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2018 - Resident Fellows Clinical Vignette Podium Presentations
California-Clinical Vignette-Podium Presentation
Caroline Chen, MD

Title: Telomerase syndrome presenting as pancytopenia in a post-lung transplant patient

Authors: Caroline Chen, MD; G. Xon Ng, MD, Department of Medicine, University of California, Los Angeles

Introduction: Pancytopenia in a post-transplant patient is commonly due to infections in the context of immunosuppression, or the immunosuppressant medications themselves. However, when these causes have been excluded, it is important to investigate less common genetic causes which may lead to a unifying diagnosis.

Case Presentation: A 52 year old former smoker presented to pulmonology clinic with a four year history of progressive dyspnea resulting in oxygen dependence. A clinical diagnosis of usual interstitial pneumonia (UIP) was made based on CT imaging. Rheumatologic work up was unrevealing. He underwent bilateral lung transplant with a course complicated by severe pancytopenias initially thought to be due to CMV viremia and his immunosuppressant medications. He was treated with anti-viral therapy and reduction of his immunosuppressant regimen without improvement. Bone marrow biopsy was performed and showed aplastic anemia. Given his history of UIP and now aplastic anemia, he was tested for telomerase syndrome which showed a lymphocyte telomere length < 1% percentile. He was started on danazol without significant clinical improvement, and was unable to undergo hematopoietic cell transplantation (HCT) due to persistent viremia. He passed away 22 months after initial presentation.

Discussion: Pancytopenia in a post-transplant patient is commonly caused by infections and medications, and less commonly by genetic disorders such as telomerase syndrome. This case demonstrates the importance of considering telomerase syndrome in patients in whom infection or medication induced pancytopenia has been ruled out, especially if they have other findings such as UIP that may be part of the underlying syndrome. Treatment options for telomerase syndrome include danazol and allogeneic HCT, the latter being the only curative option for bone marrow failure.

References

Title: If You Don't Look For It, You Won't Find It: A Case of Anomalous Right Coronary Artery Presenting As Cardiac Arrest

Authors: Stacy Tsai, MD; Stephanie Hsiao, MD; Carolyn Enders, MD; Andrew Rosenblatt, MD

Introduction: Anomalous aortic origin of a coronary artery (AAOCA) is a known and rare entity, but has clinical significance as it may lead to sudden cardiac death. This case is about a patient with an anomalous aortic origin of the right coronary artery detected on coronary CT angiography, who presented with ventricular fibrillation arrest in the setting of alcohol and cocaine use.

Case Presentation: The patient is a Laotian, 27 year old male with no past medical history, who was found down at a night club after using alcohol and cocaine. CPR was initiated by bystanders, and EMS found him in ventricular fibrillation arrest. He was defibrillated with ROSC after an unknown amount of time. At the hospital, the patient had a GCS of 3 and was intubated for airway protection. He underwent hypothermic protocol with full neurological recovery within the next 24 hours. Subsequent work-up was done to determine the cause of the cardiac arrest. EKG did not show ST/T wave changes or prolonged QTc but did show incomplete RBBB. Electrolytes were within normal limits and TTE was unremarkable. The differential diagnosis for ventricular fibrillation arrest included polysubstance use, Brugada syndrome, and anomalous coronary artery anatomy. Given the patient’s young age and his extreme presentation, it was felt that he warranted further evaluation by coronary CT angiography, which showed an anomalous aortic origin of the right coronary artery with an inter-arterial course and an intramural segment. Cardiothoracic surgery was consulted and three months after discharge, the patient underwent surgery for translocation of the right coronary artery to the right aortic sinus, completed without complications.

Discussion: The prevalence of AAOCA in the general population is difficult to ascertain and has been estimated to be 0.1-2%. Although rare, its implication is significant as it is a potential cause of sudden cardiac death. This case highlights a challenging and under-appreciated entity, which requires a high level of suspicion to diagnose as there were other possible explanations. The patient was exposed to cocaethylene, a byproduct of cocaine and alcohol, which could have led to QT prolongation ending in ventricular fibrillation arrest. Furthermore, he is of Laotian descent and his EKG was suspicious for Brugada syndrome given the incomplete RBBB. This case demonstrates the danger of anchoring on a diagnosis. The patient likely had transient compression of the anomalous right coronary artery in the inter-arterial or intramural segment in the setting of increased inotropic and chronotropic effects from cocaine use. The discovery of the anomalous right coronary artery allowed the patient to receive corrective surgery, which may reduce his risk of cardiac morbidities including sudden cardiac death.

References

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Indiana-Clinical Vignette-Podium Presentation
Kevin M Ball, MD

Title: Post CABG Infarction - Not The Kind You Might Think

Authors: Kevin Ball, MD and Laura Hampton, MD

Introduction: Failure to thrive can present a diagnostic dilemma. There are endless diagnoses that may contribute to an elderly patient’s failure to thrive. One such diagnosis that may go unrecognized is adrenal insufficiency. Adrenal insufficiency secondary to hypopituitarism can have an indolent onset, often presenting as a constellation of nonspecific systemic symptoms.

Case Presentation: Our patient is a 69 year old female with a past medical history of depression and coronary artery disease who underwent triple vessel CABG six weeks prior to admission. She was transferred to our facility from an outside hospital after presenting with profound hypoglycemia, persistent abdominal pain, diarrhea, 20 pound weight loss, nausea, vomiting, and decreased oral intake. Her symptoms began immediately after her CABG. She had undergone extensive evaluation both inpatient and outpatient by multiple providers for these complaints including specialists from gastroenterology, cardiology, psychiatry, and cardiovascular surgery. She had become quite debilitated and immobile at home, and was diagnosed with a small pulmonary embolism upon presentation. Her clinical course demonstrated persistent hypoglycemia requiring intravenous dextrose support. She had a waxing and waning mental status and was felt to be in clinical depression by the medical and psychiatry team. She previously had a history of depression and had not been taking her medications. She was unable to tolerate essentially any oral intake while in the hospital. She was started on mirtazapine and dronabinol with no improvement. Work up for reversible causes of depression included a random serum cortisol that returned as undetectable. She underwent an ACTH stimulation test that showed an inappropriate response, and her serum ACTH level was undetectable. She subsequently underwent a pituitary MRI that showed an empty sella turcica. She also had low to absent levels of LH, FSH, prolactin, and IGF-1. Interestingly, her thyroid function was preserved. She was started on steroid replacement therapy and had a dramatic improvement in symptomatology. She was able to tolerate a regular diet and her mood and mentation also improved. She eventually discharged to a skilled facility for further rehabilitation.

Discussion: Post CABG hypopituitarism has been very rarely described in case reports. The precise etiology is unknown, but it is theorized that it is related to infarction resulting from a prolonged cardiopulmonary bypass run or as the result of a previously undiagnosed pituitary adenoma that hemorrhaged during surgery. Because of its nonspecific presentation, adrenal insufficiency can often go undiagnosed for long periods of time at the expense of the patient. Adrenal insufficiency is an important, treatable diagnosis that needs to be considered in patients with failure to thrive.
Michigan-Clinical Vignette-Podium Presentation
Pranay Korpole, MBBS

Title: Injectable Naltrexone Induced Acute Eosinophilic Pneumonia

Authors: Pranay Korpole, Resident Physician, Department of Internal Medicine, St. Mary Mercy Hospital, Livonia, Michigan

Introduction: Injectable Naltrexone has the potential to become an important medication for the treatment of opiate and alcohol dependence. Acute eosinophilic pneumonia is a severe and rapidly progressive lung disease which may cause fatal respiratory failure. Eosinophilic pneumonia secondary to injectable Naltrexone use is rare. The diagnosis of acute eosinophilic pneumonia is generally overlooked given the shared clinical attributes with Acute Lung Injury due to other causes, including severe community acquired pneumonia.

Case Presentation: The patient is a 32 year old lady who presented to the Emergency Department (ED) with symptoms of dyspnea, chest pain, cough and subjective fevers since 3 days. The patient was reportedly in good health before she received a dose of intramuscular Naltrexone for treatment of substance abuse on the day of symptom onset. Patient had been smoking cigarettes consistently for the past 6 years. On arrival, she was noted to be hypoxic and oxygen supplementation was initiated via nasal cannula. Examination was pertinent for tachypnea and bilateral crackles. Patient was afebrile. While in the ED, she was placed on a non-rebreather mask because of worsening hypoxia. Chest x-ray showed diffuse bilateral pulmonary infiltrates. A CT angiogram of the chest was ordered which was negative for pulmonary embolism but showed diffuse bilateral pulmonary infiltrates as well. Initial laboratory data was pertinent for elevated WBC count with mild peripheral eosinophilia. Levofloxacin was administered for treatment of suspected community acquired pneumonia. The patient was admitted to the Medicine service and was started on intravenous steroids for management of suspected eosinophilic pneumonia secondary to injectable Naltrexone given the temporal relationship between administration of the medication and onset of symptoms. Antibiotics were discontinued upon admission. Bronchodilator therapy was initiated for wheezing. The patient’s oxygen requirements improved. Pulmonology consultation was requested and the patient underwent bronchoscopy. BAL studies showed predominance of lymphocytes with no eosinophils. Lung biopsy showed findings consistent with drug induced eosinophilic pneumonitis. Patient’s hypoxia resolved with steroid therapy. BAL cultures (including tuberculosis) remained sterile. The patient was discharged with a course of oral prednisone, albuterol inhaler and outpatient Pulmonology follow up.

Discussion: Two diagnosed and one suspected case of eosinophilic pneumonia have been reported with the use of injectable Naltrexone, however only one case report has been published revealing case details. The patient’s presentation in the previously published case report was consistent with chronic eosinophilic pneumonia while this case presentation was in keeping with acute eosinophilic pneumonia. BAL fluid eosinophil predominance, a common finding in acute eosinophilic pneumonia, was notably absent. Acute eosinophilic pneumonia should be considered as an important differential diagnosis when patients on injectable Naltrexone present with hypoxia and respiratory distress.
Title: A Fishy Case of Haff Disease

Authors: Nikhil Malhotra, MD, Saint Louis University School of Medicine, Maureen Lyons, MD, Saint Louis University School of Medicine

Introduction: Haff disease is a rarely documented illness associated with the development of rhabdomyolysis approximately 8-24 hours after consuming cooked fish. Patients often present with symptoms that may include myalgia, vomiting, dyspnea, chest pain, epigastric pain, and brown urine indicating myoglobinuria. Although Haff disease is not endemic to a specific region in the world, there have been over 1,000 cases of Haff disease globally with at least twenty-six cases of Haff disease reported in the United States from 1984-2014 with two cases in Saint Louis in 1997 and two cases in Cook County, Illinois in 2014.

Case Presentation: A 74-year-old female with a past medical history of hypertension and hyperlipidemia presented to the emergency room with persistent abdominal pain that began the morning after moderate consumption of fried Buffalo fish. Laboratory findings in the emergency room were significant for CK-Total of 3,596, CK-MB of 169.8 (ng/ml), creatinine of 1.1 and an unremarkable urine analysis. The patient’s total CK trended up to 5,714 six hours later despite aggressive intravenous hydration. The patient was admitted to the hospital for observation. A repeat CK total an hour later decreased to 4,341 and the patient was counseled on reducing her intake of buffalo fish before being discharged later that day with instructions to return to the emergency room if she develops muscle pain, increased abdominal pain, or notices dark urine. The Center for Disease Control was notified of the hospitalization for Haff disease.

Discussion: This case demonstrates the need to be aware of the association between consuming certain types of fish and the development of rhabdomyolysis. Haff disease is not specific to Buffalo fish but the fish is associated with the disease. Other implicated fish around the world include burbot (Lota lota), pike (Esox sp.) and freshwater eel (anguilla anguilla). There have been cases in the United States including Atlantic salmon, crayfish, and Buffalo fish. There are 21 similar cases in the US where samples of suspected fish or seafood were tested by the CDC and/or FDA but were negative for any known aquatic toxins. It is currently suspected that the bioaccumulation of a new heat-stable algal toxin, similar to palytoxin, but primarily myotoxic and not neurotoxic is responsible for Haff disease. While the pathophysiology of the disease process is currently unknown, it is important that these cases are documented in order to provide a foundation for further research. At the least, healthcare providers inquiring about the consumption of fish within the last 24 hours may reveal further cases of Haff disease and prevent unnecessary further testing.

References

Title: Euglycemic Diabetic Ketoacidosis: a Diagnostic Dilemma

Authors: Elixabeth Sullivan, Internal Medicine Resident, Department of Medicine, Stony Brook University Hospital, Stony Brook, NY

Introduction: Euglycemic diabetic ketoacidosis (DKA) is a constellation of history and laboratory findings associated with the use of SGLT-2 inhibitors. Its late diagnosis due to “euglycemia” makes it an extraordinary diagnostic challenge.

Case Presentation: A 59 year-old female with diabetes mellitus type 2 on metformin and canagliflozin, hyperlipidemia, and hypothyroidism presented to the emergency department with a 4 day history of generalized malaise and progressive shortness of breath associated with pressure-like chest pain that was worse after meals. The patient denied any recent fever, chills, cough, orthopnea, paroxysmal nocturnal dyspnea, abdominal pain, dysuria, diarrhea or lower extremity edema. Physical exam revealed tachycardia and tachypnea without hypoxia, Kussmaul’s breathing, but no JVD or cardiac murmurs. Laboratory Results: were significant for mildly elevated serum glucose (185 mg/dL), hypochloremia (95 mmol/L), pure anion gap metabolic acidosis (bicarbonate 8 mmol/L; ABG: 7.11/25/52/8), and elevated beta hydroxybutirate (9.57 mmol/L). Urinalysis was positive for glycosuria and moderate ketones. After some initial confusion due to the patient’s lack of significant serum glucose elevation, the patient’s medications were reviewed and she was diagnosed with severe euglycemic DKA secondary to canagliflozin use. The patient was emergently fluid resuscitated, was started on insulin drip, and was admitted to the medical intensive care unit for close monitoring.

By treating the aforementioned condition with insulin drip, the patient’s anion gap corrected appropriately. The patient was started on bicarbonate drip for persistent acidosis (bicarbonate <6 mmol/L for 12 hours). Once the patient’s serum glucose level normalized and the acidosis resolved her symptoms improved. She did continue to have glycosuria for ten more days. The patient was discharged home on insulin glargine and insulin lispro. The patient was instructed not to resume canagliflozin or any other SGLT-2 inhibitors.

Discussion: This case illustrates the importance of systematically obtaining a complete history, including a patient’s medications, understanding the mechanism of action of novel medications, and not prematurely excluding a diagnosis based on commonly expected laboratory data.
Title: Small or Big, Lupus is Lupus

Authors: Moon Kyung Choi, Michael Viray

Introduction: Discoid lupus erythematosus (DLE) is often considered as mild lupus with limited systemic involvement and good prognosis. Most cases are managed with photo protection and topical steroids. Here, we present a case of serious cardiovascular complication from DLE in an otherwise healthy young female.

Case Presentation: A 35 year-old African American female with past medical history of DLE and recurrent miscarriages presented with sudden-onset of 10/10 substernal, pressure-like pain with no alleviating factors. She was hemodynamically stable on presentation. Initial 12 lead-EKG revealed inferior ST elevation with reciprocal ST depression in the lateral leads. Emergent coronary angiography revealed a total occlusion of the proximal right coronary artery and an 80% lesion of the posterior descending artery. She subsequently underwent percutaneous transluminal coronary angioplasty with drug eluting stents placed in the RCA and PDA. In regard to her DLE, the patient presented with multiple atrophic scars and rash on her face without mucosal involvement. She denied oral ulcers, photosensitivity, joint swelling or any history of serositis. Laboratory evaluation of ANA, anti-ds DNA, anti-Sm antibody and lupus anticoagulant was negative. Coronary artery vasculitis was deemed less likely in the setting of normal C3, C4 complement levels and the absence of suggestive findings on angiography.

Discussion: DLE is the most common type of chronic cutaneous lupus erythematosus (CLE) characterized by discrete, erythematous plaques on the face with atrophy and depigmentation. Only 5-10% of localized DLE patients have co-existing systemic lupus erythematosus (SLE). Premature atherosclerotic coronary artery disease (CAD) is a well-known complication of SLE. Though there is less data regarding CAD risk in CLE, recent studies show that CLE patients also have increased mortality from cardiovascular disease and increased risk of metabolic syndrome. This young female did not have traditional risk factors such as hypertension or a family history of CAD. However, she had uncontrolled DLE not on medication, and was a current smoker with increased LDL (163) and marginal HDL (49). This case underscores the importance of aggressive risk factor modification for cardiovascular disease in CLE, even with seemingly ‘localized’ discoid lupus.
Title: Anti-NMDA-Receptor Encephalitis in a Patient with Radiographically Occult Ovarian Teratoma: A case report and Discussion of management strategies

Authors: Madeline Eckenrode, M.D.

Introduction: The purpose of this case presentation is to:

Review the presentation and treatment of anti-NMDA-receptor encephalitis for internal medicine physicians

Discuss a case of anti-NMDA-receptor encephalitis associated with occult ovarian teratoma and to propose early oophorectomy as a treatment strategy

Case Presentation: A healthy 24-year-old African-American female presented to an emergency department with several hours of visual hallucinations, fever, and agitation. During the week prior to presentation, she had headache, memory loss, and confusion. Work-up was notable only for elevated WBC count in her CSF. She was treated for meningitis with appropriate antimicrobials, but became unresponsive and began having orolingual dyskinesia, rhythmic hand jerking, and generalized seizures. She was transferred to our institution after one week. Differential included viral and autoimmune encephalitis. Another LP was performed; arbovirus and autoimmune encephalitis panels were sent. Brain MRI was unremarkable. Given the high suspicion for an autoimmune process, IVIG was initiated. She also received plasma exchange. Eventually, she was found to have anti-NMDA-receptor encephalitis. Because she had two normal abdominal CT scans, concern for ovarian neoplasm was low. A pelvic ultrasound revealed what was thought to be a cyst in the right ovary. Five weeks after presentation, due to her lack of improvement, she had a right salpingoophorectomy. Pathology showed a mature ovarian teratoma that stained positive for S-100 and GFAP. Despite treatment and teratoma resection, the patient died.

Discussion: Anti-NMDA-receptor encephalitis typically presents with non-specific symptoms such as fever and headache and progresses to cause hallucinations, seizures and motor dysfunction, autonomic instability, and catatonia. It is caused by autoantibodies to the NMDA receptor (involved in memory/neuronal plasticity).

About 80% of anti-NMDA-R encephalitis patients are women. In post-pubescent women, the likelihood that a tumor will be present is about 60%. These are often ovarian teratomas, which contain neural tissue that triggers an immune response. The diagnosis of anti-NMDA-R encephalitis should be considered in young patients who present with behavioral abnormalities or speech/motor disturbances that progress to seizures/alteration in cognition and functioning.

Diagnosis involves detection of antibodies in the CSF or serum. This is currently the only reliable means of diagnosis; CT and MRI of the brain can be normal. CSF may show a lymphocytic pleocytosis. Patients with suspected NMDA-R encephalitis should have abdominal/pelvic imaging to assess for the presence of malignancy.
IVIG and steroids are considered first line therapy. After treatment, patients can return to pre-morbid levels of functionality, although many patients continue to experience memory dysfunction/difficulties with executive functioning (25% die or remain neurologically impaired).

Black females are the most likely of any patient population to have co-existent ovarian teratoma. Males/younger patients are less likely to have tumors. Case reports exist of teratomas that were detectable only microscopically. Because patients with anti-NMDA-R encephalitis and ovarian teratomas usually experience improvement after resection, it is worth considering oophorectomy even without radiographic evidence of tumor.

References

Title: Atraumatic splenic rupture associated with apixaban

Authors: Elan Mohanty MD, Izza Mir DO, Sijan Basnet MD, Ajay Koirala MD, Niranjan Tachamo MD, Anthony Donato, MD

Introduction: Atraumatic splenic rupture is a rare but life-threatening condition requiring urgent intervention. Anticoagulants have been associated with this condition in the past. We report a case of atraumatic splenic rupture in a patient on apixaban presenting with syncope.

Case Presentation: An 86-year-old man with a history of atrial fibrillation on apixaban and aspirin for coronary artery disease presented with orthostatic syncope leading to a fall. Patient had complained of left upper quadrant abdominal pain the night prior to the fall. Patient denied trauma prior to the onset of abdominal pain, recent sore throat, fever, chills, night sweats, weight loss, fatigue, reflux, or recent travel. On presentation, blood pressure was 107/67 mm Hg and heart rate 65/minute. Physical examination was significant for diffuse bilateral abdominal tenderness without any guarding or rebound tenderness. Hemoglobin was 11.8 g/dl, down from a baseline of 14 g/dl. Coagulation panel revealed INR 1.2 (normal: 0.9-1.1), PT 14.9 seconds (normal: 11.7-14.1 seconds), PTT 28 seconds (normal: 23-34 seconds) and platelets 146,000/ul (normal: 130,000-400,000/ul). CT abdomen/pelvis with contrast showed active extravasation from the spleen with a large subcapsular hematoma with hemoperitoneum. He was treated with prothrombin complex concentrate, 2 units packed red cells, and splenic artery embolization. Despite this, several hours later the patient became acutely hypotensive. Ultrasound at bedside showed large hemoperitoneum. Patient underwent urgent exploratory laparotomy with splenectomy. Splenic pathology revealed capsular disruption with associated sub-capsular and intra-parenchymal hemorrhage, and numerous macrophages within the splenic parenchyma likely secondary to a reactive phenomenon. The patient had a good recovery and his apixaban was resumed upon discharge.

Discussion: Atraumatic splenic rupture has been reported in patients on anticoagulants like streptokinase, heparin and recently even with apixaban. While the pathophysiology remains unclear, some hypothesize that a spleen with prior microtrauma may rupture when patient is on anticoagulation. With increasing use of novel anticoagulants in patients with atrial fibrillation, this association might gain further prominence.
Title: The Challenge of Recognizing JAK Inhibitor Withdrawal Syndrome

Authors: Michael Pierro1, Arun K Singavi2, Laura Michaelis2, 1DoM, Medical College of Wisconsin, Milwaukee, WI 2Division of Hematology/Oncology, Medical College of Wisconsin, Milwaukee, WI

Introduction: Janus Associated Kinase (JAK) inhibitors have been proven to effectively manage symptoms of myelofibrosis. However, upon discontinuation of these medications, some patients can experience an acute and severe relapse of symptoms. This syndrome can mimic sepsis or a systemic inflammatory response syndrome (SIRS). The infrequency with which JAK inhibitors are encountered makes recognizing this syndrome a particular challenge.

Case Presentation: A 41 year old female was admitted to the hospital with 5-6 days of nausea, vomiting, fevers, and drenching night sweats. She has a history of polycythemia vera diagnosed at age 20, which progressed to myelofibrosis at age 37. Her vital signs were significant for tachycardia and a temperature of 102.8. She briefly required supplemental oxygen secondary to hypoxia. Physical exam was remarkable for marked splenomegaly. Laboratory evaluation noted severe anemia and thrombocytopenia, worse from her baseline values. A CT scan of her abdomen demonstrated marked splenomegaly to 32 cm with possible early infarction. There was concern for sepsis especially in her underlying immunocompromised state, so she was started on broad spectrum antibiotics. Additional history revealed she had been treated with a JAK2 inhibitor for the previous 4 years, but treatment had been stopped the day prior to onset of symptoms secondary to disease progression. Hematology was consulted and she was started on high dose corticosteroids. On hospital day (HD) two, her spleen size increased on exam and the patient was experiencing worsening abdominal pain. Thereby, her corticosteroid dose was increased. A repeat CT showed stable splenomegaly. On HD three, she was urgently restarted on a JAK2 inhibitor and began a tapered course of corticosteroids. Over the next week, her symptoms of nausea, vomiting, abdominal pain, and fevers improved. Her blood cultures remained negative and thus antibiotics were discontinued on HD 5. She was discharged on HD 6 with resolution of symptoms and a taper of JAK2 inhibitor.

Discussion: JAK inhibitor withdrawal syndrome is characterized by an acute relapse of myelofibrosis symptoms including rapidly worsening cytopenias and splenomegaly, and at times hemodynamic compromise that can present very similar to sepsis or septic shock. Importantly, this syndrome has been described in patients being treated with JAK inhibitors for myelofibrosis, but not in patients being treated with JAK inhibitors for other conditions. This case illustrates the importance of early recognition for this severe withdrawal syndrome to avoid poor outcomes. Maintaining a wide differential diagnosis and obtaining a thorough history in patients with known hematologic malignancies is central to making the correct diagnosis. Treatment includes initiating high dose corticosteroids and restarting the JAK2 inhibitor. Had it not been for the prompt diagnosis and treatment of this withdrawal syndrome, our patient could have had a quickly deteriorating clinical condition and much worse outcome.

References

2018 - Resident Fellows Clinical Vignette Poster Finalists
Title: Doxycycline Induced Pseudoporphyria Masquerading as Porphyria Cutanea Tarda

Authors: Page Axley, Department of Internal Medicine, University of Alabama at Birmingham School of Medicine, Ashwani Singal, Division of Gastroenterology and Hepatology, University of Alabama at Birmingham School of Medicine

Introduction: Pseudoporphyria is a rare photodermatitis characterized by skin fragility and cutaneous bullae, with distinct clinical and histologic features. Medications including antibiotics and nonsteroidal anti-inflammatory agents have been linked to this disease. The diagnosis of pseudoporphyria, which does not result from enzymatic absence, should be considered in patients with photosensitivity and skin blistering. We describe here an interesting case of doxycycline-induced pseudoporphyria.

Case Presentation: 28 year-old white female reported recurrent episodes of bullous lesions on sun-exposed areas for the past 3 years. Her past medical history was pertinent for gastroesophageal reflux disease and acne, for which she was taking pantoprazole and doxycycline. There was no history of liver disease, joint pain or rheumatic conditions, chronic kidney disease, or immunosuppression. She was a prior smoker and drinks 4-5 alcoholic beverages weekly. She originally presented to an outside dermatologist, and punch biopsy from left finger showed subtle dermal fibrosis with necrotic keratinocytes and hyalinized blood vessels (Figure 1A). Another punch biopsy showed deposits of IgG and C3 around blood vessels. She was started on hydroxychloroquine 200mg daily without lesion improvement. At the time of presentation to us, the patient complained of a persistent extensive bright erythematosus plaque-like lesions on the dorsal aspect of her hands and feet, forearms, and face (Figure 1B). Routine laboratory work including testing for hepatitis C and HIV infection was negative. Serum iron and ferritin levels were normal. Genetic testing for haemochromatosis (HFE) gene and uroporphyrinogen decarboxylase (UROD) mutations were also negative. Biochemical porphyrins profile showed normal total urine and plasma porphyrins with values of 99 nmol/L and 0.1 mcg/dl, respectively. She was referred back to her dermatologist for suspected doxycycline-induced pseudoporphyria with reported improvement in skin lesions once doxycycline was discontinued.

Discussion: Photosensitivity with skin blistering, a classic presenting sign of porphyria cutanea tarda, is not exclusive to the disease. Pseudoporphyria is a rare photo distributed bullous disorder with the clinical hallmarks of porphyria cutanea tarda including characteristic findings on skin biopsy, but without accompanying biochemical porphyrin abnormalities. As opposed to porphyria cutanea tarda, hyperpigmentation, hypertrichosis and sclerodermoid changes are rarely seen in pseudoporphyria. Initially described in patients with renal failure on dialysis, pseudoporphyria has been associated with numerous photosensitizing medications, hormone replacement, chlorophyll and UVA exposure, and autoimmune disorders. Nonsteroidal anti-inflammatory agents, particularly naproxen, are most frequently implicated in the development of drug-induced pseudoporphyria. Treatment consists of UV protection and cessation of the offending agent which commonly leads to resolution of symptoms in weeks to months, as was observed in our case.

References
Alabama-Clinical Vignette-Poster Finalist
Heather L Fishel, MD

Title: Whipping the Bowel

Authors: Heather Fishel, M.D., Leonel Maldonadom M.D., Caitlin Prickett, D.O., and Philip Almalouf, M.D.

Introduction: Whipple's disease (WD) is a rare infection caused by the bacterium Tropheryma whipplei that can affect multiple organs and commonly occurs in the immunocompetent host. T. whipplei occurs in the environment, is prevalent only in humans, and believed to be transmitted via oral routes and to be host dependent. It has four cardinal manifestations: arthralgias, weight loss, diarrhea, and abdominal pain. It may mimic chronic inflammatory diseases, and the diagnosis remains challenging.

Case Presentation: A 57 year old white female with a self-diagnosis of celiac’s disease, rheumatoid arthritis, multiple transient ischemic attacks, atrial fibrillation and fibromyalgia presented for generalized weakness with 150 pounds weight loss over a year’s time, chronic diarrhea, nausea, and abdominal pain. Physical exam revealed pitting edema of the extremities, generalized muscle atrophy, otherwise unremarkable. Initial workup revealed hypokalemia, normal thyroid studies, negative HIV and hepatitis panel. Stool studies for clostridium difficile, ova and parasites were negative, but a stool osmol gap indicated an osmotic diarrhea. Her diarrhea resolved as she was kept NPO for 24 hours. Patient had a flex sigmoidoscopy with sigmoid and rectal biopsies. Pathology showed inflammation with mild architectural distortion. Pancreatic insufficiency work up was negative with unremarkable pancreatic elastase level. Testing for heavy metal poisoning was negative and measurement of vitamins and minerals indicated multiple deficiencies. Antibody and antigen testings for celiac’s disease were negative. However, CT imaging of the abdomen noted abnormal appearance of the ileum with lymphadenopathy which could be consistent with celiac’s disease. Upper endoscopy was performed due to concerns for malabsorption versus an autoimmune enteropathy. EGD biopsies reported findings of PAS-positive rods in macrophages, consistent with Tropheryma whipplei. Patient was initiated on Ceftriaxone with marked improvement in her abdominal pain and resolution of her diarrhea. She was transitioned after 2 weeks to oral trimethoprim/sulfamethoxazole with outpatient follow up, and she continued to improve.

Discussion: The clinical manifestations of Whipple’s disease are nonspecific and easy to misdiagnose as a sequelae of an autoimmune process. Our patient had all classic symptoms of intermittent arthralgia, chronic diarrhea, abdominal pain and weight loss. The disease is caused by infection with T. whipplei, a bacterium that may be more common than was initially assumed as routine cultivation is not feasible. Recent advances in medical microbiology, immunohistochemistry, and PCR have improved our understanding of the clinical range and natural course. Endoscopic tests are key as they allow histopathological examination for definitive diagnosis. Periodic acid-Schiff (PAS) staining of duodenal biopsy specimens remains the gold standard, and it was positive in our patient confirming the diagnosis. Whipple's disease, although rare, is an entity that should be included in the differential diagnosis when evaluating chronic diarrhea, as its progression may be fatal if left untreated.
Alabama-Clinical Vignette-Poster Finalist
Nicholas R Ludvik, MD

Title: A case of abdominal pain warranting a nephrology consultation

Authors: Nicholas Ludvik MD, Philip Almalouf, MD

Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary form of kidney disease and exhibits diverse clinical presentations. It is attributed as the causation of up to 10% of end stage renal disease (ESRD) cases and is the 4th most common cause for renal replacement therapy. It might have extrarenal manifestations such as polycystic liver disease, thus it represents an important diagnosis to consider as early intervention is critical in optimizing outcomes.

Case Presentation: A 44-year-old African-American female, with past medical history of polycystic kidney diagnosed incidentally a decade ago during workup for nephrolithiasis, presented with 2 weeks of progressive right flank pain and accompanying fatigue. Pain was described as constant, diffuse, progressive, and associated with nausea and vomiting. Admitted to chronic right sided abdominal discomfort for which she sought no medical care. Had attempted taking ibuprofen without relief. Her mother and daughter had PKD. Physical exam was notable for BP 131/80, HR 93, afebrile, absence of jaundice, abdomen diffusely tender to palpation with numerous masses visualized and palpated along abdominal wall. Labs revealed creatinine 0.9, INR 1.14, bilirubin 1.1, alkaline phosphatase 301, and normal liver enzymes. CT of abdomen revealed innumerable massive cystic lesions in the liver extending into right pelvis. MRI of abdomen revealed an acute hemorrhagic hepatic cyst as well as complex hepatic cysts with signal alteration consistent with old blood products suggestive of prior hemorrhages. Patient was treated conservatively with improvement symptomatic improvement over a few days. Outpatient follow up was arranged to undergo partial hepatectomy to alleviate symptoms.

Discussion: Autosomal dominant polycystic kidney disease has a considerable variability of clinical manifestations, ranging from a few asymptomatic cysts to innumerable cysts causing severe hypertension ultimately leading to ESRD. Hepatic involvement has been reported, primarily in women, and was an unexpected feature of this case. Polycystic liver disease is characterized by multiple cystic lesions that range from 20 to 30 cm to small microscopic nodules. The mass effect of the liver cysts can cause life-threatening symptoms such as reduction of oral intake, weight loss, and malnutrition. Here, we present a case of chronic abdominal pain caused by hepatomegaly resulting from ADPKD and acute worsening caused by cystic hemorrhage which was confirmed on abdominal MRI. Liver transplantation is the only curative option, and preserved for severely symptomatic patients and advanced liver failure. Partial hepatectomy can be considered to alleviate mass effect and prevent recurrence of cystic rupture. That was recommended in our patient as her liver function was preserved. Given the relatively common nature of ADPKD, physicians should be aware of its various clinical presentations to provide better outcomes.
Title: When They Don’t Hear The Noise, Think Centipoise

Authors: Salam Mohammed Arif MD, Brenda Shinar MD, Richard Gerkin MD

Introduction: Waldenström macroglobulinemia (WM) is a lymphoproliferative disease, characterized by bone marrow infiltration with lymphoplasmacytic lymphoma capable of producing high serum levels of circulating monoclonal IgM that usually manifest clinically with neurologic deficits related to increased serum viscosity (Hyperviscosity syndrome).

We are presenting here a case of an exceptionally high serum viscosity secondary to WM, only presented with bilateral sensorineural hearing loss.

Case Presentation: A 63-year-old woman presents with progressive, severe bilateral hearing loss over the course of two months with complete loss of hearing over the week prior to presentation. She has associated imbalance and mild generalized fatigue. She has not received medical care for many years prior to this admission. She denies head trauma, prolonged exposure to noise or ototoxins, tinnitus, fever, ear pain, or sinus congestion. She takes no medications. Review of systems is otherwise negative. Physical exam confirms bilateral sensorineural hearing loss. Laboratory testing shows lymphocytic predominance of leukocytes, and mild anemia and thrombocytopenia. Comprehensive metabolic panel showed Cr 1.38, as well as a large protein-albumin gap (serum total protein 9g/dl and albumin 2g/dl). CT/CTA and MRI of the brain were normal.

Further diagnostic testing includes serum protein electrophoresis and Immunofixation assay demonstrating IgM 8728 and serum viscosity 27.3 centipoises (cP) (normal <1.5 cP). Bone marrow biopsy confirms lymphoplasmacytic lymphoma consistent with Waldenström macroglobulinemia (WM). The patient receives 5 days of continuous total plasmapheresis for hyperviscosity syndrome and initiates chemotherapy for B cell lymphoma. Despite reduction of serum viscosity to near normal, the patient does not regain hearing during her hospitalization.

Discussion: Waldenström macroglobulinemia (WM), first described by Jan Waldenström in 1944, is a lymphoproliferative lymphoma characterized by bone marrow infiltration by lymphoplasmacytic lymphoma cells producing high levels of circulating monoclonal IgM. The large size and pentameric configuration of the IgM molecules increase serum viscosity and impairs blood flow through the microcirculatory system. Hyperviscosity syndrome (HVS) occur in up to 30% of patients with WM, when the serum viscosity rises above 4 cP. The level of serum viscosity in this patient is among the highest reported in the literature.

Typical neurologic manifestations of HVS include visual disturbances, headache, ataxia, vertigo, tinnitus, and rarely seizure and coma. Of eleven published case reports of WM presenting with some degree of sensorineural hearing loss, we believe that this case represents the third in which bilateral sensorineural hearing loss is the only presenting symptom.

Though rare, hyperviscosity syndrome can have potentially devastating consequences if not recognized and treated immediately. Current treatment for HVS is plasmapheresis, the most effective method of removing IgM from circulation. The very high level of viscosity at presentation combined with the
duration of symptoms prior to seeking medical care likely contributed to the persistence of her symptoms despite aggressive therapy.

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Arizona-Clinical Vignette-Poster Finalist
Shilpa Junna, MD

Title: *Stenotrophomonas maltophilia*-A Rare Cause of Recurrent Necrotizing Pancreatitis

Authors: Shilpa Junna, MD1; Todd Golden, MS2; Sehem Ghazala, MD1; Vaishnavi Veerapaneni, MBBS3; Rita Wadeea, MD1; Mayar Al Mohajer, MD MBA4. 1. University of Arizona Medical Center, Department of Internal Medicine, Tucson, Arizona. 2. University of Arizona School of Medicine, Tucson, Arizona. 3. Jawaharlal Nehru Medical College, Belgaum, India. 4. Baylor College of Medicine Medical Center, Department of Infectious Disease, Houston, Texas

Introduction: *Stenotrophomonas maltophilia* is an aerobic Gram-negative bacterium that is well known for its capacity to produce biofilms. It has recently been implicated as an emerging nosocomial pathogen in cases of pneumonia, endocarditis, and septic arthritis, particularly in patients that are immunosuppressed or admitted to intensive care units. *S. maltophilia* is also known for its resistance to multiple conventional pharmacological therapies, including some cephalosporins and carbapenems. We present one of the few cases of *S. maltophilia* causing necrotizing pancreatitis.

Case Presentation: Our case involves a 73-year-old male who presented from an outside facility after undergoing a protracted five-week course of severe pancreatitis complicated by subsequent sepsis, acute hypoxic respiratory failure, and pancreatic pseudocyst formation. Due to a decline in clinical status, magnetic resonance imaging of the abdomen was obtained, which noted two enlarging pancreatic pseudocysts and new areas of pancreatic necrosis causing gastric outlet obstruction. Blood cultures were obtained and showed no evidence of bacterial growth, however endoscopic drainage of one of the pancreatic pseudocysts yielded *Enterobacter cloacae, Enterococcus faecium, and S. maltophilia*. The patient underwent recurrent endoscopic ultrasonography with debridement of necrotic tissue in attempts to relieve the obstruction, along with cystogastrostomy stent placement. The patient was initially treated with broad spectrum antibiotics including vancomycin and meropenem, and then transitioned to a regimen of linezolid and trimethoprim/sulfamethoxazole. Despite aggressive antibiotic therapy and necrotic tissue debridement, the patient did not survive.

Discussion: Recent cases have noted increasing prevalence of *S. maltophilia* in bloodstream and hepatobiliary infections, thereby increasing the possibility that it may become a common causative organism of necrotizing pancreatitis in the future. This case illustrates the importance of continued research regarding appropriate antibiotic coverage for necrotizing pancreatitis and possible need for early initiation of targeted antibiotics, as *S. maltophilia* becomes more prevalent.

References

Title: Emerging Considerations in Management of Hepatic Abscesses

Authors: Shilpa Junna, MD1; Todd Rabkin Golden, MS2; Kris Kumar, DO1; Laura Meinke, MD1; Meenakshi Dagar, MD1, 1. University of Arizona Medical Center, Tucson, Arizona, 2. University of Arizona School of Medicine, Tucson, Arizona

Introduction: *Fusobacterium nucleatum* is an anaerobic organism that has been occasionally associated with hepatic abscesses, and more recently implicated in tumorigenesis pathways in patients with colorectal carcinoma. We present one of the few cases of *F. nucleatum* causing a hepatic abscess and subsequent sigmoid diverticulitis in an asymptomatic patient.

Case Presentation: A 47-year-old male presented with a two-week history of progressive dyspnea on exertion after recent completion of treatment for community-acquired pneumonia. He had a history of type 2 diabetes mellitus, hyperlipidemia, and benign essential hypertension, and denied use of immunosuppressive medications. Chest computerized tomography incidentally noted a 6.2 x 6.7 cm ill-defined hypodense area in the liver, and magnetic resonance imaging of the abdomen confirmed this diagnosis. The patient underwent minimally invasive drainage of the abscess, with cultures noting *F. nucleatum*. Treatment was administered with a brief course of oral metronidazole and the abscess was monitored via serial abdominal imaging. Ten days after drainage, computer tomography revealed evidence of moderate sigmoid diverticulitis that had not been visualized previously, and continued extension of diverticular disease into the descending colon was observed on subsequent imaging studies over the next several months. Throughout his course, the patient denied abdominal symptomatology, and had conservative medical management of his diverticulitis with a regimen of ceftriaxone and metronidazole, to complete an eight-week course with no further complications.

Discussion: The association between *F. nucleatum* and diverticulitis has been infrequently observed, with only a few cases documented in the literature. These cases have primarily been noted in patients with severely immunocompromised states, periodontal disease, or old age. Our patient’s case demonstrates an atypical presentation of this rare infection, and emphasizes the importance of obtaining serial abdominal imaging in patients who present with *F. nucleatum* associated hepatic abscesses in order to evaluate for coexisting diverticulitis, even in the absence of abdominal symptomatology. Moreover, the increasing association of *F. nucleatum* with colorectal carcinoma demonstrates the additional significance of providing closer monitoring of patients infected with this organism.

References

Arizona-Clinical Vignette-Poster Finalist
Supreet Khare, MBBS

Title: A Perfect Camouflage

Authors: Supreet Khare, MD

Introduction: Cushing’s syndrome is a rare disorder of cortisol excess due to an adrenal tumor or prolonged exposure to cortisol. Earlier diagnosis of Cushing’s syndrome is associated with decreased morbidity and mortality. Often the clinical picture is highly variable making the diagnosis difficult.

Case Presentation: We present a case of 55 y.o Hispanic female with h/o recent onset DM and uncontrolled Hypertension who presented to us with progressively worsening bilateral lower extremity edema over the past month associated with numbness and tingling. Furosemide was started with pending evaluation of her kidney, liver, heart, and DVT due to venous stasis ulcers. On detailed interviewing, it was evident that her problems started almost two years ago after she had a breakdown and was going through job stress. She was diagnosed and treated for DM and Hyperthyroidism by her PCP for her symptoms which did not improve and the reason was assumed to be her non-compliance with medications. The patient began having disturbed sleep, fatigue, became depressed with extreme mood swings, gained a lot of weight, started losing hairs and had virilism with deepening of the voice.

Physical examination revealed uncontrolled HTN (198/110), prominent supraclavicular and dorsal-cervical fat pads with dry coarse thick skin, exophthalmos, central obesity with violaceous striae, diffuse ecchymosis, with poor memory and venous ulcers.

Initial diagnostic studies were notable for hyperglycemia, leukocytosis & hypokalemia. Based on her history and physical findings, Cushing’s syndrome was suspected. Further, dexamethasone suppression test with urine Cortisol levels and Metanephrines were ordered which showed high Cortisol levels of 37.8 µ/dl and normal Metanephrines which confirmed the diagnosis of ACTH independent Cushing’s Syndrome. MRI abdomen showed 5.8x3.8 x 5.7 cm lipid-rich right adrenal adenoma. The patient subsequently underwent laparoscopic right adrenalectomy; she was discharged and was followed up in Endocrinology clinic.

Discussion: The incidence of Cushing’s Syndrome is 10-15/million people/year in the US. Often found in the age group of 20-50 yrs with more than 70% females being affected. Diagnosis of Cushing’s Syndrome is challenging and is made by a combination of patients history and examination findings, imaging and laboratory Results: A further cause of Cushing’s Syndrome can be identified by measuring the ACTH levels. A multidisciplinary approach with long-term follow-up is required to keep an eye on the comorbidities associated with Cushing’s Syndrome. The treatment modalities should be individualized according to each individual essential for optimal control of hypercortisolemia and in decreasing morbidity and mortality associated with Cushing’s Syndrome. The present case stresses upon and establishes the importance of detailed history taking and physical examination in a patient presenting with the persistent hyperglycemia and hypertension and to consider Cushing’s syndrome as a differential in such cases.
Arkansas-Clinical Vignette-Poster Finalist
Aneesha Ananthula, MD

Title: PRIMARY CARDIAC SARCOMA PRESENTING AS BILATERAL FLANK PAIN

Authors: Aneesha Ananthula - Resident, Department of Internal Medicine, UAMS, Manogna Konda - Resident, Department of Internal Medicine, UAMS, Dinesh Atwal - Resident, Department of Internal Medicine, UAMS, Rangaswami Govindarajan - Professor, Division of Oncology, UAMS

Introduction: Primary cardiac tumors are extremely rare with a very poor prognosis, with an incidence ranging from 0.0017% to 0.28%. They typically present with cardiac symptoms that may include symptoms of heart failure, obstruction to blood flow and arrhythmias. We report a unique case of a patient with cardiac sarcoma who presented with flank pain due to renal infarcts secondary to embolization without any preceding cardiac symptoms.

Case Presentation: A 52-year-old African-American male presented with complaints of bilateral flank pain for a period of one month. Computed Tomography (CT) of abdomen and pelvis was obtained which was concerning for renal infarcts and enlargement of bilateral adrenal glands. Subsequently, a transesophageal echocardiography was obtained to look for potential embolic source. The images showed a thickened mitral valve with moderate regurgitation and a complex large echo density 5x3cm partly sessile and partly mobile in the left atrium, which was most likely a thrombus or a mass. A cardiac magnetic resonance imaging (MRI) was performed which showed a non enhancing soft tissue density most consistent with thrombus in the left atrium with no infiltration. He subsequently underwent surgery for the removal of the presumed thrombus given the risk of embolic stroke. Intraoperative findings showed that the walls of the left atrium were lined entirely with abnormal white fibrous tissue extending into the mitral valve as well as 1 – 2 cm along each pulmonary vein. Histology and Immunohistochemistry staining done suggested a diagnosis of undifferentiated pleomorphic sarcoma. During the diagnostic work-up, the patient developed disease progression with brain mets, new mediastinal and retroperitoneal lymphadenopathy, new liver lesions and progression of adrenal metastases. Patient subsequently passed away, approximately 10 months after the time of initial diagnosis.

Discussion: Angiosarcoma is the most common, while undifferentiated pleomorphic sarcomas were comparatively uncommon, as evidenced by several large series. The patients with cardiac sarcoma have a poor prognosis due to rapid proliferation of the tumor and distant metastases are often found at the time of initial diagnosis. The only mode of therapy that has been shown to prolong survival is complete surgical excision. A retrospective series spanning 25 years at the Cleveland Clinic found that the patients who received multimodality therapy (any combination of surgery, chemotherapy or radiation therapy) had a survival advantage over those who were treated with surgery, radiation therapy or chemotherapy alone. However, there is currently no consensus on the ideal timing or duration of adjuvant radiation and/or chemotherapy. The median survival rates of these patients is less than 1 year despite aggressive adjuvant chemotherapy after resection.

References
Arkansas-Clinical Vignette-Poster Finalist
Anthony Kunnumpurath, MBBS MD

Title: Chronic myelomonocytic leukemia presenting with seropositivity for rheumatoid arthritis and polyserositis

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Introduction: Chronic myelomonocytic leukemia (CMML) is a rare clonal hematopoietic stem cell disorder associated with clinical and pathologic features of myelodysplasia and myeloproliferation. Systemic autoimmune/inflammatory disorders (SAID) and polyserositis have been associated with CMML. These manifestations can be observed concomitantly, shortly before diagnosis or anytime along the course of illness. We would like to present a case of myeloproliferative CMML who presented with polyserositis and positive serology for rheumatoid arthritis.

Case Presentation: A 65 year old white female was initially evaluated in ER for right upper quadrant pain; imaging studies ruled out cholecystitis. She was treated with parenteral narcotics in short stay. WBC was elevated at 12 x10⁹/µL. Absolute monocyte count was also elevated 3.3 x 10³/µL. She was admitted 2 weeks later with dyspnea and chest discomfort. CT scan revealed bilateral pleural effusion and pericardial effusion. WBC was further elevated at 33.4 x10³/µL. Rheumatoid factor was positive (54.9 IU) and anti CCP antibody was also positive; however there was no evidence of arthritis. Pleural fluid was exudative and showed monocytic infiltrate. Bone marrow biopsy was consistent with CMML; myeloblasts were not increased. She was sent home on tapering dose of Prednisone. After 2 weeks she was initiated on Azacitidine (Vidaza); on initiation her white count was 143 x10⁹/µL with 78% monocytes. Bone marrow evaluation after 5 cycles revealed transformation to acute myeloid leukemia and cytogenetic studies revealed 46 XX, deletion (9 q13 – q 22) in 20 cells. Molecular studies revealed mutated NPM gene. She underwent remission induction following which she is on Decitabine maintenance. Allogenic stem cell transplant is being planned. She did not have any recurrence of serositis related symptom.

Discussion: CMML and autoimmunity is increasingly recognized while pathogenic mechanisms are barely studied. The increased cytokine production by monocytes include TNF- α, IL-6, (Interferon regulatory factor -1) trigger polyclonal B-lymphocyte proliferation, antibodies production, abnormal antigen presentation and global deregulation of immune response. Polyserositis is commonly associated with a C reactive protein, which suggest the involvement of IL-6 in the pathophysiology. Autoimmune manifestations does not alter the outcome in CMML.
Retrospective studies of MDS/CMML have reported 15 to 25% incidence of SAID. The most frequently observed disorders include systemic vasculitis, connective tissue diseases, polychondritis, seronegative arthritis and immune thrombocytopenia. SAID does not confer adverse prognosis in retrospective studies. Polyserositis is less common; this may result from leukemic infiltrate or result from autoimmunity. Treatment of serositis includes steroids and cytoreductive agents. Serositis may confer poor prognosis and hypomethylating therapy may improve the outcome.

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Title: A case of Cholesteryl Ester Storage Disease (CESD) - a rare storage disease.

Authors: Dr. Madhabi Karmaker, Dr. Ahmedul Kabir, Professor Dr. Khan Abul Kalam Azad, Professor Dr. Abid Hossain Mollah, Department of medicine, Dhaka medical College hospital, Dhaka, Bangladesh.

Introduction: Cholesteryl Ester Storage Disease (CESD) is a rare genetic disease characterized by accumulation of lipid material in many tissues due to deficiency of Lysosomal Acid Lipase (LAL/LIPA) enzyme. It is inherited as an autosomal recessive condition. (1) CESD affects males and females in equal numbers. (2) The symptoms and severity of CESD are highly variable and usually start in childhood with vomiting, diarrhea, nutritional deficiency and failure to thrive. Alterations of blood lipid profile, hepatomegaly, splenomegaly or adrenalomegaly may also be found. (3) In 2015 FDA (USA) approved Kanuma (Sebelipase alfa) as first treatment for LAL deficiency. (4) A hypolipidaemic diet and lipid lowering agents are other therapeutic tools against CESD. (5)

Case Presentation: 17 year-old-girl presented with episodic abnormal behaviour followed by disorientation and generalized convulsion for 6 years. She also had frequent vomiting, frequent passage of stool and failure to thrive since childhood with primary amenorrhoea. She had history of jaundice 10 years back. There was consanguinity of marriage between her parents and one brother died of liver disease at age six.

She was short statured having height below 3rd percentile with BMI 15 kg/m2. She was mildly anaemic. Axillary and pubic hair was absent. Stigmata of liver disease was not found. Higher psychic function was normal but plantar reflexes were extensor initially later flexor bilaterally. Slit lamp exam showed no Kayser - fleischer ring. Abdomen examination revealed hepatomegaly. Reproductive system examination revealed underdeveloped breast with absent pubic hair.

Complete blood count with peripheral blood film revealed normocytic normochromic anaemia with thrombocytopenia. Liver function test showed normal serum bilirubin, prothrombin time with low serum albumin (2.7 gm/dl). Fasting lipid profile was always abnormal in the form of hypercholesterolaemia (192 mg/dl), hypertriglyceridaemia (575 mg/dl) and very low HDL level (15mg/dl). USG showed hepatosplenomegaly (coarse hepatic parenchyma) with infantile uterus and small ovary. Viral markers, ANA and Penicillamine challenge test were negative. MRI showed hyperintensity in head of caudate nucleus. EEG showed generalised encephalopathy. Blood for lysosomal acid lipase was reduced in activity (12.1 nmol/hr/mg; Mean: 34.6 nmol/hr/mg). Liver biopsy showed chronic hepatitis, Fibroscan reported fibrosis score 63.9, stage F4. Endoscopy showed grade II esophageal varices.

Discussion: Young patient with chronic liver disease with neuropsychiatric symptom, our first consideration was Wilson’s disease but absence of K-F ring, normal serum copper, ceruloplasmin level and negative penicillamine challenge test virtually excluded Wilson’s disease. Next consideration was storage disease (cholesteryl ester storage disease) as her lipid profile was abnormal and measurement of enzyme activity of lysosomal acid lipase was found to be lower. We treated the patient with atorvastatin initially then fibrates and was discharged with improving symptoms and near normal lipid profile.

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Bangladesh-Clinical Vignette-Poster Finalist
Sabbiha Majumder, MBBS

Title: New Gene for First Reported Case of ‘Tree Man Syndrome’ in Female

Authors: Sabbiha Nadia Majumder, KM Furkan Uddin, Md Robed Amin, Mohammad Abdul Aleem, Atiqr Rahman, Nushrat Jahan Dity, MD Abdul Baqui, Hosneara Akter, Muhammad Mizanur Rahman, Marc Woodbury-Smith, Stephen Scherer, Mohammed Uddin

Introduction: Tree man syndrome is an ultra-rare, genodermosis. [1]Most commonly an autosomal recessive genetic disorder [3]. Sporadic, sex-linked and autosomal dominant inheritance in some cases [4] A wide range of clinical presentations are reported : most striking cutaneous bark like lesions coined the term ‘Tree Man Syndrome’. [2]. The mutations in the EVER1 or EVER2 genes on chromosome 17, lead to an abnormal susceptibility of the patients to a specific group of HPV genotypes known as EVHPV causing various clinical features

Case Presentation: 10-year-old, S.K, only issue of non-consanguineous parents presented with horn like lesion on face for 4 years. Since 1 year of age, papularrashes in different parts of the body. Past history was insignificant. Local examination revealed multiple conical tree bark like lesions. Laboratory examination was unremarkable. Surgical intervention initially removed all bark like lesions and confirmed by histopathology. Using whole genome sequencing (WGS), we identified a rare stop gain somatic mutation in ANKRD26 gene and subsequently confirmed the mutation in the resected tree bark like tissue.

Discussion: Tree man syndrome is an exceedingly rare hereditary skin disease formally known as Epidermodysplasia verruciformis (EV). It has myriad clinical presentations ranging from flat wart like lesion, flat-topped papule, plaques to verrucous or seborrhoeic keratosis like lesions and even cutaneous horn like lesion mainly seen in UV- exposed regions. Malignant transformations occur in about 25% [4]. In our case, multiple flat-topped hyperpigmented papulonodular lesion were observed. Also horn like projections found on the face are not common presentation which were present in our case making it an atypical one.

Till now it was described exclusively in males with loss of function mutations in EVER1 and EVER2 gene in 75% of cases [2]. Using whole genome sequencing (WGS), we initially screened for mutations in EVER1 and EVER2. No mutation was found in these gene. Rather a rare somatic stop gain mutation in exon 11 of ANKRD 26 was identified.

Our findings suggest a new gene for EV and a pleiotropic impact of the gene into the etiology of EV. This is the first female case of an association between EV and ANKRD26 in the world. Further research on new gene needs to be explored.

References

Title: Primary Enterolith: An Unusual Cause of Severe Gastrointestinal Bleeding

Authors: Neha Agarwal, MD and Xon Ng, MD

Introduction: Enterolithiasis is the presence of a stone in the intestine. Though the prevalence has been reported as between 0.3% and 10%, this uncommon condition can be a diagnostic challenge that must be recognized quickly due to mortality rates up to 8%.

Case Presentation: A 42 year-old male with a history of a surgically repaired congenital diaphragmatic hernia, multiple partial small bowel obstructions due to adhesions, malrotation and volvulus requiring small bowel resection, and peptic ulcer disease presented to the Emergency Department with severe abdominal pain, maroon colored stool, and fevers for the past week. Physical exam was notable for blood pressure 89/53, and a 5cm pulsatile abdominal mass with moderate tenderness to palpation. Notable labs included WBC 12.98 k/uL and Hgb 7.1 g/dL (baseline 16 g/dL). The patient was admitted and received aggressive fluid and blood product resuscitation. The gastroenterology (GI) team was consulted and recommended an magnetic resonance angiogram (MRA) to evaluate for a duodenoaortic fistula. The MRA revealed an 8.1 cm lamellated intraluminal bowel mass. A follow-up CT of the abdomen and pelvis demonstrated a partial small bowel obstruction caused by an enterolith. In an attempt to localize the bleeding, the patient underwent an esophagogastroduodenoscopy which showed no culprit lesions up to the third portion of the duodenum. Further studies recommended by the GI team included a tagged red blood cell scan, which was also negative. In light of these negative studies, the leading diagnosis was enterolith-induced ischemia and subsequent mucosal ulceration proximal to a prior surgical anastomosis. The patient underwent an exploratory laparotomy, which revealed malrotation and two full thickness injuries within the ileum. The six feet of dilated loops of bowel containing the enterolith were removed, and an end-end jejuno-ileostomy was performed. The patient’s bleeding subsequently resolved.

Discussion: This case demonstrates the potential complications of large enteroliths inaccessible through standard endoscopic interventions. Surgical resection is reserved for severe cases of ileus, but further studies are needed to investigate the efficacy of new approaches to enterolith removal. Advances in these procedural skills may decrease morbidity and mortality associated with surgical intervention. This case suggests that clinicians should consider enteroliths as a possible cause of small bowel obstructions in patients with risk factors predisposing to intestinal stasis, such as inflammatory diseases or anatomical abnormalities. A high level of clinical suspicion is required to diagnose this uncommon condition, and prevent severe intestinal ischemia and gangrene.

References

California-Clinical Vignette-Poster Finalist
Bashar Al Hemyari, MD

Title: Puzzled by the Pannus!

Authors: Bashar Al hemyari MD, Mimi Biswas MD, Riverside Community Hospital/UCR School of Medicine

Introduction: Pannus is an abnormal layer of fibrovascular tissue or granulation tissue. It is a well-known process complicating 0.1%–0.6% of prosthetic heart valves within the first 5 years, leading to valve obstruction. Clinically it may present as TIA/stroke, syncope, and can eventually lead to heart failure. There are no case reports describing pannus formation on non-prosthetic valves.

Case Presentation: A 55 year-old female with past medical history of hypertrophic obstructive cardiomyopathy status post septal myomectomy and automated implantable cardioverter and defibrillator (AICD) placement six months ago, presented with sudden onset of aphasia and left sided weakness for five minutes, followed by syncope lasting for two minutes, after which she regained full consciousness with resolution of neurological deficits. She reported having two pre-syncopal attacks in the last four weeks. She denied fever, chills, chest pain, shortness of breath, or palpitations. Her vitals revealed blood pressure of 131/73 mmHg, heart rate of 78 bpm, temperature of 36.5°C, respiratory rate of 17 breaths per minute, and oxygen saturation of 98% on room air. Her neurological exam was unremarkable. No carotid bruits or heart murmur were appreciated. Both orthostatic and table-tilt tests were negative. Laboratory tests, including CBC, CMP, and fasting lipid panel, were unremarkable. EKG showed paced rhythm. AICD interrogation failed to show any arrhythmic events. Head CT did not show any new acute infarcts, and MRI brain was not done due to incompatibility with her AICD. A carotid ultrasound didn’t reveal any significant stenosis.

A transthoracic echocardiogram showed mitral valve mass, followed by a transesophageal echo which revealed a 3.4 cm mobile echogenic vegetation attached to the anterior leaflet of the mitral valve near the left ventricular outlet tract, with preserved ejection fraction and negative bubble study. Blood cultures were negative. A tagged WBC scan failed to show any nidus for infection. Further testing including ANA, anti-dsDNA, anti-CCP and rheumatoid factor were all negative.

During her hospital stay, she remained afebrile with no new neurological symptoms/syncope. The patient was started on anticoagulation and transferred to higher level of care where she had open thoracotomy with mitral mass removal. Pathology revealed calcified pannus with no vegetations/thrombi. The patient tolerated the surgery very well and anticoagulation was subsequently discontinued. The patient continues to be asymptomatic at a six-month post-operative follow up visit.

Discussion: Pannus growth on non-prosthetic valves is very rare with minimal data in the literature. Risk factors include previous cardiac surgery, however the mechanism is unknown. Our case is unique because the patient developed pannus on a non-prosthetic valve. There is not enough data to determine the benefit of chronic anticoagulation. However, symptoms like TIA/stroke and dyspnea warrant surgical removal to prevent further morbidity and improve quality of life.
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California-Clinical Vignette-Poster Finalist
Rani Berry, MD

Title: Steatohepatitis and Acquired Acrodermatitis: Rare Complications of Roux-En-Y-Gastric Bypass-Induced Weight Loss and Malnutrition

Authors: Rani Berry, MD, Claudio A. Rivera, MD, MPH, James H. Tabibian, MD, PhD, Internal Medicine Residency Program, UCLA Ronald Reagan Medical Center, Los Angeles, CA, USA, Division of Gastroenterology, Department of Medicine, University of Pennsylvania, Philadelphia, PA, USA, Division of Gastroenterology, Department of Medicine, Olive View-UCLA Medical Center, Sylmar, CA, USA

Introduction: Non-alcoholic fatty liver disease (NAFLD) is a common syndrome that includes both simple steatosis and steatohepatitis (NASH). Most commonly associated with co-morbidities such as diabetes, obesity, and hyperlipidemia, NAFLD is often reversed following gastric bypass surgery. This case highlights the important yet rare complication of Roux-en-Y gastric bypass surgery causing rapid weight loss and fatty deposition in the liver leading to steatohepatitis. Attention to this rare outcome is vitally important given that NASH is often asymptomatic and detected by abnormal labs and imaging.

Case Presentation: A 35-year-old woman with a history of gastric bypass surgery 10 years prior, with a recent complication of proximal common bile duct stricture requiring bile duct excision and Roux-en-Y hepaticojejunostomy presented with four weeks of cutaneous lesions. Additionally, the patient reported poor oral intake, weight loss, and chronic diarrhea. Physical examination revealed erythematous, scaly papules in a reticular pattern, with some lesions coalescing to form plaques on the dorsal hands, forearms, and distal lower extremities (Figure A). A skin biopsy demonstrated prominent hyperkeratosis with alternating areas of para- and orthokeratosis as well as a zone of pallor, which was noted in the upper portion of the epidermis with associated focal loss of the granular layer, consistent with acrodermatitis enteropathica or pellagra. Serum laboratory tests were significant for low levels of albumin (<1.5 g/dL), pre-albumin (7.1 mg/dL), vitamin B3/niacin (1.50 mcg/mL), and zinc (27 mcg/dL). Additionally, total bilirubin was 3.1 mg/dL, aspartate transaminase 58 U/L, alanine transaminase 92 U/L, alkaline phosphatase 231 U/L, and international normalized ratio 1.6. Magnetic resonance cholangiopancreatography demonstrated a patent anastomosis and absence of ductal dilation, hepatobiliary mass, or features of cirrhosis but was remarkable for severe hepatic steatosis (Figure B).

The constellation of clinico-pathologic and radiologic findings was consistent with severe malnutrition manifesting as acrodermatitis acquisita with features of pellagra as well as diffuse hepatosteatosis. Treatment including total parenteral nutrition for severe malnutrition and topical corticosteroid cream for her cutaneous lesions was initiated, resulting in resolution of rash, serologic abnormalities, and hepatosteatosis (Figure C).

Discussion: Nutritional deficiencies, including zinc and niacin, are a known complication of bariatric surgical procedures and can present with dermatologic manifestations. Additionally, rapid weight loss is also a known, albeit uncommon, etiology of hepatic steatosis. Management should focus on nutritional supplementation, as was done for this patient; topical treatments may also be provided if cutaneous involvement is symptomatic. This case serves to highlight the potential multisystem sequelae of rapid bariatric surgery-associated weight loss and consequent malnutrition and the utility of recognizing cutaneous and other manifestations and their relationship to broader systemic processes. In the setting of bariatric surgery, post-operative nutritional status must be closely monitored with a multidisciplinary approach to avoid clinically-significant sequelae.
References

Title: A headache or something more? A case of Lemierre’s syndrome with central venous sinus thrombosis

Introduction: Lemierre’s syndrome is a rare condition characterized by septic thrombophlebitis of the internal jugular vein, most commonly caused by *Fusobacterium necrophorum*. When not recognized early, complications include bacteremia or septic emboli to the lungs. We present a rare complication of *Klebsiella pneumonia*‐associated Lemierre’s syndrome, with thrombosis extending from the internal jugular vein into the central venous sinuses.

Case Presentation: A 43‐year‐old man with diabetes mellitus, intravenous drug use, tobacco abuse, and a history of prior incarcerations presented to the emergency department with left ear drainage for several weeks associated with cough, myalgias, rhinorrhea, and headaches. On two prior ED visits, he was diagnosed with acute otitis media, treated with amoxicillin, and pneumonia, treated with azithromycin. His symptoms failed to improve and was admitted to the hospital days later with high grade fever, tachycardia, and agitation with labs notable for an elevated lactate and CRP. Head CT showed complete opacification of the left middle ear cavity and density within the left mastoid air cells suggesting a component of osteitis. He was diagnosed with recurring acute otitis media by Otolaryngology and started on broad spectrum antibiotics. On hospital day two, he developed left sided torticollis with tenderness of the left neck and leukocytosis. His mental status also declined, eventually becoming oriented only to self. CT Neck revealed left internal jugular venous thrombophlebitis with thrombosis of the mid and upper cervical internal jugular vein as well as the left sigmoid and transverse sinuses. Blood cultures grew *Klebsiella pneumoniae*. Chest CT revealed septic emboli to the lungs. The patient was diagnosed with Lemierre’s syndrome and underwent surgical myringotomy for source control and anticoagulation for his central venous thrombosis. His symptoms improved and he was discharged on a six‐week course of oral levofloxacin.

Discussion: Lemierre’s syndrome stems from an oropharyngeal infection that spreads to the carotid sheath vessels and specifically the internal jugular vein gradually over the course of a week and is most frequently caused by *Fusobacterium necrophorum*. Only a handful of case reports implicate *Klebsiella pneumoniae* as the causative organism. When *Klebsiella* is involved, it may be associated with thrombotic extension into the central venous sinuses. When thrombosis is localized to the internal jugular vein or with septic pulmonary emboli, the role of anticoagulation remains unclear. With thrombotic extension into the cerebral sinuses, anticoagulation is recommended. In all cases, treatment includes four weeks of antibiotics and surgical intervention if the sepsis does not improve with antibiotics alone.

References

Title: Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia: A Rare Syndrome or Under Recognized?

Authors: Andrew J. Heisel*, MD; Atul N. Sharma*, MD, MPH, CHES; & Carrie Chun, MD, *Co-first authors, Scripps Clinic Internal Medicine Residency Program

Introduction: Although the exact pathophysiology is still elusive, Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia (DIPNECH) is thought to be as result of proliferation of pulmonary neuroendocrine cells (PNE) in the bronchial wall. Often mistaken for obstructive lung disease, DIPNECH is more prevalent in females and has no clear association with smoking.

Case Presentation: 75-year-old Caucasian female with past medical history of interstitial lung disease for 12 years of unclear etiology and chronic respiratory failure with hypoxemia presented to the pulmonary clinic to transfer care from another hospital system. For the preceding 6 months, she had been experiencing progressive shortness of breath and dyspnea on exertion with associated dry cough. She was oxygen dependent and was no longer able to perform her activities of daily living. Her medications included montelukast, loratadine/pseudoephedrine, oral Prednisone and albuterol inhaler.

Her vitals were notable for oxygen saturation of 92 on 3L/min via nasal cannula. She was a chronically ill-appearing obese female in no overt distress and her lung exam revealed diffused crackles and decreased breath sounds at the bases bilaterally. No wheezes were audible. Routine and specialty labs (aldolase and alpha 1 antitrypsin) were normal. Lung biopsy was performed and specimen was sent to Mayo Clinic. Pathology results showed chronic hypersensitivity pneumonitis vs non-specific interstitial pneumonia and neuroendocrine/carcinoid tumorlets consistent with DIPNECH.

She achieved minimal relief with supplemental oxygen, short and long acting bronchodilators and oral corticosteroids, and has been referred to endocrinology for possible octreotide therapy.

Discussion: Aguayo et al first published a clinical series of 6 cases describing DIPNECH in 1992 and there are currently fewer than 200 reported cases in the scientific literature worldwide. It is considered pre-invasive/pre-neoplastic lesion for lung carcinoid tumors by the World Health Organization and is found in 5.4% of individuals undergoing surgical resection for lung carcinoid tumors.

The condition primary affects non-smoking women between 60 and 70 years of age and due to non-specific symptoms, it is often mistaken for asthma. Dry irritating cough and dyspnea are the most common presenting symptoms. Surgical lung biopsy is the gold standard for making the diagnosis and requires histo-pathological conformation of pulmonary neuroendocrine (PNE) cells confined to the epithelium of large and small airways.

DIPNECH is typically an indolent and non-progressive disorder, however, some patients can have progressive symptoms. There have been no clinical trials to determine effective treatment to date. Management includes inhaled corticosteroids, systematic steroids, somatostatin analogs, chemotherapeutics or even lung transplantation in extreme cases.
Increased recognition of this rare entity is due to advanced imaging studies and better staining methods. Diagnosis should be considered in patients with refractory cough, obstructive pulmonary physiology and even with small pulmonary nodules that are stable over many years.

References

Title: Bone to Lung: Dissemination of Foreign Bodies and Infection in a Cirrhotic Patient

Authors: Quoc-Anh Ho, MD
Department of Internal Medicine, UCSF-Fresno Center for Medical Education and Research, Fresno, CA.

Introduction: Cement embolism and osteomyelitis are known complications of vertebroplasty. Empyema originating from a hardware infection is much rarer.

Case Presentation: A 52-year-old woman with medical history of alcoholic cirrhosis and thoracic-spine compression fractures treated with vertebroplasty complicated by osteomyelitis presents to the emergency department for acute respiratory distress. Her vital signs on initial presentation were significant for a blood pressure of 84/57, heart rate of 115, respiratory rate of 32, and an oxygen saturation of 94% on 4 liters per minute nasal canula. She was afebrile on presentation. On examination, she was somnolent, overtly jaundiced, and had diminished breath sounds in the base of her left lung. Her white blood cell count was $35.9 \times 10^9/L$ with 20% bandemia. Her MELD-Na score was 34. Chest radiograph revealed a left sided pleural effusion. Her blood pressure responded to normal saline in the emergency department. The patient was admitted and started on empiric vancomycin and piperillin-tazobactam. Her mental status had improved and lactulose was held off. Computed tomography (CT) of the chest revealed a large left-sided pleural effusion collection consistent with an empyema as well as retained cement fragments consistent with cement pulmonary embolism. Surgery service was consulted and was able to place a chest tube for drainage. 200 mL of pleural fluid was extracted, which was much smaller in volume than how the effusion appeared on CT-chest. Culture of the pleural fluid grew out extended-spectrum beta-lactamase (ESBL) Escherichia coli that was sensitive to meropenem. Infectious diseases was consulted and recommended two weeks of meropenem. Pulmonology was consulted and initially recommended aggressive diuresis. However, as the effusion did not improve significantly, a three-day course of intrapleural fibrinolytics was added. During the patient's inpatient treatment, her medical records from an outside hospital were obtained and reviewed. One year prior to the current admission, she was found to have compression fracture of T9-T11 requiring vertebroplasty. Her back pain worsened months later, and she was found to have ESBL-osteomyelitis of the implanted hardware. However, the patient was lost to follow-up and did not receive appropriate care. Infectious diseases was re-consulted during this admission, and recommended a six-week course of meropenem. The patient's clinical course slowly improved over the course of her four-week hospitalization and she was discharged to a skilled nursing facility to receive the remainder of her meropenem.

Discussion: Cirrhosis, particularly when severe, is a known cause of immunocompromise as it impairs the immune surveillance function of the liver and reduces hepatic protein synthesis. Furthermore, hepatopulmonary syndrome raises the susceptibility of the lungs to circulating infection. This patient had a known untreated ESBL-osteomyelitis of her vertebral hardware that manifested in cement pulmonary embolism and ESBL-empyema one year later. This case supports the need for more vigilant treatment of chronic co-morbidities, especially infections, in cirrhotic patients.
Title: Isolated Bilateral Optic Neuropathy in Wernicke Encephalopathy: A Political Loss of Vision

Authors: David Ly MD, Lisa Gaynon MD, Jason Mok MD, Stephanie Hsiao MD, Sara L. Swenson, MD

Introduction: Wernicke encephalopathy (WE), a neurological disorder due to thiamine deficiency, is classically the triad of oculomotor dysfunction, encephalopathy and gait ataxia. We describe an unusual case of WE presenting as bilateral optic neuropathy and vision loss.

Case Presentation: An undocumented 22 year old male from Mexico with no past medical history was held in an immigration detention center for 3 months where he had poor oral intake and a self-limited diarrheal illness. In the next 6 months, he developed progressive weight loss, tinnitus and blurry vision. Ultimately, a retinal specialist noted bilateral optic neuropathy, and he was admitted for expedited work-up. He demonstrated bilateral horizontal nystagmus and vision was light perception only. Routine labs and CSF studies were unremarkable. Brain MRI/MRA revealed symmetric hyperintensity involving the periaqueductal gray and tectal plate that was concerning for Wernicke encephalopathy. He denied excessive alcohol use. An extensive evaluation excluded other causes, including negative NMO antibody studies, ANA, ESR/CRP, HIV, Quantiferon, RPR, vitamin B12, spine MRI, and upper endoscopy. Surprisingly, his vitamin B1 level was normal. The patient received parenteral thiamine and empiric high-dose methylprednisolone but, unfortunately, neither improved his vision.

Discussion: Ocular symptoms of Wernicke encephalopathy generally present as horizontal and vertical nystagmus, vertical and conjugate gaze palsies and internuclear ophthalmoplegia. Wernicke encephalopathy rarely presents with optic neuropathy. All sequelae of WE, if left untreated, inevitably lead to poor outcomes. Previous case reports have noted similar bilateral optic neuropathy in patients with thiamine deficiency secondary to ketogenic diet or malabsorption state from persistent diarrhea. Brain MRI is the imaging test of choice for WE, with a 53% sensitivity and 93% specificity for detecting WE. Lesions appear as decreased T1 signals or increased T2/FLAIR signals in the periaqueductal gray and third ventricular area and within the tectal plate, thalamus, medulla, and mammillary bodies. The sensitivity and specificity of serum thiamine levels in symptomatic patients is unclear. Though low levels suggest deficiency, serum and CNS levels are not correlated and a normal serum level does not rule out WE. Treatment with thiamine can reverse MRI abnormalities within 48 hours. Normal CSF studies narrow the differential diagnosis since a pleocytosis or protein level >100 mg/dL is not typical for WE. Sadly, our patient’s initial detention put him at risk for thiamine deficiency, and his poor access to medical care delayed his diagnosis and treatment, resulting in permanent vision loss.

The classic triad of WE includes ophthalmoplegia, ataxia and confusion. However, it is important to recognize that an alternate presentation, although uncommon, can include isolated optic neuropathy and visual symptoms. Left untreated, it can lead to severe, irreversible vision loss.
References

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California-Clinical Vignette-Poster Finalist
Lauren M Mathias, MD

Title: Disseminated Mycobacterium Avium Complex: A Rare Cause of Diffuse Alveolar Hemorrhage

Authors: Lauren Mathias, MD and Seth Politano, DO Department of Medicine, Keck School of Medicine of USC, Los Angeles, CA.

Introduction: Diffuse alveolar hemorrhage (DAH) is a life-threatening syndrome characterized by bleeding into the alveolar spaces of the lungs. DAH is most commonly caused by capillaritis associated with systemic autoimmune diseases such as anti-neutrophil antibody-associated vasculitis, systemic lupus erythematosus, and anti-glomerular basement membrane disease. DAH can less commonly result from alveolar damage due to infection, inhalation, or cytotoxic drug therapy. Here we present a case that has not been described in current literature: DAH caused by disseminated Mycobacterium avium complex (MAC).

Case Presentation: A 34 yo Hispanic male with recently diagnosed AIDS (CD4 <20) presented to our hospital with fevers, chills, shortness of breath, and hemoptysis. Patient initially presented one month ago with similar symptoms at an outside hospital (OSH). CT Thorax showed bilateral pleural effusions with diffuse patchy groundglass and consolidative opacities. CT Abdomen/Pelvis demonstrated diffuse retroperitoneal and mesenteric lymphadenopathy. Mycobacterium tuberculosis was ruled out with three negative AFB stains, and patient was discharged with a course of amoxicillin. Two weeks later, patient was readmitted to the OSH due to worsening symptoms. At this time, AFB sputum culture from initial admission returned positive. Patient was started on rifampin, isoniazid, pyrazinamide, and ethambutol (RIPE). Shortly after discharge, patient presented to our hospital for continued shortness of breath and hemoptysis. Pulmonology and Infectious Disease were consulted and agreed that AFB culture positivity one week after collection suggested disseminated MAC, which was confirmed by DNA probe. RIPE was discontinued, and treatment for MAC was initiated with rifabutin, ethambutol, and clarithromycin. Diagnostic bronchoscopy demonstrated DAH. PCP-DFA was negative, and there was no evidence of pulmonary Kaposi’s sarcoma. Broad-spectrum antibiotics were given for a possible secondary bacterial etiology, but the patient continued to deteriorate. Rheumatology was consulted to evaluate for a possible autoimmune etiology of DAH. Although serologies were negative and a urinalysis did not show active sediment, IV Solumedrol was given. Despite maximal treatment, patient’s clinical picture continued to worsen. The final diagnosis was determined to be DAH secondary to disseminated MAC.

Discussion: The most common causes of DAH are systemic autoimmune diseases. Infectious disease, however, should be considered early in the differential diagnosis of DAH, especially in patients who are immunocompromised. It is well-established that early targeted antimicrobial therapy improves survival. The most common infections causing DAH include influenza A, dengue, leptospirosis, malaria, and S. aureus pneumonia. While MAC has been associated with alveolar damage, we describe the first case of DAH secondary to disseminated MAC.

References
Title: DISSEMINATED ACTINOMYCES INFECTION: A Case of Malignant Mimicry

Authors: Khoi Pham, DO; Jeremiah Sisay, MD; Leslie Cler, MD, FACP. Methodist Dallas Medical Center, Dallas, Texas.

Introduction: Learning Objective: Recognize infection with Actinomyces as a possible mimic of metastatic cancer with abdominal masses and pulmonary nodules.

Case Presentation: A 53-year-old African American male presented with right-sided abdominal pain on and off for 9 months, worsening in the last 2 to 3 months. Associated symptoms included recent right abdominal swelling which resolved without treatment and 30 lbs weight loss over the last 6 months. The patient was a 70-pack-year smoker with poor dental hygiene. Vital signs revealed 101.5 degree Fahrenheit and pulse of 108 beats per minute. On exam, he had mild tenderness over the right upper and lower quadrants with firmness of the right upper quadrant and flank area but no hepatosplenomegaly. Abdominal CT showed a large mass lesion involving the right body wall and extending into the lateral right intra-abdominal cavity and right paracolic gutter along with a large mass lesion in the right lobe of the liver and numerous tiny pulmonary nodules. He had FNA biopsies of liver 3 different times and a liver wedge biopsy which all were negative for malignancy. An investigation into possible infectious etiologies came up negative until final pathology reported single cluster of filamentous organisms (GMS stain positive, gram positive and AFB negative), which was interpreted as Actinomyces with the masses being a reactive/inflammatory process related to the infection. During his 4-week hospital course, he was treated empirically with IV vancomycin for 10 days and piperacillin-tazobactam for 15 days. His fever resolved within 3 days of treatment and leukocytosis slowly trended down. Patient was started on amoxicillin for 6 months when final pathology result came back.

Discussion: In cases of fever and leukocytosis with metastatic-appearing lesions and pathology without evidence for malignancy, tuberculosis or fungus, keeping actinomyces in the differential, though rare, is important. The fact that this patient had poor oral hygiene with significant smoking history may have been the entry point for his disseminated disease. Treatment initially with IV antibiotics including broad-spectrum penicillin likely contributed to his clinical improvement. Once identified, 6 months of amoxicillin is appropriate for treatment of disseminated actinomyces.
Title: Fever, Ferritin, and Multi Organ Failure

Authors: Saduvala, S, MD; Ahmed, Z, MD; Wang, M, MD; Halanych, JH, MD, MSc.

Introduction: Hemophagocytic Lymphohistiocytosis (HLH) is a hematological condition, rarely seen in adults, characterized by activated macrophages acting outside normal regulatory conditions.

Case Presentation: A 24 year old man with a history of autism and hypertension presented with 4 days of nausea, vomiting, epigastric pain, diarrhea and loss of appetite. He denied tobacco and alcohol use. He had temperature of 101.9 F, heart rate 132 bpm, respiratory rate 28/min, and blood pressure 153/113 mm Hg. He was cognitively impaired due to his autism, but otherwise his physical examination was normal. His initial labs showed hemoglobin 6.9 gm/dl, WBC 6.7 cu/mm, platelets 230,000, BUN 90 units, creatinine 10.9 units, creatine kinase 1114 units and lipase 844 units. Urinalysis is positive for proteinuria and occult blood with only 7 RBC's. Iron studies were normal except for ferritin 30,900 units. His lipid panel was normal except for triglycerides 675 mg/dl. Recent outpatient lab work was normal. Chest radiograph was normal, and all cultures were negative. Given his non-infectious febrile illness and multi organ involvement with elevated ferritin, we were concerned about Hemophagocytic Lymphohistiocytosis (HLH). Patient met 5 out of 8 criteria for HLH with fever, cytopenia, elevated triglyceride, elevated ferritin, and elevated Interleukin 2 receptor (CD 25) soluble at 9790 pg/ml. Bone marrow biopsy showed minimal hemophagocytosis. Kidney biopsy showed acute tubular necrosis with focal segmental glomerulosclerosis. Further viral and infectious causes were ruled out with negative HSV, EBV, CMV, hepatitis and HIV. Due to renal involvement our final diagnosis of HLH with renal syndrome was made. After initiating IV steroids patient’s clinical status and serum ferritin levels began to improve. Patient was discharged home after 5 days with 60mg of prednisone without requiring dialysis. Unfortunately, family was unable to fill the steroid prescription and the patient returned to the ED one week later with abdominal pain, nausea, and shortness of breath. While in ED he had a cardiac arrest with pulseless electrical activity and resuscitation was unsuccessful.

Discussion: This case highlights an uncommon presentation of HLH with renal syndrome. Adult onset HLH is rare and is seen secondary to an underlying disease such as malignancy, infections or autoimmune disorders. It’s reported incidence is approximately 1.2 cases per million individuals per year. The diagnosis of HLH is particularly challenging and a combined picture of SIRS, rash, generalized lymphadenopathy, hepatosplenomegaly, cytopenia and hyperferritinemia, should raise the suspicion for HLH in the correct clinical setting. Renal involvement in HLH is poorly understood and acute kidney injury is the most common manifestation. Treatment of HLH is complex and requires removal of the triggering event, immunosuppression and supportive care. Early recognition and treatment of this rare clinical entity is imperative, to prevent fatal outcomes as seen in our case.

References
Title: Uveitis Caused By Neurosyphilis

Authors: Aditi Shrivastava, MD., Dr. Daniel Hart

Introduction: Several ocular symptoms are known to be associated with neurosyphilis. Our clinical observations suggested that uveitis is an under-recognized but common ocular manifestation of neurosyphilis that has important short-term and long-term prognostic significance. The low incidence with which it is encountered makes neurosyphilis a diagnostic challenge.

Case Presentation: A 56 year old gentleman consulted his rheumatologist and ophthalmologist because of diffuse joint pain, lip swelling, and left eye blurry vision. Additional history revealed that at age 16, he had experienced urethral discharge and dysuria, and after diagnosis with syphilis, was given one dose of penicillin upon which symptoms resolved, but never followed up. A clinical diagnosis of bilateral uveitis and syphilis was made. Upon evaluation by the rheumatologist, an X-ray of all joints was performed, which were negative, and eye drops minimally alleviated symptoms. When seen by his ophthalmologist, the patient reported seeing floaters and was diagnosed with bilateral uveitis, as he was noted to have inflammation of the eyes, associated with ciliary flush. Subsequently, the rheumatologist performed bloodwork and he tested positive for syphilis (RPR 1:128 titer and Treponema pallidum). He then presented to the hospital for IV treatment and neurological evaluation. In addition to bilateral uveitis, the physical examination revealed mild impairment of hearing in the left ear during the whisper test. A lumbar puncture revealed CSF VDRL titer 1:1, with WBC 49 (19% segments, 77% lymphocytes), protein 93, glucose 65. Serology workup, as inpatient, revealed RPR 1:256 reactivity along with FTA-ABS reactive. Antibiotic therapy with Crystalline penicillin was initiated. After his first dose of penicillin, he experienced rigors, abdominal pain, nausea and vomiting. Because cultures were negative, symptoms were attributed to a Jarisch Herxheimer reaction, in which antibiotic treatment caused release of spirochetes, resulting in a systemic inflammatory response. Supportive treatment was initiated with IV fluids, Benadryl, and ibuprofen. Subsequently, symptoms subsided and patient was discharged from the hospital with PICC line in place for antibiotic therapy for two weeks. Upon outpatient followup, RPR titers decreased to 1:32, and later 1:2, along with resolution of headaches, visual changes, hearing loss, and arthralgias.

Discussion: This case illustrates the potential for neurosyphilis to manifest as uveitis and the value of a complete history and physical exam. Although there are many causes of uveitis, syphilis remains an essential differential diagnosis to consider, despite its rare prevalence. Recognition of this manifestation is critical to institution of appropriate therapy and prevention of Inflammatory Ocular Hypertension Syndrome, where intraocular pressure acutely increases secondary to direct inflammation of the trabecula.
California-Clinical Vignette-Poster Finalist
Alan Sit, MD

Title: Syncope as a Presentation of Sclerosing Mesenteritis

Authors: Alan Sit, M.D., Walter Coyle, M.D., Enoch Wang, M.D., FACP, Department of Internal Medicine, Division of Gastroenterology; Scripps Clinic Medical Group; La Jolla, CA

Introduction: Sclerosing mesenteritis is a rare primary inflammatory and fibrotic process involving the mesentery. It presents as a broad spectrum of symptoms including abdominal pain, nausea, vomiting, weight loss, and fever through mass effect on gastrointestinal lumen and mesenteric vessels. Our case illustrates the importance of a careful history in this unusual presentation of a rare condition, sclerosing mesenteritis.

Case Presentation: A 56-year-old gentleman with paroxysmal atrial fibrillation status post two ablations and prior laparoscopic cholecystectomy presented with syncope after urination. His vitals, physical exam, labs, EKG, and transthoracic echocardiogram were unremarkable including negative orthostatic vitals and troponins. His presentation was thought to be secondary to micturitional syncope. Upon more careful questioning, he endorsed years of abdominal cramping, early satiety, and bloating that worsened during admission. A CT of the abdomen and pelvis and eventually a PET scan revealed a 15.6cm area of sclerosing mesenteritis and hypermetabolic sub-centimeter mesenteric lymph nodes compressive of the mesenteric vasculature. The mass and adjacent lymph nodes were surgically resected and reviewed by two additional outside pathologists who concluded that the histology was most consistent with non-fibrotic sclerosing mesenteritis without any evidence of lymphoma. Due to the association between sclerosing mesenteritis and autoimmune pancreatitis, an IgG4 level was checked and was normal. He was trialed on a one-month burst of prednisone with minimal improvement in his symptoms. Subsequent colonoscopy and gastric emptying studying were normal. One year later, a repeat CT and MRI of the abdomen and pelvis showed recurrent central mesenteric inflammation.

Discussion: Sclerosing mesenteritis is a rare idiopathic inflammatory and fibrotic condition that affects the mesentery or any abdominal structure including the vagus nerve which was thought to have contributed to his syncope. Its pathogenesis is unclear but there are associations with prior abdominal surgery, autoimmune conditions especially autoimmune pancreatitis, paraneoplastic syndromes (70% of cases associated with lymphoma, breast cancer, and melanoma among others), and infection. Nonetheless, CT scan is the best initial test followed by surgical biopsy to confirm the diagnosis and rule out other etiologies. Management should be tailored to symptom resolution rather than radiographic findings and consists of immunosuppressive medications. Glucocorticoids plus tamoxifen is first line followed by azathioprine, cyclophosphamide, and thalidomide for refractory cases. This case illustrates the importance of a careful history and an unusual presentation of a rare condition, sclerosing mesenteritis.
Title: The Thyroid STEMI

Authors: Jason Tso, Lisa Liu, Philip Ng

Introduction: Hyperthyroidism has numerous effects on the heart ranging from hemodynamic and rhythm changes to congestive heart failure. Thyroid storm is a rare and life-threatening decompensated thyrotoxicosis requiring intensive supportive care and antithyroid therapy. Takotsubo’s cardiomyopathy is an often reversible nonischemic cardiomyopathy elicited by emotional or physiologic stress. Here we report a rare case of Takotsubo’s cardiomyopathy in a patient with thyroid storm and a history of Grave’s disease.

Case Presentation: A 36 year old woman with a history of Grave’s disease, bariatric surgery, and chronic pancreatitis was admitted to the hospital for abdominal pain, nausea, and vomiting. The patient was status post radioactive iodine ablation six months ago and had tapered off methimazole one week prior to admission. Her hospitalization was complicated by intermittent sinus tachycardia and at one point she became acutely confused and agitated. EKG at that time revealed ST-elevations in the precordial leads with accompanying troponinemia. She had an emergent cardiac catheterization which revealed no coronary artery disease. Echocardiogram revealed an ejection fraction of 24%, down from 65% two days prior, and apical akinesis. The patient then became febrile and nonresponsive and was intubated. TSH was suppressed at 0.03 mcu/mL and thyroid hormone levels were elevated at T4 of 3 ng/dL and free T3 at 5.6 pg/mL. The patient was in clear thyroid storm based on the Burch-Wartofsky Point Scale. A diagnosis of Takotsubo’s cardiomyopathy was also made based on the sudden left ventricular dysfunction, apical ballooning on echocardiogram, and cardiac catheterization findings. The patient was treated with an esmolol drip, propylthiouracil, and dexamethasone for thyroid storm. She regained mental function and was extubated and transitioned to oral beta blockers 3 days later. Thyroid hormone levels decreased to normal range in 6 days and the patient regained normal left ventricular function with resolution of heart failure in two weeks.

Discussion: Takotsubo’s cardiomyopathy is generally thought to be due to an excess of catecholamine activity on the heart. In this case, that stimulus came in the form of thyroid hormone, which has a close interrelation with catecholamines and their adrenergic activity. Thyroid storm, often dramatic in presentation as in this case, is characterized by fever, central nervous system abnormalities, GI dysfunction, tachycardia, heart failure, and arrhythmia. Beta-blocker therapy is a mainstay of treatment but is also contraindicated in most cases of acute decompensated heart failure. However, in a case of Takotsubo’s precipitated by thyroid storm, treating the underlying thyroid pathology is the best way to improve the heart failure. Though thyroid induced heart failure is rarely this acute, rapid resolution of this condition with appropriate antithyroid medication and beta blockade may result in rapid cardiovascular recovery.

References


Title: Severe type B lactic acidosis as the initial presentation of monomorphic post-transplant lymphoproliferative disorder

Authors: Aline Zorian, UCLA, Internal Medicine

Introduction: Post-transplant lymphoproliferative disorder (PTLD) is the most common non-cutaneous malignancy in solid organ transplant recipients. However, due to its highly variable and frequently nonspecific clinical presentation, it frequently poses a diagnostic challenge for clinicians.

Case Presentation: A 67 year-old patient presented 11 years after deceased donor renal transplant with generalized weakness and malaise. His laboratory studies revealed acute renal insufficiency, pancytopenia, and most notably severe anion gap metabolic acidosis with a profoundly elevated serum lactate. His vital signs were unremarkable, and neither his review of systems nor his physical exam were suggestive of a localized source of infection. He remained normotensive; however his serum lactate continued to uptrend (peak lactate 15.2 mmol/L) despite intravenous fluid resuscitation and empiric broad spectrum antimicrobials. Extensive infectious workup including blood and urine cultures, cerebrospinal fluid studies, and viral serologies were unremarkable, with the exception of elevated EBV PCR and mildly elevated CMV PCR. Ultrasound of his renal allograft was unrevealing. CT scans of the chest, abdomen, and pelvis showed diffuse lymphadenopathy as well as scattered peritoneal nodules. Biopsy of one such nodule was diagnostic for monomorphic PTLD, Burkitt lymphoma type. Subsequent bone marrow biopsy revealed 80% lymphomatous involvement. Treatment with systemic and intrathecal chemotherapy was initiated and resulted in rapid clinical improvement, including normalization of serum lactate levels. However, his hospital course was later complicated by refractory pancytopenia, encephalopathy, bacteremia, and septic shock, ultimately resulting in death.

Discussion: This case highlights hurdles both in the diagnosis and in the management of PTLD, as well as the unique importance of maintaining a broad differential for common symptoms and common laboratory abnormalities in the care of organ transplant recipients. Though lactic acidosis due to tissue hypoperfusion is the most frequent cause of metabolic acidosis among hospitalized patients, alternative etiologies of an elevated lactate level, including malignancy, should be considered in all patients who do not have evidence of systemic hypoperfusion. Early consideration of PTLD in the diagnostic workup of an organ transplant recipient is critical, as early treatment can be instrumental in the management of this severe and often fatal complication of solid organ transplant.
Colorado-Clinical Vignette-Poster Finalist
Alexander Steinberg

Title: Syphilis Uveitis, Importance of Early Recognition with Rising Incidence

Authors: Alex Steinberg, MD, Saint Joseph Hospital, Denver CO

Introduction: Since its first description over 500 years ago, syphilis has become a rare entity thanks to the widespread use of penicillin. However, the incidence of this sexually transmitted Treponema infection has been on the rise since 2000. We present a typical case that was initially misdiagnosed and therefore allowed to progress.

Case Presentation: A forty one year old woman presented with three weeks of visual disturbances including floaters in her upper left vision and six days of a new blurry spot in the central visual field of her left eye affecting her depth perception. Over the last nine months she has had a number of new physical complaints including headaches, patchy hair loss, intermittent diarrhea, and increased cold sore-like lesions. She also noted a rash that started on her soles and palms and subsequently spread to her lower legs, upper arms, and thorax. The rash lasted three months and was diagnosed as hives by both her primary care physician and dermatologist. The patient described hard pruritic nodules below the surface of the skin that scaled over without blistering. She denied pain. Due to the spots in her vision, the patient was seen by ophthalmology. She was found to have a large, central scotoma of the left eye with significant retinal loss in both eyes. Ophthalmology ordered a RPR to “rule out syphilis” and asked the patient to return in 2 days for fundus auto-fluorescence. Interestingly, she was found to have a positive RPR and RPR titer of 1:128, diagnosing syphilis uveitis. She was treated with penicillin G and monitored for Jarish Herxheimer reaction. The patient was discharged with a full course of antibiotic therapy.

Discussion: The incidence of syphilis is on the rise. According to the CDC, in 2015, there were 117 cases of primary and secondary syphilis in Denver County, or nearly 18 cases per hundred thousand people. There were 99 cases, or just over 16 per hundred thousand in 2010. This is a modest increase compared to parts of the country such as Los Angeles and San Diego where rates have doubled during the same period. Incidence has increased in nearly every metropolitan center in the country. The rising rate of syphilis infection is most marked in men who have sex with men, however as this case demonstrates, MSM is not the only population at risk. Our patient first presented with the classic non-painful palmar rash, gastrointestinal distress, patchy hair loss, and constitutional symptoms. Unfortunately, by the time her diagnosis was made, some of the neurologic sequelae of this infection had begun to develop. With the incidence of syphilis on the rise, this case stresses the importance that all providers consider and recognize this ancient infection.
**Connecticut-Clinical Vignette-Poster Finalist**

**Deborah Akanya, MD**

**Title:** “LINES” from “Lines”: Levamisole-Induced Necrotizing Syndrome from Intranasal Cocaine Use.

**Authors:** Akanya Deborah MD, Cadan Anand MD, Jayaraman Ramya MD, Herman Edward MD, Joseph Mattana MD.

**Introduction:** Various medical conditions may result not only from illicit drug use but also from inadvertent exposure to additives used to either “cut” or dilute the drug and or to enhance the effect. Here we describe a patient with cocaine use who developed anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis from levamisole.

**Case Presentation:** A 22-year-old woman with rheumatoid arthritis (RA) and daily intranasal cocaine use presented to the emergency department for evaluation of recurrent left ankle blisters and generalized body lesions. She described the lesions, which first appeared 2 years previously, as tender blisters that would burst, turn black and become foul smelling with yellowish discharge. She stated that the lesions would resolve while taking prednisone but would reemerge once she stopped it. She had normal vital signs and palpable purpuric lesions on her ear lobes, nose tip, cheeks, trunk, finger tips and extremities were present. The lesions varied in size and were at different stages of healing. Some lesions had central necrosis and others had necrotic borders and were tender to palpation with purulent discharge. She had a white blood cell count of 12.2 X 10⁹/L with 80% neutrophils, mild anemia and an ESR of 85mm/h. Broad spectrum antibiotics were initiated along with aggressive wound care with debridement. P-ANCA was positive and ANA and RF were negative. Skin biopsy revealed a pattern of inflammation and thrombosis compatible with levamisole-induced necrotizing syndrome (LINES). She was treated with prednisone with marked improvement in the lesions. The importance of cessation of cocaine use was emphasized.

**Discussion:** LINES is a characteristic cutaneous vasculitis syndrome associated with the use of levamisole-adulterated cocaine. Levamisole, a veterinary anti-helminthic drug, has become a commonly used cocaine bulking agent which is difficult to detect and thought to intensify the ecstasy effect of cocaine through release of dopamine. Despite the fact that most of the cocaine in the US is believed to contain levamisole only 16 cases of LINES have been described. Our patient, who used intranasal (“lines” of) cocaine, manifested characteristic skin findings, supporting serologic findings including p-ANCA positivity, and confirmatory histologic features described as microvascular thrombi causing occlusive vasculopathy and or leukocytoclastic vasculitis involving the small vessels, deep dermis and superficial skin. Most patients experience resolution of lesions with cessation of illicit drug use, steroids and wound care but skin grafting and amputation may become necessary in some cases. It is plausible that our patient’s underlying autoimmune disease (RA) might have predisposed her to developing this condition but this is speculative. The diagnosis of LINES should be considered in patients with skin necrosis and illicit drug use especially given the high prevalence of levamisole contamination of cocaine.
Connecticut-Clinical Vignette-Poster Finalist
Iradat Amusa, MBChB

Title: Cytomegalovirus - A Nondiscriminatory Virus!!

Authors: Iradat Amusa MD MPH, Divyansh Bajaj MD, Venkata Manchala MD

Introduction: Cytomegalovirus (CMV) is capable of producing both congenital and acquired infections. It has the ability to produce equitably devastating and catastrophic consequences in both immune-competent and immunosuppressed patients. Here we report a case of acute CMV infection with severe manifestations in an immunocompetent patient.

Case Presentation: A 17 year old male presented to the emergency department with severe epigastric pain, headache and generalized weakness for one day. Upon arrival, vital signs were remarkable for temperature of 39.3C and heart rate of 116 bpm. Physical examination revealed an acutely ill appearing male with bilateral conjunctival injection, marked right upper quadrant tenderness and a diffuse morbilliform rash. Labs were remarkable for WBC 16.7, ALT-207, AST-171, ALP-179, total bilirubin 5.9 and direct bilirubin 1.6. He was admitted with a working diagnosis of sepsis secondary to possible hepatobiliary disease. Ultrasound abdomen revealed cholelithiasis without cholecystitis and mild splenomegaly. Initially, patient was treated empirically with cefepime and metronidazole to cover for abdominal infections. MRCP was negative for acute cholangitis. Blood and urine cultures and HIV testing were negative. Viral serologies such as EBV, CMV and HSV were sent for hepatitis work up. CMV IgM antibodies were significantly elevated at 97.7 establishing diagnosis of acute CMV infection. Hospital course was complicated by presumed CMV pneumonia with patchy, bilateral lung infiltrates on chest x-ray, despite being on broad spectrum antibiotics. Patient’s immunoglobulin levels were normal. He was given aggressive supportive care without initiation of antiviral therapy as he was immunocompetent. Over the course of a week, patient’s clinical status improved and liver enzymes trended down. Patient had complete resolution of symptoms at discharge and was doing well at 4 weeks follow up.

Discussion: CMV is a common viral pathogen with 40-100% of the general population showing prior exposure by serology (IgG antibodies). It is usually acquired early in life and remains latent and reactivates when host immune system is compromised. Majority of individuals with acute CMV infection will be asymptomatic or have mild flu like symptoms that develop after weeks after primary infection, however about 5-7% of immunocompetent patients can have severe manifestations such as colitis, encephalitis, viral pneumonia, and transaminitis. The best diagnostic test for acute CMV illness is serology for CMV IgM antibodies. Histologic finding of “owl-eye” inclusion bodies on tissue sample is highly specific of CMV organ involvement. In immunocompetent individuals, course is typically self-limiting with good prognosis even without use of antiviral medications. Antiviral treatment is typically reserved for immunocompromised patients because of its potential severe side effects.

Acute CMV infection should be suspected even in immunocompetent patients with mononucleosis like illness and Ig M CMV serology should be checked promptly to establish the diagnosis and provide reassurance and preclude further expensive work-up.
References

Connecticut-Clinical Vignette-Poster Finalist
Ayushi Chauhan, MBBS

Title: Double trouble: Pancytopenia and Aplastic Anemia secondary to Acute Hepatitis A and Felty Syndrome

Authors: Puja Shankar, Yue Lin, Brandon Moore, Ayushi Chauhan, Parin Shah, Ahmed Zaghloul, and Edgar Naut

Introduction: Hepatitis Associated Aplastic Anemia (HAAA) is a rare but potentially fatal complication occurring months after an acute infection of hepatitis. Although all the hepatitis viruses have been implicated in this complication, HBV, HCV and HGV seropositivity has been most commonly observed in reported cases of HAAA with Hepatitis A being a rare cause. Bone marrow exam in HAAA shows features of hematopoietic failure with hypocellularity. On the other hand, Felty syndrome (FS) comprises an infrequently occurring but severe subset of seropositive rheumatoid arthritis (RA) complicated by neutropenia and splenomegaly. Herein the bone marrow reveals myeloid hyperplasia with an excess of immature forms. We report an uncommon case of pancytopenia and aplastic anemia with concomitant Felty Syndrome and Acute Hepatitis A infection.

Case Presentation: A 65-year-old Caucasian female with past medical history of untreated rheumatoid arthritis presented after a mechanical fall and one-week history of melena. She was transferred for further management from another institution after workup for the mechanical fall incidentally revealed significant pancytopenia. Additional history taking revealed decreased appetite and a 15-pound unintentional weight loss over 6 months. Home medications were only NSAIDs for pain. On admission, labs showed WBC of 1000 cells/mL, ANC count of 0.6, Hemoglobin/Hematocrit of 4.9/18.9, Mean Corpuscular Volume of 74.4, and platelets of 135,000. Iron panel studies were within normal range. Further laboratory investigations revealed: positive Hepatitis A IgM, negative Hepatitis B Surface Antigen and Core IgM, negative Hepatitis C Antibody, positive Antinuclear Antibody, positive Rheumatoid Factor [>19000], negative HIV, and positive EBV IgG/negative EBV IgM. Abdominal Ultrasound showed mild splenomegaly measuring 14.9 cm and CT Abdomen/Pelvis/Chest revealed no features suggestive of malignancy. A bone marrow biopsy was done that showed decreased cellularity with suppression of tri-lineage hematopoiesis without signs of leukemia, lymphoma, or myelodysplastic syndrome, thought to be consistent with drug-induced or auto-immune myelosuppression. Subsequently, she received Granulocyte Colony Stimulating Factor 300mcg once daily for three days with improvement in WBC to 7.8, ANC to 7.2. She was then switched to IV steroids and methotrexate for treatment of FS with discontinuation of GCSF.

Discussion: Our case was unique secondary to multiple confounders to the diagnosis of pancytopenia including active hepatitis A, GI bleed and Felty Syndrome. Among these, Hepatitis A and FS were the likely culprits for bone marrow suppression. The splenomegaly and significant RF seropositivity increased the likelihood of FS, however, the bone marrow findings were more indicative of viral hematopoietic suppression suggesting hepatitis A-induced aplastic anemia. Treatment of Hepatitis A being purely supportive, it would be interesting to consider whether the same applies to a severe complication such as HAAA and whether use of Eltrombopag herein would be beneficial. Another learning objective would also be the use of LFTs and hepatitis panel as a first, cost-effective, diagnostic step in the evaluation of pancytopenia of uncertain origin.
Title: Triple Native Valve Endocarditis Due to Serratia *marcescens*

Authors: Chad Conner MD, Peter Montesano MD, Deepak Vaderha DO, Nihar Shah MBBS

Introduction: Infective endocarditis is an infection of the endocardial lining of the heart, typically involving prosthetic or native hearts valves. Typical organisms include *S. aureus*, viridans streptococci, *S. galloyticus*, HACEK organisms and enterococci. In one study infective endocarditis due to a non-HACEK organism averaged only 1.8%, and only 0.14% of cases are due to Serratia *marcescens*. Since 1980 only 20 case of *S. marcescens* endocarditis have been described.

Case Presentation: A 35-year-old male with a medical history of hepatitis C, trauma induced splenectomy and intravenous drug abuse presented to the emergency room with altered mental status and recent fall with head trauma. The patient had been feeling ill for a week prior to presentation experiencing very high fevers, nausea, vomiting and chest pain. In the emergency department he was noted to be febrile, temperature 102.8, CT scan of his head showed a subarachnoid hemorrhage, and lab values showed thrombocytopenia (platelets 26), elevated INR 4.3, and a WBC count 46.3. He was in septic shock, hypotensive with a wide pulse pressure and tachycardic not responsive to IV fluids requiring vasopressors. Physical exam was significant for altered mental status, diastolic cardiac murmur and track marks in the antecubital fossas. A bedside trans-thoracic echocardiogram reviled severe aortic insufficiency, and aortic, mitral and tricuspid valve vegetations. He was a poor candidate for transesophageal echocardiography given his liver dysfunction and increased risk of bleeding. Infectious disease was consulted and he was continued on vancomycin and ceftriaxone. During his first day in the intensive care unit (ICU) he became increasingly tachypneic requiring intubation. That night he developed a progressively worsening 1st degree AV block, PR interval 334. Day 2, his blood cultures grew gram negative lactose fermenting rods and his antibiotic regimen was switched to monotherapy with cefepime. Cardiothoracic surgery deemed the patient not a surgical candidate due to his significant coagulopathy and multiple valve involvement. Day 3, given his progressive 1st degree AV block, thrombocytopenia not responsive to antibiotics or platelet transfusions, multiple valve involvement, and not being a surgical candidate his mother decided to change his code status to CMO. Postmortem his blood cultures finalized growing Serratia *marcescens*, sensitive to cephalosporin’s.

Discussion: Endocarditis due to *S. marcescens* is uncommon and indistinguishable from that of other organisms without positive blood cultures. Of the 20 cases described 18 were associated with chronic illnesses, recent surgeries, intravenous drug use or immunocompromising conditions. Our patient had two risk factors, IV drug use and immunocompromised state. *S. marcescens* should remain a consideration for all intravenous drug using patients presenting with signs and symptoms of infective endocarditis, as it is commonly found in water used to mix and inject drugs. In our case having not being a surgical candidate and despite appropriate antibiotic coverage the patient’s disease rapidly progressed. It is important start early broad spectrum antibiotics and to explore all treatment options with an interdisciplinary team for best patient outcomes.
Connecticut-Clinical Vignette-Poster Finalist
Pankil Desai

Title: Isolated Third Cranial Nerve Palsy as a Presenting Sign of Pituitary Apoplexy: A Case Report and Review of the Literature

Authors: Pankil Desai, MD1, Sadiya Thermidor, MD1, Bismurta Misra, MD2, C. Cory Rosenstein, MD3, Department of Internal Medicine, Endocrinology and Neurosurgery

Introduction: Pituitary apoplexy is most commonly characterized by the sudden onset of headache, visual impairment, ophthalmoplegia, altered mental status, and hormonal dysfunction secondary to acute hemorrhage into or infarction of the pituitary gland/tumor. In the majority of cases, it is associated with a pituitary tumor that had not been previously diagnosed. It classically presents with a visual field defects (bitemporal hemianopsia). Ophthalmoplegia can occur later in the course of the disease. A rapid rise in intrasellar pressure leads to compression of the cavernous sinus, with secondary compression of the cranial nerves. We present a case of pituitary apoplexy presenting as isolated third nerve palsy. There are less than 10 reported cases worldwide.

Case Presentation: A 60 year-old male with history of hypertension, obstructive sleep apnea and hyperlipidemia presented to the emergency department with the sudden onset of severe frontal headache radiating to his left eye associated with photophobia, nausea and vomiting. Patient denied any prior history of migraine or ictal headaches. Initial neurologic exam was unremarkable. On presentation, electrolytes, thyroid and prolactin levels were normal. CT of head showed a 2.5 cm pituitary macroadenoma with compression of the left cavernous sinus and carotid artery. MRI of the brain showed hemorrhagic pituitary macroadenoma consistent with pituitary apoplexy with tumor extension into both cavernous sinus and abutting optic chiasm. During hospital course, the patient reported seeing “halos and rainbows” upon leftward gaze and noted to have partial left eye ptosis with dilated, sluggish left pupil, at which time a repeat CT showed no interval change. Subsequently, the patient developed binocular diplopia and complete left-sided ptosis. Vascular imaging and lumbar puncture were unrevealing. Repeat labs were significant for hyponatremia, hypothyroidism and adrenal insufficiency suggesting panhypopituitarism therefore glucocorticoid therapy was initiated. The patient underwent semi-emergent transsphenoidal resection of pituitary adenoma complicated by transient diabetes insipidus post-operatively. His headache resolved immediately post-operatively with rapid improvement of ptosis and diplopia within 24 hours.

Discussion: Isolated third nerve palsy as a presenting sign of pituitary apoplexy is extremely rare. It can mimic a number of other intracranial processes such as posterior communicating artery aneurysm, intracranial hemorrhage, bacterial meningitis and cavernous sinus thrombosis. Hence, making the diagnosis can be extremely difficult. Pituitary apoplexy should be considered early in the differential diagnosis of acute onset isolated third nerve palsy. The anatomic location of the third cranial nerve in the superior and lateral cavernous sinus makes it more susceptible to the laterally transmitted pressure by an expanding pituitary mass. The definitive treatment in these cases is expeditious surgical decompression of the tumor and glucocorticoid to avert an adrenal crisis. Early diagnosis and prompt surgical intervention can restore pituitary function and result in optimal clinical recovery of third cranial nerve function.

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Connecticut-Clinical Vignette-Poster Finalist
Arun Gautam, MBBS

Title: Nivolumab induced Diabetic Ketoacidosis.

Authors: Arun Gautam, MBBS. Department of Medicine, University of Connecticut Health Center, Farmington, CT, Richa Bhardwaj, MBBS. Department of Medicine, University of Connecticut Health Center, Farmington, CT

Introduction: Nivolumab is an anti-programmed cell death protein-1 (PD-1) antibody, which blocks the binding of PD-1 to programmed cell death ligand-1 (PDL-1) that would otherwise inhibit T cell activation. In the recent years few cases of Nivolumab associated type 1 diabetes mellitus have been reported. We report a case of new onset insulin dependent diabetes mellitus (IDDM) presenting as Diabetic ketoacidosis (DKA) after initiation of Nivolumab in a patient with metastatic clear cell renal carcinoma.

Case Presentation: 56-year-old female with history of renal cell carcinoma (RCC) presented to us with polyuria and polydipsia for 5 days. The patient was first diagnosed with RCC and underwent left sided nephrectomy in 2007, her disease was in remission until she had a recurrence in 2014 with multiple pelvic metastasis. She was initially started on a Pazopanib based chemotherapy regimen but was switched to Nivolumab in 2017 due to continued disease progression. In the ED patient was found to be hyperglycemic with serum glucose level up to 529 mg/dl. Further lab work up revealed a high anion gap (19) metabolic acidosis. She was also found to have positive serum and urine ketones and her hemoglobin A1c was 7.1. She was started on an insulin drip and admitted for further treatment of DKA. Work up was negative for Insulin Autoantibodies, Glutamic Acid Decarboxylase (GAD-65) antibody and Islet Cell IgG Cytoplasmic autoantibodies. Patient improved symptomatically and clinically and she was transitioned to a subcutaneous insulin regimen. Nivolumab was discontinued after Discussion: with oncologist. Patient was subsequently discharged home with a close outpatient follow up.

Discussion: Immune check point inhibitors are a popular choice to treat advanced cancers currently due to their durable and survival benefit. These drugs target receptors which provide inhibitory signals to T-cells, thus amplifying T-cell activity in an attempt to generate anti-tumor immune response. Due to the above mechanism of action these drugs are associated with immune related adverse events (irAE), such as type 1 diabetes mellitus (T1DM), thyroiditis, adrenal insufficiency and pneumonitis. T1DM Results: likely from destruction of pancreatic beta cells by auto reactive T-cells. Our patient tested negative for insulin antibodies, other similar case reports have not found a consistent association with antibody detection. It is likely that our patient had prediabetes that was unmasked due to the increased destruction of pancreatic B cells due to the use of Nivolumab causing her to eventually become symptomatic.

With the advent of these new chemotherapy regimens it is pertinent to be aware of these IRAE’e so that we can identify these complications and institute prompt therapy when needed.

References

Connecticut-Clinical Vignette-Poster Finalist
Abhas Khurana, MBBS

Title: Stroke as initial presentation of left ventricular non-compaction cardiomyopathy (LVNC)

Authors: 1. Abhas Khurana, Internal Medicine, University of Connecticut, 2. Rohini Manaktala, Internal Medicine, University of Connecticut, 3. Subhendu Rath, Neurology, University of Connecticut

Introduction: Left ventricular non-compaction (LVNC) is a relatively rare cardiomyopathy, frequently complicated by heart failure, arrhythmia, and thromboembolism. The echocardiographic diagnosis is often challenging and could easily be missed during stroke work-up. The aim of this report is to expand the awareness and understanding of the association between LVNC and strokes, highlighting the relevant literature about the role of anticoagulation and its indications in the syndrome.

Case Presentation: A 61-year-old female with past medical history significant of hypertension, hyperlipidemia and tobacco use who presented with sudden onset difficulty speaking and right sided weakness. Her vital signs were stable. Physical exam was significant for global aphasia, left gaze preference, flattening of the right nasolabial fold, right-sided hemiparesis, with right-sided positive Babinski reflex. Her initial Head CT scan ruled out intracranial hemorrhage, and she was administered IV-TPA. MRI brain showed multiple embolic infarcts. During stroke work-up, trans-thoracic echocardiography suggested LVNC with EF of 15-20%, later confirmed by cardiac magnetic resonance imaging showing a prominent layer of noncompacted myocardium in the left ventricle more so in the mid to apical regions with the ratio of noncompacted to the compacted myocardium of 3:1. Of note, she did not have a family history of cardiomyopathy or sudden cardiac death. Additional history revealed patient had a few episodes of exertional shortness of breath in recent past. She was started on anticoagulation with Warfarin bridged with Lovenox and medical therapy with Carvedilol, Lisinopril, and Spironolactone. The patient continued to improve and was discharged with close cardiology follow up. Later after medication optimization, repeat TTE revealed persistently reduced ejection fraction <35% and so ICD was placed.

Discussion: As illustrated in this case, thromboembolism can be one of the initial presentation of LVNC. Thromboembolism occurs in 13%–24% of patients with LVNC and presents as transient ischemic attack/stroke, renal infarction, mesenteric ischemia, cardiac ischemia and peripheral vascular occlusion. It is important to recognize it during stroke work-up for the institution of appropriate therapy. There is scarce data in the literature about the appropriate indications, risk stratification and efficacy of anticoagulation therapy in LVNC. Current recommendations are based on retrospective case series. Large controlled trials are needed to provide stronger evidence-based guidance derived from larger patient populations.

References

Connecticut-Clinical Vignette-Poster Finalist
Deepthi Kodali, MBBS

Title: Reversal of acquired resistance to anti PD-1 treatment of metastatic melanoma in a patient following discontinuation of Azathioprine treatment for ulcerative colitis

Authors: Kodali Deepthi, MD and Hegde Upendra P., MD; Department of Medicine, Neag Cancer Center, University of Connecticut Health Center

Introduction: Harnessing effector T cell activity against melanoma has become possible through blockade of T cell checkpoints resulting in improved outcomes in the treatment of metastatic melanoma. However, development of resistance to this treatment may result in treatment failures. We report a patient with BRAF wild-type metastatic melanoma receiving immune suppressive therapy for ulcerative colitis, who after initial response to anti PD-1 agent began to develop resistance to treatment. Elimination of azathioprine led to the restoration of sensitivity to anti PD-1 agent. We describe the case and discuss underlying biological mechanisms responsible for this observation.

Case Presentation: A 67-year-old woman receiving immune-suppressive agents prednisone, azathioprine and hydroxychloroquine for ulcerative colitis was treated for BRAF wild metastatic melanoma to the para-duodenal lymph nodes. Past history included surgical excision of a primary cutaneous melanoma over the left cheek 2 years prior (AJCC stage T4bNXM, IIC). Metastatic melanoma was diagnosed on a surveillance CT scan of the abdomen that showed a large para-duodenal mass, confirmed to be metastatic melanoma by a trans-duodenal needle biopsy and seen as FDG avid mass on a whole body PET scan while brain MRI was normal. As the tumor was not amenable to surgical excision due to its large size and location, treatment with immune checkpoint inhibitor was considered. Due to the relative safety of anti PD-1 agent, the patient was started on Pembrolizumab 2mg/kg dose while immunosuppressive treatment of ulcerative colitis was limited to hydroxychloroquine and azathioprine. The patient tolerated treatment without major autoimmune toxicity and a continued response to Pembrolizumab was documented by periodic imaging that revealed the tumor to be reduced to about 6-7mm size over a 14-month period. While Pembrolizumab treatment was continued, repeat imaging study after 3 months showed regrowth of the tumor now measuring 4 x 3.5 cm suggesting resistance to Pembrolizumab treatment. As no other options were considered safe, Pembrolizumab was continued while immune suppressive therapy was changed to eliminate azathioprine. Follow up clinically and with imaging studies revealed restoration of tumor regression without relapse of ulcerative colitis.

Discussion: A large number of T cell clones exist in the body that are directed against diverse tumor neo-epitopes, but their efficacy in anti-tumor immunity is restricted by inhibitory T-cell checkpoints. Although rejuvenated by the immune checkpoint inhibitor treatment, adaptive resistance occurs due to changes in tumor epitope, their presentation to the effector T-cells, functional integrity of effector T-cells as well as the influence of inhibitory tumor microenvironment. Our finding of restoration of clinically relevant anti-melanoma activity in our patient by the elimination of azathioprine treatment exemplifies existence in our patient of metastatic melanoma epitope-specific T cell clones vulnerable to azathioprine suppression that were able to be reactivated by the discontinuation of this immunosuppressive agent.
Title: Can You Have Left Ventricular Thrombus Fourteen Years After a Myocardial Infarction?

Authors: Kurdi, H MD, Anwar, M MD, Ramakar, A MD, Alexander, J MD

Introduction: Left ventricular thrombus (LVT) is frequently seen with acute anterior myocardial infarction. Due to these large infarcts causing subsequent stasis from poor muscle contractility, the risk of thrombi formation increases. Most thrombi form within the first two weeks after myocardial infarction (MI), which increases the risk of embolization [1,2]. To date, there is no record of incidental finding of LVT in surveillance echo without recent cardiac events.

Case Presentation: We present a 77 year old female with past medical history hypertension, hyperlipidemia, coronary artery disease with STEMI status post PCI of mid LAD of anterior wall myocardial infarction fourteen years ago who underwent PCI. Prior echocardiogram in 2016 showed LVEF 40-45% with chronic apical and septal akinesis. On surveillance echocardiogram, the patient was found to have an LVT of 1.8 x 1.5 cm with a new reduced ejection fraction of 30-35% and was sent to the hospital for anticoagulation.

On presentation, she was asymptomatic, hemodynamically and neurologically stable. Physical exam was unremarkable, and her EKG showed more evident T wave inversion in the lateral leads. Her troponin was negative. INR, aPTT, and PT, were within reference ranges.

She was started on a continuous infusion of unfractionated heparin and was bridged with warfarin. Her hospitalization stay was uneventful. Follow up echocardiogram six weeks later showed a decrease in the size of the thrombus to 1.5 and 1.3 cm.

Discussion: Large infarct size, LV aneurysm, Apical akinesis is a risk factor for the formation of LVT and are seen post-myocardial infarction. 90 % of LVT are formed within two weeks within acute MI. There are no prior documented cases of LVT formation in the absence of recent cardiac events. Our case report identifies the LVT that presented after fourteen years of an acute cardiac event.

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Connecticut-Clinical Vignette-Poster Finalist
Marianna Mavilia, DO

Title: Metastatic Cholangiocarcinoma in a Patient with Undiagnosed Wilson Disease

Authors: Marianna Mavilia, Aniket Sharma, Kirsten Ek

Introduction: Cholangiocarcinoma (CC) is the second most common primary hepatic malignancy. Although the frequency of malignancy is generally increased in chronic liver disease, CC rarely presents in Wilson disease (WD). The incidence of hepatic malignancy in WD is only 1.2%, with CC accounting for 0.5%.

Case Presentation: A 66 year old male with history of hypertension, diabetes, and compensated cryptogenic cirrhosis presented with acute onset dyspnea and pleuritic chest pain. He denied cough, congestion, fever, chills, orthopnea, PND. Of note, patient was recently diagnosed with metastatic cancer of unknown primary origin after suffering cord compression syndrome due to a metastatic spine lesion. He was undergoing further workup as an outpatient. Past surgical history was significant for laparoscopic cholecystectomy. He had no family history of liver disease or malignancy. Social history revealed extensive travel through Southeast Asia. He denied history of smoking and heavy alcohol use. The patient was afebrile, tachycardic to 125, tachypneic to 24 and hypoxic to 80% on room air. Physical exam was notable for diffuse bilateral rhonchi, with a normal HEENT and neuropsychiatric exam. CTA of the chest showed a 5.8 cm hypodensity in the right hepatic lobe, innumerable punctate pulmonary nodules, and diffuse bone metastasis. His laboratory Results: were remarkable for an elevated ALP to 252 IU/L, with normal AST, ALT and bilirubin. His CEA was 6.7 mg/mL and CA19-9 was 19,653 U/mL. Colonoscopy was negative. PET scan confirmed hypermetabolic uptake in the liver lesion and extensive lesions consistent with metastases throughout the lungs and axial skeleton. A liver biopsy revealed dense desmoplasia and increased mucin production with staining positive for CK 7 and CK 19, consistent with diagnosis of CC. Biopsy also demonstrated findings consistent with Wilson disease, explaining the patient’s underlying cirrhosis.

Discussion: Common risk factors for CC include viral and alcoholic hepatitis, hepatolithiasis, primary sclerosing cholangitis, parasitic liver disease and cystic liver disease. In these conditions, chronic inflammation and bile stasis stimulate cytokines, leading to formation of reactive oxygen species (ROS). ROS damage DNA and allow for neoplastic growth of cholangiocytes. Unlike other etiologies of chronic liver disease, WD has a low prevalence of CC. This may be explained by a protective effect of copper in WD, which acts to both stabilize DNA and inhibit angiogenesis. In this case, WD had not been diagnosed prior to this presentation. The liver biopsy diagnosed CC and incidentally revealed WD. He also did not display any of the typical neuropsychiatric or ophthalmic signs and symptoms of WD. It is possible that the degree of copper deposition in his liver was mild, causing cirrhosis and the chronic liver inflammation that caused his CC. However, it may not have been sufficient to “protect” against development of CC.
Connecticut-Clinical Vignette-Poster Finalist
Ahmed Zaghloul, MBBCH

Title: Lithium induced ventricular tachycardia

Authors: Ahmed Zaghloul, MD; Corina Iorgoveanu, MD; Kathir Balakumaran, MD, Leon Averbuck, DO

Introduction: Lithium serves an important role in treating various psychiatric disorders. However, what has remained constant since medical Lithium’s discovery are the dangerous side effects that improper dosing can result in due to Lithium’s narrow therapeutic window. (1)

Case Presentation: A 63 year old female with diabetes mellitus, hypertension and bipolar disorder, presented with altered mental status, slurred speech and abnormal gait. The patient was admitted two weeks prior for pneumonia and she was treated with intravenous antibiotics. Prior to discharge the patient’s lithium level was subtherapeutic (lithium level 0.5; nml 0.4 to 0.8) and thus her dose was increased to 600 mg BID from 300 mg BID. Upon presentation, her lithium level was checked was noted to be 1.8. In the emergency department, she was found to be in stable monomorphic ventricular tachycardia.(fig.1) She was given 150mg intravenous Amiodarone twice with no effect. She was subsequently cardioverted with 200J and she converted to atrial fibrillation. The patient was emergently started on hemodialysis. Transthoracic echocardiogram revealed normal left ventricular ejection fraction and no wall motion abnormalities. Coronary angiogram revealed normal coronary arteries.(fig.2) The patient did not develop further episodes of ventricular tachycardia and was discharged with discontinuation of lithium.

Discussion: The pathophysiology by which lithium affects the heart is still not that well understood and it appears that Lithium’s effects on cardiac function is multifactorial. One well documented effect of Lithium’s cardiotoxicity is its propensity to induce bradyarrhythmias. It has been proposed that the culprit for this lies in Lithium’s blockage of Na voltage gated channels. Voltage gated sodium channel disruption may cause sinus node dysfunction. However, sinus node blockage does not explain the ventricular tachycardia our patient as well as a handful of others have experienced.(2,3,4) For this, attention may need to be focused on Lithium’s effect on Na/K exchangers. It has been hypothesized that Lithium’s blockage of Na/K channels helps induce the depletion of intracellular potassium levels.(5) It has been well studied that hypokalemia induces t wave flattening and inversions, EKG effects of Lithium that have been for the most part deemed to be benign.(6) We suggest that it is possible that these EKG effects are not in fact benign but may be an early indication of ventricular irritation that may in certain individuals progress to more dangerous ventricular complications if dosing is further increased. It is conceivable that Lithium overdose may, in certain susceptible individuals more sensitive to blockage of Na/K exchangers, cause symptoms parallel to those seen in worsening hypokalemia, ultimately resulting in Ventricular tachycardia. While Ventricular tachycardia is a rare side effect of lithium toxicity, it’s EKG parallels with worsening hypokalemia suggest there may be a more significant relationship between the two than currently understood.
Delaware-Clinical Vignette-Poster Finalist
Ugochukwu Amadi, MD

Title: Legionella Endocarditis with Rapidly Progressive Glomerulonephritis

Authors: Ugochukwu Amadi, Peter Block, Mahmudul Haque, John Donnelly, Department of Medicine, Christiana Care Health System, Newark, DE

Introduction: Legionella-associated endocarditis is a rare phenomenon. To date, few cases have been reported in the literature1. In turn, there is a limited understanding of downstream complications from this infection. Here, we describe a case of legionella endocarditis complicated by rapidly progressive glomerulonephritis (RPGN).

Case Presentation: The patient was a 49-year old man with a history of congenital aortic stenosis with childhood aortic valve replacement, most recently revised at age 23, and work as a commercial plumber and A/C repair man who was referred to our hospital for acute renal failure after presenting with 4 weeks of intermittent fevers, chest pain, hemoptysis, and hematuria. Initial workup of the patient’s symptoms was unremarkable for most autoimmune or infectious etiologies, including ANCA titers, SLE, RA, HIV, HBV, HCV, M. tuberculosis, Bartonella, C. burnetii, streptococcus spp., staphylococcus, or HACEK organisms. In contrast, urine antigen for legionella was positive on two separate tests. In light of worsening renal function, culture-negative endocarditis with renal complications became a leading diagnosis. Renal biopsy revealed pauci-immune deposits suggestive of ANCA negative vasculitis2 concerning for prosthetic aortic valve association and trans-esophageal echocardiogram (TEE) showed a vegetation on the patient’s prosthetic aortic valve. Thus, a diagnosis of legionella-associated endocarditis with complicating RPGN was made, as the Modified Duke Infective Endocarditis Criteria were satisfied with the patient’s clinical and laboratory findings. The patient was subsequently placed on a 6-week course of azithromycin and levofloxacin for treatment of legionella. He was also started on hemodialysis, as well as an immunosuppressive regimen with Cytosan and corticosteroids based upon the CYCLOPS Trial3. He was ultimately discharged in clinical stability with completion of his infectious and renal treatment regimens as an outpatient, as well as follow-up with cardiothoracic surgery for potential valve replacement.

Discussion: This case describes both an unusual presentation and previously unreported complication of legionella endocarditis. Whereas most prior reports occurred during the perioperative period of cardiothoracic surgery4, the patient described here developed extra-pulmonary manifestations of legionella decades after his last valve replacement. Moreover, this is the first report, to our knowledge, of RPGN secondary to legionella-associated endocarditis4. Thus, this case highlights legionella as a viable cause of culture-negative endocarditis, illustrating a novel downstream complication of this infection.

References

Title: Swig and a Miss? Acute intoxication and diagnosis of neurosyphilis, many forgotten facts

Authors: Sanjana Bhatia-Patel DO, David M Cohen MD, Kevin K Patel MD, Jenifer Goldstein MD

Introduction: Syphilis is a chronic infection involving multiple organ systems caused by Treponema pallidum. Neurosyphilis is defined as any involvement of the central nervous system with this bacterium.

Case Presentation: This is a 54 year-old gentleman with a history of alcohol use who presented to the hospital with acute change in mental status. He had two weeks of confusion and nonsensical speech with visual and auditory hallucinations. He denied any constitutional symptoms, suicidal, or homicidal ideation. Social history included drinking six cans of beers daily and multiple sexual partners. Polydipsia was observed. Laboratory studies showed sodium of 110 mmol/L and positive blood alcohol level. CT and MRI of the head were unremarkable. Initial evaluation also included serum RPR, B12, folate, Lyme titer, and urine toxicology screen all of which were within normal limits. Hyponatremia corrected appropriately without improvement in mentation. Cerebrospinal fluid revealed CSF WBC 10 cumm, elevated protein of 97mg/dL, and CSF glucose of 42mg/dL which was noted by the primary team to be abnormal but not diagnostic for any particular condition. CSF VDRL was negative. Psychiatry was consulted for persistent psychosis however the mental health providers doubted the working diagnosis of primary psychosis due to the rarity of this condition presenting in the sixth decade of life. Repeat serum RPR was found to be positive at 1:32 (confirmed by T. pallidum particle agglutination assay). A diagnosis of neurosyphilis was confirmed. The patient was started on IV penicillin G therapy for a total of 10 days to treat neurosyphilis followed by 3 weekly IM bicillin LA injections for syphilis of unknown duration. His mental status ultimately improved.

Discussion: This case illustrates the challenge of diagnosing neurosyphilis: Despite appropriate clinical suspicion and the use of diagnostic testing the ultimate diagnosis was delayed by confounding variables. It is agreed that serum RPR has outstanding sensitivity (78-86%) and specificity (95-99%) across all stages of syphilis. However, a well-documented cause for a false negative result is acute alcohol intoxication. This detail of testing is often forgotten. Further, acute psychosis complicated by acute hyponatremia and psychogenic polydipsia has been previously described in cases of neurosyphilis. Given the complexity and diversity of CNS findings caused by T. pallidum the diagnosis of neurosyphilis is very inclusive: positive treponemal specific test plus either any CSF abnormality or positive CSF VDRL. This study well illustrates the following conclusions: Firstly, the general internist should be reminded that acute psychosis typically presents in the second and third decade of life. Secondly, neurosyphilis can develop at any stage of the disease. Lastly, neurosyphilis should be considered in the diagnosis of acute psychosis and the internist should be vigilant in his efforts to exclude this protean illness.
District of Columbia-Clinical Vignette-Poster Finalist
Muhammad Ahsan, MBBS MD

Title: Granulicatella endocarditis presenting with migratory arthralgias

Authors: Muhammad Ahsan, MD, Saira Farid, MD, Muhammad Rizwan Sohail, MD

Introduction: Granulicatella species, part of the nutritionally variant streptococci (NVS), are a rare but life-threatening cause of infective endocarditis (IE). Diagnosis may be delayed, leading to formation of large, bulky vegetations and consequent embolic lesions to brain and other organs.

Case Presentation: A 62 year old man with past medical history of hypertension, hyperlipidemia, and right hemilaminectomy for ruptured disk, presented with progressive shortness of breath on exertion of 6 months duration. He reported fevers, night sweats, chills, severe joint pains, generalized weakness, and 20 pounds unintentional weight loss over the past two months. He denied any chest pain, headaches, changes in vision, focal weakness, orthopnea, or new rashes. Review of symptoms was remarkable for intermittent, selflimited, left-sided abdominal pain associated with three episodes of non-bloody, nonbilious vomiting one day ago. His examination was significant for an apical holosystolic murmur. Laboratory investigations revealed a hemoglobin of 10 g/dl, white count of 10.7 x 109/L, B-type natriuretic peptide (BNP) of 732 ng/L, AST 17 U/L, ALT 13 U/L, Cr 0.9 mg/dL, and UA showing only trace hemoglobin. Transesophageal echocardiogram (TEE) showed flail posterior mitral leaflet and one large vegetation (2.5x1 cm) on the posterior leaflet and a smaller (1.6x0.3 cm) vegetation on the anterior leaflet with severe mitral regurgitation. Abdominal CT showed new focal splenic infarcts, and a brain MRI revealed subacute infarcts, consistent with embolic phenomenon. Blood cultures grew Granulicatella elegans. Patient underwent mitral valve replacement surgery followed by six weeks of parenteral therapy with vancomycin and gentamicin. He was doing well at one-month follow-up.

Discussion: Granulicatella species, are members of the nutritionally variant streptococci. These organisms are characterized by their dependence on pyridoxal or cysteine supplementation for growth in standard blood culture media. These organism are part of oral and a frequent cause of dental plaque. They frequently colonize human gastrointestinal as well. G. elegans is a very fastidious organism among all species of Granulicatella and a rare cause of endocarditis (12 reported cases in literature). Clinical presentation can be very non-specific. Our patient had prominent joint symptoms and muscles aches and was initially referred to rheumatology for evaluation of suspected polymyalgia rheumatica. However, presence of a murmur and echocardiographic evidence of vegetations led to diagnosis of IE. Mitral valve is the most frequently infected valve with Granulicatella species. These organisms tend to form large, bulky vegetations and are associated with high risk of embolic complications (up to 49% in one series). Reported mortality rate is 20%. The American Heart Association guidelines suggest ampicillin or penicillin G plus gentamicin for 4 to 6 weeks or vancomycin as an alternative regimen for Granulicatella IE. Prolonged antimicrobial therapy combined with valve replacement surgery (as performed in our case) is frequently required to cure these infections.
District of Columbia-Clinical Vignette-Poster Finalist
Rachel Denyer, BMBch MD

Title: Septic shock due to Citrobacter amalonaticus bacteremia and urinary tract infection.

Authors: Rachel Denyer BMBch, University of Maryland Prince George's Hospital Center., Sabeen Khan MBBS, University of Maryland Prince George's Hospital Center.

Introduction: We present what we believe is the first documented case of bacteremia and septic shock due to *Citrobacter amalonaticus* urinary tract infection. Infections caused by *Citrobacter* are rare, and the species is considered low virulence. *Citrobacter* is associated with significant morbidity and mortality due to its long survival period in hosts and frequent resistance to antimicrobials. Clinical manifestations include urinary tract infections, cerebral abscess, meningitis, intra-abdominal infections, pneumonia, and bacteremia. Infections due to *C. amalonaticus* comprised only 4 of 111 *Citrobacter* strains identified in one retrospective study (Manganello et al. 2001). There is one previous report in the medical literature of *C. amalonaticus* bacteremia in Taiwan (Lai et al. 2010). There is also a case series of four patients from France with *C. amalonaticus* urinary tract infection without bacteremia; all of the patients in question had either urinary tract abnormalities or were renal transplant recipients (Garcia et al. 2016).

Case Presentation: A 73 year-old male, with a history of recurrent urinary tract infections due to benign prostatic hypertrophy, was brought to our hospital by family members with a one-day history of fever, rigors and altered mental status. He had also developed urinary incontinence over the preceding months. His other medical history included diabetes mellitus and chronic alcohol excess. In the emergency room he was febrile at 103.2F, tachycardic at 120 bpm, and hypotensive at 94/52. On examination, he was oriented to self alone, not to person or place, and the medical team witnessed the patient having rigors. Chest, cardiovascular, abdominal skin and neurological exams were otherwise normal. Urinalysis was positive for nitrites and leukocyte esterase, microscopy revealed 6-10 white blood cells per hpf, and moderate bacteria.

The patient was treated empirically for sepsis with IV piperacillin/tazobactam and vancomycin. On Hospital Day 2, blood and urine cultures both grew *Citrobacter amalonaticus* and the patient had defervesced so antibiotics were changed to ceftriaxone alone based on sensitivities. He had a peripherally-inserted central venous catheter placed, and went to a nursing facility to complete a 10-day course of ceftriaxone. His altered mental status resolved.

Discussion: *Citrobacter amalonaticus* was first described in 1971 after it was isolated from inpatient fecal samples. It was initially thought to be part of a new genus called *Levinea* but subsequently reclassified. Its natural reservoir is unknown. *Non-freundii, non-koserii* *Citrobacter* species can be misidentified by common automated laboratory identification systems, such as Vitek II and Phoenix. Garcia et al. (2016) documented an apparent increase in *C. amalonaticus* infections following adoption of the MALDI-TOF mass spectrometry-based system for species identification. Improved detection rates will result in an apparent increased incidence of *C. amalonaticus*, underscoring the urgent need to rectify our essential ignorance of the ecology of this species.

References

Title: ‘Fonda’ Failure? A tale of multiple hypercoagulability disorders in the same patient and how to treat them

Introduction: It is a debate we have all had many times. Are we absolutely sure he/she isn’t taking their blood thinner? Or, is this clot a true failure of an anticoagulant? Hypercoagulability disorders encompass a range of syndromes causing patients to have a propensity to form venous, or sometimes arterial, thromboembolism. Depending on the cause of hypercoagulability, most patients are treated with life-long anticoagulation. In those with multiple hypercoagulable disorders, the decision for life-long anticoagulation is an easy one. However, the care of these patients is complicated when they present with recurrent thromboembolism on therapeutic anticoagulation.

Case Presentation: Here, we describe a case of a 23 year-old man with hereditary hypercoagulability who had been recently treated for lower extremity cellulitis, and who presented with acute bilateral clots. He was compliant with fondaparinux therapy, and had been without thromboembolism for >2 years prior to presentation. Vital signs were normal and physical exam showed significant lower extremity edema. Laboratory tests were normal, except for a urinalysis showing >500 protein. A 24-hour collection of urine protein revealed > 4.5 grams of protein excretion. Serologic testing revealed a positive anti-DNAse B strep antibody but negative anti-streptolysin O antibody. All other rheumatologic, hepatitis, and serologic testing was normal. He was treated empirically with high-dose corticosteroids, and his urine protein significantly decreased. Renal biopsy was deferred until acute clot could be stabilized due to his current inability to come off anticoagulation periprocedurally. Because his home dose of fondaparinux was already the maximum daily dose and new clot was formed on this medication, he was switched to therapeutic enoxaparin. Anti-Xa levels were obtained prior to discharge.

Discussion: Patients with multiple hypercoagulable disorders have long been a very difficult cohort to adequately treat. The patient is admitted to the hospital, and quite frequently, the members of the team have the age-old debate of anticoagulant-failure versus noncompliance. For this patient, it was thought that his hereditary hypercoagulable state was previously well-managed on fondaparinux therapy. But, due to his development of nephrotic syndrome, he became more hypercoagulable. In patients with nephrotic syndrome, the risk of venous thromboembolism is ~30-35%, but in this patient with a higher propensity for clots already, this risk is much higher. Schulman, et. al. reported successful avoidance of recurrent thromboembolism with a goal anti-Xa level to >20% above the upper limit of the normal therapeutic range (Blood, 2017). Prior to discharge, this patient’s anti-Xa levels were shown to be at this threshold. The general medicine physician, with the help of his/her hematology colleagues, should feel empowered to dose anticoagulants to therapeutic anti-Xa level. For patients with recurrent thromboembolism on anticoagulation, anti-Xa levels should be assessed and dosing adjusted to an anti-Xa level of 1.2 times the upper limit of normal.

References

District of Columbia-Clinical Vignette-Poster Finalist
Tushina Jain

Title: Advanced Therapy for High Clinical Suspicion Pulmonary Embolism

Authors: Tushina Jain, M.D., George Washington University Hospital, Hind Rafei, M.D., George Washington University Hospital, Raza Yunus, M.D., George Washington University Hospital

Introduction:

Case Presentation: A 74-year-old man, with chronic kidney disease and multiple prior left ankle surgeries with hardware, presented with three days of left ankle pain and shortness of breath. His physical exam revealed hypotension, hypoxia, and respiratory distress. Laboratory workup showed initially normal platelet counts that decreased significantly throughout the hospital stay as well as Staphylococcus aureus bacteremia. Imaging of the left ankle showed extensive deep venous thrombosis (DVT) and likely osteomyelitis. A ventilation-perfusion (V/Q) scan showed low probability for PE. A transthoracic echocardiogram showed new, severe right heart strain with normal left ventricular (LV) function.

The patient received pressor support, antibiotics for presumed septic shock due to osteomyelitis and infected hardware, and heparin infusion for the DVT. A few days after initial presentation, the diagnosis of massive PE was contemplated due to continued deterioration and the clinical findings.

Intravascular ultrasound (IVUS) was used to evaluate the pulmonary arteries (PA). IVUS of the proximal main PA did not reveal an embolus; however, the mean PA pressures were markedly elevated, 50mm Hg. Pigtail catheters with tissue-plasminogen activator (t-PA) were placed in the main PA.

After 16 hours of therapy, the catheters were removed. The patient had worsening hypotension again and suffered multiple pulseless electrical activity arrests resulting in death.

Discussion: This patient’s initial diagnosis was sepsis, and possible obstructive shock due to massive PE was overlooked given the V/Q scan result. However, the patient had multiple concerning signs: hypoxia, DVT, and new right heart strain. High clinical probability with a low-probability V/Q scan is indeterminate, with the chance of PE ranging from 6 to 88%. With high clinical suspicion, PE should remain on the differential, especially in critically ill patients when advanced therapy may provide early hemodynamic improvement.

Systemic fibrinolysis is the most studied advanced therapy for massive or submassive PE, defined as PE with hemodynamic instability or right ventricular dysfunction, respectively. With the risk of intracranial hemorrhage though, there is growing interest in targeted fibrinolysis. A recent study of intra-catheter t-PA in 150 patients with massive or submassive PE showed reduction in both RV/LV diameter and mean PA systolic pressure at 48 hours.

In contrast to the patients in this study, our patient did not have a proximal PE confirmed on IVUS. However, given his elevated PA pressures, high clinical suspicion, and risk of perioperative mortality with such high right heart strain, intra-catheter TPA was administered.
Further research is needed to better define the role of targeted TPA in acute PE. This case highlights the importance of weighing risks and benefits in complex patients and early consideration of advanced therapy.

References

Title: A presentation of viral meningitis highlights shortfalls in outdated zoster vaccination guidelines

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Introduction: Opportunistic viral infections are well-defined risks of immunomodulating therapy. Although cases of Varicella Zoster (VZV) have been documented with the use of adalimumab, cases of VZV central nervous system (CNS) infection in patients using adalimumab monotherapy are rare. We present a case of a 53 year-old male with psoriatic arthritis who developed a VZV reactivation as V1 dermatomal herpes zoster presenting with aseptic meningitis after initiation of adalimumab monotherapy. To our knowledge, this is the first case of opportunistic VZV meningitis infection associated with adalimumab monotherapy for psoriatic arthritis.

Case Presentation: A 53 year-old male with psoriatic arthritis on adalimumab monotherapy (40mg/0.8mL SC every two weeks) for three months initially presented to his primary doctor with headache and left eye pain and was treated with acetaminophen and artificial tears. Over the next two days, his headache and left eye pain worsened and he developed chills and night sweats prompting emergency room evaluation.

On presentation, the patient described a diffuse burning headache that worsened with cough. He was hemodynamically stable, afebrile, had no focal neurologic deficits, and his cranial nerves were intact, but he had positive meningeal signs with neck flexion. Vesicular lesions were present on the bridge of his nose and superior to his left brow. The left eye demonstrated blepharitis and episcleritis.

CBC, CMP and noncontrast head CT were unremarkable. CSF analysis revealed 112 WBCs/hpf with 92% lymphocytes, protein 87 mg/dL, glucose 80 mg/dL, negative bacterial culture, and positive VZV PCR. The patient was started on weight based IV acyclovir, and adalimumab was discontinued. He improved clinically and was discharged four days later on oral valacyclovir to complete a 14 day antiviral course. The patient recalled having chickenpox as a child, and chart review revealed no history of zoster vaccination.

Discussion: TNF-alpha blockers are known to increase risk for VZV reactivation. This case demonstrates that even monotherapy can result in severe CNS infection, which is associated with significant morbidity and mortality. Herpes zoster vaccination reduces the risk of reactivation as well as the degree of symptoms manifested in reactivation. The administration of zoster vaccination to patients already on immunomodulating therapy is contraindicated due to the infectious risk associated with live vaccines. However, herpes zoster vaccination prior
to initiating TNF-alpha blocker therapy is not specifically recommended by CDC or IDSA. This case, and the increasing literature base, suggests a role for zoster vaccination prior to the initiation of TNF-alpha blockers.
Title: Left Ventricular Noncompaction Cardiomyopathy: A Rare Case Presentation

Authors: Christopher J. Bitetzakis, Department of Medicine, University of South Florida, Tampa, FL, Emmanuel Bassily, Department of Medicine, University of South Florida, Tampa, FL, Meghana Vellanki, Department of Medicine, University of South Florida, Tampa, FL, Jessica Huang, Department of Cardiology, University of South Florida, Tampa, FL

Introduction: Often described as a genetic cardiomyopathy, a thorough history is of utmost importance and left ventricular noncompaction cardiomyopathy should be considered in all individuals, especially younger patients, who present with symptoms of heart failure.

Case Presentation: A 28-year-old African-American male presented to our institution with complaints of new-onset dyspnea, pedal edema, gait disturbances and pre-syncope of two days duration. He reported living an active lifestyle having played competitive sports in the past but had developed a progressively worsening functional capacity over the last 6 months. He had no significant past medical history and denied any history of tobacco, alcohol, or illicit drug use. He reported a significant family history of cardiovascular disease but could not recall specific information. Physical examination was remarkable for jugular venous distention, crackles on auscultation of the bilateral lung bases, tachycardia and 2+ bilateral lower extremity edema. Gait and heel to shin coordination were also abnormal. Laboratory studies revealed a Troponin I of 5.649 nanogram/milliliter and B-type Natriuretic Peptide of 2705 picogram/milliliter. The patient’s chest radiograph was significant for cardiomegaly. Magnetic resonance imaging of the brain revealed several hypodensities with restricted diffusion consistent with acute infarcts in the cerebellum. Transthoracic echocardiography noted severely dilated cardiomyopathy with prominent left ventricular trabeculations, multiple thrombi, and decreased systolic function with an estimated ejection fraction of 10-15%. Cardiac magnetic resonance imaging revealed noncompacted/compacted myocardium ratio of 3.5. Left heart catheterization revealed a normal, patent coronary anatomy with no significant atherosclerotic disease. His symptoms significantly improved during his hospitalization with diuresis and he was discharged home on optimal medical therapy, a LifeVest for primary prevention, and instructed to follow up with repeat echocardiography in 3 months to assess for improvement and need for implantable cardioverter defibrillator.

Discussion: Left ventricular noncompaction cardiomyopathy, previously described as spongy myocardium and hypertrabeculation syndrome, is a rare type of cardiomyopathy characterized anatomically by prominent ventricular trabeculations and deep intertrabecular recesses that communicate with the left ventricular cavity but not with the coronary circulation. Although left ventricular noncompaction cardiomyopathy can be asymptomatic, patients may also present with heart failure, thromboembolism, and fatal arrhythmias. Morphological features have also been identified incidentally in various healthy populations such as athletes. This case highlights the severe, and potentially fatal, complications that may arise with this cardiomyopathy. Although a relatively rare diagnosis, given significant advances in echocardiography and cardiac magnetic resonance imaging, left ventricular cardiomyopathy has been reported with increasing frequency in the literature.
Title: An Unusual Presentation of Acute Fibrinous and Organizing Pneumonia as Cavitations in the Lung

Authors: Venkatesh Gupta Cheetirala, Zuzana Talbot, Ali Vaziri, Mohamad Eid, Salman Muddassir

Introduction: Acute fibrinous and organizing pneumonia (AFOP) is a recently evolving lung pathology and rarely documented variant of cryptogenic organizing pneumonia, characterized by intra-alveolar fibrin on histology. The presence of intra-alveolar fibrin and lack of hyaline membranes (seen in Diffuse Alveolar Damage) and eosinophils (seen in Eosinophilic Pneumonia) defines the characteristic histological pattern of AFOP. It has been associated with a variety of underlying conditions that include rheumatologic diseases, drug reactions, occupational or environmental exposures, infections and several cases are idiopathic. Clinical course varies from subacute or indolent respiratory illness to rapid onset and progression to fulminant respiratory failure and death. Although cases with lower lobe infiltrates, ground glass opacification and upper lobe infiltrates have been documented so far, AFOP presenting as cavitations in lung is an infrequent and a distinguished finding.

Case Presentation: A 74 years old female with past medical history of COPD presented with five days history of high grade fever up to 103.8F, dyspnea, productive cough with hemoptysis, chills and night sweats. On physical examination she had decreased breath sounds in the right lower lobe. WBC on admission was 16.8. Chest x-ray showed right lower lobe airspace disease. CT chest without contrast showed 9.3 cm consolidation in right lower lobe with multiple areas of cavitation. Bronchoscopy with BAL was performed, and copious amount of purulent material was suctioned. Sputum cultures, blood cultures as well as AFB and cultures from bronchial wash were all negative. CT guided biopsy of right lung lesion was obtained. Patient’s symptoms significantly improved on empiric antibiotics, her leukocytosis resolved, and patient was discharged home on antibiotics and home oxygen. After discharge, the pathology of lung biopsy revealed acute fibrinous and organizing pneumonia without hyaline membranes and eosinophils. The GMS stains for fungus and an AFB for acid fast bacilli were negative. No malignant cells were identified.

Discussion: AFOP is a relatively new diagnosis with less than 120 cases reported in the literature until 2015. It is a very rare condition with a poor prognosis, accompanied by delay in diagnosis. Our case adds to the literature a new and unusual finding of multiple cavitations in AFOP which commonly presents as bilateral lower lobe infiltrates. Our case showed symptomatic improvement with antibiotic treatment suggesting a possible role of antibiotics in management of AFOP in addition to benefit from steroids and immunosuppressive therapy in some patients. This uncommon presentation warrants to be considered in the differential when encountering consolidation with multiple cavitary lesions on chest imaging. Currently, newer modalities of treatment for AFOP are under investigation.
Title: Catheter-Directed Thrombolysis For The Resolution Of A Large Device-Related Right Atrial Thrombus, A Case Report

Authors: Marcelo Fernandes, MD/MPH; Salih Greviious, MD; Alexandre Ferreira, MD

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Introduction: Implantable venous access devices (IAD) are commonly used in patients with sickle cell disease (SCD) for long-term red cell exchange. Though IADs are routinely used in the clinical setting, right atrial thrombus is a rare but potentially life-threatening complication of these devices. Current therapeutic options include systemic anticoagulation, surgical thrombectomy, and systemic thrombolysis. However, Results: stemming from these procedures are mixed, and there is currently no consensus on the optimal treatment of a device-related right atrial thrombus. We describe the case of a 25-year-old man with SCD who developed a large right atrial thrombus related to an IAD and who was successfully treated with catheter-directed, low-dose recombinant tissue plasminogen activator (tPA). To our knowledge, this is the first case in the literature describing complete resolution of a large right atrial thrombus with localized tPA in a patient with SCD.

Case Presentation: A 25-year-old man with SCD was transferred to our institution from an outside hospital after initially presenting with shortness of breath, at which time a transthoracic echocardiogram (TTE) revealed a large right atrial thrombus measuring 3.0 x 1.4 cm. He was initially started on intravenous heparin continuous infusion, with only modest reduction in thrombus size after 5 days of heparin infusion (2.5 X 1.6cm). Given the patient’s many comorbidities, his perioperative risk was too high to undergo right atrial thrombectomy. He was subsequently treated with an 18-hour infusion of tPA at 2mg/hour directly through the IAD. A follow-up TTE obtained at the conclusion of the infusion showed complete clot resolution. There were no complications related to the procedure, including major bleeding.

Discussion: The optimal management strategy for patients with a right atrial thrombus from an implantable device remains controversial. Although more investigation in the use of this novel therapy for the treatment of device-related right atrial thrombus is needed, we believe it can serve as a potentially alternative treatment option in select patients for whom current treatment modalities are not desirable or are less efficacious.
Title: Creutzfeldt-Jacob Disease

Authors: Jeffrin Joseph MD1; Priya Sharm’a B.S.2; Claudia Vallin B.S.2, 1. PGY3 resident - Department of Internal Medicine at Mount Sinai Medical Center, Miami Beach, FL, 2. Medical Student - Nova Southeastern University, Fort Lauderdale, FL

Introduction: Prion diseases or spongiform encephalopathies are neurodegenerative diseases that have protracted incubation periods and progress inevitably once clinical signs appear. Diseases currently recognized in humans are: Creutzfeldt-Jakob disease (CJD), variant Creutzfeldt-Jakob disease (vCJD), Kuru, Gerstmann-Sträussler-Scheinker syndrome (GSS), and fatal familial insomnia (FFI).

Case Presentation: 66 year old female with no medical history living in the Cayman Islands presented with unexplained left sided weakness with occasional jerking movements of her left arm and progressive cognitive decline for 1 month. Her past surgeries were 2 C-sections and a hysterectomy. Initial work up by two neurologists at the island included brain MRI’s which revealed some progression of restricted diffusion of the right cortex, EEG with delta waves bilaterally and a nerve conduction study with a possible cervical radiculopathy.

Due to her continued disease progression and worsening she was referred here to the U.S for further work up.

On arrival she had normal vital signs, exam revealed a mask like face, alert and oriented yet occasionally having an inattentive disposition. Neurological exam revealed a rigid flexed left arm and left leg with 3/5 muscle strength, 3+ reflexes, truncal ataxia and an abnormal gait needing assistance. Primitive reflexes were positive including startle with a myoclonus, glabellar tap and palmomental reflex. Differential diagnosis at the time included JC virus, Herpes encephalitis, HIV, HTLV, Wilson's disease, stiff man syndrome, rapidly progressing Parkinson's disease, CJD, and paraneoplastic encephalomyelitis. Due to her rigidity/extrapyramidal features she was started on baclofen and a carbidopa/levodopa combination which did not lead to any improvement. MRI brain revealed increase in the restricted diffusion of the right more than left parietal lobe, right insular cortex and cingulate gyrus. MRI cervical spine, thoracic spine and EEG were insignificant. She developed dysphagia, gradual mental status deterioration with confusion and inability to ambulate during the 11 day stay at the hospital. All blood and CSF tests for the various differential diagnosis came back negative except for the 14-3-3 protein from the CSF which was elevated at 4.5ng/ml (Reference <2.0) which clinched the diagnosis of sporadic CJD. She continued to decline and was taken home on request of family where she became comatose and died a month later.

Discussion: Sporadic CJD (sCJD) is a rare condition with approximately one case occurring per 1,000,000 population per year with a worldwide distribution. Although case control studies do show an association of sCJD with prior surgery, it is unknown if her multiple C-sections or hysterectomy played a role in transmission. However, it is important to consider CJD or other prion diseases in patients who received human pituitary growth hormones/gonadotropins, corneal, liver or dura graft transplants and neurosurgical procedures with rapid neurological deterioration, myoclonus and positive startle reflex. Newer tests such as RT QuIC (Real time quaking induced conversion) are prion protein conversion assays with increased sensitivity and a specificity of 98%.

References
Florida-Clinical Vignette-Poster Finalist
Hyun Woo Kim, MD

Title: Pemphigus Foliaceus: a Rare and Misdiagnosed Condition

Authors: Hyun W. Kim, MD1; Josiah D. McCain, MD1; Jason C. Sluzevich, MD2; Ricardo J. Pagán, MD3, Division of Internal Medicine, Mayo Clinic Jacksonville, FL, Division of Dermatology, Mayo Clinic Jacksonville, FL

Introduction: Pemphigus is a rare autoimmune blistering disorder involving the skin and mucosa. It is clinically characterized into two main types: pemphigus vulgaris (PV) with both mucous membrane and cutaneous involvement and pemphigus foliaceus (PF) which is solely skin limited. These differing clinical presentations reflect differences in pathogenic auto-antibodies production with desmoglein-1 being found in PF, while both desmoglein-1 and desoglein-3 autoantibodies are seen in PV. PV is the most frequent presentation, accounting for about 70% of all pemphigus cases with an estimated incidence of 1 to 5 cases per million per year worldwide. PF is uncommon with an incidence of less than 1 case per million per year.

Case Presentation: 65 year old man with history of pemphigus vulgaris who was evaluated for persistent skin erosions with surrounding erythema, resistant to his immunosuppressive therapy. He had been diagnosed with PV 5 year ago, having multiple disease flares over the first two years while on therapy with mycophenolate mofetil and prednisone. Continued flare ups led to modification of therapy and substitution of mycophenolate for cyclosporine without significant improvements. He denied noncompliance with the medications or making any particular lifestyle modifications, such as moving to different cities, changing his diet or exercise pattern, any new medications, or any identifiable life stressors. Apart from his skin complaints, the rest of the review of systems was negative. On physical examination: vital signs, cardiovascular, lung, and abdominal examinations were normal. There were extensive superficial exfoliations over face, scalp, trunk, back, and all extremities as well as some thin, crusted plaques that resembled lesions in different stages of healing. There was no mucosal or genital involvement. Nikolsky sign was negative. Laboratory evaluation revealed a macrocytic anemia [hemoglobin 10.4g/dL (13.5-17.5g/Dl) and hematocrit 96.2 fl (81.2-95.1fl)] with normal electrolytes and renal function. A 4 mm punch skin biopsy revealed extensive eosinophilic spongiosis. Perilesional direct immunofluorescence (DIF) revealed superficial intraepidermal deposition of IgG and C3. Desmoglein 1 titers were 228.9 U (<14U) with normal desmoglein 3, consistent with the diagnosis of PF. As this is was deemed refractory pemphigus foliaceus, treatment was initiated with intravenous immunoglobulin (IVIG) infusion over three days, followed by monthly rituximab. Patient has received three rituximab infusions so far and has not experienced a flare-up in three months.

Discussion: This case illustrates the importance of reconsidering and broadening the differential diagnosis based on complete history and physical exam. As a result, correct diagnosis of PF was made and appropriate therapy was instituted immediately to treat the refractory pemphigus. The presence or absence of mucosal involvement, DIF studies, and ELIZA studies for desmoglein 1 and 3 levels can aid in proper pemphigus subtyping.
Florida-Clinical Vignette-Poster Finalist
Rohit Kumar, MBBS

Title: An unexpected cause of necrotizing fasciitis: *Vibrio vulnificus* takes wing.

Authors: Rohit Kumar, MD; Kanwarpreet Tandon, MD; Alexandru Zaharcu, MD; Manoj Poudel, MD; Nemer Dabage, MD; Robert Andrews, MD; James DeMaio MD, 1- Department of Internal Medicine, Blake Medical Center, 2- Department of Infectious Disease, Blake Medical Center

Introduction: *Vibrio vulnificus* is typically acquired via two main routes of transmission. Either patients with iron overload or immune suppression may experience bacteremia then secondary necrotizing skin infections after ingesting shellfish. Or both normal and immune suppressed patients may develop necrotizing skin infections with possible bacteremia after direct exposure of open skin lesions to seawater. We present a case of *Vibrio vulnificus* necrotizing fasciitis and bacteremia from an unexpected source.

Case Presentation: Eight days after Hurricane Irma, a 43 year-old male with a history of Peripheral vascular disease and systemic lupus erythematosus on belimumab and steroids injured his leg on a tree branch while cleaning his property. He presented to our facility 5 days after the injury with a very tender, erythematous right calf laceration. The patient denied exposure to seafood, seawater or floodwaters from storm surge. Cefazolin and clindamycin were started empirically. Blood cultures drawn on admission were subsequently positive for gram negative rods and the antibiotics were switched to ceftazidime, ciprofloxacin and doxycycline. Despite this triple antibiotic regimen, the leg wound rapidly worsened over 24 hours with continued high-grade fever, increasing pain and swelling and then frank tissue necrosis. The patient was taken emergently to the OR where he required extensive debridement for necrotizing fasciitis. Both *Vibrio vulnificus* and *Oligella ureolytica* were identified from the admission blood cultures. Despite multiple further debridements and aggressive wound care, patient eventually required below knee amputation due to poor healing secondary to underlying medical conditions.

Discussion: Natural disasters pose both a clinical and a public health challenge. After hurricanes, there have been documented increases in gastrointestinal, respiratory and skin/soft tissue infections. Our patient developed a *Vibrio vulnificus* necrotizing fasciitis with no prior seawater or seafood exposure. His property is 700 meters from a brackish water bayou and 1000 meters from the Manatee River. The property was not affected by storm surge and experienced no flooding. As the eye of the storm passed due east, sustained winds of 85 mph would have been moving from the bayou/river directly towards the patient’s home. We postulate that Hurricane Irma carried brackish water droplets or brackish water contaminated debris inland. Subsequent exposure to contaminated vegetation in the face of immune suppression likely led to our patient’s life-threatening infection. Since Vibrio species are very sensitive to drying, it is surprising that the organism was able to persist inland for more than one week. The extremely wet conditions caused by the storm and the high humidity of subtropical Florida may have prolonged the survival of the organism outside its usual environment. Clinicians should remain vigilant after natural disasters for atypical presentations of unusual water associated pathogens.
Florida-Clinical Vignette-Poster Finalist
Rupesh Manam, MD

Title: Pembrolizumab-Induced Acute Inflammatory Demyelinating Polyneuropathy

Authors: Rupesh Manam MD, Josh Gross MD, Jasmine Martin MD, Dhishna Chaudhary MD, Patricio S. Espinosa MD, Sajeel Chowdhary MD and Edgardo Santos MD

Introduction: Pembrolizumab, is a monoclonal antibody against immune checkpoint programmed death receptor (PD-1) that facilitates apoptosis of cells expressing PD-1. It is FDA approved for treatment of a growing number of oncologic conditions. Reported immunological related adverse events include pneumonitis, colitis, hepatitis, nephritis, and endocrinopathies. Neurological related adverse events are less understood and have been infrequently cited in medical literature. We herein present 2 cases of acute inflammatory demyelinating polyneuropathy (AIDP) as a rare complication of pembrolizumab.

Case Presentation: 73-year-old White male with history of stage IV poorly differentiated lung adenocarcinoma with PD-L1 of 20% was started on carboplatin with pemetrexed plus pembrolizumab. Three-weeks after initiating treatment with pembrolizumab patient developed ascending generalized motor weakness requiring hospital admission. Motor strength was 3/5 bilateral (B/L) upper extremities (UE) and lower extremities (LE) with absent deep tendon reflexes. Lumbar puncture (LP) revealed albuminocytological dissociation in the CSF (68). Intravenous (IV) steroids and intravenous immunoglobulin (IVIG) were initiated. Despite five IVIG infusions, the patient’s motor strength diminished to 2/5 in bilateral UE and LE, negative inspiratory force was -20 and forced vital capacity was 1.1 prompting intensive care unit monitoring. IVIG was discontinued, and patient was started on plasmapheresis; patient received 8 sessions of plasmapheresis as well as steroids at high-dose. After 25 days of hospitalization, patient showed clinical improvement in motor function and was discharged on prednisone. Within one month his condition deteriorated, prompting hospice care.

81-year-old white male with history of stage IV metastatic melanoma with B-Raf positive mutation with metastasis was started on pembrolizumab plus dabrafenib and trametinib. PET-CT scan performed 5 months later revealed extension of metastases prompting a second cycle with pembrolizumab. Patient presented to hospital 5 weeks after with B/L LE quadriplegia. Neurological exam revealed 2/5 UE and 0/5 LE motor strength with areflexia. Lumbar puncture revealed albuminocytologic dissociation (56). He was promptly started on pulse dose solumedrol with IVIG for 5 days. Patient status continued to deteriorate requiring mechanical ventilation due to respiratory failure. Plasmapharesis was initiated but did not show clinical improvement. On day 14, patient had an intracerebral hemorrhage from bleeding brain metastasis prompting hospice consult.

Discussion: PD-1 acts by inhibiting primarily T-cell activation and limits immune effector responses when bound to ligands PD-L1 and PD-L2. Immune checkpoints negatively regulate the immune system to prevent autoimmunity. Monoclonal antibodies targeting PD-1, such as pembrolizumab prevent inhibition of antitumor response, which can lead to autoimmunity. In both cases mentioned above, patients developed AIDP following pembrolizumab. Management included IV steroids and IVIG, with subsequent escalation to plasmapheresis. Given severity of symptoms in these two cases, early recognition of pembrolizumab-induced AIDP is paramount for timely treatment.
Florida-Clinical Vignette-Poster Finalist
Julian A Marin Acevedo, MD

Title: Myositis leading to ventilation impairment: a presenting feature of chronic graft-versus-host disease

Authors: Julian A. Marin-Acevedo, M.D., Roy Vivek, M.D.

Introduction: Myositis is a rare presentation of chronic graft-versus-hot-disease (GVHD) and only few cases have been reported with respiratory-muscle involvement.

Case Presentation: A 47-year-old female presented to our hospital for dyspnea. She had a history of myelofibrosis status-post allogeneic bone marrow transplantation two-years prior complicated by chronic GVHD involving the gastrointestinal tract, liver, and skin. Her current symptoms had started two weeks prior as progressive lower extremity weakness with myalgias followed by worsening shortness of breath. She was only on low-maintenance doses of steroids for her GVHD. On admission she was in moderate respiratory distress and with an oxygen saturation of 87% on room air. Breath sounds were decreased bilaterally and muscular strength was 3/5 in both upper and lower extremities with normal reflexes throughout. Laboratory workup revealed leukocytosis of 18 x10^9/L, normal electrolytes, elevated troponin-T of 0.56 ng/mL, and arterial blood gases consistent with acute hypercapnic respiratory failure for what she was intubated and transferred to the intensive care unit. Creatine phosphokinase (CPK) was 15,000 U/L (38-176) and aldolase was 350 U/L (<7.7). Thyroid stimulating hormone was normal, antinuclear antibody, rheumatoid factor, and anti-Jo1 were negative. Chest x-rays showed diminished lung volumes and a normal heart size. Cardiac magnetic resonance ruled out myocardial involvement. A left quadriceps muscle biopsy confirmed severe inflammatory myopathy which, given her history, was considered consistent with GVHD-induced myositis. She was started on high-dose methylprednisolone. Her muscular strength improved within 48 hours followed by successful extubation and an uneventful subsequent clinical course. She was discharged on a prednisone taper and one month later, her weakness had resolved and her troponin and CPK levels had returned to baseline.

Discussion: Chronic GVHD is a complication of allogeneic bone marrow transplantation that usually affects the skin, oral mucosa, liver/gastrointestinal tract, and lungs.1 Many patients also present with a constellation of symptoms similar to those of other autoimmune diseases. Myositis is a rare presentation of chronic GVHD with an overall incidence of 2-3%.2 Often, symptoms are gradual in onset and resemble idiopathic polymyositis (symmetrical weakness affecting proximal muscle-groups). Respiratory muscle involvement has been exceptionally reported. Diagnosis requires exclusion of other causes of myositis including medications, thyroid, or autoimmune diseases.1,2 Troponin-T can be falsely elevated in the settings of high CPK levels leading to a false perception of cardiac injury.3 Muscle biopsy by itself does not establish a diagnosis of GVHD and clinical correlation is required.1,2 Management involves early use of systemic corticosteroids and cyclophosphamide or rituximab for refractory cases.3 Most patients improve within days, though others take up to 4-6 weeks.

Our case illustrates a rare, potentially life-threatening, presentation of GVHD and exemplifies how GVHD can mimic autoimmune diseases. Awareness of this is important to allow prompt diagnosis and appropriate management.

References


Florida-Clinical Vignette-Poster Finalist
Thu-Cuc T Nguyen, MD

Title: Stereotactic Body Radiation Therapy for the Treatment of Oligoprogression on Androgen Receptor Targeted Therapy in Castration-Resistant Prostate Cancer

Authors: Thu-Cuc Nguyen, Azka Ali, William P. Skelton, Long H. Dang

Introduction: Castration-resistant prostate cancer is an incurable disease. To date, six agents have shown clinical efficacy and are FDA approved: abiraterone, enzalutamide, docetaxel, cabazitaxel, radium 223, and sipuleucel-T. Patients are typically sequenced through most or all of these agents, and then eventually succumb to their disease. Development of new treatments remains an unmet need. We report a case of a patient who progressed on enzalutamide with a single enlarging metastatic lesion, was treated with ablative stereotactic body radiation therapy (SBRT) while maintaining the same systemic treatment, who then had durable complete remission. Our findings have important clinical implications and suggest novel clinical trials for this difficult to treat disease.

Case Presentation: A 44 year-old male was diagnosed with high-risk prostate cancer and he elected for primary treatment with robotic-assisted prostatectomy. He was offered androgen deprivation therapy (ADT) in the form of leuprolide which maintained his PSA at undetectable levels for about 3 years when it increased to 1.9 ng/ml. At that time, he received salvage external beam radiation therapy to the pelvic region including the obturator bed and prostatic resection bed. ADT was continued throughout radiation therapy.

One year later, his PSA going up from 0.05 ng/ml to 1.19 ng/ml due to which bicalutamide was added to ADT. His PSA continued to rise along with enlarging external iliac lymph nodes. Bicalutamide was discontinued and patient was started on abiraterone and prednisone with excellent response with undetectable PSA and complete response of his lymph nodes for the next 2 years. PSA gradually increased and he was switched to enzalutamide, which led to stable PSA for another 6 months till it increased to 0.4 ng/ml. CT scan showed an enlarging para-aortic lymph node. Patient was evaluated for SBRT to the lymph node, which was however, not considered feasible, due to its small size. Follow up scan 4 months later showed increase in the size of the lymph node to 1.5 cm with PSA rising to 2.74 ng/ml. Patient then received SBRT to the para-aortic lymph node. During the entire time, enzalutamide was continued. Six months post treatment, CT scan showed marked decrease in the size of the para-aortic lymph node and decline in PSA.

Discussion: Our case report is the first to show that SBRT can be effectively added to control oligoprogression while on androgen receptor (AR) targeted therapy for CRPC, with improved disease control. We have previously shown such treatment approach using SBRT can be applied to gastrointestinal cancer patients with oligoprogression on chemotherapy. Our data suggests that larger studies using SBRT for oligoprogression should be proposed to assess survival outcomes for the treatment of CRPC.

References
Florida-Clinical Vignette-Poster Finalist
Nicole L Pontee, MD

Title: An atypical situation: a case of complement-mediated thrombotic microangiopathy in a renal transplant patient

Authors: Daniel Watford MD, Efren A. Chavez Morales MD, Adela D. Mattiazi MD, Warren L. Kupin MD

Introduction: Complement-mediated thrombotic microangiopathy (TMA) formerly referred to as atypical hemolytic uremic syndrome, is a rare condition comprising a constellation of symptoms making diagnosis an elusive task.

Case Presentation: A 34-year-old, CMV-negative Jamaican woman successfully underwent renal transplant from a CMV-positive donor and was placed on cyclosporin and prednisone for immune suppression. Due to lack of availability in her country, antiviral therapy for treatment of CMV was not given. Two months after her transplantation, she developed left-sided flank pain and was found to have multi-drug resistant left-sided Klebsiella pyelonephritis. She was treated with Meropenem. Due to progressive worsening of her clinical status, which involved worsening renal function, hypoxic respiratory failure, acute kidney injury, hemolytic anemia and severe thrombocytopenia, she was airlifted to Miami, Florida for further medical care.
A thorough investigation of the etiology of the patient’s clinical case was undertaken. Labs were significant for thrombocytopenia and anemia with peripheral smear demonstrating schistocytes. Further evaluation revealed a normal ADAMST13 level, low C3 and C4 levels, as well as a quantitative CMV of 2.4 million copies. During this time, patient’s creatinine level steadily rose to a peak of 9.8mg/dl from her baseline of 0.9-1.0mg/dl, and she became anuric. Renal biopsy was performed and demonstrated arteriolar and glomerular thrombosis.
The patient was started on treatment for suspected primary CMV viremia-induced, complement-mediated thrombotic microangiopathy. Therapy included IV Ganciclovir and multiple, daily sessions of plasmapheresis. Immune suppression therapy with cyclosporine was discontinued and exchanged for high dose steroids, and patient also underwent five rounds of hemodialysis due to severity of her acute kidney injury.
Due to financial issues, anti-complement therapy was not immediately available; however, the patient improved remarkably with complete recovery of renal function, eradication of CMV, and resolution of hemolytic anemia and thrombocytopenia. Mycophenolate mofetil was added to her immune suppression regimen after improvement of laboratory parameters. Her response to therapy was attributed to concurrent therapeutic approaches including treatment of primary CMV viremia in an immunosuppressed patient, early and aggressive plasmapheresis therapy, as well as discontinuation of cyclosporine, which also has been identified a causative agent in drug-induced TMA.

Discussion: The clinical case emphasizes the importance of appropriate antiviral prophylaxis in the high-risk immunosuppressed patient. Though complement-mediated TMA is a relatively rare clinical entity, this case also demonstrates simultaneous therapies to eliminate the offending impetus that drove the
disease process. This multifaceted therapeutic approach was essential in the recovery of not only the transplanted kidney, but also the patient.

References


Florida-Clinical Vignette-Poster Finalist
Edin Sadic

Title: Do Not Forget Your Flu Shot: An Interesting Case of Flu Myocarditis

Authors: Edin Sadic, Angel Martin, Hugo Narvarte

Introduction: Myocarditis is a rare complication of influenza viruses which has been reported to affect up to 10-11% of patients. Patients initially present with influenza-like symptoms followed by syncope, ventricular arrhythmias, cardiogenic shock and death. The clinical hallmark is rapid hemodynamic decompensation in less than 2 weeks after development of influenza symptoms.

Case Presentation: We present a 52 year-old lady with no known heart disease who presented to clinic with sore throat, cough, and diffuse myalgias and tested positive for influenza A. Despite starting Oseltamivir, she went on to develop dyspnea, fever, and facial swelling. Upon presentation to the hospital, she was found hypotensive with a systolic blood pressure in the 80s and sinus tachycardia in the 130s. She was started on intravenous hydrocortisone for concerns for adrenal insufficiency. She again tested positive for influenza A and was continued on Oseltamivir. Initial echocardiogram demonstrated preserved ejection fraction (EF), large pericardial effusion, and dilated inferior vena cava (IVC) with right diastolic collapse, consistent with tamponade. She underwent pericardiocentesis, with removal of 130 mL of fluid; fluid studies were unremarkable. Despite this, her cardiopulmonary status quickly declined. A repeat echo demonstrated EF 35-40% with global hypokinesis. CT angiogram chest was negative for any acute process. She was intubated for respiratory support and persistent hypoxia and started on vasopressor therapy for persistent hypotension. An intra-aortic balloon pump (IABP) placed and patient was transferred to our hospital for further care. A repeat echo several days later demonstrated EF 20% and global hypokinesis. Given her progressive decline with a working diagnosis of influenza myocarditis, she was placed on extracorporeal membrane oxygenation (ECMO), with which her condition dramatically improved. IABP was discontinued on hospital day (HD) three, ECMO on HD five, vasopressors on HD six, and extubated on HD seven. Follow-up echo demonstrated EF 55-60% and no wall motion abnormalities. She was successfully discharged without complication.

Discussion: Influenza myocarditis leading to refractory cardiogenic shock has been described in the literature. Some literature describes the successful use of extracorporeal membrane oxygenation (ECMO) therapy with full recovery. This report demonstrates the importance of early recognition of influenza myocarditis and the utility of ECMO in its management, in addition to other supportive measures.
Title: The Tumpeting of Angels: A Rare Case of Anisocoria

Authors: Zhabiz Solhjou¹, MD; Jorge Morales¹, MD; David Simmons², MD, ¹University of Central Florida/HCA GME consortium of Greater Orlando, ²Orlando VA Medical Center

Introduction: Anisocoria is defined as the difference of 0.4mm or more between the sizes of a person’s pupils, and can be the presenting symptom for range of etiologies from normal physiological variants to serious, life threatening conditions such as intracranial aneurysms. Here we present a rare case of isolated toxic, yet reversible, anisocoria.

Case Presentation: A 52-year-old lady with no significant past medical history presented to the emergency department with a two-hour history of blurred vision. She reports she was gardening in her backyard when her left eye began to “feel strange”. She entered her house and looked in the mirror and found that her left pupil was markedly dilated when compared to her right. Alarmed, she came to the emergency department and was admitted for evaluation.

On admission, her vital signs revealed RR 18, HR 75, BP 146/86, temperature 98.4° and O₂ of 100%. On examination, she was normocephalic and atraumatic. Eye examination showed significant difference in pupil sizes; the left measured 15 mm and right 5 mm. The difference pupil size was more pronounced when examined in light than dark, suggesting left pupil as abnormally dilated. Reaction to light was normal on right and minimal on left pupil. The rest of the eye, neurological, and physical examination were within normal limits. CBC, CMP, PT, and INR were normal. Head CT and Brain MRI showed no evidence of mass, aneurysm, hemorrhage, or stroke.

The morning following admission, patient’s anisocoria had significantly improved, normalizing later that day. Vascular and anatomical changes being ruled out and given rapid clinical resolution without intervention, toxic exposure was now first on the differential.

Upon further questioning about her gardening, she denies exposure to any pesticides, herbicides, or other chemicals. She mentions that in her garden there is a large collection of Angel Trumpets flowers, a colloquial name for Brugmansia suaveolens, a flower known to have powerful anticholinergic properties through traces of scopolamine, hyoscyamine, atropine and other tropane alkaloids. A subsequent trip to her home showed a back yard full of Brugmansia suaveolens.

Discussion: Brugmansia is a genus of seven species of flowering plants, also known as angel’s trumpets, rich in scopolamine, hyoscyamine, and several other tropane alkaloids. Brugmansia seeds and leaves are associated with multiple toxicities, including mydriasis. Here we present a case of anisocoria with local exposure of on eye to hands contaminated with seeds causing isolated anisocoria. Given its prevalence in the United States, physicians should be familiar with angel’s trumpets as a potential cause of anisocoria especially in the absence of other neurologic symptoms or physical exam findings. It can also cause severe anticholinergic crisis if ingested and can be toxic to pets and children.
Title: An Atypical Presentation of a Typical Infection

Authors: Gemma Gulati, MD, Maulik Patel, M.D., Joseph Kramer, M.D.

Introduction: Jaundice is a complex presentation and etiologies can be categorized into factors such as overproduction of bilirubin, impaired conjugation, obstruction, or hepatic inflammation. The differential diagnosis for jaundice can range from a simple benign condition to a potentially fatal cause. The clinical presentation of a patient guides the way through each differential towards a definitive diagnosis. At times, however, the diagnosis may not always be obvious.

Case Presentation: MJ is a 20 y/o male college student with no PMH who presented with jaundice, URI symptoms and abdominal pain for a week. He denied any taking any herbal supplements, alcohol use, or excess of Tylenol. On presentation, temperature was 101.8F, MAP <65 and HR >110, not responsive to IV fluids, thus required vasopressor support in the ICU. On physical exam, he was deeply jaundiced. Abdominal exam revealed mild tenderness in the upper quadrants. Initial laboratory values were significant for WBC 14,000, bilirubin 9.1mg/dL, direct bilirubin of 3.4mg/dL, AST 54I U/L, ALT 95IU/L, and normal alkaline phosphatase. Other pertinent labs were significant for elevated GGT 162IU/L, procalcitonin 1.63ng/mL, CRP 18.6mg/dL. Coagulation factors, LDH, haptoglobin, ceruloplasmin and urine copper were normal along with negative hepatitis panel, HIV, ANA, mitochondrial Abs, anti-smooth muscle Abs, and Tylenol level. He was treated empirically for septic shock with vasopressors, vancomycin and piperclillin-tazobactam. The source of infection was unclear. Ultrasound showed a common bile duct (CBD) of 7mm with increased echogenicity of the liver. Bilirubin and transaminases continued to be elevated with persistently normal alkaline phosphatase so a liver biopsy was performed. Pathology Results: from liver biopsy showed bile ducts infiltrated by neutrophils consistent with acute cholangitis. ERCP was then performed with sludge brushed out of the CBD but no stones or strictures. Patient received a cholecystectomy prior to discharge. A week later at his hospital follow-up visit, his bilirubin and transaminases were within normal range.

Discussion: Acute cholangitis is an infection of the bile duct caused by a backflow of bacteria from the duodenum. Usually cholangitis is triggered by an obstruction in the CBD. With no obstruction on imaging and a normal alkaline phosphatase level, acute cholangitis was low on the differential. Initial treatment for acute cholangitis is IV fluids and antibiotics. However, the underlying cause for cholangitis should be eliminated, i.e. removal of obstruction. Early detection is vital to prevent progressive infection leading to irreversible shock with multi organ failure.
Georgia-Clinical Vignette-Poster Finalist
Sonali Kumar, MD

Title: A Paradoxically Grave Diagnosis

Authors: Sonali Kumar, MD, Daniel Dressler, MD

Introduction: Hyperthyroidism is found in 1% of the US population. In patients with hyperthyroidism, 10-20% develop rhythm disturbances such as atrial fibrillation or flutter. Other cardiac complications encountered in hyperthyroid patients are rare and potentially fatal, requiring early recognition and intervention.

Case Presentation: A 58-year-old male presented with left shoulder pain and dyspnea on exertion. He had a past medical history of non-ischemic heart failure with reduced ejection fraction (40%), end stage renal disease, and hypertension. Physical exam was pertinent for blood pressure 100/60 with positive pulsus paradoxus of 18-20 mmHg, pulse 70 and irregular, respiratory rate 30, oxygen saturation 93% on 4 liters of oxygen via nasal cannula. Other findings included jugular venous pressure (JVP) to 12 cm, 3-component pericardial friction rub, muffled heart sounds, and bibasilar crackles. Laboratory evaluation revealed blood urea nitrogen of 66 mg/dL, leukocytosis to 11,000/mcL, Thyroid Stimulating Hormone .02 MCIU/mL, and elevated free thyroxine and total triiodothyronine. EKG demonstrated new atrial flutter. Transthoracic echocardiogram showed a large and circumferential pericardial effusion with tamponade physiology along with a bright speckled myocardium. He underwent emergent pericardiocentesis, draining 1.5 L of bloody fluid with 1,062 nucleated cells. Pulsus measurement status post pericardiocentesis was 8 mmHg with resolution in the patient’s dyspnea and normalized JVP. Pericardial fluid stains (including acid-fast bacilli) and cultures were negative, and no malignant cells were identified. Serum and urine protein electrophoresis (SPEP and UPEP) were negative. He converted to sinus rhythm with diltiazem and carvedilol. With a CHA2DS2VASc score of 3, anticoagulation was initiated. Newly diagnosed Graves’ disease was treated with Methimazole.

