IS IT THROMBOTIC THROMBOCYTOPENIA (TTP) OR B12 DEFICIENCY?
Alex Wu, MD; Kevin Chen, MD; Charmian Sittambalam, MD
Medstar Franklin Square Medical Center, Baltimore, MD

There are an increasing number of recent publications recognizing that TTP may mask B12 deficiency, due to similar features such as hemolytic anemia, thrombocytopenia, and schistocytosis. It has been hypothesized that hemolytic anemia may be more prominent in individuals with co-existing methylenetetrahydrofolate reductase (MTHFR) gene mutation and vitamin B12 deficiency associated with elevated homocysteine levels.

A 51-year-old female with TTP and known alcohol dependence, was admitted for pancytopenia with associated lethargy. She had been diagnosed with TTP about 1 year ago based on lab work that showed WBC 2.8K, Hgb 2.6g/dL, platelet count 41,000, LDH 2731U/dL, and ADAMTS13 30%; bone marrow biopsy showed only hypercellularity. Vitals were unremarkable on admission, with no focal deficits on physical exam. After noting LDH > 4,000 U/dL, she was initially treated with steroids and plasma exchange for 5 days for a presumed flare of TTP. Despite treatment, there was minimal clinical improvement. A new ADAMTS13 was found to be 78%, which further confirmed that TTP was less likely. She also had mild DIC with elevated D-dimer to 15.09 μg/mL, FSP between 10 and 40 μg/mL, PT 16.4s, INR 1.3, but normal values of fibrinogen and PTT 35.9s, without signs of active bleeding. Additional work up excluded hepatitis, HIV, SLE, scleroderma, Sjogren and G6PD. Peripheral smear revealed schistocytes, tear cells, and ovalocytes. Other labs included positive anti-parietal antibodies, Hgb 6.6 g/dL, Hct 20%, RDW 34.3%, haptoglobin levels < 8 mg/dL, serum B12 level 142 pg/mL, folate 11.4 ng/mL, methylmalonic acid 1.19 μmol/L, homocysteine 17 μmol/L, and MCV of 89.3. Reticulocyte count was unavailable. Genetic testing also revealed MTHFR A1298C heterozygosity. Based on these results, a diagnosis of pernicious anemia was made. She was discharged to follow up with her hematologist and receive B12 supplementation for life.

This case supports that heterozygous MTHFR gene mutations may play a role in diagnosing B12 deficiency/pernicious anemia, especially when signs and symptoms can be equivocal for a more serious disease like TTP. Reaching the correct diagnosis will aid with better treatment decisions and outcomes.

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Indicate your participation in research process (4 sentences or less):

We were part of the team that collected information about the patient, researched on the topics and wrote up the case for this clinical vignette.

First Author Information:

Name: Alex Wu, MD

Institution: MedStar Health Internal Medicine, Baltimore

Daytime Phone: 410-554-2284

Co-Author(s) Associates: Kevin Chen, MD; Charmian Sittambalam, MD

Program Director’s Name: Stephanie Detterline, MD

(indicating review of abstract)
Ticks and Salt: An Atypical Case of Neuroborreliosis
Nazia Siddiqui*, Deidre St. Peter MD**, Surendra Marur MD**
* Saba University School of Medicine, Saba, Dutch Carribean
** The Greater Baltimore Medical Center, Baltimore, MD

The diagnosis of SIADH can easily be made based on laboratory criteria, but the management of the syndrome is solely dependent on the etiology. It is well documented that central nervous system (CNS) infections may lead to syndrome of inappropriate antidiuretic hormone secretion (SIADH), but diagnosing these can prove difficult in patients with atypical presentations. We present a case of SIADH and muscle weakness in a patient without typical signs and symptoms of CNS infection who was tested and diagnosed with Neuroborreliosis based largely on her likelihood of exposure. This case indicates the need for Lyme testing in patients with unexplained SIADH who live in endemic areas.

The patient was an 83-year-old female from rural Maryland with a history of type 2 diabetes and hypertension, who presented from her primary care physician’s office when her sodium was found to be 123 mEq/L. Her sole symptom was proximal muscle weakness of the arms. The diagnosis of SIADH was reached based on laboratory data. A trial of fluid restriction was initiated, but neither her sodium nor her muscle weakness improved. Lyme testing was performed as the patient lived in an endemic area, returning positive, and lumbar puncture showed evidence of neurologic involvement. After realizing the appropriate treatment for hyponatremia in this case, intravenous ceftriaxone was started, and patient’s sodium levels improved and muscle weakness resolved.

Studies show that SIADH is associated with CNS infections, likely related to the inflammatory cascade. However, the atypical presentation of Neuroborreliosis for our patient delayed the appropriate diagnosis and treatment. Our case demonstrates the need to screen for Lyme disease in endemic areas in patients presenting with neurologic symptoms and SIADH.
IS IT REALLY ANGIOEDEMA?
Lulit Dessie, MD; Saad Ali, MD; Eskandar Yazaji, MD
MedStar Union Memorial Hospital, Baltimore, Maryland

Introduction: Lymphedema is one of the complications of cancer and its associated treatments including radiation and surgery. Lymphedema can involve any part of the body but its occurrence on the face makes management much more difficult and create a significant psychological burden on patients.

Case Description: A 65-year old African American man started to have facial swelling in 2012 presumed to have been caused by medication allergy and angioedema. The patient was repeatedly admitted for similar complaints and was taking steroids and four different antihistamines. The swelling continued to worsen over time, leading to significant breathing difficulty and progressive facial swelling. The patient was admitted in January 2017 for worsening of facial edema. The medical team reviewed his prior studies and the literature concerning lymphedema in head and neck cancer patients. They also learned from his wife that he has been receiving lymphedema treatment in the past. The team decided to discontinue the antihistamines with no symptom worsening. The patient was referred for lymphedema therapy (physical therapy) with great improvement in swelling.

Conclusion: This case highlights the unique presentation of facial lymphedema in head and neck cancer survivors. It reaffirms the importance of careful documentation of patient’s history. It also underscores the considerable diagnostic challenge facial lymphedema poses and the importance of ruling out other differentials such as cancer recurrence, angioedema and allergic reactions to be able to provide proper treatment.

First Author Information:

Name: Lulit Dessie, MD
Institution: MedStar Health Internal Medicine, Baltimore
Daytime Phone: 410-554-2284

Co-Author(s) Associates: Saad Ali, MD; Eskandar Yazaji, MD

Program Director’s Name:
Stephanie Detterline, MD
(indicating review of abstract)
Myocarditis Associated with Antisynthetase Syndrome

Antisynthetase syndrome is a distinct clinical entity of myositis and the incidence ranges from 1.2 to 2.5 per million. Cardiac involvement in inflammatory myopathies has a varied presentation, however myocarditis seems to be particularly rare in antisynthetase syndromes, with only a few case reports published to date.

The patient is a 64 year old woman with a medical history of coronary artery disease (mid LAD stent 2009), HTN, prior left renal stent (2009), pulmonary fibrosis on 3L home O2 who presented to the Bayview ED on 3/29/17 with SOB and lower extremity swelling.

Of note, she had a significant history of multiple symptoms dating back to 2000. She had morning stiffness, and joint pain thought to be psoriasis treated with IM steroid injections for 12 years. She was diagnosed with pulmonary fibrosis in 2016. In the fall of 2016 she developed weakness, had trouble climbing stairs and also developed Raynaud’s Syndrome. In February of 2017 she was seen by a pulmonologist and work-up for ILD demonstrated anti-Jo1, creatinine kinase of 972 U/L, and aldolase of 18.5 U/L. PFT’s showed a restrictive pattern and she started taking azathioprine 50mg daily. In March she developed worsening shortness of breath, lower extremity edema, and RUQ pain at home and came to the BV ED for further evaluation.

During her Bayview admission, she had troponin elevation to a peak of 0.10 and a proBNP of 16,000 pg/ml (increased from 2000 pg/ml last month). TTE showed a newly depressed EF of 10-15%. She underwent RHC/LHC which showed a diffuse 70-90% mid RCA stenosis but there were no antecedent objective findings suggestive of ischemia in this distribution and this did not explain the diffuse cardiomyopathy so not stent was placed. She was seen by Rheumatology for her anti-Jo1 antisynthetase syndrome, who recommended urgent cardiac MR and endomyocardial biopsy to assess for cardiac myositis. The cardiac MRI demonstrated T2 hyperintensity and delayed enhancement in a nonvascular distribution suggestive of active myositis. Cardiac biopsy also supported the cardiac myositis diagnosis. For her antisynthetase induced myocarditis she was treated with IV methylprednisolone 1 gram daily for 3 days followed by prednisone 1 mg/kg. She was also started on mycophenolate mofetil titrating up to 1000mg twice daily.

Diagnosing myocarditis remains a challenge in patients with an autoimmune disease, particularly with inflammatory myopathies. This is because the clinical manifestations can be subclinical, nonspecific, or concealed by other clinical features, including pericarditis or lung involvement. It is, however, crucial to diagnose myocarditis, due to the need for specific treatments, and to avoid a potentially fatal early evolution or the development of chronic heart failure at a later stage.
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TAKOTSUBO CARDIOMYOPATHY MIMICKING ACUTE CORONARY SYNDROME IN SEPSIS. Byrne A, MD. Racherla, M, MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Takotsubo cardiomyopathy (TCM) is characterized by transient systolic and diastolic dysfunction precipitated by a stressor. In the acute phase, TCM can closely resemble acute coronary artery syndrome (ACS), often presenting with chest pain, dyspnea on exertion, ST segment changes on EKG and elevated cardiac biomarkers.

A 65 year-old woman with history of hypertension and short gut syndrome on chronic TPN via PICC line presented with chest pain, nausea and vomiting. Initial vital signs were notable for heart rate of 103 bpm, blood pressure of 89/53 mmHg, 34 respirations per minute with oxygen saturation of 91% on ambient air. Examination was remarkable for tachypnea. EKG revealed T-wave inversions in the precordial leads. The first troponin was 0.71 ng/mL (normal ≤ 0.06 ng/mL).

Echocardiogram revealed a mildly hypertrophic left ventricle (LV) with moderately decreased systolic function, LV ejection fraction 35% with diffuse distal and apical akinesis. An urgent left heart catheterization was performed, which revealed no angiographic evidence of coronary artery disease but severely elevated LV end-diastolic pressure of 40 mmHg (normal 4-12 mmHg). The patient was admitted, started on intravenous diuretics and placed on heart failure medications for TCM. Within 24 hours, blood cultures returned positive for coagulase negative staphylococci. She was started on antibiotics and the PICC line was exchanged. Repeat echocardiogram in 3 weeks revealed mostly normalized LV function with mild residual apical hypokinesis.

Despite a completely different pathophysiology, TCM is indistinguishable from an acute left anterior descending (LAD) territory myocardial infarction (MI). Furthermore, dysfunction in TCM is reversible whereas it is not in an acute MI. Thus, it is imperative to evaluate all patients with new apical MI with cardiac catheterization in order to definitively diagnose TCM.

First Author Information:

Name: Alexandra Byrne, MD

Institution: University of Maryland Medical Center and Baltimore VA Medical Center

Daytime Phone: 410-328-7567

Co-Author(s) Associates: Manoj Racherla, MD

Program Director’s Name: Susan D. Wolfsthal

(indicating review of abstract)
DURABLE RESPONSE WITH IMMUNOTHERAPY IN TREATMENT OF REFRACTORY RARE NON-SMALL CELL LUNG CANCER. Patel, A MD, Patel A, MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

A 61-year-old man presented with a one year history of a dry cough. Chest x-ray revealed a lobulated left lower lobe (LLL) lung mass. Biopsy of the mass showed a primary intestinal adenocarcinoma with stains initially consistent with metastatic colorectal adenocarcinoma (CRC). Given the immunophenotype, workup for gastrointestinal malignancy was recommended with upper and lower endoscopy, both of which were entirely unremarkable. Positron emission tomography (PET) scan imaging showed no evidence of extrathoracic disease. Mediastinoscopy and subsequent LLL lobectomy revealed a 13 centimeter adenocarcinoma with negative margins and negative lymph nodes. Immunostains showed +CDX2, +CK20, negative TTF1 and napsin A. This phenotypic profile confirmed pulmonary enteric adenocarcinoma (PEA).

To reduce recurrence risk of his non-small cell lung cancer (NSCLC), he was treated with docetaxol and cisplatin for four cycles. Six months after the LLL lobectomy the patient had disease progression with development of a 10 centimeter left adrenal gland nodule and two nodules in the right middle and right lower lobes. Biopsy and resection of the left adrenal gland nodule confirmed an adenocarcinoma with the same phenotype found in the initial lobectomy specimen. The patient was then started on nivolumab with repeat computed tomography (CT) scan three months later demonstrating an appreciable reduction in the right lower lung nodule and stable right middle lobe and left adrenal disease. Twenty-two weeks (11 cycles) into his treatment, he again developed progression of his left adrenal disease requiring six fractions of radiation therapy over nine months. Growth of the adrenal mass slowed. The patient is planned for repeat adrenalectomy to assess new mutational burden. While on nivolumab the patient had minimal decline in functional status and minimal change in quality of life.

