PARTICIPATING INSTITUTIONS:

- Atlantic Health System
  - Morristown Memorial Hospital
  - Overlook Hospital
- Atlanticare Regional Medical Center
- Barnabas Health System
  - Monmouth Medical Center
  - Newark Beth Israel Medical Center
  - Saint Barnabas Medical Center
- Capital Health System
- Drexel University College of Medicine, Saint Peter’s University Hospital
- HUMC Mountainside Hospital
- Meridian Health System
  - Jersey Shore University Medical Center
- Mount Sinai School of Medicine
  - Englewood
  - Jersey City Medical Center
- Palisades Medical Center
- Raritan Bay Medical Center
- Rowan University
  - Cooper Medical School
  - School of Osteopathic Medicine
- Rutgers
  - Robert Wood Johnson Medical School
  - New Jersey Medical School
- Seton Hall University School of Health and Sciences Medical Science
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It is assumed that all participants adhered to the rules as stated in the original abstract submission form. It is also assumed that the abstracts submitted were original works, represented by the true authors. The abstracts appear in no particular order. Judging was performed in an attempt to minimize bias. Judges were unaware of the authors or institutions the competitor unless they were directly involved with the associate. Although there were many excellent abstracts those selected to be presented as poster or oral presentation were chosen on the basis of content. This content was felt to be intriguing from a clinical education standpoint, thought provoking, or could stimulate debate regarding our current practice of medicine.
ORAL PRESENTATIONS

Barnabas Health - Monmouth

Accuracy of breast Specific gamma imaging in diagnosis of breast cancer; Preliminary report

Hadie Razjouyan, David Sharon, Aedon Olaso, Shahrooz Tahvilian, Jorge Pardes

Introduction: Breast cancer is one of the most frequently diagnosed cancers and the leading cause of cancer death in females worldwide. With advances in the sensitivity of mammographic screening, the diagnosis of breast tumors has changed considerably. Most screening programs are based on mammogram which has some drawbacks including radiation, false positivity, not being much reliable in breast dense patients like those young age patients. Recently, breast-specific gamma imaging (BSGI) or Molecular Breast Imaging has been introduced in literature with high sensitivity comparable to breast MRI and even better Specificity to MRI. Our center established using this modality recently. Therefore, we would like to check for its accuracy in diagnosing of breast cancer and also would like to look for important contributing factors which may refine present indications of using this modality.

Material and methods: Present study is a retrospective cross-sectional study including all patients who referred to breast center at Monmouth Medical Center between September, 2010 to January, 2013. Data on the patient’s age, date of birth, race, history of smoking or drinking, date of performing BSGI, reason for BSGI, findings of BSGI (presence or absence of uptake), personal history of breast cancer with any subsequent therapy, first degree family history of breast cancer, any prior to exam procedure on breasts, pathological results of prior to and after BSGI, type of procedure after BSGI, pathologic staging including T, N and M. estrogen, progesterone receptor status, her2neu status, other modality findings like corresponding ultrasound, mammography and MRI reports, if performed. BSGI examinations are interpreted in the clinical setting by one radiologist experienced in BSGI interpretation. The pathology report was considered as gold standard. Results: Hundred and 4 patients had BSGI at Monmouth Medical Center [Mean age (SD): 60.7 (11.9), age range: 33-89]. 61 did not have any uptake (58.7%) and 42 had suspicious uptake and one of the imaging was incomplete. 80 of these patients were Caucasian female (76.9%). Twenty seven of these patients underwent either biopsy or surgical removal of the breast. Out of 24 positive BSGI, 22 had pathology proven carcinoma and two were normal. Two of the negative BSGI, had positive findings on Pathology. The sensitivity of the test is 91.6% and because of low number of patients, currently calculation of specificity will not be accurate. Conclusions: According to our preliminary result, BSGI sensitivity in our center is acceptable and comparable to previous reports. We need more patients to have estimate of specificity of the test. We will also look for the sensitivity of the test in different subtype of breast carcinoma as we get more patients and also compare the agreement of this method with conventional screening and pre-operation imaging evaluation.
Management of gallstone related complications: The impact of admitting team, insurance and diagnosis on guideline compliance

Gregoris Komodikis, Vishal Jariwala, MBBS

Introduction: Gallstone disease with its related complications is a common emergency presentation. Current guidelines support that laparoscopic cholecystectomy should be performed at index admission. Methods: A retrospective cohort study was performed at Saint Peter’s University Hospital to determine compliance to current recommendations and test for differences in waiting time from admission to cholecystectomy based on (a) admitting service (medicine, surgery), (b) insurance (Private-Medicare, Charity-care, Self-pay, Medicaid) and (c) diagnosis (biliary colic, cholecystitis, choledocholithiasis, pancreatitis). In addition, we evaluated the readmission rate in those cases where cholecystectomy was not done at index admission and identified reasons for not performing the procedure. All patients admitted with gallstone related diagnosis from January 1, 2012, through December 31, 2012 were included. An independent sample t-test was used to test the differences in cholecystectomy waiting time between patients admitted under surgery or medicine service. A one-way ANOVA was used to test for waiting time differences based on admission diagnosis and insurance status. Results: Two hundred and twelve consecutive patients were included for analysis. A total of 166 patients (75.5%) had cholecystectomy at index admission. Nine percent (8/90) of patients admitted under surgery service did not have cholecystectomy at index admission compared to 36% (44/122) under medicine service. There was a statistically significant difference (t (120) = 4.7, p = 0.0001 p<0.05) in average admission to cholecystectomy waiting time for patients admitted under medicine service (M = 3, SD = 2.2) compared to surgery service (M = 1.6, SD = 1.2). Twenty-eight percent (34/123) privately insured patients did not have a procedure at index admission compared to 16% (8/45) with charity care, 23% (8/35) self-pay and 22% (2/9) with Medicaid. Admission to cholecystectomy waiting time based on insurance status was not statistically significant at p<0.05. Admission to cholecystectomy waiting time differed significantly based on admitting diagnosis, F (3.156) = 23.8, p = 0.00001. Post-hoc comparisons indicate that patients with biliary colic (M = 1.61, SD = 1.33) and cholecystitis (M = 1.69, SD = 1.38) waited less time than patients with choledocholithiasis (M = 4, SD = 1.8) and pancreatitis (M = 4.24, SD = 2.3). Fifty-two patients (24.5%) did not have a cholecystectomy from which nine (17.3%) were readmitted within a year. Sixteen patients (36%) that were admitted under medicine service and did not have cholecystectomy at index admission a surgeon was not consulted. Conclusions: Adherence to current recommendations for the management of gallstone related complications is of paramount importance to reduce costs. Patients admitted under medicine service experience unnecessary delays for cholecystectomy and increased risk of not having the procedure at index admission. Suspected biliary disease patients should be admitted directly to surgical service in order to reduce costs of care.
Low diagnostic yield of urinary pneumococcal antigen testing at a large tertiary hospital

Aesha Jobanputra, Shashi Kapadia, M.D., Lata Cherath, M.D., Hayder Hashim, M.D.

Background Streptococcus pneumoniae is a leading cause of community acquired pneumonia. Rapid testing for S. Pneumoniae using the Binax Now® urinary antigen test (UAT) has been used to aid diagnosis. The sensitivity of UAT varies by study but has generally been reported to be between 55 to 100%[1]. The routine use of this testing in affecting therapy or cost-effectiveness remains in question [2]. In this study, we seek characterize UAT diagnostic yield in a sample of patients from a large tertiary hospital. Methods: This was a retrospective chart review. The study population consisted of all adult patients from December 2010 to August 2011 who were admitted to Hackensack University Medical Center (HUMC) and had UAT performed. HUMC laboratory data were used to identify patients. Data collected included age, sex, sputum culture, blood culture, result of UAT, patient origin, and presence of immunocompromise. Descriptive statistics were used on all data. Results: Evaluation of 356 patients revealed that UAT was positive in only 14 samples (3.9%). Rate of positivity did not vary by age or gender. Patient’s status as being from home, hospital or other institution was documented in 319 of the 356 patients. Of this subset, 191 (60%) came from home without recent hospitalization, hemodialysis or immunocompromise. Of these 191 patients with community-acquired pneumonia, a total of 9 (4.7%) had a positive UAT. Eighty patients (25%) had healthcare-associated pneumonia based on transfer from facility, recent hospitalization, or hemodialysis. Of these, only 1 patient had a positive UAT. Forty eight (15%) had immunocompromise from either recent chemotherapy, presence of organ transplant, or HIV. Only 1 one of these had a positive UAT. Of the 14 patients with positive UAT, only 1 had culture-proven S. Pneumoniae. Four had positive cultures for non-pneumococcal bacteria. No patients had cultured S. pneumoniae with a negative UAT. Conclusion: In our study of hospitalized adult patients, UAT for pneumococcal disease had a low diagnostic yield. Given the established sensitivity of the test, this may reflect changes in pneumococcal prevalence in hospitalized patients or overuse of UAT in low-probability patients. Further investigation is needed to elucidate the most appropriate, cost-effective population for UAT testing and interpretation. References 1. Monno, R., et al., Evaluation of a rapid test for the diagnosis of pneumococcal pneumonia. J Microbiol Methods, 2013. 92(2): p. 127-31. 2. Piso, R.J., et al., The routine use of urinary pneumococcal antigen test in hospitalised patients with community acquired pneumonia has limited impact for adjustment of antibiotic treatment. Swiss Med Wkly, 2012. 142: p. w13679.
Information Hand-off and Clinical Outcomes of Critically Ill Patients Transferred Between Facilities

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Background: Transitions of care, whether within or between institutions, are an important source of medical errors, inefficiency, and unnecessary cost. Multiple studies have demonstrated that improving the information quality of patient hand-offs during care transitions is associated with more efficient and less costly care, with improved outcomes, and with improved patient satisfaction. An understudied area of transitions of care involves patients transferred between facilities. Inter-hospital transfers face multiple unique challenges including uncertainty of when the transition will take place, differing information systems, and geographical distance, which is compounded by patient acuity and complexity. This study is designed to investigate whether the quality of patient hand-off impacts outcomes for patients transferred between facilities. Methods: The Inter-Hospital Transfer study (IHTS) is a retrospective observational study of all consecutive patients transferred to Robert Wood Johnson University Hospital (RWJ) MICU, SICU, CCU from December 2011 through December 2012. Patient records were scored for the presence and completeness of discharge summary, history and physical, progress notes, consultations, images, and medication reconciliation (0 points if absent, 1 point if incomplete, 2 points if complete). The final transfer score was a percentage of the total possible points following two independent reviews. The primary outcome was in-hospital mortality. Secondary endpoints included adverse outcomes, a measure of overutilization (that included patients being transferred to another service within 24 hours and duplicated labor), and length of stay. Severity of illness was adjusted by MPM0-III, and statistical significance was inferred with a p<0.05 by multivariate logistic regression. Results: 328 patients were transferred to RWJ ICUs from 37 outside hospitals between December 2011 and December 2012. Patients were transferred with documents containing on average 66 pages with a median transfer score of 60 out of a possible 100. When controlling for severity of illness, patients transferred with higher transfer score had significantly decreased mortality by logistic regression (P=0.022, 95% CI -2.73 to -0.21). Similarly, higher transfer scores were associated with decreased adverse events within 24 hours (p<0.001, 95% CI -3.60 to -1.41) and over-utilization (p=0.001, 95% -3.18 to -0.80). These results remained significant when controlling for each individual unit they arrived. Multivariate logistic regression showed that by itself, the transfer of images was an independent predictor of reduced in-hospital mortality (p=0.017). Discussion: These data highlight the impact of information quality on outcomes during the transfer of critically ill patients between facilities. Currently, there are no guidelines detailing how to coordinate information during inter-hospital transfers. The significance of the transfer score demonstrates EMTALA forms and verbal sign-out are not sufficient on their own. Systematic improvement of the hand-off process in these cases has the potential to improve patient important outcomes and provide higher value care.
Sitosterolemia is a rare, autosomal recessively inherited lipid metabolic disorder. It is characterized by hyper absorption and decreased biliary excretion of dietary sterols leading to elevated phytosterol levels, tendon and tuberous xanthomas and premature development of atherosclerosis. We present a case of sitosterolemia in a young woman. A 45-year-old woman with history of hypothyroidism and mitral valve prolapse presented to her cardiologist because of dyspnea and precordial chest pain that worsened with physical activity and improved with rest. She never smoked, used alcohol or recreational drugs. Her mother had a myocardial infarction at age 67. Her only medication was levothyroxine 25 mcg daily. Review of systems was unremarkable. Physical exam was normal. Electrocardiogram and echocardiography were normal. Total cholesterol was 177 mg/dL, triglycerides 84 mg/dL, HDL 69 mg/dL and calculated LDL was 91 mg/dL. VLDL was normal. She achieved 94% of maximal HR on a stress test at 5.5 METS. Post-exercise EKG showed sinus tachycardia with 1 mm horizontal ST depression in inferolateral leads. She also complained of chest tightness. The stress test was suggestive of ischemia. She was referred for cardiac catheterization. The cardiac catheterization demonstrated a 30% Left main coronary stenosis, and multiple stenosis in the left anterior descending and circumflex arteries, rated at 30 to 50%, and mild stenosis in the right coronary artery. Because of the extent of the coronary artery disease in a young woman with few risk factors and normal lipids, a Boston Heart diagnostics test was ordered. This showed normal production markers of cholesterol, normal lipids levels, but high absorption markers of beta-sitosterol and campesterol absorption with values in the 99th percentile. Ezetemibe 10 mg once daily was added. A repeat test 6 weeks later showed borderline absorption markers of phytosterols. Given the very high levels of sterols indicating abnormal absorption, genetic testing was done in search of a mutation in the ABCG5/G8 proteins. She was found to be compound heterozygote for ABCG5/8 mutation. Sitosterolemia is a rare but easily treatable disease that causes premature coronary artery disease. It is a rare autosomal recessive disorder with only about 50 families being identified so far word-wide. It caused by a mutation in both copies of the ABCG 5/8 genes. Individuals with mutations ABCG5/8 proteins have dramatically elevated serum phytosterol concentrations as a consequence of increased intestinal absorption and decreased biliary excretion of phytosterols. Sitosterolemia is usually clinically characterized by xanthomatosis and premature atherosclerosis, which often causes the early onset of CVD. Treatment includes restricting food containing plant sterols. Ezetemibe is effective to reduce sterols absorption. This diagnosis should be considered in any patient with otherwise unexplained premature coronary artery disease.
Antibiotic impregnated bone-cement spacers (AIBS) are successful adjuncts therapy for the treatment of infected hip or knee arthroplasties. Aminoglycosides such as tobramycin have undetectable blood levels while still maintaining supratherapeutic concentrations in the joint space. There have been rare reports of nephrotoxicity with use of AIBS. We present a case of biopsy-proven tobramycin-induced acute interstitial nephritis with tubular injury progressing to end stage kidney disease (ESKD). A 71-year old male with congestive heart failure, coronary artery disease, hypertension, dyslipidemia, and chronic kidney disease stage three (baseline creatinine of 1.6 mg/dl) had a total knee arthroplasty for severe osteoarthritis. Within a month of surgery, he developed a left knee effusion that required incision, drainage and exchange of the polyethylene liner. Direct cultures from affected joint were positive for methicillin-sensitive Staphylococcus aureus, that was unsuccessfully treated with vancomycin and oral daptomycin. He underwent left knee prosthesis removal and implantation of AIBS. The cement spacer was impregnated with 9 grams each of tobramycin and vancomycin. A steady upward trend in serum creatinine was noted over the next few days that did not respond to fluid hydration and conservative therapy. Laboratory studies revealed creatinine level up to 3.6 mg/dl. Fractional excretion of sodium was 4%. Urinalysis showed persistent mild proteinuria, but no casts, hematuria or eosinophilia. No urinary tract obstruction was seen on ultrasound. Vancomycin level was 1.0 ug/ml while tobramycin was markedly elevated at 2.7 ug/ml. The cement spacer was removed with subsequent decline in tobramycin levels. Creatinine levels remained stable. Kidney biopsy demonstrated acute tubular injury and interstitial inflammation with numerous eosinophils. Despite initial stabilization of renal function, he developed ESKD over the subsequent few months. Tobramycin-impregnated joint spacers are helpful as adjunct therapy for the treatment of infected prosthetic joints and usually do not cause elevation of serum drug levels grater then 0.2 ug/ml at 48h or cause nephrotoxicity. Our case is one of the few reported in which AIBS caused nephrotoxicity by interstitial nephritis. We recognize the need for further studies to address optimal combinations and concentrations of AIBS to avoid kidney injury. We also recommend the routine monitoring of serum creatinine and drug levels of the aminoglycoside used in the impregnated spacer to prevent this complication.
Acute Vertebral Artery Dissection after Chiropractic Manipulation

Sujitha Nandimandalam, Dominick Zampino DO FACP

Vertebral artery dissection (VAD) is a relatively rare but increasingly identified etiology of stroke in young and otherwise healthy individuals. The annual incidence of VAD is around 1-1.5 per 100,000. We describe a rare instance of posterior inferior cerebellar artery (PICA) stroke from VAD instantly after chiropractic cervical spine manipulation. A 38 year-old female with history of hypertension presented with sudden onset of profound dizziness, moderate neck pain, nausea, vomiting, right arm weakness and ataxia immediately following cervical manipulation at a chiropractic clinic. In the emergency department she was also noted to have significant dysmetria of the right arm on finger-nose test. Stat magnetic resonance imaging of the brain revealed an acute infarct in the vascular territory of the PICA. Magnetic resonance angiography (MRA) of the neck revealed evidence of significant narrowing at the origin and distal course of the right vertebral artery along with some irregularity and narrowing about the course of the left vertebral artery. MRA of the head was unremarkable. She was anticoagulated with intravenous unfractionated heparin. Computed tomography (CT) of the head on day 2 revealed mild mass effect with slight compression of the fourth ventricle. A repeat CT of the head was obtained on day 4 which revealed stable infarct with no mass effect, midline shift or hemorrhagic conversion. Cerebral angiography done on day 9 was consistent with dissection and complete occlusion of V3 and V4 segments of the right vertebral artery. She was anticoagulated with warfarin to a goal INR between 2 and 3. She progressively showed improvement in her clinical presentation and was subsequently discharged to home with outpatient physical therapy on day 12. Chiropractic manipulation of the neck has been associated with VAD, however there is significant controversy about the level of risk of stroke from neck manipulation. The epidemiology of these injuries is difficult to ascertain. It has been estimated that 1 in 20,000 manipulations causes stroke. Although the relationship may be causative, it has been theorized that the event actually began pre-manipulation and it is the symptoms of this event that prompted the patients to seek chiropractic care. Further investigation and review may help to better understand this infrequent but potentially devastating diagnosis.
Inappropriate use of d-dimer and Wells score to diagnose Acute Pulmonary Embolism

Mihir Barvalia, Reenal Patel, Ahmed Seliem, Rahul Patel, Robert Lahita

Introduction Acute pulmonary embolism (PE) remains a life threatening condition if not recognized early. Several clinical criteria have been developed to determine pre-test probability of pulmonary embolism such as Wells score. However, it is not as frequently used and thus has led to overutilization of Computerized tomography (CT) pulmonary angiogram. The role of d-dimer testing is uncertain when it is greater than 0.5 µg/ml. Methods This study was a retrospective analysis of 588 CT pulmonary angiograms ordered between March 1, 2013 and August 31st 2013 at Newark Beth Israel Medical Center. We excluded 23 angiograms due to incomplete data or indeterminate CT pulmonary angiogram result. Imaging data was compiled using Picture Archiving and Communication System (PACS). Electronic medical record system was used to determine Wells score and clinical features such as age, sex, hypoxia and smoking status. D-dimer levels were calculated in only 258 patients and categorized into low (<0.5), intermediate (0.5 to 2) and high (>2) levels. Investigators were blinded about the CT result while performing chart review. Results PE was positive in 54 out of 565 CT pulmonary angiograms which is a positivity rate of 9.6% (8.9% in the emergency department versus 11.1% in the inpatient group). Chi-square test confirmed significant correlation of PE with both Wells score ($\chi^2=142, p<0.001$) and d-dimer levels ($\chi^2=37, p<0.001$). In patients with low, moderate, and high Wells score PE was diagnosed in 3.5%, 23.9% and 72.7% respectively. The rate of PE was 3.8% in d-dimer<2 versus 30.6% in d-dimer>2 which was statistically significant ($Z=-4.78, p<0.0001$). Mean d-dimer was 5 in patients with PE and 2.1 in patients without PE ($p<0.0001$). Furthermore, the yield of CT pulmonary angiogram was only 1.2% (2 positive out of 173) in patients with low/intermediate d-dimer levels and low Wells score. Conclusion Our study found that patients with low and intermediate d-dimer levels (< 2 µg/ml) have very low likelihood of having acute pulmonary embolism. Moreover, in patients with low wells score along with low or intermediate d-dimer level pulmonary embolism is even less likely. Hence, combining both d-dimer level and Wells score helps improve the pre-test probability of PE. A cost-effectiveness analysis would be useful to determine the d-dimer level at which it is cost-effective to not order a CT pulmonary angiogram. This can greatly reduce unnecessary use of CT pulmonary angiogram.
Introduction  Rhinoscleroma is a chronic, granulomatous, infection of the respiratory tract mucosa caused by Klebsiella rhinoscleromatosis. It is endemic to certain areas Africa, Latin America, Europe, and Asia, but rarely seen in the United States.1 We present a case of rhinoscleroma in an immigrant from Ecuador.  

Case Report  A 29-year-old otherwise healthy man from Ecuador, residing in New Jersey for seven years presented to our medical center with complaints of hoarseness, dysphagia, odynophagia, and dyspnea for three days. Inspiratory stridor and a pedunculated uvular mass were noted on examination. Laryngoscopy revealed supraglottic stenosis with granulomatous changes and limited visualization of the glottis. An emergent tracheostomy was performed due to acute decompensation of respiratory status. He was subsequently taken to the operating room for direct microscopic laryngoscopy with biopsy and mechanical debridement of the obstructing supraglottic granulomatous lesions. Biopsy revealed histiocytoid cells, plasma cells, and lymphocytes. Brown-Brenn stains revealed gram negative rods and culture grew K. Rhinoscleromatis.  

Discussion  Rhinoscleroma was first described by Hebra in 1870, 2 Patients tend to be between 15 and 35 years of age, and women are slightly more affected than men. 3 Scleromas are most often located in the nasal fossae (95%), with laryngeal scleromas, being seen in 15-80% of cases. It is endemic to parts of Africa, Southeast Asia, Central and South America, and Eastern Europe.2 Outside of the endemic regions, rhinoscleroma is rare.4 The disease typically progresses through three stages. In stage I, the rhinitis stage, patients experience catarrhal symptoms and mucopurulent nasal discharge. In Stage II, the granulomatous stage, nodular granulomatous lesions proliferate and fill the nasal fossae. Development of fibrosis and scarring occurs during stage III, the cicatricial stage. 5 The pathogenesis of rhinoscleroma is unclear but an altered CD4/CD8 ratio in lesions supports the role of impaired cellular immunity.4 Diagnosis is made by the presence of subepithelial Mikulicz cells containing rod shaped bacilli, and confirmed by positive cultures for K. Rhinoscleromatosis (most often seen in sate II).5 Once confirmed treatment with tetracycline, ciprofloxacin, or rifampin should be started promptly to reduce risk of recurrence which has been reported to occur in to 41% of patients.4 Airway compromise and severe deformity are some of the most adverse consequences of K.Rhinoscleroma and can be managed with surgery and laser therapy. 4 Conclusion  While rhinoscleroma has been reported in over 68 countries, it is rarely seen in the United States. However, the incidence appears to be increasing due to immigration from endemic areas.1 Often presentation is nonspecific and resembles chronic rhinitis. Rhinoscleroma should be considered in patients with non-responsive nasal symptoms, nasal obstruction, or polyps with a predilection for the nasal septum with relative sparing of the sinuses, particularly in immigrants from endemic areas.3
The Case of Masked Left Shoulder Tendinitis in a Patient with CAD

Andrey Samal, Phaniram Sumanam, MD; Daniel Goldsmith, MD

BACKGROUND  Evaluation of chest pain is a crucial function of general internists, and though many etiologies are benign, clinicians must be cautious not to miss serious disease such as coronary artery disease (CAD), aortic dissection, splenic rapture and others. Famously, the presence of reproducibility of the chest pain on palpation has been shown not to lower the post-test probability of coronary disease. Occasionally, however, a musculoskeletal cause can co-exist with more worrisome disease, requiring careful clinical assessment and judgement.  CASE  56-year-old caucasian male presented complaining of worsening intermittent left sided chest discomfort for three weeks, radiating to the left shoulder, aggravated by movement and alleviated by rest, associated with palpitations. Over the past three years he was diagnosed with HTN, dyslipidemia, GERD, CAD, MI, and ischemic cardiomyopathy (stage C, class 2) with EF of 25-30% and had been managed with optimal medical therapy, PCI and stent placement, and eventual pacemaker placement. The patient attributed his chest discomfort and shoulder pain to his heart condition, although on this admission his shoulder pain was somewhat worse than usual. The patient was taking carvedilol, isosorbide, simvastatin, lisinopril, omeprazole and enteric coated aspirin. The vitals were BP 132/92, RR 18, pulse 66, temperature 98.1 and oxygen saturation 97% on room air. Physical exam was unremarkable, except left sided chest wall and anterior shoulder tenderness to palpation, with moderately diminished range of motion in left shoulder. Neer’s test, cross-over impingement and Apley’s scratch tests were positive. The positive empty can test indicated mild weakness of the left supraspinatus muscle. Left shoulder X-ray showed calcific peritendinitis with mild acromioclavicular joint arthritic disease. Three negative sets of troponin ruled out myocardial infarction and 2D-echo re-demonstrated cardiomyopathy with EF of 15-20% and apical, distal lateral and septal akinesis. Nuclear stress test noted no ischemia, scarring of the anterior septal and inferior walls. Upon discharge the patient was followed up at clinic where he continued his home medications. He was administered intra-articular corticosteroid injection and was referred to physical therapy. NSAID’s were avoided because of his severe cardiovascular disease. This management resulted in significant relieve of his chest discomfort and shoulder pain and improvement of his shoulder function.  CONCLUSION This case of calcified peritendinitis illustrates the importance of clinical assessment as well radiological confirmation of musculoskeletal abnormalities in the patient with cardiac and gastro-esophageal comorbidities. Once a serious visceral etiology is ruled out, it is important to continue investigating because patients will continue to voice complaints which in the setting of known ischemic cardiomyopathy, will result in repeated admissions. In our case, a benign cause of shoulder pain was masked by significant visceral pathology, however directed evaluation and management resolved the orthopedic problem without need for more invasive testing.
Inflammatory Pseudotumor Following Anti-Thymocyte Globulin in a Patient with Severe Aplastic Anemia

Andres Bran

Introduction  Severe aplastic anemia (SAA) can be acute and fatal. Patients can present with uncontrollable hemorrhage complicated by severe sepsis, or it can be less severe presenting with only mild complications of thrombocytopenia and anemia. The preferred treatment depending on age is immunotherapy with antithymocyte globulin plus cyclosporine. If ineffective, patients can undergo bone marrow (BM) transplantation.  Case Presentation   A 43-year-old male from Guatemala with no past medical history presented to the emergency room complaining of bleeding gums, bruising and fatigue for two weeks. Two weeks prior to admission he noticed mild intermittent bleeding of his gums and excessive bruising, which gradually worsened. He did not report history of recent travel, infections and was on no medications. He worked in landscaping with no exposure to chemicals. He did have history of exposure to organophosphates for four years at a sugar cane plantation in Guatemala more than 10 years ago.  On admission patient he was afebrile, pale, with bleeding gums and petechiae on both upper and lower extremities. Laboratory studies revealed severe pancytopenia (WBC 1.1, Hb 4.8, Hct: 13.0 PLT 2), with an absolute neutrophil count (ANC) of 264 cells/mL. Peripheral blood smear confirmed markedly decreased platelet count, and anisocytosis with slight hypo and hyperchromia of RBCs. Hepatitis panel, HIV, EBV, flow cytometry, leukemia and lymphoma panel were negative. Folate, vitamin B12 and iron studies were normal. BM biopsy revealed hypocellularity with decreased hematopoiesis suggestive of aplasia. Supportive therapy with leukoreduced and irradiated blood products was given. Cyclosporine and antithymocyte globulin were started. Patient was discharged home with low but stable counts.  Three days after discharge he returned with fever, generalized malaise and diffuse lymphadenopathy. Broad-spectrum antibiotics were given empirically. The lymph node biopsy showed vascular, spindle cell, and histiocytic proliferation involving the capsule and extending focally into perinodal soft tissue. Focal collections of neutrophils were noted. Diagnosis of inflammatory pseudotumor was made.  Conclusion  Inflammatory pseudotumor of lymph nodes with distinctive pattern of nodal reaction can happen after antithymocyte globulin. The management of SAA patients remains challenging, both acutely in addressing the immediate consequences of pancytopenia and in the long term because of the disease’s natural history and the consequences of therapy that unfortunately like the disease can at times be fatal.  References:  1. Canioni. D, et al. Lymphadenopathy in renal transplant patients treated with immunosuppressive antibodies (OKT3 and anti-thymocyte globulin). A report of nine cases. Am J Surg Pathol. 1989 Feb; 13(2). 2. Marsh JC, Ball SE, Cavenagh J, et al. Guidelines for the diagnosis and management of aplastic anaemia. Br J Haematol 147(1).
Recreational Nitrous Oxide Use: A New Addiction with Serious Complications

Lloyd Centino, V. Florou MD

INTRODUCTION Vitamin B12 (VitB12) deficiency is a known cause of subacute combined degeneration (SACD) of the posterior and lateral columns of the spinal cord. The use of recreational nitrous oxide is becoming an emerging cause of this deficiency. We report two cases of myelopathy, related to VitB12 deficiency from nitrous oxide abuse. CASES Two young men, 24 and 29-years-old presented with neurologic symptoms after regularly inhaling nitrous oxide (NO) via a “whippit” for three months. The first patient complained of bilateral ascending numbness up to the nipple area for two weeks, without associated weakness and incontinence. Neurologic examination revealed diminished pinprick sensation up to the T4 level; and vibratory and proprioception deficits in both lower extremities. The second patient complained of gradually worsening of “pins and needles” sensation all over his body for two months associated with intermittent urinary incontinence. His examination revealed decreased temperature and pain sensation in both upper extremities, hyperreflexia and presence of Babinski reflex bilaterally. Both patients had no recent history of fever, respiratory infection or any episodes of diarrhea. They had ataxic gait, positive Romberg test and normal motor strength. CBC and CSF studies were normal in both. The vitB12 level were 377 and <159 (normal; 239-931 pg/mL) for the first and second patient, respectively. MRI for the first patient revealed increase intensity at the posterior column from C2-C5 level, while MRI of the second patient was unremarkable. Both patients were treated with high doses of VitB12 intramuscular injections followed by oral VitB12 supplement. They were discharged with gradual improvement of their symptoms. DISCUSSION NO is now a commonly abused drug among young individuals due to its easy availability. Currently, there is limited number of cases describing the effects of recreational NO use on VitB12 level. Biochemically, NO causes inactivation of VitB12 and causes functional VitB12 deficiency leading to SACD of the posterior and lateral columns of the spinal cord. Our first patient presented with more of posterior column myelopathy while the second patient presented with more of lateral column myelopathy. Furthermore, despite a normal VitB12 level in the first patient, his use of NO led to clinically relevant depletion of VitB12 stores resulting in neurological manifestations. CONCLUSION These case reports highlight the atypical neurological presentation of VitB12 deficiency in patients with significant recreational exposure to NO. A high clinical suspicion in the appropriate clinical context is the key to diagnosis and treatment in these cases. REFERENCE Pema, P., et.al. Myelopathy Caused by Nitrous Oxide Toxicity. AJNR Am J Neuroradiol 19:894-896, May 1998
Can the time course of SIRSS predict future organ failure in Acute Pancreatitis?

Akshat Kumar, Suresh T. Chari, Santhi S. Vege

Introduction: Systemic Inflammatory Response Syndrome Score (SIRSS) (possible score 0-4) is an easily measured bedside tool. In patients with Acute Pancreatitis (AP), we aimed to precisely quantify the association between SIRSS at day 1 and persistent SIRSS at day 3 with the various adverse outcomes of AP. Further, we analyzed SIRSS patterns for the first 14 days for any prognostic significance. Methods: We prospectively identified patients admitted to Mayo Clinic on day 1 of AP. All patients with positive SIRSS on day 1 were further followed up with daily measurement of SIRSS and organ failure status for 14 days or until discharge, whichever came first. We then correlated positive SIRSS (>2) on day 1, persistent SIRSS on day 3 and the time course of SIRSS with the development of organ failure. Results: SIRSS and persistent SIRSS were associated with all the adverse outcomes of AP with a high sensitivity and negative predictive value, ranging from 73.1 to 100.0%. Persistent SIRSS at day 3 added significantly high specificity to this association (71.7 to 80.0%). All the patients who developed late onset organ failure (3) had the highest possible value of cumulative SIRSS. Conclusions: SIRSS of <2 on day 1 has a high negative predictive value for complications of acute pancreatitis. 80% of patients with persistent SIRSS on day 3 will develop at least one adverse outcome of AP. A new variable "Cumulative SIRSS" was identified that has the potential of reliably predicting late onset persistent organ failure.

References
Thoracic Splenosis presenting as acute onset of chest pain

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OBJECTIVES: 1. Recognize the differential diagnosis of pulmonary nodules. 2. Diagnose thoracic splenosis.

CASE: A 58 year old male presented to the ED with chest pain. PMH: motor vehicle accident at the age of 22 that resulted in a splenectomy and bilateral pneumothoraces with chest tube placement. CMP, CBC, troponins and EKG were negative. Computed tomography (CT) of the chest showed multiple pulmonary nodules scattered throughout the left lower lobe with an irregularity on the left adrenal gland. A PET scan was obtained to evaluate the multiple lesions, but findings were not consistent with malignancy. A CT guided biopsy was performed yielding tissue that was described as fibrovascular with lymphocytes and histocytes, along with a rich vascular network consistent with splenic tissue.

DISCUSSION: Splenosis is a rare and benign condition involving autotransplantation of splenic tissue to another compartment of the body. The most common site is within the abdominal cavity, and the most common cause is splenic trauma with left diaphragmatic laceration resulting in splenic tissue seeding into the thoracic cavity and later developing into active splenic tissue. There have been fewer than 40 reported cases, and the majority these were incidental findings in asymptomatic patients. Because of its rarity, the diagnosis of thoracic splenosis usually involves extensive and invasive diagnostic procedures. New non-invasive imaging utilizing T-99M heat damaged erythrocytes has been used to identify this ectopic tissue. With regards to this case, the findings on the initial CT chest were suggestive of malignancy, therefore the more invasive approach was utilized. Because thoracic splenosis is a relatively benign process, the tissue does not need to be removed. It has been theorized that this ectopic tissue may be able to provide protection from encapsulated microorganisms despite being otherwise asplenic. Thoracic splenosis should be suspected in a patient with any CT or MRI demonstrating left pulmonary nodules with history of splenic trauma. Though new non-invasive studies are available and are preferred to the invasive studies, the CT guided biopsy was done to rule out malignancy found on the initial chest CT.
Do Resident-RN Rounds Reduce calls to the Oncall Person? (D. R. R. O. P.): A Quality Improvement Project

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Introduction: On call ICU/CCU residents receive many calls from nurses on their night shift regarding orders that, in theory, could have been placed by day team residents. These interrupt nursing and physician care, and could distract caregivers attention from more urgent matters. The first objective was to determine the number of, and reasons for, calls from nursing to the on-call night resident. The second objective was to identify the most common reasons, modify the process so that the day team would address these orders prior to leaving, and then resample to determine if this educational intervention reduced the number of calls made to on-call residents. Methods: The ICU night resident logged and categorized calls for 20 nights to determine the baseline. We developed a laminated card listing the common issues, distributed it to the nursing and resident staff to use as a reference during end of shift sign-out rounds, and oriented them to the process. Night calls were reassessed 2 weeks later. We coordinated with the ICU nurse manager, nurse educator, and staff nurses. Results: Baseline: 592 calls over 20 nights, Mean 29.6±8 calls/night, Range 14-46. The top 7 reasons were associated with 81% of the calls: Electrolytes 35%, medication change 15%, ventilator change 11%, order for routine morning labs 11%, insulin 5%, diet/NPO 3%, NG-tube 3%. Re-evaluation: There were 133 calls over 10 nights; a decrease of 18 calls/night [95% CI.. (10,25)]. Calls/night decreased as follows: Electrolytes drop of 3, medication decreased from 4 to 1, routine labs/imaging from 3 to 1. Discussion: A simple program educating the residents to address common orders at the end of the day shift, and nurses to remind the residents to do so, was effective and decreased calls to the night residents by 55%. This decreased the number of times nurses had to interrupt their work flow to contact a resident, and the number of times residents had to address issues at night. This decrease in calls regarding issues that should be addressed by the primary day team was noticed and appreciated by the night resident. The day shift process has been modified, and a repeat survey will be done to assess resident/nursing adherence to this modification. With continued use we anticipate a culture change will result in an additional and sustained decrease in calls for these issues.
Calciphylaxis from uremic and nonuremic etiologies

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Calciphylaxis is a life-threatening syndrome of systemic small vessel calcification and skin. It carries a significantly poor prognosis, up to 80% of patients’ progress rapidly to death after initial presentation, mainly due to infection and sepsis. A 55-year-old male presented with dry gangrene to left ring finger and glans penis, associated with pain to fingers and dysuria. The lesions started two weeks after minor trauma. Past medical history includes sarcoidosis, hypertension, diabetes mellitus, coronary artery disease, IgA nephropathy and interstitial nephritis, nephrotic syndrome, end-stage renal disease, interstitial lung disease, rheumatoid factor (RF) positive arthritis, hyperuricemia, and anemia. Patient was started on prednisone 3 months ago for sarcoidosis. On admission, vitals: BP: 180/87, Pulse: 71, respiratory rate: 18, temperature: 98.1°F. Physical findings included dry gangrene to distal and intermediate phalanges of left ring finger; black eschar to the tip of left middle finger and glans penis. Laboratory tests (reference) showed phosphorus: 6.4 (2.7-4.5 mg/dL), calcium 12.6 (8.6-10.2 mg/dL), hepatitis C antibody: 2.5 (0-0.9 s/co ratio), RF 75 (0-13.9 IU/mL), C3: 62 (90-180 mg/dL), C4: 42 (9-36 mg/dL), PTH: 175 (15-65 pg/ml), beta-2 microglobulin: 6.3 (0.6-2.4 mg/L), d-dimer: 1.03 (<0.5 ug/mL). PET/CT illustrated diffuse sarcoidosis. Mediastinal lymph node biopsy revealed noncaseating granulomas. X-ray revealed extensive vascular calcification of medium and small blood vessels in hands, feet, and penis. Kidney biopsy showed diabetic nephropathy, IgA nephropathy, and acute interstitial nephritis. Echocardiogram demonstrated left ventricle (LV) ejection fraction >55%, but abnormal LV diastolic function, right ventricular systolic pressure of 30-40 mmHg. Arterial Doppler of upper extremities digital and pulse plethysmography demonstrated bilateral peripheral arterial occlusive disease including small vessel disease in both hands. Carotid Doppler showed small amount of calcification to bilateral carotid bulbs and left carotid artery. Amputation was performed to the left ring finger. A suprapubic tube was also placed. The etiology of calciphylaxis remains elusive and may involve multiple co-morbid factors. We report a unique case where end-stage renal failure and multiple non-uremic conditions including sarcoidosis, cryoglobulinemia, and DM may directly or indirectly contribute to the pathogenesis of calciphylaxis. Given the high mortality rate and increasing prevalence, emphasis should be placed on prevention and early diagnosis of vascular calcification, and prophylaxis of secondary infection. Reduction of calcium phosphate product (<60 mg2/dL2), avoidance of prednisone usage, optimized glycemic control, and effective wound care to prevent infection would enhance quality of life and prolong the life span of patient.
INTRODUCTION: Awareness regarding the pathophysiological mechanism of calcium channel antagonist (CCA) and beta blocker (BB) toxicity and judicial implementation of insulin is limited in clinical practice. We report the role of insulin in refractory hypodynamic shock in mixed CCA and BB toxicity.

CASE PRESENTATION: A 56 year old male was admitted to hospital after he was found comatose with empty bottles of Atenolol, Amlodipine, Verapamil and Clonidine. Upon presentation, his HR was 35/min, BP 58/40 mmHg, RR 16/min, T 98.3F and So2- 98%. The patient received Naloxone, Atropine, IV fluids, Glucagon, Calcium gluconate and gastric lavage but bradycardia and hypotension persisted. Thereafter he was placed on transcutaneous pace maker, intubated and was started on vasopressors. Labs were significant for blood glucose-130 mg/dl, BUN-34 mg/dl, Creatinine-2.4 mg/dl, bicarbonate 11mEq/L. Troponins and urine drug toxicology was negative. CXR and CT head were unremarkable.

Despite the resuscitative efforts the patient had refractory hypotension and bradycardia with intrinsic HR between 30-40/min. He was eventually started on high dose insulin drip at 0.5 U/kg/h, dextrose 50% at 50 ml/h. The insulin drip was titrated to a goal MAP > 70 mmHg and heart rate > 80/min. Also serum glucose and potassium levels were monitored every 30 min and dextrose 50% titrated to maintain blood glucose between 110-140 mg/dL.

The patient responded well to the high dose insulin regimen. His heart rate, blood pressure and acidemia improved. He was taken off the pacemaker, vasopressors and insulin drip by day 5 and eventually extubated and discharged to subacute rehabilitation center. DISCUSSION:

Under physiological conditions myocardium generates energy through mitochondrial oxidation of long chain fatty acids (60–70%), glucose (20%) and lactate (10%). During the state of cardiac stress, the energy generation is switched from preferred fatty acid substrate to glucose. Calcium channel and beta blocker toxicity produces metabolic derangements that lead to hypoperfusion and acidemia. This acidemia impairs insulin release in beta cells and impairs glucose uptake and utilization in the myocardium. Therefore therapy with insulin-euglycemia is critical for optimal myocardial functioning. Insulin also increase sympathetic nervous system activity thus further supporting hemodynamics during the state of shock. Most cases of poisoning have limited information. Management decisions are based on presumed drug overdose. It is thus imperative to recognize the therapeutic role of insulin in mixed CCA and BB toxicity.
Not so thrilled by a thrill: a covert complication of arteriovenous fistulas

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Case presentation: Our patient is a 65 year-old gentleman with a history of Hepatitis C, hypertension, smoking, end-stage renal disease on hemodialysis and moderate aortic stenosis who presented with worsening dyspnea over several days. On admission, he was afebrile, with oxygen saturation of 80% on room air, blood pressure of 197/103, and heart rate 95 bpm. On exam, the patient was dyspneic while talking, had bibasilar inspiratory crackles, diminished S2, grade 3/6 systolic murmur heard throughout the precordium radiating to the carotids, 2+ lower extremity pitting edema up to mid-calf, and a right forearm arteriovenous fistula (AVF) with a significantly palpable thrill. Chest x-ray showed cardiomegaly and mild pulmonary vascular congestion. EKG showed sinus rhythm, left ventricular hypertrophy, and prominent positive T waves in the lateral chest leads compatible with a high-volume load state. The patient was placed on oxygen, given Lasix IV, and nitroglycerin drip with some improvement of his symptoms. Aortic valve replacement (AVR) was considered given previous admissions for pulmonary edema. The patient was scheduled for a coronary angiogram prior to AVR to assess for CAD. Coronary angiography showed normal coronary arteries, mean right atrium pressure of 18mmHg, mean aortic pressure 103 mmHg, right ventricular systolic pressure 68 mmHg, mean PA pressure 46mmHg, PCWP 28, normal pulmonary vascular resistance, and cardiac output estimated to be 10.6 L/min by thermodilution. Ultrasound of the AVF access showed an abnormally high velocity with flow volumes over 5 L/min. The patient was successfully treated with banding of the AVF access. Discussion: The patient was diagnosed with high-output heart failure secondary to his arteriovenous fistula. High-output heart failure is an uncommon entity associated with certain pathologic states such as hyperthyroidism, beriberi, skeletal disorders like Paget’s disease and dermatologic disorders such as psoriasis. It is defined as a high cardiac output > 8L/min, cardiac index greater than 2.5 to 4.0 L/min per m2, and low systemic vascular resistance. Cardiac catheterization is usually required for definitive diagnosis. Creation of an AVF has been associated with increased blood flow, pulmonary hypertension and increased cardiac output. Although not usually clinically significant, the increased cardiac output may result in overt heart failure in patients with underlying heart disease or if the AVF flow is greater than 2L/min. The NKF-K guidelines recommend AVF monitoring by physical exam and monthly flow measurements for patients at risk. Treatment involves flow reduction procedures or banding and peritoneal dialysis may be considered. As heart disease is a major risk factor for early death in dialysis patients, the question arises whether an AVF access may contribute to increased mortality. Clinicians should be aware of this complication; early recognition is critical as many cases are reversible (NEJM 367;23 December 6, 2012).
Recalling a Rare Case of Myonecrosis

Sima Patel, Dr. Rajendra Kapila, Dr. Maya Raghuwanshi

INTRODUCTION: A 51-year-old African American female with metastatic invasive ductal breast carcinoma (ER/PR +, Her2 +) received palliative radiation of 30 Gy in 10 fractions to her left femur and left hip-acetabulum. Two months later, she began chemotherapy with gemcitabine and herceptin. Two months after chemotherapy began, she developed worsening discomfort and swelling in her left thigh that left her unable to ambulate, at which point she was sent to the ED for further work-up. On examination, there was tenderness to palpation of the left thigh with diffuse swelling, erythema and warmth. MRI revealed myonecrosis involving the rectus femoris, semitendinosus, and adductor longus muscles as well as extensive edema in the remaining musculature. No abscess collection was seen. Further, findings consistent with metastatic disease were seen in the acetabulum, pelvic bones and proximal femurs bilaterally. Labs on admission revealed an elevated WBC count of 19.8 and elevated serum CPK level of 1128. Patient was initially started on empiric IV antibiotic treatment for possible infectious etiology of myonecrosis. Patient then underwent an ultrasound-guided left thigh muscle biopsy that revealed skeletal muscle showing myonecrosis and focal acute inflammation. Gram stain was negative for bacteria and fungi. All blood cultures were negative to date. Antibiotics were discontinued as her myonecrosis was due to radiation and subsequent chemotherapy. DISCUSSION: Radiation recall refers to a rare inflammatory reaction in a previously irradiated area, potentiated by subsequent exposure to a certain drug. The most common triggers are chemotherapeutic agents such as taxanes and anthracyclines, resulting in a cutaneous reaction. However, more recently, there have been an increasing number of reports that implicate gemcitabine as a cause of radiation recall primarily affecting internal tissues or organs. There are 17 cases reported in the literature of radiation recall associated with gemcitabine resulting in muscle damage specifically; 14 describe myositis and 3 report myonecrosis. The clinical presentation of myonecrosis may mimic or be misdiagnosed as new metastases, musculoskeletal injury, deep venous thrombosis or even a paraneoplastic myopathy. CONCLUSION: This case is presented to alert physicians to consider the rare phenomenon of radiation recall when patients present with muscle complaints following radiation and subsequent chemotherapy. Treatment often consists of withdrawal of the offending agent, although few cases of successful re-challenge have been documented in the literature. One report suggested that oral prednisone may control symptoms and allow continuation of gemcitabine therapy if necessary despite radiation recall.
Reactivation of spinal tuberculosis in a patient with rheumatoid arthritis on low-dose methotrexate

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42 year old Hispanic woman with rheumatoid arthritis who has been taking methotrexate for 4 months was admitted for severe, sharp lumbar back pain for 3 weeks. The pain radiated to both legs, worsened with walking or standing and improved at rest. It was accompanied by low grade fevers, anorexia and generalized fatigue. Patient denied cough or shortness of breath. Spine MRI showed an L4 pathologic deformity with abnormal marrow infiltration and a ring-enhancing lesion within the left psoas muscle suspicious for abscess. CT-guided biopsy of the psoas showed necrotic debris and neutrophilic infiltrates. The PPD test was reported negative. Bacterial and fungal cultures were negative after 5 days. She was discharged with an empiric 30-day regimen of intravenous ceftriaxone and oral linezolid due to high suspicion for an infectious etiology. Twenty-six days later, the patient returned due to worsening of her back pain which rendered her unable to ambulate, and with a 15 pound interval weight loss. Previous biopsy cultures that had been sent for acid-fast bacilli returned positive and another MRI was done. It showed pathologic collapse of L4 with a retropulsive fracture, superimposed phlegmons bilaterally, severe spinal stenosis, and compression of the cauda equina. Three sputum specimens for AFB smears were negative. The patient recalled having had a BCG vaccine and a negative PPD in 1995; she denied exposure to TB or international travel for the past 21 years. The patient was placed on a 6 months regimen of isoniazid, rifampin, pyrazinamide, ethambutol, and pyridoxine. She also required an L4 corpectomy with a vertebral replacement cage, an L2-L4 laminectomy, and L3-L5 fusion. Discussion: Although it is well known that biologic disease-modifying antirheumatic drugs (DMARDs) can reactivate latent Tuberculosis infection (LTBI), most cases are related to TNF-α inhibitor use. To date only 4 cases of tuberculosis reactivation have been reported in connection with methotrexate use. It is therefore imperative to screen high-risk patients for LTBI before starting any DMARDs. Published evidence on the performance of interferon-Gamma release assay (IGRA) suggests that IGRA maintain its diagnostic sensitivity for LTBI better than the tuberculin skin test (TST) in patients with immune-mediated inflammatory diseases (IMID). Routine testing with both TST and IGRA is not recommended. However, results from both tests might be useful when the initial test is negative and the risk for infection and progression to disease is high, as well as in patients with known autoimmune conditions. Since in 20% of cases tuberculosis reactivates at an extrapulmonary site, clinicians must maintain high clinical suspicion for tuberculosis even in the absence of respiratory or constitutional symptoms. Particularly in skeletal TB since there is no evidence of active chest disease in more than half of the cases.
Complication of Reversible Posterior Leukoencephalopathy in the Setting of Suspected Hypertensive Emergency Management

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Reversible Posterior Leukoencephalopathy Syndrome (RPLS) is a clinical syndrome of multiple etiologies defined by the characteristic diffuse subcortical white matter hyperdensities seen on neuroimaging. RPLS is rarely encountered, making appropriate recognition and clinical management a challenge. A 48 year old African American woman with a past medical history of poorly controlled hypertension and poor medical compliance presented with a three week history of worsening vision, leg swelling, and intermittent moderate diffuse headaches worse on the right. Vital signs were significant for a blood pressure of 265/155mmHg. The physical exam revealed only bilateral nonpitting ankle edema. Electrocardiogram showed normal sinus rhythm with left ventricular hypertrophy. Initial lab work was significant for elevated creatinine. Head CT was not performed on initial assessment. Aggressive treatment for suspected hypertensive emergency was begun with an intravenous beta blocker and the patient was admitted. Upon evaluation the next morning, the patient was difficult to arouse with altered mental status. The neurological exam revealed dysarthria as well as focal left sided motor deficits. Blood pressure at this time was 190/100mmHg. A brain attack code was called. CT of the head showed white matter changes suspicious for RPLS which were confirmed on a subsequent MRI significant for diffuse subcortical lesions worse on the right. MRA was within normal limits. There was no evidence of focal hemorrhage or infarction. Intravenous management was begun with a beta blocker, calcium channel blocker, and vasodilator. The patient’s blood pressure stabilized by hospital day three averaging 180/110mmHg. She was then transitioned to an oral antihypertensive regimen and her neurological deficits began steadily improving. By hospital day six she had returned to baseline and was discharged with a blood pressure of 160/90mmHg. The patient remained well on last follow-up ten weeks post-discharge. This case illustrates the sudden onset of severe focal neurologic deficits mimicking stroke in a patient being managed for symptomatic severe hypertension. We hypothesize that the patient had chronic RPLS at presentation, which left her unable to compensate for the rapid iatrogenic decrease in blood pressure and precipitated the observed acute decompensation.

Hypertensive RPLS is believed to develop insidiously as chronically increased pressure overcomes cerebral autoregulation. The result is endothelial dysfunction and disruption of the blood brain barrier with ensuing subcortical parenchymal edema. CT or MRI characteristically show diffuse white matter hyperdensities. MRA is typically normal. The literature recommends management of hypertensive RPLS similarly to hypertensive emergency, with aggressive reduction of the patient’s diastolic pressure to below 110mmHg without exceeding a 25% fall from the presenting mean arterial pressure. While prompt hypertensive management is recommended for the treatment for RPLS, here we present a case where rapid blood pressure correction unexpectedly precipitated focal neurological deficits due to the previously unrecognized condition.
Creutzfeldt-Jakob Disease (CJD) Presenting As Rapidly Progressive Dementia In A Middle-Aged Vegetarian Female

Mark Schwartz, Rory Shalis, MS4 and Gabriela Ferreira, MD

Creutzfeldt-Jakob Disease (CJD) Presenting As Rapidly Progressive Dementia In A Middle-Aged Vegetarian Female  Mark Schwartz, MD PGY2, Rory Shalis, MS4, Gabriela Ferreira, MD, Clinical Assistant Professor, Department of Internal Medicine, Rutgers Robert Wood Johnson Medical School, New Brunswick NJ.  Rapidly progressive dementias (RPD) are neurological conditions with a wide differential diagnosis that develop over weeks to months. In a referral population of 825 patients with RPD (1), 54% were prion related, 28% were undetermined, and 18% were non-prion related. Because of the rapidity of clinical deterioration and lack of therapeutic options for prion diseases, it is imperative that clinicians use a systematic approach to identify treatable causes. A 57 year-old female presented to the hospital with a six month history of rapid cognitive and functional decline, ataxia, and urinary retention causing recurrent urinary tract infections. One year prior to presentation, the patient experienced mood changes, fatigue, and 40 pound weight loss. A psychiatrist started anti-depressant therapy without improvement. A rheumatologist diagnosed the patient with Sjogren's syndrome after labs revealed elevated CRP, anti-RNP and anti-SSA. Steroids were started without any improvement in symptoms. The patient became more ataxic, which resulted in frequent falls. She eventually required assistance with all activities of daily living. Significant history included rheumatoid arthritis, B12 deficiency, and bilateral knee replacements. There was no history of organ transplantation, blood transfusion, or toxic exposure. The patient was a lifelong vegetarian. Family history was negative for neurodegenerative illnesses. On exam, the patient was alert with garbled speech and non-purposeful movements. The patient had a short attention span and labile affect. Neurologic exam revealed normal reflexes, muscle tone and strength without myoclonus or startle reflex. Labs including heavy metal screen and manganese were negative. MRI showed increased signal intensity in the bilateral basal ganglia on FLAIR imaging; DWI was not performed. EEG showed bilateral posterior periodic 1.5-2Hz sharp waves during wakefulness that disappeared during sleep. CSF revealed elevated 14-3-3 protein. Diagnosis of sporadic CJD (sCJD) was made based on the above findings. The patient’s family declined brain biopsy. This case illustrates the diagnostic challenges in RPD and the need for systematic evaluation in order to identify potentially treatable causes, such as autoimmune, infectious, psychiatric, vascular, toxic/metabolic or neoplastic illnesses. sCJD is a rapidly progressing and untreatable neurodegenerative illness that presents as a RPD. This disease is often associated with specific behavioral and constitutional symptoms in its earliest stages. The mechanism of this disease is misfiling of prion proteins that become infectious. Diagnosis relies on clinical suspicion, imaging, EEG, elevated CSF biomarkers, and, ultimately, brain biopsy.  Geschwind MD et al. Rapidly Progressive Dementia. Neurol Clin. 2007;25:783-807.
Anaerobic bacteria are recognized as important pathogens in pleuropulmonary infections. Iatrogenic seeding following an invasive procedure, aspiration of oropharyngeal contents, or bacteremic spread in the pleural cavity are likely routes of infection. Generally, unidentified as primary pathogens in empyema, clostridia are seldom isolated in the absence of other aerobes or anaerobes. We report Clostridium clostridioforme empyema encountered in an otherwise healthy adult who did not have any known risk factors, and clinically recovered after drainage with completion of four weeks of antibiotics. A 66 year old Pakistani male, who has been residing in the United States for three decades, a lifelong non-smoker and non-alcoholic, presented with dyspnea and diaphoresis of one day duration. Two weeks prior to presentation, he had sudden onset pleurisy, and a local cortisone injection was performed. A week later, he developed malaise and dry cough which progressively got worse. He denied symptoms of orthopnea, or hemoptysis. There was no history of recent travel or environmental exposure. He did not have any dental manipulation or history of underlying lung disease. At the time of presentation he was tachycardic, tachypneic, but afebrile and hypoxic while breathing ambient air. Diagnostic tests revealed leukocytosis in blood with predominantly polymorphonuclear cells. A chest radiograph disclosed dense opacification in the right middle and lower lobes, and a computed tomographic imaging of the chest showed a loculated fluid in the right pleural cavity. Empiric piperacillin/tazobactam and vancomycin was initiated. After cardiothoracic surgery evaluation, a tube thoracostomy drained thick, purulent material consistent with exudative pleural effusion. With persistent hypoxia and fluid collection on imaging, he eventually required a video-assisted thoracoscopic surgery with decortication. The pleural fluid culture grew gram positive rods in anaerobic medium in 48 hours identified to be Clostridium clostridioforme. Clostridia are ubiquitous, gram-positive rods which form endospores and exhibit pleomorphism. Clostridium perfringens is the species most frequently associated with human pleuropulmonary infections. Contamination of the pleural space from esophageal rupture, and necrotic lung tissue resulting from a pulmonary embolus with infarction may perpetuate anaerobic or microaerophilic growth of clostridia. Clostridium clostridioforme is primarily an indigenous flora of the lower gastrointestinal tract but has been found on occasion in the subgingival microflora of AIDS patients, vaginal flora of healthy women, and women with bacterial vaginosis. Mortality rate is comparable to that of non-clostridial infections. Most patients respond favorably to appropriate antibiotics and closed chest tube drainage. Penicillin, Cephalosporin, as well as metronidazole, have been shown to be active against all species of clostridia in vitro. C.clostridioforme, however remains a very uncommon pathogen of pleural empyema.
Introduction – Herpes Simplex virus (HSV) is infrequently recognized as a lower respiratory tract pathogen. HSV pneumonitis was first described in 1949, and has been reported mostly in immunosuppressed patients and transmitted by either localized spread from the upper respiratory tract or by hematogenous dissemination from oral or genital mucocutaneous disease. It is rare in immunocompetent individuals and may have a poor outcome if not suspected early. Case report - A healthy 58 year old male presented with fever, dry cough, myalgias and progressive shortness of breath for 1 week. He had been hospitalized a week earlier with similar complaints and treated for community acquired pneumonia with ceftriaxone and azithromycin for 3 days and discharged on levofloxacin. He denied smoking, IV drug abuse or sexually transmitted diseases. His respiratory rate was 22/min with temperature of 101ºF. The oropharynx was normal. Bibasilar rhonchi and bronchial breath sounds were noted. Heart, skin and genitalia were unremarkable. ABG was normal, white count was 20,000, and CPK was 500. CT revealed bibasilar patchy ground glass opacities with emphysematous changes; mediastinal and hilar reactive lymphadenopathy. IV vancomycin and piperacillin-tazobactam were started. Blood cultures showed no growth. Forty eight hours after admission he was still febrile and became tachycardic and tachypneic with an oxygen saturation of 88% on 3L, and was transferred to the ICU. He developed hypoxemic respiratory failure and was intubated. IV methylprednisolone 80 mg q 6 hours was started as well as antibiotics imipenem and gentamicin. Bronchoscopy revealed no ulcerations or vesicles. Bronchial wash culture was negative, and cytology was negative for pneumocystis, viral changes and malignancy. He went into septic shock with ARDS, mutiorgan failure and hemodialysis was initiated. During the third week he developed left sided pneumothorax. Bilateral chest tubes were placed. He was designated DNR and expired. An autopsy revealed HSV pneumonia with diffuse alveolar lung damage (ARDS), with immunohistochemical stains positive for HSV - 2. Discussion – The signs and symptoms of HSV pneumonia are nonspecific. In our case, the diagnosis was made only at autopsy. HSV - 1 has been isolated in critically ill patients due to spread from active lesions in the URT or the esophagus but not HSV-2. Our case was unusual as it represented an immunocompetent patient without evidence of underlying herpetic disease. The use of steroids may have exacerbated our patient’s herpetic infection. There have been theories to suggest reactivation of the virus during stress. Empiric antiviral treatment in critically ill patients is not recommended. However treatment with acyclovir may be justified when there is a high degree of suspicion - mostly in the critically ill with extensive mucosal herpes infection and diffuse chest infiltrates without another etiology.
Introduction: Sarcomas are 1% of malignant tumors that are of mesenchymal origin. They are characterized by infiltrative local growth. They are mainly hematogenous to the lungs, although lymphatic spread can occur. They can occur at any site though commonly found in extremities. With history of trauma and injury, the diagnosis could be challenging as the imaging studies could mask the underlying tumor. Presentation: A 43-year-old highly functioning female presents with acute alcohol intoxication after being found unconscious on the bathroom floor. She was very combative, agitated and in withdrawal. She was admitted to the ICU and started on ativan drip. The only significant physical finding was some discomfort on palpation of the medial aspect of the thigh near the groin area. After being stabilized and moved out of the ICU, she was being seen by physical therapy for mobilization. She did wince to palpation of the upper medial thigh without any evidence of a soft tissue mass. This was treated as trauma secondary to the fall during the intoxication. She was continued on physical therapy, pain medication and local heat therapy. Five weeks later, she came back with worsened pain, after which a radiograph and CT revealed a bulky mass involving the adductor-magnus compartment, suspicious of a organized hematoma, without evidence of calcification or ossification. An MRI was ordered, which showed a high T2 signal and a low T1 signal with areas of increased signal intensity, which was interpreted as sub-acute hematoma. A vascular Doppler was done to rule out aneurysm, which was normal. Again, conservative treatment was recommended. Patient continued to have increased pain. She was referred to radiology for an angiography, which revealed a vascularised mass with malignant characteristics. Clinical Course: Patient underwent a surgical intervention wherein a 16 X 8 X 4cm encapsulated mass was removed. Histopathology of the mass revealed, extremely pleomorphic, spindle shaped cells which was consistent with leiomyosarcoma. Post surgery the patient received radiation without any documented metastasis. Discussion: MRI remains the gold standard for evaluation of most soft tissue lesions, but findings usually overlap with benign tumors. MRI cannot histologically classify tumors. Also time dependent changes of the tumors, makes the differentiation even more difficult. MRI images of acute hematomas show low to intermediate signal intensity on T1 and low signal on T2. Intra-tumoral hemorrhage is a very rare finding, which can be found in both benign and malignant tumors. Surgery will give us the correct histological identification, thus the conclusion is intramuscular hematoma following trauma should be approached with high degree of clinical suspicion. MRI is not sensitive or specific enough to rule out malignancy. Any suspicion of high grade sarcoma should be resolved with a biopsy.
Atlantic Health - Morristown

Diagnostic Challenge in the Spleen!

Ashwini Arjuna,

Introduction: Splenic tuberculosis is an extremely rare, clinical presentation of TB. Most of the time it presents as pyrexia of unknown origin and as signs of malignancy such as weight loss, fatigue and without any prior exposure to tuberculosis. Case Presentation: 54yr old Indian male, presented with h/o ongoing fevers for the past three and half months usually starting around the evening and gradually progressing through the night which was associated with substantial weight loss. He did not have any cough, sputum production or abdominal pain. He denied any H/o TB or any history suggestive of exposure to HIV. Examination findings: He was a moderately built and nourished man, whose body temp was 37.4C and he had very mild splenomegaly. His only pertinent lab value was an elevated ESR of 73mm/hr. His chest X-ray did not reveal any abnormality. Abdominal USG showed the thickness of the spleen to be about 6.2cm with multiple hypo-echoic lesions. Ct scan of the abdomen revealed hypo dense cysts. It was initially thought to be a fungal lesion given the diffuse hypo dense lesions. CT guided splenic puncture, showed histo-pathologic report of granulomatous changes with large areas of caseation in the centre surrounded by large number of Langerhans giant cells and epithelial cells. AFB staining was positive. Course of the disease: Patient was finally diagnosed with isolated tuberculosis of the spleen with no other focus in the lung, GI tract or lymph nodes. He was started with anti TB therapy. Within a week of starting, his symptoms improved. A repeat USG of the abdomen showed marked improvement of the splenic lesions 8 months later. Discussion: Tuberculosis is a multisystem disease, primarily affecting the lungs. Extra-pulmonary manifestations especially in the spleen are very rare. It could either be primary or secondary infection of the TB in other organs. In our case, fever and weight loss were the only pertinent symptoms going in favour of Pyrexia of unknown origin or malignancy as the issue. CT could not reveal the type of lesions on the other hand, typical nodules of the splenic capsule are usually too small to be detected early on the CT. Treatment is standard for tuberculosis with rifampicin, INH, streptomycin and pyrazinamide and ethambutol. Within about two months patient’s symptoms improved and he was shifted to rifampicin and INH for the next four months. The patient’s symptoms did not recur in two years of follow up.
What is this rash?

Ashwini Arjuna,

Introduction: Chikungunya is an arthropod borne disease, transmitted to the humans by the CHIKV virus through the aedes mosquito bite. The illness is similar to dengue with an acute febrile phase, lasting for 2-5 days followed by a prolonged arthritic phase affecting the joints of the extremities. Case Description: This is a case of a 27year old Asian female, with history of recent travel to India for 5 weeks. 3 days prior to her return to the USA, she spiked a fever of 104F with right knee pain which was followed by maculo-papular rash all over the body including the palms and soles on day 3 of the fever. Initially the joint pains involved the larger joints of the body but later spread to the smaller joints . The pain and the fever subsided, only to recur after about a week with the same presentation of having a very high fever peak of 104F followed by a rash two days later. During the interim period, most of the lab works for dengue, measles, leptospirosis , lyme were negative. The presentation was also confusing with typhoid serology being equivocal, but the patient did not present with any GI symptoms nor was she exposed to any known cases of typhoid. Finally a Chikungunya IgM and IgGtiter were positive. This was sent basically because all of her initial blood work had been negative and since the fever did recur with ongoing joint pain and swelling. There were some cases of Chikungunya in India but this period was very unusual as the season did not warrant the breeding of the mosquito. Discussion: Chikungunya usually presents as a viral illness characterized by fever, arthralgia and rash. It usually follows a febrile period of 5 to 7 days associated with maculo-papular rash which rarely involves the palms and soles. The joint pains usually involve both the smaller as well as the bigger joints. Rare complications include debilitating arthropathy, myelo-radiculopathy, muco-cutaneous ulcers. The unusual presentation in this case was the bimodal febrile illness which was both followed by maculo-papular rash. The joint pains were limited to the smaller joints and involved the same joints both the times. The treatment usually is symptomatic with good hydration during the febrile illness, pain medications for the arthralgia. There is no anti-viral medication or vaccine against the disease. The other special feature was the presentation in early May when there were no cases reported in India and was unusual for the breeding period of the mosquito which is usually during late July, August and September.
INTRODUCTION: The ACGME common program requirements include the maintenance of a supportive educational environment which promotes patient safety and resident well-being. Recent research has suggested that resident mistreatment occurs at a higher rate than previously recognized, and organizations such as the AMA in collaboration with the LCME and AAMC have focused attention on identifying strategies to prevent and address medical student and resident mistreatment. Our study’s objective was to determine the prevalence of mistreatment among Internal Medicine residents training in New Jersey and to explore possible consequences of this behavior on clinical decision making and activity. METHODS: We reviewed literature on medical student and resident mistreatment and conducted a focus group of Internal Medicine residents to identify perceptions of mistreatment and issues and concerns associated with mistreatment. Based on our review, we identified four main types of mistreatment (verbal, physical, academic, and sexual), and designed a 20-item survey. We contacted the leadership of Internal Medicine residency programs in New Jersey, and received permission to email our survey to 298 residents who were enrolled in five of these programs. Most of the programs were based at mid-sized community-teaching hospitals. The survey was distributed on SurveyMonkey. RESULTS: There were 72 respondents (24.2% response rate) to our survey. 63.8% were male; 42.0% were PGY-1; 50.7% were US medical school graduates. Of the respondents, 29.2% stated that they had experienced mistreatment during their residency training. More US graduates reported mistreatment compared non-US graduates (37% vs. 15%, p=0.034); differences were not significant when comparing among training levels (p=0.62). The most common type of mistreatment was verbal (37.5%), and was most commonly committed by attending physicians and nurses. When asked if mistreatment had ever resulted in a change in how the resident cared for a patient, 22.2% of respondents answered “yes.” 18.5% of respondents felt that mistreatment had caused them to make a medical error. When asked if mistreatment had ever resulted in worsened performance on rounds, 63.0% answered “yes.” The majority of residents, 85.2%, felt that mistreatment did not improve their ability to learn. DISCUSSION: Resident mistreatment is common among Internal Medicine training programs in New Jersey. Physical, sexual, and academic abuse were less common compared to verbal abuse. Since mistreatment is perceived by residents to have negative effects on both clinical and academic performance, further study is warranted to determine actual effects on patient outcomes and board pass rates. Teaching hospitals and accreditation agencies should continue to monitor and address resident mistreatment behavior, particularly among attending physicians and nurses.
Relapsing Polychondritis as a manifestation of MDS

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History: 82 year old male with CAD, HTN who was diagnosed with Sweet’s Syndrome approximately 18 months ago. He improved on prednisone and colchicine, then maintained on colchicine. Within the next few months, bilateral mild intermittent earache, redness, and tearing of his eyes without any visual disturbances developed. Subsequent to that, nasal swelling, fatigue, weight loss, arthralgia, night sweats, intermittent fevers, and microcytic anemia developed. Bilateral skin thickening of the ears developed and became more swollen and painful. Impression & plan: Due to the inflammatory changes of the ear, right eye and a previous episode of nasal chondritis with abnormal laboratory findings, a diagnosis of relapsing polychondritis (RPC) was made. Over the course of his symptoms, he had two bone marrow biopsies. First revealed 60% hypercellular marrow with trilineage hematopoiesis. The megakaryocytes showed mild dysplastic changes, however no morphologic, immunophenotypic, and cytogenetic evidence for myelodysplastic syndrome was seen. Second revealed 80% markedly hypercellular bone marrow with trilineage dysplasia which was classified as a refractory cytopenia with multilineage dysplasia. Concurrent flow cytometry did not indicate immunophenotypic evidence for a hematolymphoid malignancy but findings were consistent with myelodysplastic syndrome (MDS). We suspected relapsing polychondritis(RPC) as an autoimmune paraneoplastic manifestation of MDS.

Discussion: The reported incidence of autoimmune disorders in MDS is about 10-13.6%. The pathogenesis of autoimmune disorders occurring as manifestation of MDS is currently unknown. Autoimmune manifestations which may come before or follow the diagnosis of MDS include acute systemic vasculitis and connective tissue disorders including SLE, Sjogren’s syndrome, polymyalgia rheumatica, and relapsing polychondritis. One series over a 14 year time period reported 0.6% of all MDS patients had RPC. This same series looked at incidence of MDS in RPC which showed about one-third of patient with PRC presented with malignancy or other autoimmune disease. Relapsing polychondritis disease has an estimated incidence of 3 per million population. It is known to be immune-mediated with an unclear etiology. However, genetic predisposition has been linked. It causes systemic inflammation of the cartilage; most commonly affecting the ear. However, it can involve other cartilages of the eyes, nose, large bronchial airways, joints, heart, and the skin. It affects genders equally, and known to be prevalent in Caucasians. The onset of age can occur between 40-60 years. There is no specific test to make diagnosis of RPC; however the use of modified Mcadams criteria can be used. Treatment of the disease depends on if there is an organ involvement. RPC associated with MDS has shown to decrease median survival compared to RPC occurring alone. Conclusion: Underlying Myelodysplastic syndrome should be suspected in immunological disorders.
Visual Recovery in an HIV Male with Cryptococcal Meningitis and a CD4 count >100?

Wendy Brandan, Gina Lacapra, MD FACP

50 year old Caucasian male with headaches for 3 weeks. On the day prior to admission developed a gait abnormality which resulted in a fall. He complained of neck stiffness, his behavior was different as per family. Exam revealed orientation to person only, tachycardia, normal temperature and normotensive, diffuse maculopapular rash, thrush, left dilated non reactive pupil, and left ptosis; and axillary adenopathy. Wbc, Hgb, CT head, CTA normal Platelets 115,000 VDRL positive Syphilis IgG positive HIV positive HIV viral load > 500,000, CD4 194 LP opening pressure:55 cmH20, lymphocytes 96%, VDRL negative Culture positive Cryptococcus Neoformans Patient admitted for Cryptococcus meningitis, started on Ceftriaxone, Vancomycin, and Amphotericin B Liposome. Fluconazole added and then both discontinued because of kidney injury. Fluconazole continued. Patient reported blurry vision, subsequent exams consistent with visual anosognosia. Acetazolamide started and ophthalmology noted multiple CN involvement without retinal disease. Neurosurgery consulted due to worsening neurological status. Pt deemed inappropriate candidate for drain because high risk for infection due to his immunocompromised state. Neurosyphilis suspected due to abnormal labs and dilated pupil. Penicillin was started. He had 5 subsequent taps-four had elevated pressures with final tap pressure of 16 cm. These taps improved his visual perception and impaired mental status. Discussion: Cryptococcus has four subtypes and three varieties. A fungal disease with a polysaccharide capsule that escapes the innate immune system via the lungs. Meningoencephalitis is the most common presentation. It is found worldwide. Infections with Cryptococcus gattii are more common among immunocompetent hosts. Aid’s patients have a more rapid course. Most cases are insidious. Signs and symptoms are fever/photophobia/stiff neck/headache and neurological signs including lethargy/behavior changes/seizures/cranial nerve palsies. Visual complaints are suggestive of a poor prognosis. Once vision loss occurs, it is often irreversible. Complications include micro-abscesses/hydrocephalus/arteritis/thrombosis and infarct. The two major causes of death are proliferation of fungi in the brain and immune reconstitution inflammatory response. Most cases are seen in patients with CD4 counts of <100. CD4 is the main mortality predictor. Other predictors of mortality are: low CSF white count, altered mental status, CSF ag titer >1:1024, CSF elevated pressures without expanding ventricular size. Treatment in HIV-infected CNS disease consists of induction with amphotericin B supplemented by fluconosine, consolidation with fluconazole/itraconazole for 8 weeks or until CSF is sterile, and maintenance with HAART and antifungals to reduce risk of opportunistic infections. Elevated ICP managed by serial lumbar taps. In patients with >40 cm opening pressures, lumbar drains are recommended. If this fails, VP shunt is indicated.
We present a 25 year-old Caucasian female with a 17-year history of eating disorder and recurrent episodes of hypotension and generalized weakness thought to be secondary to decreased oral intake/dehydration. She was well until 2 days prior to admission when she became dizzy after multiple episodes of diarrhea secondary to colonoscopy preparation. In the emergency room, the patient was hypotensive (SBP 70's) and bradycardic (HR40's). Physical exam was remarkable for somnolence and dry mucous membranes. Admission blood tests were unremarkable and EKG showed sinus bradycardia at 47bpm with QTc 472. She was admitted for dehydration secondary to poor oral intake and diarrhea. On further investigation of her medical records, she had sinus bradycardia previously with a normal ejection fraction. This was felt to be secondary to her eating disorder and a pacemaker was not indicated. She was started on intravenous fluids for several days with minimal improvement in blood pressure. Another consideration for her hypotension was the possibility of hypocortisolism. Her AM cortisol level was markedly low (0.9 ug/dL Normal: 10-20). The patient was started on dexamethasone. Her standard ACTH stimulation test revealed an adequate adrenal response, which demonstrated a pituitary/hypothalamic dysfunction as the likely etiology for her hypocortisolism. Other pituitary hormones levels i.e. LH, IGFBP-1, TSH, Prolactin were within normal limits. MRI of the brain showed no evidence of pituitary/hypothalamic abnormalities. She was switched to hydrocortisone with some improvement in blood pressure. Hydrocortisone was tapered to a maintenance dose and midodrine was added for BP augmentation. She was discharged home with an endocrine follow up after normalization of blood pressure. Anorexia nervosa is associated with several medical complications directly attributed to caloric restriction and weight loss. Traditionally, Anorexia Nervosa leads to decrease in LH, FSH, GnRH, IGF-1, ADH, T3, T4 and increase levels of growth hormone and cortisol are seen. In contrast, our patient was found to be adrenally insufficient. Although unlikely in Anorexia Nervosa, adrenal insufficiency should be considered in patients who presents with severe hypotension. Since symptoms can be vague, evaluation is needed to confirm the diagnosis and to determine the type of adrenal insufficiency. Dexamethasone, which is not measured in ACTH stimulation test, is the initial glucocorticoid of choice prior to testing. The test involves administration of synthetic ACTH IM or IV with blood cortisol levels measured at 0, 30 and 60min. In healthy individuals the response is an increase in cortisol level >20 g/dL or an increment of >10 g/dL over baseline. If the stimulation test only raises the cortisol level slightly, it would support the diagnosis of primary adrenal insufficiency. If serum cortisol level doubles or triples from baseline, secondary adrenal insufficiency is diagnosed. After conformation, hydrocortisone is the glucocorticoid of choice.
Atassic Health - Overlook

Profound DIC in unclear etiology

Nicky Desai, Gina Lacapra, MD

77 y/o AA female with CAD, DM on ASA/Plavix admitted three weeks ago to a nearby hospital for rectal bleeding. Sigmoidoscopy was normal and was told not to resume her ASA/Plavix upon discharge. She was found with asymptomatic hypotension and tachycardia at the nursing home and was transferred to our hospital. EKG demonstrated atrial fibrillation and CXR suggestive of PNA. She was started on metoprolol/vancomycin. Following day, noted to have elevated PT/PTT/INR, thrombocytopenia, low fibrinogen, elevated FDP/D-dimer. Cryoprecipitate was given and Cefepime was added for concerns over sepsis. Abdomen ultrasound suggested intra/extrahepatic, CBD and pancreatic duct dilatation. CT chest/abdomen/pelvis demonstrated thrombus in LV, thoracic/abdominal aorta. Dopplers were done revealing clots in multiple arteries/veins in upper/lower extremities. Hematologist believed underlying malignancy as etiology of DIC. She was started on argatroban. Suggested EUS to rule out pancreatic carcinoma/cholangiocarcoma but patient refused. Vascular was consulted due to large clot burden but no further intervention suggested. Discussion DIC occurs in conditions like sepsis, trauma, tissue destruction and obstetrical complications. Also associated with malignancies such as lung, prostate, pancreas and etc. Other etiologies include liver disease, snake bite, shock, heat stroke, vasculitis and an aortic aneurysm. In two large series of 346 and 118 patients with DIC, the following etiologies were found: Generalized infection (26 and 40%) Malignancy (24 and 7%) Surgery and trauma (19 and 24%) liver disease (8%) and miscellaneous (23%). The pathophysiology of DIC is defined as dysregulated coagulation and fibrinolysis resulting in clotting with bleeding. Activation of coagulation cascade yields thrombin converting fibrinogen to fibrin. Thrombin cleaves fibrinogen, leaving behind multiple fibrin clots in the circulation. These clots trap platelets leading to microvascular/macrovascular thrombosis resulting in ischemia, impaired organ perfusion and end-organ damage. Also thrombocytopenia and clotting factors are consumed in the development of multiple clots, which accounts for bleeding. The fibrinolytic system functions to break down fibrinogen and fibrin resulting in FDP. It carries an anticoagulant property contributing to hemorrhage. Patients are often acutely ill and presents with hemorrhage, petechiae, ecchymoses, renal failure and gangrene. Also manifest as hepatic/respiratory dysfunction, shock and CNS involvement. Diagnosis depends on: -Thrombocytopenia -Prolongation of PT/aPTT -Low fibrinogen -Elevated FDPs/ D-dimer The only effective treatment is the reversal of the underlying cause. Anticoagulants are given very rarely when thrombus formation is likely to lead to imminent death such as in coronary artery/cerebrovascular thrombosis. Transfuse platelets if counts are <5,000-10,000 or if massive hemorrhage is occurring. FFP may be administered to replenish coagulation factors. Prognosis varies depending on the underlying disorder and the extent of the intravascular thrombosis. Between 10%-50% of patients will die. DIC with sepsis has a significantly higher rate of death than with trauma.
MAY-THURNER SYNDROME WITH PULMONARY EMBOLISM AS THE FIRST PRESENTATION RATHER THAN DEEP VEIN THROMBOSIS (DVT)

Adebayo Fasanya, GINA LACAPRA, MD

Case Presentation: 19 year old Obese Caucasian female who was in her usual state of health until two days prior to presentation when she noted shortness of breath and pleuritic pain under her right breast. She denied recent leg edema/pain. She has no history of thromboembolic disease. CT chest with contrast showed extensive segmental and subsegmental bilateral pulmonary emboli. also right heart strain of echocardiogram. The patient was started on heparin. The patient underwent pulmonary artery thrombolysis with marked improvement in the pulmonary artery pressures. Hypercoagulability workups were negative. Lower extremity dopplers revealed acute extensive Left DVT. The patient underwent a venogram which showed stenosis of the left common iliac vein with well-established collaterals and a stent was deployed. Thrombolysis of the left lower extremity DVT, and placement of an IVC filter was done at the same time. Discussion: MTS is a rare disease that causes DVT in young females between the ages of 20-40. It is a mechanical obstruction of the left common iliac vein by the right common iliac artery causing stasis, rather than a primary hypercoagulable state. Although this condition is found approximately 22% of cadavers, it is implicated in less than 5% of lower extremity venous disorders and greater than 70% compression is needed to cause DVT. Most patients with MTS presents with sudden onset of left leg edema. Patients might not notice as it sometimes resolves with time. Many cases are recurrent with a past workup negative for other etiologies of DVT or PE. Very few cases however present as PE rather than DVT. The gold standard technique for diagnosing MTS is the femoral stick venogram. Area of compression with documentation of collateral presence confirms the diagnosis. Femoral stick venogram, although invasive, offers many advantages. This is the only technique that allows other procedures to be done at the same time such as thrombolysis, balloon angioplasty, stenting and even placing an IVC filter. Other methods of diagnosis include CT with contrast and intravascular ultrasound. Guideline for the treatment of MTS is not available in the literature due to the low incidence of the syndrome. Anticoagulation therapy alone is not enough to prevent recurrence. Short term studies and reviews recommended that a stent be placed to relieve the underlying compression of the venous system and to continue anticoagulation for 6-12 months postoperatively. Endoluminal stenting is associated with low morbidity and high patency rates. Long term studies are not presently available. Recommendation: It is important to include MTS as one of the mainstream differential diagnoses of DVT but also PE in young women. Especially, if the etiology is not obvious. Aggressive evaluation of PE and lower extremity DVT for MTS could decrease mortality and morbidity significantly.
Atlantic Health - Overlook

A Rare Case of Extrapulmonary Legionella presenting as Rhabdomyolysis

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CASE: 59 y/o Caucasian, obese, male with past medical history of COPD, who was in his usual state of health, until one week prior to admission when he developed fever, nasal congestion, and a headache. His fevers and chills persisted with a Tmax of 102.5°F which he took OTC ibuprofen to control as needed. He became lightheaded after a bowel movement and lied supine on the floor without loss of consciousness or trauma. Patient was tachycardic, tachypneic and hypoxic. Physical examination showed diminished breath sounds in the lower lobe, no crackles or wheezes. There was resonance on percussion and tactile fremitus was equal and bilateral within normal limits. Positive labs include WBC 13.51, PMNs 88.5%, Bicarb 16, BUN 27, Creatinine 2.4, AST 123. Total CK peaked at 22412. Chest x-ray showed left perihilar infiltrate with no pleural effusions or pneumothorax. EKG showed sinus tachycardia. He was hydrated and started on dual therapy with Levofloxacin 750mg IV Q24 and Azithromycin 500mg IV because his urine Legionella antigen was positive. Hospital day 5, he showed improving clinical signs, CK in 6000s. On Hospital day 8, Azithromycin was d/c and Levofloxacin was kept as monotherapy. He was discharged on Hospital day 10 with a CK of 272, continued on Levaquin 750 mg PO for 4 days. DISCUSSION: Legionnaires disease is a pneumonia with multisystemic manifestations caused by a fastidious gram-negative bacillus called Legionella pneumophila. Transmission occurs by aerosolization or aspiration of contaminated water with Legionella organisms. An estimated 8000 to 18000 cases of Legionnaires disease are reported each year in the United States. In adults, it accounts for 2-15% of all cases of community-acquired pneumonia requiring hospitalization. It is the second most common cause of severe pneumonia requiring ICU admission. Extrapulmonary legionellosis is rare and the most common site of extrapulmonary infection in adults is the heart. This was an interesting case of a 59-year old male who presented with extrapulmonary Legionaire’s disease with rhabdomyolysis. Treatment of Legionnaire’s disease is primarily with Levofloxacin or Azithromycin. ACCP studies show that there is no difference in efficacy between Levofloxacin or Azithromycin. Few case reports and laboratory studies have suggested possible benefit with combination therapy, however this approach have not been validated in observational studies. RECOMMENDATION: There are no official guidelines for management of extrapulmonary Legionnaire’s disease. However, literature recommends quinolone plus azithromycin or rifampin for severely ill patients with extrapulmonary legionellosis.
Using a Patient education Initiative to Improve COPD Outcomes

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Introduction: COPD exacerbations are a leading cause of hospital re-admissions. Each hospitalization is associated with high economic cost and progressive decline in patients’ health status. According to a recent study of about 12 million Medicare beneficiaries with COPD, the hospital readmission rate within 30 days was nearly 20%. Existing data suggests that at least three-quarters of these readmissions are preventable. Previous studies have demonstrated that simple and effective steps like patient education and adequate outpatient follow up can significantly alter COPD re-admission rates. The purpose of our study was to improve the quality of COPD management with standardized education and decrease re-admissions in this patient population.

