NEW JERSEY CHAPTER
AMERICAN COLLEGE OF PHYSICIANS

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<td>Ischemic strokes are mainly due to cardioembolic occlusion of small vessels, as well as large vessel thromboemboli. We describe a case of intrapulmonary A-V shunt as the etiology of an acute ischemic event. A 63 year old male with a past history of supraventricular tachycardia and recurrent deep vein thrombosis; who has been non-compliant on Rivaroxaban, presents with pleuritic chest pain and was found to have a right lower lobe pulmonary embolus. The deep vein thrombosis and pulmonary embolus were not significant enough to warrant ultrasound-enhanced thrombolysis by Ekosonic EndoWave Infusion Catheter System, and the patient was subsequently restarted on Rivaroxaban and discharged. The patient presented five days later with left arm tightness and was found to have multiple areas of punctuate infarction of both cerebellar hemispheres, more confluent within the right frontal lobe. Of note he was compliant at this time with Rivaroxaban. The patient was started on unfractionated heparin drip and subsequently admitted. On admission, his vital signs showed a blood pressure of 138/93, heart rate 65 bpm, and respiratory rate 16. Cardiopulmonary examination revealed regular rate and rhythm, without murmurs, rubs or gallops and his lungs were clear to auscultation. Neurologic examination revealed intact cranial nerves, preserved strength in all extremities, mild dysmetria in the left upper extremity and an NIH score of 1. Electrocardiogram revealed normal sinus rhythm, non-specific ST changes and PVCs. Transthoracic Echocardiogram revealed normal left ventricular function with mild tricuspid regurgitation, and stage 1 diastolic dysfunction. Transesophageal echocardiogram was negative for intracardiac thrombus. Echocardiography with agitated saline contrast noted microbubbles entering the left atrium from the left lower pulmonary vein indicative of Intra-pulmonary A-V Shunt. CTA of the chest revealed a connecting pulmonary vein and artery in the left lower lobe. Unfractionated heparin was continued and the patient was transferred to a tertiary center for embolization of the shunt. There the patient developed recurrent strokes while anticoagulation was temporarily on hold. CT of the abdomen for persistent abdominal pain noted an omental mass and the patient subsequently underwent exploratory laparotomy and was found to have an omentum adenocarcinoma with nodular peritoneal implants, however the primary is unknown. The patient was placed on enoxaparin, scheduled for chemotherapy. Embolotherapy of the intrapulmonary shunt was deferred. This case illustrates the embolic effect of intrapulmonary AV shunt and the characteristic appearance in those with paradoxical emboli. The main complications are hypoxia, stroke and brain abscess, as the capillary bed within the pulmonary vasculature is bypassed. Classic symptoms of dyspnea, clubbing and cyanosis are only seen in a small population of patients. These may be due to hereditary or idiopathic phenomena. Appropriate embolization, follow-up for recannulization and antibiotic prophylaxis for dental procedures is warranted.</td>
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<td>Unusual presentation of young female with SLE, Cardiomyopathy, and multi-vessel coronary aneurysms</td>
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Systemic Lupus Erythematosus is a chronic inflammatory autoimmune disease affecting multiple organ systems. Cardiac manifestations of SLE include pericarditis, myocarditis, nonbacterial endocarditis, coronary arteritis, and premature CAD. Coronary artery aneurysms (CAA) are rare among all patients undergoing coronary angiography with an estimated incidence of 0.3-5%. CAAs are an even more unusual finding in SLE patients. There have been only 17 cases reported, most with single vessel involvement in the right coronary artery. A 25-year-old African American female with a medical history significant for SLE for 10 years, CVA, and hypertension presented to the hospital with complaints of worsening orthopnea for 3 days associated with decreased appetite and generalized body aches. Physical exam was significant for bilateral basilar crackles and chronic left upper extremity weakness. BNP was elevated at 2455 and a chest x-ray was consistent with congestive heart failure. 2-D echocardiogram showed severe global left ventricular hypokinesia, estimated ejection fraction of 20-25%, and estimated pulmonary pressure of 47mmHg. A previous echocardiogram 3.5 years ago showed normal left ventricle systolic function and normal pulmonary artery pressure. The patient responded well to aggressive diuretics. Further workup included cardiac catheterization which revealed multiple large coronary aneurysms in the left circumflex and right coronary arteries, and mildly ectatic left anterior descending artery. A thrombus was also suspected in the right coronary artery. After consultation with rheumatology, large doses of pulsed steroids were started in addition to anticoagulation and antiplatelet therapy, as well as medical treatment for cardiomyopathy including beta blockers and ACE inhibitors. Further planned investigation will include cardiac MRI and possible endomyocardial biopsy. The patient will also need a prophylactic AICD implantation if there is no significant improvement in myocardial function after three months of optimal medical therapy. The most common cause of coronary aneurysms in adults is atherosclerotic disease. Other etiologies include Kawasaki disease, Marfan syndrome, Subacute bacterial endocarditis, Takayasus arteritis, Rheumatic fever, Mycosis, Syphilis, cocaine abuse, previous balloon angioplasty, and use of stents. This was a case of an unusual presentation of SLE, severe cardiomyopathy, and severe aneurysmal multi vessel coronary disease. Further workup and investigation may clarify the true etiology of this combination of cardiac manifestations and its relationship to SLE. Coronary artery bypass surgery and heart transplantation may be considered and/or needed for this patient in the future.

INTRODUCTION: Drug induced cholestatic liver disease is a subtype of liver injury that is characterized by predominant elevations of alkaline phosphatase and bilirubin secondary to the administration of a hepatotoxic agent. It can manifest itself as a cholestatic hepatitis or as bland cholestasis, depending upon the causative agent and the mechanism of injury. Drugs that typically cause cholestasis with hepatitis include psychotropic agents, antibiotics and nonsteroidal anti-inflammatory drugs (NSAIDs). Pure cholestasis without hepatitis is observed most frequently with contraceptive and androgenic steroids. We describe a case of cholestatic liver injury in a young individual following ingestion of over-the-counter muscle building supplement. Description: A 29-year-old male with no past medical history was hospitalized with rapid onset of generalized jaundice and severe pruritus. His workup showed markedly elevated total bilirubin to 11.6 mg/dl, direct 9.3 mg/dl, ALAT 121 U/L, ASAT 78 U/L, Alkaline phosphatase 552 U/L, albumin 3.5 g/dl, INR of 1.2, normal CBC and BMP parameters. His hepatitis serologies were negative. Tests for autoimmune hepatitis, primary biliary cirrhosis, sclerosing cholangitis, hemochromatosis and Wilson’s disease were negative. Ultrasound of the abdomen, MRI of the abdomen as well as MRCP were unremarkable. Upon questioning he stated that following an advice of his instructor at the local gym he used EPG “Tri-methyl platinum” dietary supplement for 2 month prior to the admission. It was suspected that patient developed drug-induced liver injury. His total billirubin continued to rise, reaching the peak of 26.1 mg/dl, however INR remained in 1.2 range. Liver biopsy was performed and showed canicular cholestasis with acute inflammation. He
was given symptomatic treatment including diphenhydramine, ursodesoxycholic acid and taper dose of prednisolone. His bilirubin, ALT, AST gradually decreased to normal values over 3 months. Puritis and jaundice successfully resolved as well. Discussion: While using dietary supplements among people involved in bodybuilding is extremely common, general public is usually not aware of the potential side effects as well as actual content of so-called “supplements”. Anabolic steroids are prohibited for non-prescriptional use, but some manufacturers are trying to market “pro-steroids or pro-hormones” which do not fall under strict regulatory scrutiny as regular steroids. However, the actual substance of these “supplements” is not very different from 17α-alkylated androgenic steroids, commonly known to use cholestatic liver injury. Supplement, which was used by our patient, contained 2 methylated derivatives of androgens. Use of such agents can lead to prolonged hepatic dysfunction with cholestatic syndrome that resembles primary biliary cirrhosis. To our mind, public awareness on such matter should be raised, as well as additional research should be performed.

### 5. Clinical Vignette

| 259 | Garabedian, Garo | Vadzim Chyzhyk, Jay Shastri | Atlanticare Regional Medical Center (Dominik Zampino) | A rare case of adult Henoch-Schonlein Purpura nephritis |

Henoch-Schonlein purpura (HSP) is a rare occurrence in adults. We report a case of suspected HSP with renal involvement in a Chinese 45 year old female, who presented with a painful rash of a few days’ duration over her lower extremities, initially described as "spots of blood". At the time of her presentation, she also complained of swelling in her legs and right hand. She did not have any fever, abdominal pain, joint pain or hematuria. On physical examination, she was found to have a bilateral lower extremity purpuric rash, dark red in color, non-blanching, from her ankles up to her mid-thighs. The surrounding skin was normal, and there were no other cutaneous involvement noted elsewhere. She was normotensive. Urine protein quantification was more than 10 g/dL. Her urine analysis also showed a few red blood cells. Laboratory workup was negative for viral hepatitis and cryoglobulinemia. Antineutrophil cytoplasmic and antinuclear antibodies were negative. Complement levels were normal. Serum protein electrophoresis was unremarkable except for low serum albumin and serum total protein. Her serum creatinine remained normal. Renal biopsy showed IgA predominant membrandoproliferative process, which together with her pupura made Henoch Schonlein her likely diagnosis. In view of the biopsy findings, the patient was started on oral prednisone, 60 mg daily. Her urine protein dropped, her edema improved and her purpuric rash resolved. Her creatinine remained around baseline. She is still following with her nephrologist. As per most recent laboratory workup, her urine protein is down to 258 mg/g of urine creatinine. Her oral prednisone regimen was tapered progressively in light if her decreasing proteinuria. HSP is a leukocytoclastic vasculitis involving small vessels, characterized by the deposition of immune complexes containing IgA. It is classically manifested as a purpuritic vasculitic rash involving lower extremities and buttocks, abdominal pain and arthragias. Renal involvement is more common and more severe in adults, resulting in deterioration of renal function in most patients, over years following initial presentation. One study notes 20% progression to end-stage renal disease. Treatment of HSP nephritis involves high dose steroids. Cyclophosphamide and other immunosuppressants do not show clear benefit.

### 6. Clinical Vignette

| 290 | Hanna, Bishoy | Kruti Parikh MD, Gurpreet Singh MD, Magdalena Szulc MD, Marcela Roger MD | Atlanticare Regional Medical Center (Dominik Zampino) | Hypertriglyceridemia, Pancreatitis and Diabetic ketoacidosis: A Familiar Triangle Seen in a New Angle |

Hypertriglyceridemia is the third most common cause of acute pancreatitis. It is reported to cause 1-4% of all cases of acute pancreatitis. The presence of DKA in the setting of hypertriglyceridemia induced pancreatitis is less common. Here, we report a case of DKA in a patient with hypertriglyceridemia induced pancreatitis treated with apheresis. A 36-year-old Hispanic man with past medical history of diabetes, hypertriglyceridemia, and tobacco abuse presented to the hospital with a chief complaint of non-
Radiating epigastric abdominal pain for 1 day. His pain was crampy in nature, 10/10 in severity, associated with nausea and decrease in appetite but no vomiting or diarrhea. The patient’s diabetes and hypertriglyceridemia were diagnosed one year earlier. He received treatment, including medications while in jail; however, subsequently stopped his treatment after release. On examination, the patient was well built, afebrile, hemodynamically stable and in no acute distress. He had mild tenderness in the epigastric region with some involuntary guarding but no rebound. The patient was noted to have diffuse erythematous papules approximately 0.5 cm in diameter scattered over his entire body; most prominent on the torso and anterior aspect of the right upper and lower extremity. The patient’s laboratory studies revealed a triglyceride level above 4000 mg/dl, blood sugar of 343 mg/dl, sodium of 131 with a corrected value of 133 mEq/L, potassium of 3.4 mEq/L, CO2 of 19 mmol/L with an anion gap of 24, and lipase of 1066 U/L. Incidentally, the blood specimen was noted to be lacticest. CT of the abdomen and pelvis with contrast showed mild swelling of the distal pancreatic body and tail with inflammatory stranding of the fat around the pancreas; consistent with acute pancreatitis. The diagnosis of acute pancreatitis secondary to hypertriglyceridemia and DKA was made. The patient was subsequently admitted for Diabetic Ketoacidosis and acute pancreatitis secondary to hypertriglyceridemia. Treatment included intravenous fluids, insulin, bowel rest and potassium supplementation. Over the next 24 hours, the patient’s symptoms did not improve and his anion gap did not close. Given his severe hypertriglyceridemia a decision was made to start the patient on apheresis. Following two cycles of apheresis, the patient’s symptoms subsided and his triglycerides came down to 298 mg/dl at the time of discharge. He was discharged on gemfibrozil and insulin with close follow-up. In DKA, insulin deficiency activates lipolysis in adipose tissue releasing free fatty acids and reducing lipoprotein lipase activity thus decreasing VLDL removal resulting hypertriglyceridemia. Although moderate hypertriglyceridemia is common during episodes of DKA; severe hypertriglyceridemia, defined as a TG level >2,000 mg/dL, is rare. In this case, rapid treatment of hypertriglyceridemia was achieved using apheresis and insulin simultaneously.

7. 301 Clinical Vignette

Hasan, Rimsha
Inga Robbins, MD
Atlanticare Regional Medical Center (Dominik Zampino)

Serendipitous finding of Giant Right Coronary Artery Aneurysm following Motor Vehicle Accident

Coronary Artery Aneurysms are not uncommon however giant aneurysms are reported with a prevalence of 0.02%. Giant Aneurysms are defined as dilatation that exceed 8mm or are 4 times the reference vessel diameter. Evidence based guidelines are limited and possible complications include rupture, thrombosis, embolization, dissection and mechanical obstruction. The presentation can vary warranting proper use of imaging and invasive techniques for diagnosis and treatment. Here we report a rare case of a giant RCA aneurysm that measured 5.7 x 6.3 cm and was discovered incidentally after motor vehicle accident. A 38 year old African American male with past medical history of Asthma presented after motor vehicle accident. Patient was evaluated following trauma series protocol that did not reveal any fractures. CT Scan of the Chest was done which revealed 5.7x6.4x6.3 cm right atrial cystic mass with mural calcifications. The electrocardiogram was consistent with Inferior wall infarction with left ventricular hypertrophy. Patient underwent 2D- echocardiogram which revealed Mild to Moderate enlargement of LV cavity size, moderate global hypokinesis involving all segments of the left ventricle. Left ventricular ejection fraction was visually estimated at 45%. cystic mass compressing right atrium was noted. No significant pericardial effusion was seen. Further history revealed that patient had episodic palpitations and rapid irregular heart beats associated with dizziness and diaphoresis. Patient denied any history of syncope. Holter monitor revealed normal sinus rhythm with heart rate between 48 to 85 beats per minutes with Isolated PAC’s and PVC’s. The patient had a subsequent Transesophageal echocardiogram which revealed ejection of 35%, large 6 x 5 cm mass with the cystic spaces present lateral to the right atrial wall with no apparent direct vascular connection to the right atrial chamber mass as noted by color-flow imaging. The patient had a cardiac catheterization subsequently with EF of 41%, inferior basilar akinesis of the left ventricle wall,
8. Clinical Vignette

Hasan, Rimsha Jingsheng Zheng, MD, John Saia, DO, Patrick O’Beirne, MD, Kenneth Khaw, MD, Gerald Ukrainski, MD

Atlanticare Regional Medical Center (Dominik Zampino)

A Rare Case Report: Apical Thrombus Mimicking Cardiac Myxoma

Echocardiography is the most common imaging modality to visualize cardiac mass. However, sometimes it is difficult to distinguish between thrombus or myxoma. Other imaging modalities should be used to delineate detailed anatomy of the cardiac mass. A 63-year-old white male with past medical history of CAD, myocardial infarct and CABGx4, was found to have a “cardiac mass” by a routine 2D echocardiogram. Two-D echocardiogram revealed a large (1.4 cm × 0.9 cm), ovoid and heterogeneous mass (large arrow) in left ventricle attached with a long narrow stalk (small arrow) to apex. Definity contrast echocardiogram revealed a non-opacified contrast defect of LV apex. Patient underwent a cardiac MRI, which revealed a nonenhancing rounded thrombus (measuring 10 mm) within the apex. There is left apical thinning/aneurysm with dyskinesis. Patient was treated with Coumadin for anticoagulation. He is doing well with current regimen.

9. Clinical Vignette

Hasan, Rimsha Nanda Nair, MD, Haitham Dib, MD

Atlanticare Regional Medical Center (Dominik Zampino)

Left ventricular pseudoaneurysm- A Rare Cause of chest pain.

Left ventricular pseudoaneurysms were traditionally described post myocardial infarction, surgery and trauma. However with recent advances in aortic valve replacement the incidence is on the rise. Common symptoms at the time of presentation include chest pain, heart failure, dyspnea and upto 10% patient can be asymptomatic. LV pseudoaneurysms are prone to rupture and a timely diagnosis is of the essence. We describe here a rare case of left apical pseudoaneurysm in an 84 year old patient post transapical aortic valve replacement. An 84 year old Caucasian male with medical history of severe aortic stenosis who underwent aortic valve replacement with an Edwards Sapien XT #26 valve via a transapical approach with additional history of diastolic congestive heart failure, severe multivessel coronary artery disease, status post CABG presented with complaint of chest pain. His symptoms were exertional and associated with dyspnea that progressively worsened over a course of one week. Electrocardiogram was done which revealed, diffuse ST-elevation with PR segment depression consistent with acute pericarditis. Patient underwent 2D-echocardiogram which revealed EF of 71%, akinetic echolucency adjacent to the apex with a narrow neck of color flow contiguous with the LV cavity. Echo contrast did not pass to the echolucent space, a finding concerning for LV apical pseudoaneurysm. Bioprosthetic AV was well seated with no obvious aortic insufficiency. Patient then had CT Angiography of the heart which revealed Left ventricular apical pseudoaneurysm, approximately 1.3 x 1.4 cm in cross-sectional diameter at the neck with the maximum cross-sectional diameter of the pseudoaneurysm of approximately 3.1 x 2.5 cm. Additional finding of a small pulmonary embolus within a branch of the right pulmonary artery leading into right lower lobe was noted. After careful discussion surgical repair was opted for. Patient was taken to hybrid cardiac catheterization laboratory / operating room for a surgical repair. Left Anterior thoracotomy with repair of left ventricular pseudoaneurysm was done. On final ventriculography there was no residual pseudoaneurysm seen. All contrast filled.
areas of the LV were trabeculated consistent with myocardium. Patient was extubated successfully however his post operative course was complicated by pulmonary embolism. He then had a cardiac arrest and family decided to withdraw care.

**INTRODUCTION:** Sickle cell crisis occurs when vaso-occlusion from sickled red blood cells leads to hypoxemia and single or multi-organ dysfunction. Patients may present with severe pain in their chest, abdominal, bone or joints or organ specific symptoms from shock liver and acute renal failure. The most common organ systems discussed in sickle cell crisis patients are lung, liver, bone, and renal. Renal dysfunction often leads to uremia which can lead to pericarditis and cardiac tamponade. Hypotension in the critically ill patient can reflexively be attributed to infection, especially in sickle cell patients with compromised immunity due to asplenia. We present a case where hemodynamic stability was restored through pericardiocentesis of a large pleural effusion causing cardiac tamponade in a sickle cell crisis patient.

**CASE DESCRIPTION:** In this case we treated a 27 year old female who presented with persistent left back and flank pain along due to sickle cell crisis who underwent multiple blood exchange transfusions and soon thereafter developed shortness of breath and tachypnea requiring mechanical ventilation. In the ICU this patient suffered from multi-organ dysfunction including shock liver and acute kidney injury likely secondary to hypoxemia from sickled red blood cells. The subsequent uremia was thought to have caused to pericarditis and pericardial effusions leading to cardiac tamponade and hemodynamic instability. Echocardiography was performed and confirmed cardiac tamponade with large pericardial effusions, diastolic compression of the right ventricle and dilated IVC without respiratory collapse. Intervention was performed in the form of pericardiocentesis removing 675 cc of exudative fluid via percardiocentesis. Post pericardiocentesis echocardiography demonstrated restoration of diastolic filling and normal cardiac function. Renal function and liver function improved over the course of the next and patient was successfully extubated and decannulated from tracheostomy collar. After a short course of rehabilitation this patient recovered from sepsis and multi-organ failure which included shock liver, acute kidney injury, bacteremia, pericarditis and successfully identified and treated cardiac tamponade.

**DISCUSSION:** Awareness of the potential cardiac manifestations in sickle cell patients can help guide appropriate management of sickle cell patients exhibiting hemodynamic instability such as this case seen at Atlanticare Medical Center in August 2014. Cardiogenic shock due to cardiac tamponade requires a different treatment paradigm than shock due to sepsis which can often be the presumed diagnosis in sickle cell patients with compromised immunity due to asplenia. Non-invasive testing such as echocardiography can help practitioners evaluate for uremic pericarditis and cardiac tamponade in sickle cell crisis patients and provide early intervention.

**Clinical Vignette**

**Probenecid as adjunct therapy with Nafcillin for the treatment of Methicillin-sensitive Staphylococcus aureus Endocarditis**

Probenecid was initially developed to competitively inhibit the renal excretion of penicillin antibiotics, significantly affecting the pharmacokinetics and pharmacodynamics – increasing plasma concentrations and prolonging the duration of action. Infectious Disease Society of America (IDSA) current treatment guidelines for endocarditis do not mention probenecid as a treatment option.
Nafcillin with the optional addition of gentamicin is the IDSA recommendation for treating endocarditis from methicillin-sensitive staphylococcus aureus (MSSA) in the absence of prosthesis. Probencid has a significant effect on nafcillin disposition resulting in decreased renal and non-renal clearance, and a decrease in its volume of distribution. The addition of probencid to nafcillin therapy results in a twofold increase in nafcillin plasma concentrations and doubles the area under the curve (AUC). A 26 year old Caucasian female with past medical history of tricuspid valve endocarditis and intravenous drug abuse presented with change in mental status, subsequently diagnosed with stroke and endocarditis. On presentation she was aphasic with right sided hemiparesis and temperature of 103.5°F. Day 1: Empiric vancomycin and cefepime were initiated after obtaining blood cultures. Day 2: Transesophageal echocardiogram (TEE) revealed a moderate sized vegetation on mitral valve and tricuspid valve. Computed tomography (CT) of chest showed bilateral septic emboli to lungs. Acute left middle cerebral artery infarct was noted on head CT. Day 3: Blood cultures 4/4 bottles revealed MSSA (oxacillin MIC= 0.5). Antibiotics de-escalated to nafcillin 2g IV Q4h. Day 5: Patient was deemed unsuitable for valve replacement surgery due to recent stroke and ongoing polysubstance abuse. Day 6: Repeat blood cultures 2/2 bottles remained positive for MSSA. One dose of gentamicin 3mg/kg was added for synergy. Day 7: Probencid 500mg Q6h was added due to persistent positive blood cultures. Day 12: Blood cultures results from Day 9 were reported as sterile in 2/2 bottles. Day 49: Patient completed 6 weeks of IV nafcillin with oral probencid and was discharged. Infections such as endocarditis often require larger and more frequent antibiotic doses in order to obtain optimal antibiotic concentrations and penetration, particularly in younger individuals with accelerated renal and non-renal drug clearance. Probencid significantly inhibits nafcillin clearance resulting in higher serum concentrations, a greater AUC, and a greater percentage of time the drug concentrations are above the MIC. This case demonstrates the value of probencid in a patient with a very high clearance (CrCl > 120mL/min). Forty-eight hours after initiating of probenecid, blood cultures sterilized. The addition of probenecid to nafcillin should be considered a treatment alternative when higher and more sustained antibiotic serum concentrations are desired.

12. 272 Clinical Vignette Nwankwor, Ifeoma Dr kruti Parikh, Dr Mathew Corcorran Atlanticare Regional Medical Center (Dominik Zampino)

A review of the Cardiac manifestations/Complications of Acromegaly, their management and outcomes

Acromegaly is a disorder characterized by Growth hormone (GH) hypersecretion, most commonly GH –secreting pituitary adenoma. Patients with Acromegaly have an increased risk of Type 2 Diabetes mellitus, cardiovascular disease, hypertension, hypertrophic cardiomyopathy and atherosclerotic arterial disease. They have a 30% higher mortality rate, with cardiovascular disease accounting for 60% of deaths. Evidence suggests that early diagnosis and treatment of Acromegaly (before the age of 40) can help prevent the progression of cardiovascular disease, improve quality of life and reduce the risk of premature mortality. A 27 year old Bangladeshi female with a past medical history of GH secreting pituitary adenoma s/p surgery and radiation is admitted three times within a span of two months; for complaints of recurrent nausea, vomiting and headaches. Hormonal assay was consistent with panhypopituitarism and increased IGF-1. An oral glucose tolerance test showed a markedly elevated GH response: >5ng/ml consistent with the diagnosis of Acromegaly. MRI showed an empty appearance of the pituitary fossa. Patient was treated with a trial of cabergoline and other hormones were replaced accordingly. However, patient was lost to follow up: she had traveled to Bangladesh and missed several appointments, and she also could not afford her medications due to lack of health insurance. She then showed up at the clinic, 17 months later with complaints of dyspnea. She was found to have elevated JVD, bibasal crackles, S3 gallop, and a grade 2 bilateral pitting pedal edema. Systolic blood pressure was 150, hemoglobin A1C was 8.5, and ECHO showed EF of 20%, severe global hypokinesia involving all segments of the left ventricle, severe enlargement of the LV cavity size, severe LV systolic dysfunction, stage 3 diastolic dysfunction, moderate mitral regurgitation and left atrial enlargement. A diagnosis of Congestive heart failure was made and she was aggressively managed with furosemide, lisinopril, metformin, Life vest and later...
carvedilol. At this time, octreotide injections were also started. Repeat ECHO done 4 months later showed marked improvement in LV function with normal LV size, normal LV thickness, no regional wall motion abnormalities, EF of 45 % and stage 1 diastolic dysfunction. IGF -1 and A1C has been trending down, patient has been asymptomatic and is being closely followed in the clinic. GH and IGF-1 both have regulatory roles in the cardiovascular system and when in excess, can lead to myocyte hypertrophy, interstitial fibrosis and both systolic and diastolic dysfunction. We emphasize the importance of early control of GH and IGF-1 excess; in ameliorating cardiac abnormalities and leading to a significant reduction of left ventricular hypertrophy with improvement in cardiac performance. Radiation and medical therapy can also be used as an adjunct to surgery in patients with residual disease.

A Complication of a Port-a-cath venous access leading to critical care admission

Port-a-cath venous access devices are commonly used for delivery of chemotherapeutic agents. Although this is a low risk procedure, rare complications have been reported. One possible complication is hemothorax. 63-year-old female with a significant history of tobacco smoking initially presented with gradually worsening cough and shortness of breath. CT chest showed a large hilar mass in right upper lobe, and massive right pleural effusion with mediastinal adenopathy. Ultrasound guided thoracentesis with right pleural fluid aspiration and cell block revealed Small Cell Lung Cancer with malignant pleural effusion and extensive liver metastasis. A decision was made to treat the patient with chemotherapy by Oncology and placement of a chemo port was planned. This procedure was performed under local anesthesia with sedation in the operating room. During the final stages of the procedure, patient became hypotensive and tachycardic. She then became bradycardic and eventually lost pulse. Resuscitative measures performed while the surgeon completed the procedure. However, patient went into cardiac arrest and she was resuscitated with chest compressions and vasopressors. The patient regained regular rhythm in less than five minutes of Cardiopulmonary Resuscitation. On the post procedure bedside chest X-ray, the catheter location was uncertain and there was an enlarging right pleural effusion. Thoracentesis was performed and a Pigtail catheter was placed which drained frank blood. Patient was intubated, placed on mechanical ventilation, and transferred to the Intensive Care Unit. CT Angiography of the Chest demonstrated the catheter extravasatorily in the right pleural space. A tremendous right pleural effusion, which appeared to contain hemorrhage, was evident. The steps were taken for immediate interventions and the port catheter was retracted under fluoroscopy. The site of vessel injury was identified. Following this, a stent graft was deployed at the site of injury. Post stent balloon dilatation was performed achieving hemostasis. A chest tube was placed and 2.5 L of bloody effusion was drained. Patient’s condition markedly improved and hemodynamic stability was achieved after these interventions. She did well and successfully weaned off the ventilator and extubated the following day. This case demonstrates the possibility of serious outcomes of the port-a-cath venous access including cardiac arrest requiring critical care admission. Although very rare, these complications do occur and awareness of such outcomes can prevent accidents and improve patient safety.

The Curious Case of Von Hippel-Lindau disease

Von Hippel- Lindau Disease (VHL) is a disease in which about 80% of the cases are due to an inherited mutation of the VHL gene and the rest are found in individuals without a family history, known as de novo mutations. Families with VHL disease are divided into 2 types based on the likelihood of developing pheochromocytoma. Type 1 has a lower risk of developing pheochromocytoma compared to Type 2. Type 2 is subdivided based on risk of renal cell carcinoma. Type 2A has a lower incidence of RCC compared to Type 2B. Patients with pheochromocytoma associated with VHL tend to not have symptoms or biochemical evidence of
catecholamine production compared to those without VHL. We report a case of a young male with reported symptoms of palpitations, chest pain, headaches and documented uncontrolled hypertension, with a diagnosis of bilateral renal masses. This is a 39 year old African American male with pmh of uncontrolled hypertension since his 20’s (on 6 medications). Congestive Heart Failure, Cardiomyopathy, CKD, who repeatedly came to the hospital on various occasions for symptoms of chest pain, palpitations, headaches, blurry vision, dizziness, and CHF symptoms. On initial exams, patient was in no acute distress. He had noted tachycardia, regular rhythm, mild +1 lower extremity pitting edema, no JVD and fine rales at both bases. Cranial nerves were intact. Laboratory findings showed 24 hr. urine metanephrines of 457ug and 24 hr. urine normetanephrines of 1737ug. Repeated 24 hr urine studies showed 317ug metanephrines and 933ug normetanephrines. CT abdomen followed by MRI of the abdomen showed bilateral renal masses. Patient has upcoming MIBG scan. This patient likely presents with Type 2B VHL disease with bilateral renal masses suspicious for renal cell cancer with likely associated pheochromocytoma. This patient presents with symptoms and biochemical evidence of catecholamine production, which is a less likely presentation as compared to pheochromocytoma in patients without VHL disease.

### Clinical Vignette

Singh, Gurpreet Lalith Premachandra, MD, Robert Matthews, MD

**Atlanticare Regional Medical Center (Dominik Zampino)**

**Multiple Myeloma Masquerading as Acute Back Pain in a 45 Year-Old Male**

**INTRODUCTION:** Multiple Myeloma is a neoplastic proliferation of plasma cells that produces a monoclonal immunoglobulin which can lead to skeletal destruction and accumulation of paraprotein in the kidney’s resulting in acute renal failure. The diagnosis is accompanied by a constellation of symptoms including osteolytic bone lesions, increased total serum protein, presence of monoclonal protein in serum or urine, anemia, and hypercalcemia.

**CASE DESCRIPTION:** A 45 Y/O M with no significant PMH presented to the ER with the chief complaint of lower back pain. The patient reported that he had initially sustained a fall from a standing position 2 weeks earlier and was evaluated in a New York City hospital at which time X-Ray imaging of the back revealed no significant findings, results of an MRI were pending. On his second presentation to the ER for worsening back pain, he was found to be in acute renal failure with a serum creatinine of 9.0 (last known baseline from 6 years prior was 1.5), hypercalcemia (13.7) and normocytic normochronic anemia (Hemoglobin: 7.8). A CT Scan of the spine revealed numerous lytic lesions across the thoracic, lumbar, and sacral spines. The patient was started on aggressive intravenous fluid hydration, decadron, and calcitonin. A full battery of serological tests were ordered including HIV/Hepatitis B,C, RPR screening, C3, C4 complement levels, serum and urine protein electrophoresis, and parathyroid hormone levels amongst other studies. Subsequently, a renal biopsy was performed and revealed lambda light chain restricted casts suggestive of myeloma cast nephropathy. The urine protein electrophoresis studies were also noted to be positive with an M spike-lambda bence jones protein. In addition a bone marrow biopsy confirmed the presence of a plasma cell neoplasm with lambda light chain restriction. The patient’s renal function stabilized as his serum creatinine leveled to 2.5-2.8 during the hospitalization, and he did not require any renal replacement therapy. The patient has since been started on a chemotherapy regimen of velcade, dexamethasone, and cyclophosphamide. Subsequent monthly follow ups have revealed stable renal function and labs.

**DISCUSSION:** This case represents a common clinical presentation of acute back pain which masqueraded the relatively rare occurrence of multiple myeloma in a 45 year old male. Multiple Myeloma accounts for 1 percent of all cancers. Its incidence is 4 to 5 people per 100,000 in the United States and Europe. Furthermore, this disease is typically known to be a neoplasm of the elderly, with a median age of 66 years at the time of diagnosis. Only 10 percent of patient’s present prior to the age of 50, and 2 percent
Severe Nephrotic Proteinuria in the setting of Primary Membranous Glomerulopathy

**INTRODUCTION:** Membranous Nephropathy (MN) is one of the most common causes of nephrotic range proteinuria in adults as it accounts for approximately one-third of biopsy proven cases. The syndrome is associated with significant morbidity and mortality with higher levels of proteinuria correlating with a greater risk of long term end stage renal disease. Complications resulting in increased morbidity and mortality are attributed to thromboembolic and cardiovascular disease.

**CASE DESCRIPTION:** A 19-year old male with no significant past medical history presented to the emergency department with the chief complaint of periorbital swelling, lower extremity edema, and associated fatigue that had been increasing over the last two weeks. On admission to the hospital, blood pressure was notably elevated (151/99), his physical exam was significant for periorbital edema, and 4+ pitting edema in the lower extremities bilaterally. Lab work revealed a serum creatinine of 0.7, albumin of 1.1, total cholesterol of 305, and a 24hr urine protein level of 42.9 grams. In view of the patient’s presentation, a CT guided renal biopsy was scheduled. Findings revealed IgG4 predominant primary membranous glomerulopathy. Treatment options were reviewed with the patient, and he declined to start cyclophosphamide therapy in view of side effects related to infertility. In this case, the patient was started on cyclosporine 75mg BID and prednisone 10mg daily for a 6 month period. In addition, the patient was also started on lisinopril 10mg daily and furosemide 40mg every other day. On the next visit one month later, the patient’s 24 hr urinary protein and serum albumin levels were 5.4 grams and 2.5 respectively. This further improved to 495 mg of 24hr urinary protein and serum albumin of 4.3. The patient reported returning back to near his baseline weight, improved energy, and stamina. Achievement of complete remission was established with this therapeutic trial.

**DISCUSSION:** This case illustrates the implementation of a calcineurin inhibitor plus glucocorticoid therapy in a 19 year-old patient with severe nephrotic proteinuria (>40 grams over 24hrs on admission). Complete remission was achieved and the patient’s dosage of lisinopril was decreased and furosemide was discontinued. Despite the aforementioned regimen having a complete remission success rate of 75-80%, there is a significant relapse rate of close to 45%. The patient was advised to adhere to a low salt diet, and follow up on a monthly basis for continual monitoring.

Clarkson’s Disease: A rare potentially fatal clinical syndrome with undetermined pathophysiology

Idiopathic Systemic Capillary Leak syndrome (SCLS), also known as Clarkson’s disease is a rare and potentially fatal disorder of unknown etiology. It is characterized by episodes of profound hypotension, generalized edema, hemoconcentration and hypoalbuminemia without albuminuria. Fewer than 150 cases have been described in the literature, since it was first reported in 1960, with a preponderance for middle aged white males. We illustrate a case of an acute SCLS episode in an otherwise healthy young Hispanic male. A 24 year old male presented to the emergency department with chest pain and shortness of breath. The patient was diagnosed with SCLS two years ago and was maintained on Intravenous Immunoglobulin (IVIG) infusions every two
weeks. On presentation, the patient was tachycardic, tachypneic, hypotensive, and afibrile. He had dry mucus membranes, delayed capillary refill, cool extremities and diminished peripheral pulses. Electrocardiogram showed sinus tachycardia without ischemic changes. He received three liters of 0.9% normal saline fluid boluses, and was continued on aggressive fluid resuscitation, with concurrent albumin and IVIG infusions. A Transthoracic Echocardiogram obtained to evaluate his cardiac function showed preserved left ventricular ejection fraction (LVEF of 60%). Although the patients’ blood pressure improved with crystalloid and colloid resuscitation, he remained tachycardic and tachypneic, eventually becoming hypoxic. A Chest X-ray revealed pulmonary vascular congestion. Pulmonary embolism was ruled out with a normal ventilation-perfusion scan. Intravenous fluids were discontinued and diuretic therapy was initiated with additional support of Bi-level positive airway pressure. Despite these measures, the patient’s respiratory status continued to deteriorate requiring intubation, mechanical ventilation and transfer to the intensive care unit. There he received Levophed and aggressive diuretic therapy. The patient was successfully extubated on day 12 and subsequently discharged home on day 13. SCLS is a challenging entity owing to its rarity, uncertain pathophysiology, and lack of validated diagnostic criteria and therapeutic modalities. The syndrome has been described as having several distinct phases. The initial leak phase, consisting of hemoconcentration and hypovolemia and the post leak phase consisting of restoration of capillary barrier function with fluid mobilization from the tissues into the circulation and auto-diuresis. As our case demonstrates, cardiopulmonary failure can occur due to overzealous fluid resuscitation during the post-capillary leak phase. Early central venous pressure monitoring to guide fluid and catecholamine therapy through each phase may have been beneficial. Other documented complications include compartment syndrome, venous and arterial thrombosis and renal failure from hypoperfusion-induced acute tubular necrosis or myoglobinuria from rhabdomyolysis. The literature shows that steroids, terbutaline, aminophyline and immunoglobulins have been used with varying degrees of success. Further investigation to determine the underlying pathophysiology and perhaps target more adequate therapeutic approaches may prove beneficial in understanding the mechanisms behind this rare and potentially fatal condition.

18. Clinical Vignette

Vargas Jr., Jesus Kruti Parikh, MD; Sujitha Nandimandalam, MD and Nadia Sadik, MD

Atlanticare Regional Medical Center (Dominik Zampino)

Atypical Presentation of Unicentric Castleman’s Disease

Castleman’s disease (CD), also known as angiofollicular lymph node hyperplasia is a poorly understood disease consisting of proliferation of lymphoid tissue with or without constitutional symptoms. CD has been associated with HIV, HHV8 and excessive production of IL-6 or related polypeptides. The initial challenge in CD is ascertaining the diagnosis. Currently, there is no screening or early detection for CD, and the etiology to date is unclear. CD has been classified into 2 groups: a multicentric form and a unicentric form. The multicentric form is a systemic disease affecting more than one group of lymph node. The other form, Unicentric Castleman’s Disease (UCD) is a localized disease affecting a single group of lymph nodes and is the more common form of the disease. UCD present as a benign solitary mass that is commonly asymptomatic, and complete surgical resection is considered the gold standard for treatment and curative in majority of cases. We report an interesting and atypical presentation of UCD. A previously healthy 42 years old female presented with sudden onset of positional dyspnea, and an inability to lie on her left side or on her back with noted hypoxemia. Patient denied any history of fever, night sweats, weight loss, fatigue, dysarthria, dysphagia and no complaints of chest pain. A chest CT with contrast revealed a large left pleural effusion and an enhancing soft tissue mass in the left superior mediastinum, with lymphadenopathy inferiorly and superiorly in the anterior mediastinum. Ultrasound guided left thoracentesis produced a total of 2425cc of bloody fluid which resulted in improvement of her respiratory symptoms. A large bulky 17.0 x 7.5 x 7.0 cm hemorrhagic soft tissue mass that was intimately involved with the left subclavian and left carotid artery was
resected by trans-sternal approach. The tissue was positive for CD20, bcl1, bcl6, CD23, CD3, CD5 with few scattered cells positive for CD23, and Bcl-2 was negative. Flow cytometry showed no evidence of B-cell or T-cell lymphoproliferative disorder. There was no evidence of carcinoma, lymphoma, or thymoma and HHV-8 was negative. The pathology report indicated the mass displayed prominent hyaline changes with follicles showing regressed germinal centers and prominent onion skin-like mantle with morphologic and immunophenotypic features consistent of a hyaline vascular variant of Unicentric Castleman’s disease. The differential diagnosis for mediastinal mass is broad and although UCD is uncommon, it should be considered as a possible etiology.

19. 312 Clinical Vignette Vargas, Jesus Hussain, Asiya; Ashraf, Afia; Vijayakumar, Ashvin; Kaur, Tarandeep, Baptist, Justin; Roger, Marcela, Atlanticare Regional Medical Center

A ring enhancing cerebral lesion that is not a tumor: Tumefactive Multiple Sclerosis, a variant of MS

Inflammatory demyelination of the central nervous system (CNS) is caused by a spectrum of disorders, of which multiple sclerosis (MS) is the most common. Features of MS include focal demyelination, variable inflammation, gliosis and axonal preservation. MS is diagnosed clinically and radiographically by evidence of two separate areas of CNS demyelination at least 1 month apart. When demyelinating disease does not exhibit classic MS presentation and radiographic findings are indistinguishable from a neoplasm, it is described as Tumefactive Multiple Sclerosis (TMS), a rare variant of MS. TMS lesions are defined as a large (>2cm) lesion with little mass effect or vasogenic edema and ring enhancement on MRI. Diagnosing TMS is cumbersome because it can mimic an abscess, a primary or secondary neoplasm, a tuberculoma, necrosis, lymphoma or an inflammatory process such as sarcoidosis or Sjogren’s syndrome. We report a 37 year old female with a complicated history of migraine who presented with three episodes of stroke-like symptoms, progressive left sided hemiparesis leading to hemiparalysis which resolved within a couple of hours. Head CT displayed a focal heterogeneous 1.8 cm lesion on the right corona radiata, above the basal ganglia. CT perfusion studies indicated no difference in mean transit time, blood volume or blood flow. Therefore no occlusion to cerebral vessels or infarction to cerebral tissue was suspected. Due to the fact that the patient’s symptoms could have been explained by onset of complicated migraine, typical treatment of acute demyelinated disease consists of pulsed dose IV steroids was not initiated. Instead, a careful, systematic diagnostic approach consisting of lumbar puncture and neuroradiologic study including MRI of the CNS was performed. The ambiguous characteristic of the lesion prompted an MR spectroscopy study as well. MRI of the brain revealed a 1.8cm ring enhancing lesion located in the right corona radiata with no mass effect or edema. No other lesions were identified in the CNS. MRI Brain Spectroscopy demonstrated presence of lactate peak with decreased NAA, elevated choline peak and creatine peak was also significantly decreased, all suggesting cerebral tissue breakdown. Serologies including Toxoplasmosis, HSV, VDRL, and AFB were all negative. Cerebral Spinal fluid analysis disclosed elevated myelin basic protein but was negative for oligoclonal bands. When comparing the images from previous presentation, the spectra pattern is similar to that of prior studies without significant change. The clinical presentation and radiologic imaging suggests the lesion is demyelination of cerebral tissue. Careful review of serial images and recognizing findings suggestive for demyelinating lesion spared the patient from an invasive biopsy procedure for diagnosis. TMS is a diagnostic challenge therefore clinical judgment along, accurate assessment of the patient’s symptoms along with accurate MRI evaluation should be sufficient to make the diagnosis.

20. 201 Research Chyzhyk, Vadzim

Correlation between Body Habitus and lipid profile in healthy firefighters

Current 2013 ACC/AHA Guideline on the treatment of blood cholesterol focuses on treatments that are proven to reduce ASCVD events, however it is not by any means a comprehensive approach to lipid management. Some recent studies actually suggested that LDL particle number was more strongly related to incident CVD events than LDL-C. To our mind this could be a very interesting
hypothesis to test as with new 2013 ACC/AHA guidelines statin therapy reduces the risk of CVD, but do not completely eliminates it and additional factors as LDL-P need to be further evaluated. Doing advanced lipid testing is not encouraged by current guidelines and can be very costly affair for the patient and/or hospital if it’s done based on individual practitioner discretion. Thus, we were looking into other surrogate measurements of the LDL-P, triglycerides status which would allow us to assess LDL-P, triglycerides status in particular patient without doing actual blood work. Some reports have suggested that measuring hight-to-abdominal circumference ratio (HTACR) is better predictor of advance lipid particles status than BMI. We wanted to test this hypothesis and prove that there is negative correlation between HTACR and level of LDL-P and Triglycerides.

**SUBJECTS:** 132 healthy, non diabetic actively working male firefighters from two coastal towns were evaluated in the cardiology office with their BMI, hight, abdominal circumference measured. Advanced lipid profile test performed by LabCorp.

**RESEARCH METHODS:** Simple linear regressions using HTACR to predict the advanced lipid profile. Subjects were adjusted by age.

**RESULTS:** We found that HTACR has statistical significant correlation with level of LDL-P, small LDL-P and triglycerides with p<0.0001. Lower HTACR, higher expected levels of LDL-P, small LDL-P and triglycerides irrespectively of subjects’ age. There was no significant correlation found between levels of LDL-C and HTACR. HDL-C correlated positively with high HTARC with p<0.0001.

**DISCUSSION:** HTACR measurement is inexpensive tool that might be helpful in identifying subjects with adverse blood-lipids profile. It might be particularly important in during assessing the need to prescribe statin therapy for primary prevention to make appropriate decision about initiation of statin therapy of ASCVD in adults with LDL–C 70-189 mg/dL without clinical ASCVD or diabetes, in adults with LDL–C <190 mg/dL who are not otherwise identified in a statin benefit group, or for whom after quantitative risk assessment a risk-based treatment decision is uncertain (1). Another potential category are patients who achieved their LDL-C reduction goals with, but progression of the CVD is still evident. Based on results of HTARC measurements these patients might require additional attention from whether providers to help them reach their weight loss goals, improve physical activity, implement dietary modification and assess needs for further testing.

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21. 45  
Clinical Vignette  
Ahmad, Nazish  
Manish Gugnani MD  
Capital Health Regional Medical Center (Saba Hasan)  
It’s hard to tell the poison from the cure: nosocomial sepsis complicating lupus nephritis

**INTRODUCTION:** Blood cultures are a sensitive method for detection of bacteremia or fungemia. Sometimes, there can be a hidden source can hinder the patient’s progress, requiring further investigation and empiric therapy. Purulent pericarditis is a rare disease and is associated with a high risk for poor outcomes. We report a case of purulent pericarditis caused by Pseudomonas aeruginosa in a patient with MSSA bacteremia and active lupus.

**CASE:** A 37 year old African American female, recently diagnosed with Systemic Lupus Erythematosus with lupus nephritis and latent tuberculosis, was admitted for shortness of breath and chest tightness. Physical exam revealed a pericardial rub. CXR showed bilateral pleural effusions. Electrocardiogram was suggestive of pericarditis, and transthoracic echocardiography showed moderate pericardial effusion. Patient was started on colchicine and ibuprofen. The patient decompensated and became hypotensive and required emergent intubation and pressor use. Blood cultures reported gram positive cocci in clusters, later found to be methicillin sensitive staphylococcus aureus (MSSA), which was treated initially with vancomycin and then nafcillin. The
patient’s hypotension worsened and required the use of four pressors. Because of no improvement on current antibiotic therapy, a CT of the chest and abdomen revealed a worsening pericardial effusion. Heart sounds became muffled, jugular venous distension was noted, and emergent transthoracic echocardiocardiography confirmed a worsening pericardial effusion, with signs of tamponade. The patient’s antibiotic coverage was broadened to piperacillin-tazobactam. Urgent pericardiocentesis revealed 600 ml of purulent, yellow, thick fluid, suspicious for empyema. Unfortunately, the patient expired, and later cultures revealed pseudomonas aeruginosa.

DISCUSSION: Purulent pericarditis is an uncommon illness, and is usually secondary to a primary cause of pericarditis. We believe our patient had lupus associated pericarditis and developed line sepsis which was identified by blood culture to be MSSA. However, she failed to improve on therapy for MSSA, and ultimately pseudomonas pericarditis was identified. Most likely, the gram negative species was introduced by the same mechanism as the MSSA, as the patient had no evidence of nosocomial pneumonia. Complex, critically ill patients frequently have multiple processes occurring simultaneously and vigilant physicians must maintain a high index of suspicion for unusual and unexpected disease manifestations. Teaching points: Although blood cultures are accurate in establishing sources and therapeutic intervention, there is a chance a secondary source can remain unidentified. At times, a high index of suspicion requires more aggressive and invasive testing to identify all the processes at work in a complex patient.

22. Clinical Vignette

Ali, MD
Bipinpreet Nagra
Capital Health Regional Medical Center (Saba Hasan)

Man’s Best Friend? Zoonotic infections associated with canines

BACKGROUND: Zoonotic diseases are those that can be transferred between animals and humans. The CDC estimates that more than 6 out of every 10 infectious diseases in humans are spread from animals; fortunately the numbers are low in North America. The risk in humans becomes greater with immunosuppression. Skin and oral flora of dogs harbor various pathogenic microorganisms including Staphylococcus intermedius. Case reports of human infections from this microorganism are relatively rare, but the true incidence is unknown because the pathogen is frequently misidentified as Staphylococcus aureus. There are only 16 cases in the literature that have described S. intermedius as a cause of infection in humans ranging from soft tissue infections to bacterial endocarditis. Most of these cases have been described in association with exposure to animals, mostly dogs. We report a rare case of S. intermedius causing bacterial endocarditis in a dialysis patient. The presumed source of infection was the patient’s dog.

CASE: A 60 year old male with a history of diabetes, hypertension, pituitary mass, hypothyroidism and end stage renal disease on dialysis presented with complaints of fever, transient confusion and lethargy. The patient denied headache, neck pain, change in vision, nausea, vomiting, shortness of breath, recent fall, chest pain or palpitations. Patient has been on long term peritoneal dialysis and recently switched to hemodialysis for which he had tunneled dialysis catheter. There were reports of patient maintaining an intimate contact with his dog, where dog was licking him on several occasions. Patient had temperature of 102° F and tachycardia on arrival. Laboratory evaluation showed leukocytosis of 18.4 with 16% bandemia. CT of head showed no change in pre-existing pituitary mass and chest X-ray was normal. Blood cultures returned positive for S. intermedius and therefore transesophageal echocardiogram was done which showed a mobile vegetation which was attached to the posterior mitral leaflet measuring approximately 18mm. The patient was initially started on nafcillin resulting in a significant improvement in his condition. The patient was eventually discharged on cefazolin during which time he was asymptomatic and repeat blood cultures were negative for the microorganism. Patient was scheduled to follow up with cardiology in 2 weeks for a TEE.
**CONCLUSION:** Although very rare in human beings, diseases caused by S. intermedius should always be considered among the differential diagnosis of serious invasive infections, especially among patients who are in close contact with dogs.

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<th>Clinical Vignette</th>
<th>Al-Jumayli, Mohammed</th>
<th>Emily Chen, MD, Daniel Goldsmith, MD</th>
<th>Capital Health Regional Medical Center (Saba Hasan)</th>
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<td>Back pain in young active adult: beyond the usual suspects</td>
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**BACKGROUND:** The epidemiology of lower back pain in active adults indicates that most cases have a self-limited, nonspecific cause. Thus, one of the goals of the evaluation is to efficiently exclude potentially serious rare causes. In this report, we hope to illustrate the unique challenges in diagnosing a rare tumor in a young patient who complained only of lower back pain.

**CASE:** A 37 year old active man with no significant medical history and a 9-pack-year smoking history, presented with worsening lower back pain for 2 months. The pain started after lifting heavy weights at work and he was treated with pain medications as outpatient. He denied any red flag symptoms or hematuria. However, his physical exam was significant for a hard palpable abdominal mass and tenderness to the lumbar spine. Labs were significant for mild anemia, high blood sugar and elevated lipase. CT-scan and MRI of the chest, abdomen, and pelvis revealed a large right renal mass invading the IVC with retroperitoneal extension. There were metastases in the lungs, pancreas and epidural space with an L2 pathological fracture. Biopsy showed poorly differentiated Fuhrman nuclear grade 4 Renal Cell Carcinoma. Immunohistochemical evaluation revealed patchy nuclear staining for TFE-3 protein. He was referred to tertiary center for radiotherapy and chemotherapy, but did not tolerate chemotherapy and died 3 months after the initial diagnosis.

**DISCUSSION:** Renal cell carcinomas (RCC) associated with Xp11.2 translocations form a new and little known entity of the WHO classification. RCC associated with TFE3 gene fusions are relatively rare tumors that are diagnosed predominantly in children and extremely rare in adult. To our knowledge, only 34 adult cases with XP11.2 TRCC have been reported so far. Despite the innocuous complaints, TFE3-TRCC is more aggressive and often diagnosed in adults at an advanced stage. The most distinctive immunohistochemical marker of XP11.2 TRCC is a detectable nuclear staining for TFE3 protein, which has (97.5%) sensitivity and (99.6%) specificity. Because the rarity and aggressive clinical course of this tumor in adults, HE morphological examination alone is not reliable for differentiation between clear-cell-type and translocation-type neoplasm. Back pain in adult can be diagnostic challenge. Although they are quite rare, clinicians should always suspect additional sinister causes when patients complain of progressive back pain and have unusual physical findings.

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<th>24.</th>
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<th>Clinical Vignette</th>
<th>Chelu, Mihaela Mher Onanyan, MD, Scott Beede, MD, Daniel Goldsmith, MD</th>
<th>Capital Health Regional Medical Center (Saba Hasan)</th>
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<td>Polyarticular Septic Arthritis due to Staphylococcus Warneri and Endocarditis in a Patient with Rheumatoid Arthritis</td>
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**BACKGROUND:** Septic arthritis due to Staphylococcus Warneri are increasingly reported recently, and not only in patients with prosthetic devices. Also, an increase number of rheumatoid arthritis patients are diagnosed with septic arthritis. Chronic use of steroids and joint damage are the most important predisposing factors. Patients with polyarticular septic arthritis (PASA) superimposed on rheumatoid arthritis (RA) have a mortality rate of 56%, therefore, early recognition and therapy is essential; to improve survival and prevent sequelae.

**CASE REPORT:** A 64-year old female was admitted with sudden onset of nausea, vomiting and high grade fevers. Her medical history was significant for advanced RA on methotrexate, etanercept and steroids. On examination she was febrile, hypotensive, with right
hip pain on attempted active and passive motion. Laboratory tests revealed a white blood cell count (WBC) of 24.8 with a left shift. She was started on broad spectrum antibiotics and required vasopressors. CT scan revealed a large effusion within the right hip, and arthrocentesis yielded purulent material with a WBC of 107,460 and 91% neutrophils. On day 2, the patient underwent an arthrotomy with irrigation and debridement of her septic hip. A trans-esophageal echocardiogram (TEE) was negative for vegetation. On day 3, she developed severe pain in both shoulders. Synovial fluid analysis of the shoulders revealed septic arthritis, which required surgical intervention as well. Her blood and synovial fluid cultures were negative. Only the cultures of the synovial tissue obtained during the surgical debridement of the right hip revealed the causative agent as being S. Warneri. The patient was discharged on day 10 to complete a total of 4 weeks of vancomycin and ceftriaxone, but within 48 hours she was readmitted with fevers and chills but no signs of persistent septic arthritis. Blood and synovial fluid cultures were again negative. A follow up echocardiogram showed small vegetation on the mitral valve. Antibiotics were changed to daptomycin and aztreonam, and she completed a total of 6 weeks of antibiotics.

DISCUSSION: Septic arthritis of the hip and shoulder is less common in rheumatic patients and the diagnosis requires a high index of suspicion. This case also emphasizes the importance of tissue sampling in septic arthritis of unknown origin. S. Warneri is known to be of low virulence and grows slowly, and this might explain the delay in the diagnosis of endocarditis. Clinicians should be aware of S. warneri as a possible low virulence but invasive pathogen, especially in immuno-compromised patients.

Teaching point Patients with RA are susceptible to developing PASA which should prompt a vigilant search for endocarditis.

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<th>Clinical Vignette</th>
<th>Chelu, Mihaela</th>
<th>Andrei Yankovich, MD, Andrey Samal, MD, Manish Gugnani, MD, Daniel Goldsmith, MD</th>
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<td>Acute Massive Pulmonary Embolism Treated with Systemic Fibrinolysis: a remarkable case of survival</td>
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| BACKGROUND: Massive Pulmonary Embolism (PE) is a life-threatening emergency. The principal criteria for categorizing PE as massive are arterial hypotension and cardiogenic shock. Early mortality in patients with massive PE is at least 15%, and the degree of hemodynamic compromise is the most powerful predictor of in-hospital death. Systemic fibrinolysis reduces the risk of death or recurrent PE by 55%.

LEARNING OBJECTIVES: 1) Recognize cases of massive PE where systemic fibrinolytic therapy may be indicated. 2) Recognize the association between atypical antipsychotic therapy and increased risk of venous thromboembolic disease.

CASE: The patient was a 59 year-old gentleman who presented after a bicycle fall. He had a history of schizophrenia and ethanol abuse, and the only medication was quetiapine. The patient was treated for delirium tremens (DTs) in the Intensive Care Unit, and then transferred to the General Medical Floor. On day 7 of admission the patient suddenly developed syncope, hypotension with systolic blood pressure of 50 mmHg, tachycardia, and 60% oxygen saturation. He appeared cold and clammy, and his skin was cyanotic. His jugular venous pressure was 12 cm water and the cardiac exam revealed a prominent S2 heart sound. After emergent endotracheal intubation, the diagnosis of massive PE was suspected and heparin infusion was initiated along with crystalloids and vasopressors. The patient subsequently developed pulseless electrical activity and was successfully resuscitated. EKG revealed sinus tachycardia, with ST elevation in lead V1 and V2, with ST depression in lead V4, V5, V6, prominent P waves, incomplete right bundle-branch block. The bedside transthoracic echocardiogram revealed the right ventricle (RV) severely dilated, with RV hypokinesis and severely reduced systolic function, paradoxical septal systolic motion, moderate tricuspid regurgitation, and pulmonary hypertension. Tissue plasminogen activator (tPA) infusion was administered, and a CT pulmonary angiogram in 24 hours revealed
bilateral segmental and subsegmental pulmonary emboli. Venous Duplex revealed bilateral extensive deep venous thrombosis (DVT) of the lower extremities. The patient stabilized hemodynamically and was successfully extubated on day 5.

DISCUSSION: Although systemic fibrinolysis is not indicated in all patients with acute PE, it is recommended as standard, first-line treatment in hemodynamically compromised patients with massive PE. As soon as massive PE is suspected, high-dose unfractionated heparin should be administered. A large British study found that people prescribed antipsychotics have an increased risk of serious blood clots. The risk was greater for people on atypical drugs, and the highest risk was seen with quetiapine, which showed a nearly three times increase on an adjusted basis. This may help explain our patient’s massive PE and extensive clot burden despite DVT prophylaxis with Heparin 5000 units subcutaneously every 8 hours from day 1 of admission.

26. Clinical Vignette
Hassanein, Mohamed
Capital Health Regional Medical Center (Saba Hasan)

Left flank and chest pain in nephrotic syndrome: heralding thromboembolic complications

BACKGROUND: Venous thromboembolic events in nephrotic syndrome are well documented in the literature and they include deep venous thrombosis, pulmonary embolism and renal vein thrombosis.

CASE PRESENTATION: A 24 year old Hispanic male with no past medical or surgical history and no known drug allergies presented to the emergency room with 3 months of left sided flank and back pain. In addition, there was more acute, progressing left sided chest pain, aggravated with inspiration and associated with dry cough and shortness of breath. Vital signs were Temp: 98.4 ºF, HR: 78bpm, BP: 147/69 mmHg, RR: 16/min, and SO2: 98% on room air. Physical exam was unremarkable. Laboratory findings showed serum total protein 4.7g/dl, albumin 2.1g/dl, cholesterol 358 mg/dl. Urinalysis showed protein: 684mg/dl, moderate occult blood, hyaline casts 3-5 per HPF, granular cast 0-2 per HPF and 24 hour urine protein: 11037mg. Computed Tomography showed a suspected filling defect in a left lower lobe pulmonary artery suspicious for pulmonary embolism, associated with a moderate to large left pleural effusion and thrombus in the left renal vein and associated IVC. Additional work up showed: Antinuclear Ab: negative, CRP: 0.9mg/dl, C3: 167mg/dl, C4: 37mg/dl, anti-GBM Ab < 1.0, p-ANCA and c-ANCA negative, HB core Ab total: non-reactive, Hep B surface Ag: negative, HB core Ab (igM) total: non-reactive, hepatitis BE Ab: non-reactive, hepatitis BE Ag: non-reactive, HIV-1 RNA PCR: not detected, RPR screen: non-reactive. Factor V activity: 91%, protein C activity: >200%, protein S activity: 118%. Renal biopsy showed stage 2 membranous glomerulopathy and focal acute tubular injury and interstitial edema associated with renal vein thrombosis. Therapeutic anticoagulation was begun. Left thoracocentesis was performed for therapeutic and diagnostic purposes which yielded 800 ml of exudative fluid and the patient reported symptomatic relief. The patient was started on ACEIs and statins as well as prednisone 15mg orally once daily and cyclosporine 100mg orally twice daily. Teaching point Pleuritic chest pain or flank pain in a patient with findings of nephrotic range proteinuria may be an alarming symptom of underlying serious thromboembolic complications of nephrotic syndrome.

27. Clinical Vignette
Imran, Uzma
Beede MS, MD.
Capital Health Regional Medical Center (Saba Hasan)

Recurrent abdominal pain, a difficult diagnosis of Behcet’s disease

BACKGROUND: Behcet’s disease (BD) is a multisystem inflammatory vasculitis, characterized by recurrent oral and genital ulcerations, uveitis and frequently positive pathergy test. There can be a variety of visceral manifestations, including articular, gastrointestinal and nervous system involvement, though not frequently seen in the US population.
CASE: A 34 year old Hispanic woman with a history of thrombocytopenia, was hospitalized multiple times in the last 2 years due to recurrent abdominal pain, diagnosed as terminal ileitis and colitis. Further investigation revealed a history of recurrent oral ulcers with a recent labial ulcer which suggested a clinical diagnosis of Behcet’s disease, although she did not have uveitis symptoms and a pathergy test was negative. Colonoscopy revealed a single punched out ulcer of the descending colon, nonspecific acute inflammation with intact crypt architecture on histopathology, anti-Saccharomyces cerevisiae IgA antibodies was minimally elevated 26.1(N <20). Labial ulcer biopsy revealed chronic vulvitis with no dysplasia which supports our diagnosis. Interestingly she had chronic mild thrombocytopenia with platelets <130,000, which can be associated with Behcet’s. Colchicine and azathiothioprine were attempted but discontinued due to intolerance. Intestinal flares have been reduced and responded well to systemic steroids.

DISCUSSION: The prevalence of BD is higher in Middle and East Asia than in Western countries. Intestinal BD is a specific subtype of BD, characterized by intestinal ulcers and associated gastrointestinal symptoms. Similar to inflammatory bowel disease, intestinal BD exhibits a fluctuating disease course with repeated episodes of relapse and remission that necessitate adequate maintenance therapy after achievement of clinical remission. Medical treatment of intestinal BD is largely empirical since well-controlled studies have been difficult to perform due to the heterogeneity and rarity of the disease. 5-aminosalicylic acid, systemic corticosteroids, and immunosuppressants have been used anecdotally to treat intestinal BD. The clinical course of intestinal BD shows considerable variability, and the exact point at which more potent agents such as immunosuppressants should be used has not been elucidated. Teaching point Refractory colitis should raise the suspicion of Behcet’s disease if Crohn’s disease is ruled out.

28. 49 Clinical Vignette Majko, Arian Lindita Shehu, Majlinda Xhikola, Mohamed Hassanein, Emily Chen, Daniel Goldsmith Capital Health Regional Medical Center (Saba Hasan)

Hearing Loss As Initial Presentation Of Thrombotic Thrombocytopenic Purpura

INTRODUCTION: Thrombotic thrombocytopenic purpura (TTP) is a rare disorder of the blood-coagulation system, causing extensive microscopic clots to form in the small blood vessels throughout the body. Patients with TTP usually report an acute or subacute onset of symptoms related to neurologic dysfunction, anemia, or thrombocytopenia. Many cases present with a non-specific symptom complex, and require a high index of suspicion to recognize TTP as the underlying disorder.

CASE PRESENTATION: A 69-year-old female with a history of hypertension, diabetes, and hyperlipidemia was admitted in the hospital with change in mental status and seizure activity which was preceded by headache and bilateral hearing loss for 2 days prior to admission. Vital signs included blood pressure 153/68 mmHg; heart rate 106 bpm; respiration rate 25/min; temperature 99.3 F; oxygen saturation 96% on room air. Neurological examination was only significant for bilateral complete hearing impairment. Her speech was fluent and nondysarthric. Motor and sensation function were intact. Initial laboratory data showed blood sugar of 790, anion gap 21, creatinine 1.40, Hg 9.8 g/dL, WBC 8800/mm3, and platelet count of 16000/mm3. Extensive neurological imaging showed no evidence of acute abnormalities. The rest of laboratory data was significant for total bilirubin of 4.6, LDH 7928, reticulocyte count 6.8 %, which were suggestive of intravascular hemolysis. A peripheral blood smear revealed normocytic, hypochromic RBCs, significant schistocytes at 5-6 per high-power field. The presence of severe thrombocytopenia associated with a history of change in mental status, hearing loss, seizure, and schistocytes on peripheral blood smear strongly suggested a diagnosis of TTP. The patient was started on steroids and underwent emergent plasmapheresis, with subsequent dramatic improvement of hearing with complete recovery within 2 days. No further seizure episodes occurred. She underwent a total of 9 cycles of plasmapheresis during hospitalization. Complete remission was achieved on day 16, and she was discharged stable to home on day 22.
DISCUSSION: TTP is a hematological emergency and diagnostic challenge. The critical determinant of outcome is timely diagnosis and treatment. TTP has a varied presentation and a tendency to mimic several disorders. In its full-blown form, the disease consists of the pentad of microangiopathic hemolytic anemia, thrombocytopenic purpura, neurologic abnormalities, fever, and renal disease, however only 20-30 % of patients present with the classic pentad. Our patient presented with an atypical neurological manifestation of TTP which made the diagnosis more challenging. It is essential to be aware of the wide range of presentations and have a low threshold in considering such diagnosis. Due to the high mortality of untreated TTP, therapy should be initiated if the diagnosis of TTP is seriously considered.

29. 54 Clinical Vignette Samal, Andrey Peter C. Wenger Capital Health Regional Medical Center (Saba Hasan) The case of bilateral upper extremity edema Paget-Schroetter syndrome or the effort thrombosis is a rare condition that alludes to a thrombotic event in axillary or subclavian venous system associated with strenuous and repetitive movements in the upper extremities. 24-year-old female with chronic neck and upper thoracic back pain, no medications, except oral contraceptive pills (OCP), presented with right greater than left upper extremity edema that started two days after strenuous TRX upper extremity workout. Initial evaluation in the office revealed 4+ pitting edema from the upper arm to the wrist on the right and 1+ edema down to the elbow on the left arm. Normal pulses were palpable over brachial, radial and ulnar arteries bilaterally. No bruises or other skin abnormalities were found and there was no lymphadenopathy noted. A STAT MR angiography of the neck and the upper chest was performed the next day and showed focal narrowing of the right subclavian artery in the region where it was crossing the first rib. A suspected filling defect was described in the adjacent subclavian vein that could represented a venous clot. The findings were worrisome for thoracic outlet syndrome with Paget-Schroetter disease. The patient was immediately referred to a vascular surgeon, who after a physical examination performed a limited in-office vascular ultrasound which was normal. A formal bilateral upper extremity arterial and venous duplex scan was performed and provocative maneuvers confirmed the diagnosis of venous impedance and TOS. No clot or other abnormality was identified at the time of this study which was approximately 4 weeks after the initial presentation. Concurrently, the work-up for thrombophilia was performed and turned out to be negative. The patient underwent the first rib resection for decompression of her thoracic outlet on the right and after a course of traditional post-operative PT her symptoms resolved. She has resumed all her normal activities without issue. Anatomical abnormalities at the thoracic outlet and repetitive endothelial injury are well-recognized causative factors in Paget-Schroetter syndrome. In the presented case, the radiographic findings seemed to point to a right-sided compression. These studies were not able to explain her bilateral symptoms. Transient compression may have occurred due to the extensive upper extremity workout. OCP might have played a contributing role.

30. 139 Clinical Vignette Sekhar, Supriya Capital Health Regional Medical Center (Saba Hasan) LOOK WHAT I FOUND! Coexistent Ehrlichia chaffeensis infection with Klebsiella pneumonia UTI, bacteremia and liver abscess.

INTRODUCTION: Pyogenic liver abscesses have been increasingly prevalent in the tri-state New Jersey area, which is also an endemic area for tick borne infections. The classic symptoms of liver abscess are fever, chills, right upper quadrant pain and altered liver function tests. Ehrlichia Chaffeensis also presents with similar symptoms of fever, chills, rash and elevated liver function tests. However the presence of a rash is very atypical for gram negative bacteremia infections. The following case was an amalgamation of the above symptoms which prompted a broader horizon of thinking.
CASE: A 48 year old Hispanic male with no medical history presented with fever and chills for more than 2 weeks, dysuria and burning urination, redness of both palms which he said was resolving. He was seen a week prior in the ER with similar symptoms and was discharged on Ciprofloxacin. The patient was employed as a landscaper. On physical examination he had a fever of 101 F, and other vital signs were normal. There was redness of both palms and the rest of the examination was unremarkable. Labs showed a WBC 13.2, Platelets of 71, and urinalysis showed WBC > 100 hpf, positive nitrates, and few bacteria, AST 85, ALT 121 and Alkaline Phosphatase 517. At the prior ER visit, his LFTs were similarly abnormal. Blood and urine cultures showed K.Pneumoniae, however, the presence of a palmar rash, altered liver function tests, thrombocytopenia and the patient’s occupation as a landscaper drove a high index of suspicion for the presence of tick borne fevers and hence titres were drawn. The patient was empirically started on cefepime and doxycycline. A right upper quadrant ultrasound showed a complex cyst measuring 3.4x2.7x3.6 cm in the liver. For better definition, a CT showed an enlarging hypodense multi-septated cystic mass in the right hepatic lobe suggesting an abscess. Metronidazole was added to the regimen. The patient improved symptomatically after an ultrasound guided percutaneous drainage of the hepatic cyst and an in situ drain left in place. The abscess aspirate also revealed K. pneumoniae. In addition, Ehrlichia chaffeensis titres returned elevated at the same time. Patient followed up 2 weeks later and had completely recovered with all lab values at baseline.

CONCLUSION: This patient was demonstrated to have coexistent Ehrlichia Chaffeensis infection and Klebsiella UTI, bacteremia and liver abscess. Although the signs and symptoms of both infections overlap, certain features such as rash, prompted further investigation with appropriate management of the two processes. Clinicians must be prepared not to stop at the first plausible diagnosis (anchoring bias) and continue until all abnormalities are explained.

INTRODUCTION: Approximately 20 % of non resolving community acquired pneumonias are due to noninfectious causes. Steroids are the cornerstone of therapy for certain interstitial lung diseases, however is difficult to determine when they are the next step. We present a case of non resolving pneumonia with clinicoradiological features of acute interstitial lung disease.

CASE REPORT: A 46 year old man with past medical history of diabetes, hypertension and smoking presented to ER with a two week history of productive cough, malaise, fever and shortness of breath. Vital signs included temperature of 99 F, HR 102 beats per minute, RR 26/min, BP 134/73 mmHg and oxygen saturation 85 % on room air. Lung auscultation revealed bibasilar crackles. Laboratory tests showed no leukocytosis and PaO2 of 59.8 on 2 L NC. CT of the chest revealed bilateral infiltrates, more severe at the bases and periphery of the lungs. Levofloxacin was started to treat community acquired pneumonia. However, patient remained tachypneic with worsening oxygenation despite a NPPV trial and required intubation. Despite 10 days of broad spectrum antibiotics, patient remained intubated with worsening infiltrates. Multiple tests excluded infectious causes and malignancy. Antibiotics were stopped and pulse steroids were started on day 11. Patient was extubated on day 16 and was discharged stable to home on a tapering dose of prednisone. Given the excellent response to steroids without an alternative etiology identified, this case represents most likely an acute onset interstitial lung disease.
DISCUSSION: Due to increased awareness, there have been several reported cases of organizing pneumonia of the acute and fulminating variety. The outcome depends on early initiation of steroid therapy. It is imperative to have a high index of suspicion for this entity in patients presenting with acute respiratory failure. There are no current recommendations on when to start steroids in these cases with high clinicoradiological suspicion for acute interstitial disease. Some cases with a fatal outcome are either associated with delayed treatment or aggravated by lung biopsy. Because of the rarity of these cases, studies assessing optimal timing, dose or duration of steroid therapy are lacking, but increased recognition and reporting may improve our knowledge and approach to these challenging conditions.

32. 50 Clinical Vignette
Shehu, Lindita
Ronnie Mantilla, Arian Majko, Majlinda Xhikola, Emily Chen, Daniel Goldsmith
Capital Health Regional Medical Center (Saba Hasan)
Superior Vena Cava Syndrome From Small Cell Lung Cancer-Endovascular Stenting As Primary Treatment Modality

INTRODUCTION: Small cell lung cancer (SCLC) is the most aggressive form of lung cancer, characterized by a rapid doubling time and early metastases. Consequently, most patients (60-70 %) will have extensive disease at diagnosis. About 15,000 cases of superior vena cava (SVC) syndrome occur in the US yearly, with 25 % of the malignant etiologies from SCLC. Traditionally, SVC syndrome in SCLC has been managed with radiotherapy and chemotherapy. We present a case of SVC syndrome with endovascular stenting as a primary treatment modality followed by combined chemotherapy.

CASE REPORT: A 55 year old male with extensive tobacco history presented with one month of progressive proximal weakness, cough, hemoptysis, pleuritic chest pain, shortness of breath and one week of progressive face and neck swelling. Vital signs included blood pressure 131/83 mmHg; heart rate 108 bpm; respiration rate 25/min; temperature 97.9 F; oxygen saturation 97% on ambient air. Physical examination was significant for symmetrical face, neck and upper extremity swelling, jugular venous distention, chest wall tenderness, decreased breath sounds on the right and 1+ deep tendon reflexes. Laboratory data was unremarkable. Chest x-ray showed a large heterogenous mass in the right upper lobe extending to the right hilum and paratracheal space. CT chest showed a paratracheal, retrocaval mass measuring 7.6 x 7.2 cm in the right mediastinum and SVC obstruction. The patient underwent endovascular stenting of SVC and thrombolysis with rapid relief of symptoms. Tissue from transbronchial biopsy was inadequate and non-diagnostic. A thoracentesis drained 2 liters of turbid fluid, and pleural fluid cytology was consistent with SCLC. Cisplatin and etoposide chemotherapy was well tolerated and the patient was discharged on anticoagulation therapy with enoxaparin.

DISCUSSION: For SCLC patients, initial chemotherapy is the treatment of choice for symptomatic SVC syndrome. Most patients with SVC syndrome secondary to lung cancer can be treated with appropriately directed chemotherapy or radiotherapy. In these settings, the clinical response to chemotherapy alone is usually rapid. However, percutaneous stenting of the SVC is increasingly used as primary treatment and is effective in SVC syndrome of neoplastic origin. Because the decision for subsequent chemotherapy or radiation therapy is not prejudiced, stenting is a very effective initial step in the overall palliative treatment of these patients. Interventional endovascular techniques may offer a safe, rapid, and durable response, especially in patients without a previous diagnosis of cancer, and for whom the histological diagnosis takes more time than symptoms permit.

33. 86 Clinical Vignette
Shehu, Lindita
Arian Majko, Majlinda Xhikola, Mohammed Al-Jumayli, Mohamed Al-Jumayli
Capital Health Regional Medical Center
Hemolytic Uremic Syndrome Associated With Clopidogrel Use – A Case Report

INTRODUCTION: The antiplatelet drug clopidogrel has largely replaced ticlopidine, due to an association between ticlopidine and
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Center (Saba Hasan)  

thrombotic thrombocytopenic purpura–hemolytic uremic syndrome (TTP-HUS). Clopidogrel has been reported to be safe and effective in reducing vascular events. Nevertheless, there is growing evidence that Clopidogrel may also be associated with TTP-HUS.

CASE REPORT: A 43 year old male with history of chronic kidney disease, hypertension, and heart failure with preserved ejection fraction presented with progressively worsening abdominal pain, nausea, vomiting, dizziness, weakness, constipation and anuria for several days. The patient underwent cardiac catheterization one month before presentation and was taking Clopidogrel since then. Vital signs included blood pressure 240/148 mmHg; heart rate 92 bpm; respiration rate 20/min; temperature 98 F; oxygen saturation 100% on ambient air. Physical examination was significant for moderate discomfort due to abdominal pain, icteric sclera, right upper quadrant and epigastric tenderness with a normal neurologic exam. Initial laboratory data revealed creatinine 9.94 mg/dL, BUN 87 mg/dl, WBC 10,900/L, platelets 30,000/L, LDH 6375 U/L, total bilirubin 4.2 mg/dL, direct bilirubin 0.7 mg/dL, INR 1.0 and schistocytes on peripheral smear. The patient underwent emergent plasmapheresis given the high suspicion for thrombotic thrombocytopenic purpura (TTP). Clinical improvement was achieved within the first 24 hours. Platelet count steadily increased until normalized. ADAMTS13 activity sent on admission was normal; therefore the diagnosis of TTP was excluded and treatment for hemolytic uremic syndrome (HUS) was started with Eculizumab. Patient was discharged stable to home carrying a diagnosis of end stage kidney disease requiring dialysis. Clopidogrel was not reintroduced and on follow up patient was clinically stable with normal platelet count.

DISCUSSION: Clopidogrel has an improved safety profile compared with that of ticlopidine; however, recent reports have implicated clopidogrel as a possible causative agent of thrombotic thrombocytopenic purpura-hemolytic uremic syndrome (TTP-HUS), which is the most severe adverse reaction associated with ticlopidine. Phase III trials involving 20,000 patients treated with clopidogrel yielded no reports of TTP-HUS. However, with the increasing number of coronary stent procedures performed annually, the potential exists for recognition of clopidogrel-associated TTP-HUS. Several case reports have implicated it as a cause of TTP-HUS. Although there are only a few documented cases of clopidogrel-induced TTP-HUS, growing awareness of the possible problem may result in increased reporting. The index of suspicion for TTP-HUS should be high when we encounter patients taking clopidogrel who present with unexplained fever, renal failure, neurologic symptoms, bleeding, purpura or thrombocytopenia. Recognition of a drug-associated etiology in a patient with TTP-HUS is critical to avoid re-exposure and recurrent illness.

34. 48  

Clinical Vignette  

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Rhabdomyolysis-induced acute renal failure associated with Legionnaire’s disease.

LEARNING OBJECTIVE: Diagnose rhabdomyolysis induced acute renal failure in patients with underlying legionnaires’ disease. Legionella is an unusual cause of rhabdomyolysis induced acute renal failure. Renal biopsy should be performed earlier in the course of illness to diagnose histopathological pattern of the disease and early hemodialysis should be considered to prevent further decline in renal function.

CASE: A 37-year-old-male, with medical history of well controlled hypertension and asthma, presented with complaints of malaise, back pain, dark colored urine, cough, fever and chills for the past several days. On physical examination, the patient’s vitals were within normal range except temperature 101.2F. Chest radiograph showed consolidation with diffuse infiltrate of both lung fields. A diagnosis of community acquired pneumonia was made and patient was admitted and treated with Ceftriaxone and Azithromycin.
On second day of admission, the patient suddenly became hemodynamically unstable and developed cardio-pulmonary arrest. Patient was intubated and placed on hypothermia protocol. Laboratory data findings: PH 6.79, PCO2 115.2, HCO3^- 17.1, Sodium 132 mmol/L, Potassium 2.9 mmol/L, AST 145, ALT 1108, SaO2 66.3%, BUN 25 mg/dl, Creatine 2.88 mg/dl, CK 58000 U/L and positive urine antigen for Legionella. The patient required frequent hemodialysis for rhabdomyolysis induced acute renal failure. Patient recovered renal function after two weeks of hemodialysis. The follow-up clinical and laboratory findings were within reference range.

DISCUSSION: Legionnaire’s disease (LD) has been known to cause pneumonia with multiple organ failure. The classic presentations of Legionnaire’s disease include malaise, myalgia, anorexia, diarrhea, weakness, cough, confusion and headache. Distinctive features of Legionnaire’s disease with statistical significance compared to other type of community-acquired pneumonia include headache, diarrhea, arthralgia or myalgia, confusion, fever to 39°C, purulent sputum, hyponatremia, hepatic dysfunction, creatine phosphokinase elevation, hypophosphatemia, proteinuria and hematuria. The mechanism of rhabdomyolysis associated with Legionella is unknown, theories include direct invasion of Legionella into the muscle itself, or the release of its endotoxin into the circulation with subsequent muscle injury. In our case, Legionella was considered for the multi-organ failure as evidenced by laboratory findings. However, renal biopsy should be considered to diagnose typical pathological pattern of acute renal failure.

Clinical Vignette

**Sumanam, Phaniram, Sudeep Dhillon, MD, Tania Calzada, MD**

**Capital Health Regional Medical Center (Saba Hasan)**

Iodine Induced Acute Hyperthyroidism with Atrial Flutter in an Elderly Patient - Are we over doing the tests?

BACKGROUND: Iodine induced hyperthyroidism is a thyrotoxic condition caused by exposure to excessive iodine. This is a case of hyperthyroidism, due to the Jod-Basedow phenomenon following administration of oral and multiple IV iodinated contrast in a patient with history of multinodular goiter. Given our aging population with possible subclinical hyperthyroidism, multinodular goiter, and the rise in contrast administration for routine diagnostic studies, this case serves to raise awareness of the risks of repeated tests administered to our patients.

CASE PRESENTATION: This is an 89-year-old, Greek-speaking female, admitted from an acute rehabilitation center with palpitations, dyspnea on exertion, and found to be in rapid atrial fibrillation. The patient reported having palpitations, increasing fatigue, heat intolerance as well as some dyspnea with exertion. She denied chest pain, SOB, but complains of difficulty swallowing. She had been recently admitted to the hospital few weeks prior, for dysphagia and dry mouth, and was diagnosed with sialoadenitis, subclinical hyperthyroidism and multinodular goiter. On physical exam, the patient was resting comfortably. Vital Signs: Blood pressure 100/60 mmHg, heart rate 130 bpm and irregularly irregular, There was a 32 lb weight loss over one month. Thyromegaly and bilateral 1+ ankle edema was noted. EKG showed atrial flutter with 2:1 conduction, left ventricular hypertrophy, and ST depressions related to rapid rate. Atrial flutter was new when compared to previous EKG. During the prior admission, the patient received 3 IV contrast loads over a 2 week period for CT scans to evaluate her dysphagia and sialoadenitis. Thyroid ultrasound showed an enlarged, heterogeneous thyroid gland, a 5.0 cm nodule in left lobe and a 3.2 cm nodule in the right lobe. Patients TSH were always less than <0.015uIU/ml, and Free T4 1.74 before treatment that normalized to 1.29 after treatment. The patient was successfully treated initially with propanolol 60 mg TID, Methimazole 10 mg TID and later tapered to atenolol 50 mg daily and methimazole 10 mg once daily.
DISCUSSION: The biggest risk factors for manifesting overt hyperthyroidism after contrast administration are older age, Grave’s disease, living in an iodine depleted area, and multinodular goiter. Our patient presented with an episode of acute hyperthyroidism manifested as atrial fibrillation and rapid weight loss following three CT scans within a short period. Many patients with subclinical hyperthyroidism, advanced age, and undiagnosed multinodular goiters undergo radiological studies with contrast. This patient serves as a reminder to clinicians that our aging patient population is at risk for arrhythmias, increased morbidity, and mortality, from common diagnostic studies. Diagnostic imaging studies must be ordered wisely not only for cost/benefit considerations, but also for risk of adverse events associated with them.

36. 43 Clinical Vignette
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Acute Retroviral Infection

BACKGROUND: The human immunodeficiency virus (HIV) belongs to the family of human retroviruses (Retroviridae) and the subfamily of lentiviruses that causes the acquired immunodeficiency syndrome (AIDS). HIV/AIDS continues to have extraordinary public health impact in the USA. More than 1.1 million people in the United States are living with HIV infection, and almost 1 in 6 (15.8%) are unaware of their infection. HIV infection is mostly asymptomatic except in the acute infection when many experience a non-specific flu-like illness, and later in the disease when opportunistic infections and other sequelae of immunosuppression appear. Recognizing acute retroviral syndrome may provide many advantages for both patient and society.

CASE REPORT: A 20 year old African-American male with a history of asthma, UTI and pyelonephritis presented with complaints of back pain, fever, chills and sweats starting four days ago. He also complained of non-radiating constant lower abdominal pain accompanied by nausea and vomiting. There was no diarrhea, no cough, no sore throat, no dyspnea. The patient smoked half pack a day, and denied alcohol or illicit drug use. Vital signs were temperature 103 F, pulse 110 bpm, RR-18/min, BP-105/68 mmHg. Physical exam showed significant neck lymphadenopathy, mild tenderness to suprapubic area, and paravertebral tenderness from distal cervical spine to the sacrum. Laboratory evaluation showed creatinine 1.72 mg/dL, WBC 12.6, platelets 148. CT abdomen/pelvis showed prominent right lower quadrant lymph nodes. He was started on ceftriaxone and fluids for possible acute UTI. HIV ELISA was reported negative. Despite antibiotics, the patient continued complaining of dysuria, back pain, fever, and migraine-like headache with photophobia. MRI of the spine showed no evidence of abscess or osteomyelitis, but revealed markedly enlarged left retropharyngeal lymph node. Urine and blood cultures were negative, as were tests for Lyme disease, West Nile virus, CMV and parvovirus. HIV viral load came back at 2,958,120 copies. Antibiotics were discontinued and the patient was discharged to follow up for initiation of HIV treatment.

DISCUSSION: Acute HIV diagnosis can be easily missed as it presentation similar to any acute viral infections. HIV antibody screen often is negative so viral load PCR become crucial in making the correct diagnosis. Identifying HIV infection at the point of acute retroviral syndrome may offer several advantages including lengthening the patient’s time of immunocompetence, decreasing HIV transmission, and avoiding misdiagnosis and unnecessary procedures and treatment. Clinicians index of suspicion for acute retroviral syndrome must be high in order to send the correct laboratory evaluation early in the patient’s course.

