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**2014
Abstracts**

**Clinical Vignette and Research
Competition for Associates**

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**59th Annual Wisconsin Scientific Meeting
Wilderness Resort, Wisconsin Dells, Wisconsin**

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Case Based Vignettes

A RARE CASE OF POST-INFARCT VENTRAL SEPTAL DEFECT WITH VENTRICULAR PSEUDOANEURYSM

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Introduction: Immediate mechanical complications of myocardial infarction (MI) include ventricular septal defect (VSD), papillary muscle rupture, and ventricular free wall rupture. Concomitant occurrence of VSD with free wall rupture, which gets sealed off by the pericardium forming a pseudoaneurysm, is an extremely rare and life-threatening complication of MI.

Case Description: A 67 year-old female with end stage renal disease on hemodialysis was admitted for evaluation of hypotension and dyspnea. Eight weeks prior, she had a NSTEMI with subsequent nuclear stress test showing no reversible ischemia; she was managed medically.

Review of systems was unremarkable, except for worsening lower limb edema. Physical examination was significant for a blood pressure of 71/54 mmHg and 3+ leg edema. Cardiac exam was notable for a pansystolic murmur. Chest x-ray revealed a moderate right pleural effusion.

Echocardiogram demonstrated severely reduced right heart function, markedly different from her previous echocardiogram. Chest CT, performed to rule out pulmonary embolism, revealed a focal outpouching along the inferior interventricular septum with possible communication between ventricular cavities, suspicious for VSD associated with an aneurysm.

TEE and MRI confirmed the diagnosis. Coronary angiography revealed 100% occlusion of the RCA, supporting the post-infarct etiology. Given the patient's comorbidities, a percutaneous approach of VSD occlusion was adopted. The patient had an uneventful post-intervention hospital course.

Discussion: Ventricular free wall rupture is a serious complication of MI. Rupture of the inferior wall contained by the pericardium forms a pseudoaneurysm. Patients can be asymptomatic or present with chest pain and hypotension. A high index of suspicion is necessary for early diagnosis. Associated with VSD, it can lead to significant hemodynamic instability and cardiogenic shock. When feasible, a surgical approach is preferable and can be life-saving.

SERONEGATIVE GRANULOMATOSIS WITH POLYANGIITIS PRESENTING AS A LUNG MASS

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Introduction: Granulomatosis with polyangiitis (GPA) is a vasculitis with systemic manifestations mainly in respiratory and renal systems. Rarely it involves other systems, e.g. nervous, cardiovascular and genitourinary systems.

Case Presentation: 49 yo male presented with sinus discharge and intermittent headache for one year. He was initially treated with oral antibiotics without improvement and subsequently developed epistaxis. MRI of the head showed faint enhancement of the extracranial right mastoid facial nerve and paranasal sinus mucosal disease. Upper airway examination revealed significant sinonasal inflammation with scarring out of proportion to regular sinus infection. Two months later he began to cough. Imaging studies revealed consolidation in the right upper lobe with a possible mass. He was treated for pneumonia with Ceftriaxone. Follow-up CT scan revealed new cavities formation. Myeloperoxidase-antinuclear cytoplasmic antibodies (MPO-ANCA) and proteinase 3- antinuclear cytoplasmic antibodies (PR3-ANCA) were both negative. Kidney function and urinalysis were normal. Bronchoscopy with transbronchial and endobronchial biopsies were not diagnostic. Right upper and middle lobectomies with lymph node sampling showed necrotizing granulomatous inflammation with geographic necrosis and vasculitis which was consistent with GPA. He was started on high dose corticosteroids and received two cycles of rituximab with significant improvement of his respiratory symptoms. He is currently being weaned of corticosteroids and transitioned to azathioprine.

Discussion: This case demonstrates the importance of considering GPA in patients with recurrent sinus symptoms combined with lower respiratory tract symptoms in spite of negative serologies. Early diagnosis and treatment is important to prevent mortality and organ loss. MPO-ANCA and PR3-ANA antibodies are positive in 82%-94% of the patients. 10% of patients with GPA can be seronegative. The diagnosis must be confirmed with biopsy, which can be obtained from the target organ.

A RARE CAUSE OF SEIZURES

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Introduction: Anaplasmosis is rarely associated with neurologic manifestations other than confusion. Rarely, meningoencephalitis has been associated in immunocompromised hosts. Anaplasmosis-associated seizures have not been reported. Described below is a gentleman afflicted with seizures after infection with *Anaplasma phagocytophilum*.

Case Description: An 81 year old male from northern Wisconsin presented to his primary care provider with progressively worsening generalized myalgias and weakness with increasing frequency of low-energy falls of three weeks duration. These were accompanied by fevers, chills, anorexia, headache and lightheadedness during the week prior to presentation. Two weeks prior to presentation, he was bitten by a tick and was unable to remove all of it until he was seen by dermatology the next day. He did not develop a rash. Physical exam revealed an afebrile, pleasant, ill-appearing man and no other abnormalities. Initial lab studies revealed leukopenia, thrombocytopenia, and borderline elevated AST and ALT. CRP was 5.0, ESR was 8. CMV and EBV IgM antibodies were negative. Lyme serology was negative. PCR detection was positive for *Anaplasma phagocytophilum* DNA and negative for *Babesia* and *Ehrlichia* species. EKG and chest X-ray were normal. He was empirically treated with doxycycline. He developed a fever of 103 on hospital day two but was otherwise stable. His symptoms improved and discharge was planned. On hospital day four, he developed acute confusion without other focal neurologic deficits. He developed a tonic-clonic seizure of one minute in duration while in CT scan. CT Head was normal. He was placed on continuous EEG monitoring and started on levetiracetam. Brain MRI was normal. Lumbar puncture was unremarkable and PCR was negative for Lyme, Arbovirus, West Nile virus, HSV, and VZV. He continued to have intermittent epileptiform activity until he was switched to divalproex sodium and his condition improved. He was discharged home with doxycycline and divalproex sodium. He had no seizure recurrence and his antiepileptic was weaned off.

INTRAVASCULAR LARGE B-CELL LYMPHOMA IN A CAUCASIAN FEMALE WITH PROFOUND THROMBOCYTOPENIA AND DYSPNEA

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Introduction: Intravascular Large B-Cell Lymphoma (IVLBCL) is an extremely rare form of non-Hodgkin's lymphoma in which malignant cells have a tropism for small and medium-sized blood vessels. In this case, IVLBCL was diagnosed in a 78 year old Caucasian woman who presented with severe thrombocytopenia, anemia, and progressive shortness of breath.

Case: The patient, a nonsmoker with a history of Sjögren's syndrome presented three months prior to admission with SOB and was told she had interstitial pulmonary fibrosis. She was treated with a prednisone taper. She returned to clinic one week prior to admission with bruising, dyspnea, and a platelet count of 8 K/ μ L. She received prednisone and gamma-globulin for presumed immune thrombocytopenia without improvement.

On admission, the patient was afebrile. Physical findings included bibasilar lung crackles, splenomegaly without adenopathy, and diffuse ecchymoses and petechiae. Labs showed a white blood cell count of 7.8 K/ μ L, hemoglobin 9.9 g/dL, and a platelet count of 3 K/ μ L. The patient's creatinine was 1.40 mg/dL, serum LDH was 1810 U/L (normal < 245 U/L), and albumin was 1.9 g/dL. The blood smear showed thrombocytopenia without schistocytes. A bone marrow aspirate showed hemophagocytosis, while biopsy demonstrated monoclonal infiltrates of CD20+ lymphoid cells confined to vascular spaces, consistent with IVLBCL.

The patient was started on R-CHOP. She required daily blood products, but after two weeks was able to maintain a platelet count of greater than 20 K/ μ L unsupported.

Discussion: IVLBCL is notoriously difficult to diagnose, with more than one half of cases proven at autopsy. Definitive diagnosis requires finding malignant cells in the vessels of affected organs. The disease is usually disseminated and has a variable presentation, often with non-specific complaints, but frequently with elevated LDH and anemia. Western patients typically present with cutaneous and neurologic abnormalities. By contrast, Asian patients are more likely to have pulmonary symptoms and bone marrow involvement with thrombocytopenia and a hemophagocytic syndrome. In this case, a Caucasian patient presented with an Asian-variant of IVLBCL. Prognosis is poor, but treatment with R-CHOP has improved overall survival to an estimated 60-80% at 3 years.

H1N1 INFLUENZA A INFECTION AS A CAUSE OF SEVERE PULMONARY COMPLICATIONS

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Introduction: Most patients with H1N1 Influenza A infection have a mild or uncomplicated clinical course. Common complications include secondary bacterial pneumonia, ARDS etc. Other pulmonary complications such as pneumatocele, bronchopleural fistula and pneumothorax are very rare.

Case Presentation: A 59 yo male, vaccinated against flu, presented with cough, fever, chest congestion and respiratory distress. He was found to be hypoxic with bilateral alveolar and interstitial opacities on CXR. Diagnosis of ARDS was made. He was intubated and started on antibiotics. Blood and sputum cultures were negative. Later, Influenza A was detected on BAL. Oseltamivir was started. Follow-up CT-chest showed diffuse and extensive airspace opacification and repeat bronchoscopy was unremarkable. High dose steroids were started for fibroproliferative ARDS. He underwent tracheostomy and was eventually transferred to an LTAC facility. Within a month, he was admitted with shortness of breath and hypoxemia. Imaging revealed bilateral infiltrates, cavitary lesions as well as pneumothorax. He was treated for possible healthcare-associated pneumonia despite negative work-up and was discharged with chest tube and home oxygen. He presented to the hospital again within three days with shortness of breath. CT chest revealed a left bronchopleural fistula with large hydropneumothorax and persistent right pneumothorax. He underwent thoracoscopy and bronchopleural fistula repair. He had an extended hospital stay before being discharged.

Discussion: This case demonstrates the devastating complications associated with H1N1 influenza A. In our patient, the delay in diagnosis and initiation of treatment might have contributed to the development of these complications. There was no evidence of superimposed bacterial infection. Maintaining a high index of suspicion for influenza, early diagnosis and prompt initiation of treatment is paramount in preventing complications and improving outcomes.

MONOCLONAL GAMMOPATHY OF MESENTERIC SIGNIFICANCE

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Introduction: Sclerosing mesenteritis, a fibro-inflammatory condition of unclear pathogenesis, consists of a spectrum of uncommon disorders including mesenteric lipodystrophy and mesenteric panniculitis. Rarely, it has been reported in patients with underlying malignancies commonly of the gastrointestinal and genitourinary tracts but rarely with leukemia/lymphomas. Herein, we present a patient with myeloma-related mesenteritis and highlight the challenges associated with non-specific symptomatology concealing a malignant process.

Case: An 83 year old female with known history of monoclonal gammopathy of undetermined significance (MGUS) and previously treated stage I breast cancer developed persistent abdominal pain with eating and alternating constipation and diarrhea. Computerized tomography (CT) of the abdomen showed mesenteric nodules. Recurrent breast cancer was suspected. A positron emission tomography/CT (PET/CT) scan showed the mesenteric nodules were metabolically quiescent and no other evidence of metastatic disease leading to the conclusion that the mesenteric changes were non-specific. The patient continued to have symptoms for 1 year and underwent multiple endoscopies and physician visits without explanation of etiology or symptomatic benefit. She was empirically treated with prednisone 20 mg/day without relief. Subsequently, she developed bone pain in addition to her abdominal symptoms, which led to a hospitalization at our center. A PET/CT scan was repeated owing to the high suspicion for an underlying malignancy and this revealed bony lytic lesions in addition to the mesenteric nodules. Further hematology-directed work up revealed low immunoglobulin levels, however her kappa light chains of 2089mg/L (3.3 – 19.4 mg/L) and a free light chain (FLC) ratio of 289 (0.26-1.65). It was noted that her previous MGUS follow up evaluation 6 months ago had shown that her FLC ratio had increased to 20.9 from a previous stable level of 9. A bone marrow biopsy confirmed multiple myeloma. She was started on treatment with lenalidomide, bortezomib and dexamethasone with prompt resolution of her abdominal symptoms.

Discussion: Sclerosing mesenteritis includes a spectrum of disorders that lead to inflammation and fibrosis of the mesentery. Rarely, it has been associated with hematologic malignancies either as a paraneoplastic syndrome or as direct infiltration by malignant cells. In our patient, the MGUS was a clue to an underlying hematologic malignancy; the fact that it was changing as observed by an increasingly abnormal FLC ratio was a clue that it had evolved into myeloma. This case highlights the need for awareness of the paraneoplastic nature of sclerosing mesenteritis and suspicion of myeloma evolution from MGUS.

HEMOLYTIC UREMIC SYNDROME (HUS) IN AN ADULT CHEMOTHERAPY PATIENT

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HUS is often overlooked as a diagnosis in adults. Additionally, because many chemotherapy regimens are associated with diarrhea, infectious etiologies of diarrhea in cancer patients can be easily missed. The time sensitive nature of starting plasmapheresis to avoid the potentially irreversible renal damage of microangiopathic hemolytic anemia (MAHA) makes this an important diagnosis to consider, even in less classic circumstances.

A 62 year old female on dasatinib for CML developed watery, non-bloody diarrhea, and non-bilious emesis without fevers during a trip to Missouri. No other family members had gastrointestinal symptoms. Her symptoms persisted for the following 2-3 weeks, despite a trial of loperamide and ondansetron. Upon return to Wisconsin, she was instructed to stop dasatinib and required IVF for dehydration. Her labs were notable for BUN 38 and Cr2.3 thought pre-renal from volume loss. Over the next 48 hours, however, she developed worsening confusion and peri-orbital edema. Admitted to the hospital, physical exam was notable for BP 159/87, pale sclera, asterixis and petechial rash on her lower back and sacrum. Labs were notable for Hgb 7.8, Plt 85, BUN 49, CR 4.82, UA with 3+ protein and FeNa 1.34 suggestive of intra-renal pathology. Peripheral smear with >5 schistocytes/hpf and renal biopsy showing thrombotic microangiopathy confirmed MAHA. Stool was negative for shigella, O157:H7, shiga toxin, however, further inquiry of outside hospital workup revealed positive stool shigella and shiga toxin. Missouri department of public health was notified. Upon urgent initiation of plasmapheresis and IVIG she improved dramatically over the following six days with normalization of cell lines and renal function.

This case illustrates the importance of re-evaluating the cause of worsening renal function when not improving after fluid resuscitation, and remembering MAHA and HUS when presented with a history of gastrointestinal symptoms - even in adults, as early recognition and treatment are critical to maximizing renal recovery.

HEROIN AND RHABDOMYOLYSIS

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We present a case of a 23 year old male who was found unconscious by friends wedged between a bed and a night stand, having been there for an unknown period of time. He was given naloxone and became transiently more responsive. Initial evaluation revealed renal failure, hyperkalemia, lactic acidosis, CPK > 250,000 and absent bilateral pedal pulses. Bilateral lower extremity fasciotomies were performed. Renal dialysis was started shortly after admission, but his metabolic acidosis and hyperkalemia continued to worsen. Bilateral above knee amputations were performed with subsequent resolution of his metabolic abnormalities. Upon waking the patient admitted long standing history heroin and poly-substance abuse.

Heroin is one of the most harmful drugs available and causes physical, emotional and monetary burdens on both patients and society. Beyond respiratory depression and addiction, it poses many different organic problems when overdosed. In addition, narcotic addiction is a problem now reaching a younger population. Among 12-13 year olds controlled prescription drugs are now the most commonly abused. In 2012, it was estimated that 10 million individuals between ages 12-29 needed treatment for drug addiction. Providers must become more judicious in their dispensing of addictive prescription medications knowing that they are often gateway drugs.

Rhabdomyolysis is a potentially life threatening condition with complications including renal failure, compartment syndrome and fatal arrhythmias. It must be suspected in any patient found down for an unknown period of time. The first line treatment of preventing these complications is heavy fluid resuscitation and frequent neurovascular checks. Although myoglobin and its byproduct are major contributors to renal failure, the pathophysiology is much more complicated and is usually preceded by pre-renal azotemia and hyper-uricemia. In a large retrospective study of patients with rhabdomyolysis, it was found that patients with pre-renal azotemia were more likely to develop renal failure versus the patients who were well hydrated.

Narcotic abuse is becoming a wide spread problem taking a huge physical toll and resulting in a large economic burden as well. In Wisconsin there have been significant recent legislative efforts with the addition of resources to prevent narcotic overdose, however more needs to be done to prevent the abuse of narcotics. It requires both increased public awareness and also provider awareness across all specialties.

EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (CHURG STRAUSS DISEASE)

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Introduction: Eosinophilic granulomatosis with polyangiitis (EGPA), previously known as Churg Strauss Disease, is a multi-organ disease usually affecting the lungs and skin. Patients typically present with chronic rhinosinusitis, asthma and eosinophilia. While classically considered to be an ANCA-positive vasculitis, the sensitivity and specificity of ANCA activity level is low.

Case: 62 year old female with chronic sinusitis and asthma presented with progressive shortness of breath which worsened over the past month and nodules on her fingers which developed three weeks prior to presentation. Outpatient use of Advair, albuterol and Spiriva, azithromycin and steroids provided minimal relief of her symptoms. Physical exam was significant for bilateral expiratory wheezing and crackles, papulo-vesicular nodules on the extensor surfaces of her elbows, DIP, PIP joints. Her labs were remarkable for WBC count of 12.2 with 30% eosinophils, markedly elevated ESR of 116 mm/hr, CRP of 16.10 mg/dL, IgE level of 470.9 IU/mL, rheumatoid factor of 28 IU/mL. C3 was mildly elevated at 186 mg/dL. C4, total complement level, ANA, c-ANCA, p-ANCA, proteinase 3 antibody, myeloperoxidase antibody, anti-CCP antibody, ACE level, urine histoplasma, blastomyces and legionella antigen were all negative. CT chest with contrast was notable for diffuse thickening of lung parenchyma and bronchial wall, hilar and mediastinal lymphadenopathy. To aid in diagnosis, a skin biopsy of the patient's papulo-vesicular nodules was performed and found erythema elevatum diutinum, a rare and poorly understood presentation of vasculitis. VATS lung biopsy revealed eosinophilic vasculitis with patchy involvement of medium-sized arteries confirming the diagnosis of EGPA. Bone marrow biopsy showed marked eosinophilia but not an increased percentage of blasts, effectively ruling out a neoplastic process leading to the hypereosinophilia. The patient was started on steroid therapy and then transitioned to a course of rituximab.

Discussion: The differential diagnosis of peripheral eosinophilia with associated skin findings included a vasculitic process such as eosinophilic granulomatosis with polyangiitis (EGPA) or sarcoidosis, neoplastic disease such as lymphoma, immunologic disease such as hyper-eosinophilic syndrome, or atypical infection with helminth or parasite. This case describes a typical presentation of a classic vasculitis. p-ANCA is present in only about 50% of patients with EGPA, however, our patient did not have this classical marker. In a patient with chronic sinusitis and asthma presenting with peripheral eosinophilia, clinicians must maintain a high degree of suspicion for EGPA since treatment can significantly reduce morbidity and mortality.

PNEUMOCOCCAL PERICARDITIS WITH TAMPONADE

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Introduction: Pneumococcal pericarditis is a rare entity in the current antibiotic and vaccination era in the US. Here we present a case of pneumococcal pericardial effusion with tamponade.

Case: A 82 YOM with no significant known PMH, was seen for fatigue of approximately a month's duration and dyspnea at rest for a week. He was hypotensive, tachypneic, tachycardic and orthostatic. His exam revealed distant heart sounds, crackles in left lung base and distended neck veins. Labs revealed leukocytosis of 197k, multiorgan dysfunction and spontaneous tumor lysis. An ECG showed diffuse ST segment elevation in the anterolateral leads. Chest xray revealed cardiomegaly with a left retro-cardiac density. ECHO showed a moderate pericardial effusion with pending tamponade physiology and he underwent emergent pericardial window placement. His pericardial fluid grew *s.pneumoniae*. He was also diagnosed with CLL, however, there were no malignant cells in the pericardial fluid. He was treated with 4 weeks of IV antibiotics with resolution of sepsis and multi-organ dysfunction.

Discussion: Bacterial pericarditis (BP) is nowadays very rare occurring mostly in individuals with previous pericardial disease treated by chemotherapy, patients who have undergone cardiac surgery or those receiving dialysis. Risk factors for getting BP are immunosuppression, alcoholism and chest trauma. BP requires prompt recognition as it is life threatening and requires immediate antibiotics and surgical drainage.

Our patient's pericardial effusion was initially thought to be from his CLL until the pathology results came back negative for malignant cells. However, there has been a case report on concurrent *S. pneumoniae* and malignant squamous cells in pericardial fluid. Further, negative cytology does not exclude the diagnosis of malignancy, particularly if there is a high index of suspicion. In this situation, a pericardial biopsy should be considered to confirm or exclude pericardial malignancy.

This case demonstrates the importance of considering pneumococcal pericarditis as the cause of a pericardial effusion in patients with pneumococcal sepsis and immunosuppression. Further diagnostic testing such as pericardiocentesis and culture of pericardial fluid, PCR, or antigen testing with early drainage is vital.

Research Based Vignettes

REPEAT LIPOPOLYSACCHARIDE EXPOSURE IS SUFFICIENT TO IMPAIR VIRAL INDUCED PRO-ATOPIC, CD49D EXPRESSING NEUTROPHIL RECRUITMENT TO THE LUNG

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Background: Severe respiratory viral infections increase the risk of developing asthma and atopic disease. In the Sendai virus (SeV) mouse model, we demonstrated this risk depends upon the early recruitment of CD49d expressing neutrophils to the lung. We also demonstrated that single intranasal (i.n.) dose of lipopolysaccharide (LPS) prior to SeV infection significantly reduced CD49d+ neutrophils in the bronchoalveolar lavage (BAL). The hygiene hypothesis suggests chronic microbial exposure prevents development of atopic disease. Our study investigated whether chronic LPS exposure would reduce SeV mediated CD49d+ neutrophil recruitment to the lung and BAL.

Methods: C57BL6 mice were treated with daily LPS (3 µg) i.n. starting 3 or 1 days before or with SeV infection (day 0). On day 3 post SeV, the BAL and lung were isolated and the frequency of CD49d+ neutrophils determined by flow cytometry.

Results: In the BAL, CD49d+ neutrophils were reduced most significantly when LPS exposure was started one day prior to or the day of SeV infection (23.6±1.8%, 10.2±1.6% [0.0002], 10.5±2.2% [0.0037], 17.4±3.6% [0.11]; mean±sem percent CD49d+ neutrophils [p value versus PBS] for PBS, LPS starting on day 0, -1, or -3; n≥3). In the lung, CD49d+ neutrophils decreased regardless of LPS starting day (42.9±3.7%, 18.7±2.0% [0.0003], 22.6±1.0% [0.013], 24.9±2.0% [0.0052]; n≥3).

Discussion: Chronic LPS exposure reduces SeV mediated CD49d+ neutrophil accumulation in the lung and the BAL, suggesting an interaction between the viral and hygiene hypotheses in driving atopic risk. Future studies will explore whether chronic LPS exposure is sufficient to prevent the development of postviral atopic disease.

THE NOVEL PI3K/AKT/MTOR INHIBITOR PALOMID 529 (P529) CAN INHIBIT HUMAN LUNG FIBROBLAST DIFFERENTIATION IN AN *IN VITRO* MODEL OF IDIOPATHIC PULMONARY FIBROSIS

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Idiopathic Pulmonary Fibrosis (IPF) is a fibroproliferative lung disease with, unfortunately, very few therapeutic options. Investigation was made into whether the PI3K/Akt/mTOR inhibitor Palomid 529 (8-(1-hydroxyethyl)-2-methoxy-3-(4-methylbenzyloxy)-benzo[c]chromen-6-one), which is a target of both mammalian target of rapamycin complex 1 (mTORC-1) and mTORC-2) could inhibit fibrosis in human lung fibroblasts in an in vitro model.

Treatment of human lung fibroblasts after stimulation with TGF β with differing concentrations of P529 led to concentration-dependent reduction of fibrotic proteins in the form of smooth muscle actin, fibronectin, and collagen-1 as well as reduction in the Akt pathway by decreasing the amount of phospho-Akt (pAkt) via Western blotting after 24 hours of treatment. Moreover, there appears to be a reduction in the cofilin pathway with a decrease in the amount of phosph-cofilin (pcofilin) as well. P529 was compared against dimethyl sulfoxide (DMSO) as well as negative control. Luciferase assays were performed to investigate protein transcription. Immunohistochemical methods on human lung fibroblasts investigated the proliferation of human lung fibroblasts with differing concentrations of P529.

These preclinical in vitro observations are promising as the mTOR pathway could be a target for future IPF research and that the PI3K/Akt/mTOR inhibitor P529 should be further explored as a candidate treatment for IPF.