Discussion: The mechanisms by which pericardial effusions develop in Graves’ disease have not been well elucidated; however, it has been postulated that the mechanism may be similar to that of immune-mediated ophthalmopathy and myxedema associated with hyperthyroidism. Prior cases of hyperthyroidism-induced pericardial effusion have resolved with thyroid-suppressive therapy. However, our case demonstrates that tamponade physiology necessitates urgent pericardiocentesis. In one study, 64% of pericardial effusions were sanguineous, with malignancy and tuberculosis causing up to 45% and 28% of those bloody effusions respectively. Half of the twelve described hyperthyroid-related pericardial effusions (including this case) were sanguineous. We describe only the 6th reported case of pericardial tamponade related to hyperthyroidism and only the 12th case of hyperthyroid-related pericardial effusion, a rare cardiac complication. Our case of new Graves’ disease complicated by atrial flutter, sanguineous pericardial effusion, and cardiac tamponade highlights the dilemma of managing atrial flutter meeting criteria for anticoagulation in the setting of sanguineous pericardial effusion. While rare, bloody pericardial effusions with cardiac tamponade can be due to Graves’ disease, and requires aggressive urgent management, diagnostic evaluation and careful monitoring (if anticoagulation is indicated) to avoid fatal or grave outcomes.
Organizing Pneumonia in Antisynthetase syndrome: A rare occurrence

Authors: Keerthi Padooru, Olanipekun Titilope, Anam Umar, Adekunbi Egwakhe, Amina Farooq, Christine Charaf, Atlanta VA Medical Center, Morehouse School of Medicine Internal Medicine Residency Program, Atlanta, GA

Introduction: Anti-synthetase syndrome (ASS) is a rare systemic autoimmune disorder, characterized by the presence of anti-aminocyl-tRNA antibodies, and often associated with polymyositis, dermatomyositis and interstitial lung disease (ILD). The most common histologic pattern of ILD in patients with antisynthetase syndrome is nonspecific interstitial pneumonia (NSIP). Case reports have also identified usual interstitial pneumonia (USIP) and diffuse alveolar damage (DAD) in some patients. However, organizing pneumonia is a relatively rare finding in ASS.

Case Presentation: 55 year old gentleman with no significant medical history presented with complaints of worsening dyspnea on exertion, fatigue, proximal muscle weakness (more pronounced in lower thighs), tightness of calf, and joint stiffness of hands and fingers since 3 months. He denied orthopnea, paroxysmal nocturnal dyspnea, chest pain, palpitations, weight loss, fever, night sweats, history of travel and prolonged immobilization. Vital signs on admission showed BP of 131/87 mmHg, heart rate of 107 beats per minute, temperature of 98.4°F, respiratory rate of 20 cycles per minute and oxygen saturation of 96% on room air. Physical examination demonstrated dry hands, with thickened skin and cracks that started around the same time as his dyspnea. His lung exam revealed bibasilar velcro crackles. The rest of his physical exam was unremarkable.

Chest X-ray showed prominence of central hilar pulmonary vasculature and interstitial markings. Similarly, a contrast CT scan of his lungs revealed extensive bilateral parenchymal opacities mostly in the lower lobes. Pulmonary function tests revealed Tiffeneau-Pinelli index of 76%, total lung capacity of 41%, and reduced diffusing capacity of the lungs for carbon monoxide - 12.7. Given the constellation of complaints, there was a high suspicion for Myositis related ILD. His laboratory investigations were significant for elevated erythrocyte sedimentation rate of 52 mm/hr, elevated Creatine phosphokinase of 2136 U/L, positive anti-nuclear anibody and positive anti anti-threonyl-tRNA synthetase (Pl-7) antibodies. Other antibodies including anti Jo-1 and Mi-2 were negative. Muscle biopsy showed mild myopathy with type 2B fiber atrophy. Transbronchial biopsy was also obtained which showed focal changes of organizing pneumonia. He was diagnosed with the rare condition of ASS and associated organizing pneumonia based on clinical symptoms, signs, imaging and histopathological findings. He was treated with Rituximab and Prednisone therapy with resultant improvement in his symptoms and was discharged home.

Discussion: Our patient had anti-synthetase syndrome with features of cryptogenic organizing pneumonia on transbronchial biopsy. Organizing pneumonia is a rarely reported pattern of ILD in patients with antisynthetase syndrome. It is necessary to determine the type of ILD in ASS because it will define treatment approach. Also, as demonstrated by our patient, a negative screen for anti-Jo-1 antibodies does not rule out ASS. Other antibodies such as Anti PL-7 should be checked when the clinical presentation suggests ASS and ILD.

References


Georgia-Clinical Vignette-Poster Finalist
Krishna Patel, MD

Title: An atypical cause of typical chest pain

Authors: Krishna Patel, MD, Zareen Vaghaiwalla, MD, Brittany Clark, MD

Introduction: Sternoclavicular septic arthritis is a rare presenting form of septic arthritis. Most cases reported are in patients with diabetes mellitus, intravenous drug abusers and rheumatoid arthritis patients. Diagnosis is difficult with similar features of cardiac chest pain. We report a case of a 47-year-old previously healthy male who presents with chest pain found to have a sternoclavicular joint abscess. This case reviews some diagnostic features to discern this infection and therapeutic options for the treatment.

Case Presentation: A 47-year-old male with no past medical history presented with substernal chest pain for 1 day that radiated down his left arm and up left side of his neck. Exertion and movement made the pain worse while rest and nitroglycerin helped alleviate pain. He denied trauma, recreational drug use, heavy lifting exceeding 250lbs, fevers or chills. Vital signs on presentation were BP 215/119, RR 24, HR 95 and temperature 98.5F, oxygen saturation 99%. On physical exam, patient was muscularly built, in mild distress, without skin lesions on chest wall. Initial lab work showed elevated CPK, elevated LDL and total cholesterol. EKG showed inverted T-waves in the lateral leads without a previous EKG for comparison.

He underwent a nuclear perfusion stress test (NPS) which revealed anteriolateral attenuation. Due to unstable hypertension requiring 2 antihypertensive medications, chest pain, EKG abnormalities and NPS results, cardiology recommended cardiac catheterization. Cardiac catheterization showed minimal left anterior descending artery luminal irregularities with normal ejection fraction. His chest pain persisted, WBC became elevated and he became febrile with temperature of 102.6F. Blood cultures were negative. Empiric antibiotics were initiated. MRI of chest showed an abnormal signal in superior mediastinum extending in an extra mediastinal location in the subclavicular soft tissue extending laterally deep in the pectoralis muscle resembling a sternoclavicular abscess. Cardiovascular surgery drained the septic joint with subsequent repair of pectoralis flap by plastic surgery.

Discussion: Septic arthritis has an incidence of 2 out of 10 cases per 100,000 in the general population and 1% found in sternoclavicular joint. A sternoclavicular joint abscess is found in a high number of these cases.1,2 This case illustrates common features of typical cardiac chest pain that was noncardiac in etiology. Clinical features of sternoclavicular septic arthritis varies, however, most commonly presents with insidious onset of chest pain localized to the sternoclavicular joint, worse with movement of shoulder, limiting range of motion with accompanying fever.1,3 In most cases, infection is usually long standing and at presentation more easily discernable.1,3,4 This case was uncommon, in that the patient had no gross lesions or palpable abnormalities. The best imaging modalities to determine the extent of the condition is with CT or MRI, as these modalities can depict joint destruction, abscess, and periarticular inflammation.1

References

Georgia-Clinical Vignette-Poster Finalist
Isaac E Perry, DO

Title: Kittens and Fleas Doxycycline Please: Immunocompetent Meningoencephalitis

Authors: Isaac E. Perry D.O., Caitlin Allen, D.O. Carlton Kemp Jr., M.D., Nenad Avramovski, M.D.

Introduction: Meningoencephalitis is concurrent inflammation of both the meninges and the adjoining cerebral tissue causing an altered state of consciousness. We present a rare case of meningoencephalitis secondary to Bartonella henselae. Atypical cases can be difficult to recognize but a detailed social history remains paramount for early and accurate diagnosis.

Case Presentation: A twenty-year-old immunocompetent African-American man with a recent axillary abscess treated with antibiotics presented with prolonged confusion after a ten-minute generalized tonic-clonic seizure. In the hospital, he did not have a fever and his physical exam was positive for resolving confusion and bilateral axillary lymphadenopathy. CT, MRI and EEG were normal. He was discharged with close outpatient follow-up without medication.

Three hours after discharge, he presented back to the emergency department with multiple seizure episodes refractory to lorazepam, fosphenytoin and levetiracetam. The patient was intubated and midazolam and propofol were initiated. His seizure activity resolved. He was febrile at 103°F. After a normal CT of the head, CSF analysis was performed revealing 158 white blood cells with 95% neutrophils, glucose 89 mg/dl and protein 56 mg/dl.

Further questioning revealed the patient had several new flea-infested kittens with a history of scratches. Bartonella henselae IgG Ab was 1:1024 and IgM Ab was 1:256. All other viral serologies (HIV, CMV, Varicella Zoster, B. burgdorferi, Mumps, RPR, HSV, pneumococcal, enterovirus, cryptococcal) were negative. The CSF culture showed many white blood cells but no organisms. He was diagnosed with B. henselae meningoencephalitis (cat scratch disease). He was started on doxycycline and rifampin for a total of 14 days and anticonvulsant therapy was continued. He improved rapidly on antibiotics, extubated and walked out of the hospital. At outpatient follow-up two weeks later, he was seizure-free.

Discussion: Cat scratch disease is a self-limited condition caused by Bartonella henselae infection. CNS involvement is rare in immunocompetent individuals and usually occurs in HIV-positive patients. Our patient had no HIV detected by fourth generation testing (HIV combo 1-2 Ab/P24 Ag) as well as HIV-1 RNA quantification by PCR. We reiterate the importance of taking a thorough history and physical exam, without which serologic testing for B. henselae would not have been ordered. In a comprehensive literature search, there are only a few reported cases of meningoencephalitis caused by Bartonella henselae in immunocompetent individuals. Early recognition can prevent severe morbidity and mortality.
Georgia-Clinical Vignette-Poster Finalist
Jeeyon Grace Rim, MD

Title: Fourth Time’s the Charm, A Case of Acquired Hemophilia A Presenting as Recurrent Hematuria

Authors: Jeeyon Grace Rim, MD, Medical Resident, Department of Medicine, Emory University School of Medicine, Atlanta GA, Ethan Molitch-Hou, MD, Assistant Professor of Medicine, Division of Hospital Medicine, Emory University School of Medicine, Atlanta GA

Introduction: Acquired Hemophilia A is a rare autoimmune disorder with an incidence of about one case per million (1). It is caused by the spontaneous development of antibodies to factor VIII. It typically presents with multiple hemorrhagic complications and is associated with a mortality rate of 13-22% (2).

Case Presentation: A 58-year-old African American male presented to the Emergency Room for sudden onset left flank pain and hematuria. He reported mild right thigh swelling and pain over the past few weeks, at the site he typically injects his insulin. Past medical history includes insulin dependent type two diabetes and prostate cancer status post radical prostatectomy two years prior with prostate specific antigen level of 0.03 ng/mL. In the three months prior to presentation, the patient had three episodes of hematuria and flank pain treated as urinary tract infections.

Physical exam revealed moderate left flank and mild right thigh tenderness, with swelling, but otherwise unremarkable. Hemoglobin was 8.9 gm/dL, white blood cells 12.9 x 10^3/mcL, platelets 248 x 10^3/mcL, international normalized ratio (INR) 1.3, and partial thromboplastin time (PTT) 75.1 seconds. Urinalysis showed 338 red blood cells per high-power field. CT abdomen demonstrated moderate left-sided hydronephrosis and hydroureter with significant left perinephric stranding. The patient was treated empirically for a suspected urinary tract infection and urology was consulted for cystoscopy. Cystoscopy revealed an extensive amount of bloody fluid within the bladder, obscuring the ability to identify a clear source of bleeding. MRI of left thigh revealed an organized, non-enhancing hematoma within the vastus intermedius muscle measuring 4.7x3.3x5.6 cm. A broad workup for hematologic disorders revealed a Factor VIII activity of 6.8% and Factor VIII Inhibitor level of 124.2 Bethesda Units/mL establishing a diagnosis of Acquired Hemophilia A.

The patient was treated with recombinant factor VIIa, however, he developed worsening right thigh pain and hematuria. Factor Eight Inhibitor Bypassing Activity (FEIBA), was initiated followed by high dose steroids, cyclophosphamide, and rituximab therapy as FEIBA was tapered off. At one week follow up, his factor VIII level was 26.2 IU/mL and factor VIII inhibitor level was 8 BU/mL. No underlying pathology was found during hospitalization.

Discussion: Hematologic disorders are a rare cause of gross hematuria; however, it should be considered in a patient with no obvious source of bleeding such as trauma or malignancy, recurrent episodes, or additional sites of bleeding. Our patient presented three previous times with hematuria that was treated as more a common disorder, with only the hematoma this admission pushing towards a more severe diagnosis. His case illustrates the importance of keeping a broad differential, as rapid diagnosis and treatment of Acquired Hemophilia A is needed given the high risk of potentially life-threatening bleeding.

References

Title: Decades of Devastating Diarrhea

Authors: Gordon Robbins MD, Department of Medicine, Heba Iskandar MD, Department of Digestive Diseases, Emory University School of Medicine, Atlanta GA

Introduction: Non-specific complaints, such as watery diarrhea and weight loss can be difficult to manage when one’s usual differential diagnosis has been exhausted. When that rare diagnosis is made, understanding a basic mechanism enables us to develop successful treatments even when guidelines do not exist.

Case Presentation: A 46 year-old woman with Hashimoto’s thyroiditis, idiopathic cardiomyopathy, and BMI of 14 presented with over 20 years of watery diarrhea previously diagnosed as celiac disease with microscopic colitis. However, a strict gluten free diet provided no relief. Stool studies were negative for infectious pathogens, including giardia, ova, and parasites. Fecal leukocytes were absent, and MRI revealed no mucosal thickening disfavoring inflammatory bowel disease. Colonoscopy was significant for edematous mucosa in the colon and blunted villi in the terminal ileum. Biopsies were negative for malignancy but did reveal chronic inflammation of the lamina propria, decreased goblet cells, and increased epithelial apoptosis. The patient was positive for anti-goblet cell antibodies, which supported a diagnosis of autoimmune enteropathy.

Previously, the patient had minimal improvement with budesonide and mesalamine but each caused her significant swelling. Following this diagnosis, high-dose prednisone was initiated and later tapered with addition of 6-mercaptopurine (6-MP). She had poor tolerance of 6-MP and developed leukopenia, requiring the daily dose to be reduced to 50mg. Nevertheless, the patient was still having approximately ten watery stools per day with significant pain and bloating. TNF-alpha inhibitors were contraindicated in her case due to the patient’s cardiomyopathy, so she was initiated on vedolizumab infusions instead, while still taking 6-MP. After two infusions, her abdominal symptoms had resolved and her stool frequency significantly reduced. Repeat colonoscopy revealed reduced inflammation and only mild edema, and the patient now has a BMI of 30 with plans to taper 6-MP.

Discussion: General internists typically have the greatest continuity with patients and must recognize when a patient is receiving no benefit from treatment and is likely misdiagnosed. Autoimmune enteropathy is a rare disease, particularly in adults, and can easily be incorrectly diagnosed as celiac disease or inflammatory bowel disease due to their similar presentations. Autoimmune enteropathy should be suspected in patients predisposed to autoimmune disease when chronic diarrhea and malabsorption are refractory to dietary modification and providers have excluded immunodeficiency, possibility of infection, and other autoimmune processes. Diagnosis requires histologic confirmation of villous atrophy and deep crypt lymphocytosis. Corticosteroids are routinely the first-line therapy. Nevertheless, many patients are either steroid-dependent or refractory and require addition of immunomodulators or biologics such as azathioprine, 6-MP, tacrolimus, and infliximab for maintenance therapy. This is the first documented case of therapy with vedolizumab or any integrin receptor antagonist. As no formal treatment guidelines exist, it is important to document and share successes of each unique therapy.

References


Title: Progressive Myeloneuropathy with Symptomatic Anemia

Authors: Charlene L. Rohm, Sara Acree

Introduction: Copper deficiency has long been associated with cytopenias, but only in 2001 was its association with myelopathy first reported. The clinical presentation of copper deficiency is indistinguishable from that of subacute combined degeneration secondary to vitamin B12 deficiency; and when coupled with its rarity, the diagnosis of hypocupremia can often be missed.

Case Presentation: A 47-year-old woman presented with worsening balance and ascending lower extremity paresthesia over the past 3 months now involving the fingers. She also had palpitations, shortness of breath, and reported a 30-pound weight loss over 8 months. She denied a history of surgeries. She has smoked 10+ pack years and denied alcohol or illicit drug use. Physical examination showed no palpable lymphadenopathy. Neurologic examination revealed decreased vibratory sensation in the lower extremities, positive Romberg test, an ataxic gait, 3+ patellar reflexes, depressed ankle reflexes, and positive bilateral Babinski sign. Laboratory studies showed profound neutropenia (WBC 1.8, ANC 0.2), macrocytic anemia (Hb 5.2, Hct 17, MCV 109), and normal platelets. Electrolytes, LDH, iron panel, vitamin B12, and folate were normal. HIV RNA was undetectable. SPEP and UPEP were unremarkable. Bone marrow biopsy to evaluate the cytopenias revealed cytoplasmic vacuolization of erythroid and myeloid precursors. This finding prompted testing a serum copper level, which was <5 (undetectable).锌 level was 111. Given these results, further history gathering revealed that the patient had been using excessive Fixodent denture adhesive cream daily for over 20 years. Treatment involved oral copper repletion and cessation of denture cream use, resulting in improved cytopenias. Her neurologic deficits stabilized but persisted.

Discussion: When separately considering myeloneuropathy and cytopenias, the differential diagnosis is extensive. However, when considered together, the differential diagnosis is limited to deficiencies in copper, vitamin B12, and folate, myelodysplastic syndromes, certain lymphoproliferative disorders, and HIV infection. By obtaining correct serum studies as an initial test, invasive procedures and incorrect diagnoses and treatments may be avoided. As reported in the literature, it is not uncommon for hypocupremia to be initially diagnosed and treated as vitamin B12 deficiency; only after continued neurologic decline despite vitamin B12 supplementation was hypocupremia later diagnosed. Studies also showed that risk factors for hypocupremia include previous upper gastrointestinal surgery, zinc overload, and malabsorption syndromes. In this patient, the excessive use of Fixodent denture adhesive cream likely caused hypocupremia. This brand contains zinc, which competitively inhibits copper absorption in the small intestine. This patient had a zinc level in the upper limit of normal, so competitive inhibition of copper absorption likely occurred. With copper replacement, hematologic abnormalities often reverse completely and promptly. However, neurologic deficits often persist with residual symptoms. Therefore, prompt diagnosis of hypocupremia is essential in preserving neurologic functioning.
References


Georgia-Clinical Vignette-Poster Finalist
Anam Umar, MD

Title: An unusual presentation of Posterior Cerebral Artery Infarct

Authors: Anam Umar MD\textsuperscript{1,2}, Eric Chang MD\textsuperscript{1,2}, Muhammad Bilal MD\textsuperscript{3}, Chinedu Ivonye\textsuperscript{1,2} MD, Department of Internal Medicine, Morehouse School of Medicine.\textsuperscript{1}, Grady Memorial Hospital.\textsuperscript{2}, Department of Pulmonary and Critical Care, Emory University School of Medicine.\textsuperscript{3}

Introduction: Posterior cerebral artery (PCA) stroke most commonly presents with vision disturbances, hemiparesis, hemisensory loss, confusion, headaches, memory impairment, or vertigo. Few cases have reported unique presentations such as sudden change in behavior, lethargy or decreased activity level. We present an unusual case of PCA stroke mimicking giant cell arteritis (GCA).

Case Presentation: A 54-year-old female with a past medical history of hypertension and diabetes presented with two months history of temporal headaches, scalp tenderness, jaw claudication, and transient left sided vision loss from her ophthalmology clinic. Prior to the presentation, the patient had been following with rheumatology for diffuse joint pain and had been referred to ophthalmology for possible GCA. On presentation, vital signs were normal. Examination revealed sluggish left pupillary response and severe tenderness to light palpation of the bilateral temporal region. Labs were significant for an elevated erythrocyte sedimentation rate of 74 and C-reactive protein of 1.89 along with microcytic anemia (Hemoglobin 9.1g/dl). The rest of her chemistry and hematology labs were within normal limits. The patient was immediately started on high dose steroids for empiric treatment of GCA. Yet, emergent left temporal artery biopsy was negative for GCA. Further infectious and autoimmune workup including Treponema IgG, ACE, ANA, HIV and Lyme disease were also negative. CXR revealed bilateral hilar lymphadenopathy with scattered pulmonary nodules, which were confirmed on CT chest, concerning for possible sarcoidosis. MRI brain, however, revealed mixed staged infarcts involving the right PCA territory. Steroids were discontinued and aspirin and clopidogrel were initiated for stroke management. The patient’s symptoms improved with medical management and she was discharged with close outpatient neurology follow up.

Discussion: PCA diagnosis requires a high level of clinical suspicion as patients may present with a diverse spectrum of symptoms. Stroke patients sometimes delay seeking medical care after experiencing vision changes or headaches. On the other hand, an atypical presentation of PCA stroke, such as in our case, can be a diagnostic challenge and may lead to a delay in the initiation of appropriate therapy. This case highlights the fact that what appears to be a classic presentation of one disease may actually be an atypical manifestation of another. Physicians should always have a broad differential diagnosis to avoid mismanagement of patients, especially when dealing with neurological symptoms.
Title: Acute Anthracycline Induced Cardiotoxicity: A Rare and Reversible Cause of Acute Systolic Heart Failure

Authors: Hoyle L Whiteside, Arun Nagabandi, Amudhan Jyothidasan, Kristen Brown, John W Thornton

Introduction: Chronic anthracycline induced cardiotoxicity (AIC) is a type 1 chemotherapeutic cardiotoxicity characterized by irreversible myocyte destruction and necrosis. Although rare, acute AIC is characterized by a transient decline in left ventricular ejection fraction (LVEF) and does not appear to be dose related. The mechanism of acute AIC is not well defined, but the condition is felt to be reversible.

Case Presentation: A 58 year old female with history of polycythemia vera and no cardiac history underwent induction chemotherapy with 7+3 cytarabine and daunorubicin (90 mg/m2) for acute myeloid leukemia. Baseline echocardiogram demonstrated normal LVEF. On day 15 of induction, she developed tachypnea, dyspnea at rest, tachycardia, and mild troponin I elevation (0.106) which prompted cardiology consultation. Physical exam findings were consistent with decompensated heart failure, BNP was elevated (2190 pg/mL), and transthoracic echocardiogram revealed an interval decline in LVEF to 42% (Teicholz). On day 17 of induction, she developed polymorphic ventricular tachycardia in the setting of prolonged QTc and multiple electrolyte abnormalities with resultant hypoxemia prompting ICU transfer. Heart failure symptoms responded to afterload reduction, IV diuresis, and subsequent initiation of a beta blocker and the patient was transferred back to the floor on guideline directed medical therapy. LVEF remained reduced (35-40%) at the time of hospital discharge and overall prognosis was grim due to severity of underlying malignancy.

Discussion: Acute AIC is a rare cause of left ventricular failure which should be considered in patients developing acute respiratory failure as early as one week following anthracycline exposure. Prompt cardiology referral and appropriate pharmacologic intervention is paramount as left ventricular dysfunction is considered to be reversible.

References


Hawaii-Clinical Vignette-Poster Finalist
Dacia Boyce, MD

**Title:** Serial Lobar Lavage and Exogenous GM-CSF: a Novel Treatment for Pulmonary Alveolar Proteinosis Tailored to Hawaii

**Authors:** Dacia Boyce, MD (Associate); John Lee, MD (Associate); Phalgoon Shah, MBBS (Member); Matthew Aboudara, MD (FACP); David Hostler, MD, MPH (FACP)

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**Introduction:** Pulmonary alveolar proteinosis (PAP) is a rare lung disease characterized by diffuse accumulation of phospholipoproteinaceous material in the alveoli. It can lead to a wide variety of clinical sequelae including hypoxic respiratory failure. Whole lung lavage (WLL) is the treatment of choice in symptomatic PAP, but salvage methods of oxygenation such as extracorporeal membrane oxygenation (ECMO) are often required due to transient worsening of oxygenation. Research indicates granulocyte macrophage colony-stimulating factor (GM-CSF) plays a role in the pathophysiology of PAP. Here we highlight a patient diagnosed with PAP and treated with exogenous GM-CSF and sequential lobar lavage.

**Case Presentation:** 36 year old Thai female was admitted with a 4 day history of fevers, chills and dyspnea. She had no significant past medical history, recent travel, or sick contacts. On presentation she was tachypneic, tachycardic, and hypoxemic; labs revealed leukocytosis and lactic acidosis. Chest CT identified diffuse ground glass opacities in a ‘crazy-paving’ pattern. Following intubation due to impending respiratory failure, a bronchoscopy with bronchoalveolar lavage was performed. The lavage return stained positive with Periodic Acid Schiff, diagnostic of PAP. With worsening oxygenation, treatment was required. However, WLL was felt to be high risk due to the lack of ECMO capability. Nebulized GM-CSF was administered without significant clinical improvement. Subcutaneous GM-CSF was administered and isolated lobar lavages of the bilateral upper lobes were performed, with rapid improvement in oxygenation. Additional outpatient lobar lavage and continued subcutaneous GM-CSF therapy resulted in complete resolution of oxygen requirement and return to normal pulmonary physiology, verified by spirometry and DLCO.

**Discussion:** The estimated incidence of PAP is 1-3 cases per million, with peak incidence in the 3rd to 4th decade of life. Etiologies include mutations in surfactant proteins, mutations in granulocyte macrophage-colony stimulating factor (GM-CSF) receptor genes, autoimmune activation of anti-GM-CSF antibodies, toxic inhalation effects, and hematologic conditions. The autoimmune form of PAP is the most common (90% of cases). The clinical presentation is nonspecific; dyspnea is the most reported symptom. Other manifestations include chest pain, weight loss, fatigue, and fever. The standard of care for symptomatic autoimmune PAP is whole lung lavage usually with ECMO backup. Alternative, but less studied therapies include exogenous GM-CSF, plasmapheresis, and rituximab.

In acute presentations requiring mechanical ventilation, alternative treatment strategies with GM-CSF and sequential lobar lavage may be equally effective without incurring the risks inherent to whole lung lavage.
References


Hawaii-Clinical Vignette-Poster Finalist
Madeleine Prat, MD

Title: Bactrim or Badtrim? A Case of Rapidly Fatal Interstitial Pulmonary Fibrosis

Authors: Madeleine Prat (Associate), Eric Szu (Associate), Jordanna Hostler (FACP)

Introduction: Drug induced lung injury is commonly encountered in clinical practice. Pulmonary toxicity can be attributed to several mechanisms including direct dose-dependent damage to pneumocytes and alveolar capillaries, or a T-cell mediated immunologic reaction. Drug induced lung injury is a clinical diagnosis that hinges on identifying a temporal relationship between the onset of symptoms and drug exposure, new radiographic changes, and resolution of symptoms with discontinuation of the offending agent. We present a rare case of rapidly progressive interstitial lung disease induced by trimethoprim/sulfamethoxazole (TMP/SMX).

Case Presentation: Our patient was a 58 year old previously healthy African American man with a remote tobacco history of 12 pack years who was diagnosed with chronic prostatitis and treated with a four week course of TMP/SMX 160/800mg BID. One week into treatment the patient developed dyspnea on exertion. He presented to the Emergency Department (ED) with vital signs remarkable for a temperature of 100.1° and a room air oxygen saturation of 96%. His examination and chest radiograph were unremarkable. He was diagnosed with atypical pneumonia and was treated with azithromycin. He returned twice to the ED with similar complaints on subsequent days. On his fourth visit, the patient arrived in respiratory distress requiring 10 liters of supplemental oxygen. Exam was significant for diffuse wheezing. He reported recent travel to Ohio, but denied any sick contacts or unusual exposures. A CT chest showed bilateral groundglass opacities with focal areas of consolidation at the bases. Laboratory studies were remarkable only for a minimally elevated ALT and alkaline phosphatase. An echocardiogram was normal. As his oxygen requirement was too high to safely perform bronchoscopy, he was treated empirically for both suspected drug toxicity and possible pulmonary histoplasmosis. The patient worsened despite these therapies and was later intubated. Histoplasmosis antigen returned as negative. He had a surgical lung biopsy which showed organizing lung injury and diffuse alveolar damage without evidence of hypersensitivity pneumonitis. The patient was started on rituximab as salvage therapy. However, despite attempts to improve oxygenation the patient’s condition continued to decline resulting in terminal extubation.

Discussion: To our knowledge this is the first case of death from TMP/SMX related pulmonary toxicity. Our patient developed rapidly progressive, fibrosing, interstitial lung disease after initiation of TMP/SMX. Manifestations of pulmonary toxicity associated with TMP/SMX include fibrinous and organizing pneumonia, pneumonitis, pulmonary infiltrates with eosinophilia, ARDS, and drug hypersensitivity. Patients typically improve with systemic steroids and withdrawal of the medication. Obtaining a diagnosis is often challenging as signs and symptoms are non-specific. We suspect that our patient’s interstitial fibrosis was secondary to antibiotics due to the temporal relationship with drug initiation and clinicopathologic correlation.
Idaho-Clinical Vignette-Poster Finalist
Jennifer Miller, DO

Title: When Physical Exam Findings and Lab Tests Disagree: Likelihood Ratios of Physical Exam Findings in Ascites of Undetermined Etiology

Authors: Jennifer Miller, DO; Justin Reed, MS3; Farah Vega, MD, Magni Hamso, MD. University of Washington, Boise ID

Introduction: Ascites is the accumulation of fluid in the peritoneal cavity commonly caused by cirrhosis, hepatitis, heart failure, malignancy, or other etiologies. A common test to determine if the ascites is due to portal hypertension is the Serum-Ascites Albumin Gradient (SAAG). The SAAG, paired with a combination of physical exam findings, history, other lab results, and/or imaging are usually sufficient to make the diagnosis and determine the etiology.

Case Presentation: An 86-year-old male with a history of HFrEF, CAD, and past prostate cancer in remission presented with increasing shortness of breath for two days. He reported increasing abdominal girth over the previous 6 months accompanied by decreasing appetite, increasing abdominal discomfort and 17-lbs weight loss in the past month. He had a history of a blood transfusion 30 years ago and denied history of liver disease, IV drug use or significant EtOH use, hepatitis, or acute/viral illness preceding the distention.

The patient was found to have bibasilar crackles, large abdominal girth with positive fluid wave, flank and shifting dullness, cherry angiomas, 3+ edema to sacrum, muscle wasting, and platypnea. There was no scleral icterus, sublingual jaundice, asterixis or itching. The initial workup was significant for anemia, thrombocytopenia, chronically elevated alkaline phosphatase 200 U/L, GGT 131 U/L, AST 23 U/L, ALT 13 U/L, total bilirubin 1.9 mg/dL, albumin 2.7 g/dL, and protein 6.5 g/dL. Other labs ordered included hepatitis serology and AFP. Paracentesis revealed a yellow clear fluid, SAAG of 0.8, total protein <3 g/dL, WBC <200 #/cm, negative gram stain & culture. Esophagastroduodenoscopy to evaluate the varices found portal hypertensive gastropathy and a mass involving one-half the circumference of the duodenum. Mass biopsy indicated tubulovillous adenoma without high-grade dysplasia. Peritoneal fluid cytology was negative for malignant cells. CA 19-9 was positive at 59 U/L. Chest X-ray and abdominal CT showed diffuse prominence of interstitial lung markings, advanced cirrhosis, esophageal varices, ascites, and no signs of metastatic disease or masses. Echocardiogram demonstrated right-sided cardiac enlargement.

Discussion: Given the above-mentioned physical exam findings, the ascites can be attributed to the portal hypertension, however the SAAG score of 0.8 indicated non-portal hypertension related ascites. In this case, the likelihood ratio (LR) of the initial findings increased the post-test probability of ascites due to portal hypertension more than the SAAG alone. Occasionally, contraindicatory findings from various tests may skew the interpretation of the clinical picture, complicating the final diagnosis. Understanding likelihood ratios and the pre and post-test probabilities of physical exam, lab, and imaging findings can help to narrow the differential. While important to rule out other causes of this patient’s ascites, such as worsening heart failure or malignancy, clinicians could have been more reassured of the ascites’ etiology by using likelihood ratios.

References:

Illinois-Clinical Vignette-Poster Finalist
Arianne Clare Agdamag, MD

Title: Multiple Cutaneous and Mucosal Lesions in A Patient with Substance Abuse

Authors: Arianne Clare C. Agdamag, MD; Ofelya Gevorgyan, MD; Sobia Hassan, MD; Marcus Juan L. Esteban, MD

Introduction: The Cocaine-Levamisole-Induced Vasculopathy Syndrome is characterized by cutaneous manifestations, neutropenia, positive ANCA antibodies and rarely systemic involvement. Levamisole, an anthelminthic agent, is often used as a cocaine adulterant due to its euphoric and stimulating properties.

Case Presentation: A 51-year-old female with history of substance abuse presented with a 5-day history of oral, perianal and lower extremity ulceration, as well as purpuric stellate lesions on the toes and fingers. Laboratory tests were remarkable for WBC 1.54 K/uL, Hb 8.9 g/dL, ANC 10, Creatinine 2.29 mg/dL. Urinalysis showed proteinuria and hematuria. Urine toxicology was positive for cocaine and urine levamisole level was found to be elevated. Infectious work-up was unrevealing. Autoimmune work-up was pertinent for elevated CRP 173.2 mg/L, ESR >140mm/hr, P ANCA titer >1:640 and Myeloperoxidase Antibody >800. CT chest revealed diffuse ground-glass opacities bilaterally and subsequent bronchoscopy with bronchoalveolar lavage was negative for hemorrhage. Skin biopsy revealed superficial and deep perivascular dermatitis with scattered eosinophils while oral ulcer biopsy demonstrated inflammatory exudates with necrosis and eosinophilia. Bone marrow biopsy was unremarkable. Due to the constellation of symptoms and laboratory findings, she was diagnosed with Cocaine-Levamisole-Induced Vasculopathy Syndrome (CLIWS). She was managed supportively with noted improvement in renal function and skin lesions. Counseling was provided against cocaine use.

Discussion: CLIWS is an IgM mediated type 2 hypersensitivity reaction that can manifest as agranulocytosis, arthralgia and cutaneous vasculitis. Common cutaneous manifestations include purpura, digital gangrene, hemorrhagic bullae and skin infarction. Multiple ulcerations are less commonly reported cutaneous manifestations. High-titer perinuclear ANCAs are present in 86%–100% cases, and about 50% of cases also have cytoplasmic ANCAs. Anti-MPO antibodies are found in almost every case at very high titers, up to 15-fold higher than in patients with idiopathic ANCA-associated vasculitis. Both cocaine and levamisole can induce the formation of neutrophil extracellular traps, which are potential sources of self-antgens. Although rare, systemic involvement was evident in our patient with pulmonary and renal involvement manifesting as ground-glass opacities on imaging and elevated creatinine with hematuria, respectively. Skin biopsy often demonstrates leukocytoclastic vasculitis which was not seen in this case. However, biopsy revealed eosinophilia which is seen in type 2 hypersensitivity reactions. CLIWS often does not require disease-modifying anti-inflammatory medications or systemic immunosuppression. Avoidance of levamisole adulterated cocaine is crucial since manifestations recur with every exposure.

Among patients with known substance abuse who present with systemic symptoms, skin manifestations and positive autoimmune testing, CLIWS should be considered as one of the possible causes. It is often a diagnosis of exclusion but treatment is mostly supportive. Early diagnosis will prevent the use of unnecessary medications. Patient education and psychosocial support are essential in preventing recurrence and disease progression.
Illinois-Clinical Vignette-Poster Finalist
Afsana Asharaf, MBBS

Title: A'DRESS'ING AN UNUSUAL CASE OF TOXIC EPIDERMAL NECROLYSIS OVERLAP SYNDROME

Authors: Afsana Asharaf MD, Harry Fuentes-Bayne MD, Muhammad Baig MD, Moa’th Nassar MD, Ahmed Al-Ogaili MD, Department of Medicine, John H Stroger Jr. Hospital of Cook County, Chicago, Illinois

Introduction: Although severe cutaneous adverse reactions to drugs are common, overlap syndromes are rare and represent a diagnostic and therapeutic challenge, linked to a high mortality rate. We present a true overlap syndrome between Toxic Epidermal Necrolysis (TEN) and Drug Related Eosinophilia and Systemic Symptoms (DRESS) associated with the use of Chlorthalidone.

Case Presentation: A 41-year-old lady with past medical history significant only for hypertension presented with 10-days history of fever and a generalized rash associated with blistering of her oral mucosa and burning of her eyes, after initiation of Chlorthalidone. On presentation, the patient was febrile, but hemodynamically stable. Examination revealed confluent erythematous maculopapular rash interspersed with blisters involving over 60% of her body with follicular accentuation of the lower extremities and the back. The patient also had mucositis of tongue, oral and vaginal mucosa and conjunctival ingestion with mucopurulent discharge. Nikolsky sign was positive. Her laboratory parameters were significant for platelet count of 63,000/microliter, serum creatinine of 3 milligram/deciliter, serum aspartate transaminase of 7,087 units/liter, serum alanine transaminase of 3,847 units/liter, and serum lipase of 811 units/liter. Skin biopsy showed many scattered apoptotic keratinocytes and mild perivascular dermal lymphocytic inflammation, consistent with TEN. Liver biopsy was performed and findings were in keeping with drug induced hepatitis. Further work up was unrevealing for viral hepatitis and autoimmune disorders. As per the European Registry of Severe Cutaneous Adverse Reactions (RegiSCAR) scoring, our patient had a score of +4 which is Probable DRESS. Overall, the patient’s clinical presentation was consistent with TEN/DRESS overlap syndrome and Chlorthalidone was identified as the inciting agent. The offending agent was stopped and the patient was supportively managed in the intensive care unit. In addition, she was started on intravenous hydrocortisone 100 milligram every 8 hours. Patient progressively improved and was discharged home on oral prednisone without any further events on follow up visits.

Discussion: End organ involvement in patients with a phenotype of TEN should raise the possibility of an overlap syndrome, which constitute only about 2.1% of severe cutaneous adverse drug reactions. When there is an equal expression of different T cell mediated inflammatory pathways, overlapping cases occur. In addition to a detailed history including recent medication changes and a through physical examination, the use of a validated scoring system such as the RegiSCAR may assist in diagnosis. Clinicians should be aware of the occurrence of overlap syndromes, as it is plausible that this subset of patients may have improved clinical outcomes with additional therapeutic interventions such as corticosteroids.
Illinois-Clinical Vignette-Poster Finalist
Sindhura Bandaru, MD

Title: Primary hyperparathyroidism with low parathyroid hormone level diagnosed by the therapeutic response to the calcimimetic agent cinacalcet

Authors: ¹Sindhura Bandaru, MD, ²Michael Jakoby, MD/MA, FACP, ³Resident, Department of Internal Medicine; ²Chief, Division of Endocrinology, SIU School of Medicine, Springfield, IL

Introduction: Reliable assays for intact parathyroid hormone (iPTH) have greatly facilitated the diagnosis of primary hyperparathyroidism (HPT). However, in 5-10% of cases iPTH levels are within the assay reference range, and in rare cases (< 0.5%) primary HPT occurs with low iPTH levels. We present a case of delayed diagnosis of primary HPT due to unusually low iPTH levels where primary HPT was confirmed by the effect of the calcimimetic agent cinacalcet on hypercalcemia.

Case Presentation: A 57-year-old ambulatory male had multiple serum calcium levels in the range of 11-12 mg/dL (8.4-10.5). Over the next six years, twelve iPTH measurements obtained by four different manufacturers’ assays in three hospital laboratories, including the Mayo Clinic reference laboratory, ranged from 15-23 pg/mL (definition of suppressed < 20). Results: were unaffected by serial dilutions of serum. Urine 24 hour calcium was elevated (477 mg, > 300 mg/d), and serum phosphorus level was at the lower end of the laboratory reference range (2.8 mg/dL, 2.5-4.9). Total protein, albumin, serum creatinine, parathyroid hormone related peptide (PTHrP), 25-hydroxyvitamin D, 1,25-dihydroxyvitamin D, TSH, angiotensin converting enzyme, PTH fragments, and electrophoresis (serum and urine) were unremarkable. Parathyroid scintigraphy showed weak but persistent Tc-99 sestamibi uptake in the left lower neck that coincided with a 1 cm soft tissue density near the thyroid gland on cervical ultrasound. Unfortunately, fine needle biopsy of the mass yielded a specimen inadequate for cytological evaluation. Empiric therapy with cinacalcet was then started to determine if the patient’s hypercalcemia was PTH-responsive. Calcium and iPTH levels at start of treatment were 11.8 mg/dL and 21.7 pg/mL, respectively. Calcium level improved to 9.9 mg/dL on 30 mg cinacalcet daily and then increased to 11 mg/dL when treatment was stopped. On resumption of cinacalcet at 60 mg daily, calcium level fell to 9.4 mg/dL. Measurements of iPTH during both trials of cinacalcet were essentially unchanged from baseline at 17.9 pg/mL. The patient declined parathyroid surgery and other interventions for management of primary HPT.

Discussion: Several factors may confound measurements of iPTH including posttranslational modifications that affect measurement but not function, unmeasured but biologically active PTH fragments, interfering antibodies, increased PTH sensitivity, and pulsatile PTH secretion. Stable, low-to-low normal iPTH measurements over many years, with multiple assays, and unaffected by serial serum dilutions are most likely due to posttranslational modification of PTH or unmeasured PTH fragments. Cinacalcet lowers serum calcium levels by activating the calcium-sensing receptor in parathyroid tissue and inhibiting release of PTH.

This case demonstrates that empiric treatment with cinacalcet can confirm PTH-dependent hypercalcemia before committing patients to surgical neck exploration in the unusual case of primary HPT where iPTH measurements do not accurately indicate PTH activity.
Illinois-Clinical Vignette-Poster Finalist
Tulika Chatterjee, MD

Title: A Lesion: Not Just Skin Deep

Authors: TULIKA CHATTERJEE, MD and LARRY LINDAHL, MD, - UNIVERSITY OF ILLINOIS COLLEGE OF MEDICINE, PEORIA

Introduction: Gestational trophoblastic neoplasia (GTN) is one of the most curable solid tumors due to its high susceptibility to chemotherapy. GTN has a tendency to cause wide spread metastasis but does not have specific symptoms or clinical signs. A high degree of clinical suspicion along with thorough obstetrical/gynecological history taking can lead to early diagnosis and successful treatment.

Case Presentation: A 41 y.o. year old female, G4P2A2, presented to clinic with a slowly growing lump on left side of her scalp associated with headache and left eye pain since 2 weeks. She had intermittent, sharp pain in the area which radiated to the side of her face and was associated with photophobia. She had deep retro-orbital pain on palpation over the scalp lesion and pain exacerbated with bending. She reported early morning blurring of vision and nausea which improved through the day. Her inflammatory markers: CRP and ESR were normal and her ophthalmological evaluation was unremarkable. USG for left scalp showed an intramuscular, soft tissue mass lying immediately superficial to the left frontal bone measuring 3.5 x 3.0 x 0.6 cm with diffuse internal blood flow.

She was evaluated by surgery and tentative diagnosis of dermoid cyst of the scalp was formulated and excision biopsy was planned. Intra-operatively, it was noticed that the soft tissue mass extended up to and invaded the frontal bone itself. The excised supracalvarial mass measured approximately 3 cm and its histopathology was consistent with metastatic trophoblastic tumor with mixed features of choriocarcinoma and placental site trophoblastic tumor. Immunohistochemistry studies revealed diffuse neoplastic cell expression of HCG and variable HPL positivity.

Pregnancy test was positive and beta HCG level was 5924 which is above discriminatory zone. USG did not show any viable intrauterine pregnancy. CT head demonstrated a dural based mass measuring 4.1 cm in oblique AP extent with infiltration and destructive changes within the overlying calvarium. Intracranial mass effect was seen with 2 mm subfalcine deviation to the right. CT chest, abdomen and pelvis and PET scan showed no definitive evidence of metastatic disease. Patient underwent left frontal craniectomy, resection of the intracranial lesion with cranioplasty. The tumor extended against the brain but did not breach the pial membrane and its pathology was similar to the previously resected extracalvarial mass. Patient was diagnosed of FIGO stage IV gestational trophoblastic disease. She was successfully treated with 3 cycles of aggressive chemotherapy- Etoposide, Methotrexate, Actinomycin-D alternating weekly with Vincristine and Cyclophosphamide. HCG level was below 2 after completion of chemotherapy.

Discussion: It is rare for skin metastasis to be the first clinical presentation of GTN. Along with a unique presentation, our patient also had a rare combination of choriocarcinoma and placental site trophoblastic tumor cells in the same tumor mass. An additional striking feature was the presence of CNS involvement in the absence of lung metastasis.
Illinois-Clinical Vignette-Poster Finalist
Ashley M Darlington, MD

Title: A Mysterious Failing Heart – A Call to Identify Rare Etiologies to Guide Management

Authors: Ashley Darlington, MD, John Birchak, MD, and Lila Glotfelty, MD, PhD, Department of Medicine, University of Illinois-Chicago, Chicago, IL

Introduction: Many patients present with cardiomyopathy of unclear etiology, yet identifying a cause is of paramount importance. This case presents an undifferentiated cardiomyopathy typical of cardiac amyloidosis, a diagnosis which dramatically changed medical management.

Case Presentation: An 84-year-old male with heart failure with preserved ejection fraction, complete heart block (CHB) and ventricular tachycardia (VT) with implantable cardiac defibrillator (ICD), chronic kidney disease (CKD), and peripheral neuropathy presented with worsening orthopnea, paroxysmal nocturnal dyspnea, and lower extremity swelling. Echocardiography revealed severe concentric left ventricular (LV) hypertrophy, decreased LV chamber size, and restrictive filling pattern. Cardiac catheterization measured a pulmonary capillary wedge pressure of 26 mmHg and non-obstructive coronary artery disease. A cardiac magnetic resonance study demonstrated LV wall thickness of 2.0 cm with septal late gadolinium enhancement. Serum protein electrophoresis (SPEP) revealed a small, non-quantifiable IgG lambda band in the gamma region. Endomyocardial biopsy revealed positive Congo Red staining. Given the patient’s constellation of symptoms, imaging, and pathology findings, a diagnosis of wild–type transthyretin (TTR) amyloidosis was made.

Discussion: Amyloidosis accounts for only 3% of cardiomyopathies, but wild–type TTR accounts for 60% of cardiac amyloidosis. It is associated with higher mortality compared to idiopathic cardiomyopathy (hazard ratio for death 7.41). Clinical clues leading to this diagnosis include LV wall thickening and a non-dilated LV cavity associated with conduction abnormalities. A low-voltage EKG in the setting of increased wall thickness is associated with amyloidosis etiology (LR+ 8, LR- 0.3). Common extra-cardiac features include peripheral neuropathy, macroglossia, hepatomegaly, and CKD. This case displays LV wall thickening, non-dilated LV cavity, CHB, peripheral neuropathy, and CKD without low-voltage EKG. Diagnosis of amyloidosis requires positive Congo Red staining on tissue biopsy with further characterization via tissue staining. SPEP is non-diagnostic since findings may represent an unrelated concurrent monoclonal gammopathy of undetermined significance. A substantial proportion of patients presenting with unexplained non-ischemic cardiomyopathy are found to have an identifiable etiology. It is important to note that cardiomyopathy from amyloidosis is a contraindication to beta blockade due to the predisposition for AV nodal disease and dependence on heart rate to maintain cardiac output in the setting of a fixed stroke volume. Our diagnosis allowed us the opportunity to discontinue his beta-blocker. We also note there is no benefit from ICD for primary prevention since amyloid infiltration causes electro-mechanical dissociation. Thus, while our patient’s device was placed due to VT, it may not have mortality benefit in the setting of amyloidosis. Cardiac amyloidosis should be considered in any adult with increased ventricular wall thickness and non-dilated LV chamber size, particularly when associated with low-voltage EKG. This rare etiology may assist in medical management.
Illinois-Clinical Vignette-Poster Finalist
Rafael Francisco Go, MD

Title: Paraneoplastic Neurologic Syndromes: Making A Case For The Uncommon

Authors: Rafael Francisco Go, MD, MBA, Robel Desta, MD

Introduction: Paraneoplastic Neurologic Syndromes (PNS) are an uncommon group of disorders associated with malignancies that cause immune-mediated neurologic dysfunction. Patients can present with a plethora of neurologic symptoms, making it difficult to consider these diseases from the outset. This can therefore create a diagnostic dilemma for clinicians who are not entirely familiar with this group of disorders.

Case Presentation: A 62-year old woman with a known history of Stage IV Ovarian Cancer on chemotherapy presented with a 1 to 2-week history of new onset vertigo, vomiting, and frontotemporal headaches. There was no fever, focal weakness, hearing loss/tinnitus, or history of head trauma. On exam, she was found to have bidirectional horizontal nystagmus. Her vital signs were within normal limits and the rest of her physical and neurological exams were unremarkable. Main diagnostic considerations at this time included brain metastases and a neurological infection. Laboratory testing and head imaging studies were performed, yielding unremarkable findings. She was admitted and started on symptomatic treatment with benzodiazepines and anti-emetics. She deteriorated clinically, developing vertical nystagmus as well as losing the ability to ambulate properly. Neurology, Oncology, and Ophthalmology were consulted. At this time, a paraneoplastic neurologic disorder was considered, and a slew of paraneoplastic antibody panels was obtained. Anti-Yo antibodies came back positive, and the patient was diagnosed with paraneoplastic cerebellar degeneration. She was promptly started on steroids and intravenous immunoglobulin therapy, and underwent multiple plasma exchange sessions. A different chemotherapy regimen was also started at this time. Despite this, she did not respond to treatment and remained highly symptomatic. After 28 days in the hospital with no clinical improvement, she was transferred to home with 24-hour hospice care.

Discussion: Paraneoplastic cerebellar degeneration is a rare form of PNS and is observed in less than 1% of all malignancy cases. It is often associated with Hodgkin’s lymphomas, lung, breast, and gynecologic cancers. Neurologic outcomes are often poor among patients with the disease, and 75-80% remain non-ambulatory. Neurologic symptoms can precede the diagnosis of a malignancy in 60% of cases. Given the steady rise of cancer incidence worldwide, it is thus important to consider the diagnosis of a paraneoplastic neurologic syndrome in patients presenting with neurologic symptoms, regardless of the presence or absence of a previously diagnosed malignancy.

References

Illinois-Clinical Vignette-Poster Finalist
Iva Golemi, MD

Title: A Rare presentation of Median Arcuate Ligament syndrome with compression of the superior mesenteric artery causing chronic mesenteric ischemia.

Authors: Iva Golemi M.D. University of Chicago (Northshore), Juan Pablo Salazar M.D. University of Chicago (Northshore), Luis Diaz Quintero M.D.University of Chicago (Northshore), Alfonso Tafur M.D. Vascular Medicine - Northshore Cardiovascular Institute, Northshore University HealthSystem.

Introduction: Median arcuate ligament syndrome (MALS) is a rare conditions that usually affects women between 40-60 years of age compressing the proximal portion of the celiac artery and can cause chronic mesenteric ischemia. Common symptoms include nausea, vomiting, and postprandial epigastric pain leading to an aversion of food and resulting in weight loss. Epigastric bruits increasing with expiration are reported in 83% of cases and may be the only clinical sign of MALS. Compression of the superior mesenteric artery in addition to celiac artery represents an unusual variant of the MALS and poses a diagnostic and treatment challenge.

Case Presentation: A 59-year-old female presented to the emergency department with intermittent sharp abdominal pain, ongoing for the last 2 days. She also reports a 8 year history of abdominal discomfort associated with meals, and chronic cough attributed to GERD, which did not improve with therapy. Abdominal CT scan showed an acute dissection of the superior mesenteric artery (SMA) with near complete occlusion of the artery, and collateral vessel formation (Fig 1). The celiac artery also appeared occluded with extensive collaterals formed (fig 2). Pain management with norco and anticoagulation with warfarin 5 mg were initiated. A complete hypercoagulability and rheumatologic workup was performed and it was pertinent only for the MTHFR mutation 677 with normal homocysteine levels (Table 1). At this point, anticoagulation was stopped and the dissection of the SMA was thought to be a result of her chronic cough and celiac plexus occlusion. An arteriogram was performed and that revealed an arcuate ligament compression and partially occluding the SMA and restoration of flow by the arc of Riolan (fig 3). In the light of these findings she underwent supraceliac – aortomesenteric bypass with resolution of her symptoms.

Discussion: MALS remains a diagnosis of exclusion and is often misdiagnosed due to its relative scarcity. In almost all cases treatment remains surgical. The cause of MALS remains unknown, but is most likely multifactorial, including compressive effects on the celiac artery and surrounding neurogenic structures. The classical symptoms including postprandial abdominal pain, weight loss and epigastric bruit are likely to be incomplete due to the wide differential diagnosis including peptic ulcer disease, gallbladder disease, appendicitis and inflammatory bowel disease (IBD). Before the correct diagnosis is made patients will have most likely undergone several radiological tests, esophagoduodenoscopy (EGD), or even diagnostic laparoscopy. MALS can indeed compromise a rare cause of chronic mesenteric ischemia, which requires surgical treatment. Lack of awareness on this condition leads often to misdiagnosis and prolongation of the disease course.
Illinois-Clinical Vignette-Poster Finalist
Shravana Deepthi Gudivada, MBBS

Title: It’s not a usual Blast!: A rare case of disseminated Blastomycosis involving skin, soft tissues, lungs, bone and CNS

Authors: Deepthi Gudivada MD; Isha Tyagi MD

Introduction: Blastomycosis is an uncommon but serious fungal infection caused by the thermally dimorphic fungus Blastomyces dermatitidis. Although it primarily affects the lungs, in 20–30% of cases extrapulmonary sites including the skin, bones and central nervous system are involved. We are presenting a case of a young male who was found to have Blastomycosis masquerading as a soft tissue abscess.

Case Presentation: A 21 y/o male from Central Illinois with no significant PMH presented to the hospital with complaints of enlarging soft tissue lesion on his left frontal scalp area. Patient also complained of cough with mild intermittent hemoptysis for the past 3 months which had resolved and also a left thigh abscess which had spontaneously drained. He reported to have been exposed to soil and rotten wood material while cleaning dilapidated houses. Physical exam was positive for a fluctuant left frontal scalp abscess, persistent left thigh abscess wound and small wart like lesion on the ala of the nose. Patient had a normal WBC count and immunological workup was negative as well. MRI showed a complex fluid collection within the soft tissues of the scalp with intracranial and epidural extension and bony destructive changes. Leptomeningeal enhancement was also noted. CT of the chest showed left lower lobe consolidation, scattered nodules in both the lungs and a thick wall cavitary lesion in the left upper lobe. The scalp abscess was drained. Gram stain from the fluid was negative but the cytology revealed numerous broad based budding yeast organisms consistent with Blastomycosis. Patient underwent partial craniectomy with drainage of abscess and debridement of the bone. He was started on liposomal Amphotericin B for disseminated Blastomycosis.

Discussion: Blastomycosis is considered endemic to North American areas surrounding the Mississippi and Ohio River Valleys and parts of Canada. Blastomycosis commonly affects the lungs, however skin, bone and CNS involvement as in this case being rare manifestations of dissemination. Outdoor exposure to organic matter is an important risk factor. Blastomycetes can be identified via tissue culture, direct visualization in pathology preparations or PCR of samples including sputum, BAL and pleural fluid among others. Empiric fungal therapy should be started immediately once Blastomycosis is suspected. Clinicians should have a high suspicion for including endemic fungal infections in the differential for patients presenting with similar findings as prompt diagnosis and treatment can lead to improved outcomes and cure of the disease.

References

Title: Hepatocellular carcinoma with intra-atrial extension presenting as hemoperitoneum

Authors: Senuri Layer, MD

Introduction: Hepatocellular carcinoma (HCC) is the 5th most common cancer in the world, with the largest risk factor being cirrhosis [1]. It is a very treatable cancer with good prognosis and screening guidelines. Here, I present a case with very aggressive HCC that grew and spread within a surveillance period, and was discovered with catastrophic consequences.

Case Presentation: A 54 year old male with HBV cirrhosis on tenofovir, and complicated by moderate ascites and non-bleeding esophageal varices, presented for routine paracentesis. The fluid was remarkable for 158,000 RBCs on cell count. The subsequent triple phase Computed Tomography (CT) scan of the abdomen/pelvis revealed a large right hepatic lobe mass with large enhancing tumor thrombus extending into the right hepatic vein, inferior vena cava (IVC), and right atrium with adjacent thrombus within the portal vein. Alpha fetoprotein (AFP) level was 4452. Surveillance MRI abdomen 5 months prior had shown multiple stable subcentimeter lesions without typical HCC findings, and AFP had also been stable at 9.3. These imaging findings and hemoperitoneum were concerning for future life-threatening capsular rupture and bleed, so a right hepatic artery embolization was performed as a palliative measure. The poor prognosis was explained to the patient, and he was referred to hospice care at home.

Discussion: Tumor thrombus extending from the hepatic vein, through the IVC and into the right atrium is a rare complication of HCC. There are several cases reported in the literature of asymptomatic patients with presentation on routine echo, or symptomatic presentations with heart failure [2], or pulmonary embolism [3]. However, this case is unique in that the patient was found to have bleeding into his peritoneum on presentation. It is also unique in that the patient has documented imaging showing no suspicious lesions 5 months prior. Typical HCC imaging findings include arterial phase enhancement with washout appearance and/or capsule appearance on triple-phase imaging [4]. A recent meta-analysis showed that MRI is more sensitive for lesions <1cm in cirrhotics than CT [5], as was done in this patient. The American Association for the Study of Liver Diseases (AASLD) guidelines also recommend against routine biopsies of indeterminate lesions, and suggest alternative imaging or repeat imaging for close surveillance. Despite guideline-based HCC surveillance in this patient of every 4-6 months, this frequency of alternative and repeat imaging failed the patient. This case highlights the necessity of considering more frequent imaging in patients with existing indeterminate lesions.

References

Illinois-Clinical Vignette-Poster Finalist  
Tryphaena Manoharan, MD

Title: Asymmetric Paralysis: Just another stroke? No.

Authors: Manoharan, Tryphaena (1st), Azad, Hema and Yen, Scott

Introduction: West Nile Virus (WNV) disease is indigenous to Asia, Europe, Australia and Africa. It migrated to the Americas in 1999; by 2003 it had spread to 46 states. The primary reservoir of WNV is birds, and cycles via arthropod vectors; usually via mosquitoes. Risk factors include outdoor activity, contact with birds, and exposure to mosquitoes. Most people infected with WNV have a self-limited flu-like illness, characterized by fever, myalgias, headaches, gastrointestinal issues, and a maculopapular rash. Less than 1% of infected people develop West Nile Encephalitis (WNE); of those only 5-10% get poliomyelitis. Here we present a case of a patient who presented with symptoms mimicking a stroke who was found to have West Nile Encephalitis with poliomyelitis.

Case Presentation: A 60-year-old Latino Male with no past medical history presented with altered mental status (AMS) and dizziness for one day, while driving. His only complaint prior was a few days of a bad headache. In the emergency department (ED), a CT of the head and abdomen was performed and was negative. He was sent home with close follow up. Later that same day he was found unresponsive on the floor and presented again to the ED. Only additional history noted was tobacco use, and smoking cocaine 1-2x a week. His physical exam was thought to be consistent with an ischemic stroke, with left sided weakness, hyporeflexia, dysphagia, and dysarthria; complicated by aspiration pneumonia. MRI however showed no acute ischemia, with abnormal signal intensity in the right periventricular region, with possible focal demyelination. EEG showed severe slowing and disorganization. Several days later he spiked a fever of 100.9, and had a WBC count of 20.6 with 5% bands. A lumbar puncture was performed. CSF results revealed protein of 208.6, LDH of 38, lymphocytes of 95%, and the West Nile IGG and IGM were positive. Patient was diagnosed with West Nile Encephalitis with poliomyelitis and antibiotics stopped and supportive care continued. Patient was extubated and was sent to a nursing home with a PEG tube with no improvement of weakness or AMS. One year later the patient is able to speak well and eat normally, but he still has the left sided flaccid paralysis which is slowly improving.

Discussion: WNE, unlike other viral encephalitis, can present uniquely with muscular weakness, or poliomyelitis, which can mimic a stroke. WNV affects the lower motor neuron which causes asymmetric flaccid paralysis, and hyporeflexia with no sensory abnormalities. Age of 50 or greater has a higher risk of getting neurologic involvement in WNV disease. Patients with poliomyelitis usually require significant ICU care as with our patient. In patients such as ours who have a significant risk for getting WNV, WNE with poliomyelitis should be in your differential diagnosis.
Title: Anti-Hu Associated Paraneoplastic Sensory Neuropathy

Authors: Sukesh Manthri MD, Katyayan Dwivedi MD, Sindhura Bandaru MD, Sherjeel Sana MD

Introduction: Paraneoplastic neurologic syndromes are a heterogeneous group of disorders caused by mechanisms other than metastases, metabolic and nutritional deficits, infections, coagulopathy, or side effects of cancer treatment. These syndromes may affect any part of the nervous system from the cerebral cortex to neuromuscular junction and muscle. We describe a case of paraneoplastic sensory neuropathy (PSN) with anti-Hu antibodies (type 1 antineuronal nuclear autoantibodies).

Case Presentation: This is a 54-year-old female (former smoker) who was in her usual state of health until she developed paresthesias in her hands, arms and proximal legs over a course of a month. This eventually progressed to involve her face and cheeks. She also developed muscle and bone pain. She was evaluated by her neurologist and work up revealed normal SPEP, vitamin b12, TSH, and vitamin D level. She was admitted to the hospital for severe weakness and continued paresthesias. At that time, she was found to have hyponatremia (Na 115). CSF was positive for Anti Hu Antibody. Subsequent CT chest showed a 2.5x2.7 cm right infrahilar mass. Bronchoscopy with biopsy revealed small cell carcinoma. She subsequently completed 4 cycles of chemotherapy with radiation. Restaging PET scan 4 months after diagnosis showed no evidence of malignancy. However, she continues to have paresthesias in her face, cheeks, hands, arms and proximal legs despite being in remission. The paresthesias are partially currently controlled by a fentanyl patch and amitriptyline.

Discussion: Our patient was clinically diagnosed as having PSN with the anti-Hu antibody. The most common underlying tumor of anti-Hu-associated PSN is small cell carcinoma. Although pathophysiology is incompletely understood in these disorders, antibody and T-cell responses against nervous system antigens have been described in the literature. Patients suspected of having paraneoplastic neurologic syndrome should be checked for paraneoplastic antibodies. The paraneoplastic syndrome may precede the diagnosis of underlying malignancy. In such instances, the identification of specific antibodies and the clinical syndrome may aid in the diagnosis of the underlying malignancy after the more common etiologies have been ruled out.

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Illinois-Clinical Vignette-Poster Finalist
Bich-Ha Nguyen, MD

Title: When A1c and Glucose Don’t Match: A Rare Case of Hemoglobin Wayne

Authors: Bich-Ha Nguyen¹, Divya Korpu¹, Ashraf Abugroun¹, Sahar Rabiei-Samani¹, Vinita Bhagia²,
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Introduction: Hemoglobin A1c (HbA1c) is one of the most commonly used criteria for diagnosis and
treatment of diabetes mellitus type II (DMII). However, it is less likely known that there exists
hemoglobin variants that can falsely elevate HbA1c. Below is a rare case of a hemoglobin variant called
Hemoglobin Wayne which is clinically silent but runs the risk of misdiagnosing a patient with DMII,
committing them to a lifetime of antiglycemic agents and its complications.

Case Presentation: A 40-year-old asymptomatic obese female with significant family history of DMII was
evaluated for elevated HbA1c. Over the past four years, she consistently had elevated HbA1c of 10.3%,
10.5% and 10.9%. However, every glucose level on her basic metabolic panel had been normal, with
point of care fasting glucose < 126mg/dL, random glucose < 200mg/dL. Oral glucose tolerance test
(OGTT) showed fasting glucose of 84mg/dL (reference: 65-99mg/dL), 2 hour glucose of 90mg/dl
(reference 65-152mg/dL). Hgb 14.0g/dL. Given the discrepancy between her HbA1c and OGTT, a
fructosamine test was ordered which revealed a normal value of 228 µmol/L (reference: 205-
285µmol/L). Fasting insulin and C peptide were normal. Hemoglobin electrophoresis showed reduced
hemoglobin A1 of 91.4% (reference 96.4-98.2%) with normal hemoglobin A2, undetected Hgb F, S or C.
Hemoglobin electrophoresis cascade revealed the patient’s final diagnosis: Hemoglobin Wayne. Because
this diagnosis was clinically silent, our patient was never started on antiglycemics.

Discussion: Hemoglobin Wayne is a rare hemoglobin variant caused by a frameshift mutation which
alters the alpha globin molecule in heme and confers a specific charge to the hemoglobin similar to the
charge of HbA1c. It exists in two different isoforms, Hemoglobin Wayne I and Wayne II. The prevalence
of Hemoglobin Wayne is unknown as it is a very rare variant. Studies have shown that Hemoglobin
Wayne modifies charge of hemoglobin and confounds HbA1c by interfering with the commonly used
cation exchange high performance liquid chromatography assay. Methods such as the boronate affinity
assay, molecular analysis and DNA sequencing which are not affected by hemoglobinopathies can be
used. In cases of discrepancies between HbA1c and fingerstick testing, further investigations are
warranted such as: serum hemoglobin, B12, folate panel to look for decreased RBC turnover;
serum fructosamine to measure glycated protein unlike glycated hemoglobin in HbA1c; fasting insulin
and C peptide to assess endogenous insulin; hemoglobin electrophoresis to look for hemoglobin
variants. Physicians should be aware of hemoglobinopathies like Hemoglobin Wayne that can falsely
elevate HbA1c to prevent the misdiagnosis of DMII that can negatively impact a patient’s quality of life.

References


Illinois-Clinical Vignette-Poster Finalist
Niti Patel, MD

Title: Brugada syndrome in the setting of cardiac arrest related to a near drowning event

Authors: Niti Patel, MD, Tarik Hanane, MD

Introduction: Brugada syndrome is an autosomal dominant arrhythmogenic syndrome characterized by coved-type ST-segment elevation in the right precordial leads, and is associated with increased risk of sudden death. It is difficult to diagnose and may be missed unless activated by triggers. We present a case of an out-of-hospital cardiac arrest related to near drowning with ECG later revealing Brugada syndrome during targeted temperature management and during a febrile episode.

Case Presentation: 18-year-old African American male with no history presented after an apparent cardiac arrest due to near drowning in a swimming pool. Chest compressions were initiated for one minute by family but pulse was confirmed by EMS on arrival. In the emergency department, he was intubated for unresponsiveness. Initial ECG showed a right bundle branch block and nonspecific ST changes. Echo showed mild concentric left ventricular hypertrophy. On arrival to the ICU, he was started on therapeutic hypothermia given his poor neurologic status. When he achieved the targeted temperature of 34°C, his ECG showed incomplete right bundle branch block and new ST elevation in the precordial leads suggestive of possible Brugada. Cardiac enzymes were normal. Therapeutic hypothermia was maintained for 24 hours, at which point he was rewarmed with resolution of Brugada pattern on ECG. By day 5, he was awake, alert and following all commands and was able to be extubated. A brief febrile episode occurred on the evening of day 5 to 38°C and Brugada pattern again was noted on ECG. EP was consulted but unfortunately, the patient denied ICD insertion.

Discussion: Brugada syndrome is rare with prevalence of 1–5 cases/10,000 in the United States and is associated with mutations in CNSA-encoded sodium channel.

Our patient experienced anoxia, hypothermia, and metabolic acidosis during a near drowning episode which are factors known to increase the risk of arrhythmia. Moreover, aspiration of water predisposed his loss of consciousness and apnea. This correlates to cardiac-rhythm deterioration with tachycardia followed by bradycardia, pulseless electrical activity, and asystole.

Brugada ECG pattern is 20 times more likely to occur in febrile patients. Of the four ECGs we found in our patient with the classic Brugada sign, two occurred during a time of fever which is consistent with the current literature. Though Brugada syndrome is not associated with a particular anatomical defect, few studies demonstrate moderate abnormalities in the right ventricle. Our patient was on Propofol for four days. Propofol and other anesthetics have sodium channel blocking effects and can unmask Brugada syndrome. Our patient underwent therapeutic hypothermia for his poor neurologial status post-arrest. Hypothermia decreases cardiac conduction and extends all measured electrocardiographic intervals.

Brugada syndrome should be suspected in cases of precordial ST-segment elevation in patients undergoing significant temperature variations such as during therapeutic hypothermia or during febrile episodes.
Illinois-Clinical Vignette-Poster Finalist
Koosha Paydary, MD

Title: A Case of Immunoglobulin G4-Related Disease (IgG4-RD) Presenting with Malignancies: A Plausible Association

Authors: Koosha Paydary, Atefeh Vafa

Introduction: Immunoglobulin G4-Related Disease (IgG4-RD) is a syndrome of unknown etiology that is characterized by the lymphoplasmacytic infiltration of the affected organs with IgG4-positive plasma cells and subsequent fibrosis. Some of the diverse clinical manifestations in IgG4-RD include autoimmune pancreatitis, chronic sclerosing dacyoadenitis, retroperitoneal fibrosis, Reidel's and Hashimoto thyroiditis, etc. Elevated serum IgG4 levels and histopathologic findings on biopsy of affected organs are the means to diagnosis; however, the diverse clinical presentations require a high index of suspicion and a multidisciplinary approach for timely treatment. Although the association between IgG4-RD and malignancies has been previously described in literature, the issue of increased risk of malignancy among these patients remains controversial. In this abstract, we present a case of IgG4-RD that initially presented with enlarged left lacrimal gland, and developed several malignancies including endometrial carcinoma, Renal Cell Carcinoma (RCC), Diffuse Large B-cell Carcinoma as well as Systemic Lupus Erythematosus (SLE).

Case Presentation: Patient is a 47-year-old female that initially presented with enlargement of the left lacrimal gland causing proptosis of the left eye as well as mediastinal and hilar adenopathy. Biopsy of lacrimal gland and elevated IgG4 levels confirmed the diagnosis of IgG4-RD, and she was treated with prednisone and cellcept. After about six months, she was diagnosed with endometrial cancer stage IA and Renal Cell Carcinoma (RCC) that were treated with total hysterectomy, bilateral salpingo-oophorectomy and right partial nephrectomy. After almost one year, she presented with polyarthritis, maculopapular skin rash and increased Anti-Neutrophil Antibody (ANA) and Rheumatoid Factor (RF) titers with negative anti Citrullinated C peptide (anti-CCP) and was diagnosed with SLE. She showed a good response to treatment with steroids and rituximab; however in the subsequent year, she presented with diffuse neck, paratracheal and internal mammary lymphadenopathy and biopsy confirmed non-germinal CD30 (+) Diffuse Large B-Cell Lymphoma (DLBCL) stage IV. She has received six cycles of chemotherapy with CHOP-R regimen and two cycles of intra-thecal methotrexate up to now, while repeat imaging has documented remission.

Discussion: Although several studies have reported increased risk of malignancies such as non-Hodgkin lymphomas (NHL) among IgG4-RD patients, the exact temporality remains unclear. Some experts believe that IgG4-RD may in fact be a paraneoplastic presentation of malignancies such as NHL. Several studies have reported increased standardized incidence ratios of malignancies among these patients; therefore, increased screening for malignancies may be a consideration among patients with IgG4-RD. Our IgG4-RD case that presented with several malignancies further illustrates the increased clinical significance of cancer screening among these patients.
Illinois-Clinical Vignette-Poster Finalist
Carlos Rabascall Ayoub, MD

Title: An unusual presentation of sepsis from Haemophilus influenzae pyelonephritis in a transplanted kidney patient

Authors: Carlos Rabascall Ayoub, MD; Udit Joshi, MD; Sagar Ranka, MD; Sharath Vipparthy, MD, 1. Department of Internal Medicine, John H. Stroger Jr. Hospital of Cook County

Introduction: Haemophilus influenzae group (HIG) colonizes the upper respiratory tract and has been known to cause sinusitis, otitis media, epiglottitis and meningitis. There have been few case reports of HIG causing urinary tract infection, epididymitis and prostatitis. Here we report an unusual presentation of sepsis from H Influenza pyelonephritis in middle-aged men with a transplanted kidney.

Case Presentation: 53-year-old male presented to the Emergency Department with complaints of subjective fever and chills, nausea, multiple episodes of vomiting and watery diarrhea for 3 days. His past medical history was significant for hypertension, diabetes mellitus and End Stage Renal Disease status post second renal transplant, currently on immunosuppressants. On presentation, he was febrile to 102°F, tachycardic to 127 with mean arterial pressure of around 70 mmHg. He was in DKA with high anion gap metabolic acidosis - pH 7.28, HCO3 14, anion gap of 23, blood glucose of 467 and ketones in urine. He also had neutrophil predominant leukocytosis at 18000. He had complaints of burning urination on presentation and infectious workup was suggestive of urinary tract infection - pyuria with WBC of 17, leucocyte esterase positive, nitrite negative and 2 + blood; chest X-ray unremarkable. He was admitted in MICU for the management of DKA and was started on cefepime, vancomycin and his immunosuppressants were held. He also got CT scan of the chest and abdomen to look for source of sepsis and was suggestive of pyelonephritis in his transplanted kidney. In the meantime, blood and urine culture were positive for Hemophilus influenza and the antibiotics were tailored down to ceftriaxone. He also got MRI brain, which ruled out any CNS involvement as he was altered on presentation, which was later presumed to be from DKA or septic encephalopathy. His DKA resolved with resolution of all his symptoms and normalization of laboratory parameters. He was switched to levofloxacin to be completed for total of 14 days and his immunosuppressants were restarted. He was seen at the post hospital follow up clinic in 2 weeks and was doing symptomatically well.

Discussion: Genitourinary infections caused by H influenza and H parainfluenza are rare and are more common in children with anatomical malformation and dysfunction with the incidence reported to be <1%. The true incidence of HIG in adults is unknown, as most of the clinical laboratories do not include appropriate culture media for the growth of HIG. It grows best in chocolate agar as it is supplemented with factor X and V (NAD - Nicotinamide Adenine Dinucleotide). HIG is likely a more common pathogen responsible for UTI than it is currently appreciated. Hence, it is important to have more sensitive culture techniques and physician awareness to increase the diagnosis of HI UTI.

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Illinois-Clinical Vignette-Poster Finalist
Juan Pablo Salazar Adum, MD

Title: Arterial and Venous Thrombosis secondary to Non-Compaction Cardiomyopathy causing Anti-thrombin and Protein C Deficiency

Authors: Salazar Adum JP, Diaz Quintero L, Tafur AJ

Introduction: Non-compaction cardiomyopathy (NCC) is a condition characterized by ventricular trabeculae and deep intertrabecular recesses. [1, 2] Its prevalence among patients with heart failure (HF) is approximately 3 to 4 percent [3, 4]. Anti-thrombin (AT) and protein C (PC) deficiency in the setting of cardiomyopathy is not well understood, but well explained in the setting of liver congestion. We present a rare case of AT and PC deficiency resulting in acute arterial and venous thrombosis in the setting of rapidly progressive HF secondary to NCC.

Case Presentation: A 27-year-old, Hispanic male patient, with no pertinent PMH presented with acute onset, R lower extremity edema (LEE), ipsilateral calf pain and foot coldness and numbness. CTA of RLE showed a thrombus to the common iliac artery and an occlusion of the popliteal artery. ECHO demonstrated a borderline EF 50-55% with moderately reduced right ventricular systolic function (RVSF). The PC and AT levels were decreased. He underwent an uncomplicated RLE embolectomy and was discharged on LMWH to bridge to warfarin.

Two years later, he presented to our institution with scrotal and bilateral LEE after discontinuing anticoagulants. VS were unremarkable. Positive findings included a palpable, pulsatile, non-nodular hepatic left lobe with LEE 2+ and palpable pulses 2+ throughout.

Relevant workup included persistent decreased AT and PC levels. ECHO showed an EF 29% with a fixed thrombus on the apical wall of the LV and severely reduced RVSF. MRI of the heart showed multiple intracardiac thrombi in both ventricles. Multiple areas of contrast enhancement were seen on the lateral wall of the LV, apex, and basal aspect of the RV free wall consistent with embolic infarcts/trabeculae. The US abdomen showed a partial portal vein thrombosis (PVT).

He required a single chamber ICD placement and was given therapeutic doses of LMWH (1mg/kg) for the intracardiac thrombi and PVT secondary to NCC and PC and AT deficiency improving his clinical condition.

A 3-month follow-up ECHO showed an EF 48%, with a persistent fixed apical thrombus.

Discussion: The mechanism of a significant reduction in AT and PC levels resulting in acute limb ischemia and venous thromboembolism in the setting of HF is unclear. Some authors have reported that the hepatic synthesis of these natural anticoagulant proteins is affected in critically ill patients with septic or cardiogenic shock. [5, 6, 7] Thrombin generation increases as a result of increased synthesis of pro-inflammatory cytokines and tissue factor. The downregulation of thrombin synthesis is obtained via activated PC that inactivates factors Va and VIIIa. The reduced hepatic synthesis of AT and PC predisposes to a hypercoagulable state that in our case is thought to be caused by a severe, rapidly progressive HF secondary to an uncommon condition such as NCC.

References
Illinois-Clinical Vignette-Poster Finalist
Keith Smart, DO

Title: Sudden Cardiac Death Averted in Brugada Syndrome Provoked by Hashimoto's Thyroiditis

Authors: Keith Smart, DO, John Wille, DO

Introduction: Brugada syndrome is an autosomal dominant arrhythmic disorder caused by an alteration in the transmembrane ion currents that constitute the cardiac action potential. Incomplete right bundle-branch block with ST-segment elevations in the anterior precordial leads are the classic EKG findings of Brugada syndrome. Patients with Brugada syndrome are prone to develop ventricular tachyarrhythmias that may lead to syncope or sudden cardiac death in the absence of structural heart disease.

Case Presentation: A 59-year-old male with no significant prior medical history presented to the hospital after a syncopal event. His wife found him on the bathroom floor after hearing him collapse. Neurological workup for seizures and stroke were negative. The patient reported a positive family history of sudden cardiac death in both his father at age 40 and his sister at age 50. EKG showed ST elevations and T wave inversions in leads V1 and V2 consistent with type I Brugada pattern. Echocardiogram demonstrated a preserved ejection fraction and no evidence of structural heart disease. The diagnosis was confirmed by a positive Brugada provocation test and subsequently an implantable cardioverter defibrillator (ICD) was placed. Thyroid function was tested given the patient’s increasing fatigue leading up the syncopal event. He was found to have an elevated TSH, low T3 and low T4 consistent with hypothyroidism. Anti-TPO antibodies were positive confirming a diagnosis of Hashimoto’s Thyroiditis. The patient was discharged in good condition on Levothyroxine.

Discussion: This case shows the impact of a careful review of family history. Our patient’s account of sudden cardiac death in multiple family members directed us to the testing needed to diagnose Brugada syndrome. This case also shows the importance of recognizing Brugada pattern on EKG because it went unnoticed on our patient’s prior EKG’s. Our patient had type 1 “coved” Brugada pattern given his EKG with ST segment elevation that descended with an upward convexity into an inverted T wave in leads V1 and V2. Many events are known to unmask Brugada syndrome including: fever, electrolyte disturbances, alcohol and specific medicines. No such triggers were identified in our patient but he did present with increasing fatigue that prompted thyroid testing consistent with newly diagnosed hypothyroidism. It is most likely that his uncontrolled Hashimoto’s Thyroiditis exacerbated his underlying Brugada syndrome resulting in syncope. Many case reports document Brugada EKG pattern caused by hypothyroidism and correction of the Brugada EKG morphology after normalization of thyroid function. However, those patients were asymptomatic and did not meet criteria for Brugada syndrome as our patient did given his inducible electrophysiology provocation testing, syncope and family history of sudden cardiac death. Therefore, our patient received ICD placement because it is indicated to prevent sudden cardiac death in patients with Brugada syndrome.

References
Illinois-Clinical Vignette-Poster Finalist
Thomas Varghese, MD

Title: The Second Look That Saved A Girl’s Sight – Rare Presentation of Graves Orbitopathy – Unilateral Ptosis in a Euthyroid Patient

Authors: Thomas Varghese, Faisal Qureshi, Shyam Chalise (Presence Saint Joseph Hospital)

Introduction: Graves orbitopathy, also known as thyroid associated ophthalmopathy (TAO), is a complex condition that can be sight threatening if left undiagnosed or untreated. We are reporting a case of a patient who was diagnosed with TAO after presenting to us with unilateral ptosis in the setting of normal thyroid function with completely normal imaging findings.

Case Presentation: Our patient is a 20 y.o. female who presented to her primary care physician with complaints of left sided ptosis. The onset was sudden and she endorsed occasional unilateral tearing in the affected eye. She was initially referred to a neuro-ophtalmologist and an extensive work up including MRI of the brain, CT of the Orbits, ANA, SSA/SSB, IgG4, ANCA and Myaesthenia Gravis panel were ordered with unremarkable test Results:. Her thyroid function tests were normal with a TSH of 0.79 (Normal 0.2-0.4 mIU/L) and Free T4 level of 0.92 (Normal 0.7-1.9 ng/dl). Interestingly, in spite of a normal initial screen, her physician decided to take a second look at her thyroid function and proceeded to check her thyroid stimulating immunoglobulin (TSI) levels which were found to be elevated at 287 IU/mL (Normal 0 to 20 IU/mL). She was then referred to the endocrinology clinic. The above history and examination findings were confirmed. Additionally, she attested to experiencing pressure behind her left eye and diplopia intermittently. She denied weight changes, menstrual irregularities, palpitations, tremors, or cold/heat intolerance. There was no obvious eye swelling, exopthalmos, proptosis, redness, lid lag, or visual field defects noted on examination. A diagnosis of euthyroid Graves orbitopathy was made. She was started on prednisone 30 mg daily. Within 48 hours of initiating therapy, the patient showed excellent response with improvement in eye size and ocular symptoms. She was advised to follow up with us in 6 weeks for a bedside clinical assessment and to repeat her TSI levels, in the meantime she was gradually tapered off the steroids. At her follow up visit, she reported complete resolution of symptoms with her TSI levels dropping down to 128 IU/mL.

Discussion: 1. Clinicians must have a high index of suspicion when evaluating patients with new ocular symptoms for thyroid disorders. 2. Absence of classic symptoms, normal thyroid levels, and normal imaging findings must not deter clinicians from initiating further tests for Graves orbitopathy, especially when other diagnoses have been ruled out. 3. While TSH alone or in combination with free T4 is the preferred initial screening test, TSH receptor assays such as TSI, TRab (TSH Receptor Blocking Antibodies), TBII (TSH Binding Inhibitory Immunoglobulin) and LATS (Long Acting Thyroid Stimulator) must be considered owing to their superior sensitivity and specificity. 4. High dose IV or oral pulsed therapy with corticosteroids such as methylprednisone is the preferred initial line of treatment.

References

Title: Atrial Myxoma: A Not So Benign Cause of Toe Gangrene

Authors: Tanvi Desai, Deep Desai

Introduction: Atrial myxomas are benign cardiac tumors that present in a myriad of ways according to their size and location. Seventy percent of patients present with intracardiac obstructive symptoms such as pulmonary edema, hemoptysis, or dyspnea. However, thirty percent of patients present with an embolic phenomenon into the systemic circulation, the most catastrophic areas being the brain and retina. Therefore, atrial myxomas can masquerade as common disease processes and must be included as part of the differential when an embolic pathology is possible.

Case Presentation: A 53 year old female with a past medical history of type 2 diabetes mellitus complicated with multiple foot ulcers, coronary artery disease, and hypertension presented to the hospital with left fourth toe discoloration of three days duration. On physical exam, the patient was a Caucasian female with normal heart, lung and abdominal examinations. Inspection of the left foot revealed poor foot hygiene with decreased sensation by monofilament testing. The dorsum of the foot revealed warmth, erythema, and +2 pitting edema, while the fourth toe demonstrated a gangrenous black discoloration and tenderness to palpation.

The patient’s labs were grossly normal except for a leukocytosis of 14,000/ microliter. Electrocardiogram demonstrated normal sinus rhythm with possible left atrial enlargement. CT of the lower extremity demonstrated just chronic degenerative changes and no evidence of abscess or osteomyelitis.

An aortogram was conducted which demonstrated patent two vessel runoff to the left foot, excluding peripheral artery disease as a cause to her gangrene. Transthoracic echocardiogram demonstrated normal left ventricular systolic function, with mild left atrial dilation, and no vegetations or thrombi. Transesophageal echocardiogram was then pursued, revealing a small pedunculated mass along the interatrial septum with atypical appearance.

The patient was then immediately started on heparin and referred to cardiothoracic surgery for mass excision. Final pathology revealed the mass as an atrial myxoma.

Discussion: Toe gangrene often considered an internist’s “bread and butter” medicine. However, an internist must sometimes look beyond the common, and search for the uncommon when the etiology cannot easily be explained.

In this case, a patient with known risk factors for toe ischemia, such as diabetes and coronary artery disease, the initial diagnostic work up did not yield the cause of her symptoms. With the pursuit of the more invasive transesophageal echocardiogram, a left atrial myxoma was identified as her embolic source, and potentially saved her a more catastrophic presentation.
Therefore, the differential of embolic atrial myxoma should be included in any patient with acute peripheral ischemia, regardless of confounding risk factors.

References


Indiana-Clinical Vignette-Poster Finalist
Ashley M Jancuska, MD

Title: Visceral Kaposi’s Sarcoma Presenting as Acute on Chronic Hypoxic Respiratory Failure

Authors: Ashley Jancuska M.D., Brent Bagley M.D., and Andrew Wiele D.O., Indiana University Internal Medicine Residency Program, Indianapolis, Indiana

Introduction: Hypoxic respiratory failure in an immunocompromised patient has numerous and diverse potential etiologies. Isolated pulmonary Kaposi’s Sarcoma is a rare but important cause to consider. It can mimic more common disease processes such as pneumonia or opportunistic infections, so a high degree of clinical suspicion is warranted.

Case Presentation: A 27-year-old male with recently diagnosed human immunodeficiency virus (HIV) and acquired immunodeficiency syndrome (AIDS) first reported dyspnea and a nonproductive cough seven months prior to presentation. His work-up revealed bilateral infiltrates on chest computed tomography (CT), an elevated lactate dehydrogenase level, and a CD4 count of eleven. He was empirically treated with trimethoprim-sulfamethoxazole and a three-week course of prednisone for presumed pneumocystis pneumonia. Confirmatory testing was inconclusive, however, and his symptoms progressed over the following months. Repeat chest CT showed progression of the bilateral opacities, necessitating bronchoscopic evaluation. Bronchoscopy showed mucosal erythema, and bronchoalveolar lavage fluid studies were consistent with lymphocytic interstitial alveolitis; thus, a prolonged course of steroids was initiated. His respiratory status continued to decline, and he was admitted to the hospital with significant dyspnea at rest and a productive cough. Physical exam revealed tachycardia, tachypnea, and hypoxemia requiring oxygen supplementation with nasal cannula. The only pertinent finding on physical exam was bilateral coarse rhonchi; no mucocutaneous findings were present. Chest CT demonstrated worsening of the perihilar ground-glass opacities, lymphadenopathy, and pleural effusions. An extensive workup was pursued, but initial viral, fungal, bacterial, and autoimmune studies were unrevealing. A transbronchial biopsy was obtained, and pathology demonstrated Kaposi’s Sarcoma with detectable human herpesvirus 8 DNA. He was continued on anti-retroviral therapy (ART); furthermore, given his symptomatic pulmonary involvement, chemotherapy with liposomal doxorubicin was initiated during his hospital stay. At his follow-up appointment two weeks after discharge, he was symptomatically much improved and breathing comfortably on room air; additionally, repeat imaging confirmed substantial reduction of the bilateral opacities.

Discussion: Kaposi's Sarcoma is a low-grade angioproliferative malignancy that is closely associated with human herpesvirus 8; while it classically presents with cutaneous findings, isolated visceral involvement is a rare but important presentation of the disease. In the era of ART, the incidence of AIDS-related Kaposi’s Sarcoma is decreasing, but there remains an increase in disease incidence during the first six months after ART initiation. Furthermore, steroids can also precipitate Kaposi’s Sarcoma by either triggering its induction or exacerbating pre-existing disease. Literature suggests that ART remains first-line treatment for Kaposi's Sarcoma, but in patients who remain symptomatic from visceral disease, systemic chemotherapy is an appropriate adjunctive measure; steroids should be avoided if possible. Respiratory failure in immunocompromised individuals is often a diagnostic challenge, and this case demonstrates that isolated pulmonary Kaposi's Sarcoma should be considered in the appropriate clinical setting.

References
Indiana-Clinical Vignette-Poster Finalist
Gloria Ong, MD

Title: The diagnosis of noninsulinoma pancreatogenous hypoglycemia syndrome 40 years after gastrectomy: a case report.

Authors: Gloria KH Ong, M.D., Robert Fick, M.D.

Introduction: Noninsulinoma pancreatogenous hypoglycemia syndrome (NIPHS) is a rare disorder of undetermined etiology uniquely characterized by postprandial hypoglycemia, and the exclusion of an insulinoma. With the majority of hyperinsulinemic hypoglycemia cases attributed to insulinomas, NIPHS with nesidioblastosis is rare, encompassing less than 5% of total cases. Although scarce, NIPHS has been observed more frequently in recently years, most commonly in months following gastric bypass or gastrectomy. We present a case report of a 77 year old patient with recurrent severe postprandial symptomatic hypoglycemia that manifests 40 years after gastric surgery.

Case Presentation: Patient is a 77 year-old African American female with a history of gastroesophageal reflux disease with previous ulcer, status-post gastric surgery (late 1970’s), who initially presented with altered mental status and a capillary glucose of 25 (mg/dl). Patient reports that recently she had been feeling weak and fatigued, specifically related with meals. Patient has no history of diabetes and denies previous episodes. Physical exam was unremarkable. Further workup revealed increased insulin (69 micoIU/L), proinsulin (95 pmol/L), and C peptide (17.9 ng/ml) accompanied with low serum glucose (18 mg/dl). Cortisol stimulation test was within normal limits. Computed tomography (CT), magnetic resonance imaging (MRI), and endoscopic ultrasound imaging showed no evidence of insulinoma. Esophagogastroduodenoscopy (EGD) showed evidence of patent Billroth II gastrojejunostomy. Selected arterial calcium stimulation test (SACST) showed no evidence of insulinoma. Patient was then diagnosed with suspected NIPHS. The patient was ultimately medically managed on acarbose 50mg three times daily, and diazoxide 50mg every 8 hours upon discharge. Since then, the patient has reported no further episodes of symptomatic hypoglycemia.

Discussion: With the overall incidence of NIPHS cases associated with gastric bypass increasing, early detection is crucial to avoid dire outcomes. Identifying and diagnosing NIPHS is a challenge due to the rarity of the condition, in addition to the complexity of diagnostic tests required. With our patient expressing NIPHS so unusually after 40 years, a correct diagnosis was preeminent to implement the appropriate treatment, and to avoid unnecessary surgical procedures. Although NIPHS is definitively diagnosed by pancreatic biopsy with histopathology showing nesidioblastosis, all diagnostic modalities for localizing an insulinoma in our patient were negative. Thus, this led to medical management which was sufficient treatment for our elderly patient.

References

Iowa-Clinical Vignette-Poster Finalist
Justin J Chau, MD

Title: What is DRESS Without the Drug? A reevaluation of viral reactivation in an apparently immunologic process

Authors: Justin Chau, MD

Introduction: Drug reaction with eosinophilia and systemic symptoms, or DRESS, is an uncommon but potentially deadly immunogenic constellation of signs often associated with drug exposure and reactivation of herpesvirus infection, including cytomegalovirus (CMV), Epstein-Barr virus (EBV), and human herpesvirus-6 (HHV-6). Though viral reactivation is typically considered a byproduct of the host immune response, little data exists documenting viral levels prior to symptom onset, resulting in an ambiguous understanding of its pathogenesis.

Case Presentation: A 26-year-old woman with no significant history presented to the emergency department complaining of abdominal pain, cough, and new rash. A week prior, she had been evaluated for nonspecific headache and lymphadenopathy, found to have a pulmonary infiltrate on X-ray and prescribed azithromycin for community acquired pneumonia. Exam on current presentation was notable for high-grade fever, nasolabial erosions, cervical adenopathy, diffuse erythematous rash, and respiratory distress. Laboratory testing yielded marked eosinophilia and transaminitis. EBV and CMV IgM and IgG returned positive; heterophile antibody and HIV tests were negative. CT showed multifocal pulmonary consolidations and diffuse lymphadenopathy.

Bronchoalveolar lavage yielded no bacteria but showed numerous eosinophils. Axillary lymph node biopsy showed atypical cells, eosinophils and lymphocytes consistent with EBV infection; flow cytometry and peripheral smear were negative for hematologic malignancy. Skin biopsy yielded an interface dermatitis with eosinophils. These findings, along with her presentation, were consistent with a diagnosis of DRESS. She was administered high-dose steroids, and her symptoms and eosinophilia quickly improved. However, the diagnosis – and subsequently, its underlying mechanism – was called into question after recognizing the patient was symptomatic prior to azithromycin use, and fulfilled diagnostic criteria for DRESS within 1 week of exposure to the drug, much earlier than the 2-6 week latency typically expected.

Discussion: As many as 20% of DRESS-consistent cases are unable to establish clear drug causality. Case reports have shown that CMV DNA levels in patients with pre-existing infection decrease upon onset of DRESS symptoms before rebounding, which may confound current interpretations of spontaneous viral reactivation. In vitro studies by Descamps have demonstrated induction of viral replication following exposure to DRESS-associated medications, and Hashimoto’s longitudinal research shows patients sometimes develop serious HHV-related complications following resolution of DRESS. These studies suggest both a host-independent mechanism between drug and viral replication, and the possibility that patients developing early-onset DRESS may be predisposed to this by higher indolent viral load or greater propensity for development of viral reactivation. The patient’s presentation is unusual with respect to her symptoms preceding exposure to the potential culprit drug, and that the drug itself, azithromycin, has rarely been reported to incite this reaction. This suggests that viral reactivation in this patient played a major role in her development of DRESS.

References


Iowa-Clinical Vignette-Poster Finalist
Nicole M Grogan, MD

Title: A Kidney’s Crisis

Authors: Nicole M. Grogan, M.D., University of Iowa Hospitals and Clinics, Katherine I. Harris, M.D., University of Iowa Hospitals and Clinics

Introduction: Acute kidney injury, thrombocytopenia, and hemolytic anemia are separate problems commonly encountered by the internist. In combination, however, these diagnoses represent a limited subset of diseases. While thrombotic thrombocytopenic purpura or hemolytic uremic syndrome may more often cause this triad of findings, this case report will focus on a rare complication of a common rheumatologic disorder that can mirror this clinical picture.

Case Presentation: A 46-year-old female presented for evaluation of two days of nausea and vomiting. She also described swelling in her legs and decreased urination over that time. Exam revealed blood pressure of 150/100 (baseline blood pressure 90/50), pitting lower extremity edema, taut skin of both hands, and bibasilar crackles. Labs revealed creatinine of 3.2 mg/dL (baseline 0.8), hemoglobin of 8.9 g/dL (baseline 12.6), and platelet count of 113 x 10^9/L (previously 492 x 10^9/L). Peripheral blood smear revealed 3+ schistocytes. Urinalysis showed 2+ protein. Further history revealed that one month prior she had been diagnosed with systemic sclerosis with limited cutaneous involvement and started on mycophenolate mofetil, with minimal change in her clinical symptoms.

Discussion: While thrombotic thrombocytopenic purpura or hemolytic uremic syndrome may initially be high in the differential diagnosis for this patient’s presentation, in a patient with a history of systemic sclerosis it is essential to recognize scleroderma renal crisis as a disorder that can present with these same findings\(^1,2\). It affects approximately 4% of patients with systemic sclerosis with limited cutaneous disease\(^2\). The differentiation between these diseases is critical, as management is vastly different and time sensitive. While there is no validated definition of scleroderma renal crisis, a consensus classification has been proposed\(^3\). This includes new onset hypertension (blood pressure > 140/90) and at least one associated feature (> 50% increase in serum creatinine over baseline, proteinuria, thrombocytopenia, hemolysis, or hypertensive encephalopathy) in patients with systemic sclerosis. Rather than pursuing plasma exchange for treatment of thrombotic thrombocytopenic purpura or providing supportive care alone for hemolytic uremic syndrome, our patient was started on captopril. Prompt blood pressure control, before irreversible renal damage has occurred, is the mainstay of treatment for scleroderma renal crisis\(^4\). Captopril is recommended initially, as it has the advantages of rapid onset and short duration of action, allowing for rapid dose titration\(^5\). The goal of initial antihypertensive therapy is returning the patient to previous baseline blood pressure within 72 hours. As immediate blood pressure control is necessary to prevent continued renal damage, it is imperative that physicians recognize scleroderma renal crisis as a rare consequence of systemic sclerosis, since management is vastly different from more common diseases that may present similarly.

References


Kansas-Clinical Vignette-Poster Finalist
William Goodman, MD

Title: An Interesting Case of Thrombocytosis.

Authors: William Goodman, MD; Quoc Troung, MD; Robert Badgett, MD

Introduction: Polymyalgia rheumatica (PMR) is one of the most common systemic inflammatory diseases of unknown cause affecting older adults. In an attempt to better discriminate PMR from rheumatoid and other inflammatory articular diseases, the European League Against Rheumatism/American College of Rheumatology (EULAR-ACR) classification criteria for PMR was developed. Individual findings such as bilateral shoulder aching, and abnormal CRP/ESR are highly sensitive with values of 99% and 96% respectively. Other common, but less sensitive findings, include morning stiffness lasting longer than 45 minutes (77%), and remission of symptoms within 4 weeks of starting steroids (71%). Unfortunately, the EULAR-ACR classification criteria fall short when individual findings are combined: sensitivity 68% and specificity 78%. Thrombocytosis occurs in more than 50% of patients as a part of a general acute inflammatory response.

Case Presentation: The patient is a 78-year-old male with a PMH of hypertension. He was traveling by car when he developed non-traumatic, acute swelling of his right lower extremity (RLE). He went to an emergency department nearby where an ultrasound confirmed the presence of a right popliteal DVT. Routine blood work was normal except an elevated platelet count of 625,000 which was attributed to acute inflammation. He was discharge home on apixaban. He was seen by his PCP two weeks later for follow up. His RLE edema had improved. However, a repeat CBC showed his platelet count increased to 850,000. He was referred to Hematology. Upon presentation to Hematology two weeks later, his platelet had count risen to 1.1 million. Workup was initiated: JAK2, calreticulin (CALR), and thrombopoietin (MPL) mutation were all negative, and bone marrow biopsy did not show evidence of a myeloproliferative disorder. He was seen again four weeks later, now presenting with joint aches, pain, and difficulty getting out of bed. Labs showed a platelet count of 1.5 million and ESR 90. A preliminary diagnosis of PMR was made. He was started on prednisone, and referred to Rheumatology were the diagnosis of PMR was confirmed. Recheck of his platelets three weeks after starting prednisone showed a normal count of 405,000.

Discussion:

- The inaccuracy of EULAR-ACR classification criteria and the diverse presentation of PMR can make diagnosis difficult
- The absence of bilateral shoulder aching, whose sensitivity is 99% makes this case unusual.
- Over 50% of patients with PMR may have thrombocytosis.
- Diagnosis is primarily clinical, supported by inflammatory markers, and response to treatment. In this case, resolution of clinical symptoms and normalization of his platelet count.

References

Kentucky-Clinical Vignette-Poster Finalist
Jonathan S Alexander, MD

Title: Debilitating lymphedema secondary to Kaposi Sarcoma in the setting of untreated HIV-AIDS

Authors: Jonathan S. Alexander, M.D.1, Hamza Hashmi, M.D.2, Samuel B. Reynolds, M.D.1, Mounika Mandadi, M.D.2, 1. Internal Medicine Residency Program, University of Louisville School of Medicine, Louisville, KY, U.S.A., 2. Division of Hematology and Oncology, James Graham Brown Cancer Center, University of Louisville School of Medicine, Louisville, KY, U.S.A.

Introduction: Stemming from an unrestrained human herpesvirus 8 (HHV8) infection, AIDS-associated Kaposi sarcoma (KS-AIDS) is the most aggressive variant of the four subtypes. We present an interesting case of AIDS-associated Kaposi sarcoma with an atypical initial presentation involving lymph nodes and axial skeleton, an unusually aggressive clinical course with a recurrent/relapsing disease pattern and debilitating lymphedema successfully managed with supportive measures and systemic chemotherapy.

Case Presentation: A 30-year-old male presented to the emergency department with violaceous appearing skin nodules of two months duration and rapidly worsening painful swelling of the scrotum and bilateral lower extremities that prevented ambulation. His past medical history was significant for AIDS-associated Kaposi sarcoma, with lymphatic and osseous metastasis upon initial presentation three years prior, successfully treated with doxorubicin. Due to noncompliance with antiretroviral therapy (ART), the patient had multiple recurrences of Kaposi sarcoma primarily with cutaneous manifestations, which responded well to systemic chemotherapy. Whole body PET/CT 3 months prior to presentation did not reveal any evidence of systemic disease.

Initial workup including echocardiogram and doppler venous ultrasound of the lower extremities did not reveal cardiac dysfunction or deep vein thrombosis. CT imaging showed significant soft tissue edema around the pelvis, abdomen, thighs and upper back without evidence of any obstructive lymphadenopathy. CD4 count was 35 cells/ mm3. Biopsy of one of the cutaneous lesions was consistent with HHV8–associated Kaposi sarcoma. Empiric antibiotics were started to cover for underlying skin and soft tissue infection in the setting of immunodeficiency. Symptomatic management included compression stockings and elevation of the legs and scrotum. The patient underwent palliative radiotherapy to his thighs and perineum without much relief. He was started on ART along with systemic chemotherapy. After undergoing two cycles of liposomal doxorubicin, there was significant improvement in his symptoms as well as resolution of the skin nodules.

Discussion: Kaposi sarcoma typically presents with cutaneous disease. Involvement of noncutaneous sites like the oral cavity, gastrointestinal tract, and respiratory system can be seen but visceral or osseous involvement as the initial manifestation is relatively uncommon. Although Kaposi sarcoma is an indolent disease, noncompliance with antiretroviral therapy can lead to a rapidly progressive clinical course with frequent recurrences. Despite the diagnosis often being clinical, biopsy should be performed to rule out other possibilities in the setting of a compromised immune system. Lymphedema, particularly in the face, genitalia, and lower extremities may be out of proportion to the extent of the disease and requires early recognition and intervention with supportive measures and systemic chemotherapy to avoid life-threatening complications.

HIV-associated Kaposi sarcoma in patients noncompliant with antiretroviral therapy can have an atypical presentation and a rapidly progressive clinical course. Prompt initiation of supportive measures and systemic chemotherapy are required to avoid significant morbidity and mortality.
References

Kentucky-Clinical Vignette-Poster Finalist
Jacqueline Danielle Leboeuf, MD

Title: High Bili is Not Always DILI

Authors: Jacqueline LeBoeuf, MD, Adam Gray, MD

Introduction: The differential for hepatic cholestasis requires a thorough history followed by abdominal ultrasound to differentiate extrahepatic from intrahepatic cholestasis. Though invasive, a liver biopsy may be required to differentiate between the various etiologies. Some of the etiologies of intrahepatic cholestasis include drugs and toxins, PBC, PSC, infiltrative liver disease, and metastatic cancer.

Case Presentation: A 43-year-old man presented with two weeks of jaundice. He reported associated right upper quadrant abdominal pain, diarrhea, anorexia, and a 40-pound weight loss. He denied any fever, herbal supplement use, alcohol or intravenous drug use, recent travel, or family history of hemolytic disorders. Past medical history included obesity and an episode of suspected diverticulitis 1 month prior to presentation for which he received 2 weeks of ciprofloxacin and metronidazole.

His vital signs were normal. He was a well appearing, obese man with jaundice and scleral icterus. He had no spider angiomas or palmar erythema. He had mild tenderness to deep palpation in the epigastrium. No shifting dullness or hepatosplenomegaly.

His aspartate transaminase was 266U/L, alanine transaminase 235U/L, bilirubin 14.7mg/dL, alkaline phosphatase 721U/L, albumin 3.6g/dL, and total protein was 9.1g/dL. Platelets were 335 K/cmm and INR was 1.23. Viral hepatitis serologies, human immunodeficiency virus, antinuclear (ANA), anti-mitochondrial antibodies (AMA), and ceruloplasmin were normal. Ferritin was 1400 ng/mL. Computed tomography of the abdomen and pelvis showed splenomegaly and retroperitoneal lymphadenopathy with a normal appearing bowel, gallbladder, and liver, and no intrahepatic ductal dilation. Abdominal ultrasound was normal. Liver biopsy demonstrated microinvasive metastatic adenocarcinoma. He subsequently had a colonoscopy revealing a five-centimeter sigmoid colon mass.

Discussion: The diagnostic approach to a patient with a cholestatic liver injury is something every internist should know. It begins with a detailed history focusing on potential medication or toxin exposures, risk factors for viral hepatitis, alcohol use, and symptoms of an associated systemic disease. Abdominal imaging, usually with ultrasound, to assess the biliary ducts is essential to separate extrahepatic from intrahepatic cholestasis. The presence of biliary ductal dilation suggests extrahepatic cholestasis. Common etiologies of extrahepatic cholestasis are choledocholithiasis, malignant obstruction, biliary strictures, and infections.

A normal biliary system on imaging favors intrahepatic cholestasis. For intrahepatic cholestasis workup begins with autoimmune markers (AMA, ANA, and anti-smooth muscle) assessing for primary biliary cirrhosis (PBC). Additional testing includes MRCP for primary sclerosing cholangitis (PSC), serology for viral hepatitis, and if appropriate pregnancy testing for intrahepatic cholestasis of pregnancy. If diagnosis is still uncertain, liver biopsy is often performed evaluating for infiltrative liver disease or metastatic cancer.
References

Kentucky-Clinical Vignette-Poster Finalist
Ralph Millett, MD

Title: Extrapulmonary Tuberculosis Mimicking Peritoneal Carcinomatosis

Authors: Ralph Millett, MD; Sirisha Tummala, MD; Farida Izzi, MD

Department of Internal Medicine, The George Washington University, Washington, D.C.

Introduction: Despite a relative uptrend in tuberculosis (TB) infections in the US, primary presentation of extrapulmonary TB remains rare and often presents a diagnostic challenge. Peritoneal tuberculosis in particular mimics more common presentations such as peritoneal carcinomatosis and often requires a high index of suspicion, invasive testing, and considerable time to make an accurate diagnosis.

Case Presentation: A 28-year-old female patient with sickle cell trait and recent puerperium presented with a two-week history of worsening abdominal distension, abdominal and lower back pain, night sweats, and unintentional weight loss. Abdominopelvic contrast-based CT scan on arrival revealed large volume ascites with multiple peritoneal nodularities as well as prominent mesenteric and pericardial lymph nodes concerning for peritoneal carcinomatosis; no liver cirrhosis was noted. Ascitic fluid aspirate was cloudy, orange, and studies revealed a low SAAG, high RBC count, neutrophilia with lymphocytic effusion, and no mesothelial cells. The initial ascitic fluid culture was sterile and adenosine deaminase was negative, while serum tumor markers demonstrated an elevated CA-125. Shortly after the initial paracentesis, the patient became persistently febrile despite initiation of broad-spectrum antibiotics. Trans-vaginal ultrasound demonstrated only a right-sided hemorrhagic cyst, however a PET scan showed markedly increased uptake in the uterus, ovaries, and adnexa with omental caking suggestive of peritoneal metastases. The working diagnosis at that time was peritoneal carcinomatosis with unknown primary. Subsequently, an ultrasound-guided omental biopsy was performed to identify the primary source and revealed no malignant cells but did show extensive necrotizing granulomata. This raised concern for tuberculous peritonitis for which the patient was started on empiric RIPE therapy (rifampin, isoniazid, pyrazinamide, ethambutol). Importantly, the patient had a low-exposure risk for tuberculosis, no pulmonary symptoms, and her pulmonary CT was unremarkable. AFB, FITE, and fungal staining on the biopsy specimen were negative, however the patient’s QuantiFeron Gold returned positive. The patient’s fevers abated and symptoms largely improved after inception of RIPE therapy. Several weeks after discharge, the ascitic fluid culture returned positive for Mycobacterium tuberculosis by DNA probe, confirming the suspected diagnosis of primary peritoneal tuberculosis.

Discussion: Peritoneal tuberculosis is often mistaken for peritoneal carcinomatosis due to similarities in symptoms and imaging as well as non-specific elevation of CA-125. Though rare in the US, consideration of this condition may allow for ready amelioration of symptoms, provide better prognosis, and can prevent otherwise unnecessary invasive procedures.
Kentucky-Clinical Vignette-Poster Finalist
Ripa K Patel, MD

Title: Eye’m Melting

Authors: Ripa Patel, M.D. and Edwin Avallone, D.O., University of Kentucky Department of Internal Medicine

Introduction: Corneal melt is a rare but serious complication of rheumatoid arthritis with potentially devastating complications. Serious eye conditions that can be sight-threatening, such as corneal melt, exhibit red-flag features that should not be missed.

Case Presentation: A 50 year-old gentleman with seronegative rheumatoid arthritis on leflunamide presented with foreign body sensation, irritation, pain and photophobia in bilateral eyes which acutely worsened to bilateral vision loss. Funduscopic and slit-lamp examination showed crusting, moderate erythema, and edema of both eyes. Further detailed exam showed mucoid discharge, bilateral corneal infiltrates, corneal neovascularization, diffuse corneal haze of right eye, and limbal thinning of left eye. Visual acuity was significantly decreased in both eyes. The patient’s history of rheumatoid arthritis and the exam findings of corneal infiltrates with neovascularization and limbal thinning established the diagnosis of acute and rapid bilateral corneal melting. Cyclophosphamide and intravenous steroids were initiated. Empiric oral and ophthalmic antibiotics were started for possible infectious etiology, which were then titrated off once infection was ruled out. Serologic workup for other autoimmune etiologies was negative. Patient’s vision improved after two days of therapy, however, on day seven, he had a left corneal perforation and subsequently underwent corneal transplantation. His vision improved after surgery, and he was discharged on ophthalmic steroids and a long course of oral prednisone.

Discussion: Corneal melt manifests between 12 and 35 years after the diagnosis of rheumatoid arthritis. It is characterized by inflammation involving the limbal part of the cornea and sclera, collagen destruction, cellular infiltration and limbal vascular changes indicative of vasculitis. Although Herpes simplex virus keratitis and retained lenticular material are the two most common causes of corneal melt, other etiologies include Sjogren’s, SLE, Granulomatosis with polyangiitis, as well as complications due to intraocular surgeries, such as cataract extraction. Symptoms are severe pain, photophobia, sensation of foreign bodies, and excessive watering.

The red flags of a red eye are moderate-severe pain, photophobia, reduced visual acuity, eye trauma, and unilateral marked redness. These are associated with keratitis, scleritis, acute glaucoma, foreign body trauma, chemical burns, and orbital cellulitis. These entities should be part of the differential diagnoses when evaluating a red eye with red-flag features.

Diagnosis is confirmed with slit lamp examination, which narrows the differential diagnosis of the red eye. Infectious etiologies must also be excluded. Treatment must be early and aggressive due to a high mortality rate. Cyclophosphamide is the treatment of choice in conjunction with steroids, and the usual surgical treatment is scleral or corneal grafting. Duration of treatment is patient-specific based on presence of underlying systemic vasculitis, response to medical treatment, need for surgical intervention, and presence of concomitant infections. Corneal perforation is a feared complication of corneal melt and is treated with corneal transplantation.

References


**Kentucky-Clinical Vignette-Poster Finalist**  
**Christopher J Redmond, MD**

**Title:** Seeing What Needs TB Done: Treatment of Scleromalacia in a Patient with Suspected Latent Tuberculosis

**Authors:** Christopher Redmond, MD

**Introduction:** Scleromalacia is an ocular manifestation of rheumatoid arthritis (RA) that can cause significant and permanent vision loss if not treated promptly. However, the immunosuppressive treatment of scleromalacia, and RA in general, carries its own risk of reactivating conditions such as a latent tuberculosis infection (LTBI).

**Case Presentation:** A 58-year-old man with a past medical history of RA presented with two months of right eye pain, redness, and blurry vision, as well as recent development of a dark blue/brown patch on his right eye. He was diagnosed with RA 25 years ago and had only received NSAIDs and prednisone due to his concerns about the side effects of disease-modifying antirheumatic drugs (DMARDs) and biologic agents.

On admission, all vital signs were within normal limits. Physical exam showed chronic deformities of both hands consistent with RA. Initial eye exam showed right eye conjunctivitis with a raised blue/brown protrusion from the right conjunctiva. Laboratory Results: were significant for an erythrocyte sedimentation rate of 26mm/hr, rheumatoid factor of 218 IU/mL, and anti-cyclic citrullinated peptide of 42.8 U/mL. Ophthalmology was consulted and a dilated fundoscopic exam found anterior scleral thinning with visualization of the uvea, consistent with anterior necrotizing scleritis and scleromalacia.

He was started on intravenous methylprednisolone for three days and experienced improvement in vision and eye pain. The initial treatment plan was to continue oral steroids and start cyclophosphamide or anti-tumor necrosis factor (anti-TNF) therapy. However, multiple serologic tests suggested the patient had an LTBI. Given the concern related to a potential LTBI, the patient was going to be started on methotrexate, and infectious disease was consulted for TB treatment recommendations. However, the patient left prior to initiating treatment.

**Discussion:** Scleromalacia peforans is a rare ocular manifestation of RA caused by the destruction of scleral matrix. If left untreated, it can lead to blindness, globe rupture, cataracts, or glaucoma in up to 60% of patients. Initial treatment typically begins with corticosteroids and immunosuppressants, such as cyclophosphamide or anti-TNF agents. Other DMARDs, like methotrexate, are less frequently used but have also been shown to be effective in treating scleromalacia. While more commonly used, cyclophosphamide and anti-TNF agents also have a well-established risk of LTBI reactivation, with methotrexate-induced LTBI reactivation being far less prevalent.

Regardless of which immunosuppressant is chosen, if an LTBI is suspected, treatment with an extended course of isoniazid or rifampin is typically completed prior to starting immunosuppression. However, in patients with significant morbidity or mortality from RA, concurrent LTBI and RA treatment is possible with isoniazid and methotrexate, with minimal side effects and little risk of TB reactivation. In balancing the risks of uncontrolled RA and reactivation of serious infections like LTBI, it is important that...
clinicians understand the risks of each treatment option so they can help their patients make an informed decision.

References

Title: Outside the (Thoracic) Box: Extracorporeal Support for Massive Pulmonary Embolism

Authors: Tripti Gupta, MBBS [1], Kevin Ergle, MD [2], [1] Department of Internal Medicine, Ochsner Clinic Foundation, New Orleans, LA, [2] Department of Cardiology, Ochsner Clinic Foundation, New Orleans, LA

Introduction: More than 250,000 Americans are hospitalized each year from venous thromboembolic disease. Those with massive pulmonary embolism (PE) have a mortality greater than 50%. Therapeutic options for these cases include thrombolysis and surgical embolectomy. Despite these treatment modalities, some patients have persistent shock and respiratory failure. This case describes use of extracorporeal membrane oxygenation (ECMO) for refractory cardiopulmonary failure after massive PE.

Case Presentation: A 42-year-old morbidly obese female with oral-contraceptive use presented with one-week history of dyspnea with exertion and mild chest discomfort. Electrocardiogram showed sinus tachycardia with S1Q3T3 pattern. Initial labs revealed elevated troponin I (0.623 ng/mL) and D-dimer (13.02 mg/L). A chest CT angiogram was ordered, but the patient decompensated prior to scan completion. Imaging was aborted. Patient had 92% oxygen saturations on a non-rebreather, was tachypneic, tachycardic, and hypotensive with a systolic blood pressure 80 mmHg. A bedside echocardiogram showed McConnell’s sign with right ventricular dilation and strain.

Systemic thrombolysis with intravenous alteplase was initiated. However, during administration, the patient developed progressively labored breathing and suffered pulseless electrical activity requiring cardiopulmonary resuscitation, intubation, and mechanical ventilation prior to return of spontaneous circulation after 10 minutes. Blood gas analysis demonstrated hypercapnic respiratory failure with pCO2 97 mmHg and lactic acid 9.5 mmol/L. The ventilator was adjusted to achieve minute ventilation >15 L/min; intravenous dobutamine and inhaled nitric oxide were used to improve right heart function. Despite these measures, pCO2 was persistently elevated and lactic acid >9 mmol/L, indicating dead space ventilation and right heart strain leading to poor perfusion. Cardiothoracic surgery was consulted for surgical embolectomy but the patient was deemed not to be a candidate for surgery.

It was determined that the patient required advanced cardiopulmonary support. Femoral arterial and venous access were obtained with 18-French and 24-French cannulas, respectively, and veno-arterial (V-A) ECMO was initiated. Over the ensuing hours, her acid-base status and lactic acid levels normalized. Creatinine levels peaked at 6 mg/dL however urine output was preserved, and this ultimately normalized. Repeat echocardiogram on post-ECMO day 4 revealed improvement in right ventricular function. She was subsequently decannulated post-ECMO day 4 and recovered without neurologic deficits.

Discussion: This case highlights successful life-saving therapy of extracorporeal membrane oxygenation (ECMO) in a case of massive PE refractory to thrombolytic therapy. By bypassing the failing right ventricle and lungs, the V-A ECMO can be utilized to provide full cardiopulmonary support as a bridge to definitive therapy or until recovery from acute pathology, as in this case. ECMO is still likely underutilized for this scenario, in part due to risks and physician unfamiliarity with this modality. Future research studies are warranted to delineate which patients may best benefit from extracorporeal interventions.
References

Title: A CASE OF MULTIPLE MYELOMA PRESENTING WITH AMYLOID ASSOCIATED MYOPATHY

Authors: Authors: Janet Manalac, Gregory Jacob, Kelvin Raybon

Department of Internal Medicine, University Hospital and Clinics, LSUHSC, Lafayette, LA

Introduction: Amyloidosis-associated myopathy is rare. Delay in diagnosis is common and there is a high rate of pathologic and clinical misdiagnosis.

Case Presentation: A 58-year-old woman presented to the hospital with acute onset, profound left lower extremity weakness and pain. She had no prominent constitutional symptoms except for generalized tiredness and weakness. On physical examination, she had mild temporal wasting, impressive left hip and knee extensor weakness, left pitting pedal edema, and good vascular perfusion. No other focal neurologic deficits were noted. She underwent extensive evaluation including comprehensive CT imaging of the lumbar spine, abdomen and pelvis that demonstrated diffuse enlargement of the musculature and a heterogenous appearing left iliacus muscle with hypodense areas, concerning for inflammatory or infectious myositis. A diagnosis of inflammatory myositis was unlikely due to normal CPK and aldolase levels. She was initially treated with IV antibiotics for presumed pyomyositis without improvement. Furthermore, routine laboratory evaluation revealed mild anemia, hypoalbuminemia, and significant proteinuria, but no hypercalcemia or renal insufficiency. Subsequent serum and urine protein electrophoresis showed prominent lambda chains in the urine with additional IgG lambda monoclonal protein in the serum and urine. Skeletal survey showed early lytic bone lesions. She underwent bone marrow biopsy that demonstrated increased plasma cells at 70% consistent with multiple myeloma. This did not completely explain her isolated left extremity weakness so amyloid infiltration was considered. Congo red stain was later performed on the same specimen, which confirmed amyloid deposition within the blood vessel walls. Treatment was initiated with dexamethasone, lenalidomide and bisphosphonates. She continues on this treatment regimen presently resulting in improvement of her symptoms.

Discussion: This case illustrates the important clinical and laboratory findings associated with multiple myeloma, and describes an uncommon complication, amyloidosis, resulting in profound myopathy. Classic presentation typically includes CRAB features (hypercalcemia, renal insufficiency, anemia, and bone pain). However, variable initial clinical findings and the absence of typical lab markers can make recognition of the disorder difficult in many patients. Amyloid myopathy should be a consideration in adults with muscular weakness of uncertain cause. Late diagnosis remains a major obstacle to initiating effective therapy. Hence recognizing the presenting syndromes is necessary for improving survival.
Title: Ochronosis: A rare cause for a common complaint

Authors: McLeod, C., Scopelitis, E.

Introduction: Ochronosis is a rare genetic disease that can cause lower back pain. It is estimated that ochronosis has a prevalence of 1 per 250,000 to 1 million cases. Ochronosis is distinguished by the triad of dark urine, ochronotic pigmentation and arthritis. Below is a case of a young man who presented with disabiling lower back pain.

Case Presentation: Patient is a 38 year old African American male with past medical history of diabetes mellitus and hypertension who presented to rheumatology clinic for progressive and worsening lower back pain over the past year. Patient described the pain as constant, non-radiating, nine out of ten pain, made worse with activity and standing and unrelieved with anti-inflammatory agents or heating pads. One year prior, patient was admitted to the hospital for right knee pain thought to be a crystal arthropathy with Xray of his knee showing calcification of his lateral collateral ligament, however arthrocentesis showed non-inflammatory cell counts with no crystals. Patient denied trauma. Family history was pertinent for a cousin with similar chronic lower back pain. Additionally patient noticed that his urine would turn black if left overnight in the toilet bowl. Physical exam was pertinent for scleral pigmentation with tenderness along lumbar spinal processes L1-L5 with a positive Schober’s test and normal chest expansion. The patient had an Xray of his lumbar spine which exhibited calcification of all lumbar spine intervertebral disc spaces consistent with possible ochronosis, hemochromatosis or hyperparathyroidism. Patient had normal parathyroid hormone level and iron saturation. The patient’s urine was sent for organic acid testing which showed markedly elevated excretion of homogentisic acid of 1351 mmol/mol creatinine which was diagnostic for ochronosis.

Discussion: It is estimated that eighty-four percent of adults will develop lower back pain throughout their lives. The signal anatomic site for ochronosis is the lower back with sparing of the sacroiliac joints, followed by the knees and hips. Ochronosis is a rare autosomal recessive disorder caused by an inborn error in tyrosine metabolism due to a lack of homogentisic acid oxidase. Excess homogentisic acid then undergoes autoxidation and polymerization leading to ochronotic pigment accumulation in cartilage and connective tissues. Many early manifestations of this disorder such as dark urine, dark cerumen and pigmentation of the earlobe skin and sclera may go unnoticed by the patient. Cases that escape detection in childhood typically are diagnosed in the third decade of life with chronic joint pain such as lower back or knee pain. We present this case of ochronosis to bring awareness to a rare and potentially disabling genetic disease that may present later in life to prevent misdiagnosis in the future.
Title: An Unexpected Culprit for Right Lower Quadrant Pain

Authors: Ronak Patel, M.D, Naga Turaga, M.D, Shashitha Gavini, D.O, Farha Khan, M.D., University Hospital and Clinics, Lafayette, Louisiana

Introduction: Abdominal pain accounts for 1.5% of office based visits and 5% of emergency room visits. Right lower quadrant (RLQ) pain is one of the most frequently encountered problems in clinical practice with many differentials like nephrolithiasis, pyelonephritis, renal cell carcinoma, appendicitis and diverticulitis. We present here a case of very unusual etiology of RLQ pain.

Case Presentation: A 54-year-old Caucasian female with a history of chronic back pain, gastric bypass surgery, and hysterectomy for fibroids presented with right sided flank pain for a month. She attributed the pain to her chronic back pain and took over the counter analgesics without any resolution. Vitals were normal and physical exam was significant for tenderness to palpation in right lower quadrant extending to lower back. Labs were significant for creatinine of 1.05 mg/dl, 11 red blood cells/hpf on urinalysis and a negative urine pregnancy test. Computerized tomography of abdomen and pelvis without contrast was performed to rule out nephrolithiasis, but instead revealed right ovarian vein thrombosis (OVT). Patient had no known thrombophilia or familial history of blood clots and workup for bleeding diathesis was normal. She was started on apixaban for 3 months and discharged home. Outpatient workup for malignancy including pap smear and colonoscopy were normal. Mammogram revealed category 4 BI-RADS lesion of right breast and biopsy was negative for malignancy. Patient was followed in the clinic in 3 months and her symptoms resolved.

Discussion: OVT is a rare condition occurring in 0.02-0.18% of pregnancies in the postpartum setting. Review of literature showed only five reported cases in non-pregnant setting. Right OVT is more common (70-90% of cases) as right ovarian vein is longer and lacks competent valves. OVT typically presents as pelvic pain (55%), fever (80%), and rarely as a right-sided abdominal mass. Delay in the diagnosis and treatment can lead to life-threatening complications such as thrombus extension into the IVC which can potentially lead to a pulmonary embolism in 25% of patients with untreated OVT. The mortality in these patients can reach about 4%.

Hence, OVT should be considered in the differential diagnosis for unexplained lower quadrant pain to prevent fatal complications. And a detailed workup for pregnancy, malignancy and bleeding diathesis should be pursued when OVT is found.
Title: Aberrant Right Subclavian Artery: A Rare Cause of Dysphagia

Authors: Corey Saraceni, MD. Tejas Joshi, MD. Wynn-C Kwan, MD. Melissa Spera, MD. Stephen Landreneau, MD FACG

Introduction: Aberrant right subclavian artery (also known as Arteria Lusoria) is the most common congenital anomaly of the aortic arch occurring in 0.5% to 1.8% of the population based on cadaveric studies. Aberrant right subclavian artery is caused by the involution of the right 4th aortic branch and proximal right dorsal aorta with a persistent distal right dorsal aorta and 7th intersegmental artery forming the right subclavian artery. The retro esophageal course of the right subclavian artery behind the esophagus, although usually asymptomatic, may cause compression and a type of dysphagia known as dysphagia lusoria.

Case Presentation: A 50 year old female with a medical history of von Willebrand disease, hypertension, asthma, and peripheral neuropathy presents with a complaint of several weeks of progressive dysphagia associated with heaviness in her chest. Her dysphagia began with solid foods and progressed to include liquids. Three days prior to presentation, she could not tolerate any solids or liquids by mouth and would have immediate regurgitation. The patient admits to about 60 lbs weight loss in the past two months.

Follow-Up Course

- CT neck angiography revealed a congenitally anomalous aortic arch with a retroesophageal aberrant right subclavian artery reaching as far superiorly as the T1 vertebral body.
- Barium Esophagram: No evidence of an intrinsic mass, polyp, diverticulum, or stricture. There is mild extrinsic mass effect on the posterior aspect of the upper esophagus however this caused no limitation of contrast passage.
- EGD was performed and revealed a normal appearing esophagus, stomach, and duodenum. There was no endoscopic evidence of esophageal neoplasia, mucosal abnormalities, or eosinophilic components on biopsy.
- Esophageal Manometry: Normal manometry. Normal LES with normal relaxation. 100% swallows peristaltic

Discussion:

Most patients with aberrant right subclavian arteries remain symptom-free throughout their lifetimes. Occasionally symptoms may develop during early childhood, usually presenting as recurrent pulmonary infections and respiratory abnormalities. The mechanism is unknown why dysphagia develops in older adults. Various proposed mechanisms including age related increased esophageal rigidity, right subclavian aneurysm formation, and elongation of the aorta.

Diagnosis of aberrant right subclavian artery as the cause of dysphagia remains challenging. Barium swallow remains an effective tool for initial evaluation usually showing a characteristic diagonal
impression in the esophagus at the level of 3rd-4th vertebra. Esophagogastroduodenoscopy (EGD) may reveal a pulsating mass at around the same level. Esophageal manometry may reveal a high-pressure zone 25-30cm from the nose. CT angiography, angiography of the aortic arch, or endoscopic ultrasound may be used for definitive diagnosis.

Initial treatment should be conservative management using prokinetic or antireflux drugs. Surgical treatment may be attempted in those who do not respond to conservative management. In patients unsuitable for a surgical procedure, endoscopic dilation may temporarily relieve symptoms.
Title: Strongyloides Hyperinfection Syndrome

Authors: Lauren Schober, MD¹ & Shoba Vootukuri, MD²,¹ LSU Health Baton Rouge Internal Medicine
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Introduction: *Strongyloides stercoralis* is a nematode capable of entering the human host by invading the skin in the filariform state through fecal transmission. It enters the blood stream and is transported to the lungs. After ascending the tracheobronchial tree, it is swallowed into the GI tract. There it reproduces and continues to re-infect the patient by translocating, often along with other gram negative bacteria, across the gut wall and into the blood stream. During autoinfection, the parasite burden can become high enough to cause systemic symptoms resulting in hyperinfection syndrome. Risk factors for developing hyperinfection syndrome include immunosuppression secondary to AIDS, underlying hematologic malignancy, and immunosuppressive medications. Here we present a case of *Strongyloides* hyperinfection in a patient without classic risk factors for an underlying *S. stercoralis* infection.

Case Presentation: A 75 year old Caucasian male from Louisiana with a history of Glioblastoma Multiforme (GBM) status post-surgical resection, chemotherapy, and radiation three years prior presented to the emergency department (ED) with cough and hypoxemia one week after an increased dose of dexamethasone for progression of GBM. On admission patient was afebrile, tachycardic, and normotensive. Physical exam was notable for bibasilar crackles in the lungs and a petechial rash in the gluteal region. Lab work was significant for leukocytosis and eosinophilia. Chest x-ray was concerning for pulmonary edema, while CT revealed bilateral mid and upper lung interstitial and ground glass opacities. There was concern for sepsis secondary to aspiration pneumonia versus acute heart failure. Patient was started empirically on vancomycin and piperacillin/tazobactam and received a low dose of furosemide. Sputum culture resulted positive for *S. stercoralis* larvae. Subsequent work up revealed increased IgE antibodies and positive Strongyloides IgG antibodies in the serum. HTLV and HIV were negative. Patient was started on Ivermectin for treatment of *S. stercoralis* hyperinfection and steroid dose was decreased. Despite treatment, patient declined and passed away during his hospitalization. The patient had no history of traveling outside the country to *S. stercoralis* endemic areas. Retrospective chart review revealed a chronic, underlying eosinophilia for several years prior to presentation that had been worked up with stool ova and parasite studies which were negative for commonly tested parasites.

Discussion: Often overlooked, the southeastern region of the U.S. is endemic for *S. stercoralis*. *Strongyloides* infections are mostly reported in veterans, immigrants, and rural populations. The low prevalence, however, may be due in part to infrequent testing. This case demonstrates that patients in southern regions of the U.S. with unexplained eosinophilia and pulmonary or gastrointestinal symptoms may benefit from a lower threshold of suspicion for *S. stercoralis* infection.

References

Title: An Unusual Infection Acquired by a Man after Biting a Dog

Authors: Naga Sai Shravan Turaga MD, Jay Patel MD, Justin Pratt MD, Babak Amrollahie MD, Karen Curry MD, Nicholas Sells MD

Introduction: Injuries from a dog bite account for 85-90% of animal bites in the United States. Some of the well-known human infections related to dogs include bacterial like pasteurella, streptococcus, staphylococcus and anaerobes native to either the canine's mouth or human's skin. We present here a significant and rare infection related to canines, where there was no incidence of an animal biting a human, but instead, the converse.

Case Presentation: A 36-year-old male presented with complaints of fever, chills, arthralgia and generalized malaise which began two days after he bit on the neck of one his pit bull to break a dog fight. Unfortunately, the dog died three days later of an unknown cause. Initial vitals and physical exam were significant for temperature of 38.3 C and a well healed scab on his right shin. Labs revealed hemoglobin 11.5 mg/dl, WBC count 7700/mcL, platelet count 17,000/mcL, lactate dehydrogenase 255 U/L, creatinine 1.56 mg/dl, BUN 30mg/dl and lactic acid 2.5 mmol/L. Blood cultures were drawn and intravenous (IV) vancomycin and piperacillin-tazobactam, oral doxycycline and rabies vaccine course were started. Vancomycin was stopped after cultures grew gram negative bacteria. Serology for ehrlichiosis and anaplasmosis was negative. Typing of the organism was difficult, so the specimen was sent to reference lab. Two-week course of IV piperacillin-tazobactam, oral doxycycline and oral levofloxacin was completed for high suspicion of Capnocytophaga infection with resolution of symptoms and normalization of blood counts and kidney function on followup in clinic after discharge. Five weeks after discharge, the Results: of typing were obtained as Capnocytophaga canimorsus.

Discussion: Capnocytophaga canimorsus, a normal oral flora of canines is usually transmitted by bites or scratches and mainly affects immunocompromised individuals. We suspect our patient acquired the infection through wound on his shin from dog cage which likely had some saliva or slobber. It can cause pneumonia, cellulitis, meningitis, endocarditis, septic arthritis, or sepsis leading to septic shock with reported fatality rates of 28-31%. Diagnosis is mainly by culture but being a fastidious organism, it can be difficult to isolate. Microscopic examination of peripheral smear oruffy coat can also be used in diagnosis, specially in asplenic patients. Beta-lactam beta-lactamase combinations or carbapenems are the antibiotics of choice, which explained our patient’s response to piperacillin-tazobactam. Amoxicillin-clavulanate is the prophalaxis of choice for high risk patients (immunocompromised patients, asplenic patients, patients on immunosuppression, alcoholics) after dog bite.

In conclusion, diagnosis of Capnocytophaga infection is challenging as identification of organism can be difficult and untimely. Hence, clinicians should have a high degree of suspicion in the right clinical circumstance and have a low threshold to start empiric therapy.

References
Maine-Clinical Vignette-Poster Finalist
Kiera M McElrone, DO

Title: Drunk When You're Not: A 67 year old male presents with feeling "off balance"

Authors: Kiera McElrone, D.O.

Introduction: The following is a case presentation of a man who presented with gait instability and a feeling of drunkenness. Review of his history revealed that he had many of the prodromal symptoms associated with Multiple-System Atrophy, a rare fatal neurodegenerative disorder. A multidisciplinary approach to management spanning multiple specialties is helpful in patients with Multiple-System Atrophy. Currently most treatment is supportive. Further research is needed to identify potential therapies for this fatal disease.

Case Presentation: A 67 year old gentleman presented in October of 2015 reporting that he was "off balance" for "awhile now" and that when he walked, "he felt like he was drunk." The gentleman said this started after a right hip injury two years prior. He had difficulty swinging his tennis racket, multiple falls, and occasional slurred speech. In the months prior to this call, he had developed nighttime urinary incontinence. Past medical history was significant for obesity, Rheumatoid Arthritis, REM sleep disorder, and Erectile Dysfunction. His only medications were sildenafil and plaquenil. On exam, his vitals were normal. He was obese. His neurological exam was significant for mild dysarthria, dysmetria with abnormal heel to shin and finger to nose right > left, stooped posture and a wide-based gait with decreased arm swing bilaterally. Laboratory investigations including B12, TSH, and CPK were all within normal limits. An urgent MRI was performed that was nonspecific. He was referred to neurology and was seen in January of 2016. An EMG was negative, and he was referred to a Movement Disorders Clinic where a DaT- SPECT scan was performed demonstrating decreased activity in the putamen and caudate nuclei. He was diagnosed with probable Multiple-System Atrophy. In May of 2017, he began to develop sudden leg weakness and was choking on meals. Vitals were significant for systolic blood pressure of 70 and he had started to develop further progressive autonomic failure.

Discussion: Multiple-System atrophy is a rare fatal neurodegenerative disease that presents in adulthood and is characterized by progressive autonomic failure, parkinsonian features, and cerebellar and pyramidal features. It poses a diagnostic dilemma across multiple specialties including neurology, cardiology, urology, gastroenterology, and sleep medicine. It is an orphan disease with a mean incidence of only 0.6 to 0.7 cases per 100,000 person-years. It often has a prodromal premotor phase characterized by sexual dysfunction, urinary urge incontinence, orthostatic hypotension, inspiratory stridor, and REM sleep disorder. Interestingly, our patient had experienced several of these symptoms months prior to his gait instability. Most patients die within 8-10 years of developing symptoms. Death is due to sepsis secondary to urinary tract infections or pneumonia, bilateral vocal-cord paralysis, or disruption of the brain-stem cardiorespiratory system. Treatment is supportive. It is important to consider Multiple-System Atrophy in patients presenting with the premotor symptoms above.
Maryland-Clinical Vignette-Poster Finalist
Khalid Hajjir

Title: A Fat Bird – A Rare Cause of Dyspnea

Authors: Khalid Hajjir MD, Ali Kadhim MD, Kanwaldeep Virk MS 3, Sapna Kuehl MD, Saint Agnes Hospital, Baltimore, Maryland

Introduction: Achalasia is a rare disease of the esophagus with prevalence and incidence is estimated to be 1.63/100,000 and 10.82/100,000, respectively. Prevalence is increasing as individuals live longer, however incidence remains stable. The commonest presentations are dysphagia, regurgitation and chest discomfort.

Case Presentation: An 89-year-old African American woman presented to the ED with a complaint of progressive dyspnea over 2 months, which was partially relieved with rest and use of an inhaler. S.O.B was associated with dry cough, wheezing and chest tightness, more pronounced at meal times. She denied any prior diagnosis of a respiratory condition, the use of supplemental oxygen at home or any prior hospitalizations for similar events. She also denied odynophagia and dysphagia, sick contacts, orthopnea or constitutional symptoms. She denied use of tobacco, alcohol or recreational drugs. Patient is active at baseline and helps take care of a disabled friend.

On presentation, Heart rate was 125/minute and respiratory rate 31/minute with mild bilateral expiratory wheezes throughout. On initial evaluation by chest X-ray, a prominent cardiome diastinal silhouette was seen. On chest CT; a torturous massively dilated esophagus of 10.6 cm was visualized. A barium swallow study showed the classic bird beak appearance. An EGD was notable for initial resistance in passing the scope at esophagogastric junction with 2 liters of residual fluid and food that were evacuated. Diffuse candidiasis was observed and no masses were found.

Discussion: Achalasia is a neurodegenerative disease that involves the nitric oxide-releasing inhibitory neurons that affect the myenteric plexus and prevents smooth muscles in lower esophageal sphincter from relaxing. It can be divided into two categories; primary (idiopathic) and secondary (autoimmune, inflammatory or infectious causes). Dysphagia for both solids and liquids is the hallmark of achalasia; patients also report regurgitation of undigested food material, difficulty belching and non-cardiac chest pain and odynophagia. Respiratory symptoms have been reported in a single center study, and ranged from cough, aspiration, hoarseness, wheezing, shortness of breath, and sore throat. In our case, the presentation was atypical as her symptoms were primarily respiratory in nature. Treatment of achalasia can be divided into surgical and nonsurgical approaches. In this case, given her age and risk factors, options were limited. She underwent botulinum toxin injections as the initial management strategy.

In conclusion, Achalasia is a rare disease that presents with non-specific symptoms and can mimic more common conditions. It is a rare but important cause of chest discomfort and high suspicion for this condition is important in the differential of chest pain and dyspnea.

References

Maryland-Clinical Vignette-Poster Finalist
An T Ho, MD

Title: A case report of multi-organ failure due to catastrophic antiphospholipid syndrome in a young woman with six normal pregnancies and potential role of eculizumab

Authors: An Ho, MD

Introduction: Catastrophic antiphospholipid syndrome (CAPS) is the extremely rare but fatal form of APS with multiple organ thromboses (1). While antiphospholipid is well-known to cause pregnancy related fetal complication. We have a case of a young woman with six children and no adverse fetal outcomes. She presents with acute pulmonary renal syndrome, acute heart failure and hemolytic anemia. This case also highlights the use of eculizumab in refractory cases.

Case Presentation: A 34-year-old woman presented with acute dyspnea. She developed dyspnea and a productive cough 2 days ago. She was in hypoxic respiratory failure and hypertension of 200/120 mmHg. She had a creatinine of 3.5 mg/dl, leukocytosis and hemoglobin of 7.7 g/dl. Chest CT scan showed airspace opacification bilaterally. An echocardiography showed a severely depressed left ventricular systolic function without valvular or wall motion abnormality. She was subsequently intubated, started on broad spectrum antibiotics and diuresis. Hemodialysis was started 2 days later. Bronchoscopy yielded fluid consistent with diffuse alveolar hemorrhage. Further work up revealed hemolytic anemia with few schistocytes, ADAMST of 55%, ANA of 1:1280, IgG1 of 1000 mg/dl, decreased C3, C4 level and elevated level of beta2-glycoprotein-1 IgM antibody of 135 U/ml. A kidney biopsy showed chronic active thrombotic microangiopathy. CAPS was diagnosed. In retrospect, she had six pregnancies with no fetal adverse outcome. Heparin, IV steroids and later, five sessions of plasma exchange then started leading to some clinical improvement, but not hemolytic anemia. Eculizumab was added after meningococcal vaccine was administered. Hemolytic anemia then resolved. She was maintained on Eculizumab weekly and following stabilization every 2 weeks. She remained in remission 3 months after.

Discussion: Our patient is a diagnostic challenge because of her complicated manifestation and prior history not suggestive of APS. Fetal adverse outcomes in APS can occur in 30% of pregnant patients with APS but do not always exist (3). She met the four CAPS criteria: microthrombosis (kidney biopsy), high titer of anti beta2-glycoprotein-1 IgM, more 3 system involvements (kidney, heart, lung, hematology) and development in less than a week (4). Atypical hemolytic uremic syndrome, malignant hypertension are unlikely because of high titer of APS antibodies and few schistocytes. The major organs involved in CAPS are renal (71%), lung (64%), brain (62%), and heart (51%)(1). Hemolytic anemia is due to thrombotic microangiopathy. Diffuse alveolar hemorrhage presents in 6% of patients with an unclear pathogenesis (2). The combination of anticoagulation, glucocorticoids, and plasma exchange achieved a recovery rate of 78% (5, 6). Recently, there is new evidence of excessively increased all three complements pathways in CAPS, finally leading to activated terminal pathway(7). Eculizumab, a terminal pathway inhibitor was effective in a few resistant case reports (7, 8). Our case represents a successful treatment of CAPS with eculizumab.

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Title: Fatal Intoxication with N-ethylpentylone

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Introduction:
Synthetic cathinones represent the latest genre of new drugs of abuse and is increasing popularity in part due to their ease of availability and inability to be detected by routine drug testing. They provide a cheaper substitute to traditional stimulant drugs and are sold on the internet and in retail establishments as "bath salts", "plant food", or "research chemicals".1 This case report details the behavior effects and clinical presentation of N-ethylpentylone drug intoxication.

Case Presentation: 21-year-old man left his house to smoke marijuana and returned, per his girlfriend, “acting crazy.” Law enforcement was dispatched to a disturbance call. He was described as combative, confused, sweating heavily and at one point attempted to break into a police vehicle. Paramedics administered 5 mg of intramuscular haloperidol. He went into cardiac arrest shortly thereafter. ACLS was initiated and ROSC was achieved within 3 minutes. In the ED his blood pressure was 95/55 mmHg, heart rate 126 bpm, respiratory rate 25 bpm and oxygen saturation 99% on 40% FiO2. There were multiple abrasions on his face, sluggish pupillary reflexes, negative vestibulo-ocular reflex, and myoclonus at the right lower extremity. Clinical laboratory analysis revealed potassium 6.8 mmol/L, glucose 28 mg/dL, CPK 451,160 IU/L, lactic acid 28 mg/dL (without an osmolar gap), AST 12,374 IU/L, ALT 7,649 IU/L, and creatinine 1.70. ABG showed severe metabolic acidosis with pH 6.80. The patient was admitted to the ICU and treated with hypothermia cooling measures, intravenous fluids, and bicarbonate. Toxicology screening was positive for cannabinoid and an ethanol level of 12 mg/dL. MRI brain showed bilateral restricted diffusion in the posterior parietal and occipital regions suggesting profound cerebral hypoxia. Day 2, the patient was started on CRRT due to oliguria and rising creatinine levels. Despite supportive treatment, he developed severe hypothermia with a temperature of 35.3°C, DIC and hypotension requiring vasopressors. Day 3, the patient arrested again. Cardiopulmonary resuscitation was performed for 30 minutes before the patient was pronounced dead. An autopsy was performed at the Office of the Chief Medical Examiner. Post-mortem toxicology testing found N-ethylpentylone in the urine using gas chromatography and mass spectrometry. Based on the autopsy findings, clinical presentation, and hospital course, the official cause of death was deemed drug intoxication with N-ethylpentylone.

Discussion: Information regarding the exact pharmacology of N-ethylpentylone is limited. Similar to amphetamines, synthetic cathinones act on the central nervous system by inhibiting monoamine neurotransmitters dopamine, serotonin, and norepinephrine via facilitation of extracellular release and reuptake inhibition.1 2 However, they differ from amphetamines in their affinity to monoamine transporters and their mechanisms of function. Until we have a better understanding of the mechanisms of this drug, it is imperative that clinicians recognize signs and symptoms of potential intoxication with N-ethylpentylone, including behavioral effects, tachycardia, acidosis, rhabdomyolysis, and multi-organ
failure. At this point guidelines for clinical management are not available; therefore, supportive measures with intravenous fluids, bicarbonate, mechanical ventilation and cooling measures are part of the current “standard” of care.

References


Title: Intrapleural Fibrinolytic and DNase for Management of Lung Entrapment due to Complicated Parapneumonic Effusion

Authors: Strumpf Z, Grier W, Kalcheim-Dekel O, Paparo M, Pickering E, Burrows W, Sachdeva A

Introduction: Lung entrapment due to active pleural inflammatory process is a known complication of pleural space infection, or complicated parapneumonic effusion. If left untreated it may progress to trapped lung as a result of mature fibrous membrane overlying the visceral pleura. Management of the resultant unexpandable lung requires video-assisted thoracoscopic surgery (VATS) with decortication of the visceral pleura and carries significant morbidity and mortality. Intrapleural fibrinolytic and DNase therapy has shown to reduce hospital length of stay, need for thoracic surgery in pleural infections; however, its effectiveness in lung entrapment from parapneumonic effusion is unknown. We present a case wherein the combination therapy was successful in treatment of lung entrapment 3-month post-pneumonia.

Case Presentation: A 76-year-old gentleman, former light smoker, presented with a 3-month history of pleuritic chest discomfort and dyspnea on exertion that started after an episode of productive cough, congestion, and intermittent night sweats that persisted despite two separate courses of oral antibiotics. Initial evaluation of the patient revealed normal vital signs including an oxygen saturation of 98% on room air with physical examination notable for decreased breath sounds on the right. Laboratory evaluation was unremarkable except for elevated C-reactive protein 3.4mg/dL. Chest radiograph demonstrated right lung opacity, confirmed on computed-tomography (CT) scan to be a loculated, right-sided pleural effusion with associated pleural thickening but without parenchymal opacities or intrathoracic lymphadenopathy. After a multi-disciplinary team discussion, the patient was admitted to the hospital and underwent tube thoracostomy. Pleural fluid analysis revealed an exudative, lymphocyte-predominant, effusion with a pH of 7.5 and negative Gram stain, AFB smear/culture, fungal culture and cytology. Repeat CT showed decreased size though persistence of a complex effusion with multiple air pockets within the collection and pleural thickening, suggestive of lung entrapment/trapped lung. The patient underwent instillation of tissue plasminogen activator (10 mg) and dornase (5 mg) with a dwell time of 1 hour followed by drainage to suction for a total of 6 doses. He experienced significant improvement in clinical symptoms including ability to take a deep breath without pleuritic discomfort. On a follow up visit, he endorsed near-resolution of his symptoms with repeat CT consistent with clinical response. He continues to do well on a two year follow up.

Discussion: To our knowledge this is the first report of therapeutic effectiveness of this combined therapy in the management of long-standing complex pleural effusion. Evolution of lung entrapment into a trapped lung is a conundrum and symptomatic patients usually require surgical management. As there is no discrete transition point between lung entrapment and trapped lung, we postulate that intrapleural fibrinolytic and DNase has a potential role in management of longer-standing, complex effusions and offers a less morbid approach. This approach deserves further study in this sub-group of patients.

References

Maryland-Clinical Vignette-Poster Finalist
Christine Zhang

Title: Pleuroparenchymal Fibroelastosis (PPFE); a Rare Disease Entity, Secondary to Methotrexate?

Authors: Christine Zhang, Ji Hyun Rhee

Introduction: PPFE is a newly described interstitial lung disease characterized by an upper lobe predominance of pleural thickening, sub-pleural elastosis, and intra-alveolar collagenous fibrosis. PPFE is typically seen in non-smokers with severe dyspnea and often misdiagnosed as asthma or pneumonia. While the etiology is unclear, PPFE has been described in association with drugs, radiation, lung/stem cell transplant, inhalation exposures, infections, and co-existence with usual interstitial pneumonia. PPFE can be rapidly progressive to involve the whole lung and carries a poor prognosis. There is no known effective therapy other than lung transplant. Here, we report a patient with rheumatoid arthritis and PPFE who had improvement after stopping methotrexate.

Case Presentation: A 59-year-old female with rheumatoid arthritis on methotrexate for 10 years and asthma presented to the pulmonary clinic with un-resolving dry cough for 6 weeks despite a few courses of steroids, nebulizers/inhalers and antibiotics. She had no occupational or environmental exposures. CT showed interstitial fibrotic changes along with scattered areas of superimposed patchy ground glass opacities. Pulmonary function test (PFT) showed mild obstruction without evidence of restriction, but low diffusion (DLCO 68%). Bronchoscopy with bronchoalveolar lavage ruled out infectious causes. VATS wedge biopsy showed areas of fibroelastotic interstitial fibrosis with pleural and subpleural scarring with elastosis, consistent with PPFE. At the follow up visit 1 month after VATS biopsy and after discontinuing methotrexate for 6 weeks, patient reported a dramatic improvement in her cough. Repeat PFT showed similar mild obstruction but greatly improved diffusion (DLCO 89%). Repeat HRCT showed previously seen scattered patchy ground glass opacities had resolved. She is currently doing well on leflunomide.

Discussion: PPFE is a newly identified entity; first described in 2004 and included in American Thoracic Society classification of idiopathic interstitial pneumonia in 2013. Approximately 130 cases have been described in the literature to date. Diagnosis is usually made with histopathology. Clinical outcome in PPFE is variable, with a significant number of patients demonstrating progressive decline and death. Survival characteristics depend on the stage of the disease at presentation. Most cases are considered idiopathic, but could be methotrexate induced as seen in this patient and should be considered as a possible adverse effect.

References

Title: “Diarrhea, The Tip of The Iceberg” a Fatal Case of Enteropathy Associated T-cell lymphoma.

Authors: Aggarwal, Abhimanyu MD; Alarcon Velasco, Sylvia MD; Murali, Rangan MD

Introduction: Enteropathy Associated T-cell lymphoma (EATL) is a rare gastrointestinal non-Hodgkin’s lymphoma, that arises from intraepithelial intestinal T-lymphocytes. It is associated with celiac disease and is usually diagnosed in terminal stages if celiac disease is diagnosed in later stages of life. We are presenting a patient with EATL with grave outcome.

Case Presentation: A 67 year old woman presented with right upper quadrant abdominal pain and was diagnosed with acute cholecystitis. She underwent elective cholecystectomy, complicated by Clostridium difficile colitis. Despite treatment with oral Vancomycin, she had persistent diarrhea and failure to thrive for a month. Repeat Clostridium difficile toxin was negative. Esophagastroduodenoscopy showed erosive gastritis and normal duodenal mucosa with biopsy showing chronic duodenitis and villous atrophy. Immunohistochemistry demonstrated increased number of lymphocytes on CD8 and CD3 stains within surface epithelium and within the crypts. Tissue Transglutaminase Antibody IgA (TTGA) was positive.

Despite being on gluten-free diet for celiac disease, she returned with watery diarrhea, severe abdominal pain and hemoccult positive stools. CT scan demonstrated spontaneous perforation of mid-jejunum and multiple "lesions" in jejunum. She underwent small bowel resection and primary anastomosis. Intra-operatively, lesions were noted along the jejunum with desmoplastic reaction on the surface, with thickened mesentery and enlarged mesenteric lymph nodes. Patient became hemodynamically unstable requiring pressor support, followed by multi-organ failure.

Due to rapid deterioration, family made patient comfort measures only and patient passed away soon after. Later, immunohistochemistry of jejunal specimens returned positive for CD3, CD43, bcl-2 and Ki-67 (60%) expression and negative for CD5. Morphologic and immunophenotypic findings were consistent with Enteropathy Associated T-cell lymphoma.

Discussion: EATL accounts for less than 5% GI lymphomas and less than 1% of all Non-Hodgkin lymphomas. Management of celiac disease at an early stage with strict gluten free diet adherence is associated with risk reduction of EATL.

Presentation range from malabsorption, diarrhea to acute abdominal pain arising from obstruction, perforation or bleeding. EATL mostly involves the small bowel and occasionally colon or stomach.

Intestinal findings include villous atrophy, crypt hyperplasia and intraepithelial lymphocytosis invading the intestinal wall at variable depths. Immunophenotype is CD3+, CD5-, CD4-, CD8+-/-, CD30+, CD56-, TCR beta +/- and CD 103+ and its associated to positive TTGA, IgA anti-endomysial or IgG Deamidated Gliadin Peptide.

There is no standard of care for newly diagnosed EATL. Multimodality approach is warranted based on clinical scenario including combination chemotherapy, surgery and autologous stem cell transplantation. Targeted therapies, like JAK/STAT inhibitors and Brentuximab vedotin (anti-CD30) are being investigated.
References


Massachusetts-Clinical Vignette-Poster Finalist
Jamal Akhtar, MBBS

Title: An Uncommon Cause of Painful Leg Ulcers

Authors: Jamal Akhtar, MD (Associate), Amos Lal, MD (Associate), Alwyn Rapose, MD, FACP, Department of Medicine, St. Vincent Hospital, Worcester, MA

Introduction: Venous, arterial, and neuropathic ulcers account for up to 90 percent of leg ulcers. Uncommon causes include physical injury, infection, vasculopathy, vasculitis, pyoderma gangrenosum, panniculitis (erythema induratum), and malignancy. Often in clinical practice we encounter cases where an uncommon disease presents with an unfamiliar staging. We present one such scenario of non-healing ulcers.

Case Presentation: A 49 year old Vietnamese female presented with painful ulcers on her right foot of 8 weeks duration. Initially she developed painful swellings that had broken down to form these ulcers. Prior to presentation she had already received three courses of antibiotics (cephalexin, trimethoprim/sulfamethoxazole, clindamycin) with no response. Physical examination revealed two ulcers on the medial aspect of right foot with irregular margins, poorly healing granulation tissue at the bases and surrounding edema, and six tender subcutaneous nodules on both thighs and groins. She was found to have bilateral ankle edema as well. There was no inguinal lymphadenopathy. She had a strongly positive skin tuberculin test. Histopathology of the ulcers revealed mixed lobular and septal panniculitis with epitheloid and multinucleated giant cells, confirming the diagnosis of erythema induratum of Bazin (EIB). The patient was treated with anti-tubercular drug regimen, resulting in complete resolution of ulcers and subcutaneous nodules.

Discussion: Cutaneous TB is a relatively uncommon manifestation of TB, accounting for only 1 to 2 percent of all cases. EIB remains one of the rarely encountered tuberculids, although tuberculosis is known to be endemic in the developing countries. Tuberculids are commonly considered to be cutaneous hypersensitivity eruptions to *M. tuberculosis* that occur in patients with a moderate or high levels of immunity against the organism. EIB is a granulomatous lobular panniculitis of the lower extremities, classically presenting with mildly tender, dull red, subcutaneous nodules on the lower legs, over the course of several weeks. The nodules are usually grouped on the lower third of the legs, especially around the ankles. The nodules are tender and breakdown of the nodules often occurs, leading to the formation of deep, draining ulcers. The diagnosis of TB-associated EIB is made based upon recognition of the clinical features, histopathologic findings, and evidence for *M. tuberculosis* infection. Bacilli are usually not detectable in tuberculid lesions. Tuberculosis is uncommon in Western countries, and to make it even more challenging, EIB is one of the uncommon presentation of tuberculosis. Although rare, it should be considered as a cause of painful leg ulcers not responding to conventional therapy, especially in a host coming from endemic areas.
Massachusetts-Clinical Vignette-Poster Finalist
Mohammed Yousef Al Mulhim, MD

Title: ANCA-Associated Vasculitis: Incriminating an Old Friend

Authors: Mohammad Al Mulhim, MD (Associate), Suzanne Martin, MD Department of Medicine, Saint Vincent Hospital, Worcester, MA

Introduction: Hydralazine is widely used as an adjunctive treatment for hypertension. One of the rare side effects of hydralazine is ANCA-associated glomerulonephritis (AAV).

Case Presentation: An 81-year-old female with a history of type 2 diabetes mellitus, hypertension and chronic kidney disease stage 3 (baseline creatinine of 1.5 mg/dL) presented to emergency department with fatigue and lightheadedness. She was being treated with hydralazine for 2 years. Physical exam was unremarkable with stable vital signs. Initial labs showed a creatinine of 3.38 mg/dL. Urinalysis was positive for moderate blood and protein of >300 mg/dL. Urine sediment showed dysmorphic RBCs. Additional labs showed a positive ANA and atypical pANCA at 1: 320 with a high anti-MPO titer. Serology was negative for anti-histone antibody, anti-GBM antibody, anti-dsDNA antibody and Smith antibody. Hepatitis B surface antigen, hepatitis C antibody, and HIV 1 and 2 antibodies were also negative. Her C3 and C4 levels were normal. Kidney biopsy was performed and showed focal proliferative, necrotizing and crescentic glomerulonephritis, immune-complex mediated with sparse deposits reactive for all Igs and C3.

She was diagnosed with hydralazine-induced AAV. Hydralazine was promptly discontinued and she was started on steroids and rituximab.

Discussion: Anti-neutrophil cytoplasmic antibody-associated vasculitis has been associated with many drugs, including hydralazine, propylthiouracil, minocycline, phenytoin, penicillamine, allopurinol, and sulfasalazine. The diagnosis of drug-induced AAV is based on the temporal relationship between clinically evident vasculitis and administration of the offending drugs, and excluding medical conditions that mimic vasculitis and other definable types of vasculitis. It usually manifests very high titers of MPO-ANCA but a small number of patients present with PR3-ANCA. Kidney biopsy shows necrotizing and crescentic GN and immunofluorescence microscopy stain positive for Ig and C3. The management includes discontinuation of the causative drug and starting immunosuppressive therapy similar to nondrug-induced ANCA-associated GN.
Massachusetts-Clinical Vignette-Poster Finalist
Nura El-Haj, MD

Title: A Tale of Two Rare Entities: Cutaneous Gamma-Delta T-Cell Lymphoma Presenting as Hemophagocytic Lymphohistiocytosis

Authors: Nura El-Haj*, Shrinkhala Khanna§, Jason Philippi*, Jessica Yuen*, Syed Ali§, * Department of Internal Medicine and Pediatrics, Baystate Medical Center, Springfield, MA, § Department of Hematology and Oncology, Baystate Medical Center, Springfield, MA

Introduction: Subcutaneous panniculitis T cell lymphoma (SPTCL) is a rare form of skin lymphoma first described in 1991, and recognized as a distinct entity in 2001 by the World Health Organization. Two phenotypic variations based on T cell receptor (TCR) gene rearrangements exist: TCRαβ and TCRγδ. These entities have histological and immunophenotypic differences which impact management and prognosis and therefore have been redefined. SPTCL refers to cases expressing TCRαβ, while those with TCRγδ are described as ‘cutaneous gamma-delta T cell lymphoma’ (CGD-TCL). CGD-TCL presents with epidermal, dermal or subcutaneous involvement with median survival reported between 15-31 months.

Case Presentation: A 42-year-old female presented with worsening ulcerating skin lesions, three days after relocating from Puerto Rico where she was treated for cellulitis. She was afebrile, had scleral icterus, ulcerating lesions and nodules on her lower extremities with pitting edema. Workup revealed pancytopenia (WBC 3.9k/mm³, Hb 8.9 g/dl, Platelet count 107k/mm³), coagulopathy (INR 1.5, fibrinogen 62 mg/dl, D-dimer 2.89 mg/l FEU) in the context of liver dysfunction (alkaline phosphatase 721 U/L, AST 529 u/l, ALT 135 u/l, total bilirubin 4.2 mg/dl, albumin 1.8 g/dl). She had elevated fasting triglyceride (328mg/dl) and ferritin levels (6,494 ng/ml). Abdominal CT showed steatosis, hepatomegaly, without splenomegaly. Hemophagocytic lymphohistiocytosis (HLH) was suspected and dexamethasone was initiated on day 2 of hospitalization. Hemophagocytes were seen on bone marrow aspirate and dose reduced etoposide was started on day 5. Soluble IL-2 receptor levels were elevated at 7910 pg/ml, fulfilling 5 of 8 diagnostic criteria for HLH. Skin biopsy exhibited nonspecific panniculitis and was repeated to sample subcutaneous tissue, revealing atypical lymphohistiocytic infiltrates and hemophagocytosis. Based on clinical context and in consultation with Massachusetts General Hospital, a diagnosis of CGD-TCL was formulated given the presence of cytologically atypical and clonal T-cells with a uniform phenotype of γδ+, CD56+, granzyme+, and perforin+. The patient started outpatient therapy with CHOEP-cyclophosphamide, doxorubicin, etoposide, vincristine and dexamethasone with plans for allogeneic transplant.

Discussion: Our case presents a combination of two rare entities, HLH and CGD-TCL, raising diagnostic challenges, complicating therapy and prognosis. Diagnosis of CGD-TCL should be guided by clinical, pathological and molecular data, noting that skin lesions may mimic other dermatological and infectious processes. A deep biopsy including the subcutaneous tissue and dermis is essential for an accurate diagnosis. In pediatric patients, HLH is often of primary nature and involves aggressive upfront therapy and allogeneic transplant in high-risk patients. In adults, it is essential to treat the underlying trigger for HLH. We present a rare diagnosis with sparse supporting literature based on anecdotal observational data regarding treatment modalities. Although this patient’s clinical course remains to be determined, it may add to current reports describing clinical behavior and response to therapy including some positive outcomes with allogeneic transplantation in limited patient subsets.
References


Title: Pseudoporphyria: Beyond a simple drug allergy rash

Authors: Omar Hadzipasic, Internal Medicine Resident, Department of Medicine, University of Massachusetts, Worcester MA., Li Wang, Internal Medicine Resident, Department of Medicine, University of Massachusetts, Worcester MA., Shahrzad Shidfar, Assistant Professor, Department of Medicine, University of Massachusetts, Worcester MA.

Introduction: Drug-induced Pseudoporphyria presents with bullae of sun-exposed skin after exposure to an inciting agent. It is an uncommon condition that can be mistaken for similar blistering disorders, and thus its diagnosis must not be missed to avoid persistence and complications of the rash.

Case Presentation: A 50-year-old woman with a history of chronic pancreatitis presented with urinary frequency and was prescribed Bactrim for a urinary tract infection. Shortly after, she developed an intensely pruritic macular erythematous rash on her hands, feet, and trunk with the simultaneous appearance of tense clear fluid-filled blisters on the dorsal surfaces of her hands and feet with lower extremity edema. She was started on a prednisone taper for presumed drug rash due to Bactrim and given Lasix for lower extremity edema. The patient had worsening of the rash with development of shallow punched out ulcers with fibrinous exudate, as well as dried eschars over previous blister sites. White blood cell count of 13000 white blood cells per microliter without eosinophilia, normal ESR, negative Anti-nuclear antibody and hepatitis panel. Lasix was discontinued due to concern for sulfa cross-sensitivity. Pathology and direct immunofluorescence study of a new blister biopsy were consistent with pseudoporphyria or porphyria cutanea tarda (PCT). Uroporphyrin was mildly elevated with normal total serum porphyrins which made PCT unlikely. Further review of her home medications revealed that she was taking Naproxen for pain. Naproxen had been reported in multiple case reports as a trigger of pseudoporphyria. Naproxen was discontinued and ulcerations were treated with local wound care.

Discussion: Pseudoporphyria is an uncommon medical disorder that must be considered in the differential diagnosis for new vesicular rash after starting a drug. The cutaneous lesions can take months to years to resolve even after the offending agent has been removed, making initial diagnosis and discontinuation of the inciting agents paramount. The most common offending agents in drug-induced pseudoporphyria include NSAIDs, various antibiotics, and diuretics, including medications such as Naproxen, Furosemide, and Bactrim, all three of which our patient was taking at various times during her acute flares. This case highlights the importance of obtaining an accurate home medication list, including over the counter medications, and its essential role in prompt diagnosis of the culprit condition. Especially in our case, discontinuation of medications was crucial to avoid persistence of rash, additional workup, and possible misguided treatment with steroids.
Title: The Lyme Deception

Authors: Zeba Hashmath MD\(^1\) (Associate), Ramses Thabet MD\(^2\) (Associate), Mark Kranis DO\(^2\),
\(^1\) Department of Medicine, Saint Vincent Hospital, Worcester, MA, \(^2\) Division of Cardiology, Saint Vincent Hospital, Worcester, MA

Introduction: Sarcoidosis is a heterogeneous, non-caseating, granulomatous disorder of unknown etiology that can involve any organ. Cardiac involvement may be detected alone and may precede, follow, or occur concurrently with other organ (e.g., lung) involvement. In the United States, 13 to 50 percent of all sarcoid-related deaths are related to cardiac involvement.

Case Presentation: A 39-year-old Caucasian female with a remote history of unspecified tachycardia presented with chest discomfort and dizziness. She owns and cares for horses after her regular work hours as a nurse.

She was doing well until a week before presentation, when she felt a pressure in her chest, aggravated with exertion. In the ED, she was bradycardic to 49 bpm with otherwise stable vital signs. The initial EKG revealed a new LBBB. Cardiac biomarkers were negative. She was given aspirin and admitted to the medical floor for further evaluation.

During nuclear testing, she was found to be in 2:1 Mobitz type 2 heart block, and the test was aborted. Later that day, she was found to have AV dissociation with complete heart block on telemetry and was transferred to the CCU.

Despite the absence of a rash, there was a strong suspicion for Lyme carditis. Recently one of her horses had been diagnosed with Lyme disease. Pending serology and PCR, she was started on ceftriaxone, and a temporary pacemaker was placed. An echocardiogram showed a normal LVEF with no wall motion abnormalities.

After three days there was no improvement. The underlying rhythm remained complete heart block, despite pacing. The eagerly awaited Lyme serology and PCR came back negative. Hence, a permanent pacemaker was implanted. ACE level and chest x ray were within normal limits. She was discharged home on a steroid taper and a planned outpatient cardiac MRI.

The cardiac MRI revealed normal heart structure and function. However, enlarged paratracheal, precarinal, subcarinal and bilateral hilar lymph nodes were detected. There was also a suggestion of lung nodules. Given these findings, the etiology of her presentation and course was likely sarcoidosis with cardiac involvement.

Discussion: Sarcoidosis is a non-caseating granulomatous disease of unknown etiology, often involving the lungs and skin. However, only 5% of patients in the US present with cardiac symptoms. Our case emphasizes the need for having a high index of suspicion for sarcoidosis in patients presenting with varying degrees of heart block.
Massachusetts-Clinical Vignette-Poster Finalist
Najia Idrees, MBBS MD

Title: A Case of Aorto-jejunal Fistula: A Diagnostic Challenge.

Authors: Najia Idrees, MD, Mohammad Almeqdadi, MD, Manish Tandon, MD

Introduction: Aorto-enteric fistula (AEF) is a life-threatening cause of gastrointestinal bleeding (GIB). It is classified as primary, where it occurs de novo, and secondary, where it arises in the setting of aortic reconstruction. It most commonly affects the duodenum; however, it rarely involves the jejunum which can be a challenging diagnosis. We present a case of aorto-jejunal fistula, with presentation of recurrent ‘Herald bleeds’, which was not visualized on multiple imaging studies.

Case Presentation: The patient is a 55 year-old man with a past medical history of coronary artery disease, hypertension, and abdominal aortic aneurysm repair 10 years prior to presentation who had hematochezia and abdominal pain. This was the fourth recurrence of his symptoms within a month, requiring multiple hospital admissions. Prior colonoscopies revealed sigmoid diverticulosis and internal hemorrhoids presumed as the source of bleeding. Upon presenting, he was hemodynamically stable and his hemoglobin was 8.4 mg/dL. His hospital course was complicated by hypotension and hematochezia, after which upper and lower endoscopies failed to reveal the source of bleeding. However, old blood in the entire colon was visualized including the right side. A video capsule study and a tagged red blood cell scintigraphy also did not identify location of the GIB. A computed tomography angiography (CTA) was done which showed no extravasation of contrast or other signs suggestive of AEF and a 3.4 x 3.6 cm aneurysm at the distal end of the surgical aortic graft in proximity to the bowel. This was compared to an earlier CTA without significant change. However, due to the high suspicion of an aorto-enteric fistula, an exploratory laparotomy was performed, and a fistula was found connecting the aortic graft with the jejunum, and subsequently, an aortic graft resection and small bowel resection was performed with cryopreserved aortic anastomosis and jejunojejunal anastomosis. He was discharged on post-operative day five in stable condition.

Discussion: AEF is a life threatening cause of GIB necessitating early diagnosis and appropriate surgical management for survival. CTA is considered the modality of choice for diagnosis, while upper endoscopy (EGD) detects a small percent of cases. The diagnosis of AEF in this case was particularly challenging as over 75% of AEFs are located in the duodenum, rather than jejunum. Negative imaging studies during recurrent admissions further added to the diagnostic dilemma. Final diagnosis was made during surgical intervention. This case highlights the importance of high index suspicion for AEF in GIB even with negative imaging studies in patients with prior aortic reconstruction. It shows that superior non-invasive modalities for accurate diagnosis of AEF are still needed and highlights the variable sensitivity of CTA in establishing the diagnosis.
Title: Bronchus Associated Lymph Tissue (BALT) lymphoma masquerading as pneumonia.

Authors: Mridula Ann Jacob, MD, MPH; Twinkle Chandak, MD, FCCP

Introduction: BALT lymphoma is a very rare but distinct subgroup of low-grade B-cell extranodal non-Hodgkin’s lymphoma, classified as marginal-zone lymphoma. Here we present a case of a woman who presented with persistent multifocal infiltrates that did not resolve with antibiotic therapy and lung biopsy confirmed BALT lymphoma.

Case Presentation: The patient is a 51 year old female, current smoker (15 pack years) with history of COPD and recurrent bilateral lung infiltrates who presented with a three day history of malaise, fatigue and chills along with shortness of breath. She was treated for left lower lobe and right middle lobe community acquired pneumonia in June 2014. Her examination was notable for diminished breath sounds over the base of the left lung. Laboratory studies were within normal limits. CT scan of the chest showed interval worsening of left pulmonary consolidation, suggesting worsening pneumonia, and she was treated with antibiotics. Repeat CT chest in eight weeks showed improved aeration of the left lung consolidation along the major fissure; with remaining infiltrates appearing similar to the prior study. Further laboratory studies, including c- ANCA and p-ANCA which were negative. Two years later, due to the persistent bilateral infiltrates, a bronchoscopy with transbronchial biopsy was done. The pathology report showed alveolar tissue that contains nodular collections of monotonous small lymphocytes that obliterate the normal alveolar architecture and seem to be centered around small pulmonary arteries. The combination of the monotony of the lymphocytes and their location associated with blood vessels (lymphangitic pattern) is suggestive of low grade marginal zone lymphoma (BALT lymphoma). B cell gene rearrangement study was positive. PET scan showed intense hypermetabolic chronic wedge-shaped airspace infiltrates and ground-glass opacities in bilateral lungs.

Discussion: A recent literature review found only four cases of BALT lymphoma (0.17%) from 2250 lung cancer specimens collected at a department of pathology from 2010 to 2014. BALT is found in the sub-mucosal membrane that contains both T and B lymphocytes. BALT is absent from the lung in physiological circumstances; known triggers for the development of BALT lymphoma include smoking induced chronic antigenic stimulation, autoimmune diseases such as Sjögren’s syndrome, and chronic infection. BALT lymphoma should be suspected in patients with chronic alveolar opacity, usually with an air bronchogram and no pleural effusions or mediastinal adenopathy. Nearly half of the patients are asymptomatic and extra-pulmonary manifestations occur in less than one-quarter. Clinical manifestations and radiological characteristics are non-specific, but prognosis is favorable2. Although there is no standard chemotherapy regimen currently in use, case reports exist about the efficacy of using rituximab and high dose macrolides.
Title: Salicylate Toxicity from Chronic Over-the-Counter Ointments and Medications

Authors: Wei Sum Li, MD Brown University Internal Medicine

Introduction: Salicylates are an extremely common active ingredient of both prescription and over-the-counter products. While patients often consider over-the-counter products benign, salicylate toxicity remains a common overdose with high morbidity and mortality. This case illustrates the risk of unintentional toxicity with concurrent use of salicylate containing products complicated by an unassuming sunburn.

Case Presentation: A 52-year-old male with a past medical history of hypertension and hyperlipidemia presented to the emergency department with acutely altered mental status. The patient was in his usual state of health after an unremarkable weekend of yard work. The following morning, the patient developed tinnitus, dizziness, and shortness of breath. By evening, he had nausea, vomiting, and somnolence. Exam was notable for tachycardia, tachypnea, diaphoresis, and a moderate sunburn on his neck and shoulders. Initial labs were notable for serum potassium of 3.5 mEq/L, serum bicarbonate of 13 mEq/L, and an anion gap of 15. Creatinine was 1.20 mg/dL. Venous blood gas showed a pH of 7.49 with pCO2 of 19. His initial salicylate level was 80.3 mg/dL (therapeutic level between 10-30mg/dL).

The patient’s mental status and salicylate level cleared remarkably rapidly with prompt treatment of sodium bicarbonate infusion, potassium repletion to support urine alkalization, and general supportive care. As his mental status improved, further history was elicited. The patient did take a daily 81mg aspirin. He adamantly refused suicidality. He was a former football player and had been using over-the-counter ointments for chronic joint and muscle pain for >30 years. He used three different muscle rubs each of which included between 0.61% and 29% methyl salicylate. The patient reported as needed use of these topicals several times weekly on his back, shoulders, knees, and hips. Sodium bicarbonate infusion was stopped when the salicylate level remained persistently <30mg/dL and hemodialysis was deferred. Patient did not require advanced airway management and was discharged home on hospital day five.

Discussion: Daily oral aspirin and topical methyl salicylate and are commonly seen outpatient regimens, but patients may not initially provide history of use of these over-the-counter medications. Notably, five mL of wintergreen oil is equivalent to approximately seven grams of salicylic acid, or 21.5 tablets of 325 mg aspirin. In particular, areas of compromised epidermis are associated with increased risk of toxicity. While signs of suicidality and oral aspirin overdose should be considered in cases of salicylate toxicity, this case illustrates the need for careful history taking to evaluate for other sources of exposure. Physicians and patients should be aware that chronic, frequent exposure to methyl salicylate and application over any area of dermal breakdown, such as sunburn, will increase overall absorption and likelihood of elevated serum concentrations. Particular care should taken with chronic or concurrent use with various formulations of salicylates.
Introduction: A prolonged QT on ECG usually brings to mind congenital causes or medication toxicities. But can love prolong the QT and lead to Torsade? We present the case of a couple: a man with symptomatic bradycardia and prolonged QT, whose girlfriend just weeks earlier received a permanent intracardiac defibrillator (ICD).

Case Presentation: A 31 year old male with no known significant past medical history was transported by the EMS to the hospital due to two episodes of seizures with transient loss of consciousness witnessed by his girlfriend.

He reported consuming lots of energy drinks and playing videogames continuously for 36 hours, but no medication abuse. He had symptomatic bradycardia (35 bpm). An ECG revealed junctional rhythm, abnormal T waves and a prolonged QT segment with a Brugada pattern (QTc: 552). CBC, BMP, troponin T, CPK MB, alcohol, salicylate, and urine toxicology screening were all negative. Atropine was given and before the temporary cardiac pacer wire was implanted, he developed seizures with associated ventricular tachycardia that degenerated into Torsades de Pointes, which self-terminated. It was thought that his seizures were due to Torsades de pointes. A cardiac catheterization and ICD placement were planned.

Before the procedure the patient conceded he was abusing loperamide. He had started opioid consumption 4 years earlier, and he wanted to quit a year prior, but started having withdrawal symptoms. He then started himself on loperamide at a remarkable total dose of 280 mg/day, achieving similar opioid effects and overcoming the withdrawal. With observation the QTc ultimately normalized. A psychiatry consultant advised starting suboxone for withdrawal and follow up with an outpatient suboxone program.

Interestingly, his girlfriend had received an ICD at a different hospital due to the same circumstances two weeks earlier, having never divulged her loperamide abuse.

Discussion: Loperamide, known as the “poor man’s methadone,” can offer opioid effects when consumed in large quantities. Its common side effects are mild compared to its deadly arrhythmic potential in toxic doses. Since the drug was approved in 1976, 48 cases of serious cardiac problems have been reported.

Is prolonged QT transmitted between couples? Definitely not infectiously, but it is a social epidemiologic problem, as seen in this young couple. Expensive and invasive procedures could likely be avoided if loperamide abuse were reported to address a prolonged QT. We would urge the FDA to monitor loperamide consumption by designating it a prescription drug in order to minimize a societal danger, particularly in the light of opiates now becoming much more restricted.
References

Title: Congenital Lipodystrophy: Can Fat Harm the Lungs?

Authors: Charles Ma, MD; Hassan Aftab, MD; Myat Soe, MD

Introduction: Lipodystrophy is associated with abnormal distribution of lipids in the body. It is a clinical diagnosis based on physical exam, associated comorbitidies, lipid profiles and genetic testing. There are two categories: congenital and acquired with the former being a much more rare presentation. As such there is a lack of guidelines in terms of the management of such patients. We present a case of a patient with suspected congenital lipodystrophy who was admitted for symptomatic hypertriglyceridemia and developed ARDS after treatment with insulin drip.

Case Presentation: AC has a hx of clinically diagnosed congenital lipodystrophy of unknown type, as well as major complications from her chronic disease including uncontrolled type 2 Diabetes mellitus, diabetic nephropathy, neuropathy, hepatosteatosis, significant vascular atherosclerosis and hypertriglyceridemia. She presented initially to the ED for significant severe epigastric pain with radiation to the back. In the ED patient was vitally stable but had significant tenderness in the epigastric area. Labs reveal a triglyceride level of 3,184 mg/dL (baseline 300mg/dL), lipase of 16u/L and amylase of 29u/L. LFTs were WNL. A RUQ ultrasound and CT abdomen showed no pancreatic anormalities. Patient was admitted to the medical wards.

On second day of admission, lipid profile study interpreted as evidence of Type V hyperlipoproteinemia. Was started on insulin drip with triglycerides falling to 1500mg/dL. On the third day, patient developed acute onset respiratory distress and chest x-ray consistent with ARDS. She was intubated and transfered to the MICU. ICU course was uncomplicated and patient continued on insulin drip with triglyerides at around 800mg/dL before transitioning to subcutaneous insulin injections. She was extubated successfully on the 5th day and discharged a few days later.

Discussion: We report a case of ARDS in a patient with history of congenital lipodystrophy after she presented with symptomatic hypertriglyceridemia treated with Insulin drip. The mechanism of this finding is not well established, however, we believe it is similar to the development of ARDS in patients with acute pancreatitis.

One of the pathogenic players in respiratory insufficiency in a long list include FFA. The free fatty acids released from triglycerides have been shown to damage capillary alveolar membrane. In this case, it was evident that the patient did not develop pancreatitis based on lipase levels and abdominal imaging. Previous literature states that the treatment of hypertriglyceridemia with insulin drip is the treatment of choice to rapidly reduce the amount of triglycerides in the blood. We speculate she likely developed ARDS from FFA mediated pulmonary endothelial damage and surfactant disruption with rapid treatment of insulin drip stimulating endothelial lipoprotein lipase conversion of triglycerides into FFA.

Future consideration in such patient may indicate a slower treatment of hypertriglyceridemia with or without continuous insulin infusion. If patient has evidence of instability or impending development of ARDS, plasmapharesis should be considered sooner to avoid further complications.
Title: "Chaos in an attempt to appease" - A case of Mycophenolate Mofetil Induced Supraventricular Tachycardia.

Authors: Nkechi Mbaebie, Sylvia Alarcon, Peter Shaw.

Introduction: Mycophenolate mofetil (MMF or Cell Cept) is a commonly used drug in the prevention of allograft rejection in patients with solid organ transplant and has continued to gain popularity in the management of recalcitrant conditions of dire autoimmune disorders especially in the setting of lack of response to first line agents.

As with many immunosuppressive therapies, it has numerous side effects but compared to other immunosuppressive agents, it still has a lower toxicity profile making it an attractive alternative when patients with severe autoimmune disorders fail to respond to first line agents.

Side effects to MMF include nausea and vomiting, afebrile diarrhea, diffuse colitis, insomnia, anxiety, easy bleeding and mental/ mood disturbances. Although fast and irregular heartbeat was identified as a serious but very rare side effect of MMF, its incidence remains very low and rarely reported.

Case Presentation: 50 year old female with history of scleroderma and mixed connective tissue disorder and multiple cardiac co-morbidities who presented with chest pain that was sharp, non-radiating with associated nausea, found to have tachycardia on examination with regular heart rhythm.

EKG revealed new onset Supraventricular tachycardia (SVT), and ST depression in V3-V6 that was present in previous Electrocardiograms. Troponin x 3 followed every 4 hourly was <0.015

She recently started treatment with MMF for severe scleroderma in the setting of non-response to first line agents. The rare side effect of MMF in causing fast and irregular heart beats was considered and MMF was discontinued with complete resolution of her symptoms. She continued to remain in sinus rhythm even when followed up 6 months later as outpatient.

Discussion: Mycophenolate mofetil has been a stable in the prevention of allograft rejection but in addition, it is used in the treatment of autoimmune conditions such as psoriasis, rheumatoid arthritis, scleroderma and autoimmune uveoretinitis especially in the setting of non-response to first line agents.

Clinically, MMF related cardiac insult may present as any form of arrhythmia, this untoward effect is very rare and only very few cases has been reported. Our patient presented with SVT post MMF, which appears to be new in this patient as patient was adjudged to be stable from the cardiology standpoint prior to start of this medication.

This case further illustrates that a detailed evaluation including detailed medical history and consideration of the possible side effects of any new medication should be considered in any patient with new onset arrhythmia.

Early recognition of this entity along with discontinuation of this medication may result in improved outcome.
References

Massachusetts-Clinical Vignette-Poster Finalist
Ahmed Nagy, MD

Title: Congenital IVC atresia, a rare culprit associated with recurrent extensive thrombosis

Authors: 1- Ahmed Y. Nagy, MD. Internal Medicine Resident, University of Massachusetts Medical School., 2- Elizabeth Ryer, MD. Internal Medicine Resident, University of Massachusetts Medical School., 3- Hannah R. Rosenfield. Medical Student. University of Massachusetts Medical School., 4- Christine Martin. Medical Student. University of Massachusetts Medical School., 5- Meghna C. Trivedi, MD. Assistant Professor. University of Massachusetts Medical School.

Introduction: A 45 years old Vietnamese male was diagnosed with left leg deep vein thrombosis (DVT) about 2 years ago in Vietnam. He took herbal supplements without significant improvement. Soon after immigrating to the United States six months ago, he was urgently admitted to hospital for bilateral leg swelling and pain, and diagnosed with bilateral DVT. CT abdomen and pelvis revealed congenital atresia of inferior vena cava and of the bilateral common iliac veins, resulting in multiple venous collaterals in the abdomen, retroperitoneum and subcutaneous tissue.