Teaching points: 1) Symptomatic acute retroviral infection can mimic other acute viral processes. 2) HIV ELISA is not a definitive test and HIV viral load has to be ordered if infection is suspected.
Clinical Vignette

Abel, Nicole

Joshua Brody, DO and Satyajeet Roy, MD, FACP

Cooper University Hospital
(Brian Gable)

Anti-coagulation improved headache associated with levonorgestrel-releasing intrauterine device

Cerebral venous sinus thrombosis has an estimated annual incidence of 3-4 cases per 1 million people, with about 75% of cases occurring in women. Worsening and gradually progressive headache is the most common clinical presentation, however stroke-like symptoms including aphasia or hemiparesis, seizures, or signs of increased intracranial pressure may also be presenting symptoms. Cerebral venous thrombosis is diagnosed with abnormal signal on brain MRI or absence of flow on a MRV. Risk factors for developing cerebral venous thrombosis include prothrombotic conditions and hormonal contraceptives. A 34-year-old woman without preexisting medical problems presented with a weeklong severe right-sided headache associated with nausea and photophobia. She had a levonorgestrel-releasing intrauterine device insertion a month before her presentation. She had normal vitals and normal physical examination, including an unremarkable neurological examination. She was treated for migraine headache with sumatriptan and ibuprofen. Her symptoms improved minimally but she returned after two days with worsening headaches and nausea. A non-contrast magnetic resonance imaging (MRI) brain showed a filling defect and a magnetic resonance venogram (MRV) of head demonstrated occlusive thrombus in the sagittal sinus and bilateral transverse sinuses, confirming a diagnosis of cerebral venous thrombosis. She was treated with intravenous heparin and continued on warfarin. The levonorgestrel-releasing intrauterine device was removed. Her headaches improved in two weeks, and a repeat MRV of head during follow up five-months later showed significant improvement in the previously described thrombi. Raising awareness of cerebral venous thrombosis may improve the diagnosis of this condition. It should be considered in a young or middle-aged patient with recent or unusual headache, those who are pregnant, have existing thrombophilia, or those on hormonal contraceptive therapy. Anticoagulation is the mainstay treatment and more than 80% of patients treated for this condition have a good neurologic outcome.

Clinical Vignette

Agarwal, Abhishek

Satyajeet, Roy

Cooper University Hospital
(Brian Gable)

Nitrofurantoin induced ANCA associated renal and skin vasculitis

INTRODUCTION: Patients presenting with fever, rash, and renal involvement offer a variety of etiologies ranging from drugs to systemic illness.

CASE: A 67-year-old Caucasian female with hypertension and hyperlipidemia on hydrochlorothiazide and pravastatin presented with non-itchy leg rashes and fever for 1 day. Three-days ago, she received nitrofurantoin for urinary tract infection. She denied insect bite, sinus pain, epistaxis, cough, hemoptysis, wheezing, or pain anywhere. She had a temperature of 101 degree F, diffuse palpable purpura on legs. The rest of her physical examination was normal. Her comprehensive metabolic panel and complete blood count were only abnormal for elevated creatinine 1.13 mg/dl, BUN 32 mg/dl, and eosinophils 0.56 K/ul. Urinalysis showed red and white blood cell casts. Chest X-ray and renal ultrasonography were normal. A diagnosis of vasculitis and microscopic polyangiitis was suspected. Her peripheral-antineutrophilic-cytoplasmic-antibody with antymyeloperoxidase specificity [P-ANCA (MPO)] was highly positive and cytoplasmic-ANCA with proteinase-3 (PR3) specificity was negative. Her ANA (anti-nuclear antibodies) was negative and her complement levels were normal. She was offered to undergo a renal biopsy which she declined. She was diagnosed as ANCA-associated drug-induced vasculitis. Eosinophilic granulomatosis with polyangiitis was considered less likely due to lack of asthma and atopy. Acute interstitial nephritis was less likely due to absence of marked eosinophilia and eosinophiluria. A final diagnosis of vasculitis and microscopic polyangiitis secondary to nitrofurantoin was established based on the temporal relationship between initiation of nitrofurantoin and the onset of symptoms. Nitrofurantoin was discontinued and patient was treated with prednisone for
10 days. Her purpura and fever resolved in 2 days, and BUN, creatinine, eosinophils and urinalysis became normal at 1 week.

**DISCUSSION:** ANCA-associated vasculitis have 3 major presentations: Granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA). GPA is characterized by granulomatous involvement of the upper respiratory tract (sinusitis, pulmonary hemorrhage) and more than 90% ANCA positivity, predominantly PR3 (example: Wegener’s granulomatosis). MPA shows more than 90% ANCA positivity, predominantly myeloperoxidase (MPO). EGPA is characterized by marked eosinophilia, asthma, atopy and only 50% ANCA positivity, mostly MPO (example: Churg–Strauss syndrome). Drug-induced ANCA associated vasculitis present with fever, arthralgia, purpura, renal or pulmonary manifestations. High titers of ANCA antibodies with MPO specificity helped establish a diagnosis of microscopic polyangiitis. According to the Naranjo Adverse Reactions Probability Scale (NADRPS) nitrofurantoin was the most probable cause of drug-induced renal and skin vasculitis, which has not been reported. In mild cases, discontinuation of the drug can be sufficient but in severe cases involving the kidneys or lungs, treatment with corticosteroids or cyclophosphamide may be necessary.

**CONCLUSION:** In the outpatient clinics, a high degree of suspicion is necessary to recognize ANCA associated skin and renal vasculitis in order to provide optimal medical management.

39. 154 Clinical Vignette

**Agarwal, Abhishek**

Samantha Lee, Karim Nathan, Samson Zarbiv, Anjali Desai

Cooper University Hospital (Brian Gable)

Liddle’s syndrome: Rare cause of hypertension and hypokalemia in young Hispanic adult

**INTRODUCTION:** In young adults presenting with hypertension, secondary causes of hypertension should be sought. Liddle’s syndrome is a rare cause of secondary hypertension and its diagnosis is important as the treatment is different from other causes of hypertension.

**CASE:** A 38 year old man with h/o hypertension diagnosed 10 years ago presented with complaints of acute onset focal bilateral lower and R upper extremity weakness. He denied any other symptoms or past medical problems. He denied any diuretic use or licorice ingestion. On examination, his BP was 184/100 with palpable pulses b/l, no asymmetry. Apart from hypotonic, hyporeflexic weakness in his affected extremities, his physical exam including cardiovascular exam was normal. The lab investigations revealed Hypokalemia (K- 1.7 mmol/l) with normal serum Magnesium and elevated bicarbonate (35mmol/l). His Transtubular potassium gradient was 7.4. His low dose dexamethasone suppression test was positive. His renin (0.08 ng/mL/h) and aldosterone (<1 ng/dL) levels were low. His 17OHprogesterone levels were normal. His 24 hour Urine cortisol was normal and 24 hour 17OH corticosteroids were low. His CT Abdomen did not reveal any adrenal enlargement or masses. The patient thus had hypertension and hypokalemia with increased renal losses and low renin and aldosterone. His weakness resolved with potassium repletion. His hypertension did not respond to spironolactone initially and with presumptive diagnosis of Liddle’s syndrome he was started on amiloride which controlled his BP and potassium. On further investigation, patient was unaware of any family history of hypertension. He was asked to undergo genetic testing but unfortunately patient never followed up.

**DISCUSSION:** About 1 in 3 people in USA suffer from hypertension. Primary hypertension is the most common form, without an underlying cause, yielding 95% of causes. But secondary hypertension should be suspected in young patients presenting with hypertension. Liddle’s syndrome is a rare hereditary cause of secondary hypertension where patients typically present with episodic uncontrolled hypertension accompanied by hypokalemic metabolic alkalosis. Clinically, Liddle’s syndrome can mimic primary
Hyperaldosteronism as patients also present with hypertension and symptoms of hypokalemia including weakness, myalgia, muscle cramps and constipation. Unlike primary hyperaldosteronism, Liddle’s syndrome is caused by an autosomal dominant genetic mutation of renal epithelial sodium channel cells which leads to an increase in sodium reabsorption and potassium secretion from the collecting tubule. Spironolactone is ineffective in this disorder as the increase in sodium channel activity is not aldosterone dependent. Potassium-sparing diuretics such as amiloride and triamterene, which directly close the sodium channels, are effective in Liddle’s syndrome.

**CONCLUSION:** It is important to consider Liddle’s syndrome in a patient with hypertension, hypokalemia and metabolic alkalosis, as these patients won’t respond to conventional antihypertensives. Identifying Liddle’s syndrome early and treating appropriately with triamterene or amiloride can prevent complications of uncontrolled hypertension.

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**A Rare Case of Rash and Fever in an Adult**

**INTRODUCTION:** Many infectious diseases may present with fever and rash. In cases of rare diseases, a detailed history and clinical examination are particularly necessary to correctly identify entities that may not typically be seen in adults.

**CASE:** A 40-year-old woman presented with a rapidly progressive, non-pruritic rash on her face, hands and feet for 3 days. The rash was preceded by 3 days of fever (101-103 degrees F), sore throat, and headaches. One week preceding the onset of her symptoms, her 8-year-old daughter suffered from fever, diarrhea and a rash on her feet. She denied coming in contact with any other persons, recent travel, or exposure to new food, soaps, or clothing. Symptoms failed to improve with diphenhydramine. On examination, she had a fever of 101 degrees F. There was an erythematous, papulo-vesicular rash on her distal upper extremities including her palms, distal lower extremities including the soles of her feet and in a perioral distribution. Oral lesions were also noted on her soft palate. The rest of her physical examination was unremarkable. Diagnostic tests showed a normal complete blood count, comprehensive metabolic panel, urinalysis, and a negative rapid plasma reagin (RPR). Based on symptomatology and clinical exam findings, she was diagnosed with Hand, Foot, and Mouth Disease (HFMD). Initially, she was treated conservatively with rest, oral hydration, ibuprofen, and contact precautions. The rash continued to progress and alternatively a five day course of methylprednisolone was started. She had a dramatic improvement within 24 hours with complete resolution of her symptoms in 3 days.

**DISCUSSION:** HFMD is an acute viral infection caused by enteroviruses, most often Cocksackie A16 and enterovirus 71, and classically presents with fever, painful oral lesions and a rash on the hands and feet. It is a disease predominantly seen in children under the age of 5, but can rarely present in adults. Clinical manifestations are only seen in less than 1% of infected adults. Diagnosis is typically based on clinical presentation, but in more severe cases, oral, skin, fecal or serum samples may be obtained for PCR analysis. The virus may still be detected in the stool for up to 4–8 weeks after the acute phase. Prognosis is typically favorable and resolution of oral and skin lesions occurs in 5–10 days. However, serious complications can occur including myocarditis, aseptic meningitis, and flaccid paralysis, nephrotic syndrome and pulmonary edema.

**CONCLUSION:** HFMD is a disease typically manifested in children but can rarely occur in adults. Diagnosis based on history and physical examination is essential to appropriately manage, treat and observe for complications that may arise.
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<th>Eno, Eben</th>
<th>Satyajeet Roy, MD, FACP</th>
<th>Cooper University Hospital (Brian Gable)</th>
<th>Episodic forgetfulness and rash – Prompt diagnosis and management can be life saving</th>
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<td><strong>INTRODUCTION:</strong></td>
<td>Presentation of transient neurologic abnormalities, icterus and purpura in an outpatient setting can represent a myriad of medical conditions. Early diagnosis can help in planning for a targeted therapy, and a successful outcome.</td>
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<td><strong>CASE:</strong></td>
<td>A 61-year-old Caucasian man with peripheral arterial disease presented with brief episodes of forgetfulness and dizziness associated with tiredness for 3 days. He had recovered from a flu-like illness a week prior to the presentation. He denied weakness or numbness, head trauma, headache, vomiting, abdominal pain, diarrhea, or blurred vision. He was taking cilostazol and aspirin. His vital signs were in the normal range. His physical exam was remarkable only for scleral icterus, skin pallor, and presence of petechiae on arms and legs. Thrombotic thrombocytopenic purpura was suspected and he was admitted to the intensive-care-unit. Diagnostic studies revealed a normal CAT-scan of head, anemia (hemoglobin 11.1 gm/dL), marked thrombocytopenia (platelets 7 k/uL), elevated BUN (35 mg/dL), elevated creatinine (1.35 mg/dL), elevated total bilirubin (6.7 mg/dL), elevated lactate dehydrogenase (2.5 mmol/L), elevated aspartate aminotransferase (77 U/L), and low haptoglobin (less than 10 mg/dL). Peripheral blood smear showed schistocytes. A diagnosis of thrombotic thrombocytopenic purpura (TTP) was made. The patient improved after 27 rounds of plasmapheresis. His HIV test was negative and he had a low ADAMTS13 activity (less than 10%). He remained symptom free at 1-year follow up.</td>
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<td><strong>DISCUSSION:</strong></td>
<td>In 1924 Dr. Moschowitz described TTP as a pentad; microangiopathic hemolytic anemia, renal failure, thrombocytopenia, fever and neurologic findings. TTP is caused by deficiency of, or antibodies to a metalloproteinase called ADAMTS13, that cleaves von Willebrand factor (VWF). Failure of cleavage results into aggregation of unusually large VWF multimers which promote platelet aggregation and thrombosis in small vessels. It could be acquired or hereditary. Acquired TTP has an autoimmune etiology with autoantibody inhibition to ADAMTS13, with incidence of 2.9 adults per 1 million per year and it differs from TTP-HUS which presents with hemorrhagic diarrhea preceding renal failure in children. The hereditary form is caused by homozygous and compound heterozygous ADAMTS13 mutations, in which patients are usually clinically asymptomatic. An ADAMTS13 of less than 10% represents poor prognosis and high recurrence rate. The treatment of choice is plasma exchange, in which cycles are continued till the platelet count normalizes and signs of hemolysis are resolved. Complicated cases are treated with glucocorticoids and relapses may warrant trial of rituximab, other immunomodulators, and splenectomy. An early diagnosis and plasma exchange can reduce mortality from 85-100% to 10%.</td>
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<td><strong>CONCLUSION:</strong></td>
<td>This case highlights the importance of detailed physical examination and a high index of suspicion for TTP in patients who present in the outpatient clinic with purpuric rash and transient neurological disturbances.</td>
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<th>Clinical Vignette</th>
<th>Green, Camille</th>
<th>Satyajeet Roy, MD FACP</th>
<th>Cooper University Hospital (Brian Gable)</th>
<th>One in A Million: A Curious Case of Ascites in a Young Male</th>
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<td><strong>INTRODUCTION:</strong></td>
<td>Common etiologies of ascites include cirrhosis, decompensated heart failure and nephrotic syndrome. In refractory or recurrent ascites, an ascitic fluid analysis and abdominal imaging studies provide vital diagnostic information.</td>
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CASE: A 30-year-old male presented with abdominal pain, distention and diarrhea for 3 weeks to the ED. He had a 20-lb weight loss over 3 months. His symptoms were associated with intermittent low grade fevers and night sweats. He worked as a commercial fencer. Twenty-one years ago, he was treated with chemotherapy, craniospinal radiation and ventriculo-peritoneal (VP) shunt for primary CNS germinoma. He denied alcohol intake or drug use. His maternal aunts had breast cancer and ovarian cancer. His vitals were normal. His physical exam was only significant for mildly tender abdominal distention and fluid thrill. He had a normal comprehensive metabolic panel and complete blood count. Hepatitis virus panel and HIV test were negative. Abdominal ultrasonography confirmed a large volume ascites. He received broad-spectrum antibiotics for suspected peritonitis and he underwent large volume paracentesis. Ascitic fluid analysis revealed an exudative fluid with white blood cell count of 5,113 (78% neutrophils), total protein 4.3, SAAG 0.2, negative cytology and negative culture results. There was no VP-shunt malfunction. Abdominal CAT-scan revealed massive ascites and enhancement of the peritoneal reflections with thickening and nodularity. Considering re-accumulation of ascites, weight loss, low grade fevers and history of exposure to craniospinal radiation, an underlying peritoneal malignancy was suspected. Peritoneal biopsy confirmed a diagnosis of malignant peritoneal mesothelioma. He underwent surgical debulking and hyperthermic intraperitoneal chemotherapy (HIPEC). Currently, he is doing well and received 6 more cycles of chemotherapy.

DISCUSSION: Ascites in the United States is most likely a result of portal hypertension from cirrhosis in 81% of cases. However, in 10% of cases, malignancy is the cause. In this case, culture negative neutrocytic ascites was an indicator of malignancy, and pointed to the ultimate diagnosis of primary peritoneal mesothelioma (PPM). PPM is a rare entity with an incidence of 1 in 1,000,000. Risk factors for this entity includes asbestos exposure, chronic peritonitis, talc exposure, diffuse lymphocytic lymphoma, familial Mediterranean fever and/or abdominal radiation. This rare entity is usually found in women. It carries a poor prognosis with 100% mortality. Median survival has been reported as 12-56 months, even with extensive surgery and chemotherapy.

CONCLUSION: The cause of PPM in our case could have been genetic, asbestos exposure while working as a commercial fencer, or radiation exposure. This case report highlights the necessity of keeping a broad differential when approaching the diagnosis of ascites and that when fluid analysis reveals culture negative ascites with high cell count, SAAG <1.1, total protein >2.5 and negative cytology, a malignancy should still be considered even in young patients.

43.  37  Clinical Vignette  Grewal, Navjot Ahmed Sesay, Dana, Byrne Cooper University Hospital (Brian Gable)  

ACUTE FEBRILE NEUTROPHILIC DERMATOSIS, TRIMETHROPRIM-SULFAMETHOXAZOLE (BACTRIM) AND PERICARDITIS

INTRODUCTION: Acute febrile neutrophilic dermatosis is a hypersensitive reaction characterized by nonspecific clinical and pathologic findings. Lesions are usually painful, red/purple papules, nodules or plaques. They are asymmetrically distributed in the upper extremity, face, neck and are usually preceded by several days of fever and leukocytosis. Histological findings are diffuse infiltration of mature neutrophils in the papillary, upper reticular dermis, epidermis and adipose tissue. Drug induced sweets syndrome represents approximately less than 5% of all cases. This case report shows a temporal association with Trimethoprim-Sulfamethoxazole and the onset of sweet syndrome in a patient with pericarditis.

CASE REPORT: Patient presented with acute onset of dyspnea, pleuritic chest pain, rash and subjective fevers. Few weeks prior to presentation, patient experienced sore throat with yellow exudates, malaise and diffuse myalgia. Azithromycin was taken for 5 days.
without resolution of symptoms. Rapid Strep test was negative for streptococcus pharyngitis. However, she received Trimethoprim-
Sulfamethoxazole for 5 days. Shortly after, patient complained of worsening myalgia, arthralgia and fevers of 103°F. She
subsequently developed vesicular, erythematous rash localized at the left forehead. The rash spread over the entire forehead, scalp,
chest and later involving the upper and lower extremities. One day prior to admission, the patient complained of severe pleuritic
chest pain and dyspnea. Hospital course notable for negative blood, urine cultures, CSF for Cryptococcus, Herpes 1 & 2, Coxsackie,
Measles, RPR, Varicella-zoster, HIV 1 & 2. Physical exam revealed diffuse skin findings of painful, purple papules, friction rub and EKG
showing PR segment depression and diffuse ST elevation in multiple leads.

**DISCUSSION:** The existing consensus is that Sweet Syndrome is a hypersensitivity reaction to medication, bacterial, viral, tumor
antigen and several other factors. The current postulations implicate leukotactic mechanism, dermal dentrocytes, autoantibodies,
immune complexes, human leukocyte antigen and cytokines. On presentation, patient satisfied all five criteria for drug induced
Sweets syndrome as per Warren and Cohen. She was febrile with t-max at 103 degrees along with erythematous vesicular lesions
and nodules that were asymmetrically distributed to the face, neck and upper extremity. A temporal relationship existed with the
use of Trimethoprim-sulfamethoxazole and onset of syndrome. Biopsy during hospitalization established a histological evidence of
neutrophilic infiltration without evidence of leukocytoclastic vasculitis and finally, there was temporal related resolution of lesions
with steroid use. However, on presentation, our differentials also included disseminated Herpes zoster virus, Coxsackie virus and
Vasculitis.

**A young pregnant woman with acute chest pain - Must think about the life threatening possibility**

**INTRODUCTION:** Acute ST segment elevation myocardial infarction is very uncommon under age 40 (about 5.6 %), especially in
women (about 0.7%). Young woman with acute chest pain can present with serious coronary artery disease especially during
pregnancy.

**CASE:** This is a 35 year-old female (gravida 3 para 2) who was pregnant at 33 weeks and 5 days, presented with sharp substernal
chest pain radiating to arms bilaterally, associated with dyspnea and nausea. Her chest pain lasted for about an hour, and was
relieved with aspirin and nitroglycerin. Her past medical history was significant for hemolysis -elevated liver enzymes-low platelets
(HELLP) syndrome during prior pregnancy, hypertension, Grave’s disease, and ascending thoracic aortic aneurysm.

Electrocardiogram showed normal sinus rhythm, normal axis, and significant ST segment elevation in anterior leads. She had
elevated troponin of 6.92 ng/mL and creatinine kinase of 1952 U/L. Echocardiogram showed hypokinesis of distal septum and distal
inferior-apical segment of the left ventricle. A cardiac catheterization showed diffuse narrowing of distal segment of LAD, with most
severe stenosis at the apical portion, representing adjacent spontaneous coronary artery dissection (SCAD). No percutaneous
coronary intervention was done at that time. She was also found to be hypertensive on admission. Her blood pressure was
controlled with labetalol. Patient was managed conservatively. After 6 days a repeat cardiac catheterization showed healing LAD
dissection. Patient remained in a stable condition.

**DISCUSSION:** Spontaneous coronary artery dissection while rare it is often a lethal condition. I has a prevalence of 1 in 20,000 to 30,
000 deliveries and has a high mortality rate for both mother and child. Some of the predisposing conditions to spontaneous coronary
artery dissection include pregnancy, fibromuscular dysplasia, trauma, drug abuse, and connective tissue disorder. Most cases are
usually seen in young, otherwise healthy females during pregnancy, with hormonal changes, hemodynamic stress, and modification in immune system as contributing factors. Clinical presentation ranges from asymptomatic to acute coronary syndrome. It is seen in the third trimester of pregnancy and in the early postpartum period. The dissection most commonly involves the LAD (80%). The overall mortality is more than 50% at presentation. Prompt recognition of this entity is imperative for appropriate management. Management of these patients can vary depending on severity, underlying condition, and confounding comorbidities. Treatment can include conservative management with monitoring, percutaneous coronary intervention, or coronary artery bypass graft.

**CONCLUSION:** A low threshold to investigate pregnant women with chest pain is vital, and should be investigated promptly to rule out life threatening diseases such as spontaneous coronary artery dissection. Our patient was managed conservatively with monitoring of fetus until delivery. She had normal spontaneous vaginal delivery at 37 weeks without any complications.

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### A rare trio: Lung carcinoid, Adrenocorticotropic hormone and Cushing's syndrome

**INTRODUCTION:** Carcinoid tumors mostly originate in the small intestine. Carcinoid tumors of lungs represent about 10% of all carcinoid tumors, and comprise about 1%-6% of all lung tumors. A very small fraction of carcinoid tumors of lungs are associated with Cushing syndrome due to ectopic Adrenocorticotropic hormone (ACTH) secretion. We present a rare case of dexamethasone suppressible Cushing’s syndrome from bronchial carcinoid.

**CASE PRESENTATION:** A 37 year-old woman who initially presented with gradual onset generalized weakness, palpitations, fatigue, and menstrual cycle irregularities. Upon further questioning, patient reported weight gain and skin hyperpigmentation for past few years. She denied headaches or visual changes. Her past medical history was significant for new onset diabetes mellitus, hypertension, and central hypothyroidism. On clinical examination she had cushingoid features. She underwent biochemical testing, and was found to have elevated 24 hour urine free cortisol, elevated morning ACTH and free cortisol, and failure to suppress with 1 mg dexamethasone suppression test. However, her plasma cortisol suppressed after overnight 8 mg dexamethasone to 5.6ug/dL. A magnetic resonance imaging (MRI) study of the brain revealed findings consistent with a pituitary microadenoma. Incidentally, she was also found to have a 1 cm peripheral right pulmonary nodule on computerized axial tomography (CAT) scan of chest. A positron emission tomography (PET) scan revealed high activity in the pulmonary nodule. She underwent a pulmonary wedge resection. The tumor cells were positive for chromogranin, synaptophysin, and CD-56, consistent with a diagnosis of carcinoid tumor. Upon further staining, carcinoid tumor cells were also positive for ACTH.

**DISCUSSION:** Ectopic ACTH production accounts for 10–20% of all cases of Cushing Syndrome and is most commonly associated with small cell carcinoma of the lung and intra-thoracic carcinoid tumors. Furthermore, the incidence of Cushing Syndrome in subjects with bronchial carcinoid is around 1%. Bronchial carcinoid tumors can be extremely difficult to localize. Since bronchial carcinoid tumors are usually 1 cm or less, a high resolution CAT scan is required to visualize these tumors. Simultaneous presentation of pituitary microadenoma usually presents a diagnostic challenge. Inferior petrosal sinus sampling, selective pulmonary arterial sampling, and Somatostatin Receptor Scintigraphy, are some of the modalities to localize the tumor. Very few case of carcinoid tumors of lungs presenting as Cushing’s syndrome due to ectopic ACTH secretion have been reported in the literature.

**CONCLUSION:** Ectopic ACTH secreting tumors present most challenging differential diagnosis and require careful clinical, biochemical, radiological and pathological investigation. Our patient was successfully treated with pulmonary wedge resection. After
Clinical Vignette

Kamath, Priya

Dr. Satyajeet Roy

Cooper University Hospital
(Brian Gable)

My heart keeps me at the edge of my seat—it races too much when I stand up!

INTRODUCTION: Postural Orthostatic Tachycardia Syndrome (POTS) describes symptoms of orthostatic intolerance characterized by an increase in heart rate of 30 bpm (or rate that exceeds 120 bpm) within the first 10 minutes of standing, not associated with any chronic conditions that can interfere with autonomic or vascular tone.

CASE PRESENTATION: A 34 year-old Caucasian woman with history of migraines, hypothyroidism, and gastroparesis developed episodic lightheadedness on standing 3-4 times for 2 months. Her symptoms corresponded with a sudden increase in her pulse rate from her baseline of 55-60 beats/minute to 96-128 beats/minute as measured by the patient’s spouse. Episodes were associated with fatigue, headaches, palpitations, nausea, diminished concentration, and tremulousness. Her medications included rizatriptan, pantoprazole, levothyroxine, and metoclopramide. Physical examination revealed blood pressure of 96/68 mmHg without orthostatic changes, supine pulse 62 beats/minute, and BMI 28 kg/m2. The rest of her systemic examination was normal. She had a normal complete metabolic panel, troponins, complete blood count, thyroid profile, catecholamines, metanephrine, electrocardiogram, echocardiogram, and MRI brain. She was admitted for telemetry monitoring. It showed 6 short periods of sinus tachycardia when the patient was upright. During such episodes the patient complained of lightheadedness and palpitations, with an average increase in the pulse rates of 38 beats/minute with an 11-point average decline in the systolic blood pressures. A diagnosis of POTS was established. She was treated with aggressive hydration, electrolyte replacement, compression stockings, and daily exercises which provided mild symptomatic relief. Subsequently she was treated with acebutolol and midodrine which provided significant symptom relief.

DISCUSSION: The most common form of POTS is “partial dysautonomic” which often present acutely after a febrile illness in women and men (5:1). It is thought to be caused by the inability of the peripheral vasculature to maintain vascular resistance under gravitational stress which can cause a compensatory increase in heart rate and ionotropy. The second and most rare form of POTS is “hyperadrenergic” characterized by migraine headaches, tremulousness, cold sweaty extremities, orthostatic hypertension, along with postural tachycardia much like our patient above. Initial therapy for both partial dysautonomic and hyperadrenergic POTS is aimed toward lifestyle modifications (exercise, 3 gram salt diet, and compression stockings). While partial dysautonomic POTS therapy is directed more towards fluid resuscitation and vasoconstrictive agents, therapy for hyperadrenergic POTS is achieved with a combination of alpha and beta blocking pharmacotherapy (pure beta blockade may exacerbate symptoms) which ultimately resolved symptoms for our patient.

CONCLUSION: POTS can be a very debilitating condition for patients. It is important to identify POTS and initiate therapy early because close to 90% of patients respond to some combination of physical therapy and pharmacotherapy.

Clinical Vignette

Khan, Maliha

Jenny Melli, MD

Cooper University

Asymptomatic Hematuria: A Rare Case of Urinary Shistosomiasis in Camden, New Jersey
INTRODUCTION: Urinary schistosomiasis (bilharziasis) is a parasitic infection caused by worms that are prevalent in Africa, the Middle East, and certain areas of the Asia, but rarely seen in the United States. There are 200 million people affected by this parasite worldwide. [i] Schistosoma haematobium specifically deposits eggs into the urinary tract and can predispose humans to hematuria, renal failure, and squamous cell cancer of the bladder [ii].

CASE DESCRIPTION: This is a case of a 32 year old Egyptian male with no past medical history who presented to the primary care clinic for more than 1 year history of asymptomatic gross hematuria. He denied any symptoms of dysuria, hesitancy, urgency, frequency, incontinence or weight loss. He denied any recent trauma. Patient moved to the United States 4 years ago. Prior to moving he had fresh water exposure in Egypt. He takes no medications. He has no past surgical history. No allergies. Social history was significant for 2.5 pack year smoking history. Family history is non-contributory. Physical examination revealed a healthy male.

Lab Results: CBC was normal with no elevation in eosinophils. Urinalysis was consistent with red blood cells. Cystoscopy was performed. A cytology report revealed numerous cohesive urothelial cells. Biopsy of the bladder revealed rare calcified eggs with a terminal spine consistent with Schistosoma, in particular Schistosoma haematobium.

DISCUSSION: Asymptomatic hematuria in a young male patient is an uncommon presentation concerning for a parasitic etiology. However, residential and travel history are important components in making the diagnosis of an uncommon infection that is rather common elsewhere in the world. This is a case of a young patient who immigrated from Egypt with chronic hematuria and had a cystoscopy and bladder biopsy revealing schistosomiasis. Patient was treated with high dose praziquantil, 30mg/kg x 2 doses.


INTRODUCTION: Abdominal discomfort is a common symptom, with a wide differential diagnosis including both life threatening and benign etiologies.

CASE: A 36-year-old Caucasian female presented with a 1 week history of abdominal discomfort and nausea. She had no fever, loss of appetite, hematemesis, diarrhea, melena or dysuria. Her last menstrual period was two weeks prior to presentation. She denied significant past medical history, use of medications, alcohol, tobacco, or illicit drugs. On physical examination, vital signs were within normal limits. There was a firm, immobile, non-tender 9 cm infra-umbilical mass on palpation, without clinical evidence of lymphadenopathy. The remainder of examination was unremarkable. Diagnostic studies including comprehensive metabolic panel, amylase, lipase, complete blood count, pregnancy test and urinalysis were also unremarkable. Abdominal and pelvic CAT-scan revealed a well circumscribed 10×9×5.5 cm mass attached to the rectus abdominis muscle. No lymphadenopathy or other abnormalities were noted. The patient underwent wide local excision of the mass followed by local reconstruction of the defect. Histopathology of the mass revealed spindle-cells with partially intact myofibers, pale eosinophilic cytoplasm embedded in collagen
network. Immunohistochemistry revealed smooth muscle cell actin. A diagnosis of desmoid tumor was established. Patient underwent a colonoscopy which showed no abnormality.

**DISCUSSION:** Aggressive fibromatosis or desmoid tumors are rare benign tumors arising from fibroblasts, commonly presenting in women of child bearing age with a peak incidence between 25-35 years of age. They account for 0.03 percent of all neoplasms making them among the rarest of tumors. Although histologically benign, with no ability to metastasize, desmoid tumors can aggressively invade local structures and be life threatening if compresses vital organs. Most occur sporadically but a minority of tumors arise from a mutation of ACP gene, and can one of the first manifestation of familiar adenomatous polyposis. Patients typically present with a painless or mildly painful slow growing mass commonly located on the extremities or torso. Diagnostic tools include ultrasound, CT or MRI however the diagnosis is made with biopsy. A wide local excision is the treatment of choice for most tumors in those deemed surgical candidates. Average rate of recurrence is 20-30% which depends on a number of factors including size, location, and degree of surgical resection. For non-resectable tumors, patient may opt for chemotherapy and/or radiation.

**CONCLUSION:** Given the rate of recurrence, patients with desmoid tumors should be followed clinically with physical examination and radiographic studies as needed. In addition, given the association of desmoid tumors with FAP it is imperative for these patients to undergo periodic colonoscopies for colorectal cancer screening.

**INTRODUCTION:** Endovascular aneurysm repair (EVAR) has largely replaced open surgical repair of abdominal aortic aneurysm (AAA). Ischemic complications associated with EVAR are infrequent but well recognized.

**CASE:** A 76-year-old Caucasian man presented with moderate dull mid-abdominal pain after walking one block for 8 months. It was associated with mild bloating and burping. He denied fever, chest pain, dyspnea, nausea, vomiting, hematemesis, melena, hematochezia, weight loss, anorexia, dysphagia, diarrhea, or constipation. Pain used to resolve after sitting down, and aggravate 2-3 hours after a meal. Out-patient upper and lower gastrointestinal endoscopies were normal. Patient had hypertension, coronary artery disease, percutaneous coronary angioplasty with stents, atrial fibrillation, hypothyroidism and abdominal aortic aneurysm. His problems were stable on lisinopril, atorvastatin, metoprolol, rivaroxaban, amiodarone, levothyroxine and aspirin. Eight -months ago he underwent endovascular aneurysm repair (EVAR). On examination, his vital were normal, he was obese (BMI=42 Kg/m2), and rest of the physical examination was unremarkable, including soft and non-tender abdomen. Based on his history and temporal relationship a clinical diagnosis of superior mesenteric artery (SMA) stenosis was suspected. A comprehensive metabolic panel, complete blood count, serum amylase, lipase and urinalysis were within normal limits. A contrast enhanced abdominal CAT-scan revealed 90% stenosis of the SMA at its origin secondary to cephalad displacement of the endovascular stent graft. Patient underwent endovascular stenting of the SMA. He remained symptom-free immediately and at 1-year follow-up. A final diagnosis of iatrogenic SMA stenosis post-EVAR was established.

**DISCUSSION:** Ischemic complications following EVAR can be immediate thromboembolism along colonic, renal, pelvic and spinal arteries, or late limb or renal ischemia due to kinking or distal migration of the graft. SMA stenosis has been reported in 1-3 % of cases, mostly early due to microembolization of thrombotic deposits and atheroma in the suprarenal aorta during placement of the
graft. Our case is a unique case where graft migrated proximally occluding the superior mesenteric artery. Patient with SMA stenosis classically presents with chronic abdominal symptoms of nausea, vomiting, early satiety, post prandial pain developing between 10 minutes and 3 hours after a meal. Patient can develop fear of eating and report weight loss. Symptoms of bloody diarrhea and rarely constipation have also been reported. Physical examination can range from normal findings to mild diffuse abdominal tenderness without rebound or guarding. Diagnosis is made by CT abdomen followed by CT angiography. MRA and duplex mesenteric ultrasonography is showing promising results but CTA is arguably the best test for diagnosis. Treatment is open or endovascular revascularization.

**CONCLUSION:** Although SMA stenosis after EVAR is rare, nevertheless it should be suspected in patients who present with chronic abdominal pain post exertion or after meals.

| 50. | Clinical Vignette | Nathan, Karim Satyajeet Roy, MD, FACP Cooper University Hospital (Brian Gable) | Uncontrolled diabetes and perfect glycosylated hemoglobin: Are we missing something? | INTRODUCTION: Hemoglobin A1c (HbA1c) may be an unreliable measure of glycemic control in patients with inherited hemoglobinopathies.

**CASE PRESENTATION:** A 36-year-old African-American woman established care with history of type-2 diabetes mellitus (T2DM) for 2 years. She was asymptomatic. She had a strong family history of T2DM. Although she was taking metformin 500 mg orally twice-a-day, her fasting blood glucose ranged between 200 and 350 mg/dL. She admitted having difficulty in limiting high calorie food intake. She denied use of alcohol, cigarette, or drugs. Her BMI was 36.5 Kg/M2. Her vital signs were normal. Rest of the physical examination was unremarkable. Relevant diagnostic studies showed fasting blood glucose 286 mg/dL, total cholesterol 232 mg/dL, LDL 115 mg/dL, triglycerides 410 mg/dL, hemoglobin 13.8 g/dL and urine microalbumin 13.2 mg/24-hour. Her HbA1c was 4.1%. Considering her longstanding history of elevated fasting glucose levels the value of HbA1c was deemed possible lab error. A repeat HbA1c was 4.0%. An underlying associated hemoglobinopathy was suspected. Her hemoglobin electrophoresis showed 60% HbA (less than 3.5% HbA2) and 40% HbS confirming a diagnosis of sickle cell trait. Metformin dosage was increased and she was counseled for low-calorie diet and exercise, in addition to genetic counseling. Her long-term glycemic control was subsequently monitored by periodic measurement of serum fructosamine levels.

**DISCUSSION:** HbA1c measures the percentage of glycated hemoglobin A, and is usually an excellent measure of glycemic control over the preceding 2-3 months. However, it can be unreliable in monitoring diabetes in people with inherited hemoglobin variants. The two most common variants, HbS and HbC, exist alongside normal HbA in the asymptomatic heterozygote forms known respectively as sickle cell trait and HbC trait. The prevalence of sickle cell trait in the African American community is 8-10% and may be as high as 25-30% in West Africa. Furthermore, the prevalence of T2DM in African-Americans over 20 years of age is about 20%, with a rising disease burden in West Africa as well. Given this epidemiology, clinicians must be aware of potential limitations of the HbA1c test, as it can either over- or underestimate average blood glucose. A hemoglobinopathy should be suspected when blood glucose measurements do not correlate with HbA1c. In such cases, an alternative test, serum fructosamine, a measure of glycated protein, should be considered. However, fructosamine has its own limitations. It measures blood glucose over a shorter period of only 2-3 weeks and is a less standardized assay. Furthermore, there is a lack of evidence validating its use as a marker for glycemic control and microvascular risk. Conclusion: Inherited hemoglobin variants should be considered in patients with discrepancies.
between HbA1c and blood glucose measurement. In such cases, serum fructosamine may be considered as an alternative test.

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<th>Nathan, Karim</th>
<th>Abhishek Agarwal, M.D., Adil Manzoor, Christina Cloke, M.D., Brian Gable, M.D.</th>
<th>Cooper University Hospital (Brian Gable)</th>
<th>Lamotrigene-induced Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS)</th>
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<td><strong>INTRODUCTION:</strong> A 59 year-old woman is diagnosed with lamotrigene-induced drug reaction with eosinophilia and systemic symptoms (DRESS).</td>
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<td><strong>CASE PRESENTATION:</strong> A 59 year-old morbidly obese, Caucasian female was transferred from another hospital for evaluation of fever and systemic rash. The patient noticed the morbilliform rash 3 weeks back diffusely throughout her body, sparing her palms and soles. She was started on levofloxacin by her PCP. The rash progressed in the next couple of weeks with subsequent desquamation and painful, erythematous, fluid filled, blister formation. She also complained of systemic symptoms like malaise, fatigue and dyspnea. 3 weeks prior to onset of rash, she had been started on lamotrigine. On physical exam, a desquamating erythematous rash was seen over her face, lips, torso and extremities. Laboratory results revealed peripheral eosinophilia (14.8%), as well as alanine and aspartate transaminase levels of 58 and 59, respectively. Her Creatinine (Cr) was 1.9 (baseline Cr was 1.3) with 9 WBCs in the urine. No RBC casts or dysmorphic RBCs were seen. A skin biopsy was obtained from her right leg which revealed subepidermal bullous formation with increased eosinophils and neutrophils. She was diagnosed with lamotrigine-induced DRESS. Lamotrigine was discontinued and she was started on systemic steroids, initially IV methylprednisolone 60 mg BID and later oral prednisone 1 mg/kg/day. Her symptoms improved drastically in 2 days after presentation, with improvement of the rash, fatigue and malaise, as well as downtrending of her Cr.</td>
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<td><strong>DISCUSSION:</strong> DRESS is associated with aromatic anticonvulsants, including phenytoin, carbamazepine and barbiturates, as well as many non-aromatic drugs, including lamotrigine, a first-line agent for epilepsy given its overall safety and efficacy. The onset is typically 2 to 8 weeks after initiation of the drug. The first lesions to appear are diffusely erythematous, or morbilliform, and may progress to a generalized exfoliative dermatitis (erythroderma) if the drug is not discontinued. Mucosal membrane involvement includes cheilitis, tonsillitis and erosions. Systemic involvement includes fever, often preceding the rash, as well as hepatitis and interstitial nephritis, which we believe caused her AKI in this case. Eosinophilia is present in 30% of cases, and peripheral blood smear may reveal atypical lymphocytes similar to those in mononucleosis. Histologic examination of the skin reveals lymphocytic infiltrate with or without eosinophils. Management consists of discontinuing the offending agent, as well as prednisone, which usually results in rapid improvement. Follow-up should be sought for resolution of any organ involvement. The pathogenesis of DRESS is not fully understood, but is thought to involve a hypersensitivity reaction to certain drug metabolites.</td>
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<td><strong>CONCLUSION:</strong> Drug reaction with eosinophilia and systemic symptoms (DRESS) is a serious multiorgan syndrome with protean manifestations which can be fatal if not properly recognized and treated with steroids and discontinuation of the offending agent.</td>
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<th>52.</th>
<th>255</th>
<th>Clinical Vignette</th>
<th>Odesanya, Temitayo</th>
<th>Temitayo Odesanya MD, Adil Manzoor</th>
<th>Cooper University</th>
<th>Macrophages Gone Wild</th>
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Macrophages Gone Wild: A 65-year-old female with stage 3B diffuse B-cell lymphoma being treated with R-CHOP was transferred to our institution for escalation of care after developing a multisystem syndrome characterized by fever of unknown origin, altered mental status, hemodilational-dependent acute kidney injury due to rapidly progressive nephrotic syndrome, bicytopenia (hemoglobin 5.6; platelets 31), coagulopathy with low fibrinogen (81 mg/dL), and elevated liver function tests (alkaline phosphatase 960; AST 586; ALT 110). The physical examination was benign with the exception of a recurrent fever and an acute encephalopathy without associated focal neurological deficit. An extensive diagnostic evaluation to assess for underlying infection including blood cultures, urine culture, cerebrospinal fluid analysis, and cross-sectional imaging was negative. There were no schistocytes on the peripheral smear to suggest a microangiopathic hemolytic process such as thrombotic thrombocytopenic purpura-hemolytic uremic syndrome or disseminated intravascular coagulation. Given the constellation of findings, a diagnosis of lymphoma-associated hemophagocytic lymphohistiocytosis (HLH)/macrophage activation syndrome was entertained. This diagnosis was further substantiated by the presence of high triglycerides (1089) and markedly elevated ferritin levels (>100,000 ng/mL). A subsequent bone marrow biopsy revealed extensive erythrophagocytosis. The compilation of findings fulfilled the HLH-2004 diagnostic criteria for hemophagocytic lymphohistiocytosis (HLH) and treatment with the HLH-94 treatment protocol consisting of dexamethasone and etoposide was initiated. Unfortunately, the patient was unable to tolerate the therapy due to her debilitated condition and she ultimately succumbed to the disease. Of note, the result for the soluble CD25 (interleukin-2 receptor) test was made available in the post-mortem period and was found to be markedly elevated at 4906 U/mL (normal range 406-1100 U/mL), further confirming the diagnosis of HLH.

Discussion: Hemophagocytic lymphohistiocytosis (HLH) is an extremely rare, life-threatening diagnostic entity that manifests as a multisystem syndrome with progressive organ dysfunction. Most cases of HLH appear to be due to lymphocyte hyperactivation resulting in a macrophage-driven SIRS-like response that leads to multi-organ failure and a high mortality rate. Potential precipitating events include a hereditary predisposition, certain inciting infections, autoimmune diseases, or neoplastic disease as in our patient. This disease must be considered in the differential diagnosis of multisystem organ failure, but it is frequently overlooked and misdiagnosed as septic shock. Diagnosis is based on 8 clinical criteria for which at least 5 has to be met: (1) fever, (2) splenomegaly, (3) cytopenias affecting ≥2 lineages, (4) hypofibrinogenemia (<150 mg/dL) and/or hypertriglyceridemia (≥265 mg/dL), (5) elevated ferritin (>500 ng/mL), (6) hemophagocytosis, (7) low or absent natural killer cell activity, or (8) elevated soluble CD25 (≥8805; 2400 U/mL). Conclusion: Although well-defined clinical criteria have been established, HLH's rare and extremely variable presentation often delays the diagnosis resulting in an overall poor prognosis.

Clinical Vignette

Patel, Akshar Satyajeet Roy, MD, FACP

Cooper University Hospital (Brian Gable)

A Lone Star in the Garden State

INTRODUCTION: Outpatient presentation of insidious onset subacute fever and absence of additional symptoms and signs can have a plethora of underlying causes which require a cost-effective and specific diagnostic approach.

CASE DESCRIPTION: A 54-year-old Caucasian man with history of hypertension and hyperlipidemia presented with fever (maximum 102.9 degree F) for 5 days, associated with intermittent chills, mild headache, and mild muscle ache. He denied nausea, visual changes, cough, sore throat, rash, abdominal pain, urinary or bowel discomfort. Patient was a native of New Jersey and he denied travelling outside the state. He worked in a wooded area about 2 weeks prior to the onset of his symptoms. Upon further questioning he shared that he removed a tick from his groin which was about 5 mm size, reddish-brown with a white spot. Clinical examination was significant only for elevated temperature (101 degree F). He had no rash, and no nuchal rigidity. Rest of the examination was normal. Blood tests showed high alanine aminotransferase (ALT=79 U/L) and high aspartate aminotransferase.
(AST=59 U/L); and low white blood cell count (WBC=1.5 k/uL), low absolute monocyte count (0.6 K/uL), and low platelets (89 K/uL).

Based on his clinical presentation, lab abnormalities and exposure to a tick that resembled a female Lone Star tick (Amblyomma americanum) a diagnosis of Human monocytic ehrlichiosis (HME) was suspected. Patient was admitted in the hospital where a Buffy coat blood examination revealed presence of morulae in the cytoplasm of monocytes, confirming a diagnosis of HME. He was started on oral Doxycycline. He became afebrile next day. Lab abnormalities returned to the normal limits in 3 days.

**DISCUSSION:** HME presents similarly to other Tick-borne diseases and viral syndromes, with vague symptoms including fever, fatigue, myalgias, arthralgias, headache, and rash. Differentiation from other tick-borne diseases is difficult without laboratory data. A thorough history is critical and suspicion should remain high during the summer when tick exposure is common. The American dog tick and Deer tick can transmit multiple tick-borne diseases (including HME), but the Lone Star tick can only transmit Ehrlichia chaffeensis and E. ewingii, both of which cause HME. The female Lone Star tick has a unique description of a white star on the back and is found in the eastern and south central states. In 2013, there were 52 cases of HME reported in New Jersey. Early treatment with doxycycline is essential for a favorable outcome. It carries a mortality rate of 2-5%.

**CONCLUSION:** Our patient presented with vague symptoms, but key historical data resulted in early diagnosis and treatment of HME, leading to an excellent outcome. HME should be on the differential in all patients who present with flu-like symptoms with a history of tick exposure.

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**Clinical Vignette**

**Background:**

Steroid-responsive encephalopathy associated with autoimmune thyroiditis (Hashimoto’s encephalopathy) is an exceedingly rare syndrome associated with anti-thyroid antibodies that is characterized by a range of neuropsychiatric symptoms. Given its variable presentation, no clear diagnostic criteria have been established. The diagnosis may be overlooked when the evaluation for other etiologies of encephalopathy has been unrevealing.

**Case Presentation:**

A 44 year-old male with a medical history significant for Hashimoto’s thyroiditis with resulting hypothyroidism, vitiligo, and rheumatoid arthritis presented to the emergency department with 3-4 months of chronic and progressive behavioral change that included increased irritability, introversion, insomnia, and the recent development of hallucinations, myoclonus, and tremors. Physical exam revealed frequent episodes of myoclonus, clonus, and symmetric hyperreflexia of the upper and lower extremities. Imaging studies of the brain revealed no significant abnormality. Laboratory studies showed an elevated thyroid stimulating hormone of 156.5 uIU/ml (normal 0.27-4.20 uIU/ml), elevated anti-thyroglobulin antibody: 27 IU/ml (normal ≤ 1 IU/ml), and elevated antithyroperoxidase antibody: 49 IU/ml (normal < 9 IU/mL). Serum vitamin B12, ammonia, cosyntropin stimulation test, lumbar puncture and cerebrospinal fluid analysis for an infectious etiology were normal. Electroencephalography revealed epileptiform activity with generalized sharp and slow waves associated with episodes of bilateral myoclonic jerks. Given the patient’s presenting neurological symptoms in the setting of significant hypothyroidism and elevated anti-thyroid antibodies, a clinical diagnosis of Hashimoto’s encephalopathy was established. The patient was started on intravenous methylprednisolone (1 mg/kg) and within three days of therapy there was complete resolution of both the clinical abnormalities and the epileptiform activity on repeat electroencephalography. The patient was subsequently discharged on oral prednisone (1 mg/kg/day) with plans to taper as an outpatient.
### DISCUSSION:
Steroid-responsive encephalopathy associated with autoimmune thyroiditis (Hashimoto’s encephalopathy) is a diagnosis of exclusion after other causes of encephalopathy have been excluded. It is most commonly encountered in women and is often associated with elevated serum levels of anti-thyroid peroxidase and anti-thyroglobulin antibodies, although the exact pathophysiologic relationship between these antibodies and the encephalopathy remains unknown. Common clinical findings include cognitive impairment, tremor, myoclonus, ataxia, seizures, sleep disturbance, hyperreflexia, and psychosis, which were encountered in our patient. Treatment is high-dose corticosteroid therapy generally tapered to clinical improvement.

### CONCLUSIONS:
Due to its potential to be effectively treated with immunosuppressive therapy with resolution of many life-altering symptoms, Hashimoto’s encephalopathy should be considered in the differential diagnosis of encephalopathy of unknown etiology.

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<th>S5.</th>
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<th>Clinical Vignette</th>
<th>Zarliv, Samson MD, MPH</th>
<th>Abhishek Agarwal, MD, Sajed Sarwar, MD, Brian Gable, MD, FACP</th>
<th>Cooper University Hospital (Brian Gable)</th>
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"Bile Ducts: Now You See Them, Now You Don’t!"

### INTRODUCTION:
Vanishing Bile Duct Syndrome (VBDS) is an extremely rare complication of HIV infection with only 6 cases documented in the literature. The exact etiology is unknown, but associations with nevirapine, antibiotics, viral co-infections, and lymphoma have been reported.

### CASE PRESENTATION:
A 50 year-old male with recently diagnosed HIV/AIDS (CD4 count 26) complicated by disseminated Mycobacterium avium-intracellulare (MAI) infection presented for progressively worsening pruritus, malaise, fatigue, and acholic stools over the preceding 6-8 weeks. Medications on admission included azithromycin, ethambutol, trimethoprim-sulfamethoxazole (TMP/SMX), emtricitabine-tenofovir and ritonavir. Physical examination revealed a severely cachectic African-American male with oral candidiasis, hepatomegaly, and diffuse jaundice with multiple areas of excoriation. The remainder of the examination was normal.

Laboratory testing revealed abnormal liver function tests with a cholestatic pattern as follows: Alkaline phosphatase 1711, gamma glutamyl transferase 2000, total bilirubin 10.1, direct bilirubin 6.8, AST 469, and ALT 162. Hepatitis serologies, Herpes simplex virus (HSV) PCR, Epstein–Barr virus (EBV) PCR, Cytomegalovirus (CMV) PCR, Cryptosporidium serology, and stool ova and parasite studies were negative. Repeat acid fast bacilli (AFB) blood cultures were negative. A right upper quadrant ultrasound and magnetic resonance cholangiopancreatography showed no evidence of extra-hepatic bile duct obstruction. Anti-mitochondrial antibody was negative. A subsequent liver biopsy revealed resolving non-caseating granulomatous hepatitis with evidence of profound ductopenia. Immunohistochemistry and cultures were negative for EBV, CMV, HSV, fungi, bacteria, and spirochetes and there was no evidence of a lymphoproliferative process on flow cytometry.

### DISCUSSION:
Vanishing Bile Duct Syndrome is an exceedingly rare group of acquired disorders characterized by intrahepatic cholestatic jaundice due to progressive destruction and disappearance of small and medium sized intrahepatic bile ducts. Unlike AIDS cholangiopathy, there is no extrahepatic obstruction. Diagnosis is made histologically and is defined by a loss of interlobular and septal bile ducts in greater than 50% of small portal tracts in a given pathological specimen containing at least 10 portal tracts. It’s been described in association with drugs, infection, Graft-Versus-Host-Disease (GVHD), liver allograft rejection, primary biliary cirrhosis, and primary sclerosing cholangitis. Only 6 cases of HIV-associated VBDS have been reported in the literature. Nevirapine, Hodgkin’s lymphoma, EBV, CMV and HCV co-infection have been implicated as potential etiologies. MAI associated hepatitis and exposure to TMP/SMX could be causative in our patient. Prognosis is poor with progressive biliary cirrhosis seen in the majority of patients. In progressive cases, liver transplantation is the only curative option which can be pursued in HAART-controlled HIV cases.
CONCLUSION: In HIV patients presenting with painless intrahepatic cholestasis, a high degree of suspicion is required to establish a diagnosis of VBDS with liver biopsy. This disease process must be distinguished from the more commonly encountered AIDS cholangiopathy, which often presents with painful extrahepatic cholestatic jaundice.

56. Quality Improvement
Khan, Maliha
Cooper University Hospital (Brian Gable)

Improvement in Influenza Vaccination Rates Following Quality Improvement Strategies In a Resident Based Medicine Clinic

BACKGROUND: Each year, the influenza virus is responsible for many hospitalizations associated with a great deal of morbidity and mortality. According to the CDC, from 1996-2000, 560 of every 100,000 adults over the age of 65 were hospitalized for influenza whereas only of every 100,000 individuals between the ages of 18 and 65 required hospitalization. During the 2011 to 2012 flu season, only 37.8% of New Jersey residents > 17 years of age were vaccinated. It was hypothesized that quality improvement and educational intervention would increase the rate of vaccination during the 2013-1014 influenza season as compared to the 2012-2013 season.

METHODS: At that start of the 2013-2014 influenza season, multiple educational modalities were implemented, including information sheets, physician counseling, medical assistant involvement in the screening process and influenza clinics, to help increase the number of patients being vaccinated. Lists of patients seen at the Cooper University Hospital resident clinic during the 2012-2013 and 2013-2014 influenza seasons (defined as September 1 to February 28). The battery of randomly chosen patients was deidentified and patients’ charts were reviewed to determine whether or not they received the influenza vaccination and to look for documentation as to whether or not counseling had been provided. In total, 184 charts from the 2013-2014 season, whereas 378 charts were reviewed for the 2012-2013 season.

RESULTS: Both immunization status and counseling data were obtained. A chi-square test was performed on each set of data. The Chi-square statistic for 2013-2014 was 11.48 with a P value of 0.000704 (significant, p<0.05). Similarly, the results for the 2012-2013 season were significant with a chi-square statistic of 8.0457 with a P value of 0.004561. Furthermore, a chi-square value on all patients was 20.252 with a significant p value of 0.000007 (<0.05); however, when the populations were compared to one another, the chi-square statistic was found to be 26.13 with a P value of 0.3047, which is not significant.

CONCLUSIONS: Although multiple educational interventions were employed in the 2013-2014 influenza season, significance in the number of individuals vaccinated was not observed.

57. Research
Agarwal, Abhishek
Namrata Baxi, Dipanshi Patel, Kenyetta Givans, Anuradha Mookerjee, Vijay Rajput
Cooper University Hospital (Brian Gable)

Evaluation of Mentors by Resident and Fellows in a Structured Mentoring Program in an academic medical center

BACKGROUND: In Graduate Medical Education (GME) there is often a lack of structured academic and professional development programs for trainee. Residents and fellows have different academic and professional growth needs throughout their career.

METHODS: Since 2011, we have had a formal mentoring program in place to foster relationship between faculty and residents and fellows in the Department of Medicine. The mentor and mentees are required to meet face-to-face for a one hour session at least
two times a year at a local restaurant. They can continue their relationship outside of this program as per their needs. Academic scholarship is a major emphasis of this program. Mentees are required to identify their academic mentor. A mentor-mentee contract is signed by both parties. Program Directors help to identify the mentors as needed. We conducted an IRB approved research project by developing an anonymous structured questionnaire, based on prior literature. We identified survey questions in 17 areas of mentor’s qualities and attributes; using a four point agreement scale. Twenty one questions were grouped into four categories, based on the mentor’s personal attributes (honesty, integrity, privacy, enthusiasm, advocacy, and communications), action characteristics (inspiration, feedback, encouragement, approachability, and availability) and the short term and long term career goals of the mentee. A total of 60 residents and 39 fellows from ten specialties of internal medicine completed the survey at the end of the academic year. We compared the perception of residents and fellows about their mentor’s personal attributes and action skills. We also analyzed the difference between residents with a known research interest versus no interest and mentor’s attributes for long term versus short term goals. We used Student’s T Test, Pearson Chi Square and Fisher Exact test for statistical analyses.

RESULTS/DISCUSSION: Overall, fellows were more satisfied with their mentors than residents (p=0.017). The fellows were more satisfied with their mentor’s action characteristics than the residents (P=.045). All residents and fellows with declared research interest were more satisfied with mentor’s attributes and skills to help them with their long term goals. (P=0.046) Junior residents perceived that their mentors were not able to challenge them enough or beyond the check list exercise. These differences between residents and fellows may be due to maturity, established goals, or professional growth. The junior residents were not able to develop as strong a connection with their mentor as their senior colleagues.

CONCLUSION: Mentoring is a dynamic dyad interaction with immediate and long term impact. The junior residents may require different skills and attributes from faculty mentors compared to fellows. This research will help in developing future faculty development and mentoring programs across GME. Residents and fellows with established research interests may benefit with help for long term career goals.

58. 29 Research Khan, Maliha Anjali Desai, MD Cooper University Hospital (Brian Gable) Did we take a sexual history? Sexual History Documentation in an Internal Medicine Resident Clinic

OBJECTIVES: To determine how often internal medicine residents at an urban university hospital ambulatory clinic are documenting sexual history on women of childbearing age, and to recognize a potential lag in this area of documentation and bring forth underlying discrepancies between differences in physician sex, post-graduate year, patient age group, and marital status. Design: Retrospective chart review.

METHODS: A total of 252 charts were reviewed from an internal medicine resident clinic at an urban university hospital. Among the charts reviewed female adult patients between the ages of 21-45 with documentation of an STI/STD, contraceptive use or discussion, family planning, information about the partner, and number of partners, was recorded and considered to be part of a sexual history. Male patients were excluded from the study. Charts documented by post-graduate year 1 and 2 were recorded, which included a total of 37 male and female resident physicians. A Pearson Chi Square test was used to compare the patient’s age group, marital status, resident PGY level, and resident’s gender.

RESULTS: Our study showed that 46% of resident physicians documented some type of sexual history. There was no significant
difference in the marital status of the patient (P 0.303), PGY level of the resident physician (P 0.743), or the gender of the physician (P 0.95). However, among the charts that documented sexual history there was a significant difference between age groups, as older females were less likely to be asked a sexual history compared to younger age group (Mean/ SD 7.34, P 0.008).

**DISCUSSION:** Resident physicians may not always ask and document a detailed sexual history during routine outpatient visits. The CDC and USPTF recommend routine sexual history taking as not only a measure for risk reduction and prevention, but for patient education and development of an effective rapport with patients. One study reports that less than 4% of physicians have patients complete a history form that includes questions about sexual orientation, or practices, and only 10% ask new patients questions specifically to identify those at high risk for HIV (Lewis, 1987).

**CONCLUSION:** Our study demonstrated a similar or higher rate of reporting sexual history taking than others reported in literature. Perhaps this is because our study excluded male patients and limited the female age group to patients of childbearing age. Overall, we found no influence on rates of documentation based on resident gender or year of training. We looked at various variables that pertained to sexual history documentation and noted that sexual activity and partner information were documented most frequently. In conclusion, our study demonstrates that future physicians in training need a greater emphasis on recognizing this area of deficiency and training programs should implement strategies to reinforce taking a better sexual history.

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<th>59.</th>
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<th>Clinical Vignette</th>
<th>Siddiqui, Muhammad</th>
<th>Arthur Rusovici MD FACC, Michael Desiderio MD</th>
<th>Englewood Hospital And Medical Center (Jon Shammash)</th>
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**STRESS INDUCED CARDIOMYOPATHY WITH MID-VENTRICULAR BALLOONING: A RARE VARIANT**

**INTRODUCTION:** Stress cardiomyopathy (SCM) also referred, as the "broken heart syndrome" is a condition in which intense emotional or physical stress can cause fulminant and reversible cardiac muscle weakness. It occurs following a variety of emotional stressors such as grief, fear and extreme anger. Patients with this syndrome have symptoms similar to patients with acute coronary syndrome including chest pain, shortness of breath, congestive heart failure and elevated cardiac enzymes. SCM is a reversible syndrome and should be managed symptomatically. SCM most commonly involves the apical segment of left ventricle but newer and rare variants have recently been seen reported. We here report a case of rare mid ventricular variant of stress related cardiomyopathy.

**CASE PRESENTATION:** A 72 year old female with past medical history only significant for SVT, taking aspirin and digoxin presented with an episode of severe sub-sternal chest pain while walking with her husband. She felt a significant heaviness in her chest and was short of breath. During her hospitalization she was found to have positive cardiac enzymes with a troponin peaking at the level of 3.0. EKG showed 1-mm down-sloping ST segment changes but otherwise unremarkable. The cardiac catheterization showed normal coronaries. However left ventriculogram revealed a very abnormal ventricular function. The mid ventricular portion was found to be dyskinetic. Patient was diagnosed with non-ischemic cardiomyopathy, consistent with mid ventricular SCM. The patient was placed on ACE inhibitor, beta blocker and discharged in a well compensated state to follow-up in 2-3 months for re-evaluation.

**CONCLUSION:** This case reports highlights the atypical or mid-ventricular variant of SCM. However clinical characteristics are similar and differences in mechanism, management, outcome and prognosis have not been reported. Left ventriculogram characteristically shows mid-ventricular ballooning and coronary angiography may be normal or show mild-moderate coronary disease. Management remains empirically treating the cardiomyopathy during this transient syndrome.
A patient with negative Quantiferon-TB gold test developed tuberculosis pleurisy after adalimumab therapy

A 60 year-old Asian female with psoriatic arthritis presented with intermittent fever and dyspnea. She was seen by rheumatologist 12 day-prior to admission and was given 5 day-course of azithromycin. She developed cough, with yellowish sputum and began to feel dyspnea. She went to urgent care clinic 3 day-prior to admission and was given levofloxacin. She was taking prednisone, methotrexate, and adalimumab injection for her psoriatic arthritis. She has been on these medications for 10 months. Prior to initiation of adalimumab treatment, the patient was tested for Quantiferon-TB Gold assay, which was negative. She denied previous history of tuberculosis. On admission, she had normal vital signs. Physical examination revealed diminished breath sound on the right side of her chest. She had white blood cell count of 5,100/µL with lymphocytopenia. Chest x-ray showed a right-sided large pleural effusion. Broad-spectrum antibiotics were started empirically for potential pneumonia. On hospital day 2, thoracentesis was done. Pleural fluid analysis showed mild leukocytosis with lymphocytic dominance, elevated LDH and ADA and negative AFB stain. Result of repeat serum Quantiferon-TB Gold was indeterminate. On hospital day 4, she underwent bronchoscopy which did not show any pathology. The patient still had intermittent fever. All the cultures remained negative including blood, sputum, pleural fluid, and bronchial lavage. On hospital day 9, she underwent right video-assisted thoracoscopy. Pleural specimen showed multiple granuloma with focal necrosis with acid-fast bacilli. RIPE regimen was initiated for treatment of TB pleurisy. Treatment of psoriasis has evolved with TNF-α antagonists including infliximab, adalimumab, and etanercept. However, their use is associated with an increased risk of Mycobacterium tuberculosis infection. There are two types of screening tests for exposure to TB: tuberculin skin test (TST) and Quantiferon-TB Gold (QFT). QFT is more sensitive and specific than TST and less affected by immunosuppression or skin hypersensitivity and also not affected by BCG vaccination. However the utility of QFT in the setting of ongoing biological therapy is questionable given the T-cell responses to QFT antigens are diminished in the presence of anti-TNF agents. Clinical manifestation of TB in patients on biologic agents is often atypical and extrapulmonary, which leads to delayed diagnosis and disseminated TB contributing to the higher rates of morbidity and mortality. This case highlights importance of screening for tuberculosis and diagnostic limitation of QFT in patients on TNF-&#945; antagonist. Physicians should consider TB in their differential diagnoses in these patients with flu-like or pulmonary symptoms. Currently the National Psoriasis Foundation recommends TB screening annually for these patients. However, the time to onset of clinically evident tuberculosis varies, an average 5 months for adalimumab, and 3 months for infliximab. We suggest screening of TB in these patients every 3-6 months with chest x-ray and QFT.

The use of a standard visit template to monitor patients on opioid prescriptions in the primary care clinic: a method for a safe practice.

BACKGROUND: For healthcare professionals, medical errors relating to opioid medications are at the heart of many lawsuits, with most stemming from monitoring-related errors. While there has been improvement in awareness of prescription medications in the US in the past few years, the abuse of prescribed pain relievers continues to rise. In our community primary care clinic, failure of appropriate documentation and monitoring was identified as a major area for improvement to reduce opioid abuse. In this paper, we present an implementation of a template for the electronic medical record system as an intervention to improve our practice in the outpatient primary care clinic.
METHODS: This is a prospective study on the implementation of a template in an electronic medical record system (EMR) to monitor patients receiving opioid prescriptions from our providers. The template was designed for internal medical residents using eClinicalWorks. It was to be used during any patient visit for refills of prescription. We also encouraged physicians to obtain access to the NJ prescription-monitoring program. All patients who used more than two opioid prescriptions in the last year were included. A total of 36 patients were identified. Data were collected after 3 months. Outcome measures included the fraction of patients who signed the opioid contract, the fraction of medical residents who used the template for documentation, the fraction of preceptor physicians who used New Jersey’s Prescription Monitoring Program and the number of patients who were denied further prescriptions for detected aberrant behavior.

RESULTS: All patients receiving chronic pain management had signed opioid contracts, up from 38% before template adoption. 92% of residents used the template during each follow-up visit. Patient compliance with urine toxicology screens increased from 58% to 65% after implementation of the template. 61% of physicians verified the patient’s medication list during pain management visits with New Jersey’s prescription monitoring program. By the end of the study, a total of 21 patients were no longer prescribed opioids for violation of contract, aberrant behaviors or referral to a pain specialist.

CONCLUSION: We developed a template for an electronic medical record system for monitoring pain regimens that include opioids in the primary care clinic. Physicians find the template easy to use. It helps bring non-adherence to opioid regimens to provider’s attention earlier, which improves patient’s safety and physician’s liability.

62. Siddiqui, Muhammad

CODRUTA CHUIZAN PHD, ABDURRAHMAN A HUSAIN MBBS, ANDREW WEISSMAN MD FACC

ENGLEWOOD HOSPITAL AND MEDICAL CENTER (JON SHAMMASH)

TEMPORAL PATTERN OF CABG AND PCI AFTER NON-ST ELEVATION MYOCARDIAL INFARCTION AMONG ELDERLY PATIENTS FROM NHDS

BACKGROUND: Optimal management of elderly patients with Non-ST Elevation Myocardial Infarction (NSTEMI) remains unclear due to underrepresentation in large-scale clinical trials and the increased risk of adverse outcomes. Recent randomized clinical trials have shown improved outcomes among NSTEMI patients receiving invasive management versus conservative medical management, however, how this is reflected in U.S clinical practice for elderly patients has not been reported.

METHODS: We used NHDS data to identify all adults with an International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) code for NSTEMI from the years 2005 to 2009. To investigate the trends in time, we defined invasive therapy as Coronary Artery Bypass Graft or Percutaneous Coronary Intervention and identified their ICD-9 procedure codes in patients diagnosed with NSTEMI. We then stratified the patients according to age >65 or ≤65 and compared the temporal trends between the two age groups.

RESULTS: Among 21,306 patients diagnosed with NSTEMI between 2005 and 2009, the median age was 73 years (IQR: 61-82 years), 54% were males and 57% were White. For all five years, the proportion of patients age>65 receiving invasive management (21%, N=13978) was significantly lower than those age≤65 (41%, N=7328) (p<0.001). Our results also show that in both age groups, the proportion of patients receiving invasive management decreased substantially over time (p<0.001).

CONCLUSIONS: Despite published guidelines promoting use of invasive management for NSTEMI patients, the proportion of
patients receiving intervention gradually decreased from 2005-2009, more so in elderly population, who made up most of the total population of NSTEMI patients; median age 73 yrs. Our future analyses will investigate if this trend maintains after adjusting for other factors thought to be associated with the management of NSTEMI in elderly patients, including sex, co-morbid conditions etc.

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<tr>
<th>63.</th>
<th>180</th>
<th>Clinical Vignette</th>
<th>Kasinathan, Niktha</th>
<th>Shumila Kashif M.D., Douglas Zaeh M.D.</th>
<th>HUMC Mountainside (Douglas Zaeh MD)</th>
<th>HTLV-1 Positive Adult T-Cell Lymphoma</th>
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<td><strong>INTRODUCTION:</strong> Human T-Lymphotrophic virus is a retrovirus that infects millions of people worldwide, however it is associated with disease in only 5 percent of these individuals. One well recognized disease association with the virus is adult T cell leukemia-lymphoma. Given the epidemiology of the virus it is rarely encountered by physicians in North America.</td>
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<td><strong>CASE DESCRIPTION:</strong> 70-year-old Haitian female previously in her usual state of health, presents with left lower quadrant abdominal pain radiating to the flank worsening over the past 3 weeks. The pain was associated with dysuria, frequency, and retention. She also had complaints of constipation, decreased appetite, and chills. She denied any night sweats, change in weight, nausea, vomiting, or diarrhea. Physical exam revealed bilateral cervical lymphadenopathy. Nodes were non-tender, fixed; ranging 1-2 cm. Skin showed an erythematous, papular, non-puritic rash covering the back and shoulders. Patient also had supracubital abdominal tenderness and left costovertebral angle tenderness. Labs on admission were significant for calcium of 16.34, ALP 218, white count of 14.5 with 31% lymphocytes including atypical lymphocytes, 20% smudge cells, and 4% bands. Hemoglobin 11.1, hematocrit 34.1, and platelet count of 241. Electrolytes were significant for mild renal insufficiency with a BUN of 24 and creatinine of 1.4. CT abdomen and pelvis, obtained secondary to abdominal pain, revealed significant bilateral axillary and inguinal lymphadenopathy suspicious for a lymphoproliferative disease, along with an 18mm non obstructing stone in the left kidney. Peripheral blood smear showed atypical hypernucleated lymphocytes, and ELISA was positive for HTLV-1. Further work up with flow cytometry confirmed T cell lymphoma. The diagnosis of HTLV-1 positive adult T cell lymphoma was made and patient underwent chemotherapy treatment. She underwent 5 cycles of combination chemotherapy consisting of etoposide, prednisone, vincristine, and cyclophosphomide. Patient is currently in remission as per her last PET scan.</td>
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<td><strong>DISCUSSION:</strong> Human T-lymphotrophic virus is prevalent in southwest Japan, Caribbean regions, South Africa, and Sub-Saharan African countries. In the United States however, the prevalence of the virus is 0.01-0.02%, and when present is mainly seen in immigrants from endemic countries. The infection remains dormant for 20-30 years before complications such as T-cell lymphoma present. Rapidly progressive skin lesions, hypercalcemia, and lymphocytosis with cells containing lobulated nuclei, or “flower cells,” dominate the clinical picture. HTLV-1 ATL is treated with combination chemotherapy similar to other forms of lymphoma. Along with this treatment, post-chemotherapy interferon alpha and zidovudine may extend survival.</td>
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<th>Clinical Vignette</th>
<th>Kashif, Shumaila</th>
<th>Sunya Ashraf, Vinit Gupta</th>
<th>HUMC Mountainside (Douglas Zaeh MD)</th>
<th>Saline sinus irrigation related to relapsing Gram negative bacilli pneumonia</th>
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<td><strong>Saline sinus irrigation is becoming more widely used to improve sinus symptoms quality of life and decrease medication use in adult subjects with a history of sinusitis. Interestingly, devices used to deliver such therapies consistently showing contamination with pathogenic organisms such as Staphylococcus aureus, Pseudomonas aeruginosa and other gram negative bacilli. A 44 years old man with a history of chronic sinusitis presented with cough with greenish, yellow sputum, generalized malaise and fatigue for two weeks in July 2014. Two months prior patient was admitted for E.coli pneumonia which he was treated for with Bactrim DS as per</strong></td>
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sensitivity but symptoms relapsed within two months. During this encounter, Chest X-Ray & CT chest showed right middle lobe consolidation. Patient was treated with broad spectrum antibiotics but continued to have fever, cough & malaise for which he had under gone bronchoscopy which revealed friable mucosa & bronchial cultures were positive for Pseudomonas & Klebsiella. Pt was continued on antibiotics for 15 days. Additional history revealed that patient has been using a feet long saline irrigation tube with bulging middle portion for months which was the probable cause of bacterial contamination & multi-microbial pneumonia in this young healthy adult. This case illustrates that sinus irrigation bottles/tubes have potential of being contaminated with bacteria if not used with sterile techniques and may result in multi microbial pneumonia. Studies have shown clinical correlation between irrigation bottle contamination and clinical outcomes in endoscopic sinus surgery patients, where Pseudomonas aeruginosa, Acinetobacter baumannii, and Klebsiella pneumoniae were frequently recovered from irrigation bottles. Contamination rate increases when bottles/tubes are used for longer than 1 week. Further studies showed that low concentration HOCl solution can be used as an effective nasal irrigation solution as it is found to have bactericidal, fungicidal, or virucidal effects.

Fibrolamellar Hepatocellular Carcinoma presenting with Extensive Portal Vein Thrombosis

INTRODUCTION: Fibrolamellar hepatocellular carcinoma (FLHCC) is a rare primary malignant tumor of the liver with distinct demographics, etiology, risks factors, tumor markers and prognosis.

CASE REPORT: A 27-year old Hispanic male was admitted to Mountainside Hospital in July 2013 because of diffuse abdominal pain for one week. He denied history of hepatitis, prior transfusions, exposure to aflatoxins, alcohol or illicit drug use. No fever, night sweats or weight loss. Physical exam was normal except for mild right upper quadrant and epigastric tenderness. His laboratory tests showed mild thrombocytopenia of 132. CT abdomen showed extensive thrombosis of left portal vein and right portal vein with multiple areas suspicious for masses in left hepatic lobe. MRI of abdomen showed extensive thrombus involving the entire left portal vein and portion of right portal venous system and main portal vein as well as multiple masses in both hepatic lobes. Alpha-fetoprotein level was 4548. Liver biopsy was consistent with hepatocellular carcinoma with rare features of fibrolamellar type. Due to its extension, tumor was considered inoperable.

DISCUSSION: FLHCC was first described in 1956. It comprises between 1% and 8% of all hepatocellular carcinoma depending on region and age. There is no strong gender or race predilection. Its etiology is still unknown, although a causal relationship with hepatitis B virus and focal nodular hyperplasia following long-term oral contraceptive use has been suggested. Just like our case, patients present with nonspecific symptoms such as abdominal discomfort, weight loss, or malaise. Palpable mass, pain in right upper quadrant, and jaundice might be seen. Uncommonly, gynecomastia, fulminant liver failure, recurrent deep vein thrombosis, Budd-Chiari syndrome, massive ascites can be seen. Diagnosis relies on clinical presentation, imaging and pathology. Liver enzymes can be normal. AFP levels are typically normal, but elevations can be seen in 27% of the cases Vitamin B12 can be elevated. On imaging studies, they are typically hypodense and heterogeneous hepatic masses on nonenhanced CT scan and a hypointense central scar can be seen on MRI. Histologically, the tumor is made up of large polygonal cells with abundant eosinophilic cytoplasm, large vesiculated nuclei and large nucleoli with lamellar fibrosis. The gold standard therapy is surgical, either hepatic resection or liver transplantation. Recurrence is common. The reported 5-year survival rate is 34%. The mean survival rate for nonresectable tumors is 32 months. There is modest long-term benefit from systemic chemotherapy. Regimens include 5-fluorouracil and recombinant interferon alfa-2b. Cases mostly present at advance stages. There are few reported cases of FLC with such an extensive portal vein thrombosis at initial presentation. We present this case because of the rarity of this tumor and to highlight that FLHCC
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<th>Clinical Vignette</th>
<th>Ahmad, Muhammad Omer</th>
<th>Jersey City Medical Center (Amer Syed)</th>
<th>Runaway Temporary Pacemaker, a rare phenomenon</th>
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<td>&quot;Runaway&quot; pacemaker is a rare entity that occurs when a cardiac pacemaker abruptly accelerates its pacing rate above the set upper limit. This can result in life threatening dysrhythmia. Rapid ventricular response to a runaway pacemaker is a potential lethal complication and has been described with permanent pacemakers. We present a case of ventricular tachycardia because of runaway temporary pacemaker. A 61 year old female with past medical history for Sarcoidosis and Rheumatoid arthritis presented with an episode of syncope. She was found to have third degree atrioventricular (AV) block, which was transient in nature. She underwent electrophysiological study to assess conduction system disease secondary to autoimmune process. During the procedure, she was given Procainamide to unmask underlying conduction disease. Subsequently she developed complete AV block with ventricular escape rhythm. A temporary pacemaker was placed and pulse generator was set to deliver 5 mv output to pace on demand at 50 beats per minute. Ventricular paced rhythm at 50 beats per minute was observed. Few minutes later, patient developed paced wide complex tachycardia at 180 beats per minute. Pulse generator settings did not show any errors. Pacing wire position was confirmed again. During the entire episode patient remained hemodynamically stable. As pulse generator was disconnected, wide complex tachycardia terminated on its own and complete AV block was observed without any escape rhythm. A new pulse generator was connected with pacing wire which subsequently showed ventricular paced rhythm without any further complications. No accessory pathway was detected on electrophysiology study. This episode of wide complex tachycardia is consistent with &quot;Runaway pacemaker&quot; which is uncommon but has been reported with permanent pacemakers. Its rarely seen with temporary pulse generators. Underlying etiology include battery failure, component failure or abnormal environmental exposures leading to pacemaker failure and excessive pacing rates. To best of our knowledge, there are only 3 case reports of runaway pacing with temporary pulse generators in the literature. Since temporary pacemakers are frequently used in patients with various bradycarrhythmias, it is very important to recognize this lethal complication of temporary pulse generators. Also as a part of safety precautions, it is important to frequently inspect temporary pacemakers for any kind of malfunction which can result in this complication.</td>
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<th>Clinical Vignette</th>
<th>Baba, Murad Khurram Malik</th>
<th>Jersey City Medical Center (Amer Syed)</th>
<th>Early treatment of Subacute Combined Degeneration of the Spinal Cord in Pernicious Anemia will improve the outcome</th>
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<td>INTRODUCTION: Subacute combined degeneration (SCD) is characterized by demyelination of the dorsal and lateral spinal cord. We report a case of 45 yearold polish man presented with symptoms and signs of SCD, due to severe vitamin B12 deficiency as a result of pernicious anemia. The SCD diagnosis was confirmed by spine MRI. CASE DESCRIPTION: 45 yearold Polish male with a past medical history of hyperthyroidism who presented to the emergency room complaining of two weeks history of upper and lower extremity numbness and stiffness. He reported fatigue and muscular pain all over his body for the last two months, and yellowish discoloration of his eyes that noticed few days ago. He denied any dietary restrictions. Physical examination reveals impaired position sense, impaired vibration sense and &quot;glove and stocking&quot; peripheral paresthesia. Lab findings were significant for macrocytic anemia, indirect hyperbilirubinemia and low level of vitamin B12. Antibodies profile showed positive anti intrinsic factor antibody. Cervical spine MRI showed heterogenous T2 hypointensity with scattered enhancement in the bilateral posterior dorsal columns of the cervical cord with an inverted &quot;V&quot; appearance extending...</td>
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DISCUSSION: SCD refers to degeneration of the posterior and lateral columns of the spinal cord as a result of vitamin B12 deficiency which is the most common cause and usually it is associated with pernicious anemia. In the stomach, cobalamin is bound to intrinsic factor, a glycoprotein produced by the parietal cells of the stomach. The cobalamin-intrinsic factor complex is transported to the terminal ileum, where it is absorbed. The common setting of vitamin B12 deficiency is pernicious anemia, an autoimmune disorder caused by antibodies against gastric parietal cells and the intrinsic factor. Pernicious anemia can present with severe hematologic conditions and even neuropsychiatric illness known as megaloblastic madness. Our patient has neurologic abnormalities that are consistent with vitamin B12 deficiency, which was confirmed with a low level of vitamin B12. In the absence of dietary restriction or a known cause of malabsorption, further evaluation is warranted; testing for pernicious anemia (anti-intrinsic factor antibodies) was positive. Parenteral vitamin B12 treatment (10 loading injections of 1000 μg daily, followed by monthly 1000-μg injections) is an effective therapy. Effective vitamin replacement will correct blood counts in two months and correct or improve neurologic signs and symptoms within six months.

CONCLUSIONS: B12 therapy is reported to stop progression and improve neurologic deficits in most patients with SCD. After three months of parenteral injections the patient reported significant improvement in his symptoms, with almost complete resolution in six months. Early identification of vitamin B12 deficiency and prompt diagnosis of SCD could avoid any irreversible neurologic damages and prevent disability by early parenteral vitamin B12 treatment.

68. Clinical Vignette
Chevli, Parag Dogra, Danny Haddad, Simon Badin

Squamous cell carcinoma of de novo kidney allograft: An extremely rare tumor

INTRODUCTION: Squamous cell carcinoma (SCC) of the renal pelvis is a rare tumor. These tumors are associated with a poorer prognosis than urothelial tumors because they tend to be sessile and deeply invasive at presentation. SCC of de novo kidney allograft is an extremely rare presentation.

CASE DESCRIPTION: A 68 year old Egyptian male came with complaints of nausea, non-bilious, non-bloody vomiting, mild right lower quadrant abdominal pain and generalized weakness for one week associated with decreased appetite and 15 pounds weight loss in 2-3 months. He denied any fever or chills. His review of systems was unremarkable otherwise. His past medical history was significant for ESRD for which he underwent allogenic renal transplant from his wife around 15 years ago and essential hypertension. His medications included metoprolol, tacrolimus, mycophenolate mofetil and prednisone. His physical examination was only significant for mild right lower quadrant around transplanted kidney. There was no lymphadenopathy appreciated. His laboratory examination was significant for normocytic anemia with Hb of 8, bicarbonate of 7, BUN of 78 and creatinine of 7.3 which was worse than his baseline creatinine of around 2.0. The urine analysis showed positive leukocyte esterase, protein of 500 and numerous RBC but no cast. Based on the available data the differential diagnoses included acute kidney injury, UTI, post-obstructive uropathy and chronic allograft rejection. The ultrasound of kidney was ordered which revealed 8.5 cm X 8-cm solid mass in superior pole of transplanted kidney. To further evaluate the mass CT scan of the abdomen and pelvis with intravenous contrast was obtained which showed an ill defined 10.7 X 7.2 X 9.1cm heterogeneously enhancing mass at the superior border of transplanted kidney and a centrally necrotic enlarged aortocaval lymph node. Based on imaging there was a suspicious for renal cell carcinoma with lymph
node metastasis. The patient underwent nephrectomy of transplant kidney. The metastatic workup did not reveal any distant metastasis. The pathology report showed squamous cell carcinoma of renal pelvis of transplanted kidney. Also the fact that patient was from Egypt and was found to have squamous cell carcinoma, he underwent cystoscopy with biopsy which was negative for Schistosomiasis or bladder cancer. Retrospectively efforts were made to find any skin lesion suspicious for cancer but it was negative. The patient remained disease free for 2 months and was planned for cisplatin based adjuvant chemotherapy.

**DISCUSSION:** Characteristics of de novo kidney allograft RCCs is currently unknown. The largest study including 41806 renal transplant patients showed an incidence of 0.19% out of which most of them were papillary carcinomas. Review of literature suggested current primary treatment of renal squamous cell carcinoma is nephrectomy. Adjuvant chemotherapy or radiotherapy is indicated in metastatic disease.

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<th>Clinical Vignette</th>
<th>Chevli, Parag</th>
<th>Prerna Dogra, Valentin Marian, Carrie Wasserman, Amer Syed</th>
<th>Jersey City Medical Center (Amer Syed)</th>
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<td><strong>DRESS SYNDROME: A DECEPTIVE DISEASE</strong></td>
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**INTRODUCTION:** The life-threatening DRESS (drug rash with eosinophilia and systemic symptoms) syndrome is characterized by the presence of at least three of the following findings: fever, exanthema, eosinophilia, atypical circulating lymphocytes, lymphadenopathy, and hepatitis. This syndrome is difficult to diagnose, as many of its clinical features mimic those found with other serious systemic disorders.

**CASE DESCRIPTION:** A 70 year old Hispanic male came with complaint of 3 days of cough with yellowish productive sputum, left sided pleuritic chest pain, fever and chills. He also complained of rash developed about 2 days ago that started on lower extremity extending to his chest and back. He was admitted in the other hospital 1 week ago for the similar complaints for which he was treated with antibiotics. His past medical history was significant for hypertension, diastolic heart failure, migraine and benign prostatic hyperplasia. His primary care physician prescribed him topiramate around 2 months ago for migraine headache. He denied any toxic habits. Physical examination was significant for diffuse macular erythematous rash over bilateral lower extremity, chest and back. His vitals signs were unremarkable. The laboratory examination revealed WBC count of 20,000 with 46% of eosinophils, mild elevation of liver chemistry test and mild proteinuria. The CT scan of the chest revealed mediastinal and hilar lymphadenopathy. Based on significant eosinophilia and lymphadenopathy, decision was made to do open lung biopsy, which revealed poorly formed granuloma and eosinophilic infiltration of pulmonary parenchyma and small vessel vasculitis. The biopsy of the lymph node showed reactive and hyperplastic lymph node. The bone marrow aspirate was performed which was normal. The differential diagnosis included eosinophilic granulomatosis with polyangiitis (EGEP), hematologic malignancies and DRESS. Based on history of recently prescribed topiramate, development of rash, significant eosinophilia, lymphadenopathy and organ involvement, the patient was given a presumed diagnosis of DRESS. The topiramate was stopped in the hospital. He was discharged home after his rash resolved and was followed in the clinic where he was found to have his eosinophil counts subsequently normalized. Relatively rapid and total resolution of all signs and symptoms in less than 3 months without any form of treatment (other than discontinuation of topiramate) argues against most of the differential diagnosis except DRESS.