The patient’s marked response to nivolumab therapy suggests that PEA may respond more as a lung primary than CRC. Nivolumab is known to have efficacy in NSCLC with some cases of durable long term responses. This highlights the exciting possibility that immunotherapy may allow cancer patients to obtain a quality of life comparable to other chronic medical conditions.
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THERE IS NO SUCH THING AS A SURE THING...CUE AUTOIMMUNE PANCREATITIS
Michael Aughenbaugh, MD; Abhinav Sankineni, MD; Charmian Sittambalam, MD
Medstar Franklin Square Medical Center, Baltimore, MD

Autoimmune pancreatitis (AIP) is a rare, but increasingly recognized disease. Type 1 AIP accounts for 80% of cases in the United States and is associated with elevated serum IgG4 levels, causing chronic inflammation that can affect multiple organ systems. In contrast, type 2 AIP is not strongly associated with elevations in serum IgG4 and involves only the pancreas. Diagnosis of AIP can be made by clinical history, as well as histologic, serologic, and imaging findings.

A 78-year-old woman presented as an outpatient with painless jaundice and weight loss. Physical exam revealed significant jaundice of the skin and sclera. The patient was without cardiac arrhythmias or murmurs. Abdominal exam was benign, and there was no JVD or edema. Laboratory tests revealed a significant hyperbilirubinemia of 6.4, as well as moderately elevated transaminases and glucose of 447. A CT of the abdomen revealed intra- and extrahepatic ductal dilation with fullness in the pancreatic head region. An endoscopic retrograde cholangiopancreatography was performed, identifying a mass in the pancreatic head and an additional mass in the body of the pancreas. A stent was placed to relieve the biliary obstruction. A fine-needle aspirate was performed which was negative for malignant cells; however, immunostaining revealed fibrotic pancreatic parenchyma decorated with IgG4. Further laboratory tests revealed IgG4 levels of 242.2, nearly three times the upper limit of normal. A diagnosis of IgG4-mediated type 1 AIP was made. The patient was initiated on steroid therapy and after approximately two months, the patient’s jaundice resolved. Laboratory tests revealed normalization of her bilirubin, transaminases, IgG4 levels, and blood glucose levels. The biliary stent was removed and the patient was maintained on steroid therapy with outpatient follow-up.

This case illustrates the importance of distinguishing AIP from other disease entities, such as pancreatic cancer. The overall prognosis of AIP is excellent, in stark contrast to that of pancreatic cancer. AIP is responsive to steroid therapy; however, relapse is common and additional immunosuppressants or immunomodulators may be needed. Biliary stenting, aggressive glycemic monitoring, and pancreatic supplementation may be necessary.
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A “WEIL” OF A TIME
Sean Abeysekera, MD; Michelle Bahrain, DO; Charmian Sittambalam, MD
MedStar Franklin Square Medical Center, Baltimore, MD

Leptospirosis is an epidemic-prone zoonotic infectious disease that occurs worldwide with higher incidence in tropical climates. It is caused by bacteria of the genus Leptospira. Although the overall incidence in the United States is relatively low, leptospirosis is considered to be the most widespread zoonotic disease in the world.

A 55 year old Caucasian male initially presented with five days of fever, rigors, sore throat, myalgias, neck stiffness, nausea, vomiting, and poor oral intake. The patient was hemodynamically unstable requiring aggressive fluid resuscitation, vasopressors, and broad spectrum antibiotics, and was transferred to the ICU on arrival. The patient had some contact with a dog which died of leptospirosis one year prior and lived in a neighborhood with an ongoing rodent problem. Preliminary infectious work up included a viral panel, cultures, syphilis screen, HIV screen, hepatitis panel, and lyme titres, all of which were negative.

Typhoid fever, brucellosis, dengue, hepatitis A, Q fever, and rickettsial infection were largely disproven by negative blood cultures and incompatible history. Given the clinical presentation, sepsis, sepsis hepatitis, renal failure, and meningitis, a diagnosis of leptospirosis was made, and the patient was continued on ceftriaxone for a total antibiotic course of seven days.

This case met clinical criteria for icterohemorrhagic leptospirosis, also known as Weil’s Disease. Additional criteria included close contact with a pet that tested positive for leptospira infection and with rodents. While viral hemorrhagic fevers and autoimmune etiologies such as Kawasaki Disease cannot be completely ruled out, the clinical presentation which included rhabdomyolysis, renal failure, hepatitis, positive IgM serologies, and improvement on ceftriaxone were more suggestive of leptospirosis. Isolation of leptospires from human tissue or body fluids is the gold standard. Leptospirochetes grow slowly in culture, recovery rates are low, and the sensitivity of acute serologic tests is low. Given these facts, the importance of initiating empiric therapy in patients with strong clinical suspicion is crucial, given that diagnostic testing options can be of low yield, expensive, and time consuming.

First Author Information:
Name: Sean Abeysekera, MD
Institution: MedStar Health Internal Medicine, Baltimore
Daytime Phone: 410-554-2284

Co-Author(s) Associates: Michelle Bahrain, MD, Charmian Sittambalam, MD

Program Director’s Name: Stephanie Detterline, MD

(indicating review of abstract)
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TENOFOVIR INDUCED FANCONI SYNDROME AND PANCREATITIS

Brittany Duchene MD, Bibi Aneesah Jaumally MD, Francois Gregoire MD

Introduction: Fanconi syndrome is a cause of type 2 proximal renal tubular acidosis. It is a syndrome of inadequate reabsorption, characterized by the excessive loss of substances in the urine including amino acids, glucose, phosphate and bicarbonate in the absence of high serum levels. It can be caused by underlying congenital or acquired diseases, toxicity or by adverse drug reactions. Tenofovir has been a documented cause of both Fanconi syndrome as well as pancreatitis.

Case Presentation: 62 yo female with recent admission 2 weeks prior for decreased UOP, hypotension, diarrhea, nausea and vomiting found to have AKI with a creatinine of 11.8 (from a baseline of 1.1), acidosis (bicarb 12), hyponatremia (130) and hypokalemia (3.1). She improved with volume resuscitation with her creatinine to 2.59 before she left AMA. She returned to the hospital 10 days later with similar complaints of nausea, vomiting and abdominal pain for 3 days. She also endorsed having diarrhea 3 days prior to admission, but has since resolved after taking anti-diarrheal medicines. Her labs were notable on this admission for a hyperchloremic non-anion gap metabolic acidosis with markedly decreased potassium, phosphate and uric acid. Due to the abdominal pain a lipase was ordered and found to be 3515. CT abdomen with contrast was consistent with pancreatitis.

Discussion: Fanconi syndrome is either hereditary or acquired. Acquired causes include multiple myeloma, light-chain deposition disease, renal transplantation, and medications. Medications implicated in the development of Fanconi syndrome include ifosfamide, didanosine, acetazolamide, aminoglycosides and tenofovir. Tenofovir has been documented in the past to be the causative agent in inducing Fanconi syndrome worldwide. In all of these cases the patients had been boosted by lopinavir-ritonavir, a component that is absent from this presentation. Tenofovir is primarily excreted from the renal tubules via MDR-2 (multi drug resistance) protein. Ritonavir inhibits MDR-2, which is what leads to the accumulation of tenofovir in the tubules allowing them to damage the tubular cells. Our patient was also being treated with elvitegravir, which is a documented strong MDR reversing agent. It is likely the elvitegravir acted in a similar method to the other cases with ritonavir use.

Unfortunately, our patient had pancreatitis as well. Although not listed as a side effect, post market case reports have found multiple cases of pancreatitis with concomitant use of tenofovir and didanosine. It is unclear whether our patient presented with an idiopathic pancreatitis or if

First Author Information:
Name: Brittany Duchene, MD
Institution: Medstar Harbor Hospital
Daytime Phone: 667-217-9251
Co-Author(s) Associates: Bibi Aneesah Jaumally MD, Francois Gregoire MD

Program Director's Name: Dr. Richard Williams

(indicating review of abstract)
AN IRON-CLAD DIAGNOSIS OF RAPID-ONSET DIABETES

Iron overload (hemochromatosis) is a disorder of iron regulation characterized by an increase in predominantly duodenal iron absorption with subsequent tissue deposition in the liver, heart, pancreas, joints, and other endocrine organs. The following clinical vignette illustrates a case of iron overload presenting as new rapid-onset diabetes, profound insulin resistance and abnormal liver enzymes.

Ms. DG is a 52 y/o African American female with hypertension and morbid obesity who presented to the hospital because of progressive abdominal pain, nausea, and vomiting for 8 days with inability to tolerate oral intake. Review of systems revealed a 15 kg weight gain in the past year, recent menopause, depressive symptoms, and diffuse arthralgias. Physical exam was notable for tachycardia, normal S1/S2, and otherwise unremarkable. Initial labs included blood glucose of 1,048 mg/dL, bicarb of 22 mmol/L, anion gap of 24 mmol/L, AST 67 U/L, ALT 40 U/L, a VBG with pH of 7.33 and pCO2 of 45 mmHg, and a UA notable for small ketones. Hemoglobin A1C was 15.0% compared to 5.1% 12 months prior. She was treated with an insulin drip for hyperglycemic hyperosmolar state, requiring greater than 2 units/kg/day, and discharged on lantus 38 U BID and 40 U nutritional aspart.

The rapid development of diabetes mellitus (DM) with insulin resistance and abnormal LFTs prompted a more extensive workup with iron studies, including iron level of 193 ug/dL, ferritin of 545 ng/mL, percent saturation of 78%, transferrin 198 mg/dL, and TIBC of 248 ug/dL, which are consistent with a diagnosis of hemochromatosis. MRI liver confirmed increased iron deposition (30 mmol/kg dry tissue) as well as hepatic steatosis. She was discharged with a plan to follow-up for genetic testing and possible liver biopsy.

Iron overload can be caused by genetic mutations (hereditary hemochromatosis) or as a secondary process such as from frequent transfusions, non-alcoholic fatty liver disease (NAFLD), and ineffective erythropoiesis. Both can manifest with symptoms of multi-organ involvement. In women who lose iron via menstruation, symptom onset can be delayed until after menopause. An elevated transferrin saturation or serum ferritin warrants an evaluation of hereditary hemochromatosis, and in the case of abnormal LFTs, a liver biopsy should be considered. In Ms. DG who presented with morbid obesity and consistent MRI findings, the most likely etiology of her iron overload is secondary to NAFLD. With new diabetes and suggestive systemic symptoms, the differential should include iron overload leading to insulin resistance from pancreatic iron accumulation. In severe cases, patients may benefit from chelation therapy or phlebotomy to prevent further organ damage.
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WEIL OF MISFORTUNE: SEVERE LEPTOSPIROSIS IN AN ACTIVE DUTY PATIENT

Gray, Jeffery, MD1(Associate), Dore, Michael, MD2(Member)
1. Walter Reed National Military Medical Center, Bethesda, MD
2. U.S. Naval Hospital Guam, Tutuhan, Guam

Leptospirosis is a zoonotic infection caused by *Leptospira*, a spirochete that thrives in tropical climates. It can be transmitted via direct contact with body fluids from an infected host, or commonly from contaminated environmental sources such as fresh water.1 Leptospirosis tends to be an asymptomatic or mild disease in humans. However, in approximately 1% of cases, it can present with severe multi-organ involvement, which is commonly known as Weil’s Syndrome. Despite the potential for serious illness, if leptospirosis is diagnosed and treated in a timely manner, the prognosis is favorable even in severe cases.

A 28 year-old previously health active duty male stationed in Guam presented to the Emergency Department with one week of worsening fatigue and malaise. He had also noted worsening diarrhea, which had progressed from a few watery, non-bloody bowel movements per day to as many as eight bowel movements on the day of admission. His history was also notable for fevers to 102 degrees Fahrenheit at home, progressive right lower quadrant pain, and subjective yellowing of the eyes. His only notable exposure was visiting a waterfall in Guam one week prior to the onset of symptoms. His physical exam was notable for diffuse abdominal tenderness. There was no scleral icterus or conjunctival suffusion noted. Laboratory studies were notable for hyponatremia to 133 mmol/L, hypokalemia to 3.3 mmol/L, acute kidney injury with a creatinine of 7.60 mg/dL, and elevated liver enzymes. A CT scan of the abdomen and pelvis on admission was notable for small bilateral pleural effusions, mild enlargement of the bilateral kidneys, and minimal free pelvic fluid. Given concern for severe leptospirosis, the patient was started on ceftriaxone. Doxycycline was also added for rickettsial coverage. He rapidly recovered with fluid resuscitation and antibiotics. Leptospira IgM serology was positive, and the patient was discharged on oral doxycycline.

Leptospirosis should be considered in all patients with fresh water exposure and a consistent clinical syndrome. Weil’s syndrome is characterized by the classic triad of a bleeding diathesis, liver injury, and acute kidney injury with electrolyte derangements. This case illustrates the potential for severe multi-organ involvement from leptospirosis, even in previously healthy patients. It also demonstrates that early recognition and treatment of leptospirosis can lead to rapid improvement in symptoms and clinical status.
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Syncope as a Presenting Symptom of Obstructive Shock.

The pathophysiology of syncope is attributed to a transient loss of cerebral artery perfusion leading to loss of consciousness. A clinical framework for approaching the differential diagnosis is to consider vasodepressor versus cardiogenic causes. Syncope is a non-specific complaint that can be a harbinger of high-risk disease with poor outcomes. We present two patients who presented with syncope who were then found to have life threatening pulmonary emboli.