PROVIDERS' EXPERIENCE WITH SEX TRAFFICKING VICTIMS

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Background: Though many healthcare providers come into contact with victims of sex trafficking, very few recognize and identify the victims, resulting in potentially significant health disparities for this vulnerable population. One study found that 28% of victims were seen by a health care provider during the time they were being trafficked. Sex trafficking (ST) victims are a unique subset of patients with specific identification and needs. Our objective was to evaluate knowledge gaps and training needs of medical providers, importance of training for a victim's specific needs, as well as barriers to the identification and response to victims.

Methods: Survey of 168 healthcare workers including physicians, nurses, nurse practitioners, physician assistants, social workers, and patient/family advocate at multiple hospitals and medical clinics in urban, suburban, and rural locations. In particular, we focused on specialties and locations that would likely encounter victims, such as social work, general pediatrics, adolescent medicine, child abuse pediatrics, internal medicine, emergency medicine, obstetrics and gynecology, sexual assault nurse examiners and urban free clinics. The survey was sent to the chairs of each of these departments for distribution to potential participants.

Results: In two clinical vignettes, only 48% correctly classified a minor as a ST victim and only 42% correctly distinguished a ST victim from a child abuse victim. 62.5% of respondents said that they had never received training on how to identify ST victims. Those with training were significantly more likely to report ST as a major problem locally ($p < 0.001$), to have encountered a victim in their practice ($p < 0.001$) and to have greater confidence in their ability to identify victims ($p < 0.001$). The greatest barriers to identification of victims reported were a lack of training (34%) and lack of awareness (21%) on ST. There were also many action steps taken once a victim was identified, which included calling child protective services (69%), contacting local police (66%), referring patient to human trafficking victims' services (42%), and calling the national hotline (31%).

Discussion: Healthcare providers demonstrate a lack of knowledge and awareness of ST that correlates with their limited experience and training. Training is vitally important to improve identification of these victims and provide appropriate care for their specific needs. There is also a need for a coordinated, uniform protocol, without which, many healthcare workers are not confident in their ability to connect patients with the necessary services. This may consequently affect their willingness to screen potential victims.

DEVELOPMENT AND VALIDATION OF A RISK SCORE TO PREDICT ACCESS SITE COMPLICATIONS AFTER PERIPHERAL VASCULAR INTERVENTIONS

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Purpose: Access site complications (ASC) after peripheral vascular interventions (PVI) are associated with prolonged hospitalization and increased mortality. The aim of this study was to create a clinical scoring tool to stratify patients according to their risk of developing post-PVI ASC.

Methods: The Society for Vascular Surgery Vascular Quality Initiative database yielded 27,997 patients who had undergone PVI from July 2007 to January 2014 at 151 North American centers. Clinically and statistically significant ($p < 0.05$) preprocedural risk factors associated with in-hospital post-PVI ASC were included in a multivariate logistic regression model with ASC as the outcome variable. A predictive model was developed with a random sample of 70% of the dataset and validated against the remaining 30%. Risk factors were assigned weighted integers based on their beta coefficients, and the sum constituted the risk score.

Results: ASC occurred in 939 (3.4%) patients. Predictors included were: female gender, age >70 , white race, bedridden ambulatory status, IDDM, prior minor amputation, procedural indication of claudication and non-femoral arterial access site (model c-statistic = 0.637). The discriminatory power of the risk model was confirmed by the validation dataset (Brier score = 0.033). Higher risk scores correlated with increased frequency of ASC: 1.9% for low risk (score 0 - 15), 3.4% for moderate risk (16 - 27) and 5.0% for high risk (28 - 34).

Conclusions: The proposed clinical risk score based on 8 preprocedural characteristics is a simple tool to stratify patients at risk for post-PVI ASC. The risk score may assist physicians in therapeutic decision-making, including selection of the appropriate bleeding avoidance strategy, to improve outcomes in patients undergoing PVI.

FACTORS AFFECTING SPECIMEN ACCRUAL IN A COMMUNITY HOSPITAL BIOBANK

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Background: Community hospital biobanks are important contributors to several cancer genome programs. In an effort to improve the specimen accrual, we sought to determine cancer patients' attitudes and knowledge about donating tissue for research.

Methods: We mailed questionnaires to 500 patients who had undergone cancer surgery at our hospital in the year 2012.

Results: Response rate was 43.4% (217/500). Only 36/217 respondents were aware of the biobank (BB), 32 of whom had consented to tissue collection. Of the 181/217 who were not aware, 120 believed that they would have consented had they been aware, 46 were not sure, and 15 would have refused. Most respondents (174/217) saw possible benefit for others as the most important reason for consenting. 41/217 patients believed there is a potential for misuse of their tissue or personal information, 17 did not want to deal with any extra issues, and 11 did not understand the concept. None cited religious or cultural beliefs as factors influencing their decision.

Conclusions: Majority of the people were unaware of the concept of BB but if made aware, most of them would have consented primarily to help other cancer patients. Lack of patient awareness is an important difference between a smaller community hospital and a bigger quaternary setting. With increasing participation of smaller centers towards international cancer genome programs, improving patient awareness could be a major step in increasing specimen accrual. Despite being unaware, most patients showed good understanding of the concept of BB. The fear of misuse or commercial use of their tissue and personal information was their biggest apprehension. Hence, even a small, informative conversation addressing this issue could help allay patient fears about tissue donation.

Displayed Posters

1) UNUSUAL PRESENTATION OF PAUCI-IMMUNE RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS

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Introduction: Rapidly progressive glomerulonephritis (RPGN) is defined as a decrease in renal function over days to months. Clinical presentation ranges from insidious to acute, severe onset of three chief features: oligouria, edema, and macroscopic hematuria. Diagnosis must be made swiftly, or end stage renal disease or death can result. Here, we present a geriatric patient with crescentic and necrotizing pauci-immune RPGN whose only presenting symptoms were asymptomatic acute renal failure and a history of minor bilateral lower extremity edema. The objective of this case is to recognize an atypical presentation of RPGN in the geriatric population, where symptoms such as edema can be caused by a range of underlying pathologies more common than RPGN.

Case: 79-year-old female with a history of hepatic steatosis presented as a direct admission for evaluation of bilateral leg pain and edema, acute renal failure (ARF), and elevated inflammatory markers. Patient was previously followed outpatient for 8 months for bilateral leg pain only minimally responsive to NSAIDs; significantly elevated sedimentation rate led to diagnosis of polymyalgia rheumatica. Treatment with prednisone led to complete remission of symptoms; however, when prednisone taper was attempted prior to admission, patient relapsed and was started on trial of azathioprine, which was ultimately discontinued due to chest pain and fatigue. On re-evaluation at this time, labs were consistent with ARF, so patient was directly admitted. Labs at time of admission were significant for elevated creatinine (3.39) and BUN (84). Liver studies were consistent with prior CT evidence of hepatic steatosis. Physical exam was remarkable for 2+ pitting edema to mid-shin; musculoskeletal exam was unremarkable. During her hospitalization, the patient received extensive laboratory workup; notable results include P-ANCA 1:320, normal C-ANCA, normal PBO Ab, and normal PR3 Ab. Renal biopsy demonstrated focal necrotizing and crescentic glomerulonephritis, pauci-immune. Patient received three days of methylprednisolone before transitioning onto daily prednisolone, and she was started on cyclophosphamide and Bactrim.

Discussion: The geriatric population is particularly susceptible to developing ARF due to age-related changes in renal structure and function. As RPGN is the most common cause of ARF in the elderly, it is important to have a high index of suspicion if these patients present with lower extremity edema. Treatment of RPGN is divided into initial and maintenance phases. Initial treatment includes cyclophosphamide and glucocorticoids (initial methylprednisolone pulse followed by daily prednisolone). Prophylaxis (typically with Bactrim) against *Pneumocystis pneumonia* (PCP) is important, as up to 6% of patients develop PCP during the initial phase of treatment. Maintenance therapy involves switching the cyclophosphamide to azathioprine or methotrexate before eventually tapering.

2) UNUSUAL MYELOID SARCOMA/AML INVOLVING BREAST AND PANCREAS

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Introduction: Myeloid sarcoma (MS) is a tumor of myeloblasts or immature myeloid cells occurring in the bone or extramedullary sites. Common sites include sub-periosteal, lymph nodes and skin. However it rarely involves the breast and pancreas.

Case Presentation: A 59 yo female presented with abdominal pain, anorexia, fatigue, weight loss and left breast mass. Examination: Jaundice, 2x3 cm left breast mass and tenderness over the epigastric area. Labs revealed elevated liver function tests and bilirubin suggestive of obstructive jaundice. CT abdomen showed mass over the pancreatic head, inferior pole of the kidneys bilaterally, uterus and left breast area. ERCP was performed with biliary stent placement. PET scan showed hypermetabolic masses over the breast bilaterally, pancreatic head and uterus. Cytologies from pancreas were negative. Diagnostic mammogram revealed bilateral breast masses with biopsy consistent with malignancy. Immuno-histochemical stains were positive for CD45, CD117, myeloperoxidase and lysozyme and negative for CD34, which were suggestive of myeloid sarcoma. BM showed normal cellular marrow with approximately 5% atypical blasts and 46XX karyotype. NPM1 was positive in the breast biopsy sample but negative in marrow. Patient completed induction chemotherapy with cytarabine and idarubicin and consolidation with HIDAC. Follow-up PET scan showed near complete resolution of masses in breast, pancreatic head and uterus.

Discussion: The development of Myeloid Sarcoma (MS) may precede or concur with AML or CML or with other myeloproliferative disorders. The involvement of the breast and pancreas with MS is rare. Our patient had bilateral MS, which is extremely rare. MS is considered a diagnostic equivalent of AML (WHO Classification, 2008). In conclusion, the diagnosis of MS is often missed especially when presenting as a breast or pancreas mass. Most patients respond favorably to chemotherapy without surgery highlighting the importance of early diagnosis and prompt treatment.

3) UNUSUAL CASE OF WERNICKE'S ENCEPHALOPATHY

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Introduction: Wernicke's encephalopathy is a neurologic syndrome caused by thiamine deficiency. The classic triad includes encephalopathy, ophthalmoplegia and ataxia. The diagnosis is often missed as patients do not present typically.

Case Presentation: A 44 yo female presented to the ED with 2-week history of progressive confusion, disorientation, decreased interaction and not being her usual self. PMH: Alcohol intoxication (hospitalized) Exam: Vitals were stable. Patient appeared weak and confused. Exam revealed distended urinary bladder, disorientation, horizontal nystagmus, left lateral rectus palsy, gait ataxia and absent DTR. Labs showed hypoalbuminemia and elevated creatinine. Straight catheterization revealed >2L of cloudy urine with debris. Urinalysis & culture showed UTI with Proteus. CTOH was unremarkable and UDS was negative. Admission diagnosis included altered mental status, acute urinary retention and UTI. UTI was treated with PO antibiotics. Medications were reviewed and urologic evaluation was negative. Patient continued to have urinary retention and confusion. On Day 3, MRI revealed abnormal distribution of signal involving medial thalami, hypothalamus, mammillary bodies, and peri-aqueductal gray matter suggestive of Wernicke's encephalopathy. Patient was treated with IV thiamine for 5 days with complete recovery to baseline mental status at discharge.

Discussion: Wernicke's encephalopathy is relatively common in alcoholics. However, diagnosis is often missed due to atypical presentation. It becomes even more challenging if patient's alcohol history is unknown. Our patient is unique in that she presented with 2-week history of worsening confusion and urinary retention which was not described in literature to the best of our knowledge. Periaqueductal grey matter is proposed to play a vital role in normal urinary reflex. In our patient, urinary retention with periaqueductal gray matter changes resolved with thiamine treatment which supports this theory. In conclusion, high index of suspicion should be maintained for Wernicke's encephalopathy in alcoholic patients who present with atypical symptoms.

4) TRANSCATHETER AORTIC VALVE REPLACEMENT FOR FAILING BIOPROSTHETIC SURGICAL AORTIC VALVE

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Case Presentation: 78 yo male presented with fatigue and leg edema. He described dyspnea with minimal activities and occasionally at rest for two months. He also had persistent lower extremity edema for several months despite multiple diuretic adjustments. He had CAD and underwent CABG and bioprosthetic Aortic Valve Replacement 6 years ago. He had two Hospital admissions for decompensated Heart Failure in the last two months.

BP 123/44, HR 70, RR 20, Temperature 97.8, Saturation 94% on room air. He had rales in bilateral lower lung fields. Cardiovascular exam revealed normal S1 and S2, an early to mid diastolic murmur at RUSB with palpable thrill, jugular venous distension and 1+ pitting edema in both legs. WBC 6.5, Hb 10.0, Cr 2.48 [baseline 1.2], BNP 2010, CXR: Pulmonary Vascular Congestion, ECHO: severe Aortic valve regurgitation with mean gradient of 21, torn right coronary cusp of Aortic valve, moderately decreased LV function with EF of 44%. Patient's clinical condition worsened with increasing fatigue, somnolence and worsening renal function. He developed cardiorenal syndrome and required renal replacement therapy with Hemodialysis. Evaluated by two cardiothoracic surgeons who both agreed patient was extreme surgical risk. STS score was 9.47% with incremental risks for elevated BNP +3%, OSA +2% and frailty. On the 5th hospital day, he underwent successful Transcatheter Aortic Valve Replacement in failed Surgical Aortic Valve. Immediately following valve deployment, intraoperative TEE showed only trivial AI and pressure recordings revealed normalization of LVEDP. After procedure, his renal function improved and did not require renal replacement therapy, dyspnea and activity tolerance improved significantly and discharged home in good condition.

Discussion: Failing bioprosthetic surgical aortic valve poses significant challenge especially in high surgical risk patients. Its incidence is likely to increase as the population ages and more aortic valve surgeries are performed. Transcatheter aortic valve replacement for failed surgical bioprosthetic aortic valve [Valve in Valve] is reasonable alternative for high and extreme surgical risk patients that can obviate the need for surgery. Prospective randomized trials are needed to evaluate its efficacy.

5) PSEUDOINSULINOMA SYNDROME IN A NON-DIABETIC ELDERLY MAN FOLLOWING INADVERTENT SULFONYLUREA USE

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Introduction: Medication management is important for avoiding significant morbidity and mortality in the elderly, especially for those with cognitive impairment. Here, we report the case of an elderly man with no history of diabetes who presented with profound hypoglycemia that illustrates the importance of medication management. Laboratory testing revealed findings suggestive of an insulinoma. The sulfonylurea screen however returned positive for glipizide.

Case Description: A 91-year old male was found unresponsive by the EMS with a blood glucose level of 24 mg/dl. He was treated with glucagon and kept on a dextrose drip due to recurrence of hypoglycemic unresponsiveness, but was weaned off of the glucose infusion over the next 24 hours. The patient could not recall details of the event, but was lucid, awake, and alert during hospitalization thereafter. He reported recent dietary restrictions in an attempt to lose weight. The patient's wife was confident that he had not taken her anti-diabetic pills accidentally. Past history and medication review did not identify an apparent cause. Examination in the hospital was unremarkable except for mild wheezing.

Laboratory testing showed elevated c-peptide and insulin levels. He underwent a 72-hour fasting glucose test with no episodes of hypoglycemia and nadir blood glucose of 81 mg/dl. An ACTH stimulation test showed an appropriate physiologic response. The hypoglycemic agent drug screen came back positive for glipizide, a prescription drug for diabetes used by the patient's wife.

Discussion: Our patient was observed during an episode of hypoglycemia with Whipple's triad warranting further etiological investigation. In normal individuals, prolonged fasting will not manifest as hypoglycemia unless there is a defect in the normal compensatory mechanisms for the maintenance of normoglycemia. Identifying possible inadvertent ingestion of oral hypoglycemic agents, especially sulfonylurea agents that function by increasing insulin secretion, can prevent unnecessary testing for insulinoma and avert recurrence of such events. Hence, in a non-diabetic individual it is important for the provider perform pill inspection, drug screen, and measurement of insulin and c-peptide levels during an episode of hypoglycemia.

6) A CASE OF PRIMARY GASTRIC MUCOSAL MELANOMA

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Introduction: Extra-cutaneous melanomas account for 4-5 per cent of all malignant melanomas. Of these primary gastric mucosal melanomas are extremely rare and the exact incidence is unknown. There are very few cases reported in the literature.

Case: We present a case of 75 y/o gentleman who presented with acute epigastric pain and long standing diarrhea. The cause of diarrhea was not determined despite adequate lab evaluation and colonoscopy. An upper GI endoscopy done for epigastric pain , revealed a Shatzki ring in the esophagus, sliding hiatal hernia, esophagitis and a stricture in the 2nd portion of the duodenum with an ulcer adjacent to it. Stricture was dilated and the ulcer was cauterized. CT scan of abdomen/pelvis revealed gastric wall thickening and a cystic lesion in the greater curvature of stomach. EUS followed by a biopsy revealed this to be a melanoma. A thorough oral, skin and eye exam along with anoscopy did not reveal any suspicious primary lesions. PET/CT was negative for any other lesions. He underwent partial gastrectomy and pathological exam of the surgical specimen confirmed this to be a malignant melanoma which measured 2.5cm with subserosal extension.

Discussion: Primary gastric mucosal melanomas are rare and only a few case reports exist. The exact incidence of the disease is not known and based on the literature review the median survival rate is around 5 months. Vague presenting symptoms lead to delay in detection and poses a challenge for early diagnosis and treatment. Surgical resection remains the mainstay of treatment and the role of chemo-radiation and immunotherapy remains obscure. Our patient underwent surgical resection, and did not have any evidence of metastatic disease. He is doing well after 6 month follow up and is on close surveillance.

7) FIRST REPORTED CASE OF CARBAPENEM-RESISTANT ENTEROBACTERIACEAE IN WISCONSIN

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Introduction: Ever since the first case of Carbapenem-resistant Enterobacteriaceae in the United States was reported in 2001, this class of “superbug” has been emerging in health care facilities across the country. The most well-publicized outbreak occurred in 2013 at the National Institutes of Health, killing six of seventeen infected patients. However, the organism had not spread to Wisconsin until now.

Case Presentation: We describe a 72 year old Indian male with a history of pancreatic mass and placement of intraduodenal stent who presented with abdominal pain, vomiting, and fever. On admission, blood cultures were positive for Enterococcus and the patient was started on Unasyn and Flagyl. Interventional radiology was consulted for suspected biliary obstruction, and the patient was taken for percutaneous transhepatic cholangiography to remove the stent. Postoperatively, the patient was sent to the ICU for management of sepsis. A few days later, the drain cultures began to grow carbapenem-resistant *Klebsiella pneumoniae* (CRKP), and blood cultures grew the same organism shortly thereafter. Antibiotics were changed to tigecycline and colistin to treat the CRKP, as well as vancomycin to treat the enterococcal infection. Although the bacteremia successfully cleared after two weeks, the patient eventually died of pancreatic cancer during the hospitalization.

Discussion: CRE has now appeared in more than 40 states, and this alarming trend heralds the need for more research into the proper treatment of these infections. Current clinical evidence suggests treatment with tigecycline, a polymyxin, and in certain cases the addition of a carbapenem as well. The appearance of CRE in Wisconsin should alert physicians in the state of the need to understand the risk factors and proper treatment of CRE.

8) METHICILLIN-RESISTANT *STAPHYLOCOCCUS SEPTICEMIA* IN AN INTRAVENOUS DRUG USER WITH UNCONTROLLED TYPE I DIABETES MELLITUS PRESENTING AS CARDIAC TAMPONADE

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Introduction: Methicillin-resistant *Staphylococcus aureus* (MRSA) is a leading cause of hospital- and community-acquired bacteremia. We describe an uncommon presentation of MRSA septicemia in an intravenous drug user with purulent pericarditis resulting in tamponade, without subsequent endocarditis, as well as metastatic spread of the infection.

Case Description: A 25-year-old Caucasian female was transferred to our hospital following positive blood and urine cultures for *Staphylococcus aureus* with persistent hypotension and fevers. She was treated with intravenous fluids, vancomycin, and piperacillin/tazobactam for urinary tract infection and left lower lobe pneumonia for approximately 48 hours prior to transfer. She reported vague abdominal pain, nausea, chest tightness, and resolving burning micturition. Past medical history was significant for uncontrolled type I diabetes mellitus with resultant retinopathy and neuropathy as well as asthma and gastroesophageal reflux disease (GERD). She worked as an exotic dancer. She admitted to smoking half a pack of cigarettes per day, occasional alcohol use, and use of injected heroin as well as other opiates. Her family raised the possibility of the patient sharing needles with her significant other.

Upon examination, the patient's temperature was 98.7°F, blood pressure was 85/64 mmHg, heart rate was 117 beats per min, and respiratory rate was 30 with oxygen saturation of 90% on room air. She had several tattoos and needle marks were noted on her left forearm. The patient's abdomen was diffusely tender without guarding or rigidity. Jugular venous pulse was elevated at 10-11 cm. S1 and S2 were heard without other abnormal sounds. Range of motion in her right hip was restricted secondary to pain. Laboratory evaluation revealed leukocytosis, elevated venous lactate, acute kidney injury, and mild transaminitis. Urine drug screen was positive for benzodiazepines, cannabis, opiates, tricyclics, and methamphetamines. Transthoracic and transesophageal echocardiogram revealed cardiac tamponade with large pericardial effusion without valvular heart abnormalities. Pericardial fluid obtained following drain placement grew MRSA. On subsequent imaging with computed tomography (CT) scan and ultrasound, the patient was found to have metastatic seeding of the infection with MRSA positive bilateral pleural effusions, right sided hip joint effusion, right elbow joint effusion, and pyelonephritis with areas of renal infarction. Throughout her complex hospitalization, she was treated with intravenous antibiotics and later switched to oral therapy to complete an eight week course.

Discussion: MRSA is a commonly seen infection among intravenous drug users. Complications include endocarditis, pneumonia, sepsis, or the involvement of any organ system. Though less common, clinicians need to be aware of the potential for disseminated spread and multi-organ involvement.

9) A “CRYSTAL” CLEAR CASE OF PULMONARY FOREIGN BODY GRANULOMATOSIS

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Introduction: Pulmonary foreign body granulomatosis is a reactive process caused by embolization of foreign material within pulmonary arterioles resulting in inflammation, granuloma formation and fibrosis. Patients with this disease typically have a history of IV drug abuse and present with dyspnea and a dry cough. If not recognized early, the disease can progress to massive pulmonary fibrosis and end-stage lung disease.

Case: A 61 year-old male with a history of end-stage renal disease, hepatitis C and drug abuse presented with three days of dyspnea and hemoptysis. In the emergency department he was found to be afebrile, tachycardic and hypoxemic. Initial laboratory data was notable for a normal white blood cell count and baseline hemoglobin, renal function and coagulation studies. A urine drug screen was positive for cocaine, benzodiazepines, and amphetamines. CT angiogram of the chest showed no evidence of PE, but was notable for ground glass opacities in the right-middle and left-lower lobes. Blood cultures were collected and broad spectrum antibiotics were started. The patient was admitted to the inpatient medicine ward where he continued to have intermittent episodes of dyspnea and hemoptysis. An extensive infectious and autoimmune work-up yielded no significant findings. Bronchoscopy with lavage was significant for bloody return from the right-middle and left-lower lobes. Lavage fluid studies were notable for alveolar macrophages with rare hemosiderin, but an infectious or malignant etiology was not identified. Video-assisted thorascopic surgical biopsy was pursued. Biopsy results were significant for scattered crystalline, polarizable foreign material with an associated perivascular and alveolar giant cell reaction, consistent with the diagnosis of pulmonary foreign body granulomatosis.

Discussion: Foreign body granulomatosis typically results from embolization of pulmonary arterioles by insoluble material, which migrates through the vessel wall into the perivascular and interstitial tissue where it is phagocytized by macrophages and giant cells. Over time a granuloma is formed. Common substances include tablet fillers like talc or microcrystalline cellulose. Patient presentation ranges from dyspnea and dry cough to hemoptysis, night sweats and weight loss. Tissue biopsy is considered the diagnostic gold standard. Histologic findings include perivascular fibrosis, multinucleated giant cells, granulomas and polarizable material. No established therapy exists for this disease. Cessation from smoking and IV drug abuse is essential to avoid progression. This case illustrates the importance of considering pulmonary foreign body granulomatosis in patients presenting with hemoptysis, dyspnea and a diffuse ground glass pattern on chest CT without a discernable infectious, malignant or autoimmune etiology.

10) HEPATIC ABSCESES SECONDARY TO DIVERTICULITIS

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Introduction: The most common source of hepatic abscess in the U.S. is the biliary tree, especially in patients with cholecystitis, choledocholithiasis, or cholangitis. However, hepatic abscess can occur as a rare complication of sigmoid diverticulitis. When treating patients with diverticulitis, it is important to consider this complication as it is often overlooked and may delay treatment.