Case Presentation: IVC atresia was believed to be strongly associated with deep vein thrombosis in our patient. He had been a lifelong nonsmoker and did not have family history of thrombotic disorders. Workup for hypercoagulable disorders including Factor V Leiden mutation, Prothrombin gene mutation, Anticardiolipin antibody panel, and antithrombin 3 activity were all unremarkable. Subcutaneous weight based enoxaparin and warfarin overlap for anticoagulation was initiated with plan for lifelong therapy. As his work schedule precluded him to obtain regular INR checks, he was recently switched to novel anticoagulant Rivaroxaban. Unfortunately, he failed treatment with Rivaroxaban and presented back to our hospital with worsening left lower extremity pain and swelling. On exam, left leg was swollen, erythematous and had superficial ulcerations. Doppler ultrasound revealed extensive thrombosis in left common femoral vein, proximal femoral vein, proximal profundal vein, and popliteal vein. Acute superficial thrombophlebitis was found in the left great saphenous vein at the proximal calf and in a branch at the left proximal calf. He was evaluated by vascular surgery and hematology teams. No surgical intervention was warranted. Due to failure of Rivaroxaban, he was restarted on an overlap regimen of enoxaparin and warfarin. As leg swelling and pain improved, he was discharged home in good condition.

Discussion: Inferior Vena Cava atresia is a rare vascular anomaly with an estimated prevalence of 1% in general population. At 4-6 weeks of gestation, regression and fusion of three sets of paired veins-posterior cardinal, subcardinal, and supracardinal veins forms IVC. Failure of these paired veins to fuse Results: in IVC anomaly. Patients with IVC atresia develop robust collateral deep venous system with or without azygous and hemizygous continuation in the IVC. During physical exertion these collaterals inadequately drain the lower limbs, thus causing venous stasis, superficial ulcerations, and deep vein thrombosis (DVT). IVC atresia should be considered as a differential diagnosis in young patients with unexplained DVTs, recurrent lower extremity ulcers, and enlarged abdominal veins. An estimated 5% of cases of DVT in patients under 30 years of age are caused by congenital IVC anomalies. CT angiography is the best study to diagnose IVC anomalies. Due to irreversible risk of thrombosis, lifelong anticoagulation may be necessary.
Title: A Spectrum within a Spectrum: The Diagnostic Conundrum of Neuropsychiatric Systemic Lupus Erythematosus (NPSLE)

Authors: Anthony Nicolas, MD, Christopher LaChance MD; Farzan Irani, MD; Venkatrao Medarametla, MD

Introduction: Due to massive disease heterogeneity, the diagnosis and treatment of NPSLE remain a clinical challenge. A high index of suspicion, even in the setting of normal imaging, is required for diagnosis combined with early recognition of clinical/laboratory diagnostic criteria of SLE in patients without an established diagnosis of lupus.

Case Presentation: A 63-year-old woman with a medical history of bipolar disorder, depression, SIADH, celiac disease and two prior venous thromboembolic events despite anticoagulation was admitted for altered mental status and generalized weakness. Prior to this illness, she had been fully ambulatory and independent. Over the course of approximately 6-8 months, her family noted several behavioral changes as well as a progressive functional decline. She had become forgetful, confused, withdrawn and required walker for the first time in her life.

Review of systems was noteworthy for generalized alopecia and chronic leukopenia of less than 4.0k/mm$^3$ for which she was referred, by her PCP, to an outpatient hematologist and had begun workup.

Physical exam revealed a thin woman with a flat affect, alopecia and rigid extremities. She was alert and oriented; her speech slow and quiet. Pertinent admission labs demonstrated moderate hypo-osmolar hyponatremia of 125mmol/L, white count 3.0k/mm$^3$ and 3+ albuminuria. Labs from her outpatient hematologist’s office included ANA titer of 1:2560 (homogeneous), high ESR/CRP and low C3.

Throughout the patient’s hospital course, her clinical status rapidly deteriorated. She became confused, nonverbal and developed widespread dystonia with fine tremors, intermittent tachycardia, and tachypnea. An extensive workup ensued which included an unremarkable brain MRI, an abnormal EEG suggestive of diffuse toxic/metabolic encephalopathy, negative infectious workup and CSF studies indicating high levels of IgG. Neurology, infectious disease, nephrology, and psychiatry consult services were all involved.

Primary team and the nephrology service suspected an underlying diagnosis of Lupus and ordered a renal biopsy to determine the etiology of her nephrotic range proteinuria (TP/Cr 3.7g/d). Autoimmune serologies returned positive for significantly elevated levels of anti-dsDNA (41) and Rheumatology input was obtained. Renal biopsy was consistent with lupus glomerulonephritis. The patient was diagnosed with lupus cerebritis and started on Cytoxan therapy following a 1-week pulse of high dose IV methylprednisolone. Her neurological status drastically improved and she eventually returned to her baseline.

Discussion: Pathophysiology of NPSLE is multifaceted and clinically presents as varying constellations of central, peripheral, autonomic and psychiatric manifestations that are often underrecognized.$^1$$^2$

Several serological tests have been described in the literature to facilitate the diagnosis of NPSLE.$^3$$^5$. The anti-ribosomal P antibody is one notable example but has limited diagnostic utility as it does not
differentiate between various disease phenotypes and has a sensitivity and specificity of 23% and 80%, respectively. Radiologically, MRI remains the most sensitive tool in supporting the diagnosis of NPSLE, yet findings may be absent or nonspecific.

References

Title: Sarcoidosis and IgA nephropathy - An unusual relationship or mere co-incidence?

Authors: Eziafa Oduah MD MPH MS¹; Lauren Dudley MD¹; Hani Erian MD, ¹Berkshire Medical Center

Introduction: Sarcoidosis associated with IgA nephropathy is rare. A clear relationship or causality remains controversial. Renal involvement is seen in 10% to 20% of sarcoidosis mostly in the form of nephrocalcinosis and nephrolithiasis. Sarcoidosis associated with IgA nephropathy has been reported in only a few case reports to date. We hereby describe another presentation of sarcoidosis in association with IgA nephropathy.

Case Presentation: A 20 year old female was seen for cervical lymphadenopathy of about 3 months duration. Her symptom was preceded by an episode of sore throat and fever treated empirically with antibiotics for suspected streptococcal pharyngitis and mononucleosis. She later developed a rash which started in her lower extremities and extended up her trunk, arthralgias, abdominal pain, night sweats, chills, fever, and epistaxis. Physical exam revealed tender, mobile 4-5cm anterior cervical lymph nodes, blanching maculopapular rash predominantly on the lower extremities, abdomen, back and arms. Extensive work up was initiated. Labs revealed IgA of 430 (normal <340) and complement 3 levels of 199 (reference 106 – 199), positive Anti-Streptolysin O antibody, ESR 34, CRP 76.9, ACE 24. Other labs including toxicology screen, ANA, RF, antiCCP, p-ANCA, c-ANCA, cryoglobulin, hepatitis, CMV, EBV, HIV, syphilis antibody, toxoplasma, infectious mononucleosis, gonorrhea, and chlamydia were negative. Urinalysis showed 2+ proteinuria. CXR was normal. Skin biopsy was negative for vasculitis. Ultrasound of the cervical lymph nodes suggested reactive lymphadenopathy. A CT scan of the neck with IV contrast also suggested a reactive lymphadenopathy. Nephrology was consulted for proteinuria and a renal biopsy was performed. Results: showed segmental increase in mesangial matrix and cellularity. Immunofluorescence microscopy demonstrated granular mesangial immunoglobulin A (IgA) deposits consistent with IgA nephropathy. There were no subepithelial deposits or characteristic IgG and C3 deposits on IF to suggest poststreptococcal GN. Excisional cervical lymph node biopsy showed multiple non-caseating granulomas suspicious for sarcoidosis. She was started on steroid therapy with improvement. She continues to be monitored clinically on an outpatient basis.

Discussion: The patient described here presented with an unusual constellation of symptoms having failed antibiotic therapy for a presumed upper respiratory infection. Her persistent lymphadenopathy remained concerning following a biopsy proven diagnosis of IgA nephropathy which did not fully explain her symptom complex. Hence excisional lymph node biopsy was pursued confirming co-existing sarcoidosis. A causal relationship between these two conditions has been proposed based on the observation that both diseases are exacerbated following upper respiratory infections; as well as shared immunologic abnormalities such as elevated serum IgA and circulating immune complexes. Notably, our patient presented with a preceding upper respiratory illness and had modestly elevated IgA. While this case does not confirm causality, it adds to the body of evidence suggesting a relationship between IgA nephropathy and Sarcoidosis.

References

Massachusetts-Clinical Vignette-Poster Finalist
Shazia Samanani, MD

Title: If Your Heart is a Mess, Think of DRESS

Authors: Shazia Samanani, MD. Osama Kandalaft, MD. Anya Filshtinsky, MD. Joshua Schilling, MD. Jean Henneberry, MD.

Introduction: Many systemic illnesses often present with non-specific symptoms and can often mimic other conditions. These conditions require a high clinical suspicion and pattern recognition to aid in diagnosis. Here, we present a case of DRESS syndrome and understand the common features to aid in diagnosis.

Case Presentation: A 57-year-old male developed sudden left-sided crushing chest pain. He had recently traveled to Missouri two weeks prior where he spent time outdoors. A few days after returning, a rash developed on his hips and his local ED prescribed Bactrim and Keflex for presumed cellulitis. Despite antibiotics, his rash progressed and he developed a fever, malaise, and headache. On examination, was temperature was 101°F, and he had a diffuse maculopapular rash. His EKG showed T-wave inversions in the inferior and lateral leads and labs revealed a troponin-I of 10.10. Echocardiogram was normal.

With his triad of fever, headache, and rash in conjunction with his cardiac findings, Rocky Mountain Spotted Fever myocarditis was suspected and he was started on doxycycline. Despite this, the patient’s symptoms worsened with a rising leukocytosis (34k/mm3) with eosinophilia. Skin biopsy was performed showing perivascular dermatitis with eosinophils, findings considered consistent with Drug Rash with Eosinophilia and Systemic Symptoms (DRESS syndrome). His antibiotics were stopped and prednisone was started with rapid improvement in his symptoms.

Discussion: DRESS syndrome is a potentially life threatening condition commonly triggered by an offending drug, usually an anticonvulsant or sulfonamide. It presents with fever, facial edema, cervical lymphadenopathy and rash. Its nonspecific symptoms make diagnosis difficult and it has been referred to as “the great mimicker”. Eosinophilia is the hallmark laboratory abnormality in >50% of cases. Mortality of DRESS syndrome is estimated to be as high as 10%, likely due to delay in diagnosis leading to systemic complications.

Myocarditis from DRESS syndrome remains under recognized and can be fatal if untreated. The gold standard diagnosis remains an endomyocardial biopsy. As this is rarely performed, the diagnosis remains mostly clinical. EKG may show ST elevations or inverted T-waves and echocardiogram may show a decreased ejection fraction. Cardiac enzymes are usually elevated.

The pathophysiology of DRESS syndrome is postulated to be a drug specific immune reaction that triggers T-cell activation leading to cytotoxic activity and hyper-eosinophilia. In the heart, involvement usually begins as a hypersensitivity myocarditis and progresses into three different stages; an acute necrotic stage, a thrombotic stage and a fibrotic stage. Biventricular failure and cardiogenic shock are potential complications.

Treatment of DRESS syndrome focuses on discontinuing the provoking agent, which in our patient was suspected as Bactrim. Early drug withdrawal has been associated with improved outcomes. A high dose of systemic corticosteroids is also recommended with a slow taper over months to prevent relapse.
References


Title: Stroke vs Contrast Induced Encephalopathy - Compare and Contrast

Authors: William Wyman, DO., Myat Soe, MD

Introduction: Contrast Induced Encephalopathy (CIE) is a rare complication of IV contrast exposure with an estimated incidence of 0.3 -1.0%. The clinical presentation of CIE is similar to ischemic events and can easily be confused for an embolic stroke. A high degree of suspicion for CIE is required to make the diagnosis based on clinical presentation and imaging findings.

Case Presentation: 42 year old female with history of left middle cerebral artery aneurysm rupture s/p coiling 1 year ago without baseline neurological deficit presented with acute onset of altered mental status after follow up computed tomography (CT) angiogram. The patient was noted to be lethargic and unable to follow commands for 2 days then admitted for evaluation. On exam, the patient was noted to be disoriented, agitated with expressive aphasia, hyperesthesia, photophobia and phonophobia. She was able to move all extremities spontaneously and was able to walk without assistance. A non-contrast head CT and CT angiogram of head and neck were negative for acute lesions. Brain magnetic resonance imaging (MRI) was significant for punctate lesions in both cerebral hemispheres, left thalamus and left lentiform nucleus. The lesions were described as hyperintense on diffusion weighted imaging (DWI) series, but no abnormality was noted in apparent diffusion coefficient (ADC).

As patient’s clinical presentation seemed inconsistent with embolic stroke based on clinical history and imaging findings, alternative diagnoses were investigated. Literature review revealed CIE after intravenous contrast (IV) exposure, which can be mistaken as embolic stroke. Given her clinical scenario and MRI findings, we felt that CIE was the most likely diagnosis and initiated treatment with pulse dose steroids. The patient had rapid recovery after 3 days and was discharged home at baseline neurologic function.

Discussion: Several case reports describe CIE as acute mental status changes with or without focal neurologic deficits after exposure to IV contrast. It is postulated that osmotic disruption of the blood-brain barrier and direct neurotoxicity of extravasated contrast media result in cerebral edema and CIE. Although clinical presentation is variable, visual loss, aphasia, and hemiparesis are commonly reported. Imaging which helps differentiate CIE from ischemia is ADC, a quantitative measure of water diffusion. There is no abnormal ADC intensity in patients with CIE whereas in ischemic lesions, hyperintensities on DWI series are associated changes on ADC. CIE usually resolve spontaneously over three to seven days. Treatment with glucocorticoids and IV fluid hydration are reported to rapidly improve symptoms and reduce time to resolution.

CIE is a rare complication of IV contrast exposure which can be easily mistaken for embolic stroke after cardiac catheterization or cerebral angiography. The key to distinguish CIE from stroke is clinical history and symptom onset after contrast exposure with no abnormal ADC signals in MRI.
Title: My Mood is Unstable and my Heart is Slow

Authors: Rizwan Ahamed, M.D., Anupam Sule MD, Ph.D., FACP. Associate Program Director, Transitional Year. St Joseph Mercy Oakland Hospital., Ammar Huq MS3, Humam Huq MS3, Ross University, School of Medicine.

Introduction: Lithium is a mood stabilizer. It is known for its narrow therapeutic index, thus making patients more susceptible to toxicity and rarely causes cardiac side effects like sinus bradycardia with frequent pauses. Clinicians should remain cognizant of bradycardia induced by Lithium, which may be overlooked as a cause due to other confounding factors.

Case Presentation: A 65-year-old male with a history of Bipolar disorder presented to the ED with generalized weakness. Two weeks ago he was seen by his psychiatrist for generalized weakness and had elevated Lithium level at 1.8. Despite lowering his dose, his condition worsened that he was unable to get out of bed. In the ED, his bradycardia ranged from 30s-40s bpm with sinus pauses. Upon examination, he had altered mental status with fine tremors when extending his arms. Labs revealed elevated Lithium level of 2.1, hyperkalemia at 5.7, BUN of 67 and creatinine of 4.9. Thyroid function test was normal. He received a dose of atropine 0.5 mg for sinus pause in the ED. ECG showed junctional rhythm with a HR of 60 bpm, QTC of 452 milliseconds. Metoprolol 12.5mg, Lisinopril 5 mg, and Lithium were discontinued. He was admitted to the ICU to receive Dopamine drip and hyperkalemia was corrected. On day two, despite a normal potassium and cessation of Metoprolol whose half-life is approximately 4 hours, his bradycardia persisted as his Lithium remained elevated at 1.6. His recurrent sinus pauses continued and he received Atropine iv pushes. Cardiology didn’t recommend pacemaker as it was suggestive of reversible bradycardia due to Lithium toxicity. Next day, Lithium levels normalized to 0.9 with patient showing normal HR devoid of pauses. He was diagnosed with bradycardia secondary to Lithium toxicity.

Discussion: Lithium is eliminated through the kidneys. The half-life of Lithium increases as the renal function declines. Any increase in the risk of toxicity can precipitate its less common effects such as bradycardia. As a cation, it can disrupt the homeostasis of electrophysiology of the heart by interacting with calcium, potassium, and sodium. Lithium replaces intracellular calcium, and lowers intracellular potassium concentrations, interrupting the normal physiology of cAMP, Na/K and Na/Ca transporters, which decreases the depolarization rate. Physicians often tend to overlook Lithium toxicity as the primary cause of bradycardia especially when there are common confounders such as beta blockers and hyperkalemia are present. However, case report demonstrates the need for clinicians to be vigilant of Lithium toxicity induced bradycardia.
Title: Scrotal necrosis mimicking Fournier’s after HIPEC

Authors: Zohaib Ahmed, MD; Joel T. Fishbain, MD

Introduction: Hyperthermic intraperitoneal chemotherapy (HIPEC) represents an advancement in chemotherapy by applying heated chemotherapy directly into the abdomen after tumor debulking surgery. We describe the case of a rare complication associated with HIPEC: epidermal ulceration and necrosis of the scrotal wall.

Case Presentation: A 47-year-old man with metastatic appendiceal carcinoma presented to the hospital with a ten-day history of scrotal pain and swelling. Four months ago, the patient underwent tumor debulking and HIPEC. The patient denied fever and had a normal complete blood count. Scrotal ultrasound showed bilateral varicocele and hydrocele and the patient was discharged home on cephalexin. The patient returned two days later with increasing pain. Physical examination revealed serpiginous, irregular necrotic skin of the scrotum with no edema or erythema. A CT of the abdomen and pelvis showed scrotal wall edema and some fluid collection in the right lower abdominal wall. His white blood cell count was normal. Debridement of the necrotic tissue revealed epidermal ulceration with necrosis of underlying tissues and obliterator vasculitis with inflammatory features suggestive of Fournier’s gangrene. Special stains were not significant, and all cultures were ultimately negative.

Discussion: HIPEC therapy is a relatively new treatment modality with commonly reported surgical complications including bowel perforation, bleeding, fistula formation, bile leakage, pancreatitis, and sepsis. Genital ulceration has been reported but as an infrequent complication of HIPEC. Sequestration from the abdomen via the processus vaginalis is one proposed mechanism. Early recognition and appropriate management are essential in this rare complication.
**Michigan-Clinical Vignette-Poster Finalist**

**Mohammad Altujjar, MD**

Recurrent Intra-Cardiac Mass in a Patient with a History of Intra-Cardiac Lipoma Presenting with AICD Firing.

**Authors:** Mohammad Altujjar M.D., Saint John Hospital and Medical Center, Detroit, Michigan, Raymond Hilu M.D. FACP., Saint John Hospital and Medical Center, Detroit, Michigan, Mashkur Husain M.D., Saint John Hospital and Medical Center, Detroit, Michigan, David Rodriguez, M.D., FACC., Saint John Hospital and Medical Center, Detroit, Michigan

**Introduction:** The majority of primary cardiac tumors are asymptomatic, benign, and do not raise concern for a malignancy. These tumors can, however, carry a risk of life-threatening arrhythmias, embolic phenomena, cardiac tamponade, heart failure and valvular dysfunction.

**Case Presentation:** A 55 year old female with PMH of intra-cardiac lipoma with a history of debulking and AICD placement for secondary prevention of ventricular tachycardia in 2002, presented with a complaint of AICD firing. Upon presentation, her vitals were stable and she had an unremarkable initial work up. She was placed in the Clinical Decision Unit for observation. Overnight, the patient developed ventricular tachycardia with a rate of 200 and her AICD fired five times. She was loaded and continued on amiodarone drip as well as an IV beta blocker. A transesophageal echocardiogram was performed and revealed a seven by six centimeter mass occupying the right ventricular apex. She exhibited no further episodes of ventricular tachycardia and remained in normal sinus rhythm. She was later switched to oral amiodarone and beta blocker and was discharged home with plans to follow-up at the Mayo Clinic for further evaluation of probable recurrence of intra-cardiac lipoma.

**Discussion:** Cardiac tumors are usually benign in nature and asymptomatic. When symptomatic it might present with features of the classical triad of cardiac symptoms, systemic embolization and constitutional symptoms. Cardiac symptoms can vary form heart failure and valvular dysfunction, to life threatening arrhythmias. (1). Arrhythmia might occur as a consequence of neural pathway infiltration. The first manifestation could be sudden cardiac death. (2)

The diagnosis depends on high suspension index and is usually made with Echocardiography. Sometimes it is hard to distinguish the tumor, myxoma in particular, form cardiac thrombus. Other imaging modalities like MRI can be used. (3)

The management is surgical in general, however, it depends on the tumor histology. Myxoma, for example, requires surgical excision even if asymptomatic. lipoma requires resection only in case of hemodynamic compromise (4). Others prefer resection in timely fashion (2). Heart transplantation might be considered in some conditions (2). Multi-dispensary teams should be involved in its management. (2)

Even in benign cardiac tumors, patients should be monitored for recurrence and risk of cardiovascular complications. They should also be educated regarding symptoms that warrant further evaluation.

**References**


Michigan-Clinical Vignette-Poster Finalist
Brett Begley, MD

Title: Upfront triple combination therapy in pulmonary veno-occlusive disease

Authors: Brett Begley MD, Reda Girgis MD, Spectrum Health/Michigan State University Internal Medicine Residency, Fred and Lena Meijer Heart Center, Grand Rapids, Michigan

Introduction: Pulmonary veno-occlusive disease (PVOD) is a rare form of pulmonary hypertension due to preferential remodeling of pulmonary venules and is currently classified as a subtype of group 1 pulmonary arterial hypertension (PAH). Although the clinical presentation is similar to that of PAH, it carries a much worse prognosis with a high mortality rate. Importantly, targeted PAH therapy has not been shown to be effective and can precipitate pulmonary edema.

Case Presentation: Our case involves a 76-year-old female who presented with profound hypoxemia and signs of right sided heart failure. Transthoracic echocardiogram showed severe right ventricular dilatation and dysfunction with normal left heart. Right heart catheterization showed elevated pulmonary arterial (92/31 mm Hg) and right atrial pressures, normal pulmonary capillary wedge pressure, and depressed cardiac index. High resolution computed tomography (HRCT) scan showed enlarged mediastinal lymph nodes, interlobular septal thickening, and diffuse centrilobular ground glass opacities. Pulmonary function showed mild obstructive disease and severe reduction in carbon monoxide diffusion capacity. A clinical diagnosis of PVOD was made. Despite diuresis of approximately 10 L of fluid, she remained hypoxemic with an arterial blood glass demonstrating oxygen partial pressure of 62 mm Hg while on 100% FiO2 by high flow nasal cannula. She was then started on triple combination therapy with sildenafil, intravenous epoprostenol, and macitentan. Signs of right heart failure resolved with normalization of NT-BNP and oxygen requirements progressively decreased to 6 L/min by nasal cannula. NYHA class decreased from IV to III. She was discharged home and followed in the outpatient clinic with continued improvement in symptoms.

Discussion: We describe a rare case of severe PVOD with clear clinical response to targeted PAH therapy. This is the first report of the use of upfront triple combination therapy in PVOD.
Michigan-Clinical Vignette-Poster Finalist
Abhishek Bhandiwad, MBBS

Title: Kounis Syndrome’ – A case of Allergic Myocardial Infarction caused by Paclitaxel

Authors: Abhishek Bhandiwad, MD; Phoo Pwint Nandar, MD; Naveed Akthar, MD; Ernie P Balcueva, MD

Introduction: Paclitaxel is a commonly used anti-neoplastic agent included in a number of chemotherapy regimens. Acute myocardial infarction is a potentially fatal cardiotoxicity of Paclitaxel. We report a rare case of NSTEMI caused by Paclitaxel induced coronary vasospasm in a patient being treated for breast cancer.

Case Presentation: A 41-year-old female with no previous coronary artery disease and no family history of coronary artery disease, with ductal carcinoma right breast presented at the outpatient chemotherapy infusion center to receive the second dose of Paclitaxel.

Fifteen minutes after infusion started, she developed sudden sub-sternal chest pressure and dyspnea and was rushed to the emergency department. On arrival the patient was hypotensive, troponin was elevated at 0.07 and EKG showed new T wave inversions in lead II, III and aVF.

Six hours later, troponin was markedly elevated at 3.5; she was started on heparin infusion and immediately taken for cardiac catheterization that showed widely patent coronaries without stenosis.

Echocardiogram showed normal systolic and diastolic function without wall-motion abnormalities. In 24 hours, troponin trended down to 0.33, chest pressure and dyspnea resolved and she was discharged.

Discussion: Kounis syndrome i.e. allergic Myocardial Infarction is an under diagnosed entity caused by coronary spasm due to a vasoconstrictor stimulus. Allergic reactions to chemicals are encountered often with a variety of manifestations. Symptoms range from a minor rash to life threatening anaphylactic reactions. Rarely, such allergic reactions can precipitate acute organ involvement; in our case this was acute coronary syndrome.

Paclitaxel has been implicated to cause histamine mediated Coronary artery vasospasm due to its suspension vehicle named Cremophor -ethylene oxide combined with castor oil. Much caution should be exercised during treatment with Paclitaxel, especially at infusion centers where resources for cardiac resuscitation are limited. This case strengthens the postulation that allergic myocardial infarction by Paclitaxel can occur in persons with no known cardiac disease and can be potentially fatal.

References

Title: Peripheral Neuropathy and Alopecia: A Case of Surreptitious Thallium Toxicity

Authors: Emily Cordes, DO (Internal Medicine/Pediatrics, Western Michigan University School of Medicine), Lauren Lamie, DO (Internal Medicine, Western Michigan University School of Medicine), Thomas Melgar, MD (Internal Medicine/Pediatrics, Western Michigan University School of Medicine)

Introduction: Thallium is a heavy metal that is best known as a rodenticide occasionally involved in intentional or accidental poisonings. On average, Poison Control Centers of the United States report one case of thallium toxicity per year. Classically, thallium toxicity presents acutely with gastrointestinal symptoms, and chronically as peripheral neuropathy with alopecia. Many cases result in neurologic impairment or death, partially due to delay in diagnosis. Several treatment regimens have been proposed for thallium toxicity, but no single approach has been advantageous. We describe a case of thallium toxicity with rising in-hospital thallium levels treated with Prussian blue and Continuous Renal Replacement Therapy (CRRT).

Case Presentation: A 41 year old man with no significant past medical history originally presented with abdominal pain and bilateral lower extremity paresthesias. He was diagnosed with atypical Guillan-Barré Syndrome and received a four day course of IVIG with some neurologic improvement. He was subsequently discharged to a neurorehabilitation facility. He presented 10 days later with personality changes, hallucinations, abdominal pain and worsening bilateral lower extremity paresthesias. Exam was significant for flat affect, plantar sensory loss, 4/5 strength in bilateral lower extremities, absent patellar and Achilles reflexes and alopecia. Initial workup, including heavy metal screen was negative. EMG demonstrated a subacute distal axonal peripheral neuropathy. After an extensive workup, a serum thallium level was noted to be elevated at 158 ng/ml (reference 0-1ng/ml). The serum thallium level increased to 197 ng/ml prior to initiation of CRRT and Prussian blue therapy. The patient received a total of 30 days of Prussian blue and 22 days of CRRT with neurologic improvement. After extensive investigation, the source of the thallium was not found.

Discussion: Although well defined, thallium toxicity is extremely rare and therefore difficult to diagnose. Thallium is also not routinely included in heavy metal screening. Although the source of thallium was not found in this case, we suspect that thallium was being surreptitiously administered while the patient was in the hospital. The rise in serum thallium was noted prior to initiation of CRRT and Prussian blue, therefore tissue redistribution cannot explain the increase. The patient also did not receive any electrolytes that would have redistributed thallium into the circulation. Finally, although thallium is usually not well dialyzed due to its large volume of distribution, CRRT with administration of Prussian blue was successful in treatment of chronic thallium toxicity. Success of CRRT is most likely due to the fact that in this case, thallium was already distributed into the tissues, whereas acute thallium toxicity is best treated with intermittent hemodialysis because thallium has not distributed to the tissues.
Michigan-Clinical Vignette-Poster Finalist
George Gennaoui, DO

Title: Guillain-Bare Syndrome Complicating Acute Retroviral Syndrome

Authors: George Gennaoui D.O, Matthew Wilkins M.D, Raymond Hilu M.D. FACP, St. John Hospital and Medical Center, Detroit, Michigan

Introduction: Acute Retroviral Syndrome (ARS) is characterized by a nonspecific “flu-like” illness manifesting 1-4 weeks after acute infection with the human immunodeficiency virus (HIV). This often resolves spontaneously without medical attention however it can present with many atypical features requiring further attention. We present a classic case of ARS complicated by uncommon neurological developments.

Case Presentation: A 26-year-old African American female presented after multiple ER and urgent care visits with a one-week history of generalized malaise, rash, and progressive lower extremity weakness. On exam, she was febrile, tachypneic, and had a lacy reticular rash over her back and shoulders. Neurological exam was noted for 3 out of 5 strengths with diminished reflexes in the lower extremities. Laboratory workup demonstrated several electrolyte derangements, pancytopenia, acute liver injury, and reactive HIV screening test. Supportive therapy was initiated, however, the patient developed worsening neurological symptoms with extraocular muscle weakness, encephalopathy, and new onset seizure requiring ICU management. Lumbar puncture with routine analysis and MRI of the brain were unremarkable. Despite aggressive supportive care and addressing many of her metabolic derangements, her neurological symptoms progressed and the patient was unable to ambulate on her own. Neurology was consulted, and after ruling out other etiologies, patient was diagnosed with Guillain-Bare syndrome (GBS) and ultimately started on a 5-day course of IVIG. The patient was transferred to inpatient rehabilitation for continued care and was subsequently discharged to home with significant improvement of symptoms.

Discussion: This patient’s neurologic symptoms brings to attention a unique presentation of ARS of which the diagnosis was missed multiple times. As her symptoms worsened, she required admission for further evaluation and more extensive work up. The first approach in management is to provide supportive care, address any metabolic derangements, and perform appropriate diagnostic testing. In this case, patient’s primary symptoms were neurological, however, etiology was unclear and required MRI and LP testing. After ruling out many potential causes, Neurology was consulted and GBS was strongly considered and was treated accordingly. Two-thirds of patients with GBS have an infection prior to symptoms. This infection is typically gastroenteritis or upper respiratory tract infection. It is rare for acute retroviral syndrome to precede GBS. However, as identified in this case, it may be worth to consider screening for HIV when GBS is the predominating diagnosis.
Michigan-Clinical Vignette-Poster Finalist
Srinandan Guntupalli, MD

Title: It's not insulin resistance - It's Glargine resistance !!: An unusual Case of Lantus resistance.

Authors: Srinandan Guntupalli MD, Dhara Patel MD, Zeina Habib MD, Juliette Perzhinsky MD, Msc.

Introduction: Insulin resistance is a well described phenomenon which is often dreaded by physicians treating insulin dependent diabetes mellitus. However, resistance to solely glargine has not been described. We report first case of selective insulin resistance to glargine.

Case Presentation: A very complaint 65 year old female with multiple co-morbidities including diabetes mellitus type II and metabolic syndrome who was noted to have hyperglycemia despite her regular insulin therapy (glargine 56 units(U) twice daily with low dose insulin sliding scale of aspart). She was seen in office multiple times for the concern of hyperglycemia with blood glucose ranging in between 380-500. Even though insulin was titrated up to 100U twice daily and aspart 70U three times daily with strict dietary restriction, she continued to have hyperglycemia. Her HbA1c steeply raised from 6 to 12.5 in 5 months duration. During this period, she had two hospitalizations for varied reasons where she had well controlled blood glucose which was related to dietary compliance in hospital.

Upon patient request, her glargine insulin was changed to detemir at same dose. Surprisingly, with-in couple days after switch, she reported hypoglycemia (45). Her current blood glucose range from 110-140s. Anti-glargine auto-bodies or selective resistance to glargine was suspected for hyperglycemia. Glargine autoantibodies was sent out which was positive. Currently, She is doing well while her Hba1c came back to 7.3 within 3 months of switching over to insulin detemir.

Discussion: This case exemplifies the importance of history taking, broad differential diagnosis and paying attention to fine details in diagnosing a rare condition. More research is needed to understand selective resistance to glargine which can impact treatment of diabetes mellitus.
Michigan-Clinical Vignette-Poster Finalist
Puja Gupta, MD,MBBS

Title: Are Breast Cancer Staging Guidelines Failing Our Patients?

Authors: Maysarra Al-Jubori, MD; Khalid Mohamed, MD; Faisal Musa, MD; Lakshmi Swaminathan, MD; Rajiv John, MD

Introduction: The US National Cancer Institute Surveillance, Epidemiology, and End Results: (SEER) has found a significant increase in breast cancer in women below 50 years of age from 1976-2014. The American Society of Clinical Oncology (ASCO) and the National Comprehensive Cancer Network (NCCN) guidelines recommend against staging CT, PET, or bone scan stage I or II breast cancer for the identification of occult metastatic disease, unless worrisome signs or symptoms are present. These recommendations, however, are based on cancers diagnosed by screening women above the age of 50, in whom the risk of metastasis at diagnosis is low. The SEER database shows us that 38.2% of women below the age of 50 have regional metastasis at the time of diagnosis. Up to 5% of asymptomatic breast cancer patients have been diagnosed with metastasis. If caught in time and treated in a timely manner, women younger than 50 years diagnosed with regional metastatic breast cancer have up to an 86.6% 5-year relative survival rate.

Case Presentation: A 47-year-old female presented with left sided, positional chest pain. Physical exam identified a tender and fixed mass in the left breast that was later diagnosed as Stage IIa invasive ductal breast adenocarcinoma T2 N0, ER/PR positive and HER II negative. Per ASCO and NCCN guidelines, no imaging for staging was warranted, because the patient was asymptomatic. Within two months of the lumpectomy, prior to start of therapy, she presented with worsening shortness of breath. Workup diagnosed metastasis with a malignant left sided pleural effusion, subcentimeter pulmonary nodules, and spinal lytic lesions.

Discussion: Policy makers must see the need for separate imaging guidelines in women below the age of 50 with breast cancer, especially with increasing incidence in that age group.

Physicians must recognize that the national guidelines recommending against further imaging studies for patients with stage II breast cancer do not comment specifically on women younger than 50, that have been diagnosed outside the screening criteria. In women younger than 50, we must consider being more deliberate with imaging, in the context of increased chance of regional metastasis.

We challenge the ASCO and NCCN guidelines recommending against CT, PET, or bone scan for staging patients with stage II breast cancer, which do not comment specifically on patients that have been diagnosed with breast cancer outside the screening criteria.

References
Michigan-Clinical Vignette-Poster Finalist
Rahul Gupta, MBBS

Title: A demyelinating mystery of the brain: Acute disseminated encephalomyelitis with recurrence in a young adult


Introduction: Acute disseminated encephalomyelitis (ADEM), sometimes referred to as post-infectious encephalomyelitis, is an acute, rapidly progressive, autoimmune process that affects the central nervous system. The literature supports that ADEM is a rare demyelinating disease in adulthood, with possible autoimmune etiology and can closely mimic other demyelinating disorders, making it difficult to diagnose.

Case Presentation: We describe a rare case of acute disseminated encephalomyelitis with recurrence, with efficacy of high dose steroids and IVIG in its treatment. Our patient is a 27-year-old African American male with past medical history of asthma. His symptoms started in March 2013, when, after 2 months of flu-like symptoms, he was found comatose and taken to the emergency department. At that time, MRI with/without contrast showed abnormal T2/FLAIR signal abnormality within the midbrain and bilateral thalami, with larger focus on left side. During that admission, he required high-dose steroids and IVIG treatment with extensive rehabilitation, but was discharged home; his symptoms resolved almost completely over two months. Patient had a relapse of his symptoms around 3 months after first episode, with MRI revealing abnormality in similar region. Subsequently, weakness, blurry vision and/or frequent falls resulted in multiple admissions and recoveries, mostly with high-dose steroids and occasional IVIG. CSF and MRI findings did not suggest multiple sclerosis. Between 2014-2016, an elaborate work-up for other demyelinating disorders and central nervous system pathologies was unremarkable. In February 2017, he developed seizures. Repeat MRI showed extensive edematous changes in similar areas, with 2.6 x 1.2 cm peripheral enhancing lesion over left capsuloganglionic region. Due to suspicion of mass lesion, a brain biopsy was performed, which showed infiltration of inflammatory lymphocytes and macrophages; findings suggestive of encephalitis with demyelination. A diagnosis of acute disseminated encephalomyelitis with recurrence was consistent based upon clinical features, MRI findings and histological examination.

Discussion: Acute disseminated encephalomyelitis is a usually monophasic demyelinating disorder with recurrences posing a diagnostic challenge to treating physicians. Recurrences can suggest an alternative diagnosis or sometimes can be overlooked. MRI is the imaging modality of choice to diagnose ADEM. Lesions are hyperintense on T2 weighted FLAIR sequences. Paucity of trials on treatment of ADEM, gives very little options but high dose steroids and/or IVIG as mainstay.

References
Michigan-Clinical Vignette-Poster Finalist
Mohamed Hassanein, MD

Title: Statin-induced necrotizing autoimmune myopathy: An uncommon complication of a commonly used medication.

Authors: Mohamed Hassanein¹ MD, Yehia Saleh¹ MD, Robert Hillman³ DO, Howard Chang⁴ MD, PHD., 1 – Department of Internal Medicine, Michigan State University, East Lansing, MI, 3 – Department of Internal Medicine, Sparrow Health System, Lansing, MI, 4 – Department of Pathology, Sparrow Health System, Lansing, MI

Introduction: Statins are one of the most widely prescribed medications. They play a major role in reducing the incidence of cardiovascular diseases in susceptible patients. Myopathy is a well documented side effect of statin therapy. The exact mechanism of statin-induced myopathy is not yet fully understood. Necrotizing autoimmune myopathy is a rare cause of statin-induced myopathy.

Case Presentation: A 64 year-old gentleman presented with muscle weakness for 4 months. His past medical history is significant for hypertension, coronary artery disease, diabetes mellitus and hyperlipidemia. His medications include metformin, carvedilol, lisinopril, and atorvastatin. On evaluation, he had stable vital signs and weakness in all extremities. Initial laboratory Results: showed elevated liver function tests, a creatine phosphokinase (CPK) of 8200 IU/L and positive urine myoglobin. Despite discontinuing atorvastatin, his CPK remained persistently elevated. Tests for antinuclear antibody (ANA), anti-Jo 1 antibody, anti-double stranded DNA antibody, anti-scl-70 antibody and paraneoplastic autoantibodies were all negative. Computed tomography (CT) of the chest, abdomen and pelvis showed no evidence of malignancy. Electromyography (EMG) showed myopathy with fibrillations, indicating an inflammatory myopathy. Left thigh muscle biopsy was consistent with necrotizing myopathy. Anti - HMG CoA reductase antibody, ordered prior to the muscle biopsy, was strongly positive.

A diagnosis of statin-induced autoimmune myopathy was made, and the patient was started on immunosuppressive therapy. Intravenous and oral steroids were started, followed by intravenous immunoglobulin (IVIG). His muscle weakness, CPK, and liver functions gradually improved, and he was eventually discharged on oral steroids.

Discussion: Statin-induced necrotizing autoimmune myopathy is an extremely rare complication, with a reported incidence of 2-3 of every 100,000 patients treated with statins. It should be considered in patients presenting with myalgias, muscle weakness, and persistent elevation in CPK especially after discontinuation of statins. It’s pathophysiology remains unknown, but is thought to be related to statin induced autoimmunity against HMG-CoA reductase in genetically susceptible individuals. Clinically, patients present with proximal myalgias and muscle weakness, which may persist or even worsen after statins are discontinued. CPK usually exceeds 2000 IU/L. EMG shows small-amplitude motor-unit potentials with increased spontaneous activity. Muscle biopsy is consistent with immune-mediated necrotizing myopathy. Diagnosis is confirmed by testing for anti - HMG CoA reductase autoantibody. Patients are treated by discontinuing statins and initiating immunosuppressive therapy. Fortunately, patients have good prognosis, with improvement in muscle weakness and CPK level, as with our patient.

References
Title: Beta Blocker Overdose - A Sweet Alternative?

Authors: Jaspreet Hehar, Enrique Soltero, Hira Iftikhar, Bhavin Dalal, Beaumont Health, Royal Oak, MI

Introduction: Overdose with beta adrenergic antagonists can lead to profound hemodynamic instability. First line therapies include atropine, intravenous fluids, glucagon and vasopressors. These measures are not always sufficient and alternative therapies are sought. One such intervention is high dose insulin-euglycemic therapy.

Case Presentation: A 55-year-old Caucasian male presented to the ED with slurred speech and lethargy. He reported taking 200 tabs of 50mg metoprolol in an attempt to end his life. His vitals were within the normal range except for sinus bradycardia at a rate of 59. Physical examination revealed cool extremities. After intubation, his blood pressure (BP) and pulse dropped to 80/40 and 25, respectively. He was given atropine and glucagon with slight improvement initially but then became progressively more bradycardic so a glucagon infusion was initiated. This was followed by an infusion of regular insulin at 2 units/kilogram/hour with dextrose infusion to maintain euglycemia with a goal mean arterial pressure greater than 60. His BP improved to 122/49 and pulse to 57. Glucagon was gradually weaned off and then the insulin infusion was decreased by 0.5 units every two hours until it was stopped. The patient was asymptomatic and returned to baseline mental status before discharge home.

Discussion: Multiple studies have demonstrated the utility of insulin-euglycemic therapy in calcium channel blocker intoxication. Our case demonstrates that this therapy can also be an effective adjunct to standard treatment for beta blocker overdose. This is consistent with other published case reports, but more investigation is needed to elucidate the true role of insulin in the treatment toolbox of beta blocker overdose.

References

Title: Cutaneous Clues: Unusual Skin Metastases in Pancreatic Cancer

Authors: Kellen Hipp DO, Shahina Patel MD, Bianca Barbat MD, Amy Hadad DO, Lyle Goldman MD

Introduction: Pancreatic cancer is a malignancy that carries a notoriously poor prognosis, principally due to its penchant to metastasize quickly. Predictable spread to the liver, lungs, bone, and brain are common, and evidence of cutaneous metastasis originating from the pancreas as the classic Sister Mary Josephs nodule is well documented. Wider dissemination of pancreatic malignancy to the skin is an unusually rare finding. Cutaneous metastatic lesions are often mistaken for other skin conditions, especially when they are multiple, and spread far beyond the umbilical region.

Case Presentation: A 55-Year-old male with a history of muscular dystrophy presented to the hospital with diffuse abdominal pain that was associated with weight loss, fatigue, and poor appetite. Initial Computed Tomography (CT) scan of the abdomen/pelvis revealed a 6.5 x 7.1 cm mass in the liver with significant surrounding adenopathy. Subsequent magnetic resonance imaging (MRI) revealed a 3.0 x 3.7 cm solid mass in the tail of the pancreas and innumerable lung nodules consistent with metastasis. Biopsy of the liver mass confirmed the diagnosis of pancreatic adenocarcinoma. Due to his long-standing muscular dystrophy and chronic debility it was determined that he would not be a candidate for chemotherapy. He instead received palliative radiation and later enrolled in hospice care. Upon completion of his radiation, he complained of a bleeding nodule that appeared on his face in the days after treatment. He attributed this lesion to the radiation, however it continued to grow and multiple new ulcerated and bleeding cutaneous lesions developed on his scalp, neck, and shoulders over the following weeks. Lesions near the umbilical region were absent. Core biopsies of a lesion located on the anterior chest wall were analyzed and consistent with cutaneous metastasis of his pancreatic adenocarcinoma by histology and immunochemical staining.

Discussion: Cutaneous metastasis portends a grave prognosis and indicates extensive dissemination of malignancy, however it has also been reported as the initial presenting feature in cases of pancreatic cancer¹. The presence of a Sister Mary Joseph node is generally recognized as the most common cutaneous indicator of advanced abdominal or pelvic malignancy while examples of multiple skin metastases far from the umbilical region are far less common. Less than five available case reports detail pancreatic metastasis to the scalp. The unusual locations and ulcerated appearance of our patient’s skin lesions share many features with malignancy-associated skin conditions such as radiation necrosis and pyoderma gangrenosum. Our patient’s lesions represent late sequelae of known metastatic disease, however including skin metastasis in the differential diagnosis and distinguishing them from common skin conditions with biopsy is useful when there is no diagnosis of malignancy or the primary tumor site is unknown.

References

Title: Listeria Hydrocephalus: a Happy Ending?

Authors: Nicole Kapral M.D.¹, Mossum Sawhney², Ziran Yang², Fnu Abhishek M.D.³, ¹St. Joseph Mercy Oakland, Department of Medicine, ²Ross University School of Medicine, ³St. Joseph Mercy Oakland, Department of Neurology

Introduction: Bacterial meningitis is a relatively uncommon disease in the United States, with a reported incidence of approximately 1.38 cases per 100,000 population/year during 2006-07. Of these cases, Listeria monocytogenes (LM) is implicated in 4-8% of cases, with a fatality rate of 17-24%. Listeria meningitis may present throughout life, but has increased incidence in extremes of age as well as immunocompromised patients. Hydrocephalus is a known complication of meningitis, more prevalent in listeria patients (approximately 14% of cases). It is a poor prognostic marker, independent of age, CSF leukocyte count and coma, and increases mortality rates to 40-60%. In addition to antibiotic treatment, extraventricular drainage may be beneficial in select patient, dropping mortality rates by 14 percentage points according to one Spanish study. Here, we describe a patient diagnosed with Listeria meningitis and found to have hydrocephalus who responded well to extraventricular drain placement concomitantly with ampicillin/gentamicin antibacterial treatment.

Case Presentation: A 55 year-old-male with pertinent history of diabetes mellitus type 2, presented to the emergency department with complaints of fever, fatigue, and progressive, intermittent confusion over the preceding five days. He endorsed headache and neck pain with movement, but had negative Kernig's and Brudzinski's signs with no other focal deficits. Initial head CT was notable only for sinusitis. Lumbar puncture was performed revealing xanthochromia, elevated white cells with 84% PMN, elevated protein and decreased glucose. CSF and blood cultures both grew Listeria monocytogenes and the patient was subsequently placed on ampicillin and gentamicin for appropriate coverage. Despite initiation of antibiotics, the patient’s sensorium continued to decline. A repeat CT of the head revealed a new obstructive hydrocephalus. He underwent urgent ventriculostomy with external ventricular drain placement on day 4 of admission. Ampicillin and gentamicin infusion were initiated due to failure to maintain adequate drug levels with intermittent IVP administration. Patient's mentation significantly improved though he will likely need a ventricular shunt for several months while he continues rehab and recovery. Our patient was fortunate in that he survived and was able to maintain much of his mental capacity.

Discussion: Meningitis is difficult to establish clinically, as the classic triad of fever, nuchal rigidity and altered mental status is present in just 45-65% of cases. Blood and CSF cultures are positive in 45-65% and 10-30% of cases respectively. It is important clinicians have a low threshold for suspecting listeria, especially in immunocompromised patients. Mortality is high, and complications such as hydrocephalus must be investigated for early diagnosis and management. Ampicillin and gentamicin remain the standard of care, and extraventricular drain may benefit select patients. Given the high mortality rate of Listeria meningitis and associated hydrocephalus, our patient was fortunate to receive early treatment and make a significant recovery.

References


Title: A Blast in the Lung: A Case of Fatal Blastomycosis Presenting with Knee Swelling

Authors: Ojbindra KC, MD, Dilip Khanal MD, Pradeep Khanal MD, Sanjog Bastola MD, Anju Adhikari MD, Ashbina Pokharel MD, Yoshiko Nito MD, Ashbita Pokharel MD

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Introduction: Pulmonary involvement is the most common manifestation of blastomycosis. However, development of ARDS due to blastomycosis is uncommon and can have deadly outcome. Approximately 20% cases also have extrapulmonary manifestations mainly involving skin and bone. We report a case of fulminant blastomycosis in an immunocompetent individual initially presenting with knee swelling and pain, with gradual progression to ARDS and death.

Case Presentation: 50-year-old man with past medical history of knee osteoarthritis and recurrent unexplained skin infections presented with pain and swelling of right knee for 3 days. He was noted to have right knee swelling with tenderness and multiple healed small skin lesions on his face. He underwent several joint aspirations with one culture growing Streptococcus infantarium. His knee pain continued despite treatment with IV antibiotics. He underwent arthotomy with synovectomy and was diagnosed with rheumatoid arthritis (RA) with positive rheumatoid factor along with mild elevation of anti-cyclic citrulinated peptide (anti-CCP). Repeat synovial culture revealed no organism. He was treated low dose prednisone with diagnosis of RA. He then started to have left shoulder pain for which MRI was done that revealed abnormal appearing bone marrow in glenoid and humerus concerning for myeloproliferative disorder or neoplasm. Biopsy was negative for malignancy. He subsequently started to have shortness of breath. CT scan of chest revealed extensive diffuse lung nodularity. He was started on IV vancomycin and piperacillin-tazobactam for possible healthcare associated pneumonia. His dyspnea progressed leading to intubation and mechanical ventilation. Sputum, blood and Bronchoalveolar lavage (BAL) fluid cultures were negative for bacterial or fungal pathogens. He was started on steroids for possible rheumatoid lung and was placed on Extracorporeal Membrane Oxygenation (ECMO) for worsening hypoxia. Finally, his urine tested positive for Histoplasma antigen. He subsequently underwent left anterior thoracotomy and lung biopsy which revealed blastomycosis. He was started on IV amphotericin B for disseminated blastomycosis with ARDS. Despite IV antifungal therapy and multiple bronchoscopies to clear secretion his respiratory status gradually worsened eventually leading to his death.

Discussion: Unlike other dimorphic fungi, Blastomyces can cause severe infection in both healthy and immunocompromised individuals. Our case shows that diagnosis can be delayed due to confusing presentation and presence of other confounding factors. Skin, bone or joint manifestations with non-resolving pneumonia should raise the suspicion for this condition, especially in persons who live in or visit endemic regions. High index of suspicion and early workup with culture and non-culture diagnostic tests may prevent delay in diagnosis and give better chances of survival in this deadly disease.
Michigan-Clinical Vignette-Poster Finalist
Sharanjit K Khaira

Title: Expect the unexpected: A case of complicated cerebral abscess caused by Gemella morbillorum in a young female

Authors: Sharanjit Khaira, MD, PGYII; Sukhmanpreet Singh, MD, PGYII; Palaniandy Kogulan, MD, FACP; Department of Internal Medicine, Central Michigan University, Saginaw, MI

Introduction: Cerebral Abscess, a focal infection within the brain parenchymal tissue, is an uncommon entity, where frontotemporal lobe is classically affected. It may result from complicated paranasal sinusitis, sequelae of cranial trauma, or hematogenous seeding from distant source, with 15% of cases being cryptogenic. *Gemella morbillorum* (GM), a gram-positive, facultative anaerobic bacterium, is a part of normal flora of human oropharynx, genitourinary and gastrointestinal system. Infections related to GM are reported in literature as endovascular, or septicemia and infrequently as CNS such as cerebral abscess. Herein, we present a case of GM-induced complicated cerebral abscess in a young immunocompetent female.

Case Presentation: 23-year-old female was transferred from an outlying facility secondary to acute onset confusion and lethargy. Over the past 4-5 weeks, patient had been complaining about frontal headaches along with nausea and vomiting. She was given antibiotics including amoxicillin/clavulanate and azithromycin as outpatient by her primary physician for presumed diagnosis of sinusitis. However, despite the treatment, she developed mental status changes on the day of admission. Initial vitals were stable, however physical exam revealed lethargic female with significant aphasia, and right hemiparesis. CT head followed by MRI brain revealed left frontal lobe abscess, along with bifrontal extra-axial fluid collections. Patient was started on ceftriaxone and metronidazole empirically for cerebral abscess and underwent burr hole evacuation for parenchymal empyema evacuation. She initially did well, however, later began having recurrent episodes of focal-seizures, uncontrolled with repeated antileptic boluses and hence, was intubated and started on lorazepam infusion. Follow-up MRI brain revealed left frontal hypodense area transformed into an encapsulated abscess with significant enlargement. Patient underwent left frontal stereotactic intraparenchymal abscess drainage, post which significant improvement in symptoms was noted. Initial cultures from the abscess had shown gram variant organism, however, final cultures revealed *Gemella morbillorum*, which was surprising for the medical team. Based on the sensitivities, patient was discharged home on 8-weeks of ceftriaxone, and levetiracetam after complex hospital course with residual neurological deficits.

Discussion: Brain abscess is a life-threatening condition with mortality-rate reaching up-to 80% without prompt treatment. GM-induced cerebral abscess is very rare, yet potentially life-threatening entity with approximately only 9 cases reported till-date. Due to insidious presentation including nausea, vomiting and headaches, it remains a diagnostic and therapeutic challenge for the clinicians with difficulty identifying the primary septic site and hence, duration of the treatment. In patient hospitalized with cerebral abscess, the all-cause-mortality may vary from 5% to 32% with frequency of neurological sequelae ranging anywhere from 20-79%. Hence, clinicians need to be aware of this immersing organism involved in life-threatening conditions such as CNS-infections, as prompt diagnosis and therapeutic intervention remains the hallmark to improve morbidity and mortality among these patients.
Title: Metachronous Inflammatory Pseudotumor of the Lungs, Calvarium and the Meninges due to IgG4 Related Disease

Authors: Aditya Kotecha, MD; Hussein Saleh, MD; AbuFazl Shaik Mohammed, MD; Christina Desousa, MD; Geetha Krishnamoorthy, MD

Introduction: IgG4 related disease causes tumefactive lesions of the organs due to lymphoplasmacytic infiltration by IgG4 positive plasma cells and leads to fibrosis, and may be associated with elevated IgG4 level.

Case Presentation: A 28-year-old male with Cryptogenic Organizing Pneumonia 7 years ago and an episode of leucocytosis with lung nodules 2 years ago, presented with seizures. MRI of the brain showed calvarial and meningeal masses, which were resected. Biopsy showed spindle cell proliferation with polyclonal IgG4 positive plasma cells with negative ALK1 staining, consistent with Plasma Cell Granuloma (PCG), a subtype of inflammatory pseudotumor, an IgG4 related disease. CT thorax showed fibrotic changes of lungs. We obtained the slides from the 2009 lung biopsy and review was consistent with another subtype of inflammatory pseudotumor, fibrohistiocytic type with predominant macrophages and fibrosis. IgG4 cells were found rarely, since IgG4 positive cells decrease after steroid therapy, which had been started before lung biopsy. We made a diagnosis of IgG4 related disease with metachronous inflammatory pseudotumor of the lungs and meninges.

Discussion: Autoimmune pancreatitis was the first IgG4 disease described. IgG4 related disease can have multiorgan involvement at presentation or organ involvement can be metachronous. Prednisone treatment is first line and is effective if given prior to significant fibrosis. Rituximab has excellent efficacy in severe cases. Internists should be aware of IgG4 related disease, and if tissue biopsy is consistent with this diagnosis, extensive surgery can be avoided since the disease in the active phase responds to steroid or rituximab.

References

Michigan-Clinical Vignette-Poster Finalist
Elise Landa

Title: The Way to A Man’s Heart Shouldn’t Be Through His Esophagus

Authors: Elise Landa MD, Amin Pasha MD, Erin Goldman DO, Robby Singh MD, Kellen Hipp DO, Irina Shanidze MD, Christian Machado MD

Introduction: Radiofrequency catheter ablation is a very commonly performed procedure used for curative treatment of atrial fibrillation (AF). Although it is an effective and safe procedure, it is not without risk. The development of an Atrioesophageal fistula (AEF) is a rare but potentially fatal complication. As per previously described cases, surgery is the definite therapy for an AEF. However, establishing the diagnosis remains extremely challenging. We describe one such case of AEF after radiofrequency ablation.

Case Presentation: A 67-year-old Caucasian male with a past medical history of recurrent AF presented to the hospital with fever, chills and fatigue of three days duration. He had undergone radiofrequency ablation four weeks prior to presentation. He had also undergone cardioversion three days earlier. Laboratory testing revealed leukocytosis and lactic acidosis. He was started on Vancomycin and Gentamicin as per the sepsis protocol. Due to deteriorating clinical condition he was subsequently transferred to the Intensive Care Unit. The blood cultures grew *Streptococcus intermedius*. CT scan of his chest and abdomen was performed which revealed outpouching along the posterior wall of the left atrium consistent with a pseudoaneurysm or AEF. The antibiotic coverage was broadened to Vancomycin, Cefepime, Metronidazole and Fluconazole to cover for gastrointestinal flora. Despite several days of conservative medical management, patient’s condition did not improve and he continued to have intermittent chest pain. Subsequent surgical exploration of his chest revealed a perforation through his left atrium and pericardium with an adjacent defect in the esophageal wall. He underwent successful repair of the perforation after which he was eventually discharged from the Intensive Care Unit.

Discussion: AEF remains a dreaded complication of radiofrequency ablation as it can be difficult to identify. AEF symptoms are not specific which can lead to a delay in diagnosis. Patients often present with hemoptysis and sepsis. The median time for presentation is between day 6 and 59 post-ablation. The reported fatality rate is between 67-100%. As per the literature, there are three strategies that have been attempted in treating AEF - conservative management, esophageal stenting and surgical repair of the fistula. Conservative management Results: a fatality rate approaching 100%. Esophageal stenting is potentially of benefit in patients in whom esophageal perforation is present without fistula formation to the left atrium. The most successful treatment for proven or suspected AEF is surgery with repair of all damaged tissue. It is recommended that a multidisciplinary approach to care be used with coordination between the surgery, infectious disease, neurology and critical care personnel. Improving the awareness of this complication among primary care physicians and emergency medical professionals is required to decrease morbidity and mortality. Early diagnosis and surgery remains the mainstay in management.
Title: A Wedge shaped pulmonary infarct which is not due to an embolism.

Authors: Asim Mohamed, MD. Hasan Rana, MD. Raghavendra Kamath, MD, FACP, Internal Medicine Department - St John Hospital and Medical Center

Introduction: When a wedge-shaped pulmonary infarct is identified on chest imaging, pulmonary embolism is on the top of the causative etiologies. However, when a young healthy individual with no risk factors for venous thromboembolism develops an infarct, other etiologies must be explored.

Case Presentation: A 34 year old male with a past medical history of right nephrolithiasis and ureteric stent presented with a complaint of right flank and lower chest non-radiating pleuritic pain for one day. He admitted to snorting cocaine three days prior to admission and sustaining a minor blunt trauma to the right chest. He denied any urological symptoms, fever, cough, or sick contacts. He denied venous thromboembolic(VTE) risks factors including prolonged travel, immobilization, recent surgery, history of cancer, personal or family history of thromboembolic disease. The patient was afebrile with normal vital signs. Physical examination was unremarkable with non-tender, non-bruised, clear chest to auscultation bilaterally. Labs included a white blood count of 12.4 (Neutrophils 79%), a normal urinalysis, negative pro-calcitonin level and positive urine toxicology for cocaine metabolites and opiates. Chest X ray showed mild right basilar linear scarring versus atelectasis. Computer tomography(CT) of the abdomen with contrast done initially to evaluate right flank pain showed trace right pleural effusion with a wedge shaped hazy airspace consolidation in the lateral right lower lobe. CT angiography of the chest was done to further evaluate for pulmonary embolus which showed a wedge infiltration of the right lower lobe associated with pulmonary emphysema, but no pulmonary embolism was identified.

The patient received supportive care with fluids, parenteral analgesia with non-steroidal analgesics, narcotics, and antibiotic coverage of possible pneumonia. After considering the findings and not identifying another etiology, it concluded that his presentation was from cocaine induced pulmonary infarct.

Discussion: A wedge shaped pulmonary opacity is usually suggestive of pulmonary infarct from VTE. However, other etiologies must be explored especially with a history of cocaine use. Cocaine induced lung injury usually presents as diffuse alveolar damage and hemorrhagic alveolitis within 48 hours of use, known as crack lung. However, solitary lesions and infarcts have been reported and attributed to direct toxicity of cocaine, cocaine metabolites and its vasoconstrictive effects.
Michigan-Clinical Vignette-Poster Finalist
Swetha Murthi, MD

Title: Sometimes it is all in their head: Acute-onset hemiparesis in a young woman

Authors: Swetha Murthi MD MPH, Hemalatha Murugan MBBS DCH MPH, Kulothungan Gunasekaran MD

Introduction: A brainstem abscess is a potentially serious intracranial infection, and if left untreated result in nearly 100% mortality rate due to compression of medullary centers. We present a rare case of a solitary pontine abscess in a young woman, requiring acute stereotactic intervention.

Case Presentation: A 39 years old female presented with a 3-day history of nausea, vomiting, diplopia, and left-sided hemiparesis. One week prior to presentation, she underwent an uneventful dental extraction procedure for dental caries. On examination, vitals were normal. Her neurological examination was significant for dysconjugate gaze, bilateral horizontal and vertical nystagmus, hyperactive reflexes, left foot clonus and positive Romberg sign. Laboratory findings disclosed a WBC count of 14,000/mm3 with a left shift. Computed Tomogram (CT) of the head and CT angiogram of the head showed unremarkable findings. Magnetic Resonance Imaging of the brain revealed a ring-enhancing right pontine mass with edematous changes. Gram stain and cultures of the CSF were negative. The differential diagnoses included an abscess, infarction, and metastatic brain tumor. She was started on intravenous Vancomycin, Meropenem and Methylprednisolone. Due to worsening of symptoms, a repeat brain MRI was done which showed an increasing size of the brainstem lesion with worsened effacement of the fourth ventricle and cerebral aqueduct. A solitary pontine abscess was then diagnosed. She underwent stereotactic aspiration of brainstem lesion with pus removal and cultures grew Peptostreptococcus, Bacteroides, Streptococcus and Eubacterium. After the procedure, the patient's neurological status improved. She was discharged on Vancomycin and Meropenem for 6 weeks. She was followed up after 6 months with complete resolution of symptoms.

Discussion: Intracranial abscesses are complex focal infections of the brain that can be caused by a wide-variety of microorganisms. There are 3 known routes of inoculation: spread from infection of contiguous structures, direct contamination from intracranial instrumentation, and hematogenous spread. The diagnosis of brain abscess secondary to dental infection has typically been as a result of exclusion. Signs and symptoms of brain abscess include those that are seen in space-occupying lesions. Magnetic resonance imaging is useful for differentiating brain abscesses from brain tumors and infarcts (1). Brain abscess caused by dental and sinus infections typically involve aerobic and anaerobic streptococci, anaerobic gram-negative rods, Fusobacterium, Staphylococcus aureus, and Enterobacteriaceae. The treatment of brain abscesses involves a multidisciplinary approach involving neurosurgery and antimicrobial treatment (2). Brainstem abscess has a favorable outcome if treated adequately. In a stroke-like patient with or without fever, a brainstem abscess should be considered even if there is no meningeal irritation.

References

Michigan-Clinical Vignette-Poster Finalist
Rohan Naik, MD

Title: Quetiapine causing Atypical Neuroleptic Malignant Syndrome (NMS) and Acute respiratory distress syndrome (ARDS)

Authors: Rohan Naik, MD; Aditya Kotecha, MD; Ajit Thakur, MD; Hicham Krayem, MD; Camelia Arsene, MD, PhD, MHS; Geetha Krishnamoorthy, MD., Department of Internal Medicine, Sinai-Grace Hospital/Detroit Medical Center, Wayne State School of Medicine, Detroit, MI

Introduction: Typical antipsychotics are associated with a tetrad of fever, muscle rigidity, altered mentation and dysautonomia, which along with supportive laboratory findings constitute NMS. With the advent of atypical antipsychotics, atypical presentations of NMS have been reported. We present a case of Quetiapine overdose causing atypical NMS and ARDS.

Case Presentation: A 34 year old woman with bipolar disorder presented to our hospital agitated, after she was found with an empty bottle of quetiapine. Examination revealed diaphoresis, tachycardia, tachypnea, and diffuse bilateral crackles. Muscle rigidity, hyperreflexia and ankle clonus were absent. Lethargy and respiratory distress developed, necessitating intubation and mechanical ventilation. Patient was started on a drip of midazolam for sedation. Laboratory studies revealed a CPK of 24,454 U/L, AST 309 U/L, ALT 79 U/L, and LDH 583 U/L. She was not on any other medication. Initial CT-Head and Thorax were unremarkable. The next day, dyspnea worsened, with hypoxemia while on 100% FiO2. Chest X-ray showed bilateral alveolar infiltrates. She became febrile, and remained febrile with a maximum temperature of 41.2C. No clinical or radiographic evidence of infection was found. Lumbar puncture, blood, urine and respiratory cultures were negative. Temperature was unresponsive to acetaminophen and ibuprofen, but responded to IV Dantrolene and Bromocriptine, with a drop from 41.2C to 36.2C over eight hours. NMS has now resolved, but patient is gradually recovering from ARDS.

Discussion: Several case reports and a growing body of evidence suggest that atypical antipsychotics can cause an attenuated NMS, without rigidity or fever. Atypical NMS is also less severe and is associated with a lower mortality. Furthermore, benzodiazepines can mask muscle rigidity. This challenges the validity of present DSM-V diagnostic criteria for NMS, which requires muscle rigidity. It is also critical to differentiate NMS from Serotonin Syndrome and Malignant Hyperthermia as there is significant overlap of symptoms, with a different approach to managing of each of these conditions. Seroquel overdose may also independently cause ARDS.
Michigan-Clinical Vignette-Poster Finalist
Olusola Ogundipe, MD

Title: I’m getting a migraine from recurrent Hospitalizations: A case of Topiramate Induced Metabolic Acidosis

Authors: Olusola Ogundipe, MD - PGY1, St. Joseph Mercy Oakland, Pontiac, MI, Usman G. Master, MD, FACP - Chairman Division of Nephrology, St. Joseph Mercy Oakland, Pontiac, MI; Assistant Clinical Professor, Michigan State University., Anupam A. Sule, MD, PhD, FACP - Program Director, Transitional Year Residency, St. Joseph Mercy Oakland, Pontiac, MI., Benjamin Diaczok, MD, FACP - Program Director, Internal medicine, St. Joseph Mercy Oakland, Pontiac, MI.

Introduction: Topiramate is commonly used for migraine prophylaxis. It has multiple mechanisms of action including enhanced activity of GABA at non-benzodiazepine sites, inhibition of the NMDA glutamate receptor, and weak carbonic anhydrase inhibitor activity. This carbonic anhydrase inhibition is an infrequently reported precipitant of non-anion gap metabolic acidosis (NAGMA) that is resistant to therapy, producing severe hypokalemia with cardiac and neurologic sequelae. Physicians need to recognize this little reported cause of NAGMA.

Case Presentation: A 59-year old woman presented with dehydration secondary to diarrhea. Laboratory data revealed severe non-anion gap metabolic acidosis with a serum Bicarbonate of 10mEq/L, Potassium 2.0mEq/L and Magnesium 2.8mg/dL. Initial attempts at correction yielded moderate but non-sustained improvement in electrolytes. Her diarrhea resolved but the acidosis persisted. Urine chemistry revealed an anion gap of 57mEq/L, excluding diarrhea as the cause of NAGMA. Multiple myeloma with Light chain proximal tubulopathy and renal tubular acidoses were ruled out. A chart review revealed: 1). Patient had been on topiramate for migraine prophylaxis since 2013 and 2). She has had multiple admissions for metabolic acidosis with serum bicarbonate as low as 6mEq/L since 2014 with at least one episode of encephalopathy requiring ICU treatment. Each admission was preceded by a brief episode of diarrhea with resultant severe NAGMA. We weaned down topiramate and saw stabilization of patient’s electrolyte values. Follow up outcomes will be obtained by contacting the patient in three months.

Discussion: Urine chemistry with a calculated anion gap is a simple yet effective tool that can be used to differentiate causes of NAGMA. A negative urine anion gap is suggestive GI losses a cause for acidosis while a positive gap of 20 to 90 is suggestive of RTA. When RTA is suspected but history is not supportive, medication reconciliation can help identify inciting agents. This case captures a little reported but dramatic side effect of topiramate which inhibits the conversion of CO2 to Bicarbonate ion, resulting NAGMA with severely low bicarbonate levels. Even though the medication is approved as first line therapy in appropriately screened patients for the prevention of migraine headaches, it may still pose a safety risk.

Conclusion: Topiramate can cause severe refractory NAGMA in the setting of diarrhea. The only effective treatment is weaning and stopping medication. We suggest that its use in patients with recurrent diarrheal illnesses should be strictly avoided.
Title: Green thumb gone bad: a case of primary cutaneous histoplasmosis in an immunosuppressed gardener

Authors: Sean Pippen, MD; Suceil Sivasammye, MD; Ross Driscoll, MD, Western Michigan University Homer J Stryker School of Medicine

Introduction: Histoplasmosis is the most common endemic mycosis in the United States and of all the endemic mycoses, it is associated with the largest number of hospitalizations and deaths. Histoplasmosis most commonly presents in three forms which are acute pulmonary, chronic cavitary, and disseminated. Skin lesions may occur in all three forms of the disease but also as a rare form of primary cutaneous histoplasmosis as in the case presented here.

Case Presentation: A 64-year-old woman with medical history significant for rheumatoid arthritis being treated with adalimumab, methotrexate, and prednisone was seen with complaints of swelling, induration, and cracking of the skin of her left hand. She noted onset of symptoms one year prior to presentation, coinciding with a newly undertaken hobby of gardening. She also admitted to having a large bird bath in her yard near her garden which she cleaned regularly. Her symptoms began with induration of the palm which progressed to involve the dorsal aspect of the hand followed by cracking and fissuring of the skin. A few months later, she noted swelling and induration moving up the wrist and into the distal forearm. Referral was made at this point to a hand surgeon who performed a tenosynovectomy. Intraoperative cultures were positive for histoplasma capsulatum. Chest X-ray was normal and the patient had no respiratory symptoms. Ultrasound of the abdomen revealed normal sized liver and spleen, CBC and CMP were normal. She was treated with itraconazole for 9 months with resolution of her symptoms.

Discussion: Histoplasma capsulatum is an endemic mycosis which in the United States is most prevalent in the Mississippi and Ohio river valley regions. The organism is found in soil, particularly where bird and bat droppings are present. Upon infection, 99% of patients will have disease limited to the lungs. Only one percent of cases will progress to become disseminated, and fewer still will present as primary cutaneous histoplasmosis. Infection occurs through local trauma or direct inoculation, as likely occurred in this patient while gardening or cleaning her bird bath. Presenting features may include nodules, ulcers, abscesses, or molluscum contagiosum-like lesions. It is important to recognize that immunosuppressed patients have a tenfold increase in risk of disseminated infection compared to the general population. This case illustrates the importance of obtaining a good history, as this patient’s recent gardening and exposure to birds were clues to the diagnosis. Also highlighted is the importance of including rare conditions such as primary cutaneous histoplasmosis in the differential for skin lesions in the immunosuppressed patient.
Title: Brothers From Another Mother? A Case of Benign Metastasizing Leiomyoma with features of Lymphangiomatosis

Authors: Elisa Quiroz, Fernando Figueroa, Daniel Keena

Introduction: Benign Metastasizing Leiomyoma (BML) is an extremely rare and fascinating disease. While histologically benign, with low proliferative index, it is known to metastasize to distant sites; most commonly the lung. It occurs in young women with a history of uterine leiomyoma and while 130 BML cases have been reported a mere 7 cases had cystic lesions. We present a case of a 54-year-old female with multiple solid lung lesions that later progressed into cystic lesions.

Case Presentation: A 54-year-old female with history of multiple large uterine leiomyomas presented with abdominal pain. Computerized Tomography (CT) revealed numerous (approximately 30) solid pulmonary nodules. Fiber-optic bronchoscopy was performed with biopsy but was nondiagnostic. Pulmonary function testing revealed a moderate to severe, partially reversible, obstructive ventilatory defect consistent with asthma and concomitant chronic obstructive pulmonary disease (COPD). Follow up CT scan demonstrated an increase in size and number of the nodules. Biopsy of the nodules revealed a smooth muscle mesenchymal lesion with low mitotic index. Immunohistochemical stains were positive for muscle markers such as desmin and smooth muscle actin (SMA) putting mesenchymal lesions such as benign metastasizing leiomyoma high on the differential. The caveat on pathological analysis is that MITF, HMB45, and MART, markers in lymphangioleiomyomatosis (LAM), were also positive. On subsequent CT scan many of the solid lesions had evolved into cysts. The patient continues to follow up regularly with her pulmonologist.

Discussion: Benign Metastasizing Leiomyoma (BML) is a rare disease that presents in young women with a history of uterine myoma. The lesions are most often found in the lung but have been reported in the skin, bones, nervous system, abdomen and mediastinum as well. The pathophysiology of BML is not well understood. Some authors suggest that surgical manipulation may predispose to bloodstream dissemination. As the tumors are typically positive for estrogen and progesterone receptors, it has also been suggested that they arise from the hormone-sensitive in situ proliferation of smooth muscle bundles. This case is particularly interesting in that the histopathology leads to an ambiguous diagnosis. While the patient has as history of uterine myoma and markers for BML are positive, the markers that would typically confirm the diagnosis of LAM were also positive suggesting that there is an overlapping pathogenesis or common progenitor cell between these two disease processes. Additionally, the evolution of the lesions from solid to cystic is the first of its kind in the literature.
Michigan-Clinical Vignette-Poster Finalist
Zabila Saeed, MD

Title: Posterior reversible encephalopathy syndrome (PRES) in a hypertensive patient with hypercalcemia, renal failure and encephalopathy

Authors: Zabila Saeed, MD, Resident Physician, Internal Medicine, Mercy Health Saint Mary’s, Michigan State University, Grand Rapids, MI.

Introduction: Posterior reversible encephalopathy syndrome (PRES) is a clinical radiographic syndrome of heterogeneous etiologies characterized by headaches, altered consciousness, visual disturbances and seizures. Multiple myeloma (MM) is characterized by the neoplastic proliferation of plasma cells producing a monoclonal immunoglobulin. This is a clinical vignette of a patient with hypertension, encephalopathy, renal failure and hypercalcemia presenting with posterior reversible encephalopathy syndrome with underlying multiple myeloma (MM).

Case Presentation: 68-year-old female with history of diabetes mellitus type II, hypertension, hyperlipidemia, chronic back pain presented to the ED with a few days of progressive altered mental status. Patient was oriented to her first name only with tachycardia, BP 207mm Hg/89 mm Hg. Labs showed Hb of 11g/dl, WBC 12.2/mcL, creatinine 2.1mg/dL, serum calcium of 14.8mg/dL. EKG displayed tachycardia. CT of the head displayed edema within the subcortical white matter of the posterior parietal and occipital lobes bilaterally. Patient received IV fluids with labetalol with IV nicardecine for slow BP control. Patient was hospitalized for 4 weeks. During the first week, MM was diagnosed with markedly elevated kappa free light chain of 1235 with low IgA, IgG, and IgM. Beta-2 microglobulin and uric acid both elevated. Skeletal survey showed compression fractures of T11, T12, L1 and L2, lytic lesions of the skull and right scapula with diffuse osteopenia. Bone marrow biopsy was positive for 80% plasma cell involvement. Patient remained encephalopathic despite improvement in blood pressure and hypercalcemia. Lumbar puncture was order to determine involvement of CNS which was negative. EEG monitoring was abnormal indicative of mild diffuse encephalopathy. MRI Brain obtained after 4 days consistent with PRES. Patient was treated with aggressive IV fluids with one dose of dexamethasone and two doses of pamidronate with improvement in serum calcium levels. During the second week of hospitalization, patient decompensated with acute hypoxemic respiratory failure secondary to pulmonary edema and concerns for pneumonia requiring intubation and transfer to ICU. She was treated with antibiotics and diuretics. She was extubated after 3 days although with persistent respiratory difficulty requiring intermittent BIPAP and O2 via nasal cannula. During the third week of hospitalization, chemotherapy was started with Velcade and dexamethasone. Subsequently, patient’s encephalopathy improved during 4th week of hospitalization and was transferred to long term rehabilitation center.

Discussion: This case illustrates a unique presentation of multiple myeloma with PRES which is commonly associated with hypertension followed by renal diseases and chemotherapy. This patient had hypertension which was potentiated in the setting of acute kidney injury. PRES associated with multiple myeloma seemed to occur with coexistent hypertension and renal failure liked due to disordered cerebral auto regulation and endothelial dysfunction. Recognition of this presentation is critical to institution of appropriate therapy and avoiding chemotherapeutic drugs that would predispose the patient to develop PRES.

References

Michigan-Clinical Vignette-Poster Finalist
Ali Sahlieh, MD

Title: Immunoglobulin-Troponin I Complex Falsely Elevating Troponin I Levels

Authors: Ali Sahlieh, MD; Michael Barnes, MD

**Introduction:** Cardiac troponins (cTn) are highly sensitive and specific biochemical markers of myocardial injury. They are the preferred serologic tests for patients with suspected myocardial infarction (MI) as recommended by European and American societies of cardiology since 2000 [1].

However, cTn levels may be elevated in clinical conditions other than MI. Rarely, false cTn elevation may occur because of analytical interferences with the troponin immunoassay.

**Case Presentation:** 48-year-old male was brought to the emergency room after a syncopal episode while running on a treadmill. The patient reported no significant past medical history but his father had coronary artery disease. His physical examination was unremarkable.

Electrocardiogram showed sinus rhythm with inferolateral ST-T wave changes concerning for ischemia. Cardiac troponin I (cTnI) was significantly elevated up to 2 ng/mL (normal range 0.00 to 0.05 ng/mL). CK-MB and kidney function were normal. Echocardiogram showed normal left ventricular ejection fraction with no distinct regional wall motion abnormalities.

Cardiac catheterization showed normal coronary arteries. Cardiac MRI illustrated small spots of delayed hyperenhancement compatible but not definitive for myocarditis. An extensive evaluation for an infectious or autoimmune cause was negative.

Upon follow up visits, the patient had an exercise stress test and repeat cardiac MRI that were normal. However, his cTnI levels were persistently elevated over several months.

When the validity of cTnI levels in our patient became questionable, the lab performed several measures to identify any interference with the assay used. Serial dilutions did not result in decreased cTnI values which supported false positivity [2]. Testing for known interfering antibodies and monoclonal proteins was unremarkable. Two other commercial testing assays for cTnI were performed but resulted in elevated cTnI as well.

The patient’s serum was sent to another hospital for further testing. cTnI level was elevated again, but concomitant cardiac troponin T level was normal. The lab performed a polyethylene glycol (PEG) precipitation technique which is used for suspected immunoglobulin bound analytes. With this technique, any immunoglobulin-bound cTnI would be precipitated by PEG [3]. There was a significant difference in cTnI levels with and without PEG treatment. It was concluded that he has an immunoglobulin-troponin I complex that increases troponin I half-life in his circulation and Results: in an elevated troponin I levels regardless of the assay used.

**Discussion:** A false positive troponin result is a reminder that although troponin plays an important role in the diagnosis of myocardial injury, it should not be the only criterion for establishing the diagnosis. Falsely elevated troponin should also be considered as a diagnosis when data do not corroborate with clinical presentation, and when other biomarkers of myocardial injury are normal. Such a diagnosis makes further testing and procedures unnecessary, and can alleviate patient worries.
References


Michigan-Clinical Vignette-Poster Finalist
Hamza N Salam, MBBS MD

Title: An unusual complication of group 1 pulmonary hypertension: Patient with polymyositis associated interstitial lung disease presented with high degree AV block.

Authors: First author: Hamza Najam Salam, MD.

Introduction: Polymyositis is part of a group of disorders which causes widespread systemic inflammation. Involvement of the lungs is not uncommon and is mostly associated with sub acute pneumonias. A minority of patients develop pulmonary fibrosis from polymyositis associated interstitial lung disease. Several studies have identified ILD progression as one of the most common causes of death in these patients. Pulmonary hypertension if diagnosed and managed early, can significantly improve morbidity and mortality in such patients. We describe a case involving a patient with well controlled polymyositis on a stable chemotherapy regimen but with advanced interstitial lung disease causing pulmonary hypertension and life threatening complications.

Case Presentation: We describe a case of a 59 year old female with interstitial lung disease and biopsy proven polymyositis. Patient presented with the chief complaint of dizziness, worsening shortness of breath, leg swelling, nausea and vomiting. She was taking mycophenolate and prednisone for her polymyositis. At baseline, patient was on home oxygen at 4-6Ls via nasal canula. She was independent with her activities but had severe exercise intolerance. On physical exam, we saw a middle-aged female with a radial pulse in 30s, bilateral pitting edema up to the knees, jugular venous distention and coarse bilateral crackles in the all lung zones. EKG findings included third degree heart block with a junctional rhythm. Chest x-ray showed severe pulmonary vascular congestion. Transthoracic 2D echocardiogram revealed a hyper dynamic left ventricle and estimated the left ventricular EF as 75-80% with grade II diastolic dysfunction. Emergent right heart catheterization was done which identified a severely dilated, hypokinetic right ventricle and elevated right ventricular systolic pressure (60mmHg). A temporary transvenous pacemaker was inserted during the procedure. High resolution CT scan was done to rule out interstitial lung disease progression. The scan showed cardiomegaly, pericardial and pleural effusions, multifocal ground-glass opacities, interlobular septal thickening and traction bronchiectasis. Initial management involved diuresis with lasix and metolazone which significantly improved her symptoms. After discharge, patient was scheduled for a permanent pacemaker implantation. She would also follow at the pulmonary hypertension clinic for the need to start vasodilators such as sildenafil and ambrisentan.

Discussion: In the chronic stage, interstitial lung disease often causes chronic pulmonary fibrosis resulting in pulmonary hypertension. Cardiopulmonary complications from pulmonary hypertension dominate the disease course and dictate survivability. There is little consensus on the ideal treatment regimen for patients with interstitial lung disease associated with polymyositis. Several studies have shown that histological sub classification of the interstitial lung disease has proven to be a better predictor of survival than radiographic appearance or the clinical presentation. Based on these observations, we deduced that the identification of interstitial lung disease and pulmonary hypertension in patients with polymyositis is frequently missed and histological sub classification along with early diagnosis is needed for optimal management.
Michigan-Clinical Vignette-Poster Finalist
Rovin Saxena

Title: Arthritis Here, Here, and Here with Sepsis - Oh, and He is Immunocompetent without Comorbidities

Authors: Rovin Saxena MD; Buddhi Hatharaliyadda MSIV; Jacky Duong DO
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Introduction: Septic arthritis is a medical emergency with a high mortality which occurs predominantly as monoarticular arthritis in the knee, elbow, shoulder, or hip. The incidence has been reported as 1:100,000 in the general population. Only 15% of these cases involve multiple joints. The most common isolated organism in non-gonococcal septic arthritis is Staphylococcus aureus which accounts for 60% of cases. Streptococcus pyogenes is a less common cause involving only 13% of patients. However, Streptococcus pyogenes septic arthritis predominantly occurs in single joints. We report an uncommon presentation of acute polyarticular septic arthritis (PASA) due to Streptococcus pyogenes, complicated with sepsis in an otherwise healthy, immunocompetent, young adult.

Case Presentation: A 27 year-old male presented with pain in the right knee, left ankle, and generalized body aches for two days. He denied intravenous drug use, urethral discharge, insect bites, or trauma. He was sexually inactive for six months. On arrival, his temperature was 103 F, pulse 128/bpm, BP 135/83 mm/hg, and respirations were 21/min. Physical examination showed erythema, warmth, and tenderness to palpation of the left ankle, left elbow, left forearm, right knee, and right second digit interphalangeal joints. Integumentary examination was negative for epidermal punctures or abrasions. His sternoclavicular and temporomandibular joints were also involved. Laboratory studies revealed leukocytosis of 34,500, BUN 26, creatinine 1.82, lactic acid 3.6, ESR 45, and CRP 47.5. Liver function was normal. Blood cultures, HIV, Hepatitis, STD, and urine drug screen panels were negative. ANA, anti-dsDNA, RF, and uric acid levels were normal. He was started on Vancomycin, Ceftriaxone, Clindamycin, and IV crystalloid fluid, subsequently admitted to the intensive care unit. Right knee was aspirated and fluid analysis revealed a WBC of 92,750. Synovial fluid cultures grew Streptococcus pyogenes, thus antibiotics were deescalated to Ceftriaxone and Clindamycin. After an eleven day hospital course, he showed significant improvement and was discharged on 3 weeks of Ceftriaxone. Follow up showed complete resolution.

Discussion: PASA is rarely seen in young and healthy, immunocompetent individuals without risk factors. In addition, Streptococcus pyogenes is an uncommon etiology, especially affecting multiple joints. Only four cases have been reported in the medical literature with those individuals being over 50-years-old. Our patient was a young, healthy adult with no personal or family history of immunodeficiencies. He did not have risk factors including, trauma, STDs, intravenous drug use, diabetes mellitus, chronic renal failure, rheumatoid arthritis, lupus, or malignancy. We confirmed this with a detailed history, thorough physical examination, and extensive testing to rule out other causes. Physicians should be aware that PASA can result from Streptococcus pyogenes in otherwise healthy, young patients and may lead to critical illness. Due to the high morbidity and mortality, clinicians should always entertain a high index of suspicion and perform a complete work-up. This allows for early recognition and administration of disease specific treatment.

References
Michigan-Clinical Vignette-Poster Finalist
Anita Shallal, MD

Title: Heartbreak in a Storm: Cardiogenic Shock due to Takotsubo Cardiomyopathy Induced by Thyrotoxicosis

Authors: Shallal, A., M.D.¹, Jacob, C., D.O.¹, Mendonca, D., M.D.², Rogers, C., D.O.².
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Introduction: Thyroid storm is a rare and potentially deadly complication of hyperthyroidism that can have profound effects on the myocardium. Takotsubo cardiomyopathy (TM) is a diagnosis of myocardial dysfunction in the absence of coronary artery disease (CAD) that is usually precipitated by a stressful event. We present a complex case of multi-organ failure that occurred in the setting of thyrotoxicosis.

Case Presentation: A 62-year-old female with a history of hypertension, diabetes, and chronic pain was found unresponsive in her home. In the emergency department, she awoke with chest pain, and initial troponin was 0.15ng/mL with an electrocardiogram (EKG) of sinus tachycardia, heart rate of 130bpm. Computed tomography (CT) of the head was negative for acute process. Thyroid-stimulating hormone was less than 0.01U/mL, with free T4 of 2.7ng/dL, and thyroid peroxidase antibody was elevated at over 1,300U/mL, consistent with a diagnosis of Grave’s disease. She was started on treatment for hyperthyroidism with methimazole and a beta-blocker. On hospital day three, she developed sudden respiratory distress and chest pain, with hypotension and hypoxia. An EKG showed a pattern suggestive of acute anterior ST-elevation myocardial infarction (STEMI). She was intubated at the bedside for hemodynamic instability and emergently taken to the cardiac catheterization lab. The Results: revealed no CAD, however there was a severely reduced ejection fraction of 25% with apical ballooning consistent with a diagnosis of TM. She was started on a milrinone drip and transferred to the intensive care unit (ICU). During her prolonged stay, complications included ischemic hepatopathy with coagulopathy, acute kidney injury, and ventilator associated pneumonia. Methimazole was temporarily discontinued due to her liver abnormalities and she was managed with propranolol and corticosteroids, along with supportive ICU care measures. She was discharged to a rehabilitation facility on hospital day 18 and subsequently made a complete recovery.

Discussion: This case illustrates a rare association of Takotsubo cardiomyopathy and thyrotoxicosis, with only 17 other cases reported. A review of the literature notes the variability in presenting signs of TM, suggesting that 2% of all patients presenting with STEMI carry this diagnosis. Cardiogenic shock is a rare initial presentation of TM, with an associated high mortality rate. This case highlights both the importance of early recognition of thyrotoxicosis, as a delay in diagnosis could lead to disastrous outcomes, as well as the complexity of treating hyperthyroidism in the setting of acute hepatic failure. This suggests an area where alternative treatment options should be investigated. Finally, it emphasizes that severe and sudden onset heart failure can be potentially reversed with both timely diagnosis and appropriate medical management. Healthcare providers should be aware of this uncommon and potentially fatal association of thyroid storm and Takotsubo cardiomyopathy.
Michigan-Clinical Vignette-Poster Finalist
Suceil Leela Sivsammye Sr, MD

Title: Unilateral Pulmonary Edema: A menacing sign

Authors: Suceil Sivsammye M.D.; Karun Badwal M.D.; Prashant Patel D.O.; Guramrinder Thind M.D.

Introduction: Unilateral Pulmonary edema (UPE) is an unusual entity which occurs in approximately 2.1% of those presenting with acute cardiogenic pulmonary edema. Almost all cases occur in association with severe mitral regurgitation as opposed to an occurrence rate of 6% in those with bilateral pulmonary edema (BPE). This makes UPE a key diagnostic clue for clinicians that is often overlooked due to a lack of familiarity.

Case Presentation: A 69-year-old man presented with chest pain that began at rest approximately 30 minutes prior to arrival. Initial EKG showed ST-segment elevations in V2 through V5, consistent with acute anterolateral infarct. The patient underwent left heart catheterization revealing 100% stenosis of the proximal left anterior descending artery for which a stent was placed. An intra-aortic balloon pump (IABP) was placed for an ejection fraction of 35% and 3+ mitral regurgitation. He was transferred to the critical care unit and started on norepinephrine, vasopressin and milrinone. A repeat EKG was performed which showed resolution of his initial ST elevations. A follow up echocardiogram the next day showed an ejection fraction of 40% with normal mitral valve function so the IABP was removed. He continued to remain on the same doses of vasopressor support following IABP removal, however his oxygen requirements worsened requiring emergent intubation. A post-intubation chest x-ray revealed bilateral consolidations that were significantly more prominent on the right, consistent with unilateral pulmonary edema. A stat echo was subsequently performed showing an ejection fraction of less than 10% and evidence of severe mitral regurgitation. Shortly after the echocardiogram, the patient developed pulseless electrical activity and resuscitation was unsuccessful.

Discussion: In the clinical setting, patients with unilateral pulmonary edema often have a delay in treatment when compared to those with BPE. They are more likely to receive inappropriate antibiotic use due to misdiagnosis and additional findings of leukocytosis in 72% of patients. When compared to those with BPE these patients have lower systolic and diastolic blood pressures, higher use of invasive and noninvasive ventilation, and higher catecholamine use due to the acuity of condition. Among those with severe mitral regurgitation and unilateral pulmonary edema, in-hospital mortality is 39% with a risk of death 6.9-fold higher than that of patients with BPE. Other causes of UPE include vascular and bronchial obstruction, congenital heart disease, and prolonged rest on one side however severe mitral regurgitation remains the main cause. Due to the anatomy of the mitral valve and the greater predisposition for the posterior leaflet to prolapse, patients are more likely to present with right-sided UPE as regurgitant flow is directed toward to the right pulmonary veins. Early recognition and use of echocardiography are of great benefit in these patients with prompt surgical intervention being essential in management.
Michigan-Clinical Vignette-Poster Finalist
Suceil Leela Sivsammye Sr, MD

Title: A Rare Congenital Cause of Myocardial Infarction in an Adult

Authors: Suceil Sivsammye M.D.; Chris Jacob M.D.; Prashant Patel M.D.; Aness Al-Khateeb M.D.

Introduction: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is an uncommon and lethal disease. It has an incidence of 1 in 300,000 live births and accounts for 0.4% of patients with congenital heart disease. It carries a 90% mortality within the first year of life making it a rare diagnosis into adulthood.

Case Presentation: A 33-year-old man presented with a complaint of acute left-sided chest pain after getting into an altercation with his boss. Pain radiated to his left upper extremity and lasted several minutes. Troponin was elevated at 0.038 ng/ml and rose to 0.928 ng/ml after the initial check. Electrocardiogram revealed sinus bradycardia with a nonspecific intraventricular conduction delay. During heart catheterization, the left main coronary artery (LCA) was not visualized on injection of the aortic root but was indirectly visualized on left ventricular angiography, as well as right coronary artery (RCA) angiography, as originating from the pulmonary artery. His ejection fraction was 40% and he had anterior wall hypokinesis. He subsequently underwent repair of the ALCAPA by tunneling of the LCA into the aorta with autologous pericardium.

Discussion: In fetal life, the origin of the LCA from the pulmonary artery is permitted because flow in both the anomalous LCA and normal right coronary is antegrade. This is due to the fact that both the pulmonary arterial and systemic arterial pressures are equal. Myocardial ischemia ensues after flow in the LCA is reversed, secondary to the decrease in pulmonary arterial pressure during the neonatal period. The left-to-right shunt causes a “coronary steal” phenomenon which leads to abnormal left ventricular perfusion. The extent to which collateral circulation between the RCA and LCA germinates during this essential period regulates the degree of myocardial ischemia. Only 10-15% of patients with ALCAPA survive into adulthood; intercoronary collaterals are a prerequisite for survival however blood supply to the subendocardial region is often not sufficient leading to chronic left ventricular subendocardial ischemia. This can be visualized as delayed subendocardial enhancement on magnetic resonance imaging and is an important finding, especially in asymptomatic adults, as it may predict an increased propensity for malignant dysrhythmias. No history of previous cardiac complaints and a nearly normal EKG are common findings. The diagnosis is made primarily by echocardiography and heart catheterization. Early surgical repair to create a dual coronary system is the standard of care for long term survival. Prognostic outcome is related to the preoperative myocardial damage as measured by the left ventricular function. Other complications that can occur in adulthood due to undiagnosed disease includes left ventricular dysfunction, mitral regurgitation and sudden cardiac death; 5% of cases may be associated with other cardiac anomalies such as atrial septal defect, ventricular septal defect, and aortic coarctation.
Michigan-Clinical Vignette-Poster Finalist
Philip Vendittelli, DO

Title: Paradoxical Thrombus-in-Transit caught in Patent Foramen Ovale

Authors: Philip Vendittelli DO, Sushruth Edla MD, Rajaninder Sharma MD, David Rodriguez MD

Introduction: Thrombus-in-transit is usually noticed in the right atrium or right ventricle. However, it’s extremely rare to visualize a paradoxical embolus in the left atrium and left ventricle. Here we present the case of a 33-year-old male with a paradoxical thrombus-in-transit originating in the right atrium, passing through a patent foramen ovale (PFO) and extending into the left atrium and left ventricle.

Case Presentation: A 33-year-old male was admitted for chest pain, dyspnea and cough for 3 weeks which suddenly worsened over 12 hours. Physical examination was remarkable for sinus tachycardia and left calf edema. A CT angiogram of the chest revealed bilateral pulmonary embolism (PE). Transthoracic echocardiogram (TTE) discovered a 10x1.5cm thrombus in the right atrium, passing through a PFO and extending into the left atrium, ventricle and outflow tract. Transesophageal echocardiogram (TEE) confirmed these findings. Lower extremity ultrasound revealed a right-sided deep vein thrombosis. The patient was urgently taken for open thrombectomy and closure of the PFO. A few days later the patient had persistent dyspnea and underwent ultrasound-assisted catheter-directed thrombolysis for treatment of the submassive PE. He tolerated both procedures well and was discharged to home in stable condition.

Discussion: An extensive thrombus-in-transit, extending across a PFO into the left atrium and left ventricle with impending paradoxical embolism is extremely rare. Imaging with TTE and TEE is crucial to evaluate the location and extent of the thrombus. Emergent open thrombectomy is paramount in preventing catastrophic systemic embolization. This is the first description of a right atrial thrombus-in-transit with paradoxical extension into the left ventricle treated with both open thrombectomy and ultrasound-assisted catheter-directed thrombolysis successfully.
Minnesota-Clinical Vignette-Poster Finalist
Ryan Balko, MD

Title: Spontaneous tumor lysis syndrome in malignant peritoneal mesothelioma

Authors: Ryan Balko MD, Lucinda Gruber MD

Introduction: Spontaneous tumor lysis syndrome (STLS) is an uncommon complication of malignancy, most often associated with hematologic malignancies. The incidence in solid malignancies is far less common. The diagnosis of STLS in patients with solid malignancies requires high clinical suspicion.

Case Presentation: A 44-year-old male with malignant peritoneal mesothelioma and recurrent ascites diagnosed 5 months earlier presented to the emergency department. Two months earlier, he had completed 3 cycles of cisplatin, pemetrexed, and bevacizumab with no clinical response. He presented to the Emergency Department with a subjective feeling of dehydration and lightheadedness when moving from sitting to standing. He did not notice any diminished urine output in the days leading up to presentation, but did state he was taking ibuprofen frequently for abdominal pain as well as diuretics for ascites. On admission, laboratory studies were significant for a creatinine of 4.7 mg/dL, sodium of 123 mmol/L, and potassium of 5.7 mmol/L. Urine studies showed calcium oxalate crystals and granular casts. CT abdomen showed a 7.1 cm enlarging adrenal mass with innumerable peritoneal and omental metastases. Findings were considered consistent with non-oliguric acute tubular necrosis, presumably due to increased abdominal pressure secondary to the abdominal masses, as well as ibuprofen and diuretic use. With aggressive fluid resuscitation, on day 2 his creatinine began trending down to 3.2 mg/dL. His urine output increased up to 3 liters per day. His potassium remained elevated at 6.1 mmol/L. His hyperkalemia was treated with IV furosemide. An abdominal ultrasound showed no significant ascites. On day 3, his creatinine improved further to 2.3 mg/dL with BUN trending down and adequate urine output. Again, his potassium was elevated, which was unexpected given his otherwise good clinical improvement. Given his persistent hyperkalemia and tumor burden, STLS was considered. His calcium returned at 8.8 mg/dL, phosphorous at 5.2 mg/dL, magnesium at 2.1 mg/dL, and uric acid significantly elevated at 18.4 mg/dL, consistent with STLS. IV fluids were continued and he was started on sodium bicarbonate, IV furosemide, and rasburicase. On subsequent days, his creatinine, phosphorous, and uric acid returned to normal limits. His pain continued to increase throughout hospitalization, and his functional status declined. Medical oncology and palliative care evaluated him, and he was ultimately transitioned to comfort cares. He passed away on hospital day 10.

Discussion: This case demonstrates the diagnosis of STLS requires a high clinical suspicion in a patient presenting with kidney dysfunction and multiple electrolyte abnormalities in the setting of solid bulky or hematologic malignancy. It also shows that working diagnoses need to be reexamined as new information becomes available. Persistent unexplained hyperkalemia in the setting of a malignancy should prompt consideration of STLS.
Minnesota-Clinical Vignette-Poster Finalist
Grace Choong, MD

Title: No horsing around with lupus: A case of cauda equina syndrome

Authors: Grace Choong, M.D. and Floranne Ernste, M.D.

Introduction: Systemic lupus erythematous (SLE) is an autoimmune disorder affecting multiple organ systems, including the central and peripheral nervous systems. Neuropsychiatric manifestations are manifold, but primarily present as transverse myelitis, meningitis, encephalitis or psychosis.

Case Presentation: A 33 year-old woman, with a history significant for SLE and antiphospholipid syndrome, presented with acute onset “saddle anesthesia”. Within the last month, she had been hospitalized for an acute lupus flare and was taking 20 mg prednisone and 400 mg hydroxychloroquine at presentation. Physical exam was significant for decreased rectal tone, decreased sensation to light touch over labia majora and perineum, and brisk patellar reflexes bilaterally, suggestive of cauda equina syndrome. Laboratory values showed a significantly elevated anti-dsDNA, low complement levels, elevated anti-phospholipid and beta-2 glycoprotein antibodies, positive lupus anticoagulant, but normal inflammatory markers. Neurology and Rheumatology were urgently consulted. A lumbar puncture revealed a glucose of 54, elevated protein, and WBC 16, with negative infectious work-up. An MRI lumbar spine indicated sacral root and conus medullaris enhancement without evidence of cord infarction. An EMG was consistent with a sacral radiculopathy. A prior thoracic MRI did not show transverse myelitis. Neurology was concerned for an HSV lumbar radiculitis and started a 10-day course of acyclovir. Yet, due to lack of confirmatory evidence for herpes radiculitis, neuropsychiatric lupus remained high on the differential and the patient was given methylprednisolone for 3 days followed by 60 mg prednisone. After 5 days of therapy, the patient began to have improvement of urinary retention and was subsequently discharged on a slow steroid taper. The patient returned to Rheumatology clinic in 4 weeks with a significant improvement of her symptoms, but continued to have sacral dermatomal hyperesthesia with worsening patellar hyperreflexia and highly elevated anti-dsDNA. An MRI cervical/thoracic spine did not reveal any inflammatory lesions. Based on these Results, it was determined her neurological symptoms were a result of neuropsychiatric lupus and the patient was started on CellCept.

Discussion: This case highlights an unusual presentation of neuropsychiatric SLE as cauda equina syndrome. Cauda equina syndrome is a medical emergency and requires prompt attention to prevent irreversible neurological deficits. Given the varied presentations of neuropsychiatric lupus, it is imperative to rule out an infectious process, especially in patients who are chronically immunosuppressed. A lumbar puncture and radiographic imaging should be part of the initial evaluation. MRI has a higher sensitivity to evaluate for other processes, such as neoplasia, ischemia, transverse myelitis or demyelination. Neuropsychiatric lupus must always remain high on the differential, especially in patients with known antiphospholipid syndrome. Once an infectious process has been ruled out, prompt initiation of high dose steroids for treatment of SLE is important, followed by maintenance therapy with CellCept or azathioprine to prevent long-term neurological sequelae.
Minnesota-Clinical Vignette-Poster Finalist
Caitrin M Coffey

Title: Dropped Gallstones Mimicking Ovarian Carcinomatosis

Authors: Caitrin M. Coffey, Alessia Buglioni, Jason D. Eckmann, Daniel C. DeSimone, Mayo Clinic, Rochester, MN

Introduction: Laparoscopic cholecystectomy is rarely complicated by dropped gallstones, which can lead to chronic intraabdominal inflammation, infections, and granuloma formation. Early recognition of the sequelae from dropped gallstones could prevent unnecessary imaging, infectious workup, and antibiotics and allow for earlier treatment with surgical retrieval.

Case Presentation: A 72 year-old female with a history of hypertension, hyperlipidemia, hypothyroidism, prior cholecystectomy, and hysterectomy with right oophorectomy for adenomyosis was referred to Infectious Diseases Clinic for consultation regarding abdominal pain, due to concern for peritonitis. Ten months prior to presentation, the patient underwent cholecystectomy for acute cholecystitis, after which she developed persistent abdominal pain, fatigue, and anorexia. Four months after the procedure, she developed redness, pain, and drainage from the umbilical trocar site. She was found to have markedly elevated inflammatory markers, and despite negative cultures and normal leukocyte count, she was treated with multiple rounds of antibiotics including cephalexin, amoxicillin-clavulanate, and minocycline, without improvement. CT scans of the abdomen and pelvis showed multiple enhancing soft tissue densities, which were biopsied with pathology showing benign inflammation, fibrosis, and reactive changes. The patient was then referred to our institution for further evaluation.

CT scan of the abdomen and pelvis was repeated at the time of Infectious Diseases consultation, and showed a left ovarian mass concerning for malignancy, with intra-abdominal inflammatory changes suggestive of carcinomatosis. The patient was referred to Gynecologic Surgery who recommended a left oophorectomy. Intraoperatively, a cystic ovarian mass was removed and inflammatory appearance with pus was noted. Pathology revealed benign ovary with benign inflammatory changes, as well as bile deposits with surrounding giant cell reaction.

Discussion: Gallstones are spilled into the abdominal cavity in 3.8-14% of laparoscopic cholecystectomies, yet only a small percentage of these cause complications, which can occur months or many years after cholecystectomy. Dropped gallstones or bile deposits that are not removed may act as inflammatory niduses in the abdominal cavity, and have been reported to cause infections, abscesses, chronic inflammation, foreign body reactions, inflammatory adhesions, sinus tracts, fistulae, dysmenorrhea, and small bowel obstruction. Risk factors for these complications include age, male sex, acute cholecystitis, spillage of pigment stones, >15 stones, large (>1.5 cm) stones, and perihepatic stones. CT imaging may show hypodense to hyperdense nodules, which can mimic peritoneal seeding from malignancy. Prevention of complications has only been reported via retrieval of dropped stones and irrigation and removal of spilled bile. Treatment varies depending on type and location of complication, but generally includes drainage of abscesses if present, and removal of stones. Intrapelvic gallstones should be included in the differential for patients undergoing a gynecologic workup who have a history of cholecystectomy, and may be misdiagnosed as malignant deposits.

References
Minnesota-Clinical Vignette-Poster Finalist
Jaime De La Fuente

Title: Is the High Worth the Torsades?

Authors: Jaime De La Fuente MD¹, Jared Bird MD², Korosh Sharain MD², Vasken Keleshian MD¹, Nandan Anavekar MBBS², 1) Mayo Clinic, Department of Internal Medicine, Rochester, MN, USA, 2) Mayo Clinic, Division of Cardiovascular Diseases, Rochester, MN

Introduction: Polymorphic ventricular tachycardia (PVT) is a life threatening arrhythmia resulting from a variety of mechanisms. For the general internist, it is important to recognize over the counter medications with abuse potential which can lead to this serious arrhythmia.

Case Presentation: A 29-year-old female with a history of bile acid diarrhea and chronic abdominal pain presented for an outpatient esophagastroduodenoscopy. Prior to the procedure, she developed severe nausea, vomiting, and dizziness, prompting direct admission to the general medicine service.

Initial negative work up included: complete blood count, extended electrolytes, pregnancy test, urinalysis, urine drug screen, and serial troponins. Electrocardiogram demonstrated a prolonged QTc interval of 549 msec. Upon further history, she revealed she had been consuming more than 200 mg (100 pills) of loperamide daily, for over a year, to relieve her abdominal pain and diarrhea. Her last dose of loperamide was 48 hours prior to presentation.

During hospital day one she developed palpitations and had a pre-syncopal episode. Review of her cardiac monitor revealed PVT. She was promptly started on IV magnesium and isoproterenol to decrease the QTc interval and reduce the probability of further PVT. This initially abated further PVTs. However, while weaning down the isoproterenol she had recurrence of runs of PVT. Thus, IV lidocaine was added to her regimen. Significant QT interval prolongation continued despite loperamide discontinuation, raising the teams suspicion for an underlying congenital long QT2 syndrome. Thus, she was transitioned to nadolol without further occurrence of PVT.

Nine days later patient was discharged from the hospital with a QTc of 460 msec on nadolol. Further outpatient work up two months later, including electrocardiogram, stress test, and genetic testing, was negative for congenital long QT syndrome. QTc at that time was 430 msec. Patient’s nadolol was discontinued.

Discussion: This case illustrates the importance of being familiar with over the counter QT interval prolonging agents, the possibility of loperamide being used as a drug of abuse, and the treatment of acquired PVT. It is believed that the mechanism of the prolonged QT by loperamide involves the inhibition of cardiac sodium and potassium channels. As demonstrated by this case, it is also critical to understand that the cardiac effects of loperamide, especially when abused and consumed for an extended period of time, will last much longer than the reported half-life of 11 hours. Additionally, congenital QT syndrome can potentially be unmasked when taking these agents, and as such should be maintained on the differential, and treated with beta blockade therapy.

References

**Minnesota-Clinical Vignette-Poster Finalist**  
**Jason David Eckmann, MD**

**Title:** An eighteen year-old male with massive pulmonary embolism and a liver mass

**Authors:** Jason D. Eckmann, M.D.¹, Kevin P. Quinn, M.D.², Laura E. Raffals, M.D.²  
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**Introduction:** Hepatocellular carcinoma (HCC) is a malignancy most commonly identified in older patients with chronic liver disease. Infrequently, a subtype called fibrolamellar hepatocellular carcinoma (FLHCC) can be seen in younger patients without underlying hepatic pathology, and for this reason is often delayed in its diagnosis. Here, we present a case of a young man with a massive pulmonary embolism who was ultimately diagnosed with FLHCC.

**Case Presentation:** An 18-year-old male with no significant past medical history presented with one week of rapidly progressive dyspnea and cough. On initial examination, he was found to be tachycardic, hypoxic and hypotensive. Laboratory evaluation noted mildly elevated aminotransferases, and international normalized ratio (INR) of 1.5 (normal, 0.9 - 1.1). CT of the chest with intravenous contrast demonstrated extensive bilateral pulmonary emboli involving the main pulmonary arteries and segmental branches. Echocardiogram showed a severely enlarged right ventricle with markedly decreased systolic function. He was given intravenous fluids, started on anticoagulation, and admitted to the intensive care unit where a search for precipitating causes was initiated. Ultimately, CT of the abdomen and pelvis showed a large heterogeneous, lobulated mass within the right hepatic lobe. Also noted was a large adjacent lymph node mass, and multiple enlarged, enhancing mesenteric lymph nodes within the right abdomen. He subsequently underwent ultrasound-guided liver biopsy with histopathology showing large polygonal cells with eosinophilic cytoplasm, surrounded by fibrous “lamellae” of collagen. Genetic analysis confirmed the presence of the DNAJB1:PRKACA fusion protein, consistent with a diagnosis of FLHCC. Due to the patient’s advanced disease, he was initiated on three months of 5-fluorouracil and interferon-alpha, with plan for subsequent consideration of surgery.

**Discussion:** FLHCC is a rare hepatic malignancy, representing between 0.6% and 8.6% of all HCC cases, with an annual incidence of 0.02/100,000. In contrast to conventional HCC, FLHCC affects young, predominantly Caucasian individuals, has no gender predilection, and is not associated with underlying liver disease. Serum aminotransferases and alpha-fetoprotein are usually normal or minimally elevated. Imaging typically reveals a large, heterogeneous, hypervascular mass, often with central scar or calcifications, in a background of normal liver parenchyma. FLHCC stains positive for both hepatocellular and biliary markers, and genetic analysis reveals the DNAJB1:PRKACA fusion protein, which is unique to FLHCC. The mainstay of therapy is surgical resection, which is feasible in greater than two thirds of patients. The prognosis for resectable FLHCC is generally favorable as compared to HCC, although recurrence is common. Chemotherapy and radiation are reserved for surgically unresectable disease, with overall poor prognosis in these patients. Clinicians should be aware that in young patients in whom malignancy is suspected, a lack of underlying liver disease or risk factors does not preclude the presence of HCC, and abdominal imaging should be performed to evaluate for FLHCC.

**References**

Title: Narrowing the differential on a narrowed esophagus

Authors: Patrick Hoversten\(^1\), Kavel Visrodia\(^2\), Jeffrey A. Alexander\(^2\), \(^1\) Department of Internal Medicine, Mayo Clinic, Rochester, MN, \(^2\) Department of Gastroenterology and Hepatology, Mayo Clinic, Rochester, MN

Introduction: The differential diagnosis for esophageal phase dysphagia is broad and includes both mechanical and neuromuscular etiologies. In some instances, a patient’s history and/or endoscopic findings can be diagnostic. However, less common causes of dysphagia can present a diagnostic challenge for multiple reasons, including the clinician’s potential unfamiliarity with the disease state, and specific aspects of work-up necessary to establish the correct diagnosis. The objective of this case is to familiarize practitioners with a rare and likely under-recognized cause of esophageal dysphagia, and illustrate the significance of appropriate utilization of multiple diagnostic strategies across various subspecialties to evaluate a common chief complaint when the etiology is not immediately evident.

Case Presentation: A 60-year-old woman with a longstanding history of ileal Crohn’s disease in remission presented with two weeks of dysphagia to solid food in the absence of odynophagia, reflux, or weight loss. She had also recently experienced her first episode of esophageal food impaction that resolved spontaneously. A barium esophagram demonstrated a diffusely small caliber esophagus without evidence of reflux. During EGD, diffuse stenosis was encountered in the proximal esophagus preventing passage of the endoscope, and required dilation. Esophageal biopsies of the stenotic region demonstrated intraepithelial lymphocytosis with rare dyskeratotic cells (Civatte bodies), a pathologic finding seen in lichen planus. On clinical exam, the patient was found to have an erythematous left buccal mucosal lesion. Given the high index of suspicion for esophageal lichen planus (ELP), the lesion was biopsied and pathology demonstrated lichenoid mucositis compatible with a diagnosis of lichen planus. The patient’s ELP was successfully managed using a combination of serial EGD dilations and oral topical budesonide therapy with vigilant attention to potential sequelae of chronic steroid use.

Discussion: Lichen planus is an idiopathic inflammatory disorder of mucocutaneous tissue, most frequently involving the skin or oral cavity and, in rare cases, the esophagus resulting in symptomatic dysphagia.\(^1,2\) ELP typically occurs in middle-aged women and has a predilection for long stricture formation beginning in the proximal esophagus, as demonstrated in this case. Establishing a diagnosis can be challenging, often requiring clues from the clinical history, esophagram and endoscopic appearance, as well pathology. However, the differential diagnosis for dysphagia secondary to a diffusely narrowed esophagus in an adult is relatively limited, and includes eosinophilic esophagus, caustic esophageal injury, and less commonly ELP. Characteristically, necrotic keratinocytes and lichenoid lymphocytic infiltrates are seen in ELP. Multiple treatment modalities have been proposed, including systemic/topical immunosuppressive agents and serial endoscopic dilation, or a combination thereof.\(^3\) Although a rare cause of dysphagia, ELP is now viewed as a likely under-recognized etiology of dysphagia and should be considered by clinicians when encountering middle-aged patients presenting with esophageal dysphagia secondary to proximal and/or diffuse esophageal strictures.\(^4\)

References


Title: Drug-Induced Rhabdomyolysis with Tenofovir and Atorvastatin

Authors: Brenden Ingraham, Michael Klajda, Paras Karmacharya, Vaidehi Chowdhary

Introduction: Severe rhabdomyolysis resulting in debilitating proximal muscle weakness and acute kidney injury in a patient with hepatitis B on tenofovir and chronic statin use.

Case Presentation: 70-year-old male with CLL and hepatitis B reactivation after treatment with a BTK inhibitor (subsequently stopped) presented with one week of progressive proximal muscle weakness. He takes diltiazem and aspirin for chronic atrial fibrillation, atorvastatin for hyperlipidemia, and tenofovir (nucleotide reverse transcriptase inhibitor; initiated 6 weeks prior) for hepatitis B. He was previously active but could not lift his arms above his head or rise from a chair at presentation. He could ambulate with a walker by taking small, shuffling steps. Mild pain in the pelvic/shoulder girdles. No distal weakness, diplopia, dysphagia, sensory abnormalities, or rashes. Review of systems otherwise unremarkable. Pertinent exam findings include jaundice, lower extremity proximal strength 3/5, upper extremity proximal strength 3-4/5, and distal strength 5/5. Pertinent admission labs include creatine kinase 27,500, aldolase 279, LDH 1360, creatinine 1.4 (baseline 1.3), AST 1942, AL T 943, total bilirubin 9.2, direct bilirubin 6.2, albumin 3.0, INR 1.0, and urinalysis revealed large amount of hemoglobin but < 3 RBCs. Anti-Jo-1, MyoMarker 3, anti-HMGCR, and ANA were negative. He was tentatively diagnosed with rhabdomyolysis secondary to drug-induced myopathy and acute kidney injury, and tenofovir and atorvastatin were discontinued. Despite this, his weakness worsened, CK rose to 112,000, and creatinine was 2.0 on hospital day 4. There was concern for immune-mediated necrotizing myopathy related to atorvastatin, so methylprednisolone 1000 mg daily for 5 days was started empirically. EMG showed severe proximal myopathy with features suggestive of necrosis. Triceps biopsy revealed rare necrotic fibers associated with a slight myopathy and no inflammatory changes making immune or auto-immune-mediated etiology less likely. He began to improve by hospital day 6 with a CK of 3,175 and a creatinine of 1.6. He was discharged home with an oral prednisone taper and physical therapy. He is doing well now off the tenofovir and atorvastatin.

Discussion: The incidence of tenofovir-induced rhabdomyolysis is extremely rare only being described in case reports. The incidence of atorvastatin-induced rhabdomyolysis is 0.6/10,000 person-years. The risk of rhabdomyolysis increases with the concentration of the statin. Atorvastatin is metabolized through the CYP450/3A4 pathway in the liver, which is not affected by tenofovir. However, he had decreased liver function related to his hepatitis B, which would decrease the clearance of atorvastatin. It is unclear if his acute episode was the result of initiating tenofovir 6 weeks prior or if the statin was the culprit in the setting of deteriorating liver metabolism. Regardless, it is imperative to be mindful of the medication list, rare adverse reactions, and ever-changing physiology in complex patients.
Minnesota-Clinical Vignette-Poster Finalist
Claire L Jansson-Knodell, MD

Title: Feeling the Heat: A Fever of Unknown Origin Solved

Authors: Claire Jansson-Knodell, M.D., Resident, Internal Medicine, Mayo Clinic, Tariq Azam, M.D., Resident, Internal Medicine, Mayo Clinic, Isabel Hujoel, M.D., Resident, Internal Medicine, Mayo Clinic, Xin Zhang, M.D., Resident, Internal Medicine, Mayo Clinic, Patricio Escalante, M.D., Assistant Professor, Pulmonary and Critical Care Medicine, Mayo Clinic

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is a rare and life-threatening occurrence that results from excessive immune activation. It is primarily a syndrome seen in pediatrics and is considered unusual in adults.

Case Presentation: A 26-year-old female with a medical history remarkable for an outside diagnosis of an undetermined autoimmune disease presented with fevers, rash, arthralgia, and dyspnea.

On exam, her heart rate was 124 bpm, blood pressure was 124/59 mmHg, temperature was 38.5° C, oxygen saturation was 97% on 2 L of nasal cannula. Her exam was significant for anterior cervical lymphadenopathy, hyperpigmented rash, no synovitis, tachycardia with normal S1 and S2, and inspiratory crackles in the bases of her lungs bilaterally.

Laboratory Results: showed a leukocytosis, CRP 176.8 mg/L, ferritin 9010 mcg/L, Triglycerides 539 mg/dL, AST 72 U/L, ALT 24 U/L, and LDH 883 U/L. Antiphospholipid antibodies, complement levels, cryoglobulins, serum proteins were unrevealing. Viral, bacterial, and fungal testing was negative. A soluble (CD25) interleukin-2 receptor level was elevated at 2262 u/mL. IL-18 levels were severely elevated. Significantly decreased spontaneous natural killer (NK) cell cytotoxicity was seen.

Echocardiography showed an RVSP of 81 mmHg consistent with pulmonary hypertension. Chest imaging showed extensive reticular interstitial and interlobular septal thickening in both lungs, diffuse adenopathy, and splenomegaly. PET CT showed diffuse moderately increased FDG uptake suggestive of a systemic inflammatory process involving spleen, bone marrow, lungs, and lymph nodes. The spleen was measured at 14.5 x 6.5 x 16.5 cm. Bone marrow biopsy demonstrated scattered histiocytes with hemophagocytosis.

She was initiated on steroids and anakinra for adult-onset Still's disease as well as macitentan for pulmonary hypertension and did well clinically. Her ferritin decreased to 3100 mcg/L and CRP to 13 mg/L. She was discharged from the hospital.

Discussion: Common triggers for HLH in adults include hematologic malignancies, infections, and immunosuppression. Rheumatologic diseases can rarely be a trigger (only 8% of cases).\(^1\) HLH is diagnosed when 5 of 8 diagnostic criteria are met: fever, splenomegaly, cytopenias in 2 cell lines, hypertriglyceridemia, hemophagocytes in bone marrow, low or absent natural killer cell activity, serum ferritin >500 mcg/L, and elevated soluble IL-2 (CD25) levels.\(^2\) Our patient met 7 of 8 criteria.

Pulmonary hypertension is an unusual manifestation of Still's disease. Pleural and pericardial disease resulting in cough, chest pain, and dyspnea are more frequent causes of cardiopulmonary disease with adult-onset Still’s, but pulmonary hypertension is rare and documented only in a few case reports. Anchoring bias and diagnostic momentum were astutely avoided in order to properly diagnose...
and treat the underlying etiology of her symptoms. Over time, with treatment of her rheumatologic condition and with an endothelial receptor antagonist, her RVSP improved to 39 mmHg, her exercise capacity increased, her fevers diminished, and her pain became manageable.

References

Title: “Shall We Thromboylze?”

Authors: Ronstan Lobo¹, Eelco FM Wijdicks², Adam P Sawatsky¹

¹Department of Medicine, Mayo Clinic, Rochester, Minnesota.
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Introduction: Acute hemiparesis should prompt immediate investigation. One of the most common reasons for acute onset hemiparesis is an ischemic stroke. Time is of the essence as early thrombolysis reduces morbidity and mortality. However, it is important for physicians to consider other causes based on careful history and physical examination.

Case Presentation: An 82-year-old lady with a history of type II diabetes mellitus, hyperlipidemia and hypertension presents with a sudden onset of left arm and leg weakness. She was sitting in church when she developed left-sided neck and shoulder pain, followed shortly thereafter by left-sided hemiparesis. She had no trauma and was not taking any antithrombotic medications. On examination, she had 0/5 power in the left upper and left lower limbs. Sensation was preserved. Other neurological examinations, including cranial nerves was normal. Her calculated NIH Stroke Scale was 8. Blood tests revealed normal platelet counts, APTT and INR. A non-contrast CT scan of the head revealed no abnormalities.

A telestroke consultation was initiated to address whether IV thrombolytics should be administered. Because of the neurologist’s concern about the ongoing left neck and shoulder pain, a CT angiography of the neck and chest was performed, which excluded arterial dissection or intramural hematoma. Because of the ongoing clinical suspicion, an MRI of the cervical spine was performed, revealing a left epidural hematoma extending from C2-C5 with associated compression of the spinal cord. The patient began to spontaneously recover neurologic function after the MRI and was managed conservatively.

Discussion: Epidural hematomas are usually seen in the setting of trauma, even minimal trauma. When it occurs spontaneously, it is seen usually in the setting of a bleeding predisposition, such as thrombocytopenia or antithrombotic therapy. There are case reports in the literature of spontaneous cervical epidural hematoma with no identifiable cause. This case highlights the importance of good history taking and being suspicious when the history ‘does not make sense’. The presence of persistent neck pain in the setting of acute hemiparesis should prompt evaluation for etiologies other than ischemic stroke. If this patient had received IV thrombolytics, it certainly could have led to disastrous consequences. Primum non nocere.
Minnesota-Clinical Vignette-Poster Finalist
Joshua C Pritchett, MD

Title: Immaculate ComPLEXion: A case of dermatomyositis flare, and the art of the rheumatologic “Hail Mary”

Authors: Joshua Pritchett, MD1, Cyril Varghese, MD1, Mohamad Adada, MD, PhD1, Robert Ward, MD1, Max Martin, MD1, Nicholas Canzanello, MD1, Delamo Bekele, MBBS1, Anthony Sidari, MD2, Kevin Moder, MD1, 1Mayo Clinic, Rochester, MN, 2San Antonio Military Medical Center, San Antonio, TX

Introduction: In addition to the classic cutaneous manifestations of dermatomyositis (Gottron’s papules/sign, heliotrope eruption, shawl sign) and frequent association with underlying malignancy, disease “flares” can be associated with devastating end-organ manifestations including interstitial lung disease (ILD). Unfortunately, at this time no formal guidelines are available to inform the management of dermatomyositis-associated ILD, for which the prognosis is exceedingly poor.

Case Presentation: We present the case of a 52-year-old female, past medical history significant for Hashimoto’s thyroiditis and multiple sclerosis, with recent diagnosis of triceps biopsy-proven dermatomyositis featuring primary cutaneous involvement of the bilateral hands/digits, elbows, and buttocks. Serum enzyme immunoassay with immunoprecipitation revealed strongly positive anti-melanoma differentiation-associated protein 5 (MDA5) antibody. Unfortunately, despite two weeks high-dose prednisone (60mg/day), cutaneous lesions progressed and she was admitted for urgent inpatient evaluation and management. Exhaustive screening for occult malignancy, including PET-CT, was unremarkable. On admission, she received cyclophosphamide and 3 days IVIG + methylprednisolone. While her cutaneous lesions remained relatively stable, the patient developed progressive hypoxemic respiratory failure, ultimately requiring intubation on hospital day 17 despite adequate diuresis and empiric antibiotic coverage. MDA5+ associated rapidly progressive ILD was presumed to be the cause of her respiratory decline.

After an exhaustive search of available literature and consultation with several experts as well as members of the interdisciplinary care team, it was determined that evidence was significantly lacking to guide urgent management in this patient. Ultimately, following discovery of an electronic abstract entitled: “Plasma Exchange (PLEX) for Refractory MDA5 Myositis and ILD,” it was decided to initiate PLEX in our patient based largely on extensive conversation with the Authors: of this successful case. She responded rapidly, with successful extubation <48 hours later. After 7 cycles of PLEX (conducted every-other-day) her respiratory status has stabilized, cutaneous lesions have markedly improved, and she no longer requires ICU level care.

Discussion: Patients with MDA5+ antibody subtype of amyotrophic dermatomyositis are at increased risk of developing rapidly progressive ILD, which is associated with an exceedingly high mortality rate. Currently there is no defined treatment for this disease entity. Numerous techniques including IVIG, cyclophosphamide, rituximab and polymixin hemoperfusion have been attempted and documented with limited success. After review of literature, we are likely the second case report where PLEX was successful in treating this disease. While the exact mechanism is not understood, it can be presumed that while immunosuppressive agents reduce the population of autoimmune cell subtypes, PLEX may aid by direct removal of circulating pathogenic MDA5+ autoantibodies and/or other pathogenic agents which have yet to be fully characterized in patients with this condition. As such, PLEX may be offered as a rescue therapy in refractory cases of this disease.

References
Title: Not for kids only: new-diagnosis Henoch-Schönlein purpura in an adult

Authors: Wil L. Santivasi, M.D. and John C. Lieske, M.D.

Introduction: While IgA vasculitis, or Henoch-Schönlein purpura, is most often considered a pediatric disease, up to 10% of cases occur in patients over the age of 18. Compared to children, adult patients with IgA vasculitis are more likely to present with palpable purpura, are more likely to demonstrate renal involvement, and are less likely to experience gastrointestinal effects.

Case Presentation: A 40-year-old man presented to the emergency department with a month-long history of “foamy” urine and two-week history of a worsening rash on his hands and feet. On presentation, he was found to be hypertensive to 145/87, with heart rate 97 and temperature 36.8 C. Physical examination disclosed non-blanching, violaceous palpable purpura in a centripetal distribution. Initial laboratory evaluation revealed normal kidney function (serum creatinine 0.9), elevated blood glucose (281), serum albumin of 2.7, and total cholesterol of 241. Urinalysis revealed large occult blood, 3+ protein, 31-40 red blood cells (>25% dysmorphic), 4-10 white blood cells, hyaline casts, granular casts, fatty casts, free fat, and oval fat bodies. C-reactive protein was elevated to 16 ( nl < 8.0). A 24-hour urine collection confirmed 3.3 grams of total protein. A biopsy of a skin lesion revealed leukoclastocytic vasculitis by light microscopy with IgA deposition within papillary dermal vessels by immunofluorescence. A kidney biopsy revealed mild segmental mesangial proliferative glomerulonephritis with significant mesangial IgA deposition. There were no crescents or diabetic changes. Thus the diagnosis of IgA vasculitis (Henoch-Schönlein purpura) was made. A tapering course of oral prednisone was initiated (40 mg to 10 mg over 4 weeks). On follow up at 3 weeks the rash had resolved. On follow up at 8 weeks, proteinuria had improved substantially (24-hour excretion 819 mg).

Discussion: This case illustrates a surprising, though not uncommon, diagnosis in an adult patient that presented with rash and heavy proteinuria. While the most common causes of nephrotic-range proteinuria in adults are diabetic nephropathy, focal segmental glomerulosclerosis, membranous nephropathy, and minimal change disease, IgA nephropathy is an important differential diagnosis to consider. These patients most often present with a palpable, purpuric rash and normal-to-mildly elevated serum IgA levels. Renal involvement in adults is usually manifest by mild proteinuria and hematuria without red cell casts, although heavy proteinuria and more active urine sediment- as was present in this case- can occur. Renal and skin biopsies are necessary to confirm the diagnosis. The prognosis is typically good, with complete remission not uncommon, with or without a short course of steroids. In other cases, stronger immunosuppression may be necessary using prolonged corticosteroids, mycophenolate mofetil, and/or calcineurin inhibitors.

References

Mississippi-Clinical Vignette-Poster Finalist
Nathalie S Malcolm, MD

Title: Bilateral obstructive renal failure caused by nephrolithiasis secondary to post traumatic osteolysis

Authors: Nathalie Malcolm, MD, Pradeep Kumar Vaitla, MD, Prakrati C. Acharya, MD, Ashton Davis, MD

Introduction: Acute renal failure secondary to bilateral obstructive nephrolithiasis is rare. Posttraumatic osteolysis is a recognized phenomenon but never reported to cause obstructive renal failure. Early recognition is important for intervention to prevent irreversible damage.

Case Presentation: 18-year-old Caucasian male with recent admission after motor vehicle collision sustaining left femur and pelvic fracture managed by intramedullary nailing presented with left lower extremity pain, lower abdominal pain, and decreased urine output. Associated symptoms included decreased food intake and non-bilious, non-bloody emesis. Patient was transferred from an outside hospital for concern of soft tissue extravasation at the level of the pubis on CT pelvis scan with contrast. In the absence of prior history of kidney disease, serum creatinine was initially 0.6 mg/dL and rose to 5.15 mg/dL in a 4-day period. Patient denied prolonged use of NSAIDS or apparent nephrotoxins with the exception of a one-time dose of 325 mg Aspirin. No evidence of potential nephrotoxic medications given intra-operatively or post operatively. Urine drug screen was negative. Basic metabolic panel indicated high anion gap metabolic acidosis, calcium (9.3 mg/dL), hyperuricemia (9.7 mg/dL) from traumatic cell injury. CT abdomen demonstrated mild bilateral hydronephrosis with hyperdense foci along the renal papilla bilaterally consistent with Randall’s plaques. Urinalysis revealed pyuria and hematuria while urine culture revealed no growth. Urgent cystoscopy with bilateral retrograde pyelogram was performed with placement of bilateral ureteral stents. Obstructive debris was collected for analysis, which revealed 95% calcium phosphate and 5% calcium oxalate stones. The patient’s urine output and serum creatinine improved briskly after stent placement and renal function returned to baseline in 3 days.

Discussion: Urinary tract obstruction secondary to bilateral renal calculi is rare but can result in a favorable prognosis if reversed early. Nephrolithiasis begins with the formation of Randall’s plaques which are subepithelial calcifications located at the renal papilla (anchors for stone formation). Supersaturated urine can induce crystal nucleation, aggregation, and epitaxy. Post traumatic osteolysis is a known risk factor for hypercalcemia but not a well-recognized cause of nephrolithiasis. In our patient with baseline normal renal function, he was able to tolerate rising serum calcium level efficiently by renal excretion, putting him at risk for bilateral nephrolithiasis secondary to risk factors of volume depletion.

References
Mississippi-Clinical Vignette-Poster Finalist
Madison H Williams, MD

Title: Denosumab Associated Severe Hypocalcemia in a Patient with Chronic Kidney Disease

Authors: Madison H. Williams, MD, Ryan A. Williams, MD, Sohail Abdul Salim, MD, and Natale Sheehan, MD.

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Introduction: Denosumab is a monoclonal antibody against receptor activator of nuclear factor kappa B ligand (RANKL). Denosumab has been shown to reduce the risk of skeletal-related events (SREs), including spinal cord compression, pathologic fracture, and hypercalcemia of malignancy, in patients with bone metastases. Hypocalcemia is a known side effect of denosumab, occurring in an estimated 8-14% of patients. Here we present an asymptomatic patient with severe hypocalcemia who was treated with denosumab one month prior.

Case Presentation: A 70 year old white male with hypertension (HTN), chronic kidney disease (CKD) stage V, and metastatic adenocarcinoma of the lung presented from oncology clinic for severe hypocalcemia. The patient reported fatigue and a poor appetite, but otherwise had no complaints. Physical exam revealed a thin, ill-appearing male with dry, flaky skin and dry mucous membranes. Laboratory findings revealed a serum calcium of 3.6 mg/dL (corrected to 4.6 mg/dL for hypoalbuminemia), an ionized calcium of 0.62 mmol/L, blood urea nitrogen (BUN) of 38 mg/dL, creatinine (Cr) of 4.2 mg/dL, an elevated parathyroid hormone (PTH) of 401.9 pg/mL, and a low serum 25-OH vitamin D of 17 ng/mL. Electrocardiogram (EKG) showed a prolonged corrected QT (QTc) interval of 551 milliseconds. The patient was treated with a total of 8 g of intravenous calcium gluconate, followed by a 10 g continuous infusion of calcium gluconate given over 10 hours. Once his ionized calcium was greater than 1.0 mmol/L, he was transitioned to oral calcium supplementation (calcium carbonate 2500 mg three times daily), along with calcitriol (1 mcg twice daily). His severe hypocalcemia was thought to be secondary to denosumab, of which he received a single 120 mg dose approximately one month prior to presentation, with secondary hyperparathyroidism from CKD also contributing. The patient was discharged on oral calcium carbonate with continued close monitoring of serum calcium at his nursing home.

Discussion: With denosumab use, frequent monitoring of calcium levels and aggressive replacement of calcium and calcitriol is necessary to prevent hypocalcemia. Renal impairment does not affect the pharmacokinetics of denosumab, and therefore, does not necessitate dose adjustment of the drug. However, patients with CKD stages IV and V need to be monitored more closely, as they experience higher rates of hypocalcemia. This case demonstrates the importance of being aware of the adverse side effects of denosumab and monitoring for them regularly, as even patients at risk of life-threatening arrhythmias may be asymptomatic.

References


Missouri-Clinical Vignette-Poster Finalist
Joyce Achenjang, MD

Title: Miss Understood: A Case of Hashimoto’s Encephalopathy

Authors: Joyce Achenjang MD, Hiral Choksi MD

Introduction: Hashimoto’s encephalopathy, also known as steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT), is a rare neurologic disorder characterized by psychiatric and neurologic findings in the setting of thyroiditis. Not well understood, it can be a challenging diagnosis to make, especially given its non-specific presentation. As such, it can be hard to distinguish from other inflammatory disorders, thus early recognition and diagnosis are essential for patient survival.

Case Presentation: A 75-year-old African-American female with a past medical history of hypothyroidism, hypertension, and Alzheimer’s dementia presented to the emergency room with altered mental status and concern for possible stroke; family also reported new seizure-like activity. Vitals on admission were significant for severe hypertension however, she was afebrile. Physical exam was limited due to altered mental status but revealed conjugate gaze, equal and reactive pupils, no murmurs, diminished strength in the left upper extremity, benign thyroid exam, and diffusely warm extremities. Pertinent labs revealed elevated levels of white blood cell count, creatinine kinase, creatinine, thyroid stimulating hormone (TSH), and anti-thyroid peroxides antibody (anti-TPO). Following a head commuted tomography, which was negative for an acute process, brain magnetic resonance imaging was completed and showed left sided enhancement, negative for acute stroke, but concerning for an underlying inflammatory process. Further work up, including two lumbar punctures, for rheumatologic and infectious etiologies remained unrevealing. The patient continued to seize during the early part of her hospitalization, in addition to experience aggressiveness and agitation. With elevated anti-TPO and TSH levels, she was resumed on levothyroxine and started on a high dose of steroids with concern for hashimoto’s encephalopathy due to thyroiditis in the setting of new onset neurologic and psychologic changes. Throughout the remainder of her hospitalization, the patient showed significant signs of mental status improvement, eventually returning to baseline, and remained seizure free. Her anti-TPO levels continued to decline and she had resolution of her agitated and aggressive behavior as well. Following continued improvement, she was discharged home with a steroid taper and gradual increase in her levothyroxine dose.

Discussion: This case provides an example of a manifestation for an uncommon disease that is not fully understood. Other diseases, such as vasculitis and myxedema coma, remain a possibility when making this diagnosis given the patient’s presenting symptoms and significant response to steroids. However, imaging and lab findings assisted in ruling out an alternative etiology; following an involved diagnostic work up for altered mental status in the setting of thyroiditis, it is highly suggestive of Hashimoto’s encephalopathy. Treatment guidance thus hinges on patient response to steroids and to an extent, an improvement in laboratory findings over time.
Missouri-Clinical Vignette-Poster Finalist
Haytham A Allaham, MD

Title: Right Sided Hydrothorax: A Peritoneal Dialysis Dilemma

Authors: Dania Hudhud MD, Haytham Allaham MD, Danielle Severns, William Salzer MD, Internal Medicine Department, University of Missouri, Columbia

Introduction: Right sided hydrothorax due to a pleuroperitoneal leak is considered a rare complication that tends to develop 4-8 weeks after initiation of peritoneal dialysis in patients with end stage renal disease (ESRD).[1-2] A high index of suspicion is the key to establish an early diagnosis. Several diagnostic tools are present to confirm the presence of a diaphragmatic defect connecting both compartments. This case report highlights the challenges regarding the diagnosis and therapeutic approach of such a complication.

Case Presentation: We present a 23 year old female patient with progressively worsening dyspnea of 2 days duration. She denied any history of lower extremity edema, orthopnea, cough, or fever. Past medical history was significant for ESRD. A peritoneal dialysis (PD) catheter was placed 8 weeks prior to her presentation. Vital signs were significant for severe hypoxemia. Physical exam was remarkable for right sided basal crackles with no other signs of fluid overload. Laboratory Results: revealed normal levels of white cell count and brain natriuretic peptide. A chest X-ray demonstrated the presence of a large right-sided pleural effusion. Right sided thoracentesis was performed with subsequent pleural fluid analysis concerning for a possible pleuroperitoneal leak (fluid protein: 0.2 g/dl, fluid LDH: 16 units/L, fluid pH 7.64, fluid glucose: 240 mg/dl with a serum glucose of 87 mg/dl). CT peritoneography confirmed the diagnosis as contrast material leaked through the inferior vena cava (IVC) diaphragmatic foramen and into the right pleural space. Surgical intervention was deferred in light of the close proximity of the defect to the IVC. The patient was transitioned to hemodialysis for temporary cessation of peritoneal dialysis.

Discussion: The hallmark of hydrothorax secondary to a pleuroperitoneal defect is the development of hypoxemia and a unilateral pleural effusion in a euvelomic PD patient. CT peritoneography is the imaging modality of choice for the diagnosis, which provides details regarding the size and location of the defect.[3] The management of pleuroperitoneal leak is still considered to be challenging despite the presence of several well-established therapeutic techniques. The challenge stems from the lack of randomized controlled clinical trials comparing the different management strategies. Interruption of PD with temporary transfer to hemodialysis (HD) for a period of two to six weeks has proven to be effective in almost half of the patients suffering from such a complication. Surgical interventions for management of pleuroperitoneal leaks include either administering chemical pleurodesis via an intercostal chest tube or performing video-assisted thoracoscopic interventions, such as direct pleurodesis and surgical diaphragmatic repair. However, surgery is not considered the first-line treatment option for PD patients with pleuroperitoneal leak. Surgical interventions are usually reserved for young patients with persistent diaphragmatic defect despite trials of temporary dialysis interruption. In such patients, the risk of surgery should always be weighed against transitioning to long term HD.[4]

References

Missouri-Clinical Vignette-Poster Finalist
Anoushiravan Hakim, MD

Title: Posterior Orbit Compressive Mass: A unique presentation of Sweet’s syndrome.

Authors: A. Hakim, MD (Associate), F. Hoque, MD (Associate), J. Paul, MD (Fellow)

Introduction: Sweet’s syndrome is a rare inflammatory disease, characterized by sudden onset painful and erythematous plaques, fever and neutrophilia. It can be associated with extra cutaneous involvement. We report a case of Sweet’s syndrome presenting as a posterior orbital compressive mass. To our knowledge, there has been only one similar case report by Koay et al in 2013. They presented a case of compressive optic neuropathy and blurry vision.

Case Presentation: A 31-Year old Caucasian female with a history of skin biopsy proven Sweet’s syndrome presented with rapid worsening of left-sided periorbital redness, swelling, proptosis, and binocular diplopia. No fever or chills. On physical exam, extra ocular movements were intact. Pupils were equal, round and reactive to light and accommodation. Visual acuity and fields were normal.

Lab tests were negative for ANA, ds DNA, SSA/SSB, SCL 70, Jo 1, Ribosomal P, CCP antibodies. Normal CPK /RF levels/sed rate/CRP. Normal IgG subclass levels.

MRI of brain and orbit demonstrated 27x12x8.2 mm mass-like enhancement in the superior part of the left orbit involving the left superior rectus muscle. This exerted mass effect on the infraorbital segment of the left optic nerve. Infectious work up was negative. CT of the neck, chest, abdomen, and pelvis were negative for lymphadenopathy or similar lesions.

Patient was diagnosed with compressive neutrophilic lesion, a rare complication of Sweet’s syndrome. Biopsy was not performed because of difficulty with access to the lesion.

Treatment was started with high dose of oral prednisone at 80 mg daily and intraocular Kenalog. Patient’s ophthalmic symptoms improved. Treatment continued with Methotrexate as steroid sparing agent along with Hydroxychloroquine and colchicine. 6 months later, patient relapsed and had difficulty tapering prednisone below 20 mg. She has been treated with rituximab with good response.

Repeat MRI of brain showed regression of the orbital mass.

Discussion: Posterior orbital lesion is a unique manifestation of Sweet’s syndrome. Patient can present with diplopia and proptosis (secondary to extraocular muscles involvement) or blurry vision (secondary to compressive optic neuropathy). The first step in management is to exclude other diagnoses such as lymphoma. Corticosteroid is first line treatment for Sweet’s syndrome. Local and small lesions can be treated with topical or intralesional corticosteroids. In our patient, clinical finding, lab Results; and quick improvement after steroid treatment further supported the diagnosis of orbital lesion secondary to Sweet’s syndrome. Recent data have shown efficacy of biologic agents such as Anakinra, Rituximab in treatment of recurrent or refractory Sweet’s syndrome.

References


Title: A Deadly Tunnel – Not to Be Mistaken

Authors: Farzana Hoque, MD, Anoushiravan Hakim, MD, Anthony Pearson, MD

Introduction: This is a unique case of left atrial-esophageal fistula, which developed after ablation for medically refractory atrial fibrillation. The incidence of left atrial-esophageal fistula is between 0.03% to 1.5% but mortality rates are high, ranging from 67% to 100%.

Case Presentation: A 78-year-old white man with a past medical history of paroxysmal atrial fibrillation status post 3 radiofrequency catheter ablations (the most recently 1 month prior to this admission) presented with hematemesis, fever (temperature, 39.4°C) and confusion. Labs were significant for WBC of 5.3 with 21% bands, lactic acid 3.7. CT head without contrast showed no acute abnormalities. Emergency esophagastroduodenoscopy (EGD) revealed a 1 cm superficial ulcer in the mid-esophagus. Unfortunately, he developed left hemiparesis 2 hours after the upper endoscopy. MRI of the brain demonstrated multiple bilateral embolic infarcts in both cerebral and cerebellar hemispheres. Meanwhile, a blood culture was positive for Streptococcus viridans. Infective endocarditis was high among differentials. Subsequently, a transesophageal echo (TEE) was performed which did not show vegetations or valve regurgitation. Surprisingly, it revealed multiple spontaneous air bubbles inside the left atrium, an unusual finding highly suggestive of left atrial-esophageal fistula. After that, he was sent immediately for thoracotomy where the presence of a fistula was confirmed, and he underwent repair of the esophagus and posterior wall of the left atrium. He survived but he still has minor neurologic deficits at the left side.

Discussion: Left atrial-esophageal fistula is the 2nd most common cause of mortality following atrial fibrillation ablation as it can cause septic or air emboli to the brain, generalized sepsis or massive gastrointestinal bleeding. An atrial-esophageal fistula should be strongly considered in any patient with recent left atrial ablation who presents with fever, neurological symptoms, chest discomfort, or sepsis. This potential lethal complication can easily be misdiagnosed as infective endocarditis, which can lead to performance of a transesophageal echo (TEE). However, if the fistula is suspected, esophageal manipulation with TEE, EGD, or NG tube placement is relatively contraindicated because such procedures can cause cerebral air or food embolism. Chest CT with intravenous contrast is the preferred diagnostic test to reveal an air pocket or fistula in the mediastinum.

Conclusion: This case emphasizes the importance of early identification of atrial-esophageal fistula as prognosis strongly depends on the time interval between the onset of symptoms and diagnosis. Immediate surgical intervention is paramount to the reduction of mortality.

References

Title: Persistent hypokalemia of unknown etiology.

Authors: Christopher Dietrich, DO and Margaret Beliveau-Ficalora, MD

Introduction: Hypokalemia is a not uncommon presentation in both the inpatient and outpatient setting. It is present in up to 21% of patients presenting to the hospital, and in 2-3% of patients presenting to their primary care physician. Often offending medications or other causes can be found in the patient’s history, but occasionally further evaluation is necessary to determine the underlying cause so it may be adequately treated.

Case Presentation: A 32 yo F presented to her gynecologist for tubal ligation. The patient is G2P2, and had both children via vaginal deliveries. The pregnancies, deliveries and postpartum courses were normal.

She underwent preoperative evaluation and was noted to have a significant hypokalemia with a potassium of 2.6. She was asymptomatic. On historical review of her records, she had not had her potassium checked previously during her pregnancies.

She was taking no prescription medications, but was taking several herbal supplements. These were stopped, but her potassium remained low. Her BMI was 19, and a diuretic screen was done on her urine to rule out abuse for weight loss purposes, and this was unremarkable. She trialed potassium supplementation, but upon stopping supplementation her potassium rapidly dropped to 2.7.

Additional laboratory evaluation was undertaken, showing a spot urine calcium of 15, spot urine chloride of 229, spot urine creatinine of 307, spot urine potassium of 60 and a spot urine sodium of 204. This was in the setting of her potassium being 2.6 on serum laboratory evaluation.

Given her inappropriately high urine potassium coupled with her low urine calcium it is suspected she has Gitelman’s syndrome. She has been maintained on potassium supplementation, and has had no additional findings of hypokalemia.

Discussion: Hypokalemia is commonly encountered in medicine. Often the culprit is a medication with diuretics, laxatives, antibiotics and many others. Multiple other causes, including surreptitious vomiting, renal tubular acidosis, salt wasting nephropathies, dialysis and plasmapheresis may all contribute as well. An algorithmic approach to determining the underlying etiology helps avoid unnecessary testing and cost.

If medications are thought not to be contributing, a thorough history for gastrointestinal losses, including self-induced vomiting and diarrhea needs to be obtained. Obscure causes, such as licorice consumption, herbal remedies and others also needs to be evaluated in history.

Urine testing for calcium, chloride, creatinine, potassium and sodium will help guide diagnosis. Here the patient was found to have inappropriately high urine potassium with a low urine chloride, and in combination with her other laboratory findings and history was suggestive of Gitelman’s syndrome. Bartter’s syndrome is also on the differential, but this typically does not cause a low urine calcium. The treatment is lifelong potassium supplementation. For this poster, I have created a novel algorithm for hypokalemia evaluation.
Nebraska-Clinical Vignette-Poster Finalist
Daniel Ermann, MD

Title: Primary Mediastinal Large B-Cell Lymphoma Presenting with Dyspnea and Superior Vena Cava Syndrome

Authors: Daniel Arthur Ermann MD, Creighton University
Victoria Vardell Noble, Creighton University
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Mahmoud Abu Hazeem MBBS, Creighton University

Introduction: Primary Mediastinal Large B-cell Lymphoma (PMBCL) is a distinct, and uncommon, subtype of Diffuse Large B-cell Lymphoma (DLBCL), accounting for approximately 2% of patients with Non-Hodgkin Lymphoma (NHL).

Case Presentation: A previously healthy 21 year old female presented with fatigue and progressive shortness of breath for 3 months with a several week history orthopnea and swelling in her right upper arm and lower face. On presentation she had a normal WBC count and an elevated LDH at 935 (Normal 84-246). A CT scan revealed a large heterogeneous lobular soft tissue mass throughout the superior anterior mediastinum that measured 14.1 x 9.4 x 11.5 cm, with partial encasement of the SVC. Additional findings included bilateral hilar, pericardial, and supraclavicular adenopathy, as well as bilateral pleural effusion, atelectasis, and splenomegaly.

Biopsy demonstrated large lymphoid cells with characteristic centroblastic morphology amid fibroblastic proliferation with loose fibrosis. Immunohistochemistry showed positivity in CD19, CD20, PAX5, CD30 (weak/variable), with subset positivity in CD23. Negativity was shown in CD10, CD15, and surface/cytoplasm light chain.

Discussion: PMBCL is an uncommon subtype of NHL that is most commonly identified in females in the 3rd to 4th decade of life. It commonly presents as a fast-growing and aggressive tumor of the anterior mediastinum with symptoms related to local invasion or compression due to bulky disease. SVC syndrome may be present in up to half of patients with this disease, and was the clinical sign of concern in our patient. LDH is found to be elevated in 70-80 % of cases, and may-often be the only laboratory abnormality. Our patient presented with a large bulky mass, described as a tumor greater than 10cm in its largest diameter, which may be present in 75% of cases. Constitutional symptoms are relatively rare, and present in less than 20% of cases. To achieve a correct diagnosis careful consideration must be made regarding clinical presentation, tumor cell markers, and cellular morphology. The disease can present very similarly to classical Hodgkin Lymphoma or conventional DLBCL with secondary mediastinal involvement. Confined mediastinal involvement, lack of surface immunoglobulin expression, and positivity for pan-B cell markers (including CD19, CD20, and CD23) are distinguishing factors for PMBCL. Neoplastic cells are likely derived from thymic medullary B-cell origin, and are seen as large cells with pale abundant cytoplasm and heterogenous nuclei within characteristic fibrotic collagen bands. Immunohistochemistry for CD30 is weakly positive and CD10/CD15 is negative. These traits define PMBCL as a distinct clinical and histopathological entity. Due to the rare nature of the disease, the optimal chemotherapeutic treatment remains unknown. However, recent clinical trials using chemoimmunotherapy have demonstrated a favorable prognosis.
Title: Double bolus tissue plasminogen activator (t-PA) therapy during cardiopulmonary resuscitation (CPR) for cardiac arrest due to massive pulmonary embolism (PE) guided by focused bedside echocardiography

Authors: Hafiz B. Mahboob, MD, Bruce W. Denney, MD

Introduction: Massive pulmonary embolism (PE) frequently leads to cardiac arrest with an extremely high mortality rate. Available randomized trials did not show survival benefit from thrombolytics, therefore traditional advanced cardiac life support (ACLS) has been the mainstay of treatment. Thrombolytics have been used successfully during resuscitation for PE related cardiac arrest in multiple case reports and in retrospective studies, mostly late into the cardiopulmonary resuscitation (CPR) after initial unresponsiveness to traditional ACLS. Therefore, recent resuscitation guidelines recommend using t-PA for PE related cardiac arrest but do not offer a standardized treatment regimen. Most consistently applied approach in the literature is intravenous (IV) bolus of 50mg t-PA early during CPR, advised initially by British Thoracic Society. There is no consensus on the subsequent dosing and approach. We present an approach of double bolus of t-PA (two 50 mg IV boluses of t-PA were administered 15 minutes apart during CPR) guided by bedside echocardiogram showing persistent right ventricular (RV) dysfunction.

Case Presentation: A 56 year old, previously healthy Caucasian female presented to emergency room with acute severe shortness of breath and pleuritic chest pain. She was emergently intubated due to persistent hypoxic respiratory failure. Shortly after intubation, she underwent cardiac arrest with pulseless electrical activity (PEA). Emergent bedside echocardiogram during CPR showed a severely dilated RV with normal left ventricular (LV) function, and positive McConnel sign, which was likely due to a massive PE. First bolus of t-PA was given during the first round of CPR (5 minutes into the code) with ongoing chest compressions. She had return of spontaneous circulation (ROSC) one minute following the t-PA bolus administration. However, she went into cardiac arrest again, approximately at 15 minutes into the code with recurrent PEA. Echocardiogram was still showing persistent evidence of RV dysfunction with normal LV function. Therefore, a 2nd bolus of 50 mg t-PA was given approximately 20 minutes into the code. She received double bolus t-PA therapy guided by bedside echocardiogram, with ongoing CPR with favorable clinical outcome without any neurologic sequelae or any bleeding complications. On 3 months follow up, she had complete normalization of RV function without any residual pulmonary hypertension.

Discussion: There is no standard treatment regimen for t-PA administration for PE related cardiac arrest in current literature or resuscitation guidelines. This case demonstrates the benefit of utilizing bedside focused echocardiography to guide administration of a second bolus of t-PA in cardiac arrest due to massive PE, if there is a) evidence of persistent RV dysfunction in cardiac arrest despite achieving ROSC after initial bolus, b) recurrent arrest despite achieving ROSC, c) failure to achieve ROSC after first bolus

Future multi-center trials are warranted to help establish guidelines for t-PA use in cardiac arrest to maximize safety and efficacy.
Nevada-Clinical Vignette-Poster Finalist
Arhama A. Malik

Title: A Fatal Case of Donor Transmitted Tularemia in a Renal Transplant Recipient

Authors: Arhama A. Malik, Pallavi Satuluri , Wen Yuan Yu, L. Medina Garcia, M.D., S. Asad, M.D

Introduction: Infection remains an important cause of mortality and morbidity in solid organ transplant recipients with the majority being nosocomial. However, patients may develop a donor-transmitted infection that can be challenging to diagnose and manage given its diverse nature as well as the intense immunosuppression during this period. We report a fatal case of donor transmitted Tularemia in a renal transplant recipient.

Case Presentation: A 46-year-old gentleman with a history of ESRD due to Ig A nephropathy and FSGS underwent a deceased donor renal transplantation. His pretransplant workup was positive for CMV and EBV serology. Post transplantation, he received Thymoglobulin for induction and was started on Prednisone, Tacrolimus and Mycophenolate for maintenance. He was started on antimicrobial prophylaxis with Trimethoprim/sulfamethoxazole, valganciclovir and fluconazole. Postoperatively he required hemodialysis for renal dysfunction. On POD# 4, he had a febrile episode without any localizing signs of infection. Blood and urine cultures as well as chest imaging were unremarkable. A renal ultrasound of the graft demonstrated normal parenchyma and vasculature. Over the next 12 hours he deteriorated clinically with temperature up to 102.9*F, tachycardia, hypotension requiring vasopressors and was intubated for respiratory distress and hypoxia. Antibiotics were empirically broadened to vancomycin, meropenem and micafungin. The patient expired that evening due to septic shock, DIC and multiorgan failure. Subsequently, his blood cultures drawn prior to his demise revealed a gram-negative coccobacillus, which was later identified by PCR as Francisella tularensis. Patient’s autopsy demonstrated positive CSF as well as the transplanted kidney specimen for Francisella tularensis. CDC and Organ Procurement Agency were informed immediately. Other transplant recipient health care teams were notified and started on treatment with doxycycline. The recipient of the other kidney also had positive blood cultures for Francisella tularenis. The heart transplant recipient never developed an infection as he was on doxycycline for several weeks due to a LVAD infection. Final diagnosis was confirmed as donor transplant related Tularemia.

The donor tissue that was available for CDC tested positive for Francisella tularensis. Donor was from Indian reservation in northeast Nevada and experienced a near drowning episode followed by febrile episodes and cough two weeks prior to his demise from cerebral hemorrhage, which was attributed to consumptive coagulopathy from alcoholic cirrhosis.

Discussion: The majority of infections in the early post transplant period are health care associated or surgical site complications. Rarely, patients may present with a donor-transmitted infection. These infections can progress rapidly secondary to intense immunosuppression. The diagnosis requires a high index of suspicion as well as collaboration with the OPA. Although there are few cases reported in literature of transplant recipients who have developed Tularemia, to the best of our knowledge, this is the first case of donor transmitted tularemia.
Nevada-Clinical Vignette-Poster Finalist
Pallavi Satuluri, MBBS

Title: A Unique Case Report of a Non Cardiac Tumor Metastasis into the Primary Cardiac Tumor.

Authors: Pallavi Satuluri- UNLV School of Medicine, Las Vegas, Nevada, Kasaiah Makam- Newark Beth Israel Medical Center, Newark, New Jersey, Erik Marshall- Christiana Care Health Services, Newark, DE, Anand Kenia- Christiana Care Health Services, Newark, DE

Introduction: Cardiac tumors are extremely rare. We report a unique case of a coexisting atrial myxoma and micro metastasis of breast carcinoma into the myxoma.

Case Presentation: A 55-year-old female presented with worsening shortness of breath with minimal exertion and significant lower back pain. Her vital signs were within normal limits on admission. On initial evaluation, lumbo-sacral X-ray demonstrated lytic lesions and chest computed tomography angiography demonstrated a mass in her left atrium. Later on that day, she went into acute pulmonary edema requiring intubation. 12 lead EKG showed sinus tachycardia. CXR findings were suggestive of pulmonary edema. Subsequently, transthoracic echocardiography demonstrated classic appearing myxoma with stalk attached to inter-atrial septum.

She underwent surgical excision of left atrial mass (presumable myxoma). Frozen section of the atrial mass demonstrated gelatinous structure with foci of hemorrhage characteristic of myxoma. Electron microscopy of specimens had typical histology of myxoma with large areas of extracellular myxomatous matrix and thin delicate stellate cells embedded within it confirming the diagnosis of myxoma. Further analysis of biopsy specimen showed small foci of epithelial cells with malignant nuclei growing in a nodular fashion. The tumor cells were positive for cytokeratin AE1/AE3, GATA 3, estrogen receptor and HER-2/neu. This was consistent with micro metastasis of ductal carcinoma of the breast into the myxoma.

Later, biopsy of lytic lesions of the right iliac bone also demonstrated positive staining for keratin AE1/AE3 and an immunohistochemical stain for GATA-3 confirmed metastatic breast cancer.

Patient was initiated on treatment for metastatic breast cancer per oncology recommendations.

Discussion: Primary cardiac tumors are extremely rare and among them atrial myxoma is the most common primary tumor. It commonly occurs in the left atrium. Metastasis of non-cardiac tumors into the cardiac myxoma has never been reported before and micro metastasis into a myxoma is considered to be a stage 4 equivalent of breast carcinoma even in the absence of other lesions. This is the first ever-reported case of a non-cardiac tumor metastasis into the left atrial myxoma.
Title: The Perplexity of the Autoimmune Disease: There is More than Meets the Eye

Authors: Jill Sharma, MD; Internal Medicine Resident, UNLV School of Medicine, Nitasha Khullar, MD; Internal Medicine Resident, UNLV School of Medicine, Dodji Modjinou, MD, FACR; Assistant Professor of Medicine/Rheumatology, UNLV School of Medicine

Introduction: Multiple autoimmune syndrome (MAS) is a manifestation of three or more autoimmune diseases in one patient, as defined by Humbert and Dupond in 1988. Up to 25% of patients with one autoimmune disease may have another concurrent diagnosis. We report the case of a patient who had such an occurrence, with rapid onset of symptoms noted during a single hospital stay.

Case Presentation: A 65-year-old Hispanic female with a history of asthma, arthritis not previously specified otherwise, hypothyroidism and PTSD presented to the hospital with persistent cough, lower extremity maculopapular rash, associated with edema for two months. She reported having occasional oral bleeding, which she thought to be due to a chipped tooth causing lacerations of her tongue. Given the non-blanching maculopapular lesions noted upon presentation, an autoimmune workup was pursued. Her initial labs were unremarkable except 3+ blood with 50-99 RBCs on high-power field. Within the next several days, ANA was found to be markedly positive at 1:1280 and she subsequently developed respiratory distress initially thought to be due to asthma exacerbation. However, further evaluation showed ground-glass opacities on CT chest. Pulmonary function test confirmed restrictive lung disease; shortness of breath and cardiopulmonary symptoms improved with glucocorticoids. Yet, she later developed dry throat and was unable to produce tears upon crying. On the sixth day of her admission, her teeth began to decay. Further inpatient labs showed positive +P-ANCA, Anti-MPO, Anti-proteinase 3, anti-CCP, rheumatoid factor, SSA and SSB antibodies. Ultimately, the patient was found to have granulomatosis with polyangiitis with ANCA associated active and crescentic glomerulonephritis, in the setting of mesangial lupus nephritis based on kidney biopsy. Her physical exam and clinical presentation also were suggestive of seropositive rheumatoid arthritis, and likely with secondary Sjogren’s syndrome. The patient had remarkable clinical improvement on cyclophosphamide and tapering dose of glucocorticoids.

Discussion: While we may not commonly encounter autoimmune conditions as new diagnoses during a routine hospital admission, it is crucial to be aware of these diagnoses and their implications. Multiple factors of genetics, infections and immunology have been implicated in the development of MAS, however the pathogenesis is not yet established. MAS is subdivided into three categories based on observation of co-occurrence of autoimmune conditions, one of which is usually a dermatological manifestation. Although case reports have demonstrated the presence of this syndrome, a skin finding secondary to a vasculitis has not been well-reported. It is imperative to know that a patient with one autoimmune condition has increased propensity to develop others. A full rheumatologic evaluation should be entertained in these settings.

References


Nevada-Clinical Vignette-Poster Finalist
Mokshya Sharma, MD

Title: Pembrozulimab Induced Collagenous Colitis

Authors: Mokshya Sharma 1, Santhosh Ambika 2

Introduction: Immune modulating therapy that targets PD1 pathway such as pembrozulimab is at the frontier of breakthrough cancer treatment. As these immune modulating therapies become more common, it will be important to have treatment guidelines to monitor and treat potential side effects with early intervention. Although pembrozulimab is associated with immune mediated colitis, to the best of our knowledge, this is the second reported case of pembrozulimab induced collagenous colitis and first case with development of side effects early on in the treatment cycle.

Case Presentation: We present a case of collagenous colitis that initially presented as a grade 3 diarrhea in a 78 year old female after 2 cycles of pembrozulimab. Our patient had poorly differentiated metastatic small cell cancer of the lung with 90% PD-L1 expression, and was initiated on pembrozulimab therapy after resection of local mass and radiation therapy. After development of diarrhea, the patient was initially treated with imodium and prednisone for suspicion of immune mediated colitis. However, as she was not improving, a colonoscopy was performed which showed friable mucosa suspicious for collagenous colitis, later confirmed with pathology. She was initiated on treatment with budesonide 9mg/day and cholestyramine 8g/day which led to resolution of her diarrhea. To the best of our knowledge, this is the second reported case of pembrozulimab induced collagenous colitis and first case with development of side effects early on in the treatment cycle (cycle 2 vs. cycle 14 in the previous case report).

Discussion: As pembrozulimab gets approved for use in a variation of different cancers, with recent approval in September for gastric cancer as per keynote study 0592, it will be important to monitor for its side effects and have effective surveillance and treatment strategies moving forward in the future. Usually, PD1 inhibitor associated colitis is thought to be immune mediated and prednisone is initiated which controls the symptoms. However, if not improving, low threshold for obtaining a colonoscopy to rule out collagenous colitis as treatment regimen would vary.
Title: A Case Of Plasmablastic Lymphoma: A Rare Disease Entity

Authors: Nino Aleksidze, MD; Emmanuel Adomako, MD; Heidi-Anne Hanson; Tanganyika Barnes, MD; Maxwell Janosky, MD;

Introduction: Plasmablastic lymphoma (PBL) is a rare and aggressive variant of non-Hodgkin’s lymphoma (NHL), which is strongly associated with human immunodeficiency virus (HIV) and is now described as a new disease entity in the 4th WHO classification. We report an unusual case of HIV-associated PBL with initial involvement of liver and multiple vertebrae.

Case Presentation: 57 y/o Male presented to ED complaining of back pain for past 3 weeks. Past medical history was significant for HIV and renal cell cancer treated with resection 16 years ago. Vital signs were significant for BP 190/86 and HR 110. Physical exam was notable for conjunctival pallor and scleral icterus. Laboratory data revealed hemoglobin 6.4g/dl, MCV 107.8, platelet count 68000/ul, WBC count 7.79 k/ul, reticulocyte count 1.4%, LDH 10,000 U/L, fibrinogen 744mg/dL, uric acid 13.3mg/dL, haptoglobin 207mg/dL. Ionized calcium 1.59mmol/L, ALP 422U/L, AST 125U/L, total Bilirubin 3.2mg/dl, urine random protein 30mg/dl, plasma Cr 2.8, BUN 47mg/dl; SPEP and UPEP showed no evidence of monoclonal spike. AFP was 6.3ng/mL.

MRI spine revealed diffuse marrow replacement with posterior epidural involvement concerning for diffuse metastatic disease. Hypodense hepatic nodules were seen on CT scan consistent with metastatic disease. Patient underwent bone marrow biopsy: Immunostaining demonstrated strongly positive CD138 cells, which were negative for CD20, CD3 and CD34. These finding were consistent with Plasmablastic Lymphoma.

Patient underwent chemotherapy with attenuated EPOCH regimen and eventually achieved resolution of back pain, as well as normalization of kidney and liver function.

Discussion: Diagnosis of PBL remains challenging because of its morphologic and immunophenotypic overlap with plasma cell neoplasms and lack of expression of pan-B-cell antigens. Median age at diagnosis of PBL is 50 years with strong male predominance. The most common site of involvement is the oral cavity, followed by the gastrointestinal tract, lymph nodes and the skin. Because of its aggressive and relapsing clinical course, it poses challenges to therapy, complicated by high rates of disease progression and fatality. Moreover, given rare incidence of PBL, no standard of care has yet been established.

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New Jersey-Clinical Vignette-Poster Finalist
Sevil Aliyeva, MD

Title: Unrecognized thyrotoxic periodic paralysis in hyperthyroidism.

Authors: Sevil Aliyeva, MD, Natalia Plotskaya, MD, Bushra Saleem, MD, Nooraine Mazhar, MD, Daniel Goldsmith, MD, Sunil Thomas, MD

Introduction: Thyrotoxic periodic paralysis (TPP) with severe hypokalemia leads to progressive muscle weakness and life threatening cardiovascular complications when diagnosis and treatment are delayed. The muscle weakness and tachyarrhythmia result from markedly reduced serum levels of potassium. Potassium is not actually lost from the body, but increased Na+K+-ATPase activity leads to shift of potassium into tissues, depleting circulating levels in hyperthyroidism. In other types of potassium derangements, the acid-base balance is usually disturbed, with metabolic alkalosis and metabolic acidosis often present.

Case Presentation: A 44 year old Hispanic male with history significant for radiculopathy and intermittent proximal muscle weakness for 18 months, presented with substernal chest pain associated with intermittent dyspnea, nausea and diaphoresis. He reported 50 pounds weight loss over one year associated with soft bowel movements. He had no eye signs or symptoms. His sister had a history of thyroid disease, unsure of the diagnosis. Physical examination revealed tachycardia, bilateral weakness in all four extremities, inability to raise legs with intact sensation and no urinary or stool incontinence.

ECG shown sinus tachycardia at 110 bpm, deep ST segment depressions in inferolateral leads, ST elevation in aVR and aVL. Subsequent ECG demonstrated atrial fibrillation with rate of 160 bpm and severe ST depression in anterior leads with episodes of non-sustained ventricular tachycardia. During transfer to cardiac catheterization laboratory, critically low potassium of 1.2 mmol/l was reported. Total of 60 mEq potassium chloride was infused over 3 hours, chest pain subsided and code PCI was cancelled. Four hours later his potassium level was 6.9 mmol/l and next day 4.5 mmol/l. Troponins were gradually decreasing from 0.481 to 0.109 ng/ml. The thyroid panel showed TSH less than 0.015 uIU/ml, Free thyroxin 4.16 ng/dl, Total triiodothyronine 321 ng/dl. Treatment with propranolol and methimazole was started and his symptoms were gradually improving.

Discussion: The protean features of hyperthyroidism together with the rarity of this condition makes thyrotoxic periodic paralysis a difficult condition to diagnose at presentation.

In the acute phase of the attack, administration of potassium will quickly restore muscle strength and prevent complications. However, caution is advised as the total amount of potassium in the body is not decreased, and rebound hyperkalemia can occur, which happened in our patient. Slow infusions of potassium chloride are recommended while other treatment is commenced for thyrotoxicosis.

Muscle weakness involves the lower limbs and girdle muscles and then upper limbs. Sensory function, bowel and bladder function are not affected. This occurs only in the presence of hyperthyroidism and is abolished when thyroid hormone levels are normalized. The attacks of weakness are similar to those of familial hypokalemic periodic paralysis except for the presence of hyperthyroidism.
Title: Acute renal failure from hypercalcemia as a presenting feature of sarcoidosis; an overview of calcium metabolism and tissue histology in sarcoidosis

Authors: Ali Aziz MD, Saint Barnabas Medical Center, Rahul Thampi MD, Saint Barnabas Medical Center

Introduction: We discuss a case of sarcoidosis who presented with hypercalcemia and hypercalciuria resulting in tubular injury and acute renal failure. Our patient had no other manifestations of sarcoidosis and had an extensive diagnostic work up to rule out other pathologies. A high clinical suspicion of sarcoidosis in similar presentations can prevent unnecessary diagnostic testing. We discuss the metabolism of calcium in sarcoidosis as evidenced in this case by hypercalcemia, hypercalciuria, suppressed Parathyroid hormone, elevated Vitamin D levels, along with lymph node and renal histological findings in sarcoidosis.

Case Presentation: Our patient is a 55 year old male with past medical history of hypertension and diabetes mellitus type 2, who was admitted with calcium of 13.7 mg/dl and creatinine 3 times his baseline levels. He endorsed polyuria for several months prior to presentation but was otherwise asymptomatic. Parathyroid hormone was suppressed to 12 pg/ml. Vitamin D level was 74 pg/ml. Urine calcium was 420 mg/24 hour. Parathyroid hormone related protein was low. Initially patient was treated with IV fluids but calcium and renal function showed only minimal improvement. His tumor markers, autoimmune work up, protein electrophoresis and flow cytometry were negative. Imaging showed extensive mediastinal and hilar lymphadenopathy. Initial subcarinal lymph node biopsy was negative for any pathology. His kidney biopsy showed acute tubular injury, severe interstitial fibrosis, moderate arteriosclerosis, diabetic glomerular disease and scarring. Later patient had mediastinal lymph node biopsy that was positive for granulomatous changes consistent with sarcoidosis. Patient was started on glucocorticoids outpatient. His renal function subsequently improved likely due to recovery from acute tubular necrosis and development of normocalcemia.

Discussion: The most common causes of hypercalcemia are hyperparathyroidism and malignancy. Renal disease in sarcoidosis can present as acute tubular necrosis from hypercalcemia, nephrocalcinosis, nephrolithiasis, acute interstitial nephritis with granuloma formation, glomerular disease or obstructive uropathy. The most common kidney lesion in sarcoidosis is non-caseating granulomatous interstitial nephritis but the most common cause of significant renal disease in sarcoidosis is hypercalcemia and hypercalciuria. Patients may have polyuria resulting from hypercalcemia and hypercalciuria resulting in reduced responsiveness to antidiuretic hormone. Treatment of hypercalcemia in sarcoidosis is by reducing dietary calcium intake and administering steroids. In our patient, kidney disease was consistent with hypertensive and diabetic nephropathy as well as acute tubular injury from hypercalcemia. His acute kidney injury resolved with normalization of calcium levels after administration of steroids. Sarcoidosis without typical pulmonary manifestations can be a diagnostic challenge but should always be kept in mind when dealing with non-parathyroid driven hypercalcemia. A high clinical suspicion of sarcoidosis in similar presentations can prevent unnecessary diagnostic testing.
New Jersey-Clinical Vignette-Poster Finalist
Amad Choudhry, MBBS MD

Title: Persistent Diarrhea with Theophylline Toxicity, looking in all the wrong places

Authors: Amad Choudhry MD; Douglas H. Weinstein MD, MD; Zia Durrani MD, Daniel Goldsmith MD

Introduction: Theophylline, although not a first line agent is very effective in treating COPD through its mechanism of bronchodilation and anti-inflammatory effect. However, given its narrow therapeutic index, and major side effect profile, the drug has fallen out of favor in treating COPD and asthma. Serious toxicities may occur despite regular daily dosing within the normal range for weight. Although nausea and vomiting are common even with therapeutic levels of theophylline, osmotic diarrhea has been rarely reported.

Case Presentation: An 87-year-old female with past medical history of chronic obstructive pulmonary disease, diabetes mellitus, hypertension, who comes to the hospital with a 2-week history of nausea, vomiting and frequent loose stools. Patient was seen by Gastroenterology outpatient one week prior to coming to the hospital for similar complaint, when she had stool cultures collected which were all negative for any infectious etiology including Clostridium difficile. Patient stated her diarrhea has been persistently getting worse with increased frequency of multiple loose non-bloody watery stools daily, despite no changes made in her medication. On physical exam abdomen was soft with minimal epigastric tenderness, no left lower quadrant tenderness, rebounding or guarding. On labs patient had no leukocytosis on presentation nor any fevers throughout the hospital stay. Stool cultures which were collected again on admission were negative. CT scan of abdomen and pelvis with contrast suggested possible sigmoid diverticulitis versus colitis, no abscess or free air was noted. After ruling out all causes of infectious and secretory diarrhea, attention was turned towards a possible side effect of medication. Serum theophylline levels were checked which were greater than 22mg, (acceptable normal therapeutic level range being 10-20 mg). Flexible sigmoidoscopy was done which revealed congested mucosa in the rectum and sigmoid colon. Biopsies were taken, revealing colonic mucosa with mild edema, congestion and focal surface erosion. Given elevated Theophylline levels, suspicion for the Theophylline toxicities was the most likely etiology. After stopping theophylline, patient’s diarrhea resolved.

Discussion: Although, nausea and vomiting are common side effects while on theophylline (even at therapeutic range), persistent watery diarrhea is not commonly reported as a side effect of theophylline. Although, not clear how theophylline causes diarrhea, one possible theory is by blocking phosphodiesterase, theophylline leads to increase in adenylyl cyclase causing an increase in cyclic-AMP at the enterocytes which releases water and electrolytes in gut lumen leading to osmotic diarrhea. Theophylline has a very narrow therapeutic index, and can easily accumulate in the blood reaching toxic levels. In general, the drug should be monitored every 3-12 months to see if levels are within therapeutic range. Theophylline induced diarrhea will present as persistent watery loose stools with mild edema, congestion and focal surface erosion on biopsy. Discontinuation of the drug will lead to resolution of diarrhea immediately.

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New Jersey-Clinical Vignette-Poster Finalist
Mario Gioia, MD

Title: A rare case of acute renal failure secondary to intravenous immunoglobulin associated interstitial nephritis

Authors: Mario Gioia MD, Bilal Saleem MD, Sashi Raja MD, Shubho Sarkar MD

Introduction: The use of intravenous immunoglobulin (IVIG) has been proven beneficial in a multitude of disease processes, but it has been associated with a host of complications including fever, nausea, chills, hypotension/hypertension, hemolysis, aseptic meningitis, anaphylaxis and many more. In rare circumstances, it has been associated with acute renal failure, though the exact mechanism of renal failure in these patients is unknown and has not been well studied. Sucrose nephropathy has been a well-defined etiology behind patients developing renal failure after IVIG. Sucrose causes osmotic stress to the proximal tubules, resulting in extensive vaculation of the tubules, consistent with osmotic injury. This case describes a different and unique mechanism behind the development of renal failure after exposure to IVIG.

Case Presentation: A 64-year-old woman with biopsy proven linear IgA bullous dermatosis presented to our institution with acute onset hematuria, suprapubic tenderness, and fever after undergoing her second round of IVIG for her rare autoimmune disease. On presentation, her temperature was 39.5 degrees celsius while her other vitals were normal. On physical exam, she had moderate tenderness to palpation of the suprapubic region along with costovertebral angle tenderness bilaterally. A urinary catheter was inserted revealing frank hematuria. Labs revealed a creatinine of 2.4 mg/dL (baseline of 0.8 mg/dL). Over the next two days, her creatinine had risen to 8.9 mg/dL and she became oliguric. Hemodialysis was initiated. Her hospital stay was further complicated by hepatic injury, hemolytic anemia, and myocardial infarction; all thought to be adverse reactions from IVIG.

Since she remained oliguric after a few days of hemodialysis, a renal biopsy was performed revealing acute tubulointerstitial nephritis with the presence of eosinophils within the interstitium. She was subsequently started on a prednisone taper, and over the next few days there was noticeable improvement in her creatinine and urine output along with her many other lab and clinical abnormalities. She continued to improve on the outpatient basis, with a creatinine that had trended down to a new baseline of 1.5 mg/dL. At six month follow up, she has since been off hemodialysis and is doing well.

Discussion: Acute renal failure after the use of IVIG is a rare complication, with an incidence of less than 1%. Our patients renal failure initially was thought to be caused by sucrose nephropathy, resulting in significant swelling of the kidneys leading to obstruction. This mechanism of kidney injury is due to sucrose containing IVIG formulas, and has been a well-defined etiology seen in multiple case series. As our patient developed her symptoms and lab abnormalities acutely after IVIG, it was deemed to be the culprit of her clinical deterioration. Since there was no clinical improvement on hemodialysis, a biopsy was performed revealing acute tubulointerstitial nephritis (AIN). After steroids were initiated for AIN alongside hemodialysis, her clinical status improved. There have only been one or two more case reports that describe this phenomenon.
New Jersey-Clinical Vignette-Poster Finalist
Nagham S Jafar

Title Raise The Awareness Cardiac Arrest Due To Cannabinoid Use

Authors: L. Jafar, T. Gyaltsen, A. Mihali, N. Mazhar, R. Nagra, B. Nagra

Introduction: While cannabinoids can be associated with cardiac arrhythmias, cardiac arrest is a rare but possibly fatal consequence of Marijuana use. Public awareness should be raised by extensively promoting all potential complications associated with its use. We present a case of cardiac arrest after the use of cannabinoids in an 18 year old man.

Case Presentation: An 18-year-old Caucasian male with history of Marijuana use suffered a witnessed cardiac arrest. There were no prior symptoms of palpitations or syncope and no family history of sudden death. The patient was found in VF and was shocked 4 times after which he regained sinus rhythm. He was intubated and started on TTM protocol. His initial labs were normal, Cardiac troponin I was 0.02 ng/mL, and toxicology screening was positive for cannabinoids. Echocardiogram revealed an EF of 55% to 60% with no structural abnormality. EKG showed atrial ectopic rhythm and QTc of 425 msec. After 3 days the patient was extubated and remained in stable condition with no neurological deficiencies. EP studies, TEE and genetic testing were all negative. An Implanted Defibrillator placement was placed for secondary prevention of fatal arrhythmia.

Discussion: The use of synthetic cannabinoids is gradually increasing in our country. It was reported that 11% of high school students used marijuana in the last 12 months due to its availability and affordability. Cardiovascular manifestations of cannabis result from a biphasic dose-dependent physiological effect on the autonomic nervous system: low to moderate doses tend to cause tachycardia and raise blood pressure by increasing the sympathetic activity, whilst high doses produce bradycardia and hypotension by increasing the parasympathetic activity.

Marijuana in conjunction with alcohol and drugs such as cocaine creates more synergistic effects on the myocardium and can trigger significant tachycardia that leads to ischemia, infarction and even death. Arrhythmias due to the use of Marijuana are reported in literature like ventricular tachy-arrhythmias and atrial fibrillation.

Cannabinoids can also unmask the Brugada syndrome. Hence proper counseling to the patient and general population is crucial by clinicians.

Learning points:

1. Marijuana use can be associated with serious cardiovascular complications including cardiac arrest as result of arrhythmia.
2. Patient with cardiac arrest after using Marijuana should undergo investigation for Brugada syndrome and consider Implanted Defibrillator placement for secondary prevention.
3. The concurrent use of Marijuana with Alcohol and / or other drugs can increase the risk of cardiovascular adverse events.
References

New Jersey-Clinical Vignette-Poster Finalist
Alex F Lazo, MD

Title: Variceal bleeding as initial presentation of sarcoidosis

Authors: Alex F. Lazo, MD. Morristown Medical Center, Department of Internal Medicine.
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Introduction: Sarcoidosis is a systemic disease characterized by non-caseating granulomas in the affected organs including skin, lungs, heart and liver(1, 2). Hepatic involvement occurs in half the patients suffering of sarcoidosis, but <1% of patients suffer serious complications(1, 3). These complications are hepatic dysfunction, liver cirrhosis, portal hypertension(PHTN) and esophageal varices rupture(1, 4, 5). This case report should raise physician awareness of variceal bleeding as a complication of sarcoidosis.

Case Presentation: A 52-year-old man presented to the hospital with hematemesis. Four years prior he had presented with thrombocytopenia and splenomegaly. Whole body computed tomography(CT) scans revealed hilar and mesenteric adenopathy without evidence of PHTN. He was initially followed without therapy but due to increasing arthralgias he had recently begun prednisone and methotrexate. An upper endoscopy showed Grade III varices found in the lower two-thirds of esophagus which were treated with banding. An abdominal CT scan showed irregular liver contours and PHTN which were new when compared with previous scans. Serology for hepatitis B and C and anti-smooth muscle antibodies were negative. A biopsy of the liver was performed and was negative for sarcoidosis or cirrhosis. Iron stains and flow cytometry were negative. Diagnosis was sarcoidosis with PHTN and esophageal variceal bleed with negative liver biopsy.

Discussion: Sarcoidosis usually presents with chest involvement manifesting as mediastinal lymph nodes and pulmonary fibrosis(6), but it is estimated that 50% of patients have involvement of organs outside the thorax(2, 6, 7). Liver involvement is rare, and the clinical presentation can vary. Some can rapidly progress and complicate with cirrhosis, PHTN, cholestasis and Budd-Chiari syndrome(7-9). PHTN can develop without cirrhosis and or fibrosis on liver biopsies(10). Our case did not show evidence of either. Granulomas are found in 24-94% of liver biopsies and autopsies(11, 12). Granulomatous formation can produce parenchymal heterogeneity and surface nodularity, as well as lymphadenopathy and splenomegaly, simulating cirrhosis and PHTN(13, 14). Treatment consists of steroids, but it’s role in hepatic sarcoidosis is unclear(2) and it does not seem to alleviate PHTN(10) and may in fact worsen fibrosis(15). Hepatic sarcoidosis can be a rare and clinically challenging presentation. Guidelines for the management and treatment of this type of presentation are unavailable and the pathophysiology of the development of PHTN is not fully understood. The clinician should be aware of PHTN as a presentation of extrapulmonary sarcoidosis and monitor patients for complications such as variceal bleed. Further studies and guidelines are needed to elucidate the optimal management of this disease.

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New Jersey-Clinical Vignette-Poster Finalist
Ahmed Khaled Ahmed Mohammed, MD

Title: Human herpesvirus-6: An emerging etiology of encephalitis

Authors: Ahmed Hossary M.D., Kathleen Capaccione, M.D., Ph.D, and Jacqueline Darcey M.D.

Introduction: Encephalitis in the adult population is a common presentation, with an incidence between 1 in 250,000 and 1 in 500,000 yearly in industrialized nations. Clinical manifestations are protean, including headache, fever, confusion, impaired judgment, irritability, weakness, drowsiness, and unsteady gait. In immunocompetent adults, it is commonly caused by herpes simplex virus (HSV) type 1 or type 2. Human herpesvirus-6 (HHV-6) is an established cause of encephalitis in immunocompromised patients, but several cases have now been reported in the immunocompetent population.

Case Presentation: Here, we present the case of a patient with a complex medical history, including prior HSV infection, who developed encephalitis during prolonged hospitalization. Her mental status waxed and waned, oscillating between extreme agitation, lethargy, and frank confusion to alertness without orientation. Imaging and blood cultures showed no obvious etiology of her encephalopathy. After progressive clinical deterioration, the decision was made to perform a lumbar puncture to obtain CSF for analysis. A viral panel demonstrated infection with HHV-6, and was negative for HSV-1 and HSV-2. Gancyclovir, an established treatment of HHV-6 encephalitis, was initiated and the patient experienced total resolution of her encephalitis within days.