Case 1: A 54-year-old diabetic man presented to the Emergency Department (ED) with lightheadedness and syncope. Physical examination revealed hypotension to 80-90/50-60 despite receiving 5L normal saline, “borderline tachycardia” to 85-97, and hypoxia requiring oxygen supplementation. Laboratory data revealed blood glucose of 300, leukocytosis of 14.8, lactate of 5.5, and lateral lead ST depressions on EKG. Empiric treatment for sepsis was started. A CT head and CT Angiogram of the chest were obtained due altered mentation and progressive hypoxia. The latter revealed a saddle Pulmonary Embolism (PE). He was placed on extracorporeal membrane oxygenation, and discharged from the hospital 3 weeks later.

Case 2: A 79-year-old woman presented to the ED with vomiting and syncope while having lunch with co-workers. Review of systems was positive for leg swelling. Physical examination was significant for cool clammy skin, tachycardia, and hypotension. RBBB, T-wave inversion, and right heart strain were present on EKG. In the ED, she yelled out for help, lost consciousness, and went into cardiac arrest. She underwent cardiopulmonary resuscitation with successful resuscitation and intubation. Post resuscitation EKG revealed sinus tachycardia with resolution of RBBB. A post-cardiac arrest CTA confirmed a diagnosis of PE. Thrombus retrieval and intra-arterial thrombolysis were attempted. She expired 2 days later from intra-abdominal hemorrhage secondary to traumatic CPR and thrombolysis.

These cases illustrate the importance of considering pulmonary embolism in the differential diagnosis of patients with syncope. As with many cases of PE, initial presenting symptoms can be vague, and thorough histories are not always available. Evaluation for pulmonary embolism in cases of syncope, when appropriate, can be life-saving.

First Author Information:
Name: Suehyb Alkhatib, MD

Institution: University of Maryland Medical Center
Midtown Campus

Daytime Phone: 410-225-8790

Co-Author(s) Associates: Seyi Adesope, MD, Afua Ntem-Mensah, MD

Program Director’s Name: Robert Chow, MD
(indicating review of abstract)
TAKOTSUBO CARDIOMYOPATHY IN THE SETTING OF ACUTE RESPIRATORY FAILURE
Sujan Thapaliya, M.D.; Louai Naddaf, MS-III; Debajit Roy, M.D.; Tomas H. Ayala, M.D.

INTRODUCTION: Takotsubo Cardiomyopathy (TCM) was first described in the medical literature in early 1990’s, and in two decades it has gained worldwide recognition. With most recent retrospective analysis, about 2% suspected acute coronary syndrome is secondary to TCM. However, current knowledge on pathogenesis, management and outcome of TCM is largely unknown. Here we report, an elderly woman who initially presented with respiratory failure and later diagnosed with TCM.

CASE PRESENTATION: A 61 year old female with a past medical history of paroxysmal atrial fibrillation on anticoagulation, asthma, diabetes mellitus type 2, pulmonary hypertension, stroke, hypertension, and obstructive sleep apnea presented with severe shortness of breath, wheezing, orthopnea and increased lower extremity edema. She was put on non invasive positive pressure ventilation transiently. Chest x-ray was done, showing vascular congestion, and an echocardiogram was done, revealing a reduced ejection fraction (EF) of 30%. She was seen by cardiologist and diuresed with 40 mg intravenous furosemide. On Day-2 of admission, she suddenly developed 10/10, intermittent chest pain radiating to her neck, that lasted for an hour. ECG revealed new T-wave inversions in leads II, AVL, V4-V6. Troponin was found to be 3.1. At this time, she was immediately transferred to University of Maryland for cardiac catheterization. Catheterization findings included normal coronary arteries, left ventricle with hyperkinetic basal segments and an akinetic apex. After about 10 days of hospital stay, she was discharged with EF-25-30% and was asked to follow up with cardiology.

DISCUSSION:
Takotsubo cardiomyopathy was termed “broken heart syndrome” as most presentations of the syndrome occur after severe emotional stress. Our patient presented with symptoms of acute respiratory failure, which could have lead to physical and emotional stress. The hallmark of TCM that differentiates it from MI is the ballooning of the left ventricular apex with a hypercontractile base with no coronary artery obstruction. However, even though clinicians attribute this syndrome to an emotional trigger as an inciting event, the pathogenesis of Takotsubo CM is still not well understood, and here we see an example of a patient with Takotsubo who had no obvious severe emotional stressor.

CONCLUSION:
It is important to keep Takotsubo CM in the differential diagnosis list when dealing with chest pain patients with ECG changes and troponinemia, despite the lack of an emotional stress trigger.
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HERBAL TEA AND VITAMIN C: A MYSTERY OF ACUTE RENAL FAILURE
Jennifer Yeh, MD, Daniel Ambinder, MD, Catherine Handy, MD, John Sperati, MD
Johns Hopkins Medical Institutions, Baltimore, MD

A 54-year-old man presented with fatigue, headache, and nausea after a recent trip to Cameroon. He was found to have acute kidney injury with a serum creatinine of 18.9 mg/dL from a baseline of 1.5 one year prior. Past medical history was notable for diabetes mellitus and hypertension, and relevant medications included ascorbic acid 1000 mg daily. Urinalysis demonstrated 11 WBC per high power field but no hematuria or proteinuria. Kidney biopsy revealed acute tubular injury, interstitial edema, and numerous intraluminal polarizable calcium oxalate crystals, consistent with a diagnosis of calcium oxalate nephropathy [Fig]. He was initiated on hemodialysis. His history and age of presentation were not consistent with primary hyperoxaluria. He endorsed consuming significant quantities of herbal cleanses/teas purchased in Cameroon. He did not knowingly consume ethylene glycol, star fruit, or cucumber tree fruit, all of which have been associated with calcium oxalate nephropathy. He did not have a history of malabsorption or gastric bypass surgery, which increases absorption of enteric oxalate and thus urinary oxalate excretion. His kidney injury was likely caused by consumption of the herbal cleanse/tea and ascorbic acid, both of which lead to secondary hyperoxaluria. Indeed, oxalate nephropathy has been reported with excessive iced tea consumption, as black tea contains high levels of oxalate [1]. Additionally, ascorbic acid is metabolized to oxalate and can lead to oxalate nephropathy at doses less than 2 g/day [2].

This case illustrates calcium oxalate nephropathy from secondary hyperoxaluria, likely due to herbal cleanses/teas in combination with daily ascorbic acid supplementation.

Fig:

Coincidence vs. Association??

Case: A 70-year-old man with history of parkinsonism, complicated left eye cataract surgery with repair presented to emergency room with three days of left eye periorbital swelling, redness, proptosis, pain and double vision, associated with fevers, diaphoresis, malaise, cough, and shortness of breath. Orbit CT established the diagnosis of left periorbital cellulitis with mild orbital cellulitis and no evidence of sinusitis. He was started on Vancomycin and Piperacillin-Tazobactam and admitted to the floor. Initial review of systems was negative except for above mentioned. On physical examination, he was afebrile (98 F), tachycardia (110), normotensive (126/70), respiratory rate of 16 with oxygen saturation of 95% on room air. Left eye examination was pertinent for chemosis, injection, pain with extraocular movements, proptosis, binocular diplopia. No visual acuity defect, afferent pupillary defect or disc edema in both eyes. Cardiopulmonary exam is unremarkable. Abdominal exam revealed mild right upper quadrant tenderness.

Laboratory results: leukocytosis of 36000 with neutrophilia, lactic acid of 2.7, total bilirubin of 1.2. Despite clinical improvement of the eye findings, patient continued to be in sepsis. Repeat evaluation accentuated the abdominal pain, triggering a CT and US of the abdomen which showed acute cholecystitis with large amount of stranding surrounding the gall bladder and in porta hepatitis. Blood cultures were negative. On further questioning, family remembered a complaint of intermittent abdominal pain starting 3 days prior. His infection was definitively controlled with cholecystectomy.

Discussion: This is a case of severe sepsis initially thought to be secondary to orbital cellulitis but eventually found to be acute cholecystitis. Lack of history of abdominal pain as primary symptom led the academic team think of other possible explanation of sepsis. A thorough review of systems and detailed physical examination might have led to earlier diagnosis and management by identifying acute cholecystitis as primary cause of sepsis. In our case the presence of mild orbital cellulitis led to early closure leading to delay in management.

First Author Information:

Name: Swaroopa R. Nalamalapu
Institution: GBMC

Daytime Phone: 469-386-4720

Co-Author(s) Associates: Gurkeerat Singh
Program Director’s Name: Paul Foster

(indicating review of abstract)
2017 Mulholland Mohler Resident Meeting

ALL THAT SHAKES IS NOT STROKE
Malaz Alissa, MD; Nelly Arnouk, MD; Charmian Sittambalam, MD
MedStar Franklin Square Medical Center, Baltimore, Maryland

Introduction:
Cerebellar degeneration can be caused by a variety of different conditions, including stroke, chronic alcoholism, infection, and autoimmune disease. We describe a case of a middle-aged woman who suffered progressive cerebellar decline, due to a rare cause, resulting in multiple instances of an incorrect stroke diagnosis.

Case:
A 58-year-old Caucasian woman presented to the Emergency Department with slurred speech, abnormal gait, and vertigo. Past history was only notable for diabetes mellitus type 1, and HTN. She had had multiple hospitalizations in the past 8 years for similar symptoms and was consistently diagnosed with stroke, despite multiple MRIs which were negative for diffusion restriction or flair abnormalities. The patient progressed from mild ataxia in 2009 to wheelchair bound by 2016. She had no history of alcoholism or family history of ataxia. Her vitamin B12, TSH, homocysteine, CRP, HIV, and syphilis testing were within normal limits. The only finding on the multiple head imaging was mild cerebellar atrophy. A serum paraneoplastic panel demonstrated a higher level of AChr ganglionic neuronal Ab. Additional workup was done to rule out false positive results of the AChr Ab, that returned with a positive ANA but negative DS-DNA, Sjogren’s AB, C3, C4, and RA Factor. Work up for underlying malignancy was negative.

Conclusion:
Although AChr related paraneoplastic cerebellar degeneration has been described in the literature, it is almost unknown to occur in an isolated fashion, as in this case. Unfortunately for our patient, Purkinje cell damage and neuronal loss had already occurred and treatment such as immunotherapy for underlying immune disorders and further work up for malignancy were felt to have little yield. This case signifies the importance of investigating alternative causes of ataxia and dysarthria early once stroke is ruled out. Delay in the work up and potential treatment can lead to rapid progression and irreversible neurological damage.
SUBMANDIBULAR SIALOLITHIASIS IN A PATIENT WITH SJOGREN’S SYNDROME.

Nida Maham MD¹, Rajarajeshwari Ramachandran MD², Robert Ferguson MD³, Greater Baltimore Medical Center, Baltimore, Maryland.

INTRODUCTION: Sjogren’s syndrome is an autoimmune disease that targets the exocrine glands. Patients with Sjogren’s syndrome have reduced salivary secretions and are predisposed to salivary gland sialolithiasis.

CASE REPORT: 80-year-old woman with Sjogren’s syndrome and seronegative rheumatoid arthritis was admitted with jaw pain and altered mental status for one day. On the day of admission, she started having jaw pain with mastication and as the day progressed, she became lethargic and was brought to the hospital by her friend. She uses ophthalmic lubricants and mouth washes for Sjogren’s syndrome and was receiving Methotrexate for rheumatoid arthritis. On physical examination, she was afebrile and hemodynamically stable. Cardiac, pulmonary and abdominal exam were unremarkable. She was noted to have fullness over the right side of the neck and had tenderness to palpation over the right submandibular gland. Her oral mucosa was dry and a 3 mm tan, firm structure could be palpated at the opening of the right Wharton’s duct. She was somnolent, but her neurological exam was non-focal. With the physical examination findings and patient’s history of Sjogren’s syndrome, submandibular sialolithiasis was suspected. It was confirmed by a CT scan of the neck, which showed several stones within the right Wharton’s duct (largest measuring 4 mm in size). When the right Wharton’s duct was gently milked, a 3 mm stone was expressed with some pus. Her condition improved with sialogogues and antibiotics for staphylococcal coverage.

DISCUSSION: Submandibular glands lie beneath the floor of the mouth and the Wharton’s duct arises from the medial surface of the gland and drains into the floor of the mouth near the frenulum of the tongue. The submandibular glands are more prone to stone formation because of the slow flow of saliva against gravity. Sjogren’s syndrome is characterized by diminished lacrimal and salivary gland secretions and can predispose to sialolithiasis. In patients with sialolithiasis, anti-cholinergic medications must be discontinued. They must be advised to use sialogogues (eg: tart candies). If super-infection is suspected, anti-staphylococcal antibiotics can be prescribed for 7-10 days. Stones measuring less than 2 mm often pass. If the patient’s condition did not improve with conservative management, endoscopy, lithotripsy, wire basket extraction or surgical removal of the stone can be performed.

Program Director’s Name: Dr. Paul N Foster, MD

(indicating review of abstract)
FIBROSING CHOLESTATIC HEPATITIS IN THE SETTING OF AIDS. Stonesifer, E. MD. Da, B. MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Fibrosing cholestatic hepatitis (FCH) is a potentially fatal complication of hepatitis C (HCV) recurrence after liver transplantation, affecting approximately 10% of patients post-operatively and presenting with cholestasis, jaundice and high levels of HCV viremia. FCH has also been described in other immunosuppressed states such as chemotherapy and bone marrow transplantation.