Methods: The control group, identified using the University Healthsystem Consortium database, included patients admitted to Overlook Medical Center with a diagnosis of acute COPD exacerbation from February to June 2011 and the study group included similar patients studied prospectively from April to July 2012. The study group received standardized education by trained respiratory therapists based on a written non-branded handout offered by Glaxo-SmithKline. Areas covered in the education included symptom understanding, correct inhaler use and medication understanding/compliance. 30 day readmission data were recorded for patients in both groups as well as baseline demographic and clinical factors. Study group patients completed a pre and post education Likert-scale type survey to assess level of disease awareness and self-management. The two groups were compared in terms of 30-day readmission rate using the Chi-square test. A multivariate logistic regression model was used to predict adjusted readmission rate by group. Study group survey results were compared pre and post education using the McNemar test on paired proportions.

Results: A total of 105 patients were included; 64 in the control group and 41 in the study group. Average age was 74 years (SD = 10.5) with 57% females. Median length of stay was 6 days, range: 1-19 days. Overall 30-day readmission rate was 23%; 28% in the control group vs. 15% in the study group (p = 0.11 by Chi-square).

Results from logistic regression showed a significantly lower 30-day readmission rate in study group compared to control group (OR = 0.29, p = 0.036, 95% CI 0.9-0.92). Higher likelihood of 30-day readmission was marginally related to short acting anticholinergics at admission (OR = 2.98, p = 0.051, 95% CI 1.00-8.92). No significant difference in mortality was noted. Control group patient survey results showed significant improvement in disease awareness post intervention (median score post-intervention 17 vs. 13.5 pre-intervention; p <0.001, by the Wilcoxon Signed Ranks test).

Conclusion: We conclude that standardized patient education during hospitalization reduced the 30 day readmission rate for COPD patients and significantly improved disease awareness.
A qualitative study for improving effectiveness of the evaluation of patients with syncope

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Introduction: Syncope is a common clinical problem defined as transient loss of consciousness with rapid onset, short duration, and spontaneous complete recovery. Due to the facts that there is a wide range of conditions that could cause syncope and there are no strict published guidelines, the evaluation of patients with syncope is usually inefficient and expensive. We aim to improve the efficiency of evaluation by focusing on processes with high effect-cost ratio, decreasing un-necessary test orders, and shortening length of hospital stay (LOS) in patients with low risk. Methods: Multiple-step interventions were applied to improve the evaluation of patients with syncope who were seen by residents at our hospital. 1. A syncope protocol was distributed to ED to guide the triage of patients with syncope into 3 groups: discharge, admission, and observation. 2. Residents were given lectures for history taking (including medication review) and physical exam in patients with syncope. 3. For patients with syncope who are admitted to hospital or under observation, a syncope observation/admission order set was trialed by the residents to guide the ordering of efficient tests. 4. Residents coordinated with the cardiovascular lab to expedite the echocardiogram. Before and after those interventions, 9-10 charts were reviewed in each group-the admission group and the observation group. Data were collected on orthostatic blood pressure measurements, document of medication review, orders such as telemetry, echocardiogram, CT of head, carotid Doppler, cardiology or neurology consults, and LOS were compared before and after interventions in admission and/or observation groups. Results: Orthostatic blood pressure measurement and medication review were performed in 100% of patients in both the admission group and the observation group after interventions compared with less than 20% before interventions. The echocardiogram time (from time of order to time of official result report) was 24-52 hours in the observation group and 14-72 hours in the admission group. Orders of consults and carotid Doppler decreased more than 50% after interventions in both groups. There were no change in the number of tests such as cardiac enzymes and CT of head which were usually done at the emergency department. In the observation group, LOS decreased from 1-4 days (most 2 days) before interventions to 11-48 hours (60% < 24 hours) after interventions. Conclusion: Repeating resident education and use of syncope protocol help to decrease cost by decreasing un-necessary orders and LOS, thus to improve the efficiency of the syncope evaluation.
A Rare Case of Effusion Based Lymphoma in a HIV/HHV-8 Negative Patient

Trishala Meghal, Dr. Michael Wax MD, Dr. Gina Lacapra MD, Dr. Bhagavatula MD

An 81 year old male with a past medical history of end stage renal disease on hemodialysis presented with shortness of breath that started two months ago and got progressively worse. Review of systems was significant for loss of appetite. On examination, he was tachypneic. His lung exam revealed decreased breath sounds and dullness to percussion in the lower right half of the lung fields. Chest x-ray and cat scan revealed a large right pleural effusion No enlarged lymph nodes were felt on exam and there was only one precardinal lymph node measuring 1.2 cm in size on chest cat scan. Cat scan of neck and abdomen was negative for masses or enlarged lymph nodes. A diagnostic and therapeutic thoracentesis was performed, which revealed an exudative pleural effusion with a pleural fluid to serum protein ratio of 0.55 and a pleural fluid LDH of 4971. Cytology revealed CD20 and CD45 positive diffuse large B cell lymphoma. The neoplastic cells were negative for Human Herpes virus-8(HHV-8), EBV, CD138 and CD10. The patient was HIV negative. He was diagnosed with an effusion based large B cell lymphoma. Lymphoma usually presents as weight loss, fevers, loss of appetite along with enlarged nodes on exam or imaging. Our patient had lymphoma cells in his pleural fluid without any significant lymphadenopathy or masses which is classified as primary effusion lymphoma.(PEL)( WHO classification). These patients are HIV positive and are typically positive for HHV-8. The malignant cells of PEL are monoclonal B cells that express cell surface CD 38 and contain genomic material from HHV-8 and in many cases EBV. The cells correspond to a stage of B-cell development intermediate between that of immunoblasts and plasma cells. Over 90% of cases demonstrate expression of CD45. Plasma cell related markers such as CD30, CD38, CD71, CD138 are usually present. Other B cell markers such as CD19, CD20, CD79 and T cell associated antigens are typically negative. Recently there is another entity called PEL like lymphoma or effusion based lymphoma in patients who are HHV-8 negative and are generally HIV negative. I have found only 45 other cases of patients that fit these criteria. These patients are generally older and often have been found to have an underlying medical condition leading to fluid overload. Its cytomorphology resembles PEL with some distinct differences. It is usually a germinal center B cells unlike PEL in which the cells are typically activated B-cell type. The lymphoma cells express CD20 in 70% of cases. The therapy for both PEL and effusion based lymphoma includes chemotherapy and recurrent paracentesis. The prognosis for effusion based lymphoma is better than PEL and there have been case reports of remission with steroids or recurrent paracentesis.
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A Rare Cause of Joint Disease in Adults

Michael Mobasser,

HISTORY  This is a 40 year-old Haitian female who moved to the USA in 1996, with no significant PMH who was well up until 5 months prior to admission after returning from Haiti. She developed severe polyarthritis and fever. It subsided then recurred few months later when she also developed weakness and fatigue. These symptoms started to worsen and she developed myalgias. It got to the point where she was unable to work or eat well, she noticed that she lost 15-20 lbs, and she presented to our hospital. PHYSICAL EXAM  Her clinical exam revealed a chronically ill-appearing woman lying in bed in pain. She had vitiligo in the periorbital regions, periauricular regions, and her upper back; but no rashes or nodules. Musculoskeletal exam showed evidence of discomfort with attempted range of motion of both shoulders. Muscle strength 4/5 proximally in the bilateral deltoids, biceps, and quadriceps and distally in the flexor digitorum profundus, tibialis anterior and extensor hallucis longus. Tenderness and swelling of both wrists, left 5th proximal interphalangeal joint, right knee, left ankle, and the right first, second and third metatarsophalangeal joints. All other joints were non-tender and non-swollen. Cranial Nerves are intact. The bilateral patellar, biceps, and brachioradialis tendons are 1-2+ symmetrically and the plantar response is downward bilaterally. DIAGNOSTICS  White cell count 17,500 with myelocytes, metamyelocytes, rouleaux formation, teardrop cells and nucleated red blood cells. Hemoglobin 8.6, Albumin 1.4, ALT 50, LDH 410, CPK 19, alkaline phosphate 184, Erythrocyte sedimentation rate 103, c-reactive protein >190 Ferritin 23,996 An extensive infectious and immunologic serum panel including antibodies to: Anticardiolipin, Anti-CCP, ANA, Smith, dsDNA, SSA/SSB, Histoplasma, Blastomyces, Coccidioides, RNP, Rheumatoid Factor were negative. Other Serologic markers: HIV negative Hepatitis A reactive Hep B core IgG Reactive Histoplasma Ag urine negative Blood parasites negative Total, C4, and C3 complement= Normal CT scan of chest, abdomen, and pelvis were done to rule out any infectious process and results were small bilateral pleural effusions and bibasilar atelectasis but no significant infiltrates or adenopathy. There is evidence of fatty liver but no splenomegaly, retroperitoneal masses, adenopathy or ascites. Bone marrow biopsy showed myeloproliferative disease. EMG was consistent with polymyositis MRI of the bilateral lower extremity showed myositis. The impression was that she most likely had Adult-onset Still’s Disease based on the clinical picture of fever, diffuse joint pain, very high serum ferritin levels and the exclusion of other infectious and inflammatory etiologies. She was started on high dose IV steroids with 3 days of pulse dosing, then an oral taper. She was also given an intravenous infusion of Rituximab, with the plan of a repeat dose in 1 month. Her fevers and muscle pain improved.
Atlantic Health - Overlook

Quality Improvement Project on the efficiency of cardiac enzymes testing on patients in ICU/CCU of Overlook Medical Center

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Introduction: Cardiac Troponins (cTn) are highly sensitive and specific for the diagnosis of acute myocardial infarction (AMI). All high-sensitivity cardiac troponins (hs-cTn) are substantially more sensitive and specific than creatine kinase-myocardial band (CKMB) assays. Current ACCF/AHA practice guideline recommends performing cTn as the preferred test for diagnosing AMI, CKMB should not be used unless the hospital does not have troponin assay available. Creatine kinase (CK) separately is not recommended anymore for diagnosing AMI. Usual practice at Overlook Medical Center (OMC) consists of ordering serial hs-cTnl together with CK (with reflex CKMB) for diagnosing AMI. This practice of ordering CK provides no benefit while burdening laboratory staff and increasing cost for hospital and patients. The aim of this quality improvement study is to examine the impact of resident and nursing staff education on reducing the rate of CK testing done simultaneously with hs-cTnI measurement for diagnosing AMI in ICU/CCU patients. Methods: The database was obtained from the OMC Laboratory to determine the numbers and rates of simultaneous testing of hs-cTnI and CK (with reflex CK-MB) in the ICU/CCU of OMC in one month period. We excluded CK assays that were done alone without hs-TnI as this testing pattern was usually ordered for assessment of skeletal muscle pathology. An educational intervention designed to decrease use of unnecessary tests was implemented in the subsequent month in the form of resident and nursing staff education (short lecture and fliers distributed throughout our ICU/CCU). Numbers and rates of testing during the month of educational campaign were collected from the Laboratory. The rates of simultaneous troponin and CK/CKMB testing were compared pre and post intervention using the chi-square test. Results: Cardiac enzyme testing data were gathered for 178 patients [98 pre-intervention (55%) and 80 post-intervention (45%)]. The total number of tests done pre-intervention was 181, out of which 48 were troponin only tests (27%), and 133 simultaneous troponin and CK/CKMB (73%). The total number of tests done post intervention was 188, out of which 108 were troponin only tests (57%), and 80 simultaneous troponin and CK/CKMB (43%). The rate of simultaneous troponin and CK/CKMB testing decreased significantly post intervention (73% pre-intervention vs. 43% post-intervention; p <0.001). Conclusion: Simple educational interventions of resident and nursing staff were effective in reducing unnecessary CK/CKMB testing by more than 40% in ICU/CCU settings. Based on our success we now intend to generalize this throughout the hospital.
A 30-year-old Caucasian male with history of hypertension presented with four days of right upper quadrant and epigastric pain worsening with food intake. He had occasional nausea and vomiting, but denied fever, chills, diarrhea, melena, jaundice, burning pain in throat or chest, or recent surgeries. Vitals were within normal limits. The abdomen was soft with normal bowel sounds; but tender in the epigastric region with positive Murphy’s sign. No rebound tenderness, guarding, organomegaly, or hernias. The rest of the exam was unremarkable. Laboratory testing was within normal limits. An abdominal ultrasound showed normal appearing gallbladder. Hepatobiliary scintigraphy with cholecystokinin revealed a borderline low ejection fraction of 38%. He was admitted, kept npo, and given intravenous hydration. The gastroenterologist diagnosed biliary dyskinesia and recommended cholecystectomy. Surgery was consulted and advised outpatient surgery since the ejection fraction was above 35%. The patient was discharged on ursodeoxycholic acid with surgical followup. Sphincter of Oddi dysfunction (SOD) is the term used to describe a syndrome of functional or structural abnormalities involving the common bile duct, pancreatic duct, or any sphincter involved with these ducts. Various other names synonymous with SOD are biliary dyskinesia, acalculous biliary disease, postcholecystectomy syndrome, and papillary stenosis. Dyskinesia and/or stenosis lead to outflow obstruction at the ampulla of vater. Dyskinesia is abnormal tonic or phasic sphincter motor activity revealed on manometry. The structural abnormality involves stenosis of one of the ducts and/or sphincters. It results from chronic inflammation and fibrosis from recurrent pancreatitis, choledocholithiasis, trauma, or tumors. Patients must meet all Rome III criteria: • Epigastric and/or right upper quadrant pain • Episodes lasting >8805; 30 minutes • Pain not alleviated by bowel movements, postural changes, antacids, H2 blockers, Proton pump inhibitors, or antispasmodic agents. Then classify SOD into three entities: functional gallbladder disorder, biliary SOD, or pancreatic SOD: a) Functional gallbladder disorder patient has intact gallbladder, normal liver and pancreatic enzymes, normal conjugated bilirubin, but decreased gallbladder ejection fraction (<40%). Treatment: cholecystectomy b) Biliary SOD patients s/p cholecystectomy with biliary type pain: • Type 1: liver enzyme elevation and common bile duct dilation > 8mm on ultrasound. Treatment: sphincterotomy • Type 2: either liver enzyme elevation or common bile duct dilation > 8mm on ultrasound. Treatment: ERCP/manometry to decide medical (calcium channel blockers, nitrates, ursodeoxycholic acid) vs sphincterotomy • Type 3: no liver enzyme elevation or common bile duct abnormality on ultrasound. Treatment: medical management c) Pancreatic SOD patients s/p cholecystectomy with pancreatic type pain and/or recurrent pancreatitis: • Type 1: serum amylase/lipase elevation and a dilated pancreatic duct on ultrasound • Type 2: serum amylase/lipase elevation or dilated pancreatic duct on ultrasound • Type 3: no amylase/lipase elevation or dilated pancreatic duct. Treatment for all three types: ERCP/manometry to decide medical vs sphincterotomy.
WHAT IS THE BEST APPROACH TO ACCURATE MEDICATION RECONCILIATION

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Introduction: Adverse events caused by medication errors have a significant impact on patient safety and pose financial burden on patients and the health care system. In 2005, it became National Patient Safety Goal No.8. The systemic implementation of this is proving to be extraordinarily difficult.

Objectives: Utilize a residency driven quality improvement process to decrease errors in medication reconciliation upon admission to an acute care hospital by 1) Quantitative investigation of discrepancies between the three sources for medication reconciliation on the chart upon admission (the ED database, history and physical, and the medication reconciliation form completed by nursing) 2) Use of a tracer methodology to investigate how the medication list was obtained for these three sources and if it was verified 3) Investigation and development of a strategy to improve the accuracy of the medication reconciliation process.

Results: 25 randomly selected inpatient cases were investigated. 92% (23/25) of the patients had some discrepancy between the sources of information in the chart regarding a patient’s medications. In 18 of these 23 patients, all three medication lists were different. Most of these discrepancies were different medications documented; although incomplete or incorrectly documented doses or frequencies were not unusual. We joined an interdisciplinary team to further investigate and help improve medication reconciliation from the various hospitals in our system and were comprised of physicians, nurses, administration, pharmacy, and information technology. The tracer revealed that most medication information was obtained either from the patient, family, or sometimes the actual pill bottles in the ED. The admitting floor nurse verified this information with the patient most of the time if the family went home. Doctors verified this information with the doctor’s office medication list, a previous discharge summary from the same hospital, or the patient’s pharmacy if time allowed.

Conclusions: There are frequent errors in medication reconciliation which lead to incorrect information on patient illnesses and incorrect admitting orders. The problem is partly due to multiple sources without a unified system and variability in the extent to which an individual health care provider will track down the appropriate information. Based on these findings, our recommendation was to have a single source in the medical chart that can be verified by each person taking care of that patient. We also want to add verification by a pharmacist. Currently a pilot program is being put together after a computer upgrade to allow for medication reconciliation to be initiated in the ED, followed by nursing verification, pharmacy verification, and physician verification all in the same program. This program will also allow doctors to write admitting orders right from this medication list.
Moyamoya disease is a progressive, occlusive disease of the cerebral vasculature with particular involvement of the circle of Willis and the arteries that feed it. Collateral circulation develops around the blocked vessels to compensate for the blockage, but the collateral vessels are small, weak, and prone to hemorrhage and thrombosis. Moyamoya-like syndromes, usually autosomal recessive disorders, have primarily been described in children. We present an interesting case of a middle-aged man with Moyamoya-like syndrome as a complication of previously undiagnosed polycythemia vera. A 52 year old male with history of hypertension presented to the Emergency Department complaining of slurred speech and numbness, tingling and weakness in his left hand for 2 days. He was unable to extend his left hand. He had a similar episode 2 weeks prior that lasted for an hour and resolved on its own. Review of the systems was unremarkable. Physical examination revealed transient weakness in fourth and fifth digits of his left hand. He was alert and oriented with no other focal neurologic deficits. Cranial nerve examination was normal. There were no carotid bruits. Laboratory studies revealed an elevated hemoglobin of 18 g/dl, elevated RBC count 8.20X10^6 /dl and hematocrit of 58.7%. Non-contrast computed tomography (CT) of the head showed mild chronic small vessel disease with a cortical focus of non-specific low attenuation in the right frontal lobe. No embolic source was identified on 2D echocardiogram. CT brain perfusion showed an area of infarction within the right frontal lobe with no significant penumbra. CT angiography of the neck showed occlusion of right internal carotid artery (ICA). Magnetic resonance angiogram of the brain revealed “string” sign in the supraclinoid portion of the right ICA. Further workup for the polycythemia revealed elevated leucocyte alkaline phosphatase levels, increased folate and vitamin B12 with low erythropoietin levels. No other cause for the polycythemia was identified. Magnetic resonance imaging of the abdomen revealed splenomegaly. Clinical picture was diagnostic for polycythemia vera by PVSG criteria. A transfemoral angiogram confirmed the Moyamoya-like syndrome with stenosis of the cervical segment of the right internal carotid artery at the C2-3 interspace distal to the carotid bulb with right middle cerebral artery filling via right posterior cerebral artery. He was scheduled for encephalo-duro-arterio-synangiosis (EDAS) procedure as an outpatient. This procedure uses a branch of the superficial temporal artery which is laid directly on the surface of the brain without any anastomosis. This will eventually grow new arteries in to the brain and provide blood flow. He was discharged on aspirin and Plavix for secondary stroke prevention.
Energy drink-induced Ventricular Arrhythmias, QT Prolongation and Syncope

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QT interval represents the electrical depolarization and repolarization of both ventricles. A prolonged QT interval can be associated with ventricular tachyarrhythmia, such as torsade de pointes, and a possible cause of sudden death. A 29 year old female with a past history of asthma, bipolar disorder and depression, on citalopram, carbamazepine, and buspirone presented to the emergency department after a syncopal episode, which occurred while performing household chores. She reported drinking one bottle of 5-hour Energy Drink® (1.93oz), and two Monster® energy drinks (16oz) earlier that day. She also reported a previous syncopal episode one year ago, after consuming similar quantities of energy drinks, however declined medical treatment at that time. Twenty minutes after consuming the second bottle of Monster energy drink, she had palpitations, diaphoresis, and nausea, followed by syncope. Her significant other reported the patient was unconscious for three to five minutes without seizure activity or postictal state. On admission, blood pressure was 113/82 and heart rate was 98 bpm. Cardiopulmonary examination revealed regular rate and rhythm, without murmurs, rubs or gallops, and expiratory wheezing. Electrocardiogram revealed an accelerated idioventricular rhythm (AIVR) at a rate of 98 bpm with atroventricular dissociation and QT prolongation (QTc= 670). Echocardiogram revealed normal left ventricular function with mild mitral regurgitation. Serum magnesium was 1.6 mg/dL and serum potassium was 3.9 mg/dL. After admission, AIVR resolved spontaneously, while QT prolongation persisted >550 msec, despite withholding all medications. Magnesium and potassium were replaced to maintain levels >2 mg/dL and >4 mg/dL respectively. She remained asymptomatic. Her QTc improved, but remained mildly prolonged at 513 msec, raising the question of possible underlying congenital QT prolongation syndrome (LQTS-1). This case illustrates the additive effect of energy drinks with the use of citalopram on the QT segment. Several observational studies have noted that energy drinks alone prolong the QT interval by 10 msec, and increase the systolic blood pressure and heart rate by 8%. There have also been reports that excessive consumption of caffeinated energy drinks may also trigger life-threatening QT-prolongation in patients with LQTS-1. Several cases of cardiac arrest, seizures, psychiatric effects, orthostatic intolerance, restlessness, and irritability have been reported.
A patent foramen ovale (PFO) is a less common, often benign communication between the right and left atria that occurs in approximately 25% of the general population. Occasionally, this patency can allow for a paradoxical embolism, where a venous thrombus can make its way into the arterial circulation with resulting embolization in the branches of the cerebral arterial circulation. We report a case of a patient who experienced 2 stereotyped ischemic neurological events, the second of which resulted in an ischemic stroke secondary to a patent foramen ovale with an atrial septal aneurysm (ASA). A 31 year-old male with no past medical history presented to the emergency department complaining of numbness to his left wrist and hand for 14 hrs. His symptoms occurred acutely during a valsalva maneuver. He also complained of transient drooling from the left side of the mouth and a headache, 5-6/10 that worsened with rapid shifting of his head to either side. He experienced one similar self-limiting episode in the past. Vital signs were within normal limits. Physical examination was unremarkable. Neurological examination revealed decreased sensation on the left hand. Admission laboratory testing was normal. Computed tomography of the head without contrast was unremarkable. Magnetic resonance imaging revealed an acute infarction involving less than one third of the right middle cerebral artery territory. Transesophageal echocardiography revealed a small PFO with left to right shunt with an ASA. He was started on Aspirin 325mg daily and was scheduled for an elective percutaneous closure of the PFO. Current literature reports the established relationship between PFO or PFO with ASA and paradoxical embolism resulting in systemic embolism such as transient ischemic attack or ischemic stroke. Currently controversy exists in management of such patients. The current treatment options include medical management via anticoagulation, antiplatelet therapy, or a combination of both versus closure of the PFO. As per the Randomized Evaluation of Recurrent Stroke Comparing PFO Closure to Established Current Standard of Care Treatment (RESPECT) trial, the final conclusion determined no significant benefit with closure of a PFO as compared to medical therapy in an intention to treat analysis. However, closure of the PFO was associated with a decreased rate of recurrence and size of ischemic strokes. Secondary to these findings, we suggest a larger study with a lengthier follow up period as this could possibly reveal a significant benefit with closure of PFO versus medical management alone.
Arteriovenous malformations (AVMs) are considered the most dangerous congenital vascular anomaly. Cerebral AVMs occur in 0.1% of the general population. Ninety percent of brain AVMs are supratentorial. Symptoms typically present between the ages of 10 and 30, but older patients can be affected. It is more common in men and presents as intracerebral hemorrhage in 50% of cases. It may also present as a hemicranial, throbbing headache. We report a case of cerebello-pontine AVM presenting as an intracerebral aneurysm. A 51-year old female with history of osteoarthritis presented with dizziness, numbness and tingling of the left side of her face, left arm and leg associated with diplopia and blurred vision in her left eye of one day duration. Computed tomography (CT) of the head done at another facility revealed hyperdensity in the mid part of the cerebellum suspicious for hemorrhage or mass. On examination she had decreased sensation on the left side of the face. Muscle power was 4/5 on the left side and normal on the right. Magnetic resonance (MR) imaging and MR angiogram showed prominence of the left anterior inferior cerebellar artery. Three hours after admission she suddenly developed left facial droop and worsening of her headache. Repeat CT revealed a large aneurysm, confirmed by CT angiogram, extending off a left-sided branch of the basilar artery within the cerebello-pontine angle adjacent to the left side of the pons. Cerebral angiogram subsequently revealed an arteriovenous malformation. Multiple onyx embolizations were performed. Postoperatively her diplopia, left facial numbness and blurry vision in her left eye persist. She is unable to fully close her left eye but has full strength and improved sensation in her left side. She will be transferred to a rehabilitation facility. Arteriovenous malformations in the cerebello-pontine angle are extremely rare. Factors that influence the decision to treat include patient age, lesion size, location and prior history of intracerebral hemorrhage. Persistent symptoms are caused by facial nerve palsy due to compression at the cerebello-pontine angle. The risk of hemorrhage in brain AVMs is between 3 to 5 percent. Many studies have focused on the treatment of AVMs leading to technological advances that utilize a multidisciplinary approach including surgical, endovascular and radiosurgical techniques.
Drug-induced subacute cutaneous lupus (DI-SCLE) was first described in 1985 by Reed et al. He formulated an association between hydrochlorothiazide with DI-SCLE in five patients who had SSA/anti-Ro autoantibodies. Numerous medications have been implicated in drug-induced lupus syndromes, including commonly prescribed drugs such as calcium channel blockers (CCBs) and proton pump inhibitors (PPIs). We present an unusual and severe case of DI-SCLE in a patient taking both CCB and PPI chronically. A 60-year old female with a past history of chronic migraines/cluster headaches, gastritis, hypothyroidism, hyperlipidemia, chronic back pain presented with a generalized rash for two months. The first lesion appeared as a small circular, erythematous, papular, pruritic lesion on the left thigh, eventually becoming a scaly, crusty ulcer. Other similar lesions developed on the right thigh, arms, abdomen, chest, neck, back and face. She reported allergy to sulfa drugs and was taking verapamil and pantoprazole for over 15 years. Other medications were levothyroxine, oxycodone-acetaminophen, atorvastatin, cyclobenzaprine, topiramate, morphine and melatonin. She was initially treated with clindamycin, hydroxyzine and prednisone. Upon discontinuation of the steroids, the rash became more confluent. Multiple painful, pruritic, annular plaques developed with irregular papular-erythematous desquamated borders and central pigmentation, involving most of the body but sparing the nasolabial area, palms, and soles. No mucosal ulcers were seen. A skin biopsy reported interface dermatitis with focal epidermal necrosis. She was started on prednisone. Anti-Ro/SSA antibody was positive with negative anti-nuclear antibody (ANA), anti-Smith antibody, anti-La/SSB, anti dsDNA, HIV, and hepatitis panel. C-reactive protein was 13.5 mg/L and sedimentation rate was 19mm/h. Serum complements were normal. In light of the skin biopsy, verapamil and pantoprazole were discontinued and hydroxychloroquine started. Eventually oral steroids and hydroxychloroquine were tapered off as the lesions resolved without scarring, resulting in no further relapse. DI-SCLE is not a common diagnosis. The lower extremity skin lesions with positive anti-Ro/SSA antibodies, and the improvement when the offending agents were discontinued strongly support the diagnosis, despite the negative ANA. Only five cases of verapamil-induced SCLE have been reported in the past, with a mean latent period of 3.2 years and one case developing after six years. Our patient was on verapamil and pantoprazole for over 15 years. In previous reports, latent time between initiation of a PPI and onset of DI-SCLE varied between three and four months, therefore we believe pantoprazole is unlikely the causative agent, although this cannot be completely excluded.
Rhabdomyolysis caused by Total Body Potassium Depletion from Inappropriate Diet and Laxative Abuse.

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Rhabdomyolysis has been reported with severe hypokalemia (serum potassium levels<2.5 mEq/L), however serum potassium levels can be spuriously high during initial presentation, due to the leakage of intracellular contents secondary to cell breakdown. This may confound the diagnosis. We present a case of rhabdomyolysis caused by total body depletion of potassium secondary to chronic malnutrition from inappropriate diet and laxative abuse. A 64-year-old obese female with history of hypertension, diabetes, gout, osteoarthritis and bipolar disorder was brought to the hospital after being found lying in bed, with altered mental status, lethargy, drowsiness, generalized weakness and myalgia. She had lost about 50 pounds within a year, on a strict self-guided diet and chronic use of over the counter laxatives to avoid insulin therapy for her diabetes. On admission she was hypotensive and hypovolemic. Laboratory studies revealed serum creatinine 3.0mg/dl, BUN 34 mg/dl, potassium 2.9 mEq/L and creatine kinase 17000 units/L. Urinalysis was positive for myoglobin. Electrocardiogram, chest radiograph, computed tomography of the head and kidney/abdominal ultrasound were unremarkable. Fluid hydration and potassium replacement were initiated. Her clinical condition improved as total body potassium was repleted. She was discharged on day eight of hospitalization to a rehabilitation center. The total body depletion of potassium caused by inappropriate oral intakes and abuse of laxatives, was the determining factor for the development of rhabdomyolysis and subsequent acute kidney injury. Although our patient’s potassium level was above 2.5 mEq/L we recognize that potassium levels are increased by conditions associated with cell breakdown and could mask identification of hypokalemia as a cause for rhabdomyolysis. This case also emphasizes that unsupervised diet and chronic laxative use can lead to life-threatening conditions and as physicians we have to be vigilant and properly educate our patients.
Silicosis is an uncommon pulmonary disease, associated with high-risk occupations, but occasionally seen in other settings. We report a case of silicosis with resultant progressive massive fibrosis, likely secondary to remote cocaine use. A 64 year old male smoker, with a history of emphysema and productive cough for the past four years presented to the emergency department with dyspnea at rest over the past few weeks. One week prior, the dyspnea was treated with a course of moxifloxacin, without response. He had significant hypoxemia, was placed on BiPAP and transferred to the intensive care unit. Chest radiograph revealed bilateral hilar masses and fibrosis of the upper lung zones. Chest computed tomography disclosed bilateral conglomerate hilar masses, extensive fibrosis of the upper lung fields, with scattered nodules and lower lobe emphysema. Bronchoscopy revealed a markedly shiny bronchial endothelium, with compression from the hilar masses. Brushings were negative for infection or malignancy. Bronchial biopsy revealed birefringent crystals in a hyalinized granuloma consistent with a silicotic nodule. Further extensive social history failed to reveal any environmental exposure to silica, aside from a remote history of cocaine use. He has been referred for lung transplantation. The pathognomonic finding is the silicotic nodule of concentric hyalinized collagen with surrounding reticulin and birefringent particles in the periphery. Nodules may coalesce causing a variant known as progressive massive fibrosis (PMF), which results in significant clinical deterioration, and is likely the variant seen in our patient. Larger nodules are seen, with decreased DLCO, decreased PaO2, and a restrictive pattern on pulmonary function tests. Patients present with dyspnea, weakness, and weight loss, from months to years after exposure. Radiographs show lower airspace disease, in contrast to the other subtypes. Accelerated silicosis has a latency of 5 to 10 years, and progresses rapidly. Chronic or classic silicosis presents with a latency of more than 15 years, with sub-centimeter opacities in the upper zones. Findings of systemic autoimmune and connective tissue diseases are not uncommon. In all forms of silicosis, bronchitis and mycobacterial infection are seen. The likely pathogenesis of silicosis is due to the radical oxidative property of silica. This interacts with alveolar macrophages, generating inflammatory mediators, and sustained inflammation. As a result, there is an association with infection and autoimmune disorders. Radiographic findings include hilar masses and upper lobe nodules, representing silicotic nodules and surrounding parenchymal changes. The classic finding of “egg-shell” calcification of the lymph nodes is uncommon. PMF presents with large opacities in the upper lobes, with a compensatory emphysema of the lower lobes and periphery. Aside from lung transplantation, there is no effective treatment for silicosis, and management is directed towards support of residual lung function, and surveillance and treatment of the infective and autoimmune complications.
Dabigatran, a direct thrombin inhibitor, is approved for use as an alternative to warfarin for treatment for nonvalvular atrial fibrillation. Soon after its release, several concerns were raised about its safety, specifically reports of high incidence of life-threatening hemorrhage. With the availability of dabigatran and newer oral anticoagulants, it is extremely important to be aware of safety profiles, because of lack of tests that monitor the level of anticoagulation and unavailability of antidotes. There is consensus regarding the efficacy of dabigatran in stroke prevention as well as its lower incidence of intracranial hemorrhage. We chose to study the incidence of gastrointestinal hemorrhage at our institute with use of dabigatran and define associated risk factors. Methods: In a single center, retrospective cohort study, 148 eligible patients were selected from all inpatient visits over a 6 month period starting 1/1/2011. All patients were on dabigatran for secondary prevention of cardioembolic stroke. Patients with creatinine clearance <30ml/min and those with episode of gastrointestinal bleeding in 6 month period prior to admission date were excluded. Electronic medical record was reviewed for admission notes, discharge summaries, gastroenterology consult notes and laboratory and endoscopy reports. Standardized definitions were established to avoid inter-observer bias. Data were analyzed using SPSS 17.0. Results: A total of 148 patients aged 71±14 years were included, of which 75% were on 300mg daily dose of dabigatran. Concomitant use of aspirin and clopidogrel was noticed in 69% and 24% respectively. Incidence of gastrointestinal bleeding was similar to the RELY trial (8.1% vs 6.0% respectively, p>0.05), however major hemorrhages requiring intervention or transfusion were much higher (4.0% vs 1.6%, p<0.05). Incidence of bleeding was higher in older patients. Dose response relationship was not observed. Antiplatelet agents did not increase likelihood of hemorrhage. Discussion: Dabigatran is associated with high number of major gastrointestinal hemorrhages especially in the elderly. Oral anticoagulants which lack monitoring parameters and do not have established reversal protocols should be used with caution.
An otherwise healthy 66 year old male presented with seven episodes of right monocular vision loss that began shortly after taking a warm shower. Each episode lasted about 5-10 minutes, occurring over several hours, during which he experienced visual stimuli described as a “lightning-like” mix of colors. He denied any previous similar episodes. He takes no medications and denied alcohol, tobacco or substance use. There was no history of heart attack, stroke, hypertension, diabetes, heart disease or previous episodes of dizziness, syncope or chest pain. Prior yearly eye exams were unremarkable. He saw an optometrist the day the deficits began and was told his visual acuity was decreased in the left eye. In the Emergency Department he had no residual visual deficits. Physical examination revealed difficulty with accommodation to finger approaching nose test due to apparent strabismus. Visual acuity was diminished in both eyes without glasses but with lenses was 20/30. Blood pressure was 175/88, pulse 59, respiratory rate 15, temperature 98.2 F, oxygen saturation 99% on room air. Neurologic examination was normal. EKG showed normal sinus rhythm at rate of 59 with ST-T abnormalities in the anterolateral leads and left ventricular hypertrophy without previous EKG available for comparison. Two-dimensional echocardiography showed concentric moderate left ventricular hypertrophy with ejection fraction estimated at 60-65%. Magnetic resonance imaging of the brain without contrast showed diffuse small vessel disease. Magnetic resonance angiogram of the head without contrast found mild to moderate stenosis in both cavernous internal carotid arteries (the artery’s course where it is situated between the layers of the dura mater forming the cavernous sinus but covered by the lining membrane of the sinus) and fetal origin of the left posterior cerebral artery (PCA). Ultrasound of the carotids did not demonstrate any significant stenosis in either internal carotid artery. Computed tomography of the head was unremarkable. The patient was discharged on an aspirin and statin regimen for cerebrovascular protection. Uhthoff’s symptom/sign/phenomenon is a transient temperature-dependent weakness, numbness or loss of vision secondary to impaired nerve conduction. This can occur when the temperature becomes elevated. In patients with damaged nerves due to demyelination, the temperature that impulses can be successfully conducted becomes lowered. Often this transient neurologic dysfunction occurs following a hot shower, exercise, or during fever. Although this symptom is commonly associated with multiple sclerosis demyelinating optic neuropathies, it has also been observed in Leber’s hereditary optic neuropathy and sarcoidosis as well as neurologic syndromes including Freidreich’s ataxia, posterior cerebral arterial insufficiency, and intrasellar/parasellar tumors. Can a fetal origin PCA, a common variant seen in roughly 30% of the population, have a role in mimicking symptoms of optic nerve demyelination?
A 31 year old male with no past medical history presented with intermittent left-sided flank pain for three weeks. Associated symptoms included nausea, vomiting, chills without fever, intermittent frothy urine and bilateral lower extremity edema. He denied gross hematuria or recent respiratory illness. The remainder of the review of systems and family history were non contributory. On examination blood pressure was 163/89mmhg, pulse rate 100 bpm. He was afebrile. He was in mild distress due to pain, with left costovertebral angle tenderness and 4 (+) pitting edema. Laboratory tests showed BUN 18 mg/dL, creatinine 1.1 mg/dL, total serum protein 5.5 gm/dL, albumin 1.2 gm/dL. Total cholesterol was 288 mg/dL and LDL was 255 mg/dL. Urinalysis showed protein > 500, moderate blood and hyaline casts 7/lpf. 24-hour urine collection revealed 10 grams of protein. Computed tomography of the abdomen and pelvis showed retroperitoneal and iliac lymphadenopathy, right cardiophrenic lymph nodes, and moderate left hydronephrosis without renal/ureteric calculus likely secondary to lymphadenopathy. He received a left percutaneous nephrostomy tube. Alpha-feto-protein, HCG tumor markers, serum protein electrophoresis, testicular ultrasound and HIV screen were normal. Lymph node biopsies were inconclusive for lymphoma. ANA was 1:2560 with negative anti-DS DNA. ASO titer was 772 with positive streptozyme but negative streptococcal throat cultures. Serum complement levels C3, C4 and CH50 were all decreased. Right kidney biopsy revealed lupus nephritis stage IV and V. He was started on prednisone, mycophenolate mofetil and lisinopril. His obstructive uropathy resolved and the nephrostomy tube was removed. Five months later he was readmitted with acute renal failure and anasarca, secondary to non-adherence to the medication regimen because of gastrointestinal side effects. Admission creatinine was 6.1 mg/dL, BUN 68 mg/dL and potassium of 5.7 mmol/L. He received intravenous pulse methylprednisolone and was started on hemodialysis. Subsequent right kidney biopsy showed thrombotic microangiopathy and crescentic glomerulonephritis with type V features. Beta-2 glycoprotein antibodies and lupus anticoagulant were negative. IgM anticardiolipin antibody was minimally elevated at 13 (normal less than 12). He is currently on oral prednisone and monthly intravenous cyclophosphamide with outpatient hemodialysis. Anticoagulation was withheld because of a post-renal biopsy hematoma. Although lymphadenopathy is a feature of SLE, it rarely causes obstructive uropathy as the initial presentation with lupus nephritis. Our patient’s obstructive uropathy resolved with immunosuppressive therapy with subsequent removal of his percutaneous nephrostomy drain. Induction therapy for lupus nephritis includes glucocorticoids with either high dose mycophenolate melfitol or intravenous cyclophosphamide. Aggressive management of hypertension with angiotensin-converting-enzyme inhibitors and control of hyperlipidemia with a statin is also recommended. Patients should also be placed on prophylaxis for opportunistic pneumonias, gastro esophageal reflux and osteoporosis.
Anabolic steroid use has been around for decades, especially with the cultural breakthrough of modern day body building, with reported prevalence as high as 67% in body builders. The medical community has expressed reservations regarding this practice, with many reports of their effects on health. Kidneys are rarely mentioned as targeted organs. Few cases can be found documenting nephrotic syndrome in anabolic steroid users. We present the case of a 30-year old man with history of testicular failure, who presented with new onset anasarca. He has been using anabolic steroids Masteron® and Sustanon® for six months, in addition to daily pure whey protein powder. He has been on testosterone injections since age 12. Initial examination showed he was hypertensive with a blood pressure of 199/97 mmHg and had anasarca. 24 hour urine creatinine was 2.15 mg, with a 24 hour urine protein of >4800 mg. He was diagnosed with nephrotic syndrome, and kidney biopsy showed podocyte effacement on electron microscopy. No evidence of glomerular sclerosis could be appreciated, hence the diagnosis of minimal change disease (MCD). He was started on intravenous steroids, furosemide, and his blood pressure was controlled with medical therapy. He agreed to stop his anabolic steroids. The patient was discharged on oral prednisone, with improved kidney function and decreased proteinuria. Rhabdomyolysis, drug-induced cholestasis, interstitial nephritis and acute tubular necrosis have been previously described in cases of acute kidney injury in anabolic steroid users. Nephrotic syndrome has been previously reported in anabolic steroid users, however, all concerned cases were diagnosed to be focal segmental glomerulosclerosis (FSGS). No cases of new onset MCD exist in this population to our knowledge. The diagnosis of MCD in our patient could well be unrelated to anabolic steroid use, or could be a misdiagnosed early FSGS. The fact the he responded to steroid therapy weighs in favor of MCD, although FSGS in its early stage can be hypercellular and respond to steroids. More studies are necessary to elucidate the mechanism of nephrotic syndrome in anabolic steroid users.
Recurrent syncope in a patient with mediastinal seminoma

Garo Garabedian, Amit Ray, Le Wang, Tome Nascimento

Seminoma is a malignant germ cell tumor of the testes but rarely originates directly from extra-gonadal locations such as the mediastinum. It is a rare malignant neoplasm; however, it represents one of most treatable and curable cancers with 95% cure rate if discovered early. We report a rare case of extragonadal seminoma presenting with recurrent syncope. A 53-year old man with no known significant past medical history presented with recurrent syncope and increasing chest fullness which was constant and increasing in intensity over a few weeks, unrelated to exercise. His symptoms were accompanied by intermittent shortness of breath and palpitations. On physical examination, he was hypotensive with blood pressure 70/50 mmHg. He was stabilized with intravenous fluids. There was no cardiac murmur or gallop and his rate was regular. There was no jugular venous distention, facial plethora or distended chest veins. Lungs were clear to auscultation. There was no palpable cervical lymphadenopathy, and his abdominal examination was unremarkable. He did not have peripheral edema. Initial chest radiograph revealed mediastinal enlargement. Computed tomography (CT) of the chest revealed large mediastinal lymphadenopathy, pressing against the superior vena cava and the right main pulmonary artery. CT scan of the abdomen/pelvis demonstrated bulky retroperitoneal and celiac axis lymphadenopathy. Echocardiogram revealed a 4.4 cm x 6.3 cm echogenic structure impinging on the left atrium. Absence of respiratory variation in the inferior vena cava suggested elevated right atrial pressure. Serum tumor markers showed markedly elevated lactate dehydrogenase and human chorionic gonadotropin with normal alpha-feto-protein. CT-guided biopsy of the parailiac lymph node showed malignant seminoma. Chemotherapy with cisplatin and etoposide was initiated. After completion of 4 cycles of chemotherapy, he was in remission. Follow-up echocardiogram one month later showed a reduction in the size of the mass impinging on the left atrium to 3.4 cm x 3.5 cm. He feels well, has not had any new syncopal episodes and continues to see his oncologist and cardiologist regularly. Mediastinal tumors occur from a wide range of conditions, including Hodgkin and Non-Hodgkin lymphomas, thymic mass and extragonadal germ cell tumors. The latter occur mostly in the anterior mediastinum and constitute 10-20% of mediastinal masses. Of those, more than one third are seminomas, and are predominant in young males. Chemotherapy is the mainstay of treatment as most seminomas are exquisitely sensitive to chemotherapy. Rarely, radiation or surgical intervention plays a role in the management of extragonadal germ cell tumors. This case illustrates an important point of making the diagnosis first before initiating treatment.
Gallstone and alcohol are the leading causes of acute pancreatitis which results in 100,000 hospitalizations annually. Drug-induced pancreatitis has also been well reported in the literature. We report a case of cannabis-induced acute pancreatitis in a young male. A 24-year old male with a history of Crohn disease presented to the Emergency Department with a one week complaint of abdominal pain. He reported intermittent, sharp, right lower quadrant and back pain, stabbing in nature, lasting less than a minute with no aggravating or alleviating factors. He also had painless diarrhea with a single episode of bloody stool and nausea. There was no vomiting, fever, tenesmus, heartburn, chest pain or shortness of breath. He denied alcohol use but admitted to smoking 5 marijuana cigarettes daily. His last adalimumab treatment was six months ago. He has never required steroids. On examination his body mass index was 19.83. There was no pallor or jaundice. Abdomen was soft, non-distended with right lower quadrant tenderness. Bowel sounds were present. Complete blood count, basic metabolic panel and hepatic function were unremarkable. Serum lipase was 1871 U/L. Urine drug screen was positive for cannabinoids and opiates. Abdominal ultrasound revealed mild dilatation of the pancreatic duct at 3mm suggestive of acute pancreatitis. The gallbladder and common bile duct were visualized with no evidence of cholelithiasis, gallbladder wall thickening or choledocholithiasis. Computed tomography of the abdomen revealed marked thickening of terminal ileum and cecal wall. ANA and IgG 4 Antibodies were negative. He was treated with intravenous fluid resuscitation and pain management. His symptoms improved, the diarrhea resolved and the serum lipase trended to within normal limits. He was subsequently discharged to home in stable condition. Cannabis has been reported to be associated with 146.2 cases per 100,000 ED visits in 2011 as per the Drug Abuse Warning Network, USA. The exact mechanism resulting in acute pancreatitis through marijuana use remains obscured. Recently, agonism of CB1 and CB2 receptors are thought to play a role in the pathogenesis. Both of these receptors are expressed on the pancreas. Our case suggests marijuana use as one of the confounding factors to consider in a patient with acute pancreatitis.
Sarcoidosis is an immune-mediated disease of unknown cause with multi-organ association. While it may present with pulmonary, lympho-cutaneous or ocular manifestations in approximately 90% of cases, renal involvement is less frequent. Because of its rarity, the actual incidence of sarcoid-related renal dysfunction cases is not known. We present a very rare case of isolated renal sarcoidosis presenting as end stage kidney disease. A 43 year-old male with hypertension and dyslipidemia presented with anorexia, fatigue, weight loss and mild urinary retention. He had not seen a physician in ten years. On examination his blood pressure was 157/93 mmHg. Physical examination was unremarkable. Laboratory results revealed serum creatinine of 8.0 mg/dL (ref range 0.7-1.2 mg/dL), BUN 87 mg/dL (ref range 8-21 mg/dL), hemoglobin 9.9 g/dL, serum bicarbonate 14 mmol/L, with normal electrolytes. Further work up revealed mild hematuria without casts, and protein/creatinine ratio of 21.29 (ref range <0.15). Renal biopsy revealed non-caseating granulomas, advanced scarring, and chronic interstitial nephritis consistent with renal sarcoidosis. Acid-fast and fungal stains were negative. Chest radiograph was unremarkable. Serum angiotensin converting enzyme was elevated at 73 U/L (ref range 12-68 U/L) and sedimentation rate was 102 mm/hr (ref range 0-15 mm/hr). He was started on prednisone 1mg/kg/day and prepared for long-term renal replacement therapy. Upon initiation of dialysis, his uremic syndrome resolved with marked improvement in clinical condition, but he remained dialysis dependent without renal recovery after 3 months of hemodialysis. There are very few reported cases of renal sarcoidosis that progressed to end stage renal disease. In addition, to our knowledge, there are no known reported cases of isolated renal sarcoidosis-induced end-stage renal disease at presentation. The majority of documented cases of renal sarcoidosis had shown improvement in renal function with steroids. In our patient, it is likely that late treatment prevented recovery, hence early recognition of renal sarcoidosis may be crucial to prevent progression to end stage renal disease.
Medical management can be simple and sometimes complicated. This case illustrates how an adverse effect of a medication can create another process that indirectly worsens the patient’s underlying problems. A 67 year-old male farmer with a past history of chronic hyponatremia due to syndrome of inappropriate anti-diuretic hormone (SIADH), atrial flutter, and hypertension presented with a sudden onset of dizziness while at work. He denied palpitations, syncope, dyspnea, chest pain or gastrointestinal symptoms. He had a history of remote alcohol abuse. Nine months prior he was admitted for the same complaint and was found to have hyponatremia due to SIADH. He was also started on amiodarone and dabigatran for atrial flutter, and lisinopril for hypertension. On examination his blood pressure was 143/101 mmHg with negative orthostasis, heart rate 72 bpm with normal cardiopulmonary, abdominal and neurologic exam. There was trace edema in lower extremities. Serum sodium was 118 mmol/L, potassium 3.8 mmol/L, creatinine 0.7 mg/dL, thyroid stimulating hormone 127 uIU/ml, amiodarone level 1.2, urine osmolality 241; urine chloride 105, urine potassium 15.4, urine sodium 101, urine creatinine 54.7. Urine toxicology was negative. Thyroglobulin and thyroid peroxidase antibodies were negative. Computed tomography of the head was unremarkable. Electrocardiogram revealed a normal sinus rhythm rate of 72 bpm. Prior to start of amiodarone, his serum sodium was 130 mmol/L and his thyroid stimulating hormone was 3.75 uIU/ml. He was diagnosed with hypo-osmotic hyponatremia and hypothyroidism secondary to amiodarone. Through treatment of water restriction and Democlocycline, his sodium level is back at base line. He is currently on levothyroxine. In conclusion, amiodarone is a class III antiarrhythmic agent with high iodine content that has some structural similarity to thyroid hormone. Hypothyroidism occurs in 8-10 % of patients due to the suppressive effect of iodine overload which can be managed with levothyroxine therapy without a need to stop amiodarone. Our patient had chronic hyponatremia that was well controlled until amiodarone was added into his regimen leading to hypothyroidism and hence an acute on chronic hyponatremia picture. This case exemplifies the complexities of medical management.
AtlantiCare

Ertapenem for Clostridium innocuum bacteremia

Bhagyashri Navalkele, Manish Trivedi, Joseph Reilly

Clostridium innocuum is a spore-forming gram-positive anaerobic rod which is a part of the normal oral and gastrointestinal flora. We present a case of C.innocuum bacteremia in a 104-year old male with a successful outcome. There have been 13 identifiable case reports published in the literature regarding C.innocuum as a pathogen. A 104-year old male was admitted after a fall. Once hospitalized, he was found to be in septic shock with blood pressure of 73/43mmHg, heart rate of 101 beats per minute, and a temperature of 99 degrees Fahrenheit. He had an altered mental status with otherwise unremarkable physical exam findings. Laboratory examination revealed bicarbonate of 12mmol/L, aspartate transaminase of 190U/L, alanine transaminase of 113U/L, hemoglobin of 8.1g/dL, hematocrit of 24.6%, white blood cell count of 26,000/mm3 with bands of 50 and 71% polymorphonuclear leukocytes, and platelets of 79,000/mm3. Blood, sputum, and urine cultures were obtained. Urine analysis was unremarkable. He was empirically started on vancomycin with ertapenem. Two anaerobic blood cultures were positive for Clostridium innocuum while only one bottle being positive for Candida albicans and Prevotella loescheii. Computed tomography of the abdomen and pelvis were unremarkable. The presence of normocytic normochromic anemia and thrombocytopenia raised suspicion for underlying malignancy. He declined any further investigation. He received ertapenem with vancomycin for 10 days. The patient improved with negative blood cultures after 72 hours of starting ertapenem. He was discharged on day 20. Clostridium innocuum was first identified as a pathogen in humans in 1962. The bacterium name was attributed as “innocuum” because of it’s lack of virulence. Clostridium infections are typically polymicrobial in 40-50% of the cases and commonly associated with soft tissue, gastrointestinal tract, biliary tract, and female genital tract infections. Clostridium bacteremia constitutes 0.7-2.6% of all bacteremia episodes; with Clostridium perfringes being the most common isolate. Clostridium innocuum are unique in that they have decreased susceptibility to cephalosporins, clindamycin, and quinolones. The bacteria are intrinsically resistant to vancomycin (MIC 4-16mcg/mL). Mortality related to C. innocuum bacteremia is 30%, with in-hospital mortality of 15-86%, and greater than 40% of those infected are cancer patients. Poor outcomes have been attributed to advanced age and malignancy. To our knowledge, this is the first case report where ertapenem was used with successful treatment outcome for C. innocuum bacteremia. Ertapenem has excellent coverage for most anaerobic pathogens including clindamycin resistant isolates. It offers favorable in vitro activity against anaerobic bacteria when compared to piperacillin-tazobactam or metronidazole. We emphasize the importance of appropriate antimicrobial therapy for successful treatment outcomes in C. innocuum bacteremia.
Recurrence Hemangioblastoma in Von Hippel-Lindau Disease

Ifeoma Nwankwor, Castaneda, Jose MD, Hamaty Jr D O, Edward G

Von Hippel-Lindau (VHL) disease is a hereditary autosomal dominant syndrome with multisystem neoplastic disorder. It is characterized by hemangioblastomas (HGB) primarily of cerebellum, retina, brainstem and spinal cord, as well as visceral tumors including renal cysts, pheochromocytomas, and pancreatic tumors. Hemangioblastoma (HB) is rare benign tumor that account for 2.5% of intracranial neoplasms, and one quarter of all hemangioblastomas are associated with VHL. We present a case of recurrent HB in a patient with possible VHL disease. A 28-year-old female presented with a two week history of constant posterior pulsatile headaches. She had a past medical history of a brain “vascular tumor” and right eye tumor that were resected about 10 years prior, resulting in right eye blindness. She also had a positive family history of “tumors”. Computed tomography of the head and magnetic resonance imaging of the brain revealed a 3.3 x 3.1 x 3.5 cm cerebellar mass with mixed cystic and solid components and evidence of hemorrhage in the cystic portion. There was significant compression of the brainstem with vasogenic edema in the cerebellum. Cerebral angiogram showed an extensive vascular network supplying the mass. Due to the extent of the cerebral edema, hypertonic saline, levetiracetam and intravenous steroids were started in the neuro-intensive care unit. She underwent a therapeutic embolization of the feeder vessels, with subsequent sub-occipital craniotomy during which the mass was completely resected. Abdominal ultrasound showed a 1.8 cm right renal lesion and a 2cm left renal cyst. Plasma free metanephrines were negative. She had a normal recovery with a mild left upper limb dysmetria, and was discharged to a rehabilitation center on day 13 post surgery. Pathology of the cerebellar mass was consistent with hemangioblastoma. At subsequent follow-up, she did not have any neurological deficits and is being evaluated by the geneticist and radiation oncology for VHL disease. Hemangioblastomas are benign tumors, but reoccur in up to 27% of cases after surgical excision, as we saw in this case. Recurrence usually occurs with tumors that present initially at younger age (before the age of 30), and in VHL patients (60 to 70 % of cases). Acute hemorrhage is rare in HB, and can cause obstructive hydrocephalus, brain stem compression and cerebellar tonsillar herniation. VHL occurs due to inactivation of the tumor suppressor gene 3p25. The presentation of HB should prompt physicians to be vigilant of the possibility of VHL by inquiring about the history of other tumors in the patient and family members. The diagnosis of VHL is done by genetic testing and should be recommended in all similar cases.
Polyglandular Autoimmune Syndromes (PGAS) are caused by an immune-mediated destruction of endocrine glands and nonendocrine organs. They are classified into three major types: types 1, 2, and 3. Type 3 was subsequently subcategorized into type IIIa, IIIb, IIIc. We report an interesting case of a male patient showing features of both type IIIb and IIIc PGAS. A 42 year old male recently diagnosed with Hashimoto thyroiditis and hypothyroidism presented with weakness and numbness of upper and lower extremities, unsteady gait, fatigue, dyspnea on exertion, constipation and erectile dysfunction over several months. He had no known history of recent tick bites. Physical exam revealed normal speech and intact cranial nerves. Muscle tone was normal, Reflexes were +2 in all extremities. Strength in both lower extremities was 3-4/5, with decreased sensation to touch, pinprick and vibration in both lower extremities. His gait was ataxic with a positive Romberg’s sign and intact finger to nose test. Skin hypopigmentation consistent with vitiligo was noted. Laboratory evaluation revealed hemoglobin of 8g/dl; vitamin b12 108pg/m, intrinsic factor antibody positive, TSH 100.4uIU/ml, FT4 0.61ng/dl, FT3 1.60ng/dl, anti-microsomal antibody 600 iu/ml, anti-thyroglobulin antibody 76 iu/ml, total testosterone 279ng/dl; free testosterone 8.17ng/dl; LH of 9.94mIU/ml and FSH 5.56mIU/ml. He had normal levels of blood glucose, calcium, parathyroid hormone, cortisol and aldosterone. Cosyntropin stimulation test was negative. ELISA and Western Blot were positive for Lyme disease. He was treated for hypothyroidism, B12 deficiency and acute Lyme disease. He currently has autoimmune thyroid disease, pernicious anemia and vitiligo and has not demonstrated adrenal insufficiency or any other autoimmune disease. PGAS types IIIa, IIIb, IIIc have immune thyroiditis along with immune-mediated diabetes, pernicious anemia, vitiligo or alopecia respectively. Approximately 3.5-4% of the total population has PGAS type 3 with 5% being women and 1.5% being males. In a case study done of patients with thyroiditis, 14% had PGA type III and all were women. PGA type 3 has both genetic and environmental components. Environmental factors such as viral infection may exaggerate the ongoing immune response and precipitate glandular failure; similar to a well recognized link between congenital rubella infection, hypothyroidism, and type 1 diabetes. This case demonstrates that one may have variant forms of PGAS. Further more it also illustrates that atypical environmental factors can possibly play a role in the development of these syndromes. Viruses have been proposed as possible factors but infections caused by other organisms should also be considered.
Vibrio parahaemolyticus septicemia in a Patient with Newly Diagnosed Multiple Myeloma

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Vibrio parahaemolyticus (VP) is a gram-negative anaerobic facultative bacillus with halophilic properties. It commonly presents as gastroenteritis, with an incubation period ranging from 4 to 96 hours, leading to dehydration in most cases. On rare occasions it leads to septicemia (about 5%), mainly in patients with underlying medical conditions or immunosuppressive states. We report a case of Vibrio parahaemolyticus septicemia (VPS) in a patient with newly diagnosed multiple myeloma (MM) who presented with a massive ischemic stroke. To our knowledge this is the first such case reported. A 64-year old male with history of hypertension, uncontrolled non-insulin dependent diabetes mellitus, paroxysmal atrial fibrillation, coronary artery disease, ischemic cardiomyopathy and chronic kidney disease was admitted after being found unresponsive. He was last seen two days prior, and complained of diarrhea and vomiting after sea food consumption. Physical exam revealed an intubated male with temperature of 101.4°F, BP 133/103 mmHg, irregular pulse 123 bpm, 100% oxygen saturation on 60% FIO2 with ventilator support. Orogastric tube was in place. Pupils were 2 mm equally reactive to light. He withdrew to noxious stimuli moving his right upper extremity more than his left. Blisters and abrasions were noted on both legs. Computed tomography revealed a large ischemic stroke in the right middle cerebral distribution involving 2/3 of the right brain with edema and mass effect. Serum creatinine was 1.6mg/dl, BUN 29 mg/dl, glucose 232mg/dl, bicarbonate 28mmol/l, calcium 7.8mg/dl, total protein 8.7g/dl, albumin 2.9g/dl, albumin to globulin ratio (A/G) of 0.52, white blood cell count 10.0 x 103, hemoglobin 11.6g/dl, platelets 252,000 with normal liver function tests and lactate level of 2 mmol/l. Hepatitis panel and lymphocyte profile were unremarkable. Initial blood cultures revealed Gram-negative rods for which he was started on piperacillin/tazobactam. Final blood cultures grew Vibrio parahaemolyticus. He developed septic shock with acute respiratory distress syndrome (ARDS). An IgG monoclonal protein with lambda light chain specificity and urinary Bence Jones protein were identified confirming the diagnosis of multiple myeloma. He required a short course of hemodialysis due to worsening renal failure. He gradually improved and was transferred to a long-term acute care facility. VPS is rare and conveys a mortality of up to 29%. MM is a plasma cell malignancy with an estimated life expectancy of 10 years depending on treatment. Multiple myeloma produces an immunosuppressive state putting patients at higher risk for bacterial infections. We speculate that the initial dehydration caused by the gastroenteritis may have also produced a hyperviscosity state in this patient playing an important role in the development of the stroke.
Adrenocortical carcinoma (ACC) is a rare malignancy with poor prognosis. The incidence of ACC is estimated at 0.7 to 2.0 cases per million population each year. Surgery is the only curative therapy available. We report a rare case of ACC with cardiac metastasis causing symptoms due to dynamic obstruction. A 59-year old female presented with complaints of increasing lower extremity edema, progressive dyspnea, and increasing abdominal girth with early satiety. There was no weight loss or chest pain. During an outpatient echocardiogram a large pedunculated mass was seen in the right atrium extending from the inferior vena cava (IVC) and flopping through the tricuspid valve, causing dynamic obstruction consistent with myxoma. Computed tomography of the chest, abdomen, and pelvis showed distal left lobe pulmonary emboli. The right kidney was displaced by a large heterogeneous mass arising from the right adrenal gland. The IVC was displaced anteriorly and compressed by the mass with hypodensity noted in the IVC potentially representing tumor thrombus. Magnetic resonance imaging demonstrated the 13 x 9.5 x 10 cm mass in the suprarenal region, deforming the right hepatic lobe. A prior ultrasound of the abdomen performed in October 2011 revealed no lesion in the kidneys or adrenals. She had persistent hyperglycemia, hypokalemia, and hypertension. Hypercortisolemia was found with low ACTH (<1.1 pg/mL), elevated random serum cortisol (34.3 ug/mL), a non-suppressible cortisol after 1mg dexamethasone, and 24-hour urinary free cortisol of 1350 ug/24 hr with elevated testosterone (329 ng/dL) and androstenedione (2654 ng/dL). Aldosterone, renin, urinary and fractionated metanephrines, DHEA, 17-hydroxyprogesterone, and estradiol were within normal limits. She underwent a right adrenalectomy en bloc with right nephrectomy, cardiopulmonary bypass for atriotomy with tumor extraction, and wedge resection of the left lateral segment of the liver for metastases detected during surgery. Tumor histopathology suggested adrenocortical carcinoma with central necrosis and vascular invasion. It was strongly positive for inhibin and focally for calretinin and negative for S100 and chromogranin. The atrial mass was also suggestive of adrenocortical carcinoma. Post-operatively, her symptoms improved. 24 hour urinary free cortisol decreased to 20 ug/24 hr and androstenedione, though still elevated, dropped to 336 ng/dL.

After surgical resection, current NCCN guidelines recommend consideration of systemic therapy, preferably in clinical trial, for metastatic adrenocortical carcinoma. The only pharmaceutical agent approved for adrenocortical carcinoma is mitotane, which is often used in combination with several different chemotherapeutic agents, including streptozocin or etoposide, doxorubicin, and cisplatin. Unfortunately, among patients with metastatic disease, the five year survival rate is 15%.
Takotsubo cardiomyopathy, also known as stress-induced cardiomyopathy, is an increasingly reported syndrome typically characterized by chest pain and transient systolic dysfunction manifesting as apical ballooning of the left ventricle in the absence of obstructive coronary artery disease. We report a case of Takotsubo cardiomyopathy after a near-drowning event. A 54-year-old woman with a history of borderline diabetes and sinus tachycardia presented with severe dyspnea after being caught in a riptide. She was able to swim to shore and did not recall aspirating salt water. She was very hypoxic and intubated in the field. On admission, systolic blood pressure was in the 70’s and norepinephrine was initiated. Examination did not reveal neck vein distention. Rales were heard bilaterally. Cardiac examination was unremarkable. Electrocardiogram showed sinus tachycardia with subtle ST-elevation in the anterior leads. Chest radiograph demonstrated extensive bilateral airspace disease. Echocardiogram demonstrated severe mid-cavity hypokinesis, slightly better at the base with the apex poorly visualized. Initial troponin was 3.3 ng/mL. Cardiac catheterization showed a variant Takotsubo cardiomyopathy with moderate to severe pulmonary hypertension and moderate diagonal disease. Her pulmonary capillary wedge pressure was 22 mmHg and an intraaortic balloon pump was inserted. Left ventriculography showed mid-cavity hypokinesis with preservation of the base and apex typical for mid-cavity variant Takotsubo. After 48 hours, vasopressors were discontinued and intraaortic balloon pump was removed. On day three, repeat echocardiogram revealed an estimated ejection fraction of 50%. She was successfully extubated and discharged home on day seven. Takotsubo cardiomyopathy should be suspected in patients presenting with acute coronary syndrome, particularly in postmenopausal women after exposure of intense psychological or physical stress. Typical diagnostic criteria include transient wall motion abnormalities extending beyond a single coronary distribution, absence of obstructive CAD, presence of new electrocardiogram abnormalities or modest troponin elevation, and absence of pheochromocytoma or myocarditis. Only a few cases of Takotsubo have been reported associated with near drowning. Our patient presented with dyspnea but without chest pain. Pulmonary edema in a near-drowning patient may be attributed to aspiration of salt water. However, in a patient presenting with unexplained left ventricular systolic dysfunction, the diagnosis of Takotsubo should be considered. As far as we are aware, the mid-cavitary variant, which represents less than 20% of Takotsubo cardiomyopathy, has not been previously reported in near-drowning patients. Despite the severity of the acute illness, most patients recover their normal left ventricular function within one to four weeks. Management of stress-induced cardiomyopathy is largely supportive including alleviation of triggering physical or emotional stress. Intraaortic balloon pump should be considered in patients with hypotension refractory to volume resuscitation and medical therapy.
5-Fluorouracil (5-FU) is commonly prescribed for treatment of a variety of cancers, including breast, ovarian, head and neck, and gastrointestinal. The enzyme dihydropyrimidine dehydrogenase (DPD) is responsible for the rate-limiting step in 5-FU catabolism and therefore is necessary for clearance of 5-FU. When enzyme activity is low, myelosuppression, stomatitis, diarrhea, dermatitis, neurotoxicity, and cardiotoxicity can result. In rare cases, these can prove fatal, as evidenced by our case. A 59-year old female was admitted with a three day history of nausea, vomiting, and diarrhea, four days after starting neoadjuvant modified FOLFOX-VI followed by 46 hours of continuous 5-FU infusion for stage IVA moderately differentiated invasive adenocarcinoma of the descending colon with metastases to the liver. She was afebrile with pulse of 113 and blood pressure of 89/58 mmHg. Poor skin turgor was noted with grade IV mucositis, oral thrush, and facial swelling of the eyes and the lower lip with drooling. Her platelets were 158x10^3 cells/µL (baseline 401x10^3 cells/µL) and white blood cell count was 7.2x10^3 cells/µL. She received intravenous fluconazole, normal saline, nutrition support, and GI/DVT prophylaxis. Given the patient’s early systemic reaction following induction of chemotherapy, her oncologist recommended testing for the DPD gene mutation to establish if 5-FU should be used at a reduced dose, if at all, in her cancer treatment regimen. Three days after admission (day ten post initiation of FOLFOX-VI) her platelets dropped to 50x10^3 cells/µL and white count dropped to 0.4x10^3 cells/µL with an absolute neutrophil count of 216 cells/µL. Daily filgrastim and neutropenic precautions were instituted. Stool studies and cultures of blood, sputum, and urine were unremarkable. Chest radiograph showed left sided pneumonia treated with cefepime and vancomycin. She developed refractory hypotension, hyperglycemia, coagulopathy, severe metabolic acidosis and hypoxemia, requiring intensive care and intubation with mechanical ventilation. Because of her terminal illness, comfort measures were instituted and she was terminally extubated. Results of the dihydropyrimidine dehydrogenase test confirmed that she had two copies of the IVS14+1 G>A mutation of the DPD gene. Unfortunately, without DPD, catabolism of 5-FU is greatly reduced and the drug can reach toxic levels. Few patients with DPD deficiency will have a positive family history, so identifying patients who would benefit from costly genetic testing is difficult. Thus far, a cost-effective solution for screening the entire population of patients receiving 5-FU has yet to be developed and as this case illustrates, could be potentially lifesaving.
Hyperviscosity-related hemorrhagic stroke in patient with Waldenstrom macroglobulinemia

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Waldenstrom macroglobulinemia is an indolent B-cell lymphoma characterized by bone marrow infiltration of lymphoplasmacytic cells and elevated IgM monoclonal gammopathy. Hyperviscosity occurs from pathologic elevation of pentameric IgM molecules, resulting in decreased flow and impaired microcirculation in critical organs, such as central nervous system, retina, and heart. It can rapidly manifest as medical emergency, causing significant morbidity and mortality. Symptoms are usually associated with neurological dysfunction, retinal hemorrhage, or aggressive heart failure. Early recognition and plasmapheresis to remove IgM from the circulation is the key in the management of hyperviscosity state. We report a rare case of hyperviscosity-related hemorrhagic stroke in patient with newly diagnosed Waldenstrom macroglobulinemia. An 82-year old Caucasian man with well-controlled hypertension and monoclonal IgM spike was seen in emergency room with severe headache, stupor, slurry speech, and weakness. On exam, blood pressure was 200/102mm Hg, with decreased muscle strength in all extremities and blurry vision. WBC 5.9x 10^3, hemoglobin 7g/dl, platelets 243x10^3, LDH 234U/L, AST 343U/L, ALT 497 U/L, PT 14.5sec, INR 1.4, and PTT 32.4 sec. Computed tomography revealed right thalamic bleed along with large intraventricular hemorrhage. He was transfused with 2 units of packed red blood cells and 4 units of fresh frozen plasma. Emergent ventriculostomy was placed for external blood drainage. Serum IgM was 3g/dl. Serum viscosity, however, was too thick to measure, leading to the clinical suspicion of hyperviscosity-related hemorrhagic stroke. Emergent plasmapheresis was initiated. Serum viscosity subsequently dropped to 8.4 centipoises (CP) after first cycle of plasmapheresis, and continued to fall with additional plasmapherases until complete resolution of neurological symptoms. Immunofixation confirmed the monoclonal IgM with lambda specificity. Bone marrow biopsy was positive for infiltration of small lymphoplasmacytic cells, consistent with Waldenstrom macroglobulinemia. In this case, we believe that the elevated serum viscosity was the ultimate cause for the intracranial bleed. The transient elevation in blood pressure was merely a secondary effect from the intracranial event. In retrospect, blood transfusion should be avoided in patient with hyperviscosity. Most patients become symptomatic when serum viscosity exceeds 6 CP. However, the decision to initiate plasmapheresis is based on the clinical scenario, rather than serum viscosity. Asymptomatic Waldenstrom macroglobulinemia does not require treatment. Indications for disease-modifying therapy are anemia (hemoglobin <10g/dl), constitutional symptoms, hyperviscosity symptoms or bulky disease. Rituximab, a monoclonal antibody against CD20 molecules on malignant cells, has been shown to be the most effective treatment of choice. While the role of chemotherapy is less well defined, its combination with Rituximab may provide a new way of treating this disease.
A 69-year old male presented with sharp, stabbing right upper quadrant and lower abdominal pain which radiated to the back for four months, associated with nausea and vomiting. He had decreased appetite and a 15-pound weight loss for the past two months. He had a long-standing history of alcohol abuse but none in the past two months. There was no history of lye ingestion. On examination there was no jaundice. The abdomen was distended with right upper quadrant tenderness and guarding. There was ascites and diminished bowel sounds. There was no edema. A nasogastric tube was placed for small bowel obstruction. Paracentesis yielded 5 liters of fluid. Cytology suggested mesothelial proliferation versus metastatic carcinoma. A virtual computed tomography/colonoscopy revealed moderate amount of ascites with nodularity of the mesentery and possible carcinomatosis. Endoscopy showed diffuse moderate inflammation of the gastric body characterized by erythema, friability, and granularity. Cold forceps biopsies showed invasive poorly differentiated adenocarcinoma with focal signet ring cell differentiation. Gastric biopsy was positive for linitis plastica. There was no evidence of Helicobacter pylori. He was evaluated by hematology oncology. He received dexamethasone and palonosetron for severe nausea and vomiting. He elected not to undergo any further treatment and was discharged on hospice care.