Case: A 58 year old male with past medical history significant for recurrent diverticulitis presented with chief complaint of constipation and left costal margin/upper quadrant pain after having a syncopal episode at home. He had developed classic symptoms of diverticulitis including constipation a week prior to admission. At the hospital, he underwent workup for the pain, which included a chest CT that incidentally showed multiple liver lesions. On physical exam, patient was afebrile and in no acute distress. Lab workup revealed an elevated white count of 11.8k. Patient also complained of a subjective fever that spiked periodically throughout the day. He underwent US guided drainage of the largest lesion and was treated with ceftriaxone based on the liver aspirate culture growing *Strep anginosus*. Despite antibiotics and drainage, patient continued to experience fevers, night sweats, and chills. His antibiotic coverage was then broadened to Ertapenem due to concerns of a polymicrobial infection. A repeat CT scan of the abdomen and pelvis identified multiple worsening loculated liver abscesses that were inaccessible for percutaneous drainage, and also a mural abscess in the sigmoid colon as the source of infection. Surgery was consulted for surgical drainage of the abscesses and a partial sigmoid resection. Patient was discharged on IV Ertapenem for a treatment duration of 17 days total.

Discussion: Patients who develop hepatic abscesses secondary to recurrent diverticulitis can present with vague symptoms. Fever is the most common presenting sign of liver abscesses. Only 50% of patients develop hepatomegaly, right upper quadrant abdominal pain, or jaundice. It is important to recognize that diverticulitis can seed the abscesses through the portal venous system. Invasion through this route is frequently associated with the formation of multiple abscesses that may require surgical drainage. Therefore, in a patient with recurrent diverticulitis, who presents with vague symptoms like chills, fevers, or upper quadrant abdominal pain, further workup should be performed to rule out hepatic abscesses as a complication. A high degree of clinical suspicion and appropriate medical and surgical intervention can provide a better prognosis.

11) DIABETIC KETOACIDOSIS: WHY HERE, WHY NOW?

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Diabetic ketoacidosis (DKA) is an acute metabolic derangement and commonly encountered complication of diabetes mellitus in the hospital. When susceptible type II diabetics present in DKA it is important to consider sinister underlying causes, like MI or infection, which may be masked by signs and symptoms of DKA. A 61 year-old man with DMII and multiple episodes of DKA attributed to medication non-adherence presented with a 10-day history of abdominal pain, vomiting, anorexia, polyuria, polydipsia, and chills. The patient reported not taking his insulin over this period, and his recent glycohemoglobin was 12%. He appeared somnolent with Kussmaul respirations, left abdominal tenderness, and a 1x1cm eschar without surrounding cellulitis on his left foot. The remainder of his exam was unremarkable. His chemistries were consistent with DKA. He was afebrile but had a WBC of 20,000. Chest x-ray and urinalysis did not reveal infection. Blood cultures were drawn, but antibiotics were deferred in the absence of clear infection. He was admitted to the ICU on DKA protocol. On day 2 DKA had resolved with slight improvement in overall symptoms despite persistent abdominal pain and nausea. WBC remained elevated and vancomycin was started for gram-positive cocci in clusters. Transthoracic echocardiogram showed no signs of cardiac infection, and CT abdomen was negative for abscess. On day 3 the organisms in the initial blood cultures were identified as methicillin-sensitive *Staph. aureus*. The patient then reported complaints of vague, dull posterior thorax pain that had been present for days. On day 4, the patient complained of ongoing nausea and chills. Antibiotic coverage was narrowed to cefazolin. Daily blood cultures became negative on this regimen. MRI showed a paraspinal fluid collection concerning for abscess dissecting the right posterior paraspinal and superficial muscles measuring 1x3x10cm (AP-transverse-cranio-caudad) from T1 to T4 vertebrae. Treatment suggestions were solicited from general surgery, neurosurgery, and interventional radiology, though incision and drainage was delayed two days due to precarious location of the abscess and lack of clarity in which team would take responsibility for intervening in this anatomic location. Unfortunately, the patient developed acute respiratory distress syndrome and septic shock requiring mechanical ventilation and vasopressors. General surgery then drained purulent fluid from the paraspinal region. However, ultrasound showed persistent fluid collections, and he was transferred to a quaternary center for further care. This case illustrates the importance of early, thorough evaluation of patients who present in DKA. Poorly controlled diabetes can precipitate DKA, but we must remember bacterial infections can trigger DKA or develop as a consequence of hyperglycemia. Patients obtunded from DKA also may not be able to clearly point out areas of pain. Here an aggressive team-based approach may have resulted in a better patient outcome.

12) BACK PAIN PRESENTING AS SOLITARY BONE PLASMACYTOMA

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Introduction: Back pain is a common chief complaint. Therefore, it is imperative that clinicians have a high index of suspicion for mimics of common back pain. The following case of solitary bone plasmacytoma demonstrates how clinicians must remain vigilant to alarming symptoms of back pain and escalate management appropriately.

Case Study: A 57 year old woman with no relevant past medical history presents with a 2 week history of focal low back pain with acute onset. History and exam did not reveal any particular trigger or trauma, red flag symptoms, or neurological deficits. The patient failed conservative therapy with scheduled NSAIDs and had acute worsening of pain one week after the initial onset. She subsequently had a CT scan of her back which revealed a L2 lytic lesion with posterior vertebral body fracture. The lytic lesion was suspicious for multiple myeloma, plasmacytoma, or lymphoma. A PET scan revealed a solitary non FDG avid L2 lytic lesion and no evidence of widespread disease. She was then transferred to our hospital for biopsy and vertebroplasty. Labs on admission showed normal calcium, creatinine, and hemoglobin levels. Biopsy of the L2 lesion revealed normal hematopoietic elements admixed with sheets of numerous plasma cells. A bone marrow biopsy followed, which did not demonstrate evidence of multiple myeloma. The patient was diagnosed with plasmacytoma and was scheduled for radiotherapy. Although she continues to require pain management she is able to exercise and perform activities of daily living.

Discussion: Back pain is often self-limited and resolves within 6 weeks with conservative management. Therefore, diagnostic imaging is usually withheld for one month and is then indicated in the absence of reduction of symptoms. However, if a fracture, tumor, infection, or cauda equina syndrome is suspected, then the patient should urgently undergo appropriate imaging. A dialogue between the clinician, the radiologist, and any appropriate consultation should ensue to correlate imaging findings with patient presentation in order to rule out high-morbid conditions and to order any further testing. Patients diagnosed with solitary bone plasmacytoma should be followed regularly, as 54% of patients ultimately progress to multiple myeloma even after radiation therapy. Appropriate testing includes urine and serum protein electrophoresis with immunofixation, CBC, serum creatinine, and serum calcium. Follow-up should be every 4 to 6 months for the first year after diagnosis, and then annually afterwards.

13) EARLY DIAGNOSIS AND TREATMENT OF PERICARDIAL ACTINOMYCOSIS IMPROVES OUTCOME

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Introduction: Actinomycosis pericarditis is a rare condition with good prognosis if diagnosed and treated early and appropriately. Because of its subtle presentation, it is important to consider it in the differential diagnosis presenting with pericarditis with risk factors.

Case Description: This is the case of a 55-year-old male with a history of benign lung mass as well as gunshot wound to the left upper chest with remaining bullet fragments. He presented with weakness and hypotension which was unresponsive to initial fluid resuscitation. His physical exam revealed very poor dentition and posterior neck scarring. His chest was clear to auscultation while his cardiovascular exam was otherwise normal. The ECG was significant for low voltage and his Echocardiogram had features of tamponade. Urgent pericardiocentesis was done resulting in 550 ml of yellowish fluid which stabilized the patient hemodynamically. He required minimal vasopressor support. He was initially empirically treated with IV vancomycin and meropenem. This was eventually changed to IV ertapenem once culture demonstrated branching anaerobic, filamentous bacteria suggestive of actinomycosis. Collagen vascular work up was negative. He was discharged on a three-month course of IV ertapenem. He did require readmission for a pleural effusion requiring thoracentesis. In follow up, echocardiogram revealed no evidence of constrictive pericarditis while clinically his hemodynamic status completely normalized. His predisposing factors for actinomycosis included poor dentition, prior gunshot wound to the chest with remaining bullet fragments and, in a retrospect a benign lung mass most likely attributable to thoracic actinomycosis.

Discussion: This case stressed the importance of early diagnosis and treatment of a rare pericardial disorder that can lead to an improved outcome and decrease the risk of developing constrictive pericarditis.

14) DIFFUSE LARGE B CELL LYMPHOMA WITH SECONDARY CUTANEOUS MANIFESTATIONS IN A 64-YEAR OLD MALE

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Introduction: Diffuse Large B Cell Lymphoma (DLBCL) is the most common lymphoma with an incidence of about 7 cases per 100,000 people per year. DLBCL will typically present with a fast growing symptomatic mass but can also present with extranodal involvement. This case demonstrates that diffuse large B cell lymphoma can present as a cutaneous manifestation along with systemic B symptoms.

Case: 64-year-old male with a 6-day history of ventral and dorsal truncal rash. 1-2 weeks prior to admission patient had multiple episodes of shaking and weakness. His family reported he was not acting like himself and was often confused. He was seen at an OSH where an MRI brain was negative. The shaking and tremors started to improve however he noticed an erythematous rash in his axillae bilaterally and had continued generalized weakness. On day of admission, rash had progressed from axillae, across the chest and down to the groin. The rash was also present on his back and buttocks. Patient complained of weakness, fatigue, lymphadenopathy, and low grade fever. He was found to be pancytopenic with downtrending of his platelets, Hgb, WBC throughout hospitalization. A CT chest/abd/pelvis was completed which showed diffuse lymphadenopathy and hepatosplenomegaly. Labs were drawn including SPEP, UPEP, immunoelectrophoresis, CPK, LDH, HIV, EBV, CMV, parvovirus, anaplasma, erlichiosis, and RMSF serology. Dermatology performed skin biopsy of rash and surgery biopsied the inguinal node. Skin biopsy showed atypical lymphoplasmacytic infiltrate with lambda immunoglobulin light chain restricted plasma cells. Lymph node biopsy was performed showing CD5-/CD10- B cell non-Hodgkin lymphoma with plasmacytic differentiation. Heme/Onc was consulted to perform a BM biopsy, which demonstrated B cell non-Hodgkin lymphoma with plasmacytic differentiation.

Discussion: From our literature review, we have not found cases and treatments of cutaneous manifestations secondary to DLBCL. Most published literature focus on primary cutaneous B-cell lymphoma (PCBCL). PCBCL is a lymphoma with a B-cell phenotype that present only in the skin. Usually, there is no evidence of systemic or extracutaneous disease at initial presentation. However, based on our patient's biopsy and hemotologic results, it was discovered that he suffered from systemic B-cell lymphoma with secondary skin involvement versus a primary cutaneous pathologic process. This case serves as a good example to further study the clinical features, prognosis, and treatment options for DLBCL with secondary cutaneous manifestations.

15) A CASE OF ANTIBIOTIC INDUCED IGA VASCULITIS

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Introduction: IgA vasculitis is a systemic small vessel vasculitis characterized by IgA deposition in the tissue. It is most commonly seen in children after an upper respiratory infection. This case demonstrates antibiotic-induced IgA vasculitis in an adult male.

Case: A 55-year-old male with a history of poorly controlled type 1 diabetes mellitus with peripheral neuropathy, chronic kidney disease, and a chronic diabetic right foot ulcer complained of worsening fatigue.

On admission, he was afebrile with stable vital signs. Pertinent physical exam findings included right fifth metatarsal head ulcer with associated erythema. Distal pulses were intact. Pertinent labs included leukocytosis with left shift and negative blood cultures. The patient was started on Vancomycin, Ciprofloxacin, and Metronidazole. Three days later, he complained of bilateral hand pain and swelling. A purpuric, bullous rash to the hands, spreading to the upper extremities, trunk and lower extremities, was noted on exam. Biopsy revealed perivascular neutrophilic infiltrates with leukocytoclasia and extravasated red blood cells. Direct immunofluorescence staining was positive for IgA, C3, and fibrinogen. ANCA panel, hepatitis panel, liver function tests, streptococcal ASO titer, C3, and C4 were all normal. A urinalysis was positive for red blood cells. Antibiotics were changed to Clindamycin after a bone culture grew Methicillin-resistant *Staphylococcus aureus*. Within one week, the skin lesions began to resolve. The patient was diagnosed with antibiotic-associated IgA vasculitis. This eventually resolved, and he did not require steroids. With multiple debridements, amputation of the 4th and 5th metatarsal, hyperbaric O₂ and Clindamycin, he improved without further complications.

Discussion: Antibiotic-induced IgA vasculitis is rare, but must be recognized quickly. Offending agents should be discontinued immediately, and close monitoring for multi-organ involvement is essential. Treatment is usually symptomatic, but may require systemic steroids if multiple organ failure occurs.

16) SEGMENTAL ARTERIAL MEDIOLYSIS AS A CAUSE OF CATASTROPHIC INTRA-ABDOMINAL HEMORRHAGE

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Introduction: Segmental arterial mediolysis (SAM) is a non-atherosclerotic, non-inflammatory arteriopathy of unknown cause. It generally causes lysis of the medial layer of the arterial wall, which can result in arterial dissection, stenosis, occlusion or aneurysm formation.

Case: A 79 year old female with a history of atrial fibrillation is transferred to the ICU with hemorrhagic shock of unknown cause. The patient had been feeling well until she suddenly developed severe low back and abdominal pain. She presented to an outside hospital and was found to be hypotensive. Initial labs revealed a transaminitis and a decline in hemoglobin to 10.0 g/dL. A CT abdomen/pelvis was unrevealing apart from a small, non-specific lesion in the right hepatic lobe.

On transfer, the patient remained hypotensive and tachycardic. She appeared pale and had diffuse tenderness to palpation throughout her abdomen, worst in the epigastric region & RUQ. Labs revealed a worsening anemia with a Hgb of 5.7 g/dL and a supra-therapeutic INR of 5.0. She was placed on a norepinephrine drip and transfused with large quantities of pRBC's and FFP. A repeat CT abdomen/pelvis revealed massive intra-abdominal hemorrhage likely originating from the previously seen lesion in the right hepatic lobe. An angiogram of the common & proper hepatic arteries revealed diffuse narrowing, pseudoaneurysm and active bleeding involving the left & right hepatic arteries. The right, common & proper hepatic arteries were successfully embolized. After the procedure her blood pressure improved and her blood counts remained stable. Rheumatology was consulted and felt that the clinical course and radiologic findings were consistent with SAM. She was discharged shortly thereafter and made a full recovery.

Discussion: SAM is a rare and life-threatening arteriopathy of unknown cause. We present a case with a classic presentation of life-threatening intra-abdominal hemorrhage. Other common presentations include acute retroperitoneal or intra-cranial hemorrhage. Diagnosis is can be made by via angiography (CT or MR), but is often made at autopsy. Treatment usually requires either emergent endovascular or surgical intervention. Prognosis is poor with fatality rates near 50%.

17) DISSEMINATED STRONGYLOIDIASIS IN HIV PATIENT AFTER STEROID USE: A CASE REPORT

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Introduction: *Strongyloides stercoralis* is an intestinal nematode acquired in tropical and subtropical areas. Most often it causes chronic asymptomatic infection but a change in immune status can lead to disseminated strongyloidiasis which carries a very high mortality rate if left untreated.

We report a case of an HIV patient with disseminated strongyloidiasis after receiving steroids.

Case Presentation: The patient is a 51 yo female HIV positive patient originally from Puerto Rico with past medical history of recurrent GI symptoms, adult onset asthma, and recurrent pruritic skin rash who was treated with high dose steroids for assumed contrast allergy. Six days later, she presented to the hospital with abdominal pain, severe hypotension, respiratory failure, and renal failure. She was admitted to ICU and blood cultures grew gram negative rods and clinically diagnosed meningitis. Ultimately Strongyloidiasis was diagnosed by the presence of larvae in the stool. The patient was subsequently treated with Ivermectin 200 mcg/kg daily and responded well.

Discussion: Disseminated strongyloidiasis is a serious complication of intestinal strongyloidiasis. It occurs mainly in immunocompromised patients and has a significant mortality rate. To have an impact on early diagnosis and treatment, clinicians need to have high clinical suspicion in patients who present with certain clinical features and once lived in an endemic area.

18) A CASE OF RIGHT HEART FAILURE SECONDARY TO AORTA TO RIGHT PULMONARY ARTERY FISTULA

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Fistulization of prosthetic vasculature is a known complication of vascular repair, however, the fistulization of aortic root repairs is a rarer phenomenon.

A 65-year-old male with a history of Bentall procedure repairing his ascending aortic aneurysm with associated mechanical aortic valve replacement for severe aortic stenosis presented with nonproductive cough for 1 week which was worse when sitting concerning for congestive heart failure exacerbation. The patient was found to have a troponin elevation, and given this, underwent transthoracic echocardiography (TTE) which revealed a normal ejection fraction but with new, severe pulmonary arterial hypertension and dilated right ventricle. Due to these findings, the patient underwent thoracic computed tomography (CT) with intravenous contrast showing a perigraft collection around his prosthetic aortic segment of 9.1 cm x 8.7 cm with marked mass effect on the main right pulmonary artery. Plan was made for urgent Cardiac Surgery, however, the patient decompensated with cardiogenic shock and pulseless electrical activity arrest with return of spontaneous circulation. He was found to have a fistula between his aorta and right pulmonary artery on thoracic CT angiography. He was taken to the operating room for repair. Post-operatively, he required continuous renal replacement therapy (CRRT) and multiple vasopressors which could not be weaned. The decision was made to transition him to comfort measures with him passing away shortly thereafter.

This case illustrates a rare complication of the Bentall procedure with cardiogenic shock and fistulization occurring leading to a surgical emergency. This complication is, unfortunately, known in other uses of prosthetic material. Recognition of this syndrome is imperative as time is limited.

19) KAPOSI SARCOMA PRESENTING AS ASCITES IN A RENAL TRANSPLANT PATIENT

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Introduction: Kaposi sarcoma (KS) is a vascular neoplasm caused by human herpes virus 8 (HHV-8). Demographics and risk factors differentiate the four clinical KS subtypes, namely classic, endemic African, immunosuppressive therapy-associated, and AIDS-related. Dependent on the subtype, KS can have a variable course, ranging from the classically localized cutaneous lesions to widely disseminated disease. We report a case of disseminated Kaposi sarcoma presenting as ascites in a renal-allograft recipient.

Case: A 34 year-old African-American male with history of end-stage renal disease secondary to Alport's syndrome s/p 6/6 HLA-matched kidney transplant eight months prior presented with gradual-onset abdominal bloating. He also complained of epistaxis, painful gum-line swelling, and easy bruising. At the time of transplantation, he was induced with thymoglobulin. He received tacrolimus, mycophenolate mofetil, and prednisone for maintenance; tacrolimus levels ranged from 7 to 12 ng/dL. Recently, he had developed CMV and BK viremia, which responded to valganciclovir and reduction in immunosuppression, respectively. Physical exam revealed a thin male with tender, peri-molar violaceous swellings in the oral cavity. His abdomen was softly distended and nontender with shifting dullness. A large darkened macular lesion was noted on his left shin. Basic labs were significant for new thrombocytopenia, mild anemia, and a rise in creatinine from 1.2 to 1.8 mg/dL. Liver function tests were normal. Paracentesis removed 1.3 L of grossly bloody ascites, with SAAG of 0.9. Ultrasound, CT, and MRI revealed innumerable nonspecific liver lesions. Transjugular liver biopsy confirmed a diagnosis of Kaposi sarcoma by morphology and immunohistochemistry positive for HHV-8. PET scan suggested disease dissemination to the oropharynx, lungs, and para-aortic lymph nodes. The patient's tacrolimus was promptly replaced by an mTOR inhibitor (sirolimus); mycophenolate and prednisone were continued. Paclitaxel chemotherapy was initiated. Three months later, the patient's abdominal bloating and oral, skin, and liver lesions are vastly improved.

Discussion: This case illustrates a severe and rare complication of immunosuppression, while highlighting the diagnostic challenge often posed by unusual disease manifestations in the transplant patient. KS affects an estimated 0.5-5% of renal transplant recipients, in whom cutaneous signs are arguably unanimous at presentation. However, post-transplant KS is often more aggressive than classic KS, with extracutaneous involvement also common. Optimal treatment and prevention strategies of post-transplant KS are ill-defined and require close monitoring due to increased risk of organ rejection.

20) RECURRENT BACTERIAL MENINGITIS

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Introduction: Recurrent bacterial meningitis (RBM) is a rare phenomenon which poses a considerable diagnostic challenge, often requiring multi-disciplinary care. With the mortality rate for acute bacterial meningitis (ABM) approaching 25%, the need for prompt initiation of empiric treatment and subsequent etiological workup is crucial to improve outcomes. Included in the differential for RBM are (broadly): anatomical defects (congenital, acquired, or iatrogenic), immune compromise (suppression or deficiency), and seeding via parameningeal infections. Below we present a case of recurrent bacterial meningitis secondary to an acquired anatomical defect following non-penetrating head trauma.

Case: We discuss a 63-year-old Caucasian female with a history of a basilar skull fracture secondary to a motor vehicle collision at the age of 11, recurrent CSF leaks s/p endonasal repair at age 59, and RBM (x4) who presented to the ED with a 12 hour history of frontal headache, fever (101.3 F), chills, nausea/vomiting, nuchal rigidity, and photophobia. She denied SOB, dysuria, cough, rash, vision changes, syncope, rhinorrhea, and otorrhea. She denied previous sick contacts, recent travel, and illnesses. Lumbar puncture was performed in the ED and CSF analysis showed a white count of 8,600/ μ L with 97% PMNs, no bacteria, a glucose of 52 mg/dL (versus 183 mg/dL in serum), and a protein of 340 mg/dL. She was started on empiric IV vancomycin, ceftriaxone, dexamethasone, and admitted to the floor. CSF and blood cultures had no growth in five days. HSV PCR of CSF was negative. Infectious disease, ENT, and neurosurgery were consulted. MRI which showed; post-surgical ethmoid changes from previous patch repair in 2009, no dural enhancement, no evidence of active leak, and no dural lymphatic fistula. Nasal endoscopy was performed and found no evidence of active CSF leak. On day two of IV vancomycin, ceftriaxone, and dexamethasone she had near complete resolution of fever, nuchal rigidity, photophobia, and frontal headache. Dexamethasone was discontinued on day three, ceftriaxone on day seven, and she was discharged on day eight to complete a 10-day course of IV vancomycin.

Discussion: Recurrent bacterial meningitis (RBM) is a rare entity. An estimated 2-9% of acute cases will have recurrent episodes. Interestingly, the mortality rate in recurrent meningitis is less than that of single case meningitis (15% versus 34%). A recent case series of 34 patients with RBM, found that 24 patients (76%) had identifiable predisposing factors (i.e. head trauma, CSF leak, and immunosuppression), making a thorough history paramount in guiding management. A recent meta-analysis found that, of RBM cases with known etiology, 59% were due to underlying anatomical defects. Of those with anatomical defects, 48% had acquired basilar skull fractures (or 28% of the total cases). Some 36% of cases had underlying immune compromise, of which complement deficiency and HIV/AIDS comprised 55% and 33%, respectively.

21) AZATHIOPRINE INDUCED SEVERE ENTERITIS IN A PATIENT WITH ULCERATIVE COLITIS

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Introduction: Azathioprine (AZA) induced adverse reaction affecting gastrointestinal tract is very rare. There is only one reported case with AZA induced colitis to our knowledge. We present a case of severe enteritis from AZA mimicking the flare of inflammatory bowel disease.

Case Presentation: 52 YO male with 9-month history of ulcerative colitis (UC), on mesalamine, presented with severe diarrhea. He was evaluated in ER 12 days prior to his hospitalization for diarrhea. His stool test was positive for *C.difficile* PCR but negative for *C.diff.* toxins. The result was considered indeterminate given high sensitivity and increased possibility of false positive PCR test for *C.difficile*. Decision to treat him for *C.diff* was made and he was treated initially with metronidazole followed by oral vancomycin as he did not respond to metronidazole. Due to refractory symptoms colonoscopy was done which showed severe pancolitis with no evidence of pseudomembrane. CT enterography revealed colitis with no small bowel involvement. He was diagnosed with UC flare. Steroid was started and patient was placed on AZA in conjunction with Adalimumab. TPMT level was checked before starting him on AZA which was normal. His symptoms improved temporarily. After 10 days his diarrhea came back. He was readmitted with severe diarrhea and dehydration. He was afebrile. Labs revealed leukocytosis (19,000), normal inflammatory markers (ESR, CRP) and negative work up for infectious etiologies (*C. difficile*, HSV, CMV, HIV). Colonoscopy showed improvement in previously seen pancolitis. Small bowel capsule endoscopy (SBCE) was performed for diarrhea refractory to the treatment and with the concern of Crohn's disease, revealed severe erosive enteritis confirmed with biopsy. His adalimumab was switched over to infliximab with no benefit. Given the low possibility of UC flare in the light of improving colitis and negative infectious work up, concern of AZA induced enteritis was raised. AZA had been stopped. We continued infliximab and oral steroids. He has had dramatic improvement in his diarrheal symptoms with gradual reduction in his stool frequency. Follow up SBCE 14 days after discontinuation of AZA was normal.