A 37 year-old man with history of untreated HIV, CD4 count of 65 mm³, HCV with cirrhosis and active drug use presented with leg pain after a loss of consciousness on to a hard floor. He was diagnosed with rhabdomyolysis, acute kidney injury and community acquired pneumonia, and started on ceftriaxone, azithromycin and vancomycin. Liver function tests (LFTs) 1 year prior included aspartate aminotransferase (AST) 170 IU/mL, alanine aminotransferase (ALT) 110 IU/mL, alkaline phosphatase 109 IU/mL and total bilirubin 1.1 mg/dL. Admission LFTs included AST 304 IU/mL, ALT 101 IU/mL, alkaline phosphatase 82 IU/mL, and total bilirubin 9.9 mg/dL. Initially it was thought that his abnormal LFTs were due to sepsis. There was no improvement despite adequate resuscitation and antibiotic therapy and the degree of cholestasis was felt to be out of proportion to cirrhosis. Total bilirubin peaked at 44.2 mg/dL 7 days after admission. Workup for alternative causes of cholestasis including ferritin, serum iron and transferrin, anti-nuclear antibody, smooth muscle antibody, acute hepatitis panel, ceruloplasmin and 24-hour urine for copper was negative. HCV viral load was 6,779,262 IU/mL. Multiple imaging studies including computed tomography (CT), ultrasound, and magnetic resonance imaging (MRI) described a cirrhotic-appearing liver with patent vasculature but no evidence of biliary dilatation or cholestasis to the degree he experienced serologically. Transjugular liver biopsy showed cirrhotic architecture and findings consistent with chronic hepatitis C and significant ductal proliferation suggestive of FCH. The patient was offered direct-acting antiviral (DAA) therapy for HCV but in the setting of progressive liver and kidney failure, he declined and was discharged to hospice.

Often discussed in post-liver transplant literature, FCH has rarely been described in the setting of HIV. In the era of anti-retroviral therapy, cases of such profound immunosuppression are less common. This case demonstrates that in untreated HIV with HCV co-infection, FCH may be the cause of rapidly progressive cholestatic hepatitis.
2017 Mulholland Mohler Resident Meeting

ARTHRITIC SHOULDER PAIN: A UNIQUE MANIFESTATION OF ACUTE PROMYELOCYTIC LEUKEMIA. Nakhoda S, MD, Christopher K, MD, Emadi A, MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Acute promyelocytic leukemia (APL) is a subtype of acute myeloid leukemia characterized by the presence of translocation (15;17), coding for the PML-RARA gene. Without treatment, APL carries significant morbidity, with a median survival of less than one month, largely due to development of disseminated intravascular coagulopathy. With urgent therapy consisting of all-trans retinoic acid (ATRA) and arsenic trioxide, APL is associated with the highest cure rate within AML subtypes.

A 52 year-old Caucasian woman with a history of hypertension and osteoarthritis presents with chronic right shoulder pain for several years which had acutely worsened over the preceding 3 weeks and new onset arm swelling over the previous 2 days. An outpatient MRI which showed an abnormal lesion on her humeral greater tuberosity. She was referred to an orthopedic surgeon who performed a fine needle biopsy. This revealed acute leukemia cells concerning for acute promyelocytic leukemia. She was urgently admitted for initiation of ATRA and arsenic trioxide. Fluorescence in situ hybridization of the lesion biopsy was positive for translocation (15;17), confirming the diagnosis.

Upon admission, she was largely asymptomatic aside from mild ache in her right shoulder. She denied fevers, chills, weight loss, or fatigue. Lab work revealed a white count of 3.5 K/mcL, hemoglobin of 13.7 g/dL, and platelet count of 121 K/mcL. In addition, flow cytometry on peripheral blood revealed no abnormalities. A bone marrow biopsy was obtained from the posterior iliac which revealed normocellular bone marrow with trilineage hematopoiesis and no APL involvement. PET scan revealed increased FDG uptake in the appendicular skeleton but otherwise no abnormalities. She displayed no signs of disseminated intravascular coagulopathy or tumor lysis syndrome on lab work and tolerated treatment with no significant side effects.

Acute myeloid leukemia manifests in 2-8% of all cases as a myeloid sarcoma, a prominent extramedullary malignant lesion presenting with or prior to development of bone marrow disease. Due to the rarity of this diagnosis, large retrospective data is lacking. Current literature suggests cases of isolated myeloid sarcoma have a better prognosis than patients with AML without myeloid sarcoma and treatment is largely the same. This case highlights the diversity of oncologic disease manifestations and the success of multidisciplinary collaboration in management of an oncologic emergency.
When in Doubt, Think Viral
HSV-1 Epiglottitis

Introduction: Epiglottitis is most commonly caused by bacterial pathogens but seldom cultures remain negative in approximately 44-77% of cases. Viral epiglottitis is rare. We hereby report a case of epiglottitis that failed antibiotic treatment and was found to be caused by Herpes simplex virus-1 (HSV-1).

Case: A 49-year-old previously healthy male presented with a 4 day history of sore throat, odynophagia, subjective fever and globus sensation followed by progressive dysphagia and shortness of breath that developed a few hours before admission. On physical examination vital signs were: temperature 38.1°C, pulse 122 bpm, respiratory rate 18/min, and oxygen saturation 98% while on a non-rebreather face mask. He appeared dyspneic and was sitting in an erect position. He had dysphonia, a normal appearing oropharynx, and mild tenderness to palpation over the anterior cervical triangle. No palpable lymphadenopathy or skin lesions were noted. Laboratory results included a white blood count of 18,000 cells/microL with 91% neutrophils. Neck computed tomography (CT) scan with contrast revealed diffuse swelling of the epiglottis with decreased airway patency at the level of the hypopharynx, extending to the level of false cords; there was no evidence of abscess and the true cords appeared normal. Flexible nasopharyngeal laryngoscopy revealed edema of the lingual and laryngeal side of the epiglottis as well as the aryepiglottic folds and arytenoids with normal appearing true cords. The patient underwent endotracheal intubation due to acute airway compromise and impending loss of airway patency. He was started on antibiotics for presumed bacterial epiglottitis after collection of blood culture samples. The patient failed to improve and on the fourth day of hospitalization video laryngoscopy revealed severe epiglottic edema with purulent secretions. No ulcerative or vesicular lesions were identified. The epiglottitis was swabbed and specimen was sent for bacterial cultures and viral serologies. HSV-1 PCR came back positive. Sputum and blood cultures remained negative after several days. The patient was started on valacyclovir and was successfully extubated on the seventh day of hospitalization and discharged on day 10 to complete a total of ten days of antiviral therapy.

Discussion: Whilst in children epiglottitis is readily recognized and treated with standard antibiotic therapy, in adults epiglottitis can be mistaken for more common upper respiratory infections, which can insidiously or abruptly progress to airway obstruction requiring emergent endotracheal intubation. Approximately one third of patients have been seen by a primary care physician and received antibiotics for an upper respiratory infection before the diagnosis is made. Worrisome signs of airway obstruction such as stridor and sitting erect are present in only 12% and 16% of patients respectively. While viral epiglottitis is rare, the diagnosis should be considered when ulcerations or vesicles are identified on laryngoscopy; unfortunately, severe edema with abundant secretions may limit visualization of such lesions, as in this patient. Four cases of HSV epiglottitis in adults have been published in the English literature since the original report by D’Angelo et al. in 1990. Of these, three patients presented with ulcerative lesions on laryngoscopy which prompted the diagnosis and two presented with polypoid masses with histopathologic findings consistent with HSV. None of these patients were diagnosed with immunosuppressive states or evidence of disseminated herpes infection. PCR HSV-1 has higher rate of detection, faster turnaround times, and higher sensitivity and specificity as compared to IgM detection and cultures. Conservative management is advocated by some authors; however, the mortality in adults has been reported to be up to 7.8 times higher as compared to children.

Conclusions: This case highlights the importance of becoming familiar with HSV-1 as a potential etiology of epiglottitis, suspecting a viral infection when patients fail to respond to antibiotic therapy, and using videoendoscopy in the assessment of epiglottitis and its response to treatment.
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<th>LUPUS OVERLAP WITH POLYMYOSITIS PRESENTING WITH RHABDOMYOLYSIS</th>
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<td>Praveena Sunkara, MD; Rozi Khan, MD; Brent Harris, MD; Irfan</td>
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<td>Shaukat, MD; Maimoona Inayat, MD</td>
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<td>MedStar Good Samaritan Hospital, Baltimore, Maryland</td>
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Myositis is a known complication of Systemic Lupus Erythematosus (SLE); however, lupus presenting as myositis is extremely rare. This is a case of a middle aged man with severe myositis causing rhabdomyolysis as the initial presentation of SLE.

A 58y/o man with h/o hypertension presented with extreme weakness of bilateral biceps, which he attributed to an increased exercise regimen in the past few weeks. The patient also stated that he felt weaker in his legs. The weakness had gotten progressively worse to the point of involving both shoulders and thighs. He also complained of dark, tea colored urine during the same period, although he denied dysuria. At the time of admission, creatinine was markedly elevated with severe rhabdomyolysis, with CK of 24,000. Physical examination revealed decreased strength in bilateral upper and lower extremities. Bilateral upper extremities were swollen left > right. There was no evidence of infection or compartment syndrome. Treatment was initiated with aggressive intravenous hydration, with concurrent infusions of bicarbonate to alkalinate the urine. Despite aggressive hydration, his condition worsened and he continued to have worsening weakness with decreased in functional status for several days. Additional history revealed family history of SLE (patient's mother), which prompted autoimmune work-up, resulting in ANA that was positive at 1:640. Extensive work up revealed negative anticardiolipin, anti-Jo, and anti-smith antibodies. Muscle biopsy was done which revealed changes associated with inflammatory myositis. The patient was diagnosed with possible lupus overlap with polymyositis. He was pulsed with steroids and received IVIG and plasmapheresis and was discharged on azathioprine. Over the course of the next 4 months, the patient’s weakness had considerably improved with simultaneous decrease in CPK to 309.

This case illustrates the importance of keeping a broad differential, taking a good family history, and recognizing that myositis could be a possible initial presentation of SLE in men. In this case, prompt recognition and institution of appropriate therapy prevented further deterioration of the patient’s functional status.

<table>
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<th>First Author Information:</th>
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<tbody>
<tr>
<td>Name: Praveena Sunkara</td>
</tr>
<tr>
<td>Institution: MedStar Health Internal Medicine, Baltimore</td>
</tr>
<tr>
<td>Daytime Phone: 4105542284</td>
</tr>
</tbody>
</table>

Co-Author(s) Associates: Rozi Khan, M.D., Brent T. Harris MD, Irfan Shaukat, M.D., Maimoona Inayat M.D.

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Program Director’s Name: Stephanie Detterline, MD

(indicating review of abstract)
MORE THAN A SINUS HEADACHE: A CASE OF CAVERNOUS VENOUS SINUS THROMBOSIS
Abhilasha Singh, MD; Syed Rameez Jafri, MD; Sadaf Mustafa, MD
MedStar Union Memorial Hospital, Baltimore, Maryland

Introduction:
Septic cavernous venous sinus thrombosis (CVST) is a rare condition with high morbidity and mortality. Although the pathogenesis of CVST is poorly understood, the two most well defined mechanisms that may contribute to its development include either thrombosis of the cerebral veins and dural sinuses or occlusion of the dural sinuses leading to decreased CSF absorption. In 85% of adults at least one or more risk factors can be identified.

Case Summary:
A 67-year old female with hypertension was sent to the ED from her PCP’s office with sore throat, flu-like symptoms, sinus pain and pressure for 1 week, worsening headaches, left eye ptosis, chemosis, and diplopia for 12-24 hrs. She had not been prescribed hormone replacement therapy or OCPs in the past and had no history of prior thrombotic event. On examination, her vital signs were stable and she was noted to have left eye proptosis, ptosis and diplopia. Oral and neurological examination was unremarkable. Tonometry was negative for raised intraocular pressure. Labs demonstrated leukocytosis (WBC count of 27.8) with bandemia. CT of the head was negative for intracranial pathology but revealed left maxillary acute sinusitis. CTA of the head revealed bilateral cavernous sinus thrombosis. Blood cultures were obtained and the patient was started on broad-spectrum antibiotics (ceftriaxone, vancomycin, and metronidazole), anticoagulation, and steroids, which were tapered shortly thereafter. Her blood cultures turned positive for *Aggregatibacter*, *Corynebacterium* and *Streptococcus constellatus*. Testing was negative for Factor V Leiden mutation and an echocardiogram was negative for thrombus or vegetation. Antibiotics were narrowed to ceftriaxone 2g q12h for a total of 21 days. Anticoagulation was bridged to warfarin.

Discussion:
The presentation of bilateral septic CVST may be very nonspecific. A thorough history and physical examination focusing on the potential contributing factors along with dedicated imaging in a timely manner is necessary for establishing the diagnosis. Prompt treatment is essential and is usually directed towards identifying the precipitating factor and eradicating it. Treatment includes antibiotics, anticoagulation and anticonvulsive treatment, if indicated.
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UNEXPECTED DIABETIC KETACIDOSIS AS A WARNING SIGN FOR PANCREATIC ADENOCARCINOMA
Travis Weaver D.O. (Associate), Michael Godar M.D. (Associate),
Brett Sadowski M.D. (Member)
Walter Reed National Military Medical Center

Pancreatic cancer is a devastating diagnosis for many patients and ranks as the fourth leading cause of cancer mortality in the U.S., as well as the second most common GI malignancy. Although surgical resection can be curative, the majority of patients present beyond the point when curative resection is possible. This is owed to the fact that they usually present with vague symptoms and are often identified at a late stage. These factors, along with relatively few recent advances in treatment, give pancreatic cancer the distinction of having the highest mortality rate of all major cancers.