Approximately 21,600 cases of gastric cancer are diagnosed annually in the United States. Most patients are symptomatic but have advanced incurable disease at time of presentation. Weight loss and persistent abdominal pain are the most common initial symptoms. Patients may also complain of nausea or early satiety. Linitis plastica is a morphological variant of diffuse (or infiltrating) stomach cancer. It can occur from lye ingestion or metastatic infiltration of the stomach, particularly from breast and lung carcinoma. The risk factors are undefined, except for rare inherited mutations in E-cadherin, which are found in about 50% of diffuse-type gastric carcinomas. They spread by direct extension through the gastric wall to perigastric tissue occasionally adhering to adjacent organs such as pancreas, colon, or liver. These tumors tend to infiltrate the submucosa and muscularis propria yielding negative superficial mucosal biopsies. Diagnostic esophagogastroduodenoscopy (EGD) is usually preferred over barium studies. However, a barium swallow may be superior to EGD in linitis plastica, showing decreased distensibility of the stiff, “leather-flask” appearing stomach. Complete surgical removal of the tumor with resection of adjacent lymph nodes offers the only chance of cure, however, is possible in less than one third of patients. A subtotal gastrectomy is the treatment of choice for patients with distal carcinomas, while total or near-total gastrectomies are required for more proximal tumors. Combination chemotherapy administered before and after surgery as well as post-operative chemotherapy combined with radiation reduces recurrence rate and prolongs survival.
Myopericarditis is an underdiagnosed cause of fulminant heart failure, conduction abnormalities, malignant arrhythmias, sudden cardiac death and chronic dilated cardiomyopathy. The true incidence of the disease is not known owing to its heterogeneous presentation and the lack of universally accepted and standardized diagnostic criteria. It may manifest as chest pain with regional ST-segment changes on electrocardiogram, and elevated cardiac biomarkers suggesting myocardial ischemia. We report a case of viral myopericarditis mimicking ST elevation myocardial infarction confirmed by cardiac magnetic resonance imaging. A 19 year old male had symptoms of acute viral upper respiratory infection, with fever, cough, fatigue, sore throat, poor appetite and watery diarrhea for one week. He presented with acute onset retrosternal chest pain associated with dizziness and exertional dyspnea. An electrocardiogram (ECG) showed ST-segment elevation in inferolateral leads. Chest X-ray was unremarkable. Serum troponin I was 7.660 ng/ml (normal < 0.779 ng/ml) and CKMB was 16.90 ng/ml (normal <5 ng/ml). Trans-thoracic echocardiography showed mildly impaired left ventricular ejection fraction of 54% with no significant pericardial effusion. Due to ongoing chest pain and rising cardiac biomarkers, coronary angiography was performed the following day which was normal. Cardiac magnetic resonance imaging (CMR) with intravenous injection of gadolinium showed delayed enhancement in left ventricular apex confirming the diagnosis of viral myopericarditis .Viral serological analysis detected IgG antibodies against Coxsackie virus, parvovirus, Epstein bar virus and herpes-simplex virus I and II. His symptoms resolved after receiving supportive treatment and he was discharged four days later. Follow-up echocardiography and ECG were to be repeated in 3 months. Myopericarditis in young patients with suspected myocardial infarction may be more common than previously thought. Complex pathogenesis includes direct viral myocardial damage as well as autoimmune reactions against cardiac epitopes. The diagnosis is aided by clinical presentation, physical examination, laboratory testing, electrocardiographic changes, and the lack of epicardial coronary disease revealed by cardiac angiography. Echocardiography serves to evaluate regional or global left ventricular dysfunction in such cases and to rule out other causes of heart failure, such as valvular, congenital or amyloid heart disease. Although endomyocardial biopsy is the diagnostic gold standard, it has a low sensitivity and considerable periprocedural morbidity and mortality. Thus, cardiac magnetic resonance imaging (CMR) is emerging as an important tool for not only diagnosing myopericarditis but also in predicting the extent of myocardial injuries with delayed contrast enhancement. Further, it can guide tissue sampling of an endomyocardial biopsy to increase diagnostic yield.
Painless Jaundice: An atypical presentation of NASH

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Non-Alcoholic Fatty Liver disease (NAFLD) is a spectrum of disorders, ranging from simple fatty liver to nonalcoholic steatohepatitis (NASH) and NAFLD-associated cirrhosis. It is a diagnosis of exclusion. It was originally described in obese females with diabetes, but can occur in normal weight people of both genders. Although its presentation is mostly asymptomatic, it is an important diagnosis to make in order to prevent liver cirrhosis. Cholestasis is not classically associated with NASH, hence our interest in reporting this unusual case. A 61-year old obese female with past history of hypertension, dyslipidemia and diabetes, presented with one day onset of painless jaundice. She denied experiencing abdominal pain, pruritus, fevers, chills, fatigue, malaise, or weight loss. Physical examination revealed scleral icterus and hepatomegaly with a liver span of 25 cm, but no abdominal pain or tenderness. There were no signs of cirrhosis. She denied alcohol luse and was not taking any known hepatotoxic medications. Computed tomography of the abdomen revealed mild fatty liver and cholelithiasis without evidence of cholecystitis. Direct bilirubin was markedly elevated at 19.5 mg/dL, total bilirubin elevated at 23.0 mg/dL, alkaline phosphatase elevated at 628 U/L, gamma glutamyl transpeptidase (GGT) elevated at 1739 U/L, hypoalbuminemia 2.9 mg/dL, and moderate transaminitis with aspartase aminotransferase (AST) of 163 U/L of and alanine aminotransferase (ALT) of 230 U/L. She had a normal prothrombin time at 10.9 seconds, with international normalization ratio of 1.0. Magnetic resonance cholangiopancreatography demonstrated non-dilated intra- and extra-hepatic ducts. The common bile duct was within normal limits and no duct stones were identified. Mild fatty infiltration of the liver was noted. The pancreas appeared to be normal. An endoscopic ultrasound was unremarkable. Serological markers for primary biliary cirrhosis, primary sclerosing cholangitis, auto-immune hepatitis, and acute viral hepatitis were all negative. Liver biopsy showed mild acute and chronic hepatitis with predominantly macrovesicular steatosis, involving approximately 30% of the hepatocytes, with mild cholestasis and ballooning degeneration and nuclear inclusions. These findings were consistent with steatohepatitis, most likely from NASH. NASH typically presents with moderate elevations in AST and ALT and normal bilirubin levels. Cholestasis occurs in setting of cirrhosis when NAFLD is advanced, but our patient did not have cirrhosis. This case is unique in its atypical presentation. The treatment of NASH involves weight loss and management of the metabolic syndrome with aggressive treatment of underlying disorders like diabetes mellitus and dyslipidemia. The goal of treatment is prevention of cirrhosis and liver related death, that can be as high as 20% and 8% in NAFLD patients.
Rectal Mucosal Necrosis in a Patient with a Stool Management System

Ronald Yglesias, Manoj Patel MD, Rimsha Hasan MD, Domick Zampino DO, Nancy Higgins MD

Rectal necrosis due to pressure is a known adverse effect of stool management systems for patients in the intensive care unit (ICU). Commercially available stool management systems claim that rectal necrosis is reduced by using more modern flexible tubes with inflatable cuffs. We present a case of rectal mucosal necrosis after the insertion of a stool management system. A 63-year old male was transferred from a nearby medical center with a severe lower gastrointestinal bleed and hemorrhagic shock. He had complained of intermittent bright red blood per rectum for several months. He had no allergies and was not taking any medications. There was no family history of bleeding disorders. On physical examination he was alert, awake and oriented. Vitals signs: temperature 97.6 degrees Fahrenheit, blood pressure 95/78 mmHg, heart rate was 149 beats per minute, respiratory rate was 16 respirations per minute. Bowel sounds were present without distention. There was mild diffuse discomfort upon palpation of the abdomen. Skin turgor was decreased. He was resuscitated with packed red blood cells and saline and transferred to ICU. He continued to bleed actively with associated dizziness, lightheadedness, tachycardia and blood pressure instability. A stool management system was placed to quantify the amount of bleeding. A bleeding scan revealed active bleeding in the descending colon. He underwent a mesenteric angiogram for possible embolization but no active source for the bleeding was found. Colonoscopy showed mucosal ulceration with necrosis of the entire rectum and diverticulosis but no active source of bleeding. No obvious alternative etiology for the rectal necrosis was found. The stool management system was discontinued. His hemoglobin stabilized without further rectal bleeding. He was subsequently discharged home with recommendations for a follow up colonoscopy. We present a unique case of rectal mucosal ulceration and necrosis likely caused by the associated pressure on the rectal wall secondary to the inflated cuff after insertion of a stool management system. Our patient did not have major complications arising from the placement of this system; however we must stress the importance of the warnings and precautions of this equipment. To our knowledge, no literature has been published describing this rare but definitely life threatening event.
A rare case of rabbit anti-thymocyte globulin induced disseminated intravascular coagulation

Vasanthi Balaraman, Anup Patel M.D, Sunil Sapru M.D

Introduction: Rabbit anti-thymocyte globulin (RATG) is a purified polyclonal immunoglobulin used for induction therapy in renal transplantation. Known adverse effects include cytokine release syndrome, leukopenia, thrombocytopenia and late serum sickness reaction. To date, only a few case reports of RATG induced coagulopathy exist in the literature. We report a case of renal transplant (RT) recipient who developed severe disseminated intravascular coagulation (DIC) and post-operative bleeding after RATG induction therapy. Case presentation: Patient is a 61 year old female with history of end stage renal disease secondary to diabetes mellitus who received a deceased donor renal transplant. Her operative course was uncomplicated. 150 mg of RATG in 500 ml normal saline was initiated intraoperatively which was scheduled to run over 10 hours. Postoperatively, the patient became hemodynamically unstable with excessive bloody output from the surgical drain. She received aggressive fluid and blood product resuscitation. Exploration of the RT revealed diffuse oozing but no evidence of bleeding from the arterial, venous anastomosis, renal hilum or ureteric mesentery. A coagulation profile revealed an elevated prothrombin time of 27.0 sec, thrombin time of 36.4 sec, INR of 2.5 and d-dimer more than 5250 ng/ml and reduced fibrinogen level of 135 mg/dl and platelet count of 66000/cmm consistent with DIC. Her preoperative coagulation profile was within normal limits. Despite aggressive transfusion, patient continued to have persistent bloody output from the surgical drain. A reexploration revealed an intact anastomosis but persistent diffuse oozing which necessitated packing the transplant bed with gauze and administration of additional blood products. Subsequently, the patient's hemodynamic status improved and output from the surgical drain decreased. Her coagulation parameters showed improvement. The abdominal packs were removed with improvement in the appearance of the operative field. Discussion: Although the inciting cause of DIC is not entirely certain in this case, we suspect it may be secondary to RATG. The only significant medications the patient received were RATG, mycophenolate and methylprednisolone. Only a few case reports of RATG induced coagulopathy exist in the literature. Two cases of coagulopathy with equine ATA therapy which is no longer used in RT were reported previously. Another study reported a series of twelve hematopoietic stem cell transplant patients having an increase in D-dimer, tissue factor, thrombin-antithrombin III complex, and thrombomodulin after RATG therapy. None of the patients had bleeding or thromboembolic complications which was attributed to non-overt DIC. Our patient had DIC with severe post-operative bleeding as a consequence. The precise mechanism of this observation remains to be understood. We contemplate that DIC is a rare but potential life threatening complication of RATG induction and an awareness of this potential complication is essential.
Barnabas Health - St. Barnabas Medical Center

Bath salts induced Myocarditis

MOHSIN HASEEB, Saba Farooq, Anjum Tanwir, Sunil Sapru, Mindy Houng

A 20 year old man with a history of marijuana abuse presented to the ER with generalized abdominal pain, vomiting and diarrhea and shortness of breath for one day. The patient had a heart rate of 120 BPM and a blood pressure of 93/75 mm Hg. Bilateral inspiratory crackles and a loud S3 gallop were present. A 12-lead electrocardiogram demonstrated bidirectional wide complex tachycardia. Subsequent EKGs showed right bundle-branch block followed by a left bundle-branch block type pattern. The patient’s Troponin-T level was also elevated at 22.09. His creatine phosphokinase level was 741 IU/L. Urine drug screen was positive for cannabis. Emergent bedside transthoracic echocardiography revealed a left ventricular ejection fraction of 35-40% with severe right ventricular dilation and hypokinesis. Coronary angiography demonstrated no obstructive coronary artery disease. Patient’s ventricular tachycardia did not respond to IV amiodarone and Lidocaine. After approximately 2 hours into the care episode, the patient’s HR increased to 160 BPM with marginal hemodynamics. Multiple attempts of defibrillation were made without any success. The patient sustained a cardiac arrest and was resuscitated with ACLS protocol. The patient had a second cardiac arrest and died within 6 hours of presentation. An autopsy confirmed myocarditis and bath salt intoxication. “Bubbles”, “Bonzai fertilizer”, “White Surge” and “Cloud 9” are the street names for a synthetic stimulant called methylenedioxyxpyrovalerone (MDPV). MDPV is marketed as a “bath salt” that is widely available in convenience stores and smoke shops. MDPV is one member of a family of over 25 chemicals that inhibit norepinephrine-dopamine reuptake and thus act as central nervous system stimulants. MDPV is related in chemical structure to cathinone, an active alkaloid found in the khat plant, methamphetamine, 3, 4-methylenedioxymethamphetamine, and other schedule I phenethylamines. The most common route of administration is nasal insufflation (snorting), but drug can be ingested, inhaled, or injected. There are various hazardous side effects reported with its use including cardiovascular, respiratory and neurological. Sympathomimetic effects are noticed due to the surge in catecholamine levels. The typical neurological findings include agitation, paranoid delusions, hallucinations and self-injurious behaviors. Our case outlines the cardiovascular manifestations of MDPV abuse. Usual complaints associated with MDPV abuse. There is currently no way to routinely test for these substances. These substances are not detected in routine drug testing. They can be detected in urine and hair analyses using gas chromatography-mass spectrometry. The diagnosis is usually made on the basis of a history of abuse and symptoms and signs of a sympathomimetic syndrome. Intoxication with bath salts should be considered in any case of myocarditis with unclear etiology.
Concomitant C.Diff and CMV Infection in Immunocompromised Patients

Komal Hussain, Nida Habib, Navdeep Dhillon

INTRODUCTION: As the incidence of Clostridium difficile (C.diff) continues to rise over the past decade, physicians are encountering increasingly varied and often unique presentations of the illness. Although, most commonly C.diff is associated with an elevated white blood cell count, there is evidence in the literature that suggests C.diff can also present with leukopenia. One of the circumstances where this is seen is with a co-infection CMV in immunocompromised patients and has been shown to be a poor prognostic factor. CASE REPORT: We present the case of a 47 year old lady with past medical history significant for lupus nephritis, pulmonary embolism and living unrelated renal transplant in 2008, recipient positive for CMV, currently on tacrolimus, mycophenolic acid and prednisone for immunosuppression. She presented to the hospital with a four week history of persistent post-parandial watery diarrhea. When questioned, she denied any abdominal pain or fevers. Her physical exam was unremarkable and did not demonstrate abdominal tenderness on palpation. In the ED, she was found to have an elevated creatinine (2.26) and pancytopenia. Her white blood cell count was 1.6K/ccm with absolute neutrophil count of 1.2k/ccm, hemoglobin was 8.3 g/dL and platelets were 76 k/cmm. The differential showed 41% bands and 8% lymphocytes. Her baseline white cell count had been in the range of 3.4-41. k/ccm. She was admitted to the hospital for IV hydration and further work-up. Several stool studies were sent including E.Coli 0157:H7, Shiga toxin, Campylobacter, Shigella and Salmonella, which were all determined to be negative. However, Stool C.diff by PCR did come back positive. Therefore, the patient was placed on contact isolation and metronidazole was initiated. The following day, CMV PCR titers returned with >37,000 copies. She was then started on gancyclovir. During her admission, the patient continued to be afebrile and did not complain of abdominal pain. Her diarrhea began to improve gradually with complete resolution in three days. In addition, her leukopenia improved significantly following the administration of gancyclovir. On the other hand, her creatinine continued to increase despite IV hydration during her hospital stay. Kidney biopsy was performed which ruled out transplant rejection but was suggestive of recurrence of lupus nephritis. DISCUSSION: Immunocompromised patient are at a higher risk than the general population to develop infections, especially multiple simultaneous ones. CMV is one such infection that can be concealed by other concomitant infections such as C.diff and can often be missed. Therefore, if an immunocompromised patient presents C.diff and has leukopenia, this should warrant further work-up for co-infection.
Ascites as a presenting sign of hypothyroidism is very rare; 16 case reports have appeared in the literature in the past 20 years. We present a case of ascites in a patient with undertreated hypothyroidism that resolved completely with levothyroxine replacement. The patient is a 55 year old African-American woman who presented to the emergency department with recurrent symptomatic ascites requiring repeated paracentesis. Past medical history includes asthma, leiomyoma status post total hysterectomy and bilateral salpingo-oopherectomy and hypothyroidism diagnosed six month ago. She had been taking levothyroxine 125 mcg/day, but discontinued it four weeks prior to the onset of ascites. She reported progressive abdominal distention, but no abdominal pain, change in bowel movements, nausea or vomiting. On admission there was no evidence of myxedema; TSH was 51 UIU/mL and free T4 was 0.21 ng/dL. The etiology of the ascites was unclear. The patient underwent paracentesis four times; 1.4 to 8 liters was removed at each procedure. The ascitic fluid was clear and yellow, with elevated total peritoneal fluid protein ranging from 4.1-4.9 mcg/dL; the serum-ascites albumin gradient was 0.8 g/dL. Cytology and culture of the ascitic fluid were negative for malignancy and infection. Liver function tests, hepatitis B and C serology, HIV and RPR testing, carcinoembryonic antigen and alfa-fetoprotein were within normal limits. A CT scan of the abdomen and pelvis with intravenous and oral contrast did not reveal any structural abnormalities. Erythrocyte sedimentation rate was slightly elevated at 19 mm/hr. CA 125 was elevated at 41 U/Ml, but has been stable for more than six months. Diagnostic laparoscopy was performed and liver biopsy demonstrated mild macrovesicular steatosis, mild sinusoidal dilatation, mild hemosiderosis and no fibrosis. ECG and MUGA scan were unremarkable. The patient was treated with levothyroxine 125 mcg/day and three days of 20 mg/day of oral furosemide during the admission and was discharged home with levothyroxine 125 mcg/day. Follow up abdominal ultrasounds at 2.5 months and 6 months revealed no ascites. In conclusion, ascites associated with hypothyroidism is a rare but reversible condition; thyroid hormone replacement is the definitive treatment.
A case of chronic autoimmune vestibulopathy improved with steroids

Li Jiang, Sapru Sunil, Wen Xiong

The most common causes of vestibulopathy are ototoxic aminoglycosides, Meniere’s disease and meningitis. The diagnosis of autoimmune vestibulopathy is rare. We present a case of autoimmune vestibulopathy in a 31 year old African-American woman with severe intermittent chronic vertigo and dizziness for fourteen months that improved with the treatment of steroids. The dizziness and vertigo affected her daily life and work. Symptoms were sometimes associated with headache and nausea but not with hearing loss, weakness, numbness or tingling. There was no history of intravenous antibiotic use or head trauma. Past medical history included Vitamin D deficiency and iron deficient anemia.

Physical exam and routine labs such as complete blood count with differential and comprehensive metabolic panel were unrevealing. Treatment was initiated with meclizine and vestibular physical therapy without much improvement. Dizziness improved briefly but recurred one week after she was started on topiramate 15mg bid. The patient was then referred to neurology and rheumatology for further evaluation and treatment. Rheumatoid factor, cyclic citrullinated peptides, anti-neutrophil cytoplasmic antibodies, antiphospholipid antibodies, beta 2-glycoprotein-antibody, anti-jo-1 antibody and complement C3 were within normal limits; erythrocyte sedimentation rate was elevated at 48 mm/hr, C-reactive protein was elevated at 6.5 mg/L (normal limit is less than 4.9 mg/L), antinuclear antibodies was 1:80, anti-SSA was positive (higher than 8.0 AI) and anti-SSB was negative. Sialogram was unremarkable. MRI of the brain with and without intravenous contrast revealed non-specific nodular hyperintensities in bifrontal and left frontal parietal subcortical white matter and normal bilateral seventh and eighth intracranial nerve complex. Electronystagmogram and rotator chair test confirmed bilateral central vestibular dysfunction and left peripheral vestibular dysfunction.

Autoimmune vestibulopathy was diagnosed and the patient was started on hydroxychloroquine 200mg daily. Methylprednisolone 64 mg per day was added tapering the dose 8 mg every 5 days until 16 mg daily within 4 weeks. She was also placed on calcium and vitamin D. Four weeks after steroids treatment the patient reported significant improvement of dizziness and complete resolution of vertigo. In conclusion, autoimmune vestibulopathy is a relatively rare condition. The diagnosis can be difficult, however a short term treatment trial of steroids may be effective and possibly diagnostic.
Superior Mesenteric Artery Syndrome and Its Associated Gastrointestinal Implications


The superior mesenteric artery (SMA) syndrome is a rare but potentially life-threatening gastrointestinal condition. Over the years, it has been referenced by several names, the most common of which is Wilkie’s syndrome. These numerous terminologies have made it difficult to estimate its true frequency in the general population. Common symptoms associated with this syndrome include intermittent postprandial abdominal pain, nausea, and bilious vomiting. Our review revealed that although it is currently well-defined in the literature, the diagnosis of SMA syndrome remains challenging as other disorders can mimic its presentation. However, CT angiography is currently favored in the literature for diagnosis as it can not only show the narrowed aorto-mesenteric angle and distance, but also the extent of duodenal obstruction. In addition, we found no consensus on the preferred mode of therapy once SMA syndrome is diagnosed. The agreement among authors is that the treatment options should be based on severity of the disease, using conservative measures as the first line of therapy in mild SMA syndrome. Duodenojejunostomy is the preferred surgical approach when conservative management fails, or in severe cases. Clin Anat. 2013 Aug 20. doi: 10.1002/ca.22249. [Epub ahead of print]. PMID: 23959808
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Fatal Malignant Cerebral Ischemia from Spontaneous Carotid Dissection: Would intervention have helped?

Michael Meadows, Sunil Sapru, MD

Introduction Patients with large space-occupying hemispheric infarctions carry a poor prognosis, with mortality rates of up to 80% secondary to herniation of cerebral or brainstem structures. Evidence has been shown that decompressive hemicraniectomies within two days of onset of symptoms have reduced mortality in these patients. However, controversy surrounds the decision to perform these procedures. Case Report A healthy 56 year-old gentleman with no significant medical history developed frontal headaches two days prior to admission. On the day of admission, the patient’s wife noticed that his left face appeared mildly swollen. When the patient stood from a seated position he fell forward onto the floor. He did not lose consciousness but could not pick himself up off the floor. Emergency medical services were called and he was evaluated in the emergency room. Initial physical findings showed left-sided facial droop, dysarthria, left hemiplegia, left neglect, and impaired sensation on the left side. Computed tomography (CT) of head was negative for bleeding or mass effect and tissue plasminogen activator was administered as per protocol. The patient subsequently underwent a cerebral angiogram where a right-sided carotid dissection and a right middle cerebral artery occlusion were identified. Successful embolectomy was performed and the patient was transported to the neurological intensive care unit for monitoring. Post embolectomy, he underwent serial head imaging over the course of several days which showed a stable 3.8-mm midline shift. Neurovascular checks during this period remained unchanged. Discussions were held between the family and neurosurgeons about prophylactic decompression hemicraniectomy but it was decided to treat him medically. Six days after admission, the patient became unresponsive and began having seizure activity involving his right limbs. The seizures were refractive to intravenous administration of levetiracetam, fosphenytoin, and ativan. Midazolam infusion was initiated and he was intubated for airway protection. He continued to have breakthrough seizures albeit less frequently. A repeat CT head showed an increase in midline shift from 3.8-mm to 11-mm with no hemorrhagic transformation. A decompressive hemicraniectomy was again contemplated but it was ultimately decided to treat with hypertonic saline infusion. His condition continued to deteriorate; he was placed under hospice care soon thereafter, and subsequently died two days later. Discussion A meta-analysis of randomized clinical trials has shown reduced mortality in patients who have surgical decompression within 48 hours of stroke onset with malignant cerebral ischemia and severe swelling. However, functional outcome does not improve when decompression is delayed up to 96 hours after initial symptoms. The decision to perform this potentially life-saving procedure, despite evidence of benefit, continues to be dependent on surgeon and family preferences with regards to perceived long term functional outcome and quality of life.
NSTEMI in TTP, a curious occurrence

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NSTEMI in TTP, a curious occurrence  Adrien Melikian, MD, Daivesh Patel MD, Mindy Houng, MD  Saint Barnabas Medical Center, Livingston, NJ  Thrombotic Thrombocytopenic Purpura (TTP), a thrombotic microangiopathy, causes occlusions of microvasculature commonly leading to neurological and renal manifestations; however, cardiac ischemia is not frequently associated with TTP. We describe a patient presenting with non-ST elevation myocardial infarction, found to be with acute TTP.  A 73 year-old African-American female arrived in the Emergency Department complaining of chest pain. She had been on aspirin and clopidogrel and had a recent normal coronary angiogram for a similar presentation. She appeared comfortable, mildly jaundiced, and her chest pain was reproducible. Her hemoglobin was 7.6, platelets 18,000, white blood cell count 8.5 with bands 14%, reticulocyte count 3.1, total bilirubin 3.2, direct bilirubin 0.6, and leukocyte dehydrogenase 965. Haptoglobin was undetectable. Her EKG was unchanged from prior admissions, with no ST segment or T wave changes; however, her troponin T was 0.205 which peaked 24 hours later at 0.768. The patient was admitted to the cardiac care unit, where upon arrival she developed confusion, slurred speech, and a hemifacial droop which lasted a mere ten minutes. Computed tomography of the head revealed no acute pathology. She was started on high dose methylprednisolone and underwent urgent plasmapheresis which improved her hemolysis and immediately improved her chest pain. After one week of twice daily plasmapheresis, her TTP resolved. She experienced no more transient neurologic abnormalities. Her cardiac enzymes trended down and a subsequent echo revealed no wall motion abnormalities. Her ADAMTS13 level drawn upon admission returned very low, less than two. Overview of the literature describes acute myocardial infarction as a relatively uncommon occurrence in TTP and etiologies suggested range from an autoimmune and inflammatory effect to thrombosis of the coronary microvasculature and rarely, macrovasculature. Prompt diagnosis and treatment of TTP causing MI is imperative. Plasma exchange is the treatment of choice to halt and reverse the responsible pathological process and improve clinical outcomes.
**Barnabas Health - St. Barnabas Medical Center**

**Intracranial hemorrhage with Prasugrel use in a patient treated with induced hypothermia for anoxic encephalopathy**

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**Background:** Prasugrel is known to increase risk of intracranial hemorrhage in patients with previous history of ischemic stroke. There is however no current data of such an increase in risk in patients who have history of anoxic encephalopathy resulting from events such as a ventricular arrhythmia with cardiac arrest who had gone to receive therapeutic hypothermia protocol for anoxic encephalopathy as with our patient. Case Presentation: 69-years-old gentleman with no past medical history who earlier on the day of presentation reported experiencing chest pain and diaphoresis and subsequently presented to Emergency Room after having sustained an out-of-hospital ventricular arrhythmia with cardiac arrest. He was unresponsive on arrival to ER. ER work-up demonstrated an anterior wall ST segment elevation myocardial infarction. Prior to departing for the angiography he also underwent a Computed Tomography scan of head, which was negative for any intracranial pathology. Cardiac catheterization had demonstrated a 100 percent blockage of proximal left anterior descending artery and a 90 percent occlusion in the middle right coronary artery. Both lesions were treated with drug-eluting stents. Following his cardiac catheterization, he was started on dual anti-platelet agent therapy with aspirin and prasugrel. Additionally, therapeutic hypothermia protocol was initiated as he had suffered an out-of-hospital cardiac arrest with possible resultant anoxic encephalopathy. 16 hours after starting hypothermia protocol patient developed ventricular fibrillation and had cardiac arrest. He was resuscitated as per ACLS protocol and received defibrillation with return of spontaneous circulation briefly after starting resuscitation. Several days following this event our patient was extubated and returned to his baseline mental status. He was subsequently discharged home on a medication regimen which included aspirin 81 mg daily and prasugrel 10 mg daily. He presented to ER again one month after his discharge with severe headache and worsening of vision in left eye. Imaging with CT of the head revealed a right sided occipital lobe hemorrhage with subdural extension and 3mm midline shift. He was admitted to intensive care unit and managed conservatively by the neurosurgical team. He had repeat CT scan of the head, which showed stable, bleed. Patient was started on clopidogrel instead of aspirin and prasugrel. Patient also had CT scan of head after clopidogrel was initiated, which demonstrated stable changes consistent with earlier intracranial bleed. Conclusion: Prasugrel’s known risk of intracranial bleed after ischemic stroke may carry over to patients who have also had global anoxic encephalopathy – specifically in this case following a cardiac arrest. Further studies will help to determine if greater degree of caution should be exercised prior to initiating prasugrel in patients who have such a history of anoxic encephalopathy.

Concomitant pulmonary embolus and ischemic stroke: A rare clinical scenario

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Introduction: Concomitant pulmonary embolus and ischemic stroke is not common. It can be explained by paradoxical embolism, a clinical condition in which emboli originating from the venous system reach the systemic arterial circulation most commonly via a patent foramen ovale (PFO). Case presentation: A 68 year old man with a history of paroxysmal atrial fibrillation and hyperlipidemia presented to the emergency department (ED) with the acute onset of exertional dyspnea. One day earlier he had been hiking. In the ED he was tachypneic, hypoxic (PaO2 of 59 mmHg on 100% oxygen) and was therefore intubated. Chest computed tomography angiography (CTA) revealed multiple large intraluminal filling defects consistent with the presence of large bilateral pulmonary emboli. While in cat scan, the patient developed sudden left-sided weakness and a left facial droop. A head CT scan revealed no hemorrhage. The patient was treated with tissue plasminogen activator (tPA) 100mg IV over two hours. He was transferred to the intensive care unit and started on vasopressors because of hemodynamic instability. Venous dopplers of the lower extremity revealed an acute thrombus in the right common femoral vein. Transesophageal echocardiogram (TEE) did not demonstrate any structural abnormalities. The patient became progressively agitated; a repeat CT scan revealed intracranial hemorrhage involving the right fronto-temporal lobe and right basal ganglia with significant mass effect and extensive subarachnoid hemorrhage with intraventricular extension. The patient expired two days later. Discussion: This patient had bilateral pulmonary emboli and an ischemic stroke. Both events can be explained by a single process (eg, embolism through a PFO or an intrapulmonary shunt). In this case, TTE did not detect any structural cardiac abnormalities. Transesophageal echocardiogram (TEE), a more sensitive tool to detect a PFO, could not be performed for the patient was unstable. It is possible that two independent events occurred concomitantly. The patient did have atrial fibrillation and had discontinued anticoagulant therapy; emboli from the left atrial appendage could have engendered the ischemic stroke and deep vein thrombosis could have caused the pulmonary embolism. The patient met the criteria for treatment with tPA both for his ischemic stroke and PE the dosing for which is tPA 0.9mg/kg over one hour (ie 72mg) and 100mg over two hours respectively. Unfortunately the patient developed a massive intracranial hemorrhage following tPA treatment. While there are existing guidelines for tPA dosing for PE and stroke occurring independently, there are no recommendations for concomitant PE and stroke. Further studies are needed to better understand this rare phenomenon and its optimal treatment.
“BURNett’s syndrome: Hypercalcemic crisis as a result of calcium carbonate ingestion to self-treat heartBURN”

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Introduction: Incidence of Burnett’s syndrome (Milk Alkali Syndrome), as a cause of hypercalcemia in patients treated with Sippy regimen to neutralize gastric acidity, decreased after introduction of H2 receptor antagonists and proton pump inhibitor (PPI). However, it has more recently reemerged in the form of Calcium Alkali Syndrome, caused in part by ingestion of large quantities of calcium carbonate present in over-the-counter antacid and as a calcium supplement to treat or prevent osteoporosis. Hypercalcemic crisis is a rare life-threatening condition caused by severe hypercalcemia leading to renal failure and altered mental status. We present a case of 59 year old gentleman who presented with hypercalcemic crisis with serum calcium level of 18 as a result of calcium carbonate ingestion for self-treatment of peptic disease. Case Report: A 59-year-old male with past medical history of hypertension, reflux disease, dyslipidemia, and left carotid endarterectomy presented with 4-day history of dull aching chest pain, generalized weakness and muscle cramps. His medications included lisinopril, simvastatin, aspirin, and tums (3-4 tablets every few hours for the past 4 days - equivalent to 400 mg of elemental calcium per tablet). He was lethargic and had mild diffuse tenderness on abdominal exam. Initial labs revealed serum calcium of 18.4 mg/dl with ionized calcium of 9.8 mg/dl, serum bicarbonate of 33 mEq/L, BUN of 55 mg/dl with creatinine 2.26 mg/dL. Serum phosphorus, total protein and albumin level were normal. Extensive work up for occult malignancy, hyperparathyroidism, hypervitaminosis D and multiple myeloma was negative. Patient’s serum calcium and bicarbonate levels returned to normal after aggressive fluid resuscitation followed by diuresis, calcitonin and pamidronate. Esophagastroduodenoscopy (EGD) showed reflux esophagitis, and pathology was negative for malignancy. He was instructed to reduce his intake of calcium carbonate and started on a PPI. His calcium level and renal function after one month were normal. Discussion: Hypercalcemia from ingestion of calcium and vitamin D supplements is the third most common cause of hospital admission for hypercalcemia after hyperparathyroidism and hypercalcemia of malignancy. Our diagnosis was based on timing of ingestion of calcium carbonate and exclusion of other causes. Hypercalcemia crisis due to calcium carbonate ingestion should mandate an extensive work up to rule out hyperparathyroidism and occult malignancy while prompting a detailed history and review of medications and laboratory findings for a more definite diagnosis of calcium alkali syndrome. References: 1) Hardt et al. Toxic manifestations following the alkali treatment of peptic ulcer. Arch. Int. Med. 31:171-180,1923 2) Sippy et al. Gastric and duodenal ulcer; medical cure by an efficient removal of gastric juice corrosion. J. Am. Med-Assn. 64:1625-1630,1915 3) Patel AM et al. Got calcium? Welcome to calcium alkali syndrome. J Am Soc Nephrology 2010;21:1440-1443
Acute interstitial pneumonia as an early feature of polymyositis -

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Introduction Most patients of interstitial lung disease (ILD) associated with autoimmune disorders have a chronic indolent course. However, a few case reports have documented acute progression of ILD in patients with polymyositis-dermatomyositis. These cases were mostly resistant to immunosuppressive therapy and associated with a high mortality rate. We present a case of severe, rapidly progressive acute interstitial pneumonia as the initial presentation of polymyositis. Case This is a 55 year old man with no significant medical history. His symptoms started with generalized body aches associated with low grade fevers and dyspnea, without prior history of viral or bacterial infection. Two weeks later, he developed multiple joint pain and swelling, located in the proximal interphalangeal joints, metacarpophalangeal joints, and Wrists, with progressive proximal muscle weakness. An outpatient laboratory study showed an elevated ESR of 54 and abnormal CPK of 7000. Subsequently he was admitted to a local hospital, laboratory workup showed normal complete blood count and basic metabolic panel, abnormal liver function tests, elevated CPK at 25000, positive Anti-nuclear antibody, rheumatoid factor at 71, and elevated anti-CCP greater than 250; anti-Jo-1 was negative. A muscle biopsy of the quadriceps was performed. CT scan of the chest showed bilateral multifocal consolidations and atelectasis. The patient was started on intravenous antibiotics for suspected pneumonia and aggressive hydration for assumptive rhabdomyolysis. Rheumatology consult recommended starting intravenous steroid with methylprednisolone 125mg every 8 hours for underlying rheumatologic condition; the dose was decreased gradually. Patient’s respiratory status worsened rapidly and required intubation and mechanical ventilation. Hypoxemia continued to worsen and he was transferred to a tertiary center for extracorporeal membrane oxygenation (ECMO) therapy. Low dose IV steroids therapy was continued in addition to supportive measurements. Muscle biopsy later reported features suggestive of inflammatory myopathy, and lung biopsy showed features compatible with acute interstitial pneumonia. The patient then received a trial of methylprednisolone 1g intravenously daily for 3 days and intravenous immunoglobulin (IVIG) 2g/kg over 2 days. Plasmapheresis also was initiated but was discontinued shortly due to hemodynamic instability. Patient's condition deteriorated rapidly and he expired, approximately 8 weeks after his initial symptoms. Discussion Most patients with polymyositis initially present with symmetric proximal muscle weakness. We report a case of polymyositis that presented with acute interstitial pneumonia. Our patient had several features consistent with antisynthetase syndrome, a rare subset of polymyositis. ILD associated with polymyositis generally presents with an insidious phase that usually responds to immunosuppressive therapy. Only about 5 cases in the literature have reported acute interstitial pneumonia associated with polymyositis/dermatomyositis. Majority of patients died of the acute phase of the disease within 2 months, the rest had significant mortality and morbidity later because of progressive pulmonary fibrosis. One patient reportedly survived with cyclophosphamide/cyclosporine combination therapy.
Chronic Neutrophilic Leukemia: A Case Report

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Chronic Neutrophilic Leukemia: A Case Report James Orsini MD, Shyam Ravisankar MD, Sari Jacoby MD, Amit Gokhale MD  Abstract: Introduction: Chronic neutrophilic leukemia (CNL) is a rare BCR/ABL negative myeloproliferative disorder characterized by persistent neutrophilia and splenomegaly. It is a rare disease, with only about 33 case reports being identified that appeared to have met the diagnostic criteria. It is often a difficult diagnosis to make based on current WHO guidelines however it is an important diagnosis to make as mean survival time is only 21 months. Newer therapeutic targets have been identified and these may help improve clinical outcomes. Objective: Our objectives are 1. To detail the clinical case of a 49 year old female school teacher who presented with leukocytosis, arthralgias, and fatigue. 2. To review the current diagnostic criteria of CNL and newer molecular testing. 3. To review current and future treatment strategies of CNL and 4. To briefly review the current literature on CNL. Case Presentation: KS is a 49 year lady with hypothyroidism who presented with leukocytosis, fatigue, and arthralgias. A full workup of her neutrophilia was performed eventually leading to a diagnosis of Chronic neutrophilic Leukemia based on the current guidelines from the WHO. She was treated with Hydroxyurea and then switched to Interferon because of poor hematological response. CSF3R gene mutation was tested in the hope that if it was positive, we could enroll her on a clinical trial for Ruxolitinib or Dasatinib but the test was negative. She has shown good hematological and clinical response so far. Interferon therapy is being continued and she is being followed closely every two weeks. Discussion: The term “true” CNL introduced by Reilly highlights the need for more experience and dissection of “true” CNL cases to aid in the development of diagnostic criteria, we examined a single case of a newly diagnosed patient with CNL and reviewed current diagnostic criteria, treatment approaches, and outcomes. We compared this case to the cases currently in the literature we explored newer diagnostic tools including use of CSF3R gene and its impact on the diagnosis and possible treatment of CNL. Conclusion: CNL is a rare but fatal BCR/ABL negative myeloproliferative disorder with such few cases currently in the literature it is paramount that each case be presented in current medical literature to assist in development of new diagnostic and possible treatments strategies.
A case of cryptogenic hemoptysis

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In about 30% of patients with hemoptysis, a cause cannot be found even after careful evaluation by chest x-ray, computed tomography (CT), bronchoscopy and other modalities. The term cryptogenic hemoptysis is often used in these cases. There is a significant incidence of malignancy in these patients and they often present a diagnostic challenge. Bronchial artery embolization is considered the gold standard for treatment. A 67 year old woman with chronic obstructive pulmonary disease (COPD), chronic kidney disease, and coronary artery disease on aspirin; presented with moderate hemoptysis. Physical examination was unremarkable and initial hemoglobin was 12.6 gm/dL. CT scan of the chest showed mild emphysematous changes and a 3 mm right middle lobe nodule. Bronchoscopy revealed mucosal edema of the lateral segment of the right middle lobe with minimal active bleeding which was treated with local epinephrine. She had recurrent bleeding during the hospital stay. Aspirin was held; she was treated with platelet transfusions, DDAVP and epsilon-aminocaproic acid and discharged home. She returned after two days with a bout of massive hemoptysis at which time she was intubated, treated with DDAVP, platelet and FFP transfusions. Emergent bronchoscopy showed active bleeding from the right middle and lower lobes. Angiography revealed extensive hypervascularity as well as irregularity and tortuosity of the branches of the bronchial artery overlying the medial aspect of the right lower lobe. Bronchial artery embolization was successfully completed and the bleeding stopped. She also received empiric antibiotic therapy in view of new infiltrates seen on a repeat CT scan of the chest. Post extubation, she developed a stridor and needed a repeat bronchoscopy which revealed a subglottic stenosis from inspissated secretions that responded promptly to suctioning and steroid therapy. Patient improved clinically after the embolization and made a complete recovery. The work up for cardiac causes, bleeding disorders and vasculitis was all negative. A comprehensive evaluation of hemoptysis includes detailed history and physical examination, chest X-ray, CT scan, laboratory assessment for bleeding and coagulation disorders. Common risk factors for cryptogenic hemoptysis include smoking and the presence of COPD. Recent study by Hearth and colleagues demonstrated a substantial incidence (6%) of bronchial carcinoma in patients with cryptogenic hemoptysis. A focused work up for infection, neoplasm and cardiac valve abnormalities is required. A combination of bronchoscopy and high resolution computed tomography (HRCT) is considered the optimal work up to obtain the highest yield for cryptogenic hemoptysis. Bronchial artery embolization (BAE) is the treatment of choice with 77%-98% success rate. This case illustrates the difficult diagnostic and therapeutic challenge involved in managing a patient with cryptogenic hemoptysis and underscores the utility of bronchial artery embolization in successful treatment.
Diabetes Outcomes following establishment of a Diabetic Education Program in an Internal Medicine Faculty Practice

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Introduction: Type 2 diabetes mellitus is an increasingly prevalent disease and is associated with substantial morbidity, mortality and health care costs. The purpose of our study was to evaluate and compare the outcomes in meeting diabetic evidenced-based treatment goals in diabetic populations managed by the standard resident/attending care team to those managed by multidisciplinary team as well as comparing both to national benchmark data provided by the National Committee of Quality Assurance (NCQA) State of Health Care Quality Report (SHCQR). Methods: A retrospective analysis of the diabetic patient population seen at the internal medicine faculty practice (IMFP) was conducted between January 1, 2012 to December 31, 2012, one year after the practice became a recognized NCQA Level 3 Patient Centered Medical Home (PCMH). Patients were assigned one of two groups: The control group patients were managed by the IMFP residents/ faculty physicians. Those managed by a multidisciplinary team (clinical pharmacist/certified diabetic educator and the IMFP residents/ faculty physicians) were assigned to the experimental group. A total of 60 patients were evaluated with 30 patients assigned to each group. The primary objective was measurement of glycemic control defined as having a glycosylated hemoglobin A1C (HbA1C) value less than seven percent. Secondary outcomes identified were having a HbA1C value less than eight percent, low density lipoprotein (LDL) control defined as less than 100 gm/dL and blood pressure (BP) control defined as less than 140/80 mmHg. All groups were compared to the results of the 2012 NCQA’s SHCQR. A Fishers Exact test was used for statistical analysis. Results: Control group demonstrated statistically significant superiority compared to the goals set by SHCQR with 80 percent of patients attaining a HbA1C level less than eight percent (p = 0.02). However the control group did not reach statistical significant superiority in percentage of patients attaining a HbA1C less than seven percent (p = 0.12), BP control (p = 0.7) and LDL control (p = 0.17). The experimental group surpassed the goals set forth by the SHCQR and demonstrated a statistically significant superiority with 72 percent of patients attaining a HbA1C less than seven percent (p < 0.01), 80 percent of patients attaining a HbA1C less than eight percent and 76 percent of patients reaching LDL control (p = 0.02). Experimental group did not show statistical significant superiority with respect to BP control (p = 0.15) as compared with SHCQR. Conclusion: The internal medicine faculty practice met the evidence based treatment goals for diabetes as identified in NCQA’s State of Health Care Quality Report (SHCQR). However, a multidisciplinary care approach surpassed national diabetic care performance measure outcomes as defined within our primary and secondary objectives.
A SEIZURE THAT WAS NOT A SEIZURE – PROLONGED QT WITH ATRIAL TACHYARRHYTHMIA

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Introduction: Congenital long QT syndrome (LQTS) is a hereditary cardiac disease that manifests as prolonged QT interval and life threatening ventricular arrhythmias in relatively healthy young individuals. The most severe and rare variant, Jervell and Lange-Nielsen syndrome, is associated with congenital deafness. We report a patient with Jervell and Lange-Nielsen syndrome who had multiple episodes of atrial tachyarrhythmia and was initially thought to have seizures. Case Report: A 25-year-old African American woman, with bilateral hearing loss since birth, presented to the ER after a cardio-respiratory arrest at home which required AED delivered shocks. Six months prior, the patient had presented with transient loss of consciousness and after extensive workup was diagnosed with possible seizures. On evaluation in the ER, she was alert, in no distress and physical examination was unremarkable except for tachycardia. Electrocardiogram showed sinus tachycardia at 125 bpm with QTc interval of 534 msec and incomplete right bundle branch block. One day after admission, she had two episodes of atrial tachycardia with hemodynamic instability. Repeat electrocardiogram showed persistent QT prolongation with frequent atrial extrasystoles. Transthoracic echocardiogram and cardiac catheterization did not reveal any abnormalities. A diagnosis of JLN was made, she was started on metoprolol, and an AICD with dual chamber pacemaker was implanted, and she was discharged with a plan for outpatient follow up. Genetic testing for LQTS mutations was negative. Six months later, she was again admitted with reported defibrillator shocks and, on interrogation of the defibrillator, was found to have atrial tachyarrhythmias at rates of about 200 with 1:1 conduction. Metoprolol dose and frequency was increased. Two months later, she had recurrence of atrial tachyarrhythmia and electrophysiological study revealed multiple foci of atrial tachycardia with different cycle lengths arising close to the coronary sinus. Ablation therapy was instituted and she remained in junctional rhythm with no further events. Discussion: JLN and other LQTS syndromes are caused by mutations in the genes (KVLQT1) encoding for potassium and sodium channels. Abnormality in these currents results in prolongation of repolarization with prolonged action potential duration. This prolonged action potential, in the presence of triggering events, characteristically causes the development of a special type of polymorphic ventricular tachycardia called Torsades de pointes. The same ionic channels were found to be implicated in atrial repolarization causing polymorphic atrial tachycardia. In a study, which measured action potential durations in a group of LQTS patients, more than 50% presented as polymorphic AT. Although beta-blocker therapy reduces the risk of cardiac arrest in patients with LQTS, the efficacy is limited in patients with JLN and early AICD placement is recommended. Our patient represents the rare occurrence of clinically significant atrial tachyarrhythmia in a patient with LQTS.
A case of heparin-induced skin necrosis detected at early stage with severe pain

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INTRODUCTION: Heparin-induced skin necrosis is a rare complication of heparin use mostly seen in mid-aged women. It occurs at injection sites and points distal, and is often associated with heparin-induced thrombocytopenia (HIT). Thirty-nine cases have been reported in the literature. Although its underlying mechanism is not fully understood, it often owes to an intravascular thrombus composed of heparin-antibody coated platelets or to vasculitis caused by a type III hypersensitivity reaction. In most cases, skin necrosis occurs five to eleven days after initiation of heparin therapy. It may present shortly after heparin use if a patient has been previously sensitized, or very rarely, several months after treatment. The skin lesion may begin as a painful but nonspecific erythematous plaque before sharply demarcating necrosis develops. Delayed recognition of skin necrosis may necessitate future debridement and precipitate life-threatening complications in other organ systems. CASE: We report a case of a 57 years old woman with hemodialysis dependent end stage renal disease. She developed painful lesions of her thighs, lower abdomen, and left arm five days after right femoral bypass surgery. Unfractionated heparin had been injected subcutaneously to the periumbilical region before and after surgery for the purpose of deep vein thrombosis prophylaxis. She also received heparin intraoperatively during bypass surgery. Pain at the medial left thigh and lateral left arm was associated with irregular dark red plaques, measuring 7 cm X 10 cm and 5 cm X 7 cm respectively. No skin changes were noted at the lower abdomen or lateral right thigh. The pain was sharp, constant and intense and exacerbated by pressure. There were also similar painful but without skin lesion. The patient had no fever. Laboratory studies showed stable platelets at 187,000/µl, PT 13.4s, INR 1.2, and PTT 30s. A punch skin biopsy of the left thigh was performed. Analysis revealed a single blood vessel with a fibrin thrombus and ischemic changes in the surrounding tissue. Heparin was discontinued and the patient was treated with aspirin. No further skin necrosis developed and the pain was relieved. DISCUSSION: This case describes the early presentation of heparin-induced skin necrosis, with severe pain and nonspecific skin lesions. Rapid discontinuation of heparin may lead to prompt recovery and possibly avoid skin necrosis. Clinical diagnosis is easier once the the typical demarcating necrosis occurs. There should be a low threshold for considering heparin induced skin necrosis should be applied to middle age women with history of thrombosis and heparin use, presenting with acute onset painful maculopapular lesion.
INTRODUCTION  Myocarditis poses a diagnostic challenge given that its presentation is highly variable, ranging from sub clinical to severe cardiac failure and complex arrhythmias. We report a case of myocarditis in a previously healthy man who presented with ventricular tachycardia and syncope.

CASE REPORT  An active 61 year old male of Italian descent with hyperlipidemia, 20 pack year smoking, and daily alcohol use, presented with a syncopal event at home while running on his treadmill. Evaluation by EMT revealed a monomorphic ventricular tachycardia at 220bpm and he was cardioverted. Physical examination was unremarkable except for facial contusions and forearm lacerations. Cardiac enzymes peaked at Troponin T of 1.760 and CKMB of 42. ECG showed incomplete right bundle branch block with ST depressions in inferior and anterolateral leads. Echocardiogram showed normal biventricular size and wall thickness with normal systolic and diastolic function. Urine toxicology screen was negative. Chest CT was unremarkable. Coronary Angiography showed normal coronaries. Cardiac MRI showed focal fibrosis/necrosis in the lateral wall of the left ventricle with no suggestion of Arrhythmogenic Right Ventricular Dysplasia. Electrophysiologic study revealed reproducible induction of rapid ventricular tachycardia that terminated with overdrive pacing and was consistent with reentry. A detailed work up for possible etiology of myocarditis was completed. Recent history was negative for any viral illnesses, bug bites, rashes or known sick contacts. Inflammatory markers as well as serologies for EBV, HHV6, HSV, CMV, Coxsackie, Coxiella, Lyme, HIV, Parvo B19, and leptospira were all inconclusive. He was treated with amiodarone and ICD placement and remained asymptomatic since then.

DISCUSSION  The most common etiology of myocarditis is viral infections or post-viral immune reactions. Diagnosis is challenging because the presenting symptoms, ECG, Echocardiogram, and cardiac biomarkers may all be non specific. Cardiac MRI is becoming the primary non-invasive tool in investigating myocarditis. Diagnosis is established by using three tissue markers - edema, hyperemia/capillary leak and necrosis/fibrosis; with an accuracy of more than 80% if 2 or more are present. For our patient, abnormal delayed enhancement was consistent with fibrosis/necrosis but T2 images were not available to evaluate edema. While endocardial biopsy remains the gold standard in diagnosing viral myocarditis, it was not performed considering the risk of complications, the possibility of false negative results, and given that a biopsy proven diagnosis would not affect the definitive management. The constellation of symptoms, cardiac MRI findings, and absence of other identifiable etiology was helpful in diagnosing our patient with viral myocarditis.
Two cases of non-falciparum malaria in the same night: how often do you see that?

Abiy Tessema, Tianhua Guo, Anthony Carlino

While the global incidence of malaria is declining, the United States had the highest incidence in more than 40 years in 2011. Of the 1,925 newly diagnosed people, twenty-five became infected in the U.S. Outbreaks of locally acquired malaria in the U.S. have been small and rare, but the potential for the re-emergence of the disease is present due to the abundance of competent vectors (e.g., Anopheles quadrimaculatus). We describe two patients with non-falciparum malaria, both living in the same northern New Jersey city, who presented to our emergency department the same night. Case 1: A 49 year old Guyanese woman presented with fever, chills and rigors occurring every 48 hourly. Additionally she had headache and nausea with vomiting for two weeks. She arrived in the USA from Guyana one month prior. A few weeks before her arrival, she was treated for malaria of unknown type with unspecified medications. In our hospital, blood smear analysis revealed P. vivax with parasitemia of 7%. Serology was positive for non-falciparum malaria. She was treated with atavaquone-proguanil. Twelve hours later, parasitic load improved to 3.9%, and was <0.1% after a day. Fever resolved and she was started on primaquin. Case 2: A 51 year old Nigerian male presented with fever, rigors, headache and vomiting for one week. He was a U.S. resident who returned from a year long stay in Nigeria six months prior. Analysis of thick and thin smears showed Plasmodium sp, consistent with either P. vivax or P. ovale. Serology was positive for non-falciparum Plasmodium infection. PCR analysis done by the CDC identified the organism as P. ovale. Parasitic load was initially 0.8%. He was treated with atavaquone-proguanil; primaquin was not offered owing to glucose-6-phosphate dehydrogenase deficiency. Parasitemia improved to <0.1% after one day; however his fever persisted for three days. Blood and urine cultures were negative; CT of the abdomen and pelvis did not reveal an intra-abdominal abscess. He was treated with chloroquin and his fever subsided. He was discharged with a plan to continue chloroquin for one year. In our hospital, we see a case or two of malaria every few months. The presentation of these two patients in the same night, both living in the same city in New Jersey, initially raised a question about a possible link. Their history of recent travel from different parts of the world and the identification of different organisms in the two cases allowed us to dismiss that possibility. The presence of an infectious condition at a higher than expected rate should always raise a suspicion for an unusual outbreak.
An unusual presentation of Cerebral Toxoplasmosis in a patient with HIV

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Introduction Toxoplasmosis is a zoonotic infection caused by the intracellular protozoan parasite, Toxoplasma Gondii. It is the most common opportunistic infection involving Central Nervous System in patients with AIDS. Head ache, fever, and confusion are the most common presenting features. We report a case with an unusual presentation of cerebral toxoplasmosis in the form of non specific gastrointestinal symptoms in an otherwise healthy individual. Case Presentation A 55 year old Haitian male, with no significant past medical history, initially presented to the emergency room with nausea, vomiting and abdominal discomfort. Physical examination was unremarkable except for mild epigastric tenderness. Serum lipase and transaminases were elevated. CT abdomen and pelvis was negative for any acute pathology. He received symptomatic treatment and was discharged home. Four weeks later, he was admitted with worsening of previous symptoms and 20 lb weight loss. Repeat imaging and laboratory work up were consistent with previous results. One day after admission, he developed severe vomiting and increasing lethargy. Head CT and MRI revealed an aggressive, enhancing mass centered within the deep left frontal lobe with a midline shift, ipsilateral transtentorial herniation, and ventricular effacement. He was started on intravenous steroids and anti-seizure medication. A whole body CT scan ruled out an underlying malignancy. At this point, the patient admitted to having been diagnosed with HIV 20 years ago and having received treatment for 9 years in California. He underwent craniotomy and excision of the frontal mass. Further workup was positive for Toxoplasma Ig G, CD4 count was 33 and HIV viral load was more than 100,000 copies/ml. Brain biopsy showed pseudocysts filled with multiple toxoplasma organisms and focal occlusion of vessels by dense lymphocytic infiltrates. He was initially treated with Bactrim and then switched to a sulfadiazine, pyrimethamine and leukovorin regimen due to acute kidney injury. He showed clinical improvement and HAART therapy was initiated before discharge. Discussion Latent infection with toxoplasma persists for life and the disease reactivates in the immunocompromised host by affecting mainly the central nervous system, presenting most commonly with headache (52%), fever (52%), and confusion (47%). In the absence of prophylaxis, patients with AIDS have an approximately 30% probability of developing reactivated disease. In the late 1990s, a sharp decline seen in the annual hospitalizations was likely related to the advent of HAART therapy and adequate prophylaxis. A considerable slowing down of this decline after 2001 was attributed to the late diagnosis of HIV among Hispanics and African Americans. Inspite of remarkable improvement in the prophylaxis and management of opportunistic infections in HIV patients, it is not uncommon to encounter a case like this. Physicians should maintain a high index of suspicion and patient education is invaluable.
An unusual case of Giant-Cell Myocarditis in a patient with Polymalgia Rheumatica and Myeloproliferative disorder

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Giant-cell myocarditis (GCM) is a rare disease with high mortality from progressive congestive heart failure and refractory ventricular arrhythmia. The association of GCM with autoimmune disorders has been reported but association with hematological malignancy has been very rare. We report an autopsy case of GCM in a patient with longstanding polymalgia rheumatic (PMR) who developed a myeloproliferative disorder. A 59 year old Caucasian female with PMR for six years, presented to the emergency room with two days of fever, chills and diffuse muscle pain. The patient was on oral prednisone, recently tapered to 15 milligrams daily. Prior to admission, she had persistent leukocytosis for one month and had extensive work up with no etiology found. Physical exam was unremarkable except for obesity and mild bilateral ankle edema. Initial laboratory studies showed leukocytosis, low platelet count and elevated LDH. Chest X-Ray showed minimal interstitial infiltration with vascular congestion. A recent echocardiogram had revealed mild left ventricular hypertrophy with enlarged atria. She was empirically treated with vancomycin and cefepime. Four days after admission, the patient was noted to have severe dyspnea with worsening hypoxia which led to elective intubation. Later that night, she developed pulseless electrical activity and could not be rescued. Autopsy revealed a diffuse lymphohistiocytic with multinucleated giant cell infiltration of the myocardium. She was also found to have marked splenomegaly with the white pulp filled with immature myeloid cells. The immature myeloid cells also infiltrated the bone marrow, liver, myocardium, and blood vessels. Immune stains showed positive CD45, CD68, and myeloperoxidase, suggestive of myeloproliferative disorder with monocytic differentiation in blast transformation. GCM is characterized by florid myocarditis with large numbers of multinucleated giant cells. Etiology and pathogenesis is unclear but many cases have been associated with “altered immune states” like thymomas as well as autoimmune diseases. In animal study, immunization with cardiac myosin fraction reproduced the features of GCM. Combined immunosuppression therapy improved GCM patients’ outcomes. No previous case reported of PMR associated with GCM, although it is also an autoimmune disease and is associated with giant cell arteritis. It is of interest to note that the autopsy did show giant cell infiltration in the arteries in our patient. Multiple organs infiltration with immature myeloid cells suggestive of a myeloproliferative disorder is interesting but whether this represents an association remains unclear. There are no previous reports of GCM associated with myeloid neoplasia but two cases showed an association with lymphoma. The multinucleated giant cells in GCM express monocytic markers and appear to be of monocytic and macrophage origin. This case report suggests that a myeloproliferative disorder with multiple organs infiltration, in a patient with altered immune status, may lead to the development of fatal GCM.
RELAPSE OF GRANULOMATOSIS WITH POLYANGIITIS (WEGENER’S) AFTER A 15-YEAR REMISSION

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Introduction: Granulomatosis with polyangiitis (GPA, Wegener’s) is a granulomatous systemic necrotizing vasculitis involving multiple organs. Despite successful treatment, relapses may occur, most within the first 12 to 18 months after the cessation of immunosuppressive therapy. Here we present a patient who was initially treated for upper airway GPA with steroids and cyclophosphamide and achieved complete remission; GPA recurred 15 years later with nasal, pulmonary and renal manifestations. Case: A 64 year old Caucasian male with a history of hypertension, coronary artery disease, bladder cancer, and GPA diagnosed 15 years ago presented with a dry cough and shortness of breath for one week. One month earlier the patient had a bilateral lower extremity rash, biopsy of which revealed leukocytoclastic vasculitis. He was treated with prednisone and the skin rash improved. Renal function was normal at that time. Three weeks later he was seen by an otolaryngologist for epistaxis and nasal endoscopy revealed necrotic tissue with crusted blood. The following day he presented to the emergency department with a fever of 100.4 °F, a pulse of 114 beats/min, respirations of 20/min, blood pressure 98/59 mmHg, and oxygen saturation of 92% on 4 L/min via nasal cannula. Physical exam demonstrated decreased breath sounds with rales at the bases bilaterally. Laboratory data showed a hemoglobin 8.2 g/dL, white blood cell count 8.3x10^3/uL with 20% bands, creatinine 4.45 mg/dL, and a significantly elevated ESR (135 mm/hr) and CRP (27.16 mg/L). Arterial blood gas showed pH 7.41, pCO2 27 mmHg and pO2 61 mmHg on oxygen at 4 L/min via nasal canula. CT of the chest demonstrated extensive bilateral alveolar infiltrates with consolidation most pronounced in the lower lobes bilaterally. He was treated empirically with broad spectrum antibiotics and IV steroids. On day 2, his respiratory status improved but creatinine increased to 4.98 mg/dL. On day 3, a renal biopsy was performed and pathology revealed pauci-immune necrotizing glomerulonephritis with focal global glomerulosclerosis, consistent with GPA (Wegener’s). Proteinase-3 ANCA was positive. The patient was treated with rituximab (375 mg/BSA weekly x 4 doses), plasmapheresis (every other day x 7 exchanges), steroids and hemodialysis. On day 12, his respiratory status was significantly improved and he was saturating well on room air. He was discharged home with continued outpatient plasmapheresis, hemodialysis, rituximab and steroids. On week 8, hemodialysis was discontinued due to improved renal function. Discussion: GPA may recur many years following successful therapy. This case emphasizes the importance of long term follow up of patients with GPA. Physicians must be aware that GPA can recur and that prompt diagnosis of GPA recurrence is important to permit early initiation of therapy that may be life saving and organ sparing.
Discharge Outcomes May Indicate Missed Opportunities For Thrombolysis In Patients With Acute Ischemic Stroke With Mild Or Rapidly Improving Symptoms: Findings From A Community-Based Stroke Center

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Introduction: Active reduction of clot size by thrombolysis is the only FDA-approved treatment for improving neurologic outcome after acute ischemic stroke. However, despite known benefits of thrombolytic therapy, overall rate of use of r-TPA in the USA remains very low. Rapid improvement of stroke symptoms continues to be one of the most common reasons for deferring thrombolysis in patients who present within 3 to 4.5 hours of symptom onset. Our goal in this study was to determine whether these patients might have benefited from thrombolytic therapy. Methods: We retrieved data from the Stroke database of Monmouth Medical Center to assess the frequency, patient characteristics, and discharge outcomes of patients with acute ischemic strokes who presented within 4.5 hours of symptom onset and did not receive thrombolytic therapy due to rapid improvement of stroke symptoms. Results: During the period from January 1, 2008 to July 15, 2012, eighty-four patients met criteria for r-TPA. Forty-four did not receive r-TPA due to mild or rapidly improving symptoms. These 44 patients had a wide range of NIHSS on admission, from 0 to 15, with average NIHSS of 1.5. On discharge, 30 patients (68.18%) were able to be sent home, 13 patients (29.55%) still needed rehabilitation placement, and 1 patient (2.27%) died. Conclusion: Our findings suggest that reluctance to treat mild or rapidly improving stroke is a missed opportunity to improve outcomes. The NIHSS score may be a poor measure of a patient’s functional disability. Despite low NIHSS scores, patients with mild symptoms should still be strongly considered for thrombolytic therapy.
INTRODUCTION: Adult T-cell leukemia-lymphoma (ATL) is an uncommon highly aggressive T-cell lymphoma associated with human T-cell lymphotropic virus type 1 (HTLV-1) infection. It is endemic in Japan and the Caribbean basin. It is rarely encountered during pregnancy and is particularly challenging to treat. CASE PRESENTATION: A 27-year-old female Jamaican immigrant, at 28 weeks of age of gestation, presented with 1-week history of progressive confusion, poor appetite, vomiting and joint pains. During admission, she was noted to have small nodular lesions around the mouth and neck, multiple bilateral tender cervical lymphadenopathy and rib tenderness. Pertinent laboratory findings include leukocytosis, hypercalcemia, hyperuricemia and elevated PTH-related protein levels. HIV antibody test was negative. Chest radiograph revealed diffuse lytic lesions and fractures in the clavicles and ribs. Cautious emergent hemodialysis was instituted to control hypercalcemia. Core biopsy of the largest cervical lymph node revealed intermediate to large peripheral T-cells with immunophenotype classic for HTLV-1. Peripheral blood showed occasional atypical polylobulated T-lymphocytes. Qualitative HTLV-1 DNA test was positive. In order to initiate chemotherapy promptly, the team of specialists decided to deliver the baby via Cesarean section. Betamethasone was not given before delivery due to a high risk of tumor lysis. She gave birth to a baby girl, weighing 1280 grams, with an Apgar score of 1/2/3, who was admitted to the neonatal ICU. MRI of the brain revealed scattered subcortical T2-hyperintense foci and enhancing nodules in the calvarium, consistent with metastatic disease. Bone marrow biopsy did not show evidence of involvement. The patient was started on E-CHOP chemotherapy. She underwent Omaya reservoir placement for intrathecal chemotherapy. She was referred for hematopoietic stem cell transplantation upon completion of six cycles of chemotherapy. DISCUSSION: Treatment of ATL is challenging because of the lack of robust data on optimal chemotherapeutic regimens. Whenever possible, patients with ATL should be encouraged to enroll in clinical trials. ATL is uncommonly seen in pregnancy and it presents a dilemma with regards to prompt institution of chemotherapy to the mother without potentially harming the fetus. Delivery of the fetus when fetal lungs are mature followed by chemotherapy would be the best course of action. However, in cases where fetal lung maturity is not optimal, administration of intravenous steroids prior to delivery is not a reasonable option due to high risk for tumor lysis. A worse scenario is anticipated when ATL presents early in pregnancy when significant fetal prematurity precludes prompt delivery. Furthermore, maternal hypercalcemia and other concomitant metabolic derangements, as well as the HTLV-1 infection may have potential short term and long term adverse effects to the fetus.
"SERENDIPITY IN TESTING THE THYROID"

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The apparently excessive number of Ashkenazi Jewish women who experienced post-partum thyroiditis was previously noted (Freeman, Ruth. Incidence of Thyroid Dysfunction in an Unselected Postpartum Population. Arch Intern Med. 146(7):1361-4. 1986). Working at a clinic serving primarily that population, one of the authors again noted an extraordinary number of patients presenting with that problem. We questioned whether we could distinguish any environmental factors contributing to that apparent excess. After reviewing 35 charts, we found no specific characteristics that would help us define any such factors. We found that 15 patients were diagnosed with a thyroid problem by the obstetrician/gynecologists during pregnancy, 12 after pregnancy and 8 at a time uncertain with respect to the pregnancy. Of the 35 patients, 10 were asymptomatic and presented with abnormal laboratory studies diagnostic of hypothyroidism during pregnancy, 8 had abnormal studies post-partum (6 hypothyroid and 2 hyperthyroid), and 5 were found to have subclinical hypothyroidism at a time unspecified in relation to pregnancy. These patients presented with no symptoms that could be attributed specifically to the thyroid and not to the pregnancy or its subsequent effects. On October 1, 2007, the American Congress of Obstetricians and Gynecologists (ACOG) stated that “Thyroid disease testing is not a routine part of prenatal care and should be limited to women with symptoms of thyroid disease and those with a history of thyroid disease of other medical conditions associated with it such as diabetes.” (http://www.acog.org/About_ACOG/News_Room/News_Releases/2007/Routine_Thyroid_Screening_Not_Recommended_for_Pregnant_Women; ACOG Practice Bulletin Clinical Management Guidelines for Obstetrician-Gynecologists Number 37, August 2002). Our research indicated that monitoring the TSH during and after pregnancy can uncover a significant number of patients with previously undiagnosed thyroid disease who were either asymptomatic or had symptoms which could be ascribed to the pregnancy or post-partum state. We, therefore, believe that TSH levels should be monitored routinely by their obstetricians in all women who are pregnant or in a recently post-partum state.
Successful early carotid endarterectomy for critical carotid artery stenosis following thrombolysis in acute ischemic stroke: A case report

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Introduction We present the case of a 43 year old male who presented with symptoms of acute stroke, was given tPA within the recommended time frame, but was subsequently found to have high grade carotid stenosis and fluctuating symptoms. We follow with a review and discussion of recent literature showing that in select cases, carotid endarterectomy can be done early with no increase in peri-operative complications or adverse events. Case description A 43 year old male with no significant comorbidity presented to our Emergency Room after he was found to be restless in bed by his wife at 12:30am on the day of admission. He was also unable to express himself and was noted to have had a right sided facial droop. He was last observed to be asymptomatic 10:45pm prior to going to bed earlier that evening. The patient did not have any other neurological findings. He had no focal weakness, urinary or bowel incontinence. He did not have any loss of consciousness, seizure like activity or recent trauma. 

1:15 AM Arrival in ER, code stroke called. CT head without contrast revealed no acute intracranial hemorrhage. Initial NIH stroke scale was 5 with aphasia, disorientation, and right-sided facial droop. 

1:45 AM On re-evaluation NIH score improved to 2. 

1:50 AM NIH score of 6 with aphasia and confusion. 

2:10 AM Given fluctuating nature of symptoms and being within window period tPA administered in 3 hours 25 minutes from the onset of symptoms. 

Patient admitted to ICU under stroke protocol with neuro checks and close monitoring of vitals. 

6 AM Patient developed weakness of right upper and lower extremities. His aphasia and facial droop persisted. 

Repeat stat CT head revealed no bleed. 

CT angiogram revealed nearly occlusive thrombus in the left internal carotid artery approximately 2.7 cm distal to the left common carotid bifurcation. An intraluminal thrombus was suspected in the distal left middle cerebral artery segment. 

Advised to undergo early carotid endarterectomy given the fluctuating nature of his symptoms. 

Patient consented after having understood the risk and benefits. 

Patient underwent carotid endarterectomy in 21 hours and 55 minutes of administration of tPA. 

Following carotid endarterectomy, the patient reported improvement of his right sided weakness. 

At the time of discharge he had minimal facial droop and mild aphasia with no motor deficits. 

Discussion Timing of carotid endarterectomy following acute ischemic stroke and administration of tPA has been of debate. In selected candidates with no significant co-morbidities carotid endarterectomy can be done within a 48 hour period with no major complications. Use of peri-operative CT scan will help to select appropriate candidates for early carotid endarterectomy.
OBJECTIVES: We have started an open-access outpatient TIA Rapid Evaluation Center (TREC) at our community medical center. Patients referred to the TREC are seen on the next weekday and undergo a diagnostic evaluation then consultation with a stroke neurologist. The decision to refer a TIA patient to the TREC is left to the discretion of the attending physician. METHODS: We collected prospective data from all TREC patients, including follow-up contact at 3 and 12 months to check for future cerebrovascular events. We performed a retrospective chart review on all patients admitted to the hospital with a primary diagnosis of TIA during the same period (June 2012-June 2013). This study had IRB approval. RESULTS: We saw 74 TREC patients within an average of 1.25 days of referral during its first year of operation, 56 from the emergency room and 18 from physician offices. Only 2 TREC patients required admission to the hospital, the remainder completed their evaluation as out-patients. Only 1 TREC patient had a follow-up cerebrovascular event. During this same time period, 88 patients were admitted to the hospital with a primary diagnosis of TIA. Patients referred to the TREC had lower ABCD2 scores (average of 1.8 vs. 3.8, p <0.001) and were less likely to have a final diagnosis of TIA (19% vs. 77%, p<0.001) than the hospitalized patients. More hospitalized TIA patients were seen on weekdays than weekends (74% vs. 27%). Nearly all patients underwent CT scan, lipid panel and EKG. However, TREC patients were more likely to undergo carotid ultrasound (99% vs. 84%, p=0.001) and MRI of the brain (89% vs. 68%, p=0.001). We estimate that the hospital cost to Medicare is $750 for a TREC evaluation and $5000 for a TIA admission. CONCLUSIONS: 40% of all TIA patients at our hospital were referred to the TREC and seen within 1.25 days during its first year of operation. TIA patients referred to the TREC had lower ABCD2 scores and were less likely to have a final diagnosis of TIA. Day of the week seen did not influence the referral decision. TREC patients were more likely to have a complete diagnostic evaluation at a cost savings of $4250. Out-patient evaluation of TIA can be an efficient, safe, and less costly alternative to hospital admission.
Introduction Heart Failure is the leading cause of hospitalization and readmissions worldwide. It affects quality of life of the patient as well as huge burden on caregiver. In present study, we decided to look for the readmission rates among our patient populations and also look for possible factors which may have contributory role on increasing risk of readmissions. Material and Methods This retrospective cohort study conducted in Monmouth medical Center, in which all patients who were discharged with a principal diagnosis of heart failure were followed for a period of one year. All patients above 18 year old age who were discharged from Monmouth Medical Center with a principal diagnosis of heart failure from January first 2008 till January first 2012 were included in this study. Results: From January 2008 till December 2011, 684 patients [mean age (SD); 76.2 (13.9); age range: 21-103; male: 334 (48.8%)] were admitted to Monmouth Medical Center due to heart failure with a total number of 991 encounters. The admission ratio for each patient was 1:4. Thirty three (4.8%) were readmitted to sister hospitals of Barnabas health system within 30 days and 144 (21.1%) within one year. Fifty-nine patients were started on Telehealth following index admission. 31 patients had just one admission and two patients had two admissions. 29.2% of those who were readmitted had Telehealth compared to 18.2% of those who did not (p=0.19). 97 patients (14.2%) had just one admissions and 30 (4.4%) patients had 2 admissions and the highest number of admissions was 6 which happened just for one patient. 21.1% of those who were readmitted had Telehealth compared to 18.3% of those who did not (p=0.56). Coagulopathy [OR: 2.01, 95% CI: 1.08-3.74, P: 0.02] and Complicated hypertension [OR: 1.90, 95% CI: 1.28-2.82, P: 0.001] were independent factors increasing the one year readmission rate. Discussion: Our preliminary analysis shows that 4.8% and 21.1% of patients were readmitted at least one time within 30 days and one year, respectively. There was no difference among demographic variables in those who get readmitted, however, in regression analysis complicated hypertension and coagulopathy were determined as independent factors increasing risk of readmission within one year. A small portion of present study was on Telehealth home monitoring, although this factor was not included in our regression analysis. These findings need to be investigated with more data to determine the impact of Telehealth home monitoring, as well as intensive heart failure management programs, on admissions in patients with significant risk factors for readmission. This information will help our center to take appropriate steps to avoid, or at least decrease, readmission rate in this vulnerable patient population.
Flushing, Diarrhea, Dyspnea, right sided heart failure; importance of critical thinking in unifying diagnosis

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Case presentation: A 63 year old gentleman presented with progressive shortness of breath, bilateral leg swelling, abdominal distention and weight gain of 30 pounds for 4 weeks. On further questioning, he reported experiencing abdominal pain, diarrhea and flushing for the last two years. His abdominal pain was responsive to omeprazole initially prescribed for dyspepsia. A diagnosis of lactose intolerance was given for diarrhea. Additional symptoms of dyspnea were treated with albuterol for a probable diagnosis of asthma. Past medical history includes hypertension, hypothyroidism, alcohol dependence, “enlarged heart” according to the patient, anemia, gastro-esophageal reflux disease and no past surgical history. Family history was only positive for heart failure. He has been heavy drinker for many years. He denied drug abuse and quit smoking nine years ago. Physical exam was remarkable for respiratory rate of 20; distended jugular vein at the jaw; systolic ejection murmur of 2/6 at the right lower sternal border; abdominal ascites; anasarca edema. Lab works showed normochromic normocytic anemia, negative hepatitis panel, normal PT and liver enzymes but elevated alkaline phosphotase (240). BNP was 225. Patient was initially treated with diuretic as decompensated heart failure. An echocardiogram showed an ejection fraction of 60% with severely dilated right ventricle, thickening of tricuspid leaflets with wide open severe regurgitation, elevated right ventricle systolic pressure. Based on this constellation of findings, carcinoid syndrome was suspected. Further workup, abdominal sonogram, revealed a diffusely heterogeneous liver with multiple hepatic masses, abdominal ascites. More specific studies including serum gastrin, pancreatic polypeptide, chromogranin A and 24 hour urine 5-HIAA were requested with values of 119 pg/ml, 169 pg/ml, 1800 ng/ml, and 130 mg/24 hrs, respectively. Further axial imaging with a CT of chest, abdomen and pelvis with contrast showed a 3.0 cm by 2.5 cm mass with a stellate appearance and central calcification in the appendix region consistent with an appendiceal carcinoid. Additional findings included innumerable hypo- and hyper attenuating masses within the liver. The patient’s symptoms including dyspnea improved significantly with diuresis and initiation of sandostatin therapy, 100 mcg three times a day. Conclusion: Having indolent nature and presenting features similar to other more common diseases such as functional gastrointestinal disorders as well as irritable bowel syndrome obscure diagnosis. On the side of physician, always looking for horses and overlooking zebras could be another reason for late diagnosis of these cases. These remind importance of critical thinking and being creative mindful especially approaching patients who experience multiple, persistent, and disabling physical symptoms to unify their symptoms. This strategy could significantly improve patient care and quality of life and prevent unwanted complications.
Introduction IgA nephropathy is one of the most common causes of glomerulonephritis (GN) worldwide but is rare in African Americans. The clinical presentation is diverse with hematuria being the most common complaint. Crescentic GN is rare, approximately 7% of patients with IgA nephropathy. We are reporting a case of 58 year old African American male who was diagnosed and successfully treated for rapidly progressive GN secondary to crescentic IgA nephropathy. Case Presentation He initially presented in November 2012 with worsening dyspnea on exertion, nose bleeds and hematuria for 3 months. He was hypertensive with blood pressure 217/117 mm of Hg with bibasilar crackles on auscultation. Laboratory data revealed anemia with hemoglobin 9.2 g/dl, creatinine 4.32 mg/dl (1.27 mg/dl in August 2012), and estimated glomerular filtration rate (eGFR) of 17 ml/min/1.73 m2. Urinalysis showed nephrotic range proteinuria and serology testing was positive for anti-myeloperoxidase antibody. In addition, ground glass opacities were seen on computerized tomography of chest. Due to this clinical picture, Granulomatosis with Polyangiitis was a likely differential diagnosis. Hence, renal biopsy was performed to determine the etiology of acute kidney injury. Renal biopsy consisting of 29 glomeruli showed extensive mesangial expansion with hypercellularity, fibroepithelial crescents and mild interstitial fibrosis along with tubular atrophy. Immunofluorescence revealed mild mesangial IgA staining along with mild IgM and C1q staining. Electron microscopy was consistent with numerous mesangial and segmental subendothelial electron dense materials suggestive of IgA nephropathy pattern instead of granulomatosis with polyangiitis. Patient was initially started on an angiotensin converting enzyme inhibitor, intravenous pulse steroid therapy and one dose of intravenous cyclophosphamide. eGFR improved to 31 ml/min/1.73 m2 and hematuria resolved completely prior to discharge from hospital. His renal function gradually improved to eGFR >60 ml/min/1.73 m2 (creatinine 1.14 mg/dl) by February 2013 after two doses of cyclophosphamide and maintenance oral prednisone daily. Unfortunately he also developed idiopathic pulmonary fibrosis, diastolic dysfunction and pulmonary hypertension leading to marked impairment in his functional status. His subsequent admissions to the hospital were complicated with hypoxic respiratory failure and fluid overload requiring higher oxygen support and diuresis. He was eventually transferred to hospice care where he passed away in June 2013. Discussion Recognizing crescentic IgA nephropathy is critical since it can rapidly progress to acute renal failure. Treating crescentic IgA nephropathy still remains challenging. There is only one small prospective study supporting the use of intravenous pulse glucocorticoids and cyclophosphamide (Tumlin, 2003). Larger randomized clinical trials are urgently needed to determine best treatment options for this condition. Other immunosuppressive agents such as rituximab are still in trial or inconclusive so far.
A RARE CASE OF MASSIVE GASTROINTESTINAL BLEEDING AS INITIAL MANIFESTATION OF MULTIPLE MYELOMA.

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Introduction: Plasma cell neoplasms (plasma cell dyscrasias) are a group of entities characterized by the neoplastic proliferation of a single clone of plasma cells, typically producing a monoclonal immunoglobulin. Plasma cell neoplasms can present as a single lesion (solitary plasmacytoma) or as multiple lesions (multiple myeloma). Solitary plasmacytomas most frequently occur in bone (plasmacytoma of bone), but can also be found outside bone in soft tissues (Extramedullary Plasmacytoma EP). EP is seen in 7% of patients presenting with multiple myeloma. Gastric plasmacytomas are uncommon, accounting for only 2% of all EP. Massive upper gastrointestinal bleeding secondary to gastric plasmacytoma is extremely rare, and have been reported in only a few case reports. Case Description: A 54 year old African American female with no past medical history presents to the emergency room with acute onset of altered mental status and upper gastrointestinal bleeding. Laboratory data on admission showed a White blood count of 6.2 K/UL, Hemoglobin 3.0 g/dL, Platelet count of 83 K/UL, and creatinine of 5.87 mg/dL. Patient was hemodynamically unstable and was therefore transferred to Intensive Care Unit for aggressive resuscitation with fluids and blood products. Physical exam revealed melanotic stools and a large palpable mid-abdominal mass. CAT SCAN showed a 12.3 x 11.3 x 8.3 lobular soft tissue mass in the abdomen, a 4.9 cm mass in the lingula of the left lung, retroperitoneal lymphadenopathy, and a destructive lytic lesion in the left iliac bone. On upper endoscopy, an actively bleeding ulcerative mass was visualized. Biopsy of the mass demonstrated monoclonal plasma cell proliferation, consistent with plasma cell neoplasm. Similar histopathology was confirmed on review of the bone marrow biopsy and aspirate. In an attempt to control the bleeding, angiographic embolization of left gastric artery was performed. Bleeding persisted; the patient remained critically unstable and therefore external beam radiation and chemotherapy with VCD regimen (Velcade, Cytoxan, and Decadron) was initiated. The patient remained stable with no further bleeding, and was discharged home with outpatient follow up for continued treatment. Discussion: Multiple Myeloma is a neoplastic proliferation of monoclonal plasma cells leading to bone marrow suppression and end organ damage. EP is plasma cell tumor that arises outside of the bone marrow. Most patients present with symptoms related to the location of the mass. EP should be part of the differential in patients with multiple myeloma presenting with acute gastrointestinal bleeding. EP can be treated with a combination of radiation, chemotherapy, and surgery. One case report showed treatment with VCD resulted in complete remission at 13 months. Understanding the pathogenesis of multiple myeloma spread and drug therapies continue to be an area of increased research.
A case of chronic autoimmune vestibulopathy improved with steroids

Li Jiang, Sapru Sunil, Wen Xiong

The most common causes of vestibulopathy are ototoxic aminoglycosides, Meniere’s disease and meningitis. The diagnosis of autoimmune vestibulopathy is rare. We present a case of autoimmune vestibulopathy in a 31 year old African-American woman with severe intermittent chronic vertigo and dizziness for fourteen months that improved with the treatment of steroids. The dizziness and vertigo affected her daily life and work. Symptoms were sometimes associated with headache and nausea but not with hearing loss, weakness, numbness or tingling. There was no history of intravenous antibiotic use or head trauma. Past medical history included Vitamin D deficiency and iron deficient anemia. Physical exam and routine labs such as complete blood count with differential and comprehensive metabolic panel were unrevealing. Treatment was initiated with meclizine and vestibular physical therapy without much improvement. Dizziness improved briefly but recurred one week after she was started on topiramate 15mg bid. The patient was then referred to neurology and rheumatology for further evaluation and treatment. Rheumatoid factor, cyclic citrullinated peptides, anti-neutrophil cytoplasmic antibodies, antiphospholipid antibodies, beta 2-glycoprotein-antibody, anti-jo-1 antibody and complement C3 were within normal limits; erythrocyte sedimentation rate was elevated at 48 mm/hr, C-reactive protein was elevated at 6.5 mg/L (normal limit <4.9 mg/L), antinuclear antibodies was 1:80; anti-SSA was positive ( > 8.0 AI ) and anti-SSB was negative. Sialogram was unremarkable. MRI of the brain with and without intravenous contrast revealed non-specific nodular hyperintensities in bifrontal and left frontal parietal subcortical white matter and normal bilateral seventh and eighth intracranial nerve complex. Electronystagmogram and rotator chair test confirmed bilateral central vestibular dysfunction and left peripheral vestibular dysfunction. Autoimmune vestibulopathy was diagnosed and the patient was started on hydroxychloroquine 200mg daily. Methylprednisolone 64 mg per day was added tapering the dose 8 mg every 5 days until 16 mg daily within 4 weeks. She was also placed on calcium and vitamin D. Four weeks after steroids treatment the patient reported significant improvement of dizziness and complete resolution of vertigo. In conclusion, autoimmune vestibulopathy is a relatively rare condition. The diagnosis can be difficult, however a short term treatment trial of steroids may be effective and possibly diagnostic.
Capital Health

It’s hard to tell the poison from the cure: nosocomial sepsis complicating lupus nephritis

Nazish Ahmad, Manish K. Gugnani,

Introduction: Blood cultures are a sensitive method for detection of bacteremia or fungemia. Sometimes, there can be a hidden source can hinder the patient’s progress, requiring further investigation and empiric therapy. Purulent pericarditis is a rare disease and is associated with a high risk for poor outcomes. We report a case of purulent pericarditis caused by Pseudomonas aeruginosa in a patient with MSSA bacteremia and active lupus. Case: A 37 year old African American female, recently diagnosed with Systemic Lupus Erythematosus with lupus nephrits and latent tuberculosis, was admitted for shortness of breath and chest tightness. Physical exam revealed a pericardial rub. CXR showed bilateral pleural effusions. Electrocardiogram was suggestive of pericarditis, and transthoracic echocardiography showed moderate pericardial effusion. Patient was started on colchicine and ibuprofen. The patient decompensated and became hypotensive and required emergent intubation and pressor use. Blood cultures reported gram positive cocci in clusters, later found to be methicillin sensitive staphylococcus aureus (MSSA), which was treated initially with vancomycin and then nafcillin. The patient’s hypotension worsened and required the use of four pressors. Because of no improvement on current antibiotic therapy, a CT of the chest and abdomen revealed a worsening pericardial effusion. Heart sounds became muffled, jugular venous distension was noted, and emergent transthoracic echocardiography confirmed a worsening pericardial effusion, with signs of tamponade. The patient’s antibiotic coverage was broadened to piperillin-tazobactam. Urgent pericardiocentesis revealed 600 ml of purulent, yellow, thick fluid, suspicious for empyema. Unfortunately, the patient expired, and later cultures revealed pseudomonas aeruginosa. Discussion: Purulent pericarditis is an uncommon illness, and is usually secondary to a primary cause of pericarditis. We believe our patient had lupus associated pericarditis and developed line sepsis which was identified by blood culture to be MSSA. However, she failed to improve on therapy for MSSA, and ultimately pseudomonas pericarditis was identified. Most likely, the gram negative species was introduced by the same mechanism as the MSSA, as the patient had no evidence of nosocomial pneumonia. Complex, critically ill patients frequently have multiple processes occurring simultaneously and vigilant physicians must maintain a high index of suspicion for unusual and unexpected disease manifestations. Teaching points: Although blood cultures are accurate in establishing sources and therapeutic intervention, there is a chance a secondary source can remain unidentified. At times, a high index of suspicion requires more aggressive and invasive testing to identify all the processes at work in a complex patient.
**Capital Health**

**A Rare Case of Rhabdomyolysis Associated Acute Kidney Injury complicating Legionella Pneumonia**

Salman Azim, Waqar Siddiqui, Manish Gugnani

**INTRODUCTION:** Legionella is a common cause of community acquired pneumonia with frequent extrapulmonary signs and symptoms, mostly neurologic and gastroenterologic. Musculoskeletal complaints are not typically associated with legionella infection, however, we encountered a case of non-traumatic rhabdomyolysis and acute kidney injury (AKI) associated with Legionella pneumonia.

**CASE PRESENTATION:** A 37-year-old-man, with past history of well controlled hypertension and asthma, presented with complaints of malaise, back pain and dark colored urine. He also complained of cough, fever and chills for several days. On physical examination, vitals were normal except temperature 101.20F. Chest radiograph showed consolidation with diffuse infiltrate of both lung fields. The patient was admitted for treatment of community acquired pneumonia. On day 2, however, the patient suddenly became hemodynamically unstable and developed cardio-pulmonary arrest. Laboratory investigation revealed creatinine 2.88 mg/dl, CPK 58000 U/L and positive urine antigen for Legionella. The patient required hemodialysis for rhabdomyolysis associated AKI. After two weeks, the patient recovered both renal and pulmonary function and had complete relief of symptoms. The follow-up clinical and laboratory findings were normal. **DISCUSSION:** Legionnaire’s disease (LD) has been known to cause pneumonia with multiple organ system involvement. The classic presentations of LD include malaise, myalgia, anorexia, diarrhea, weakness, cough, confusion and headache. Distinctive features of LD with statistical significance compared to other type of community-acquired pneumonia include headache, diarrhea, arthralgia or myalgia, confusion, fever to 39°C, purulent sputum, hyponatremia, hepatic dysfunction, creatine phosphokinase elevation, hypophosphatemia, proteinuria and hematuria. The mechanism of rhabdomyolysis associated with Legionella is unknown, theories include direct invasion of Legionella into the muscle itself, or the release of its endotoxin into the circulation with subsequent muscle injury. In our case, Legionella was considered for the multi-system failure as evidenced by laboratory findings. **CONCLUSION:** Legionella is an unusual non-traumatic cause of rhabdomyolysis associated AKI.
INTRODUCTION: Whipple’s triad of 1) demonstrated hypoglycemia, 2) associated symptoms and 3) relief of symptoms with normalized blood glucose is commonly associated with pancreatic insulinoma. However, because insulinoma is rare, indolent and difficult to identify, many patients with symptomatic hypoglycemia may be diagnosed with other disorders before an investigation is complete. CASE: A 53-year-old man, with past history of recurrent neuroglycopenic symptoms, presented to ED with complaints of feeling sweaty, dizzy, and shaky. The patient also had a chronic non-healing ulcer over great toe with superinfection worsening over several weeks. The patient had at least seven year’s history of symptomatic hypoglycemic episodes, with investigations at several institutions, with normal biochemical testing and imaging. He also had numerous admissions for investigation of the non-healing ulcer and treatment of superinfection, with no definitive diagnosis to explain the ulceration. On this admission, laboratory findings revealed: C-peptide 8.38 ng/ml (0.80-3.10 ng/ml) and Insulin 98 microIU/ml (<23 microIU/ml). CT of the abdomen was normal, however, endoscopic ultrasound (EUS) showed a 1.5 cm mass within the head of pancreas. Immunohistochemical stains of a biopsy specimen were positive for PanCK, synaptophysin and chromogranin, confirming insulinoma. Pre-operative MRI showed a hyper-vascular mass in the uncinate process. The patient underwent surgical resection of the mass and had uncomplicated post-operative recovery with complete relief of symptoms. The patient’s follow-up C-peptide and insulin level were normal. DISCUSSION: Insulinomas are usually small (90% are < 2cm) and solitary, and can be too small to identify on CT scan. Further, biochemical testing can be normal, especially if blood is drawn at times other than the nadir of glycemia. Clinicians involved in our case were suspicious of insulinoma for a significant time before the actual diagnosis was made, however the diagnostic tests of EUS and MRI were not ordered until the biochemical testing was positive. Advanced testing in high suspicion patients should not wait for abnormal biochemical testing. The temporal relationship between the insulinoma and the chronic ulcer suggests a pathophysiologic link, and did add to the suspicion of an underlying malignancy. This case may suggest a pathophysiologic role of hyperinsulinemia in formation of foot ulcers in addition to the neuropathic and vascular processes familiar in Type 2 diabetes. CONCLUSION: A high index of suspicion is required for accurate diagnosis of insulinoma in a patient who presents with the Whipple’s triad even if CT and biochemical testing are normal.
A CASE OF NORWEGIAN SCABIES IN AN IMMUNOCOMPETENT MULTIPLE SCLEROSIS PATIENT WITHOUT EXACERBATING NEUROLOGIC SYMPTOMS.