Discussion: Adverse reactions to AZA can be classified as dose related toxic reactions (myelosuppression and hepatotoxicity) and dose-independent idiosyncratic reactions. For our patient, we consider enteritis was due to AZA use, since it occurred early and no infectious etiologies were found. Furthermore, Enteritis resolved completely with discontinuation of AZA. The pathophysiological mechanism is unknown. This reaction was unrelated to TPMT activity. This case emphasizes the fact that AZA induced enteritis could present like a flare of IBD or GI infection.

22) ENDOSCOPIC YIELD AND APPROPRIATENESS OF ESOPHAGOGASTRODUODENOSCOPY (EGD) IN A COMMUNITY HOSPITAL BASED GASTROENTEROLOGY PRACTICE

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Background: The appropriateness of EGD has become an important issue as the use of EGD becomes more widespread. Aim of our study was to determine the adherence to American Society of Gastrointestinal Endoscopy (ASGE) indication guidelines and diagnostic yield of EGD in a gastroenterology practice within an integrated health care system.

Methods: A random sample of all EGDs completed in 2013 were retrospectively reviewed for demographics, hospitalization status, indication, findings (relevant to indication or not), and urgency. Appropriate indication was determined by adherence to ASGE guidelines. Appropriateness and diagnostic yield were calculated separately within these categories. The diagnostic yields between appropriate and inappropriate EGDs were compared using the chi-square test.

Results: 451 EGDs were included. The patients' mean age was 57.3 years (71.2% greater than 50 years,) more Female (56.4%), more Outpatient (86.4%), and more elective (83.4%). Indications were appropriate in 377 cases (83.6%). Inappropriate indications were more likely if the patient was Female, 68.9% vs. 54.1% ($p=0.019$) or <50 years old, 62.1% vs. 22.2% ($p<0.001$). Relevant findings were more likely if indication was appropriate, 57.8% vs. 17.5% ($p<0.001$). Of the exams with appropriate indications, the relevant yields by diagnosis were as follows: Refractory gastroesophageal reflux disease (GERD) 38.2%, Abdominal Symptoms 42.5%, Portal Hypertension 75%, Iron Deficiency Anemia 59%, Dysphagia 75%, Barrett's screening 2%, vomiting 57.1%, and Upper gastrointestinal (GI) bleeding 82.6%. Relevant findings were more likely if the patient was Male, 54.6% vs. 48% ($p=0.017$); Inpatient, 77% vs. 47% ($p<0.001$); and age >50 , 52.6% vs. 47.7% ($p=0.003$).

Conclusion: This study demonstrated good adherence to ASGE guidelines for EGD indications within this practice. Higher yields of relevant findings were obtained in patients who were male, inpatients, and older than 50 years. It confirms that most of the guideline recommended indications predict the identification of relevant lesions on EGD in fairly high percentages. EGDs for surveillance of malignancy in Barrett's esophagus had lowest diagnostic yield.

23) CHEST PAIN NOT A NORM IN AORTIC DISSECTION

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Introduction: Aortic dissection is a relatively uncommon, though catastrophic, illness often presenting with severe chest pain and acute hemodynamic compromise. Prompt and accurate diagnosis with early intervention are crucial for survival.

Case Presentation: 61 yo male with HTN was found lying on a sidewalk. On arrival to the ED, patient was agitated and complaining of left leg pain for two hours. Physical exam was notable for BP 257/138, no palpable pulse in the left pedal and dorsalis pedis arteries. Abnormal labs include creatinine of 1.3, lactic acid of 2.6 and CPK of 14,405. Chest x ray showed mild pulmonary congestion, enlarged and tortuous aorta. He was admitted for hypertensive encephalopathy and limb ischemia. BP was controlled but patient became more obtunded and left leg became cold and cyanotic. Doppler arterial U/S revealed near complete occlusion of common femoral artery with no visualization of right iliac artery. With the impression of ilio-femoral thromboembolism and compartmental syndrome of the legs, patient was taken to the OR for limb revascularization and fasciotomy. Prior to the surgery, another high BP raised suspicion for aortic dissection and TEE was obtained. TEE, and later on CT angiography, showed aortic dissection extending into the femoral vessels. Besides for limb revascularization and fasciotomy, patient had replacement of the ascending and arch of aorta with elephant trunk. Despite the delay in treatment of aortic dissection, patient survived though he remained dialysis dependent on discharge.

Conclusion: Diagnosis of aortic dissection requires a high index of clinical suspicion, especially in the absence of the classic symptoms. Limb ischemia should be approached with wide differentials on top of local thrombo-embolic events.

24) CEREBELLAR STROKE DUE TO PARADOXICAL EMBOLISM VIA PATENT FORAMEN OVALE

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Learning Objective: Identify patients with posterior circulation stroke due to paradoxical embolism.

Introduction: Cerebellar infarction can result from vertebral artery dissection, cardioembolic event, hematologic disturbances or complicated migraine. Paradoxical embolism is an uncommon cause of posterior circulation stroke.

Case Presentation: A 24 yo male with remote history of migraines presented with one day history of severe occipital headache, vertigo, nausea and vomiting. On exam BP was 150/100. Neuro exam showed no focal deficits. Rest of exam was unremarkable. CT brain showed hypo attenuation of left inferior cerebellum. Brain MRI showed cerebellar infarct in PICA distribution. CBC, CMP, Drug screen, Hypercoagulability workup, ANA were unremarkable. TEE showed patent foramen ovale with atrial septal defect. Lower Extremity Venous Doppler, CTA of head and neck were normal. He was initially treated with ASA, atorvastatin. Symptoms improved on Day 2 and he was discharged on Day 3 with clopidogrel and atorvastatin. Patient was given the option to enter a PFO closure study for paradoxical embolic stroke.

Discussion: Cerebellar infarctions account for approximately 1.5% of elderly population with stroke. Incidence in younger population is unknown. Several studies have suggested that embolic infarcts may affect the posterior cerebral artery and cerebellum in approximately 35% and 54% of cases respectively and an increased incidence was noted in patients with PFO or ASD. Based on recent studies, there is no significant benefit of PFO closure over medical therapy. Individualized approach should be used especially in a young patient considering life long antiplatelet or anticoagulant therapy vs PFO closure. This case highlights the atypical presentation of posterior circulation stroke, the importance of thorough evaluation (including TEE) in younger population and the need for personalized approach in management.

25) NEUROENDOCRINE TUMOR METASTASIS TO EXTRAOCULAR MUSCLE

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Introduction: Neuroendocrine tumors are rare, accounting for 0.5% of all malignancies. They are typically slow-growing and do not frequently metastasize. However, patients who present with symptoms from this disease typically have metastases with the most common sites being the lymph nodes, liver and bone marrow. In addition, a review of the literature reveals that this tumor has a predilection for metastasizing to extraocular muscles causing eyelid swelling, proptosis and limiting extraocular motion.

Case: 71 year old male with history of metastatic neuroendocrine tumor status-post chemotherapy presented with progressively worsening left eye redness, decreased visual acuity, and pain since cataract surgery three months prior. Several days prior to admission, he started ciprofloxacin eye drops along with oral amoxicillin/clavulanate for presumed pre-septal cellulitis. Despite this treatment, his eye symptoms worsened and he was referred for admission. Physical exam revealed left upper eyelid warmth and tenderness, left eye proptosis and severe conjunctival injection. Other notable findings included elevated intraocular pressure, afferent pupillary defect and absent corneal sensation in left eye. Extraocular movements were severely limited on left, normal on the right. Vision exam revealed 20/20 vision in right eye and only light perception ability and appreciation of gross hand motion in left eye. Laboratory data was unremarkable. MRI of orbits and brain revealed diffuse extraocular muscle enlargement most significant in left medial rectus with heterogeneous contrast enhancement consistent with metastatic neuroendocrine tumor. He underwent emergent surgical ocular decompression and extraocular muscle biopsy and had improvement in left eye proptosis and vision. Pathology confirmed neuroendocrine tumor metastasis. Radiation therapy was initiated along with dexamethasone.

Discussion: Metastatic neuroendocrine tumors are a rare occurrence and very few cases have described orbital involvement with metastatic neuroendocrine tumors; even fewer have described extraocular muscle involvement. MRI and CT are excellent choices to evaluate the orbits if metastatic disease is suspected. It should be noted that other entities, such as thyroid orbitopathy, infectious myositis or idiopathic orbital inflammatory syndrome, may present with similar findings on imaging making biopsy essential for diagnosis. Although there is no clear consensus for treatment, radiation therapy has been successful. In patients with a history of neuroendocrine tumor presenting with proptosis and limited eye mobility, it is important to consider non-infectious causes such as tumor metastasis, to rapidly perform imaging, and obtain biopsy since early diagnosis and initiation of radiation therapy may spare vision.

26) BACTERIAL ENDOPTHALMITIS IN PATIENT WITH CROHN'S DISEASE

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Background: Extraintestinal manifestations of Crohn's disease are broad. Though rare, transmucosal seeding and bacteremia should be considered in an immunocompromised patient with poorly controlled Crohn's disease and symptoms consistent with septic emboli.

Case: A 72 year-old woman with history of coronary artery disease and refractory Crohn's disease was admitted with acute painless bilateral vision loss. The patient had been hospitalized at an outside hospital for demand ischemia myocardial infarction secondary to anemia from refractory Crohn's disease. During that admission she suffered vision loss first in her left eye and consequently in her right eye within hour to follow and was transferred to our institution for further management. Physical exam on admission, including complete neurological assessment, was notable for marked bilateral hyperemia on external eye exam and vision limited to finger movements and light. Full ophthalmologic exam demonstrated severe intraocular inflammation and haze, but no obvious retinal detachment. Intraocular pressures were normal. Her initial work up included a complete blood count and complete metabolic panel which were notable for leukocytosis to 13, baseline anemia, platelet count of 80, and albumin of 1.3. CT of her head revealed mild age-related cortical atrophy with no acute intracranial process. Blood and vitreous cultures were drawn at the bedside and ultimately both were positive for group B Streptococcus. She was immediately treated with intravitreal and intravenous antibiotics, and Atropine and Prednisone ophthalmic solutions without improvement of vision on discharge. Streptococcal bacteremia and resultant endophthalmitis was suspected secondary to seeding across an inflamed and compromised bowel mucosa.

Discussion: Crohn's disease is an inflammatory process of unknown pathogenesis characterized by transmural inflammation of any segment of the gastrointestinal tract. While symptoms are often localized to the gastrointestinal system, many patients also present with various extraintestinal manifestations and complications. These most commonly include fistulization, strictures, perforation, intra-abdominal abscess formation and various hepatobiliary disorders; however, in patients with severe immunologic compromise and malnutrition, atypical presentations can occur. Bacterial endophthalmitis is rare and most commonly occurs following intraocular surgeries, with endogenous endophthalmitis only accounting for 2 to 8% of all cases. While initiated by bacterial infection, much of the damage in endophthalmitis comes from the ongoing inflammatory process, which in part explains the lack of clinical improvement despite adequate antibiotic treatment.

27) Q AND A

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Introduction: Aortic aneurysms are not commonly infected. When they are, staph aureus and salmonella are the prevailing pathogens. Here is a mysterious case of a mycotic aneurysm caused by a worldwide pathogen that was once unidentified and is now uncommonly suspected.

Case: A 78 year old female former smoker with diabetes mellitus and hypertension presents with back and abdominal pain and constitutional symptoms of subjective fevers, chills, night sweats, diminished appetite, and fatigue. CT imaging at an outside hospital revealed a 4.3cm mycotic infra-renal abdominal aneurysm. The patient denied any recent infections or localizing signs of infection. Blood cultures obtained prior to the initiation of broad spectrum antibiotics (vancomycin and meropenem) were negative. Fungal work-up and salmonella antibodies were also negative. A TEE showed no evidence of endocarditis. The patient underwent aneurysm repair with a rifampin-impregnated graft after extensive debridement of pus and inflamed tissue. Intra-operative bacterial, fungal, and AFB cultures were all negative. Six days after surgery, 16S ribosomal RNA came back positive for *Coxiella burnetii*, the causative agent for Q fever. Q fever IgG phase I titer was significantly elevated (4000) and higher than phase 2 titer (2000), indicative of chronic infection. Upon further interview, the patient has been living in a rural valley near Richland Center, WI for several decades downwind of nearby goats and cows and is an owner of horses, cats, and rabbits. Her post-operative course was complicated by wound dehiscence. Upon discharge, she was started on long term therapy with doxycycline and hydroxychloroquine.

Discussion: Chronic Q fever, *Coxiella burnetii* infection lasting more than 6 months, is a rare form of an already uncommon zoonotic infection. After endocarditis, mycotic aneurysm is the second most common manifestation of chronic infection. This case highlights the enduring role of history taking, the importance of understanding host-environmental factors, and the indispensable value of new ribosomal 16S techniques in establishing the elusive diagnosis.

28) DRUG-SEEKING? AN IMPORTANT ALTERNATE CONSIDERATION FOR PATIENTS WITH CHRONIC ABDOMINAL PAIN AND WEIGHT LOSS

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Introduction: Superior mesenteric artery (SMA) syndrome is a rare cause of vomiting and weight loss, characterized by compression of the third part of the duodenum by the SMA and the aorta. Numerous medical and psychiatric problems, including eating disorders, cause an initial rapid weight loss, which results in loss of fat tissue surrounding the SMA and narrowing of the aortomesenteric angle. The patient will have obstructive symptoms, leading to reduced food intake, which further narrows the aortomesenteric angle and perpetuates the process.

Case: A 31-year-old lady with history of Marfan syndrome and narcotic abuse presented with 2-month history of abdominal pain worse with eating, bloating, vomiting, and 40 pounds of unintentional weight loss over 18 months. Prior workup (including EGD, colonoscopy, CT scan of chest and abdomen, gastric emptying study, RUQ ultrasound, and EKG) was unremarkable. Subsequent stool culture, stool ova and parasites, *C. difficile* testing, UA, LFT, TSH, PTH, and TTE were also normal. She was referred to an endocrinologist, who considered opioid-induced suppression of the pituitary-adrenal axis. Her ACTH-stimulation test, however, was normal. She was advised by a gastroenterologist to try erythromycin, nortriptyline, and promethazine, but the medications did not help her symptoms. At this point, her BMI had dropped to 14.2 kg/m². Her gastroenterologist went back over all of her past imaging results, which included a CT scan report that identified “a relatively narrow angle between the abdominal aorta and the SMA.” She was referred to surgery and underwent a duodenojejunostomy. In her postoperative clinic visit, she reported still eating small portions, but for the first time in months, she was able to eat without getting sick or vomiting.

Discussion: The diagnosis of SMA syndrome is elusive for many reasons. Symptoms are insidious, and physical exam and lab findings are non-specific. The diagnosis is made when the aortomesenteric angle is less than 25 degrees, but imaging studies are not specific and other etiologies of bowel obstruction still need to be considered before undergoing surgery. Literature shows that the biggest confounders of diagnosis of SMA syndrome are psychosocial conditions, such as substance abuse and eating disorders. Case series suggest that the presumption of the weight loss having a psychiatric etiology may contribute to a time to diagnosis that can be as lengthy as 18 months from symptom onset. Early recognition of risk factors is important to ensure typical symptoms are not marginalized or ignored in these patients. This will also help in an earlier diagnosis.

29) 'THE FORGOTTEN DISEASE': LEMIERRE'S SYNDROME

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Introduction: Lemierre's Syndrome has an incidence of 3.6 cases per 1 million per year. It is characterized by Jugular vein thrombophlebitis and bacteremia occurring after pharyngeal infection. It mostly affects young healthy adults.

Case description: A 19 year old male presented to the ER with fever, worsening odynophagia, left neck pain for 1 week. On exam, Temp 103.5 F, HR 120 B/M, BP 110/70 mmHg. Head and neck exam revealed tenderness and swelling of the left side of the neck and bilateral tonsillar erythema. Labs revealed WBCs of 14.8 K/mcL with a left shift and platelets of 58 K/mcl. CT scan showed left peritonsillar abscess, left IJ vein thrombosis and pulmonary emboli. Patient was started on intravenous Ampicillin/sulbactam. He underwent left quinsy tonsillectomy then blood culture grew *Fusobacterium Necrophorum*. Patient was diagnosed with Lemierre's disease. His hospital stay was complicated by multifocal pneumonia and Empyema. Repeat ultrasound demonstrated extension of the IJ thrombus necessitating anticoagulation with Heparin. Patient received 2 weeks of IV antibiotics then was discharged on oral Augmentin and Xarelto for 3 months.

Discussion: Lemierre's Syndrome is characterized by IJ thrombophlebitis, bacteremia, metastatic abscesses and septic emboli occurring after Streptococcus pharyngitis. It is mostly caused by normal oral flora with *Fusobacterium necrophorum* being the commonest. This disease was first described in 1936 by Andre Lemierre with a mortality rate of 90%. Its incidence dropped significantly after the advent of antibiotics. However, in the last decade it was reported more frequently possibly due to the limited use of antibiotics in pharyngitis. Many physicians are still unaware of its existence. The mortality of this disease can be 15% when correctly diagnosed and treated. Most useful diagnostic tool is CT scan. Treatment must include a beta-lactam antibiotic or a Carbapenem for 4 weeks with a minimum of 2 weeks IV therapy. Surgery and anticoagulation roles are controversial.

Conclusion: Lemierre's syndrome has been reported more commonly in the last 15 years. High index of suspicion should be maintained in any young patient presenting with non resolving pharyngitis and neck pain followed by pneumonia or sepsis. Extended antibiotic therapy possibly combined with anticoagulation or surgery is the foundation of treatment with low mortality rates being achieved with proper diagnosis and treatment.

30) HYPOGLYCEMIA & LACTIC ACIDOSIS IN A PATIENT WITH MCL

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Introduction: Mantle cell lymphoma (MCL) is a subtype of B-cell non-Hodgkin lymphoma, characterized by overexpression of cyclin D1 due to a t(11:14)(q13;q32) chromosomal translocation. MCL patients are predominantly males with a median age of 65 and typically present with extensive disease involving multiple lymph nodes, spleen, and bone marrow. Relapse usually occurs within 2-3 years and overall survival is less than 5 years. We report a case of newly diagnosed mantle cell lymphoma complicated by refractory hypoglycemia and lactic acidosis.

Case: A 75 year old male with end stage renal disease secondary to hypertension and prostate cancer status post radiation presented with a 2 week history of lymphadenopathy, decreased appetite, chills, night sweats, and weight loss. He denied fevers. Physical exam revealed a cachectic male with non-tender submandibular, anterior cervical, supraclavicular, axillary, and inguinal lymphadenopathy. Initial labs were notable for elevated ESR and ionized calcium, low albumin, and normal WBC. CT of the head/neck and abdominal ultrasound confirmed extensive lymphadenopathy including the retroperitoneal and periaortic regions. Excisional lymph node biopsy was positive for CD5 and cyclin D1, consistent with MCL. On hospital day 5, the patient was transferred to the ICU for increased O2 requirements and altered mental status. An ABG showed respiratory acidosis and a POC glucose of 23 mg/dL. The patient became more responsive immediately after 1 amp of D50. However, his hypoglycemia persisted despite continuous D10 infusion, with finger stick glucose measurements ranging from <20 to 292. Lactate levels subsequently increased to 4.0 and peaked at 4.8. His glucose drip was discontinued on hospital day 11 after his blood glucose stabilized. After starting MCL therapy with rituximab, cyclophosphamide, vincristine, prednisone, and methotrexate, he had no further episodes of hypoglycemia and his lactic acidosis improved. A PET scan 3 months after initial presentation showed a malignant mass in the portal region, but was otherwise negative.

Discussion: This case illustrates an unusual presentation of lymphoma with hypoglycemia coupled with lactic acidosis. One proposed mechanism is the Warburg effect, where tumor cells utilize glycolysis at a high rate even in aerobic conditions, leading to lactic acid fermentation. A 75% mortality rate has been reported in such patients. Treatment of the hypoglycemia with glucose infusion may exacerbate the Warburg effect leading to worsening lactic acidosis. The initiation of chemotherapy usually resolves both of these metabolic derangements as was the case with our patient.

31) DELAYED GEMCITABINE-INDUCED PULMONARY TOXICITY DURING TREATMENT FOR RECURRENT PANCREATIC ADENOCARCINOMA IN A PATIENT WITH LIFRAUMENI SYNDROME

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Introduction: Pulmonary side effects of gemcitabine based therapy are usually mild and self-limiting. Here, we present a case of delayed and life threatening lung toxicity following initiation of treatment in an 81-year-old gentleman with recurrent pancreatic adenocarcinoma.

Case Description: An 81-year-old Caucasian male with LiFraumeni syndrome and concurrent primary malignancies, including diffuse large B cell lymphoma, invasive grade 3 pancreatic adenocarcinoma status post partial pancreatectomy, invasive grade 3 gastric adenocarcinoma, and poorly differentiated esophageal squamous carcinoma, was started on gemcitabine/paclitaxel therapy for recurrence of metastatic pancreatic adenocarcinoma. The patient was admitted to the hospital 10 weeks later for symptoms of dyspnea at rest, chills, night sweats, and severe, generalized weakness for 5 days. On examination, he was afebrile and hypoxic with an oxygen saturation of 68% on room air. Harsh breath sounds and dry crackles were present bilaterally. Laboratory examination showed a white cell count of 6700/ μ l and platelet count of 321,000/ μ l. C-reactive protein and procalcitonin obtained on the day of admission were 13.0 mg/dl (normal: 0-1 mg/dl) and 0.24 ng/ml, respectively. Computed tomography (CT) of the chest revealed extensive ground-glass opacities in a crazy-paving pattern within both lungs, with a predominance in the upper lobes, consistent with acute respiratory distress syndrome (ARDS) or interstitial pneumonia. An infectious work-up, including sputum for Gram-stain and culture, blood cultures for bacteria and fungi, and Legionella urinary antigen, was negative. Cultures from bronchoalveolar lavage were negative for aerobic culture, acid fast bacilli, Pneumocystis jiroveci, and Legionella pneumophila, but grew Candida albicans later on. The clinical presentation was thought to be secondary to gemcitabine-induced ARDS. Following discontinuation of chemotherapy, the patient was treated with high dose steroids and supplemental high flow oxygen therapy. Empiric antibiotics were discontinued on day 3 of admission and he continued to improve on a prednisone tapering dose during the hospital course and upon discharge.

Discussion: This case illustrates the rare side effect of severe pulmonary toxicity associated with gemcitabine-based chemotherapy with increased incidence when combined with bleomycin or a taxane (e.g. paclitaxel, docetaxel). Discontinuation of the drug and supportive treatment along with steroids and/or diuretics remain the mainstay of management.

32) UNUSUAL PRESENTATION OF ANTI-GBM DISEASE WITH LINEAR IGA STAINING

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Introduction: Anti-glomerular basement membrane (anti-GBM) disease is an aggressive form of glomerulonephritis usually mediated by circulating IgG autoantibodies to the non-collagenous (NC1) domain of alpha3 subunit of Type IV collagen. There have been few case reports about an IgA variant of anti-GBM disease.

Case Report: A 72-year-old white male with a history of hypertension presented with one episode of gross hematuria and abdominal pain. He was found to have acute kidney injury (AKI) with a creatinine (Cr) of 2.7 mg/dL (baseline 0.9-1.0 mg/dL), which rapidly increased to 5.6 mg/dL over 3 days. His urinalysis was significant for 1+ protein and 2+ hematuria. Complement levels were normal. Serologic studies were negative for anti-nuclear antibodies, anti-neutrophil cytoplasm antibodies, cryoglobulins, hepatitis B and C panels, and anti-GBM antibodies. Renal biopsy yielded 3 glomeruli with cellular crescents and fibrin deposits. Immunofluorescence revealed glomerular capillary walls with 3+ linear staining for IgA. Electron microscopy revealed a mild increase in mesangial matrix, normal cellularity, and normal thickness of the glomerular basement membrane with no electron-dense deposits. The patient was treated with IV methylprednisolone for 3 days, then transitioned to oral prednisone, cyclophosphamide, and received a 6-week course of plasmapheresis. His renal function improved gradually from 5.6 mg/dL to 3.2 mg/dL after 3 weeks of treatment.

Discussion: We report a case of anti-GBM disease with linear staining for IgA; thus far, there are only 13 other reported cases in the literature. Outcomes of these cases varied, although 5 of the 13 went on to develop ESRD, and only 3 of the 13 survived with Cr of <2. Pulmonary hemorrhage (PH) was absent in our patient; only five of the 13 other cases (38%) had PH, lower than the reported 60% of patients with co-existent pulmonary and renal disease in anti-GBM disease. The antigenic targets of IgA antibodies remain poorly defined, but the heterogeneity of anti-GBM disease demonstrates the necessity of renal biopsy for reliable diagnosis of this atypical presentation, as serological assays are likely to give false negative results.