Discussion: The unexpected finding of HHV-6 in a clinical context where HSV-1 or HSV-2 was expected highlights the importance of recognizing the possibility of this emerging pathogen in the immunocompetent population. Previously only seen in the immunocompromised population, HHV-6 has now been reported multiple times in immunocompetent patients. In light of the fact that treatment of HHV-6 is different than that of the more common HSV, it is critical that clinicians consider HHV-6 in their differential diagnosis of encephalitis and for them to obtain diagnostic confirmation of the etiology of encephalitis before initiating treatment. Although tempting to treat empirically based only on the clinical presentation, this case highlights the need for further testing to direct management. As HHV-6 emerges as a causative agent of encephalitis in the immunocompetent population, clinicians must both recognize and appropriately treat this potentially deadly pathogen.
New Jersey-Clinical Vignette-Poster Finalist
Muhammad Ahad Nabil, MD

Title: A Case of Anabolic Steroid-Induced Cardiomyopathy Complicated by an Apical Thrombus

Authors: Muhammad Ahad Nabil MD, Syed Jaleel MD

Introduction: The use of anabolic-androgenic steroids (AAS) is well known, not just by professional athletes but also by casual fitness enthusiasts and sub-elite sportsmen. People who abuse AAS for a prolonged time period are at risk for hypertension, cardiovascular disease, cerebrovascular events and lipid metabolism disorders. Some major cardiac adverse effects include ventricular remodeling, myocardial infarction and sudden cardiac death. We present a case where prolonged use of AAS resulted in cardiomyopathy and heart failure which was further complicated by left ventricular apical clot formation.

Case Presentation: Our patient was a 45-year old male, with no known medical history, who presented to the hospital with scrotal swelling for 4 days. He also complained of exertional dyspnea for last few weeks. He confessed to using anabolic steroids for many years but otherwise denied any other recreational drug or alcohol abuse. On examination, he was noted to have jugular venous distension as well as a positive hepato-jugular reflux. Chest-xray was consistent with enlarged cardiac silhouette and BNP was 2621. Patient was subsequently admitted for acute heart failure. Trans-thoracic echo (TTE) showed severe global hypokinesis with an ejection fraction of 10-15%, stage-III diastolic dysfunction along with a large apical thrombus. Ischemic cardiomyopathy was ruled out with a cardiac catheterization which revealed only mild disease of the left anterior descending artery. Viral myocarditis was ruled out as serology for Parvo virus, Coxsackie virus, Hepatitis B and C viruses turned out to be negative. Lyme disease was similarly ruled out. His TSH was normal. Urine drug screen was negative for all substances as well.

Patient was diagnosed with anabolic steroid-induced cardiomyopathy which was further complicated by an apical thrombus. He was started on warfarin with a goal INR of 2.5. After the patient was clinically euvolemic and his symptoms had significantly improved, he was discharged with a LifeVest and advised to follow up with his primary care physician as well as a cardiologist.

Discussion: Use of supra-physiological testosterone doses have shown to result in increased fat-free mass and muscle size. However, it also has growth-promoting effects on cardiac tissue followed by release of apoptogenic factors and cell death. Direct myocardial injury can result from increased fibrosis of the myocardium mediated by aldosterone-like effects. AAS enhance platelet aggregation and thrombus formation by increasing platelet production of thromboxane A2, decreasing production of prostacyclin and increasing fibrinogen levels. In our patient, intra-cardiac thrombus could have been the result of apical hypokinesia, steroid-induced predisposition to clot formation or both. Steroid induced cardiomyopathy is a known complication of AAS but rarely has been associated with an intra-cardiac thrombus. Such patients are at high risk of thrombo-embolism and is important to recognize them in early stages of their disease for closer monitoring and follow up.
New Jersey-Clinical Vignette-Poster Finalist
Jay Naik, MD

Title: Two Wrongs Make a Right? A Lesson in Patient Safety

Authors: Jay Naik MD, Julie Kanevsky MD, Montefiore Medical Center/Albert Einstein College of Medicine, Bronx, NY

Introduction: In recent years, medication reconciliation has become the focus of many patient safety initiatives. However, there is no consensus on what constitutes adequate medication reconciliation. In this case, we discuss an important, but often overlooked aspect of medication reconciliation.

Case Presentation: A 72-year-old man with a history of diabetes and hypertension presented with syncope. He felt progressively lethargic, became unresponsive, and woke up in the ED. He had a similar event 2 weeks prior, with an unrevealing syncope work-up, including brain CT/MRI. Social history was negative for alcohol and drug use. Exam was notable for lethargy and transient hypotension to systolic BP of 80. Labs and brain CT were unremarkable. Due to prominent aortic contour on CXR, a CT chest/abdomen was obtained and ruled out aortic dissection, but detected an 8-cm renal mass. Patient was admitted, and his home medications, including chlorthalidone and fosinopril were held.

The next morning the patient appeared well, but his wife became lethargic. She was euglycemic, with BP of 160/90. She had already taken her daily dose of chlorthalidone and fosinopril and attributed elevated BP to being in the ED all night with her husband. Over the next hour, she became progressively difficult to arouse and had slurred speech. A stroke code was called, and right eye exotropia and impaired upward and downward gaze were identified. After a negative head CT, she was given TPA for suspected right midbrain CVA. Within a few hours, her mental status returned to baseline. Brain MRI/MRA were unremarkable.

The similarity of the patient’s and his wife’s symptoms raised the suspicion of a unifying cause, especially since the patient’s wife took her dose of BP medication from his bottle while he was in the ED. When the patient’s medications were examined using an online pill identifier, it was discovered that the bottle labeled “Chlorthalidone 25mg” actually contained clozapine 50mg tabs, which neither the patient nor his wife was ever prescribed. Clozapine prescribing is tightly restricted due to its severe potential side effects, including agranulocytosis. It is normally started at 12.5mg/day with slow titration because the risk of orthostatic hypotension, bradycardia, lethargy, syncope, cardiotoxicity, and cardiac arrest is highest during the initial titration period.

Discussion: In this case, both the patient and his wife were exposed to very high doses of clozapine due to a dispensing error and had neurologic manifestations consistent with clozapine toxicity. Fortuitously, this medication error allowed for the early diagnosis and resection of the renal cell carcinoma discovered incidentally in this asymptomatic patient with a normal urinalysis. Dispensing errors account for about one-fifth of all medication errors, which are a leading cause of mortality in the US. It is unusual and impractical for physicians to routinely examine bottle contents, which is one of the few ways to detect dispensing errors. Therefore, a high index of suspicion, as with this case, is required to identify such errors.
New Jersey-Clinical Vignette-Poster Finalist
Kennedy C Ukadike, MD

Title: Bilateral Giant Cell Arteritis with Renal ANCA-Associated Vasculitis

Authors: Kennedy C. Ukadike, MD, MS1,2; Marnie G. Aguasvivas, MD1,2; Louis C. Jan, MD1,3; Marc S. Zelkowitz, MD1,3, 1Department of Internal Medicine, Englewood Hospital and Medical Center, Englewood, NJ, 2Graduate Medical Education, Seton Hall University School of Health and Medical Sciences, South Orange, NJ, 3Division of Nephrology, The Valley Hospital, Valley Health System, Ridgewood, NJ

Introduction: Giant cell arteritis (GCA) is categorized as a medium-to-large vessel vasculitis. Although an elevated ESR and/or CRP in combination with the clinical findings are highly suspicious of GCA, temporal artery biopsy is highly recommended in all suspected patients. However, a negative biopsy or absence of giant cells does not rule out the disease given the possibility of skip lesions. In contrast to small vessel vasculitides which can be linked to ANCA, large vessel vasculitides have no known associated antibody expression patterns. Here we present an extremely rare phenomenon in a patient with biopsy-proven bilateral GCA with renal failure in whom kidney biopsy was consistent with AAV.

Case Presentation: 72-year-old Caucasian woman with PMH of cervical spinal stenosis presented with one-month history of worsening generalized weakness and soreness in all her extremities. Three months prior she had complained of persistent headaches for which she took ibuprofen with limited relief. Suspected of having GCA based on ESR 75 mm/hr with no biopsy at the time, she was started on prednisone 60mg oral daily with tapering which provided relief. However, when the prednisone dose was down to 10mg, the headaches started to reoccur along with her new complaints. On physical exam, her vital signs were normal, and findings were notable for tenderness in bilateral temporal areas and in the muscle groups of all extremities with weakness of lower extremities. Initial workup revealed BUN 62mg/dl, creatinine 2.8mg/dl, random urine protein 76mg/dl with 24-hour urine protein 576mg but no urine RBCs, ESR 81mm/hr, CRP 170mg/l, total CPK 245u/l, and positive MPO-ANCA with titer >8.

Bilateral temporal artery biopsies revealed chronic inflammation on the left, and acute on chronic inflammation with rare giant cells on the right, both with elastic stains showing disruption of the internal elastic lamina, all consistent with GCA. Renal biopsy revealed necrotizing vasculitis, extensive tubulitis and interstitial inflammation, all consistent with MPO-ANCA-associated vasculitis. Immunofluorescence showed no evidence of immune complex glomerular disease. Recommended treatment included steroid taper and cyclophosphamide with consideration for rituximab if adequate response not achieved later.

Discussion: Renal involvement in patients with GCA is quite rare, and to the best of our knowledge, only two cases have been reported with clear documentation of association with AAV. Majority of patients with renal limited vasculitis are seropositive for ANCA, mostly MPO-ANCA. The patient in this case being MPO-ANCA positive with renal vasculitis and GCA represents an extremely rare clinical overlap between small, medium and large vessel vasculitis. In all patients diagnosed with GCA, care should be taken to closely evaluate for possible associated renal disease. Doing so has important therapeutic implications as GCA has good clinical response to steroids, but the presence of renal vasculitis requires the addition of another immunosuppressant such as cyclophosphamide.

References


New Jersey-Clinical Vignette-Poster Finalist
Shivani Vekaria, MD

Title: An Unusual Cause of Hypercalcemia in a Patient with AIDS

Authors: Shivani Vekaria, MD, Gabriela Ferreira, MD, Department of Medicine, Rutgers Robert Wood Johnson Medical School, New Brunswick, NJ.

Introduction: Hypercalcemia is common among hospitalized patients and determining the cause requires a systematic approach. Most cases are due to elevated intact parathyroid hormone (iPTH) or paraneoplastic parathyroid-related peptide (PTHrP) levels, or to osteolysis from bone metastases. We report an unusual case of severe hypercalcemia due to disseminated Mycobacterium Avium Complex (MAI) in a patient with AIDS.

Case Presentation: A 37-year-old male with HIV/AIDS presented with delirium, abdominal pain, and 10-pound weight loss for one week. He denied fevers, chills, or night sweats. He appeared thin, somnolent and dehydrated. Labs were significant for total calcium 14.3 (8.6-10.4 mg/dL), ionized calcium 7.0 (4.6-5.3 mg/dL), creatinine 5.1 (0.5-1.2 mg/dL), HIV viral load 9950, and CD4 70 (424-1509 #/mm). Computed tomography of chest, abdomen, and pelvis demonstrated mediastinal, mesenteric, retroperitoneal and supraclavicular adenopathy with severe constipation. Subsequent Results: included: iPTH 5 (9-76 pg/mL), PTHrP 0.4 (<0.2 pmol/L), Vitamin D25 31.7 (25-80 ng/mL) and Vitamin D1,25 44 (18-64 pg/mL). After aggressive hydration, his symptoms resolved, and electrolytes and renal function improved. A fine needle aspirate and core biopsy of a cervical lymph node both stained strongly positive (> 3+) for acid-fast bacillus, suggestive of MAI, which was confirmed on culture. Polymerase chain reaction for Mycobacterium tuberculosis was negative. Biopsies revealed diffuse histiocytic infiltration suggestive of granulomatous inflammation. Once his serum calcium stabilized, he was discharged home on ethambutol and azithromycin therapy for MAI. Unfortunately, he was readmitted frequently over the next two months with hypercalcemia, and each time improved with hydration. He admitted medication nonadherence, despite receiving financial assistance and free medication.

Discussion: Hypercalcemia occurs in many hospitalized patients, and internists must be familiar with its presentation, etiologies and treatment. One first confirms the diagnosis with an ionized calcium level. In order to determine etiology, one then measures iPTH. If iPTH is low or normal, then check PTHrP level and evaluate for bone metastases from malignancy. If these are negative, a systematic approach will help identify less common causes, such as vitamin D excess, medications, endocrine disorders, immobilization with increased bone turnover, and rhabdomyolysis. Rarely, hypercalcemic patients without malignancy will have elevated PTHrP, as reported in cases of HIV-associated adenopathy.1,2 Initial treatment of hypercalcemia is resuscitation with isotonic fluid, followed by bisphosphonate therapy in refractory cases.3 Once the acute hypercalcemia resolves, management is focused on treating the underlying cause to prevent recurrence.

In this case, hypercalcemia was attributed to MAI, and recurrences to medication noncompliance. Interestingly, although biopsies showed histiocytic infiltration suggestive of granulomatosis, both Vitamin D25 and Vitamin D1,25 were normal. The absence of granulomas was postulated to be due to lymphopenia from poorly treated HIV. However, it is possible that hypercalcemia was from the diffuse lymphadenopathy associated with his HIV, since his PTHrP level was elevated.

References
New Jersey-Clinical Vignette-Poster Finalist
Kristina Zarkua, MD

Title: The Reverse Side Of ICD

Authors: Kristina Zarkua MD, Naziha Slimani MD, Hannah James MD, Georgy Zarkua MD

Introduction: As the prevalence of cardiovascular diseases is increasing in the population, the number of patients requiring implantable cardioverter-defibrillators (ICDs) is significantly increasing. ICD leads are known to be a predisposing factor for thrombosis. It is a rare, but potentially fatal condition. We present a rare case of a subclavian vein thrombus manifested after an ICD lead replacement.

Case Presentation: This is the case of a 41-year-old Caucasian male with history of Brugada syndrome with AICD placement, recent change of the leads and battery secondary to malfunction and breakage, presented to the hospital because of feeling of fullness in his head, face swelling and redness along with prominent neck veins, shortness of breath started gradually few days ago. The patient reported that two months prior he had inappropriate ICD shock related to lead malfunction, so leads were extracted and replaced.

On arrival vital signs were normal, prominent neck veins, facial redness, and swelling were noted on physical exam. Initial laboratory data were within normal limits, EKG showed Brugada pattern, an echocardiogram showed normal ejection fraction and LV size. CT chest with contrast revealed possible stricture and thrombosis in superior vena cava. The patient was started on anticoagulation immediately.

Shortly after admission patient was seen by vascular surgeon, who recommended against thrombectomy due to thrombus location, but to continue long-term anticoagulant therapy, consider thrombolysis and AICD lead reevaluation. SVC angioplasty was performed by Interventional Radiology Department in the area of stenosis, residual thrombus adherent to ICD leads did not resolve, the patient was considered as a poor candidate for tPA treatment. After Discussion: with patient’s cardiologist was decided to continue with an extraction of AICD, restratification of risks of fatal arrhythmias and further decision making regarding device replacement.

Discussion: SVC thrombosis associated with ICD leads is an unusual complication of ICD insertion. Previous studies reported that significant thrombotic or embolic events occur in 0.6-3.5% of patients with permanent transvenous pacing leads. Pacemaker lead-associated thrombus formation is multifactorial and several possible mechanisms may contribute to endothelial injury, inflammation, hypercoagulability and foreign body-type reaction. Heart failure, atrial fibrillation, and hypercoagulability may also cause thrombus formation. Implantation procedure per se probably causes a varying degree of venous endothelial injury, which can be further exacerbated by inflammation and irritation induced by the friction of the transvenous lead over time. The type of treatment should be established according to the dimension, mobility, and location of the thrombus.

In conclusion, an ICD placement per se can be a significant risk factor for serious complications, including thrombi formation, and requires weighing of all possible pros and cons of the procedure.
New Mexico-Clinical Vignette-Poster Finalist
Amir Anabtawi, MD

Title: Can coronary artery aneurysmal fistula present with acute coronary syndrome?

Authors: Amir Anabtawi\textsuperscript{1}, Abed Anabtawi\textsuperscript{1,2}, Wael Al-Husami\textsuperscript{2}, 1. University of New Mexico, Albuquerque, NM, USA, 2. Brown University, Providence, RI, USA

Introduction: Coronary arteriovenous fistula is a rare defect that is mostly congenital in origin. It usually represents a communication between a coronary artery and a great vessel, vena cava or a cardiac chamber. This is a case report of a patient who presented with very unusual fistula which originates from proximal left anterior descending coronary artery aneurysmal drainage into the pulmonary artery.

Case Presentation: We are presenting a 69 year old female patient with history of hypertensive renovascular disease status post bilateral renal artery stenting, stable angina, hyperlipidemia and glucose intolerance. She presented with exertional left arm heaviness. Vital signs were stable. Heart, chest and neurologic physical examinations were unremarkable. Electrocardiogram showed sinus rhythm, RBBB, and no significant ST-T changes. Ejection fraction was normal by echocardiogram without evidence of wall motion abnormalities or significant valvular heart disease. Troponin I was elevated. She was diagnosed with non-ST elevation myocardial infarction. Antiplatelet therapy, anticoagulation, ACEi, beta blocker and statin were started. Left heart catheterization showed a proximal left anterior descending coronary artery-to-pulmonary artery fistula with severe aneurysmal drainage.

The rest of LAD showed mild diffuse disease and was tortuous artery. The circumflex and right coronary arteries showed no significant disease. The patient was evaluated by cardiothoracic surgery and aggressive medical therapy was recommended. She remained asymptomatic on follow-up visit.

Discussion: A coronary arteriovenous fistula is a congenital or acquired coronary artery anomaly in which blood is shunted into a cardiac chamber or great vessel, superceding the myocardial capillary network. Clinical presentation may be heralded by the development of dyspnea, congestive heart failure, angina, dysrhythmias or myocardial infarction. Incidence is 0.5 % of all cardiac catheterizations, out of which 15 % of these fistulas open to the pulmonary artery and is rarely associated with aneurysmal dilatation of the coronary artery. Although noninvasive imaging may facilitate the diagnosis and identification of the fistula origin and insertion; coronary angiography and hemodynamic assessments are still the gold standard for evaluating the coronary anatomy and the presence of atherosclerosis or other structural anomalies. Recently, cardiac CTA scan drew more attention to such cases and added a tremendous amount of information. Treatment is indicated in symptomatic patients who failed medical therapy and for those who are asymptomatic with future risk complications. Minimally invasive percutaneous transcatheter embolization has been used routinely and in rare occasions surgical correction could be an option. Our case presented with acute coronary syndrome either due to coronary aneurysmal fistula steal phenomenon or distal embolization to the LAD.
New Mexico-Clinical Vignette-Poster Finalist
Chih-Wei Chang

Title: Left Ventricular Metastasis in Renal Cell Carcinoma Without Vena Cava or Right Heart Involvement: an Unusual Risk for Wall Perforation

Authors: Chih-Wei Chang MD, Ihab Ajaaj MD, Alex Schevchuck MD

Introduction: Cardiac metastasis of renal cell carcinoma (RCC) in the absence of vena cava and right heart involvement is extremely rare with no report on strategy for lowering risk of wall perforation.

Case Presentation: A 79-year-old female with history of left-sided pT2a papillary RCC type II, status post left open radical nephrectomy nine months ago presented with bilateral chest pain. Physical exam and laboratory findings were unremarkable. EKG revealed normal sinus rhythm with left axis deviation, and T wave inversion in anterior leads. Mammogram showed a mass in the left breast and multiple masses in the right breast, highly suggestive of malignancy (BI-RADS of 5). A CT chest/abdomen/pelvis with contrast revealed two metastatic transmural lesions in the left ventricular wall measuring 1.5 cm by 1.7 cm and 1.3 cm by 1.2 cm, mediastinal adenopathy and retroperitoneal adenopathy, and metastatic lesions to the lungs, liver, left adrenal gland, subcutaneous fat of the right breast, musculature of the proximal lower extremities, and left rib. An ultrasound guided needle biopsy of the right breast was consistent with metastatic renal cell carcinoma. A transthoracic echocardiogram revealed normal left ventricular systolic function with ejection fraction (EF) of 56% and two highly vascular infiltrating tumors at the mid and distal segments of the antero-lateral and infero-lateral walls that appeared to be nearly transmural only sparing a thin layer of endocardium. This was concerning for risk of wall rupture. The patient was started on lisinopril and carvedilol to decrease left ventricular wall stress.

Discussion: Primary cardiac tumors are rare, and most cardiac tumors are secondary (metastatic). RCC comprises about 3% of all malignant tumors. Cardiac metastasis was present only in 11% of patient who died of RCC. However, cardiac metastasis in the absence of vena cava and right heart involvement is exceedingly rare with only a few cases reported in the literature. Medical treatments include tyrosine kinase inhibitors such as pazopanib, monoclonal antibody nivolumab, and mammalian target of rapamycin inhibitors such as everolimus. There are several reports of ventricular wall perforation associated with treatment-induced necrosis when the metastatic tumor was embedded within the myocardium. Medical management strategy for lowering risk of wall perforation in this situation has not been established, with no prior report on this topic. Based on the limited observation in this unique case, we suggest using angiotensin-converting enzyme inhibitors (ACE-I) and beta-blockers to reduce ventricular wall stress for lowering the risk of wall perforation as long as there is no baseline systolic or diastolic dysfunction. During treatment, patient’s cardiac function needs to be closely monitored with serial echocardiograms. A longitudinal follow-up will provide valuable information on the efficacy and long-term safety of ACE-I and beta-blocker utilization in this clinical scenario.

References


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New Mexico-Clinical Vignette-Poster Finalist
James T Dean, DO

Title: Where's the source: An atypical case of Veillonella bacteremia.

Authors: James Dean III DO. Diedre Hofinger MD, FACP.

Introduction: *Veillonella parvula* is a gram-negative anaerobic cocci that is found as part of the normal human oral, vaginal and gastrointestinal flora (1). It is rarely found to be pathogenic in humans but has been implicated in various infections including osteomyelitis, meningitis, pneumonia, endocarditis and bacteremia. We present a case of *Veillonella* bacteremia.

Case Presentation: A 58-year-old male with a history of alcohol dependence, hepatic cirrhosis, chronic cystitis, urinary outflow obstructions and chronic diarrhea with intermittent melena presented following a fall at home. Physical exam was significant for fever 102.1°F, pulse 123 bpm, poor dentition, mild abdominal distention but non-tender. The laboratory Results: were remarkable for WBC 17 K/mm$^3$ with left shift 89% neutrophils, lactic acid 2.4 mmol/L and a urinalysis consistent with a urinary tract infection. Urinary cultures were positive for *Escherichia coli*. One set of blood cultures was positive for *E. coli*, however both sets of blood cultures were positive for *Veillonella* species. Patient was started on amoxicillin/clavulanate and cefazolin. To work up source of infection, a positron emission tomography (PET) scan was performed which was negative for osteomyelitis, esophagastroduodenoscopy (EGD) demonstrated mild gastropathy and biopsies were positive for *Helicobacter pylori*, colonoscopy demonstrated diverticulosis and a panorex of the teeth was negative. Diagnostic paracentesis demonstrated no growth on cultures. Without an identified source of infection it was felt the *Veillonella* bacteremia originated in urine or due to poor dentition. The patient clinically improved on amoxicillin/clavulanate and cefazolin and was discharged home to complete 14 days of antibiotics.

Discussion: This case demonstrates the need for a comprehensive yet focused work up of a rare pathogen, especially when patients have symptoms consistent with infection. There is little written about *Veillonella* infections. *Veillonella* spp are part of the normal oral, vaginal and gastrointestinal flora of humans (1). There are 13 species of *Veillonella* identified but only 4 species causing human infections including *V. parvula*, *V. dispar*, *V. montpellierensis* and *V. alcaliens* (2, 3). There are no standard susceptibility studies for *Veillonella*, however, in vitro data demonstrates susceptibility to beta-lactams, cephalosporins, metronidazole and clindamycin (4). Resistance has been demonstrated to penicillin in recent reports (5). This patient had multiple risk factors for both a GI and oropharynx source for his *Veillonella* bacteremia due to his poor dentition, alcohol history with cirrhosis and diverticulosis. We did not find a clear source for this case of *Veillonella* bacteremia which is not uncommon based on literature review. Although *Veillonella* is a rare bacterium, it should be considered a pathogen in patients with risk factors, and a comprehensive workup should be initiated.

References

New Mexico-Clinical Vignette-Poster Finalist
Imaneh Fallahi

Title: Beyond Typhoid Mary: Optimal Management of Salmonella Aortitis.

Authors: Dr. Imaneh Fallahi, Dr. Charles Pizanis

Introduction: Salmonella infection of the aorta and its adjacent arteries is a rare but life-threatening medical condition. It usually involves the abdominal aorta and femoral arteries with very few cases arising above the renal arteries. The pathogenesis involves bacterial invasion and seeding of atherosclerotic plaque within an arterial wall or an embolic phenomenon as a complication of infective endocarditis.

Case Presentation: A 65-year-old woman with a past medical history of diabetes mellitus, non-alcoholic fatty liver disease and recent laparoscopic cholecystectomy for acute cholecystitis presented from a referring hospital with one-month history of gradual and worsening abdominal pain and a CT scan concerning for aortitis. The patient’s abdominal symptoms started four weeks after her cholecystectomy. Two weeks later she was diagnosed with urinary tract infection secondary to Salmonella infection which was treated with a ten-day course of amoxicillin. However, her symptoms persisted prompting her to seek medical reevaluation. Upon admission to our institution, two sets of blood culture were drawn with one returning positive for Salmonella spp. Treatment was commenced with intravenous ceftriaxone and vascular surgery was consulted. To rule out endocarditis, a transthoracic echocardiogram was completed which did not show any valvular vegetation or significant valvular dysfunction. A repeat CT scan of the abdomen demonstrated abnormal soft tissue surrounding the infra-renal abdominal aorta consistent with aortitis with no clear pseudo-aneurysm. The patient underwent resection of the infra-renal aortic infection, debridement of retroperitoneal phlegmon and aorta grafting with a rifampin-soaked graft. The aortic tissue and the purulent material in the retroperitoneum were sent for culture which were similarly positive for Salmonella spp. Her postoperative course was uneventful, and she was eventually discharged with a six-week course of intravenous ceftriaxone followed by lifelong suppressive antibiotic, ciprofloxacin daily.

Discussion: Studies done in the past have demonstrated improved survival with combined early surgical and medical intervention rather than medical therapy alone in treatment of Salmonella aortitis. Because of high incidence of recurrent infection, extra-anatomic bypass has been shown to be superior to in situ graft replacement especially in patients with infra-renal abdominal aortic infection. However, more recent studies have indicated that in situ repair is still successful for infrarenal aortic infection if there is a prompt confirmation of infection, debridement of infected tissue and use of prolonged antibiotic therapy. In our case, a combination of surgical and lifelong antibiotic therapy was chosen to manage the aortitis with no apparent post-operative complications. This case shows the importance of timely surgical intervention and comprehensive medical management of patients with such a rare vascular disease.

References
Title: Yamaguchi Syndrome: A Rare Cause of T-wave Inversions on Electrocardiogram

Authors: Madhura Myla, University of New Mexico School of Medicine; Saira Samani, University of New Mexico School of Medicine; Jerome Yatskowitz, University of New Mexico School of Medicine

Introduction: Yamaguchi syndrome is a rare form of hypertrophic cardiomyopathy. T-wave inversions in the anterolateral leads on electrocardiogram (ECG) are considered as one of the typical features of this syndrome. Yamaguchi syndrome should be ruled out in a patient presenting with chest pain and T-wave inversions before pursuing cardiac catheterization.

Case Presentation: A 63-year-old male with a history of hypertension was referred for an exercise stress echocardiography for chest pain, to evaluate for cardiac ischemia. Baseline ECG demonstrated significant T-wave inversion in the antero-apical leads. During the stress test, he developed symptoms of chest discomfort, which were non-limiting, with no changes on his ECG. The stress echocardiogram demonstrated normal resting wall motion, with no exercise-induced wall motion abnormality. Apical hypertrophy was noted in the contrast-enhanced echocardiogram. Despite having symptoms of chest discomfort during the stress test, it was decided not to pursue any further work-up for cardiac ischemia based on the fact that his symptoms were atypical. It was determined that the T-wave inversion on the ECG was due to the apical hypertrophy that was evident on the echocardiogram. The patient was prescribed Hydrochlorothiazide-Lisinopril 12.5mg-10mg, aspirin 81mg and discharged to home.

Discussion: Yamaguchi syndrome is also known as apical hypertrophic cardiomyopathy (AHCM). It is a rare form of hypertrophic cardiomyopathy. The course of this disease is usually benign and associated with good prognosis. The typical features of Yamaguchi syndrome include audible fourth heart sound, and giant (>10mm) T-wave inversions in the anterolateral leads on the electrocardiogram. A spade-like configuration of the left ventricular cavity is seen on ventriculography due to the apical left ventricular thickening. The imaging diagnostic criteria include left ventricular wall thickness of 15mm or more during diastole or apical to basal left ventricular wall thickness ratio of 1.5 or more. The goals of the treatment for symptomatic patients are a reduction in the heart rate and left ventricular afterload. β-blockers, calcium-channel blockers, and angiotensin-converting enzyme inhibitors are the classes of drugs typically used to achieve the treatment goals. Heart transplant and apical myectomy are the treatment options available for patients with severe heart failure refractory to medical therapy. Given that the patients with the apical variant of hypertrophic cardiomyopathy may have deep T-wave inversions in the anterolateral leads, interpretation for cardiac ischemia is particularly challenging. A careful history taking and imaging will help determine the etiology, prior to pursuing a more invasive strategy.

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Title: Novel Oral Anticoagulants - An Internist's perspective

Authors: Payal Sen, MD ; Uddalak Majumdar, MD ; Patrick Rendon, MD.

Introduction: Synergism and potentiation exists between anticoagulants, including the Novel Oral Anticoagulants. This effect can further be enhanced by certain antibiotics, often used for treating common respiratory and gastrointestinal infections. Failure to remember this can lead to life threatening bleeding and fatal consequences.

Case Presentation: A 90-year-old Caucasian female with non-valvular atrial fibrillation, on therapeutic anticoagulation with warfarin was admitted with 3 days of pleuritic chest pain and dyspnea. She had never smoked, had no sick contacts, and had recently undergone HIV testing which was negative. Physical exam revealed bronchial breath sounds in the middle lobe of the right lung. White count was elevated at 18,000 per mcl and CXR showed focal consolidation in the right lung. She was diagnosed with community acquired pneumonia and was treated with Levofoxacin. She underwent cardioversion for symptomatic atrial fibrillation with rapid ventricular rate and was switched from warfarin to Rivaroxaban 20 mg. After the second dose of Rivaroxaban, the patient developed acute onset of quadriplegia. Repeat INR was 6 and PT was 64.2 seconds. CT scan of the neck revealed posterior cervical epidural hematoma from C2 to C7 with cord compression. Unfortunately the patient sustained injury to the phrenic nerve roots causing paralysis of the diaphragm, and eventually died.

Discussion: Warfarin and Rivaroxaban have a synergistic anticoagulant effect, usually seen shortly after switching from warfarin to Rivaroxaban. Antibiotics also potentiate the effects of warfarin by inhibiting the metabolizing isoenzyme, CYP2C9. It is hypothesized that these two effects led to the fatal cervical spinal hematoma in our patient.

General internists are increasingly preferring rivaroxaban to warfarin, for the approved indication of thromboembolism prophylaxis, due to the convenience of not needing frequent INR checks, as well as the lack of multiple food and drug interactions with rivaroxaban. Rivaroxaban does not require laboratory monitoring; interacts less with food or other drugs; causes less intracranial or fatal bleeding; and is overall non-inferior to Warfarin. There is paucity of data on how to safely switch from oral vitamin K antagonists (VKA) to Rivaroxaban, and there is little research regarding the physiology of the potentiation effect. Expert opinion is to switch 24 hours after the INR is less than 3. A single observational matched-cohort study of switching VKA to Rivaroxaban supports this practice. In another study of in silico effects, a post-switch synergistic anticoagulant effect has also been observed and a nomogram has been developed for switching to Rivaroxaban. INR is affected variably by Rivaroxaban and cannot be used as a marker for its anticoagulant effect.

This case report teaches us that the synergistic effect and interactions with antibiotics should be kept in mind.
mind during the transition from VKAs to NOACs. Patients must be carefully monitored to prevent fatal consequences.

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New York-Clinical Vignette-Poster Finalist
Bhavna Abbi

Title: Cutaneous Microscopic Polyangiitis Resembling Cellulitis: A Case Report

Authors: Bhavna Abbi, M.D., Michael DiGiorno, D.O, Jacobo Futran, M.D., Department of Internal Medicine, St. John’s Riverside Hospital, Yonkers, NY, USA

Introduction: Microscopic Polyangiitis (MPA) is a necrotizing small vessel vasculitis that affects venules, capillaries and arterioles systemically, through blood vessel inflammation. (1) Cutaneous MPA occurs in 20-70% of patients. (2) We report a case of cutaneous MPA that was initially diagnosed as cellulitis. It is important for clinicians to consider MPA in the differential diagnosis of a patient presenting with lower extremity skin lesions to facilitate early treatment.

Case Presentation: A 76 year old African American female presented with chief complaint of edema and ulceration over her left medial malleolus for three days, and a non-healing right medial malleolus ulcer for the past year. The patient’s medical history was significant for Polymyalgia Rheumatica and chronic venous stasis.

At the time of presentation, her vital signs were within normal limits and physical examination was significant for 1+ dorsalis pedis pulses bilaterally. Bilateral ulceration was noted on the left and right medial malleoli.

Laboratory studies were significant for a leukocytosis of 13.8 k/mm³, hemoglobin and hematocrit of 9.7 g/dL and 29.6% respectively, ESR of 53 mm/hr, BUN of 25 mg/dL, creatinine of 1.5 mg/dL. Urinalysis showed 2+ proteinuria and 3+ hematuria. Wound culture of the left medial malleolus ulcer grew Pseudomonas aeruginosa, Escherichia coli, and methicillin-resistant Staphylococcus aureus (MRSA). Chest radiograph revealed opacities in the lower lungs bilaterally. The patient was presumed to have left lower extremity cellulitis and was started on vancomycin and piperacillin/tazobactam.

On hospitalization Day 4, the patient developed hemoptysis and became increasingly hypoxic. Chest computed tomography (CT) scan showed bibasilar subsegmental atelectasis, suggestive of a worsening right-sided pneumonia. Considering multi-system involvement, rheumatologic workup was performed and revealed positive antinuclear antibody, perinuclear anti-neutrophilic antibody titer of 1:160, positive myeloperoxidase antibody, total complement level greater than 65, and negative glomerular basement membrane antibody.

Renal biopsy revealed pauci-immune vasculitis with necrotizing arteritis, consistent with MPA. She was then started on IV pulse steroids, followed by Rituximab 375 mg/m² once per week for four doses. On discharge, pt was placed on Prednisone and subsequently improved.

Discussion: As seen in our case, cutaneous MPA can mimic the presentation of cellulitis. A possible explanation could be that, similar to bacterial exotoxins in cellulitis (3), myeloperoxidase-antineutrophil cytoplasmic antibodies (MPO-ANCA) interact with fungal antigens from pre-existing wounds or venous stasis, leading to ensuing inflammation. We propose that those with cutaneous manifestations typical for cellulitis and systemic involvement, undergo a rheumatologic workup to rule out MPA.

References


New York-Clinical Vignette-Poster Finalist
Tatsiana Aleksandrovich, MD

Title: The First Reported Case Of Quadruple-Valve Bartonella Endocarditis

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Introduction: Bartonella spp. are fastidious Gram-negative bacteria that cause blood culture-negative endocarditis and are the source of up to 3% of all cases of infective endocarditis. Bartonella endocarditis with all four valves involvement has never been described in the literature. We present a case of native quadruple-valve endocarditis in immunocompetent middle-aged man caused by Bartonella.

Case Presentation: A 52-year-old homeless man with a history of a remote stroke with residual left sided hemiparesis and dysarthria presented with fever, left sided body pain and altered mental status for two days.

The initial vitals: tachycardia and temperature of 101.2 F. On exam, he had poor dentition, dysarthria, left-sided facial paresis and spastic hemiparesis. Otherwise physical exam was unremarkable. The laboratory values on admission included Hb of 9.4 g/dl and WBC of 7000/mcL with 14% bands. The comprehensive metabolic panel was normal. HIV and Influenza tests were negative. Urinalysis was within normal limits. ECG was remarkable for sinus tachycardia. The chest radiograph showed clear lungs. CT-head demonstrated a right frontal-insular hypodensity and the brain MRI confirmed an acute right MCA-territory infarct.

Transthoracic echocardiography (TTE) revealed thickening of aortic, mitral, and tricuspid valves and possible mobile, soft-tissue masses on the aortic and mitral valves. Transesophageal echocardiography (TEE) revealed soft-tissue masses consistent with vegetations on all valves (Aortic valve 12 x 3 mm, Mitral valve 13 x 3 mm, Tricuspid valve 14 x 19 mm and Pulmonic valve: ill-defined mass). Empiric broad spectrum antibiotics were started for infectious endocarditis. Blood cultures were negative on multiple occasions. Further Infectious workup revealed a strongly-positive serology for Bartonella henselae IgG (>1:1024) and quintana Ig G (> 1:1024) and antibiotics were tailored accordingly.

Causes of nonbacterial thrombotic endocarditis were ruled out by negative workups for malignancy, hypercoagulable state and autoimmune processes.

After 6 weeks of the treatment, TTE showed the vegetations had significantly decreased in size and serum inflammatory markers had normalized.

Discussion: Bartonella endocarditis is a clinical and diagnostic challenge and commonly missed due to its subacute clinical course, presence of nonspecific symptoms and lack of criteria for diagnosis. This diagnosis must be considered in the patients with infectious endocarditis when routine bacterial cultures remain sterile. Echocardiography plays a pivotal role in the diagnosis of endocarditis as a source of cardiac embolism.
In this case, cardioembolic stroke was suspected and TTE was performed which revealed possible vegetations leading to TEE finding of quadruple-valve endocarditis. Diagnosis of Bartonella endocarditis was established based on serological tests and failure to isolate an alternative organism. The patient was successfully treated with antibiotics and did not require surgical treatment.

References

New York-Clinical Vignette-Poster Finalist
Gabriela Andries, MD

Title: Role of PCR to Diagnose Paradoxical Emboli from Porphyromonas Gingivalis Tricuspid Valve Endocarditis

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Introduction: Porphyromonas gingivalis is a gram-negative, anaerobic oral bacterium, which is widely known to cause periodontitis. Here we presented a case report illustrating tricuspid valve endocarditis and brain abscess from Porphyromonas gingivalis and how we diagnosed this bacterial infection using unconventional method.

Case Presentation: 55 year-old-man presented with tonic-clonic seizure. Physical examination revealed multiple missing teeth without sign of gingivitis or periodontitis and grade 4/6 systolic murmur at the left lower sternal border. MRI brain showed multiple small enhancing lesions in both hemispheres, most prominent in the left frontal lobe. CT-scan of chest and abdomen demonstrated numerous lesions in liver, lungs, and kidney. During his hospitalization, patient became aphasic, and subsequent brain MRI showed significant increase in the size of mass in the left frontal lobe. Transesophageal echocardiography revealed multiple large vegetations on tricuspid valve (TV) causing flail anterior leaflet and severe tricuspid regurgitation, along with large patent foramen ovale (PFO). Given the findings, paradoxical emboli from IE was highly suspected. Multiple blood cultures done beginning on admission consistently showed no growth. Patient was started on broad spectrum antibiotic Vancomycin, Ceftriaxone, and Metronidazole. Due to progressive decline in neurological status, patient underwent surgical evacuation of brain abscess. Pathology of brain tissue sample showed organizing abscess, but no growth from culture. However, PCR of brain tissue identified Porphyromonas gingivalis. After he recovered from first surgery, patient underwent TV replacement and PFO closure. Patient was switched to IV Ceftriaxone and oral Metronidazole. His condition slowly improved with resolving aphasia and right hemiparesis. Patient was discharged from the hospital on day-34 of admission with the plan to finish antibiotic for 6-8 weeks.

Discussion: TV endocarditis accounts for 5-10% of IE case and commonly associated with rheumatic heart disease, IV drug use, congenital heart disease, and foreign device in right side of heart. IE from odontogenic bacteremia has been described in the past, but to our knowledge, this is the first case report illustrating TV endocarditis from Porphyromonas gingivalis causing brain abscess from paradoxical emboli. This case especially highlights the challenge in diagnosing culture-negative endocarditis and the role of PCR of tissue sample in identifying the species. Porphyromonas gingivalis has the ability to form biofilm, locally invade periodontal tissues causing destruction of surrounding tissue, which facilitates the entry of bacteria into bloodstream and lead to endocarditis. Aside from our case, there are few other case reports of intracranial abscess due to this organism without prior signs of periodontitis. The possibility of odontogenic source of endocarditis should always be on differential when no other obvious origins of infection could be identified.

References
Title: Caliber persistent artery: A rare but interesting cause of acute massive gastrointestinal bleeding

Authors: Tehseen Haider MD1, Gaurav Bhardwaj MD1, Harish Guddati MD2, Nejat Kiyici MD2, and Hilary Hertan MD, FACG2, 1. Department of Internal Medicine, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, NY, 2. Division of Gastroenterology, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, NY

Introduction: Dieulafoy’s lesion is an uncommon but significant cause of massive, life threatening acute gastrointestinal (GI) bleeding. Its rarity and presence of no ulcerations surrounding it, with life threatening hemorrhage makes it a diagnostic and therapeutic challenge.

Case Presentation: A 56 year old male with history of Hypertension and NSAID use for chronic back pain was admitted to intensive care unit (ICU) for abdominal pain and hematemesis. His BP was 133/83 mm of hg and heart rate was 92 beats/min. Abdomen was nontender to palpation. Initial laboratory data showed blood urea nitrogen (BUN) 22 mg/dL, creatinine 1.47 mg/dL, hemoglobin 12 gm/dL, hematocrit 34.5 %, white blood cell (WBC) 17.0 k/uL and normal coagulation profile. Computed tomography (CT) of abdomen and pelvis revealed large central density in the stomach representing a blood clot. He underwent emergent esophagogastroduodenoscopy (EGD) which showed clots and food in fundus with no obvious lesions in the esophagus, body, antrum and duodenum. He was started on IV proton pump inhibitor (PPI) therapy. He had recurrent episode of hematemesis and melena with drop in hemoglobin and hematocrit (H&H) to 6 gm/dL and 18 % respectively and received two packed red blood cells (PRBC). Repeat EGD next day showed Dieulafoy’s lesion with oozing blood in the gastric fundus. Hemostasis was achieved with epinephrine injection and five hemoclips. He remained stable in ICU, but two days later he had recurrent bleeding with drop in H&H requiring five PRBC transfusions and vasopressors. Repeat EGD showed oozing blood from same site with intact hemoclips from previous endoscopy, epinephrine injection and four hemoclips were placed again followed by left gastric artery collaterals embolization. He improved clinically with no further episodes of bleeding and was discharged home.

Discussion: Dieulafoy’s lesion is responsible for ~ 1.5% of acute nonvariceal upper GI bleeding. It can be located anywhere in GI tract but most commonly is found in stomach 75-90%, mostly within 6 cm of gastroesophageal junction. It is called caliber persistent artery as its diameter is around 1-3 in mucosa which is 10 times the normal caliber of mucosal capillaries. Bleeding is mostly seen in males. Endoscopy is the diagnostic modality and treatment of choice. It appears as raised nipple or visible vessel with no ulcerations. Mechanical therapy, hemoclip placement, epinephrine injection and thermocoagulation are used to control bleeding. If endoscopy fails, angiography is used for diagnosis and treatment. Rebleeding can be treated with repeat endoscopy, angiographic embolization or surgical wedge resection. The long term rate of rebleeding is low once Dieulafoy’s lesion is completely treated.

Learning Objectives:

1. To recognize rare causes of acute massive life-threatening GI bleeding.
2. To understand pathophysiology, diagnosis and management of Dieulafoy’s lesion.
Title: Auto Brewery Syndrome induced ‘Alcoholic’ Pancreatitis

Authors: Chokshi, S, M.D., Arulhasan, M. MD, Nfonoyim, J, MD, Internal Medicine Resident, Richmond University Medical Center, NY

Introduction: Auto Brewery Syndrome also known as Gut Fermentation Syndrome is a rare yet interesting condition in modern medicine. In this condition, individuals produce endogenous alcohol levels after eating carbohydrate-rich foods due to fermentation by yeast in the gut. These high levels of endogenous alcohol have the same pathophysiology effect as exogenous alcohol on the pancreas. Thus individuals are susceptible to alcoholic pancreatitis, as seen in our patient.

Case Presentation: A 45-year-old Italian-American male known to have Auto Brewery Syndrome presented complaining of vomiting for 3 weeks, which had worsened over the past few days prior to presentation. Over the years, the patient has had multiple falls resulting in numerous nasal trauma eventually requiring corrective surgery which was complicated by sinus infections. The infections were treated with Vancomycin, Ciprofloxacin, and steroids which promoted nausea, vomiting, diarrhea and uncontrolled Diabetes Mellitus. Nine months later, Augmentin was used during a laser dental surgery which once again led to uncontrolled diabetes with weight gain (50lbs), peripheral edema and sleep apnea. In 2015 he developed recurrent seizures with slurred speech, glazy eyes, abnormal ideology, and alcohol breathe. This presentation was consistent with alcohol withdrawal symptoms. The next year he was arrested twice for driving under the influence, even though he continuously denied consuming alcohol; blood alcohol level was 42.2 mg/dL. Patient and his family were determined to find the true cause of his condition, and in 2016 a Diagnosis of Auto Brewery Syndrome was confirmed using carbohydrate challenges and timed blood alcohol levels. Patient was treated with Fluconazole and Nystatin as outpatient to treat Candida albicans as the causative organism.

Discussion: However, despite being on anti-mycotic treatment patient presented complaining of intolerable nausea, vomiting, and abdominal pain for 3 weeks prior to admission. Combining alcohol level, lipase level, radiological imaging and clinical examination diagnosis of acute alcoholic pancreatitis was made, along with pancreatic calcification of chronic pancreatitis. Acute pancreatitis despite Fluconazole meant persistent fermentation of carbohydrates leading to further investigation. Stool cultures showed Saccharomyces cerevisiae as the causative organism. Infectious disease consult recommended discontinuing Fluconazole and starting Micafungin 100 mg IV daily for broad coverage as C.glabrata or C. krusei can also contribute to gut fermentation along with Saccharomyce. As there are no guidelines for treatment, the patient was treated with Micafungin intravenously for approximately 6 weeks, until repeat fungal studies were negative.

Auto Brewery Syndrome is rare, but should be evaluated in patients with intoxication symptoms who stand by sobriety. As seen our patient, Auto Brewery Syndrome can lead to serious medical conditions along with social consequences if not treated in a timely manner. Definitive tests and studies to confirm the diagnosis need to be done and antymycotic therapy should be started based on the causative agent.
New York-Clinical Vignette-Poster Finalist
Amanda Dowden, MD

Title: Organizing Pneumonia: A Delayed Diagnosis

Authors: Amanda Dowden, MD;

Introduction: Organizing Pneumonia (OP), presents similar to infectious pneumonia, requiring a careful review of history and evaluation of disease to make a definitive diagnosis. We report a case that was misdiagnosed as bacterial pneumonia, but later recognized as organizing pneumonia. This case highlights the complexity in diagnosing organizing pneumonia and the importance of recognizing non-infectious etiologies for respiratory failure and pulmonary infiltrates.

Case Presentation: A 53-yr female with a history of bipolar disorder and multiple prior PNAs was admitted to the ICU for dyspnea and cough for three days. On presentation, patient was afebrile, hypotensive, tachycardic, and hypoxic. Physical exam notable for bilateral crackles on pulmonary exam. Initial laboratory data notable for Na of 125 mmol/L, no leukocytosis, ALT/AST of 91IU/L/94IU/L, and Lactate of 3.01 mmol/L. Initial imaging notable for extensive severe diffuse multifocal consolidative opacities throughout both lungs, with bi-lateral lower lobe consolidation on CT Chest. Patient was treated with empiric antibiotics including vancomycin, azithromycin, and cefepime and intubated for hypoxic respiratory failure secondary to multifocal pneumonia. Patient continued to have fevers with an uptrending leukocytosis in the setting of gastric aspiration. She was broadened to Zosyn. There was no improvement in lung consolidations and oxygenation, despite broad-spectrum antibiotics on HOD 10. A bronchoscopy was performed, which showed minimal secretions and BAL with no growth on gram stain. Given lack of secretions expected in extensive infectious pneumonia, patient underwent a video-assisted thoracoscopic (VATS) and L wedge lung biopsy. Pathology revealed early OP and no cellular evidence of an infectious process. She was started on high dose steroids with improvement in her respiratory status. She was successfully extubated and discharged on a steroid taper.

Discussion: Recognition of OP can be difficult because the clinical presentation often mimics that of infectious pneumonias and interstitial lung disease. The patient described here had a history of pneumonias and high NG tube output concerning for aspiration pneumonia. However, there was no improve with antibiotics prompting a lung biopsy to evaluate for non-infectious etiologies. OP is a clinicopathologic syndrome described as a result of lung injury, most commonly, from lung infections, drugs, connective tissue disorders, or toxic environmental exposure. It is classified as Cryptogenic Organizing Pneumonia (COP) if the initial insult is unknown. OP is a rare disease, with an unknown incidence and prevalence because a lung biopsy is essential for a diagnosis. The clinical symptoms of OP include dyspnea, cough, fever, and malaise over several weeks. The most common CT manifestations include multiple, peripheral, patchy opacities, ranging from ground glass to consolidation distributed bilaterally. A high clinical suspicion for organizing pneumonia is crucial in individuals. Delay in diagnosis, as in this patient, can lead to persistent symptoms and extended hospital stay.

References

Title: CMV Spindle Cell Pseudotumor in an Immunocompetent Patient: A Rare Entity

Authors: Zahava Farkas, DO Lavneet Chawla, MD Shalom Frager, MD Brad Dworkin, MD

Introduction: Cytomegalovirus (CMV) infection of the colon is common in both immunocompetent and immunocompromised patients. However, CMV-induced pseudotumor or mass-like lesions in the colon are a rare entity in the immunocompetent population. We report a case of CMV related spindle cell pseudotumor in an immunocompetent patient who presented with severe lower gastrointestinal bleeding in the setting of systemic anticoagulation for LVAD therapy.

Case Presentation: A 63-year-old man with no past medical history was transferred to our institution for management of cardiogenic shock. On arrival he was placed on venous-arterial extracorporeal membrane oxygenation (ECMO). He had multiple episodes of diarrhea with microscopic blood on stool occult testing. Computed tomography (CT) showed pan-colitis, most severe around the cecum. He later had a left ventricular assist device (LVAD) placed at which time an exploratory laparotomy was performed to evaluate for ischemic bowel; none was found. While on systemic anticoagulation to prevent LVAD pump thrombosis, he began having multiple episodes of hematochezia. Esophagogastroduodenoscopy (EGD) and colonoscopy were performed. Multiple firm masses (ranging from 0.5 to 1.5 cm in largest diameter) were noted in the ascending colon and hepatic flexure. Repeat colonoscopy with additional tissue sampling was performed due to recurrent bleeding. Initial pathology on the biopsies was suggestive of gastrointestinal stromal tumor (GIST) with a high mitotic index and he was started on Imatinib (Gleevec). Due to persistent lower GI bleeding with hemodynamic instability, he underwent a hemi-colectomy with end-ileostomy. The final pathology report on the colonic biopsies revealed a diagnosis of CMV-associated spindle cell pseudotumor. Histopathology showed ulcerated, inflamed colonic mucosa with two cellular spindle cell fragments. The cells were morphologically consistent with CMV inclusions and negative for CD117, DOG-1, smooth muscle marker and CD34, excluding the diagnosis of GIST. Immunostain for CMV was positive in these cells. Workup up for underlying immunodeficiency was negative. The patient recently received a heart and kidney transplant and is planned for possible reversal of his ileostomy.

Discussion: CMV infection is common in both immunocompetent and immunocompromised patients and may be the result of primary infection or a secondary reactivation due to impaired T-cell function. The GI tract is a frequent site of CMV infection and most commonly manifests as erosive colitis or esophagogastritis with prototypical punched out ulcerations and erosions on endoscopy. Inflammatory pseudotumors associated with CMV virus in an immunocompetent patient, as in the above reported case, is rare. CMV-associated pseudotumors should be considered in the differential of tumorous lesions of the colon, even in immunocompetent hosts. Maintaining clinical suspicion is key, as it is amenable to intravenous antiviral therapy and may prevent unnecessary pharmacologic or surgical interventions.

References
New York-Clinical Vignette-Poster Finalist
Michael Grzeskowiak

Title: Cyclic Cushing’s Disease mimicking PCOS

Authors: Michael Grzeskowiak MD, Shitij Arora MD FACP, Department of Medicine, Division of Hospital Medicine, Montefiore Medical Center, Bronx, NY

Introduction: Intermittent or periodic hypercortisolism, as seen in Cyclic Cushing’s disease (CCD), is an interesting and a rare phenomenon. The clinical presentation varies from a single outstanding symptom, such as hirsutism, to a complex clinical syndrome affecting nearly all systems; thus, making it a diagnostic challenge. We report CCD in a patient previously identified as having polycystic ovarian syndrome (PCOS) who presented with intermittent facial swelling.

Case Presentation: A 37-year-old female with a past medical history of PCOS (obesity, hirsutism, secondary amenorrhoea) and multiple psychiatric hospitalizations for anxiety who presents to the hospital with facial swelling, facial erythema, palpitations, and headaches for 1 day. She denied diarrhea. She described recurrent similar symptoms for the past 4 years. Her only medication included Metformin for PCOS. Review of her self-portrait photographs showed a near doubling of her facial measurements during episodes. Upon exam, she was an obese female with moderate facial erythema, moon facies, hirsutism, negative Darier’s sign, and no abdominal striae. We ruled out carcinoid syndrome (normal 24h urine 5 HIAA), pheochromocytoma (normal plasma metanephrines), and mastocytosis (negative Darier’s sign, normal tryptase levels); all of which can present as intermittent facial swelling. Her laboratory data showed a low normal AM cortisol and normal ACTH levels but review of her records over the past year showed a variability in 24h urine free cortisol with at least 3 peaks and 2 troughs. We suspected CCD. A repeat MRI Brain (she had a normal MRI brain within the past year) was performed which showed a 4mm pituitary adenoma. She is now scheduled for a trans-sphenoidal hypophysectomy.

Discussion: Cyclic Cushing’s disease is rare and often underdiagnosed. Indeed, studies have shown that the spontaneous remission in patients with Cushing’s disease could likely be explained by cyclical hormonogenesis[1]. Any etiology of Cushing’s disease can produce cyclic patterns of hypercortisolism. There are limited explanations for the periodicity but some suggest hypothalamic causes as mediators of pulsatile ACTH hypersecretion including neurotransmitters such as noradrenaline, dopamine, acetylcholine and GABA[2]. We suspect the multiple admissions she had for anxiety could have been due to hypercortisolism from CCD, however, it can also be argued that the anxiety was resulting in hypercortisolism through the hypothalamic regulation of the anterior pituitary[3].

In patients with hypercortisolism phenotype and an initial negative biochemical profile repeating measurements of urinary cortisol is a reliable test when CCD is suspected[4,5]. Dexamethasone suppression tests may give spurious Results: owing to spontaneous variability in serum cortisol at the time of testing. When CCD is biochemically confirmed, further imaging and laboratory studies should be guided by the ACTH dependency as indicated by the levels.

References


New York-Clinical Vignette-Poster Finalist
Garima Handa

Title: A Young Male Presenting With Complete Heart Block Due To Lyme Carditis: A Case Report

Authors: Garima Handa, MD; Salman Haq, MD

Introduction: Lyme disease is a tick-borne disease caused by the spirochete Borrelia Burgdorferi. It encompasses several systemic manifestations, of which cardiac involvement is relatively rare. It usually presents as varying degree of heart blocks and/or myopericarditis. The spirochete infects the conduction system around the AV node, membranes of the heart, valves, muscle, and the vasculature. We present a case of 31-year-old male who presented with complete heart block due to lyme carditis that resolved with antibiotics.

Case Presentation: A 31 year old male with no significant past medical history presented with intermittent palpitations and exertional dyspnea. His symptoms started a week prior to admission, at which time he also had a near syncopal episode. It was preceded by palpitations and lightheadedness and followed by loss of consciousness. He was only able to walk 1-2 blocks before experiencing shortness of breath. He denied chest pain, orthopnea, leg swelling, headache/dizziness, tick bites or a family history of sudden cardiac death. He reported a history of frequent visits to upstate NY. The vitals and physical exam were within normal limits. Electrocardiogram showed a third degree AV block with wide QRS, R'R pattern and T wave inversion in the precordial leads. Cardiac enzymes were within normal limits and the pro-bnp was 479. Transthoracic echocardiogram, cardiac MRI and left heart catherization were negative for any structural abnormalities, except mild mitral and tricuspid regurgitation. Given the history of frequent visits to upstate New York, lyme carditis was suspected. The lyme titers, IgG and IgM, were positive. The patient was started on IV Ceftriaxone 2gm for 21 days. Over the course of the treatment, conduction improved to normal sinus rhythm. Complete resolution of symptoms occurred with no further episodes of syncope, palpitations or exertional dyspnea.

Discussion: This case serves to be a unique case in that the only manifestation of lyme disease was the carditis symptoms, which alone is very rare. In the United States, lyme carditis occurs in about 1% of the patients with lyme disease. The deaths from it are rare as the antibiotic therapy for erythema migrans (EM) and other symptoms in the early-localized disease prevents the progression to carditis. Unfortunately, not all the patients with lyme carditis have symptoms of early disease. For example, only 40% of the patients with carditis have EM rash. Furthermore, of all the carditis symptoms of lyme disease, the third degree heart block is the most rare and severe form. Literature review showed only 45 published cases of the third degree heart block associated with lyme carditis. CDC has reported 3 deaths from November 2012 to July 2013 in patients with unrecognized lyme carditis. This case highlights the need for prompt recognition and early antibiotic therapy, as it can be fatal if untreated.
New York-Clinical Vignette-Poster Finalist  
Isaac Huml, MD

Title: Hemophagocytic lymphohistiocytosis in a patient with SLE and B cell lymphoma

Authors: Isaac Hum MD, Bhashkar Madhira MD, Bhavana Jami DO, Arslan Khan MD. Department of Internal Medicine, SUNY Upstate, Syracuse, New York

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is a rare clinical entity in adults that is usually secondary to autoimmune disease or hematological malignancies, resulting in dysregulation of the immune system. Here we describe a case of a young woman with systemic lupus erythematosus (SLE) and Sjogren's who developed HLH and was ultimately diagnosed with a rare B-cell lymphoma.

Case Presentation: A 44-year-old female presented to our hospital with persistent fevers, chills, headaches, nausea and vomiting for 15 days prior to presentation. She presented to her primary care one week before and was given a 7-day course of levofloxacin for bronchitis, but continued to have persistent fevers and chills. On presentation she complained of joint pain and was noted to have moderate arthralgias of small and large joints as well as the neck. There was originally concern for an infectious etiology, specifically meningitis, as well as a lupus flare. Her labs showed moderately elevated inflammatory markers (ESR, CRP), but lumbar puncture was unrevealing and complement levels were normal. She received CT imaging of her head, thorax and abdomen, which was significant only for hepatosplenomegaly. After several days, all of her cultures yielded no growth and she started to develop worsening cytopenia. All cell lines dropping steadily with no laboratory or histological signs of hemolysis. The patient continued to spike intermittent fevers above 39°C. There was increasing concern for HLH; therefore, levels for ferritin, triglycerides and IL-2 receptor were obtained. The Results: were markedly abnormal meeting diagnostic criteria for HLH. A bone marrow biopsy was obtained for confirmation of the diagnosis. This showed macrophage activation with focal hemophagocytosis consistent with the diagnosis of HLH. Bone marrow biopsy also revealed a large B-cell lymphoma, which was confirmed with cytogenetics.

Discussion: This case reemphasizes the importance of a bone marrow biopsy when evaluating a patient with HLH. This patient has moderately severe SLE, which could easily be considered the causative factor of this patient’s disease. However, because of the input from the bone marrow biopsy we have a different source that we can attribute to causing this patient’s HLH and were able to diagnose this patient with a treatable hematological malignancy.
Title: CEREBROSPINAL FLUID LEAK: A Diagnostic Algorithm For A Rare Illness

Authors: Rami Jabbour¹ MD, Shaheen Alvi¹ MD, Charbel Ishak¹ MD, Sridhar Chilimuri¹ MD

Introduction: Cerebrospinal fluid leak (CSF leak), although rare, is a well-recognized cause of headache. The annual incidence of spontaneous CSF leak was estimated to be 5/100 000. Since headache is a common condition it is unclear when to suspect CSF leak requiring additional workup. We propose a diagnostic algorithm based on our experience. We present four illustrative clinical cases to explain this algorithm.

Case Presentation: Case 1: A 57 year-old man presented with postural headache associated with neck, shoulder and back pain. Six months prior to presentation, he had history of a closed head trauma. Computed tomography (CT) Cisternography revealed CSF leak in left maxillary and frontal sinuses.

Case 2: A 50 year-old immunocompetent woman presented with headache, nausea and vomiting. She was diagnosed to have Cryptococcal meningitis. She reported a remote history of closed head trauma and on further questioning reported intermittent rhinorrhea. CT Cisternography revealed right nostril CSF leak.

Case 3: A 55 year-old man presented with postural headache and photophobia one-month after cervical laminectomy. A diagnostic spinal tap revealed meningitis. CT cisternography revealed a 7x6x15 cm fluid collection overlying the cervical spine compatible with CSF leak.

Case 4: A 49 year-old woman presented with postural headache. She had recurrent spontaneous right ear fluid leak and right nostril rhinorrhea of 6 months duration. There was no history of trauma. CT Cisternography revealed CSF leak.

Discussion: Discussion: Postural headache in an important feature of CSF leak. However, by itself, lacks the sensitivity or specificity to suspect a CSF leak. Postural headache with a history of head trauma/surgery appears to increase the possibility of a CSF leak. The presence of postural headache, with history of head trauma and the presence of unusual meningitis should prompt a search for CSF leak. It is well recognized that postural headache with rhinorrhea should prompt search a CSF leak. CT cisternography remains the gold standard for diagnosing CSF leak. CSF leak can lead to life threatening complications including meningitis in 25-50% of cases.

Conclusion: Both spontaneous and head trauma related CSF leaks are rare conditions. However, when unrecognized could lead to life threatening complications. Early recognition could lead to interventions with excellent outcomes. Postural headache, history of trauma/surgery (recent and remote) and the presence of unusual causes of meningitis should prompt work up for CSF leak. CT cisternography remains the gold standard for diagnosing CSF leak.
Title: An Exaggerated Immune Response

Authors: Khan, Mahmuda; Solomon, Philip

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is a disorder manifested by excessive immune activation. The two most common triggers are immune deficiency and immune activation, such as infection. Persistent activation of cytotoxic lymphocytes and natural killer (NK) cells leads to a cytokine storm via high levels of interferon gamma, tumor necrosis factor, and interleukins (IL). This cascading response can lead to rapid multiorgan failure. HLH is more common in infants than adults, and carries significant mortality risk without prompt treatment. We present an atypical case of suspected primary HLH in an elderly patient with a subacute presentation.

Case Presentation: A 62-year-old male with cryptogenic cirrhosis presented with nonspecific complaints of fatigue, unintentional weight loss, and nausea for five months. He was found to be hypotensive and febrile. Exam was notable for splenomegaly. Labs were significant for pancytopenia, with hemoglobin of 8.2 g/dL, platelets of 65,000/mcL, and absolute neutrophil count of 400/mm^3. Patient was admitted to the intensive care unit and initially treated for septic shock; however, extensive infectious workup was negative. His hospital course was complicated by continued fevers with pancytopenia. Autoimmune and vasculitis workup was unremarkable. CT of the chest, abdomen, and pelvis was unrevealing for underlying infection or malignancy. Bone marrow biopsy was performed, which showed normal trilineage maturation and evidence of hemophagocytosis. Follow-up labs revealed a ferritin of 7960 ng/mL and elevated IL-2 level of 17714, in addition to hypertriglyceridemia and hypofibrinogenemia. Given the patient met seven out of eight diagnostic criteria for HLH, he was started on treatment with etoposide and dexamethasone. The patient slowly improved, and was discharged with outpatient hematology follow-up.

Discussion: HLH is an acute illness that commonly presents with hepatomegaly, lymphadenopathy, neurological symptoms, and rash [1]. Secondary HLH is associated with underlying malignancy and rheumatologic disorders, while primary HLH is due to gene mutation. Diagnosis is made by identification of HLH-associated gene mutation or five of the eight criteria of fever, splenomegaly, cytopenias, hypertriglyceridemia/hypofibrinogenemia, hemophagocytosis in bone marrow/spleen/lymph node/liver, low NK cell activity, elevated ferritin, and elevated CD25/IL-2. Treatment includes weekly administration of dexamethasone and etoposide. In secondary HLH, treatment of the underlying trigger, such as infection or malignancy, is essential. When patients fail to improve, allogeneic hematopoietic cell transplantation is offered.

HLH should be considered in patients with persistent fevers and pancytopenia. Given no underlying precipitating factor was identified, our patient may have primary HLH, a rare presentation for an adult. He will benefit from follow-up genetic testing.

This is a unique case of a subacute presentation of HLH. Patients often present with rapidly progressive multiorgan failure. The delay in diagnosis and treatment was due to our patient’s unusual presentation. This case is an important example of an atypical subtle presentation of this rare disease.

References
New York-Clinical Vignette-Poster Finalist
Tien Lau, DO

Title: In a neurological differential dilemma? Think Left Atrial Myxoma.

Authors: Tien Lau DO, Yashodhan Chivate MD, Myat Myo MD, John T. Mather Memorial Hospital, Port Jefferson, NY.

Introduction: Left atrial myxomas (LAM) present in a protean fashion with a variety of non-specific signs and symptoms (Table 1). The variety of presentations can make LAM a daunting diagnostic challenge and any delay in the appropriate treatment carries several complications (Table 2).

Case Presentation: On physical examination, she appeared to be in no acute distress, BP 146/94 mmHg, HR 90 BPM, 97.8F, 94% O2 saturation on room air. Neurological exam revealed 4/5 strength in her left arm and leg, with preserved sensation and reflexes. No heart murmurs were audible. A CT scan of the head was negative for acute intracranial pathology, but a MRI of the brain with and without contrast revealed several foci of impaired diffusion in the right frontal lobe and right post-central gyrus, consistent with multiple acute infarcts (Figure 1). She was started on anticoagulation and underwent an extensive workup to identify the underlying cause of her cerebral vascular accident (Table 3). Laboratory studies for vasculitis, hypercoagulability, and autoimmune disorders along with cerebrospinal fluid analysis for Multiple Sclerosis, and Lyme’s all returned negative by the third day of admission. A transthoracic echo with bubble study was performed, and while there was no right to left shunt, a large mass (3.7 cm x 2.4 cm) adherent to the anterior leaflet of the mitral valve and intra-atrial septum was identified (Figure 2) consistent with left atrial myxoma. The anticoagulation was stopped, and our patient was subsequently transferred for emergent cardiothoracic surgery for removal of the myxoma.

Discussion: LAM can manifest as commonly observed symptoms and signs (Table 1), but may present more discretely as in our case and make the diagnosis challenging. LAM causes symptoms by inducing mitral valve disease or obstructing blood outflow, creating secondary pulmonary hypertension and/or heart failure. In addition to impeding the circulation, LAM may release tumor fragments or thrombi into the systemic circulation, often causing neurological deficits as initial complaints. This case elucidates the potential for severe complications (Table 3) in unusual LAM presentations without immediate medical intervention.
Title: Labetalol can be a Heart Breaker

Authors: Kyu-In Lee¹, Anoshia Raza¹, Dipak Chandy¹,¹ New York Medical College, Westchester Medical College

Introduction: Labetalol is an alpha and nonselective beta adrenergic receptor blocker. In addition to its negative chronotropic and inotropic effects, it is also a powerful peripheral vasodilator. When given intravenously (IV), the proportion of beta:alpha antagonism of labetalol is 6.9:1. Beta antagonists are generally contraindicated in asthmatics as they can cause bronchospasm. However, severe respiratory acidosis secondary to such bronchospasm and subsequent cardiac arrest has rarely been reported with the administration of labetalol.

Case Presentation: A 57 year old man with a 15 pack-year smoking history, type A aortic aneurysm status post Bentall procedure four years ago, hypertension and mild asthma presented with shortness of breath and chest pain. Computed tomography angiogram (CTA) was concerning for a possible type B aortic dissection involving his ascending aorta, aortic arch and left subclavian artery. He was transferred to the intensive care unit where his mean arterial pressure (MAP) was between 110-120 mmHg and oxygen saturation was 99% on 3L/minute via nasal cannula (NC). Examination was significant for an obese habitus while the pulmonary exam revealed no wheezing. Initial labs were all within normal limits. An arterial blood gas (ABG) collected on oxygen at 3L/minute via NC was pH7.38, pCO₂ 49 mmHg and pO₂ 84 mmHg.

In view of the hypertension in a possible type B aortic dissection, metoprolol 5mg IV was administered followed by labetalol infusion at 0.5mg/min. Approximately 30-40 minutes later, patient began experiencing severe shortness of breath, MAP decreased to the 50s and he became bradycardic to 33 beats/minute. He subsequently went into a systolic cardiac arrest. Return of spontaneous circulation was achieved within 10 minutes. Immediately post-intubation, his ABG was pH6.98, pCO₂ 110 mmHg and pO₂ 84 mmHg.

Cardiothoracic surgery interpreted the CTA as a residual effect of the Bentall procedure. Transthoracic-echocardiography revealed moderate left ventricular (LV) hypertrophy, hyperdynamic LV systolic function with an ejection-fraction of 75%, mildly dilated left-atrium, dilated right-atrium and pulmonary-arterial-systolic-pressure (PASP) in the range of 33-48 mmHg. Electrocardiogram revealed normal sinus rhythm.

Pulmonology was urgently consulted for further evaluation of the acute hypercarbic respiratory failure. Initial assessment of the patient showed evidence of severe bronchospasm (large difference between peak and plateau pressures) and air-trapping (auto-positive-end-expiratory-pressure of 13cm H2O). He was started on high-dose IV steroids, bronchodilators and settings on the ventilator were adjusted to reduce air-trapping. Over the next 48 hours, patient’s condition dramatically improved. He was extubated and discharged on an oral steroid taper and inhaled bronchodilators.

Discussion: Our case is remarkable for a temporal relationship between the infusion of a nonselective beta-blocker and the rapid development of severe bronchospasm in a known asthmatic resulting in severe respiratory acidosis, culminating in cardiac asystole. Nonselective beta agonists such as labetalol should be used with a great deal of caution in patients with asthma.

References
New York-Clinical Vignette-Poster Finalist
Matthew C Lee, MD

Title: A Rare Initial Presentation of Lung Cancer Metastasis

Authors: Matthew Lee, Internal Medicine Resident, Department of Medicine, North Shore University Hospital-Long Island Jewish Medical Center, Manhasset, NY, Shikha Sheth, Internal Medicine Resident, Department of Medicine, North Shore University Hospital-Long Island Jewish Medical Center, Manhasset, NY, Syed Ahmad, Assistant Professor at the Department of Medicine, North Shore University Hospital, Manhasset, NY

Introduction: Lung cancer is both the second most common malignancy and the leading cause of cancer-associated mortality. It typically metastasizes to the brain, bone, liver and adrenal glands. In rare cases, lung cancer may metastasize to the skin and has been reported in 1-12% of lung cancer cases (1-4). Overall, in men who present with skin metastases, the most common primary site is lung cancer. Furthermore, 20-60% of these cases presented initially with cutaneous findings before the diagnosis of primary lung cancer was made (3). Here we present a rare initial presentation of skin metastases in zosteriform pattern from a primary lung adenocarcinoma.

Case Presentation: A 78 year old male with a history of atrial fibrillation, type 2 diabetes and former significant tobacco use, presented with a one month history of left sided neck pain. The pain was progressive and he subsequently developed left upper extremity swelling along with a rash involving the neck, left anterior chest and back. In the hospital, the patient reported no recent infections or travels, fevers, dyspnea, or weight changes. On physical exam, patient was noted to have a firm, indurated mass on his left upper back along with a vesicular rash in a T3-4 dermatomal pattern concerning for shingles. Laboratory findings were significant for leukocytosis, normal calcium level and HIV was negative. Ultrasound confirmed a thrombus in the left internal jugular vein along with left cervical, supraclavicular and axillary lymphadenopathy, and he was started on Lovenox for anticoagulation. Due to the unknown cause of the hypercoagulability, a CT Chest was performed which revealed a right upper lobe heterogenous enhancing mass concerning for malignancy. A skin biopsy was subsequently performed which revealed primary adenocarcinoma of the lung. Immunohistochemical staining was positive for TTF-1, a marker that is sensitive and specific for primary adenocarcinoma, as well as PD-L1, CK20 and KRAS mutation.

Discussion: This is a rare case of metastatic lung adenocarcinoma presenting initially with skin metastases in a zosteriform dermatome pattern. Commonly, skin metastases are seen on the chest, back and neck but usually are not distributed in a dermatome area (5). Our case is similar to a previously reported case report by Li et. al (6) in which the proposed the possible pathogenesis of the zosteriform skin metastasis may be due to emboli from the cancer that causes lymphatic congestion and blockage. This could then lead to the skin lesions and left upper extremity swelling as seen in this patient. This case highlights the importance of recognizing new skin findings in high risk patients such as those with significant smoking history as one of the first clinical manifestation of a possible occult malignancy.

References


Title: Post Polio Syndrome presenting as Acute Respiratory Failure. Expanding the differential diagnosis of restrictive lung disease

Authors: Fidel Martinez, Resident, Department of Medicine, SUNY Upstate, Syracuse, NY, David F Lehmann, MD, PharmD, Distinguished Service Professor of Medicine, Department of Medicine, SUNY Upstate, Syracuse, NY

Introduction: Post Polio Syndrome is the delayed sequelae of polio infection. The incidence ranges from 22-68% in survivors, and may too easily be attributed to common comorbidities in older patients. Diagnosing can be challenging or missed. On literature review, no case reports were found of acute respiratory failure as initial presentation.

Case Presentation: A 90 y/o male presented with progressive weakness and dyspnea on exertion over one month. On presentation chest radiography was unremarkable and his oxygen saturation was 88%. Hypoxemia progressed and was associated with hypercapnia via ABGs: pH 7.28, pCO2 61.6, pO2 74.7 and bicarbonate 28.3. CTA did not show pulmonary embolism. He was placed on bilevel positive airway pressure (BiPAP) and transferred to the intensive care unit. Respiratory panel was positive for rhino/enterovirus and he was treated empirically for post viral bacterial pneumonia. Echocardiogram showed an ejection fraction of 55 to 60% and Grade II diastolic dysfunction. Despite treatment, patient continued to depend on BiPAP at nights. Attempts to wean him off BiPAP failed as he became delirious and agitated. Pulmonary function test could not be performed due to acuity of his condition. An EMG showed chronic, widespread disorder of the motor neurons or their proximal axons, without evidence of ongoing axon loss, compatible with history of polio. Chronic diaphragmatic weakness most likely had became pronounced due to his age and recent upper respiratory infection. The patient had been diagnosed with Polio as a child, and recalled having the some respiratory sequela, for which he was given a harmonica to exercise his lungs. He had recovered fully, had an active and productive life, and had forgotten of the disease. He was kept on BiPAP at nights, his delirium improved. He was discharged to a rehab facility on BiPAP and was encouraged to keep playing the harmonica.

Discussion: This case illustrates the importance of obtaining a complete medical history to broaden the differential diagnosis to account for the patient’s age.

References

New York-Clinical Vignette-Poster Finalist
Fitsumberhan Medhane, MD

Title: Where There is a Smoke There is Thrombosis

Authors: Fitsumberhan Medhane, MD, MPH; Hanane Ben Faras, MD; Amit Kakkar, MD

Introduction: Carbon monoxide (CO) poisoning is one of the most common types of poisoning. It can be underdiagnosed as it can present with non-specific symptoms: nausea, vomiting, headache, chest pain and fatigue. We present a case study of a couple who presented with CO poisoning to emphasize the importance of early recognition of CO poisoning and its potentially fatal pro-thrombotic complications.

Case Presentation: Case 1: 53-year-old woman with hypertension and anxiety was transferred to our facility for CO poisoning. A few days prior to presentation, the patient was experiencing headache, dizziness and fatigue. She was evaluated at an outside ED and was diagnosed with viral syndrome. The following day she was found down in her apartment. The patient recalled turning on the gas furnace earlier that week. She was anxious, confused and complaining of chest pain. Initial CoHgb was 14.3% (normal 0-5%). Troponin was elevated and the EKG showed ST depression in II, III, aVF, and V4-V6, and ST elevation in aVR. She was treated for acute coronary syndrome and transferred to our facility for hyperbaric oxygen treatment. The patient received hyperbaric treatment with a significant improvement in her mental status but continued to have chest pain. She underwent cardiac catheterization that revealed significant one vessel disease of mid LAD and moderate distal CFX disease. A single drug eluding stent was placed in the LAD. The patient eventually improved and was discharged home.

Case 2: 53 year-old male with hypertension who presented with his partner (described in case 1) with CO poisoning. The patient was initially confused but later gave history of progressively feeling weak and dizzy a few days prior to presentation. His mental status improved after receiving hyperbaric treatment, but later he reported bilateral thigh pain, left leg pain and pleuritic chest pain. Physical exam was unremarkable. Labs and imaging were consistent with rhabdomyolysis, bilateral DVT and sub-massive PE. Otherwise, troponin, BNP, EKG and coagulopathy studies were all unremarkable. He was treated with anticoagulation and discharged on Apixaban.

Discussion: These cases demonstrate acute CO poisoning with thrombotic complications. Carbon monoxide is directly toxic to the mitochondria and endothelial cells, consequently triggering ischemia, as well as arterial and venous thrombosis. The risk of DVT is 3.85 times higher in patients with CO poisoning than in the general population. Common presentations of CO poisoning include dizziness, confusion, headaches and flu-like symptoms. Larger exposure can lead to significant neurological, myocardial, renal and pro-thrombotic complications.

Clinicians should be trained to recognize the early signs of CO poisoning and to detect life threatening complications (hypoxic and pro-thrombotic). Thorough history taking can prevent discharging patients back to harmful environments which can further worsen their prognosis.

New York-Clinical Vignette-Poster Finalist
Ahmed A Mohammad, MD

Title: Longitudinally Extensive Acute Transverse Myelitis ‘ATM’ following Tdap vaccination

Authors: Ahmed Mohammad MD, Sheelan Karim MD, Roxana Lazarescu MD

Introduction: Acute Transverse Myelitis is a rare acquired neuro-immune spinal cord disorder that can present with rapid onset of weakness, sensory alterations, and bowel or bladder dysfunction. It can be an isolated entity or a manifestation of other neuroinflammatory conditions.

Case Presentation: Patient is a 40 year-old male with no past medical history who presented with bilateral lower extremity weakness, unbalanced gait and numbness starting at the level of the waist which all started on the day of admission. He also reported difficulty urinating for one day.

He received tetanus vaccine 9 days prior to presentation.

Patient was initially mildly febrile at 38.5 Physical examination revealed a sensory level starting at the level T8, Motor power was 5/5 in both lower extremities with bilateral hyperreflexia and clonus that was more pronounced on the right side.

Sensory and motor functions were intact in the upper extremities and all cranial nerves were intact.

CT scan of the head showed no acute intracranial pathology, MRI of the whole spine showed signal abnormality throughout the cervical and upper thoracic cord with faint enhancement.

CSF analysis showed slightly elevated CSF protein with no oligoclonal bands preset. VDRL, Enterovirus by PCR, Toxoplasma IgG, West Nile Virus Abs, HSV 1&2 DNA PCR and Cryptococcal Ag were all negative.

Patient was started on pulse steroids methylprednisone 500 mg IV BID. with symptoms improvement after the first dose of IV pulse steroids by his 3rd day of admission, all his symptoms resolved but he continued to show mild residual hyperreflexia and clonus on neurological examination. On outpatient follow up, he reported inability to ejaculate as his last remaining deficit.

Discussion: Our patient met diagnostic criteria for ATM as per the Transverse Myelitis Consortium Working Group ‘2’

The absence of Multiple sclerosis like lesions on the Brain MRI, the absence of oligoclonal bands in the CSF and CSF pleocytosis all made this diagnosis highly unlikely.

He also failed to meet the diagnostic criteria of Neuromyelitis optica which is very well known to cause Longitudinally extensive ATM, The negative serology Results: coupled with the negative bacterial, viral, and fungal culture Results: on the cerebrospinal fluid excluded the possibility of an infectious cause.

ATM is a rare condition, with an incidence of about 1-8 per million per year ‘3’ and it is even rarer to be reported after vaccination. It has been reported that ATM can occur after administration of several types of vaccines including, Hepatitis B, Tdap, oral poliomyelitis vaccine, Japanese encephalitis vaccine, cholera vaccine, typhoid vaccine, rabies vaccine, and seasonal influenza virus vaccine ‘4 - 5’
In a recent literature review, 37 cases of ATM following administration of various vaccines ‘6’, very rare but significant.

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New York-Clinical Vignette-Poster Finalist
Anthony Pasquarella, MD

Title: CMV Thrombosis: A rare constellation of thromboembolic findings in an immunocompetent patient.

Authors: Anthony Pasquarella, MD1; James Ciancarelli DO1; Anthony Calio, MD1; David Well MD2, NYU Winthrop Hospital Department of Internal Medicine1, Department of Radiology2

Introduction: Viral infections can cause a transient hypercoagulability. Cytomegalovirus (CMV) is one virus that causes thrombosis in both immunocompromised and more rarely immunocompetent patients. Here we present the first immunocompetent patient that has thrombotic features found in both patient populations.

Case Presentation: 60-year-old female presented with 2 weeks of fever, night sweats, weight loss, left upper quadrant (LUQ) abdominal pain and malaise. Her history includes hypertension, hyperlipidemia, hypothyroidism, sarcoidosis, and gastroesophageal reflux disease. Initial examination, was notable for mild tenderness to palpation in the LUQ and linear ecchymosis on her upper thighs bilaterally, consistent with superficial thrombophlebitis. Her vital signs were notable for a temperature of 100.3°F that increased to 102.5°F within 24 hours of admission.

Labs were significant for a WBC 12.0 k/uL, CRP 25.36 mg/L, ferritin 353 ng/mL. A CT scan of the abdomen showed an enlarged spleen measuring 15.2cm with an area of infarction. A lower extremity doppler was ordered, revealing left gastrocnemius venous thrombosis and subsequent CTPE showed pulmonary embolus within the right lung.

Outpatient labs showed an elevation in CMV IgM and IgG as well as elevation in EBV IgM and IgG. PCR revealed a positive CMV and negative EBV. After discharge the patient presented to her hematologist, and was found to be heterozygous for Factor V Leiden.

Discussion: Acute CMV infections are associated with individual venous thromboemboli. In those who are immunocompetent, vascular phenomena is the least likely complication to develop. Where the thrombosis occurs is dependent upon immunological state; immunocompetent individuals are more likely to develop splenic vein thrombosis and subsequent infarct when compared to immunocompromised patients, who have a higher incidence of thromboembolic phenomena. It has been recommended that for those with a febrile illness and splenic infarction, an active CMV infection be considered. Curiously for those with VTE and an acute CMV infection, there is a much higher incidence of underlying hereditary thrombophilia. While the correlation for transient hypercoagulability and CMV infection is apparent, the pathophysiology is poorly understood. It has been suggested that CMV induces a transient elevation of anti-cardiolipin antibody, thereby increasing risk of thrombosis. Others have speculated that it is due to vascular endothelium damage as CMV directly enters the endothelium and release factors that activate the coagulation cascade. Another explanation is that CMV can increase thrombin production and decrease the production of prostaglandin and interleukin-2, which increases platelet adhesion.

Regardless of the mechanism it is important to recognize the association between CMV and venous thrombosis. Patients with repeat thrombosis may be placed on lifelong anticoagulation, which can lead to adverse events. By recognizing CMV as the cause, the clinician may consider early discontinuation of anticoagulation to prevent unnecessary anticoagulation.
Introduction: Thymectomy is an accepted treatment modality for myasthenia gravis. However in rare instances it has not been demonstrated to be a total preventative or curative measure for myasthenia gravis. Patients who undergo thymectomy should be evaluated for autoimmune conditions associated with thymomas. We discuss a case where a thymectomy did not prevent potentially life threatening complications.

Case Presentation: A 69-year-old Asian female presented with new onset progressively worsening ptosis, dyspnea and dysphagia along with profound weakness with exertion. The patient was admitted to the medical intensive care unit due to potential airway compromise. The patient was found to have positive acetylcholine receptor and binding antibodies confirming a diagnosis of type II-A myasthenia gravis. The patient responded to treatment with high dose corticosteroids and pyridostigmine avoiding the need for mechanical ventilation. After further investigation it was discovered the patient previously underwent a thymectomy for an incidentally discovered type B1 thymoma. Post-operative CT scan did not demonstrate residual mediastinal mass or possible metastatic lesions. The patient’s symptoms resolved with treatment and the patient went into remission.

Discussion: An integral component of treatment for myasthenia gravis related thymomas is thymectomy. A review of the literature reveals that thymectomy and thymoma excision are not entirely preventative for myasthenia gravis development. Previously asymptomatic patients, as the one discussed above, develop generalized myasthenia gravis despite undergoing thymectomy. Excluding cases with clinical or radiographic evidence of tumor recurrence, the disease symptom onset has been documented from six days to ten years post-thymectomy. The mean interval of disease onset post-thymectomy was noted to be greater than eighteen months. Previous reviews estimated myasthenia gravis symptoms manifest in 2% of previously asymptomatic patients. These patients tend to respond well to standard treatment modalities with a generally optimistic prognosis. Published studies noted an 83% remission rate in patients treated with steroids only, 40% in those treated with anticholinesterase inhibitor alone and 86% with anticholinesterase inhibitors and steroids. Patients who develop myasthenia gravis earlier post thymectomy have a better prognosis. The mechanism of post-thymectomy myasthenia gravis development has yet to be fully elucidated. Research has demonstrated the expulsion of both CD4+ and CD8+ T-lymphocytes from the thymus prior to thymectomy. T-lymphocyte expulsion and persistence has been documented in patients with thymoma. It is theorized that expelled potentially autoreactive T-lymphocytes have an extremely long life span and may persist years after removal of the tumor. Thus, myasthenia gravis can develop despite removal of all thymic tissue. It is universally agreed that thymectomy is not a trigger for myasthenia gravis onset. There are many instances of myasthenia relapse post thymectomy independent of tumor recurrence. Myasthenia gravis relapse rates in patients post thymoma removal have been as high as 45 %, while the relapse rate is estimated at 14% in non-thymomatous patients.

References

Title: Got the ‘GIST’ of It: Gastrointestinal Stromal Tumor Presenting with Acute Gi bleed and Syncope

Authors: Neelesh Rastogi MD and Sarah MacArthur MD

Introduction: Gastrointestinal stromal tumor (GIST) is a common nonepithelial benign neoplasm involving the GI tract, most commonly located in the stomach. Here, we describe a challenging case of GIST that presented with acute GI bleed and syncope.

Case Presentation: A 53 year-old male with no significant past medical history presented to the ER with loss of consciousness for several minutes earlier in the day. He arrived in New York three days prior to admission after a long day of travel from Iowa during which he began to experience general malaise, chills, and had poor intake per os. On the day of presentation, the patient awoke feeling weak. He got out of bed and fell down with loss of consciousness for seconds to minutes, witnessed by his wife. Other review of systems was negative.

Physical exam revealed normal vital signs and a well-appearing gentleman without obvious abnormalities. Initial labs were notable for a hemoglobin of 9.1, BUN/Cr ratio of 29/0.7, and a respiratory viral panel positive for influenza A. The syncope was initially attributed to hypovolemia from his influenza and the team planned to discharge the patient from the ER. He then had a bowel movement with a moderate amount of bright red blood. Vital signs and Hgb remained stable. The patient was then admitted to medicine for further work-up.

The patient continued to have bright red blood followed by melenic stools in the morning and was taken urgently to the endoscopy suite. EGD revealed a large ulcerated mass oozing blood in the gastric body. Biopsies were non-diagnostic. Endoscopic ultrasound was subsequently performed showing a mass arising from the muscularis propria protruding 4-5 cm into the mid gastric body. The patient underwent a partial gastrectomy and resection of his mass. Pathology confirmed the diagnosis of gastrointestinal stromal tumor, mixed-type. The patient improved and was discharged on Hospital Day 11.

Discussion: While gastrointestinal stromal tumors tend to be benign, their presentations can be quite variable. In general, GISTs are characterized by non-specific symptoms like early satiety or bloating. In this case, our patient presented with an acute GI bleed and syncope from a GIST that had grown to be large and ulcerated. The patient’s stable vital signs and his viral illness paved the way for anchoring bias, diverting the team from further investigating his anemia, and its potential role in his presentation.
Title: Atypical McCune-Albright Syndrome Presenting as Prolactinoma and Café-au-lait Spots

Authors: Jeffrey Rico II, MD, Olayinka Wilhelm, MD, FACE, and Thomas Genese, MD, FACP, United Health Services Hospitals, Johnson City NY

Introduction: McCune-Albright Syndrome (MAS) is a rare, sporadically occurring syndrome caused by somatic mutations of the gene encoding the α-subunit of the trimeric G protein which stimulates adenylyl cyclase. It is classically characterized by the triad of café-au-lait spots, polyostotic fibrous dysplasia, and GnRH independent sexual precocity. MAS is widely diagnosed by the presence of two of the three classic manifestations; despite this, MAS has been frequently reported to present incompletely. The varied clinical findings can be partly attributed to the mosaic pattern of the somatic constitutive activating mutation implicated in MAS. Autonomous endocrine hyperfunction such as prolactinoma is a known feature of the syndrome.

Case Presentation: A 20-year-old male was diagnosed with the syndrome after he was found to have a macroprolactinoma and café-au-lait spots on the right cheek. At diagnosis, he was experiencing transient vision loss of his right eye for 1 year, progressive weight gain despite lifestyle modifications for 4 years, and gynecomastia. There was no history of precocious puberty, no lytic lesions on prior radiographs, no documentation of abnormal skin lesions in his medical record, and no evidence of bone lesions on whole body bone scan. MRI of the brain showed a 1.7 x 1.2 x 1.1 cm pituitary macroadenoma with suprasellar extension. Comprehensive endocrinological work up confirmed prolactinoma with Results: showing prolactin level of 449.3 ng/dL (normal < 20 ng/dL), LH level of 0.39 mIU/mL (normal = 1.8-8.6 mIU/mL), total testosterone level of 58 ng/dL (normal = 348-1197 ng/dL), but normal TSH, free T4, FSH, and IGF-1 levels. He was started on cabergoline 0.5mg twice a week but a repeat MRI of the brain done 10 months later showed the tumor did not change in size. He subsequently underwent transsphenoidal hypophysectomy to remove the pituitary tumor 3 weeks later.

Discussion: Prolactinoma, such as in this patient, is a well-documented manifestation of MAS, but does not fit the traditional criteria for diagnosis. Thus, we suggest that in the presence of café-au-lait spots, a diagnosis of MAS can be supported by the presence of endocrine hyperfunction. A comprehensive endocrinological work up to detect endocrinopathies should be considered in patients with only one of the three classic findings of MAS. Establishing the diagnosis definitively is important as surveillance for long term complications of McCune-Albright Syndrome should be done.
New York-Clinical Vignette-Poster Finalist
Gregory Rubinfeld, MD

Title: A Rare Interstitial Lung Disease Presenting As Neutropenic Fever

Authors: Gregory Rubinfeld MD, David Rhee MD, Gabriel Perreault MD, and Mark Sloane MD, Department of Medicine, NYU Langone Medical Center, New York, NY

Introduction: Rituximab, an anti-CD20 antibody, is frequently used in the treatment of B cell lymphomas. Rituximab induced interstitial lung disease (R-ILD) is a rare but deadly complication of rituximab therapy that may often go unrecognized.

Case Presentation: A 66 year-old man presented with fever, dyspnea, and cough productive of white sputum for three days. Three months prior to his presentation he was diagnosed with diffuse large B cell lymphoma and started on R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). His aforementioned symptoms began four days after his fourth cycle of R-CHOP.

On admission, his temperature was 102.4°F and lung fields were clear to auscultation. His initial absolute neutrophil count (ANC) was zero, blood, urine, and sputum cultures were negative, and chest x-ray showed no acute abnormalities. Vancomycin, piperillin/tazobactam, micafungin, and filgrastim were initiated for neutropenic fever. A few days after admission his ANC increased, and chest x-ray and CT showed diffuse bilateral infiltrates. He was intubated for hypoxemic respiratory failure and had a bronchoalveolar lavage that was negative for fungi, bacteria and mycobacteria. Methylprednisolone (125 mg IV every 6 hours) was initiated to address a non-infectious pneumonitis. Thoracoscopic lung biopsy revealed diffuse interstitial pneumonitis without evidence of infection, suggesting a diagnosis of R-ILD.

Within two days of initiating steroids the patient’s pulmonary infiltrates significantly improved and he was extubated. He continued to improve on a steroid taper and upon discharge it was recommended that rituximab be removed from his future chemotherapy regimen.

Discussion: Initially, the patient’s neutropenic fever and non-specific symptoms were concerning for an underlying infection. However, his clinical deterioration with the recovery of his ANC and despite broad-spectrum antibiotics led us to consider less common diagnoses with overlapping presentations. R-ILD presents with fever, dyspnea, and cough beginning days to weeks after successive rituximab infusions (typically after the fourth cycle), as was seen in our patient. Diffuse bilateral infiltrates on imaging and interstitial pneumonitis on biopsy are characteristic of R-ILD as well. While the pathogenesis of R-ILD remains poorly understood, cell lysis and cytokine release provoked by rituximab therapy are thought to play a key role. This may offer insight into why our patient decompensated as his ANC recovered. Corticosteroids are the treatment of choice for R-ILD and often lead to prompt clinical improvement as seen in our patient, however numerous fatalities from R-ILD have been described.

With the growing utility of rituximab for hematologic and rheumatologic disorders the incidence of R-ILD has risen in tandem. Given the nonspecific presentation of R-ILD, a high index of suspicion is fundamental to the recognition and treatment of this rare but potentially fatal entity.

References


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New York-Clinical Vignette-Poster Finalist
Sylvana Salama

Title: How The Times Have Changed: Pulmonary Kaposi’s Sarcoma in the Post-HAART Era

Authors: Salama, Sylvana M.D., Ravish Singhal M.D., Keerthana Keshava M.D., New York Presbyterian Brooklyn Methodist Hospital

Introduction: Pulmonary Kaposi’s Sarcoma is a rare entity, and poor prognostic sign affecting immunocompromised patients. With the advent of HAART, the incidence of Kaposi’s Sarcoma has decreased significantly.

Case Presentation: The patient is a 27 year old male with recent diagnosis of AIDS (last known CD4 count of 64) who presented with inability to turn his neck, dry cough, fatigue, and shortness of breath for two weeks. Two months prior to presentation, upon diagnosis of HIV, he was started on Elvitegravir/Cobicistat/Emtricitabine/Tenofovir (Genvoya), Acyclovir, and Trimethoprim/Sulfamethoxazole. He denied any fevers, chills, night sweats, recent travel, or exposure to sick contacts. He is a homosexual male who reported having multiple sexual partners over the past 1 year. On physical exam, he was noted to have decreased breath sounds at the bilateral bases, rhonchi in mid-lung fields, violaceous skin lesions on the lower lip and anterior chest, and marked posterior cervical and axillary lymphadenopathy. Infectious workup as well as flow cytometry were non-contributory. Chest x-ray revealed bilateral lung parenchymal infiltrates and pleural effusions. Given his presentation and radiographic findings, he underwent bronchoscopy. This revealed diffuse, erythematous airway inflammation extending from the lower part of the trachea to the distal airways and diffuse granular non-obstructing lesions throughout the airways bilaterally. He then underwent biopsy of the skin lesions and excisional cervical lymph node biopsy. Bronchoalveolar lavage and skin biopsies confirmed the diagnosis of Kaposi’s Sarcoma, for which he was started on Liposomal Doxorubicin.

Discussion: Kaposi’s sarcoma (KS) is a vascular tumor affecting the blood vessels and lymph nodes associated with Human Herpesvirus 8. It is an uncommon diagnosis but the risk is 10,000 times greater among those with a CD4 count less than 150, and especially common in young to middle aged males. Up to 40% of homosexual males presented with KS at the time of initial AIDS diagnosis in the 1980s. Ten years later, this number had decreased to approximately 10-20%. More recently, with the adoption of HAART, this has further decreased by 59% with triple therapy. The incidence of AIDS-related KS is now down to approximately 2,500 cases each year (as of 2015). Patients with pulmonary KS have median survival of 3-10 months, but with chemotherapy and HAART, survival improves to 18 months. Although Kaposi’s sarcoma is much less common today, it is still an important diagnosis to be mindful of when evaluating an immunocompromised patient with mucocutaneous lesions.

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New York-Clinical Vignette-Poster Finalist
Neha Sharma

Title: The Case of Pembrolizumab and A Drowning Heart

Authors: Neha Sharma, DO (Department of Internal Medicine, Baystate - University of Massachusetts Medical School), Myat Soe, MD (Department of Internal Medicine, Baystate - University of Massachusetts Medical School)

Introduction: Pembrolizumab, included in a class of drugs known as immune checkpoint inhibitors, is a monoclonal antibody that inhibits the programmed cell death protein 1 (PD-1) of cytotoxic T lymphocytes. Immune checkpoint inhibitors have shown antitumor responses in multiple cancers including melanoma, non-small cell lung cancer (NSCLC), and renal cell cancer. With increasing use of checkpoint inhibitors, a unique spectrum of side effects called immune-related adverse events (irAEs), including thyroiditis, hypophysitis, rash, hepatitis, pneumonitis and type 1 diabetes have been reported. Pericardial effusion as an irAE of pembrolizumab has not been reported in the literature before diagnosis of this case. We report a case of NSCLC with hemorrhagic pericardial effusion secondary to pembrolizumab.

Case Presentation: A 66-year-old female was diagnosed with squamous cell carcinoma of the left lung with mediastinal lymphadenopathy and left adrenal metastasis, and underwent the first round of immunotherapy with pembrolizumab. Two weeks later, patient was admitted for progressive shortness of breath and chest pain with significant hypoxia. Physical exam was remarkable for reduced breath sounds at the left lung base and muffled heart sounds. Computed Tomography of the chest showed left lung mass with concern for post-obstructive pneumonia and large pericardial effusion without signs of tamponade on echocardiogram. Antibiotics were started for possible post-obstructive pneumonia. Pericardiocentesis was performed with drain placement. Fluid was hemorrhagic and consistent with lymphocyte predominant exudate. To increase diagnostic yield, three separate pericardial fluid samples were sent for cytology and no malignant cells were seen. Cultures and connective tissue disorder workup, including anti-neutrophil cytoplasmic antibodies, antinuclear antibody, and rheumatoid factor were negative. Patient continued to have significant output from pericardial drain for a few days. Based on the workup and clinical scenario, irAE from pembrolizumab was suspected and patient was started on high dose prednisone. After starting a steroid, there was a marked and sustained decrease in pericardial drain output and shortness of breath. The drain was then removed. Prednisone was continued for a few weeks with a taper and immunotherapy was held.

Discussion: Pembrolizumab has been associated with fatal myocardial infarction and myocarditis in some case reports. There was no case of pericardial effusion related to pembrolizumab reported in the literature at the time of diagnosis, although massive pericardial effusion has been reported with nivolumab. Our case highlights the importance of recognizing pericardial effusion as an irAE of pembrolizumab for prompt and appropriate treatment with steroids. Clinicians should also keep in mind that pneumonitis, pleural effusion, and pericardial effusion in lung cancer patients treated with immune checkpoint inhibitors could be pseudoprogression of tumor from irAEs rather than actual progression of tumor. Thorough workup should be pursued for diagnosis as treatment of moderate to severe irAEs requires interruption of the checkpoint inhibitor and the use of corticosteroid immunosuppression.
New York-Clinical Vignette-Poster Finalist
Yael Simons, MD

Title: Influenza A Induced Pancytopenia

Authors: Yael Simons MD, Neena Malik MD, Jenny Placido Disla MD, Wan Ling Lam MD, Icahn School of Medicine at Mount Sinai Beth Israel

Introduction: The following case highlights a rare complication, pancytopenia, in a common disease entity, Influenza A.

Case Presentation: A 73-year-old Mandarin speaking male with a past medical history of diabetes presented to the emergency department following an outpatient visit which found abnormal laboratory values. On initial assessment, the patient had stable vital signs and physical exam was solely notable for clear nasal discharge. Laboratory evaluation displayed leukopenia of 1,400 K/uL with lymphocytic predominance, normocytic anemia of 8.5 g/dL, and thrombocytopenia of 15,000 K/uL. On further history, the patient had been in his usual state of health until two days prior to presentation when he developed rhinorrhea, sore throat, dry cough, and decreased appetite. In the ED, nasopharyngeal swab was obtained that confirmed Influenza A virus. The patient was initiated on oseltamivir and supportive treatment with improvement of viral symptoms. Multiple laboratory and imaging tests were obtained to evaluate for alternate infectious, malignant, or immunologic etiologies that could be causing the pancytopenia, of which all Results: were normal. The hematologic abnormalities gradually improved as the viral symptoms resolved, suggesting that the pancytopenia was secondary to Influenza A.

Discussion: Influenza is a major cause of morbidity and mortality worldwide, with the CDC estimating an approximate of 425,000 influenza-related hospitalizations annually since 2010. Individual hematologic abnormalities are often noted in patients with influenza; however, pancytopenia has rarely been documented in the literature, with majority of cases seen in the pediatric population. The mechanism of pancytopenia in influenza may involve viral inhibition of erythroid and granulocytic cell lineages via infection of the bone marrow, however, additional research is needed to verify this finding. Regardless of etiology, pancytopenia in influenza is a serious complication of a common condition that must be recognized, with the understanding that laboratory values will recover with improvement of the viral syndrome.

References

New York-Clinical Vignette-Poster Finalist
Bushra Tbakhi, MD

Title: Can Multiple Myeloma Lyse before treatment?

Authors: Bushra Tbakhi, Sandrine Hanna, Yazan Samhouri, Deerajnath Lingutla, Department of Internal Medicine, Rochester Regional Health- Unity Hospital, Rochester, NY

Introduction: Tumor Lysis syndrome (TLS) is an oncologic emergency characterized by the lysis of tumor cells and subsequent rapid release of intracellular contents into the bloodstream. It is typically associated with highly proliferative tumors like high-grade lymphomas and leukemias, usually after initiation of treatment. Multiple myeloma (MM) is a malignancy classified as low risk to progress to TLS given the plasma cells' low proliferative rate. Patients that progress to TLS generally do so after the commencement of chemotherapy. Spontaneous TLS prior to the initiation of chemotherapy is an exceedingly rare occurrence in MM. We report an interesting case of spontaneous TLS present before the diagnosis of his MM.

Case Presentation: A 52-year-old male with history of bipolar disorder presented to our hospital with a three-day history of lethargy and abdominal pain. He denied any back pain. On physical examination, he appeared to be dehydrated along with mild generalized abdominal tenderness; otherwise unremarkable exam. Initial labs revealed a calcium level of 17.4 mg/dL, creatinine of 4.5 mg/dL, uric acid of 19.2 mg/dL, Phosphorus 6.3 mg/dl, LDH of 559 U/L, total serum protein 8.8 g/dL, mild normocytic and normochromic anemia with Hb of 11.6 g/dL, and a lipase of 1198 U/L. He was treated for spontaneous TLS, hypercalcemia, and hypercalcemia induced acute pancreatitis with aggressive IV fluid hydration, Rasburicase, Pamidronate and Calcitonin. Uric acid improved to 9.9 mg/dL and Calcium to 14.7 mg/dL. Further workup included serum protein electrophoresis showing an M-spike (2.1 g/dL paraproteins) in the Beta region, serum immunofixation revealing IgA k Myeloma, with a κ/λ ratio of 3.37. Plasma cells comprised 55% of the bone marrow biopsy. No lytic lesions were observed on a skeletal survey. The patient was further treated with plasmapheresis and initiation of hemodialysis due to worsening of acute renal failure secondary to myeloma kidney. Subsequently, he was started on Dexamethasone and Bortezomib. His most recent labs showed normalization of calcium and gradual improvement of his creatinine.

Discussion: As per the Cairo-Bishop definition, this MM case meets both the laboratory and clinical criteria for TLS. The timeline described indicates that TLS was present prior to MM diagnosis and treatment with Bortezomib. While treatment induced TLS in MM is rare, there are only five reported cases of TLS occurring prior to MM treatment. Constituting the sixth case worldwide, this case adds to the sparse literature available regarding spontaneous TLS in MM. High tumor burden, unfavorable cytogenetics, and elevated LDH (as displayed in this case) are risk factors associated with progression to TLS. This case highlights the importance of considering this rare yet life-threatening possibility in the initial workup of MM. A higher index of suspicion amongst clinicians will aid in prompt recognition and intervention, facilitating improvement of patient outcomes.

References
New York-Clinical Vignette-Poster Finalist
Syed Arsalan Zaidi, MBBS MD

Title: Cerebrofacial Arteriovenous Metameric Syndrome (CAMS) Type II with unusual involvement of temporal lobe

Authors: Syed Zaidi M.D.(a), Tushar Bajaj (b), Elkin Rene M.D.(a) (c), (a)Department of Medicine, Bronx Lebanon Hospital Center, (b) American University of the Caribbean School of Medicine (c) Chief of Division of Neurology, Bronx Lebanon Hospital Center, Bronx, NY, 10457. USA

Introduction: Cerebrofacial Arteriovenous Metameric Syndrome (CAMS) is a term that encompasses craniofacial arterio-venous malformation (AVM) syndromes into a classification system depending on location of AVMs. CAMS is classified into 3 subgroups:

- CAMS I: medial prosencephalic group with involvement of nose and hypothalamus
- CAMS II: lateral prosencephalic group with involvement of occipital lobe, optic chiasma, optic tract, thalamus, retina and maxilla
- CAMS III: rhombencephalic group with involvement of cerebellum, pons and mandible

Case Presentation: We discuss a case of a 24-year-old male patient who presented to the hospital with twitching of left side of face and left arm for two weeks and worsening for past four days. He had significant comorbidities of Wyburn-Mason Syndrome, right cerebral AVM status-post coiling, history of stroke 10 years ago; with residual left sided weakness (patient is hemiplegic and wheel chair bound), hypertension, hypothyroidism, and seizure disorder. Patient was born with hydrocephalus, and has been having seizures since he was 5 years old. His physical exam was remarkable for left arm tremors, left facial twitching in the area of the zygomaticus, right sided ptosis, dense hemiplegia of left side and slightly decreased sensation on left side of body as compared to right. Computerized tomographic scan of head showed Large AVM involving the right cerebral hemisphere. Contrast enhanced magnetic resonance imaging of the head showed marked signal abnormality involving the right cerebral hemisphere with extensive pathologic flow voids compatible with extensive vascular malformation with extension to the right orbit. Flow-Voids involving most of right temporal lobe, optic chiasma, optic tract, thalamus, retina, and slight extension to right frontal lobe. Also seen was mildly increased diffusion weighted signal involving central medulla concerning for tiny acute/subacute lacunar type infarct. Carotid Ultrasound showed no significant hemodynamic stenosis of carotid and vertebral arteries. No surgical intervention was recommended due to chronic nature of most findings. Patient was started on levitirecimat and valproic acid for his seizures, and was discharged home. Patient was followed as outpatient with clinical improvement and good response to anti-seizure medications.

Discussion: Our case has a rare variant of Type II CAMS, which involves the temporal lobe as well, it is an unusual finding and has potential for research to investigate possible genetic linkage between lateral prosencephalic group of CAMS and other arterio-venous malformations.
Title: Acute Cytomegalovirus Mononucleosis as a Cause of Generalized Lymphadenopathy in an Immunocompetent Female

Authors: Syed Arsalan Zaidi M.D. (a), Tushar Bajaj (b), Efrain Gonzalez, M.D. (a,c), Anupama, Menon M.D., M.P.H (a,c), (a) Department of Medicine, Bronx Lebanon Hospital Center, Bronx, NY, USA, (b) American University of the Caribbean School of Medicine, (c) Division of Infectious Diseases, Bronx Lebanon Hospital Center, Bronx, NY, 10457. USA.

Introduction: Cytomegalovirus infection is a well-documented infection in immunocompromised hosts; however, there are limited reports in immunocompetent individuals because patients are usually asymptomatic or the infection is discovered on routine testing incidentally. Mononucleosis has a myriad of clinical manifestations such as low-grade fever, pharyngitis, fatigue, generalized lymphadenopathy and splenomegaly.

Case Presentation: We discuss a case of a 48-year-old immunocompetent female who presented with a one-week history of parotid swelling, sore throat, with associated low-grade fever. She had a medical history significant for well-controlled type 2 diabetes mellitus, hypertension, hyperlipidemia, hepatic steatosis, parathyroidectomy, and history of non-obstructing renal calculus. Physical exam revealed diffuse bilateral preauricular, postauricular, and submandibular swelling. Also noticeable was mild bilateral parotid gland swelling, supraclavicular, axillary and inguinal lymph node enlargement. Rest of the examination was unremarkable. Pertinent laboratory values included a white blood cell count of 8.2 k/ul, with 64% neutrophils, 14.5% lymphocytes, 13% monocytes, and 8% eosinophils; C-reactive protein of 69.5mg/L. The initial differential diagnosis included acute parotitis, reactive lymphadenopathy, acute retroviral conversion syndrome, and EBV mononucleosis. The patient was initially treated with ketorolac for pain control, given intravenous fluid bolus, and begun empirically on clindamycin and dexamethasone.

Further evaluation with contrast-enhanced computed tomography revealed generalized lymphadenopathy in the neck, submandibular region, axilla, mediastinum, perihiilar, periaortic, obturator, mesenteric, portocaval, periceliac, inguinal, and external iliac regions. Fine needle aspiration of a left supraclavicular lymph node revealed numerous mature appearing lymphocytes and no evidence of malignancy or other diagnosis. Immunological workup revealed a negative heterophile agglutination assay; negative Epstein-Barr virus early antigen; immunity to mumps; negative antibody to HIV-1 and HIV-2; negative antibody to HTLV-I and -II; negative RPR assay for syphilis; negative Strongyloides IgG antibody, undetectable HIV-1 viral load; negative Rheumatoid factor; and negative cryoglobulin assay. A titer of >4.0 of Cytomegalovirus-specific IgM antibody was detected via enzyme-linked immunosorbent assay (ELISA). A level for CMV IgG obtained 7 days after initial presentation revealed a positive result, level of 1.88. In view of pertinent history, clinical features, laboratory Results: and computed tomography findings, the patient was diagnosed with Cytomegalovirus mononucleosis. Antibiotics were discontinued after preliminary bacterial blood cultures revealed no growth; only symptomatic management was continued. The patient’s lymphadenopathy dramatically improved within the one-week inpatient hospital stay. One month after hospitalization the patient was seen in clinic, where she reported that her symptoms had completely resolved and denied any recurrence of lymph-node enlargement or tenderness.
Discussion: Although Epstein-Barr virus is the most common cause of mononucleosis, and is confirmed in over 90% of cases of the syndrome in patients who present for evaluation; however, Cytomegalovirus has been documented in approximately 5 to 7% of evaluated patients and should be considered as a rare but important cause of generalized lymphadenopathy in immunocompetent adults.
New York-Clinical Vignette-Poster Finalist

Wen Zhang, DO

Title: A rare presentation of an ancient disease - a case of Rapidly Progressive Syphilitic Lumbosacral Polyradiculitis

Authors: Wen Zhang, DO, Mount Sinai St Luke's and Mount Sinai West Hospitals

Bertin Salguero, MD, Mount Sinai St Luke's and Mount Sinai West Hospitals

Introduction: Neurosyphilis is known as a late stage complication of syphilis, often presenting as part of secondary/tertiary syphilis with a wide range of neurologic manifestations. The neurologic clinical syndromes vary with disease stage, with meningitis, cranial nerve palsies and ocular syphilis arising within one year of the initial infection; whereas, meningovascular and parenchymal syphilis occurs in later stages. In this case, we present a patient with well controlled HIV that developed rapidly progressive lumbosacral polyradiculitis in the absence of previous syphilis infection.

Case Presentation: 58 year old male with well controlled HIV [current CD4 699, 33%, VL undetectable]. Three weeks prior to presentation, he reported a fall after which he hurt both knees and began noticing some mild weakness in both legs. His primary doctor ordered a MRI, which was negative for injuries. Over the following weeks, he developed worsening weakness in his lower extremities (right > left) to the point of needing crutches for ambulation, associated now with low back pain and lower extremity paresthesias. On admission, he was noted to have noticeable proximal lower extremity weakness and areflexia, however was otherwise asymptomatic.

Head CT was unremarkable and a lumbar MRI showed leptomeningeal enhancement in the distal lumbar region. An extensive workup including TSH, creatinine kinase, aldolase, antinuclear antibody, returned negative, and electromyography was planned. However, RPR returned positive, with a titer of 1:128, confirmed with a positive FTA-ABS test. Due to suspicion of neurosyphilis, a lumbar puncture was done that showed a WBC count of 140, protein of 264, and a positive VDRL with a titer of 1:8.

The patient had well controlled HIV, had had negative RPRs on multiple tests every year since 2010, the last one being just four months prior, demonstrating that his current presentation was a newly acquired infection. The patient stated he had only one partner 6 months ago, where no anal penetration took place, but did engage in oral sex. He was treated with IV Penicillin, his weakness slowly improved during hospitalization and he was discharged to rehabilitation.

Discussion: While it is known that syphilis usually presents in stages that progress in a stepwise manner, our case represents an atypical presentation, where the patient’s primary symptom was rapid new onset weakness. Late syphilis and neurosyphilis is uncommon in individuals with adequate CD4 counts, and cases of symptomatic neurosyphilis are especially rare, taking years to develop even in immunocompromised individuals. Therefore upon arrival, syphilis was not on our differential given recent negative RPR Results: and well controlled HIV. However, our case remind us that syphilis always needs to be considered as a possible etiology for neurologic symptoms, and that in certain cases, can develop rapidly.
No Chapter - Clinical Vignette-Poster Finalist
Muneer Al-Husseini

Title: Cough and air under the diaphragm; Chilaiditi syndrome as a rare cause of pneumoperitoneum

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Introduction: Pneumoperitoneum poses an important diagnostic sign determining the urgency of management of patients in the Emergency Department (ED). Chilaiditi syndrome is a rare condition of symptomatic large intestines transposition between the diaphragm and the liver. We present a rare case of a 49-year-old male who presented to the emergency department complaining of cough and vague abdominal discomfort who was found to have Chilaiditi syndrome diagnosed radiologically by CT scan.

Case Presentation: A 49-year-old male presented to the ED with a 48-hour history of cough. The cough was productive of a small amount of sputum and caused abdominal discomfort. He denied a previous similar episode. He was fatigued but recalled no chest pain, emesis, fever, chills, night sweats, melena, constipation or diarrhea. His past history was only significant for laparoscopic Roux-en-Y gastric bypass. He denied tobacco, alcohol, or illicit drug use. His family history was not significant. On ED presentation, he was afebrile, with a blood pressure of 152/74 mmHg, pulse of 98 beats/min, respiratory rate of 18 beats/min, and oxygen saturation of 98% on room air. On physical examination, the cardiovascular and respiratory examinations were unremarkable. His abdomen was soft, nontender, nondistended, no organomegaly, stool guaiac negative, and inspection showed scars from a previous laparoscopic surgery. Troponin-I and electrolyte levels were normal. Complete blood count was unremarkable. Kidney function tests were within normal limits except for a low Urea (1.52 mmol/L). Imaging studies showed air under the diaphragm in the chest X-ray. Further imaging by a CT abdomen with contrast was obtained and showed that the supposed air underneath the raised right copula of the diaphragm was a loop of colon. These findings were suggestive of Chilaiditi syndrome. The patient was conservatively managed with IV fluids, cough suppressants, and pain management. The pain resolved and the patient was stable. After discussing the diagnosis and the imaging with the patient he chose to be discharged home.

Discussion: This case highlights the importance of treating the patient as a human rather than numbers and images. Medical students are taught that air under the diaphragm is always a surgical emergency. It almost always is, but a thorough physical examination that does not show signs of peritonitis should prompt further investigations to understand the underlying pathology. Chilaiditi’s sign is a radiographic evidence of air filled loops of large intestines between the right copula of the diaphragm and the liver. When the patient becomes symptomatic and complains of abdominal pain, constipation, vomiting, cough, dyspnea, anorexia, and/or volvulus; it is then referred to as Chilaiditi’s syndrome. If colonic loops are mistaken for air under the diaphragm on imaging, that might lead to undergoing unnecessary serious procedures increasing the patient morbidity.
Title: Less myeloid cells than expected.

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INSTITUTION: CEDIMAT, Santo Domingo, Dominican Republic.

Introduction: Multiple myeloma requires very specific findings for its diagnosis, some of them are anticipated by the physician while waiting for tests but, if resulted otherwise, our approach to the patient must not necessarily change, as seen in this case.

Case Presentation: A 51-year-old man consulted his doctor with a history of one-month mild chest pain in the right hemithorax when leaning forward. CT scan of the chest reported a mass between his right pleural cavity and ribs. With a CT-guided biopsy the diagnosis of Solitary Extramedullary Plasmocytoma (SEP) was made; he underwent surgery without further investigation. Four months later, he came to the emergency room with a 6-week history of headache worsened in the last 5 days, dysphagia to solids and dysarthria aggravated when standing or with head turns. Physical examination was unremarkable. After extensive evaluation by the neurology department, head-CT depicted osteolytic bone lesions at clivus, occipital condyle, atlas and axis; Dexamethasone was started. Given the imminent risk of pathological fracture, radiotherapy was begun at the third day of admission. Several diagnostic tests were obtained: hemoglobin 10.8 g/dL, hematocrit 31.9%, serum calcium 8.6 mg/dL, creatinine 1.02 mg/dL, BUN 6.07 mg/dL, IgG 4,314 mg/dL, total proteins 9.1g/dL, globulins 6,000g/dL, Beta 2 microglobulin 2,753 mg/L. Serum protein electrophoresis revealed a single narrow peak of monoclonal gamma protein of 46% (3.7g/dL) and the bone marrow biopsy identified approximately 5% of myeloid cells, concluded as non-diagnostic. Chemotherapy was started by day six with thalidomide, bortezomib and zoledronic acid. Further testing showed osteolytic bone lesions at multiple levels of the spine. Due to persistence of dysphagia despite treatment, a gastrostomy tube was placed. Six months later, the patient continued receiving therapy with significant dysarthria improvement.

Discussion: Everybody recognizes multiple myeloma at the finding of more than 10% of myeloid cells in the bone marrow, but it is not so easy to discard or to confirm when having a patient with this clinical presentation. A biopsy-proven plasmacytoma is sufficient to make this diagnosis coupled with evidence of end organ damage (such as CRAB)\(^1\); some will even skip bone marrow biopsy since the presence of the long-awaited 10% of myelocytes is not essential on this case. The medullar compression caused by the osteolytic bone lesions lead to an inaccurate initial approach by the neurology department on a patient with a previous diagnosis of SEP. These symptoms should rise a suspicion of progression of the disease or allow us to disregard the first diagnosis. The relevance of this case is highlighted by the finding of a non-diagnostic bone marrow biopsy and a spinal cord compression related to bone lesions.

References

No Chapter-Clinical Vignette-Poster Finalist
Pamela Pina Santana, MD

Title: A not so simple back pain.

Authors: Pamela Piña S., PGY-1 Internal Medicine, CEDIMAT., William Castro, Neurologist, CEDIMAT., INSTITUTION: CEDIMAT, Santo Domingo, Dominican Republic.

Introduction: Infective endocarditis is a disease of the endocardial surface of the heart, primarily affecting heart valves or intracardiac devices, which has multiple systemic repercussions. It should be considered in the context of a patient with cardiac findings, fever and multiorgan symptoms.

Case Presentation: A 75-year-old male patient was admitted to the hospital after being evaluated in the emergency room for a 7 days lower back pain, 9 of 10 on pain scale, non-radiating but limiting his ambulation. Neurological exam unraveled sensitive aphasia with no other focal neurological deficits. A CT scan of the brain reported a left parieto-occipital hypo-density with no edema or midline deviation, also seen on MRI. His past medical history was unremarkable. Cardiac examination revealed a holosystolic murmur III/VI heard best at the apex radiating to the axilla. An echocardiogram exhibited severe acute mitral regurgitation related to a 13mm mitral valve vegetation with posterior leaflet rupture. No skin or eye findings were noted. CBC showed mild leukocytosis.

A Lumbar MRI was obtained due to the persistence of back pain, where a diffuse hyperintensity at L2-L5 was seen, defined as a probable infectious diskitis. On further investigation, non-medicated fever, predominantly nocturnal during the last 5 days, was elicited. He was taken to surgery by the third day of his admission for mitral valve replacement with a biological prothesis. Surgeons described a prolapsed, thick, spongy and friable valve suggestive of Barlow's disease and vegetations extending throughout the valvular and subvalvular apparatus. Blood cultures reported streptococcal (undefined specie) growth. The patient had a favorable postoperative recovery without new neurological deficits and completed 4 weeks of antibiotics.

Discussion: We took the chance of naming as embolic stroke the findings at head CT scan and MRI, and as vertebral osteomyelitis from metastatic infection those on Lumbar MRI, and conducted the treatment in concordance.

Infective endocarditis produces embolic stroke (40%), septic embolism (25%) with infarction of kidneys, lungs, and spleen; and metastatic infection such as vertebral osteomyelitis, septic arthritis and psoas abscesses. The diagnosis should be suspected in patients with cardiac signs on physical exam and a history of fever although not well characterized in our first encounter. Septic embolism produces severe multiorgan damage and to ensure patient's recovery, we need to achieve a proper diagnosis and start treatment immediately. Vegetations greater than 10mm in size and posterior leaflet rupture producing acute mitral regurgitation are clear indications of surgical intervention. Uncommon manifestations of a common disorder made this case worth discussing.

References


Title: Familiarity Bias Delaying the Rare Diagnosis of Prostatic Neuroendocrine Carcinoma

Authors: Ashley Choe, MD - Internal Medicine-Pediatrics Residency Program, Vidant Medical Center/East Carolina University Brody School of Medicine, Jonathan Garber, DO - Internal Medicine-Pediatrics Residency Program, Vidant Medical Center/East Carolina University Brody School of Medicine, Arul Vigg, MD - Internal Medicine, Vidant Medical Center/East Carolina University Brody School of Medicine

Introduction: Neuroendocrine carcinomas of the prostate, which comprise a small subset of prostate malignancies, are a rare and varied group of tumors with differing management and outcomes. We present a case of delayed diagnosis of prostatic neuroendocrine carcinoma in a middle-aged male with rapidly progressive lower urinary tract symptoms initially attributed to benign prostatic hyperplasia (BPH) and prostatitis.

Case Presentation: A 55-year-old male without prior urological history presented with dysuria and was empirically treated for a urinary tract infection; however, the next day he went to the emergency department for worsening urinary retention. CT scan showed a markedly enlarged (10x11x12 cm), heterogeneous appearing prostate and several small, sclerotic pelvic bone lesions noted to be possible metastases. With a normal PSA at 1.68 ng/mL, the patient was diagnosed with BPH and prostatitis and was started on tamsulosin and antibiotics again. Symptom progression prompted referral to urology for further management of BPH and insertion of an indwelling catheter for worsening retention. During outpatient urological workup, development of severe constipation and sepsis presumed to be secondary to prostatitis prompted hospital admission and eventual transfer to a tertiary care center for further management. Repeat CT prior to transfer revealed further pelvic mass enlargement, and a second PSA check was normal. Given his constipation and a large palpable rectal mass on physical exam, flexible sigmoidoscopy was performed for evaluation of suspected colorectal malignancy. However, this did not reveal an intra-rectal mass. An MRI of the pelvis for further mass characterization showed an enlarged prostate (13.4x11 x17.5 cm) with no significant internal enhancement, a fluid density suggestive of prostatic abscess, and re-demonstration of pelvic bone lesions suspicious for metastases. Subsequent biopsy revealed a highly aggressive stage IV prostatic neuroendocrine carcinoma six weeks after the original label of BPH with significant growth of the tumor during that time.

Discussion: Familiar symptoms lead physicians down comfortable diagnostic pathways: most men over the age of 50 presenting with hesitancy, straining, dribbling, and dysuria have BPH. However, clinicians must guard against cognitive biases (familiarity, availability, and anchoring biases), which can impede consideration of other diagnostic possibilities. The distinct features of this unusual malignancy made diagnosing this patient difficult. He represents the kind of familiar presentation that we must guard against when reaching out for readily available answers and anchoring ourselves in them. Without guarding against cognitive biases, physicians with good intentions can easily be led down the wrong diagnostic pathway. We believe this case report serves as a useful reminder of the need for vigilance in this area.

References


4. Pathology image courtesy of Shuhua Ma, MD, Department of Pathology, East Carolina University.
North Carolina-Clinical Vignette-Poster Finalist
Sean M Hernandez, MD

Title: Look! It’s a mass! It’s a fungus! It’s legionella? The Atoll Sign as a Diagnostic Aide for Pneumonia in Immunosuppressed Patients

Authors: Sean Hernandez, MD, Resident, Department of Medicine, Wake Forest University, Winston-Salem NC, Caryn Brehm, MD, Resident, Department of Medicine, Wake Forest University, Winston-Salem NC, Aaaron Lampkin, DO, Fellow, Department of Pulmonary, Critical Care, Allergy and Immunologic Medicine, Wake Forest University, Winston-Salem NC, Rita Bahkru, MD, Associate Professor, Department of Pulmonary, Critical Care, Allergy and Immunologic Medicine, Wake Forest University, Winston-Salem NC

Introduction: The reverse halo sign, or the “atoll sign”, is a chest imaging finding normally suggestive of a fungal pneumonia. However, studies have shown that the atoll sign is a nonspecific finding in immunosuppressed individuals as we’ll see in the following vignette.

Case Presentation: A 48 year-old man with a history of systolic heart failure status post pacemaker placement and end stage renal disease status post donor kidney transplant (on tacrolimus, mycophenolate and prednisone) presented to the nephrology clinic with a fever. His only symptom was mild chest pain over the surgical site of his pacemaker. Physical exam was normal except for two vesicles (1 cm and 5mm respectively) noted on his right forearm which were not painful. Labs were remarkable for a leukocytosis of 22,000 with a neutrophilic predominance. Chest X-ray showed a rounded consolidation within the left upper lobe. In addition to pneumonia, there was concern for infarct or hemorrhage. CT imaging revealed central ground glass opacifications with a surrounding dense consolidative peripheral ring (atoll’s sign). This sign, given his immunosuppression, was concerning for fungal pneumonia.

Pulmonology was consulted and recommended a bronchoscopy which revealed normal anatomy without overt concern for hemorrhage, infarct or infection. With the two vesicular lesions on the right forearm and the pulmonary findings in an immunosuppressed patient, disseminated fungal infection was the top differential and dermatology was consulted. Punch biopsy done on both lesions was consistent with mixed-cell infiltrate seen in sepsis. Urine, blood, serum fungal, and sputum cultures revealed no growth. However, both urine and sputum legionella antigen were positive for legionella pneumophila.

On admission to the hospital he was started on broad spectrum antibiotics including piperacillin/tazobactam and vancomycin. He continued to have intermittent fevers and sustained leukocytosis until day 3 of admission when doxycycline was added to the antibiotic regimen. He responded well to the addition of doxycycline and was discharged with a goal of two weeks total of doxycycline therapy. Clearance of both the pneumonia confirmed with follow up imaging at a follow-up office visit with his primary nephrologist.

Discussion: This case highlights both an atypical presentation of Legionnaire’s pneumonia and the low specificity of atoll’s sign. Although atoll’s sign is common with fungal pneumonias, it is not exclusively seen with fungal infections. In immunosuppressed and immunocompromised patients, it can be a result of a bacterial infection (mostly pneumococcal pneumonia but also psittacosis or Legionnaire’s pneumonia as in our patient). The literature supports working up fungal etiology when the atoll’s sign is present. However, the atoll sign alone is not sufficient to diagnosis fungal pneumonia and treatment for a fungal infection should not be initiated based on the imaging findings alone.

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References

Title: A Case of Ominous Hiccups

Authors: Saleen Khan MD, Myra Chai MD, Sowmya Nagaraj MD

Introduction: Little is known about hiccups (singultus). They can be a transient and benign phenomenon, hardly noticeable or recognizable, but they can also be a harbinger of more serious pathology. We present a rare case of unremitting hiccups marking a potentially lethal illness.

Case Presentation: A 50-year-old man with a history of hypertension presented to the emergency department with a three-day history of nonstop hiccups. His systolic blood pressure was more than 200 and chest x-ray imaging was unremarkable. Over the next several days, his blood pressure remained elevated despite multiple medication additions, and he continued to complain only of hiccups. Workup for secondary causes of hypertension was unremarkable. An echocardiogram ordered for evaluation of cardiac abnormalities demonstrated a dissection flap in the descending thoracic and abdominal aorta and possibly into the aortic arch. Computed tomographic scan showed aortic dissection with an intramural hematoma extending from the left subclavian origin into bilateral common iliac arteries without mesenteric or renal vasculature involvement, consistent with a Stanford type B aortic dissection. On review of admission chest x-ray, widened mediastinum had been missed in the setting of severe scoliosis. The patient was initially managed medically with tight blood pressure control and complete resolution of his hiccups. Several days later, his hiccups returned and he was found with continued aneurysmal degeneration of the descending aorta in the setting of new lower extremity motor and sensation loss, for which he underwent successful thoracic endovascular aortic repair without residual deficits.

Discussion: Aortic dissection is a catastrophe that, if undiagnosed, carries a 50% mortality rate within the first 48 hours. In fact, 20% of patients will die before even reaching the hospital. Complicating matters, initial imaging studies may not show characteristics signs of a widened mediastinum on chest x-ray, and patients may present without typical symptoms of chest and upper back pain. In the previous case, the misread of the initial chest x-ray led to delayed ordering of the echocardiogram that ultimately established the diagnosis. Asymptomatic presentation of type B aortic dissection is exceedingly rare, and to date only two other cases in the literature have reported hiccups as the presenting symptom. The mechanism behind this phenomenon is unclear, however it is hypothesized that irritation of the phrenic nerve by the distention of the aorta leads to recurrent hiccups, as seen with this patient. Although the patient ultimately did not suffer residual neurological deficits on discharge, earlier diagnosis of the aortic dissection could have decreased risk for further aneurysmal degeneration. This case emphasizes the importance of maintaining high clinical suspicion of rare catastrophes despite a negative initial work up and to consider aortic disease in the setting of prolonged hiccups.

References
Title: Extreme Insulin Resistance Responsive To Immunosuppressive Therapy

Authors: Koromia GA, Bassi S, Azad MR, Houston C, Cook F

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Introduction: Type B Insulin Resistance (TBIR) Syndrome is a rare condition in which autoantibodies to the insulin cell surface receptor cause severe hyperglycemia, and occasionally, dangerous hypoglycemia. TBIR typically occurs in the setting of other autoimmune diseases. Treatment of TBIR is evolving. The NIH has published its success with an immunosuppressive protocol using rituximab, cyclophosphamide, and dexamethasone. We present a case of TBIR and highlight the outcome of treatment with this immunosuppressive protocol.

Case Presentation: A 42-year old Afro-Jamaican female with a past medical history of newly-diagnosed systemic lupus erythematosus (SLE) was transferred to our facility for severe hyperglycemia without diabetic ketoacidosis that was unresponsive to high doses of insulin. She had initially presented with a four month history of progressive generalized weakness, weight loss, facial hyperpigmentation, and patchy alopecia. The hyperglycemia was managed with Metformin and increasing doses of IV insulin and SC U-500 regular insulin, as high as 2,200 Units per 24 hour period. Rheumatologic workup was consistent with SLE. A presumptive clinical diagnosis of TBIR was made based on her severe hyperinsulinemia, lack of ketoacidosis, normal adiponectin, low triglycerides, and a high c-peptide-to-insulin ratio. Therapy was initiated using the NIH protocol: two 14-day cycles of rituximab, dexamethasone, and cyclophosphamide. Meanwhile, serum was sent to a research lab in Cambridge, UK, and was found to be strongly positive for insulin receptor antibodies. Subsequently, due to cytopenia on cyclophosphamide, azathioprine was substituted. After treatment she was able to taper off insulin therapy. She was discharged with euglycemia on metformin 2gm daily and azathioprine 100mg daily.

Discussion: Primary insulin signaling defects are extremely rare and result from either insulin receptoropathies or autoantibody activity against the insulin cell-surface receptor. TBIR is the only described syndrome involving anti-insulin receptor antibodies (AIRAs). TBIR is so uncommon that the exact prevalence is unknown. The largest cohort was followed by the NIH and included 24 patients over the course of 28 years. That cohort demonstrated that patients are typically female, middle age, have an African ancestry and concomitant autoimmune conditions (most commonly SLE, Sjogren syndrome or MCTD). Patients presenting with insulin resistance refractory to large doses of insulin and a history of autoimmune disease should prompt consideration of a Primary insulin signaling defect. Diagnosis of TBIR is based on the demonstration of AIRAs. The clinical course of TBIR is biphasic: an initial hyperglycemic phase, characterized by weight loss and acanthosis nigricans; and a subsequent hypoglycemic phase, characterized by a rapid reduction in exogenous insulin requirements and high risk for hypoglycemia. Management should be in consultation with an Endocrinologist. There is no standard therapy but various immunotherapies have been studied. To our knowledge, this is the fifth reported case of successful treatment using this protocol outside of the NIH.

References
North Carolina-Clinical Vignette-Poster Finalist
Maggie C Moses, MD

Title: A Perplexing Case of Paralysis in a Patient with Myasthenia Gravis

Authors: Maggie Moses, M.D., Neil Stafford, M.D.

Introduction: When a patient with an established neuromuscular disease presents with an atypical distribution of weakness you must search for an alternative diagnosis. Periodic paralyses are a rare group of inherited or acquired ion-channel myopathies that result in acute attacks of paralysis during times of hypokalemia or hyperkalemia. We report a case of periodic paralysis in a patient with myasthenia gravis.

Case Presentation: A 22-year-old male presents to the Emergency Department after developing acute onset weakness associated with three episodes of non-bilious, non-bloody emesis. He has a history of seropositive myasthenia gravis status post thymectomy 15 years ago and now off therapy for the past seven years without any flare of disease. He reports that the day prior to presentation he felt diffuse myalgias of which he attributes to recent exercise, something he hadn’t done in the past year. Physical exam is pertinent for sinus tachycardia, his thyroid gland is mildly enlarged without nodules, and he has distal greater than proximal muscle weakness with 3/5 strength in bilateral distal muscle groups. His labs are remarkable for a potassium of 1.4 mmol/L and he is given intravenous and oral potassium supplementation before admission to the hospital. Working differential diagnosis at this time included hypokalemic periodic paralysis, atypical flare of myasthenia gravis, or thyrotoxic periodic paralysis. Thyroid studies reveal a TSH of <0.01 mIU/mL, Free Thyroxine (T4) of 4.32 ng/dL (reference range 0.52-1.21 ng/dL) and Free Triiodothyronine (T3) of 13.07 pg/mL (reference range 2.20-3.80 pg/mL). Thyrotropin Receptor Antibodies return back positive and thyroid ultrasound scan is consistent with thyroiditis (heterogenous echotexture and hypervascular vascularity). He is started on methimazole without accompanying beta-blocker therapy out of concern for worsening his myasthenia. The patient’s potassium is monitored closely, requiring 120mEq of supplementation to normalize and luckily not resulting in rebound hyperkalemia. After initiation of methimazole and with improvement in serum potassium levels the patient regains full motor strength in all extremities. He is discharged with close endocrinology follow up.

Discussion: This case shattered numerous diagnostic biases in order to achieve an accurate and lifesaving diagnosis for our patient. Thyrotoxic periodic paralysis is a rare variant of hypokalemic periodic paralysis in which hyperthyroidism Results: in transient neuromuscular weakness. Precipitating factors include exercise and this is thought to have triggered our patient’s paralytic attack. Restoration of a euthyroid state is curative of the transient paralysis and thus highlights the importance of checking thyroid studies in patients presenting with hypokalemia and weakness.
North Carolina-Clinical Vignette-Poster Finalist
Jeffrey P Roan, DO

Title: Chest Wall Rigidity in the ICU after Fentanyl administration "A Rare Side Effect of a Common Medication"

Authors: Jeffrey Roan, DO; Navin Bajaj, MD; Field Davis, DO; Natalie Kandinata, B.S.

Introduction: Intubation and mechanical ventilation are often necessary in the critically ill. Commonly used drugs during mechanical ventilation includes opioids, propofol, dexmedetomidine and benzodiazepines. It's imperative to be able to trouble-shoot ventilatory alarms and uncommon physical exam findings in an unstable patient. We present a rare side effect of a commonly used drug in the ICU.

Case Presentation: A 56-year-old female was admitted for acute hypercapnic respiratory failure secondary to COPD. She was started on non-invasive positive pressure ventilation(NIPPV), however her respiratory status worsened and was transferred to the ICU. She persisted in respiratory distress despite NIPPV and was intubated and placed on mechanical ventilation. Fentanyl 100 mcg, Versed 4mg, and Rocuronium 50mg were used for induction. Chest radiograph confirmed placement of endotracheal tube(ETT) and showed no acute cardiopulmonary disease. Fentanyl drip was subsequently initiated at 50mcg/hr.

Repeat examination revealed jaw clenching and generalized muscle stiffness. The patient was afebrile and labs revealed normal renal function, creatinine kinase, and electrolytes. Versed infusion was initiated at 4mg/hr for concern of insufficient sedation, however, her new findings persisted. The patient became hypotensive with elevated peak and plateau pressures 35-45cmH2O. Placing an ETT bite block failed to improve airway pressures. Differentials were discussed regarding common causes of high airway pressures. Ultimately the team pursued opioid-induced muscle rigidity as the cause. Fentanyl was discontinued and Naloxone was administered. Subsequently, the patient had complete reversal of masseter rigidity, skeletal stiffness and hypotension. Peak and plateau pressures decreased to 10-25cmH2O. She was extubated 2 days later after resolution of her respiratory illness and transferred to the medical floor.

Discussion: It was initially thought the paralytic agent had caused the generalized rigidity. However, the patient remained afebrile with normal electrolytes and creatinine kinase. Hypotension is commonly experienced during intubation due to medications used for induction and the increased intrathoracic pressures from positive pressure ventilation. Despite the elevated Peak and Plateau pressures, there was no Auto-PEEP. Since hypotension had persisted, the possibility of an anaphylactic reaction causing bronchospasms was considered. Absence of wheezing, tachycardia, or cutaneous findings made this less likely.

Recently it was shown that even low fentanyl doses can cause thoracic rigidity. Dimitriou revealed muscle rigidity following an unusual low dose of fentanyl with onset of symptoms within 40 seconds of administration, similar to our patient. Risk factors have been identified including dose/rate of injection; extremes of age(newborns, elderly); critical illness with neurologic or metabolic diseases and use of dopaminergic medications.

Treatment includes discontinuing the offending agent, using reversal agents(naloxone), short-acting neuromuscular blockade and continued support with mechanical ventilation. Pre-administration of dexmedetomidine has also been shown in animal studies to decrease the incidence.
In our case, chest wall rigidity was manifested with decreased chest wall compliance, leading to elevated plateau pressures. This resulted in hypotension and ventilatory failure.\textsuperscript{16,18,19} Opioids are commonly used for continuous analgesia, it’s imperative for practitioners to be cognizant of this rare phenomenon and its early recognition to avoid complications.

References

North Carolina-Clinical Vignette-Poster Finalist
Ana Snelling

Title: Eye Missed the Diagnosis

Authors: Ana Snelling, PGY-1, Joanna Young, PGY-2, Thomas Montgomery, Internal Medicine, Carolinas Medical Center, Charlotte, NC

Introduction: Von Hippel-Lindau (VHL) is a familial neoplastic condition characterized by both benign and malignant tumors. Due to the many organ systems it affects, VHL requires multiple surveillance strategies and, if undiagnosed, could have fatal consequences.

Case Presentation: A 63-year-old woman with a past medical history of left eye hemangioblastoma status post eye implant and hypothyroidism, presented to the emergency room for ataxia and abnormal neuroimaging. She underwent extensive evaluation for disequilibrium by an Otorhinolaryngologist and Neurologist. Neurology ultimately ordered a CT of her head for further evaluation as she could not undergo a MRI due to a left eye implant. The head CT showed a three-centimeter left superior cerebellar mass with resulting obstructive hydrocephalus and mild upward cerebellar herniation. She was advised to go to the hospital for further workup of the cerebellar mass. The patient was up to date on her cancer screenings, including colon, breast, and cervical cancer. To rule out metastatic etiology, a CT chest, abdomen, and pelvis was ordered; it was within normal limits. Following the CT, the patient underwent a suboccipital craniotomy to characterize the tumor and to resect the mass. The pathology was consistent with a hemangioblastoma. Based on the patient’s past history of retinal hemangioblastoma at age 17 and now a cerebellar hemangioblastoma, it is highly likely that she has VHL.

Discussion: Von-Hippel Lindau is mainly an autosomal dominant disorder, however 20% of cases arise from de novo mutations. Patients are screened for VHL if they have a single manifestation of VHL or if they have a close relative with VHL. The diagnosis is confirmed through detection of a mutation in the VHL gene. The most common tumor associated with VHL is a hemangioblastoma. VHL patients are screened annually for hemangioblastomas, as these tumors can often be a major cause of mortality from the mass effect on nearby central nervous system structures. Other tumors associated with VHL include renal cell carcinoma, pheochromocytoma, and pancreatic neuroendocrine tumors.

Even though these tumors can occur independently from VHL, our case serves as a reminder for internists to screen for VHL in patients who have tumors associated with the disease. There is no cure for VHL, therefore the primary goal of disease management is through surveillance strategies. These surveillance strategies can only be implemented in patients with a known diagnosis of VHL. Following screening guidelines can lead to early recognition of tumors and early intervention to reduce morbidity and mortality. This patient went 46 years without being diagnosed with Von Hippel-Lindau. She was fortunate to not have suffered fatal complications from undiagnosed VHL, however other patients who go unrecognized might not have a similar outcome.
North Dakota-Clinical Vignette-Poster Finalist
Mahammed Z Khan Suheb, MBBS

Title: Rare cause of refractory lactic acidosis with a fatal outcome

Authors: Khansuheb MZ, MD; Koponen M, MD; Khan H, MD; Phadke G, MD; Weiland T, MD

Introduction: Systemic inflammation and lactic acidosis are commonly associated with sepsis, and hematologic malignancies mimicking this presentation can be a challenge. Elevated lactate levels are often thought to be indicative of relative tissue hypoxia or type A lactic acidosis. Shock, severe anemia, and thromboembolic events can all cause elevated lactate due to tissue hypoperfusion, as well as mitochondrial dysfunction thought to occur in sepsis and other critically ill states. Malignancy can also lead to elevation in lactate, a phenomenon described as type B lactic acidosis.

Case Presentation: • A 66-year old gentlemen presented with a 12-week history of fatigue, intermittent fevers, pedal edema, and a 2-week history of dysuria, urgency and hesitancy.
• Physical exam revealed splenomegaly. No skin rashes or inflamed joints were noted.
• Initial labs included CBC with peripheral blood smear showed Hgb 10.4 g/dL, platelets 129 k/uL, peripheral blood smear showing microcytic anemia without schistocytes. Renal function panel showed creatinine 2.4 mg/dL, and BUN 65 mg/dL. Urinalysis showed hematuria and proteinuria (500 mg/day). Serum Electrophoresis was unremarkable. With worsening renal function a renal biopsy was performed revealing acute tubular necrosis (ATN) (Fig. 1). Cultures and infectious serology along with Autoimmune workup were negative. Patient received a steroid course with concern for secondary hemophagocytic lymphohistiocytosis (HLH). Bone marrow biopsy was negative for malignancy and other disorders.
• Lactic acid continued to rise despite adequate fluid resuscitation and antibiotic therapy (Fig. 2). In search for the potential source of localized organ ischemia, CT imaging of abdomen and pelvis and angiography of the mesenteric vessels was negative. On Day 7 patient deteriorated with hypoxic respiratory failure and shock needing mechanical ventilation with vasopressor support. Patient had declining kidney function, metabolic acidosis and anasarca. Continuous renal replacement therapy (CRRT) was initiated. Ongoing CRRT was unable to stop rising lactate, peaking at 26 mmol/L. His shock state persisted despite several interventions. Consequently, patient experienced a fatal cardiac arrest. Autopsy revealed intravascular large B-cell lymphoma (IVLBCl)(Fig 3&4).

Discussion: This case report illustrates that type B lactic acidosis must be considered, especially in situations of elevated lactate without hypotension as was the case with this patient on initial admission. Fever, Hyperferritinemia, elevated CRP and Serum LDH, and kidney failure initially pointed towards an infections etiology. With Cultures and imaging negative both infectious etiology and ischemia were ruled out. His lactic acidosis continued to worsen with clinical course complicated by hypoxic failure and Circulatory shock. Despite attempts at resuscitation, lactate remained persistently elevated. IVLBCl is a rare extranodal lymphoma that may present as fever of unknown origin with neurologic, pulmonary, and hematologic manifestations. Bone marrow biopsies like in our patient can be normal in IVLBCl, and random skin biopsies often help with diagnosis. Type B lactic acidosis from malignancy is believed to be multifactorial including altered lactate metabolism from liver and kidney dysfunction and also lactate production by tumor cells due...
to growth factors promoting a high rate of glycolysis despite adequate tissue oxygenation (Warburg effect), i.e. type B lactic acidosis.

References


Title: Ipilimumab-induced hypophysitis in a patient with metastatic melanoma.

Authors: Hind Alameddine MD, Vincent Kang DO, Kim Jordan MD, Daryl Cottrell MD

Introduction: Immune checkpoint inhibitors include nivolumab, an anti-PD-1 antibody, and ipilimumab, an anti-CTLA-4 antibody. These new medications promote effector T-cell response to tumors, and are used to treat melanoma, lung, prostate, and renal cell carcinoma. Unfortunately, immune-related adverse events may occur with alteration of immune signaling pathways, including dermatologic, gastrointestinal and endocrine toxicities. We report a rare case of ipilimumab-associated hypophysitis in a patient with metastatic melanoma.

Case Presentation: A 67-year-old Caucasian female with stage IV esophageal melanoma presented with headache and intractable nausea. Medical history included papillary thyroid carcinoma, renal cell carcinoma, and synovial sarcoma of lung, all resected. PTEN, TP53, and BRAF mutation testing were negative. Due to progressive metastatic melanoma despite 18 months of nivolumab, therapy was changed to ipilimumab. Her symptoms started after the 3rd session of ipilimumab. Admission blood chemistries, CBC, and head CT were normal. However, brain MRI showed pituitary gland enlargement of 11 mm, consistent with hypophysitis. Hormonal evaluation included: luteinizing hormone 2.2 mIU/mL, TSH 0.14 uIU/mL (on suppressive therapy for thyroid carcinoma), and normal IGF-1(36 ng/mL), ACTH (22 pg/mL), and prolactin (13.7 ng/mL). Random cortisol level was 8.6 ug/dL, and she had no evidence of diabetes insipidus. Symptoms dramatically improved with prednisone, 100 mg PO daily, and she was discharged after 4 days on a gradual steroid taper. Repeat brain MRI at 2 months showed resolution of pituitary enlargement.

Discussion: Hypophysitis is estimated to occur in 1 in 7-9 million persons annually\(^1\), but pituitary inflammation associated with new immunotherapeutic agents is increasingly reported, estimated to occur in 9-15% of patients taking ipilimumab\(^1-3\). Male sex and older age appear to increase risk\(^2\). Symptoms related to pituitary enlargement and/or impaired secretion of target hormones generally occur 2-3 months after therapy initiation (range 5–36 weeks)\(^3\). Headache is most common, followed by fatigue, nausea and vomiting. Both transient and permanent disruptions of pituitary function are reported; symptoms of impaired hormone secretion vary, dependent on involved axis. Unlike hypophysitis from other causes, diabetes insipidus and visual disturbances from optic chiasm compression are unusual with ipilimumab toxicity\(^1-3\). Biopsy is not required if immunotherapy is the suspected cause of hypophysitis, but biochemical testing and imaging are needed. Pituitary and target tissue hormones should be measured. Gadolinium-enhanced MRI usually shows pituitary enlargement, but enhancement of pituitary stalk and normal pituitary are reported\(^4,5\). Management depends on severity: for low grade toxicity, drug continuation and close surveillance is recommended while more severe toxicity mandates medication discontinuation and high dose steroid therapy (1 mg/kg of prednisone or equivalent), followed by a 4-week taper\(^6\).

Teaching Point: Rare adverse effects may become more frequent with increasing use of immune checkpoint inhibitor therapy. Early recognition of hypophysitis is important for best outcomes.

References
Title: Necrotizing pneumonia secondary to Nocardia in a patient with myasthenia gravis

Authors: Hind Alameddine, MD and Kim Jordan MD, FACP

Introduction: Nocardia pneumonia is rare, and occurs primarily in patients who are significantly immunocompromised from disease and/or treatment with immunosuppressive drugs or long-term high-dose corticosteroids. Diagnosis is often delayed, up to 45 days in some reports, secondary to variable clinical presentation. Additionally, physicians may be less likely to consider nocardiosis early in immunocompetent patients; however, one-third of affected patients have normal immunity. We describe fatal Nocardia pneumonia in a patient with myasthenia gravis (MG) treated with low dose prednisone.

Case Presentation: A 72-year-old male with MG, atrial fibrillation and congestive heart failure was admitted to an outside hospital with pleuritic chest pain, cough productive of green sputum, and fever of 103.2 F. Medications included pyridostigmine and prednisone 10 mg daily. Amoxicillin-clavulanate was initiated, but his condition worsened prompting transfer to our hospital. Chest CT showed pleural effusions, atelectasis versus pneumonitis, and changes suggestive of volume overload. Despite therapy with amoxicillin-clavulanate and intravenous diuretics, he developed respiratory failure requiring mechanical ventilation within 48 hours. Thoracentesis yielded 250cc of cloudy fluid consistent with exudative neutrophil-predominant process, but cultures and cytology were negative. A parapneumonic process was suspected, and prednisone was increased to 40 mg daily, but he did not improve. On day 10, thoracic ultrasound showed multiple areas of fluid loculation, prompting chest tube insertion and intrapleural lytic therapy. All cultures remained negative, but he developed shock, and right lower lobe lung collapse. Thoracotomy revealed necrotizing pneumonia. Two liters of putrid, brown pus were removed and grew Nocardia otitidiscaviarum at 5 days. Antibiotic therapy was changed to trimethoprim-sulfamethoxazole and meropenem; however, he failed to improve, could not be extubated, and his family eventually withdrew care.

Discussion: Nocardiosis can present as acute, subacute or chronic disease. Symptoms are variable and include: productive or nonproductive cough, dyspnea, chest pain, hemoptysis, fever, sweats, weight loss, and fatigue. Radiographic findings also vary, with nodular, alveolar, or interstitial pattern. Cavitation and pleural effusions occur in 30% and 35% of cases, respectively. Only a few cases of severe nocardiosis are reported in patients with MG treated with high dose steroids or other immunosuppressant therapy. Though our patient was taking 10 mg daily prednisone, the CDC describes “immunosuppressive steroid dose” as ≥2 weeks of prednisone, or equivalent, at 20 mg daily or 2 mg/kg body weight. MG likely contributed to respiratory failure and unsuccessful extubation. Combination therapy (TMP-SMZ plus a second agent like amikacin, imipenem, or meropenem) is recommended until clinical improvement occurs and because of emerging drug resistance, especially among N. farcinica and N. otitidiscaviarum isolates. Following species confirmation and antimicrobial susceptibility testing, single-drug therapy may suffice (6-12 months if immunocompetent; 1 year if immunosuppressed). Mortality rates range from 40% to 64%.

References

Ohio-Clinical Vignette-Poster Finalist
Katlyn Baxter, DO

Title: Is That Elevated Creatinine Really Acute Kidney Injury?

Authors: Katlyn Baxter, DO, Ohiohealth Riverside Methodist Hospital; Ryan Lauf, DO, Ohiohealth Riverside Methodist Hospital; Kim Jordan, MD, Ohiohealth Riverside Methodist Hospital

Introduction: Acute kidney injury (AKI) is defined as an abrupt and potentially reversible decline in the glomerular filtration rate (GFR), with resultant elevated serum blood urea nitrogen (BUN) and creatinine (SCr), as well as other metabolic waste products. AKIN Criteria for AKI include: 1) an absolute increase in SCr ≥0.3mg/dL from baseline; 2) an increase in serum creatinine concentration of ≥50%; or 3) oliguria of <0.5mL/kg per hour for 6 hours. We present an unusual diagnosis in a young woman admitted one week post-gynecological surgery with symptoms of oliguria, ascites and serologic criteria for AKI.

Case Presentation: A 27 year old female underwent uncomplicated laparoscopic right oophorectomy for an ovarian cyst. Her post-procedural recovery was uneventful and she was discharged home the same day. Over the following week, she noticed increasing abdominal distention, diffuse abdominal pain, severe nausea, and poor oral intake. She also reported minimal urine output for approximately 3 days. Her only medication was oxycodone-acetaminophen which was prescribed for post-procedural pain. She sought evaluation in her local emergency department 6 days post-procedure. Physical examination revealed a diffusely tender, distended abdomen. Cardiopulmonary examination was unremarkable, no peripheral edema. Initial laboratory studies found hyperkalemia 5.2 mmol/L, bicarbonate 11mmol/L and elevated SCr to 9.77 mg/dL with BUN of 81 mg/dL. Urinalysis Results: were significant for 30mg/dL protein, small blood, leukocyte esterase and 1 granular cast but no nitrites.

A urinary catheter was placed with immediate return of 200cc of clear fluid; however, over the next 2 hours, urine output was 1300cc. Computed tomography (CT) of her abdomen and pelvis revealed protrusion of the urinary catheter through the dome of the bladder into the intraperitoneal space, large ascites and free intraperitoneal air. The kidneys appeared normal without hydronephrosis. She was transferred to our facility for emergent surgery which revealed two injuries to the bladder, one anteriorly and one posteriorly, immediately below the suprapubic port site incision suggesting a trocar injury to the bladder. A copious amount of clear fluid was drained from the peritoneum consistent with urinary ascites. Within 24 hours of surgical repair, the patient was significantly improved, demonstrated normal urine output, normal serum creatinine of 0.82 mg/dL with BUN of 19 mg/dL. Electrolytes and anion gap normalized within 48 hours.

Discussion: This case reminds clinicians to consider bladder injury in patients presenting with post-operative ascites, clinical and laboratory parameters suggestive of AKI. Though our patient’s symptoms were consistent with uremia, her markedly elevated creatinine and BUN were related to peritoneal absorption of urea and creatinine from urinary ascites due to bladder injury, previously referred to as “reverse auto-dialysis” or “pseudo-AKI”. The metabolic derangements associated with urinary ascites underscore the importance of early identification and correction of the bladder injury.

References


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Ohio-Clinical Vignette-Poster Finalist
Chantelle Carneiro, MD

Title: Rare Presentation of Primary Adrenal Insufficiency in a Systemic Lupus Erythematosus (SLE) Patient without Antiphospholipid Syndrome (APLS)

Authors: Chantelle Carneiro, MD, MS1, Pichaya O-charoen, MD2, Soumya Chatterjee MD, MS, FRCP2, 1. Department of Internal Medicine, Cleveland Clinic, Cleveland, OH, 2. Department of Rheumatic and Immunologic Diseases, Cleveland Clinic, Cleveland, OH

Introduction: Primary adrenal insufficiency has rarely been reported in systemic lupus erythematosus (SLE) with antiphospholipid syndrome (APLS) resulting in bilateral adrenal hemorrhage. We report a case of SLE without APLS that presented with hypotension, fatigue, hyperpigmentation and gastrointestinal symptoms with elevated adrenocorticotropic hormone (ACTH) and a failed ACTH stimulation test.

Case Presentation: A 42-year-old male presented with abdominal pain, nausea, vomiting, hypotension, ischemic hepatitis, acute kidney injury secondary to acute tubular necrosis, polyarthralgia, pericardial effusion, anemia and severe thrombocytopenia. The patient had a positive antinuclear antibody (ANA), anti-Smith, anti-ribonucleoprotein (RNP), and anti-chromatin antibodies. Anti-phospholipid serologies, and his infectious work-up were negative. Based on his serologies the patient was diagnosed with systemic lupus erythematosus. However, the causes of his gastrointestinal symptoms and hypotension were unclear. His symptoms improved with high dose intravenous methylprednisolone, but his platelet count continued to decrease despite being on IVIG. The patient was subsequently treated with rituximab, which normalized his platelet counts and other lupus symptoms over a 3-month period. Prednisone was gradually tapered off over the course of a year. When his prednisone dose was below 5 mg daily, he developed severe fatigue, anorexia, weight loss, abdominal pain, nausea, vomiting, diarrhea, postural hypotension, and increasing hyperpigmentation of his face, extremities, palmar creases, lips, tongue, and hard palate. He also had relatively low blood glucose. Initially, hydroxychloroquine-induced hyperpigmentation was considered. However, given his other concurrent symptoms, further work up for adrenal insufficiency was pursued. He had an elevated ACTH level of 193 pg/mL [normal: 8-42 pg/mL] and a failed cosyntropin stimulation test. Computed tomography of the abdomen and pelvis did not reveal adrenal gland enlargement, calcification, mass, hemorrhage, or infarction. Interferon gamma release assay, thyroid function tests, calcium, blood glucose, HbA1C, and testosterone levels were normal. He did not have candidiasis, vitiligo or any other evidence of autoimmune polyendocrine syndrome. He was diagnosed with primary adrenal insufficiency. His symptoms significantly improved after being given high dose systemic glucocorticoids. He was re-treated with rituximab for thrombocytopenia. His prednisone was again tapered to 10 mg daily. ACTH level was re-tested with similar findings. Renin and Aldosterone levels were normal. Even though adrenal antibody and 21-hydroxylase antibody tests were negative, in the absence of an alternative explanation, his primary adrenal insufficiency was thought to be the result of an autoimmune process. Prevalence of adrenal autoantibodies reported in the literature ranges from 25-84% (1). During subsequent visits, his postural hypotension persisted requiring the addition of fludrocortisone. It is likely that this patient's autoimmune primary adrenal insufficiency was associated with his SLE.

Discussion: In patients with SLE, autoimmune primary adrenal insufficiency can present concurrently with a lupus flare. A high index of suspicion is needed for prompt diagnosis and treatment of this condition.

References
Ohio-Clinical Vignette-Poster Finalist
Vincent Kang, DO

Title: Pegloticase-associated Hemolytic Anemia in an Elderly Patient

Authors: Vincent Kang DO, Hind Alameddine MD, Kim Jordan MD

Introduction: Pegloticase was approved in 2010 for treatment of chronic gout refractory to conventional therapy. It promotes metabolism of uric acid to allantoin, thus, reducing plasma uric acid and crystal formation. The elimination half-life of 10-14 days allows for infrequent dosing and decreased immunogenicity to the medication. We present a rare case of severe hemolytic anemia and methemoglobinemia in a female treated with pegloticase.

Case Presentation: A 73 year-old African-American female with heart failure, chronic refractory gout, gastrointestinal reflux, chronic anemia, and obstructive sleep apnea presented with 3 days of worsening dyspnea, fatigue and profuse, watery non-bloody diarrhea. One day prior to symptom onset, her rheumatologist administered her first pegloticase infusion for gout refractory to allopurinol. On admission, she required intubation for worsening respiratory status. Her hemoglobin was 3.9 g/dL (baseline 9.9 g/dL), with WBC 28.06 K/mcL and normal platelets. A methemoglobin level was 13%, haptoglobin < 10.0 mg/dL (33.0-171.0 mg/dL), LDH 4370 U/L (100-250 U/L), and total bilirubin of 5.6 mg/dL (direct 1.3 mg/dL), consistent with hemolysis. Direct Coombs test was negative. Peripheral blood smear showed occasional schistocytes with rare bite cells. She improved with packed RBC transfusion, and was extubated the next day. Stool was positive for Clostridium difficile, successfully treated with vancomycin. Non-oliguric acute kidney injury, thought secondary to heme pigment-induced acute tubular injury, responded to hydration. On day 6, she required prednisone for acute gout affecting her right 3rd PIP and right wrist. Eventually, a screening glucose-6-phosphate dehydrogenase (G6PD) level (ordered on admission) returned at 7.2 U/gHb (normal 8.8 - 13.4). She was discharged after 9 days of hospitalization, on prednisone for acute gout, and plans for confirmatory G6PD deficiency testing.

Discussion: Though rasburicase-induced methemoglobinemia and hemolysis are well-described in patients with G6PD deficiency, few reports of pegloticase-associated methemoglobinemia and hemolysis exist; we found 3 cases\(^1\)-\(^3\). Importantly, the G6PD enzyme protects RBCs against oxidative injury via NADPH production; if deficient, certain foods and known oxidant medications, including pegloticase, may cause harm. With pegloticase, uric acid conversion to allantoin generates hydrogen peroxide; this reactive oxygen species damages RBCs, causing hemolysis. Oxidation of ferrous iron to ferric iron leads to methemoglobinemia. Our patient had a history of chronic anemia, but no prior diagnosis or screening for G6PD deficiency. Patients at high risk, such as African or Mediterranean ancestry, require G6PD screening prior to therapy with rasburicase or pegloticase\(^4\), but some have suggested that all patients be screened prior to treatment\(^5\). Importantly, hemolytic anemia following pegloticase has been reported in one patient with initial negative G6PD testing, mandating close follow-up after treatment initiation. Treatment for pegloticase-associated hemolytic anemia includes transfusion and plasmapheresis in cases unresponsive to transfusion alone. Ascorbic acid has been used to treat pegloticase-induced methemoglobinemia in G6PD deficient patients\(^6\).

References

Title: Enterococcus Faecalis Infective Endocarditis and Colorectal Carcinoma: Case of New Association Gaining Ground.

Authors: Zubair Khan MD [1], Nauman Siddiqui MD [1], Wasif M. Saif MD [2]. 1. University of Toledo Medical Center, Toledo, Ohio, 2. Tufts Medical Center, Boston, Massachusetts

Introduction: Mostly Streptococcus Bovis (S. Bovis) bacteremia and endocarditis has been found to be associated with underlying colorectal cancer (CRC). The prevalence of CRC in patients with S. Bovis bacteremia undergoing colonoscopy is approximately 60%. Enterococcus Faecalis (E. Faecalis) is the most common cause of infection among enterococci, and is responsible for 63–81% of cases. E. Faecalis endocarditis is the third leading cause of infective endocarditis, being responsible for 5% to 15% of cases. The source of bacteremia in majority of these cases is undetermined and if identifiable, it is mostly Genito-urinary tract. There has been some evidence in favor of association between E. Faecalis endocarditis and hidden CRC. Here we report a case of E. Faecalis endocarditis associated with Stage 1 CRC.

Case Presentation: A 55 years old gentleman known to have non-ischemic cardiomyopathy with implantation of Automated Implantable Cardioverter Defibrillator (AICD) & atrial fibrillation presented with the complaint of low grade fever, productive cough, back pain, fatigue & weight loss. His labs revealed normocytic anemia (Hb 11.4 g/dl), Anti-HCV positive, blood cultures Positive for E. Faecalis 2 out of 2. Trans-esophageal echocardiography for work up of bacteremia revealed aortic valve vegetation. His CT abdomen and pelvis was negative. His social history was positive for being active smoker and 20 pack years of smoking history. Further work up done for the back pain revealed osteomyelitis of T7-T8 vertebrae and L4-L5 vertebrae. His history of unintentional weight loss of 30 pounds in two months with normocytic anemia prompted the work up for hidden malignancy. His AICD was also removed because of the high suspicion of seeding during bacteremia. He subsequently underwent colonoscopy that showed large 2cm x 2cm pedunculated polyp with 5 cm stalk found in sigmoid colon at 30 cm. The pathology showed well differentiated invasive mucinous adenocarcinoma, with focal squamous differentiation. The colorectal surgery team did a sigmoid colectomy. The pathology from that surgery showed minimal residual adenocarcinoma, negative nodes (18/18), and distal margin of 8.6 cm with tumor classification of T1N0M0. Meanwhile he was treated for endocarditis and osteomyelitis with IV antibiotics and started treatment for HCV after surgery. The stage 1 CRC was thus the source of E. Faecalis bacteremia and the resultant complications of endocarditis and osteomyelitis.

Discussion: The association between E. Faecalis endocarditis and CRC is yet to be established. Some data even suggest that the E. Faecalis may be responsible for mutagenesis of colonic cells and resultant malignancy and intestinal lesions that then lead to translocation of E. Faecalis into blood stream.

Conclusion: We conclude that in cases of E. Faecalis bacteremia and endocarditis with unidentified source, colonoscopy should be considered if feasible to rule out the diagnosis of CRC.
Title: Cervical spinal osteomyelitis with epidural abscess following an Escherichia coli urinary tract infection in an immunocompetent host.

Authors: Abdelmoniem Moustafa, Rowida Kheieldin, Hussam Alim, Zubair Khan, Mohammad Saud Khan

Introduction: Spinal Epidural Abscess (SEA) is uncommon with an incidence reported as 0.33–1.96 abscesses per 10,000 hospital admissions per year. The leading bacterial pathogen causing SEA is Staph aureus, which accounts for about two-thirds of cases caused by pyogenic bacteria. Escherichia coli (E. Coli) is a less common cause of SEA and it is usually after urinary tract infection. Patients in these cases usually have pre-existing risk factors such as diabetes, obesity, alcoholism, trauma and bone degeneration.

Case Presentation: A 69-year-old male with a past medical history significant for prostatitis was admitted with fever, altered mental status, progressive lower extremities weakness and frequent falls for 7 days. Upon admission, his WBC count was 21.8 Thou/mm3 with 89% segments. Urinalysis showed trace leukocyte esterase, few WBCs, and many bacteria. CRP and ESR were elevated at 113 and 110, respectively. He was pan cultured and then started on Vancomycin, Cefepime, and Acyclovir. Both blood and urine cultures grew E. Coli. Lumbar Puncture was also done 16 hours after presentation and CSF had 94 RBCs, 24 WBCs (16 % neutrophils and 46 % lymphocytes). It also showed significantly elevated protein level at 1140 mg/dl with no bacterial or Acid-Fast Bacilli growth. Myelin basic protein was elevated at 7.45 ng/ml. Glucose level was 66 mg/dl.

Neurology team recommended C-Spine MRI which demonstrated epidural abscess along the anterior and right lateral margin of the cord causing cord compression from C5 through C7, anterior peri-vertebral abscess from C4 through T2, marrow edema involving C6 and C7 vertebral bodies with increased signal in the intervertebral disc space at C6-C7, and some enhancement of the vertebral bodies at C5 and C6. These findings consistent with osteomyelitis discitis. On the same day, the patient developed respiratory distress which required intubation and transfer to the MICU. Next day, anterior cervical decompression with evacuation of anterior epidural abscess with fusion were done. The culture from the epidural abscess grew E coli. He was switched to IV ceftriaxone and was extubated successfully. He was then discharged to rehabilitation facility for physical therapy and completion of antibiotics.

Discussion: The classical diagnostic triad of SEA consists of fever, spinal pain, and neurologic deficits. However, only a small proportion of patients have all three components at presentation. This is the first case described of a spontaneous cervical spinal osteomyelitis and epidural abscess caused by E coli following a UTI; presenting with progressive weakness of both lower extremities, decreased sensation in the upper extremities and bowel and urinary retention, in a previously healthy individual with no risk factors.
Ohio-Clinical Vignette-Poster Finalist
Jillian Pattison, DO

Title: Nephrotic Syndrome Secondary to Waldenstrom Macroglobulinemia: An unusual occurrence

Authors: Jillian Pattison DO, Ruta Arays MD, Kim Jordan MD FACP

Introduction: Waldenstrom macroglobulinemia (WM) is a rare lymphoplasmacytic lymphoma with a reported incidence of approximately three per million people per year. Disease classically presents in white males in their 6th decade of life. Onset is often insidious; patients may be asymptomatic, or complain of weakness, anorexia, and weight loss. Additionally, symptoms related to hyperviscosity can occur. Renal involvement from WM is reported, but is unusual. This case highlights nephrotic syndrome as the initial clinical presentation of WM in a middle aged woman.

Case Presentation: A 54-year-old white female was admitted for acute onset of bilateral lower extremity swelling, and foamy urine. She admitted to recent viral illness, with lingering fatigue. Her medical history included adult-onset type 1 diabetes mellitus, diagnosed at age 43. Her diabetes was well-controlled, with recent HbA1C of 5.7%, and she had no known nephropathy or retinopathy. Laboratory studies showed normal serum creatinine of 0.6 mg/dL and BUN of 12mg/dL. Urine protein to creatinine ratio was 15.7 and hyperlipidemia was noted, confirming a clinical diagnosis of nephrotic syndrome. Additional evaluation included elevated ANA 1:80, homogenous pattern, and normal C3 and C4 levels. A kidney biopsy was completed and immunofluorescence stains revealed prominent IgM and kappa immune deposits. Electron microscopy displayed focal foot process broadening, consistent with minimal change disease (MCD). There was no evidence of nodular diabetic nephropathy. Serum protein electrophoresis found elevated gamma globulin at 2.6 g/dl, and subsequent bone marrow biopsy revealed heavy infiltration by small B-cell lymphoma. Morphology and immunophenotype were compatible with either lymphoplasmacytic lymphoma or marginal zone lymphoma. Computed tomography of the chest, abdomen, and pelvis found mild intraperitoneal and retroperitoneal adenopathy without splenomegaly. Hematology/oncology physician initially recommended prednisone and furosemide therapies. Gene mutation testing revealed MYD88 mutation suggesting a diagnosis of WM, and rituximab was initiated. Cryoglobulin testing was positive and blood viscosity testing was within normal limits.

Discussion: This case describes a very rare presentation of MCD and nephrotic syndrome as the presenting sign of WM, and emphasizes the diagnostic importance of renal biopsy based on clinical scenario. Renal manifestations in WM are uncommon; when reported, microhematuria and mild proteinuria are most seen. Nephrotic syndrome is only reported in about 7% of WM cases, and almost always occurs secondary to amyloidosis; MCD secondary to WM is quite rare. Renal biopsy was important in establishing our patient’s diagnosis. Though she was a known diabetic, it is important to consider that moderate increased albuminuria (microalbuminuria) generally occurs after a mean duration of 10-15 years in 20-30% of patients, and not all progress to macroalbuminuria. Patients generally have poorly controlled blood glucose levels, and diabetic retinopathy is usually present.
Title: Dilaudid IV for Me: Genetic Variation in Opioid Metabolism Affecting Analgesia

Authors: Gabrielle Sabino, DO and Julie Coffman, MD

Introduction: Increasing opiate abuse and overdose demand physician vigilance when prescribing. Adequate treatment of patients with chronic pain can be challenging as various factors contribute to differences in opiate efficacy. Pharmacogenomics and genotype testing have identified genetic differences in rates of opioid metabolism. Thus, select patients may have valid reasons for opiate preference or need for increased dosing. The following case demonstrates application of genetic testing in assessing a patient’s increased opioid requirements.

Case Presentation: A 39-year-old female with chronic pain from C4 transverse myelitis was admitted for obstructing nephrolithiasis and urosepsis. Her home pain management regimen included hydromorphone 40mg orally every 4 hours. Because of NPO status, she was transitioned to scheduled intravenous hydromorphone, 6mg IV every 3 hours with 5 mg IV every 3 hours as needed (PRN). However, concern about high opioid requirements resulted in palliative care consultation. Patient-controlled analgesia with hydromorphone 7mg IV every 3 hours and 4mg IV every 2 hours PRN was prescribed. Pharmacogenomic testing for 2D6 and 2C19 cytochrome p450 activity was completed and demonstrated extensive ultrarapid metabolism at 2C19, justifying high opioid requirements. Her pain improved over the next few days, she was converted to her home regimen, and discharged to follow-up with her pain physician.

Discussion: Many opioids are metabolized via the cytochrome p450 enzyme; variations in gene expression and function, particularly cytochrome 2D6, 2C9 and 2C19, can affect metabolism, efficacy and elimination. Pharmacogenomics testing classifies patients as normal, overactive, underactive, or minimally active/inactive opioid metabolizers. Identification of “metabolizer type” allows individualization of pain control. For example, overactive metabolizers experience decreased level and/or duration of analgesia and often require higher, more frequent dosing; minimally active or inactive metabolizers with poor elimination have higher risk of overdose secondary to increased drug levels. Improved, safer analgesia may occur with opiates undergoing glucuronidation for activation (oxymorphone, morphine, hydromorphone, tapentadol) rather than cytochrome p450 metabolism. Adjuvant analgesics or routes of administration that bypass liver metabolism (buccal, sublingual, intrathecal, intravenous, subcutaneous) may be considered. Candidates for pharmacogenomics testing for opioid metabolism include patients who use 150mg oral morphine equivalent daily or repeatedly report poor analgesic effect despite appropriate opioid adjustment. Additionally, consider testing in patients who require increased dental/surgical anesthesia, have adverse reactions to alcohol, have limited relief from short-acting opioids (1-2 hr), or require higher than expected opioid dose. Not all variations in cytochrome P450 metabolism have a significant clinical impact, but it is important to recognize those patients who may be affected. Correct identification of these select patients avoids labeling as “drug seekers”, and validates opiate requirements and preferences for best analgesic relief.
**Ohio-Clinical Vignette-Poster Finalist**  
**Divya Sachdev, MBBS**

**Title:** An Interesting Case of Heart Block  

**Authors:** Divya Sachdev, MD. Vivek Nagaraja, M.D.

**Introduction:** 64-year-old lady presented with gradually progressive forgetfulness, confusion, dyspnea on exertion, non-productive cough and fatigue. In addition, she had poor appetite and weight loss (10 lbs). Due to the deteriorating mental status, her husband brought her to the hospital.

**Case Presentation:** On examination, BP-83/54mmHg, P- 53/min, she had a flat affect and answered to questions vaguely. She scored 14/30 on Montreal cognitive assessment indicating cognitive impairment. She had posterior cervical and left axillary lymphadenopathy, and raised hyper-pigmented and erythematous lesions on the anterior chest. EKG revealed a 2:1 AV Block and intermittent complete heart block. Laboratory work-up revealed anemia, (Hb-9.8g/dL) and high TSH (27.28 µIU/mL) comprehensive metabolic panel- unremarkable. Cardiac MRI showed delayed myocardial enhancement of the septal wall and anteroseptal wall, and preserved ejection fraction (70%) indicative of probable infiltrative cardiomyopathy. CT scan of the brain showed chronic small microvascular changes. CT angiogram of the chest showed mediastinal and axillary lymphadenopathy, small bilateral pleural effusions, and no pulmonary emboli. Due to the heart block and probable infiltrative cardiomyopathy, she underwent an implantation of dual chamber implantable cardioverter defibrillator. Sarcoidosis was the working diagnosis at this time given mediastinal lymphadenopathy and infiltrative cardiomyopathy. A core biopsy of the left axillary lymph node showed lymphocytic proliferation with necrotic changes, but no granulomas or histiocytes.

For mental status changes, she underwent a lumbar puncture; CSF studies revealed elevated protein (52 mg/dL), normal glucose,(42mg/ dL) no white cells, negative oligoclonal bands, and normal IgG index. An electroencephalogram showed severe diffuse encephalopathy with an epileptogenic focus in the posterior cortical structure. Neurology team was consulted and she was placed on anti-epileptic therapy. In parallel to the above evaluations, she had serologic evaluations for connective tissue disorders, to explore other potential systemic conditions. She had an elevated ANA(1:2560, homogenous pattern), Anti-dsDNA (1:640), positive anti-Smith, low complements, and polyclonal hypergammaglobulinemia. She had non-nephrotic range proteinuria (2 g/24hrs). Given the proteinuria and the serologies, a renal biopsy was performed, and the histopathology revealed changes consistent with a combination of class III and class V lupus nephritis. The final diagnosis was systemic lupus erythematosus (SLE); she was started on high dose oral prednisone, hydroxychloroquine and mycophenolate mofetil.

**Discussion:** Conduction defects in SLE have been described in infants born to mothers with anti-Ro antibodies. However, conduction disturbances in adults with SLE are rare but documented, in the form of all types of atrioventricular (AV) block, bundle branch block, sinus tachycardia, atrial fibrillation, and atrial ectopic beats. The pathology is thought to be nodal artery occlusive lesions with secondary collagen degeneration and fibrosis of AV and SA nodes. In our case, the lady had complete heart block,
neurological and renal involvement, and lymphadenopathy due to SLE. She had significant improvement in her cognition after the initiation of immunosuppressive therapy.
Ohio-Clinical Vignette-Poster Finalist
Andrew Schaefer, MD

Title: The Use of Extracorporeal Carbon Dioxide Removal in Status Asthmaticus

Authors: Andrew Schaefer, MD; Derrick Herman, MD; Emily Amin, MD; Maria Lucarelli, MD

Introduction: Status asthmaticus is a life-threatening asthma exacerbation characterized by hypoxemic and/or hypercapnic respiratory failure. Management for status asthmaticus has changed little over the past several decades with inhaled bronchodilators and corticosteroids forming the backbone of therapy while magnesium, heliox, neuromuscular blockade, ketamine and bilevel positive airway pressure ventilation have been used as adjunctive therapy. Extracorporeal carbon dioxide removal (ECCO2R) has the potential to correct hypercapnia and/or hypoxia while providing a lung protective mechanism. ECCO2R is an artificial respiratory support in which carbon dioxide is removed from the blood through an extracorporeal gas exchanger. Herein, we present a patient with status asthmaticus refractory to traditional treatment who was successfully managed with ECCO2R.

Case Presentation: A 69 year old male with a history of moderate persistent asthma treated with oral steroids due to medication noncompliance developed acute onset dyspnea. Paramedics administered nebulized albuterol and intramuscular epinephrine without improvement. In the emergency department, the patient was tachycardic, tachypnic, and oxygenating at 99% on bilevel positive airway pressure ventilation. He was unable to speak in complete sentences, and had accessory muscle use. Lung auscultation revealed decreased breath sounds bilaterally with diffuse wheezes. Chest radiograph revealed hyperinflated lungs. The patient received three nebulized treatments of albuterol and ipratropium bromide, intravenous (IV) methylprednisolone, and IV magnesium with poor response. After a failed trial of bilevel positive airway pressure ventilation, the patient was endotracheally intubated for respiratory failure and admitted to the intensive care unit (ICU). His arterial blood gas upon ICU admission demonstrated a pH of 7.142, a pCO2 of 85.0, pO2 of 109.0 and HCO3 of 28.4. He received continuous nebulized albuterol, heliox, IV ketamine infusion, and continuous neuromuscular blockade with cisatracurium without improvement in his acute respiratory acidosis over six hours. Cardiothoracic surgery was consulted for venovenous ECCO2R. Within two hours of initiation of ECCO2R, the patient’s respiratory acidosis improved and pH normalized. In less than four hours, the patient’s carbon dioxide was within normal ranges. His albuterol, methylprednisolone, and his ECCO2R settings were weaned. He was decannulated and successfully extubated 48 hours after initiation of ECCO2R. He was discharged on room air seven days after he presented.

Discussion: This case illustrates the impact of ECCO2R in improving hypercapnia and relieving respiratory acidosis in cases of status asthmatics refractory to traditional treatment. While case reports have demonstrated the safety and efficacy of ECCO2R in hypercapnic respiratory failure, future research is needed to cement its safety and efficacy for hypercapnic respiratory failure. Studies should also evaluate the optimal timing of ECCO2R. For example, early ECCO2R with bilevel positive airway pressure ventilation may be an effective alternative to mechanical ventilation. Current evidence suggests that ECCO2R for hypercapnic respiratory failure should be saved until maximal conventional therapy has failed to correct the patient’s hypercapnia.

References


Ohio-Clinical Vignette-Poster Finalist
Daniel Shonebarger, DO

Title: A Full-House is Not Always the Best Hand

Authors: Daniel Shonebarger, DO¹ Mitchell Stelzer, DO¹, Jacob Koopman, MS4², John Hatanelas, DO¹, Lucia Chowdhury, MD¹, Henry Wehrum, DO¹, Thomas Keeling, MD¹, OhioHealth Doctors Hospital, Columbus, OH, ™Ohio University Heritage College of Osteopathic Medicine, Athens, OH

Introduction: Infection of the endocardium, the innermost layer of the heart, is referred to as Infective Endocarditis (IE). It can be a serious, life-threatening illness with potential downstream health complications such as myocardial infarction, valvular insufficiency, congestive heart failure, glomerulonephritis and acute renal failure.

Case Presentation: A 52-year-old caucasian male with no known chronic medical conditions, presented to our hospital with complaints of gradually worsening dyspnea accompanied by lower extremity edema, orthopnea and dark urine. He was in notable respiratory distress and there was jugular venous distention, respiratory crackles and lower extremity edema on physical examination. Laboratory examination demonstrated leukocytosis, anemia and acute kidney injury. A urinalysis with microscopy showed proteinuria, hematuria, pyuria and red blood cell casts. Blood cultures were obtained and the patient was treated empirically with antibiotics.

The nephrology service was consulted due to the worsening renal function and ordered a renal biopsy as well as serum studies to evaluate potential causes of glomerulonephritis. The only positive or abnormal result was a positive anti-streptolysin O antibody titer. Examination of the renal biopsy specimen on light microscopy revealed cellular crescents in 10/11 glomeruli and 5/11 were fibrotic. Immunofluorescence showed “full-house” nephropathy, in which IgA, IgG, IgM, C3, and C1q all stained positively. The final diagnosis was rapidly progressive glomerulonephritis (RPGN). Transthoracic echocardiogram revealed a lesion on the aortic valve consistent in appearance with a vegetation. Blood cultures from admission grew Streptococcus mutans. Antibiotics were altered based on sensitivities and the infectious disease service was consulted. The patient subsequently required continuous renal replacement therapy (CRRT) for worsening hypoxia and volume overload. The cardiothoracic surgery service evaluated the patient and determined that the aortic valve required replacement. The patient underwent cardiac catheterization and subsequent coronary artery bypass grafting with aortic valve replacement. Despite excellent post-operative recovery, renal function never improved and he continues to require intermittent hemodialysis.

Discussion: The negative outcome of this case illustrates the need to broaden the treatment approach in cases of immune complex-mediated organ damage in the setting of infection. The dearth of treatment options is likely owed to the rarity of the condition as RPGN is a rare, but serious complication of IE and ours is the only documented case of infection with Streptococcus mutans resulting in RPGN with “full-house” nephropathy on immunostaining. This pattern is most commonly associated with Lupus Nephritis, class IV, which is treated with immunosuppressive therapy. Future studies will need to evaluate the utility of current immunosuppressive therapies in the setting of infection-induced “full-house” nephropathy.
Title: A Misleading presentation of pulmonary tuberculosis in an immunosuppressed patient

Authors: Kavitha Mattaparthi MD, Carmen Vesbianu MD

Introduction: It is not uncommon for pulmonary tuberculosis to be misdiagnosed as pneumonia. The diagnoses becomes particularly challenging when presenting in a chronically immunosuppressed patient, specifically one on a TNF-alpha antagonist. Many such patients have inconsistent physical findings, atypical radiological findings, and/or negative tuberculin skin tests. Given the growing use of biologic therapies to treat chronic inflammatory diseases, it is imperative we include active pulmonary tuberculosis in our differential to ensure early diagnosis and timely treatment.