Salman azim, Waqar Siddiqui, James Ware, Omar Ahmad

Introduction: Commonly immunodeficient patients such as individuals infected with HIV, HTLV infections, immunosuppressive therapy or living in unsanitary conditions present with opportunistic infections such as Norwegian Scabies. Its widely acknowledged as scientific fact that infections exacerbation multiple sclerosis (MS) patients, but a systematic review detailing a decrease in exacerbations in MS patients with parasitic infections makes an exception to that rule. Case Presentation: A 60 yo AAM diagnosed with Multiple Sclerosis twelve years prior not on immunosuppressive therapy and chronic bilateral venous insufficiency presented with severe lower extremity edema with hyperkeratosis and foul smelling friableskin. Patient denied any fever or chills or new neurologic deficits. Due to the severe edema in the lower extremities the patient had some ambulatory dysfunction. The patient had prior history of scabies infections 4 months ago with failure of past treatment. Bacterial cellulitis and deep venous thrombosis were ruled out. The patient was treated initially with Permethrin with no clinical improvement. The patient was then started on Ivermectin which effectively resolved the illness. Discussion: Patients with Multiple Sclerosis are normally vaccinated to prevent infections that can exacerbate their condition. This particular patient had a case of Norwegian Scabies in which millions of mites had infested in his lower extremities. Despite this severe infection the patient did not have any new sensory, motor or ophthalmologic symptoms. A 2007 double blind trial published in the Annals of Neurology demonstrated increased production of IL-10 and TGF-beta, together with induction of CD25+CD4+ FoxP3+ T cell. In this study patients had a decreased incidence of relapse. It suggests that regulatory T cells induced during parasitic infections might alter the course of MS.
Hypertriglyceredemia-induced acute pancreatitis treated with plasmapheresis

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Background: Acute pancreatitis is a common condition with multiple etiologies. While alcohol and gallstones are the most common etiologies in the United States, hypertriglyceredemia (HTG) is responsible for 7% of all cases and has been recognized as the third most common cause of acute pancreatitis. Compared to acute pancreatitis from other etiologies, HTG-induced acute pancreatitis has a higher degree of severity and complications. Therefore, rapidly lowering the triglyceride (TG) level is crucial. Removal of TG by plasmapheresis is a safe and beneficial treatment when conventional methods are inadequate, improving the outcome and preventing complications. Learning Objective: Demonstrate plasmapheresis as a definitive treatment for severe hypertriglyceredemia and pancreatitis.

Case: A 49 year-old African American obese man, presented to the emergency department with complaints of severe upper abdominal pain for approximately ten days. Phlebotomists who drew his blood were alarmed that the blood was milky white in color. Laboratory findings showed: Triglycerides 7313 mg/dL Total cholesterol 1153 mg/dL Lipase 2071 units/L Amylase 271 units/L Glucose 347 mg/dL White blood cell count 17,200 cells/mm3 A CT scan of the abdomen revealed infiltration and haziness around the pancreatic head, body and uncinate process, consistent with acute pancreatitis. On day 2, the patient developed diabetic ketoacidosis (DKA) and was transferred to Intensive Care Unit. Insulin infusion was initiated and maintained for 3 days with lowering of his TG level by 24%. Despite insulin and gemfibrozil therapy for 5 days, the patient's TG level dropped only to 5019 mg/dL, and he continued to report abdominal pain. Being at very high risk of necrotizing pancreatitis, the patient was started on plasmapheresis on the general medical floor and the TG dropped dramatically by 78% in a single session. Two additional plasmapheresis sessions were done lowering his TG level below 500 mg/dL. The patient was discharged in stable condition, pain-free and tolerating a low-fat diet on day 9 with TG 408 mg/dL and total cholesterol 118 mg/dL. The patient was discharged on Lantus 25 units daily, gemfibrozil 600 mg twice daily, and pravastatin 40 mg daily along with lifestyle modifications. Discussion: HTG-induced acute pancreatitis may be definitively treated with plasmapheresis, rapidly and dramatically lowering TG level, improving outcomes and preventing complications. Plasmapheresis is indicated in emergency situations with TG level greater than 1000 mg/dL and should be continued until levels are lowered to less than 500 mg/dL. Medical therapy alone for severe HTG can be slow and severe complications such as pancreatitis may progress before it can take effect. Further, medical therapy may not be effective in fully lowering severely elevated TG levels. After the acute episode, rigorous pharmacological regimen and lifestyle modification are imperative, as is a discussion regarding screening of family members for lipid disorders.
**Capital Health**

**Hepatorenal Syndrome successfully treated with Vasoconstrictors, Albumin and Pentoxifylline**

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Background: Acute kidney injury in the setting of advanced liver disease is both common and very challenging to evaluate and treat. The hepatorenal syndrome (HRS) is one of the most serious complications of liver failure and is especially difficult to manage and is associated with a poor prognosis. Medical therapy with pentoxifylline is thought to prevent onset of HRS in some patients, but may also have a role in treatment of acute decompensations. Case Report: A 51-year old man with cirrhosis Child-Pugh Class C due to hepatitis C and ethanol abuse was admitted with a three day-history of abdominal pain and distention, and bilateral lower extremity edema. The patient had not received antibiotic prophylaxis for prevention of spontaneous bacterial peritonitis (SBP) and had not been listed for liver transplantation due to recent alcohol abuse and non compliance. Daily medications included spironolactone and furosemide. On clinical examination, the patient showed low grade fever, intense jaundice, large ascites, diffuse abdominal pain, hepatosplenomegaly and bilateral lower extremity edema. The pertinent laboratory data were: serum albumin 2.7 g/dL, total bilirubin level 8.2 mg/dL, INR 2.1, WBC 11.5 X10^9/L, bands 39%. Paracentesis removed 960 ml of cloudy peritoneal fluid with 3,986 leucocytes/µl (74% polymorphonuclear cells) and yielded a culture positive for Escherichia coli. Antibiotic therapy with ceftriaxone 2 gm daily was started and continued for 10 days. 24 hours after admission, his serum creatinine increased to 1.69 mg/dL. Diuretics were stopped and an intravenous fluid trial was initiated. His creatinine increased to 3.34 mg/dL. Urinalysis was unremarkable, urine sodium was less than 5mmol/l and the renal ultrasound was negative for hydronephrosis or nephrolithiasis. A diagnosis of type 1 hepatorenal syndrome (HRS) triggered by SBP was made. Pharmacological therapy with octreotide 200 mcg subcutaneous three times a day, midodrine 7.5 mg per mouth three times a day, and albumin 1 gm/kg intravenous daily was started and continued for 12 days. In the first 48 hours his serum creatinine level continued to increase to a peak of 4.52 mg/dL. Pentoxifylline (PTX) 400 mg per mouth three times a day was added and continued for 28 days. His renal function returned to normal by the tenth day of therapy. The patient’s renal function remained normal after the therapy was discontinued, and at 45 days follow-up. Discussion: This case emphasizes the role of infections, especially SBP as trigger for developing HRS and the efficacy of therapy with octreotide, midodrine, albumin, and PTX in the management of this condition. PTX is efficacious in prevention of HRS in patients with severe alcoholic hepatitis; however, based on this case report we suggest that PTX could potentially be used also in the treatment of HRS complicating advanced liver disease.
THE CROUCHING TIGER AND THE HIDDEN DRAGON - AN UNUSUAL CASE OF HEPATOCELLULAR CARCINOMA WITH CONCOMITANT METHICILLIN-RESISTANT STAPHYLOCOCCUS AUREUS LIVER ABSCESS

Background: Methicillin-resistant staphylococcus aureus (MRSA) is an extremely uncommon cause of pyogenic liver abscess. In the presence of hepatic tumors, identifying concomitant liver abscesses can be challenging due to the equivocal nature of radiographic testing and clinical signs. Failure to diagnose the hidden abscess can lead to life-threatening complications and increase morbidity and mortality. We present an unusual case of MRSA associated liver abscess in a patient with newly diagnosed hepatocellular carcinoma (HCC).

Case presentation: A 63-year old Caucasian male presented to the emergency department after having fallen in the bathroom and hitting his head on the sink. On examination the patient complained of headache and intermittent abdominal pain. Past medical history was significant for seizure disorder, hepatitis C, pulmonary hypertension, chronic obstructive pulmonary disease, intravenous drug abuse and coronary artery disease. Patient was admitted to rule out possible seizure and imaging was ordered to rule out any trauma-related injury. Incidentally, the computed tomography scan of the abdomen revealed an ill defined mass in the liver suggestive of HCC. On magnetic resonance imaging, micronodular cirrhosis with multiple suspicious masses of varying sizes again suggestive of HCC was reported. Subsequent serum marker testing revealed alpha fetoprotein level of 88.3 nanograms/milliliter, which was deemed nonspecific. Finally liver biopsy was ordered to confirm the diagnosis. Surprisingly, during the biopsy, 15 ml of purulent material was aspirated and sent for analysis which came back positive for strains of MRSA. Interestingly, blood work done did not reveal any signs of leucocytosis or left-shift. The histopathology report confirmed poorly differentiated HCC. The patient was subsequently started on intravenous vancomycin to complete six weeks of therapy and was scheduled for follow-up.

Discussion: Biopsy of HCC is usually avoided to prevent tumor seeding, unless there is diagnostic uncertainty. In the initial stages of abscess formation, hepatocytes undergoing inflammatory change make it difficult to distinguish it radiologically. The etiological source of MRSA in the abscess remains unclear. Although intravenous drug abuse is certainly a risk factor, a trans-esophageal echocardiogram done was negative for valvular endocarditis. Conclusion: MRSA associated liver abscess may manifest differently in comparison to garden variety pyogenic liver abscesses and may follow a more insidious course. Such patients may not express obvious signs of an underlying infection. The presence of large hepatic tumors makes diagnosing an underlying MRSA abscess difficult and may often be missed if relied solely on radiological imaging.
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Myoclonus following cardiac arrest: what’s the prognosis?

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Background: prognosticating after cardiopulmonary arrest is difficult since patients are at high risk for poor outcomes, and those who improve tend to do so slowly over time. Especially concerning is the risk of anoxic encephalopathy, and worrisome neurologic findings are common in these patients. However, one finding that should not be misinterpreted as a poor prognostic sign is the development of the Lance-Adams Syndrome. Case Presentation: A 76 year old African American male with COPD and polysubstance abuse, was brought to the ED after having severe respiratory distress followed by cardiac arrest. EMS found the patient at home in asystole. Chest compressions with ACLS protocol and mechanical ventilation was started in the field. While on route to the hospital, he had a return to spontaneous circulation. After exclusion criteria, the patient was started on the hypothermia protocol and admitted to the ICU. Per the protocol, cisatracurium, continuous sedation with propofol, and continuous analgesia with fentanyl was administered. During the rewarming phase, the patient started to have multiple episodes of jerking movements of the extremities suspicious for seizure like movements. Lorazepam and levetiracetam were given urgently and the jerking movements improved. He was placed on standing levetiracetam, topirimate and valproic acid. Video EEG monitoring did not show any clinical or electrographic seizures activity, but rather a pattern consistent with metabolic encephalopathy. Paralytics, propofol infusion and analgesia were discontinued once the hypothermia protocol was completed. Patient was weaned and extubated after 8 days. On day 10, patient was alert, awake and able to follow commands, but when he tried to move all 4 extremities, there were myoclonic movements most prominently in the upper extremities. After 3 weeks of physical therapy, he recovered his strength in all extremities and the jerking movements resolved. Patient was transferred to the rehab to continue physical therapy. Discussion: Lance-Adams syndrome is characterized by intention myoclonus without loss of consciousness and is a complication of successful resuscitation after cardiac arrest. It is very important to distinguish Lance-Adams syndrome from myoclonic status epilepticus which is considered to indicate a poor prognosis which can lead to inappropriate withdrawal of life support. Lance-Adams syndrome indicates a relatively good prognosis after cardiac arrest and patients should be referred to a comprehensive rehabilitation program.
Acute urinary retention in previously healthy young patient

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INTRODUCTION: Acute urinary retention is most often related to outflow obstruction secondary to trauma, infection, neoplasm or benign prostate hyperplasia. Less commonly it could be caused by neurological diseases, such as spinal cord injuries or inflammatory demyelinating disorders. Acute urinary retention in young patients raises the suspicion for an unusual disorder, such as an acute neurologic condition. CASE REPORTS: A 38-year-old man with recent history of an upper respiratory viral infection was seen in emergency room with complaints of tingling sensations in the upper body. At that time, he did not have any significant findings and was discharged home with a prescription for naproxen. Two days later, he returned to the emergency room with the inability to urinate and severe suprapubic pain. He also complained of exhausting general fatigue, myalgia and malaise. Physical and detailed neurological examination did not reveal any significant abnormalities. Foley catheter was placed and 1000 mL of urine returned with immediate relief. CBC, CMP, basic screening for autoimmune diseases (ANA, Sjogren Abs) and urinalysis were normal. MRI of the brain, C-spine, T-spine, L-spine did not show any evidence of spinal cord compression, tumor, abscess, or multiple sclerosis. Lumbar puncture showed a colourless liquid under normal pressure, WBC 206 /mm3; Lymph 95%; Neutrophils 5%; Glucose 52 mg/dl; Protein 61 mg/dL; no Oligoclonal bands; Synthesis rate, IgG -2.2; non-reactive VDRL; negative Lyme Ab; negative HSV 1 and 2 DNA PCR. Given the clinical symptoms of viral syndrome, isolated acute urinary retention and pleocytosis in CSF, the patient was diagnosed with idiopathic sacral myeloradiculopathy and started on empiric treatment with corticosteroids. The Foley catheter was removed on third hospital day and the patient was discharged symptom free on the fifth day. DISCUSSIONS: Acute urinary retention due to sacral myeloradiculitis is a rare neurological syndrome. Initial workup of acute urinary retention includes evaluation for possible outflow obstruction. Next step will be MRI of the spinal cord and CSF analysis to rule out possible structural spinal cord lesions and demyelinating disorders, such as Guillain- Barr syndrome or multiple sclerosis. Blood tests can be helpful in diagnosing SLE, HIV infection or Lyme disease related CNS disorders. If none of these tests suggests a specific cause, sacral myeloradiculitis should be considered in young patients with a viral syndrome, isolated acute urinary retention and pleocytosis in CSF.
Severe Hypertriglyceridemia – Plasmapheresis or Insulin infusion therapy for treatment?

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INTRODUCTION: Severe Hypertriglyceridemia (HTG) may present clinically with signs of acute pancreatitis, acute cholecystitis or remain asymptomatic. Sometimes, the clinical presentation of HTG may be misinterpreted as Diabetic Keto-acidosis (DKA). The current mainstay of treatment for severe HTG includes Plasmapheresis, Insulin infusion, heparin infusion, and lipid lowering agents. There are hardly any studies done to compare effectiveness of these treatment modalities. AIM: To compare effectiveness of plasmapheresis vs. high dose Insulin infusion therapy for the treatment of severe HTG.

CASE PRESENTATION: We reported two cases of severe HTG. One patient was treated with plasmapheresis and the second patient with high dose insulin infusion. Case 1: A 49-year-old man presented to ED with complaints of severe epigastric pain, nausea and vomiting. The patient’s vitals were within reference range. Patient’s laboratory findings included: blood glucose 347 mg/dl, Sodium 136 mmol/L, Potassium 4.5 mmol/L, Chloride 104 mmol/L, CO 2 16 mmol/L. Detailed laboratory investigations were not possible because of milky blood. CT abdomen showed findings compatible with acute pancreatitis. Initial diagnosis of DKA was made and the patient was treated with DKA protocol, however, the patient did not improve. On second day of admission, the laboratory investigation showed triglyceride 7313 mg/dl, amylase 159 U/L, lipase 926 U/L, total cholesterol 477 mg/dl. He was treated with plasmapheresis, and triglyceride level improved to 1141 mg/dl after first cycle and 348 mg/dl after second cycle. The patient showed signs of complete recovery without any complication. Case 2: A 34-year-old man, with past history of Diabetes Mellitus, Dyslipidemia and alcohol dependence, presented to the ED with complaints of nausea, vomiting, back pain and severe epigastric pain. A CT Abdomen did not reveal any abnormality. The laboratory investigation showed: PH 7.16, Triglyceride 6388 mg/dl, Cholesterol 1053, Amylase and Lipase within reference range, blood glucose 247 mg/dl. Initial diagnosis of DKA and severe HTG was made. The patient was treated with high dose Insulin infusion therapy (Fixed dose: 10U/hour) with 10 % Dextrose. Follow up laboratory investigation showed triglyceride level of 2879 mg/dl on second day, subsequently a level of 487 mg/dl on the following days. His pH improved to 7.30, Bicarbonate 18.5 mmol/L with complete clinical recovery. CONCLUSION: We report that both Plasmapheresis and Insulin infusion therapy are effective in treatment of severe HTG. However, in either strategy, early diagnosis and prompt treatment should be considered to prevent fatal complications. The use of Insulin infusion therapy can be considered to treat severe HTG in remote places where there are no facilities for plasmapheresis. However, given the relatively low prevalence of severe HTG, a multicentric approach is required to study comparative effectiveness in an adequately large population.
Neurocysticercosis mimicking the presentation of normal pressure hydrocephalus

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Background: Neurocysticercosis is the most common parasitic infection of the nervous system in developing countries. It is acquired through feco-oral contact with carriers of the adult tape worm Taenia solium with reported cases of autoinfection. Incidence rates in the US are rising because of increasing immigration. Seizures are the most common presentation but intracranial hypertension, hydrocephalus, chronic meningitis and cranial nerve abnormalities may also occur. The presenting symptoms may resemble other neurologic conditions, so a high index of suspicion and early imaging are the key to implement appropriate management.  

Case: A 64 year old Hispanic male with past medical history of diabetes and hypertension presented to the emergency department with three months of frequent falls and imbalance with new onset of urinary incontinence and loss of orientation to place. Two previous visits to the ED with similar symptoms and CT of the head showed enlargement of the ventricles out of proportion to the sulci. On this presentation, he had worsening ambulatory dysfunction. Physical exam revealed ataxic gait and loss of orientation to place. CT of the head was unchanged. Normal pressure hydrocephalus was considered as the primary working diagnosis. On lumbar puncture, the opening pressure was normal, but there was a WBC of 135/cubic mm with 85 % lymphocytes and protein of 83mg/dL. MRI of the head with gadolinium was then done revealing a 1.5cm non-enhancing cystic lesion within the medial right occipital lobe with communicating hydrocephalus. The radiological findings of the cyst suggested acute stage neurocysticercosis without any calcifications. Serum antibody to taenia solium was positive. He received a four week course of albendazole, and oral steroid taper. In addition, the interferon gamma release assay for M. tuberculosis was positive. Initially, he began four drug anti-tuberculous therapy with isoniazide, rifampin, ethambutol and pyrizinamide, which was then stopped when the cerebrospinal fluid cultures were negative for tuberculosis. He continued only isoniazide therapy for total of nine months. On follow up as outpatient, repeat magnetic resonance imaging of the head showed resolution of the cystic lesion to 8mm. Patient reported marked improvement of his gait and urinary incontinence. His family reported improving orientation. Ventriculoperitoneal shunt placement was planned for the patient by neurosurgery, but was subsequently cancelled because of the marked clinical improvement.  

Discussion: Neurocysticercosis can be missed in favor of more common diagnoses if the classical presentation of large cystic lesions with CSF lymphocytosis is not present. Further imaging with magnetic resonance imaging and serum testing for Taenia solium antibodies are helpful diagnostic modalities along with a detailed history including exposure to areas with endemic disease.  

Teaching Point: Symptomatic neurocysticercosis can mimic other common neurological diseases such as normal pressure hydrocephalus requiring a high index of suspicion to identify correctly.
Severe legionnaires disease complicated by interstitial nephritis and rhabdomyolysis

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Learning Objective: Atypical manifestations of pneumonia should raise the concern for Legionella since the mortality without treatment with appropriate antibiotics is as high as 60-70 percent. Case: A 54 yo AAM was brought to the ED due to neurological changes, weakness and a fall. Family members reported that the patient was confused, hallucinating, and talking to himself. On his way to bathroom he fell and could not get up due to weakness and laid in the hallway for unknown period of time. The family also reported some diarrhea. PMH was positive for alcoholic neuropathy and HTN. He had a fever of 99.1 F, tachycardic at 117 and increased RR of 25-30. On physical exam crackles were heard on the left lower base of lung with equal entry of air on both lungs. Neurologically pt was alert and awake but not oriented. He was trying to communicate but speech was incoherent. He followed simple verbal commands. There was increased muscle tone and drift of bilateral upper extremities. CXR showed LLL infiltrate and CT chest confirmed extensive left lower lobe pneumonia with interstitial component. CBC showed leucocytosis of 16.6 x10^9/L with bandemia of 21%. Additionally there was acute kidney injury with a creatinine of 5.93 mg/dl and BUN of 41mg/dl with bicarbonate of 16 mmol/L and anion gap of 25. CPK was >161600 U/L, AST was 3619 U/L, and ALT 482 U/L. ABG showed a 7.44 U/20.5 mmHg /68.7 mmHg /13.6 mmol/L. Considering all the labs with clinical constellation, Legionella pneumonia was suspected and levofloxacin was started with renal dosing. Hemodialysis was started urgently for the AKI associated with rhabdomyolysis. A urine legionella antigen test from admission was positive for legionella pneumophila serogroup 1 Ag. Urinalysis revealed urine eosinophils suggesting of interstitial nephritis. The patient required mechanical ventilation but successfully weaned and was discharged after 26 days. Creatinine on discharge was 4.95 mg/dl which returned to 1.39 mg/dl at 2 months follow up. Discussion: Legionella is an important cause of community acquired pneumonia, and is associated with a variety of extra-pulmonary manifestations. Most commonly, neurologic and gastroenterologic systems are affected, which occurred prominently in our patient. The rhabdomyolysis and acute kidney injury may have been associated with the patient’s fall and alcohol use, however the urinalysis would have been more likely to exhibit an ATN picture rather than a nephritic pattern. Acute interstitial nephritis is a well described though pathophysiologically poorly understood manifestation of Legionella infection. Aggressive resuscitation and treatment including mechanical ventilation, hemodialysis and antibiotic management carried out in intensive care frequently results in good outcomes, reducing the mortality rate from 60-70% to 10-20%. Atypical multi–organ manifestations in patients diagnosed with pneumonia should raise concern for Legionella pneumonia.
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Introduction Anomalous coronary arteries are rare but potentially life threatening abnormalities of the coronary circulation. Many of the variations can be benign in nature, however some may lead to myocardial infarction or sudden cardiac arrest. Dynamic functional testing may be necessary to identify those requiring intervention before complications occur. Case Report A 34 year old African American male with a past history of hypertension and smoking presented with a two day history of stabbing epigastric pain while eating that radiated to the mid sternum. Initial ECG showed normal sinus rhythm, with left axis deviation and early repolarization. A nuclear stress test showed evidence of lateral wall ischemia and mild diffuse left ventricular hypokinesis. Cardiac catheterization revealed an ejection fraction of 50% and the right coronary artery having an anomalous origin directly from the left sinus of Valsalva/left main coronary with a malignant course between the aorta and pulmonary outflow track. Patient further underwent a dobutamine cardiac catheterization that led to compression of artery during stress and was thus scheduled for a single vessel bypass graft. The rationale for surgical repair was based on a significantly increased risk for sudden cardiac arrest and myocardial infarction during exercise and stress secondary to dilatation of the pulmonary artery and aorta leading to compression of the proximal portion of the anomalous vessel leading to ischemia, arrhythmias and sudden death.

Discussion Sequelae of coronary artery anomalies may range from severe events such as sudden cardiac death, dyspnea, angina pectoris, dizziness, palpitations, and syncope to being entirely asymptomatic. Various mechanisms have been postulated for the occurrence of symptoms including, origin in an acute angle and folding or occlusion caused by angulation at the point of the coronary artery emergence, coronary vasoospasm resulting from its torsion movement, mechanical compression of the anomalous artery between the pulmonary and aortic trunks during physical exertion, and an intramural origin of the coronary artery from within the tunica aortic media. The majority of the complication may be exacerbated during or immediately after exercise, as this leads to compression of the coronary arteries as well as increasing the preexisting angulation of the proximal portion of the anomalous vessel. Treatment of the anomalous vessel may need surgical reconstruction, decompression, and reimplantation in the correct sinus, or bypass with ligation of the native vessel to eliminate competitive flow in the graft. Particularly if the vessel courses between the aorta and the pulmonary trunks, has a narrow ostium or arises from the pulmonary vasculature, it is highly susceptible to repeated episodes of myocardial ischemia. These patients benefit from surgical intervention. Those patients with asymptomatic or non-significant anomalous coronary arteries may be observed and managed medically.
The Syndrome of Irreversible Lithium – Effectuated Neurotoxicity – A SILENT cause of prolonged altered mental status

Jina Makadia, Manish Gugnani MD, Tania Calzada MD, Daniel Goldsmith MD

Learning objective: Persistent sequelae of lithium intoxication gained clinical attention in the 1980’s and was named Syndrome of Irreversible Lithium-Effectuated Neurotoxicity (SILENT). It is important to consider the diagnosis in patients with refractory altered mental status after Lithium Poisoning.

Introduction and background: Lithium has been commonly used for the treatment of several mood disorders particularly bipolar disorder since the 1870’s and gained much popularity as a potent mood stabilizer. Lithium has a relatively narrow therapeutic index that predisposes patients on chronic lithium maintenance treatment to poisoning with relatively minor changes in dose or health status. Well known side effect of long term lithium use include Nephrogenic Diabetes Insipidus (NDI), teratogenicity and neurologic sequelae including coarse tremor, muscle twitching, and convulsions. It is unclear to what extent its long-term use may result in either neuroprotective or toxic consequences, however persons who survive a poisoning episode may develop persistent neurotoxicity. Case Presentation: A 57 year old Caucasian male with past history of schizophrenia presented with polyuria, tremors, observed imbalance and a fall. Lithium levels were found to be 1.4 mmol/L. Poison Control was contacted and aggressive hydration was advised. The patient was initially alert, awake and oriented only to self, which was his reported baseline as per his long term care facility. On day 2 of admission, he developed altered mentation, becoming less responsive, but agitated and constantly moving all extremities in a choreoathetoid way. Lithium levels were below normal range (0.4 mmol/L; range 0.6-1.2). Serum and urine electrolytes were consistent with NDI and was well managed. The altered mental status persisted despite normalization of blood chemistries. Further workup including CT head, MRI head, EEG, LP, and blood cultures were all normal. Patient’s clinical picture did not improve and continued to have the choreoathetoid type movements, and being non-verbal. Patient was transferred to a nursing home in the same state. Discussion: A syndrome of persistent neurologic sequelae of lithium intoxication was recognized in the 1980s and dubbed the Syndrome of Irreversible Lithium-Effectuated Neurotoxicity (SILENT). Although the biologic mechanism remains unclear, the authors hypothesize that the putative cause of SILENT is demyelination caused by lithium at multiple sites in the nervous system, including the cerebellum. Recent advances in the understanding of the molecular basis of lithium-induced neurotoxicity may be able to provide a means of defining a pathway associated with the long-term prophylactic properties of lithium, distinct from its toxicity profile. It has also been found that there is a long term persistence of the symptoms months after the level of lithium is below normal, including cerebellar dysfunction, extra-pyramidal symptoms, brainstem dysfunction, and dementia. SILENT can continue for months and, in rare cases, effects persist for years.
All that Falls: A simple non invasive test to reveal carotid mass as etiology of syncope

Anubha Mishra Tewary, Sergey Gerasim, MD, Bipinpreet Nagra, MD

Introduction: Current strategies for the evaluation of syncope were outlined in guidelines published by the European Society of Cardiology (ESC) in 2009, and in a 2006 scientific statement from the American Heart Association/American College of Cardiology (AHA/ACC). These however, do not include a trans carotid Doppler ultrasound, an easy and cost effective diagnostic tool. Case Presentation: The patient is an 83-year-old female who was admitted to the hospital after recurrent syncopal episodes. She had a history of TIA, and prior work up from two years ago included a carotid ultrasound which showed no significant disease present on the left and a mild 1% to 15% disease on the right, and no mention of any carotid body mass. CTA of neck was normal. At the current admission, carotid duplex ultrasound showed 1% to 15% of stenosis bilaterally and a small mass located within the carotid bifurcation on the right. CTA of the neck showed the bifurcation mass measuring approximately 2.6 cm x 1.8 x 3.5 cm in length and vertebral artery stenosis on the right. Excisional biopsy of the carotid body paraganglioma was performed. Discussion: Current guidelines for evaluation of patients with a syncope episode do not include carotid ultrasound - a benign, non invasive study. In our case the ultrasound revealed the carotid body tumor at the bifurcation of the right carotid artery as the cause of recurrent syncopal episodes. This should prompt clinicians to have a low threshold for doing a trans carotid Doppler ultrasound in the appropriate clinical setting, and recognize the limitations of guideline documents as not always addressing the diagnostic needs of every patient.
Acute Cough following shoulder surgery

Anubha Mishra Tewary, Dr Manish Gugnani

Introduction: Migration of orthopaedic fixation wires and pins into the thoracic cavity occurs infrequently, but can have dire consequences. Although rare, intrathoracic migration is a serious complication that warrants immediate removal of the foreign body. Reports have shown that all types of pins can migrate even asymptotically, and postoperative course is greatly variable. Case Presentation: The patient is an 85-year-old female with a known history of COPD, and a history of recent (1 week ago) right shoulder reduction with internal fixation, who presented to the ER with cough for 3-4 days. A chest x-ray from the first day of her cough showed a likely right middle lobe infiltrate. She was treated with IV antibiotics and bronchodilators for health care associated pneumonia but her cough persisted and evolved into hemoptysis. Repeat chest radiograph showed re-dislocation of the right shoulder and migration of the Steinmann pin. CT scan of the chest showed the pin extending through the humerus into the chest cavity and into the lung parenchyma. The patient underwent a prompt right thoracotomy with removal of the foreign body and evacuation of hemothorax. Fortunately, the patient did well and recovered successfully. Discussion: Migration of fixation wires and pins within the chest is uncommon, but is a known complication, particularly with fixation wires or pins around the shoulder and may occur as early as day 1 to as late as years to decades after the original procedure. The process may be asymptomatic or rarely catastrophic cardiovascular events and cardiac tamponade may ensue. Serious, non-fatal complications include pericardial tamponade, arrhythmia, pericarditis, pseudoaneurysm, aortopulmonary fistula, pneumothorax, haemoptysis (as in our patient), subclavian steal syndrome, hemianopia, hemiplegia, paraplegia, radicular pain, dysphagia and splenic hematoma. Therefore, for patients with a history of internal fixation devices, time notwithstanding, the clinician should have a suspicion for migration of the device as the implicating cause for the symptoms (even as innocuous as a cough) and an early imaging helps in recognition of the offending body. If suspected, a CT scan clearly evaluates the exact location of the migrated pin or wire, as well as defining complications. Any fractured pins or wires must be removed immediately to prevent dangerous migration. The purpose of this report is to emphasize that such migration may begin early or late, and it should always be considered by clinicians, as they tend to progress rapidly, and successful outcomes warrant an urgent recognition and removal of the migrated foreign body.
Introduction: In the current literature there is no consensus how to treat a symptomatic floating aortic thrombus, however, detection of this type of aortic pathology has increased considerably due to the increasing use of TEE after any embolic event. Nevertheless, therapeutic management of an intraluminal mobile thrombus of the aorta remains controversial. Case Presentation: A 49 year old female with recent hospitalization for TIA and visual changes was found to have bilateral cerebellar strokes, and also a large mobile mass in the aortic arch. She had history of HTN. Investigations showed a negative screening for thrombophilic disorders, no significant family history and no hyperlipidemia. A CTA of the neck reveal a filling defect in the aortic arch and a subsequent TEE revealed the presence of a highly mobile 1.2 x 0.9 cm mass in the aortic arch. In agreement with the cardiologist, neurologist and thoracic surgery, the patient was started on heparin and discharged with the plan to anticoagulate and repeat a TEE in 4-6 weeks, with a surgical removal planned if the mass persisted. However, fortunately for this case, the mass was significantly reduced to a grade 1 plaque on repeat TEE. Further monitoring for this patient at this time includes long-term anticoagulation with repeat TEE in 6 months. Discussion: Current literature lacks agreement about diagnostic and therapeutic approaches to mobile aortic thrombi. Comparative studies have not been performed to achieve consensus so therapeutic strategies are influenced by a variety of factors, including the co-morbidities of the patient and the patient’s and physician’s preferences. Based on case reports and series, it is not unreasonable to initiate a trial of anticoagulation in the appropriate clinical setting, until good quality data are available. Thoracic mural thrombi are a dynamic pathological process with evolving therapeutic approaches and long-term anticoagulation therapy currently represents a viable first line strategy, with surgical or endovascular treatment as a backup option.
Introduction  Evaluation of chest pain is a frequent and essential function of general internists, but it can be challenging given the frequency of non-specific ECG findings and borderline elevated biomarkers. Most cases can be successfully approached with non-invasive testing, however, practitioners must recognize high risk syndromes that preclude exercise stress testing and require urgent invasive evaluation.  

Case  A 56 year-old Hispanic male presented with moderate intensity pressure-like retrosternal chest pain at rest, which began 18 hours before. The pain radiated to the left shoulder, accompanied by shortness of breath and subsided after 10 minutes. He has never experienced this pain before. The pain reappeared twice, 4 hours, and then 1 hour before presentation. These episodes lasted 10-15 minutes and were more intense, and also resolved spontaneously. There was no history of diabetes, hypertension, smoking or familial premature coronary artery disease. Aside from bradycardia of 52 bpm, and discomfort on chest wall palpation, the physical exam was unremarkable. Chest X-ray was normal. Troponin was slightly elevated at 0.04 (units). EKG showed sinus rhythm and biphasic T waves with preserved R waves in the precordial leads (V2,V3). A clinical diagnosis of unstable angina was made and the patient was treated with heparin, clopidogrel, aspirin, high dose statin and nitroglycerine paste. Beta-blockers were withheld due to the bradycardia. Several hours later, the troponin had increased to 0.1 (units) and a repeat EKG demonstrated resolution of the biphasic T-waves. Cardiac catheterization revealed triple-vessel disease including a critical stenosis of the proximal LAD, 60% stenosis of the LMCA, and diffuse RCA disease. The patient underwent successful coronary bypass surgery with resolution of his symptoms.  

Discussion  In 1982, Wellens’ group first described certain criteria by which critical stenosis in the proximal LAD could be diagnosed from specific T-wave changes on an ECG. There are two variants of precordial ST-T wave abnormalities that constitute Wellens’ syndrome. The more common abnormality has deeply symmetrically inverted T waves in leads V2 and V3, with frequent involvement of V1 and V4. The smaller subset demonstrates biphasic T waves in leads V2 and V3, similar to those seen in our patient. Wellens' syndrome is considered a dangerous pre-infarction state as 75% of patients go on to have an acute MI within 21 days. Up to 80% of patients do not manifest elevated troponin levels until frank MI, so recognition of the ECG changes is crucial to identifying these high risk patients. Treadmill stress testing is not indicated in patients with this ECG presentation, as it places them at risk for acute anterior wall MI. Patients should proceed directly to cardiac catheterization because of the high risk of impending anterior MI, and the poor response to even intensive medical therapy alone.
INTRODUCTION: DRESS syndrome (Drug Rash with Eosinophilia and Systemic Symptoms) is a rare drug hypersensitivity reaction with a significant mortality. We would like to report a rare case of DRESS syndrome who developed acute fulminant liver failure who required liver transplantation after vancomycin therapy. CASE PRESENTATION: A 37-year-old man, with history of inguinal abscess after hip fracture surgery two weeks before, presented to the ED with complaints of low grade fever and rash over his body for about one week. The patient’s medications included vancomycin and metronidazole. On physical examination, the rash was progressive and involved the upper half of the body and extremities. Vitals were within reference range. Laboratory investigations showed: eosinophils 12%, AST 1025 U/L, ALT 738 U/L, PT 83 seconds, PTT 50 Seconds, INR 11 and increased D-dimer. Serum ceruloplasmin, anti-nuclear antibody, anti-smooth muscle antibody and ammonia were within reference range. A diagnosis of acute fulminant liver failure and DRESS syndrome due to vancomycin was made. Patient’s vancomycin and metronidazole were discontinued. The patient was treated with intravenous Dextrose, Vitamin K, Fresh Frozen Plasma and N-Acetylcystine. The patient was transferred to a transplant center for liver transplantation. DISCUSSION: The hypersensitivity syndrome, described as DRESS syndrome, is a severe, acute, drug reaction, defined by the presence of fever, cutaneous eruption, and systemic findings including enlarged lymph nodes, hepatitis, or hematologic abnormalities with eosinophilia and atypical lymphocytes. The differential diagnosis includes Stevens-Johnson syndrome (SJS), a life-threatening, cutaneous adverse reaction. Precise diagnostic boundaries between SJS and DRESS have not been well established. The two syndromes overlap clinically, but have different characteristics, treatments and prognosis. This reaction can be life threatening, with a mortality rate of approximately 10%, most commonly secondary to liver failure. The pathogenesis of DRESS syndrome is not fully understood, and may be multifactorial. Histological and immunological investigations suggest an important role of granzymeB and FasL mediated cell death in DRESS associated hepatitis. CONCLUSION: A high index of suspicion and early diagnosis of fulminant liver failure is required for urgent need of liver transplantation to prevent fatal life threatening complications.
INTRODUCTION: The Coronary artery fistula is an abnormal communication between a Coronary artery and either a cardiac chamber or a major vessel. We report a case of Coronary – Bronchial artery fistula who presented with symptoms of angina and shortness of breath. CASE PRESENTATION: A 59-year-old woman, with past history of Hypertension, Asthma and Ectopic Atrial Tachycardia, presented to the Emergency Department with complaints of chest pain and palpitations for several days. The chest pain was substernally located, intermittent, did not radiate and lasted for about 15 minutes. The patient’s home medication included oral sotalol. On physical examination, the patient's vitals were within normal range except blood pressure 152/72 mm Hg. Initial electrocardiogram showed atrial tachycardia. In the emergency room, the patient was treated with intravenous metoprolol. A stress test showed mild-to-moderate apical ischemia. Cardiac catheterization showed a large fistula from right and left circumflex coronary arteries, although the drainage point of fistula was not obvious from coronary angiogram, and normal left ventricular function. A CT Angiogram showed a large branch arising from the right coronary artery that extended posteriorly between the aorta and right ventricle. This branch gave multiple spider-like collateral branches extending into both hila and create a tangle of vessels, connecting them to the bronchial artery, consistent with Coronary- Bronchial artery Fistula. The patient underwent coiling of the fistula from right coronary artery to bronchial artery. The patient had an uncomplicated post-operative recovery with complete relief of palpitation, shortness of breath and angina indicating that steal phenomenon was responsible for patient’s symptoms. DISCUSSION: Coronary- Bronchial Artery Fistulae are usually present from birth with few hemodynamic consequences. Most of the fistulas do not cause symptoms until the patient is in the fourth or fifth decade of life. Once dilated, they cause coronary steal phenomenon as was seen in our patient. The majority of these fistulae arise from the right coronary artery and the left anterior descending coronary artery; the circumflex coronary artery is rarely involved. Clinical manifestations vary considerably and the long-term outcome is not fully known. The patients with coronary fistulae may present with hemoptysis, congestive heart failure, angina, endocarditis, arrhythmias, or myocardial infarction. CT Angiogram and cardiac catheterization are used for accurate diagnosis and for assessment of coronary hemodynamics. Therapeutic options include surgical correction, transcatheter coil embolization and placement of stent graft. CONCLUSION: Coronary- Bronchial Fistula is a very rare condition. A high index of suspicion is needed for accurate diagnosis and prompt treatment to avoid potentially life threatening complications.
INTRODUCTION: Adverse drug reactions are an important cause of morbidity and mortality, accounting for an estimated 5% of all hospitalizations and 0.03% of hospital deaths. Bactrim induced lung injury is a rare event seen in clinical practice. There are hardly few cases with Bactrim induced diffuse lung disease (DLD) have been reported. We would like to report such an unusual case. CASE PRESENTATION: A 59-year-old woman, with past history of buttock abscess treated with incision and Drainage one week before, who presented to ED with complaints of sudden onset of worsening dyspnea for past few days. The patient was treated with Bactrim for seven days for an abscess. The patient also complained of itching and redness in both eyes. On physical examination, patient’s vitals were: Temperature 101.10F, PR 105/min, RR 24/min, BP 103/55 mm Hg, SaO2 90%, and decreased air entry on right lung base. Initial laboratory investigation showed 29% bandemia and lactic acid level of 2.4 mg/dl. A Chest radiograph showed mild increase interstitial lung markings. Initial diagnosis of Pneumonia was made and the patient was started on Rocephin and Zithromax. Two days after admission, patient did not improve. A CT Scan showed marked diffuse reticulation involving both lung fields. The patient suddenly became hemodynamically unstable and was transferred to ICU for intubation and chest tube placement. Interestingly, lung biopsy revealed diffuse alveolar damage with mild-to-moderate anthracosis and bronchiolitis without evidence of granulomatous inflammation or malignancy. The diagnosis of Bactrim induced DLD was made and patient was treated with high dose corticosteroids. The patient had uncomplicated recovery with complete relief of symptoms. DISCUSSION: Lung injury is an increasing cause of morbidity and mortality in patients treated with cytotoxic and noncytotoxic drugs. Over 450 drugs are now recognized as being implicated in drug-induced lung diseases. It is extremely rare to find Bactrim induced diffuse interstitial lung injury. At present, there is no consensus for a definite diagnostic workup approach in patients with a suspicion of DLD caused by bactrim. A carefully obtained history that includes medications (including those sold over-the counter) is essential to suspecting a drug induced reaction. Recognition of drug-induced DLD is difficult because the clinical, radio-logic, and histological findings are nonspecific. The diagnosis is based on a history of drug exposure, histological evidence of lung damage, and exclusion of other causes of lung injury. CONCLUSION: A high index of suspicion is required for early diagnosis and treatment of DLD caused by medications to prevent potential life threatening complications.
Spinal Cord Infarction: A Difficult Diagnosis

Rexanne Caga-anan,

INTRODUCTION Spinal cord infarction is rare and often misdiagnosed. Because of the rich anastomotic blood supply and the system of collaterals, the presentation is varied and unpredictable. The sensitivity of the initial MRI of the cord is limited, contributing to the difficulty of this diagnosis. CASE An 81 year old male with coronary artery disease, diabetes and hypertension had an acute onset of left upper extremity weakness, with rapid progression to quadriparesis in 24 hours. He also had numbness starting from the lower half of his abdomen. He did not have back pain, respiratory or bulbar symptoms. He had no preceding surgery, trauma, infection or illnesses. He had asymmetric quadriparesis with decreased sensation from T8 and downwards. CT scan of the head was unremarkable. On his second hospital day, he lost deep tendon reflexes and developed Babinski reflex with autonomic dysfunction. MRI of the head, brain and cervical spine did not show evidence of compression, inflammation or infarction. EMG was suggestive of generalized demyelination. He was started on IVIG for a presumed atypical presentation of Guillain-Barre Syndrome. However, he did not show improvement upon completion of treatment. CSF analysis was unremarkable. Because of the persistent Babinski reflex and sensory level disturbance, a repeat MRI of the cervical spine was done and it revealed an anterior spinal cord infarct from C5 through T1. Possible etiologies such as vasculitis, aortic disease, cardioembolism, infection and prothrombotic states were ruled out. The patient was then discharged to a rehabilitation facility for aggressive physical and occupational therapy. DISCUSSION Spinal cord infarction has a wide array of pathologic etiologies, but is most often idiopathic. The diagnosis is mostly clinical, but because of the presence of collateral networks, the presentation is highly variable and is often missed. In 17-45% of patients, the initial MRI is unremarkable. If the index of suspicion is high, serial scans should be performed. CONCLUSION Spinal cord infarction is a rare and devastating disease that is often missed because of varying clinical presentations and the low sensitivity of the initial MRI. Serial scans should therefore be performed if the diagnosis is suspected. REFERENCES 1- Sivadasan, A., et. Al. Spectrum of Clinicoradiological Findings in Spinal Cord Infarction. Ann Indian Acad Neurol. 2013 Apr-Jun; 16(2): 190–193.
STAGHORN CALCULI IN PRIMARY HYPERPARATHYROIDISM

Raj Akula,

INTRODUCTION: The prevalence of Primary Hyperparathyroidism (0.003%) and staghorn calculi (1.2%) in general population is significant and is easily diagnosed. However, coexistence of the two conditions together with normal lab values is a rare coincidence. CASE: A 63 year old female with a past medical history of renal stones came to the hospital with complaints of constipation since two weeks. Her symptoms include decrease in appetite, depressed mood and polyuria. On presentation her vitals were within normal limits. Her blood work revealed a calcium level of 10.2mg/dl and phosphorous of 2.5mg/dl. Urine microscopy and analysis showed 10-20 RBC, large leucocyte-esterase, blood and protein. An abdominal CT with contrast showed large Right staghorn calculus and multiple renal calculi on both sides with multiple dilated calyces. Urine culture grew Klebsiella and appropriate antibiotic was started. A serum PTH was elevated at 185.6. A diagnosis of primary hyperparathyroidism was made in the setting of his symptoms. She received IV hydration as treatment for the hypercalcemia. A nephrolithotomy was performed and the staghorn calculus was removed. A sestamibi scan performed revealed a parathyroid adenoma and she subsequently underwent a parathyroidectomy. DISCUSSION: Though individually primary hyperparathyroidism and staghorn calculi are prevalent and easily diagnosed, a mixed picture can pose a diagnostic challenge. In a small percentage of cases long standing hyperparathyroidism can present atypically with a staghorn calculus(1). This can easily be overlooked especially in the background of normal lab values. Thus a physician has to have an inquisitive approach when diagnosing primary hyperparathyroidism and needs to take into consideration all the atypical ways it may present. CONCLUSION: This case reinforces the fact that hyperparathyroidism is a benign disease and a physician should have a keen eye and consider all the subtle ways of presentation to make an accurate diagnosis. Reference: 1) Philip George AJ et al. Brown tumor and staghorn calculi in primary hyperparathyroidism. Urology. 2013 Aug;82(2)
Introduction: Scurvy is an uncommon clinical problem present in industrialized countries. Case: A 44-year-old male from Guyana living in the United States for the last nine years presented to the emergency room complaining of acute on chronic low back pain and sciatica. His presentation was complicated by a generalized tonic-clonic seizure. He had been unemployed for six months and was drinking 3-4 alcoholic drinks daily. His last drink was two days prior to admission. He also revealed bleeding of the gums while brushing his teeth. On physical examination, the patient was anxious and irritable. Skin was dry with extensive ecchymotic lesions and perifollicular hemorrhages on all extremities, the trunk and lower back. Besides a positive straight leg raising test and 4/5 muscle strength on the extremities, the rest of the physical exam was unremarkable.

Labs

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Treatment: In addition to being treated for alcohol withdrawal seizures, he was started on vitamin C 2,500 units and multivitamin. There was marked improvement in his skin lesions over the seven-day course after receiving only high-dose vitamin C, multivitamins and intravenous fluids.

Discussion: Studies have shown Vitamin C deficiency is present in 25% of men from low-income populations in the United Kingdom1 and 7.1% of healthy middle-income men and women in the U.S.2 Risk factors include alcoholism, smoking and fad diets and it presents with non-specific signs. Manifestations of scurvy like poor wound healing, petechial hemorrhages, follicular hyperkeratosis and bleeding gums have rarely been reported in the U.S. in the past 30 years, possibly because it is often unrecognized. This patient had an extensive history of alcoholism that contributed to his hemorrhagic manifestations. His response to high-dose vitamin C confirmed the diagnosis. Conclusion: Scurvy still exists in industrialized populations and may go unrecognized because of non-specific symptoms. Thorough history taking and physical examination in the proper clinical context may lead to a diagnosis and can avoid unnecessary hospitalization. Treatment is simple and readily available.

References

A rare case of Chilaiditi Syndrome with a long and complicated clinical course

Gupta Anupam,

INTRODUCTION Chilaiditi sign is a rare gastrointestinal anomaly, occurring in about 0.02 percent of the population, wherein a loop of the large intestine, usually transverse colon, is abnormally interposed between the liver and the right hemidiaphragm (1). When impacted, it may rarely cause a chronic, intermittent intestinal obstruction along with unusual symptoms like lung compression and cardiopulmonary compromise, which typically subside with conservative bowel decompression, laxatives, or surgical correction. Presented here is one of the first reported cases where Chilaiditi syndrome was uncorrectable and therefore fatal. CASE A 71 year old male with a history of hemorrhagic stroke in 2007 with residual left hemiplegia, aphasia, dysphagia status post gastrostomy, and seizure disorder; developed episodic abdominal distension, pain, dyspnea, projectile vomiting, constipation, hiccups, and a peculiar gurgling cough (presumably from reflux of gastric and lung secretions, and diaphragmatic irritation) in 2009, which all usually resolved with magnesium citrate administration via Gastric (G) tube. Extensive investigations for differentials of such intermittent abdominal obstruction and dyspnea were negative until a year later, when radiologists picked up Chilaiditi sign in his abdominal CAT scan (CT) in 2010. His multiple comorbidities prohibited surgery, while the anatomy was not amenable to laparoscopic colopexy. These episodes kept recurring every 2-3 months, until his presentation in 2013, with accompanied labored breathing. On examination, his vitals were stable. He had decreased breath sounds in right lower lung, with distended abdomen. Labs were normal. CXR showed right hemidiaphragm elevation with subdiaphragmatic loops of colon, and a right lower lobe opacity concerning for aspiration pneumonitis. CT Abdomen confirmed the Chilaiditi and showed a partial colonic malrotation with cecum abnormally positioned in the right upper quadrant. Patient was treated with conservative bowel decompression and magnesium citrate via G tube. Gradually, his breathing improved, gurgling cough resolved, and he passed a bowel movement. G tube feeds were resumed and patient was discharged in his baseline health to nursing home. However 10 days later he had a repeat aspiration, developed severe respiratory distress and died. DISCUSSION In the period from his stroke until the Chilaiditi onset, patient was able to keep his G tube feeds down. But had had multiple aspirations since the Chilaiditi reflux episodes started, which eventually proved fatal in combination with his aspiration predisposition from the residual neurologic deficit of stroke. CONCLUSION This unfortunate case offers a unique opportunity to study the chronic ramifications of an uncorrected Chilaiditi anomaly. REFERENCES 1. Chilaiditi D. Zur frage der hepatoptose und ptose im allgemeinen im anschluss an drei falle von temporärer, partieller leberverlagerung. Fortschritte auf dem Gebiete der Röntgenstrahlen. 1910;16:173–208
IVIG- dose dependent response in Toxic Epidermal Necrolysis Syndrome

INTRODUCTION Toxic epidermal necrolysis (TENS) is a severe mucocutaenous reaction very commonly triggered by medications. It manifests with fever, malaise, rash, bulla, and sloughing of the epidermis involving more than a third of the body surface area. It is associated with 35% morbidity. Mainstay treatment is supportive care and IVIG therapy.

CASE A 75 year old Caucasian man with a past medical history of allergies (to Penicillins, Sulfa, Doxycycline, and vancomycin), knee replacements with subsequent MRSA superinfection, and type 2 Diabetes Mellitus, presented with complaints of 1 week of painful red rash on his face and arms. These subsequently developed into vesicles and blisters on his lips, hands, arms and buttocks. There was no preceding fever, malaise or significant sunlight exposure. Prior to this, he was being treated for a knee infection with Daptomycin, followed by Bactrim for 2 weeks. He reported a similar but more severe episode 3 years prior with vancomycin use. On physical examination, vital signs were unremarkable. There were diffuse, ill defined, and tender, erythematous, macules and bulla of the face, trunk, back, upper, lower extremities, and buttocks (with positive Nikolsky’s sign) and skin sloughing noted in more than 40% of the body surface area. Blistering and crusting on his lips were present, but no oropharyngeal mucosa, ocular, or urethral involvement. The diagnosis of TENS secondary to Bactrim was made. Bactrim was discontinued on admission to the ICU. Supportive care was initiated with sterile handling, wound care, fluid and electrolyte repletion, nutritional support, pain control, and reverse-isolation for prevention of super-infections. Due to the strong evidence of systemic glucocorticoid-induced risk of sepsis, decreased rate of epithelialization, and overall increased morbidity and mortality (particularly in patients with TENS), the patient was started on intravenous immunoglobulin (IVIG) therapy. Unexpectedly, after 4 days of IVIG at 1gm/kg/day, there was no improvement in his skin lesions. As a result, IVIG dosage was increased to 2gm/kg/day with significant symptomatic improvement.

DISCUSSION Lack of response to the initial IVIG infusion dose was attributed to suboptimal doses of IVIG especially with poor prognostic factors such as older age, greater extent of skin detachment, and greater length of symptoms prior to initiation of therapy.

CONCLUSION Early infusion of high dose IVIG is associated with early improvement and decreased TENS- associated morbidity and mortality

REFERENCES
**INTRODUCTION:** Ovarian hyperstimulation syndrome (OHS) is the most severe iatrogenic complication of modern fertilization methods. The pathogenesis of ovarian hyperstimulation syndrome is unknown but is generally believed to represent the overproduction or altered expression of vasoactive substances of ovarian origin that are critical for follicle release or neovascularization of the developing corpus luteum.[1] OHS can range from mild to severe based on the severity of symptoms, signs, and laboratory findings. In its most severe presentation, OHS is characterized by massive ovarian enlargement, ascites, as well as hemoconcentration caused by intravascular volume depletion, and in some patients, pleural and/or pericardial effusion.[2]

**CASE:** This is a 31 y/o lady who presented with progressive dyspnea of 4 days duration. Six days prior to admission, she went to and in-vitro fertilization (IVF) clinic where she had her eggs harvested, and received a gonadotropin releasing hormone (GnRH) agonists to induce ovulation. In the emergency department, her vital signs were: blood pressure 127/78, pulse 126 bpm, respiratory rate 16, and 99% oxygen saturation on room air; she was in moderate distress. Physical exam showed, decreased air entry on the right lower lung base, dullness to percussion, positive egophony, and decreased tactile fremitus suggestive of pleural effusion. Her abdominal exam revealed a soft, non-tender, non-distended abdomen with no fluid wave, no hepatosplenomegaly, and no shifting dullness. No leg edema was noted. Lab studies showed a white blood cell count of 26.0, no bands, hemoglobin 16.9, hematocrit 50.9, red blood cell count 6.15, platelets 527, albumin 3.5, and electrolytes showed sodium 128 and potassium 5.0. Her chest X-ray showed a large right-sided pleural effusion, and her CT of abdomen/pelvis demonstrated enlarged ovaries with follicles and cysts, and moderate to large amount of free fluid in pelvic region. She was diagnosed with OHS, treated in the intensive care unit with chest tube drainage, and improved clinically.

**DISCUSSION:** Ovarian stimulation with injectable gonadotropins is commonly used for assisted reproductive treatments, such as IVF. A recognized complication of gonadotropin use is OHS. Early diagnosis and treatment is critical in severe cases such as this patient. **CONCLUSION:** OHS needs to be suspected in a young patient undergoing IVF who presents with increasing shortness of breath. **REFERENCES:** [1] Am J ObstetGynecol 1999;180:1468-71. [2] The Pathogenesis of the Ovarian Hyperstimulation Syndrome Ursula Brigitte Kaiser, M.D.N Engl J Med 2003; 349:729-732
Amulya Belagavi,

Introduction  Asbestosis related neoplastic and non neoplastic diseases of the lungs and pleura range from pleural effusion and pleural plaques to lung cancer and malignant mesothelioma. Pleural effusions are typically hemorrhagic exudates of mixed cellularity but do not always contain asbestos bodies.

Case  A 55 year old male from Dominican republic, living in the United states for the past 20 years presented with right sided pleuritic chest pain that started 5 days prior to admission. The pain was associated with shortness of breath and dry cough. He did not complain of fever, hemoptysis, or weight loss. There was no exposure to pulmonary toxins, drugs or recent travel. On physical exam, his vitals were stable. There was dullness and decreased air entry in the right lung base without digital clubbing. Chest X ray confirmed moderate right pleural effusion. CT Angiogram of the chest ruled out pulmonary embolism. The patient was placed in isolation until 2 sputum samples were negative for acid fast bacilli. Thoracentesis was done with removal of 600 ml of amber colored pleural fluid that was exudative in nature, cytology being negative for malignant cells and asbestos bodies. Investigations for rheumatological conditions were done with ANA screen being negative, rheumatoid factor being positive with a titer of 1:64, ESR of 26 and CRP of 34. On reviewing the CT films, pleural plaques were noted bilaterally along with linear comet tail of rounded atelectasis. The possibility of asbestosis was then entertained from the features of the CT scan consistent with asbestosis.

Discussion  The pleural plaques were not identified in the initial CT scan. It was only noted on subsequent review of the films. Pleural effusions are the earliest pleural based phenomena in asbestosis. It is important to consider asbestosis as a differential in a patient with unilateral pleural effusion even when there is no definite history of exposure since there is increased risk of developing malignant pleural mesothelioma.

Conclusion  Pleural plaques may be easily missed if one is not looking for them in the subpleural space. Despite recent advances in the treatment of other types of cancer, patients with mesothelioma currently face a poor prognosis. Therefore, it is highly important to develop an early diagnostic method with the greatest challenge on screening techniques to detect the disease at a subclinical stage.

Allen Test: Is It Really Necessary Before Transradial Catheterization?

Marinos Charalambous,

Background: The Allen test (AT) was described in 1929, by doctor Edgar V. Allen, as a method of diagnosing thromboangiitis obliterans. With the advent of radial artery grafting, surgeons started using this method to assess for adequate hand perfusion, before harvesting the radial artery. The use of AT for transradial catheterization (TRC) was recommended as a way to identify those patients that are at risk for the extremely rare complication of hand ischemia. Currently, there is poor evidence to support routine use of AT for all patients undergoing TRC. Our aim is to examine if TRC can be safely performed without this test. Methods: We performed a total of 3006 transradial catheterizations between January 1st, 2006 and December 31st, 2009. Baseline characteristics of our patients and all procedural information were collected through a computerized database and close follow up of the current study population was performed until June 2013. We evaluated the incidence of asymptomatic radial artery occlusion in patients undergoing TRC for the first time (Group I) and in patients undergoing repeated TRC (Group II). In both groups we also examined the incidence of symptomatic hand ischemia. Results: In total, 2,817 patients (96.9%) underwent a single TRC (Group I) and 91 patients (3.1%) underwent two or more TRC (Group II). While the incidence of asymptomatic radial artery occlusion increased from 0.5% in Group I to 3.3% in Group II, there were no cases of symptomatic hand ischemia either on periprocedural or long term follow up. Conclusion: More than 3000 catheterizations were performed safely in our center without the Allen test and that included 91 patients that underwent repeated catheterizations from the same artery without having any signs of hand ischemia. The incidence of hand ischemia is very rare. Even in the case where radial artery occlusion occurs, this is mostly asymptomatic due to collateral circulation. Based on our experience, we conclude that initial or repeated radial artery catheterization can be safely performed without the Allen test. REFERENCES 1. Ghuran AV, Dixon G, Holmberg S, de Belder A, Hildick-Smith D. Transradial coronary intervention without pre-screening for a dual palmar blood supply. Int J Cardiol 2007;121:320 322. 2. Greenwood MJ, Della-Siega AJ, Fretz EB, et al. Vascular communications of the hand in patients being considered for transradial coronary angiography: is the Allen’s test accurate? JAm Coll Cardiol 2005;46: 2013–7. 3. Agostoni P, Biondi-Zoccai GG, de Benedictis ML, et al. Radial versus femoral approach for percutaneous coronary diagnostic and interventional procedures: systematic overview and meta-analysis of randomized trials. J Am Coll Cardiol 2004;44:349 –56.
Marinos Charalambous,

Background: The Transradial Approach (TrA) is increasingly used as an alternative to transfemoral approach for coronary interventions but despite its advantages, there are several factors that limit its widespread use. One of those, is the anatomical variations of radial artery (RA) that are considered to be a major contributor to difficulties in the learning curve and in failures. In particular, radial loop is considered by many as an important cause of access failure. Methods: We performed 4970 transradial catheterizations between Jan 2006 and Dec 2012. Baseline characteristics, procedural success rates and major complications were recorded. We examined the incidence of RA loop and identified the rate of procedural failure that was attributed to this anatomic variation. We routinely tried to gain access from the right RA. If unsuccessful our next approach was the left RA. We only proceeded to femoral access, if we were unable to perform the procedure through both radial arteries. Results: We identified 21 cases of RA loop in 4970 transradial procedures. 3 cases (14.2%) were bilateral RA loops. In 9 cases (42.9%), catheterization was successfully completed by overcoming the loop and without the need to change access site. In 9 cases (42.9%) catheterization was successfully completed through a second access site which was always the left RA. In 3 cases (14.2%), both right and left radial access was unsuccessful and transition to right femoral access was required. The prevalence of RA loop in our cathlab was 0.4%. If a RA loop was encountered, there was a 42.9 % chance of successfully performing the procedure without the need to change the access site. There was an 85.8% chance of completing the procedure with one of the 2 radial arteries and only 14.2 % chance that an eventual femoral puncture would be required. Conclusion: In our opinion, RA loop is not a contraindication for transradial catheterization and efforts should be made to overcome the loop since there is > 40 % chance to overcome the loop. If this is not succesfull, contralateral RA should be tried, since there is <15% chance to encounter a bilateral artery loop and > 85 % chance to complete the procedure through radial approach. References: 1. Sarji R, Sricharoen N. Beyond the bend: a literature review and case report of radial artery loop. J Invasive Cardiol. 2011 Nov;23(11):E271-2. 2. Valsecchi O, Vassileva A, Musumeci G, Rossini R, Tespili M, Guagliumi G, Mihalcsik L, Gavazzi A, Ferrazzi P. Failure of transradial approach during coronary interventions: anatomic considerations. Catheter Cardiovasc Interv. 2006 Jun;67(6):870-8.
Carotid Artery Stenting: Single Center Experience

Marinos Charalambous, Tatiana Michaelidou1, Elpidoforos Soteriades1, Savvas Constantinides1, Christos Christou1

Background: Recent, updated, guidelines for the management of patients with carotid artery disease have indicated carotid artery stenting (CAS) as an alternative to carotid endarterectomy for selected patient groups. The less invasive nature of CAS constitutes an important advantage, as we examine the potential of this treatment modality. Our single-center experience is presented. Methods: Carotid angiography was performed in all patients with a history of a transient ischemic attack (TIA) or Stroke who were admitted to the American Heart Institute from 1999 to 2011 as well as to those who had abnormal carotid Doppler. Following angiography, patients underwent percutaneous intervention for carotid artery stenosis based on specific criteria. CAS was performed among asymptomatic patients with more than 90% stenosis, whereas in symptomatic patients, CAS was performed if stenosis was 80% or higher. Results: A total of 106 patients underwent diagnostic angiography. The majority were male (77.3%). Mean age was 67.7 and ranged from 28 to 86 years old. Of those, 41.5% were smokers, 93.4% had hypertension, 38.6% had diabetes, and 85.8% had coronary artery disease confirmed by coronary angiography. Of those, 62.2% underwent carotid stenting (66 stents in 61 patients were used). Among patients with carotid intervention, 41% underwent CAS on the left and 59% on the right side. The majority had unilateral (91.8%) and 8.2% had bilateral stenting. Among patients with CAS, 23% had coronary artery angioplasty and 26.2% had coronary artery bypass surgery (CABG) either prior or after the CAS procedure. A protection device was used in 97% of the cases. One patient (1.6%) had TIA in the lab that resolved within minutes and there was one common carotid artery dissection (1.6%) treated with an additional stent. No strokes or deaths were noted up to 2 years of follow up. Conclusion: Our 12 year experience shows that CAS, when performed by experienced operators, with proper selection of patients, can be accomplished successfully with a very low complication rate and provide a safe and less invasive alternative to carotid endarterectomy. References: 1. Brott TG, Halperin JL, Abbara S et al. Guideline on the Management of Patients With Extracranial Carotid and Vertebral Artery Disease: Executive Summary. J Am Coll Cardiol. 2011 Feb 22;57(8):1002-44. 2. Brott TG, Hobson RW 2nd, Howard G, Roubin GS, et al. (CREST Investigators). Stenting versus endarterectomy for treatment of carotid-artery stenosis. N Engl J Med. 2010 Jul 1;363(1):11-23
Introduction: Evans syndrome (ES) is a rare autoimmune disorder characterized by the presence of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP). Many of the cases diagnosed with Evans syndrome are later found to be thrombotic thrombocytopenic purpura (TTP). TTP is a life-threatening condition characterized by microangiopathic hemolytic anemia, thrombocytopenia and multiorgan failure. Only a minority of patients presents with classic symptoms. The management of these conditions varies significantly. Case presentation: A 31-year-old African-American female presented to the emergency department with 7 days of fatigue, abdominal discomfort, difficulty concentrating and tremors. On physical exam she had ecchymosis and petechial rash in her oral mucosa and lower extremities. Fine tremors were noticed on her upper extremities. Laboratory findings were remarkable for low hemoglobin at 8.5g/dl, white blood cells: 10x3/cumm, platelet: 13x3/cumm, LDH: 3067 U/L, indirect bilirubin 1.5 mg/dL, reticulocyte count: 13.8%, haptoglobin: <8mg/dL, direct Coombs test was positive. The differential diagnosis of Evans syndrome versus TTP was considered. However, the presence of schistocytes on the peripheral smear favored the diagnosis of TTP and plasmapheresis was initiated. Further laboratories were positive for fibrinogen degradation product, supporting the presence of microangiopathic hemolysis. HIV, hepatitis B, hepatitis C, parvovirus, malaria smear were negative. ANA titer were 1:640 (nucleolar and speckled pattern), anti-double stranded DNA and anti-histone antibodies were negative, complement C3 and C4 were normal, anticardiolipine and Sjogren antibodies were negative. VWF PROTEASE ACTIVITY was <1 %. The patient clinically improved after multiple sessions of plasmapheresis and blood transfusions. After 10 days of hospitalization, the patient was discharged with a platelet count of 242 x103 and normal hemoglobin and LDH levels, without persistent hemolysis. Discussion: This case illustrates a rare presentation of TTP and autoimmune hemolytic anemia. The differential diagnosis was Evan’s syndrome, although the presence of microangiopathic hemolytic anemia drove us to the decision to begin immediate plasmapheresis, which was then supported by a low protease activity. Conclusion: It is important to acknowledge that Evan’s syndrome is a rare condition and TTP should always be considered a possibility. Recognition of TTP is important in order to establish prompt and appropriate management. References 1. Mannucci PM, Peyvandi F. TTP and ADAMTS13: When Is Testing Appropriate? Hematology Am Soc Hematol Educ Program. 2007:121-6. . James N. George Ten patient stories illustrating the extraordinarily diverse clinical features of patients with thrombotic thrombocytopenic purpura and severe ADAMTS13 deficience. Journal of Clinical Apheresis Volume 27, Issue 6, pages 302–311, 2012 . Han-Mou Tsai. Disorders of the Platelets Thrombotic Thrombocytopenic Purpura and the Atypical Hemolytic Uremic Syndrome. Hematology/Oncology Clinics of North America, 2013-06-01, Volume 27, Issue 3, Pages 565-584
Choose wisely companion” for high risk patients? –A case study of atypical chest pain with elevated Troponins

Yingzi Deng

Introduction: Recent “Choosing Wisely” initiative by the American Board of Internal Medicine Foundation developed lists of “Five Things Physicians and Patients Should Question” in recognition of the importance of physician and patient conversations to improve care and eliminate unnecessary tests and procedures. However, most of the recommendations are for healthy population or lower risk patients. We are presenting a case of atypical chest pain with elevated troponin levels in an attempt to discuss the necessity of “Choosing wisely” in high risk patients. Case: This is a 44-year old female with past medical history of hyperlipidemia, hypertension and coronary artery disease, who underwent single vessel CABG surgery in 2009. She is a current smoker who has smoked half pack per day for 30 years. In addition, she consumes 20 drinks per day and more than $100 worth of cocaine daily. Since August of 2012, she has presented to ER for left-sided chest pain and discomfort 4 times and resulted in three hospitalizations. Each time her urine drug screening test was positive for cocaine and she admitted to heavy alcohol drinking prior to each hospitalization. On all three admissions, her EKG remained unchanged from her prior ECGs on record: normal sinus rhythm, septal infarct of undetermined age and no acute ST-T changes. Her serial troponins were elevated moderately. She received therapy under ACS protocol for her chest pain and has been worked-up for possible ischemia/restenosis. Two echocardiograms and one Lexi-scan have been done all negative for ischemia. She was treated for alcohol withdrawal and cocaine and was discharged home. Discussion: This patient suffered several episodes of chest pain secondary to cocaine abuse, which is a well-known drug that can cause coronary spasm. Due to her high risk profile for ACS, each time she was hospitalized and received multiple tests for her chest pain including serial ECG, cardiac enzymes, and cardiac imagine tests. However, studies have shown that cocaine users with a normal or non-diagnostic ECG are less likely to experience cardiac events, but no “Choosing wisely” recommendations are currently available for such a patient population. There is an imminent need for studies evaluating tests that could help risk stratification of high risk patients and could significantly avoid unnecessary hospitalizations and reduce health care costs. References 1. Choose Wisely. http://www.choosingwisely.org/doctor-patient-lists/ 2. Hendel RC, Ruthazer R, Chaparro S et al. Cocaine-using patients with a normal or nondiagnostic electrocardiogram: single-photon emission computed tomography myocardial perfusion imaging and outcome. ClinCardiol. 2012 Jun; 35(6):354-8. 3. Radensky PW, Hilton TC, Fulmer H et al. Potential cost effectiveness of initial myocardial perfusion imaging for assessment of emergency department patients with chest pain. Am J Cardiol. 1997 Mar 1;79(5):595-9.
Hemiataxia-hypesthesia syndrome

Introduction: Lacunar strokes classically present as: pure motor hemiparesis, pure sensory stroke, ataxic hemiparesis, sensory-motor syndrome and dysarthria-clumsy hand syndrome. More than twenty other lacunar syndromes have been described. Hemiataxia-hypesthesia is one such variant. Case: A 74yo gentleman, with a previous Karnofsky score of 90 and history of hypertension and chronic bronchitis, came to the emergency room with complaint of unsteady gait, slurred speech and difficulty in writing since one day. On the night before admission, while going to the bathroom, he was unable to coordinate his gait, and had to use the support of the wall to prevent a fall. Next morning, symptoms improved slightly and he was able to drive himself to the grocery store, where he experienced slurring of speech. Later in the day, he had difficulty signing a check and could not shave using his dominant right hand. He also noticed some perioral numbness on the right side, but did not have any weakness or vertigo. On admission, his vitals were: BP-188/84, T-97.6F, Pulse-81, RR-16, SpO2-98% on room air. Neurological exam: Motor – 5/5 power in all four extremities, Sensory - Diminished sensation to touch, pain in the maxillary & mandibular distribution on the right side of the face, right upper arm & right lower arm; DTR 2+ in all extremities; Cerebellar signs: Gait-normal, Finger-to-nose: positive for past pointing on right, Romberg test: negative, but he was unable to do Tandem walking. Remainder of physical exam was normal. CT head without contrast showed an age-undetermined radiolucenty in the left thalamus without any haemorrhage. He was started on Aspirin and Atorvastatin. MRI brain was done, which showed acute left thalamic lacunar infarct. By next morning, all his symptoms had resolved completely, except for peri-oral numbness, which improved only partly. Twenty-four hours after admission, he was started on Amlodipine 5mg, and discharged home. Discussion: Hemiataxia-Hypesthesia is postulated to be caused by small infarcts in lateral thalamus (thalamogeniculate territory) or the immediately adjacent internal capsule (anterior choroidal artery territory). Ataxia and other cerebellar signs in such cases, would make one think of cerebellar infarcts, but the associated hypesthesia points towards the thalamus. Conclusion: Ataxia can be the presenting sign of thalamic infarcts. Acute hemiataxia with hypesthesia should make one consider the possibility of a thalamic infarct. References: 1) Melo TP, Bogousslavsky J. Hemiataxia-hypesthesia: a thalamic stroke syndrome. J Neurology, Neurosurgery, and Psychiatry. 1992;55:581-584 2) Lee N, Roh JK, Myung H. Hypesthetic Ataxic Hemiparesis in a Thalamic Lacune. Stroke 1989;20:819-821 3) Carrera E, Michel P, Bogousslavsky J. Anteromedian, Central, and Posterolateral Infarcts of the Thalamus: Three Variant Types. Stroke. 2004;35:2826-2831
Arterial thrombosis while on Heparin drip in Prothrombin G20210A gene mutation carrier

Komal Dumaswala

INTRODUCTION: Certain inherited hypercoagulable states can modestly increase the risk for arterial thrombosis1, in addition to the increased risk for venous thromboembolism (relative risk 2-5)2. CASE: An 86 year old female presented with progressive shortness of breath since month. It was initially only exertional but later occurred even at rest. She had no chest pain, cough, palpitation, fever. She was admitted with similar complaint 2 months ago when pulmonary embolism was ruled out by negative CT chest angiogram and cardiac work up was negative. Her history was significant for pulmonary embolism and right ventricular thrombus diagnosed 7 years prior, when she received Coumadin for 6-7 months followed by aspirin, which she was taking until one week before admission when she suffered an episode of epistaxis requiring nasal packing. She had a strong family history for pulmonary embolism. On admission, she was hemodynamically stable, saturating well on nasal cannula. CT angiogram showed multiple bilateral pulmonary emboli and was therefore restarted on anticoagulation. Her lower extremity Doppler showed a thrombus in right peroneal vein. Given this was her second episode of pulmonary embolism, she would require long-term anti-coagulation. Due to her recent episode of epistaxis, an IVC filter was placed and heparin drip was continued. During this time, she developed acute right lower extremity pain with cold and pale foot. CT angiogram showed right popliteal artery occlusion with some involvement of right superficial femoral, right anterior tibial artery and left anterior tibial arteries. A hypercoagulability work up was negative for Protein C & S deficiency, Factor V Leiden and lupus anticoagulant. Echocardiogram with bubble study was negative for intra-cardiac shunt. As she was on heparin, measuring anti-thrombin III levels was not reliable, so Prothrombin gene mutation was ordered which showed Heterozygous G20210A MUTATION in the prothrombin/Factor II Gene.

DISCUSSION: Prothrombin G20210A gene mutation heterozygotes are at increased risk for arterial thrombosis1, however development of such thrombi while on therapeutic anticoagulation has not been studied. CONCLUSION: This case highlights the possibility of developing arterial thrombus in prothrombin gene mutation carriers, especially in elderly patients, which could be a combination of thrombophilic state and age related arteriosclerotic predisposition. Also, striking is the development of such arterial thrombi while on therapeutic anticoagulation.

Acute Fatty Liver of Pregnancy

Devendra Enjamuri

Introduction: Acute fatty liver of pregnancy (AFLOP), characterized by microvesicular fatty infiltration of hepatocytes, is a disorder which is unique to human pregnancy. It was described in 1940 and was initially thought to be universally fatal. However, early diagnosis and prompt delivery have dramatically improved the prognosis, and maternal mortality should now be the exception rather than the rule. Case: 24 year old pregnant lady from India G1 P0, at 35 weeks presented with vaginal bleeding, headaches, blurry vision and yellow discoloration of skin for 1 day. Fetal monitor showed recurrent decelerations. Initial labs showed WBC count of 18.4/cumm, hemoglobin of 11.1g/dl, Platelet count -94/cumm glucose of 50mg/dl, creatinine of 2.82 mg/dl, total bilirubin- 14.8, AST-230,ALT-222, Alkaline phosphatase – 529 LDH-2224 , PT-63.7, INR-5.74, APTT-78.5, Uric acid-10.6mg/dl ..Hepatitis A, B ,C , E virus serology is negative. She underwent C-section as an emergent procedure and patient was intubated during surgery. Patient was transferred to ICU after surgery and she remained unresponsive for 3 days secondary to hepatic encephalopathy. She was transferred to Liver Transplant facility for advanced care. Her liver enzymes and functions initially worsened and gradual improvement was noted after 5 days after which patient was extubated. Discussion: AFLOP is a rare clinical entity unique to pregnancy that can lead to hepatic failure and encephalopathy and, if the diagnosis is delayed, to death for the baby and the mother. The characteristic histological picture demonstrates microvesicular fatty infiltration of hepatocytes. AFLOP is a disease of the third trimester of pregnancy. The most significant clinical findings are nausea or vomiting, abdominal pain, jaundice, hepatic encephalopathy, significant hypoglycemia,elevated WBC count increased transaminase levels, decreased platelet count, increased prothrombin time, and renal failure. Hypertension and proteinuria are common. Liver biopsy is not always necessary for diagnosis but may be useful in atypical cases (1). Although the mechanism is not clear, polyuria and polydipsia are noted in about 5% of cases and are almost pathognomonic for AFLOP once diabetes has been ruled out. Maternal mortality has decreased from 90% to less than 10% over the past 30 years. Swansea criteria (vomiting, leukocytosis, elevated transaminases,elevated bilirubin,polydipsia, coagulopathy etc) is used to diagnose AFLOP clinically. Delivery results in resolution of symptoms and hepatic recovery for the mother, but close monitoring is required for the child because of the risk of an associated fatty acid oxidation defect. (2) Reference: 1. Bacq Y, Riely CA, Acute fatty liver of pregnancy: the Hepatologist"s view ,Gastroenterologist. 1993 ;1(4):257-64. 2. Leila Kia, and Mary E. Rinella, Interpretation and Management of Hepatic Abnormalities in Pregnancy , Clinical Gastroenterology and Hepatology 2013 ;11: 1392-1398
**Drexel - St. Peter's**

**Vector-Borne Infection Case Complicated by Social Factors**

Peter Fish

Introduction Tick-borne diseases have an insidious onset and variable symptoms. The often unnoticed bite can co-transmit multiple pathogens. Mild initial symptoms belie the consequences of improperly treated cases. Even with a proper diagnosis and treatment plan, failure to address social issues can harm a patient. Case A 58-year-old Ukrainian male presented to the hospital complaining of 4 days of fatigue and constant, diffuse, dull 6/10 abdominal pain. He attributed this to a plum that had “gone down uncomfortably”. He indicated his pain was throughout his abdomen, did not radiate, and was unaffected by food, defecation, or position. He took no medications, had no appetite, and vomited once. The pain woke him up, and he had been feverish. His history and physical were complicated by his stoicism, his poor English, and further by the fact that though his primary language was Ukrainian, providers kept using Russian translators. Though the patient denied medical problems, his medical record revealed CHF (EF 20%), an AMI s/p CABG, atrial fibrillation, hyperlipidemia and two recent visits to the ED for a rash on his arm (diagnosed as cellulitis, treated with Ceftriaxone). Physical exam revealed a large pulse deficit, an irregularly irregular heartbeat and diffuse abdominal pain, but no rash. At this point, the differential included mesenteric ischemia, atypical angina, and GERD. Labs Platelets: 51, LDH: 751, UA: 2+bilirubin, Blood Smear: “Intracellular organisms c/w Babesia”. Labs for Erlichia and Lyme were sent out. The patient was admitted and empiric treatment with Atovaquone and Azithromycin were initiated to cover Babesia and Erlichia. At discharge, the Lyme titer was also returned as positive, so Doxycycline was added. Telephone follow-up revealed that the patient had not filled the prescriptions because he could not afford them (Atovaquone costs $200/week), whereupon charity care was arranged and he was able to resume his complete course of antibiotics. At follow-up, his symptoms had resolved.

**Discussion**

This case illustrates the dangers of anchoring heuristics when making differentials, assumptions about ethnicities and languages, the value of combing through medical records, effective teamwork, and the importance of patient follow-up. The vigilance of the laboratory staff immediately found the source of the patient’s symptoms by initiating a thin prep on their own volition—a disease process that was not even on the initial differential. Language barriers, the patient’s stoic personality, and vague symptoms further obscured the medical presentation and had confounded physicians during previous visits to the ED resulting in improper care. Proper follow-up enabled social work to procure necessary medications.

**REFERENCES**

Pulmonary Hemorrhage Masquerading as Pneumonia in a case of Myelodysplastic Syndrome

Vedavyas Gannamani

INTRODUCTION: Myelodysplastic syndrome encompasses a heterogeneous group of closely related clonal hematopoietic disorders which can present with a multitude of clinical manifestations. Pneumonia is a common complication seen in these patients1, while others like pulmonary hemorrhage, alveolar proteinosis2 are uncommon. CASE A 68 year old female with past history of COPD, mycobacterium avium infection and untreated MDS presented to the emergency department with progressive shortness of breath and productive cough with pink colored sputum of one week duration. She had no fever, chest pain or palpitations. Vital signs include temp: 98.8F, BP: 148/78mm Hg, HR 108, RR 24, O2 Saturation: 98% on 3L nasal cannula. On chest and lung examination she had tachycardia, tachypnea, aegophony and crackles at left lung base. Initial lab studies showed WBC 1.2, Hemoglobin 6.9, Platelets 11, BUN 23, Serum Creatinine 0.89, lactic acid 2.2, PT 13.1, INR 1.19. Chest x-ray (CXR) showed left lower lobe infiltrate suggestive of pneumonia. She was initially treated with antibiotics for healthcare associated pneumonia. She received multiple blood and platelet transfusions. However she continued to have dyspnea, cough and developed hemoptysis and respiratory insufficiency requiring BiPaP. Further workup including Doppler ultrasound of lower limbs, CT angiography of chest, 2D Echocardiography and sputum culture were inconclusive. Bronchoscopy and lung biopsy were not attempted due to low platelet count. Repeat CXR showed new infiltrates involving both lower lobes and large left-perihilar infiltrate. Treating reactivation of MAI and other opportunistic infections did not improve the clinical situation and her hemoptysis worsened. She had 2 episodes of dark stools further during the course of hospitalization. Aggressive management with platelet transfusion for her severe thrombocytopenia keeping the platelet count ≥ 50,000 resulted in improvement of her clinical symptoms of hemoptysis, cough & dyspnea which was highly suggestive of pulmonary hemorrhage.

DISCUSSION: Pulmonary hemorrhage is a serious complication seen in patients with Myelodysplastic syndrome (MDS). It might obscure the clinical picture of other pulmonary complications seen in MDS like infections and impede diagnostic procedures as well. Prompt management with platelet transfusions would potentially reverse the course while avoiding unnecessary use of antibiotics.