**DISCUSSION:** This case represents how DRESS syndrome can mimic other malignant or autoimmune condition. Aromatic anticonvulsant agents and allopurinol are the most frequently reported causes although nonaromatic anticonvulsants such as topiramate are considered safe. As per our knowledge this was probably the first case of topiramate induced DRESS. Discontinuation
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| 70. | 319         | Clinical Vignette Chevli, Parag Krithika, Suri, Valentin, Marian     | Jersey City Medical Center (Amer Syed)                                  | Pulmonary arterial hypertension in a patient with anti-CCP antibodies without clinical signs of Rheumatoid Arthritis  
INTRODUCTION: Anti-CCP antibodies are highly specific for Rheumatoid arthritis (RA). Citrullinated extracellular fibrin in the RA synovium may be one of the major autoantigens driving the local immune response, suggested by the discovery of local production of anti-CCP and anti-citrullinated filaggrin antibodies in the joint. A similar mechanism is involved in the pathogenesis of interstitial lung disease in RA. We describe a case of a patient with interstitial lung disease and pulmonary arterial hypertension with anti-ccp antibodies without clinical signs of RA.  
CASE DESCRIPTION: A 57 year old African American female came with complaints of progressively worsening exertional shortness of breath for the last 6 months associated with chest pain. She had a dry cough for several years. She also noticed bilateral leg swelling and an unintentional weight loss of around 10lbs in the last 3 months. On review of systems, she complained of non specific joint aches in her hands, shoulders and knees although denied morning stiffness. She was an ex-smoker with a 22 pack-year smoking history and had quit 14 years ago. She worked in the metro station and had a history of exposure to fumes. Her physical examination revealed a raised JVD, loud S2 and bilateral lower extremity pitting edema. There was no evidence of synovitis, tender joints, joint deformities, raynaud’s phenomenon or dilated capillaries in the nail fold. The laboratory examination revealed an elevated CRP, normal ESR, positive RF and anti-CCP antibody at a titer >250 units. Other studies including complement levels, ANCA screen, Cryoglobulin, ScI-70 Ab, JO-1 Ab, Anti-Centromere Ab were negative. Chest X-ray showed no signs of infiltrate or effusion. Echocardiography revealed dilated right sided structures with normal LV size and function. PFTs showed mild obstructive and restrictive defect but was unable to perform the DLCO. To investigate the cause of her right heart failure further we obtained a V/Q scan and a CT thorax. The V/Q scan showed multiple perfusion defects. The CT scan of the chest showed no evidence of pulmonary emboli, but revealed bilateral diffuse ground glass opacities making vasculitis a possibility. A right heart catheterization with adenosine vaso-reactivity was performed that showed an RVSP of 62mmHg and a pulmonary vascular resistance of 11.75 Woods units that was reduced to 6.68 Woods units after the administration of 400 mcg of adenosine. This confirmed the strong presence of a pulmonary arterial hypertension component to the pulmonary hypertension. The patient was started on inhaled prostacyclin with subsequent improvement in symptoms.  
DISCUSSION: Rheumatoid arthritis is known to be associated with ILD but pulmonary arterial hypertension in seldom recognized in these patients. Echocardiography and subsequent right heart catheterization must be a part of the diagnostic workup of these patients to ensure adequate treatment and symptom improvement. |
CASE SUMMARY:
A 20 year old African American Male presented with 1 day history of bilateral throbbing headache, nausea, vomiting. On questioning, he reported experiencing flu like symptoms 2-3 weeks ago with fever, sore throat and fatigue that lasted for around 5 days. He was found to have a blood pressure of 210/110 at presentation and lab works were significant for BUN of 24, creatinine of 5.5 and proteinuria of 13.4 grams in 24 hours urine collection. Renal biopsy was consistent with a diagnosis of Focal Segmental Glomerulosclerosis- collapsing variant. A detailed work up for HIV, CMV, Parvo Virus B19 and connective tissue disorder was negative. IgM for EBV was positive at a level of 1.49 and IgG was negative suggesting acute infection with EBV. Prominent adenoid tonsils were found on CT Head. A diagnosis of acute EBV infection related collapsing glomerulopathy was made based on the temporal relation with the infection, kidney biopsy finding and presence of IgM antibodies in the absence of IgG antibodies against EBV.

DISCUSSION:
The association between acute infection with EBV and renal disease is well known with some studies showing renal involvement in as high as 15% of the cases of acute EBV infection. The renal manifestations of EBV infections are mostly limited to mesangial proliferative glomerulonephritis, crescentic glomerulonephritis, minimal change disease, membranous nephropathy and interstitial nephritis. Although the association between EBV and collapsing FSGS has been mentioned in literature, the association has been too weak to establish a casual relationship. We report one such rare case of collapsing glomerulopathy with a fairly acceptable casual relation with acute infection with EBV and emphasize that collapsing FSGS secondary to EBV should be suspected in clinically relevant cases.

DRAMATIC RESPONSE TO INFliximAB IN REFRACTORY NEUROSARCOIDOSIS COMPLICATED BY CRYPTOCOCCAL MENINGITIS

INTRODUCTION:
Neurologic involvement occurs in approximately 5% of patients with sarcoidosis and can be the presenting feature. Corticosteroids are the first line agents for the treatment of neurosarcoidosis. Patients who deteriorate in spite of aggressive corticosteroid treatment, who cannot tolerate corticosteroids, or who have a primary contraindication to corticosteroid treatment may benefit from alternative therapies. We report a case of refractory neurosarcoidosis complicated with cryptococcal meningitis that showed dramatic improvement in clinical and radiological manifestation after treatment with Infliximab.

CASE DESCRIPTION: A 44 year old African American female presented with two weeks of progressive left sided hearing loss and facial droop associated with gait imbalance. MRI of brain showed multiple ring enhancing lesions and CT scan of the chest was significant for bilateral hilar lymphadenopathy. After intensive work up including biopsy of right lung showing non-caseating granulomas and excluding other inflammatory, infectous and malignant etiologies, patient was given a diagnosis of pulmonary and neurosarcoidosis. She was started on treatment with prednisone and discharged. After two weeks, she presented with lethargy and fever of 102 F. MRI of brain showed improvement in the brain lesions but lumbar puncture was significant for positive Indian Ink stain and elevated Cryptococcal Antigen (CAg) titers. She was started on two weeks of Amphotericin and Flucytosine as induction therapy. Due to failure to improve after 10 days, it was planned to continue induction therapy for another four weeks along with serial lumbar puncture every 48 hours. After four weeks of induction therapy, there was no improvement along with persistently high CAg titers and opening pressures. MRI of Brain was performed which showed worsening brain lesions and it was believed that patient had worsening of neurosarcoidosis along with cryptococcal meningitis leading to failure in improvement. It was decided to start treatment with Infliximab infusion. After first dose of Infliximab, slight improvement in mental status was noted with decreasing...
CAg. After the second and third infliximab infusion patient showed dramatic improvement in mental status along with resolving menigitis. A repeat MRI of brain revealed significant improvement along with resolution of lesions in multiple areas. The induction therapy was completed and maintenance therapy with fluconazole was started. Patient was discharged to rehabilitation center with outpatient infliximab infusions.

DISCUSSION: This case represents the difficulty in managing neurosarcoidosis especially in patients with disease itself or treatment related complications occur. While the etiology of the disease is still unknown, we now understand that tumor necrosis factor-α (TNFα) plays a pivotal role in the development of the granulomas and it is believed to be involved in the pathogenesis of the disease. Taking advantage of this better understanding of disease pathogenesis, anti-TNFα agents are being increasingly used to treat refractory sarcoidosis.

73. 155
Clinical Vignette
Paudel, Robin
P Dogra, S Suman, L Dominguez, C Wasserman, S Badin
Jersey City Medical Center (Amer Syed)
A Hematological Menace: Multiple Venous Thrombosis complicated by Acquired Factor VIII Deficiency

INTRODUCTION: Acquired Hemophilia A is a disease entity with a classic presentation of spontaneous bleeding leading to large hematomas, ecchymosis and mucosal bleeding. But due to its rarity, diagnosing this condition can be a challenge particularly in a setting where the patient has been on anticoagulation which can delay the proper treatment for this life threatening condition. We present a case with life threatening bleeding caused by acquired factor VIII deficiency complicating the treatment of multiple venous thrombosis.

CASE SUMMARY: A 21 year old African American male with no past medical history presented with Gunshot wounds causing multiple organ injuries leading exploratory laparotomy, repair of abdominal wall, colonic resection, splenectomy, and gastric repair. 13 days post-surgery, patient developed right femoral vein thrombosis while receiving prophylactic dose of enoxaparin. Therapeutic dose of enoxaparin was started and continued with warfarin until therapeutic INR was achieved after which he developed bilateral cephalic vein thrombosis. Before any changes could be made, patient started bleeding from multiple mucosal sites with abdominal hematoma and was found to have a deranged coagulation profile. Warfarin was held and multiple bags of FFP were given but the bleeding failed to improve with lab works now showing elevated PTT (>100) with fairly normal PT and marginally elevated INR. Extensive workup ruled out other common causes of bleeding and coagulopathy and a diagnosis of acquired factor VIII deficiency was made. Factor VIII activity was <1% and factor VIII inhibitor level of 12 Bethesda unit. Patient was treated with high dose steroids and recombinant factor Vila with immediate improvement after which treatment with Rituximab was initiated. Follow up lab works showed decreasing levels of factor VIII inhibitor.

DISCUSSION: Acquired factor VIII deficiency is caused by autoantibody against Factor VIII. Although it generally presents with a classic picture of spontaneous or post surgical bleeding, it is a diagnostic menace to physicians because of its extreme rarity, especially when patients are on anticoagulation. As a result, there is often a delay in diagnosis of this potentially life threatening condition which presents as a major bleed 87% of the time and with 22% mortality. Elevated PTT with normal PT and INR on a patient with previously normal PTT or no prior history of bleeding diathesis can be a strong clue. Therefore specific lab tests like mixing studies followed by factor VIII activity and Factor VIII inhibitors should be considered in the proper clinical setting as early diagnosis and treatment can prove to be life-saving.
| 74. | 278 | Clinical Vignette | Paudel, Robin | Saurav Suman, Prerna Dogra, Jyoti Matta | Jersey City Medical Center (Amer Syed) | Acute Liver and Renal Failure: A rare adverse effect exclusive to intravenous form of amiodarone

INTRODUCTION: Limited knowledge about the mechanism behind acute liver and renal failure secondary to intravenous amiodarone always puts a physician in a dilemma whether to start a potentially lifesaving medication once the acute effect has resolved or to withhold it altogether owing to the adverse effects exclusively seen with the intravenous (IV) form. We report an unusual case of acute liver and renal failure within 24 hours of initiation of IV amiodarone which reversed after discontinuation of the medication.

CASE SUMMARY: 65 years old male with past medical history of ischemic cardiomyopathy with Automatic Implantable Cardioverter Defibrillator (AICD) presented with complaints of multiple AICD shocks. Lab works at presentation were within normal limits. AICD interrogation showed 16 episodes of Ventricular Tachycardias with 10 episodes of appropriate AICD firing. Patient was admitted to intensive care unit for IV loading of amiodarone over 24 hours after which his lab work revealed acute liver failure and also acute renal failure requiring emergent dialysis. Amiodarone was discontinued and a detailed work up ruled out any other possible etiology behind liver and renal failure. Patient was monitored in the hospital for 5 days for downward trend of Liver Function Test and creatinine and was discharged home with arrangements for further follow-up. After one month, oral amiodarone was started which patient tolerated well with follow up lab work done at 6 months showing normal values.

DISCUSSION: Amiodarone is an antiarrhythmic drug highly effective against a wide spectrum of ventricular tachyarrhythmias. Amiodarone is notoriously known to cause various adverse effects. While most of the known toxic effects of amiodarone is seen on prolonged oral use, for reasons unknown, few of the rare side effects that include acute liver failure, cardiac arrest, Acute Respiratory Distress Syndrome (ARDS), renal failure and hypotension are almost exclusively seen with the IV administration of amiodarone and not with oral loading or maintenance dosing of amiodarone. The most widely accepted mechanisms are ischemic liver injury secondary to relative hypotension in the setting of congestive hepatopathy, E-ferol syndrome or cardio-renal syndrome causing renal failure. Irrespective of the etiology, acute toxicity of IV amiodarone is a distinctly different entity from the chronic toxicities of oral amiodarone, and this case supports the recommendation of not withholding the potentially lifesaving oral amiodarone after the acute toxicity due to IV amiodarone has resolved, although further monitoring of the liver enzymes and renal function is recommended.

75. | 314 | Clinical Vignette | Shaik, Imam H | Jennifer Mouri, Rumana Khan, David Flores, Valentin Marian, Amer Syed | Jersey City Medical Center (Amer Syed) | Moyamoya syndrome associated with Thyroid Storm and Diabetic Ketoacidosis: A debilitating condition

INTRODUCTION: Moyamoya syndrome is characterized by progressive narrowing of the bilateral intracranial and extracranial arteries leading to ischemic and hemorrhagic strokes in young adults. Although first described in Japan, there have been numerous cases in the US. Approximately 10-14% of these cases occur in Hispanics. Its association with Graves’s disease is well documented in case reports, but the mechanism is still unclear. Elevated thyroid antibodies in these patients suggest an autoimmune phenomenon. Here we report a young female with an acute onset of hemorrhagic stroke in the setting of severe thyroid storm and diabetic ketoacidosis (DKA) with MRA findings consistent with Moyamoya syndrome.

CASE DESCRIPTION: A 30-year-old Hispanic female was brought to the ED for altered mental status. She had suffered from fatigue,
insomnia and a subjective feeling of being hot for two weeks prior to admission. Her past medical history is consistent with Grave's disease and insulin dependent diabetes mellitus (IDDM). Her home medications include Insulin, Propylthiouracil, Metoprolol but not compliant with her medications. Laboratory tests revealed severe diabetic ketoacidosis and thyroid storm (TSH <0.01; T4 9.10; Thyroglobulin Antibodies 26; Thyroid peroxidase Antibodies >100) with EKG changes consistent with supraventricular tachycardia. Poor neurological status required brain imaging. The CT-head revealed a hemorrhagic stroke in the patient’s left basal ganglia. A MRI/MRA showed stenosis of right Middle Cerebral Artery, reduced flow bilaterally in the Posterior Cerebral Artery, an occlusion of the right Internal Carotid Artery (ICA) and reduced caliber of the left ICA with multiple areas of infarctions. The diagnosis is consistent with Moyamoya syndrome with multivessel stenoses and collateralization on MRA in association of Thyroid storm. She was treated with insulin, beta-blockers, antithyroid, anti-platelet medications with improved DKA and Thyroid storm but required long term ventilator support with poor neurological status requiring tracheostomy and tube feeding support.

DISCUSSION: Decreased cerebral blood flow resulting from stenosis of the distal internal carotid arteries and several other Circle of Willis vessels, including the proximal ACA and MCA is seen in Moyamoya disease. Collateral vessels develop around the areas of stenosis to compensate for the impeded blood flow. Early recognition of symptoms and prompt treatment with antithyroid medications, plasmapheresis, antiplatelets, and steroids has some benefit and in refractory cases surgical revascularization improves the outcome.

76. Clinical Vignette
Suman, Saurav
Robin Paudel, Matthew Delfiner, Rao Mikkilineni, Amer Syed
Jersey City Medical Center (Amer Syed)
Hyperammonemia: Its not always liver

INTRODUCTION: Altered mental status (AMS) is one of the leading causes of elderly patients presenting to the emergency department (ED). Hyperammonemia is often missed as a cause of AMS in patients with no prior history of liver dysfunction. We are reporting a case of a patient with AMS that was found to have hyperammonemia with a normal liver.

CASE PRESENTATION: This is the case of a 68-year-old Hispanic woman with a history of hypertension, non-insulin dependent diabetes mellitus and dyslipidemia who presented to the ED with AMS for 1 day. Collateral history from the patient’s daughter revealed that the patient had been having chronic constipation for some time with intermittent episodes of diarrhea. Her home medications included losartan/hydrochlorothiazide, simvastatin, glimepiride, and sitagliptin/metformin. She had no history of alcohol consumption or liver dysfunction. In the ED, her vitals were stable. Her Glasgow Coma Score was determined to be 5 (E1V1M3) and physical examination was significant for generalized abdominal tenderness to palpation and hyperactive bowel sounds. Initial lab values were significant for a white blood cell count of 22.3, BUN of 30.0 and creatinine of 1.5. The patient’s aspartate aminotransferase was 66 IU/L while her alanine aminotransferase was 38 IU/L. The alkaline phosphatase was 135 IU/L, total protein 7.6, and albumin 3.3. Her lactic acid was 5.1 and ammonia was 102 UMOL/L. A stool sample was positive for Clostridium difficile antigen but negative for the toxin. The patient was admitted to the intensive care unit for further evaluation. A CT of the abdomen with PO and IV contrast was done and demonstrated splenic and gastric varices with a splenorenal shunt and a diminutive but patent portal vein. This finding suggested that venous blood draining from the intestines, and thus rich in ammonia from gut flora (especially during chronic constipation), would bypass liver detoxification because of the splenorenal shunt and enter systemic circulation through the renal veins into the inferior vena cava. The ammonia could then enter the cerebrospinal fluid and produce an AMS. Ultrasonography of liver did not show any evidence of parenchymal abnormality of the liver or increased portal...
vein pressure. The patient’s hyperammonemia was managed with lactulose and C. difficile was treated with metronidazole and vancomycin. Her mental status improved, and she was discharged home on lactulose.

DISCUSSION: The presence of hyperammonemia without liver dysfunction can go unnoticed. There have been case reports of congenital splenorenal shunts, which can bypass the excess ammonia from the liver directly to the systemic circulation. Enzyme deficiency in the urea cycle can also produce hyperammonemia in a normal liver.

CONCLUSION: The presence of splenorenal shunts should be considered in patients with elevated ammonia level in the absence of significant liver dysfunction.

77. Clinical Vignette

Suman, Saurav Soniyashri Koochana, Preerna Dogra, Valentin Marian, Amer Syed Jersey City Medical Center (Amer Syed)

Diffuse Proliferative Lupus Nephritis with Thrombotic Microangiopathy: A Management Nightmare

INTRODUCTION: Diffuse proliferative glomerulonephritis (DPGN) is the Class IV of ISN/RPS classification model for glomerulonephritis in SLE. Association of thrombotic microangiopathy (TMA) in the setting of Anti Phospholipid antibody Syndrome (APS) with diffuse proliferative glomerulonephritis further accelerates the kidney damage and dramatically worsens the prognosis of DPGN. We report a case of DPGN with TMA that possessed various management challenges despite technological advances.

CASE PRESENTATION: 43 y/o Filipino female with past medical history of Anemia and unprovoked left leg DVT presented to ER with 3 weeks history of sore throat, painful swallowing, bilateral lower extremity swelling and multiple small joint pain in the fingers. Physical examination was significant for mucosal ulceration on hard palate, small infarcts on fingertips, bilateral lower extremity 3+ pitting edema and nail fold capillaroscopy revealed dilated capillary loops. Patient was normotensive with unremarkable vitals. Initial lab work revealed pancytopenia (WBC: 1.9, Hb: 6.3, Platelets: 96), elevated ESR, elevated PTT (which did not correct on mixing study and confirmed by dRVVT, consistent with the presence of Lupus inhibitor effect), high-level multiple anti phospholipid antibodies and normal creatinine. Patient’s history of DVT along with positive lupus anticoagulant was suggestive of APS associated with SLE. In addition, the presence of synovitis, mucosal lesions, pancytopenia, high positive dsDNA and depleted complement proteins were indicative of a severe Lupus flare. In the meanwhile, patient was became hypertensive with proteinuria; kidney biopsy was performed which revealed class IV lupus nephritis with diffuse thrombotic microangiopathy. Decision was made to start anticoagulation (five days post renal biopsy) in addition to corticosteroid therapy for severe APS, perhaps imminent catastrophic (CAPS) form, giving the abrupt onset of disease with thrombosis in the kidney, skin and deep vein. Further course was complicated with large perinephric hematoma requiring endovascular embolization of culprit renal vessel and anticoagulation was stopped. Thereafter patient was managed medically on aspirin, mycophenolate mofetil, prednisone and plaquenil. Patient stayed in the hospital for 4 weeks and her symptoms resolved with repeat labs showed downward trend of dsDNA level and normalization of complements.

DISCUSSION: Diffuse proliferative Glomerulonephritis (Class IV Lupus Nephritis) is the most aggressive form of lupus nephritis. The association of TMA with DPGN has been found in around 8% of the cases and it portends bad prognosis. There are no standard treatment guidelines regarding the management of DPGN with TMA. For this case we decided to add anticoagulation to corticosteroids for imminent CAPS, but it was complicated with retroperitoneal hemodynamically significant bleeding and thus patient was treated with immunosuppressive medications alone without the anticoagulation. Current recommendations for
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<th>Tarar, Omer</th>
<th>Murad Baba, Amer Syed</th>
<th>Jersey City Medical Center (Amer Syed)</th>
<th>A Rare Case of Spontaneous Pneumocephalus Associated with Non-traumatic Cerebrospinal Fluid Leak</th>
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<td><strong>INTRODUCTION:</strong> Pneumocephalus (PNC) is defined as pathological collection of gas within the cranial cavity accumulating in the epidural, subdural, subarachnoid, intraventricular, or intraparenchymal compartments. Spontaneous non traumatic PNC and non traumatic spontaneous cerebrospinal fluid (CSF) leaks are both very uncommon conditions. We report a rare case of spontaneous pneumocephalus associated with CSF leak secondary to right sphenoid sinus bony defect without history of trauma.</td>
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<td><strong>CASE DESCRIPTION:</strong> 51 year old Hispanic female with past medical history of hypertension presented to the emergency room with three week history of progressively worsening right side headache associated with clear discharge from right nostril which was aggravated by bending forward and straining. She denied any history of trauma. She reported having flu with frequent sneezing episodes around the same time. She had similar symptoms four months ago which resolved spontaneously, CT scan of the brain at that time was unremarkable. Physical examination was significant for right frontal sinus tenderness and clear discharge from right nostril. Initial lab work did not reveal any significant abnormality. CT scan of the brain showed moderate amount of extra-axial air within the right cerebral hemisphere indicative of pneumocephalus. CT scan of facial bones showed focal bony defect along thin roof of right sphenoid sinus with abnormal CSF collection immediately above and within the lateral recess of the right sphenoid sinus. Whole body bone scan, CT chest, abdomen and pelvis were done and malignancy was ruled out at the time. The patient was started on intravenous antibiotics for meningitis prophylaxis and subsequently underwent trans-sphenoidal repair of cerebrospinal fluid leak with abdominal fat graft and lumbar drain placement. CSF rhinorrhea stopped completely after the surgery, the lumbar drain was removed after six days and repeat brain CT scan showed near complete resolution of right hemispheric pneumocephalus.</td>
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<td><strong>DISCUSSION:</strong> PNC is associated with several etiological factors, including head injury, surgical procedures, infection and neoplasm. PNC is particularly frequent after head injury with concomitant skull base fractures and CSF leakage. Persistent or tension PNC may cause headache, lethargy, and neurological deterioration. Persistent PNC reflects an abnormal communication between the intradural space and external environment, creating a risk factor for central nervous system infection. Spontaneous, non-traumatic pneumocephalus is very uncommon, most cases result from nose blowing, sneezing or valsalva maneuver and will require a surgical intervention. Conclusions: Few cases were described in the literature and most were treated surgically. Our rare case presented with non-traumatic CSF leak and spontaneous pneumocephalus, early identification of such cases and surgical intervention can help decrease the morbidity and the complications that can happen. Keywords: Spontaneous Pneumocephalus, Non-traumatic CSF leak.</td>
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<td>Clinical Vignette</td>
<td>Tarar, Omer</td>
<td>Muhammad Ahmad, Salman Arif, Ramsha Javed, Muhammad Javed, Saleem Mahmood</td>
<td>Jersey City Medical Center (Amer Syed)</td>
<td>Anemia leading to diagnosis of paravalvular leak</td>
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<td><strong>INTRODUCTION:</strong> Hemolytic anemia secondary to paravalvular leak has been reported commonly with mechanical valves but is relatively rare with bioprosthetic valves. We present a case of severe persistent hemolytic anemia due to paravalvular leak two months after porcine bioprosthetic mitral valve replacement requiring surgical intervention.</td>
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**CASE DESCRIPTION:** A 65 year old African American Male with past medical history of Hypertension, Chronic Kidney disease, Gout and Mitral valve prolapse status-post porcine bioprosthetic mitral valve replacement 2 months ago presented to the emergency department with sudden onset left upper extremity weakness. Review of system was positive for generalized weakness and fatigue since one month. He was recently discharged from the hospital a month ago after he presented with symptomatic anemia and gastrointestinal bleed was ruled out at the time. Physical exam did not reveal any focal neurologic abnormality at time of presentation. Magnetic resonant imaging (MRI) of brain showed tiny acute right parietal infarct. Labs were significant for severe anemia with hemoglobin ranging between 7 to 8 g/dl despite blood transfusions. His baseline hemoglobin prior to surgery was 12 g/dl. At this time further hematologic workup was done which revealed LDH of 3576 and haptoglobin of less than 20 with some schistocytes on peripheral smear suggestive of hemolytic anemia. All other workup for Multiple Myeloma and Autoimmune hemolytic anemia was negative. Tran esophageal echocardiogram was done at this time which showed small paravalvular leak around the prosthetic mitral valve. Patient was transferred to University Hospital where he underwent mitral valve excision with debridement of surrounding tissue followed by mitral valve replacement with a 29mm Medtronic Hancock bioprosthetic valve. On one month follow up, his symptoms had markedly improved.

**DISCUSSION:** Paravalvular leak (PVL) refers to blood flowing through a channel between the structure of the implanted valve and cardiac tissue as a result of a lack of appropriate sealing. It is considered to be relatively rare complication of surgical mitral and aortic valve replacement. PVL can lead to Heart failure, hemolytic anemia and infective endocarditis. Hemolytic anemia secondary to paravalvular leak has been reported commonly with mechanical valves but is rare in bioprosthetic valves. Echocardiography is the primary modality for diagnosis of PVL in high risk patients. Transesophageal echocardiography (TEE) is usually more effective and important in diagnosis and characterization of PVL. Current treatment options for PVL include surgical or transcatheter deployment of occlude devices.

**CONCLUSION:** Patients presenting with anemia after prosthetic valve replacement should be evaluated for hemolysis and paravalvular leak should be considered as a differential diagnosis in such patients as early identification and surgical correction can decrease the morbidity and mortality associated with this condition.

**BACKGROUND/PURPOSE:** Results of serologic tests for autoantibodies, including tests for Antinuclear antibodies (ANAs) and antibodies to specific nuclear antigens such as double-stranded DNA (dsDNA), play an important role in the diagnosis of connective tissue disorders (CTDs) such as systemic lupus erythematosus (SLE). ANAs are often detected in many healthy individuals without CTDs (~13%). Although a negative ANA test makes SLE highly unlikely, the positive results without significant clinical and laboratory features will lead to inappropriate tests and misdiagnoses.

**METHODS:** This is a retrospective chart review of patients on whom ANA test has been performed at a 330-bed community hospital in U.S. over a period of one year. All relevant details like demographics, locations, physician service, clinical features, history of CTDs, prior ANA results, additional tests and their results were noted. The justification for ordering the ANA test was compared against
clinical and laboratory parameters included in the 2012 SLICC classification criteria for SLE. Subsequently, true and false positive incidence was calculated. For all the negative or positive ANA tests, special attention was given to the indications of testing based on chart analysis. The 2012 SLICC clinical classification criteria were applied retrospectively to all cases regardless the ANA results; more than two positive parameters by SLICC criteria were proposed as a justification to order ANA test. The results are compared using 2x2 chi-square test.

RESULTS: During one year period, ANA was ordered for a total of 465 patients (Male=151, Female=314). Among them, 12.47% (n=58) had prior history of CTDs and 0.98% (n=4) had prior ANA positivity. In the remaining 403 patients, ANA was found positive (titers ≥1:80) in 6.94% (n=28) and negative in 93.05% (n=375). By applying 2x2 chi-square test, was shown that ANA positivity or diagnosis of CTD is very unlikely if less than 2 SLICC parameters are present with a p value <0.05 (Table 1) Out of all 465 cases, only one new case of Anti phospholipid antibody syndrome was identified. A total of $39,297 was spent on ANA, and $87,165 on additional tests ordered in conjunction or following a positive ANA. It was noted that a large number of cases where ANA sub-serologies are ordered without knowing the ANA. This ordering pattern is against recommendation by "Choosing Wisely" campaign endorsed by ABIM and ACR.

CONCLUSION: Testing for ANA and related serology had cost approximately $126,000/yr for a medium size hospital and lead to no new SLE cases. ANA sub-serologies had cost the hospital $87,165 and according to "Choosing Wisely" campaign, were not indicated in more than 93% of cases, as these were negative ANA by IIF. In our hospital the lab was instructed to cancel the additional sub-serologies unless the ANA turns positive.

81. 271
Clinical Vignette
Abbassi, Sonja
Sushil K. Mehandru, MD, Shivendra Pandey, MD, Michael Carson, MD, Vikas Singh, MD
Jersey Shore Medical Center (Mayer Ezer)
The First Reported Case of Bartter’s Syndrome with Atypical Kaliopenic Nephropathy

OBJECTIVE: Recognize that Bartter’s syndrome can result in typical renal biopsy findings as well as kaliopenic nephropathy.

CASE: A 37 year old female with a chronic history of Bartter’s syndrome presented with severe persistent hypokalemia. Her current medications include spironolactone, aliskiren, and potassium chloride. Exam: 1+ peripheral edema. LABS: potassium 2.3 mEq/L, chloride 87 mEq/L, bicarbonate 38 mmol/L, rennin 13.6, aldosterone of 11.6, magnesium of 2.2 mg/dl, and GFR of 41 cc/min. Urinalysis was normal. Despite treatment with a potassium-sparing diuretic and potassium supplementation, the patient’s serum potassium level remained depressed. Because of progressive renal failure and refractory hypokalemia, a renal biopsy was obtained and not surprisingly was consistent with Bartter’s syndrome (juxtaglomerular hyperplasia), however it also showed findings of atypical kaliopenic nephropathy (interstitial fibrosis and tubular atrophy), and did not show the classic kaliopenic findings of tubular vacuolization and cysts.

DISCUSSION: Bartter’s Syndrome is a rare autosomal recessive disorder of the nephron that has the features of polydipsia, polyuria, decreased urine concentrating abilities, hypokalemia, hypochloremic metabolic alkalosis, hyperreninemia, hyperaldosteronism, normal to mildly reduced serum magnesium, and normal to low blood pressure. The most consistent pathological finding is juxtaglomerular hyperplasia. Kaliopenic nephropathy is often found in Bartter’s patients and involves tubular vacuolization/cystic changes. Atypical kaliopenic nephropathy will have interstitial fibrosis and tubular atrophy caused by chronic hypokalemia, as in this case. A pubmed search (terms: Bartter, kaliopenic, hypokalemic, nephropathy) did not return any citations associating Bartter’s with this pathological finding. This case illustrates the importance of anticipating other pathological findings that are heavily associated with a particular syndrome. In this case Bartter’s syndrome caused chronic hypokalemia that led to kaliopenic nephropathy and worsening of her renal function. When Bartter’s syndrome is suspected due to hypokalemia and renal failure, aggressive control of
82. 91 Clinical Vignette
Abu Homoud, Ahmad
Deborah R. Alpert MD, PhD
Jersey Shore Medical Center (Mayer Ezer)

Recognizing the pattern: Relapsing Polychondritis presenting with recurrent otitis externa and scleritis

LEARNING OBJECTIVES: 1. Recognize common signs and symptoms of relapsing polychondritis presenting over time. 2. Understand the diagnostic criteria for relapsing polychondritis.

CASE SUMMARY: A 37 year-old black female presented with recurrent left eye and ear swelling and redness over the past 1.5 years. She was hospitalized several times and treated with various antibiotics without significant improvement. She presented with one week of left eye and ear swelling, pain and redness, photophobia and blurry vision. Her exam revealed conjunctivitis and scleritis of the left eye, and left otitis externa. She was treated with intravenous ampicillin/sulbactam and dexamethasone with clinical improvement, then discharged home with oral antibiotics and a rapid steroid taper. She was readmitted several days later with symptom recurrence, subjective fevers, headache and progressive hoarseness. She denied arthralgias, sick contacts, sexually transmitted infections, contact lens use or swimming. Laboratory evaluations revealed ESR 59 mm/hr (0-20). Antinuclear antibody, rheumatoid factor, anti-neutrophil cytoplasmic antibody, Lyme, human immunodeficiency virus, hepatitis and syphilis serologies were all negative. Urinalysis was normal, and all cultures, including eye culture, were negative. CT scan of the facial bones revealed marked soft tissue swelling of the left external auditory canal. CT scan of the neck and chest demonstrated mild soft tissue thickening along the anterolateral trachea without stenosis, and clear lungs. She was diagnosed with relapsing polychondritis (RP) and treated with prednisone 60 mg daily, leading to clinical improvement. She was scheduled for close outpatient follow-up.

CONCLUSION: RP is a rare autoimmune disorder affecting various cartilaginous structures, mostly seen in males between the ages of 40-60. RP may begin with subtle inflammatory symptoms affecting the ears, nose, eyes, joints, and/or respiratory tract. The average delay to diagnosis is four years. If unrecognized, RP may lead to life-threatening complications including airway collapse or cardiac involvement. Our patient presented with recurrent otitis externa, scleritis, conjunctivitis and tracheitis, and was treated unsuccessfully with various courses of antibiotics over more than one year. The diagnosis of RP is based solely upon clinical manifestations. McAdam’s criteria require three or more clinical features including: (1) bilateral auricular chondritis, (2) nonerosive, seronegative inflammatory polyarthritis, (3) nasal chondritis, (4) ocular inflammation, (5) respiratory tract chondritis, or (6) cochlear or vestibular dysfunction. Modified McAdam’s criteria require one clinical manifestation with pathologic evidence of chondritis at separate anatomic locations, and response to steroids and/or dapsone. Although our patient presented with unilateral auricular chondritis, she manifested other typical features of RP over time that were steroid-responsive. This case reiterates the importance of pattern recognition in a patient manifesting an unusual constellation of symptoms over time. The clinical diagnosis of RP unified this patient’s recurrent otitis externa, eye inflammation and tracheitis, thus allowing for appropriate treatment.

83. 179 Clinical Vignette
Kassab, Ihab
Casey MD, Michael P. Carson MD
Jersey Shore Medical Center (Mayer Ezer)

First Reported Case of Group B Streptococcus Mediastinal Abscess in an Adult: Not Your Usual Chest Pain

LEARNING OBJECTIVES: 1) Diagnose primary mediastinal abscess presenting as chest pain in the absence of other head/neck infection 2) Recognize that a primary mediastinal abscess can be caused by Streptococcus agalactiae (Group B Strep: GBS).
CASE: A 43 year-old male with Type 2 DM and hypertension presented with a two week history of chest pain at rest and worse at exertion. One week prior he was diagnosed with costochondritis and sent home on NSAIDS. In Our ED he presented with 10/10 midsternal chest pain and tenderness over the left sternal border to left nipple area and down to the upper epigastrum. ROS: subjective fevers and night sweats for 2-3 days, and he shaves his chest weekly. EXAM: BP 189/84 mmHg, HR 117, Temperature 99.3oF, RR 16, room air pulse oximetry 97%. Right anterior chest wall was swollen, erythematous, tender to palpation, and indurated over the chest and upper abdomen. No rashes or skin lesions that could serve as a portal for infection. Normoactive bowel sounds, no hepatosplenomegaly, normal cardiac exam. LABS: glucose 359mg/dL, Albumin 1.9g/dL, Hemoglobin 12.4gm/dL, WBC 19.6K/uL, 86% neutrophils, troponin 0.02ng/mL. CT chest: “Enlargement of the pectoralis muscle bellies, inflammatory change within the fat deep to the pectoralis muscle bellies and adjacent to the anterior chest wall, soft tissue density deep to the chest wall within the anterior mediastinum.” Day #1: preliminary blood culture: Gram positive cocci in pairs and chains, vancomycin was started empirically and changed to ampicillin on Day #3 when the blood cultures grew GBS. CT guided mediastinal biopsy/culture also grew GBS. The abscess was explored and drained. After 6 weeks of IV ampicillin a follow-up CT scan showed complete resolution. 2D-echocardography did not show vegetations or vascular anomaly.

DISCUSSION: A pubmed.org search using the terms “mediastinal, abscess, anterior, group B, streptococcus” found that most mediastinal abscesses occur in relation to other head/neck infections (dental, epiglottitis, pharyngeal, cervical), trauma, medical instrumentation, or procedures. The reported organisms include Strept milleri, Group A and Group G. There was one report of GBS, but that was related to extension of infection from the sternocleidomastoid muscle. There are 14 reported cases of primary atraumatic mediastinal abscess in children, but we did not identify any reports of GBS causing primary mediastinal abscess in adults. In summary, this is, to our knowledge, the first reported case of atraumatic primary GBS mediastinal abscess in an adult. The presentation of this unusual problem can be subtle, so a high index of suspicion and consideration of the proper differential diagnosis is likely the best way to avoid delays in identification and treatment.

84. 81 Clinical Vignette Mamarabadi, Mansoureh Lumeng J. Yu BA, Alan Klein MD, Michael P. Carson MD Jersey Shore Medical Center (Mayer Ezer)

Red Herring: Ipsilateral Unilateral Papilledema in Meningioma

OBJECTIVES: 1)Recognize unusual presentations of meningioma 2)Review causes of unilateral papilledema

CASE: A 41 year old right-handed female was sent to the ER for a finding of right sided papilledema associated with 4 weeks of progressive hazy vision in the right eye. PMH: cranial trauma (2003), TAHBSO (2010). MED: Conjugated estrogens. ROS: denied any eye pain, seizures, anosmia, nausea/vomiting, headache, weakness, numbness, change in mental status, personality, mood, or color perception. EXAM: Visual acuity 20/20 OS, only able to count fingers at 20 cm OD. Right relative afferent pupillary defect. Funduscopy: right optic disk edema, left normal. No sensory deficits, pathologic reflexes, or long tract signs. MRI: 6.2x5.6x5.5 mass arising from right sphenoid wing, with dural tail, crossing midline, large mass effect, and intratumoral calcifications. Compression of optic nerve at level of orbital apex, but no orbit involvement. Dexamethasone was followed by craniotomy for removal. Pathology: atypical sphenoid wing meningioma, WHO grade II, with some degree of brain invasion. She did well postoperatively with gradual improvement in right visual acuity.

DISCUSSION: Meningiomas account for 15% of intracranial tumors. Risk factors include radiation exposure, head trauma, hereditary
predisposition, and hormonal factors. Population studies suggest that coexistence of trauma and meningioma is likely due to chance (PMID: 460549). However, many meningiomas express ER/PR, and estrogen therapy may be associated with development of meningiomas (PMID: 23800670). Papilledema is typically bilateral and secondary to globally increased intracranial pressure (ICP). The differential for the unusual finding of unilateral papilledema includes pre-existing unilateral optic disc atrophy, interruption of the peri-optic meningeal space by increased ICP or tumor, or Foster-Kennedy syndrome (ipsilateral vision loss with optic nerve atrophy due to compression of the optic nerve by a tumor on that side, rather than papilledema, but contralateral papilledema due to the tumor causing increased ICP). In this case, the papilledema and vision loss were on the same side. Case reports of unilateral disc swelling have been published with a range of etiologies, but a pubmed.org search revealed unilateral papilledema with meningioma is typically discussed with overt Foster-Kennedy syndrome. The isolated ipsilateral papilledema in a meningioma compressing the optic nerve could be secondary to stasis of axoplasmic flow causing prelaminar axon swelling, resulting in optic disc swelling and/or disruption of perioptic nerve subarachnoid space by mechanical pressure as well as ophthalmic vein return. It is possible, if the diagnosis were delayed, that our patient would have presented with Foster-Kennedy syndrome. It is important to note that unilateral papilledema has lead to the misdiagnosis of an isolated optic nerve lesion such as optic neuritis and multiple sclerosis, so when this unusual finding is detected, cranial imaging must be performed.

85. 62 Clinical Vignette Midha, Monica
Kenneth Nahum, DO; Ira Strauss, MD; Avais Masud, MD
Jersey Shore Medical Center
(Mayer Ezer)
Methotrexate Induced Nephrotoxicity
LEARNING OBJECTIVES: 1) Recognize parameters for early detection of nephrotoxicity in patients receiving methotrexate. 2) Understand how to prevent and treat methotrexate induced nephrotoxicity.

CASE SUMMARY: A 67 year old male with large B cell lymphoma of the CNS had neurosurgical resection of one mass, 22 radiation treatments, and was transferred to rehab. He returned one month later for chemotherapy with high dose methotrexate (MTX; 15,760mg). He received naproxen for pain control and ciprofloxacin for UTI. Urinary pH was monitored, the patient received bicarbonate to alkalinize the urine, and IV fluids were administered. Leucovorin treatments were started on hospital Day 3. Admission creatinine was 0.81mg/dL, which rose to 1.46mg/dL on Day 2. On day 3, creatinine was 3.15mg/dL and the patient had a change in mental status, appearing agitated and restless with no relief from anxiolytics. MTX levels were 397.20uMol/L on Day 1, 3.62uMol/L on Day 2, and 1.38uMol/L on Day 3 (goal < 1uMol/L at 48 hours). On exam, the patient was alert, awake and oriented but agitated and anxious. He was transferred to the intensive care unit for emergent high-flux hemodialysis to treat the altered mental status and acute renal failure, with a peak creatinine of 4.71mg/dL. Naproxen was also stopped and ciprofloxacin was changed to another antibiotic. In the ICU, the patient developed respiratory failure resulting in intubation. He received a week of hemodialysis and over two weeks of leucovorin, after which his renal function normalized and his mental status improved moderately; however, he was unable to be weaned off the ventilator, resulting in tracheostomy and PEG tube placement, after which he was transferred to a long term rehabilitation facility.

DISCUSSION: Though the subject has been discussed in literature, cases of methotrexate induced nephrotoxicity continue to occur. High dose MTX is used to treat CNS lymphoma, and typically leucovorin is administered 24-48 hours after MTX to rescue normally dividing cells, in conjunction with urine alkalization and IV hydration. To improve early detection of MTX induced nephrotoxicity, serum creatinine, urine output and urine pH should be monitored. Plasma MTX concentration should also be monitored, with values greater than 1uMol/L after 48 hours of administration being predictive of toxicity. Certain medications should be avoided to prevent
nephrotoxicity when MTX is administered including NSAIDs, phenytoin, amiodarone, ciprofloxacin and PPI’s. Hemodialysis has limited efficacy in treating nephrotoxicity because it removes the drug from the blood but not from tissues, and therefore cannot reverse organ damage. Glucarpidase is an enzyme that has been shown in limited studies to rapidly lower serum MTX concentrations, but is exceedingly expensive and difficult to obtain. Prevention is key for patients receiving high dose MTX as treatment may not always reverse renal damage.

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<th>Midha, Monica</th>
<th>Anuradha Sharma, MD; Marnie Rosenthal, DO, MPH, FACP</th>
<th>Jersey Shore Medical Center (Mayer Ezer)</th>
<th>Group G Streptococcal Myositis and Early Compartment Syndrome</th>
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<td>OBJECTIVES: 1. Recognize that streptococcal infections cause a wide spectrum of disease, with myositis being rare but carrying a high mortality rate. 2. Understand that a high index of suspicion is vital to early diagnosis and treatment in this highly fatal infection.</td>
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<td>CASE SUMMARY: A 57 year old male with myeloproliferative disorder (MPD) diagnosed 8 months prior to admission supported with platelet transfusions, presented with fevers and body aches for several days. He complained of myalgias, bilateral leg and calf pain, nausea, and decreased oral intake. He reported dental extraction two weeks before admission, and grandchildren with upper respiratory tract infections who had also recently traveled to Mexico; he denied insect bites or skin lesions. Temperature on admission was 104.1°F, blood pressure 96/64 and pulse 158. Laboratory studies revealed WBC count of 20.0K/uL (38% bands) and platelets of 23K/uL. CPK peaked at 11,607(iU)/L. Empiric sepsis treatment was initiated with imipenem, vancomycin and gentamicin. Exam showed swollen lower extremities with tenderness, decreased range of motion, and development of clear bullae. The patient complained of worsening calf tenderness, pain and swelling; emergent CT scan showed indistinct margins of calf musculature with no soft tissue gas. Compartment pressures were obtained and peaked at 29mmHg, borderline for compartment syndrome. In the setting of severe thrombocytopenia, medical management with serial compartment pressure measurements were performed. Antibiotics were narrowed to ampicillin and clindamycin when blood cultures grew Group G streptococci. Subsequently his condition improved, and repeat imaging as well as blood culture results confirmed a diagnosis of myositis with a resolving phlegmon, and improvement of early fasciitis and compartment syndrome.</td>
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<td>DISCUSSION: Most reported cases of streptococcal myositis are linked to Lancefield Groups A and B, with a rising incidence in reported cases of Lancefield Group G streptococcus (GGS) causing severe, life-threatening infections. A majority of these infections occur in patients with underlying immunocompromised states, such as our patient whose medical history includes MPD. Myositis, defined as degeneration and inflammation of muscle tissue without abscess formation, is distinct from necrotizing fasciitis, which is characterized by involvement of deep tissues and skin involvement. The presentation of this case is unique as there is no clear source for streptococcal infection. Predisposing factors for our patient’s development of this serious condition include immunosuppression secondary to malignancy, and the possibility of viral infection contracted from sick contacts. Streptococcal myositis is a rare infection that carries a high mortality rate if left untreated or if diagnosis is delayed. Our patient’s clinical condition improved with aggressive medical management and targeted antibiotics, including the addition of clindamycin for protein-mediated down-regulation of toxin synthesis. Having a high clinical suspicion for infectious myositis is critically important to prevent mortality, as early treatment is paramount to survival.</td>
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<th>Morgan, Mina</th>
<th>Dr. Michael P. Carson, Dr. Sushil Mehandru</th>
<th>Jersey Shore Medical</th>
<th>Diarrhea as Presenting Symptom of Minimal Change Disease</th>
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OBJECTIVES: 1) Recognize that diarrhea due to bowel wall edema can be a presenting symptom of nephrotic syndrome. 2) Recognize the differential diagnosis of bowel wall thickening on CT scan in a patient with nephritic syndrome.

CASE: 24 year old woman presented to the emergency department with 1 day of watery diarrhea associated with right upper and lower quadrant abdominal pain. ROS: 20 pound weight gain in one month. CT Abdomen/Pelvis with PO/IV contrast: small amount of ascites; circumferential wall thickening of the cecum and entire ascending colon. Her history and imaging findings led to a working diagnosis of right sided colitis, and she was discharged home on ciprofloxacin and metronidazole. She was called back two days later when 1/2 blood cultures grew gram negative bacilli. EXAM: BP: 130/64 Periorbital swelling, +1 sacral/+2 pretibial pitting edema. LABS: Albumin <1g/dL, total cholesterol 329mg/dL, triglycerides 158 mg/dL, LDL 221mg/dL, HDL of 52mg/dL, 24-hour urine protein 8.39 grams. C3 190 mg/dL(85-170), C4 30.2mg/dL(16-40), ANA negative, ASO Screening negative (153IU/mL). Urine protein 222.65mg/dL, urine protein 987mg/dL, Protein/Creatine Ratio 4.43. HIV not detected by enzyme immunoassay. Urinalysis >300mg/dL protein, WBC's 8-10/HPF, +nitrates. Normal kidneys on CT imaging. Antibiotics were continued for the bacteremia, the diarrhea improved during the hospitalization, possibly due to the use of low doses of a loop diuretic to palliate the symptoms of the diffuse peripheral edema. Renal biopsy revealed Minimal Change Disease. The patient is followed as an outpatient and was started on oral prednisone 60mg/day.

DISCUSSION: Nephrotic Syndrome is diagnosed in the presence of proteinuria greater than 3.5 grams per day, weight gain, edema, hypoalbuminemia and hyperlipidemia. Gastrointestinal symptoms are not commonly described and a pubmed.org search (terms: nephrotic, bowel, diarrhea, edema) only revealed a case of chylous ascites theoretically due to lymphatic impairment due to bowel wall edema, but no mention was made that bowel wall thickening was documented on imaging studies [PMID 17044480]. The failure of the diarrhea to improve until the admission where low-dose diuretics were given in addition to the antibiotics, and the presence of severe peripheral edema strongly suggest that the CT findings were edema and not the less likely simultaneous presence of concurrent inflammatory colitis in a young woman. It is likely that the bacteremia was transmigration across edematous bowel wall. To our knowledge, this is the first reported case of nephrotic syndrome presenting with diarrhea due to hypoalbuminemia related colon edema leading to decreased absorption of water. Clinicians should be aware that in the presence of nephrotic syndrome with anasarca, the differential of bowel thickening on CT scan includes edema as well as other more common conditions.
underwent video assisted thoracoscopic surgery (VATS) with wedge resection of lung and visceral pleura; frozen sections were negative for malignancy. Histological staining revealed large interstitial amyloid deposition especially around bronchioles, blood vessels, interlobular septa and pleura along with focal calcification and a foreign body giant cell reaction to amyloid deposits. These findings were consistent with severe diffuse alveolar septal amyloidosis. Multiple myeloma was diagnosed via an M spike and bone marrow biopsy with > 10% plasma cells.

DISCUSSION: Amyloidosis is characterized by extracellular deposition of amyloid fibrils. The main two types of amyloidosis are AL (primary) and AA (Secondary). AL, caused by a plasma cell dyscrasia, is due to deposition of protein derived from immunoglobulin light chain fragments. Calculated overall amyloidosis incidence is 15 cases per million person-years at most. This is on the basis that the AL type represents two thirds of all cases, and it’s incidence in the United States is 6-10 cases per million person-years (approximately 2,200 new cases of AL amyloidosis could occur annually in the USA). Pulmonary amyloidosis seems to be rarer, but the exact incidence is unknown. It is typically sub-classified into tracheobronchial, nodular parenchymal, diffuse parenchymal (interstitial/diffuse alveolar septal), and senile. We are reporting a case of diffuse alveolar septal pulmonary AL amyloidosis; which is by far the rarest subtype. A PubMed search using terms: “alveolar, septal, pulmonary, amyloidosis, multiple, myeloma” revealed only 24 reported cases of this condition of which only 5 cases of diffuse alveolar septal pulmonary amyloidosis were associated with multiple myeloma. Treatment for such cases is mainly directed towards the underlying plasma cell dyscrasia. Our patient is currently receiving Cyclophosphamide, Bortezomib and Dexamethasone (CyBorD).

89. 77 Clinical Vignette Agrawal, Abhinav Dennis Lourdusamy, Neil Holland Monmouth Medical Center (Margaret Eng)

‘Neuro-myelopathy Optica’: NMO spectrum disorder presenting as Brown-Sequard syndrome

INTRODUCTION: Brown Sequard syndrome (BSS) presents with features of ipsilateral motor function loss and contralateral loss of pain and temperature. Patients presenting with features of either myelitis or recurrent/bilateral optic neuritis with positive NMO/anti-aquaporin-4 (AQP4) antibodies they are classified under Neuromyelitis Optica Spectrum Disorders (NMOSD). Rare cases of non-traumatic BSS caused by spontaneous epidural hematomas or transverse myelitis have been described in literature. Here, we are presenting a rare case of NMOSD presenting as Brown Sequard syndrome.

CASE DESCRIPTION: A 70-year-old female presented the emergency room with a 2 day history of weakness in her left leg. On examination, she had 3/5 strength in the left leg and normal strength in the right leg. Temperature and pain sensitivity was markedly diminished in the right leg up to a truncal sensory level at the umbilicus. The patellar and ankle reflexes were brisk on the left side. Proprioception was diminished in the left leg. Magnetic resonance imaging (MR) of the brain was normal. MRI of the spine showed T2 hyperintense signal seen within the upper thoracic cord centered at T2-T5 level with mild expansion of the cord. Spinal fluid analysis was normal – no pleocytosis and normal IgG index with no oligoclonal bands. Computed tomography (CT) imaging of the chest was negative for aortic dissection and trans-esophageal echocardiography was negative for embolic sources. Serum NMO (AQP4) antibody titers were found to be significantly positive with levels of 160 U/ml, suggesting a diagnosis NMO spectrum disorder. She was treated with high dose intravenous methyl prednisone, and her symptoms showed some improvement after a few days of therapy. She was ultimately transferred to an acute rehabilitation facility.

DISCUSSION: Neuromyelitis Optica (NMO) is an inflammatory/autoimmune disorder of the central nervous system usually
characterized by the features of myelitis and optic neuritis. Patients who are seropositive for NMO antibodies, but do not present with the symptoms of classical NMO are classified NMOSD. These patients can present with involvement of either spinal cord alone, recurrent attacks of optic neuritis alone, and even non-optico spinal manifestations like brain stem lesions with intractable hicups and nausea/vomiting. Our patient can be classified under NMOSD, because of positive NMO antibodies with a spinal cord lesion affecting three contiguous vertebrae. Our patient had a unique presentation with unilateral involvement of spinal cord, presenting as BSS. The absence of pleocytosis during the attack in our patient is another interesting. Knowledge about the clinical presentation of NMO and NMO Spectrum disorder continues to evolve. The case teaches us that a diagnosis of NMOSD should be considered even in atypical presentations like in our patient presenting with Brown- Sequard syndrome in the absence of CSF pleocytosis.

Clinical Vignette

Amor, Martin Miguel I
Abhinav Agrawal MD, Deepa Iyer MD, Marc Cohen MD
Monmouth Medical Center (Margaret Eng)

AORTICO-LEFT ATRIAL FISTULA: A RARE COMPLICATION OF INFECTIVE ENDOCARDITIS

INTRODUCTION: Paravalvular aortic root abscess with intracardiac fistula formation is an exceedingly rare complication of infective endocarditis. This complication places patients at increased risk for congestive heart failure, heart block and death.

CASE DESCRIPTION: A 68-year-old Bosnian female with prior aortic valve replacement with a bioprosthetic valve for aortic regurgitation was admitted for worsening shortness of breath, fever and lethargy. She was recently on a vacation in Bosnia, where she fell ill and was hospitalized for 1 month for sepsis and renal failure. She had an extensive past medical history, pertinent for coronary artery disease, s/p PCI and stenting, diastolic congestive heart failure, atrial fibrillation, chronic kidney disease, systemic hypertension, multiple prior cerebrovascular accidents and chronic UTI. In the ED, she became markedly hypotensive and hypoxic. She was intubated and started on dopamine infusion. A bedside transthoracic echocardiogram revealed a paravalvular leak around the bioprosthetic valve, raising concern for an aortic root abscess. EKG revealed atrial fibrillation with low voltage QRS, without evidence of bundle branch blocks or conduction delays. The patient developed septic shock and was started on broad spectrum antibiotic therapy and pressor support. A transesophageal echocardiogram revealed an extensive aortic root abscess. The abscess had ruptured into the left atrium, with a fistula connecting the aortic root to the left atrial cavity. The abscess was located around the bioprosthetic aortic valve which had a large vegetation and severe paravalvular aortic regurgitation. She became hemodynamically unstable during the procedure and was brought to the operating room for emergent surgery. She underwent homograft aortic root replacement, VSD repair and ligation of the aortico-left atrial fistula. Two sets of blood cultures grew Enterococcus faecalis. Postoperatively, she developed worsening septic shock, requiring multiple pressors, disseminated intravascular coagulation, and anuria, requiring CVVHD. She eventually expired 5 days later.

DISCUSSION: Intracardiac fistula formation is a rare and particularly problematic complication of periannular spread of infective endocarditis, with high mortality despite adequate therapy. In prosthetic valves, this process usually begins on the prosthetic cuff, and often extends outside the valvular apparatus, resulting in valvular dehiscence, abscess formation, and myocardial involvement. Operative treatment remains the cornerstone of management. Surgical treatment involves removal of all infected tissue including annular elements, followed by reconstruction of the annulus for safe anchoring of a valve conduit. Early rather than delayed surgical intervention has been shown to improve survival. In our patient, a delay in the diagnosis, which in turn led to a delay in surgical intervention, contributed to the poor outcome. This case illustrates that a high index of suspicion, prompt diagnosis by echocardiography, and early rather than delayed surgical intervention, are crucial to improving treatment outcomes for this rare condition.
RECURRENT SINUS PAUSES: AN ATYPICAL PRESENTATION OF TEMPORAL LOBE EPILEPSY

INTRODUCTION: Epileptiform disorders are known to alter autonomic function at both sympathetic and parasympathetic levels. These effects may give rise to a broad spectrum of cardiovascular abnormalities. While ictal tachycardia is more common, ictal bradycardia and conduction delays may also be encountered.

CASE DESCRIPTION: A 58-year-old Haitian female with known history of hypertension was admitted for severe bilateral lower extremity weakness. CT scan of the thoracolumbar spine revealed severe kyphosis of T10-T11 secondary to anterior collapse of the T11 vertebral body. She received intravenous steroids and underwent T11 corpectomy, fusion of T10-T12, implantation of biomechanical device at T11, anterior instrumentation of T10-12 and posterolateral fusion of T8-L3. Bone biopsy revealed evidence of osteomyelitis. She was started on a 42-day course of antibiotics. After surgery, she was noted to be increasingly lethargic and confused. Rapid response was called when she developed a complex partial seizure with secondary generalization. The seizure was terminated upon administration of intravenous antiepileptic medications. EKG monitoring during the seizure episode revealed sinus bradycardia, which eventually progressed to a 10-second sinus pause, approximately 20 seconds after seizure onset. She had 2 more similar seizure episodes during the same day. In each seizure episode, she would develop sinus bradycardia, followed by sinus pauses a few seconds after seizure onset. Interictal EKGs revealed normal sinus rhythm. EEG revealed periodic lateralized epileptiform discharges and a single seizure emanating from the right posterior temporal region. The seizure observed during the EEG focally originated from the T6 area, and then had secondary generalization. It lasted around 75 seconds and clinically manifested as blank staring. MRI revealed a large area of gyrical edema, sulcal effacement and cortically based diffusion restriction involving the right occipital lobe, right posterior temporal and parietal lobes. Lumbar tap revealed normal findings. She did not have any further seizure episodes. Thereafter, she underwent DDD pacemaker insertion and did not develop any more pauses. Repeat EEG revealed no lateralizing or epileptiform discharges. She was discharged to an acute rehabilitation facility after 26 days of hospital stay.

DISCUSSION: This case illustrates that epileptiform activity from the amygdala, anterior cingular cortex and insula of the temporal lobe can produce cardiac rate and rhythm abnormalities. Ictal tachycardia is more common, and can precede EEG seizure onset by approximately 13 seconds. Ictal bradycardia, on the other hand, occurs in fewer than 2% of seizures, is usually of temporal lobe origin, and may progress to cardiac asystole when left untreated. While increases in heart rate are more common, ictal bradycardia should also be carefully detected and addressed, to prevent progression into cardiac asystole. Early suspicion and recognition of these events may aid the clinician in averting sudden, unexplained death in epilepsy.

Exfoliative Erythroderma as a Paraneoplastic presentation of Adenocarcinoma of the Gall bladder

Gall bladder carcinoma (GBC) is a rare GI malignancy. Worldwide, there is a prominent geographic variability in GBC incidence that correlates with the prevalence of cholelithiasis. High rates of GBC are seen in South American countries as well as some areas of India, Pakistan, Japan and Korea. We are presenting a case of gall bladder adenocarcinoma presenting with a paraneoplastic syndrome in the form of exfoliative erythroderma. This is a very rare presentation of gall bladder malignancy, which by itself is an uncommon malignancy. North America is considered a low incidence area. We report a case of a 71 year old Caucasian male
presenting with an exfoliative skin rash all over his body. The patient was also found to have a huge gall bladder mass extending into the liver that turned out to be adenocarcinoma of the gall bladder on biopsy. Gall bladder cancer usually presents with abdominal pain or jaundice. We report only the second case in literature of a gall bladder adenocarcinoma presenting with exfoliative erythroderma as its paraneoplastic presentation. The patient as an outpatient also went to a dermatologist who prescribed him a course of oral prednisone without any improvement. The patient thus decided to come to the emergency room for further management. Interestingly, the patient had presented to the hospital 6 months ago after an accident. At that time, he had an evaluation in the emergency department where a CAT scan of chest and abdomen was performed which showed fracture in different places that were treated appropriately. Coincidentally at that time a liver mass was also seen on the CT scan in the right hepatic lobe abutting the gall bladder. High attenuation with nodularity was seen in the gallbladder fundus. The patient was instructed to follow up for that mass at that time but was lost to follow up. The patient also noted weight loss. No abdominal pain or obstructive jaundice were noted apart from the rash and itching. When the patient had a second CT scan on the second time, it showed enlargement of the previously seen mass. Patient was referred to surgery service and gall bladder mass was removed with dramatic resolution of the skin rash after 2 weeks from the surgery. The pathology of the removed mass turned back to be adenocarcinoma of the gall bladder. Skin rash can provide the first clues to a diagnosis in 1% of internal malignancies. Erythroderma secondary to malignancies in general has been reported. A study was performed on 135 cases with erythroderma found that about one fifth of the patients had lymphomas and mycosis fungoides. Exfoliative erythroderma is rare secondary to GI malignancy. Keywords: Exfoliative erythroderma, Gall bladder carcinoma, Paraneoplastic syndrome, Skin rash.

93. 199 Clinical Vignette Eltawansy, Sherif Shil Patel MD, Mana Rao MD, Samaa Hassanien MD, Mihir Maniar MD Monmouth Medical Center (Margaret Eng) Acute renal infarction presenting with acute abdominal pain secondary to newly discovered atrial fibrillation

BACKGROUND: Acute renal infarction is one of the rare causes of acute abdominal pain and it can be easily missed as it is unexpected. It has to be expected in the patients with risk factors. Most accurate diagnostic tool is the helical CT scan of abdomen. Preferred therapy lies between percutaneous endovascular therapy, anticoagulation and thrombolysis. Missing the diagnosis can compromise the case and may affect the kidney function and the overall outcome. Our case was a challenging one given that she presented with acute abdominal pain with no previous history of kidney disease and in the same time, accidentally discovered atrial fibrillation. We report an 85 year old female with known history of recurrent diverticulitis that required multiple admissions for repeated acute diverticulitis episodes. Patient this time came with abdominal pain and was started on the routine treatment for diverticulitis as usual. The CT scan of abdomen showed renal infarcts bilaterally that were confirmed by injecting a contrast. The atrial fibrillation was discovered accidentally while doing EKG. Patient was started on anti coagulation and discharged. Renal infarction is rare. In a study of 14,411 autopsies published in 1940, the incidence of renal infarction was 1.4 percent. Acute renal infarction diagnosis can be missed. The entity is often misdiagnosed. Unilateral flank pain in a patient with an increased risk for thromboembolism should raise the suspicion of renal infarction. In such a setting, hematuria, leucocytosis and an elevated lactate dehydrogenase level are strongly supportive of the diagnosis. Obviously, Delay in diagnosis is much too long and points to a lack of physician awareness regarding the entity. This also applies to the radiologist. Since unenhanced CT is now used almost routinely in the investigation of acute flank pain, it is imperative to remember that contrast enhancement is essential for the diagnosis of acute renal infarction

CONCLUSION: Renal infarction is one of the rare causes of acute abdominal pain that has to be suspected and managed appropriately specially in patients with risk factors and it has to be on the differential diagnosis of the admitting physician.
**LEARNING OBJECTIVE:** Our case represents an acute renal infarction presenting only with abdominal pain that was thought to could be one of her recurrent diverticulitis episodes that turned out to be bilateral multiple renal infarctions. In the same time, atrial fibrillation was discovered accidentally although it is known to be one of the most common causes of renal infarction. Although the atrial fibrillation was discovered, it was not one of her problem list before the recent presentation. Renal infarction should be put on the differential diagnosis list of the admitting physician and the radiologist. Key words: Renal infarction-atrial fibrillation-abdominal pain.

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<th>Eltawansy, Sherif Chandni Merchant MD, Paavani Atluri MD, Sukrut Dwivedi D.O</th>
<th>Monmouth Medical Center (Margaret Eng)</th>
<th>Gas forming pyogenic liver abscess secondary to Clostridium Perfringens ending in multi-organ failure following a self-limited episode of acute gastroenteritis</th>
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<td><strong>BACKGROUND:</strong> Pyogenic liver abscess is a critical infection that can lead to ICU admission. Identifying the organism causing that liver abscess is critical for diagnosis and management. Clostridium perfringens is unusual to cause gas forming pyogenic liver abscess. Clostridium perfringens septicemia has a poor prognosis and high mortality rate. We report a case of an 81 year old female who developed a pyogenic liver abscess with gas formation that occurred within a short duration spontaneously after a self limited acute gastroenteritis with an interval period of 5 days. She used to live with her husband and an aide. She was brought to the emergency department with vomiting and diarrhea for few days associated with lethargy and confusion. Jaundice was also noticed. Patient was aphasic secondary to the previous CVA. Medication list included aspirin, clopidogril, paroxetine, clonazepam, Olanzapine, esoterodine, atorvastatin, levotryoxine and lisinopril in addition to lactulose and senna for constipation as needed. She was on Leviteracetam for history of seizures. Her surgical history included cholecystectomy 10 years ago and thyroidectomy and this was long time ago also patient stayed for 3 days in a regular floor and transaminitis with elevated bilirubin were noticed but overall condition improved on intravenous fluids with no antibiotic used. EGD was done and unremarkable. She was sent home on the same previous medications. 5 days only later, She came back with toxic appearance and was vitally unstable in the ED with a picture of septic shock. CT abdomen showed a big right hepatic abscess with gas formation. CT guided drainage of the abscess was done and culture showed C.perfringens that was found in the blood culture also patient was put on maximum vasopressers and wide spectrum antibiotics and was kept on the mechanical ventilator. Case did not respond treatment and died of septic shock. Patient suffered from multi-organ failure including respiratory, renal and hepatic failure in addition to the persistent shock status and coma. Most common organisms to cause pyogenic liver abscess are klebsiella pneumonia, Escherichia coli and gram positive cocci like Enterococcus and staph aureus. The most common precipitating factors are invasive procedures like endoscopy of the biliary tract or any invasive treatment like trans-arterial hepatic embolization to treat hepatocellular carcinoma.Chemotherapy or diabetes mellitus can precipitate to liver abscess through immuno-suppression. It is uncommon to isolate C.Perfringens from a liver abscess. C. perfringens is normal inhabitant of human intestine and a common cause of food poisoning. It causes tissue necrosis and gas gangrene. Few cases were reported to have a liver abscess secondary to C. perfringens; for example following a laparoscopic cholecystectomy, pancreatectomy or spontaneously. Key words: pyogenic liver abscess- clostridium perfringens- gas gangrene.</td>
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<th>Monmouth Medical Center (Margaret Eng)</th>
<th>Syndrome of inappropriate anti-diuretic hormone secondary to Non-Cirrhotic Primary Hepatocellular Carcinoma</th>
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<td>**The syndrome of inappropriate antidiuretic hormone secretion (SIADH) is a disorder of impaired water excretion caused by the inability to suppress the secretion of antidiuretic hormone (ADH). It is usually seen in pulmonary malignancies, central nervous</td>
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system disorders, and secondary to medications. SIADH has very rarely been encountered in primary hepatocellular carcinoma. Two cases were reported in Japan and one case in Spain after extensive investigation of the medical records. We report a case of a 71-year-old Guatemalan man who presented with confusion, cachexia, and abdominal symptoms in the form of vomiting and abdominal discomfort. He did not have any significant past medical history although he has not had any regular medical assessment and he was not on any medication. On the initial work-up, SIADH diagnosis was made in the setting of hypotonic euvolemic hyponatremia with high urine osmolality. Patient was found to have normal thyroid hormone levels and had normal cortisol levels, as well. He had poor response to normal saline infusion but responded adequately to water restriction, hypertonic saline, and eventually to the ADH receptor antagonist Tolvaptan with normalization of the sodium levels in three days. After an extensive work-up that included an abdominal CT scan and ultrasound-guided liver biopsy, the reason for SIADH turned out to be a newly diagnosed primary hepatocellular carcinoma. The precipitating factor for the cancer was not identified by history or by work-up. No metastasis was identified. Liver functions were preserved but patient was severely malnourished. SIADH can occur as a para-malignant feature of certain malignancies. In primary hepatocellular carcinoma, the most common para-neoplastic syndromes (PNS) associated with the disease includes hypercholesterolemia, hypercalcemia, hypoglycemia, and erythrocytosis. In our case, SIADH was the main PNS related to the primary hepatocellular carcinoma, which is a malignancy very rare to cause SIADH.

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### ACUTE PANCREATITIS WITH NORMAL LIPASE AND AMYLASE: A CASE SERIES

**INTRODUCTION:** Lipase and/or amylase elevation greater than 3 times upper limit of normal (ULN) is one of the three diagnostic criteria for the diagnosis of AP as per the American College of Gastroenterology practice guidelines. Both lipase and amylase are extremely sensitive for the diagnosis of AP with sensitivity in the range of 85-100% for lipase and 81-95% for amylase. AP with either lipase or amylase being normal is a rare entity but has been reported before. AP with both lipase and amylase normal is an extremely rare event with only isolated case reports in literature. We report a series of 9 cases with imaging proven AP who presented with normal lipase and amylase.

**CASE DESCRIPTION AND DISCUSSION:** All the 9 cases were from our institution, admitted with abdominal pain consistent with AP. All the cases had CT abdomen findings consistent with AP. None of the patients had imaging or clinical features of chronic pancreatitis and one patient had recurrent pancreatitis with 3 episodes in the past, the last episode being several months ago. At the time of admission lipase and amylase levels were normal in all the 9 patients. The enzyme levels were repeated within 24 hours in 8 out of the 9 cases and were found to be normal. In 2 cases, the normal enzyme levels could be explained by late presentation, however in majority of the cases, the reason for lack of enzyme elevation was not clear. The mean (±SD) time from the onset of symptoms to the presentation in the Emergency department was 69.33 hours (±1.4). In 6 out of the 9 cases, the etiology of AP was unclear and 1 case was due to gall stones. All the cases were mild as per Modified Atlanta Classification and Determinant based classification.

**CONCLUSIONS:** Imaging confirmed AP can occur in patient in the absence of pancreatic enzyme elevation. Specific scenarios where enzyme levels are more likely to be normal include: a. Severe necrotizing pancreatitis b. Very early or late in the course of the disease. In any patient having risk factors for AP, with abdominal pain consistent with AP, the diagnosis of AP should be considered even if the pancreatic enzyme levels are normal at the time of presentation.
| 97. | 87 | Clinical Vignette | Merchant, Chandni | Mana Rao | Monmouth Medical Center (Margaret Eng) | **A Case of Erythromelalgia treated with Gabapentin and Propranolol**

Erythromelalgia is a neurovascular disorder characterized by a triad of intense burning pain, redness and warmth of the skin; it frequently affects the lower extremities but may involve other areas such as the hands or face. Primary erythromelalgia is believed to be idiopathic and may have an autosomal dominant inheritance. Erythromelalgia can also be secondary to autoimmune disorders, diabetes mellitus (DM), hypertension, essential thrombocytosis, myeloproliferative disorders and venous insufficiency; or may occur as a paraneoplastic presentation of an underlying malignancy. This is the case of a 34 year old Asian Indian female with a medical history of type 1 DM, end stage renal disease on hemodialysis, hypertension and chronic hepatitis C. She presented to the office with an 18 month history of redness and burning pain affecting her feet. She was noted to have symmetrical blanching erythema over the planter aspects of her feet and insteps, which was accompanied by warmth. The rest of her physical exam including all vital signs were within normal limits. A detailed work up revealed normocytic normochromic anemia (hemoglobin 8.4 g/dl), a normal total leukocyte count with differential (6000 cells/mcL), normal platelet count (2,00,000/mcL), elevated blood sugar at 146, elevated creatinine (although at baseline - 2.8), ALT and AST were elevated at 56 and 58 respectively. ANA was weakly positive (1.1). ESR and CRP were reportedly normal. Serum cryoglobulin and complement levels were within the reference ranges as were anti neutrophilic cytoplasmic antibodies excluding cryoglobulinemia and small vessel vasculitis. A skin biopsy from the right instep showed a perivascular infiltrate. The patient was started on gabapentin 300mg/day and was advised cool compresses with leg elevation; these alleviated her pain but the redness and warmth persisted. She experienced a recurrence of the same symptoms 8 months later when the dose of gabapentin was increased to 900mg/day. Propranolol 10mg/day was added to the treatment regimen. The patient subsequently reported complete resolution of symptoms and was noted to be symptom free at 6, 12, 18 and 24 months of outpatient follow up. With this vignette, we showcase the usual presentation of secondary erythromelalgia, an infrequently reported diagnosis in the internal medicine outpatient setting. The challenge in defining a pathophysiological process persists, one of the chief reasons for this being lack of specificity among biopsy findings of previously reported cases. Thus management is targeted at symptom alleviation with the many physical, pharmacological and surgical interventions tried to date. Controlled studies remain to be performed while an answer to curative therapy via hard evidence remains obscure.

| 98. | 264 | Clinical Vignette | Parikh, Manan | Martin Amor MD, Madhu Paladugu MD | Monmouth Medical Center (Margaret Eng) | **Small bowel obstruction masquerading as Acute ST elevation myocardial infarction: A catastrophe avoided**

**INTRODUCTION:** Acute myocardial infarction is known to frequently present with atypical symptoms. On rare occasions, abdominal or thoracic conditions present with EKG findings consistent with acute myocardial infarction, raising suspicion of atypical presentation of a life threatening condition.

**CASE DESCRIPTION:** A 86-year-old male presented to the ER with complaint of vomiting for 24 hours and epigastric pain upon presentation. Patient had known medical history of Atrial Fibrillation, Hypertension, Chronic kidney disease, Barrett’s esophagus, esophageal rupture and pneumo-mediastinum requiring surgical repair. Patient’s initial evaluation showed elevated amylase and lipase and gallstones with dilatation of common bile duct on ultrasound. An EKG showed ST segment elevation in Inferior leads with reciprocal ST segment depressions in anterior chest leads consistent with acute inferior wall ST segment Elevation myocardial infarction. Patient was taken for an emergency cardiac catheterization. Patient was found to have 70 % stenosis of right coronary artery as well as diffuse calcific disease of posterior lateral branch with 40% left anterior descending artery stenosis. None of the
vessels showed rupture of plaque or an acute thrombus. Serials troponins remained normal. Intervention for non-critical lesion of RCA was deferred in light of impending gastrointestinal catastrophe. A CT scan of the abdomen showed small bowel obstruction with marked distention of stomach and a transition point in jejunum. Small bowel obstruction and gastric distention was conservatively treated with nasogastric tube placement over next 2 days. As patient’s gastric distention improved, his EKG findings of ST elevations in inferior leads also resolved. Patient further had aspiration pneumonia treated with intravenous piperacillin-tazobactam. He underwent a laparoscopic cholecystectomy for gallstones pancreatitis and was discharged with a stable follow up.

**DISCUSSION:** A variety of illnesses are known to mimic myocardial infarction including acute pericarditis, myocarditis, aortic dissection or acute pancreatitis. EKG changes consistent with ST elevation suggest trans-mural ischemia caused by acute thrombus formation or plaque rupture. However, structural abnormalities or altered configuration of thoracic or abdominal organs may cause variations on a surface EKG mimicking ST elevation myocardial infarction. A possible for ST elevation in this care may be that massive distention of stomach caused obstruction of the previously diseased right coronary artery without any thrombus formation giving the mentioned EKG changes. This is further supported by the complete resolution of ST changes after improvement in gastric distention. In summary, this a case of a 86 year old male presenting with EKG changes suggestive of ST elevation Myocardial Infarction due to small bowel obstruction in absence of acute coronary thrombus formation. It illustrates a rare presentation of a gastrointestinal condition mimicking a impending cardiovascular catastrophe.

**INTRODUCTION:** Temporo-parietal headache as the lone presenting symptom of massive aortic dissection

**CASE DESCRIPTION:** A 71-year-old Caucasian male presented to emergency room with severe temporo-parietal headache. Patient had known medical history of well controlled hypertension, dyslipidemia, coronary artery disease and Choroideremia. CT scan of head showed no intracranial hemorrhage or territorial infarct. Patient was bradycardic with heart rate ranging from 36 to 40. Blood pressure in left and right upper extremities was 128/53 mm Hg and 131/61 mm Hg respectively. Later in the emergency room he also complained of chest discomfort described as pressure on the left side of the sternum without radiation. His pain worsened with deep inspiration and in supine position and improved on sitting up. A chest radiograph showed mild enlargement of cardimediastinal silhouette and mild cardiomegaly. Patient’s EKG showed sinus bradycardia with non specific ST - T changes in anterolateral leads. He was immediately taken for a CT scan of chest to exclude pulmonary embolism. He was found to have thoracic aortic dissection involving ascending and descending aorta with intramural hematoma extending above the level of superior mesenteric artery consistent with Stanford type A and Debakey type 1 dissection. The patient was taken to the operating room and underwent an ascending aortic replacement, hemi-arch replacement as well as a suspension of the aortic valve. Post-operatively the patient had episodes of atrial fibrillation. He recovered within 9 days and was discharged with a stable follow up.

**DISCUSSION:** Aortic dissection is a life-threatening condition typically presenting with acute onset severe, sharp chest or abdominal pain. More than 90% of cases show presence of one or more of the following findings: 1) Abrupt onset sharp chest or abdominal pain 2) chest radiograph suggestive of aortic or mediastinal widening 3) A variation in pulse or blood pressure. Though neurological
symptoms of stroke have commonly been described as a presenting symptom, headache is an uncommon presentation. This case represented an unusual presentation of aortic dissection with headache being the only presenting complaint. Furthermore, the patient’s quality of chest pain as well as unremarkable chest radiograph and physical examination demonstrates the rare instances where subtle clues like bradycardia and atypical chest pain represent possibility of a grave medical catastrophe. In summary we are presenting a case of 71 year old male presenting with severe tempo-parietal headache found to have Stanford type A aortic dissection. This case illustrates how aortic dissection can present with subtle atypical manifestations and high index of suspicion is required to avoid any delay in diagnosis of such cases.

Exchange Transfusion in Sickle Cell Intrahepatic Cholestasis

Hepatic dysfunction in sickle cell disease may be caused by various clinicopathologic entities such as acute hepatic sequestration, sickle cell intrahepatic cholestasis (SCIC), benign hyperbilirubinemia, gallstones, iron overload and viral hepatitis. SCIC happens to carry a high case fatality rate. A 31 year old African American male known to have sickle cell anemia (homozygous - HbSS) presented to the emergency room with bilateral lower extremity and back pain. His past history included recurrent admissions for acute painful crises, multiple blood transfusions leading to iron overload and cholecystectomy. On exam, he was noted to have scleral icterus with all vital signs being within normal limits; a left sided chest wall port was noted to be intact. His laboratory studies revealed the following values: Hemoglobin 6.2, reticulocyte count 16, total bilirubin 4.0 (direct component 1.8), ALT 78, AST 132 and alkaline phosphatase 274. With a presumptive diagnosis of acute painful vaso-occlusive crisis the patient was admitted to the medical floor and prescribed intravenous fluids, hydromorphone for pain relief and one unit of packed red cells. Over the following 48 hours, he developed severe right upper quadrant abdominal pain and his bilirubin rose to 50 mg/dl with direct component measured at 37mg/dl. He was also noted to have renal failure at this time with serum creatinine measuring 3.5 mg/dl. A flat plate and ultrasound of his abdomen were unrevealing and an MRCP done only showed diffusely decreased signal intensity throughout the liver (suggestive of hemosiderosis) and periporal edema. Viral serologies excluded acute viral hepatitis. With the differential of SCIC in mind, the patient was placed on exchange transfusions which dramatically led to an improvement in his overall clinical status and serum bilirubin downtrended to 10mg/dl during his ensuing hospital course. An improvement in his renal function was also recorded. This case brings to our attention the infrequently encountered complication of Sickle Cell Disease, namely SCIC. The pathophysiology is believed to be sickling of the red cells within the hepatic sinusoids which leads to congestion of the vascular bed; this is followed by ballooning and necrosis of the surrounding hepatocytes thereby causing conjugated hyperbilirubinemia. Accompanying renal failure prohibits excretion of bilirubin, compounding the existing problem. The management includes RBC exchange, correction of coagulopathy and encephalopathy if any; and in warranted cases, Liver transplantation. Maintaining an index of suspicion, timely diagnosis and institution of therapy are paramount given the mortality attributed to SCIC in previously reported literature.

BiPAP with AVAPS in a Super Obese Pickwickian with Ogilvie’s Syndrome

The choice between non invasive and invasive ventilation is based on the cumulative of patient history, clinical status, arterial blood gas chemistries and physician judgement. A 41 year old obese male with pickwickian syndrome, chronic abdominal pain from a prior pannus, opiate dependence and diabetes was encountered in the emergency room in an obtunded state. His Glasgow coma scale
was calculated to be 10 and vital signs were normal except for oxygen saturation, noted to be fluctuant between 84 and 89% on a nonrebreather mask. Physical examination revealed a morbidly obese male (body mass index 63.27) whose neurological exam was limited given his presentation. Arterial blood gas (ABG) analysis revealed primary respiratory acidosis with metabolic compensation. As naloxone failed to reverse his altered mentation, he was placed on mechanical ventilation due to persistent obtundation with worsening ABGs to pH 7.03, pCO2 >125, pO2 60 and HCO3 35. A nasogastric tube yielded 2 litres of bilious output, adding ileus to the existing scenario. The patient extubated himself the following day and was subsequently obtunded again necessitating noninvasive ventilation by Bilevel Positive Airway pressure (BiPAP) with average volume assured pressure support (AVAPS). In a few hours, the patient was noted to be alert and was able to hold meaningful conversation. ABGs were noted to have corrected to pH 7.25, pCO2 71, pO2 100 and HCO3 36, on 35.00 FiO2. An insightful history revealed the administration of 16 mg of oral hydromorphone for abdominal pain which perhaps led to these events. AVAPS is a BiPAP mode that utilizes and ensures the delivery of a preset tidal volume as a result of an algorithm that automatically adjusts inspiratory positive airway pressure as per the patient's ventilatory and positional needs. It has been studied in subgroups of patients with chronic obstructive pulmonary disease (COPD), chronic respiratory insufficiency (CRI), obesity hypoventilation syndrome (OHS) and is reported to improve minute ventilation in CRI and ventilation quality during sleep thus efficiently decreasing pCO2 in OHS. A recent study reported the superiority of BiPAP/AVAPS in time to recovery from hypercapnic encephalopathy in COPD patients(1). While the final common pathway of carbon dioxide retention leading to hypercapnic respiratory failure and thence encephalopathy remains the same, the use of AVAPS during recovery from altered mentation in those with hypercapnic encephalopathy from opiate overuse and obesity hypoventilation remains to be reported.

compared to the standard FDA approved guidelines for quantities of caffeine. The stimulant drug that our patient used contained a number of other constituents along with caffeine as mentioned above with potentially harmful side effects. It is unclear how these components may have interacted or the arrhythmia is the sole effect of consuming high quantity of caffeine. Irrespective of which, consumption of such over the counter stimulant drugs containing caffeine and other components have shown to have increased risk in cardiovascular complications. Treatment of severe caffeine intoxication is generally supportive, but if the patient has very high serum levels of caffeine, then renal replacement therapy may be required. Management of seizures, replacement of electrolytes and fluids.

CONCLUSION: Cardiac Arrhythmia can be a rare but fatal complication secondary to the consumption of over the counter stimulant drugs containing high amounts of caffeine. The presence of such severe symptoms has led to the inclusion of Caffeine toxicity, Caffeine withdrawal and Caffeine dependence among substance abuse disorders in DSM – 5. Hence it is advised that manufactures must clearly state the amount of caffeine, the health risks associated with consumption of caffeine along with the long term effects and the public should be made aware of this.

| 103 | 72 | Quality Improvement | Agrawal, Abhinav | Paavani Atluri, Koteswararao Thella, Anar Modi, Mana Rao, Imran Ismail, Tisha Tan, Madhu Paladugu | Monmouth Medical Center (Margaret Eng) | Are we being an Oxy-moron: The overuse of oxygen in a community hospital setting |
| INTRODUCTION: Oxygen is one of the most important and yet the most misused therapy in an in-patient hospital setting. In spite of having clear indications physicians often tend inadvertently order oxygen on every patient being admitted. This leads to wastage of resources and increases the cost of healthcare. More importantly, oxygen therapy is not benign and has deleterious adverse effects. Our objective was to study the overuse of oxygen and institute an intervention to prevent the wastage of resources and prevention of such potential adverse events. |
| METHODS: We designed a pilot project and implemented on one of our telemetry units. We first assessed the patients on a single inpatient floor at bedside and looked into their electronic health records for indication of oxygen, co morbidities, orders for oxygen therapy, orders for titration, actual implementation of the physician orders by nurses and respiratory therapist. Our intervention was to educate the residents, nurses on the targeted floor and respiratory therapists about the potential adverse effects of overuse of oxygen, importance of titration of oxygen to a set goal, indications and expenses involved in the usage and wastage of oxygen therapy. After 4 weeks of intervention, we collected post-intervention data using the same parameters on the same floor. |
| RESULTS: The total number of patient’s in the pre-intervention and post intervention arms were 40 each. In the pre-intervention arm, 28 patients had active orders of oxygen of which 24 had indicationsto be on oxygen therapy. 18 patients were using oxygen. 12 patients were using oxygen without being titrated to the goal of saturation >92%. In the post-intervention arm 22 patients were on oxygen of which all the patient’s had an indication to use oxygen (p-value - 0.6825). 20 of these 22 patients were using NC thus with a significant P-value of 0.0447 after an intervention. Only 4 of these patients were on therapy without titration (p value – 0.0761). Based on our calculations, the total annual saving after 1 intervention on a floor having 40 patients was $2441.12. |
| CONCLUSION: Based on our results, we concluded that oxygen is often used a placebo because of lack of awareness of its potential hazards as mentioned above and its expenses involved. This involves: (1) Oxygen therapy being initiated without an appropriate
indication. (2) Wastage of oxygen and oxygen delivery devices in patients who are off the floor or are doing well without oxygen therapy (due to lack of titration). (3) Lack of awareness about adverse effects of oxygen. By the means of education of the physicians, trainee physicians, nurses and ancillary staff, we calculated that we can save a significant amount of expense and also avoid the preventable adverse effects of overuse of oxygen therapy.