Our patient is a 77 year old male with a history of well-controlled type II diabetes mellitus who presented to our outpatient clinic for follow up two months after he was admitted to the intensive care unit (ICU) for diabetic ketoacidosis (DKA). During his outpatient appointment, he was noted to have a new right upper quadrant abdominal mass as well as significant liver function test abnormalities with an alkaline phosphatase of 696 U/L, compared to 59 U/L during his ICU admission. He was admitted to the medical ward where abdominal computed tomography (CT) revealed a mass in the head of the pancreas, common bile duct dilation, significant gallbladder distention into the abdominal wall, in addition to diffuse liver metastases. Biopsy via endoscopic ultrasound and fine needle aspiration revealed adenocarcinoma. The patient elected to receive hospice services and passed away two months later.

This is a case of newly diagnosed pancreatic cancer with an atypical initial presentation. Primary pancreatic malignancy classically presents with painless jaundice and is frequently metastatic at the time of presentation. This patient with previously well controlled type II diabetes mellitus presented with DKA 12 weeks prior to his cancer diagnosis. DKA in this patient could be viewed as a harbinger of his malignancy and may have lead to earlier diagnosis. Diabetogenic changes to pancreatic beta cell function have been previously described. Identification of these diabetogenic factors may have a future role in early detection of pancreatic malignancy. When a patient presents with unexpected and unexplained DKA, considering pancreatic malignancy in the differential diagnosis could lead to an earlier diagnosis of this devastating disease.

Program Director’s Name: William Shimeall

(indicating review of abstract)
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A Rare Cause of Intussusception

Introduction: Intussusception in adults is rare, accounting for only 5% of all cases, and 1% of cases involving the small bowel. There is a dearth of literature on adult intussusception, but current evidence suggests that malignancy, polyps, and diverticulae are the usual culprits. We present a case of adult ileocecal intussusception secondary to carcinoid tumor.

Case: A 53-year-old male presented with hematochezia and subsequently developed non-radiating constant left upper quadrant pain accompanied by nausea and vomiting. CT pelvis showed a pathognomonic “target” sign, consistent with ileocecal intussusception and early small bowel obstruction. Two years prior to this current presentation, he had an episode of hematochezia for which he underwent colonoscopy and polypectomy, which was negative for colon cancer. He denies diarrhea, constipation, weight loss, decreased appetite or skin flushing. During the exploratory laparotomy, a white-colored, chalky mass indicative of penetrating tumor was noted 6-7 inches distally from ileocecal junction. An extended right hemicolectomy followed the discovery of the mass. Pathology showed a well-differentiated neuroendocrine tumor consistent with Carcinoid tumor. Evaluation for metastatic disease using serum 5’HIAA and CgA was fortunately unremarkable, and the resection of the right colon carcinoid tumor was felt to be curative.

Discussion: Adult ileocecal intussusception secondary to carcinoid tumor is a rare clinical presentation, as only a handful of cases have been reported. The pathognomonic “target” sign lesion will typically be demonstrated on the CT. Carcinoid tumors may present without the typical symptoms of carcinoid syndrome. Intussusception may be the only manifestation of carcinoid tumor; therefore, the cause of intussusception should always be assiduously uncovered. As Carcinoid tumors secrete 5-hydroxyindole acetic acid (5-HIAA) and chromogranin A, serial measurement of 24-hour urinary 5-HIAA can aid in evaluating for possible metastatic involvement.

Conclusion: It is uncommon for adults to present with intussusception; it is crucial to rule out malignancy as an underlying cause. Among the secondary causes such as inflammatory bowel disease, colon adenocarcinoma and lipomas, Carcinoid tumor should be considered among the malignancies that can cause intussusception.

First Author Information:

Name: Jennie Zhang, DO

Institution: UMMC Midtown Campus

Daytime Phone: (410) 225-8790

Co-Author(s) Associates: Naomitsu Watanabe, MD, Dena Tran MS III

Program Director’s Name: Dobbin Chow, MD

(indicating review of abstract)
Gerstmann syndrome with left Corona Radiata and Basal Ganglia infarct
A Jat, M.D., S Limani, M.D., B Waseem, M.D., M LaMonte, M.D., U Khan

Introduction: Gerstmann syndrome or angular gyrus syndrome is a psychoneurologic syndrome with four cardinal features: dysgraphia, acalculia, finger agnosia, and left-right confusion; this is known as the Gerstmann tetrad. It is commonly associated with lesions involving the dominant parietal lobe; however, other areas of the brain have also been found to be involved. We present a case of Gerstmann syndrome in a patient with a left corona radiata and basal ganglia stroke.

Case description:
Chief Complaint: Impaired speech
History of presenting illness: A 73-year-old, right handed female, with uncontrolled hypertension, presented complaining of impaired speech that had started 6 hours before presentation. At baseline patient was independent on all activities of daily living and had normal speech
Physical exam:
Blood pressure: 247/124 mm Hg
Neurologic exam: alert, cooperative and oriented in four spheres. Minor telegraphic speech impairment. No apraxia. Unable to answer any simple calculations correctly despite her educational level. Unable to name fingers. Although she did not display neglect or extinction, she was unable to correctly identify body parts and objects in the room as localized to the right or left. While she could comprehend and follow all commands, read and repeat, she could not write. Thus she demonstrated acalculia, finger agnosia, right-left confusion, and agraphia without significant language or other processing deficits.
Imaging:
Brain computed tomography (CT): no acute intracranial process
Brain CT angiogram: no large artery occlusions
Brain magnetic resonance imaging: acute to subacute infarct in the left corona radiate and basal ganglia.

Patient was admitted on neuro-tele floor. She was managed per stroke protocol with permissive hypertension. She received Aspirin and statin. Anti-hypertensives were started on day 3 of admission. Her language and the components of her Gerstmann syndrome were improving by day 5 and she was discharged to subacute rehabilitation.

Discussion: Pure Gerstmann syndrome is a rare disorder as typically one or more components of the tetrad may be pseudodisplayed as a function of a co-existing language, other cognitive visuospatial processing disorder, or apraxia. Given this some experts have challenged the existence of Gerstmann syndrome as an independent entity. Possible underlying causative disorders that have been postulated include: impairments in spatial orientation and constructional apraxia, selective disorders of the body schema in the hand, language impairments, general intellectual deficits, a disconnect between verbal and visuospatial functions, defective horizontal mapping, and impairment in the manipulation of mental images which require visuospatial processing. None of these theories have panned out as there have been patients with the Gerstmann tetrad who test normally in the aforementioned areas. In this case the patient presented with telegraphic aphasia which is usually seen with dominant subcortical lesions; however, her comprehension, repetition, and naming were normal. Therefore, the presence of Gerstmann syndrome was clearly evident and we can surmise her symptoms were not merely a consequence of her aphasia.

Conclusion: Gerstmann syndrome can present with lesions involving left (dominant) basal ganglia. This case adds to the evidence that Gerstmann syndrome may be more than just a manifestation of aphasia or other primary brain process dysfunction.
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General Classification:

( x ) Clinical Vignette
( ) Research Competition
( ) Basic Science
( ) Evidence based medicine review
( ) Quality/Safety
( ) Clinical Research

Indicate your participation in research process (4 sentences or less):

First Author Information:

Name: Rosalie Yan
Institution: GBMC

Daytime Phone: 469-386-4720

Co-Author(s) Associates: Amit Kulkarni, Mustafa Mohammed, Surendra Marur
Program Director’s Name: Paul Foster

(indicating review of abstract)
Black Esophagus: A Cause or Effect of Diabetic Ketoacidosis (DKA)?

Despite having an incidence of 0.01 – 0.28%, Acute Esophageal Necrosis (AEN) should be considered when a patient with DKA presents with symptoms of an upper GI bleed. Our case report highlights one such presentation.

A 30-year-old woman with a history of Type 1 diabetes (anti-GAD65 positive) diagnosed at the age of 25 in the setting of DKA was brought to the ED by paramedics for altered mental status. On the day of admission, the patient contacted her sister via instant messaging to say that she had vomited a dark substance. She continued to send messages to her sister during the day, but her messages progressively became less coherent. As a result, her sister went to check on her and found her unresponsive on the floor. Upon admission, the patient was relatively hypotensive (BP 90/50 mmHg), tachycardic (HR 132 bpm), and tachypneic (RR 32/min). Exam was notable for diffuse abdominal pain and Kussmaul respirations. Laboratory results were remarkable for leukocytosis (WBC 45 K/cu mm), hyperglycemia (BG 1294 mg/dL), and high anion gap metabolic acidosis (Bicarbonate 4 mmol/L, Anion Gap 32 mmol/L, Venous Blood pH <6.8). She was admitted to the intensive care unit for DKA and shortly thereafter was noted to have 2 episodes of coffee ground emesis. Infectious work-up did not reveal a potential cause for her DKA. She denied feeling ill prior to the day of her admission and reported adherence to her insulin regimen. Her white count down-trended without the use of antimicrobials and her blood glucose was controlled on a slight adjustment to her home insulin regimen. The patient underwent an esophagogastroduodenoscopy (EGD) which revealed an area of necrosis with overlying black eschar in the distal esophagus – diagnostic of acute esophageal necrosis. Her diet was advanced as tolerated and she was discharged on proton pump inhibitor therapy. Plans were made to repeat EGD within an eight-week period to check for healing of her esophageal mucosa.

This case illustrates the association between DKA and AEN. Black esophagus is a rare syndrome that arises due to a combination of factors including ischemic insult seen with hemodynamically compromised states, corrosive injury from gastric outlet obstruction or gastroparesis, and decreased function of mucosal barrier systems present in malnourished and debilitated patients. It tends to occur in the distal third of the esophagus, which is relatively hypo-vascular compared with other esophageal segments. The cause and effect relationship of DKA and AEN has not been cited in the literature. One might think that given the chronology of this case, the trigger for this patient’s DKA is AEN; however, the mechanism remains unclear.
2017 Mulholland Mohler Resident Meeting

Title: HEPATIC ABSCESS: AN UNUSUAL COMPLICATION OF KLIPPEL-TRENAUNAY SYNDROME

Introduction: Klippel-Trenaunay Syndrome (KTS) is a rare disease that affects multiple bodily structures including blood vessels, bone, soft tissue, and lymphatics; patients often have vascular abnormalities such as port wine stain, bleeding due to malformations and limb hypertrophy.  

Case Description: A 49-year-old woman with history of KTS presented with severe abdominal as well as neck/shoulder pain and massive hepatosplenomegaly. Her surgical history was notable for prior embolization of vascular malformations including laser ablation of the bladder and alcohol embolization of buttock lesions. Vital signs on admission were T 37.3, HR 104, BP 120/68, RR 24, SpO2 93%. Her physical exam was notable for crackles in the right lung field as well as a distended abdomen with hepatosplenomegaly and diffuse abdominal tenderness. Laboratory studies demonstrated WBC count 7.3K/cu mm, hemoglobin 8.2g/dL, AST 10U/L, ALT 5U/L, alkaline phosphatase 127U/L and INR 1.3. MRI imaging revealed two lesions in the right lobe of the liver, the larger measured at 12.6cm x 12.1cm and additional abdominal US was concerning for hematoma/hemangiomma vs. cystic process. Percutaneous drainage of the lesion produced greater than 500 cc of purulent fluid. The patient subsequently developed septic shock requiring transfer to the intensive care unit. Culture of abscess was positive for E. Coli. The patient was stabilized via fluid resuscitation and broad-spectrum antibiotics and was eventually discharged on IV antibiotics with drains in place.  

Discussion: The patient’s multiple cystic lesions in the liver appear to have been a nidus for infection. Hepatic abscesses more commonly arise from biliary obstruction, infection in the abdomen and secondary to procedures. They occur less often from idiopathic cystic processes. This case is unique as complications of KTS more often involve bleeding; however, hepatic abscess is noted in other dural malformation phenomena such as Caroli’s disease.  

Works Cited:  
AN INCIDENTALOA: A CASE OF ASYMPTOMATIC LOA LOA. Manski, S MD, Patel D, MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Loa loa filariasis is a parasitic infection endemic in the rainforests of Western and Central Africa. Though Loa loa is rarely seen in the United States and Europe, immigration from endemic countries has led to higher rates of physicians encountering the disease in recent decades. Here we report a case of Loa loa, which was incidentally found. The patient’s low parasite burden and concurrent diagnosis of Gastrointestinal Stromal Tumor (GIST) provided a unique opportunity to treat both conditions with imatinib (Gleevec).

A 34-year-old man with a history of recent immigration from Cameroon presented with severe symptomatic anemia. The patient noticed bright red blood upon wiping after bowel movements, which later changed to melena. He then began experiencing fatigue and chills followed by a syncopal episode shortly after. On presentation, the patient denied vision changes, extremity pain, or hematuria.