A rare case of Chilaiditi Syndrome with a long and complicated clinical course

Anupam Gupta

INTRODUCTION Chilaiditi sign is a rare gastrointestinal anomaly, occurring in about 0.02 percent of the population, wherein a loop of the large intestine, usually transverse colon, is abnormally interposed between the liver and the right hemidiaphragm(1). When impacted, it may rarely cause a chronic, intermittent intestinal obstruction along with unusual symptoms like lung compression and cardiopulmonary compromise, which typically subside with conservative bowel decompression, laxatives, or surgical correction. Presented here is one of the first reported cases where Chilaiditi syndrome was uncorrectable and therefore fatal. CASE A 71 year old male with a history of hemorrhagic stroke in 2007 with residual left hemiplegia, aphasia, dysphagia status post gastrostomy, and seizure disorder; developed episodic abdominal distension, pain, dyspnea, projectile vomiting, constipation, hiccups, and a peculiar gurgling cough (presumably from reflux of gastric and lung secretions, and diaphragmatic irritation) in 2009, which all usually resolved with magnesium citrate administration via Gastric(G) tube. Extensive investigations for differentials of such intermittent abdominal obstruction and dyspnea were negative until a year later, when radiologists picked up Chilaiditi sign in his abdominal CAT scan(CT) in 2010. His multiple comorbidities prohibited surgery, while the anatomy was not amenable to laparoscopic colopexy. These episodes kept recurring every 2-3 months, until his presentation in 2013, with accompanied labored breathing. On examination, his vitals were stable. He had decreased breath sounds in right lower lung, with distended abdomen. Labs were normal. CXR showed right hemidiaphragm elevation with subdiaphragmatic loops of colon, and a right lower lobe opacity concerning for aspiration pneumonitis. CT Abdomen confirmed the Chilaiditi and showed a partial colonic malrotation with cecum abnormally positioned in the right upper quadrant. Patient was treated with conservative bowel decompression and magnesium citrate via G tube. Gradually, his breathing improved, gurgling cough resolved, and he passed a bowel movement. G tube feeds were resumed and patient was discharged in his baseline health to nursing home. However 10 days later he had a repeat aspiration, developed severe respiratory distress and died. DISCUSSION In the period from his stroke until the Chilaiditi onset, patient was able to keep his G tube feeds down. But had had multiple aspirations since the Chilaiditi reflux episodes started, which eventually proved fatal in combination with his aspiration predisposition from the residual neurologic deficit of stroke. CONCLUSION This unfortunate case offers a unique opportunity to study the chronic ramifications of an uncorrected Chilaiditi anomaly. REFERENCES 1. Chilaiditi D. Zur Frage der hepatoptose und ptose im allgemeinen im Anschluß an drei Fälle von temporärer, partieller Leberverlagerung. Fortschr Röntgenstrahlen 1910;16:173–208
Drexel - St. Peter's

Intractable Diarrhea Associated with Olmesartan

Alejandra Gutierrez, Shivank Madan MD, Marinos Charalambous MD

Introduction: Olmesartan is an angiotensin II receptor antagonist, introduced in 2002 and is currently used as an antihypertensive medication. In 2012, reports suggested an unusual side effect described as severe sprue-like enteropathy. These patients have intractable chronic diarrhea that can’t be explained by any other disease process but resolves after discontinuing olmesartan. Case: A 67 year old male with hypertension presented with severe diarrhea associated with a 70 pound weight loss over a 12 months period. His symptoms did not resolved despite a gluten free diet, treatment with antibiotics for suspected infectious causes and TNF-a inhibitors for suspected Crohn’s disease. Despite multiple colonoscopies and upper endoscopies the etiology of his condition remained undiagnosed for about one year. Identification of a possible relation of his intractable diarrhea with olmesartan resulted in discontinuation of the drug. This resulted in immediate relief of his symptoms which confirmed the causal relationship between olmesartan and his diarrhea. Discussion: This illustrated a case of intractable diarrhea caused by olmesartan, The severe diarrhea resolved only after discontinuing olmesartan. There have been clinical reports supporting the association between olmesartan and severe sprue-like enteropathy. A study of 22 patients with unexplained severe enteropathy, symptomatically resembling celiac disease. These patients were referred to Mayo clinic for evaluation of presumed refractory celiac disease or unexplained sprue and were noted to lack IgA tissue transglutaminase and never responded to a gluten free diet. On the contrary, the symptoms of diarrhea and weight loss improved after discontinuation of olmesartan. These patients were taking olmesartan for months to years prior to the onset of diarrhea. Baseline intestinal biopsies revealed villous atrophy and variable degrees of mucosal inflammation. In 5 patients evidence of Microscopic colitis were also noted. Follow up biopsies were performed in the majority of patients after discontinuation of olmesartan and histologic recovery was observed. The mechanisms mediating this severe form of enteropathy are still unclear. Possible explanation could be the result of cell mediated immunity rather than type 1 hypersensitivity and/or the result of the inhibitory effect of angiotensin receptor blockers on TGF-b that may alter the intestinal immune homeostasis. Identification of this side effect, which is considered rare so far, will allow the study of these patients and will help in further understanding of the pathophysiology of this disabling drug reaction. References: 1. Olmesartan medoxomil. Warner GT, Jarvis B. Drugs. 2002; 62(9):1345-53; PMID:1207618. 2. Severe spruelike enteropathy associated with olmesartan. Rubio-Tapia A, Herman ML, Ludvigsson JF, Kelly DG, Mangan TF, Wu TT, Murray JA. Mayo Clin Proc. 2012 Aug; 87(8):732-8. PMID: 22728033
THE WATER BOTTLE SHAPE IN A MAN

Vivian Igilige

Introduction: Cardiac tamponade can be fatal if not recognized and treated promptly. The three most common causes of cardiac tamponade are neoplastic disease, idiopathic pericarditis and renal failure. Others include radiation, trauma, autoimmune, drugs and metabolic. Beck’s triad consists of hypotension, soft or absent heart sounds and jugular venous distention. CASE: This is a 47 years old man with history of non-small cell lung cancer, with extensive brain, lung and bone metastases, who presented with worsening shortness of breath for three days, which was aggravated by exertion and lying down and had no relieving factors. He also had orthopnea. On presentation, vitals were as follows BP 104/70, PULSE 80, TEMP 97 RR 28 O2 SAT 95 rmail. Physical exam was positive for cachexia, increased jugular venous distention, diminished breath sounds at lung bases, right more than left side, prescence of a parasternal heave. Chest Xray showed enlarged cardiac silhouette (WATER BOTTLE SHAPE), hazy opacity at the right lung base. EKG showed electrical alternans, sinus tachycardia and low voltage. Ultrasound showed large pericardial effusion. Echo showed right atrium and right ventricle diastolic collapse, large pericardial effusion and cardiac tamponade. Pericardiocentesi was done and 2.5 liters of fluid was drained. Cytology of the fluid was positive for malignant cells of (Adenocarcinoma). Considering the pathology, patient would get a pericardial window placed. DISCUSSION: It’s been estimated that 10% of patients with metastatic cancer will develop cardiac tamponade. In some patients, the classic clinical signs of Beck’s triad is absent, as in our patient. Pericardial fluid removal can be done by catheter pericardiocentesis, open surgical drainage with or without pericardiectomy (pericardial window) or video-assisted thoracoscopic pericardiectomy (VATS). (1, 2). Conclusion: In a study of patients with effusion who were clinically suspected of having hemodynamic compromise, 64% had tamponade. Pericardial effusion and subsequent cardiac tamponade may be missed if physicians are limited by traditional clinical signs (Beck’s Triad), they should be familiar with use of other subtle findings and the importance of other modalities that may potentially enhance diagnosis. (1). Reference: 1. Jacob S, Sebastian JC, Cherian PK, Abraham A, John SK., Pericardial effusion impending tamponade: a look beyond Beck’'s triad. Pericardial effusion impending tamponade: a look beyond Beck’s triad. Am J Emerg Med. 2009 Feb;27(2):216-9. doi: 10.1016/j.ajem.2008.01.056. 2. Uramoto H, Hanagiri T., Video-assisted thoracoscopic pericardiectomy for malignant pericardial effusion., Anticancer Res. 2010 Nov;30(11):4691-4.
Drexel - St. Peter's

Variant Lateral Medullary syndrome

Ovais Khan

INTRODUCTION Lateral medullary infarction (Wallenberg syndrome) is the most common and important syndrome related to intracranial vertebral artery occlusion. Vestibulocerebellar signs and symptoms are nearly always present in patients with lateral medullary infarcts. Case: 51 year old Caucasian male came to the Emergency room complaining of altered sensation in his right hand and foot for 4 days. At home he realized he could not feel the cold temperature of water bottle he was holding compared to his left hand. He also had a scratchy feeling in his throat with difficulty in swallowing solid foods but none with liquids. On examination patient was noticed to have altered temperature sensation in his right arm and right leg. No changes were noted in touch perceptions. Cranial nerves examination was normal. Romberg sign was positive, gait was normal; rest of the neurological examination was unremarkable. On admission his vitals were- BP: 227/139, HR-104, regular rhythm, RR- 20, Oxygen sat- 98% on room air. Labs were, WBC- 7.3, Hgb – 16.8, Hct- 48.9, MCV- 83.8, and Platelets – 206. BMP, Na – 139, K- 3.7, Cl-95, carbonate- 30, Anion gap- 14, BUN- 10, Cr- 0.62 and blood glucose of 398. Imaging: MRI report suggested sub-acute infarct in left medullary region. An MRA showed occlusion of the left vertebral artery with retrograde filling of the most distal aspect of the left vertebral artery and the posterior inferior cerebellar artery by the right vertebral. There was mild stenosis within the mid portion of the basilar artery. DISCUSSION: A diagnosis of lateral medullary syndrome was entertained. He was monitored for progression of symptoms and was treated with aspirin, antihypertensive medications such as ramipril, hydrochlorothiazide and hydralazine and appropriate management of diabetes with insulin NPH and aspart. Usual presentation of lateral medullary syndrome consists of constellation of vertigo, numbness of ipsilateral face and contralateral limbs, diplopia, hoarseness, dysarthria, dysphagia and ipsilateral Horner’s syndrome. CONCLUSION In my case the difference in presentation highlights the importance of looking into the subtle signs and symptoms of presentation of a medullary infarct. As it may be difficult to diagnose clinically as the spectrum of symptoms may vary, as in this case where retrograde filling of the most distal aspect of the left vertebral artery and the posterior inferior cerebellar artery by the right vertebral may have caused variation in the usual presentation. REFERENCES: 1. Kim JS, Lee JH, Lee MC. Patterns of sensory dysfunction in lateral medullary infarction. Clinical-MRI correlation. Neurology. 1997;49(6):1557–1563. [PubMed] 2. Neal M. Rao, MD, Tritia Yamasaki, MD, PhD, […], and Catherine Yim, MD, Neurohospitalist. 2013 April; 3(2): 98–99.
Liver Transaminase elevation Predicts More Severe Coronary Artery Disease in Octogenarians

Shivank Madan, Kiran Guthikonda, MD; Dinesh Singal, MD, FACC, FSCAI; Capecomorin Pitchumoni, MD, MACG, MACP, FACC.

Background: Non alcoholic fatty liver disease (NAFLD) is positively correlated with coronary artery disease (CAD). The objective of this study was to evaluate the relationship between serum transaminase levels (AST, ALT) and the prevalence of angiographic CAD in a cohort of octogenarians. Methods: In this IRB approved, retrospective cohort study, of the 524 octogenarians who underwent elective cardiac catheterization from 2008 to 2012, 373 had liver transaminase levels checked prior to catheterization, lacked a history of significant alcohol consumption, lacked a history of STEMI and were included. These were divided into 2 cohorts based on normal and elevated transaminase levels (ALT>55 U/L, AST>40 U/L). Baseline characteristics and coronary angiographic findings were recorded. Significant CAD was defined as >50% stenosis. CAD was subdivided into 1, 2 and 3-vessel CAD. Cohorts were compared with Chi sq & unpaired t tests, using STATA IC 12.1 software. Results: • Both cohorts had similar prevalence of traditional cardiovascular risk factors including age, sex, BMI, diabetes, dyslipidemia and smoking. • Elevated transaminase levels were found to be associated with: a) Greater prevalence of 3-vessel CAD. (34.6% vs 23.9% in normal cohort, p=0.038) b) Higher mean number of vessels involved. (Mean vessel score of 1.77+0.14 vs 1.45+0.08 in normal cohort , p=0.037) c) Increased prevalence of cerebrovascular disease, which nearly reached statistical significance (14.8% vs 8.4% in normal cohort, p=0.07) • No significant correlation was found between transaminase levels and single, double or left main CAD. Conclusions: • Elevated serum transaminase levels were predictive of greater prevalence of triple vessel CAD and a higher mean number of vessels involved. • Evidence of NAFLD in octogenarians as determined by elevated AST and ALT levels may indicate more severe CAD. Further studies are needed to evaluate these findings in other age-specific populations.
Obesity, not so bad in Octogenarians with Coronary Artery Disease: the “Obesity Paradox”

Shivank Madan, Kiran Guthikonda, MD; Dinesh Singal, MD, FACC, FSCAI; C.S. Pitchumoni, MD, MACG, MACP, FACC

Background: Obesity is increasing worldwide and is a major risk factor for hypertension, diabetes, dyslipidemia and cardiovascular (CVS) diseases [1]. However, recent studies have shown an obesity paradox in two important patient populations, a) those with established cardiovascular diseases [1] and b) cohorts of elderly patients [2]. In these two patient cohorts, overweight and obese patients have had better clinical outcomes than their normal weight counterparts. We aimed to study the association between Body Mass Index (BMI), and angiographic severity of coronary artery disease (CAD) in a cohort of octogenarians.

Methods: 524 octogenarians who underwent coronary catheterization at our hospital between 2008 to 2012 for various indications were included. The subjects were grouped according to BMI into three groups, BMI < 25, BMI: 25-30, and BMI > 30. Baseline characteristics and angiographic data were obtained from New Jersey Cardiac Catheterization Registry. A stenosis of >50% was defined as significant CAD. Variables were compared with chi-square tests, unpaired t tests and ANOVA using STATA software.

Results:
- Total number of study patients= 524
- Patients with CAD= 381 (72.7%)
- The three groups (BMI<25, n1=181), (BMI25-30, n2= 194 ) and (BMI>30, n3=149) were similar in age, race, history of prior congestive heart failure, hypertension and smoking status
- Across the three BMI groups, increase in BMI was associated with increased prevalence of diabetes: (30.4% vs. 33.5% vs. 45.6%, p=0.011) and dyslipidemia (72.9% vs. 76.8% vs. 85.9%, p=0.016)
- However, increase in BMI was associated with a decrease in prevalence of CAD in the three groups: 76.8% vs. 75.7% vs. 63.8% (p=0.010) and a decrease in prevalence of triple vessel CAD: 35.9% vs. 23.7% vs. 20.1% (p=0.002)
- Also, Octogenarians with CAD overall, had a lower mean BMI (27.4+ 0.29) when compared to patients without CAD (29.2+ 0.81)(p=0.013)
- The three groups of BMI were similar in terms of prevalence of left main coronary disease.

Conclusions: In our large cohort of “octogenarians”, although increase in BMI was associated with increased prevalence of diabetes and dyslipidemia, BMI was found to be inversely related to the prevalence of CAD and triple vessel disease. This indicates towards the existence of another “cardiovascular obesity paradox” in the elderly. One explanation could be that BMI is a measure of overall obesity and does not accurately measure visceral fat content or abdominal obesity which are probably more important cardiovascular risk factors.

References:
Adenocarcinoma of Unknown Primary Obscured by Hematologic Manifestations: A Case Report

Shripad Pande

Introduction: Hematologic paraneoplastic phenomena are known to occur with solid tumors. Disseminated intravascular coagulation (DIC) as a manifestation of cancer of unknown primary site obscuring the symptoms of tumor is a rare occurrence. Case Report: A 66 year-old woman noticed pain in central abdomen. About a month later, workup in an ER revealed positive fecal occult blood test. She was transfused with blood for hemoglobin of 6 g/dL. She had black stools and hematochezia in next the month. Computed tomographic (CT) scan of abdomen revealed colonic diverticula and retroperitoneal lymphadenopathy (RPLN). Colonoscopy and Esophago-gastroduodenoscopy (EGD) were normal. She required transfusions of blood again. She presented to us a monthly later for melena, hematuria and bleeding from mouth. CT abdomen revealed RPLN, and multiple renal and splenic wedge-shaped infarcts. Blood was transfused at our center. Outpatient RPLN core biopsy earlier in the same month revealed metastatic adenocarcinoma. Immunohistochemistry, EGD, push enteroscopy and colonoscopy didn’t reveal source of adenocarcinoma. Anemia with reticulocytosis, indirect hyperbiliurbinemia, schistocytes, thrombocytopenia, decreased haptoglobin, hypofibrinogenemia, positive fibrin degradation products (FDPs), prolonged PT and aPTT with intermittently active bleeding, in the presence of preserved kidney function led to the diagnosis of chronic DIC with microangiopathic hemolytic anemia (MAHA). In her Hospital course, she had acute embolic stroke in the MCA territory. Transthoracic 2D echocardiography (TTE) and transesophageal echocardiography (TEE) showed vegetations on the mitral valve. Repeated blood cultures, including cultures for HACEK organisms, and serologies for uncommon organisms were negative. She was diagnosed with nonbacterial thrombotic endocarditis (NBTE/marantic endocarditis) as a complication of chronic DIC. Subsequently, she developed proximal and distal deep vein thrombosis of right lower extremity, for which an inferior vena cava filter (IVC) was deployed. Discussion: DIC is a pathologic syndrome in which the manifestations are in large part a consequence of thrombin formation. Thrombin catalyzes activation and consequent consumption of fibrinogen and other coagulant proteins, along with production of fibrin thrombi. Tissue injury occurs in the malignant tumors in which procoagulant material, usually tissue factor, is expressed on circulating tumor cells or on vessel surface. Tissue factor, in conjunction with factor VII through IX, can activate the intrinsic and extrinsic pathway. Some other factors implicated are tumor cysteine protease, trypsin-like substances in prostate cancer, and mucin in gastrointestinal tumors. Conclusion: The index case had chronic DIC presumably from mucin in adenocarcinoma. Presence of malignancy was overshadowed by hematologic syndromes that eventually led to the discovery of malignancy.

“Why Can’t I Swallow my Food?!”

Shyam Patel

Introduction: Bulbar Myasthenia Gravis is a rare neuromuscular disorder, but has been reported. And it should be considered in patients presenting with symptoms of oropharyngeal dysphagia and change of voice. Case: A 66 y/o Caucasian male with history of hypertension on lisinopril for a number of years, presents to the Emergency Department (ED) with 3-4 weeks of subacute onset dysphagia, dysarthria, muffled voice, and perioral numbness while on a cruise on the Danube River. Vital Signs included Temperature of 96 F, pulse 71, blood pressure 136/71, respiratory rate 16, pulse ox 100% room air. Physical exam was significant for dysarthric speech, minimal erythema in the posterior pharynx, minimal soft palate movement, without tonsillar exudates, cervical lymphadenopathy, lip or tongue swelling. Cranial nerve examination was normal, and no focal neurological deficits were found. Lab studies for CBC, CMP, TSH were normal. He had ENT laryngoscopy which showed pooling of saliva in the posterior pharynx but no edema. He was initially treated as a case of angioedema due to lisinopril use. As his symptoms did not improve with standard treatment for angioedema, he had a CT scan and MRI which were negative. The differential then broadened and he tested (+) for Acetocholine Receptor Antibody but negative for Ganglioside GQ1B. EMG was also conducted and showed high repetitive stimulation of CN 7 on the right showed decrement between sets of stimulation that improved with time, which is highly suggestive of neuromuscular disease. He was given a Pyridostigmine challenge that showed improvement in movement of soft palate, and was placed on Prednisone 20mg and Pyridostigmine 60mg Three Times Daily. His condition improved after therapy. Discussion: Myasthenia Gravis is an autoimmune disorder that directs antibodies to the postsynaptic receptor that prevent stimulation of that nerve and nerve conduction. It can present as generalized or focal (ocular-bulbar). About 15% of cases are found to be ocular-bulbar. Diagnosis is made via history and physical with positive Anti-ACh antibodies or positive anti-MuSK antibodies. The first line therapy for mild to moderate disease is pyridostigmine whereas plasmaphoresis is reserved for more severe/generalized disease. Conclusion: This case highlights the importance of considering myasthenia gravis in patients that present with nasopharyngeal dysphagia and voice changes. Scherer K, Bedlack RS, Simel DL. Does this patient have myasthenia gravis? JAMA. 2005;293(15):1906-1914.
A case of first motor neuron disease after spinal manipulation

Sivagamavalli Ramasundaram

Introduction: Spinal manipulation is a technique used by chiropractors, physical therapists, and osteopathic physicians, primarily to provide relief for musculoskeletal pain related to the back and neck. It is a form of manual therapy that involves the movement of a joint beyond its usual end range of motion but not past its anatomic range of motion, an area that has been termed the "paraphysiologic zone." This movement of the joint is frequently accompanied by an audible cracking or popping sound. Minor complaints following spinal manipulation are common, with rates variably reported between 33 and 60 percent[1]. The most common serious adverse events associated with spinal manipulation include disk herniation, the cauda equina syndrome, and vertebrobasilar accidents. Estimates of the incidence of these complications range from 1 per 2 million to 1 per 400,000 manipulations[2].

Case: A 42 year old male from Dominican Republic, with no significant past medical history presented to the emergency room with complaints of right arm weakness, numbness and tingling sensation for two days. The night prior to admission, his family noticed that his right eye was drooping and hence was brought to the hospital. In the Emergency room, patient had right eye ptosis, myosis, right arm strength 4/5 and diminished sensation to light touch. The rest of the physical examination was normal. Laboratory studies were normal. Initially a cerebrovascular accident was suspected and hence a CT scan was done which was unremarkable. In retrospect, the symptoms began one day after the patient had his ‘neck adjusted’ by his chiropractor. A neurology consult was obtained and a first order neuron disease was diagnosed. The patient’s symptoms improved gradually and completely resolved.

Introduction: Staphylococcal scalded skin syndrome is rarely found in adults. Distinguished by skin desquamation and erythema. Caused by staphylococcal exfoliative toxins, specifically phage group I and III and histologically found to have epidermal splitting of the stratum corneum. In adults can have almost 60% mortality. It can be associated with immunosuppression, renal failure, malignancy or alcohol abuse. Case: A 54 year old male with chief complaint of left lower extremity swelling, redness and pain worsening for 1 week. Four months prior he noticed erythema and pain between his toes. He went to multiple ER and podiatrists, was treated with PO antibiotics – amoxillin, clindamycin, topical antifungals – and given the diagnosis of “Athlete’s foot.” As his foot became worse he presented to our ER complaining of worsening symptoms involving his entire left foot and ankle. There was no history of trauma, bug bite or immobilization. On exam he was afebrile with normal vitals. He appeared comfortable but in pain. Heart, lung and abdominal exam were unremarkable. Extremities revealed the left foot with diffuse erythema from ankle to toes; Desquamation of skin with yellow discharge. The toes were swollen and sausage-like. The whole foot was exquisitely tender with a beefy red appearance. Toe nails had a yellow hue, thickened, and Nikolsky’s sign positive. Labs were unremarkable and blood cultures were sent. He was diagnosed with Scalded Skin Syndrome and started on IV Vancomycin. He was evaluated by ID who continued Vancomycin and started prednisone. Day 7 he was discharged home with 3 days of clindamycin and a prednisone taper. Blood cultures were negative. Conclusion: Staphylococcal scalded skin syndrome is caused by epidermolytic toxins produced by various strains of Staphylococci. The toxin is spread systemically resulting in necrosis of keratinocyte attachments in the stratum granulosum - upper layer of epidermis - and sloughing of the skin. Generally the infection is diffuse involving the whole body sparing mucous membranes. It is thought that because of the ability to metabolize the staphylococcal toxin and excrete it through the kidneys, it is rare in adults; the incidence being <0.1% with a mortality rate reaching near 60%. It also has been suggested that immunosuppression increases one’s risk. References: 1. K.M. Acland et al. Staphylococcal Scalded Skin Syndrome in an adult associated with MRSA. British Journal of Dermatoligy. (1999) Vol 140 pg 518-520 2. S. Ladhani. Recent Developments in Staphylococcal scalded skin syndrome. Society of Clinical Microbiology and Infectious Diseases. (2001) Vol 7 pg 301-307 3. MajaMockenhaupt et al. Epidemiology of Staphylococcal Scalded Skin Syndrome in Germany. Society of Investigative Dermatology. (2005) Vol 124 Pg 700-703.
“MADNESS” A CASE OF MYXEDEMA

David Reyes

Introduction: Hypothyroidism is a medical condition encountered in a variety of clinical settings. 4.1/1000 women and 0.6/1000 men suffer from hypothyroidism. The somatic and neuropsychiatric presentations of thyroid hormone deficiency are often overlooked. Missing the signs and symptoms of hypothyroidism in our clinical practices may lead to severe consequences. Case: A 56 year-old female was brought to the hospital for ‘odd behavior” reported by her teenage children. According to them this problem had worsened over the period of one month. They noted a decline in her capacity, as she had stopped adequately performing her normal activities. Examples given were an inability to successfully pay her bills, cook, or dress herself. She could not understand why leaving her house without wearing pants was not correct. She was able to identify her children but she could not recall their names, and when asked for any date she would answer with her birthday. She was not oriented to place or time. Vital signs revealed a heart rate of Forty-nine, with her blood pressure and temperature being within normal limits. During the examination she was alert but lacked insight. On physical examination she had an edematous face and eyelids. Her lower extremities showed non-pitting edema, her reflexes were normal. Laboratory analysis revealed a raised TSH and trace levels of Triiodothyronine and Thyroxine. CT of the brain revealed frontal lobe and central atrophy, with agenesis of the corpus callosum, while neuropsychological assessment identified deficit in memory processing and executive functions. A tonic-clonic seizure occurred during her hospitalization. Despite appropriate treatment for primary hypothyroidism with Intravenous levothyroxine and corticosteroids. The patient remained delusional and kept responding with the date of her birthday for any date asked. Although she showed some improvement in memory capacity, she did not return to her usual state of mental function and her self-care remained poor; she was subsequently transferred to a rehabilitation center. Discussion: Myxedema madness was an alliteration coined by Asher in 1949. There is little doubt that thyroid hormone plays a major role in the regulation of, cognition, behavior and mood. Persons with thyroid dysfunction frequently experience a wide variety of neuropsychiatric symptoms. Profound hypothyroidism has been associated with irreversible structural damage; particularly to the frontal lobe of the brain, and has been reported several times in the literature. Conclusion: This case is presented to highlight the importance of identifying neuropsychiatric and somatic changes of profound hypothyroidism. References: 1. Zzopardi L, Murfin C, Sharda A, De Silva N. BMJ Case Rep. 2010 Sep 17;2010. doi:pii: bcr0320102841. 10.1136/bcr.03.2010.2841. 2. Heinrich TW, Grahm G.Prim Care Companion J Clin Psychiatry. 2003 Dec;5(6):260-266.
**Respiratory Distress in Myotonic Dystrophy**

Trisha Saha

Introduction: Myotonic dystrophy is a widely varied multisystemic disorder characterized by progressive muscle wasting, early cataract formation, cardiac abnormalities, and insulin resistance among others. Patients with myotonic dystrophy may present to any number of specialists requiring a high index of suspicion for a clinical diagnosis. Case Presentation: A 50 y/o female with history of pneumonia, myocarditis, and cataracts presented with acute onset of shortness of breath for five days. Initially the shortness of breath was only on exertion, which quickly progressed to shortness of breath at rest. Of note there was no history of orthopnea, postural nocturnal dyspnea, prolonged immobility, heart failure, or recent infection. Vitals on admission included O2 saturation of 96% on room air and blood pressure of . Pulmonary embolism was suspected as history revealed a five hour trip to Rochester. A CT angiography was done which revealed no evidence of pulmonary embolism. A transthoracic ECHO showed an ejection fraction of 50% and normal left ventricular function. She was sent to the floors on antibiotics. By the next day the her saturation had decreased to 92% on room air. Upon further questioning she mentioned she was diagnosed with Myotonic dystrophy at the age of 33. She was then sent to the ICU to observe the patient for respiratory failure. The patient’s symptoms were relieved significantly with administration of BiPAP during the night. Her oxygen saturation improved to 97% off BiPAP and her symptoms improved significantly. Discussion: Myotonic dystrophy is an autosomal dominant systemic disorder characterized by skeletal muscle weakness, cardiac conduction abnormalities divided into two broad categories, DM1 and DM2. DM 1 results from an unsteady repeat in CTG on chromosome 19, while DM2 is caused by an expansion of CCTG on chromosome 3¹. Conclusion: Many patients with Muscular Dystrophy survive well into middle age with new age medical care. Patients may present with symptoms of shortness of breath with a clinical picture that may suggest heart failure or an embolic event. Once they have been ruled out it is essential to consider respiratory distress secondary to muscle weakness as a cause of shortness of breath. Overnight mechanical ventilation such as BiPAP can provide significant symptomatic relief. As of yet there is no disease modifying agent for myotonic dystrophy therefore treatment is symptomatic. References: 1. Day JW, Ricker K, Jacobsen JF, et al. Myotonic dystrophy type 2: molecular, diagnostic and clinical spectrum. Neurology 2003; 60:657.
Drexel - St. Peter's

**Rickettsial pox in the Greater New York City Region: A Case Report.**

JANEESH SEKKATH VEEDU

Introduction - Rickettsia akari is still endemic to the region where it was first described in 1946 in the borough of Queens, New York City. Rickettsial pox should be included in the differential of patients with eschars, papulovesicular rashes and aseptic meningitis. Particular care should be exercised to develop a thorough history that includes a description of living conditions and highlights signs of recent infestation with the house mouse (Mus musculus). Case - 19-year-old African American female presented with 2 days of fever and gradual onset constant throbbing bifrontal radiating to the occiput headache, increasing in intensity with exposure to light not relieved by Tylenol or rest. Associated symptoms included: • Hyperosmia with a new onset olfactory sensitivity to soaps and perfumes. • One episode of vomiting, loss of appetite and decreased fluid intake. • Fever with diaphoresis. • Intense lethargy She lives on the second floor in a group home in Somerset, NJ with her 11-month-old daughter and 8 other women and their infants and the child attends daycare in the basement of the facility. She reports seeing many insects in the house and was bit on the chest by a bug “a few days before” admission. However, she has never seen rodents. Labs showed thrombocytopenia of 139. Lumbar Puncture: Opening pressure 60mm Hg WBC 32 with 99% lymphocytes, 1% PMN RBC 15000 Protein 131 Glucose 53 Supportive treatment was given. On day 3 – Symmetric, papulovesicular rash emerged on trunk, flank, lateral neck and wrists which was mildly pruritic on flank and neck 4 hours after onset. Headache resolved completely. Clinical diagnosis of Rickettsial pox was made and was treated with doxycycline and discharged home subsequently. Discussion Rickettsial pox should be on the differential of any patient in the greater New York City region that presents with: • History of cramped living quarters actively or recently infested with the common house mouse. • A systemic illness that includes high fever, severe headache, and a generalized papulo-vesicular rash approximately one week after eschar eruption. • Thrombocytopenia throughout the course of the illness. Confirmatory serologic analysis and/or IHC from eschar biopsy, offer the greatest opportunity to accurately diagnose rickettsial pox. Clinicians and the public should be increasingly aware of these poorly recognized but widespread infections. This case illustrates that clinical diagnosis without serology may be sufficient to warrant treatment. Ref- • Paddock, CD, et al, Isolation of Ricketsia akari from Eschars of Patients with Rickettsial pox. Am. J. Trop. Med. Hyg., 75(4), 2006, pp. 732-738. • Roos KL, Tyler KL. Chapter 381. Meningitis, Encephalitis, Brain Abscess, and Empyema. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson JL, Loscalzo J, eds. Harrison"s Principles of Internal Medicine. 18th Ed. New York: McGraw-Hill; 2012.
INTRODUCTION Lemierre syndrome is a rare but serious illness that includes throat infection and thrombosis of the internal jugular vein (IJV) with subsequent distant septic emboli. The most frequently isolated pathogen is Fusobacterium necrophorum. The infection usually begins with a sore throat followed by fever, septicemia, thrombosis and metastatic abscesses.

CASE An 18 year old African American male presented with one week history of sore throat, high grade fever associated with chills, neck pain, nausea and vomiting. He presented multiple times over the course of one week to his Primary care physician and to the ER. Rapid strep test was negative and was treated symptomatically with NSAIDS, Tylenol and zofran. He was called to come to the hospital for a positive blood culture. His vital signs included Temp 102.9F, pulse rate – 109 bpm and BP 86/32 mmHg. On physical exam patient appeared sick- lying in fetal position covered with many blankets and had rigors. Oral cavity- revealed bilateral tonsillar enlargement with erythema in the tonsillar pillars and posterior pharyngeal wall. Tenderness on the left side of the neck was present. Liver and spleen were palpable. Laboratory studies showed WBC 12.9 with 86% Polymorphs Hemoglobin 11.8, platelets 56 total bilurubin 2.2, AST 333, ALT 206 Alkaline phosphatase 126, Albumin 2.9, LDH 680. Blood culture was positive for gram negative bacilli, anaerobe- Fusobacterium Necrophorum. CT neck showed thrombosis of the left jugular vein with surrounding edema and infiltration consistent with thrombophlebitis. Small foci in the lung were suggestive of septic emboli. CT Abdomen and pelvis showed hepatosplenomegaly. Patient clinically improved with IV antibiotics and was discharged home on flagyl for 4 weeks.

DISCUSSION Lemierre’s syndrome is a rare illness affecting young adults 16 and 25 years. Males are more frequently affected. It develops in two clinical phases. Initial phase corresponds to oropharyngeal infection of the mucosa with or without peritonsillar abscess, fever and cervical lymphadenopathy. Second phase corresponds to infection spread to the pharyngeal space with thrombophlebitis of the IJV and sepsis.

CONCLUSION This case is presented to highlight the importance of a rare but still poorly understood serious condition. Delay in the diagnosis and initiation of treatment is responsible for increased morbidity and mortality. Diagnostic studies include blood culture and cervical and chest CT scan. Treatment is primarily medical with antibiotics for 2-4 weeks duration.

Drexel - St. Peter's

Post neurosurgical meningitis

Riddhi Shah

INTRODUCTION Gram negative bacterial meningitis is an important cause of health care associated Central Nervous System infections, most commonly occurring after a neurosurgical procedure. The presentation can be subtle and may easily be missed unless a high index of suspicion is executed by the treating physician.1

CASE 42 year old Indian lady was evaluated for gradually worsening headache and chills for one week and new onset swelling at the nape of the neck. Her symptoms started 1 year ago when she started experiencing recurrent episodes of left sided facial weakness, loss of voice, choking sensation and twitching of left eyelid; each lasting for few seconds and resolving spontaneously. A few weeks later she developed weakness of the right arm and leg; neuroimaging was done which showed Arnold-Chiari type 1 malformation. She underwent a decompressive craniotomy. One week after the surgery, she started experiencing occipital headache and chills. She was afebrile and other vital signs were stable. A 4 X 4 cm nontender, fluctuant swelling was noted at the nape of the neck. The neurological examination was remarkable for signs of meningeal irritation (positive Kernig, Brudzinky and Jolt sign). Laboratory studies showed leukocytosis. Noncontrast MRI of brain and cervical spine revealed presence of cervico-occipital meningocele. Lumbar puncture revealed polymorphonuclear leukocytosis, hypoglycorrhea and high protein, consistent with bacterial meningitis. CSF culture remained sterile; however blood and urine cultures grew β-lactamase producing E.Coli. Patient was treated with cefazolin and meropenem for 10 days with clinical improvement and resolution of the symptoms.

DISCUSSION Among the gram negative bacilli that cause meningitis, most frequently implicated are E. Coli, Klebsiella Pneumoniae, P. Aeruginosa and Acinetobacter. The former two account for more then 50% of cases of gram negative meningitis in adults after neurosurgery.2 Most cases of post neurosurgical meningitis occur 10 or more days after surgery. Clinical presentation can be subtle with an insidious onset.3 Gram negative meningitis is commonly accompanied by bacteremia. In fact, bacteremia is an important prognostic feature of gram-negative bacterial meningitis.

CONCLUSION The present case reviews the management of Gram negative bacterial meningitis as a complication of a neurosurgical procedure.

Acute Symptomatic Hyponatraemia Following Colonic Bowel Preparation

Emily Shay

Introduction: Hyponatraemia is a common electrolyte abnormality presenting with neurological symptoms that can range from mild and relatively nonspecific to devastating and deadly. Case: A 65 year old Filipino female with a past medical history of diabetes, hypertension, fatty liver and erosive gastritis presented with complaints of nausea, facial and upper body muscle twitching involving the eyelids, perioral region and spasms involving the feet and legs and mild confusion which began the morning of admission. Two days prior, she had undergone an upper and lower endoscopy as part of a work up for a possible pancreatic malignancy. Her review of systems was positive for unintentional weight loss of 20 lbs over two months. Her home medications included glimepride 2mg daily, omeprazole 20mg daily, and losartan/hydrochlorothiazide 100/12.5mg daily. On physical examination her blood pressure was 140/90 mmHg, heart rate was 123 beats per minute. She was awake, alert, fully oriented. Neurological exam was positive only for perioral twitching. Electrocardiogram was normal. Labs revealed a sodium of 113; potassium 4.0; chloride 76; bicarbonate 24; BUN 11; Cr 1.10; blood glucose 154; CPK 268. She was admitted to the ICU for treatment of symptomatic hyponatraemia with 3% hypertonic saline intravenously (IV) at 30 ml/hr, Furosemide 20 mg IV, and free water restriction of 800 ml/day. Her serum sodium within the first 10 hours reached 119 and her muscle twitching had resolved. By hospital day 3 her sodium had reached 132. Discussion: Acute hyponatraemia following colonic bowel preparation has a reported incidence of 7.6%1 and 8.8%2 based on two studies. It is due to the combination of excess free water ingestion and the normal physiologic action of Vasopressin (ADH). Medications such as diuretics and underlying colonic disorders potentiate the action of ADH, while polyethylene glycol-containing products can cause further increase its release. Abdominal manipulation, pain, nausea and stress – all of which are or can be part of the colonoscopy process – are also known to increase ADH release1. Conclusion: Patients – especially the elderly and those with medically managed comorbid conditions – should be made aware of the dangers of ingesting large amounts of electrolyte free water. Physicians should be aware of the potential of this adverse effect and recognize it in the context of a patient preparing for or having recently undergone colonoscopy.

Post traumatic syringomyelia: Dual challenge

Tejas Sheth

Introduction  Syringomyelia is characterized by presence of fluid-containing cavities in the parenchyma of the spinal cord. Post-traumatic syringomyelia (PTS) is the most common acquired form of this condition. The incidence ranges from 0.3 to 3.2%. There is currently no optimal intervention capable of producing satisfactory long-term clinical results. Case report  A 29 year old Mexican gentleman was evaluated for stiffness in bilateral lower limbs for past 7 months. Patient had history of fall from bike 3 years ago, followed by lower back pain and left sided knee pain immediately after the fall. A year later he started noticing weakness of left hand grip along with tingling and numbness and inability to perceive hot/cold sensation. Vital signs were stable. On examination all cranial nerves were intact. Patient had wasting of intrinsic muscles of left hand with flattening of hypothenar eminence resulting in ulnar “claw hand”. Bilateral lower limbs had clasp knife spasticity with 4/5 strength. Sensory examination revealed “dissociative anesthesia” in the upper extremities with intact light touch and grossly impaired temperature senses. Reflexes were exaggerated and there was sustained clonus at knee and ankle, bilaterally. MRI revealed a central syrinx cavity from C4 to C8, displacing the cord dorsally. The diagnosis cervical central cord syndrome due to PTS was made. Patient was managed surgically by decompressing lateral laminectomy followed by syringo-subarachnoid-pleural shunt. Three week post-op, patient reported marked improvement in balance, spasticity and coordination with no further worsening of the weakness. Discussion  PTS is the occurrence of longitudinal cavities within the spinal cord following trauma, typically in the chronic phase. The proposed mechanisms include narrowing or obstruction of the subarachnoid space, central canal occlusion, myelomalacia and alterations in intramedullary water permeability. The literature supports a strong recommendation for surgical intervention in the setting of motor neurologic deterioration and several surgical options exist including shunting the syrinx to the subarachnoid space or to either the pleural or peritoneal cavities via a valveless tube; spinal cord untethering with or without expansile duraplasty; and cordectomy. However, the proper management strategy for patients with spinal cord injury with respect to PTS has not been established. Conclusion  The present case describes the clinical and radiological improvement in PTS with internal decompression and syringo-subarachnoid-pleural shunting. References 1. Ko HY, Kim W, Kim SY, et al. Factors associated with early onset post-traumatic syringomyelia. Spinal cord. Sep 2012;50(9):695-698. 2. Shields CB, Zhang YP, Shields LB. Post-traumatic syringomyelia: CSF hydrodynamic changes following spinal cord injury are the driving force in the development of PTSM. Handbook of clinical neurology. 2012;109:355-367. 3. Bonfield CM, Levi AD, Arnold PM, Okonkwo DO. Surgical management of post-traumatic syringomyelia. Spine. Oct 1 2010;35(21 Suppl):S245-258.
Hypervitaminosis D induced hypercalcemia in Osteoporosis

Gagan Singh

Introduction: Vitamin D intoxication has been found in all age groups and is rare but treatable cause of hypercalcemia. It can be iatrogenic due to self medication or accidental with over fortification of milk. Patients especially immigrant and foreign population often get treated with higher than recommended doses of vitamin D for various diseases and on occasions doctor-patient communication error can lead to doses intake that far exceed the requirements of the patients. Case: 67 year old lady visiting from India with a history of Diabetes Mellitus, Coronary Artery disease, Chronic kidney disease, Hypothyroidism, Diastolic Heart Failure, Osteoporosis taking Vitamin D 60,000 IU (1.5 mg) daily since 6 months presented to Emergency Department with 2 weeks history of nausea, vomiting, abdominal pain, constipation, polyuria and polydipsia. On examination her blood pressure was 158/73 mmHg, pulse 79 beats per minute, temperature 98.6 F, and oxygen saturation 99 % on room air. Mild epigastric tenderness was present on abdominal examination, heart and lung examination were unremarkable. Laboratory tests showed leukocytosis (11,200/cumm with 79.2% neutrophils), Calcium(Total 13.1, Ionized 5.6), Potassium 3.1, magnesium 1.2, PTH 7.1 pg/ml, 25-Hydroxy-Vitamin D total 406 ng/ml, TSH 1.58 IU/ml. EKG and Chest X-Ray were unremarkable. Intravenous fluids and lasix were started, insulin for DM, home medications for hypertension and hypothyroidism were continued and vitamin D was discontinued. Potassium and magnesium were replaced. Patient symptoms and hypercalcemia improved with iv fluids and lasix and patient was discharged after 2 days with calcium level 10.7. Discussion: Vitamin D supplementation is recommended for prevention and treatment of osteoporosis with maximum doses of 250 micrograms (10,000 U) per day (1,3). Doses more than 50,000 U per day for several months or more than 300,000 U in 24 hr period raise levels of 25-OH Vit D more than 150 ng/ml and are associated with hypercalcemia. (2) Conclusions: Education should be provided to health care providers for prescribing recommended doses of vitamin D and patients about risks and symptoms associated with hypervitaminosis D. Health care provider and patient communication errors should be avoided by electronic prescribing of drugs along with doses, simplify dosing schedules and follow up with home medication list References: 1. Takako Araki, Michael F. Holick, Bianca D Vitamin D Intoxication due to labeling error J Clin Endocrinol Metab, December 2011, 96(12):0000–0000 2. Parvaiz A. Koul,* Sheikh Hilal Ahmad, Feroze Ahmad, S.U. Shah, and Umar H. Khan Vitamin D Toxicity in Adults: A Case Series from an Area with Endemic Hypovitaminosis D 3. Kerstens PJ, van Ditzhuijsen TJ, van Tongeren JH Mega-dosages vitamin D: progressive medicine] Ned Tijdschr Geneeskd. 1990 Oct 6;134(40):1959-61.
Löfgren’s Syndrome

Yojna Singh

Introduction Sarcoidosis is a multisystemic granulomatous disease affecting all races and ages, with incidence of 6.3/5.9 cases per 100,000 in women/men respectively. Löfgren’s syndrome: a variant, presents more acutely with erythema nodosum, periarticular inflammation, bilateral hilar lymphadenopathy and fever. As most patients with sarcoidosis do not die of the disease, medical challenge is to help them live well with their symptoms. Case A 56-year-old female from Honduras presented with one week history of dry cough, fever with chills and chest pain progressing to development of generalized arthralgia in multiple joints, involving wrist, elbows, shoulders, knees. She presented to hospital with pronounced skin rash, mostly in trunk, arms and extremities. On exam her temperature was 99.1 F, pulse 110 bpm and BP 118/64 mmHg. Lung exam reveled bilateral wheezing and faint rales at the bases. She had erythroderma over face, upper trunk, bilateral upper and lower extremities with areas of intensely macular rashes scattered on right lower shin. Right wrist was warm and tender. Lab studies included: WBC 29500, 4% bands, 85% polymorphs, MCV 84.8, Hemoglobin 11.1, platelets 507000, ESR 96, CRP 85, LA 1.8, Uric acid 3.5. Chest X Ray showed bibasilar linear densities with mediastinal fullness. CTA revealed Mediastinal and hilar lymphadenopathy with right paratracheal adenopathy with differential of tuberculosis, sarcoidosis and lymphoma. She was initially treated with antibiotics for possible pneumonia, without clinical improvement. Blood, Sputum, and AFB cultures yielded no growth. ACE test was deferred due to low sensitivity and specificity. Patient was started on prednisone 30mg daily which led to rapid resolution of her symptoms. Discussion Löfgren’s syndrome is a self-limiting form of sarcoidosis, with a good prognosis. Elevated levels of ACE are reported in 60% patients with acute disease and only 20% with chronic disease. It may also be seen with other disseminated granulomatous diseases, leprosy, Gaucher’s disease and hyperthyroidism. Histologic confirmation is not necessary in typical cases. More than 90% of patients experience disease resolution within 2 years. Steroids help in acute settings as in our patient and may shorten the recovery period. Conclusion Because of its nonspecific presentation, the diagnosis of sarcoidosis can be challenging and is often delayed. Our patient had compatible clinico-radiologic features highly suggestive of Löfgren’s syndrome and was managed appropriately with steroids without extensive pathological testing. References 1. Grunewald J, Eklund A: Löfgren’s syndrome: Human leukocyte antigen strongly influences the disease course. Am J Respir Crit Care Med 179:307, 2009 2. Ziegenhagen MW et al: Exaggerated TNF-alpha release of alveolar macrophages in corticosteroid resistant sarcoidosis. Sarcoidosis Vasc Diffuse Lung Dis 19:185, 2002
**Pancreatic adenocarcinoma-induced cardiopulmonary complications.**

Rushab Vakharia

Introduction: Some complications of pancreatic adenocarcinoma can be foreseen, but regardless of the medical care, they are not always preventable. This case presents one of the latter situations. Case Description: A 78 year old man, status post ERCP for obstructive jaundice and diagnosis of pancreatic cancer, was admitted for a Whipple procedure. Due to invasion of the Superior Mesenteric Vein and Portal Vein, the procedure was aborted as the mass was deemed unresectable. His postop course was complicated by the new onset of atrial fibrillation which was treated with a Cardizem drip. Chest x-ray done at the time revealed a small pleural effusion on the right as well as some pulmonary vascular congestion. Clinical suspicion for PE prompted a CTA of the chest which revealed a small right sided embolism. He was started on anticoagulation with heparin. The patient consequently developed bradycardia with a significant pause lasting 4.2 seconds requiring the discontinuation of the Cardizem and Metoprolol. He did not require any invasive management as he converted to sinus rhythm on his own. Over the next week, he continued to show sinus node dysfunction with episodes of atrial fibrillation and asymptomatic sinus bradycardia. An additional complication arose in the form of acute hypercapneic respiratory failure requiring diuresis, BIPAP, and ICU stay. He improved but was later found in respiratory distress hypotensive, unresponsive. He was intubated for respiratory failure due to aspiration. This improved with the use of antibiotics, pressors, and critical care management. In light of his current condition and that he was asymptomatic through this tachy/brady episodes, it was determined that placement of a pacemaker was not indicated at this moment for his sick sinus syndrome and this would be readdressed again in the future. His condition improved in the following days and he was able to be extubated and transitioned to warfarin for his PE. Conclusion: This case shows the importance of management of cardiopulmonary complications following surgery in a patient with a malignancy. Although steps were taken to treat each complication, the subsequent one was unable to be prevented, even with appropriate medical care. This case also shows a side of sick sinus syndrome that doesn’t require pacemaker placement in the acute setting. References: Horsted F, West J, Grainge MJ. Risk of venous thromboembolism in patients with cancer: a systematic review and meta-analysis. PLoS Med. 2012;9(7):e1001275  Stein PD, Beemath A, Meyers FA, Skaf E, Sanchez J, Olson RE. Incidence of venous thromboembolism in patients hospitalized with cancer. Am J Med. 2006;119(1):60–68
Gastric Bypass (Bariatric) Surgery Triggered Hyperammonemic Encephalopathy

Shuang Wang

Introduction: The number of bariatric procedures is rapidly growing as the prevalence of obesity is increasing. Complications following surgical treatment of severe obesity vary based upon the procedure performed and can be as high as 40 percent (1). Here we present a rare case of hyperammonemic encephalopathy after gastric bypass surgery.  Case: A 65-year-old woman with a past medical history of asthma, obstructive sleep apnea, and morbid obesity status post Roux-en-Y gastric bypass surgery 19 years ago with 170 pounds weight loss presented with acute altered mental status and a fall. She had multiple similar episodes in the last year. The patient was evaluated by a neurologist, seizures and CVAs were ruled out. She was found to have a pancytopenia, mild abnormal liver function test and non-cirrhotic liver with fatty infiltration from CT scan of abdomen. Her serum ammonia levels were persistently elevated during these episodes being 110microg/dL at this admission. Metabolic encephalopathy secondary to hyperammonemia was diagnosed after pharmacologic, microbial, and autoimmune causes were excluded. Patient responded well to traditional ammonia-reducing therapies which included lactulose and rifampin. An urea cycle disorder was suspected but further work up revealed a normal Carnitine level.  Discussion: Encephalopathy has been described as a rare complication of gastric bypass surgery, but it has generally been attributed to deficiencies of thiamine and other nutrients (2). There have been a few other case reports of hyperammonemia in patients who have undergone gastric bypass (3). Some patients died shortly after surgery, and some had a steady decline that led to coma or death due to elevated ammonia levels. A number of patient were found to have a previously undiagnosed genetic mutation (ornithine transcarbamylase deficiency) resulting in a urea cycle disorder. Other cases were considered to have secondary or acquired urea cycle disorder due to malnourished states. Further studies are needed to establish the mechanisms resulting in hyperammonemic encephalopathy after gastric bypass surgery. Ornithine transcarbamylase geneDNA analysis should be performed in these patients.  Conclusion: It is important that clinicians are aware of hyperammonemic encephalopathy which can occur after gastric bypass surgery. Early recognition will lead to appropriate testing, diagnosis and treatment. This may prevent a fatal outcome in these patients.  References: 1. Longitudinal Assessment of Bariatric Surgery (LABS) Consortium, Flum DR, Belle SH, et al. Perioperative safety in the longitudinal assessment of bariatric surgery. N Engl J Med 2009; 361:445. 2. Juhasz-Pocsine K, Rudnicki SA, Archer RL, Harik SI. Neurologic complications of gastric bypass surgery for morbid obesity. Neurology.2007;68:1843-1850. 3. Limketkai BN, Zucker SD. Hyperammonemic encephalopathy caused by carnitine deficiency. J Gen Intern Med. 2008;23(2):210-213.
HUMC Mountainside

A Case of Extremely High Triglycerides Resistant to Therapy

Soemiwati Holland, Lee Loewinger, M.D

A Case of Extremely High Triglycerides Resistant to Therapy Soemiwati W Holland, M.D, Lee Loewinger, M.D.  Hypertriglyceridemia (HTG) is the third most common cause of acute pancreatitis (AP), after alcohol and gallstones. Hypertriglyceridemia is considered to increase the chance of acute pancreatitis in a level-dependent manner through concentrations of greater than 1000 mg/dL (11.2 mmol/L). We report a case of pancreatitis associated with an extremely high triglyceride level, despite pharmacologic therapy.  CASE REPORT A 42-year-old mildly obese male presented to our ER with progressive abdominal pain.  His history included a prior bout of pancreatitis 8 years ago, attributed to severe hypertriglyceridemia, for which the patient had been prescribed Fenofibrate 130 mg daily and Simvastatin 40 mg daily.  He had been diagnosed with diabetes six months prior to admission, and was taking Metformin 500 mg BID.  The patient described minimal alcohol use.  Exam revealed a soft, diffusely tender abdomen.  CT scan showed liver is diffusely fatty. There is acute pancreatitis with peripancreatic edema from the tail all the way to the head. There is mesenteric engorgement. Ultrasound revealed a large, fatty infiltrated liver and a bulky heterogeneous pancreas, indicating pancreatitis.  Lipid panel drawn at time of presentation revealed a triglyceride level of 4404, total cholesterol of 758, HDL of 18, and LDL was incalculable, Lipase 5335.  The patient was admitted to the hospital and started on an insulin drip.  Standard supportive care was provided.  The patient improved clinically, and after five days his triglyceride level was 600.  He was discharged home on a regimen of Fenofibrate 134 mg daily, Gemfibrozil 600mg BID, and Simvastatin 40 mg daily.  DISCUSSION Hypertriglyceridemia is the likely etiology for almost 10% of cases of acute pancreatitis, although it is present in the majority of cases of pancreatitis from any cause.  The most common clinical scenario for a patient to present with triglyceride-induced pancreatitis is a patient with a triglyceride level over 1000 mg/dL and uncontrolled diabetes.  Our patient had a history of severe HTG that was controlled for many years with a Simvastatin and Fenofibrate, but once he became diabetic that control was inadequate. Patients with extremely high triglyceride levels must be monitored closely throughout their lives. Introduction of another major risk factor to a patient with HTG is the most typical scenario in which these patients experience HTG-related acute pancreatitis.  The most common risk factor to be so associated is uncontrolled diabetes. When the patient with a history of severe hypertriglyceridemia develops diabetes, their current medical regimen, even if effective up until that point, may become inadequate.  The primary physician must keep this in mind and in addition to aggressively controlling glucose levels, must follow these patients’ lipid panels and increase lipid therapy as needed.
Atrial fibrillation is a common medical problem and is frequently complicated by thromboembolism, most notably to the cerebral circulation. Emboli to the kidney, liver, and spleen also occur, although these are rarer. We report a case of a cardioembolic event caused by atrial fibrillation, wherein the patient presented with acute abdominal pain and was found to have concurrent embolus-induced infarction of these three organs.  

CASE REPORT  
An 87 year-old female presented to the ER with complaints of abdominal pain, nausea and vomiting. She had recently been diagnosed with atrial fibrillation and started on Lopressor 25 mg bid and Rivaroxaban 15 mg daily. Rivaroxaban had been held because of persistent diarrhea and she was put on aspirin 325 mg daily. Computed Tomography Angiogram (CTA) of the abdomen showed infarction in the lower half of the left kidney, splenic infarct in the lower pole measuring 2.5-3cm, and a linear peripheral density in the liver, representing small peripheral infarcts. A transthoracic echocardiogram showed no visible clot in the left atrium. The patient was started on anticoagulation and improved clinically.  

DISCUSSION  
Renal infarction is rare and likely underreported. The incidence is 1.4% on autopsy, but the condition is underdiagnosed in living patients since the presenting symptoms of abdominal and flank pain resembles many more common clinical scenarios. Atrial fibrillation seems to be involved in approximately one quarter of renal infarctions. In an autopsy series of 96 consecutive cases of splenic infarction only 10 % had been suspected clinically. Infarction of the spleen was reported in 60 of those cases, involving the kidneys in most cases (62%). Another study recognized 205 autopsy cases of renal infarcts, of which only 1 % had been diagnosed antemortem, with infarct of the other organs present in 69 % of the cases. The spleen was involved in 91 cases. The liver has a dual blood supply, making hepatic infarction less common than other forms of ischemic injury. Even in our case the CTA demonstrates a linear peripheral density, which represents small peripheral infarcts as opposed to a major infarct. The use of CTA in the Emergency Department diagnosed this patient early with her triple infarct. CTA was adequate to diagnose triple infarct. Stroke is the most known and feared complication of atrial fibrillation, but this case demonstrates that patients with this disease are vulnerable to all manner of thromboembolic events, and that aspirin is often inadequate protection. Physicians must keep these more exotic embolic events in mind when treating any patient with atrial fibrillation, as the presentation can be highly variable.
Herpes Esophagitis as a Complication of Epidural Steroid Injection

Riffat Jafrin, Vinit Gupta MD

Herpes Esophagitis as a Complication of Epidural Steroid Injection Riffat Jafrin MD, Vinit Gupta MD, Department of Medicine, HackensackUMC Mountainside, Montclair, NJ Introduction Epidural steroid injections are common therapeutic interventions in the management of musculoskeletal pain. However, the systemic risks of these procedures, including immunosuppression, are underappreciated. Case Description A 51 year old male with well-controlled HIV was admitted to HackensackUMC Mountainside Hospital with dehydration and acute kidney injury. Four days prior, he had received an epidural steroid injection into the cervical spine for chronic neck pain from disc disease and scoliosis. He had been fully compliant with a regimen of emtricitabine, tenofovir, and ritonavir-boosted atazanavir for many years with a recent CD4 count of 337 and undetectable viral load. Physical examination on admission revealed evidence of oropharyngeal candidiasis. Blood tests confirmed an undetectable viral load, but with a CD4 count of 54. Fluconazole therapy was initiated, but the patient developed progressively worsening odynophagia. He underwent upper endoscopy which showed severe esophageal inflammation. Biopsy specimens were consistent with herpetic esophagitis. The patient was subsequently treated with intravenous acyclovir, which resulted in resolution of his symptoms. Discussion While it is not unusual to see herpes esophagitis in HIV patients, it is typically associated with persistently depressed CD4 counts. We hypothesized our patient’s CD4 count dropped acutely as a result of the epidural steroid injection. Moreover, discussions with his primary physician confirmed the patient had been well-controlled on antiretroviral therapy for several years. This case illustrates the potential for systemic immunosuppression as a consequence of an epidural steroid injection. Given the high frequency with which epidural steroid injections are administered, physicians should be aware of the potential for systemic side-effects.
INTRODUCTION: Neurocysticercosis (NCC) is the most common parasitic infection of the central nervous system worldwide. It is an uncommon, yet growing, problem in the United States, especially among certain immigrant populations. Racemose Cisternal Cysticercosis is a rare form of NCC which usually presents with signs and symptoms of meningitis and raised intracranial pressure. We report a case of RCC manifesting as headache and diplopia in an immigrant from Guatemala. CASE REPORT: A 32 year old male with no significant past medical history presented with complaints of severe headache and diplopia for three weeks. Headache was associated with photophobia, blurred vision, nausea, and vomiting. The patient came from Guatemala eleven years prior and had not returned since. Physical examination was remarkable for bilateral abducens nerve palsy. MRI revealed subtle deformity of the suprasellar and perimesencephalic region with dilation of the basal cisterns, and mass effects on the cranial nerves with septations. An EEG was normal. Lumbar puncture revealed low glucose and high protein with 95 white blood cells that were predominantly lymphocytes. Serum cysticercosis antibodies are positive. The patient was started on albendazole, steroids, and levetiracetam. His headache and diplopia improved rapidly and repeat MRI one month later showed improved radiological findings. DISCUSSION: NCC is usually transmitted by eating undercooked pork containing cysticerci. While it is not quite as prevalent in the United States as in other countries, it is estimated that 1000 new cases occur annually. This is attributable to an increase in immigration from countries with endemic NCC and improvements in CNS imaging. CT and MRI findings are quite characteristic and diagnostic for this condition. RCC is a rare subtype of NCC and often presents with grape-like clusters of multiple cysts located in the basal cisterns, sylvian fissures, or ventricles. Seizures are the most common presentation of NCC. Unlike parenchymal NCC, RCC can raise intracranial pressure from an intense inflammatory reaction, fibrosis, and progressive thickening of the leptomeninges at the base of the brain, leading to hydrocephalus and cranial nerve palsies. Differential diagnosis includes lesions such as epidermoid or arachnoid cysts. Treatment of RCC is multidisciplinary including medications such as albendazole, steroids, and levetiracetam. Surgical cyst excision with shunt placement can be performed if the patient has signs of hydrocephalus. Neurocysticercosis is expected to increase in incidence in the United States. Physicians should have a high index of suspicion for this disease when treating a patient from an endemic country with refractory headaches, seizures, and cranial nerve palsies.
Primary Pancreatic Sarcoma: A Rare Cause of Pancreatic Neoplasm

Sonja Abbassi, John Mikhail, MD, Reza Akhtar, MD

Learning Objectives: 1) Identify a rare case of primary pancreatic sarcoma. 2) Review the etiology, pathology, and histology of pancreatic sarcoma. Case: A 66-year-old male presented with mid-epigastric pain for 2 weeks. PMH: hypertension, hyperlipidemia, iron deficiency anemia, and benign prostatic hyperplasia. He reported worsening constipation and pain with eating during this time. There was no fever, chills, nausea or vomiting, and no alcohol use. Initial labs: amylase 104 U/L, lipase 171 U/L, hemoglobin 11.1 GM/dL, and MCV 69.1 FL. CT of abdomen and pelvis revealed a 7.5 x 8.5-cm soft tissue mass at the level of the pancreatic head and body, worrisome for primary pancreatic neoplasm. In the liver, at least two enhancing lesions were seen. Bone scan showed tracer activity in the left sacroiliac joint, suggesting possible metastases. Additional lab values were checked, including CEA and CA 19-9, which were normal at 1.2 NG/ML and 12 U/mL, respectively. Subsequently, EGD with endoscopic ultrasound (EUS) and fine needle aspiration (FNA) revealed a large, heterogeneous, uncinate pancreatic mass. The lesion was mostly solid with some cystic areas, with suggestion of involvement of the SMA and SMV. FNA revealed neoplastic cells positive for vimentin stain, equivocal for pan-melanoma stain and negative for S100, cytokeratin AE1/AE3, CAM5.2, E-cadherin and TTF1 stains. This profile was consistent with high-grade malignant neoplasm favoring undifferentiated sarcoma. Immunohistochemistry stains showed that the tumor cells were negative for inhibin, EMA, CK-18, and synaptophysin. The decision was made at a tertiary referral cancer center to treat with initiation of gemcitabine and dicetaxel; a regimen for carcinoma with sarcomatoid features. The regimen has been well tolerated and the patient''s abdominal discomfort has completely resolved. Conclusion: Primary pancreatic sarcomas represent an exceedingly rare breed of pancreatic tumors which account for less than 0.1% of all pancreatic malignancies. According to literature, pancreatic sarcomas occur frequently in younger individuals, and the pancreatic caput is most commonly involved followed by the tail and the body. The origin of such tumors is often argued considering the rarity of the pancreas being the primary source. Clinically patients present with nonspecific complaints such as abdominal pain, nausea and vomiting. The presentation and diagnostic strategies are similar to those employed with more common pancreatic malignancies. Axial imaging complemented by EUS allowed appropriate staging and diagnosis of this lesion. EUS additionally aided in investigating the lesion''s involvement with the gastric wall layers; an important consideration in light of the histology. Treatment regimens may be varied and considering the rarity of this neoplasm, consultation with tertiary referral cancer centers should be considered.
LIVEDO RETICULARIS: RARE PRESENTATION OF AORTIC DISSECTION AND THROMBUS

Sonja Abbassi, Heather Boakye, MD, Mayer Ezer, MD

Learning Objectives: 1) Recognize the presentation of an aortic thrombus. 2) Discuss the differential diagnosis of distal atheroemboli. Case: A 49 year old male presented with left lower extremity (LLE) crampy pain and rash for three weeks. PMH: hypertension, hyperlipidiema, 30 pack year smoking, daily alcohol consumption, and cardiac catheterization five months prior to admission. He initially presented to another hospital only with leg pain; there the workup was negative for venous or arterial occlusions and he was discharged with narcotic pain medications. The intensity of the pain increased, localized to the toes, and a purplish discoloration developed his lower leg. He returned to the same hospital and was started on prednisone for possible vasculitis. The pain persisted, so he decided to come to our hospital. Vital signs were normal. Exam: blanching, mottled, and erythematous macular rash that extended from mid calf to the fifth digit on the LLE consistent with livedo reticularis. He had 1+ dorsalis pedis and posterior tibialis pulses and skin was cool and painful to light touch. CMP, CBC were normal. Repeat LLE arterial and venous dopplers were negative for arterial or venous occlusion. CT angiogram of the abdominal aorta and lower extremities demonstrated a 2.6 cm long subtotal intraluminal occlusion of the infrarenal abdominal aorta along with total occlusion of left anterior tibial artery. Further CT imaging of the ascending and descending thoracic aorta was negative for any other aortic pathology. Hypercoaguable workup was negative and transthoracic and transesophageal echocardiograms were negative for intracardiac pathology. He underwent open laparotomy with aortic exploration and embolectomy. Pathological analysis of the thrombus demonstrated a heterogeneous mosaic with the presence of thrombin consistent with a white thrombus. There were no post-op complications and he was discharged on aspirin and warfarin. Discussion: Aortic dissections are more commonly seen in the elderly population and associated with uncontrolled hypertension. Dissections involve a tear in the internal layer (intimia) of the aorta due to weakening of the surrounding layer (media) producing a false lumen. The false lumen contains degenerative tissue which is prone to rupture. 30-50% of aortic ruptures are fatal despite surgical intervention. Prior literature recognizes that underlying inflammation is involved in aortic dissections; and it this inflammation along with turbulent blood flow that may promote blood clot formation. The proposed etiology of the patient's aortic thrombus is thought to be secondary to the underlying dissection. The origin of the dissection is also unclear, however his recent cardiac catheterization could have caused endotheliam damage that triggered a cascading event. Without surgical repair of the dissection, the damaged area is exposed promoting clot formation. Anticoagulation is warranted to prevent a repeat event.
Meridian Health-Jersey Shore

Title: What Happened to My Platelets! Development of Thrombotic Thrombocytopenic Purpura Without Significant Schistocytes in the Setting of Pregnancy and Scleroderma

Heather Boakye, Michael Carson MD, Eric Costanzo DO

Objectives: 1.) Recognize the differential diagnosis for acute thrombocytopenia. 2.) Diagnose thrombotic thrombocytopenic purpura. Case Summary: A 37 year old G7P0242 at 15 weeks gestation with scleroderma presented with headaches and epistaxis for two weeks. BP 109/68 mmHg. Initial labs: platelet (PLT) count 9 K/uL (140-450), hemoglobin (Hgb) 8.6 GM/dL (12.0-160), LDH 765 IU/L (91-200), haptoglobin < 6 mg/DL (30-225), creatinine 0.61 mg/DL (0.6-1.0), AST 63 IU/L (15-37), ALT normal, INR 1.04, fibrinogen 385 mg/DL (232-519), Coomb’s negative, proteinuria absent. Initial peripheral blood smear (PBS) had 1+ schistocytes but was not considered severe enough by the hematologist to be consistent with microangiopathic hemolytic anemia (MAHA). The working diagnosis upon admission was idiopathic thrombocytopenic purpura (ITP) and dexamethasone 40mg PO was administered for two days without improvement in the PLT count. Intravenous immunoglobulin (IVIG) was added for two days, again without improvement. Fetal ultrasounds, daily fetal surveillance and renal function remained normal. Day 5: her speech became slurred prompting a change of the admission diagnosis to thrombotic thrombocytopenic purpura (TTP). Plasmapheresis was initiated during which she had a tonic-clonic seizure requiring intubation, and she had an incomplete spontaneous abortion. Brain imaging and fetal/placental pathology were normal. After four plasmapheresis treatments, her PLTs improved to 135 K/uL, Hgb stabilized at 9.4 GM/dL, and LDH decreased to 330 IU/L. She was extubated without any neurologic sequelae. PLTs (221 K/uL) and Hgb (12.1 GM/dL) normalized after seven more plasmapheresis treatments. ADAMSTS-13 level prior to initiating plasmaphoresis was 7 units (>67).

Discussion: TTP is characterized by the pentad of MAHA, thrombocytopenia, neurologic abnormalities, fever, and renal disease. The exact etiology is unknown; however it associated with ADAMSTS13 deficiency, resulting in large PLT multimers that promote intravascular thrombosis at the microvasculature level. While MAHA is a hallmark finding for TTP, the criteria do not define a severity that is diagnostic. Commercial assays are imperfect and ADAMSTS13 levels do not come back quickly enough to establish the diagnosis upon admission. In this case, Evan’s syndrome (autoimmune hemolytic anemia and thrombocytopenia possibly associated with scleroderma) was excluded by the negative Coomb’s test, and normal BP and renal function excluded Scleroderma renal crisis. Plasmapheresis is not without its risks, but neither is delaying treatment for TTP. A literature search identified a similar case of TTP without MAHA that responded to plasmapheresis. Our case demonstrates that the diagnosis of TTP can be challenging in the presence of other confounding variables (i.e. Scleroderma). Pregnancy is associated with 10-25% of cases and despite underwhelming MAHA, elevated LDH and low haptoglobin clearly demonstrated hemolysis. The inclusion of biochemical evidence to support MAHA could have avoided the delay in utilization of plasmapheresis, the treatment that ultimately reversed her disease.
Incidentaloma: Testicular Seminoma

Megha Chitkara, Hegazi, Mohamed MD; Mencel, Peter J. MD, FACP

Learning Objectives 1) Recognize the uncommon presentations of testicular seminoma 2) Understand the management of incidentalomas

Case  A 37 year old male with no significant past medical history presented to our hospital with complaints of bright red blood per rectum (BRBPR) for one day with associated lower abdominal cramping. He denied weight loss, fevers, diarrhea or a change in bowel habits. The patient had not traveled out of the United States and did not have any significant exposure. On admission, he was afebrile with normal vital signs. His physical exam and laboratory findings were within normal limits. He was admitted for a gastrointestinal bleeding. His hemoglobin and hematocrit was checked serially. Endoscopy was deferred until the patient began to show a drop in H/H or had another episode of BRBPR. A CT scan of the abdomen and pelvis was done which incidentally showed a left para-aortic pathologic retroperitoneal lymph node which was biopsied. Pathology of the lymph node returned as a germ cell tumor (pure seminoma). A testicular ultrasound showed echogenicity. The patient underwent a left inguinal radical orchiectomy; pathology was consistent with pure seminoma.

Discussion  Testicular malignancy is the most common type of malignancy in men between the ages of 15 and 35. 80% of cases present with disease limited to testicles and 15% present with extension to retroperitoneal lymph nodes. The remaining 5% present with spread beyond retroperitoneal lymph nodes. The most common presentation is unilateral, painless testicular swelling. This case was unique due to the fact that the patient presented with a lower GI bleed and the retroperitoneal lymphadenopathy was found incidentally on imaging. After the lymph node biopsy showed pure seminoma, a full testicular physical exam was performed which revealed no abnormalities. Our literature search did not show any reports describing an incidental finding of retroperitoneal lymphadenopathy leading to the diagnosis. Furthermore, compared to other case presentations, this case did not have duodenal lymph node metastases, but rather retroperitoneal lymph node metastases. GI blood loss as an initial presentation of pure seminomas is rare, and in this case the described GI blood loss was the incident leading to the unexpected finding of retroperitoneal lymphadenopathy in a patient with no complaints related to his testicles.
Learning Objectives 1) Diagnose a subtle presentation of fungal endocarditis. 2) Recognize that arterial embolic events should prompt a workup for fungal endocarditis.

Case Summary A 23 year old woman G1P0 at 30 weeks gestation presented with right groin pain for 2 days. PMH: hepatitis C and prior IV drug use. Exam: 2/6 systolic murmur and mild tenderness to palpation of her right inguinal region. Lab workup revealed a CRP of 41.8 mg/l (N <10 mg/l) and a normal CBC and chemistry. Ultrasound of the right leg revealed a profunda femoral arterial thrombosis attributed to the hypercoagulable state of pregnancy. The murmur noted on the physical exam prompted a transthoracic echocardiogram (TTE) which showed a large aortic valvular lesion. Blood cultures were drawn. Given preliminarily negative blood cultures and lack of fever or leukocytosis in a non-toxic appearing patient, this was considered to be a sterile vegetation rather than acute infective endocarditis. The decision was made to treat conservatively and no antibiotics were started. On day 3, her blood cultures grew Candida parapsilosis and the patient was started on liposomal amphotericin B. During the initial infusion she developed hypotension and fetal heart rate decelerations. Once stabilized, she was transferred to our hospital for further management. Discussion Infective endocarditis (IE) refers to an intracardiac infection that resides on one or more cardiac valve surfaces. Arterial embolization is more common in fungal endocarditis than in bacterial, given the larger size of the vegetations associated with fungal endocarditis. In this case, as pregnancy is considered to be a hypercoagulable state and the patient had no signs or symptoms of infection, the etiology of her arterial thrombus was unknown. With symptomatic arterial clots, embolization from IE should be considered. In a literature review of 270 patients with fungal endocarditis, peripheral arterial embolization occurred in 45%, with 16% of them to the femoral artery. At our institution, the patient was premedicated with IV hydrocortisone and benadryl prior to restarting infusions with liposomal amphotericin B with close maternal and fetal monitoring. She was referred to cardiothoracic surgery for valve replacement. Nonobstetric surgery during pregnancy has been associated with preterm labor and delivery, but poses no teratogenic risk after the first trimester. However, prior to initiating maternal cardiac bypass, the decision was made to deliver the fetus preterm via cesarean section prior to valve replacement. The patient was then transferred to an outside institution where she underwent cesarean section, followed the next day with an aortic tissue valve replacement with no compromise to the neonate. Four months after aortic valve replacement, she was alive and her fungal endocarditis had not recurred after completing a course of diflucan.
Sterile polyarticular arthritis and tendonitis associated with Group C Streptococcus bacteremia: Diagnostic and Treatment Dilemma

Angelina Hristov, Deborah R. Alpert MD, PhD; and Marnie Rosenthal DO, MPH

Objectives: 1) Recognize the association of Group C streptococcus (GCS) bacteremia with polyarticular arthritis and tendonitis. 2) Appreciate the challenge of managing inflammatory arthritis in the setting of sepsis.

Case Review: A 48 year-old Hispanic female with prior Chlamydial urethritis presented with one week of additive joint pain and fever. Symptoms began with right shoulder pain, followed by right knee, left elbow and left knee pain with swelling. She was hypotensive with blood pressure 82/54 mmHg and febrile to 102.3o F. On exam there was right subacromial pain with limited range of motion, left lateral epicondyle warmth, fullness and tenderness; and bilateral knee effusions and tenderness with limited range of motion. No rash was observed. Labwork revealed white blood cells (WBC) 20.5 k/μl (4.5-11 k/μl), hemoglobin 6.3 gm/dl (12-16 gm/dl); sedimentation rate 111 mm/hr (0-20 mm/h) and C-reactive protein 36.23 mg/dl (0-0.744 mg/dl). ANA, rheumatoid factor, Lyme serology, and human immunodeficiency virus serology were negative. Urine polymerase chain reaction for C. trachomatis and N. gonorrhoeae was negative. Two blood cultures grew group C streptococcus (GCS). Right knee joint aspirate revealed WBC 34,900 k/μl (0-2000 k/μl), normal glucose and no crystals; Gram stain and culture were negative. Transthoracic echocardiogram was negative for vegetations. A triple phase bone scan was negative for multifocal osteomyelitis. Nonsteroidal anti-inflammatory drugs (NSAIDs) were withheld due to anemia, ultimately attributed to menorrhagia from uterine fibroids. After convincingly ruling out endocarditis, osteomyelitis and septic arthritis, the patient was diagnosed with sterile polyarticular arthritis and tendonitis, felt to be reactive to GCS bacteremia. She completed a 21 day course of various targeted antibiotics, including ceftriaxone, and slowly defervesced. Discussion: Group A beta-hemolytic streptococcus may be associated with arthritis, as seen in acute rheumatic fever. There is significant overlap between Lancefield group A and group C streptococci species but direct association between invasive GCS and arthritis is unclear. Four cases of polyarthritis following group C and group G beta-hemolytic streptococci infection following pharyngitis are described in the literature. It was imperative to rule out joint space infection in this patient, as treatment of transient bacteremia, septic arthritis, and reactive arthritis differ. In our patient, polyarticular septic arthritis from hematogenous GCS translocation was ruled out with subsequent negative blood and negative joint fluid cultures. The case raised a treatment dilemma, as administering both NSAIDs and corticosteroids was contraindicated due to anemia and bacteremia, respectively. In this patient, GCS bacteremia resolved with antimicrobial therapy, and joint symptoms gradually improved. The case highlights an association of GCS bacteremia with sterile inflammatory arthritis and tendonitis, and demonstrates the challenge of diagnosing and managing these conditions simultaneously.
Meridian Health-Jersey Shore

The Golden Ticket: Novel idea for improving Transition of Care

Adam Kaplan, Carmela Rocchetti MD, Mayer Ezer MD

Background: Smooth transition of care (TOC) between the hospital and outpatient settings is important for patient safety and can lower hospital re-admissions. We developed a “TOC Resident Rotation” at Jersey Shore University Medical Center to address the additional barriers that complicate this for uninsured or under-insured patients. The goal was to identify patients in need of close follow up and decrease the “no show” rate for patients scheduled to be seen at our outpatient Family Health Center (FHC) after a recent hospitalization. Methods: Eligibility: uninsured or under-insured patients admitted to our service who would be seen at the FHC. The TOC resident would interview patients prior to discharge and schedule a FHC appointment. If a patient was a no-show for the outpatient visit, the TOC resident attempted to contact them and identify any barriers. After 3 months we identified that conflicts with the scheduled appointment date/time was an issue (20 occurrences), and the protocol was modified so that patients were given a separate paper or “Ticket” that listed the date, time, reason for the appointment, and the TOC resident contact information. Primary Outcome: A comparison of the historical no-show rate at the FHC to the no-show rate of the patients scheduled in this program. Results: The historical no-show rate at the FHC was 60%. After 3 months, 169 patients were scheduled over 37 days with 86 no-shows (51% no-show rate). 3 of the top 5 reasons for no-shows were communication barriers such as providing false contact information, agreeing on an acceptable appointment date/time, and importance of follow-up visit, upon discharge. In response we developed the personalized discharge Ticket. After one month utilizing the Ticket, 26 patients were scheduled over 8 clinic days, there were 9 no-shows (35%). Excluding the 2 patients still in the hospital, and 1 who rescheduled, the rate was 23%. Conclusion: Implementation of a TOC resident rotation allowed us to determine that the standard discharge documents, while thorough, did not facilitate patient awareness regarding the follow-up appointment. Addition of the discharge Ticket decreased the no-show rate from 51% to 35%, both an improvement over the historical rate of 60%. While the discharge documents are comprehensive regarding the follow-up plan and medications, discharge appointment information was apparently lost in the “noise” of the large set of paperwork. We will consider reformatting to help patients identify information crucial to efficient follow-up. In the meantime, we will continue what we found to be a successful program and subsequent studies will look at the readmission rates after implementation of the TOC program.
Case Report of Myotonic Dystrophy with Variable Atrial Flutter

Sehrish Memon, Faiza Fakhar MD, Abiodun Laoye, MD, Edmund T. Karam, MD.