**104. Quality Improvement**

Amor, Martin Miguel I  
Michael Edward Chan MD, Nagakrishnal Nachimuthu MD, Florence Armour MS, Neil Holland MBBS  
Monmouth Medical Center (Margaret Eng)

**IMPROVED EFFICIENCY AND COST SAVINGS FROM TRIAGING SELECTED TRANSIENT ISCHEMIC ATTACK PATIENTS TO OUTPATIENT URGENT CARE**

**BACKGROUND:** Although most patients presenting with suspected transient ischemic attack (TIA) in the United States are currently hospitalized for urgent evaluation, many are later found to have alternate diagnoses, and it is not clear that hospital admission is either necessary or cost effective in all cases.

**OBJECTIVE:** To report the outcomes, diagnostic efficiency and cost savings from triaging selected transient ischemic attack (TIA) patients with lower expected stroke risks into an outpatient TIA rapid evaluation center (TREC) to avoid hospitalization.

**METHODS:** We started an open-access ABCD2 score-based outpatient TIA Rapid Evaluation Center (TREC). Patients referred to the TREC are seen on the next weekday and undergo a diagnostic evaluation then consultation with a stroke neurologist. We collected prospective data from all TREC patients seen during its first year, and compared them to the patients who were still admitted to the hospital with a primary diagnosis of TIA during the same period.

**RESULTS:** We saw 74 TREC patients within an average of 1.25 days of referral during its first year of operation (56 from the emergency room and 18 from physician offices). Only 2 TREC patients needed admission to the hospital, the remainder completed their evaluation as out-patients. Only 1 TREC patient had a follow-up cerebrovascular event. Patients referred to the TREC had lower ABCD2 scores (1.8 vs. 3.8, p <0.001) and were less likely to have a final diagnosis of TIA (19% vs. 77%, p<0.001). Nearly all patients underwent CT scan, lipid panel and EKG. However, TREC patients were more likely to undergo carotid ultrasound (99% vs. 84%, p=0.001) and MRI of the brain (89% vs. 68%, p=0.001). Based from our financial analysis, TREC patients were evaluated at significant cost savings. Both average hospital charges ($2,270 vs. $6,232, p=0.03) and average hospital costs ($666 vs. $6,523, p<0.00001) were significantly lower in TREC patients compared to hospitalized patients. In its first year, institution of the TREC resulted in average cost savings of $340,000 at our community medical center.

**CONCLUSION:** Our TREC program allowed us to avoid hospitalization for selected TIA patients, and still offer timely and efficient diagnostic evaluations at significant cost savings.

**105. Research**

Elmergawy, Hesham  
Takeddin, Juman, MD, Rochester Regional Health System  
Monmouth Medical Center (Margaret Eng)

**Raynaud’s phenomenon could be more serious than it looks!**

Chronic myeloid leukemia (CML) is a clonal hematopoietic stem cell disorder characterized by myeloid proliferation that invades the blood. The National Cancer Institute estimates 33,990 people in the United States are living with CML, with another 5,980 new cases expected in 2014, we present a case of Raynaud’s phenomenon as an uncommon presentation of chronic myelogenous leukemia in a 27 year male. A 27 year old Caucasian smoker male with past medical history of intermittent asthma not taking any medications, who has presented with complaint of intermittent painful bluish discoloration of his left thumb and index finger with numbness,
tingling and intermittent low grade fever and night sweats, review of systems was negative, and his physical examination was unremarkable except for bluish discoloration of his left thumb, no splenomegaly or lymphadenopathy. Initially during previous ED visits, he was considered of having Berger’s disease and was told to stop smoking, during his second presentation with acute onset of painful bluish discoloration of his left thumb, he has had CT angiography of left upper extremity without evidence of acute ischemia, and he has had abnormal leukocytosis WBC 30.2, segmental 68%, lymphocytes 21%, metamyelocytes 3%, myelocyte 1%, Platelet 236, hemoglobin 13.8, blood and urine culture came negative, OXR was unremarkable, Urine analysis was normal, he had negative vasculitis work up with negative ANA, C ANCA, normal RF, normal ESR, LDH was high 353, peripheral blood smear reveals neutrophilia and left shifted granulopoiesis. Eventually the patient has had cytogenetic testing with fluorescence in situ FISH analysis using DNA probes revealed BCR/ABL1 translocation in 89.5% of his nuclei results from the classic t(9:22) which was consistent with chronic myelogenous leukemia, and the patient was treated with BCR/ABL inhibitor imatinib. This case illustrates the potential of Raynaud’s phenomenon to precede the diagnosis of chronic myeloid leukemia.

INTRODUCTION:
Observation status, as delineated under CMS-1599F- Inpatient Prospective Payment System, which encompasses the "two midnight rule," requires hospitals to determine whether a patient is likely to stay for greater than two midnights in the hospital. Determination of whether the patient is either an inpatient status or an observation status has a significant financial impact on both the hospital and the patient. A scale that can reliably and consistently distinguish between patients who are likely to stay for less than 48 hours and who are likely to stay for more than 48 hours would be invaluable. Correct stratification early on in the course of admission can help hospitals adopt strategies that can partially mitigate the substantial financial losses incurred in caring for general internal medicine observation patients. At the same time, it can also facilitate the interaction between the admitting physician and patient regarding the patient status. Objective: To develop and retrospectively validate a scale that can reliably predict length of stay less than 48 hours.

METHODS:
The scale was devised at the Department of Medicine, Monmouth Medical Center. In devising the scale, experience gained by reviewing several thousand observation cases over several years by the faculty member mentoring the project, along with the insight obtained by reviewing the current literature and about 1000 recent observation admissions at MMC by the authors was utilized. The scale was validated retrospectively. A list of 400 consecutive patients on medicine service who were discharged by a physician from MMC in less than 48 hours after being admitted between the months of January 2013 and December 2013 was generated from medical records. Patients who were transferred to other hospitals for inpatient care, expired, left against medical advice (AMA), transferred to hospice, transferred to other departments were excluded from the list. Similarly, a contemporaneous consecutive list of 414 patients who were discharged by a physician from MMC after greater than 48 hours was generated. The scale was then applied to these patients by reviewing their electronic medical records and the data was tabulated and analyzed with the help of MS office Excel sheet and QuickCalcs-Graphpad software.

RESULTS AND CONCLUSION:
MEET score of greater than or equal to 3 predicts a length of stay less than 48 hours with 90.5% sensitivity (95% CI between 87.10%-93.15%) and 93.47% specificity (95% CI between 90.64%-95.51%). MEET score is easy and quick to calculate as it utilizes parameters readily and routinely available in the ED records at the time of admission. Based on the high sensitivity and specificity of the MEET scale as shown by retrospective validation, it is recommended that it be studied in a
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| 107 | 15 | Research | Jasdanwala, Sarfaraz | **Impact of diagnostic cut off levels on the sensitivity and comparison of sensitivities of serum lipase and amylase for the diagnosis of acute pancreatitis**

**INTRODUCTION AND OBJECTIVES:** Laboratory testing of serum amylase and/or lipase levels is central to the diagnosis of acute pancreatitis (AP) as these tests are quick, cheap, reliable and perhaps the only objective criteria available at the point of initial patient contact. This study attempts to answer two questions: 1. What are the sensitivities of lipase and amylase at various cut off levels with imaging (either CT or MRI) as gold standard. 2. What are the implications of using 3 times upper limit of normal (ULN) elevation of lipase/amylase for the diagnosis of AP in terms of impact on sensitivity of these two tests and clinical outcomes.

**METHODS:** In this retrospective chart based study, after approval from the institutional review board, a total of 455 cases that met the ACG diagnostic criteria for the diagnosis of AP were included. All the information was obtained from the electronic medical records. The serum lipase and amylase levels along with imaging findings on either CT and/or Ultrasound Abdomen and/or MRI/MRCP were recorded for these cases along with time since onset of symptoms, possible etiology and severity of the disease based on Modified Atlanta Classification and Determinant Based Classification. Data was recorded and analyzed with the help of Microsoft excel sheet and IBM SPSS statistic standard software.

**RESULTS:** Overall sensitivity of lipase is superior to amylase at all cut off levels in our study and the difference between the sensitivity of lipase and amylase was statistically significant. The sensitivity for lipase and amylase at ULN and 3 times ULN were 96.85%, 76.09% and 77.27%, 50.19% respectively. Out of the 455 cases under study, a total of 42 (9.22%) had non-diagnostic lipase elevation. Out of these 42, only 2 cases (4.7%) had amylase elevations that were diagnostic. A total of 148 (32.52%) cases on the other hand had a non diagnostic amylase elevation. Out of these 148 cases, 140 (94.59%) had a diagnostic lipase elevation.

**CONCLUSIONS:** Lipase has greater sensitivity as compared to amylase. With the existing knowledge that testing for lipase elevation alone offers the same degree of diagnostic accuracy as testing for both lipase and amylase, many Emergency Rooms are testing for only lipase elevation in patients with suspected AP. This approach seems to be justifiable and is recommended based on our study. If the clinical suspicion of AP is high, non diagnostic lipase or amylase elevations cannot rule out the diagnosis of AP, as a significant number of moderate cases of AP can present with non diagnostic enzyme elevation between ULN and 3 times ULN. This recommendation is concordant with the current ACG practice guidelines.

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| 108 | 120 | Clinical Vignette | Elfandi, Ali | **Is it truly eosinophilic gastroenteritis or celiac or both?**

Eosinophilic gastroenteritis (EG) is a rare disorder commonly presenting with abdominal pain, nausea, vomiting, and diarrhea. It is characterized by inflammation and eosinophilic invasion of the gastrointestinal tract, in the absence of a clear cause of the eosinophilia. We present a 23 year old Caucasian male with PMH of systemic lupus erythematosus (SLE) with nephritis. The patient was doing until 3 weeks prior to admission when he developed nausea, vomiting, and non-bloody diarrhea with diffuse abdominal pain.
pain and fevers. He lost 15 Lbs. of his weight within the last 3 months. He was placed on ciprofloxacin and metronidazole as an outpatient, with improvement after 2 days. On presentation the patient had tenderness in his epigastric and left lower quadrant areas. His initial workup revealed a WBC of 16.47 (30.8% eosinophils). His random fecal fat and low serum albumin suggested malabsorption, so celiac disease (CD) was considered. IgA tissue transglutaminase was positive (22.7), with a duodenal biopsy revealing mild blunting of villous architecture, he was started on a gluten free diet. There was some improvement in his white cell count but his eosinophils were still high. The diagnosis of eosinophilic gastroenteritis was made after being confirmed with multiple biopsies from the third portion of the duodenum, the antrum and body of the stomach all revealing > 20-30 Eosinophils/ high power field. The patient markedly improved after he was started on steroids and was symptom free within 2 weeks. The prevalence of EG in the United States is estimated to be 22 to 28 per 100,000 persons. Although the etiology of EG is idiopathic; it is believed that an allergic mechanism is responsible in a subset of patients. Any part of the GI tract can be affected in EG, but the stomach and small intestine are usually involved. It usually presents with nausea, vomiting, abdominal pain and sometimes diarrhea and weight loss. Some of the presenting symptoms and laboratory findings seen in EG can be similar to the presentation of some malabsorption syndromes, which can make the diagnosis of EG challenging. The evaluation of patients suspected with EG should be directed toward excluding other possible causes of eosinophilia, given the broad differential diagnosis such as intestinal parasites, malignancies, inflammatory bowel diseases and CD. Having both EG and CD in the same patient, or an overlapping type of picture has been described in a few case reports. We believe our patient had EG with underlying CD. After being treated with steroids, he will need to be kept on a gluten free diet with a possible follow up endoscopy to demonstrate the improvement of the villous blunting and complete resolution of his symptoms.

109. 275 Clinical Vignette Elfandi, Ali Oxana Ovakimyan, M.D. Overlook Medical Center (Jeff Brensilver) Reverse Takotsubo Cardiomyopathy: A rare disease with an unusual presentation

Takotsubo cardiomyopathy (TC) is a heart disease, described in its most classic form as an acute reversible systolic dysfunction, with ballooning of the apical segment of the left ventricle, and a hyperdynamic base. A rare subtype known as reverse or inverted TC is another variant characterized by akinesis of the heart base with a normal contracting apex. This is a 35 year old female diagnosed with multiple sclerosis 3 months prior to her hospitalization. The patient woke up in the early morning suddenly having shortness of breath, with pink frothy sputum coming out of her mouth. On arrival to the emergency department she was hypoxic with a pulse oximetry saturation of 88% and hypotensive. She was intubated and transferred to the Coronary Care Unit. Her EKG was normal sinus rhythm with nonspecific T wave changes in the anterolateral leads. The chest x-ray and CT scan showed increased interstitial lung markings consistent with pulmonary edema. Her initial troponin was 0.105 ng/mL, with a peak troponin of 1.81 ng/ml. Her BNP was 18 pg/ml, that later increased to 918 pg/ml. A transthoracic echocardiogram (ECHO) revealed a normal left ventricular size, basal anterior, basal inferior and basal lateral hypokinesis, with a reduced ejection fraction (EF) of 40%. Coronary angiography revealed normal coronary arteries. The patient was weaned off vasopressors and was extubated after 24hours in the CCU, with an uneventful recovery. A follow up ECHO obtained 2 days after, showed improvement of the EF to 45% with basal hypokinesis. Takotsubo cardiomyopathy, also known as stress cardiomyopathy is typically observed in patients after an acute medical illness or after a period of great physical or emotional stress. The underlying pathophysiology is poorly understood, but evidence suggests a major role of catecholamine excess and exaggerated stimulation of the sympathetic nervous system. Most studies have shown that TC is more commonly seen in post-menopausal women, rarely seen in younger patients such as in our case. Although a small retrospective study by Ramaraj etal identified 60 patients with different types of TC, demonstrated that patients with reverse-TC were significantly younger compared with those with other types of TC. The presentation of TC varies but it commonly mimics an
acute coronary syndrome, with ST elevations on the ECG and elevated troponins, with normal coronaries on angiography. In this case the patient was in cardiogenic shock with no preceding chest pain, or ST elevations on the EKG. Therefore it is important to have a high index of suspicion for this temporary cardiomyopathy. The management in most cases is supportive with expectant recovery. In some cases it can be more challenging and life-threatening, requiring invasive and urgent intervention as with our patient.

**Clinical Vignette**

**Patel, Kriya Gina LaCapra MD, FACP**

**Overlook Medical Center (Jeff Brensilver)**

**Pericardial effusion: Rare consequence of long-standing diabetic nephrotic syndrome**

**CASE:** A 53-year-old man with history of long-standing uncontrolled type 2 diabetes, hypertension, hyperlipidemia, coronary artery disease, PVD s/p lower extremity stent, developed significant leg edema eight months ago and was diagnosed with nephrotic syndrome and moderate pericardial effusion. Leg edema improved with Lasix but he was lost to follow-up until the recurrence of leg edema three weeks prior to admission. On physical exam he was hypertensive, heart sounds were normal, no JVD, +3 pitting edema in lower extremities bilaterally up to the knees. Creatinine clearance 56, 24-hour urine had 5 grams of protein. ANA and RF negative and complements were normal. ECHO revealed large pericardial effusion and partial collapse of RV free wall. 1,750 ml of serosanguineous fluid was drained from pericardium and window was placed. Fluid consisted of LDH 2,258U/L, total protein 3.9g/dl, glucose 80mg/dl, WBC 36,000 with 48% neutrophils, 43% lymphocytes. Pericardial biopsy showed chronic pleuritis, no evidence of malignancy. AFB, fungal, aerobic, and anaerobic cultures were negative. It was thought the pericardial effusion was most likely secondary to nephrotic syndrome from diabetic nephropathy. He was treated with torsemide and metolazone with improvement in lower extremity edema and renal function.

**DISCUSSION:** Pericardial effusion is an abnormal amount of fluid in the pericardial space. It can develop acutely or gradually in patients with any condition that affects the pericardium, including acute pericarditis and multiple systemic disorders. Common causes are infection, malignancy, radiation, collagen vascular disease, uremia, and idiopathic. With pleural effusions, unlike pericardial effusion, light's criteria is helpful with identifying exudative vs transudative effusions and therefore establishing etiology. It is less helpful in identifying etiology for pericardial effusion because most fluids become labeled as exudative. When patients have renal disease not severe enough to present with uremia, i.e. nephrotic syndrome in this patient, the incidence of pericardial effusion is extremely rare. Uremia and dialysis-associated pericarditis, however, is frequently associated with pericardial effusion and accounted for 12% of cases in the US. In literature, there were two studies which looked at association of nephrotic syndrome and pericardial effusion. The first study concluded nephrotic syndrome leading to pericardial effusion was entirely secondary to SLE or another secondary cause not associated with extra-cellular volume expansion. The second study, however, concluded pericardial effusion in childhood NS was more frequent in SLE than non-SLE patients but occurred in both groups. There was only one isolated case report of 67-year-old man with nephrotic syndrome from long-standing diabetes who developed pericardial effusion and presented similar to our patient.

In conclusion, patients with long-standing uncontrolled diabetes mellitus who develop nephrotic syndrome and are not uremic can in rare instances also develop pericardial effusions. Most do not present with any overt signs or symptoms of pericardial disease.

**Clinical Vignette**

**Ramirez, Roberto Gina LaCapra MD**

**Overlook Medical**

**Is all Cardiac Amyloidosis from AL Amyloidosis?**
Center (Jeff Brensilver)

63 years old African American female with ESRD, congestive heart failure, poorly controlled hypertension was admitted for increasing shortness of breath. Patient had BP 208/112, JVD, crackles bilateral bases, and edema. EKG: ST elevations(V3, V4) BNP: >5000 Patient was diagnosed with STEMI, CHF and hypertensive crisis and treated accordingly. She developed upward gaze, confusion and jaw trismus which progressed to nonresponsiveness to verbal stimuli. EEG revealed nonconvulsive status refractory to all antiepileptics. She suffered a cardiac arrest which led to responsiveness only to painful stimuli. Macroglossia was then noticed.

Echocardiogram: thickened left ventricle/dense myocardial speckling suggestive of amyloidosis. Protein electrophoresis and immunofixation: normal making AA and AL less likely. Genetic testing for TTR was not sent. She became more alert, but then developed a complication from her tracheostomy and passed away. Autopsy was declined.

Discussion

Amyloidosis frequently goes unrecognized for long periods of time due to its wide variety of nonspecific clinical manifestations like fatigue, dyspnea, arthralgia, numbness, macroglossia, and its associations with different diseases (AL: plasma cell dyscrasias and AA: chronic inflammation). Macroglossia is seen in AL, although could also be present in TTR, Cardiac involvement is most commonly seen in AL and TTR amyloidosis which is the most common hereditary type of amyloidosis (Val122Ile allele). 50% of AL patients have clinically significant cardiac involvement but there is limited data on TTR. The most common cardiac manifestation is heart failure in both AL and TTR, atrial fibrillation, poor LV diastolic function, and poor atrial mechanical function. TTR is more severe, more prone to develop atrial fibrillation, and tends to present later in life. The TTR variant has a prevalence of 3.5% in the African-American and African-Caribbean population. 25% of cardiac amyloidosis in African American and African Caribbean present with this allele mutation.

Echocardiography is the initial noninvasive test of choice although hypertensive cardiomyopathy can appear similar. Most common findings in amyloidosis are increase in left ventricular wall thickness and diastolic dysfunction with E/e' ratio >13. Increased echogenicity, described as “sparkling” or “granular.” Definitive diagnosis is amyloid deposits by Congo red stain on an endomyocardial biopsy or another organ. The genetic test for TTR is helpful in differentiating the two entities. Loops diuretics are a mainstay for AL and TTR cardiac amyloidosis. ACE inhibitors, calcium channel blockers and beta blockers have not shown proven benefit. In AL amyloidosis therapy involves chemotherapy and/or autologous stem cell transplantation (ASCT). In TTR amyloidosis treatment is liver transplantation. Cardiac amyloidosis should be considered in any adult with unexplained heart failure and an echocardiogram showing increased wall thickness. TTR amyloidosis should be suspected in any patient of African American/African Caribbean descent over the age of 50 who has unexplained left wall thickening on echocardiogram.

Overlook Medical Center (Jeff Brensilver)

Clinical Vignette

Ross, Andrew

Inflammatory myopathy vs. Rhabdomyolysis

SH is a 41 year old female with no past medical history, no recent medications, with a strict exercise regimen who was well up until two months ago when she developed proximal bilateral lower extremity soreness that progressed to weakness to the point of trouble walking. She is a fit, African American female with decreased strength in the proximal muscles of her bilateral lower extremities. No rash. Diagnostics: • Creatine Kinase(CK): 10,132 U/L • ANA: positive • SSA: positive(363) • Aldolase: elevated(142.1) • AST-490, ALT-320 • Acetylcholinesterase receptor binding antibody: negative Initial impression was rhabdomyolysis secondary to exercise and was hydrated aggressively. The soreness improved but not her weakness. CK remained around 7000 throughout admission. Due to persistent weakness, CK elevation, and MRI revealing asymmetric abnormal signal and edema within the posterior compartment musculature and subcutaneous fat bilaterally, impression was inflammatory myopathy. Steroids were started and muscle biopsy revealed skeletal muscle with a few necrotic fibers. Rheumatology felt she had polymyositis and the lack of a large lymphocytic infiltrate and more necrotic fibers was secondary to steroids started 5 days before the biopsy. Weakness improved and her steroids were tapered. Discussion: Inflammatory myopathies are a group of disorders.
including polymyositis (PM), dermatomyositis (DM), and inclusion body myositis (IBM). DM and PM occur between 40-50, female predominance of 2:1, with a sub-acute onset lasting several months. IBM is often males, average age 60 with an insidious onset of symptoms lasting several years before diagnosis. Proximal muscle weakness is hallmark clinical finding in all three although distal muscle can be predominant in some patients with IBM. Skin manifestations only occur in DM, gottron’s papules and the heliotrope eruption being pathognomonic. Muscle enzymes (creatine kinase, aldolase, aminotransferases, lactate dehydrogenase) can be elevated in all. ANA is positive in up to 80% of PM and DM cases. IBM is not associated with a positive ANA. There is overlap in MRI findings including active inflammation, fibrosis, and calcification. However, IBM tends to be more anterior compartment and asymmetrical. Capillary injury and perifascicular myofiber atrophy and fibroses are characteristic to DM. PM biopsy demonstrates a cellular infiltrate with necrotic and regenerating muscle fibers. IBM shows vacuolated muscle fibers and inclusions that have staining characteristics of beta-amyloid deposits. Rhabdomyolysis is a syndrome characterized by muscle pain, weakness, increased total CK (as high as 100,000), and necrosis with leaking of muscle cell contents into the serum. Inflammatory myopathies and rhabdomyolysis can be difficult to distinguish. The lack of decline of CK, proximal muscle weakness, and abnormal autoimmune testing are good indicators of inflammatory myopathies. History of insidious course, no clear cause, muscle weakness, skin manifestations, and mildly elevated CK, hydrate the patient while working up for inflammatory myopathy.

113 27 Clinical Vignette Scott, Amanda Dr. Jenny Cabas-Vargas Overlook Medical Center (Jeff Brensilver)

**Adult Onset Still’s Disease Presenting with Pulmonary and Pleuropericardial Manifestations**

A 26-year-old Romanian man presented with 1 week of odynophagia, non-productive cough, pleuritic chest pain, cyclic high grade fevers (103°F), night sweats, arthralgias of both ankles, diffuse myalgias and hypoxemia with oxygen saturation of 80-90%. He failed outpatient treatment with amoxicillin for presumed community acquired pneumonia based on chest X-ray that documented diffuse pulmonary infiltrates and bilateral pleural effusions. At time of admission had leukocytosis, thrombocytosis, transaminitis with AST and ALT twice the limit of normal and elevation of CRP (190 units) and ESR (51 mm/h). CT chest documented bilateral pulmonary infiltrates, pleural effusions and large pericardial effusion, was confirmed by echocardiogram. CT abdomen revealed mild ascites and small retroperitoneal lymphadenopathies. Inpatient treatment with ceftriaxone and azithromycin failed to control his fevers and empiric treatment for pneumocystis carinii with bactrim and intravenous methyprednisolone was started. Colchicine and ibuprofen were added for pericardial effusion. He had improvement, but as soon as his steroids were tapered, fever and pleuritic chest pain recurred. Infectious work up including serologies for legionella, streptococci, influenza A and B, Coxsackie, respiratory viral panel, HIV, CMV, Hepatitis B and C and Lyme disease were negative. Diagnostic thoracenthesys yielded a mononuclear transudate, negative for AFB. RF, anti-ccp and ANCA antibodies were negative, ANA came back equivocal and ferritin was elevated at 7735. C3 and C4 levels were normal. B2 microglobulin was normal and flow cytometry was negative for lymphoma. In view of negative infectious and malignancy work up and findings suggestive of Adult Onset Still’s Disease, IV steroids were continued. His chest pain, ferritin, ESR and CRP improved only slightly and methotrexate was added and titrated to 15mg weekly. AOSD is a rare systemic inflammatory disorder affecting both genders equally, with majority of patients between ages 16 and 35. Etiology is unknown. T-helper (Th1) predominance with elevation of IL-2, IL-6, IL-8, interferon-γ and TNF-α has been reported in persons with active AOSD. According to the Yamaguchi criteria, this patient met the requirements for diagnosis. Though he never developed the characteristic salmon-colored rash, he had diffuse muscle pain, ankle joint tenderness, significantly elevated ferritin, some lymphadenopathy, pericardial and pleural effusions, leukocytosis, pharyngitis, transaminitis and 1 to 2 high-grade fevers per day. The presentation can be very heterogeneous and we must keep it in mind when other infectious, malignant and inflammatory processes have been ruled out. Corticosteroids are the mainstay of therapy. Hence, there is no surprise that each time we tried to wean this patient off the prednisone his pleuritic chest pain worsened. Data has also shown that methotrexate decreases disease
activity and can be added as a steroid-sparing agent. For refractory cases, biologic therapy with agents blocking IL-1, IL-6, and TNF-α has shown to be beneficial.

### Hypereosinophilic Syndrome: What’s It All About?

A 37-year-old female with asthma/seasonal allergies was well until 2 months ago when she reports productive cough and dyspnea. She was found to have diffuse bilateral infiltrates, leukocytosis with 60% eosinophils, ESR 99, BNP 564, IgE 2790, RF 17.5, troponin 20. She failed courses of azithromycin/levaquin. CT chest confirmed the abnormal CXR findings along with minimal hilar lymphadenopathy and moderate to severe pericardial effusion. EKG revealed sinus tachycardia, low voltage, small q waves inferiorly. Physical exam: T100.0; HR123 regular; RR18; BP88/64. No apparent distress, no carotid bruits, positive Kussmaul's sign; S1S2, Grade I/II systolic murmur heard best at the apex, Pulsum paradoxicus: 5mmHg, diffuse rhonchi but minimal wheezing, no edema. 2D echo: moderate hypertrophy of posterior wall (1.5cm), which is characteristic of eosinophilic myocarditis, EF 40-45%, mild to moderate mitral regurgitation and moderate to severe pericardial effusion. Stool for ova and parasites: negative Bone marrow biopsy: normocellular marrow with maturing trilineage hematopoiensis, mild myeloid hyperplasia & moderate to marked eosinophilia (15-20%). She was admitted with idiopathic hypereosinophilic syndrome with eosinophilic myocarditis and was given solumedrol. Her dyspnea, cough, eosinophilia, and pericardial effusion improved and was discharged home. Hypereosinophilia is defined as: a) eosinophil level >1.5x10⁹/L on 2 examinations at least 1 month apart and/or b) tissue hypereosinophilia: eosinophils >20% of all nucleated cells on bone marrow, and/or tissue infiltration that is extensive according to a pathologist, and/or marked deposition of eosinophil granule proteins in tissue. Hypereosinophilic syndrome is the association of hypereosinophilia, with eosinophil-mediated organ damage, provided other causes for the damage have been excluded. Hypereosinophilic Syndrome Categories: 1) Primary/neoplastic: underlying stem cell, myeloid, or eosinophilic neoplasm; considered clonal. 2) Secondary/reactive: overproduction of eosinophilopoietic cytokines by other cell types, and is polyclonal (parasitic infections, certain solid tumors, and T cell lymphoma). 3) Idiopathic: underlying cause of hypereosinophilia remains unknown despite extensive work-up. Presentation of hypereosinophilic syndrome can be very heterogenous. Pulmonary involvement is seen in 25% of patients; most common symptoms are dyspnea, cough which our patient had; as well as wheezing. Abnormal CXR/CT findings include patchy ground glass parenchymal infiltrates, pleural effusion, intrathoracic lymphadenopathy and pulmonary emboli. Cardiac involvement is seen rarely; in only 5%; although it was the main presentation for our patient. Some patients with sustained eosinophilia never develop cardiac involvement, and the severity of cardiac injury does not correlate with the degree of peripheral eosinophilia. Treatment for patients who are symptomatic or have end organ damage who do NOT have the FIP1L1-PDGFRA fusion or another imatinib-sensitive tyrosine kinase mutation is glucocorticoids. Hydroxyurea, interferon-alpha or anti-IL-5 agents is utilized for patients who have not responded to glucocorticoids or as a steroid-sparing agent with lower dose glucocorticoids. Allogeneic hematopoietic cell transplantation is an option for those who have failed these interventions.

### AN UNUSUAL CASE OF BACTEREMIA COMPlicated BY CHOleCYSToduodenAL FISTULA - A CASE REPORT

A 79-year-old Caucasian man with Diabetes Mellitus type 2, hypertension, gastroesophageal reflux disease, presented with one day of fever, chills and mental status changes described as “not being himself.” He denied headache, photophobia, abdominal pain, vomiting or diarrhea. There was no reported cough, shortness of breath, or chest pain. He had a routine dental cleaning procedure two months prior to his presentation. In E.D he was tachycardic at 108 bpm, with a temperature of 104.7 &####8304;F. He had bilateral
diffuse crackles, regular heart sounds with II/VI systolic murmur loudest in aortic area and chronic bilateral lower extremity edema.

His abdomen was soft, non-tender with no organomegaly. He was alert and oriented to time; place and person, with no signs of meningeal irritation or focal motor/sensory deficit. His complete blood count with differential, basic metabolic panel and urinalysis were within normal limits; chest X-ray and CT head revealed no abnormalities. He had a procalcitonin of 38 ng/ml. Liver function tests were within normal limits. His blood culture yielded Streptococcus mitis sensitive to Cephalosporins. TEE was negative for valvular vegetations. His CT abdomen showed contracted gallbladder with multiple stones, and HIDA scan was consistent with acute cholecystitis. He was treated with Ceftriaxone; he underwent cholecystectomy which revealed a cholecystoduodenal fistula with communicating stone, which was closed surgically without complications. The pathology report concluded severe acute cholecystitis with cholelithiasis. The patient went home on IV antibiotics, and repeat cultures 2 weeks later were negative.

**DISCUSSION AND RECOMMENDATION:** Streptococcus mitis is a mouth commensal, member of the Viridans group of the Streptococcus genus. It is present in dental plaque and most commonly causes subacute endocarditis or prosthetic joint infection. Acute cholecystitis linked to this bacterium is not commonly reported in the literature. Treatment of choice is Penicillin G or Amoxicillin, or Erythromycin for Penicillin allergic patients. Incidentally, our patient also had a biloenteric fistula, itself a rare complication of gallbladder disease with calculus. It is mostly seen in the elderly and usually identified during surgical procedures given its nonspecific symptoms. These fistulae are noted to develop secondary to the erosion of gallbladder wall by necrosis caused by chronic irritation, followed by the passage of the stone through adjacent viscera. Most common site of passage is through the duodenum, known as a cholecystoduodenal fistula. An acute attack of cholecystitis, in which the adjacent serosa becomes inflamed and adheres to the gallbladder is even rarer, yet a known mechanism of biloenteric fistula formation. We recommend that physicians remain suspicious of Streptococcus mitis seeding in sites other than the heart valves, while appropriate workup and treatment not be delayed.

**Clinical Vignette**

A 66-year-old woman with medical history of hypertension, rheumatoid arthritis, chronic back pain with sciatica was brought to our hospital for worsening confusion, weakness, nausea, vomiting and diarrhea. She had seen her primary care physician two weeks prior for worsening back pain and was prescribed a Prednisone taper, Acetaminophen with Codeine and Gabapentin. The patient was a retired librarian who lived at home with her husband and was a former smoker of ½ pack of cigarettes for 34 years. She denied drug use. Her family history was pertinent for pancreatic cancer in her father. On physical examination she was noted to have moderate epigastric tenderness, hypotension and abnormal laboratory data. Her blood work showed Hematocrit of 34.5, BUN/Cr 70/3.9, AST 351, ALT 229, ALP 830, TBili 10.7, DBili 8.2, Lactic acid 6.6, and Ammonia level of 66. She was admitted to the ICU with severe sepsis suspected of biliary source, and she was started on broad spectrum antibiotics. Esophagogastroduodenoscopy showed diffuse gastritis and she was started on antacids. Ultrasound of the abdomen reported heterogeneity of the liver, dilated CBD, contracted gallbladder with wall thickening and sludge without sonographic Murphy's sign. She underwent ERCP which revealed cystic duct obstruction and narrow CBD. A biliary stent was placed with good bile efflux. The recommendation was made for cholecystectomy when more stable. Her condition deteriorated and she was intubated for respiratory failure. Tumor markers showed CA 125 of 173, CEA was 37.6, Ca 15-3 was 99, Ca 19-9 was 242, and AFP was 0.97. Her bone marrow biopsy resulted in metastatic high-grade neuroendocrine carcinoma. Given her grim prognosis, life sustaining measures were withdrawn on hospital day 11.
DISCUSSION AND RECOMMENDATION:

Neuroendocrine tumors (NETs) are rare and develop in the cells of the neuroendocrine system. These cells are dispersed throughout the body and their most common sites are the gastrointestinal tract and respiratory mucosa. NETs are classified into well differentiated neuroendocrine tumor, or poorly differentiated neuroendocrine carcinoma (NEC). The WHO classification for these tumors depends on their level of differentiation (histological resemblance of tumor to counterpart cell) and grading (proliferative activity of the tumor). The clinical diagnosis is based on history and physical, radiographic findings, tumor markers and imaging looking for a source of origin. Neuroendocrine carcinomas are poorly differentiated which means it may be challenging to locate the site of origin, which was the case with our patient. In biopsy specimens NECs have less intense immunoexpression of neuroendocrine markers. Because of their advanced nature at the time of diagnosis, treatment consists of palliative chemotherapy with Cisplatin or Etoposide. We encourage physicians to carry some level of suspicion for malignancy in cases of non-resolving or worsening organ-specific symptoms despite aggressive medical intervention.

A 65 year-old female with past medical history of hypertension, presented to our hospital in the month of September with worsening back pain and lower extremity bilateral weakness. The pain had started 24 hours prior, noted after she had gone swimming with her sister. She had visited a local Urgent Care where she received Oxycodone and muscle relaxants and sent home. Due to inability to walk, she returned to our hospital E.R, where her systolic blood pressure was over 190, C.T chest and abdomen was negative for dissection, however given her severe complaint she was admitted for further workup. Her lumbosacral MRI showed foramina disc herniation at the L2-3 and L3-4 levels. On her first night of admission a Rapid Response was called for agitation and desaturation. She had a history of alcohol use, and it was presumed she was undergoing withdrawal. She was given Dilaudid and Ativan IV. Her saturation improved to 95% on face mask. She also had 1200cc of urine removed from her bladder via foley catheter. The next morning, the patient was found obtunded, an arterial blood gas revealed severe metabolic acidosis and the patient was intubated and transferred to ICU. The initial thought was aspiration pneumonia secondary to medication for EtOH withdrawal and she was started on empiric antibiotics. Head C.T was negative for acute intracranial hemorrhage. Her deconditioning and failed ventilator weaning attempts prompted further workup. Neurology was consulted and a lumbar puncture was performed. Her CSF total protein was 223, Glucose was 93, WBC 12 with 2% Neutrophils and 90% Lymphocytes. Her West Nile virus IgM and IgG were positive in both serum and CSF. She was treated with 10 day course of IV Immunoglobulin. She received tracheostomy for respiratory failure and PEG tube and was discharged to long term care facility with some upper extremity motor improvement.

DISCUSSION AND RECOMMENDATION:

West Nile virus is a mosquito-borne flavivirus human pathogen. It commonly causes febrile disease with headache, abdominal pain, arthralgias and a maculopapular rash on the trunk, but it can also present as meningitis or encephalitis. Neuromuscular manifestations of West Nile virus have been reported in the literature. Its most common presentation is a poliomyelitis syndrome with asymmetric paralysis with or without respiratory failure. The inflammation may also involve skeletal or cardiac muscles, and peripheral nerves presenting as a Guillain-Barre Syndrome (GBS). Autonomic instability is seen in patients with spinal sympathetic neuron involvement. The diagnosis is based on serum or CSF IgM. The treatment for West Nile virus is conservative; however a therapy of IVIG has been proposed for GBS as a complication. We recommend physicians to keep a broad differential of infectious etiologies in patients with acute respiratory failure.
| 118 | 34 | Quality Improvement -- Changed per Challa request | Challa, Sridevi S. Challa MD*, E.A. King Pharm D*, S. Anghel MD*, J. Brensilver MD*, L. Bielory MD *#, Overlook Medical Center (Jeff Brensilver) | Impact of penicillin skin testing (PST) on antibiotic (abx) use in patients with a penicillin allergy (PA) |

**RATIONALE:** PA (Penicillin Allergy) generally leads to the use of broad-spectrum abx (antibiotic) that may increase complications and cost. A PST pilot protocol was developed at Overlook Medical Center to help reduce the use of broad-spectrum/high-cost (BS/HC) abx and determine the cost effectiveness of PST (Penicillin Skin Testing).

**METHODS:** A retrospective analysis was conducted on patients who had PST performed by an allergist between November 2013 – May 2014. PST included prick and intradermal testing with major (Pre-Pen™) and minor (penicillin G) determinants followed by an oral amoxicillin challenge. Abx were de-escalated based on results of PST. Reduction in cost of abx, and adverse reactions were assessed. Cost analysis was based on the material cost of abx and skin testing components. Study was IRB approved.

**RESULTS:** A total of 38 adult patients had PST. Median age was 64 yrs; 97%(n=37) were allergic to penicillin alone; piperacillin/tazobactam (n=1), and a history of multiple abx allergies (n=9). Sixty three percent had a reaction >20yrs ago with cutaneous reaction being the most common reaction (45%). Aztreonam was the most common initial abx used prior to PST (55%).Urinary tract (26.3%) and intra-abdominal infections (18%) were the most common indications for abx treatment. Skin testing was negative in 38 (100%) patients but one patient had anaphylaxis to oral amoxicillin challenge following a negative skin test. A cost analysis was performed in 29 patients who had antibiotics switched to a beta-lactam after negative PST. Fourteen patients (48%) were switched to a narrow-spectrum agent (i.e. aminopenicillins, oxacillin, and non-pseudomonal cephalosporins). Total cost of therapy with a beta-lactam after PST was $3,538. Total cost of therapy if initial BS/HC abx prior to PST were continued was $14,670. Approximate cost for PST materials for 38 patients was $3,631. Overall cost savings was $7,521. The additional savings of impact on developing adverse reactions and superinfections (i.e. Clostridium difficile) was not assessed.

**CONCLUSIONS:** Following the implementation of a PST protocol, we observed a decrease in BS/HC antibiotic use in patients with previously documented PA. PST is a safe and cost-effective procedure to serve as a negative predictor test for penicillin hypersensitivity mediated by IgE.

| 119 | 64 | Clinical Vignette | Aldabag, Saddam Melissa G, Katherine A, Talina F, Mohamad H, Mahmad A, Islam M Raritan Bay Medical Center (Abdalla M Yousif) | Skin mass as a presenting feature of Renal Cell Carcinoma: need for high index of suspicion |

**INTRODUCTION:** About one third of Renal Cell Carcinoma patients develop metastasis at the time of diagnosis affecting the lung, lymphatic, liver, bone, contralateral kidney, adrenal glands, and brain. We report a case of skin mass as an initial presentation of Sarcomatoid Renal Cell Carcinoma.

**HOSPITAL COURSE:** A 46 year old female was healthy until five months PTA when she noticed a painless skin mass over her right shoulder. MRI study reported a possible skin sarcoma. Patient was in denial and refused treatment. Within the next few months patient developed similar skin masses on her trunk and limbs. Eventually patient presented to the ED with recurrent episodes of dizziness, dyspnea, and productive non-bloody cough. Her vital signs were stable on admission. Relevant laboratory studies included hemoglobin 7.9 g/dL, hematocrit 25.8%, and platelets 568x103 per µL, MCV 63.1fL, BUN 29mg/dL, Creatinine 1.7 mg/dL, calcium
17.9 mg/dL, and Alk Phos. 367 U/L. Physical examinations revealed a left upper arm mass, a right shoulder mass, bilateral breast masses, a left hip swelling, and right foot mass. Patient was admitted to the ICU for management of the hypercalcemia and other electrolyte disturbance. CT scan revealed a mass in the upper pole of the left kidney. Breast ultrasound showed 2 cm masses on both breasts. Surgery and oncology consults were called to rule out the possibilities of multiple sarcoma, breast cancer, and kidney cancer with metastasis. Incisional biopsies and subsequent pathology report indicated sarcomatoid renal clear cell carcinoma with skin and breast metastasis.

DISCUSSION: Clinical presentation of sRCC varies widely and is dependent on the stage at diagnosis. In most published series, sarcomatoid tumors are usually extremely large, and 90% are symptomatic at presentation. The incidence of metastatic disease is extremely high at presentation, with 45%–84% having evidence of systemic disease. Metastases typically occur at the lungs, bone, nodes, liver, and brain. Cutaneous metastasis in sRCC is rare; accounting between 1 and 3% of all metastases incidence. The presentation is usually a single lesion located most commonly in the face, scalp, and trunk. The rich vascular component of cutaneous metastasis in sRCC may cause clinical confusion with hemangiomas, pyogenic granulomas, and Kaposi’s sarcoma. It can also imitate cutaneous cysts, cutaneous horns, lymphomas, or abscesses. In most published sRCC cases, skin metastases took place within six months to five years of the initial diagnosis. Our case report of multiple cutaneous metastases as presentation of sRCC, highlights the absolute importance of initial precise histological diagnosis to permit the correct identification of the skin lesion.

Clinical Vignette

INTRODUCTION: Spontaneous intracranial hypotension (SIH) is characterized by postural headaches secondary to low CSF pressures, which are not attributable to lumbar puncture (LP), trauma, surgery or any other inciting event. SIH is typically due to spontaneous spinal CSF leaks, and are an important and often misdiagnosed cause of headache in young and middle-aged individuals, especially women.

CASE: Case of a 31 year-old female with no significant past medical history c/o sudden onset intense 10/10 frontal headache worsened by sitting upright and alleviated by lying supine, a/w nausea and vomiting. Vitals and labs were stable, physical exam, including neurological exam, was within normal limits. No history of LP in the past, and patient refused an LP in ED. MRI w/wo contrast showed nonspecific fluid/edema within the C1-C2 retrospinal region, and dilatation of the right transverse sinus. LP under fluoroscopic guidance yielded an opening pressure of 0 mmH2O. Patient was diagnosed with SIH and improved with fioricet and caffeine tablets.

DISCUSSION: SIH is an uncommon, but important cause of headache in the young to middle-aged population, especially females. SIH is classically described as an orthostatic headache that worsens when the patient is sitting upright or standing and resides when lying flat, and can be accompanied by many symptoms, including nausea, vomiting, anorexia, neck pain, dizziness, photophobia, etc. SIH is usually caused by a persistent occult CSF leak, which presents with a postural headache as the most prominent symptom. Recognizing SIH in patients is essential and may prevent costly unnecessary procedures. A retrospective study done by Schievink involving 18 patients with intracranial hypotension demonstrated that 17, or 94%, were incorrectly diagnosed with a diagnostic delay ranging from 4 days to 13 years. Diagnosis of SIH is obtained with MRI of the head with gadolinium, which often shows diffuse meningeal enhancement, engorgement of the venous sinuses, subdural fluid collections, or descent of the cerebellar tonsils, often misdiagnosed as a chiari malformation. Lumbar puncture with opening pressures is confirmatory and are expected
to be 0-60 mmH2O\textsuperscript{7,8}; however, it is important to note that lumbar puncture may worsen the patient’s symptoms. Initial preferred management of SIH is conservative treatment with bed rest, fluid replacement and caffeine in the form of caffeine tablets and/or high caffeine dietary intake, which due to its vasoconstrictive properties has proven to work well\textsuperscript{7,9-11}. In the event that conservative treatment is found to be ineffective, a CT myelogram with focal epidural blood patching is indicated\textsuperscript{7,11}.

CONCLUSION: While SIH is an uncommon diagnosis, misdiagnosis of SIH is quite commonplace and is often confused with migraine, chiari malformation, pseudotumor cerebri or aneurysms\textsuperscript{7,11,13}, which subsequently leads to wrong and potentially harmful procedures including craniotomies and brain biopsies\textsuperscript{7,11,13}.

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Clinical Vignette

Arshed, Sabrina Shoab Zaffar, Ping Zhang, Waqas Jehangir, Shuvendu Sen, Mohammed Islam

Raritan Bay Medical Center (Abdalla M Yousif)

Idiopathic Sudden Sensorineural hearing loss in primary HIV infection

INTRODUCTION: Sudden Sensorineural hearing loss (SSHL) can be caused by a multitude of etiologies, infection being the most dominant. While there are multiple documented cases of neurologic and otologic manifestations in patients with established HIV/AIDS infection, it is rare for SSHL to present as a manifestation of an acute primary HIV infection, which is not accountable by any autoimmune, congenital, metabolic, or infectious causes.

CASE: We report a case of a 35 y/o male, with a past medical history of uncontrolled diabetes mellitus, who presented to the emergency department with complaints of sudden onset nausea, vomiting, dizziness, unsteady gait and hearing loss in the left ear associated with ear fullness and tinnitus. Patient reported multiple behavioral risk factors. Otoscopic exam was within normal limits. CT of the head was negative. MRI of the head only demonstrated a congenital neuroglial cyst. HIV test was done; while awaiting the results, the patient’s condition rapidly improved, and he was discharged home. After discharge, the patient’s rapid HIV test returned with a presumptive positive result; confirmatory testing proved it to be positive. The patient was notified, and followed up at the HIV clinic.

DISCUSSION: There are multiple causes of unilateral SSHL, most often caused by temporal bone injury leading to facial nerve palsy, trauma causing perilymph leakage or fistula and Meniere’s disease. However, SSHL may be idiopathic, with speculative theory implicating viral infection as the cause. There are multiple otologic complications of HIV/AIDS, which accounts for 21-49% of HIV-induced otologic manifestations. HIV-related SSHL may be attributed to primary infection by HIV of the central nervous system (CNS) or localized to the vestibulocochlear nerve. Studies have shown approximately 88% of HIV patients have evidence of CNS involvement on autopsies, and roughly 10% present to their primary physician with complaints of neurologic symptoms. Furthermore, SSHL in HIV patients may occur with or without the presence of an opportunistic infection or any clinical evidence of progression to AIDS. Lin, et al concluded that HIV infection in patients between the ages of 18-35 years had a significantly greater risk of developing SSHL than the general population without HIV (P=0.03); however, the underlying etiology of remains unclear. The majority of the research done on SSHL has been done on patients with known cases of HIV whose symptoms began after diagnosis. Our case demonstrates an individual who had no previous diagnosis of HIV and no previous symptomology commonly seen and suggestive of acute HIV infection. It becomes imperative therefore to take into consideration the varied presentation of a primary viral infection of HIV, including as the case report suggests, sudden sensorineural hearing loss.
122. Clinical Vignette

Arshed, Sabrina
Mohammed A Hossain, Rafay Khan, Qiang Nai, Mohammed Osman, Soad Enakkua, Nazir Raoof, Abdalla Yousif
Raritan Bay Medical Center (Abdalla M Yousif)

Facial Vein Thrombosis: A rare case of atypical Lemierre’s Syndrome

INTRODUCTION: Lemierre’s syndrome begins with an acute oropharyngeal infection followed by unilateral septic thrombophlebitis and multiple metastatic mostly pulmonary abscesses resulting from septic emboli. This is a rare but severe infection originating from microbes within the mouth and pharynx most commonly by Fusobacterium necrophorum.

CASE: A 29 years old hypertensive male was evaluated for 3 day history of progressive, tender left sided facial edema and fever that began by bursting an intranasal left-sided pimple. Oropharyngeal exam was notable for left sided pharyngeal erythema and tender left submandibular lymphadenopathy. A CT scan of Facial bones with contrast showed thrombosis of the second order branch of the left facial vein. Coagulopathy workup done as per hematology consultation came back negative. CT scan of chest with contrast revealed bilateral septic emboli. Patient was treated with intravenous antibiotics, and discharged on oral antibiotics.

DISCUSSION: Lemierre’s syndrome, also known as postanginal sepsis, while relatively common in the pre-antibiotic era, is a rare disease mostly affecting the young adult population. It usually presents with an acute oropharyngeal infection resulting in septic thrombophlebitis of the ipsilateral internal jugular vein; however, rare cases of thrombophlebitic spread to the facial vein have been reported by Lemierre himself. The usual causative organism is Fusobacterium necrophorum. Diagnosis is initially made via radiographic identification of the thrombophlebitis of the IJ vein, or one of its tributaries, including the facial vein, as in the case of our patient. Positive blood cultures are also a key factor in the diagnosis of Lemierre’s syndrome; however, in many cases blood cultures are found to be negative, usually due to pretreatment with antibiotics, as is the case in our patient. Furthermore, typical laboratory abnormalities include leukocytosis, thrombocytopenia, abnormal liver function and elevated C-reactive protein. Metastatic infections have also been found to be very common among those suffering from Lemierre’s syndrome accounting for approximately 63%-100% of patients, the lungs being the most common site, followed by liver, muscle, pericardium and skin, all of which are identifiable by CT. Treatment of Lemierre’s includes long term treatment with the appropriate antibiotics, and if needed, surgical drainage. The use of anticoagulation in Lemierre’s syndrome remains controversial; however, possible indications for anticoagulation therapy include lack of improvement despite adequate antibiotic therapy, thrombophilia, and advancement of the venous thrombosis. Although mortality rates range from 4%-18%, the overall prognosis of Lemierre’s syndrome is good with the prompt administration of antibiotics.

CONCLUSION: While it remains a rare disease, the incidence of Lemierre’s syndrome may be increasing, and may represent a potentially fatal illness. Primary care providers should be aware of this syndrome and have a high clinical suspicion for this syndrome in their younger patients with oropharyngeal infections with systemic illness and pulmonary involvement.

123. Clinical Vignette

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USE OF SYNTHETIC MARIJUANA AMONG YOUNG ADULTS

INTRODUCTION: Synthetic cannabinoid (SC) or “spice” refers to a variety of herbal/chemical mixtures, which mimic the effects of marijuana. This drug is generally marked as “herbal incense” and best known by the brand names of “K2”, “spice”, “aroma”, “Mr. nice guy” and “dream”. Little data is available on the actual psychopathological and physical effects of SC but some reported presentations include high blood pressure, nausea, vomiting, anxiety, agitation and seizures. Here we highlight the case of a patient presenting with both abnormal psychopathological and physical manifestations thus illustrating the importance of recognizing a new
### Alternate Form of Marijuana as a Risk Factor for Such a Presentation

**CASE DESCRIPTION:** A 28-year-old male, was brought to the emergency department by EMS 2 days prior to admission for agitation, aggressive behavior and auditory hallucinations. As per ER note patient was a habitual synthetic marijuana user. Patient was then given Ativan 4 mg IM, Ziprazidone, 1 L of NS and Benadryl on first presentation to ER. Patient was sent to crisis unit after psych evaluation for new onset psychosis. Internal medicine was then called for medical evaluation of elevated WBCs and CK. Physical examination including vital signs were normal. Internal Medicine consult was called when her Lab reports showed leukocytosis and an elevated CK. Her urine drug screen was positive for benzodiazepine and THC. Patient was treated with fluids and antipsychotics.

**DISCUSSION:** Easy access and the misperception that SC products are “natural” and therefore harmless have likely contributed to their popularity. This along with the fact that the chemicals used in Spice are not detected in standard drug test are causing it to be one of the most used by high-school seniors, second to marijuana. The U.S. Substance Abuse and Mental Health Services Administration stated that toxicity due to synthetic marijuana resulted in 11,400 cases of emergency room visits in 2010. Synthetic marijuana abusers who have been taken to Poison Control Centers report symptoms that include rapid heart rate, vomiting, agitation, profuse sweating, confusion, and hallucinations. SC can also raise blood pressure and cause reduced blood supply to the heart (myocardial ischemia), and in a few cases it has been associated with Acute Coronary Syndrome. It thus becomes imperative for the medical fraternity to raise awareness among in the society about the deleterious effects of this apparently ‘natural’ and harmless synthetic product.

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### Paranoid Personality Masking an Atypical Case of Frontotemporal Dementia

**INTRODUCTION:** Frontotemporal dementia (FTD) is a debilitating disease that is well described in the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5), and typically presents with memory impairment, progressive decline in cortical functioning, and behavioral changes. It is a relatively uncommon disease and most often misdiagnosed as a psychiatric disorder. The age of onset is generally in the fifties, and usually the first presentation involves a change in behavior and emotional blunting.

**CASE:** We present a case of a 57 years old female who presented with signs and symptoms of paranoia and frank persecutory delusions for about a year. She was socially active and had worked for an insurance agent for about 30 years prior to her developing the delusional symptoms. She has a paranoid personality but no previous psychiatric admission. Her past medical history was significant for hypertension which was well controlled. Other than speech fluency that was reduced, her physical examination was unremarkable with normal vital signs. On mental status examination, she scored 14 on the Montreal cognitive assessment test. She was neatly dressed but had body odor. She smiled frequently and seemed somewhat distant. A Complete blood count and complete metabolic profile were within normal limits. Vitamin B12 level and Thyroid stimulating hormone were within normal limits. A CT scan of the brain was unremarkable. A diagnosis of paranoid and delusional disorder non-otherwise specified was made and she was treated with Olanzapine without any significant improvement. At this point a diagnosis of FTD was considered taking into account her age and the time frame of her symptoms. A subsequent MRI of the brain was obtained which showed atrophy of frontal and temporal lobes bilaterally (left more prominent than right) which confirmed the diagnosis of FTD. Patient was appropriately treated. Treatment of FTD involves management of any neurobehavioral symptoms while trials of atypical antipsychotics are ongoing but suggest some efficacy.
**DISCUSSION**: Diagnosis of FTD is mainly clinical but MRI of the brain often plays a key role. However, misdiagnosis of FTD can be seen when patients present with atypical features. This case illustrates that FTD can present with atypical signs and symptoms such as paranoia and delusions and having a high index of suspicion in the relevant setting thus becomes imperative for appropriate diagnosis and avoidance of improper management.

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### 125. 157

**Clinical Vignette**

Iroka, Nneka

M. HOSSAIN, J. MIDDLETON

**Raritan Bay Medical Center (Abdalla M Yousif)**

**ATYPICAL PRESENTATION OF ROCKY MOUNTAIN SPOTTED FEVER IN AN YOUNG ADULT**

Rocky Mountain spotted fever (RMSF) is a curable tick-borne disease that can be potentially lethal. Its causative agent Rickettsia rickettsii is a gram-negative intracellular bacterium with a tropism for vascular endothelial cells. Classic symptoms of RMSF include fever, which is almost always present, headaches, and rash; however, all of these diagnostic clues may not be present which can lead to delay in diagnosis and appropriate antibiotic therapy leading to poor outcomes in certain cases. RMSF very rarely may involve the myocardium but solely presenting with cardiac signs and symptoms without any of the typical features—fever, rash, or headaches—is even rarer and may pose a diagnostic challenge. We report a case of an unusual presentation of RMSF in a 20 years old male patient with no past medical history who presented with severe retrosternal chest pain, electrocardiographic changes, and troponin elevation without any fever, rash, or headaches. The chest pain was associated with diaphoresis, 5 episodes of vomiting but no shortness of breath, abdominal pain. He is a non-smoker, non-alcoholic and denies use of recreational drug. He went camping a month earlier. On admission his cardiac enzymes showed a CK level of 939 IU/L (normal 40-300 IU/L), CKMB 58.62 ng/mL (normal 1.0-5.0 ng/mL). Initial troponin I was 17.50 ng/mL (normal <0.30) and 20 hours later was 54.33 ng/mL. An electrocardiogram showed a 2 mm ST elevation in inferior leads and V6. It also showed mild ST depression in leads V1-V3. Patient was given aspirin, clopidogrel, and heparin. A left heart cardiac catheterization was done, result of which revealed normal coronaries and normal left ventricular ejection fraction. At this point presumptive diagnosis of Myocarditis due to RMSF was made and diagnostic testing was further pursued. Results of RMSF IgG titre was elevated at 1:128 indicating recent infection with Rickettsia rickettsii which is the etiologic agent of RMSF. The patient was subsequently treated with Doxycycline. This case demonstrates the fact that RMSF can present solely with signs and symptoms of Myocarditis without any of the typical features of RMSF. It also strongly emphasizes the need for physicians to have a high index of suspicion in the appropriate setting and recognize the complication of Myocarditis secondary to RMSF and prevent unnecessary invasive cardiac procedures especially in young patients with no significant personal or family history.

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### 126. 310

**Clinical Vignette**

Iroka, Nneka

N. REGEVIK, P. SEN

**Raritan Bay Medical Center (Abdalla M Yousif)**

**PROLACTINOMA IN A TRANSGENDER MALE-TO-FEMALE HIV POSITIVE PATIENT-A RARE OCCURRENCE WITH A THERAPEUTIC DELIMA**

Pituitary adenoma has been reported in patients treated with estrogen or transsexual male-to-female patient. Estrogen directly stimulates the cells of the lactotroph and this can lead to lactotroph hyperplasia and even prolactinoma. Also, there have been literatures and reports that Human immunodeficiency virus infection (HIV) is associated with an increase risk of development of adenomas including endocrine adenoma such as prolactinoma. When prolactinoma is diagnosed in a patient with HIV infection and a transgender male-to-female, the question becomes is the prolactinoma an association of HIV infection or is secondary to estrogen

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exposure. Another challenge arises when it comes to management of such a patient as there have been reported significant interactions between antiretroviral drugs and drugs use for treatment of prolactinoma. We report a case of a rare occurrence of prolactinoma diagnosed in a patient who was treated with estrogen therapy and was also found to be HIV seropositive. To date there have not been any reported case as such. A 22 years old transsexual male-to-female presented with complaints of breast tenderness and galactorhea for a couple of months. Prior to this presentation she patient underwent a transgender procedure including breast and buttocks estrogen pellet implant. She took estrogen injections twice a month intermittently for 2 years. She has no other medical history besides being HIV seropositive. She was taking once a day regime of rilpivirine-emtricitabine-tenofovir combination. On physical examination vital signs were unremarkable, normal female appearing, no acute neurological deficit was seen and cranial nerve examination was grossly intact. Breast exam disclosed milk discharge from the right nipple and bilateral breast tenderness. All other exams were unremarkable. CBC and CMP were unremarkable. Serum Prolactin was 1147ng/ml (normal 2-29ng/ml). An MRI of the brain showed a 2.5 x 2.2 x 2.3cm Suprasellar mass. Given her history, laboratory findings and MRI finding, a diagnosis of Pituitary Macroadenoma was made. She was treated with Cabergoline and 3 months later her serum Prolactin level decreased to 96ng/ml. Although there has not been a documented interaction between Cabergoline or Bromocriptine and Rilpivirine, Rilpivirine like most other non nucleoside reverse transcriptase inhibitors such as Efavirenz is metabolized through the CYP isoenzyme system and may increase levels of Cabergoline by decreasing its metabolism and this can lead to toxicity. In patients who have high viral load (>100,000 copies of viral RNA/ml) at the time of starting antiretroviral therapy, it is better to start such patients on an Efavirenz-based regimens to suppress viral load before switching them to Rilpivirine-based regime. In such patients using dopamine agonist can be a real challenge due to drug interactions.

127. Clinical Vignette Jehangir, Waqas Shilpi Singh, Mohammed A. Islam, Shuvendu Sen, Abdalla Yousif Raritan Bay Medical Center (Abdalla M Yousif)

**Chronic glomerulonephritis causing Atypical Hemolytic Uremic Syndrome**

**INTRODUCTION:** Atypical hemolytic uremic syndrome (aHUS) is a life threatening disease that is usually characterized by hemolytic anemia, thrombocytopenia and kidney failure. It is caused by abnormalities in complement system, triggered by certain environmental factors or genetic mutations unlike typical HUS which is usually caused by infection with certain strains of Escherichia coli. Atypical HUS can be genetic, acquired, or idiopathic. Mutations in at least 10 genes identified so far (C3, CD46, CFB, CFH, CFHR1, 3, 4 and 5, CFI and THBD) appear to increase the risk of developing aHUS. CFH mutation is most common accounting for approximately 30 percent of all cases. The pathophysiology of aHUS is proposed to be microangiopathic in nature, where platelet thrombi occlude microcirculation resulting in organ dysfunction. It is believed that aHUS causes renal dysfunction but we propose that either condition can precede the other.

**CASE:** We report a case of a 26 year old male without any past medical history who presented to E.D. with malaise for one week. Laboratory studies and imaging were consistent with hemolytic anemia, thrombocytopenia and renal insufficiency prompting the presumptive diagnosis of atypical TTP/HUS. Testing for ADAMTS13, complement C3 and C4 levels were normal. Renal ultrasound discovered atrophic kidneys consistent with severe CKD and kidney biopsy revealed focal segmental collapsing glomerulosclerosis without any evidence of microthrombi. He was treated with emergent plasmapheresis and hemodialysis. Genetic studies for CFH, C3, CFB and CFI mutations were sent, and heterozygous common deletion of CFHR1–CFHR3 was identified. The patient was subsequently referred to a specialized center for treatment with Eculizumab and possible renal transplant.

**DISCUSSION:** The sequence of events and kidney biopsy findings for our patient suggest that he had an underlying chronic
glomerulonephritis which was complicated by aHUS, which has been reported previously. We propose that some common genetic mutations may predispose patients to glomerulonephritis and subsequent development of aHUS. Till now it has been postulated that underlying pathophysiology in aHUS is chronic, uncontrolled activation of the complement system. In this case normal complement levels and underlying chronic glomerulonephritis suggests an alternate mechanism for development of aHUS.

Understanding the fatal consequences of this syndrome, we emphasize the absolute importance and necessity for further testing and research into the mechanism of aHUS and possible genetic linkage of chronic glomerulonephritis to aHUS.

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<th>Jehangir, Waqas Tarek Aly, Shuvendu Sen, Mark L. Niemiera, Abdalla Yousif</th>
<th>Raritan Bay Medical Center (Abdalla M Yousif)</th>
<th>Multi-vessel Spontaneous Coronary Artery Dissection: A Diagnostic Challenge in an Unlikely Setting</th>
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<td>As a healthcare professional, anchoring heuristic is a diagnostic error that one must be aware of when approaching every patient. This diagnostic error is the healthcare professional’s tendency to rely on a previous diagnosis, and in situations where a set of symptoms might mask a rare and deadly condition, this error can prove fatal for the patient. One such condition, Spontaneous Coronary Artery Dissection, is an uncommon and malefic presentation of coronary artery disease that can lead to myocardial infarction and sudden death. Spontaneous Coronary Artery Dissection SCAD is seen predominantly in women with a mean age above 40. In a young, athletic male with a SCAD, the danger of diagnostic error is overwhelming due to the broad symptomatology and the betraying demographic. A 27 year old man without any significant past medical history presented with a two week history of intermittent, retrosternal, stabbing chest pain that radiated to his left arm and shoulder. The pain was associated with nausea and diaphoresis. As the pain was pleuritic and positional, and the patient had an upper respiratory tract infection with fever and chills prior to the onset of the pain, his primary care physician had made the clinical diagnosis of costochondritis. While engaged in a sporting activity, the pain became exacerbated and the patient was admitted due to concern for a cardiac cause of the distress. On physical examination, patient was tachycardic and tachypneic while his blood pressure and other findings were unremarkable. His initial lab results showed leucocytosis. Follow up ECG showed ST elevation in anterolateral leads which prompted a cardiology consultation. After consultation, the emergency physician called a Code Heart and the patient was sent to the cardiac catheterization lab for rescue angioplasty. He was found to have a 95% mid Right Coronary Artery (RCA) dissection with a total Left Anterior Descending (LAD) artery spontaneous dissection while all other remaining vessels were found to be normal. Percutaneous Coronary Intervention to the LAD and mid RCA was performed with intra-aortic balloon pump placement. The initial diagnosis of costochondritis made by the primary care provider was the diagnostic formulation that could have influenced future judgments made in the therapeutic plan. This example presents the danger of anchoring heuristic as this patient did not fit the clinical demographic of SCAD and the diagnosis of costochondritis could have delayed pivotal interventions that could have been life saving. Chest pain must always be taken into serious consideration and Acute Coronary Syndrome should always be ruled out regardless of the presenting demographic.</td>
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<th>Clinical Vignette</th>
<th>Khan, Rafay Abdalla Yousif, M.D., Abdul Mahmad, M.D. Hiyam Ibrahim M.D.</th>
<th>Raritan Bay Medical Center (Abdalla M Yousif)</th>
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<td>mucinous adenocarcinoma of the bladder: tumor markers in a young adult with unclear etiology</td>
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<td>Introduction: Primary mucinous adenocarcinoma of the bladder is a rare urologic entity, which is found in less than 2% of all bladder tumors. The average age of bladder cancer incidences being in the late sixties, it is unusual to find a young adult diagnosed with mucinous adenocarcinoma of the bladder. Some epidemiologic studies have been able to identify chemical carcinogens</td>
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believed to be associated with Bladder Cancer, with cigarette smoking being reported in half of the cases. We highlight a case of Mucinous Adenocarcinoma with positive tumor markers in a young adult with no risk factors, with possible correlation with other malignancies including Colon Cancer.

**CASE DESCRIPTION:** A 27-year-old Hispanic male presented with new onset painless hematuria following a 4-month history of abdominal pain. Patient was born in Peru, is a non-smoker, social drinker, with a family history significant for colon cancer in the father. Patient has been working at UPS for a few years and his job required him to load trucks, with no known chemical exposure. Patient underwent CT abdomen and pelvis that demonstrated a calcified mass at the dome of the bladder infiltrating adjacent lower abdominal mesentery. Multiphasic post-contrast CT showed a calcified mass at the dome of the urinary bladder with reticulation of mesenteric fat concerning for peritoneal carcinomatosis. Cystoscopy with partial transurethral resection of bladder tumor was performed. Following dome-biopsy, diagnosis of Urachal primary Mucinous Adenocarcinoma with prominent Signet Ring Features was made with extension into subepithelial connective tissues. The tumor cells although negative for p53 were found to be strongly positive for CK20 and CDX2. Post resection, patient is currently undergoing chemotherapy at a Cancer Institute.

**DISCUSSION:** Although the oncogenesis of urothelial tumors in young patients is unclear, multiple environmental and genetic factors may contribute to the etiology. The amount of tobacco smoking and certain occupation exposure have been documented to be known risk factors for urothelial tumors. The causes of Bladder Cancer in the young age group however have not been well reported in the literature. CK20 and CDX2 were tumor markers found to be present in our patient and have been shown to be prominent in some other malignancies, especially colorectal cancer. It thus brings into question and certainly opens up further research possibilities of a potential genetic linkage between colon cancer and early onset bladder cancer and whether a patient with a positive family history of colon cancer with above mentioned markers markers should have early screening for bladder cancer. Furthermore, further research should determine whether a young patient diagnosed with Bladder Cancer with relevant tumor markers may necessitate early colonoscopy screening.

130. 89 Clinical Vignette Luo, Hongxiu Harsh Bhatt, Eric Uhrik, Shaza Mohamad, Teena Mathew, Abdalla Yousef Raritan Bay Medical Center (Abdalla M Yousef) Acute Pancreatitis: unexpected side effect of dimethyl fumarate for treatment of relapsing-remitting multiple sclerosis

Multiple Sclerosis (MS) is a chronic inflammatory immune-mediated disease of the central nervous system. Relapsing/Remitting MS (RRMS) accounts for nearly 85% of the cases. Dimethyl fumarate (Tecfidera) was approved by FDA in March 2013 for treatment of RRMS. Although the side effects, on the basis of phase three studies, include headache, episodic flushing, and gastrointestinal disturbances, decreased white blood cell and lymphocyte cell counts, and elevated liver enzymes, no reports of acute pancreatitis has ever been reported. We present a case of acute pancreatitis believed to be induced by this new medication. Our patient is a 43-year-old Hispanic female with past medical history significant for Relapsing/Remitting MS, hypertension and cholecystitis status post cholecystectomy 10 years prior to the presentation, who presented with three days of epigastric abdominal pain. Her pain was sharp, on a scale of 10/10, intermittent, worse with eating and associated with multiple episodes of non-bloody, non-projectile vomiting. She denied trauma, insect bite, history of prior episodes of pancreatitis, alcohol or drug abuse. Her current medications included Diltiazem 100 mg per os (PO) daily, aspirin 81 mg PO daily, and Tecfidera 240 mg PO twice a day. Tecfidera was started 4 weeks prior to the admission. On physical exam, she was hemodynamically stable except moderately dehydrated. The serum alcohol level and urine drug screen were negative. Her white blood cell count was 8.3 with decreased absolute lymphocyte count 800/
ul(900-2900/UL), amylase 172 U/L (28-100 U/L), lipase levels 1407 U/L (13-60 U/L), triglyceride level 100 mg/dl (<150 mg/dl), calcium level 9.4 mg/dl (8.6-10.2). The magnetic resonance cholangiopancreatography (MRCP) turned out to be negative for cholecdocholithiasis. She was treated as acute pancreatitis, with nothing by mouth (NPO) and intravenous fluids. Tecfidera was discontinued since Day 1 of admission. Her symptom improved and she was begun on oral diet on day 2. The lipase and amylase level decreased to normal range gradually after three days. She was discharged on day 4 being symptom free. In this case, patient developed acute pancreatitis 4 weeks after initiating Tecfidera, without any other known precipitating causes for acute pancreatitis such as biliary tract diseases, alcohol, trauma, hyperlipidemia, hypercalcemia or medications as literature reported. We postulate that Tecfidera induced her acute pancreatitis. The mechanism of acute pancreatitis may be related to its immunosuppressive activity. Since gastrointestinal-related symptoms are common side effects of Tecfidera, acute pancreatitis is easily masked and misdiagnosed. Importantly, since Tecfidera is a newly-developed oral medication, further clinical study is imperative to identify whether these side effects are dose dependent or formulation related.

131. Clinical Vignette
Luo, Hongxiu
Geeta Tadepalli, Abdul Mahmad, Mark Niemiera, Tenen Mathew
Raritan Bay Medical Center (Abdalla M Yousif)
A Rare Case of Heterotaxy syndrome: Eisenmenger Syndrome with Dextrocardia and Situs Inversus in an Adult

Heterotaxy syndrome is rare with an incidence of 1:10,000 worldwide and is associated with at least 3% of cases of congenital heart defects. This syndrome presents with some degree of visceral malposition and disporphism (within both thorax and abdomen) with broad-spectrum variation. Most patients with Heterotaxy syndrome die in childhood secondary to their congenital heart defects. We present the first case of an adult who presents with an associated Eisenmenger Syndrome with Dextrocardia and Heterotaxy syndrome with polysplenia. A 43-year-old female with a past medical history significant for cyanotic congenital heart disease presented to the emergency room with a two-day history of worsening dyspnea. Due to limited access to health care, she was unable to have appropriate cardiology evaluation despite persistent exertional dyspnea. Due to limited access to health care, she was unable to have appropriate cardiology evaluation despite persistent exertional dyspnea. She denied smoking, drinking, or illicit drug use. She denied any family history regarding congenital heart disease and was on no medication. On admission, she was hemodynamically stable except hypoxia. Her pulse oxygen saturation was 76% on room air, and only improved to 80% with 100% oxygen via a non-rebreather mask. On her physical examination, she has obvious clubbing and cyanotic extremities, bilateral diffuse wheezes along with basal rales on lung auscultation, and dextrocardia with a 3/6-holosystolic murmur on the right lower sternal border. Laboratory investigations were as follows: Hemoglobin – 20.9 g/dL, Hematocrit – 65.8%, WBC-count – 5.5 k/µL, Platelets – 70 k/µL, Sodium – 138 mmol/L, Potassium – 3.8 mmol/L, Chloride – 104 mmol/L, CO2 – 23 mmol/L, BUN – 16 mg/dL, Creatinine – 0.7 mg/dL, Glucose – 84 mg/dL. BNP was 1306 pg/mL (normal range: < 450 pg/mL in those <50 year of age). Her chest x-ray showed dextrocardia with evidence of cardiomegaly and mild pulmonary vascular congestion. Her echocardiogram (using left-sided chest leads) showed dextrocardia with a dilated left ventricle, a large ventricular septal defect (VSD) with left-to-right shunting. A CT-angiogram of the chest showed bilateral bi-lobed lung with both main stem hyparterial bronchi, left-sided dominant liver, right-sided stomach and right-sided polysplenia. The patient was diagnosed with Heterotaxy syndrome with dextrocardia and situs inversus, polysplenia-subtype, and Eisenmenger Syndrome. She was treated with supportive care and awaiting for surgical interventions for the large VSD. This case illustrates the rare concurrence of Eisenmenger Syndrome with Dextrocardia associated with Heterotaxy syndrome, polysplenia type. Heterotaxy syndrome has complex variation in clinic presentation and is associated with multiple thoracic and abdominal organ disposition and dysfunction. It is crucial for clinicians to recognize this and conduct a thorough investigation for the possible associated severe multi-organ abnormalities in Heterotaxy syndrome.

132. Clinical Vignette
Luo, Hongxiu
Rafay Khan, Abdalla Yousif
Raritan Bay Medical
May-Thurner Syndrome: more than anticoagulation to treat deep venous thrombosis
May-Thurner Syndrome is a rare condition in which the left common iliac vein is compressed by an overriding right common iliac artery, thus increasing the risk of deep venous thrombosis (DVT) in the left lower extremity. Without correction of this mechanical compression, patients will be at continued risk for recurrent DVT and post thrombotic syndrome despite anticoagulation. Although this anatomic variant is seen in 22% of the population, it remains an under-recognized cause of left iliofemoral DVT. We present a case of a 34 year-old African male with no significant past medical history who complained of left lower extremity swelling and pain. He denied any chest pain or dyspnea. The patient lacked any risk factors for DVT including recent surgery, smoking, prolonged immobilization, extended travel or previous thrombotic disease. The venous Doppler revealed DVT in the left lower extremity, extending from the left popliteal vein to the left common femoral vein. The further hypercoagulable workup, including protein C, protein S, antithrombin deficiency, anti-phospholipid antibody, lupus anticoagulant antibody, factor V Leiden, prothrombin gene mutation, and hyperhomocysteinemia, were found to be negative. The patient was initiated on oral Coumadin therapy. Although the patient’s INR was therapeutic, with a range of 2-6, the swelling of his left lower extremity continued to get worse even after 3 months of treatment. CT of the abdomen with intravenous contrast demonstrated the left common iliac vein being compressed by the right common iliac artery against the vertebral body. This level of compression resulted in complete thrombosis of the involved vein. At this point he was treated with pharmacomechanical thrombectomy, waiting for the stent to prevent recurrent DVT and possible pulmonary embolism. In our case, the idiopathic and unprovoked nature of the patient’s left lower extremity DVT was found to be caused by an anatomic abnormality, rather than a hypercoagulable etiology which would have resolved with empiric anticoagulation therapy. It is important for clinicians to recognize May-Thurner Syndrome as a possible underlying cause for DVT in left lower extremity and provide prompt endovascular management to prevent the recurrence and possible complication of left lower extremity extensive DVT.

An Unusual Cause of Acute Stridor in an Adult Patient

INTRODUCTION: Squamous Cell Papilloma (Recurrent Respiratory Papillomatosis) typically identified in infancy or childhood which usually presents as hoarseness or stridor and is characterized by extensive involvement of the larynx. It can have frequent recurrences and associated maternal genital warts. However, in adults respiratory papillomatosis is a rare medical condition. The major risk factors for head and neck cancers are smoking, smokeless tobacco use, alcohol consumption, human papillomavirus (HPV) infection (especially for oropharyngeal cancers), and Epstein-Barr virus (EBV) infection. Here we report an unusual cause of stridor in an adult patient who was diagnosed with HIV infection.

CASE PRESENTATION: A 49-year-old Caucasian male with past medical history significant for hypertension, COPD, asthma on steroid inhaler, HIV, pulmonary embolism, and a long standing history of hoarseness of voice presented to the ER with acute shortness of breath. On examination his blood pressure was 169/96 mm hg, pulse 117 beats per minute, respiratory rate 28 per minute, temperature 97.6 F, with an Oxygen saturation of 91% on 2 liters/min nasal cannula. Lung examination revealed bilateral wheezes and stridor. Further examination showed a distended abdomen with negative fluid thrill and evidences of pink striae. Laboratory studies showed: Hemoglobin 13.7 gm/dl, hematocrit 41 %, WBC 8,800 cells per cubic millimeter of blood, Platelets 158,000 cells per cubic millimeter of blood, ABG showed a pH of 7.38, PCO2 60mmhg, P-O2 76, Glucose 136 mg/dl, BUN 33 mg/dl, creatinine 0.7 mg/dl, sodium 140 mmol/L, potassium 4.2mmol/L, chloride 98mmol/L, bicarbonate 35.5 mmol/L. His CD4 was 27, with an undetectable viral load. Due to patient’s level of respiratory distress intubation was required. However it failed and tracheostomy was done along with direct Laryngoscopy which revealed a hard laryngeal mass. Vocal cord biopsy showed Squamous Cell Papilloma
which was partially resected. Subsequent pathology report was found to be HPV related with a diffusely positive P16.

**DISCUSSION:** Along with oropharyngeal cancers, other clinical sequela of HPV infection include Cervical, vulvar, vaginal, and anal cancer. HPV DNA detection in oropharyngeal cancers varies from 13% to 56%. Dysphonia (hoarse voice) is a common complaint among users of inhaled glucocorticoids (ICS) as in our patient. However there is an approximately two- to threefold increase in the incidence of Squamous Cell Carcinoma of the head and neck for individuals infected with HIV. It is thus imperative to keep a high level of suspicion for HPV related Squamous Cell Papilloma in patients presenting with hoarseness of voice with or without shortness of breath. It is equally important to consider laryngoscopy screening for HIV patients who present with such atypical symptoms.

**INTRODUCTION:** Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) has mortality rate of 10-20%. It affects multiple organs including liver, heart, lung, pancreas, and skin. We present two cases where DRESS was highly suspected but with opposite outcomes.

**CASE PRESENTATIONS:** Case 1: A 45 year old female with history of asthma presented to ED for flu like symptoms and right neck swelling. She was given Levofloxacin and Amoxicillin, both resulted in severe reactions: upper body rash, perioral and tongue swelling, dysphagia, corneal icterus, cervical adenopathy, RUC pain, diarrhea, finger and foot edema. MRTIs showed myositis in upper body. Tests showed eosinophilia, elevated IgE, EBV IgG and PCR, Parvovirus IgM/IgG, CPK and aldolase. The symptoms improved markedly, leukocytosis and eosinophilia resolved after prednisone. Case 2: A 45 year old male presented with extensive pruritic macular rash with scale and eruption, facial and pedal edema. Tests showed mild LFT elevation, INR 1.5, Albumin 1.5, creatinine 0.8, Eosinophil 6%, IgE 502, LVEF >55%. Home medication hydralazine was discontinued for possible drug reaction. Seven months later, patient was readmitted for anasarca, corneal icterus, tense ascites, skin rashes. Patient was again found on hydralazine. The test results showed mild LFT elevation, creatinine 2.2, INR 2.2, Albumin 0.8, IgE 502, Eosinophil 9%, pro-BNP 50,429, LVEF 30%-35%. He also had atrophic pancreas, superior mesenteric and portal thrombosis. The symptoms deteriorated, patient expired eventually due to multiple organ failure.

**DISCUSSION:** Here we presented two cases of highly suspected DRESS with opposite outcomes. Antibiotics and hydralazine might be the culprits in case 1 and 2, respectively. Both cases had elevated eosinophil and IgE levels, which might be seen in a variety of differentials, such as parasite infection, Kimura’s Disease/angiolymphoid hyperplasia with eosinophilia, Churg–Strauss syndrome, Eosinophilic cellulitis, etc. Based on the temporal relationship between symptom onset and medications and certain diagnostic tests, DRESS was strongly suspected in both cases. If diagnose made and appropriate treatment started early as in case 1, the symptoms and reaction might be well controlled. If a delay in diagnosis and treatment occur as in case 2, multiple organ failure might occur and result in death. In addition to maintain a high vigilance, a meticulous history and physical examination, especially the medication list can not be overestimated in the process of making a correct diagnosis. Subsequently, corticosteroids might help arrest the clinical progression, reverse the organ damage, and could be lifesaving.
Chiari networks are remnants of embryologic structures whose incomplete involution creates a net-like membrane in the right atrium. When discovered it is normally of little clinical significance and is found in 2% to 10% of the population. Nevertheless, there are reported cases of this formation having causation in atrial arrhythmias, thrombus formation, tumor development, tricuspid regurgitation, and embolus and catheter entrapment/entanglement during invasive procedures. We present a case of a patient with an incidental finding of Chiari network on echocardiography and discuss the importance of detecting these formations before certain cardiac procedures. A 92 year-old female was admitted to the hospital for congestive heart failure decompensation. She has a past medical history of type II diabetes mellitus, hypertension, coronary artery disease with stent placement, and peripheral vascular disease with femoral artery embolectomy, history of DVT, pace maker placement, atrial fibrillation, seizures, and dementia. Upon initial examination her blood pressure 116/60, pulse 90 and irregular, respirations is 16, O2 saturation of 95% and was afebrile. Echocardiogram demonstrated an abnormal whip-like structure in the right atrium consistent with a Chiari network. Additional echo findings were mildly calcified aortic valve, and severe pulmonary hypertension that was estimated to be >60 mmHg. As this patient was identified as a candidate for cardiac catherization (right and left), we were subsequently compelled to inform the cardiac team of the prominent presence of the network. To date there is no surgical indication to remove an asymptomatic Chiari network. Previous case reports have described massive right atrial thrombus formed on a Chiari network, leading to intermittent tricuspid obstruction. In one report thrombus had formed after the use of a Hickman catheter for the administration of cytotoxic chemotherapy. Another Case reports cardiac catheterization in a 51-year-old man with secundum atrial septal defect, and catheter entrapment within the area of the inter-atrial defect. At thoracotomy, the catheter was found to be trapped by and looped within a Chiari network involving the septal defect, a hitherto unreported complication of cardiac catheterization. A guide wire having been entrapped by the Chiari network prior to an intended radiofrequency ablation procedure has also been reported. Central venous catheters (especially right atrial catheters), right heart pressure catheters, ablation catheters, and electrophysiological study catheters are used frequently utilized in the hospital setting. Procedures deploying these devices present their own set of risks without anatomic abnormalities. When considering inserting and removing guide wires, sheaths, and other parts of intra-cardiac catheters, the threat of getting entangled within the varied construct of a Chiari network is high. Therefore, careful pre - procedure echocardiographic evaluation is paramount in mitigating potential adverse effects of cardiac - based procedures which may occur as a result of this not uncommon finding.

INH resistant TB in US born patients, a rare but dangerous emergence of home grown resistant TB

According to Centers for Disease control (CDC) a total of 10528 TB cases were diagnosed in the United States. Of these cases 3981 (38%) were patients born in the United States. Of these cases, only 124 in the same year were found to have drug resistant strains, of which only one was born in the United States. We present a 48-year-old female with a past medical history of asthma, hypertension, who was admitted to the hospital after outpatient bronchoscopy culture was positive for M. tuberculosis. Several weeks ago an outpatient chest x-ray and a subsequent CAT scan of the chest showed a hilar mass. She underwent bronchoscopy, and cultures came positive for M tuberculosis. On Admission, Infectious disease consultation was requested for further care. She denied any fevers, chills, nausea, vomiting, diarrhea, dysuria, hemoptysis, night sweats or weight loss. She denied any known contacts with tuberculosis and gave no history of travel outside the country. She denied any HIV related sick behaviour and was tested HIV negative a year ago. She was never incarcerated and was never homeless. She does not smoke. On Examination, vital signs were normal. She was nontoxic, and was in no acute distress. Lungs were clear to auscultation bilaterally. Abdomen was soft,
no organomegaly or any lymphadenopathy appreciated. Patient was started on four drug anti-tuberculosis therapy. Culture results confirmed mycobacterium tuberculosis resistant to isoniazid. The presence of TB cases in the United States is by no means a thing of the past, and despite increasing efforts towards its eradication its prevalence has only seen minor decrease over the last 10 years. TB case rates vary by well recognized factors such as age, race and ethnicity, and country of origin and the increasing immigrant population remains a very important source of infection. However, incidences of drug resistant TB have been very limited, and is usually diagnosed in patients who were not born in the United States. Other risk factors that increase the risk of this disease are incarceration to prisons, homelessness, as well as history of travel. It is of particular interest to find cases like the one we presented here that exhibited "home grown" INH resistant TB, with none of the risk factors mentioned above. It’s imperative to have a heightened awareness of these emerging cases. We aim to alert clinicians to their presence. Foreign-born individuals have accounted for the majority of TB cases in the United States every year since 2001. But we should not discount TB, and even drug resistant strains of TB in US born patients. More research is at this point necessary to probe beyond established cases of resistant TB.