33) CENTRAL VENOUS CATHETER PLACEMENT: COMPLICATIONS IN THE INTENSIVE CARE UNITS VS MEDICAL-SURGICAL FLOORS

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Purpose: To decrease admissions to the intensive care unit (ICU) for the purpose of placing central venous catheters (CVCs); to reduce the use of unnecessary peripherally inserted central catheters (PICCs).

Description: Our institution requires CVC placement to be within an ICU to have increased monitoring of the patient throughout the procedure. We propose that with the use of ultrasound guidance, standard precautions, and standardized training, CVC insertion outside the ICU carries no increased risk of complications. To accomplish our objective, we start by comparing major and minor complication rates associated with CVC placement between two hospitalist led procedure teams, one in the ICU setting and one on Medical Surgical Floors.

Results: We compared 127 CVC placements performed at John H. Stroger Hospital to 15 CVC placements at Froedtert & the Medical College of Wisconsin between March 2012 and January 2014. There was one major and one minor complication reported in the Medical-Surgical floor setting and no complications reported in the ICU. Both complications arose from internal jugular CVC placement. Average values were similar between both hospitals aside from a creatinine of 6.33 (Std Dev: 5.8) at John H. Stroger Hospital compared to 1.01 (Std Dev: 0.85) at Froedtert & the Medical College of Wisconsin.

Conclusions: Review of current literature shows no objective data to indicate a benefit to placing CVCs in an ICU compared to medical surgical floors. In this study, we showed that placement of CVCs on Medical-Surgical floors can be safely performed when under the supervision of an experienced hospitalist. We recommend CVC placement on the floor can be safely used to decrease PICCs.

34) SEVERE CONTACT DERMATITIS MASQUERADING AS BULLOUS PEMPHIGOID

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Introduction: Allergic contact dermatitis (ACD) is a presentation of T cell mediated delayed hypersensitivity reaction. Presentation can be acute, after an exposure to the allergen, or can be chronic, after chronic exposure to a given allergen. Here, we present a case of a patient with severe contact dermatitis that on physical exam appeared as bullous pemphigoid. The objective of the case is to identify allergic contact dermatitis as a cause of a bullous and vesicular rash.

Case: 60 year old female with history of rheumatoid arthritis currently on methotrexate presenting with pruritic erythematous patches with vesicles. Two weeks prior to the onset of rash, patient began using lavender/argan oil cleanser. Symptoms began with isolated labial swelling which progressed to swelling, redness and itching along the anterior aspect of both thighs. Cortisone cream provided moderate relief. Prior to admission, she was prescribed cephalixin for presumed cellulitis. Upon starting this, blisters and vesicles formed on anterior thighs bilaterally which ruptured and drained serous fluid. The rash spread to abdomen, web space of fingers and left axilla. Patient discontinued cephalixin due to concern for drug-induced dermatitis. On exam patient had bright pink partially blanchable erythematous patches studded with vesicles and bullae weeping serous fluid on the anterior and inner thighs and pink patches in the bilateral axillary folds and web spaces of the fingers. Nikolsky sign was negative. Dermatology was consulted given concern for bullous pemphigoid. A biopsy revealed findings consistent with an acute exematoid hypersensitivity reaction likely due to contact dermatitis or drug eruption. The lavender/argan oil cleanser was discontinued and she was started on high dose prednisone with a three week taper. At discharge, pruritus was resolving and rash was improved.

Discussion: ACD classically presents with erythematous, indurated scaly plaques. However, vesicles and bullae may be seen in severe cases. Edema can be prominent in areas with thin skin such as genitalia, lips and eyelid. Rash is usually localized to areas in contact with allergens but patchy diffuse distributions can be observed. Treatment involves identification and avoidance of offending allergen and topical corticosteroids are the first line treatment for localized ACD. For ACD which involves greater than 20% of the body surface area or involving face, hands, feet or genitalia systemic corticosteroids are indicated. It is important to consider ACD as a cause of skin rashes involving vesicles and bullae and punch biopsy can be a useful tool in evaluating its etiology, as contact dermatitis can progress to skin necrosis if untreated.

35) DAPTOMYCIN-INDUCED EOSINOPHILIC PNEUMONIA

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Introduction: Eosinophilic pneumonia (EP) is characterized by lung infiltrates on chest x-ray, increased eosinophils in the lung parenchyma or airway and may include peripheral eosinophilia. Inciting causes include parasites or fungal infection, drug ingestion or idiopathic causes. Common drugs associated with EP include daptomycin, ACE inhibitors, amiodarone, isoniazid, hydralazine, methotrexate, penicillin and sulfa drugs. Clinical presentation generally involves fever, cough, dyspnea, urticarial or papular rash. The objective of this case is to recognize the association between EP and daptomycin.

Case: 81 year old male with a history of BPH status-post TURP complicated by MRSA bacteremia treated with IV daptomycin infusions developed eosinophilia discovered on routine labs (WBC 9.4 with 35% eosinophils). Daptomycin was discontinued and he was switched to IV vancomycin six days prior to admission. Due to inadequate vancomycin trough levels he was switched back to daptomycin.

On the day of admission, his wife reported the patient had shaking chills with altered mentation, a non-productive cough and temperature of 100.3F. Laboratory findings were significant for WBC 18.4, neutrophils 79%, eosinophils 12%. UA was positive for 3+ leukocyte esterase and bacteria. Blood cultures were negative.

On chest x-ray he was found to have a new left lower lobe infiltrate that was not present on CXR from prior hospitalization; given that the infiltrate developed after initiation of daptomycin therapy, the pneumonia was presumed to be daptomycin-induced eosinophilic pneumonia. Daptomycin was discontinued and he was started on vancomycin, cefipime and ciprofloxacin for empiric coverage of the MRSA bacteremia, pneumonia and UTI. Pulmonology and ID were consulted for management and deferred invasive diagnostic procedures such as bronchoscopy or BAL since the patient improved clinically. WBC count and eosinophil count continued to downtrend over the course of his hospital stay.

Discussion: Eosinophilic pneumonia (EP) should be suspected in patients with new lung infiltrate and either peripheral eosinophilia, BAL eosinophilia, or lung tissue biopsy eosinophilia. In vitro studies which have shown that daptomycin interacts with pulmonary surfactant, inhibiting antimicrobial activity and causing accumulation of the drug. This may cause epithelial injury. Clinical improvement is usually observed with removal of offending agent. However, in cases where clinical symptoms persist after discontinuation of the drug corticosteroids may be indicated. Given the potential for pulmonary toxicity and progression to respiratory failure by drugs such as daptomycin, early recognition and consideration for corticosteroid therapy of EP is imperative.

36) WHAT'S GROWING IN THAT PLEURAL FLUID??

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Introduction: A Plasmacytoma is a discrete, solitary mass of neoplastic monoclonal plasma cells in either bone or soft tissue. The upper aerodigestive tract is the most common site for extramedullary plasmacytoma, pleural involvement is extremely rare. A patient who does not have diagnosis of MM before may present with pleural involvement, and the involvement may be bilateral.

Case Presentation: 42 yo male presented after a fall with complaints of headaches, dizziness and myalgia. He also reported a tick bite prior to the onset of symptoms. Creatinine was mildly elevated at 1.31 with a normal GFR. He was mildly anemic with hemoglobin of 10.7. A febrile tick panel was found to be positive for Anaplasma. He was treated with doxycycline for 10 days. After a period of initial improvement, his symptoms recurred, with persistent headaches, blurriness of vision and myalgias, for which he sought medical attention. Further lab work revealed hemoglobin of 10, LDH of 474, C-reactive protein of 5.51, calcium of 10.5, albumin of 4.6, ESR of 33 and creatinine worsened to 1.72 with a GFR of 44. SPEP showed elevated monoclonal lambda protein at 0.1. Serum lambda free light chains were high at 263 mg/dl; 24 hour urinary protein was >11.5 gm and beta 2 microglobulin 8.95. Chest x-ray showed bilateral pleural effusions with a suspicious right lung nodule. CT chest showed bilateral pleural effusions with multiple pleural based nodules. His pleural fluid smear was studded with multinucleate neoplastic plasma cells and Pleural fluid cytology showed neoplastic plasma cells consistent with plasma cell myeloma with lambda light chain restriction. Peripheral smear was negative for plasma cells. Skeletal survey showed multiple lytic lesions. PET showed diffuse marrow involvement. Bone marrow showed high cellularity with plasma cells involving 80 % of the marrow. He was started on steroids and Bortezomib and is awaiting autologous hematopoietic cell transplant.

Discussion: Plasmacytomas can have varying presentations. In our patient, pleural fluid analysis confirmed the diagnosis of Myelomatous Pleural effusion with pleura based plasmacytomas. Serum albumin and beta 2 microglobulin help stage the disease and predict prognosis. Our patient was found to have Stage III disease which carries a 29 month survival without treatment. FISH and other genetic studies further help in Risk stratification. Our patient had t(1;16), t(6;14), t(8;14;20) and t(18;14) placing him in the high risk category.

37) HYPOXEMIC RESPIRATORY FAILURE FOLLOWING INITIATION OF HIGHLY ACTIVE ANTIRETROVIRAL THERAPY IN HIV/AIDS PATIENT

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Introduction: Despite the advent of Highly Active Antiretroviral Therapy (HAART), *Pneumocystis jirovecii* pneumonia (PCP) remains the most common opportunistic infection in the United States in patients with HIV/AIDS with CD4 counts <200 cells/ μ L. We present a case of a 23-year old male with HIV/AIDS presenting with severe PCP after recent initiation of HAART.

Case: 23-year-old male with HIV/AIDS (CD4 109/15% and viral load 128,215 copies/mL on admission, down from 881,018 copies/mL at diagnosis) diagnosed one month prior to admission presented with three days of worsening myalgias, dyspnea, and fevers. The patient had started HAART (Efavirenz/Emtricitabine/

Tenofovir) 10 days prior to admission. Social history was significant for MSM and employment as a flight attendant with recent travel to Texas, California, and Mexico. Vital signs on admission were significant for blood pressure of 89/62 mmHg, heart rate of 126/minute, and temperature of 39.1 degrees Celsius. Examination was significant for bilateral bronchial breath sounds and accessory respiratory muscle use with nasal flaring. Arterial blood gas on room air showed a pH of 7.54, pO₂ of 57 mmHg, and pCO₂ of 29 mmHg. Chest radiograph (CXR) demonstrated extensive bilateral patchy infiltrates with ground glass opacities. The patient was treated for septic shock with intravenous fluid boluses, required a continuous norepinephrine infusion, and was intubated for hypoxemic respiratory failure. Therapies for both PCP and community-acquired pneumonia (CAP) were started. On hospital day 3, direct fluorescent antigen from bronchoscopy returned positive for PCP and antimicrobials for CAP were stopped. Other bacterial, fungal, and viral cultures were negative. Mechanical ventilatory support was discontinued on hospital day 4. HAART (Dolutegravir/Emtricitabine/Tenofovir) was restarted on hospital day 5. The patient was discharged home on hospital day 9 on treatment doses of Trimethoprim-Sulfamethoxazole (TMP-SMX) and prednisone for a total of 21 days. One month after discharge the patient is doing well (CD4 281/26% and viral load 494 copies/mL) and is returning to work as a flight attendant.

Discussion: Risk factors for PCP include CD4 <200 cells/ μ L, CD4 cell percentage $<14\%$, and HIV RNA level $>100,000$ copies/mL. The most common clinical manifestations are subacute onset of dyspnea, fever, and non-productive cough. CXR typically shows diffuse bilateral interstitial infiltrates emanating from the hila. The preferred therapy is TMP-SMX for 21 days and adjunctive prednisone for patients with moderate-to-severe disease, defined by room air pO₂ <70 mmHg or alveolar-arterial oxygen gradient ≥ 35 mmHg. The differential diagnosis in this patient included Immune Reconstitution Inflammatory Syndrome (IRIS), defined as paradoxical worsening of pre-existing infections (in this case PCP) within weeks to months of starting HAART in HIV-infected patients.

38) A CASE OF IDIOPATHIC TRACHEAL STENOSIS

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Introduction: Idiopathic tracheal stenosis is a rare condition. Around 5% of cases of upper airway narrowing (subglottic stenosis) are due to an unknown cause (Idiopathic). ISS occurs almost exclusively in women, with only a small number of cases reported in men. It usually occurs between 30-50 years of age, but has been known to occur in a population ranging from children to elderly patients. There are various theories for etiology of ITS including chronic GERD, chronic cough causing microscopic trauma, hormones, but none have been conclusively proven. We present a case of a young 24 y/o Female with idiopathic tracheal stenosis.

We report a case of idiopathic tracheal stenosis in a 24 year old female patient who presented with complaints of worsening dyspnea, stridor and pleuritic chest pain. Pulmonary function tests revealed a FEV1 (50-70% predicted) and a reduced FEV1/FVC with markedly blunted inspiratory and expiratory flow loops, findings consistent with obstructive lung disease in the spirometry. Lack of response to beta agonists was suggestive of fixed airway obstruction. Computed tomography of chest and neck showed nodular, circumferential thickening of the upper trachea extending for approximately 2.5 cm and causing areas of 50% or greater stenosis. Suspension micro laryngoscopy revealed severely inflamed subglottic/ superior tracheal mucosa with friable inflammatory debris. Biopsy specimen revealed ulcerated squamous mucosa with nonspecific inflammatory changes.

Treatment: steroid injection with balloon dilation provided immediate relief. Workup for rheumatologic causes, GERD, infectious causes, malignancy was all negative. She had to undergo repeat dilatation about a year after the first one with relief of symptoms. She was also treated with oral steroids and methotrexate, both of which were eventually tapered off.

Conclusion: ITS is a uncommon cause of dyspnea in a young adult. Common presentation would be dyspnea on exertion, unresponsive to inhalers, stridor, and fatigue. PFT with blunted inspiratory and expiratory curves with lack of response to beta agonist is indicative of ITS. When these findings are present otolaryngology must be consulted for further evaluation and treatment.

39) A CASE OF ACONITE TOXICITY WITH BIDIRECTIONAL VENTRICULAR TACHYCARDIA .

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Introduction: Aconite, derived from the root of the aconitum plant, is used throughout eastern Asia, including India and China, as an analgesic and an anti-inflammatory agent. Serious neuro- and cardiotoxic effects can occur if taken at a high dose. Aconite toxicity is common in Southeast Asia, but rare in the U.S.

Case: A 62-year-old Hmong man with no significant cardiac history came to the emergency department with chest pain, dizziness, and palpitations. Systolic blood pressure was 60-80 mmHg, and heart rate was 150-220 bpm. Cardiac rhythms were variable with runs of ventricular tachycardia (VT) and supraventricular tachycardia, couplets with right bundle branch block-like morphology, and left axis deviation suggestive of left posterior fascicular origin. Multiple cardioversions did not resolve his arrhythmias. Amiodarone load was given, also without improvement. Signs compatible with coronary ischemia and lactic acidosis prompted coronary angiography and ventriculography, which showed nonobstructive coronary atherosclerosis with slow coronary flow and hyperdynamic left ventricular function with normal wall motion. The patient's wife told an interpreter that the patient had taken multiple doses of decoction made from aconitum roots for stomach upset before having cardiac symptoms. Literature review confirmed symptoms compatible with aconite toxicity. Over 24 hours the patient improved both hemodynamically and electrically. Supportive care was weaned successfully over 48 hours. Discharge ECG was identical to an ECG obtained 3 years prior when he presented with mild chest pains.

Mechanism of toxicity: Aconite interacts with the voltage-dependent sodium channel present on cell membranes of excitable tissues, including the myocardium, striated and smooth muscle, and neurons nerves, altering membrane depolarization and repolarization.

Management: Mainly supportive. Vasopressor support and cardiopulmonary bypass have been used to maintain perfusion in refractory shock. Ventricular arrhythmias are often refractory to cardioversion and only minimally responsive to antiarrhythmic like sodium channel blocking agents

Conclusion: Aconite poisoning may be on the rise in the U.S. with a growing Asian population in which use of alternative medicine is high. Our case illustrates that herbal remedies may result in critical illness and highlights the need to recognize the signs and side effects of alternative remedies used in the locale. This case also highlights the importance of having interpreter services in emergency services to overcome language barriers, especially for locally prevalent languages

40) LEFT UPPER QUADRANT PAIN AND SPLENOMEGALLY IN A SICKLE CELL PATIENT

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Introduction: Acute splenic sequestration crisis (ASSC) is a disease associated with sickle cell patients in their first decade of life. There are a few reports of ASSC occurring later in life with sickle cell beta thalassemia patients.

Case: A 30 year old African American gentleman with sickle cell beta and pancreatic pseudocyst presented with a sore throat and diffuse achy pain. Physical exam showed tenderness to palpation of 1st left lower molar and a 2x2 cm firm nonfluctuant indurated right cheek mass. Abdominal exam showed tenderness in the left upper quadrant. CBC, BMP, and CXR were within normal limits other than patient's chronic anemia. Patient was admitted for pain crises and was started on penicillin for the cheek abscess. On the subsequent day an incision and drainage was done on the indurated area of the right cheek and antibiotics were broadened to Vancomycin, Piperacillin and Tazobactam.

Patient continued to spike fevers and on day 3 there was a sudden drop in hemoglobin to 5.5 g/dL with a severe leukocytosis of $23.5 \times 10^3/\mu\text{L}$ and thrombocytopenia. Labs showed rises in liver function tests, alkaline phosphatase, reticulocyte percentage count and bilirubin. Patient was immediately transfused three units of packed red blood cells. Hemoglobin stabilized back to previous level over the next few days. A CT scan with contrast of abdomen and pelvis showed a markedly enlarged spleen. Calculated spleen volume increased from 412 cm³ three months ago to 726 cm³. Leukocytosis continued to worsen and doxycycline was added to the antibiotic regimen. White blood counts began to improve. A complete infectious workup was negative and patient was discharged on doxycycline.

Discussion: Acute splenic sequestration (ASSC) was diagnosed based on the acute and significant increase in size of the spleen, along with the sudden drop in hemoglobin and platelet count. There are very few reported cases of ASSC in adults with sickle cell beta thalassemia. The mean age of ASSC in this patient group was 29 years of age. Mortality of ASSC in adult sickle cell beta thalassemia was 33% based on reported cases. The significant morbidity and mortality associated with ASSC require clinicians to have a high index of suspicion and prompt management. All the patients who survived ASSC and who did not have a splenectomy had a recurrence of ASSC. Though splenectomy was not done in our patient, prophylactic splenectomy in this subgroup of patients may be considered.

41) SPONDYLODISCITIS HIDDEN BEHIND THE C-REACTIVE PROTEIN

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Introduction: Spontaneous infectious spondylodiscitis (SIS) (also known as vertebral osteomyelitis) is a rare form of osteomyelitis, with over 50% of cases due to staphylococcus aureus. Patients often present solely with back pain, without any other common signs of infection, such as fever or leukocytosis. C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) are highly sensitive tools that can help guide diagnosis and treatment.

Case Report: A 71-year-old male with no pertinent past medical history presented to our outpatient clinic with severe, debilitating back pain. Three weeks prior at an outside hospital he was evaluated for acutely worsening back pain and fever, with blood cultures positive for Klebsiella pneumonia. At that time, MRI of lumbar and thoracic spine revealed no clear etiology of back pain. He was noted to have elevated ESR and CRP. Infectious work-up and additional imaging failed to identify the source of bacteremia. After a week of treatment with piperacillin/tazobactam, his inflammatory markers trended down and subsequent blood cultures were negative. The patient was discharged on ciprofloxacin and pain medications.

At outpatient follow up a week later, the patient reported worsening back pain with limited mobility. Vital signs were within normal limits, and the patient's exam demonstrated no neurological deficits. CRP was markedly elevated from discharge, but fevers and leukocytosis were absent. He was re-admitted, and a repeat MRI of lumbar spine demonstrated spondylodiscitis of L2 through L3. Repeat blood cultures were negative and echocardiogram showed no valvular vegetations. He was started on a six-week course of ceftriaxone to cover for Klebsiella, given the initial positive blood culture.

Discussion: SIS due to Klebsiella pneumonia is relatively rare. ESR and CRP are elevated in 98% and 100% of cases of SIS, respectively. Often, back pain and increased CRP may be the only markers of disease. Imaging is required for diagnosis, with MRI being more sensitive than CT for early detection. In the absence of positive blood cultures, a biopsy is required prior to initiating treatment in otherwise stable patients. There are no established guidelines for length of treatment; however patients are typically treated with intravenous antibiotics for at least 6 weeks. This case highlights the challenges of early diagnosis of SIS. Though not specific, CRP is a highly sensitive tool for identifying infections, such as SIS. Early diagnosis may help prevent significant sequela, such as chronic back pain or even sepsis.

42) HENOCH-SCHÖENLEIN PURPURA WITH RAPID PROGRESSION TO RENAL FAILURE IN AN ADULT PATIENT

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Introduction: Henoch-Schöenlein Purpura (HSP; also called IgA Vasculitis) is a leukocytic vasculitis which primarily affects small vessels. Clinical presentation involves palpable purpura without thrombocytopenia and frequently, GI and joint involvement. However, renal damage is the primary source of long-term morbidity and mortality. While HSP is well known as a pediatric disease, much less is known about adult HSP leading to a problematic lack of evidence based treatment options for use in clinical practice. Adult HSP is believed to have a higher rate of long-term complications including progression to renal failure. It also appears to be associated with malignancy. Renal impairment, ranging from microscopic hematuria to nephrotic syndrome, is detected in 45-85% of cases, with risk of progression to renal insufficiency in 30% of patients. There are few concrete data available for adult HSP including the rate of progression to renal failure.

Case: We discuss a 23-year-old woman who presented with 10/10 abdominal pain and associated anorexia, refractory to naproxen, APAP, or PO opioids. Review of systems was positive for diarrhea, hematuria, myalgias, and progressive lower extremity skin lesions. Of note, the prodrome also included sore throat. After many visits to the ED for these symptoms, Rheumatology evaluation led to diagnosis of HSP. Approximately two weeks later, she had no improvement in her symptoms but her creatinine was 3.58, from a value of 0.66 ten days prior. Renal biopsy was significant for proliferative/focal necrotizing and crescentic IgA nephropathy, ultimately confirming the diagnosis of HSP. The patient was started on IV methylprednisolone for 3 days before switching to PO prednisone 80 mg/d. Her abdominal pain incrementally improved with progressive PO tolerance and her creatinine improved to 2.09 by time of discharge.

Discussion: A striking feature of this case was the rapid onset of renal involvement in a patient with no kidney disease at time of HSP diagnosis. Renal involvement almost always begins within 6 months in children and most often within 4 weeks. In children, at least, the American Academy of Family Physicians recommends monthly monitoring via UA, with some investigators advocating for weekly screening; our case suggests that this may be applicable to adult HSP as well.

Supportive therapy is the mainstay of HSP therapy. Treatment of adult HSP nephritis is generally not evidence based, due to the paucity of randomized controlled trials. Nearly all clinicians use corticosteroids for symptomatic relief and in cases of severe renal disease although their benefit is unclear. The small clinical trials performed to date do not support the use of cyclophosphamide in adults. Overall, few options exist when encountering this frustrating and sometimes life-threatening disease.

43) AN UNUSUAL CASE OF FEVER

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Fever of unknown origin (FUO) frequently requires a multispecialty approach guided by patient symptomatology.

A 65-year-old female was referred to our institution's hepatology clinic for evaluation of FUO. She had a long history of rheumatoid arthritis (RA) previously treated with various immunosuppressant medications including methotrexate, infliximab, prednisolone. Eight months prior to presentation, she began to experience fevers up to 103°F (39.4°C), sweats, chills, nausea, vomiting, and a persistent dull epigastric abdominal pain. Since the onset of symptoms, she had significant anorexia resulting in a 30-pound weight loss. Her symptoms seemed to follow a waxing and waning pattern over three day cycles. She underwent a thorough work-up at a local community hospital including laboratory and imaging studies, an inguinal lymph node biopsy, a bone marrow biopsy, two colonoscopies, an EGD, and a liver biopsy. Of these, the most remarkable finding was the ultrasound-guided liver biopsy showing chronic inflammation of the portal tracts and a granuloma, which prompted referral to our center. On arrival, she was found to be febrile with physical exam findings consistent with long-standing RA but was otherwise normal. A multispecialty diagnostic approach was pursued. Laboratory findings showed isolated elevations in alkaline phosphatase (439 U/L) and gamma glutamyltransferase (452 U/L), negative autoimmune serologies (including ANA and anti-mitochondrial antibody M2), and normal immunoglobulin levels (including IgG4). A thorough infectious work-up was unrevealing. Ultimately, MRCP followed by ERCP was performed and showed findings consistent with primary sclerosing cholangitis (PSC). She was started on low-dose ursodiol and high-dose prednisone for possible overlap syndrome. Her symptoms markedly improved within days and her laboratory markers normalized.