1) Define Myotonic Dystrophy and review the associated cardiovascular complications 2) Discuss pathophysiology at histologic level of myopathy, conduction abnormalities and arrhythmias. Case: 29-year-old male with gradual muscle weakness, clinically diagnosed with myotonic dystrophy (MD) by a neurologist, was sent to the hospital for abnormal EKG. He was asymptomatic. EKG revealed atrial flutter with 2:1 block progressing to variable AV block from 2:1 to 8:1. He was started on low intensity heparin; underwent transesophageal echocardiogram, which was negative for left atrial appendage thrombus, showed LVEF 50% with mild tricuspid regurgitation. He underwent cardioversion for clockwise cavotricuspid isthmus dependent atrial flutter restoring sinus rhythm and then ablated with radiofrequency energy. Post ablation, anticoagulation was initiated for 30 days. Discussion: MD is a genetic and clinically heterogenous disease with two major forms, MD1 and MD2; MD2 is the milder form. MD1 is an autosomal dominant neuromuscular disorder caused by abnormal expansion of CTG trinucleotide repeats on chromosome 19 manifesting second through fourth decade of life. Trinucleotides are present in non-coding region of DMK(myotonin protein kinase) gene altering transcription and translation and affecting structures of adjacent proteins. It is a multi-organ disorder with clinical manifestations including myotonia, muscle weakness in a classic distribution of facial, temporal, sternomastoid and distal limb muscles, cataracts, endocrine, CNS, GI, and cardiac conduction abnormalities including arrhythmias with variable blocks. Respiratory and cardiovascular disease remains the most common cause of mortality during a 10 year follow up study of 367 patients accounting for 40% and 30% of fatalities respectively. Cardiac complications include AV conduction blocks, supraventricular and ventricular tachyarrhythmias, left ventricular systolic and diastolic dysfunction, ischemic heart disease and mitral valve prolapse. An investigational study of 12 patients and 7 controls found reduced levels in glucose metabolism and phosphorylation which is hypothesized as the mechanism by which cell damage and fibrosis occurs leading to conduction abnormalities and arrhythmias. A study by Nguyen et el analyzing autopsy of the cardiac conduction system in MD1 patients revealed fibrotic changes and atrophy of sinus/AV node, left bundle branch, fatty infiltration of bundle system and lymphocytic infiltration. Predominant histopathologic changes of cardiac muscle biopsies showed hypertrophied myocardial cells, fatty infiltration, interstitial fibrosis and myofibrillar degeneration. Atrial flutter and fibrillation are the most frequent tachyarrhythmia, occurring in about 25% of patients with MD1. If not treated promptly, it could be life threatening. Due to high incidence and mortality rate of cardiac disease, MD1 patients require a standard 12 lead EKG on first visit and at least every 12 months; a 24 hour Holter monitoring is indicated in symptomatic patients with fainting, dyspnea, palpitations or blackouts.
**Meridian Health-Jersey Shore**

**Disseminated herpes simplex virus 1 in a patient with systemic lupus erythematosus**

Janaki Patel, Sonja Abbassi MD; Marnie Rosenthal DO, MPH, Deborah R. Alpert MD, PhD

Objectives: (1) Understand the differential diagnosis of hepatic failure. (2) Recognize herpesviruses as a cause of hepatic failure in an immunosuppressed patient. Case: A 25 year-old black male with recently diagnosed systemic lupus erythematosus presented to the hospital for seizure-like activity. The patient was discharged two weeks prior following a diagnosis of Class IV/V lupus nephritis treated with mycophenolate mofetil (MMF) 3 g/day and prednisone 80 mg/day. Admission temperature was 101.7°F. Physical exam was unremarkable, with no skin rashes, oral or genital lesions. There was a history of recently treated Chlamydia urethritis. Labs were remarkable for AST 135 IU/L (10–42 IU/L), ALT 162 IU/L (10–60 IU/L), and serum creatinine 1.67 mg/dL (0.61–1.24 mg/dL). Blood cultures were negative. Brain MRI demonstrated multiple nonspecific foci of elevated signal intensity and electroencephalography was unremarkable. MMF was discontinued due to transaminitis. Over 5 hospital days, liver function tests increased exponentially, with peak AST 8603 IU/L and ALT 5846 IU/L. Viral hepatitis, cytomegalovirus and human immunodeficiency virus serology were negative; Epstein-Barr virus serology revealed prior infection. Acetaminophen level was undetectable. Anti-double-stranded DNA was marginally elevated and total hemolytic complement was slightly reduced. A liver ultrasound was normal. By hospital day 5 the patient developed anemia, thrombocytopenia (platelets 23 K/μL; 140–450 K/μL) and brisk upper gastrointestinal bleeding. Hypotensive shock, disseminated intravascular coagulation and hepatorenal syndrome necessitated emergent hemodialysis. The patient was treated with pulse steroids for possible lupus flare. On hospital day 6, herpetic-like vesicles were noted on the patient’s forehead, and there were new genital ulcers. He was emergently transferred to a hepatic transplant center and empirically treated with intravenous acyclovir. He rapidly expired due to a presumed diagnosis of disseminated herpesvirus infection. Vesicle viral culture and direct fluorescence antibody to HSV1 subsequently returned positive. Discussion: Herein we describe a young male on high-dose immunosuppression presenting with seizure activity and hepatitis, leading to fulminant hepatic failure. Workup for various causes of fulminant hepatitis was unrevealing. In the absence of cutaneous lesions, HSV hepatitis was not initially considered. Although a liver biopsy was not performed, this patient likely had fulminant hepatitis due to disseminated HSV1. Immunocompromised patients are at increased risk for developing complicated herpesvirus infections, and HSV hepatitis may develop in the absence of coincident rash. HSV hepatitis accounts for 1% of acute liver failure cases, with a mortality rate of 75%. Main features at presentation are fever (98%), coagulopathy (84%), and encephalopathy (80%). Due to its low incidence, HSV hepatitis is not often considered immediately in the differential diagnosis of acute hepatitis. This case illustrates the need for heightened awareness of herpesviruses as a cause of fulminant hepatitis in immunosuppressed patients, so that prompt empiric antiviral therapy may be initiated.
Atypical presentation of Epstein Barr Virus (EBV) meningoencephalitis in a young immunocompetent patient

Janaki Patel, Eric Costanzo DO, Marnie Rosenthal DO, MPH

Objectives: 1. Recognize an atypical presentation of EBV encephalitis with generalized seizures 2. Understand the limitations of the Heterophile Antibody test (Monospot)

Case: A 20 year old male with a substance abuse history was transferred to our facility with altered mental status and generalized tonic clonic seizures. The patient developed a sore throat and generalized weakness one week prior to admission and reported his girlfriend was diagnosed with mononucleosis. Two days prior to admission, the patient reported polysubstance abuse (ecstasy, cannabis, LSD, and ketamine). He became increasingly confused and sustained a witnessed generalized tonic clonic seizure. On admission the patient had an oral temperature of 100.6°F. Neurologic exam showed equal and reactive pupils with an intact corneal reflex. Gag reflex and response to verbal stimuli were absent, but withdrawal to painful stimuli was intact. Lumbar puncture and video electroencephalography was obtained and antiepileptics were initiated. Labs were only significant for leukocytosis of 17K/uL with 45% lymphocytes. Initial monospot on presentation was negative. Video electroencephalography revealed diffuse slowing with burst suppression pattern. Cerebrospinal fluid showed pleocytosis, 84/CUmm with 77% lymphocytes. Epstein-Barr Virus (EBV) PCR was positive and EBV Antibody to Early D Antigen was also elevated to >149.9 U/mL (normal 0.0 – 10.9). Computed tomography of the head showed exaggerated gray white matter differentiation at the bilateral cerebrum with poor CSF attenuation. Magnetic Resonance Imaging of the brain showed elevated signal intensity on the FLAIR-weighted sequence in the frontal and temporal lobes suggesting infectious inflammatory process. Acyclovir was initiated immediately. The patient was transferred to complete a slow recovery.

Discussion: Epstein-Barr virus (EBV) infection most often manifests as an acute febrile illness in the pediatric/young adult population with tonsillopharyngitis, lymphadenopathy and hepatosplenomegaly. Leukocytosis with lymphocytic predominance is seen. Neurological involvement occurs in 1-10% of patients with infectious mononucleosis. Central nervous system (CNS) symptoms can be the sole manifestation of EBV infection. Neurologic complications include serous meningitis, encephalitis, encephalomyelitis, transverse myelitis, neuritis, polyradiculoneuritis, and seizures. Encephalitis and meningitis are the most commonly encountered neurologic complications associated with infectious mononucleosis. Seizures and status epilepticus have been reported and most adults have a complete recovery. Pathogenesis of neural involvement is not completely understood but believed to be due to direct viral invasion into central nervous system, infiltration of cytotoxic CD8 lymphocytes into neural tissue or deposition of antibody-antigen complexes. Diagnosis is made by the presence of EBV DNA in CSF and/or positive serum antibody titer. Early in disease, the Heterophile Antibody test (monospot) may be negative. Acyclovir and corticosteroids have been recommended for treatment of EBV encephalitis, but their effectiveness is uncertain.
Objective: Recognize that CA 19-9 levels can be elevated in hepatic abscesses and can show progressive decline with appropriate treatment. Case Summary: An 85 year old female presented with left sided chest pain. PMH: breast cancer, modified radical mastectomy 20yrs ago, A-fib and hypertension. CTA chest showed no pulmonary embolism but incidentally showed cystic lesions in the liver, including one large complex cystic mass in the left lobe of the liver. Exam: Temp 101.8 F, BP:95/53 mmHg, Cardiac: irregularly irregular rhythm. Abdomen: generalized abdominal tenderness. Labs: AST:56 IU/L, WBC:12.8 K/uL, Neutrophils:80%. Cardiac workup was negative for ischemia on cardiac enzymes and Dipyridamole Stress Test. Metastatic disease to the liver was suspected. CEA and alpha fetoprotein were normal, but the CA19-9 was 1387 U/mL(normal 0-35). Liver ultrasound revealed right lobe simple cyst and a complex left lobe cyst containing thick septations, and soft tissue lobularity as well as internal echoes. MRI revealed no increased enhancement within the previously visualized complex cyst and no other abdominal pathology. Blood cultures were positive for Klebsiella pneumoniae. She underwent percutaneous drainage of the hepatic cyst, and the culture grew MRSA and Klebsiella. The hepatic cyst wall and abscess cytology study was negative for malignancy. She responded well to antibiotics. Serial Measurements of Serum CA19-9 showed progressive decline with antibiotic treatment. Discussion: High elevations of CA 19-9 can be seen in various benign diseases but elevations associated with liver abscess are thought to be rare. High CA 19-9 levels are usually associated with biliary and pancreatic malignancies, in the latter it is considered an accepted tumor marker and high concentration in serum signifies a poor prognosis. Benign hepatobiliary conditions can cause moderately increased levels except for acute cholangitis where sometimes high elevations can be seen. There are very few documented cases of liver abscess associated with high CA 19-9 levels and our Pubmed search did not identify prior reports associating it with MRSA liver abscess (search terms CA 19-9, liver abscess, MRSA). Most pyogenic liver abscesses are polymicrobial; mixed enteric facultative and anaerobic species are the most common pathogens. Staph aureus is only found in around 7% and usually results from hematogenous dissemination from a preexisting infection elsewhere in the body. The biliary tree is the most common source of pyogenic liver abscess and biliary epithelial cells can produce CA 19-9 constitutively. In our patient, abscess fluid culture came back positive for MRSA and CA 19-9 levels decreased after treatment with appropriate antibiotics which suggests that CA 19-9 might be a useful marker of treatment response in some cases of liver abscesses.
Interstitial Nephritis as an Extraintestinal Manifestation of Inflammatory Bowel Disease

Lucas Bazi, Aditi Shah

Inflammatory bowel disease comprises of two types of chronic intestinal diseases, Ulcerative Colitis and Crohn’s disease. A variety of other conditions are associated with inflammatory bowel disease, also known as extra-intestinal manifestations. Commonly associated findings include uveitis, hepatobiliary, pulmonary and skin complications. Renal disease is not a well-reported extra-intestinal finding. Some case reports have highlighted this association; however it remains unclear if the renal manifestation developed as an adverse effect of 5-ASA compound or from inflammatory bowel disease. It is important to differentiate whether nephritis is subtending from the disease or its therapy. We present a case of interstitial nephritis in a patient diagnosed with inflammatory bowel disease on no previous medications. A 32-year-old male patient, with no past medical history presented with a 3-year duration of intermittent non-bloody diarrhea, fatigue and progressive 14 Kg weight loss. Patient was evaluated by his primary care physician few days prior to presentation. On the routine lab work-up he was found to have hemoglobin of 6.8 and creatinine of 7.3 and was referred to the hospital. He was admitted with a preliminary diagnosis of chronic intestinal disease and acute kidney injury secondary to pre-renal azotemia. He had extensive workup, including urine and stool studies, renal ultrasound and serologic markers to rule out glomerulonephritides. Renal ultrasound revealed normal size kidneys and no hydronephrosis. His renal and gastroenterology work up, including ANCA antibodies, anti-GBM, HIV, serum complement levels, hepatitis panel, C.diff toxins, H.pylori serology and Celiac panel were all negative. He also underwent Esophagogastroduodenoscopy and Colonoscopy. The pathology from the small and large bowel biopsy were notable for mild acute ileitis with non-necrotizing granulomata and active moderate chronic colitis suggestive of inflammatory bowel disease. Over the hospital course, patient’s kidney function continued to deteriorate despite aggressive fluid hydration. Creatinine level increased from 7.3 to 8.2. At that point patient underwent kidney biopsy. Renal core biopsy revealed chronic moderate to severe active interstitial nephritis, moderate to severe tubular atrophy and interstitial fibrosis. Patient was started on mesalamine and prednisone for treatment of inflammatory bowel disease and concomitant acute interstitial nephritis. Renal complications may occur and in the case previewed, the progressive decline of the patient’s renal function occurred with no prior 5-ASA therapy. It is reasonable to conclude that the main precipitating cause of the patient’s severe interstitial nephritis was inflammatory bowel disease.
A rare form of vasculitis associated with hairy cell leukemia

Pierre El Hachem, Sharan Sharma, MD, Ramsay Farah, MD, Justin Louie, MS

Introduction: Malignancy-associated vasculitis is rare and occurs more often with hematologic malignancies than with solid malignancies. The associated hematologic malignancies are typically lymphoproliferative, or less frequently, myelodysplastic. In patients with hematologic malignancies, the most common presentation is a small vessel cutaneous leukocytoclastic vasculitis. Lesions manifest as palpable purpura that are preferentially located in dependent areas. We report a very rare case of granulomatous vasculitis associated with hairy cell leukemia. Case Description: 57 year old Caucasian male with a one year history of untreated hairy cell leukemia and bipolar disorder presented to the hospital for fever and rash. The rash is described as non-itchy, distributed over the bilateral arms and legs, and appeared one month prior to presentation. The fever was high-grade, intermittent and started two weeks prior to presentation. The review of systems was positive for one month history of headaches for which he started taking Tramadol 50 mg tablet three times a day. He also takes Zyprexa 2.5 mg a day. Physical examination was remarkable for a fever of 102.5 and a non-blanching papular erythematous rash distributed over his bilateral arms and legs. Laboratory findings demonstrated neutropenia with absolute neutrophilic count of 600. The patient was admitted for the management of neutropenic fever. Empiric antibiotic treatment, Cefepime, was given and within 5 days his fever resolved. Blood and urine cultures were negative. His rash was first thought to be drug-induced (possibly due to Tramadol), but no remission was noted after stopping the medication. ANA and ANCA serologies were negative. A punch biopsy of the skin lesion demonstrated granulomatous vasculitis with few eosinophils noted. Patient was discharged on methylprednisone tapering dose for 6 days and followed as outpatient by his dermatologist. During his follow-up visit 2 weeks later, the rash had 90% remission. Discussion: Patients with hairy cell leukemia occasionally develop vasculitis, more often polyarteritis nodosa or cutaneous leukocytoclastic vasculitis are described. Cutaneous granulomatous vasculitis is rare in the setting of a hematologic malignancies and it could be successfully treated with a short course of steroids.
Mount Sinai - Englewood

Invasive Liver Abscess Syndrome with Multiple Organ Involvement

Kourtney Ricks, Sharan Sharma

Invasive Klebsiella pneumonia liver abscess syndrome (IKPLAS) was first described in Southeast Asia more than two decades ago. IKPLAS, has shown increased emergence in Europe and North America. We describe a case report of a 60 year old male of Filipino descent with past medical history of new onset diabetes and hypertension admitted to our hospital with a 3-week history of fevers and chills who was diagnosed with IKPLAS. His workup showed multifocal liver abscesses with the largest one being 9.5x7.5x6.5cm, multiple pulmonary septic nodules, left thigh abscess, septic emboli in brain and bilateral endophthalmitis. He was treated with drainage of thigh abscess, intravitreal antibiotics and prolonged course of intravenous antibiotics. Five months after discharge his hepatic lesions have resolved and he does not have any residual symptoms. With only a handful of documented cases in the United States, our case adds to the emerging incidence of IKPLAS in North America. Host factors such as Diabetes mellitus has been shown to predispose patients to the development of pyogenic liver abscesses and its metastatic spread. However, microbiologic risk factors including the hypermucoviscosity phenotype and the presence of rmpA and magA genes also play a role in the virulence of this syndrome. Microbiologic risk factors were not known in our case. Past studies have shown that appropriate antimicrobial treatment with percutaneous drainage of liver abscess decreases mortality. However, the presence of multiple abscesses in our patient precluded the use of percutaneous drainage. Our case describes a rare, yet emerging syndrome of IKPLAS in the United States. Our case underscores the importance of early diagnosis and appropriate antibiotic treatment in order to prevent long term sequelae. Further, this case highlights the importance of including IKPLAS in the differential diagnosis when presented with patients of Asian ancestry with a history of diabetes and with unexplained fevers.
An Abrupt and Rapid Decline: A Case of Possible CJD

Prabin Sharma, Michelle Didesch, Nour Khouzam, Yelena Galumyan

We present a case of a 74-year-old female with medical history of hypertension, atrial fibrillation, fibromyalgia, and peripheral neuropathy referred to us with progressively worsening mental status over two years. She was working as a part time cashier until six months ago when she reached a state where she was not able to get out of bed without assistance. Her husband noticed increased anxiety, sensitivity to sounds, rapidly progressive dementia, diminished gait balance, personality change and inability to perform ADLs. She was evaluated by a neurologist in an outside hospital and had a negative work up. She didn’t have any family history of dementia or neurocognitive disorder. She didn’t have history of transfusions, hormone supplements, organ transplants or implants. On our initial evaluation, she was unable to carry on a conversation though she remained verbal with hypophonic, slow speech and was able to follow simple commands. She was able to name objects but unable to read sentences. She had normal cranial nerves, normal strength and sensation, no fasciculations, tremors or myoclonus, but she had increased tone throughout the body. CT head showed no intracranial abnormality. Work up including CBC, Chem-7, RPR, ESR, TSH, Vitamin B12, folic acid, Chest XRay were all normal. MRI brain with and without contrast showed mild bilateral cerebral periventricular, subcortical hyperintensities sparing the basal ganglia and thalamus. CSF studies were inconclusive in the background of a traumatic tap. However it revealed increased WBCs with predominantly PMNs, increased CSF protein and decreased glucose. Herpes PCR from the CSF was negative. EEG showed generalized slowing without epileptiform activity. During her hospitalization, she continued to refuse food and medications. A PEG tube couldn’t be inserted due to a large hiatal hernia. With the help of the surgical team, a J-tube was inserted. CSF studies revealed positive tau protein of 3517 pg/ml and the 14-3-3 test was also positive. The family decided to perform autopsy after her death if needed for confirmation of the diagnosis.

Based on our findings, we diagnosed our patient with a possibility of Creutzfeld-Jacob disease (CJD), a rapidly progressive fatal neurodegenerative prion disease. Despite a supportive clinical picture, MRI findings, positive tau and 14-3-3 protein, our patient didn’t meet all the WHO criteria for a definitive diagnosis of CJD in the absence of neuropathological confirmation. Patients presenting with progressive dementia are often falsely labeled with more common causes of dementia. We conclude that in patients presenting with rapidly progressive dementia with personality changes, CJD should be considered as a diagnosis even though it is a rare condition affecting less than one person in a million per year.
We present a case of 52 year-old male who was diagnosed with stage IV colorectal carcinoma 6 months prior, after he presented with changes in his bowels, decreased appetite and weight loss. His initial workup revealed a rectosigmoid mass with metastasis to the retroperitoneal lymph nodes. Systemic chemotherapy was initiated using FOLFOX-Avastin one month after diagnosis. He tolerated the regimen well. An interval CT scan 3 months ago had documented significant decrease in the size of his metastatic disease and, at the same time, his CEA had normalized. Approximately 1 month ago, the patient started developing mild headache, which was not associated with any other neurological symptoms. A brain MRI was performed, which showed no evidence of intracranial metastasis, but was remarkable for severe opacification of the maxillary ethmoid and sphenoid sinuses. On the morning of admission, the patient’s wife noticed the patient was slightly agitated. Subsequently, she witnessed a tonic-clonic seizure that lasted for 5 minutes following loss of consciousness for 10 minutes. EMS was called, and the patient was given 2 mg of lorazepam. In the emergency department, the patient continued to have tonic-clonic seizures, and 2 mg of midazolam was given. A CT scan of the head was done, which was entirely unremarkable. Physical exam was notable for elevated blood pressure (195/123), and the patient was post-ictal. Laboratory assessment revealed a WBC count of 21.8 mg/dL. During admission to the hospital floor, patient developed another episode of tonic-clonic seizure and he was given repeated doses of benzodiazepines. Despite medical intervention, the patient continued to experience generalized tonic-clonic seizure and was intubated for airway protection and subsequently transferred to the intensive care unit. An MRI of the brain revealed extensive cortical and subcortical T2 hyperintensity and edema predominantly in the parietal-occipital, frontal lobes, gangliocapsular region and cerebellum, characteristic of (Posterior Reversible Encephalopathy Syndrome) PRES. Involvement of the cerebellum suggested severe disease. The patient was taken off chemotherapy, and blood pressure was controlled medically. Subsequently, the patient did not experience any more generalized tonic-clonic seizure episodes and was discharged from the hospital. Status epilepticus is a life-threatening condition, where the brain is in a state of persistent, unremitting seizure. There is a plethora of etiologies of status epilepticus, including posterior reversible encephalopathy syndrome (PRES), and many can be reversible. Therefore, understanding the cause and early medical intervention is pivotal to more favorable outcomes. PRES is a clinical-radiological abnormality which should be considered as one of the differential diagnosis of status epilepticus, especially in patients on immunosuppressive therapy presenting with severely elevated blood pressure, confusion and lethargy. Immediate withdrawal from immunosuppressive medications and adequate blood pressure control has shown to resolve symptoms of PRES.
PULMONARY EDEMA: A LATE MANIFESTATION OF SALICYLATE TOXICITY

Prabin Sharma, Michelle Didesch, Nour Khouzam, Yelena Galumyan

We present a case of a 57 year-old male with significant medical history of previous back surgeries and chronic back pain maintained on high dose opioid analgesics who presented with complains of acute onset of shortness of breath, headache, diarrhea, diaphoresis, tremors, ringing in both ears and chest pain which was worse on inspiration. A week prior to arrival, he stopped his opioids abruptly and started taking multiple bottles of excedrin, advil, aspirin and tylenol. He was anxious and restless on examination but not tachypneic and was maintaining normal oxygen saturation. His vitals were stable. Blood work revealed a supratherapeutic level of salicylate (40.7) with normal acetaminophen level. Blood gas analysis revealed respiratory alkalosis without evidence of metabolic acidosis. CBC, chem-7, coagulation studies, liver panel, cardiac enzymes, EKG, alcohol level, lactate, urinary toxic drug screen were all normal. Chest XRay on admission didn’t reveal any infiltrates or pulmonary edema. He was treated with intravenous hydration, electrolytes supplementation and serum and urinary alkalinization. Blood gases, urine PH, serum PH, salicylate levels were repeated every 1-2 hours until the values were normal. Serum salicylate levels normalized within 18 hours and he improved clinically with resolution of most of his symptoms. The rate of intravenous hydration was gradually reduced in order to prevent fluid overload and then stopped. On the fourth day of hospital admission, he desaturated in room air and was febrile upto 103 deg F. Blood works with CBC, lactate and procalcitonin were all normal. Chest Xray showed bilateral pulmonary edema. He was treated with supplemental oxygen and diuresis. During the rest of the hospital course, he remained afebrile and his follow up chest Xrays showed resolution of pulmonary edema. Pulmonary edema is one of the severe complications of salicylate toxicity at supratherapeutic level and usually needs intensive management often with dialysis. On review of previous reports on salicylate toxicity, pulmonary edema is usually an early manifestation when the serum salicylate levels are high. Based on this case we would like to conclude that pulmonary edema could present as a late complication in cases of salicylate toxicity even after the salicylate levels in the serum have normalized.
Myxedema Case Presenting with Cardiac Tamponade

Murad Baba, Saurav Acharya

Case report Abstract  Introduction: We are reporting a case of severe hypothyroidism presenting with pretibial myxedema, infected wound, anemia due to GI bleed and impending cardiac tamponade. Background: Hypothyroidism is a well-known cause of pericardial effusion, cardiac tamponade and massive pericardial effusions rarely occur with severe hypothyroidism and myxedema. The accumulation of the pericardial fluid is gradual, and often does not compromise cardiac hemodynamic function. There is a relationship between the severity and chronicity of the disease with the presence of pericardial effusion. Case presentation: 41 year old Caucasian male patient with no past medical history, presented to the emergency room complaining of left lower limb swelling, ulceration and foul smelling discharge. His left leg has been swollen for the past 5 months. On presentation, vital signs were stable except for bradycardia. Physical examination revealed left lower limb swelling with deep infected ulcerations. Initial investigations revealed microcytic anemia, guaiac test was positive. CXR showed cardiomegaly suggestive for pericardial effusion. Echocardiography was ordered immediately and demonstrated moderate to large pericardial effusion with pre-tamponade physiology. A pericardial window procedure was performed and levothyroxine therapy was initiated. Severe anemia was treated with blood transfusion and he was worked up with upper and lower GI endoscopy. Conclusions: Impending cardiac tamponade is a rare initial manifestation of hypothyroidism. Timely diagnosis of pericardial tamponade is important for prompt intervention. Recurrent pericardial effusions are common, necessitating close follow-up. Myxedema-associated pericardial disease responds slowly to appropriate therapy with levothyroxine. Keywords: Myxedema, Pericardial effusion, Cardiac tamponade.
**Mount Sinai - Jersey City**

**Thrombotic Thrombocytopenic Purpura: A Case Presenting with Splenic Infarction**

Murad Baba, Saurav Acharya

Background: Microangiopathic Hemolytic Anemia (MAHA), thrombocytopenia, fever, renal failure, and neurologic symptoms comprise the cardinal features of thrombotic thrombocytopenic purpura (TTP). However recognition of thrombotic thrombocytopenic purpura can be difficult because of the variety of presentations. Case presentation: We present a case in which 79 year old hispanic male with past medical history of HTN and Osteoarthritis presented to the Emergency Room complaining of left side abdominal pain for 3 days. On presentation, vital signs were stable and the patient was afebrile. Physical examination revealed confusion and left flank tenderness. Initial investigations revealed Anemia, thrombocytopenia, elevated bilirubin, but normal liver enzymes and lipase. Abdominal CT demonstrated wedge-shaped hypo-enhancing lesions in the spleen suspicious for splenic infarcts, and the Brain CT scan was negative. Peripheral Blood smear showed Schistocytes. Plasma exchange was initiated immediately along with systemic steroids. Platelets counts improved but the confusion persisted. Brain MRI was done and showed multiple bilateral acute lacunar infarcts. Transthoracic echo revealed pulmonary artery hypertension. Both these brain MRI and echo findings suggests complications due to microvascular thrombosis. Patient’s hospital stay was complicated with gram negative hospital acquired sepsis due to E.Coli, the possible source being central venous catheter for plasma exchange. He maintained acceptable platelet count until being discharged from hospital on systemic steroids. Conclusions: Abdominal pain due to splenic infarction can be an atypical presentation of TTP. Prompt recognition of TTP is important because the disease responds well to plasma-exchange treatment. Mortality of around 70 to 90% without treatment can be reduced to less than 20% with plasma exchange. In addition to hemolytic anemia and thrombocytopenia, other microvascular complications like brain infarcts and pulmonary hypertension could manifest during the clinical course. Catheter related sepsis could complicate the treatment and hence prevention and early identification of which is important along with the plasma exchange. Keywords: Thrombotic thrombocytopenic purpura, Splenic infarction, Microangiopathic hemolytic anemia (MAHA), Plasma exchange
Role of exchange transfusion in acute chest syndrome: A case report and literature review

Parag Chevli, Rumana Khan; Arlinda Elezi; Angela Smolarz; Simon Badin; David Flores; Jyoti Matta

INTRODUCTION: Acute chest syndrome (ACS) is currently defined as a new infiltrate on chest radiograph in conjunction with 1 other new symptom or sign: chest pain, cough, wheezing, tachypnea, and/or fever. ACS have been a leading cause of hospitalization and death in adults with sickle cell disease. Here we presented a case of an acute chest syndrome who was successfully treated with exchange blood transfusion. CASE DESCRIPTION: A 27 year old African American male with history of sickle cell disease presented with complaint of 3 days of productive cough associated with right sided pleuritic chest pain and difficulty breathing. He also complained of subjective fever and chills. He denied any recent travelling, sick contacts, flu-like symptoms or similar complaints in the past. He was never admitted to the hospital for sickle cell crisis. He was an active smoker with 4-5 cigarettes/day for 5 years but denied alcohol abuse or illicit drug use. He was only taking Folic acid and denied any drug allergies. Physical examination was significant for scleral icterus and decreased air entry along with dullness on percussion over right middle and lower lobes of the lung. The vital signs were unremarkable. The laboratory examination showed leukocytosis with WBC count of 32,000. The hemoglobin was 9.7 and total bilirubin was 6.9. The chest X-ray showed right middle and lower lobe infiltrate with small pleural effusion. The patient was treated with antibiotics for pneumonia and possible acute chest syndrome. While patient was on the floor his condition deteriorated and he developed acute respiratory failure requiring intubation and transfer to intensive care unit. The CT scan of the chest was done which failed to show any evidence of pulmonary embolism but showed bilateral infiltrate more on the right side. Patient received broad-spectrum antibiotics, IV hydration, narcotics for the pain control and received simple blood transfusion. The bronchoscopy was also done which failed to show significant findings. Because patient’s condition failed to improve despite all the other measures; decision was taken to do exchange blood transfusion. After the second exchange transfusion, patient’s oxygen requirement on the ventilator decreased. Even though he underwent tracheostomy due to prolonged intubation; he was discharged to acute rehabilitation only on tracheostomy collar without any ventilator support. DISCUSSION: There have been no randomized trials to determine the optimal ACS therapy in adults with SCD; however, the mainstay of acute treatment is transfusion therapy. The need for transfusion and the modality of transfusion (simple versus exchange transfusion) depend upon the severity of the ACS episode. For moderate to severe episodes involving >1 lobe and with an oxygen requirement &gt;4 liters nasal cannula to maintain PaO2 &gt;70 mmHg or signs of clinical deterioration it is recommended to do exchange transfusion.
INTRODUCTION: Drug reaction with eosinophilia and systemic symptoms (DRESS) is a rare, potentially life-threatening, drug-induced hypersensitivity reaction that includes skin eruption, hematologic abnormalities, lymphadenopathy and internal organ involvement. Here we presented a patient who came with significant eosinophilia and subsequently was diagnosed with DRESS. CASE DESCRIPTION: A 70 year old male African American male came with complaint of 3 days of cough with yellowish productive sputum, left sided pleuritic chest pain, fever and chills. He also complained of development of rash about 2 days ago started on lower extremity extending to his chest and back. He was admitted in the other hospital 1 week ago for the similar complaints for which he was treated with antibiotics. His past medical history was significant for hypertension, diastolic heart failure, migraine and benign prostatic hyperplasia. His medication included aspirin, carvedilol, furosemide, ranitidine and tamsulosin. His primary care physician prescribed him topiramate around 2 months ago for migraine headache. He denied any toxic habits. Physical examination was significant for diffuse macular erythematous rash over bilateral lower extremity, chest and back. His vitals signs were unremarkable. The laboratory examination revealed WBC count of 20,000 with 46% of eosinophils, mild elevation of liver chemistry test and mild proteinuria. The CT scan of the chest revealed mediastinal and hilar lymphadenopathy. Based on significant eosinophilia and lymphadenopathy, decision was made to do open lung biopsy which revealed poorly formed granuloma and eosinophilic infiltration of pulmonary parenchyma. The biopsy of the lymph node showed reactive and hyperplastic lymph node. Based on history of recently prescribed topiramate, development of rash, significant eosinophilia, lymphadenopathy and organ involvement, the patient was given a presumed diagnosis of DRESS. The topiramate was stopped in the hospital. He was discharged home after his rash resolved and was followed in the clinic where he was found to have his eosinophil counts subsequently normalize. DISCUSSION: DRESS is a rare, potentially life threatening drug induced hypersensitivity reaction. Antiepileptic agents and Allopurinol are the most frequently reported causes. Although we think that this was probably the first case of topiramate induced DRESS. Identification and prompt withdrawal of offending drug is the main treatment of DRESS and that is why it is very important to have a high suspicion of this condition.
Increased completion of advance directives after an interdisciplinary team intervention


Background: Only one third of Americans have an advance directive (AD). Our study was conducted at an acute care teaching facility to determine if allowing other members of the health care team to participate in this discussion would increase the number of completed AD. The objectives of the study were to increase the number of patients completing an AD and to increase awareness concerning the importance of AD. Methods: We compared the number of AD obtained by physicians versus the number of AD obtained on a separate medical inpatient unit in which all nurses, social workers and resident doctors were educated by the palliative care team on a weekly basis and then encouraged to ask a patient about AD and follow-up with completion of the AD document by the patient representative. Results: During the first quarter of 2012 a total of 52 pts were identified on the specific medical unit and referred to the patient representative. Of the 52 patients, 24 completed the document. A retrospective analysis of medical records showed the average ADs completed on that same medical ward in the previous 6 months was 10-15%. After the termination of data collection, the increase in the number of AD completed was around 46%. Two medical wards, selected for comparison, had averages of around 5-15% of completed AD in the previous six months and after termination of data collection, the average number of AD completed was 5-10%. Conclusions: Currently in the United States the general knowledge about AD and the clinical implications in regard to end-of-life care in critically-ill patients is poor and has not significantly changed in the past decade. The present study demonstrates that initiating a directed discussion between patients and other medical personnel increases the number of ADs obtained among inpatients and shows a clear-cut benefit in the inpatient setting.
Introduction: A Dieulafoy’s lesion (DL) is an aberrant dilated torturous large-caliber arteriole in the submucosa, which protrudes through overlying mucosa making it vulnerable to rupture and bleeding. These lesions account for less than 5% of all upper GI bleedings, but with a significant mortality risks. There are cases reported, who required more than 100 units of packed red blood cells (PRBC), multiple endoscopies, hence a significant cause of economic burden for the institutions. High index of suspicion is often necessary to diagnose DL in otherwise healthy people with no risk factors and repeated GI bleeding. Herein we report a middle aged man with multiple admissions and multiple blood transfusions for recurrent upper GI bleed, who failed conservative approach, treated with partial gastrectomy as a last resort. Case Presentation: A 66 year-old African American male with history of Hypertension, Chronic gastritis presented to the ER with two episodes of dark colored vomiting associated with palpitations, dizziness and exercise intolerance. Patient denied of any other symptoms, alcohol and NSAID abuse. Physical examination revealed sinus tachycardia, dyspnea and orthostatic hypotension. Labs showed Hemoglobin: 5.3, Hematocrit: 16.9, Platelets: 592, BUN: 43, Creatinine: 1.3, positive stool guaiac with normal amylase, lipase and electrolytes. Patient was admitted to ICU, stabilized with intravenous fluids, 4 units of PRBC, 2 units of FFP transfusion, protonix drip, beta blocker and octreotide. Emergency Upper GI Endoscopy revealed erosive gastritis and a large blood clot in fundus which was irrigated with Epinephrine. In the next few days, patient had multiple episodes of hematemesis and malena with dropping hematocrit for which he undergone repeated endoscopies, IR-guided left gastric artery embolization which did not improve his condition. He received a total of 11units of PRBC, 6 units of FFP and 2 bags of platelets. Patient was taken to operating room and endoscopy performed to mark the site of bleeding with methylene blue and partial gastrectomy was performed. Patient’s symptoms resolved after 72 hour follow-up period with stable hematocrit. Pathology of specimen revealed a large caliber submucosal tortuous arteriole consistent with Dieulafoy’s lesion. Conclusion: Dieulafoy’s lesions remain an unusual, but significant cause of severe, uncontrollable, recurrent upper GI bleeding. Initial diagnosis of DL as the cause of an upper GI bleed can be quite trivial, causing delays, warranting a multidisciplinary approach. Endoscopy has replaced surgery as the single most powerful modality for diagnosing and managing Dieulafoy’s lesions. However, due to the intermittent nature of the bleeding, repeated endoscopies are warranted in order to establish the diagnosis. Angiography is an acceptable alternative in case of failed management with endoscopy. Laparoscopic surgery is only reserved for cases refractory to Endoscopic management.
Disseminated Extrapulmonary Tuberculosis in an Immuno-competent Patient: A case report

Rumana Khan, I. Shaik, S. Samineni, K. Malik

Introduction: Mycobacterium Tuberculosis (MTB) is the commonest of infections worldwide and can affect almost any part of the body. Pott’s disease of the spine with psoas abscess is currently a rare form of extra-pulmonary tuberculosis in developed countries; Intracerebral Tuberculoma is even more unusual. Early recognition is a challenge because clinical manifestations and radiology are confounding and can mimic malignancy, which is encountered more frequently. This case report is a rare entity of spinal osteomyelitis with epidural abscess and bilateral psoas muscle abscesses associated with multiple intracerebral Tuberculomas secondary to tuberculosis, in an otherwise healthy immunocompetent individual with no other traceable source. Case Report: A previously healthy middle aged African American female presented with 3-month history of low back pain and left lower extremity weakness associated with subjective fevers, chills and headaches, but no weight loss. She was immigrated to United States 15 years ago, since then had no TB exposure. Physical exam was significant for lumbar spinal tenderness with a positive straight leg raise test in the left side. Neurological exam revealed decreased motor strength and hypoparasthesia of lateral aspect of left lower extremity with no meningeal signs. MRI of the brain showed 8 ring-enhancing lesions varying from 2 mm to 5 mm at the gray-white matter junction. MRI of lumbar spine revealed disco vertebral osteomyelitis at L4 and L5, with epidural, extensive paraspinal, bilateral retroperitoneal and psoas abscesses. CSF and Sputum gram stain, AFB stain and cultures were sterile with negative cytology. HIV test was negative. Chest radiograph and CT scan showed no pulmonary infiltrates or fibrosis in the lung ruling out current or past pulmonary TB. Abdominal CT scan was normal. Pathological analysis of the psoas abscess aspirate revealed non-caseating granulomas, which grew MTB in culture. She was successfully treated with laminectomy and fusion, drainage of the epidural abscess in combination with percutaneous CT-guided needle aspiration of the psoas abscesses. Following surgery, the patient was relieved of pain with satisfactory recovery of power in her lower limbs. The patient was discharged on antituberculosis therapy for 9 months. After 2 months follow-up, MRI showed resolution of brain lesions. Conclusion: Extra-pulmonary Tuberculosis has emerged as a big challenge in developed countries with growing HIV population. Although the transmission is less, it can cause significant morbidity, mortality and economic burden. It can manifest without a pulmonary focus, therefore it is important to maintain a high index of suspicion in every person and not only in those who are immunocompromised. Early initiation of antituberculous treatment is the key to successful management.
Mount Sinai - Jersey City

**Multiple cryptogenic hepatic abscesses in a young healthy female treated with percutaneous intervention: A case report**

Rumana Khan, I. Shaik, A. Syed, S. Bellomo

**Introduction**  Hepatic abscess is a relatively rare condition with incidence rate of 8-15 per 100,000 persons in the United States. Cryptogenic pyogenic abscess with no evidence of infection is even less common. Developments of new microbiologic and radiologic techniques have decreased mortality rates drastically, yet the prevalence of liver abscess has remained relatively unchanged. If untreated, this infection remains uniformly fatal. The mortality ranges up to 30% even with treatment.  

**Case Presentation:**  Herein, we report a previously healthy 29 y/o African American female, who presented to the ER with complaints of Right upper abdominal pain and high grade fevers for 3 days. Pertinent medical history includes no allergies, drug or alcohol use, travel or occupational, contagious exposure. Review of systems was significant for gastrointestinal complaints such as nausea and vomiting, but no significant change in bowel movements. Physical exam was significant for toxic appearing, febrile young woman, hemodynamically stable with tachycardia. Abdominal exam revealed severe tenderness at right upper quadrant with guarding but no peritoneal signs. Laboratory evaluation was remarkable for leukocytosis with a left shift. Liver function tests showed elevated transaminases and INR. Abdominal ultrasound revealed multiple hypo-echoic hepatic lesions, some of which were complex cysts. Abdominal CT Scan confirmed 4 target lesions within the right and medial segment of left hepatic lobes. Patient was empirically commenced on broad spectrum intravenous antibiotics. The patient underwent radiologically guided percutaneous drainage of the abscesses. Aspirate from the abscess, blood cultures and amebic serology were negative. Furthermore, cultures for AFB and fungus were negative. Colonoscopy revealed no pathology. Retrospectively, an abdomen CT scan five years ago showed no focal abnormalities of liver excluding any possibility for superinfection of a previous cyst. Patient was treated with 12 weeks of IV Antibiotics with good clinical improvement.  

**Conclusion**  Pyogenic abscess constitute 80% of hepatic abscess, most of them are a direct causation of ascending biliary or portal tract sepsis, traumatic implantation of bacteria through the abdominal wall and superinfection of cysts or necrotic tissue. The most common pathogens are E. Coli, Klebsiella, Streptococcus and Staphylococcus species. Cryptogenic abscess constitutes up to 50% of liver abscesses for which the etiology is obscure even with extensive work up. Colonoscopy should be considered to rule out malignancy as a source of infection. Long term IV antibiotics and radiological intervention is considered as first line therapy. Open surgical intervention is appropriate if no clinical response achieved with 4-7 days of continuous indwelling catheter drainage or multiple, large or loculated abscesses are present with the risk of rupture.
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Mycobacterium Avium Complex- Immune Reconstitution Syndrome (MAC-IRIS) presenting as Chylous Ascites in HIV/AIDS: A rare case report


Introduction: Chylous Ascites (CA) is defined as the accumulation of chylous fluid in the peritoneal cavity secondary to lymphatic disruption. Most often it is associated with malignancy, surgery, trauma, cirrhosis and various inflammatory conditions of the abdominal cavity. Among HIV patients, Chylous Ascites has rarely been reported in association with Mycobacterium Avium Complex (MAC), Mycobacterium tuberculosis (MTB), Lymphoma, Kaposi sarcoma and, or as a complication of Immune Reconstitution Inflammatory Syndrome (IRIS). Case Report: A 44-year-old HIV infected African American male presented with three month history of progressive abdominal distension, chronic back ache and exertional dyspnea. His past medical history is significant for HIV/AIDS acquired by having same sex contact and disseminated MAC of duodenum and bone marrow. His home medications include Norvir, Preziesta, Truvada, Rifabutin, Fuzeon, Azithromycin, Ethambutol, and Mepron. On physical examination, vitals were stable with mild respiratory distress on lying position. Abdominal examination is consistent with ascites without any signs of portal hypertension and congestive heart failure. Abdominal imaging revealed extensive ascites, mesenteric and retroperitoneal adenopathy. Diagnostic and therapeutic paracentesis was done with a removal of five liters of grossly turbid milky fluid. The ascetic fluid analysis showed: Glucose: 98mg/dL, Neutrophils: 18%, lymphocytes: 31%, monocytes: 51%, Amylase: 37, Fluid LDH: 196mg/dL, Fluid Total Protein: 6.1mg/dL, Fluid Albumin: 2.3mg/dL, Triglycerides: 239mg/dL. Cytology was negative for malignant cells, gram stain and cultures were negative for bacteria, fungus and acid-fast bacilli after six-weeks. The diagnosis of Chylous Ascites was made based on visual appearance and triglyceride level of more than 110mg/dl. The previous medical records and labs reviewed which showed improvement in absolute CD4 counts from 90 to 130 to 222 and change in HIV-1 RNA PCR log copies from 4.99 to 2.42 despite of his worsening symptoms, which is consistent with Immune Reconstitution Inflammatory Syndrome (IRIS). This patient was successfully treated with repeated therapeutic abdominal paracentesis, HAART and MAC therapy and dietary modifications. Conclusions: IRIS manifesting as Chylous ascites in patients with HIV/MAC infection is an extremely rare condition with a significant morbidity and mortality. Antiretroviral drug-naive patients who start HAART in close proximity to the diagnosis of an opportunistic infection and have a rapid decline in HIV RNA level should be monitored for development of IRIS. The management depends on treating primary disease, repeated therapeutic paracentesis and dietary modifications. Some case reports have shown benefit of short course steroids which has to be proven in further studies.
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Pregnancy to CABG: A deadly case of Postpartum Myocardial Infarction in a young woman


Introduction: Pregnancy-related acute myocardial infarction is one of the rare but very important causes of morbidity and mortality in young, childbearing women. In a U.S based, large population study by James et al. reported the incidence of 6.2 cases per 100,000 deliveries with case fatality rates up to 30%. The occurrence is more common during postpartum period, manifests as unstable angina, myocardial infarction and sudden cardiac death. The most common pathophysiology in this cohort was atherosclerotic occlusion with or without intracoronary thrombus, spontaneous coronary artery dissection and vasospasm. The diagnosis and management is often delayed because of the low pretest probability in these young females, which carries a significant mortality risk.

Case presentation: Here, we report a 36 years old Filipino female with no significant past medical history presented to the ER with complaints of severe chest tightness started twenty minutes prior to arrival. She delivered her first baby twelve days ago via C-section secondary to oligohydramnios. It was around 6 am, she was breast feeding her baby and felt sudden onset of severe retrosternal chest tightness associated with shortness of breath. In the ER, patient was found in significant painful distress which was partially relieved with sublingual nitrate and morphine. Labs were unremarkable with normal first troponins.

Electrocardiogram revealed diffuse ST segment elevations in anterolateral leads and reciprocal changes in inferior leads. Code Heart was initiated with emergent catheterization. Angiogram showed severe two-vessel disease involving thrombotic occlusion of proximal and mid left anterior descending artery and proximal left circumflex artery. The Catheterization was complicated by coronary artery dissection and difficulty in advancing the catheter. Cardiothoracic surgeons were contacted immediately and Two-vessel bypass was performed. Further hospital course is complicated with cardiac arrest, shock and multiorgan failure. Patient was remained on ventilator in cardiac care unit and extubated on day 11.

Patient was discharged to rehabilitation facility on sixteenth-day and recovered fully after six-week follow-up.

Conclusion: Peripartum myocardial infarction very rare but has a significant mortality risk in young females of childbearing age. Irrespective of other risk factors, Pregnancy itself carries a high risk for thrombotic events. Pregnancy induced hormonal changes were believed to damage tunica intima and media leading to spontaneous coronary artery dissection in these patients. Due to fragile nature of intima, catheterization poses a high risk for dissection. Without proper identification and treatment, the mortality rates are very high. This case illustrates the importance of high suspicion for thromboembolic events, early recognition of symptoms, urgent diagnosis and immediate revascularization in peripartum women.
An Unusual Cause of Dilated Cardiomyopathy

Rotana Tek

HPI: 59 y/o w, f presents w/ intermittent CP, SOB, and palpitations for one year duration. S/Sx worsening in the past week. Dyspnea now with 3-4 blocks of walking. Resting improves S/Sx. Pt is a poor historian and non-compliant.  PMHx: Chronic Atrial fibrillation, Herniated Discs, COPD, MVA X8 Allergies: States she’s allergic to steroids Meds: Denies any herbal, OTC or Rx meds Surgical Hx: Cervical steroid injections Family Hx: No family recollection of early heart conditions Social Hx: Heavy tobacco smoker since early 20's; Denies ETOH and illicit drugs  PHYSICAL EXAMINATION: VS: 128/89, P: 102, RR: 20, O2: 94% (RA) & 98% (2L of O2), T 98.9F GEN: Alert and oriented but in mild distress HEENT: No JVD, No LN, No enlarged or nodular thyroid glands CV: S1+S2 Tachycardia; irregularly irregular; 3/6 pan-systolic murmur noticeably in mid-clavicular 4th ICS w/o radiation to axilla LUNGS: Bilateral crackles especially in lower posterior bases ABD: Unremarkable NEURO: Unremarkable MS: Unremarkable SKIN: Unremarkable EXT: +2 PTE; No cyanosis or clubbing; Cap refill < 2 sec LAB FINDINGS: CBC: Unremarkable CMP: Unremarkable PT/INR: Therapeutic after bridging and adequate anti-coagulation ACP: Negative x 3 BNP: 369 ABG : Unremarkable Thyroid Panel: Unremarkable D-Dimer: Unremarkable Urine, Blood, & Sputum Cx: Unremarkable  ECG: Afib/Flutter with RVR CT-Angio: cardiomegaly with pericardial effusion and B/L pleural effusion. No PE.  Echo: EF 20%, Cardiomyopathy with systolic and diastolic dysfunctions; segmental and global wall motion abnormality; mild pericardial effusion without tamponade; severe MR with enlarged LA.  Cardiac Cath: Indicates there is a fistula branching off the left main coronary artery going into the left atrium as indicated the arrow.  DISCUSSION: Dilated Cardiomyopathy (DCM) is associated with one third of CHF cases. DCM is characterized by reduced EF and dilated one or two ventricular chambers. DCM is classified into ischemic DCM and non- ischemic DCM. The most common cause of DCM, however, is idiopathic. Here, we will discuss a rare cause of DCM that has not been extensively described in the literature. The patient had a cardiac catheterization to rule out ischemic DCM, but a peculiar coronary anomaly was discovered as shown in the image on the left. Although no stenotic lesions were present. As depicted by the arrow, the patient has a rare coronary-cameral fistula, connecting the left main branch into the left atrium, causing abnormally enlarged LA, severe MR, and consequently DCM.  CONCLUSION: We demonstrated that a rare congenital coronary fistula that results in an abnormality of coronary circulation and a back-flow into the left atrium. This can be the etiology of DCM with an abnormally enlarged LA.
Psychosis is a well established complication of long-term, high-dose corticosteroid treatment, most notably in rheumatological cases. The presence of steroid-induced psychosis in pregnancy, a state saturated with stress hormones, represents a novel setting which poses unique diagnostic and management challenges for clinicians. We report the case of a 28-year-old primigravida at 25 weeks pregnant, without a significant psychiatric history admitted for increasingly bizarre behavior, agitation and pressurized speech with hallucinations suggestive of acute psychosis. Three weeks prior, she was treated with methylprednisolone intravenously, with a total dose of 40 mg twice daily for 5 days, for uncomplicated acute exacerbation of asthma at an outlying facility. Despite an appropriate taper upon discharge, she developed a psychosis with delusions and hallucinations over the next two days. During this time, on routine follow-up for asthma, she was noted to be hypotensive and readmitted. Corticosteroids were re-initiated but psychosis worsened. After early discharge against medical advice, the patient prematurely halted her remaining course of steroids mid-taper, three days prior to presentation to our facility. Clinical course in our facility was notable for gradually improving psychosis despite one notable episode of agitation controlled with haloperidol. Patient returned to baseline as defined by her family six days later, without the use of steroids or antipsychotics. She was discharged home per recommendation from psychiatry. It was argued that her psychosis was caused by the corticosteroid, since psychiatric disturbance is a well-known complication of corticosteroid therapy. To our knowledge, psychosis during pregnancy as a result of treatment with corticosteroids has not been reported previously. This is a surprising finding considering both are independently linked to psychogenesis. Their mutual occurrence suggests a dynamic therapy whereby additive steroid therapy may entail permissiveness of underlying predisposition to psychogenesis in pregnancy. That permissiveness may be related to the elevated stress drive in pregnancy primarily mediated by enhanced placental CRH secretion as a scheme to increase fetal glucose deliverance. This may induce psychiatric symptoms by increasing subcortical dopaminergic outflow and steroid-induced hypothalamic permissiveness of irrelevant stimuli. The reported case represents a likely case of steroid induced psychosis and reveals an important and novel addition to the differential diagnosis of acute onset psychosis in pregnancy.
Simultaneous Occurrence of Spontaneous Coronary Artery Dissection and Embolic Stroke in a Young Male Patient with history of Radiotherapy

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Acute coronary syndrome (ACS) due to spontaneous coronary artery dissection (SCAD) is a rare condition; moreover, the concurrent presence of ACS with a cerebrovascular accident would be considered generally implausible. We report a case of a young man with a history of Hodgkin’s lymphoma treated with chemo-radiotherapy ten years ago who presented with acute coronary syndrome caused by an extensive dissection of the right coronary artery, together with acute ischemic stroke. A 39 year-old male with a past medical history of hypertension, morbid obesity and Hodgkin lymphoma presented with severe typical chest pain at rest. Patient denied intense physical activity, smoking, alcohol intake or illicit drug use. Patient denied family history of coronary artery disease. Vitals: BP:151/93, HR:99, RR:14, T: 98.2°F, BMI:51. Cardiovascular exam showed no jugular venous distention, normal S1 and S2, normal heart sounds, no murmurs. Lung was clear. Extremity exam revealed palpable symmetric peripheral pulses with no pedal edema. EKG showed sinus rhythm, Q waves in inferior leads and tall R waves in leads V1-V2 consistent with posterior wall involvement. CBC and BMP were within normal limits. CK: 302, CK-MB: 17.42, Troponin I: 3.71. The patient was initially placed on IV unfractionated heparin, dual antiplatelet therapy, statin, &beta;-blocker, ACEI therapy and morphine. One-hour later, patient reported numbness and weakness in his left arm associated with left-sided facial numbness. Neurological exam revealed 1/5 muscular strength in the left upper and lower extremities, decreased pain sensation in the left side of the body, and normal deep tendon reflexes with a left upward Babinski reflex. CT head without contrast did not reveal any acute findings. CT angiography of head and neck did not show carotid or aortic dissection. Brain MRI confirmed two small bilateral non-hemorrhagic embolic infarcts over the right frontal lobe and left anterior parietal cortex. Cardiac catheterization revealed 70–80% occlusion in mid LAD, 30% in left main and spontaneous dissection of proximal to distal RCA; left ventricular systolic function was mildly impaired with ejection fraction of 40%. After cardiovascular surgery, interventional cardiology and neurology evaluation, patient was advised to follow conservative medical therapy as the use of thrombolytic therapy in stroke within the first three hours of presentation has shown to improve neurologic deficit, whereas the use of fibrinolytic agents could be detrimental in the case of SCAD. Two months later, the patient denied recurrent symptoms but persistence of left sided hemiplegia was noted. In conclusion, we believe the treatment of SCAD should be individualized depending upon clinical presentation and concomitant conditions like stroke, as presented in our clinical case. It is our intention to familiarize the medical community with this difficult clinical scenario and emphasize its evidence-based management.
An Unusual Case of Hypereosinophilic Syndrome (HES)

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Currently a rare condition, Hypereosinophilic Syndrome is considered a myeloproliferative disorder which may ensue into fatal multi-organ damage. Often asymptomatic or clinically vague, this syndrome results from a persistent Eosinophilia. We report a case of HES due to its unique and valuable educational contribution. Case Presentation: 45 year old male and recent immigrant from Dominican Republic with no significant past medical history, presented with six week history of non productive cough, fever, nasal congestion, and myalgia. Patient reported no relief with over the counter medications and initial physical examination was otherwise normal. Laboratory workup revealed a WBC of 25,000 µ/L and a significant Eosinophilia of 20,500 Cells/mm³, which further increased from 50% to 69% following admission. In addition, Hemoglobin and Hematocrit were normal as well as the rest of the basic metabolic panel. Further testing showed elevated ESR (57 mm/hr) and IgE levels (> 1,600 IU/mL), but normal IgA levels. Cultures for blood and urine resulted negative. Analysis for stool and sputum showed no Ova, Parasites or any other organisms. Dengue, Strongyloid, TST and HIV showed negative serologies including a negative blood smear for Malaria. Furthermore, nasal secretions were negative for Influenza Antigen. Imaging testing illustrated atypical pneumonia and mastoid disease on CXR and Head-CT scan, respectively. Upon admission to the regular floor, the patient was treated for community acquired pneumonia using IV Unasyn® and given respiratory treatment with Albuterol. On third day of stay, patient developed severe respiratory distress needing immediate transfer to the ICU. He was promptly treated with IV steroids and responded positively. In consideration of persistent unexplained Eosinophilia, Trans-bronchial biopsy, Bronchioalveolar Lavage and Duodenal Aspirates were performed, which tested negative for ova, parasites or malignancy. Consecutively, peripheral blood smear showed marked eosinophilia with no evidence of atypical or malignant cells. After numerous inconclusive studies to determine the etiology of eosinophilia, diagnosis for HES was met by exclusion criteria. After 6 days of IV steroid therapy, WBC as well as Eosinophil count decreased to normal ranges. He continued to be asymptomatic and was followed up for a period 6 months without any complications. Discussion: According to literature, HES manifests with eosinophilia ≥ 1500/µL on two occasions without any recognizable etiology. Incidence of HES is estimated at 0.5-1.0 cases per 100,000 persons per year and is 9-fold higher in men than women. Usually asymptomatic, HES is incidentally detected on a CBC performed for other purposes. However, HES could be fatal as a result of multi-organ failure if clinically missed. Physician’s recognition of this syndrome accompanied with prompt diagnosis and management could decrease mortality in this seldom identified condition.
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An Atypical Case of Acute Myocardial Infarction Linked to Neuroleptic Malignant Syndrome (NMS)

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Psychiatric patients often develop NMS when taking Neuroleptic medications. Current literature indicates a mortality rate of 10-20% in NMS alone, which significantly rises if linked to acute MI. However, very few reports have been documented regarding this association. We report a case of NMS causing acute MI due to its rarity and great educational contribution. A 49 year old African American female with a significant history of Schizophrenia and Renal Insufficiency presented to ED with a 2-day history of SOB aggravated by a new onset of high grade fever for the last two hours. Consecutively, she developed severe tightening chest pain with radiation to the left arm accompanied with marked sweating. She also reported feeling progressive stiffness throughout her body. She has no history of CAD or pertinent risk factors. Medications included Risperidone, Trazodone, Aripiprazole and Benztropine. In the ED she had a BP 104/60 mmHg, HR 88 bpm, RR 23/min, T 101.1F and SO2 95% [3L NC]. Physical exam showed marked diaphoresis and limited range of motion in all extremities. EKG revealed acute inferior STEMI. During cardiac catheterization her RCA was > 90% occluded and consequently stented; patient remained markedly febrile with mild generalized stiffness for which broad spectrum antibiotics was empirically initiated. Upon admission to ICU patient became tachypneic, lethargic, diaphoretic and had clear chest on auscultation with BP 97/46 mmHg, HR 156 bpm regular, SO2 95% [3L NC], T 102.4F but no chest pain. Fever persisted in spite use of antibiotics and Tylenol. ABG’s showed Respiratory Alkalosis, Urine/Blood cultures and Urine Drug Screen were negative, Glucose (POC) normal, Ca level was 8.5 and CXR was clear. CPK level and WBC increased from 98 to 1343 and 28.5 to 40.7 (3-4 hrs), respectively, and Renal function was clearly decreased. Post-Cath repeat EKG was unremarkable. She developed generalized body stiffness compromising her normal breathing along with autonomic instability. NMS was immediately identified, aggressive IV hydration started, Bromocriptine along with Benztropine given and Neurology/ Psychiatric consults were placed. Patient was closely monitored and improved considerably few hours later; body stiffness and breathing improved without the need for intubation. She regained full motion of all extremities, renal function improved, cultures continued to be negative, CPK trended down to 733 and finally she became stable (HR 110 bpm, RR 20, SO2 99% [2L NC] and T 98F). NMS is not a diagnosis to be missed. It is a medical emergency that warrants immediate intervention. However in association with acute MI mortality is increased significantly.
Lynch syndrome is an autosomal dominant inherited cancer predisposition syndrome. Patients inheriting this predisposition are susceptible to colorectal, endometrial and other extracolonic tumors, with an average age of onset of 45 years for colorectal cancer. Lynch syndrome is associated with a germ line mutation in one of several DNA mismatch repair (MMR) genes. DNA mismatches commonly occur in regions of repetitive nucleotide sequences called microsatellites. The expansion or contraction of these regions is termed microsatellite instability and is a molecular hallmark of Lynch associated cancers. Tumors can be tested for MSI using polymerase chain reaction and immunohistochemistry usually helps in detecting the loss of MMR genes thus prompting further genetic testing. We report a case of 42 year old male with history of splenectomy at five years of age who presented with abdominal pain and nausea. CT scan of abdomen/pelvis revealed a cecal mass. Colonoscopy was done and he underwent right hemicolecotomy. Pathology report confirmed invasive mucinous adenocarcinoma of cecum (T2N0M0). Tumor tissue tested positive for high microsatellite instability (MSI-H) with abnormal immunohistochemistry. The laboratory data showed anemia (Hemoglobin 6.5) and thrombocytosis (Platelets 1059) at presentation. His hospital course was complicated by pulmonary embolism, portal and mesenteric vein thrombosis. In light of persistent thrombocytosis and extensive thrombotic disease, a bone marrow biopsy was done and showed megakaryocytic hyperplasia. Chromosomal analysis revealed absence of JAK2 V617F mutation and absence of Philadelphia chromosome. Further workup included Fluorescence in situ hybridization (FISH) analysis using a myelodysplastic syndrome specific set of FISH probes, which were all within normal range, thus prompting the diagnosis of essential thrombocytemia. The patient was started on hydroxyurea and eventually discharged for further follow up in specialized center. Our traditional understanding of what is considered the extracolonic manifestations of Lynch syndrome continues to change as our knowledge evolves. The most common extracolonic tumor is endometrial cancer and other sites of increased risk for neoplasm development include the ovary, stomach, hepatobiliary system, small bowel, renal pelvis and ureter. However, over the last few years, there have been reports of hematologic malignancies in families with Lynch syndrome. Some of those reports included leukemia, non-Hodgkin’s lymphoma and myeloma. Our case report describes a patient with Lynch syndrome and essential thrombocytemia. Recent studies have investigated the role of novel mutations in myeloproliferative disorders. One of those mutations is IKZF1 located on the short arm of chromosome 7, which also carries the PMS2 mismatch repair gene found in 14% of Lynch syndrome cases. A genetic predisposition encompassing both the diseases seem very likely and does call for further studies for better comprehension and understanding of the possible different hematologic manifestations of this syndrome, thereby increasing our index of suspicion and advancing our patient care.
**Raritan Bay Medical Center**

**Bulbar Post-Polio Syndrome**

Jenil Gandhi

Introduction - Post-polio syndrome is a rare condition that affects approximately 25-50% of individuals who have previously recovered from acute poliomyelitis. Symptoms include new onset of weakness, fatigue, and pain that typically appear decades after the initial bout. A 70-year-old man with Bulbar Poliomyelitis and Marfan Syndrome was admitted for sepsis secondary to aspiration pneumonia. Since two months, patient had complained of worsening fatigue and general malaise. Weakness later followed as patient started to have difficulty swallowing and articulating speech. The mother reported finding the patient lying on the floor the night following admission. On arrival he was toxic-appearing, pale, and in mild respiratory distress. He denied any headache, loss of consciousness, seizure, or dizziness. White sputum productive cough was significant for two months. On physical examination, prominent muscular atrophy, kyphosis, and pectus excavatum was accompanied by harsh rapid breath sounds and bibasilar coarse crackles on auscultation. Neurological exam was significant for intact motor (4/5) without fasciculations. He had a fever of 102.4oF, leukocytosis (17.0 K/uL), hyponatremia (130 mmol/L), elevated creatine kinase (1802 U/L), high cortisol (26.9 ug/dL), and bilateral infiltrates on chest x-ray. EKG and EEG respectively showed sinus tachycardia and signs of mild encephalopathy. Non-contrast CT of the head was unremarkable. He was subsequently started on IV antibiotics and steroids. One day later, a computed tomography (CT) of chest was performed to confirm previous finding in addition to bronchiectasis, mediastinal/hilar lymphadenopathy, and evidence of underlying emphysema. Rapid HIV testing and blood/urine cultures were negative. Evaluation of dysphagia yielded poor clearance of both solids and liquids, which led to insertion of percutaneous endoscopic gastrostomy (PEG) tube. He was eventually discharged to nursing home in stable condition with a plan to follow up. Discussion - Represented by 2% of paralytic polio cases, bulbar polio occurs when poliovirus destroys cranial nerves within the bulbar region of the brain stem. The result is weakening of muscle groups that are supplied by these nerves leading to respiratory distress, dysarthria, and dysphagia. Regardless of its precise pathophysiology, therapy strategies remain to be largely compensatory. Small portion of food intake while sitting down may show symptom relief. PPS with respiratory involvement necessitates special management such as breathing exercises and chest percussion with a stethoscope on regular occasions for surveillance of secretions. Severe cases may entail chronic ventilation support or tracheostomy. Failure to properly manage PPS with respiratory involvement can increase the risk of missing aspiration pneumonia. Sleep apnea can also arise as a complication. Smoking cessation and vaccination against influenza virus is strongly recommended for drastic improvement. In summary, clinicians should be vigilant of potentially fatal complications, especially respiratory compromise, arising from this progressive debilitating disease.
Multisystem diseases with Autoimmune and Autoinflammatory features in patient with Vitiligo and Familial cafe au lait spots


Autoinflammation and autoimmunity are pathological processes direct against self and cause monogenic or polygenic systemic diseases affecting a large number of organs, frequently musculoskeletal system. Autoinflammation employs innate immune system to cause tissue damage directly; whereas autoimmunity initiates with innate immune system that subsequently activates the adaptive immune system to cause tissue damage. Autoinflammatory and autoimmune diseases might be considered two ends of a large spectrum of immunopathology in light of their similarities. Both cause chronic activation of immune system and eventually result in tissue inflammation in genetically susceptible individuals. A 46 year old female with vitiligo, rheumatoid arthritis (RA), diabetes mellitus with features of latent autoimmune diabetes in adults (LADA), hypertension, hyperlipidemia, glaucoma, familial café au lait spots, presented with abrupt onset of severe respiratory distress due to orofacial angioedema without associated itchiness, pain, or urticaria in other body parts. Patient had similar symptoms one month ago. Home medications include methotrexate, folic acid, lisinopril (started 1 year ago), adalimumab, simvastatin, omeprazole, metformin, neurontin. Family history is significant for rheumatoid arthritis in grandfather, café au lait in grandmother, mother and sister. She is the only family member with vitiligo and angioedema. On admission, vitals: BP 164/95 mmHg, pulse: 86, respiration: 18, temperature: 98.3°F, oximetry: 98%. Patient was given famotidine, methylprednisolone, diphenhydramine, epinephrine and admitted to critical care unit. She was discharged three days later. This report not only reemphasized the delayed side effect of ACEI, which may manifest months to years after administration; but more importantly the intriguing coexistence of multiple diseases that are more or less related to autoimmune/autoinflammation. The concomitance of vitiligo, RA and LADA strongly suggests one or more common genetic component of susceptibility to autoimmunity/autoinflammation. The NALP1 represents one major susceptibility gene for vitiligo and associated autoimmune disorders. The activation of its product NACHT leucine-rich-repeat protein 1 (NALP1), a regulator of innate immune system recruits other proteins to the NALP inflamasome, which controls the activation of interleukin-1β (IL-1β). IL-1β then subsequently recruits adaptive immunity. The pathogenesis of hypertension, hyperlipidemia, glaucoma, and angioedema may also involve aberrant immunological reactions. Therefore, it is reasonable to presume that the multi-system disorders in our patient were direct or indirect resultant of immunopathology. Further investigation and clarification of roles played by autoimmune/autoinflammation in different diseases will not only facilitate the understanding the pathogenesis and classification, but promote the development and early incorporation of immunomodulatory therapies in a large number of patients and effectively reduce complications.
Being one of the most critical endocrine emergencies, thyroid storm is rare but often fatal without immediate and aggressive management. The overproduction of thyroid hormones fosters an increased metabolic status encompassing all systems, which may eventually cause failure of multiple organs, with cardiorespiratory failure being the most common cause of death.  A 51 year old female with past medical history of hypertension, myocardial infarction (MI), and asthma presented with sudden onset of severe respiratory distress. Detailed history could not be obtained on admission due to her clinical instability. The vital signs upon admission were blood pressure: 180/135 mmHg, heart rate: 170 bpm, respiration rate: 44/minute, oxygen saturation: 70 % on room air, temperature: 101 °F. She was agitated and using accessory muscle for breathing. On physical examination, there were mild wheezing and rales. She was intubated and admitted to intensive care unit. Biochemistry demonstrated Troponin I: 0.51 (<0.3 ng/ml), NT-proBNP: 2380 (<300 pg/ml). Chest X-ray revealed pulmonary edema. Initial medical treatment was aimed at cardiorespiratory failure likely related to non-ST segment elevation MI and presumed asthma exacerbation. Medications included methylprednisolone, furosemide, heparin drip, aspirin, clopidogrel, metoprolol. However, further evaluation after extubation revealed symptoms implying hyperthyroidism, including chronic palpitations, tremors, anxiety, insomnia, heat intolerance (2 years), and weight loss of 100 lbs (3 years); physical features of proptosis, goiter, tachycardia, and hand tremors. Thyroid function tests demonstrated (with reference) TSH: <0.005 (0.45-4.50 uIU/ml), free T4: 5.05 (0.82-1.77 ng/dL), free T3: 10 (2.0-4.4 pg/ml), and total T3: 269 (71-180 ng/dL); thyroid stimulating immunoglobin: 446 (0-139 %), anti TPO antibody: 67.0 (0-34 IU/ml). Thyroid imaging studies illustrated multinoduler goiter and increased I123 uptake. Thyroid storm was diagnosed according to the point system developed by Burch and Wartofsky (1993), the score was 75 (temperature 15 points, agitation 10 points, heart rate 25 points, pulmonary edema 15 points, suggestive history 10 points). A score of ≥45 is highly suggestive of thyroid storm. Methimazole was subsequently administered. The cardiorespiratory failure was likely related to tachycardia-mediated congestive heart failure and thyrotoxic myopathy-related respiratory muscle fatigue and failure which were reversible upon early correction of thyrotoxicosis. Acute respiratory distress in an asthmatic patient with hyperthyroidism in the absence of significant wheezing may be a sign of underlying worsening thyroid pathology with imminent thyroid storm. A high clinical suspicion and early recognition is crucial in patient management.
Mesenteric cyst versus Lymphangioma: a clinical conundrum

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Introduction Mesenteric cysts and cystic mesenteric tumors are very rare abdominal growths. They may be localized all over the mesentery, from duodenum to rectum, however, they are mostly found in the ileum and right colon mesentery. Case report 43 year old Hispanic male presented to ER complaining of abdominal pain and difficulty in breathing in recumbent position. Patient reported no significant past medical, surgical or family history. Physical examination was unremarkable except for an abdomen that was firm and distended with no fluid thrill. Bowel sounds were present but diminished. All laboratory tests were normal except a mild normocytic, normochromic anemia. CT scan of abdomen revealed a 16.5 x 25 x 23 cm rounded, well circumscribed, hypo dense lesion/mass within the peritoneal cavity. A further MRI of abdomen and Pelvis with and without contrast revealed a 25.3 x 16.7 large complex proteinous cyst in the midline of abdomen centered in the mesentery/ peritoneum without any findings to suggest that it is originating from any adjacent structure. Radiological differential diagnosis included a mesenteric cyst or an intra abdominal cystic lymphangioma. Laparotomy was done & 30 cm intra-abdominal cyst was removed. Histopathological examination revealed hemorrhagic intra-abdominal cyst which is unilocular thin fibrous pseudo-capsule containing fibrin debris & bloody fluid.

Discussion Mesenteric cysts are quite rare intra-abdominal lesions. Fortunately they behave mostly as benign tumors, while malignancy accounts for 3% of the cases, arising gradually or de novo. This is in sharp contrast to intra abdominal cystic lymphangiomas who often present clinically and radiologically similar to mesenteric cysts. However histologic and ultrastructural evidence suggests that they are pathologically different entities. Differentiation of these lesions is imperative since lymphangiomas may follow a proliferative and invasive course. Accurate pre-operative diagnosis is quite tough due to the rarity of the entity and the lack of specific symptoms and signs. Clinical imaging (ultrasound, CT scan or Magnetic Resonance Imaging-MRI) may aid in the correct diagnosis. Careful interpretation of images is thus vital in making the correct diagnosis and formulating further management decisions.
Troponin-I elevation due to alcoholism in absence of acute coronary syndrome

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Troponin-I elevation due to alcoholism in absence of acute coronary syndrome

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Cardiac Troponin I (cTnI) is a sensitive and specific marker for myocardial injury and for the diagnosis of an acute myocardial infarction (AMI). cTnI is known to be elevated in a variety of clinical scenarios without evidence of acute coronary syndrome (ACS). Although ethanol intake is known to cause myocardial damage in susceptible patients; elevation of high sensitivity cTnI has not been reported with alcoholism specially when there is no objective evidence of myocardial damage. We present a case of a 55-year-old Hispanic male who was admitted for alcoholic intoxication. He has hypertension for 5 years, which has been well controlled with medication. He has been drinking alcohol for about 10 years (about 43g/d). The alcohol level on admission was 375.2 mg/dL (normal range < 10 mg/dL). The liver function and renal function tests were normal. Incidentally he was found to have elevated cTnI [0.36 ng/mL on presentation and 0.42 ng/mL (after 8h), 0.38 ng/mL (after 16 hours)], normal range <0.3 ng/mL]. The patient denied symptoms of chest pain, shortness of breath, or palpitations. Serial electrocardiograms (EKGs) did not show any dynamic ST changes. He had normal left ventricle function on Echocardiogram. The single-photon emission computed tomography pharmacological stress was also normal, therefore cardiac catheterization was not deemed necessary. He was treated as non-ST segment elevation myocardial infarction (NSTEMI). Two days later patient was discharged home. Five months later, this patient was admitted again because of nausea and vomiting after binge drinking of alcohol. He had no ACS symptoms and was hemodynamically stable. His liver function and kidney function were normal. The cTnI level was again found to be elevated in range of 0.31-0.34 ng/mL in three-set serials (<0.30 ng/mL). His EKGs and Echocardiogram were again normal. One day later patient became symptom-free and then discharge home. In this case, patient had elevated cTnI with normal liver and renal function, without pulmonary disease, or other medical conditions which may induce elevations in cTnI as literature reported. We postulate that chronic alcoholism induced elevations of cTnI in this patient. The mechanism of elevated cTnI may be related to alcohol induced-myocyte injury, very early myocardial damage, independent of any coronary artery disease. This finding may help clinicians to better manage patients with elevated cTnI by looking for the true underlying reason and avoid unnecessary invasive cardiac testing.
Introduction

Spontaneous hydropneumothorax is a rare, but possible presentation of malignant pleural mesothelioma (MPM) related to asbestos exposure. A high index of suspicion of malignancy should be maintained in older asbestos-exposed patients with hydropneumothorax. Also, the strong inflammatory and atherogenic properties of asbestos may play a central role in the pathogenesis of cerebrovascular and ischemic heart disease in such individuals. Case report

An 80 year old male presented with dizziness and dyspnea. Sixty-six years prior to admission (PTA) patient started asbestos exposure in high school and laboratory. Fifteen years PTA patient developed myocardial infarction, atrial fibrillation, and subsequently heart failure with impaired left ventricular ejection function, and AICD was placed 5 years ago. Medical history also included hypertension, hypercholesterolemia, coronary artery disease, and dilated cardiomyopathy. Vitals: BP: 112/72, Pulse: 61, Respiration: 15, Temperature: 98 &#61616;F, O2 saturation: 93% on Room air. Chest X-ray revealed bilateral pleural effusions, and left-sided loculated hydropneumothorax. CT chest demonstrated multiple air fluid levels in the left pleural space, round atelectasis and pleural thickening consistent with asbestos exposure. EKG illustrated atrial fibrillation. Echocardiogram showed calcified mitral annulus, dilated left ventricle, dyskinetic apex, impaired left ventricle systolic function, and pulmonary hypertension. Calcification of carotid and abdominal aorta were also demonstrated by imaging studies. Blood test showed RBC: 3.82 M/ul, WBC: 2.4 K/ul, BNP: 2715. Thoracentesis collected 650 ml cloudy yellow fluid. Pleural fluid tests showed LDH: 114 IU/L, protein: 2.8 gm/dL (serum protein 5.5 g/dL), glucose: 77 mg/dL, pH: 8.0, RBC: 7920 /ul, WBC: 1188 /ul, slightly blood-tinged, no evidence of malignancy. Discussion

We present a rare spectrum of asbestos-related cardiac and pleural pathology presenting as hydropneumothorax and multiple cardiovascular disorders. Chest CT-scan may help rule out an underlying malignancy. In cases of recurrent or persistent pneumothorax, pathological study of pleurectomy samples may be employed. In addition, asbestos increases the risk of cardiovascular diseases; regular screening tests are warranted in patients with this condition. The underlying pathophysiological basis may involve inflammation and oxidative stress with derangement in molecular signaling and gene expression profiles by asbestos exposure, such as activation of Ras-related C3 botulinum toxin substrate 1, which is not only related the initiation of A-fib, but also the pulmonary fibrosis, development and invasiveness of a variety of cancers.
INTRODUCTION: Stent placement is one of the most common procedures in relieving obstruction and maintaining patency of various outflow tracts. Nevertheless, infection of these stents is also an emerging concern for the physicians. We present a rare case of stent infection which highlights the successful treatment strategy for such scenarios.

CASE PRESENTATION:
64 year old male presented to us with fever of 101-102 F, myalgia and chills for eleven days. One year prior to admission the patient was diagnosed with metastatic pancreatic cancer to liver and underwent liver ablation, chemotherapy, biliary drainage and stenting. MRI one month later had no evidence of residual tumor. Past medical history was significant for hypertension, dyslipidemia and pancreatic carcinoma with liver metastasis. No history of smoking, alcohol or drug abuse. He had no history of recent travel, no pets at home. His vaccinations were up to date. On admission P-78/min, BP-100/60, T-102.2F, RR-18/min, Sat-98% on room air. Patient was alert, looked toxic. No jaundice, no oral candidiasis, no parotid enlargement, no adenopathy. There was a porto-cath on left upper chest which was clean. Abdomen was soft, bowel sounds were active, liver span–11 cm. There was no tenderness, distention or ascites. Labs were significant for WBC-55.2k/μL, and Bands-13k/μL. Liver function tests were within normal limits. CT Abdomen showed air within biliary tree, a biliary stent and an 8.6 cm x 7.0 cm intrahepatic abscess. The patient underwent CT-guided drainage for the liver abscess. The abscess cultures grew Escherichia coli, Pseudomonas aeruginosa, Streptococcus viridians. The patient was successfully treated with six week course of Imipenem.

DISCUSSION:
Bacterial adherence and growth on solid surfaces as biofilms are naturally occurring phenomena. A biofilm is a structured consortium of bacteria embedded in a self-produced polymer matrix consisting of polysaccharide, protein and DNA. Biofilm formation occurs on devices placed in the human body such as urethral, ureteric, prostatic and biliary stents. It has major impacts, with potentially devastating consequences such as resistant infection and occlusion, which sometimes necessitates the removal of the device.

In experimental studies non cardiac stents become occluded by six bacterial species, designated as Leitbakteria which include Pseudomonas aeruginosa being the pioneer colonizer followed by Klebsiella pneumoniae, Enterococcus faecalis, Enterobacter aerogenes, and two unculturabe bacteria distantly related to Escherichia coli and Shigella sonnei. Therefore we summarize that anti microbial choice for stent biofilm infection should be directed against Leitbakteria specifically pseudomonas aeruginosa which acts as the initial colonizer.
INTRODUCTION:
Fahr’s Syndrome is a rare progressive unremitting neurological disorder. It is characterized by abnormal bilaterally symmetric calcium deposits in brain especially basal ganglia. We report first case of Fahr’s syndrome associated with hypoparathyroidism, hypothyroidism and thrombocytopenia.

CASE PRESENTATION:
51-year-old female presented to the ER with an episode of tonic clonic seizures preceded by an aura characterized by spasms of her hands and feet. She denied urinary or stool incontinence, post-ictial confusion, fever and headaches. She also complained of recent weight gain, cold intolerance and memory loss. She was born in Puerto Rico and had a family history of mental retardation. On physical examination, P-88/min, BP-111/76, RR-16/min, T-98 F. Patient was alert but lethargic. There was no thyromegaly, nuchal rigidity or lymphadenopathy. Tendon reflexes showed delayed relaxation phase. Initial labs were significant for Platelets-97 k/μL, Calcium-5.2 mg/dL, Ionized Ca-0.59 mmo/L, Albumin-4.2 mg/dl, Phosphorous-6.7 mg/dl, Mg-1.4 mg/dl, TSH-21.82 uIU/ml, UDS-negative, CT- Head showed symmetric calcification involving the basal ganglia, corona radiata, centrum semiovale, subcortical white matter and cerebellar hemispheres bilaterally. Further investigations revealed PTH-3pg/ml, Vitamin-D – 1,25-OH 21ng/ml, T3 64ng/dl, T4 0.63ng/dl, Urine Ca, cortisol, Ldh, pre and post exercise lactic acid levels, RPR, Iron studies, LFTs were normal. Heavy metal screening was negative. Thyroid and parathyroid sonography was normal.

The patient was diagnosed with Fahr’ syndrome. She received IV and oral calcium. She was discharged on calcium acetate, vitamin D and phenytoin.

DISCUSSION:
Fahr’s syndrome typically manifests in third and fourth decades of life and is associated with various endocrinopathies, primarily hypoparathyroidism. PTH deficiency disrupts bone resorption, phosphaturic effect, renal distal tubular calcium reabsorption and 1,25-dihydroxy vitamin D-mediated dietary calcium absorption leading to hypocalcemia and hyperphosphatemia. This abnormal calcium and phosphorus hemostasis leads to tetany, paresthesias, convulsions and abnormal symmetric calcification in brain, which is one of the core features of Fahr’s disease.