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<th>Federico Sanabria, BS, Mohammed A. Islam, MD, FACP</th>
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<td><strong>INTRODUCTION:</strong> According to the American Diabetes Association, Diabetic Ketoacidosis (DKA) is defined as a patient presenting with a plasma glucose greater than 250 mg/dl, arterial pH less than 7.3, serum bicarbonate less than 18 mEq/L, positive urine or serum ketones, and an anion gap greater than 10. DKA is a serious condition that can lead to many complications including coma and death. Thought rare, DKA has been reported with blood glucose levels less than 180 mg/dl. We report a rare case of a patient with euglycemic ketoacidosis precipitated by starvation and alcohol intoxication that highlights the need for careful assessment of diabetic patients despite normal glucose levels.</td>
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<td><strong>CASE REPORT:</strong> A 59-year-old male with a past medical history of diabetes, coronary artery disease, and hypertension presented to the emergency room with a three day history of shoulder pain following a fall, and a two day history of coffee ground emesis. Patient stated that he had been drinking vodka for three days to relieve his shoulder pain. Patient was initially admitted to the ICU for an upper GI bleed to rule out esophageal varices or Mallory-Weiss tears secondary to alcohol intoxication. Labs demonstrated a drop in Hgb from 10.4 to 8.3, an anion gap of 28, lactic acid 1.3, betahydroxybutyrate greater than 8 mmol/L and an arterial pH of 7.32. Ketones were also found in urine. IV insulin was started despite his continued euglycemic status, which caused improvement biochemically and clinically. Patient received thiamine, B12, folic acid and IV fluids as well as blood transfusion for his anemia.</td>
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<td><strong>DISCUSSION:</strong> Although the mortality rate associated with DKA has decreased significantly over the years, DKA remains responsible for a significant amount of hospital admissions for patients with diabetes. Eughlycemic DKA is a very rare variation of DKA occurring in about 1% of patients that fit the criteria for DKA. In order to receive a diagnosis of euglycemic DKA, blood glucose levels should be less than 180 mg/dL. Euglycemic DKA has been observed in pregnant patients, fasting or starving patients, alcoholic patients and patients with glycogen storage disorders. It is believed that diabetic patients that have been fasting, and still receiving insulin, will form ketones while maintaining euglycemia. These patients will become acidic and can present with DKA with minimal to no elevation of blood glucose. The case report thus raises the awareness and highlights the absolute necessity to be cognizant that normal or near normal glucose levels should not discourage the use of IV insulin in DKA.</td>
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### A rare case of transient ST-segment elevation on ECG during stress test

**INTRODUCTION:** While atherosclerotic plaque and induced stenosis in the coronary artery develop over a period of time, rupture of a vulnerable plaque can lead to sudden thrombosis formation, blockage of blood perfusion and subsequent myocardial ischemia and necrosis relatively fast. Often the stress test could induce the transient ST segment depression due to external myocardial ischemia. Here we report a rare case, in which, with the typical transient ST-segment elevation during stress test, 50% stenosis of coronary artery was revealed from the first cardiac catheterization and 99% stenosis from the second one 3 months later with eventual stent placement.

**CASE:** A 65 year old male with past medical history of hypertension and diabetes, was referred to the clinic because of exertional chest pain with associated palpitation. He denied syncope or shortness of breath. He had no significant family history or personal history of smoking, alcohol or drugs. His medications included Metoprolol, Lisinopril, Aspirin and Metformin. His physical examination including vital signs were all normal. All blood tests were within normal range. ECG showed normal sinus rhythm while Echocardiogram showed an ejection fraction of 55% with mild diastolic dysfunction. When patient took the first stress test, he developed chest pain and significant transient ST elevation on ECG, lasting about 3 minutes that caused a premature termination of the test. Patient’s cardiac catheterization showed the coronary artery (anterior ascending artery) with 50% stenosis. Patient was asymptomatic with no active issues and was subsequently discharged home with continued medication treatment. Three months later, patient came back with similar chest pain. Intriguingly, a repeat stress test stress test evoked similar symptoms and ECG changes. The second cardiac catheterization showed 99% stenosis this time. The stents were placed and patient experienced no chest pain thereafter. The third stress test was normal 2 months later.

**DISCUSSION:** In this case, the transient rapid ST-segment elevation likely resulted from the severe exertional ischemia due to transient coronary artery spasm. Another explanation is that the plaque rupture induced thrombosis temporarily completely occluded the blood flow and then dissolved spontaneously by the thrombolytic systems in the body. The increased severity of the coronary artery stenosis from 50% to 99% in three months probably resulted from the dynamic rupture of the vulnerable plaque and thrombosis formation. This thus raises concern and beckons further research about the necessity of stent placement in early stenosis for best management and prevention of an acute coronary syndrome.

### Superior mesenteric artery syndrome in a patient on chronic use of morphine

**INTRODUCTION:** Superior mesenteric artery (SMA) syndrome is a rare cause of upper gastrointestinal obstruction due to the duodenum compression with the SMA and aorta. Early recognition, institution of the appropriate conservative measures and timely selection of a definite surgical method are critical to prevent severe complications and death. There have been no previous report about SMA syndrome on patient with use of opioid and here we report a case with SMA syndrome on chronic use of morphine over three years.

**CASE:** Patient was a 64-year-old man with past medical history of hypertension and chronic back pain, who was sent to hospital with chief complaint of abdominal pain, nausea and vomiting with constipation and shortness of breath for one day. No abdominal
surgery history before. He was using morphine for more than 3 years. No history of smoking, alcohol or drugs. Physical examination: Bp: 134/100, HR: 102, RR: 23, T: 97.5 SaO2: 100% on non-breathing mask. He looked sick and cachectic. There were crackles on the bilateral lungs, Heart rhythm and rate regular with sinus tachycardia. Abdomen was distended and tender with diminished bowel sounds. Patient was alert and awake. Labs: Hb: 13.8, Hct: 41.4, WBC: 9000, Band: 23%, Plt: 284, Lactic acid: 9.9, HCO3 18, ABG: pH 7.28, PCO2: 30, PO2 59. CXR: pneumonia. CT on abdomen showed Massively dilated stomach and proximal duodenum, with transition point in the third segment of the duodenum near where it crossed underneath the SMA. Patient was immediately admitted to the critical care unit and treated with antibiotics and intravenous fluid. Nasal gastric tube was inserted with suction of three liters of fluids from the stomach. The endoscopy showed patient had gastritis while UPPER GI SERIES showed duodenal obstruction. Later patient improved and could take diets normally.

**DISCUSSION:** Normally the third part of the duodenum is suspended by the Treitz ligament in the angle between 28° and 65° between the aorta and the SMA on CT. In SMA syndrome, the angle could be below 25° with decreased space between SMA and the aorta finally leading to the third part of duodenum compression. In our case, the angle is less than 25°. Our patient had poor appetite for a long time due to chronic use of morphine with emaciated-looking and BMI of 17.7 kg/m². The poor appetite led to malnutrition and loss of fat tissue in the whole body, esp. in the mesenteric fat, which led to the narrowed angle between SMA and aorta. Treatment included the medical conservative treatment and surgery. In our case, after suction and antibiotics for pneumonia, patient improved and could take meal normally and continue the medical treatment.

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**INTRODUCTION:** Vitamin B12 deficiency is estimated to affect 10%-15% of people over the age of 60. Besides malabsorption and pernicious anemia, prolonged usage of certain medications have been linked with Vitamin B12 deficiency.

**CASE PRESENTATION:** 65 year old female patient with PMHx of HTN and hyperlipidemia presented to ER for one syncope episode. She denied head trauma, no headache, no vision or hearing changes, no palpitation, no weakness or numbness, no abdominal discomfort, no diarrhea or weight loss. She eats balanced diet all her life and she was on HCTZ-candesartan and atenolol for hypertension, and pravastatin was started for hyperlipidemia 10 months ago. Upon admission, her vitals were HR 70, BP 106/53, RR 16, O2 saturation 100% on room air, temperature 99.0. Physical examination including neurological examination was within normal limits. Lab values: Hb 13 g/dl, and MCV 103 fl, compared to Hb 14.2 g/dl and MCV 90.2 fl 10 months ago, Vitamin B12 88 pg/ml, and folic acid was 17.1 ng/ml, homocystine was 27.9 µmol/L, and methylmalonic acid was 1541 nmol/l. Further investigation including MRI, carotid ultrasound, electrolytes and EEG were re-assuring. Vitamin B12 deficiency was diagnosed and pt was prescribed with PO Vitamin B12 and discharged home.

**DISCUSSION:** Prevalence of vitamin deficiencies increases with age. Prolonged usage of medications such as metformin and H2 blocker have been shown to associated with Vitamin B12 deficiency. One previous study has shown a correlation of simvastatin with vitamin B12 deficiency. In the past, screening for vitamin B-12 deficiency was indicated only for the evaluation of those with relevant symptoms and signs, such as anemia, neuropathy, or cognitive impairment. However, elderly people who have Vitamin B12 deficiency frequently lack the classical signs and symptoms. Statins are widely prescribe as a long term therapy for hyperlipidemia and our case and other studies have shown possible correlations of acquired Vitamin B12 deficiency with chronic statins’ usage.
Therefore, it seems reasonable to draw baseline CBC and/or Vitamin B12 level and monitor vitamin B12 levels periodically in patients taking stains.

**Case Presentation:**

The patient was a 21-year-old female with borderline personality disorder who had initiated lamotrigine 25 mg daily six weeks prior to admission and was appropriately titrated up to 50 mg daily three weeks later. She began having high-grade fevers up to 105°F with myalgias and fatigue ten days prior to admission and subsequently developed an extensive rash across her face and body. On admission, she had a temperature of 103.3°F and a blood pressure of 67/44. Physical exam revealed a confluent, morbilliform, erythematous rash with desquamation affecting her face, torso, back, and all extremities, and cervical and inguinal lymphadenopathy. Initial laboratory findings included a white blood cell count of 23,200 with 13% eosinophils, AST 818, ALT 670, and alkaline phosphatase 208. She had discontinued her lamotrigine on onset of symptoms and it was not restarted. Given the concern for severe sepsis, the patient was volume resuscitated and started on broad-spectrum antibiotics. However, blood and urine cultures were negative and blood pressure improved after fluid administration. Antibiotics were promptly discontinued after a diagnosis of drug reaction with eosinophilia and systemic symptoms (DRESS) syndrome was made. The patient was evaluated by dermatology and prescribed hydrocortisone topical cream. However, given worsening liver function tests and persistent symptoms, she was started on prednisone 1 mg/kg/day, resulting in dramatic improvement in liver function abnormalities and near complete resolution of her rash.

**Discussion:**

DRESS is a rare and potentially deadly syndrome that has been estimated to occur in 1 in 1000 to 1 in 10,000 exposures to many frequently prescribed medications, including antiepileptics, sulfonamides, and allopurinol. Reported mortality is up to 10 percent, most commonly in severe hepatic involvement, which was of concern with our patient. The diagnosis can be made using the RegiSCAR scoring system, which requires hospitalization for acute rash in the setting of an associated medication, with at least three of the following additional features: (1) fever, (2) lymphadenopathy, (3) visceral organ involvement, and (4) hematologic abnormalities, including eosinophilia. This unusual case of DRESS syndrome highlights several important issues in clinical practice. First, hypotension is not commonly seen in DRESS syndrome and, consequently, the patient appeared to have severe sepsis on admission despite meeting RegiSCAR diagnostic criteria. Thus, clinicians should know that DRESS can masquerade as severe sepsis and be able to recognize when it does so. Furthermore, lamotrigine has been established as a cause of DRESS syndrome when it is initiated at high doses or titrated up in sooner than two week intervals, but our patient’s medication had been properly managed. This case emphasizes that the prompt diagnosis and treatment of DRESS syndrome relies on the appropriate index of suspicion in the correct clinical setting, even with uncommon presentations.

**Case Presentation:**

A thorn puncture from a rose bush is common, especially in patients who work in outdoor gardening. A healthy 49 year old man developed Alternaria osteomyelitis as a complication of thorn penetration into the radial aspect of the second MCP (MCP) joint. He was successfully treated with joint debridement and irrigation and a prolonged course of itraconazole. In the last two decades, fungal infections have become an important cause of morbidity and mortality in humans, especially in immunocompromised patients. We report a healthy 49yr old man with osteomyelitis of the second MCP joint caused by the dematiaceous fungus...
Alternaria. This infection was acquired via thorn penetration into the radial aspect of the 2nd MCP joint while gardening. To our knowledge, this is the first reported case of Alternaria osteomyelitis as a septic joint in a healthy adult male.

143 59 Clinical Vignette Schwartz, Mark Sabiha Hussain, MD, Amanda Kaveney, MD Rutgers - New Brunswick (Ranita Sharma)

Essential Mixed Cryoglobulinemia Presenting As A Fever Of Unknown Origin In A Middle-Aged Woman

Fever of unknown origin (FUO) is defined as a temperature greater than 38.3 degrees Celsius for at least three weeks duration and no established diagnosis after a one-week workup(1). The workup of FUO includes a thorough history, physical, laboratory studies, and imaging. The most common causes of FUO include infections, malignancies, and connective tissue disorders. Essential Mixed Cryoglobulinemia (Essential CG) is a vasculitis of the small and medium vessels secondary to deposition of cryoglobulins, which are polyclonal IgG and IgM immune complexes that precipitate at low temperatures. Essential CG can be associated with viral illnesses, hematologic malignancies, and rheumatologic diseases. The gold standard for diagnosis is a positive serum cryocrit. Management depends on the severity of disease and includes treating the underlying cause, plasma exchange, and immunomodulators (2,3,4). A 47 year-old female with history of hypertension, type 2 diabetes, asthma, and non-alcoholic liver disease presented with 3-week history of fever and erythematous rash of the head and trunk. Workup at another hospital prior to admission was significant for negative blood cultures and mediastinal adenopathy on CT chest. She was treated with a seven day course of antibiotics with no improvement. On presentation to our hospital, significant findings included blood pressure of 79/48, pulse of 120, fever of 39 degrees Celsius, and altered mental status. Labs were pertinent for leukocytosis, anemia, hyponatremia, transaminitis, lactic acidosis, and elevated inflammatory markers (ESR and CRP). Blood and urine cultures were negative. She was admitted to the ICU and remained febrile despite treatment with broad spectrum antibiotics. She was placed on a cooling blanket and developed a diffuse purple macular papular rash. Additional infectious workup including HIV, hepatitis, cytomegalovirus (CMV), Epstein Barr virus (EBV), adenovirus, human herpesvirus 8 (HHV-8), and tick-born illnesses was negative. Bone marrow biopsy showed pancytopenia without evidence of malignancy. The patient was found to have positive serum cryocrit and immunofixation suggestive of cryoglobulinemia. The diagnosis of Essential CG was made and the patient was treated with Methylprednisolone, plasmapheresis, and Rituximab. The patient developed ARDS, acute renal failure, and DIC. The patient passed away from a large intracerebral hemorrhage. This case illustrates the extensive workup a primary care physician should perform on patients presenting with FUO and a review of Essential CG. When a patient remains febrile on broad spectrum antibiotics, less common causes of FUO should be considered. Patients diagnosed with Essential CG should be evaluated for other infectious and rheumatologic diseases. In patient’s experiencing arthralgias or fatigue, the treatment involves non-steroidal anti-inflammatory drugs. When evidence of end organ damage is present, steroids and immunomodulators are indicated. In severe cases where hyper viscosity syndrome is present, treatment involves plasma exchange.

144 114 Clinical Vignette Sedhom, Ramy Ranita Sharma Rutgers - New Brunswick (Ranita Sharma)

The Importance of Patient Education and Communication at Hospital Discharge: A Case of Rapid Recognition and Readmission for Guillain-Barre Syndrome as a Result of the Teach-Back Method

Physician-patient communication is fundamental to the practice of medicine and is especially important at discharge. Several studies have demonstrated disconnect between physician information giving and patient understanding. Despite the recognized importance of a comprehensive discharge summary, it often is not given to patients in a timely manner nor communicated at their literacy level. Thus, absent is critical information about diagnosis, complications, medications, test results and follow-up. We report a case of Guillain-Barre Syndrome (GBS) where effective communication and education at discharge provided for early recognition and timely
intervention for a well established sequel of Campylobacteriosis. A 65-year old female with no past medical history was admitted with 4 days of cramping abdominal pain, non-bloody diarrhea (10 episodes/day) and fever (101.5). She denied antibiotic use, sick contacts, travel, nausea, vomiting, hematochezia, or melena. She lived alone and denied smoking, alcohol, or illicit drug use. At presentation, she was afebrile and not hypotensive or orthostatic. Physical exam revealed dry mucus membranes, hyperactive bowel sounds, and marked tenderness to palpation in the RUQ. Murphy’s sign was negative. No evidence of ascites, hepatosplenomegaly or manifestations of chronic liver disease was noted. Stool occult blood testing was negative. Prerenal AKI was suspected with a creatinine 1.3. CBC and hepatic function panel were normal. CT abdomen revealed mesenteric adenitis and right-sided colitis. C. difficile toxin was negative. A diagnosis of Campylobacter infection was made from stool culture. The patient recovered following a 5-day course of azithromycin. She left with discharge paperwork and educated about manifestations of a rare well-documented association of a neurologic illness characterized by ascending weakness. She was advised to return if such symptoms were noted. One week later, she developed progressive weakness in her distal lower extremities with difficulty ambulating and fine motor weakness in her hands. She returned to the hospital and requested her prior doctors. Preceding campylobacter infection, new ascending paralysis, symmetric areflexia in the lower extremities and CSF albuminocytologic dissociation confirmed the diagnosis of GBS. Sensory symptoms, cranial nerve involvement, respiratory failure or bladder/bowel dysfunction were notably absent. She completed a 5-day course of IVIG with motor function improvement from 2/5 to 4/5 strength in lower extremities at discharge. She regained full function with rehabilitation. The transition from hospital to home is a vulnerable recovery period that requires astute self-monitoring, attention to modified medication regimens, ability to gauge recovery and recognize delayed complications. The absence of reliable social/family support complicates the scenario. Appropriate patient education pre-discharge is the best tool available to minimize adverse events. This case illustrates the importance of health care provider driven education. Using the teach-back method, discharge instructions were effectively communicated. The patient articulated an appropriate understanding of the complications of her diarrheal illness, including GBS.

### Sinus sagittal thrombosis in setting of Graves’ disease and recent radioactive iodine ablation

**CASE:** Cerebral venous thrombosis (CVT) is a distinct cerebrovascular condition with an annual incidence of 5 cases per 1 million individuals. Typical risk factors for CVT include inherited thrombophilia, pregnancy, oral contraceptives (OCP), vasculitis and malignancy. Hyperthyroidism is rarely recognized as a risk factor for CVT, however it has been shown to be a predisposing factor in 1.7% of cases due to an increased hypercoagulable state. Here we present a woman with Graves’ disease who presented after radioactive iodine ablation in a thyrotoxic state and was found to have straight, transverse and sagittal sinus thrombosis. A 34-year-old women with past medical history of Graves’ disease and migraines, presented with one day of worsening headache, blurred vision, nausea, vomiting, palpitations, and flushing. Headache was described as constant, frontal pain above her eyes bilaterally, which were inconsistent with her normal migraine symptoms. No complaints of neurologic deficits. Patient was recently diagnosed with Graves’ disease and was being managed with propranolol 20mg tid. Five days prior to this admission, patient underwent radioactive iodine ablation, and reported worsening palpitations and flushing since the ablation. Patient takes OCPs, but denies history of drug abuse, no family history of hypercoagulable states or prior history of thromboembolic events or malignancy. On examination, patient was tachycardic; neurological examination was unremarkable with no signs of exophthalmoplegia or goiter. Basic laboratory work up was within normal limits. A CT scan of her head revealed a hyperdensity involving the left transverse and sagittal sinuses. Subsequently an MRV brain was performed which demonstrated thrombosis involving the straight left transverse and sagittal sinuses. Patient was managed with an increased dose of propranolol 40mg tid for thyrotoxic symptoms. She was
started on anticoagulation with enoxaparin injections at a dose of 1mg/kg bid, and transitioned to warfarin for long term anticoagulation. Tests for protein C deficiency, Protein S deficiency, factor V leiden mutation, Antithrombin III deficiency, and von Willebrand Disease all returned negative.

**DISCUSSION:** This patient presented with symptoms of thyrotoxicosis following radioactive iodine ablation for Graves’ disease and a worsening headache that was found to have CVT. Hyperthyroidism has shown to be a predisposing factor for the development of venous thrombosis due to reversible pro-coagulant and anti-fibrinolytic effects. Studies have demonstrated increased serum levels of von Willebrand Factor, fibrinogen, factor VIII, factor IX, antithrombin and plasminogen activator inhibitor levels, and decreased levels of TPA and protein C in hyperthyroid states. This leads to an overall hypercoagulable state, which resolves with treatment of hyperthyroidism. This case is particularly interesting given the patient presented following a radioactive iodine ablation with worsening thyrotoxic symptoms and CVT. The increased hyperthyroid state from ablation likely caused a pro-coagulant state predisposing the patient for this thrombotic event.

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**Quality Improvement**

**Mora, Natalie**

**Rutgers - New Brunswick (Ranita Sharma)**

**The Prediabetes & Diabetes Epidemic in the Hispanic Population: An Implementation Science Model to Improve Outcomes**

**INTRODUCTION:** Prevalence of T2DM among Hispanics is 11.8%. The overall objective was to understand why certain issues hinder implementation of a diabetes education project (In-DEP) in an outpatient urban clinic.

**METHODS:** We conducted a pilot intervention in the University Hospital primary care clinic in Newark, NJ. Eligible criteria included the following: prediabetes, Hispanic, ages 21-65, family history of T2DM, BMI >27 kg/m2. The public health practice basis of this study was to evaluate the process indicators for the initial steps involved in the In-DEP project. Prospective patients were identified through clinic EMRs and daily schedules were used to track prospective subjects. The aim of the In-DEP program was to ultimately decrease cardiovascular complications. The overall timeline of the program included 4 weekly workshops to discuss the AHA’s Simple 7 principles. However, the implementation science objective of this study emphasized the recruitment and enrollment phases of the In-DEP project; workshop assessment was not included in this analysis.

**RESULTS:** Total eligible participants for this study totaled 137 patients from the clinic database. Percentage of approached patients for enrollment in the In-DEP project was 40.1%. From the pool of approached patients, 11 participants were enrolled in the study (18.2%). Of the patients unable to be approached, 20.4% included no show appointments, and 29.1% were patients who were unable to be located in clinic on the day of their appointment. Implementation science analysis illustrates how issues with recruitment and program participation impacted the overall enrollment rate (18.2%). Inability to travel to clinic for the workshop sessions accounted for 14.5% of participation issues, and 32.7% accounted for work schedule conflicts. Enrolled participants were predominantly female (63.6%), and only spoke Spanish (63.6%).

**CONCLUSION:** Some aspects of the In-DEP program can be improved to encourage overall patient recruitment, enrollment, and participation. By evaluating process indicators of the implementation phase of this study, specific methods and strategies to effect changes at this level emerged. Do we need a different model for patient health education programs? Addressing language barriers, minimizing travel for sessions, and capitalizing on the time the patient spends in clinic (e.g. “on the spot” intervention) are potential methods that can correct the lack of participation. Same day intervention would allow for enrolled subjects to participate in
intervention sessions before/after their appointment to limit return visits/travel. The addition of alerts on prediabetic and diabetic medical records is also another potential solution. This method would then allow for the distribution of data to the patient regarding their own health status (e.g. HbA1C, BMI, cholesterol, blood pressure) that they can then review while waiting for their appointment. Identifying and correcting the implementation of this research study is key to improving disease outcomes in a Hispanic, prediabetic population.

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<td>The American Board of Internal Medicine (ABIM) expects trainees to develop competency related to the knowledge and understanding of a limited set of procedures but does not specify a minimum number of procedures needed to demonstrate competency or to satisfy training completion requirements. Graduating residents pursuing procedural based specialties (PBS) rank procedural access and skills as an important aspect of their career decision. There is little information in the current literature related to subspecialty interest among internal medicine residents and its influence on the number of procedures performed by residents. We conducted a retrospective analysis to determine the number of procedures performed by internal medicine residents who began training between 2006-2011. Preliminary year interns and residents who transferred into or out of the program during the study period were excluded. The primary end point was the difference in the total number of procedures performed over 3 years by residents pursuing PBS versus those pursuing NPBS. Invasive procedures were defined as arterial lines, femoral, jugular, and subclavian central line insertions, lumbar punctures, paracentesis, and thoracentesis. 119 residents who began training between 2006-2011 performed an average of 25.4 procedures (SD 20.2) during their three year training. Residents entering PBS (n=29) performed more procedures than residents entering NPBS (n=90) (30.7 vs 23.7, p&lt;0.05). Residents entering Pulmonary/Critical Care Medicine performed the highest number of procedures amongst all residents with an average of 55.7 (statistically significant). Residents entering Cardiology and Gastroenterology performed an average of 23.1 and 23.3 procedures respectively. Residents entering Nephrology, a NPBS, performed an average of 31.9 procedures and those pursuing general medicine (hospital medicine versus outpatient medicine not specified) performed an average of 24.7 procedures. The number of procedures per resident declined by 3.0 procedures per year over the study period with no statistical difference between the PBS and NPBS groups. Procedures performed by men entering PBS declined less than women (0.7 vs 4.7 procedures/year, p&lt;0.01). Procedural skills are recognized as an essential part of internal medicine training. Our findings suggest that residents entering PBS performed more procedures than residents pursuing NPBS. Residents entering Pulmonary/Critical Care Medicine perform the most procedures compared to residents entering other fields. This data correlates with information obtained in post graduation surveys in which residents entering PBS viewed access and ability to perform procedures as an important part of their fellowship decision. We report an overall decline in the total number of procedures performed by residents throughout their three years of training from 2006-2011. Further research is needed to determine whether the ACGME duty hour restrictions, the increasing use of Interventional Radiology for procedures, access to simulation training and/or changes to the ABIM training requirements related to procedural competencies are responsible for these trends.</td>
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<td>BACKGROUND: Since the recognition of the important role the intestinal microbiome is playing in our well being, research of its</td>
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Anne Tyno, Roger Strair Sharma) complex nature is gaining momentum. We performed a pilot study to determine the safety of administering the probiotic lactobacillus GG following hematopoietic engraftment after allogeneic hematopoietic stem cell transplant (HSCT). This retrospective analysis was designed to analyze the effects of the probiotic on the incidence of intestinal GVHD.

METHODS: We evaluated eighty patients who underwent an allogeneic HSCT between 1/3/2006 and 1/31/2010 through our bone marrow transplant registry. Thirty patients were supplemented with the probiotic, and fifty patients were chosen as historic controls. The primary end point was the development of acute GVHD (aGVHD) or chronic GVHD (cGVHD).

RESULTS: Among patients who were treated with probiotics, at the three-month interval, 14 (47%) developed grade I aGVHD, 6 (20%) developed grade II aGVHD, and 10 (33%) were free of disease. At six months, 12 patients (40%) had limited cGVHD, 7 (23%) had extensive cGVHD, and 4 (13%) were unverifiable. For patients who did not receive probiotics, at the three-month interval, 21 (42%) developed grade I aGVHD, 5 (10%) had grade II, and 6 (12%) had grade III or IV aGVHD. Seventeen (34%) patients were free of disease, and 1 (2%) was unknown. At six months, 17 patients (34%) had limited cGVHD, 10 (20%) had extensive cGVHD, and four patients (8%) were unknown or expired. At the 12-month period, 10 patients (20%) had limited cGVHD, 21 (42%) had extensive cGVHD, 15 (30%) had no evidence of disease, and 4 (8%) were unknown or expired.

CONCLUSION: Our findings suggest that supplementing therapy with probiotics may modulate risk of GVHD with the potential for lower rates of grade III- IV aGVHD. This study demonstrates the safety and feasibility of administering probiotics in post-allogeneic stem cell transplant patients.

149. Research Lee, Nara Valerie Francescutti, Joseph Skitzki Rutgers - New Brunswick (Ranita Sharma) Peritoneal Carcinomatosis in Colorectal Cancer: Proposed Algorithm for Cytoreduction and HIPEC

BACKGROUND: As a natural progression of colorectal cancer, transcoelomic spread to the peritoneum result in peritoneal carcinomatosis (PC). Historically, this was approached as generalized disease with systemic therapy, but locoregional approach with cytoreduction and hyperthermic intraperitoneal chemotherapy (HIPEC) has gained wide acceptance. Given the inherent morbidity and mortality, patient selection is paramount, yet challenging due to limited practical clinical tools available. It is our attempt to reduce the confusion and assist in the decision process by providing a treatment algorithm.

METHODS: Published literature in the PubMed database regarding peritoneal carcinomatosis in colorectal cancer was reviewed. Based on the available evidence, algorithm is proposed to aid in patient selection for cytoreduction/HIPEC.

RESULTS: Peritoneal Carcinomatosis Index (PCI) of 20 is widely accepted as the threshold above which cytoreduction/HIPEC should be reconsidered as a treatment option. If PCI < 20 and complete cytoreduction can be achieved through peritoneal stripping, cytoreduction/HIPEC should be considered. If PCI <20, but prospect of complete cytoreduction is unlikely, systemic therapy is recommended with restaging CT afterwards to determine candidacy for cytoreduction/HIPEC. For PCI >805; 20, systemic chemotherapy is preferred over locoregional therapy for achieving complete cytoreduction is more difficult with increased risk of surgical complications. If restaging CT shows favorable response to systemic therapy and complete cytoreduction is probable,
cytoreduction/HIPEC may be considered at that time. If unresponsive or progressive disease on systemic therapy, locoregional therapy is of limited benefit. In synchronous peritoneal metastases with primary tumor, if primary tumor is asymptomatic and PCI < 20, resection of the primary tumor and cytoreduction/HIPEC for PC should be considered with curative intent. If asymptomatic and PCI & #8805; 20, systemic therapy with repeat CT to determine tumor response is recommended. If favorable, primary tumor resection with cytoreduction/HIPEC may be considered. For symptomatic primary tumor, mixed data exists regarding primary tumor resection thus decision should be made on an individual basis. If resection of symptomatic tumor is performed, we suggest systemic therapy with repeat CT to assess response. Based on the treatment response and PCI, cytoreduction/HIPEC may be considered as discussed above.

CONCLUSION: Although the cornerstone of treatment decisions involves a thorough discussion between the multidisciplinary team and the patient, we present treatment algorithms to assist in patient selection for cytoreduction/HIPEC. These algorithms should be used in conjunction with clinical expertise, patient’s comorbidities, performance status and surgical risk stratification.

150. 137
Clinical Vignette DeRose, Joseph
Rutgers - Newark (Neil Kothari)
Human Granulocytic Anaplasmosis in the Setting of a Hunting Injury
Human Granulocytic Anaplasmosis is a traditionally tick borne illness characterized by non-specific symptoms. Most cases occur in spring/summer in the Northeastern and northern Midwest US. 63 year old man with medical history of HIV on ART, presented to E O-VAMC with a complaint of cyclic fevers, chills, and diarrhea. Symptoms began with generalized weakness one week prior to presentation. Patient revealed he was a hunter and experienced trauma to his hand while butchering a deer. Diarrhea began four days prior to presentation with multiple bowel movements associated with lack of appetite, abdominal discomfort and nausea. He denied vomiting, hematemesis, sick contacts, changes in diet, shortness of breath, recent travel or known tick exposure. On physical exam patient was tachycardic, ill-appearing and over short period of time became hemodynamically unstable and was admitted to MICU. Initial labs were significant for thrombocytopenia, coagulopathy and transaminitis. He was fluid resuscitated and Infectious Disease service was consulted. He was started on broad spectrum antibiotics given his recent hunting injury and possible tick exposure. The patient’s clinical status improved throughout his hospital stay and he was discharged on Azithromycin, Atovoquone and Doxycycline. The patient’s clinical status improved throughout his hospital stay and he was discharged on Azithromycin, Atovoquone and Doxycycline. The patient returned to ID clinic two weeks after discharge and reported complete resolution of symptoms. Titers at that time revealed (+) IgM against HGA and re-examination of the peripheral smear revealed intraleukocytic morulae characteristic of Ehrlichia/Anaplasmosis infection. Given patient’s history of trauma while butchering a deer, timeline of symptoms and positive Anaplasmosis serology, diagnosis of HGA with transmission via handling of infected butchered deer meat was made. This case illustrates an uncommon mode of HGA transmission and emphasizes value of obtaining thorough history. Although the mode of transmission in this case is rare, the disease is common to the northeast US and recognition, prompt treatment is necessary to prevent complications.

151. 215
Clinical Vignette DeRose, Joseph
Rutgers - Newark (Neil Kothari)
CUTANEOUS HANSEN’S DISEASE IN A YOUNG HIV POSITIVE GUYANESE WOMAN
Hansen’s disease (AKA Leprosy) is a chronic bacterial infection caused by Mycobacterium lepromatosis traditionally involving the peripheral nerves and skin. While it continues to manifest as a common disease in many developing countries, cases diagnosed in the United States remain rare. 19 year old Guyanese woman with a past medical history of AIDS, HPV and latent tuberculosis
presented to Rutgers-NJMS infectious disease clinic with complaints of skin lesions on her upper and lower extremities for two weeks. She reported having these same lesions in the past which had spontaneously resolved over a period of three weeks, leaving residual hyperpigmentation. She was recently restarted on antiviral treatment after a period of medication non-compliance. The lesions were tender, pruritic and dark with well demarcated but irregular borders. They involved the arms and legs but the joints were spared. They were not associated with any swelling or lesions elsewhere. She associated arthralgia, myalgia and subjective fevers for one month preceding the lesions. She denied night sweats, chills, weight loss, cough, shortness of breath, abdominal pain, diarrhea, recent travel, exposure to sick contacts or insect bites, recent hospitalizations or history of opportunistic infections. On physical examination, the patient was noted to have two indurated erythematous nodules measuring 4mm-1cm on the right arm and multiple hyper-pigmented tender macules with scaling noted on bilateral anterior shins. Given the patient’s history of latent TB, a PET scan was performed to rule out occult TB infection which was found to be negative. She was subsequently referred to dermatology for a skin biopsy of her lesions which revealed cutaneous mycobacterium infection. The sample was referred to the National Hansen’s Disease Program who identified active lepromatous Hansen’s disease in the tissue. The patient was treated with Dapsone, Clotazimine and Thalidomide however she developed severe side effects to the thalidomide and it was discontinued. The patient continued to have painful skin lesions at the time of last contact with the Rutgers-NJMS clinic. This case illustrates a case of immune reconstitution following initiation of ART manifesting as active lepromatous Hansen’s disease. It is important to note that despite commonly held beliefs that Hansen’s disease is highly contagious and devastating, prompt recognition and treatment leads to favorable outcomes in many patients.

152. 98 Clinical Vignette Duffy, Margaret Mandeep Samra, MD, John Kern, MD, Neil Kothing, MD, Alluru Reddi, MD Rutgers - Newark (Neil Kothing) Polycystic Kidney Disease- An Incidental Finding in a patient who presented with subarachnoid hemorrhage and renal failure A 54 year-old man with history of chronic kidney disease (baseline Cr 2-3), previous arteriovenous fistula placement, uncontrolled hypertension, and polysubstance abuse (cocaine, heroine) presented to an outside hospital with acute-onset headache and dizziness over the course of 24 hours. A CT of the head revealed acute subarachnoid hemorrhage for which he was given a loading dose of phosphenytoin and transferred to a university center with neurosurgical capability. Upon presentation he denied fevers, chills, loss of consciousness, changes in vision, chest pain, shortness of breath, abdominal pain, nausea, vomiting, decreased urination, hematuria, or swelling in his lower extremities. He did admit to recent cocaine use one day before presentation. In the emergency room he was afibrile, BP 171/114mmHg, HR 82 bpm, RR 14, and O2 sat 99% room air. In the emergency room he was afibrile, BP 171/114mmHg, HR 82 bpm, RR 14, and O2 sat 99% room air. On physical exam he appeared lethargic, had a palpable left-sided AV fistula with thrill, a notable left renal bruit and palpable kidneys on exam. Lab work was notable for Na 133, K 4.0, Cl 98, HCO3 19, BUN/CR 67/6.5 (calculated GFR 11), glucose 164, WBC 15.6 (95% neutrophils), Hgb 13.8 (baseline 12), plt 150, and albumin 3.4. Admission urinalysis showed specific gravity 1.013, pH 6.0, protein >500mg/dL, no leukocyte esterase or nitrates, and 3 RBC. Urine toxicology was positive for cocaine and opiates. A CT without contrast revealed acute subarachnoid hemorrhage of the left frontal, temporal, and parietal lobes without evidence of midline shift or hydrocephalus. Neurosurgery was consulted and ordered a CT angiogram. Findings revealed a small 3x2 mm aneurysm arising from the left middle cerebral artery (MCA) bifurcation with small lobulations, which were highly suggestive of site of rupture. The patient underwent a successful emergent craniotomy, endovascular clipping of his ruptured aneurysm, and hemodialysis. In order to further characterize his renal function and physical findings a renal ultrasound was ordered. The renal ultrasound revealed grossly enlarged kidneys (15-16 cm) with innumerable cysts (largest 3 cm), consistent with polycystic kidney disease (PKD). Ruptured aneurysms leading to intracranial bleeding are one of the most serious complications of PKD with an estimated prevalence of 5-20% and associated mortality rate as high as 52%. Uncontrolled hypertension and cocaine abuse are independent risk factors for hemorrhagic stroke that are more common than...
underlying PCKD. In patients presenting with a ruptured intracerebral aneurysm and renal failure, the diagnosis of polycystic kidney disease should always be included in the differential diagnosis.

REFERENCES:

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<td>Mesentric neuroendocrine tumor : A Diagnostic mystery</td>
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**INTRODUCTION:** Neuroendocrine tumors (NETs) arise from secretory cells found throughout the body, which makes them biologically and clinically diverse—a diagnostic challenge for both researchers and practitioners. It is not uncommon to see patients with these tumors to take a long time to be identified or all together be misdiagnosed and miscategorized as having some other medical condition. This case exemplifies the diagnostic dilemma and extensive workup that was required to make the diagnosis.

**CASE:** A 67 year old Caucasian male with no significant medical history came to the ED with an episode of severe constipation. An abdominal CT scan was done which revealed a 1X2.2 cm necrotic mass adjacent to the uncinate process of pancreas, anterior to abdominal aorta and contiguous with multiple loops of bowel. Patient underwent an endoscopic biopsy; FNA for cytology and flow cytometry were performed and were non-diagnostic. Patient’s symptoms improved until 4 months later, when the severe constipation returned. Repeat abdominal CT scan revealed that the mass had increased in size to 7.4x4.1 cm, with two new paraaortic lymph nodes (2x1.2 cm and 3x1.9 cm in size), not seen on previous CT scan. PET scan showed only mild hypermetabolism corresponding to above locations. On exploratory laparoscopy it was found that the base of the mass invaded the superior mesenteric artery and vein and only a partial resection was possible. Histology showed low grade (G1) neuroendocrine tumor of probable gastrointestinal tract origin. Ki-67 nuclear labeling was very low (<1%). Serum somatostatin, gastrin, calcitonin and chromogranin A levels were within normal limits. Serotonin was markedly elevated at 991 ng/ml and 24 hour urine 5-HIAA high at 25.3. The patient underwent an octreotide scan that showed metastatic lymphadenopathy in the mesentery, retroperitoneum and left supraclavicular region. He was started on Sandostatin therapy and is currently undergoing treatment.

**DISCUSSION:** With an incidence of 5.25 cases per 100,000 people in the US, neuroendocrine tumors are extremely rare. Patients with neuroendocrine tumors may or may not have symptoms attributable to hormonal secretion; this presents a considerable diagnostic challenge. After revisiting our patient’s prior CT scans, it was found that the original size of the tumor had been stable at 8 cm. Carcinoid tumors are highly vascular and can appear isodense with liver and pancreas on conventional CT scans, leading to diagnostic errors. Because most carcinoid tumors express high-affinity receptors for somatostatin, radiolabeled somatostatin receptor scintigraphy is useful for diagnosis. Resection with regional lymphadenectomy is the primary treatment for most localized tumors. This case illustrates the fact that carcinoid tumors can be silent and, albeit rare, should be considered in the differential diagnosis of a mesenteric mass.

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<td>A baffled mind: a mysterious presentation of small cell carcinoma of the bladder</td>
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We report the case of a 72 year-old man with a history of prostate cancer status post radiation and hormonal therapy several years
| 155 | Clinical Vignette | Imran MD, Tasnim | Rutgers - Newark (Neil Kothari) | The chicken or the egg: pleuropericarditis complicated by a pericardial cyst

We report the case of a 36 year-old woman with no significant past medical history who presented with worsening chest pressure and nausea for five days. Associated symptoms included inability to lie flat. Symptoms improved with leaning forward. The patient reported a recent upper respiratory illness two weeks prior to presentation. Physical exam and laboratory studies were unremarkable, cardiac enzymes were within the normal range, and electrocardiogram showed no evidence of ischemia. Computed tomography scan of the chest revealed a 6.9 x 4 x 2.5cm pericardial cyst adjacent to the right heart border. The patient was diagnosed with pleuro-pericarditis with an underlying pericardial cyst, and was treated with ibuprofen and colchicine. Pericardial cysts are rare mediastinal lesions that are often congenital, but may be inflammatory in a few cases. Diagnosis is confirmed by transthoracic echocardiography, computed tomography scan of the chest or chest magnetic resonance imaging. If symptomatic, surgical resection or percutaneous aspiration of the cyst may be considered. Our patient was treated for pericarditis with reduction in the size of the cyst and improvement in symptoms, which suggests an inflammatory component. It is unclear whether pericarditis led to the formation of an inflammatory cyst or if an underlying pericardial cyst predisposed the patient to developing pericarditis. We describe the first reported case of reduction of a pericardial cyst shortly after treatment of pericarditis due to a presumptive viral infection. In most previous cases, patients had surgical excision of the pericardial cysts to avoid further complications. Our case illustrates that perhaps a more conservative approach may be undertaken for certain patients with pericardial cysts with an inflammatory component. Non-operative management may be appropriate in these select patients.

| 156 | Clinical Vignette | Jain, Shashank and Shuchie Jain, DO | Rutgers - Newark (Neil Kothari) | A Rare Case of Reversible Delirium

**INTRODUCTION:** Delirium is a common problem encountered in various settings, occurring in 30% of older hospitalized patients, 70% of intensive care patients, and 10-50% of surgical patients. It requires a detailed workup and investigation of the underlying condition. However there are cases where a cause is not apparent. We present a rare and commonly under-recognized case of delirium, secondary to autoimmune thyroiditis, which is characterized by cognitive decline and elevation in anti-thyroid antibodies without evidence of stroke, tumor or central nervous system infection.
**CASE PRESENTATION:** A 54 year old female with past medical history of morbid obesity and hypothyroidism presented to the emergency department for rectal bleeding. The patient was unable to respond to questions and only groaned on occasion. Patient’s family described that the patient was responsive and verbal a month prior to this presentation. Pertinent positives on physical exam were vertical nystagmus, 1+ pitting edema in bilateral lower extremities and external hemorrhoids. She was admitted for sepsis due to urinary tract infection (Urinalysis with WBC >182) and rectal bleeding deemed to be from external hemorrhoids. She was also noted to have severe hypothyroidism with a TSH of 36.9 (0.27-4.0uu/ml), Free T4 of <0.4 (0.7-1.5ng/dl), T3 of 0.3 (0.6-1.6ng/dl), thyroid peroxidase of 67 (0-34IU/ml) and anti-thyroglobulin antibodies of 73.6 (0-0.9IU/ml). Throughout the next few days of the hospital course, despite the use of appropriate antibiotics and stable hemoglobin, she remained somnolent and unresponsive with a Glasgow coma scale of 7 (E4V2M1). An Electroencephalogram was obtained showing a low amplitude study with moderate slowing with no epileptiform activity. CSF was positive for elevated protein of 166 (15-45mg/dl) Despite administration of the adequate dose of intravenous (IV) levothyroxine changed from 25 mcg orally (outpatient) to 100 mcg IV, patient’s mental status did not improve. Her TSH improved to 4.99. A CT scan of the head showed mild cerebral volume loss while MRI was without obvious structural brain abnormality although consistent with atrophy of white matter. Patient was subsequently started on IV steroids, which led to significant improvement in mental status. It was decided to keep her on long-term oral steroid therapy.

**CONCLUSION:** Steroid-responsive Encephalopathy associated with Autoimmune Thyroiditis (SREAT) also known as Hashimoto’s encephalopathy, should be considered as a potential cause of delirium of unclear etiology in a patient with hypothyroidism whose mental status fails to improve with adequate dose of oral or intravenous levothyroxine, particularly if there is elevated protein level in CSF in the absence of CNS infection. The prompt treatment with corticosteroids is warranted as residual cognitive impairment occurs in up to 25 percent with long-standing untreated disease.

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<td>An Unusual Pathogen in an Unusual Circumstance: A Case of Progressive Unilateral Vision loss</td>
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**INTRODUCTION:** Vision loss is a frightening experience for patients, and always requires an urgent ophthalmological evaluation. Early diagnosis and treatment generally translates to preservation of vision. One cause of rapid vision loss is acute retinal necrosis (ARN). We present a case of ARN where the pathogen and underlying pathophysiology are very uncommon.

**CASE PRESENTATION:** A 32 year old woman with history of multiple sclerosis (MS), presented for left eye pain and vision loss for 10 days. The symptoms started with intermittent blurring in her left eye and associated sharp, 4/10 pain with no radiation. The patient sought medical care and was told the vision loss was secondary to MS flare with some mild anterior uveitis, for which she was treated. The pain progressed to chronic, 10/10 pain with complete vision loss in the left eye after 4 days. Pain was exacerbated by movement, without relieving factors. Patient denied fevers, rashes, chills, nausea, vomiting, headaches, recent travel, insect bites or sick contacts, and no previous vision involvement with MS flares. Of note, the patient had a MS flare 2 weeks prior, when she was hospitalized and given a 5 day course of intravenous (IV) steroids. Exam revealed no light perception, sluggish pupillary responsive to light, active panuveitis, and dense viteritis in left eye. Vitals, Right eye exam and remainder of the physical exam was normal. Lab studies including CBC, BMP and HIV screening were all normal. Magnetic resonance imaging (MRI) showed new edema involving the entire left optic nerve. Lumbar puncture was done, cerebrospinal fluid analysis including bacterial and fungal cultures were negative. Viral studies showed only a positive HSV-2 IgG. Patient underwent left vitrectomy, and vitreal tissue was positive for HSV-2 by PCR.
Other viral studies, and cultures were negative. Post-operative fundoscopy showed micro hemorrhages, diffuse retinitis, periarteritis and inflammation indicating ARN. Patient was treated with 7 days of IV acyclovir, and discharged with oral valacylovir. 1 month later patient continued to have no light perception in the left eye, with normal vision in the right eye. Repeat MRI imaging showed mildly improved left optic neuritis.

CONCLUSION: ARN is an uncommon but a devastating, potentially blinding disease, most commonly caused by Varicella Zoster virus. Traditionally ARN caused by HSV-2 infection was seen in children generally with neonatal exposure, but incidence is growing in adults. Vitrectomy is a key component in management with recent studies showing early and prophylactic vitrectomy may improve outcomes. Clinical suspicion for diagnosis is paramount, especially if patient has received oral or IV steroid therapy 2-3 weeks prior to start of symptoms. Early diagnosis and treatment is essential in order to prevent worsening of vision, and prevent co-infection on the other eye.

Successful Management of MGUS Associated Neuropathy with Rituximab

BACKGROUND: Monoclonal gammopathy of undetermined significance (MGUS) is a premalignant clonal plasma cell disorder which is most commonly asymptomatic but may sometimes have diverse presentations such as hemolytic anemia, hypercalcemia, skin rashes, neuropathy etc. We present a case of MGUS with an unusual clinical presentation of isolated peripheral neuropathy.

CASE: A 44 year old Caucasian female with no significant past medical history presented to the hospital with progressively worsening numbness, tingling sensation and pain in the bilateral lower limbs for 3 months. Physical examination revealed bilateral below knee paresthesia and dysesthesia. EMG and nerve biopsy showed evidence of moderate to severe demyelination suggestive of Chronic Inflammatory Demyelinating Polyneuropathy (CIDP). Patient was initially treated symptomatically with Gabapentin with no significant improvement of symptoms. Further workup revealed an elevated serum IgM of 380U with normal IgA (166) and IgG (986) levels with a serum M spike of 0.22 g/dL. Bone marrow biopsy revealed <1 percent monoclonal plasma cell population. Apart from the neuropathy, the patient had no other symptoms, electrolytes were in normal range and PETCT showed no evidence of lytic bone lesions or pathologic lymphadenopathy. A diagnosis of MGUS was made and the patient was treated with four cycles of Rituximab 375 mg/m² IV once a week for 4 weeks followed by a maintenance dose of Rituximab 375 mg/m² IV once every 2 months. Patient showed significant clinical improvement with the Rituximab therapy. IgM levels dropped to normal range and the ‘M’ spike on electrophoresis was no longer present.

DISCUSSION: MGUS is defined by the presence of a serum monoclonal protein at a concentration <3g/dl, a bone marrow with <10 percent monoclonal plasma cells and absence of end organ damage. MGUS occurs in over 3% of the Caucasian population above 50 years of age. Due to this high prevalence we believe MGUS associated neuropathy is a relatively underdiagnosed cause of isolated neuropathy, especially in older adults. Standard symptomatic management with Gabapentin and immunomodulatory agents usually used for CIDP like steroids, intravenous immunoglobulin and plasmapheresis have not shown to be efficacious in MGUS but instead, like in our case, studies show good response with Rituximab. Moreover, the number of MGUS patients progressing to a plasma cell neoplasm or related disorders was 7.3 times than that expected in the general population and this number is higher in patients with MGUS neuropathy, hence all patients with MGUS neuropathy need to be followed up with serial blood work to monitor for
malignant transformation. We suggest that all patients with idiopathic peripheral neuropathy who do not respond to symptomatic management should be investigated for MGUS and all patients with MGUS and neuropathy should be treated with Rituximab.

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<td><strong>Spontaneous Hemoperitoneum as an Initial Manifestation of Plasma Cell Leukemia</strong></td>
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<td><strong>INTRODUCTION:</strong> Plasma Cell Leukemia (PCL) is a rare but aggressive variant of multiple myeloma which either presents as a progression of multiple myeloma or as primary PCL. It typically presents with anemia, leukocytosis, acute renal failure, hypercalcemia, lytic bone lesions and in some cases with hepatosplenomegaly. We present a case of plasma cell leukemia presenting with spontaneous splenic bleeding.</td>
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<td><strong>CASE REPORT:</strong> A 49 year old African American male with past medical history of hypertension, diabetes and anemia presented with one day of acute onset abdominal pain that woke him from sleep. Patient denied any preceding trauma or associated nausea, vomiting or diarrhea. He had visited the emergency department one month before for medication refills, and had been diagnosed incidentally with normocytic anemia (hemoglobin of 8.5 gm/dL). On physical examination, he was borderline hypotensive; his abdomen was soft but diffusely tender with voluntary guarding. Stool was guaiac negative. Initial hemoglobin was 6.9 gm/dL which decreased to 5.1 gm/dL hours later. Laboratory studies were notable for hyperproteinemia with a protein gap, hypercalcemia, elevated LDH and acute renal failure. Abdominal imaging revealed hemoperitoneum with greatest amount of blood near the spleen, which appeared enlarged but without any active bleeding or laceration. The patient received emergent fluid hydration and blood transfusion without adequate response in hemoglobin, and a splenic angiogram was performed which did not reveal any active extravasation. The source of the bleeding was thought to be secondary to a slow but persistent splenic venous bleed and the proximal splenic artery was embolized to prevent further bleeding. Serum electrophoresis was significant for IGG monoclonal gammopathy lambda type. A lymphoma panel of peripheral blood revealed clonal population of plasma cells (17%) that expressed CD38, CD138 and lambda, but were negative for CD45, CD56, CD19 and CD20 expression. Patient had elevated serum free lambda light chains. Further imaging revealed supraclavicular lymphadenopathy and retroperitoneal and intra-abdominal lymphadenopathy with lytic lesions in thoracic vertebral bodies. The patient was transferred to an outside specialty center where a bone marrow biopsy revealed CD138 positive plasma cells which were about 70% of marrow cellularity. Cytogenetic studies were significant for t(14;20) translocation, gain of long arm of chromosome 1, loss of chromosome 13 which indicate poor prognosis.</td>
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<td><strong>DISCUSSION:</strong> PCL usually presents with signs and symptoms which are often seen in multiple myeloma and other leukemias. It is unclear whether our patient had underlying myeloma prior to presentation which progressed to PCL vs. primary PCL. Spontaneous splenic rupture is a rare occurrence but it has been reported to occur in acute and chronic leukemia. Only four case reports of spontaneous splenic ruptures have been reported with PCL, making it a rare but potentially fatal complication of an already aggressive and deadly disease.</td>
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<th>Kileci, John</th>
<th>Anthony Kim, MD and Rafael A Teran Vargas, MD</th>
<th>Rutgers - Newark (Neil Kothari)</th>
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<td><strong>Endocarditis and Mucormycosis in an elderly lady after Intravitreal Kenalog (Triamcinolone Acetonide) Injection</strong></td>
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<td><strong>INTRODUCTION:</strong> Kenalog (triamcinolone acetonide) is a common medication used to prevent diabetic macular edema. We present a case where an outpatient procedure of intravitreal triamcinolone acetonide injection was associated with our patient developing mucormycosis and sinusitis leading to bacteremia and endocarditis.</td>
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CASE: A 78 year old female with past medical history of diabetes mellitus type 2, macular degeneration, osteoporosis, hyperlipidemia presented to the hospital for 2 weeks of right sided headaches, fevers and uncontrolled glucose levels. Her headaches started behind the eye, felt like electric shocks and encompassed the entire right side of her head all the way to the occiput. These headaches began after a Kenalog injection into her right eye for macular degeneration by her ophthalmologist. Over the last week, her fingersticks at home were over 500 which was a change to her previously somewhat well controlled diabetes (HbA1c 6.9%). On exam, she was tender from her right eye all along the right side of her face, temporal head, down to her neck. She also had blurriness in her vision in her right eye and a new holosystolic murmur. Her initial glucose level was 609 and blood cultures taken in the Emergency Room were positive for methicillin sensitive staphylococcus aureus (MSSA). A CT scan of head was obtained which showed bilateral sphenoid sinus congestion. Transesophageal echocardiography showed a 0.9x0.8cm vegetation on the mitral valve. She was started on oxacillin. An MRI head was obtained that reconfirmed the bilateral sphenoid sinus congestion and also showed acute-subacute right caudate, right frontal and right cerebellar infarcts that were thought to be septic emboli. Her headaches were very difficult to control and carbamazepine was the only modality which would minimally decrease symptoms. She underwent bilateral spheniodotomy that showed mucor species and MSSA and was started on liposomal amphotericin B for mucormycosis. Due to toxicity of amphotericin and her septic state patient eventually required hemodialysis. Repeat sphenoidotomy showed resolving mucormycosis after 9 days of treatment and patient was switched to posaconazole. Although on appropriate treatment, she eventually deteriorated requiring mechanical ventilation and vasopressor support. After a 37 day stay in the hospital, per family’s request she was made comfort care and deceased.

DISCUSSION: Diabetic Macular Edema is one of the major complications of diabetes mellitus type 2. One treatment option is intravitreal glucorticoids. To our knowledge this is the first case report describing a treatment session with kenalog resulting in multiple complications including mucormycosis and sinusitis leading to bacteremia and endocarditis. Given recent intravitreal injection and both the right sphenoid sinuses and her heart valves growing MSSA, one could argue that there may be a direct correlation between the injection and patient’s symptoms.
**DISCUSSION:** Renal disease is a frequent manifestation of systemic amyloidosis. Without treatment, it usually progresses to ESRD. Diagnosis is made by biopsy and Congo Red stain. Staining for a specific protein is essential as different forms of amyloid require different treatment. Secondary amyloidosis is caused by AA protein deposition and is found in patients with chronic inflammatory states such as rheumatoid diseases, inflammatory bowel disease and chronic infections. Although rarer than primary amyloidosis, renal cell carcinoma has been associated with secondary amyloidosis as well. Treatment of AA amyloidosis is focused on decreasing inflammation. In our case, patient was treated, however still progressed to end stage renal disease and hemodialysis.


**INTRODUCTION:** Renal cell carcinoma (RCC) is an uncommon tumor, accounting for approximately 3% of all adult malignancies. Most cases are asymptomatic at initial presentation and detected incidentally on abdominal imaging.

**CASE:** A 77 year-old Caucasian man with a past medical history of diabetes mellitus, hypertension and obesity presented with the complaint of constant, progressively worsening bilateral neck pain for 4 weeks. There was no history of recent trauma or similar pain in the past. Review of systems was otherwise negative. On physical examination, there was decreased range of motion of the neck due to pain. The remaining physical examination was unremarkable. Computed tomography (CT) scan of the neck revealed a destructive, unstable lytic lesion at the C2 level, with a pathological fracture of the dens [D1] and surrounding edema. There was no mass effect on the spinal cord. MRI of the cervical spine showed a heterogeneous lesion involving the entire C2 vertebral body, with diffuse enhancement. These findings triggered a search for a primary malignancy. CT scan of the abdomen showed an ill-defined nodular mass arising from the mid-pole of the left kidney, with increased attenuation. No other metastatic lesions were identified. Percutaneous biopsy of the renal mass showed fibrosis containing clusters of granular cells and focal papillary areas, diagnosed as papillary sarcomatoid neoplasia of the kidney. The patient underwent surgical stabilization of the cervical spine, followed by 10 fractions of radiation therapy. The patient then underwent cytoreductive radical left nephrectomy. He is now asymptomatic and doing well one year after diagnosis, with ongoing surveillance for tumor recurrence.

**DISCUSSION:** Patients with RCC typically present with painless hematuria, flank pain or abdominal mass. Sarcomatoid renal cell carcinoma (SRCC) is a highly aggressive RCC subtype that is poorly responsive to chemotherapy and immunotherapy. Metastases most commonly occur to the lung, lymph nodes, liver and brain. The reported average survival time is less than six months. Survival is longer for patients with localized disease, single metastatic site, partial response to systemic therapy, and nephrectomy. In our patient presenting with neck pain, the initial differential diagnosis was broad, with osteoarthritis as the leading diagnosis. This report highlights the importance of a high index of suspicion for bone metastases in patients with localized and progressively severe musculoskeletal pain. The most likely primary cancer sites are lung, prostate, multiple myeloma and lymphoma. Although bone metastases occur in up to one-third of patients with RCC, an isolated bony lesion is very rarely the tumor’s initial presentation. Early diagnosis and initiation of therapy may prolong survival for patients with this highly aggressive tumor.
### 163. Clinical Vignette

**Nizami, Sobia**  
Srikanth Thiruvadsothy MD, Dina Khatreeb DO, Slawomir Sender MD  
Rutgers - Newark (Neil Kothari)

**Coarctation of the abdominal aorta: A very rare cause of adult acute abdominal pain**

**INTRODUCTION:** Coarctation of the abdominal aorta is a rare vascular entity. As a congenital condition, it generally presents in children and young adults with lower extremity claudication, renovascular hypertension, and abdominal angina from compromised regional blood flow. Coarctation of the abdominal aorta may also be associated with acquired conditions in adults such as large vessel vasculitis, fibromuscular dysplasia, retroperitoneal fibrosis, and atherosclerosis.

**CASE:** A 50 year-old African American woman with a history of hypertension controlled on hydrochlorothiazide, celiac disease, asthma, and hysterectomy for fibroids, presented to the emergency department with the complaint of severe lower abdominal pain. The abdominal pain had progressed over four days, was described as sharp, severe, and localized to the right lower quadrant. It was associated with nausea and non-bilious vomiting. She also reported a long-standing history of intermittent bilateral leg claudication. She denied similar past episodes of abdominal pain. On examination the patient was afebrile, normotensive, and without tachycardia. Right lower quadrant tenderness was found on abdominal examination. Complete blood count, comprehensive metabolic panel, and inflammatory markers were normal. CT scan of the abdomen/pelvis revealed markedly unusual tortuosity of the aorta and great vessels of the abdomen, but was otherwise unremarkable. CT angiogram demonstrated aortic narrowing starting below the celiac axis level, with a minimum lumen of 4 mm consistent with coarctation. The left renal artery originated above the coarctation, while the inferior mesenteric artery (IMA) and right renal artery originated below. She was managed conservatively with intravenous hydration, analgesia and bowel rest. She improved with supportive care after four days and was discharged home with vascular surgery follow-up.

**DISCUSSION:** There are very few reported cases of congenital abdominal aortic coarctation presenting in adulthood. It commonly involves renal artery stenosis, resulting in severe resistant hypertension. Visceral artery occlusion can also occur, with development of tortuosity and collaterals of the abdominal circulation. If left untreated, its prognosis is poor with 90% mortality by age 60. Death is usually from complications of uncontrolled hypertension such as stroke, renal failure, and congestive heart failure. In our patient, the history of claudication, the finding of extensive tortuosity of abdominal vessels, and lack of clinical or laboratory evidence of vasculopathy, strongly suggested congenital aortic coarctation to be the cause of abdominal pain. Although, this is difficult to prove in the absence of prior imaging. We suggest the patient developed bowel ischemia in IMA territory, causing abdominal pain and vomiting. It is unclear what triggered her symptoms and why they resolved spontaneously. The case is an atypical presentation of abdominal aortic coarctation, presenting at a later age with controlled hypertension. Despite being rare, abdominal aortic coarctation should be considered a differential for abdominal pain in adults as highlighted by this report.

### 164. Clinical Vignette

**Ogbonnaya-Odor, Chinonye**  
Ramneek Nakai MD, George Nahas MD, Srithi Srinivas MD, Ahmed Shahida MD  
Rutgers - Newark (Neil Kothari)

**ADENOCARCINOMA OF UNKNOWN PRIMARY CAUSING CONSTRUCTIVE HEART DISEASE**

Cardiac involvement by cancer, leading to pericardial and sometimes myocardial disease, is a well-known complication of advanced metastatic disease. Cardiac tamponade and pericarditis as a result of metastatic carcinomatosis without initial manifestation of primary tumor is uncommon. We present a case of cardiac tamponade followed by rapid cardiac constriction as initial presentation adenocarcinoma of unknown primary site. Patient is a 45 year-old black man with history of HIV, compliant with HAART and...
hypertension, who presented for medical care, complaining of a 6 week history of worsening shortness of breath at rest which was increased with exertion, night sweats, non-productive cough and 10 lbs weight loss. On examination, he was found to have a blood pressure of 101/67 mmHg, pulse of 99 beats per minutes, oxygen saturation of 94% on room air, decreased breath sounds on the left chest, without jugular venous distention, abnormal heart sounds, ascites or lower extremity edema. Laboratory studies showed WBC of 15 x 10^9/L, sodium of 132mEq/L, LDH of 268IU/L. A Chest x-ray showed left pleural effusion and cardiomegaly. A computed tomography (CT) pulmonary angiogram revealed left pleural effusion, moderate pericardial effusion and significant lymphadenopathy. Electrocardiogram (EKG) showed sinus tachycardia. Transthoracic echocardiogram (TTE) suggested early cardiac tamponade. A pericardial window was performed; a pericardial drain and a left chest tube were placed. Pericardial tissue analysis revealed metastatic adenocarcinoma. The specimen was positive for cytokeratin 7, abundant mucin, and ber-EP4 but negative for cytokeratin 20, CDX-2, TTF-1, PSA, PSAP, mesothelial markers and inhibin. Despite negative stain for TTF-1, origin was speculated to be from the lung. After initial progress, patient developed dyspnea, increased abdominal girth, and peripheral edema. On day 20 of admission, he developed new chest pain. Atrial flutter with rapid ventricular response was seen on EKG and treated with cardioversion. Readings determined by Swan-Ganz catheter were consistent with constrictive pericarditis. The patient decided to discontinue therapy for comfort measures. He died two days later. Autopsy revealed diffuse metastasis to multiple organs including the pleura, pericardium, epicardium, myocardium and lymph nodes. Meticulous search for the primary malignancy did not reveal one. Pericardial disease is an important cause of mortality in cancer patients, it is has however been identified as a late finding in malignancy. This case reinforces the importance of increased suspicion for malignancy in a patient presenting with acute onset of pericardial effusion, tamponade or constrictive pericarditis, as it may be the first presentation of malignancy especially in patients with HIV where malignancy has been documented to be rather aggressive and presents at younger ages.

**INTRODUCTION:**

The introduction of highly active antiretroviral therapy (HAART) has extended the life expectancy of patients with HIV due to immune reconstitution, and decreased opportunistic infections. It is well known that certain malignancies, including plasma cell disorders, have an increased incidence in patients with HIV, but multiple myeloma (MM) cases have been infrequently described. The exact molecular mechanism to account for this is unknown and optimal treatment for these patients is still undefined.

**CASE PRESENTATION:**

A 62 year old male with a medical history of HIV and HTN presented with worsening back pain for 4 months. The pain was sharp, non-radiating, localized to the midline lumbar spine, exacerbated by movement, and worst when supine, and alleviated moderately by acetaminophen/oxycodone and tramadol. The patient had not been able to ambulate for the past 2 months due to pain, and required admission to a rehabilitation facility. Patient denied fevers, chills, recent travel, bowel or urinary incontinence, numbness, or tingling. In addition to the diffuse tenderness noted in the lumbar spinal region, there was decreased lower extremity strength with brisk patellar reflexes bilaterally. Patient was found to have total protein of 12.2 (normal 6.4 - 8.6 gm/dl) and albumin of 2.6 (normal 3.9 - 5.2 gm/dl). CD4 count was 589 cells/ml with viral load <20. An x-ray showed multiple areas of lucency throughout the bony calvarium consistent with diffuse lytic lesions. A CT scan showed extensive lytic lesions seen in lower thoracic, lumbar vertebral, and sacrum, with a severe compression deformity of L5, with mild compression of L2-L4. Peripheral blood smear showed rouleaux formation. Bone marrow aspirate showed hypercellularity, plasma blasts with distinct nucleoli and areas of perinuclear clearing. Serum protein electrophoresis (SPEP) revealed an M spike in the beta-2 region. Immunofixation was
consistent with monoclonal gammopathy restricted to IgA heavy chain and kappa light chain. Cytogenetics showed a monosomy of chromosome 13 (55.5% of cells), and a translocation between chromosomes 4 and 14 (t(4;14)/FGFR3-IGH). Patient received radiation therapy to the lumbar spine and was discharged to receive outpatient treatment of bortezomib and dexamethasone.

**DISCUSSION:** This case highlights the radiological and laboratory findings seen in MM with well controlled HIV. Although plasma cell disorders are seen at a higher incidence in HIV patients, MM is not commonly described. It is not known if this is related to increased life expectancy of HIV patients on HAART, immune reconstitution, or unknown molecular mechanisms. Early recognition and treatment are paramount for improved outcomes, especially since these patients may have an increased risk of complications, including intractable hypercalcemia, hyperviscosity, cytopenia, and renal failure. Optimization of HAART is also critical, as HAART therapy may also have some therapeutic effect on MM.

**166.**

**Clinical Vignette**

Patel, Monali

Raad Tashman, MD

Rutgers - Newark (Neil Kothari)

**Brugada phenocopy induced by intracranial hemorrhage**

Brugada phenocopy is a clinical subgroup of Brugada syndrome that demonstrates EKG findings consistent with Brugada syndrome in patients who do not have true congenital Brugada syndrome. It is well known that this EKG finding is induced by metabolic disturbances, mechanical compression, ischemia, myocardial/pericardial diseases, heroin and ethanol use. This case report illustrates that acute intracranial hemorrhage can also unmask Brugada phenocopy in susceptible patients. A 57-year-old previously healthy male was brought to the hospital after having syncopal episode with subsequent head trauma while walking. He had brief lucid interval but then he was found obtunded and did not follow commands. He also had witnessed generalized, tonic-clinic seizures en route to the hospital. He does not have any past medical history and does not take any medications at home. CT scan of the head exhibited extensive fractures involving left occipital bone, left mastoid and petrous temporal bones, and left sphenoid sinus roof extending to left carotid canal. It also demonstrated diffuse bifrontotemporal subarachnoid hemorrhages (SAH), subdural hemorrhages along the frontal convexities, temporal convexities and falk cerebri, as well as epidural hematomas. A 12 lead EKG revealed ST segment elevation with Brugada type-2 morphology in leads V1, and V2. ST segment elevations were also noted in V3, V4, V5 and V6. Serial cardiac troponins were negative. Echocardiogram was within normal limit. Serial EKGs over the following days showed gradually resolving ST segment elevations and complete resolution to baseline after 48 hours of the initial event. Repeat CT scan of the head showed improving bilateral SAH and subdural hematoma. Based on the clinical picture and previous medical history, it was concluded that the patient had Type-2C Brugada phenocopy that was unveiled by acute intracranial hemorrhage. To our knowledge this is the second reported case of Brugada phenocopy induced by acute intracranial hemorrhage in literature. We are postulating that intracranial hemorrhage is another clinical entity that can provoke Brugada phenocopy in susceptible patients, which can lead to sudden cardiac death. Hence, extensive cardiac workup is crucial in these patients to avoid morbidity and mortality.

**167.**

**Clinical Vignette**

Patel, Sima

Shreya Patel, Eugenio Capitle

Rutgers - Newark (Neil Kothari)

**A Case of An Allergic Reaction To Omalizumab**

**RATIONALE:** Omalizumab is an injectable prescription medication that has been approved for use in patients over the age of 12 diagnosed with moderate to severe persistent allergic asthma caused by perennial aeroallergens. Allergic reactions to Omalizumab are rare, especially a delayed type hypersensitivity reaction. Here we present a patient who developed an allergic reaction to
Omalizumab three days after her third dose.

METHODS: Skin Prick and Intradermal skin testing performed.

RESULTS: A 59 year old African American female was originally evaluated in our clinic for allergic asthma. Skin testing revealed sensitivity to trees, grass, weeds, molds, dust mite and dog. Total IgE was noted at 711. Patient was enrolled and approved for Omalizumab. Three days after her third Omalizumab injection, she developed facial swelling with angioedema of lips. She denied tongue and throat swelling as well as shortness of breath. She was treated in the emergency department with benadryl, famotidine and prednisone prior to arrival to the clinic. Skin prick testing and intradermal testing to Omalizumab was found to be negative but clinical history suspicious for reaction to Omalizumab.

CONCLUSIONS: Although rare, allergic reactions have been reported to occur after administration of Omalizumab. Thus, health care providers administering the medication should be cognizant of such reactions. Due to this risk of allergic reaction, patients are observed closely after Omalizumab administration. However, as demonstrated in this case, it is also important to be aware of delayed reactions to Omalizumab.

A young male with Systemic Lupus Erythematosus presenting with seizures secondary to Posterior Reversible Encephalopathy Syndrome (PRES)

RATIONALE: Posterior reversible encephalopathy syndrome (PRES) is an under recognized condition in systemic lupus erythematosus (SLE) that can mimic neuropsychiatric lupus. Identification of distinct clinical and radiographic patterns is important, as one would need to escalate rather than decrease immunosuppressant therapy in neuropsychiatric lupus. The majority of PRES cases secondary to SLE reported in the literature involve female patients. We report a young male patient presenting with PRES secondary to uncontrolled hypertension due to lupus nephritis.

METHODS: MRI brain demonstrated findings consistent with PRES. RESULTS: Patient is a 20-year-old male with history of SLE (lupus colitis, lupus nephritis), HTN, and end-stage renal disease (creatinine 2.9) who presented to the emergency department (ED) with new onset tonic-clonic seizures. His blood pressure in the ED was 197/114 for which he was started on a Cardene drip and admitted to ICU for further management. Subsequent MRI brain revealed multiple foci of increased FLAIR and T2 weighted signal intensity in the subcortical and periventricular matter bilaterally, and bilateral cerebellum; edema in the parietal and occipital regions consistent with PRES.

CONCLUSIONS: Systemic lupus erythematosus should be considered in the differential diagnosis of young patients who present with PRES. Clinicians should have a low threshold for MRI especially when neurological symptoms occur in young patients with SLE; even more so in those with active lupus, lupus nephritis, renal failure, and/or poorly controlled hypertension. We feel that consideration of this diagnosis in young adults with lupus, including males should be recognized, as prompt recognition is crucial to deliver appropriate management.
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<th>Patel, Sima</th>
<th>M. Kang</th>
<th>Rutgers - Newark (Neil Kothari)</th>
<th>A Lethal Cause of Nasal Pain</th>
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<td>INTRODUCTION: Patient is a 39 year old Hispanic female with no significant past medical history who presented to the clinic with the complaint of burning pain in her bilateral nares for the past 5 months. She further reports feeling blister-like lesions in her nose bilaterally as well as occasional clear nasal discharge with scant nosebleeds. She denies any sinus pain, sneezing, headaches, ear pain, sore throat, or cough. Patient was seen by multiple physicians previously, and was diagnosed with persistent nasal infections. She was subsequently treated with multiple antibiotics including Augmentin, Keflex, and Clindamycin. On physical exam, patient’s vital signs were unremarkable. Pertinent positive exam findings included ulcerated lesion of the right anterior nasal septum and medial nasal vestibule with erythema of columella skin. Tenderness of the septum and nasal dorsum were also present. Lab results including CBC, CMP, ESR, and CRP were all within normal limits. ANA, RF, c-ANCA, p-ANCA, anti-SSA, anti-SSB, quantiferon gold, RPR, and HIV were negative. Patient underwent a nasal endoscopy by ENT that revealed right-sided ulcerated, friable mucosa on the anterior, inferior nasal septum. Additionally, CT scan of the sinuses revealed diffuse paranasal sinus mucosal thickening and marked enhancement on the right middle and inferior nasal turbinate suggestive of bone erosion along the medial wall of the right maxillary sinus. Biopsy of the septum revealed necrotic debris with lymphocytes and epithelioid histiocytes. During the work-up, patient was noted to have microscopic hematuria and underwent renal biopsy which returned negative for granulomatosis with polyangiitis (Wegener’s). The above clinical findings are consistent with idiopathic midline necrotizing granulomatous disease. Patient was started on oral steroids, followed by Rituximab and Methotrexate with poor response. Consequently, she was changed to Cyclophosphamide, which has provided a very good response.</td>
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<td>DISCUSSION: Midline granulomatous disease (aka lethal midline granuloma) is a mutilating process produced by a variety of diseases that progressively destroy the nose, paranasal sinuses, and palate. Neoplastic, infectious, autoimmune, and idiopathic forms of this disease have been described in the literature. Despite improved understanding of these various disease processes, there continues to be difficulty in establishing an early diagnosis. This becomes problematic as there are subsequent delays in prompt treatment to minimize loss of function and cosmetic deformity. The specific etiology of this disease must be ascertained as well, as the treatment varies depending on the underlying cause.</td>
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<td>CONCLUSION: This case is presented to alert physicians to consider the rare phenomenon of midline granulomatous disease when patients present with persistent nasal pain and rhinorrhea. It is imperative to recognize this clinical entity and begin treatment immediately to prevent progressive destruction of the facial midline structures. Treatment depends on the etiology of the disease process and often consists of immunosuppressive agents.</td>
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<th>Clinical Vignette</th>
<th>Pievsky, Daniel</th>
<th>Narjust Duma</th>
<th>Rutgers - Newark (Neil Kothari)</th>
<th>An uncommon clinical presentation of a rare lung cancer</th>
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<td>Lung cancers can present in unique ways and frequently do not follow the hemoptysis and weight loss stereotype. This is especially true for uncommon histological subtypes like sarcomatoid lung cancer, a type of non small cell cancer which is a diagnostic challenge in its own right due to its histological heterogeneity. We present a case of undifferentiated sarcomatoid carcinoma of the lung with dysphagia as the initial presentation. A 67 year old African American male with past medical history of hypertension, hyperlipidemia, surgically resected prostate cancer and a 35 pack year smoking history was initially evaluated as an outpatient for complaints of dysphagia and shortness of breath. Preliminary chest x-ray showed a double density along the left heart boarder. A</td>
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follow up chest CT revealed a 7.5cm soft tissue mass in the middle mediastinum encasing the aorta, displacing the esophagus to the right and the heart anteriorly. He presented to the emergency room several days later with worsening shortness of breath. An endoscopic ultrasound with needle biopsy was performed which demonstrated malignant sarcomatoid neoplasm with giant cell features not further classified. PET scan showed intense hypermetabolism within the mediastinal mass with central necrosis of the tumor. The neoplasm was staged as IIIA. The patient was treated with multiple cycles of systemic chemotherapy and radiation. Despite treatment, disease progression was noted with metastases to the left humerus, right femur and spine which resulted in compression of the cauda equina. He underwent decompression and tumor debulking by neurosurgery; however, his clinical status rapidly deteriorated precluding the possibility of further treatment. The patient was made comfort care 24 months after initial presentation. Sarcomatoid carcinoma is a rare subtype of lung cancer, accounting for 0.3 – 3% of all non-small cell lung carcinomas. It is a very aggressive cancer with a 5 year survival rate ranging from 17.4-24.5% compared to 46.3% for matched patients with other types of non-small cell lung cancer. It is usually associated with men who have a history of heavy smoking, and the typical presentation is identical to that of other typical non-small cell cancers with cough, hemoptysis and dyspnea. While our patient did have dyspnea, his primary complaint of dysphagia is an extremely unusual presentation for this type of cancer. Sarcomatoid carcinomas are a major diagnostic challenge because of their histological heterogeneity resulting in a preoperative misdiagnosis rate as high as 70.6%. It is also exquisitely difficult to treat and has a median time of recurrence of 11.3 months compared to 61.4 months for other non-small cell lung cancers. Surgical resection, if possible, is the only curative treatment as adjuvant chemotherapy and radiotherapy appear to have little benefit.

171. Clinical Vignette

Pievsky, Daniel

Igor Eyzner, Leia Rispoli, Samit Desai

Rutgers - Newark (Neil Kothari)

The cat's in the bag (of dialysate)

Pasteurella multocida is a gram negative coccobacillus that is commonly found in the oral cavity of many wild and domestic animals, specifically cats and dogs. The bacterium may be transmitted to humans via a bite, scratch or lick from a colonized host. While soft tissue infections from Pasteurella are common, cases of peritonitis are rare and can be a diagnostic challenge. A 73 year old female presented to the emergency department with a one day history of worsening abdominal pain which was sudden in onset, constant, non-radiating and initially diffusely dull but became sharp and centralized. She had a past medical history significant for diabetes and end stage renal disease for which she was on continuous cycling peritoneal dialysis (CCPD) via a Tenckhoff catheter which had been placed 16 months prior to the current presentation. She denied any fevers, chills or any changes with her dialysis site or dialysate solution. Her vital signs were notable for a low grade fever of 100.1°F and a blood pressure of 154/78 mmHg. Her physical exam was unremarkable except for a distended abdomen with mild diffuse tenderness. There was no exudate or erythema noted around her peritoneal dialysis (PD) catheter site. Her labs were significant for a WBC of 11,000 cells/mm3 with 73.95% neutrophils on the differential. The peritoneal dialysate was noted to be cloudy and the fluid analysis showed 4,230 WBCs with 92% neutrophils but no organisms on gram stain. A diagnosis of PD catheter associated peritonitis was made and the patient was given empiric Tobramycin, Vancomycin and Fluconazole therapy. Subsequent peritoneal dialysate cultures grew out Pasteurella multocida which was sensitive to Ampicillin, Cefazolin and Gentamicin. Blood cultures remained negative. Further history obtained from the patient revealed that she had two cats at home which sleep with the patient in the same bed. The patient's peritoneal cell count normalized over three days and subsequent cultures remained negative on intravenous Tobramycin therapy. The intravenous therapy was chosen over the intraperitoneal therapy based on discussion with the renal service and the ease of administration after discharge. Prior to discharge the importance of good hygiene and appropriate PD catheter site care was discussed in detail with the patient. This case illustrates the potential concerns for PD patients with pet exposures and the importance of good hygiene and sterile technique when
Performing self care on long term catheters. Pets are an important part of the family for many patients and while their presence has many noted health benefits it is paramount to stress to patients that even domestic animals may be a source of life threatening infection.

172. 298 Clinical Vignette
Razi-Akmal, Rabail
Rajendra Kapila, MD
Rutgers - Newark (Neil Kothari)

HIV Vasculopathy and Varicella Vasculitis in a patient with HIV/AIDS

INTRODUCTION: HIV vasculopathy and varicella vasculitis are two infectious causes of strokes in HIV/AIDS patients. Early identification would help treat and prevent recurrence.

CASE PRESENTATION: A 52 year-old woman with HIV/AIDS (CD4 count of 4) presented with altered mental status and being non-verbal for seven hours. Family denied any seizure activity, tongue biting, or bowel or bladder incontinence. Two months prior to admission she had developed an erythematous, vesicular rash on her extremities. Over the past month she had been losing weight with no apparent hematemesis, abdominal pain, hematochezia, melena, fevers, or chills. She appeared cachectic with hyper-reflexia and increased muscle tone in both the upper and lower extremities. Initial labs showed leukopenia with bands of 20%. CT scan of the head showed multiple low-density foci in left frontal lobe and right internal capsule. An MRI showed multiple foci of acute and sub-acute infarction along multiple vascular territories in both cerebral hemispheres. She was started on empiric treatment for meningitis with Ceftriaxone, Vancomycin, Ampicillin, and Acyclovir. An electroencephalogram was obtained that showed moderate slowing and no epileptiform activity. An echocardiogram showed no vegetation. A lumbar puncture was performed which showed clear fluid with an opening pressure of 18 cm H2O, 5 white blood cells (79% lymphocytes), glucose 34, and protein 88. Further cerebrospinal fluid studies detected both Varicella and HIV DNA. She was also seen by Ophthalmology for retinitis which was initially misidentified as CMV. A CT angiogram showed irregularity of left anterior cerebral and right middle cerebral arteries consistent with vasculitis. She was thus diagnosed with HIV vasculopathy and Varicella vasculitis along with vesicular rash and retinitis.

DISCUSSION: Cerebral infarction may be present in 4% to 29% of patients with AIDS based on autopsy results. An HIV-associated vasculopathy has been described in AIDS patients upon autopsy showing small vessel thickening, pigment deposition, and perivascular inflammatory infiltrates in the absence of an identifiable cause (lymphoma, embolism, or non-HIV central nervous system infection). Varicella zoster vasculopathy is often chronic with lesions occurring at the grey-white matter junction and mostly involving both large and small arteries. CSF mononuclear pleocytosis and a shingles rash may not be present in a third of the cases hence a high index of suspicion is necessary, and early and aggressive antiviral therapy may improve the outcome in such patients.

173. 97 Clinical Vignette
Riggs, Jessica
Aileen Tlamsa
Rutgers - Newark (Neil Kothari)

Recurrent Intractable Epistaxis in the ICU, a Case of Carotid Blowout Syndrome

Carotid blowout syndrome is not an uncommon adverse effect of post-radiated nasopharyngeal cancer patients, and is often a life-threatening medical emergency requiring rapid and appropriate critical care management. A 63 year-old veteran with squamous cell nasopharyngeal cancer status-post high-dose radiation to the primary tumor and neck, and 2 cycles 5-Fluorouracil/Cisplatin chemotherapy, was emergently transferred to the medical intensive care unit after an episode of massive nasal and oral hemorrhage onset suddenly while sitting in clinic. Since initial cancer diagnosis 10 months prior, the patient had suffered from severe intractable headaches, sensorineural hearing loss, and dysphagia. Over the course of treatment he reported several episodes of spontaneous epistaxis and hematemesis that resolved without intervention, occasionally waking him from sleep. The patient had progressive...
weight loss and intolerable fatigue, requiring hospital admission up to thrice weekly for intravenous hydration and/or blood transfusion. Comparison of post-radiation MRI to that on initial diagnosis was concerning for non-regression and hypervascularity of the mass, with extensive necrotic tissue. The patient was scheduled for rebiopsy and staging, but was detoured to the ICU before the procedure even began. In the unit, Otolaryngology specialists performed blind balloon tamponade to the anterior and posterior oropharynx, and the patient was emergently intubated for airway protection with chemical sedation and paralysis. His blood pressure was controlled to avoid shearing forces, and the patient received packed RBC and fluid resuscitation prior to transfer to a nearby facility for endovascular embolization. CT angiogram revealed maxillary artery dominance and outpouching consistent with pseudoaneurysm and carotid blowout, with stenosis of an intact internal carotid artery. Patient successfully underwent particle embolization to the suspected culprit maxillary artery, was later extubated, and on post-operative day 6 was pending discharge to home. Abruptly and without forewarning, the patient began coughing with immediate progression to pulsatile exsanguination from the oral cavity. Within minutes, he was obtunded and bradycardic with transition to asystole, and was pronounced dead at that time due to hemorrhagic shock and asphyxia. This case illustrates the unpredictable nature of carotid blowout syndrome in irradiated nasopharyngeal cancer survivors, and the importance of pre-emptive triaging in the medical care unit, coordination of endovascular intervention, and forthright discussion of advanced directives. Although a surgical emergency, it is imperative for the medical specialist to anticipate this potentially lethal complication, at any stage of oncologic treatment.

174. Clinical Vignette

Tlamsa, Aileen P. Jessica Riggs, Michael Yudd, RoseMarie Pasmantier, Allen Blaivas

Rutgers - Newark (Neil Kothari)

Early Plasmapheresis in the Treatment of Severe Hypertriglyceridemia-Induced Pancreatitis

INTRODUCTION: Severe hypertriglyceridemia (SHTG), referring to triglyceride levels >1000mg/dL, is well documented to be a critical condition requiring immediate intervention. SHTG has been reported to account for approximately 10% of episodes of acute pancreatitis, the third most common cause after gallstones and alcohol. Evidence shows these patients with SHTG suffer a higher complication rate and disease severity during pancreatitis. The use of plasmapheresis has been shown to be safe and effective in rapidly lowering triglyceride levels in patients with SHTG.

CASE PRESENTATION: This is a 33 year old Hispanic man with a medical history of familial hypertriglyceridemia, non-insulin dependent diabetes mellitus, obesity, and sleep apnea who presented to the emergency room with a chief complaint of severe abdominal pain. The patient reported he had missed doses of his medication for 5 days prior. Additionally, he reported noncompliance with dietary regimen over the preceding two months. Upon arrival to ED, the patient reported progressively worsening abdominal pain with subsequent nausea, anorexia, and vomiting. He was found to be tachycardic, diaphoretic and hypertensive with a low grade temperature. He denied chest pain but admitted to feeling severe abdominal pressure. His abdomen was diffusely tender with hypoactive bowel sounds. His exam was otherwise unremarkable. Serum chemistry revealed a triglyceride level of 9,312 mg/dL. His amylase and lipase were 342 and 375, respectively. Lactic acid was 2.01 mg/dL. CT of the abdomen and pelvis showed nonspecific inflammation of the pancreas without evidence of necrosis. Patient was initiated on continuous insulin infusion and concomitant dextrose 5% in normal saline. He was hydrated aggressively and placed on insulin infusion for enhancement of lipoprotein lipase activity, with subcutaneous heparin for synergistic effect. Decision was made for the patient to undergo plasmapheresis for rapid reduction of triglyceride level. Patient underwent plasmapheresis for 3 daily sessions with exchange of 4 liters plasma per session with 5% albumin replacement. The triglyceride level dropped by 87% after the first session, with concomitant insulin infusion and gemfibrozil 200mg twice daily. The patient improved and was discharged to home in stable condition.
DISCUSSION: This case exemplifies the utility of rapid plasmapheresis (<24 hours) in the management of SHTG, particularly during episodes of acute pancreatitis. In patients with critically high triglyceride levels, as seen in our patient, rapid reduction may not be feasible by standard interventions, including insulin infusion, heparin, and fibrate administration. Removal of triglycerides from plasma has been shown to prevent relapses of SHTG, in addition to strict dietary and medication compliance. Presently there are no guidelines for the use of plasmapheresis in the treatment of SHTG, however current practices have shown this to be a safe and effective tool.