Case Presentation: A 47-year-old Native-American male presented with persistent fever, cough, exertional dyspnea, confusion, night sweats, myalgias, and generalized malaise. Patient had a history of documented HLA-B27 positive spondyloarthropathy, latent TB appropriately treated in the 1990s, chronic right lower lobe densities dating back to 2012, and recurrent pneumonia treated with multiple courses of antibiotics. His medications included Ceroluzimab pegol, Leflunomide and Prednisone. Initial vitals significant for high-grade fever, tachycardia, and hypoxia. Exam revealed an ill-appearing male with inspiratory/expiratory wheezing and coarse breath sounds. Pertinent labs: leukopenia with normal absolute neutrophil count, mildly elevated CRP/ESR. Atypical panel showed elevated IgG levels to Influenza A, Influenza B, RSV, Chlamydia, Mycoplasma, and Adenovirus (all IgM levels normal). Serum/urine histoplasmosis and serum HIV antibodies were negative. PPD with no detectable induration. CXR showed right lower lobe bronchiectasis. CT chest with contrast showed persistent and worsening right lower lobe nodular densities. The patient was treated initially with IV Vancomycin and Meropenem, however due to persistent cough and fevers, this was changed to Azithromycin and Fluconazole for atypical and fungal coverage. CT biopsy-lung revealed necrotizing granulomas with no organisms identified on staining; bronchoscopy with BAL completed with cytology was positive for scattered acid-fast bacilli. Molecular probe testing confirmed Mycobacterium tuberculosis with AFB culture consistent with growth of pansensitive M. tuberculosis. Therapy for active pulmonary TB with isoniazid, rifampin, pyrazinamide, and ethambutol was initiated prior to obtaining final cultures.

Discussion: Pulmonary tuberculosis can be a challenging diagnosis, particularly in the immunosuppressed population. One study indicated that misdiagnosis of TB (often as pneumonia) may be occurring in more than 20% of TB patients diagnosed in inpatient and emergency department settings. Although typically associated with upper lobe infiltrates, lower lung field infiltrative disease is an atypical presentation of pulmonary tuberculosis, particularly in immunocompromised patients. All biologics predispose patients to a higher risk of infection; interestingly the risk of acquiring TB may be higher in specific anti-TNF agents like Ceroluzimab pegol or Golimumab and risk for reactivation may further increase with concomitant DMARD use. With a surge in the administration of biologics for chronic inflammatory diseases, we must aim to identify TB early for timely treatment and prevention of its infectious spread in the healthcare setting and community.

References

Oklahoma-Clinical Vignette-Poster Finalist
Marcus A Toschi, MD

Title: A Case of Granulomatosis with Polyangiitis in Late Pregnancy

Authors: Marcus Toschi, MD, Sarah Alvarez, MD, Matlock A. Jeffries, MD

Introduction: Granulomatosis with Polyangiitis (GPA), formerly known as Wegener’s Granulomatosis, is a vasculitis defined by necrotizing granulomatous inflammation, usually involving the upper and lower respiratory tract, predominantly affecting small and medium-sized vessels. Renal involvement with glomerulonephritis is also common. Among patients with GPA, approximately 90 percent have nasal, sinus, or ear involvement. The most common lower respiratory tract symptoms in GPA are cough, hemoptysis, dyspnea, and pleuritic chest pain. Most patients with GPA have c-ANCA, characterized by diffuse cytoplasmic staining and autoantibodies directed against proteinase 3 (PR-3). We present a case of GPA in late pregnancy.

Case Presentation: A 30 year-old Caucasian female at 31 weeks of pregnancy presented with a non-healing, left-sided hip abscess, productive cough, pleuritic chest pain, and sepsis. Initial x-ray showed multiple basilar opacities and a 4cm density in the right upper lobe concerning for pneumonia or septic emboli. The hip abscess was believed to be secondary to a steroid injection for streptococcal pharyngitis, 6 weeks prior. She had complaints of greenish-yellow phlegm, but did not endorse hemoptysis. On review of systems, she recalled multiple sinus infections and, during pregnancy, episodes of epistaxis. While admitted, the patient developed oral ulcers, small joint arthritis of bilateral hands, blood-tinged sputum, and deep vein thrombosis. A bronchoscopy revealed inflammatory fluid with neutrophil predominance. Serial x-rays demonstrated mediastinal widening and a new necrotic mass in the right upper lobe, despite continued antibiotics. Two weeks later, an ANCA titer by indirect fluorescent antibody demonstrated a low titer positive c-ANCA pattern at a 1:40 titer. Rheumatology was consulted. A confirmatory ANCA revealed high titer c-ANCA pattern (1:180) and strongly positive proteinase 3 antibody by ELISA at 117. Urinalysis did not reveal proteinuria. A diagnosis of Granulomatosis with Polyangiitis (GPA) without renal failure was made. Given the concern for pregnancy complication with high dose corticosteroids and rituximab, she underwent a caesarean section. After delivery, pulse steroids were administered, followed by an oral steroid taper and rituximab infusion, consisting of 1000mg every 14 days for a total of two doses. Her dyspnea, sputum, and arthritis resolved. In rheumatology clinic, the hemoptysis, pleurisy, and bloody nasal discharge had abated, though she did continue to have some persistent arthralgia.

Discussion: This case demonstrates an unusual presentation of GPA in a young, pregnant woman. At presentation, her history of productive cough, sepsis, and hip abscess was most concerning for a primary infectious process. The development of hemoptysis, arthritis, and oral ulcers hinted of a potential vasculitic process, although continued antibiotic therapy and additional invasive testing was performed. At the time of rheumatology consultation, the patient had seen multiple specialties including cardiothoracic surgery, infectious diseases, and pulmonary medicine. This case highlights the utility of vigilant history taking and the consideration of unusual rheumatologic conditions in young women.

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Ontario-Clinical Vignette-Poster Finalist
Eric Coomes, MD

Title: Persistent Profound Lactic Acidosis: An Unusual Case

Authors: Eric Coomes MD¹, Mark Cheung MD FRCPC², Steven Shadowitz MD FRCPC², ¹Department of Medicine, University of Toronto, Toronto, ON, Canada, ²Department of Medicine, Sunnybrook Health Sciences Center, Toronto, ON, Canada

Introduction: Lactic acidosis is often used as a surrogate for hypo-perfusion amongst critically-ill patients. However, as a product of anaerobic metabolism, its production may be driven by either hypoperfusion or fundamental abnormalities in aerobic metabolism. We present an unusual case of persistent profound lactic acidosis to elucidate this dichotomy.

Case Presentation: A 45 year-old-man presented to the hospital with exertional dyspnea. He was promptly referred to the internal medicine for laboratory abnormalities—most strikingly, a lactic acid of 22 mmol/L. He reported a several week history of progressive dyspnea, myalgias, acute on chronic abdominal pain, significant weight loss, and general malaise, felt to be initially precipitated by a viral infection. Aside from blindness from congenital cataracts, he had no past medical history. He was adopted, and his family history was unknown. Despite his metabolic derangements, he appeared well and hemodynamically stable. He had neither peritonitis, evidence of ischemia, sepsis, nor hypoperfusion. His examination was only significant for widespread pain with muscle palpation and mild proximal hip weakness.

His initial laboratory investigations demonstrated a profound lactic acidosis, concomitant respiratory acidosis, rhabdomyolysis, transaminitis, pancreatitis, pre-renal acute kidney injury, and mild hypoglycemia. His abdominal imaging demonstrated chronic pancreatitis.

He was treated with high-volume intravenous D10W/NS and thiamine. He remained clinically stable and his renal, pancreatic, and hepatic abnormalities resolved, yet his rhabdomyolysis and lactic acidosis persisted despite therapy. In consultation with his primary care provider and reviewing available laboratory data, it was identified that his rhabdomyolysis and anion-gap metabolic acidosis had been ongoing for at least a decade.

Extensive endocrine, hepatology, rheumatologic and toxicology investigations failed to yield an etiology. Electromyogram and nerve conduction studies demonstrated a mild myopathy. MRI brain was normal. Biochemical testing for inborn errors of metabolism was negative. A muscle biopsy was performed prior to discharge. Muscle pathology identified ragged red fibers, mitochondrial accumulation, and mosaic abnormalities in electron transport chain staining; pathognomonic features of mitochondrial myopathies.

Discussion: Mitochondrial diseases are uncommon and heterogeneous disorders arising from mutations of either nuclear or mitochondrial DNA-encoded genes.¹ The resultant dysfunction of oxidative phosphorylation can produce characteristic lactic acidosis and rhabdomyolysis.² Unequal cellular distribution of mutated mitochondria can produce highly variable clinical presentations ranging from isolated myopathies to multi-system disorders.¹ While patients with classical phenotypes may be diagnosed with upfront genetic evaluation, those with protean manifestations may have significant diagnostic delay.¹ Therapeutic regimens include electron transport chain cofactor supplementation and correction of downstream biochemical deficits.¹²
References


Ontario-Clinical Vignette-Poster Finalist
Eric Coomes, MD

Title: When Steroids are the Answer: A Case of Profound Hypereosinophilia

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Introduction: Eosinophilia is an uncommon hematologic abnormality with a diversity of causes ranging from allergic to neoplastic, but has the potential for significant end-organ impact. We present a case of profound eosinophilia in a patient with respiratory failure, highlighting an important fibroinflammatory etiology.

Case Presentation: A 76-year-old man with a history of asthma and heart failure was admitted to the intensive care unit with hypercapnic respiratory failure and profound eosinophilia > 10 x 10\(^9\) cells/L.

He had multiple preceding admissions for respiratory failure and eosinophilia, each treated as presumed pneumonia. Prior to admission he reported progressive dyspnea and edema. Review of systems was otherwise unremarkable and he had no significant exposures.

Examination revealed evidence of volume overload and diffuse lymphadenopathy. CT Chest demonstrated bilateral pulmonary consolidations and pronounced mediastinal lymphadenopathy.

To investigate the eosinophilia, extensive hematologic, infectious, and rheumatologic investigations were pursued. Flow cytometry and bone marrow biopsy demonstrated increased eosinophil precursors, but no evidence of malignancy. Lymph node fine needle aspirate demonstrated reactive hyperplasia. Bronchoscopy cultures failed to identify an infectious agent. Serologies were unremarkable besides an elevated rheumatoid factor. Immunoglobulins showed polyclonal IgG and IgE elevations.

Despite diuresis and broad-spectrum antimicrobials, he remained intubated without clinical improvement. With a tenuous respiratory status, the patient was unsuitable for lung biopsy. With a presumptive diagnosis of hyper-eosinophilic syndrome, the patient was started on pulse IV methylprednisolone with dramatic improvement; within 24 hours their eosinophilia resolved, and within 48 hours they were successfully extubated.

Results: of the lymph node core biopsy subsequently returned unexpected findings – sheets of plasma cells, with 50% of plasma cells staining positive for IgG4. Serum IgG4 level was also significantly elevated at 4.1 g/L.

In retrospect, our patient had pulmonary infiltrates, diffuse lymphadenopathy, hypereosinophilia, elevated RF, IgE, IgG, and serum IgG4 levels, and lymph-node invasion by IgG4-staining plasma cells – a diagnosis of IgG4-related disease was made.

Discussion: IgG4-related disease is an increasingly recognized multi-systemic fibroinflammatory disorder characterized by infiltrative IgG4 plasma cells with resultant storiform fibrosis and obliterative phlebitis. IgG4-related disease may effect any organ system with protean manifestations, but must be considered in patients with unexplained tissue or organ enlargement. The clinician may be alerted by characteristic laboratory clues: eosinophilia, elevated IgE, hypergammaglobulinemia, or hypocomplementemia. While diagnostic criteria require the combination of organ swelling or masses, elevated serum IgG4 levels, and
tissue infiltration with IgG4-positive plasma cells, other inflammatory and neoplastic disorders may elevate IgG4 levels and must be considered in the differential diagnosis. While the optimum treatment has yet to be established, initial treatment consists of glucocorticoid therapy, usually with significant clinical response, although certain patients may benefit from adjunctive immunosuppressive therapies.

References

Ontario-Clinical Vignette-Poster Finalist
Ankur Goswami, MD

Title: Primary adrenal insufficiency unveiled by a corticosteroid taper.

Authors: Ankur Goswami, Kevin Kumar Singh, Conor James Cox, Samir Raza, McMaster University, Department of Medicine, Hamilton, ON, Canada

Introduction: Primary adrenal insufficiency, or Addison’s disease is a debilitating endocrinopathy presenting with hyponatremia, hyperkalemia, orthostatic presyncope and skin pigmentation. The etiology behind Addison's disease ranges from autoimmune adrenalitis, tuberculosis and neoplasia - however, the majority of Addison's disease in the Western hemisphere is a result of autoimmune pathology. However, the path to identifying adrenal insufficiency is fraught with misdiagnoses due to its insidious natural history, and can potentially culminate in a life-threatening Addisonian crisis.

Case Presentation: We report the case of a 65 year old female presenting with hyponatremia in the context of worsening orthostatic syncope, generalized fatigue and malaise ongoing for 2 years. Her medical history was significant for multiple sclerosis and polymyalgia rheumatica, the latter of which was diagnosed a few months prior. At that time she presented with proximal myalgias and hyponatremia incorrectly attributed to SIADH. She was prescribed prednisone 20 mg daily, which abated her symptoms and corrected her hyponatremia. However, as her prednisone dose was tapered, her symptoms of lassitude and malaise recurred, which led to multiple emergency room visits.

Physical examination was remarkable for orthostatic hypotension and hyperpigmentation. She had a low serum sodium with an elevated urinary sodium, incongruent with a previous diagnosis of SIADH. This prompted us to interrogate adrenal function, which showed an undetectable cortisol level in response to a cosyntropin stimulation test. Her ACTH level was elevated at 55.7 pmol/L, confirming a diagnosis of primary adrenal insufficiency. Computed tomography of her abdomen revealed diminutive adrenals, suggesting destructive autoimmune adrenalitis as the likely cause of her adrenal insufficiency. She began prednisone and fludrocortisone supplementation, and experienced rapid resolution of both her symptoms and hyponatremia. At that point, she was discharged with internal medicine follow-up, and instructions regarding stress dosing her steroids during acute illness.

Discussion: This case illustrates the potential of misdiagnosis in Addison's disease, given its non-specific symptoms outside of an adrenal crisis. Primary adrenal insufficiency is an uncommon but critical diagnosis to consider when evaluating hyponatremia. In our patient’s case, the presence of concomitant autoimmune pathology necessitating steroids masked the symptoms of adrenal insufficiency temporarily, making diagnosis a challenge. Given the relative ease of glucocorticoid and mineralocorticoid replacement, and the severity of a potential Addisonian crisis, clinicians should bear a high index of suspicion for primary adrenal insufficiency.

References

Title: Let’s Not Be Rash...

Authors: Jessica J. Dreicer, MD; Stephanie A.C. Halvorson, MD

Introduction: Neutrophilic dermatoses are uncommon skin conditions associated with malignancy, rheumatologic, and autoimmune conditions. They manifest as sterile neutrophilic pustules and are often misdiagnosed as an infection.

Case Presentation: A 74-year old man with a history of myelodysplastic syndrome (MDS) and recent diagnosis of biopsy-proven giant cell arteritis (GCA) still on moderate-dose steroids presented with subacute, progressive skin and mucosal lesions. About one month prior the patient cut his left fourth finger and then attempted to remove a splinter. Subsequently his finger became dusky and painful. Around this same time he noticed a painful "marble" in his left axilla followed by formation of lesions on his left chest and right neck. He also stopped using his upper dentures because of pain. Finally, he developed pain and redness of his left eye which prompted him to present to an outside hospital. Despite empiric treatment with broad-spectrum antimicrobials and anticoagulation, the patient’s lesions worsened and he continued to have intermittent fevers. He was transferred to our hospital for further evaluation.

On arrival the patient was found to have erythema of the left eyelid and ulcero-necrotic skin lesions in his left axilla, left chest, right anterior cervical region and necrosis of his maxillary gums. His left fourth finger was necrotic. Biopsies of the chest lesions demonstrated “suppurative neutrophilic dermatitis with necrosis.” Atypical infections with sporotrichoid spread were considered given the distribution of skin involvement however repeat cultures and a universal polymerase chain reaction test for a wide range of infectious etiologies were all negative. Sweet’s syndrome was considered given the biopsy results however this seemed less likely since he was already receiving steroids. Due to his recent diagnosis of GCA, vasculitis was considered. Computed tomography (CT) angiography of the head and neck returned normal. With his history of MDS, a positron emission tomography scan and CT of the chest, abdomen, and pelvis were obtained to evaluate for malignancy. All were unremarkable.

Given his overall unrevealing work-up, his presentation was re-evaluated and a diagnosis of a rare form of neutrophilic dermatosis called pyoderma/pyostomatitis vegetans (PD-PSV) was rendered based on the classic finding of neutrophilic necrotic lesions involving both skin and mucosa. His steroids were increased to 1 mg/kg. PD-PSV is reported almost exclusively in patients with inflammatory bowel disease (IBD) so the patient underwent colonoscopy with biopsies which were negative for inflammation. At a follow-up visit five months later, the patient’s lesions were completely resolved.

Discussion: PD-PSV is a rare subtype of neutrophilic dermatosis distinguished by involvement of mucosa and flexural areas of the skin. The fact that the patient tested negative for IBD makes his presentation even more unusual. Sometimes PD-PSV can predate the manifestation of IBD. In this case, the underlying MDS was thought to be a predisposing factor for both PD-PSV and GCA.
References


Oregon-Clinical Vignette-Poster Finalist
Angela M Gibbs, MD

Title: Cocaine-induced Brugada Syndrome

Authors: Gibbs, Angela MD and Irene Hendrickson, MD, Providence St Vincent Internal Medicine Residency, Portland, Oregon

Introduction: Brugada syndrome was first described in 1992 based on eight case reports of patients with recurrent episodes of aborted sudden cardiac death. These patients shared an electrocardiographic (ECG) pattern of a pseudo-right bundle branch block, ST-segment elevation in V1, V2, and V3, with a normal QT interval. All 8 had no structural heart disease or other known risk factors for arrhythmias and all had sustained polymorphic ventricular tachycardia.

Case Presentation: A 42-year-old man with a history of marijuana use was brought to the hospital after a ventricular fibrillation cardiac arrest. He was defibrillated twice, with return of spontaneous circulation. Upon transfer to the intensive care unit he had six subsequent ventricular fibrillation arrests requiring defibrillation. His ECG showed a down-sloping ST elevation in V1 and V2 with a saddle-back appearance. Angiography revealed normal coronary arteries. His left ventricular function was mild reduced with an ejection fraction of 45% by echocardiogram. Treatment included therapeutic temperature management and lidocaine. His urine drug screen was positive for cocaine and cannabinoids. Upon achieving normothermia the patient awoke and had a complete neurologic recovery. He acknowledged using cocaine daily for several months. His echocardiogram and ECG normalized, and he was discharged home in good condition.

Discussion: Cocaine has several cardiac effects, most notably, its sympathomimetic effect which can cause tachycardia, hypertension, and coronary vasospasm. However, there is an additional mechanism that is implicated here, as cocaine is a direct sodium channel blocker similar to class IC antiarrhythmic drugs.

Brugada syndrome has been widely described since the few original case reports mentioned above. It is more common in males and usually occurs in patients age 22-65, with a peak in the 40s. Sodium channel defects in cardiac myocytes lead to the characteristic ECG pattern and clinical syndrome. A specific genetic mutation may be implicated in up to 25% of patients. The inherited form of the mutation has been seen more commonly in Asian populations.

Two different types of Brugada syndrome have since been described, with distinct ECG changes. Type 1 has a high take-off ST elevation and Type 2 has a saddle-back appearance. Numerous drugs have recently been reported to cause drug-induced Brugada including Class IA and IC antiarrhythmics, calcium channel blockers, beta blockers, nitrates, psychotropic drugs, and others.

My patient’s ECG changes are consistent with a type 2 Brugada syndrome. His ECG changes resolved after the cocaine had been metabolized, suggesting an inducible Brugada syndrome from the cocaine’s sodium channel blockade. While all internists should be familiar with the Brugada pattern on ECG and its associated Brugada syndrome that puts patients at risk for sudden cardiac death, we should also be aware that these same changes can be drug-induced.
Title: Antipsychotic-Induced Hypothermia

Introduction: Antipsychotic drugs (APDs) are often used in the treatment of acute delirium in the elderly. Hypothermia as an adverse drug reaction secondary to APDs poses a diagnostic challenge.

Case Presentation: An 86-year-old man with a past medical history notable for Alzheimer dementia and Bipolar disorder type 1 was admitted for acute delirium and psychosis. Prior to admission, the patient was unsuccessfully treated with lithium and risperidone. Upon admission, risperidone was discontinued and he was started on haloperidol for management of his psychosis. He remained agitated and was subsequently started on quetiapine, 12.5mg nightly.

Approximately 48 hours after initiation of quetiapine, he was noted to have asymptomatic hypothermia to 35.1°C (94.1°F,) which resolved without intervention. Three days later, his quetiapine dose was increased to 25mg nightly. This was followed again by hypothermia to 34.1°C (93.3°F) approximately 48 hours later. Extensive workup for infection and abnormal renal, adrenal, and thyroid function was unrevealing. ECG revealed worsened bradycardia, prolonged PR interval, and J-waves in the lateral leads. A diagnosis of antipsychotic-induced hypothermia was made. Quetiapine was stopped, haloperidol was continued, Bair Hugger therapy was initiated, and his temperature normalized over one week.

Discussion: Hypothermia is defined as a core temperature below 35°C (95°F.) Mild hypothermia ranges from 32 to 35°C (90 to 95°F,) moderate from 28 to 32°C (82 to 90°F,) and severe below 28ºC (82°F.) The differential diagnosis is broad, but the most common causes are exposure to cold, CNS failure (CNS trauma, Parkinson's disease, hypothalamic dysfunction,) endocrine failure (hypothyroidism, hypoglycemia,) infection (severe sepsis,) uremia, and medications/toxins [1.] Older adults are particularly at risk of developing hypothermia due to low physiological reserve, chronic co-morbid disease and medications that impair compensatory response [2.]

Thermoregulation represents the complex integration of chemical (serotonergic) and afferent axonal inputs in the pre-optic hypothalamus. Typical and atypical APDs achieve clinical antipsychotic effect through CNS modulation of dopaminergic (D1, D2) and serotonergic (5-HT-2a) receptors [1,2.] As a result, APDs may interfere with CNS thermoregulation, and atypical APDs may have a greater risk of hypothermia than typical antipsychotics [3,4,6.] Furthermore, APD-mediated inhibition of pathways which modulate physiologic compensation in the setting of hypothermia (alpha adrenergic) may potentiate decreased core temperatures [4,7.]

In elderly patients with concurrent dementia and agitation, hypothermia represents a clinically significant reaction to APDs, likely due to unintended interference with serotonergic and alpha-adrenergic thermoregulatory mechanisms. Dose-dependent hypothermia and ECG changes (bradycardia, prolonged PR interval, J-waves) are clues to this diagnosis [5.] Rapid withdrawal of the APD and active re-warming therapy after full diagnostic work-up Results: in resolution of hypothermia.

References

Title: When Percutaneous coronary intervention alert was called for infective endocarditis

Authors: Shaik Abdul Rashid, Shaik Abdul Samad, Gajanan Ganesh, Bhagatwala, Kunal, Munshi, Braun, Todd - Abington Hospital Jefferson Health, Abington PA

Introduction: Infective endocarditis (IE) is an obscure disease that manifests itself in various ways. Rarely do we encounter IE presenting as acute coronary syndrome (ACS) and even rarer is Lactobacillus (L) species causing IE. We present one such case of IE caused by L. paracasei presenting as ACS.

Case Presentation: A 51-year-old male with bicuspid aortic valve and essential hypertension was diagnosed with ST elevation myocardial infarction elsewhere and was transferred to our hospital for emergent catheterization. Auscultation revealed a 2/6 systolic murmur radiating to the axilla. Electrocardiogram showed ST-segment elevation in the inferolateral leads and troponin was elevated. Left heart catheterization showed 100% occlusion of distal left anterior descending artery. Thrombectomy was unsuccessful. Percutaneous transluminal coronary angioplasty was attempted but flow could not be restored. Patient spiked a fever overnight and blood cultures were drawn. Empiric antibiotics were started. The setting of fever, non-stentable coronary lesion and cardiac murmur was highly suspicious for IE causing coronary septic embolus. TEE showed multiple echo densities predominantly on the anterior mitral leaflet concerning for valvular vegetation, and paravalvular abscess. Blood cultures were positive for a slow growing “Streptococcus-like” organism. We started empiric vancomycin and gentamicin. Patient underwent aortic and mitral valve replacement. Extensive bivalvular infection and paravalvular abscess was noted and tissue samples were sent for culture. The slow growing Streptococcus was concerning for a nutritionally variant Streptococci like Abiotrophia and Granulicatella. Finally, on the eleventh day, we received the final identification of the gram-positive cocci as L. paracasei susceptible to penicillin and clindamycin. He was treated with a 6-week course of aqueous penicillin G with excellent response.

Discussion: ACS can be the presenting feature of IE. Embolic occlusion of coronary artery secondary to IE is very uncommon (reported in one study to be 2.9%, with a 64% mortality rate). Coronary occlusion in the setting of clinical features suspicious for IE should raise concern for a septic coronary embolism.

Lactobacillus causing IE is rare and L. paracasei is even rarer. Interestingly, upon further questioning, our patient revealed that he regularly consumed products containing Lactobacillus (Kombucha). Though there was no clear association, the source of Lactobacillus could have very likely been the probiotic drinks. The diagnosis and management was difficult in this case due to the unexpected organism which was resistant to empiric antibiotics.

We believe this is the first reported case of coronary septic embolus secondary to L. paracasei endocarditis. Lactobacillus as a cause of bacteremia should be kept in mind in a patient on probiotics. Also, the presence of paravalvular abscess in this case was surprising as Lactobacillus is not known to cause such a severe valvular disease.
Pennsylvania-Clinical Vignette-Poster Finalist
Taimoor Adnan, MBBS

Title: A Tumor Never Reported Before

Authors: Taimoor Adnan, MD, Kainat Saleem, MD, Kevin Kane, MD, UPMC McKeesport Hospital

Introduction: This clinical vignette presents the first reported case of a primary primitive liver epithelial cell neoplasm with hepatocellular and neuroendocrine components, which was highly aggressive and resistant to treatment.

Case Presentation: A 66 year old female presented to the primary care clinic with discomfort in the right upper quadrant of the abdomen, associated with weight loss. Physical examination was significant for a palpable liver several centimeters below the costal margin. CT scan of the abdomen and chest revealed multiple bilobar hepatic metastases and bilateral lung metastases. Needle biopsy of the liver mass revealed histological and immunocytochemical features most consistent with primary primitive hepatocellular neoplasms with divergent lines of differentiation of mostly hepatocellular and neuroendocrine features. AST, ALT and ALK were deranged. Aplha fetoprotein was 19872 ng/ml and chromogranin A was 58 ng/ml. Treatment was started with sandostatin and sorafenib, first line therapy for the individual components of this mixed type of tumor. Despite treatment, the patient continued to lose weight, felt more fatigued and alpha fetoprotein continued to rise, consistent with progression of tumor. Repeat CT scan showed interval progression of liver and lung metastases. Nivolumab was added at that point. Personalized cancer mutation panel was performed, which revealed no actionable mutation. Immunohistochemical studies for DNA mismatch repair proteins indicated that tumor is microsatellite stable and PDL-1 expression also came back negative. FOLFOX and Bevacizumab were also started later in the course of disease. Despite trying these treatment modalities, the patient’s condition deteriorated and she passed away within 6 months following the diagnosis of cancer.

Discussion: This is a very unfortunate woman diagnosed with primitive liver epithelial cell neoplasm with hepatocellular and neuroendocrine differentiations which has not been previously reported. The patient did not have risk factors for hepatocellular carcinoma, like Hepatitis B or C, NASH or hemochromatosis. Chemotherapy, immunotherapy, and biological agents all failed to induce a response. Current treatment modalities for both hepatocellular and neuroendocrine carcinoma were not effective, and an alternative approach may be necessary in the future for this type of cancer.
Title: Adult Subaortic Stenosis: An Unusual Etiology of Preventable Heart Failure

Introduction: Left ventricular outflow tract (LVOT) obstruction and elevated aortic valve (AV) velocities measured by echocardiography can be due to subaortic stenosis (SAS), aortic valvular stenosis, supravalvular aortic stenosis, hypertrophic cardiomyopathy or coarctation of aorta. The most common mechanism is AV stenosis, however it is important to consider other less common causes such as adult congenital heart disease in the differential diagnosis. Adult SAS is estimated to occur in approximately 6.5% of patients with adult congenital heart disease and is most commonly associated with other cardiac anomalies.

Case Presentation: A 69 yr old female with past medical history of hypertension initially presented five years ago with dyspnea and leg swelling. A transthoracic echocardiography (TTE) at that time showed hyperdynamic LV function with aortic valve velocities of 4.57m/s, mean gradient 46mmHG, and what appeared to be a normally opening aortic valve. Transesophageal echocardiography (TEE) was recommended but the patient declined. She represented five months ago with heart failure. TTE showed normal LV function, aortic valve velocities >4m/s, and mild aortic regurgitation. She presents a third time with heart failure and consented to TEE which showed a subaortic membrane and normally opening aortic valve. In addition, she had developed severely decreased left ventricular systolic function and moderate-severe aortic regurgitation (AR). She was referred for cardiac surgery and resection of a discrete subaortic membrane primarily located under the left and right aortic valve leaflets and adherent to the underside of the noncoronary leaflet limiting its mobility was undertaken. There was extension into the LVOT and anterior mitral leaflet. Release of fibrosis trigones was also performed. The AV appeared structurally normal and was therefore preserved. Repeat echocardiography showed improvement in LV function, decrease in aortic valve velocity to 2.8 m/s, and mild AR.

Discussion: Our patient had a fibrous subaortic membrane which caused outflow tract obstruction and eventually aortic insufficiency and rapid decline in LV function. Aortic regurgitation due to subaortic membrane is common but usually mild and nonprogressive. Damage to the aortic valve due to the subvalvular systolic jet appears to be the main cause of AR but as in our patient, a direct extension of subvalvular fibrous tissue into the aortic valve has been rarely described. Resection of the membrane leads to restoration of normal aortic valve geometry and improved the AR without valve replacement. Membrane resection is recommended for patients with subaortic stenosis and a peak gradient of 50 mm Hg or a mean gradient of 30 mm Hg on echocardiography. Valve replacement may be necessary depending on the amount and etiology of the AR.

It is important to consider subaortic stenosis in the differential of elevated aortic outflow velocity and prompt early diagnosis and treatment as per guidelines to prevent heart failure.
Title: Stiff person Syndrome Masquerading As Psychosomatic Illness

Authors: Reema Andrade MD, Justin Chacko DO, Bojana Milekic MD

Introduction: Stiff person syndrome (SPS) is a rare disease with progressive, disabling axial and truncal rigidity causing painful spasms, postural deformities, and ambulatory impairment. SPS is due to neural inhibitory loss causing increased muscle activity. Reduced inhibition of the central nervous system occurs from the blockade of glutamic acid decarboxylase (GAD), an enzyme crucial to inhibitory pathways, as well as a decline in the levels of Gamma-Aminobutyric Acid (GABA). Estimated prevalence is one to two cases per million, predominantly in 30 to 50 year old women. Clinical presentation of the disease can be vague which can make diagnosis challenging.

Case Presentation: A 34-year-old Hispanic female with a history of generalized seizure disorder presented to the emergency department with complaints of back and leg stiffness. Medical records revealed frequent ER visits and prolonged hospitalizations at other institutions for similar complaints over the last six months. She was admitted in the past with severe muscle spasm, back and leg stiffness and recurrent falls. During the previous admissions she was also found to have tachycardia and high temperature. Differential diagnosis of conversion disorder and reflex sympathetic disorder was noted by several neurologists, epileptologists and psychiatrists during prior encounters. Her seizure disorder was without an episode in two years on Phenytoin and Levetiracetam. She was hemodynamically stable, afebrile and examination was remarkable except for stiffness in bilateral lower extremities and inability to bend at the waist with normal strength, sensation and reflexes. She was unable to sit up in bed and when attempted would fall backwards. Laboratory studies showed an abnormal CK (377 unit/L) but were otherwise normal (CBC, CMP, TSH, ESR, CRP). Review of external records noted unremarkable noncontrast imaging of the brain and electroencephalography (EEG). Repeat MRI of the brain along with MRI of the thoracic and lumbar spine showed no acute process. Labs for anti-GAD 65 antibody were strongly positive with titers > 250 IU/ml (normal < 5 IU/ml). She was subsequently started on Diazepam 5mg three times a day. There was a significant improvement in the stiffness and the patient was able to walk with help and discharged home for outpatient neurology follow-up.

Discussion: SPS is a rare disease which is often overlooked or misdiagnosed due to its variable presentation. High titers of anti-GAD 65 antibody along with characteristic muscular rigidity and truncal spasticity are required to diagnose stiff person syndrome. It should be considered in any patient with unexplained stiffness because early diagnosis and treatment can prevent debilitation. Barbiturates and benzodiazepines remain the treatment of choice due to their GABAergic properties.

References
Title: Rapid development of Severe Aortic Regurgitation from *Streptococcus pyogenes* infection: An Atypical Culprit

Authors: Timothy Gaw DO, Akanksha Agrawal MD, Toni Anne de Venecia MD

Introduction: Group A Streptococcal endocarditis is a rare form of endocarditis most commonly reported in intravenous drug users and in children following varicella infection. We report a case of infective endocarditis due to *Streptococcus pyogenes* with development of severe valvular regurgitation within 24 hours.

Case Presentation: A 50-year-old African American female with no known prior medical history presented with two weeks of subjective fever and lower back pain. She returned from Mali two months back, and denied any sick contacts or intravenous drug use. Her vitals were remarkable for low grade fever and tachycardia to 120 beats per minute. Initial lab work was significant for leukocytosis of 13,600/ mcL, lactic acidosis and kidney dysfunction with blood urea nitrogen of 60 mg/dL and creatinine 5.8 mg/dL. Admitting blood cultures grew *Streptococcus pyogenes* in two sets of anaerobic and aerobic bottles. She was started on intravenous ceftriaxone. Transthoracic echocardiogram showed concern for a mobile mass at the aortic root with trace aortic regurgitation (AR) and a left ventricular ejection fraction (LVEF) of 50%. A transesophageal echocardiogram was performed to evaluate the mass in aortic root, which interestingly showed severe AR with prolapse of left cusp, without any perforation or vegetation. With the new acute valvular regurgitation and positive blood cultures, patient was diagnosed with infective endocarditis due to *S. pyogenes* per Modified Duke Criteria. Patient then became symptomatic with mild shortness of breath and worsening tachycardia. Cardiothoracic surgery was consulted for evaluation for replacement of the aortic valve. Clindamycin was added to dampen the necrotizing effect of *S. pyogenes* in addition to intravenous ceftriaxone. On day 7, the patient was noted to have a tender, open wound between the right 4th and 5th toe with purulent drainage. Wound culture from the cloudy drainage grew *Streptococcus pyogenes* which was then believed to be the source of the bacteremia. Her blood cultures after the initial 2 sets remained negative, creatinine improved and she was discharged on intravenous antibiotics for outpatient aortic valve replacement.

Discussion: This case illustrates that *Streptococcus pyogenes*, though an atypical cause of IE, can cause rapid destruction of the valve leaflets. Our case also demonstrates the importance of daily thorough physical examinations in providing clues towards the source for the infection.
Title: Superior and Inferior Ophthalmic Vein Thrombosis in the setting of Lung Cancer

Authors: Nawal Habib MD, Kimberly Lessard DO

Introduction: Superior Ophthalmic Vein Thrombosis is a rare diagnosis that is generally associated with infectious conditions and hypercoagulable states related to cancer and/or autoimmune conditions. Although there are limited management guidelines available for this medical emergency, prompt diagnosis and treatment are essential to prevent permanent blindness. We present a case of superior ophthalmic vein thrombosis with left cavernous sinus thrombosis in the setting of newly diagnosed malignancy.

Case Presentation: An 82-year-old female with past medical history of uncontrolled hypertension, dementia and tobacco abuse presented with complaints of blurry vision and frequent falls for the past few weeks. She also noted recent unintentional weight loss and productive cough. She was afebrile, cachectic, had decreased left-sided breath sounds, mild left-sided proptosis, chemosis and bilateral cataracts on admission. Visual acuity was R 20/25, L hand motion, with normal pupillary function and extraocular movements. Lab Results: and initial CT Head were within normal limits. Chest XR and CT alike showed left-sided atelectasis with ipsilateral mediastinal shift due to mucus plugging, a moderate left pleural effusion and multiple spiculated cavitating nodules in the right lobe concerning for malignancy. Two days following admission, she developed acute left orbital pain and visual loss prompting a brain MRI. MRI revealed enlargement of the L superior and inferior ophthalmic veins without contrast enhancement concerning for thrombosis. Following initiation of heparin infusion, a CT Venogram confirmed the presence of thrombosis along with a filling defect in the L cavernous sinus.

In the absence of signs, symptoms or risk factors for underlying infection, malignancy remained the primary differential and antibiotics were not initiated. Unfortunately, several barriers to confirmation of malignancy arose. Bronchoscopy was performed which, despite removal of a mucus plug, resulted in minimal improvement in the atelectasis. Pathology from brush cytology and trans-bronchial biopsy were inconclusive. A transthoracic lung biopsy of right lung nodules was deemed high risk given persistent atelectasis of the left lung and presence of underlying emphysema. A thoracentesis of the left effusion was performed, however, cytology was negative for malignancy. Given the negative autoimmune and relevant hematologic work up for hypercoagulability and the high likelihood of underlying lung cancer she was started on Lovenox with subsequent improvement in visual symptoms over the next few weeks. Serial chest CT will be required to monitor for progression.

Discussion: Risk factors for superior ophthalmic vein thrombosis are multifactorial and include infections, autoimmune disorders, and malignancy. Clinical features include abrupt onset of painful proptosis, chemosis, ophthalmoplegia and diminished visual acuity. Urgent MRV/CTV are required to confirm the diagnosis and exclude cavernous sinus thrombosis. The severity of symptoms and underlying systemic condition guide prompt management to prevent permanent blindness. The use of anticoagulation for malignancy associated cases is to be determined after risk-benefit analysis.
Pennsylvania-Clinical Vignette-Poster Finalist
Stephanie Hart, AHP MBBCH

Title: Unilateral pulmonary edema: An unusual manifestation of systolic heart failure

Authors: Stephanie Hart, MD, Lankenau Medical Center Internal Medicine Residency Program, Niraj Patel, MD, Lankenau Medical Center

Introduction: Unilateral pulmonary edema is a rare finding and represents a difficult clinical scenario as it can be mistaken for other causes of unilateral infiltrates on chest imaging. Below we present a case of unilateral pulmonary edema in a young adult male as the first manifestation of systolic heart failure.

Case Presentation: A 37-year-old male with a history of paroxysmal atrial fibrillation presented to the hospital with a cough productive of pink, frothy sputum, dyspnea on exertion, and central chest tightness that began abruptly three hours prior to admission. He endorsed increased dietary salt intake. In the emergency department, his blood pressure was 221/102 mmHg and oxygen saturation was 88% on room air. Other vital signs were normal. Examination was notable for obesity and bibasilar coarse crackles, worse on the right. He had no jugular venous distention or ankle edema. Labs were unremarkable with the exception of B-type natriuretic peptide of 249 pg/ml. A chest x-ray and confirmatory CT revealed cardiomegaly and extensive scattered patchy groundglass/vaguely nodular opacities throughout the right lung with minimal findings on the left, concerning for an atypical infectious or inflammatory process. He was initially managed with intravenous nitroglycerin, furosemide, enalapril, ceftriaxone, and azithromycin, as well as supplemental oxygen, and his symptoms rapidly improved. Treatment for community-acquired pneumonia was continued overnight. Tests for Legionella and Mycoplasma were negative. Pulmonology was consulted and believed his presentation was consistent with atypical pulmonary edema rather than an infectious process, precipitated by uncontrolled hypertension and excessive salt intake. A transthoracic echocardiogram was performed, which confirmed mild left ventricular systolic dysfunction. He had no evidence of diastolic dysfunction or valvular pathology. Antibiotics were discontinued and he received one additional day of intravenous diuresis prior to being discharged on a thiazide and calcium-channel blocker. An ApneaLink performed during his hospitalization confirmed an apnea-hypopnea index of 24. He was discharged with instructions for a repeat CT in six weeks to ensure resolution of the abnormal lung findings as well as an outpatient sleep study to confirm sleep apnea.

Discussion: Unilateral pulmonary edema is a rare entity, usually right upper lobe predominant and associated with severe mitral regurgitation. It has also been seen in individuals with pre-existing pulmonary disease. Review of the literature reveals mostly case reports of this unusual condition. This case illustrates unilateral pulmonary edema as the initial manifestation of systolic dysfunction in a young adult male with no known history of heart failure. Interestingly, he had no evidence of mitral valve disease on echocardiogram. Unilateral pulmonary edema conveys a higher mortality risk compared with bilateral pulmonary edema, possibly related to the delay in adequate treatment due to diagnostic uncertainty. Prompt diagnosis is important in patients presenting with unilateral pulmonary infiltrates to ensure appropriate and timely management.

References

Pennsylvania-Clinical Vignette-Poster Finalist
Jana Havranova, MD

Title: A rare case of extreme immune activation

Authors: Jana Havranova - Internal medicine, St.Lukes University Hospital, Bethlehem PA; Renee Tehrani - Internal medicine, St.Lukes University Hospital, Bethlehem PA; Thong Le - Infectious disease, St.Lukes University Hospital, Bethlehem PA

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is an uncommon hematologic disorder. Although more common in children under the age of one, it can be seen in older children and adults 30% of the time. In this life threatening condition, there is severe systemic hyperinflammation caused by aberrant T lymphocyte function, leading to “cytokine storm”. Proposed mechanisms of immune activation include primary genetic syndromes, underlying malignancies especially lymphoid, autoimmune conditions, bacterial infections and viral infections, especially EBV, CMV, HIV and HCV. Patients with HLH usually present initially with nonspecific symptoms and findings of fever, encephalopathy, hepatosplenomegaly, pancytopenia and hypertriglyceridemia. Highly elevated ferritin is a both sensitive and specific marker. Hemophagocytosis is seen on bone marrow biopsy. Lack of familiarity, nonspecific presentation and high mortality make HLH a diagnostic challenge.

Case Presentation: A 54-year-old male with PMH of ankylosing spondylitis presented with fever, rigors, myalgias and severe sore throat for 1 week. On examination, he had pharyngeal exudates, pharyngeal and tonsillar necrosis, petechial rash on his abdomen and bilateral lower legs and inguinal lymphadenopathy. Initial laboratory studies revealed AKI, elevated CK, thrombocytopenia, leukopenia and hypofibrinogenemia. He was initially treated for severe pharyngitis with IV ampicillin/sulbactam. Due to lack of response after 48 hours, further work up with chest, abdomen and pelvis CT revealed diffuse lymphadenopathy. At this point, the diagnosis of HLH was considered. Ferritin level was elevated at 27,564 ng/ml. Bone marrow biopsy confirmed the diagnosis showing a hypercellular marrow and increased histiocytes with the presence of hemophagocytosis. There were no blasts, granulomas or metastatic malignancy. Extensive infectious workup was largely negative with the exception of positive EBV IgG with negative IgM. Patient was felt to have chronic active EBV mononucleosis versus underlying lymphoid malignancy as precipitating factor for HLH. While awaiting transfer to a regional tertiary care hospital, therapy with IV dexamethasone was initiated. At the tertiary care hospital, patient was treated with chemotherapy. Unfortunately, patient deteriorated. He developed worsening encephalopathy and acute respiratory failure requiring mechanical ventilation. He subsequently developed fungemia and despite antifungal treatment, expired 3 weeks after symptom onset.

Discussion: This case illustrates the importance of early recognition and treatment of HLH. This diagnosis should be considered in critically ill febrile adults with multisystem involvement. A serum ferritin level should be obtained early and if highly elevated, is suggestive. HLH is a rare life-threatening disorder of unregulated immune activation resulting in extreme systemic inflammation and requires prompt recognition and early introduction of appropriate therapy.
Pennsylvania-Clinical Vignette-Poster Finalist
CPT Kelly M Kalovcak, USA

Title: A Case of Vedolizumab-associated Hypophysitis with Panhypopituitarism and Diabetes Insipidus

Authors: Kelly Marie Kalovcak, MD; Jodie A. Reider, MD; John W. Kennedy, MD - Geisinger Endocrinology Division, Danville, PA

Introduction: Improved understanding of immune dysregulation in autoimmune diseases has led to development of monoclonal antibody based therapies, which target the aberrant immune response. While significant benefits are seen with these therapies, they may increase the risk of infection and new autoimmune conditions by blocking pathways that protect the host. Immune-related adverse events involving the endocrine system have been reported, including hypophysitis, thyroid dysfunction and adrenal insufficiency. These events have been reported most commonly in association with immune checkpoint inhibitors involving cytotoxic T-lymphocyte antigen-4 (CTLA-4) and programmed cell death-1 (PD-1) receptors. Vedolizumab is an anti-integrin α4β7 antibody with gut-specific immunosuppressive effects, approved for use in patients with inflammatory bowel disease. Here we report the first case in the literature of a life-threatening presentation of vedolizumab-associated hypophysitis with panhypopituitarism and diabetes insipidus (DI).

Case Presentation: A 57-year old male with ulcerative colitis presented with extreme fatigue, polyuria, polydipsia, anorexia and 40-pound weight loss. Symptoms began 6 months following initiation of therapy with vedolizumab. Initial laboratory studies showed cortisol <0.1 µg/dL and TSH 0.02 ng/dL. He was admitted for further evaluation. Pituitary hormone analysis showed panhypopituitarism (ACTH <5.0 pg/mL, FSH 0.6 mIU/mL, LH <0.1 mIU/mL, total testosterone <2.5 ng/dL, PRL 24.3 pg/mL). Sella MRI revealed an enlarged pituitary gland with displacement and elevation of the optic chiasm, diffuse heterogeneous contrast enhancement, and markedly thickened stalk, suggestive of hypophysitis.

Suspected hypophysitis prompted treatment with high-dose steroids. Thyroid hormone and testosterone replacement were later prescribed. Shortly following initiation of steroids, he developed hypernatremia and inappropriately dilute polyuria, consistent with DI, likely unmasked by steroid treatment. This was treated successfully with desmopressin (DDAVP).

He was seen shortly after hospital discharge and noted significant symptomatic improvement. He is scheduled for repeat sellar MRI in 1 month to determine the pituitary response to treatment with steroids.

Discussion: The development of hypophysitis occurred in temporal association with initiation of therapy with vedolizumab in this patient, suggesting a causative effect. It is also possible that the presence of another autoimmune disease, ulcerative colitis, predisposed this patient to hypophysitis. The development of hypophysitis in association with monoclonal antibodies has been well described, particularly with ipilimumab and nivolumab, which target CTLA-4 and PD-1 T-cell receptors respectively. This is the first reported case to describe hypophysitis in association with vedolizumab, an anti-integrin α4β7 antibody. Since vedolizumab’s primary mechanism of action is felt involve gut-specific immunosuppression, this case should prompt further research into a possible mechanism for immune-related adverse effects on the pituitary gland. It also highlights the importance of increasing provider awareness of immune-related adverse events that may occur in patients treated with monoclonal antibodies. Patients should be screened for symptoms of endocrine dysfunction with hormonal testing performed if indicated.
Pennsylvania-Clinical Vignette-Poster Finalist
Venkata Sowjanya Kanakadandi, MBBS

Title: Bubbles in Liver-Clostridium Perfringens Abscess

Authors: Sowjanya Kanakadandi, Simran Dhillon, Angela Lucero, Naveen Parva

Introduction: Pyogenic liver abscess from clostridium perfringens is a rare occurrence. Risk factors for clostridium septicemia include being elderly, having poorly controlled diabetes mellitus, and having concomitant or history of cirrhosis and malignancy, especially gastrointestinal and genitourinary malignancies. Clostridium is a normal inhabitant of the human bowel and a common cause of food poisoning leading to tissue necrosis and gas gangrene. Only few cases have been reported so far. However, our case is peculiar in that the patient did not have any known malignancy and had been treated with antibiotics prior to diagnosis of the abscess.

Case Presentation: A 88-year-old female with past medical history of coronary artery disease, hypertension, hyperlipidemia, uncontrolled diabetes, hypothyroidism who had flu-like symptoms about a week ago presented with loss of appetite. The following vitals were measured: blood pressure 70/50 mmhg, temperature of 39.1 F, pulse rate of 130 beats per minute, respiratory rate of 30 beats per minute. She was confused and had a tender abdomen. Laboratory studies indicated white blood cell count 16.60/microliter, and lactic acid 9.2 mmol/L. Computerized tomography scan of the abdomen revealed a round, gas-containing collection within the right hepatic lobe measuring 4.6 x 4.3 x 4.7 cm, consistent with hepatic abscess. The patient was started on aztreonam, metronidazole, and vancomycin, and hepatic drain was placed using ultrasound guidance. Abscess cultures revealed polymicrobial infection with clostridium perfringens, bacteroides fragilis, streptococcus viridans, klebsiella oxytoca, and enterococcus. She received six weeks of IV vancomycin, IV ceftriaxone and PO metronidazole. On follow up, the hepatic abscess decreased in size to 1.2 x 1.1 x 1.3 cm. The patient was transitioned to PO antibiotics and will remain on antibiotics until complete resolution of the abscess.

Discussion:

In summary, liver abscess due to C. perfringens is a rare and often fatal event. A high degree of suspicion for the diagnosis and immediate treatment with drainage and antibiotics are required to avoid catastrophic consequences. This type of abscess is commonly seen in patients with immunosuppression or malignancy. However, since our patient did not have either, the question arises as to whether the increased mortality in such hepatic abscess is likely from the infection itself or the underlying etiology related to immunosuppression.
Title: Anti-coagulating a Supratherapeutic PTT: A cirrhotic story

Authors: Latorre, Johan, Pizzola, Christopher, and Beck, Alyssa.

Introduction: Anti-coagulation in a cirrhotic patient is practically impossible. Gone are the days where we thought their elevated INRs provided protection against thromboembolic events. We now know that in fact, the opposite is true. There are no guidelines for which to follow as we do not have a good way to monitor whether or not the patient is anticoagulated\textsuperscript{1,2}.

Case Presentation: Mr. DS was a 51 year old gentleman with no past medical history when he suddenly developed abdominal pain and nausea. The pain gradually worsened over a month before he went to his primary care physician. Labs revealed elevated LFTs and an abdominal ultrasound showed splenomegaly, ascites, and cirrhosis. Abdominal CT confirmed the ultrasound findings in addition to showing a thrombosis in the SMV, splenic vein, proximal portal vein, and varices, suggestive of Budd Chiari Syndrome (BCS). He was immediately transferred to a Tertiary center. Hepatology, Vascular and General Surgery were consulted, but no interventions were performed because a repeat MRI showed extensive progression into the hepatic veins. Hematology was consulted because cbc showed an elevation in all cell lines. A hypercoagulable work-up was initiated. Our team ordered an EPO level, and continued the heparin drip for anti-coagulation in case of the need for urgent intervention, and planned for a bone marrow biopsy. EPO was high normal, and JAK2 assay was positive for Polycythemia Vera, and confirmed with a bone marrow biopsy. We then began to question whether he was truly anti-coagulated because his PTT was heavily elevated from his cirrhosis, which was used to titrate the heparin drip. An anti-Xa level was 0.13, indicating he was sub-therapeutic. We discussed the use of Apixiban with Hepatology for long-term anti-coagulation, but unfortunately, the patient decided to stop all treatment and proceed with comfort measures.

Discussion: The current recommendations for anticoagulation in a patient with cirrhosis is an INR goal of 2.0-3.0, but with the elevated INR due to the decrease synthesis of most clotting factors, it becomes difficult to assess whether anticoagulation is therapeutic\textsuperscript{1,2}. Low Molecular Weight Heparin (LMWH) is preferred over unfractionated heparin since bleeding risk is less in LMWH\textsuperscript{2}. Unfortunately, compliance is an issue as patients do not like to inject themselves subcutaneously\textsuperscript{3}. Secondly, Anti-Xa assays cannot reliably be used for monitoring cirrhotic patients as they have low anti-thrombin levels, though is recommend for individuals with prolonged PTT at baseline with levels between 0.5 – 0.8\textsuperscript{3-5}. Kunk et al. (2016) showed that compared to historic controls, the incidence of major bleeding decreased and clot resolution increased with use of direct oral anticoagulants (DOACs). DOACs are a promising treatment as there is no need for monitoring, but unfortunately more data is needed to determine their efficacy in these circumstances\textsuperscript{6-8}.

References


Pennsylvania-Clinical Vignette-Poster Finalist
Daniel Lefler, MD

Title: Try, Try Again: A Case of Non-Resectable Rosai-Dorfman Disease

Authors: Daniel Lefler MD, Andrew Orr MD

Hospital of the University of Pennsylvania

Introduction: Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive lymphadenopathy, is a rare, nonmalignant disorder of histiocyte proliferation. There have only been approximately 600 reported cases. Diagnosis of RDD can also be logistically difficult, and treatment modalities are limited.

Case Presentation: A 41-year-old woman presented to care with lower extremity edema and progressive dyspnea on exertion. She was found to have right heart failure secondary to left pulmonary artery occlusion and severe narrowing of the right pulmonary artery by an amorphous superior mediastinal mass. A PET scan showed marked fluorodeoxyglucose (FDG) avidity of the mass, and she underwent intravascular biopsy to determine its etiology. At that time, the radiologic appearance was concerning for angiosarcoma. However, histology revealed only thrombus material and a minute fragment of unremarkable intima, and she was then lost to follow-up until re-presenting three months later for worsening symptoms. She was admitted to the hospital for expedited workup and underwent two further biopsies: one via bronchoscopy and another by repeat intravascular approach. Unfortunately, histology from these biopsies was again non-diagnostic, revealing only fibrous tissue with a mixed inflammatory infiltrate and vessel wall fragments with myxoid degeneration, respectively. Additionally, the cardiothoracic surgery team did not believe her mass was accessible from an anterior approach. Given her progressive clinical decline, chemotherapy was considered to treat for angiosarcoma, as this was still the leading clinical diagnosis. In the meantime, she underwent stenting of the right pulmonary artery to relieve the pressure on her right heart, with almost immediate improvement of symptoms. During the procedure a fourth biopsy was performed, and histology revealed fibrous tissue with a lymphohistiocytic and plasma cell infiltrate. There was positive staining for S100 and CD68, negative staining for CD1a, emperiplois, and molecular testing for a BRAF V600E mutation was negative. These findings were consistent with a diagnosis of Rosai-Dorfman disease (RDD). Though surgical management is standard of care, she was considered to be a high-risk surgical candidate due to her comorbidities and extent of disease, and the patient and medical team opted instead to treat with involved-field radiation. Six weeks after beginning radiation therapy, a follow-up CT showed a decrease in the infiltrative mediastinal mass reflecting partial therapeutic response, a widely patent pulmonary artery, and interval improvement of right atrial and ventricular enlargement. Six months later, a repeat CT was stable.

Discussion: This case presents only the fifth reported case of RDD involving the pulmonary arteries. It also demonstrates the difficulty in the diagnosis of RDD, and should provide a warning to clinicians about planning treatment without a tissue diagnosis. Finally, though surgical therapy is preferred for RDD, radiation can provide at least an initial benefit. Further evaluation will be needed to ensure this response is durable.

References

Pennsylvania-Clinical Vignette-Poster Finalist
Kyle Macaulay, DO

Title: A case of lupus in the elderly- a diagnostic dilemma.

Authors: Kyle Macaulay, DO1; Sijan Basnet, MD1; Pragya Shrestha, MD1; Oluwaseun Shogbesan, MD1; Justin Johannesen, MD1

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Introduction: Systemic lupus erythematosus (SLE) is an autoimmune disorder with diverse manifestation but presents commonly with fever, fatigue, malar rash, weight changes and arthralgia. Predominantly affecting women of child bearing age however about 15 % of cases develop later in life. We present a case of late onset SLE diagnosed in an elderly female with obscure clinical presentation and life-threatening complications.

Case Presentation: An 81-year-old woman with longstanding history of pancytopenia presented to the emergency department with several months of progressive nausea, fatigue, and dizziness. She was afebrile with blood pressure 102/51 mm Hg and heart rate of 55 beats per minute. She denied photosensitivity, rash, fever, chills, mucositis, chest pain, abdominal pain, joint pain or joint swelling. There was no known family history of connective tissue disorders. Physical exam was benign except for faint malar rash on the face. Labs were remarkable for WBC 3,000/µl, hemoglobin 7.8 g/dl and platelets 63,000/µL. Liver and renal function tests were within normal limits with normal urinalysis. Bone marrow biopsy for pancytopenia in the past revealed mildly hypocellular marrow without evidence of lymphoproliferative disorder, and flow cytometry was unremarkable. Chest X-ray showed slight cardiomegaly with left basilar opacity and a small left pleural effusion. Computed tomography revealed moderate pericardial effusion. Transthoracic echocardiogram confirmed pericardial effusion with evidence of early tamponade physiology. Patient underwent video-assisted thoracoscopy surgery with left pericardial window formation and chest tube placement. Pericardial fluid analysis was remarkable for mildly elevated lymphocytes, normal glucose and LDH without malignant cells. Autoimmune workup revealed positive ANA at 1:160 titer, otherwise normal C3/C4/total complement levels, and negative RF and anti-CCP antibodies. Anti-histone and single stranded DNA antibodies were positive with negative antibodies for Smith, centromere, RNP, double stranded (ds) DNA, Ro and La antigens. Possibility of drug induced lupus erythematosus was considered with patient’s history of hydralazine use for 7 months, however this was considered less likely as hydralazine was stopped one month prior to presentation of her current symptoms and pancytopenia predating use of medication. Patient was diagnosed with SLE based on SLICC criteria (≥4/11 criteria- thrombocytopenia, leucopenia, pleuro-pericardial effusion, positive ANA, malar rash). Hydroxychloroquine was initiated after which patient had improvement of her symptoms and is planned for outpatient follow up.

Discussion: Diagnosis of elderly onset lupus is challenging due to its insidious onset, atypical presentation and similarity to more common disorders in the elderly. Clinical and serological features may differ from lupus in younger individuals, as arthritis, fever, serositis, and lung disease are more common than skin and renal involvement. Most patients will be ANA positive, but the prevalence of anti-ds DNA and hypocomplementemia is lower.

References


Pennsylvania-Clinical Vignette-Poster Finalist
Roman Marchenko, MD

Title: A rare case of hyperkalemia mimicking ST-elevation myocardial infarction

Authors: Marchenko R, Nazir S, Diaz Fraga JN

Introduction: Profound hyperkalemia may be associated with a variety of well-known and described in literature EKG manifestations including tall peaked T-waves, PR-interval and QRS prolongation, disappearance of P-waves, sine wave pattern, and literally almost any type of dysrhythmia. ST-segment elevation is an extremely rare and unusual manifestation of hyperkalemia.

Case Presentation: A 56-year-old male with a past medical history of end stage renal disease on hemodialysis presented to the hospital with generalized weakness and fatigue. Patient was noted to be very diaphoretic at presentation. Stat EKG showed a 7 mm horizontal ST-segment elevation in leads V1-V2 and tall peaked T-waves. STEMI alert was activated. Initial labs were remarkable for hyperkalemia to 8.7 mEq/L but negative troponin. Due to lack of anginal symptoms patient did not undergo emergent cardiac catheterization and a stat bedside transthoracic echocardiogram was performed. The study showed normal left ventricular systolic function with no wall motion abnormality. EKG changes were attributed to extreme hyperkalemia. Patient was appropriately treated with IV calcium, bicarbonate, insulin and glucose, as well as emergent hemodialysis after which his potassium level normalized and EKG changes resolved. Further troponin trend did not reveal any myocardial damage. Patient was discharged from the hospital in stable medical condition 2 days later.

Discussion: Mechanisms of ST-segment elevation in hyperkalemia are poorly understood and might be related to high level of potassium affecting action potential during repolarization. Case reports describing this phenomenon are scarce. Clinicians should be aware of such an unusual EKG manifestation of a fairly common electrolyte abnormality since the delay in appropriate therapy aimed at correction of hyperkalemia may lead to life threatening complications.
Pennsylvania-Clinical Vignette-Poster Finalist
Ana Martinez-Tapia, MD

Title: Eructation as the Main Presentation in a Patient with a Gastrointestinal Stromal Tumor

Authors: Ana Laura Martinez-Tapia, M.D, Fabio Dorville, M.D, Neeraj Khiyani associated with St. Luke's University Health Network

Introduction: Gastrointestinal stromal tumors (GISTs) are tumors arising from subepithelial cells throughout the gastrointestinal tract. Approximately 60-70% of GISTs are found in the stomach. Clinical presentation for GISTs are usually nonspecific symptoms such as bloating, abdominal mass or abdominal pain. However, larger GISTs tend to ulcerate and bleed once they have grown a certain size and can present with obstructive symptoms.

Case Presentation: A 50-year old man with no significant past medical history presented to his primary care physician for increased eructation after meals for one month in duration. Physical examination was unremarkable. Abdominal ultrasound showed a solid mass below the left hepatic lobe potentially arising from the stomach. CT abdomen and pelvis with contrast showed a 9.5 cm exophytic mass arising from the inferior body of the stomach with mass effect on adjacent viscera. No lymphadenopathy or distant metastasis was seen. Further endoscopy evaluation showed indeterminate submucosal fullness on greater curvature of stomach. Gastric biopsy of the lesion was negative for Helicobacter pylori and histologic malignancy. A linear endoscopic ultrasound with fine needle aspiration was performed and this revealed a lesion arising from the serosa with focal involvement into the muscularis propria. Biopsy revealed positive markers for CD117, DOG-1, and S100 that were conclusive for a GIST. Patient underwent exploratory laparotomy with partial gastrectomy and excision of the tumor. No adjuvant chemotherapy was given due to negative tumor margins and low risk for recurrence.

Discussion: GISTs arise from the Interstitial Cells of Cajal (ICC), which are the pacemakers of the gastrointestinal system. These cells are responsible for the peristaltic movement within the stomach. Other functions of these cells, such as mechanosensation of the stomach have been less frequently described. During belching, accumulation of air in the stomach increases its volume and activates receptors within the gastric wall. This in turn relaxes the lower esophageal sphincter and Results: in eructation. Certain gastrointestinal disorders, such as functional dyspepsia and gastrointestinal esophageal reflux disease have been described relating with dysfunction of ICC. Whether gastric belching is related to ICC disorders has not been clinically defined. Our patient presented with increased eructation about a month in duration with no other cause found except a GIST.
Pennsylvania-Clinical Vignette-Poster Finalist
Don Mathew

Title: Value of Lung Ultrasound in Differentiating Between a Giant bulla and a Pneumothorax

Authors: Don Mathew, M.D, John Tso , M.D, Ibrahim Ghobrial , M.D , UPMC McKeensport

Introduction: Bedside ultrasound is gaining widespread clinical application in the emergency and intensive care settings. Its portability and availability has made it a very useful supplement to clinical examination. Here, we present a case where bedside lung ultrasound helped to accurately differentiate a bulla from a pneumothorax.

Case Presentation: A 79-year-old man with history of bullous emphysema presented with unsteadiness of gait and intermittent change of sensorium. During our clinical encounter, he was alert, oriented to time, place and persons. He denied shortness of breath or chest pain. He was found to be hemodynamically stable with adequate oxygen saturation of 92% on room air. Physical examination was remarkable for diminished breath sounds bilaterally, especially on the left side. Neurological examination showed ataxia but with no other focal motor or sensory deficits. Chest X-ray showed new left basilar pneumothorax of 20-25%, which was confirmed by chest CT, reporting the size of the pneumothorax to be at least 50%. Both reports were produced by licensed radiologists. However, due to the discrepancy between the size of the reported pneumothorax and hemodynamic status, bedside ultrasound was performed which showed a positive sliding sign indicating the presence of lung tissue thereby ruling out pneumothorax.

His neurological symptoms were found to be related to glioblastoma multiforme in the splenium of corpus callosum.

Discussion: Chest CT is the most accurate imaging for the detection of pneumothorax and is used as the reference standard. The sensitivity and specificity of lung ultrasound in detecting pneumothorax ranges from 86 to 98% and 97 to 100% respectively. Our case is unique because the lung ultrasound was more accurate than the chest CT in ruling out pneumothorax. Pneumothorax is identified on lung ultrasound by the absence of lung sliding on 2-D view with the presence of a lung point. Placing a chest tube in a patient with a giant bulla can result in pneumothorax, hemothorax, broncho-pleural fistula, hemorrhagic shock, or death.
Pennsylvania-Clinical Vignette-Poster Finalist
Hassan Mehmood, MD

Title: Opana ER (oxymorphone) Induced Thrombotic Microangiopathy- An Atypical Presentation in a patient with Hepatitis-C

Authors: Hassan Mehmood, MD, Asghar Marwat, MD, Ali Hussain, MD, Muzammil Khan, MD, Medha Joshi, MD, Varun Malhotra, MD

Introduction: Oxymorphone is a semi synthetic extended release opiate used to treat moderate to severe chronic pain. The Food and Drug administration approved oral form of oxymorphone available as Opana and Opana ER since 2006. Despite the warning issued by FDA and CDC, Opana ER (extended release) continues to be used by recreational users in intravenous (IV) form leading to severe adverse effects like thrombotic microangiopathy (TMA).

Case Presentation: We herein, present an interesting case of a 37-year-old female with a history of active IV drug abuse and asthma who presented to our hospital for shortness of breath of 3 weeks duration. Her vitals were unremarkable except for respiratory rate of 22/min. Her physical exam was normal. Her labs suggested renal failure (creatinine 2.2 mg/dL) with baseline creatinine of 1.8 mg/dL. Urinalysis showed RBC sediments, many dysmorphic RBC casts along with nephrotic range proteinuria of 12 g/dL per day. Kidney biopsy was obtained that suggested microscopic thrombotic angiopathy (TMA) involving the glomeruli and vessels. Further work up was undertaken for TMA and apart from mildly elevated LDH of 380 (normal<243) and slightly low ADAMTS 13 (55%) there was no other lab evidence of TMA with normal platelet count, normal haptoglobin, mildly high reticulocyte count and no schistocytes in the peripheral smear. Scleroderma antibodies, antiphospholipid antibodies, anti GBM antibodies, C3 and C4 complements were negative. Hepatitis B and HIV was negative and hepatitis C antibody was positive, but cryoglobulin was negative. On literature search, we found that intravenous injection of chronic Opana ER has been reported to cause TMA resulting in chronic kidney disease. Our patient admitted with a history of ongoing intravenous Opana ER use for the last five years, therefore TMA was considered drug induced. She had a normal platelet count and an absence of schistocytes which makes it an atypical presentation of TMA resulting in CKD in an opiate user. As there was no evidence of thrombotic thrombocytopenic purpura(TTP), plasma pheresis was not done and eventually ADAMTS 13 was not low enough to suggest TTP for that level should be less than 10 %. The patient was offered opiate rehabilitation counseling, however, she left against medical advice.

Discussion: The Opioid epidemic is a growing public health concern. The diagnosis should be kept in mind in an injection drug user who presents with symptoms like TTP, HUS and subacute or chronic kidney disease of unclear etiology. We recommend health care professional to take a detailed history with focused questions on recent intravenous Opana ER abuse. In a nutshell, treatment is mainly supportive along with the treatment of underlying infection and avoidance of drug in the future that could be challenging in intravenous drug abusers like ours.
Pennsylvania-Clinical Vignette-Poster Finalist
Elan Mohanty, MBBS

Title: Bitter melon tea causing cola colored pee!

Authors: Elan Mohanty MD, Niranjan Tachamo MD, Izza Mir DO, Sijan Basnet MD, Ajay Koirala MD, Sharon Swierczynski, MD, PhD, Daniel Forman, DO

Introduction: Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common enzyme deficiency in humans, affecting 400 million people worldwide. While several drugs, foods and chemicals can trigger hemolysis in G6PD deficient individuals, the association between herbal and dietary supplements and hemolysis is not clear.

Case Presentation: A 42-year-old Vietnamese-American gentleman presented to the Emergency Department with generalized fatigue and intermittent dark urine for 1 week and fever, chills, vomiting and loose black stools for 1 day. He denied any rash, joint pain, strenuous exercise, recent travel or trauma, new medications or supplements and tick or mosquito bites. There was no history of valve replacement surgeries. Physical examination revealed icteric sclera and jaundiced skin. Pertinent labs included hemoglobin 9.7 g/dL with reticulocyte production index of 1.7 %, total bilirubin 5.2 mg/dL, direct bilirubin 0.7 mg/dL, lactate dehydrogenase 544 IU/L (normal: 94-202 IU/L) and haptoglobin 18 mg/dL (normal: 36 - 195 mg/dl). Urinalysis showed large amount of blood but no red blood cells on microscopy. Peripheral blood smear revealed bite cells, blister cells and polychromasia consistent with hemolysis. Direct Coombs test was negative. Blood smear for parasites, monospot test and heavy metal screen were negative. His hemoglobin dropped to 5.7 mg/dl and he was transfused 2 units of packed red blood cells. G6PD level was < 0.3 U/g Hb (normal: 8.8-13.4 U/g Hb). On revisiting the history, he mentioned increased consumption of Chinese bitter melon tea for his hyperlipidemia for 11 days prior to hospitalization. The patient’s symptoms improved with supportive care, his hemoglobin stabilized and he was discharged with a list of foods and medications to avoid.

Discussion: Bitter melon seeds contain vicine-like compounds which can induce favism. The severity of hemolytic anemia varies among individuals with G6PD deficiency and can be diagnostically challenging. Detailed history including all herbal and nutritional supplements is required to identify rarer causes of favism in susceptible individuals. Patient education regarding safe and unsafe food and medications is the cornerstone in preventing further episodes of hemolysis.
Pennsylvania-Clinical Vignette-Poster Finalist
Andres Mora Carpio, MD

Title: When an image is worth a thousand parasites: A case of neurocysticercosis in the city of brotherly love

Authors: Andres Mora MD, Daniela de Lima MD, Jessica Stempel MD, Antonette Climaco MD.

Introduction: Neurocysticercosis is an infection caused by the cysts of the parasite Taenia solium, which is endemic to Central and South America, Asia, India and sub-Saharan Africa and is associated to poor hygienic conditions. Infection develops after ingesting the parasite’s eggs by eating food contaminated with human feces from individuals infected with intestinal T. solium. The ingested eggs mature in the intestine and the larvae sets into different tissues forming cysticerci that cause asymptomatic infection that can last for years as they evade the host’s immune response. Eventually, the cysticerci degenerate, which triggers an immune response causing inflammation around the cysts. This inflammation is ultimately responsible for the symptoms. When cysticerci invade the brain, the most common manifestation is seizures, but hydrocephalus, vasculitis, meningitis, strokes and a myriad of different symptoms can be seen depending on the location.

Case Presentation: We present a case of a 74-year-old Brazilian female who lived her first few years in rural areas of Brazil and moved to the US in 1990. Medical history was pertinent for breast cancer status post chemotherapy and radiation, hyperparathyroidism and non-ischemic cardiomyopathy. She denied any history of intestinal or extra intestinal parasitosis.

She presented complaining of 4 years of intermittent vertigo that had progressed to constant nausea and ataxia with several falls, requiring multiple ED visits and eventually hospitalization. During her hospital stay a CT head and MRI brain showed a 2.2x2.5 cm cystic brain lesion with septations at the suprasellar cistern. Given this finding a possible differential diagnosis including cryptococcosis, neurocysticercosis, or echinococcosis was developed and serum crypto antigen, and ELISA for cysticercosis and echinococcosis were sent for diagnosis. Results: showed positive Antibodies to echinococcus and cysticercus. Western blot was negative for echinococcus and positive for cysticercus.

Treatment with prolonged course albendazole and dexamethasone was started, with addition of prophylactic levitiracetam. The treatment course was complicated by the development of cutaneous zoster that required hospitalization and IV acyclovir with appearance of post-herpetic neuralgia. Albendazole treatment was continued for 2 months as per department of health recommendations and serum cysticercus antigen was sent to CDC for titer evaluation which returned positive. Nausea and emesis resolved and vertigo markedly improved. Follow up MRI showed decrease in size of the cysts.

Discussion: Neurocysticercosis is an extremely uncommon condition in the US, that if unrecognized can leave lasting sequela as the cysts calcify and damage the brain’s architecture. Although the most common presentation is seizures, uncommon presentations must not be missed. Symptoms depend mostly on the area of brain involved. Our case shows the importance of having a broad differential diagnosis and evading anchoring as well as for thinking of atypical causes of symptoms when presentation of disease is atypical.
Pennsylvania-Clinical Vignette-Poster Finalist
Joseph V Moran, DO

Title: AMPA-R Antibody Positive Autoimmune Encephalitis: An under recognized cause of acute mental status changes

Authors: Joseph Moran, DO; George Prousi, MD; Stacey Smith, MD

Introduction: Autoimmune-/Paraneoplastic encephalitis presents with a variety of manifestations ranging from limbic encephalitis to neuropsychiatric symptoms and usually occurs in the presence of cancer. Autoimmune or paraneoplastic causes are often overlooked initially because symptomatology overlaps with metabolic and infectious causes of encephalitis which are exceedingly more common. This case presents a patient who was diagnosed with Anti-AMPA-Receptor Encephalitis after a one-month history of altered mental status.

Case Presentation: A 60-year-old female with rheumatoid arthritis presented with a one-month history of altered mental status and abnormal behavior. Initially presenting to an outside hospital, she underwent metabolic, infectious, and neuroimaging studies that were unremarkable. Her mental status did not improve over her 15 day hospital course. On follow-up presentation to our facility she was uncooperative and not oriented to person, place or time. Repeat MRI revealed increased DWI intensity in the cerebral cortices and increased FLAIR signal intensity in bilateral hippocampi. Differentials included CJD, vasculitis and paraneoplastic-/autoimmune encephalitis. Extensive work up for underlying malignancy was negative; however, studies showed positive Anti-AMPA-R antibodies. The patient made significant improvement with plasma exchange therapy, cyclophosphamide and steroids. The patient’s mental status and behavior improved and she was discharged in stable condition.

Discussion: Studies at the outside hospital were narrowly focused on common causes of encephalitis. Consideration of an autoimmune or paraneoplastic cause with corresponding studies could have led to an earlier diagnosis. Literature suggests a 60-yo woman with known autoimmune disease presenting with limbic encephalitis should be evaluated for anti-AMPA-R encephalitis when other causes have been ruled out. Prompt diagnosis is critical as 64% of patients may have underlying malignancy. Immunotherapy is the treatment of choice for patients without malignancy. In the present case, steroids were added due to the degree of inflammation seen on MRI. Overall, the patient responded well and prognosis is favorable with literature showing a 100% 5 year survival in patients with no underlying malignancy compared to 50% in those with malignancy. Close follow up is imperative as literature states a relapse rate of 50% and one study shows a 28% incidence of primary tumor diagnoses during relapse.

References


Pennsylvania-Clinical Vignette-Poster Finalist
Ritu Nahar, MD

Title: Cat Scratching your Valve: an Elusive Case of *Bartonella* Endocarditis

Authors: Ritu Nahar, Evan Caruso, Department of Medicine, Thomas Jefferson University Hospital, Philadelphia, PA

Introduction: *Bartonella Henselae* is an uncommon, but significant cause of “culture-negative” endocarditis. However, the spectrum of symptoms with which it manifests, the rarity of the disease, and limitations of diagnostic testing make this entity a diagnostic challenge.

Case Presentation: A 53-year-old female with a history of dysautonomia requiring pacemaker, membranoproliferative glomerulonephritis (MPGN), and chronic pulmonary embolism presented to an outside hospital with two years of fatigue, weakness, and weight loss, complicated by fevers of unknown origin (FUO) for 2 weeks. Persistent fevers despite broad spectrum antibiotics and negative infectious work up, prompted transfer for work-up of a presumed rheumatologic cause for FUO. Upon delving further into her history, the patient endorsed extensive contact with cats, assisting in kitten birthing 6 months prior to presentation. Physical exam revealed a 2/6 systolic murmur at the left lower sternal border, louder with inspiration. On transthoracic echocardiography a 2.2 x 1.9 echodensity was visualized adjacent the tricuspid valve in close proximity to the right ventricular (RV) pacemaker lead, with smaller echodensities on the atrial side of the RV pacemaker lead. Broad spectrum antibiotics were initiated but the patient continued to spike fevers as high as 104. Blood cultures remained sterile despite prolonged incubation periods. Transesophageal echocardiography 5 days later demonstrated progression of lesions, now revealing a 3.9 x 2.5 cm mass stemming from the RV pacemaker lead. On the 5th hospital day the patient developed severe pleuritic chest pain. Computed tomography (CT) of the chest demonstrated wedge shaped peripheral opacities suspicious for lung infarct from septic emboli versus thromboemboli. *Bartonella henselae* immunoglobulin G was found to be elevated at 1:512. The patient was switched to doxycycline and deemed appropriate for pacemaker removal with initiation of anticoagulation given evolving lung infarcts on CT. Management was complicated by heparin resistance requiring antithrombin concentrate pre and intra-operatively. On hospital day 10 she underwent successful cardiopulmonary bypass surgery for pacemaker and mass removal. Post-operatively she was initiated on a 6-week course of doxycycline, rifampin, and ceftriaxone for *Bartonella* endocarditis. Gentamycin was avoided given renal impairment from MPGN.

Discussion: This case highlights the importance of eliciting epidemiologic risk factors when working up FUO. Systemic complications our patient endured from lack of prompt intervention include recurrent hospitalizations and septic lung emboli. Additionally *Bartonella* endocarditis is associated with development of immune-complex glomerulonephritis. Thus her MPGN may also be a manifestation of her untreated endocarditis. Furthermore, while endocarditis is an established predisposing factor for decreased heparin responsiveness during cardiopulmonary bypass, the mechanism of this phenomena and how this alters management pre and intra-operatively is an area of research requiring further evaluation. Recognizing *Bartonella* endocarditis is critical in preventing systemic complications via earlier surgical intervention for source control augmented with adequate antibiotic coverage.
Title: The Disappearing Lung Mass

Authors: Shilpa Pedapati, MD; Crystal Duran MD; Muhammad Asif, MD; Deepa Kuchelan, MD; Kim Norville, MD

Introduction: PET Scans are an indispensable tool in the diagnosis and staging of cancer. However, several inflammatory conditions including trauma can cause intense uptake on a PET scan. A knowledge of these non-malignant causes of positive PET scans is important.

Case Presentation: A 33 y/o woman presented to the ED with sudden onset right-sided chest pain described as sharp, severe, non-radiating, non-pleuritic. She also had minimal nonproductive cough without hemoptysis. Medications included Mirena for contraception. She was an active smoker with a 5 pack year history. On examination, she was afebrile, BP 118/82, HR 101, RR 16, and O2 sats 97% on room air. Lungs were clear to auscultation bilaterally. EKG showed sinus tachycardia. D-Dimer was elevated at 1963. Contrast enhanced Computed tomography of the chest showed a 5 x 3 cm opacity in the right para-mediastinal region and right hilar lymphadenopathy. Malignancy was suspected and she was referred to Pulmonology for further work-up. PET scan was performed 10 days later, which showed an intense FDG uptake in the same right para-mediastinal region and anteromedial right upper lobe (SUV max of 10.8) and moderate FDG uptake within the right hilar lymph nodes. Interestingly the size of the opacity had decreased (now 4.0 x 1.9 cm) compared to the CT scan done 10 days prior. She underwent bronchoscopy. BAL and biopsies were negative for infection or malignancy. The bronchoscopy was complicated by a small pneumothorax which resolved. Within a few weeks, her symptoms completely resolved. On further questioning, she revealed that she is a recreational hunter and had recently started using a cross bow to hunt. A CT scan was repeated a month after the bronchoscopy for follow-up and showed complete resolution of the opacity as well as the hilar lymphadenopathy. The etiology of the CT findings were attributed to a cross bow injury to the chest resulting in a lung contusion.

Discussion: PET scans are highly sensitive (90%), but their specificity is 80% leading to false positive tests. Standardized uptake value (SUV) is a measure of the tumor’s metabolic activity — a high SUV(>2.5) indicates robust FDG uptake due to high metabolic glycolytic activity and suggests malignancy or active inflammation. Metabolically active infectious or inflammatory lesions including traumatic lesions can lead to substantial FDG uptake and thereby high SUV due to an increase in the glucose transporters on the cell membranes of the neutrophils and macrophages and cytokines that recruit these cells. In one systematic review, benign conditions causing PET positive lesions constituted about 25% of all the studies and half of them were markedly intense. This case is an interesting example of traumatic injury leading to a false positive PET scan and emphasizes the importance of history in the evaluation of pulmonary pathology.

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Pennsylvania-Clinical Vignette-Poster Finalist
Mary Rodriguez Ziccardi, MD

Title: Resistant Hypoglycemia with Hyperkalemia: An Unusual Cause

Authors: Mary Rodriguez Ziccardi, Maria Veronica Bandres, Alexander Pop, Sohaib Basharat, Ejaz Mahmood, Valentine Ezekwem.

Introduction: Hypoglycemia in the elderly is a common reason for admission which can present with symptoms ranging from dizziness, pre-syncope to seizures and coma. The predisposition to polypharmacy in this population and underlying comorbidities make them high risk for developing hypoglycemia. Interaction of medications, underlying nutritional and health status need to be evaluated in cases of hypoglycemia.

Case Presentation: A 64-year-old African American female with Past Medical History of Diabetes Mellitus type 2 on glipizide, Chronic hepatitis C, Arthritis and vascular ulcers in lower extremities with recent debridement and on treatment with Trimethoprim-Sulfamethoxazole was transferred from a nursing home for lethargy and hypoglycemia. In the emergency department, she was found to be confused, diaphoretic and hypoglycemic with a point-of-care blood glucose reported as “low”. She denied any prior symptoms including nausea, vomiting, fever, diarrhea or pain. Vital signs were stable. Physical exam was unremarkable except for morbid obesity and multiple clean base ulcers with erythematous borders in lower extremities.

Initial laboratory showed hypoglycemia with a blood glucose of 29mg/dl, creatinine of 1.7 mg/dl (baseline 1.3), BUN 40mg/dl and potassium of 8.4mmol/l. Electrocardiogram showed sinus rhythm with diffuse peaked T waves. Bilateral tibial and fibular X-rays showed diffuse soft tissue edema without evidence of osteomyelitis or emphysema.

Despite multiple glucose infusions and hyperkalemic measures (kayexalate, sodium bicarbonate, calcium gluconate, furosemide, and continuous albuterol nebulization), severe hypoglycemia and hyperkalemia persisted. Vancomycin and Cefepime were started for initial concern of concomitant sepsis. Only Dextrose solutions (5%, 10%, 50%) were used for the hypoglycemia. Glucagon and octreotide were not used due to hyperkalemia. Nephrology team recommended emergent hemodialysis. Hyperkalemia started to resolve after hemodialysis. Glycemic control was achieved 24 hours later.

Discussion: Hospitalizations for hypoglycemia are increasingly more common than hyperglycemia with associated higher morbidity. Interactions with certain antibiotics are a major cause of hypoglycemia in elderly patients with Diabetes Mellitus treated with sulfonylureas. Association with Trimethoprim-Sulfamethoxazole has been reported. The mechanism is believed to be interaction with hepatic metabolism of the sulfonylurea (CYP2C9 inhibitor) increasing this drug’s levels. Care should be taken with use in this population.
Title: Isolated celiac artery vasculitis- a rare cause of abdominal pain in young adult

Authors: Muhammad Hassaan Sattar, MD, Crozer Chester Medical Center, Muhammad Usman Ali, MD, Crozer Chester Medical Center, Sai Bhaskara Srinivas Sajja, M.D., FACS, Crozer Chester Medical Center

Introduction: Isolated celiac artery vasculitis is an unusual and rare etiology of abdominal pain. Vasculitis of the GI tract may occur in isolation or in the form of single-organ vasculitis (SOV). Isolated vasculitis tends to have a good prognosis, although it can also progress to a systemic illness. There have been limited data in the literature regarding isolated vasculitis of the GI tract and etiology is not well understood.

Case Presentation: A 43-years-old African American male with past medical history of hypertension and chronic back pain presented to the emergency department with 4 days of worsening sharp epigastric pain radiating to right lower quadrant, nausea and food intolerance. Initial vitals were within normal limits except blood pressure of 180/100. Notable examination includes mild epigastric and right lower quadrant tenderness and positive bowel sounds. There was no rebound tenderness or rigidity. Rest of the examination was within normal limits. Initial lab work including CBC, CMP, lactic acid and lipase was unremarkable. He received symptomatic treatment. Initial CT scan of abdomen showed no evidence for acute intra-abdominal or pelvic inflammatory process. No evidence of bowel obstruction and normal appendix. Patient was reassured and was sent home with ranitidine, Zofran and GI follow up. Upon further review of CT scan, a possible celiac artery dissection was diagnosed and patient was requested to come back to the emergency department. Patient continued to complain of nonspecific abdominal pain on revisit as well. He underwent CTA abdomen which showed findings of inflammatory soft tissue encasing the celiac artery origin consistent with vasculitis and causing significant narrowing of the common hepatic artery. The splenic artery and left gastric arteries appear patent. Further labs include ESR 29, normal complement levels and negative ANA/ANCA screen. The patient was started on high dose steroids and admitted to intensive care unit. Abdominal pain resolved dramatically with steroids and patient started to tolerate food. Patient discharged on oral steroids regimen and recommended to make outpatient follow up appointment with rheumatology.

Discussion: We present a case of isolated celiac artery vasculitis that was diagnosed via imaging studies in a young adult. Significant inflammation can cause narrowing of lumen of artery and compromise the blood flow leading to abdominal pain. Laboratory tests are nonspecific and ESR is usually below 30 in most of these patients (1). The diagnosis is usually based on characteristic radiographic imaging studies and/or histopathological examination of surgical specimens (1). There have been no clear guidelines; however steroids are the mainstay of treatment.

Although rare, isolated vasculitis can be a possible etiology of undiagnosed abdominal pain and high level of suspicion is required to evoke the diagnosis by further investigations. Localized vasculitis of gastrointestinal tract can be associated with significant morbidity and mortality if remain undiagnosed.

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Pennsylvania-Clinical Vignette-Poster Finalist
Joseph J Scuorzo

Title: Primary Hyperparathyroidism Presenting as Osteitis Fibrosa Cystica

Authors: Joseph Scuorzo DO, Dr. Angela Magdaleno; Dr. Gretchen Perilli

Introduction: Primary hyperparathyroidism is the third most common endocrine disorder, commonly presenting as an incidental finding of hypercalemia in the United States. Geographical regions marked by vitamin D deficiency result in patients presenting with more severe symptomatic disease often with normocalcemia. Osteitis Fibrosa Cystica (OFC) is uncommon with prevalence 1.5 to 1.7% and is characterized by large cystic bone lesions filled with giant cell osteoclastic tumors.

Case Presentation: A 54-year-old Egyptian female with history of chronic back pain and diabetes mellitus presented with a pathologic fracture of her left femur. Initial labs showed mild corrected hypercalcemia (10.6 mg/dL), hypophosphatemia (0.9 mg/dL), hypomagnesemia (1.0 mg/dL), hypokalemia (2.7 mmol/L), normal kidney function (creatinine 0.36 mg/dL) and elevated alkaline phosphatase (399 U/L). CT scan chest/abdomen/pelvis revealed thyroid masses and nephrolithiasis. Further imaging revealed diffuse lytic bone lesions. The femur fracture was treated with surgical fixation with bone biopsy to evaluate lytic lesions. Diagnostic labs included extremely elevated intact parathyroid hormone (PTH) (1962.5 pg/mL), severely low Vitamin D 25-OH (5 ng/mL), normal TSH, worsening corrected calcium (12.9 mg/dL), normal urine and serum protein electrophoresis, normal serum free light chains and normal PTH-related-peptide (<2 pmol/L). Pamidronate, calcitonin, and intravenous fluids were administered to treat the hypercalcemia. Parathyroid nuclear medicine scan showed a left lower lobe parathyroid adenoma. Bone biopsy Results: revealed giant cell lesion consistent with brown tumor of hyperparathyroidism. Thyroid fine needle aspiration of thyroid nodule showed benign nodular hyperplasia. Therapeutic parathyroidectomy was performed to treat OFC from primary hyperparathyroidism. To prevent hungry bone syndrome, the patient was treated with 50,000 units of vitamin D prior to surgery.

Discussion: The diagnosis of primary hyperparathyroidism is based on persistent hypercalcemia in the presence of elevated or inappropriately normal PTH. Our patient presented with a pathologic fracture and markedly elevated PTH level in the setting of severe vitamin D deficiency, masking her hypercalcemia. OFC is a rare presenting feature of hyperparathyroidism in the United States. OFC is characterized by pathological fractures and diffuse lytic bone lesions. After treatment with parathyroidectomy, the patient’s lytic bone lesions should heal and resolve within three months.
Pennsylvania-Clinical Vignette-Poster Finalist
Brianna J Shinn, MD

Title: Spontaneous Pneumothorax Affecting Three Generations in a Birt-Hogg-Dubé Syndrome Family: A Unique Pattern of Inheritance?

Authors: Brianna Shinn, MD, Department of Internal Medicine, Thomas Jefferson University Hospital, Philadelphia, PA, Gregory Kane, MD, Department of Internal Medicine, Thomas Jefferson University Hospital, Philadelphia, PA

Introduction:

Case Presentation: A 30-year-old male patient presented from an outside hospital where he was found to have a left-sided spontaneous pneumothorax and multiple bilateral pulmonary cysts in the basilar medial regions on high-resolution CT. The patient explained that he had been at work when he felt a pop in his left chest and heard gurgling. The patient underwent video-assisted thoroscopic surgery (VATS) and pleurodesis. We saw the patient in pulmonology clinic after hospital discharge.

Upon reviewing the patient’s family history, it was discovered that the patient’s sister, mother and maternal grandmother all suffered from repeated spontaneous pneumothoraxes and approximately twelve aunts, uncles and cousins suffered from renal cell carcinomas, skin lesions and pneumothoraxes. These clinical manifestations within a family are consistent with an autosomal dominant genetic disease called Birt-Hogg-Dubé syndrome (BHD) which is characterized by follicular hamartomas, pulmonary cysts, spontaneous pneumothoraxes and kidney neoplasms. [1] Five family members were genetically tested and found to be positive for BHD syndrome.

Discussion: BHD syndrome is an autosomal dominant condition caused by a germline mutation in the folliculin gene (FLCN) which is thought to act as a tumor suppressor gene. [2] The incidence of BHD syndrome is unknown but there are approximately 200 families in the world that have been identified. [3] Pulmonary cysts are reported in 80% of affected members of BHDS families but only 24% of affected members suffer from spontaneous pneumothoraxes. [2] Our patient’s immediate family history which demonstrates three generations with spontaneous pneumothoraxes is unique and suggests a more complicated interfamilial pattern of inheritance of the FLCN mutation, especially because of the different predominant manifestations in distant relatives.

Beyond discovering the inheritance of BHD syndrome and the variations that possibly exist between and within families with BHD, it is important for physicians and practitioners to understand the wide variety of symptoms that can be seen in BHD syndrome. Because of this variety, it has been suggested that BHD is likely underdiagnosed. [3] It is important for physicians to have a thorough understanding of the symptoms seen in BHD so that patients can get screened for renal neoplasms and undergo genetic testing for BHD syndrome. It is also important for patients and physicians to understand that several other neoplasms that have been associated with BHD syndrome such medullary thyroid cancer, thyroid adenomas, angiolipomas, intestinal colorectal adenomas, parotid oncocytesomas and parathyroid adenomas. [4]

References


Pennsylvania-Clinical Vignette-Poster Finalist
Karanveet Somel

Title: Erythema Annulare Centrifugum with relapsing Polychondritis

Authors: Somel K, Jajoria P, Kandala H, Parva N, Nookala V

Introduction: Erythema annulare centrifugum (EAC) is a chronic skin condition which manifests as the rash that is scaly, erythematous, ring-shaped and spreads centrifugally. Relapsing polychondritis is a rare connective tissue disease with symptoms that usually manifests as recurrent inflammation of cartilage throughout the body, including auricular, nasal, and tracheal cartilages. We herein present a rare case that initially presented with only a cutaneous manifestation of EAC, and eventually showcased relapsing polychondritis.

Case Presentation: A 58-year-old male with past medical history of EAC diagnosed in 2015, presented with pain in his hands and feet, left ear swelling and nasal congestion with mild nasal septal pain. He was diagnosed with polychondritis in 2016 when he presented with similar complaints as now, which was treated with multiple IM steroid injections and topical betamethasone. On examination, he had mild tenderness of his left ear cartilage with swelling and mild erythema over nasal septum. Labs showed elevated erythrocyte sedimentation rate (ESR) 89 mm/hr, C-reactive protein (CRP) 3.9 mg/L and angiotensin-converting enzyme (ACE) 80µL. Diagnosis of relapsing polychondritis was made, and immunosuppressive therapy was recommended, however, the patient wished to wait until his rash recurred. The patient came back in a month, with a new complaint of epistaxis and mild redness over nasal septum. ESR and CRP increased to 99 and 4.7 respectively. The patient was then started on Methotrexate 12.5mg once a week and folic acid 1mg daily. Prednisone was tapered. After one month, his rash and swelling improved. Methotrexate was increased to 15 mg once a week and continued with Prednisone at 22.5mg. ESR and CRP were within normal limits. Two months later he complained of rash recurring especially over his chest and increased pain in his hands, shoulders, hips, and knees. ESR and CRP remained within normal limits, his lab Results: were significant for low white blood cell count 3.4/microliter. Prednisone taper was at 10mg, folic acid was continued, and Methotrexate was increased to 17.5mg once a week. Despite this, his rash relapsed after two months. His prednisone was 2.5mg at this time and ESR was 23. The patient did not wish to increase steroid dose at that time. Methotrexate was discontinued, and Azathioprine 50 mg was started and after two weeks was increased to 75mg. This patient failed to follow up after this visit keeping us in dilemma about the efficacy of the treatment.

Discussion: The EAC association with relapsing polychondritis is rare and can present very early before the onset of polychondritis, as in this case. This may suggest a similar autoimmune or inflammatory process related to a concurrent systemic disease. We wanted to focus on the need for further investigations to rule out polychondritis and vigorous treatment with immunosuppressants in a patient with Erythema Annulare Centrifugum.
Pennsylvania-Clinical Vignette-Poster Finalist
Jian Liang Tan, MD

Title: An Ambiguous Choice of Anticoagulant in Atrial Fibrillation with Factor X Deficiency - A Management Dilemma

Authors: Jian Liang Tan, MD1, Kheng Joe Lau, MD2; Anshul Fnu, MD1; Arezoo Ghaneie, MD3, 1.
Department of Internal Medicine, Crozer-Chester Medical Center, PA, 2. Department of Internal Medicine, John H. Stroger Jr. Hospital of Cook County, IL, 3. Department of Hematology/Oncology, Crozer-Chester Medical Center, PA

Introduction: Amyloid light-chain (AL) amyloidosis is associated with coagulation abnormality such as factor X (FX) deficiency. FX binds to amyloid fibrils, hence shortened its half-life in the plasma. This is a report of a patient with a bleeding disorder (acquired-FX deficiency) who needs an anticoagulation for paroxysmal atrial fibrillation (PAF).

Case Presentation: A 64-year-old woman with past medical history of PAF, acquired-FX deficiency, AL amyloidosis, hypertrophic cardiomyopathy (HCM), metastatic lung adenocarcinoma on Erlotinib, hypertension, and diabetes was in a hematology clinic for a follow-up. She offered no new symptoms other than self-limited epistaxis and superficial bruising. In 2013, she was incidentally diagnosed with acquired-FX deficiency secondary to AL amyloidosis with a decreased FX coagulation activity of 45% (reference 70-150%). In 2015, she developed an episode of symptomatic PAF. Coagulation profile: prothrombin time (PT) of 16.3 seconds, international normalized ratio (INR) of 1.3, activated partial thromboplastin time (aPTT) of 33 seconds and FX coagulation activity of 34%. As she has HCM, her CHA2DS2-VASc score was 4 and the repeated FX level was 34% (not adequately self-anticoagulated). Hence, she was started on apixaban 5 mg twice daily with close outpatient follow-up.

Discussion: Nonetheless, there is presently insufficient evidence regarding the appropriate choice of anticoagulant in a patient with FX deficiency. The use of FXa inhibitor, in this case, raised a handful of concerns: 1.) there is no reliable way to monitor the level of anticoagulation; 2.) the efficacy of FXa inhibitor remains uncertain; 3.) patient with FX deficiency has an unpredictable bleeding risk. Alternative options: use 1.) FXa inhibitor at a lower dose; 2.) dabigatran, (a direct thrombin inhibitor) which has an antidote in case of a bleeding event; 3.) warfarin, monitor INR closely with a conservative goal of 2-2.5; 4.) no anticoagulant if FX level <20-30% to avoid bleeding event. However, more data or experience is needed to support such practices. As for our case, she has been on apixaban for the past 1.5 years without any bleeding or thromboembolic event. Her most recent implantable loop recorder did not show recurrence of atrial fibrillation. Upon reviewing the case, a decision has been made. Taking into consideration of her future unpredictable bleeding risk and no recurrence of atrial fibrillation, she was discontinued from apixaban with continuous heart rhythm monitoring and close follow-up.

This is a complex case with no clear answer on the appropriate choice of anticoagulant. Having a better understanding of the coagulation cascade would lead to a better selection of the anticoagulant. This case also highlights the importance of weighing the risk of bleeding and thromboembolic event prior to committing a patient on long-term anticoagulation.

References


Title: Recurrent Cerebrovascular Accidents In A Patient With Cardiac Calcified Amorphous Tumor: An Urgent Need For Timely Diagnosis

Authors: Anh Tran, DO¹ and Simona Opris, MD², (1) Internal Medicine, Drexel University College of Medicine, Philadelphia, PA, (2) Internal Medicine, Abington-Jefferson Health, Abington, PA

Introduction: Cardiac Calcified Amorphous Tumor (CAT) is one of the uncommon primary cardiac tumors. Despite their benign pathologic features of calcification and amorphous material, cardiac CATs can cause life-threatening conditions, one of which is embolic cerebrovascular accidents (CVAs). This prompts the need for urgent timely recognition, diagnosis and treatment of the condition.

Case Presentation: A 63 year-old Caucasian male, R.B. presented with worsening dysarthria and right hemiparesis. Patient had previous CVAs from 2007 to 2017, especially 5 new acute strokes over a period of 3 months from March to May 2017. R.B. has a long-standing history of cocaine abuse (last reported use in 03/2017) and uncontrolled hypertension with medication non-compliance. He also had 14 episodes of syncope prior to 03/2017, thought to be related to ongoing cocaine abuse at the time. During his previous hospitalizations, cardiology was consulted, stating no clear cardioembolic events with an unremarkable TTE and no further recommended cardiovascular testing.

Upon arrival at our hospital, MRI/MRA demonstrated chronic small vessel ischemic changes, 2 foci of acute small vessel infarction in the left parietal periventricular white matter, and multiple prior microhemorrhages. TTE demonstrated hyperdynamic left ventricles, normal right ventricular function, no valvular lesions. Given his multiple CVAs and high pre-test probability of cardiac embolic source, TEE was performed, illustrating a focal echodense mobile mass, 5x0.6mm, on left coronary cusp, which could have been a source of emboli.

Subsequently, R.B. underwent surgery for tumor removal. Pathological report of the resected mass demonstrated a fragment of degenerated hyalinized tissue with calcifications, consistent with cardiac CAT. Cultures from blood and tumor were negative, ruling out infective endocarditis. R.B. was later discharged to rehabilitation.

Discussion: There are approximately 42 case reports of CAT from 1997 to 2014 with a mean age at detection in the 50s and female predominance. Recurrent embolic CVA within the short time period caused by CAT is a rare entity.

TTE is a common and widely used tool in majority of hospitals. However, its sensitivity and specificity is not as good as TEE for the detection of cardiac tumors. When patient’s story is consistent with intracardiac emboli, invasive TEE is the test of choice despite the normal TTE. Could our patient’s CAT have been diagnosed earlier as the cause of recurrent CVA if his cocaine use or uncontrolled hypertension did not obscure the picture of possible cardiac embolism?

This case demonstrated the need for essential, timely diagnosis of CAT in particular and heart tumor in general so that early surgical intervention can be performed to prevent further serious sequelae of cardiac embolism.
Pennsylvania-Clinical Vignette-Poster Finalist
Benjamin Ware

Title: Relapsing Fevers Leading to Neurologic Collapse in the New York City Area

Authors: Benjamin Ware MD, Robert Ulrich MD, Paawan Punjabi MD, Department of Medicine, New York University School of Medicine, New York, New York

Introduction: Viral encephalitis can be difficult to diagnose because symptoms are often non-specific and laboratory tests can take several days to weeks to result. West Nile Virus (WNV) is a mosquito-borne infection that arrived in New York in 1999 and can now be seen during the summer and fall in all 48 contiguous states and Canada. Most infections are asymptomatic, but 1 in 4 develop fever and 1 in 150 develop neuroinvasive disease.

Case Presentation: A 73 year-old man from northern New Jersey with well-controlled diabetes presented in August 2017 with 2 weeks of relapsing nightly fevers, drenching sweats, frontal headaches, and malaise. Exam revealed temperature 101°F, no neurologic deficits, and no rashes. Initial labs showed leukopenia (WBC 3.1). Chest x-ray, blood cultures, and respiratory pathogen PCR panel were negative. Vancomycin, piperacillin-tazobactam, and azithromycin were started on admission but nightly fevers continued. After infectious disease consultation on hospital day (HD) 2, these were discontinued and doxycycline was initiated for suspected tick-borne infection. Blood smear was negative for babesiosis and PCR for anaplasmosis and ehrlichiosis were sent. Symptoms initially improved, but late on HD 3, high fever (103.1°) and headache recurred. Vancomycin, cefepime, and ampicillin were started for presumed meningitis. Leukopenia evolved to leukocytosis (WBC 12.1) with a lymphopenic differential (5%). Lumbar puncture revealed neutrophilic pleocytosis (1213 WBC, 83% PMN), elevated protein, and normal glucose. CSF PCR for common meningitis pathogens (7 viral, 6 bacterial, and 1 yeast) was negative. Despite broad spectrum antibiotics, by HD 6, the patient remained febrile and had become progressively somnolent. Right sided weakness developed. Brain CT and MRI did not show neurovascular changes. By HD 7, he was unresponsive to commands and required intubation. Ultimately, plasma WNV IgM and RNA PCR returned elevated on HD 9, and CSF viral RNA PCR on HD 13. Subsequent course was complicated by seizures. Supportive care was continued and intravenous immunoglobulin (IVIG) was trialed for 5 days without improvement. After a 45 day hospital course, patient was unable to achieve meaningful cognitive recovery and was discharged to a long-term acute care facility.

Discussion: Relapsing fevers, headache, and cytopenias can be caused by a variety of infections. In the northeastern United States, tick-borne infections are classic examples. However, WNV should be on the differential diagnosis for any unexplained summertime fever. This case evolved into neuroinvasive disease. While neutrophilic CSF initially rose concern for bacterial causes, early neuroinvasive WNV can cause neutrophilic predominance. The relative peripheral lymphopenia is also suggestive of WNV in the literature. This case illustrated focal neurologic deficits in WNV. Treatment for WNV is largely supportive and specific therapies, such as IVIG, lack evidence. WNV neuroinvasive disease needs more effective treatment options.

References
Title: Giant Non-Traumatic Abdominal Wall Hematoma in an Anticoagulated Patient as a Complication of Acute COPD Exacerbation

Authors: Bradley F. Woodman, M.D., Darren Morris, B.S., Tiffany Becker, M.D., Jennifer Arena, PA-C, Rohit Jain, M.D.

Introduction: The spontaneous abdominal wall hematoma is a rare cause of acute abdominal pain [1, 2]. Risk factors include advanced age, anticoagulation, and increased abdominal straining from cough. Here we describe the case of a geriatric woman on therapeutic anticoagulation that developed a giant, non-traumatic oblique abdominal wall hematoma secondary to cough while hospitalized for a COPD exacerbation.

Case Presentation: An 82 year old woman with a past medical history significant for COPD on home oxygen and atrial fibrillation on apixaban was initially admitted to the intensive care unit with hypoxic and hypercarbic respiratory failure secondary to a COPD exacerbation. Her respiratory status improved and she was transitioned to the intermediate care unit. She continued to have significant coughing despite therapy with antitussives, and she began to strain more vigorously for bowel movements despite bowel stimulation. On hospital day 5, the patient complained of mild left sided abdominal pain. On examination her left abdominal quadrants were slightly tender with no palpable masses. The following morning her hemoglobin had dropped from 11.7 to 8.4 g/dL, and the left side of her abdomen appeared swollen and ecchymotic with increased tenderness to palpation. She had a computed tomography scan of the abdomen and pelvis with IV contrast which showed a 22.6 x 8.7 x 7.3 cm hematoma along the left ventral abdominal wall with no evidence of active bleeding. Her apixiban was discontinued, she remained hemodynamically stable however when her hemoglobin fell to 7.1 g/dL she was transfused one unit of packed red blood cells. Her hematoma was conservatively managed with bedrest, analgesics, and warm compresses. She remained hemodynamically stable, the hematoma gradually began to regress, and she remained off anticoagulation when she was discharged from the hospital.

Discussion: A spontaneous abdominal wall hematoma is rare clinical entity [1]. Clinicians should consider abdominal wall hematomas in their differential for abdominal pain, particularly in geriatric patients on therapeutic anticoagulation. Increased coughing, such as with COPD exacerbations, is a risk factor for developing a spontaneous hematoma. The most common symptoms are acute abdominal pain, a palpable abdominal mass, and anemia. A CT scan is more sensitive than ultrasound for establishing the diagnosis [3]. Conservative treatment is appropriate in hemodynamically stable patients, which includes rest, analgesia, transfusions, and correction of coagulopathies. Most patients are able to resume anticoagulation following resolution of the hematoma, as recurrence rates are low [4].

References

Puerto Rico-Clinical Vignette-Poster Finalist
Aliana Bofill, MD

Title: Cavitary Disease Is Not Always Tuberculosis

Authors: Aliana Bofill, MD; Stefano Coppola, MD; Joel Matos, MD; Glenda Gonzalez, MD, Ivan Caraballo, MD

Introduction: Cavitary pneumonia is a rare complication of community-acquired pneumonia. Streptococcus pneumonia is one of the most common causative agents in this type of lower respiratory tract infection. However, cavitary lesions are extremely rare with this pathogen. It is more frequently reported among patients with concurrent bacteremia, which may reflect the greater severity of disease among bacteremic patients. Treatment is supportive with antibiotics however, it has become more challenging since antibiotic-resistant strains are becoming more prevalent. Fluroquinolones have been extensively used to treat S.pneumoniae with increasing reports of resistant strains. Here we present a case of acute cavitary pneumonia with multi-drug resistant S.pneumoniae in an immunized patient.

Case Presentation: This is a case of a 70-year-old man with past medical history of hepatitis C, diabetes mellitus, treated for latent tuberculosis in 1993, who came to the emergency room due to nasal congestion, cough productive of green sputum, and night sweats since 1 week ago. He denied weight loss, chills, fever, hemoptysis, recent travel. Initial vital signs were 99.4F, 89bpm, 20bpm, 96%O2. Laboratory findings remarkable for chronic thrombocytopenia. Computed tomography of the chest was performed revealing a cavitating paramediastinal parenchymal lesion between the anterior of the left upper lobe and superior segment of the lingula. Patient was admitted to internal medicine ward with airborne isolation. During the admission, patient remained clinically stable, sputum cultures, blood cultures and three acid-fast bacilli sputum were obtained. Both, blood and sputum cultures reported S.pneumoniae resistant to fluoroquinolones and penicillin but, susceptible to vancomycin and cephalosporin. He was started on ceftriaxone. The three acid-fast bacilli sputum smear taken were negative. Bronchoscopy was done revealing an erythematos and edematous mucosa bilaterally greater over left lung with abundant purulent secretions. No lesions were identified. Bronchial washings negative for growth and cytology with inflammatory cells but no malignant cells. Following 2 days of antibiotics patient showed clinical improvement and was discharged home.

Discussion: Cavitary lesions are often seen in tuberculosis and malignancy. They can also be present in a variety of infectious processes but, those attributable to S.pneumoniae are extremely uncommon; thus, to date, epidemiological studies are lacking. In our case, the patient had hepatitis C and diabetes which are risk factors for developing cavitating pneumonia. Possibility of Streptococcus C6 serotype was considered giving immunization status. However, he had a favorable progression with medical therapy despite being a multi-resistant strain. Diagnosis can be difficult since sputum and blood cultures are positive in only half of the cases. Thus, it is important that S. pneumoniae must be considered in the differential diagnosis of the patient with acute cavitary pneumonia and routine testing of pneumococci for susceptibility to fluoroquinolones should now be considered since it is widely use as empiric treatment of pneumonic process.
Puerto Rico-Clinical Vignette-Poster Finalist
Nicolle Canales Ramos, MD

Title: Henoch-Schönlein Purpura Nephritis: Not Only a Childhood Disease

Authors: Nicolle Canales-Ramos, MD; Krystahl Andújar-Rivera, MD; Ileana E. Ocasio-Melendez, MD; Sharlene Medina, MD; Fatima Cintrón-Rosa, MD; Naomi Collazo, MD

Introduction: Henoch-Schönlein Purpura Nephritis (HSPN) is often regarded as a childhood disease and is rarely seen in adults. Patients are at risk of developing chronic kidney disease if treatment is delayed. Thus, it is vital that clinicians be vigilant for the symptoms of this rare disease.

Case Presentation: A 51-year-old Puerto Rican male without known medical history complained of sore throat and chills for three days. He was prescribed Azithromycin by primary physician. Four days later, he developed a rash on his lower extremities initially thought to be a drug eruption. Despite discontinuing the antibiotic, the rash rapidly progressed over the course of 24 hours involving buttocks, mid-back and abdomen. Associated symptoms included arthralgia, gross hematuria, abdominal pain and bloody stools. On physical examination, palpable purpura was present on legs, back and buttocks. Vital signs were stable. Laboratory results showed 2.0 mg/dL of serum creatinine. Microscopic examination of urine revealed dysmorphic red blood cells. Proteinuria of 1,988 mg/g was quantified using urine protein-creatinine ratio. Serum complement levels C3 and C4 were normal. Hepatitis B surface antigen, hepatitis C antibody, anti-neutrophil cytoplasmic antibodies, antiglomerular basement membrane antibodies, anti-nuclear antibody and serum cryoglobulins were negative. Renal ultrasound showed kidneys of adequate size, shape and echotexture. A renal biopsy was performed revealing increased mesangial cellularity and mesangial staining with IgA on immunofluorescence. Focal areas of tubular epithelium and vessels were positive for C3. He was diagnosed with Henoch-Schönlein purpura nephritis. After a three-day course of intravenous methylprednisolone pulse, he was started on oral prednisone and azathioprine. Upon clinical improvement, the patient was discharged home. Two weeks later, serum creatinine level returned to baseline, there was improvement of proteinuria and resolution of palpable purpura and abdominal complaints.

Discussion: HSPN is a rare kidney disease and is most prevalent in the first decade of life, making our adult case even more unusual. Among children, the incidence per 1 million is 15-70 patients while in adults it can decrease to 4 patients. Renal involvement in adults with HSPN is usually more aggressive than in children and up to forty percent of patients can progress to chronic kidney disease. One of the challenges the physician encounters is balancing the cost of immunosuppressive treatment versus the actual risk of developing chronic kidney disease. Likewise, choosing between immunosuppressive agents can be difficult since more evidence-based recommendations are needed to pinpoint which agent is the choice of therapy for HSPN. In a clinical trial by Bergstein et al, observations suggest that corticosteroid and azathioprine therapy is beneficial in treating HSPN. In our case, treatment with glucocorticoids and azathioprine therapy proved to be effective in eliminating abdominal symptoms, decreasing proteinuria and improving renal function.

References


Title: An Unusual Presentation of Small Bowel Obstruction

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Introduction: Eosinophilic gastroenteritis is an inflammatory disorder characterized by eosinophilic infiltration of the esophagus, stomach, duodenum, and colon without any known etiology. It is a rare digestive disease characterized by the triad of eosinophilic infiltration of segments of the gastrointestinal tract, abnormalities of gastrointestinal function (can range from dyspepsia and obstruction to diarrhea and ascites) and exclusion of other diseases with peripheral eosinophilia.

Case Presentation: Case of a 72-year-old male who presents to the emergency room with complaints of abdominal pain that began the day before. Patient states having had abdominal pain one week prior. Patient denied fever, chills, nausea, emesis, diarrhea, or sick contacts. Initial evaluation remarkable for soft and depressible abdomen with localized periumbilical pain upon palpation. Initial laboratories revealed negative U/A, CBC with marked leukocytosis with an elevated eosinophil count of 28%. Chemistries with preserved renal function, no electrolyte abnormalities, normal pancreatic enzymes. CT of abdomen and pelvis was performed which revealed small bowel obstruction with wall thickening suggestive of pneumatosis as well, suspicious for ischemic bowel. Patient was admitted to the Surgical Ward and patient underwent abdominal exploratory laparotomy in which partial enterectomy with primary anastomosis was performed, without any complications. Pathology report revealed focal acute enteritis, erosion and a marked eosinophilic infiltrate involving the mucosa and full thickness of the bowel wall, extending into the serosa, with no microorganisms or granulomas seen. Ova and parasites including Schistosoma and Strongyloides were negative. Tryptase levels normal for hypereosinophilic syndrome or mast cell disease. Bone marrow biopsy performed for further evaluation and revealed no abnormal pathology. Endoscopic studies failed to reveal any abnormalities as well. All other causes of eosinophilia were eliminated leaving eosinophilic enteritis as the cause of this patient's small bowel obstruction.

Discussion: Eosinophilic enteritis is a rare disease of exclusion but must always be entertained when a patient presents with gastrointestinal symptoms and eosinophilia.

References

Puerto Rico-Clinical Vignette-Poster Finalist
Yamila Goenaga Vazquez, MD

Title: Catastrophic Consequences: An Undiagnosed Atypical Presentation of Neuromyelitis Optica in an Otherwise Healthy Woman

Authors: Yamila Goenaga Vázquez MD1, José Ávila Ornelas MD2, Ana C. Méndez Gómez MD3, José C. López Puebla MD4, Amanda C. Doval López MS4, Doriann González Rodríguez MS4, Lucia Rivera Matos MS4, 1Med-Peds Residency Program, University of Puerto Rico Medical Sciences Campus, 2Neurology Residency Program, University of Puerto Rico Medical Sciences Campus, 3Transitional Year Residency Program, Damas Hospital, 4Internal Medicine Residency Program, University of Puerto Rico Medical Sciences Campus, 5University of Puerto Rico Medical Sciences Campus, School of Medicine, 6Universidad Central del Caribe, School of Medicine

Introduction: Neuromyelitis Optica (NMO) spectrum disorders (NMOSD) are inflammatory disorders of the CNS characterized by immune-mediated axonal demyelination predominantly targeting optic nerves and spinal cord. The presence of immunoglobulin G (IgG) that binds to aquaporin receptor 4 (AQP4) known to be key in the pathogenic process of this disorder, distinguishes NMOSD from other CNS inflammatory disorders. NMO has a relapsing course in 90% or more of cases. In some patients, optic neuritis (ON) and transverse myelitis (TM) occur concurrently; in others, clinical episodes are separated by a variable time delay, which typically is no longer than 5 years. The postpartum period is a particularly high-risk time for severe relapses. Currently, there is no effective treatment for this disorder. However, diagnosis of NMOSD calls for an early and aggressive treatment with immunosuppressive therapy which reduces the frequency and severity of attacks. Failure to identify this entity may result in severe clinical disability demonstrated by permanent blindness and quadriplegia.

Case Presentation: 61-year old female presented to the hospital with ascending subacute progressive quadriparesis and paresthesias one week prior to evaluation. Patient had a history of abrupt bilateral vision loss at age 17 that resolved spontaneously. At age 35 blindness became permanent in the immediate postpartum period. Upon neurologic examination, she had bilateral decreased visual acuity with afferent pupillary defect. She was arreflexic with spastic quadriplegia and anesthesia to all modalities below C3 sensory level. Cerebrospinal fluid (CSF) showed 13 erythrocytes/mm3 and 102 leukocytes/mm3 with 99% mononuclear predominance; glucose was 101mg/dL, protein 147mg/dL and negative VDRL. Gram stain showed no organisms and no oligoclonal bands were present. Cervicothoracic MRI revealed long segment diffuse abnormal signal intensity of the spinal cord from cervical medullary junction to T7 with patchy contrast enhancement favoring an acute/subacute non-specific myelitis. Patient was started on Solumedrol 1 gram IV daily for 5 days. Hospital course was complicated by respiratory failure requiring mechanical ventilation. In view of unchanged neurologic status, five courses of plasmapheresis were given. Subsequently, AQP4-IgG antibodies came back positive confirming diagnosis of NMO. Today, after 2 months of initial diagnosis, patient continues to be critically ill at the ICU with quadriplegia and blindness.

Discussion: In retrospect, there were multiple clues suggesting the diagnosis of NMOSD in this patient, including two episodes of bilateral vision loss, the second of which led to severe residual blindness. However, the diagnosis was not made in a timely matter. Our patient’s clinical presentation was atypical in regards to the sequence of events. There was a long gap between initial presentation (ON) and TN of almost 4 decades, which further misled the patient’s physicians. Had this patient been treated initially with immunosuppression, probably it wouldn’t have progressed to the current severe clinical disability and dismal expectation of recovery that she faces at this moment.
References

Puerto Rico-Clinical Vignette-Poster Finalist
Yamila Goenaga Vazquez, MD

Title: Acute Retinal Artery Ischemia and Profound Cytopenias in a Young Patient with Coexisting Systemic Lupus Erythematous and Paroxysmal Nocturnal Hemoglobinuria.

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Introduction: In many patients with Systemic Lupus Erythematous (SLE), the natural course of the disease is punctuated by hematological complications such as cytopenias and higher risk for thrombosis. While cytopenias in SLE are thought to be multifactorial, thrombosis is usually attributed to antiphospholipid antibodies, among other known risk factors. In cases where these are not identified, exclusion of rare causes of thrombosis is essential. Paroxysmal nocturnal hemoglobinuria (PNH) is one of such rare causes and is characterized by the deficiency of glycosylphosphatidylinositol (GPI) anchor proteins which protect cells from complement mediated lysis. The relationship between SLE and PNH have been described only in rare instances. It is possible that the coexistence of both disorders may aggravate hematological complications in these patients.

Case Presentation: 26-year old female without systemic illnesses presented to the hospital with a 6-week history of generalized weakness, malaise, anorexia, ten-pound weight loss, headaches, myalgias, arthralgias, dyspnea, and decreased visual acuity in left eye. On initial evaluation, physical examination revealed alopecia and joint tenderness involving several joints. Ophthalmology evaluation was remarkable for left retinal artery ischemia. Chest X-rays showed large bilateral pleural effusions. Initial laboratory workup was remarkable for pancytopenia with a white blood cell count of 1,570 cells/microL, platelet count of 82,000 cells/microL, and hemoglobin concentration of 11 g/dL. Serologic tests revealed positive anti-nuclear antibodies (titer 1:320), anti-Ro antibodies, and hypocomplementemia with reduction in both C3 and C4. Thrombophilia workup including anti-thrombin deficiency, protein C and S deficiency, prothrombin gene mutation, and antiphospholipid antibodies (anticardiolipin IgG and IgM, anti-beta 2 glycoprotein IgG and IgM, and lupus anticoagulant) were negative. Bone marrow aspiration and biopsy, were remarkable for loss of CD24 and Fluorescent Aerolysin (FLAER) on neutrophils, loss of CD14 and FLAER on monocytes, and PNH clonal expansion of 27%. The patient was diagnosed with SLE and was treated with intravenous high-dose corticosteroids followed by oral prednisone, hydroxychloroquine, and azathioprine. She had a remarkable clinical improvement and was eventually discharged with resolution of clinical findings. When followed at rheumatology clinics she has continued to demonstrate sustained clinical improvement with no evidence of organ-threatening disease.

Discussion: Diminished expression of GPI anchored proteins and small PNH clone could be associated with enhanced complement-mediated lysis of blood cells causing cellular depletion and thrombosis at unusual sites in this patient with SLE and PNH. Since she responded well to standard immunosuppressive therapy we suggest these patients should be treated by targeting the underlying autoimmune disorder. However, it is important to closely monitor the clone because expansion have been described creating devastating consequences. To the best of our knowledge, this is the first time that the relationship between PNH to both cytopenias and thrombosis is reported in a patient with SLE.
References

Puerto Rico-Clinical Vignette-Poster Finalist
Eduardo J Gonzalez Bonilla, MD

Title: “Mosquito bite hurts so much that I couldn’t walk”

Authors: González, E. MD; Santiago, P. MD; Rodríguez, R. MD; González, S. MD

Introduction: Guillain-Barre Syndrome (GBS) is an immune mediated disorder affecting the peripheral motor and sensory nerves and nerve roots, and is the most common cause of rapidly progressive generalized paralysis. The condition is rare, but is extremely rare if is produced after mosquito bite, especially if Zika Virus causes it.

Case Presentation: Case of a 78 year old woman with past medical history of DM type 2, Neuropathy, Hypertension, Peripheral Vascular disease, Osteoarthritis and Hypothyroidism that is brought to emergency department after falling at home. Patient refers being well of being until 2 days ago when started with general fatigue and extreme weakness. Had previously a urinary tract infection treated with cephalaxin and thought symptoms were associated with current illness. She describes the fall, as feeling numbness and tingling sensation at hands and feet that made it difficult to ambulate and was the reason for falling. Patient reports having lumbar pain 8/10 intensity that radiated to bilateral lower extremities. Vital signs were normal and no fever reported. Physical exam was unremarkable except for extremities and neurologic exam which demonstrated decreased pulses bilaterally and cranial nerves 2-12 was intact, decreased deep tendon reflex on patellar and Achilles’ tendon sites, decrease of motor strength bilaterally at lower extremities. When laboratories arrived, WBC: 9.8, Hemoglobin: 14.4g/dl, Hematocrit: 42.4% and Platelet 272 with normal differential. No abnormalities seen on basic metabolic panel and all imaging studies arrived negative for acute changes. During the first two days of admission ascending flaccid paralysis started with neurologic deterioration continues and lumbar puncture was perform and Results: were: Colorless, clear with WBC: 1mm3, RBC 6-8mm3/hpf, Lymphocytes 100%, Protein 166mg/dl consistent with Guillain-Barre Syndrome. Patient is initially started on Intravenous Immunoglobulin for treatment of syndrome. Final result from Cerebrospinal fluid arrived and Culture, Gram stain, Glucose: 145mg/dl, Streptococcus group B, Streptococcus pneumonia, Haemophilus influenza, type b, Neisseria meningitides ACYW135, E. coli and Indian ink were negative. Family members were interviewed and they refer that 5 days earlier of when weakness started, patient developed upper respiratory symptoms and diffuse rash on whole body that wasn’t evident at the moment of initial evaluation. Chikungunya and Zika titer were ordered. We received a call from CDC to report a positive Zika Virus result on PCR. After 15 days of hospitalization patient was discharged to rehabilitation center.

Discussion: An estimated 3,000 to 6,000 people, or 1-2 cases for every 100,000 people, develop GBS each year in the US. GBS is strongly associated with Zika; however, only a small proportion of people with recent Zika virus infection get GBS. CDC is continuing to investigate the link between GBS and Zika.
Puerto Rico-Clinical Vignette-Poster Finalist
Deyson Lorenzo-Rios, MD

Title: Eschars as a sign of a deadly disease

Authors: Deyson Lorenzo-Rios, MD, Department of Internal Medicine, Veterans Affairs Caribbean Healthcare System. San Juan, Puerto Rico US.

Introduction: Cutaneous aspergillosis is an invasive skin infection caused by *Aspergillus* species occurring mostly in the immunocompromised patients. The infection can be classified as primary, when is associated to interrupted skin integrity; or secondary, when is developed during disseminated disease.

Case Presentation: A 67-year-old man with chronic lymphocytic leukemia and hypogammaglobulinemia presented to the emergency room with progressive fatigue for 2 weeks. He completed chemotherapy with fludarabine, cyclophosphamide and rituximab 4 years ago, and continued ibrutinib and prophylactic trimethoprim/sulfamethoxazole. On initial examination, he was afebrile and pale, with irregular pulse, and generalized xerosis. Laboratories showed severe anemia, thrombocytopenia and lymphocytosis. Patient was admitted to the medical intensive care unit under the diagnostic impression of autoimmune-hemolytic anemia and atrial fibrillation. Eventually, a 3.5-cm round shaped, well-demarcated necrotic eschar, surrounded by minimal edema and erythema, was found on his right distal lateral leg. The lesion started as a tiny pink spot, similar to an insect bite, that rapidly darkened and increased in size over 3 weeks. Patient denied fever, chills, local trauma or pruritus. A perilesional punch biopsy demonstrated extensive necrosis, hemorrhage and acute inflammatory infiltrate involving epidermis, dermis and subcutaneous fat. Gomori methenamine-silver staining showed bullous forms within septated hyphae infiltrating into subcutaneous tissue, consistent with chronic cutaneous aspergillosis. Fungal morphology from isolated tissue culture was compatible with *Aspergillus flavus*. No microorganisms were identified in blood cultures. Patient was started on voriconazole and discharged home to continue steroid therapy for hemolytic anemia. He died at home 1 week later during a febrile episode.

Discussion: Lymphoproliferative disorders and medical regimens, such as corticosteroids, ibrutinib and rituximab, have been associated to invasive aspergillosis. Cutaneous invasion by aspergillosis can be locally destructive, and after overcoming mechanical and immunological barriers, it may serve as a portal for developing systemic disseminated disease. The combination of these factors may have potentiated the skin invasion into a systemic disease in our patient. Customized sensitive diagnostic tools, such as serum galactomannan antigen detection, and aggressive combined therapy with azoles and echinocandins should be considered in high risk patients with an alarming skin finding like ours. Eschars should never be underestimated as they may indicate disseminated disease and high mortality risk.

References


Puerto Rico-Clinical Vignette-Poster Finalist
Jonathan Rivera, MD

Title: Recognizing the Cause of Digital Ischemia While Presenting Similarities With Two Different Entities. –Case Report

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Introduction: Septic emboli are commonly associated with multiple medical conditions, including infective endocarditis -characterized by its heterogeneous presentation. The risk of embolization in these patients is approximately 50%. Within the population at risk of developing infectious endocarditis are intravenous drug users. Amongst them, patients that are medically supervised for opioids withdrawal. Most of these patients are on sublingual buprenorphine enrolled in opioid detoxification programs.

The buprenorphine is a partial agonist of mu opioid receptors and reduces the symptoms of opiate withdrawal. A recent practice is to dissolve the buprenorphine film, and inject it intravascularly. Such practice can affect the artery and compromise endothelial recovery. Buprenorphine excipients can directly damage the endothelium, releasing vasoactive substances that induce vasoconstriction, and ultimately, limb ischemia. This clinical presentation in an IV drug user can be misdiagnosed as a septic embolus.

Case Presentation: 39-year-old male, patient with history of IV drug use presents with sudden-onset of severe pain in the left hand since 3 days prior to admission. Patient indicates he was removing a syringe from left forearm, a self-accessed IV point for injection of dissolved buprenorphine film, when he gradually developed localized sharp pain in his first three fingers of his left hand, along with pallor and flushing, followed by a blue discoloration and severe pain. He indicates that when he attempted to move the digits, their purpuric hue worsened. He admits to some shortness of breath, general malaise and unquantified fever, but denies chills, chest pain, loss of appetite, dizziness or weight loss. He was admitted under Internal Medicine for further evaluation. An arteriogram was done by Interventional Radiology that revealed digital arteries in digits 1-2 are patent, at least proximally, but markedly reduced in caliber suggestive of severe vasoconstriction. No evidence of frank thrombosis.

Patient was moved to ICU, treatment was initiated with a heparin drip and analgesics without significant improvement on symptoms. Calcium channel blockers were administered with slight relief. Sublingual nitrates were added as well for vasodilation. After a few days of continued treatment, digits regained color and movement. Unfortunately, afterexperimenting significantly improvement of symptoms, patient decided to leave against medical advice prior to coordinating transesophagic echocardiogram. He left the hospital with adequate functioning of the left hand. Blood cultures were negative, and no fever was reported during the hospital stay.

Discussion: It is of utmost importance to distinguish between severe artery vasospasm and septic emboli. Since IV buprenorphine use is a relatively recent trend, increased awareness of this possible diagnosis is necessary in order to treat accordingly and establish standard, adequate management to prevent further complications, limb compromise and even life-threatening clinical situations.
Puerto Rico-Clinical Vignette-Poster Finalist
Frances Rodriguez Berrios, MD

Title: When Mast Cells Attack

Authors: Rodriguez-Berrios, Frances, M.D.; Jordan, Patricia, M.D.; Valle-Cancel, Viviana, M.D.

Introduction: Mastocytosis is a rare condition characterized by massive mast cell proliferation causing a spectrum of different possible clinical presentations. Cutaneous involvement may present as flushing, rash and pruritus while systemic manifestations include allergic reactions, anaphylaxis and gastrointestinal complaints. The differential diagnosis is vast as symptoms are broad and non-specific, making diagnosis challenging and delaying adequate treatment.

Case Presentation: 78 year-old male with past medical history of hypertension, diabetes mellitus type 2, coronary artery disease, atrial fibrillation, and myelodysplastic syndrome presented to the emergency department after developing unintentional weight loss, chronic diarrheas and dyspepsia of 2 months duration. Vital signs upon initial evaluation were found within normal limits. Physical examination demonstrated a cachectic patient, with normal cardiopulmonary and abdominal examination, but no lymphadenopathy or rash was observed. Laboratory Results: showed pancytopenia, white blood cell at 4.2 X10^3/ul, hemoglobin 9.2 L g/dL, platelet 86.0 L X10^3; worsening renal function, and hypokalemia. Patient’s daughter and son had a prior history of colon cancer, for which an upper gastrointestinal tract (UGIT) endoscopy and colonoscopy were performed; UGIT endoscopy demonstrated a hiatal hernia and peptic ulcer disease, and colonoscopy was essentially unremarkable. Further imaging studies included abdominal computed tomography (CT) scan and positron emission tomography/CT scan demonstrating gastro-hepatic, portacaval, pericaval, retroperitoneal and abdominal mesenteric lymphadenopathy, with SUVs ranging from 2.1 to 2.7. Mild hypermetabolic spleen and skeletal bone marrow were also noted, for which and ultrasound core biopsy of the spleen was performed; pathology preliminary report described fibrosis with increased eosinophilic, monocytoïd cell infiltrate, as well as hyalinized granulomas. In view that diagnosis was inconclusive, bone marrow biopsy was warranted and analysis showed mast cell tryptase (+), CD117(+), CD34(-), CD2(-), CD30(-). Patient was started on imatinib treatment, however was discontinued due to side effects of persistent diarrhea. Later, treatment was changed to interferon alpha 2b and response was monitored. Three months later, recurrent ascites started to develop, requiring frequent diagnostic and therapeutic paracentesis. Fluid cellularity had abundant macrophages, occasional mast cells, and eosinophils. Within the multiple systemic mastocytosis classifications, this case most consistent with aggressive mastocytosis, due to the presentation with cytopenias, ascites, and palpable splenomegaly.

Discussion: Aggressive mastocytosis subtype, as presented in this case, is a rare condition that may cause organ dysfunction due to excessive mast cell infiltration. Bone marrow, liver, spleen and gastrointestinal tract are most commonly involved. Pathogenesis of this condition is crucial when determining its treatment, making its diagnosis essential.
Puerto Rico-Clinical Vignette-Poster Finalist
Janiabeth Vega Maldonado, MD

Title: Oh Oh my foot just dropped! A case of Mononeuritis Multiplex Associated with Hepatitis C Infection

Authors: Janiabeth Vega Maldonado M.D., Damaris Ortiz M.D., Jazmin Feliz M.D., Raúl Llinás M.D., Milton D.Carrero Quiñones M.D.

Introduction: Mononeuritis multiplex is a painful, asymmetrical, asynchronous sensory and motor peripheral neuropathy involving isolated damage to at least 2 separate nerve areas. As the condition worsens, it becomes less multifocal and more symmetrical. Foot-drop is the most common manifestation of mononeuritis multiplex. Common causes of damage include a lack of oxygen from decreased blood flow or inflammation of blood vessels causing destruction of the vessel wall and occlusion of the vessel lumen of small epineurial arteries. Mononeuritis multiplex can be associated with infections such as hepatitis C, acute viral hepatitis A, hepatitis B, AIDS and HIV infection.

Case Presentation: This is a case of a 31 y/o male patient with a past medical history of intravenous drug abuse (IVDA) with heroin. Patient arrived at the emergency department after being found unconscious in the sidewalk with evidence of multiple insect bites in the lower extremities. Patient was admitted due to rhabdomyolysis, upon arrival to ED patient he was found to have volume depletion and acute kidney injury, initially elevated total CPK levels of 47,098 and continued to increase until they reached 92,172, after aggressive hydration CPK levels decreased until resolution. During hospitalization the patient started complaining of bilateral foot pain and paresthesia, upon physical examination he was found to have left lower limb decreased sensation in a stocking distribution and left foot drop. An electromyography and nerve conduction studies were done and the patient was started on physical therapy and gabapentin. Since the patient was a former IVDA and was presenting with signs and symptoms of peripheral neuropathy we suspected an infection as the most likely cause of his current symptoms, therefore a hepatitis panel was sent. Results: came positive for hepatitis C infection. A consult with an ID specialist was ordered and a workup for genotyping was initiated. Results: of his electromyography and nerve conduction studies showed an asymmetric peripheral axonal sensory-motor neuropathy involving the bilateral lower extremities, being the left side more involved. Patient EMG Results: were compatible with a mononeuritis multiplex. Therapy for hepatitis C was initiated.

Discussion: The prevalence of peripheral neuropathy among patients with hepatitis C virus (HCV) varies from 8% to 10.6%. We can learn from this case the importance of obtaining a thorough history and physical examination in patients with suspected hepatitis infection, presenting with lower limb pain and paresthesia, as with our patient who was diagnosed with mononeuritis multiplex using electromyography and nerve conduction studies and the importance of treating the underlying for improvement of the patient symptoms.
Title: Tricky Treponema: An unusual presentation of syphilitic hepatitis in a HIV positive patient

Authors: Lena Fan, MD Second Author: Philip A. Chan, MD, Timothy P. Flanigan, MD

Introduction: Syphilis is a sexually transmitted disease with significantly increased rates over the last several years. Given that syphilis is increasing, physicians should be aware of unusual presentations. We describe a case of hepatitis as the initial presenting symptom of secondary syphilis.

Case Presentation: A 34-year-old HIV positive male on antiretroviral therapy (CD4 count 275, viral load nondetectable) without a history of syphilis, presented with 10 days of intermittent fevers (max temperature of 102 F), chills, night sweats, headache, 14-lb weight loss, foul smelling watery brown stool, and total body rash. The rash was a non-pruritic, non-painful, maculopapular rash beginning in his bilateral extremities before spreading to his trunk and back, sparing his face and palmar/plantar surfaces of hands and feet. He was initially treated with a 3-day course of azithromycin 500mg daily with no improvement. Outpatient work up was notable for a cleared hepatitis C infection, prior Epstein Bar Virus and Cytomegalovirus infections, as well as a transaminitis with Aspartate aminotransferase 179 IU/L, Alanine aminotransferase 213 IU/L and Alkaline phosphatase 570 IU/L. He denied any sick contacts, tick bites, exposure to wooded areas, chemical exposure, new medications, or over the counter supplements. Sexual history was notable for only one male partner in the past six months with whom he had anal receptive sex using condoms, but he later revealed he also had oral sex with another male partner (unknown syphilis status) without condoms. Physical exam on admission was notable for hepatomegaly, resolving rash, and no penile lesions.

A right upper quadrant ultrasound showed cholelithiasis with decompressed gallbladder. Abdominal MRI showed hepatomegaly without cirrhosis or gallbladder pathology. His Rapid Plasma Reagin was found reactive (1:256) with reflex treponema pallidum IgG Antibody >8.0 with negative serologies six months prior. Given positive antibody, his symptomatology was determined to be attributable to a new diagnosis of secondary syphilis. He was immediately treated with a single dose of intra-muscular Penicillin G Benzathine 2.4 Million Units. He was monitored for a Jarisch-Herxheimer reaction, which included fevers and chills. Upon resolution of symptoms he was discharged home. At his one-month follow up with his outpatient immunologist, his transaminitis had completely resolved.

Discussion: According to the CDC, in 2015, Rhode Island reported 64 new HIV diagnoses and the incidence rate of primary and secondary syphilis was 7.3 per 100,000 persons, a dramatic increase from 2011. This case illustrates the importance of considering syphilis as a potential diagnosis in patients who present with abnormal hepatic function tests regardless of their medical history. Early recognition of syphilitic hepatitis can help prevent further transmission of syphilis, especially in the HIV positive population.
Title: Postoperative fevers of unknown origin: a diagnostic challenge

Authors: Marya Haq, MD, MHA; Amr El Meligy, MD; Fatima Zeba, MD; Taro Minami, MD, FACP; John Miskovsky, MD

Introduction: Systemic lupus erythematosus (SLE) is an autoimmune multisystem disorder characterized by a relapsing and remitting course. Patients are classically women of childbearing age who present with low grade fever, arthralgia, and a malar rash. This report describes an unusual case of SLE in a patient whose first presentation was fever precipitated by surgical stress.

Case Presentation: A 48-year-old woman who successfully underwent L4-L5 laminectomy and fusion for lumbar radiculopathy developed a dry cough and persistent fever on postoperative day two. Blood cultures obtained on postoperative day four were positive in one of two bottles at 22 hours with Methicillin-sensitive Staphylococcus Aureus (MSSA). Broad spectrum antibiotics were initiated, initially with vancomycin and piperacillin-tazobactam, and later with cefazolin and daptomycin. However, fever persisted and worsened in severity. All subsequent blood cultures remained negative.

On postoperative day seven, she developed worsening non-productive cough, oral mucositis, leukopenia (3200-3800/mcL), anemia, myalgias, arthralgias, malar and petechial rashes, and acute kidney injury (serum creatinine rose from 0.70 to 1.64). Computed tomography (CT) scan of the chest revealed ground-glass opacities and bilateral pleural effusions. Serologic testing was performed to evaluate for potential infectious, malignant, and autoimmune causes of patient's presentation. Results: were remarkable for low serum complement levels (C3, C4) and positive antinuclear antibodies (1:640), anti-smith, anti-SSA/Ro, anti-U1RNP, anti-dsDNA antibodies. Of note, anti-histone antibodies were also positive, and this classical finding of drug-induced lupus can be seen in up to 80% of idiopathic SLE (Tetikkurt, C.,2016).

Skin biopsy of the patient’s petechial rash revealed perivascular lymphocytic infiltrate without eosinophilia; immunofluorescence revealed a weak lupus band of C3 as well as IgG and IgM deposition in basement membrane. A diagnosis of SLE was made and the patient was started on IV methylprednisolone. All of her symptoms improved markedly with complete resolution of her fevers, arthralgias, myalgias, cough, rash and acute kidney injury within forty eight hours. Renal biopsy was performed subsequent to this hospitalization and demonstrated mesangial lupus nephritis.

Discussion: Postoperative fever is usually attributed to infection, deep venous thrombosis, or drug-related causes. Inflammatory disorders are much less common etiologies and are placed lower on the differential diagnosis. While there have been instances of patients with SLE experiencing flares after surgical stress, to our knowledge, this is the first reported case of new-onset SLE triggered by surgical stress. In patients experiencing a postoperative fever without an identifiable source that is unresponsive to antibiotics, a thorough history and physical examination should be done to identify stigmata of SLE, with early initiation of corticosteroids to prevent end-organ damage.

References

Rhode Island-Clinical Vignette-Poster Finalist
Lolita S Nidadavolu, MDPHD

Title: A novel approach to managing central diabetes insipidus refractory to desmopressin: long-term administration of a free water sliding scale without serum sodium levels

Authors: Lolita Nidadavolu, MD, PhD; Marissa Martz, MD; Rebekah Gardner, MD

Introduction: A 41-year-old woman with Human Immunodeficiency Virus (HIV) on highly active antiretroviral therapy presented with altered mental status. Magnetic resonance imaging (MRI) of the brain revealed a complex cystic suprasellar mass, hydrocephalus and ventriculomegaly.

Case Presentation: Craniotomy for mass resection was performed with pathology demonstrating grade 1 benign craniopharyngioma, for which she received six weeks of radiation. Her hospital course was complicated by delirium, secondary hypothyroidism, secondary adrenal insufficiency, and severe central diabetes insipidus (CDI). A percutaneous endoscopic gastrostomy (PEG) tube was placed to help meet nutritional goals. Her urine output (UOP) increased to 1300 milliliters per hour and she was started on desmopressin twice daily. Despite continuous adjustment of the desmopressin dose and free water flushes via PEG tube, her UOP remained unpredictable, leading to fluctuations in serum sodium from 120 to 150 millimoles per liter. Persistent delirium and impaired thirst mechanism prevented her from drinking sufficient water while hypernatremic, despite having access to fluids. We created a sliding scale for water flushes administered via PEG tube based on her urine output and successfully stabilized her serum sodium to normal range. The free water sliding scale was designed to mimic the commonly used insulin sliding scale used in long-term care facilities and to account for limited access to serum sodium measurement after hospital discharge. One month post-discharge, she continues on this regimen at a long-term care facility.

Discussion: This case highlights the difficulty of managing CDI in a patient with altered mental status and impaired thirst mechanism, or adipsia. Patients with CDI with a preserved thirst mechanism, access to free water and ability to drink, are able to maintain normal serum sodium levels. General approaches to CDI with adipsia include administering fixed doses of desmopressin, scheduling fluid intake with a sliding scale based on body weight and serum sodium changes and frequent monitoring of serum sodium levels. A review of the literature shows that a free water sliding scale has been used for children with adipsic central diabetes insipidus; however, it was based on home serum sodium monitoring. Our initial inpatient CDI management strategy measured serum sodium every four hours, adjusting free water flushes accordingly. This strategy was not feasible at the long-term care facility as they were unable to frequently monitor serum sodium and recalculate free water deficit throughout the day. We developed a free water sliding scale administering flushes via PEG tube based off of urine output measured every four hours. We developed this plan in collaboration with the long-term care facility, incorporating their staffing and laboratory capabilities. This approach led to sustained eunatremia. This case demonstrates a successful approach for administering free water to match urine output in CDI patients with adipsia who reside in long-term care facilities.
Rhode Island-Clinical Vignette-Poster Finalist
Reema Qureshi, MD

Title: Aortoesophageal Fistula: A rare complication of Left Atrial Ablation

Authors: Reema O. Qureshi; Sena Kilic; Juliet Yirerong; Paulette Pinargote

Introduction: Esophageal injury is a rare complication of left atrial ablation procedures. Undiagnosed, it can progress to ulceration, perforation and aortoesophageal fistula (AEF) formation and has a 100% mortality rate.

Case Presentation: We present a rare case of AEF. A 60 year old man with a history significant for atrial fibrillation and bicuspid aortic valve (AV) with mechanical AV replacement presented to the emergency department (ED) 5 weeks after surgical revision of his AV and a MAZE procedure. He presented with chest pain, palpitations and shortness of breath for 5 weeks and fever and neurological deficits for 1 day. His physical exam was remarkable for tachycardia, cannon A waves, and a systolic ejection murmur. Electrocardiogram showed Atrioventricular Nodal Reentrant Tachycardia. Lab-work and imaging were unremarkable. His ED course was remarkable for hematemesis, hypotension and fever. An esophageogastroduodenoscopy (EGD) revealed a 1cm luminal defect concerning for a fistula in the middle third of the esophagus. A computed tomography scan did not confirm this finding.

On the morning of Hospital Day 2, he developed projectile hematemesis. A repeat EGD was done revealing oozing from the previously identified fistula site. The patient developed pulseless ventricular tachycardia and the scope was withdrawn. After 59 minutes of resuscitation he was pronounced dead. Autopsy revealed an AEF.

Discussion: AEF is a rare complication of left atrial ablation procedures with a high mortality rate. The clinical presentation is vague and variable. Rapid diagnosis and surgical therapy are imperative.
Rhode Island-Clinical Vignette-Poster Finalist
Jose R Ruiz, MD, MPH

Title: A BADAS Initial Presentation of Crohn’s Disease

Authors: Jose R. Ruiz, MD, MPH. Jennie Johnson, MD. Steven Moss, MD. Department of Medicine. Warren Alpert Medical School, Brown University.

Introduction: Bowel Associated Dermatosis-Arthritis Syndrome (BADAS) is a rare, systemic presentation of an underlying gastrointestinal abnormality. BADAS has classically been described as a complication following ileo-jejunal bowel surgery. A number of case reports have described BADAS in patients with inflammatory bowel disease (IBD). BADAS usually presents with rapidly progressing skin eruptions, diffuse polyarthralgias, low grade fevers and gastrointestinal complaints.

Case Presentation: A 55-year-old woman with no known past medical history presented with two weeks worsening facial rash and one day of fevers. The rash started as facial flushing and small erythematous papules over her cheeks that spread to her forehead. She was evaluated by dermatology and treated with topical ivermectin for rosacea and acne. The papules evolved into honey-crusted pustules and she was then treated with cephalexin for presumed impetigo, without improvement. On initial physical examination, the patient had honey crusted plaques and draining pustules on her cheeks and forehead and papules on her chest, shoulder and abdomen. The rest of her physical exam was unremarkable. No significant laboratory abnormalities were noted. She also reported a 3 month history of intermittent non-bloody diarrhea. A Tzanck smear showed multinucleated cells. The patient was started on acyclovir and broad spectrum antibacterials. Gram stain of pustule fluid showed many neutrophils but no organisms; bacterial cultures were negative. VZV and HSV were undetectable by PCR. Her rash evolved into large oozing, crusted pustules. She developed malaise, erythematous, tender subcutaneous nodules on her shins, tenderness and synovitis of her wrist, elbow and ankle joints and worsening, now bloody, diarrhea. Skin biopsy revealed intradermal neutrophilic inflammation with ulceration and intracorneal neutrophils with negative PAS. Bacterial and fungal stains and cultures remained negative.

The patient underwent colonoscopy, which demonstrated moderate colonic inflammation in the sigmoid colon surrounded by normal mucosa. Biopsies showed focal mildly active ileitis and severely active colitis with ulceration, basal cell lymphoplasmacytosis, crypt architectural distortion and Paneth cell metaplasia, consistent with Crohn’s Disease. She was started on steroids and then transitioned to rituximab therapy. Several weeks after initiation of treatment, the patient’s facial rash and other symptoms had almost completely resolved with only scant patchy erythematous discoloration of her cheeks remaining.

Discussion: This is the first known report of BADAS presenting prior to a diagnosis of IBD. BADAS is presumed to result from a Type III immune-mediated response to increased systemic circulating levels of sterile bacterial antigens. Recently, this syndrome has increasingly been associated with IBD. However, it may be initially overlooked, resulting in extensive antimicrobial therapy and expensive diagnostic work up prior to initiating appropriate IBD treatment. Therefore, it is important to be aware of this syndrome and able to recognize it early in its course.

References


"Bowel-associated dermatosis-arthritis syndrome. Immune complex-mediated vessel damage and increased neutrophil migration."

Title: A Case of Idiopathic Pulmonary Fibrosis Associated with Atypical Anti-Neutrophil Cytoplasmic Antibodies

Authors: Roy Souaid, MD, Faeq Kukhon MD, Andre Kharabi MD, Patricia Russo-Magno MD, Abdullah Chahin MD, Memorial Hospital of Rhode Island, Providence, RI., Warren Alpert Medical School, Brown University, Providence, RI., Kent Hospital, Warwick, RI.

Introduction: Idiopathic pulmonary fibrosis (IPF) is chronic, restrictive, fibrosing interstitial pneumonia of unknown etiology. Anti-neutrophil cytoplasmic antibodies (ANCAs) are important diagnostic tools in detecting many autoimmune disorders especially systemic vasculitis. Because of the overlapping symptoms between connective tissue disorders secondary to vasculitis affecting the lung and interstitial lung diseases (ILD), ANCA assay is obtained as part of serologic studies to ascertain connective tissue disorders are not overlooked during the diagnostic work-up. We present a case of IPF that was found to be associated with atypical p-ANCA.

Case Presentation: An 83-year-old man with past medical history of hypertension, hyperlipidemia, and advanced dementia presented to the Emergency Department (ED) with dyspnea and hemoptysis for one month duration. Prior to his presentation, he was treated for the same complaints with a course of azithromycin for presumed acute bronchitis. In the ED, he was found to be hypoxemic with vitals as follow: Temperature of 36.1, blood pressure 143/83, pulse 66, respiratory rate 18. O2 saturation 90% on 4 liters.

Physical examination with impressive bibasilar crackles on auscultation. Initial blood work-up with elevated Wbc at 13.9 with 71.8% neutrophils, no bands and negative procalcitonin.

Chest radiograph showed bilateral ground-glass opacities scattered in both upper and lower lobes more prominent on the right side. The patient was started on antibiotics, steroids, and bronchodilators, and was admitted to the hospital for presumed COPD exacerbation and pneumonia given his history of smoking although was never diagnosed with COPD.

Extensive work-up including sputum culture, sputum acid-fast bacilli (AFB) smear, β-D-glucan, C-ANCA, anti-nuclear antibody (ANA), and anti-glomerular basement membrane antibodies were all negative. p-ANCA came back positive with negative proteinase-3 and myeloperoxidase staining patterns. Patient was started on cyclophosphamide. However, he became more hypoxic and was transferred to the Intensive Care Unit (ICU). He was started on pulse steroids. Computed tomography (CT) scan of the chest showed mediastinal adenopathy and was suggestive of evolving fibrotic process compared to a prior scan done 3 years earlier. Bronchoscopy was then performed and a biopsy was taken which showed diffuse alveolar damage very consistent of Idiopathic pulmonary fibrosis in an accelerated form.

Given the biopsy results, patient’s advanced age, and baseline dementia combined with poor performance prior to admission, the family was informed of his poor prognosis. The family requested that comfort measures take place. Patient peacefully expired.

Discussion:

IPF is a multifactorial condition that is still extensively studied for better understanding of its pathophysiology. In the literature, many reports described IPF cases that were associated with ANCA. In
in our case, atypical p-ANCA titers were found to be elevated in association with IPF. This might suggest a possible role for this autoimmune process, resulting in IPF.
Title: SPONTANEOUS CORONARY ARTERY DISSECTION IN A MALE WITH INTENSE COUGHING SPASMS

Authors: Juliet Yirerong 1, Reema Qureshi 1, Paulette Pinargote 1, Heather Hurlburt 2, 1 - Memorial Hospital of Rhode Island/Brown University, 2 - Brigham and Women's Cardiovascular Associates at Care New England

Introduction: Spontaneous Coronary Artery Dissection (SCAD) is a rare cause of acute myocardial infarction, accounting for 0.1-0.4% of all acute coronary syndrome pathologies, with about 80% of cases occurring in young women in the peripartum period, without a history of heart disease or coronary artery disease risk factors, and less than 10-15% of cases occurring in men. We report a case of SCAD in a male who presented with chest pain.

Case Presentation: 55 year old male with a history of hypertension, remote tobacco use, presented with a sudden onset of 10/10 substernal chest pain radiating to the back. A week prior to this, he had developed shortness of breath with an associated violent, ongoing, productive cough shortly after inhaling thick, black petroleum-based fumes in an enclosed warehouse. He denied cocaine use.

Physical exam notable for hypoxia on room air, bilateral rhonchi on lung auscultation, no chest wall tenderness, regular heart rate and rhythm with no murmurs, rubs or gallops.

Laboratory investigations revealed an initial negative troponin, with EKG showing no evidence of dynamic ischemic changes. Chest Computed Tomography Angiography ruled out pulmonary embolism and aortic dissection. Troponin peaked at 29.9, subsequent EKG obtained showed non-specific T wave flattening in the lateral leads. NSTEMI was diagnosed. Aspirin, clopidogrel and enoxaparin were initiated.

Transthoracic echocardiogram reported normal left ventricular wall motion and contractility, Ejection Fraction 65%. Left heart catheterization with coronary angiography was significant for a filling defect throughout the second obtuse marginal branch of the left circumflex artery, consistent with a linear dissection with intramural thrombus. Rheumatologic screening was negative. Patient was managed conservatively with dual antiplatelet therapy, high intensity statin, an ACE inhibitor and a beta blocker. He was doing well on follow up at the cardiology outpatient clinic, 2 weeks after discharge.

Discussion: Though seemingly rare, SCAD should be considered as a differential for patients presenting with acute myocardial infarction in the setting of frequent, intense Valsalva-like activities such as coughing or retching, even in the non-classic male population.

Recognition of SCAD is key, as patient characteristics and management differ considerably from typical acute coronary syndrome cases. An increasing number of SCAD cases are being identified due to recent heightened awareness, use of invasive angiography, and advanced intravascular imaging. This calls for outlined guidelines regarding SCAD management.

Cases reported thus far suggest that in stable patients with preserved coronary blood flow, conservative management is associated with spontaneous healing of dissection the majority of the time. Our patient’s favorable outcome goes to further buttress the recommendation of a conservative approach in uncomplicated, clinical situations.
Rhode Island-Clinical Vignette-Poster Finalist
Fatima Zeba

Title: Not always a contaminant: An interesting case of prosthetic valve endocarditis due to Lactobacillus rhamnosus

Authors: Fatima Zeba MD, Juliet Yirerong MD, Geetika Tewary MD

Introduction: Lactobacilli are relatively avirulent organisms forming part of the normal flora of the oral, gastrointestinal and genitourinary tracts. At times misdiagnosed as a contaminant, Lactobacillus has uncommonly been reported to cause intra-abdominal abscesses, bacteremia, pneumonia and endocarditis. We present a rare case of Lactobacillus prosthetic valve endocarditis.

Case Presentation: A 74 year old male with a history of coronary artery disease status post coronary artery bypass graft with bioprosthetic aortic valve replacement (AVR) a year prior, diabetes mellitus, presented to the Emergency Department with acute onset confusion and disorientation. Notably, he had been admitted three months prior for acute encephalopathy ultimately thought to be secondary to Lactobacillus acidophilus bacteremia and treated appropriately with antibiotics. Workup for septic foci including computed tomography (CT) head, lumbar puncture (LP), transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), computed tomography angiography (CTA) of the head was negative at the time. Prior to this, he had been treated for an Escherichia coli urinary tract infection with ciprofloxacin.

Physical exam was notable for fever (38.6ºC), fair dentition and a grade 3/6 ejection systolic murmur over the aortic area.

Laboratory investigations showed leukocytosis of 20,300. CT Head, LP, urinalysis, chest X-Ray, were unrevealing. TTE showed a thickened aortic valve leaflet, ejection fraction (EF) 40-45% which was reduced compared to a prior study. Blood cultures isolated gram positive coccobacilli so patient was started empirically on ampicillin-sulbactam and vancomycin. He developed a Mobitz type 2 AV block requiring transvenous pacing. TEE showed EF 30-35%, severe aortic valve leaflet thickening and a vegetation on the aortic bioprosthetic leaflet. Coronary CTA revealed findings consistent with an aortic root abscess. Blood cultures isolated Lactobacillus rhamnosus, antibiotics were deescalated to ampicillin-sulbactam.

Patient underwent redo AVR with patch closure of aortic root abscess. Intraoperative specimen of the prosthetic valve vegetation isolated Lactobacillus rhamnosus, sensitive to penicillin, for which he completed a 6-week course of therapy. Workup for infection focus or associated underlying illness including Panorex, CT chest, abdomen and pelvis, colonoscopy was negative except for esophagogastroduodenoscopy (EGD) that showed white esophageal plaques suspicious for candidiasis. He was discharged on postoperative day 17; afebrile, with improved mentation and resolved leukocytosis.

Discussion: Of the identified predisposing factors for Lactobacillus bacteremia/endocarditis, our patient had a history of recent surgery, recent endoscopy (TEE), recent use of antibiotic therapy resistant to Lactobacillus, and prosthetic valvular disease. Cancer, cirrhosis, diabetes mellitus are among the underlying illnesses noted to be associated with clinically significant Lactobacillus infection. Our patient was diabetic; workup for malignancy was negative. Given EGD findings, gut translocation was considered...
a possible etiology. This case highlights the importance of avoiding dismissing Lactobacillus as a blood culture contaminant, especially in the presence of identified predisposing factors. A diagnosis of Lactobacillus bacteremia should always prompt work-up for an underlying disease condition.

References

South Carolina-Clinical Vignette-Poster Finalist
Mohamed Faris, MBBCH

Title: A 23 Year Old Female with Stroke: Complications of Severe Uncontrolled Hypertension in Neurofibromatosis type 1

Authors: Mohamed Faris MD, Michelle Baliss, OMS-IV student, Robert Coni DO, Donald Eagerton MD, Baby Kodali MD, Vinod Nambudiri MD

Introduction: Objective: Recognize the consequences of uncontrolled hypertension in a patient with neurofibromatosis type 1 (NF1).

Case Presentation: A 23-year-old female with a history of difficult-to-control hypertension since age 12 presented with acute onset of left sided weakness, left sided facial droop, slurred speech, and a severe headache.

On presentation, her initial blood pressure was markedly elevated at 240/140 mmHg. On examination, the patient had a blunted affect, and was oriented to person, time, and place. She had a left-sided hemiparesis and sensory loss, left sided hemineglect, slurred speech, positive Babinski on the left foot, flexion contracture of the left upper extremity, left sided nasolabial fold flattening, and hyperreflexia on the left side.

Skin findings included widespread large café-au-lait macules, neurofibromas, and axillary freckling as well as multiple pigmented iris lesions consistent with Lisch nodules.

The patient had a known family history of neurofibromatosis (NF-1) and had had an optic glioma diagnosed at age 5.

Non-contrast head CT revealed a hemorrhagic stroke affecting right parietal lobe and right basal ganglia.

Urinary catecholamines and metanephrines were measured, CT and MRI of the head, neck, and abdomen were performed, and outside imaging including an MRA one year earlier were reviewed; there was no tumor, vascular structural abnormality, or other contributory etiology.

Blood pressure control required using several antihypertensives including labetalol, nicardipine, lisinopril, and clonidine.

The patient was discharged to a stroke rehab facility following her stay.

Discussion: Neurofibromatosis-1 (NF1) is an autosomal dominant disorder with a prevalence of 1/3000 that is characterized by various cutaneous and neurological manifestations.

16% of NF1 patients develop hypertension, which may be essential or secondary. Patients with NF1 have a higher incidence of bilateral renal artery stenosis, pheochromocytoma, and coarctation of the abdominal aorta than the general population.

The association of NF1 with multiple conditions related to hypertension makes the evaluation of hypertension in patients with NF1 a noteworthy diagnostic challenge.
When compared with the general population, the odds of any type of stroke are significantly increased for patients with NF1, both adult and pediatric. This risk is most notable for hemorrhagic strokes.

Physicians should be aware of the increased risk of stroke in this population, and consider stroke as a potential cause of new neurological symptoms.

This case illustrates the importance of recognizing and managing hypertension in patients with NF1, given potentially devastating hemorrhagic stroke at a young age.

The association of NF1 with pheochromocytomas, bilateral renal artery stenosis, and coarctation of the aorta should prompt evaluation for these possibilities in patients with NF1 to ensure institution of treatment and prophylaxis as appropriate.

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South Carolina-Clinical Vignette-Poster Finalist
Navya Nambudiri, MD

Title: Visual Diagnosis of Amyloidosis: Multiple Clinical Manifestations of Extensive Plasma Cell Dyscrasia

Authors: Navya S. Nambudiri MD, Mo Faris MD, Vinod Nambudiri MD MBA

Introduction: Visual diagnosis based on physical examination is a critical component of an internal medicine physician’s skillset. Cutaneous manifestations of systemic disease can help guide clinicians toward an appropriate workup for patients.

Case Presentation: A 55-year-old female presented for evaluation endorsing “being ill for over a year.” She noted extensive fatigue, and was particularly concerned about progressive lower extremity edema bilaterally. She reported developing “pouches of fluid” that traveled up to her thighs at night. Additionally, she noted a diffuse rash on her trunk and extremities that had been present for over a year. Her past history was notable for hypertension, for which she had not been taking medication due to cost. She worked at a beach-goods store with damp conditions and was concerned her symptoms were due to occupational exposure. She denied chest pain, shortness of breath, fevers, chills, or recent travel.

On examination, she appeared much older than stated age. Her scalp was notable for diffuse alopecia and thinning hair. Her oral examination included striking macroglossia with a diffusely nodular and hemorrhagic tongue; she also had poor dentition. Her trunk and arms were notable for multiple diffusely scattered petechiae and purpura. Her axillae and inframammary regions had multiple whitish papules. Diffuse blistering was present on all extremities. The lower extremities were notable for extremely doughy, redundant skin with 3+ edema to the thighs giving an elephantiasis-like appearance.

Initial labs were notable for urinalysis showing >500mg/dL of protein, total serum protein 5.8 with total albumin 1.7, and a microcytic anemia. Serum immunofixation showed markedly elevated serum free light chains (1166 mg/L). Renal biopsy was performed and confirmed involvement by AL type amyloidosis with light chain restriction.

Discussion: Systemic amyloidosis is an uncommon plasma cell dyscrasia. Clinical manifestations are related to the underlying pathophysiology, which involves deposition of abnormal protein fibrils in affected tissues. Common organs of involvement include the kidneys, liver, skin, and other soft tissues. The most frequent form of primary systemic amyloidosis is AL amyloidosis, where immunoglobulin light chains are abnormally produced, as in our patient. Patients often present with an elevated serum globulin gap, and may present with renal failure or proteinuria.

Cutaneous manifestations of systemic amyloidosis are variable but offer important diagnostic clues. Our patient exhibited several characteristic cutaneous findings including diffuse alopecia and easy bruising (so called “pinch purpura” that can be induced by minor trauma). In addition, she had macroglossia, reflecting diffuse infiltration of the tongue by abnormal protein. Finally, her extreme lower extremity edema, proteinuria, and blistering reflect profound renal involvement of her advanced disease.

We present this case to highlight the importance of information gleaned from a thorough physical examination in making a diagnosis of amyloidosis.
Title: A Solitary Left Lower Extremity Mass: Diffuse Large B-Cell Lymphoma

Authors: Amir Yosef, MD. Emily Touloukian, MD. Vinod Nambudiri, MD.

Introduction: Diffuse large B cell Lymphoma is the most common lymphoid malignancy in adults. It is biologically and clinically heterogeneous with median age of onset in the 7th decade. Typically presents with fever, night sweats, and weight loss (B symptoms). The International prognostic index (IPI) helps with determination of prognosis and tailoring therapy. Cure rate varies from 20% in advanced disease with high IPI score, to 80% in localized disease with low IPI score (as in this case). R-CHOP chemotherapy has been the standard of treatment for years. This case highlights an uncommon presentation of a common disease.

Case Presentation: A 73 year old woman presented initially to the primary care clinic with a solitary mass on the shin of the left leg that had been progressively enlarging for several weeks. She was otherwise asymptomatic and denied weight loss, leg pain, fractures or history of DVT. She had no prior history of cancer or similar conditions and was up to date with her mammogram and colonoscopy. On review of systems, she denied anorexia and weight loss. On examination, no cervical, axillary or inguinal lymphadenopathy was present. The mass on the left shin was firm, measured 6x3 cm, and was non tender but mildly inflamed. The growth was initially evaluated with an ultrasound which demonstrated a 6 cm lesion with surrounding soft tissue edema. MRI of left lower leg showed multiple intramedullary lesions with a soft tissue component corresponding to the growth measuring 3.2x1.5x5.9 cm. A biopsy of the lesion was done and pathology was consistent with diffuse large B cell lymphoma with Bcl-6 expression and Ki67 expression greater than 90%. Subsequent CT scan of the chest, abdomen and pelvis demonstrated scattered non-pathologically enlarged lymph nodes in the abdomen and pelvis. PET scan indicated abnormal activity only in the leg mass without additional adenopathy activity. Bone marrow aspiration and biopsy were performed and showed no evidence of lymphoma and a normal female karyotype. Patient initially received 3 cycles of R-CHOP based chemotherapy. Follow-up PET scan showed interval resolution of the left proximal tibial mass. She subsequently received external beam involved field radiation therapy over 33 days. PET scan upon completion of therapy showed complete resolution with no evidence of recurrence throughout extended follow-up.

Discussion: Diffuse large B cell lymphoma is the most common lymphoid malignancy in adults. Approximately 20% of patients present with a localized lesion, and extranodal disease is less frequent. Despite the odd site of the mass and absence of B symptoms, the progressively enlarging character of the mass in an elderly patient raises the suspicion for malignancy. Diffuse large B cell lymphoma, leg type, is an uncommon variant of the disease with skin-limited findings. We present this case to highlight this rare presentation.

References
Title: “Don’t Ignore the Family History”

Authors: Sheikh MR, Khan MS, Klippenstein K, Desai C, Pham S.

Introduction: Arrhythmogenic Right Ventricular Cardiomyopathy is an inherited and potentially fatal cardiac condition that must be considered in otherwise healthy young adults.

Case Presentation: A 22-year-old, 25 week, pregnant female presented to the emergency department with three-day history of nausea, vomiting and palpitations. Vital signs revealed tachycardia; physical exam was within normal limits. Electrocardiogram (EKG) was performed and patient received IV push of adenosine. Heart rate did not slow and patient became hypotensive. Electrophysiology was consulted. Based on the EKG, monomorphic ventricular tachycardia was suspected. The patient was cardioverted and reverted to normal sinus rhythm with normalization of her blood pressure. Initial labs revealed significantly elevated troponin, elevated liver enzymes, proteinuria and thrombocytopenia.

Patient history was significant for no prenatal care. Obstetrics was consulted and diagnosed HELLP (hemolysis, elevated liver enzymes, low platelets) syndrome based on clinical picture and laboratory studies. The patient underwent emergent Cesarean-delivery. Subsequent comprehensive history revealed that the patient and her family had undergone genetic testing three years ago as part of a population screening study in the Hutterite colonies. The patient was found to be homozygous for known Hutterite variant for Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) (DSC2, p.Q554X). Both of her parents and her baby are carriers for the pathogenic variant, two of her siblings are homozygous for the trait; however, no family member is currently undergoing subsequent follow up for this condition.

The patient’s echocardiogram on admission revealed left ventricular ejection fraction of 40-45% and severe right ventricular dilation with moderate decrease in right ventricular systolic function. Cardiac MRI showed classic findings for ARVC and intracardiac defibrillator was placed.

Discussion: Arrhythmogenic right ventricular cardiomyopathy is characterized by progressive degeneration of the right myocardium leading to electrical instability. It has a strong genetic association and presents in the second to fifth decade of life. ARVC has variable presentation including atypical chest pain, palpitations, dizziness, syncope and sudden cardiac death. Our patient presented with monomorphic ventricular tachycardia, the most common arrhythmia associated with ARVC. This is a well-documented and often under recognized pathology. This case has great educational value as it provides classical transthoracic echocardiogram and EKG findings of ARVC. In addition, this condition and associated complications have been infrequently described in pregnant females. This patient’s clinical course was further complicated by HELLP syndrome, which upon review has not been previously associated with ARVC.
Tennessee-Clinical Vignette-Poster Finalist
Emmanuel Addo-Yobo, MD

Title: Syncope evaluation using point of care ultrasound

Authors: Emmanuel Addo-Yobo, MD, Jennifer Treece, MD MBA, Vandana Pai, MD, Mariam Alawoki, MD, Neha Kakkar, MD, Christian Rosero, MD, Shelby Shamas, DO, Department of Internal Medicine, East Tennessee State University, Johnson City, TN, Department of Internal Medicine, VA Mountain Home, Johnson City TN

Introduction: While syncope represents a common presentation of cardiovascular compromise, it is rarely the first presenting symptom of a pulmonary embolism (PE). If syncope occurs, delay in therapy can quickly become fatal. We describe a complex presentation of syncope in which bedside point-of-care echocardiogram would have directed appropriate management of a PE.

Case Presentation: Eighty seven year old male with no significant cardiac history presented with three syncopal episodes. There were no prodromal symptoms or evidence of seizure activity. Blood pressure of 76/42 mmHg and pulse of 118bpm were recorded by paramedics. Electrocardiogram (EKG) showed atrial fibrillation with rapid ventricular response. Upon arrival at our facility, he was hypoxemic and hypotensive. Labs showed serum troponin of 3.7 ng/mL and acute renal insufficiency. EKG showed no acute ischemic changes. Head imaging showed no acute intracranial pathology.

Acute coronary syndrome protocol with low intensity heparin drip was initiated. The patient refused cardiac catheterization and opted for medical management. A few days later he was transitioned to apixaban for stroke prophylaxis due to the new onset of atrial fibrillation. Transthoracic echocardiogram (TTE) on the fourth day showed characteristic signs of right heart strain. This prompted a computer tomography (CT) with pulmonary embolism (PE) protocol, which then showed a large central pulmonary embolus in the right lung with additional multifocal segmental and sub segmental PE throughout both lungs. Subsequent lower extremity ultrasound showed a non-occlusive deep vein thrombosis (DVT) in the left lower extremity. Treatment teams all agreed to continue the current management with apixaban since the patient was oxygenating well on nasal cannula and was hemodynamically stable.

Discussion: Syncope is an uncommon presentation for PE. Even when an alternative reason for syncope exists, it is estimated that PE is present in 12% of patients with syncope. Our patient presented with syncope in the setting of ACS, new onset atrial fibrillation and hypotension. His pretest probability for PE was low; a calculated wells' score of 1.5 for tachycardia. The echocardiography showed the Mcconnel sign, a finding which has a 94% specificity for a pulmonary embolus. The Mcconnel sign is a pattern of right ventricular dysfunction with akinesia of the mid wall and hyper contractility of the apical wall. If point-of-care ultrasonography (POCUS) was part of the initial evaluation, the patient’s presentation would have been appropriately managed for a hemodynamically unstable PE. This would have included consideration for thrombolytic therapy or embolectomy. Fortunately, this patient did well despite being treated for the confounding atrial fibrillation and non-ST segment myocardial infarction. In hindsight, application of POCUS would have made the biggest difference during his complex initial presentation. It would have shown evidence of right heart strain or the Mcconnel sign and this would have ultimately prevented the delay in treatment of an unstable pulmonary embolism.

References

Title: Complicated infective endocarditis limited to a Chiari network.

Authors: Olufolahan Carrena, Oluchukwu Oluoha, Amr Wahba, Maria Endsley, Henry Okafor.

Introduction: The Chiari network is an uncommon vestigial structure of the heart that is often clinically insignificant. We present a case of infective endocarditis affecting only the Chiari network in a patient who presented with septic emboli to lungs and brain.

Case Presentation: A 61-year-old man was evaluated for a 2-month history of hemoptysis and pleuritic chest pain. He also reported right upper extremity numbness and weakness. Vital signs were notable for a heart rate of 100/min. Physical examination was normal except for decreased sensation and motor strength (4/5) in the right upper extremity. Laboratory studies revealed leukocytosis and an elevated C-reactive protein. Chest radiography showed multifocal bilateral pulmonary opacities. Electrocardiogram was normal. Non-contrast head CT scan revealed a possible infarct of left parietal lobe. MRI of the brain, however suggested an abscess collection in the same anatomic location. Blood cultures grew Streptococcus intermedius. Transthoracic echocardiogram (TTE) was normal. Subsequent transesophageal echocardiogram (TEE) revealed an isolated 8.3X4.6 mm vegetation arising from a Chiari network, close to the right atrial appendage, without atrial or ventricular septal defects. He was treated with appropriate antibiotics with improvement of symptoms. Repeat chest imaging showed improvement of the lung opacities. He was transferred to an outside hospital for neurosurgical intervention for the brain abscess, and was later lost to follow up.

Discussion: The Chiari network is a mobile, fenestrated anatomic variant seen in the right atrium of about 2 percent of the population and has been described as an embryonic remnant of the right valve of the venous sinus. It is usually discovered incidentally at autopsy or during TEE for other indications and associated with patent foramen ovale, atrial septal aneurysms and recurrent arterial embolic events. Infective endocarditis affecting the Chiari network is rare and isolated involvement of the Chiari network, sparing the other heart valves is even more uncommon. Our patient was found on TEE, to have a vegetation attached solely to the Chiari network, sparing the valves on both the right and left sides of the heart. This is in contrast to many previously reported cases of infective endocarditis affecting a Chiari network, in which other heart valves were involved. Complications such as the lung and brain abscesses, from hematogenous spread from the primary focus of infection, have similarly been reported and are thought to be facilitated by atrial septal defects or other cardiac shunts. The lack of an anatomic demonstrable shunt however does not preclude hematogenous spread, as demonstrated in our index case.

Conclusion: In Chiari network endocarditis, a medical approach to treatment can be explored, while monitoring with imaging, the vegetation itself and any sites of complications from septic emboli, which may warrant a multidisciplinary approach, including surgical management.

References


Title: Fulminant Ehrlichiosis: An Unusual Cause of Myocarditis

Authors: Cherie N. Dahm, MD¹ and W. Chad Armstrong, MD²

Introduction: Ehrlichiosis is a tick-borne illness endemic to the southeast/south central United States, and when in humans, is an intracellular bacteria that grows within leukocytes. It causes a wide range of clinical manifestations from a low-grade febrile illness to multisystem organ failure.¹

Case Presentation: A previously healthy 41-year-old male presented to the Emergency Department with several days of high fevers up to 104°F, headache, nausea/vomiting, “weakness,” and a dry cough. When discussing exposures, he mentioned being an avid trail runner and removing non-attached ticks on his extremities but none recently. His vital signs were significant for temperature of 102.7°F. On initial exam, he was ill appearing in moderate distress with ongoing rigors. He had a faint petechial rash on his hard palate and lower extremities with conjunctival hemorrhages bilaterally but no splenomegaly, edema, rales, jugular venous distention, or extra heart sounds. His initial labs were notable for sodium 128, creatinine 1.34, aspartate aminotransferase 2244, alanine aminotransferase 1067, WBC 1.5, hematocrit 33, platelet count 37,000, partial thromboplastin time 59, lactate dehydrogenase 2960, haptoglobin 15, troponin 1.02, creatine phosphokinase 219, ferritin 57,356, and normal triglycerides. Electrocardiogram (EKG) was notable for T wave inversions in the inferior leads. He was immediately started on intravenous doxycycline for treatment of possible tick borne illness. Within his first day of admission, infectious disease, hematology, and cardiology were consulted. On hospital day 2, his troponin peaked at 1.6. He had a transthoracic echocardiogram completed demonstrating normal left ventricular systolic function (LVSF) with focal hypokinesis of mid-lateral wall frequently seen in myocarditis. Also on hospital day 2, he remained afebrile with downtrending liver enzymes, and ehrlichia PCR preliminary result was positive. Blood cultures were notable for streptococcus viridans treated with ceftriaxone and gentamicin. With progressive symptomatic and clinical improvement, MRI, myocardial biopsy, and bone marrow biopsy were not performed. On hospital day 4, further lab testing resulted with decreased natural killer cell function and elevated soluble interleukin-2 receptor confirming the diagnosis of secondary Hemophagocytic Lymphohistiocytosis (HLH) fulfilling 5 out of 8 diagnostic criteria. He was discharged home on hospital day 5 with plans to complete a 10-day course of doxycycline and a 14-day course of ceftriaxone and gentamicin. At his one-month follow up with Cardiology, a repeat EKG showed normal sinus rhythm without T wave inversions, and his repeat echocardiogram showed normal LVSF with normalization of previous wall motion abnormality. Most recently, he is feeling well and continuing to recover.

Discussion: This is an interesting case of Ehrlichiosis leading to myocarditis, pancytopenia, acute transaminitis, coagulopathy, and secondary HLH that with early recognition and antibiotic treatment in an otherwise healthy individual completely resolved.

References

Tennessee-Clinical Vignette-Poster Finalist
Christina A Kozlovski, MD

Title: Pseudohyperparathyroidism—Location Matters!

Authors: Sister Teresa Mary (Christina) Kozlovski, RSM, MD1, Aaron Bussey, MD1,2, 1University of Tennessee Graduate School of Medicine, 2Endocrinology Consultants of East Tennessee

Introduction: Primary hyperparathyroidism in the setting of multiple endocrine neoplasia (MEN) requires surgical intervention as definitive management. One surgical option is a total parathyroidectomy with forearm auto-transplantation of remnant tissue. Post-operative surveillance of parathyroid hormone (PTH) that reveals an elevated level in this population suggests disease persistence or recurrence. The following cases discuss an important but underrepresented alternative etiology for elevated PTH levels in patients with MEN and prior parathyroid surgery.1,2

Case Presentation: A 72-year-old female with MEN1 and primary hyperparathyroidism status post total parathyroidectomy with left forearm auto-transplantation followed with her endocrinologist for surveillance of disease activity. She was asymptomatic. PTH monitoring revealed levels from 76.6 to 5,751.1 pg/mL (reference range: 14-72 pg/mL) over several months despite normocalcemia. Given the inconsistency of her laboratory values and her clinical picture, her physician recognized a source for discrepancy and ordered bilateral upper extremity blood samples. PTH from her right arm measured 72 pg/mL, while PTH from her left arm, the side of the auto-transplantation, measured 884.5 pg/mL.

Similarly, a 69-year-old female with MEN2A and primary hyperparathyroidism status post total parathyroidectomy with left forearm autotransplantation was asymptomatic and underwent surveillance monitoring. Her PTH post-operatively was found to be 510.4 pg/mL, a specimen taken from her left arm. While she did have mild hypocalcemia, a same day sample drawn from her right arm revealed a PTH of 14.1 pg/mL.

Discussion: Patients with primary hyperparathyroidism in the setting of MEN who undergo parathyroidectomy require subsequent post-operative surveillance of PTH levels to monitor for disease persistence or recurrence. If PTH levels are elevated, further work-up and additional surgical interventions may be indicated. However, it is imperative to consider alternative explanations, particularly in an asymptomatic patient with normocalcemia. Our cases highlight the important lesson of blood sample location in patients after total parathyroidectomy with auto-transplantation. Fluctuations and significant elevations in the PTH levels simply reflect the proximity of the blood sample to the site of auto-transplantation of the remnant parathyroid tissue rather than active disease. In fact, a gradient of PTH greater than 1.5 between the ipsilateral and contralateral arm to auto-transplantation is associated with graft function and survival in the surgical literature.3 Physician awareness and patient education to ensure blood draws are performed on the contralateral side from autotransplantation can decrease costs and patient harm associated with unnecessary work-up and interventions.

References

Title: Desert Heat: Mystery Fever in a Traveler

Authors: Sagal Mohamed, MD, Celestine Wanjalla, MD, PhD, Sir Norman Melancon, MS, Matthew Greene, MD, Johnson Wong, MD.

Introduction: Q fever, a zoonotic infection caused by Coxiella burnetii through aerosolized particles or contaminated animal products, is an uncommon disease that can have a varied presentation, especially when acute. This case of a traveler with fever but otherwise nonspecific symptoms, who was diagnosed with acute Q fever through serological testing, highlights the importance of keeping a wide differential diagnosis when evaluating patients with perplexing symptoms and otherwise negative work-ups.

Case Presentation: The patient is a 51-year-old female civilian contractor working as a field engineer for the US military living on base in Afghanistan, who presented to the hospital after approximately 3 weeks of worsening fatigue and daily fevers for 1 week. She had a history of remote smoking and cholecystectomy with accidental splenectomy, status post vaccinations. Her first symptom was of extreme fatigue and feeling dehydrated despite increased fluid intake. As she was not authorized to leave the base, she did not venture outside of it. Prior travel was to UAE and Iraq. She presented to a civilian clinic after 2 weeks of fatigue and was hypotensive on arrival. After fluid resuscitation, she was transferred to a military hospital where a cardiologist suspected a large AAA on exam, confirmed on CT imaging at 6.7 cm diameter. She was flown to a hospital in Dubai for repair. While there, she had daily fevers prior to surgery and received broad-spectrum antibiotics. She was not aware of fever. Labs were notable for elevated CRP, leukocytosis, and transaminitis. Multiple sets of blood cultures were negative and a partial infectious and rheumatologic work-up was unrevealing. Fevers persisted after graft surgery despite antibiotics and she was discharged with Levaquin. She flew to Nashville and presented at the hospital the next day, febrile to 101.2 F. Physical exam and labs were otherwise unremarkable. PET scan showed no uptake at endograft material and low suspicion of post-operative infection. No antibiotics were administered. Blood cultures and multiple serologies were negative and fevers resolved, so she was discharged. Q fever titers returned significantly elevated and she was diagnosed with acute Q fever and treated with doxycycline.

Discussion: This case illustrates the complexity of evaluating a traveler with fever and the importance of conducting thorough questioning and examination and maintaining a wide differential. This patient had acute Q fever with a nonspecific presentation and no significant exposure history. It is also, despite having a wide reservoir, rare in travelers who present with fever. In addition, several case reports have discussed Q fever and rare vascular complications such as aortic aneurysm. It is unknown whether this was a long-standing aneurysm or an acute complication of Q fever, but if the latter, this would be a rare possible complication.

References

Title: A Rare Case of Aggressive Intravascular Large B cell lymphoma

Authors: Moka.N1, Balagoni.H1, Arikapudi.S1, Bokhari.A1, Shah.R2

Introduction: Intravascular Large B cell lymphoma (ILBL) is a subtype of extranodal Large B-cell lymphoma which is an extremely rare, aggressive and has poor prognosis. Incidence is one case per million. ILBL is rarely on the differential, since it is not well known. Most common symptoms are fever, fatigue, weight loss and symptoms based on the systems involved. Primary presentation in the lungs is highly uncommon. Herein, we present a unique case of ILBL presenting as pulmonary nodules.

Case Presentation: A 73-year-old non-smoker, diabetic male presented with recurrent dry cough, progressive dyspnea, intermittent episodes of fever, drenching night sweats and chills for 2 months. He denied any weight loss, sick contacts, recent travel to caves. Physical exam was unremarkable. Computed tomography (CT) chest showed vague 4-6mm nodular densities in right lung and mediastinal and hilar lymphadenopathy. Laboratory workup was remarkable for increased monocytes 14% (Normal: 0-8%) and elevated Angiotensin I-converting enzyme (ACE) at 81 (Normal: 9-67 U/L). The patient underwent endobronchial ultrasonography which was non-diagnostic. Subsequent video-assisted thoracoscopy with biopsy of the lung and mediastinal lymph node, revealed ILBL with the classic appearance of large malignant lymphocytes within the lumina of small and intermediate-sized vessels of right upper, middle and lower lobes, with positive CD20, CD79a, CD10, Pax-5, Mum-1. Oncological workup showed Stage-4 disease with involvement of spine and bone marrow. Cerebrospinal fluid (CSF) was negative for malignancy. The patient was given prophylactic intrathecal methotrexate and started on chemotherapy with rituximab, cyclophosphamide, hydroxydaunorubicin, vincristine, and prednisolone (R-CHOP). First cycle of chemotherapy was well tolerated, and the patient continues to follow with oncology.

Discussion: ILBL is most commonly seen in males with median age of 70 years. 91% of ILBL patients present at stage III or IV disease. Symptoms depends on the organ system involved. Two variants of clinical presentation are Western form, characterized by symptoms related to central nervous system (CNS) and skin. Asian variant, in which the patients present with multiorgan failure, hepatosplenomegaly, pancytopenia and hemophagocytic syndrome. Histopathology remains the gold standard for diagnosis. Staging is based on imaging, bone marrow biopsy and CSF analysis. R-CHOP plus CNS prophylaxis (PPX) is the treatment of choice. Initial treatment should include CNS prophylaxis, since CNS recurrence is very high. Agents used for CNS PPX are high dose methotrexate, high dose cytarabine. ILBL presents with variable and nonspecific symptoms, thus making it a challenging diagnosis. In our patient, Clinical and radiological findings were suggestive of sarcoidosis. However, the B-symptoms were concerning and the subsequent biopsy showed ILBL. Our case highlights the atypical presentation of ILBL as pulmonary nodules and hilar lymphadenopathy and the importance of history taking which led us to consider malignancy in the differential.