PSC is a form of cholestatic liver disease without well-defined medical therapies. Here we present a case of glucocorticoid-responsive PSC in the setting of a patient on chronic immunosuppressant therapies for RA.

44) A RARE CASE OF ANAPLASTIC SMALL BOWEL CARCINOMA

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Introduction: Small bowel carcinoma is a rare disease and requires a high index of suspicion and clinical knowledge for prompt diagnosis and treatment. Small bowel cancers account for only 3% of gastrointestinal (GI) malignancies but are responsible for worse outcomes than colorectal cancer.

Case: A 65-year-old man with a past medical history of hypertension, diabetes, hyperlipidemia, gastroesophageal reflux disease and depression presented with intermittent exertional chest pain and dyspnea. He also reported having black stools, abdominal pain and constipation for two weeks. He had been taking ibuprofen for back pain relief and had a colonoscopy in 2010, which was positive for a hyperplastic polyp and diverticulosis. The patient also had a 45-pack-year smoking history and drank 2 alcoholic beverages per night. Initial physical examination was positive for pale skin and melena. Initial laboratory studies revealed a hemoglobin of 4.8 g/dl, blood urea nitrogen of 20 mg/dl, and creatinine 1.12 mg/dl. The patient was started on IV pantoprazole and transfused with red blood cells. Upper endoscopy revealed mild antral erythema and no ulcerations. Subsequent colonoscopy revealed normal colonic mucosa, mild diverticula in the sigmoid colon, and green/black liquid in the ascending colon and exiting the ileocecal valve. Having found no source of GI bleed an abdominal CT was ordered and revealed asymmetric focal wall thickening in the proximal small bowel and enlarged adjacent lymph nodes but no bowel obstruction. The patient then underwent upper endoscopy with a pediatric colonoscope which identified 2 large friable masses in the proximal jejunum. Surgical intervention was then performed and the distal duodenum was resected from the 3rd portion to the mid-jejunum. Pathology confirmed poorly differentiated adenocarcinoma with limited lymph node involvement.

Discussion: Small bowel cancers account for only 3% of GI malignancy and is primarily caused by carcinoid tumors, adenocarcinoma, GI stromal tumors, and lymphomas. However, anaplastic small bowel carcinoma is a rare variant with only 20 cases in the English literature. The incidence of small bowel carcinoma in the US has doubled in the last 30 years and causes almost 1100 deaths per year. Obstruction, hemorrhage, and metastasis are the major complications of small bowel carcinoma accounting for many of the symptoms the patient will present with. Patients usually present with insidious clinical signs, including abdominal pain, weight loss, anemia, and melena. Diagnosis involves various methods of endoscopy with biopsy, CT, capsule endoscopy, and biological markers. Patients are usually managed surgically with resection of the effected bowel, and adjunct chemotherapy and radiation if metastasis is found. Prognosis of small bowel adenocarcinoma is poor overall (approximately 10%), with 3-year survival dependent on stage of tumor.

45) A RARE CASE OF PNEUMATOSIS INTESTINALIS

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Introduction: The presence of gas in the wall of the small or large intestine is referred to as pneumatosis intestinalis (PI). There are multiple theories as to the pathogenesis of this condition. Several diseases are associated including: intraabdominal catastrophes, infections, and immunologic disturbances. When associated with bowel necrosis, PI may have high mortality rates.

Case: This patient is a 62-year-old female with a diagnosis of ovarian cancer involving the left ovary, uterus, colon and mesentery. Initial surgical management included bilateral salpingoophorectomy, hysterectomy and sub-total colectomy. One month post-op, she was treated with IV paclitaxel, IP paclitaxel and IP cisplatin. Nausea and vomiting prophylaxis included dexamethasone. Two months following the induction of chemotherapy, the patient developed diarrhea and belching at home and was later found unresponsive by her husband. The patient was transported to the ED. She was minimally responsive, hypotensive, tachycardic and hypothermic at 34.1 C. Physical exam was significant for dry oral mucosa, clear lungs, non-tender abdomen and no focal neurologic deficits. Initial investigations included a negative CT-head and chest x-ray revealing an air filled esophagus and dilated stomach. Labs revealed an anion gap of 24, WBC-24 and lactate- 9.9. The patient passed bloody stool in the ED and was admitted to critical care with a consult to gastroenterology. Abdominal x-ray revealed marked dilation of the stomach. EGD was performed with the findings of esophagitis, severe gastritis and duodenal ulceration suggestive of mucositis and inflammation. A CT-abdomen was recommended, revealing pneumatosis involving the stomach, duodenum and most of the small bowel consistent with bowel necrosis. In the ICU, the patient quickly improved with conservative management. Her acute GI bleed resolved and lactate normalized within 24 hours. She received a 7 day course of piperacillin-tazobactam and was discharged home on hospital day 10.

Discussion: Pneumatosis intestinalis and gastric pneumatosis are rare but documented complications of immunological disturbances secondary to steroids or chemotherapy. Corticosteroids are the most common drugs associated with PI. There has been one case reported of PI during paclitaxel treatment and one case reported involving cisplatin in combination with irinotecan. Although the exact pathogenesis of this case may never be known, it adds to the collective data of documented causes of PI and side effects of these specific chemotherapeutic agents.

46) CO-PRESENTATION OF SARCOIDOSIS AND SJÖGREN'S

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Introduction: Sarcoidosis is a multi-system granulomatous disorder of unknown etiology. It typically presents with bilateral hilar adenopathy, pulmonary reticular opacities, and lesions of skin, joints, and/or eyes. Sjögren's syndrome is a chronic inflammatory autoimmune disorder resulting in decreased lacrimal and salivary gland secretions which can also affect lungs and lymph nodes. We describe a young man with both Sjögren's syndrome and sarcoidosis and reviewed the medical literature to determine whether the coexistence of these relatively rare diseases had previously been described.

Case: A 34-year-old Caucasian man was diagnosed with Sjögren's syndrome in 2007, when he presented with dry mouth, dry eyes, joint pain, poor dentition and fatigue. The diagnosis was supported by a positive labial biopsy, and significant SSA and ANA titers. He was treated with hydroxychloroquine, methotrexate, cyclosporine eye drops, and tear duct plugs. In September 2013 he returned from a trip to Albania with cervical lymphadenopathy and anterior pleuritic chest pain associated with cough and shortness of breath. A chest CT, bronchoscopy, endobronchial ultrasound, and biopsy showed granulomatous inflammation consistent with sarcoidosis. He was referred to our rheumatology clinic for further evaluation. Quantitative SSA/SSB confirmed the diagnosis of Sjögren's syndrome. We felt he most likely had both Sjögren's syndrome and sarcoidosis, but was most symptomatic currently from his sarcoidosis. As a steroid sparing agent, he was started on azathioprine. He continues to take 15mg/day of prednisone to control his pulmonary symptoms.

Discussion: A review of the literature found dozens of case reports describing a similar coexistence of Sjögren's syndrome and sarcoidosis. One recent meta-analysis described 59 such cases, but deemed true coexistence in only 28 cases. Several possible explanations exist for the coexistence of sarcoidosis in patients with Sjögren's syndrome. Perhaps the treatment for Sjögren's could predispose a person to sarcoidosis. This is less likely since we do not see an increase in sarcoidosis in patients similarly immunosuppressed for other rheumatologic conditions. Both Sjögren's syndrome and sarcoidosis are inflammatory diseases with a Th1-like cytokine secretion profile and they could share a pathogenetic mechanism. Several groups have postulated that sarcoidosis is related to an inhaled environmental antigen. Yet another possibility is that the dryness associated with Sjögren's syndrome decreases the effectiveness of mucosal barriers and could promote antigen access to the lungs. Further studies of the pathogenesis of both these diseases may shed light on the rare but fascinating co-occurrence of these two syndromes.

47) MARKERS FOR PERFORMANCE STATUS IN CHART REVIEW OF LUNG CANCER PATIENTS

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Background: Retrospective analyses are useful to conduct comparative effectiveness research. Most are conducted using databases such as the SEER Medicare Database or the VA Cancer Registry. The advantage of the VA Cancer Registry is that it includes clinically important variables, such as lab data, vital signs and comorbidities. We have previously established that baseline hemoglobin, albumin and weight loss obtained from this database are significant factors associated with overall survival in patients with lung cancer. However, to confirm whether anemia, albumin and weight loss accurately predict performance status a chart review was needed.

Methods: As part of a larger study, hemoglobin, creatinine, albumin, and weight change were recorded on a sample of Stage III and Stage IV NSCLC patients. One of the authors (AM) conducted a chart review of 281 patients to determine the performance status before starting chemotherapy.

Results: A multivariate analysis of overall survival was conducted which included hemoglobin, weight loss, albumin, EGFR and performance status. Performance status was found to have an independent effect when added to these variables on multivariate analysis.

Discussion: Variables such as anemia, albumin, and weight loss are useful to stand in for how sick cancer patients are when conducting retrospective chart reviews. This is especially useful when using the VA Cancer Registry as these variables can be obtained without reviewing each individual chart allowing for a larger sample size. However, as performance status had an independent effect on overall survival in multivariate analysis, a study including performance status would require individual chart review.

48) SEVERE HYPOCALCEMIA AS INITIAL PRESENTATION OF CELIAC DISEASE

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Introduction: Celiac disease can present with typical gastrointestinal symptoms (diarrhea, steatorrhea, weight loss, bloating, flatulence, abdominal pain) or non-gastrointestinal abnormalities. Many individuals may not have any symptoms at all and can present later in life.

Case Presentation: A 58 yo male was referred to emergency department after routine lab showed severe hypocalcemia. Patient had perioral tingling, numbness, metallic taste, muscular cramping, 10 pounds weight loss for three months. No history of diarrhea, flatulence or abdominal pain. On physical examination, initial vital showed BMI 20.6 temperature 98.2 PR 78 BP 181/88 RR 18 SpO2 100%. Physical examination was unremarkable. HB was 10.9 with RDW of 16.6. Total and ionized calcium were 5.3 (L) and 0.71 mmol/L (L) respectively while phosphorus was 5 (H). PTH was 217 (H) and vitamin D-25 measured at 4.0 (L). Patient was worked up for secondary hypoparathyroidism. Renal function test was normal. Transaminases and alkaline phosphatase were elevated. Folate, iron and ferritin level were low. Urine calcium level was normal. Celiac panel showed high Gliadin Ab IgA, Gliadin Ab IgG, and TTG IgA which were consistent with celiac disease. The diagnosis was confirmed with biopsy from the duodenum that showed villous atrophy with infiltrating lymphocytes. ECG showed prolonged QT. Patient was treated with calcium gluconate, calcitriol with oral calcium and vitamin D supplementation. He showed improvement of all symptoms and correction of calcium level. Bone density scan showed osteoporosis.

Conclusion: We recommend celiac disease should be considered as the cause of hypocalcaemia with secondary hyperparathyroidism from vitamin D deficiency despite lack of GI manifestations. Patient with celiac disease presenting with hypocalcemia should be screened for metabolic bone diseases for appropriate interventions.

49) ACUTE MYOCARDITIS WITH NEGATIVE ENDOMYOCARDIAL BIOPSY

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Introduction: Myocarditis is an inflammatory disease of the heart frequently resulting from viral infection. Endomyocardial biopsy (EMB) provides the most specific diagnosis for myocarditis.

Case Presentation: An 18 yo male started to have a dull type, intermittent, retro-sternal chest pain radiating to the bilateral upper extremities which woke him up from sleep. Pain worsened in the next two days. On physical examination, initial vital showed temperature 98.8oF, PR 64, RR 18 and BP 129/79. Examination including cardiovascular system was unremarkable. Diagnostic work up showed WBC of 12.8 with left shift. The rest of hemo-gram and toxicology were normal. EKG was unremarkable and TTE showed LVEF of 60%. With chest pain persisting, CT angiography was obtained and showed normal coronaries. Indomethacin was started for possible pericarditis. However, after few days repeated TTE showed LVEF of 26 % with global LV hypokinesis with superimposed akinetic infero-lateral wall. At this time, ECG showed ST segment elevation of II, III aVF, V5 and V6 leads. Troponin and CKMB peaked at 38.89 and 64.7 respectively. Viral studies were negative. Cardiac MRI showed delayed enhancement involving the left ventricle, lateral wall and inferoseptal wall and severe hypokinesis/akinesis of the apical septum. EMB showed no evidence of evidence of myocarditis. He was managed medically conservatively and showed improvement. On discharge, TTE showed LVEF of 55 % with normalization of regional wall motion abnormalities.

Conclusion: Based on this case, we suggest that EMB is unnecessary if there are signs of recovery on imaging studies. However, EMB can be considered for patient worsening or no recovery despite treatment.

50) ASYMPTOMATIC INFECTED GIANT BULLA

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Introduction: Giant bulla is defined as a dilated airspace that occupies at least 30% of a hemithorax. The most common cause of lung bullae is chronic obstructive lung disease.

Case Presentation: A 43 yo male presented with lightheadedness which has been progressively worsened over 2 days due to severe anemia. He had an unintentional weight loss of 50-60 pounds over six months. No history of fever, chest pain, shortness of breath or sweating. On physical examination, he was not in respiratory distress and initial vitals showed BP 115/76, PR 112, Temp 97.6 O°F (36.4 O°C) RR 16, and SpO2 100%. Patient had pale conjunctiva and buccal mucosa. Lung examination showed hyperresonant to percussion on the lower 2/3 left lung field with no air entry on auscultation. The rest of physical examination was unremarkable. Diagnostic workup showed HGB/HCT- 6.2/20.9, WBC- 17.6 with left shift and platelet of 665. Chest X- ray was showed large lucent region in the left mid and lower lung. CT showed large left hemithorax bulla measuring up to 15.1 X 9.5 cm with some peripheral debris within the bulla. Left thoracotomy with total decortications showed left emphyema with trapped lung. Surgical pathology showed acute and chronic pleuritis with no organism identified. Flexible bronchoscopy with bronchial washings was done and grew candida albicans and mycobacterium avium complex (MAC). However, repeat Culture for MAC was negative. Patient was treated with Unasyn while inpatient and etrapenem after discharged.

Conclusion: Infected giant bulla is normally associated with structural lung disease, lung cancer or other malignancy but can occur isolated. Infected giant bulla should be managed surgically and medically despite the patient being asymptomatic.

51) UNILATERAL MOYAMOYA SYNDROME IN ASSOCIATION WITH SYSTEMIC LUPUS ERYTHEMATOSIS

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Introduction: Japanese doctors were the first to describe a phenomenon causing the formation of collateral blood vessels in areas of cerebrovascular occlusion in 1957. These tangled vessels were said to resemble a “puff of smoke”, which is called moyamoya in Japanese. This exceedingly rare syndrome has an incidence of approximately one in a million in the US.

Case Presentation: We describe the case of a 46 yo African-American female with a history of polysubstance abuse, pancytopenia, and unsubstantiated SLE who presented following a new-onset seizure. On admission, vital signs were stable; she was lethargic with no signs of incontinence or oral trauma. Neurological assessment revealed no discernible deficits. Lab work was positive for pancytopenia and cocaine in urine but otherwise unremarkable. While being admitted she experienced yet another seizure. CT showed an indeterminate hypoattenuation near the left basal ganglia. Immunologic workup revealed grossly elevated ANA titers and anti-dsDNA, with low C3 and C4. MRI demonstrated T2 hyperintensity and significant vessel recruitment within the left MCA territory, along with a severely diminutive left ICA. Findings were highly suggestive of diffuse lupus vasculitis and concurrent unilateral Moyamoya syndrome. Highly diagnostic CTA subsequently confirmed the diagnosis. She remained clinically stable and was discharged with close follow-up.

Discussion: MRA and CTA are the preferred modalities to diagnose Moyamoya (high diagnostic yield and noninvasive nature), although conventional CTA remains the gold standard. The occurrence of Moyamoya in people of non-Asian descent is extremely rare. Although the etiology remains unknown, Moyamoya appears to be more common in patients with autoimmune diseases. Clinicians should maintain a broad differential to include this rare phenomenon, especially when encountering cerebrovascular events in young patients or those with autoimmune diseases.

52) A CASE OF METHICILLIN SENSITIVE *STAPHYLOCOCCUS AUREUS* LIVER ABSCESS IN A 55 YEAR OLD WOMAN WITH PRE-EXISTING HEPATOBILIARY DISEASE

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Introduction: *Staphylococcus aureus* liver abscesses are usually associated with hematogenous spread. Isolated liver abscesses are rare and associated with specific circumstances. We describe a case of methicillin sensitive *Staphylococcus Aureus* (MSSA) liver abscess in a 55 year old female with history of multiple abdominal procedures.

Case Description: A 55-year-old Caucasian female with history of recurrent pancreatic stricture, chronic pancreatitis s/p pylorus-sparing Whipple in 2004 with splenectomy, failed pancreatic islet cell transplant in 2004, presented with acute on chronic right upper quadrant abdominal pain over past 2 week. The pain was non-crampy, worse with movement and eating, without relief from over the counter pain medications. She reported nausea, vomiting and diarrhea for the past one week. Patient was allergic to codeine and morphine. Patient's father had chronic pancreatitis of unclear etiology. She was a chronic cigarette smoker and denied any illicit drug or alcohol use. Vital signs showed temperature was 98.1F, pulse was 138, blood pressure was 171/67 mm Hg, respiratory rate was 20, and saturation was 93% on room air. Physical exam showed diffuse abdominal tenderness worse in the right upper quadrant. Significant labs were WBC of 20,600 cells/microliter, mild transaminitis, Alkaline Phosphatase 523 Units/Liter, venous lactate 4.5 mmol/Liter, Procalcitonin 0.2 nanogram/ml.

CT abdomen/pelvis showed multiple irregular, thick walled cystic lesions in the right hepatic lobe and dome. The largest lesion was 3.5 x 4.8 x 3.6cm with protrusion against the diaphragm. There were findings of persistent pneumobilia with intra and extrahepatic biliary ductal dilation, suspected intrahepatic biliary ductal stones, but no definite choledocholithiasis. Ultrasound guided drainage was performed on two lesions with a catheter left in the largest lesion. Cultures were positive for MSSA and light growth of *Streptococcus pneumoniae*. The patient was treated with Ampicillin-Salbamand had decreasing amounts of fluid from the drain on subsequent days; her pain improved, and was discharged home with a total 4 weeks of intravenous antibiotics.

Discussion: Though most Pyogenic liver abscesses are polymicrobial involving the Enterobacteriaceae family, *Staphylococcus aureus* infection should be suspected in patients with pyogenic liver abscess carrying history of prior hepatobiliary tract surgeries/disease. Treatment involves percutaneous drainage of the abscess with two-six weeks of antibiotics.

53) GRS CONSTRUCTED WITH TOP HITS FOR CLINICAL CAD PREDICTS CAC IN ELDERLY PATIENTS

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Coronary artery calcification (CAC) is a marker of subclinical coronary atherosclerosis and a strong independent predictor of clinical complications related to coronary atherosclerosis. We investigated whether newly identified susceptibility SNPs from the CARDIoGRAMplusC4D consortium study of clinical CAD predict the presence and extent of CAC in the ADVANCE study of elderly healthy controls. Individual genotype data in ADVANCE was first imputed with 1000 genomes. We then calculated a genetic risk score (GRS) for all individuals as an unweighted sum of the number of high risk alleles. Subjects were considered “cases” if their CAC score was greater than 75th percentile for their sex (n cases = 159, n controls = 474). We then used logistic regression to determine whether the GRS predicted case/control status. Odds ratio of GRS for the top 34, 49, 152 SNPs respectively were 1.34 ($p < 0.005$), 1.34 ($p < 0.005$), 1.55 ($P < 0.00001$). Thus, patients with a higher GRS were up to 55% more likely to have a CAC score greater than 75th percentile suggesting that susceptibility alleles for clinical CAD predominantly work through the promotion of subclinical atherosclerosis. The larger OR for the third GRS that included SNPs that have not reached genome wide significance suggests that a majority of these SNPs will likely be validated in the future. Further investigation is necessary to determine whether these trends can be confirmed in other cohorts.

54) SMALL CELL LUNG CANCER PRESENTING AS CRMP-5-MEDIATED PROGRESSIVE MOTOR AND SENSORY DEFECIT

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Background: Paraneoplastic syndromes encompass a variety of disorders associated with malignancy but not directly caused by metastasis or metabolic derangement. Neurological paraneoplastic syndromes are believed to be a T Cell mediated autoimmune attack of nervous tissue. It is thought that there may be epitope similarity between neuroendocrine tumor proteins, as in small cell lung cancer, that ultimately lead to the formation of autoantibodies and autoimmune attack of the nervous system. Therefore, treatments are focused on immunomodulation. Specifically, high dose steroid, plasmapheresis, IVIG, and T Cell modulating medications are typically employed in the treatment of neurological paraneoplastic syndromes.

Case presentation: A 67-year-old man presented to the emergency department with bilateral lower extremity weakness and dizziness that had been progressively worsening for 2 days. A small cell lung cancer was discovered and CRMP-5 antibody was found on paraneoplastic panel. He has since progressed to total bilateral lower extremity paraplegia with sensory loss to the T4 level, bladder dysfunction, and muscular atrophy in bilateral lower extremities despite treatment with steroids, plasmapheresis, and IVIG. Electromyography shows marked abnormalities in sensory and motor nerve conductions without evidence of demyelination. There was marked denervation on needle exam with no recruitable units in the distal lower extremity.

Conclusion: The identification and diagnosis of a neurologic paraneoplastic syndrome is complicated by both shared symptomatology between different antibodies and the ability of one antibody to present with a variety of complaints. Complicating matters further, paraneoplastic syndromes can often cause the presenting complaint of a malignancy. CRMP-5 most typically presents as some combination of symptoms resulting from encephalomyelitis, cerebellar degeneration, chorea, and peripheral neuropathy and is most likely to be seen in small cell lung cancer or thymoma. After addressing the primary malignancy, treatment of these syndromes is typically focused on immunomodulation with varying degrees of success.

55) LEFT VENTRICULAR NON-COMPACTION: ETIOLOGY, PATHOLOGY AND CLINICAL SIGNIFICANCE

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29 yo AA male presents with clinical symptoms of congestive heart failure the etiology of which was unclear. Pt was incidentally diagnosed with severe left ventricular (LV) dilation and HF when he came to ED for an abdominal gunshot wound at the age of 22. His left heart catheterization was negative for acute coronary disease to explain his systolic dysfunction. He had an ejection fraction of about 20% and had an AICD placed. In addition, pt was unaware of any family history of cardiovascular disease.

During this admission pt complained of increasing DOE, PND, weight gain and feeling as though his ICD had fired. He was diagnosed with CHF exacerbation secondary to poor diet and medication non-compliance. Pt was diuresed and TTE was performed. TTE showed trabeculation seen within the LV endocardium concerning for left ventricular non-compaction (LVNC). Furthermore a small thrombus within the trabeculation could not be ruled out.

LVNC also known as spongy myocardium is a rare disease that, occurs in the 12th week in utero when the spongy myocardium fails to transform into mature compact myocardium. This results in deep recess and trabeculations which form within a dilated left ventricle. This is a rare disease but should be considered in the differential for diagnosis of new onset HF in young individuals.

These individuals are at an increased risk for arrhythmias, thrombus formation and heart failure. The diagnosis is made either by TTE or Cardiac MRI (not performed in this patient given that he had a subcutaneous ICD lead). Anticoagulation should be considered given their increased risk for thromboemboli caused by their dilated left ventricle. Our patient was started on coumadin given potential for LV thrombus formation. This is also a congenital disease, and so family members should be offered an opportunity to be screened. This did not occur in this case because of the pt's financial constraints. Keeping LVNC in the differential for new HF in younger individuals is critical as it helps clinicians risk stratify the patient's outcomes for strokes, arrhythmias and their children's risk for heart disease.

56) BEZEX SYNDROME: AN UNUSUAL PRESENTATION OF AN ESOPHAGEAL MALIGNANCY

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Introduction: Bezex syndrome is a rare neoplastic syndrome associated with upper gastrointestinal or oropharyngeal malignancies. While its histology is non-specific, recognition is important as it can be the presenting feature of its associated cancer. Identification of Bezex syndrome should prompt a full cancer work up and can result in faster tumor diagnosis, treatment initiation and survival.

Case: A 57-year-old male was admitted due to 3 weeks of painful, blistering rash with desquamation on his dorsal and palmar hands and feet, bilateral knees, elbows, and helices of ears. One week prior to presentation, he underwent endoscopy for dysphagia, weight loss and a mass seen on chest CT. Biopsies from his esophagus demonstrated a poorly differentiated carcinoma. Dermatology, who he had seen just prior to admission, was suspicious of Bezex syndrome (acrokeratosis paraneoplastica) due to his mass. He was treated with oxycodone, oral prednisone, topical steroids and Epsom salt soak. Skin biopsy demonstrated non-specific findings of an acanthotic and mildly spongiotic epidermis with eosinophils, parakeratosis in the stratum corneum with rare neutrophils and increased eosinophils in the dermis. Bullous pemphigoid was ruled out. The patient's disease progressed despite chemotherapy, and he ultimately developed refractory seizures due to brain metastasis and died of multi-organ failure.