**175. An Unusual Presentation of Small Cell Lung Cancer: Unique histology and paraneoplastic syndromes**

**INTRODUCTION:** Small cell lung carcinoma (SCLC) manifests in a variety of ways, often with distinctive paraneoplastic syndromes. We present a case of SCLC with unique symptoms and liver infiltration pattern, and dual paraneoplastic syndromes.

**CASE DESCRIPTION:** A 60 year old African American male smoker (50+ pack year history) with history of COPD and untreated hepatitis C, presented with abdominal pain, loss of appetite, and 7 kg weight loss for 3 weeks prior to admission. His physical exam was significant for hepatomegaly. Initial laboratory evaluation showed profound hypophosphatemia, hyponatremia (123 mmol/L) and liver failure (AST 170 IU/L, ALT 141 IU/L, Total Bilirubin 4.0 mg/dL, Direct Bilirubin 2.89 mg/dL, INR 1.38). Further studies showed decreased serum osmolality 256 mOsm/kg [normal 285-295], inappropriately elevated urine osmolality 966 mOsm/kg [normal 50-1050], and inappropriately elevated urine sodium concentration 79 mEq/L [normal<20], suggesting SIADH rather than hyponatremia associated with liver disease. He was Hypophosphatemic, phosphate level 0.8 mg/dL, which remained unchanged despite repletion. 24 hour urine phosphate was 1269.1 mg/24h [normal <1100 mg], fractional excretion of phosphate was measured at 82% and Fibroblast Growth Factor 23 (FGF23) was elevated at 577 RU [normal <180 RU]. Chest x-ray showed hilar lymphadenopathy and subsequent computed tomography showed a soft tissue lesion surrounding the right lower lobe bronchus with right hilar lymph node necrosis and a newly enlarged liver without focal intrahepatic lesions. Bronchoscopic biopsy showed SCLC, and liver biopsy showed sinusoidal pattern of infiltration by small cell carcinoma. Palliative treatment with Cisplatin was started. The patient’s liver function tests continued to worsen, peaking at Total bilirubin 15.3 mg/dL, Direct Bilirubin 10.8 mg/dL, AST 1580 IU/L, ALT 539 IU/L and INR 3.1. The hyponatremia and hypophosphatemia initially improved with therapy but after 48 hours, renal function began to decline and chemotherapy was stopped. Comfort care measures were started, and the patient expired 10 days after presentation. Autopsy showed a right lower lobe necrotic mass, diffuse pulmonary hemorrhage, and massive hepatomegaly with nodularity. Staining of hepatic and lung tissue was positive for FGF 23 in cancer cells.

**DISCUSSION:** While SIADH has been commonly associated with small cell lung cancer, oncogenic osteomalacia is usually seen in patients with benign tumors of mesenchymal origin, and this is the fourth case we are aware of where oncogenic osteomalacia has been associated with SCLC. Immunostaining demonstrated FGF23 production by cancer cells, never before reported in our literature search. Sinusoidal hepatic variant of small cell lung cancer is a recently recognized entity usually diagnosed postmortem, and this is the first case where both paraneoplastic syndromes have been associated with the sinusoidal variant of SCLC. Heightened clinical suspicion, earlier diagnosis and prompt initiation of therapy may play a role in improving prognosis.
A huge retroperitoneal mass, a rare neoplasm

INTRODUCTION: The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumors and malignant neoplasms that can be either primary or metastatic lesions.

CASE PRESENTATION: A 77 year old man with a history of autoimmune hepatitis treated with azathioprine and prednisone presented with fatigue and involuntary weight loss of 30 pounds for 4 months. Laboratory work-up revealed WBC 26.9, neutrophil 76%, lymphocytes 14%. A CT scan revealed a 7.0 x 5.9 x 10.3 cm retroperitoneal mass which either arose or eroded into the duodenum resulting in ulceration. The mass surrounded the superior mesenteric artery as well as the right renal artery, right renal vein and inferior vena cava; associated with abdominal pathologic lymphadenopathy. Endoscopy revealed a large mass in the 2nd portion of duodenum with significant necrosis, ulceration and friability. While the pathological report of EGD biopsy was pending, the patient experienced sudden onset of severe back pain and one episode of syncope. He then vomited a large amount of bright blood and went into cardiopulmonary arrest and CPR was unsuccessful. The patient died, 20 days after admission. The pathology was consistent with a EBV-positive B-cell lymphoproliferative disorder, with features of polymorphic B-cell lymphoma at the 2nd portion of duodenum. EBER in-situ hybridization was positive in the atypical B-cells. Whole blood EBV PCR detected (A), EBV CP: 451, EBV IgG: >=1:2560, EBV IgM: <1:10, EBV Early Ag Ab: >=1:160, EBV Nuclear Ag Ab: >=1:8, HTLV I/II Abs: Negative, HIV 1/2 Ab: Negative.

DISCUSSION: Retroperitoneal masses usually present with few local symptoms, unless the mass is large enough to compress or invade surrounding structures. Approximately 80% of retroperitoneal masses are malignant. Based on CT findings of this case, differential diagnoses include retroperitoneal soft tissue sarcoma (STS), gastrointestinal stromal tumor (GIST), metastatic lymphadenopathy, primary duodenal adenocarcinoma and primary duodenal lymphoma. Gastrointestinal lymphomas are rare, with the incidence of 0.8-1.2 per 100,000 persons per year. Primary duodenal lymphomas are extremely rare, only accounting for 0.8-2% of all gastrointestinal lymphomas. The direct association between autoimmune hepatitis and B cell lymphoma is not clear and rarely reported. Interestingly, in this case, EBER in-situ hybridization is positive in the atypical B-cells, whole blood EBV positive as well. EBV was likely secondary to prior immunosuppressive treatment for autoimmune hepatitis and may have contributed to the pathogenesis of B cell lymphoma by working as a sustained antigen drive. In summary, this was a rare huge retroperitoneal mass that was a primary duodenal lymphoma in a patient with history of autoimmune hepatitis status post immunosuppressive therapy with azathioprine and prednisone. It provides more insight in the association between autoimmune hepatitis, EBV infection and lymphoma pathogenesis.
caused by mycobacterial tuberculosis which was diagnosed postmortem.

**CASE PRESENTATION:** A 70 year old Ecuadorian woman with HTN, DM type II, ESRD on hemodialysis presented with generalized weakness, nausea, diffuse vague abdominal pain, poor oral intake and increasing abdominal girth for three weeks. Initial laboratory work showed lactate 16.4 mmol/l, bandemia 38%, albumin 1.6 g/dl, AST 160 U/L, ALT 49 U/L, INR2.3. Three months earlier her liver function tests were normal. A diagnostic abdominal paracentesis showed cloudy fluid, glucose 38mg/dl, protein 3.6mg/dl, albumin 1.0 g/dl. There were 570 RBC, 260 WBC with 86% neutrophils.PMN (polymorphonuclear leukocyte) was 223 cells/mm3. SAAG(Serum ascitis albumin gradient) was 0.6 g/dl.Chest X ray showed normal cardiomedstinal silhouette, no infiltrate or pleural effusion. The patient was treated with IV fluid, broad spectrum antibiotics for presumed sepsis most likely SBP( spontaneous bacterial peritonitis) with Vancomycin and Meropenem. Albumin was also administered for hypoalbuminemia. Ascitic fluid culture was negative. Abdominal ultrasound showed large amount of complex ascites with multiple septations and slightly nodular hepatic contour. A therapeutic paracentesis was performed on hospital day 8 with 1.8L amber fluid removed. The fluid study showed: glucose 25mg/dl, protein 4.0mg/dl, albumin 1.5g/dl. RBC was 847, WBC 594 with 54% neutrophil, ADA 6.9 U/L. The patient’s clinical condition continued to deteriorate and was made comfort care by family. The patient expired on hospital day 9. Four weeks later, the peritoneal fluid AFB culture returned positive for mycobacterium tuberculosis.

**DISCUSSION:** Peritoneal tuberculosis is an uncommon site of extrapulmonary mycobacterium tuberculosis infection. Clinical presentation can be variable. Most common clinical features are ascites, abdominal pain. Diagnosis can be very difficult since the onset is often insidious. In patients with cirrhosis, peritoneal tuberculosis is often not suspected as the ascitic fluid is often related to spontaneous bacterial peritonitis and mycobacterial culture is not routinely sent. Mortality rate is up to 50% if there is delay in diagnosis and treatment. Clues in peritoneal fluid analysis include protein >3g/dl, SAAG< 1.1g/dl if there is no underlying cirrhosis. Peritoneal fluid ADA (adenosine deaminase) measurement has a high sensitivity and specificity in non-cirrhotic patients. Its sensitivity is substantially lower in patients with cirrhosis due to impaired humoral and T cell mediated responses. Peritoneal tuberculosis should be suspected in all patients with SAAG < 1.1g/dl, especially in patients from areas endemic for tuberculosis. Early detection and treatment will certainly decrease mortality.

**SYNTHETIC CANNABINOIDS INDUCED RHABDOMYOLYSIS**

**BACKGROUND:** Synthetic cannabinoids are analogs of naturally occurring chemicals found in marijuana including THC (tetrahydrocannabinol), cannabidiol and cannabinol. Common street names are “Spice”, “K2”, “Spice Gold”, “Spice Diamond”, “Chill Out”, and “Chill X”. These compounds are classified as Class I controlled substances by the United States Drug Enforcement Administration. Synthetic cannabinoids are illegal to possess in the United States. However, illicit use remains significant. In 2012, up to 11 percent of United States high school seniors reported use of synthetic cannabinoids. Here we are reporting a case of K2-induced rhabdomyolysis.

**CASE REPORT:** A 27 year old African American man with no past medical history was found unresponsive in bed with K2 wrappers around him. The patient was initially confused, very agitated and combative. He required both chemical and physical restraints for safety concerns. Laboratory work showed WBC 26.4, lactate 5.9, creatinine 1.3, AST 84, ALT 37, creatine kinase 3173u/L. Urine toxicology was negative. The patient was sedated overnight. He woke up the next day alert and oriented x3 and did not remember what happened. He was treated with intravenous hydration and sodium bicarbonate infusion to maintain urine output >
200ml/hour. His kidney function returned to normal range one day later. However his CK continued to rise over the next two days and peaked at 136700 u/L. He had no musculoskeletal complaints and was ambulatory without any difficulty. The patient was discharged on hospital day 4 in stable condition.

**DISCUSSION:** Clinical presentation of synthetic cannabinoids use may include reddening of the eyes, tachycardia, anxiety, aggressive behavior, psychosis, and seizures. Sympathomimetic effects are often more pronounced than that observed after use of natural marijuana. Treatment is mainly supportive with intravenous fluid hydration and benzodiazepines for agitation and combative behaviors. Standard toxicology tests currently used is only able to detect naturally occurring THC and not able to detect many synthetic cannabinoid-like substances. The duration of acute synthetic cannabinoid toxicity depends upon the exposure dose. In a small case series, the duration was reported ranging from 4-24 hours. It is important to keep synthetic cannabinoids overdose in mind when treating patients with similar presentations since urine toxicology cannot help with diagnosis. Severe rhabdomyolysis and acute kidney injury can occur as a consequence. It is surprising that our patient did not report any myalgia or weakness given this high level of CK.

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**179. Quality Improvement**

**Kim, Anthony**

Leonard Genovese MD, Roopa Roy MD, Chinonye Ogbonnaya-Odor MD; Mirela Feurdean MD; Alex Tentler MD

Rutgers - Newark (Neil Kothari)

**Treating Osteopenia in a Resident Run Clinic**

**BACKGROUND:** Osteopenia is defined as a DEXA T score between -1.0 and -2.5. Many clinicians are attuned to the benefits of initiating therapy in patients suffering from osteoporosis to decrease fracture risk, but the absolute number of fractures is higher in patients with osteopenia than osteoporosis due to increase in prevalence. Therefore, a low bone mass density should not be the sole marker of fracture risk, and the use of a FRAX score, which takes into account other risk factors can be used to calculate a 10 year probability of hip and major osteoporotic fracture. Our study aims to examine if proper treatment was being offered to patients with osteopenia based on calculated FRAX score at UMDCare. Our resident run clinic.

**METHODS:** A randomized, retrospective chart review was conducted at University Hospital in Newark, NJ’s UMDCare outpatient clinic. A list was generated of 1187 patients being seen at UMDCare who had a DEXA scan ordered between January 1, 2008 and December 31, 2013. From that list, 200 patients were randomly selected. DEXA scan results were reviewed and patients were included in our usable data if their T-score was between -1.0 and -2.5. The chart was then reviewed for the following information: age, gender, T-score, osteopenia documentation and patient treatment. FRAX score was generated for each patient and the appropriateness of the treatment was assessed.

**RESULTS:** Of the 200 randomly selected patients, 81/200 had osteopenia. Of those 81 patients, 7 were lost to follow up. 43 of 74 (58%) had been appropriately treated, however, of 6 patients that needed to be on bisphosphonate therapy by FRAX score, only 2 of 6 were placed on the appropriate regimen of Calcium Vitamin D and bisphosphonate. Of note, from the 74 patients, 45 of 74 had been documented inaccurately but 22 were treated appropriately regardless with Vitamin D and Calcium.

**CONCLUSION:** We found that treating osteopenia mostly with calcium and vitamin D would be adequate. Only 8% of patients had FRAX high enough to warrant treatment with bisphosphonate therapy. However, only 33% of these patients were treated appropriately. We found that even when osteopenia was not mentioned in the chart, it was still treated appropriately simply by
Early identification of patients with osteopenia and risk factors of fracture by using the FRAX score is an easy first step to implementing more appropriate, WHO-based treatment guidelines to de-mystify the appropriate treatment of osteopenia.


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<th>Improving High Value – Cost Conscious Care in the Laboratory Workup of Thrombophilia at an Academic Institution</th>
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<td>Dina Khateeb, DO;</td>
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<td>In 2010 the American College of Physicians announced a high value cost conscious care initiative identifying practicing physicians as stewards of health care resources. Soon after, the American Board of Internal Medicine Foundation recognized thrombophilia testing as an area in medical stewardship in need of quality improvement. The patient selection, setting, and timing of tests must be considered to maximize their accuracy and utility. Current guidelines recommend against testing at the time of acute thrombosis as test results can be altered in the setting of the acute event. Common anticoagulants such as warfarin and heparinoids can alter Protein C/S and antithrombin III levels making, these tests difficult to interpret and the immediate treatment of acute thrombosis is not influenced by results. To understand how this area can be improved in our hospital, we did a retrospective chart review of all patients with thrombophilia workup over the past year and collected data regarding the setting and patient selection in consideration of accuracy and utility of thrombophilia testing. We found that over the course of 1 year, 181 patients at our hospital had laboratory thrombophilia workup. The average age of the patients was 45 (range 21 to 84). Venous thromboembolism (VTE) was the most common associated diagnosis (59%), followed by cerebrovascular accidents (21%). Of all the patients who had a thrombophilia workup, 70% of tests were completed during hospitalization, whereas 30% were ordered as outpatients. At the time of lab work, 88 patients (49%) were on anticoagulation. Physicians in Internal medicine (and subspecialties) ordered tests for 61% of the patients, followed by Neurology (15%) and OB/Gyn (8%). The remaining 16% were ordered by ophthalmology, surgery, and emergency physicians. 48 patients (33%) had at least 1 reversible risk factor for VTE, such as prolonged immobilization, recent surgery, cancer, or pregnancy. We found that the majority of our patients had thrombophilia work up during hospitalization at the time of the acute event and about half of these subjects were on anticoagulation at the time, making the results of these tests difficult to interpret. Also, 33% of the patients had at least 1 reversible risk factor for VTE, which made thrombophilia work-up unnecessary according to current best practice guidelines. Although the high value cost conscious care initiative has been implemented in 2010, there continues to be need for improvement in medical stewardship. Internal medicine and its subspecialties ordered most of the workup for thrombophilia followed by neurology at our hospital. Interventions aimed at improving timing and selection of tests can be aimed at this group.</td>
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<th>Poor adherence to AASLD guidelines for suspected NAFLD in an urban hospital: under-evaluating an overly prevalent disease</th>
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<td>Jill Deutsch MD, Henry S. Dinneen DO, Jason Zucker MD, Mirela Feurdean MD</td>
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<td>INTRODUCTION: In June 2012, the American Association for the Study of Liver Diseases (AASLD) published guidelines for the diagnosis and management of non-alcoholic fatty liver disease (NAFLD). These guidelines included a grade IA recommendation that patients with hepatic steatosis detected on imaging with either elevated liver function tests or signs and symptoms attributable to liver disease should have competing etiologies for liver disease excluded. The purpose of this pilot study was to assess our</td>
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METHODS: We performed a retrospective chart review of patients with hepatic steatosis detected on ultrasound, CT, or MRI after June 2012 seen at University Hospital in Newark, NJ. We reviewed demographic, clinical, laboratory, radiologic, and pathologic data. We included patients with elevated transaminases and/or symptoms attributable to liver disease. Patients with acute liver failure, known history of alcohol abuse/dependence, known history of viral hepatitis, or late stage malignancy were excluded. We defined a “complete” evaluation as screening for the following 7 components: alcohol use, hepatitis A, hepatitis B, hepatitis C, hemochromatosis, Wilson disease, and autoimmune hepatitis. We calculated the percentage of the complete evaluation ordered for each patient. We performed a z-test to determine if ALT elevation or the specialty of the physician seen were associated with completeness of evaluation ordered.

RESULTS: 130 patients with radiologic evidence of hepatic steatosis were identified, of which 55/130 (42%) met criteria for further evaluation. 38 patients were female and 17 male, with a mean age of 45.8 years. The mean AST was 36 (13-190) and mean ALT 47.2 (8-258). Following detection of steatosis, 19/55 (34.5%) patients were seen in our medicine clinic only and had a mean 22% of the complete evaluation ordered. 19/55 (34.5%) patients were seen in both medicine and GI clinics, with a mean evaluation of 28%. 1/55 (1.8%) patient was seen in our medicine and hepatology clinics, with 14% of the evaluation ordered. 1/55 (5.5%) patients were seen by medicine, GI, and hepatology, with a mean evaluation of 67%. 12/55 (21.8%) patients were lost to follow-up, with a mean evaluation of 13%. 1/55 (5.5%) patient had follow-up with a non-medicine subspecialty. Patients with an elevated ALT were likely to have a higher percentage of the complete evaluation ordered (39% vs. 17%, p = .0002). As expected, patients seen for follow-up had a higher percentage of the evaluation ordered than those lost to follow-up (28% vs. 13%, p = 0.0032).

CONCLUSION: These preliminary results indicate a very poor adherence to AASLD practice guidelines for the evaluation of suspected NAFLD. Although elevated ALT was associated with a more complete evaluation, it was still largely inadequate with a mean completeness percentage of only 39%.

BACKGROUND: Mild pulmonary hypertension (PH) may occur in patients with obstructive sleep apnea (OSA), even in the absence of cardiac or lung disease. There is limited data on the development and severity of pulmonary hypertension in patients with obstructive sleep apnea without underlying cardiac or lung pathology, and the response of continuous positive airway pressure (CPAP) treatment on pulmonary artery (PA) pressures.

METHODS: The Pubmed, Medline, Cochrane reviews, Central Registry of Controlled Trials, Web of Science, and EMBASE databases were searched (the latest search date: October 2014) with the following keywords: obstructive sleep apnea and pulmonary hypertension, sleep apnea, pulmonary artery pressure. The pooled mean PA pressure was 33 mmHg ± 9.7 for patients with OSA who had PH. OSA patients with PH have a mean deviation of
3.29 +/- 0.68 higher BMI than those without PH (t-value: 5.37, p<0.0001). Five studies measured PA pressures pre and post continuous positive airway pressure (CPAP) therapy. The mean reduction in PA pressures was 6.69mm/HG (95% CI -12.63, -0.74) after CPAP treatment.

CONCLUSIONS: We conclude that OSA may be associated with PH (mean PA pressures of 33 mmHg ± 9.7) even in patients without coexisting cardiovascular or lung disease and in the absence of significant daytime hypoxemia. Even patients with severe OSA were noted to have only a modest increase in PA pressures. Potential mechanisms leading to daytime PH in patients with OSA include hypoxic pulmonary vasoconstriction, hypoxia induced endothelial dysfunction, and pulmonary vascular remodeling. OSA patients with PH tend to have higher BMIs than those without PH. Pulmonary hypertension is an independent predictor of mortality in patients with obstructive sleep apnea. CPAP may reduce pulmonary vessel reactivity to hypoxia and improve pulmonary endothelial function, which may reduce PA pressures to near normal. It may potentially lead to reversal of PH in some patients, but further studies with larger sample sizes and longer duration are needed to confirm this hypothesis.

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183.

Clinical Vignette

Dsouza, Ria MD;Jeffrey Sohn;Kezia Alberto MD

Saint Barnabas Medical Center (Sunil Sapru)

Uremic Frost

INTRODUCTION: Uremic frost is a rare cutaneous manifestation, occurring in the setting of severe azotemia and found only 3% of the time in non-dialyzed advanced chronic renal failure patients. Uremic frost is a visible collection of yellow-white urate crystals, appearing when blood urea levels rise to levels greater than 200mg/dL; urate crystals excreted from eccrine glands typically deposit on the face, neck and upper extremities. Once a relatively common manifestation of advanced chronic renal failure, it is now a rare physical finding since the advent and nearly ubiquitous implementation of hemodialysis in patients with chronic kidney disease.

CASE PRESENTATION: A 52-year-old Hispanic male presented with a two-week’s history of generalized pruritus, shortness of breath, and chest pain on exertion. Laboratory analysis demonstrated hemoglobin of 5.6, creatinine (Cr) of 14 and blood urea nitrogen (BUN) of 111. His past medical history was significant for insulin dependent diabetes and hypertension (non-medicated). His BUN/Cr from one year prior to presentation was 45/3.25. Physical examination was significant for multiple tracks of excoriation with superficial bleeding, uremic fetor and white crystalline deposits found on his arms, chest and face. At the time of initial presentation, he was anemic, elevated BUN and Cr were likely evidence of advancing chronic kidney disease. Renal ultrasound showed bilateral renal cortical thinning and increased echogenicties without hydronephrosis, further supporting a diagnosis of advancing renal disease. Echocardiogram performed showed moderate concentric left ventricular hypertrophy with an ejection fraction of 60-65%, grade 1 diastolic dysfunction and a moderately dilated left atrium. Patient was dialyzed in the first twelve hours and subsequently experienced an improved electrolyte profile with a reduction in BUN and Cr of 87 and 12 respectively. Prior to dialysis, he received a transfusion and anti-hypertensives were initiated. The patient’s stay was complicated by MRSA sepsis and appropriate treatment was administered. Prior to discharge, an AV fistula was placed for permanent dialysis access. He was scheduled for outpatient dialysis three times a week. On discharge his hemoglobin had improved to 8.1, with a BUN of 38 and Cr of 7. The patient’s clinical course continued to improve with resolution of both uremic frost and fetor. He was instructed to follow-up with his nephrologist upon discharge.

CONCLUSION: The most common dermatological findings of advancing chronic renal failure include xerosis, pallor and pruritus. Other rare dermatological manifestations of severe azotemia are rarely observed today in clinical practice due to increasing use and...
early initiation of renal replacement therapy. Sweat concentration of urea dramatically increases when BUN levels are high, with deposition of urea crystals occurring with evaporation of sweat.

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<td><strong>Streptococcal Toxic Shock Syndrome in Diabetic Ketoacidosis Patient</strong></td>
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| **INTRODUCTION:** Streptococcal toxic shock syndrome (TSS) is a rare clinical condition characterized by shock, isolation of group A Streptococcus from a normally sterile body site and multi-organ failure such as acute respiratory distress syndrome, coagulopathy, liver or renal failure. It occurs due to capillary leak and tissue damage associated with release of inflammatory cytokines from T-cell lymphocytes induced by streptococcal toxins. We report a case of classic presentation of TSS with diabetic ketoacidosis.

**CASE REPORT:** A 45 years old African American male with past medical history of Diabetes Mellitus Type II, Hypertension and Hyperlipidemia presented to ER with fever, sore throat, chills, sweats, nausea, vomiting and diarrhea. He was discharged the same day with diagnosis of acute viral gastroenteritis. Two days later, he had persistent fever and generalized weakness. He was tachycardic, hypotensive with swelling and erythema of both hands. Initial blood work revealed leukocytosis with bandemia, anion gap acidosis and elevated blood sugar levels. He was admitted to ICU for management of Diabetic ketoacidosis (DKA). Clinical diagnosis of TSS was made. He was initially treated with vancomycin and clindamycin to cover both staphylococci and streptococci.

Blood culture grew colonies of group A beta-hemolytic streptococci. He also developed Adult Respiratory Distress Syndrome for which he was placed on mechanical ventilation, and Acute Renal Failure which was treated with Continuous Renal Replacement Therapy (CRRT). Coagulopathy and liver dysfunction was noted as well. He was treated with Intravenous Immunoglobulin (IVIG) for 3 days, Penicillin G and clindamycin for 10 days. Extensive work up for source of infection was negative. Subsequently his clinical status improved. He was extubated, taken off CRRT and his coagulopathy resolved. He had desquamation of skin on his hands after few days. He was stable at time of discharge to home.

**DISCUSSION:** Risk factors of TSS include diabetes, alcoholism, minor trauma and bruising, and severe streptococcal infection. No source of infection is reported in about half of patients. Renal failure is reported nearly in all patients within 2-3 days. It has a high case fatality rate of 30-60%. Early and aggressive exploration of soft tissue swelling and possible source of infection is mandatory. Once suspected, treatment is hemodynamic support with intravenous fluids, vasopressors and empirical antibiotic coverage with clindamycin plus a carbapenem or penicillin with beta-lactamase inhibitor. Once diagnosis is confirmed, treatment with Penicillin G and clindamycin should be instituted. Treatment with IVIG 1 g/kg on day one, followed by 0.5 g/kg on days two and three is recommended to increase antibody concentration in severe infection, neutralize streptococcal toxins, and inhibit T-cell proliferation causing tissue injury.

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<th>Sargis Khoobiar M.D., Sunil Sapru M.D., FACP</th>
<th>Saint Barnabas Medical Center (Sunil Sapru)</th>
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<td><strong>Infective Endocarditis of prosthetic valves – Room for Improvement</strong></td>
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| **INTRODUCTION:** Infective endocarditis, especially of prosthetic valves, remains a considerable challenge due to the associated significant morbidity and mortality. The problem is further complicated by the rise of antibiotic resistance among common pathogens and increasing reports of antibiotic prophylaxis failures. Although endocarditis of prosthetic valves is uncommon, it is a serious complication and overall mortality is as high as 59%.
CASE REPORT: We report the case of a 63 year old Caucasian gentleman with past medical history of hypertension, hyperlipidemia, coronary artery disease status post one vessel coronary artery bypass graft and bicuspid aortic valve with subsequent valve replacement with a bioprosthetic valve. He presented approximately 6 months following his aortic valve replacement surgery with a four week history of fevers with temperatures up to 102°F, progressive fatigue and myalgias. He denied any chest pain, chest pressure, discomfort or shortness of breath. He also denied palpitations, lightheadedness or syncopal events. Upon further questioning, the patient revealed that he had a dental procedure performed recently. He was not informed nor given antibiotic prophylaxis prior to the dental work. His physical exam was unremarkable except for a Grade II/VI systolic murmur and a grade I/VI diastolic murmur appreciated best at the aortic area. There was no evidence of splinter hemorrhages, Osler’s nodes or Janeway lesions. Lab work was significant for an elevated ESR of 79 mm/hr and CRP at 4.17 mg/L. Blood cultures were positive for Streptococcus anginosus. An initial transthoracic and transesophageal echo (TEE) was negative and the patient was started on IV ceftriaxone. His course was complicated by a transient slurred speech and drooling from right side of mouth. MRI of the brain was positive for a small infarct of the left anterior insula. This was consistent with an embolic event and a repeat TEE revealed a 0.5 cm mobile homogeneous echo dense mass from the annulus at the non-coronary cusp position consistent with vegetation. He was offered a valve repair surgery but the patient refused and he continued medical management with IV ceftriaxone for six weeks. However, a repeat TEE revealed that the mass had grown to 1.6 cm following which he undergone surgical valve repair.

DISCUSSION: The importance of prophylaxis and prompt identification of infective endocarditis cannot be emphasized enough as it has a profound effect on morbidity and mortality. Data suggests that most cases of endocarditis are due to inappropriate prophylaxis use rather than antibiotic failures. Therefore, a multidisciplinary approach is necessary including the involvement of dentists and primary physicians, who must be educated on the updated ACC guidelines for prophylaxis, diagnosis, and treatment of infective endocarditis.

**Clinical Vignette**

**Jiang, Li**

Saint Barnabas Medical Center (Sunil Sapru)

**Chronic Autoimmune Vestibulopathy Improved with Hydroxychloroquine and Steroids**

The most common causes of vestibulopathy are ototoxic aminoglycosides, Meniere’s disease and meningitis. Reports on autoimmune vestibulopathy are rare. We present a case of autoimmune vestibulopathy in a 32 year old African-American woman with severe intermittent chronic vertigo and dizziness for fourteen months that improved with steroids and hydroxychloroquine. The dizziness and vertigo affected her daily life and work, sometimes associated with headache and nausea but not with hearing loss, weakness, numbness or tingling. There was no history of antibiotic use or head trauma. Past medical history included Vitamin D deficiency and iron deficient anemia. Physical exam and routine labs such as complete blood count with differential and comprehensive metabolic panel were unrevealing. Treatment was initiated with meclizine and vestibular physical therapy without improvement. Dizziness improved briefly but recurred one week after she was started on topiramate 15 mg bid. The patient was then referred to neurology and rheumatology. Rheumatoid factor, cyclic citrullinated peptides, anti-neutrophil cytoplasmic antibodies, antiphospholipid antibodies, beta 2-glycoprotein-antibody, anti-jo-1 antibody, complement C3 and C4, anti-ds DNA antibodies, antiscleroderma-70 antibodies, CPK, TSH and RPR were within normal limits; erythrocyte sedimentation rate was elevated at 48 mm/hr; C-reactive protein was elevated at 6.5 mg/L; antinuclear antibodies was 1:80; anti-SSA was positive and anti-SSB was negative. Sialogram was unremarkable. MRI of the brain with and without intravenous contrast revealed non-specific nodular hyperintensities in bifrontal and left frontal parietal subcortical white matter and normal bilateral seventh and eighth intracranial nerve complex. Electronystagmogram and rotator chair test confirmed bilateral central vestibular dysfunction and left
Peripheral vestibular dysfunction. Autoimmune vestibulopathy was diagnosed and she was started on hydroxychloroquine 200mg daily. Methylprednisolone 64 mg per day was added tapering the dose 8 mg every 5 days until 16 mg daily within 4 weeks. She was also placed on calcium and vitamin D. Four weeks later the patient reported significant improvement of dizziness and complete resolution of vertigo, remaining stable on hydroxychloroquine for twelve months so far. Autoimmune vestibulopathy is a relatively rare condition. The diagnosis can be difficult as there are no universally accepted diagnostic criteria or diagnostic tests. Our patient had only isolated signs and symptoms of bilateral vestibulopathy. An auto-immunological etiology was likely based on the fact that other causes had been excluded and the good response to steroids and hydroxychloroquine. The treatment trials on autoimmune inner ear disorders that have been published have focused on hearing loss. Very few cases on isolated autoimmune vestibulopathy were reported in the past. From the clinical course and response to treatment of our patient, we conclude that a short term treatment trial of steroids can be diagnostic for autoimmune vestibulopathy and should be started early on to preserve or even improve vestibular function.

Clinical Vignette

Jiang, Li, Carlino, Anthony, MD, Sapru, Sunil, MD, Xiong, Wen, MD, Saint Barnabas Medical Center (Sunil Sapru)

Autoimmune Vestibulopathy Improved with Hydroxychloroquine and Steroids

The most common causes of vestibulopathy are ototoxic aminoglycosides, Meniere’s disease and meningitis. Reports on autoimmune vestibulopathy are rare. We present a case of autoimmune vestibulopathy in a 32 year old African-American woman with severe intermittent chronic vertigo and dizziness for fourteen months that improved with steroids and hydroxychloroquine. The dizziness and vertigo affected her daily life and work, sometimes associated with headache and nausea but not with hearing loss, weakness, numbness or tingling. There was no history of antibiotic use or head trauma. Past medical history included Vitamin D deficiency and iron deficient anemia. Physical exam and routine labs such as complete blood count with differential and comprehensive metabolic panel were unrevealing. Treatment was initiated with meclizine and vestibular physical therapy without improvement. Dizziness improved briefly but recurred one week after she was started on topiramate 15 mg bid. The patient was then referred to neurology and rheumatology. Rheumatoid factor, cyclic citrullinated peptides, anti-neutrophil cytoplasmic antibodies, antiphospholipid antibodies, beta 2-glycoprotein-antibody, anti-jo-1 antibody, complement C3 and C4, anti-ds DNA antibodies, antiscleoderm-70 antibodies, CPK, TSH and RPR were within normal limits; erythrocyte sedimentation rate was elevated at 48 mm/hr, C-reactive protein was elevated at 6.5 mg/L, antinuclear antibodies was 1:80; anti-SSA was positive and anti-SSB was negative. Sialogram was unremarkable. MRI of the brain with and without intravenous contrast revealed non-specific nodular hyperintensities in bifrontal and left frontal parietal subcortical white matter and normal bilateral seventh and eighth intracranial nerve complex. Electronystagmogram and rotator chair test confirmed bilateral central vestibular dysfunction and left peripheral vestibular dysfunction. Autoimmune vestibulopathy was diagnosed and she was started on hydroxychloroquine 200mg daily. Methylprednisolone 64 mg per day was added tapering the dose 8 mg every 5 days until 16 mg daily within 4 weeks. She was also placed on calcium and vitamin D. Four weeks later the patient reported significant improvement of dizziness and complete resolution of vertigo, remaining stable on hydroxychloroquine for twelve months now. Autoimmune vestibulopathy is a relatively rare condition. The diagnosis can be difficult as there are no universally accepted diagnostic criteria or diagnostic tests. Our patient had only isolated signs and symptoms of bilateral vestibulopathy. An auto-immunological etiology was likely based on the fact that other causes had been excluded and the good response to steroids and hydroxychloroquine. The treatment trials on autoimmune inner ear disorders that have been published have focused on hearing loss. Very few cases on isolated autoimmune vestibulopathy were reported in the past. From the clinical course and response to treatment of our patient, we conclude that a short term treatment
trial of steroids can be diagnostic for autoimmune vestibulopathy and should be started early on to preserve or even improve vestibular function.

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<th>Khan, Nasir</th>
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<th>Lyme Disease: The New Great Imitator?</th>
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<td>INTRODUCTION: Lyme disease is a multisystem vector-borne illness caused by the spirochete Borrelia burgdorferi. The disease manifestations owe not only to localized or disseminated infection, but also to the host immune response. B. burgdorferi is a highly neurotropic organism and causes a wide spectrum of central nervous system (CNS) derangements. Cerebral vasculitis is a rare complication of Lyme disease (0.3% of neuroborreliosis cases). We describe a case of Lyme disease induced CNS vasculitis.</td>
<td>CASE: A 30 year old caucasian female landscaping consultant frequently working in the Atlantic Northeast presented to Saint Barnabas Medical Centre with a five day history of gradual onset generalized headache, nausea, mild photophobia and right upper extremity tingling. Routine laboratory tests were within normal limits except for an ESR of 68 mm/hr and C-Reactive Protein of 8 mg/dl. Extensive head and neck imaging studies including computed tomography, magnetic resonance (MR) imaging and MR-angiography of the head and neck vessels were unrevealing. ELISA assay for Lyme disease was positive and the result was confirmed by Western blot analysis. CSF analysis showed pleocytosis; no organisms were noted on microscopy and PCR analysis of the CSF for Lyme disease was negative. The autoimmune workup including ANA titer, Compliment levels (CH50) and ACE levels were all within normal limits. The patient was started on intravenous Ceftriaxone and high dose steroids. Subsequently, a diagnostic cerebral angiogram confirmed small and medium vessel CNS vasculitis. Her symptoms responded rapidly to high dose steroids and she was discharged home asymptomatic on antibiotics and steroids.</td>
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<td>Clinical Vignette</td>
<td>Melikian, Adrien</td>
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<td>Saint Barnabas Medical Center (Sunil Sapru)</td>
<td>Migraine or Moyamoya?</td>
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<td>Moyamoya disease (MMD) is a rare cerebral vasculopathy characterized by bilateral stenosis or occlusion of the arteries around the Circle of Willis. The lenticulostrate arteries subsequently develop an extensive collateral circulation which resembles a “puff of smoke” (moyamoya in Japanese) on angiography. MMD predominantly affects patients of Asian heritage, with increasing detection of the disease throughout the world including American and European populations. Females are affected nearly twice as often as males. Patients with MMD frequently present with ischemic or hemorrhagic stroke, transient ischemic attacks, or seizures, however, headache is also a common manifestation and can often be a misleading presentation. We present a 54-year-old female of Asian descent with known long-standing history of MMD who presented to the Emergency Department with severe headache associated with recurrent emesis. She was hemodynamically stable with no focal deficits noted on neurological exam. A non-contrast head computed tomography was obtained and the preliminary results described no acute pathology. She was discharged home after</td>
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receiving supportive therapy. The patient was called back to the hospital the following day after expert review of the imaging revealed a mild hyperdensity of the right frontal midline region, suspicious for a subarachnoid hemorrhage. The patient was admitted to the intensive care unit and underwent cerebral angiography. Angiography illustrated absent left middle and anterior cerebral artery, multiple small abnormal vessels at the M1/A1 junction, and a collateral filling vessel between the right and left anterior cerebral artery, corresponding with the area of subarachnoid hemorrhage. All findings were consistent with moyamoya disease. No intervention could be made and the patient was managed medically. Headache associated with moyamoya disease is a recognized phenomenon; however, we must maintain a high suspicion of intracerebral hemorrhage as the presentation may be equivalent. There is no cure for moyamoya disease. Secondary prevention in those with symptomatic disease largely depends upon surgical revascularization techniques. Despite the equivocal evidence, anti-platelet agents are used in those who are asymptomatic, have mild ischemic symptoms, or are poor surgical candidates.

**Subcutaneous Coil Placement in a patient with failed Defibrillation testing**

**INTRODUCTION:** Implantation of an implantable cardioverter-defibrillator (ICD) is a recognized method of prevention of sudden cardiac death in patients at risk for malignant ventricular arrhythmias. Post-implant defibrillation threshold (DFT) testing is done to affirm that shock energy required to terminate arrhythmia is safely under the maximum output of the device. Some high risk patients e.g. patients with extremely low Left Ventricular Ejection Fraction (LVEF), high left-ventricular size or mass have high DFTs. In such patients a variety of measures are tried to lower DFT including changing polarity of shock vector, adjustment to tilt of Biphasic shock vector, usage of single vs dual coil, changing the position of Right Ventricular (RV) coil to apical or anterior interventricular position, addition of a new lead and implanting it under the skin/ in the azygous vein or in the coronary sinus. We are presenting a case where an additional subcutaneous electrode was successfully used to lower DFT.

**CASE REPORT:** Our patient is a 40 year old male with Coronary Artery disease status post coronary artery bypass grafting in 2007, hypertension, hyperlipidemia, Type II diabetes mellitus, End Stage Renal Disease on hemodialysis, old right bundle branch block, NYHA Stage III Congestive Heart Failure (CHF). He is a former smoker and a former heavy drinker. The patient was found to be having recurrent episodes of CHF exacerbation and found to have a LVEF of 25-30% on Echocardiogram. He was found to be having a former smoker and a former heavy drinker. The patient was found to have a normal sinus rhythm and had elevated DFT. He had to be given a rescue shock externally. It was then decided to implant a subcutaneous coil/electrode to attempt to decrease the patient’s DFT. During the procedure, a curved tunneling rod introducer was advanced through the standard infraclavicular incision into the subcutaneous tissue until the tip rested lateral to the spine across the midline. Then the Subcutaneous electrode was advanced to this position through the introducer and then connected to the ICD device/can. On repeat DFT testing the device was able to successfully shock the patient out of induced Ventricular fibrillation.

**CONCLUSION:** Thus, in patients with an elevated DFT post-ICD insertion, it is possible to implant an additional subcutaneous array electrode in an attempt to lower the DFT. Former studies have proved that such a solution offers a mean DFT reduction of 20% to 60%, depending on the electrode model.

**Infertility: A Rare Presentation of Tuberculosis**

**INTRODUCTION:** Implantation of an implantable cardioverter-defibrillator (ICD) is a recognized method of prevention of sudden cardiac death in patients at risk for malignant ventricular arrhythmias. Post-implant defibrillation threshold (DFT) testing is done to affirm that shock energy required to terminate arrhythmia is safely under the maximum output of the device. Some high risk patients e.g. patients with extremely low Left Ventricular Ejection Fraction (LVEF), high left-ventricular size or mass have high DFTs. In such patients a variety of measures are tried to lower DFT including changing polarity of shock vector, adjustment to tilt of Biphasic shock vector, usage of single vs dual coil, changing the position of Right Ventricular (RV) coil to apical or anterior interventricular position, addition of a new lead and implanting it under the skin/ in the azygous vein or in the coronary sinus. We are presenting a case where an additional subcutaneous electrode was successfully used to lower DFT.

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**CONCLUSION:** Thus, in patients with an elevated DFT post-ICD insertion, it is possible to implant an additional subcutaneous array electrode in an attempt to lower the DFT. Former studies have proved that such a solution offers a mean DFT reduction of 20% to 60%, depending on the electrode model.
INTRODUCTION: In the United States (US), reported tuberculosis (TB) cases have declined from 1993-2013. In 2013, foreign-born persons accounted for the majority of all reported cases. Female genital tuberculosis is rare; it usually presents in patients undergoing evaluation for infertility. We present a case of a 29 year old woman who presented with genital tuberculosis.

CASE: A 29 year old Indian woman, who immigrated to the US six years ago, underwent in vitro fertilization (IVF) treatment for infertility in New Jersey. After four unsuccessful cycles of IVF, an endometrial biopsy was performed that revealed granulomatous pathology. A subsequent chest X-ray showed upper lobe infiltrates and cavitory lesions and she was admitted to Saint Barnabas Medical Center for presumed TB. Her review of systems was negative and her physical examination was unremarkable. She was afebrile throughout her admission and her first two AFB sputum smears were negative. Upon questioning, she recalled being ill three years earlier, with persistent high fevers that lasted three to four weeks. A local physician treated her with a fluoroquinolone antibiotic for three to four weeks with resolution of her fever. Additional workup included a Chest CT scan that confirmed the cavitory lesions and documented calcified mediastinal, axillary lymph nodes and splenic calcifications. A BAL with biopsy was performed and she was then started on standard four drug TB therapy. BAL smears were AFB smear and MTB-PCR negative. Biopsy revealed non-caseating granulomas. She was discharged home on directly observed therapy with a diagnosis of probable tuberculosis. A few weeks later, all sputum cultures grew Mycobacterium tuberculosis, drug-susceptible.

DISCUSSION: In 2013, 9582 cases of TB were reported in the US. Genital tuberculosis accounts for only about 1-2 % of all reported cases and usually presents with infertility. Diagnosis is usually established by culture (often at another site) and tissue biopsies. Treatment is the standard treatment for TB. Post treatment fertility results are disappointing; the most effective method is IVF with embryo transfer. Several clinical points about this case are important to stress: 1) Genital TB needs to be considered as a cause of infertility in a foreign-born patient who is undergoing an infertility evaluation; 2) A foreign-born patient with a protracted fever needs to be carefully evaluated for TB and should not be treated with a fluoroquinolone antibiotic. When used alone, this antibiotic has been shown to delay the diagnosis of TB by partially treating the patient and promoting smear negative tuberculosis.


Euglycemic Diabetic Ketoacidosis (DKA) is characterized by serum glucose < 200mg/dl, anion gap metabolic acidosis, and ketonemia. Although it is extremely uncommon in patients with type II Diabetes Mellitus (DM II), the advent of new classes of drugs may result in more frequent occurrence of this complication. Canaglifozin, a Sodium Glucose Co-Transporter (SGLT2) inhibitor approved as a first line agent for treatment of DM II in 2013, lowers serum glucose levels by increasing its excretion in the urine. We describe a patient on Canaglifozin who developed euglycemic DKA. Case: A 31 year old lady with past medical history significant for DM II being treated with metformin, glimepiride and canagliflozin was admitted for robotic subtotal pancreatectomy for pancreatic mucinous cystadenoma. She was switched from her oral medications to insulin prior to the surgery. On day 2 of the surgery, she had poor oral intake secondary to abdominal pain and was persistently tachycardic. Blood work revealed profound anion gap metabolic acidosis. An urgent diagnostic laparoscopy didn’t show any significant surgical findings. Serum lactic acid and creatinine levels were steadily normal. Her serum glucose was in the range of 150-200mg/dl. However, urinalysis showed ketonuria with 1:25 dilution positivity.
and 1000+ glucose. She was suspected to have euglycemic DKA. She was treated with aggressive intravenous fluids and insulin with close monitoring of her metabolic panel. Her serum glucose levels remained below 200mg/dl. She was then switched to subcutaneous insulin after resolution of metabolic acidosis and was discharged home in stable condition. Her HbA1c was noted to be 10.1%, suggesting poor glycemic control. Discussion: Euglycemic DKA was first reported by Munro et al in 1973. However, in most of these patients, urine glucose was either low or absent. Ireland et al identified a subgroup of patients with Euglycemic DKA who had massive urinary loss of glucose. Since ketoacidosis is generally regarded as the metabolic outcome of excessive gluconeogenesis coupled with increased fatty acid release, it seems that euglycemia in these patients results from greater urinary loss. As Canaglifozin causes glucosuria by working independent of insulin, it can lead to lower serum glucose levels than would be expected in patients with DKA. Euglycemia does not rule out DKA in these patients and a high index of suspicion is warranted. Physicians should be aware of the mechanism of action of newer anti diabetic medications as patients may have normal glucose levels yet still have life threatening ketoacidosis. Assessment of their acid base status is crucial for timely detection and proper treatment.


A demyelinating disorder masquerading as Multiple sclerosis

INTRODUCTION: Neuromyelitis Optica (NMO) and Neuromyelitis Optica spectrum disorder (NMOSD) are uncommon idiopathic autoimmune demyelinating disorders that affect the spinal cord and optic nerve. Although NMOSD is often misdiagnosed as progressive multiple sclerosis (MS), the disease is distinct from MS and carries a worse prognosis. Early diagnosis and appropriate management might be associated with a better outcome.

CASE PRESENTATION: Thirty year old lady without any past medical history, presented with progressively worsening low back pain, bilateral lower extremity weakness and numbness for 6 days. The numbness progressed to involve her perianal and genital areas and eventually she developed urinary retention. She denied any febrile illness, recent vaccination, travel or sick contact. No family history of any neurologic diseases. Neurological exam revealed increased tone in the lower extremities with power 3/5 in all muscle groups; all reflexes were absent. Sensory level was established at T10. Blood work was negative for any infectious or autoimmune disease. No signs of heavy metal poisoning or micronutrient deficiency were evident. MRI of the spinal cord showed diffuse mild hyperintensity in the central portion of the lower thoracic spinal cord. CSF studies showed WBC of 122 with 85% lymphocyte, elevated protein, and no oligoclonal bands. High dose of steroid didn’t improve her condition. Subsequently, she complained of hearing difficulty and a repeat MRI of brain revealed a demyelinating lesion in the left cerebellar peduncle. She received IVIG and plasmapheresis but the spinal cord lesions continued to spread proximally. The CSF Anti NMO antibody was positive. She was started mycophenolate mofetil with a modest improvement in her symptoms over the next six months.

DISCUSSION: NMO and NMOSD usually present with optic neuritis, myelitis and positive anti NMO antibodies. About 15% of patients present with demyelinating lesions involving other areas of the brain. The revised diagnostic criteria for NMO require the presence of optic neuritis, myelitis and 2 out of 3 supportive criteria: Spinal cord MRI with contiguous signal abnormalities extending over 3 or more vertebral segments; MRI of the brain with normal findings or with findings not consistent with MS; and NMO-IgG seropositivity. NMOSD is defined as NMO-IgG–seropositive status in association with a clinical syndrome compatible with a limited
form of NMO. The prognosis for recovery of vision and muscle power in NMOSD is often poor when compared with MS. Initial therapy with high dose steroid is recommended. Resistant cases respond to plasmapheresis in about 50% of cases. Immunosuppression with other agents like mycophenolate or Azathioprine is the last resort.

CONCLUSION: NMO and NMOSD should always be considered in the differential diagnosis of demyelinating disorders. Early diagnosis and prompt management are crucial and appropriate treatment might be life changing in patients affected with this dreadful disease.

194. Clinical Vignette
Saha, Biplab
Mistry, Nirav Sapru, Sunil Saint Barnabas Medical Center (Sunil Sapru)
Cancer related Thrombotic Microangiopathy: A deadly disease

INTRODUCTION: Thrombotic microangiopathy (TMA) is a group of disorders characterized by microangiopathic hemolytic anemia, thrombocytopenia and microvascular thrombosis. Cancer related TMA is an uncommon but potentially fatal disease in cancer patients. This form of TMA differs from thrombotic thrombocytopenic purpura (TTP) or hemolytic uremic syndrome (HUS) in clinical presentation and it is not associated with ADAMTS-13 deficiency. Survival is weeks to months after diagnosis.

CASE PRESENTATION: Fifty two year old female with history of widely metastatic breast cancer on paclitaxel, presented to the emergency room with complaints of worsening lightheadedness, dyspnea on exertion, and fatigue for the past 2 weeks as well as hematuria for 2 days. She denied chest pain, palpitation, recent weight change, cold intolerance or focal neurological symptoms. The patient had received her last dose of chemotherapy 2 weeks ago. Her vital signs revealed BP 126/52, pulse 132, respiratory rate 22, temperature 99.7 and oxygen saturation 98% on room air. Physical examination was significant for marked conjunctival pallor and tachycardia. The rest of the examination was within normal limits. Laboratory data showed hemoglobin of 3.1, WBC 9.8, platelet 31000, INR 1.2, PTT 24, BUN 18, creatinine 0.8, reticulocyte 19, low haptoglobin, elevated fibrinogen, LDH 3021 and a negative coomb’s test. Peripheral blood film showed more than 50% schistocytes in a high power field. Urinalysis was positive for large amount of blood with very few RBCs on microscopic examination. The patient was started on corticosteroid for treatment of acute hemolytic anemia. She had received 3 units of PRBC transfusion with repeat hemoglobin of 7.3. Subsequently, the repeat hemoglobin was 5.3 and the platelet count dropped further to 8000. Plasmapheresis was started for thrombotic microangiopathy. The patient received 2 cycles of plasmapheresis. Eventually, she became febrile with altered mental status. Plasmapheresis was discontinued due to massive hemolysis and coagulopathy. Patient received maximal medical management but despite all efforts, the patient died within a week of admission.

DISCUSSION: Among solid tumors, TMA is most common in metastatic gastric, breast, prostate and lung cancer. Lymphoma and Myeloma are also known to cause TMA, and it may be the initial presentation of occult malignancy. The extent of hemolysis and resultant LDH elevation are usually much worse than in TTP or HUS. Since there is no deficiency of ADAMTS-13, this condition is usually refractory to plasmapheresis, steroids, or immunotherapy. Initiation of chemotherapy might be associated with better outcome and survival.

CONCLUSION: Although uncommon, TMA is associated with a very high mortality rate. As the clinical course is different from other thrombotic microangiopathies and prognosis is extremely poor, early recognition and initiation of chemotherapy, in addition to conventional management, might be lifesaving.
Clinical Vignette
Saraceno, Leonardo
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Saint Barnabas Medical Center (Sunil Sapru)

A Case Report of Dysautonomia: Guillain-Barré Syndrome or Sjögren’s Autonomic Neuropathy?

INTRODUCTION: Acute dysautonomia can be a manifestation of both Guillain-Barré Syndrome (GBS) and Sjögren’s Syndrome. Both diseases can cause injury to nerves regulating the coordination of heart rate, blood pressure, respiration, and gastric motility.

CASE PRESENTATION: A previously healthy 40-year old woman presented to the emergency department for abdominal pain and diarrhea. Routine laboratory studies and an abdominal CT scan were unrevealing. She was sent home with analgesics for putative gastroenteritis. She returned to emergency department less than a week later owing to syncopal episodes, persistent abdominal symptoms, new onset urinary frequency, sore throat with intermittent fevers, diffuse paresthesia and a headache. On exam, the patient was afebrile, tachycardic and became hypotensive upon standing which induced a syncopal event. Neurological exam revealed anisocoria. There was decreased sensation to light touch over the entire face, upper and lower extremities with decreased reflexes throughout. The abdomen was soft with mild tenderness. Initial lab work was negative for leukocytosis and electrolyte abnormalities. The initial cerebrospinal fluid (CSF) analysis was unremarkable with 0 cu/mm white cells and 33 mg/dl protein. CT of head and neck and MRI of the brain were negative for acute pathology and EEG was negative for seizures. Further lab studies revealed she had positive serum anti-SSA >8 AI, and positive ANA of 1:360. Repeat CSF analysis eight days later demonstrated an increased protein level of 129 mg/dl. Orthostatic hypotension persisted despite treatment with midodrine and fludrocortisone. Laryngoscopy demonstrated sensory loss of the larynx. Peripheral nerve conduction studies of her extremities showed sensory demyelinating peripheral polyneuropathy. She was initially treated with doxycycline, ampicillin-sulbactam and acyclovir for a presumed underlying central infection; antibiotic therapy was discontinued after negative cultures and titers. She received a five day course of intravenous immunoglobulins with no improvement and was eventually discharged to rehab. She was readmitted to the hospital days after discharge from a rehabilitation facility for persistent symptoms and was treated with plasmapheresis. Eventual sural nerve biopsy revealed severe axonal degeneration.

DISCUSSION: Guillain-Barré and Sjögren’s Syndrome may both cause sympathetic, parasympathetic and enteric system dysfunction. In this case of severe autonomic dysfunction, there was evidence supporting both diagnoses. It is important that repeat CSF analysis be performed since protein levels are normal when symptoms are present for less than 48 hours. It is also suggested patients with primary Sjögren’s Syndrome be tested for autonomic neuropathy.

Clinical Vignette
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Saint Barnabas Medical Center (Sunil Sapru)

Successful Bundled Treatment of Streptococcal Toxic Shock Syndrome

INTRODUCTION: Infection by highly invasive Group A Streptococcus (GAS) can give rise to Streptococcal Toxic Shock Syndrome (TSS) which manifests with shock and multiple organ failure. Although it presents with clinical symptoms identical or similar to Staphylococcal TSS, Streptococcal TSS is typically a more severe disease with a mortality rate of 30-60 %. Early recognition and timely bundled treatment with specific antibiotics, IVIG infusion and supportive care are critical to decrease mortality and morbidity.

CASE PRESENTATION: A 48 year old man with a medical history of diabetes mellitus type II, hyperlipidemia, and hypertension was admitted to the hospital with two days history of bilateral hand swelling and feeling unwell. On exam, his pulse rate was 119/min,
respiratory rate 30/min, blood pressure 123/67 mm Hg, and he was febrile to 38.4 °C. Blood glucose was 453 mg/dL and positive for ketones. He was admitted for infection induced diabetic ketoacidosis to the intensive care unit, where he received insulin therapy and ceftriaxone plus clindamycin treatment, in addition to aggressive hydration with normal saline. Despite fluid repletion, renal function declined from his normal baseline; muddy brown casts were present in the urine and anuria followed. Creatinine was elevated at 1.78 mg/dL. He also developed a coagulopathy with an INR of 1.6. Other laboratory values were within normal limits. Blood cultures at 36 hours were positive for Group A Streptococcus supporting the diagnosis of Steptococcal TSS. He immediately received IVIG at 1 gram/kg. Antibiotics were changed to penicillin G 3 million units every 4 hours with clindamycin 900 mg every 8 hours. Overnight, he developed respiratory failure with ARDS like changes on CXR and was intubated. His creatinine further elevated to 5.13 mg/dL and anuria persisted. He was started on continuous renal replacement therapy. IVIG was continued for two more days after which he was successfully extubated and hemodynamically improved. Exfoliation of skin was observed at 10th day of admission.

DISCUSSION: In the spectrum of diseases caused by GAS, Streptococcal TSS is associated with the highest mortality. The pathogenesis of TSS owes to Streptococcal exotoxins. The GAS infection was confirmed by blood culture. Evidence for the use of IVIG in the management of Streptococcal TSS is limited due to its low prevalence, however multiple groups have the efficacy of IVIG infusion in decreasing Streptococcal TSS mortality with an odd ration from 2 to 8.3.

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Clinical Vignette
Takavarasha, Shandurai
Saint Barnabas Medical Center (Sunil Sapru)
Rapid manifestation of Refeeding Syndrome with Dronabinol Use: A Case Report
Dronabinol is a cannabinoid well known as an orexigenic agent in the treatment of cancer-associated anorexia – while not as effective as megestrol, it has the added benefit of helping control chemotherapy-induced nausea and vomiting. Given the side effect profile of Megestrol, the use of dronabinol is becoming increasing popular. We present a case report illustrating the potentially fatal adverse reaction to use of dronabinol in treating cancer-associated anorexia. ME is an 80 year old woman with metastatic papillary serous ovarian adenocarcinoma being managed with Topotecan – a topoisomerase inhibitor, with her last infusion being 5 days prior to presentation to the ED. She was admitted with weakness and dizziness secondary to a 3 day history of diarrhea, accompanied with nausea and vomiting leading to decreased oral intake. Initial workup showed hypotension responsive to intravenous fluid hydration. Lab work was significant for potassium of 2.9 and Hb of 6.8 (baseline 8.0). Despite symptomatic improvement following packed red blood cell infusion, and potassium repletion and resolution of diarrhea, her hospital course was complicated by continued poor oral intake attributed to poor appetite. Patient was noted with total serum protein of 4.5 mg/dL, with an albumin level of 1.5g/dL. Metabolic consult was obtained and decision made to initiate dronabinol 2.5mg twice daily prior to meals. The patient had experience with dronabinol as an antiemetic prior to chemo infusion with no ill effects. She demonstrated rapid appetite improvement on day1of dronabinol use resulting in increased oral caloric intake. On day 2, she reported weakness, lightheadedness and palpitations. Telemetry showed multiple episodes of nonsustained ventricular tachycardia. Lab work revealed gross electrolyte derangement consistent with refeeding syndrome: Phosphorus 0.4mg/dL, magnesium 0.6mEq/L, Potassium of 3.3mEq/L. Aggressive intravenous electrolyte repletion was initiated, along with thiamine infusion and multivitamin support. Refeeding syndrome manifests as severe electrolyte and fluid shifts associated with metabolic abnormalities upon refeeding of malnourished patients, with incidences as high as 25% in cancer patients who are nutritionally supported. The electrolyte disturbances involve intracellular shifts of mainly phosphorus, magnesium and potassium and can result in potentially lethal cardiac arrhythmias as well as other serious clinical consequences including neurological and respiratory complications. Refeeding syndrome is a well described
phenomenon, though often under recognized, particularly in those patients who are being refeed orally rather than via parenteral or enteral means. Although it usually occurs within several days to a couple of weeks of refeeding, this case is atypical in that it manifested after one day of initiating dronabinol as an appetite stimulant. This case teaches us that it is important to be aware of Refeeding Syndrome and prepare for potential complications particularly following dronabinol use.

Corticosteroids for the Treatment of Bleomycin Lung Toxicity

INTRODUCTION: Bleomycin is used to treat a number of solid organ malignancies (eg. Hodgkin’s lymphoma and testicular cancer). It accumulates in the skin and lungs and can cause skin ulceration and pulmonary toxicity; bleomycin induced pneumonitis (BIP) is the most dreaded complications. The incidence of pulmonary toxicity is 10% with 1-2% fatality. Standard therapy includes discontinuation of bleomycin and a trial of steroids. We present a case of bleomycin-induced pneumonitis (BIP) that was unsuccessfully treated with high dose steroids.

CASE REPORT: A 32 year old woman with Hodgkin’s lymphoma treated with six cycle ABVD therapy presented to ED with a three week history of non-productive cough and dyspnea. For the sixth cycle, bleomycin was withheld owing to suspicion of BIP and oral steroids were initiated. On physical examination, she was dyspneic. BP was 110/70 mmHg, pulse 100/m, respiratory rate 28/min and oxygen saturation 88% on room air. Lung exam revealed crackles at the bases bilaterally. A fiberoptic bronchoscopy with BAL was performed; Gram stain, bacterial and fungal cultures, acid fast bacilli smear were unremarkable. Moreover, there was no evidence of malignancy. CTA of the chest showed diffuse airspace opacity, but no evidence of pulmonary embolism. The patient was treated with high dose IV methylprednisolone (1000mg/d in divided doses), broad spectrum antibiotics with antifungals and antiviral. The patient expired after 12 days of mechanical ventilation and ECMO support.

DISCUSSION: Bleomycin, originally isolated from the fungus Streptomyces verticillus is an antibiotic agent with antitumor activity. Bleomycin exerts its antitumor effect by inducing free radicals leading to tumor cell death and inhibiting tumor angiogenesis. Bleomycin use is somewhat limited due to toxicity primarily involving the lungs and the skin. Directly after administration, fever, chills and hypotension are not uncommon, however the most feared and dose limiting side effects are due to pulmonary toxicity. The risk factors for BIP include advanced age, high cumulative dose, renal impairment, bolus administration, smoking and giving concurrent high flow oxygen and radiotherapy. Our patient was a younger female who received moderate doses of bleomycin (10 units/m2), was a nonsmoker and did not receive radiotherapy. Clinically our patient had a presentation 3 weeks after the last dose of bleomycin, with a sub acute presentation and typical physical exam findings and gas exchange abnormalities. There are no controlled studies of use of glucocorticoids for the treatment of BIP although case reported have described substantial recovery when a significant inflammatory pneumonitis was present. Bleomycin Lung toxicity remains a major limitation and cause of mortality and morbidity. One should always be vigilant of its onset and initiate aggressive steroid and supportive treatment promptly.


Sulfonylurea induced severe hypoglycemia in a diabetic with renal failure - A case report.
Renal insufficiency is a concerning risk factor for hypoglycemia in diabetic patients on sulfonylurea therapy. A case of severe hypoglycemia in a diabetic with acute on chronic renal insufficiency while on glimeperide is presented here.

**BACKGROUND:** Hypoglycemia is a recognized complication of sulfonylurea overdose [1]. Conditions such as acute or chronic renal insufficiency can potentiate these effects and may result in life threatening hypoglycemia even with therapeutic doses of sulfonylureas [1].

**CASE REPORT:** A seventy six year old man presented to the emergency department with extreme weakness, shortness of breath and lightheadedness. His medical history was significant for congestive heart failure, type 2 diabetes mellitus, hypertension and a remote history of cervical spine injury resulting from an aortic aneurysm repair. Blood sugar at home was reported to be 22 and prompted this ED visit. The patient had last taken his dose of glimeperide earlier that morning. At the time of presentation, the patient was afebrile, normotensive but tachypneic, diaphoretic and tachycardic. Examination revealed bi-basilar rales and moderate dependent edema. Initial glucometer reading was 36 and blood glucose on BMP was 49. Patient serum creatinine was 2.50, increased from a baseline of 1.80. While in the ED, patient was started on D10 IV fluids and received a total of 5 pushes of 25 grams of dextrose IV over 6 hours. The patient was also found to have acute decompensated congestive heart failure and was started on IV diuretics. Despite the above treatment, the patient remained persistently hypoglycemic. In light of the persistent hypoglycemia, 50 micrograms of octreotide were administered subcutaneously. In approximately two hours, the patient's blood sugar started to improve and the D10 IV fluids were discontinued. Eight hours later the patient received another dose of 50 micrograms of octreotide. By now, his sugars were consistently in the normal range. The patient underwent optimization of his cardiac function with diuretics during the remainder of his hospital stay. At the time of discharge, he was asked to discontinue glimeperide and was started on repaglinide.

**CONCLUSION:** Severe refractory hypoglycemia is a serious complication of sulfonylurea use in diabetics with renal insufficiency [1]. Prompt recognition and initiation of octreotide therapy appears to be effective in reversing the hypoglycemia rapidly and safely [2]. Emerging data suggests that octreotide should be used as first line therapy in treatment of cases such as above [3].

CASE: The patient is a 40 year old Caucasian woman with a history of chronic anemia secondary to menorrhagia, smoking and premature atrial contractions. Medication included oral contraceptives. She suddenly developed dysarthria and weakness of the left lower extremity (LLE) while dancing. In the last six months, she reports having had night sweats, intermittent low-grade fevers to 100°F and body aches and chills for which she self-treated with ibuprofen 400 mg four nights per week. Other history was notable for a right visual field deficit about two months ago lasting 30 minutes. In the emergency department (ED) vital signs and physical examination were within normal limits except for an erythematous rash of the thenar eminence bilaterally, mild dysarthria and LLE weakness. Laboratory data were within normal limits except for a hemoglobin of 10 g/dL. CT of the head was negative for hemorrhage. While in the ED, the dysarthria and her ability to ambulate independently improved but was not restored to baseline. Transthoracic echocardiogram revealed a 2.7x3.5 cm echogenic mass within the left atrium associated with the inter-atrial septum. Transesophageal echocardiogram demonstrated a large 3.7x3.0x3.0 cm lobulated heterogeneous mass on the medial aspect of the left atrium. The morphology suggested a myxoma and/or vegetation. MRI of the brain showed approximately 40 bihemispheric acute lacunar infarcts. She underwent surgery to remove the LAM. Post-operatively, she complained of left calf and right foot pain. Arterial duplex scan revealed emboli in the right profunda artery and the left popliteal and peroneal arteries. Embolectomy of the right femoral and left popliteal arteries was performed, where myxomatous tissue was removed. The patient was discharged home with physical therapy.

DISCUSSION: Cardiac myxoma is a source of emboli to the central nervous system and elsewhere in the vascular tree. Non-specific constitutional symptoms and transient neurological ones may be over-looked in the absence of a cardiac history. In this scenario, cardiac investigations may not be performed and the diagnosis of this rare condition may be delayed until the onset of more significant embolic disease, such as stroke with functional impairment.

INTRODUCTION: Exercise capacity is a major determinate of cardiovascular risk and can be expressed as estimated metabolic equivalents of task (METs). An exaggerated blood pressure response to exercise is also known to be associated with various cardiovascular risk factors and adverse cardiovascular outcome, even in normotensive individuals. The purpose of this study was to determine whether the blood pressure changes during exercise independently predict the exercise capacity and diastolic function in patients with and without hypertension.

METHODS: All patients had transthoracic echocardiography and treadmill exercise test between April and September, 2014. The patients with negative stress test and normal EF on echocardiogram in one month were reviewed. The other exclusion criteria included known coronary artery disease, inability to tolerate exercise stress test, severe structural heart disease, and chronic renal disease. The blood pressure response was presented as the change of systolic blood pressure during the exercise divided by the METs.

RESULTS: Among 61 normotensive patients, 16 exhibited an exaggerated blood pressure response (&gt;880;210mmHg for men and &gt;880;190mmHg for female, or &gt;916;SBP/METs &gt;880;7.5mmHg, or &gt;916;DBP&gt;880;20mmHg) while the same was seen in 13 out of 61 patients with hypertension. There was no significant difference in age and sex between patients with or without
exaggerated blood pressure response. In hypertensive patients, echocardiographic doppler derived E/E′ was high in patients with exaggerated blood pressure response (E/E′ 8.31±2.67 vs 10.92± 7.06, P<0.045). But there was no significant difference in E/E′ among normotensive patients (E/E′ 6.79±2.12 vs 7.15± 1.70, P>0.05). There was no difference in METs between patients with and without exaggerated blood pressure response in hypertensive patients (METs 9.18±2.83 vs 8.56 ±2.70 P=0.05) or in normotensive patients (METs 10.56±2.49 vs 10.51 ±2.87 P>0.05). There was no difference in METs between patients with and without exaggerated blood pressure response in hypertensive patients (METs 9.18±2.83 vs 8.56 ±2.70 P=0.05) or in normotensive patients (METs 10.56±2.49 vs 10.51 ±2.87 P>0.05). However, the METs did correlate with the baseline SBP (r=0.292), DBP/METs (r=0.229), left atrial volume (LAV) (r=-0.231), LA volume index (LAVI) (r=-0.224), left ventricular septal wall thickness (LVSd) (r=-0.196), diastolic function (E/A r=0.324, E′ r=0.276, E/E′ r=0.261), age(r=0.477) and sex (r=0.385)(all p<0.05). The baseline DBP, change of DBP, left atrial diameter, left ventricular mass index (LVMI) did not correlate with excise capacity. By stepwise liner regression analysis, the baseline SBP and SBP change per METs, and LAV were shown to be independent determinant of the patients’ exercise capacity besides the traditional parameters of age and sex. R2 was 0.516.

CONCLUSIONS: Hypertensive patients with exaggerated blood pressure response to exercise stress test had worse diastolic dysfunction in comparison to patients without an exaggerated blood pressure response. Besides age and sex, baseline SBP, ΔSBP/METs, LAV independently predicted the exercise capacity in patients with and without hypertension.

202 158 Clinical Vignette Aung, Myo Myo Aung MM, MD, Ursani MA, MD, Iyer P, MD, Muddassir S, MD, Barn G, MD, Seinfeld F, MD, Share Saint Francis Medical Center (Sara Wallach) Recurrent CVA in low risk patient secondary to Large Atrial Myxoma INTRODUCTION: Primary cardiac tumors are extremely rare. Metastatic cardiac lesions are twenty times more common than primary lesions. Cardiac tumors can present with embolization, heart failure, heart blocks, arrhythmias, pericardial effusions, mitral regurgitation, secondary pulmonary hypertension and neurological symptoms.

CASE PRESENTATION: 46 year-old Hispanic male with no significant past medical history complaining of slurred speech, change in mentation, blurry vision, and right limbs weakness. Patient’s symptoms began one day prior to presentation. Change in speech and mentation progressively worsened. Patient also has a history of intermittent palpitations and dyspnea on exertion. The patient denied having any significant family history of hypercoagulable state, coronary disease or presence of atrial myxoma. On exam, the patient was found to have a 4/6 systolic murmur auscultated loudest at the 5th intercostal space midclavicular line. General neurological exam displayed expressive aphasia and dysarthria. Furthermore, right extremities strength +4/5 with appropriate tone. No change in sensation and hyperreflexia was noted. Cerebellar signs were absent. Patient underwent CT head displayed bifrontal hypodense brain lesion without significant regional mass effect. MRI Brain displayed acute left frontal infarction and well as old right frontal, left pontine and left cerebellar infarction. Patient was given high dose aspirin. 2-D Transesophageal Echocardiogram displayed a large mobile left atrial myxoma attached to the lower atrium and mitral annulus area. The myxoma transversed the mitral valve and encroached on the left ventricle, leading to severe mitral regurgitation. The patient underwent an emergent excision of the myxoma which was found to be 4.5 cm in size. The patient did not have any post-operative complications and recovered well.

DISCUSSION: Atrial Myxoma is a benign tumor and is commonly associated with recurrent cerebral accidents. The etiology is unknown, 20% of cases are asymptomatic. Others present with ischemic stroke, syncope, psychiatric manifestations, unspecified headaches and seizures. It is important to note that the tumor emboli are not related to size of the myxoma, rather, are related to the mobility and friability of the mass itself. Cerebral aneurysm is rare complication. Young individual with secondary cause of multiple cerebral infarcts, should rule out left atrial myxoma. We present this case to increase awareness to stress an appropriate...
### Clinical Vignette

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<td><strong>Of Anomalous Left Coronary Artery from Right Sinus Valsalva: An Anomaly with Fatal Outcomes.</strong></td>
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Coronary anomalies are not uncommon. Patients with coronary anomalies can have a wide array of presentations varying from chest pain to sudden cardiac death. In this case, a middle-aged woman presented with atypical chest pain and was admitted for unstable angina. Cardiac catheterization identified an anomalous origin of the left coronary artery from the right coronary sinus. CT angiogram identified a high-risk course. The patient’s chest pain was attributed to anomalous origin of the left coronary artery with subsequent compression due to inter-arterial course between the aorta and pulmonary trunk. She underwent successful surgical revascularization with left internal mammary artery to left anterior descending with resolution of symptoms.

### Clinical Vignette

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<td><strong>Disseminated Kaposi Sarcoma In Haart Era, a Rare Occurrence and Delayed Entry to HIV Care</strong></td>
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**INTRODUCTION:** Aids-related Kaposi Sarcoma can present in a wide spectrum ranging from incidental findings to aggressive neoplasms resulting in significant morbidity and mortality. KS commonly affects the lower extremities, face, oral mucosa, genitalia, gastrointestinal tract and lungs. The features associated with poor prognosis include involvement in the GI tract, non-nodal viscera or extensive oral disease, tumor-associated edema or ulceration, a CD4 count < 200, history of opportunistic infection or oral thrush, B symptoms and a Karnofsky performance status <70

**CASE REPORT:** 29 year-old Hispanic male presented to ER for sore throat, fever and rash over face, neck and upper chest for 2 days. He was empirically treated for pharyngitis. He returned after 9 days for persistent fever, rash & dyspnea on exertion. He was septic and had anemia, thrombocytopenia. Chest X-ray showed right costophrenic angle infiltrate. CT of chest and abdomen showed diffuse lymphadenopathy. HIV test was positive. He was treated for community acquired pneumonia and discharged. Upon follow up in Infectious Diseases clinic, he complained of dyspnea, diarrhea and vomiting. His CD4 counts were 33/uL and viral load was 187,096 copies/mL. He was re-admitted to ICU. Patient appeared dyspneic with worse violaceous rash over neck, face and upper chest. He had septic shock, persistent anemia and thrombocytopenia. Repeated imaging revealed new bilateral pleural effusions, diffuse lymphadenopathy, dependent band like opacities and new right upper lobe ground-glass opacity. Because of bilateral pleural effusion on CT chest, the diagnosis was less likely PJP; preliminary diagnosis of pulmonary Kaposi’s sarcoma was made. Diagnostics for Mycobacteria, Cytomegalovirus, Ova and parasites were all negative. Skin biopsy confirmed with Kaposi’s sarcoma and final diagnosis of disseminated Kaposi sarcoma ( stage T1I1S1 ) involving skin, lungs and GI tract was made. HAART was initiated along with empiric MAI coverage. He started on weekly Doxorubicin after his sepsis was resolved. He initially improved, but subsequently developed Pneumonia, Septicemia and continued to deteriorate. Patient expired during the hospital course.

**DISCUSSION:** Our patient presented with diffuse Kaposi Sarcoma involving skin, pulmonary and GI tract; which is unusual in the...
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Spontaneous Cerebrospinal Fluid Rhinorrhea:

**INTRODUCTION:** CSF rhinorrhea is a clinical condition that requires surgery to prevent possible complication such as meningitis and brain abscess. This condition could be traumatic and nontraumatic (spontaneous), with the latter being very rare. To be categorized as spontaneous or idiopathic, there can be no history of Dural puncture or other cause of CSF fistula. The usual presentation is a unilateral clear persistent nasal discharge with orthostatic headaches. The misdiagnosis of these leaks can have serious consequences.

**CASE REPORT:** We report a case of a 36 year-old male who presented after one episode of a witnessed generalized tonic-clonic seizure lasting 2 mins. He had no focal deficits on presentation and had no prior history of seizures. He had fevers, chills, neck stiffness and severe persistent orthostatic headache preceding the seizures. He also had a one year history of clear nasal discharge from his left nostril which had been treated with various OTC allergy medications with no resolution of symptoms. Initial clinical exam revealed a temperature of 101.2°F, positive neck stiffness but negative kerning’s or brudzinski sign. He had an unremarkable neurological examination. An atraumatic lumbar puncture revealed mildly elevated protein and normal glucose, red and white blood cell counts. A gram stain and culture showed Streptococcus mitis/oralis and he was treated for meningitis. His CSFopening pressures were low, measuring 20mm H2O. Initial Head CT demonstrated mildly diffuse ventricles with prominence of the temporal hons bilaterally. There were also findings suggestive of a small sphenoid bone defect involving the roof of the left sphenoid sinus air cell, potentially representing a point of communication between the sphenoid sinus and the left middle cranial fossa. On further imaging assessment with nuclear cisternography, it demonstrated CSF leaking into the left sphenoid sinus. The Nasal discharge was sent to the laboratory for analysis and it tested positive for beta-2-transferrin. The patient was transferred to a tertiary center for further management.

**DISCUSSION:** Nasal Discharge is a condition often trivialized and a common reason for visits to the primary care physician. CSF Rhinorrhea, though rare, is an important diagnostic consideration in patients who present with "unilateral clear nasal discharge". Increasing awareness of this condition can prevent some of the devastating consequences of Misdiagnosis.

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Renal Artery Infarction Associated with Prothrombin 20210G/A Gene Variant

**INTRODUCTION:** Prothrombin G20210A gene mutation is an inherited thrombophilia common in people of European descent. Having one abnormal gene (called heterozygous) results in an increased risk for thrombosis. The risk is even much higher with two abnormal prothrombin genes (called homozygous). This mutation has been associated with venous thrombotic events; however its involvement in arterial thrombosis remains controversial.

**CASE REPORT:** We present a 44 year-old male with sudden onset of severe sharp right flank pain radiating to his right lower quadrant and groin associated with nausea and non-bloody emesis. He denied fever, chills, dysuria or hematuria. Patient admitted using phencyclidine about 2 hours prior to onset of symptoms. CT abdomen/pelvis with intravenous contrast showed an acute thromboembolic occlusion of the lower arterial branch to the right kidney with discrete cortical infarct of the right inferior pole. He
was then evaluated for a systemic etiology of the thrombotic event. He was found to have low HDL levels but normal cholesterol, triglyceride and LDL levels. He had normal blood glucose, denied tobacco use and complete blood count showed a leukocytosis of 16.8 and LDH level of 510. His urine drug screen was positive for PCP. He had no symptoms of autoimmune disease, had a negative rheumatoid factor, antinuclear antibody and normal erythrocyte sedimentation rate. 2D-Echocardiogram and agitated saline study showed a normal study with no patent foramen ovale, intracardiac or intrapulmonary shunting. Screening tests for thrombophilia including tests for protein C and S deficiency, Factor V Leiden, lupus anticoagulant, antiphospholipid antibody, antithrombin III and fasting plasma homocysteine were all negative. He was found to be heterozygous for the G20210A mutation in the Prothrombin/Factor II gene. He was treated with 38 unfractionated heparin and Coumadin. His INR was maintained at 2-3. After one year of follow up, he has had no recurrent thromboembolism.

DISCUSSION: Screening for thrombophilias including Prothrombin 20210G mutations should be considered in select patients who present with arterial thrombosis. Although Prothrombin 20210G mutation is known to be associated with venous thrombosis, our case report demonstrates its relationship to arterial thrombosis.

207 311 Clinical Vignette Hussain, Asim Dr. Grace Tseng, Dr. Muhammad Awan, Dr. Oleg Chebotarev Saint Francis Medical Center (Sara Wallach) Adult Patent Ductus Arteriosis in Two Siblings

INTRODUCTION: The ductus arteriosus is a fetal vascular connection between the main pulmonary artery and the aorta which usually obliterates upon birth of the fetus. A patent ductus arteriosus (PDA) occurs when the DA fails to completely close within one week after birth. PDA is common in neonates, but is a rare finding in the adult population. In addition, having a genetic association adds an even more unique angle to this pathology.

CASE DESCRIPTION: A 44 year old female with no medical history was referred to the cardiology clinic with palpitations and exertional dyspnea. She stated that she has been dyspneic since child-hood with poor exercise tolerance. Her symptoms have worsened in the past 6 months. Her initial electrocardiogram shows normal sinus rhythm with right axis deviation and ischemic pattern in anterior leads. A TTE showed increased right ventricular volume and diastolic pressure. A TEE confirmed pulmonary hypertension with a communication between the right and left pulmonary artery and descending aorta consistent with a patent ductus arteriosus. Her elder sister, who has a history of hypertension, presented to the medical clinic complaining of chest pain. Upon physical exam, a loud 4/6 pan-systolic machinery like murmur was appreciated. A suspicion of PDA was raised. A few weeks later, the patient presented to the ER for acute chest pain and exertional dyspnea. Her cardiac enzymes and EKG ruled out acute coronary syndrome. Subsequent 2D echo and TEE were inconclusive. The following CT angiogram of the chest revealed a 8mm caliber artery connection between left main pulmonary artery to the proximal descending thoracic aorta, consistent with a PDA.

DISCUSSION: There has been an increase in the incidence of PDA secondary to the survival rate of preterm infants. Nonetheless the un-repaired patent ductus arteriosus in the adult is rare in developed countries. The case also highlights a complication of the disease if it persists until adulthood; namely that the patient may not be a candidate for repair. In addition, this abstract highlighting a PDA in two siblings may suggest a genetic link in the family which has not been identified yet. Previous studies have suggested that genetic abnormalities in chromosome six may predispose individuals to PDA among other clinical features.
Elevated Protein S Antigen and Ischemic Stroke

INTRODUCTION: The term "stroke" denotes a group of disorders in which there is usually abrupt onset and causes neurologic damage. The vast majority of disorders are ischemic in origin and are caused by insufficient blood flow to all or part of the brain. While ischemic strokes are largely secondary to thromboembolic causes, there are other etiologies such as vasculitis, coagulopathies, myoxma, hemoglobinopathies etc. This abstract highlights an unusual and rather rare form of coagulopathy induced CVA.

CASE DESCRIPTION: A 50 year old female with past medical history of depression, anxiety, and bipolar disorder and a pertinent social history of approximately 10 pack year history of tobacco use, presents to the emergency room with a complaint of being weak on the right side of her body. She was thus unable to walk. She also admitted to slurring of the speech. On exam, patient had mild dysarthria, and had decreased motor strength on the right upper extremity. In addition RUE displayed pronator drift. Sensation and reflexes were intact with absent cerebellar signs. A CT head/brain without contrast on admission was negative for any pathology. An MRI of the brain revealed an acute non-hemorrhagic infarct in the left basal ganglia and adjacent corona radiata. An ekg and telemetry monitoring revealed regular sinus rhythm. A transthoracic echo, carotid doppler, and transcranial doppler study were negative for any pathology predisposing to the stroke. Given the lack of largely significant risk factors and negative cardiovascular workup, a hypercoaguable panel was ordered. This resulted in an elevated protein S antigen.

DISCUSSION: Coagulopathies are an established risk factor and etiology for strokes. However in this case one would expect a protein S deficiency leading to the neurologic outcome. Rather it seems an elevation of protein S antigen is the culprit. In the literature there are very few references to this phenomenon. There is mention that an elevation of protein S antigen predisposes one to adverse cerebro-vascular events, including coronary heart disease and stroke. Further research is warranted to further explore this relationship especially given the heavy burden of disease from cerebro-and cardiovascular pathology.

A Case of Crigler- Najjar Syndrome Type II

INTRODUCTION: Crigler-Najjar syndrome (CNS) is a rare cause of unconjugated hyperbilirubinemia resulting from mutations of UGT1A1 gene encoding bilirubin-UDP-glucuronosyltransferase. CNS type II is characterised by reduced enzyme activity (i.e, 10-30% of the normal range), responses to phenobarbital, and serum bilirubin levels between 5.8 and 20.0 mg/ dL. Although CNS type II is considered a benign condition, the health effects of elevated unconjugated hyperbilirubinemia are not established. We present a potential case of CNS type II.

CASE REPORT: A 24 year-old Haitian woman presented to the clinic for pre-employment physical exam. She stated that her eyes have been consistently yellow since birth without any significant changes. No other family members had similar symptoms. She had never seen a physician; however, recalls that her mother was told to expose her to the sun because of her yellow eyes. She reported no pain, pruritus, paresthesia, discharge from eyes, prior trauma to the eyes, fevers, chills, diaphoresis, fatigue, weakness, loss of appetite, weight loss, clay-colored stools, dark urine, or history of blood transfusion. Past medical history included vaginitis and urinary tract infection which were treated with antibiotics. She denied past surgery and use of medications, alcohol, or illicit drugs. She had no family history of hematologic or hepatic disorder. She immigrated from Haiti three years ago. On examination, besides obvious jaundice in palms and sclera, there were no significant abnormalities including hepatosplenomegaly, ascites, or neurological
abnormalities. Complete blood count, coagulation, comprehensive metabolic panel and thyroid function tests were unremarkable except for elevated serum total and indirect bilirubin levels of 16.6 mg/dL and 16.4 mg/dL, respectively. Haptoglobin, LDH, peripheral blood smear, and reticulocyte counts were normal. Hepatitis panel, urine toxicology, and HIV antibody were negative. Urine bilirubin was not detected. Computed tomography of abdomen and pelvis revealed no significant abnormalities in liver, bile duct, pelvic or retroperitoneal lymph nodes, and spleen. Patient was planned for genetic testing and phenobarbital treatment.

DISCUSSION: Although CNS type II is considered a benign condition, adverse effects including neurotoxicity of elevated unconjugated hyperbilirubinemia are not established. Given that phenobarbital effectively reduces bilirubin levels, confirmation tests including bilirubin-UDP-glucuronosyltransferase enzyme activity as well as genetic counseling and testing should be initiated when CNS type II is suspected.

210. Clinical Vignette

Imayama, Ikuyo
M. Ejedike, MD; N. Metupalli, MBBS; R. Blanco, JR; H. Bhatti, MD; S. Nadeem, MD

Saint Francis Medical Center (Sara Wallach)

A Case of Ovarian Adenocarcinoma Associated with Hypercalcemia

INTRODUCTION: Hypercalcemia is an uncommon paraneoplastic syndrome in gynecologic neoplasms. It is caused by (1) production of humoral factors including parathyroid hormone-related peptide (PTHrP), 1,25-(OH) dihydroxyvitamin D and PTH and (2) bone metastases. Although there are several case reports of hypercalcemia in ovarian clear cell carcinoma, few studies have reported hypercalcemia in ovarian adenocarcinoma, non-small cell type neuroendocrine carcinoma admixed with endometrioid adenocarcinoma. We report a case of ovarian adenocarcinoma which presented with hypercalcemia.