The combination of Fahr’s syndrome with hypoparathyroidism and hypothyroidism is established in rare cases, nevertheless association between the above mentioned three phenomena with thrombocytopenia has not been reported. An autoimmune-related mechanism is one of the likely etiologies. In Fahr’s syndrome, calcification develops within the vessel wall and in the perivascular space, ultimately extending to the neuron. Also free radical production leads to tissue damage and friable intracranial vessels. In patients with Fahr’s syndrome this thrombocytopenia may lead to fatal intracranial hemorrhage. Therefore it is crucial to recognize thrombocytopenia in these patients.
Introduction Reactive eosinophilia can be caused by a wide variety of causes including infections, allergies, connective tissue disorders, autoimmune and inflammatory disorders, malignancy, and medications. Eosinophilia has also been increasingly related to thromboembolism, especially in individuals with other autoimmune or autoinflammatory conditions. However, it remains unclear whether eosinophilia is causal, secondary, or coincidental phenomenon in such events. Case report A 46 year old female presented with chest pain and dyspnea, she had a past medical history of vitiligo, rheumatoid arthritis (RA), diabetes mellitus with features of latent autoimmune diabetes in adults, hypertension, hyperlipidemia, glaucoma, and familial café au lait spots. Home medications include methotrexate and adalimumab. Initial laboratory showed a WBC of 11.3 K/uL with eosinophil increasing from 3% upon presentation to 5% and 4 % afterward, over a period of more than one week. Arterial gas blood test showed pH 7.402, PCO2 40.7, PaO2 143, HCO3 24.8. CT chest with contrast illustrated filling defect in segmental right and left lower lobe pulmonary arteries suggesting pulmonary embolism. Patient was then admitted to critical unit. She was put on heparin drip and subsequently warfarin. She was then discharged after achievement of therapeutic INR levels. Discussion Here, we report a rare case where coincidence of eosinophilia and pulmonary embolism was found in a patient with vitiligo and RA. Since there was no evidence of other apparent causes, it is very likely that the patients’ eosinophilia was secondary to the underlying vitiligo and/or RA flares. The association of eosinophilia with the severity of other autoimmune disease including psoriasis and autoimmune esophagitis has been reported. Eosinophilia that has been related to vasculitis and endothelial damage, may also induce a systemic hypercoagulable state. Toxins released from eosinophils may activate platelets and suppress natural anticoagulation activities, which promotes the development of thromboembolism in arteries and veins. Since a large number of medical conditions can result in eosinophilia, further systemic and large size studies are required to clarify the relationship between eosinophilia and thromboembolism.
Oral contraception: A purported cause in patients with Pseudotumor Cerebri

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Pseudotumor Cerebri or Idiopathic intracranial hypertension describes elevation in the intracranial pressure. In the absence of abnormal cerebrospinal fluid composition, space occupying lesion, or other cause of intracranial hypertension. We report a 20 year old female who presented 9 months after an uncomplicated delivery, with periodic episodes of headache for three weeks. Headache was severe; bi-occipital, radiating anteriorly to bi-temporal and lateral frontal areas, associated with nausea and consistent blurred vision. She was referred to ophthalmology 2 days PTA with worsening blurred vision and new onset sight loss, she was sent to the ER after discovery of papilledema on exam. She had started depot injectable contraception and switched to Oral contraceptive pills CPs 4 months PTA. PMH was unremarkable. On admission blood pressure was 116/97, pulse was 84. Pupils were reactive and EOM were intact, with significant photophobia and light sensitivity. Fundus examination showed bilateral papilledema. Physical exam was otherwise unremarkable. Head CT showed asymmetrical mild prominence of the right optic nerve, otherwise unremarkable for any masses, bleeding or stroke. However, head CT two day prior to admission was unremarkable. Patient also had an MRI, MRA, and MRV of the head all of which were unremarkable. CBC showed leukocytosis. Patient had a lumbar puncture which showed elevated cerebral spinal fluid (CSF) pressures at 43 mm H2O, and normal CSF analysis. Patient improved and was discharged with Acetazolamide for outpatient follow up. OCP was discontinued. 3 weeks after discharge patient remains asymptomatic. To date the natural history of Pseudotumor Cerebri remains poorly understood. Identifying patients at risk is becoming vital especially those at risk of severe permanent vision loss. Use of Oral contraceptives has been formally linked to the disease, and case reports have enforced this association. However the duration of its use that might provoke such an attack as well as patient predisposition remains poorly defined. Our patient has seen significant improvement in the signs and symptoms after discontinuing the pills. Although we still lack extensive data to establish how and when OCP use becomes a risk factor, at the current time it is imperative to stress the importance of thorough neurological and ophthalmological evaluation of females on OCP who present with headache and visual disturbances. Utilizing simple diagnostics as fundoscopy and subsequent Lumbar puncture can detect this dangerous condition and prevent fatal complications.
Propionibacterium Acnes is part of normal flora of the skin and mucosal surfaces. Discovery is only clinically significant in the setting implanted foreign bodies. Positive culture can be true infection or result of contamination. Presentation usually is mild and found to be in the setting of previous surgery and can be delayed given the low virulence of the organism. A 64 year old man was referred to the ID clinic with abnormal operative wound culture results, he has a history of Post Lumbar fusion cauda equina syndrome and urinary retention. He was admitted for a second lumbar fusion 3 weeks ago. A spacer, metallic Rods and screws were utilized in the fusion. He was referred to the ID clinic for abnormal CSF culture results. The patient is asymptomatic, review of systems only positive for urinary retention. Patient reports multiple lumbar fusions, Right Total Hip Replacement, and emergency surgery for Cauda Equina Syndrome. On examination he did not look toxic, was vitally stable, and afebrile. Neurological exam was consistent with cauda equina syndrome. Neck was supple. A surgical scar is seen in the lumbosacral area. No redness, pain or induration. ESR was 55. Cultures of the deep wound fluid grew Propionibacterium on the 11th day of incubation. Patient was treated with a 6 week course of Ceftriaxone, and remains asymptomatic on long term follow up. Symptoms of presentation can include fever, local swelling, effusion, tract formation and purulent discharge. Late presentation can be with pain, stiffness, and prosthesis dysfunction or loosening. Diagnosis of Propionibacterium Acnes infection can be challenging since the organism is slow growing and false negative results can be seen in cases with early termination of incubation. The significance of this case is due the rare occurrence, and more importantly the proper action that is to be taken upon discovery of the organism in patients with implanted devices. In asymptomatic patients, the decision to treat the patient can be a hard one to make, this is because treatment almost always requires removal of the hardware since bio film formation makes eradication extremely difficult without removal of the implant. Although antimicrobial therapy is feasible with the wide range susceptibility of the organism. Therapy is prolonged and not without risks and complications. On the other hand leaving these patients untreated can be invalid, since the organism can in time cause many complications up to prosthesis loosening and malfunction. We aim to present Propionicaberium infection as rare but serious complication that should be addressed seriously, and question if no treatment can be valid.
Toxic Shock Syndrome by Clostridium difficile - A Case Report

Shilpi Singh, Rahaf Almasri, Nisheet Prasad, Abdalla Yousif, Grethchen Bowling

Toxic Shock Syndrome (TSS) has been reported with Clostridium sordellii in Obstetric/Gynecology cases following obstetric procedures with use of misoprostol and mifepristone, but never with Clostridium difficile. Characteristics of TSS with Clostridium sordellii differ from that caused by traditional staphylococcus and streptococcus species, which include pleural and peritoneal effusions, profound leukocytosis, hemoconcentration and notable absence of fever. Characteristics that are similar in both are thrombocytopenia, anemia and laboratory abnormalities reflecting multiorgan failure such as elevated BUN and creatinine, and elevated LFTs. We report a case of 60 year old female who presented to ER complaining of diffuse abdominal pain, nausea, vomiting and dizziness for one day. She had diarrhea for past two weeks that started after taking an antibiotics following a dental procedure. The diarrhea progressed until she became incontinent of stool. She inserted a tampon in her rectum the previous night in an attempt to contain the diarrhea. She did not recall extracting the tampon or passing it in stool. On examination in the emergency department, temperature was 97.4°F, blood pressure 100/65 mm Hg, pulse 105, respiratory rate 18, and oxygen saturation 99% on ambient air. Laboratory data included hemoglobin of 16.1, hematocrit 49.4, WBC 33.4, bands 15, platelets 232, INR 1.4, PTT 36, BUN 57 and creatinine of 3.7. LFTs were normal. CT scan of abdomen/pelvis revealed severe pancolitis, ascites and small pleural effusions. Patient was admitted to ICU. Her condition deteriorated, with rapid rise in LFTs, ammonia levels and worsening kidney function requiring temporary dialysis. She also developed anemia (Hemoglobin 16.1 to 8.9) and thrombocytopenia (Platelets 232 to 62). She persistently remained afebrile and tachycardic and required aggressive volume resuscitation to maintain her blood pressure. She had altered mental status. ABGs showed CO2 retention and subsequently she was intubated. Stool studies came back positive for Clostridium difficile toxins A and B. She was started on intravenous metronidazole and oral vancomycin. Serial KUBs were obtained thereafter which showed improvement in bowel dilatation. She was successfully weaned off ventilator and her condition improved over the period of 10 days with return of baseline normal labs. She was eventually discharged home in stable condition. This is the first reported case of TSS by Clostridium difficile. Our patient exhibited all of the well-documented features of TSS by Clostridium sordellii, in absence of risk factors for Clostridium sordellii and stool test positive for Clostridium difficile toxins. Previous studies have reported TSS by clostridial toxins in the same manner as staphylococcal toxins although in dose-dependent manner. Our patient might have been exposed to higher concentrations of Clostridium difficile toxins due to inadvertent use of rectal tampon. This case may help other clinicians in recognizing similar cases in future.
Zieve’s Syndrome-Revisited

Shilpi Singh, Abdalla Yousif, Nisheet Prasad, Soad Enakuaa, Jenil Gandhi, Zubair Ahmed

Zieve’s syndrome is typically defined by constellation of abdominal pain, jaundice, hemolytic anemia, and hyperlipidemia in alcoholic patients. The main feature of this rare syndrome is acute hemolysis, which differentiates it from acute alcoholic hepatitis. It has been postulated that hyperlipidemia induced red cell membrane instability as the probable cause of hemolysis; but the underlying pathophysiological mechanism is yet to be elucidated. Of note is the fact that not all cases reported so far can fit into the current definition of Zieve’s syndrome. We report a case of 42-year-old male with history of chronic alcohol abuse who was admitted with peripheral edema and worsening jaundice following an episode of binge drinking. Physical examination revealed scleral icterus, anasarca, and multiple vesicular hemorrhagic rash located on both lower extremities. Laboratory data showed elevated LFTs (total/direct bilirubin 14.0/6.6 mg/dL, AST 72 U/L ALT 38U/L) and macrocytic anemia (Hemoglobin 10.1, hematocrit 30.7). Elevated LDH with low haptoglobin consistent with intravascular hemolysis were also noted. Hepatitis panel and GI tumor markers were negative. Lipid panel showed total cholesterol of 65 mg/dL, HDL 20 mg/dL, triglycerides 61 mg/dL, and LDL 33 mg/dL. Abdominal ultrasonography showed cirrhosis of the liver, splenomegaly and a recanalized umbilical vein. Skin biopsy of the hemorrhagic lesions revealed bullous disease significance of which, in relation to Zieve’s syndrome is unclear although one case of Zieve’s syndrome presenting as coagulopathy and subcutaneous hemorrhage has previously been reported. Patient was admitted and received supportive treatment along with diuretics and multivitamins. Patient condition slowly improved over period of two weeks following which he was discharged home. This case is consistent with Zieve’s syndrome with patient’s history of chronic alcohol abuse presenting with acute hemolytic anemia. However, absence of hyperlipidemia in this case goes against the current spectrum of the syndrome. Hemolysis can occur even without hyperlipidemia, as is illustrated in this case. One more case has been reported as atypical Zieve’s Syndrome with normal lipids. First described in 1958 by Dr. Leslie Zieve, only about 200 cases have been reported so far. It is crucial that clinicians are aware of the likelihood that hemolysis can occur even without hyperlipidemia, so that actual cases are not misdiagnosed. It is our impression that this may have contributed to lower incidence of cases worldwide. Balcerzak, et. al. (1968) reported that both native and donor erythrocytes demonstrated hemolysis during the acute syndrome. We postulate that blood transfusions should be avoided as it may worsen the hemolysis. We propose that hemolysis in setting of alcohol abuse alone should suffice the diagnosis of Zieve’s Syndrome, until the exact mechanism of hemolysis can be elucidated.
Is there possible genetic mutations linking the pathogenesis/susceptibility of Depression and Takotsubo Cardiomyopathy?

Daniel Torino, Dr. Shuvendu Sen

Introduction: Takotsubo Cardiomyopathy is an increasingly reported syndrome generally characterized by transient systolic dysfunction of the apical and/or mid segments of the left ventricle that mimics myocardial infarction (MI), but in the absence of obstructive coronary artery disease [1-15]. Described below is a case of typical variant Takotsubo Cardiomyopathy presenting as NSTEMI with positive troponins. Case: 53-year-old caucasian female with a past medical history significant for hypertension, Major Depressive Disorder, diabetes, presents to the emergency department complaining of retrosternal chest pain. Prior to admission, the patient was denied disability and became very upset. The chest pain was described as midline pressure, 8 out of 10 severity, with radiation to both sides of the upper chest, worsened on exertion. Patient admitted palpitations, diaphoresis, emotionally disturbed. ROS was otherwise unremarkable. Troponin was 0.97, 0.31, 0.39. CK was 151, 112, 96. CKMB was 7.07, 5.06, 4.10. EKG showed normal sinus rhythm. Echocardiogram demonstrated mildly dilated left ventricle without concentric LVH and significant hypokinesis of mid IV septum and midventricular segment with extension to the apex. Discussion: Takotsubo Cardiomyopathy is a newly comprehended phenomenon that may further be grasped by analyzing its pathogenicity and possible genetic factors increasing susceptibility. The search for candidate genes has been the subject of considerable study, much of which focus upon neurotransmitter systems influenced by the medications used in the clinical management of this disorder. Linkage studies have suggested a role for the tryptophan hydroxylase 2 (TPH2) gene [19]. In addition, genes involved in the pathogenesis of bipolar disorder may have pleiotropic effects and confer risk for other types of psychopathology [18]. As an example, a meta-analysis of genome-wide association studies (33,332 patients with bipolar disorder, unipolar major depression, schizophrenia, autism spectrum disorders, or attention deficit-hyperactivity disorder, and 27,888 controls) identified three single-nucleotide polymorphisms on chromosomes 3 and 10 that were associated with all five disorders [22]. Many of the neurotransmitters involved in the pathogenesis of depression have been implicated separately in Takotsubo Cardiomyopathy, namely serotonin and dopamine. These concepts bring to light a novel proposition involving the possible converging pathogenesis between depression and Takotsubo Cardiomyopathy, consisting of a mutation in the gene encoding for the enzyme in the second synthetic step of both Dopamine and Serotonin that is additionally involved in the synthetic pathway of Norepinephrine and Epinephrine. These catecholamines share the aromatic L-amino acid decarboxylase (AADC) enzyme, also known as 5-hydroxytryptophan decarboxylase or dopa decarboxylase. Investigation into individuals with specific genetic profiles could potentially prove beneficial in identifying susceptibility to the above mentioned heart disease or even previously established cardiac disease since analogous permanent (rather than transient) apical outpouchings develop in patients with hypertrophic cardiomyopathy and mid-ventricular obstruction.
Raritan Bay Medical Center

Enteric Fistulous communication with Aortoiliac graft fistula permitting Eggerthella Lenta Colonization

Daniel Torino, Dr. Shuvendu Sen

Introduction: Anaerobic, non-sporulating, gram-positive bacilli from the normal human gut flora can occasionally be implicated in invasive diseases. Notably, Eggerthella lenta, formerly known as Eubacterium lentum, often causes clinically significant bacteremia (5). Here we report a novel case of Eggerthella colonization. Case: Patient is 77-year-old male with a past medical history significant for insulin resistant diabetes mellitus, abdominal aortic aneurysm status post iliac graft repair two years ago, hypertension, congestive heart failure who presented to the hospital complaining of dizziness and lethargy. The patient reported recording a temperature of 101. He denied cough, chest pain, palpitation, nausea, vomiting or constipation. The patient admitted fever, chills, dysuria, polyuria, burning sensation during micturition for two days. Physical examination revealed periumbilical tenderness with deep palpation without peritoneal signs. Laboratory studies confirmed Eggerthella lenta septicemia and E.coli ESBL positive urinary tract infection. Subsequent abdominal CT with contrast demonstrated an aortoiliac bypass graft with stranding around the aorta and near the iliac components with presence of an air bubble between these components. Discussion: Eggerthella is a bacterial genus of Actinobacteria, in the family Coriobacteriaceae. Members of this genus are found in the human colon and feces and have been isolated from various clinical specimens including blood, abscesses, wounds, obstetric and genitourinary tract infections, and intra-abdominal infections [3, 4]. As PCR and sequencing techniques are becoming more readily available in clinical laboratories, 16S rRNA gene analysis will be a more reliable and practical approach to identify Eggerthella to the species level [6]. All Eggerthella species including E. lenta, E. hongkongensis, and E. sinensis have been associated with bacteremias of relatively high mortality [5, 6]. Eggerthella lenta identification in blood culture should be considered serious and warrants evaluation of a source that can include skin and soft tissues, obstetric-genitourinary tract, and intra-abdominal infections. Based on agar dilution and E-test methods susceptibility studies, in general, E. lenta are susceptible to amoxicillin-clavulanic acid (MIC &#8804; 2/1), metronidazole (MIC &#8804; 2), and clindamycin (MIC &#8804; 0.5) [4, 8, 11]. With its known anaerobic activity, moxifloxacin also demonstrated good activity against E. lenta (MIC &#8804; 1) [11]. Although ertapenem susceptibilities were not found in the literature, imipenem demonstrated good activity (MIC &#8804; 0.5) [11]. Surprisingly, the MIC for penicillin (&#8804; 1) is in the intermediate breakpoint, and one recent report noted resistance to piperacillin-tazobactam (MIC = 32) [4]; hence clinicians should refrain from the use of penicillin. E. lenta also appears to be uniformly resistant to cefotaxime (MIC > 256) [4]. This case illustrates a unique occurrence in a patient with many comorbidities leading to E. lenta disseminated infection. Further studies should be performed to improve our knowledge of the epidemiology and pathogenicity of these slow growing anaerobic bacteria.
Possible role of Hydrocortisone in late stage of septic shock

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Traditional management of septic shock included vasopressor and intravenous fluid to maintain blood pressure so that organs could get enough blood perfusion. Addition of corticosteroid is also suggested at an early critical stage if the blood pressure still cannot be maintained at MAP > 65 or SBP>90 according to The Surviving Sepsis Campaign Guidelines 2012. Here we report a case, in which hydrocortisone was added in the later stage of septic shock dependent on vasopressors. Case: A 36 year-old female nursing home resident with past medical history of ventilator dependent respiratory failure, congenital mental retardation and seizure, ,was admitted to the hospital with chief complaint of high fever (Tmax =103.4). Patient’s vital signs included BP 95/59, HR 98, RR 18. T 102.9 °F. SaO2 98% at 50% oxygen. The lungs were clear to auscultation and heart rhythms were regular with normal S1 and S2. Patient had increased abdominal sound, and diarrhea. Hb 8.7, Hct 25.6, WBC 37.6, Platelet 224,000, BUN is 44, Creatinine 0.7, Sodium 138, Potassium 3.4, Magnesium 2.1, TSH 1.9. Initially patient was admitted to telemetry ward. Later patient’s blood pressure dropped to 80/42, and was then transferred to the critical care unit. Patient was started with Dopamine intravenously to maintain the blood pressure after fluid challenge with normal saline. Patient continued her other treatment. Patient was also started with broad antibiotics. Later after pneumonia (pseudomonas aeruginosa) and C-difficile colitis were diagnosed, patient was started with Amikacin and Metronidazole intravenously. Over two weeks’ treatment, patient’s temperature and WBC became within normal range and diarrhea stopped, but once Dopamine were titrated down, patient’s systolic blood pressure dropped below 90 with corresponding sinus tachycardia. Finally after hydrocortisone was started, patient’s blood pressure gradually improved to115/71 over next five days and Dopamine was successfully stopped. Patient subsequently was downgraded to telemetry. Discussion: In acute phase of septic shock, hydrocortisone was suggested when vasopressor and fluid are not enough to maintain normal blood pressure, but there has been no suggestion about the use and efficacy of hydrocortisone on the later stages of septic shock based on The Surviving Sepsis Campaign Guidelines 2012. We postulate that our patient most likely developed relative adrenal insufficiency during septic shock and that hydrocortisone helped maintain blood pressure to wean off the vasopressors by increasing the sensitivity of resistance vessels to catecholamines. While more strict studies are needed to validate our postulation, our case study strongly raises the awareness of the possibility of more extended and comprehensive use of hydrocortisone during septic shock.
Case Report: Uterine fibroid causing an extensive left leg venous thrombosis

Nicole Abel, Krysta Contino and Elizabeth Cerceo

Introduction: Uterine fibroids, or leiomyomatas, are a common benign tumor in women, occurring most frequently during the fifth decade of life. Because of their slow growth rate, uterine fibroids rarely cause acute complications, including venous thromboembolisms (VTE), which are generally more common in hypercoagulable states, such as pregnancy or gynecological malignancy(3). When VTEs present in the setting of a leiomyomata, they usually form secondary to pelvic vein compression(1). Below is the case of a patient who presented with extensive lower extremity VTE in the setting of uterine fibroids.

Case Report: A 44-year old African American female with a history of uterine fibroids, iron deficiency anemia, and nicotine dependence presented to the hospital with an acute onset of left leg/buttock and lower back pain for five days duration. On physical exam, she had a large, contoured abdominal mass and marked edema of the left leg with associated erythema and warmth. An ultrasound of the left lower extremity discovered an occlusive thrombus in the greater saphenous vein, superficial femoral vein, and popliteal veins, with a non-occlusive thrombus in the common femoral vein, deep femoral vein and upper superficial femoral veins, with concurrent thrombus in the posterior tibial and peroneal veins. A CT of the abdomen/pelvis showed an enlarged uterus measuring 16 cm by 8 cm above the umbilicus. Hypercoagulable work-up was negative. Endometrial biopsy was negative for malignancy. She was started on warfarin therapy and underwent a total abdominal hysterectomy for removal of uterine fibroid. Conclusion: Virchow’s triad consists of hypercoagulability, endothelial injury and stasis of blood flow. It is not uncommon for women to present with VTEs in the setting of a hypercoagulable state or with structural abnormalities, such as May-Thurner syndrome. However there are few case reports demonstrating an association between DVTs and uterine leiomyomatas. In one report by Stanko, et al, a 49-year old woman with history of large uterine leiomyomata presented with acute left lower extremity edema and was found to have thrombosis of the left iliofemoral vein(4). Ippolito, et al cited the case of a 51-year old woman with large fibroids who presented with syncope and dyspnea. She was found to have pulmonary embolism along with extensive lower extremity VTE(2). Although there was no clear cause of endothelial injury in the woman we presented above, venous stasis resulting from large uterine fibroids was the most likely pathophysiologic diagnosis. Leiomyomata are common in middle-aged women but have not heretofore been recognized as a risk factor for DVTs. Future areas of research including studies evaluating the utility of screening women with large uterine fibroids for DVTs and clarifying the role for VTE prophylaxis in such individuals may be warranted.
WHAT DO SHOPPING CARTS AND FOOTBALL HAVE IN COMMON?

Namrata Baxi, Megan Blessinger, Nikunjkumar Patel, Anuradha Mookerjee

Testicular cancer commonly presents between 20-39 years of age and incidence is rapidly increasing. Trauma is a well documented association of testicular cancer. The question remains whether trauma is a causative factor in the development of the tumor or is it a trigger for diagnosis. There are conflicting studies regarding the association of trauma and the role it plays in the development of testicular cancer. A 39 year old male presented to the clinic with low back and right testicular swelling and pain associated with right lumbar area paraspinal pain, fevers and night sweats. One week prior to presentation, patient sustained a direct trauma to his testicles by a shopping cart. Patient reported trauma to the genital area six months prior to presentation while playing recreational football. After this incident he noted mild swelling, redness and pain, which resolved over time. Patient denied penile discharge and history of epididymitis, dysuria, pyuria, and weight loss. Physical exam revealed a right testicle measuring at 8cm, surrounded by erythematous and warm overlying skin, and tender right inguinal lymphadenopathy. Ultrasound revealed a large heterogeneous mass in the right testis with extensive right testicular microlithiasis. CT scan revealed right-sided enlarged necrotic retroperitoneal lymphadenopathy (pelvis and iliac chain- measuring at 4.6 x 3 cm). AFP was 1.3 and a beta HCG is elevated at 36 (normal <5). Patient underwent a right radical orchiectomy. The fevers and night sweats were attributed to the tumor and discharged on a short course of antibiotics. Pathology was consistent with 11 x 9 x 6 cm seminoma classic type with necrosis and fibrosis with lympho-vascular invasion. Patient was subsequently diagnosed with Stage III seminoma and has completed 3 cycles of bleomycin, etoposide, and cisplatin, with no complications. The rapidly enlarging tumor within one week of trauma illustrates an interplay between trauma and testicular cancer. Early diagnosis of testicular cancer is crucial since the doubling time of testis tumor is estimated to be 10 to 30 days. Even though evidence based guidelines recommend against screening for routine testicular cancer in age appropriate males; it is important to have testicular tumors in the differential for young males presenting with acute onset of scrotal pain and swelling. Commonly, tumors have been misdiagnosed and treated ineffectively with antibiotics, resulting in the time frame of a correct diagnosis to range from 4 months to 5 years. Studies have persistently failed to identify trauma as an important risk factor for testicular cancer due to recall bias. Therefore, additional studies are needed to better elucidate the role of trauma in testicular cancer.
A Cold Case of Recurrent Leg Edema after a Successful Treatment of Hepatitis C Liver Disease

Prianka Bhattacharya, Satyajeet Roy, MD, FACP

Introduction: Hepatitis C infection occurs worldwide and can manifest with multi-system complications. Clinical manifestations may include nonspecific constitutional symptoms, hepatic manifestations (e.g. chronic active hepatitis, cirrhosis, fulminant liver failure), or extrahepatic complications (e.g. vasculitis).

Case Presentation: A 51 year old male presented with recurrent bilateral lower extremity edema for 6 months. Workup and treatment for common causes was undertaken without revelation or resolution. His past medical history was significant for chronic active hepatitis secondary to Hepatitis C virus (HCV), genotype 3a. Two years ago the patient used to have persistent leg edema which resolved after successful treatment of HCV with peg-interferon and ribavirin. Patient attained a sustained virologic response (SVR), plus normalization of elevated serum aminotransferases and cryoglobulins. We measured serum cryoglobulin level with the present episode of leg edema. It was significantly elevated. His serum HCV RNA and aminotransferases were also elevated. Patient subsequently underwent treatment for recurrent HCV hepatitis with peg-interferon and ribavirin, which resulted into undetectable HCV RNA, normalization of cryoglobulin level, and complete resolution of leg edema.

Discussion: Patients with HCV infection are at risk for mixed cryoglobulinemia. After SVR recurrence of HCV and mixed cryoglobulinemia are rare. A study identified 8 cases of relapsed infection in patients after they had achieved SVR. This patient’s unexpected relapse was evidenced by an uncommon resulting complication—mixed cryoglobulinemia. More than 50% of patients with HCV have cryoglobulins, although only 10% to 15% tend to develop clinical disease. Cryoglobulins are cold-precipitating immunoglobulins that typically remain soluble at body temperature but may precipitate in peripheral vessels, manifesting as palpable purpura or edema, usually in the lower extremities. There are 3 types: type I (monoclonal), type II (mixed monoclonal), and type III (polyclonal), with the latter two being associated with HCV. Other associated disorders include autoimmune diseases and hematologic malignancies. Cryoglobulinemic vasculitis associated with HCV can also cause glomerulonephritis, neuropathy, arthritis, and pulmonary inflammation. Treatment of mixed cryoglobulinemia may entail immunologic therapy, but foremost is treating the underlying cause. Eradication of HCV is the only treatment for mixed cryoglobulinemia and leg edema like in our patient. Conclusion: Our patient presented with a rare recurrence of mixed cryoglobulinemia and HCV after a previously successful eradication of HCV, presenting indolently with persistent lower extremity edema. The edema resolved only after resolution of cryoglobulinemia and HCV infection after further anti-HCV therapy. Presence of persistent leg edema in patients with history of successfully treated HCV infection may indicate presence of cryoglobulinemia. In turn, presence of cryoglobulinemia may indicate recurrence of HCV infection, treatment of which results in resolution of leg edema.
Hypereosinophilia as initial presentation leading to diagnosis of HIV.

Prianka Bhattacharya, Brian Gable, MD

Introduction: Hypereosinophilia is defined as a peripheral eosinophil count of 1500/cc or higher. Eosinophilia can be seen with a number of causes, including parasitic (helminthic) infections, allergic or drug reactions, malignancies, and vasculitis. Manifestations of hypereosinophilia can arise in multiple systems. Examples of associated conditions include pulmonary (i.e. Churg-Strauss), gastrointestinal (i.e. eosinophilic esophagitis), skin (i.e. eosinophilic cellulitis), and neurologic diseases (i.e. eosinophilic meningitis).

Case presentation: A 41 year old African American female presented with intractable nausea, vomiting, abdominal pain for four months, and diffuse pruritus for nine months. She later developed blurred vision and severe bilateral lower extremity pain and paresthesias. CBC showed eosinophilia, with absolute eosinophil count (AEC) persistently greater than 3000. Tissue biopsies (skin, foregut) showed significant eosinophilic infiltration. Extensive additional workup undertaken to rule out secondary causes of eosinophilia was unrevealing, including initial negative HIV testing. In evaluation of clonal eosinophilia, FIPL1-PGFGRA/B mutation testing was negative. Bone marrow biopsy revealed essential absence of CD4+ T-cells and markedly reduced CD4:CD8 ratio. This prompted repeat testing for HIV, resulting positive with HIV viral load greater than 500,000 and absolute CD4 count of 6. Upon treatment of HIV infection with HAART, patient''s CD4 count, HIV viral load, eosinophilia, and symptoms all improved. Discussion: The algorithm for evaluation of elevated blood eosinophilia starts with evaluation for reactive eosinophilia (i.e. parasitic infection, allergic disease, drug reaction). Secondary workup includes evaluation for primary clonal eosinophilia (i.e. FIPL1-PGFGRA mutation, leukemia with eosinophilia), and other diseases associated with eosinophilia as mentioned. If through thorough workup secondary causes and clonal eosinophilia are excluded, a diagnosis of idiopathic hypereosinophilia is considered. This patient''s presentation with symptomatic moderate eosinophilia, initially without obvious cause and with evidence of end-organ damage (i.e. gastrointestinal, neurologic), signaled concern for hypereosinophilic syndrome. This is a subset of idiopathic hypereosinophilia with evidence of potentially devastating resulting organ damage (i.e. cardiac infiltration with endocarditis), and therefore necessary to recognize when present. With the ultimate finding of HIV positivity on retesting, and improvement in patient''s eosinophilia with treatment of HIV, it appears her hypereosinophilia was related to HIV infection. HIV with eosinophilia may clinically manifest with organ damage from eosinophilic infiltration, with examples of intractable pruritus, eosinophilic folliculitis, eosinophilic gastroenteritis, and nerve palsies, all noted in sporadic case studies. Conclusion: A presentation of isolated hypereosinophilia of unknown cause requires diligent pursuit in diagnosis, as resultant manifestations can range to life-threatening. While this is a rare presentation of a common epidemic disease (HIV), awareness and further study is necessary given possible severe complications if either is left untreated.
A broken heart that resembled an octopus pot

Effat Jabeen, Satyajeet Roy, MD, FACP

Introduction: Broken heart syndrome, apical ballooning syndrome, stress cardiomyopathy are some of the names of a reversible cardiac syndrome that is brought on by physical or emotional stress, such as trauma, stressful medical condition, or death of a loved one. It is mostly seen in postmenopausal women. Case Presentation: A 66 years old Caucasian female with history of hypertension underwent an elective small bowel surgery under general anesthesia. She had a good performance status and a normal cardiac evaluation. Her left ventricular ejection fraction was 55-60% prior to the surgery. Her postoperative course was significant for tachyarrhythmias, shortness of breath, worsening pressure like sensation in her chest, and wheezing. Physical exam revealed elevated jugular venous pressure, left ventricular S3, and bilateral ankle edema. Her chest x-ray revealed congestive heart failure. An echocardiogram showed severe left ventricular systolic dysfunction, systolic apical ballooning, akinesis with basal hypokinesis that resembled a Japanese octopus trap (locally known as "Takotsubo"), and a left ventricular systolic function of 25-30%. Her pro-BNP was 31,370 pg/ml. She was diagnosed with Takotsubo Cardiomyopathy. She was treated with furosemide, lisinopril, and oxygen via mechanical ventilator support. Cardiac catheterization showed normal coronary arteries. She recovered fully and required medical therapy for 2 weeks. A repeat echocardiogram at 3 months showed a normal size left ventricle, normal wall thickness, and left ventricular systolic function of 55-60%. She remained asymptomatic at 1 month follow up. Discussion: Takotsubo Cardiomyopathy presents very much like acute coronary syndrome. Patients typically present with acute onset chest pain, elevated troponin, and ST elevation on EKG. However, cardiac catheterization does not reveal any coronary artery disease or obstruction. About 1.7-2.2% patients who present with suspected acute coronary syndrome are diagnosed with Takotsubo Cardiomyopathy. Takotsubo is a Japanese word that translates to “octopus pot”; resembling the shape of left ventricle during systole on imaging in stress cardiomyopathy. Hypokinesis or akinesia of the apex of heart with preserved function of the base causes bulging of apex during systole. Most recent diagnostic criteria, proposed by researchers at Mayo Clinic, include 1) hypokinesis, akinesis, or dyskinesis of left ventricle extending beyond a single epicardial vascular distribution, 2) absence of obstructive coronary disease, 3) new EKG abnormalities or elevated troponin, and 4) absence of pheochromocytoma and myocarditis. Recognition of Takotsubo cardiomyopathy is important because these patients’ hearts generally return back to their baseline with prompt supportive treatment. Since recovery is usually spontaneous, patients do not require long term medical therapy. Conclusion: Our patient had a post surgical stress induced Takotsubo Cardiomyopathy that was promptly diagnosed and managed, resulting in an excellent outcome. Takotsubo Cardiomyopathy should be suspected in all patients who present with suspected acute coronary syndrome.
Incidentalomas of the gastrointestinal tract - A case report of appendiceal mucocele resected by laparoscopic right hemicolecction

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Introduction: Appendiceal tumors are a rare but well-known clinical entity. These are most commonly identified incidentally as a “mucocele”, either during screening studies in asymptomatic individuals, or during the workup of unrelated non-specific abdominal complaints. These constitute only 0.2 to 0.3 % of all resected pathology specimens. Case Presentation: A 52 year-old woman was found to have an enlarged appendix with a wide base that prolapsed into the cecum during a routine colonoscopy. CT-abdomen revealed a 3.0 x 1.2 cm uniform hypodense lesion involving the appendix suggestive of a “mucocele”. As the lesion involved the base of the appendix and prolapsed into the cecum, a decision was made to perform laparoscopic hemicolecction for resection. Histopathology revealed a mucinous cystadenoma of the appendix with no evidence of dysplasia. Further, a detailed intra-operative exploration of the gastrointestinal tract, ovaries and peritoneum for coexisting colorectal and ovarian malignancies was unremarkable. Post-operative course was uneventful and the patient recovered well. Follow-up exams at 6, 12, and 24 months were unremarkable, and further confirmed that the patient was doing well. Discussion: The earliest accounts in the literature come from Rokitansky in 1842, in which he described a “mucocele” as a cystic mass with a dilated appendiceal lumen due to abnormal mucus accumulation. While this traditional definition still holds true; much headway has been made in understanding the underlying etiologies and the diagnostic and the management approach has evolved considerably since that time. Although a presumptive pre-operative diagnosis can be made with appropriate imaging studies, a definitive diagnosis is principally made after histology, which correlates strongly to the course and prognosis. Given that lesions that seem benign on imaging could harbor changes of a cystadenocarcinoma, the treatment of choice is surgical resection. However, no consensus guidelines still exist about the surgical approach. While appendiceal resection by laparotomy remains the conservative and favored approach; with technological advances, laparoscopic resections have become safer and offer another feasible option. Given the risk of spontaneous or accidental rupture of a “mucocele” with progression to “pseudomyxoma peritonei”, a condition that is morbid and recurrent, use of laparoscopy still remains controversial. Patients with simple or benign neoplastic mucocele have an excellent postoperative prognosis, with 5-year survival rates of 91%–100%. Conclusion: We report a case of a low-grade appendiceal mucinous neoplasm that was successfully treated by laparoscopic hemicolecction. Due to significantly high association of coexistent colorectal and pelvic malignancies, patient with incidental “mucocele” of appendix should be periodically screened for colorectal cancer, and gynecological malignancies.
Are Internal Medicine Residents Still Burning Out?

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Introduction: The Accreditation Council for Graduate Medical Education (ACGME) work-hour restrictions for the residents were meant to keep residents from experiencing burnout. The prevalence of resident burnout in our setting is unknown. It is also unclear whether there is a change in the prevalence of burnout with advancement through three years of residency. Methods: Sixty residents of the Internal Medicine program were asked to complete a burnout assessment survey form (Maslach Burnout Inventory: MBI). The collected data was analyzed. Pearson correlation was used to correlate the emotional exhaustion score, and depersonalization score. Pearson Chi Square test was used to compare the frequency of burnout among our Internal Medicine residents to the available literature. Results: Thirty-nine (65%) residents completed the survey forms. Demographics showed 61.5% males, 38.5% females; age group: 5% in 25-29, 54% in 30-34, and 41% were greater than 35 years of age. First, second, and third year residents were 44%, 25%, and 31% respectively; 56% married; 97% international medical graduates. Ninety percent had family support, 87% had close friends, 51% had close friends within their residency program, and 66% were comfortable with their plans after residency. Ninety five percent of the participants had scores indicative of burnout. All experiencing depersonalization, while only 10.8% of those who experienced depersonalization also experienced emotional exhaustion. There was no significant difference in burnout prevalence between first, second, and third year residents. Prevalence of burnout in our first year residents was 100%, compared to 34% in another study. (1) A systemic review revealed that the overall prevalence of resident burnout is 27%–75%, depending on specialty. (2) Discussion: In 2003, the ACGME announced that residents were no longer allowed to work greater than 32 hours consecutively and more than 80 hours per week. (3) Despite the regulations placed on duty hours, burnout continues to be a significant problem amongst Internal Medicine residents. Our data reveals that 95% of the Internal Medicine residents at Cooper University Hospital met criteria for burnout even in the setting of strict work hour regulations and good family as well as social support. Such a high rate of burnout suggests that the number of hours spent in training and patient care may not be the sole cause of burnout. In fact, this data warrants questioning whether duty hour regulations are leading to some additional stressors for individual residents as patient care is handed off more frequently and medical education is potentially compromised. Conclusion: The prevalence of burnout among Internal Medicine residents is high even with implementation of duty hour regulations. Despite good family and friend support, and security with the future plans, high prevalence of burnout suggests additional undefined causative factors that need to be explored and addressed.
A Retrospective study assessing the impact of supplemental calcium in critically ill trauma patients receiving total parenteral nutrition.

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Background: Total parenteral nutrition (TPN) is an integral part of the patient care, particularly in the intensive care unit. Recently, there have been national drug shortages including many basic additives for TPN such as calcium, phosphate and multivitamins. After receiving Internal Revenue Board’s approval, we initiated a Quality Improvement Project (QIP) in the Trauma Intensive Care Unit (TICU) to assess whether the absence of calcium in TPN has any clinically significant impact. Method: A retrospective chart review of all TICU patients who received TPN from March 2011 to December 2011 was performed. Exclusion criteria included malabsorption, sepsis, parathyroid disorders and patients requiring renal-replacement-therapy. We compared the length of stay in the hospital, duration of ventilator assistance and mortality among the patients who received TPN with and without supplemental calcium. Results: A total of 34 patients received TPN in the TICU during the selected time period who fulfilled our criteria. Mean age of the patients was 45 years and 42% of them were females. 19 patients received TPN with calcium-glucuronate and 15 patients received TPN without any supplemental calcium. 2 patients in the calcium group and 5 patients in the non-calcium group died during the hospital admission. This mortality data didn’t reach any statistical significance. We did not find any significant difference in the length of hospital stay (p=0.888) or the duration of ventilator assistance (p=0.811). Conclusion: Theoretically, the absence of calcium in the TPN may be well compensated by the large stores of calcium in the human body in the form of bone. But the current study indicated that there is a trend towards increased mortality for those patients who didn’t receive supplemental calcium in their TPN compared to those who did. This didn’t reach statistical significance, but this pilot study was small and not adequately powered to assess for this. It is unclear why there is a difference in mortality in the non-calcium group. But some considerations include increased cardiac arrhythmogenicity due to hypocalcemia. As a result, we are now expanding the study to include all surgical, medical and coronary care ICU patients. Hopefully, this ongoing study will help us determine if the absence of Calcium in the TPN has a clinically adverse impact.
ST Segment Elevation Myocardial Infarction Following Assault

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Aims: To describe a case of ST segment elevation myocardial infarction following blunt trauma to the chest wall, including diagnosis and management of such a presentation. Background: Early recognition of myocardial ischemia and infarction as a cause of post-traumatic chest pain is often confounded by the clinical history and patient characteristics. However, early diagnosis and institution of appropriate therapy can improve outcomes given these same factors. It is important for physicians to be aware of and to entertain a high suspicion for this disease entity. Case: A 38 year old male with a past medical history of hypertension, tobacco abuse and human immunodeficiency virus suffered blunt chest wall trauma during assault. He presented to an emergency department for evaluation of subsequent severe chest pain. Initial work-up included computed tomography imaging of the chest, abdomen and pelvis. This revealed fractures in the L2 and L3 transverse processes. A routine electrocardiogram (EKG) was subsequently performed and the patient was found to have ST segment elevations in precordial leads V1 and V2 with associated reciprocal changes. Biochemical testing revealed elevation of cardiac troponin levels. Myocardial infarction was diagnosed based upon these findings and the patient was referred to our institution for emergent percutaneous coronary intervention. Coronary angiography revealed acute thrombosis, occluding 100% of the proximal left anterior descending artery (LAD). Manual thrombectomy was performed, followed by placement of a drug eluting stent. Subsequent to this intervention, there was complete resolution of chest pain and ST segment elevations. Discussion: Ischemic coronary artery pathology must be considered in the setting of blunt chest wall trauma in all patients, regardless of age or risk factors. ST segment elevation myocardial infarction following blunt chest wall trauma occurs most frequently following injury to the LAD. Compression of this vessel against the unyielding chest wall makes it vulnerable to shearing forces. Intimal injury can predispose to arterial dissection, or serve as a nidus for activation of the coagulation cascade and lead to acute thrombosis. Additionally, vascular injury may incite a spasmodic event leading to downstream myocardial damage. Appropriate management of a patient following blunt chest wall trauma includes a screening EKG in all patients presenting with chest pain or whose history is suggestive of heart disease. Serial cardiac enzymes may be indicated if there are abnormal findings on EKG, if chest pain is typical for ischemic etiology, or there is a history of coronary artery disease. Treatment is based upon findings, but includes urgent revascularization if there is evidence of ischemic pathology. Prompt diagnosis as well as early and appropriate treatment can minimize cardiac damage, limit morbidity and be life saving.
**Infective Endocarditis: Duke Doesn’t Always Have Your Back**

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**Introduction:** Infective endocarditis seems straightforward to diagnose when it fits the Modified Duke’s Criteria, but in some cases, it does not. Case Presentation: An 83-year-old man with a past medical history of atrial fibrillation and aortic valve replacement (bioprosthetic) in 2011, presented with two days of progressively worsening abdominal pain with radiation to the lower back. He described the pain as severe and diffuse. He stated he had not had a bowel movement in 4 days, but was passing gas. He denied fevers, nausea, and vomiting. On exam, his abdomen was distended and tympanic with decreased bowel sounds. Laboratory evaluation was unremarkable except for an elevated BNP. Chest radiograph showed bilateral infiltrates and a CT scan of the chest and abdomen was normal except for a right pleural effusion. Treatment for bacterial pneumonia was initiated with ceftriaxone and azithromycin. Blood cultures x 2 bottles were positive for Enterococcus faecalis that was susceptible to ampicillin and gentamicin. Transesophageal echocardiography did not demonstrate vegetations, valvular dysfunction, or myocardial abscess. His antimicrobials were changed to IV ampicillin and gentamicin. His abdominal pain resolved with a bowel regimen, but his back pain continued. Blood cultures remained positive for a week despite appropriate antimicrobial therapy. He remained afebrile and hemodynamically stable with an unchanged physical exam. Lumbar spine MRI with contrast showed no evidence of osteomyelitis, discitis, or abscess. Thoracentesis revealed transudative effusion with negative cultures. After 3 consecutive sets of negative blood cultures, the patient was discharged to a rehabilitation center on hospital day 17 with a diagnosis of aortic prosthetic valve enterococcal endocarditis. His back pain was minimally relieved with Percocet. He completed a 6-week course of ampicillin and gentamicin. Discussion: This patient was classified as possible endocarditis using the Modified Duke's Criteria; meeting only one major and one minor criterion. A clinical diagnosis of infective endocarditis was made due to persistently positive blood cultures with enterococcus in the setting of a bioprosthetic valve. In searching for a source of infection, we were reminded that infective endocarditis can be masked by vague or nonspecific symptoms such as acute arthritis, back pain, arthralgias or myalgias, and may not always meet the criteria for definitive diagnosis. In some cases, these complaints can be the first symptoms to appear in infective endocarditis.
Legionnaires’ secret operation: unexpected complication of Legionella infection

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A 51 year old man with hypertension, diabetes and history of tobacco use presented with fatigue, myalgia, fever and chills of one week duration. He also reported decreased appetite, weight loss (8 lbs. in three weeks) and sweats. Four months prior he had been prescribed five-day course of azithromycin for presumed community acquired pneumonia, but completed only the first day of treatment. His medications included metformin, triamterene/hydrochlorothiazide, amlodipine, lisinopril, metoprolol, hydroxyzine, ranitidine, and aspirin. He denied herbal supplements, over the counter medications, or NSAIDs. He had no history of kidney disease. He worked in a car sales office at a desk positioned below the cooling vent. Objectively, patient was febrile (101.4F) and normotensive, with right sided basilar rales on lung auscultation. Chest X-ray showed linear opacities in the posterior right lower lobe.

Laboratory analyses were consistent with acute renal failure (ARF): serum creatinine 4.2mg/dL (baseline 0.8mg/dL), blood urea nitrogen 43mg/dL, mixed anion gap metabolic acidosis, respiratory and metabolic alkalosis, and hyperphosphatemia. He was normoglycemic, without ketosis. There was no leukocytosis or eosinophilia. Urine was dark yellow, specific gravity of 1.009, pH 5.5, without signs of infection. Urine sediment microscopy demonstrated hematuria with dysmorphic red blood cells (RBC), pyuria and RBC casts. Random urine protein/creatinine ratio was 1.07, FENa 4.4% and FEurea 31%. C3 and C4 levels were normal, with ANCA negative. Renal ultrasound suggested medical renal disease with normal sized kidneys. Patient was started on intravenous fluids and levofloxacin but the renal function further deteriorated (creatinine 6.4mg/dL), despite adequate urine output. Given the presentation suggestive of rapidly progressive glomerulonephritis (RPGN), patient was started on pulse intravenous methylprednisolone therapy. However, the urine Legionella antigen returned positive, thus regimen was changed to high dose intravenous steroids (methylprednisolone 1mg/kg/day for 2 days), with immediate improvement in creatinine (5.9mg/dL) and gradual return nearly to baseline (1.1mg/dL) over the following three weeks. The patient never underwent renal biopsy, given the clinical response to steroids. ARF is an uncommon complication of Legionella infection, usually caused by acute tubular necrosis. However, our patient most likely had tubule-interstitial nephritis (TIN) or crescentic glomerulonephritis (CGN), given his sterile pyuria and hematuria with RBC casts. While cases of oliguric TIN with rapidly increasing creatinine have been reported, non-oliguric, yet rapidly progressive course in this patient raises the possibility of CGN. To our knowledge, only two cases of RPGN with CGN accompanying Legionellosis have been published. One of those was treated with pulse methylprednisolone; both evolved into end-stage renal disease. In conclusion, even though ARF from TIN or CGN is an extremely infrequent complication of Legionellosis, it should still be in the differential in patients with ARF and pneumonia, as prompt high-dose steroid treatment may significantly improve prognosis.
Acute appendicitis in the setting of infectious mononucleosis

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Acute appendicitis is a rare complication of infectious mononucleosis (IM). We present a 35-year-old male who presented with intermittent fevers, malaise, diffuse mild headache, and sore throat for 12 days. His physical examination was negative for axillary, supraclavicular or inguinal lymphadenopathy, and abdomen was soft, non-tender without rigidity, guarding or rebound tenderness with no hepatomegaly or splenomegaly. Lab at admission was significant for WBC count of 14.4 x10³/µL; the manual differential revealed 15% atypical lymphocytes, along with ALT 420, AST 295, and Alk Phos 346. Blood work was positive for heterophile antibody. EBV titers revealed EBV IgM: >1:10, EBV IgG: >1:1280, EBV Early Ag AB: >1:80, and EBV Nuclear Ag AB: >1:8. CT abdomen and pelvis with intravenous contrast revealed splenomegaly (13.3cm) and abnormally enlarged appendix tip (diameter of 9 mm), consistent with appendicitis. Clinical assessment showed no signs of appendicitis, so no surgical intervention was needed. The patient was discharged on the fourth hospital day with outpatient follow-up. His acute appendicitis, likely secondary to the infiltration of lymphocytes causing the luminal obstruction, resolved over a few weeks. His elevated transaminases at presentation continued to trend down upon discharge and were back to baseline at two month’s follow-up visit. Based on a review of published literature, this may be the first reported case of simultaneous presentation of IM and acute appendicitis, where no surgical intervention was performed in order to treat appendicitis. In the past 10 years, there has been one reported case of simultaneous presentation of IM and appendicitis: a previously healthy homosexual man with appendicitis caused by CMV as evidenced by pathological findings and CMV seroconversion. Interestingly, IM with a positive monospot and EBV IgM were also seen. Kanafani et al. speculated that their patient acquired EBV and CMV during the same time period. For several decades, the preferred therapeutic option for acute appendicitis has been appendectomy. In reported cases of acute appendicitis with IM, clinical indications for surgical intervention were secondary to a variety of abdominal complications, including abdominal peritonitis from perforation, abscess formation, mesenteric adnitis, or persistent fevers with significant leukocytosis. In our case, no surgical intervention was performed because the patient did not have any clinical signs of appendicitis or hemodynamic compromise secondary to appendicitis throughout his hospital stay and two months after discharge. In conclusion, recognition of appendicitis based on CT imaging does not necessarily require surgical intervention in a clinically asymptomatic patient with IM. As was true with our patient, asymptomatic patients with IM and radiographic evidence of appendicitis may be monitored medically with appropriate follow up.
HLH: A Case Study of a Sepsis-Like Syndrome

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Introduction: Hemophagocytic lymphohistiocytosis (HLH) is a rare sepsis-like clinical syndrome that exists in two forms. The primary or familial form typically presents early in life, is often fatal, and is thought to occur in the setting of one or more gene mutations. The secondary form can occur at any age and is induced by an underlying trigger such as infection or malignancy. We present a case of HLH that met all diagnostic criteria for the diagnosis. Case Description: A 28 year old previously healthy gentleman presented with fever for three weeks, associated with chills, diaphoresis, fatigue, and anorexia. Physical examination was significant for a temperature of 99.4 degrees Fahrenheit, heart rate of 105 beats per minute, and hepatosplenomegaly. Work-up revealed a pancytopenia, elevated liver function tests, and CT demonstrating several enlarged lymph nodes. An extensive infectious diseases workup was negative except for an EBV IgG titer of 1:320 and nuclear antigen antibody of >1:8. Additional testing revealed an elevated triglyceride, ferritin, and IL-2R alpha level. A diagnosis of HLH was made, and he was started on chemotherapy. Initial response was excellent; however, one month later, he returned with fevers and chills. Work-up revealed a diffuse large B-cell lymphoma, marked histiocytes, and prominent erythrophagocytosis in the sinusoidal area of the spleen. Despite chemotherapy, his clinic status continued to deteriorate, and life support was eventually withdrawn. Conclusion: The patient met all eight diagnostic criteria for HLH including fever, splenomegaly, cytopenia affecting two or more lineages in the peripheral blood, hypertriglyceridemia (>265 mg/dL), ferritin >500 µg/L, decreased NK-cell activity, hemophagocytosis demonstrated on biopsy, and soluble IL-2 receptor >2400 units/mL. Because HLH may clinically present as part of a sepsis-like syndrome, the diagnosis should be considered in any patient who presents with a systemic inflammatory response, abnormal LFTs, and high triglycerides after an infectious diseases workup is non-diagnostic.
Organizing Pneumonia in a patient with Antiphospholipid and Sjögren's syndromes. An uncommon presentation

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Organizing pneumonia (OP) is a diffuse interstitial lung disease that affects the distal bronchioles, alveolar ducts and alveolar walls. In addition to the cryptogenic form, secondary organizing pneumonia has been associated with connective tissue diseases, such as rheumatoid arthritis, polymyositis – dermatomyositis and only rarely with Sjögren’s syndrome (SS). Furthermore, there are only a few cases reported in association with anti-phospholipid syndrome (APS). We present a unique case of organizing pneumonia in the context of both antiphospholipid and Sjogren’s syndrome. A 36 year old male with history of morphea, deep vein thrombosis (DVT) and pulmonary embolism (PE) presented with a 3 month history of dyspnea on exertion, dry cough, night sweats and weight loss. A month prior to admission a chest CT scan revealed pulmonary embolism and diffuse bilateral peribronchial consolidations. Bronchoalveolar lavage with biopsy showed chronic inflammation. He was managed with antibiotics, anticoagulation and discharged with home oxygen. One month later he was admitted for worsening exertional dyspnea. Initial vitals were significant for oxygen saturation of 88% on oxygen at 4 L/min by nasal canula. He was in mild respiratory distress, with bilateral inspiratory crackles on lung exam. CT scan of the chest showed worsening peribronchial and lower lung parenchymal disease. Serologic testing revealed antinuclear antibody titer >1:640 (speckled) and presence of rheumatoid factor, anti-cardiolipin antibody, lupus anticoagulant, anti-SS-A and anti-SS-B antibodies, as well as absence of anti-double stranded DNA, anti-citrullinated protein, anti-centromere B, anti-ribonucleoprotein and anti-smith antibodies. Based on clinical presentation and serology the patient was diagnosed with concomitant Sjogren’s syndrome and antiphospholipid syndrome. Open lung biopsy was consistent with OP with mild to moderate active interstitial pneumonitis. His symptoms improved dramatically with steroid treatment. On follow up, the patient had continued clinical improvement, now independent of home oxygen. Organizing pneumonia (OP) is a relatively rare disease that typically presents as a non-resolving pneumonia despite adequate treatment. When associated with an autoimmune disease, it rarely presents as the initial manifestation of the condition. Almost 15% of the patients with SS can be asymptomatic and only present with positive antibodies, in which case a salivary gland biopsy confirms the diagnosis. In addition, many patients can have autoantibodies present years before clinical onset of the disease. APS has been rarely associated with OP; to our knowledge there are only 2 cases reported in the literature to date, and none with concomitant SS. Identification of possible underlying autoimmune causes of OP –even if they’re silent initially, like SS –is key for proper management, since the pulmonary symptoms generally respond to the specific treatment of the primary disease process.
Hip Pain From the Gut: A Rare Source of Skeletal Metastasis

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Introduction: Skeletal metastasis as the presenting sign for colon carcinoma, especially of the cecum, has been rarely described in the literature and usually indicates advanced disease with a poor prognosis. This case describes a patient who was diagnosed with multiple skeletal metastatic lesions before detection of his primary tumor. Case: A 51 year old African American man presented to the orthopedic clinic with progressively worsening right hip and groin pain for 3 months. The pain was worsened by weight-bearing and alleviated by rest, but often woke him at night. He also reported an unintentional weight loss of 8 pounds over the same time but denied gastrointestinal symptoms. On exam, there was minimal tenderness to palpation superior to the right greater trochanter. A plain radiograph of the pelvis revealed a right femoral neck fracture also seen on CT, demonstrating an associated lytic lesion. Lab work revealed a normocytic anemia, elevated ESR and CRP, and a mildly elevated alkaline phosphatase; there was no M spike and CEA and PSA were within normal limits. The pathology from bone biopsy revealed a metastatic poorly differentiated adenocarcinoma. Further imaging with bone scan and PET/CT was consistent with the right hip lesion and revealed extensive bony involvement, as well as uptake in the ascending colon, presumed to be the primary. At this time, the patient was seen in GI clinic for screening colonoscopy with biopsy of a cecal tubulovillous mass. Pathology later revealed a poorly differentiated adenocarcinoma with focal signet ring cell features. The patient underwent radiation to the right hip and is being followed by surgical oncology for tumor resection and chemotherapy. Discussion: Colorectal cancer is the third most common cancer among adult men in the United States and the fourth most common cause of death from cancer, however, skeletal metastases as the primary presentation is rare. In two case series analyzing the site of primary cancer in skeletal metastases of unknown origin, only 2 out of 104 patients (1.9%) with identified primary lesions had carcinoma of the colon. Both of these case series recommended imaging including plain radiographs of the chest and affected bone in addition to a whole body scan and thoracic and abdominal CT, but did not recommend CT scan of the pelvis or an examination of the gastrointestinal tract because these seldom revealed the primary lesion. This case is unique because of the presence of skeletal metastasis without evidence of vital organ involvement in a primary poorly differentiated cecal adenocarcinoma with focal signet ring cell features. Though this entity has been described in the literature, it remains exceedingly rare and should be noted for the thorough diagnostic workup despite literature recommendations against a gastrointestinal source of primary malignancy in skeletal metastasis.
COMORBIDITY AND SURVIVAL IN PATIENTS WITH ADVANCED LUNG CANCER


Background: Non-small cell lung cancer (NSCLC) predominantly affects older patients with a median age of diagnosis of 70, and median age of death 71. Increasing age is associated with increasing co-morbidities, and prior studies have demonstrated that multiple co-morbidities, in addition to age, interfere with the selection of optimal medical and surgical therapy for patients with NSCLC. We examined three commonly used co-morbidity indices to assess how they predict survival in patients with stage III or IV NSCLC. Methods: We performed a retrospective chart review of 198 veterans (V) diagnosed with stage III or IV NSCLC from January 2004 to December 2008 at the Veterans Affairs New Jersey Health Care System (VANJHCS). We reviewed demographic, clinical, laboratory and pathology data. Co-morbidity was assessed using the Charlson Comorbidity Index (CCI), Respiratory Kaplan-Feinstein Index (KFIr) subscale, and Cumulative Illness Rating Scale (CIRS). Cox regression analysis was performed using STATA 11. Results: There were 198 veterans with median age 70 years (47-91). 18 veterans were stage IIIA, 50 stage IIIB, and 113 stage IV. The median LDH was 200.5 IU/DL (104-763). Mean CCI was 5 (1-15), KFIr 1 (0-3), CIRS15 5 (0-10), CIRS16 8 (0-17), CIRS 17 1.65 (0-3.8), CIRS 18 0 (0-2), and CIRS 19 0 (0-1). The most common histologies were squamous cell (72 pts, 37%), adenocarcinoma (63 pts, 33%), and NSCLC (29 pts, 15%). The median CCI 5 (1-15), KFIr 1 (0-3), CIRS15 5 (0-10), CIRS16 8 (0-17), CIRS 17 1.65 (0-3.8), CIRS 18 0 (0-2), CIRS 19 0 (0-1). Median survival (days) for stage IIIA pts 348 (23-2411), stage IIIB pts 288 (6-3306) and stage IV pts 147 (3-2664). (p <.01). Median survival (days) for squamous pts 254 days (13-2790), adenocarcinoma 216 (12-3306), and others 124 (3 -1453 (p <.01) Conclusions: By univariate analysis, our results show that stage and the CIRS subscales were significant predictors of survival in patients with stage III or IV non-small cell lung cancer. Neither the CCI nor the KFI were significant predictors of survival. In multivariate analysis with stage and LDH, only the CIRS 15 through CIRS 18 were statistically significant (p <.001) predictors of survival. Elderly patients with late-stage NSCLC are less likely to receive aggressive chemotherapy despite studies showing that fit elderly patients have acceptable tolerability of multi-drug chemotherapy with comparable response and survival to younger patients. Proper assessment of comorbidities may improve our understanding of a patient’s prognosis and tolerance to treatment thereby allowing us to better select patients suitable for aggressive treatment regimens regardless of age. The CCI, CIRS, and KFI are three recognized tools used to assess comorbidities. More research is needed to determine their usefulness in predicting survival of patients with late stage NSCLC.
Background: Osteoporosis is characterized by low bone mass and micro-architectural deterioration of bone tissue leading to enhanced bone fragility and an increased risk of fracture. Osteoporosis-related fractures affect half of all postmenopausal women during their lifetime leading to chronic pain, disability, and increased mortality. Dual-energy x-ray absorptiometry (DEXA) to predict short-term risk for osteoporotic fractures is recommended by the United States Preventative Services Task Force (USPSTF) in women above age 65 years or younger women with increased fracture risk. The goal of our study was to determine the rate of osteoporosis screening using DEXA scan for women aged 65-85.

Methods: This was a retrospective chart review of female patients aged 65-85 seen in the Ambulatory Care Center internal medicine clinic at University Hospital in Newark, NJ from July 1st, 2011 to June 30th, 2012. Primary endpoint was the number of patients appropriately screened for osteoporosis using DEXA scan. Secondary endpoint was the number of patients initiated on appropriate medical therapy for osteopenia or osteoporosis. Results: 100 charts were reviewed of female patients aged 65 – 85 years old. The mean age of the patients was 71.5. The mean number of visits in the year for each patient was 5.9. 72 patients had documentation of a DEXA scan being done or ordered appropriately according to USPSTF guidelines. Of these 72 patients, 29% were found to have DEXA proven osteoporosis. The rate of compliance with DEXA ordering was plotted against the number of visits. The correlation factor (r) was found to be 0.43, a weak correlation. Initiation of bisphosphonate therapy and calcium and vitamin D supplementation was seen in 42% of cases. Conclusions: We found a 72% compliance rate with USPSTF screening guidelines for osteoporosis for women aged 65-85 using DEXA scan. 29% of these patients were found to have osteoporosis. 42% of patients with osteopenia or osteoporosis were initiated on appropriate medical therapy with either calcium and vitamin D supplementation or bisphosphonates. There was not a significant correlation found between the number of clinic visits and osteoporosis screening. Some limitations to our study include a relatively small sample size (n=100) and variability of clinicians practicing in the clinic. It is estimated that approximately 12 million Americans older than 50 years have osteoporosis. One half of all postmenopausal women will have an osteoporosis related fracture during their lifetime; 25% of these women will develop a vertebral deformity, and 15% will experience a hip fracture. Osteoporotic fractures often lead to patient suffering and a remarkable cost burden. When one considers that this is preventable through appropriate screening and treatment, it is apparent that we must intervene now to improve our screening rate and therapeutic intervention.
Cushing’s Syndrome- A Missed Diagnosis

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Case presentation: A 61-year-old woman with hypertension and recent immigration from India presented with progressive shortness of breath along with generalized weakness and swelling. As per family, she underwent extensive cardiac work-up in India, all of which was unremarkable. Home medications included losartan and metoprolol succinate for hypertension. She also admitted to a 9-kg weight gain during the past year. She otherwise denied headaches, changes in vision, focal weakness, fevers, chills, chest pain, palpitations, cough, wheezing, nausea, vomiting, abdominal pain, heat or cold intolerance, or recent illnesses. She denied use of tobacco, alcohol or illicit drugs. She recently was started on torsemide with minimal relief of her symptoms. Upon evaluation, vital signs included BP 137/97 mmHg and BMI of 26. Physical exam was notable for facial plethora, truncal obesity, dorsocervical prominence and 1+ pitting edema bilaterally. Laboratory studies revealed K 2.9, Cl 92, HCO3 36, BUN 44, Cr 1.5 (baseline unknown), and TSH 25.8 g/dL (elevated). Levothyroxine was started for hypothyroidism. Urinalysis was negative for protein or signs of infection. EKG revealed LVH and anterior fascicular block. Further cardiac work-up including TTE and carotid dopplers was normal. Chest x-ray was unremarkable and CT head without contrast showed no pathology. For her shortness of breath a CT PE protocol was performed but showed no pulmonary embolus. It did, however, reveal a 2.9 cm left adrenal hypodensity, suspicious for adrenal adenoma. Work-up for Cushing’s syndrome was initiated, and studies revealed an elevated AM serum cortisol (32.1 μg/dL; normal 5-25 μg/dL). After a 2 mg overnight dexamethasone suppression test serum, AM cortisol level remained elevated (31.2 μg/dL). Three days later, serum ACTH was low (<0.5 pg/mL; normal >0.5 pg/mL) and PM cortisol was elevated (32.5 μg/dL). A 24-hour urine cortisol level was 110 μg/24 hr (normal 3.5-45 μg/24 hr), consistent with a diagnosis of Cushing’s syndrome. Within one month, the patient underwent a left adrenalectomy and was discharged home on steroid replacement therapy with outpatient Endocrinology follow-up. Surgical pathology later revealed evidence of an adrenal cortical adenoma with low proliferation rate. Discussion: Cushing’s syndrome represents a constellation of symptoms consistent with longstanding exposure to supraphysiologic levels of glucocorticoids. Untreated Cushing’s syndrome results in high morbidity and mortality (up to 50% in 5 years), thereby necessitating prompt recognition and intervention. Though our patient presented with clinical findings of hypercortisolism, the diagnosis was made much later. This case illustrates the importance of considering Cushing’s syndrome in the differential diagnosis of a patient with classic cushingoid features such as hypertension, weight gain, and generalized edema.
Incidence of Contrast-Induced Nephropathy in medicine in-patient setting: Analysis of current practices at a Veteran Administration Hospital

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Background: Contrast-induced nephropathy (CIN) is a form of acute kidney injury that occurs after administration of radio-contrast material. CIN is associated with adverse clinical outcomes including increased hospital stay and significant morbidity and mortality. The aim of the study was to determine the incidence of CIN and renal recovery in a veteran’s affairs (VA) population and to evaluate the adherence of CIN prevention guidelines in patients who received intravenous (IV) contrast. Methods: We conducted a retrospective analysis of 206 consecutive patients admitted to the medicine floors at the VA hospital from Sept 2010 – Dec 2012 who received non-ionic low-osmolol IV radio-contrast material (Iohexol). Serum creatinine (sCr) was measured at least once per 24 hours. Only patients with a minimum of 3 sCr readings were included in the analysis. Patients with end-stage renal disease were excluded. CIN was defined as a rise in sCr ≥ 0.5 mg/dL or a 25% relative increase from baseline sCr within 48 hours. The primary endpoint was to calculate the incidence of CIN and renal recovery. The secondary endpoint was to examine the adherence to CIN prevention guidelines, including use of IV fluids and N-acetylcysteine (NAC). Renal recovery was defined as: complete- return of sCr to within &##8805; 20% of baseline sCr; partial- sCr > 20% from baseline; and no recovery as dialysis dependence within a follow-up period of 3 months. IV fluids were defined as isotonic normal saline or sodium bicarbonate received within 1 hour prior to and 6 hours after administration of contrast. Results: The mean (SD) age of the study population was 66.6±12.1 years and 100% were male. The mean (SD) baseline sCr was 1.0±0.5 mg/dL. Sixteen patients (16/206, 7.8%) developed CIN. The mean increase in sCr levels in patients who developed CIN was 0.4±0.1 mg/dL. 30/206 (14.6%) of patients had GFR<60 mL/min/1.73 m2 (1.9% stage 4, 4.4% stage 3b, and 7.8% stage 3a). Patients who developed CIN had a greater prevalence of liver disease as compared to non-CIN patients (37.5% vs. 17.9%, respectively, p=0.02). 3/14 (21.4%) CIN patients had partial renal recovery and 1/16 patients (6.3%) required dialysis during hospital admission. Only 5/16 (31.3%) of patients who developed CIN were documented to be receiving fluids. None of the 16 CIN patients were prescribed NAC and 9/16 patients (56.3%) were continued on nephrotoxic drugs at the time of procedure. Conclusion: Based on our preliminary data, the incidence of CIN in hospitalized male VA medicine patients is high. Furthermore, the adherence to CIN prevention guidelines was suboptimal. Future studies would include the development of a risk stratification schema and implementation of CIN prevention protocol as part of an “electronic alert system” in high-risk patients.
**Rutgers- New Jersey Medical School**

**Blind-ended case of malignant ascites**

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Ascites is most commonly associated with underlying end stage liver disease. However, malignancy associated ascites can be a challenging proposition in the setting of an unknown primary source. The patient is a 54 years old Brazilian woman, with a past medical history of hypertension and hyperlipidemia, who presented to the hospital with a two month history of increasing abdominal girth. During that span, she noticed that her pant size grew from a size eight to a size twenty. The patient also reported dull right and left upper quadrant abdominal pain, early satiety, nausea, vomiting, and diarrhea. She denied any recent travel or sick contacts. Her prior surgical history was significant for total hysterectomy for myomas and breast lumpectomy. Family history was significant for multiple first degree relatives with ovarian, breast, and renal cancers. On physical exam, she was found to have a distended abdomen with positive fluid wave, shifting dullness on percussion, and reducible umbilical hernia. CT scan of the abdomen and pelvis revealed moderate ascites and nodular infiltration of the greater omentum. She underwent a CT-guided paracentesis, which showed a bloody fluid with a SAAG <1.1 indicating absence of portal hypertension. Tumor markers for colorectal cancer and ovarian cancer (CEA and CA-125) were elevated. Over the next two months, she underwent further testing of the omental infiltration found on abdominal CT scan. Transvaginal ultrasound of the pelvis demonstrated complex ascites and omental thickening, suspicious for carcinomatosis. She subsequently underwent surgical exploratory laparotomy. Pathology was consistent with metastatic mucinous adenocarcinoma of the appendix. Intra-operative chemotherapy was administered. Ascites caused solely by cancer accounts for only 7% of patients who initially present with ascites. This case describes a patient whose ascites was secondary to a metastatic adenocarcinoma of the appendix. Diagnostic work up for all patients who present with ascites of unclear etiology is important, as it will guide therapy. Another aspect to consider is the importance of her significant family history of cancer, particularly breast and ovarian cancer. Given her history of 3 affected relatives, 2 first generation relatives, and 1 diagnosed under the age of 50, this patient meets diagnostic criteria for Lynch Syndrome. Properly identifying and screening of these patients is paramount as it can lead to earlier treatment and overall increased survival.
Wegener’s Polyangiitis presenting with symptoms of the common flu

Enoc Fernandez Ginorio, Kevin Gershuny, DO, Neil Kothari, MD, Dan Matassa, MD, Lindsay Thornton, MD

The first sign of a severe underlying disease entity, such as vasculitis, can present with nonspecific findings. Prompt recognition and treatment are sometimes difficult but can prevent future sequelae of this disease. The patient is a 30-year-old man, with no past medical history, who first presented 2 month prior to this hospital admission to an ENT physician complaining of nasal congestion, epistaxis, ear fullness, and tinnitus. At the time, he was prescribed a short course of antibiotics and nasal saline spray. His symptoms initially improved until 4 days before this admission, when he developed a dry cough, subjective fevers, chills, pleuritic chest pain, diaphoresis, night sweats, arthralgias, and 1 episode of hemoptysis. His symptoms did not improve and he was seen in the ED where he was prescribed a seven day course of levofloxacin for suspected bronchitis. The following day, he was seen by an outpatient pulmonologist. A CT scan of the chest showed patchy infiltrates and pleural involvement. He again presented to the ED after his symptoms did not subside with antibiotic treatment. On this current hospital admission, the patient was found to be afebrile, in no acute distress, and saturating 99% on room air. His physical examination was remarkable for decreased breath sounds throughout the right lung field. His lab work showed elevated liver enzymes and alkaline phosphatase. He underwent radiological studies of the chest, which showed peripheral nodular infiltrates bilaterally on x-ray and mediastinal adenopathy on CT scan. The patient was then started on azithromycin and ceftriaxone for presumed pneumonia. Infectious etiologies, such as HIV, hepatitis panel, tuberculosis, and respiratory viral panel, were ruled out. On day 3 of admission, the patient continued to have worsening myalgias and arthralgias. CRP and anti-PR3 were elevated, consistent with the diagnosis of granulomatosis with polyangiitis (Wegener’s). He was started on a three day course of high dose steroids and his symptoms rapidly improved. He was discharged with rheumatology follow up for rituximab therapy. Granulomatosis with polyangiitis is a rare disease occurring in 3 cases per 100,000 people in the US. This case demonstrates the importance of broadening the differential diagnosis when current treatment is not working. It is imperative to understand and recognize the diagnostic criteria, such as abnormal radiologic studies, to adequately diagnose and treat these patients to prevent renal progression of the disease.
Chilaiditti’s syndrome is the rare finding (incidence of 0.025-0.28%) of symptomatic colonic interposition between the liver and diaphragm. Its ability to mimic surgical emergencies including pneumoperitoneum and subphrenic abscess on plain film, however, presents a diagnostic challenge to both medical and surgical clinicians. Additionally, complaints associated with Chilaiditti’s syndrome may overlap with clinical signs of acute abdomen, further obfuscating diagnosis. We present a case of metastatic prostate cancer presenting initially with Chilaiditti’s syndrome in order to underline the importance of this atypical entity. Case: The patient is an eighty-eight-year-old man with a past medical history significant for heart disease, chronic constipation, and diabetes, who presents with severe, crampy abdominal pain. The patient endorsed a history of similar episodes of abdominal pain, which have been increasing in intensity over past year. Additionally he reports a 20 lb weight loss and anorexia for the past year. Initial physical exam revealed a thin man with normoactive bowel sounds and abdomen which was nondistended and nontender to palpation. Laboratory results were significant for elevated alkaline phosphatase of 694 and PSA of 880.87. CT scan of the abdomen on admission and confirming bone scan revealed extensive osteoblastic lesions suggestive for malignancy. On the second day of hospitalization the patient developed increasing subjective abdominal pain with new tenderness of physical exam. Chest xray obtained on the same day revealed increased lucency noted in the projection of the right upper quadrant the abdomen and which suggested a loop of bowel or, less likely, free air. Left lateral decubitus plain film of the upper abdomen also revealed area of lucency in the right upper quadrant the abdomen which was suspicious for free air. Emergent CT scan of the abdomen was obtained and showed eventration of the right hemidiaphragm with interposition of the liver and colon, with no evidence of free intraperitoneal air and therefore no surgical intervention was attempted. Patient was treated for constipation with supportive therapy consisting of laxatives and enema with improvement in abdominal pain. TRUS guided biopsy confirmed prostate cancer. The patient declined surgical intervention and was started on leuprolide/bicalutamide treatment for malignancy and discharged in stable condition. Discussion: The pathogenesis of Chilaiditti’s sign is associated with intestinal, hepatic, and diaphragmatic etiologies, including anatomic variations including absence, laxity, or elongation of the suspensory ligaments of the transverse colon or the falciform ligament, as well as dolichocolons or congenital malpositions. Physical exam findings may range from mild to emergent (acute abdomen). While management of Chilaiditti’s syndrome includes bowel decompression, the rare finding can mimic surgical emergencies including subphrenic abscess, pneumoperitoneum, or bowel obstruction. High diagnostic suspicion should be maintained for patients with such symptoms and appearance of free air on imaging to prevent unnecessary surgical procedures.
Azathioprine-Induced Life-Threatening Myelotoxicity: Are We Aware of TPMT Activity Testing to Identify Susceptible Individuals?

Larysa Gromko

Azathioprine (AZA) is an immunosuppressive antimetabolite used as a steroid-sparing agent in a number of autoimmune conditions. AZA is well known to cause bone marrow suppression. The degree of drug-induced myelotoxicity differs among individuals according to thiopurine methyltransferase (TPMT) activity, which regulates AZA metabolism. A 29-year-old female with medical history significant for relapsing-remitting multiple sclerosis (MS), presented with a 4 day history of epistaxis, fatigue, fever and chills. The patient had been hospitalized 1.5 months prior to presentation for an acute exacerbation of MS and at that time was started on AZA 100mg/day. On admission, the patient was febrile (102.6 F), tachycardic (113/min), hypotensive (BP 96/57), and tachypneic (20/min). The patient was exhibiting rigors and had gingival bleeding. Laboratory studies revealed profound pancytopenia: WBC 0.8 with 6% neutrophils (ANC 48), hemoglobin 4.4, and platelets 4. She was started empirically on Cefepime/Vancomycin for neutropenic fever and was transfused PRBC/platelets to a goal Hb>7 and goal Plt>10. AZA was discontinued as pancytopenia was deemed to be secondary to drug-induced myelotoxicity. On day 3, the patient was hemodynamically stable with Hb 7.7 and Plt 19, without active bleeding. However, she continued to spike fevers daily and remained profoundly neutropenic, WBC ranging from 0.8-1.3 and ANC ranging from 26-72. Filgrastim 400mcg/day was started, with a total of 10 doses administered during hospitalization. All cultures remained negative. Given unremitting fevers, empiric coverage was expanded to include Acyclovir on Day 5 and Micafungin on Day 10. The patient required additional transfusions of PRBC/platelets throughout admission. On Day 16, the patient became afebrile, Hb and Plt counts stabilized and WBC count began to recover, with WBC of 1.6 and ANC of 272. On Day 19, the patient was discharged on Levofloxacin, Fluconazole, and Acyclovir with close outpatient follow-up. Approximately 1 month later, the patient’s TPMT activity was measured and was 9.1 units/mL, which signifies heterozygosity for low TPMT variant. At that time, the patient’s blood counts had not yet normalized, with WBC 3.9 with 30% neutrophils, Hb 7.4, and Plt 124. Discussion: Research has found that individuals who are heterozygous/homozygous for low TPMT activity exhibit a significantly increased risk of developing AZA-induced myelotoxicity due to increased concentration of cytotoxic metabolites. It is suggested that heterozygous individuals be started at 30-70% of the full AZA dose, whereas in homozygous individuals alternative agents should be considered. The FDA recommends that TPMT activity testing be performed prior to starting AZA therapy. If our patient had undergone TPMT activity testing, she could have been spared AZA-induced life-threatening pancytopenia. This case raises an important question: are physicians aware of TPMT activity testing and are they utilizing it to identify susceptible individuals?
Title: The Effects of Rising Time on School Performance in Adolescents

Background: This study analyzes the effect of rising time on the school performance of adolescents. Objectives: The authors aimed to determine whether there is a relationship between rise time and school performance, a constellation of daytime sluggishness symptoms (after school naps, perception of inadequate sleep, daytime sleepiness). The authors also sought out to measure the relationship between changes in sleep schedule since 5th grade and 7th grade and their effects on changes in school performance.

Methods: Anonymous questionnaires were distributed to three New Jersey public high schools and two private schools to students in grades 9-12 during school hours to be completed on a voluntary basis. The questionnaire consisted of 16 categories assessing school day and weekend sleep schedules; sleep duration; daytime sluggishness symptoms; sleep schedule changes since 5th and 7th grades; and current, 5th, 7th grade school performance.

Results: 2100 students completed the survey. Data showed that earlier rise times on school days are associated with better performance (P=0.001). The authors observed that in students getting less than 7.5 hours of sleep on school days, students rising before 6AM were less likely to experience daytime sluggishness symptoms than those rising 6-6:30AM (P=0.02), 6:30-7AM (P=0.047), and 7-7:30AM (P=0.001). Those with at least 7.5 hours of sleep, rising before 6AM were more likely to experience daytime sluggishness symptoms than those rising 6-6:30AM (P=0.01), 6:30-7AM (P=0.003), and 7-7:30AM (P=0.01). Changes in sleep schedule from prior grades were not associated with changes in school performance except changes in sleep duration.