On admission, the patient was febrile to 38.0°C. Ocular exam was negative for parasites under the conjunctiva. The patient had no evidence of subcutaneous non-erythematous areas of swelling on the extremities (calabar swellings). Laboratory data was significant for a microcytic anemia with a hemoglobin of 3.9g/dL and an MCV of 75 fl, requiring multiple transfusions. An endoscopy was performed, showing an 8-10cm gastric fundal ulcerative mass. A biopsy demonstrated a Gastrointestinal Stromal Tumor (GIST). Blood smears were also performed due to concern for malaria and revealed a microfilaria identified as Loa loa. The patient’s parasite load was calculated to be 5 microfilariae/mL.

In endemic areas of Western and Central Africa, 13 million individuals are estimated to be infected with this parasite. However, a large majority of those infected are clinically asymptomatic. Diethylcarbamazine (DEC) is regarded as standard treatment for symptomatic individuals with a parasite load <8000 microfilariae/mL. In our case, Loa loa was incidentally found and did not require treatment. However, a recent study demonstrated the filaricidal activity of the tyrosine kinase inhibitor imatinib in vitro. The patient was then discharged with planned follow up with Oncology for Gleevec therapy. This case of Loa loa illustrates a unique encounter and a rare opportunity to treat both a parasite and tumor with the same drug.
IS THIS GLUTEN FREE? A CASE OF LIFE-THREATENING TYPE II REFRACTORY CELIAC DISEASE. McCusker M, MD, Christian K, MD. The University of Maryland School of Medicine and Baltimore VA Medical Center, Baltimore, MD.

Refactory celiac disease (RCD) is an uncommon, but severe complication of celiac disease (CD). Celiac disease is a chronic immune-mediated enteropathy precipitated by exposure to dietary gluten that affects 1 to 2% of the United States population. The disease is diagnosed by evidence of malabsorption, positive celiac-specific antibodies, and villous atrophy in duodenal biopsies. The disease is treated with life-long gluten free diet (GFD), which leads to disease regression. RCD is defined as persistent or recurrent signs and symptoms of malabsorption, with small intestinal villous atrophy despite a strict GFD for more than 12 months, in the absence of other causes of villous atrophy. It is divided into two types based on the absence (Type I) or presence (Type II) of abnormal intraepithelial lymphocytes (IEL). Type II RCD (RCD-II) is exceedingly rare and entails a worse prognosis. The disease is often complicated by severe malnutrition, ulcerative jejunoileitis, and enteropathy associated T-cell lymphoma (EATL).

A 55-year-old man with well controlled CD over 12 years who presented to the emergency department with a 25-pound weight loss, diarrhea, fatigue, and swelling over one month in the setting of strict gluten-free diet. The patient's work-up revealed negative transglutaminase antibodies and endomysial IgA. Endoscopy and colonoscopy revealed crypt hyperplasia in the duodenum and ulceration in the ileum. Capsule endoscopy then showed ulcerative jejunoileitis and balloon enteroscopy with biopsy also revealed ulcerative jejunitis with villus blunting. Immunohistochemical staining was performed on jejunal biopsies and showed evidence of IEL clonal population. The patient was started on budesonide, urgently referred to Mayo Clinic to be evaluated by a small group of RCD experts, and is currently being considered for novel investigational treatments.

RCD is a rare, though severe condition developing in 1 to 5% of the adult-onset CD patients. The diagnosis of RCD-II has a poor prognosis with a 5-year survival rate of 33 to 52%. Treatments are poorly outlined and are targeted toward elemental and/or total parenteral nutrition and immunosuppression. This case presents an unusual situation of a patient diagnosed with CD under the age of 50 years with 12 years of diet-controlled disease and acute onset of life threatening RCD-II.
Generalized Weakness in a Young Male: A case of Thyrotoxic Periodic Paralysis

Introduction: Thyrotoxic periodic paralysis (TPP) has traditionally been a well-known complication of thyrotoxicosis in Asian populations; however, it is becoming more in Western countries. Therefore, it is important to consider TPP if the clinical picture is compatible with the condition regardless of the patient's ethnicity. Early diagnosis and treatment may prevent life-threatening cardiopulmonary complications associated with hypokalemia and muscle weakness. We hereby report a case of TPP in an African American patient caused by Graves' disease.

Case: A 30-year-old African American male with no significant past medical history presented to the emergency room with acute onset generalized muscle weakness. The night before presentation he experienced sudden lower extremity weakness while going up a flight of stairs and almost fell. He went to sleep, woke up with generalized weakness and was unable to stand up. When questioned about food intake he reported that dinner the night before had a high-carbohydrate load. Review of systems was positive for intentional weight loss (almost 100 pounds over the last year), hand tremor, excessive sweating and heat intolerance. He denied taking medications on a regular basis as well as smoking, ethanol or illicit drug use. Family history was significant for Graves' disease in mother and maternal aunt. No family history of weakness or paralysis. Physical examination was significant for blood pressure 138/74 mmHg, heart rate 140 beats/minute, respiratory rate 18 per minute, temperature 36.5°C; he was alert, well-developed, well-nourished, and not in distress. No lid lag, lid retraction or edema, and no exophthalmos were noted. Thyroid was non-tender on palpation, no nodules were appreciated, its size was 30-40g, and a bruit was auscultated over the right thyroid lobe. On cardiovascular auscultation he had an irregularly irregular rhythm and tachycardia. Strength was 4/5 in all four limbs. Pertinent laboratory investigations included TSH 0.007 mIU/L, free thyroxine 7.76 ng/dl, total tri-iodothyronine >650 ng/dl, total thyroxine 19.6 ug/dl, thyroid stimulating immunoglobulin 46%, thyrotropin binding inhibitory Ig 21.52 IU/L, potassium 2.2 mEq/L, phosphorus 3.0 mg/dl, magnesium 1.4 mg/dl.

A diagnosis of TPP associated with Graves’ thyrotoxicosis was made. He was treated with potassium supplementation, propranolol and methimazole. Within hours strength had returned to normal, potassium levels had normalized and heart rhythm had converted back to sinus.

Discussion: TPP has an estimated incidence of approximately 0.1-0.2% among the hyperthyroid population in North America. The pathogenesis of TPP remains unclear, previous reports revealed that high levels of thyroid hormone increases the number and activity of the Na+/K+/ATPase pump increasing the shift of potassium into the cell, leading to hypokalemia and loss of muscle excitability and paralysis. Androgens also increase the activity of the pump, which might explain the male predominance of this condition. Insulin similarly activates the Na+/K+/ATPase pump; therefore, anything that promotes hyperinsulinemia, such as high-carbohydrate meals, can trigger or exacerbate episodes of paralysis. TPP only occurs in a small minority of patients with hyperthyroidism. Thus additional factors, such as a genetic predisposition, are presumed to contribute. Nonselective beta-blockers attenuate Na+/K+/ATPase pump stimulation and are recommended as part of the management during acute attacks combined with antithyroid medications. Clinicians should exercise caution when replacing potassium as high doses may lead to rebound hyperkalemia given the hypokalemia is due to compartment shifts and not a true potassium deficit. Patients should be educated about triggers and to avoid activities that may precipitate episodes of paralysis. Awareness is a key-element for early diagnosis and treatment.
Sudden Lost of Sensation in a Patient with Recent Back Surgery

Eric Feng DO, Michelle Le DO, Tim Brennan MD, Tushar Sharma MBBS, Olufunmilayo Ogundele MD, Lee-Gardie Jean MD, Department of Medicine, Sinai Hospital of Baltimore, Baltimore, Maryland

Phlegmasia cerulea dolens (PCD) is a rare condition that results from extensive deep vein thrombosis of an extremity, and is associated with high morbidity and mortality rates due to obstruction in arterial blood flow with critical limb ischemia and circulatory collapse. A 51-year-old woman with history of recurrent unprovoked deep vein thromboses (DVT) and pulmonary emboli (PE) without underlying thrombophilia who has an IVC filter and is on lifelong rivaroxaban, presented to the ER for altered mental status and hypotension. She had taken large amounts of opiates earlier for pain related to spinal laminectomy performed 7 days before presentation. After treatment with naloxone and intravenous fluids, she showed appropriate improvement. On waking up, she complained of right leg pain, and mentioned that she has been off rivaroxaban for 8 days, and it is to be resumed in 2 days. A venous doppler of right leg was done that was negative for DVT. Twenty four hours later, she started complaining of worsening pain and loss of sensation in both legs. On exam, she was hypotensive and tachycardic. Both lower extremities demonstrated edema, tenderness and violaceous discoloration. Emergent venous doppler was done that revealed extensive deep vein thrombosis involving external iliac, common femoral, femoral, and popliteal veins of bilateral lower extremities. Immediate anticoagulation with dabigatran was begun, followed by multiple attempts at catheter-directed thrombolysis which proved to be futile due to heavy clot burden. She died 2 days later.

PCD is considered a precursor of frank venous gangrene, and is characterized by sudden severe pain, swelling, cyanosis and edema of the affected limb. Severe venous congestion causes impairment of arterial supply, frequently followed by shock and circulatory collapse. Associated risk factors include malignancy (in up to 50% cases), surgery, trauma, pregnancy and presence of IVC filter, but up to 10% cases are thought to be idiopathic. Mortality rates can be as high as 40%, and amputation rates of up to 50% have been reported in those who survived. Early recognition and prompt treatment are a must to prevent loss of limb or death. PCD is a widely accepted indication for thrombolysis and/or thrombectomy, and patients should be managed with systemic anticoagulation in the interim. Intravenous unfractionated heparin has been used most commonly and should be promptly initiated to improve outcomes of this rare but life-threatening condition.
Title: IS IT ALWAYS LUPUS?
Authors: Rachit Vakil, MD (ACP Member); Ruth Tamrat, MD (ACP Member); Sarath Raju, MD (ACP Member)
Introduction: This is a unique case of a woman with a previously undifferentiated connective tissue disorder who presented with progressive dyspnea on exertion and a new onset cardiomyopathy.
Case Description: The patient is a 55-year-old African American female with a history of ESRD, recently diagnosed vaginal SCC, and an undifferentiated connective tissue disease presenting with two months of DOE and 2 weeks of PND. In 2002, the patient was diagnosed with lupus and MCTD based on positive ANA and high anti-RNP antibodies. She was briefly on hydroxychloroquine before self-discontinuing. The patient restarted hydroxychloroquine about one year prior to presentation, but stopped treatment when cisplatin therapy for recurrence of vaginal squamous cell carcinoma was initiated in mid-2016. The patient is a former smoker, worked as a programmer, and has no family history of autoimmune conditions. On presentation, the patient had mild tachycardia, distended JVP, bibasilar crackles and no edema. Lab abnormalities included a mildly elevated troponin, paraprotein gap, low complements, and high titer anti-RNP antibodies. TTE showed severe global hypokinesis of the LV with EF 20-25%. PET imaging revealed no evidence of myocardial ischemia during stress testing. Most significantly FDG metabolic images of her heart demonstrated patchy, abnormal FDG uptake suggestive of myocarditis with active inflammation.
Discussion: Major risks for non-ischemic cardiomyopathy in this patient included lupus and chemotherapy use. Cisplatin chemotherapy is not classically associated with non-ischemic cardiomyopathy. Differentials for inflammatory and autoimmune causes applicable to this patient included lupus, other rheumatologic disorders, and sarcoidosis. Given her negative stress test, clinical history of lupus with evidence of active disease on labs, and inflammation on PET/CT, the patient's heart failure was diagnosed as lupus myocarditis (LM). Biopsy was not pursued given her that her imaging findings and active lupus were enough to confirm the diagnosis. Clinical LM is rare, occurring in 6-9% of lupus patients. In a recent case series of 29 patients with LM, 81% of those treated with steroids and immunosuppressives showed significant improvement in EF and clinical cardiac recovery. Our patient received a methylprednisolone pulse, and was transitioned to oral prednisone and monthly cyclophosphamide. Optimal treatment for LM has yet to be established, and warrants further study.
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First Author Information:

Name: Alissa Werzen, MD

Institution: University of Maryland Medical Center and Baltimore VA Medical Center

Daytime Phone: 410-328-7567

Co-Author(s) Associates:

2017 Mulholland Mohler Resident Meeting

MYOCARDITIS AS A CAUSE OF CHEST PAIN IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS. Werzen A, MD.
The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Myocarditis in patients with systemic lupus erythematosus (SLE) is a major cause of morbidity and mortality. The autoantibodies implicated in the pathogenesis of SLE can directly damage all three layers of the heart – the pericardium, myocardium and endocardium, as well as lead to arrhythmias and conduction system disturbances. While lupus myocarditis remains a largely subclinical entity, it nevertheless should be considered in the differential diagnosis of lupus patients presenting with chest pain.

A 66-year old woman with a history of SLE on hydroxychloroquine, diabetes mellitus, and hypertension presented with 2 days of chest pain that improved with leaning forward. She also reported a 2-week history of fatigue, dyspnea on exertion, orthopnea, and bilateral lower extremity edema. The patient reported a history of a heart murmur of unknown etiology, but denied any other cardiac history. At the time of presentation, cardiac exam was significant for a systolic ejection murmur at the right upper sternal border and a systolic murmur at the left lower sternal border. She was also found to have crackles in the bilateral lung fields and 2+ pitting edema of the lower extremities. Labs revealed an elevated troponin, which peaked at 4.48 μg/L and an elevated creatinine kinase, which peaked at 4304 units/L. Her electrocardiogram did not have evidence of ischemic changes. Transthoracic echocardiogram showed a hyperdynamic left ventricle with left ventricular outflow tract obstruction, a moderate pericardial effusion without evidence of tamponade, and an ejection fraction of 70%. Subsequent cardiac catheterization revealed hypertrophic cardiomyopathy without significant aortic stenosis or coronary artery disease.