References

Title: A Qurious Case of the Heart: A Report of Q Fever Endocarditis

Authors: Johnson Wong MD, Monica Jimenez, MD, Sagal Mohamed MD, Peter Jelsma MD, Joy Gary DVM PhD, and Steven Embry MD.

Introduction: Endocarditis is not an uncommon diagnosis in the hospital setting, with 82% of cases secondary to Staphylococci and Streptococci species (Murdoch DR, 2009). Beyond these two species, there can be many other pathogens that can cause endocarditis including Enterococcus spp., HACEK organisms, various fungal species, and even Coxiella burnetii, a bacterium that causes the rare zoonotic infection Q fever. Q fever is a very uncommon cause of endocarditis and may be challenging to diagnose.

Case Presentation: The patient is a 59-year-old Caucasian male with past medical history significant for bovine aortic valve replacement in 2011, who initially presented to an outside hospital with crampy, progressively worsening right lower extremity pain and numbness located from distal thigh to foot. Right lower extremity pulses were absent. He also developed progressive numbness and a cool limb during this time frame. An aortogram with runoff showed evidence of occluded distal right external iliac and common femoral arteries. He underwent an urgent right iliofemoral embolectomy with successful restoration of blood flow. He was given a dose of ceftaroline, started on heparin and transferred to our facility for further care. Careful history taking revealed that the patient recently retired as a Kentucky state park manager. He was an avid hunter of deer and wild boar and frequently participated in the slaughtering process as well. Echocardiogram was performed because of suspicion of endocarditis and demonstrated a mobile lesion which prolapsed into the left ventricular outflow tract consistent with a vegetation. He was started on vancomycin and gentamicin and days later had a repeat TEE. The vegetation continued to enlarge despite antibiotics and a decision was made to perform an open aortic valve replacement by CT surgery. The valve and vegetation were sent to pathology for analysis. All fungal, bacterial, and viral cultures were all negative. Following multiple rounds of special stains, features suggestive of either Bartonella or Coxiella species emerged. Serology testing was initiated for both bacterial species were performed and revealed elevated titers to both. Results: were sent to the CDC who confirmed final diagnosis of Coxiella burnetii. Treatment of hydroxychloroquine and doxycycline was instituted. Duration of treatment, 1.5 years.

Discussion: This case demonstrates an uncommon pathogen of endocarditis. Q fever endocarditis is the most common manifestation of chronic Coxiella burnetii exposure, a disease caused by inhalation of aerosolized particles. Our patient’s risk factors of prosthetic valve and persistent exposure to aerosolized particles through hunting and slaughtering processes places him at high risk of contracting disease. Q fever can have a subclinical or asymptomatic presentation to some patients and diagnosis can be particularly challenging as highlighted in our patient’s hospital course. Treatment of Q fever endocarditis is particularly lengthy of at least two years in patients with prosthetic valves.

References


Title: Tugging on your Heart Strings: A Case of Primary Cardiac Angiosarcoma

Authors: Amna Ahmed MD, Ethan Burns MD, Anusha Sunkara MD

Introduction: Angiosarcoma is a rapidly proliferating, soft tissue sarcoma derived from anaplastic endothelial cells lining the blood vessel walls. Primary cardiac sarcoma is extremely aggressive, and is often overlooked due to its rarity. Despite ongoing efforts to improve mortality rates, prognosis of cardiac angiosarcoma remains poor, with a mean life expectancy of only a few months after diagnosis.

Case Presentation: The patient is a 23-year-old Chinese female with past medical history only recently significant for a positive home pregnancy test prior to admission, presented with a two week history of worsening nausea, vomiting, abdominal pain, and a three month history of infrequent sharp chest pain. Upon initial examination, patient was afebrile and hemodynamically stable. Physical exam was notable for tachycardia and muffled heart sounds, and generalized abdominal swelling and tenderness to palpation. EKG on admission showed sinus tachycardia with low voltage QRS. Labs were significant for elevated liver function tests, so an abdominal ultrasound was done, which demonstrated several masses on the liver. Further workup with an MRI demonstrated mild enhancement of these lesions on T2, as well as cardiomegaly. An echocardiogram was ordered, which showed ejection fraction of 60-65% and a large pericardial effusion with evidence of pericardial tamponade. She had placement of pericardial window with mediastinal drain as well as bilateral chest tubes and paracentesis. Diagnostic work up revealed no malignant cells from pericardial fluid with negative flow cytometry and negative pericardial biopsy. Echo five days after initial echo showed right ventricle systolic dysfunction that was moderately to severely depressed, with a reduced ejection fraction of 40-44%. Subsequent Cardiac MRI demonstrated a large, ill defined mass anterior and superior to the right atrium, isointense on T1, hyperintense on T2. PET/CT scan done showed increased uptake in the right atrium. Patient subsequently underwent right atrial cardiac biopsy that showed high grade cardiac angiosarcoma, and liver biopsy showing anastomosing hemangiomas. The patient had elective termination of pregnancy and was started on Adriamycin and Ifosfamide chemotherapy.

Discussion: Primary cardiac angiosarcoma is a rare tumor with an extremely poor prognosis. It most often presents in males in the 3rd to 4th decade of life, on the right side of the heart, and often mimics other pathologies. Patients may manifest through symptoms related to obstruction, local invasion, or embolic phenomena, and are typically found in advanced stages. Cardiac MRI and echocardiogram have become the mainstay in detecting this malignancy, and these patients must often be evaluated for distant metastases. The prognosis remains poor, with estimate survival approaching 3.8 months in those without surgical resection. Clinicians should maintain a low threshold of suspicion for this condition, since an early diagnosis can prolong survival.
Title: Neurocysticercosis Induced Subarachnoid Hemorrhage Without Aneurysm or Vasculitis

Authors: Hema Bohra, PGY2 Internal Medicine, Medical City Fort Worth, Syed Tariq Ahsan, PGY3 Internal Medicine, Medical City Fort Worth, Dr. Frood Eelani DO, Internal Medicine Attending, Dr. Dr. Ryan Gianatasio MD, Neurology Attending, Texas Stroke Institute, Dr. Machaiah Madhira MD, Program Director, Medical City Fort Worth

Introduction: Increasing globalization facilitates travel of disease from underdeveloped areas. Often attributed to eating undercooked pork, the zoonotic vector *Taenia solium* also infects travelers to endemic areas who consume contaminated food or water. The ingested eggs cause havoc in muscles, skin, eyes and brain as they grow and develop into larvae. Neurocysticercosis effects functions of the central nervous system as a result of the immunologic process that accompanies cyst degeneration. Sequelae include seizures, intracranial hypertension, hydrocephalus and inflammatory peri-lesional edema that can manifest as infarcts causing ischemic or hemorrhagic events.

Case Presentation: A 45-year-old Hispanic female arrived from an outside facility after falling and hitting her head while arising from the bathroom commode. This occurred after one day of nausea and emesis concomitant with temporary left leg weakness, following several days of progressive bilateral headache. Past medical history significant for hypertension, GERD and increasing urinary incontinence for one month. Patient emigrated from Mexico in 1999, but visits often; her last trip was in July 2017. Patient denied any changes in vision, vertigo or ringing in her ears, speech or memory issues, fevers or cough. CT imaging noted subarachnoid hemorrhage, obstructive hydrocephalus, and multiple intracranial lesions concerning for neurocysticercosis. MRI described enhancing cystic lesions in the basilar cisterns, left frontal lobe and left basal ganglia consistent with neurocysticercosis and further described the subarachnoid hemorrhage as an extension of neurocysticercosis into the posterior fossa. Four vessel cerebral angiogram was negative for vasculitis, aneurysm and arteriovenous fisulas or malformations. Placement of a ventriculostomy catheter was done to reduce increasing cerebrospinal fluid pressure. Treatment with Albendazole and Praziquantel was started for an extended 28-day course in addition to dexamethasone. Four days after admission, patient noted increased headaches and a temperature of 100.0. Patient was empirically started on vancomycin and cefepime until cerebrospinal fluid gram stain returned negative for bacterial causes. Symptoms were attributed to placement of external ventricular drain. Transcranial doppler noted vasospasm on day six of admission. Nimodipine had been initiated after day one of admission. Patient was without any new neuronal deficits and continues to improve with physical therapy.

Discussion: Most presentations of intracranial hemorrhage resulting from neurocysticercosis are secondary to aneurysm rupture, inflammatory arteriopathy or subarachnoid hemorrhage, often with evidence of dense inflammation with thickened-leptomeninges. Our patient manifested a very unusual presentation of subarachnoid hemorrhage in the posterior fossa, without evidence of prior aneurysmal or vasculitic events. Pathogenesis theories denote the large parasite burden perpetrating an inflammatory cascade, leading to the erosion of a degenerating cyst into neurovascular causing endarteritis and endothelial hyperplasia that Result: with aneurysm. This leads to the conclusion that cysticercosis manifests in different types of intracranial hemorrhage that need further research. When evaluating young patients without vascular risk factors but with travel to endemic areas, neurocysticercosis should be a differential.
References

Texas-Clinical Vignette-Poster Finalist
Elizabeth C Brewer, MD

Title: Acute Liver Failure from Disseminated Varicella Zoster Infection

Authors: Elizabeth Caitlin Brewer, MD, Internal Medicine PGY-3, Methodist Hospitals of Dallas; Leigh Hunter, MD, FACP, Program Director Internal Medicine Residency and Hospitalist Programs, Methodist Hospitals of Dallas

Introduction: Numerous causes of acute liver failure (ALF) are reported with the most common being acetaminophen toxicity and viral infections. We present the rare case of Varicella zoster virus (VZV) induced ALF with review of the literature.

Case Presentation: A 66-year-old Caucasian woman with past medical history of dermatomyositis, dysphagia, gastro-esophageal reflux and hypertension presented to the emergency department with several days of mid-epigastric, constant, non-radiating abdominal pain and rash that began on her face and chest, then spread to her arms and abdomen. She also reported white “spots” in her mouth, nausea, constipation, and worsening dysphagia. Her medications included prednisone, mycophenolate, trimethoprim/sulfamethoxazole, and nystatin.

Physical examination revealed an alert and oriented patient with normal vital signs. Significant findings included oral thrush with petechial, macular, and vesicular skin lesions on face, trunk and extremities. Significant laboratory data included: AST 1389 U/L, ALT 1570 U/L, alkaline phosphatase 68 U/L, INR 1.6 and PT 18 seconds. Skin biopsy performed 2 days prior to admission showed multinucleated giant cells with viral inclusions suggestive of “herpes virus” infection. The patient was initiated on IV acyclovir, micafungin, vancomycin, aztreonam and solumedrol. Subsequently, AST and ALT increased to the 4000s, INR to 1.8 and PT to 20.6. Due to worsening ALF, she was transferred to our facility for liver transplant evaluation.

Upon transfer, her skin lesions were consistent with VZV, which skin biopsy cultures later confirmed. Liver biopsy revealed hepatic necrosis with positive VZV PCR and blood PCR also showed high levels of VZV. Laboratory evaluation for other causes of ALF was negative. Her hospital stay was complicated by multidrug resistant Enterobacter pneumonia and bacteremia, respiratory failure requiring intubation and multiple organ failure. VZV PCR copies decreased with treatment, but her severity of illness and active infection prevented liver transplantation. The patient’s code status was eventually changed to “do not resuscitate” and she expired.

Discussion: Reactivation of VZV as “shingles” is common, but ALF due to VZV is exceedingly rare with high mortality. The differential diagnosis of ALF includes ischemia, venous obstruction, medications, toxins, autoimmune hepatitis, metabolic and infectious causes. In the setting of VZV hepatitis, definitive diagnosis is made by liver biopsy, histopathology, culture and VZV PCR.

In review of the literature, only 8 adult cases of ALF from VZV were found of which only 2 survived. In these cases, IV acyclovir was the staple of treatment. Other therapies include VZV immune globulin, liver transplant, IVIG and supportive care. Since ALF due to VZV has such high mortality rates and early treatment is critical to survival, VZV should be considered in the differential diagnosis of all patients with ALF who present with skin lesions.
Texas-Clinical Vignette-Poster Finalist
Hira I Cheema, MBBS

Title: Hide and Seek

Authors: Hira Cheema MD. PGY3. TTUHSC at The Permian Basin.

Faculty advisor: Kalyan Chakrala MD.

Introduction: Aortic stenosis, anemia and angiodysplasia related GI bleed are all very common presentations, especially in the elderly. It is very important to put them together in light of Heyde’s syndrome. A prompt diagnosis will help us treat the patient appropriately which is by aortic valve replacement.

Case Presentation: We report a 76-year-old Caucasian male with medical history of chronic obstructive pulmonary disease (COPD) on home oxygen 2L, hypertension, carotid stenosis, and SLE (not on steroids). He was first referred for evaluation of multiple episodes of anemia that required monthly blood transfusions. At time of referral, patient denied usage of ASA, NSAIDs or blood thinners. EGD and colonoscopy done 2 weeks prior to time of referral were negative for source of bleeding. Labs showed hemoglobin 8.7, hematocrit 26.9, RDW 16.4, platelets 237,000. Patient underwent second look EGD and colonoscopy and was found to have several actively bleeding arteriovenous malformations (AVM) in stomach and second part of duodenum. The bleeding AVMs were successfully coagulated with argon plasma coagulation (APC).

A month later, patient was admitted to the hospital due to sudden onset of shortness of breath. During this hospital stay, patient had been hemodynamically stable (HGB 8.0) and required no transfusion. Repeat EGD again showed actively bleeding AVMs that were again coagulated with APC. Decision was made to start the patient on IV octreotide treatment.

Patient was again admitted to the hospital for hypoxia and shortness of breath secondary to GI bleed after one month. Laboratory showed HGB 6.7, Hct 21.5. Physical exam revealed grade 3 systolic murmur best heard at right second intercostal space. Echocardiogram showed severe aortic stenosis (AS) with valve area 0.92 cm². A diagnosis of Heyde’s syndrome was made. Patient subsequently underwent bioprosthetic valve replacement. Patient also received 2 drug-eluting stents for coronary artery disease and was started on anti-platelet therapy (Plavix).

Over the course of 9 months as outpatient follow up patient reported lesser events of GI bleed. From March-July 2016 his HGB improved form 6.6 to 10.7. In the next 5 months his HGB improved steadily from 10-12 with minimal to no bleed in stools. He did not require repeat endoscopy over the next one year as outpatient follow up.

Discussion: Heyde’s syndrome is defined as a triad of aortic stenosis, acquired coagulopathy (vWS type 2A) and anemia due to angiodysplasia related bleed. When blood passes through a stenosed aortic valve it leads to mechanical disruption of vWS. The enzyme ADAMTS13 then attacks these smaller multimers. These further cleaved multimers are unable to bind to exposed collagen in bleeding arterioles and hence
clot does not form. Aortic valve replacement cures the acquired coagulopathy and remains treatment of choice.
Texas-Clinical Vignette-Poster Finalist
Jerry Fan, MD

Title: To Close or Not to Close? A Study of Moving Emboli through a PFO Causing MI, Stroke, and PE

Authors: Jerry Fan¹, MD, Denisse Sangha¹, MD, Christopher Chiles¹, MD, Baylor Scott and White Health System, Temple Memorial Hospital

Introduction: A 75-year-old female that was diagnosed with multiple paradoxical emboli after presenting with deep vein thrombosis, bilateral pulmonary emboli, right MCA stroke, and RCA/LAD occlusion due to a stretched patent foramen ovale.

Case Presentation: A 75-year-old generally healthy female presented with acute left sided paralysis and chest pain that was preceded by left leg pain and swelling. The patient was initially evaluated in ED with imaging that revealed right MCA infarct. Simultaneously she developed acute hypoxia and CTA showed bilateral pulmonary emboli. The initial EKG showed anterior injury with troponins 0.02, then 66.88. She underwent tPA administration and neuro-interventional radiology directed tPA and eptifibatide which resolved her focal neurological symptoms. After tPA, her chest pain, ST-segment elevation, and troponins also resolved. A TTE revealed large right-to-left shunt suggesting patent foramen ovale and an atrial septal aneurysm. Lower extremity ultrasound revealed a popliteal vein thrombosis. Her hospital course was complicated by a large right intramuscular thigh hematoma and hemarthrosis which caused a dramatic drop in hemoglobin requiring transfusion. An IVC filter was placed to prevent further embolic transit. Cardiac catheterization showed no significant coronary artery disease. The patient’s PFO was closed and life-long anticoagulation was started. Before discharge, an IVC venography demonstrated moderate sized DVT in the IVC filter and it was left in place.

Discussion: Patent foramen ovale is common, occurring in approximately 27% of the general population, but most do not cause problems, yet there is significant morbidity and mortality that can occur from transiting emboli. Paradoxical embolism presents a challenging differential diagnosis and often confusing initial presentation since it requires a right-to-left shunt leading a transiting thrombus into systemic. This case, represents the only report of simultaneous pulmonary embolism, and arterial embolism due to a DVT traveling across a PFO.

References

Bibliography


Texas-Clinical Vignette-Poster Finalist
Olubadewa Adeyemi Fatunde, MD

Title: Integrated coronary revascularization in a patient with an Inferior STEMI from a LCX lesion

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Introduction: Inferior wall ST-segment Elevation Myocardial Infarction (STEMI) is most commonly localized to the right coronary artery (RCA). In approximately 40% of individuals, however, the left circumflex artery (LCX) supplies the sinoatrial nodal artery and in an even smaller minority (10-20%), the atrioventricular nodal artery in a left dominant heart. Left dominance occurs in approximately 8% of individuals and is associated with higher in-hospital mortality.

Integrated or hybrid coronary revascularization refers to utilizing a combination of percutaneous coronary intervention (PCI) and coronary artery bypass grafting (CABG) to restore perfusion to a heart with critical multivessel disease, while optimizing the benefits of both procedures.

Case Presentation: We report a case of a 62 year old Caucasian male with a past medical history of hypothyroidism, hypertension and hyperlipidemia complaining of chest pain approximately 35 minutes prior to presentation. He experienced crescendo angina in the preceding two weeks. His electrocardiogram showed evidence of an inferior wall STEMI with 0.5 to 1mm STEMI, and he was taken to the catheterization lab. Immediately prior to procedure, patient became unresponsive, developing ventricular fibrillation. He responded appropriately to two rounds of electrical cardioversion, CPR, and atropine. Images showed 70% occlusion of the acute marginal branch arising from the proximal portion of the LCX, 90% stenosis of the proximal LAD at the bifurcation of a large diagonal branch and 100% occlusion of the LCX (midway). After multiple passes, manual thrombectomy was performed of the LCX lesion, with cath images confirming restoration of flow. Of note, patient had a dominant LCX, giving branches to the PDA and PLV.

It is well documented that the longest lasting intervention providing the best survival benefit in a critical LAD lesion is to bypass the LAD with a left internal mammary artery (LIMA) graft. After consultation with CV surgery, bare metal stents (BMS) were placed in the proximal and mid LCX, in addition to inserting an intraaortic ballon pump to improve perfusion of the critical LAD lesion. Patient recovered well from the LCX stent and was optimized on medical therapy. Four weeks later, he received a LIMA to LAD graft for his critical LAD lesion.

Discussion: This case underscores an uncommon, but important case of inferior wall MI secondary to a LCX lesion, as well as ideal patient selection for hybrid revascularization. Inferior STEMI secondary to LCX lesions, while relatively infrequent, is more commonly found in left dominant patients. Studies show that these patients have a worse prognosis compared to an RCA-related infarction, possibly due to imbalanced ischemia and greater shear stress. Selection for hybrid revascularization or CABG depends primarily on patient characteristics. The 30 day rate of major adverse cardiac & cerebrovascular events (MACCE) is comparable for both procedures in similar patients.
Texas-Clinical Vignette-Poster Finalist
Brian A Karahalios, MD

Title: A Case of Letting Sleeping Dogs Lie

Authors: Courtney Hatcher, Brian Karahalios, Manjulatha Badam

Introduction: Rocky Mountain Spotted Fever (RMSF) is associated with high mortality and requires prompt identification and treatment to ensure better outcomes.

Case Presentation: A 45-year-old woman from Kingwood, Texas presented to the ER with a 7-day history of fevers associated with headache, arthralgias, nausea, fatigue and neck pain. On exam she was tachycardic but afebrile. Labwork was WNL and CT Head was unremarkable. Clinical diagnosis of viral illness was made and she was discharged. Two days later, she presented to our ER with worsening confusion, combativeness, dyspnea and ataxia. The patient’s spouse mentioned that the patient had sustained multiple recent fleabites from her pet dogs sleeping in the bed. The patient was febrile, tachycardic and hypotensive. A maculopapular rash was noted on her abdomen. No bites or ticks were noted on the patient. The patient was admitted to the ICU for further management. MRI of her brain was unremarkable and an LP showed 16 WBC/HPF, 49mg/dL protein, 51mg/dL glucose, 32.91mg/dL albumin and 4 RBC/HPF. The patient was started on vancomycin, meropenem, acyclovir and doxycycline for suspected meningitis. Despite these interventions, her condition worsened and she required intubation for airway protection. A full infectious workup was performed. Positive findings were R. typhi IgM 1:1024 as well as R. Rickettsii IgM 1:1024 and IgG 1:128. The titers were repeated for possible cross-reactivity and R. typhi antibodies were noted to be negative. Antibiotics were de-escalated to doxycycline alone for the remainder of her hospitalization with clinical improvement. Fever defervesced and the patient was able to be extubated. She was discharged home with two more weeks of doxycycline after 7 days.

Discussion: The case illustrates that despite living in an urban setting, exposure to domestic dogs can increase the chance of acquiring RMSF outside the usual geographic regions of distribution. The highest incidence is noted in Oklahoma, Arkansas, Missouri, Tennessee and Virginia. The reported incidence in Texas was 2.3–6.6 cases per million in Texas in 2014. Patients become symptomatic 2–14 days post exposure. Classic presentation is febrile illness with headache, rash and a history of tick bite. A rash is seen in 90% of cases, but the classic involvement of the palms and soles is noted much later in the disease. Major complications include encephalitis, non-cardiogenic pulmonary edema, ARDS, coagulopathy, and seizures. Early treatment is key as fatality rates reach 30% in some areas if left untreated. The case illustrates the importance of keeping RMSF in the differential in patient populations outside of the usual geographic areas of incidence.

References

Texas-Clinical Vignette-Poster Finalist
Farah Ladak, MD

Title: Something’s Fishy: Manifestations of a Deadly Bacteria in a Middle-Aged Liver Patient

Authors: Farah Ladak, MD- UT Health Science Center San Antonio, Cynthia Cantu, DO- UT Health Science Center San Antonio, Natascha Tuznik, DO, FACP- UT Health Science Center San Antonio, Christopher Moreau, BSBME- UT Health Science Center San Antonio

Introduction: *Vibrio vulnificus* is a gram negative bacillus typically found in coastal environments such as the Gulf of Mexico. Consumption of raw or undercooked shellfish, handling of contaminated seafood or exposure of open wounds to water in which the organism lives may result in *Vibriosis*. Manifestations range from mild gastroenteritis to primary septicemia which has a mortality rate of approximately 50%. Patients with underlying chronic illnesses such as chronic liver disease, hemochromatosis, AIDS, malignancy or any immunocompromised state are particularly vulnerable to *Vibrio vulnificus* infections. These patients should be promptly educated on the dangers of raw seafood consumption upon diagnosis.

Case Presentation: A 61 year old male with a history of alcoholic cirrhosis, hypothyroidism, and hypertension was transferred from an outside hospital for a pre-transplant evaluation and complaint of dyspnea on exertion, bilateral lower extremity edema and worsening abdominal distension. His vital signs were within normal limits and findings on physical exam included diffuse anasarca with a non-tender distended abdomen. His initial laboratory Results: were notable for a sodium of 122, creatinine of 1.30, mildly elevated alkaline phosphatase and aspartate aminotransferase, total bilirubin of 6.3 and leukocytosis of 34,000. His bedside paracentesis fluid tested negative for spontaneous bacterial peritonitis. A right upper quadrant ultrasound showed significant cirrhosis and chest X-ray revealed a possible infiltrate. The patient was given Cefepime for possible hospital-acquired pneumonia. The following day, the admission blood culture grew gram-negative rods and the patient abruptly developed a non-tender, erythematous maculopapular rash extending from his right buttock to his right lateral thigh. He became hypotensive and was unresponsive to fluid resuscitation though other vital signs remained normal. Repeat labs were stable and lactate was normal. Broad-spectrum antibiotics were started. Both a CT of his right lower extremity and an emergent exploratory dissection showed no necrotizing fasciitis. His admission blood culture returned positive for *Vibrio vulnificus*. Upon further questioning, the patient recalled consuming large portions of raw oysters in the last week. He was treated with Ceftriaxone and Doxycycline for fourteen days and his rash improved significantly throughout his course. He was discharged with strict counseling regarding raw seafood and referred for hepatology follow-up.

Discussion: This patient was unaware of the dangers of consuming raw seafood given his underlying cirrhosis. This case highlights the importance of educating patients with chronic liver disease or any immunocompromised state on avoiding consumption and handling of raw seafood or swimming in *Vibrio* infested waters. It also illustrates the importance of prompt imaging and surgical evaluation for necrotizing fasciitis. While this patient had a favorable outcome, delayed differential diagnosis could result in significant morbidity and possibly death. Clinicians should be aware of the clinical manifestations associated with *Vibrio vulnificus* infections and should educate vulnerable patients to avoid raw seafood consumption.
Texas-Clinical Vignette-Poster Finalist
Passisd Laoveeravat, MD

Title: Unusual presentation of perforated gastric ulcer with ST-segment elevation

Authors: Passisd Laoveeravat, MD; Wasawat Vutthikraivit, MD; Subhanudh Thavaraputta, MD; Pavida Pachariyanon, MD.

Introduction: Perforated gastric ulcer is a common surgical problem which mostly presents with acute abdominal pain and an abnormal x-ray. We report an uncommon case of a perforated gastric ulcer presented with ST-segment elevation with documented normal coronary arteries. The ST-segment elevation reverted to normal after the surgery.

Case Presentation: A 78-year-old man with no past medical history presented to the hospital with sudden epigastric pain for 3 hours. His vital signs were within normal limits. An electrocardiogram and it disclosed ST-segment elevation in leads II, III, aVF, V2, and V3, consistent an acute anterior and inferior myocardial infarction (MI). Auscultation of the chest and heart showed normal breath sounds and no cardiac murmurs. There was no jugular vein distension and pitting edema. Examination of abdomen disclosed hypoactive bowel sounds, generalized abdominal guarding, and rebound tenderness. Cardiac markers test revealed troponin-T less than 0.01. The remaining tests, including complete blood count, electrolyte, blood urea nitrogen, and creatinine, were within normal limits. Transthoracic echocardiography revealed normal wall motion with an ejection fraction of 74%. No vegetations or valvular disease were noted. An acute abdomen series was performed for a suspected acute surgical condition. An upright abdominal x-ray showed free air under both hemidiaphragms. He underwent emergency exploratory laparotomy. A perforated gastric ulcer 1 cm in diameter was found. Simple suture and placement of drainage were performed. The follow-up 12-lead electrocardiography performed in the 24 hours after the operation revealed normalization of ST-segments in lead II, III, aVF, V2, and V3. CTA coronary obtained after the patient stabilized revealed no significant stenosis of coronary arteries. Repeat cardiac marker remained normal. There was no complication during the postoperative course, and the patient was discharged 7 days later.

Discussion: In conclusion, an acute abdomen is one of the non-cardiac causes associated with pseudo-myocardial infarction, but it is rare. Perforated gastric ulcer should be recognized as a rare presentation mimicking ST-segment elevation MI. It is vital for the clinicians to be aware of acute abdominal condition as one of the causes of ECG change because a delay in diagnosis will lead to serious complications.
Texas-Clinical Vignette-Poster Finalist
Luna Liu, DO

Title: A Rare Case of Dyspnea on Exertion Due to Left Atrial (LA) Sarcoma

Authors: Luna Liu, D.O., Aman Patel, D.O., Ahsan Khan, D.O., Attending: Giridhar Mundluru, M.D.

Introduction: Primary cardiac sarcomas are extremely rare (frequency rate of 0.001% to 0.03%) and usually malignant with a very poor one-year prognosis due to their limited response to chemo-radiation, high recurrence rate, and propensity to metastasize. Here, we report a rare case of this deadly tumor in a healthy elderly patient presenting as an insidious onset dyspnea on exertion. 1-2

Case Presentation: A 70-year-old caucasian female with a PMHx of HLD, hypothyroidism, and Factor V Leiden presented with progressive SOB and cough for 2 weeks. She was treated for a sinus infection prior but continued to have worsening SOB with minimal exertion. Prior to this, she was treated for a sinus infection but continued to have worsening SOB with minimal exertion. CT-chest and TTE demonstrated a 5 cm left atrial mass partially obstructing the LV and flow through the mitral valve. Cardiothoracic surgery was consulted for further evaluation. Surgical approach revealed a large myxoid mass with attachments to the LA wall and septum. Complete resection of mass and attachments with subsequent reconstruction was successful. Pathology confirmed the diagnosis of sarcoma. Following resection, the patient’s symptoms improved with minimal postoperative complications. The patient was discharged on POD#7 to an acute inpatient rehabilitation facility for further strengthening and conditioning.

Discussion: As the majority of cardiac tumors are the result of metastases from elsewhere, primary cardiac tumors are uncommon and likely (~75%) to be myxomatous tumors. Primary cardiac sarcomas are scarce and constitute a very poor prognosis as they present late in the course of the disease due to its early asymptomatic nature. 1-5 Per Hsieh et al., clinical presentations vary depending on the size and location of the tumor. Although commonly found in the upper chambers (left > right), primary cardiac sarcomas can develop in any cardiac chamber, from the cardiac wall, pericardial surface or even valvular surface. Because cardiac sarcomas can mimic myxoma, tissue biopsy is essential in proper management of its malignant nature. 6-7

This case underlines the importance of considering rare causes of sudden onset CHF. Furthermore, it stresses the necessity of tissue biopsy in the management of cardiac tumors as cardiac sarcomas have a poor prognosis. However, with early complete resection and adjuvant chemotherapy, the patient can overcome the poor prognosis and maintain a good quality of life. 5-8

In short, primary metastatic cardiac sarcomas have a poor survival rate due to their variable and nonspecific presentations leading to delay in diagnosis. Echocardiography is advantageous in early detection and thus improving morbidity. Due to its poor response to chemo-radiation, palliative surgical excision as well as a referral to a tertiary center for cardiac transplant may help in improving prognosis. 1-7

References


Texas-Clinical Vignette-Poster Finalist
Juan J Menjivar, MD

Title: “Keep your eyes wide open when your patient's eyes are wide shut”.

Authors: Juan J. Menjivar MD, Carlos A. Paris MD, Bilal Nazif DO, Rex Huang MD, Juan Castano MD.

Introduction: Horner's syndrome is a well known neurological syndrome that includes signs of ptosis, miosis, and anhidrosis. It is also called oculosympathetic paresis and may be caused by a lesion anywhere in the sympathetic pathway which supplies the head, eye, and neck. Lesions affecting the neck most commonly compromise the second and/or third cranial nerves. The syndrome is usually unilateral and there are few reports of bilateral involvement. Here we present a case of Bilateral Horner's Syndrome secondary to bilateral lymph nodes metastasis due to Squamous Cell Carcinoma of the lower esophagus.

Case Presentation: A 48 year-old male and former smoker of 1 pack of cigarettes a day for 20 years with no other medical history developed symptoms of dysphagia to solids, odynophagia and 25 lbs. weight loss 6 months before he consulted a gastroenterologist. On physical exam patient was found to be cachectic. He had an esophagogastroduodenoscopy (EGD) performed which showed an obstructive mass occluding the lower esophagus. Biopsy of the mass revealed Squamous Cell Carcinoma. He began chemotherapy however his dysphagia progressed to liquids, and he was referred for percutaneous endoscopic gastrostomy (PEG) tube placement. During his hospitalization he developed new onset bilateral weakness of both eyelids (left greater than right) associated with diplopia. On physical exam he had 2 mm pupils bilaterally with pupillary dilation lag, ptosis (left greater than right) and diffuse bilateral adenopathy in the neck in segments II-III-IV and V. A CT of the neck showed the marked enlarged lymph nodes compressing the carotid arteries and sympathetic trunk just posterior to the carotid sheath. The patient was subsequently discharged home after PEG tube placement; he passed away several months later from his underlying disease.

Discussion: This case illustrates the rare presentation of bilateral Horner's Syndrome secondary to esophageal metastasis. The level of the lesion belongs to a third order syndrome or postganglionic which indicates lesions of the internal carotid artery such as an arterial dissection, thrombosis, or cavernous sinus aneurysm. Other cases are related to malignancy such as in our patient, as we confirmed the presence of neck masses on CT of the neck. It is important to always have a broad differential on bilateral ptosis patients. The examiner always has to have a high index of clinical suspicion for diagnosis, the confirmatory test is the use of pharmacological agents and also imaging studies. This case challenges the way we learned this syndrome in medical school, and due to the fact that is difficult to find it bilaterally, clinicians should be aware of this clinical presentation.

References

Texas-Clinical Vignette-Poster Finalist
Whitney Sharp, DO

Title: Loperamide Toxicity Mimicking Brugada Syndrome

Authors: Whitney Sharp, D.O., Houston Methodist Hospital

Introduction: Loperamide, an over the counter anti-diarrheal medication, targets opiate receptors in the gastrointestinal tract with poor blood brain barrier penetration. However, analgesic or euphoric effects are produced in supra-therapeutic doses. Due to increasing incidence of occult loperamide abuse, clinicians need to be aware of its potentially fatal cardiotoxicity. Adverse cardiac effects of loperamide include widening of the QRS interval, prolongation of the QTc and dysrhythmias, with a black box warning in place for torsades de pointes and sudden cardiac death in higher than recommended doses.

Case Presentation: A 49 year old female presented to the emergency department with complaints of generalized weakness, numbness and tingling in her arms and legs, multiple pre-syncopal episodes and constipation. She reported symptoms for several months that had progressed to the point of impairing ambulation, which led her to seek medical attention. Further history revealed occult loperamide abuse with the patient ingesting up to 40 tablets daily that had resulted in a small bowel obstruction two years prior to presentation. Electrocardiogram (ECG) obtained in the emergency department demonstrated multiple abnormalities including prolonged QTc of 584 ms, right bundle branch block, widening of the QRS complex to 126 ms, and first degree AV block. Initial electrolyte studies including calcium, phosphorus and magnesium were normal. On admission, the patient had extensive neurological evaluation including neuroimaging with magnetic resonance imaging of the brain which was negative for acute pathology. She also underwent electromyogram and nerve conduction studies which showing a mild sensory neuropathy in the lower extremities without evidence of myopathy. Echocardiogram revealed no structural or wall motion abnormalities with preserved ejection fraction. Electrophysiology was consulted over concern for Brugada pattern seen on ECG and recommended placement of an implantable cardioverter defibrillator for Brugada syndrome, which patient refused. Serial ECGs throughout her hospital stay demonstrated gradual improvement of her QTc interval and narrowing of her QRS complex following the withholding of loperamide. Loperamide levels in the serum were sent and eventually resulted 14 times the therapeutic level.

Discussion: As a result of the opiate crisis, loperamide abuse has been seen with increasing frequency in recent years with the majority of the adverse effects seen on the gastrointestinal tract and cardiac conduction system. This case underscores the importance of taking a thorough history and the need for clinicians to recognize reversible cardiac toxicity of loperamide in a patient with unexplained conduction abnormalities and any history of substance abuse, as an occult opiate abuse disorder may be present and require concomitant treatment.
Texas-Clinical Vignette-Poster Finalist
Heidi Torres, MD

Title: When the Timeline is Wrong: A Missed Early Post – MI Ventricular Septal Defect

Authors: Heidi Torres MD, Matt Lelegren MSIV, Oscar Garza Ovalle MD, Paula Montana De La Cadena MD, Rushit Kanakia, MD

Introduction: Infarct related VSDs formerly occurred in 1% to 2% of patients in the prethrombolytic era. The incidence has decreased dramatically to 0.2% with modern reperfusion. Our knowledge of structural MI complications often follows a classic timeline where VSDs occur 1-2 weeks post MI. Though it is known structural complications may present earlier, our often strict adherence to this timeline may cause us to dismiss the manifestations when encountering them in an unexpected part of the disease process.

Case Presentation: A 59 year old man with a longstanding history of DM type II who presented to the emergency department with a two day history of chest pain, nausea, diaphoresis, and dyspnea on exertion. On initial physical exam a 3/6 holosystolic murmur was heard at the mitral area and left lower sternal border, which was diagnosed as a flow murmur by multiple physicians. He was diagnosed with DKA and NSTEMI with troponin I peaking at 5.3 ng/mL, glucose >350 mg/dL, and significant ketosis, and ketonuria. Transthoracic echocardiogram report identified a normal ejection fraction and mild mitral regurgitation but no other structural cardiac abnormalities were noted. DKA was first treated and once resolved patient underwent LHC. Obstructive coronary artery disease was found in his RCA with subsequent stent placement. He was discharged on optimized medical therapy. Three days later he returned to the ER with worsening shortness of breath. On physical exam JVP was elevated, he had crackles in bilateral bases, and florid pulmonary edema was seen on chest x ray. New echo and reassessment of echo performed at previous admission confirmed a missed inferior basal infarct and a muscular VSD. Patient was hemodynamically stabilized and then underwent surgical repair of his VSD.

Discussion: Our patient presented with an MI and an early VSD, a life threatening complication. This case is an example of how our strict timeline adherence can sometimes guide clinical decisions erroneously. It should call attention to the importance of a thorough daily physical exam with particular emphasis on any murmur heard despite findings on echocardiogram. Also, acute MI is seen with increased frequency in diabetics and is associated with higher morbidity and mortality due to the increased rate of complications. In the case of our patient, whom presented with DKA, there should be a lower threshold for suspicion of early complications due not only to increased myocardial stress but also due to the need for delayed reperfusion. This experience raises awareness of the need to search for complications early after an MI, the value of a proper physical exam, and most importantly to trust our clinical instincts regardless of timelines or tests.
Texas-Clinical Vignette-Poster Finalist
Heidi Torres, MD

Title: A Rare Case of Anaerobic Septic Arthritis

Authors: Heidi Torres, MD, Emily Wasson, MD

Introduction: Anaerobic bacteria cause septic arthritis in only 1% of cases. Fusobacterium species are strict anaerobic bacterium and part of oral, vaginal, and intestinal flora. It has been the causative agent of septic arthritis in cases with trauma, dental infections, oropharyngeal surgeries, and oropharyngeal sepsis to include Lemierre’s syndrome. Fusobacterium has very rarely been the cause of isolated septic arthritis without provoking trauma, oral infection, or surgery. The few reported cases have typically been seen in patients with chronic debilitating conditions.

Case Presentation: 48 year old male with a history of HTN, HLD, RA, and uncontrolled diabetes mellitus type 2 with associated neuropathy, retinopathy, and bilateral blindness presented with a two week history of fever, chills, and left knee pain such that he was unable to ambulate. Pain began suddenly without related trauma. After one week of pain he presented to a clinic and underwent steroid injection which did not relieve the pain. Physical exam in the ED showed a significantly swollen and tender left knee and calf with pain on passive and active range of motion. Orthopedic surgery performed arthrocentesis which revealed purulent fluid with 87,661 neutrophils and arthrotomy which revealed murky frank pus. Vancomycin and Zosyn were started. That night the patient had increased pain and swelling and underwent a second arthrocentesis. At 72 hours the cultures from initial arthrocentesis revealed Fusobacterium species in synovial fluid, only on thioglycollate medium. Flagyl was added. Second arthrocentesis confirmed Fusobacterium species at 96 hours on thioglycollate medium and anaerobic cultures. Vancomycin and Zosyn were discontinued. Panorex revealed no dental caries. On day 7 a third arthrocentesis was performed for swelling and unresolved pain. It revealed purulent fluid and a neutrophil count of 101,878. Arthrotomy was performed, again with frank pus. All cultures from this arthrotomy were negative. On day eight the patient was able to ambulate. He was discharged home on day twelve on a four week course of Flagyl.

Discussion: The clinical course of anaerobic septic arthritis is one of slow improvement often requiring more aggressive medical and surgical management than that of typical causative organisms. Though rare, patients can have devastating Results: such as joint destruction and loss of mobility if not treated appropriately early. Anaerobic septic arthritis should be considered in patients with atypical presentations, failure to respond to empiric treatment, persistent frank pus on repeat procedures, or chronic disease such as uncontrolled diabetes or RA as in our patient. These clues should prompt early addition of anaerobic coverage. The improvement and recovery of this patient can likely be attributed to multiple factors including the prompt initiation of Flagyl after giving credence to culture data based on the patient’s clinical picture as well as aggressive Orthopedic management and physical therapy.
US Air Force-Clinical Vignette-Poster Finalist
CAPT Matthew L Bezzant, USAF

Title: Unique case of penile Rhizopus Oryzae infection in an unexpected patient

Authors: Matt Bezzant, Kathryn Lago, Tatjana Calvano, San Antonio Military Medical Center

Introduction: Penile mucormycosis is an unusually rare infection which is not easily recognizable. To further complicate the matter, delayed diagnosis of mucormycoses can lead to significant morbidity and mortality due to the aggressive nature of the disease. The majority of reported penile mucormycoses have been in patients with hematologic malignancies or solid organ transplants. This case highlights an atypical presentation of a rare disease in a diabetic patient who is neither immunosuppressed nor in ketoacidosis. It also offers an alternative therapy to the poorly tolerated gold standard of amphotericin in patients who are uniquely susceptible to its side effects.

Case Presentation: An 80-year-old man with poorly controlled type II diabetes mellitus presented to his primary care doctor with dysuria, pain with foreskin retraction, erythema on the head of the penis and a hemoglobin A1C of 13.6 in Nov 2016. After failing topical hydrocortisone, he was diagnosed with fungal balanitis and started on topical nystatin. He failed to follow up and inexplicably discontinued all his diabetes medications. He returned to the clinic in late June 2017 with a complaint of one week of penis pain and an evolving lesion on the glans. At this point, his A1C had risen to >20%. He subsequently underwent debridement of the lesion. Cultures from the biopsy grew mucorales, and histopathology demonstrated angioinvasive fungal elements with non-septate hyphae, which were later confirmed by gene sequencing to be Rhizopus oryzae. After multiple attempts to contact the patient, he was admitted to the hospital to initiate therapy with liposomal amphotericin B. He tolerated it well initially, but then developed an elevated alkaline phosphatase and rising creatinine consistent with the adverse effects of amphotericin. During this time, he underwent multiple debridements culminating in a partial penectomy. After 7 days of therapy amphotericin was replaced with isavonazonium, which he tolerated well. After 3 days the pathology from his penectomy came back with clear margins and therapy was discontinued. One month later he was healing well without evidence of infection and his creatinine was returning to baseline.

Discussion: Our case demonstrates the importance of early recognition of invasive mucormycosis at a rare site of infection in a patient without clear risk factors. Given the high mortality of untreated mucormycoses, this case highlights the importance of considering them in diabetic patients, even those who are not immunosuppressed or in ketoacidosis. Given the negative side effect profile of amphotericin, patients with underlying renal disease are often poor candidates for therapy even though they can be at high risk for contracting mucorcycosis. For our patient, who did not tolerate amphotericin, isavuconazole provided a more tolerable alternative and appeared to be equally efficacious. This supports the possibility of using isavuconazole as first line therapy for patients with underlying kidney disease, but additional research is required to confirm these findings.
Title: Statin-Induced Necrotizing Autoimmune Myopathy: A Case of Delayed Onset Following Statin Discontinuation

Authors: Andrea N. Keithler, DO¹, Brian C. Pomerantz, MD¹, Diane-Ngan H. Trang, MD², Mary Barbara, MD², Rosco S. Gore, MD¹. ¹San Antonio Uniformed Services Health Education Consortium, San Antonio, TX, ²University of Texas Health Science Center at San Antonio, San Antonio, TX

Introduction: Statin-induced necrotizing autoimmune myopathy (SINAM) is an immune mediated myopathy characterized by proximal muscle weakness and necrosis in the presence of autoantibodies to hydroxyl-methyl-glutaryl-coenzyme-A (HMG-CoA) reductase. It is a rare side effect of statin use that must be recognized quickly in order to prevent the development of irreversible muscle weakness. Most cases occur within the first year of statin use, although some may present sub-acutely with progression of symptoms following withdrawal of therapy. We report a rare case of an individual who developed SINAM two years after cessation of therapy.

Case Presentation: A 60-year-old Hispanic man with history of hypertension, hyperlipidemia, and hypothyroidism presented with several months of progressive myalgias and proximal muscle weakness. He had a history of atorvastatin use, but had discontinued it two years prior due to financial difficulties. Physical examination was notable for tenderness of the bilateral proximal lower extremity muscle groups as well as bilateral proximal upper and lower extremity muscle weakness. Initial laboratory evaluation was remarkable for a creatine kinase (CK) of 10,737, erythrocyte sedimentation rate (ESR) of 54, and C-reactive protein (CRP) of 11.30. Despite fluid resuscitation, muscle weakness and CK elevation persisted. Electromyogram (EMG) was concerning for muscle fiber necrosis; quadriceps muscle biopsy ultimately confirmed myopathic changes with necrosis. Subsequently, enzyme-linked immunosorbent assay (ELISA) revealed antibodies to HMG-CoA reductase, which, in concert with his clinical presentation and an extensive evaluation for alternate myopathic processes, suggested SINAM as the cause of his weakness. Pulse dose steroids and intravenous immune globulin (IVIG) were initiated with profound improvement in his weakness, after which he was transitioned to rituximab for continued immunosuppression.

Discussion: SINAM is an immune mediated myopathy manifested by muscle weakness in the setting of statin exposure. In contrast to other forms of statin-associated myotoxicity, including myalgias, myopathies, and rhabdomyolysis, SINAM is mediated by an autoimmune response and is associated with persistent or progressive symptoms despite statin discontinuation. SINAM generally occurs in the absence of risk factors for other statin-associated myotoxicities, but a genetic predisposition in individuals with certain HLA alleles has been described. Characteristic biochemical, EMG, and histologic findings support the diagnosis. Importantly, the irreversible muscle weakness seen in SINAM can be prevented with cessation of the offending agent and initiation of immunosuppression, so early and accurate diagnosis is critical to good clinical outcomes. This case is unusual in that onset of myopathy occurred in a delayed fashion, although, with the increasing availability of ELISA testing for HMG-CoA reductase autoantibodies, it is felt that recognition of late onset SINAM may increase.

References


US Air Force-Clinical Vignette-Poster Finalist
CAPT Diana Le

Title: Enteroviral meningoencephalitis as a Complication of Rituximab Therapy for Rheumatoid Arthritis

Authors: Authors: D. Le, MD1; J. Scott, DO1; A. Ford2, MD; D. Lindholm, MD1, Programs: 1Department of Internal Medicine, Wright State University, Boonshoft School of Medicine. 2Department of Neurology, Wright Patterson Medical Center.

Introduction: Enteroviral meningoencephalitis is typically observed in patients with congenital immunodeficiencies. However, there have been rare cases of this infectious complication of rituximab in the treatment of hematologic malignancies.1 To our knowledge, this is the first case of enteroviral meningoencephalitis in a patient receiving rituximab for rheumatoid arthritis (RA).

Case Presentation: A 37-year-old female with severe treatment resistant RA, currently managed with rituximab, presented to the emergency department. Presenting symptoms included 7 day history of intermittent fever, abnormal gait, headache, and confusion. Exam was notable for disorientation to time, facial and upper extremity tremor, and antalgic gait.

MRI of the brain showed no acute intracranial pathology or abnormal enhancement. Laboratory evaluation included normal CBC, metabolic panel, ESR, and CRP. Cerebrospinal fluid (CSF) analysis revealed normal protein, normal glucose, and a lymphocytic pleocytosis with 31cells/mCL and 83% lymphocyte predominance. Commercial multiplex CSF PCR was only positive for enterovirus. Patient had progressive neurologic development of agitation, delirium, and aphasia. Treatment was initiated with intravenous immunoglobulin (IVIg) at 0.4g/kg/d for five days with significant improvement in her agitation, confusion, tremor, gait, aphasia, and headaches.

Discussion: Rituximab is a monoclonal anti-CD20 antibody that Results: in B-cell depletion and hypogammaglobulinemia for 6-12 months after administration.2 Initially used for treatment of lymphoma and immune cytopenias, the list of indications has grown. In 2006, rituximab was approved for RA that had an inadequate response to tumor necrosis factor (TNF) antagonists.

The rituximab-induced hypogammaglobulinemia has well known, though rare, neurological complications due to reactivation of JC virus. However, enteroviral meningoencephalitis is a relatively unknown entity, with only 11 reported cases of enteroviral meningoencephalitis associated with treatment of lymphomas and autoimmune cytopenias with rituximab.1 The nonspecific clinical syndrome of headache, lethargy, tremor and weakness can vary in severity and time from exposure to the drug.3 Like our patient, most cases are diagnosed with enterovirus PCR of the CSF. To our knowledge, there are no known cases of enteroviral meningoencephalitis in a patient receiving rituximab for a rheumatologic disease.

While there is no known treatment for enteroviral meningoencephalitis, IVIg has been used2. Reported treatment outcomes are variable ranging from resolution of neurologic symptoms to progression of disease and death. There are several possible reasons for the wide spectrum, including delays in diagnosis due to nonspecific disease presentation as well as differences in the quantitative content of enteroviral antibodies between IVIg preparations.4
We present this case to draw attention to the idea that patients on rituximab, regardless of the indication for use, are susceptible to severe complications of otherwise typically self-limited pathogens. Furthermore, our case supports the potential utility of treatment with IVlg in enteroviral meningoencephalitis in patients treated with rituximab.

References

Title: Draft - Chapter Winner

Diffuse Erythrodermic Psoriasis in a Patient without Psoriasis

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Introduction: Erythrodermic psoriasis (EP) is a rare, severe form of plaque psoriasis with sudden development of inflammatory, erythematous skin plaques and edema. Diagnosis requires involvement of over 75% of the total body surface area (TBSA), and may lead to metabolic and systemic compromise. Untreated, the patient may succumb to high output heart failure secondary to cutaneous water loss. We present a case of EP as a patient’s first presentation of psoriasis.

Case Presentation: The patient is a sixty-eight year old morbidly obese female who developed a papulosquamous eruption under her breast one month after receiving a solumedrol injection for a presumed insect bite. Over one week, the rash spread to involve large areas of the trunk, proximal extremities and scalp. A skin biopsy found psoriaform dermatitis involving 40% of her TBSA, and topical triamcinolone cream was initiated. Two weeks later she presented to the emergency room with tachycardia and hypotension with diffuse skin pain, leukocytosis and an acute kidney injury. She had weeping, coalescing violaceous plaques on the chest, abdomen, back, and upper and lower extremities involving greater than 90% TBSA. There were pustules scattered across separated plaques on both legs. With no identifiable triggers, personal nor familial history of psoriasis, empiric antibiotic coverage with clindamycin and miconazole was initiated for possible superinfection. Tuberculin skin test, chest x-ray, and hepatitis panels were unremarkable. She was continued on triamcinolone cream with wet skin wraps. Her acute kidney injury resolved with IV fluids and she developed diffuse edema in the setting of continued weeping plaques. The pustules on her legs formed vesicles prior to rupturing with gradual coalescence of diffuse erythematous plaques. Direct immunofluorescence was negative for IgG, IgA, IgM and fibrin along the dermal-epidermal junction without evidence of IgA pemphigus. Skin biopsy returned as pustular psoriasis. The patient was started on infliximab infusions. After the third infusion six weeks later, she had 10% TBSA still affected, and was lesion free after five months.

Discussion: Erythrodermic psoriasis is a rare and potentially fatal variant of plaque psoriasis believed to affect less than 1% of patients with psoriasis. Treatment includes adequate hydration, topical steroids and vitamin D analogs, and immunosuppression for uncontrolled symptoms. With a high degree of morbidity associated with it, EP is rare as the initial presentation of psoriasis, and was triggered in this patient by steroid withdrawal after a single solumedrol injection.
US Air Force-Clinical Vignette-Poster Finalist
CAPT Jason M Thomas, MD

Title: Scurvy: When Life Gives You Shock, Make Lemonade

Authors: Jason Thomas, MD, PGY 3, Wright State University Boonshoft School of Medicine, Dayton, OH, Christopher Madison, MD, PGY 2, Wright State University Boonshoft School of Medicine, Dayton, OH, Kathryn Burtson, MD, attending physician, Wright State University Boonshoft School of Medicine, Dayton, OH

Introduction: Scurvy is an old disease, documented even in ancient Egypt. This disease can be devastatingly debilitating and be life threatening. Surprisingly, scurvy plagues a significant percentage of populations in resource-rich nations.

Case Presentation: A 69-year-old female with a history of hypertension and depression presented with hemodynamic instability and anemia. She complained of three weeks of bilateral lower extremity bruising and pain, preceded by a 4-month history of weakness and 20-pound weight loss. She denied alcohol use. Her diet was limited to canned green beans, burgers, eggs, and bananas. On exam, the patient was hypotensive and had poor dentition, perifollicular hemorrhages, and extensive ecchymoses in different stages of healing over her posterior thighs and calves which were tender to palpation. Laboratory studies revealed a hemoglobin of 9.5 g/dL, albumin of 2.3 g/dL, and ESR of 75 mm/hr. Platelet count, INR, B12, folate, and TSH were within normal limits. During her work-up and treatment, the patient was stabilized after 4 liters of crystalloid. With suspicion of scurvy, a vitamin C level was drawn and resulted at 0.0. Patient was advised to take supplemental vitamin C. On a follow-up appointment two weeks later, the patient endorsed improvement in pain and exercise tolerance and was noted to have marked improvement in skin findings.

Discussion: While animals produce Vitamin C from glucose, humans lack a functional copy of a crucial enzyme. Thus, humans are required to attain vitamin C from food intake. People who consume diets lacking in vitamin C can develop scurvy. Many believe scurvy to be a historical disease, however nearly 10% of the US population suffers from vitamin deficiency. Due to integral role of vitamin C in numerous pathways, scurvy presents with a variety of symptoms including, bleeding diathesis, anemia, fatigue, myalgias, perifollicular palpable purpura, and gingival swelling. Rarely, patients can present with hemodynamic instability attributed to the lack of intrinsic catecholamines. The treatment for this condition is very simple. Patients often have resolution of symptoms within 48 hours of supplementing vitamin C. Scurvy is a devastating disease with a very simple cure. Clinicians must be aware of signs and symptoms of vitamin deficiencies and counsel patients on the importance of a healthful diet.

References

US Army-Clinical Vignette-Poster Finalist
CPT Brennan R Cebula, MC USA

Title: Recurrent Ganciclovir Resistant Cytomegalovirus Colitis Managed With Intravenous Immunoglobulin Monotherapy

Authors: Brennan R. Cebula, CPT, MC, USA, Christina M. Schofield, MD, FACP Madigan Army Medical Center, Tacoma, WA

Introduction: Cytomegalovirus (CMV) is a significant cause of morbidity in patients with impaired cellular immunity including those on immunosuppressive therapy. Development of CMV antiviral resistance is a challenging complication with limited evidence to guide management.

Case Presentation: A 45 year-old male with history of common variable immune deficiency complicated by granulomatous-lymphocytic interstitial lung disease (GL-ILD) treated with azathioprine and one cycle of rituximab 6 months previously presented with fever, abdominal pain, and bloody diarrhea. CT-abdomen/pelvis revealed pan-colitis and colonoscopy revealed colonic ulcerations. Tissue histology was consistent with CMV colitis. The patient underwent induction with ganciclovir for 14 days with improvement in symptoms. He was transitioned to valganciclovir 900mg twice daily for an additional week to complete treatment followed by 450mg twice daily for suppression to be continued until completion of rituximab therapy. Over the next 2 years the patient experienced multiple recurrences of CMV colitis in the setting of variable adherence to suppressive valganciclovir therapy. Antiviral resistance was eventually suspected and confirmed with gene sequencing revealing a UL97 kinase H520Q gene mutation conferring high-level ganciclovir resistance. Valganciclovir was discontinued and on his next recurrence he underwent induction with foscarnet along with one cycle of CMV immunoglobulin (CMVIG) which resulted in remission. He was transitioned to foscarnet 80mg/kg daily for suppression, but subsequently developed nephrotoxicity necessitating its discontinuation. Valacyclovir was started to attempt suppression while avoiding inducing further antiviral resistance. He thereafter experienced recurrence of symptoms consistent with CMV colitis, but this was confounded by detection of Campylobacter and entropathogenic Escherichia coli on stool PCR. He received a five-day course of ciprofloxacin for potential bacterial infectious diarrhea without improvement in his symptoms. Determining that this presentation represented recurrent CMV colitis, it was decided to attempt management with a repeat treatment cycle of CMVIG. The patient’s symptoms improved, and he was transitioned to monthly suppressive CMVIG. He has experienced no recurrence of symptoms suggestive of CMV colitis, and a follow-up colonoscopy with tissue biopsy revealed no evidence of active CMV colitis.

Discussion: Ganciclovir is the mainstay of therapy for CMV disease, but resistance necessitates therapy with antivirals associated with limiting toxicity as seen in our patient. Additionally, no controlled trials to guide the management of CMV antiviral resistance exist. CMVIG has shown promise as an adjunct to antiviral therapy for CMV disease and as monotherapy for asymptomatic CMV infection and non-severe disease in transplant patients, but the evidence base for this is very limited. This case represents a unique presentation of ganciclovir-resistant CMV colitis in a patient with CVID complicated by GL-ILD requiring immunosuppressive therapy successfully managed with CMV IVIG monotherapy, and highlights the need for further studies to guide the management of antiviral resistant CMV and determine the optimal role of CMVIG.

References


Title: Coil Embolization of Intrapulmonary Sequestration: An Alternative to Surgery

Authors: John Ellis, MD, Daniel Desmond, MD, Brian Ching, DO, Jordanna Hostler, MD (FACP)

Introduction: Symptomatic bronchopulmonary sequestration (BPS) is uncommon. It is seen more frequently in the pediatric population than in adults. It has traditionally been treated with surgical resection; however, a handful of cases have been treated with angiographic embolization. Given the inherent risks of cardiothoracic surgery, embolization of the anomalous vessel is an enticing alternative treatment. We present a case of a 56-year-old female with known, symptomatic, intralobar pulmonary sequestration (ILPS) that was successfully treated with coil embolization.

Case Presentation: A 56-year-old woman with a history of chronic lymphocytic leukemia in remission was admitted to the ICU with an episode of hemoptysis. CT scan of the chest demonstrated left lower lobe (LLL) intralobar sequestration which was fed by a large tortuous vessel branching off of the descending thoracic aorta. At the origin of the aberrant vessel it measured approximately 1 cm. No alternative source of bleeding was detected. Resection of the LLL sequestration is the current standard treatment strategy of symptomatic ILPS. Cardiothoracic surgery noted that surgical resection would be difficult, risky, and would require a thoracotomy and lobectomy. Interventional radiology offered embolization of the lesion as a safer alternative to surgery. Multiple coils, size 6mm - 13mm, were used to embolize the sequestration. No considerable flow distal to the coils was noted post-embolization. Her post-procedure course was notable for pleurisy that responded to oral analgesia and the brief use of bronchodilators. At her 9-month follow up she reported no pulmonary symptoms, cough, or hemoptysis.

Discussion: ILPS is a rare condition that typically requires surgical management. This case demonstrates the efficacy of coil embolization as an alternative management strategy. To date, limited case reports of adults treated with endovascular embolization exist. Treatment of symptomatic pulmonary sequestration with embolization can be considered as an alternative to surgical resection, in cases where surgery would have significant morbidity and mortality risks. A multidisciplinary team should assess the patient to determine which treatment course provides the best risk to reward balance and likelihood of a durable response.
**US Army-Clinical Vignette-Poster Finalist**  
**Kimberly Fabyan, MD**

**Title:** Heartbreaking Cellulitis: A Rare Case of Purulent Constrictive Pericarditis from Community Acquired MRSA Cellulitis in a Healthy Adult

**Authors:** Kimberly D. Fabyan, MD (1); Geoffrey J. Cole, MD (2)

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**Introduction:** Heartbreaking cellulitis: A rare case of purulent constrictive pericarditis secondary to community-acquired methicillin resistant staphylococcus aureus (MRSA) cellulitis in a healthy adult.

**Case Presentation:** A 48-year-old man with no significant medical history developed cellulitis around his elbow while deployed. An abscess formed and was drained. He was started on amoxicillin-clavulanate. Two days later, cultures were positive for MRSA, and antibiotics were changed to linezolid. After two doses, a pruritic rash developed concerning for an allergic reaction; antibiotics were stopped. Prednisolone and desloratadine were prescribed, and the rash improved.

Thirteen days after original presentation, he presented to the Emergency Department following a syncopal event with worsening dyspnea, chest pain, cough, and fever. Chest x-ray was concerning for a pericardial effusion. Echocardiogram confirmed pericardial effusion with signs of tamponade and heart failure with a left ventricular ejection fraction of 25%. A pericardiocentesis was performed. Blood and pericardial fluid cultures grew MRSA. He was started on intravenous vancomycin and gentamicin, and medevac’d to our hospital.

Upon arrival, he had persistent tachycardia with conversational dyspnea. Exam was also notable for jugular venous distension to 14cm and muffled heart sounds. Echocardiogram showed a moderate, loculated effusion with pericardial thickening, severely reduced bi-ventricular function, and a septal bounce concerning for constrictive pericarditis. Constrictive physiology was confirmed with heart catheterization. Cardiothoracic surgery was consulted, and the patient subsequently underwent pericardiectomy. Though his post-operative course was tumultuous, he was discharged two weeks later on intravenous antibiotics and continues to recover.

**Discussion:** MRSA related community-acquired infections among immunocompetent adults are on the rise. While the majority of these infections are mild, invasive and life-threatening infections do occur. Purulent pericarditis is a known complication of staphylococcus infections; however, in the era of antibiotics, purulent pericarditis is a rarity in patients without significant immunosuppression and other risk factors. For our patient, the suppression of his immune system with the use of steroids coupled with insufficient antibiotic treatment allowed the MRSA infection to spread to the pericardium. This severe infection was complicated by constrictive pericarditis. There are a small number of cases of
community-acquired MRSA purulent pericarditis in otherwise healthy adults reported, none of which were complicated by pericardial constriction or associated with steroids. This case demonstrates a rare complication of a mild skin and soft tissue infection which could have possibly been prevented had antibiotics been continued. This case argues that caution should be used when steroids are administered the setting of MRSA infections.
Title: Milliseconds Between Life and Death: A Case of Recurrent Torsades

Authors: Joel Guess, MD (Associate); Kinsley Hubel, MB BCH BAO (Member); Jacob Hurley, MD (Associate); Jessica Bunin, MD (FACP)

Introduction: QT prolongation is a common, often multifactorial problem which can lead to severe cardiac conduction disturbances, most notably Torsade de Pointes (TdP). Acquired QT prolongation is often treated with electrolyte replacement and discontinuation of QT prolonging drugs. In the presence of Torsades, magnesium sulfate with or without lidocaine is often added for myocardial stabilization. If these interventions fail to suppress further dysrhythmias, the next intervention can include transcutaneous pacing and beta agonists. We present the case of a gentleman with acquired QT prolongation treated with isoproterenol.

Case Presentation: The patient is a 54 year old male was admitted to the ICU following a ventricular fibrillation (VF) arrest. After ROSC was obtained, the patient regained consciousness without residual deficits. There was no evidence of ischemia on his post-arrest ECG and troponin-I levels were undetectable initially. The patient’s ECG did, however, demonstrate a likely etiology in a QTc of 917ms. The patient was placed on an amiodarone drip prior to his transfer to the ICU, but, shortly after arrival, he developed Torsade des Pointes (TdP) with subsequent VF arrest with ROSC again achieved after one defibrillation. All QT prolonging drugs were discontinued, including amiodarone, and electrolyte abnormalities were corrected with initial improvement in QTc (688ms). Despite these interventions, the patient had recurrent TdP with VF arrest requiring defibrillation to achieve ROSC. He was started on both magnesium and lidocaine drips to prevent further episodes of torsades; however, this resulted in his QTc increasing to 816ms. An isoproterenol drip was initiated to achieve a goal heart rate of at least 90 bpm. Isoproterenol administration resulted in immediate improvement in his QTc to 490ms and he had no further episodes of TdP. All drips were stopped within 48 hours of admission. He had persistent mild QTc prolongation 500-550ms ultimately leading to AICD placement. Subsequent genetic testing revealed a polymorphism of undetermined significance in the KCNQ1 gene associated with Long QT Syndrome type 1.

Discussion: TdP is precipitated by early after depolarizations (EAD), often arising from myocardial Purkinje cells and propagated in the setting of QT prolongation. Isoproterenol is a non-selective β-agonist that increases both chronotropy and inotropy. Increased chronotropy induces more rapid repolarization and decreases the degree of regional repolarization heterogeneity that acts as the substrate for reentrant tachyarrhythmias. Isoproterenol was the only treatment that shortened the QTc in our patient. It is indicated as a bridge to pacing or when pacing is unavailable in patients with acquired prolonged QT resulting in TdP. Isoproterenol is contraindicated in many forms of congenital long QT syndrome because it can increase QT duration and risk of TdP.
Title: Immune complex glomerulonephritis secondary to anti-tumor necrosis factor α therapy for inflammatory bowel disease

Authors: Alexandra Stewart, DO, ACP Associate; Jorge Martinezosorio, MD, ACP member; MAJ Sarah Gordon, MD, ACP member, Department of Medicine, Tripler Army Medical Center, Hawaii

Introduction: Glomerulonephritis among patients with comorbid inflammatory bowel disease (IBD) is a well-documented occurrence. Various IBD therapies are also associated with development of abnormal renal function. Anti-tumor necrosis factor α (TNF-α) therapy is one commonly used treatment for IBD. Cases of lupus-like syndromes associated with anti-TNF-α therapy have been reported, rarely with renal involvement. We report an unusual case of lupus-like glomerulonephritis associated with anti-TNF-α therapy for Crohn disease.

Case Presentation: A 39-year-old Caucasian female with a history of Crohn disease, leukocytoclastic vasculitis, and transverse myelitis was evaluated for hypertension, microhematuria, and rising serum creatinine. She underwent renal biopsy for proteinuria of 1.6 grams with preserved renal function (creatinine 0.8 mg/dL), and was diagnosed with IgA nephropathy. She was treated with ACE inhibitors and prednisone. Her Crohn disease was managed with sulfasalazine and infliximab. One year later she presented with hypertension and acute kidney injury with creatinine at 1.5 mg/dL. A second renal biopsy revealed a proliferative lupus-like immune complex glomerulonephritis (ICGN). Serologic workup was notable only for weakly positive anti-double stranded DNA. She did not meet criteria for systemic lupus erythematosus. The biopsy findings were attributed to infliximab which was then transitioned to adalimumab. Her renal disease transiently improved, and worsened six months later, with peak SCr of 3.4 mg/dL. Imaging demonstrated normal renal size and cortical thickness. A third renal biopsy was completed for clarification, which confirmed an active ICGN with diffuse segmental sclerosis. The biopsy demonstrated significant chronicity with 50% interstitial fibrosis and tubular atrophy. Electron dense deposits were found in subepithelial, subendothelial and mesangial compartments. There were no tubuloreticular inclusions. Immunofluorescence demonstrated 1+ staining for IgG, IgM, C1q, kappa and lambda, and C3 with trace IgA in a “full house” pattern. Adalimumab and sulfasalazine were discontinued and serum Cr improved to 2.0 mg/dL. She was felt to have a lupus like glomerulonephritis induced by anti-TNF therapy or sulfasalazine.

Discussion: The spectrum of potential renal disorders among patients with IBD includes glomerulonephritis, most commonly IgA nephropathy. We identified around 50 case reports describing drug-induced lupus associated with anti-TNF therapies, but only six reports of lupus glomerulonephritis. Our patient’s biopsy was consistent with a World Health Organization class IV pattern of lupus-like nephritis. This case adds to existing reports of this unusual potential side effect of anti-TNF-α therapy. Monitoring of renal function and urinalysis is warranted in individuals receiving anti-TNF-α therapy in order to identify early the development of drug-induced glomerulonephritis.

References


Title: “Lyme Eye in the Blind Guy: An eye catching case of Neuroborreliosis presenting with bilateral Optic Neuritis”

Authors: Unger, Jason A, MD, (Resident ACP Member), Woodmorris, Robert, MD, Mekenon, Dawitt, MD, Camillie Costan-Toth MD, (ACP Member)

Introduction: Optic neuritis can sometimes be an early finding of many systemic conditions that are encountered by Internists. It has an incidence of ~1-5/100,000 per year, and is most commonly caused by demyelinating neuropathies. Bilateral presentations are rare, but are more likely to have an atypical etiology. Our unique case of a military service member with bilateral vision loss, who was later diagnosed with Neuroborreliosis, helps to illustrate this concept.

Case Presentation: A 33 year old male Active Duty Marine, presented to his local emergency department over the summer, reporting two weeks of worsening bilateral eye pain with vertical gaze, and blurry vision, worse on the left side. He reported that prior to this, he had returned from a two week hiking trip in Massachusetts, in which he endorsed multiple insect bites, but no rash. His was otherwise in good health and had no history of regular medication use. He was seen in the Ophthalmology clinic the next day, where his visual acuity was noted to be 20/100 OD, and 20/200 OS. His funduscopic exam showed bilateral optic disk swelling. A battery of serum studies was ordered. An MRI of the orbits showed bilateral T2 enhancement of both optic nerve sheaths, and post contrast enhancement of the intra-conal fat. He was evaluated by Neurology the next day and underwent lumbar puncture which revealed a mild lymphocytic pleocytosis. A week later, a Borrelia burgdorferi antibody ELISA (from his initial serum studies) returned positive. Follow up Western blot testing showed both a positive IgG and IgM band pattern, (5/8 expected IgG, and 3/3 expected IgM antibodies). The rest of his initial work up including CBC, CMP, ESR, HIV, RPR, Nuclear-Ab panel, TSH, SPEP, B12, and folate, were all unremarkable. Additional CSF studies were all negative as well. Based on his outdoor exposure history, his objective findings on eye exam and MRI, and a grossly positive serum Lyme Western Blot, the diagnosis of Neuroborreliosis was made. He was admitted to the Internal Medicine service on 23 Aug and evaluated by the Infectious Disease Consultant. A PICC line was placed, and Ceftriaxone 2GM IV daily was started for 4-6 weeks of intended therapy.

Discussion: Optic neuritis most often presents with unilateral visual loss, and pain with eye movement. Characteristic imaging findings support the diagnosis. Demyelinating conditions remain the most likely cause, but the differential is broad. The associated pain pattern, laterality of vision loss, and timing of symptom progression, are most helpful to narrow the differential. This patient’s bilateral vision loss, and sub-acute presentation over two weeks, were clues to his atypical etiology. In areas where Lyme is endemic, such as the north eastern United States, it remains an important consideration in this expanded differential.

References

Title: VZV reactivation with associated meningitis in a patient receiving Infliximab: Time for a new look at treatment.

Authors: Allison Bush, MD, LT USN (Member) Shannon Wood, MD CPT USA (Member); Joshua Hartzell, MD LTC USA (Fellow) Walter Reed National Military Medical Center, Bethesda,

Introduction: The increased risk of varicella zoster virus (VZV) reactivation in the form of shingles is well established in patients receiving TNF-alpha inhibitors; however, neurologic complications are rarely reported and best treatment practices are not well defined. We present a case of VZV meningitis in an adult male on Infliximab successfully treated with rapid transition to oral antiviral therapy.

Case Presentation: A 56 year old male with long-standing ulcerative colitis (UC) treated with Infliximab presented with 15 days of a new, severe, bi-temporal headache accompanied by neck pain, photophobia, phonophobia, fevers and a painful vesicular rash on his right anterior chest. The patient received Infliximab three weeks prior to the onset of symptoms. Physical examination revealed a non-toxic, afebrile male with a resolving herpes zoster rash over the right anterior chest wall in the T4 dermatome distribution and pain with neck flexion. Basic labs demonstrated a white blood cell count (WBC) of 6.0 x 10^3/mcl. Computed tomography of the head without contrast was negative for intracranial pathology. Cerebral spinal fluid (CSF) parameters and cell count demonstrated elevated protein, normal glucose and a leukocyte predominant pleocytosis. Rapid polymerase chain reaction (PCR) of CSF detected VZV DNA. Treatment was initiated with intravenous acyclovir dosed at 10mg/kg IV q8h with rapid improvement in symptoms in less than 24 hours. The patient was transitioned to valacyclovir 1000 mg PO three times a day on hospital day 2 and discharged home with a 14-day course of therapy. Approximately 48 hours after initiation of anti-viral therapy the patient reported complete resolution of symptoms.

Discussion: VZV reactivation is three times more likely to occur in patients being treated with TNF-alpha inhibitors than the general population, but concomitant meningitis is rarely reported. While treatment for severely ill individuals invariably involves the use of intravenous acyclovir for several weeks, treatment strategies for stable, non-toxic patients presenting in the outpatient setting remains unclear. This case demonstrates that VZV meningitis in a non-toxic patient can be treated successfully with rapid transition to oral antiviral therapy. With close follow-up, similar patients could avoid the need for prolonged intravenous antiviral therapy.
Title: A Case of Sneaky Sarcoid: Neurosarcoidosis Presenting as Panhypopituitarism

Authors: Hillary A. Chace, D.O. (Associate), Sarah D. Hodges, D.O.

Introduction: Sarcoidosis is a chronic inflammatory disease that is most often characterized by non-caseating granulomatous inflammation and systemic involvement. Although no definitive test for sarcoidosis exists, three components are required: clinical/radiographical manifestations, exclusion of other diseases, and histological evidence of non-caseating granulomas. Neurosarcoidosis occurs in approximately 5 to 10% of patients with cranial neuropathies the most common manifestation. MRI findings range from an intraparenchymal tumor-like mass to diffuse leptomeningeal enhancement. CSF analysis is non-specific, but lymphocytic pleocytosis with elevated protein is often seen in active disease.

Case Presentation: A 36-year-old black male presented to his primary care physician six months prior to ultimate diagnosis with fatigue and weight loss. He was found to have low testosterone which prompted an endocrine workup significant for decreased FSH and LH with increased ACTH, cortisol, and prolactin. MRI of the brain revealed a 0.3 cm pituitary lesion, thought to represent a microadenoma. Within the next several months, he developed ataxia, diplopia, nausea, vomiting, and headaches, prompting a second MRI which showed diffuse leptomeningeal enhancement and pituitary gland enlargement. Robust meningeal enhancement of the left cerebellar hemisphere, anterior pons, right middle cranial fossa, right cavernous sinus, and the optic chiasm was also seen. Faint leptomeningeal and nodular root enhancement throughout the cervical, thoracic, lumbar and cauda equina was found on a spinal MRI. Chest CT revealed bilateral hilar lymphadenopathy and a bronchoscopy-guided biopsy revealed non-caseating granulomas. CSF angiotensin converting enzyme (ACE) was elevated to 13.2 (reference 0-2.5), with a normal serum ACE and normal serum calcium. CSF was also significant for increased protein, low glucose, and high IgG at 80 (reference 10-30). He was diagnosed with probable neurosarcoidosis and initial management included high dose steroids and DDAVP. Since diagnosis, his treatment has been refined, with the addition of levothyroxine, testosterone, and immunosuppressive therapy with mycophenolate mofetil. Neurologic function has returned to baseline and he has noted marked improvement in his neuroendocrine function.

Discussion: Neurosarcoidosis often presents with multi-focal findings in both the peripheral and central nervous system. This patient had manifestations of cranial neuropathy, neuroendocrine dysfunction, myelopathy, and cerebellar dysfunction. There was a high level of ACE and IgG in the CSF, but the serum ACE was normal. It is uncommon for neurosarcoidosis to present as panhypopituitarism prior to frank neurologic manifestations, which may explain the delay in diagnosis. This case is unique and reinforces that the differential diagnosis of any patient with panhypopituitarism should include sarcoidosis.
US Navy-Clinical Vignette-Poster Finalist
LCDR Hillary A. Chace, MC USN

Title: A Case of Novel Nivolumab in Elderly Hodgkin’s Lymphoma

Authors: Hillary A. Chace, D.O. (Associate), Justin A. Harder, M.D. (Fellow)

Introduction: Hodgkin’s lymphoma (HL) arises from germinal center B-cells (unique cellular composition containing Reed-Sternberg cells) and is either classified as classical or nodular lymphocytic based on tumor immunophenotype. It has a bimodal age distribution, first in young adults approximately 20 years of age, and again in older adults approximately 65 years of age. In classical HL, the Reed-Sternberg cells typically express CD15 and CD30 and lack global expression of CD20, but 95% are weakly positive for PAX-5. PD-1 is a programmed cell death protein 1 and acts as an immune checkpoint and guards against autoimmunity. PDL-1 is a transmembrane protein ligand that suppresses the immune system with upregulation and allows cancers to evade the immune system. PD-1 overexpression proposes that relapsed or refractory disease may have a genetically determined vulnerability to PD-1 blockade. Novel studies have shown Nivolumab, a fully human monoclonal IgG4 antibody directed against PD-1, can be successfully used in these patients with refractory HL.

Case Presentation: An 89 year old male presented with lethargy, anorexia, chest pain, and shortness of breath and was found to have bulky retroperitoneal, supraclavicular, and pelvic sidewall lymphadenopathy. He underwent a needle core biopsy revealing HL Stage IIIB. Pathology demonstrated Reed-Sternberg cells that were weakly PAX5+, CD30+, CD15+, CD20-. He was initially treated with Brentuximab, a monoclonal antibody targeting CD30. After 3 cycles, the patient showed a mixed response with resolution of left supraclavicular lymphadenopathy and improvement in his B-symptoms, but new inguinal lymphadenopathy. Brentuximab was discontinued due to a Grade III neuropathy and maculopapular rash. Given the suboptimal response and side effects, he was started on Nivolumab. Nine months into therapy a surveillance PET-CT demonstrated complete response with resolution of inguinal, cervical, and retroperitoneal lymphadenopathy. The patient gained 45 pounds, had minimal chemotherapy side effects, and no current evidence of recurrence.

Discussion: Hodgkin's Lymphoma represents 10% of all cases of malignant lymphoma. In most cases, HL is a curable disease when treated with chemotherapy with or without radiation. Curative treatments are too toxic for the elderly, leaving few options to treat advanced disease. In recent novel studies it was shown that Nivolumab was highly effective for HL with a more favorable toxicity profile, making it an attractive option for elderly patients with relapsed or refractory HL.
Title: A Perfect Storm: Hemophagocytic Lymphohistiocytosis in the Setting of HIV, EBV, and Hodgkin Lymphoma

Authors: Matthew T Nelson, MD, Member ACP, Jaime P Gastwirt, MD, Member ACP, Evan C Ewers, MD, Member ACP

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is an aggressive overactivation of the immune system, which is often associated with infection, malignancy, or rheumatologic disorders. It is marked by fever, organomegaly, cytopenias, elevated ferritin, and hemophagocytosis on bone marrow aspirate. Here, we present a case of hemophagocytic lymphohistiocytosis in the setting of HIV, classical Hodgkin lymphoma, and Epstein-Barr virus.

Case Presentation: A 45-year-old HIV-positive male with previously well-controlled disease on antiretroviral therapy, but with recent noted CD4+ T-cell decline (from 705/µL to 298/µL over the previous year) in the setting of continuously undetectable viral load, presented with fatigue, anorexia, nausea, vomiting, dark urine, and fever to 102°F. Workup only revealed mildly elevated transaminases, and he improved with intravenous fluids. Blood and urine cultures were negative. A repeat CD4+ count was 134/µL (17%) with undetectable viral load. His acute symptoms resolved, but developed worsening fatigue, recurrent fevers, sweats, and unintentional weight loss. Two months later, he re-presented with fever to 101.8°F, nausea, vomiting, and malaise. Work up was notable for leukopenia, mild transaminitis, elevated CRP and ESR (14.42 and 91, respectively), ferritin of 2070 ng/mL, and microcytic anemia with a hemoglobin of 12.6 g/dL. Serologic workup for viral and autoimmune hepatitis, Wilson's disease, and alpha-1-antitrypsin disease were negative. Extensive workup for opportunistic pathogens was negative. EBV PCR demonstrated 107,177 copies/mL. Abdominal CT showed hepatic steatosis and a non-specific splenic lesion. His symptoms worsened over the next few days, and he was started on empiric antibiotic therapy. Trimethoprim-sulfamethoxazole and azithromycin were added for Pneumocystis and M. avium complex prophylaxis, respectively. He clinically improved but developed bulky left anterior axillary lymphadenopathy. Left axillary lymph node biopsy showed nodular sclerosis classical Hodgkin lymphoma. A bone marrow biopsy showed hypercellular marrow with megakaryocytic hyperplasia, T-cell infiltrate, and poorly formed granulomas, consistent with a severe inflammatory process. Several weeks later, a repeat bone marrow biopsy was performed to evaluate worsening pancytopenia, increase in ferritin to 18,800 ng/ml and EBV viremia to 1,039,888 copies/ml occurring in the setting of continued fevers, which revealed marrow involvement of his Hodgkin lymphoma and evidence of histiocytic phagocytosis concerning for HLH. His pancytopenia and hemophagocytosis were felt to be due to his lymphoma, and he was started on chemotherapy with doxorubicin/bleomycin/vinblastine/dacarbazine (ABVD). He completed six cycles of ABVD and follow up PET/CT scan showed no evidence of disease. Five months after completion of therapy he remains in clinical remission and his CD4 count has returned 444 (17%). His viral load remained suppressed throughout his chemotherapy.

Discussion: This case highlights the need for vigilance and close monitoring of HIV-positive patients with immunologic failure in the setting of suppressed viral load, as this can be a harbinger of Hodgkin lymphoma, and highlights the need for interdisciplinary management of complex patients.

References

Title: Emphysematous Gastritis: An Unusual Cause of an Unusual Diagnosis

Authors: Sara Robinson, MD, MS (Associate); Emily Ward, MD (Associate); Brett Sadowski, MD; Christa Eickhoff, MD; Patrick Young, MD, FACP, FACG, FASGE, MD; Edward Mitre, MD

Introduction: Emphysematous gastritis (EG) is a rare diagnosis with a high mortality rate. EG is defined by air in the stomach wall with systemic infection. We describe a case of EG that is unique in both the clinical stability of the patient and the proposed etiology of the infection.

Case Presentation: A 62 year-old woman with a history of poorly-characterized cyclic vomiting syndrome presented with 24 hours of nausea, vomiting, and diarrhea. She presented with tachycardia, fever, abdominal pain, and leukocytosis to 19x10^3/mcL. On admission, she was started on oral ciprofloxacin and metronidazole for possible bacterial gastroenteritis. Although the patient was clinically stable, on hospital day (HD) 2 the patient’s antibiotics were broadened to vancomycin, cefepime, and metronidazole because of persistent fever and the high leukocytosis. CT of the abdomen/pelvis revealed significant circumferential gastric wall thickening, edema, and hyper-enhancement of the mucosa with associated portal venous gas and gastric pneumatosis. Endoscopy was performed and revealed extensive necrotic tissue in the body of the stomach. Pathologic biopsy of the gastric mucosa exhibited suppurative gastritis with ulceration and necroinflammatory debris. Overall her clinical picture with the radiographic and endoscopic findings was consistent with the diagnosis of infectious emphysematous gastritis with associated necrosis of the stomach mucosa.

Surgical consultation was obtained and, due to a reassuring clinical presentation, the patient was managed conservatively with continued broad spectrum antibiotics. Tissue culture obtained during endoscopy grew Streptococcus mitis as well as normal flora and fungal culture was negative. Leukocytosis resolved by HD 5 and repeat imaging on HD 9 revealed marked interval improvement of the previously seen gastric wall thickening, edema, and hyper-enhancement as well as resolution of gastric pneumatosis and portal venous gas. The patient was transitioned to oral antibiotics and continued on a proton pump inhibitor. She was also initiated on prophylactic therapy for abdominal migraines which may have been the precipitating factor for the development of EG.

Discussion: In this case, we speculate that the patient developed mucosal barrier disruption in the setting of recent episodes of emesis and retching, and this served as a portal for polymicrobial bacterial infection of the gastric mucosa. The patient remained stable despite gastric pneumatosis, necrosis of the gastric mucosa visualized on endoscopy, and portal venous gas, which is usually an ominous finding consistent with bowel necrosis. We suspect that the rapid initiation of antibiotics in this case contributed to resolution without surgery. This case demonstrates that patients with EG that are clinically stable can, in some instances, be managed conservatively with broad spectrum antibiotics, serial imaging, endoscopy, and close monitoring by the gastrointestinal and surgical services. While EG is an uncommon diagnosis, recognizing potential risk factors such as cyclic vomiting may improve diagnosis in the appropriate clinical scenario.
Title: Cardiac Myxomas in Atypical Locations: A Case Series

Authors: Aaron Tallant¹, Zachary Junga¹, Zorana Mrsic¹, Geoffrey Cole¹

1.) Walter Reed National Military Medical Center, Bethesda, MD

Introduction: Primary tumors of the heart are rare (total incidence 0.1%) with myxomas making up the majority of benign cardiac tumors. Most myxomas are either located in the left atrium (75-80%) or right atrium (10-20%). Left ventricle myxomas are exceedingly rare, making up 1.7% of all cardiac myxomas. Here we present two cases of myxomas located in atypical locations with differing clinical presentations and outcomes.

Case Presentation: Patient 1: A 58 year old woman with a past medical history of hypertension presented to our hospital with profound dyspnea on exertion, palpitations, and lightheadedness. Acute coronary syndrome was ruled out and arrhythmias were not seen on telemetry. Transthoracic echocardiography (TTE) was performed and notable for a mass in the left ventricle (LV). Cardiac magnetic resonance imaging (MRI) demonstrated a 2 cm pedunculated mass in the LV with evidence of outflow tract obstruction. Surgery was recommended for removal. In the operating room, the mass was visualized in the LV attached to the papillary muscle and excised. Pathology confirmed the mass as a cardiac myxoma. The patient did well after surgery with marked improvement in her symptoms.

Patient 2: A 78 year old man with a past medical history of atrial fibrillation, on anticoagulation for a known left atrial appendage thrombus, and a previous ischemic stroke with residual motor deficits presented to our hospital with a two day history of left upper extremity incoordination and weakness. Physical exam was notable for new motor deficits from his baseline and a 2/6 early-peaking systolic murmur at the left upper sternal border. A head computed tomography scan without contrast demonstrated a new right frontal lobe ischemic stroke. A 1 cm mobile mass was noted in the left ventricle on TTE. Cardiac MRI demonstrated a 7 mm T2 hyperintense, pedunculated mass favored to represent a LV myxoma. Extensive coronary disease was found on cardiac angiography precluding the patient from surgery. He was medically optimized and discharged from the hospital in stable condition.

Discussion: Heart myxomas are most commonly located in the atrial septum near the fossa ovalis but can rarely occur in either ventricle. Symptoms of myxomas are predominately of cardiovascular and cerebrovascular origin but can vary significantly. Specifically with myxomas located in the left ventricle, patients are more prone to present with symptoms of systemic embolization due to chamber pressure and flow variation. Left ventricular myxomas can also cause tachyarrhythmias (most commonly atrial fibrillation) and LV outflow tract obstruction. Here, we present two cases of left ventricle myxomas with differing presenting symptoms, highlighting the above-mentioned clinical heterogeneity. Our case series
highlights the importance of echocardiography in diagnosing primary cardiac tumors and the utility of cardiac MRI in the diagnosis when surgical resection is not an option.

References


Title: TB or Not TB, That is the Question

Authors: Noopur Goyal, MD

Introduction: Clinical history plays an important role in consideration for Mycobacterium tuberculosis (TB) workup as incidence of TB is exceedingly low in the United States. Presentation can mimic many other disease processes including malignancy and autoimmune disease. Further, extrapulmonary TB is highly difficult to diagnose as sites of infection can vary including lymph nodes, pleura, or solid organ. High clinical suspicion can lead to appropriate workup to diagnose and treat extrapulmonary TB.

Case Presentation: An 82 year old Cambodian male with a history of hypertension and diabetes, presents with a six month history of fevers, chills, sweats, and weight loss. He notes painful axillary lymphedema that had developed over the last three months. A CT abdomen/pelvis showed widespread metastatic disease of the mesentery, omenta, and retroperitoneal lymph nodes, with concern for disseminated lymphoma. Given consolation of symptoms, further workup included negative HIV, hepatitis panel, EBV, and ANA. Surprisingly, patient had a positive RPR and Quantiferon Gold. Patient denied known TB exposure, and his last visit to Cambodia was twenty years ago. He further denied recollection of chancre, rash, and diagnosis or treatment for syphilis in the past. This became a curious case of disseminated lymphedema with workup to evaluate etiology of lymphoma versus tuberculosis, with concurrent syphilis findings. An excisional lymph node biopsy confirmed TB with PCR, and later grew positive fungal cultures, consistent with tuberculosis lymphadenitis. There was no evidence of underlying autoimmune disease or malignancy given peripheral smear and cytology, and no evidence of pulmonary TB on CT chest.

Discussion: This patient originally presented with concern for metastatic disease given his B-symptoms and significant metastatic findings on initial CT. Patients with these findings should be considered for TB workup in the setting of known risk factors including immune suppression, immigration status, and extremes of age. TB lymphadenitis is the most common presentation of extrapulmonary TB, often presenting as cervical lymphadenopathy, and can be diagnosed with FNA of lymph node or excision biopsy. Confirming diagnosis was important prior to initiating RIPE therapy given high rates of drug resistance. This patient also had findings of a positive RPR. Subsequently, testing indicated a positive confirmatory FTA, and a negative CSF RPR. Given asymptomatic presentation, with lack of suspected exposure in last twelve months, patient was diagnosed with late latent syphilis. He was started on Penicillin G IM, with total course of three injections, one per week, to complete treatment. This case demonstrates an interesting, atypical presentation of two infectious processes, both with longterm implications if undiagnosed, and untreated.

References


Vermont-Clinical Vignette-Poster Finalist
Louis-Bassett Porter, MD

Title: A “Retro” Case: Critically Advanced HIV/AIDS Presenting with Five Simultaneous Opportunistic Infections and Brain Lymphoma

Authors: Louis-Bassett Porter*, MD, Elena Kozakewich*, MD, Andrew Hale, MD

Introduction: The management of multiple simultaneous HIV/AIDS-related opportunistic infections and associated malignancies can represent profound clinical challenges. Until a cure for HIV is developed and widely implemented, knowledge of such diseases will continue to hold a fundamental place in general internal medicine.

Case Presentation: A 56 year-old man was found with tonic-clonic movements. He had complained of URI for several weeks, as well as altered mental status described by his husband as "slowed cognition, unlike himself". History was notable for hepatitis B (HBV) and HIV, acquired in the 1980s. He had previously taken HIV and HBV medications but had discontinued all medications three years prior, against medical advice. His CD4 T-cell count at that time was 250 cells/µL.

Vital signs were remarkable for temperature 38ºC and heart rate 127. He was cachectic and lungs were clear to auscultation bilaterally. Laboratory values revealed sodium of 114 mEq/L, lactic acid 7.5 mmol/L, hemoglobin 12.0 gm/dl, WBC count 5.8 K/µL, and platelets 143 K/µL. CD4 count was 19 cells/µL and HIV viral load was 468,999 copies/mL. Brain MRI showed a rim-enhancing lesion, 20x24mm, involving the right basal ganglia. Cytopathology from LP confirmed B-cell lymphoma.

Further work-up revealed positive hepatitis B surface antigen with HBV viral load >8.23 log IU/mL, mycobacterium avium complex (MAC) on blood cultures, positive Epstein Barr virus (EBV) in cerebrospinal fluid, and serum cytomegalovirus (CMV) viral load of 2770 IU/mL with fundoscopic findings of creamy exudates with associated dot-blot hemorrhages, consistent with CMV retinitis. PET scan showed strong fludeoxyglucose avidity within the CNS lesion, consistent with HIV-associated primary CNS B-cell lymphoma. BAL demonstrated Pneumocystis jiroveci pneumonia (PJP). Thus, this patient’s presentation was ultimately consistent with critically advanced AIDS with CD4 count of 19 cells/µL, active HBV, HIV-associated primary CNS lymphoma, PJP, CMV retinitis, and MAC bacteremia.

Treatment was initiated, but the patient had ongoing encephalopathy. By hospital day 36, he had not improved and his husband requested that all anti-infective medications be stopped. At the request of the patient’s husband he was discharged home with palliative care.

Discussion: For many clinicians in the US, treatment of advanced HIV/AIDS may be evocative of a past era. However, the CDC estimates that in 2014 over 1.1 million people in the US were living with HIV, and it remains a leading cause of death nationally. This case illuminates how the treatment of advanced HIV/AIDS can present profound treatment dilemmas. Numerous first-line therapies presented conflicts with the treatment of other concurrent disease processes. For instance, treatment of CNS lymphoma with rituximab and steroids would increase risk of fulminant HBV. Additionally, while initiation of ART leads to improvements in cellular immunity and significantly improves overall mortality, this patient’s multiple infections represented substantial risk of immune reconstitution inflammatory syndrome (IRIS).

References


Vermont-Clinical Vignette-Poster Finalist
Umer Syed, MD

Title: Thrombotic Thrombocytopenic Purpura Associated with Pazopanib

Authors: Umer Syed, Daniel R Douce, Julian R Sprague

Introduction: Thrombotic thrombocytopenic purpura (TTP) is a life-threatening thrombotic microangiopathy (TMA) characterized by platelet microthrombosis of the microvasculature due to decreased activity of the ADAMTS13 enzyme responsible for cleaving von Willebrand factor. Impaired ADAMTS13 activity can be due to hereditary enzyme deficiency or due to the acquisition of an auto-antibody inhibitor[1].

This has been differentiated from drug-induced TMA (DITMA) which is defined as microangiopathic hemolytic anemia, thrombocytopenia and microvascular thrombosis with characteristic vasculature abnormalities[2, 3]. DITMA has generally been defined to have two different mechanisms: immune-mediated and toxicity-mediated. In immune-mediated DITMA, antibodies are formed that bind with numerous cells including platelets, neutrophils, and endothelial cells and causes the adverse effect which is independent of dose and occurs within 2-3 weeks of drug exposure[4]. One example of this is TMA associated with quinine. In toxicity-mediated DITMA, there is a direct tissue injury from the drug with severity dependent on dose and route, typically occurring over weeks to months[5]. Chemotherapeutics may lead to TMA via this mechanism.

We present a case of a patient who presented with inhibitor-negative TTP following the initiation of a chemotherapeutic agent for renal cell carcinoma (RCC).

Case Presentation: A 76 year old male with metastatic renal carcinoma on day 24 of pazopanib was admitted with complaints of emesis, confusion and hematuria. Laboratory testing showed acute kidney injury, hyperbilirubinemia, and thrombocytopenia. Scattered schistocytes were seen on peripheral smear and he was diagnosed with thrombotic microangiopathy (TMA). He was started on daily, one-volume plasma exchange with rapid improvement in thrombocytopenia.

ADAMTS13 activity returned as undetectably low with no inhibitor detected. After cessation of plasmapheresis, repeat ADAMTS13 activity returned as normal. Unfortunately, his platelet count started to downtrend within four days after he developed septicemia thought to be due to a catheter-associated infection. He was placed on comfort care measures after discussion with his family. An autopsy listed the major cause of death as metastatic renal cell carcinoma.

Discussion: According to two separate systematic reviews, there have been no cases of proven drug-induced TMA where decreased ADAMTS13 activity was the identified mechanism. While pazopanib is also associated with TMA, this unique case suggests a novel potential mechanism for TMA associated with pazopanib and brings forth “drug-induced thrombotic thrombocytopenic purpura” that quickly responds to plasmapheresis as a possible new diagnostic entity requiring prompt recognition and treatment.
References


Virginia-Clinical Vignette-Poster Finalist
Mai Abdelnabi, MD

Title: A fatal case of disseminated Mycobacterium bovis infection involving the kidneys and a prosthetic hip after therapeutic BCG administration

Authors: Mai Abdelnabi, MD; Daniel J. Stein, MD; Luke Luetkemeyer, MD; Quanjun Cui, MD; Christopher Moore, MD

Introduction: Bacillus Calmette-Guérin (BCG) is first-line therapy for the treatment of superficial bladder cancer. Although generally well tolerated, BCG-related systemic infectious complications can occur.

Case Presentation: A 76-year-old man with a history of bladder cancer (s/p resection and 11 BCG treatments) and total hip arthroplasty was transferred to our institution for evaluation of possible prosthetic joint infection. He was initially admitted to a local facility due to altered mental status and fevers. His fever was eventually attributed to C. difficile colitis, after positive PCR testing, and he was discharged to a rehabilitation facility on oral metronidazole. He then developed recurrent fevers and was re-admitted to the outside institution where a psoas abscess was identified on imaging. Given concern for possible communication between the psoas abscess and the hip prosthesis, he was transferred to our institution for further management. On admission, bacterial and mycobacterial blood cultures were obtained. Hip aspiration was obtained out of concern for joint prosthesis involvement. Given our clinical concern for disseminated BCG, we sent fluid for acid fast bacilli (AFB) staining which revealed 1+ AFB, mycobacterial culture which was ultimately identified by the Centers for Disease Control as M. bovis, and a direct M. tuberculosis complex polymerase chain reaction (PCR) probe which was positive. He subsequently underwent operative removal of his infected hardware; his operative findings demonstrated chronic infection with granulation tissue and gross pus in the hip and pelvis behind the acetabular cup and lateral to the iliactus muscle. Pathology revealed marked acute inflammation. While no acid-fast bacilli were identified on the smear, cultures ultimately grew AFB. Given his prior BCG treatment, he was treated for disseminated M. bovis infection with rifampin, ethambutol, and isoniazid immediately post-operatively. Unfortunately, his mental status continued to decline and he was transferred to the ICU. After a discussion of his goals of care with the patient and his family during a period of lucidity in the ICU, he was transitioned to inpatient hospice. He expired approximately two weeks later. An autopsy was conducted and demonstrated disseminated mycobacterial infection as the final cause of death. The anatomic exam demonstrated a 5 x 4 cm abscess involving the left psoas muscle and extending to the left iliac bone with microscopic granulomas noted in the bilateral kidneys, liver, and prostate. Microscopic examination was notable for scattered small and occasionally necrotizing granulomas in the renal cortex. Organisms were identified within a granuloma on Fites stain consistent with his known M. bovis infection.

Discussion: This is a case of disseminated BCG with M. bovis identified in the renal parenchyma, combined with the atypical combination of renal, prosthetic joint and musculoskeletal involvement. It highlights the importance of prompt recognition and management of serious infectious complications related to BCG therapy.
Virginia-Clinical Vignette-Poster Finalist
Mariam A Assi, MD

Title: Leukocytosis in a Patient with Shock: a Lesson in Antimicrobial Stewardship

Authors: Mariam Assi, Department of Internal Medicine, Virginia Commonwealth University, Richmond, VA

Introduction: Extreme leukocytosis, otherwise known as a leukemoid reaction, is often the harbinger of a hematologic malignancy, but not always. The diagnostic challenge lies in the few cases where a leukemoid reaction is caused by a benign process.

Case Presentation: This is the case of a 60-year-old African American female with marked leukocytosis in the setting of cardiogenic shock. This patient had a history of viral myocarditis one year earlier, followed by development of cardiomyopathy that led to severe heart failure. Long-term inotropic support had been initiated as a bridge to anticipated heart transplantation. Patient’s condition had been relatively stable until she presented to us in acute cardiogenic shock; an echocardiogram revealed acute mitral regurgitation secondary to chordae tendineae rupture as the cause. On admission, patient’s white blood cell count showed a new leukocytosis of 15 x 10^9/µL. Evaluation of peripheral blood smear revealed a predominance of mature neutrophils at 90% with left shift, and absence of basophilia, eosinophilia, monocytosis and immature cells. There were no red blood cell or platelet abnormalities. Patient was afebrile and had no focal symptoms or signs of infection. Standard infectious workup, including blood and urine cultures, chest radiograph, and polymerase chain reaction (PCR) testing for Clostridium difficile, did not reveal an infectious cause. In the absence of an alternative explanation, it was assumed that the patient had an occult infection, and she was maintained on broad spectrum antibiotics from the day of admission. The patient’s only home medications were dobutamine and furosemide, both of which she had been taking for a few months prior to admission with normal white blood cell counts documented during that time. Patient’s white blood cell count continued to rise despite antibiotics, reaching 58 x 10^9/µL over the course of a week. Meanwhile, patient’s cardiogenic shock had failed to respond to medical support. Antibiotics were discontinued as our differential diagnosis shifted toward either a myeloproliferative neoplasm or a leukemoid reaction to physiologic stress. Cytogenetic analysis of peripheral blood did not reveal a BCR-ABL fusion gene or a JAK2 mutation, rendering a myeloproliferative neoplasm unlikely. Leukocytosis resolved in parallel with improvement in patient’s clinical condition after initiation of mechanical cardiac support with an intra-aortic balloon pump, followed shortly by a left ventricular assist device. The clinical course was consistent with a leukemoid reaction secondary to cardiogenic shock.

Discussion: It is important that the internist be cognizant of the non-infectious causes of leukemoid reactions that have been described in the literature. These include paraneoplastic phenomena, certain medications, and conditions associated with inflammation, ischemia and extreme physiologic stress. The importance of prompt recognition of these causes lies in avoiding unnecessary use of antibiotics, which translates to improved antimicrobial stewardship among internists.
Virginia-Clinical Vignette-Poster Finalist
Mariam A Assi, MD

Title: Emerging Pathogens in the Immunocompromised Host: a Case of Disseminated Disease with Graphium species

Authors: Mariam Assi, Department of Internal Medicine, Virginia Commonwealth University, Richmond, VA, Oveimar De La Cruz, Division of Infectious Diseases, Virginia Commonwealth University, Richmond, VA

Introduction: In the current era of chemotherapy and immunosuppressive therapy, microorganisms previously not known as pathogens are gaining recognition as causes of severe infection, particularly in the immunocompromised host. We describe a case of invasive infection with Graphium species manifesting as skin and soft tissue infection with high clinical suspicion for dissemination to the lungs in an immunosuppressed patient with acute myeloid leukemia.

Case Presentation: A 27 year old man with acute myeloid leukemia was admitted for induction chemotherapy. While neutropenic, patient was on prophylactic levofloxacin, acyclovir and fluconazole. Around day +26, he developed a violaceous non-tender lesion, with erythematous margins, on his right thigh. A punch biopsy of the skin lesion showed yeast and pseudohyphae in the deep dermis with clots and edema suggestive of invasive fungal infection. Workup including serum (1-3)-β-D-glucan and galactomannan antigen testing, CT of the chest and sinuses and MRI of the thigh showed no evidence of disseminated disease. Voriconazole (VOR) was begun on day +34. The lesion progressed rapidly with development of an ulcerated center, prompting switch to posaconazole (POS) on day +37. Tissue from the biopsy was insufficient for culture, prompting surgical debridement on day +42. Fungal blood culture obtained on day +35, and deep tissue culture obtained surgically on day +42, grew Graphium spp. Patient achieved neutrophil count recovery on day +41 and was discharged on day +44 on POS.

Patient was readmitted on day +84 with respiratory distress and febrile neutropenia, still on POS. Chest CT revealed ground glass opacities in both upper lung lobes. Cultures grew ESBL-producing Klebsiella pneumoniae in sputum and Streptococcus viridans of intermediate susceptibility in blood. Patient was maintained on meropenem and vancomycin respectively. Fevers and respiratory distress continued despite broad spectrum antimicrobial coverage. Chest CT on day +98 showed interval worsening of ground glass opacities, now involving all lobes. Bronchoscopy with bronchoalveolar lavage revealed no macroscopic airway lesions; bacterial, fungal, viral and AFB cultures never grew any organisms. On day +100, antifungal susceptibilities of the Graphium species grown in blood and soft tissue cultures from previous hospitalization, were finally reported: MIC 0.5 mcg/ml for amphotericin B, MIC above 16 mcg/ml for flucytosine and all azoles tested. Patient was switched from POS to amphotericin B (AMB) with subsequent clinical and radiologic improvement.

Discussion: Most clinical isolates of Graphium spp. are synanamorphic forms of Pseudallescheria boydii or secondary forms of Scedosporium apiospermum. Severe infections have been described in immunocompromised hosts, including fungemia, mycetoma, pneumonitis, osteomyelitis, arthritis, meningitis, brain abscess, endocarditis, thyroid abscess and cutaneous and subcutaneous granulomas. Our case adds to the evidence that this pathogen can prove challenging to treat owing to its wide resistance pattern. Surprisingly, however, our isolate was susceptible to AMB, which is often reported resistant.
Title: An Unusual Cause of Acute Heart Failure: A Case Report of Iliocaval Venous Stent Migration

Authors: Sherif Elmahdy, MD, Muhamad Alhaj Moustafa, MD

Introduction: The reported incidence of stent migration as a complication of endovascular stenting is reported to be as low as 3%. Majority of cases reported with stent embolization to the right heart were from the central thoracic veins, dialysis fistulas, and renal veins. Stent migration to the right heart from iliac veins occurs more seldom. Endovascular extraction is usually the initial intervention with surgery reserved for complicated cases.

Case Presentation: A 61-year-old female with a medical history of hypertension, diabetes mellitus type 2, and CKD V initially presented to an outside facility with progressive dyspnea on exertion, orthopnea, and bilateral lower extremity edema. She was diagnosed with new onset heart failure and treated with diuretics. A transthoracic echocardiogram revealed an intracardiac foreign body. She was subsequently transferred to our institution for further management. Upon transfer to our facility a transesophageal echocardiogram revealed a long stent straddling the tricuspid valve from the right atrium with the other end lodged in the trabeculation of the right ventricle with severe tricuspid regurgitation. We learned that the patient suffered from May-Thurner syndrome and had undergone peripheral endovascular intervention with placement of a self-expanding Nitinol Protege (14mm x 60mm) stent to the left iliac vein six months prior. A percutaneous endovascular approach with a Gooseneck snare was attempted to retrieve the migrated stent. However, the stent fractured leaving behind two fragments. After ensuring tricuspid valve integrity with intracardiac ultrasound, the patient was referred for surgical extraction. During the operative procedure the stent was found to be densely adherent to the tricuspid leaflets and the subvalvular apparatus, with majority of the primary chords to the anterior and posterior leaflets ruptured. She underwent valvuloplasty with a 29mm Carpentier-Edwards bioprosthetic valve. Post-operative echocardiogram revealed trace tricuspid regurgitation.

Discussion: May–Thurner syndrome is an acquired stenosis of the left common iliac vein. It results from compression of the vein against the lumbar vertebrae by the overlying right common iliac artery. It can cause pain, edema, or deep venous thrombosis mostly in middle-aged women. Balloon dilation with stenting is superior to conventional surgical treatment and should be considered first line therapy. Intravascular stents can migrate into cardiac cavities or pulmonary arteries when they are deployed in the venous system. These complications might be due to stent undersizing, increased venous vascular compliance in comparison to the arterial system, the natural venous flow from smaller to larger diameter vessels, and the possibility of arterial pulsatility over the vein causing stent dislodgement. Even though stent migration is a rare occurrence, embolization to the right atrium, ventricle, and pulmonary circulation can be catastrophic. Standardized sizing techniques as well as serial imaging should be considered to confirm adequate long-term placement and to avoid life-threatening outcomes.
Virginia-Clinical Vignette-Poster Finalist
Nicole Habel, MD

Title: Unusually large exophytic penile mass

Authors: Nicole Habel, MD; Carilion Clinic and Virginia Tech Carilion School of Medicine, Roshan Bhowansingh, MD; Salem VA Medical Center and Virginia Tech Carilion School of Medicine

Introduction: Penile cancer is an uncommon cancer in North America and Europe with an incidence of < 1 per 100,000 and its diagnosis is often delayed due to embarrassment, fear and personal neglect. Variants, such as Buschke-Löwenstein tumor and leiomyosarcoma must be considered, particularly when the mass is slow growing and has a bulky appearance.

Case Presentation: An 87 year old Caucasian male presented to the hospital for respiratory complaints and was found to have an exophytic mass involving the entire penile circumference and shaft, which had been slowly progressing over the previous 12 months. The tumor was dense, rubbery and irregular with a necrotic area of 2.8cm x 2cm at the glans penis and smaller shallow ulcerations at the shaft and scrotum. The mass was accompanied by erythema of the groin and scrotum, however warmth or tenderness to suggest superficial infection were absent. No inguinal lymph nodes were palpable. Broad-spectrum antibiotics administered for unrelated reasons did not alter overall appearance. Buschke-Löwenstein tumor, a pre-malignant verrucous tumor, was considered in the differential diagnosis given its slow growth, large and infiltrative appearance as well as locally destructive behavior. However, a punch biopsy revealed common squamous cell carcinoma, which is the most frequent histologic subtype of penile carcinomas with HPV 16, 6 and 18 as likely carcinogens in approximately one third of cases. The patient declined surgical intervention or radiation treatment. A suprapubic catheter was placed with palliative intent in anticipation of eventual complete urethral obstruction.

Discussion: While penile cancer is an infrequent malignancy it is associated with devastating morbidity and high mortality once metastatic spread has occurred. Risk factors for penile cancer include HPV infection, lack of circumcision, poor hygiene, phimosis and smoking, suggesting that inflammation in addition to oncogenes plays a critical role in tumor carcinogenesis. The majority of penile cancers are comprised of squamous cell carcinoma with long-term prognosis being dependent on location, stage and grade. Treatment modalities are aimed at tumor removal with organ preservation. Superficial non-invasive disease can be treated with topical chemotherapy, such as 5-fluorouracil or laser therapy. Invasive disease on the other hand requires surgical resection or radiation therapy. Lymph node involvement necessitates multimodal treatment (surgery with either adjuvant or neo-adjuvant chemotherapy) as long-term survival is drastically reduced in advanced disease.

This case brings to light the importance of early diagnosis and treatment in order to avoid significant morbidity and mortality. Creating a supportive and judgement free environment for discussing men’s health issues in the primary care setting is a vital task for the present and future.
Title: Paraneoplastic Cushing’s Syndrome in a Hypertensive, Hypokalemic Patient

Authors: Elora Majumder, MD¹, Shant Ayanian, MD¹, ¹Department of Medicine, George Washington University Hospital

Introduction: Paraneoplastic Cushing’s Syndrome is a rare but well known complication of certain neuroendocrine tumors that can be difficult to diagnose. Through this case, the reader will learn how to diagnose paraneoplastic Cushing’s syndrome in patients with persistent hypertension and hypokalemia, and how to distinguish it from other causes of secondary hypertension.

Case Presentation: A 61 year old woman presented to the hospital with dyspnea and chest pain. She had been gaining weight, and experiencing night sweats and lower extremity swelling for several months. She had a 30 pack year smoking history. Physical examination revealed fingernail clubbing, 1+ pitting edema, and a blood pressure of 220/97—despite taking Hydralazine and Spironolactone for months. Labs revealed a potassium level of 2.6 mmol/L, a bicarbonate level of 31.0 mmol/L, a hemoglobin of 15.2 gm/dL, and a WBC count of 15.66 x10³ cells per µl. A PET/CT scan showed a suspicious mass that was hypermetabolic, and a bronchoscopy with biopsy revealed small cell carcinoma of the lung that was already metastatic.

Despite treatment, her hypertension and hypokalemia persisted. A urine potassium level of 30.7 mmol/L indicated renal potassium wasting. An abdominal MRA showed patent renal arteries, ruling out renal artery stenosis. Normal renin and aldosterone levels ruled out primary aldosteronism. However, cortisol levels were elevated at 67 mcg/dL, prompting a 24-hour urinary-free cortisol excretion, which showed a urinary cortisol level of 3336 mcg/24 hr. An overnight dexamethasone suppression test showed persistent high morning cortisol levels. These tests, coupled with the patient’s persistent hypertension and known cancer prompted a diagnosis of paraneoplastic Cushing’s syndrome.

Discussion: Ectopic adrenocorticotropic hormone production is a rare cause of Cushing’s syndrome, and has been found in approximately 1%-5% of small cell lung cancers.¹ Although not a classic symptom of Cushing’s syndrome, hypokalemia can affect up to 57% of patients with Cushing’s syndrome, as excessive cortisol can act as a weak mineralocorticoid.² The initial work-up of hypokalemia includes obtaining a urine potassium level, which can distinguish urinary potassium losses from others. If urinary potassium is inappropriately increased, then hypertension may suggest a primary or secondary hyperaldostone state.³ Obtaining renin and aldosterone levels can help in further classification. If renin and aldosterone levels are normal, one should investigate serum cortisol levels. Elevated cortisol levels warrant further work-up, including a 24-hour urinary cortisol collection and a dexamethasone suppression test to confirm the diagnosis of paraneoplastic Cushing’s syndrome.

In a patient with hypertension and malignancy with evidence of metastasis, the differential diagnosis is broad, including renovascular disease, a renin secreting tumor, primary aldosteronism, and Cushing’s syndrome. Diagnosing paraneoplastic Cushing’s syndrome early is important as it alters the patient’s prognosis and requires additional treatment.

References


Title: Anemia without Actual Anemia... a very Pernicious Process

Authors: James Mendoza, MD, Department of Internal Medicine, Eastern Virginia Medical School, Ian Chen, MD, Professor of Medicine, Department of Internal Medicine, Eastern Virginia Medical School

Introduction: In the 1840s, Dr. Thomas Addison described “a slow and insidious pallor” where “the mind occasionally wanders... the debility becomes extreme... and at last (he) expires.” Addison’s anemia, commonly known as pernicious anemia, is an auto-immune atrophic gastritis characterized by the presence of anti-intrinsic factor antibodies. The resulting decreased cyanocobalamin absorption manifests as psychologic, neurologic, and hematologic symptoms. Even in the absence of hallmark findings, failure to include the disease in differential diagnosis can be as harmful as its name suggests.

Case Presentation: A 52 year-old white male with a PMHx of remote alcohol and drug abuse presents with progressively worsening confusion, forgetfulness, and ataxic gait for one month. Family members said his gait was shuffling, as if the feet were heavy, experiencing several falls. The patient related it to walking a tightrope. Vital signs were normal. On physical examination, he appeared tremulous and was oriented only to person. Cerebellar exam, including rapid alternating movements and finger-to-nose testing, were impaired. His gait was broad-based with outward rotation of the feet. Vibration-sense in the distal lower extremities was markedly diminished. There was no glossitis. CT scan of the head was normal as well as full chemistry panel, including sodium. CBC showed a hemoglobin of 17.1 g/dL and MCV of 88.1 fL. Cerebrospinal fluid analysis was unremarkable. Urine drug screen, RPR and HIV screen were negative, and serum ethanol, ammonia, TSH, and folate were normal. Cyanocobalamin (Vitamin B12) was 226 pg/mL (reference range 221-911). The patient was given 1000 µg of intramuscular Vitamin B12 daily for three consecutive days, and on hospital stay day 5, his symptoms improved enough for discharge. After discharge, intrinsic factor antibody returned positive, and on follow-up phone call two weeks later, the patient had reported almost complete resolution of memory and gait symptoms.

Discussion: Pernicious anemia can occur without anemia or macrocytosis, and may only manifest as neurological symptoms. Others have shown this anomaly in 28% of a small subset of patients with cyanocobalamin deficiency. Furthermore, it can present with only borderline-low Vitamin B12 levels. Diagnosing the disease is no longer as cumbersome as the Schilling test—the intrinsic factor antibody assay provides an efficient and convenient means for diagnosis. As such, in the appropriate setting, clinicians should not hesitate to test for pernicious anemia. Indeed, Dr. Addison’s description of his patient teaches us that a keen physical exam may surprisingly outweigh laboratory findings.

References

Title: Bloody Bladder: Where Do We Go from Here?

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Introduction: Microscopic hematuria is a common incidental finding on routine urinalysis. Common causes include renal disease, infection, medications, and malignancy. The prevalence of urologic malignancy in patients with microscopic hematuria ranges widely from 0.19% to 16.1%, depending on the age, sex, and risk factors of those studied.¹ The indications for further evaluation of microscopic hematuria are not well-defined, and ultimately, 40% to 90% of these patients are not referred for further evaluation.² We report the case of a man who presented with hematuria and was subsequently found to have epithelioid angiosarcoma of the bladder.

Case Presentation: A 57 year-old healthy Asian man presented to a Traditional Chinese Medicine practitioner with a four-week history of hematuria. He denied a history of smoking, malignancy, or exposure to chemicals or radiation. He received a herbal medicine, but over the next three weeks, experienced increasingly painful voiding episodes with gross hematuria, prompting admission to the hospital. Physical exam was notable for pallor, cachexia, and bilateral lower extremity edema. Abdominal exam was unremarkable. Laboratory Results: were notable for hemoglobin 6.1 g/dL, sodium 129 mEq/L, creatinine 1.4 mg/dL, protein 5.6 g/dL, and albumin 2.7 g/dL. Liver function tests and coagulation studies were unremarkable. Microscopic urinalysis showed “large” blood, too numerous to count red blood cells per high powered field (HPF), and 11-25 white blood cells per HPF. CT of the abdomen/pelvis revealed a mass in the left upper renal pelvis extending to the renal pole and proximal ureter and bladder expanded with blood clots. Bone scan showed a metastatic focus in the ischium. He underwent a partial cystectomy, with pathology revealing muscle-invasive epithelioid angiosarcoma. He elected to pursue palliative chemotherapy and passed away several months later.

Discussion: Microscopic hematuria may be seen in 9% to 18% of healthy individuals in a normal population.¹,⁵ The prevalence of urologic malignancy in those with microscopic hematuria is variable. Consequently, the indications for further evaluation are controversial.

Initial evaluation should involve ruling out benign causes of hematuria. Risk factors for urologic malignancy including male gender, age greater than 35 years, occupational exposures to chemicals/dyes, or history of smoking or pelvic irradiation should be noted.⁴ Once benign causes have been excluded, average-risk patients should undergo evaluation with CT urography.¹,²,⁴ Higher-risk patients should also undergo cystoscopy.⁴

This case highlights the importance of recognizing when further evaluation of microscopic hematuria is warranted. Although our patient was a healthy man, his gender and age should have prompted further evaluation of any microscopic hematuria seen prior to the development of gross hematuria. Microscopic hematuria is commonly encountered in practice and may be an early manifestation of urologic malignancy. Due to the increased mortality in the advanced stages of urologic cancers, early detection, beginning with appropriate evaluation, is of paramount importance.²

References


Title: Pain in the Neck: A case of IgG4-Related Disease

Authors: Ivan Pena

Introduction: IgG4-related disease is a rare immune-mediated condition with a multitude of presentations, including pancreatitis, kidney dysfunction, pneumonitis and generalized lymphadenopathy. This case highlights an uncommon presentation of this condition: tender cervical lymphadenopathy.

Case Presentation: A 44 year-old African-American man presented to his primary care physician with a five day history of neck swelling and pain associated with chills, night sweats and weight loss. Three days prior to presentation, he was evaluated at a nearby emergency department and prescribed amoxicillin-clavulanic acid without improvement. Physical and laboratory examinations were normal except for four round, tender, right-sided cervical mobile masses. Computed tomography(CT) of the neck with intravenous contrast revealed bilateral, right greater than left cervical adenopathy consistent with possible lymphoma. Flexible fiberoptic laryngoscopy was normal and a fine needle aspiration was inconclusive. He completed a second course of antibiotics with trimethoprim-sulfamethoxazole, again without improvement. Given concern for malignancy, a positron emission tomography(PET) scan was performed and revealed fluorodeoxyglucose uptake in bilateral cervical, axillary and external iliac nodes. As a result, he underwent excisional lymph node biopsy and pathology was consistent with reactive lymphadenopathy without evidence of malignancy. He subsequently underwent an infectious workup which was negative. During his extensive workup, patient developed symptomatic anemia, acute kidney injury and a decrease in his serum albumin. Given his negative malignancy and infectious workup, autoimmune disease was considered. Further staining of the biopsy samples was performed, specifically for IgG4 and HHV-8. HHV-8 staining was negative, however there were greater than 50% IgG4 cells in the sample. Laboratory examination was also notable for an elevated serum IgG4 level. A diagnosis of IgG4-related disease was made and he was initiated on steroids, a mainstay in the treatment of this condition. On a subsequent visit, his symptoms had improved, but unfortunately, patient was lost to follow up.

Discussion: This case illustrates the importance of including IgG4-related disease in the differential diagnosis of tender cervical lymphadenopathy. This is most commonly caused by local head and neck pathology such as infections, or systemic pathology such as tuberculosis or malignancy. With prompt diagnosis, end organ dysfunction and morbidity would have been avoided. This case also illustrates a physician’s duty to constantly re-evaluate a case, particularly when a patient is not responding to what is presumed to be appropriate therapy. This patient was initially treated with antibiotics for presumed local infection, however, after minimal response to therapy, workup was continued, the correct diagnosis was made, and appropriate therapy was initiated.
Virginia-Clinical Vignette-Poster Finalist
Zahra Rehman, MD

Title: Oh My-"Eye"-Loma!

Authors: Zahra Rehman, MD and Ian Chen, MD - Eastern Virginia Medical School, Norfolk, VA

Introduction: Extramedullary spread in multiple myeloma can either result in plasma cell leukemias or soft-tissue plasmacytomas (1). Soft-tissue plasmacytomas are most often found in the head and neck region, but may also present in the GI tract, bladder, CNS, thyroid, breast, testes, parotid gland, lymph node, skin, and rarely eyes. These tumors can present at any time during the course of multiple myeloma and account for 3% of all plasma cell tumors (2). The incidence of extramedullary plasmacytomas is 7% to 18% at the diagnosis of multiple myeloma and risk escalates to 20% at relapse (1). Diagnosis is made by biopsy exhibiting clonal plasma cells. Although multiple myeloma and plasmacytomas are made up of identical cells, treatment differs. Treatment for plasmacytomas includes systemic steroids with surgical resection and/or radiation therapy. Chemotherapy does not improve relapse rate or increase disease-free survival in plasmacytomas as it does in multiple myeloma.

Case Presentation: This patient was a 45 year old Caucasian male with a previous history of IgG lambda multiple myeloma diagnosed in 2013 with a history of testicular plasmacytoma s/p orchiectomy and left clavicle plasmacytoma s/p radiation. He had previously been treated with six cycles of chemotherapy (Thalidomide, Dexamethasone, Velcade) in 2013 and in 2016 with CyBord and VDT-PACE. At our facility, he presented with bilateral eye swelling, erythema, and blurry vision for about 3 weeks. Prior to admission, it had been treated as conjunctivitis with steroid and decongestant eye drops with no improvement. On admission, CT scan showed temporal mass and orbital soft tissue swelling and proptosis. This was followed by MRI head (See Picture), which revealed multiple bilateral temporoccipital and intra-orbital enhancing masses with concern for metastases. He was started on steroids and IV/topical antibiotics without improvement. Ultimately, orbital biopsy was done which indicated plasmacytoma.

Discussion: Our patient presented with orbital plasmacytoma, which had never been witnessed by the ophthalmologist and oncologist at our facility. The patient had worsening proptosis during hospitalization (See Picture) and eventually transitioned to hospice care due to no response to treatment. If the plasmacytoma had been suspected earlier and steroids had been started upon presentation, the course of the disease may have been different. It is important to consider plasmacytomas when pathology of the eye presents in a patient with a history of multiple myeloma.

References

Title: Omalizumab Anaphylaxis in Chronic Urticaria and Systematic Desensitization

Authors: Matthew D. Straesser, MD and Timothy T. Kyin, MD, 1Department of Medicine, 2Division of Allergy and Clinical Immunology, University of Virginia Health System, Charlottesville VA

Introduction: Omalizumab is a monoclonal IgG directed against the Fc receptor of IgE. Omalizumab was first approved by the FDA for the treatment of moderate-severe asthma in 2003 and for chronic urticaria in 2014. Although documented, anaphylaxis is rare with an estimated 0.1% occurrence. Considering that omalizumab can be highly effective in controlling disease symptoms, drug desensitization even in the setting of anaphylaxis could potentially be beneficial. Previous reports of omalizumab desensitization are very limited and not in the setting of anaphylaxis. Dreyfus and Randolph reported a successful premedication with NSAIDS to abate an omalizumab anaphylactoid reaction. Owens and Petrov reported 3 cases of omalizumab desensitization using an escalating desensitization protocol; however, the reactions were hypersensitivities and not anaphylactic in nature. Here, we report a successful omalizumab desensitization in a 21 year old woman who previously experienced omalizumab-induced anaphylaxis. To our knowledge, this is the first report of omalizumab desensitization in the setting of a prior anaphylaxis.

Case Presentation: We present a 21 year old woman with a past medical history of Ehlers-Danlos syndrome, irritable bowel syndrome, polycystic ovary syndrome, interstitial cystitis, mast cell activation syndrome, and chronic urticaria who presented to the allergy clinic for her chronic urticaria. She was refractory to high-dose antihistamines and was prescribed omalizumab 300 mg. Within 30 min of the first omalizumab injection (without premedication), she developed hives, lip angioedema, wheezing, hypotension, and tachycardia. Epinephrine 0.3 mg was administered IM and her anaphylaxis resolved. She experienced rebound anaphylaxis 12 hours later and required a brief observation admission with a steroid taper. Her chronic urticaria continued to persist and omalizumab desensitization was sought 3 months later in the outpatient allergy clinic. An 18 gauge IV was placed and she was administered omalizumab subcutaneous every 30 minutes at the following escalating intervals: 1.25 mg, 2.5 mg, 5.0 mg, 10.0 mg, 12.5 mg, 18.75 mg, 25.0 mg, 31.75 mg, 43.75 mg, 50.0 mg, 100 mg, 150 mg. After reaching the target dose of 150 mg, she transition to 150 mg every 14 days and then finally to 300 mg every 30 days. She tolerated the desensitization well without any complications or drug reactions. After 4 months of omalizumab therapy, her chronic urticaria symptoms had significantly improved.

Discussion: Omalizumab is a highly effective treatment for moderate-severe asthma and chronic urticaria; however, it carries a small risk of anaphylaxis. Our case demonstrates that omalizumab hypersensitivities can systematically be desensitized, even in the setting of anaphylaxis, and may potentially serve as a model for monoclonal antibody desensitization.
Introduction: Melanoma is the sixth leading malignancy. Melanomas frequently metastasize to the lung, presenting as multiple nodules. Primary pulmonary melanoma (PPM), usually presents as a single mass like lesion and often in an advanced stage upon diagnosis. Solid melanoma of the pancreas is almost always a metastatic condition. In this case we have a patient who presented with probable PPM metastasize to the brain and pancreas.

Case Presentation: A 69-year-old male with history of prostate adenocarcinoma presented initially with left sided hemiparesis. On CT head he was found to have right basal ganglia and internal capsule hemorrhagic stroke. Admission CXR showed RLL lung mass. Further work up showed, 5cm pleural based RLL lung mass, a 1.5cm pancreatic mass in the uncinate process, and multiple mass with sub-acute hemorrhage concerning for metastases to the brain.

Subsequently CT guided Lung biopsy findings were consistent with melanoma. To differentiate the pancreatic uncinated process mass from a melanoma metastatic deposit versus incidental primary pancreatic mass, he underwent EGD with FNA. The neoplastic cells were found to have similar morphology, as the lung biopsy.

A vigorous search for a primary skin/mucosal melanoma did not reveal any suspicious lesions and given no prior history of treated melanoma, our patient was diagnosed with probable metastatic PPM. Subsequently, he underwent radiation to the brain, and started on immunotherapy with Nivolumab, followed by Pembrolizumab and Ipilimumab.

Discussion: PPM usually presents as a single mass like lesion, estimated to be less than 0.01% of all lung cancers and 4% of all melanomas. Airways as well as parenchyma and pleura can be involved. Roughly about a third of these patients have no pulmonary symptoms at time of diagnosis. Our patient had only a single lung lesion that is extremely rare as a metastatic lesion. The 5-year survival of PPM is about 10%. However, case reports with successful surgical resection of a single PPM, have been reported. Lobectomy and or pneumonectomy are preferred over tumor resection with clear margin to avoid recurrence. Surgical resection is not recommended in patients with advanced metastatic lung disease, and our patient was not a surgical candidate.

Since the introduction of immune checkpoint inhibitors, there have been a growing interest in using them for treatment of metastatic melanomas, currently at least there are 7 clinical trials. To our knowledge, there is no clinical trial targeting non-cutaneous metastatic melanomas. Our patient was started on immunotherapy with Nivolumab, followed by Pembrolizumab and Ipilimumab. Our case although involving only one patient it seemed to show a benefit in treatment with PD-1 and CTLA4 inhibitors. At the time of writing this case report, over 24 months after diagnosis, patient still alive and able to perform daily activities. In conclusion, patients with PPM who are not surgical candidate should receive PD-1 inhibitor or a combination of PD-1 and CTLA4 inhibitor as it may improve survival.
Title: Statin-Induced Necrotizing Autoimmune Myopathy as an emerging cause of proximal muscle weakness

Authors: Douglas D Thaggard, Internal Medicine Resident, University of Virginia Medical Center

Introduction: Statin Induced Necrotizing Autoimmune Myopathy (SINAM) is a rare but emerging toxicity related to cholesterol-lowering therapies in the statin pharmacologic class. Distinct from the more common statin myopathy, which is hypothesized to be related to the reduction in ubiquinone, coenzyme Q10, and muscle energy production, this type of myopathy occurs in less than 0.1% of patients. Further, symptoms and morbidity are more striking in SINAM, typically with progression despite removal of the offending medication. Treatment involves immunosuppression, and prognosis is often guarded with frequent relapse.

Case Presentation: A 51 year old male with past history of coronary artery disease with prior MI on statin therapy (self-discontinued 6 months prior) presenting with subacute, symmetric, painless, proximal muscle weakness primarily affecting his shoulder and hip girdles. Examination revealed an ability to move all limbs against gravity but not examiner resistance. He was not able to rise from a seated position by any means on his own power. Notable presenting labs include an AST/ALT of 433/660 as well as a CK of 32,000. Standard autoimmune serology was unremarkable for any autoimmune process, and the patient underwent muscle biopsy, which was consistent with severe, chronic necrotizing and inflammatory myopathy. Given his history and lack of clear diagnosis, an anti HMG CoA reductase antibody was sent which was strongly positive at >200 units (normal range: <20 units). Steroid therapy was started without any appreciable improvement in symptoms. IgG therapy was deferred given patient preference (Jehovah’s Witness), and the patient underwent successful treatment with rituximab and azathioprine with improvement in symptoms.

Discussion: Statin Induced Necrotizing Autoimmune Myopathy is important and likely under-diagnosed cause of proximal muscle weakness related to statin therapy. This case represents a typical presentation of this condition with onset of muscle weakness months after initiation of statin therapy with progression despite discontinuation of said therapy. Steroid therapy alone is often not sufficient to reverse symptoms, and intravenous immunoglobulin seems to have a beneficial role in conjunction with immunomodulating therapies. Because of the clear mortality benefits of statin therapy, this pharmacological class of medications has become commonplace, almost ubiquitous. For this reason SINAM is an important clinical condition to recognize with potentially devastating side effects and should serve as a reminder to clinicians to continue discussing potential adverse effects of all new medications before starting therapy.

References:
West Virginia-Clinical Vignette-Poster Finalist
Mohammed A Alshehri, MBBS

Title: A rare case of successfully treated Enterococcal Durans infective endocarditis on a mechanical mitral valve

Authors: Mohammed Alshehri, MD - West Virginia University, Internal Medicine , Peter Farjo - West Virginia University, Internal Medicine , William Shocker, MD - West Virginia University, Internal Medicine

Introduction: Enterococcal infective endocarditis (IE) accounts for up to 20% of all cases. However, IE secondary to Enterococcus Durans is very rare with only 7 cases reported in the literature. This report describes a case of successfully treated E.durans infective endocarditis and is the first case describing the pathology on a mechanical valve in a female patient.

Case Presentation: A 56-year-old female presented from an outside facility with acute renal failure (ARF). She initially presented with a 3-month history of generalized fatigue, weight loss, and back pain. Her medical history included hypertension, diabetes, mechanical mitral valve, atrial fibrillation, coronary artery disease, and peripheral artery disease with a recent carotid endarterectomy. At the time of admission she was afebrile, normotensive with a HR of 56 bpm and oxygen saturation of 98% on room air. A chest examination revealed a 4/6 diastic murmur on the apical area along with significant tenderness on the left upper chest wall that radiated to the back. A further examination showed trace pretibial edema but no other significant physical findings. Admission blood tests revealed a Hgb of 10.0, WBC of 2.9, and platelet count of 105,000; creatinine of 5.9 and a C-reactive protein of 61.8. A CT scan of her abdomen showed splenomegaly with splenic infarction suggesting a possible septic embolic event. Blood cultures grew gram positive cocci in pairs and chains which eventually speciated as E.durans. A transesophageal echocardiogram (TEE) showed: bi-leaflet mechanical mitral valve with small mobile vegetation, 0.8 X 0.3 cm, on the medial aspect of the annulus and an ejection fraction of 55-60%. A kidney biopsy was performed which revealed proliferative infectious glomerular nephritis.

With the significant ARF the treatment with Daptomycin was empirically initiated. However, after the speciation of the blood culture and the findings of TEE the treatment with Daptomycin changed to renal dosed Vancomycin for late prosthetic IE. Antibiotics susceptibility testing showed E.durans susceptible to ampicillin, ciprofloxacin, Gentamicin, and Vancomycin. On the basis on these Results: and patient’s persistent AKF the decision made to change the antimicrobial therapy to Ampicillin and ciprofloxacin. The patient clinically improved and the creatinine level continued to trend down. Treatment was adjusted again to Daptomycin instead of ampicillin due to development of eosinophilia. Repeated Transthoracic Echocardiogram after 6 weeks did not show any vegetation.

Discussion: Prosthetic Enterococcal IE is typically treated with a cell wall active agent plus gentamicin for synergy. In this patient, owing to the renal failure, gentamicin was avoided and ciprofloxacin was combined with ampicillin based on the sensitivity Results:. Ciprofloxacin as a part of the therapeutic regimen for E.durans IE was associated with full recovery in a previous case report. This case also shows that therapeutic challenges were almost always associated with IE secondary to E.durans due to its association with an elderly population with other comorbidities.

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West Virginia-Clinical Vignette-Poster Finalist
Obadah S Aqtash, MD

Title: Extensive Cutaneous Reaction and Tongue Raynaudes with Tenofovir

Authors: Obadah Aqtash MD, Brent Thornhill MD, Ryan Carroll MD, Adee Elhamdani MS3, Eva Tackett MD.

Introduction: Drug reaction with eosinophilia and systemic symptoms (DRESS) is a potentially fatal disease. Tenofovir-induced DRESS is extremely rare and has only been previously reported once. Raynaud’s of the tongue is also an exceptionally uncommon phenomenon usually reported with connective tissue diseases. We present a case of a patient who developed Tenofovir-induced DRESS that manifested as liver failure, skin eruption, and Raynaud’s of the tongue.

Case Presentation: A 65 year-old male patient was transferred from a local hospital with the complaint of generalized skin rash of five weeks duration. He was hemodynamically stable with blood pressure 135/96, heart rate 95, and temperature 97.7. On physical exam, the patient had icteric sclera and a diffuse exfoliative maculopapular rash involving his entire body. On laboratory evaluation, complete blood count and basic metabolic panel were found to be normal with the exceptions of white blood cell count of 11.3 and sodium level of 133. Liver function testing was significant for alkaline phosphatase level of 1505, AST 376, ALT 595, bilirubin of 15.4, and direct bilirubin of 12.8. Ultrasound of the abdomen and duplex scan were both unremarkable. The patient indicated that he was diagnosed with hepatitis B two months earlier and was started on Truvada (emtricitabine /tenofovir). Records obtained from the outside hospital showed that the patient’s liver enzymes were normal 3 weeks earlier but revealed an eosinophil count of 25% with a repeat of 33%. The patient was managed with normal saline, steroids, and Benadryl. His Tenofovir was discontinued. Skin biopsy was eventually obtained and it showed spongiotic dermatitis with eosinophilia. During his hospital stay, the patient developed Raynaud’s of the tongue following ingestion of cold water. This was followed by Raynaud’s of the nose and left fifth finger. The patient remained in the hospital for close observation. Over the span of 10 days, the patient’s rash resolved and his liver enzymes normalized.

Discussion: DRESS is a rare drug-induced hypersensitivity reaction that includes skin eruption and internal organ involvement. Onset of symptoms occurs 2-6 weeks after initiation of the offending agent. The liver is most frequently affected but all organs are potentially susceptible. Various medications have been implicated in triggering DRESS with anticonvulsants being most common. However, Tenofovir-induced DRESS has been reported only once in the literature and that patient succumbed to his disease. Additionally, DRESS-induced Raynaud’s of the tongue has never been reported. Considering that the patient was started on Tenofovir two months earlier, developed liver failure and skin eruption shortly thereafter, was found to have elevated eosinophils in his blood and skin biopsy showing spongiotic dermatitis with eosinophilia, and that his symptoms completely resolved within 10 days of discontinuation of the medication, we state that our patient developed DRESS secondary to Tenofovir.
West Virginia-Clinical Vignette-Poster Finalist
Muhammad Mustafa Bhaty, MD

Title: UNEXPLAINED ASCITES: THINKING OUTSIDE THE LIVER

Authors: Mustafa Bhaty, M.D.
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Introduction: Cerebrospinal fluid (CSF) ascites after ventriculoperitoneal (VP) shunt is rare. It is generally diagnosed in the pediatric population within two years after shunt procedure. We report a rare case of an adult patient who presented with CSF ascites 18 years after VP shunt placement, the longest interval reported.

Case Presentation: A 32-year-old female presented with a several week history of shortness of breath and abdominal distention. As an infant, she had undergone VP shunt placement for hydrocephalus, and subsequently required a shunt revision at the age of 14 years. She had no prior history of shunt infection, heart failure, hepatic, or renal disease. The results of routine blood and urine tests were normal. CT of the abdomen and pelvis showed normal liver and extensive ascites. VP shuntogram did not show any narrowing or disconnection. Doppler abdominal ultrasound and echocardiography were normal. Paracentesis revealed straw-colored fluid, no malignant cells. Protein level was 2.3g/dl and serum-ascites albumin gradient (SAAG) was 2.6. Given a normal shuntogram and high SAAG (not characteristic of CSF ascites), the cause of ascites was thought to be most likely of hepatic origin. Diuretic therapy was initiated and numerous therapeutic paracentesis were required, totaling 16 liters over a period of 4 months. Liver etiology was eventually excluded after comprehensive evaluation including trans-jugular liver biopsy which was normal. Subsequently, the peritoneal catheter was removed and a ventriculo-atrial shunt was placed. Within 1 week her abdominal distention resolved. No ascites was evident on ultrasound two months later, thus confirming the diagnosis of CSF ascites.

Discussion: This case represents what appears to be the longest reported interval for the development of V-P shunt associated ascites. If cause of ascites is otherwise unclear, CSF ascites must be suspected in V-P shunt patients regardless of age, duration of shunt placement, or ascitic fluid chemistries.
West Virginia-Clinical Vignette-Poster Finalist
Monica Chowdhry, MD

Title: Cavitary Necrotizing Pneumonia in a Young Female – A Rare Presentation of an Occult Bronchial Carcinoid Tumor

Authors: Monica Chowdhry MD, Ahmad Khan MD, Brandon Rose DO, Hesham Mohamed MD

Introduction: Carcinoid tumor of the lung is a rare neuroendocrine neoplasia, accounting for less than 2% of all the lung cancers. They are mostly asymptomatic and diagnosed incidentally on chest imaging. When symptomatic, they can present with cough, wheezing, hemoptysis or with symptoms of ectopic hormone production. Because of their atypical presentation, the diagnosis is challenging and often delayed. We report a rare case of bronchial carcinoid tumor in a young female presenting as cavitary necrotizing pneumonia.

Case Presentation: A 31-year-old immunocompetent female, non-smoker presented with fever, shortness of breath, nonproductive cough and fatigue for four days. She had two episodes of right lower lobe pneumonia in the last three years, which responded well to antibiotics alone. Older chest CT scans did not show any visible lesion. On physical examination, the patient was tachypnea and tachycardia. Lung examination revealed diminished breath sounds in the right lower zone. Blood work showed significant leukocytosis of 23,400, lactic acidosis and mild hyponatremia. Chest X-Ray revealed a right lower lobe consolidation with an air-fluid level. Chest CT showed large right lower lobe cavitary necrotizing pneumonia without any obvious lesion. Bronchoscopic examination showed an obstructive endobronchial tumor in the right lower lobe bronchus. Biopsy revealed a well-differentiated typical carcinoid tumor (low grade, G1) with a mitotic index < 2 and Ki-67 index of 2-3%. She was started on antibiotics and right lower lobectomy was performed. Surgical pathology confirmed a 2.5 cm carcinoid tumor (pT2a N0) confined to the lung with no evidence of necrosis or lymphovascular invasion. Pathology also showed severe acute cavitary necrotizing pneumonia with hemorrhage and pleural empyema. She did well postoperatively and was discharged in a very good condition.

Discussion: Lung carcinoid tumors or “Karzinoid” were reported in 1907 to describe tumors pursuing a more indolent clinical course. They are rare and tend to grow slower than other types of lung cancers, usually presenting in the fifth decade. Carcinoid tumor in our patient presented as cavitary necrotizing pneumonia which is extremely rare. Diagnosis of carcinoid tumor with this rare presentation can be challenging and requires a high level of suspicion. In our patient diagnosis was made by bronchoscopy and biopsies; this highlights the importance of early bronchoscopy in a young patient with recurrent pneumonia at the same location. Timely diagnosis and treatment can prevent morbidity and mortality related to this rare condition.

References

Wisconsin-Clinical Vignette-Poster Finalist
Ryan Logue, MD

Title: Multiple cranial nerve palsies mimicking acute stroke syndrome

Authors: Logue RJ, Worku D, Biswas A

Introduction: Sarcoidosis is a non-caseating granulomatous disorder that predominantly affects the pulmonary system, however virtually any organ system can be affected. Approximately 5 percent of patients with systemic sarcoidosis tend to develop neurologic complications. Of these patients, 25 to 50 percent develop either peripheral facial nerve palsy or optic neuropathy. Few cases have been presented with isolated cranial nerve III, IV, V, or VI palsies. Presentation with multiple cranial nerve palsies is rare and often masquerades as an acute stroke syndrome.

Case Presentation: A 45-year-old woman presents experiencing rapidly progressing left facial paralysis, left gaze paralysis, vision loss, gait unsteadiness, and generalized weakness. In the emergency department, NIH stroke scale was 9. Physical exam was significant for slurred speech, left facial droop, left eyelid lagophthalmos, left corneal injection, bilateral vision loss, and gross ataxia. A CT scan of the brain was negative for acute ischemia or hemorrhage. The patient then underwent a contrast-enhanced MRI of the brain and orbits with results significant for punctate focal enhancement in the cisternal segment of the left third cranial nerve and increased T2/flair signal in the right optic nerve. Distant medical records were reviewed, and she was found to have been diagnosed with pulmonary sarcoidosis approximately 10 years ago. Since this diagnosis she has not been compliant with prednisone therapy. Subsequently, we made the diagnosis of neurosarcoidosis with optic neuritis, oculomotor palsy, facial nerve palsy, and cerebellar ataxia. She was started on high-dose prednisone and within one week her symptoms improved rapidly. Urgent ophthalmology consultation was also obtained and in addition to multiple cranial nerve palsies, she was found to have bilateral uveitis for which she was started on prednisolone drops. At the time of discharge her vision had returned.

Discussion: Patients with multiple cranial nerve palsies can present the clinician with a significant diagnostic challenge. The most common explanation for such symptoms is often acute cerebral vascular accident, malignancy, infection, or trauma. In the patient we presented, symptoms were consistent with ventral pontine syndrome (previously, Millard-Gruber syndrome) given ipsilateral gaze palsy, ipsilateral face weakness, vision loss, weakness, and ataxia. However, in patients with known pulmonary sarcoidosis, neurosarcoidosis must be strongly considered on the differential diagnosis. Most patients improve significantly with corticosteroids, however in some cases immunotherapy may be necessary. In cases of suspected optic nerve and oculomotor nerve involvement, urgent ophthalmology consultation is warranted.
Title: Dermatoses and nutritional deficiency in patient with underlying Hodgkin’s lymphoma

Authors: Xiaoxiao Qian, Siri Neelati, Ankoor Biswas, Department of Internal Medicine, Aurora Health Care, Milwaukee, WI

Introduction: Paraneoplastic dermatoses are the second most common type of paraneoplastic syndrome, and are related to underlying visceral or hematological malignancy. Nutritional deficiency can be the result of dermatoses but also can be the cause of dermatoses.

Case Presentation: The case involved a 73-year-old male presenting with generalized weakness, weight loss, diarrhea and diffuse rash for a duration of several weeks. A diagnosis of Grover’s disease was made by his dermatologist weeks ago after the initial presentation with diffuse erythematous macular rash on the trunk and extremities. This time patient’s rash was mostly located on the face and scalp with the involvement of ocular mucus. Based on the scaly nature of the rash, this patient was diagnosed with exfoliative dermatitis and the rash was considered to a separate process from Grover’s disease. CT of chest, abdomen and pelvis was obtained due to the high suspicion of underlying malignancy based on the clinical picture and the fact that Grover’s disease and exfoliative dermatitis are often paraneoplastic presentations. CT revealed significant abdominal lymphadenopathy and lymph node biopsy confirmed the diagnosis of Hodgkin’s lymphoma. At the same time, the patient was found to have an extremely low folate acid level while working up for his anemia though he had a previous normal folate level. This was thought to be mostly caused by the exfoliative dermatitis as there is excessive consumption of folate due to rapid skin turnover and increased keratinocyte activity. In the content of persistent diarrhea and dermatitis, a niacin level was ordered with a result of low normal. A second skin biopsy on the same site was obtained to evaluate the complexity of dermatitis weeks after the initial biopsy and it revealed similar dermatitis to the previous one but with additional component of nutrition deficiency. As the patient received chemotherapy and nutritional supplement, the rash improved significantly.

Discussion: This case illustrates the potential for multiple dermatological processes caused by a single underlying malignancy. Recognizing these skin manifestations are essential for early diagnosis. Moreover, checking for possible nutritional deficiency that could be related to these dermatoses would provide us with valuable information for prompt institution of comprehensive therapy.
Wisconsin-Clinical Vignette-Poster Finalist
Payal Sharma, MD

Title: “DUST-OFF” SPRAY CAUSING TOXIC MYOCARDITIS

Authors: Payal Sharma MD, Prakash Nallani MD, Ankoor Biswas MD

Introduction: Myocarditis has several etiologies including hypersensitivity reactions, infections, medications and toxins. Specifically, catecholamines, antipsychotic agents, alcohol, cocaine, and hydrocarbons have previously been described to cause myocarditis. Cardiac magnetic resonance imaging has proven to be a useful diagnostic tool in suspected cases of acute myocarditis. Here we present a case of myocardial injury diagnosed on cardiac MRI believed to be due to toxic inhalant abuse.

Case Presentation: A 38-year-old male with a medical history significant for hypertension and type 2 diabetes mellitus presented to the ER with diffuse myalgias and arthralgias, especially prominent in the lower extremities. He had been recreationally inhaling “Dust-Off”, a dust-cleaning spray containing difluoroethane for the past week. Initial labs showed severe metabolic acidosis along with acute renal failure, rhabdomyolysis, and elevated troponin. The patient was subsequently transferred to the ICU with suspected acute substance poisoning. Troponin level peaked at 78 mg/wL and echocardiogram showed mildly decreased left ventricular systolic function with global hypokinesis. Cardiac MRI demonstrated myocarditis and fibrosis as evidenced by patchy diffuse myocardial edema and abnormal extracellular volume fraction throughout the myocardium. MRI revealed delayed mesocardial enhancement involving 20% of the myocardium. Given the history of substance abuse with complications, the patient’s symptoms were deemed secondary to toxic myocarditis. The patient gradually improved with supportive care. Repeat cardiac MRI four months later showed resolution of previously seen myocardial edema and systolic dysfunction; there was improved, minimal residual patchy delayed enhancement of the basal lateral wall with portions of the anterior and inferior wall.

Discussion: It is essential to include substance-induced cardiomyopathy in the list of differential diagnoses for patients presenting with cardiac dysfunction of unknown cause. The pathophysiological mechanism is related to hydrocarbons, which can precipitate sudden death and cause myocardial sensitization to catecholamines by inhibition of calcium signaling this produces ventricular arrhythmias and myocardial dysfunction. Cardiac MR is a non-invasive tomographic imaging modality which can be utilized to assess the structure and function of the heart with high accuracy. CMR is able to demonstrate typical features of acute inflammation such as dysfunction, edema, and necrosis. Injured cardiac myocytes lose their membrane integrity and gadolinium based contrast agent (GBCA) that normally remains in the extracellular space, can freely diffuse into the cell. GBCA affects the magnetic parameter T1 relaxation time, and regional differences in contrast concentration are depicted with late gadolinium enhancement (LGE). LGE imaging has been shown to be an excellent diagnostic tool in suspected myocarditis.
Wisconsin-Clinical Vignette-Poster Finalist
Kurtis Swanson, MD

Title: Sweet’s Syndrome (Acute Febrile Neutrophilic Dermatosis)

Authors: Kurtis Swanson MD, Lara Voigt MD, Pinky Jha MD, MPH

Introduction: Sweet’s Syndrome (acute febrile neutrophilic dermatosis), cited as a “para-inflammatory” phenomenon, is a rare disease marked by painful eruption and pathergy. This condition is important for internists to recognize, diagnose, and manage, as it avidly responds to steroid therapy and can be a harbinger for malignancy.

Case Presentation: Patient presented to the ED with 7 days of progressively worsening painful upper extremity rash. 1 day prior to onset, the patient underwent left wrist carpal tunnel release. Interestingly, the rash emerged on the dorsum of her right hand at a venipuncture site used during the procedure. 1 day later, the rash had progressed to both forearms, manifesting as tender nodular lesions with surrounding erythema. She sought care at Urgent Care, where she was prescribed antibiotic therapy for presumed non-purulent cellulitis. Patient sought further care several times prior to presentation, due to worsening pain/rash despite appropriate empiric antimicrobial therapy. Intractable pain led to her presentation to the Emergency Department.

She was non-toxic appearing and in no acute distress. Patient was afebrile, tachycardic to the low 100s and otherwise hemodynamically stable and saturating well with normal respiratory rate on room air. Physical exam was remarkable for multiple annular lesions on dorsum of hands/forearms ranging from 5-6mm to 3cm with central eschar present in some along with blanching perilesional erythema. Patient was neurovascullarly intact.

Labs notable for leukocytosis to 11K with 77% PMN, ESR 50. Imaging notable for wrist plain film with evidence of soft tissue swelling without obvious osteomyelitis. She was subsequently admitted. Initial management included Linezolid + clindamycin, opiate analgesia and Dermatology consultation. Lesional punch biopsy revealed dermal neutrophilic infiltrate consistent with Sweet’s Syndrome (acute febrile neutrophilic dermatosis).

Discussion: Sweet’s syndrome (acute febrile neutrophilic dermatosis) is a rare eruption associated with infectious, inflammatory and neoplastic processes. It occurs predominantly in women aged 30-60 years. Originally discovered in 1964 by British physician Dr. Robert Douglas Sweet, classical criteria exist to delineate this condition from similar eruptions including 2 major criteria: 1) Abrupt onset of painful erythematous plaques/nodules, 2) Histopathologic evidence of a dense neutrophilic infiltrate without signs of vasculitis as well as 4 minor criteria: pyrexia, association with malignancy, infection, inflammatory process, excellent response to steroids or potassium iodide, as well as 3 of 4 abnormal labs: ESR >20, positive CRP, >8K WBCs and >70%PMN. The patient met diagnostic criteria fulfilling both major and 2/4 minor criteria (steroid response and laboratory abnormalities).

She was treated with 60mg Prednisone for 1 week with resolution of her symptoms.

Sweet’s Syndrome (acute febrile neutrophilic dermatosis) is a rare eruption that internists ought to be mindful of particularly in young women with possible cellulitis refractory to antibiotics. Key criteria exist to identify this condition, which responds well to steroid therapy.

References
Visual Diagnosis. Sweet’s Syndrome.
Resident / Fellows Research – Podium Presentations
Title: Reducing the Ordering of Inappropriate Echocardiograms in a VA Medical System

Authors: Cecil A Rambarat MD, George Dibu MD, Garret Brown MD, Anas Al-Ani MD, David E Winchester MD

Introduction: Healthcare in the United States is disproportionately expensive when compared with other developed nations. One reason for this is inappropriate ordering of diagnostic tests. Echocardiograms, which vary in cost from $400 to $4000 across the United States, are frequently ordered, some of which for inappropriate reasons. Reducing the ordering of inappropriate echocardiograms could reduce healthcare costs through decreasing need for extra echocardiogram lab personnel and reducing physician time spent reading these studies. Our goal was to reduce the ordering of inappropriate echocardiograms by 30%, in a period of 6 months, by incorporating a tested questionnaire, which guides appropriateness of imaging, into the echocardiogram order menu in CPRS.

Methods: A four-question yes/no questionnaire was developed based on a prior study [1] (echocardiogram within the past year, echocardiogram for endocarditis in the absence of new murmur or positive blood cultures, echocardiogram for surveillance of a known problem, echocardiogram in the absence of new symptoms/change in clinical exam) and added to the CPRS echocardiogram ordering menu. Following the four yes/no questions we added a statement that if the provider answered yes to 2/4 questions, there was an 80% chance that the echo order was inappropriate. The ordering provider was left at liberty to proceed with the echocardiogram order if he/she decided to do so at that point. A total of 294 CPRS charts were reviewed prior to the implementation of the questionnaire for incidence of inappropriate echo orders. Appropriateness was determined based on the American College of Cardiology Foundation Appropriate Use Criteria Task Force guidelines [2]. A total of 293 CPRS echocardiogram orders were reviewed 6 months after the implementation of the questionnaire for appropriateness.

Results: Prior to our intervention, we found 16.3% (48/294) echocardiograms ordered for inappropriate reasons. After instituting our questionnaire into the echocardiogram ordering menu we found a decrease in the incidence of ordering of inappropriate echocardiograms to 11.6% (34/293). The absolute reduction in inappropriate echocardiograms was 4.7% and the relative reduction was 29% (p=0.12, 95% CI 0.46-1.09).

Conclusion: Institution of a questionnaire into the CPRS echocardiogram ordering menu may reduce the ordering of inappropriate echocardiograms. Refinement of the questionnaire may be necessary to produce more reliable Results.

References

Illinois-Research-Podium Presentation
Keith Smart, DO

Title: Post-Pulmonary Embolism Syndrome Development in Patients Evaluated by a Pulmonary Embolism Response Team

Authors: Keith Smart1 DO, Iva Golemi1 MD, Oscar Garza2 MD, Omer Iftikhar2 MD, Alfonso Tafur3 MD,
1Department of Internal Medicine, NorthShore University HealthSystems, 2Department of Cardiology, NorthShore University HealthSystems, 3Department of Vascular Medicine, NorthShore University HealthSystems

Introduction: Approximately 100,000 Americans die each year from venous thromboembolism (VTE). The rapidly evolving management of complex cases of pulmonary embolism (PE) has sponsored the creation of Pulmonary Embolism Response Teams (PERT) as an infrastructure to streamline management of patients with PE. Post PE syndrome is a clinically significant spectrum that is characterized by persistent right ventricle (RV) dysfunction and diminished functional status in survivors of PE. We describe the implementation, design and outcomes of the PERT protocol at NorthShore University HealthSystems and aim to create a prospective registry to assess Post PE syndrome development.

Methods: The PERT protocol was initiated after diagnosis of PE with high-risk features including RV dilatation on CT scans concerning for sub-massive PE. We developed a PERT registry that includes initial activation and outpatient follow up data. A follow up ECHO was ordered on patients with persistent symptoms after 3 months. Patients were classified as post-PE syndrome if the imaging right ventricular systolic pressure (RVSP) increased along with clinical evidence of decreased functional capacity not attributed to any other disease pathology. Continuous variables are reported as median and categorical as percentages.

Results: A total of 58 patients required activation of PERT, median age of 70.5, most (65.5%) were female, mean PESI class was III and 32.7% had cancer (Table 1). The RV was dilated by CT criteria in 48.9% of the patients and 40% had troponin elevation on presentation. PERT decided to do thrombolysis in 10%. A reported 8.5% of the patients died. Among survivors, 11% had an abnormal right ventricular systolic pressure on ECHO. Right heart ECHO variables are tabulated in Table 2. Positive troponin, increased RV/LV ratios and increased RVSP at diagnosis were not associated with post-PE syndrome incidence. Patients with PESI class V were more likely to have an abnormal ECHO at follow-up, but this result was not statistically significant (OR 2.1 95%CI 0.3-13.4).

Table 1: Basic demographics and clinical parameters from NorthShore's PERT registry. (N=58)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Count</th>
<th>Percentage</th>
</tr>
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<tbody>
<tr>
<td>Median Age</td>
<td>70.5</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>38/58 (65.5%)</td>
<td></td>
</tr>
<tr>
<td>Active Malignancy</td>
<td>19/58 (32.7%)</td>
<td></td>
</tr>
<tr>
<td>Concomitant DVT</td>
<td>37/58 (66.6%)</td>
<td></td>
</tr>
<tr>
<td>RV/LV ratio &gt;1</td>
<td>23/58 (48.9%)</td>
<td></td>
</tr>
<tr>
<td>Troponin Elevation</td>
<td>22/58 (40%)</td>
<td></td>
</tr>
<tr>
<td>Pro-BNP / BNP Elevation</td>
<td>28/58 (68.2%)</td>
<td></td>
</tr>
</tbody>
</table>
**Table 2: ECHO Variables (N=10)**

<table>
<thead>
<tr>
<th></th>
<th>Median RVSP at Diagnosis</th>
<th>RV Dilation at Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis</td>
<td>30 mm Hg</td>
<td>5/10 (50%)</td>
</tr>
<tr>
<td>Follow Up</td>
<td>40.5 mm Hg</td>
<td>5/10 (50%)</td>
</tr>
</tbody>
</table>

**Conclusion:** Our PERT algorithm does successfully select high-risk PE patients with a high incidence of post-PE syndrome. Given our preliminary Results: and other literature reports approximating the development of post PE syndrome in 20-30% of patients, we estimate that a 500 patient comparative trial may define if there is clinical utility of PERT initiatives, and is needed as to justify its growing acceptance.

**References**

Title: A Successful Congestive Heart Failure Management Program: Quality Improvement Initiative by Internal Medicine Residents

Authors: Naga Sai Shravan Turaga MD, Shashitha Gavini DO, Himani Manthena MD, Samiya Yasin MD, Jay Patel MD, Gregory Jacob MD, Imran Shuja MD, Ashwin Narayana MD, Syed Saad MD, Avaneesh Jakkoju MD, Elizabeth Borrero MD

Introduction: Congestive heart failure (CHF) is a major healthcare issue all over the world with an ever-increasing morbidity and economic burden. It is associated with a high 30-day readmission rate. However, detailed description of the causes and preventability of readmissions are lacking. We performed a quality improvement study to understand the high impact factors causing CHF readmissions to formulate effective strategies to reduce readmissions.

Methods: A multidisciplinary team consisting of internal medicine residents, internal medicine staff, cardiology staff, case managers and nurses was formed, and the study was done in four phases. In the first phase retrospective chart review of patients admitted for CHF exacerbation between July 2016 and November 2016 was done. Patients readmitted within 30 days were identified and the potential causes of readmissions were analyzed. During the second phase, interventions to address the high impact factors identified in first phase were formulated and the most feasible ones were selected with common consensus of the team members. Third phase was the implementation phase in which the interventions were implemented. Fourth phase involved analyzing the logistics and problems associated with project implementation as well as studying the impact of the interventions. Plan-Do-Check-Act (PDCA) methodology was used as a quality improvement tool for each phase to help identify factors and processes in our institution that required change.

Results: Retrospective chart review in first phase revealed 1 month readmission rate of 22% and 1 week readmission rate of 11%. The major patient related factors impacting readmissions were medication unaffordability (75.5%), medication noncompliance (28.5%), and transport (10%). While the major health care system related factors were lack of 1 week post discharge follow up (100%), medication reconciliation and patient education. Interventions selected for implementation were peer-to-peer education of the multidisciplinary team about their role, selected interventions, CHF management guidelines, home health evaluation and medication reconciliation, establishing 1 week post discharge clinic follow up, post discharge nurse follow up phone call in 3 days, $40 financial assistance for patients at discharge and providing a revised educational material to the patients. Other interventions which were identified but not selected due to non-feasibility were transportation assistance, substance abuse management and forming patient support groups for education and counselling. The interventions were implemented from August 2017. Four months after the interventions were implemented, 1 month readmission was 9.7% (11 readmissions out of 113 admissions) and 1 week readmission rate was 1.7% (2 readmissions out of 113 admissions).

Conclusion: Every medical facility has its own unique patient population and hurdles in providing health care. An individualized approach to identify and address those hurdles can direct towards successful strategies to reduce CHF readmissions and thereby improve quality of life of the patients and reduce the economic burden on the society.

References

Maryland-Research-Podium Presentation
LT Sara Robinson, MC USN

Title: High Value Cost-Conscious Care: Multiple Interventions for Sustained Reduction of Unnecessary Diagnostic Tests

Authors: Sara Robinson, MD (Associate); Nora Maddy, MD (Associate); Alison Lane, MD; Rebecca Wetzel, DO (Associate); Brett Sadowski, MD

Introduction: Inappropriate lab testing contributes to the growing problem of health care costs in the Unites States. Teaching hospitals are particularly at risk for contributing to “wasted care” that is of little clinical yield in the interest of academic development. Cost-conscious care is a key topic in medical education, including specific instructions to avoid repetitive lab testing in patients with clinical and lab stability. We designed a three-cycle high-yield, systems-based intervention to reduce the number of diagnostic laboratory tests ordered for patients admitted to the internal medicine wards at a single academic center.

Methods: We isolated the computer-based, automated admission order sets as the target for a sustainable intervention. Prior to our interventions, the order set allowed for basic labs such as complete blood count, basic and comprehensive metabolic panels, coagulation and mineral panels to be easily ordered daily with a ‘qAM’ selection as the default for frequency in the admission order set. In the first cycle of our systems-based intervention, we eliminated the daily frequency as the default option within the order set. The second cycle of our intervention changed the lab order display to include the cost of the laboratory test in the name of the order to make cost more transparent to ordering providers. The third cycle eliminated the option of ordering morning labs indefinitely with the frequency of ‘qAM’ and replaced it with ‘qAM x 3’, to stop the order after three days.

Results: Data for each cycle was collected over the two month period after each intervention was initiated. The number of labs per inpatient day was calculated and compared to a similar two month period from the previous year. After the first cycle’s intervention of removing ‘qAM’ from the default order set, the absolute number of labs was reduced by 19% when controlled for number of inpatient days. After the second cycle’s intervention of displaying cost, the absolute number of labs was not significantly reduced further, but the initial reduction was sustained. After the third cycle of replacing ‘qAM’ with ‘qAM x 3’ for the frequency option, an absolute reduction in the number of labs of 22% was observed. Over the three cycles there was an overall reduction of 35% in all laboratory orders when controlled for inpatient bed days.

Conclusion: These interventions were highly effective at limiting unnecessary lab testing and reducing cost of lab tests among inpatients on the internal medicine wards. We demonstrate that multiple step-wise interventions can have an additive effect to refine interventions using the Plan-Do-Study-Act cycle model. We found that the most effective method of decreasing unnecessary laboratory testing was by reducing automaticity and encourage active clinical decision making, which led to improved delivery of cost-conscious care in a teaching hospital.
Introduction: Although the benefits of stress ulcer prophylaxis (SUP) have been demonstrated in select ICU patients, it is often provided to non-critically ill patients despite the absence of appropriate indications as described in the American-Society for Health-System Pharmacists (ASHP) guidelines for SUP in 1999. In addition to the health risks associated with overuse of PPIs like increase in the risk of pneumonia and clostridium difficile colitis, another implication is an increase in healthcare expenditure with minimal benefit.

Studies showed that factors associated with misuse of PPI included physicians' poor knowledge of indications for appropriate use and lack of awareness of potential side effects of therapy. Not enough studies have been done in an attempt to evaluate successful interventions that improve prescribing behaviors among physicians.

Our project aims to evaluate the use and the indications of PPI as GI prophylaxis at Crittenton Hospital Medical Center, identify the gap in inappropriate use of PPI, analyze the root causes, design, implement, and evaluate an intervention to close this gap.

Methods: The IHI model was used to guide and format this project.

Plan: A retrospective chart review of 100 patients whom PPI was prescribed during the hospital admission was performed for the month of October in 2012 to identify the indication for initiation of PPI in each patient. A significant gap was identified that necessitates an intervention.

Do: Using root-cause analysis, we identified a system-based problem where the patient can be prescribed a PPI without an indication; a new order tab was developed and implemented in the EMR for use of PPI in the non-ICU setting that requires the prescriber to specify their indication for use of a PPI as per ASHP guidelines for SUP.

Study: Post implementation of the new tab, chart review of another 100 patients over a one month period was done to assess any change.

Act: Full implementation of the tab order was adapted and used as a model for future interventions.

Results: of the retrospective chart review during October, 2012 showed that 21% of inpatient PPI prescriptions had no appropriate indication. Results: of the chart review during October, 2016, post implementation of the new tab, showed that only 5% of the patients were prescribed a PPI without an indication of use, which is a significant decrease with p-value of 0.0026 (p-value <0.05). There was a relative risk reduction of 76% in the inappropriate use of PPI.

Conclusion: Overuse of PPIs has been associated with health risks and increase in healthcare expenditure. It represents one example of over utilization and treatment the health system is suffering from. Utilizing of informational technology as an intervention, not only to educate the prescriber, but to
establish a valve to reduce or even eliminate inappropriate utilization of resources was clearly successful in this QI project.

References

8. Thomas L, Culley E, Gladowski P, Goff V, Fong J, Marche S. Longitudinal Analysis of the Costs Associated with Inpatient Initiation and Subsequent Outpatient Continuation of Proton Pump Inhibitor Therapy for Stress Ulcer Prophylaxis in a Large Managed Care Organization. JMCP. 2010;16(2):122-129
Anoushiravan Hakim, MD

Title: Opioid-induced Constipation: An Educational Intervention to Improve an Overlooked Cause of Patient Discomfort!

Authors: A. Hakim, MD (Associate), M. Tibuakuu, MD, MPH (Associate), J. Xi, MD, MPH (Associate), N. Ali, MD (Associate), D. Murphy, MD (Associate), S. Sattar, MD (Associate), L. Kalvala, MD (associate); M. Tamal, MD (Associate); S. Sheikh, MD (Associate), S. Nadella, MD (Associate), C. Yang, MD, PhD (Associate), and H. Kawsar, MD, PhD (Fellow), Department of Internal Medicine, St. Luke’s Hospital, Chesterfield, MO

Introduction: Opioids are the most commonly used analgesics for pain management in hospitalized patients. Approximately 40% to 81% of patients on opioid analgesics experience opioid-induced constipation (OIC) which is associated with significant patient discomfort. Prophylactic scheduled laxative therapy is recommended to prevent OIC. The purpose of this study was to improve appropriate use of laxatives to prevent OIC.

Methods: We conducted a retrospective study on patients admitted to the medical floor who received any type of opioid analgesics for at least two days, from October 2016 to August 2017. Patients on surgical, CVICU, Ob/Gyn, and emergency department were excluded. After completion of pre-intervention data collection, a 5-week educational intervention was conducted for the physicians taking care of patients in teaching service. Post-intervention data was collected and analyzed using appropriate statistical tools.

Results: In all, 146 and 198 qualified patients were included, before and after the intervention, respectively. Patients characteristics were similar in both pre- and post-intervention period. The most common reason for narcotic analgesic use was pain due to musculoskeletal etiology. Although marginally significant, educational intervention resulted in 10% improvement in the overall (scheduled or PRN) use of laxatives (p=0.05). However, no significant difference was observed in the proportion of patients who received scheduled laxatives between pre- and post-intervention groups. Among patients who received scheduled laxatives, there was about 25% improvement in administration of the recommended laxative (Senna or Bisacodyl) after the intervention (58.1% vs 82.7%) (p<0.001). There was a significant improvement in the administration of the recommended laxative among patients admitted to the teaching service (intervention group), compared to non-teaching service (81.4% vs 18.6%) (p<0.001).

Conclusion: An appropriately designed physician education program is effective in improving proper prophylactic use of laxative for patients taking opioid analgesics to prevent OIC. Ongoing educational program is required to sustain the improvement.

References

New York-Research-Podium Presentation
Arun Kumar

Title: Demographic trends and seasonal variations in Eosinophilic Esophagitis (EoE) hospitalizations: Analysis of Nationwide Inpatient Sample from 2008 to 2012.

Authors: Arun Kumar M.D, Shantanu Solanki M.D, Khwaja Haq M.D, Akshay Khatri M.D, Amirta Devi M.B.B.S, Christopher Nabors M.D;

Department of Medicine, New York medical college at Westchester Medical center, Valhalla, N.Y.

Introduction: Eosinophilic Esophagitis (EoE) is a newly recognized autoimmune condition, which presents as dysphagia and food bolus obstruction events in adults. It is considered a form of food allergy and many patients respond favorably to dietary elimination therapy. Currently, there are limited and sometimes conflicting data regarding whether changes in dietary patterns or other factors such as seasonal factors influence rates of diagnosis and hospital admissions for the condition. We reviewed the NIS database from 2008 to 2012 to assess the demographic trends and seasonal variations in EoE of inpatient hospitalizations.

Methods: We used the nationwide inpatient sample (NIS) database 2008-2012 to identify patients with EoE using International Classification of Diseases, Ninth Edition, Clinical Modification procedure codes. Regional differences in EoE incidence, survival to hospital discharge, and resource utilization (total hospital cost and discharge disposition) were analyzed.

Results: We identified 15,275 adult hospitalizations for EoE from 2008 to 2012. These increased from 566 in 2008 to 4620 in 2012. We found significant seasonal variation in EoE hospitalizations, with fall admissions increasing from 1134 (September) to 1451 (October) (p<0.0001). October to December admission rates (5241/14596) were significantly higher than during the remainder of the year (9355/14596) (p<.0001). Most patients were less than 65 years old, more commonly male (60.2%) than (39.5%) female, with no change in gender-related trends over time. More patients were Caucasian (72.1%) than African American (8.1%), Hispanic (4.8%) or other race (3.7%). Admissions varied according to regions: South (30.22%), West (26.37%) and Midwest (26.08%) and Northeast (17.33%). Most urban hospitalizations were in teaching (66.4%) rather than non-teaching hospitals (28.4%), with 4.5% of cases in rural hospitals. Weekday admissions were more common (78.8%) than weekends (21.2%). Mean length of stay increased from 3.6 days in 2008 to 4.7 days in 2012; overall cost per hospitalization increased from $8,548 in 2008 to $11,119 in 2012. Despite an increase in hospitalizations, all-cause mortality declined from 0.83% in 2008 to 0.22% in 2012. We didn’t find any statistically significant seasonal variation in regards to age, gender, race or region.

Conclusion: In this study, hospital admissions for EoE increased over time, were more common in urban teaching centers, most commonly involved Caucasian males and were more frequent in the fall/winter season. Further study will be needed to explore those factors which contribute to these trends.
New York-Research-Podium Presentation
Avinash Singh, MBBS MD

Title: Reducing Inappropriate Proton-Pump Inhibitor (PPI) Therapy in Patients Seen by Internal Medicine Residents in Primary Care Clinics

Authors: Avinash Singh\textsuperscript{1}, Heather Viola\textsuperscript{1}, Elijah Verheyen\textsuperscript{1}, Lorenz Leuprecht\textsuperscript{1}, Luba Uslar\textsuperscript{1}, David Weininger\textsuperscript{1}, Dipal Patel\textsuperscript{2}, Tamara Goldberg\textsuperscript{2}, Ranjan Ginde\textsuperscript{2}, \textsuperscript{1}Internal Medicine, Mount Sinai St. Luke's & West Hospitals, New York, NY, United States, \textsuperscript{2}Internal Medicine Residency Program, Mount Sinai St. Luke's & West Hospitals, New York, NY, United States

Introduction: Proton-pump inhibitor (PPI) prescribing has been increasing, with long term PPI therapy recommended only in certain medical conditions. Prior studies suggest that 50\% of PPI prescriptions are inappropriate. Recent findings on the risks of prolonged therapy, including kidney disease and infection, have led to several initiatives to discontinue PPI therapy when not indicated. The goal of this Quality Improvement (QI) project was to identify inappropriate PPI use and to deprescribe therapy where not indicated, based on symptoms and established guidelines.

Methods: This was a cohort-based QI initiative across three federally-qualified urban community health clinics, during a 7-week intervention phase. 2,687 patients were seen in total by Internal Medicine resident providers of which 429 (16\%) were previously prescribed and currently taking a PPI. Patient encounters that were spent addressing other active medical issues were excluded, as well as encounters that were not reported by the physician. Medical reconciliation at each visit would identify patients on a PPI. A deprescribing algorithm was created for providers to assist decision making, and information regarding indications and risks of the medication was provided to all patients. Indication for therapy, ideal prescribing action, and action taken were recorded for all patients included in the study. Data was pooled from all three clinic sites to analyze the percent reduction of PPI prescriptions that qualified as inappropriate.

Results: Sixty-nine patients were included in our study. The average age of the intervention population was 59.7 years (SD 10.9). The most common indication for a PPI prescription was Gastroesophageal Reflux Disease (GERD) (47 patients, 68\%) followed by Peptic Ulcer Disease (PUD) (4 patients, 6\%). 56 patients (81\%) were identified as inappropriate users per the prescribing guidelines. Prescriptions of 34 (49\%) patients were discontinued, 11 (16\%) were initiated on a taper with the aim of discontinuing the medication over the following weeks, and 13 (19\%) patients met indications to continue PPI therapy, and were encouraged to do so. PPI-therapy was deprescribed in 65\% of all patients, representing 80\% of those who were identified as inappropriate users.

Conclusion: This QI initiative reveals that a vast majority of our patients are on inappropriate PPI therapy, with GERD being the most common indication for the initial PPI prescription. Our intervention has helped reduce PPI use in a significant proportion of inappropriate users. The strategy adopted can serve as a valid approach to reduce unnecessary PPI prescriptions. Further studies are needed to assess symptom recurrence, reduction in long term PPI-related complications, and the efficacy of our method compared to other models in patients in whom PPI therapy is being discontinued.
Pennsylvania-Research-Podium Presentation
Shivtaj Mann, DO

Title: Resveratrol Increases Cathelicidin Expression and Offers a Novel Approach to Combat Staph aureus Invasion

Authors: Shivtaj Mann, DO, Kyungho Park, PhD

Introduction: Cathelicidin (CAMP), an antimicrobial peptide, is commonly described in literature as a byproduct of the Vit D receptor pathway and is a key component of the epidermal barrier. The epidermal barrier is a refined product of billions of years of evolution that serves to protect vital tissues from microbial pathogens and mechanical damage.

We previously described a novel, VitD receptor independent pathway that showcased the role of sphingosine-1-phosphate (S1P) in de novo production of cathelicidin. Moreover, we have also shown that keratinocytes prevent apoptosis by de novo production of sphingomyelin from ceramide. Here, we investigate if ceramide expression can be induced by resveratrol (RESV), a polyphenol known to increase ceramide expression, and if induced ceramide can modulate downstream cathelicidin production in order to enhance the innate skin barrier and combat staph aureus invasion.

Methods: We used the well established oxazolone (Ox) model to perturb the epidermal skin barrier and study the effects of cathelicidin production in restoring the skin barrier and combating staph aureus penetration/invasion. Six mice model groups were created using the hairless SKH1 mice, including two control groups: EtOH and Sphingosine-kinase-inhibitor (SKI), and four experimental groups: Ox+EtOH, Ox+SKI, Ox+RESV, Ox+RESV+SKI. RESV was applied to the flanks of the mice 2x/day for 21 days. Skin samples were obtained at the end of the 21 day period. qT-PCR and Western blot techniques were used on homogenized skin samples in order to quantify cathelicidin expression. Further, NF-κB receptor assay was utilized to assess whether RESV increased activation of the NF-κB nuclear transcription factor. Finally, skin tissue biopsies were gram stained to investigate the effects of cathelicidin in preventing invasion of staph aureus into and through the innate skin barrier. Statistical comparisons were performed using an unpaired Student t-Test.

Results: Topical RESV increased CAMP production in murine skin compared to control, p<.01. Moreover, we show that this increased production is secondary to activation of NF-κB pathway as revealed by 1) increased phosphorylation of NF-κB1(marker of activation) in RESV group, 2) attenuation of phospho-NF-κB by introduction of SKI, 3) attenuation of CAMP production with introduction of BAY11-7082, a NF-κB inhibitor, in cultured keratinocytes; p<.01. Lastly, gram stain of skin biopsies of control vs experimental groups were compared. Gram staining revealed that RESV prevented invasion of staph aureus into the epidermal barrier.

Conclusion: Our Results: display strong evidence that 1) RESV increases CAMP production, 2) this upregulation likely follows the activation cascade ceramide→sphingosine→S1P metabolism mediated by NF-κB nuclear transcription factor, and most importantly 3) upregulation of CAMP expression may offer a novel method to combat staph aureus invasion in an era of ever-increasing antimicrobial resistance.
Title: Application of a 21-gene Recurrence Score Assay in Node Negative, Hormone Receptor Positive Breast Cancer Patients in the Military Population

Authors: Wan, WingYee. Dela Cruz, Wilfred. Aden, James. Terrazzino, Sandra. Villarreal, Sarah., Department of Internal Medicine, San Antonio Military Medical Center, Fort Sam Houston, TX

Introduction: Genomic characterization of breast cancer tumors offers useful prognostic information. The Oncotype DX Recurrence Score is a reverse-transcriptase-polymerase chain reaction assay of 21 genes, the Results: of which are used to calculate a score that describes the risk of breast cancer recurrence. Low scores indicate low recurrence, for which the use of hormone therapy alone is adequate, while higher scores may require adjuvant chemotherapy. Available since 2004, the use of the Recurrence Score has become incorporated into standard of care of breast cancer management. This study aimed to analyze the use of the Recurrence Score since its adoption at our institution to characterize this unique population subset, to describe this institution’s breast cancer tumors clinicopathologically, to analyze the use of the Recurrence Score to establish treatment, and to evaluate the effect of this genetic assay on patient outcome.

Methods: This was a retrospective cohort study. Our Oncotype DX database, which spanned from 2007-2017, contained scores for 149 patients, of which 148 were included in this study based on the presence of hormone-positive, node-negative breast cancer. Data regarding patient and tumor characteristics, resultant adjuvant therapy, and outcomes (both rate of recurrence and mortality) were collected.

Results: Out of 148 patients, 47% were low risk (score <18), 41% were intermediate risk (score 18-30), and 12% were high risk (>30), consistent with prior validation studies. There was no correlation between patient age or ethnicity and score severity, while higher scores were associated with larger tumor size (p= 0.025), higher histologic grades (p<0.001), and rates of recurrence (p = 0.044). Interestingly, patients who had tumors with receptor status other than estrogen receptor positive (ER+), progesterone receptor positive (PR+), human epidermal growth factor receptor 2 negative (HER2-) were all in the high risk category (n=8). Only 7% of low risk patients were offered chemotherapy, compared to 52% of intermediate risk patients and 100% of high risk patients. Overall 5 year progression-free survival was 94.5%: low risk patients had 98.6% survival, intermediate risk patients had 93.3% survival (p = 0.114 compared to that of low risk), and high risk patients had 88.2% survival (p = 0.034 compared to that of low risk).

Conclusion: This characterization of practice within our military health system showed that the Recurrence Score has guided use of chemotherapy in patients with node-negative, hormone positive breast cancer, with 7% of low risk patients being offered chemotherapy, 52% of intermediate risk patients, and 100% of high risk patients. The rate of chemotherapy offered to intermediate risk patients is slightly above that of published literature.
Resident / Fellows Research – Poster Finalists
Alabama-Research-Poster Finalist
Joanna Zurko, MD

Title: Impact of Infectious Disease Consultation on Guideline Adherence and Mortality in Patients with Candidemia

Authors: Joanna Zurko MD, Rachael Lee MD

Introduction: Candida species are extremely virulent pathogens in blood stream infections with crude mortality rates as high as 61%1. It is well established that infectious disease consultation (IDC) is associated with reduced mortality and improved guideline adherence in patients with Staphylococcus aureus bacteremia2,3. The aim of this study was to determine how IDC impacts guideline adherence and mortality in patients with candidemia.

Methods: Using a retrospective cohort design, we included patients with a diagnosis of candidemia seen at our institution from January 2015 to July 2016. Patients who died within two days of positive blood cultures were excluded. Differences between groups were analyzed using t-test and chi square testing. Logistic regression was performed to assess for predisposing factors for IDC as the outcome variable. A p-value of <0.05 was considered significant.

Results: 163 patients met the case definition. 13 patients were excluded due to death within two days. Of the remaining patients, 111 (74%) received IDC and 39 (26%) did not. Baseline demographics and comorbidities were similar between groups. Patients who received IDC were more likely to receive: definitive antifungal therapy (100.0% vs 92.3%, p=0.003), repeat cultures documenting clearance (92.8% vs 46.2%, p<0.001), dilated retinal examination (64.9% vs 28.2%, p=0.001), echocardiogram (74.8% vs 53.8%, p=0.015), appropriate antifungal duration (87.9% vs 71.4%, p=0.02), and removal of central venous catheter (90.3% vs 58.3%, p<0.0001). 30-day mortality was significantly lower in the IDC group (18.9% vs 51.3%, p=0.0001). APACHE II scores were significantly lower in the IDC group (21.6 vs 27.2, p=0.01); therefore, patients were further divided into two groups: those with low-intermediate (<24) and high (≥24) APACHE II scores. In those with high scores, IDC was associated with a significant 30-day mortality benefit compared to no IDC (37.5% vs 75.0%, p=0.004) with similar average scores (34.48 vs 34.08, p=0.830). In those with low-intermediate scores, IDC was not associated with a significant mortality benefit compared to no IDC (8.45% vs 13.33%, p=0.55) with similar average scores (16.3 vs 16.2, p=0.28). There was no significant difference in hospital readmission, time to definitive therapy, time to clearance of cultures, duration of hospital stay, or recurrent candidemia between groups. On logistic regression analysis, a higher APACHE II score was associated with a decreased likelihood of obtaining IDC (OR 0.96, p=0.01).

Conclusion: In this study, IDC was associated with lower 30-day mortality and improved guideline adherence in patients with candidemia. This mortality benefit was seen primarily seen in those with greater severity of illness; however, those with higher APACHE II scores were also less likely to receive IDC. Therefore, those who may benefit most from IDC are often not receiving these services. These Results: suggest that IDC may lead to better outcomes in candidemia, but further investigation is needed.

References

Arkansas-Research-Poster Finalist
Joseph H Holthoff, MD PhD

**Title:** Zoster meningoencephalitis: two cases of an uncommon diagnosis

**Authors:** Holthoff J.H., Enuganti S., Burger B., Johnson T. Hopkins R.H.

**Introduction:** Varicella zoster virus (VZV) is the most common infectious cause of both central and peripheral nervous system disorders. Primary VZV infection spreads throughout the body via the bloodstream, causing chicken pox in children, after which the virus remains latent in ganglia of the central nervous system (CNS). With increasing age there is a resultant decline in cell-mediated immunity, allowing the virus to directly invade the neuronal and glial cell bodies. The immune response to this invasion produces the characteristic vesicular skin lesions and neurological damage classically seen with Zoster reactivation. Other manifestation of zoster reactivation can include meningoencephalitis, cerebellitis, and ocular disease.