CASE REPORT: A 62 year-old Caucasian woman with a past medical history of ovarian cancer after undergoing total abdominal hysterectomy with bilateral salpingo-oophorectomy and chemotherapy 7 years ago, depression, anxiety disorder, and mitral valve prolapse presented with the complaint of frequent falls. The patient stated that earlier in the morning she experienced bilateral leg weakness without dizziness or visual problems. She had been experiencing generalized fatigue, memory problem, irritation, constipation, and occasional confusion for one month. She denied weight loss, joint, leg or back problems, urinary or bowel incontinence, recent medication or supplement changes. Physical exam including neurological exam was unremarkable except for a Grade III/VI systolic ejection murmur at apex and diminished bowel sounds. Serum calcium was markedly increased to 14.8 mg/dL. No specific ECG changes were observed. Head CT without contrast did not show any acute pathology. Further testing revealed suppressed PTH and 25-hydroxyvitamin D, and normal levels of angiotensin converting enzyme, rheumatoid factor, c-reactive protein, serum protein electrophoresis, CA-125, and CEA. Abdominal/pelvic CT showed a large pelvic mass which revealed ovarian-type adenocarcinoma. Chest CT showed benign chronic granulomatous process (i.e., non-calcified granulomas of lungs and bilateral calcified hilar lymph nodes), and three lung nodules and a lesion in the liver suspicious for metastatic disease. Bone scan did not show any osseous metastatic disease. The hypercalcemia was treated with hydration, furosemide, calcitonin and pamidronate. Ovarian cancer was treated with carboplatin. Serum calcium decreased to 8.6 mg/dL and the symptoms resolved.

DISCUSSION: While small cell and clear cell carcinomas of the ovary are known to be associated with hypercalcemia, few cases were reported on PTHrP-producing ovarian adeno carcinoma resulting in hypercalcemia. Further investigation on molecular mechanisms of how ovarian adenocarcinoma produces PTHrP is warranted.
**Vignette**

Krathen, DO

**Medical Center (Sara Wallach)**

**INTRODUCTION:** Metastatic esophageal cancer has a poor prognosis and is more common in age group of 65-74 years. It’s rare in younger population below 35 years. We report a case of 28 year-old Caucasian male with history of Gastro-esophageal reflux disease, who was found to have extensive metastatic esophageal cancer. Liver is the most common site of distant metastasis. However, our patient had metastasis to the lung. Also, elevated tumor markers were seen in this patient.

**CASE REPORT:** A 28 year-old Caucasian male with past medical history of gastro-esophageal reflux disease for 10 years and history of eight pack year smoking, was sent to hospital for progressive dysphagia, mild shortness of breath and 40 pound weight loss since the past two months. Initial chest X-ray showed enumerable bilateral lung nodules. A subsequent CAT scan of chest/abdomen/pelvis with oral contrast revealed lower esophageal mass with mediastinal adenopathy and diffuse bilateral pulmonary nodules which were consistent with metastatic disease. Upper GI endoscopic exam was performed and multiple biopsies from the diseased portion were obtained. The biopsies showed moderately differentiated adenocarcinoma of proximal, mid, distal esophagus and cardia of stomach. Alpha fetoprotein and beta-HCG levels were ordered, to rule out testicular cancer. Alpha Fetoprotein levels were markedly elevated with mild elevations in Beta-HCG levels. Testicular Ultrasound done during hospital course, showed no testicular masses. Patient’s sputum was sent for Acid Fast Bacilli (AFB) stain and culture, which were negative. HIV ELISA test was negative too. Patient was treated with one cycle of chemotherapy with Docetaxel and Cisplatin. A recent CAT scan (chest/abdomen/pelvis) with contrast showed increased bilateral pulmonary nodules with new periaortic lymphadenopathy, consistent with worsening esophageal cancer. Patient’s prognosis is deemed to be poor.

**DISCUSSION:** There is a reported upward trend in the incidence of esophageal adenocarcinoma. This case report portrays a young individual with moderate risk factors who presented with end stage disease and extensive bilateral lung metastasis. Also, there was an atypical elevation in tumor markers observed in this patient.

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**Clinical Vignette**

Lopez Moncayo, Javier

W. Siddiqui, MBBS; M. Aung, MBBS; J.M. Smith, MD

Saint Francis Medical Center (Sara Wallach)

**Ogilvie Syndrome**

**INTRODUCTION:** Acute colonic pseudo-obstruction (ACPO), also known as Ogilvie’s syndrome, is a disorder characterized by gross colonic distension of the cecum and right hemicolon in the absence of mechanical – anatomic obstruction. Patients presenting with this syndrome often have underlying medical or surgical predisposing conditions. The precise mechanism by which colonic dilation occurs in these patients is unknown but impairment of the autonomic nervous system is proposed as a possible etiology. Management includes conservative treatment, pharmacologic therapy, or invasive procedures.

**CASE REPORT:** This is 89 year-old Caucasian male with a past medical history of hypertension, hyperlipidemia, COPD, and hypothyroidism. He also had severe triple vessel coronary artery disease and underwent CABG without complications one week prior to presentation. He had no previous history of ileus nor bowel obstruction. One week post-op the patient complained of diffuse abdominal pain and distention associated with constipation. WBC count was 11,000. Serum electrolytes were normal. Abdominal films showed distended loops of bowel. CT scan detected more than 10cm dilatation of the colon. Angiography ruled out mesenteric ischemia. Stool for occult blood was negative. The patient was made NPO and hydrated intravenously. Considering more up dated protocols, first was consider starting pharmacological approaches, but the size of the bowel distention was the strongest clinical factor for posterior perforation and possible bradycardia. Colonoscopic decompression was also deemed, but interventionist did not
agreed with the risk of insufflating more pressure inside the colonic wall making it prone for acute perforation. Subsequently, a diverting loop colostomy was performed. He was started on parenteral nutrition until he was able to tolerate enteral feedings. Interestingly, further history obtained from the patient revealed two posterior episodes of pseudo-obstruction.

**DISCUSSION:** Ogilvie’s syndrome is a clinical syndrome arising with marked abdominal distension without evidence of mechanical obstruction. Prompt treatment is important to avoid the complication of perforated cecum, including trial of conservative measures with nasogastric decompression, bowel rest, and correction of electrolytes and cessation of medications with the potential of exacerbation.

**INTRODUCTION:** Bicuspid aortic valve (BAV) is the most common congenital cardiac defect. Associated findings including dilation of the proximal ascending aorta secondary to abnormalities of the aortic media are independent of whether the valve is functionally normal, stenotic or incompetent. Although symptoms often manifest in adulthood, there is a wide spectrum of presentations ranging from severe disease detected in utero to asymptomatic disease in old age. Because it is a disease of valve and aorta, surgical decision making is complicated.

**CASE REPORT:** A 28 year-old Hispanic male presented to the ER with acute-onset chest pain. While at work as a roofer, the patient drank “a Red Bull energy drink”. Within minutes the patient developed substernal chest pain, headache, nausea and shortness of breath. There was no prior history of personal or family illness. In the ER he was found to be diaphoretic and pulseless in all 4 extremities. He was hypertensive and bradycardic with a 2/4, harsh, systolic murmur in the second intercostal space on the 60 right. Chest radiograph showed a widened mediastinum. Computerized tomography demonstrated an ascending aortic aneurysm with aortic root dissection, hemopericardium associated with a descending thoracic aortic aneurysm without dissection, and polycystic kidney disease. 2D-echocardiography confirmed severe concentric left ventricular hypertrophy, a bicuspid aortic valve, a severely dilated aortic root dissection, and hemorrhagic pericardial effusion. Emergent cardio thoracic surgery was performed for aortic root and aortic valve replacement with reimplantation of the coronary arteries and placement of a St. Jude mechanical valve. Pathology confirmed myxoid medial degeneration of the aortic root but otherwise unremarkable valve histology. There were no immediate post surgical complications, but days later the patient reported persistent headaches. Axial computerized tomography revealed multiple aneurysms in the left internal carotid artery, left posterior communicating, and right middle cerebral trifurcation. Anticoagulation was started. Complications included a benign hematoma at the surgical access site.

**DISCUSSION:** Consumption of energy drinks carry potential danger. We offer insight into a patient at high risk of cardiovascular collapse, while promoting valve-preserving surgeries for BAV associated with dilatation of the aorta. Recent studies show this approach has excellent results, including stabilization of the aortic root while improving valve durability.

**INTRODUCTION:** Elevated Serum Ceruloplasmin In Hepatolenticular Degeneration

**INTRODUCTION:** Progressive autosomal recessive hepatolenticular degeneration (aka, Wilson’s Disease), a copper metabolism genetic disorder, usually presents in older children/young adults with hepatic, neurologic, and/or psychiatric manifestations clinically. 80-95% of cases have a low serum ceruloplasmin level on laboratory data, a key diagnostic value. A high index of suspicion
is necessary to pursue potential cases in patients with high ceruloplasmin levels. Failure of prompt diagnosis and treatment can be fatal.

**CASE REPORT:** 47 year-old non-alcoholic African-American female with past medical history of hypertension, insulin dependent diabetes, chronic obstructive pulmonary disease and mitral valve regurgitation presented to our SFMC hospital for a scheduled cardiac catheterization to get cardiac clearance to undergo valve replacement. While gathering more information, the patient complained of right upper quadrant pain (moderate intensity, non-radiating, described as discomfort, without alleviating or aggravating factors) for more than two weeks. Accompanying symptoms included tiredness, tremors and ataxia. Physical examination revealed yellowish skin and mucosae discoloration, protopiosis, jugular vein distention (2/4), displaced cardiac maximal impulse point, mitral systolic murmur (2/4), decreased inspiratory effort, bibasilar rales, abdominal wall secondary circulation, present bowel sounds, mildly tender right upper quadrant abdomen and hepato-splenomegaly. Asterixis, bradykinesia and imbalanced abnormal gait were also noted. Psychiatric history query revealed liable mood and impulsivity, subsequently confirmed by psychiatry evaluation. Laboratory examinations recorded elevated transaminases, direct hyperbilirubinemia, hyperammonemia, normal lipid panel, seronegative complete viral hepatitis panel, negative anti-mitocondrial and anti-smooth muscle antibodies, high ceruloplasmin (66mg/dL) and high urine cooper levels (182mcg/24hrs). Tomographic images noted a cirrhotic appearing liver, portal hypertension with dilated intra- and extra-hepatic bile ducts described as hepatic steatosis. Further testing showed negative autoimmune etiology and HIV seronegativity. Posterior genetic testing was planned as outpatient with proper follow up.

**DISCUSSION:** Untreated Wilson’s disease becomes progressively worse, eventually fatal. Conversely, early detection and treatment yields most living relatively normal lives. Liver/neurologic damage occurring prior to treatment is often permanent. High serum ceruloplasmin levels, although rare, must not dissuade the clinician from a diagnosis work up which may save their patient.

215. Clinical Vignette

**INTRODUCTION:** Diagnosing a stroke (CVA) in a young adult female with hypertension should not be difficult - deciphering its etiology however can be challenging. Thromboembolism and intracranial small vessel disease are unlikely in young adults, raising probable causes of cardiac abnormalities, vascular lesions, hematologic abnormalities, infection, head/neck trauma, genetic conditions or substance misuse. Cocaine-induced ischemia etiology is multifactorial (e.g. vasospasm, thrombus formation, cerebral vasculitis, hypertensive surges) further compounding etiology determination.

**CASE REPORT:** 33 year-old female with past medical history of cocaine abuse, hypertension, hyperthyroidism, depression and previous CVA (one month prior) with residual left-sided weakness was brought to the ED after found lying on kitchen floor. Patient was AAOx2 with partial gaze palsy, complete hemianopia, partial lower facial palsy, RUE + RLE drift, left hemiparesis, one limb ataxia, no sensory deficit, moderate aphasia, mild dysarthria equating to an NIH Stroke Scale score of 18. Urine toxicology was positive for marijuana + cocaine, cardiac enzyme markers negative and normal echocardiogram. Two weeks ago, the patient was admitted for CVA one day after smoking “crack” cocaine and found to have 30-49% right carotid artery stenosis. Patient’s aunt had a CVA at 46 years old. Admitted for right middle cerebral artery (MCA) infarct CVA secondary to cocaine use, TPA was withheld since symptoms were outside the therapeutic window. Non-contrast CT(head) scan demonstrated a large abnormal hypoattenuation concerning for acute right MCA infarct without ICH, confirmed by non-contrast MRI (head). Non-contrast MRA (head) demonstrated a high-grade
right MCA M1 segment stenosis-occlusion while MRA (neck) found no gross evidence of cervical internal carotid artery stenosis, bilaterally patent distal common + external carotid arteries and patent vertebral and basilar arteries. Neurologic symptoms gradually improved allowing ICU-to-Ward patient transfer on day #3. On-going differential work-ups include cocaine induced, vasculitis, and fibromuscular dysplasia (type II).

**DISCUSSION:** While the etiology of an ischemic CVA in a “crack” cocaine smoking, young, hypertensive female adult may be as simple as an induced hypertensive surge, other independent or concomitant causes may exist. Simple cocaine abstention can prove fatal should another causes exist. The clinician must to rule out other causes.

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**216. 231 Clinical Vignette**

Meloro, Ralph Lopez Moncayo, J.; Amba, S.; Mettupalli, N.; Mukkavilli, G.; Wallach, S.

Saint Francis Medical Center (Sara Wallach)

**Persistent Uncontrolled Diabetes = Hereditary**

**INTRODUCTION:** Hereditary hemochromatosis, an autosomal recessive disorder classically from a HFE gene mutation, causes increased intestinal iron absorption resulting in progressive heme and non-heme iron deposition in body tissues, most commonly the heart, liver, pancreas and pituitary. In contrast to normal physiologic iron absorption, iron absorbed in individuals with hemochromatosis is not regulated by iron stores and there is no normal mechanism for excess iron excretion once absorbed. Diagnosis may elude clinicians who don’t have a high degree of clinical suspicion. 63

**CASE REPORT:** 52 year-old African American female with past medical history of diabetes mellitus, hypertension, thrombocytopenia, holosystolic cardiac murmur, chronic lower back/left hip/leg pain presents to our SFMC medicine clinic for routine follow up as she has for the past several years. Laboratory HbA1c of 32.4 and recent hospitalization for diabetic ketoacidosis underscored our challenge to help her gain glycemic control. Prior hospitalization earlier this year yielded a diagnosis of myotropic dermatitis of the face, trunk and bilateral upper extremities. Current physical examination revealed normal appearing skin, hepatomegaly, positive SLRT on left lower extremity with no alarming symptoms of cord compression. Alternate/concomitant diagnoses were pursued through extensive rheumatologic and metabolic work up studies. Serum iron profile studies ultimately pointed us in the right direction with elevated iron level of 213 mcg/mL (normal 37 -145), ferritin level of 487ng/mL (normal 37-150) and iron saturation of 59.8%. Elevated liver function tests (hyperbilirubinemia, elevated transaminases and ALP) and a CBC significant for elevated hemoglobin/hematocrit with a low platelet count helped support our working diagnosis of hemochromatosis.

**DISCUSSION:** Although the common manifestations of diabetes mellitus, impotence, arthropathy, skin pigmentation and cardiac enlargement occur in hemochromatosis, these often don’t abruptly present simultaneously. This creates the challenge to the clinician to make the diagnosis. Clinical suspicion of hereditary hemochromatosis may prove useful in evaluating patients with uncontrolled diabetes.

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**217. 223 Clinical Vignette**

Mettupalli, Neeha S. Amba, MD; S. Muddassir, MD

Saint Francis Medical Center (Sara Wallach)

**Bartter Syndrome**

**INTRODUCTION:** Bartter syndrome is an autosomal recessive disorder that often presents in childhood. It is caused by a defect in the genes that direct synthesis or membrane insertion of the transporters in sodium chloride reabsorption in the thick ascending limb of the loop of Henle. Its prevalence is 1 in 1,000,000 individuals. There are 5 types of Bartter syndrome. Variation in disease severity depends upon different types. It has been associated with developmental disabilities, hypokalemia, hypochloremia, metabolic...
alakalosis, polyuria and polydipsia.

**CASE REPORT:** A 32 years old Hispanic male was admitted to hospital for evaluation of a witnessed generalized tonic-clonic seizure at work. He had a similar episode 1 month prior to admission. He denied taking any medications and did not have seizures as a child. There was no significant family history and he denied alcohol or illicit drug use. Vitals signs and physical examination including neurologic examinations were unremarkable. Urine drug screen was negative. An MRI brain showed a 2.0x2.6 cm mass in the left posterior temporo-parietal lobe. He underwent resection of the mass and biopsy of the mass revealed possible Glioma. Since hospitalization he was normotensive or hypotensive. He had persistent refractory hypokalemia (Potassium ≈ 2.8 to 3.0 mMol/liter) and hypochloremia (Chloride ≈ 89 to 97 mMol/liter) with metabolic alkalosis. Work up for hypokalemia revealed normal BUN/Creatinine, magnesium and urinalysis, urine potassium 28.7 mMol/liter, urine creatinine 55.6 mg/dl and urine osmolality 448 mOsm/kg. Calculated transtubular potassium gradient was close to 7. Urine chloride was 130 mEq/liter which ruled out vomiting or gastrointestinal loss. Urine potassium to creatinine ratio was 51.6 mEq/gm (Diagnostic for Bartter syndrome which is >20 mEq/gm). 24 hour urine calcium excretion was 258 mg which ruled out Gitelman syndrome where urinary calcium excretion is less than 100 mg/day. The suspicion of Bartter syndrome was confirmed.

**DISCUSSION:** Bartter syndrome is not curable but the prognosis is able to lead fairly normal lives. Long term prognosis remains guarded because of the slow progression to chronic renal failure due to interstitial fibrosis. The initial therapy is often combination of NSAIDs and potassium-sparing diuretic, supplementation with potassium chloride and magnesium.

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**A Case of Thrombotic Thrombocytopenic Purpura**

**INTRODUCTION:** Thrombotic Thrombocytopenic Purpura (TTP) is a rare blood disorder characterized by clotting in small blood vessels of the body (thrombosis), resulting in a low platelet count. In its full-blown form, the disease consists of the pentad of microangiopathic hemolytic anemia, thrombocytopenic purpura, neurologic abnormalities, fever, and renal disease. The ultimate cause of TTP is unknown; however, recent research has uncovered some clues about the pathophysiology.

**CASE REPORT:** 44 year-old female with extensive past medical history of TTP, ITP, hyperlipidemia, marijuana abuse, depression and lung nodule s/p thoracoscopy and excision presented with the chief complain of sudden onset of headache and blurry vision for 21 days associated with fever, pruritus, vomiting and rhinorrhea with no aggravating or alleviating factors for which she had visited the ER several times over the last few days. Patient stated that for the past 2-3 days she was having some rashes on her belly and chest and on the day of admission she developed some on the shins. She consulted her PCP who advised her to get have blood work done. On physical exam, petechial rashes were noted on the chest, abdomen and some purpura on the right shin. 82 The rest of the physical examination was unremarkable. Patients CBC showed platelet count of 26000 and hemoglobin of 10.9, both of which dropped to 21000 and 9.6 respectively in the subsequent six hours. Considering the history of TTP in the patient and 4 out of 5 criteria of the pentad being fulfilled, the diagnosis of TTP was made and the patient was immediately started on IV methylpredniso1ne and plasmapheresis. In the next 3 days patient's platelet count increased to 130000 and Hb also came up to 12.5. Patient was clinically improved and was subsequently discharged and outpatient plasmapheresis was arranged.

**DISCUSSION:** The mainstay of treatment for TTP is high dose steroids and plasmapheresis. The identification of ADAMTS13 deficiency as the cause of TTP has major implications for the treatment, as the recombinant ADAMTS13 enzyme could now
potentially be produced and might provide a “safer and more effective” alternative to plasma exchange.

Clinical Vignette

Tseng, Grace

Awan M, Iyer P, Dessalines N.

Saint Francis Medical Center (Sara Wallach)

Collapsing Type of Focal Sclerosis

INTRODUCTION: Collapsing FSGS is most often seen in association with HIV infection but is increasingly recognized in non-HIV infected patients. Collapsing glomerulopathy is a distinct clinicopathologic entity with significant proteinuria, poor response to immunosuppressive therapy, and rapid progression to renal failure. It is characterized by the collapsing feature of glomerular damage with frequent tubulointerstitial involvement and rapid progression to ESRD. We have observed one case of collapsing FSGS in a patient with untreated SLE.

CASE REPORT: A 30 year-old African American male with ten-year SLE present with joint pain for four months, acute weakness, SOB, fatigue, myalgia for 5 days. He was not on any therapy and he had refused Lupus treatment because of reproductive issues. He has had a facial discoid lupus since he was teenager. The laboratory analysis revealed severe anemia, leucopenia, uremia, anion gap metabolic acidosis, hyponatremia and suggested stage-V renal failure, however the chronicity of his renal disease was unclear. The renal ultrasound showed extremely echogenic and slightly enlarged kidney, compatible with advanced renal disease. CT of abdominal and chest showed possible lupus related peniculitis in abdominal wall. His random urine collection and twenty four hour urine collection showed nephrotic-range proteinuria. Urinalysis under microscopy presents RBC cells but no RBC casts founded. Rheumatology workup showed positive anti-ds-DNA antibody and positive ANA. HIV serum test was negative. Cryoglobulin work up was negative. His complement C4 and Cardiolipin antibodies IgG were slightly elevated. Renal biopsy was performed and the renal pathology showed collapsing type focal sclerosis segmental glomerulonephritis. The patient, despite treatment with high dose corticosteroids, progressed to end stage renal disease and required hemodialysis within the course of hospitalization.

DISCUSSION: This patient was diagnosed with Collapsing type of Focal Segmental Glomerulosclerosis likely caused by his chronic untreated lupus nephritis. This case demonstrated an unusual type of FSGS found in advanced stage of nephritis. SLE has not been reported to be related with this variant type of FSGS until recent years.

Clinical Vignette

Ursani, M. Ali

Sasinowska S, Hussain A, Karabulut N

Saint Francis Medical Center (Sara Wallach)

Job’s Syndrome: A Rare Cause of Immunodeficiency

INTRODUCTION: Autosomal Dominant Hyper IgE Syndrome—also known as Job’s Syndrome—is a rare immunodeficiency syndrome that is classically known to present with the triad of eczema, recurrent fungal lung and bacterial skin infections, and an elevated IgE level (>2000 IU/ml). Physical signs may also include retained primary dentition, deep-set eyes, prominent forehead, hyper-extensibility, and scoliosis. The etiology is due to a mutation of the STAT3 gene, which encodes for a signal proteins involved in angiogenesis, wound healing, and immunity.

CASE REPORT: The patient is a 23 year-old male with a past medical history of deafness, mental retardation and recurrent pneumonia with spontaneous pneumothorax who presented to the emergency department with complaint of cough and shortness of breath. On exam, the patient was found to have a prominent jaw, deep-set eyes, and hyper extensibility of the MCP/ DIP/wrist/talocural joint. On chest x-ray, the patient was also found to have a chronic large abscess of the left upper lobe (consistent
with previous imaging). Vancomycin was administered for coverage of community acquired methicillin-resistant Staph Aureus. The sputum cultures returned with a growth of Aspergillus fungi. Vancomycin was discontinued and voriconazole was administered. After the initiation of antifungal treatment, the abscess persisted, and the patient experienced minimal relief of cough, and shortness of breath. Ultimately, the patient underwent pleurodesis of the left upper lobe. Given recurrent nature of his fungal pneumonia, we were suspicious of the patient being in an immunocompromised state. Upon further questioning, the mother of the patient stated that he had to undergo several dental procedures in the past due to retention of his primary teeth into his early adulthood. Additionally, the patient was frequently in the hospital as a child due to recurrent skin infections and pneumonia. X-ray of his spine confirmed scoliosis and IgE levels were found to be significantly elevated (>15,000 IU/ml). Upon discharge, the patient was referred to the National Institute of Health for further work-up.

DISCUSSION: Job’s Syndrome is an under diagnosed syndrome in which patients present with recurrent infections and have specific physical characteristics. Thus far, the treatment is based on symptomology; immunomodulators have not been shown to be beneficial. The case is presented to increase awareness of the condition in the medical community.

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Saint Francis Medical Center (Sara Wallach)

"I Just Can't Catch My Breath!" Clinical Manifestations of Idiopathic Pulmonary Fibrosis

INTRODUCTION: Idiopathic Pulmonary Fibrosis represents a unique subset of interstitial lung disease which presents with progressive respiratory decline over months to years and characteristic radiographic findings of diminished lung volumes with ground glass infiltrates and honeycombing. Patients have poor prognosis with median survival of approximately 3 years. Current therapeutic options are currently limited to just transplant at the moment with active research ongoing into fibrotic agents. Corticosteroids and cytotoxic agents at best provide a 15% response rate.

CASE REPORT: A 58 year-old Hispanic male with a past medical history of COPD (25 pack years), IVDA (currently using methadone) and anxiety arrived to the emergency department with complaint of progressively worsening dyspnea. The patient has been on home oxygen therapy and has had multiple visits to another institution for acute dyspnea—the most recent discharge six days ago. During that stay, the patient was treated for community acquired pneumonia and sent home after four days of IV antibiotics. The patient became more dyspneic periodically over the next two days prior to current admission. On exam, patient was tachypneic, tachycardic and hypoxic with oxygen saturation of 85% on 6L oxygen via nasal cannula. He admitted to chronic cough with clear sputum, denied fevers or chills. Patient states that he was previously a car mechanic and may have been exposed to asbestosis in the past. On admission, chest x-ray was read as increased interstitial markings suggestive of chronic interstitial lung disease without signs of infiltrates, pleural effusion and pneumothorax. Day two post admission, patient had an episode of acute desaturation and was subsequently intubated. High resolution CT of the Chest was done which revealed extensive bilateral ground-glass opacity and honeycombing. Right-side lung biopsy displayed honeycomb fibrosis, chronic inflammation and intracellular foreign particles—the findings were deemed as nonspecific but could not rule out idiopathic pulmonary fibrosis. The patient is currently being managed in the intensive care unit with a poor prognosis.

DISCUSSION: Idiopathic pulmonary fibrosis is a subset of interstitial lung disease. The diagnosis is indicated with the presence of ground glass infiltrates and honeycombing on CT Chest. Aside from lung transplantation, there is no cure, although cytotoxic agents
Baclofen Induced Brain Death?

Baclofen, a widely used central acting muscle relaxant, can mimic brain death in overdose. While cases published are rare, here we describe a patient who overdosed on Baclofen, met brain death criteria, who subsequently was discharged fully functional.

Introduction: Brain death implies the permanent absence of cerebral and brainstem functions—usually described as a persistent vegetative state. In adult patients, trauma and subarachnoid hemorrhage are the most common etiologies of brain death. Before the clinical exam for brain death can be done, a few pre-requisites have to be cleared: core temperature > 36 C, SBP >100, no drug or poisoning which confound the clinical assessment, and exclusion of medical conditions which may confound the exam. All the following must be present to clinically diagnose brain death: absent motor response, pupillary light reflexes, dilated fixed pupils, absent gag reflex, sucking reflex, caloric responses. Furthermore, apnea as demonstrated by the apnea test must be done, as well.

CASE REPORT AND DISCUSSION: Patient is a 30 year Hispanic male with past medical history of paraplegia status post traumatic injury one year prior causing fracture of the T7 vertebrae arrived to the Emergency Department when a friend found him unconscious, nonreactive to verbal and painful stimuli. He was last witnessed the night before carrying out his normal activities of daily living. Status post field intubation, ED examination found him unresponsive to touch, pain and vibration. Reflexes were 2+ bilateral patellar, brachial, bracioradialis and Babinski was down-going. Cough, gag, and corneal reflexes were not able to be elicited but pupils 1mm were fixed, constricted and midpoint unreactive to light and accommodation. Furthermore, caloric testing was done and found to be absent reflex. Tone of upper and extremity limbs was normal. Otherwise, patient’s exam was within normal limits. Head CT without contrast displayed mild sinusitis, otherwise normal. Glasgow Coma Scale Score was E1V1M1. EKG was also found to be normal sinus rhythm. Home medications were reviewed: Lyrica 3 mg BID, Neurontin 800 mg TID, Omeprazole 20 mg daily, baclofen 10 mg daily and duloxetine 30 mg daily. The first brain death exam was performed the following day which fulfilled brain death criteria. The second exam was scheduled to be done 24 hours later. 12 hours after the first Head CT without contrast was done, imaging was repeated—results were within normal limits. The night of Admission Day 2, the patient self-extubated and was immediately able to speak, and function at his baseline. After further discussion with the patient, he stated to have attempted suicide with baclofen. Any patient on Baclofen meeting brain death criteria should be re-evaluated frequently to rule out reversible metabolic cause secondary to drug overdose.

Urinary Tract Infection Caused by Raoultella planticola with Associated Staghorn Calculus

Raoultella planticola is a gram-negative bacillus, formerly known as Klebsiella planticola and Klebsiella trevisanii. It is a member of the Enterobacteriaceae family; although it is primarily an environmental commensal, there has recently been a surge in pathogenic cases. A 75-year-old male with a history of diverticulosis, enlarged prostate, nephrolithiasis, and hypertension, presented with encephalopathy three weeks after resection of a thoracic intradural meningioma. Physical exam was significant for lower abdominal pain and urinary retention. On laboratory evaluation acute kidney injury, uremia, and hyperkalemia were observed. Abdominal ultrasound revealed a large, non-obstructive staghorn calculus in the left kidney. Urine culture was positive for ampicillin-resistant R. planticola. Historically R. planticola has not presented a pathogenic threat to humans. The incidence of K. planticola was reported to be less than 0.5%. Since the first reported pathogenic case in 1984 R. planticola has only been implicated in sparsely reported cases.
of conjunctivitis, prostatitis, cystitis, cholecystitis, necrotizing fasciitis, cholangitis, soft tissue infection, and pancreatitis. Among these cases only that of prostatitis and cholangitis occurred in immunocompromised patients. Some authors, including Podschen et al postulate that K. planticola is becoming more prevalent as a human pathogen than previously speculated. The incidence of both pathogenic and drug resistant cases of R. planticola infection is increasingly being reported in the literature. The most recent epidemiologic studies have described pathogenic cases of drug resistant R. planticola. The first case of R. planticola carrying the blaIMP-8 gene that codes for imipenem resistance was published in early 2014. Additionally, pathogenic strains isolated in New Jersey and Ohio carried blaKPC gene, conferring resistance to beta-lactams. In our case, susceptibility testing indicated ampicillin resistance. This reinforced recent speculation that indicates the emergence of pathogenicity and drug resistance. To the best of the authors' knowledge, this is the second case documenting urinary tract infection (UTI) and the first associated with a staghorn calculus. Staghorn calculi are typically associated with urease producing bacteria such as Proteus and Klebsiella; which our patient has no prior history of. Among the exceedingly rare cases of pathogenic infection caused by R. planticola we present an ampicillin resistant case associated with a staghorn calculus. Our case highlights the emergence of R. planticola as a drug resistant pathogen that must now be considered in the differential for a variety of infections, particularly UTI.

Legalization of Marijuana and Cyclical Vomiting Syndrome

INTRODUCTION: Marijuana-derived compounds are frequently utilized as anti-emetics. However, it is well described that cannabinoids may also provoke emesis. Cannabinoids contribute to the pathogenesis of cyclic vomiting syndrome (CVS) in up to 50% of adults with this disorder. While the underlying pathophysiology is unclear, mechanisms involving the CB1 receptor and disordered allostasis have been proposed.

CASE REPORT: A 28 year old African American male presented with intractable nausea accompanied by non-bloody, non-bilious vomiting and dull epigastric pain of one week’s duration. He had a history of reflux and a hiatal hernia with a normal recent upper endoscopy. No travel, sick contacts or consumption of new or unusual foods was reported. He did have seven similar episodes within a three-year time span. Previous treatment with Zofran and Pepcid did not alleviate his symptoms or prevent recurrent episodes. Our patient associated onset of symptoms with physical exertion (ie: playing basketball) and noted mild relief by washing his face with cold water. Generally, the symptoms resolved within a few days and the patient experienced a multiple month period during which he was asymptomatic. On assessing the patient’s social history, he admitted to marijuana use at least twice daily, over a 12 year period. His blood work, laboratory evaluation and abdominal ultrasound were all unremarkable. A urine drug screen confirmed cannabinoid abuse. He was treated conservatively, with intravenous 0.9% NaCl, Zofran, Nexium and gradual diet progression; with improvement of symptoms prior to discharge.

CONCLUSION: Cyclic vomiting syndrome is a recently described condition that was &64257;rst documented in the literature in 2004. It is an idiopathic illness which is characterized by recurrent, self-limiting episodes of severe nausea and vomiting, separated by asymptomatic periods of varying duration, in the absence of an organic etiology, in chronic cannabis users. The diagnosis of CVS depends on recognizing the typical symptom constellations. Lack of familiarity with this disorder within the medical community leads to frequent misdiagnosis, often as psychogenic vomiting. Additionally, the absence of laboratory, radiographic and endoscopic markers makes CVS a difficult entity to diagnose. As the prohibition of marijuana use is progressively withdrawn, it is likely that an increase in its usage will result. This will ultimately increased number of cases of CVS seen by physicians. Clinicians should therefore
be aware of this condition and maintain a high level of suspicion in patients with episodic hyperemesis and a history of cannabis abuse.

**Asthma and the Procoagulant State**

**INTRODUCTION:** Asthma is a common chronic disease characterized by airway inflammation, intermittent airflow obstruction, and bronchial hyper-responsiveness. Recent evidence has shown that asthma is associated with a procoagulant state in the bronchoalveolar space. There is further suggestion that patients with severe asthma have a high risk of pulmonary embolism, which may be further increased by oral corticosteroids. We report a case of venous thrombosis with pulmonary embolism in a 39-year-old male with severe persistent asthma.

**CASE REPORT:** A 39-year-old man with a long standing history of asthma and non-compliance presented with a 3 day history of cough, difficulty breathing and audible wheezing of insidious onset. Concomitantly, the patient reported right leg pain and right foot numbness as well as tingling that started one day prior to presentation. He denied any increase in sputum production, hemoptysis, fevers, chills or associated chest pain. He also denied any history of trauma. The patient has a 10 pack-year history of smoking, however denied alcohol or illicit drug use. His asthma is poorly controlled on albuterol; which he takes on an as needed basis. Family history is non-contributory. Physical examination revealed a tachycardia and tachypnea with relative hypoxia of 95% on room air. Diffuse inspiratory and expiratory wheezing and crackles could be auscultated in all lung fields. Homan’s sign was positive in the right lower extremity but no swelling or erythma could be noted. Initial bloodwork including complete blood count, and basic metabolic panel were normal. Arterial blood gas was 7.4/40/78/24.8. Chest X-ray was clear showing no infiltrates or consolidation. CT angiogram showed multiple bilateral filling defects compatible with pulmonary emboli. Venous Doppler revealed a thrombus in the right posterior tibial vein. Hypercoagulation workup including Protein C & S, Homocysteine, Antithrombin III, Anti-Cardiolipin and Factor V Leiden are negative. The patient was anticoagulated with Lovenox and bridged to coumadin. He was also placed on intravenous steroids and nebulized Albuterol with subsequent improvement in peak flows. He was discharged home on coumadin after achieving a therapeutic INR.

**DISCUSSION:** Patients with asthma are predisposed to acquired hypercoagulable states. A high clinical suspicion must be maintained for pulmonary emboli in asthmatics presenting with frequent exacerbations. Additionally it should be noted that asthma exacerbation may mask the clinical picture and subsequent diagnosis of pulmonary embolism. Further evidence is needed to definitively determine the association between the severity of asthma and the predisposition to thromboembolic disease.

**GLOBAL AMNESIA: RARE CLINICAL PRESENTATION OF POSTERIOR CIRCULATION STROKE**

**INTRODUCTION:** Beauty of cerebrovascular disease in neurological sciences is ability to localize anatomical lesion based on clinical characteristics. Ability to deduct the anatomical correlation of posterior infarcts is challenging because majority of the time the clinical presentation in these cases is a symptom salad with a syndrome like picture rather than clear cut clinical deficits. Posterior circulation strokes contribute one fifth of all ischemic stroke cases. One of the rarest forms of presentation is by amnesia. Currently there is no consensus on the etiology of the stroke associated with amnesia. We report a case of posterior circulation stroke presenting with an amnesic presentation.

**CASE REPORT:** An 88 year old Hispanic female was brought to the hospital due to confusion of unknown duration. She was last seen normal 2 days ago. Past medical history is significant for hypertension, hyperlipidemia, chronic systolic heart failure, bio prosthetic valve with bio prosthetic aortic valve replacement and permanent pacemaker. Medical and surgical history was unremarkable. Physical exam revealed a tachycardia with an INR of 4.9. A computed tomography angiogram of the chest and abdomen was normal. A non-contrast computed tomography of the head was performed which showed a large area of hypodensity with surrounding vasogenic edema in the left occipital lobe extending posteriorly to the left parietal lobe. A computed tomography angiogram of the head was normal. A diffusion weighted magnetic resonance imaging of the head demonstrated a large area of restricted diffusion within the left occipital lobe extending posteriorly to the left parietal lobe. Conventional magnetic resonance imaging revealed a large hyperintense area of acute/subacute infarction in the left occipital lobe extending posteriorly to the left parietal lobe with surrounding vasogenic edema. The posterior cerebral artery was occluded. The patient underwent a right internal carotid angiogram which showed complete occlusion of the left posterior cerebral artery. This was managed medically with anticoagulation and best supportive care. The patient was discharged in a stable condition to a skilled nursing facility.
mitral valve replacement, atrial fibrillation currently on anticoagulation. History could not be obtained from patient however family reports no recent history of fever, fall or similar experiences in past. Patient was living independently prior coming to the hospital. On physical examination, patient was found to have profound global amnesia with predominant retrograde memory loss. Neuropsychological testing revealed normal executive functioning, intelligence, attention, language, and mood. Her short term working memory was also intact. Her confusion was attributed solely because of her memory loss to the extent where she was unable to identify her family. She had no other complaints. No gross motor, sensory, cerebellar deficits were noted. Initial lab tests revealed patient was sub therapeutic despite being on Coumadin for anticoagulation. Computed tomography of brain without contrast was unremarkable. Magnetic resonance imaging of brain revealed multiple tiny infarcts in bilateral occipital lobes, right cerebellum and posterior pons. The multiplicity of infarcts, regionalization of vascular supply, prominent cardiac risk factors, state of sub therapeutic anti-coagulation point towards etiology of thromboembolism to distribution of posterior circulation. Patient was treated for Ischemic stroke and given higher doses of Coumadin to achieve therapeutic range of INR. At 6 weeks of follow-up patient completely regained her memory and currently has no neurological deficits. One very important differential to consider in this case would be transient global amnesia (TGA), condition whose etiology is still questioned.

CONCLUSIONS: The clinical presentation of vertebra-basilar cerebrovascular ischemic events is radically different from our usual encounters. Since the presentations are very different from each other and have a whole spectrum of symptoms to present with, we should be familiar with these cases for effective diagnosis and management. One step further from stroke would be basilar artery occlusion itself with the similar etiology and pathophysiology and has very high rate of fatality and mortality. This case once again underlines the unique nature of posterior circulation strokes thus helping us in future practice.

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Clinical Vignette

Patel, Dimple

Yamini, Sundermurthy M.D., Manda, Jayaprakash M.D.

Saint Joseph’s Medical Center (M. Anees Khan)

A RARE NEW ONSET JUNCTIONAL BRADYCARDIA WITH RETROGRADE CONDUCTION AND ATAXIA RESULTING FROM ORAL PHENYTOIN TOXICITY: THE ECLECTIC SIDE EFFECT PROFILE CLINICIANS SHOULD LOOK OUT FOR

Introduction

Phenytoin has a very narrow therapeutic index and a hepatic metabolism implies numerous drug-drug interactions. It’s side effects involve almost all the organ systems making it a diagnostic dilemma. We present a rare case of an elderly patient who had neurologic and cardiac symptomatology resulting from oral Phenytoin toxicity.

Case Description

85 y/o Caucasian female with hypertension, hypothyroidism, seizure disorder, left frontal meningioma post resection, previously independent, was brought from home with complaints of progressive bilateral lower extremity weakness, multiple falls and decline in mentation for 6 weeks. Prior to presentation, a friend noted her to be staring into space for several seconds. At baseline she remained aphasic after meningioma resection. Home medications included Phenytoin 100 mg twice daily, Levothyroxine 150 mcg and Irbesartan 150 mg. On physical examination reflexes were diminished to absent with a narrow based gait, however she was alert and oriented. Also blood pressure was elevated at 200/99 mm of Hg but she was bradycardic in 40s. Patient was admitted to telemetry with suspected complex partial seizures versus neuro-cardiogenic cause of her falls given the bradycardia. She was started on Hydralazine and Irbesartan which brought the BP down to normal range but asymptomatic bradycardia persisted in 40s. Neurology and Electrophysiology services were consulted. MRI of brain, CT of head and thyroid function tests were all within normal limits. EEG showed possible partial focal seizures. EKG showed junctional bradycardia with retrograde conduction at 42bpm. Troponins were negative and 2D Echo showed no wall motion abnormality. Free and total Phenytoin levels were elevated at 3.45.
and 27.2 respectively, so it was switched to Lacosamide. With free levels finally down to <0.3, patient’s ataxia and bradycardia resolved completely as verified by repeat EKG. All the consultants concurred with our diagnosis of dilantin toxicity and patient was discharged to subacute rehab in stable condition.

**DISCUSSION:** Multiple studies have shown the involvement of almost all organ systems in IV Phenytoin toxicity but only three case reports have been published with bradycardia as the cardiac side effects of oral Phenytoin. In our case, accurate diagnosis was challenging due to baseline aphasia and involvement of two organ systems in toxicity. Recognizing its harmful side effects involving multiple organ systems, prompt discontinuation and substitution with other anti-epileptic drug if needed is critical as rarely death has been known to occur and there is no antidote.


Bleeding is a common complication of cardiac surgery, accounting for a significant proportion of the total transfusions performed in the United States and Europe. The relationship between platelet reactivity, bleeding and other adverse events after coronary artery bypass graft surgery (CABGS) has been incompletely characterized. This study investigated the relationship between platelet reactivity and bleeding as a clinical outcome after successful CABGS. A total of 238 patients underwent CABGS were retrospectively followed for postoperative bleeding. Platelet reactivity unit (PRU) values for all patients were obtained preoperatively to assess the platelet reactivity. The data showed that a range of 180-200 PRU suggests the likelihood of bleeding after CABGS (P=0.004), with a statistically significant association only for dual antiplatelet therapy with aspirin and clopidogrel. In conclusion, using PRU values as a method to assess platelet reactivity and antiplatelet responsiveness, our findings suggest that it may be possible to stratify patients undergoing CABGS for the risk of postoperative bleeding particularly patients on dual antiplatelet therapy.

| 229 | Research | Patel, Hiren | Jeewanot Randhawa, Sushant Nanavati, L. Randy Marton, Walid J. Baddoura, Vincent A. Debari | Saint Joseph’s Medical Center (M. Anees Khan) | Laboratory and Clinical features of Toxin-positive and Toxin-negative Community-acquired Clostridium difficile Infection

Studies have described the clinical course of patients with Clostridium difficile infection (CDI) with positive enzyme immunoassay (EIA) for toxins A and B. Limited information is available for the patients with negative EIA but positive for the toxin B gene by the PCR. The aim of our study is to determine if there are any differences that exist among the clinical and laboratory parameters in the patients tested to be positive by EIA for toxin and those who were negative. This is a retrospective cohort study conducted in a 700-bed teaching hospital. We reviewed charts of the patients with presumptive CDI between January 2006 and July 2013. We divided these patients into two groups, Tox (+) and Tox (-) based on result of EIA for toxins A and B. The Tox (+) group had significantly higher white blood cells counts (p < 0.001), with significantly greater percentage of bands (p < 0.0001). Albumin and total protein both exhibit significantly (p < 0.0001, both comparisons) lower values in the Tox (+) group. Among clinical findings, the Tox (+) group had significantly longer length of hospital stay (p = 0.010). These data suggest that infection with a toxin (-) negative strain of C. difficile presents laboratory markers closer to those of healthy subjects and clinical features suggesting considerably less severity than infection with toxin (+) C. difficile.

| 230 | Clinical | Jumean, Khalid | Ahmad Abu Arqoub, | Saint | Mondor’s Disease of the Penis: A Forgotten Entity
**Vignette**

**MD. Katherine Hanify, DO.**

**Michael's Medical Center (John W. Sensakovic)**

**INTRODUCTION:** Superficial vein thrombosis was first described by Mondor in 1939 when it involved subcutaneous veins of the anterolateral thoracoabdominal wall. In 1955, Braun-Falcó described penile involvement, and in 1958, Helm and Hodge described an isolated superficial penile vein thrombosis. Mondor’s disease of the penis is an under-reported condition. This case report describes the symptomatology, diagnosis, and treatment of thrombosis of the superficial dorsal vein of the penis.

**PRESENTATION:** 41-year old male with no past medical history, he presented with swelling and pain of penis for 3 weeks. Our patient noticed the appearance of painful cord on the dorsum of his penis, near the penis root, being more painful during erections. There was no associated itching, discharge, hematuria, or dysuria. He denied any history of recent trauma, vigorous sexual activity, or use of constriction devices. He admits to have one sexual partner. He also denied any history of fever or lower urinary tract symptoms. Physical examination revealed a slightly anxious but physically healthy man with a dorsal cord-like swelling, extending from the pubic symphysis to mid-shaft of his penis, mild tenderness during palpation. Genitourinary examination was normal. There was no associated inguinal lymphadenopathy and the laboratory tests including blood and urine were within normal limits. The patient underwent ultrasonography examination which revealed a non-compressible portion of superficial dorsal vein as well as the lack of venous flow signals in Doppler ultrasonography. Provisional diagnosis of thrombosis of the superficial dorsal vein of the penis was made, and conservative treatment was prescribed in the form of nonsteroidal anti-inflammatory drugs (diclofenac) and 500 mg of ciprofloxacin twice daily for 5 days for prophylaxis. The patient was reassured of the benign nature of his condition and was instructed to abstain from sexual activity and was advised to follow up within three weeks. On follow-up visit his physical examination revealed a complete resolution of the swelling.

**DISCUSSION:** Mondor’s disease of the penis is a benign and, usually, self-limited disease. Patients complain of cord-like indurations on the dorsal aspect of the penis. The etiology of this condition is unknown. The diagnosis of the disease is mainly clinical, supplemented with Doppler ultrasonography. The differential diagnosis includes sclerosing lymphangitis, Peyronie’s disease, and a fractured penis. Treatment is essentially conservative. Anticoagulation with aspirin, heparin, or other antiplatelet agents will not expedite healing and is not necessary to prevent additional thrombosis. Antibiotics can be used prophylactically. NSAIDs can be used for pain relief. Patients should also be informed about the avoidance of sexual intercourse or masturbation. In most of the cases, symptoms resolve completely within 6 to 8 weeks. In cases with no resolution, despite conservative treatment, thrombus excision or vein excision has to be done.

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**Clinical Vignette**

**Sen, Parijat**

**Abhishek Kumar, Tejas Modi, Amol Shah, Hamid Shaaban, N Parikh**

**Saint Michael’s Medical Center (John W. Sensakovic)**

**HAIRY CELL LEUKEMIA: WHEN IT TURNED UP AT AN UNLIKELY PLACE**

**INTRODUCTION:** Hairy Cell Leukemia (HCL) is a chronic B cell lymphoproliferative disorder that obtains its name from the fine hair like projections seen on the cell surfaces under microscopy. Clinically, HCL is associated with splenomegaly and pancytopenia. Tartrate Resistant Acid Phosphatase (TRAP) positivity is diagnostic of HCL. While usual sites of spread are bone marrow, liver and spleen, rare involvements such as Skin metastases, sweet syndrome and panniculitis have been reported. We hereby report a rare case of duodenal involvement in HCL.

**CASE REPORT:** A 48-year-old Hispanic male presented to the ER with colitis in March 2008. Hematological workup revealed pancytopenia (hemoglobin-6.9g/dL, Leukocyte count- 4800/µL, platelet count- 23,000/µL). CT scan abdomen revealed a massive splenomegaly (17cm). Bone marrow aspiration and biopsy showed hypercellular marrow diffusely infiltrated by small lymphocytes with round to bean shaped nuclei with reticulated chromatin pattern and abundant cytoplasm. Immunohistochemical staining...
showed monoclonal kappa expressing B cell population positive for CD-19, CD-22, and CD-45 and TRAP while negative for CD-23, CD-10, CD-5 and CD-3. Reticulin stain showed diffuse increase in reticulin fibers in the marrow. A subsequent diagnosis of Hairy Cell Leukemia (HCL) was made and the patient received outpatient 2-Chlorodeoxyadenosine (CDA) 0.1 mg/kg for 5 days. He was readmitted three days after completion of chemotherapy with febrile neutropenia. Infectious etiology work up was negative. Broad-spectrum antibiotics and antifungals were started empirically. CT scan abdomen revealed thickening of the duodenum. Subsequently an Esophagogastroduodenoscopy (EGD) with biopsy was performed. The duodenal biopsy was positive for chronic inflammatory infiltrate, primarily consisting of atypical lymphocytes and plasma cells with TRAP positivity and hence a diagnosis of duodenal involvement with HCL was made. Repeat bone marrow biopsy done 2 weeks after finishing chemotherapy revealed remnant disease. At 3 months follow up patient was asymptomatic with a normocellular marrow and no residual disease. Labs showed leukocyte count-2800/uL, hemoglobin- 10gm%, platelet count- 246,000/uL. Repeat CT Abdomen in March, 2009 showed resolution of duodenal thickening and spleen size of 12 cms. Currently patient is in clinical remission for past 6years.

DISCUSSION: Hairy cell leukemia is rare B-cell lymphoproliferative disorder with annual incidence of 3 cases per million populations. HCL arises from late activated memory B-cells. Most common sites for organ involvement are liver, spleen and bone marrow. However, unlike most mature B-Cell Lymphomas, HCL shows minimal lymph node infiltration. The nature of its spread is attributed to its unique homing properties which is perceived to be limited to the blood compartments. Involvements of other sites in HCL is extremely rare and would change the way we perceive the pathology of the disease and the properties of hairy cells. We here report the first case to our knowledge of HCL infiltrating the gastrointestinal tract.

INTRODUCTION: Acute pancreatitis is one of the most commonly seen conditions accounting for over 250,000 hospitalizations every year. The diagnosis of acute pancreatitis is based on the presence of two of the following three criteria: clinical, serological and radiological. Gallstones and alcohol consumption are the most common etiologies causing up to 80-90% of the cases seen in practice. However, other etiologies like hypertriglyceridemia, smoking, invasive procedures like ERCP and a variety of medications have been well documented to contribute to disease burden. We, hereby, report a case of drug induced pancreatitis with normal serum lipase.

CASE DESCRIPTION: 34 year old lady came to the ER with complaints of acute epigastric pain associated with nausea and vomiting which was worsening for last 4 days. She was noted to have past medical history significant for hypertension, diabetes mellitus, HIV infection on HAART and chronic kidney disease. She underwent work up in ER, which revealed high WBC count with elevated creatinine, blood glucose and normal serum amylase and lipase. Due to high suspicion of acute pancreatitis and to evaluate for other causes of acute epigastric pain she underwent CT scan of the abdomen which showed peripancreatic fat stranding and induration with fluid collections around paracolic gutters and right upper abdomen consistent with acute pancreatitis. Further work up for etiology of pancreatitis showed no gallbladder stones and common biliary duct was 3 mm as seen on ultrasound. On detailed history patient reported no alcohol use but was found to be on HAART medications that are a well-known cause of pancreatitis. Medications were subsequently stopped and aggressive intravenous fluid resuscitation was initiated on admission. She continued to improve during the course of admission and was discharged as symptoms improved.
DISCUSSION: A wide variety of medications have been implicated in causing pancreatitis and the list is expanding at exponential rate. However, very few cases have been reported for drug induced pancreatitis with normal lipase raising a question if the diagnosis is being overlooked. We, with the help of this case report, would like to increase awareness and consciousness about this rare presentation of the disease.


233 99 Clinical Vignette Akula, Raj Saint Peter's University Hospital (Nayan Kothari)

POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME AND ISCHEMIC STROKE

INTRODUCTION: Posterior reversible encephalopathy syndrome(PRES) is a well described clinico-neuro-radiological condition described first time in 1996. It is a syndrome consisting of altered mentation, seizures, cortical blindness. It is diagnosed by CT/MRI findings in an appropriate clinical setting. The few main etiologies identified are eclampsia, immunosuppressive drugs, sepsis, Hypertension and recently there have been numerous case reports about its association with SLE, Porphyria and other autoimmune diseases. PRES involves the white matter, predominantly the water-shed areas of the posterior circulation territory, mainly the parieto-occipital regions though lesions have been described in other regions of the brain. The Pathophysiology is not clear but the current hypothesis suggests a vasogenic edema, vasospasm and endothelial dysfunction as the mechanisms causing this syndrome. Though stroke and respiratory failure have not been described consistently with the syndrome can they be the effect of the same pathophysiological mechanism?

CASE: A 36 y/o F in 38 wks gestation was bought with history of seizures and loss of consciousness. She had blood pressure in 200’s, low platelet count, elevated WBC and proteinuria. She became hypoxic and unconscious, subsequently went into respiratory failure. She was intubated and ventilated. An Emergency C-section was done and two healthy babies were delivered. An MRI and MRA of the brain showed hyperintense lesions in bilateral parieto-occipital and right cerebellar regions consistent with diagnosis of PRES. Also found was a restricted diffusion in the right posterior occipital region consistent with infarction. A repeat MRI Brain was done 2 days later which showed the findings were resolving but complete evolution of the infarction with no hemorrhage. After extubation and return of the patient to her baseline mental status, a thorough exam did not reveal residual neurological deficits.

DISCUSSION: This case exemplifies the etiology, clinical and radiological spectrum of PRES. The main pathophysiological mechanisms described include vasogenic edema causing leakage of proteins and fluid, Vasospasm and endothelial dysfunction. These mechanisms can also explain the respiratory failure and infarction found in this patient. This raises the question if PRES is a condition only associated with changes in the brain or is it a systemic process causing endothelial dysfunction and vasospasm across all organ systems which can be fatal.

CONCLUSION: Posterior reversible encephalopathy syndrome is a new entity and has not been well described. A deeper understanding into the pathophysiology of the disease is needed to better understand the nature of the disease and the spectrum of presentation. The multitudes of clinical, laboratory and radiological findings suggest that PRES might be a systemic process whose presentation can be varied and high index of suspicion must be maintained when diagnosing it.
| 234 | 127 | Clinical Vignette | Alcid, Arthur | Saint Peter's University Hospital (Nayan Kothari) | A case of albuterol-induced lactic acidosis  
**INTRODUCTION:** Although many medications are known to cause lactic acidosis, albuterol is not a commonly recognized etiology.  
**CASE:** A 61-year-old female with history of hypertension and a thyroid nodule presented to the hospital complaining of worsening productive cough and shortness of breath for four days. Her symptoms were associated with subjective fever, chills and chest tightness. In the emergency department she was afebrile, hypertensive and in respiratory distress. Oxygen saturation was 93 percent on room air. She was treated with methylprednisolone 125 mg IV, four albuterol-ipratropium nebulizer treatments (3 mL each), magnesium sulfate 2 gm IV and placed on bilevel positive airway pressure. Serum anion gap was 15, serum bicarbonate was 24 mmol/L. Lactic acid was 3.6 mEq/L. She was admitted to the intensive care unit for severe acute bronchospasm and respiratory distress and was treated with albuterol nebulizer therapy around the clock every four hours (2.5 mg each). The following day the patient’s repeat lactic acid level was 7.8 mEq/L despite her improved clinical status which did not correlate with this finding. Subsequent levels over the remainder of her admission were 2.4 mEq/L and 4.0 mEq/L, however the patient continued to clinically improve until her discharge.  
**DISCUSSION:** Lactic acidosis is caused by impaired tissue oxygenation, toxin-induced impairment of cellular metabolism resulting in the generation of lactic acid in the absence of hypoxia or hypoperfusion and the formation of D-lactate by bacteria in the gut. This patient’s initial presentation suggests impaired tissue oxygenation for which she was treated with aggressive albuterol nebulizer therapy. Despite clinical improvement, her lactate level increased. She was not in shock or sepsis. There was no history of liver disease, use of metformin or antiretroviral therapy. She had no history of bowel resection. CT scans of the thorax, abdomen and pelvis were negative for malignancy. Based on these findings, albuterol was suspected to be the most likely cause of the patient’s lactic acidosis. Albuterol causes lactic acidosis by enhancing glycogenolysis and gluconeogenesis, resulting in increased pyruvate production. Simultaneously, albuterol enhances lipolysis, causing inhibition of pyruvate dehydrogenase. This prevents pyruvate from entering the Krebs cycle, resulting in pyruvate reduction to lactate.  
**CONCLUSION:** Use of beta-2 agonists may cause lactic acidosis through metabolic effects. Recognizing this as a cause of lactic acidosis in the absence of other etiologies of can prevent unnecessary examinations and treatment.  

| 235 | 128 | Clinical Vignette | Ayangade, Tolulope | Saint Peter’s University Hospital (Nayan Kothari) | Not So Tonic Water after all – A Case of Quinine-induced Neurotoxicity  
**INTRODUCTION:** Quinine is an alkaloid derived from the bark of the cinchona tree. Its medicinal use in history includes as an antimalarial, treatment of muscle cramps, and in tonic water, a carbonated soft drink, for its bitter taste. Tonic water contains 83mg of quinine/L compared to 2100mg/day of quinine in antimalarial drugs. The use of quinine is limited today due to its cardiotoxicity, neurotoxicity, hematologic toxicity; occurring from hours to years after quinine ingestion.  
**CASE:** A 27 year old African lady with a history of chronic microcytic anemia, and a 1 year history of habitual consumption of 3 liters of tonic water daily presents with a 1 week history of gradual onset, episodic, mild to moderate intensity, non-radiating diffuse head aches. She was found to have a positive Babinski sign and dense bilateral Babinski. The patient’s cerebrospinal fluid analysis was unremarkable. The patient was treated with 100mg of benzodiazepine and 4 gm of magnesium. The patient’s neurological examination returned to normal on repeat testing.  
**DISCUSSION:** Quinine is a non-competitive inhibitor of the GABA-A receptor, which is a chloride channel. Quinine’s binding to the GABA-A receptor causes a chloride ion flux, which results in hyperpolarization of the neuron. This excessive intracellular chloride causes a decrease in the number of calcium ions. Calcium ions play a key role in the release of neurotransmitters and ultimately the transmission of nerve impulses. The patient’s sudden onset of diffuse head aches was most likely due to the action of quinine on the GABA-A receptors. The patient’s Babinski sign was likely due to the same mechanism. The patient was treated with a benzodiazepine and magnesium sulfate to alleviate the symptoms. This treatment was effective and the patient’s neurological examination returned to normal on repeat testing.  
**CONCLUSION:** The case presented above is a rare example of quinine-induced neurotoxicity and highlights the importance of recognizing this condition and treating it promptly.
heaviness aggravated by exertion and movements and relieved with rest. Her symptoms were associated with lightheadedness, nausea, hyperacusis, tinnitus, gait imbalance, and loss of postural tone without loss of consciousness. Symptoms were preceded by a 3 week history of blurry vision. She did not have vertigo, hearing loss, phonophobia, and no decrease in fluid intake. She was hospitalized after having a sudden loss of postural tone. Her vital signs were significant for orthostatic hypotension. Neurological exam was negative for Dix hall pike maneuver. She had a normal ear examination, negative Romberg test, and normal cerebellar function tests. Head, ear, nose, throat, neck, cardiac, respiratory, abdominal, extremity, and skin examinations were normal. Ophthalmologic examination showed worsening of visual acuity. Laboratory studies showed quinidine levels of 0.5mg/L (normal range 2-5mg/L). There were no EKG abnormalities and the MRI of her brain was normal. With IV fluid resuscitation, orthostatic hypotension resolved, and with discontinuation of tonic water, patient reported gradual resolution of her above-mentioned symptoms over the course of 3 weeks.

**DISCUSSION:** Quinine-induced neurotoxicity is known as cinchonism and it may include ataxia, headache, vertigo, orthostatic hypotension, syncope, confusion, and delirium. Tinnitus is common, occurring in 38% of reported cases. Deafness may occur in severe poisoning. The mechanism of neurotoxicity remains controversial. CONCLUSION Quinine-induced neurotoxicity are not limited to high doses of quinine as is seen in antimalarial drugs, but in low quinine doses as found in tonic water, with their effects present even at normal quinine serum levels.

**REFERENCES**

**Clinical Vignette**

**Bandeira, Joseph Saint Peter’s University Hospital (Nayan Kothari)**

**Cryptococcal meningitis and tuberculosis as AIDS defining illnesses in a newly diagnosed HIV infected patient**

**INTRODUCTION:** Opportunistic infections are common complications of advanced immuno-deficiency in individuals with Human Immunodeficiency Virus (HIV) infection. This is a case of concurrent cryptococcal meningitis and pulmonary tuberculosis (TB) as Aquired Immune Deficiency Syndrome (AIDS) defining illnesses in a previously undiagnosed HIV-infected 36 year old male.

**CASE REPORT:** This is a 36 year old male from Honduras who presented with obtundation, confusion and agitation. Vital signs showed temperature 102.4 F; otherwise normal. Physical exam revealed neck stiffness, equal & reactive pupils without papilledema, and no focal signs. CT of the head was normal. Spinal fluid (CSF) analysis showed significantly elevated protein, decreased glucose and 115 cells, all of them lymphocytes. Given the patient’s symptoms and CSF findings, subacute meningoencephalitis was suspected. Further investigation included CSF India Ink and culture showed encapsulated yeast, confirming the diagnosis of Cryptococcal meningitis. Sputum culture revealed AFB, confirming the diagnosis of TB, w/ normal CXR. HIV Ag/Antibody was reactive, with viral load <20 copy/mL, <1.3 log, and CD4 showed 4% helper, w/ absolute CD4 of 69. For Cryptococcal meningeal encephalitis, the patient was treated with Amphotericin B and Fluocytosine for 14 days, and discharged on Fluconazole. For active TB, quadruple therapy with Rifampin, Isoniazide, Pyrazinamide, Ethambutol was initiated, with addition of Pyridoxine and vitamin B6. Lastly, antiretroviral therapy for HIV was postponed to outpatient care given the risk of immune reconstitution inflammatory syndrome (IRIS).
DISCUSSION: TB is the most common opportunistic infection in patients with HIV, and when untreated, can accelerate the course of infection because plasma levels of HIV RNA increase in the setting of active TB. In contrast, plasma levels of HIV RNA decrease in the setting of successful TB treatment. Cryptococcosis occurs worldwide, mostly affects immuno-deficient individuals, and is the most common life threatening fungal infection in AIDS, with meningitis being the most common manifestation. Conversely, the initiation of effective cART therapy carries the risk of immune IRIS, in which a paradoxical worsening of pre-existing opportunistic infections may occur. This condition is particularly common in patients starting therapy with CD4 Tcell count <50 cells/μl who show a rapid decline in HIV load following initiation of cART. Therefore, it is recommended that initiation of HART be delayed in antiretroviral naive patients until 2-8 weeks following the initiation of treatment for TB.

CONCLUSION: Immunosuppression secondary to HIV has resulted in a growing number of clinical cases harbouring multiple opportunistic infections such as TB and cryptococcosis. Overlapping of symptoms and a delay in diagnosis are the main causes of increased mortality in such cases, and a high level of clinical suspicion is required. Early diagnosis and treatment is critical, and often lifesaving as in the case of this patient.

Resolution of Group A Beta-Hemolytic Streptococcus Sepsis-Induced Cardiomyopathy after IVIG and Steroid Treatment

INTRODUCTION: Myocarditis is clinically and pathologically defined as inflammation of the myocardium and is a common cause of acute cardiomyopathy. Its clinical presentation ranges from nonspecific systemic symptoms to fulminant hemodynamic collapse, cardiogenic shock and sudden death. Intravenous immunoglobulin (IVIG) therapy in acute myocarditis has been widely investigated and its use remains controversial.

CASE REPORT: A 41-year-old active female with history of hypothyroidism and no prior cardiac history presented with shortness of breath. Three days earlier, she had developed fever, chills, myalgias and joint pains followed by several episodes of vomiting and diarrhea. Initially her dyspnea was only on exertion but it progressed to being present at rest. She stated that her daughter had been recently diagnosed with streptococcal throat infection and was currently on antibiotics. She did not smoke, would drink a glass of red wine a month, her only medication was levothyroxine and she had no known drug allergies. On admission, she was in septic shock and treated with aggressive fluid hydration, which resulted in pulmonary edema and intubation. Broad-spectrum antibiotics, inotropes and stress dose steroids were started. She had elevated cardiac enzymes, chest x-ray showed a moderate right pleural effusion, EKG showed sinus tachycardia and non-specific T wave changes. Two sets of blood cultures grew group A Beta-hemolytic streptococcus. Transthoracic echocardiogram (TTE) showed left ventricular heart failure with global hypokinesis with a left ventricular ejection fraction of 40%. The patient remained in critical condition for the next 26 days as her course was complicated by multi-organ failure needing hemodialysis and other supportive measures. Two therapeutic thoracentesis were required for her recurrent pleural effusions refractory to diuretics. The severity of her condition prompted the administration of IVIG 85 g via intravenous bolus and methylprednisolone 60mg every six hours, followed by daily IVIG 40 g treatments that were discontinued after two doses. Patient was diuresed, inotropes were weaned, antibiotics were stopped and IV steroids were tapered down. On hospital day number 32 patient was discharged home on a beta blocker. Follow up TTE 6 months after discharge showed a normal left ventricular ejection fraction of 55% with no areas of hypokinesis.

DISCUSSION: Myocarditis can be regarded as a precursor to cardiomyopathy. Treatment for acute myocarditis is primarily supportive
following evidence based recommendations. Some clinicians still use steroids and IVIG to treat infectious myocarditis. While several case studies have shown some benefits of steroids and IVIG in some forms of myocarditis, randomized controlled trials have failed to show any benefit. Although IVIG treatment is controversial and its use cannot be routinely recommended, it is possible that in select cases of sepsis induced cardiomyopathy where there is no response to intensive supportive measures, IVIG and steroids may be used.

**Clinical Vignette**

Dumaswala, Komal
Saint Peter’s University Hospital (Nayan Kothari)

**Everolimus (Afinitor) induced Interstitial lung disease**

INTRODUCTION: Mammalian target of rapamycin inhibitors (m-TORi) well known as immune-suppressants for organ transplantation, are now also approved for use in various malignancies. Everolimus (Afinitor) was the first drug amongst this class to be approved by FDA for advanced hormone receptor positive and HER-2 negative breast cancer in post-menopausal women.

CASE: A 76 year old female presented to ER with worsening shortness of breath for four days associated with pleuritic chest pain, left arm and shoulder pain. The patient has a history of left side breast carcinoma 20 years ago status post mastectomy followed by chemotherapy, radiation and five years of tamoxifen therapy. She had recurrence of her malignancy with bone metastasis for which she was recently started on Everolimus (Afinitor) with Letrozole (Femara) two months ago. Two weeks after initiation she noticed shortness of breath with minimal exertion and hence after reading the drug information manual, stopped taking Everolimus. One week after being off the treatment, her symptoms improved. After consulting her oncologist, Everolimus was resumed at half the dose. Five to seven days after resuming the therapy, she again noted dyspnea which this time did not improve with discontinuation of the drug. On admission, her SaO2 was 96% on room air. Physical examination revealed velcro-like crackles in lower two-third of lung zones bilaterally. EKG as well as troponins were negative. She underwent CT angiogram of the chest revealing no evidence of pulmonary embolism but bilateral diffuse ground glass opacities. Everolimus was not restarted and patient was started on oral prednisone 40mg daily. On the third day of treatment, her symptoms, oxygen requirement on ambulation (SaO2 94% post-ambulation) and her crackles significantly improved. She was discharged home with a total five day course of oral steroid.

DISCUSSION: Like other m-TORi, Everolimus can also lead to interstitial lung disease. The time interval from exposure to development of pulmonary toxicity ranges from 3.4 to 36.7 weeks (median – 15.4 weeks) (2). NCI has graded it into 4 grades based on clinical severity of pulmonary involvement (3). Given the wide spectrum of pulmonary toxicity, prompt diagnosis and early non-aggressive management by drug discontinuation and steroids can prevent respiratory failure. CONCLUSION: This case highlights the possibility of developing non-infectious pneumonitis while on Everolimus therapy, which should be kept in mind given the increasing indications and uses of Everolimus in various cancers.


**Clinical Vignette**

Enjamuri, 
Saint Peter’s

**Sessile Serrated Adenoma- A deviant from Vogelgram**
### INTRODUCTION

Colorectal Carcinoma is the third most frequent cancer worldwide with more than one million incident cases. Approximately 10% of colorectal cancers arise from serrated polyps, most of which have hypermethylation of CpG-rich promoter regions that leads to inactivation of the DNA mismatch repair gene MLH1, resulting in microsatellite instability, and activation of mutations of the BRAF gene.

### CASE

58 year old Caucasian male with significant history of alcohol intake came to the emergency room with one week history of bright red blood mixed with stools. He did not have abdominal pain, dizziness. Review of systems is otherwise normal. Examination revealed conjunctival pallor. Digital rectal examination was normal. His heart rate was 84/min and blood pressure was 124/82. Labs revealed hemoglobin of 6.8mg/dl. His Hemoglobin two months ago was 8.2mg/dl. Esophagogastroduodenoscopy did not reveal active bleeding. Colonoscopy revealed 3mm sessile polyp in ascending colon with no active bleeding. The polyp was removed with piecemeal technique and injection chromoscopy was done. Histopathology had shown serrated adenoma. No active source of bleeding was identified.

### DISCUSSION

Serrated polyps includes hyperplastic polyps, sessile serrated adenoma (SSA), traditional serrated adenoma (TSA) and a combination of two or more characteristics, formerly classified as mixed polyps. Sessile serrated adenomas account for 3%–9% of all colorectal polyps and are frequently found in ascending colon. Serrated pathway as opposed to Vogelstein Adenoma carcinoma sequence involved BRAF and KRAS mutations. There is lack of studies of follow up intervals of serrated polyps and more accurate information about management of serrated polyps of the colon is not yet available. Randomized controlled studies need to be done to establish surveillance guidelines for SSA.

### REFERENCES

consult was ordered, and the patient was discharged the next morning after being setup with another internist. The etiology of this patient’s symptoms is still unknown, but she is now with a single provider who will gather the information from all of the other specialists, and assess the composite data along with input from the nephrologist, and will determine the etiology and therapeutic plan after a thorough and targeted evaluation is completed. When symptoms don’t easily fit into a recognizable pattern, intellectual rigor must be applied to determine the correct unifying condition. This patient should have been evaluated by an internist who would be dogged enough to rule out each of the potential etiologies, but humble enough to seek consultation from appropriate specialists. Her testing should have included a 24-hour urine collection for a Urine K-to-creatinine ratio to distinguish renal from extra-renal etiology. A BMP with concurrent ABG for acid/base status would further refine the etiology. Finally, antibody titers and renal biopsy can be pursued if an autoimmune condition adequately ties together the rest of the symptoms.

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Clinical Vignette
Florou, Vaia
Komodikis Gregoris
Saint Peter’s University Hospital (Nayan Kothari)
Moyamoya syndrome: not as rare as thought to be?

INTRODUCTION: Moyamoya syndrome is a rare vasculopathy characterized by bilateral stenosis or occlusion of the arteries around the circle of Willis, associated with a network of abnormally dilated collateral vessels. The incidence is 0.086 per 100,000 persons in Caucasians and 4.6 in Asian Americans. Here, we describe two cases diagnosed with Moyamoya syndrome.

CASES: Two middle-aged adults presented with neurologic symptoms. The first case, a 47-year-old Filipino male complained of a brief episode of anarthria and perioral numbness one day prior to presentation. Two weeks before, the patient complained of peripheral vision loss and headache lasting for a few minutes. His past medical history included hyperlipidemia and gout treated with allopurinol and gemfibrozil. His family history included CVA in his brother and father at 40 and later age respectively. He had quit smoking after 25 years and consumed alcohol occasionally. He did not have any focal weakness and was otherwise completely asymptomatic. Our second case, a 54-year-old Caucasian female presented with right-sided weakness and facial droop for three days. She did not have any speech or vision changes. Her past medical history included untreated DM. Her family history included embolic CVA in her father. She did not smoke or drink any alcohol. She was noted to have only right-sided hemiplegia on presentation. Brain imaging of both patients showed evidence of Moyamoya syndrome. The brain MRA of the first patient showed occluded left internal carotid artery (ICA), middle cerebral artery (MCA) and decreased flow within the left posterior cerebral artery (PCA). He had severe occlusion of the proximal right MCA with moyamoya collaterals. The CT head of the second patient showed a large, non-hemorrhagic left PCA distribution infarct with mass effect, 5 mm midline shift to the right and mild uncal herniation. An MRA of the head showed diffuse right ICA narrowing, occlusion of the right M1 segment with multiple moyamoya collaterals at the carotid terminus. There was complete occlusion of cervical and petrous segments of the left ICA. The left MCA branches were diffusely narrowed and the left PCA was completely occluded. Immediate medical treatment was initiated to both patients. The first patient remained asymptomatic, whereas the second patient gradually improved until she was discharged to a rehab facility.

DISCUSSION Moyamoya syndrome is a rare but increasingly recognizable cause of strokes. The natural history is highly variable. Prompt initiation of treatment achieves best outcomes. Surgical approach may be beneficial in selected cases.

CONCLUSION: These case reports highlight the atypical vascular findings of Moyamoya syndrome. Neurologic status more than age, at the time of presentation, is a long-term outcome predictor. Recognition and immediate initiation of treatment is critical.

REFERENCE R. Scott, E. Smith Moyamoya disease and Moyamoya syndrome.
Lupus enteritis: Uncommon presentation in patients with SLE

INTRODUCTION: Lupus enteritis (LE), an uncommon presentation in patients with SLE (systemic lupus erythematosus) manifests with abdominal pain, ascites, vomiting and diarrhea with characteristic imaging abnormalities of the bowel. It is defined as either vasculitis or inflammation of the small bowel, with supportive image and/or biopsy findings(1, 2).

CASE DESCRIPTION: 25 year old female presented with diffuse spasmodic abdominal pain, nausea with vomiting and diarrhea but no fever. Vital signs on admission included BP 107/79 with no orthostatic changes, HR 82/min, temp 98.4F. Physical examination was unremarkable except for abdominal distention. There was no jaundice, abdominal tenderness or organomegaly. Laboratory studies included WBC 15.1cells/cumm, 81% neutrophils, platelets 478/cumm, Hb 12.6gm/dl, MCV 87.7fl, ESR 39mm/1st hr, CRP 17mg/dl, BUN 25mg/dl, Sr. Creatinine 0.9mg/dl. UA showed 1.023 sp gravity, 10-20 WBC/HPF, trace blood, 0-2 hyaline casts LPF and 300mg/dl protein. Patient was hospitalized 3 weeks prior to this admission with similar complaints when she was 22 weeks pregnant. Patient had miscarriage during that admission and had no prior history of abortions. Imaging studies of chest and abdomen showed ascites and thickening of mucosal wall at ileum and jejunum. Ascitic fluid was aseptic exudate. Patient was diagnosed with possible SLE based on small joint pains, positive ANA and anti-DNA antibodies and was discharged home on prednisone. During this admission patient had a second CT of abdomen showing resolution of ascites however pronounced thickening of small bowel in ileum and jejunum was observed. Imaging findings included bowel wall thickening with target sign. Her symptoms resolved with high dose of steroids.

DISCUSSION: The clinical features of LE include abdominal pain, ascites, vomiting and diarrhea in the setting of lupus diagnosed by established criteria of SLE and characteristic imaging findings on CT of abdomen. Other clinical features include active SLE features and fever associated with laboratory abnormalities namely anemia, leukopenia or leukocytopenia, thrombocytopenia, moderately raised CRP and proteinuria. Typical CT findings include focal or diffuse bowel wall thickening, bowel dilatation and abnormal bowel wall enhancement (target sign), engorgement of mesenteric vessels (comb’s sign) and attenuation of bowel wall fat. Potential complications include bowel wall necrosis and perforation. Lupus enteritis is responsive to high dose steroids and immunosuppressant(1).

CONCLUSION: Lupus enteritis is a rare presentation of SLE with serious complications and morbidity if left untreated. Response to steroid therapy is excellent. Diagnostic criteria need to be established.

CASE: A 47 year old woman was admitted with a 4 day history of nausea, vomiting, fever and left lower quadrant (LLQ) pain. Medical History was significant for Limb Girdle Muscular Dystrophy (LGMD) since the age of 5, and scoliosis surgery at age 17. Family History was remarkable for LGMD in her father, who died of same. At presentation, she was febrile (101F), tachycardic (115/min), with BP of 102/65. Physical exam was normal except mildly tender LLQ. Labs were normal except leukocytosis of 13,000 with 14% bands, and mild anion gap metabolic acidosis. Blood culture grew Escherichia Coli. CT abdomen showed chronic sigmoid diverticulitis with adjoining mesenteric abscess corresponding with stage 1 of Hinchey classification of perforated Diverticulitis. Failure to improve on IV antibiotics prompted a repeat CT which showed an interval development of HPVG. Anticipating a difficult post-op extubation because of her muscular dystrophy, she was managed non-surgically, with broader spectrum antibiotics. Absence of clinical or radiologic signs of a graver etiology of HPVG such as gangrenous bowel was a reassurance. A follow up CT revealed increased HPVG along with extensive thrombosis within the intra-hepatic portal vein, Superior Mesenteric Vein, and Splenic Vein. Patient underwent laparotomy and sigmoid colectomy with Hartmann’s end colostomy. Resected abscess as well as a subsequent blood culture grew Candida albicans which is a lesser known etiology – 7% of pylephlebitis (1). She was treated with Micafungin, besides Heparin infusion for Portal Vein Thrombosis. Patient could not be weaned off ventilator because of her muscular dystrophy and eventually underwent tracheostomy that was gradually reversed two months later.

DISCUSSION: Pylephlebitis occurs due to an abdominal infection draining into the portal vein radicles, that initiates the clotting cascade. It may be preceded by HPVG. CT based early diagnosis has lately proved that in absence of a graver etiology such as bowel necrosis; Diverticulitis is a rather benign cause, and can be safely observed. (2)

CONCLUSION: The case illustrates a rare finding of a common disease, and reviews the available treatment options.

bloating. Significant on physical examination was body mass index 36.6 and tenderness to palpation on RLQ. Firstly, irritable bowel syndrome was suspected, however symptoms continued despite adequate dietary modification. Patient underwent multiple imaging and procedures, amongst EGD and colonoscopy. Results were normal except for Helicobacter pylori infection. Initial abdominal CT scan showed prominent lymph nodes in RLQ. Pelvic US and DISIDA scan were unremarkable. Subsequent abdominal CT scan reported a new rim of hypoattenuating tissue around infrarenal aorta. Pertinent laboratory data include: ESR 93 mm/h, CRP 76 mg/L, low serum iron and saturation and normal renal function test. Prednisone 50 mg by mouth daily was started and symptoms significantly improved, as well as dramatic decrease in ESR and CRP levels. The onset of CP is often characterized by non-specific signs and symptoms requiring a high clinical suspicious. Etiology is likely to be multifactorial and involve genetic and environmental factors and immune-mediated mechanisms. One popular theory suggests local inflammatory reaction against an antigen localized in the atherosclerotic plaques of the abdominal aorta. Both medical and surgical approaches have a role in the management of CP patients, but no established therapeutic algorithm exists. Autoimmune tests should always be evaluated in CP. Histopathologically findings such as adventitia inflammation and clinically by variable involvement of different arteries raise the issue of whether CP should be considered a large- vessel vasculitis. Therapy is largely empirical, based on the use of corticosteroids and immunosuppressants. Randomized controlled multicenter trials are needed.

INTRODUCTION: Hemophagocytic lymphohistiocytosis (HLH) is an aggressive, life-threatening syndrome of uncontrolled immune activation, excessive inflammation and tissue destruction. It is characterized by the presence of red blood cells, platelets, or white blood cells (or fragments of these cells) within the cytoplasm of macrophages.

CASE REPORT: A 54 year old woman who presented with 2 month history of worsening daily urticarial rash, diffuse bilateral cervical lymphadenopathy, fever and night sweats. She had consulted various physicians and got treated with antibiotics without improvement. She had been hiking on the east coast and she had tooth extraction and root canal a month ago. She or her family didn’t have allergy or immunodeficiency disease. On physical examination, she had significant bilateral lymphadenopathy in her neck, supraclavicular and axillary region. Oropharynx was clear. Lung sounds were normal and she didn’t have stridor or wheezing. Laboratory showed anemia with Hg 8.4, thrombocytopenia with platelets 105, ferritin 2062, CRP 88 and ESR 28. Blood cultures were negative. Ct neck with contrast showed extensive lymphadenopathy involving supraclavicular, pretracheal, Para tracheal, Para tracheal, axillary, intraparathyroid lymph node, jugular chain nodes, and diffuse lymphadenopathy around neck. She was tested for HIV, babesiosis and ehrlichia, all of which were negative. She was diagnosed with hemophagocytic syndrome. All of her inpatient antibiotics was stopped and started her on steroid treatment.

DISCUSSION: HLH is classified as primary genetic disorder or secondary HLH phenomenon. HLH can be triggered by infection or other immunologically activating events. Male-to-female ratio is 1:1. HLH can affect all age group. The cell types involved in the pathogenesis of HLH are Macrophages, Natural killer cells and cytotoxic lymphocytes. Phagocytosis can be seen on biopsies of immune tissues (lymph nodes, spleen, liver) or bone marrow aspirates. Most common presentation are diffuse lymphadenopathy, neurological symptoms, rashes, fever, multi organ involvement, anemia, thrombocytopenia, hypertriglyceridemia, elevated ferritin > 500 and Hemophagocytosis in bone marrow, spleen, lymph node, or liver. Treatment of HLH are immunosuppressive etoposide, steroids, methotrexate, and refractory cases needs to treat with anti CD52 monoclonal antibody. The prognosis is guarded with
overall mortality of 50%. Awareness of the clinical symptoms and of the diagnostic criteria of HS is important to start life-saving therapy in time.


246 188 Clinical Vignette John, Febin Oleg Alekseev, Trisha Saha Saint Peter’s University Hospital (Nayan Kothari) Acute Cholecystitis in Kawasaki disease

INTRODUCTION: Kawasaki disease (KD) is a medium vessel vasculitis with systemic and mucocutaneous manifestations. Involvement of gall bladder (GB) in KD disease is rare and its management is not clear1. We are presenting a case of acalculous cholecystitis in a young boy with KD.

CASE: A 4 year old previously healthy boy of Nigerian descent was brought to the ED with a 4-day history of fever (103.3°F) and a papular rash that started on his hands and feet and further spread to the trunk. Physical examination revealed edema of the extremities, bilateral non-exudative conjunctivitis, cracked lips, strawberry tongue, RUQ tenderness and 3/6 systolic murmur in left sternal border. Laboratory values on admission revealed a microcytic hypochromic anemia, Hb 10.8 g/dL (11.5-13.5), amylase 391 units/L (30-131), lipase 264 units/L (11-82), CRP 153 mg/L (0-5.0) and ESR 37 mm/hour (0-15). Amoxicillin therapy, initiated for suspected scarlet fever, failed to improve his condition. Persistent fever with the accompanying symptoms raised high suspicion of KD. Echocardiogram revealed mild dilatation of the coronary arteries and pericardial effusion. His liver function tests showed AST 106 units/L (17-59), ALT 111 units/L (21-72), direct bilirubin 2.2 mg/dL (0.1-0.5), and INR 1.3 (0.9-1.1). Although the initial RUQ ultrasound (US) was normal, the repeat US showed enlargement of the GB and pericholecystic edema without common bile duct dilatation suggestive of acalculous cholecystitis. HIDA revealed severe intrahepatic cholestasis. MRCP confirmed the aforementioned findings. Patient was started on high dose aspirin (80mg/kg) and IVIG was administered on the second and fifth day post-admission. Serum transaminase levels responded to therapy, yet the fever and hyperbilirubinemia persisted. A 3-day course of pulsed solumedrol (30 mg/kg) was initiated on the seventh day post-admission. Patient became afebrile with steady decline of total bilirubin and marked clinical improvement, although, his echocardiogram revealed progressive coronary artery dilatation. He was discharged on the twelfth day post-admission.

DISCUSSION: KD, also known as mucocutaneous lymph node syndrome, is a vasculitis presenting as persistent fever (>5 days), bilateral bulbar conjunctival injection, oral mucous membrane changes, peripheral extremity changes, polymorphous rash and cervical lymphadenopathy. It is common in infants and children. About 15% of patients have GB involvement secondary to GB wall vasculitis1. GB distension in early KD is associated with IVIG resistance and increased risk of coronary aneurisms. Steroids help to prevent coronary aneurism when IVIG resistance is suspected. Currently, there are no guidelines for the treatment of cholecystitis in KD; this case of cholecystitis was successfully managed conservatively. Conclusion Acalculous cholecystitis, a marker of severity, can mislead the clinical diagnosis of KD and delay the administration of IVIG.
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<td>Tropical pulmonary eosinophilia (TPE) results from a hypersensitivity reaction to Wuchereria bancrofti and Brugia malayi. It usually affects people living in Southeast Asia, India and certain parts of China and Africa. The diagnosis is often delayed in the non-endemic area because the clinical presentation is often confused with acute or refractory asthma. A 45 year old man from South India presented with complaints of chest pain associated with shortness of breath. Chest pain started a day back and was more on the left side. He had progressive shortness of for one week, presented to urgent care and was prescribed quinolones without response. His symptoms had been increasing for the past 3 months. He has also noticed 3 kg weight loss over the past 3 months. Past medical history is significant for asthma like symptoms comprising of nocturnal dyspnea and occasional wheezing. Lung examination revealed decreases breathe sounds bilateral with intermittent expiratory wheezing. LABS: wbc - 15.8, HB- 12.2, platelets- 233 and eosinophil count - 67. Ig E serum- 499. ESR- 80. Imaging: CT Angio- Bilateral ground glass, airspace and interstitial opacities. Relatively centrally located rather than peripherally. Discussion: Patient’s presentation of chest pain with elevated troponins but no EKG changes related to ST segment elevated MI or acute coronary syndrome. CT angiogram ruled out pulmonary embolism. Patient was initially admitted to ICU and was started on steroid. He responded well to the treatment diagnosis of tropical pulmonary eosinophilia was entertained considering that patient was from endemic area for filariasis. He underwent lung biopsy which did not show any signs of vasculitis ruling out Churg- Strauss disease. Patient was treated with doxycycline as Diethyl Carbamazine(DEC) is not available in USA, his condition improved on follow up visits. Conclusion: Pulmonary infiltrates with peripheral eosinophilia can be caused by a variety of conditions including infectious, inflammatory, and allergic etiologies: •Allergic bronchopulmonary aspergillosis •Chronic eosinophilic pneumonia •Vasculitis such as Churg-Stauss syndrome, PAN and Wegener’s granulomatosis •Idiopathic hypereosinophilic syndrome (IHES) •drug reactions Although these diseases share similar clinical presentation and laboratory features, there are certain additional criteria that must be met in order to diagnose TPE. These include: •Residence in a filariasis-endemic area like the Indian subcontinent and Africa, marked peripheral eosinophilia, •Absence of microfilaria in peripheral blood due to rapid clearance of opsonized microfilaria, •Increased anti-filarial antibody titer, •Elevated serum IgE level and a •Response to Diethylcarbamazine (DEC), which is the recommended treatment. It is important to diagnose and treat this condition early because it may lead to long-term sequelae of pulmonary fibrosis or chronic bronchitis with chronic respiratory failure.</td>
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<th>Kovaleva, Alexandra</th>
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<td>INTRODUCTION: Babesiosis is one of the underdiagnosed parasitic disease. Risk factors for severe babesiosis include an age of &gt; 50 years, male gender, asplenia, HIV/AIDS, malignancy and immunosuppression. The fatality rate is 5% among all hospitalized patients but it is much higher (20%) among immunocompromised patients. The growing rate of reportable cases in endemic regions including New Jersey should be aware of this disease especially among patients with risk factors of severe babesiosis.</td>
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<td>CASE DESCRIPTION: An 89 y/o male was presented at first admission with increase fatigue, generalized weakness, forgetfulness, fever, symptoms of dysuria and blood in urine for one week since patient’s trip to Connecticut. One of the patient’s hobbies was visiting graves. Patient has known history of Benign prostatic hyperplasia, Hypertension, Hyperlipidemia, Diabetes Mellitus type 2 and Sick Sinus Syndrome s/p pacemaker. On physical examination generalized pallor present, mini mental status was consistent with mild dementia; no lymphadenopathy, no rashes or lesions. Pedal edema 2+ was present on lower extremities b/l. Patient was admitted to general medical floor, initially with increased Total bilirubin and history of fever. Right upper quadrant US was</td>
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unremarkable. Was suspected hemolytic anemia: LDH was elevated, reticulocytes was elevated, platelets count was in normal range, Coombs was negative. By hematology consult ANA was positive and autoimmune hemolysis versus possible Paroxysmal Nocturnal Hemoglobinuria was suspected. Peripheral smear did not reveal any abnormalities at that time. The patient was discharged to subacute rehab with close follow up Hematology Oncology for possible bone marrow biopsy. Four days later was readmitted due to low Hb 6.5 and worsening of previous symptoms. At this time peripheral smear was positive for intracellular organisms with babesia species 2% RBCs infected. Patient was started on Atovaquone and Azithromycin regimen for total 10 days, was discharge after 3rd day of treatment in good condition with significant improvement in general.