The patient was diagnosed non-ST elevation myocardial infarction secondary to myopericarditis in the setting of hypertrophic cardiomyopathy. She was treated with intravenous hydration, beta-blockers, and a steroid burst. Subsequently, her chest pain resolved and muscle enzymes returned to normal. This case presentation reiterates the importance of considering lupus myocarditis as a cause of chest pain in patients with SLE and its favorable response to treatment with steroids.

Program Director’s Name: Susan D. Wolfsthal

(indicating review of abstract)
**Suffocating kidney**

A Saeed, M.D., S Limani, M.D., S M. Bagnasco M.D.,2 R Pullman, M.D.

**Introduction:** Solitary extramedullary plasmacytoma (SEP) accounts for 3% of plasma cell malignancies. Retroperitoneal involvement is very rare and renal involvement is exceptionally rare. We present a case of SEP leading to renal failure via two rare disease processes.

**Case:** A 77-year-old man presented complaining of progressive weakness and decreased urination for more than one month. Physical examination was significant for blood pressure of 90/40 mmHg. Laboratory investigations revealed hemoglobin 8.5g/dL, potassium 6.8mEq/L, bicarbonate 5.2mEq/L, BUN 159mg/dL, creatinine 16.7mg/dL, phosphorus 11mg/dL, arterial blood gas pH 7.05. Urinalysis revealed 2+ protein and 1+ RBC. Additional urine studies: urine creatinine 153mg/dL, urine sodium 77 mg/dL, urine protein 642mg/dL. Renal ultrasound showed mild right hydronephrosis and evidence of medical renal disease. The patient was volume resuscitated with isotonic bicarbonate and blood pressure improved to 105/55 mmHg; unfortunately urine output remained low and he ended up requiring hemodialysis. A left kidney biopsy revealed moderate diffuse tubular atrophy with tubular cell vacuolization, bleeding and flattening of epithelium. Serum protein electrophoresis showed an M-spike in the gamma region (0.29 from 0.71 g/dl of protein in gamma region), sFLC kappa 385mg/dL, lambda 1.56mg/dL, and ratio 246.79. 24-hr urine collection revealed 2200 mg of protein, 68.7% (1.5 g) being kappa light chain. Bone marrow biopsy did not reveal any phenotypically abnormal cell populations. At this time the working diagnosis was ischemic acute tubular necrosis and monoclonal gammopathy of undetermined significance. After 3 weeks there was no meaningful renal recovery and the diagnoses were put into question. An abdominal computed tomography (CT) of the abdomen demonstrated prominent right hydronephrosis and a heterogeneous soft tissue mass anterior to the right psoas muscle compressing the ureter. A CT guided biopsy of the mass revealed sheets of mononuclear cells with central necrosis suggestive of a plasma cell neoplasm with kappa restriction. Flow cytometry confirmed the presence of an abnormal plasma cell population. Reevaluation of the kidney biopsy with immunostains and electron microscopy showed Kappa light chain positivity in the cells of the proximal tubules with a droplet inclusion pattern. The patient was started on dexamethasone and bortezomib. Kappa levels decreased dramatically (greater than 10 fold). After 6 weeks and 3 cycles of therapy his creatinine had settled at 2.5-3.0 mg/dL and hemodialysis was discontinued.

**Discussion:** Renal disease associated with SEP is exceptionally rare; there are only 3 cases reported in the literature. In one of these cases the tumor originated from the urethelium and was causing local obstruction, in the second one a retroperitoneal mass was causing urethral compression, in the third case light chain proximal tubulopathy (LCPT) was responsible for the renal dysfunction. LCPT represents 5% of paraproteinemia-related kidney disease. LCPT is further classified into two forms; the crystalline type represents 87% of cases, the non-crystalline type 13%. It is thought that free light chains pass through the glomerular basement membranes and undergo endocytosis by the proximal tubular epithelial cells. Following endocytosis the variable region of the light chain might resist degradation, accumulate within the lysosomes and undergo spontaneous crystallization, or it might possess resistance to degradation by lysosomal enzymes but lack the ability to spontaneously crystallize. Tubular casts are usually absent, this might be explained by the fact that these free light chains have a relatively low affinity for Tamm-Horsfall protein7,9,10 which results in a lower propensity to form casts in the distal tubules. Treatment of LCPT is directed at eliminating the responsible B-cell clone. In cases of plasmacytoma chemotherapy alone or in combination with radiotherapy is highly effective. Kidney function stabilizes or improves in most treated patients. Independent predictors of final eGFR include the initial eGFR and percent interstitial fibrosis highlighting the importance of early detection.

**Conclusion:** LCPT is a rare and challenging diagnosis that should be suspected in cases of dysproteinemias as treatment of the underlying plasma cell disorder results in significant renal recovery.
MALIGNANT MESODERMAL TUMOR: A RARE GYNECOLOGICAL TUMOR OF MESENCHYMAAL AND EPITHEIAL TISSUE

Introduction: Malignant mixed mesodermal tumors are rare gynecological malignancies which typically present in elderly, postmenopausal women and account for ≤2% of gynecologic cancers. They are composed of both carcinomatous and sarcomatous tissue and can originate anywhere in the female genital tract (ovaries, fallopian tubes, uterus, vagina) and, rarely, extragenital sites including the peritoneum and gastrointestinal tract. Radical surgery may be curative but there is a poor prognosis and average 5 year survival is 26-34%. Metastasis most commonly occurs in the abdominal wall, lung, and bone. We report a case of a 69 year old African American female with a history significant for a pelvic mass and evidence of metastatic disease to the colon, lung, and lymph nodes in multiple locations throughout the body.

Case Presentation: A 69 year old African American female presented to GI clinic with complaint of new-onset diarrhea, nausea, and vomiting. These symptoms led to a loss of appetite and subsequent 30 lb unintentional weight loss. Diagnostic colonoscopy discovered a submucosal mass in the terminal ileum causing partial luminal compression. Small bowel biopsies was consistent with poorly differentiated adenocarcinoma.

Discussion: Ovarian cancer has the highest mortality rate of the gynecologic cancers and is the fifth most common cancer in women worldwide. Malignant mixed mesodermal tumors (MMMTs) account for ≤2% of gynecologic malignancies, and those originating in the ovaries account for <1%. MMMTs are growths containing both malignant mesenchymal and epithelial tissues. They can be further subdivided by their sarcomatous component: Mullerian system tissue is found in homologous tumors, and non-Mullerian tissue (bone, adipose, striated muscle) is found in heterologous tumors. Primary ovarian MMMTs often present in later stages and have a particularly poor prognosis, with median survival of less than two years. Due to the low prevalence of the disease, optimal treatment regimens are controversial although aggressive surgical resection is often favored; the role of chemotherapy and radiation therapy remains unclear.

Unique to our case is the direct involvement of the tumor to the terminal ileum. This led to an initial incorrect diagnosis of adenocarcinoma. Since our patient had MMMT which contained both mesenchymal and epithelial tissues, the superficial nature of small bowel biopsy during colonoscopy missed deeper mesenchymal component needed for the right diagnosis. Furthermore, tumor invasion to the terminal ileum likely caused disruption of the enterohepatic circulation of bile salts. Excess deposition of bile acid into the colon resulted in post-prandial diarrhea which was one of the presenting symptoms by the patient.

Conclusion: Although rare, MMMTs are serious malignancies with poor prognosis and limited treatment options. As primary, aggressive surgical resection is currently the best treatment option, it is of utmost importance to catch these tumors as early as possible.
THE BEAT GOES ON: Atrial Tachycardias after Fontan Procedure
The Fontan procedure is a surgical treatment for congenital heart diseases
with single-ventricle physiology, such as tricuspid atresia and hypoplastolic left
heart syndrome. With Fontan procedure, systemic venous blood is directed
to the lungs without passing through a ventricle. These procedures have allowed
increasing numbers of patients with congenital heart disease to survive into
adulthood. Twenty year survival now exceeds 60%. It is thus increasingly
important for internists to be familiar with long-term complications of Fontan
procedures.

Our patient is a 29 year-old male with a history of tricuspid atresia, palliated
with a Classic Fontan (direct surgical connection between the right atrium and
pulmonary arteries) at age three, who presented to the emergency department
with palpitations. Mr. T had a history of intraatrial tachycardia (a form of
macoreentrant tachycardia in the atria) beginning 1.5 years prior to
presentation, which was treated with anticoagulation and electrical
cardioversion. His arrhythmia recurred shortly thereafter and was treated with
dofetilide, which suppressed subsequent arrhythmias after one dose. Nine
months prior to presentation, the patient terminated his dofetilide due cost,
and developed intermittent palpitations. He remained on anticoagulation with
rivaroxaban. On the day prior to presentation, he developed palpitations while
at rest, which persisted for over 12 hours, prompting his presentation to the
emergency department. Physical exam revealed a non-pulsatile, distended
jugular veins. Cardiac exam showed an irregular tachycardia at a rate of
around 130, and a single S2. EKG showed atypical atrial flutter with variable
(2:1 - 3:1) block and right atrial enlargement with so-called Himalayan p-
waves. Transthoracic and transesophageal echocardiograms were performed
and showed Fontan anatomy with a markedly enlarged right atrium filled with
near-stagnant blood flow and dense spontaneous echocardiographic contrast
(smoke). A thrombus could not be excluded. Given concern for potential
thrombus and increasing risk for thrombus formation with ongoing
arrhythmia, anticoagulation was escalated to warfarin with a goal of 2.5-3.5
and the addition of aspirin 81mg. Dofetilide was reinitiated with close QTe
monitoring. Mr. T chemically cardioverted after one dose. His recurrent
arrhythmia were felt to represent Fontan failure. To prolong Fontan function
and reduce the burden of arrhythmias, Mr. T was encouraged to pursue a
Fontan conversion (use of a graft to directly connect the IVC to the
pulmonary artery, bypassing the right atrium) with MAZE procedure. Mr. T
declined reoperation due to concern about operative risks. Screening for
additional complications of Fontan anatomy included spot stool alpha-1
antitrypsin testing for protein losing enteropathy and liver elastography for
cirrhosis.

This case demonstrates a common, long-term complication of Fontan
procedures and challenges in its management. 40-50% of patients who had
Fontan procedures performed 30 years ago are alive today, however 50% of
these patients have severely impaired functional capacity. As the right atrium
dilates, it becomes a source of atrial tachyarrhythmias. Slow blood
flow through the circuit becomes a source of thrombus. Atrial
tachycardias have a particularly strong association with poor
outcomes after Fontan, and when recurrent, are a marker of Fontan
failure. Fontan conversion is indicated to offload the right atrium and
MAZE procedures to reduce arrhythmias. Anticoagulation is crucial to
long-term management, but TEEs are required to assess for
intracardiac thrombus as this can form despite therapeutic
anticoagulation. Extracardiac complications include cirrhosis, protein
losing enteropathy, recurrent pleural effusions, and require treatment
by multidisciplinary providers. Internist caring for adults with
congenital heart disease should be alert to these long-term
complications.
CELLULITIS WITH CONCURRENT AUTOECZAMITIZATION
Ramachandra P Bongu, MD. PGY-1 Resident, Department of Medicine
GBMC Baltimore MD.

Introduction: 'ID' Reaction are secondary immunological reactions to microbial antigens. It can be triggered by fungal, bacterial, viral and parasitic infections. Superficial fungal infections are the most common cause of id reactions. They are only a few reported cases of bacterial infections causing id reactions.

Case Report:
41-year-old man with history of reactive airway disease presented to the emergency room with 10-day history of left leg pain redness, swelling with scaling and crusting of the skin. This was followed by pruritic rash on his entire body sparing the palm, soles and face. Rash spared the mucosal surfaces. The patient did not report any previously known allergies or recent exposure to drugs, poison ivy or occupational exposure.

Physical exam revealed 4+ tense edema extending to the knee with erythema and extensive crusted erosions. Symmetrically distributed dense monomorphic papular eruptions with circular scales were noted on thighs, abdomen, chest, neck, back and arms.

Treatment was initiated with broad-spectrum antibiotics Vancomycin, Piperacillin-Tazobactam and anti-histaminic diphenhydramine. Expert opinion from dermatologist provided more insight into the clinical condition suggesting two different dermatologic pathologies. It was hypothesized that cellulitis of the left leg from an infectious etiology led to a concurrent hypersensitivity reaction manifested as popular eruptions. Prednisone was started and punch biopsies of the skin lesion revealed interface dermatitis most likely consistent with a ID reaction. Following treatment the patient's condition improved and he was discharged on a prednisone taper and Trimethoprim and Sulfamethoxazole.

This case illustrates the importance of recognition of two different dermatoses occurring concurrently that require different treatment modalities.

First Author Information:
Name: RAMACHANDRA P BONGU
Institution: Greater Baltimore Medical Center
Daytime Phone: 443-849-3760

Co-Author(s) Associates:
Racheal Moss

Program Director's Name: Paul Foster MD

(indicating review of abstract)
Title: Intralobar Bronchopulmonary Sequestration with Unusual Presentation of Recurrent Abdominal Pain

INTRODUCTION Bronchopulmonary sequestration (BPS) is a rare congenital malformation characterized by non-functioning lung tissue that does not communicate with the bronchial tree or pulmonary arteries, but typically has its own arterial supply arising from the aorta. It can be divided into extralobar sequestration (ELS), which is outside of the lung, has its own visceral pleura, and manifests earlier in life; and intralobar sequestration (ILS), which is within the pulmonary lobe, shares a visceral pleura, and often manifests later in life. Usually BPS in patients is unidentified unless infected or diagnosed incidentally on imaging. When symptomatic, patients can present with fever, cough, chest pain or hemoptysis. Here, we report a rare case of a young male who presented with recurrent left upper quadrant (LUQ) pain, diagnosed with ILS.