**Methods:**

**Patient #1:** 73-year-old male with a history of large B-cell lymphoma with CNS involvement and Parkinson disease. He underwent autologous bone marrow transplantation (BMT) in 2012. He received Zostavax® approximately 2 years prior to BMT and 6 years prior to presentation. He presented with headache, fever, confusion, and worsening of his baseline tremor.

**Patient #2:** 72-year-old male with history of inflammatory bowel disease (not on active treatment), hypertension, and benign prostatic hypertrophy who presented to the outpatient clinic with fever, headache, and general malaise for 2 days. He denied the presence of rash and physical examination was unremarkable. He was discharged from the clinic with recommendation of symptomatic care for a likely viral illness. Ten days later, he suffered a generalized tonic-clinic seizure while driving and was admitted to an outside hospital.

**Results:**

**Patient #1:** Basic laboratory analysis including a complete blood count (CBC), basic metabolic panel (BMP), hepatic panel, urinalysis (U/A) with culture, and blood cultures revealed no significant abnormalities. CT head and MRI brain without contrast were both negative for acute intracranial process. Cerebrospinal fluid (CSF) analysis revealed leukocytosis (303 WBC), which was predominantly lymphocytic (74%). CSF glucose (60 mg/dl) was normal and protein levels were mildly elevated (110 mg/dl). Some atypical leukocytes were noted in CSF, but further analysis with flow cytometry of CSF was negative for recurrence of malignancy. Polymerase chain reaction (PCR) for viral pathogens detection was performed and detected 477,000 copies/ml of VZV.

**Patient #2:** Laboratory analysis showed a normal CBC, hepatic panel, and U/A, but was remarkable for lactic acidosis (9.5 mMol/l) and hyponatremia (126 mMol/l). Blood and urine cultures revealed no growth. CT head and MRI brain were negative for acute pathology. A lumbar puncture was performed and CSF analysis showed predominantly lymphocytic (79%) leukocytosis (328 WBC), elevated protein (224 mg/dl), and normal glucose (44 mg/dl).
**Conclusion:** This presentation emphasizes the need for heightened clinical suspicion for VZV encephalitis in older adults, and emphasizes the waning effectiveness of the live-attenuated zoster vaccination over time.
Title: ADVERSE PATIENT EVENT DUE TO FORMAT OF COMPUTERIZED ORDER SET: A PATIENT SAFETY CASE REPORT

Authors: Madhav Chopra, Indrajit Nandi, Edward Maharam, Rustan Sharer, Jordan Coulston, Rhonda Roberts, Rita Tassinari, Samantha Woosley

Introduction: Medical error is the third leading cause of death in the United States. Incorrect medication administration is one of the most common errors that occur in the inpatient setting. Studies have indicated that almost twenty-five percent of all hospitalizations have at least one medication administration error. As electronic medical records continue to evolve, it is increasingly important to minimize any chance of error. We present the case of an Electronic Patient Event Report (EPER) filed on a patient who received a continuous infusion of hydromorphone instead of the intended single bolus of hydromorphone followed PCA boluses at fixed lock out interval.

Methods: Our patient safety consult team, performed a causal analysis was performed using a fishbone, gemba walk and process map. It was determined that a contributing source of error and target for intervention was the formatting of the computerized order and order printout. It displayed the order instructions on the same line, which led to misreading of the intended PCA dose as a continuous infusion rate and resulting in inappropriate medication administration. An inter-professional team surveyed a variety of stakeholders including physician, pharmacists, and nurse. Using the information from the survey the order set was then revised so that each component of the order was displayed on a separate line.

Results: Our intervention resulted in a process improvement measure that resulted in simplifying the order set to for all parties involved in medication administration. The multidisciplinary approach resulted in a permanent change with buy-in from all the stakeholders involved in the process.

Conclusion: From this process change we can infer that outcome measure of decreasing medication administration errors has also occurred. Due to the reliance of self-reporting nature of the EPER system we were unable to accurately quantify how often there were errors in the administration of PCAs. However, given the severity of this event and potential for catastrophe the prevalence of this error is less important. This case highlights the benefit of a formalized process reporting errors and the advantages of using an interdisciplinary team to perform causal analysis and perform system level interventions.

References


Title: Snakebites in an Ecuadorian Hospital in the Amazon Rainforest: Epidemiological profile

Authors: Francisco E. Mora, MD; Isabel Freire, MD

Introduction: Between 1.2 and 5.5 million people are bitten by venous snakes annually worldwide. It is a public health problem, affecting the population in rural areas, with difficult access to health care. In rural Ecuador, hospitalist or general practitioners manage snakebites. In Ecuador, the real impact of this problem is not well known due to limited data. Some government statistics report around 1200-1600 cases annually, with mortality estimated at 1%; although, there are independent studies that showed mortality as high as 5.4%. The aim of this report is to show the epidemiological profile of snakebites in an Ecuadorian rural hospital.

Methods: This is a retrospective, descriptive study, carried out in a remote 50-bed community hospital in the eastern rainforest region of Ecuador. All venous snakebites admissions between January 2012 and December 2014 were recorded. Epidemiological data and cost were analyzed. Adherence to management protocol and outcomes were also evaluated and are reported in a separated document.

Results: During a 36-month period, 211 cases were admitted to the hospital. The average incidence during this period was 190 per 100,000 inhabitants. 126 patients were male and 85 female. Mostly of patients were farmers of indigenous origin. 64% were between 15-65 years old. 97% of patients were bitten on the extremities. 58 (27.4%) cases were classified as without poisoning. Envenomation cases were classified as 65 mild, 80 moderate; and 8 severe. The type of snake was identified in 66% of the cases, Bothrops atrox accounted for 84 cases (40%). The average hospitalization days for mild cases were 4 days, moderate cases were 5 days, and severe cases were 7 days. Time from bite to reaching the hospital averages 6 hours. Nine patients (6%) presented mild allergic reaction to antivenom. The cost of each antivenom bottle, depending of the market and availability, range from 42 to over a 100 US dollars. Each patient received between 4-12 bottles in average, depending of the envenomation severity.

Conclusion: The incidence of snakebites in southeast Ecuador is very high and affects mainly to the population involved in agriculture and livestock. Extremities are the most affected body part. There has been identified 200 species of snakes in Ecuador, with 44 of this been venomous. Bothrops atrox was the main snake type involve in envenomation in this study. Treatment is very expensive, causing hardship in an already finance limited health system. Further studies need to be carried out to establish the real economical impact of this disease and better ways to prevent it.
Title: Opioid Taper Initiative: An Interdisciplinary Quality Improvement Project

Authors: Michelle Faierman, MD; Ingrid Block-Kurbisch MD, FACP; Stephanie Hsu, BS, Department of Internal Medicine, St. Mary’s Medical Center, San Francisco

Introduction: In a national sample of VA patients with chronic pain, death due to overdose occurred in those prescribed an average of 98 MME (morphine milligram equivalents)/day, while low risk was associated with an average dose of 48 MME/day. In response to these findings, the Sister Mary Philippa Clinic (SMPC) launched an interdisciplinary quality improvement initiative to enhance the safety and care of patients with chronic pain through standardized monitoring, education, and tapering of high-dose opioids.

Methods: In 2017, SMPC initiated a collaboration among residents, attending physicians, and clinic staff to consistently adhere to CDC guidelines for monitoring patients on chronic opioids. To avoid bias inherent in selective patient testing, all chronic pain patients undergo initial evaluation to assess high-risk behaviors, as well as urine toxicology screens and “Patient Activity Report” (PAR) reviews every 3 months or more. EHR templates were created to standardize documentation of pain management visits. Tapering regimens were initiated for patients on ≥ 90 MME/day. Patients were offered a free course on mind-body skills and yoga at SMPC. Additionally a resource binder, which lists many free and low-cost non-pharmacological therapies in the community, was created. Education about risks of high-dose opioids was provided through training workshops, interdisciplinary behavioral medicine conferences, handouts, and posters displayed in the exam and waiting areas. A baseline survey was administered to residents, assessing their confidence in managing patients with chronic pain, and this was repeated after a year to evaluate for correlation between effective implementation of the opioid taper initiative and residents’ reported confidence with chronic pain management. A patient questionnaire will be administered (within patient focus groups or during individual interviews with patients) to explore patient perspectives on opioid tapering. We hope to use the Results: to improve psychosocial support for patients and increase patient engagement with their providers.

Results: Of the 90 clinic patients on chronic opioid therapy at project initiation, 30 (33%) were prescribed ≥ 90 MME/day. Of these 30 patients, 11 (37%) are undergoing tapering, 3 (10%) have completed tapering, 3 (10%) have died, 3 (10%) are taking methadone from an outside substance abuse clinic, 4 (13%) have discussed a taper but have not started it, and 6 (20%) have not yet discussed a taper. The project goal is a 10% reduction in MME/month for all patients on chronic opioid therapy, with 85% of patients below 90 MME/day within a year. Residents and faculty now more regularly follow recommended screening procedures as compared to prior practice.

Conclusion: The expected outcome of this initiative is that systematized monitoring and an interdisciplinary, collaborative approach to pain management will improve patient-provider relationships, change the opioid prescribing practices of our residents, increase patient safety, and improve quality of life for our chronic pain patients.
Title: Delirium is a major predictor of hospital readmission

Introduction: Delirium is an acute change in mental status affecting more than 7 million hospitalized patients in the United States annually. While prior studies suggest an association between delirium and worse clinical outcomes, these investigations have not focused on post-discharge health care utilization.

Methods: The study population included all adults at least 65 years old hospitalized from September 2010 to March 2015 at a Kaiser Permanente (KP) hospital and were discharged alive. Subjects with alcohol withdrawal were excluded. Nursing staff screened the majority of hospitalized patients for delirium with a standard tool; a staff psychiatrist evaluated most of those screening positive. Patients diagnosed with delirium were compared to patients not diagnosed with delirium during the study period. Study data were derived from the KP electronic medical record, which uniquely detailed hospitalization and post-hospitalization data due to integration of insurance coverage and medical care for KP members. To account for differences between groups that might affect outcomes, we calculated propensity scores for delirium based on patient demographics (age, sex), and admission clinical characteristics (admission type, urgency and ward, illness classification and severity indices), for the primary analyses, which involved inverse propensity of treatment weighted (IPTW) logistic regression.

Results: 718 delirious patients (mean age 83±7 years, 57% women) and 7927 non-delirious patients (mean age 77±8 years, 57% women) were included. Delirium during admission was significantly (p<0.001) associated with the following outcomes: hospital readmission within 30 days of discharge (9.7% versus 3.4%; relative risk, RR, 2.9), emergency department visit within 30 days of discharge (28.1% versus 14.7%; RR 1.9), discharge to skilled nursing facility or hospice rather than home (42.6% versus 8.4%; RR 2.3), as well as death within 30 days (1.9% versus 1.3%; RR 1.4) and within one year of discharge (13.7% versus 8.6%; RR 1.6). Delirium was not associated with death during hospitalization.

Conclusion: Delirium is a significant predictor of hospital readmission and emergency department use within 30 days of discharge, as well as discharge to nursing facility and post-hospitalization mortality. Delirious patients are a vulnerable group that should be targeted to reduce post-discharge health care utilization.
California-Research-Poster Finalist  
Rachita Navara, MD

**Title:** Investigating Mechanisms of Persistent Atrial Fibrillation: Are Local Sources Stable?

**Authors:** Rachita Navara, MD, Christopher Kowalewski, George Leef, MD, Fatemah Shenasa, BS, Gabriela Meckler, BS, Tina Baykaner, MD, MPH, Mahmood Alhusseini, MS, Samir Hossainy, Vijay Joshi, Albert J. Rogers, MD, MBA, Junaid Zaman, MA, BMBCh, MRCP, Shirley Park, MD, Paul Zei, MD, PhD, Paul J. Wang, MD, and Sanjiv M. Narayan, MD, PHD, FHRS.

**Introduction:** There is interest in whether persistent atrial fibrillation (AF) is driven by sources, which may be targeted to improve pulmonary vein isolation (PVI). Yet, while many studies show focal or rotational AF drivers, it is unclear if these sources fluctuate over time or differ by proximity to PVs. We hypothesized that local sources may fluctuate even at sites where they are important (e.g. termination of persistent AF).

**Methods:** In 55 patients with persistent AF (61±11 years, 62% men), bi-atrial contact basket/phase maps identified sites where ablation terminated AF before PVI. Sources were localized on electroanatomic maps near (<2 cm) or remote from PVs.

**Results:** Patients had 4.6±1.9 sources identified by mapping: 64.3% rotational and 35.7% focal. One fourth (25.5%) of termination sites were near PVs. Localized sources were identified at sites of AF termination and while individually fluctuating over time, they were present for 68±20% of a 1 minute time interval (300-400 cycles). Rotational activity profile (RAP) quantified the stability of phase singularities, and comparing PV and NPV sources revealed no difference in mathematical presence of phase singularities or spatiotemporal stability (p=0.84)

**Conclusion:** In patients with a defined event - termination of persistent AF by ablation before PVI - sources were found near and remote from PVs. Driver stability by phase was similar for PV and NPV termination sites, and these sources were detectable for roughly two thirds of a given time interval. Fluctuations in local sources over time can explain why studies may or may not show AF drivers, and help reconcile the AF mechanistic debate.
Title: Resident Physician’s Perceived Barriers in Communication and Challenging Authority: A Cross-Cultural Comparison between America and Japan

Authors: Marisa E Rivera, MD1; Chi-Mei Liu, PhD2; Michiko Mizobe MD3; Takashi Shiga MD3, Danny Sam, MD.1, 1 Department of Internal Medicine, Kaiser Permanente Santa Clara, 2 Department of Research, Kaiser Permanente Santa Clara, 3 Department of Emergency Medicine, Tokyo Bay Urayasu Ichikawa Medical Center

Introduction: Effective communication is necessary to reduce medical error and ensure patient safety. This skill is one of the essential competencies for resident physicians during their training, yet it is difficult for resident physicians to question or challenge their superiors. Limited studies explore whether residents feel confident in effective communication, especially in challenging authority during their graduate medical education. Presumably, developing effective communication skills is one major goal of graduate medical education (GME) across different countries. There are few studies comparing cross-cultural GME training on residents’ perception of challenging authority using a theoretical approach. This study has two objectives to fill this gap. First, it compares whether the degree of challenging authority has significant differences among residents in Western or Eastern culture. Second, it identifies factors associated with residents’ willingness to challenging authority using the Health Belief Model (HBM), a model used to help explain and predict behaviors.

Methods: A multi-center cross-sectional study was conducted during April 2016 and June, 2016. Self-administered questionnaires were sent out to Japanese and American resident physicians in the hospital setting from six and one teaching hospitals in Japan and California respectively. This questionnaire was modified from the Operating Team Resource Management Survey and used in previous published literature. We regrouped the measurements of the questionnaire into 6 constructs of the HBM, including perceived susceptibility, perceived severity, perceived benefits, cue to action, and self-efficacy. Wilcoxon rank sum test was used to examine whether there is any differences in response trends between Japanese and American resident physicians. A logistic regression model with a generalized estimating equations approach was used to explore whether there is a significant association between HBM constructs with residents’ willingness to take risk of challenging authority. P value < 0.05 represents significant differences. All statistical analyses were performed with Stata 14.0.

Results: The survey was completed by total 196 resident physicians (Japanese=161 American=35), with 81% response rate. Around 33% (19 out of 36 items) of the responded questions showed statistical significances between two different culture-based residents. Perceived high level of severity (OR=2.42, CI=1.21-5.17), and high level of self-efficacy (OR=2.90, CI=1.22-6.90) would have higher threshold for taking risks to challenging authority, after controlling other HBM constructs and demographic characteristics.

Conclusion: Perceived Severity and Self-efficacy play a significant role in resident physicians’ willingness to challenge authority. Developing sensitive strategies for resident physicians to feel comfortable questioning their supervisors could be a future goal for the GME curriculum to decrease medical error and improve overall communication skills in a hospital-based setting.
California-Research-Poster Finalist
Paniz Vafaei, MD

Title: Using Lean Methodology to Standardize Intake, Output and Daily Weight Measurements in Hospitalized Heart Failure Patients

Authors: Paniz Vafaei, Paul D. Marcus, Robert M. Airoso, Mark D. Macbayne, Thomas N. Hackford, Nicole H. Tran, Nardine M. Riegels, Jamal S. Rana

Introduction: The importance of accurate measurement of fluid intake and output (I/O) and daily body weight has been well established as part of the American Heart Association Guidelines for the management of Heart Failure (HF). Variation in consistency and accuracy of I/O and daily weight measurements for hospitalized HF patients remains an operational challenge and leads to increased mortality, length of stay, and readmissions.

Methods: At our tertiary care medical center, we applied Lean methodology to improve the I/O and weight measurement workflow of patients with HF admitted to medicine services. Process maps were used to identify key decision-making points and areas of waste and variation. Using a multi-disciplinary approach, a team of physicians, nurses, pharmacists, dieticians and case managers performed a rapid improvement event (RIE). During the RIE, best practices were used to improve each stakeholder’s workflow. This was then disseminated using visual management techniques that tracked the teaching and application of workflows to staff members. Nursing rounds and audits were implemented as part of a daily management system to encourage sustainment. One floor, where standardization and sustainment efforts had been implemented, was selected as a pilot floor. The remaining three hospital floors were used as reference floors.

Results: In the study period from April to June 2017, the frequency of I/O and weight measurements in HF patients on the pilot floor (n=19) was compared to HF patients on the reference floors (n=70). Daily weights were recorded in 90.6% of patients on the pilot floor compared to 77% on the reference floors. Additionally, daily intake and output was documented in 100% of patients on the pilot floor compared to 64% and 66% respectively on the reference floors.

Conclusion: Our initial data suggests a trend towards improvement of I/O and weight measurements after implementation of new workflows. Current efforts are focused on ongoing data collection, staff education and visual tracking in order to encourage sustainment, while future steps include expansion to additional hospital floors. Future studies to assess impact on outcome measures such as 30-day readmission rates and length of stay are planned.
California-Research-Poster Finalist
Starleen Frousiakis, MD

Title: Cardiac conduction in the setting of mitochondrial dysfunction in Leber’s hereditary optic neuropathy (LHON)

Authors: Frousiakis SE1,7, Tran JS1,11, Asanad S1,2, Karanjia R1,2,3,9, Gale J1,8, Conrad GL10, Moraes Filho M4, Salomao SR5, Belfort R5, Chicani F5, Quiros PA1,2, Christianakis S7, Carelli V6, Sadun AA1,2,1. Doheny Eye Institute, Los Angeles, CA, 2. Doheny Eye Centers, Department of Ophthalmology, David Geffen School of Medicine at UCLA, Los Angeles, CA, 3. Department of Ophthalmology, University of Ottawa, Ottawa, Ontario, Canada, 4. Instituto de Olhos de Colatina, Colatina, Brazil, 5. Department of Ophthalmology, Federal University of Sao Paulo, Sao Paulo, Brazil, 6. Dipartimento di Scienze Neurologiche, Università di Bologna, Italy, 7. Department of Internal Medicine, Huntington Memorial Hospital, Pasadena, CA, 8. Department of Surgery and Anaesthesia, University of Otago, Wellington, New Zealand, 9. Ottawa Hospital Research Institute, Ottawa, Ontario, Canada, 10. Department of Cardiology, Huntington Memorial Hospital, Pasadena, CA, 11. Department of Internal Medicine, Keck School of Medicine of USC, Los Angeles, CA

Introduction: Leber’s hereditary optic neuropathy (LHON) is a maternally-inherited mitochondrial DNA (mtDNA) mutation resulting in impaired expression of Complex I, an integral membrane protein of the electron transport chain, and leading to rapid onset of bilateral vision loss. LHON commonly presents in young men as subacute painless central vision loss in one eye, followed by loss in the second eye weeks to months later, and visual acuity often falling to 20/200 or worse. Systemic neurological and cardiovascular manifestations of LHON have been described, such as a history of palpitations, syncope or abnormal electrocardiography (EKG) findings, but are rare relative to other mitochondrial optic neuropathies. The aim of this study was to compare cardiac conduction in subjects with LHON 11778 mtDNA mutation with controls using EKG.

Methods: From the largest-studied pedigree of LHON, 11778 mtDNA mutation (haplotype J, sharing a common Italian maternal ancestor), two patient cohorts comprised of affected patients (n1=42) and asymptomatic carriers (n2=44) were established, and these individuals were compared to a Brazilian-derived database of healthy subjects (n3=100). Each participant underwent EKG testing with measurement of PR interval and QTc duration. QTc was calculated using the Bazett formula, QTc = QT/√RR, where RR = 60/HR; HR= heart rate in beats per minute. EKGs were interpreted by a blinded cardiologist and assessed for the presence or absence of a delta wave.

Results: The PR and QTc intervals in both affected and carrier populations of the LHON 11778 mtDNA mutation demonstrated non-normal distributions. Non-parametric ANOVA analysis of PR and QTc intervals demonstrated significant associations between various groups, including between the affected and carrier groups. Mean PR intervals in affected, carrier and control populations were 135.5±31.6 ms, 129.3±37.3 ms and 134.7±25.0 ms, respectively. The majority of subjects in the carrier group demonstrated a PR interval of 120 ms, but there was a clustering of both affected and control groups with PR interval < 120 ms. None of the subjects demonstrated a delta wave. Mean QTc intervals were 384.7±58.7 ms, 402.0±34.1 ms and 302.0±111.9 ms, respectively.

Conclusion: The data support previous findings of an association between cardiac conduction abnormalities and LHON, however, this study identified significant differences between affected and carrier groups. The exaggeration of effect seen in carrier subjects, including PR interval shortening and QTc prolongation, may provide insight into understanding the pathophysiological link between mitochondrial dysfunction and cardiac conduction. Mitochondrial DNA copy number and mitochondrial
mass have been shown to be greater in LHON carriers of the current pedigree, when compared to affected subjects with vision loss. In turn, increased mitochondrial biogenesis in carrier subjects could dampen the pathogenic effect of this mutation. Sub-clinical cardiac conduction abnormalities within a minority of the LHON pedigree suggests that intermittent cardiac assessment may be warranted.

References

Title: The Voice of the Veterans: A Focus Group Approach to Improving Veterans' Inpatient Experiences at an Academic VA Medical Center

Authors: Kevin Hsu MD-VA Greater Los Angeles Healthcare System, Cedars-Sinai Medical Center, Shanon Peter MD-VA Greater Los Angeles Healthcare System, UCLA Medical Center

Introduction: The mission of the Veterans Health Administration (VHA) is to “serve and honor the men and women who are America’s Veterans.” Healthcare teams strive to provide a satisfactory experience for Veterans, and they continually seek to improve the quality of inpatient care at VA hospitals. However, quantitative data in patient surveys don’t offer concrete suggestions. Therefore, we utilized qualitative focus groups at our institution, VA-Greater Los Angeles (VAGLA), both to generate hypotheses for local improvement and demonstrate the applicability of this methodology towards inpatient quality improvement.

Methods: Quantitative surveys have universally pinpointed Communication as a poor-performing domain at the VA. We decided to hone in and formulate a list of questions based on this target domain to guide our focus group. The meeting was a two-hour session at VAGLA in January 2017, consisting of eleven participants led by one non-medical facilitator, along with two non-participating physician observers who kept record. Inclusion criteria were former inpatients recently discharged from VAGLA within the previous six months. Exclusion criteria included those unfit to provide fruitful discussion (patients with dementia, mental health issues or severe delirium). The physician observers subsequently analyzed the discussion contents.

Results: Four overarching themes arose from our focus group discussion. The first theme detailed physicians’ disconnect from communicating the plan of care to the patient. Participants didn’t feel they were “let in” on the entirety of the conversation with the healthcare team. The second theme addressed the uncomfortable feeling when the entire healthcare team arrived into the patient rooms as a “herd,” a term coined by one member and used avidly by others. Participants didn’t understand why there were so many healthcare providers involved. The third theme involved a feeling of being treated like “guinea pigs”, not being told the rationale behind treatments and feeling abandoned with no idea how to follow up. The final theme covered poor communication amongst healthcare providers. Participants felt like the various medical and surgical teams didn’t talk to each other, and had doubts about them all following a unified plan of care.

Conclusion: The “Voice of the Veterans” focus group study was a hypothesis-generating qualitative exercise dedicated to improving the inpatient experience for Veterans at VAGLA, from which four thematic areas for improvement were gleaned, as detailed above. Progress can be made via promoting awareness and educating our physicians. Future directions include holding workshops to encourage proper communication habits amongst the residents training at VAGLA. Although the effects of such a workshop might be difficult to precisely quantify, it may be most appropriate for disseminating our qualitative conclusions. Altogether, the focus group methodology proved effective in gaining important insights into the patient experience.
Title:
Heterogeneity of Treatment Effect in SPRINT by Age and Baseline Comorbidities: The Greatest Impact of Intensive Blood Pressure Treatment is Observed among Younger Patients without CKD or CVD and in Older Patients with CKD or CVD

Authors: Ara H Rostomian, MD, MBA, MPH; Jonathan Soverow, MD, MPH

Introduction: Subgroup analyses from SPRINT demonstrated that older patients (age≥75) treated with an intensive blood pressure control regimen as opposed to standard therapy have a significantly lower risk of cardiovascular events. This was not observed in patients younger than 75 years. Similarly, the risk of cardiovascular outcomes did not differ between those treated with the intensive intervention versus standard therapy among patients with CKD or CVD at baseline. However, it is unclear whether subgroups defined by age and pre-existing comorbidities would benefit from the intensive blood pressure control intervention. Given that the elderly are more vulnerable to adverse effects of intensive blood pressure control, it is important to understand whether the benefits of intensive blood pressure control outweigh the risks in elderly patients with comorbidities. The objective of this study was to evaluate the impact of the intervention on the risk of cardiovascular events in subgroups of patients defined by age and the presence of CKD or CVD.

Methods: As part of the NEJM SPRINT data challenge, we performed two separate subgroup analyses, where subgroups were defined by age and CKD, and age and CVD (i.e., <75 years with CVD; <75 years without CVD; ≥75 years with CVD; ≥75 years without CVD; <75 years with CKD; <75 years without CKD; ≥75 years with CKD; ≥75 years without CKD). Baseline characteristics were assessed in subgroups to determine whether they were balanced between the intensive intervention and standard therapy exposure groups. We used Cox proportional-hazards regression to model time to the primary outcome of cardiovascular events (i.e., first occurrence of MI, ACS, stroke, HF, or CVD death) with the intervention variable as the sole predictor and stratification by clinic site. Hazard ratios with 95% confidence intervals and P-values were reported.

Results: A total of 9,361 patients from the SPRINT trial were included. Intensive blood pressure control was associated with a lower risk of cardiovascular outcomes compared to standard therapy among younger patients without CVD (HR=0.68, 95% CI: 0.51-0.91; p=0.68), younger patients without CKD (HR=0.67, 95% CI: 0.51-0.88; p<0.01), older patients with CVD (HR=0.50, 95% CI: 0.31-0.80; p<0.01), and older patients with CKD (HR=0.65, 95% CI: 0.46-0.94; p=0.03).

Conclusion: The addition of CKD or CVD to age subgroups may modify the impact of intensive blood pressure control intervention on cardiovascular events. Specifically, younger patients without CVD or CKD, may benefit from intensive blood pressure control. Conversely, intensive therapy was associated with decreased risk of cardiovascular outcomes among older patients with CVD or CKD at baseline, but not those without CVD or CKD. It is important to consider patient-specific characteristics, including age and pre-existing comorbidities in order to determine whether patients may benefit from different blood pressure reduction goals and to tailor clinical practice to patients according to their profile.
Title: Smart Prescription Consumer: Comparing Medication Prices Between the U.S. and Mexico

Authors: Henderson Lopez, MD (Internal Medicine Resident), Deyanira Galvan, MD (Internal Medicine Resident), James Hanley, MD (Program Director), Blake Hanley (Overland High School student), Carlos Ramos, MD (Assistant Clinical Professor), Laura Garcia, MD (Associate Program Director), Internal Medicine Residency Program-University of Texas Rio Grande Valley, Harlingen, Texas, Overland High School, Aurora, Colorado

Introduction: Prescription medications are a driver of high cost in our health care system. This is of particular concern for uninsured patients. Elevated prices are associated with medication non-adherence, leading to frequent hospitalizations, readmission and uncontrolled multi-morbidities. An option for patients in Cameron County, Texas, is to obtain medications across the border in Mexico, where it is presumed to be more affordable. These observations prompted us to compare the costs of medications in the U.S. and Mexico.

Methods: A total of 100 patient charts were reviewed. The cost difference for non-OTC prescriptions was established using high cost and low cost prescriptions using Good-Rx® website (proxy for U.S. prescription prices), and compared with the lowest price from either Mexipharmacies (Mexican mail order service) or Farmacias Similares (Mexican pharmacy website). We excluded U.S. medications without a Mexican equivalent version.

Results: 620 unique prescription combinations (different drug types and dosages) were analyzed. 70% of the prescriptions had a Mexican equivalent for price comparison. The difference between high and low Good-Rx prices averaged $238.21 per patient (range: $0 to $2,086.72). The average difference between high Good-Rx and low equivalent cost was $836.30 (range: $0 to $21,057.44). The average difference between low Good-Rx and low equivalent was $598.09 (range: $45.26 to $18,970.72).

Conclusion: Prescription medications can be expensive, and physicians need to be cost conscious. Our analysis showed widely varying price patterns between US commercial pharmacies. For self-pay patients, the difference could represent hundreds of dollars. Price comparison tools can help direct patients to more affordable resources, aid in medical compliance, and perhaps reduce patient multi-morbidities, hospitalizations and readmissions. When adding Mexico to the variables, our hypothesis was not completely correct. Certain medications were more affordable in the U.S., while other medications were more affordable in Mexico. The comparison was not straightforward because there are variations in formulations of common medications (e.g. aspirin 81mg is available in the U.S. while 100mg is available in Mexico) and some medications were not available for comparison. It is important that we educate our patients on how to be a “smart” consumer, physicians should attempt to access cost-effective prescription procurement. We cannot at this time advise patients to obtain prescriptions in Mexico however given our location and demographic our patients do utilize this option.
Title: Tuberculosis Therapeutic Response in Patients with Diabetes Mellitus

Authors: 1. Diego Ramirez-Urizar, Internal Medicine Department, Hospital General San Juan de Dios, Guatemala.
2. Ana Batres, Internal Medicine Department, Hospital General San Juan de Dios, Guatemala.

Introduction: Tuberculosis (TB) and Diabetes (DM) are heavy healthcare burdens nowadays. The close relationship between these illnesses is well known, but there are only a few studies on the therapeutic response. Therefore, this study looks to find the differences, if any, in the therapeutic response at the fifth month (therapeutic failure) to the first line treatment for tuberculosis among patients with DM and without DM.

Methods: Retrospective cohort study.

Medical records from 174 patients with DM and pulmonary TB were evaluated. They were treated with first line treatment at Hospital San Vicente, between the years 2013 and 2015. The medical records from patients with pulmonary DM but without DM were extracted in a 2:1 ratio.

The variables evaluated were time to respond to treatment (by Ziehl-Neelsen stain and Lowenstein-Jehnens), sputum culture, age, and sex.

Results presented as mean time in months, Relative Risk (RR) and Hazzard Ratio (HR)

Results: Out of the 174 medical records from patients with DM, 139 entered the study. The medical records from 278 patients without DM were selected randomly. The therapeutic response time was evaluated among patients with DM and without DM, showing mean response times of 2.58 (+/- 1.62) vs 1.96 (+/- 1.34) months p < 0.05. The RR for therapeutic failure in patients without DM was 0.92 (CI 95% 0.86 - 0.98). The RR between the sexes was 0.95 (CI95% 0.90 - 1.01) p = 0.069. Cox multivariable analysis indicated a HR of 0.74 (CI 95% 0.58 - 0.63) for patients without DM, regardless of age and sex.

Conclusion: This study accept the research hypothesis showing that DM is associated to longer time to therapeutic response and treatment failure, regardless of age and sex. These results can be related to changes to the immune system in a patient with DM. Further prospective studies should evaluated glycemic control with therapeutic response.

References


Connecticut-Research-Poster Finalist
Abhay Singh, MD

Title: Effectiveness of a Cancer Survivorship Clinic: An Evaluation Project

Authors: Abhay Singh MD MPH,¹ ² Tara Sanft MD³

¹Department of Internal medicine and Preventive Medicine, Griffin Hospital, Derby, CT; ²School of Public Health, Yale University, New Haven, CT; ³Medical Oncology, Yale School of Medicine, New Haven, CT

Introduction: In 2014, there were 14.5 million cancer survivors in the United States. In 2024, number is projected to be 19 million. As the number of cancer survivors continue to increase, millions of Americans will require long-term cancer survivorship care. With the increasing number of cancer survivors in US, how important is long term survivorship care? And how effective is it? To answer this question, we conducted evaluation of flagship Yale Cancer Survivorship Program.

Methods: Data collection in this study included both quantitative and qualitative (mixed) measures. For quantitative data, questionnaires were used. These focused on demographics of individuals, improvement in fatigue symptoms, distress levels and improvement in quality of life. Applicability of these questionnaires to clinic cohort was measured using Cronbach’s alpha.

Analysis

Quantitative analysis of patient satisfaction, distress, fatigue and quality of life was reported as mean changes in scores analyzed with paired T test for detecting significant differences in pre- and post-program means.

Results: Demographics The sample size for the retrospective analysis was 366. The clinic primarily cares for the patients with a mean age of 55 years ± 10 years. 90.1% of the survivors are females. Breast cancer survivors are the most commonly seen cancer at the clinic, accounting for 75% of all cancers. 78.4 % of the population served at the clinic is white, followed by blacks and hispanics. The most common cancer types among survivors at clinic included Breast (75.1%), Lymphoma (4.6 %) and Lung (3%).

Brief fatigue inventory We compared the data from the fatigue scale (BFI) between the two visits. On the second visit, participants reported a significant reduction in fatigue levels and effects of fatigue on almost all the measured aspects of daily life.

Distress thermometer We found improvement in distress levels after visit, with a mean difference observed as 2.4, p =<0.001.

FACT-B (Quality of Life) Mean changes pre and post program could not be reported, as clinic offers FACT-B only at the first visit. Several interesting findings were observed with the use of FACT-B; 60% patients didn’t feel they had ample family support, several patients had the fear of recurrence, 70% were experiencing an unsatisfied sex life, 26% felt they had low levels of functionality and most felt they had poor quality of life.

Pilot finding Social media survey – Only 33% survivors attend survivorship clinic after diagnosis.
**Conclusion:** There remains an unmet need to increase awareness regarding survivorship clinics. The provision of a Survivorship clinic results in lesser distress, lesser fatigue and aids in addressing several crucial concerns related to quality of life of survivors. With only 30% survivors seeking long-term survivorship, incorporation of such care models is prudent to increase reach and address issues such as cancer related anxiousness, disturbed personal lives. Whether this results in improved patient-reported outcomes in the long-term needs further research.
District of Columbia-Research-Poster Finalist
Ordessia Charran, MD

Title: Fluid Resuscitation In Critically Ill Patients With Shock: The Impact Of Co-Morbidities

Authors: Ordessia Charran, MD, Sherif Elmahdy, MD, Matthew P Schreiber, MD

Introduction: Achieving adequate filling pressures is a major goal in the treatment of patients with shock. However, specific patients (e.g. those with heart failure (CHF) or end-stage renal disease (ESRD)) may often be under-resuscitated. While prolonged volume depletion from any etiology may progress to distributive shock, other studies demonstrate that a positive fluid balance is also associated with mortality. Our study assesses the association of comorbidities with resuscitation practices in shock.

Methods: We conducted a retrospective study of consecutive medical intensive care unit (MICU) patients requiring vasopressor support over 6 months, from January 2016 until June 2016. The primary outcome was the total resuscitation volume (TRV) administered at 24 and 48 hours. This was analyzed for associations with time to hemodynamic stability, length of stay (LOS) and mortality. Regression analyses were performed to further assess associations with patient characteristics (CHF, ESRD, ARDS, cirrhosis, DNR/DNI status) with TRV and patient outcomes.

Results: 536 charts with 129 subjects requiring vasopressor support at ICU admission were assessed. Septic shock accounted for 78% of shock etiology. 41% of subjects had a known history of CHF and among those patients 1.4L (95% CI 0.06-2.8L) and 1.5L (95% CI 0.1-3.2L) less TRV was administered at 24 and 48 hours respectively. Patients with a primary diagnosis of sepsis received an additional 1.6L (95% CI 0.4-2.9L) and 2.1L (95% CI 0.6-3.8L) TRV at 24 and 48 hours, respectively. The odds ratio for mortality (OR, [95%CI]) in those given > 20cc/kg of volume at 24 and 48 hours was reduced (0.2, [0.1, 0.6] & 0.3, [0.1, 0.7] respectively). There was no difference observed in mortality, LOS or hemodynamic stability at 48 hours when stratified by comorbidity.

Conclusion: Administration of at least 20cc/kg volume resuscitation was associated with improved survival among patients with shock requiring vasopressor support. Comorbidities such as CHF may impact resuscitation practices and additional study is needed to assess if disease-specific resuscitation practices in patients with shock impact all-cause mortality.
Title: A Novel Way of Teaching Focused Cardiac Ultrasound Using 3D-printed Heart Models: a Pilot Study

Authors: Jonathan Segal, MD, Sebastian Ochoa-Gonzalez, MD, Noah Garcia, Ernest Fischer, MD

Introduction: As conventionally taught, focused cardiac ultrasound (FCU) requires mental reconstruction of anatomy using a 2D image. We explored a novel way of teaching FCU using 3D-printed heart models in a hands-on workshop to see if this method could facilitate learning FCU.

Methods: We created 3D-printed heart models in parasternal long and short axis, 4-chamber, and subcostal views. Students were randomly assigned to a traditional lecture or 3D heart workshop (each one hour), then practiced image acquisition on standardized patients and took a written exam assessing their ability to identify cardiac structures on FCU. Three months later, each group had a refresher training session and took a second written exam and an image acquisition practical exam. Primary outcomes were exam scores. Student’s T and Mann-Whitney tests were used to compare groups.

Results: Twenty students were randomly assigned, 10 to each group. On the first written exam, the traditional and 3D heart groups had median scores of 74% (interquartile range [IQR] 27) and 90% (IQR 42), respectively (P = 0.7). On the second written exam, the traditional and 3D heart groups scored 56% and 58%, respectively (95% confidence interval [CI], -23.8 to 18.8; P = 0.81). Results: were similar on the practical with scores of 3.3/5 vs. 2.7/5 (95% CI, -0.17 to 1.36; P = 0.117) for the traditional and 3D heart groups, respectively.

Conclusion: There was no significant difference between the groups. Both methods lead to adequate structure recognition but these skills waned over time. We plan to conduct a larger study enrolling participants at different stages of training to assess the utility of 3D-printed heart models in teaching FCU. Additionally, we would like to identify the ideal retraining interval to avoid skill decline.
Florida-Research-Poster Finalist
Azka Ali, MD

Title: Are we using too much Vancomycin? A single center review of management of febrile neutropenia

Authors: Azka Ali MD¹, Bently Doonan MD¹, Grant Jester MD¹, Jess Delaune MD¹, Jason Starr DO²

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Introduction: Neutropenic fever (NF) is an oncologic emergency, commonly associated with high morbidity and mortality. While mild cases can be treated safely as outpatient, historically NF is associated with inpatient treatment. The Multinational Association for Supportive Care in Cancer (MASCC) can be used to identify high-risk NF patients from low-risk NF patients; however, it is not universally adopted in clinical inpatient practice. As most cases of NF are aggressively treated with inpatient hospitalization and prolonged use of intravenous antibiotics, this results in a significant amount of cost to the healthcare system. The purpose of this study is to identify areas of strength and areas of deficiency in our management of inpatient febrile neutropenia.

Methods: We performed an IRB-approved single center retrospective chart review at University of Florida Health to identify inpatient management of neutropenic fever. We reviewed patients over a two-year span, 2015-2016, admitted to UFH through the Emergency Room. Inclusion criteria included active cancer diagnosis, patients with subjective fevers at home or objective fevers in the Emergency Room (ER), and diagnosis of neutropenic fever by a physician in the ER. Exclusion criteria included prior history of bone marrow transplant. Eighty patients (n= 80) met the above criteria and were included in this study. We used the Infectious Disease Society of America (IDSA) guidelines as the standard of care to analyze appropriateness of antibiotic use.

Results: Our results showed that neutropenic patients and MASCC score > 21 (low-risk group), 55 out of 56 (93.8%) received either a 3rd or a 4th generation cephalosporin or piperillin-tazobactam, and 47 out of 56 (83%) received vancomycin. Of the patients who received vancomycin, only 8 out of 65 (12%) were hypotensive, 44 out of 66 (69%) had an indwelling picc or a port, and 39 out of 67 (58%) had an infectious source identified by the time of discharge. There was no significant use in vancomycin use by cancer type, and there was no significant difference in hospital length of stay.

Conclusion: Our preliminary data show overuse of intravenous vancomycin, potentially leading to increased cost from the use of antibiotic. We aim to develop a NF algorithm at UFH with the contribution of oncology pharmacy and the ER decrease the use of IV vancomycin. We plan to obtain prospective data after our intervention, and compare the outcomes in terms of in-hospital morbidity and mortality, hospital length of stay, and overall cost.
Title: Management of Iron Deficiency Anemia in Free Clinics: A Retrospective Study of Uninsured Patients in Tampa, Florida

Authors: Sean Amirzadeh, Abu-Sayeeff Mirza, Saagar Majethia, Michael Jaglal, Smitha Pabbathi. Department of Internal Medicine, University of South Florida Tampa, Florida

Introduction: It is estimated that IDA occurs in 2-5% of adult men and post-menopausal women in the developed world.1 Another study revealed a 18.5% of pediatric patients who presented with occult anemia in the ED were uninsured.2 The Women, Infants, and Children (WIC) program has historically targeted Iron Deficiency Anemia (IDA) in children by focusing on nutritional and supplementation, but no such program exists for the uninsured in free clinics.3 There is concern that Iron-deficiency anemia (IDA) is poorly managed in resource poor locations and can lead to poor outcomes.4 For example, 9% of patients older than 65 years with iron deficiency anemia have a gastrointestinal cancer when evaluated.5 Free clinics are often faced with economic restraints that limit therapeutic options, diagnostic equipment, and patient’s ability to follow up. There is limited research on the prevalence of anemia, and its management in the free clinic setting.

Methods: A retrospective study was designed to estimate the prevalence of IDA among the uninsured population of Tampa, FL and study its management in a free clinic setting. Demographics, vitals, medications, anemia diagnoses were extracted from medical charts of patients managed in seven free clinics in Tampa, FL, between January to December 2016. Proportion for prevalence, logistic regression adjusted odds ratio, and 95% confidence intervals for associations between socioeconomic factors and anemia are also reported.

Results: From a total of 4670 uninsured patients managed in free clinics in 2016, 382 (5.5%) were diagnosed with some form of anemia. The average age was 38.93 (SD 15.73) with only 12 patients (3.1%) being age 18 and below. These patients had an average of 3.05 visits in 2016. Among these 382, 85.5% (201) were IDA, 5.1% (12) had sickle cell anemia, 3.4% (8) were considered anemia of chronic disease, 3.8% (9) autoimmune hemolytic anemia. Among the 201 patients diagnosed with IDA, only 65.4% (212) were receiving iron supplementation.

Conclusion: Uninsured patients from free clinics are considered outside the health care system and represent a unique population that is understudied in the literature. Adult patients with anemia are more often managed than pediatric patients in free clinics. Patients with IDA are not always receiving supplementation that could improve health outcomes. Education regarding the mechanisms of anemia to free clinic providers may increase awareness and improve management of anemia. Education has shown to increase adherence to iron supplementation and prevent long term effects of untreated anemia.7,8

References

Title: Prognostic significance of dynamic change of PD-L1 expression in Multiple Myeloma patients

Authors: Syed Askari Hasan, MD; Jian Guan, MD, PhD; Renching Wang, MD; Mohammed Wazir, MD; Akriti Jain, MD; Shahram Mori, MD, PhD; Jeff Chang, MD, PhD

Introduction: Inhibition of PD-1/PD-L1 signaling pathway has been shown to be one of the most exciting targeted therapy in fighting both solid organ cancer and hematological malignancies. The expression of PD-L1 in other cancers including squamous lung cancer or Diffuse Large B cell Lymphoma can also serve as a prognostic marker. However, the expression of PD-L1 and its prognostic significance in multiple myeloma is unknown.

Methods: Immunohistochemistry staining of PD-L1 was performed in bone marrow biopsy samples in 39 myeloma patients admitted to one tertiary referral center receiving autologous stem cell transplant (ASCT) at various time points: at the time of diagnosis, before ASCT, post ASCT and/or at relapse after ASCT. >1% cells with PD-L1 staining was considered as PD-L1 positive samples. Data on age, gender, free light chain concentration, cytogenetics, hemoglobin, platelet, calcium level, albumin, b2-microglobulin and survival time were compared according to the status of PD-L1 expression. One way ANOVA test were performed to assess difference in overall survival among different subgroups: PD-L1 negative group, PD-L1 negative switched to positive group, PD-L1 positive group and PD-L1 positive switched to negative group.

Results: In this pilot study, a total of 11 patients out of our cohort (39 patients) were positive for PD-L1 expression (Figure 1). A dynamic change of PD-L1 expression in myeloma is noted (Table 1). Among the patients who were initially negative for PD-L1 staining, 5 patients became positive during the disease course. In contrast, there were 4 patients initially positive for PD-L1 expression became negative during the disease course. 28 patients were persistently negative for PD-L1 expression and 2 patients were persistently positive for PD-L1 expression during the disease course. No significant difference was identified in the basic characteristic including age, gender, initial laboratory workups among these four groups (Table 2). There is a significant difference in overall survival between PD-L1 negative group and PD-L1 negative switched to positive group (p<0.005)(Figure 2). The survival time of PD-L1 negative switched to positive group after switching of PD-L1 status is very comparable to PD-L1 positive group at 14 months.

Conclusion: Our result indicate that immunohistochemistry can be reliably used for measuring PD-L1 expression in decalcified marrow core biopsy materials. This is the first report demonstrating the dynamic change of PD-L1 staining in multiple myeloma. More importantly, this dynamic change has a prognostic significance as shown by the fact that both group with persistently positive PD-L1 expression or group with initial PD-L1 negative but becoming positive during the disease course are associated with shorter survival time. Periodically measuring PD-L1 expression in patients with multiple myeloma may be a good marker for monitoring and prognosis.
Title: Health Outcomes of Uninsured Cancer Patients: A Retrospective Study of Free Clinic Patients in Tampa, Florida.

Authors: Sarah Mushtaq, Abu-Sayef Mirza, Noura Ayoubi, Aldenise Ewing, Michael Jaglal, Smitha Pabbathi

Introduction: There is limited research on the health outcomes of uninsured patients who have been diagnosed with cancer. Free clinics manage patients with a diversity of diseases but few studies document the prevalence of malignancies and follow-up surveillance. Prior studies have revealed Medicaid patients are more likely to be diagnosed with more advanced cancer and less likely to receive cancer-directed surgery and diagnosed with advanced cancer. As a result, they also have a greater risk of death. Socioeconomic determinants such as gender and race have long been researched, especially emphasizing poor outcomes for African Americans.

Methods: Data involving demographics and chronic disease measures were extracted from medical charts of patients managed in seven free clinics between January to December 2016 in the Tampa Bay Area. Proportion for prevalence, logistic regression adjusted odds ratio, and 95% confidence intervals for associations between participant characteristics and cancer diagnoses and screening practices are reported.

Results: There were 172 patients (3.8%) who were identified to have been diagnosed with cancer. Their average age was 54.24 years (SD 13.43). Most common malignancies included breast cancer (51, 31.5%), prostate (13, 8%), melanoma (11, 6.8%), and lung cancer (9, 5.6%). The average length of survival of approximately was 6 years. Regarding management, patients experienced surgery (76, 71%), chemotherapy (5, 4.7%), radiation (6, 5.7%), or just observation (15, 14.2%). Regarding prevention practices, 72 (74.2%) patients completed routine breast cancer screening, 47 (48.5%) routine Papanicolaou smears, 31 (32%) colonoscopies, and 8 (8.2%) routine melanoma skin checks.

Conclusion: Uninsured patients from free clinics are considered outside the health care system and represent a unique population that is understudied in the literature. Cancer patients who are often well connected to resources during the treatment phase of their disease can become lost to follow-up and end up in free clinics for further management of chronic morbidities. The most common malignancies identified in free clinics correlate with national epidemiology. Though most patients received some form of treatment, rates of screening practices are likely lower when compared to their age equivalent counterparts with employment-based health insurance.
Georgia-Research-Poster Finalist
Haris Hatic, DO

Title: Quality Improvement Initiative To Optimize Laboratory Tracking & Patient Notification in Resident Clinic

Authors: Haris Hatic DO, Maighan S. Guffey DO, Nidhip Patel DO & Kimberly Bates MD

Introduction: Laboratory tests hold a vital role in the daily decision-making-process in medicine and it can help shape future therapeutic interventions and identify the cause of an ailment. However, with increased dependence on diagnostic studies there has been an observed prevalence of errors related to missed or unreported test results. The aim of this quality improvement project was to employ PDSA cycle to identify our resident clinic’s average laboratory call-back time and call 90% of all our patients within 10 days for regular test results and 100% with critical results within 1 day.

Methods: This was a multifaceted quality improvement (QI) project with emphasis on the Plan-Do-Study-Act (PDSA) cycle. This study was conducted at our resident clinic, Academic Internal Medicine Partner (AIMP), from July 2016 to June 2017. Any ambulatory patients that had blood work ordered were tracked in this study and this included critical lab values (primarily INR results). Our study was divided in 10 blocks. Blocks 1-4 primarily created a way to track all the laboratory data to provide a baseline analysis of test call-backs. Block 5 residents identified test changes to address any deficiencies observed in prior blocks and created unique PDSA cycles. Blocks 6-10 data was collected on prior changes and new PDSA cycles. During the last block, 40 patients were randomly selected to establish the 10-day follow-up rate for regular test results. For critical lab results, 20 patients were also selected and these values were also compared to the baseline measurement.

Results: Baseline measurement showed there was 82.7% call-back rate for basic labs within 10 days. For critical labs, only 35% of patients were called back within 1 day and 65% within 3 days. 25% patients were not contacted past 2 months. After the PDSA cycle with the necessary clinic modifications a new call-back rate of 95% was achieved within 10 days. For critical labs the 1-day call back rate was 100%.

Conclusion: As errors are becoming more prevalent in the ambulatory setting there needs to be a concerted effort by primary providers to undertake standardized reviews to identify the scope of the problem and initiate solutions to improve patient safety. The reasons why labs are not followed-up are not singular and require detailed examination of once practice for long term process and quality improvement.

References


Title: Are Residents Treating Heart Failure Successfully?

Authors: Timothy Kim, MD; Venkata Gogineni, MD; Nikoloz Shekiladze, MD; Desh Nepal, MD

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Introduction: There is a dire lack of studies assessing the performance of residency-run clinic. Explicit guideline directed therapy (GDT) for heart failure with reduced ejection fraction (HFrEF) are reviewed annually by the American College of Cardiology (ACC) and American Heart Association (AHA). These explicit guidelines offer a way to assess how successfully HFrEF patients are treated in continuity clinics. Continuity clinic present a unique challenge for both the resident physician and the patient population. Resident physicians are challenged with dynamic treatment settings with complex schedules. The patient population often have advanced, chronic diseases with genuine socioeconomic barriers. These are real obstacles to providing optimal care. Thusly, we suspect there is room for quality improvement in treating HFrEF with GDT.

Methods: This is a single center, cross-sectional analysis of patients seen in a resident-run continuity clinic. This analysis is a retrospective chart review through EPIC. Patients were selected for HFrEF < 40% (defined by ACC/AHA Heart Failure Guidelines) and last visit to continuity clinic within the last 3 years. Outcome variables included appropriate guideline directed therapy, appropriate dosing of medications, heart failure treatment teams, and mean dosages compared to their landmark trials. This cross-sectional analysis will measure prevalence of appropriate intervention and will be using co-variance analysis to control for other possible confounding variables.

Results: 4,554 patients were seen in continuity clinic in the past 3 years, 661 patients’ charts were analyzed for diagnoses related to “heart failure”, and of those, 114 patients were analyzed for HFrEF. The average age was 60, 63% (72/114) male and 36% (42/114) female, and 68% (77/114) black, 30% (35/114) white, and 2% (2/114) other. Ultimately, 35% (40/114) of patients’ heart failure regimen were not optimized. Among those not optimized, two variables were statistically significant: 1) most recent clinic visit and 2) the CHF treatment team. Patients were less likely to be optimized when most recent visit was >1 year (46% of patients, p = 0.0395), or when the treatment team does not include all cardiologists, CHF clinic, and internal medicine clinic (24% of patients, p = 0.0360).

Conclusion: There is room to improve in treating heart failure in resident-run continuity clinic. Though there are confounding variables to this analysis, it is clear that assessing performance is the first step in improving patient care. We are compelled to ask why resident-run clinic’s patients are optimized at such a low rate (35%), even with the assistance of a dedicated heart failure clinic. Systemic levels of improvement, as well as individualized resident education are likely the next steps in quality improvement of this study.
References


Title: Low Rates of Osteoporosis Treatment after Hospitalization for Hip Fracture

Authors: 1Edward Nguyen, 2Therese Posas-Mendoza, MD, 3Andrea, Siu, MPH, RAC, 4Jun Ahn Hyeong, Ph.D, 5Sian Yik Lim, MD, 1Lewis and Clark College, 2Department of Internal Medicine, University of Hawaii, Honolulu, HI, USA, 3Hawaii Pacific Health Research Institute, Honolulu, HI, USA, 4Department of Complementary and Integrative Medicine, University of Hawaii, Honolulu, HI, USA, 5Straub Clinic, Hawaii Pacific Health, Honolulu, HI, USA

Introduction: After initial non-traumatic hip fracture, patients are seldom treated with osteoporosis medication for the secondary prevention of osteoporotic fracture. The objective of this study was to understand the trend in osteoporosis care after an admission for hip fracture at 3 community hospitals in Hawai’i within a single health care system.

Methods: A retrospective chart review was conducted (n=428) of patients > 50 years and hospitalized for hip fractures between January 1, 2015 and May 31, 2016 at Straub, Pali Momi, and Wilcox Medical Centers. Basic demographics were collected and medications prescribed were quantified and described within one year of hip fracture. Descriptive statistics were calculated and logistic regression was used to evaluate the association between collected variables and the odds of treatment with medication.

Results: The mean age was 80 years old and 68% were female. Within a year of hospitalization for hip fracture, 26.9% of patients were prescribed medication for osteoporosis, most of which were oral bisphosphonates. Treatment facility and gender were found to be predictive factors for treatment. Patients treated at Pali Momi Medical Center were more likely (OR 2.16, 95% CI: 1.15, 4.16) to be treated than patients at Wilcox Medical Center. Females were more likely (OR 2.46, 95% CI: 1.24, 5.02) to be treated than males.

Conclusion: The use of osteoporosis medication for secondary prevention of osteoporotic fractures was low. Even within the same health care system, there were significant differences in treatment. Efforts need to be made to improve treatment rates, especially among males.
Illinois-Research-Poster Finalist
Widad Abou Chaar, MD

Title: Disseminated cryptococcosis and ibrutinib: A causal relationship?

Authors: Widad Abou Chaar

Introduction: Ibrutinib is a new biologic agent used to treat lymphoproliferative disorders. Recent case reports have suggested that this drug could increase the risk for cryptococcal infections, which may be potentially life threatening.

Methods: Our patient is a 72-year-old man with chronic lymphocytic leukemia (CLL), treated previously with steroids and IVIG, who was admitted for a 2 day history of worsening dyspnea and fatigue. The patient had been started on ibrutinib 420mg daily 3 days prior to admission. On presentation, the patient had high grade fever and decreased breath sounds over the right lung field. He was noted to have purpuric macules and papules on the chest, back, and neck, along with petechiae on the forehead and arms. CT chest revealed a right pleural effusion and mucous plugging. Labs were consistent with thrombocytopenia and neutropenia with absolute neutrophil count decreasing from 1000 at initiation of ibrutinib therapy to 500 on admission. He was started on broad spectrum antibiotics for pneumonia and sepsis, and ibrutinib was held. Cryptococcus was isolated from blood and pleural fluid cultures, and cryptococcus antigen in the serum was positive at 1:32. The patient was started on IV fluconazole 400mg daily and liposomal amphotericin B 550mg daily for fungemia. During his hospital stay, the patient developed septic shock and multi-organ failure; he had ARDS requiring mechanical ventilation and acute renal failure requiring dialysis. He was eventually offered comfort care measures and passed away 9 days after admission.

Results: This case sheds light on many key points. Mainly, it highlights a rarely reported association between ibrutinib and disseminated cryptococcosis. While CLL and treatment with immunosuppressive agents are independent risks factors for developing invasive fungal infections, to the best of our knowledge there are only 2 case reports in the literature that discuss the association between ibrutinib and invasive cryptococcus infection. Ibrutinib, a new biologic agent, works by inhibiting Bruton’s tyrosine kinase, a kinase critical for B-cell survival and proliferation. It is hypothesized that by blocking this enzyme, immunodeficient patients are unable to limit infection to the lungs, with reduced uptake by macrophages, leading to systemic disease progression and dissemination to the brain. However, our patient had neutropenia on admission, which could be another risk factor for developing a severe fungal infection. Therefore, more studies are needed to investigate the exact immune implication of Ibrutinib in the dissemination of cryptococcal infections.

Conclusion: This case illustrates the importance of considering fungal infections early in the course for patients on ibrutinib and other biological agents. Our case suggests it might be crucial for patients on ibrutinib to be started on anti-fungal therapy early on, as disseminated Cryptococcus infection carries with it a high morbidity and mortality risk.

References

Title: Effective communication between AMS (Anticoagulant Management Service) and Internal Medicine residents

Authors: Muhammad Ajmal, MD Holly Rosencranz, MD James Kumar, MD

Introduction: Anticoagulant management service (AMS) clinics are designated to follow the patients who are taking anticoagulants, primarily warfarin, for various indications. The international normalized ratio (INR) is used to monitor the therapeutic level of warfarin. These clinics follow the INR and communicate Results: to the designated physicians caring for patients on this anticoagulant regimen. This is a critical aspect of patient safety and quality health care. Internal medicine residents follow a significant number of out-patients and are responsible for the management of their anticoagulation. For various reasons, including resident clinical assignments and other scheduling challenges, the communication between residents and AMS staff had not been as effective as compared to attending physicians in our program. Specifically, nurses found delays in residents’ response to pages regarding INR values, and confusion regarding responsibility for the Results::

Methods: We conducted a pre and post-survey.

In the pre-survey, we distributed the questionnaire to both AMS staff and residents to find out the top factors which are leading to a delay in response to a given INR lab result. After implementing following interventions we did post survey from AMS staff.

1. Introductory informational lecture by AMS staff to the new intern class and available senior residents in July at the start of the academic year.
2. Identification of a resident liaison who was always available as a backup to address lab Results::
3. Residents assigned to a half day rotation at AMS clinic.

Results:

Pre-Survey

We found out that the main reasons were: 1. Lack of knowledge among the residents about the AMS clinic, 2. No identified back up resident who would be responsible for the Results: and who could be approached if there is no response from the resident.

Post Survey

1. AMS staff indicated a significant improvement in communication which was mostly attributed to the existence of the resident liaison.
2. Residents also indicated a better understanding of anticoagulant management in their outpatients which was mostly attributed to the half day spent at AMS clinic.

Conclusion: We conclude that we can significantly improve optimizing communication between residents and AMS staff. Our data reveal several strategies to assure this. While residents benefited by gaining education from AMS staff through a didactic information session, they credited experience in a dedicated half-day session at the AMS clinic, working directly with AMS staff and patients. However, it was the assignment of a designated resident liaison, who was always available as a back-up to provide
orders and recommendations regarding INR, which was the most significant factor in the improved overall communication. This increased the communication without delay and overcame inherent challenges regarding resident availability and schedules. We predict that these strategies can benefit other training programs that may encounter similar challenges.
Illinois-Research-Poster Finalist
Clark T Bach, MD

Title: Clinicians’ Learning Sources and Adherence to Guidelines Regarding Individualizing Glycemic Goals

Authors: Clark Bach, Dragana Radovanovic, Syed Zia Shah, Thalia Kamel, Charles Lawler, John D. Yoon, Neda Laiteerapong

Introduction: ADA guidelines recommend individualizing glycemic (A1C) goals for patients with diabetes based on patient characteristics and preferences. Effects of glycemic control vary with diabetes duration and history of complications. Intensive control soon after diagnosis is associated with reduced complications for two to three decades. In contrast, in patients with diabetes for ten years, predisposed to cardiovascular disease, intensive control may cause harm and mortality. We looked at how clinicians' self-reported practices relate to current guidelines.

Methods: In 2015, a survey was mailed to endocrinologists and primary care physicians at an academic medical center (AMC; University of Chicago) and the AMC-affiliated community health system (North Shore University CHS). Main outcomes were physicians' 1) Source of learning about individualizing goals 2) Degree of difficulty individualizing goals 3) Reasons for difficulty 4) Factors considered when managing goals. We also considered these variables: clinical site, gender, specialty, years in practice, practice size, and percent of patients 65 or older. Statistics were calculated using means and proportions as appropriate. Bivariate relationships between physician/practice characteristics and outcomes were tested using chi-square tests.

Results: Response rate was 73% (156/213). Nearly 40% reported learning about individualizing goals from journal articles; 30% each learned from educational seminars and guidelines. About 20% each learned online or from colleagues. Individualizing goals was described as very challenging by 45% because of patient preferences (26%), vague recommendations (22%), or insufficient patient information (21%). There were significant differences depending on practice site. AMC physicians were more likely to learn from colleagues (41% vs. 16%, p=0.001), and were also more likely to report learning from published guidelines: American Diabetes Association (45% vs. 20%, p=0.001), American Geriatrics Society (14% vs. 4%, p=0.016), and Choosing Wisely (10% vs. 2%, p=0.026). AMC physicians were more likely to report that individualizing A1C goals was a challenge due to vague recommendations (41% vs. 16%, p=0.001), not knowing patients' duration of diabetes (24% vs. 5%, p=0.001) and hypoglycemia risk (33% vs. 8%, p<0.001), and not knowing how to estimate life expectancy (43% vs. 7%, p<0.001). AMC physicians were more likely to consider life expectancy (79% vs. 50%, p=0.002), while CHS physicians were more likely to consider diabetes duration (23% vs. 5%, p=0.009) and history of diabetic complications (57% vs. 38%, p=0.04). Only the difference in clinical site (but no other physician/patient characteristics) was consistently associated with the outcome measures.

Conclusion: In our survey, adherence to guidelines regarding individualizing A1C goals varied significantly by clinical site. This may be because of closer interactions between specialists and primary care physicians in AMC settings. Interestingly, the AMC physicians reported greater difficulty individualizing glycemic goals, possibly due to higher complexity of patients or to inherent differences in physician characteristics. Developing system-level interventions may help decrease physician-level variation in adherence to guidelines for individualizing glycemic goals.

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Acknowledgements: Special thanks to Dr. Catherine Meyer.
Illinois-Research-Poster Finalist
Negar Faramarzi

Title: The Effect Of Modified Early Warning Scoring System (MEWS score) On Rapid Response Team Call Outcome

Authors: Negar Faramarzi, Amishi Parekh, Ekta Shrestha, Hugo Macchi Cattoni, Shahriar Dadkhah

Introduction: Rapid response teams (RRT) have been implemented since 2004 in many hospitals across the country. However, the usefulness of this intervention has been in question. A modified early warning system (MEWS) was implemented in 2007 and has been a useful tool to enable the early recognition of patient deterioration. In 2006, we completed a study which demonstrated that implementation of RRT’s did not affect mortality at our institution. The objective of this study was to determine if MEWS implementation had an effect on outcomes.

Methods: We completed a retrospective chart review of every RRT since the implementation of our electronic medical record from November 2011 through June 2017. We evaluated the data before implementation of MEWS from November 2011 to January 2015 and after the implementation of MEWS on Non-ICU floor from January 2015 to Jun 201

Results: There was a total of 976 RRT’s called on 898 patients during this period. The average patient age was 68 years old and 51% were Male. 639 RRT’s (65%) were called on patients admitted to the floor from the ED, 206 RRT’s (27%) were called on patients transferred to the floor from the ICU and 53 were called on patients admitted directly to the hospital. A significant proportion of RRT’s was called within 24 hours of transfer, occurring in 37% of ED transfers, 36% of ICU transfers and 32% of direct admissions. Of the 898 patients who were RRT’d, 17% died, 41% of patients were discharged home with self-care, and the remaining went to hospice or to a nursing home by study end. After implementation of MEWS, there were proportionally more RRT’s called, however, there was a statistically significant increase in patients discharged home to self-care and there was a trend towards reduction in mortality. There were proportionally less RRT’s during the night shift (11 pm to 7 am) where there was a lower nurse to patient ratio.

Conclusion: An analysis of RRT’s at our community teaching hospital demonstrated that implementation of MEWS increased the total number of RRT’s called but increased overall patient’s discharged home to self-care and decreased mortality. Therefore, we recommend MEWS score implementation in the ED and ICU prior to transferring patients to the floor.
Illinois-Research-Poster Finalist
Mohammad Luqman, MD

Title: Using the 4Ts Score to Reduce Unnecessary Heparin Induced Thrombocytopenia Antibody Testing; Pre and Post iFORM Implementation in the Electronic Medical Record System at a Community Hospital

Authors: Mohammad Luqman, MD, Sarah Mallet, MD, Priyank Shah, PharmD, Nina Undevia Yedavalli, MD, MPH

West Suburban Medical Center, Oak Park, IL

Introduction: In the United States, one-third of patients are exposed to heparin products while hospitalized. Heparin induced thrombocytopenia (HIT) is a drug reaction that can increase the risk of thrombotic events by 50% with a mortality rate of 20%. The incidence is low at 0.2%. When HIT is suspected, it is recommended to calculate a pre-test probability score, called the 4Ts. 4Ts is a well validated test with a high negative predictive value, therefore a score of 3 or less excludes HIT and no further testing is needed. In those with an intermediate or high score, HIT antibody testing can be considered. We believe at our institution, HIT antibody testing is done in many whose 4Ts score is low. An iFORM that allows for the calculation of the 4Ts was implemented in the electronic medical record system (EMR) to remind physicians when trying to order the HIT antibody test. We also reviewed the patient charges associated with the improper ordering of HIT antibody before and after the iFORM was implemented.

Methods: Retrospective chart review identifying patient that were admitted to our hospital from January 1, 2015 to December 31, 2016 (pre-iFORM) and February 1, 2017 to June 30, 2017 (post-iFORM) who had HIT antibody testing. The iFORM was implemented in January 2017. 4Ts were calculated by evaluating the timing and degree of thrombocytopenia, timing of the decrease in platelet count, presence of thrombosis, and other potential causes of thrombocytopenia. Patient costs associated with testing for HIT antibody were calculated.

Results: 57 patients were identified in the pre-intervention cohort of which 45 (79% of patients) had a low 4Ts score. Total patient costs were laboratory testing was $43,542.45. 12 patient had intermediate and high score. Patient costs for those patient were $12,116.31. In the post-intervention group, 12 (57% of patients) had a low score and patient charges totaled $11,905.84. 9 patients had a score in the intermediate and high score and patient charges totaled $8,708.49.

Conclusion: Prior to the iFORM, $31,931.13 were incurred due to improper testing. Post iFORM revealed that there is significant HIT antibody testing being done, despite a iFORM that guides the ordering physician to complete the 4Ts testing prior to proceeding with HIT antibody testing to avoid testing in those with low scores. These was a waste of $11,905.84 within 5 months. The average 4Ts score was 0.8 in the pre-intervention group and 3.09 in the post-intervention group. ABIM’s Choosing Wisely Campaign recommends not to test or treat for suspected HIT in patient with a low pre-test probability of HIT. In
our continued focus on high value care, we need to continue to educate physicians on unnecessary testing and the downstream costs associated with it that burdens our already fragile healthcare system.

References


Illinois-Research-Poster Finalist
Sreelakshmi Panginikkod, MD

Title: A Survey on Gout-Related Knowledge among Internal Medicine Residents

Authors: Sreelakshmi Panginikkod, Rasiya Hashim, Alvaro Altamirano Ufion, Roshanak Habibi, Ahmad Raja, Ehsan Rajabirostami, Niyati Gupta, Venu Gopalakrishnan

Introduction: The prevalence of gout has risen in recent decades despite advancements in therapeutic options due to several reasons, including lack of adherence to treatment guidelines by physicians and patients’ poor perception and adherence to therapy. We aim to assess the beliefs and knowledge of gout management in Internal Medicine residents, to investigate the gaps, so that education programs can target these gaps and address them in a comprehensive fashion.

Methods: A cross-section survey of Internal Medicine residents from three residency programs in Chicago was conducted using a 20-item questionnaire based on current ACP guidelines on gout management. Residents were considered to have “good” knowledge if 70% of the questions are answered correctly. Survey performance was compared to respondent’s’ year in residency, information on guidelines, number of teaching sessions attended, and number of gout patients cared for. We obtained adjusted relative risks (RRs) of good knowledge by estimating a multivariable Poisson regression model with robust variance estimates, adjusted for covariates. Analyses were conducted using Stata, version 14.2.

Results: Of the original sample of 150 residents, 126 (84%) responded to the survey. Good knowledge was demonstrated by only 40% of the respondents. In our survey, two-thirds of the residents reported that their teaching on gout management is inadequate. For acute gout attacks, 70% of the respondents recognized the right therapeutic options. Only half of the residents knew the correct dose of colchicine and other half opted regimens accounting for higher dose of >2 mg/day. During an acute attack, urate-lowering therapy (ULT) was continued by approximately three-quarters (73%) of the residents. Half of the respondents (50%) were aware that anti-inflammatory prophylaxis (54% colchicine) was indicated while initiating ULT, but only one-third offered the prophylaxis for ≥8 weeks. Approximately 60% of residents reported that allopurinol is initially dosed according to the renal function, but only one third (33%) were aware that it must be titrated to the target serum urate level. Less than half (46%) considered the target serum urate level to be less than 6 mg/dl. In multivariable regression analysis, gout related knowledge was found to be higher among residents who attended more than 3 teaching sessions (RR-3.1; P=0.03; 95% CI, 1.09 to 10.9) and who read the guidelines (ACR/ACP/EULAR) on management (RR-1.8; P=0.04; 95% CI, 1.87 to 4.00).

Conclusion: Our study suggests that better dissemination of knowledge on gout management to Internal Medicine physicians in training is needed. We have identified several areas that should be focused: 1) avoidance of high-dose colchicine; 2) initiating anti-inflammatory prophylaxis while starting urate lowering therapy and its duration; 3) initial dosing of allopurinol 4) target serum urate level; and 4) the need to titrate allopurinol to target serum urate level. Education programs targeting these knowledge gaps may lead to better management practices of the upcoming physicians and help in reducing the prevalence of this burdensome disease.

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Illinois-Research-Poster Finalist
Melby Philip

**Title:** Thrombolytic Therapy in Stroke Patients within tPA window: “Are we FAST or merely getting by standards?”

**Authors:** First Author: Resident Dr. Melby Philip
Additional Authors: Dr. Mir Yadullahi, Heather Beckstrom, Devonne Issac, Laura Gallagher, Dr. Olga Zavelsky, Dr. Nadew Sebro

**Introduction:** American Heart Association’s target of Tissue Plasminogen Activator (tPA) administration within 60 minutes to eligible stroke patients is achieved only in 30% of US patients and there is currently a national momentum to improve this rate to at least 50%. Studies have shown that for every 15 minute reduction in door-to-needle times, there was a 5% lower risk-adjusted in hospital mortality and 4% increase of independent function at discharge. Therefore, it is imperative to assess these barriers and implement best practices to reduce door to tPA times.

The aim of the study was to assess the impact of structured changes to our existing hospital stroke care system (intervention) on door-to-needle times for IV tPA administration (outcome).

The interventions were implemented on key barriers to target tPA administration time, that were identified using root cause analysis and focus group discussions on a separate retrospective study, a prelude to our current study.

**Methods:** The study design was a quasi interventional outcomes study with a pre (June 2014-November 2016) and post (December 2016 to July 2017) system intervention comparison. Subjects were patients who arrived to the Emergency Department with clinical suspicion of acute ischemic stroke within 3.5 hours of last known well and were eligible for tPA. Exclusion criteria included hemorrhagic strokes and uncontrolled blood pressure.

Primary outcome of the study was door-to-needle time for IV tPA administration. System interventions to the key barriers were: Telestroke robot implementation, replace full NIHSS assessment by a shorter and more efficient neurological assessment, and a new "Code Stroke Triage Order Set" for ED RN’s to place with CT brain.

**Results:** Our current analysis includes 70 pre-intervention (baseline) and 18 post-intervention subjects. The percentage of tPA eligible stroke patients who met goal of tPA administration within 60 minutes of arrival improved from a pre-intervention rate of 36% to a post-intervention rate of 83%. Pre-intervention mean door to tPA time was 83.2 minutes compared to post-intervention time of 49.4 minutes (33.8 minute reduction).

**Conclusion:** The analysis concludes that by correctly identifying limiting barriers and implementing steps to decrease delays, we can improve door to tPA times. Additional analysis would be useful to examine secondary outcomes such as: increasing the odds of independent function at discharge and reducing the risk of in hospital mortality. Limitations include unequal pre and post sample sizes and geographic bias in regard to patient population.
Title: Quality Improvement Project: Improving the Internal Medicine Resident Paging System

Authors: Anand Ramadorai DO, Katarzyna Mikrut MD, James Schneider MD, Department of Internal Medicine, Advocate Lutheran General Hospital, Park Ridge, IL

Introduction: Quality improvement projects have become an important standard of practice in many hospitals. Many individuals in a hospital, including administration, attending physicians, residents, and nursing staff, are encouraged to identify problems and help come up with solutions ultimately leading to improvement in patient care.

We observed, from anecdotal incidents reported by our fellow residents and nursing staff and from personal experience, that nurses in our institution frequently have difficulty finding the appropriate resident covering a specific patient, which has led to a delay in patient care. There has been a large turnover of nurses at our institution, with almost 30% new nursing staff in the last year, which we think contributes to the problem together with unfamiliarity with the terms that identify which on-call team is covering a specific patient.

Methods: A Plan-Do-Study-Act (PDSA) strategy was implemented. The hospital’s nursing staff and the internal medicine residents were surveyed and discovered that the pager names in our hospital have been confusing to nurses, attending physicians, and residents working with different services at our institution. The confusion worsens as not every patient in our hospital is resident covered. Through our observations, we identified that not all residents title their daily progress notes in the electronic medical record in a consistent manner.

We implemented new pager names to better describe the covering service and contacted the paging system company to update them. We also developed a streamlined reference sheet for the nurses with recommendations on how to quickly find the appropriate covering resident. We instructed our residents to title their daily progress notes in a uniform manner that clearly indicates what service they are representing. We also encouraged nursing staff, residents and attending physicians to report any issues with paging the internal medicine residents via the online safety event reporting system.

Results: One month after implementation of the changes we asked our residents for any feedback and issues with the new changes. They reported an improvement in the amount of appropriate pages they received throughout the day. They did not experience any issues with signing into the appropriate pagers and felt as though the changes were seamless. We also followed up with the nursing staff to see if the new changes helped optimize patient care. Most nurses were familiar with the reference sheet and felt that it helped them identify the appropriate covering resident. There were a couple departments that were not utilizing them so further education has been required.

Conclusion: A Plan-Do-Study-Act (PDSA) strategy is very instrumental in quality improvement projects. This strategy allowed us to identify multiple areas of improvement that involved different members of the healthcare team. This is a very practical approach to implement change and ultimately improve patient care.
Title: Physician Knowledge and Beliefs of Prescription Drug Costs

Authors: Ryan Nguyen, Yao-Wen Cheng, Ann Zerr

Introduction: According to the 2013 National Centers for Health Statistics survey, almost 8% of US adults did not take their medications as prescribed to save money and 15.1% asked their doctors for lower-cost medications. Adverse outcomes may occur when patients are unable to afford medications, and is further exacerbated by inadequate physician knowledge of drug prices. Our objective is to quantitatively assess physician views about prescription drug costs and compare differences in estimates of ten common medication costs between faculty physicians and medical residents.

Methods: Forty-two physicians from two primary care clinics at Indiana University (IU) Hospital were surveyed. Respondents were asked to grade four statements on a 100-point Likert scale (0, strongly disagree; 50, no opinion; 100, strongly agree) pertaining to their understanding of drug costs. They were also asked to estimate the lowest available cash price in Marion County for ten commonly prescribed medications. Price estimates were compared with data from GoodRx.com. Additionally, rates of identifying the four free medications of the ten were measured.

Results: The survey was completed by 57% of those surveyed (n=24). Faculty physicians self-reported a higher understanding of drug costs compared to residents (mean scores 60 vs. 38). Both faculty and resident physicians agreed that it is worthwhile to sacrifice medication efficacy to ensure affordability (mean score 77) and that a patient’s insurance status should effect prescribing habits (mean score 66).

Overall, 40% of estimated drug costs were within 25% accuracy and 44% were within 50% accuracy. Estimates from faculty physicians were within 25% accuracy at a rate (60%) more than twice that of estimates from PGY (post-graduate year)-1 residents (27%). No statistical difference was found between cost estimates from PGY-1 vs PGY2/3 residents. Faculty physician estimates identified 65% of free medications compared to 0% of PGY-1 and 8% of PGY-2/3 resident estimates.

Conclusion: Our survey demonstrates a willingness among physicians to modify prescription medications to fit patients’ financial capacity. However, compared to faculty physicians, residents were significantly less accurate in estimating the cost of ten commonly prescribed medications. Interventions are needed to educate training physicians on drug costs as well as discounted drug lists in Marion County to ensure better patient compliance and outcomes.
Title: Skin Sympathetic Nerve Activity During Therapeutic Hypothermia Predicts Neurological Recovery

Authors: Issa Kutkut, MD; Awayneesh Kumar, MD; Johnson Wong, BSBME; Keith C. Wright, BS, MA; David Adams, BSEE; Michelle Deckard, CNS; Richard Kovacs, MD; Thomas Everett IV, PhD; Peng-Sheng Chen, MD

From the Division of Cardiology, Department of Medicine, Indiana University School of Medicine, Indianapolis, Indiana

Introduction: Therapeutic hypothermia (TH) improves neurologic outcome in patients after cardiac arrest. However, it usually takes several days and multiple modalities of costly neurological testing in the intensive care unit to determine the prognosis. Accordingly, it is highly desirable to develop a marker to predict early prognosis of patients undergoing TH. Our research laboratory previously developed a non-invasive method to simultaneously record electrocardiogram (ECG) and skin sympathetic nerve activity (SKNA) using conventional ECG patch electrodes. Pre-clinical and clinical studies have shown that this new method (neuECG) can be used to estimate sympathetic tone.

The purpose of this study was to record neuECG in patients undergoing TH during the warming phase. SKNA and heart rate are expected to be correlated in a normal sympathetic nerve response. We hypothesize that the correspondence between SKNA and heart rate changes during the warming phase is a good prognostic sign, while an absence of such response predicts significant brain injury or brain death.

Methods: Patients undergoing TH for cardiac arrest were enrolled in the study after consent was obtained from a family member. Conventional ECG patch electrodes on the chest and forearm were used to record neuECG. Clinical data pertaining to patients’ presentation and hospital course were collected from chart review.

Results: A total of 18 patients were included in the study. Of these patients, 6 had neurological recovery (a score of 1-2 on the Cerebral Performance Category Scale; group 1), and 12 had brain death or were terminally weaned for no neurological recovery (group 2). Changes in SKNA corresponded with changes in heart rate during warming in group 1 but not in group 2. The Spearman’s correlation coefficient between SKNA and heart rate was 0.56 ± 0.08 in group 1 and 0.01 ± 0.10 in group 2 (p= 0.003).

Conclusion: One third of the patients undergoing TH achieved neurological recovery. These patients had a significantly better correlation between SKNA and heart rate than patients who failed to recover. Correlation between SKNA and heart rate may be used as a biomarker to predict neurological recovery in patients undergoing TH after cardiac arrest.

References
Title: Safe handoff of high risk patients at the completion of residency

Authors: Maxia McEachron, MD, Grace Griest, MD, Stephen Knaus, MD

Introduction: Resident handoff periods are associated with increased mortality [1,2]. At the completion of residency, each out-going third year resident transitions care of their continuity patients to an upcoming resident. Outpatient handoffs are not well studied and there are no specific guidelines for this process [3]. The last 3 years, we have instituted and evaluated a formal process for safe and effective handoffs of our high-risk patients at the completion of residency.

Methods: At the end of the academic year, outgoing 3rd year residents were provided a list of their assigned patients. Each resident identified at least 5 high-risk patients. High-risk patients could include patients with multiple medical problems, pending test results, polypharmacy, taking high-risk medications or complicated psychosocial issues. An upcoming PGY-2 resident was assigned to receive the patient handoff. A formal handoff process was implemented. Residents were surveyed after the handoff sessions. Patient charts were audited 2 months and 6 months after the handoffs. The chart audit included whether the patient was seen within 2 months of the transition as recommended, whether the patient was seen by the correct PCP, if the PCP was correct in the EHR, and if there was a hospital admission or emergency department visit during the transition period.

Results: For the past 3 years, 90% of residents who completed the survey felt the handoff process was safe and effective. Most residents agreed that having direct communication regarding high risk patients was a useful tool. In 2017, 90% of residents agreed or strongly agreed the process would improve patient care.

Regarding patient care, in 2017, 75 patients were included in this process. After 2 months, 95% of patients had the correct PCP designated in their chart. Thirty-three patients had a visit within the recommended 2 month transition period. Of the 33 patients seen within the transition period, 42% saw the correct designated PCP. Only 12% of patients had an ED visit or admission in the first 2 months.

Conclusion: Each year we do not have full resident participation in the handoff or survey process. This is related to scheduling conflicts. Our data regarding patient care is descriptive and there is no control cohort for comparison. Overall, the handoff process continues to be viewed favorably by residents who participate. For the patients included, 44% were seen during the 2 month transition period and only 12% of the high risk patients had an ED visit or hospital admission during this time. Both of these metrics are promising. We plan to perform the 6 month patient care audits for this year in December of 2017.

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Title: Physician Knowledge and Beliefs of Prescription Drug Costs

Authors: Ryan Nguyen, Yao-Wen Cheng, Ann Zerr

Introduction: According to the 2013 National Centers for Health Statistics survey, almost 8% of US adults did not take their medications as prescribed to save money and 15.1% asked their doctors for lower-cost medications. Adverse outcomes may occur when patients are unable to afford medications, and is further exacerbated by inadequate physician knowledge of drug prices.

Methods: Forty-two physicians from two primary care clinics at Indiana University (IU) Hospital were surveyed. Respondents were asked to grade four statements on a 100-point Likert scale (0, strongly disagree; 50, no opinion; 100, strongly agree) pertaining to their understanding of drug costs. They were also asked to estimate the lowest available cash price in Marion County for ten commonly prescribed medications. Price estimates were compared with data from GoodRx.com. Additionally, rates of identifying the four free medications of the ten were measured.

Results: The survey was completed by 57% of those surveyed (n=24). 21% of respondents were faculty physicians, 46% were PGY-1 residents, and 33% were PGY-2/3 residents. Faculty physicians self-reported a higher understanding of drug costs compared to residents (mean scores 60 vs. 38). Both faculty and resident physicians agreed that it is worthwhile to sacrifice medication efficacy to ensure affordability (mean score 77) and that a patient’s insurance status should effect prescribing habits (mean score 66). Overall, 40% of estimated drug costs were within 25% accuracy and 44% were within 50% accuracy. Estimates from faculty physicians were within 25% accuracy at a rate (60%) more than twice that of estimates from PGY (post-graduate year)-1 residents (27%). PGY-2/3 residents also had higher rates of drug cost accuracy compared to PGY-1 residents within 25% (42% vs 27%) and 50% (47% vs 32%) accuracy, although no statistical difference was found with either range. Faculty physician estimates identified 65% of free medications compared to 0% of PGY-1 and 8% of PGY-2/3 resident estimates.

Conclusion: Our survey demonstrates a willingness among physicians to modify prescription medications to fit patients’ financial capacity. However, compared to faculty physicians, residents were significantly less accurate in estimating the cost of ten commonly prescribed medications. Interventions are needed to educate training physicians on drug costs as well as discounted drug lists in Marion County to ensure better patient compliance and outcomes.

References
Title: Effectiveness of Vagal Maneuvers in Supraventricular Tachycardia: a Network Meta-Analysis

Authors: Jeet Mehta, MD1; Madhu Reddy, MD, FACC, FHRS2; Milan Bimali, PhD1; Robert Badgett, MD1

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Introduction: Vagal maneuvers are recommended as first-line treatment for paroxysmal supraventricular tachycardia (PSVT) before medical intervention to terminate PSVT by the 2015 American College of Cardiology/American Heart Association/Heart Rhythm Society guidelines, however, these guidelines note an absence of a "gold standard" for vagal maneuvers. We evaluated the effectiveness of different types of vagal maneuvers in treatment of PSVT, specifically comparing modified Valsalva versus standard vagal maneuvers.

Methods: Two existing meta-analyses were updated with a search for newer trials using a mix of methods as described at openMetaAnalysis (https://openmetaanalysis.github.io/Supraventricular-tachycardia-treatment-with-vagal-manuvers/). Randomized controlled trials were added that included the Valsalva maneuver for treating supraventricular tachycardia. Studies were abstracted into standardized tables of PICO attributes, Cochrane risk of bias, and results. The strength of evidence was qualified with a Grading of Recommendations Assessment, Development and Evaluation (GRADE) Profile.

Results: Results are online at https://openmetaanalysis.github.io/svt/. Five trials were included: three trials from before 2000 that compared the standard valsalva to carotid sinus massage (CSM) and two recent trials that compared the modified and standard valsalva maneuvers. When the two newer trials were pooled together, the modified Valsalva maneuver is favored with an RR of 2.60 (95% confidence interval: 1.90-3.58) and heterogeneity of I² = 0%. Network meta-analysis indicated that the modified valsalva had a 93% probability of being the most effective vagal maneuver and will convert almost 50% of episodes. The next most effective maneuver was the supine Valsalva; least effective maneuvers were the standing Valsalva and CSM. Using the GRADE framework, the quality of evidence was high due to consistent results in the two trials and the high quality of the larger trial.

Conclusion: The modified Valsalva was the most effective vagal maneuver for achieving normal sinus rhythm. With the evidence is of high quality, we encourage trial of the modified Valsalva maneuver due to its safety and low cost. Execution of the maneuver can be standardized by simply blowing into a 10 cc syringe with sufficient force to move the plunger. If the equipment is not available, a supine Valsalva maneuver should be tried for hemodynamically stable patients.

References


Title: Colorectal Cancer Screening: A Shift Towards a “No Preference” Strategy

Authors: Fredy Nehme MD, Kyle Rowe MD, Layth Al Attar MD, William Salyers MD, Nathan Tofteland MD, Robert Badgett MD - Department of Internal Medicine, University of Kansas School of Medicine

Introduction: Current guidelines for colorectal cancer (CRC) screening offer conflicting recommendations regarding the preference of one screening strategy over another, specifically fecal immunochemical test (FIT) and optical endoscopy (OE). Given that colonoscopy is the most accurate tool for CRC detection, many guidelines recommend a sequential approach for CRC screening starting with colonoscopy before moving to other strategies. However, more recent guidelines are moving towards a “no preference” strategy. Our aim was to perform a systematic review and meta-analysis to evaluate adherence and detection rate of CRC by FIT compared to OE in effectiveness trials. For OE, we included studies of either sigmoidoscopy or colonoscopy.

Methods: We included randomized trials comparing the effectiveness of FIT to OE for CRC detection. We searched Medline, Google Scholar, and Cochrane through May 2017. Effectiveness trials are defined as studies that measured the performance of an intervention under “real world” conditions. Eligible studies were searched for variables of interest including attendance rate and CRC detection. All statistical analyses were performed using the R Programming Language. Outcomes were reported as Relative risks (RR) with 95% confidence interval and heterogeneity was reported as I² statistics.

Results: A total of 7 randomized controlled trials were eligible and included in the analysis. There were a total of 59912 subjects in the FIT group and 63100 in the OE group. Attendance rate was significantly lower for OE compared to FIT (RR= 0.62, 0.49-0.78; I² = 99%). CRC detection was comparable between the 2 groups regardless of OE technique used (RR= 1.25, 0.61-2.57; I² = 44%). Restricting the analysis to subjects who complied with testing, no statistically significant benefit was found for colonoscopy or sigmoidoscopy over FIT for CRC detection. However, a trend towards significance was noted when both OE techniques were combined (RR=1.69, 1.00-2.85; I² = 74%).

Conclusion: While colonoscopy is the most accurate screening test for CRC in efficacy trials, FIT is as effective as colonoscopy for CRC detection under real world circumstances. The effectiveness of screening depends not only on the sensitivity for neoplasia but also on population attendance and other factors that are undetected in efficacy trials. This supports the shift of more recent guidelines towards offering multiple screening options for the patient, specifically FIT and colonoscopy rather than a sequential approach. However, given the heterogeneity of the studies and the absence of long-term randomized trials assessing mortality of FIT vs colonoscopy testing, preference of one strategy over another is controversial and further studies are needed.

References

Title: Procalcitonin Guided Antibiotics in Sepsis: A Rapid Review to Guide Clinical Quality Improvement

Authors: Fredy Nehme MD, Habiba Hassouna MD, Robert Badgett MD - Department of Internal Medicine, University of Kansas School of Medicine

Introduction: Early antimicrobial therapy for severe sepsis and septic shock is associated with decreased mortality. However, overuse of antibiotics is also problematic. Procalcitonin (PCT) is a biomarker proposed for guiding antibiotic therapy. 'Rapid reviews' of studies are increasingly used to underpin decisions for population health when evidence has not been clarified. Currently, local hospitals do not incorporate PCT into treatment algorithms for sepsis. The aim of this project is to use a rapid review to determine the best role for PCT in our septic patients given the discrepancy of available data.

Methods: We performed a living systematic review of the reduction in days of antibiotic usage and other clinical outcomes with PCT-guided antibiotics among patients with sepsis. We used meta-regression to identify cofactors that modulate the association. We searched PubMed, Google Scholar, and the Cochrane Registry to identify randomized trials.

Results: We included 13 randomized controlled trials. Pooling of all trials indicates that PCT-guided therapy, compared with usual care, significantly reduces days of antibiotic therapy (mean 1.26 days; 2 to 0.50). Heterogeneity of Results: and guidance was substantial. Meta-regression suggests that benefit is confined to settings where PCT-based guidance allows antibiotics cessation within 4 days and a trend towards benefit when usual care is seven days or more days of antibiotics. There was no correlation with outcome when a threshold PCT value was used to discourage antibiotics. Mortality was reduced among patients with PCT-guided care. PCT-guided care did not differ from care guided by C-reactive protein. We found no harm from PCT-guided care.

Conclusion: Low quality evidence suggests that PCT-guided therapy may aid antimicrobial stewardship and improve mortality in sepsis. Meta-regression indicates importance of the criteria used in guidance. The lack of benefit in the trial comparing PCT-guided therapy to an alternative protocol, suggests that this finding may be due to “suboptimization” of care in the control group. This rapid review suggests that PCT-guided algorithms may play a role in guiding antibiotic therapy in our hospitals if antibiotics are allowed to be stopped within 4 days according to PCT levels. However, data is heterogenous and further studies are needed.
Title: An Unmet Need: Assessing Financial Education in Medical Residents

Authors: Jesse Richards DO, Caleb J. Scheckel DO, Jessica Newman DO, Kenneth Poole MD MBA, 1. University of Kansas, Internal Medicine, 2. Mayo Clinic Arizona, Internal Medicine

Introduction: Medical education debt is a growing concern for new physicians. The AAMC reported in 2016 average indebtedness of $189,000 for medical school graduates. A paucity of current research exists examining resident physician’s current financial education needs and whether these are being met. In this multicenter cross-sectional study we analyze these uncertainties.

Methods: An IRB-approved anonymous survey was disseminated to Internal Medicine Residency Program Directors. This instrument assessed resident personal loan burden, financial knowledge, formal financial education, perceived importance and desire for financial education, and ability to save for retirement. Data was obtained from 13 programs, with a response rate of 69.6%.

Results: Of 403 unique respondents, 80.2% reported medical education debt, with 64.5% of those reporting debt greater than $200,000. Mean and median indebtedness for respondents were $193,623 and $225,000, respectively. Respondents totaling 42.9% reported education debt was a significant source of stress, with more residents in the top quartile of debt reporting high stress levels (67.2% vs. 27%, p <0.0001).

While 59.1% of residents reported saving for retirement, of those who were not, 87.1% thought they should be. A total of 60.1% of respondents reported their level of indebtedness strongly influenced their decision to save for retirement in residency.

Of respondents, 71.4% noted they would use debt counseling if provided, with significantly more utilization indicated by residents with high loan burdens (89.2%, p = 0.0009). In a concerning trend, residents with high debt (>75%ile) were nearly three times as likely to carry revolving consumer debt as those with low debt (58% vs 20.3%, p=0.0001). Additionally, residents with debt were much more likely to have a budget with little flexible discretionary funds (47.8% vs. 26%, p = 0.0002).

Finally, almost all (95.1%) of residents replied they received little to no finance education in residency (<4 hours) and the large majority (91.6%) of residents thought personal finance education should be included in residency curriculum.

Conclusion: Education debt is a significant stressor for medical residents. Residents with high educational debt loads are also more likely to carry other forms of debt. Internal Medicine Residency training currently offers a trivial amount of education in financial matters and almost all responding residents felt that more should be included. Financial stress is one of a multitude of factors currently contributing to resident stress levels and is an area not currently addressed in a systematic manner by graduate medical education. These stressors need to be addressed as part of a comprehensive response to ever increasing levels of resident burnout.
Title: Lung Cancer Patients ‘Migrate’ to Seek Better Care

Authors: Danh Pham, Christina Pinkston, Malgorzata Oechsli, Maiying Kong, Jorge Rios, Goetz Kloecker

Introduction: Every year a significant population exists of those diagnosed with NSCLC who do not receive initial treatment upon diagnosis and then “migrate” to additional hospital systems before ultimately getting treatment. Migration to different hospitals may play a role in the decision to treat or not-to-treat, and we aimed to evaluate the potential factors that lead to treatment.

Methods: As part of the Kentucky Lung Cancer Education Awareness Detection Survival (LEADS) Collaborative, 29 of 32 Kentucky hospital registries contacted provided NSCLC data of 6,212 out of 10,471 patients from 2012-2014. Variables collected included hospital accreditation status by the Commission on Cancer, patient zip codes, age at diagnosis, stage, overall survival (OS), sex, race, education, income, and insurance status. Treatment included any combination of surgery, radiation, or chemotherapy. Hospital records were matched to Kentucky Cancer Registry records to determine the number of hospitals visited for treatment. Patient treatment and migration patterns were analyzed with a logistic regression model along with additional post-hoc analysis.

Results: Treatment, initially or via migration, was more likely when an accredited hospital was visited (84% vs. 61%). Most patients were treated at their initial hospital (73%). However, among the remaining patients, 36% migrated to a different hospital where most then received treatment (93%). Initially treated vs untreated was significantly associated with Stage I-II disease, insurance status, younger age (66.8 vs 70.1 years), and longer OS (505 days vs 303). Migrating to another hospital was associated with Stage I-III disease, younger age (66.4 vs 72.2 years), longer OS (561 vs 153 days), but also notably associated with initial hospitals missing treatment modalities and patients having private insurance. Patients who were treated after migrating were associated with Stage I-II disease, younger age (65.8 vs 72.8 years), and longer OS (591 vs 151 days). Compared to patients who were treated initially, patients treated via migration lived longer (591 vs 505 days) and particularly had longer survival with stage III and IV disease (563 vs 495 days) and (379 vs 300 days) respectively.

Conclusion: This analysis demonstrates a survival benefit for initially untreated patients who migrate to another hospital. This migration is significantly associated with stage, missing treatment modalities at diagnosis site, and insurance status which suggests that patients intending to seek better care will frequently migrate. They are more likely to receive treatment and live longer if they are insured. Considering the current landscape of changing healthcare policy, it is notable that insurance status plays such a significant role in enabling lung cancer patients to find effective treatment.
Title: Diagnostic Yield of Non-Invasive Testing in Response to Asymptomatic Non-Sustained Ventricular Tachycardia Detected on Routine Pacemaker Interrogation

Authors: Seunghwan Byun, MD^1,2, Ganesh Venkataraman, MD^1, Marc Wish, MD^1, Kevin Bliden, BS, MBA^1, Ted Friehling, MD^1, and S. Adam Strickberger, MD^1; ^1INOVA Heart and Vascular Institute, Electrophysiology, Fairfax, VA, USA; ^2Sinai Hospital of Baltimore, Baltimore, MD, USA

Introduction: In patient with permanent cardiac pacemakers, asymptomatic non-sustained ventricular tachycardia (NSVT) detected on routine pacemaker interrogation can lead to further cardiac testing such as an echocardiogram and/or cardiac stress test to evaluate for deterioration in cardiac function, ischemia, or new valvular dysfunction. The diagnostic yield of such workup has not been previously reported. Since these interventions bear significant cost, indiscriminate use of such testing in this patient population may lead to increased healthcare expenses for the patients and medical system as a whole. The goal of this study is to identify the diagnostic yield of non-invasive testing in response to asymptomatic NSVT detected on routine pacemaker interrogation.

Methods: A total of 178 patients underwent non-invasive testing in response to asymptomatic NSVT on routine pacemaker interrogation between July 2014 and April 2017. Of those, 169 patients were included in final analysis. 9 patients were excluded due to baseline left ventricular ejection fraction of less than 50%. Data were collected and analyzed from prospectively maintained databases, with retrospective chart review, as required.

Results: Of the 169 patients with asymptomatic NSVT on pacemaker interrogation, 137 patients (81%) had an echocardiogram and 86 patients (51%) underwent cardiac stress test, with 68 patients (40%) undergoing both tests.

Among the 137 patients tested with an echocardiogram, 9 (6.5%) were found to have a decrease in ejection fraction greater than 10% compared to a previous echocardiogram. 3 patients (2.2%) underwent an upgrade to either a biventricular pacemaker (1 patient) or biventricular defibrillator (2 patients).

Among 89 patients who underwent a stress test, 8 (8.9%) had a new perfusion defect. 3 patients (3.3%) were prescribed a change in medications and 5 patients (5.6%) underwent a cardiac catheterization, of which 1 patient (1.1%) underwent a percutaneous coronary intervention.

Of the 169 patients, 98 (58%) had right ventricular (RV) pacing % > 90. Eight out of 9 patients (89%) who were found to have a 10% or more decrease in ejection fraction and all 3 patients (100%) who underwent device upgrade had RV pacing % > 90.

Conclusion: Non-invasive testing in response to asymptomatic NSVT detected on routine pacemaker interrogation has a very low diagnostic yield overall. Patients with a high % of RV pacing may represent a population in which such non-invasive testing may be considered.

References
Maryland-Research-Poster Finalist
Yi Zhen Lee, MD

Title: Wild-type Transthyretin Cardiac Amyloidosis is Associated with Increased Physical Activity

Authors: Yi Zhen Joan Lee¹,², Emily Brown¹, Daniel P. Judge¹,³, ¹Johns Hopkins University, ²Sinai Hospital of Baltimore, ³Medical University of South Carolina

Introduction: Wild-type transthyretin cardiac amyloidosis (wtATTR) is a progressive disorder caused by misfolded amyloid fibrils without any known mutations that cumulatively deposit in the heart. This eventually leads to heart failure, often with preserved ejection function. It is seen predominantly in Caucasian men in their seventh or eighth decade. It is unknown whether environmental factors or lifestyle behaviors play a role in the disease pathogenesis. This study aims to identify the effects of physical activity (PA) on the disease.

Methods: In this retrospective cohort study, all patients with wtATTR seen at Johns Hopkins University were sent an invitation to participate in a survey. Each patient was surveyed for their varying levels of physical activity in three different settings: occupational, recreational, and transport (i.e. traveling from work to home or vice versa). We compared data from wtATTR patients (n = 26) with data from healthy controls from the National Health and Nutrition Examination Survey (NHANES) population between 2007-2014 (n = 3370). Recreational PA was divided into vigorous and moderate intensities; all PA were measured by Physical Activity Questionnaire (PAQ) survey and converted to metabolic equivalents in minutes per week (MET-min/week). Two-sample Wilcoxon rank-sum test was used to compare PA between diseased and healthy cohorts. Linear regression was then performed to look for associations between PA in the disease cohort and the age of onset of disease.

Results: We found statistically significant differences in vigorous recreational PA (z = 7.2, p < 0.001), moderate recreational PA (z = 7.4, p < 0.001), and transport PA (z = 2.9, p < 0.003), between diseased cohort (n = 26) and healthy cohort (n = 3370). Analysis of the age of onset of disease, however, did not show any association with the degree of PA in all categories.

Conclusion: This preliminary study demonstrates differences in the amount of PA done between patients with wtATTR and healthy controls. In this small cohort, the increase in PA does not have any effect on the age of onset of disease. Larger studies may help to delineate the associations between PA and wtATTR. If present, this would help better understand the underlying pathogenesis of wtATTR, and may influence management of patients with this condition.
Title: 30 Day readmission reduction and application usage among acute myocardial infarction patients: preliminary findings from the Myocardial Infarction COCombined-device Recovery Enhancement (MiCORE) study

Authors: Francoise A. Marvel1, Erin M. Spaulding2, Matthias Lee3, Jane Wang1, Helen Xun1, Lochan Shah1, William E. Yang1, Ryan C. Demo3, and Seth S. Martin1, 4

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**Introduction:** Digital and mobile health technologies have the potential to transform health care by providing cost-effective patient-centered tools that are accessible to the majority of patients, and empowering patients with easy access to what they need to do and how to do it. To this end, Apple CareKit is an open source framework that allows developers to build patient-centered apps for the iPhone and Apple Watch to help patients learn more about their health and effectively implement their care plan. Our interdisciplinary team of Hopkins clinicians and Whiting School engineers built the 1st cardiologyCareKit application (“Corrie”), which is focused on guiding patients through each step of the acute myocardial infarction recovery process, with an overarching goal of improving outcomes including reducing hospital readmissions.

The objective of this study is to determine whether providing a prevention-focused cardiology software application (app) and smartwatch to post-acute myocardial infarction (AMI) patients during inpatient admission and 30 days post-discharge reduced 30 day readmissions compared to regional readmission rates of 20%.

**Methods:** This is an observational study of AMI patients at the Johns Hopkins Bayview and Johns Hopkins Hospital Cardiac Units. Early in admission, AMI patients are provided with an app and smartwatch, which provides functionality allowing patients to develop medication self-management skills, coordinate follow-up appointments, learn about critical preventive cardiology topics via videos, facilitate pharmacy interaction to access medication, and connect with health resources across the continuum of care. Chart review of electronic medical records at Johns Hopkins Hospital and Johns Hopkins Bayview, and review of CRISP regional data were conducted to determine whether patients were readmitted at 30 days.

**Results:** There was a significant difference between the baseline AMI 30 day readmission in the USA, which is on average at least 20%, and the Corrie-enrolled readmission rate, which was 0% (n = 25). Patients’ data usage shows robust use during the 30 day period, with an average of 24 logins, some signing on as often as six times per day and some every day for over 60 days. A significant number of users are exploring features of the application, with 30% visiting four or more screens per session. An initial cost-saving model demonstrates a saving of $44,000 per 10 AMI patients in which the standard 20% (two patients) are not readmitted at 30 days (based on Center for Medicare and Medicaid reimbursement and penalty costs).
**Conclusion:** Digital health technologies have the potential to transform healthcare by providing cost-effective patient-centered tools that are accessible to the majority of patients, can be developed for responsiveness to a patient’s healthcare needs, and provide real-time information.
Maryland-Research-Poster Finalist
Niloofar Yahyapourjalaly, MD

Title: Genetic Mutation Testing and Smoking Are Changing the Etiologic Profile of Acute Recurrent Pancreatitis and Chronic Pancreatitis: Results from a Multidisciplinary Pancreatitis Clinic

Authors: Niloofar Yahyapourjalaly 1,2, Tina Boortalary 2, Robert A. Moran 2, Mahya Faghih 2, Vikesh K. Singh 2, Internal Medicine Department, Medstar Health 1, Division of Gastroenterology, Johns Hopkins University School of Medicine 2, Baltimore, Maryland, USA

Introduction: The incidence of idiopathic pancreatitis has been previously estimated to be 10-30%. With increasing recognition of the role of smoking and genetic determinants of pancreatitis, the estimates of idiopathic pancreatitis are likely to change. There has been no known well-designed study to fully investigate the etiologic factors of acute recurrent pancreatitis (ARP) and chronic pancreatitis (CP) considering newer genetic mutations. The aim of the present study is to evaluate the prevalence of different etiologies in a cohort of ARP and CP.

Methods: All adult patients with ARP and CP who were referred to the multidisciplinary pancreatitis clinic from 2010 to 2016 were enrolled in the study. ARP was defined as ≥ 2 episodes of acute pancreatitis (AP) with resolution of symptoms between episodes. CP was defined as the presence of acute recurrent pancreatitis and/or chronic abdominal pain with calcification(s) on CT scan and/or moderate to severe ductal changes by Cambridge classification on MRCP/ERCP. Etiologies were classified as: heavy alcohol consumption and smoking ≥ 12 PPD per the NAPS2 study; hypertriglyceridemia; positive pathogenic genetic variant(s) on 4 panel gene testing [PRSS1, CFTR, SPINK1 & CTRC]; biliary; idiopathic; and other (drug induced, autoimmune, neoplastic cyst, trauma). All idiopathic patients underwent gene mutation testing. The presence of pancreas divisum on imaging was recorded but this was not considered a cause of pancreatitis.

Results: A total of 1549 patients were evaluated in pancreatitis clinic at Johns Hopkins Hospital between July 2010 to December 2016. Among 179 patients with ARP and 342 patients with CP, 91 (50.84%) and 140 (40.93%) were diagnosed with idiopathic ARP and CP and offered genetic testing, respectively. A total of 135 patients with ARP and 288 patients with CP were enrolled after excluding 98 idiopathic pancreatitis patients who did not undergo genetic testing. Among patients with ARP, pathogenic genetic variants were identified as most common etiologic factor 31 (23%). Other etiologies in descending order were biliary 20 (15%), idiopathic 18 (13%), smoking 18 (13%), alcohol and smoking 14 (10 %), hypertriglyceridemia 14 (10%) and alcohol 7 (5%). Out of 288 patients with CP, alcohol and smoking were the most common identified etiology for CP 72 (25%), whereas other etiologies in descending order were smoking 64 (22%), pathogenic genetic variant 45 (15%), idiopathic 41 (14%), alcohol 29 (10%), hypertriglyceridemia 8 (3%) and biliary with 4 (1%). Out of 68 patients with pancreas divisum, 18 (26.5%) have been classified as idiopathic pancreatitis while 50 (73.5 %) of patients had other risk factors for ARP/CP.

Conclusion: After consideration of genetic variants and smoking, idiopathic pancreatitis is only found in 13-14% of patients with ARP and CP. This will likely decrease further as more genetic variants are available for commercial testing.
Maryland-Research-Poster Finalist
Wendy Ying, MD

Title: Characterization of Clinical Overlap between Lamin A/C-Associated Cardiomyopathy and Arrhythmogenic Right Ventricular Cardiomyopathy

Authors: Wendy Ying, MD; Cynthia A. James, PhD, ScM, CGC; Brittney Murray, MS, CGC; Emily E. Brown, MGC; Nuria Amat, MS; Julia Cadrin-Tourigny, MD, MSc; Jan M. Griffin, MB BCh BAO; Crystal Tichnell, MGC; Allison Hays, MD; Hugh Calkins, MD; Daniel P. Judge, MD

Introduction: Lamin A/C (LMNA) associated cardiomyopathy (CM) is a genetic disorder caused by mutations in the LMNA gene, characterized by dilated cardiomyopathy, ventricular arrhythmias, and conduction disease. Arrhythmogenic right ventricular cardiomyopathy (ARVC) is also an inherited disorder that shares certain characteristics with LMNA CM, including arrhythmias and heart failure. ARVC is most often associated with desmosomal mutations; however, LMNA mutations in ARVC have been reported. The significance of LMNA mutations in ARVC is debated, and the extent of clinical overlap between LMNA CM and ARVC is not well characterized. Our aim was to investigate the prevalence of LMNA mutations in ARVC and to compare the heart failure (HF) and arrhythmic outcomes of LMNA CM and ARVC.

Methods: To identify LMNA variants in ARVC patients, we retrospectively reviewed 282 patients diagnosed with ARVC by the 2010 Task Force Criteria who underwent LMNA testing using next generation sequencing and confirmatory Sanger analysis. To compare the clinical characteristics of LMNA CM and ARVC, we identified 17 patients with LMNA mutations and cardiac manifestations without ARVC and 34 age- and gender-matched patients with definite ARVC without LMNA mutations.

Results: Among our ARVC cohort, one patient had a LMNA variant (p.Arg399His) at a highly conserved locus that was considered pathogenic or likely pathogenic. p.Arg399His has never been previously associated with cardiomyopathy.

Comparing the clinical outcomes of LMNA CM and ARVC, HF was more prevalent among LMNA CM patients than ARVC patients (71% vs 38%, p = 0.03). Among patients who developed HF, mean left ventricular ejection fraction was 41% in those with LMNA CM and 54% in those with ARVC (p = 0.02). LMNA CM patients experienced more supraventricular tachycardia (77% vs 18%, p < 0.001), whereas ARVC patients experienced more ventricular arrhythmias (85% vs 35%, p < 0.001). 82% of LMNA CM patients versus 44% of ARVC patients experienced conduction disease (p = 0.009). Overall mortality was similar between the two groups. However, the combined outcome of death from any cause and malignant ventricular arrhythmias was worse among ARVC patients (hazard ratio 2.65; 95% confidence interval 1.15-6.09; p = 0.01).

Conclusion: LMNA mutations are a rare cause of ARVC. p.Arg399His is a novel likely pathogenic cause of ARVC that has not been previously reported. Atrial arrhythmias and conduction disease are more prevalent in LMNA CM, and ventricular arrhythmias occur more frequently in ARVC. Recognition of these characteristics which may aid in early diagnosis and treatment of these inherited cardiomyopathies.
Introduction: The convergent procedure is a minimally invasive procedure, developed as an option for management of refractory atrial fibrillation (AF). It involves epicardial ablation of the posterior wall of the left atrium via a transdiaphragmatic approach by a cardiothoracic surgeon and catheter based endocardial pulmonary vein isolation by an electrophysiologist. Our objective is to assess recurrence of arrhythmias within one year in patients who underwent the convergent procedure for AF at a tertiary healthcare facility.

Methods: A retrospective review of electronic medical records at St Elizabeth's Medical Center was performed to identify patients who underwent the convergent procedure. Demographic characteristics, medical history, type of AF (paroxysmal vs persistent) and procedural data including complications were obtained. Our primary outcome was the recurrence of AF or atrial flutter within one year of the intervention using Cox proportional survival analyses. We used Kaplan-Meier non-parametric analyses to depict the event-free probability for this outcome. Secondary outcomes were the need for repeat catheter ablations, antiarrhythmics and anticoagulants. The follow-up period for all outcomes was one year after the procedure.

Results: 17 patients were included in the study. The mean age was 65.4± 7.4 with a male predominance (77%). More patients presented with persistent (76.5%) than paroxysmal (23.5%) AF. The average CHA2DS2VASc was 2.56 ± 1.6. No patients with cardiac tamponade, major bleeding, phrenic nerve injury, or death were identified. Complications were liver laceration (5.9%), and acute kidney injury (11.76%). Length of stay in the hospital was 4.9 ± 1.9 days. When compared to pre-procedural paroxysmal AF, persistent AF was not associated with recurrence of atrial flutter [hazard ratio (HR) 0.60; 95% CI 0.05, 6.66; p = 0.680], AF (HR 0.42; 95% CI 0.07, 2.53; p = 0.344), or either (HR 0.54; 95% CI 0.10, 2.96; p = 0.479). In similar analyses restricted to events after a three-month blanking period, no recurrence of arrhythmia was identified.

Conclusion: The convergent procedure appears to be safe and effective. Success rates are higher than reported success rates in catheter ablation studies including a high percentage of patients with persistent AF.
Massachusetts-Research-Poster Finalist
Ioannis Koulouridis, MD

Title: A Multidisciplinary Approach to Atrial Fibrillation: The Convergent Procedure

Authors: Joe Aoun, MD¹; Ioannis Koulouridis, MD, MS¹,²; Aleem Mughal, MD¹; Maxwell Eyram Afari, MD¹; Caroline Zahm, MD¹; John V. Wylie, Jr, MD¹

¹ Department of Medicine, St. Elizabeth's Medical Center, Boston, MA
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Introduction: The convergent procedure is a minimally invasive procedure, developed as an option for management of refractory atrial fibrillation (AF). It involves epicardial ablation of the posterior wall of the left atrium via a transdiaphragmatic approach by a cardiothoracic surgeon and catheter based endocardial pulmonary vein isolation by an electrophysiologist. Our objective is to assess recurrence of arrhythmias within one year in patients who underwent the convergent procedure for AF at a tertiary healthcare facility.

Methods: A retrospective review of electronic medical records at St Elizabeth’s Medical Center was performed to identify patients who underwent the convergent procedure. Demographic characteristics, medical history, type of AF (paroxysmal vs persistent) and procedural data including complications were obtained. Our primary outcome was the recurrence of AF or atrial flutter within one year of the intervention using Cox proportional survival analyses. We used Kaplan-Meier non-parametric analyses to depict the event-free probability for this outcome. Secondary outcomes were the need for repeat catheter ablations, antiarrhythmics and anticoagulants. The follow-up period for all outcomes was one year after the procedure.

Results: 17 patients were included in the study. The mean age was 65.4 ± 7.4 with a male predominance (77%). More patients presented with persistent (76.5%) than paroxysmal (23.5%) AF. The average CHA2DS2VASC was 2.56 ± 1.6. No patients with cardiac tamponade, major bleeding, phrenic nerve injury, or death were identified. Complications were liver laceration (5.9%), and acute kidney injury (11.76%). Length of stay in the hospital was 4.9 ± 1.9 days. When compared to pre-procedural paroxysmal AF, persistent AF was not associated with recurrence of atrial flutter [hazard ratio (HR) 0.60; 95% CI 0.05, 6.66; p = 0.680], AF (HR 0.42; 95% CI 0.07, 2.53; p = 0.344), or either (HR 0.54; 95% CI 0.10, 2.96; p = 0.479). In similar analyses restricted to events after a three-month blanking period, no recurrence of arrhythmia was identified.

Conclusion: The convergent procedure appears to be safe and effective. Success rates are higher than reported success rates in catheter ablation studies including a high percentage of patients with persistent AF.
Mexico-Research-Poster Finalist
Carlos Nava-Santana, MD

Title: Risk Factors Associated with In-hospital Mortality: A Retrospective Analysis of 80,330 Hospitalizations at the National Institute of Medical Sciences and Nutrition.

Authors: Nava-Santana Carlos A*†, Kershenobich-Stalnikowitz David1†, Sifuentes-Osornio José*, Gulias-Herrero Alfonso*†, Marfil-Garza Braulio*†, *Division of Medicine. National Institute of Medical Sciences and Nutrition Salvador Zubiran., †Department of Internal Medicine. , †General Director.

Introduction: In-hospital mortality (IHM) is one of the primary measures to evaluate the quality of medical care. The National Institute of Medical Sciences and Nutrition is a tertiary care center that attends to adult patients with complex cases from all over Mexico, and is one of the Mexican National Institutes of Health. The aim of this study was to identify the different risk factors associated with IHM at our institution.

Methods:

Retrospective analysis of hospital discharges from 2000-2015 using databases of our institution’s medical records. We compared survivors vs non-survivors using descriptive and inferential statistics.

Results: We analyzed 80,330 episodes of hospitalization. We found a gross mortality rate of 2.4% (n: 1921). Older age, residence outside Mexico City, weekend admission, hospitalization in a shared room, lower socioeconomic level, and the number of comorbidities per patient were associated with higher risk of IHM in a multivariate logistic regression analysis. Starting March 2008, changes in hospitalizations occurred at our institute (the main change was a reduced number of patients per medical team). This change was associated with a decrease in IHM of 19% and a lower risk of IHM after multivariate logistic regression analysis (OR 0.73, CI 95% 0.66-0.82, p<0.001). “Pneumonia” (J10.0-J18.9) and “Complications of Diabetes Mellitus” (E11.0-E11.9) were the most common diagnosis for survivors and non-survivors, respectively. Infectious diseases had the lowest OR for IHM overall. Chronic conditions such as diabetes mellitus, COPD, liver cirrhosis and cardiovascular disease, as well as hemato-oncologic conditions had the greatest OR for IHM. Surgery was associated with a lower risk of IHM.

Conclusion: Information regarding risk factors for IHM in Latin America is scarce and this is the first study analyzing specific risk factors for IHM at the Mexican National Institutes of Health. Herein, we describe key sociodemographic and disease-related risk factors for IHM. This could facilitate, in the future, early identification of patients with a higher mortality risk that could warrant specific measures of care, which could improve outcomes and decrease mortality.
Michigan-Research-Poster Finalist
Ariful Alam, MD

Title: Transforming The Structure and Process of Peripherally Inserted Central Catheter (PICC) Utilization and Ordering Practices: A Quality Initiative

Authors: Alam, A; Ezwawi, S; Zoroya, J; Lilley, K; Misra, P

Introduction: Variability in peripherally-inserted central catheter (PICC) line appropriateness can decrease quality and safety. Our aim in this project was to assess PICC line utilization and ordering practices at St. Mary Mercy to meet the appropriate utilization goal per HMS guidelines.

Methods: A total of 228 patients were studied in this initiative. Inclusion criteria consists of patients at St. Mary Mercy Hospital in Livonia, MI aged 18 years or older who were hospitalized between 8/1/16-12/31/16 with an active order for PICC line placement. Of the 228 patients, 126 underwent proactive chart review where ordering practitioners were contacted for PICC line insertion orders that were for inappropriate reasons per Hospital Medicine Safety (HMS) Consortium guidelines. Immediate feedback via webexchange or DocHalo including positive reinforcement and guidance was provided for order modification.

Results: Of the 228 patients, 49.56% (n=113) were between the ages of 50 and 75, 35.96% (n=82) were over 75 years and only 14.47% (n=33) were younger than 50 years. Of note, 50 % (n=29) of patients over the age of 75 and 41.38% (n=24) of patients between ages 50 and 75 had GFR < 45 compared to only 8.62% of the patients (n=5) below the age of 50 had GFR < 45. Furthermore, 53.07% (n=121) of the orders were placed for patients on the general medical floor as compared to only 12.28% (n=28) were placed on ICU and Step-Down patients. Based on analysis of the data, there was a significant decrease in inappropriate PICC EMR orders from 61% to 38% (p<0.0007). Proactive chart review follow-up with ordering practitioner occurred only in 41% of cases. Of orders requiring practitioner follow-up, there was 21% order modification. We observed a significant decrease in patients with a PICC line indwelling duration of 5 days or less from 19% to 17% (p<0.0370). Furthermore, there was a non-significant decrease in patients with PICC lines with GFRs<45 from 27% to 24% (p<0.53).

Conclusion: Proactive chart review in addition to follow-up with ordering practitioner significantly improved PICC line ordering practices. Specifically, there was a significant decrease in unacceptable ordering reasons for PICC line as well as in number of patients who had a PICC line indwelling duration of 5 days or less. There was also a reduction of PICC line insertion for patients who had a GFR<45 although this was not statistically significant. It is therefore important to educate ordering practitioners about appropriate HMS guidelines for PICC line use. This quality initiative also identifies the need for and availability of a modified and extensive order set in the EMR for practitioners to avoid inappropriate PICC placement in the absence of proactive order review. Furthermore, practitioners should seek alternative options for patients with poor venous access.
Michigan-Research-Poster Finalist
Fatima Asif

Title: Will The Wrong Choice Of Dialysis Modality Kill First Or The End Stage Renal Disease ?!

Authors: F Asif, P Misra, D Steinberger

Introduction: In the US there are approximately 615,000 patients with end stage renal disease, majority of whom require dialysis. US mortality rates for ESRD dialysis patients remain high with a 5 year survival rate of 35-40%. My PICO question aimed to answer mortality benefit of hemodialysis (HD) versus peritoneal dialysis (PD) in ESRD patients.

Methods: Literature research was conducted using PubMed, Cochrane Library and Google Scholar. Filters applied were english language, full text, humans,10 years. Two observational studies, the MacRae study and Han study were selected. The Mac Rae study included a cohort of 458,329 adult dialysis patients in the US on Medicare. Statistical analysis used χ2 test, t test, ANOVA. Univariate and multivariate logistic regression analyses and Poisson regression was performed. In the Han study 13,065 dialysis Korean patients (age≥65 years) were included. Multivariate Cox model was used. Meta-analysis of 15 observational studies was also performed.

Results: In the Mac Rae study out-of-center HD was initial modality in 0.4% of participants. These patients were younger, nonwhite, had fewer co-morbidities and higher median income. They had a higher risk of death compared to in-center HD or PD patients (HR 1.10; 95%CI 1.04, 1.17), however, relative risk of death was lower in younger, healthier patients (HR 0.78; 95% CI 0.61,1.00). In the Han study the Korean PD group had a higher mortality rate than the HD group (HR 1.20; 95% CI 1.13,1.28). In the meta-analysis, PD versus HD, HR was 1.10 (95% CI, 1.01, 1.20).

Conclusion: There is a higher risk of death in elderly patients receiving PD compared to HD. Relative risk of death associated with out-of-center HD is lower than for in-center HD when younger, healthier patients are considered. When a patient has diabetes mellitus or has been undergoing dialysis longer, HD is preferred to PD with respect to survival. Choice of modality should take into consideration patient values and preferences.
Michigan-Research-Poster Finalist
Jasmeet K Bal, MBBS

Title: Process to Improve Cardiac Telemetry Use in A Community Hospital

Authors: Sana Chams, Firas Ido, Mulham Hamdon, Vesna Tegeltija, and Sarwan Kumar

Introduction: Inpatient telemetry monitoring is commonly used to identify arrhythmias, QT prolongation and ischemia. ACC/AHA guidelines identify groups in which telemetry use is indicated (class I), may be beneficial (class II), and unlikely to benefit (class III). Strict adherence to guidelines can improve cost without harm to patients. In PDSA cycle one, a checklist, following ACC/AHA guidelines, was integrated into the EMR. Post implementation review revealed no improvement in ACC/AHA guided checklist due to physician ability to bypass the checklist.

The purpose of the second PDSA cycle is to incorporate documentation for telemetry use and telemetry continuation.

Methods: IHI model to guide and format this study:

Plan: We created a small team that reviewed 30 charts over one month period at our academic institution. Chart review revealed inappropriate telemetry use with zero documentation of physician telemetry review.

Do: We provided an education session describing ACC/AHA guidelines. We evaluated the effect of education on appropriateness of telemetry initiation and continuation.

Study: Post implementation, chart review of another 30 patients over a one month period was done to assess any change

Act: The small pilot study.

Results: After one month of further education to residents, chart review of 30 patients was done; 9 out of the 30 (30%) indicated that telemetry was reviewed and included any abnormalities or alarms noted; a significant difference as compared to chart review prior to education with pvalue of 0.0028 (p-value <0.05).

Conclusion: Using resident-based educational sessions led to improved compliance of ACC/AHA guidelines. The primary objective of our project was to limit the use of cardiac telemetry as per the guidelines. Our team proceeded with this project after reviewing the cost efficiency, the feasibility of limiting inappropriate and unnecessary alarms that may undermine both patient safety and quality of care, and the reduced extent of nurses’ fatigue while improving the patient flow between the emergency department (ED) and other units throughout the hospital.
Michigan-Research-Poster Finalist
Seifeldin Hakim, MD

Title: A Comparison of ERCP Performed Over the Weekend versus Weekday: A Reduction in Length of Stay, Hospital Costs, and Patient Charges.

Authors: Seifeldin Hakim, Resident, Internal Medicine Department, Oakland University William Beaumont, Royal Oak, MI, Andrew M. Aneese, Resident, Internal Medicine Department, Oakland University William Beaumont, Royal Oak, MI, Ahmed Edhi, Resident, Internal Medicine Department, Oakland University William Beaumont, Royal Oak, MI, Mitchell S. Cappell, Chief, Gastroenterology Department, Oakland University William Beaumont, Royal Oak, MI

Introduction: Endoscopic retrograde cholangiopancreatography (ERCP) is a procedure that can be diagnostic or therapeutic for many biliary and pancreatic diseases. The weekend effect is a phenomenon used to describe worse outcomes when patients were admitted on the weekend versus the weekday. Weekend effect has been studied for ERCP with no significant change in outcomes. Similar to other surgical procedures, performing an ERCP requires a significant use of resources including nurses, specialized technicians, anesthesia services, and endoscopist with advanced training. Frequently, hospitals do not have these resources available over the weekend, which leads to postponing the procedure to the first available weekday, generally Monday or Tuesday. Currently there is a paucity of data regarding the ERCPs performed on the weekend vs postponing the procedure to the first available weekday. Our hypothesis is performing an ERCP over the weekend instead of waiting until the next available weekday will positively impact length of hospital stay (LOS), and hospital costs, and patient charges.

Methods: Retrospective review of patients from January 2011 through December 2016 where queried form our institution electronic medical record who had a gastroenterology consult on Friday or Saturday before 12:00 PM, which resulted in an ERCP performed on the weekend (WE) versus postponing it to the first available weekday (WD), Monday or Tuesday. LOS, hospital costs and patient charges were compared between both groups as well as readmission rates. All categorical and continues variables were analyzed with Pearson, chi-squared/Fisher exact test and non-parametric Wilcoxon rank tests respectively.

Results: 535 patients were identified with 315 (59%) patients constituted the WE group and 220 (41%) patients constituted the WD group. Both groups were similar at baseline regarding demographics, medical, and surgical history, and basic laboratory analyses. The median for LOS in WD group was 6.9 days (25th, 75th percentile: 5, 11) compared to 4.5 days (25th, 75th percentile: 3, 7) in WE group, (P<0.0001). Total hospital costs per patient are $9,208 (25th, 75th percentile: 6,288, 12923) in the WE group compared to $11,657 (25th, 75th percentile: $8,454, $18,045) in the WD group, (P<0.0001). Total hospital charges per patient are $28,026 (25th, 75th percentile: $19,698, $39,503) in the WE group compared to $37,899$ (25th, 75th percentile: $26,786, $61,167) in WD group (P<0.0001).
Conclusion: Performing ERCPs on the weekend significantly reduces the length of hospital stay, hospital costs and charges compared to ERCPs that were postponed to the first available Monday or Tuesday.

References

Title: Evaluation of a Pharmacist-Driven Protocol to Reduce Inappropriate Use of Acid Suppressive Medications in the Non-ICU Setting

Authors: Chris Jacob¹, Suceil Sivsammye¹, Tracey Mersfelder¹,²,³, Christin Campbell³, Kevin Kavanaugh¹.
1. Western Michigan University Homer Stryker MD School of Medicine. 2. Ferris State University. 3. Borgess Medical Center

Introduction: Overutilization of proton pump inhibitors (PPIs) and histamine-2 receptor antagonists (H2 blockers) related to stress ulcer prophylaxis (SUP) commonly occurs. Problems associated with the use of acid suppressive medications are well known, such as increased risk of Clostridium difficile infection, pneumonia, electrolyte and vitamin abnormalities, and drug interactions. Adverse events from SUP also create an economic burden to the healthcare system. Our study aims to evaluate the effectiveness of a pharmacist-driven termination protocol to limit inappropriate use of acid suppressive medications in the non-ICU setting.

Methods: Patients were included if they met the following criteria: 18 years of age or older, prescribed a PPI or H2 blocker, and were admitted to the hospitalist service. Patients were excluded if the PPI or H2 blocker was a home medication or if they were admitted to the intensive care unit. Phase 1 evaluated patient charts for appropriate use of PPIs or H2 blockers. Phase 2 was similar to phase 1, except that pharmacists contacted health care providers for medication discontinuation if the acid suppressant use was deemed inappropriate. The primary outcome of this study was the proportion of patients that had acid suppressive medication discontinued before and after implementation of the pharmacist-driven termination protocol. Outcomes were evaluated by the Fisher’s exact test. Descriptive statistics were used to determine the frequency of the acid suppressive medication prescribed and the time the pharmacist spent making the intervention.

Results: In Phase 1, of 132 patients evaluated, 95 were excluded due to home medication use. Ten patients met the criteria for appropriate indications, with eight treated for GERD, one for GI bleed, and one for PUD. Acid suppressive medications was inappropriately prescribed in 26 patients. Of these patients with inappropriately prescribed medications, one had their medication discontinued during the inpatient admission, while nine were prescribed a PPI or H2 blocker at discharge. In Phase 2, of 132 patients evaluated, 113 were excluded due to home medication use. Ten patients met the criteria for appropriate indications, with four treated for GERD, three for GI bleed, one each for PUD, pancreatitis, and gastritis. Acid suppressive medications were inappropriately prescribed in nine patients, eight of which were discontinued based on the pharmacist-driven termination protocol, and none were prescribed on discharge. The difference was statistically significant (p<0.0001) in the use of the medications in the inpatient setting, but it was not significant at discharge (p=0.18).

Conclusion: Inappropriate medication use may cause patient harm and increase overall healthcare costs. This study shows an effective method to reduce the rate of inappropriately used acid suppressive medications using a pharmacist-driven termination protocol with little impact on pharmacist workflow.
The implementation of a pharmacist-driven termination protocol may help decrease the inappropriate use of acid suppressive medications on an inpatient hospital service.

References


Michigan-Research-Poster Finalist
Karthik Kailasam, MBBS

Title: Regional differences in hospitalization outcomes for sepsis across the United States: A National In-patient sample database analysis

Authors: Karthik Kailasam MD1, Marie Ravi Chandar MBBS2, Guramrinder Thind MD1, Prashant Patel DO1, Suceil Sivsammue MD1, Ross Driscoll MD1, 1Western Michigan University Homer Stryker MD School of Medicine 2Madras Medical college

Introduction: Sepsis is the leading cause of death in US hospitals. According to the CDC, one in three patients who die in a hospital have sepsis. The goal of this analysis is to study the differences in hospital outcomes for sepsis, across the different geographical regions in the United States.

Methods: An observational analysis was performed using National Inpatient Sample (NIS) database for the years 2010-2014. NIS represents 20% of all hospital data in the US. Patients were identified by Clinical Classifications Software code (CCS) 2 for Septicemia. The hospital's census region was obtained from the AHA Annual Survey of Hospitals. Z-test was used to statistically compare for differences in incidence, length of hospital stay, mortality and hospitalization cost across the census regions. Graphs were plotted with time (in years) on the x-axis and variables (incidence,length of hospital stay, mortality and cost) on the y-axis. The error bars on the graph represent a 95% confidence interval (CI).

Results: Over the years 2010-2014, the incidence of sepsis in the United States has increased steadily at the rate of 44.97 per 100,000 population. However, there was no statistically significant difference in the incidence of sepsis across the various census regions. The average length of hospital stay was 8.84 days in the Northeast, 7.28 days in the Midwest, 7.96 days in the South and 7.3 days in the West. The average length of stay was 1.56 days (95% CI 1.22-1.90; p<0.05) shorter in the Midwest compared to the Northeast. There was no difference in the length of hospital stay between the Midwest and the West. The average mortality was 15.32% in the Northeast, 11.57% in the Midwest, 13.44% in the South and 12.24% in the West. The mortality was 3.75% (95% CI 2.42-5.07; p<0.05) lower in the Midwest compared to the Northeast. There was no difference in mortality between the Midwest and the West. The average cost of hospitalization was $19,365 in the Northeast, $17,077 in the Midwest, $16,583 in the South and $22,484 in the West. The cost of hospitalization in the Midwest was $5,406 (95% CI 4069-6743; p<0.05) lower compared to the West and $2,287 (95% CI 339-4236; p<0.05) lower compared to the Northeast. There was no difference in the cost of hospitalization between the Midwest and the South.

Conclusion: Hospitals in the Midwest and the West have lower in-patient mortality and shorter length of hospital stay compared to the Northeast and the South. However, the cost of hospitalization is higher in the West compared to other regions. The reason for these regional differences is unclear. Further studies to identify for contributing factors might help improve hospitalization outcomes throughout the United States.
Title: Impella RP as a Bridge for Right Ventricular Support in Patients with Massive and Submassive Pulmonary Embolism.

Authors: Marvin Kajy MD, Nimrod Blank MD, Amir Kaki MD, Cindy Grines MD, Mahir Elder MD, Tamam Mohamad MD, Theodore Schreiber MD, FACC

Introduction: Massive and submassive pulmonary emboli (PE) are the most common causes of acute right ventricular failure[1]. Ultrasound assisted catheter directed thrombolysis (UACDT) is an established treatment for massive & submassive PE[2]. Despite UACDT, some patients remain hemodynamically impaired.

The Impella RP is a temporary right ventricular assist device that supplies mechanical circulatory support to the right heart. The use of this device is indicated in right ventricular failure resulting in cardiogenic shock[3]. In the setting of an acute PE, the right ventricle is subjected to abnormal and increased loading that varies in timing, magnitude, and duration[4]. Consequently, RV dysfunction ensues leading to hemodynamic instability. By providing mechanical support to the heart (via the Impella RP) during the thrombolysis process, the damage to the right ventricle may be minimal or even reversible.

Methods: We present a series of five consecutive patients with massive PE who sustained cardiogenic shock refractory to volume expansion, inotropic support and UACDT. These patients were assessed with right heart catheterization and treated with Impella RP. Heart catheterization parameters and echocardiogram of the heart were obtained before and after Impella RP deployment.

Results: At presentation, all patients had a low cardiac index (mean 1.69 l/min/m²) without left heart abnormalities on echocardiography. An echocardiogram for all five patients prior to Impella RP insertion was remarkable for right ventricular severe hypokinesis to akinesis and severe right ventricular dilation. Left ventricular ejection fraction in the cohort ranged from 50-80%.

The Impella RP mechanical circulatory device was used for an average of 3.2 days (range of 1-6 days). During Impella RP treatment, all patients improved clinically and hemodynamically. To illustrate, the average baseline cardiac index at presentation was 1.69/min/m². This improved to 2.5 l/min/m² and 3.1 l/min/m² at 24 hours and 48 hours of treatment, respectively. Furthermore, the average systolic blood pressure increased and average heart rate decreased. Echocardiography at index day and follow up after three to four days, demonstrated improvement in right ventricular basal diameter, tricuspid annular plane systolic excursion and fractional area change (parameters of global right ventricular function). No significant changes were found in renal function, hemoglobin and platelets level during device use. None of the patients required a blood transfusion, although one patient treated with Impella RP for 6 days experienced a hemoglobin drop from 13.7 to 7.3 gm/dl. All patients survived to discharge.

Conclusion: Acute PE patients may present or deteriorate into hemodynamic instability and cardiogenic shock despite optimal management. Our study suggests a possible utility of Impella RP as a bridge to recovery for such patients because of the immediate clinical improvement noted after device placement and a low rate of adverse events. Further research is needed to establish whether the combination of Impella RP and UACDT can be adopted widely.

References


Title: Assessing the Rate of Providing Anti-Coagulation Management Recommendations Post-Endoscopy: a quality improvement initiative

Authors: Vivek Mendiratta, MD; Christopher Fernandez, MD; Mohammad Elbatta, MD; Salwa Hussain, MD

Introduction: Over 20 million esophagogastroduodenoscopies (EGD) and colonoscopies were performed last year. Many of the patients undergoing these procedures receive anti-thrombotic therapy for a variety of clinical conditions which increase the risk of blood clots, potentially leading to strokes, coronary events, deep venous thrombosis (DVT), and pulmonary embolism (PE). Major gastroenterology societies publish guidelines periodically regarding appropriate pre-endoscopy anti-coagulation management so that patients can be provided with explicit, evidence-based, pre-procedure instructions. However, our clinical observations suggest inadequate physician to patient communication in regards to anti-coagulation recommendations post-endoscopy. Thus, this quality improvement project was devised to assess the actual rates of written communication of anti-coagulation recommendations post-EGD or colonoscopy and the possible adverse effects of inadequate communication.

Methods: After IRB approval, a retrospective HIPAA compliant database search of medical records was performed from 1/2014-7/2017. Medical record review of the first 1000 patients with atrial fibrillation or a hypercoagulable state (i.e. Protein C and S deficiency, Factor V Leiden mutation, anti-phospholipid antibody, recurrent PE or DVT) was performed. 100 patients met inclusion criteria, which included active anticoagulation/antiplatelet therapy (except aspirin) at the time of an EGD or colonoscopy. Charts were then reviewed to assess for documentation of post-procedure anti-coagulation management recommendations, as well as complications related to inappropriate management of anticoagulation.

Results: Of the 100 patients (43 female, 57 male) meeting inclusion criteria, 44% underwent biopsy and/or polypectomy during endoscopy. 14% of patients (8 males, 6 females) received explicit, written post-procedure instructions on how to manage their anti-coagulation therapy post-endoscopy. None of these patients had adverse events occur post-procedure. 86% of patients were not given post-procedure anti-coagulation instructions, of which 3 adverse events were seen, including a TIA, stroke, and GI bleed within 90 days of their procedure.

Conclusion: Our Results: indicate poor physician to patient written communication regarding anti-coagulation recommendations post-EGD or colonoscopy. While these procedures are typically low risk for bleeding events, certain interventions (such as biopsy or polyp removal) can portend a higher risk of adverse events, thus requiring the need to temporarily hold anticoagulation therapy pre-procedure. Many of these patients carry a high risk of thrombosis and thus poor post-procedure communication regarding timing of anti-coagulation resumption may have serious and potentially life-threatening consequences. As a result of this data, a medical record best practice advisory or reminder will be implemented as a quality improvement initiative to improve physician compliance with providing written recommendations on anti-coagulation management post-endoscopy.
Title: Neutrophil-to-Lymphocyte Ratio and Platelet-to-Lymphocyte Ratio as Predictive Markers for Deep Vein Thrombosis

Authors: Jason Mouabbi, MD; Susan Szpunar, PhD; Louis Saravolatz, MD; Zyad Kafri, MD; Tarik Hadid, MD

Introduction: The neutrophil to lymphocyte ratio (NLR) and platelet to lymphocyte ratio (PLR) are known indicators of systemic inflammation. Thrombotic disorders are pro-inflammatory in nature. NLR and PLR have not yet been established as predictive markers of deep vein thrombosis (DVT).

Methods: We conducted a retrospective chart review of patients who presented with lower extremity swelling at St. John Hospital and Medical Center from 1/2010 to 12/2014. Patients with a diagnosis of DVT confirmed via ultrasound Doppler (DVT group) were compared to patients with a negative Doppler (Control group). The NLR and PLR were calculated based on a complete blood count (CBC) done on the same day of the Doppler study. Values of NLR ≥ 3.4 were considered positive; values of PLR ≥ 260 were considered positive. For comparison, D-dimer was also assessed with values ≥ 500ng/dl considered positive. We assessed the sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) of NLR, PLR, positive on both NLR and PLR and D-dimer. All data were analyzed using SPSS v. 24.0.

Results: We reviewed 708 charts; 51 charts met the inclusion criteria for the DVT group and 51 charts were randomly selected for the controls. There was no difference in mean age between the DVT and the Control group (60.5 ± 20.4 years vs. 60.4 ± 18.7 years, respectively). There were also no differences between groups by sex or race. NLR showed a sensitivity of 90% (NPV of 89%) whereas PLR showed a specificity of 98% (PPV 97%). D-Dimer showed a sensitivity of 88% (NPV 75%). When we look at NLR and PLR in parallel when both are positive the sensitivity for the presence of DVT is 88%, specificity 100%, PPV 100% and NPV 90%.

Conclusion: Both NLR and PLR are better predictors of the presence or absence of DVT compared to D-Dimer. NLR can be useful to rule-out DVT when it is negative; whereas PLR can be useful in ruling-in DVT when it is positive. Furthermore, a positive NLR and positive PLR yielded the best prognostic value for predicting DVT. NLR and PLR ratios offer a new powerful, affordable, simple and readily available tool in the hands of clinicians to help them in the diagnosis of DVT.
Michigan-Research-Poster Finalist  
Alicja Sobilo, MD

Title: Inappropriate Use of IV Anti-Hypertensives

Authors: Alicja Sobilo, MD - Wayne State University, Internal Medicine PGY-3, Jacob Salman, MD - Wayne State University, Internal Medicine PGY-2, Daymon Peterson, MD - Wayne State University, Internal Medicine PGY-1

Introduction: There are no clear guidelines for the proper use of IV anti-hypertensives (IVAHs) in the hospital setting for the treatment of asymptomatic hypertension. Studies have shown that aggressive treatment can lead to hypotension, acute kidney injury, and other complications. Ambiguity of when to use IVAHs prompted two PDSA cycles which were focused on education. Initial goal was to reduce the use of IVAHs by at least 20%. The intervention of educating physicians, nurses and pharmacy staff decreased the use of IVAHs in the setting of non-emergent hypertension by much more than the goal and as a direct result also decreased patient complications and absolute cost.

Methods: Over a three-month period, a retrospective chart review of 134 hospitalized patients who received IV Labetalol, IV Enalapril, and IV Hydralazine was done. Exclusion criteria included hypertensive emergency, stroke and NPO patients. Data showed 34% complications with IV Enalapril, 23% with IV Hydralazine and 35% with IV Labetalol. In all of these cases an IVAH was not strictly indicated and over 80% of patients had oral options available. Intervention included two rounds of education to physicians, nurses and pharmacy.

Results: Education focused on hypertension guidelines had significant impact. Total dispensed doses of IV Enalapril decreased from 1,857 to 1,282 (31% reduction) and total cost decreased from $8,414.62 to $6,929.10 (18% reduction). IV Hydralazine doses decreased from 663 to 250 (62% reduction) and total cost decreased from $10,266.48 to $3,810.24 (63% reduction). IV Labetalol use decreased from 411 doses to 287 (30% reduction) and total cost decreased from $4,503.69 to $2,921.18 (35% reduction). In a 3-month interval, doses of IVAHs was reduced by 1,112 and total cost by $9,524.27.

Conclusion: Currently, data is being collected for the next PDSA cycle for which an EHR pop-up questioning the indication for IVAH was implemented. Promising Results: are so far encouraging and show further decrease in inappropriate IVAH use; ultimately decreasing patient complications and secondarily, absolute cost.

References

Title: The Effect of the Selective Inhibitor of Nuclear Exports, Selinexor (KPT-330), on T-cell Non-Hodgkin Lymphoma

Authors: Jithma P. Abeykoon1, Mary J. Stenson2, Jonas Paludo2, Joshua M. Lawson2, Niraj K. Shenoy2, Xiaosheng Wu2, Thomas E. Wizig2; 1Department of Internal Medicine, 2Division of Hematology, Mayo Clinic, Rochester, MN, USA

Introduction: Communication and molecular trafficking between the nucleus and the cytoplasm are vital to cells survival and have an additional demand in rapidly dividing cancer cells including lymphoma. Chromosome region maintenance 1(CRM1) mediated nuclear export, is a major pathway responsible for the nuclear efflux of proteins. KPT-330 is a novel drug that covalently binds to and inhibits CRM1 thereby disabling a key nuclear export pathway used by the tumor cell. KPT-330 is in clinical trials for B-cell malignancies; however, pre-clinical data on T-cell lymphoma (TCL) and mantle cell lymphoma (MCL) are limited. To provide rationale for future trials, we studied KPT-330 on aggressive non-Hodgkin lymphoma cell-lines with a special emphasis on TCL.

Methods: Representative cell-lines of TCL (SU-DHL-1, Karpas299, SR-786), MCL (Joko, Mino, JVM-2) and DLBCL (LY-1, SU-DHL-2) were treated with various concentrations (0.1uM to 10 uM) of KPT-330 to assess viability, proliferative potential and cell-cycle arrest. Cell viability was evaluated by an apoptosis assay using Annexin V and propidium iodide (PI) after KPT-330 treatment for 48h using flow cytometry. 3[H] labeled thymidine labeling was used to assess cell proliferative potential in the presence of various concentrations (0.1 uM to 0.5 uM) of KPT-330 after 48h treatment. For cell-cycle analysis, Jeko cell line was incubated with 0.5 uM, and 1.0 uM concentrations of KPT-330 for 24h and subsequently underwent analysis using flow cytometry. Proliferation assay was also used to evaluate the synergistic effect of bortezomib and gemcitabine when added to KPT-330, separately. Synergy in drug combination was analyzed by CalcuSyn and the combination index <1 was considered to be synergistic.

Results:

Cell apoptosis

The mean viability of the control cell-lines was 91% in TCL and MCL cell-lines and decreased to 53%, (p=0.03) and 66%, (p=0.04), respectively when treated with 1.0 uM KPT-330 concentration (lowest concentration where a statistically significant difference was observed). No such effect was seen in diffuse large B-cell lymphoma (DLBCL), when treated with the highest KPT-330 concentration (10 uM).

Cell Proliferation, Cell-cycle and Synergy

In TCL and MCL cell-lines, the mean thymidine labeling decreased from 276,385 cpm to 50,699 cpm, (p<0.0001) and 525,448 cpm to 433,499 cpm, (p=0.03) at 0.1uM KPT-330, respectively and no such significant response was observed in DLBCL cell-lines at the highest concentration of KPT-330. Cell-cycle analysis showed a G0-G1 arrest when cell line was treated with KPT-330. Synergistic effect was seen in TCL and MCL when treated with gemcitabine or bortezomib combined with KPT-330 in concentrations ≤0.5 uM.

Conclusion: KPT-330 imposes pro-apoptotic and anti-proliferative activity in TCL and MCL at concentrations ≤ 1uM; a concentration (< 35-40 mg/m²), shown in previous clinical trials to have a good
safety profile.\textsuperscript{1,2} KPT-330 deserves further clinical investigation to assess its potential in treating TCL and MCL.

References


Title: A Novel Volumetric Laser Endomicroscopy Computer Algorithm for Landmark Identification and Delineation of Barrett’s Esophagus Dysplasia

Authors: Amrit K. Kamboj, MD1; Liam Zakko, MD2; Kavel Visrodia, MD2, Daniel K. Chan, MD2; Lori S. Lutzke, CCRP2; Kenneth K. Wang, MD2; Cadman L. Leggett, MD2; 1Department of Internal Medicine, Rochester, MN, 2Division of Gastroenterology and Hepatology, Rochester, MN

Introduction: Volumetric laser endomicroscopy (VLE) is an emerging technology used for detection of Barrett’s esophagus (BE) dysplasia. Thorough review of VLE data can be challenging due to the subtle image characteristics and the large volume of data to interpret (1200 cross-sectional frames). We hypothesized that a whole scan en-face VLE view, where the segmentation of clinically relevant tissue structures is conveniently visualized, would aid in recognition of anatomical landmarks. The aim of this study was to generate an automated computer aided diagnostic (CAD) algorithm that provides an en-face VLE view with superimposed features of interest.

Methods: The first step in the proposed computer algorithm is to generate an en-face image of a full VLE scan. This is achieved by digitally ‘unwrapping’ the circumferential scan (tubular esophagus) and presenting the data from the top down (filleted and flattened esophagus), i.e. enabling visualization of the entire esophageal surface in one image. The second step is to overlay automatically segmented VLE features onto the en-face view. A feature that distinguishes normal squamous epithelium from BE is the presence of a layered architecture. Two other features of interest include the identification of high scattering tissue and glands, as these have some association with BE dysplasia. The algorithm first segments the tissue surface and measures the signal intensity decay at every imaging point (A-line) in a cross-section and displays a graded color map onto the en-face view. The algorithm then performs segmentation of areas that contain glandular structures. The final output is an en-face view of the entire VLE scan with superimposed color graded information on layered architecture, signal intensity and glandular structures.

Results: The CAD algorithm was applied to 10 VLE scans obtained from a U.S. multicenter VLE registry. The gastroesophageal junction (GEJ) was identified by the en-face visualization of the top of the gastric folds. The squamocolumnar junction was identified by the transition of high signal intensity (associated with gastric epithelium) to lower signal intensity (associated with squamous epithelium). Both anatomical landmarks were easily identified on en-face imaging. BE showed lack of a layered architecture with glandular structures that extended beyond the GEJ. A VLE scan contained an area concerning for BE dysplasia delineated by VLE laser markings. Laser markings were clearly visible on en-face imaging and delineated a region with higher surface signal intensity compared to surrounding BE.

Conclusion: The proposed computer algorithm can potentially simplify VLE image interpretation by providing a comprehensive en-face view with superimposed color graded imaging features. Further validation of this algorithm is needed to determine its diagnostic performance for detection of BE dysplasia.
Minnesota-Research-Poster Finalist
Nasreen Syeda Quadri, MD

Title: Elevated Blood Glucose Prevalence in Newly Arrived Refugees

Authors: Nasreen Quadri, MD - University of Minnesota, Departments of Internal Medicine and Pediatrics
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Introduction: The prevalence of diabetes in newly arrived refugee patients varies according to country of origin, age and BMI. Data currently suggests pre-departure prevalence of diabetes in refugee populations is roughly 2-3% and typically diagnosed in symptomatic individuals. Other studies of diabetes prevalence in this population has been estimated between 3-8%. The prevalence is important in recognizing future risk for cardiovascular disease and complications of diabetes. This risk increases over time as lifestyle changes after US arrival as early as one to five years after resettlement. The objective of this study is to describe the prevalence of elevated glucose in newly arrived refugee patients and correlate associations with demographic data.

Methods: This was a retrospective chart review of the electronic medical record of 2,332 refugee patients from May 2009 to February 2016 looking at markers of elevated glucose (random glucose from basic metabolic panel, hemoglobin A1c) and demographic information (age, sex, language as proxy for country of origin, BMI).

Results: Roughly half of the patients in this study were male (52%). Of the 2,332 newly arrived refugee patients undergoing new arrival screening, 1.8% had elevated glucose defined as glucose > 199 mg/dL. The average glucose in the elevated group was 313 mg/dL and the average hemoglobin A1c was 9.4%. About 95% with elevated glucose were under the age of 65. When compared to other refugee populations in the study, Karen/Burmese speakers had a lower prevalence of elevated glucose (1.1%, p = 0.01) and Bhutanese Nepali speakers had a higher prevalence (4.1%, p = 0.005). Upon comparing BMI categories with elevated glucose prevalence, there is a statistically significant relationship with higher BMI and elevated glucose prevalence (3.7% vs 1.0%, p < 0.001).

Conclusion: Refugee patients should be viewed as a subset of immigrant patients based on their common forced migration as a social determinant of health impacting their access to preventive health care obtained prior to resettlement in the United States. Though the prevalence in the this study population (1.8%) was lower than other estimates (2.3%), it was in accordance with the varied prevalence from 2.5% to 14% based on country of origin as demonstrated in other studies. There is more discussion to be had regarding the appropriate screening test (fasting plasma glucose, random blood glucose, HbA1c) to identify disease burden in a cost-effective manner on a large scale. These data suggest universal blood glucose screening of new arrival refugees greater than age 40 and those within BMI categories of overweight and obese ranges may be advisable as part of the domestic medical examination to diagnose and treat diabetes to prevent morbidity and mortality. Asian refugees should be screened at lower BMI levels (overweight BMI 23-27.4, obese BMI >27.4) given higher prevalence at relatively lower BMI cut points.

References

**Minnesota-Research-Poster Finalist**  
**Nicholas A Zorko, MD,PhD**

**Title:** University of Minnesota Resident Primary Care Continuity Clinic Prompt Notification of Patient Results: Strategies for Improvement after Adaption of the 4+4 Continuity Clinic Schedule

**Authors:** Nicholas Zorko, MD, PhD; Susan Lou, MD; Demitri Andrisani, MD; Jessica Boarini, MD; James Campbell, MD; Ruben Crespo-Diaz, MD, PhD; Matthew Matsunaga, MD; Daniel Murphy, MD; Sasha Prisco, MD, PhD; Sravanti Rangaraju, MD; Katarina Wrzos, MD; Alisa Duran, MD; Heather Thompson, MD; Charles Moldow, MD; Briar Duffy, MD; Tanya Melnik, MD

**Introduction:** Communication is key to the doctor-patient relationship and impacts patient safety and satisfaction. Studies have demonstrated no notification in 75% of normal and ~1/3 of abnormal results. Similar issues were identified in the Internal Medicine resident clinic with the transition from a traditional half-day of clinic weekly to a full day weekly for 4 weeks followed by inpatient only duties for the subsequent 4 weeks. A new method for improving the timeliness and completeness of result reporting was needed given the changes brought about due to the new clinic structure.

**Methods:** Our goal was to improve total results with documented patient notification in Epic and time to patient notification from final result. Each resident encounter on specific date was assigned a number in a linear fashion. Records from each resident clinic date within a PDSA cycle were then queried at random using a web-based random number generator. Records without a lab result requiring follow-up were excluded from analysis. Time of final result, presence of a result note, and time of result note were compiled using pre-intervention, pooled, and paired resident result systems over the course of 3 PDSA cycles with multiple clinic dates included in each PDSA cycle.

**Results:** Mean time to result note decreased from 9.78 days pre-intervention to 0.98 days with paired intervention strategy ($p<0.0001$, 1-way ANOVA). Percent of results with notes increased from a mean of 50% in the pre-intervention group to 100% in the paired intervention group ($p<0.0001$, 1-way ANOVA).

**Conclusion:** This study showed significant increase in numbers of results with notes and decreased time to result note when comparing pre-intervention to paired result reporting strategies. Modifications in result reporting had the desired results, which may lead to improved patient satisfaction and safety.
Title: Autologous bone marrow transplantation for multiple myeloma: an analysis of risk factors that affected patient outcomes at the University of Mississippi Medical Center

Authors: Hansen DK, Williamson T, Hsu J, Buck T and Milner CP

Introduction: Objectives: To examine prognostic factors that affected Mississippian outcomes in autologous hematopoietic stem cell transplantation (HSCT) for multiple myeloma (MM) as opposed to historical cohorts. To implement institutional changes for MM patients undergoing HSCT at the University of Mississippi Medical Center (UMMC) based on our study results.

Methods: This retrospective chart review included 128 patients with MM who received autologous HSCT at UMMC from 2009 to 2014. IRB approval was obtained. Nine parameters were evaluated including age, sex, ethnicity, time of diagnosis to transplantation, disease status at the time of transplantation and at day + 100, Durie-Salmon stage at diagnosis, type of induction chemotherapy received, Karnofsky performance status, cytogenetics and post-transplantation maintenance therapy.

Results: Mean age of our patients was 57 with the majority being African American (54%) and female (52%). The majority of patients had IgG kappa disease (41%), with stage 3A, and a Karnofsky score > 80%. Induction chemotherapy received was either revlimid/dexamethasone (RD 27%) or revlimid/velcade/dexamethasone (RVD 20%). 60% of patients underwent maintenance therapy after HSCT while 33% were observed. Disease status at HSCT was partial response (40%), very good partial response (23%) or complete response (20%). 37% of patients had a complete response at day 100 following HSCT. Most patients underwent HSCT 6 months from initial diagnosis. Mean time to progression was approximately 24 months. There were statistical differences noted for higher Karnofsky scores (p=0.004), disease status at transplant (p<0.01), disease status at day 100 (p<0.01) with improved progression free survival (PFS) in patients with a complete response. There was a trend toward improved overall survival (OS) with earlier HSCT (p=0.38), earlier stage disease (p=0.14) and low-risk cytogenetics (p=0.09) but our study was underpowered to definitively show these differences.

Conclusion: MM remains an incurable malignancy, however, there are several prognostic factors that can affect patient survival. Regarding practice changes at UMMC, we believe that our study as per national guidelines, would advise using a three drug induction chemotherapy regimen such as RVD rather than RD, earlier transplantation and standard maintenance chemotherapy for all patients following HSCT. Furthermore, we need to ensure that cytogenetics are obtained in all MM patients as they are prognostic of survival benefit. While a few areas of improvement are needed and currently in
the process of being implemented, our institution appears to be at par with national standards regarding disease PFS despite healthcare disparity in the state of Mississippi.
Title: Let sleeping bugs lie: Assessing inappropriate treatment of asymptomatic bacteriuria amongst residents.

Authors: John O. Kolawole MD, Nancy Harrison MD, Nikhil Patel MD, Sarika Bedi MD, Frankie Pedigo MD and Calvin Thigpen MD.

Introduction: Urinary tract infections are a common medical problem encountered in the inpatient and outpatient settings. However, overtreatment of asymptomatic bacteriuria frequently occurs and is contributing to antibiotic overuse and resistance patterns. Typical symptoms of UTI, including both cystitis and pyelonephritis include: dysuria, frequency, urgency, suprapubic pain, hematuria, fever (>38ºc), chills, flank pain, costovertebral angle tenderness and/or nausea/vomiting. This project identifies patients with urinary symptoms and appropriate documentation of symptoms as well appropriate treatment.

Methods: We reviewed patients on Internal Medicine Resident teams at the University of Mississippi Medical Center from December 2016 to May 2017. On March 1, 2017 education was provided to residents about appropriate guidelines for diagnosing and treating a urinary tract infection versus asymptomatic bacteria. We compared data prior to intervention and after intervention to assess if patient were appropriately being treated for UTIs. Data gathered included urine culture results, associated urinary symptoms, antibiotic choice and duration.

Results: Over a 6 month period 152 patients were treated for UTI. Of those patients 26 patients had indwelling Foley catheters and were excluded from the project. 54 patients had altered mental status and/or fit criteria for Systemic Inflammatory Response (SIRS) and were treated broadly with antibiotics for sepsis rule out. These patients were considered positive for symptomatic bacteriuria. Prior to intervention 8 out of 66 patients (12%) were inappropriately treated for asymptomatic bacteriuria. After intervention 3 out of 60 patients (5%) were inappropriately treated for asymptomatic bacteriuria (p value 0.21 by Two-tailed Fischer Exact Probability Test).

Conclusion: Educating clinicians about overtreatment of asymptomatic bacteriuria is important to help reduce antibiotic resistance and potential adverse effects of antibiotics on otherwise asymptomatic patients. Although, our intervention did not show statistically significant results, ongoing education about appropriate treatment of asymptomatic bacteria will improve patient care. As shown in the results, we are proficient in identifying patients with UTIs versus asymptomatic bacteriuria. This study was conducted in the inpatient setting where many variables such as concurrent infections and sepsis rule out complicated the overall outcome. If we were to replicate this study in the outpatient setting the results will likely show a higher percentage of patients being over treated for asymptomatic bacteriuria. However, further studies are necessary to confirm this hypothesis.
Title: Long lasting androgen-induced cardiometabolic effects in a model of Polycystic Ovary Syndrome

Authors: Edgar Torres-Fernandez, Kristen Adams, Maryam Syed, Damian G. Romero and Licy L. Yanes Cardozo

Introduction: Polycystic Ovary Syndrome (PCOS), the commonest endocrine disorder in young women, is characterized by hyperandrogenemia and ovarian dysfunction. We recently showed that exposure to the androgen dihydrotestosterone (DHT) causes obesity, hypertension and activation of renin angiotensin system (RAS) in a model of PCOS. We tested the hypothesis that androgen withdrawal reverses the cardiometabolic effects previously triggered by hyperandrogenemia in PCOS.

Methods: Four week-old female Sprague Dawley rats were randomized to DHT (83 mcg daily) or placebo (n=10/grp). After 6 months, DHT administration were discontinued (Ex-DHT). During additional 6 months, food intake, body weight, body composition (Echo-MRI) and insulin resistance were analyzed. At 12 months mean arterial pressure (MAP), baseline and after enalapril administration was measured by radiotelemetry. At the end of the study, insulin, leptin, adiponectin, plasma androgens and estradiol, RNA expression of renal and adipose tissue of RAS components and androgen receptor were determined.

Results: After 6 months of DHT withdrawal, body weight (330±9 vs. 265±4 g, p<0.001), fat mass (44.60±4.1 vs. 26.6±1.5 g, p<0.001), lean mass and food intake were significantly higher in Ex-DHT rats. Fasting plasma insulin, leptin, adiponectin, insulin resistance and MAP (122±1 vs. 110±1 mmHg, p<0.001) were increased in Ex-DHT rats. Plasma DHT and testosterone were decreased in Ex-DHT and no significantly changes were observed in estradiol levels. Enalapril normalized MAP in ex-DHT. Renal injury, glomerular sclerosis, renal RNA expression of RAS components were significantly higher in Ex-DHT. Androgen receptor RNA expression in renal medulla and visceral/subcutaneous adipose tissue were higher in Ex-DHT.

Conclusion: Despite normalization of hyperandrogenemia, Ex-DHT animals exhibited long lasting negative cardiometabolic effects triggered by previous DHT exposure. Moreover, persistent renal RAS and renal/adipose androgen receptor activation after DHT withdrawal may play a major role in PCOS hypertension. Excess androgen-mediated cardiometabolic effects may be irreversible and therapies to overcome target-organ deleterious effects should be used even after normalization of hyperandrogenemia.
Title: Monitoring the Use of Telemonitor: Evaluation of Adherence to Choosing Wisely® Campaign

Authors: T. Maduke, MD, MPH (Associate), K. Gandhi, MD (Associate), Y. Goite, MD (Associate), S. Dhital, MD (Associate), B. Qureshi, MD (Associate), F. Bofarrag, MD (Associate), S. Khan, MD (Associate), S. Basnyat, MD, MPH (Associate), L. Liu, MD (Associate), M. Suazo-Martinez MD (Associate) and H. Kawsar, MD, Ph.D (Fellow)

Introduction: In 2004, American Heart Association (AHA) released guidelines on use of telemonitor, and divided patients into 3 classes: class I (telemonitor use indicated), class II (telemonitor may be beneficial) and class III (telemonitor not indicated). Studies have shown that telemonitor is often used inappropriately, and can negatively impact patient care. As a result, The Choosing Wisely® campaign has adopted this guideline to improve appropriate use of telemonitor in patients outside of ICU.

Methods: We conducted a retrospective study of patients who were admitted between January and March 2017 and were on telemonitor. We documented the indication(s) and duration of telemonitor use, event(s) recorded in telemonitor and outcome of the event(s). The data was analyzed using appropriate statistical tools.

Results: Among 329 qualified patients, 162 were in teaching service and 167 in non-teaching service. Mean age was 75 years and 52% patients were male. Mean duration of telemonitor use was 4.4 days on the teaching service and 3.6 days on the non-teaching service. On the teaching service, 62% had class I, 25% had class II, and 13% had class III indications. In non-teaching service, 47% had class I, 38% had class II, and 15% had class III indications. Syncope, altered mental status, chest pain, atrial fibrillation, and heart failure were the most common indications. Ten events were recorded in teaching patients and 13 in non-teaching patients. None of the events occurred in patients with class III indications. Two patients died. Based on our data, we estimated that the hospital spends approximately $2,5 million per year by using telemonitor for class III (not indicated) patients.

Conclusion: Our results highlight the need to improve guideline-based utilization of telemonitor in hospitalized patients. An educational intervention is underway to achieve this goal. An educational intervention is underway to reduce telemonitor use for class III indication, and thereby reduce patient discomfort, false alarm and save money for the patients. Final post intervention results will be available and will be shown in the poster presentation at the time of the national ACP conference meeting.

References


Missouri-Research-Poster Finalist
Rachna Rawal, MD

Title: Stop. Think. Does My Patient Need Those Labs?: A Quality Improvement Study to Promote Cost-Conscious Care

Authors: ¹Rachna Rawal. ¹Oluwasayo Adeyemo. ²Ara Vartanyan. ¹Jennifer M. Schmidt.

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Introduction: Cost of healthcare in the United States is a challenge to hospitals and healthcare providers. Cost-conscious, high-value care education should begin early in residency training so that it becomes an integral component of residents’ future practice. Routine lab ordering for hospitalized patients is a well-known cause of high costs. This study’s goal was to decrease the number of labs ordered by residents on the general medicine service.

Methods: Subjects were Internal Medicine residents rotating through the inpatient Medicine service at a tertiary-care, academic medical center. Pre- and post-study surveys, created by the project team, assessed knowledge of lab costs, ordering practices and global perceptions of physicians’ role in healthcare cost. The number of BMPs, CMPs and CBCs (with and without differential) ordered by each team was obtained from the electronic medical record (EMR) during a control block and discrete 4-week cohorts. Educational interventions included presentations at noon-conference, posters in team rooms, weekly emails (lab cost information, high-value care literature) and monthly emails to on-service attending physicians to alert them of the project. After four months of educational interventions (phase I), data continued to be collected via EMR for an additional four months (phase II). Additionally, the order of lab tests in the Admission Order Set was changed during the first month of phase II.

Results: During phase I, the total number of labs/patient day decreased by 7%. Post-survey data showed 92% of residents think about whether labs are indicated for their patient, however, 50% continue to feel uncomfortable when their patients do not have morning labs. 100% of the residents think about the difference between CMP and BMP. Phase II showed a 0.9% decrease in labs per a patient day and increased most blocks 3-4%. Across both phases, the proportion of CBC without differential compared to CBC with differential increased from 10% to 25%. Free responses from residents’ surveys revealed reasons for over-ordering included: concern for missing something clinically relevant, and reprimands from attending physicians.

Conclusion: Our data shows that residents are thinking about lab costs, but these thoughts are not always reflected in their lab ordering practices. Phase II’s smaller decrease in labs ordered suggests the importance of regular educational interventions in maintaining a cost-conscious culture. Culture change must occur at all levels of the team as evidenced by residents’ identification of attending physicians as barriers. These results have framed Phase III that includes intern and senior resident orientation sessions in addition to new educational interventions focused on case-based high-value care education. We believe that we are starting to create a cultural shift based on resident surveys and anecdotes. We anticipate a continued shift towards cost-conscious care in our residency program.
New Jersey-Research-Poster Finalist
Aayah Fatayerji

Title: Improving the Quality of Resident-Run Code Blues in a Community Teaching Hospital

Authors: Aayah Fatayerji, DO, MPH, Stephanie Vuong, DO, Vivek Singh, DO, Jaron Schaumberg, DO, MS, Abraham Lo, DO, Sameh Elias, MD

Introduction: Across most hospitals, a Code Blue is announced when an adult patient is in cardiopulmonary arrest and requires resuscitative efforts. Traditionally, hospital policy has enforced Code Blues to be led by Hospitalists and Intensivists. There has not been a policy to allow for senior residents to lead Code Blues at Hackensack Meridian Health - Palisades Medical Center (PMC). PMC is a 202-bed community teaching hospital where Residents in the Internal Medicine program work closely with the in-house Intensivist or Hospitalist present at all Code Blues. The incorporation of lectures, mock-code simulations, and direct experience has been used to improve knowledge of up-to-date guidelines, closed-loop communication, and overall confidence in the leader’s ability to run a code. The purpose of this study is to identify opportunities for improvement during a code to increase the quality and effectiveness of Code Blue management. The first stage of this study was to increase involvement of resident physicians by updating the hospital policy to facilitate resident-run codes. An anonymous survey was performed which identified self-reported deficiencies and overall lack of confidence when leading a Code Blue. A comprehensive curriculum was devised which consisted of monthly lectures, with pre- and post-tests, two mock Code Blue simulations, with post-code debriefings, and a Code Blue simulation exam where each individual resident was scored on a standardized grading system. Upon completion, resident confidence and knowledge will be re-examined, stratified, and statistically analyzed. A post-code debriefing tool will be used to review the events of the resuscitation efforts by the code teams. Data obtained will be presented to the ICU Committee to implement future improvements in code dynamics and policy updates. By interpreting retrospective and prospective data of code performance, the effectiveness of interventions will be studied in accordance with the “AHA-Get with the Guidelines-Resuscitation” recognition measures. While other studies have predominantly focused on nursing roles, the purpose of this project is to improve patient safety, hospital awareness, accountability for quality improvement, and confidence among resident physicians, with the goal of earning AHA-Gold Accreditation Status. We predict that over the course of twelve months under this curriculum, self-reported resident confidence and the ability to successfully run a Code Blue will improve. Ultimately, the results of this study will be used to increase quality and efficacy of Code Blues in our community teaching hospital and provide a framework to be reproduced at other residency programs.
New Jersey-Research-Poster Finalist
Nagham S Jafar

Title: Hypertriglyceridemia - Induced Pancreatitis Management Community-Based Teaching Hospital Experience

Authors: Nagham Leanne Jafar, Ola Adams, Nebeel Mesih, Daniel Goldsmith

Introduction: Severe hypertriglyceridemia (triglyceride >1000 mg/dl) occurs at a rate of 0.4% and is associated with a range of medical complications including severe pancreatitis and its complications like acute kidney injury. Hypertriglyceridemia (HTG) is the third most common cause of acute pancreatitis especially in levels above 1000 mg/dl. It has been suggested that insulin and plasmapheresis have a role in the management of acute HTGP, but there are no clear guidelines on the best treatment option in terms of better clinical outcomes. We present a retrospective study of patients who received active treatment in the form of insulin drip, plasmapheresis or a combination of both versus supportive treatment alone.

Methods: We reviewed medical records on patient’s age > 18 years, admitted with the primary diagnosis pancreatitis and had TG level above 1000 mg/dl between Jan 2014 and Oct 2017.

We gathered information on demographics, co-morbidities, incidence of pancreatitis associated with alcohol and type of treatment received. Outcomes were hospital length of stay (LOS) and recurrence of HTG induced pancreatitis (HTGIP).

Results: 89 Patients had a HTG level of 1000 mg/dl and above, and from that 36 had a diagnosis of acute pancreatitis. The Incidence of HTGIP among Patients with ETOH abuse is 41%. The mean age of presentation was 44.8; there was a predominance of men (88%) and Hispanic population (44.8%). 54.3% (19) were treated with supportive treatment (ST) and had a mean TG of 1614.79 mg/dl versus 45.7% (16) were given active treatment (AT) with a mean TG of 3066.19 mg/dl. Total LOS was less in AT group with a mean of 3.3 days versus 5.0 days in ST group (p=0.001). The days needed to achieve TG level below 500 was significantly less in the AT group (4.56 versus 1.56 in ST group) (p= 0.010). The 30 day readmission rate in ST group was 54.4% versus 8.33% in AT group (P= 0.016).

Conclusion: Hospital LOS in patients receiving active treatment was shorter than supportive treatment. They also have a lower readmission rate and faster achievement of TG lower than 500 mg/dl, suggesting more effective therapy in the AT group. We also observed high percentage of comorbid alcohol use with TG level above 1000 mg/dl.

We suggest starting active treatment in any patient with HTG- induced pancreatitis if TG level is equal or above 1000mg/dl, which may favor insulin drip in diabetes patients and plasmapheresis in non-diabetics. Further, we propose checking TG level in any diagnosis of acute pancreatitis even if alcohol abuse is present as HTG is frequently found.

References


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Title: Vedolizumab Induction May Prevent Celiac Enteritis in Patients with Celiac Disease

Authors: Vishal D. Patel, MD, Melissa Diener, MD, Barry Kaufman, MD, Theresa Stevens, APN

Introduction: Celiac disease (CD) is characterized as an autoimmune disorder where a protein known as gluten induces an immunological response in patients. The prevalence of CD is estimated up to 1% of the population worldwide. Small amounts of gluten can trigger hematological process in the intestines leading to diarrhea and abdominal pain and places significant stress on patients. Long-term complications include lymphoma, malabsorption, weight loss, anemia, vitamin deficiencies, osteoporosis, and other autoimmune diseases. Antigen receptors HLA DQ2 and DQ8 exhibit an exaggerated response in genetically susceptible patients. Tissue transglutaminase (TTG) is an enzyme that degrades gluten proteins and by-products then bind to these receptors with high affinity causing T-cells within the intestines to trigger cytokine production and induce inflammation. Transglutaminase also stimulates specific B-cell to produce anti-TTG immunoglobulins. The only treatment available is lifelong adherence to a strict gluten-free diet. Unfortunately there is significant cross-contamination of gluten in foods and medications that makes adhering to such diet difficult.

Methods: Vedolizumab is a monoclonal antibody developed for the treatment of Ulcerative Colitis and Crohn's disease. It binds to integrin α4β7 resulting anti-inflammatory activity selective to the gut. We hypothesize that vedolizumab induction may inhibit T-cell migration and therefore prevent immune mediated intestinal inflammation in patients with CD after a 2-week gluten challenge. All patients enrolled will have an established diagnosis of CD for greater than 6 months and an abnormal MARSH score on initial duodenal biopsy at time of diagnosis, positive celiac serologies including anti-TTG and anti-gliadin antibodies and positive HLA DQ2 and DQ8. At the time of enrollment, all patients will have negative celiac serologies and a MARSH score of 0 indicative of serologic and histologic remission. Patients will then be given three vedolizumab infusions two weeks apart. After the final infusion, patients will be given a gluten challenge consisting of 3g of gluten daily for 14 days. After completion of the gluten challenge, patients will undergo endoscopy with biopsy and repeat blood work consisting of TTG IgA, gliadin peptide IgA and IgG, endomysial antibody IgA and total immunoglobulin.

Results: The current study is ongoing with one patient enrolled. She has completed the full course of vedolizumab and completed a gluten challenge for 14-days. Patient reported no complaints after ingesting gluten. Follow up lab work confirmed serological remission and endoscopic biopsy confirmed histological remission.

Conclusion: Celiac disease has an estimated prevalence of up to 1% of the population worldwide with no pharmaceutical therapy. The only treatment available is adherence to a strict gluten-free diet. While this study is currently ongoing, it does suggest that vedolizumab has potential to be a treatment option for CD. With further research, patients with celiac disease may finally live without dietary restrictions.
Title: Effectiveness of Blood Transfusion Guidance Alert System in preventing inappropriate transfusions at a Community Hospital

Authors: Muhammad Sardar MD¹, Ananta Subedi MD¹, Muhammad Azharuddin MD¹, Prateek Ghatage MD¹, Doantrang Du MD¹, Arpad Szallasi MD², ¹Dept. of Internal Medicine, Monmouth Medical Center, 300, Second Avenue, Long Branch, New Jersey, 07740, ²Dept. of Pathology, Monmouth Medical Center, 300, Second Avenue, Long Branch, New Jersey, 07740

Introduction: Various strategies have been employed by hospitals to limit inappropriate blood transfusion practices including physician education programs and alerts in CPOE( computerized provider order entry system). A transfusion guidance alert system adopted in the electronic medical record system at our hospital has shown a decrease in the number of transfusion orders.¹ We aim to analyze the effectiveness of the alert system, not by comparing the number of transfusions before and after its adoption, but by seeing how frequently activation of an alert resulted in cancellation of the order. We identified the common causes of inappropriate transfusion and stratified them according to the ordering physician’s department.

Methods: A retrospective analysis of packed red blood cell transfusion orders that activated the alert (transfusion orders for hemoglobin above 7g/dL) was done from November 2016 to January 2017. Appropriateness of blood transfusion was determined using American Association of Blood Bank (AABB) 2016 guidelines by reviewing documented indication in the patient charts. Duplicate orders and orders with inconclusive indications were excluded.

Results: In a 3 month period, 138 packed red blood cell transfusion orders were filtered out by the hospital’s EMR system.100 orders were included in the final analysis after excluding orders that were duplicate (24) or with inconclusive indication (14). 44% (44/100) of the orders that were transfused above the cut off of 7g/dL could be justified using AABB 2016 guidelines. Of the 56 orders that were unjustified, 21.42 % (12/56) were cancelled by the physicians after seeing the alert. The top 3 causes of inappropriate blood transfusion were; pre-operative/post- operative patients with anemia- 31.7% (13/41); asymptomatic drop of hemoglobin with unclear etiology – 26.82% (11/41) and asymptomatic GI blood loss- 14.63% (6/41). Orthopedic department was most liberal in transfusing as 70% (7/10) of their transfusion orders that activated the alert system were not indicated, followed by department of internal medicine 44.44% (4/9) and anaesthesia 42.8% (15/35). A rough estimate showed that around $80000 worth of resources were spent on blood transfusions that were inappropriate during the time period of the study.

Conclusion: Physicians tend to ignore blood transfusion guidance alerts as a result of which we still have a very high percentage of inappropriate blood transfusions at our hospital. We propose that the ordering physician should be obligated to provide justification to the blood bank if the alert is activated before the packed red blood unit is released. We also encourage educating and reminding physicians about the latest guidelines.

References
New Jersey-Research-Poster Finalist
Ramy Sedhom, MD

Title: Does Section A of the NJ POLST Form Offer a Physicians a Greater Understanding of Patient Goals of Care in Advanced Illness

Authors: Ramy Sedhom MD, Daniel Sedhom MD, Justin Lebenthal MSIII, Daniel Schaer MD, David Barile MD

Introduction: The POLST “Practitioner Orders for Life-Sustaining Treatments” form addresses patient goals, scope of therapies, artificial nutrition, and resuscitation status. In New Jersey, Section A offers a unique opportunity for patients to describe in their own words overall goals of care. We investigated if completion of Section A influences overall care for patients with chronic illness.

Methods: All POLST forms completed in 2015 at the University Medical Center of Princeton were retrospectively reviewed. Patients 18 years or older who had their POLST form uploaded electronically by their physicians were eligible for inclusion. All other responses on the POLST form were collected. Chart review was used to collect admitting diagnosis, number of consultants used, admission to the intensive care unit, palliative care and hospice use, length of stay, number of readmissions and unplanned deaths in the hospital. Discharge summaries were reviewed to assess if advance care planning discussions were documented.

Results: 490 charts were reviewed: 54% were female, age 41 to 99 (mean 82). Section A was complete for 276 out of 490 patients (56%). Patients with section A completed more likely to choose limited or symptom treatment compared to full care (86% vs. 70%, p=<0.001, OR 0.3742, CI: 0.24 – 0.59). Patients with section A complete were also more likely to select a DNH (112 vs. 68, p= 0.32, OR 1.46, CI: 1.07 to 2.13), were more likely to choose no artificial nutrition (116 vs. 71, p=0.45, OR1.46, CI: 1.01 – 2.12). There was no statistical difference in choice of DNAR/DNAI status. In addition, patients who completed section A were less likely to be referred to the ICU, had a shorten length of stay (4 vs. 6 days), fewer readmissions, number of consultants participating in their care and unexpected deaths in the hospital (p<0.05). In addition, those completing section A were more likely to utilize both palliative and hospices services. Though physicians rarely documented code status on discharge summaries (38%), patients who completed section A were more likely to have advance care planning documented in discharge paper work.

Conclusion: Use of the POLST form provides an opportunity to discuss end-of-life care. Patients completing section A were more likely to utilize palliative care, hospice services, and have documented advance care planning. They experienced fewer days in the hospital and were less likely to be admitted to an intensive care unit. Completion of section A may lead to more conservative care, though qualitative studies are needed to further evaluate physician behaviors. The benefits of the POLST form extends beyond patient outcomes and may positively impact the care patients by allowing providers greater insight into the overall goal of individual patients. The importance of individualized preferences seems logical, but larger studies are needed to validate our findings.
New Mexico-Research-Poster Finalist
Nasim Pourtabatabaei, MD

Title: ZZZs Without Zolpidem: Cognitive Behavioral Therapy to Improve Sleep

Authors: Nasim Pourtabatabaei, MD, PGY2, University of New Mexico Department of Medicine, James Dean, DO, PGY3, University of New Mexico Department of Medicine, Carol Morales, MD, University of New Mexico Department of Medicine, Katherine Belon, PhD, New Mexico VA Health Care System, Aaron Joyce, PhD, New Mexico VA Health Care System, Michael Webster, PharmD, New Mexico VA Health Care System, Gregory Toney, PharmD, New Mexico VA Health Care System, Julie Foster, PharmD, New Mexico VA Health Care System, Linda Macdonald, MD, Staff Physician, New Mexico VA Health Care System, Associate Professor, University of New Mexico Department of Medicine

Introduction: Insomnia is a common medical complaint, affecting large numbers of individuals and resulting in numerous office visits. Zolpidem is approved by the FDA for treatment of short-term insomnia. A large proportion of NMVAHCS patients who are prescribed zolpidem are prescribed a nightly dose for a longer duration than recommended. The goals of this quality improvement project are to:

1. Reduce the number of patients who are taking zolpidem at doses higher and/or for a duration longer than recommended by the FDA;
2. Increase the number of patients who are using evidence-based psychological and behavioral treatments such as cognitive behavioral therapy for insomnia (CBT-I); and
3. Give physicians information about effective treatments for insomnia and strategies to help them talk to patients about the risks of zolpidem and alternatives to zolpidem.

Methods: PCPs were trained during three educational and role playing sessions to discuss the risks of chronic zolpidem use and the benefits of CBT-I with patients. Physicians completed a pre- and post-education questionnaire regarding their familiarity with CBT-I and its role in treating insomnia, comfort discussing CBT-I with patients, and plans to refer patients for CBT-I. Each PCP was given a list of his/her patients with a prescription for nightly zolpidem. Patients who agreed to participate in CBT-I were enrolled in an eight-week CBT-I class run by two behavioral psychologists. An internal medicine physician attended each class to monitor tapering of the zolpidem dose. Pre- and post-class zolpidem dose and insomnia severity score were measured.

Results: All resident physicians received all components of the education program. Compared to pre-education, the post-education data showed an improvement in score for all parameters measured. Forty-four patients met criteria for this intervention. PCPs discussed the risks of chronic zolpidem use and offered CBT-I to 28 patients (64%). Twenty-eight percent of the original cohort discontinued zolpidem use (twelve patients), 17% based solely on education by their PCP without CBT-I (seven patients). Of the 11 who enrolled, five completed the class and six dropped out. For the five patients who completed the CBT-I class, mean nightly zolpidem dose decreased from 10mg to 0mg and mean insomnia severity score dropped from 19.8 to 5.75 over 8 weeks.

Conclusion: Providing PCPs with information about the risks of chronic zolpidem use, effective non-pharmacologic treatments for insomnia, and strategies for having difficult conversations with patients in better care for patients with insomnia. Patients who attended the CBT-I class showed objective improvement in their insomnia, with a drop in mean insomnia severity score from 19.8 (moderately severe insomnia) at initiation to 5.75 (no clinically significant insomnia) at completion.
significant reduction in insomnia occurred in the setting of complete cessation of zolpidem. Efforts to increase the number of CBT-I enrollees are underway.
Nevada-Research-Poster Finalist  
Harmeet S Mashiana, MBBS

Title: Comparison of outcomes for supine versus prone position for ERCP: A systematic review and meta-analysis

Authors: Harmeet S Mashiana1; Mahendran Jayaraj1; Babu Pappu Mohan2; Syed R Shah1; Banreet Dhindsa1, Gordon Ohning1; Douglas G Adler3 1) Department of Internal Medicine, University of Nevada Las Vegas School of Medicine, Las Vegas, NV, 2) Department of Internal Medicine, University of Alabama, Tuscaloosa, AL, 3) Division of Gastroenterology, University of Utah School of Medicine, Salt Lake City, Utah

Introduction  Traditional textbook description of ERCP recommends doing routine ERCP in the prone position. Supine ERCP has been recommended for special situations like morbid obesity, the presence of abdominal wounds, drains etc. Many high-volume centers for ERCP have adopted doing ERCP in the supine position by default. Studies comparing the technical success and safety outcomes have shown variable results. We aimed at doing a systematic review and meta-analysis of studies reporting the comparison between the two positions for ERCP outcomes.

Methods: We conducted a comprehensive search of multiple electronic databases and conference proceedings including PubMed, EMBASE, and Web of Science databases (from inception through May 2017) to identify studies that reported the comparison of technical success and safety outcomes between supine and prone ERCP. The primary outcome was to estimate the pooled rates of technical success. The secondary outcome was to estimate the risks of complications, such as cardiopulmonary and post-ERCP pancreatitis (PEP).