Discussion: Bezex syndrome was first described in 1965 as a paraneoplastic syndrome causing hyperkeratosis of the extremities. It is characterized by erythematous psoriatic plaques most commonly affecting the ear helices and nose. Bezex syndrome is most commonly associated with squamous cell carcinomas of the upper gastrointestinal tract or oropharynx, less commonly with cervical lymph node metastasis and rarely with adenocarcinomas.

While the cutaneous lesions of Bezex syndrome have non-specific histological features, generally the lesions follow a predictable progression. The first stage typically starts while the associated tumor is asymptomatic. Poorly defined macules erupt on the fingers, toe and ear helices. In stage two, when the tumor is causing local signs and symptoms, lesions evolve to include the palms, soles and cheeks. The third stage involves systemic tumor symptoms with further lesion progression to the arms, legs and trunk. Plaques often coalesce and are erythematous with a fine adherent scale. Symptomatic treatment of Bezex syndrome is primarily pain control as it typically only resolves with tumor treatment.

Recognition of Bezex syndrome is important as it is often the first sign of an otherwise asymptomatic cancer. Because of this, the diagnosis of Bezex should prompt a full cancer screening. Faster initiation of screening leads to earlier diagnosis and initiation of treatment which can result in improved outcomes.

57) A TRIPLE ACID-BASE DISTURBANCE DUE TO SELF-MEDICATION FOR ABDOMINAL PAIN

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Introduction: Studies show that alkalemia can be associated with significant morbidity and mortality, especially in cases of mixed metabolic and respiratory alkalosis. Determining cause is important both for treatment choice and maximizing outcomes. This can be challenging, however, especially in the context of complicated acid-base disorders: we describe such a case.

Case Report: A 39 year old man with history of chronic kidney disease and alcohol dependence presented with persistent nausea and emesis, altered consciousness, and tetanic spasms following consumption of a large volume of alcohol one day prior. Laboratory showed hyponatremia (133 mmol/L), hypochloremia (71 mmol/L), hypomagnesemia (1.0 mg/dL), normal total calcium (10.0 mg/dL), decreased ionized calcium (0.89 mmol/L), and elevated bicarbonate (39 mmol/L), blood urea (24 mg/dL) and creatinine (3.13 mg/dL). Arterial blood pH was elevated (7.553), arterial carbon dioxide within reference range (41.5 mmHg), anion gap elevated (23), salicylates negative, cocaine positive and acetone present in the blood. This represents a metabolic alkalosis, an anion gap and concomitant respiratory alkalosis.

The degree of acid-base disturbance seemed inconsistent with acute alcohol intoxication/ withdrawal: further history taking revealed ingestion of 2 cups of apple cider vinegar with baking soda as a home remedy for nausea. Suspected causes of metabolic alkalosis include ingestion of bicarbonate (baking soda) and vomiting. Causes of his metabolic acidosis may include lactic acidosis, known to be associated with respiratory alkalosis in the context of anxiety, uremia, and acetonemia, likely due to conversion of acetic acid in apple cider vinegar to acetone during digestion. Sources of his respiratory alkalosis may include anxiety and pain. Elevated creatinine represented prerenal acute kidney injury on chronic kidney disease based on fractional excretion of sodium. Tetany likely resulted from low ionized calcium due to increased albumin binding in the context of alkalemia. He received supportive care with electrolyte replacement, treatment for nausea and alcohol withdrawal, and discharge home after a week. He was advised against large amounts of apple cider vinegar and baking soda and informed of treatment options for alcohol dependence.

Discussion: In cases of mixed acid-base disturbance in patients, prompt etiologic identification is important since it may lead to severe morbidity and mortality and source-specific treatment should not be delayed. Supportive treatment may also be warranted: as always, “do no harm” is an operative principle. Causes may be multi-factorial, so thorough history and laboratory work-up merits careful attention. This case demonstrates the importance of good history taking in such cases as well as the need for a systematic approach to acid-base disorders as the metabolic acidosis could have been easily overlooked given his very high bicarbonate level.

58) SUCCESSFUL TREATMENT OF DIFFUSE ALVEOLAR HEMORRHAGE WITH RITUXIMAB IN A PATIENT WITH HENOCH-SCHONLEIN PURPURA

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Introduction: Diffuse Alveolar Hemorrhage (DAH) is a rare and life threatening complication of HSP with reported prevalence 0.8-5%. Current protocols use pulse methylprednisolone and cyclophosphamide but new treatment options including Rituximab have shown to be effective in refractory cases.

Case Presentation: A 36 year old woman was seen in the Nephrology clinic for evaluation of microscopic hematuria and proteinuria. She also had purpuric rash both legs. Biopsy of the rash showed leucocytoclastic vasculitis. She had a kidney biopsy. This showed necrotizing glomerulonephritis with crescent formation. There were mesangial IgA deposits seen. Based on this clinical presentation, a diagnosis of Henoch- Schonlein purpura (HSP) was made. She was treated with IV steroids and pulse IV cyclophosphamide. 3 weeks later she was admitted with shortness of breath, hypoxemia and fever. Her chest X-ray showed bilateral alveolar infiltrates. She also had worsening hematuria and proteinuria with acute kidney injury. She was started on antibiotics for presumed pneumonia in an immunocompromised host. Patient's respiratory status continued to deteriorate. She was intubated and placed on a mechanical ventilator. Continuous renal replacement therapy was started. As the patient did not respond to antibiotics, bronchoalveolar lavage (BAL) was performed. This showed a bloody effluent. Laboratory evaluation for infectious etiologies on BAL sample was negative. Given positive histochemical staining for hemosiderin laden macrophages, a diagnosis of vasculitis causing alveolar hemorrhage was made. As the patient had failed cyclophosphamide therapy, she was started on high dose steroids and Rituximab, with significant improvement in renal function and respiratory status. She was extubated and later taken off dialysis. Her serum creatinine normalized over time.

Discussion: Diffuse Alveolar Hemorrhage (DAH) is a rare and life threatening complication of HSP. Current treatment options include the use pulse methylprednisolone and cyclophosphamide. Rituximab has been shown to be efficacious in the treatment of anti-neutrophil cytoplasmic antibody associated vasculitis. This is one of the few case reports of successful treatment of refractory HSP nephritis with pulmonary vasculitis causing diffuse alveolar hemorrhage with Rituximab.

59) CUTANEOUS VASCULITIS SECONDARY TO LEVAMISOLE TOXICITY

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Introduction: Levamisole is increasingly being used as an additive in cocaine. As such, vasculitic cutaneous lesions from levamisole toxicity are being diagnosed more frequently. It is important to differentiate these lesions from those caused by other autoimmune vasculitides due to significant differences in how they are managed.

Case Presentation: A 49-year-old male with history of ongoing cocaine abuse with previous levamisole-toxicity presented with painful skin lesions. Lesions had been present intermittently for the past four years, occurring on the extremities and ears, spontaneously resolving after approximately one week. The current lesions were particularly painful with purulent discharge. Recurrent levamisole-induced cutaneous vasculitis was diagnosed. Intermediate-stage lesions with black eschar were seen over the extremities and ear. Advanced stage lesions with central ulcerations and necrotic borders were also present. The patient was treated with high-dose daily prednisone and was taken to the operating room for wound debridement. The patient was also noted to have acute kidney injury and positive p-ANCA and c-ANCA. Renal biopsy showed crescentic glomerulonephritis with underlying membranous nephropathy, a rare result of levamisole vasculitis. Renal function improved with steroid treatment. He was discharged on an extended steroid taper and counseled to stop using cocaine.

Discussion: Levamisole is an immunomodulatory agent previously used for pediatric nephropathy, rheumatoid arthritis, and colon cancer. It was withdrawn from the market in 2000 because of its side effect profile. Approximately 70% of cocaine in the United States contains levamisole. Patients with levamisole toxicity present with characteristic cutaneous manifestations that are classically seen on the helix of the ear and on the extremities. These lesions begin as purpura, progress to bullae, and eventually necrose. Patients often have arthralgias, oral sores, fever, and neutropenia. Treatment is typically symptomatic after discontinuing levamisole exposure. Steroids can expedite recovery. Wound care and surgical debridement may be required if lesions are extensive. Levamisole-induced vasculitis is usually a diagnosis of exclusion. Renal involvement is rare, but several case reports have described pauci-immune focal necrotizing and crescentic glomerulonephritis. As levamisole-induced vasculitis becomes increasingly common, it is imperative that physicians recognize the characteristic cutaneous lesions, so that appropriate treatment can be expedited and more serious complications avoided.

60) THORACIC AORTIC ANEURYSM 30 YEARS AFTER AORTIC VALVE REPLACEMENT

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Introduction: Aneurysms of the proximal thoracic aorta (TAA) are increasingly recognized and often diagnosed on imaging studies performed for other indications. Proximal TAA has been frequently associated with a bicuspid aortic valve (BAV), one of the most common congenital heart anomalies diagnosed in adult patients. Some patients with BAV demonstrate progressive dilation of the proximal ascending aorta even after elective aortic valve replacement (AVR). This observation suggests that the pathophysiology of BAV “aortopathy” is likely multi-factorial and not strictly limited to increased wall stress due to abnormal flow characteristics across the aortic valve.

Case: A 54 year old man received regular follow-up in the Anticoagulation Clinic at the Milwaukee VAMC in order to monitor his chronic warfarin therapy following placement of a 27 mm Bjork Shiley aortic valve prosthesis in 1976 for aortic insufficiency that was attributed to a BAV. When the patient began to notice increased dyspnea on exertion and generalized fatigue in the fall of 2006, a transthoracic echocardiogram (TTE) was ordered to evaluate his left ventricular (LV) function. The TTE showed normal prosthetic valve function, mildly decreased LV ejection fraction but severe dilation of the aortic root. CT angiography demonstrated a fusiform aneurysm of the ascending aorta measuring 7.3 cm in AP dimension – the aortic arch, great vessels and descending aorta were all normal in caliber. The patient subsequently underwent aortic root replacement in February 2007 using a St. Jude 27 mm composite graft – his continued follow-up in the outpatient internal medicine clinic has been uneventful.

Discussion: Aneurysms of the proximal thoracic aorta have multiple potential etiologies including genetic syndromes (Marfan and Ehlers-Danlos), vasculitis/inflammatory diseases (Takayasu and Giant Cell Arteritis) and degenerative disorders. Clinicians should be aware that BAV has also been strongly linked to the development of proximal TAA. The aortic dilatation associated with BAV tends to occur more frequently and at a younger age than in patients with tri-leaflet aortic valves. BAV patients with isolated aortic insufficiency may be at increased risk of late aortic events compared with BAV stenosis patients even after the aortic valve has been replaced. Further studies are needed not only to ascertain the detailed pathophysiology but also to clinically determine the optimal timing and method of assessing the ascending aorta in BAV patients, including those patients who have previously undergone AVR.

61) DIGITAL ISCHEMIA SECONDARY TO LOW GRADE DIC DUE TO ADENOCARCINOMA

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Introduction: Disseminated Intravascular Coagulation (DIC) is an acquired syndrome and typically a systemic process producing both thrombosis and hemorrhage. DIC can be caused by exposure of the blood to procoagulants such as cancer procoagulants. The key to successful management of DIC is often effective treatment of the underlying disease.

Case: We discuss a 65-year-old Caucasian male with low grade (non-overt) DIC. He had end stage liver disease secondary to primary sclerosing cholangitis and ulcerative colitis status post total colectomy and end ileostomy decades earlier. Patient was receiving large volume abdominal paracentesis every fourth day for comfort. Two weeks prior to presentation, patient had a lower GI bleed at an outside hospital thought to be from peristomal varices. TIPS was not performed due to patient's high MELD scores in the upper 20's to low 30's. Patient presented in February 2014 to a major Milwaukee teaching hospital with acute on chronic renal failure noted on blood work and progressive fingertip ischemia and ulcerations over four weeks. In the hospital, bilateral upper extremities duplex showed normal proximal arterial flow. CT of the abdomen and pelvis for work up of liver transplant showed new upper abdominal lymphadenopathy with largest nodes measuring 4.7 cm. His platelet counts were between 55 and 134 with gradual decrease during patient's two week hospitalization. Extensive laboratory workup for the fingertip ulcerations had pertinent positives including elevated d-dimer levels between 5.2 and 7.2, prolonged PT and PTT, and low fibrinogen levels. Patient had endoscopic ultrasound and fine needle aspiration of a hepatic lymph node. Pathology showed malignant cells positive for CA19.9 and concerning for pancreatic adenocarcinoma. Patient opted to be discharged home with palliative care.

Discussion: Malignancy, particularly solid tumors, is the most common cause of chronic or non-overt DIC. In chronic or compensated DIC, blood is continuously or intermittently exposed to small amount of procoagulant(s), and most patients are asymptomatic but with laboratory evidence of low-grade activation of coagulation and fibrinolysis. Thrombotic manifestation may be present and include arterial thromboses producing digital ischemia. Risk factors for development of symptomatic chronic DIC in patients with cancer include male sex, age over 60, tumor necrosis and advanced stage disease. Survival is reduced in patients with DIC. Detection of unexplained low grade DIC should prompt an evaluation for occult cancer, especially an adenocarcinoma.

62) CAREFUL PHYSICAL EXAM REMAINS A KEY STEP TO DIAGNOSIS

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Introduction: A thorough history can often lead one down the correct path towards a diagnosis, but even in the most straightforward cases it is crucial that a careful physical exam take place. In this exciting time of advancing diagnostics, the art of physical exam remains a skill to be honed and trusted.

Case Presentation: A 62 year old man with excellent health presents for evaluation of two weeks of abdominal distension with a sense of bloating. Outside of a slight increase in stool frequency, from one to two formed stools a day, he was feeling in his usual state of health. He had been seen in urgent care prior to clinic visit, at which time a CBC and abdominal x-ray were obtained. As abdominal x-ray did evidence some increased stool burden, initial conservative management with bowel regimen was started by urgent care provider. However, he was training for a triathlon and as the sense of abdominal fullness was impacting his cycling he again sought care when he had not improved a week later. Careful palpation of his abdomen was performed as part of his thorough physical exam, and was remarkable for a superficial sense of nodular fullness on light palpation, which disappeared with deep palpation. Due to this unusual finding on abdominal exam, CT scan of the abdomen was performed which confirmed omental caking and peritoneal nodularity with a thickened and lobular appendix. Prompt referral to a surgical oncologist resulted in expedited surgical evaluation and exploratory laparotomy which confirmed the diagnosis of metastatic appendiceal mucinous adenocarcinoma.

Discussion: The vast array of diagnostic options available is truly impressive, and there are certainly times when the newest technology is the best method for diagnosis. At the same time, there is much to be said about a thoughtful physical exam for each patient encounter. As a new generation of physicians beginning practice, it is crucial that we learn to blend the old with the new and learn to trust our own two hands as much as our imaging modalities.

63) A SELF-LIMITED VIRUS TURNED DEADLY

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Presentation: A previously healthy 22 year old male college student presented to the emergency room (ER) with one week of progressive respiratory symptoms: shortness of breath, productive cough, and fever. A chest x-ray revealed a right upper lobe infiltrate. Levofloxacin was prescribed and he was discharged. Within 24 hours, he re-presented to the ER for worsening dyspnea. He was still febrile, tachycardic and hypoxic. Chest x-ray showed a progressive right upper lobe infiltrate. He was admitted and continued on levofloxacin.

Hospital Course: Hypoxia progressively worsened, fever continued, and on hospital day 3, he was transferred to the ICU and started on BiPAP. Microbiology was nondiagnostic and legionella testing negative. Antibiotics were changed to ceftriaxone and azithromycin due to a possible reaction to levofloxacin. Chest CT revealed right sided patchy nodular airspace disease. Due to progressive illness and lack of response to antibiotics, non-bacterial sources were suspected. Voriconazole was added for empiric treatment of blastomycosis on day 4. He became more hypoxic and required intubation on hospital day 5. A bronchoscopy with bronchoalveolar lavage (BAL) was performed with lymphocytic predominance but no yeast forms. On hospital day 6 he underwent an open lung biopsy; pathology was consistent with acute respiratory distress syndrome. The adenovirus PCR from his BAL returned positive and all other viral studies were negative. Fungal studies were negative and voriconazole was stopped. With literature supporting in vivo antiviral activity of cidofovir to adenovirus, he was treated with cidofovir on hospital day 7. Antibiotics were stopped on hospital day 8 when bacterial cultures remained negative. His oxygenation improved, and he was extubated on hospital day 10. He was discharged home on hospital day 19.

Discussion: Adenovirus is a DNA virus that causes various infections, most commonly conjunctivitis, mild respiratory infections and gastroenteritis. Infections are typically self-limited in the immunocompetent host, but can be life-threatening in immunosuppressed hosts. Cidofovir is an antiviral agent approved to treat CMV infections, and has also shown to have in vivo activity against adenovirus. It has been used to treat adenovirus infections in immunocompromised patients in varying dosages. There are case reports of severe adenoviral disease in healthy adults treated with supportive care, but there is no data on use of cidofovir in this population. This case illustrates potential successful use of cidofovir in an immunocompetent adult with life-threatening adenovirus pneumonia.

64) A RARE CAUSE OF TYPE 2 MYOCARDIAL INFARCTION

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Presentation: A 59 year old previously healthy male presented to the emergency room for headache, nausea and chest pain. Several hours earlier, he had a bilateral hydrocele repair under general anesthesia; the surgery was uneventful. Blood pressure on arrival to the emergency room was 222/118. An electrocardiogram showed inferior ST depression and troponin was positive. Chest CT was negative for pulmonary embolism. He was admitted to the coronary intensive care unit.

Hospital Course: Hypertension was controlled with a nitroglycerin drip. Symptoms resolved, and troponin levels peaked at 3.4 ng/mL. Echocardiogram performed the following morning showed severe basal and inferior hypokinesis, but cardiac catheterization was normal. On hospital days 2 and 3, he had multiple episodes of severe hypertension, tachycardia, diaphoresis and headache which raised clinical concern for pheochromocytoma. Abdominal CT showed a 7 centimeter heterogeneous adrenal mass measuring 60 hounsfield units. Plasma and urine metanephrines were markedly elevated, and plasma aldosterone to renin ratio and urinary free cortisol were normal. He was transitioned to metoprolol and phenoxybenzamine which effectively controlled his hypertension. He became orthostatic and was treated with a high salt diet. Three weeks later, he returned for a laparoscopic adrenalectomy which was uneventful; pathology confirmed pheochromocytoma. Phenoxybenzamine and metoprolol were discontinued.

Discussion: A pheochromocytoma is a neuroendocrine tumor that arises from the chromaffin cells of the adrenal medulla. Pheochromocytomas secrete catecholamines and classically cause symptoms of headache, diaphoresis, and tachycardia. Hypertension occurring during surgery is a common presentation. Pheochromocytoma should be also suspected in patients with resistant or early onset hypertension, hyperadrenergic spells, and incidental adrenal adenomas. Occasionally, the initial presentation is an acute coronary syndrome. Case reports in the literature describe both ST-elevation and non-ST-elevation myocardial infarction as the presenting finding in these patients. Takotsubo cardiomyopathy has also been reported. When coronary catheterization is negative, clinical history must be reexamined to evaluate for another cause.

65) EMPTY SELLA SYNDROME PRESENTING AS EPISODIC HYPOGLYCEMIA

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A 38-year-old female with a history of Crohn's disease and hypothyroidism presented with dizziness after she skipped a meal due to nausea. On arrival, her blood glucose level was 41 mg/dL, which was corrected with IV glucose. Over the past 18 months, she had 5 previous episodes of symptomatic hypoglycemia. Her family history is positive for diabetes mellitus in her father. Physical exam was unremarkable. Laboratory findings included an elevated TSH, a low free T4, and a normal hemoglobin A1C. During the hospital course she continued to have episodes of hypoglycemia that ranged between 40-60mg/dL. Endocrinology was consulted and a cosyntropin stimulation test was performed, which was positive for adrenal insufficiency. An ACTH level drawn at the start of the test was low indicating secondary adrenal insufficiency. An MRI of the sella was then performed and it showed a partially empty sella with a thin sliver of pituitary tissue at the floor. The patient was started on hydrocortisone for her secondary adrenal insufficiency and levothyroxine for her hypothyroidism. The patient's hypoglycemic episodes resolved after the initiation of steroid therapy.

Empty sella syndrome is a condition in which the sella turcica appears empty on imaging. This syndrome has been reported in 6-20% of people. Many patients are asymptomatic, however between 19-50% of patients present with various endocrine abnormalities. The most common endocrine abnormalities are decreased ACTH, TSH, and GH as well as elevated prolactin, all of which were seen in our patient. When the pituitary gland fails to produce ACTH, secondary adrenal insufficiency occurs. This can result in hypoglycemia because cortisol normally acts to increase blood sugar by increasing gluconeogenesis and by blocking the uptake of glucose into cells throughout the body. Adrenal insufficiency can be diagnosed by the cosyntropin stimulation test, where a bolus of cosyntropin is given to the patient and plasma cortisol levels are measured over time. In primary adrenal insufficiency, cortisol levels will fail to increase due to atrophy of the adrenal glands, and plasma ACTH levels will be high. In secondary adrenal insufficiency, cortisol levels will fail to increase because the adrenal glands are not used to being stimulated, but they will begin to produce cortisol within 4-5 days. In secondary adrenal insufficiency, plasma ACTH levels will be low. Treatment of empty sella syndrome is replacement of the deficient hormones, which included hydrocortisone and levothyroxine for our patient. Of note, with secondary adrenal insufficiency, the renin-aldosterone system is intact so mineralocorticoid treatment is not necessary.

66) DASATINIB IN ACUTE MYELOID LEUKEMIA WITH C-KIT POSITIVITY AND ASSOCIATED SYSTEMIC MASTOCYTOSIS

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Introduction: Systemic mastocytosis and associated hematological non-mass cell lineage disease (SM-AHNMD) is a rare disorder with particularly poor prognosis. In patients who present with acute myeloid leukemia (AML) with c-KIT mutations, dasatinib may help achieve remission. We present a case of SM-AHNMD where the hematologic malignancy was C-KIT positive AML and where dasatinib was added to combination chemotherapy.

Case: A 22-year-old male presented to an outside hospital with productive cough, fevers, and nights sweats of two months' duration. Work up revealed white blood cell count of 65,000/uL with 80% circulating blasts, severe anemia, and thrombocytopenia. Bone marrow biopsy (BM) revealed AML and mastocytosis; cytogenetics and molecular analysis demonstrated translocation (8; 21) and c-KIT D816V mutation, respectively. He was diagnosed with SM-AHNMD and underwent standard induction with cytarabine and idarubicin, complicated by bilateral pre-retinal hemorrhage and R molar dental abscess. Patient's fevers and symptoms improved with induction and supportive care. Mid-cycle BM revealed residual disease with persistent mastocytosis. Patient was transferred to our facility for further workup and treatment. He received re-induction with cytarabine and idarubicin, and after informed consent dasatinib was added given his CKIT-positive disease. Day 28 BM revealed no evidence of AML, although mastocytosis persisted. He is undergoing consolidation therapy with plan for early stem cell transplant.

Discussion: AML with t(8;21) occurs in 5-8% of cases of AML and is typically associated with a favorable prognosis. However, c-KIT mutation is also present in 20-25% of these patients and this combination is associated with poorer prognosis. SM-AHNMD is an extremely rare disorder with few case series and reports in literature, but there are case reports of SM-AHNMD where the hematologic disorder is AML with t(8;21) and C-Kit mutations. In a series of 10 patients, seven patients achieved remission after induction or consolidation with only two patients maintaining a durable remission. Both of these patients proceeded to early allogeneic hematopoietic transplant. Dasatinib is reported to inhibit c-KIT and its downstream signaling, with published literature demonstrating significant benefit for patients with c-KIT positive AML. A prospective study is under way to determine the efficacy of dasatinib in the setting of c-KIT positive leukemia. In patients with SM-AHNMD, prognosis is poor and early transplant may provide the best opportunity to achieve durable remission.

67) ACQUIRED ANGIOEDEMA IN A PATIENT WITH A HISTORY OF CLL

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Introduction: Acquired angioedema is a rare disorder distinct from hereditary angioedema. It is most commonly associated with lymphoproliferative disorders. The presentation includes recurrent episodes of self-limited swelling of the skin, upper respiratory tract, and GI tracts without urticarial lesions. It is often under recognized disorder that can result in frequent hospitalizations, and is potentially fatal.