CASE PRESENTATION A 29 year old male with no significant history presented to ED with recurrent LUQ pain. He had 2 prior ED visits spanning 6 months where no specific diagnosis was made. He denied chest pain, cough, and fever. Vital signs, physical exam, and labs including CBC, LFTs and lipase were unremarkable. Chest x-ray and EKG were within normal limits. On the third ED visit, CT abdomen was performed. This showed no abnormalities in the abdomen but a mass in the left lower lobe, concerning for infection, atelectasis, congenital malformation, or malignancy. Further evaluation showed a 6x4x7cm mass supplied by an aberrant artery off the descending thoracic aorta. Patient was referred to thoracic surgery, had a pre-operative angiogram and embolization of the feeding artery, and underwent left thoracotomy with complete resection of BPS, which resolved his abdominal pain. Pathology confirmed the diagnosis.

DISCUSSION BPS is very rare, comprising only 0.15-6.4% of all pulmonary malformations. It is typically within the lower lobe, on the left side, involving the posterior basal segment. However, clinical presentation can vary depending on its type, size, and location, and sometimes presentation can be unusual, making it difficult to diagnose, as seen in our case where inflamed BPS caused diaphragmatic irritation resulting in referred abdominal pain.

CONCLUSIONS We report a rare case of ILS presenting with recurrent abdominal pain later in life. Screening for thoracic etiologies should be considered when no obvious abdominal cause is found to explain for the presenting abdominal pain.
2017 Mulholland Mohler Resident Meeting

Trimethoprim-sulfamethoxazole induced DRESS Syndrome
Siddharth Patel MD, Shyam Kolangara MD, Joshua Birnbaum MD, Sinai Hospital of Baltimore

Introduction:
Trimethoprim-sulfamethoxazole is a routinely used antibiotic for various types of bacterial infection. Skin rash is a frequently reported adverse drug reaction with it. However, a systemic allergic reaction with multi-organ involvement is rare.

Case:
A 64-year old Caucasian man came in with complaints of diffuse morbilliform rash, fever, chills and several episodes of non-bloody diarrhea for 3 days. Three months ago, the patient had an open displaced fracture of the ankle after a mechanical fall which was then managed with open reduction and internal fixation. However, 2 months after the initial surgery he developed a polymicrobial wound infection. Incision and drainage of the wound was performed and he was discharged home on PO Trimethoprim-sulfamethoxazole and IV Piperacillin-Tazobactam. He had also been on subcutaneous enoxaparin since then for DVT prophylaxis.

On admission, the patient was febrile (38.3-38.7°C), tachycardic (110/min), normotensive (130/70 mmHg) and breathing well on room air. Physical exam revealed diffuse, non-blanching, non-itchy, morbilliform macular rash involving entire body except palms, soles and face. Laboratory investigations were remarkable for acute kidney injury, leukopenia, thrombocytopenia, eosinophilia, transaminitis, elevated ESR and elevated CRP. Sepsis from wound infection/abscess, allergic drug reaction, drug induced lupus or eosinophilic vasculitis were some of the possible differential diagnosis considered at that time. He was treated with intravenous fluids, his home antibiotics were stopped, blood cultures were drawn and he was started on Aztreonam, Vancomycin and Metronidazole empirically for suspected sepsis from the wound. Oral prednisone was also added to the regimen for suspected allergic reaction to his home medications. On day 2, he started feeling better and his rash improved. He was afebrile and not tachycardic. His kidney function started to recover. His leukopenia and thrombocytopenia improved and his eosinophilia trended down. Blood cultures remained negative and his antibiotics were discontinued. Work-up for heparin induced thrombocytopenia, anti-nuclear antibodies and acute viral hepatitis returned normal. A final diagnosis of drug reaction with eosinophilia and systemic symptoms (DRESS syndrome) was made. The patient was discharged home with a 5-day course of prednisone.

Case discussion:
DRESS syndrome is a rare, potentially life-threatening drug induced hypersensitivity reaction that manifests as a skin rash, eosinophilia, lymphadenopathy, and/or internal organ involvement. Anti-epileptics, sulfonamides, Dapsone, allopurinol etc. are the most common drugs associated with it. In this case, Trimethoprim-sulfamethoxazole was the most likely medication responsible. In most patients, the reaction occurs within two to six weeks after the initiation of the offending medication. Kidney, liver and lungs are commonly involved. Diagnosis is based on cutaneous findings, clinical features and laboratory abnormalities. Discontinuation of the offending drug is the mainstay of treatment.
Sequela of Untreated Basal Cell Carcinoma
Michael Adugna, M.D., Keval Patel MD, Aksa Khan, Ammer Bekele M.D.

Introduction: Basal cell carcinoma (BCC) is the most common malignancy in whites, it accounts for 70-80% of all cutaneous cancers. BCC has a very low rate of metastasis ranging from 0.0028%-0.54%; however, it tends to be locally invasive, aggressive, and destructive. We hereby present a case of long standing, untreated, highly destructive and locally invasive BCC.

Case description: A 90-year-old White male was referred from an outside facility for evaluation of new onset atrial fibrillation and a very elevated white blood cell count (40,700 cells/µL). His only symptoms were fatigue and decreased energy. Medical history was significant for unspecified skin cancer of the face and anterior chest wall which he reported having since 1984. He had undergone Mohs surgical excision and multiple follow up excisions through 1994 for recurrent lesions. He was lost to follow up as he felt surgery was not helpful. He had been caring for these wounds at home with local cleansing and hydrogen peroxide. Physical examination was significant for a large locally erode lesion over the right maxilla, triangular in shape, with irregular borders, measuring 3×5 cm, with superficial necrosis, extending onto the lateral canthus. Additionally, a full thickness ulcer with necrosis, purulent and foul smelling discharge and fibrinous exudate was found over the upper third of the sternum, measuring 9×10×4 cm; the manubrium, sternal body and ribs were exposed. Laboratory investigations: WBC 31,800 cells/µL, 87% neutrophils, CRP 322 mg/L, alkaline phosphatase 161 U/L. Chest computed tomography (CT) revealed a deep midline chest wound with comminuted displaced fractures of the manubrium and sternum, a mediastinal abscess, and a large right pleural effusion. Pleural fluid analysis was unremarkable. The patient underwent local debridement and removal of a segment of the sternal body and adjacent tissues, pathology findings were consistent with osteomyelitis and locally invasive BCC. Chest wound cultures isolated MRSA and Pseudomonas sp. Osteomyelitis was initially treated with vancomycin and piperacillin-tazobactam but later on transitioned to oral doxycycline and levofloxacin. Facial lesion was thought to represent a synchronous primary BCC; biopsy was not pursued here but records from surgery back in the 1994 obtained later on confirmed this was so. Serial debridement with subsequent combination chemotherapy and radiation was offered; however, due to the complexity and intensity of the treatment the patient and family opted not to pursue aggressive measures and resorted to hospice care.

Discussion: BCC is a common surface malignancy which makes it very amenable to early detection. Sun exposure seems to be the most significant risk factor which explains why BCC occurs most commonly on the face and neck (70-90%). About 15% of cases occur on the trunk; however, invasive chest wall BCC has rarely been reported. Early recognition and treatment result in cure in the majority of patients (cure rates are >95%). Patients who present late are challenging as treatment is often more complicated and less likely to be effective. The treatment of choice is usually surgical excision; however, the risk of recurrence partially depends on the quality of the surgery (clean margins). Other treatment options include topical chemotherapy or immunomodulation (for minor, low-risk lesions) and radiation. A relatively new medication vismodegib, a Hedgehog pathway inhibitor, has been used to treat advanced basal cell carcinoma when both surgery and radiation are not viable options.

Conclusions: BCC can present as an indolent malignancy with significant morbidity when not treated and followed up appropriately as demonstrated by this case.
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First Author Information:

Name: Ann Miller, MD

Institution: University of Maryland Medical Center and Baltimore VA Medical Center

Daytime Phone: 410-328-7567

Co-Author(s) Associates: Rana Malek, MD

Program Director's Name: Susan D. Wolfsthal

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THE ROLE OF CT ANGIOGRAPHY IN DIAGNOSING A RARE CASE OF HYPER-EMBOLIC STATE

A 61-year old woman with hypertension and remote history of provoked deep venous thrombosis with pulmonary embolism presented to an outside facility with a cold, painful right lower extremity. One month prior, she was diagnosed with an infra-renal aortic thrombus with arterial emboli to both lower extremities, managed with thrombolysis and alteplase infusion. She was discharged on warfarin anticoagulation, and her bilateral lower extremity pain had been improving until the day prior to this presentation.

Physical examination revealed the absence of right dorsal pedal and posterior tibial pulses; pulses were also undetectable on Doppler ultrasound. Ankle brachial index in the right lower extremity was 0.2. She underwent angiography and embolectomy in the R external iliac artery, resulting in improvement of her symptoms. On post-operative day 2, however, she developed nausea and vomiting. A computed tomography angiogram (CTA) of the abdomen revealed a new embolus in the superior mesenteric artery. Upon radiologic review of the aortic mass, primary angiosarcoma of the aorta was suspected as the source for these emboli, accompanied by an adrenal mass that was suspected to be a metastasis. Pathologic examination of the right lower extremity emboli confirmed the diagnosis of aortic intimal sarcoma, and the sarcoma-containing segment of the descending aorta was ultimately excised and replaced with an interposition graft. After a hospital course involving multiple operative interventions, the patient was discharged to a rehab facility with plans for systemic chemotherapy.

Careful consideration of this patient’s clinical course should have prompted reassessment of the initial diagnosis of aortic thrombus, especially given the progression of the aortic mass despite adequate anticoagulation. In addition, the patient had a negative hypercoagulability work-up and minimal atherosclerotic plaque elsewhere in the vasculature. Finally, certain features of her imaging are hallmarks of aortic sarcoma – a mass rapidly growing from the wall of the descending aorta with necrosis and a frond-like polypoid appearance. The adrenal mass on the abdominal imaging also raises suspicion for metastasis of a sarcoma. This case illustrates classic radiographic features of this rare tumor and the importance of maintaining a broad differential, especially for patients with a complicated clinical course.

First Author Information:

Name: Brian Burkett MD
Institution: University of Maryland Medical Center - Midtown Campus
Daytime Phone: 504-701-6316
Co-Author(s) Associates: Michael Morris, MD, Babak Saboury, MD
University of Maryland Medical Center

Program Director’s Name: R. Dobbin Chow, MD MBA MACP

(indicating review of abstract)
61 year old female with history of sacral osteomyelitis, left hip contracture, DVT s/p anticoagulation, cognitive impairment, who presented with 2 week history of diffuse body pruritis followed by blistering eruption. Rash initially was on bilateral lower extremities before spreading to all body areas. No new medication or contact exposures. She was seen by PMD and prescribed Benadryl for symptomatic relief, but given progression of her rash, her caretaker brought her to the hospital. On exam she was tachycardic, hypotensive, and appeared very malnourished. She had diffuse excoriations with erythematous base, a few flaccid bullae on her right hand and left foot, small vesicles on her upper lip, as well as scaly plaques on her feet. Total body surface area involved was about 80%, with <15% open wound. There was no oral or genital involvement. She was seen by dermatology and skin biopsy was performed. Initial labs notable for lactic acidosis of 8 mg/dl and she was admitted to MICU for distributive shock. She was given IVF and started on broad spectrum antibiotics and acyclovir for possible HSV superinfection. She was also started on high dose IV steroids for bullous pemphigoid, which was later confirmed by direct immunofluorescence on the biopsy, and her whole body was covered with Acticoat (antimicrobial barrier dressing). Peripheral blood cultures were positive for Klebsiella pneumoniae, methicillin-susceptible S. aureus (MSSA), and group B streptococcus. Skin biopsy culture showed Proteus mirabilis, MSSA, group B streptococcus, and corynebacterium species. She was also HSV-1 positive on the skin biopsy and in her serum. A TTE was attempted but was unable to adequately visualize the valves. She had an endoscopically placed nasogastric tube for nutrition. On the EGD, she was found to have severe erosive and candida esophagitis and was started on fluconazole. Despite IV steroids, her lesions did not markedly improve. Given concurrent sepsis, IVIG was added on day 6 with plan to transition to Rituximab once acute illness resolves. Follow-up blood cultures were negative but she remained hypotensive, eventually requiring norepinephrine. Her mental status declined with the addition of pain medication for her wounds, and she was intubated for hypercarbic respiratory failure. After failure to wean from the vent and likely poor functional status outcome, family discussion was held and she was transitioned to comfort care and terminally extubated on hospital day 14.

Bullous pemphigoid is generally thought to be a chronic inflammatory blistering disease caused by auto-antibodies to components of skin basement membrane. However, this disease can be fatal in the debilitated and chronically ill. This case delineates the severe complications from refractory bullous pemphigoid and the importance of assessing and treating infections in the setting of massive skin denudation.