DISCUSSION: This case demonstrates the missing diagnosis in elderly person with suspected malignancy and autoimmune hemolysis without evidence of parasites at first admission. Although babesiosis is rare, but should always be considered for any patient who presented with flu like symptoms and has recently resided or travel to an endemic area at any group age, especially elderly, immunocompromised patients due to severity of the disease and increased mortality rate.


A Difficulty in Approach to Signet Ring Cell Gastric Cancers

INTRODUCTION: Signet Ring Cell Carcinoma (SRCC) is a histological type based tumor of epithelial origin that is most commonly found in the glandular cells of the stomach. It has historically been known to be a highly aggressive and deadly disease, but recent studies have shown that it’s prognosis is comparable to other types of gastric adenocarcinoma (1).

CASE: A 50yr old Male from Costa Rica with no past medical history presented to the emergency room with abdominal pain that is associated with nausea, vomiting and weight loss of 25 pounds in the last 3 months. Abdominal CT scan showed a markedly distended stomach with circumferentially thickened antrum and pylorus. Gastroduodenoscopy revealed an obstructing exophytic mass in the prepyloric region. Biopsy of the mass showed poorly differentiated invasive adenocarcinoma with signet ring cells. This tumor was staged as T3N2Mx with the help of CT scan and Endoscopic Ultrasound. He was started on neoadjuvant chemotherapy with Epirubicin, Cisplatin and 5-Fluorouracil and to be reassessed for operative management after the first cycle of chemotherapy.

DISCUSSION: According to WHO Classification, SRCC contains more than 50% extracellular mucinous pools. Signet Ring cell gastric cancers may have inherent chemo resistance leaving many clinicians unsure of the right approach of management. As per the PRODIGE Trial, the strategy of primary surgery for SRC tumors, followed by adjuvant chemotherapy, will improve overall survival at 2 years when compared to a standard peri-operative chemotherapeutic strategy(4). Messager et al showed that perioperative chemotherapy was found to be an independent predictor of poor survival with HR = 1.4, 95% CI 1.1-1.9, P = 0.042(5). Many surgeons have a divided opinion about the proper approach of SRC cancers and further studies are still needed to support this data.

### Clinical Vignette

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**A unique presentation of cryptococcal meningitis**

**INTRODUCTION:** Cryptococcal meningitis is an invasive fungal infection due to Cryptococcus neoformans that has become increasingly prevalent in immunocompromised and HIV infected patients who are at risk for this infection when their CD4 count declines to less than 100 cells/mm³. The most common presenting symptoms are fever, malaise, photophobia, vomiting and headache. The incidence of cryptococcal meningitis has declined in patients who have access to antiretroviral therapy.

**CASE REPORT:** A 33 year old Mexican gentleman came to the emergency room because of headache for 9 days duration. Patient stated that the headache started 9 days prior, with a dull aching pain in the frontal forehead, symmetrical on both sides, 1/10 in intensity did not cause any discomfort in performing his daily activities, it was gradual in onset, constant without progressing over the duration of this episode and non radiating. Headache was not aggravated by light, sound, chewing or lying flat, and Tylenol relieved the headache completely. No Nausea, Vomiting, Fever, blurred visual disturbance, neck stiffness, confusion, no history of trauma. Headache worsened progressively over the next few days and was not relieved with medications and patient came to ER.

Past history is significant for HIV diagnosed in 2007, took highly active antiretroviral therapy till 2010, TB lymphadenitis treated for 6 months. Admissions vitals were stable. Physical exam was normal. Labs showed leukocytosis, hyponatremia and hypokalemia. A lumbar puncture was done, and his csf was positive for cryptococcal antigen, also were the cultures positive for C. Neoformans. Indian ink was positive for encapsulated yeast. Imaging was normal. His CD4 count was 9. Patient was started on Amphotericin and Flucytosine as induction therapy for two weeks. Later he was started on fluconazole as maintainence therapy for eight weeks. His later CSF cultures did not show any growth and patient was subsequently discharged after a week.

**CONCLUSION:** This is a unique presentation of cryptococcal meningitis because of the duration taken for the presentation of the symptoms as the patient was off highly active antiretroviral therapy for four years. In general Cryptococcal meningitis is known for the quicker onset of symptoms within 2-4 months post discontinuation of highly active antiretroviral therapy. This case demonstrates that there is a sudden inciting factor for Cryptococcal meningitis in a patient who are off highly active antiretroviral therapy.

### Clinical Vignette

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**ATYPICAL PRESENTATION OF PYLORIC STENOSIS WITH SEVERE HYPONATREMIA**

**INTRODUCTION:** Pyloric stenosis is narrowing of the opening from the stomach to the duodenum, due to the hypertrophy of the pylorus muscle. It is seen in children but also occurs rarely in adults, secondary to scarring from chronic peptic ulceration. It manifests with vomiting and dehydration with metabolic alkalosis, hypokalemia and hypochloremia. Pyloric stenosis causing severe
hyponatremia is a rare clinical presentation. Hyponatremia is a decrease in serum sodium < 136 meq/L. Clinical manifestations of hyponatremia are primarily neurologic including headache, confusion, stupor, seizures and coma.

CASE REPORT: A 78 year old Indian male presented with abdominal discomfort which progressed to a feeling of abdominal distension over 10 days. Discomfort was worsened by eating at night and relieved by using PPIs. He also reports associated nausea and decreased oral intake. No fever, chills, constipation, diarrhea, sick contacts, unintentional weight loss, NSAID or alcohol use. Past history is significant for Polio, GERD, Hypertension and Hyperlipidemia. GERD was diagnosed with complaints of heart burn, bitter taste in the mouth and he has been using Ranitidine but never had an endoscopy. Admissions vitals were stable. Physical exam was normal. Labs showed anemia and leukocytosis with Sodium of 90. Serum and urine osmolality, specific gravity and urine analysis were normal. X ray abdomen showed distension of bowels with no air fluid levels. Three liters of fluid was removed from the abdomen by Nasogastric lavage. CT abdomen showed mild to moderate distension of abdomen. Patient initially was volume resuscitated with 3 liters of normal saline and then progressively switched from 3% NS to 2%NS to NS with a goal correction 4-6mEq/24 hours. Neurological checks for seizures and obtundation were done. Sodium increased from 90 to 142 over two weeks. Patient had UGI endoscopy which showed H pylori, pyloric stenosis with one non-bleeding gastric ulcer. Patient was treated with triple therapy and had gastro-jejunostomy.

CONCLUSION: This is an unusual presentation of severe hyponatremia secondary to pyloric stenosis. Pyloric stenosis usually manifests as vomiting leading to dehydration and contraction metabolic alkalosis. This patient had no vomiting and had severe hyponatremia out of proportion with the dehydration. In general, older patients with hyponatremia develop more symptoms than younger healthy patients. Symptoms are also more severe with faster-onset hyponatremia. This patient had a Glasgow Coma Scale of 14/15 with no neurological deficits in spite of having sodium of 90. Hyponatremia was most likely acute on chronic, hypovolemic and multifactorial secondary to poor oral intake, diuretic use and pyloric obstruction.

Lung biopsy revealed nonmucinous adenocarcinoma without angiolymphatic, pleural or stromal invasion, consistent with the diagnosis of bronchoalveolar carcinoma. She had a relentless disease progression thereafter, and she succumbed to the disease approximately 9 months after initial cancer diagnosis.

**DISCUSSION:** The overall incidence of pemetrexed-related pulmonary toxicity is 3.5%. One study revealed that preexisting interstitial lung disease is a risk factor for pemetrexed-related pulmonary toxicity (2). Since the patient had an exposure to pemetrexed, we were led to entertain the diagnosis of interstitial lung disease secondary to pemetrexed delaying the revelation of bronchoalveolar carcinoma. This case exemplifies how bronchoalveolar carcinoma can masquerade as interstitial lung disease. It arises from type II pneumocytes and may manifest radiologically as a solitary peripheral nodule (SPN), multifocal disease, or a rapidly progressing pneumonic form, which can spread rapidly from one lobe to another. Pretreated lung cancer can be potentially treated with tyrosine kinase inhibitor if ERBB mutation is present. Our patient's disease was progression was very rapid and encompassing both lungs and multiple lobes, due to which, no definitive treatment could be instituted. Conclusion: Bronchoalveolar carcinoma can occasionally mimic interstitial lung disease. In a patient with obvious risk factors for interstitial lung disease, such as medication in our patient, diagnosis of cancer may be rendered difficult.

**REFERENCES:**

**Autoimmune Hemolytic Anemia - from undiagnosed asymptomatic to symptomatic spectrum: A case report**

Autoimmune hemolytic anemia (AIHA) is a rare disease, characterized by the presence of autoantibodies, most frequently of the IgG isotype, directed against erythrocyte surface antigens. It affects women more often than men. The disease may be primary (idiopathic), or secondary to another underlying illness. Idiopathic AIHA accounts for approximately 50% of cases. The most common causes of secondary AIHA include lymphoproliferative disorders (e.g., chronic lymphocytic leukemia, lymphoma) and other autoimmune disorders (e.g., systemic lupus erythematosus, rheumatoid arthritis, scleroderma, ulcerative colitis). Rarely it follows the use of certain drugs, such as penicillin or #945;-,methylhipp. The management of secondary AIHA is often challenging. A 20-year-old female presented to our ER with headaches from past one-week. She has no history of fever, night sweats, cough, chest or abdominal pain. There was no history of recent travel or close sick contacts. She had no urinary symptoms. Her menstrual cycle was regular. She did not use tampons. She also did not give history of sun sensitivity or hair loss. She worked in a hair salon and was in a monogamous relationship and used protection for birth control. Family history was negative for any blood disorder. On physical examination:
- **GENERAL:** She did not look unwell.
- **HEENT:** Pale conjunctiva, coated tongue, rest within normal limits.
- **CHEST:** Revealed good air entry bilaterally. Percussion of the spine did not elicit any tenderness.
- **HEART:** Showed a regular rate and rhythm. No murmurs or gallops.
- **ABDOMEN:** Flat, soft and nontender. No hepatosplenomegaly. No inguinal adenopathy.
- **SKIN:** No petechiae or hematomas. No rashes.

**Pertinent lab findings:**
- **Hgb:** 4.1. She was found to be Direct coomb's positive when tested with Anti-IgG and Anti-C3b, C3d and had low haptoglobin and LDH was elevated at 390 and bilirubin was high at 1.8. The iron study was normal with a high ferritin of 319. Her ESR was 119 and Reticuloscyte count was 18. Her Hgb electrophoresis showed Hgb A1 of 66.4%, Hgb C of 32.6 %. She was also found to be ANA(1:640) and Anti SSA positive. A review of her blood smear showed marked anisopoliokilocytosis with hypochromic cells. There were some spherocytes. There was abundant polychromasia and some nucleated red blood cells as well orthochromic normoblasts were seen. She was symptomatic during her
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<td>Clinical Vignette Penigalapati, Damodar Kanishk Agnihotri, Pratik Patel Saint Peter’s University Hospital (Nayan Kothari)</td>
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<td>Hospital (Nayan Kothari)</td>
<td>INTRODUCTION: Germ cell tumors (GCTs) are classified as extradural if there is no evidence of a primary tumor in the testes or ovaries. In adults, the most common extradural sites are the anterior mediastinum, retroperitoneum, as well as the pineal and suprasellar regions. Mediastinal seminomas constitute approximately 2 to 4 percent of mediastinal masses and one-third of malignant mediastinal GCTs. Mediastinal seminomas occur predominantly in men between the ages of 20 and 40. The common presenting symptoms of mediastinal seminoma are chest pain, dyspnea, cough, weight loss and superior vena cava syndrome. CASE: A 22 y/o male presented with 1 week history of neck swelling, difficulty in swallowing, SOB on exertion and dry cough. No Hx of weight loss, fever/chills. His vital signs included temperature 97.7, BP 133/73, HR 99, RR 16, O2 saturation 99% at room air. His physical examination revealed prominent neck swelling along with bilateral cervical lymphadenopathy. Labs WBC 7.1, Hgb 13.6, Hct 41.5, Platelets 233, Na 144, K 5.1, Cl 101, Co2 32, BUN 18, Cr 0.94, AFP 506, Tumour HCG 41, LDH 876. CXR showed mediastinal widening, CT soft tissues of neck with contrast showed large mediastinal mass extending into the base of the neck which encases the trachea as well as the great vessels of the neck, bilateral supra-clavicular and jugular lymphadenopathy. US of the scrotum and testes showed no evidence of testicular mass. US guided biopsy of the left jugular lymph node revealed malignant germ cell tumour consistent with typical seminoma. Immunohistochemistry studies showed the tumor cells are focally positive for HCG and negative for AFP. Flow cytometry leukemia and lymphoma panel showed no evidence of a B-cell or T-cell clonal lymphoproliferation. The patient was treated with chemotherapy. He has completed 2 cycles, repeat CT thorax after chemotherapy shows residual activity and the tumour was inoperable. DISCUSSION: Pure seminomas do not cause an elevated serum AFP, but can have positive beta HCG in one third of patients. However, molecular studies have demonstrated AFP mRNA in minute quantities in pure seminoma. Several case reports have described pure seminoma with borderline elevations in serum AFP (10.4 to 16 ng/mL). Higher serum AFP concentrations are considered diagnostic of a nonseminomatous component of the tumor or hepatic metastases. If the presence of elevated serum AFP is confirmed, patients should be treated as if they had an NSGCT. Serum AFP concentrations &gt; 10,000 microg/L are found almost exclusively in patients with NSGCTs or hepatocellular carcinoma. CONCLUSION: This case is presented to highlight the importance of AFP elevation in patients with nongonadal seminoma in determining treatment strategy. An elevated serum AFP in pure Seminoma indicates that nonseminomatous elements present. Such tumors are treated as nonseminomatous GCTs.</td>
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failure with creatinine of 8.29 mg/dL. Colonoscopy showed a large (5.2 cm x 3.3 cm x 1.7 cm), multi-lobulated polyp mass in the mid-transverse colon, histopathology report indicated tubulovillous adenoma with high-grade dysplasia and this was later resected. The renal failure did not improve after transfusing 4 PRBC and completing 3 sessions of hemodialysis, reason why additional investigation was warranted. In subsequent laboratory investigations markers for different autoimmune diseases were normal, viral tests were negative, and urine immunofixation showed monoclonal free kappa band though serum immunoglobulins were normal. The renal biopsy revealed the presence of kappa light chain deposition disease with moderate to severe tubular atrophy and interstitial fibrosis. A bone marrow biopsy and aspirate showing 27% plasma cells and a high serum β2-microglobulin of 30.8 mg/L, confirmed the diagnosis of Multiple Myeloma (Durie-Salmon stage IIB); a bone scan did not show lytic lesions and he was started on outpatient chemotherapy.

**DISCUSSION:** Multiple myeloma accounts for approximately 10% of all hematologic malignancies. In recent case reports, it has been presented with synchronous solid tumors involving lung, colon, breast and prostate. The development of a colon adenoma is mediated by a mutation in the Adenomatous Polyposis Coli (APC) tumor suppressor gene, the same gene that is a negative regulator of the Wnt/wingless signaling pathway that has been recently shown to play a role in the proliferation of multiple myeloma cells. Proper clinical management in cases like this, due to its uncommonness, is not well established. In the present case, the objective was to control and solve the gastrointestinal involvement in order to proceed with targeted therapy against multiple myeloma; stabilizing the associated kidney failure in the meantime.


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257 96 Clinical Vignette Ramos, Esther Alejandro Herrera Saint Peter’s University Hospital (Nayan Kothari)

"Pied Tabetique": A Case of Charcot’s Foot

An often overlooked complication of diabetes, Charcot’s foot or neuropathic arthropathy, involves a long and challenging course in management. First described in syphillis patients in 1883 by Jean-Martin Charcot and his colleague Feré as “pied tabeticum”, Charcot’s foot is a neuropathic joint disease involving progressive destructive arthritis after sustained mechanical stress associated with a loss of pain sensation, proprioception, or both. Diabetes mellitus is now the most common cause of Charcot’s Foot. This is a case of a 58 year old woman with an average HgA1c of 10, suffering from retinopathy, neuropathy, vasculopathy, recurrent cellulitis and osteomyelitis from chronic foot ulcers resulting in multiple wound debridements, toe amputations and bilateral Charcot foot stemming from uncontrolled type 2 diabetes. The patient was referred to podiatric surgery for immobilization of lower extremities via total contact casts (TCC) on alternate lower extremities. She subsequently underwent reconstructive surgery of her right foot with an external fixator. The differential diagnosis for Charcot’s foot includes infection, such as osteomyelitis, cellulitis, abscess, deep tissue infection, or other inflammatory processes such as acute venous thrombosis or gout. The diagnosis is a clinical one. Acute presentation of Charcot foot is usually an erythematos, swollen mid-foot, with a greater than 3° C temperature difference between the affected and the contralateral foot. Pain may not be a prominent feature due to neuropathy. Studies have indicated that PET/CT scanning in conjunction with MRI may be useful to help diagnose acute Charcot ostearthropathy as well as monitoring for resolution. Management is focused on immobilizing and offloading in a total contact cast (TCC) for a recommended initial 6 to 8 weeks, with biweekly change of cast, for acute Charcot ostearthropathy. Alternative modalities for which a clear benefit still needs
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to be established include bisphosphonates and intranasal calcitonin. Surgical correction of the deformity rather than longitudinal management with accommodative bracing has been found to be more favorable in patients with recurrent ulceration or unstable foot as it can achieve limb salvage and eventual ambulation. However, surgical correction is best avoided in most patients given that correction of skeletal deformity following nonunion of a fracture in a neuropathic foot has a relatively high failure rate (over one-quarter of patients). This occurs mainly in patients who have active ulceration relating to the deformity at the time of operation; thus, surgery should be postponed until the ulcers have healed.

Clinical Vignette

Rastogi, Ujjwal
Saint Peter’s University Hospital (Nayan Kothari)

Lyme Carditis in an avid golfer: A diagnostic challenge and a therapeutic dilemma

Cardiac manifestations occur in about 5% of Lyme infections and stem from the involvement of the cardiac conduction system, resulting in varying degrees of sino-atrioventricular block. Occasionally, Lyme infection may also present with myopericarditis. Unlike isolated conduction node disease, myocardial involvement presents a great diagnostic and therapeutic dilemma for the physician. Case Report: A 68-year-old male Cardiologist, with history of Prediabetes and Benign Prostatic Hyperplasia, presented with new onset exertional dyspnea and palpitations. Of note, he remained fairly active with excellent exercise tolerance until the day of presentation. The patient was an avid golfer and ran on the treadmill 5 days a week. He did not recall any tick bites, rashes or fever. Review of systems was otherwise negative. Physical examination revealed a well-nourished male who appeared his stated age. Cardiac exam revealed irregularly irregular rhythm, and a variable intensity of S1. The remainder of the physical examination was unremarkable. Electrocardiograms revealed intermittent Wenckebach with markedly prolonged PR interval varying between 290-350ms and a transient episode of atrial fibrillation/flutter with AV block. On admission, his vitals were blood pressure 136/67, pulse 75, 98% oxygen saturation on room air, temperature 98.0 F. Lyme antibodies titers IgM (3/3 bands) and IgG (5/10 bands) were positive. Rest of the complete blood count and comprehensive metabolic panel were within normal limits. The patient was promptly treated with intravenous Ceftriaxone, and within 36 hours, the patient’s arrhythmias began to resolve. By 48 hours the patient’s PR interval had shortened to 230ms. The patient was discharged on three weeks course of oral Doxycycline. Discussion: Lyme infection should be suspected in any patient who engages in high-risk activities or lives in endemic areas. Our patient, an avid golfer living in a Lyme endemic area, presented with atrial fibrillation, and complete AV block with no prior cardiac disease or risk factors. Management of Atrial fibrillation in the setting of Lyme carditis poses many challenges, as many of our traditional algorithms used to treat arrhythmias would be deemed unsafe. Electrical Cardioversion in the setting of an inflamed myocardium could be pro-arrhythmogenic and fatal. Pharmacological cardioversion may uncover underlying AV nodal disease caused by Lyme and induce complete heart block. For the same reasons, Class I, II, III, and IV antiarrhythmic drugs traditionally used for rate control, cannot be used in this setting. Finally anticoagulation traditionally used for stroke prevention, would be contraindicated if concomitant pericardial involvement is present for the fear of causing hemopericardium. The broad range of clinical manifestations often result in a delay of diagnosis or misdiagnosis. Thus the treatment algorithm is different, and depends upon prompt diagnosis and antibiotics.

Clinical Vignette

Sarasan, Ashley
Saint Peter’s University

Severe Anemia causing Grade III dyspnea and pitting edema in profound Hypothyroidism
BACKGROUND: Anemia in hypothyroidism is multi-factorial. There may be hypoproliferation in addition to Fe deficiency secondary to menorrhagia in women. Severe anemia can present with dyspnea and pitting edema often being confused as CHF presentation. Here we present a case where profound hypothyroidism masked the clinical presentation of severe anemia.

CASE: 50 yo F came to the hospital with c/o swelling of b/l legs and severe dyspnea on exertion which had been progressing over the past couple of years. Since last couple of weeks, she could barely walk 10 mins before getting short of breath. She did not have a history suggestive of cardiac etiology. She has had very heavy menstrual flow since the past several months which were initially attributed to her perimenopausal state. Physical exam showed hypothyroid facies, coarse voice, peripheral pitting edema of lower extremities and non pitting edema of upper extremities, delayed relaxation phase of DTR, and cardiovascular exam showing a functional grade II ejection systolic murmur. On evaluation she was found to have severe anemia with a hemoglobin of 3 with microcytic hypochromic anemia. Thyroid profile showed a TSH of 58.5 and Free T4 of 0.29. At this point, the dyspnea as well as the pitting edema was attributed to her severe anemia. She was transfused 4 pack units of blood before addressing her myedema and after her condition was stabilized, she was started on Levothyroxine IV at first and then switched over to oral dose. Subsequently the patient’s condition improved, pitting edema drastically reduced in 2 days and she was no more dyspneic on exertion. She was discharged home on oral levothyroxine 125mg daily.

CONCLUSION: Severe hypothyroidism can cause severe life threatening anemia. Thyroid hormones directly or indirectly, through erythropoietin, stimulate the growth of erythroid colonies. Deficiency of the hormone leads to hypoproliferation; with microcytic anemia while 10% develop macrocytic anemia. Pernicious anemia develops in association with autoimmune thyroiditis. And severe Fe deficiency from menorrhagia in women can lead to severe anemia resulting in dyspnea and pedal edema. Checking thyroid profile is important in these patients who do not have a cardiac etiology for the pitting edema and dyspnea.

INTRODUCTION: Dermatomyositis has been described as paraneoplastic syndrome. Paraneoplastic symptoms caused by a malignancy but not directly related to tumor invasion are the result of a wide variety of tumor-derived biologic mediators, such as hormones, peptides, antibodies, cytotoxic lymphocytes, autocrine and paracrine mediators.

CASE: A 73 year old male presented with sudden onset of weakness of bilateral upper and lower extremities. He woke up with an inability to move his arms and to get himself out of bed. He reported no sensory loss or loss of bowel or bladder function. No history of dysphagia, diplopia or dysarthria. He reported rash involving his face, arms and back two weeks prior to developing weakness which he attributed to poison ivy and was treated with oral steroid taper. No history of tobacco use or alcohol. No family history of cancer. Physical exam revealed a moderately built and nourished male oriented to time, place and person. Cranial nerve exam was normal. Severe weakness of bilateral deltoid, biceps and triceps in the upper and severe weakness of quadriceps in lower extremities was noted. Reflexes absent in upper and lower extremities. Normal position sense and light touch in all four extremities. Lab studies include WBC 12.0, Hemoglobin 11.7, platelets 284, ANA positive with titer of 1: 2560, aldolase 40, anti DNA- negative, anti smooth muscle negative, calcium 7.9, C-RP 60, ESR-23, C3-normal, HIV nonreactive, Hepatitis B &C non reactive, Lyme negative, Anti mitochondrial Antibody negative, SS-A and SS-B negative, TSH 10, thyroid peroxidase Antibody- negative, Vitamin B12 341, CPK 5046. EMG was consistent with inflammatory myopathy. Muscle biopsy showed acute severe inflammatory myopathy consistent
with dermatomyositis. CT Abdomen and pelvis showed cecal mass suspicious for adenocarcinoma with regional pericecal adenopathy.

**DISCUSSION:** Dermatomyositis a rare spectrum of inflammatory myopathy has been associated with malignancies. Predictive signs of malignancy include older age at onset, male sex, elevated ESR, initial higher serum CPK level, higher AST/ALT and low albumin.

**CONCLUSION:** Recognition of paraneoplastic syndromes is important, as it may lead to an early diagnosis of cancer. The clinical severity of the symptoms can be used as a guide to the extent of response to underlying tumor therapy. The quality of life of the patient is affected, therefore the palliative treatment of paraneoplasia is very important.

**REFERENCES**
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261 135 Clinical Vignette Shah, Jatin

Saint Peter’s University Hospital (Nayan Kothari)

**Naproxen Induced isolated uvular angioedema: Quincke’s disease**

**INTRODUCTION:** Angioedema is characterized by submucosal or subcutaneous swelling, which affects lips, tongue, oral cavity, larynx, pharynx and subglottic tissues. The pathogenesis of angioedema is thought to involve generation of bradykinin and complement-derived mediators that increase vascular permeability and subsequent extravasation of fluid. Diagnosis of angioedema is made clinically, based on a suggestive history and physical findings.

**CASE REPORT:** A 30 year old gentleman complains of discomfort in his throat for 1 day. He describes seeing a ball in his mouth. He does not have skin rash, drooling of saliva, difficulty in breathing or change in voice. He has neck pain 2 days before this and has taken naproxen for pain relief. He has never taken naproxen in the past. He or his family doesn’t have allergies or angioedema. He has never smoked, does not drink alcohol and never used any illicit drugs. His medication is naproxen for pain relief. On examination, his uvula was edematous and erythematous. Lung sounds were normal and he doesn’t have stridor or wheezing. His C4 complement level was normal. Uvular angioedema or Quincke’s disease was diagnosed and treatment with glucocorticoids, antihistamines and antacids were started. Improvement was rapid and his uvular edema resolved completely within 24 hours. He was discharged and underwent outpatient follow up. He was advised to avoid naproxen in the future and keep EpiPen with him.

**DISCUSSION:** Isolated uvular angioedema is immediate type 1 hypersensitivity reaction. It can be seen as a complication of general anesthesia, ACE inhibitor use or NSAIDs use. It should be promptly recognized and treated as it can lead to obstructive respiratory symptoms which may require emergency intubation. Hereditary angioedema is commonly seen in patients with family history of allergies or angioedema and suspected in patients with low complement C4 level. Treatment with glucocorticoids results in rapid resolution.

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<td><strong>A Case of Bird Fancier’s Lung</strong></td>
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<td><strong>INTRODUCTION:</strong> Extrinsic allergic alveolitis, is an immunologically induced inflammatory disease involving lung parenchyma and terminal airways secondary to repeated inhalation of an inciting agent in a sensitized host.</td>
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<td><strong>CASE DESCRIPTION:</strong> A 23 year old, non-smoking, white male, without significant past medical history, was admitted for worsening shortness of breath and intermittent fever. His symptoms started as a “respiratory tract infection” four months prior to admission. He was initially treated with beta agonists followed by an oral prednisone taper with significant relief. However, his symptoms returned soon after discontinuing steroids. He next presented to the emergency department with fever, shortness of breath and an oxygen saturation of 86%. On exam, he had inspiratory crackles. A CT showed diffuse ground glass nodularity in both lungs. The patient was questioned regarding respiratory exposures and noted that he kept pet parakeets. A bronchoscopy with cell counts and transbronchial biopsies showed evidence of an inflammatory process. Cultures were negative with the working diagnosis of extrinsic allergic alveolitis, the patient was started on prednisone 60 mg daily. He experienced improvement in symptoms over the first week of treatment. His oxygen saturation improved to 97% on room air. The patient was counseled against exposure to suspicious antigens. In a few weeks, after weaning off of the steroids, the patient again returned with shortness of breath and hypoxia. He was still keeping parakeets. He underwent a thorascopic biopsy with findings characteristic of extrinsic allergic alveolitis. He is currently on a six week steroid taper and doing well.</td>
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<td><strong>DISCUSSION:</strong> Extrinsic allergic alveolitis is characterized by a monocytic infiltrate in the small airways and lung parenchyma. Inciting agents may be derived from fungal, bacterial, or animal proteins or reactive chemical sources and have been recognized in a variety of occupations and recreational activities. Imaging reveals diffuse ground glass nodules or infiltrates. Pathological findings include non-caseating granulomas and interstitial and luminal alveolitis. Treatment is avoidance of antigen exposure as well as corticosteroids. Based upon his exposure history, we believe our patient suffered from “bird fancier’s lung” due to exposure to avian antigens from his pet parakeet.</td>
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<td><strong>CONCLUSION:</strong> Extrinsic allergic alveolitis is an inflammatory lung disease which can occur due to a wide variety of inhaled antigens. Physicians should be aware of the key clinical findings of this process and perform a thorough exposure history in a patient with suggestive radiographs and symptoms. The cornerstone of treatment is exposure avoidance.</td>
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<td><strong>Ocular Myositis</strong></td>
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|     |     | **INTRODUCTION:** Ocular Myositis: a benign idiopathic inflammatory disease capable of affecting any orbital structure is a subgroup of the “idiopathic inflammatory orbital syndromes.” A non-infective, non-specific orbital inflammation without identifiable local or systemic causes (1) with two major forms: limited oligosymptomatic ocular myositis and severe exophthalmic ocular myositis. There
is ongoing discussion as to whether it is a manifestation of other systemic inflammatory diseases.

**CASE:** A 37 year old South Korean woman diagnosed with bipolar disorder, left corneal ulcer, and scleritis presented with worsening right eye pain for three months. The pain was sudden in onset, beginning along the medial aspect of the right eye, gradually involving the entire orbit with radiation to the right scalp and forehead. She experienced horizontal double vision on rightward gaze which progressed to involve her entire visual field. On examination there was mild proptosis of the right eye with minimal periorbital erythema and sclera injection; the globe was nontender. Pupils were equally reactive to light with intact visual fields, there was no nystagmus and dilated fundoscopic examination was normal. Pain was elicited with any right lateral gaze and the right eye did not cross the midline. There was no scalp tenderness or temporal bruits. The remaining exam, including the thyroid, was normal. MRI of the orbits revealed right medial rectus muscle enlargement with retro-bulbar fatty infiltration. Solumedrol 60 mg IV for Ocular Myositis was begun with immediate improvement. She was discharged on Prednisone with follow-up with a primary physician and her ophthalmologist. During the steroid taper she began to have symptom recurrence resulting in a dose increase providing symptom relief.

**DISCUSSION:** The reported incidence of Ocular Myositis is 4.1−6.3% of all orbital disorders. The true incidence is unknown due to lack of a universally accepted definition of the disease (2). Before diagnosing Ocular Myositis it is important to rule out more common diseases such as: Giant Cell Arteritis, Glaucoma, cranial nerve palsy, Thyroid-associated orbitopathy, orbital cellulitis, etc. Contrast-enhanced orbital MRI is the most sensitive imaging study demonstrating swelling, signal hyperintensity, and enhancement of isolated orbital adnexa(3). Standard therapy is corticosteroids 1-1.5 mg/kg/d for 1 to 2 weeks with a prolonged taper, with improvement within days. Select patients experience recurrences and may require immunosuppressive therapy involving TNF, high dose IVIG or rituximab(4).


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**Clinical Vignette**

Singh, Gagan

Saint Peter’s University Hospital (Nayan Kothari)

**Orthostatic Hypotension secondary to Autonomic Dysfunction in Type 2 Diabetes**

**INTRODUCTION:** Orthostatic hypotension (OH), defined as decrease in systolic blood pressure > 20 mm Hg or a decrease in diastolic blood pressure > 10 mm Hg during the first 3 minutes of standing or a head-up tilt on a tilt table, is a classic manifestation of sympathetic vasoconstrictr (autonomic) failure in patients with diabetes mellitus (DM). In adults with DM in whom orthostatic hypotension has been well categorized, it may reflect the impact of duration of DM, poor diabetic control or both and prevalence has been reported to be between 3% and 35%.

**CASE:** 51-year old male with a history of Hypertension and Diabetes Mellitus presented with a chief complaint of dizziness for 3 days, described as lightheadedness aggravated on standing from a supine position along with occipital headache, weakness and fatigue impairing his daily activities. On physical examination his supine blood pressure was 113/83 mmHg. The remainder of examination,
except for peripheral sensory neuropathy was normal. Laboratory showed Hemoglobin of 13.1, Blood Glucose of 447 and HbA1c of 13.4. The patient was resuscitated with fluids and antihypertensive medication was discontinued. On day 2 of hospitalization the patient’s dizziness mildly improved, his supine BP was 162/99 with heart rate of 94, standing BP was 81/57 with no change in heart rate. Autonomic dysfunction secondary to uncontrolled diabetes mellitus was diagnosed. The patient was started on pyridostigmine and was counseled regarding non pharmacologic strategies such as performing physical counter-maneuvers, raising the head of the bed, and ingesting adequate salt and fluid.

DISCUSSION: BP changes to posture have been observed to be a reliable indicator of sympathetic autonomic dysfunction even in diabetic patients. Long-standing poor glycemic control predisposes patients to microvascular complications and autonomic neuropathy, of which OH is one. Variability in heart rate, a compensatory mechanism, probably declines at about three times the normal rate. Among the agents used to treat orthostatic hypotension, only midodrine has been subjected to a large, multicenter, placebo-controlled trial (1) The presence of both elevated SBP and OH in the same individual makes treatment difficult as measures to treat the former will exacerbate the latter. In the index case, pyridostigmine was the only reasonable option along with adequate control of diabetes mellitus (2).

CONCLUSION: OH is a frequent occurrence in DM. In a patient, with contraindication to the use of midodrine or fludrocortisone, pyridostigmine is a potential choice. Autonomic dysfunction seems particularly resistant to the restoration of normoglycemia, thus measures targeted at preventing this terrible complication should be emphasized.


265 136
Clinical Vignette Singh, Yojna Amrit Misra PhD Saint Peter’s University Hospital (Nayan Kothari)

Post Colonoscopy Ischemic Colitis: Is it real after colonoscopy?

INTRODUCTION: Ischemic colitis (IC) is the most common subtype of Ischemic Bowel Disease. It is the result of an acute, often self-limiting, compromise in colonic blood flow, leading to reflex mesenteric vasoconstriction and ischemia. IC is typically seen in elderly patients with underlying cardiovascular risk factors. However, recently, a rare variant of IC: post-colonoscopy ischemic colitis (1-2), has been documented, though only a few case reports exist. These patients tend to be younger and lack many of the traditional risk factors.

CASE: A 50 y/o female with no underlying medical condition and no acute gastrointestinal complaints underwent screening colonoscopy. A standard bowel preparation was performed: 2 doses of Polyethylene Glycol (238g) at 6 hour intervals and Bisacodyl (10mg) the night prior. Colonoscopy revealed normal colonic mucosa with a single 6mm polyp, which was snared without complication. Over the next 2 days, she experienced an abrupt onset of intermittent but copious rectal bleeding, with lower abdominal cramping, and passage of a single solid blood streaked stool. Her vital signs were normal with BP of 106/56 without orthostatic change. Physical examination was unremarkable except for lower abdominal tenderness. Digital rectal examination showed no signs of occult or overt bleeding. Laboratory studies were notable for leukocytosis with left shift (WBC 15.2, Neutrophils 82%) and a mildly accelerated prothrombin time at 9.9 seconds. Repeat colonoscopy confirmed no bleeding from polypectomy site. However, a region of colon between 20 and 40 cm from the rectum showed ulceration and bleeding, interspersed with normal
mucosa both proximal and distal. In the previous colonoscopy this region had showed no signs of colitis. She was managed conservatively with intravenous fluid hydration and bowel rest and within next 24 hours, experienced remarkable improvement. Histology of the colonic mucosal biopsy confirmed the diagnosis of IC. However, the stool also tested positive for Clostridium Difficile (C.Diff) toxin A/B. Although the histology was characteristic for ischemic colitis, metronidazole therapy was given for possible Clostridium Difficile Associated Disease (CDAD).

DISCUSSION: This report describes development of transient colitis in a patient with both clinical presentation and histological analysis consistent with ischemic colitis. Despite positive stool toxins, her rapid recovery prior to starting metronidazole, and the lack of prior precedence of iatrogenic C.Diff infection post colonoscopy, we propose that the patient was likely colonized, making the stool toxin finding incidental.

CONCLUSION: While rare, ischemic colitis should be considered in the diagnosis of post colonoscopy lower GI bleed, even in the presence positive stool antigens.

however not in all cases. Other causes for headache in this setting include pneumocephalus, subdural hematoma, and subdural hygroma. With re-expansion of the brain, the subdural hygromas may resolve. Complications of subdural hygromas are transformation into a chronic subdural hematoma. If initial treatment measures fail the clinician should widen the differential diagnosis, as any delay in treatment can be detrimental.


267 123 Clinical Vignette Thurston, Rhea Saint Peter’s University Hospital (Nayan Kothari)

DUODENAL CARCINOMA, A RARE GASTROINTESTINAL TUMOR

INTRODUCTION: The incidence of primary small bowel carcinoma is extremely infrequent (1% of all GI malignancies) and the subset of duodenal carcinomas is even more uncommon (0.3% of all GI malignancies). This case represents an elderly female presenting with nonspecific symptoms and the ultimate discovery of a tumor that was initially unsuspected.

CASE DESCRIPTION: An 88 y/o F was brought to the emergency room with a day history of vomiting associated with vague abdominal pain. The vomiting was described as several episodes of bilious material, small in volume followed by small amounts of dark red blood. The abdominal pain was sudden in onset, generalized and not associated with any other symptoms. Her co-morbid conditions included hypertension, atrial fibrillation, metallic aortic valve replacement, hypothyroidism, remote history of breast cancer and ischemic cerebrovascular accident with residual left sided hemiparesis. On presentation, vital signs were within normal limits. Positive physical examination findings were limited to dark red blood seen in the mouth. Gastrointestinal exam was unrevealing. About 700 cc bilious fluid was aspirated by NG tube. Admission hemoglobin was 10.9. CT scan of the abdomen and pelvis showed a dilated stomach and duodenum with no dilated loops of bowel distal to the duodenum. Upper GI endoscopy revealed a large near obstructing malignant looking mass with no active bleeding in the 2nd part of the duodenum. The biopsy results: well differentiated adenocarcinoma. The family was informed and given the patient’s multiple co-morbidities, a decision was made not to undergo any further aggressive treatment measures.

DISCUSSION: Primary duodenal adenocarcinoma is rare and as such, there is very limited data in the literature. The annual incidence is estimated to be 5,300 cases with 1,100 deaths each year. The most common location of all duodenal carcinomas is in the ampullary region of the 2nd part of the duodenum which was where the primary tumor of this patient was located. The peak incidence is usually in the sixth decade of life and presenting symptoms in order of decreasing frequency include abdominal pain, weight loss, nausea and vomiting, jaundice and hemorrhage. There was no need to suspect metastatic disease in this patient and ideally, patients with limited disease may benefit from a pancreaticoduodenectomy with negative margins. However, given her current Karnofsky of 30-40%, major surgery may not have provided an additional sustained benefit and was therefore avoided. The five year
survival rate is >40% for patients that undergo curative surgery but for non-resected is estimated to be about 0%.

**Clinical Vignette**

**Vakharia, Rushabh**

**Saint Peter’s University Hospital (Nayan Kothari)**

**Segmental Motor Paresis After Herpes Zoster Infection**

**INTRODUCTION:** There are many complications of a Herpes Zoster infection, however some are hard to recognize. This case describes one of the latter situations.

**CASE DESCRIPTION:** An 82 year old man with a history of atrial fibrillation presents for isolated weakness of his left lower extremity. About 3 weeks prior to presentation, the patient experienced pain and rash in the L5-S1 dermatome region in the same limb. He was diagnosed with shingles and underwent the appropriate treatment for the same. Around that time period, he started developing frequent falls and had to start using a walker because he felt weak in the left leg. This weakness progressively worsened and so he presented to the hospital. The initial physical exam revealed decreased strength of the left hamstrings, plantar flexion, inversion and eversion of foot, and the tibialis anterior. It also revealed a rash in his posterior thigh, and the dorsal aspect of the foot. He never had any involvement of his face or arm. A CT Head done at the time of presentation did not show any signs of ischemic or hemorrhagic stroke. Although the presence of atrial fibrillation makes the possibility of a stroke very high, the onset of the symptoms and gradual nature of the findings indicate an alternate etiology of the problem.

**CONCLUSION:** This case shows a complication of a Herpes Zoster infection that is not commonly encountered. With symptoms of a stroke and a setting of Afib, this can be a difficult diagnosis to make. Nerve conduction and EMG studies can be useful in this scenario. Recovery from this is common, however the length of time required is variable and treatment mainly consists of physical therapy.


**PLURALITAS NON EST PONENDA SINE NECESSITATE**

**Veedu, Janeesh**

**Saint Peter’s University Hospital (Nayan Kothari)**

**PLURALITAS NON EST PONENDA SINE NECESSITATE**

**CASE:** 30 year old female presented to the hospital with weight loss, diarrhea, nausea and vomiting. CT abdomen showed markedly distended stomach with thickening of distal stomach. Endoscopy showed an infiltrative non circumferential mass in pre-pyloric stomach which was a poorly differentiated adenocarcinoma with signet ring cell features. She also had Krukenberg tumor and biopsy confirmed metastatic disease in bladder. She received a palliative bypass surgery and three cycles of chemotherapy. A review of her history showed that she had six ER visits in 28 months with four CT and one MRI of the abdomen, two RUQ ultrasound and one trans-vaginal ultrasound. During each visit and imaging she was given diagnoses of cholelithiasis, biliary colic, pelvic pain, hydronephrosis, ureteral stones, retroperitoneal fibrosis, ovarian cyst, gastritis, bacterial gastroenteritis, and epigastric pain of unclear etiology. She had a cholecystectomy for epigastric pain because an ultrasound showed gallstones without evidence of cholecystitis or abnormal LFTs. She also received a ureteral stent for the hydronephrosis.

**DISCUSSION:** High rates of gastric cancer (GC) are seen in South America mainly on the Western coast while mortality rates are the highest in the mountainous regions (Pacific coast). The prevalence of highly conducive conditions for H pylori infection i.e. poor
sanitation, overcrowding and dietary factors such as reduced fruit and vegetable intake together with restricted access to refrigeration are the possible factors.1,2 High altitude regions such as Columbia have a higher incidence and mortality which could be due to clustering of genetic risk factors and H pylori virulent strains.3,4 Reports suggest 300% rise in proximal tumors as well as diffuse type in some parts of the continent.

CONCLUSION: Our case emphasizes that plurality should not be posited without necessity (“PLURALITAS NON EST PONENDA SINE NECESSITATE”). The patient was given different diagnoses each visit and no single unifying diagnosis. A gastric outlet obstruction was never considered even though numerous imaging showed gastric dilatation and wall thickening. We also intend to highlight the higher incidence of gastric cancers in young South Americans.


Improving Telemetry utilization in Medicine Teaching Service

INTRODUCTION: Telemetry is a commonly used tool to monitor patients at high risk for arrhythmia or sudden death. While the American Heart Association (AHA) has issued guidelines for appropriate use of telemetry, unnecessary use continues to lead to wastage of resources. The goal of our study was to develop and assess the efficacy of an intervention to reduce inappropriate utilization of telemetry.

METHODS: We performed chart audits of patients (N = 122) admitted to Cardiac Progressive Care Unit (CPCU) or Med/Surg unit with cardiac monitor for the month of April 2014. Residents and faculty were educated regarding the AHA classification (Classes 1, 2 and 3) and indications for telemetry. Using pre-designed forms, data were again collected for patients (N = 77) admitted to Cardiac Progressive Care Unit (CPCU) or Med/Surg unit with cardiac monitor for the month of September 2014. The residents were required to complete the forms on daily basis. Data were compared across intervention (pre vs post), based on location (CPCU vs Med/Surg). Primary endpoints included number of patients on telemetry and length of stay (LOS). Data were compared using two-sample t-test or chi-square.

RESULTS: In the Med/Surg unit, there was a significant decrease in the total number of patients (61 vs 32, p<0.002), driven mostly by a reduction in the number of patients admitted with Class 3 indications (34 vs 8, p<0.001). LOS, however, was not significantly decreased [4.29(1.35) vs 3.65(0.48), p=0.27]. In the CPCU there was no significant difference in number of patients (61 vs 45, p=0.12) or LOS [3.68(0.30) vs 4.47(0.43), p=0.14] following intervention.

DISCUSSION: The effects in this study were modest, however they highlight the importance and efficacy of our intervention.
Reduction in the number of patients on the Med/Surg unit prevents unnecessary testing and downstream costs that the hospital would otherwise incur from continuous telemetry monitoring. We observed a trend towards decrease in number of patients admitted to CPCU during the post-intervention period, reducing hospital cost by approximately $200,000. Additionally, the increase in LOS on CPCU, though not statistically significant, could be due to patients with appropriate diagnoses being admitted to CPCU.

**CONCLUSION:** Simple intervention like educating physicians can play a crucial role in increasing awareness of the telemetry guidelines. Together with frequent monitoring it has the potential to significantly improve hospital resource utilization.

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**Research**

Agnihotri, Kanishk

Nilay Patel, S Arora, N Patel, A Badheka, C Shah, P Patel, D Singal

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**Drug Eluting Stents utilization in the United States from 2006 to 2011**

**BACKGROUND:** Concerns about long term safety of drug eluting stent led to decline in their usage in 2006. Recovery of usage occurred in 2009 after long term data was reinvestigated. We attempt to address the question of DES usage post 2009, up till 2011.

**METHODS:** We queried the Healthcare Cost and Utilization Project’s Nationwide Inpatient Sample (NIS) between 2006 -2011 using ICD-9-CM procedure code, 36.06 (bare metal coronary artery stent) or 36.07 (drug eluting coronary artery stents) for Percutaneous Coronary Intervention (PCI). All procedures performed in patients >= 18 years were included. The NIS is the largest all payer datasets which represents 20% of all the hospitals in the US. All analyses were performed using the designated weighting specified to the NIS data base to minimize bias.

**RESULTS:** A total of 665,804 procedures were analyzed representative of 3,277,884 procedures in the United States. Safety concerns arising in 2006 reduced utilization from 90% of all PCIs performed in 2006 to 72% in 2007 and reached a nadir of 69% in 2008. After publication of salutary outcomes data in 2008, utilization increased to 76% of all stents in 2009 and plateaued (74% in 2010 and 75% in 2011).

**CONCLUSION:** After initial acceptance, panic and recovery, the utilization DES has reached a plateau since 2009. Currently DES is used in three fourth of all stents in the United States.

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**Research**

Ahmed, Nazir

Raghuvansh P Sah, Naoki Takahashi, Suresh T. Chari

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**Prevalence and Significance of Elevated serum IgG4 levels in Primary Sclerosing Cholangitis**

**BACKGROUND:** IgG4-related disease (IgG4-RD) is an emerging multi-system disorder characterized by tumefactive lesions, IgG4-rich lymphoplasmacytic infiltrates, storiform fibrosis, elevated serum IgG4 levels and steroid responsiveness. Involvement of the biliary tree, known as IgG4-related Sclerosing Cholangitis (IgG4-SC), presents as biliary strictures that mimic biliary involvement in Primary Sclerosing Cholangitis (PSC), cholangiocarcinoma or pancreatic cancer. While the diseases it mimics have a uniformly poor prognosis, IgG4-SC itself is an uncommon but steroid-responsive disorder. However, untreated IgG4-SC can rapidly progress to biliary cirrhosis needing liver transplant; therefore it is imperative to identify and differentiate IgG4-SC from PSC. Currently, in the absence of histology or concomitant AIP, IgG4-SC is difficult to diagnose. In a large cohort of subjects managed as PSC we determined if: (A) those with elevated IgG4 differ in their profile from PSC (B) the subset of PSC with elevated IgG4 would more likely have other organ involvement as an indicator of IgG4-SC (C) frequency of transplant was different in those with elevated IgG4.
METHODS: From Mayo Clinic’s electronic medical record database we retrospectively identified patients aged >18 years seen from 2001 to 2014 who had a clinical diagnosis of PSC and had serum IgG4 measured (n=803). We reviewed their medical records, imaging and lab values. Radiologic images (MRI/CT) were reviewed in patients with elevated IgG4 and their age-gender matched PSC patients with normal IgG4 levels.

RESULTS: Of the 803 PSC patients who had serum IgG4 levels measured (age 46±15 yrs, 66% males, serum IgG4 70.4±97.2mg/dl, median 41.5), 102 were seropositive with elevated serum IgG4 levels (13%; age 46±17 yrs; 75% males, mean IgG4 259.4±160.7mg/dl), 30% (n=31) of whom had IgG4 >2-fold upper limit of normal (2xULN). Prevalence of inflammatory bowel disease (IBD) among seropositive (55%) was similar to seronegative patients. 14 (14%) of seropositive patients underwent liver transplantation, which was similar to the frequency of liver transplantation in patients with normal IgG4 (11%). Among the 86 seropositive patients who had initial imaging (MRI or CT scan) available for review, 17 (20%) had evidence of pancreatic or renal manifestations of IgG4-related disease, among which 7 had >2X ULN IgG4 elevation. In contrast, the prevalence of pancreatic manifestation was 0 (p<0.0001) and renal manifestation was 3% (p<0.0001) among age- and gender- matched sample of 88 seronegative patients in whom initial imaging was reviewed.

CONCLUSION: Elevation of serum IgG4 is seen in 13% of patients with PSC. IgG4 seropositive and seronegative PSC patients are similar in demographics and seem to have similar rate of liver transplantation. Evidence of other organ involvement on imaging suggestive of IgG4-related disease is seen in only 20% of seropositive patients. To our knowledge, this is the largest cohort of IgG4 seropositive patients followed for Primary Sclerosing Cholangitis.

273. 129 Research Charalambous, Marinos Savvas S. Constantinides, Panayiotis Marellos, Elpidoforos S. Soteriades, Christos P. Christou Saint Peter’s University Hospital (Nayan Kothari)

Transradial Approach for Coronary Angiography and Intervention in Octogenarians

BACKGROUND: The transradial approach has been shown to have lower complication rates when compared to the transfemoral approach especially in the elderly, where vascular complications are more common. However, it may be more challenging due to age related variations in vascular anatomy. We report our experience with the transradial approach in octogenarians undergoing coronary angiography and intervention.

METHODS: We retrospectively reviewed 2,455 consecutive patients over a 30 month period (June 2006 – December 2008) at our center. A total of 108 octogenarians were identified. Baseline characteristics, procedural success rates and major complications were recorded.

RESULTS: About half underwent coronary angiography (49/108=45%) and the remaining underwent PCI (59/108=55%) via either the femoral (18/108=17%) or the radial approach (90/108=83%). Two thirds (72/108) were male. The mean age was 82.7 years (range 80 – 93). A significant proportion of the above procedures were urgent (42/108=39%). Previous CABG was documented in 11% (12/108). Adjunctive therapies were used as deemed necessary (IABP in 1 case, temporary pacing wire in 3). Only 2/90 radial procedures (2.2%) had to be aborted and were successfully performed via the femoral route, both due to extreme subclavian artery tortuosity and spasm. The use of the radial access increased dramatically over the study period (9/18=50% in the first 6 months,
36/43=84% in the following 12 months and 45/47=96% in the final 12 month period). No significant access related complications occurred either in the radial or the femoral group. All, but one PCIs (chronic total RCA occlusion) were successful. Three (3%) in-hospital deaths occurred, all three in patients that presented with cardiogenic shock and acute myocardial infarction.

CONCLUSIONS: Coronary angiography and intervention is safe in octogenarians. The radial access is equally feasible as in the younger age groups, with very low rates of failure in the hands of experienced operators (2.2%). Radial access should be the preferred route in the elderly to avoid femoral access site complications in this vulnerable group of patients.

274. 130 Research Charalambous, Marinos Elpidoforos S. Soteriades, Savvas Constantinides, Christos Christou Saint Peter's University Hospital (Nayan Kothari) Left Versus Right Radial Approach for Accessing Left Internal Mammary Artery Grafts

BACKGROUND: Transradial approach (TrA) has now been established as the routine method for coronary angiography and percutaneous coronary intervention (PCI), in many centers around the world. Despite its benefits, TrA was considered to be relatively contraindicated in patients with a history of coronary artery bypass grafting (CABG) involving the left internal mammary artery (LIMA). Our aim was to examine the feasibility and safety of the TrA in this group of patients and evaluate any potential benefits when performing the procedure through the Left Radial (LR) versus the Right radial (RR) artery.

METHODS: We performed 5,479 transradial catheterizations between Jan 2006 and Dec 2013. In our center, we established TrA as the routine method for elective, urgent and emergency procedures (primary or rescue PCI). Baseline characteristics, procedural success rates and major complications were recorded.

RESULTS: A total of 247 transradial catheterizations were performed on patients with previous history of CABG involving the LIMA. Among these catheterizations, the initial approach was through the LR artery (209 cases, 84.6%), the RR artery (33 cases, 13.4%) and the Right Femoral (RF) artery (5 patients, 2%). The LIMA graft was successfully accessed in all 209 cases performed through the LR artery (100% success rate), in 32 out of 33 cases performed through the RR artery (97% success rate) and in all 5 RF artery cases (100% success rate). In 1 case, it was not possible to access the LIMA graft through a RR approach but this was possible after crossing over to a LR approach. No major complications were noted in any of the procedures involving access to the LIMA graft.

CONCLUSIONS: Our findings indicate 100% procedural success rate when attempting access to the LIMA graft through the LR artery as compared to 97% success rate through the RR artery. Although, both approaches are associated with a high success rate, we identified a preference of our operators to perform such procedures through the LR (84.6%) instead of the RR artery (13.4%). In our opinion, TrA is feasible in patients with history of CABG involving the LIMA and our study provides supportive evidence for its efficacy and safety.

275. 263 Research Florou, Vaia G Komodikis, D Penigalapati, N Patel, H Rana, CS Pitchumoni Saint Peter’s University Hospital (Nayan Kothari) Can liver tests predict prognosis in septic patients without pre-existing liver disease?

INTRODUCTION: Multiple prognostic scores are routinely used in assessing disease severity of acutely ill patients. APACHE II and SAPS II scores have been successfully validated for septic patients. Liver dysfunction is usually associated with overall poor prognosis but there are no studies that correlated liver tests abnormalities with APACHE-II and SAPS-II scores. Most studies evaluating liver
METHODS: A retrospective cohort study was performed at Saint Peter’s University Hospital to assess the prognostic significance of liver tests in septic patients in regards to in-hospital mortality and total length of stay. In addition we evaluated liver tests in septic patients and their correlation with APACHE-II and SAPS-II scores. All patients admitted to ICU from emergency department with severe sepsis from November 2011 through October 2013 were included. Patients with known liver disease, solid tumors with liver involvement, hepatobiliary source of infection and re-admitted patients were excluded from further analysis. Pearson’s correlation method was used to understand the association of two continuous variables. Liver tests were divided in 3 equal tertiles. Chi square and Wilcoxon rank sum test was used to compare categorical and continuous variables respectively. SAS 9.2 and SPSS 22 were used for data analysis. P value <0.05 was considered significant.

RESULTS: 305 consecutive patients were included for analysis. Among the liver tests low albumin levels were significantly associated with poor outcomes. The lowest tertile of albumin (< 2.4 g/dl) was significantly associated with higher in-hospital mortality (33.46%) compared to other tertiles [2nd tertile (2.5 g/dl - 3.1 g/dl): 23.53% and 3rd tertile (>3.2 g/dl): 13.46%; P Value <0.01]. Low albumin (<2.4g/dL) was also associated with higher ICU length of stay (Median, Interquartile range [lowest tertile: 3 days (2-6) compared to highest tertile: 2 days (1-3); P value: <0.01] and higher overall hospital stay [lowest tertile: 8 days (6-12) compared to highest tertile: 6 days (4-11)]. Lowest tertile of albumin was significantly associated with higher APACHE II score (Median, Interquartile range [lowest tertile: 25 (22-30) compared to highest tertile: 19.5 (14-25); P value: <0.01]) and higher SAPS II score (Median, Interquartile range [lowest tertile: 49 (41-58) compared to highest tertile: 38 (28-48); P value: <0.01]).

CONCLUSION: Our study shows that after excluding patients with pre-existing liver disease, low albumin is an independent predictor of mortality in septic patients requiring ICU. Low albumin levels are also associated with increased hospital length of stay and higher APACHE II and SAPS II scores. This study differs from others which included patients with preexisting liver diseases and suggested that bilirubin elevation indicated poor prognosis.

INTRODUCTION: Non-Alcoholic Fatty Liver Disease in type 2 diabetes mellitus contributes to increased diastolic dysfunction

INTRODUCTION: Type 2 diabetes mellitus (T2DM) is known to be associated with cardiac diastolic dysfunction1. NAFLD is the surrogate marker for inflammatory state seen in T2DM associated with metabolic syndrome (MS). In MS, diastolic dysfunction is hypothesized to occur from abnormal active myocardial relaxation and increased passive stiffness due to lipotoxicity and increased cytokine activity1. It is not known whether NAFLD in T2DM increases the risk for diastolic dysfunction. The aim of this study was to evaluate the effect of NAFLD on cardiac function by echocardiography, in patients with T2DM.

MATERIAL AND METHODS: We retrospectively analyzed electronic medical records of 2,561 patients with T2DM who were treated at a tertiary care centre for diabetes in southern India between 1991 to May 2014, and had a liver ultrasound (US) with cardiac echocardiogram done for various reasons. Patients with history of alcoholism and hepatitis were excluded. NAFLD was diagnosed by liver US. The two cohorts, NAFLD (n=1618) and non-NAFLD (n=943) were compared for echocardiographic findings. All statistical analysis was done using SPSS software.
RESULTS: The baseline demographics of the study population were compared between the two cohorts. Diabetic patients with NAFLD were found to have a higher inter-ventricular septal wall thickness (9.4±1.9cm vs. 9.0±1.8cm, p<0.001), indicative of increased cardiac hypertrophy. Even though ejection fraction was higher in T2DM patients with NAFLD (61.9±8.1% vs. 60.9±8.5%, p=0.001), they had a lower end diastolic volume (94.2±28.7cc vs. 110.1±70.9cc, p<0.001) and lower end systolic volume (35.8±17.8cc vs. 43.5±24.6cc, p<0.001), indicating presence of diastolic dysfunction. T2DM patients with NAFLD were associated with increased cardiac hypertrophy and diastolic dysfunction compared to those without NAFLD.

CONCLUSIONS: NAFLD is associated with diastolic dysfunction probably due to cardiac hypertrophy in this population of T2DM patients of south-east Asian origin. With a large south-east Asian population with T2DM in New Jersey, this study is clinically relevant to our practice.


Effect of Initial Training Months on PCI Outcomes Over 11 Years: A National Perspective

BACKGROUND: The relationship between the initial months of training on outcomes in patients undergoing percutaneous coronary interventions (PCI) is unclear.

METHODS: An 11 year cross-sectional study based on the Nationwide Inpatient Sample from 2000-2010 was performed. Subjects were identified by International Classification of Diseases, 9th Revision CM procedure code, 36.06 and 36.07. Teaching hospitals were considered if they had an American Medical Association approved residency program. The study population was divided into 3 month quartiles. Hierarchical multivariate mixed models were created. Primary outcome was in-hospital mortality and secondary outcome was a composite of in-hospital mortality and peri-procedural complications.

RESULTS: A total of 867,380 PCIs were identified representing a total of 4,311,469 PCIs done at teaching hospitals in the US during the study period. Overall, in-hospital mortality was 1.2% and complication was 7.2%. Primary and secondary outcome in the initial quarter of the training period (July – September) were lower (odds ratio (OR): 0.89 (0.83-0.95) and 0.95(0.93-0.98)), compared to January-March (p<0.001). Similarly there was a decrease in length of stay by 0.1 days (0.13 days-0.07 days) but an increase in cost of hospitalization by $238 ($80-$395).

CONCLUSION: The initial months of training in a teaching hospital are associated with lower in-hospital mortality, peri-procedural complications, length of stay but an increased cost in patients undergoing PCI.

Impact of hospital volume on in-hospital mortality, peri-procedural complications and outcomes in renal artery stenosis stent placement in the United States

CONCLUSION: The initial months of training in a teaching hospital are associated with lower in-hospital mortality, peri-procedural complications, length of stay but an increased cost in patients undergoing PCI.
| Patel, D Singal | (Nayan Kothari) | BACKGROUND: Numerous studies are available to assess the impact of hospital volume on in-hospital mortality, length of stay (LOS) and hospitalization costs for coronary intervention, while similar studies for renal artery stenosis with stent placement are lacking. Methods: Using information from the HCUP's Nationwide In-patient Sample (NIS), we examined the trends in hospitalization stay and cost of care associated with renal artery stenosis stent placement between 2000 -2011. Designated weight specified analysis was performed to minimize bias. Only patients >18 years were included. The primary outcome was in-hospital all-cause mortality, and the secondary outcome was a composite of in-hospital mortality and peri-procedural complications. Hierarchical multilevel regression models were generated to determine independent predictors of LOS and hospitalization costs. LOS was calculated only in patients who were alive. Cost to charge ratio files were merged with NIS to calculate cost of care. The cost of care was adjusted for inflation with reference to 2011.

RESULTS: Total 18,355 procedures were analyzed which were representative of 89,607 procedures in US (42.5% females and 62.7% whites). In-hospital mortality rates and peri-procedural complications decreased significantly with increasing hospital volume. In-hospital mortality decreased significantly with higher hospital volume. [2nd tertile (0.2%); 3rd tertile (0.1%); when compared to 1st tertile(0.4%) p-value=0.001]. Secondary outcomes showed the similar results. Subgroup analysis in high-risk population (Charlson-Deyo Comorbidity index ≥ 2 and age > 80) also showed improvement in outcomes with increased hospital volume. The average LOS 2.13 ± 0.03 days and cost of care $11,893 ± 63. As hospital volume increases hospital LOS and cost of care decreases. Multivariate analysis showed similar findings of increased hospital volume, associated with decreased LOS in days (Adjusted OR, 95% CI, p-value) (3rd tertile: -0.74, -0.94 - -0.55, p = 0.001; 2nd tertile: -0.56, -0.71 - -0.40, p = 0.001 when compared to 1st tertile) and cost of care (3rd tertile: -$935, -$1468--$402, p = 0.004; 2nd tertile: -$803, -$1,215 -- -$391, p = 0.01 when compared to 1st tertile). Emergent/urgent admissions were associated with increase in LOS (p < 0.001) (1.49 days, 1.36 – 1.62) and hospitalization cost (p < 0.001) ($2037, $1735 – $2338) and Charlson comorbidity score >2 was associated with an increase in the cost hospitalization (p value <0.001) ($1829 , 1539 – 2119).

CONCLUSION: Patients undergoing renal artery stent placement procedures can significantly reduce their risk of in-hospital death and complications, along with disposition outcomes by selecting a high-volume hospital. High-volume hospitals had significant reduction in hospital length of stay and hospitalization cost.

| 279 | 241 | Research Veedu, Janeesh | Saint Peter’s University Hospital (Mayan Kothari) | OPIOIDS- CARCINOGENESIS AND RECURRENCES OF CANCER

INTRODUCTION: Opioids are widely used to alleviate malignant and non-malignant pain. Recently there have been several reports advocating that these drugs may alter cancer recurrence or metastasis. It is thought to be due to the modulation of the immune response or cellular pathways that control the survival and migratory behavior of cancer cells. There have been numerous studies on the effects of opioids on tumor cell growth via direct (receptor stimulation) and indirect (immunosuppression and endocrine modulation) pathways. We intend to look at current evidence that question the role of opioids in analgesia. A literature search of PubMed of the terms “opioids and cancer progression,” “anesthesia and cancer outcomes,” “Carcinogenesis and recurrences of cancer,” was used for our review.

DISCUSSION: Opioids promote angiogenesis, cancer cell proliferation and metastasis. Four different opioid receptors (mu, delta, kappa, and nociceptin) are expressed in the body with endogenous and exogenous opioids binding in the central nervous system
(CNS) modulating downstream effectors and the perception of pain. μ-opioid receptor (MOR) is overexpressed in several human malignancies. One study of 34 lung cancer patients showed an association between the expression of MOR and disease progression. Also opioids interfere with almost all hormonal axes but little evidence exists on effect of opioids on endocrine system in cancer patients. Retrospective studies suggest that regional anesthesia reduces the risk of tumor metastasis and recurrence. Analgesics affect the cellular milieu of malignant and nonmalignant cells and may influence cancer outcomes by directly stimulating tumor growth and inhibiting immune surveillance. It has been shown that open prostatectomy surgery with general anesthesia, substituting epidural analgesia for postoperative opioids, was associated with substantially less risk of biochemical cancer recurrence. Peri-operative techniques and analgesics may stimulate cancer cells and suppress host anticancer immunity. If the observation that opioid agonists promote metastases and recurrence of cancers, it will be worthwhile to examine the therapeutic potential of peripheral opioid antagonists against malignancy.

CONCLUSION: Current evidence does not support the hypothesis that opioids are associated with metastasis or recurrence of cancers. Prospective randomized controlled trials are currently underway which may or may not prove the hypothesis. Opioids will stay the mainstay of treatment of cancer pain until better treatment options or convincing data on the harmful effects are established. Our review is of importance in view of the increasing usage of opioids for pain, rising incidence of cancers and the rising incidence of prescription narcotic addictions in the country. The use of opioids must be judicious and must always be within the current guidelines.


HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS IN A PATIENT WITH GOODPASTURE’S SYNDROME: A RARE CLINICAL ASSOCIATION

INTRODUCTION: Hemophagocytic lymphohistiocytosis (HLH) is a life threatening clinical syndrome. HLH can occur in the setting of an autoimmune disease, chronic immunosuppression, malignancy and infection. Patients may succumb to death from sepsis and multiorgan failure. Goodpasture’s syndrome is a rare autoantibody-mediated disorder that typically presents with rapidly progressive glomerulonephritis (RPGN) and pulmonary hemorrhage. This is mediated by an anti-glomerular basement membrane (anti-GBM) antibody. We discuss a case of a young lady who was diagnosed with Goodpasture’s syndrome that was complicated by HLH.

CASE REPORT: A 31 year old female who was diagnosed with Goodpasture’s syndrome presents on this admission with febrile neutropenia. She had originally presented with frank hemoptysis and acute renal failure seven years ago. She eventually progressed to end stage renal disease, now dialysis dependent. She was receiving treatment with hydroxychloroquine and steroids. Over the last seven years, she had presented with polyarthritis involving multiple small and large joints. She was found to have antibodies to cyclic citrullinated peptide (Anti-CCP), rheumatoid factor (RF) and myeloperoxidase anti-neutrophil cytoplasmic antibody (MPO-ANCA). Due to multiple complications from hydroxychloroquine, she was placed on azathioprine and prednisone. On this admission, her clinical status deteriorated from persistent pancytopenia despite the use of broad spectrum antibiotics for underlying sepsis. Her pancytopenia was initially attributed to the immunosuppressive effect of hydroxychloroquine and azathioprine. A bone marrow biopsy was initially performed and revealed a hypercellular bone marrow. Due to prolonged pancytopenia of autoimmune etiology,
she was initiated on high dose steroids. Further work up was performed which showed an elevated ferritin. With the persistent febrile episodes, hepatosplenomegaly found on examination and pancytopenia, HLH syndrome was suspected. A repeat bone marrow biopsy was performed and confirmed this diagnosis. Hemophagocytosis was demonstrated by the presence of histiocytes engulfing erythroid cells. CD68, a marker of for histiocytes, was also found to be positive. Patient eventually expired despite aggressive treatment with high dose solumedrol and intravenous antibiotics.

**DISCUSSION:** Five of eight of these criterias must be fulfilled to make a diagnosis of HLH: Fever, splenomegaly, cytopenias, hypertriglyceridemia, hypofibrinogenemia, hyperferritinemia, hemophagocytosis in bone marrow, spleen or lymph nodes and low/absent NK cell activity. Treatment involves suppression of the inflammatory response in order to prevent impending organ failure and death. With the reporting of this case, we hope to emphasize the known association of autoimmune conditions with HLH. Here we consider one possible cause of secondary HLH to be that of Goodpasture’s syndrome, a disease entity with a well known autoimmune pathophysiology. Early recognition of this cause as a possible inciting factor to this disease state may lead to decreased morbidity and mortality in future cases through early treatment interventions.

**Clinical Vignette**

**281.16 Clinical Vignette**

**Basnet, Alina**

**Trinitas Medical Center (William Farrer)**

**PARATHYROID HORMONE RELATED PEPTIDE MEDIATED HYPERCALCEMIA IN CLASSICAL HODGKIN’S LYMPHOMA: AN UNCOMMON PRESENTATION**

**BACKGROUND:** The incidence of hypercalcemia is estimated to be one to five percent in classical Hodgkin’s lymphoma (HL). Although uncommon, it has been ascribed to be mediated through various factors; osteoclast activating factors (OAFs), overproduction of 1, 25 (OH) D3 (calcitriol), and infrequently prostaglandin synthesis. HL associated parathyroid hormone related peptide (PTH-rP) related hypercalcemia is very uncommon. We report a case of HL in a patient who presented with hypercalcemia and was found to have elevated PTH-rP and elevated calcitriol.

**CASE REPORT:** We are presenting an 81-year-old female, lifelong non smoker, with past medical history significant for recurrent renal calculi, hypertension and history of melanoma surgically resected thirty years ago, presented to us with complaints of inability to ambulate and altered mental status. Family members did report some weight loss and progressive worsening weakness over last few months. She denied history of fever, chills, night sweats, cough and abdominal pain. Physical examination was essentially benign except for palpable left axillary and left supraclavicular lymph nodes. Laboratory studies revealed severe hypercalcemia of fifteen mg/dl with phosphorus level of three mg/dl on admission. Work up for hypercalcemia, revealed abnormally elevated parathyroid hormone related peptide (PTHrP), normal serum protein electrophoresis and appropriately suppressed intact parathyroid hormone. Twenty four hour calcium excretion was also elevated. Imaging studies including bone scan revealed increased activity in the thoraco lumbar vertebrae and pelvic bones. Computerized tomography (CT) scan of the pelvis showed subtle area of cortical scalloping within the left iliac wing that could represent underlying lesion. CT of the chest and abdomen demonstrated abnormal very bulky left axillary lymphadenopathy with extension into left subpectoral space. Enlarged retroperitoneal lymph nodes and shotty pelvic and inguinal lymph nodes were demonstrated. A bone marrow biopsy was done which did not show any abnormality in the marrow. A left supraclavicular lymph node was biopsied which demonstrated classical HL on histopathological examination. She was then started on twelve weeks cycle of chemotherapy.

**DISCUSSION:** Of the various etiologies of hypercalcemia described in patient with HL, it has been rarely associated to be mediated
through increased PTH-rP. There is a well established evidence that 1,25-dihydroxycholecalciferol is the factor produced by the tumor and result in hypercalcemia in lymphoid tissue. PTH-rP and calcitriol level should respond to the treatment for the disease with chemotherapy. Literature review revealed one other case where HD with hypercalcemia was secondary to PTH-rP and that was proven by immunohistochemistry of liver biopsy.

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<th>Clinical Vignette</th>
<th>Basnet, Alina</th>
<th>Gerardo Capo</th>
<th>Trinitas Medical Center (William Farrer)</th>
<th>Malignant triton tumors in sisters with clinical Neurofibromatosis type- 1.</th>
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<td>INTRODUCTION: Malignant triton tumors (MTTs) are uncommon sarcomas, a subgroup of malignant peripheral nerve sheath tumors (MPNSTs). They arise from Schwann cells of peripheral nerve or existing neurofibromas and have elements of rhabdomyoblastic differentiation. Clinically, they have been associated with neurofibromatosis type 1 (NF-1). MTTs are associated with a high incidence of local recurrence and distant metastases and there is no treatment consensus yet exists.</td>
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<td>CASE REPORT: We report two sisters, both of whom were diagnosed with MTTs. Firstly, a 36 yr. old female presented with left sided chest wall swelling. On examination, she was found to have auxiliary freckling, café au lait spots and mild cognitive impairment. Her examination findings were consistent with clinical neurofibromatosis-1(NF-1). On imaging, a 10-cm soft tissue mass in the left lower anterolateral chest wall was seen. Debulking of the mass was done and histopathological examination showed high grade malignant peripheral nerve sheath tumor with skeletal muscle differentiation (Triton Tumor). The patient was treated with Ifosfamide and Adriamycin along with radiation. She still has no evidence of disease recurrence 4 years later. Her sister subsequently presented to us at age 42 yr. with left sided lateral chest wall pain. On examination, left flank fullness was palpated; café-au-lait spots were noted on her as well along with mild cognitive impairment. Imaging showed a multi-compartmental retroperitoneal cystic mass with left psoas involvement, displacement of the left kidney and loco regional lymphadenopathy. The tumor was resected and, similarly to her sister, it showed high grade malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation (MTT). The plan for her is to start on chemotherapy and radiation as described above.</td>
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<td>DISCUSSION: The incidence of MPNSTs is one per 100,000 cases. MTTs account for &lt;10% of MPNSTs. MTTs are a subgroup of MPNSTs that follow a particularly aggressive course. Around 50% of MPNSTs cases arise in patients with neurofibromatosis type 1 (NF1). NF1 association with MTTs is a negative prognostic factor. MTTs occur predominantly in the head, neck and trunk regions. The 5-year survival rate is 5–15% for MTTs, compared to 50–60% for MPNSTs. Radical surgical excision is the only curative intervention known so far. Adjuvant radiotherapy combined with chemotherapy has been used. No consensus for treatment yet exists. We want to report these two siblings presenting with this rare aggressive tumors in the setting clinical neurofibromatosis type 1. We hope to emphasize further the association of NF-1 with MTTs and that the first sibling is disease free four years after diagnosis.</td>
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<th>Clinical Vignette</th>
<th>Dsouza, Ria</th>
<th>Nahla Osman, Clark Sherer</th>
<th>Trinitas Medical</th>
<th>DRESS syndrome associated with Carbamezepine</th>
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DRESS syndrome is a rare, life-threatening, drug-induced immune response with frequent association with reactivation of a latent human herpesvirus infection. Clinical presentation usually involves skin eruption, hematologic abnormalities involving eosinophilia and atypical lymphocytes, lymphadenopathy, and internal organ involvement including liver, kidney, and lungs. Latency period of two to eight weeks between drug exposure and disease onset is described in most of the cases. We report a case of DRESS syndrome secondary to Carbamazepine as offending drug, with severe acute hepatitis as a systemic manifestation, and also human herpes viral reactivation documented. Complete recovery noted after withdrawal of offending drug and treatment with systemic corticosteroids. A 29 year old African American female with past medical history significant for bipolar disorder for which she was started on Carbamazepine a month prior to admission. Patient presented to our hospital with one week of fever, diffuse maculopapular rash, facial edema, cervical lymphadenopathy, and watery diarrhea. No drug allergies were reported on admission. Laboratory studies revealed elevated serum transaminases, positive serological testing for HHV6 and HHV7, atypical lymphocytes on peripheral smear, and marked eosinophilia on bone marrow biopsy. Early diagnosis of DRESS syndrome, withdrawal of Carbamazepine, and prompt treatment with steroids resulted in full recovery within a month. DRESS syndrome has a similar presentation to various other systemic disorders. Hence, early diagnosis, withdrawal of offending drug and management with corticosteroids can be life saving in cases with multiple organ involvement. ER physicians, Internists, and Allergists should have a low threshold for suspecting this rare clinical syndrome when a patient presents with skin rash and unusual systemic symptoms, and a new drug has been initiated recently. Prompt diagnosis and management are crucial to prevent fulminant liver failure associated with a mortality rate of 10% in DRESS syndrome.

A Susceptible Bowel: The Natural Course after Disseminated Histoplasmosis

INTRODUCTION: Histoplasmosis is the most prevalent endemic mycosis in the United States entering the human body by inhaling the microconidia, with gastrointestinal (GI) tract involvement reported in about 70-80% of cases. Plenty has been said in regards to isolated episodes of progressive disseminated histoplasmosis (PDH) affecting the intestine, yet to be determined is the natural course of a disrupted bowel rendered susceptible to either a possible structural intestinal complication, or a superimposed infection.

CASE DESCRIPTION: A 49 year-old heterosexual male from Honduras with AIDS (CD4 count of 87 and viral load of >10 E6 copies/ml off HAART) presented with fever, abdominal pain, and bloody mucoid diarrhea. He appeared toxic, wasted, but hemodynamically stable. There was no rash, peripheral lymphadenopathy, and watery diarrhea. No drug allergies were reported on admission. Laboratory studies revealed elevated serum transaminases, positive serological testing for HHV6 and HHV7, atypical lymphocytes on peripheral smear, and marked eosinophilia on bone marrow biopsy. Early diagnosis of DRESS syndrome, withdrawal of Carbamazepine, and prompt treatment with steroids resulted in full recovery within a month. DRESS syndrome has a similar presentation to various other systemic disorders. Hence, early diagnosis, withdrawal of offending drug and management with corticosteroids can be life saving in cases with multiple organ involvement. ER physicians, Internists, and Allergists should have a low threshold for suspecting this rare clinical syndrome when a patient presents with skin rash and unusual systemic symptoms, and a new drug has been initiated recently. Prompt diagnosis and management are crucial to prevent fulminant liver failure associated with a mortality rate of 10% in DRESS syndrome.
The risk of relapse or complications are significantly reduced by maintenance therapy after 12 to 24 months. Nevertheless, PDH sometimes takes an unexpected turn when co-infections such as parasitic, viral or bacterial agents are present leading to an increased Histoplasma pathogenicity. As seen in our case, C difficile could have potentially precipitated a fulminant presentation based on markers of intestinal inflammation and the emergence of strains such as the virulent C difficile NAP1/027. Should this occur, be alert to manage differently as it could affect the long-term outcome.

Clinical Vignette Peralta, Paloma
KUNAL PATEL MD, JOAQUIM J CORREIA, FAYEZ SHAMOON MD, Trinitas Medical Center (William Farrer)
DTAP-VACCINE INDUCED MYOPERICARDITIS MIMICKING ST ELEVATION MYOCARDIAL INFARCTION

INTRODUCTION: Vaccine related myocarditis is one rare cause of myocarditis. Smallpox vaccination-induced myocarditis has been well-reported in literature however only two cases of myocarditis related to DTaP vaccination have been reported. Both of these cases were documented in the pediatric population.

CASE DESCRIPTION: We present a 37 year old Portuguese male with past medical history only significant for drug abuse that came to the emergency room with complaints of pressure like chest pains, pleuritic in nature associated with fevers and chills for five days. The patient had a DTAP vaccine placed five days prior to the admission after which his symptoms began. Upon arrival to the emergency room an electrocardiogram was performed which showed ST elevations in leads II, III, AVF, v4 and v5. An emergency echocardiogram done at bedside showed an ejection fraction 40% with moderate global hypokinesis but no evidence of pericardial fluid. Immediate cardiac catheterization revealed normal coronary arteries. First Troponin I level was elevated at 30. The patient was transferred to CCU with a presumed differential diagnosis of Myocarditis vs Coronary Vasospasm. Rheumatologic screening and serum viral antibody titers for suspected acute infectious causes were all negative. This included Coxsackie virus group B, Human immunodeficiency virus (HIV), Cytomegalovirus, Ebstein-Barr virus, Hepatitis virus family, and Influenza viruses. In addition, urine and hair samples were sent for drug screen; they were negative for recent cocaine or amphetamine use. Cardiac MRI with and without gadolinium was performed for definitive diagnosis and showed delayed myocardial enhancement involving the myocardium of the mid-inferior and mid-inferolateral wall of the left ventricle. These findings were consistent with edema, inflammation, and myocarditis. Troponin I values were trended to monitor extent of myocardial damage and serial values reached 30, 46.4, 17.8, 0.19, respectively. The patient was treated with Colchicine and NSAIDs and his symptoms improved significantly over the following 3 days.

DISCUSSION: Myocarditis has multiple etiologies however vaccine related causes are rare. In a thorough review of literature, we found only two cases of myocarditis induced by tetanus vaccine. Both were reported in the juvenile population: one in a 3 month old after a DTAP vaccine and the other in a 13 year old male after tetanus vaccination. Our case may be the first one seen in an adult. DTAP induced myocarditis should be suspected in patients with chest pains and fevers with an antecedent of the vaccine and promptly evaluated as it can cause detrimental repercussions.

LEUCONOSTOC INDUCED ENDOCARDITIS IN A PATIENT WITH NO RISK FACTORS

INTRODUCTION: Infective endocarditis is an infection of the endocardial surface of the heart, which may include heart valves, mural endocardium, or a septal defect. Its complications include severe valvular insufficiency, intractable congestive heart failure and myocardial abscesses. Leuconostoc species are members of the Streptococcaceae family, which were thought to be non-pathogenic but only recently were recognized as potential pathogens after few cases in the literature reported as a cause of endocarditis. We
are presenting a case of a 65-year-old male with no risk factors that developed Leuconostoc acute endocarditis with severe aortic valve insufficiency.

**CASE PRESENTATION:** This is a 65 year old male originally from Trinidad with past medical history of chronic obstructive pulmonary disease and atherosclerotic peripheral vascular disease, who was admitted to the hospital for evaluation after a syncopal episode at home with prodrome of abdominal pain, gas sensation and intermittent fevers for 2 months prior to admission. Upon evaluation in the emergency room, electrocardiography showed ST segment depressions in lateral leads and T wave inversions with initial troponins of 0.5. Trans-thoracic echocardiography revealed a calcified mobile echo density attached to the ventricular surface of the aortic valve consistent with vegetations which was corroborated with a trans esophageal echocardiography that revealed two vegetations on the right and left leaflet respectively with severe aortic insufficiency and a pressure gradient of 125 milliseconds.

Three different blood cultures grew Leuconostoc SP resistant to vancomycin for which the treatment was switched from the empiric ceftriaxone and vancomycin to intravenous ampicillin. Para nasal computed tomography (CT) failed to show any evidence of dental infection, abscess or other abnormality. His course was complicated with ventilator dependent respiratory failure secondary to acute congestive heart failure with further extubation within 36 hours and clinically improvement of patients' condition after which was transferred to a rehabilitation center for completion of antibiotics. Cardiac spect scan showed a reversible defect in distal anterior wall and cardiac catheterization was planned in the future. After completion of antibiotics, he was transferred to another hospital for coronary artery bypass graft (CABG) and valve replacement. Unfortunately, the patient died with post-surgical complications.

**DISCUSSION:** Leuconostoc species are gram-positive coccobacilli, which usually demonstrate vancomycin resistance. Had always been thought to not clearly play a role in human infections but associated with gastrointestinal symptomatology. Only three cases have being reported in the literature of leuconostoc acute endocarditis, but to our knowledge this is the first case reported in a patient with no previous valve disease and no risk factors for endocarditis. This will be an important entity to consider as new cases arise, especially because of its detrimental effects, inherent vancomycin resistance and consideration of different antibiotic coverage.
with sinus tachycardia. CT of the abdomen with IV contrast showed hepatosplenomegaly and wedge-shaped splenic infarct. Further evaluation included positive mono-spot test, elevated antiphospholipid IgM >100 [<25U/mL] and anti-TPO Antibodies >1000 [<35 IU/mL], low Protein C 37 [70-140%] and a normal transthoracic echocardiogram. Neck sonogram displayed heterogeneous hypervascular thyroid gland. She was discharged home. Further work up confirmed the diagnosis of Garve’s Disease.

**DISCUSSION:** Constellations of facts are quite unique in this case. We describe splenic infarction in a patient with acute IM due to EBV, transient elevation of antiphospholipid antibodies, protein C deficiency and hyperthyroidism with positive anti-TPO antibodies. Splenic infarction represents a rare complication of IM due to EBV infection. In one case of IM, the patient had underlying hereditary spherocytosis and protein C deficiency and another case described transient induction of antiphospholipid antibodies associated with acute EBV infection and splenic infarction. One study reports three cases of Grave’s disease associated with IM due to EBV infection while EBV is associated with an increased risk of autoimmune diseases such as SLE, RA and MS. Our case reinforces the literature which associates acute IM due to EBV infection with splenic infarction but proposes that EBV -as a source of chronic immune stimulation- may initiate a destructive autoimmune process in the thyroid gland.