Results: 6 studies reporting on 309 supine and 1415 prone ERCPs were identified. The pooled technical success rates for completion of ERCP in supine and prone positions were 89.1% (95%CI=80.9-94.0) and 95.6% (95%CI=91.5-97.7), respectively. The pooled rates for complications- Cardiopulmonary and PEP in supine position were 37.5% (95%CI=19.1-60.3) and 3.5% (95%CI=1.6-7.3), respectively. The pooled rates for complications- Cardiopulmonary and PEP in prone position were 41.0% (95%CI=20.9-64.8) and 3.9% (95%CI=2.4-6.4), respectively. The mean time required for completion of the procedure was 30 mins and 29.8 mins for supine and prone positions, respectively. Substantial heterogeneity noted in the analysis.

Conclusion: Prone ERCP’s have a higher technical success rate with a slightly lower mean duration and fewer adverse events. The decision regarding patient position should be made after evaluating the overall clinical scenario.
New York-Research-Poster Finalist
Aishwarya Bhardwaj, MD

Title: Guideline Directed Medical Therapy for Heart Failure Optimization in a Primary Care Setting

Authors: Aishwarya Bhardwaj, MD, Huy Phan, MD, Shane Varghese, MD, Smita Bakhai, MD, MPH

Introduction: At the Internal Medicine Clinic (IMC) in Erie County Medical Center (ECMC), roughly 15% of the patient population has a diagnosis of heart failure (HF). Due to lack of a consistent methodology for classifying HF in clinic electronic medical record (EMR), many of these patients are not on guideline directed medical therapy (GDMT) based on ACC/AHA/HFSA 2013 guidelines. This, in turn, has a significant negative impact on patients’ quality of life (QOL), increased mortality and number of avoidable hospitalizations, which pose a huge burden on limited hospital and national healthcare resources. The aims of this Quality Improvement (QI) project are to appropriately specify type of cardiac dysfunction based on ejection fraction (EF) and increase the use of GDMT by 10% from baseline within 12 months in patients aged 40-75 years with a diagnosis of HF at ECMC IMC.

Methods: To design this QI project, we employed Institute of Medicine’s STEEEP model (Safe, Timely, Effective, Efficient, Equitable, Patient-Centered). We used root cause-analysis to identify system-, provider-, and patient-based barriers in providing optimal GDMT to our patients with HF. In order to implement various interventions and overcome barriers toward improvement of care, we used the Plan Do Study Act (PDSA) model. Furthermore, we identified a family of measures, as adopted from the Institute for Healthcare Improvement, namely, outcome, process, and balance measures. Data analysis is performed using monthly run charts.

Results: Initial chart review revealed that less than 5% of eligible clinic patients with a diagnosis of HF were explicitly classified based on their EF and were on appropriate GDMT based on type of cardiac dysfunction. In September 2016, 26% of patients with HF were classified based on the type of cardiac dysfunction. After two PDSA cycles and 6 months into initiation of the project, in February 2017, 78% of patients were appropriately classified.

Conclusion: So far, two PDSA cycles have been implemented comprising 1) resident education reviewing ACC/AHA/HFSA 2013 guidelines 2) collaborating with the clinic Information Technology department to create a database for HF classification and to keep track of ECHO reports. Next PDSA cycle will focus on creation of pamphlets which will serve as reminders for providers and patients for optimization of HF therapy.

In the last 6 months, there has been a clear trend toward rise in appropriate HF classification leading to guideline directed treatment of our clinic patients. In the next 6 months, our overarching goal will be to further augment GDMT through use of these diagnostic categories.
Title: Impending “death” of autopsy: Analysis of resident’s attitude towards autopsy at a tertiary care center.

Authors: Pallavi Kopparthy MD, Amit Dhamoon MD, PhD; SUNY Upstate Medical University.

Introduction: “Tracking the necrotic footprints of our own missteps would teach us lessons far more memorable than any text could.”

Autopsy was once considered the gold standard for medical diagnosis. Recent data from United States National Center for Health Statistics (NCHS) has shown that autopsy rate of all deaths decreased from 19.1% in 1972 to 8.3% in 2003. At our institution, for the past year, number of deaths to autopsies ratio was 945/50 which is less than 5%. A number of reasons for this decline have been postulated, including improvements in diagnostic technology, fear of litigation and removal of defined minimum autopsy rate standards. Resident physician attitudes towards requesting an autopsy may also play a major role in declining rate of autopsies in recent years. The purpose of this study was to elicit attitudes of residents from various departments at a tertiary care center for decline in autopsy rates so that barriers for this valuable teaching tool can be addressed.

Methods: A voluntary 20-question survey regarding resident perceptions towards autopsy was distributed through email to 269 residents from Internal Medicine (IM), Surgery, Emergency Medicine, Family Medicine and Neurology.

Results: 155 of 269 residents responded to the survey with majority responses from IM. Among residents who responded, 84% believed autopsies improved management of subsequent patients and revealed medical conditions that affect the health of surviving relatives whereas 1.4% felt that autopsies did not improve management. 71% residents agreed that autopsies were an effective use of health care resources with <2% believing that they were not. When asked to identify barriers for not obtaining an autopsy, lack of proper training on how to request an autopsy (66%) was the most common reason cited. Other common reasons include the unpleasantness of requesting an autopsy from families (54%), perceived religious/moral beliefs of families (56%), advanced age of patients (59%) and a perceived obvious cause of death (56%). Interestingly majority of residents (54%) believed that concern for malpractice litigation was not a huge barrier as compared to other studies. Although autopsy was considered a useful tool by residents, 92% reviewed <3 autopsies in their training so far. Furthermore, 80% of residents from the above specialties did not have any formal education on how to communicate with family members while requesting an autopsy and about 83% were not aware of the steps involved in an autopsy.

Conclusion: From our multi-departmental single institution survey, it is evident that residents report educational value in obtaining autopsies; however several barriers, including inadequate training in how to have this difficult conversation with grieving families and inadequate information about autopsy procedures remain. A possible solution to this could be implementation of autopsy orientation program along with monthly interdisciplinary case conferences that include autopsy reports to “revive” this dying educational tool.

References
1. Sir William Osler
Title: Regulation of reactive oxygen species formation in red cells of sickle cell anemia patients using Vitamin C: an in-vitro study

Authors: Ogechukwu Egini¹ - first author, Edouard Guillaume¹, Eric Jaffe¹, Titilope Adeyemo², ¹Interfaith Medical Center, Brooklyn, NY 11213, ²Lagos University Teaching Hospital, Lagos, Nigeria

Introduction: Sickle red cells produce greater amounts of reactive oxygen species (ROS) than normal cells, leading to increased membrane damage. Under stress conditions, a multiplier effect is induced leading to generation of tremendous amounts of ROS which then lead to more widespread damage. The average red cell span is estimated at 1/5th the normal red cell. Theoretically, reducing ROS production will spare sickle red cells and improve life span.

Vitamin C is a free radical scavenger and has been shown to reduce ROS generation and improve recovery of pulmonary function in experimental mice model with induced pulmonary contusion. This in-vitro study investigated the effect of Vitamin C on ROS generation in stressed sickle red cells and also analysed demographic factors that might influence ROS formation in sickle red cells at baseline.

Methods: 27 HbSS patients were randomly recruited from the outpatient clinics of Lagos University Hospital, Nigeria. Inclusion included patients of all ages not on any multivitamin or blood transfusion within preceding 3 months and those using or not on hydroxyurea. If taking hydroxyurea, must be on stable dose for at least 3 months. With patient consent, demographic information and EDTA blood samples were collected. Individual samples were centrifuged and washed thrice in phosphate-buffered saline (PBS) before 500,000 cells from each sample were incubated individually in: (1) cellrox orange, a fluorophore stain followed by ROS counting on flowcytometer for baseline ROS documentation; (2) 80uM and (3)100uM Vitamin C concentrations followed by tertbutylhydroperoxide (TBHP) incubation then staining and cytometry. TBHP is an oxidizing agent. The control were red cells incubated in PBS and later TBHP followed by staining and flowcytometry. The test samples were the red cells incubated in Vitamin C.

Primary measure was comparison of amount of ROS generated in test samples vs matched control for all 27 samples. Secondary analyses evaluated the effect of gender, age, BMI and hydroxyurea use on baseline ROS formation.

Results: ROS counts per ul for Vitamin C treated cells were significantly lower than matched controls (p<0.001). Average ROS count for the 80uM test samples was 27.5ul (95% CI, 17.5-72.5) and 3.9ul for 100uM test group (95% CI, 1.9-5.9). Male gender was associated with significantly higher ROS formation than females (p<0.05). When a score of 1 was assigned each to male gender, hydroxyurea use, BMI>18.5 and age>20 in order to predict baseline ROS count, no significant association was noted. However, those with scores of 3 or more had higher ROS generation.

Conclusion: This study showed that ROS formation in sickle red cells treated with 80 and 100uM of Vitamin C before stressing was significantly reduced. It also showed that the male gender is an independent risk factor associated with ROS generation at baseline.

References

Title: Pharmacogenetic Association Study on Clopidogrel Response in Caribbean Hispanics with Peripheral Artery Disease

Authors: Dagmar F. Hernandez-Suarez, MD, MSc; Karid Nieves, MD; Hector Núñez-Medina, MD; Stuart A. Scott, PhD; Kyle Melin, PharmD; Angel Lopez-Candales, MD; Jorge Duconge, PhD

Introduction: Antiplatelet therapy with clopidogrel is recommended to reduce cardiovascular events in patients with peripheral artery disease (PAD); however, clopidogrel efficacy has not been adequately studied in this patient population. Therefore, we aimed to determine the effects of cilostazol therapy on platelet reactivity among PAD patients on clopidogrel.

Methods: We performed a cross-sectional pilot study of 46 Puerto Rican patients diagnosed with PAD. The cohort was divided based on use of clopidogrel and cilostazol (n = 24) or clopidogrel alone (n = 22). Platelet function was measured ex vivo using the VerifyNow P2Y12 assay. Genomic DNA was extracted from peripheral blood samples using the QIAamp DNA Blood Midi Kit, which was subjected to candidate variant genotyping (CYP2C19, ABCB1, PON1 and P2RY12) using TaqMan quantitative polymerase chain reaction assays. All analyses were performed using SAS version 9.4 (SAS Institute).

Results: Among all enrolled patients, 18 (39%) had high on-treatment platelet reactivity (HTPR). The mean platelet reactivity was 207 ± 53 (range, 78–325) with higher P2Y12 reaction units in the non-cilostazol group, 224 ± 45 vs. 191 ± 55 on the cilostazol group (p = 0.03). No significant differences were observed in the clinical or genetic variables between the two groups. A multiple regression analysis determined that history of diabetes mellitus (p = 0.03), use of cilostazol (p = 0.03) and hematocrit (p = 0.02) were independent predictors of platelet reactivity.

Conclusion: In Puerto Rican PAD patients on clopidogrel therapy, history of diabetes mellitus, use of cilostazol and hematocrit are independent predictors of platelet reactivity. Adjunctive cilostazol therapy may enhance clopidogrel efficacy among PAD patients with HTPR.
Puerto Rico - Research - Poster Finalist
Dagmar Fredy Hernandez Suarez, MD

Title: CYP2C19*2 and PON1 p.Q192R polymorphisms are associated with platelet reactivity to clopidogrel in Puerto Rican Hispanics with cardiovascular disease

Authors: Dagmar F. Hernandez-Suarez, MD; Stuart A. Scott, PhD; Matthew I. Tomey, MD; Mario J. Garcia, MD; Jose M. Wiley, MD; Pedro A. Villablanca, MD; Kyle Melin, PharmD; Angel Lopez-Candales, MD; Jorge Duconge, PhD

Introduction: High on-treatment platelet reactivity (HTPR) with clopidogrel imparts an increased risk for ischemic events in adults with coronary artery disease. Although more potent antiplatelet agents are available, clopidogrel remains the most commonly used P2Y12 inhibitor in Puerto Rico. Platelet reactivity varies with ethnicity and is influenced by both clinical and genetic variables; however, no clopidogrel pharmacogenetic studies with Puerto Rican patients have been reported. Therefore, we sought to identify clinical and genetic determinants of on-treatment platelet reactivity in a cohort of Puerto Rican patients with cardiovascular disease.

Methods: We performed a retrospective study of 111 Puerto Rican patients on 75 mg/day maintenance dose of clopidogrel. Patients were allocated into two groups: Group I, without HTPR; and Group II, with HTPR. Clinical data was obtained from the medical record. Platelet function was measured ex vivo using the VerifyNow® P2Y12 assay and HTPR was defined as P2Y12 reaction units (PRU) ≥ 230. Genotyping of CYP2C19, ABCB1, PON1, PY2R12, B4GALT2, CES1 and PEAR1 was performed using Taqman® Genotyping Assays.

Results: The mean PRU across the cohort was 203 ± 61 PRU (range, 8-324), and 42 (38%) patients had HTPR. One in four individuals carried at least one copy of the CYP2C19*2 variant allele. Hematocrit and PON1 p.Q192R variant were inversely correlated with platelet reactivity (p < 0.05). Multiple logistic regression showed that 27% of the total variation in PRU was explained by a history of diabetes mellitus, hematocrit, CYP2C19*2, and PON1 p.Q192R. Body mass index (OR = 1.15; CI: 1.03-1.27), diabetes mellitus (OR = 3.46; CI: 1.05-11.43), hematocrit (OR = 0.75; CI: 0.65-0.87) and CYP2C19*2 (OR = 4.44; CI: 1.21-16.20) were the only independent predictors of HTPR.

Conclusion: In a representative sample of Puerto Rican patients with cardiovascular disease, diabetes mellitus, hematocrit, CYP2C19*2 and PON1 p.Q192R were associated with on-treatment platelet reactivity. These factors may identify a subset of patients at higher risk for adverse events on clopidogrel in the Hispanic population.
New York-Research-Poster Finalist
Chayakrit Krittanawong, MD

Title: Association between Coffee Consumption and Atrial Fibrillation Risk: A Systematic Review and Meta-Analysis

Authors: Chayakrit Krittanawong, MD

Introduction: The etiology of atrial fibrillation (AF) is multifactorial including comorbid like hypertension, diabetes, or advanced age. Coffee is increasingly consumed around the world, particularly in the United States and it becomes necessary to assess the impacts of coffee on cardiovascular health. To date, there have been discrepant findings on whether coffee consumption is associated with the rate of developing AF. The purpose of this study was to explore the associations between coffee consumption and AF.

Methods: We conducted a comprehensive search of MEDLINE, MEDLINE In-Process & Other Non-Indexed Citations, EMBASE, Scopus, and the Cochrane Central Register of Controlled Trials from database inception through April 2017. Observational studies were included if the studies reported hazard ratios (HRs) or odd ratios (ORs) of the associations between coffee consumption and AF. Data were extracted by one reviewer and then reviewed by two independent reviewers. Conflicts were resolved through consensus. Using the DerSimonian and Laird random effects models, we calculated pooled HRs and pooled ORs with 95% confidence intervals (95% CIs). Subgroup analyses were performed to explore potential sources of heterogeneity. The quality of the included studies and publication bias were assessed.

Results: Out of 497 retrieved articles, we identified 2 cross-sectional and 9 prospective studies with a total of 339,692 individuals with 18,228 total AF events. The highest category of coffee consumption (≥5 cups/day) was not associated with increased or decreased risk of AF (pooled OR, 0.98; 95% CI: 0.88-1.09; p<0.001; I²= 59.21%), compared to the lowest category (1 cup/day).

Conclusion: We found no evidence that coffee consumption is associated with increased risk of AF.
Title: Comparing Surgical and Nonsurgical Management of Gallstone Disease in Geriatric Patients

Authors: Yousef Nassar, Seth Richter

Introduction: Choledocolithiasis or gallstones obstructing the common bile duct is a frequent complication of gallbladder stones. It may be associated with significant morbidity and mortality in elderly patients. Management of this condition in the geriatric population poses a clinical challenge as periprocedural complications increase as patients get older. We set out to investigate the difference in inpatient mortality and length of hospital stay in geriatric patients with complicated gallstone disease who underwent Endoscopic retrograde cholangiopancreatography (ERCP) alone compared to those who underwent both ERCP and cholecystectomy (EAC).

Methods: The Healthcare Cost and Utilization Project – Nationwide Inpatient Sample (HCUP-NIS) data from the year 2001-2014 were used in this study. Patients that were admitted to HCUP-NIS hospitals with the ICD 9 diagnostic codes of cholelithiasis and associated complications (574.xx, 575.0x-575.3x and 577.0), greater than 60 years of age were included in this study. Statistical analysis was performed using multiple logistic regression to compare inpatient mortality of patients managed with ERCP alone compared to those who underwent EAC. Ordinary linear regression was used to compare the length of hospital stay (in days) in the studied procedural groups. Results: were analyzed controlling for age, gender and cause of death.

Results: A total of 46,270 patients above age 60 who underwent ERCP or EAC in the management of complications of gallstone disease were included in our study. Patients between ages 60-70 who underwent ERCP only had a higher inpatient mortality rate when compared to the EAC group with an odds ratio (OR) of 4.068 (p <0.001). Patients between ages 70-80 who underwent ERCP alone had increased inpatient mortality rates compared to the EAC group, OR 1.65 (p <0.002). There was no statistically significant difference in the inpatient mortality rate in patients > 80 who underwent ERCP alone compared to the EAC group with OR 1.28 (p=0.09).

For patients aged 60-70, the mean length of hospital stay was shorter in the ERCP group compared to the EAC group by 0.5 days (p= 0.77) which was not statistically significant. Patients between ages 70-80 who underwent ERCP alone had a mean hospital stay that was 1.14 days (<0.001) shorter than the EAC group. Patients above age 80 who underwent ERCP alone had a hospital stay 2.29 days (p <0.001) shorter than the EAC group.

Conclusion: Management of choledocolithiasis in geriatric patients poses a challenge to clinicians. We investigated the use of ERCP alone compared to using EAC in their management. There was a lower inpatient mortality rate in the EAC group compared to ERCP alone in patients between 60-80 years of age. There was no significant difference in inpatient mortality in patients above 80 indicating the possibility of a more conservative management in this age group.
New York-Research-Poster Finalist
Vikashsingh Rambhujun, MBBS

Title: Effect of Statin Therapy on Severity of Illness in HFrEF

Authors: K. Wong, MD (1), V. Rambhujun MBBS (1), A Maidhoff DO (1), R. Davino MD (2), C. Healey MD (3), B Ray (2), (1) NYU Winthrop internal medicine department, (2) NYU Winthrop Cardiology department, (3) Albany Cardiology group

Introduction: Recent literature has demonstrated a possible role for inflammation in the development of heart failure with preserved ejection fraction (HFrEF). Statins have known anti-inflammatory, anti-oxidative and endothelial protective effects. This study examines the effects of statins on the severity of HFrEF. We hypothesize statin users (SUs) will have less severe outcomes than non-statin users (NSUs).

Methods: We retrospectively evaluated 273 consecutive patients with the primary diagnosis of acute HFrEF at Winthrop University Hospital in 2012. The patients were divided into 2 cohorts based on their use of background statins on admission. The 2 cohorts were compared for total hospital length of stay, number of readmissions for HF, admissions requiring intensive medical care, and length of stay (LOS) in intensive care units. Demographics and data were analyzed using Sigmaplot v.11.

Results: Our cohort had an average age of 77 years; 64% were female and the majority were Caucasian (81%). Approximately half were on statins at admission (48%). SUs had higher levels of serum BNP than NSUs (498 vs. 489, p = 0.002). SUs had decreased LOS compared to NSUs (10 days vs. 12 days, p = 0.04). There was no difference in CHF readmission rates between the two groups, but SUs had a longer period of time between admissions (259 vs. 243 days, p = 0.025). There was no difference in number of patients requiring intensive care (22% SUs, 28% NSUs), but NSUs required prolonged ICU time compared to SUs (6 vs. 4 days, p < 0.05).

Conclusion: Our study suggests that statin therapy may reduce LOS and need for intensive medical care, when comparing SUs with NSUs during acute HFrEF admissions. This suggests that statins may decrease severity of acute HFrEF. A prospective multi-center trial, investigating other markers of HFrEF severity and morbidity is warranted.
Title: Assessing the Impact of the Community-based Transitions Care Program on 30-day readmission rates at a tertiary care hospital: a retrospective study

Authors: Charles Ramkishun, Stony Brook University Hospital, Getu Teressa, Stony Brook University Hospital, Leanne Merril, Stony Brook University Hospital, Jere Freeman, Stony Brook University Hospital

Introduction: The 2007 Medicare Payment Advisory Commission Report identified that 13.3% of Medicare readmissions occurring within 30 days after the index hospitalization were potentially preventable and accounted for $12 billion in excess Medicare expenditures.¹ The finding led to increasing use of readmission rates as a proxy measure of overall hospital performance and prompted Medicare payment reforms to adjust reimbursement based on readmission rates.

As a response to excessive unplanned 30-day hospital readmissions, the Affordable Care Act established the Community-based Care Transitions Program (CCTP) to allot funding to community based organizations for providing care transition services to Medicare patients at high risk of readmission.²

Stony Brook University Hospital is a 603-bed tertiary care academic medical center located in Suffolk County, NY with a catchment area of 1.6 million, which experienced reduced readmission rates after implementing CCTP services to augment pre-existing discharge support services.

Methods: A retrospective chart review was conducted comparing readmission rates of patients receiving care transition services via a pre-existing telephone based nurse and pharmacist led intervention known as the BOOST Program and that of controls to patients receiving a combination of services from the BOOST program and the CCTP. Cases were identified by review of consecutive discharges of patients age 65 and older presenting to the hospital from home on a minimum of 5 medications prior to hospitalization admitted between January 1st to December 31st, 2015. The primary outcome measured was the relative difference in percentage of patients among the three groups that required readmission within 30 days of an index hospitalization as determined by Pearson’s Chi Square test.

Results: A total of 676 patients were included [364 patients (53.8%) in the BOOST group, 153 (22.6%) patients in the BOOST/CCTP group and 159 (23.5%) in the control group. The 30-day readmission rate for the BOOST group, BOOST/CCTP and control groups were 62 (17%), 12 (7.9%) and 26 (16.4%), respectively, indicating significantly less readmissions among BOOST/CCTP recipients (p=.024). The relative risk of readmission for BOOST/CCTP patients was 0.47 compared to controls suggesting that patients receiving BOOST/CCTP intervention were 52% less likely to be readmitted within 30 days as compared to controls (95% CI 8.4-75). Relative to the control group, the number needed to treat with the combined BOOST/CCTP intervention to prevent one readmission within 30 days is 12 patients (95% CI 6-82).

Conclusion: The study demonstrated a significant decrease in readmission rates in the BOOST/CCTP group as compared to the control and BOOST group, indicating that the CCTP program was an effective intervention whether by direct effect or through synergy with the BOOST program. The CCTP program focused primarily on patient empowerment and improving patient health literacy, which we feel is a major reason behind it’s efficacy. Use of its care transition model and careful selection of recipients may help reduce readmission rates in similar academic institutions.
References

New York-Research-Poster Finalist
Felix M Reyes, MD

Title: Potentially Inappropriate Medications at the time of admission in elderly COPD patients increase length of stay and resource utilization.

Authors: Felix M. Reyes M.D.¹, Justin Lee M.D.¹, Miguel Ramirez M.D.², Mafuzur Rahman M.D.², 1 Department of Medicine, SUNY Downstate Medical Center, Brooklyn, NY., 2 Clinical Assistant Professor, Department of Medicine, SUNY Downstate Medical Center, Brooklyn, NY.

Introduction: Aging populations and better management of chronic diseases have resulted in increasingly complex elderly patients. The elderly are frequently exposed to polypharmacy and any prescribed medication can lead to adverse events. To help clinicians decide the appropriateness of prescriptions, the American Geriatrics Society (AGS) published a set of criteria to follow in this population. To what extent Potentially inappropriate medications (PIM) affect the care of elderly patients with Chronic Obstructive Pulmonary Disease (COPD) patients is largely underreported.

Objective: To determine the PIM frequency in elderly COPD patients admitted to the general medicine wards at an urban teaching hospital.

Methods: A retrospective chart review was performed on all patients aged 65 years or older admitted to the medical wards from the period of July 2015 to June 2016. The AGS’s Beers criteria were operationalized and charts were screened for these prescriptions at the time of admission, during hospital stay and discharge.

Results: Of 229 patients, 42 patients (18.34%) had an existing diagnosis of COPD, out of which 14 patients (33%) were admitted under the diagnosis of COPD exacerbation. Of 14 COPD exacerbations 4 had no PIMs prescribed on admission and 10 patients had at least one PIM prescribed on admission (LR: 2.5). Admitted COPD patients without PIM prescription on admission had an average length of stay of 7.69 days, compared to 11.5 days on those prescribed PIMs at the time of admission. COPD patients with PIMs were less likely than those without PIMs to be discharged home without services (LR 0.25). COPD patients taking PIMs were more likely to be discharged to a Skilled Nursing Facility (LR: 9).

Conclusion: Use of PIMs by elderly COPD patients was associated with longer hospital stays and a discharge to a SNF. Elderly patients can have difficulty understanding new medications and long medication list which will result in more complicated discharge plans.

References

New York-Research-Poster Finalist
Daniel Sartori, MD

Title: New Curriculum Leads Toward Higher Value Care for Acute Pancreatitis

Authors: Christopher Sonne, Daniel J Sartori, June Ha, Shivani Thanawala, Jonathan Whitehouse, Kevin Hauck, Andrew Dikman

Introduction: Acute pancreatitis is the leading gastrointestinal cause for hospitalization in the US and thus represents a significant clinical resource burden for providers. Diagnostic criteria require two of the following: 1) abdominal pain; 2) serum lipase >3x the upper limit of normal (ULN); 3) characteristic findings from abdominal imaging. Imaging is thus not required to establish a diagnosis, however is still widely performed. We participated in a novel value-based care curriculum for trainees through which we sought to define the scope, patient characteristics, and provider trends associated with inappropriate utilization of abdominal CT at our institution, with the ultimate goal of implementing a targeted intervention to curtail unnecessary imaging.

Methods: We performed a single institution retrospective chart review of all patients with ICD-10 diagnoses of acute pancreatitis in the first two quarters of 2016 at NYU Langone Medical Center (NYULMC). Exclusion criteria consisted of absence of documented abdominal pain, lipase less than 3x ULN, and complicated pancreaticobiliary anatomy. Remaining patients thus met diagnostic criteria by lipase and clinical exam. Utilization of abdominal CT was then assessed. Multivariate regression analysis was used to predict which clinical characteristics and outcome data were associated with CT utilization.

Results: Eighty-six patients were admitted with acute pancreatitis at NYULMC over the study period, 37 of whom met inclusion criteria. Of these, 26 patients (70%) underwent abdominal CT. CT utilization was not associated with increased severity of pancreatitis (mean APACHE 7.6 +/- 5.2 vs. 8.2 +/- 6.9 for CT and no CT respectively, p = 0.876); (mean SAPS-II 24.5 +/- 10.4 vs.18 +/- 11, (p = 0.129). Average observed length of stay (OLOS) was longer for those undergoing CT (4.4 +/- 2.1 days vs. 3.6 +/- 1.7 days, p = 0.20), however this difference was not statistically significant. No variables, including severity of pancreatitis or etiology of pancreatitis, were independent predictors of OLOS. CT utilization was associated with higher mean serum lipase (4694 +/- 3279 U/L vs. 2848 +/- 1836 U/L, p = 0.025).

Conclusion: In this single-center retrospective chart review, nearly three quarters of patients with acute pancreatitis underwent unnecessary CT imaging. CT utilization was not associated with clinical severity however was associated with increased lipase levels, suggesting a perceived bias toward imaging patients based on degree of lipase elevation. CT utilization additionally reflected a trend toward increased OLOS, and was independent of the etiology of pancreatitis, suggesting that this effect was not driven by gallstone pancreatitis. These findings support development of value-based improvement initiatives to target the use of lipase in diagnosis in acute pancreatitis in an effort to reduce unnecessary imaging and OLOS in this patient population.
New York-Research-Poster Finalist
Christopher Su, MD

Title: Concurrent NSAID and warfarin use is associated with decreased time to therapeutic INR and increased transfusions in hospitalized patients

Authors: Christopher Su MD MPH, William Southern MD MS, Shitij Arora MD FACP

Introduction: Warfarin is commonly used in long-term anticoagulation and has a narrow therapeutic index. Dosing is dependent on intrinsic patient variables including cytochrome P450 2C9 (CYP2C9) and VKORC1 polymorphisms. High protein binding and CYP-dependent clearance mechanisms of NSAIDs also affect warfarin serum levels. This study investigates the clinical interaction of warfarin and NSAIDs administered concurrently on the time taken to reach therapeutic INR in hospitalized patients.

Methods: A retrospective, matched cohort study was performed on inpatients initiated on warfarin as part of a heparin “bridge” from January 2006 through January 2016 at an academic medical center. After patients exposed to NSAIDs while on warfarin (“cases”) were identified via the electronic medical record, patients receiving warfarin who were not exposed to NSAIDs (“controls”) were matched randomly to cases by age and gender. Paired t-tests were conducted for primary outcomes including time to therapeutic INR (>2), cumulative warfarin dose administered to achieve therapeutic INR, and the value of the first therapeutic INR. Additional tests were performed on secondary outcomes including change in hemoglobin from warfarin initiation to therapeutic INR, blood transfusions given during hospitalization, and differences between patients who received IV (ketorolac) and PO (ibuprofen or naproxen) NSAIDs. Multivariate linear regression models were generated to control for BMI, renal function, hepatic function, concurrent aspirin use, and other variables.

Results: One hundred and seventy pairs of matched cases and controls were identified, with decreased time to therapeutic INR (3.8 days vs. 4.6 days, p<0.001), decreased cumulative warfarin dose (27 mg vs. 35 mg, p<0.001), and higher first therapeutic INR (2.5 vs. 2.3, p=0.02) observed in cases who were exposed to NSAIDs. Cases experienced a significant decrease in hemoglobin levels while receiving warfarin (-0.71 g/dl vs. -0.45 g/dl, p=0.02) and increased transfusions during hospitalization (p<0.001). Additionally, patients who received IV NSAIDs experienced decreased time to therapeutic INR (3.5 days vs. 4.0 days, p=0.05) and decreased cumulative warfarin dose (23 mg vs. 30 mg, p=0.006) relative to patients who received PO NSAIDs. Multivariate analyses confirmed that concurrent NSAID use is significantly correlated with time to therapeutic INR (p=0.03) and cumulative warfarin dose (p=0.004).

Conclusion: Patients exposed to NSAIDs while on warfarin experience significantly decreased time to therapeutic INR, decreased cumulative warfarin dose, and higher first therapeutic INR, suggesting a phenotypic manifestation of NSAIDs delaying warfarin clearance. These conclusions are further preserved when stratifying for the route of administration of NSAIDs, signifying a potency-mediated effect. Finally, increased need for transfusion and significant interval decrease in hemoglobin among cases demonstrate the increased bleeding risk associated with concurrent warfarin and NSAID use. These findings illustrate a major clinically-relevant adverse drug interaction among two routine medications in the inpatient setting and underscore the importance of careful warfarin dosing in patients already receiving NSAIDs.
Title: Money Down the Drain: Opportunity to Reduce Unnecessary Urine Cultures

Authors: Don Bambino Geno Tai, MD; Amrah Hasan, MD; Sanchit Panda, MD; Robert Goldstein, MD, FACP

Montefiore New Rochelle / Albert Einstein College of Medicine, New Rochelle, New York

Introduction: United States healthcare faces a crisis of wasteful spending [1]. Urine cultures are often ordered together with urinalysis, regardless of results of the latter. Reducing unnecessary urine cultures can cut costs and workload for healthcare systems. Additionally, it could reduce treatment of asymptomatic bacteriuria, misdiagnosis of catheter-associated urinary tract infection, and curtail inappropriate antibiotic use [2,3].

Methods: The design was a retrospective study of patients in a community-based teaching hospital. All patients ages 12 years and older who had a urinalysis and urine culture done at the same time from January 1, 2016 to March 31, 2017 were included in the study.

Positive urine culture was defined as cultures with growth of more than 10,000 colony-forming units/mL of any bacteria. Multiple bacterial growths deemed as contamination by the microbiology laboratory were considered negative results. The cut-offs for variables in urinalysis considered positive were white blood cell (WBC) count of more than 10 per high-power field, any leukocyte esterase and nitrite other than negative, and any bacteria on microscopy. A urinalysis was considered high-risk if at least one of the variables was positive. A low-risk urinalysis was defined as a urinalysis which was negative on all the variables.

Results: There were 2,995 patients included in the study. Majority were female (60%, n=1789) and the average age was 65 years old (range 12-105). Majority of tests were ordered by medicine (74%, n=2210), followed by emergency medicine (16%, n=490). 74% (n=2203) of cultures were negative while 26% (n=792) were positive.

Among the four variables, presence of bacteria on urinalysis was the most sensitive in predicting a positive urine culture (88%). Nitrite had the highest specificity at 97%. Leukocyte esterase and bacteria had the highest negative predictive value (90%). All four had dismal positive predictive value, highest of which was nitrite (69%).

There were 2,203 high-risk urinalyses (74%) and 792 low-risk urinalyses (26%). The combined sensitivity of urinalysis was 94% while the negative predictive value was 94%.

There were 46 cases of false negative urinalysis. Only two cases had indication for treatment. Two other cases were treated with antibiotics but without indication. The rest were not treated.

Conclusion: Using the variables noted, urine cultures can be automatically cancelled if the urinalysis is deemed low-risk. This would have resulted in a 26% reduction in urine cultures with an estimated
savings of $8,000 not including cost of antibiotics and length of hospital stay. False omission rate was low (6%). By excluding asymptomatic bacteriuria, only 0.25% (n=2) would have been falsely omitted. Cancellation rate can be as high as 35-39% in the emergency department [4,5].

This study reinforced that reflex cancellation of urine cultures can significantly reduce unnecessary urine cultures even in the inpatient setting.

References


New York-Research-Poster Finalist
Linda-Marie Ustaris, DO

Title: Analyzing Hospital Readmissions in End-Stage Renal Disease Patients with and without Heart Failure

Authors: Linda-Marie Ustaris\(^1\), Nirvani Goolsarran\(^1\), Sandeep K. Mallipattu\(^1\), \(^1\)Stony Brook University Hospital, Stony Brook, NY

Introduction: Among Medicare beneficiaries above 66 years old, end-stage renal disease (ESRD) patients are two times more likely to be re-hospitalized within thirty days of discharge than persons without kidney disease. Heart failure is one of the most common diagnoses for readmission in the ESRD population. Although prior studies have identified ESRD as a high risk population for readmission, no studies have evaluated the likelihood of 30-day hospital readmission in patients with ESRD and heart failure. We aim to analyze the 30-day readmission rate in ESRD patients with heart failure compared to subjects without heart failure. We aim to report readmission rates related to sub-categories of heart failure.

Methods: We conducted a retrospective cohort study of 1533 ESRD patients at a suburban academic medical center with greater than one hospital visit within 30-days (total 18788 encounters). Outpatient encounters were excluded. 1156 ESRD patients and 4117 inpatient encounters met inclusion criteria. We categorized each visit by reason for admission as dialysis related or non-dialysis related. We collected baseline demographics and clinical data, including presence of reduced ejection fraction of <50% (HFrEF) and presence of left ventricular (LV) diastolic dysfunction with preserved ejection fraction of >50% (HFpEF). We calculated the proportion of ESRD related visits within each aforementioned echocardiogram sub-groups. Chi-squared Fisher’s exact test was used to calculate the \(p\)-value to establish statistical significance.

Results: Preliminary data was collected for 130 ESRD patients accounting for 928 encounters. 105 patients had evidence of heart failure and 25 patients had no evidence of heart failure \((p<0.05)\). Sub-group analysis of patients with heart failure showed: 72% (76/105) had HFpEF only, 26% (27/105) had HFrEF with LV diastolic dysfunction and 2% (2/105) had HFrEF only \((p<0.05)\). 81% of readmissions were in patients with heart failure compared to 19% in the group without heart failure \((p<0.05)\). More than half (54%) of total readmissions were from patients with HFpEF only. Of the 753 readmissions in patients with heart failure, 331 readmissions were dialysis related (vascular access issues, hypertension, electrolyte abnormalities, volume overload or other) and 422 readmissions were non-dialysis related \((p:<0.05)\). Overall, there was no statistically significant difference in the number of 30-day readmissions for dialysis related versus non-dialysis related visits, regardless of LV function \((p = 0.36)\).

Conclusion: Our preliminary data suggests that 30-day readmissions in the ESRD population are related to a high incidence of heart failure, particularly HFpEF. ESRD patients with heart failure represent a high risk and vulnerable population for readmission. Therefore, an effective readmission intervention that targets this group is likely to improve patient outcomes and health expenditure. Our next steps include a comprehensive cost analysis related to the ESRD and heart failure population, and assessment of a readmission tool that integrates ESRD to predict 30-day readmission.

References

Title: Amyloid-Beta Protein Impacting the Insulin Signaling Pathway in the Central Nervous System

Introduction: Alzheimer’s disease (AD) is characterized by amyloid-β (Aβ) toxicity, tau pathology, and neuroinflammation, all of which play roles in neurodegeneration. Insulin has polytrophic effects on neurons and may be at the center of these pathophysiological changes. It is well known that insulin receptor is involved in complex signaling pathways in cells of the central nervous system (CNS) and thus modulates downstream tau phosphorylation as well as cell survival. Within this context, we investigate how CNS insulin signaling pathway responds to Aβ in vitro.

Methods: Our group uses an immortalized mice neuroblastoma 2a cell line (N2a). A subset of cells was stably transfected with human APP gene (N2a-APP), which upregulates amyloid precursor protein (APP) expression. Baseline insulin receptor signaling proteins including IRS1, Akt, GSK3β, mTor, tau and tubulin levels were determined by ELISA in the original N2a and transfected N2a-APP cells. Both were subsequently treated with solubilized human recombinant insulin at concentrations of 0.5nM and 5nM for different time intervals up to 8 hours. IRS1 and phosphorylated Akt levels, early responders in the signaling pathway, were contrasted between two cell lines. Lastly, both groups were treated with Aβ Peptide binding peptide (ABP) for APP removal to measure the responses of insulin receptor signaling pathways in N2a and N2a-APP.

Results: The over-expression of APP and the production of Aβ peptides in N2a-APP cells alter the levels/phosphorylation states of insulin pathway proteins. This process dampens the cellular response to insulin treatment as compared to parental N2a cells, indicating insulin insensitivity or resistance. However, removal of this extra Aβ in N2a-APP cells with ABP recovered the sensitivity or response of the cells to insulin treatment, as demonstrated by increased phosphorylation levels in IRS-1, Akt, mTor and p70S6K proteins.

Conclusion: Our study has proved that Aβ can competitively binds at insulin receptors in CNS and thus, leading to insulin resistance. This condition dysregulates the function and metabolism of neural cells. Enhancement of insulin signaling by the application of ABP relieved the insulin-resistant phenotype and the dysregulated insulin signaling pathway, and inhibited the pro-inflammatory response. Alleviation of insulin resistance in AD brain may be a future direction of therapeutic development to relieve or delay the progression of the disease and neurodegeneration.
New York-Research-Poster Finalist
Srikanth Yandrapalli, MD

Title: Association of Comorbidity burden with Survival after In-hospital Cardiac Arrest

Authors: Srikanth Yandrapalli, MD¹, Venkat Lakshmi Kishan Vuddanda, MD², Amanda Lloji, MD¹, Gabriela Andries, MD¹, Syed Zaid, MD¹, Shantanu Solanki, MD¹, Christopher Nabors, MD, PhD¹, ¹ Department of Medicine, New York Medical College at Westchester Medical Center, Valhalla, NY., ² Harvard Medical School, Boston, MA.

Introduction: Whether or not to undergo cardiopulmonary resuscitation (CPR) during a hospitalization is a question that an increasing number of patients and providers consider as the United States (U.S.) population ages. Limited data are available for guidance. At one European center, increasing disease burden, as defined by the Age-combined Charlson comorbidity Index (ACCI) score, was associated with significantly lower survival following in-hospital cardiac arrest (IHCA). These findings could have broad use as an aid to decision-making related to code status. We therefore sought to determine if a similar association exists in hospitalized patients in the U.S.

Methods: We identified hospitalizations in patients ≥ 18 years who underwent CPR for IHCA in the U.S. National Inpatient Sample databases 2013-14 using relevant ICD-9-CM codes. Cases with primary diagnosis of cardiac arrest were removed to exclude possible out-of-hospital cardiac arrests. ACCI is among the widely used indexes of comorbidity. Comorbidity burden was assessed using ACCI scores with degree of disease burden stratified into low (0–4 points), moderate (5–9) and high (≥10) categories. The cohort was stratified by primary discharge diagnosis into sepsis, cardiac or respiratory related hospitalizations – as they constituted 64% of the sample. Association of comorbidity burden with survival to discharge was assessed with survey design logistic regression models adjusted for year, sex, and the presence of a shockable rhythm. Results: are presented as adjusted odds ratios (ORs) with 95% confidence intervals (CIs).

Results: Of the 211,260 hospitalizations with an IHCA (mean age 66 years, 43% women), patients survived to discharge in 65,395 (31%; mean age 64 years, 43% women). Overall, 37.4% had low ACCI comorbidity burden, 53.5% had moderate burden and 9.1% had high burden. Survival to discharge among the respective groups was 35.2%, 29.3%, and 23.4% (P<0.001). Compared to those with low comorbidity, patients with moderate (OR 0.76, 95% CI 0.72 - 0.79) and high comorbidity burden (OR 0.57, 95% CI 0.52 - 0.62) had significantly lower rates of survival to discharge. Among those hospitalized for sepsis, survival was significantly reduced only in the high comorbidity group (OR 0.71, 95% CI 0.58 - 0.87). The odds of survival were significantly lower in moderate and high comorbidity groups compared to the low comorbidity group in patients hospitalized for either pulmonary (moderate comorbidity: OR 0.81, 95% CI 0.73 to 0.90; high comorbidity: OR 0.69, 95% CI 0.56-0.84) or cardiac cause (moderate comorbidity: OR 0.64, 95% CI 0.59-0.69; high comorbidity: OR 0.44, 95% CI 0.38-0.52).

Conclusion: In the U.S., patients with high age-combined comorbidity burden were significantly less likely to survive IHCA, an effect which varied modestly based on the reason for hospitalization. These findings may facilitate shared decision making regarding code status between patient/family and provider, especially in patients with higher comorbid disease burden.
Title: Assessing Clinical Outcomes in Colorectal Cancer with Assay for Invasive Circulating Tumor Cells

Authors: Kevin Zarrabi MD, Wei Hou PhD, Wen-Tien Chen PhD, Yue Zhang MD MPH

Introduction: Colorectal carcinoma (CRC) is the second leading cause of cancer-related mortality in the United States. Lack of diagnostic and prognostic biomarkers remains an area of urgent unmet need. Circulating tumor cells (CTCs) are believed to be cells that have detached from the primary tumor and entered the circulatory system. CTCs have been identified in the venous blood of patients with a variety of cancers and they are considered to be responsible for the metastatic process. The aim of this study is to utilize a novel cell CTC enrichment assay to evaluate the association between the levels of invasive CTCs with CRC patient outcomes.

Methods: Peripheral blood from 93 patients with stage I-IV CRC was obtained and assessed for the presence of invasive CTCs. CTC isolation and enrichment was accomplished by our novel CTC isolation assay, Vita-Assay™. The invasive CTCs were identified through cell surface expression of epithelial markers (Epi+) and by their ability to invade a collagen adhesion matrix (CAM+). Patients were followed prospectively and assessed for overall survival.

Results: Of 93 patients enrolled in the study, 88 (94%) had detectable CTCs in serum samples. Patient CTC’s ranged from 0 – 470 CTCs/mL. Patients with stage I, II, III and IV disease exhibited mean CTC counts of 8.6 CTCs/mL, 35.8 CTCs/mL, 65.9 CTCs/mL, and 144.8 CTCs/mL, respectively (p<0.001). Kaplan-Meier curve analysis demonstrated a significant survival benefit in patients with low CTC counts compared to patients with high CTC counts (log-rank p<0.001). Multivariable Cox model analysis revealed that CTC count is an independent prognostic factor of overall survival (p= 0.009). Disease stage (p =0.01, Hazard Ratio 1.66; 95% CI: 1.12-2.47), and surgical intervention (p =0.03, Hazard Ratio 0.37; 95% CI: 0.15-0.92) were also independent prognostic factors. Gender, race, age, chemotherapy treatment, radiation treatment, and primary tumor location (colon or rectal) did not show survival difference.

Conclusion: Invasive CTC’s isolated from the serum of patients with CRC can be identified through the novel CAM assay. Moreover, the number of invasive CTCs inversely correlated with overall survival. Invasive CTCs are a promising prognostic tool in patients with CRCs.
Title: The FaceTime Fraction: Making Rounds Shorter while Increasing Patient-Physician Interaction

Authors: Kathryn Haroldson, MD, MPH; Hitesh H Patel, MD, MBA; Anthony Mazzella, MD; Rosanne Tiller, MD; and Debra Bynum, MD, MMEL, Department of Internal Medicine, University of North Carolina Hospitals, Chapel Hill, North Carolina

Introduction: The current interaction between daily rounding teams and patients on inpatient medicine wards is ineffective, repetitive, and not patient-focused. In modernized healthcare settings with increasing patient complexities, electronic health records, and multidisciplinary resources, traditional rounding strategies lack efficiency. Restructuring rounds to focus more on the patient is important to increase the team’s productivity and decrease the patient’s sense of vulnerability.

Methods: Changes to daily morning rounds on one general medicine service at a tertiary academic hospital were implemented for an 8 week period. All team members were expected to review patient data prior to formal rounds. Discussions using the traditional SOAP format outside of the patient’s room were discouraged and instead the team initially entered the patient’s room to involve them more in the decision-making process of their own medical care. The presenter used an assessment and plan based presentation at the bedside while sitting in a chair to emphasize eye-level communication. Time motion analysis was performed to determine how these changes effected daily workflow. A designated team member tracked the amount of time spent walking, talking inside each patient’s room, and discussions outside each patient’s room. Timing data was documented on a standardized spreadsheet. The patients were given “face sheets” with pictures of the team members including names and role descriptions. Timing data were compared to a traditional rounding strategy (SOAP presentation outside the room followed by an in-person update to the patient from the medical team) obtained in the same manner for 2 weeks preceding the implementation of the new bedside rounding strategy. Pre and post-pilot surveys were distributed to residents, attendings, and nurses. Patient satisfaction surveys were analyzed as well.

Results: The primary concern raised by residents regarding bedside rounds was that it would prolong rounding time. Average time spent per patient encounter decreased from 11’45” to 9’22” (p<0.0001), with an average increase in time spent with the patient from 4’43” to 6’31” (p<0.0001). The FaceTime Fraction, or the fraction of time spent in front of a patient compared to the total time spent discussing the patient, increased from 40.2% to 69.6%.

Conclusion: The new bedside rounding method has significantly decreased the total rounding time while increasing the amount of time spent with the patient. The initiative has received positive feedback from physicians, nurses, and perhaps most importantly, patients.
Ohio-Research-Poster Finalist
Amer Aldamouk

Title: S-Nitrosohemoglobin, Organ Oxygenation and Micro-circulation in Human After Brain Death

Authors: Amer Aldamouk, MD, Maroun Matta, MD, Silvia Perez-Protto, MD, Lin Zhu, MD, PhD, Alfred Hausladen, PhD, Jonathan S Stamler, MD and James D. Reynolds, PhD

Introduction: Transplantable organ function following brain death is negatively impacted by reduced perfusion and increased inflammation, processes regulated by protein S-nitrosylation; this dysregulation continues during storage. We will depict the effects of BD on protein nitrosylation, particularly on S-nitrosohemoglobin and micro-vascular blood flow in human donors.

Methods: Once BD is confirmed and consent for organ donation is obtained, a standard donor support regimen is enacted. Serial sub-lingual images of the micro-vasculature will be obtained at regular intervals during the support phase. Arterial blood samples were collected at the same intervals for determination of red blood cell S-nitrosohemoglobin levels, using mercury-coupled photolysis-chemiluminescence. Arterial and venous blood gas parameters were measured to determine tissues oxygen utilization. In addition, we will use NIRS probes for continual monitoring of tissue oxygenation and blood flow. Tissue oxygenation was measured in the buccal mucosa using a spectrophotometric monitoring system (T-Stat). Therapy and monitoring will continue through organ procurement.

Results: Circulating nitric oxide bioactivity (S-nitrosohemoglobin) was markedly disrupted after confirmation of brain death in human donors—a disruption that was associated with decline in tissue blood flow and oxygenation.

Conclusion: Maintenance of endocrine nitric oxide bioactivity after brain death may provide a notable means to improve the quality of organs available for donation by improving tissue blood flow and oxygenation.
Ohio-Research-Poster Finalist
Arjan S Flora, MD

Title: Development of a Body Surface Area Classification of Obesity Using Pulmonary Physiology

Authors: Arjan S. Flora, MD1; Hermann Simo, MD2; Aaron Baugh, MD2; Mohammad Saud Khan, MD2; Carson Oostra, MD2; Divya Sachdev, MD2; Dawn-Alita Hernandez, MD, MSPH1, 1Division of Pulmonary, Critical Care, and Sleep Medicine, Department of Internal Medicine, University of Toledo, Toledo, OH, 2Department of Internal Medicine, University of Toledo, Toledo, OH

Introduction: Severity of obesity has traditionally used body mass index (BMI); however, variables such as age, gender, and lean muscle mass may cause misclassification of disease. BMI does not distinguish the distribution of fat among individuals. Body surface area (BSA) is a better index for metabolic mass in the obese and has been used in body fat estimation1-2. A grading system has not been made to determine the severity of obesity using BSA. We propose a preliminary classification system based on carbon dioxide (CO2) retention and lung volumes in patients without central airway obstruction.

Methods: Retrospective review of awake arterial blood gas measurements for CO2 and pulmonary function testing (PFT) performed on adult patients at the University of Toledo between 2010 and 2017. Measured BMI and BSA (via the DuBois equation) were compared. Exclusion criteria included incomplete data, pH <7.35 or >7.45, HCO3 <23 mEq, and FEV1/FVC ratio <70%.

Results: Of 108 patients who met criteria, 57.8% female, 70.6% white, 21.7% black, average age 55.2 (18-90) years.

BSA (m2) for normal CO2 (≤44mmHg): 2.15±0.24 male, 1.91±0.25 female; elevated CO2 (45-49mmHg): 2.16±0.31 male, 2.02±0.23 female; severely elevated CO2 (>50mmHg): 2.28±0.47 male, 2.17±0.36 female.

Class was determined from above and known normals (1.9 m2 for males and 1.6 m2 for females), with BMI, CO2, and PFTs compared to confirm physiologic changes with obesity (Table 1).

Table 1. Comparison of Pulmonary Physiology Parameters between BSA Obesity Classes

<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>High Normal</th>
<th>Large</th>
<th>Very Large</th>
<th>Super Large</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Male BSA &lt;1.9</td>
<td>Male BSA 1.9-2.09</td>
<td>Male BSA 2.1-2.19</td>
<td>Male BSA 2.2-2.29</td>
<td>Male BSA &gt;2.3</td>
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<tr>
<td></td>
<td>Female BSA &lt;1.6</td>
<td>Female BSA 1.6-1.89</td>
<td>Female BSA 1.9-2.09</td>
<td>Female BSA 2.1-2.19</td>
<td>Female BSA &gt;2.2</td>
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<td></td>
<td>(n = 22)</td>
<td>(n = 55)</td>
<td>(n = 43)</td>
<td>(n = 26)</td>
<td>(n = 34)</td>
</tr>
<tr>
<td>BMI (kg/m2)</td>
<td>24.72 ± 3.12</td>
<td>28.75 ± 3.89</td>
<td>34.58 ± 4.48</td>
<td>37.68 ± 5.53</td>
<td>44.29 ± 7.74</td>
</tr>
<tr>
<td>pCO2 (mmHg)</td>
<td>40.82 ± 4.16</td>
<td>40.22 ± 3.03</td>
<td>39.44 ± 3.62</td>
<td>40.4 ± 5.00</td>
<td>41.45 ± 6.14</td>
</tr>
<tr>
<td>FVC (% predicted)</td>
<td>98.09 ± 24.01</td>
<td>96.24 ± 18.67</td>
<td>93.30 ± 19.36</td>
<td>92.38 ± 15.6</td>
<td>85.55 ± 17.46</td>
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<tr>
<td></td>
<td>SVC (% predicted)</td>
<td>ERV (% predicted)</td>
<td>RV (% predicted)</td>
<td>TLC (% predicted)</td>
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<td></td>
<td>102.64 ± 26.58</td>
<td>99.55 ± 19.27</td>
<td>96.74 ± 17.56</td>
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<td></td>
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<td>96.88 ± 17.56</td>
<td>97.50 ± 14.46</td>
<td>86.88 ± 15.92</td>
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<td></td>
<td>99.55 ± 19.27</td>
<td>76.33 ± 50.611</td>
<td>96.74 ± 17.56</td>
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<td></td>
<td>96.88 ± 17.56</td>
<td>77.42 ± 44.49</td>
<td>97.50 ± 14.46</td>
<td>86.88 ± 15.92</td>
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<td></td>
<td>97.50 ± 14.46</td>
<td>60.48 ± 48.01</td>
<td>97.50 ± 14.46</td>
<td>86.88 ± 15.92</td>
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</tbody>
</table>

**Conclusion:**

The proposed classification system reflects the physiologic changes with obesity in the medical literature\(^3\)\(^-\)\(^5\). BSA, especially in the morbidly and super obese, can better distinguish extraparenchymal thoracic wall restriction and CO\(_2\) retention with cut-offs of 2.3 in males and 2.2 in females. Our main limitation is a small study population. Future studies correlating other physiologic measurements and morbidity and mortality may further delineate the usefulness of BSA for clinicians treating obese patients.

**References**

Ohio-Research-Poster Finalist
Garima Gupta, MD

Title: Non-mosaic somatic HIF2A mutations associated with late onset polycythemia-paraganglioma syndrome

Authors: Garima Gupta1 M.D., Ying Pang2 M.D., Ph.D., Chunzhang Yang3 M.D., Ph.D., Zhengping Zhuang1 M.D., Ph.D. and Karel Pacak2 M.D., Ph.D., D.Sc., FACE, 1 Department of Medicine, The Jewish Hospital of Cincinnati, Cincinnati, OH, 2 Section on Medical Neuroendocrinology, Eunice Kennedy Shriver National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, MD, 3 Neuro-Oncology Branch, Center for Cancer Research, National Cancer Institute, Bethesda, MD

Introduction: There is increasing evidence to support the role of pseudohypoxia and hypoxia in erythropoiesis and tumorigenesis. Somatic mutations in hypoxia-inducible factor 2α (HIF2A) and germline mutations in prolyl hydroxylase type 1 and 2 (PHD 1 and 2) are associated with polycythemia-paraganglioma syndrome. Specifically, the classic presentation of female patients with recurrent paragangliomas, congenital polycythemia and somatostatinomas has been described as Pacak Zhuang syndrome. Studies have demonstrated that HIF2A mutations occur as post zygotic events and therefore may be associated with somatic mosaicism affecting hematopoietic and other tissue precursors. This phenomenon explains the development of early onset polycythemia in the absence of erythropoietin secreting tumors.

Methods: Whole exome sequencing (WES) was applied to paraganglioma DNA obtained from three female patients with a history of polycythemia and recurrent paragangliomas to screen for novel genetic alterations associated with this syndrome. All variations were filtered by population frequency, functional predictability and pathogenicity. Sanger sequencing was performed to validate the variations. To detect mosaicism of the target variations in blood genomic DNA, we performed PCR assay followed by TA cloning, which has previously detected mosaic variants with an allele frequency as low as 4%.

Results: Somatic HIF2A mutations (p.A530V, p.P531S and p.D539N) were identified in DNA extracted from paragangliomas from all three patients. No somatic mosaicism was detected through deep sequencing of blood genomic DNA. While polycythemia was diagnosed at an earlier age (30, 17 and 15) in all three patients, paragangliomas developed later at age 34, 55 and 30, respectively. No positive family history was reported. None of the patients were noted to have somatostatinomas or other neuroendocrine tumors. All three paragangliomas were mainly secreting norepinephrine. One out of the three patients developed metastatic disease localized to the retroperitoneum. Two patients were also noted to have ophthalmological findings including proliferative retinopathy and juxtapapillary retinal capillary hemangiomas.

Conclusion: These findings suggest that newer techniques need to be developed to detect somatic mosaicism in patients with this syndrome. Absence of mosaicism in patients with somatic HIF2A mutations is associated with late onset of the disease, milder clinical phenotype and an improved prognosis compared to patients with mosaicism.

References

Title: Better Utilization of Lab Testing to Reduce Medical Costs


Introduction: As part of the ABIM Choosing Wisely initiative, the Society of Hospital Medicine has emphasized repetitive complete blood count (CBC) and basic metabolic panel (BMP) testing among its top five practices that physicians and patients should question.

Reduction of unnecessary laboratory testing is important not only for cost containment, but also for improved patient experience and avoidance of iatrogenic harm.

Methods: Education and feedback tailored for internal medicine residents including: Lectures, Monthly faculty reminders, Email updates, Lab cost badge displays.

Residents were encouraged to evaluate each patient daily for the need for daily CBC and BMP.

CBC and BMP orders per patient/per day on General Med/Surg floors order by IM residents. Data from ICU excluded. CBC and BMP orders per patient/per day on General Med/Surg floors order by IM residents. Data from ICU excluded. Pre-intervention data from 07/2015 to 09/2016. Post-intervention data from 09/2016 to 12/2016. Interventional Maintenance 01/2017 to 06/2017

Pre-and post-intervention compared using two-sample Poisson rate.

Results: Pre-Intervention (2015)= 1.18 BMP/patient/day. 1.19 CBC/patient/day.

Post- Intervention (2016)= 0.91 BMP/patient/day. 22.8% reduction (p<0.001). 1.06 CBC/patient/day. 10.9% reduction (p = 0.014).

Overall (2016-2017). 0.93 BMP/patient/day. 21.2 % reduction (p < 0.001) 1.03 CBC/patient/day. 13.4 % reduction (p < 0.001).

Conclusion: Most importantly, less patient harm incurred. IM Residency average daily census: 33.9 patients/day. Interventions have reduced 3,093 BMPs/ year and 1,979 CBCs/year. This results in $194,964 per calendar year in direct testing costs. There is likely a reduction in Phlebotomy and Physician work hours.

Intervention only took 16 to 20 person-hours. There was a small cost to make lab cost badge displays. With more time and further education, we believe our goal reduction in CBC/BMP ordering can be reached, expanded, and more specifically targeted. We will continue to perform analysis for targeted interventions, future directions.

References


5. Mount Carmel West Charge Database
Title: Bowel Irrigation Lengthens Hospital Stay in Patients with Foodborne Botulism

Authors: Hoda Ilias DO, Anokh Kondru DO, Elizabeth Walz MD, Andrew Murry MD FACP, Mike Tobin CSSBB(ASQ), Jarrod Bruce MD

Introduction: Foodborne botulism occurs from exposure to preformed neurotoxin produced by the anaerobic bacterium *Clostridium botulinum*. Symptoms range from gastrointestinal upset to autonomic dysfunction and symmetric muscle paralysis. Guidelines for diagnosis and management of foodborne botulism are based predominantly on case reports. The administration of cathartics and enemas to remove unabsorbed botulism toxin from the gastrointestinal system of infected patients is thought to reduce disease severity and duration. In April 2015, the largest foodborne botulism outbreak in the United States in decades was reported in Lancaster, Ohio secondary to contaminated potato salad from home canned potatoes at a local church potluck. The aim of this study was to determine if bowel irrigation in botulism infected patients decreased their hospital length of stay (LOS).

Methods: This was a retrospective chart review examining 33 patients with confirmed or suspected botulism and concerns of exposure to the contaminated potato salad who presented to the local emergency room at Fairfield Medical Center (FMC) in Lancaster, Ohio. The majority of patients received care at FMC. Botulism patients that were transferred to 4 surrounding facilities due to hospital capacity were also included in the chart review. Charts of patients that underwent Golytely bowel irrigation were reviewed. LOS was compared between bowel treated patients and non-bowel treated patients. We hypothesized that patients who underwent bowel irrigation would have reduced hospital LOS.

Results: Of the 33 patients included in the study, 14 underwent bowel irrigation and 19 did not. Initial hospital LOS ranged from 1 to 21 days. Data was analyzed with a 2-sample t-test and Chi-Square. 2-sample t-test with a p value of 0.040 showed that the mean LOS for bowel treated patients was significantly higher at 5.9 days than the patients without bowel irrigation with an average LOS of 2.1 days. 71% of bowel treated patients had a hospital LOS greater than 2 days, while only 37% of patients who had no bowel irrigation had LOS greater than 2 days. Chi-square analysis yielded a p-value of 0.049 indicating statistical difference.

Conclusion: Botulism treatment guidelines are rooted in pathophysiology and review of previous outbreaks due to the rarity of the disease. The botulism outbreak in Lancaster provides a unique opportunity to study botulism management and treatment. Our data indicate that bowel irrigation of foodborne botulism infected patients is associated with increased hospital LOS. Guidelines do not currently recommend bowel irrigation as a treatment option and our data are consistent with this recommendation. Our study is limited by size and lack of standardization of bowel irrigation timing in clinical course. There is concern that the sickest patients received bowel irrigation.

References

Ohio-Research-Poster Finalist
Sravani Kamatam, MBBS

Title: BMI or Waist Circumference: Which is the better predictor for Hypertension in adult rural Indians?

Authors: Sravani Kamatam MD, Arunbabu Sankaranarayanan MD, Keyvan Ravakhah MD

Introduction: Overweight and obesity status of all populations is not adequately reflected by the BMI cut-offs defined by the World Health Organization (WHO). Thus, universal applicability of BMI among various ethnic groups, age, gender is raising a concern and becoming a less reliable predictor of cardiovascular risks especially hypertension. The aim of our study is to determine the predictive role of BMI and Waist Circumference (WC) in association with Hypertension (HTN) among adults in rural India.

Methods: A cross-sectional survey was conducted among adult patients who participated in the outpatient medical camp in rural India. A total of 137 participants were randomly included in the study and exclusion criteria include h/o diabetes, hyperlipidemia, alcohol use and smoking. BMI, WC and Blood Pressure (BP) with appropriate cuff size were measured at the time of visit to the medical camp. Continuous variables were used and expressed as mean ± SD. A Pearson correlation was used to test the correlation between BP with BMI and WC. Linear regression analysis was done to predict Systolic BP based on WC and BMI. The level of significance was 5% and confidence interval 95%.

Results: Among 137 participants, the overall prevalence of HTN was 45.3%, overweight 82.5%, and abdominal obesity was 39.4%. Pearson correlation analysis showed a statistically significant (p<0.000) positive correlation of 0.434 between systolic BP and WC and no correlation between systolic BP and BMI (p<0.997). Linear regression analysis was done to predict Systolic BP based on WC and a statistically significant regression equation was found for WC (F (1,134) = 31.108, p<0.000), with a R2 of 0.188, whereas regression equation for BMI was not statistically significant (F (1,134) = 0.000, p<0.997), with a R2 of 0.000. From the analysis, cut off of WC for HTN diagnosis with a systolic BP of 140 mm Hg was 38 inches for both males and females.

Conclusion: From the analysis, we have found 18.8% (p<0.000) of the variation in systolic BP was due to WC and had no relation with BMI. Due to significant variations in body fat composition and distribution in target populations, WC can be used other than BMI in assessing risk for hypertension. There is a strong statistically significant correlation between HTN and WC, when compared to BMI and the cut off values for WC is same for both males and females. WC provides a simple method for assessing the risk factors of HTN especially in targeted populations like the medically underserved Asian population in rural India. From this study, we recommend that WC can be used as a predictor variable for HTN than BMI and more research needs to be done on larger population across various demographics on WC and determining cut off values as cardiovascular risk factors.
Title: The T786C Mutation in the Endothelial Nitric Oxide Synthase (eNOS) Gene, a Pathoetiology of Osteonecrosis

Authors: Amir M. Khan MD, Joshua Choi MD, Richard A. Freiberg MD, Charles J. Glueck MD, Naila Goldenberg MD, and Ping Wang PhD

From the Jewish Hospital—Mercy Health, Graduate Medical Education, Cincinnati, OH

Introduction: Mutations in the endothelial nitric oxide (NO) synthase gene (eNOS T786C) have been associated with osteonecrosis (ON) and Prinzmetal’s angina (PMA). NO is necessary for normal bone health and ameliorates angina in PMA patients who fail to respond to conventional therapy. We compared T786C eNOS mutations in primary and secondary ON, normal controls, and PMA.

Methods: PCR analysis for eNOS T78C mutations were performed in 146 patients with primary ON (not secondary to high-dose-long term steroids or alcohol), 96 secondary ON, 114 PMA, and 83 normal controls.

Results: Compared to controls, patients with primary ON were more likely to have eNOS hetero-homozygosity (p<.0001) and had more mutant alleles (42% vs 22%, p<.0001). Patients with secondary ON differed from controls, being more likely to have eNOS hetero-homozygosity (p=.044) and more mutant eNOS alleles (32% vs 22%), p=.045. Primary ON and PMA patients did not differ for eNOS hetero-homozygosity (p=0.7) or for mutant eNOS alleles (42% vs 41%, p=.7). PMA patients differed from controls, being more likely to have both eNOS hetero-homozygosity (p=.0001) and mutant eNOS alleles, 41% vs 22%, p =.0001. Of 146 patients with primary ON, 65 (45%) had none of the five thrombophilias (Factor V Leiden heterozygosity, high Factors VIII and XI, high ACLA IgM, high homocysteine) which otherwise separated ON patients from controls, p<.05. Of those 65, 32 (49%) had eNOS heterozygosity and 9 (14%) eNOS homozygosity vs 76 controls with no thrombophilias, 28 (37%) of whom had eNOS heterozygosity and 2 (3%) homozygosity, p=.0012. The 65 ON patients with no thrombophilia-hypofibrinolysis were more likely to have mutant eNOS alleles (38%) than the controls (21%), p=.0013. There were no associations between eNOS hetero-homozygosity and the 5 major thrombophilias in primary ON patients. Of 146 patients with primary ON, 41 (28%) had eNOS hetero-homozygosity as their only abnormality, as did 30 of 96 (31%) secondary ON patients.

Conclusion: The high frequency of eNOS T786C hetero-homozygosity in patients with primary ON without thrombophilia raises the question of whether treatment with L-arginine or L-citrulline would slow or stop progression of ON, in a manner similar to anticoagulants halting the progression of ON when associated with thrombophilia. The T786C eNOS mutation is common in patients with primary ON (51% GA, 17% AA), not different than in PMA where the mutation is a known pathoetiology (50% GA, 16% AA), and much more common than in controls (37% GA, 4% AA). eNOS T786C hetero-homozygosity is the only identifiable etiology in 28% of primary ON patients and 32% of secondary ON patients.
Whether treatment with 9 g/day of L-arginine or 800 mg/day L-citrulline, both shown to be effective in PMA, would be beneficial in ON remains to be determined by placebo-controlled trials.
Ohio-Research-Poster Finalist
Morgan M Manley, DO

Title: Improving Vaccination Status in Solid Tumor Patients Receiving Chemotherapy

Authors: Morgan Manley, DO; Susannah Cooper, MD; Daniel Cybulski, MD; Ronald Markert, PhD

Introduction: Pneumonias are estimated to cause or complicate nearly 10% of hospital admissions among patients with cancer. The Centers for Disease Control and Prevention lists the efficacy for pneumococcal vaccines as 75% effective against invasive pneumococcal pneumonia for the conjugate vaccine and 60% effective for serotypes included in the polysaccharide vaccine. The National Comprehensive Cancer Network (NCCN) recommends that solid tumor patients receive pneumococcal and influenza vaccinations before chemotherapy initiation. Prior to October 2015, the Wright-Patterson Air Force Base (WPAFB) Hematology-Oncology clinic had no formal process to ensure patients receive these vaccinations. Our objective was to improve the rates of pneumococcal and influenza vaccinations among solid tumor patients presenting to the WPAFB Hematology-Oncology Clinic for chemotherapy by using a two-step process.

Methods: Retrospective analysis of patients diagnosed with solid tumor malignancies who underwent chemotherapy from August 2014 to October 2015 was performed to establish baseline rates of vaccination. Patients were identified using the WPAFB tumor registry system. Vaccination status was identified via the Aeromedical Services Information Management System. In October 2015, upon receiving pre-chemotherapy education in the clinic, patients were given a letter which detailed the NCCN vaccination recommendations and were advised to obtain them. Review of the data after the initial intervention showed an increase in vaccinated patients, but still room for improvement. In October 2016, step two of the improvement process, patients were also given a blank vaccination card which recorded dates of administration as well as a dual prescription for both conjugated (PCV 13) and polysaccharide (PPSV 23) pneumococcal vaccinations. Compliance rates were reviewed and percentages were calculated and reviewed.

Results: 60 solid tumor patients were used as baseline rates prior to interventions. 182 patients were analyzed after step one (letter) and 28 patients were included after step two (letter+vaccine card+prescription). Comparing the patients after both interventions to the baseline patients, 52% vs 22% of patients had received influenza plus PCV 13 (p= 0.042), 24% vs 18% had received all three vaccines (p=0.001), and 10% vs 38% had no vaccines (p= 0.001), which meant that 90% of our patients had received at least one vaccine.

Conclusion: Prior to October 2015, the Hematology-Oncology Clinic at WPAFB was not optimally vaccinating solid tumor patients prior to initiation of chemotherapy. After the two-step process we started in the cancer clinic, we have vaccinated more patients, and therefore likely decreasing the rates of preventable infections. Although we were successful in increasing the number of patients vaccinated, continued surveillance and improvement is needed to further patient care.
Title: Validation Of RACE vs VAN Scores To Predict Large Vessel Occlusion in the Prehospital Setting

Authors: Mohamad Alhoda Mohamad Alahmad1, Victoria Calderon2, Osama Ziadat2, 1Department of Internal Medicine, Mercy St Vincent Medical Center, Toledo, OH, 2Department of Neurointervention, Mercy St Vincent Medical Center, Toledo, OH

Introduction: Stroke is the leading cause of disability and fifth leading cause of death in United States. Early identification of acute ischemic stroke (AIS) due to large vessel occlusion (LVO) is vital to provide timely thrombectomy treatment that has been shown to lead to improved clinical outcome. Two prehospital LVO screening tools are commonly used- the Rapid Arterial occlusion Evaluation (RACE) and the Vision, Aphasia and Neglect (VAN) scores. The purpose of this study is to compare RACE and VAN in identification of LVO in a community prehospital setting when used by trained personnel.

Methods: Mobile stroke unit (MSU), a special ambulance equipped with CT scan and run by neuro-trained team, is structured to identify, manage, and transport AIS patients in the field. All MSU personnel are formally trained in both RACE and VAN scores and performed these tools routinely unaware of the current study design. Patients were prospectively evaluated by the MSU nurse and VAN and RACE scores were calculated on the scene. LVO was defined as an occlusion in any of the following arteries: MCA M1, M2, PCA, ACA, ICA, or basilar artery, as identified by a computed tomography angiography (CTA), magnetic resonance angiogram (MRA), or digital subtraction angiography (DSA) taken within 24 hours of RACE or VAN. The MSU nurses were unaware of the vascular imaging Results:

Results: A total of 22 male and 25 female were identified initially. 34 evaluable patients were included in the final analysis, of those 34% of patients had RACE > 4 and 24% had positive VAN. By the gold standard definition, only 26% of patients had LVO. VAN scoring system had higher sensitivity (33% compared to 22%) and higher specificity (80% compared to 61%) in comparison to RACE tool.

Conclusion: In patients with suspected acute ischemic stroke; VAN had more favorable diagnostic test statistics for predicting LVO compared to RACE, although statistical significance cannot be determined with the small sample size. Further research is needed to determine the best LVO detection screening tool to be used by the first contact provider in the field.
Ohio-Research-Poster Finalist
Ilana M Schlam, MD

Title: The Use of the 4T’s Score as a Primary Screening Tool for Heparin Induced Thrombocytopenia

Authors: Ilana M Schlam MD (1), Sarah H Min MD (1), Eithan Orlev-Shitrit MD (1), Chelsea Beaton (1), Ping Wang PhD (2), Miguel A. Islas-Ohlmayer (3), 1, The Jewish Hospital of Cincinnati, Internal Medicine Department, 2, Cholesterol and Metabolism Center, The Jewish Hospital of Cincinnati, 3, OHC – Blood Cancer Center

Introduction: The aim of was study was to determine if the 4T’s score for heparin induced thrombocytopenia (HIT) has been used appropriately in our institution. The 4T’s score is a validated tool with a very high negative predictive value and by using it systematically we can decrease unnecessary laboratory testing, consequently decreasing costs and length of stay.

Methods: In 2015, 57 PF4 ELISA were ordered in our institution. T4’s score was determined retrospectively for each patient and patients were categorized into low, moderate and high pretest probability for HIT (low if ≤3, intermediate 4-5, and high if ≥6).

Results: 28 patients were male and 24 were female, and they were between 25 and 89 years of age. 7 (13.4%) did not receive heparin (in within 100 days), 11 (21.1%) received therapeutic doses of unfractionated heparin, 11 (21.1%) received prophylactic doses of unfractionated heparin, and 23 (44.2%) received prophylactic low molecular weight heparin. The 4T’s score was calculated for each patient; 40 were low, 10 intermediate and 2 high risk (76.9, 19.2, 3.8% respectively). All the patients had HIT screening antibodies ordered; only 10 (19.2%) were positive and from those only 1 (1.9%) was confirmed by the serotonin release assay. This confirmed patient had a 4T’s score of 6. Of the 7 tested patients who did not receive heparin, none had a positive screening test (all had 4T’s score of 1 or 2). By using the McNemar’s test for matched pairs, we found that a 4T’s score ≤2 predicted a negative HIT-screening test, with a NPV of 75% (p=0.0018). Ten of the patients received argatroban as treatment for HIT, 1 was high risk, 3 intermediate, and 6 low.

Conclusion: Only 12% of the patients had a 4T’s score ≥4 that would warrant laboratory work up, therefore we concluded that PF4 ELISA screening tests have been over utilized in our institution. The cost of PF4 ELISA in our institution is $251, 40 tests were ordered during 2015 in low risk patients ($10,040). Additionally, based on previous studies, the mean cost of argatroban treatment is $7,440, it was given to 6 patients without indications ($44,640). This unnecessary testing and exposure to medications as well as the increased cost could be avoided by calculating the 4T’s score before obtaining any further laboratory tests. We will continue to work with our staff to improve education on HIT and adequate diagnosis based on current guidelines, the next step is to build in the 4T’s score into our electronic medical record in order to make it more easily available for our physicians.

References

Title: Evaluating and Improving Compliance with Breast, Cervical, Lung, and Colorectal Cancer Screening in the Primary Care Setting

Authors: Ilana Schlam MD*, Samantha M. Buszek MD*, Matan R. Rothschild MD, Natasha Dhawan MD, Ali Huda MD, Amir Khan MD, Jason Douglas MD, The Jewish Hospital of Cincinnati

Introduction:

- Cancer is one of the leading causes of death in the United States, second only to heart disease. Early diagnosis leads to less aggressive treatment regimens and better outcomes for patients. This can be achieved with screening tests.
- The United States Preventive Task Force (USPTF) issue guidelines for cancer screening with different grades of recommendations.
- The aim of this study was to assess compliance and to improve cancer-screening rates in the primary care setting in an internal medicine resident clinic.

Methods:

- 553 patients were identified and their compliance rates for age indicated cancer screenings, according to USPTF guidelines, were recorded on a retrospective chart review to determine rates of compliance with current recommendations (Grade A and B, Table 1).
- Over a six-month period educational interventions were implemented. Continued education was the main intervention method used in this project. Residents were also encouraged to add a "Health Maintenance" section to their progress notes and to provide clear documentation of completed screenings and when further screenings are required.
- After six months, a second retrospective chart review was done to determine post-intervention compliance rates.

Results:

- Statistical Analyses were completed using Chi-squared and Fisher’s-exact tests. These showed improvement in screening for breast, colorectal, cervical and lung cancer (Table 2, Figure 1) with statistically significant improvement in breast, colorectal, and cervical cancer screenings.
- Mammogram compliance improved 29% (from 52% to 67%) after the intervention (p=0.006).
- Colonoscopy compliance increased by 29% (from 48% to 62%) (p=0.0002).
- Pap-smear compliance significantly increased by 56% (from 32% to 50%; p=0.0005).
- Lung cancer screening compliance was found to increase by 38% (from 26% to 36%), post-intervention; however these findings were not statistically significant.

Conclusion:

- Residents at The Jewish Hospital (TJH) were able to achieve compliance similar to national current compliance levels according to the Office of Disease Prevention and Health Promotion (ODPHP); for breast cancer (TJH 67% vs ODPHP 71.6%), for colon cancer (TJH 62% vs ODPHP...
62%); and for cervical cancer (TJH 50% vs ODPHP 81.2%) There are not reported rates for lung cancer.

- Unfortunately, patient willingness to complete screening exams was found to be one of the major barriers for resident cancer screening compliance. Therefore, patient education will need to be optimized in our subsequent studies.
- Simple and inexpensive methods of education were effective in increasing cancer-screening rates and had a significant impact in the care we provide for our patients.
Ohio-Research-Poster Finalist
Rajpreet Singh, MD

Title: LEADER (Learning Expedited through Audiovisual Directed Education by Residents)

Authors: Rajpreet Singh MD, Perry Lin MD, Lynn Shaffer PhD, Rishi Kora MD, Sagar Gandhi MD, Kelly Hanson DO, Diana Zellner DO, Karampal Mand MD, Jonathan Burton DO, Johonna Asquith MD

Introduction: Poor health literacy is more prevalent in patients with lower socioeconomic status and education, and contributes to unnecessary health care expense. Diabetic patients with limited health literacy have been shown to have an A1c 1% higher than those with proficient literacy. Residency run clinics see an above average percentage of both patients below the poverty level and those with diabetes, making them uniquely poised for literacy interventions. We developed an educational intervention to improve health literacy in our patients towards an ultimate goal of secondary prevention.

Methods: All patients with an outpatient visit at MetroWest Clinic 11/1/2026 – 1/15/2017 from November 1st to January 15th received a questionnaire at check in which covered diabetes-related knowledge, perception and self-care behavior topics. Patients with a chart diagnosis of diabetes also viewed an educational five-minute video developed by our team prior to interaction with a physician. After video education patients with diabetes answered the same survey prior to leaving clinic. Their pre- and post-video answers were compared using McNemar’s test.

Results: 296 unique patients visited the clinic and completed the survey; of these 51 (17.2%) had diabetes. Three of four knowledge questions showed improvement among patients with diabetes, including: 1) use of A1c for screening (78% to 94%, p=0.0047); 2) A1c target of 6 (39% to 59%, p=0.0075); and 3) A1c should be checked every 3 months (41% to 63%, p=0.0076). Additionally, 27.4% of patients with diabetes either did not check blood sugar at home or had daily readings >200. Only 21.57% of the diabetic patients exercised daily, and 21.57% that never exercise. 18% of patients without diabetes stated they would like more information about diabetes while 9.8% of patients with diabetes agreed with this statement.

Conclusion: Patients both with and without diabetes have poor understanding of the disease process, screening, diagnosis and monitoring needed. Our intervention, albeit with a small sample size, has clearly shown that already-scheduled patient visits to the PCP can be used to provide health education with the goal towards improving outcomes and limit health care costs.
Oklahoma-Research-Poster Finalist
Taylor D Jenkins, MD

Title: Traumatized Residents: An Initial Look at PTSD in Internal Medicine Residents

Authors: Taylor D. Jenkins MD1, Theresa Jackson MD2, Jacob Murray DO1, Chelsea Therrien MD1, Kristin Rodriguez MPH1, Martina Jelley MD, MSPH, FACP1, Michael Truitt MD3, 1Department of Internal Medicine, University of Oklahoma School of Community Medicine, 2Department of Surgery, University of Oklahoma School of Community Medicine, 3Department of Surgery, Methodist Dallas Medical Center

Introduction: Current interest in physician wellness and burnout has largely excluded post-traumatic stress disorder (PTSD). Originally recognized in military personnel, PTSD has been documented in police officers, trauma surgeons, and other first responders1. Although not thought as likely to occur in primary care physicians, little data exists. Given the negative effects of PTSD on wellness, we sought to examine the prevalence of PTSD in internal medicine (IM) residents in the United States (U.S.).

Methods: A cross-sectional online survey of U.S. IM residents was conducted from September 2016 to May 2017 and yielded 550 respondents following an emailed invitation to participate. At-risk participants were identified using the four-question Primary Care PTSD (PC-PTSD) screen. Demographic, occupational, and work satisfaction risk factors were also assessed; occupational and work satisfaction variables were developed by the originators of the study. Results: were analyzed using IBM SPSS v. 23.

Results: 466 IM residents representing all regions of the U.S. completed the PC-PTSD screen. 19.7% of respondents screened PTSD positive (at least 3 screening questions positive). There were no significant differences between demographic variables and screening positive for PTSD. IM residents who reported working 81 or more hours/week on average were more likely to screen PTSD positive than those who worked 80 hours/week or less (p<0.001). IM residents who reported duty hour violations once/week or more were more likely to screen PTSD positive (20.0%, p=0.001). Although the majority of respondents reported having poor work-life balance, only 17.1% of respondents indicated unhappiness with their careers. IM residents screening positive for PTSD were more likely to report poor work-life balance in 4 areas (p<0.001) than those not screening positive for PTSD. Additionally, IM residents who screened positive for PTSD were more likely to be dissatisfied with 9 measured workplace variables than those screening negative for PTSD (p≤0.001). Those who screened positive were also more likely to report feeling less satisfied with their jobs (85.1%, p<0.001) and unhappy with their careers (40.0%, p<0.001) than those who did not screen positive for PTSD.

Conclusion: Increasing national efforts to improve physician and resident wellness highlight the need for understanding PTSD in IM residents. In this national study, IM residents screened positive for PTSD at strikingly higher rates than the general population (19.7% compared to 1 - 6.8%)2-3. Multiple work-life and work-related factors were found to be associated with screening positive for PTSD. In addition, significant associations between positive PTSD screens and being at higher risk for components of burnout exist. Resident wellness improvement efforts need to recognize PTSD and its effects in resident physicians.

References

Title: Improving A1c above 9.0 in an academic Internal Medicine ambulatory clinic: A quality improvement project

Authors: Henry Le MD, Kevin Smith DO, Todd Thomas DO, Chelsea Therrien MD, Michael Cloud MD, Khanh Le MD, Mitchell Fahrenkrog MD, Brent Beasley MD, Kristin Rodriguez MPH LSSGB, Carmen Vesbianu MD

Introduction: Diabetes management can be challenging in the ambulatory academic setting due to multiple factors including the discontinuous nature of resident clinics and a patient population that experiences social disadvantages that frequently result in limited engagement in their health. In order to better serve our patients, we designed a quality improvement project aiming to decrease the percentage of patients with an HbA1c above 9.0 seen in the last year in the OU-Tulsa Internal Medicine practice by 50% within one year.

Methods: Patients were identified by using ICD-10 codes related to diabetes and narrowed down to those with an associated HbA1c of 9.0 or above. We performed a root cause analysis to identify some of the reasons patients have elevated HbA1c as well as generate ideas for improvement. Our project consisted of three PDSA cycles. The first PDSA cycle included a small sample of patients and consisted of calling patients and scheduling appointments for a standardized diabetes visit. Care managers and patient service representatives facilitated this cycle. For the second PDSA cycle, we created a diabetes check list to be used at a regular office visit in order to guide and standardize physician-patient interaction. For the third cycle, we added the diabetes standardized check list to the office visit note in the EMR, making it easily accessible to all providers in the practice.

Results: 39% of the patients diagnosed with diabetes and seen in the last year in the OU – Tulsa Internal Medicine practice had an HbA1c above 9% prior to the implementation of our project. The percentage decreased to 36% by the end of the project. We were not able to reach the 50% targeted reduction. The first cycle failed to get patients scheduled; only 10% of the patients targeted were seen for a “diabetes visit”. The second cycle yielded disappointing results as the diabetes checklist was utilized less than 20% of the time at a regular office visit. We have not been able to measure the “diabetes quick text” utilization to date.

Conclusion: Management of poorly controlled diabetes in our patient population can be very challenging. The creation of an easy-to-use check list can help clinicians target areas most fruitful in improving a patient’s HbA1c. We encountered multiple difficulties throughout our project life cycle. Learning from these challenges can help us improve our future QI projects, clinic practices, and ultimately patient care.
Title: Improving blood pressure control in an academic Internal Medicine clinic

Authors: Thao Tran DO, Mary Mao MD, Andy Nguyen DO, Arjun Ghodasara MD, Christina Molumby MD, John Gemma MD, Andrea Kellogg MD, John Carment MD, Kristin Rodriguez MPH LSSGB, Carmen Vesbianu MD

Introduction: High blood pressure is a well-established risk factor for stroke and heart disease (1). Treating hypertension in the primary care office is a complex process that involves a multidisciplinary team. Our quality improvement project aims to increase the percentage of patients seen in Internal Medicine Clinic whose blood pressure is adequately controlled from 64% to 75% by May 2017.

Methods: We identified patients 18 and older with an active diagnosis of hypertension and at least one visit in the last year. For our project, we defined uncontrolled blood pressure as that above 140/90. We chose this target to align with the CMS Clinical Quality Measures reports; however, in our clinic, hypertension is managed according to the JNC 8 recommendations (2).

We mapped out the current process for measuring and treating high blood pressure to identify areas for improvement and found three: sub-optimal blood pressure measurement techniques, physician inertia in high blood pressure management, and insufficient follow-up of patients with hypertension. Three PDSA cycles were designed based on these areas. To improve the measurement technique, we recommended procurement of electronic blood pressure monitor devices; these were purchased towards the end of our project and are now used throughout the clinic. Nurses were trained in proper blood pressure measurement techniques. To overcome physician inertia, we designed a “follow-up” protocol for residents to use. This protocol targets patients who had not achieved their target blood pressure and involves scheduling a visit with a nurse in 1-2 weeks after the last abnormal blood pressure reading. Additionally, patients without a diagnosis of hypertension but with new high blood pressure readings were managed with the “follow-up” protocol.

Results: In July 2016, 64% of patients 18 and older with diagnosis of HTN were found to have blood pressures below 140/90 and therefore considered to be in control. In May 2017, 65% of patients with hypertension had blood pressures below 140/90. The majority of the changes we designed were implemented towards the end of the project. Despite this, and likely due in part of administrative changes and increased nursing and physician awareness of the deficits in hypertension management, we were able to significantly increase the percentage of patients with hypertension who had blood pressures considered in control. We were not, however, able to reach our aim of 75% of hypertensive patients with controlled blood pressure. We have extended the duration of this project through January 2018 and hope to see substantial improvement by that time.

Conclusion: Improving blood pressure control requires a multidisciplinary approach. By designing a clinic hypertension protocol, we increased awareness of hypertension management and believe that in the upcoming months will be able to see a significant improvement in our outcome measurement.

References

Mediation Analysis with a Time-to-Event Outcome: a Review of Use and Reporting in Clinical Research

Authors: Lauren Lapointe-Shaw MD, Zachary Bouck MPH, Nicholas A. Howell MSc, Theis Lange PhD, Ani Orkanian MISt, Peter C. Austin PhD, Noah M. Ivers MD, Donald A. Redelmeier MD MSHSR, Chaim M Bell MD PhD, and others.

Introduction: Mediation analysis is used to explain the relationship between two variables by a third variable, the mediator. This method is increasingly utilized in healthcare research, where questions of how an exposure leads to an effect abound. Survival outcomes are of frequent clinical interest, yet mediation analysis with a time-to-event outcome poses some unique challenges. In particular, the popular Cox Proportional Hazards model is ill-suited to testing for or measuring a mediator effect using traditional methods. We sought to describe the usage and reporting of mediation analysis with time-to-event outcomes in published healthcare research.

Methods: We undertook a systematic search of Medline, Embase, and Web of Science for applications of mediation analysis to healthcare research involving a clinically relevant time-to-event outcome. We summarized usage over time and reporting of important methodological characteristics using counts and frequencies.

Results: Our search yielded 1,991 unique abstracts, of which 321 were selected for further review. Of these, 8 were excluded as they did not relate to human healthcare, 110 because they did not include mediation criteria, a test, or measurement of the indirect effect. Another 12 were excluded because they did not include a clinically meaningful outcome, and 41 because the outcome of mediation analysis was not time-to-event. Further, 1 full text could not be reviewed as it was in Arabic.

This left 149 eligible studies, published from 1997 to 2016. Most studies were published after 2011 (n=110, 74%), and the number of studies nearly doubled in the last year (from n=21 to n=40). A traditional approach (causal steps or change in coefficient) was most commonly taken (n=87, 58%), and the majority of studies (n=117, 79%) used a Cox Proportional Hazards regression to conduct mediation...
analysis. An estimate of the indirect effect was reported in 55 studies (37%) while proportion mediated was reported in 83 studies (56%). Most studies reporting an indirect effect size included a measure of uncertainty (n=53 of 55, 96%) around the estimate; however, only 36 studies (of 83, 43%) included a measure of uncertainty around the proportion mediated. Few studies (n=52, 35%) mentioned any of the assumptions or limitations fundamental to a causal interpretation of mediation analysis.

**Conclusion:** There is increasing use of mediation analysis with time-to-event outcomes in healthcare research. Current usage is limited by reliance on traditional methods and the Cox Proportional Hazards model, and low rates of reporting of underlying assumptions. In addition, reports of proportion mediated frequently lack a measure of precision around the estimate. There is a need for formal criteria to aid authors, reviewers, and readers reporting or appraising such studies.

**References**

Title: An investigatory Biofire sputum film array PCR panel improves rapid diagnostics in patients hospitalized with community acquired pneumonia (CAP).

Authors: Katie Hendrickson, MD, Gita Gelfer, DO, David Gilbert, MD, James Leggett, MD

Introduction: With standard diagnostic methods, the etiologic pathogen of community acquired pneumonia (CAP) is detected in ≤ 50% hospitalized CAP patients. In previous studies a diagnostic “bundle” detected etiologic pathogens in ≥ 70% of the patients. Our bundle consisted of nasopharyngeal swab for Biofire film array that detects 9 viral genera, nucleic acid amplification testing (NAAT) for *Streptococcus pneumoniae* (*S. pneumo*) and *Staph. Aureus*, urine antigens for *Legionella* and *S. pneumo*, and cultures of sputum and blood. The current study is designed to determine if a next generation PCR Biofire Multiplex Investigational Sputum Pneumonia Panel (BiPP) improves detection of candidate pathogens.

Methods: Starting January 2017 we compared the standard bundle to BiPP. Herein we report Results: in the first 120 patients admitted with CAP. The BiPP requires sputum or a sputum equivalent. The panel includes probes for 9 viral Genera, 18 bacterial pathogens, and 7 antibiotic resistance genes. Serum procalcitonin (PCT) levels were used to separate bacterial colonization from invasion.

Results: Of the first 120 patients 64 were evaluable. Pts. With inadequate data collection or an alternative diagnosis were non-evaluable. Influenza was detected in 19 of the 64 (29.7%) patients with the standard bundle and 29 of 64 (45.3%) patients with the BiPP, p < 0.07. Similarly, 38 potential bacterial pathogens were detected with the standard bundle vs 52 with the addition of BiPP. Detection of both potential bacterial and viral pathogens occurred in 19 of 64 (29.7%) patients with the standard bundle as compared to 41 of 64 (64.1%) patients with the BiPP, p < .0.0001.

The median PCT level in 8 virus only patients was 0.07 ng/ml. In comparison there were 14 patients wherein BiPP detected a mixture of virus plus bacteria (V+B) but the median PCT was 0.09, similar to the virus only patients, p 0.70. In contrast, 23 patients had V + B detection with a PCT median value of 3.4, which was significantly higher than virus only patients, p 0.001, and similar to bacterial only patients, p 0.6. Hence, if a concomitant PCT was < 0.25 ng/ml, the detected bacteria were interpreted as colonizing and not invading. Overall the standard bundle detected a pathogen in 84% of evaluated patients as compared to 98% when BiPP Results: were included.

Conclusion: With a rapid diagnostic bundle, a candidate etiologic pathogen can be detected in 80% of the patients with CAP and using the BiPP platform, the detection rate increases to 98%. Moreover, PCT levels successfully adjudicated whether detected bacteria were colonizing or invading. Thus, the increase in rapid pathogen detection coupled with PCT levels offers great potential for improvements in antibiotic stewardship, including reduction in overuse of antibiotics and allowing for targeted treatment in patients with CAP.
Oregon-Research-Poster Finalist
Hayden Oldham, MD

Title: A Breath of Fresh Care: Increasing Referrals for Pulmonary Rehabilitation after Admission for Acute Exacerbation of COPD

Authors: Hayden Oldham, MD; Jacob Luty, MD; Brady Wright, MD; Blake Lesselroth, MD

Introduction: Hospitalizations related to ambulatory care sensitivity conditions (ACSCs), a group of medical conditions that include chronic obstructive pulmonary disease (COPD), are preventable if high-quality outpatient care is provided. At the Portland Veterans Affairs Health Care System (PORVAHCS), admissions for acute exacerbation of COPD (AE-COPD) are significant higher than expected. Further, readmissions account for a large portion of these AE-COPD admissions. Evidence suggests that pulmonary rehabilitation, when started within 28 days of discharge, can significantly reduce readmission rates. An analysis at the PORVAHCS revealed that this intervention was rarely offered at discharge after an AE-COPD admission. The aim of this project is to decrease readmission rates of veterans admitted for AE-COPD by increasing the number who start pulmonary rehabilitation within four weeks of discharge.

Methods: To understand why pulmonary rehab was not consistently utilized at PORVAHCS, we performed process mapping via user observations and identified significant gaps in the referral process. To address these gaps, we first created a standardized referral pathway for pulmonary rehab by building a consult order and template in the computerized patient record system (CPRS). We then inserted decision support into the physician discharge workflow to remind the discharging provider to order pulmonary rehabilitation when indicated. Finally, we developed and distributed educational materials to inform providers of the system changes. Tracking mechanisms in CPRS along with the VA’s corporate data warehouse were used to monitor use of the decision support and the consult order, as well as readmission rates for AE-COPD. Chart reviews were necessary to determine if veterans actually started pulmonary rehab within the 28-day period.

Results: After three months, the percentage of AE-COPD admissions with pulmonary rehab ordered at discharge has increased 0% to 19%, and patients who actually start pulmonary rehab within 28 days increased from 0% to 12.5%. Changes in AE-COPD admission and readmission rates were difficult to assess due to seasonal variation but remained essentially unchanged.

Conclusion: Although our primary goal of improving the ACSC quality indicator is not yet achieved, we demonstrate that the introduction of a standardized process for ordering pulmonary rehab along with decision support and user education resulted in a significant increase in number of veterans that receive this evidence-based intervention. Future quality improvement cycles involving workflow optimization and decision support tweaks may further increase the number of patients who are referred and start pulmonary rehab within 28 days and may ultimately yield fewer AE-COPD readmissions.
Title: Implementation of a Delirium Prevention Program – Barriers and Successes

Authors: Ben Pedroja, MD, Lisa Sanders, MD, Tricia James, MD

Introduction: A wealth of evidence has accumulated to support comprehensive non-pharmacologic interventions in the prevention of delirium in hospitalized patients. Implementation can prove difficult, however, especially in community-based hospitals that often lack the resources and specialized staff to advance such a program. We developed a resident-led quality improvement project with the goal of demonstrating the impact of delirium prevention in a small high-risk population in effort to fuel broader institutional investment.

Methods: Our population included all patients aged 70 and above admitted to our orthopedic unit with an acute fracture, a population that is well known to be at exceptionally high risk for delirium. The project included four main components. Our first aim was to establish a process for measuring delirium (CAM assessments) in order to establish a baseline rate in our hospital and track our progress going forward. Second was to involve our clinical pharmacists to conduct medication review and provide recommendations to the treatment team. Third was to recruit and train “goodnight volunteers” to assist nursing with patients going to sleep. And fourth was to develop nursing protocols and a physician order-set with an eye toward hospital-wide and system-wide implementation.

Results: We encountered many of the common barriers to quality improvement projects, including obtaining stakeholder buy in, lack of project-specific staff, multidisciplinary engagement, and the difficulties of aligning with system-wide priorities. Our most immediate challenge was engaging the nursing staff to incorporate CAM assessments into their routine intake protocols. Leadership was at first resistant to asking more of their nurses, but by identifying and working closely with nursing champions, and emphasizing education and patient safety, we were able to improve our CAM assessment rate to >80%. CAMs are now regularly discussed during safety huddles and our estimated in-hospital delirium incidence is ~30-40%, in line with published estimates. We also struggled initially to find enough volunteers, in part due to a lack of awareness about the problem. With the help of our volunteer coordinator we now have nightly volunteers that have been well received by patients and care providers. Pharmacists are entering timely recommendations in our EMR, but the response rate by the treatment teams has been inconsistent. We’re currently exploring ways to improve communication. While slow to develop, discussions are ongoing at the system level to establish formalized nursing protocols and a physician order set targeted at delirium prevention.

Conclusion: This QI project successfully piloted a delirium prevention program at a community hospital and may help pave the way toward hospital-wide and system-wide adoption.
Title: A Quality Improvement Intervention: A high-value Approach to Heparin-Induced Thrombocytopenia

Authors: Fnu Aparna, Jian Liang Tan, Andrew Kunkel, Fnu Anshul, Naveed Jan, Kshitij Thakur

Introduction: Heparin-Induced Thrombocytopenia (HIT) is an antibody-mediated pro-thrombotic disorder which occurs after exposure to heparin product. Diagnosis of HIT type II is rather challenging as affected individual often has other causes of thrombocytopenia and a delayed discontinuation of heparin in HIT type II is associated with a mortality rate of 20-30%. 4Ts score is a validated screening tool with a high negative predictive value in excluding HIT type II when a low probability score is present. Hence, this clinical scoring system (4Ts) was adapted by our hospital in November 2014 as an effort to reduce the number of inappropriate HIT antibody testing. The aim of this study was to assess the outcomes of pre- and post-implementation of the 4Ts scoring system.

Methods: We conducted a retrospective chart review of patients who had HIT antibody tests ordered. There was a pre-intervention phase (January 2013 – June 2014) which served as a control, and a post-intervention phase (August 2015 – February 2017) which served as a comparison. Pre-intervention: A total of 154 patients had the HIT antibody testing ordered prior to the hardwiring of the 4Ts scoring system in the EMR. Then, there was an extensive in-house education provided to the healthcare providers regarding the need of completing 4Ts score prior to ordering the HIT antibody tests over the 13 months period. Post-intervention: A total of 103 patients had the HIT antibody testing ordered after the 4Ts scoring system was created in the EMR. Inclusion criteria: Patients who were suspected to have HIT type II and HIT antibody tests sent out. Exclusion criteria: Age under 18-year-old. The number of 4Ts scores documented and the appropriateness of HIT antibody tests ordered between pre- and post-intervention were compared.

Results: Pre-intervention, only 1.3% (2/154 patients) had their 4Ts score documented in the chart, which significantly improved to 34% (35/103 patients) post-intervention [1.3% vs 34%, p < 0.00001]. Pre-intervention, 62.3 % (96/154 patients) with low probability score had unnecessary HIT antibody testing ordered, which significantly reduced to 40.8% (42/103 patients) post-intervention [62.3% vs 40.8%, p = 0.00068]. The total average cost of HIT antibody testing per patient was around $5,000. As the test takes 3 – 5 days to return, the average length of stay in hospital ($7,000/day) will inadvertently be prolonged. Hence, if the physician complies with the use of 4Ts score, an estimated cost savings per patient is between $26,000 - $40,000.

Conclusion: It is evident that the implementation of the 4Ts scoring system in EMR had significantly reduced the number of inappropriate HIT antibody testing in low probability group. Clinicians are encouraged to utilize the 4T scoring system prior to sending HIT panels to reduce unnecessary testing and practice cost-effective medicine.

References

Title: Outcome of Inherited Thrombophilia in Patients Admitted with Acute Coronary Syndrome

Authors: Sijan Basnet, Rashmi Dhital, Biswaraj Tharu, Dilli Ram Poudel, Anthony Donato

Introduction: According to the American Heart Association update in 2017, an estimated 695,000 will have a new coronary event and 320,000 will have a recurrent coronary event. Many coronary risk factors such as age, cholesterol, systolic blood pressure, smoking, etc. have been associated with coronary events. However, studies demonstrating role of inherited thrombophilia in overall outcome of patients admitted with acute coronary syndrome is limited. It was our aim to explore this relationship.

Methods: We utilized the National Inpatient Sample (NIS) databases from 2009 – 2011 to select non-pregnant adult hospitalizations (≥18 years) with ACS and history of IT. We chose patients with ACS diagnosis in first 2 positions and history of IT (cases) based on International Classification of disease-9 (ICD-9) and Clinical Classification Software-Diagnoses codes supplied by HCUP. The control group consisted of ACS patients without history of IT. We studied the differences in overall cost of hospitalization, length of stay (LOS) and mortality rate between our cases and controls. We used STATA version 13.0 (College Station, TX) for our analyses.

Results: Patient with history of IT who were admitted with ACS (cases) were younger and predominantly Caucasians. Obesity was more frequent in cases whereas diabetes mellitus, hypertension and dyslipidemia were more frequent in control group. Other comorbid factors such as chronic kidney disease and smoking were not significantly different between the two groups. Multivariable logistic regression failed to show difference in mortality between control and cases but showed greater cost and length of stay in cases.

Conclusion: In our study, patients with IT present with ACS at a younger age but carry the same mortality risk as the older, general population. They also have a longer hospital stay and greater cost of hospitalization. Inherited thrombophilia other than antiphospholipid syndrome do not increase risk for arterial thrombi. They have been linked with risk for venous thromboembolism. Identification of IT as an established risk factor is a matter of further longitudinal cohort studies.
Pennsylvania-Research-Poster Finalist
Ronan W Hsieh, MD

Title: Oral Fluoroquinolone and the Risk of Aortic Dissection and Aortic Aneurysm: A Population-Based Case-Crossover and Disease Risk Score Matched Case-Time-Control Study

Authors: Ronan Hsieh, M.D.¹, Meng-tse Gabriel Lee, Ph.D.², Lorenzo Porta, M.D.³, Wan-Chien Lee, M.S.², Sie-Huei Lee, M.D.⁴,⁵, Shy-Shin Chang, M.D., Ph.D.⁶, Chien-Chang Lee, M.D., Sc.D.², ¹Department of Medicine, Albert Einstein Medical Center, Philadelphia, PA, USA, ²Department of Emergency Medicine, National Taiwan University Hospital, Taipei, Taiwan, ³Dipartimento di scienze Biomediche e Cliniche, Ospedale "L. Sacco", Università degli Studi di Milano, Milan, Italy, ⁴Department of Rehabilitation and Physical Medicine, Taipei Veteran General Hospital, Taipei, Taiwan, ⁵Department of Medicine, National Yang Ming University, Taipei, Taiwan, ⁶Department of Family Medicine, Taipei Medical University Hospital and School of Medicine, Taipei Medical University, Taipei, Taiwan

Introduction: Fluoroquinolones have been increasingly prescribed despite continued uncertainty about their association with aortic aneurysm (AA) and aortic dissection (AD). Two recent studies showed a two to three-fold increased risk of AA/AD in fluoroquinolone treatment as compared to non-treatment (1, 2). However, limited by the study designs, previous studies could not completely obviate the possibility of residual confounding, since many confounders, such as lifestyles, were not registered in administrative databases. Therefore, we conducted a case-crossover and case-time-control study using a nationwide population-based database. A case-crossover design uses cases as their own controls to match out all intra-individual time-invariant covariates, minimizing the risk of bias due to unmeasured confounding. A disease-risk score matched case-time-control study not only addressed the time-varying factors but also provided a robust control to minimize the risk of time-trend bias, which was associated with the case only design (3).

Methods: We performed a case-crossover study using hospitalized AA/AD patients, identified from the Taiwan’s National Health Insurance Research Database. Exposure to fluoroquinolones was assumed whenever there was a fluoroquinolone prescription filled for three or more days during the predefined 60-days exposure window. We determined the association between fluoroquinolone usage and AA/AD by a conditional logistic regression model comparing case and control periods. Potential time-varying confounding variables were adjusted by multivariate analysis. In addition, potential temporal-changes in fluoroquinolone exposure were investigated using a disease-risk score (DRS) matched time control design, where each case was matched 1:1 to the general population by DRS. In the sensitivity analysis, we used different exposure periods from 60 to either 120 or 180 days.

Results: We identified a total of 1,213 patients with AA or AD. In the case-crossover analysis, use of fluoroquinolone was associated with a substantial increase of the risk of AA/AD before (OR, 2.52; 95% CI, 1.44,4.44) and after multivariate adjustment of potential time-varying confounders (OR, 2.05; 95% CI, 1.13,3.71). An increased risk of AA/AD was observed for 3-14 days of fluoroquinolone use (OR, 2.41; 95% CI, 1.25,4.65) and >14 days of fluoroquinolone use (OR, 2.83; 95% CI, 1.06,7.57), when compared with <3 days of fluoroquinolone prescription. In the sensitivity analysis, we found that the risk of AA/AD was highest within the 60 days’ time period (OR, 6.33; 95% CI, 1.87,21.40), and there was no significant association for the two other time periods. The calculated number needed to harm (NNH) was 11,111. However, in the DRS-matched time control analysis, we did not find any significant association both before (OR, 0.98; 95% CI, 0.50,1.92) and after adjustment for potential time-varying confounders (OR, 0.83; 95% CI, 0.42,1.64).
Conclusion: Exposure to fluoroquinolone was substantially associated with AA/AD. This risk seemed to be modified by the duration of fluoroquinolone use, and the length of the exposure period.

References

Pennsylvania-Research-Poster Finalist
Amir Hossein Gougol, MD

Title: Association of Dietary Habits with Severe Acute Pancreatitis

Authors: Amir Gougol, Mohannad Dugum, Pedram Paragomi, Adam Slivka, Dhiraj Yadav, David C. Whitcomb, and Georgios I. Papachristou

Introduction: The revised Atlanta classification stratifies acute pancreatitis (AP) based on the development of local complications and/or organ failure into mild, moderate, and severe disease. The relation between diet and risk of AP has been suggested by prior studies, but the association of dietary habits with AP severity has not been previously evaluated.

Aim: Assess differences in severity of AP based on reported dietary habits.

Methods: A prospectively maintained cohort of patients with AP admitted to a tertiary medical center between 2008 and 2016 was utilized. A questionnaire with details on dietary habits was completed by interviewing enrolled subjects during their hospitalization. Patients were stratified into two groups: mild/moderate AP and severe AP. Dietary habits were categorized based on the overall type of diet, fruits/vegetables servings, fat content, diary consumption, and fluid intake. Multivariate analysis was used to determine whether dietary habits have an independent association with severity of AP. P-value \( \leq 0.05 \) was considered statistically significant.

Results: A total of 407 prospectively enrolled patients had available dietary habits questionnaires: 202 (49.6%) male, mean age was 51 years. Seventy-nine (19.4%) developed severe AP, 103 (25.3%) moderate AP, and 225 (55.3%) developed mild AP. No differences in etiology of AP were present between both groups. Patients who developed severe AP were more likely to consume more than few servings of meats per week (84.3% in severe AP versus 67.4% in moderate, versus 71% in mild AP, \( p=0.04 \)) and less likely to be vegetarian prior to the onset of AP. No differences in the diet fat content (\( p=0.59 \)), or dairy consumption (\( p=0.55 \)) were present between both groups. Multivariate analysis controlling for gender, age, race and BMI showed an independent association between diet type (odds ratio:2.30 (95% CI 1.14-5.09, \( p=0.03 \))) with the development of severe AP. Diet type is the only variable significant in predicting RAC severe adjusting for demographic. Spearman correlation coefficient among diet related questions showed that only diet fat and diet type is moderately correlated (\( \rho=0.35 \)).

Conclusion: A diet high in meats and low in vegetables associated with severe disease in patients with AP. These important findings require further evaluation and may be useful in patient counseling and risk stratification.

References

Pennsylvania-Research-Poster Finalist
Yinn Shaung Ooi, MD

Title: Delta QRS distinguishes Ito-mediated J waves from "Pseudo-J waves" produced by Conduction Delays on Body Surface ECGs.

Authors: Yinn Shaung Ooi, MD, Aman Khaji, MD, Catherine Prince, DO, Gan-Xin Yan, MD, PhD

Introduction: On a 12-lead body surface electrocardiogram (ECG), it is often difficult to definitively distinguish Ito-mediated J waves seen in J wave syndromes from QRS deflections mimicking J waves ("Pseudo-J wave") produced by delayed intraventricular conductions, such as those observed in right bundle branch block (RBBB) and intraventricular conduction delay (IVCD). We hypothesize that the measured difference between the maximum QRS duration (QRS including Ito-mediated J wave or "Pseudo-J wave") and the minimum QRS duration identified on a 12-lead tracing is significantly larger in Ito-mediated J waves, and can be used as a reliable tool to make the distinction. This theory was tested in the present study.

Methods: A retrospective analysis was performed on patients 18 years or older with one the following four ECG manifestations: Osborn wave associated with hypothermia, early repolarization pattern with J wave, and RBBB and IVCD with QRS deflections mimicking J wave in leads other than V1 to V3 that increased in amplitude with an acceleration of heart rate. All ECGs were assessed individually and the maximum and minimum discrete QRS deflections on 12-lead tracings, defined as QRSmax and QRSmin, were identified. The measured difference between QRSmax and QRSmin, designated by delta QRS, was calculated and compared across the studied populations.

Results: A total of 60 patients (mean age of 55.5 years) consisting of 15 patients in each arm were included in the study. QRSmax was significantly smaller in the IVCD group (P <0.0001), while QRSmin was significantly larger in the RBBB group (P <0.0001). These differences were in such that a significantly larger delta QRS was observed in the hypothermia and early repolarization groups, compared to the RBBB and IVCD groups (P <0.0001), with the following mean delta QRS, hypothermia 54.3±13.7 ms, early repolarization pattern 47.3±15.3 ms, RBBB 19.3±6.5 ms and IVCD 16.0±6.6 ms.

Conclusion: Delta QRS serves as a reliable ECG parameter to distinguish Ito-mediated J wave seen in J wave syndromes from “Pseudo-J wave” produced by delayed intraventricular conduction.

References


Pennsylvania-Research-Poster Finalist
Sneh P Pandey, MD,MBBS

Title: A Retrospective Analysis of Pulmonary Function Tests in Patients with COPD and Studying Their Association with COPD-associated Pulmonary Hypertension

Authors: Sneh Pandey, Smitha Krishnan, Sofiya Rehman, Shikha Gupta

Introduction: Pulmonary hypertension (PH) associated with chronic obstructive pulmonary disease (COPD) is a common type of PH. Presence of PH in COPD patients has significant clinical implications such as requirement for long-term oxygen therapy, increased mortality, and limitations in physical activity. Hyperinflation due to destruction of alveoli in COPD causes reduction of area available for oxygen exchange and therefore should be expected to cause hypoxemia. Hypoxic vasoconstriction has been vastly accepted as the most important mechanism leading to development of PH in COPD patients. We hypothesized that patients with more hyperinflation will have more severe elevations of mean-pulmonary-artery pressure (mPAP) regardless of the degree of obstruction.

Methods: A retrospective clinical chart review of the patients with COPD patients listed for lung transplantation was done and patient demographics, pulmonary function tests (PFTs), and RHC values were collected. K-means cluster analysis was done to classify the mPAP in to 3 clusters. FEV1% predicted, DLCO% predicted and RV/TLC were the three main PFT variables which had significant differences between three clusters.

Results: Our study included 177 patients with age range: 33-77 (median 63), of which 55% were males and 45% females. The mean mPAP in our study was 17, however median was 26. The prevalence of PH was 52.5 %. Three major means of mPAP were observed with most values clustering around them. On the basis of those means, the patient population was divided into three groups in order of increasing mPAP as shown below. No significant difference between LVEDP occurred between these 3 groups, however, PaO2 was lower (p<0.05) in group 2 vs group 1.

<table>
<thead>
<tr>
<th>Groups</th>
<th>mPAP</th>
<th>FEV1%</th>
<th>DLCO%</th>
<th>RV/TLC</th>
</tr>
</thead>
<tbody>
<tr>
<td>No PH-Group 1</td>
<td>Mean</td>
<td>23.07692</td>
<td>28.84615</td>
<td>50.5</td>
</tr>
<tr>
<td>SD</td>
<td>5.098428</td>
<td>7.019258</td>
<td>11.90403</td>
<td>12.20435</td>
</tr>
<tr>
<td>Median</td>
<td>23</td>
<td>29.5</td>
<td>48</td>
<td>45</td>
</tr>
<tr>
<td>Min</td>
<td>14</td>
<td>11</td>
<td>30</td>
<td>25.54257</td>
</tr>
<tr>
<td>Max</td>
<td>34</td>
<td>44</td>
<td>75</td>
<td>64.64471</td>
</tr>
<tr>
<td>Mild PH-Group 2</td>
<td>Mean</td>
<td>26.70787</td>
<td>21.58067</td>
<td>32.24719</td>
</tr>
<tr>
<td>SD</td>
<td>7.081028</td>
<td>5.8116</td>
<td>8.810222</td>
<td>10.1212</td>
</tr>
<tr>
<td>Median</td>
<td>26</td>
<td>22</td>
<td>33</td>
<td>58.5</td>
</tr>
<tr>
<td>Min</td>
<td>15</td>
<td>10</td>
<td>13</td>
<td>36</td>
</tr>
<tr>
<td>Max</td>
<td>57</td>
<td>35</td>
<td>52</td>
<td>73.34218</td>
</tr>
<tr>
<td>Moderate to Severe PH- Group 3</td>
<td>Mean</td>
<td>30.3913</td>
<td>64.08696</td>
<td>34.26087</td>
</tr>
<tr>
<td>SD</td>
<td>10.61619</td>
<td>15.23427</td>
<td>10.88877</td>
<td>9.184623</td>
</tr>
<tr>
<td>Median</td>
<td>32</td>
<td>60</td>
<td>31</td>
<td>72</td>
</tr>
<tr>
<td>Min</td>
<td>15</td>
<td>42</td>
<td>21</td>
<td>36</td>
</tr>
</tbody>
</table>
Conclusion: Based on above findings, it appears that hyperinflation and obstruction might occur through independent pathological processes and the process causing hyperinflation is the primary pathology leading to development of PH. Commonly used anti-inflammatory therapies such as inhaled steroids don't have significant role in preventing hyperinflation and further research into hyperinflation specific therapies and their role in prevention and reversal of COPD associated PH is warranted.
Pennsylvania-Research-Poster Finalist
Sneh P Pandey, MD

Title: Evaluating the Role of Echocardiography in Detecting Severe Pulmonary Hypertension in COPD Patients

Authors: Sneh Pandey, MD; Smitha Krishnan, MD; Sofiya Rehman, MD; Vikas Singh, MD, Shikha Gupta, MD

Introduction: About half of the COPD patients listed for transplant have mean pulmonary arterial pressure (mPAP) ≥25 mm Hg as determined by right heart catheterization (RHC). Most COPD patients have mild to moderate pulmonary hypertension (PH) (mPAP 25-35). However, about 5-10% of them have mPAP >40 mm Hg, described as severe PH and they might benefit from PH specific intervention and could be candidates for early transplant. RHC is an invasive procedure with risks of complications thus necessitating a screening tool. Echocardiography is a simple and non-invasive tool which ascertains pulmonary artery systolic pressure (E-PASP) which is used as surrogate for mPAP. However, the role of echo in detecting COPD associated severe PH is still not clearly defined. The aim of the study is to evaluate echo as a screening test to detect severe PH in COPD patients.

Methods: A retrospective chart review of the patients with COPD listed for lung transplantation was done to tabulate demographics, echocardiography, pulmonary function tests (PFTs), and RHC values. Using Pearson’s correlation, relationship between echo derived pulmonary artery systolic pressure (E-PASP) and mPAP was computed. We plotted receiver operating characteristic (ROC) curve and using ROC curve and table, we chose cut-off of E-PASP>=50 for severe PH which we defined as mPAP>=40.

Results: We studied 177 patients with age range 33-77 (median 63) of which 55% were males and 45% females. PH was found in 52.5% patients while severe PH was noted in 7.8% patients. The correlation co-efficient between E-PASP and mPAP was 0.595 (p<0.0001). The area under ROC curve for E-PASP was 0.89 for detection of severe PH. We compounded a efficacy table listing positives and negatives and evaluated E-PASP as a tool for evaluating PH in COPD patients:

<table>
<thead>
<tr>
<th>Statistic</th>
<th>Value</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>81.82%</td>
<td>48.22% to 97.72%</td>
</tr>
<tr>
<td>Specificity</td>
<td>82.81%</td>
<td>75.14% to 88.90%</td>
</tr>
<tr>
<td>Positive Likelihood Ratio</td>
<td>4.76</td>
<td>2.97 to 7.63</td>
</tr>
<tr>
<td>Negative Likelihood Ratio</td>
<td>0.22</td>
<td>0.06 to 0.77</td>
</tr>
<tr>
<td>Positive Predictive Value</td>
<td>29.03%</td>
<td>14.22% to 48.04%</td>
</tr>
<tr>
<td>Negative Predictive Value</td>
<td>98.15%</td>
<td>93.47% to 99.77%</td>
</tr>
</tbody>
</table>

Conclusion: PH in COPD patients. Although PASP>=50, appears a good screening test, it still needs to be validated in a larger and different dataset to further ascertain its accuracy but for now, we advocate avoiding RHC in patients with E-PASP <50.
Pennsylvania-Research-Poster Finalist
Robert Park, MD

Title: Review of the Safety and Efficacy of Echocardiographically Guided Pericardiocentesis: A Retrospective Single-Center Analysis, 2009 - 2016

Authors: Robert Park, MD; Michael Valentino, MD, Ph.D; Rajiv Kabadi, MD; Matthew DeCaro, MD*; Gregary Marhefka, MD*, *Both contributed equally. Thomas Jefferson University Health System.

Introduction: For almost four decades, percutaneous pericardiocentesis guided by two-dimensional echocardiography or echo-guided pericardiocentesis (EGP) has been employed in the management of pericardial effusions. Since its inception, it has become internationally recognized as a safe, effective, and time-conscious technique (1, 2). Our team sought to investigate the safety and efficacy of EGP at our institution given the relatively limited data of this modality’s safety profile in American healthcare centers over the past decade. While our investigation primarily focused on outcomes associated with EGP, we also identified the most common etiologies underlying pericardial effusions necessitating EGP at our institution.

Methods: We performed a retrospective review of electronic medical records of patients that underwent EGP at Thomas Jefferson University Hospital (Philadelphia, PA) between March 1, 2009 and July 31, 2016. We identified eligible patients using a database provided by the two cardiologists at our institution who performed EGP during the study period. Demographic and clinical parameters as well as procedural details were collected and analyzed. A successful pericardiocentesis was defined as entering the pericardial space and fluid drained either for analysis or achieving symptomatic relief or both. Outcomes of interest were minor and major complications related to EGP—the former describing those requiring monitoring or non-invasive management, and the latter necessitating a subsequent invasive intervention.

Results: Over the seven-year study period, EGP was performed 175 times on 174 patients (98 males and 76 females) with a mean age of 57.7 ± 15.2 years (range 1 week to 93 years). EGP was successful in 174 cases (99.4%) with complications occurring in six (3.4%). Minor complications such as transient arrhythmia and puncture site cellulitis occurred in 5 cases (2.8%). Only one major complication was identified (0.6%) involving post-procedural hypotension and ST-segment elevation with an unremarkable left heart catheterization. There were no procedure-related mortalities within 30 days. Interestingly, the most common etiologies underlying pericardial effusions in our cohort included malignancy (73 cases; 42%), complications from invasive cardiothoracic procedures (46 cases; 26.4%), and infection (17 cases; 9.8%). The remaining cases were attributed to rheumatologic disease, uremia, and idiopathic causes. Analysis of pericardial fluid obtained via EGP also led to new diagnoses of malignancy in 12 patients (6.9%).

Conclusion: Our data demonstrate the overall safety and efficacy of EGP performed at our hospital over a seven-year period. Our favorable outcomes are also in part due to experienced operators and their proficiency in EGP after performing a myriad of cases spanning many years. Ultimately, our findings are compatible with preexisting literature describing outcomes associated with EGP at other institutions, and further support its role in the routine management of pericardial effusions today.

References

Title: Improving Compliance with Continuous Positive Airway Pressure (CPAP)/Bi-Level Positive Airway Pressure (BPAP) in Patients with Obstructive Sleep Apnea (OSA): An Initial Step

Authors: Faeq Kukhon MD 1,3, Muhammad Ehtisham MD 1,3, Ahmed Mohamed MD 1,3, Fatima Zeba MD 1,3, Maen Assali MD 1,3, Taro Minami MD FCCP 2,3, 1Department of Medicine, Memorial Hospital of Rhode Island, Pawtucket, RI., 2Division of Pulmonary and Sleep Medicine, Memorial Hospital of Rhode Island, Pawtucket, RI., 3The Warren Alpert Medical School of Brown University, Providence, RI.

Introduction: Compliance with CPAP/BPAP is an essential part of treatment success in patients with OSA. However, not all OSA patients are compliant with CPAP/BPAP. Many interventions had been proposed to improve CPAP/BPAP compliance. We designed a quality improvement (QI) project aiming to improve CPAP/BPAP compliance in patients with OSA. As an initial step, and before applying interventions on noncompliant OSA patients, we started our QI project by trying to identify possible factors that could be associated with poor compliance with CPAP/BPAP to help in identifying patients at-risk for being noncompliant.

Methods: This QI project was started in a community-based teaching hospital and as a follow-up to a previously presented project. OSA patients were screened through outpatient electronic medical records. Identified patients were those who visited our Sleep Clinic from December 2016 to July 2017. Patients were included for the following criteria: (1) Age of 18 years or older; (2) Established diagnosis of OSA or mixed OSA and central sleep apnea; (3) Use of either CPAP or BPAP; (4) Available compliance report. Patients were excluded if: (1) There was no compliance report available; or, (2) There was no sleep study report to confirm the diagnosis. Compliance was defined as CPAP/BPAP usage for at least four hours or more (≥ four-hour compliance) for 70% of the time or more. Examined demographic and clinical characteristics were: age, gender, body-mass index (BMI), sleep apnea type (OSA or mixed), apnea hypopnea index (AHI), and comorbidities (diabetes mellitus [DM], hypertension, obesity, atrial fibrillation [AF], and stroke). Two-tailed t-test was used to test the association between ≥ 4-hour compliance and continuous variables. The association between ≥ 4-hour compliance and categorical variables was tested using Chi-square test.

Results: Out of 675 initially-screened patients, 100 patients were included. The mean age of the patients was 59.8 years. 56% of the patients were male and 43% were female. Mean BMI was 36.6. 57% of the patients had hypertension, 36% had DM, 79% had obesity, 10% had stroke, and 16% had AF. OSA was identified in 80% and the rest had mixed type. 61% of the patients were compliant. There was a strong trend, though not a statistically significant association, between having DM and being non-compliant (p=0.0538). Interestingly, there was a significant association between having AF and being compliant (p=0.0238). The rest of the variables, including hypertension, did not have significant association with CPAP/BPAP compliance.

Conclusion: AF is associated with good CPAP/BPAP compliance. On the other side, there was a strong trend between DM and non-compliance with CPAP/BPAP treatment in OSA patients. Those Results: would make it easier to identify OSA patients at-risk of being non-compliant. Identification of such patients will help in targeting them with the appropriate interventions to improve their CPAP/BPAP compliance.

References

Title: Utilization of Prediction Models for Pulmonary Embolism in a Community E.D.

Authors: Edward G Medeiros, Jr. D.O and Alisa Merolli, MD

Introduction: Pulmonary embolism (PE) remains a challenging diagnosis given lack of specific signs and symptoms. In 2015, the American College of Physicians (ACP) released clinical practice guidelines for PE diagnosis. Using the prospectively validated Wells’ Criteria, patients are stratified into Low risk, Moderate risk, or High risk groups. Low risk patients are subdivided using the “Pulmonary Embolism Rule-out Criteria” (PERC); “PERC negative” patients require no further workup, while those meeting 1+ PERC and all moderate risk patients should have d-dimer testing. Individuals with elevated d-dimer subsequently undergo diagnostic imaging, such as CT angiography. High risk patients have diagnostic imaging without laboratory testing. The American College of Emergency Physicians (ACEP) also support a “two step” Wells’ Criteria. Patients in the “PE unlikely” group use PERC and d-dimer, while “PE likely” patients undergo imaging. Our hypothesis suggests the increased availability of CT imaging has led to reduced Wells’ Criteria utilization and inappropriate CT imaging. This retrospective chart review in an Emergency Department (ED) setting evaluated utilization of Wells’ Criteria and CT imaging appropriateness for suspected PE based on the aforementioned risk model.

Methods: A retrospective chart review was conducted in a community hospital ED. We selected patients whom underwent CT Angiography of the Chest for suspected acute pulmonary embolism. Patient encounters ranged from 01/01/14 – 12/31/14. Wells’ Criteria was retrospectively applied, with each patient placed into Low risk, Moderate risk, or High risk groups. Moderate risk patients were further stratified based on the “two-step” Wells’ Criteria. CT scan appropriateness and Wells’ Criteria utilization were determined based on each patient’s diagnostic workup. CT scans performed for indications other than acute PE were excluded from our study.

Results: 369 CT scans were included, with 159 of the scans (43.1%) determined to be inappropriate based on Wells’ Criteria. Inappropriate CT scans in each population included 109 of 216 (50.5%) in Low risk patients, 50 of 120 (41.7%) in the Moderate risk category and 0 of 33 (0%) High risk. An additional 25 CT scans were deemed inappropriate using the more recently studied Age-Adjusted D-Dimer criteria. Additionally, Wells’ Criteria was documented in only 164 of 369 (44.4%) patient encounters, which included any documentation of risk stratification. CT scan was positive for pulmonary embolism in 7 of 369 (1.9%) cases.

Conclusion: Despite supporting evidence, Wells’ Criteria remains underutilized in the workup of pulmonary embolism. While research shows increasing rates of CT imaging, our study suggests it may be associated with inappropriate usage. Wells’ Criteria was not documented in more than half of the cases, supporting our hypothesis of underutilization. Increasing the usage of previously validated prediction models may lead to more appropriate diagnostic testing, limiting the unnecessary complications and healthcare costs associated with CT imaging.
References


Saudi Arabia—Research—Poster Finalist
Hussam Alhamidi, MBBS

Title: The Direct Oral Anticoagulants in Antiphospholipid Syndrome

Authors: Hussam Alhamidi, Anjad AlJurais, Esra Abdullatif, Thara alWathnani, Nada AlShraim, Ohuod Alarfaj, Reem Bahmaid, Mukhtar Alomar Dr. Mohammed Al Sheef

Introduction: Antiphospholipid Syndrome (APS) is an enigmatic and rare autoimmune disorder of unknown etiology that may occur as an isolated disorder (primary APS) or in association with another autoimmune disorder such as systemic lupus erythematosus (secondary APS). APS is defined by clinical manifestations that include thrombosis and/or fetal loss or pregnancy morbidity in patients with antiphospholipid antibodies (aPL).

Methods: Study design: a retrospective cohort study. This study will be conducted in single tertiary care hospital, thrombosis clinic at department of internal medicine, King Fahd medical city (KFMC) Riyadh, Saudi Arabia.

Study Subjects: All APS patients who were objectively diagnosed to have venous or arterial thrombosis by Doppler ultrasonography, computed tomography or magnetic resonance result and started on anticoagulant treatment. Patients will be considered to have APS if met (Modified Sapporo criteria). We will recruit all patients who fulfill the study criteria from March 2010 until March 2016.

Inclusion criteria: Adult patients with age >= 18 years, who has APS (venous or arterial thrombosis) and were started on DOACs during the defined study period.

Exclusion: All patients who do not meet the inclusion criteria.

Sample size: 73 patients.

Data Collection Methods: Instrument Used, and Measurements: It will be through a comprehensive case report form.

Data Management and Analysis Plan: Data will be entered and analyzed using SPSS version 19. The analysis will consist mainly of descriptive analysis.

Results: In our study we investigated 73 patients diagnosed with antiphospholipid syndrome. The age of diagnosis was between 26-40 (52%). 83.5% was female gender and 97.2 % of patients had no family history. 67% of the cases were diagnosed in the form of DVT while the rest had arterial or unusual site venous thrombosis. 43% of patients have systemic lupus disease. Thrombosis was unprovoked in 90% of the cases. There is 20.5% of the cases had single positive aPL profile, 17.8% had double positive and 49.3 had triple positive aPL. 68.5% of the cases treated with warfarin while 31.5% of the cases were warfarin switchers to rivaroxaban. Safety profile was monitored after initiation of anticoagulant. 15% of patients had major and clinically relevant non-major bleeding (2 patients had major bleeding in the warfarin group and 4 patients in the rivaroxaban group). Recurrence of thrombosis (arterial and venous) was 43% with rivaroxaban and 30% with warfarin. Duration from starting Rivaroxaban to development of complication (bleeding and Thromboembolic events) was mostly after 1-2 years in (70%).
Conclusion: There is a strong association between thrombosis and a full positive aPL profile (i.e. triple positivity) in our study. Rivaroxaban should be used with extreme caution in APS patients in particular with full positive aPL profile and arterial thrombosis

References

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Title: Beyond Bates: The Advanced Physical Exam Series

Authors: Ryan Dean, MD; Steven Connelly, MD, FACP, FHM

Introduction: The physical exam is often taught in the first two years of medical school without much clinical context. As a result, students struggle with how to use maneuvers for diagnosis later in their careers. The purpose of this project was to develop a curriculum for medical students designed to teach proper technique and choice of both routine and advanced physical exam maneuvers as they relate to medical decision making in clinical practice.

Methods: We created a pilot course in partnership with the University of South Carolina School of Medicine – Greenville with 4th year medical students matching into clinical specialties. The two week course used a combination of Attending Physician and Resident-led didactics, student-led journal discussions, bedside ultrasound, and bedside physical exam rounds for direct application of skills.

Results: Five 4th year medical students matching into Internal Medicine, Family Medicine, and Emergency Medicine enrolled in the pilot course. Articles from JAMA: The Rational Clinical Exam and chapters from Evidence-Based Physical Diagnoses, by Steven McGee, MD were used to develop the curriculum and didactics. The curriculum focused on topics most relevant to Internal Medicine including cardiac, pulmonary, abdominal, neurologic, and endocrinologic exams. Both faculty and residents in the Internal Medicine and Internal Medicine-Pediatrics Departments were recruited to facilitate all portions of the courses. A 21 question pre-course assessment and a 20 question post-course assessment were administered.

Pre-course assessment average: 12.4 out of 21 points (59.1 ± 12.9%)

Post-course assessment average: 16.8 out of 20 points (84.0 ± 11.4%)

All students demonstrated an improvement in their percent score by an average of 25 ± 10.6%. A final oral case-based final assessment was also developed to assess skill and decision-making in four lifelike clinical situations using the Greenville Health System Simulation Center and trained professional patients. Grading was based on a four-point rubric in five different categories (40 available points).

Average oral score: 34.8 out of 40 points (87 ± 8.9%).

Constructive feedback on performance was also provided to the students. They were also asked to provide feedback for course. The data showed students found the afternoon physical exam rounds and the ultrasound didactics most helpful for learning. Students also requested more structured teaching on technique of exam maneuvers and high value patient care.

Conclusion: After participating in the advanced physical exam pilot course, students demonstrated improved knowledge of physical exam operating statistics and enhanced implementation of the physical exam for clinical decision making. They also reported greater comfort using the physical exam to help answer relevant clinical questions. Preliminary tools in data tracking were developed to monitor their improvement. Results from the pilot will be used to improve a follow-up course that has already been expanded to include 10 students with a growing waitlist.

References


Texas-Research-Poster Finalist
Yvette Achuo-Egbe, MD

Title: Fostering Wellness and Preventing Burnout Among Resident Physicians: A Pilot Study

Authors: Yvette Achuo-Egbe (ACP Associate), Elena Iglehart (ACP associate), Sam Hooper, Emmanuel Elueze (ACP Fellow), Department of Internal Medicine, University of Texas Health Science Center at Tyler / CHRISTUS Good Shepherd Health System, Longview, TX

Introduction: Maslach and colleagues defined burnout to encompass emotional exhaustion, cynicism and reduced personal accomplishment negatively affecting performance efficacy and effectiveness. Meta-analyses have shown higher suicide rates with male and female physicians at 40% and 130% higher risk respectively, compared to gender and age-matched individuals in the general population. The Accreditation Council for Graduate Medical Education in 2015 emphasized the importance of wellbeing by adding “Physician Well-Being” to its core initiatives. The Wellness Program was implemented at this residency program in 2016 to raise awareness and organize activities to promote resident physician wellness and decrease burnout.

Methods: A pilot prospective cohort study with 42 residents from the UT Health Northeast/CHRISTUS Good Shepherd Internal Medicine Residency program over a 7-month span to improve resident wellbeing. First, the residents anonymously completed the National Center for Health Statistics General Well-Being Scale (GWBS) questionnaire at 3 different time intervals. The GWBS has a total of 110 points ranging from “severe” to “positive wellbeing.” Secondly, a series of 4 lectures with applicable lifestyle changes were given that addressed each of the wellness domains: Diet and Nutrition, Activity and Exercise, Social Health, and Mental Health. Thirdly, residents got involved in appropriate community social functions and volunteering at a free clinic. Lastly, residents obtained a Body Composition Analysis at the beginning and end of the study. The primary goal was to have a 10% improvement in GWBS and 1% reduction in body fat at the end of this study with at least a 50% overall participation.

Results: The average GWBS score (and percent participation) at the 3 different intervals were 71 (52%), 66 (52.3%), and 78 (57%). The pre-intervention to post-intervention change was calculated to be 6%. This correlates to an initial GWBS category of “marginal wellness” that improved to “low positive wellness.” Based on Body Composition Analysis, the average body fat score (and percent participation) at the beginning and end of the study were 27.9% (45%) and 23% (23%) respectively, with a calculated change of -4.9% between the first and second measurements. There was more than 50% resident participation during the lecture sessions and volunteer opportunities.

Conclusion: The Wellness Program was created to educate residents on the importance of balance between physical and psychosocial aspects of life to improve resident wellbeing and in turn, decrease emotional exhaustion and improve performance efficacy. Although, the goal of a 10% change in GWBS score was not achieved, there was an overall positive improvement in resident wellness with increasing resident participation, surpassing the 50% goal. Moreso, the goal of 1% improvement in percent body fat was achieved. In the future, the goal will be to continually increase the percentage of resident participation and foster wellness. By so doing, decrease burnout, create a culture of wellness and improve patient care.

References
Title: Unnecessary Testing for *Clostridium difficile* among Hospitalized Patients at a Tertiary Care Community Teaching Institute

Authors: Anam Hameed, Davis Huang, Andrew Herman, Hamza Aziz, Gabriel Aisenberg

Introduction: *Clostridium difficile* (C. diff.) infection is one of the most common healthcare-associated infections and a significant cause of morbidity and mortality among adult hospitalized patients (1). Because of this, the test and treat strategy has become increasingly common. However, not all patients have symptoms consistent with *C. diff.* infection (CDI) prior to testing. A few studies have been conducted in the past to determine the unnecessary testing of repeat *C. diff.* within 7 days of a prior test (3,4, 5). No study has so far been conducted assessing the initial *C. diff.* test as being unnecessary or whether actually required.

Methods: We conducted a retrospective review of the electronic medical record of all adult patients admitted to the medicine floor from October-December 2016 who underwent testing for *C. diff.* at a tertiary care teaching hospital. Data was collected for demographics, results of *C. diff.* testing, and the use of antibiotics for the treatment of *C. diff.* Statistical analysis was performed using SPSS.

Results: 191 patients (99 females and 92 males) were included in the 3-month study period. All patients were tested using the stool PCR test for *C. diff.* Mean age of the patient population was 51.32 ± 14.58 years. Most patients were Hispanic (n=112) followed by African American (n=50), Caucasian (n= 24) and Asian (n=5). Diarrhea was the most common indication for ordering the test (110/191, X%). 17/191 (9%) patients tested positive for *C. diff.* (Table 1). None of the patients with HIV or inflammatory bowel disease who had presented with diarrhea (7/18, X% and 7/11, X% respectively) tested positive for the pathogen.

Conclusion: The Results: of our study show that only 9% of the patients tested for *C. diff.* in a three-month study period at a tertiary care community teaching hospital tested positive. The most common reason for ordering the test was diarrhea; however, only 6% of these patients tested positive. Testing for *C. diff.* should not be a reflexive order for patients who present with complaints of diarrhea. Better documentation of the presenting symptoms may help identifying factors independently associated with an increasing risk of *C. diff.* infection. The creation of an effective risk score module for the prediction of *C. diff.* may enable clinicians to deliver higher quality care.
Title: Continuing IV antibiotics after hospital discharge: a retrospective review of outpatient parenteral antimicrobial therapy (OPAT) outcomes at a teaching hospital

Authors: Christopher Kim, MD, Ruth Serrano, MD, Jordan Abel, MD, Jose Cadena-Zuluaga, MD, Jason E. Bowling, MD, Heta Javeri, MD, MPH

Introduction: Background: Outpatient parenteral antimicrobial therapy (OPAT) provides patients with appropriate intravenous medical treatments without the need to remain in the hospital. Outcome measurements are important for such programs to assess the efficacy, safety and overall quality of patient care delivered.

Aim: To review the OPAT service within University Health System in San Antonio, TX.

Methods: Methods: We performed a retrospective chart review of patients discharged with intravenous antibiotics from January 2011 to August 2013 at University Hospital in San Antonio, TX. Variables including patient demographics, type and duration of infection, choice of antibiotics and complications during therapy were obtained from the electronic medical database. Primary outcomes looked at rates of relapse, reinfection and cure. Secondary outcomes looked at adverse events during treatment course and the severity of these adverse events.

Results: Results: 148 patients were included in the study (mean age 48.2; 31.8% female; mean Charlson Comorbidity Index score 3.6). The most frequent indications for intravenous antibiotics were bacteremia (n=45, 29.8%), skin and soft tissue infections (n=35, 23.2%), and osteomyelitis (n=34, 22.5%). Staphylococcus aureus represented the most frequent organism targeted (MRSA n=33, 22.4%; MSSA n=31, 21.1%), followed by Gram negatives (n=44, 29.9%) and Enterococcus spp. (n=25, 17.0%). The most common antibiotics used were vancomycin (n=58, 38.7%) and a beta-lactam plus beta-lactamase inhibitor (n=30, 20.0%). Cure, defined as lack of recurrence at one year of follow-up, was achieved in 76.1% of patients. From the subset of patients who experienced failure (n=34, 23.8%), 58.8% had relapse of infection and 41.2% had reinfection, with median time to failure being 53 days. Clinical complications related to therapy occurred in 14.8% of patients, the most common being GI disturbances (n=11). Lab-related complications occurred in 17.5% of patients, with the most common being anemia (n=10). Line-related complications requiring early line removal and replacement occurred in eight patients.

Conclusion: Conclusions: Our results add to the growing observational data showing that OPAT is a safe and effective tool providing curative care for a variety of infections, with the majority of complications being minor.
Texas-Research-Poster Finalist
Dheepa Sekar

Title: Pilot Study of a Half-Day Mini-Curriculum to Improve Resident Confidence In Conducting Goals of Care Discussions

Authors: Dheepa R. Sekar¹, Lauren N. Smith¹, Caitlin Siropaides¹, Oanh K. Nguyen¹

¹Department of Internal Medicine, UT Southwestern Medical Center, Dallas, Texas

Introduction: Although goals of care discussions are an integral component of patient care, and require proficiency in patient communication skills, residents receive limited observation, feedback, or experiential training in these skills. Existing goals of care curricula include training modules spanning multiple days, limiting broad dissemination and implementation given the time constraints of many residency programs. Thus, we sought to develop, pilot, and evaluate the effect of a focused, half-day mini-curriculum on resident attitudes and confidence in conducting goals of care discussions.

Methods: We conducted a single-site pilot study in the internal medicine residency program at UT Southwestern in Dallas, Texas from March through April 2017. Our curriculum consisted of: 1) a didactic lecture to introduce the ‘SPIKES’ framework for breaking bad news (1-1.5 hours); and 2) small group breakout sessions for guided practice in: a) succinctly summarizing a complex hospital course for patients and families; b) anticipating and responding to emotional responses with empathy; and c) eliciting patient priorities to make code status recommendations (1.5-2 hours). We conducted a pre-post survey analysis of resident attitudes towards training and confidence in leading goals of care discussions, with survey responses reported on a five-point Likert scale.

Results: Of 184 total residents, 109 residents (59%) participated in the curricular intervention. Residents reported an improvement in their overall confidence in leading goals of care discussions after the intervention (3.6±0.9 vs. 4.1±0.6, p<0.001), with the largest improvements in confidence responding to emotions (3.5±0.9 vs. 3.9±0.6, p=0.005), making care recommendations to families (3.5±1.0 vs. 3.9±0.7, p<0.001), and quickly conducting an effective code status discussion (3.6±1.0 vs. 4.0±0.7, p<0.001). Additionally, residents reported an increased in desire for supervision (3.7±1.0 vs. 4.0±1.0, p=0.03) and feedback (4.0±0.9 vs. 4.2±0.8, p=0.05) during goals of care discussions.

Conclusion: A single session mini-curriculum improves resident confidence in goals of care discussion skills and attitudes towards supervision and feedback during these discussions, creating a foundation for additional training interventions. Additional experiential training opportunities and observation are likely needed to influence resident competence in these communication skills.
Title: Development of a Hepatocellular Carcinoma Tumor Regression Grading System to Assess Treatment Response to Transcatheter Arterial Chemoembolization (TACE)

Authors: Judy A Trieu, MD, MPH, Adam L. Booth, MD, Heather L. Stevenson, MD, PhD, Department of Internal Medicine, Department of Pathology, University of Texas Medical Branch

Introduction: Hepatocellular carcinoma is the third most common cause of cancer-related death in the world and seventh in the United States. Mean survival is approximately 6 to 20 months without intervention and liver transplantation offers a potentially curative treatment. Transcatheter arterial chemoembolization (TACE) is used to increase survival by preventing hepatocellular carcinoma (HCC) progression while patients are on the transplant wait list, as well as to maintain or down-stage patients within criteria for orthotopic liver transplantation. At this time, however, a histologic tumor regression grading system that evaluates the impact of TACE on HCC progression and/or recurrence does not exist. Our main goal is to develop an HCC tumor regression grading (HTRG) system that can be used to easily score treatment responses in both hepatectomy and liver resection specimens from patients that have previously received TACE treatment.

Methods: We performed a retrospective institutional search for livers explanted from 2016-2017. Eligibility criteria included patients who received TACE at least once prior to transplantation. Baseline demographic and clinical data were collected. One section per centimeter of the tumor nodules that received TACE were evaluated histologically. Histopathologic findings included fibrous capsular breach by viable tumor, total percent viable tumor versus tumor necrosis, and presence or absence of microscopic lymphovascular invasion.

Results: Twenty-eight explanted livers were reviewed and 9 cases met eligibility criteria with 14 total nodules evaluated. All patients were male, with average age of 63 years. Hepatitis C infection was the etiology of HCC for 8 of the 9 patients. Average listing model for end-stage liver disease (MELD) for liver transplantation was 21.7. Wait time from TACE to transplantation averaged 6.4 months. Percentage of viable tumor versus necrosis was estimated according to our developed HTRG system. Majority of nodules were graded HTRG I (<10% viable tumor) or IV (60-90% viable tumor); none were HTRG V (>90% viable tumor). Among nodules with lymphovascular invasion, there was an average of 50% viable tumor, whereas only 32% viable tumor was identified among nodules without lymphovascular invasion. Capsular breaching was seen in 87.5% of moderately-differentiated HCC, as opposed to 50% of well-differentiated HCC.

Conclusion: Residual viable tumor as assessed with HRTG, fibrous capsular breaching by tumor, and microscopic lymphovascular invasion may be prognostic markers for tumor recurrence post-transplantation. Our developed HTRG scoring system will provide pathologists with a simple method that assists in evaluating these characteristics. Use of the HTRG system will increase consistency in pathology reporting and synoptic data, which then can be correlated with clinical outcomes in larger studies.
Introduction: Malnutrition is a common cause of impeding recovery in patients with acute alcoholic hepatitis (AAH). Previous reports have shown that appropriate nutritional supplementation reduces short and long-term mortality in patients with AAH. Current guidelines by the American Gastroenterology Association (AGA) and American Association for the Studies of Liver Disease (AASLD) recommend a “1-1.5 g protein and 30-40 kcal/kg body weight” daily intake for adequate recovery. Despite these clear recommendations, the element of nutrition in AAH is often neglected. At the University of Texas Medical Branch, our baseline compliance with guideline-directed treatment of AAH was 17%. We designed a quality improvement project aimed to increase the compliance to 20% in a 3-month period by providing resident education, with an overall goal of 30% over the subsequent 6 months.

Methods: We retrospectively reviewed charts of all patients admitted with AAH between December 2015 and December 2016. Data analysis was performed and based on Results; an intervention plan was developed and executed. Post-intervention data was collected during a 3-month period. Our interventions included physician education, coordination with dietary and nutrition services to recommend guideline based diet for these patients, and encouraging use of electronic medical record (EMR) based order set for “high-protein/high-calorie” diet.

Results: A total of 53 patients were included in the pre-intervention group and 24 patients were included in the post-intervention group. The presence of appropriate nutrition in the pre-intervention group was only 17%, which improved to 21% in the post-intervention group.

Conclusion: Despite strong evidence, adherence to appropriate nutrition in AAH is low. Although our post-intervention data collection is ongoing, there has been a mild increase in adherence to appropriate nutrition at our institution. Our initial investigation showed that hepatology and nutrition consultation improved compliance with appropriate nutrition. We are developing additional Plan-Do-Study-Act cycles to continue implementation of protocolized care for appropriate nutrition in AAH by incorporating consultation of hepatology and nutrition services, and creation of alert/hard-stops in the EMR once patient is diagnosed with AAH. We plan to collect further data on adherence on nutrition in AAH and assess its impact on outcomes including length of stay and readmission rates.
Texas-Research-Poster Finalist
Sarah E Tuthill, MD

Title: Transfer Bundle Improves Transfer Time from Intensive Care Unit to Medicine Floor

Authors: Sarah Tuthill, MD, Michael Shieh, MD, Drew Emge, MD, Taher Mandviwala, MD, Khaled Sanber, MD, Eric Wu, MD, Sumit Kapoor, MD. Baylor College of Medicine.

Introduction: Efficient transfer of stabilized patients out of the intensive care unit (ICU) is an important aspect of patient care in hospitals. Studies suggest that delays in transfer time have multiple deleterious effects, including increased re-admission rates and increased in-hospital mortality. The purpose of this study was to improve transfer time out of medical ICUs by implementing a transfer bundle that included earlier transfer order placement and a multi-disciplinary approach between the intensivist, hospitalist and nursing staff.

Methods: Baseline data was collected on the time transfer orders were placed, when patients left the ICU, and type of bed requested (floor, telemetry, or step-down) for every patient who was transferred out of a 17 bed ICU over 62 days (n= 136). A transfer bundle was implemented, stipulating: 1) transfer orders be submitted prior to 9 am, 2) hospitalists be involved in patient care 24 hours prior to transfer, 3) transfer note template be completed prior to transfer, 4) outgoing patients highlighted on nursing unit board and 5) ICU transfer huddles be held with the charge nurse twice daily. For thirty days following bundle implementation, the original pre-implementation variables were again collected for all patients transferred (n=122). The number of ICU transfer notes written was used as a surrogate for measuring bundle compliance. An unpaired two-tail t-test with Welch’s correction was used to determine statistical significance comparing average transfer times. All p-values were calculated using Prism 6 (GraphPad).

Results: In the 30 days following initiation of the ICU transfer bundle, orders placed prior to 12 pm increased by 18% and average transfer time decreased by 3.7 hours (p=0.09). Transfer time from ICU to floor beds decreased by 3.9 hours (p = 0.03). Transfer times decreased for step-down beds by 5.8 hours (p = 0.18) and increased for telemetry beds by 1.8 hours (p = 0.27). Transfer note compliance was 71%.

Conclusion: Multiple transfer parameters improved following implementation of the ICU transfer bundle, including a significantly reduced overall average transfer time by 3.7 hours, and a significantly reduced average transfer time to floor beds by 3.9 hours. The decrease in transfer time to step-down by 5.8 hours likely did not approach significance due to a low number of step-down transfers during study period. Study Results: were likely limited by a change to our hospital's bed request system that coincided with ICU bundle initiation. Following these early promising Results: of the ICU transfer bundle, we plan to continue our study with further aims directed at increasing bundle compliance and increasing the efficiency of the new bed request system.
Title: Examining the Initial Episode Leading to Long-Term Opioid Use at a Military Medical Treatment Facility

(Podium presentation of original research)

Authors: Capt Micheal A. Massoud, USAF, MC, ACP Associate
Capt James D. Dizmang, USAF, MC, ACP Associate
Ronald J. Markert, Ph.D., WSU SOM
Maj Joshua N. Scott, USAF, MC, ACP Member
Maj Kathryn M. Burtson, USAF, MC, ACP Member

Introduction: The purpose of this study is to examine the most commonly prescribed opiates, and any characteristics or diagnoses which make a provider or patient more likely to receive chronic pain medication. This study reviewed the initial prescription opioid episode which lead to chronic opioid use, then described characteristics of the provider and patient. It is unique in its scope—to identify, evaluate and offer some analysis in a military population being served at a military medical center.

Methods: This single-center retrospective study evaluates initial opioid episodes leading to chronic use in patients at least 16 years of age between January 2012 and May 2017 enrolled at Wright Patterson Medical Center (WPMC) located in Dayton, Ohio.

This study will analyze data from the electronic health records for patients that have been taking any oral medication containing: hydrocodone, dihydrocodeinone, or oxycodone for greater than 12 weeks. The population includes Active Duty Military, dependents of Active Duty, Veterans/Retired, dependents of Veterans/Retired, and other, with exclusion criteria including cancer patients or hospice patients.

Results: In a period of just over 5 years 11,635 total studied opioid prescriptions for 3,701 patients were examined, representing 19.86% of the total population of such patients. Of these 3,701 there were 348 which met inclusion criteria (of examined: 9.4%).

Remainder of results are complete, however manuscript is still in process. Variables were as follows:

• Primary outcome
  – Specialty of the prescriber
• Secondary outcomes
–Provider credentials
–Clinic location
–Pain diagnosis
–Patient characteristics

•Age, rank, gender, psychiatric history, perioperative

Conclusion: It is beyond the intent of this description and analytical study to determine utility of long-term opioid in non-cancer patients at this military medical center, rather the intent is to identify the population and analyze trends which could affect prescription habits. It is known, however, that a rise in the opioid epidemic is a concern. This study helps to describe a unique population in a unique medical system. A population mix of active duty military members, their spouses and dependent family, and veterans and their spouses.

We have identified patient’s initial episode which resulted in long-term opioid use at this medical treatment facility. To date, no other study has examined characteristics of mixed active duty military and veteran population with their families included in regard to these common opioids.

Characteristics of age, gender, and diagnosis of initial episodes leading to long-term opioid use were discovered during this study. This sample, nearly 20% sampled of the population, resulted in similar findings to a large civilian study in California.
Title: Palliative Care Utilization in Patients with Newly Diagnosed, Incurable Solid Tumor Malignancies: Experience of an Institution New to Palliative Care

Authors: Josh Romain, Wilfred Delacruz, Adrian Bersabe

Introduction: Early palliative care (PC) referral can enhance quality of life (QOL) in patients with metastatic cancer and even improve overall survival in non-small cell lung cancer. Despite compelling data showing benefits of early integration of PC, institutional barriers to care exist. Our institution recently established an outpatient PC service. To improve utilization of this new service, we identified clinicians’ attitudes and barriers to PC and provided education on the PC referral process. The effects of this intervention on PC utilization were assessed.

Methods: A retrospective chart review was completed on outpatient medical oncology referrals to identify patients referred for newly diagnosed, solid tumor malignancies with no curative options. Hospice referrals were excluded. Clinicians participating in multi-disciplinary tumor boards were surveyed to identify PC referral practices and attitudes/barriers to PC referrals. Education on PC and the PC referral process was provided during the survey period. Post-intervention review of PC referrals over four months was conducted.

Results: There were 53 new, pre-intervention consultations meeting study criteria, and only three (5.7%) patients were referred to PC. Our survey revealed that 66% (31/47) of oncology providers did not know the institution had onsite PC available and only eight (17%) knew how to place a referral. After education was provided, a second cohort revealed 24.1% (7/29) of patients were referred for outpatient PC representing an 18.4% absolute increase.

Conclusion: We significantly increased PC referrals for medical oncology patients with advanced or metastatic disease. However, other efforts to improve institutional awareness and change the culture of care are needed to ensure that majority of these patients are appropriately referred. Longitudinal evaluation will further ensure that timely adjustments are made and that effects are permanent.

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Introduction: Leishmaniasis is a zoonotic parasitic disease transmitted by sand fly bites. Visceral leishmaniasis (VL) is a chronic intracellular infection which, when symptomatic, can be fatal without therapy. Subclinical or latent VL may occur in a majority of those infected with lifelong risk of activation when immunosuppressed. Symptomatic VL has been described in Soldiers deployed to Operation Iraqi Freedom (OIF). We report the prevalence and risk characteristics of latent VL infection in OIF Soldiers.

Methods: Healthy Soldiers deployed during summer months (2002-2011) to VL endemic areas of Iraq were recruited from Fort Bliss, Texas. Responses to a risk factor survey and blood samples were obtained. Leishmania research diagnostics were performed on serum and/or white blood cells to include ELISA, rk39 immunochromatography, qPCR, and interferon gamma release (IGRA) assays. Analyses included descriptive percentages and other summary statistics. Fisher’s Exact test and logistic regression were used for group comparisons.

Results: Out of 88 subjects enrolled, 76/88 (86%) were male with median age 39 years and deployment duration of 365 days. The prevalence of latent VL was 10.2% (CI 4.8%-18.5%) with 7 IGRA positive and 2 ELISA positive. Travel to Ninewa governate correlated with VL, p<0.05. No significant differences were noted in occupation, personal protective measures, deployment timeframe, or sleeping conditions between VL positive and negative individuals. In persons with latent VL, 4/9 (44.4%) and 6/9 (66.7%) deployed to Ninewa and Baghdad respectively, 7/9 (77.8%) were outdoors most nights, 5/9 (55.6%) slept on the ground during deployment, 5/9 (55.6%) were medical personnel, 7/9 (77.8%) slept in less than full uniform, and 8/9 (88.9%) never or rarely used insect repellent.

Conclusion: Latent VL was identified in asymptomatic OIF Soldiers (10.2%). Travel to Ninewa governate correlated with VL latent infection. In the latent VL group, many were healthcare workers, slept on the ground or in less than full uniform, and rarely used insect repellent. Further studies are needed to inform risk of reactivation disease in latently infected US Soldiers and to target measures for broader surveillance and safety, such as the screening of military blood donors.
Title: Quality Improvement of 24 hour Ambulatory Blood Pressure Monitor

Authors: Alexandra Stewart, DO, ACP Associate; Catherine Miloni, RN; COL Troy Denunzio, DO, ACP Member; MAJ Sarah Gordon, MD, ACP Member; Department of Medicine, Tripler Army Medical Center, Hawaii

Introduction: Traditionally, the diagnosis of hypertension has been based off of in-office blood pressure measurements. A variety of factors can affect these values and lead to inaccurate diagnoses. Literature suggests 24 hour ambulatory blood pressure monitoring (ABPM) predicts organ damage and cardiovascular events better than office blood pressure readings. It is now considered the gold standard by professional societies. However, it is an often underutilized tool. We instituted a quality improvement project to increase the number of patients referred for ABPM and monitor patient outcomes.

Methods: At Tripler Army Medical Center, 24 hour ABPM was formally organized and monitored beginning in January 2017. We identified lack of physician education as well as patient time and travel constraints as referral barriers. We therefore began education for Referring clinics on the literature supporting use of 24 hour ABPM and institutional availability in August 2017. We also modified the referral process to include patient contact directly by clinic staff to explain the reason for the study and to assist the patient in scheduling. At completion of the study, patients are provided a brief meeting with the interpreting physician to receive appropriate education. We tracked the number of patients referred, demographic data, and number of resulting changes in clinical management.

Results: Between January 2017 and November 2017 280 patients were referred. 28 were excluded due to incomplete data resulting in 252 completed ABPMs for analysis. The most common reasons for referral were suspected white-coat hypertension (WCH) and medication titration. 55% were male and 45% female. Average age was 52 (19-91). Average body mass index was 28.48 (17.18-49.90). 29.9% self-identified as Asian/Pacific Islander, 5.2% Hispanic, 13.2% African American, 8.4% other, 2.8% unknown and 40.6% White. After implementation of primary care outreach, an average of 10 additional patients per month (50% increase) were referred. We identified 93 patients as under or over-treated. 88 patients were referred for suspected WCH with greater than 25% of these subsequently diagnosed with hypertension. 73 patients, including 65 diagnosed with WCH, were prevented from taking unnecessary medications. All but two patients had prompt primary care follow up and received the recommended medication changes.

Conclusion: Following implementation of primary care outreach and direct patient contact by our clinic staff, number of ABPM referrals per month increased. Greater than 35% of patients who completed an ABPM had a resulting change in clinical management. These findings support existing evidence that the use of 24 hour ABPM is a potentially more accurate tool for assessment of hypertension than office blood pressure. It may also prevent adverse outcomes associated with undertreatment or overtreatment of blood pressure. When home and office blood pressure discrepancies exist, referral for 24 hour ABPM is an invaluable tool for patient care.
Title: Inpatient Emergency Department Admissions: One Institution’s Response to The Joint Commission’s ORYX Quality Measures

Authors: Cornelia Willis, CPT, MC, USA (ACP Associate), Diana Schofield ARNP, Cristin Mount, LTC (P), MC, USA, FACP, Madigan Army Medical Center, Tacoma, Washington.

Introduction: Prolonged Emergency Department (ED) stays contribute to overcrowding and increased preventable errors as well as delays in definitive treatments for patients. The Joint Commission’s ORYX initiative, ED-2, established the national standard of 90 minutes from the Emergency Department’s decision to admit (DTA) to the patient’s arrival to the floor. In collaboration with the ED, the Madigan Internal Medicine (IM) service initiated a 60 minute goal for placing holding orders with an abbreviated review of the patient’s chart. The aim of this study was to evaluate the clinical impact and safety of this intervention.

Methods: The IM Inpatient service at one Department of Defense (DOD) teaching hospital initiated a 60 minute time limit for the placement of holding orders. An educational intervention with the IM Residency and Hospitalist Services was conducted to examine current practices and encourage improvements in efficiency with ED evaluations. Monthly feedback was given to both services on performance in terms of DTA to Admit Order Time. Daily data collected by the hospital ED on patient acuity and movement to the Internal Medicine Service was reviewed for a one month period prior to initiating educational intervention and the 60 minute limit on physician admissions. This was then compared to data collected 1, 6, 18, 19, and 20 months post-intervention. Statistical analysis with Student T-test and linear regression analysis was performed.

Results: One month prior to the intervention, the IM Service spent on average 75 minutes before placing orders. One month after the intervention, DTA time decreased to 34 minutes (p=<0.001) with 40% of patients spending longer than 90 minutes in the ED. The decreased DTA time was sustained at all follow up intervals (p=<0.001). The number of patients spending longer than 90 minutes in the ED decreased to 3.4% in 18 month follow up. These improvements were independent of patient acuity and number of daily admits (R² linear=0.002, 0.00002). There was no increased reporting in patient safety issues or sentinel events following the policy change.

Conclusion: An in depth chart review is not always necessary for a safe admission from the ED. Placing holding orders for floor admissions prior to or while evaluating the patient is a safe practice for many hospital admissions and facilitates patient movement to help meet ORYX measures.

References

Title: Cost-Effectiveness of Norovirus Vaccine Acquisition Strategy Against Diarrheal Disease in the Peruvian Military

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Introduction: Worldwide Norovirus (NoV) is a significant cause of diarrheal disease in both civilian and military populations, presenting with debilitating diarrhea and vomiting. Previous studies have shown the negative impact due to duty days lost (DDL) in both garrison and deployment settings, as well have described how the public health sector in a developing country, such as Peru, is profoundly affected by diarrheal disease caused by NoV. Given the burden of diarrheal disease in Peru one might suggest that the Peruvian military may benefit from a NoV vaccine acquisition strategy (VAS), as their military is circulated throughout the country, possibly carrying infectious disease along with them and/or becoming infected through contact with the local communities, as previous studies have suggested.

Methods: An economic model that had been developed by Riddle, et al. to assess the cost-effectiveness of a vaccine acquisition strategy (VAS) against NoV within the US Department of Defense (DoD) was applied to the Peruvian military. The model compared the cost-effectiveness of a VAS to current medical management, including no treatment, NoV-associated resource consumption and aeromedical evacuation in instances of severe disease. Given the large burden of NoV disease in Peru and the medical exchange that occurs between the Navies of the United States and Peru, this model was applied to answer the question regarding the cost effectiveness of obtaining a VAS against NoV for the Peruvian military in regards to the value of a DDL.

Results: It is estimated that a VAS for NoV for the Peruvian military would cost $4,038,170, while cost of care without a NoV vaccine is estimated to be $995,153. Acquisition of a NoV vaccine by the Peruvian military is estimated to result in a cost-effectiveness ratio per duty day lost to NoV diarrheal illness (CER_{DDL}) of $337 to avert a DDL.

Conclusion: NoV diarrheal disease poses a burden in deployment and garrison setting in Peru, affecting both the military and civilian populations. At this time, the monetary cost of a VAS is more than what is spent on current treatment strategies. However, the cost-effectiveness of obtaining a VAS may be found when considering the value of a duty day lost due to diarrheal disease in terms of mission readiness.
References


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Title: Individualized and Interprofessional Cross-Continuum Complex Care Maps© are associated with Reduced Utilization and Cost for Frequent High-Need Users of Emergency and Inpatient Services

Authors: Adam Kilian MD 1, 2, Lauran Hardin MSN, RN-BC, CNL 1, 1 Trinity Health-Michigan d/b/a Mercy Health Saint Mary’s, Grand Rapids, MI, USA, 2 University of Utah Health Care, Salt Lake City, Utah, USA

Introduction: High-need, high-cost (HNHC) patients can over-use acute care services, a pattern of behavior associated with many poor outcomes that disproportionately contributes to increased U.S. healthcare cost. Our objective was to reduce healthcare cost and improve outcomes by optimizing the system of care. We targeted HNHC patients and identified root causes of frequent healthcare utilization. We developed a cross-continuum intervention process and a succinct tool called a Complex Care Map (CCM)© that addresses fragmentation in the system and links providers to a comprehensive individualized analysis of the patient story and causes for frequent access to health services.

Methods: Using a pre-/post-test design in which each subject served as his/her own historical control, this quality improvement project focused on determining if the interdisciplinary intervention called CCM© had an impact on healthcare utilization and costs for HNHC patients. We conducted the analysis between November 2012 and December 2015 at Mercy Health Saint Mary’s, a Midwestern urban hospital with greater than 80,000 annual emergency department (ED) visits. All referred patients with three or more hospital visits (ED or inpatient [IP]) in the 12 months prior to initiation of a CCM© (n=339) were included in the study. Individualized CCMs© were created and made available in the electronic medical record (EMR) to all healthcare providers. We compared utilization, cost, social, and healthcare access variables from the EMR and cost-accounting system for 12 months before and after CCMs© implementation. We used both descriptive and limited inferential statistics.

Results: ED mean visits decreased 43% (p<0.001), inpatient mean admissions decreased 44% (p<0.001), outpatient mean visits decreased 17% (p<0.001), computed tomography mean scans decreased 62% (p<0.001), and OBS/IP length of stay mean days decreased 41% (p<0.001). Gross charges decreased 45 % (p<0.001), direct expenses decreased 47% (p<0.001), contribution margin improved by 11% (p=0.002), and operating margin improved by 73% (p<0.001). Patients with housing increased 14% (p<0.001), those with primary care increased 15% (p<0.001), and those with insurance increased 16% (p<0.001).

Conclusion: Individualized CCMs© for a select group of patients are associated with decreased healthcare system overutilization and cost of care.
Title: Initiating a protocol for thorough neuroprognostication of the post-cardiac arrest patient

Authors: Avneet Singh M.D., Michael Theriault, M.D., Ryan Clouser D.O., Gilman Allen, M.D.

Introduction: Advanced cardiac life support emphasizes the importance of post-cardiac arrest care which includes targeted temperature management. Therapeutic hypothermia (TH) or targeted temperature management protocols (TTM) may halt or prevent ischemic brain injury\(^1\). TH was shown to be beneficial in improving both mortality and neurologic outcomes following out of hospital VF/VT arrest, however, it has also been shown that moderate hypothermia to 33-34\(^\circ\)C alters the validity of traditional prognostic tools which could lead to premature withdrawal of support. Nielsen et al. demonstrated that cooling to 36\(^\circ\)C was non-inferior to 33\(^\circ\)C. TTM to 36\(^\circ\)C is unlikely to impact the ability to prognosticate, allowing for more rapid neuroprognostication by 72 hours following return of spontaneous circulation. Data collected from record review suggested that patients treated with TH did not show any standardized method of prognostication and were unable to tolerate TH and did not receive aggressive temperature management post arrest. The Medical Intensive Care Unit, in conjunction with the Neurology department implemented a protocol which delineated TH/TTM post-cardiac arrest care to maximize prognostic information for patients and families and prevent premature withdrawal of support.

Methods: A protocol was implemented to ensure appropriate neuroprognostication as well as the option to cool to 36\(^\circ\)C (TTM) for patients that were unable to tolerate TH. Non-contrast CT brain on admission, cEEG, Somatosensory evoked potentials and MRI were also standardized in cases of persistent encephalopathy. All cardiac arrests from Feb 2012-Dec 2014 and August 2016-October 2017 requiring TH/TTM protocol underwent chart review to determine protocol compliance and outcomes.

Results: Cardiac arrests prior to the initiation of the protocol (2/2012 to 12/2014; total of 121 patients) were analyzed; of these only 50 (41.6%) completed the full protocol and 12 patients were discharged to sub-acute rehab, 9 home, and 28 to the morgue, (1 lost to follow up). Prior to creation of our new protocol, the average time to comfort measures orders (CMO) for patients treated with TH (33\(^\circ\)C) was 4.75 days after return of spontaneous circulation. 71\% of patients had cEEG and 24\% of patients had an MRI. Patients treated with TTM (36\(^\circ\)C) had a mean time to CMO of 2 days. Post protocol creation, 23 patients were treated. 65.2\% completed the protocol, 91.3\% underwent CT imaging while 30.4\% underwent MRI imaging. cEEGs were completed on 78.2\% and the mean time to CMO was 3 days for those treated with 33\(^\circ\)C protocol and 3.33 days for those treated with 36\(^\circ\)C protocol.

Conclusion: Data following implementation of the protocol continues to be collected. Analysis of the first 23 cases demonstrates that more patients received the benefit of temperature management post cardiac arrest and the time to withdrawal of support was increased thus potentially preventing premature withdrawal of support.

References
**Virginia-Research-Poster Finalist**  
**Sumera A Bukhari, MD**

Title: Lung Cancer Screening: Need for Better Compliance and Broader Guidelines

Authors: Sumera Bukhari, MD (Virginia Tech-Carilion Clinic), Ahmed Dirweesh, MD (St. Francis Medical Center), Afolarin Amodu, MD (St. Francis Medical Center), Areig Awad, MD (St. Francis Medical Center), Rishita Yelisetti, MD (St. Francis Medical Center), Rabia Soomro, MD (St. Francis Medical Center), Mohammad Y. Ibrahim, MD (St. Francis Medical Center), Ali Mustansar Mir, MD (St. Francis Medical Center), Mohammad Arsalan, MD (St. Francis Medical Center), Vincet DeBari, PhD (St. Francis Medical Center), Anad Kaji, MD (St. Francis Medical Center), Sara Wallach, MD (St. Francis Medical Center)

**Introduction:** Lung cancer is the leading cause of cancer-related death in the United States. Many patients report no symptoms whatsoever and the majority of them become symptomatic only with advanced local or metastatic disease. For clinicians, it becomes a diagnostic challenge at times to have an index of suspicion regarding lung cancer in patients, particularly smokers, who don’t present with typical respiratory or systemic symptoms. The United States Preventive Services Task Force (USPSTF) recommends annual low-dose computed tomography (CT) of the chest in asymptomatic adults aged 55 to 80 years with a 30 pack-year smoking history for current smokers or those who quitted smoking within the past 15 years. The purpose of this study is to compare the incidence of lung cancer in patients who fitted the USPSTF guidelines to those who did not fit the screening criteria and to assess the prior compliance among those who fitted the screening guidelines.

**Methods:** We retrospectively reviewed records of all patients with a biopsy proven primary lung cancer at our hospital from 2011 to 2017. All patients aged 18 to 80 years with available chest CT scans and histopathological reports were included. Patients were evaluated for baseline characteristics, smoking history, and prior screening if indicated.

**Results:** Of the 104 patients included, 55 (53%) fitted the USPTSF guidelines and 49 (47%) did not. None of this population underwent screening prior to diagnosis. Males constituted 73% of patients who fitted the guidelines, and 51% of those who did not (p-value 0.48). Both groups matched for racial distribution (26% black, 67% white, and 7% other races in the fitted group; and 23% black, 63% white, and 14% other races in the non-fitting group; p-values: 0.49, 0.49, and 0.53 respectively). Non-Small Cell Lung Cancer (NSCLC) occurred in 98% of patients who fitted the screening guidelines and in 96% of those who did not (p-value 0.47).

**Conclusion:** All patients who met the screening guidelines were never screened before the diagnosis. This warrants the need for a strategy as an institution and community to get better compliance for screening. There was no statistical significance in the incidence of NSCLC between the study groups. This emphasizes the need for an effective smoking cessation programs in the community and the need for further studies to determine the usefulness of the current guidelines.

**References**

Title: An Educational Intervention Increases Indicated Telemetry Use

Authors: Charles Cui, MD; Danielle Johnson, MD; Chris Nguyen, MD; Ariel Sindel, DO

Introduction: The Problem:

Inpatient telemetry monitoring is a valuable diagnostic tool; however, when used inappropriately, it can lead to increased cost as well as patient discomfort. During residency, trainees receive little to no training on indications for telemetry and often fail to realize when telemetry is not appropriate. In 2004, the American Heart Association (AHA) published practice standards that help define appropriate telemetry use and could potentially be a useful tool to teach trainees.

Project Goals:

This quality improvement study aims to use educational intervention to improve the percentage of adherence to recommended telemetry practices. This intervention will educate the trainee on practice standards as well as increase the number of re-evaluations during hospital course for appropriate use of telemetry.

Methods: Data was collected from medicine teams prior to intervention looking at number of patients on telemetry and the percentage of those patients who met practice standards for appropriate use. Teams participated in a pre-intervention survey to assess knowledge of indications and attitudes toward telemetry. Medicine teams received individual education sessions and were given pocket cards summarizing the guidelines. Post-intervention data on team use of appropriate telemetry was obtained and the survey was repeated.

Results: Pre-intervention, the mean percentage of telemetry indications meeting AHA practice standards was 66%; this increased to 79% post-intervention. Post-intervention, trainees stating they had “full confidence” in using telemetry increased from 8% to 11%, “mostly confident” increased from 15% to 33%, while “not confident” decreased from 15% to 0%. Post-intervention, trainees stating they checked telemetry “always” increased from 8% to 11%, “sometimes” increased from 15% to 45% and “less than half the time” decreased from 69% to 33%.

Conclusion: Educational interventions can objectively improve appropriate use of telemetry among resident trainees. Additionally, these interventions can subjectively increase confidence in use of telemetry and the number of times telemetry monitoring is followed up.

Next Steps:

Further studies would focus on a larger population of trainees receiving the intervention, encompassing the intern class during orientation as well as residents during night float curriculum. In addition, appropriate telemetry use can be tracked across trainee levels to see if there is improvement from intern year up to the third year of residency.
Title: Patterns and Perceptions of Healthcare Usage in Pampas Grande, Peru.

Authors: Ariella Krones, Carolyn Zin, Sean McKenna, Sarika Modi, Michael P. Stevens

Introduction: Since 2009 the Richmond Global Health Alliance (RGHA) has traveled to the district of Pampas Grande in Peru at least yearly to provide supplemental medical care to the local health post. The health post is the point of first access for the local population when they need healthcare, and is theoretically covered by the Comprehensive Health Insurance (SIS) from the Ministry of Health. Anecdotal experience from mobile RGHA clinics demonstrates that the community may prefer alternative sources of healthcare and has limited access to health insurance. A needs assessment survey was created to inform RGHA outreach efforts in the region.

Methods: A nineteen-question IRB approved survey was constructed from validated tools with similar aims, including the CDC’s national Behavioral Risk Factor Surveillance system and a previous RGHA needs assessment that focused on community morbidity and effect of distance on health access. Fifty participants who attended RGHA clinics in June 2017 were surveyed. After consent was obtained the survey was administered verbally by an experienced translator.

Results: In response to questions about where interviewees received care in the last year, 44% (22/50) responded that they attended the local health post at least once, while the remaining attended private or public clinics and hospitals. When asked about insurance, 88% (44/50) of interviewees said that they did have insurance. Questions focusing on medication availability yielded that 54% (27/50) could reliably obtain medication. 48% (24/50), said their medications were free. Of the thirty-seven interviewees who answered questions about barriers to health and community needs, medication cost (17/37, 44%), access to healthcare (17/37, 44%), and economic security (16/37, 41%) were identified most frequently.

Conclusion: Based on these data, there is a cohort of individuals in Pampas Grande who have health access outside of the RGHA clinics. Barriers to accessing healthcare were identified, including access to medications and specialists. Economic security was also a major issue identified. These data will inform RGHA efforts in the region.
Title: Right Atrial to Left Atrial Volume Index Ratio is Associated with Mortality in Patients with Pulmonary Hypertension

Authors: Manu Mysore MD, Kenneth Bilchick MD, Benjamin Ruth MD, William Harding MD, Christiana M. Jeukeng MD, Jamie Kennedy MD, Andrew Mihalek MD, and Sula Mazimba MD

Introduction: Pulmonary hypertension (PH) is a progressive condition characterized by increased pulmonary vascular resistance leading to right heart failure. Elevated right atrial (RA) pressure reflects right ventricular (RV) pressure overload in patients with PH and is an established risk factor for mortality. We hypothesized that in patients with PH, an increase in the ratio of RA to LA volume index, RA-LAVI, would be associated with increased mortality.

Methods: We evaluated the association between RA-LAVI with long-term clinical outcomes after adjustment for REVEAL Risk Score, an established risk score in pulmonary hypertension. Patients in this study were from a tertiary academic center, with pulmonary hypertension, and had data available for both right heart catheterization (RHC) and transthoracic echocardiogram (TTE) within one year of each other. LA and RA volume indices were measured using the Biplane Disc summation Method in the four and two Chamber Views by two independent researchers. Multivariable logistic regression was used to model RA-LAVI ratio to mortality.

Results: Among 124 patients (mean age 62 ± 12.7 years, 68.6% female), each unit increase in the RA-LAVI ratio was associated with a nearly two-fold increase in mortality (OR 1.912, 95% CI 1.20-3.04). In the multivariable logistic regression model adjusted for the REVEAL Risk score, each unit increase in the RA-LAVI ratio was associated with a nearly two-fold increase in mortality (OR 1.734, 95% CI 1.003-2.998). Furthermore, RA-LAVI ratio in the highest quartile (>1.42) was significantly associated with elevated right atrial pressure (RAP) to pulmonary wedge capillary pressure (PWCP) ratio (RAP/PWCP) (0.76±0.41, p=0.02) compared to the lowest quartile (<0.77), suggesting an interaction between invasive hemodynamic data, atrial structural changes and mortality in PH patients.

Conclusion: Increased RA-LAVI ratio in PH is associated with mortality, and accounts for atrial structural remodeling related to invasive hemodynamics. These findings support further study of this index for guiding risk stratification in PH patients.
West Virginia-Research-Poster Finalist
Obadah S Aqtash, MD

Title: Inappropriate Testing for Clostridium Difficile Infection in Hospitalized Patients, a Closer Look

Authors: Obadah Aqtash MD, Hisham Awad MD, Chris Fitzpatrick PharmD, Derek Evans PharmD, Kara Willenburg MD

Introduction: Clostridium difficile (C-Diff) infection is a known cause of mortality and morbidity in hospitals, adding nearly 4.8 billion dollars a year to the health care burden. Testing for this organism is governed by guidelines highlighted in the Infectious Diseases Society of America (IDSA). Only patients with clinically significant diarrhea benefit from testing as a positive test can represent colonization or active disease. Our objective is to minimize unnecessary testing for C-Diff by providing health care clinicians with the latest evidence based guidelines in managing patients’ diarrhea. We also aim to outline the financial benefits that result from minimizing inappropriate testing.

Methods: Prospective study performed at a university tertiary medical center. A clinically significant diarrhea was defined by the IDSA guideline for C-Diff testing “as passage of 3 or more unformed stools in 24 or fewer consecutive hours”. Unless this was clearly documented in the patient’s file, a bedside patient interview was held for verification. Complete profile review was performed for all patients who were hospitalized for greater than or equal to seven days and tested for C-diff. Our aim was to detect any potential medications or interventions that could lead to developing diarrhea; Including antibiotics and proton pump inhibitor (PPI) use within 7 days prior to development of diarrhea, a bowel regimen for constipation or tube feeding within 48 hours of onset of diarrhea.

Results: Results were collected over a one month period. A total of 53 C-Diff DNA amplified tests were performed, of those 53 cases, only 16 cases (30%) met the guidelines for collection while 37 cases (70%) did not meet these guidelines. Of the 16 cases that met the guidelines, only 5 (31%) cases came back positive for C-Diff, which is equal to 9.4% of the total sample. Of the 37 cases that didn’t meet the guidelines, 13 cases didn’t undergo any testing as patients were unable to provide a stool sample, the other 24 cases were tested for C-diff and all were found to be negative. The financial burden of the negative tests that did not meet the guidelines was around $7519 ($314 per test). Projected over one year, the price would be $90228. Taking into account, that this number doesn’t include the cost of sample containers, transport, testing kits, prophylactic antibiotics, and isolation rooms/equipment’s. The data also showed 50% of the patients tested were on laxatives.

Conclusion: Testing for C-Diff infection in patients with no clinically significant diarrhea has an extremely low yield. Additionally, testing those patients has a significant negative financial impact, as well as a negative psychological impact that results from being placed in isolation rooms. In addition, there is risk of side effects with the unnecessary use of prophylactic antibiotics.
West Virginia-Research-Poster Finalist
Hassaan Yasin, MBBS

Title: Emergency Room Visits & Hospital Admission Rates After Adjuvant Chemotherapy for Breast Cancer at Charleston Area Medical Center

Authors: Hassaan Yasin, Lance M. Workman, Steven J. Jubelirer, Christine A. Welch

Introduction: Breast cancer is the number one diagnosed cancer among women. It is the second leading cause of cancer deaths in West Virginia and worldwide. Adjuvant chemotherapy can lead to an increase in survival rate and decrease the risk of recurrence of breast cancer. However, adjuvant chemotherapy can result in side effects which may result in emergency room (ER) visit or hospitalizations (HA). The purpose of our study is to examine the reasons for and the factors associated with ER visits and HA after chemotherapy in patients with early stage breast cancer.

Methods: A retrospective cohort study of all patients with early stage breast cancer who completed at least one cycle of curative chemotherapy for breast cancer between January 2010 and December 2016 at Charleston Area Medical Center.

Results: After completion of 234 records it has been found that 94% of the population was Caucasian, 71% married and the mean age was 55 ± .64 (range 26 to 81). Comorbidities included hypertension (51%), depression or anxiety (43%), Hyperlipidemia (32%), hypothyroidism (23%) and diabetes (17%). The most commonly used chemotherapy drugs were adriamycin, cytoxan and taxol (28%), adriamycin and cytoxan (21%), and taxol and cytoxan (18%). Twenty-eight percent of the patients were stage I, while 56.7 were stage II and 14% were stage III. In the 30 days after completion of chemotherapy 28% of the patients had had a health problem in which they sought assistance either by telephone, clinic visit or ER. Of those who had a health problem 78.8% had one contact, while 21.2% had two or more contacts. The oncology clinic saw 1% of the population, while 13.6 % visited the ER and 6.4% were hospitalized. The reasons for the visits for which some patients had more than one included neurologic (43 %), diarrhea (24%), swelling (24%), fever alone (19%) febrile neutropenia (19%), atrial fibrillation (14%), nausea (10%), cellulitis (10%), pain (10%) and dysuria (4.8%).

Conclusion: In this study, 28% of the population sought assistance for a health situation in the 30 days post chemotherapy. No definite factors thus far have been associated with ER visits or HA such as stage of disease, tumor size, comorbidities or chemotherapy regimen used. Other factors are being studied such as use of G-CSF or geographic home location (urban versus rural). More research is needed to identify patient, provider and health system factors associated with ER visits and HA so these can be minimized in the future to lessen the burden on the healthcare system.
Title: FLUOROQUINOLONE RESTRICTION DECREASES RATES OF CLOSTRIDIUM DIFFICILE: RESULTS OF A MIXED-METHODS STUDY

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Introduction: Clostridium difficile infection (CDI) is a leading hospital acquired infection and costs our health care system $7 billion annually. Antimicrobial stewardship is an important component of a multifaceted infection prevention program to decrease CDI rates. To maximize impact, front-line provider perceptions of barriers and facilitators of successful interventions must be understood and used to inform stewardship policies. We conducted a mixed-methods study of a fluoroquinolone restriction aimed to reduce rates of hospital-acquired CDI on high-risk units.

Methods: We instituted a fluoroquinolone restriction policy in our medical-surgical intensive care and transplant units. This policy required approval from our antimicrobial stewardship team prior to prescribing fluoroquinolones on the identified units. We compared pre- and post-intervention rates of CDI, fluoroquinolone use, use of alternative antimicrobials, hospital length-of-stay, readmissions, and crude mortality. A chart review of randomly selected patients receiving fluoroquinolones pre- and post-intervention was performed to determine indications for fluoroquinolone prescribing. Semi-structured interviews were conducted with providers on these units after the institution of the restrictive policy to identify barriers and facilitators to fluoroquinolone use and the successful implementation of the restrictive policy. Content was coded and analyzed using the Systems Engineering Initiative for Patient Safety (SEIPS) model.

Results: The restriction policy reduced fluoroquinolone usage by an average of 68% on the two units. CDI rates decreased by 42% with no increase in crude mortality. Fluoroquinolone use before and after restriction was directed towards empiric double coverage of suspected gram negative sepsis and transition to oral therapy in anticipation of discharge. Readmissions and hospital length of stay on the two units did not differ significant in the pre- and post-intervention periods. Qualitative analysis of interviews with 12 clinicians and pharmacists revealed beta-lactam allergy and pending discharge encouraged fluoroquinolone use; a history of CDI and pharmacist involvement in antimicrobial decision-making discouraged fluoroquinolone use.

Conclusion: A fluoroquinolone restriction policy reduced hospital-acquired CDI rates and significantly reduced fluoroquinolone use with no increase in crude mortality, readmission rate or length of stay. Knowledge of front-line barriers and facilitators to fluoroquinolone use can inform an effective antimicrobial stewardship intervention. Lessons from this pilot have been applied to an institution-wide restriction policy.