Case: The patient is an otherwise healthy 55 year old who was diagnosed with chronic lymphocytic leukemia in 2012, when routine mammography revealed axillary lymphadenopathy. She denied any constitutional symptoms and had normal red blood cell and platelet counts. She followed with an oncologist regularly with routine blood counts and had never received chemotherapy. Between June 2013 and December 2013 she presented to the hospital four times with the same symptoms of abdominal pain and dizziness. Each time she was found to be hypotensive and received large volumes of intravenous fluids. Her dizziness and hypotension would resolve after volume resuscitation and abdominal pain would self-resolve after a few days. During each hospitalization the patient received a variety of diagnostic studies. Broad infectious laboratory work up was negative. ANA was negative. Complement levels including C3, C4, and CH 50 were low. Abdominal CT revealed non-specific duodenal and jejunal wall thickening with stable lymphadenopathy. EGD with biopsy of the duodenum and jejunum were unremarkable. Eventually a C1 esterase inhibitor level was ordered, which was low, revealing a diagnosis of acquired angioedema associated with a history of CLL. The patient was subsequently started on chemotherapy and has had no recurrence of symptoms since that time.

Discussion: Acquired angioedema is characterized by an acquired deficiency in C1 esterase inhibitor, an inhibitor of the first component of the complement pathway. It is less well known than hereditary angioedema. Acquired angioedema presents in middle age in patients with lymphoproliferative disorders while hereditary disease presents in otherwise healthy youth. It typically involves edema of GI tract or upper respiratory tract and self resolves after a few days. No urticarial lesions are present. Patients with acquired angioedema have low complement levels and identifiable antibodies to C1 esterase on lab analysis. The relationship between the production of these antibodies and concurrent lymphoproliferative disease is incompletely understood. The diagnosis often evades recognition for years. Some reasons for this include a lack of knowledge of the acquired form of the illness, uncertainly regarding lab analysis, and fluctuant complement levels during the early development of this illness.

68) PORTAL VEIN AND SMV THROMBOSIS ASSOCIATED WITH CMV INFECTION IN AN IMMUNOCOMPETANT HOST

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Introduction: Portal Vein Thrombosis and its association with CMV infection is a known but not frequently reported phenomenon. PV/SMV thrombosis is commonly associated with cirrhosis, portal hypertension, and/or thrombophilia. This case illustrates the risk of CMV as another risk factor for PV and SMV thrombosis.

Case Details: We present a case of a 30-year-old otherwise healthy female who presented with a 3-week history of general malaise and several constitutional symptoms including chills, night sweats, myalgias, headaches, nausea, and fatigue and subsequently abdominal fullness. The week prior to admission she went to urgent care with fever of 101.5F and had an elevated D-dimer at that time. She had a negative Lyme PCR, monospot, and MRA to r/o PE, and was diagnosed with viral illness. Five days prior to admission, she endorsed dull left sided abdominal pain. She saw her PCP and had an extensive workup performed 2 days after her ED visit, which included an infectious workup (EBV, CMV, HIV, ehrlichia). Lab testing at that encounter was notable for: LD (440), thrombocytopenia (140), leukocytosis (13.6) with lymphocytosis (9790, with reactive lymphocytes), Alk phos (197), ALT (175), AST (158).

Past medical history was notable for obesity (BMI 40kg/m²). Her Only medication was estrogen-containing OCP. Family history was notable for Factor V Leiden on father's side. She was a non-smoker.

On day of admission, abdominal CT showed complete thrombosis of the right portal vein, as well as nearly occlusive thrombus present within the superior mesenteric vein, and no evidence of cirrhosis. She was initiated on therapeutic LMWH, and bridged to warfarin, with improvement in her abdominal pain. Hypercoagulability workup was unrevealing, including a negative factor V Leiden and JAK2. CMV IgG and IgM antibodies were both detectable on day 1 of hospital stay. CMV PCR resulted positive at 57,000.

Discussion: This case illustrates the pro-coagulant risk of an active CMV infection. Recognizing that CMV infection can present like a worrisome disease and lead to clinically significant venous thrombosis with high morbidity and mortality could result in a more rapid, tailored workup and less risk for patients. Establishing a connection between CMV and VTE could lead to a more standard approach for anticoagulation.

69) MUNCHAUSEN SYNDROME IN A CASE OF BORDERLINE PERSONALITY DISORDER

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Factitious disorders can be some of the most frustrating and difficult conditions to deal with as a medical professional, especially when the presented disorder is exceptionally rare and cannot be easily tested for. Patients who have these rare illnesses, real or feigned, can feasibly know more about them than the treating physician, which can be either a blessing or a barrier.

We present here a case of a 26-year-old woman who was transferred from a community hospital to Froedtert on a Friday afternoon for care of an Acute Intermittent Porphyria (AIP) exacerbation with paralysis of her right-sided extremities. Other than morbid obesity, her physical exam was remarkable only for 0/5 strength and loss of sensation in her right upper and lower extremities. Initial diagnostic workup including a brain and C-spine MRI and urine porphyrins were negative. She was empirically started on IV hematin for 4 days without clinical improvement. She also received IV Dilaudid and Benadryl for self-reported severe abdominal pain. From the outset, she was emotionally labile, drawing a picture of the attending with “#1 Physician!” written on it. Outside records revealed not only multiple negative AIP tests, but it was also discovered that by the age of 15 her mother had hospitalized her over 120 times at least 26 institutions. Because of this, she missed over 50% of school, and did not complete high school. When confronted with this information, she became hostile towards staff. She still demanded continued treatment for her AIP, but it was held secondary to high suspicion of factitious disorder. When her demands were no longer being met, she walked out of the room without any difficulty, and left AMA five days after admission.

Munchausen Syndrome is a condition that is somewhat rare, although prevalence data is not readily available. As in this case, it is frequently associated with borderline personality disorder, but can also be seen in antisocial and passive-aggressive personality disorders. A past history of Munchausen Syndrome by Proxy is a strong predictor of future development of Munchausen Syndrome. Presentations of s Syndrome can vary widely, but can classically take the form of insulin-induced hypoglycemia or laxative-induced diarrhea. There is no curative treatment for this condition, and it is exceptionally difficult to manage. These patients are often manipulative and can induce significant countertransference. The most important management strategy includes setting limits on visits, and limiting care to a single PCP as much as possible. This PCP should know the patient well, and must take the patient’s complaints seriously, but not order any unnecessary diagnostic testing and avoid iatrogenic harm.

70) HEPATITIS C INDUCED ACUTE LIVER INJURY

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Introduction: Most patients exposed to hepatitis C develop chronic infections (80%) and usually are asymptomatic or may present with nonspecific symptoms such as myalgia or arthralgia. Rarely, patients develop acute hepatitis C and present with jaundice, nausea, abdominal pain and elevated transaminases 10-20x the upper limit of normal. Here, we present a patient with hepatitis C induced acute liver injury. The objective of this case is to present an atypical presentation of hepatitis C induced acute liver injury.

Case: 33 year old male with hepatitis C (newly diagnosed based on hepatitis C NAAT) and heroin abuse (last use one week prior to admission) presented with abdominal pain, and jaundice of 2 weeks duration. Laboratory findings were significant for ALT of 2869, AST of 1739, total bilirubin of 5.4 and direct bilirubin of 4.1. Physical exam was remarkable for jaundice, scleral icterus and bilateral upper quadrant abdominal pain. Gastroenterology was consulted for further evaluation of transaminitis and jaundice. Viral studies, including EBV IgM, CMV, varicella, HSV and hepatitis E serologies, were negative. Hepatitis C genotype was 1a and viral load was greater than 76 million copies/mL. Transjugular liver biopsy revealed grade three acute hepatitis, indicating marked inflammation with severe focal liver cell necrosis and no significant fibrosis. MRCP revealed diffusely thickened gallbladder, periportal edema, perihepatic fluid with portal nodes consistent with acute viral hepatitis. Transaminases continued to downtrend during admission and at discharge ALT was 1056 and AST was 157.

Discussion: Here we report a case of acute Hepatitis C in a patient with significant transaminitis and jaundice. Initially it was hypothesized that the patient's heroin use could have contributed to his acute liver injury. However, heroin does not cause significant liver injury and would not account for the level of transaminitis seen in this patient. Given the MRCP and liver biopsy results consistent with acute viral liver injury, acute Hepatitis C is the most likely etiology of this patient's transaminitis and clinical picture. With IV hydration, the patient's transaminitis and clinical symptoms improved over the course of his hospital stay. Although hepatitis C typically presents with asymptomatic chronic liver injury, it is important to recognize that it can be the cause of acute liver injury. Given the risk of hepatitis C induced fulminant liver failure, patients with signs and symptoms of hepatitis C should be monitored closely. Viral load and genotype should be determined in order to initiate the appropriate course of treatment. If transaminases remain elevated for 12 weeks after inoculation or symptom onset, consider initiation of therapy with peginterferon alfa and ribavirin.

71) ATYPICAL PRESENTATION OF MAC INFECTION

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Mycobacterium avium complex (MAC) refers to an infection caused by either *M. avium* or *M. intracellulare*. MAC infections typically affect the respiratory or GI tract and symptoms include fevers, night sweats, abdominal pain, diarrhea and weight loss. Here, we present a patient with disseminated MAC (DMAC) causing granulomatous hepatitis and bone marrow suppression who has remained asymptomatic throughout her illness.

28 year old female with history of AIDS presented for evaluation of elevated liver transaminases. Laboratory findings were significant for AST elevation of 964, ALT of 220, Alkaline phosphatase of 871, CD4 count of 8, WBC count of 500 and platelets of 87. Patient appeared comfortable and was not complaining of any respiratory or GI symptoms. Physical exam and review of systems were unremarkable. Over the course of hospitalization, patient underwent an extensive microbiology work up as well as imaging of the abdomen, the hepatobiliary tree and biopsies of the bone marrow and liver. Bone marrow biopsy showed non-necrotizing granulomatous inflammation with acid fast positive organisms and liver biopsy revealed granulomatous hepatitis with acid-fast bacilli. Laboratory results were significant for a positive AFB blood culture. Based on biopsy results and positive blood culture, disseminated MAC with multiorgan involvement was thought to be responsible for the pancytopenia as well as the hepatocellular liver injury. Azithromycin, Rifabutin, and Amikacin were chosen as triple therapy for DMAC as the patient had advanced AIDS with a low CD4 count. WBC count increased with Neupogen administration. During hospital course, patient continued to remain asymptomatic however her liver function tests significantly improved after the initiation of antibiotics.

DMAC is a life threatening infection that carries up to a three-fold risk of death in patients with AIDS. It is typically acquired through inhalation or ingestion of organisms from soil or water after the CD4 count drops below 50 cells/mm³. First line agents for treatment are clarithromycin and azithromycin followed by ethambutol and rifampin. IDSA guidelines suggest that at least two drugs should be initiated for the treatment of DMAC due to concern for development of resistance. In our patient, however, we elected to use a three drug therapy of Azithromycin, Rifabutin, and Amikacin due to the patient's severe immunocompromised state. Ethambutol was avoided due to patient's history of optic neuritis. Therapy will be continued for at least a year and until the CD4 count remains above 100 cells/mm³ for at least 6 months.

72) CASE OF HYPOGLOSSAL NERVE PALSY IN A PATIENT WITH HIV

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Introduction: Burkitt Lymphoma (BL) is an aggressive B-cell non-Hodgkin lymphoma with three recognized clinical forms: African (endemic), sporadic and immunodeficiency-associated. We describe a case of hypoglossal nerve palsy secondary to BL in the setting of HIV.

Case Summary: Mr. P is a previously healthy 52 year-old who presented with several months of urinary hesitancy, six weeks of night sweats with 15 pound weight loss and several days of change in voice quality. Exam revealed muffled voice, stridor, and prognathism in addition to suprapubic and prostatic tenderness. Admission labs revealed WBC 9.8K without lymphopenia, BUN 66 and Cr 5.0. Laryngoscopy revealed supraglottitis; he was also diagnosed with prostatitis, both of which improved on antibiotics. HIV screen was positive and confirmed by Western Blot; CD4 count was 184 and viral load 35500 copies/mL. He was discharged on antibiotics with planned outpatient infectious disease and urology follow-up.

Mr. P returned in three days with worsening stridor and was found to have impaired tongue protrusion and lateral motion. Repeat laryngoscopy showed adenoidal and lingual tonsillar hypertrophy, with apparent resolution of supraglottitis. Empiric glucocorticoids were started for worsening stridor. MRI of the head showed osseous lesions throughout the calvarium and skull base with right greater than left hypoglossal canal involvement and diffuse soft tissue enhancement. Diffuse lymphadenopathy and lytic bone lesions were found on CT chest/abdomen/pelvis. Subsequent cranial nerve exam demonstrated left tongue deviation. A tracheostomy was ultimately required for worsening airway obstruction. An axillary lymph node was biopsied and found to be consistent with BL. He immediately began chemotherapy in addition to beginning antiretroviral therapy while inpatient.

Approximately 3.5 months after induction chemotherapy, Mr. P achieved remission with resolution of tongue paresis and stridor. HIV viral load became undetectable three months after initiation of antiretroviral therapy.

Discussion: Immunodeficiency-associated BL is most often associated with HIV, and those with HIV are at >200x greater risk for BL. EBV is associated with nearly all cases of African BL but only 40% of immunodeficiency-associated BL. In patients with HIV, BL is observed more commonly when CD4 counts are >200, and unlike most other HIV-associated lymphomas, the incidence has not decreased with the advent of antiretroviral therapy. Immunodeficiency-associated BL confers high risk for CNS involvement and is highly aggressive.

73) CASE REPORT OF AN AGGRESSIVE LYMPHOMA WITH MIXED PLASMA CELL AND T CELL FEATURES IN A PATIENT WITH CROHN'S DISEASE RECEIVING TUMOR NECROSIS FACTOR ALPHA INHIBITORS (TNFI)

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Case History : Our patient is a 62 year male with Crohn's disease who presented with fever, drenching night sweats and increased frequency of watery diarrhea. Patient also reported severe anorexia, cachexia and weight loss. This was his fifth hospital admission in past 30 days with similar symptoms. He has had multiple laparotomies in the past with a total of 128 cm's of small bowel removed till date. Due to disease progression on initially methotrexate and later Adalimumab, he was recently started on a combination of 6 mercaptopurine (6 MP) and Certolizumab. He underwent colonoscopy few days ago, which showed mild active disease at the anastomotic site with normal C- reactive protein on all admissions. Biopsy was negative for cytomegalovirus and inclusion body disease. His stool cultures had repeatedly come back negative for routine bacteria, ova and parasites including giardia and cryptosporidium. Other tests that had come back negative were serum HIV, Epstein - Barr virus, hepatitis panel, cryptococcal antigen and fungal assays. 6 MP levels were found normal as well and discontinuation of this drug did not improve his symptoms. His symptoms were persistent despite empirical treatment with steroids for Crohn's flare and use of cholestyramine. Physical exam during this admission revealed left axillary adenopathy and a 2x2 cm erythematous skin lesion on his scalp. He was also found anemic with hemoglobin of 9 and lactate dehydrogenase was found mildly elevated at 400. Computed tomography (CT) scan of abdomen showed a left flank mass measuring 6 cm x 6 cm with mild splenomegaly and mesenteric adenopathy, latter was also noted on the magnetic resonance imaging (MRI), 2 years ago and was thought to be reactive at that point. Biopsy of the left flank mass showed an unusual combination of cell surface markers with presence of both T cell (CD 3) and plasma cell (CD 38, 138 and Mum 1) markers. This sample was also sent for second opinion at a quaternary center where findings were confirmed and was diagnosed as "aggressive plasmablastic lymphoma with aberrant expression of T cell markers". Bone marrow and cerebrospinal fluid analysis came back negative for lymphoma. Biopsy of scalp lesion revealed squamous cell skin cancer. Patient opted for comfort care after carefully evaluating all his treatment options and expired peacefully within next 2-3 days.

Discussion: The activity of tumor necrosis factor alpha (TNF) against tumors in laboratory models and, potentially, in humans raises the possibility that TNFI might potentiate the clinical risk of malignancy. There are studies supporting increased incidence of cancers espacially skin cancer and lymphoma in patients receiving TNFI, but their accuracy is questionable due to mixed patient population and frequent co-administration of immunomodulators like 6 MP and azathioprine which are also implicated in the causation of lymphoma. Thus, a more homogeneous sample consisting of patients with crohn's disease treated solely with TNFI is needed to reach accurate results. Our patient's unusual lymphoma with rare atypical characteristics and presence of a concomitant squamous cell skin cancer makes us strongly suspicious of TNFI playing a role in the pathogenesis of these cancers.

74) A CASE OF SUPERIOR SAGITTAL SINUS THROMBOSIS IN PAROXYSMAL NOCTURNAL HEMOGLOBINURIA (PNH)

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Case Presentation: A 21 year-old male presented with a 2 week history of posterior headache located behind his eyes that progressed with muscle spasm of both shoulders, photophobia, and diplopia. Pain was intermittent and worsened with head and neck movement and supine position. He was seen 3 times in clinic and 2 times in local ED. A non-contrast head CT was negative for any acute process. He received symptomatic treatments without relief. The patient had no prior medical history and took no medications. There was no report of prior headache, fever, chills, recent camping/hiking, tick bites, viral illnesses, or sick contact. Physical exam revealed a young man with normal vital signs in mild distress. Notable findings include bilateral papilledema, limited abduction and adduction in right eye, and horizontal diplopia worse in rightward gaze. Gait was slow and cautious. Initial labs were notable for hemoglobin 12.9 g/dL, hematocrit 35%, platelet $48 \times 10^3/L$, creatinine 0.63, fibrinogen at 507, PTT 23.5, INR 0.9, D-dimer 1.62. Head MRI showed acute thrombosis of posterior superior sagittal sinus. EKG, CXR and UA were normal. Upon admission, a thorough hypercoagulable workup was performed with assistance from hematology service. He was placed on therapeutic LMWH. Workup showed negative ANA and ANCA, Hep B and C, CMV, EBV and monospot. Haptoglobin was elevated and LDH, Anti-cardiolipin and beta-2-glycoprotein were normal. Absolute retic, % retic were 65 and 1.6% respectively. A peripheral blood smear showed RBC with fragments and spiculated cells. Ultrasound did not show splenomegaly. MRI of cervical and thoracic spine showed patchy and diffusely abnormal marrow signal in C2, T2 and T8 concerning for malignancy. Bone marrow biopsy showed diffuse hypocellularity without dysplasia or increased blast cells. PNH was suspected and confirmed on peripheral flow cytometry. The patient was transitioned to oral rivaroxaban and began induction with eculizumab.

Conclusion: Paroxysmal nocturnal hemoglobinuria is a rare disease with manifestations including hemolysis, mostly venous thrombosis with bone-marrow failure. In patients presenting with unexplained venous thrombosis, PNH should be on the differential. Diagnosis is easily made with peripheral blood by flow cytometry showing deficiency of GPI-APs of blood cells. Treatment includes long term anticoagulation and the FDA approved humanized monoclonal antibody eculizumab.

75) CRYPTOGENIC BRAIN ABSCESS FROM A UNIQUE FOCUS

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Introduction: Patent foramen ovale (PFO) is a common congenital heart defect (35% of the population). Brain abscess is a relatively uncommon but potentially devastating disease. 20-30% of cases have an unknown primary source (Cryptogenic Brain abscess – CBA) and some of these patients have PFO with right-left shunt possibly allowing bacteria to bypass the pulmonary circulation.

Case Presentation: We present the case of a 56 yo male presenting with altered mentation, septic shock and elevated liver function tests. Abdominal imaging revealed a 9.8 cm heterogeneous liver mass. Despite appropriate empiric antibiotics, his neurologic status deteriorated. Brain MRI revealed numerous peripheral enhancing lesions suggestive of brain abscesses. Blood and liver aspirate cultures grew Alpha hemolytic Streptococci. TEE revealed a large PFO with a bidirectional shunt and no evidence of endocarditis. He made a full recovery after a prolonged course of intravenous antibiotics. PFO closure was done several months later after discussing with the patient.

Discussion: Upon literature review, only 15 other cases of CBA have been published in the past 20 years. While 60% of cases had a primary source of infection in the oral cavity, to the best of our knowledge, ours is the first case with a primary infectious liver nidus. All the cases reviewed were treated with IV antibiotics but there was no consensus on PFO closure. Randomized studies in cryptogenic stroke (similar pathophysiology) suggested no benefit with PFO closure. As such, we feel that PFO closure after CBA should be considered for primary prophylaxis of cryptogenic stroke in patients at high risk, as calculated by RoPE score and/or echocardiographic criteria.

Conclusion: Routine PFO closure is controversial and likely not beneficial. Future prospective studies are necessary to clarify optimal management.

76) A SHOT THROUGH THE HEART

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An estimated 60,000 people receive prosthetic heart valves every year in the United States. This life-saving intervention is associated with several life threatening complications including complicated infections.

A 72 year old Caucasian male with diabetes and a prosthetic aortic valve for aortic stenosis had increasing somnolence and weakness resulting in multiple falls over the previous several weeks. He was taking aspirin, warfarin and several anti-hypertensives. During this time, he had a slowly improving diarrhea illness and his diet was relatively minimal. Recently, he had been seen for chest pain determined to be rib fractures sustained from a fall. At presentation he was febrile with significantly decreased mental status and non-fluent aphasia. Initial workup up demonstrated a WBC of 15, troponin I of 18, creatinine of 2 and an INR of 3.6. A non-contrast head CT was performed and read as acute vs subacute ischemic stroke of the left temporal lobe. A transthoracic echocardiogram was obtained which was poor quality. Multiple blood cultures identified *Enterococcus faecalis*. IV antibiotic treatment was optimized for likely prosthetic valve endocarditis and a transesophageal echocardiogram was scheduled. Prior to this, he began to have episodes of junctional tachycardia with a new bundle branch block. TEE confirmed prosthetic valve endocarditis and showed extension of the infection into the intervalvular fibrosa with a large mobile vegetation on the mitral annulus. Further medical stabilization was needed before surgical intervention was an option. However, soon after the extent of his infectious process was discovered, he developed right upper quadrant abdominal pain and distension. CT identified multiple splenic lesions representing septic emboli. Unfortunately, he died within 8 weeks of initial diagnosis.

Multivalvular endocarditis is rare and represents an estimated 15% of all cases of infectious endocarditis. The aortic and mitral valves are the most common sites among these. Multivalvular endocarditis has similar in-hospital mortality to single valve endocarditis but is associated with a significantly increased morbidity including heart failure, perivalvular abnormalities and need for surgery. Notably, neurologic complications significantly increase the perioperative risk for hemorrhagic conversion making acute management decisions increasingly difficult. This case underscores the importance of prompt diagnosis and high clinical suspicion when evaluating any patient with prosthetic heart valves and positive blood cultures.

77) A RARE CASE OF DELIRIUM ASSOCIATED WITH CROWNED DENS SYNDROME

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Introduction: Crowned dens syndrome (CDS) is a rare and under-recognized cause of acute neck pain. CDS is characterized by severe cervico-occipital neck pain associated with deposition of calcium pyrophosphate dihydrate (CPPD) around the odontoid process (or dens) to give the appearance of a crown on imaging. Because CDS is often associated with neck stiffness, fever, and elevated inflammatory markers, it can be misdiagnosed as other conditions such as meningitis, cervical spondylitis, or polymyalgia rheumatica, thus delaying diagnosis and appropriate treatment.

Case: An 87 year old male with history of hypertension, diabetes, and chronic kidney disease presented with acute delirium on top of gradual cognitive decline. Patient's family noted increased confusion and severe neck pain in the past week. On exam, the patient had fever to 101oF and was oriented only to self. He had significant neck tenderness and stiffness with restricted cervical range of motion but no focal neurological deficits. He also had active synovitis to the bilateral knees and wrists. Labs revealed normal white count and elevated acute phase reactants with sedimentation rate of 82 mm/hr and C-reactive protein 22.8 mg/dl. Infectious workup with urinalysis, blood cultures, and chest radiography were negative. A lumbar puncture was attempted but unsuccessful. Due to the patient's persistent severe neck pain, CT and MRI cervical spine were performed, revealing calcium deposition around the odontoid process, consistent with crowned dens syndrome. Left knee arthrocentesis was subsequently performed with CPPD crystals on synovial fluid analysis suggestive of acute pseudogout. The patient was started on Prednisone 30 mg daily with subsequent taper for treatment of crowned dens syndrome related to CPPD and polyarticular pseudogout with rapid and significant improvement in mental status and neck and joint pain.

Discussion: Crowned dens syndrome can present with a pseudo-meningitis picture with acute neck pain and fever. CT of the cervical spine is the gold standard imaging modality with classic finding of calcium deposition in a crown around the dens. The majority of patients with CDS also have chondrocalcinosis with CPPD deposition within primary sites (eg knees, wrists, ankles). Treatment consists of NSAIDs or steroids, with dramatic improvement in symptoms and excellent prognosis. Thus, CDS should be considered in the differential with this clinical picture to avoid unnecessary invasive procedures and allow for appropriate diagnosis and rapid initiation of targeted treatment.



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