Conclusions: Students that rise earlier are more likely to perform better academically as compared to their peers that rise later. Daytime sluggishness is affected by more than just sleep duration and rising time. Chronotype may be a more important factor than specific sleep stage duration on academic performance. Changes in sleep schedules from previous years are minimally predictive of performance changes.
Cardiovascular disease is the leading cause of death in the United States; men tend to have a higher risk of developing coronary artery disease and usually present at a younger age. Women present at an older age and are more likely to die from myocardial infarction (MI). One simple, overlooked, and easily available therapy can reduce this risk: aspirin. Low dose aspirin reduces the risk of first time myocardial infarction in men. In primary prevention trials among women, aspirin lowered the risk of ischemic stroke (CVA). Randomized trials have also found that aspirin has been associated with 25-32% risk reduction in developing myocardial infarction. The US preventive services task force strongly recommends that clinicians consider aspirin for patients who are at risk for developing coronary artery disease, including all men between ages of 45-79 and all women aged 55-79. The aim of our study was to determine adherence to guidelines with regard to aspirin for primary prevention of myocardial infarction and stoke in our resident clinic. A retrospective chart review of randomized patients with risk factors for CAD and CVA seen at Ambulatory Care Center in Newark, NJ, between May 1st, 2012 and October 29th, 2013 was conducted to determine whether residents placed at-risk patients on aspirin. All patients with cardiovascular risk factors were included, based on documentation of at least one of the following: diabetes mellitus, hypertension, hyperlipidemia, smoking, or family history of premature coronary disease. We excluded patients with established CVA, MI, coronary artery disease or peripheral vascular disease, those allergic to aspirin, those with no follow-up and those documented to lack medical decision making capacity. Over 400 charts were reviewed; 292 met inclusion criteria. 63% of patients were women, 37% men, ages 29-91, 43% diabetic (58% of them had HbA1c>7%), 44% had a smoking history (78% current smokers), LDL ranges: 42% <100, 51% 100-160, 6% 160-200; 31% were overweight, and 43% obese. Per chart documentation, only 24% of all at-risk patients were taking aspirin. This study revealed poor adherence to guidelines with regards to use of aspirin for primary prevention of cardiovascular disease in our resident clinic. Data from the National Ambulatory Care Center survey indicates that only 21.9% of adult patients with diabetes report regular aspirin therapy. Even though in our clinic adherence to aspirin therapy is slightly above this average, it is still very low. However, our estimate may be falsely low due to improper documentation of over the counter medications. We propose interventions to increase awareness of aspirin use such as educational campaigns for patients, mini-lectures for ambulatory resident cohorts, automated EMR reminders and adding aspirin therapy to health maintenance smart texts to achieve our goal of 100% compliance.
THINKING OUTSIDE THE LIVER- AN UNUSUAL CASE OF TRANSAMINITIS

Shuchie Jain, Weekon Choi

Introduction: Gastrointestinal involvement is seen in 0.1 to 0.9 percent of patients with sarcoidosis. We present a patient with presentation of failure to thrive with transaminitis and hypercalcemia found to have non-caseating granulomas on liver biopsy. Prompt diagnosis and treatment is pertinent for these patients to prevent permanent organ damage. Case: 62 year old male with past medical history of nephrolithiasis, bronchiectasis, CABG and aortic valve replacement secondary to endocarditis presents to the emergency department for generalized weakness and failure to thrive. Patient reported a 40 pound weight loss in the last 4 months secondary to decreased appetite with increased right upper quadrant pain. He denied any coughing, night sweats or fevers. He did not have a history of smoking but reported drinking in the past. He was also hypotensive and tachycardic on presentation. On physical examination patient appeared cachetic and mildly lethargic and the rest of the exam was unremarkable. Patient was mildly anemic at 11.8. He had hypercalcemia of 12.1 and transaminitis with AST/ALT of 236/170 with alkaline phosphatase of 1116. Other pertinent labs included prolonged PT/PTT of 16.6/42.2 with an INR of 1.4, albumin of 2.0 and an elevated lipase of 610. Chest X-ray showed a chronic right lower lobe infiltrate with no adenopathy. Abdominal ultrasound showed fairly marked hepatomegaly without mass or ductal dilatation. CT abdomen without contrast showed hepatomegaly and a right lower lobe peripheral infiltrate. Patient was started on aggressive IV hydration which did not improve his hypercalcemia. PTH and UPEP/SPEP for evaluation of the hypercalcemia and both were within normal limits. Vitamin K was also administered for the elevated INR. On day 4 of hospitalization, an elevated ACE of 183 was noted. A liver biopsy was conducted which showed results of moderate non-caseating granulomatous inflammation diagnosing sarcoidosis infiltration in the liver parenchyma without pulmonary involvement. Patient was started on Prednisone 40mg for one month with a gradual taper. His liver function tests improved and symptoms resolved months later. Discussion: Sarcoidosis is a systemic disease affecting many organs however rarely infiltrating the gastrointestinal organs. Hepatomegaly is the most common presenting symptom however transaminitis is seen in only 20-40 percent with hepatic sarcoidosis. This case demonstrates classic laboratory findings of sarcoidosis such as hypercalcemia and elevated ACE levels however with no pulmonary manifestations. Treatment with steroids is important to be started for prevention of any further liver damage and reduction of symptoms.
A CASE OF THYROID CANCER WHERE IT’S LEAST EXPECTED

Shuchie Jain, Maya Raghuwanshi

Introduction: Tumors metastatic to the mandible are infrequent and exceedingly rare if they are of thyroid origin. Recognition of papillary carcinoma is crucial for early treatment. We present a rare case of metastatic papillary carcinoma to the mandible with no detected primary cancer in the thyroid. Case: 44 year old female with no significant past medical history who presented with an increasing right sided mandibular mass. Patient underwent biopsy of the mandibular mass and a right sided enlarged neck node. Biopsy of the mandible showed metastatic carcinoma consistent with a thyroid origin. Tumor invasion through the bone with involvement of the adjacent skeletal muscle was noted after resection of the mass. The morphology of the tumor was consistent with papillary carcinoma of the thyroid. Immuno-histochemical staining revealed positive staining for thyroglobulin which thus supported the above diagnosis. Benign salivary gland tissue was obtained from a fine needle aspiration of the right parotid. Patient underwent fine needle biopsy for the search of the primary cancer which revealed scant groups of follicular cells in rosette like clusters suggestive of a follicular lesion. Total thyroidectomy was conducted 2 months after diagnosis of the metastatic lesion with a high suspicion of primary cancer in the thyroid gland. However, pathology of the thyroid and para-tracheal lymph node resection were negative for neoplasm. Thyroid gland only showed nodular hyperplasia in the right and left lobe. Radioactive iodine uptake was also performed on the patient after the total thyroidectomy which showed increased uptake in the zygomatic region. Patient underwent a second radioactive iodine ablation 2 years later. Patient has been cancer free for greater than 10 years. Discussion: Most papillary thyroid cancers present with a palpable thyroid abnormality. In this case, the primary presentation was mandibular bone metastases with no overt thyroid anomaly. Metastases are uncommon beyond the neck in this cancer type. Only 25 percent of those patients with metastases beyond the neck have skeletal involvement. This case is especially rare given that there was no primary cancer found in the thyroid gland. Though metastases of papillary carcinoma to the orofacial region are rare it should be considered when a patient presents with a suspicious lesion. Early detection and treatment may decrease mortality in this patient population.
A RARE CASE OF NMDA-R ENCEPHALITIS WITH NEW ONSET BRUGADA SYNDROME

Shuchie Jain, Nidhiben Anadani, Ramneek Nakai, Peter Ricketti, Enrique Feoli

Introduction: Delirium in a younger patient is seldom of autoimmune origin. We present a case of increasing unresponsiveness with catatonia, psychosis, seizures leading to respiratory instability in a young male. NMDA receptor encephalitis is a rare neurological emergency which should be suspected with the above manifestations. Co-existence of brugada syndrome with NMDA-R encephalitis is also an uncommon occurrence with possible link to SCN5A channel mutation causing recurrent seizures and arrhythmias. Case presentation: 26 year old Philippino male with past medical history of a childhood febrile seizure presented with a new onset generalized tonic-clonic seizure. Vitals signs and physical examination on admission were unremarkable. Patient was alert and oriented. CBC, chemistry, urine toxicology was within normal limits. Patient was started on Tegetrol. CT head and MRI brain without contrast showed no intracranial pathology. Electroencephalography showed right temporal slowing and sharps with a right temporal lobe seizure. Patient was discharged home after 3 days. 7 days later patient presented to the hospital after a syncopal episode and heart palpitations. On admission patient was disoriented with no focal neurologic signs. Electrocardiogram showed convex ST elevation and T wave inversion in V1-V3 consistent with type -1 brugada syndrome. Video EEG revealed background slowing and a seizure was captured that originated in the left temporal lobe. Few hours after presentation, he was combative, biting himself and others. Tegetrol was stopped and Depakote was initiated. Multiple crises were called for agitation and abusive behavior during the hospitalization. On day 9 of admission, new oral dyskinesia, drooling and flat affect were seen. Lumbar Puncture showed lymphocytic predominance and was negative for viral and fungal serology. Repeat brain MRI with contrast did not show any abnormalities. Patient was intubated to protect the airway and was transferred to the ICU for further management. Video EEG continued to show occasional seizures with bilateral temporal spikes and sharps. Keppra and Vimpal were started and Depakote was discontinued. Treatment with Solumedrol 1gm for 5 days did not improve his condition. Secondary to increased self-extubations he was paralyzed with vecuronium. Plasmapheresis for 5 days did not alleviate symptoms. After day 14, tracheostomy placement was pursued. CSF for NMDA receptor antibody send out returned positive. Rituxamab and Cytoxan treatment was initiated and improved his mental status and dyskinesias. Discussion: Anti-N-methyl-D-aspartate receptor(NMDAR) encephalitis is a newly recognized disorder seen mostly in children and adolescents. The condition is confirmed by NMDA antibody in the CSF. There are some described cases of coexisting epilepsy and brugada syndrome. However, brugada syndrome with anti-NMDA receptor encephalitis has not been described previously. We want to explore their relationship and the possibility of brugada syndrome being part of the autonomic instability seen in NMDA receptor encephalitis.
Introduction Congenital Long QT syndrome type 2 is a rare inherited cardiac abnormality resulting in QT prolongation associated with the risk of polymorphic ventricular tachycardia. This patient’s risk of a cardiac event was increased due to her postpartum state. Case Description - A 21 year old, two month post partum female presents following a three day history of syncopal episodes. The first occurred when her son set off her cell phone alarm, second with the son playing a loud video game. During each event the husband reported that the patient was pulseless and perform 2-3 minutes of CPR prior to return to spontaneous circulation. She was place on standard monitoring in the emergency department. During this time she had a “syncopal” episode which correlated with polymorphic ventricular tachycardia on telemetry monitoring. EKG revealed QTc of 530. The patient was also found to be hypokalemic at 3.2. Her QT interval remained prolonged following repletion. The patient’s only medication was depot provera. She failed therapy with beta blockade and on the fourth day of admission a dual chamber pacemaker with ICD was placed for secondary prevention of sudden cardiac death. She was monitored without an event and was discharge home to follow up with electrophysiology. Genetic testing was sent for analysis. Discussion - Long QT syndrome type 2 is associated with alarm sounds as a precipitating factor, as it was in this case. The hormonal impacts have been recently described with estrogen resulting in QTc prolongation and progesterone resulting in QTc shortening. Thus the dramatic shifts in estrogen/progesterone levels in the post partum state has resulted in the initial cardiac presentation of long QT type 2 in several patients with this underlying congenital disease. While endogenous progesterone provides protection the synthetic depot provera does not activate the endothelial nitrogen oxide synthetase and thus provides no cardiac protection during the post partum period. The post partum state and potentially the use of medroxyprosterone acetate, which has been implicated as a potential exacerbating factors in recent literature, precipitated her cardiac events. This is a rare cause of syncope that should be considered in the differential when assessing post partum patients presenting with syncope.
Introduction: Tuberculosis in the United States usually results from reactivated latent disease and at-risk populations should be screened. While tuberculosis has a myriad of manifestations, oral tuberculosis is very rare with an incidence of 1.4%. We describe a case of disseminated tuberculosis in an immunocompromised patient who presented with lingual disease. Case description: A 59 year-old Haitian woman with a history of recently diagnosed dermatomyositis treated with glucocorticoids, presented with a painful ulcer and mass of the tongue in addition to persistent right shoulder pain. The shoulder pain was present for four months, and previously treated with intravenous antibiotics for suspected septic arthritis. The painful tongue lesion began after biting her tongue and was progressing over two months. The tongue ulceration was treated as an outpatient with famcyclovir without improvement. On review of systems the patient denied fever, chills, night sweats, cough, or shortness of breath. On examination, she had a three by three centimeter ulcerative lesion of the left lateral aspect of the tongue with irregular indurated borders and white exudative material as shown in Figure 1. Additionally, she had limited range of motion in the right shoulder due to pain. A chest radiograph demonstrated diffuse micronodular densities consistent with a miliary pattern. Biopsy of the tongue lesion was obtained, and stained positive for acid-fast bacilli. Multiple induced sputum specimens were positive for Mycobacterium tuberculosis. She was started on an anti-tubercular regimen of rifampin, isoniazid, pyrazinamide, and ethambutol with marked improvement of both the tongue lesion and shoulder pain. Discussion: This is a rare case of disseminated tuberculosis in a patient with dermatomyositis, where the main manifestation of disease was of the tongue. However, it is presumed that there also was tubercular involvement of the shoulder. The index of suspicion for tuberculosis should remain high in patients who are immunosuppressed. Extrapulmonary and disseminated tuberculosis occur frequently in this population. While oral tuberculosis is very rare, it is likely to occur when pulmonary disease is present and infected sputum enters breaks in mucosal surfaces or there is hematogenous spread. Often times, patients who have an oral lesion will report prior trauma to the area as reported in this patient. Maintaining a high index of suspicion for unusual manifestations of tuberculosis is essential as clinicians continue to increasingly and aggressively treat rheumatologic conditions.
Inhibition of Histone Deacetylase Enhances 1,25(OH)2D3 Induction of Cathelicidin Transcription

Ki-Yoon Kim

Purpose of Research: We earlier reported that 1,25(OH)2D3 induces the expression of Cathelicidin Antimicrobial Peptide (CAMP) gene in airway epithelial cells and results in an increase in antibacterial activity. This study demonstrates for the first time that the short chain fatty acid butyrate, a histone deacetylase inhibitor (HDACi), enhances 1,25(OH)2D3 induction of CAMP transcription in A549 human lung epithelial cells. Brief Overview of Methods: Transient transfection of A549 cells A549 (adenocarcinomic human alveolar basal epithelial cells) cells were transfected with promoter constructs linked to the firefly luciferase reporter gene and treated. Luciferase assay was conducted and the results were represented as fold induction compared to basal levels of the control group. Statistical t-test analysis was used to test for significance. Reverse-Transcription PCR Total RNA was extracted from cultured A549. Reverse transcription and PCR amplification of target gene were performed. The resulting expression bands were quantified, corrected with GAPDH and normalized to the control group. Results: Transcription assays in A549 cells revealed that 1,25(OH)2D3 (10 nM, 24h) or butyrate (2 mM, 24h) stimulates CAMP transcription (hCAMP promoter -693/+17) by 2 and 6 fold, respectively. Combined treatment by butyrate and 1,25(OH)2D3 resulted in a 21.5 +/- 3.7 fold induction in CAMP transcription, indicating that decreased histone deacetylation amplifies the effect of 1,25(OH)2D3 on CAMP transcription. RT-PCR demonstrated that CAMP mRNA levels, when compared to control, were also found to be increased by 1,25(OH)2D3, C/EBPα, butyrate and combined treatment of C/EBPα and 1,25(OH)2D3. Additional transcription assays in A549 cells revealed that 1,25(OH)2D3, C/EBPα; butyrate also significantly stimulate the transcription of CD14 (p227 CD14-luc), thus demonstrating 1,25(OH)2D3 and butyrate induction of another component of innate immunity. Butyrate was also found to induce the transcription of 24(OH)ase (5.7+/0.9 fold). However, unlike induction of CAMP and CD14 transcription, induction by butyrate of 24(OH)ase transcription was not as potent as 1,25(OH)2D3 induction. Conclusion: Butyrate enhances 1,25(OH)2D3 induction of cathelicidin transcription in human lung epithelial cells. C/EBPα; as well as 1,25(OH)2D3, induces cathelicidin mRNA expression. Lastly, 1,25(OH)2D3, C/EBPα; and butyrate stimulate transcription of CD14, which is known to amplify the antimicrobial response. These findings suggest the importance of epigenetic control by histone acetylation of 1,25(OH)2D3 regulated antimicrobial activity in lung epithelial cells. Therefore, these findings, related to the enhancement of antimicrobial capabilities, have long reaching implications for the treatment of infection and for modulation of host immune responses.
HIV/AIDS-associated Burkitt lymphoma with isolated extranodal presentations in gastrointestinal tract

Xiangli Li, Ying Tang, Chithra Balasingham, Tatyana A. Feldman

HIV-associated Burkitt lymphoma has low incident among patients with ≤50 CD4 lymphocytes/µl and involves lymph node and extranodal organs. Here we reported a case of HIV/AIDS-associated Burkitt lymphoma with isolated extranodal presentations in gastrointestinal tract. A 46 year old woman with history of HIV for 24 years without antiretroviral therapy (currently CD4 count 46) and hypothyroidism presented with recurrent synoptic episodes, abdominal pain, nausea, weight loss and decreased appetite. Patient was found to have anemia, GI bleeding and underwent EGD/Colonoscopy with biopsies by which she was diagnosed Burkitt lymphoma in gastric, small intestine and ileocecal valve, positive for EBV and c-MYC (8q24) rearrangement. CT scan of abdomen and pelvis, chest and head did not reveal lymphadenopathy and other extranodal involvements. Skin biopsy and lumbar puncture results showed negative for Burkitt lymphoma. She received 6 cycles of chemotherapeutic regimen, CODOX-M then hyper-CVAD, and ended up with EPOCH-Rituximab. Awareness of the clinical settings contributes to early detection and early treatment of aggressive Burkitt lymphoma.
Rutgers- New Jersey Medical School

**Tryptophan Metabolism in Hemodialysis Patients**


Background: Tryptophan (TRP) levels and augmented concentrations of its metabolites have been shown to be significantly lower in animal models of renal insufficiency. We sought to compare the levels of TRP and its metabolites between hemodialysis (HD) patients and healthy subjects and to examine the extent to which TRP metabolites are associated with inflammation in HD patients. Methods: Venous blood samples were drawn in healthy subjects and HD patients. TRP, para-cresol sulfate (PCS) and kynurenine (KYN) metabolites were measured by reverse-phase high-performance liquid chromatography (HPLC), with appropriate detection wavelength for each compound. C-reactive protein (CRP) was measured with an immunoturbidometric technique. We used Spearman rank test for correlational analysis. Results: We studied 30 HD patients (70% male; mean age 58.8±13.3 yrs) and 10 healthy control subjects (60% male; mean age 43.9±12.4 yrs). KYN and KYN/TRP (6.0±2.5 uMol/L vs. 2.1±0.5 uMol/L; p=0.002 and 0.17±.06 vs. 0.03±0.01; P<0.0002, respectively) were significantly higher and TRP levels were significantly lower (35.3±8.1 uMol/L vs. 67.9±9.8 uMol/L; p<.0001) in the HD patients than those in the controls. KYN/TRP ratio and CRP levels were positively correlated in HD patients (r²= 0.14; P=0.05). Conclusions: Low TRP and high metabolite levels were observed in HD patients, indicating increased TRP catabolism, possibly related to immune activation/inflammation. Further studies exploring the biological and functional consequences of increased TRP catabolism in HD patients are warranted.
Rutgers- New Jersey Medical School

Omeprazole-induced Acute Tubulo-Interstitial Nephritis

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Background: Proton pump inhibitors (PPIs) are widely prescribed to treat acid-related gastrointestinal disorders. We report a case of reversible acute renal failure due to acute tubulointerstitial nephritis (AIN), confirmed by histology of a renal biopsy, associated with taking omeprazole. Body: A 70-year-old female presented with complain of generalized weakness and abdominal pain for last 2 months. She had a past medical history for HTN, diabetes, and CKD (baseline sCr 1.6 mg/dL). Physical examination revealed pale conjunctivae. There was no pedal edema, skin rash, petechiae or purpura. Lab results showed elevated BUN of 53 mg/dL and sCR of 2.4 mg/dl. Her Hb was 11g/dL. Urinalysis revealed proteinuria and eosinophiluria. Complements were normal. Serological markers for Hep B, Hep C, and HIV were negative, and no cryoglobulins were detected. Renal ultrasonography was normal. Patient was started on IV fluids however her sCr kept worsening with peak sCr of 4.1 mg/dL. Nephrology consult was obtained and kidney biopsy performed. Biopsy results showed interstitial inflammation with active tubulitis and acute tubal injury. Biopsy also showed chronic features including interstitial fibrosis and tubular atrophy. Overall, findings were consistent with severe interstitial nephritis. The patient had initiated treatment with omeprazole 3 months prior to admission. Omeprazole was discontinued in view of the diagnosis of AIN. Patient was started on oral prednisone for 6-8 weeks with further tapering. The patient sCr gradually improved with subsequently serum creatinine concentration leveled off at 1.2 mg/dl. Conclusions: The pathogenesis of omeprazole-associated AIN is unclear and may be related to the involvement of both humoral and cellular immune mechanisms. Physicians should be aware of this disorder. Accurate and timely diagnosis and withdrawal of the offending drug can prevent potentially life-threatening renal failure.
Introduction  The American Diabetes Association has guidelines for the outpatient management of diabetes. These guidelines include risk factor monitoring, diabetes control, and management of end-organ complications. In 2012, we assessed the degree to which residents in the RUCare Internal Medicine clinic documented evidence based management of type 2 diabetics. In reaction to our findings, we created an optional Smart Text template checklist in the electronic health record and educated residents on its use. In this follow-up study, we assess resident use of ADA guidelines after implementation of this checklist. Methods We conducted a retrospective chart review of 268 encounters of patients with type 2 diabetes between August and November 2013. Residents had previously been educated on management guidelines and on how to use the Smart Text. A laminated reminder about the Smart Text had been attached to the monitor in each exam room in July 2013. Patients were sampled evenly across different resident groups. Progress notes were evaluated for documentation of ADA outpatient guidelines. These results were compared to data from our previous study (before the Smart Text was implemented). The difference between the results was assessed via z-score and difference in confidence intervals. Results The clinic requires blood pressure measurement and finger stick glucose documentation in 100% of encounters, recorded by medical assistants. There was significant improvement in acknowledgement of A1C values over the past six months, in 98% of the post intervention encounters versus 85% of the pre-intervention encounters (p<0.0002). Significant improvement was also found in documentation of eye exams (82% vs. 53%, p<0.0002), foot exams (80% vs. 53%, p<0.0002), urine microalbumin (70% vs. 34%, p<0.0002), and LDL cholesterol (82% vs. 56%, p<0.0002). There was improvement in counseling for both diet (52% vs. 41%) and exercise (43% vs. 35%), though these values did not meet significance. The Smart Text was used in 20% of encounters. Smart Text users met 98% of the measured goals while non-users met 74%. Even non-users, however, showed significant improvement in several of the categories above compared to pre-intervention data. Discussion Adherence to ADA guidelines for diabetes management have improved since education, introduction of the Smart Text and laminated cards. Use of a Smart Text checklist ensured 98% acknowledgement of measured goals. Three months after introduction of the laminated Smart Text card, the adoption rate is still low at 20%. However, overall adherence to ADA guidelines has increased in both users and non-users. This multifaceted intervention is associated with improved care and documentation of management for type 2 diabetes.
Lumbar Drain Placement in a Case of Cryptococcal Meningitis With Transient Cranial Nerve Palsy

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Cryptococcal meningitis is an AIDS defining opportunistic infection characterized by elevated intracranial pressures (ICP). Relief of ICP can be lifesaving and in conjunction with antifungal therapy is an important part of the treatment of this disease. Serial lumbar punctures are the mainstay method for relief of ICP, but in certain cases lumbar drain placement may be indicated. We report the case of a 24 year old man with AIDS who presented to the emergency room with severe headache, neck stiffness, fever, and malaise that had slowly progressed over a 1-2 week course. The patient was found to have elevated opening pressures of 40 cm H20 on lumbar puncture, yeast present in CSF, and cryptococcal antigen titer of 1:1024 in CSF fluid. He was admitted for induction therapy with amphotericin B, flucytosine, and serial lumbar punctures. On day 18 of hospital course the medical team was conducting bedside rounds when the patient complained of blurry vision and acute worsening of headache. On physical exam the patient had ptosis of the left eye. The left pupil was dilated to 5 mm, nonreactive to light, and the patient was unable to adduct his left eye past the midline. The right eye and remainder of the neurological exam were normal. The left eye deficits resolved spontaneously within 10 minutes. A CT scan of the head and dilated eye exam done the same day were both normal. Despite aggressive medical therapy the patient continued to have severe headaches and episodic third cranial nerve (CN III) palsy witnessed by members of the medical team. Serial lumbar punctures were performed, revealing high opening pressures of 40 to > 55 cm H20. The lumbar spine area at the level of L4/L5 became tender and an MRI scan of the region showed a hematoma attributed to repeated lumbar punctures. Neurosurgery was consulted for placement of a lumbar drain for relief of persistent ICP. After drain placement, the patient symptomatically improved and did not experience any further episodes of CN III palsy while completing induction therapy. This case illustrates the successful use of a lumbar drain for relief of persistent ICP in the setting of cryptococcal meningitis complicated by transient CN III palsy. Persistently elevated ICP is a poor prognostic factor in cryptococcal meningitis and despite aggressive treatment some of these patients do not do well. The indications for lumbar drain placement in place of serial LPs are not well established but may include cranial nerve deficits or local soft tissue trauma at the L4/L5 level. Advantages of drain placement include ongoing removal of cerebral spinal fluid and leveling of ICP during induction therapy, while the risk of infection and complications related to drain placement is relatively low.
Severe Alcoholic Lactic Acidosis without Anion Gap

Ramneek Nakai, Suniya Khan

Intro: Lactic acidosis is a major metabolic abnormality that occurs frequently in chronic alcoholism, especially in the setting of malnutrition. These patients are often found to have a concurrent ketoacidosis; however, lactic acidosis in the absence of ketoacidosis or an osmolal gap is rarely reported in literature. We present a patient with profound lactic acidosis secondary to alcohol use who recovered with intensive care therapy. Case presentation: A 56 year old Caucasian man presented to the emergency department complaining of weakness and nausea for 3 days. He reported drinking a bottle of wine (750ml) every day for the past seven days while on a cruise, along with a history of chronic alcohol use. He denied having any other symptoms. On physical examination he was tachycardic and appeared dry. Labs were notable for a hemoglobin of 12, hematocrit 41, MCV 105.2, Cr 1.5, CO2 level < 5, total bilirubin 4.5, AST 213, ALT 57, lactic acid > 24, serum osmolarity 329 without an osmolal gap, and an anion gap of > 57. Arterial blood gas showed pH 6.65, PCO2 11.3, and HC03 1.2. Urine and blood samples were negative for ketones. Ethylene glycol was also negative. He was diagnosed with a severe alcoholic lactic acidosis and transferred to the intensive care unit. Aggressive fluid resuscitation was initiated along with IV sodium bicarbonate, thiamine, and folic acid. Although subsequent arterial blood gases demonstrated improvement, the patient rapidly deteriorated on day two of his admission requiring intubation for airway protection and hemodialysis. By day 3, he demonstrated clinical and laboratory improvement, and repeat arterial blood gas confirmed resolution of his acidosis. Dialysis was discontinued after 5 days and the patient was successfully extubated. Discussion: Although the patient presented with very mild symptomatology, the gravity of his acidosis was discovered on labs obtained as the result of a suspicious history. Lactic acidosis should be considered in any chronic alcoholic patient with a history of binge drinking and low caloric intake who presents with symptoms of nausea, vomiting, and abdominal pain, as prompt recognition and aggressive management is the key to survival. Ann Intern Med. 1990 Oct 15;113(8):580-2. Increased osmolal gap in alcoholic ketoacidosis and lactic acidosis. Schelling JR, Howard RL, Winter SD, Linas SL. Clin Toxicol (Phila). 2005;43(3):161-6. Does ethanol explain the acidosis commonly seen in ethanol-intoxicated patients? Zehtabchi S, Sinert R, Baron BJ, Paladino L, Yadav K.
**Rutgers- New Jersey Medical School**

**Tuberculous Leptomeningitis in a Previously Healthy Camper**

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**Introduction:** Tuberculosis meningitis (TBM) is a rare infection of the central nervous system with devastating consequences if not recognized and treated early. TBM occurs due to the rupture of a small intracranial tuberculoma (Rich focus) that causes a basal meningitis, leading to hydrocephalus, cranial nerve palsies, and ischemic brain injury secondary to tuberculous vasculitis. Diagnosis is challenging due to non specific symptoms and limited sensitivity of existing laboratory techniques. We present a case of a healthy woman who rapidly developed tuberculous leptomeningitis. Case: 40 year old Hispanic woman with no past medical history presented with persistent back pain for about a week which began two weeks after returning from a camping trip in Delaware. She denied any other symptoms or evidence of a rash, insect, animal, or human bite. As her laboratory and radiological workup was unremarkable she was discharged home. Patient returned to the ED after one week complaining of headaches, left sided weakness, blurry vision and increased lethargy. Her laboratory workup was negative except a low sodium of 126. CT scan on admission was unremarkable and LP performed revealed a leukocytosis with lymphocyte predominance (90%), markedly elevated protein and decreased glucose. 2% NS and empirical vancomycin, ceftriaxone, ampicillin, doxycycline, and acyclovir were initiated. She was intubated due to worsening hypoxia. Diagnostics of note HIV, Hepatitis C, Lyme IgM, ANA, ESR and drug screen were all negative. Initial MRI brain revealed small cerebellar infarcts but no temporal lobe involvement and EEG showed moderate, diffuse, cerebral dysfunction, consistent with encephalopathy. Patient remained afebrile and normocytic but in proceeding 3 days brain function deteriorated. Repeat CT head showed diffuse cerebral edema and repeat MRI revealed severe global meningoencephalitis with patchy white matter hyperintensities. Brain perfusion scan confirmed brain death and patient was terminally extubated. Autopsy performed revealed significant necrotizing meningoencephalitis with demonstrable acid fast bacilli, confined to the base of the brain and entire spinal cord, with fibrinoid necrosis of leptomeningeal blood vessels that had lead to focal infarction of the CNS parenchyma. **Discussion:** CNS tuberculosis is the most severe form of this disease and must retain high suspicion as early diagnosis and prompt treatment are key to survival. Due to rarity and non-specific clinical presentation, this condition is often missed in developed countries. CSF typically shows lymphocytic leukocytosis, elevated protein, and low glucose. Imaging often reveals hydrocephalus and diffuse basal enhancement. Hyponatremia is also a common complication. If suspected treatment should be started empirically. Current WHO guidelines recommend treatment with the standard 4-drug regimen (rifampicin, isoniazid, pyrazinamide, and streptomycin or ethambutol) for 2 months, followed by a two-drug regimen for 10 months. Corticosteroids have also been found to reduce mortality.
CMV encephalitis in a Patient with DLBCL Treated with Rituximab-based Chemotherapy

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Introduction: Rituximab has revolutionized the treatment of antibody-mediated and antibody-associated diseases. There is increasing evidence that the use of rituximab is associated with infectious complications. We present the case of a patient with primary CNS DLBCL and AIDS who developed CMV ventriculoencephalitis while on therapy with rituximab. This case adds to the growing literature of potentially fatal CMV infections associated with rituximab-based chemotherapy.

Case description: A 55-year-old man was transferred from an outside hospital after developing acute left-sided hemiplegia. Neuroimaging demonstrated a right frontal lobe mass, blood tests revealed HIV (with a CD4 count of 60/uL), CMV and biopsy was consistent with diffuse large B cell lymphoma (DLBCL). He was treated with high-dose methotrexate and rituximab for DLBCL along with HAART/ganciclovir for HIV/CMV, respectively. Due to the development of thrombocytopenia, ganciclovir was held; at that time CMV PCR was undetectable in the blood. Three months and three cycles of chemotherapy later, restaging MRI showed decrease in the right frontal lobe mass but suggested dissemination of lymphoma along the ependymal surfaces intracranially. The patient reported mild fatigue but was otherwise asymptomatic. LP was negative for malignancy but returned CMV PCR positive with 29,300 copies/ml. Ophthalmological exam was unrevealing for CMV retinitis. Encephalitis was treated with foscarnet after which CMV by PCR in the blood and spinal fluid decreased appropriately. He was eventually transitioned to valganciclovir and cytarabine consolidation therapy for DLBCL.

Discussion: CMV encephalitis is common in patients with HIV/AIDS, typically those with a CD4 count less than 50/uL. Our patient had newly diagnosed HIV/AIDS as well as CMV infection complicating primary CNS DLBCL. He demonstrated progression of CNS lesions despite being on chemotherapy, albeit with a CD4 count of 81/uL at that time. Differentials included progression of lymphoma but also included CMV ventriculoencephalitis. The MRI demonstrated typical findings of curvilinear high-signal intensities along the ventricular wall that prompted lumbar puncture and assessment of CMV viral load. An increase incidence of viral infectious complications have been reported in patients taking rituximab for treatment of malignancies.[1] Although Lanini et al. initially reported no overall increase in incidence of severe infection, a follow-up study suggested HIV status and aggressiveness of lymphoma may place patients at greater risk of infection during therapy with rituximab.[2] Thus there may be a role for active screening and management of viral infections in patients with HIV and aggressive lymphoma.

Prophylaxis Against Inappropriate PPI Use: A Follow-Up

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Introduction

The use of proton pump inhibitors (PPIs) has long been used as “gastrointestinal prophylaxis” in hospitalized patients. The effectiveness of these drugs in reducing stress ulcers in critical care patients is well documented and supported by published literature; however, as we demonstrated in an earlier study, PPIs are often inappropriately prescribed in the inpatient, non-ICU setting based on the FDA prescribing recommendations. In this follow-up study, we attempt to reduce the number of inappropriate prescriptions by instituting an educational intervention.

Methods

A resident lecture was held outlining the current FDA approved indications for instituting pantoprazole. We then performed a retrospective chart review on 100 randomly selected patients admitted to the medical service over the next 3 months. The major outcome was the appropriate or inappropriate ordering of pantoprazole upon admission to the medical service, based on criteria from FDA prescribing guidelines. These data were then compared to data collected prior to the intervention.

Results

In our follow-up study, eight patients (8%) met the FDA criteria for pantoprazole administration. Seven of these patients received pantoprazole upon admission to the medical floor. Twenty-seven patients (27%) received pantoprazole despite not having any clear indication for its use. Sixty five patients (65%) did not have an indication for pantoprazole administration and were not placed on pantoprazole. Overall, guidelines were followed in 72% of patients admitted to the general medical service over the three month period following our intervention compared to 51% of patients prior to our intervention (p=0.001).

Conclusion

The results of our follow-up study showed statistically significant improvement in the appropriate use of pantoprazole in non-critically ill patients admitted to the medical ward. With further educational interventions to both resident and attending physicians, we expect to see further adherence to pantoprazole prescribing information. The recent institution of an electronic ordering system may be of further benefit by implementing a hard stop if pantoprazole is ordered without an indication or by having an order set available for those patients with a history warranting its use.

References

Introduction: Asthma is a common chronic inflammatory disease of the airway characterized by reversible airflow obstruction and bronchospasm. The prevalence of asthma has increased significantly since the 1970s; as of 2011, more than 235 million people worldwide were afflicted with asthma, resulting in 250,000 deaths annually. It is thought to be caused by a combination of genetic and environmental factors. The most common symptoms include: cough, wheezing, chest tightness, and shortness of breath. It is clinically classified in 4 categories of severity according to frequency of symptoms, forced expiratory volume in one second (FEV1), or peak expiratory flow rate. A reliable system for diagnosis and severity classification is essential to guide treatment and reduce risk of death.

Methods: Use of asthma severity classification to guide step-wise treatment has been shown to decrease the risk of exacerbations and death (S. Tual, et al. Allergy. April 2008.). Our study aimed to examine adherence to National Guidelines in our clinic with regard to diagnosis and treatment of asthma. This was a retrospective chart review of consecutive patients seen for asthma between January to June 2013. Results: A total of 197 unique visits were reviewed. The majority of patients were female (78%) and the mean age was 50 years (standard deviation 13). Smoking history was identified in 42% of patients, and 55% of them were males. Only 22% of the notes recorded the categorization of asthma according to the severity of symptoms. Pulmonary function tests (PFTs) were documented in 30% of the patients. COPD was an accompanying diagnosis in 15% of the encounters, of these 76% had PFTs reported. Standardized clinical evaluation (e.g. weekly use of albuterol, recent hospitalizations and presence of night symptoms) were documented only in 26%, 17% and 11% of the clinic notes, respectively. Conclusion: It is evident that asthma is inadequately characterized in University Hospital primary care clinic. An implementation to improve classification of, and ultimately treat, patients is to develop a Smart Set in the electronic chart system that includes applicable questions. In addition, a brief lecture on the appropriate management of asthma will be given to all residents. Future studies include evaluating a larger number of patients and analyzing charts post-implementation of the Smart Set.
A Case of CMV Hepatitis in a Post-Liver Transplant Recipient presenting with Tacrolimus Neurotoxicity

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Introduction Two of the most common complications following orthotopic liver transplant are acute transplant rejection and opportunistic infections, most commonly viral. The most common viral infection to affect post-transplant recipients is CMV. CMV can cause direct effects such as bone marrow suppression or tissue invasion. It may also indirectly increase allograft rejection, recurrence of Hepatitis C, and opportunistic infections. We report a case of biopsy-proven CMV hepatitis in a post-liver transplant recipient. Case Presentation A 56 year old Caucasian female with a past medical history of orthotopic liver transplantation for HCV cirrhosis presented with a 1 week history of lower abdominal pain and a 1 day history of confusion and subsequent seizure. At the time of transplant, both the donor and recipient were CMV positive. At presentation, the patient was four months post liver transplant and was on dual immunosuppressant medications with tacrolimus and mycophenolate mofetil. Furthermore, she completed three months of CMV prophylaxis with valganciclovir post-transplant. Her tacrolimus dose was increased 1 week prior to presentation after she was found to be neutropenic. Upon initial evaluation, the patient was noted to have a general tonic-clonic seizure, with a CT Head showing posterior reversible encephalopathy syndrome. Her tacrolimus level was checked and found to be supratherapeutic. Her encephalopathy and subsequent seizure were attributed to tacrolimus toxicity. Later, her liver function tests were noted to be elevated, which were not explained by further abdominal imaging. She then had a bedside percutaneous liver biopsy done to rule out allograft rejection, but histology revealed changes consistent with acute CMV hepatitis, later confirmed with immunostain and serology. The patient was then started on Valganciclovir and discharged in stable condition. Discussion CMV is the most common viral pathogen to influence the outcome of orthotopic liver transplantation. It has many direct and indirect effects in liver transplant patients. In our case, the direct effect of tissue invasion leading to acute hepatitis was observed. A review by Lee SO et al. reports the incidence of CMV infection within 12 months of liver transplantation in a CMV-seropositive donor and recipient is 2.7% if CMV prophylaxis is given and 18.2% in patients that are not given CMV prophylaxis. In our case, both the donor and recipient were CMV-seropositive and the patient received CMV prophylaxis following liver transplantation. Therefore, her risk of having CMV hepatitis should have been minimal. The balance between adjusting immunosuppressant medications and risk of opportunistic infection should always be considered in transplant patients and something that physicians of these patients should be aware of.
Lung cancer usually presents with pulmonary symptoms, but it can have diverse and dramatic presentations, such as retrograde invasion of the mediastinal nodes to the pericardial plexus sac, resulting in cardiac tamponade. We present a case of cardiac tamponade as the initial presentation of lung cancer. An 85 year-old female was admitted for lethargy and dyspnea. She was hypotensive (82/61 mmHg) and had renal failure. She received 3 liters of normal saline with minimal response and was subsequently started on vasopressors. The echocardiogram revealed pericardial effusion with possible clots; the chest CT confirmed pericardial effusion, small pleural effusions and a 5 mm left lower lobe calcified granuloma. The patient deteriorated rapidly, requiring bedside pericardial window and intubation. Bacterial and viral cultures from the pericardial effusion were negative. Pericardial biopsy showed benign fibro-connective tissue with focal neutrophilic infiltration without malignant cells. Her condition improved over several days and she was discharged home. 29 days later the patient returned with shortness of breath and chest pain due to re-accumulation of the large pericardial effusion this time with right ventricular collapse. Chest CT showed moderate-size pericardial effusion, bilateral pleural effusions, extensive thoracic adenopathy and diffuse scattered pulmonary nodules, not previously seen in the first CT scan. The patient underwent pericardiectomy to prevent effusion re-accumulation. A second pericardial biopsy and fluid cytology showed metastatic adenocarcinoma consistent with lung origin. Later, brain MRI showed multiple supra- and infra-tentorial metastases measuring 4-5 mm. The patient deteriorated rapidly and required bilateral chest tubes. Her critical status precluded the possibility of systemic chemotherapy. Patient died 2 months after the initial pericardial effusion. Discussion: Malignant involvement of the pericardium is detected in 1 to 20 percent of cancer cases in autopsy studies. The most common metastatic tumors involving the pericardium are lung, breast and esophageal cancer. Newly found primary cancer on autopsy is responsible for 18% of the cases; however, the likelihood of diagnosing the primary malignancy pre-mortem is only 4-7%. The sensitivity of cytology for the diagnosis of a malignant effusion is 67-92 %, but a negative cytology does not exclude malignancy, particularly if the index of suspicion is high. This case is an example of an unusual initial presentation of lung adenocarcinoma with negative histology and no obvious cancer on chest CT during the first admission. Neoplastic pericardial effusion should be included in the differential diagnosis of cardiac tamponade. Patients whose first clinical presentation of malignancy is pericardial effusion usually have a short median survival (2-6 months). However, it is still necessary to search for occult malignancy in otherwise “healthy” patients with pericardial effusion, due to growing evidence that chemotherapy/radiation might prolong survival in some subgroups of patients, giving special value to an early diagnosis.
Rutgers- New Jersey Medical School

Gastric cancer: Clinical differences among Hispanic and non-Hispanic whites at the John Theurer Cancer Center (JTCC), Hackensack University Medical Center.

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Background: Higher gastric cancer-related mortality in Hispanic patients has been described in diverse studies but just a few have targeted the Hispanic population in New Jersey. We aimed to compare the clinicopathological differences at the time of diagnosis in Hispanics vs. non-Hispanic Whites with gastric cancer. Methods: Records of all the patients with gastric adenocarcinoma who visited JTCC from 09/01/2011 to 09/01/2013 were reviewed. Two ethnic-groups were formed: Hispanics and non-Hispanic Whites. Data was computed by age, sex, histologic type, anatomic site, biomarkers, previous GI pathologies, risk factors including: alcohol, tobacco, family history, and mortality rate among both groups. Tissue diagnosis and established tumor locations were required. Results: Case records of 56 eligible patients, consisting of 28 Hispanics and 28 non-Hispanic Whites were analyzed. The average age at the time of diagnosis was 54 among Hispanics and 66 in the non-Hispanic White patients. Among non-Hispanic Whites, 57% had a prior smoking history compared with 32% of Hispanic patients. Hispanics were more likely to have a stage IV, HER2 negative tumor at the time of diagnosis (57% vs. 41%) and less likely to have family history of gastric cancer. Non-Hispanic Whites were more likely to have a proximal tumor (50% vs. 25%), and less likely to have history of Helicobacter Pylori infection. During the 2 year period, 35% of Hispanic patients died within a 12-14 month average interval after the diagnosis, while 46% of non-Hispanic Whites died within an average of 18-20 months after the diagnosis. Conclusions: Hispanics were less likely to have family history of gastric cancer, but they were diagnosed at an earlier age and with advanced diseases stage. Overall survival in Hispanic patients with gastric cancer was shorter when compared with Non-Hispanic Whites. This warrants the need for more research in this topic, as this could potentially impact screening recommendations.
Ebstein’s Anomaly: Silent congenital defect raises havoc in an otherwise healthy man.

Jessica Riggs, MD, Suman Manchireddy, MD.

Introduction: Ebstein’s anomaly is a congenital malformation of the heart characterized primarily by abnormalities of the tricuspid valve and the right ventricle. Occurrence is roughly 1/200,000 births, with occasional cited risk being lithium use in the first trimester. In the adult population, emergence of this anomaly is extremely rare, as only 5% afflicted survive into the 5th decade of life. Recognition of symptoms, accurate diagnostics, and risk reduction, are essential in order to prevent morbidity and mortality. We present a case of a healthy adult without known cardiac history, who was found with atrialization of the right ventricle and a left-to-right shunt, consistent with Ebstein’s anomaly. Case: A 45 year old man presented to the ED with progressively worsening stabbing chest pain located substernally. He initially noted a nagging discomfort in the upper abdomen 1 month prior, which gradually spread up the chest and increased in severity daily. This man, who delivered food via bicycle and rode up to 12 miles/day, was incapacitated by his symptoms. Pain was worse with leaning forward, exertion, and deep inspiration, and was unaffected by eating, stress, or chest percussion. The patient indicated severe dyspnea on exertion and easy fatiguability, as well as transient palpitations. He otherwise denied associated symptoms. On cardiac exam, S1 and S2 were fixed and widely split, with a holosystolic 5/6 murmur with a palpable thrill, loudest in early systolic, with blowing at the left sternal border and more intense with inspiration. EKG showed a right bundle branch and 1st degree AV block, as well as right atrial enlargement. Labs including cardiac enzymes and ABG, were not indicative of any pertinent pathology, but chest xray did prove cardiomegaly without effusion or consolidation. Transesophageal echocardiography revealed atrialization of the right ventricle and markedly dilated right atrium and ventricle, severe tricuspid regurgitation, and pulmonary hypertension. Finally, a bubble-study unveiled an atrial septal defect with left-to-right flow, all agreeing with a final diagnosis of Ebstein’s anomaly and significant right heart failure. Scheduled for emergency surgery, however, the patient signed out against medical advice, and was lost to follow up. Discussion: New found congenital heart defects in the adult population, though rare, present unannounced with non-specific symptoms, and lead to devastating consequences if not identified immediately. In Ebstein’s anomaly, alteration of the ventricle permits conduction abnormalities, including Wolff-Parkinson-White syndrome, AV blocks, reentrant tachycardias, atrial fibrillation, and sudden cardiac death. 50% of patients also have septal defects, which predisposes to embolic events, seeded infections, and severely high pulmonary pressures. Transesophageal echocardiography is the gold standard of diagnosis. Treatment depends on severity of dysfunction, and ranges from afterload reduction, anti-coagulation, and pulmonary vasodilation, to necessitated surgical repair as seen in this case.
Thrombotic thrombocytopenic purpura: a hemolytic crisis, leaving a healthy woman on dialysis

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Introduction: Thrombotic thrombocytopenic purpura (TTP) is an acute attack on multiple organ systems, classically presenting with a subset of the cited pentad of thrombocytopenia, renal failure, microangiopathic hemolytic anemia, neurologic changes, and fever. It affects roughly 5 per one-million annually, and about 1/25,000 pregnancies. Presentation may be non-specific leading to misdiagnosis. Without immediate plasma exchange, adult mortality approaches 90%. We present a case of a healthy woman with significant mental decline and rapidly deteriorating nephropathy, with a diagnosis of TTP and irreversible renal failure. Case: The patient is a 62 year old Hispanic woman with past medical history of untreated hypertension, who presented from an outside hospital with suspected TTP, and need for imminent plasmapheresis. The patient was admitted with 10 days of abdominal pain, with workup and CT imaging showing free fluid consistent with a ruptured ovarian cyst, as well as active diverticulosis. There was rigidity and abdominal tenderness, but otherwise the physical exam was normal. On hospital day 2, however, the patient had a 5-minute tonic-clonic seizure and waning of mental status which continued to decline well beyond the post-ictal period. Head CT and MRI were negative for acute pathology, and a lumbar puncture ruled out infection or bleed. Patient was given Cerebyx and started on IV fluids. On day 3, reported more somnolent and difficult to arouse, and was found oliguric. Lab analysis revealed thrombocytopenia (94K), hemoglobin of 8 (baseline 11.5), elevated LDH and decreased haptoglobin, but no schistocytes on peripheral smear or fibrin degradation products indicative of DIC. Creatinine had risen to 5.4 (baseline 1.1), and potassium was 5.6. Emergent hemodialysis initiated, and transfer arranged to tertiary center for suspected TTP. ICU team commenced plasma exchange with fresh frozen plasma on arrival, with goal of reversal of microvascular thrombi formation and subsequent organ damage. But as often seen with this syndrome, this woman’s case did not follow a typical pattern. ADAMTS13 levels were normal, and though we did see neurologic and hematologic improvement after weeks of daily plasmapheresis and rituximab, renal function never returned. Kidney biopsy revealed only subclinical spotty glomerular microthrombi, unable to explain dysfunction. By Day 30, the patient regained full cognition, and would go home with plan for AV fistula and chronic hemodialysis. Discussion: TTP is a spontaneous systemic disorder due to endothelial injury from platelet-rich microthrombus deposition. Etiologies range from drug-induced (12%) to pregnancy related (7%) to complete idiopathy (40%). Diagnosis rests on clinical presentation plus supporting serum tests, and must be considered in every differential. Gold-standard treatment is immediate plasmapheresis, even when TTP diagnosis is uncertain. If untreated, there is high likelihood of extremely poor outcome including stroke, cardiac ischemia, and death. Steroids and immunosuppression are considered when plasmapheresis chronically fails.
Osmotic Demyelination Syndrome due to Rapid Correction of Hypernatremia

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Osmotic demyelination syndrome (ODS) is a rare condition usually associated with the rapid correction of hyponatremia. However, ODS has been sporadically reported in association to hypernatremia (only 33 cases reported in the literature since 1994). We present a case of ODS linked to correction of extreme hypernatremia in a patient with multiple risk factors for elevated serum sodium. A 59 year old female with a history of diabetes and HTN was transferred for confusion and burns on her feet that were debrided and dressed while abroad. She had been vacationing in the Dominican Republic before admission to a local hospital for acute confusion. Initial labs at the Dominican facility showed extreme hypernatremia (202 mEq/L), hyperglycemia (624 mg/dL), azotemia, elevated lipase, amylase and CPK, and she received fluids, insulin, bicarbonate, and Lasix. Her sodium was corrected by 12meq/L on day one, by 13meq/L on day two, and by 15meq/L on day three to 158 meq/L. Her mentation did not improve, so she was transferred to our hospital. On admission her sodium was 167mEq/L, CK was 7012IU/L, and glucose was 274mg/dL with elevated anion gap and ketones on urinalysis. CT of the head was normal. She was treated for DKA and acute kidney injury. Her elevated CPK was thought to be secondary to her burns. Further workup was negative; her elevated sodium seemed due to multiple unrelated metabolic derangements. Despite continued correction of her sodium at a rate of 8meq/L/24hrs, her mentation didn’t improve. A brain MRI showed central and extra-pontine demyelination consistent with ODS. The pathophysiology of ODS due to hypernatremia is not well understood. The inciting event is thought to be either aggressive correction of sodium or merely the acute rise in sodium by more than 20meq/L/24 hrs. Our patient’s extreme rise in serum sodium was likely secondary to free water loss from her burns, osmotic diuresis from hyperglycemia, an inability to ask for water due to confusion and diuretic administration. The ODS that resulted could have been due to the acute rise in her serum sodium; however, aggressive correction could have played a role. The suggested rate of correction for acute hypernatremia is 8-12mEq/L/d, but this does not necessarily prevent ODS. There are reports of more aggressive correction of serum sodium without neurologic sequelae. Moreover, in one review article the patient outcomes did not correlate with sodium level at presentation, severity of symptoms, or rate of correction. Given the high overall incidence of hypernatremia seen in adult hospitalized patients (0.3-5.5%), and its association with high mortality rate (30-48% in ICU patients with a serum sodium over 150meq/L), prompt recognition and adequate treatment is therefore essential to prevent long term disability due to ODS.
“Milk in the Belly? Chylous Ascites: An Atypical Manifestation of Cirrhosis”

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Introduction: Chylous Ascites, or chyloperitoneum, is the accumulation of milky appearing triglyceride-rich peritoneal fluid, which can result from multiple etiologies including post-operative complications, lymphatic obstructions, lymphoma, solid tumors, and mycobacterial infections. Case Presentation: A 57 year old cachectic Caucasian man with Hepatitis B, Hepatitis C, and chronic alcohol dependence with cirrhosis presented with progressive diffuse abdominal pain and distention over a five month course, which was accompanied by a 90lb weight loss and fatigue. On presentation, physical exam revealed a hemodynamically stable, however chronically-ill appearing man with bi-temporal wasting, cachexia, and abdominal distention accompanied by shifting dullness and fluid wave. Additional findings included significant hepatojugular reflex, caput medusa, thenar wasting, gynecomastia, easy bruising, and digital clubbing. Despite stigmata for severe liver disease, initial laboratory studies were only significant for serum albumin level of 2.8g/dl however a MELD score of 7 was calculated. Computed tomography of the abdomen and pelvis revealed a heterogeneously enhancing nodular liver along with massive ascites, with no evidence of malignant mass or adenopathy. Due to suspicion of spontaneous bacterial peritonitis, a paracentesis was performed which yielded 4L of milky-appearing triglyceride-rich fluid. Lab analysis of abdominal fluid revealed a triglyceride level of 329, albumin of less than 1, and leukocyte count of 33.7, inconsistent with peritonitis. Fluid cytology was negative for malignancy and serum AFP was determined to be within normal limits. Pain and abdominal distension improved after paracentesis, aggressive diuresis with furosemide and spironolactone, and water restriction of 1 to 1.5 L per day. Intravenous antibiotics which were initially started for empiric coverage of spontaneous bacterial peritonitis were stopped. Transthoracic echocardiogram was obtained to rule out right heart failure, which revealed no diastolic or systolic dysfunction. An abdominal ultrasound with doppler displayed no signs of hepatic vein thrombosis, and PPD testing was negative for tuberculosis. Further evaluation with esophagogastroduodenoscopy revealed columnar metaplasia of the esophagus consistent with Barrett’s Esophagus, along with esophageal varices, and portal hypertensive gastropathy. Colonoscopy was also performed which was significant for a 3cm rectal mass, suspicious for malignancy, with biopsy specimen sent for pathologic diagnosis. Discussion: Chylous ascites is a rare manifestation of cirrhosis, and its finding can be associated with other etiologies including malignancy, lymphoma, and mycobacterial infections. Spontaneously occurring chylous ascites in the absence of portal hypertension or iatrogenic causes is rare, nonetheless presents a diagnostic dilemma for clinicians. While cirrhosis may contribute up to 11% of cases of non-traumatic chylous peritoneal fluid, other causes such as mycobacterium infection and malignancy should be ruled out in patients presenting with chylous ascites. Accurate diagnosis of etiology of chylous ascites is necessary to guide effective treatment.
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Methanol accounts for 1% of poisonings in US, with 1000-2000 ingestions annually. The mortality ranges from 8-36%, but can be up to 80% with severe acidosis. This infrequent, potentially fatal ingestion requires immediate identification and treatment to prevent permanent vision loss and death. 42-year old man presented to the emergency department 12 hours after ingesting an unknown cleaning agent in a suicide attempt. After ingestion, he went to sleep and awoke with severe generalized body pain, dyspnea and complete vision loss. In the emergency department, he rapidly developed respiratory failure requiring intubation. Physical exam revealed tachypnea, tachycardia and hyperemic fundi. Arterial blood gas demonstrated severe metabolic acidosis- pH- 7.027, CO2- 9, PO2- 51, HCO3- 6. Concurrent chemistry revealed a bicarbonate- 4, anion gap- 32, measured serum osmolality- 342, osmolal gap- 60. He received 2 ampules of sodium bicarbonate and fomepizole was ordered. Nephrology was consulted for suspicion of methanol toxicity and emergent hemodialysis was arranged. He received 4 hours of hemodialysis and two doses of fomepizole 12 hours apart. He clinically improved and was extubated the next day with resolution of all symptoms except vision loss, subsequent visual field testing documented 20/400 bilaterally. His methanol level from admission later returned as 85 mg/dL. In this case, fomepizole and emergent dialysis were indicated based on the strong clinical suspicion of methanol overdose with severe acidosis, and an elevated anion and osmolal gap. Toxicity from methanol is not from the methanol itself, but the toxic metabolite, formate (formic acid). Formate is toxic to the optic disk leading to permanent injury in up to 30% of cases, therefore a fundoscopic exam is imperative. Initially after methanol ingestion, the alcohol causes an elevated osmolal gap. As the methanol is metabolized, the osmolal gap decreases and the anion gap rises due to formate accumulation. Depending on the time of presentation, the serum osmolality may be at or close to normal. This may confuse the clinical picture and result in a delay in diagnosis. In cases of known or suspected methanol ingestion communication with the poison center is essential. Treatment includes sodium bicarbonate, fomepizole and possibly hemodialysis. Indications for treatment include: methanol >20 mg/dl, osmolar gap >10 or strong clinical suspicion with pH <7.3, bicarbonate <20, osmolal gap >20. Fomepizole inhibits the oxidation of methanol into formate via alcohol dehydrogenase. Hemodialysis removes methanol and formate, and is indicated in cases of severe metabolic acidosis, end organ damage or methanol level >50mg/dl. As our case highlights, it’s important to institute immediate treatment if there is strong clinical suspicion as confirmatory methanol levels may take several days to return.
Evaluation of Clinician’s Compliance with ACG Guideline in Diagnosis of Acute Pancreatitis: A Comparison between The University Hospital and Hackensack University Medical Center

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Background: ACG Diagnostic Guidelines require two of three criteria: 1) characteristic abdominal pain; 2) amylase and/or lipase greater than three times of upper limit; 3) characteristic findings of acute pancreatitis on abdominal CT scan to diagnose acute pancreatitis. An abdominal CT scan is not always necessary for diagnosis in certain clinical situation. Purpose: This study is to evaluate whether clinicians are using guidelines suggested by ACG to make the most appropriate imaging decisions for acute pancreatitis. A comparison is also made between a State-managed inner-city hospital and a privately-managed suburban hospital to assess associated factors of compliance. Methods: This is a retrospective cohort study. Out of the hospitalized patients diagnosed with acute pancreatitis from 01/2010 to 01/2011, a group of 50 were randomly chosen at Hackensack University Medical Center (HUMC), NJ and a group of 64 was chosen at The University Hospital in Newark, NJ (UH). Each case was categorized into one of four clinical situations based on whether a CT scan was indicated by the ACG guidelines. Situation #1 does not require abdominal CT for diagnosis. Situations #2 to #4 are indicated for certain imaging study. Compliance with the guidelines was assessed using the Z-ratio for the significance of the difference between two hospitals. Results: For the cohort at HUMC, 49% presented as situation #1; 16% as situation #2; no cases as situation #3 and 35% as situation #4. For the cohort at UH, 64% presented as situation #1; 9% as situation #2; 24% as situation #3 and 3% as situation #4. Situation #1 was the most frequently encountered presentation. It was found that overall compliance was 42% at HUMC vs 61% at UH (p=0.03). For situation #1, the compliance was 19% at HUMC vs 44% at UH (p=0.02). For situation #2, compliance was 89% at HUMC vs 83% at UH, but was not statistically significant. For situations #3 and #4, the small number of cases precluded statistical analysis. Conclusion: The overall compliance for the ACG guidelines was found to be very poor, which are less than 60% for both hospitals. In Situation #1, (the most frequently encountered presentation of acute pancreatitis), compliance was even worse, at less than 44% for both sites. Clinicians did better in the less frequently encountered Situation #2, with compliance for both sites at 83-89%. Situation #3 and #4 are rarely encountered clinical presentations. Since the major difference between Situations #1 and #2 is whether an abdominal CT scan should be ordered on initial presentation, our study results indicate that clinicians often order unnecessary CT scans despite ACG guideline. It seems, however, that compliance with the guidelines was better at the State-run hospital than the private one.
Decompensated heart failure due to cardiac non-compaction associated with myocarditis

Mahaa Umapathi, Adam Raskin, Pallavi Solanki

Left ventricular non compaction (LVNC) is defined as a spongy, trabeculated appearing myocardium which results from failure of normal myocardial compaction during embryogenesis. This clinical condition has only recently been described (1990), and can often be misdiagnosed without confirmatory testing. A 51 year old female with no significant past medical history presented with shortness of breath, orthopnea and decreased exercise tolerance for 2 weeks. She denied cough, fevers, chills, chest pain, palpitations, or lower extremity edema. Transthoracic echocardiogram was consistent with severely depressed left ventricular systolic function (LVEF <15%), global hypo kinesis and LV non-compaction. A cardiac MRI was performed to confirm the diagnosis and was significant for increased trabeculation of the LV myocardium measuring 22 mm in thickness. Additionally, there was abnormal delayed enhancement along the inferior wall and septum, consistent with myocarditis. A myocardial biopsy was performed with findings consistent with lymphocytic infiltration and myonecrosis. The patient was started on a heart failure regimen that included a beta-blocker, ACE-inhibitor and aldosterone blockade. Due to an episode of sustained ventricular tachycardia, an intracardiac defibrillator was implanted for secondary prevention. This case illustrates a clinical presentation of acute decompensated heart failure due to severely reduced systolic function caused by 2 concomitant etiologies. The echocardiogram performed during her hospital course initially seemed clear that isolated LV noncompaction had resulted in the patient’s heart failure syndrome. While the cardiac MRI confirmed LV non-compaction, it was also consistent with myocarditis, which was later confirmed by biopsy. The combination of noncompaction and myocarditis is an uncommon finding; our case suggests that in the appropriate context one should consider investigating further to rule out alternative/coexisting causes for decompensated heart failure.
The unexpandable lung-lung entrapment re-visited

Mahaa Umapathi, Andrea Isaacs, MD

The unexpandable lung is a clinical entity encountered frequently in pulmonary medicine. Etiologies for impairment of complete expansion of the lung can be connected to endobronchial obstruction, pleural disease or chronic atelectasis. Clinical symptoms may range from dyspnea and chest pain during therapeutic thoracentesis, to the development of pneumothorax post-pleural fluid removal. The diagnostic challenge of differentiating lung entrapment from trapped lung becomes essential for appropriate treatment. A 72-year-old woman with a history of COPD, hypertension and hyperlipidemia presented post syncope and confusion for one day. She had experienced increasing fatigue, lethargy for the prior 2-3 weeks and decreased exercise tolerance secondary to shortness of breath. On hospital admission she was noted to be acidemic, hypercapneic and hypoxic with a large, loculated effusion and accompanying lung collapse on her left side. She was started on empiric antibiotics and steroids for clinical diagnoses of COPD exacerbation and community-acquired pneumonia. An echocardiogram was significant for left ventricular ejection fraction of 70% with severe pulmonary hypertension. She underwent bilateral video assisted thoracoscopy with wedge resection, drainage of effusion and left sided pleurodesis with doxycycline and chest tube placement. A post procedure CT scan of the chest showed bilateral pneumothoraces despite proper chest tube placement and no interval expansion of the lung over her hospital course despite complete removal of her effusion. Cytological studies of the fluid were consistent with lung adenocarcinoma. Despite appropriate positioning of the chest tubes lung re-expansion did not occur. Her respiratory status continued to decline and given her clinical prognosis the patient and her family agreed with the institution of comfort measures. This case illustrates the clinical relevance of differentiating trapped lung and lung entrapment. The trapped lung occurs due to defective pleural healing in setting of remote inflammation, which results in a visceral, fibrous peel that mechanically prevents the expansion of the lung. Lung entrapment is due to active pleural disease. Ascertainment of a diagnosis of lung entrapment is useful in preventing procedures such as pleurodesis, which is invasive and relatively unsuccessful in these cases. Many patients with trapped lung are asymptomatic or have minimal dyspnea on exertion. If there is persistence of pleural effusion, repeated thoracenteses are not recommended and in patients with incapacitating dyspnea pleural decortication is an option. The literature also argues for attempting lung re-expansion with thoracostomy tubes in poor surgical candidates.
The incidence of digitalis toxicity has declined in recent years due to decreased use of this drug along with improved laboratory monitoring and awareness of contraindications. However clinicians need to remain sensitive to the polymorphic presentation of digoxin toxicity. Maintaining a high index of suspicion is essential given the almost exclusive clinical nature of diagnosing the condition, which can be fatal in the absence of treatment. A 50 year old male with history of non-ischemic cardiomyopathy, atrial fibrillation, stroke, hypothyroidism and hypertension presented with a 3 day history of nausea and vomiting associated with blurry vision, mild diarrhea and transitory paresthesia of the fingertips. He also complained of non radiating, non exertional chest pain worsened with inspiration. His homedications included digoxin 0.25 mg daily, tramadol 50 mg every 6 hours as needed, lisinopril 20 mg daily, simvastatin 40 mg daily, carvedilol 25 mg twice daily and warfarin 5 mg daily. He had been on a stable dose of digoxin for over a month and denied taking more than his prescribed dose. On presentation to the ED his vital signs were normal (BP 134/94, pulse 71, temperature 98.8 F, respiratory rate of 18 and oxygen saturation of 100% on room air). Physical exam was remarkable for mild epigastric tenderness. ECG demonstrated atrial fibrillation and signs of early repolarization. Compared to prior ECG the only difference was the ventricular rate decrease from 109 bpm. Lab results revealed normal electrolytes, acute renal failure (creatinine 2.1 from baseline 1.2 to 1.4), serum digoxin level of 4.5 ng/ml and INR of 3.9 with normal liver function tests. The patient was administered IV fluids and a weight based dose of digibind. He underwent successful direct current cardioversion and was placed on amiodarone. After therapy all symptoms resolved. Kidney ultrasound revealed possible renal artery stenosis. Subsequent MRA demonstrated 90% proximal right renal artery stenosis. The patient underwent successful angioplasty and stent placement in the right renal artery. Digoxin is the only cardiac glycoside commercially available in the US and is still prescribed regularly in the management of atrial fibrillation and heart failure. The clinical utility of this drug has been declining given its narrow therapeutic window and the possibility of multiple drug interactions leading to serious toxicity. In 2008 the US poison control centers were called for 2632 cases involving digoxin toxicity and 17 cases resulting in death. We report a case of symptomatic digoxin toxicity in the setting of acute renal failure. Administration of digoxin antibodies results in improved symptoms. The case re-iterates the sole clinical nature of diagnosing digoxin toxicity and highlights the narrow therapeutic index of the drug especially in the setting of acute renal failure.
A Case of Type A Aortic Dissection with Intimal Flap Prolapse into Left Ventricular Outflow Tract Presenting as Left Main ST Segment Elevation Myocardial Infarction

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Acute aortic dissections are life-threatening emergencies requiring appropriate diagnosis and urgent management. Although extension of aortic dissections to coronary ostia are often described in literature, the right coronary ostium is much more commonly affected than the left, especially with presentation of symptoms similar to those of a myocardial infarction. A 66 year-old man presented with sudden-onset substernal chest pain that radiated to his neck and left arm and was associated with vomiting and diaphoresis. He was found to be hypotensive, and the electrocardiogram revealed diffuse ST depressions in all precordial and limb leads, with ST segment elevation in aVR. On suspicion of a left main coronary artery myocardial infarction causing cardiogenic shock, he was taken for immediate cardiac catheterization and started on inotropic support. An aortogram performed after unsuccessful attempts to engage the left main coronary artery revealed a dilated ascending aortic root, severe aortic regurgitation, and a right-sided dissection plane. Transthoracic echocardiography showed an ascending aortic dissection with intimal flap prolapse through the aortic valve and into the left ventricular outflow tract. He was then taken to the operating room for emergent dissection repair. The dissection was found to have involved the entire circumference of the aorta, extending into the proximal portions of the left and right main coronary artery ostia. Aortic root and valve replacement with coronary re-implantation and distal anastomoses were performed. This case presents a rare mechanism of myocardial infarction secondary to obstruction of the left ventricular outflow tract by an intimal flap. Additionally, there was unusual involvement of both coronary arteries in this Stanford Type A aortic dissection. Inclusion of aortic dissection in the differential diagnosis for myocardial infarction underscores the importance of determining an efficient method to eliminate this possible cause, especially when considering the use of thrombolytic agents.
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Outcomes after Surgery and Radiotherapy for Spinal Myxopapillary Ependymoma

Yucai Wang, Chiaojung Jillian Tsai, Pamela K. Allen, Paul D. Brown, Jing Li

Outcomes after Surgery and Radiotherapy for Spinal Myxopapillary Ependymoma Yucai Wang,1,2 Chiaojung Jillian Tsai,2 Pamela K. Allen,2 Paul D. Brown,2 Jing Li2 1Department of Medicine, Rutgers New Jersey Medical School, Newark, New Jersey; 2Division of Radiation Oncology, The University of Texas MD Anderson Cancer Center, Houston, Texas Objective: To evaluate long-term outcomes after surgery and radiotherapy (RT) for spinal myxopapillary ependymoma (MPE). Methods: Fifty-one patients with spinal MPE treated at The University of Texas MD Anderson Cancer Center in 1968-2007 were included. Overall survival (OS), progression-free survival (PFS), and local control (LC) rates were calculated by Kaplan-Meier estimates. Clinical and treatment variables (age at diagnosis, sex, tumor location, symptom lateralization, initial neurologic function, primary treatment modality, extent of surgical resection) were analyzed for prognostic value with log-rank tests and Cox regression analysis. Statistical analysis was performed using SPSS. All tests were two-sided, and P ≤ 0.05 was considered statistically significant. Results: Median age at diagnosis was 35 years (range 8-63). Thirty-two patients were male and 19 female. Tumors were in the thoracolumbar cord in 14 patients (27%) and lumbosacral cord/cauda equina in 37 patients (73%). Primary treatment was surgery in 20 patients (39%), surgery plus RT in 30 patients (59%), and RT alone in 1 patient (2%). Of the 50 patients who had surgery, 28 (56%) had gross total resection, 18 (36%) subtotal resection, and 1 (2%) biopsy only. At a median follow-up time of 11 years (range 0.2-37), 10-year OS, PFS, and LC rates were 93%, 63%, and 67%, respectively. Nineteen patients (37%) had disease recurrence, all in the neural axis. In multivariate analyses adjusting for resection type, age > 35 at diagnosis and receipt of adjuvant radiation were associated with improved PFS (HR = 0.14 [95% CI = 0.04-0.51, P = 0.003] and HR = 0.45 [95% CI = 0.25-0.79, P = 0.009], respectively), and improved LC (HR = 0.22 [95% CI = 0.06-0.80, P = 0.02] and HR = 0.45 [95% CI = 0.24-0.82, P = 0.009], respectively). Among 28 patients treated with gross total resection, 10-year LC rates were 56% after surgery alone versus 92% after surgery plus radiation (P = 0.14); median time of LC was 4.75 years for patients receiving resection only and 10.5 years for patients receiving resection plus RT (P = 0.03). Among 16 patients treated with subtotal resection with sufficient follow-up data, 10-year LC rates were 0% after surgery alone versus 65% after surgery plus radiation (P = 0.008). Conclusions: Patients with spinal MPE had favorable long-term outcomes after surgery and radiotherapy. Age > 35 at diagnosis and use of adjuvant radiation conferred better PFS and LC. Surgery and adjuvant radiotherapy should remain as primary treatment modalities for spinal MPE.
Myoclonus in Renal failure: Two Cases on Gabapentin Toxicity

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Antiepileptic drugs (AEDs) are used in the treatment of epilepsy, pain, and psychiatric disorders. Renal status may impact the efficacy and toxicity associated with AEDs which requires awareness by clinicians in multiple specialties. Gabapentin is cleared solely by renal excretion and dosing requires consideration of the patient’s renal function. We report 2 cases of myoclonic activity associated with gabapentin toxicity in the setting of renal disease and address treatment with dialysis.

Case 1: 78-year-old woman with congestive heart failure, history of thromboembolism, hypertension, diabetes mellitus, asthma and diabetic peripheral neuropathy presented with tremors involving her upper extremities for 3 days prior to admission. Evaluation revealed acute kidney injury (AKI) secondary to increased furosemide and lisinopril with hyperkalemia and azotemia. The patient was noted to have severe myoclonus. Prior to admission, the patient was treated with gabapentin 900 mg total daily dose for neuropathic pain. The patient had no history of renal disease, but presented with an estimated glomerular filtration rate of 13 mL/min/1.73 m2. With discontinuation of gabapentin and initiation of hemodialysis (HD), marked improvement in her myoclonus occurred. Patient received 2 session of HD, and was discharged with normal renal function and resolved myoclonus. Gabapentin was held on discharge.

Case 2: 55-year-old man with end-stage renal disease on peritoneal dialysis (PD), diabetes mellitus, hypertension, neuropathic pain, and peripheral vascular disease with toe gangrene on long term antibiotics presented for evaluation of diffuse body tremors, altered mental status and worsening leg infection. Gabapentin 600 mg total daily dose was initiated for neuropathic pain 3 days prior to presentation with myoclonus. The patient’s PD treatment was increased from 4 to 6 exchanges daily. With increased dialysis and discontinuation of gabapentin, myoclonus resolved.

Myoclonic activity may occur as a complication of gabapentin toxicity, especially in the setting of renal dysfunction. In the cases reported, both HD and PD were effective in treating myoclonic activity in acute and chronic renal dysfunction. Gabapentin requires renal dosing in patients with chronic kidney disease and in patients and in patients at risk for developing AKI. As gabapentin has multiple indications, an understanding of such renal dosing is important to clinicians in multiple specialties.
Inter-hospital Transfers of Uninsured Patients: the Potential for a Medical Limbo

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Background: Insurance coverage is an important contributor to health disparities in the United States. Uninsured patients are less likely to be hospitalized, receive less intense care overall, and have increased mortality among the critically ill. This occurs despite the EMTALA act, which mandates patient stabilization and treatment regardless of a patient’s insurance coverage and is designed to prevent premature transfer of patients between facilities for financial reasons. The EMTALA act is balanced by recent privatization of medical care, which may increase incentive to transfer patients without insurance to other facilities as early as possible. This study aims to determine to what degree insurance status impacts inter-hospital transfers and outcomes in the health care climate.

Methods: We conducted a retrospective observational trial, the Inter-Hospital Transfer study (IHTS), wherein we evaluated consecutive patient transfers from outside hospitals to Robert Wood Johnson’s (RWJ) three intensive care units (medical, surgical, and cardiac) during December 2011 to December 2012. We compared insurance status against the population of patients admitted to ICU from the wards or emergency department. Outcomes included in-hospital mortality, adverse events, and adjusted for severity of illness by MPM0-III. Outcomes and insurance status were compared by two tailed ANOVA, t-test, and Fisher’s exact test, with statistical significance inferred by p<0.05.

Results: 328 patients were transferred to RWJ three ICUs from 37 outside facilities between December 2011 and December 2012 versus 3706 patients admitted from the wards or emergency department during that same time period. Self-pay or charity care patients were underrepresented in the inter-hospital transfer group when compared with those transferred from the wards or emergency department (8% versus 14% of transfers, p=0.005). Patients with private insurance were conversely overrepresented in the transferred patients (p=0.06). This difference was not explained by differences in race, age, gender, or regional variations in insurance rates. Of the cohort transferred from outside hospitals, uninsured patients had a reduced length of stay at the prior hospital (P<0.0001, 95% CI -2.786 to -1.189) and had a reduced severity of illness (p<0.0001) when compared against patients with insurance. Severity adjusted outcomes including in-hospital mortality, adverse events, length of stay, and over-utilization were similar between the groups.

Discussion: These data illustrate the complex effect of insurance status on a patient’s likelihood of being transferred between facilities. Patients without insurance are transferred earlier and are of lower acuity than their insured counterparts, which likely reflects a motivation to move patients who cannot pay to other hospitals. On the other hand, the under-representation of uninsured patients who are actually transferred is likely based on the lack of desire of hospitals to accept the uninsured. This creates the potential for a medical “limbo,” which would exacerbate health disparities in underserved populations.
TUMOR-INDUCED OSTEOMALACIA CAUSED BY A FGF-23 PRODUCING MESENCHYMAL TUMOR

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Introduction: Tumor-induced osteomalacia (TIO) is a rare and reversible form of osteomalacia and hypophosphatemia caused by overproduction of Fibroblast Growth Factor 23 (FGF-23). We present a case of TIO caused by a FGF-23 producing mesenchymal tumor, where surgical resection of this tumor corrected the patient’s osteomalacia and hypophosphatemia. Case Presentation: A 52 year-old female was in good health until after her first pregnancy in 1986, when she developed muscle cramps with exercise. Over a few years, these cramps became debilitating myalgias, and she developed a waddling gait. Initial labs showed chronic hypophosphatemia 1.2-1.9 mg/dL, mild hypocalcemia 8.4-9.2 mg/dL, hyperphosphaturia 1506 mg/24 hr (normal 400-1300), and normal levels of 25-OH Vitamin D 37 pg/mL and 1, 25-OH Vitamin D 60 pg/mL. N-terminal PTH was 13 pg/mL (normal 4-19), mid-molecule PTH was 379 pg/mL (normal 50-330), and no glycosuria nor amino aciduria were noted. Large doses of neotrofos (1.5-4 g/day) and calcitriol were initiated. Transiliac crest bone biopsy findings were consistent with severe osteomalacia, at which time she was diagnosed with vitamin D resistant hypophosphatemic osteomalacia. She continued to have symptoms of bone pain, myalgias, fatigue, depression, with elevated creatinine 1.2-2.8 mg/dL, and remained hypophosphatemic 1.6-2.3 mg/dL despite daily phosphate and calcitriol replacement. About 20 years after initial presentation, she noted a mass on her forearm. In November 2009, she underwent resection of the mass with pathology showing phosphaturic mesenchymal tumor (4.5x3.5x2 cm), mixed connective tissue variant. Subsequently, her phosphate levels gradually increased to 3.2 mg/dL, creatinine improved to 1.5 mg/dL, and urine phosphate normalized to 948 mg/24 hr. FGF-23 was measured to be 157-182 RU/mL (normal <180), however there were no preoperative values for comparison. In March 2011, she was titrated off all phosphate supplements and phosphate levels have remained normal. Discussion: TIO is a rare acquired disorder characterized by osteomalacia and severe hypophosphatemia. In this recently described disease, mesenchymal tumors produce FGF-23, a phosphaturic hormone which inhibits renal phosphate reabsorption and reduces renal 1,25-dihydroxycholecalciferol production, resulting in hypophosphatemia, phosphaturia and eventual osteomalacia. The pathways involved and clinical presentations are similar to the inherited forms of vitamin D resistant rickets, which include X-linked hypophosphatemic rickets (XLH) and autosomal dominant hypophosphatemic rickets (ADHR). In all of these conditions, phosphate and calcitriol replacement are the cornerstone of treatment. However, unlike XLH and ADHR, TIO is acquired and can potentially be reversed once the FGF-23 producing tumor is localized and resected. In our case of TIO, her hypophosphatemia and osteomalacia completely resolved with tumor resection. Interestingly, 20 years have elapsed between the initial presentation and the discovery of this rare FGF-23 producing mesenchymal tumor.
We describe a unique case of lymphoma presenting with bilateral adrenal masses, hypercalcemia, and non-oliguric renal failure secondary to lymphomatous renal infiltration. The patient is a 71 year old female with a past medical history of aortic stenosis (valve area of 0.7cm²), osteoporosis with vertebral compression fractures, hypertension, and hyperlipidemia who presented with one week of progressive generalized weakness and fatigue. The patient was normotensive with a BMI of 51.9 and had an exam remarkable only for a III/VI holosystolic murmur radiating to the carotid arteries and trace lower extremity edema. She was found to have non-oliguric acute kidney injury (AKI) (creatinine of 1.8 mg/dL; GFR 28 mL/min) and hypercalcemia (corrected calcium of 11.1 mg/dL). Patient was initially treated as AKI secondary to prerenal and give aggressive fluid hydration. On the second day of admission the patient developed severe left sided back pain. Imaging studies showed large bilateral adrenal masses and a small left perinephric subcapsular hematoma; kidney size and architecture was normal without evidence of obstructive uropathy. She became progressively uremic over the next several days, necessitating dialysis on hospital day 10. Extensive non-invasive evaluation during this time was non-diagnostic, including an essentially bland urinalysis, non-nephrotic range urine, with a spot protein-to-creatinine ratio of 0.67gram/24hours, several small restriction bands on immunofixation, and elevated serum beta-2 microglobulin & kappa free light-chains. Evaluation of her adrenal masses for neuroendocrine activity was negative. Additionally, hypercalcemia investigation was non-diagnostic with a suppressed parathyroid hormone level, normal 1,25-OH vitamin D level, and negative parathyroid hormone-related peptide. Kidney core biopsies were obtained and demonstrated diffuse infiltrative B-cell lymphoma with minimal normal kidney architecture remaining. Renal or adrenal involvement is a common occurrence in several types of advanced lymphomas. A few of these patients go on to develop acute kidney injury and only rarely is lymphomatous renal infiltration the underlying cause. There have only been 29 reported cases of patients initially presenting with AKI secondary to lymphomatous renal infiltration. A search of the literature reveals that all of these cases reported an active urine sediment, diffuse kidney enlargement, or multiple kidney masses. All of these findings were absent in this case. This case underscores the diagnostic value of early renal biopsy when conservative evaluation and treatment of renal failure is unsuccessful. Additionally, it demonstrates that lymphomatous renal infiltration can counter-intuitively present with a combination of non-oliguric acute progressive renal failure and bland urinalysis.
Oh Crap-A Case of Enteric Fever

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Introduction Fever in the returning traveler is a common clinical problem faced by general internists. Gastrointestinal symptoms often coincide with fever in these scenarios. Many times illnesses acquired overseas are self-limited and require supportive care only(1). However, failure to recognize the distinguishing features of more sinister diseases will often result in poor clinical outcomes for a subset of patients. We present a case of enteric fever in a traveler returning from India. Case A 47 year old female with no medical history presented to her doctor’s office with profuse diarrhea, intermittent fevers and chills, weakness, and dehydration for 3 weeks. Approximately 4 weeks prior to presentation the patient traveled to Delhi, India with no pre-travel care. A week later she began to have profuse watery diarrhea, intermittent chills, and then began to develop abdominal pain over a 2 week period. Chills became more frequent the week prior to presentation and the patient became weaker, nearly collapsing at work leading her to present for medical care. On presentation, she appeared ill and dehydrated with diffuse abdominal tenderness. Vital signs were temperature 101F, pulse 81, and blood pressure 119/55. She was hospitalized and cultures of blood and stool were sent as well as ova and parasite examination, Entamoeba serologies, and C. difficile PCR. CBC was unremarkable, BMP showed non-gap metabolic acidosis, and CT showed only diffuse small bowel enteritis. Empiric ciprofloxacin and metronidazole were initiated with aggressive hydration. Gram negative rods were isolated from the blood on hospital day 2, with S. typhi isolated in the stool on day 3. Entamoeba serology and C. difficile PCR were negative. Due to quinolone resistance in Delhi and her lack of improvement, ceftriaxone was added and metronidazole discontinued. She defervesced and was discharged on oral ciprofloxacin to complete a 14 day course after culture showed quinolone sensitivity. She re-presented with fever of 104 and chills on day 9 of antibiotics from initial presentation and gram negative rods were again isolated from the blood with quinolone sensitivity. Both isolates (initial and subsequent) were resistant to nalidixic acid, which can predict quinolone resistance(2). She then completed 21 day course of ceftriaxone therapy and is relapse free. Discussion While most diarrheal illnesses in a returning traveler are self-limiting and don’t require antimicrobials, the presence of fever and rigors along with prolonged course should prompt further investigation(1). Once a diagnosis of typhoid is made, it is imperative to prevent relapse by choosing appropriate therapy. Nalidixic acid resistance can predict quinolone failure(2) and should prompt choosing an alternate antimicrobial. This case emphasizes the need for communication between clinicians and the microbiological lab as well as for internists in the community to recognize signs of serious travel-acquired illnesses that require treatment.
Use of FFP in patients with mildly elevated INR undergoing invasive procedures

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Background Fresh frozen plasma (FFP) is frequently transfused to patients with mild coagulation abnormalities, but there is limited data documenting its efficacy in correcting INR or reducing bleeding complications. Methods We reviewed the medical records of consecutive inpatients with pre-procedure INR between 1.3 and 1.9 who underwent low-risk procedures and received FFP transfusion within a 12-month period. We recorded for each patient the age, gender, procedure performed, INR, hemoglobin level, and amount of FFP, and red blood cells transfused. Results Of the 38 patients included in our analysis, 32% (n=12) received FFP transfusion for central line insertion. Other common procedures included paracentesis (n=7, 18%), thoracentesis (n=4, 11%), lumbar puncture (n=4, 11%), and closed liver biopsy (n=4, 11%). A median of 2 units of FFP were transfused for a mean pre-procedure INR of 1.65. The post-procedure mean INR fell by 0.11 to 1.54. Ten (26%) patients also received a median of 1 unit of red blood cells. Corrected for amount of red blood cell transfusions, 55% of patients (21) had a decrease in hemoglobin level of 0.5 mg/dL or less, and 71% (27) had a decrease of 1.0 g/dL or less. Mean fall in hemoglobin level was 1.0 g/dL. Conclusion Transfusion of FFP for mild coagulation abnormalities does not appear to affect INR in a clinically significant manner although there was modest fall in hemoglobin level. Clinical trials are needed to definitively evaluate the efficacy of FFP.
Rutgers- RWJMS

Acute Hepatotoxicity from Sitagliptin

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Sitagliptin belongs to a class of oral hypoglycemic agents known as dipeptidyl peptidase-4 (DPP-4) inhibitors. They work by increasing activity of glucagon-like peptide 1, a gut hormone that increases glucose dependent insulin secretion thus improving glycemic control. Clinical trials have proven its efficacy in the treatment of type 2 diabetes mellitus (T2DM). It is a very well tolerated medication with minimal adverse effects and drug-drug interactions. Unlike other hypoglycemic agents such as acarbose and sulfonylureas, it undergoes mostly renal excretion and is not associated with hepatotoxicity. We present a rare case of acute hepatitis from sitagliptin in a patient with T2DM. A 38 year old man with T2DM, hypothyroidism and schizophrenia presented for follow up of his diabetes which had previously been controlled with lifestyle modification. His medications were quetiapine, paliperidone, fluoxetine, levothyroxine and simvastatin. Physical exam showed a BMI of 37. Abdominal exam was normal. Labs showed a HbA1c of 7.2% which was up from 6.8% four months prior. His AST and ALT were 44 and 58 units/L, respectively, unchanged from prior values. The decision was made to start the patient on sitagliptin 100 mg daily. At a four month follow up, his HbA1C was 6.0%. His AST and ALT had increased to 134 and 232 units/L with a total bilirubin of 1.2 mg/dl. Simvastatin was stopped at the time. Repeat labs one month later revealed AST and ALT of 405 and >500 units/L at which point sitagliptin was stopped. An abdominal ultrasound showed no gallstones or biliary ductal dilatation. Acute hepatitis panel revealed a positive hepatitis B surface antigen with negative surface antibody, negative E antigen and positive core antibody. Viral load was 5 million copies. Hepatitis A and C antibodies were negative. One month after stopping sitagliptin, his AST/ALT improved to 134 and 381 units/L and returned to baseline a month later. The patient was referred to a hepatologist for chronic hepatitis B and was started on entecavir. Drug induced liver injury due to anti-glycemic agents has been associated with sulfonylureas and acarbose but traditionally has not been associated with DPP-4 inhibitors. Multiple clinical trials assessing safety and tolerability have not shown hepatotoxicity associated with these agents. On the contrary, sitagliptin has been shown to be safe and effective in patients with chronic hepatitis as it undergoes minimal hepatic metabolism, unlike acarbose or sulfonylureas. Moreover, DPP-4 inhibitors have shown efficacy in reversing nonalcoholic fatty liver disease (NAFLD) and steatohepatitis (NASH). Our patient had incidentally diagnosed chronic hepatitis but it is unlikely the cause of his acute hepatotoxicity given the temporal relationship of sitagliptin and hepatocellular injury followed by improvement of the liver enzymes after discontinuation. Sitagliptin should be recognized as a rare cause of drug induced acute hepatitis.
Tyrosine Kinase Inhibitors (TKIs) are oncologic agents used for Philadelphia chromosome positive chronic myelogenous leukemia (CML) and acute lymphoblastic leukemia (ALL). They work by blocking the action of BCR-ABL, a fusion oncogene associated with the 9;22 translocation that encodes for a constitutively active tyrosine kinase leading to unregulated tumor activity. When combined with other chemotherapeutic agents, patients achieve higher rates of complete remission and overall improved outcomes. However, side effects of TKIs can limit their use. TKIs have been associated with pancreatitis, possibly related to TKI induced ischemia or reflux of enzymes causing pancreatic injury. In most cases, discontinuation of the TKI leads to prompt recovery. We present the first case of delayed pancreatitis three months after discontinuation of ponatinib in a patient with ALL. A 30 year-old Hispanic male with a history of relapsed Philadelphia chromosome positive ALL presented with fevers and night sweats. A bone marrow biopsy was consistent with CML with lymphoid blast crisis(ALL). The patient began standard chemotherapy along with imatinib which was subsequently changed to nilotinib. A screening lumbar puncture a year later showed CNS involvement indicating worsening disease. The patient was started on ponatinib 45 mg. A restaging bone marrow biopsy 5 months later showed further progression of disease and ponatinib was discontinued. Three months after discontinuation of ponatinib, he presented with acute onset mid-upper abdominal pain, nausea and vomiting. He denied history of alcohol use and had undergone a cholecystectomy a year prior. His leukocyte count was 0.7 k/uL, lipase 5,707 units/L and normal triglyceride levels. Abdominal ultrasound revealed a nonspecific echogenicity of the pancreatic head. The patient improved with bowel rest, pain medications and intravenous hydration after which he was discharged. TKIs have improved outcomes and survival in patients with Philadelphia chromosome positive leukemias. Their use, however, has been limited by adverse events including gastrointestinal toxicity. Hepatitis, pancreatitis and GI bleeding have been described. Pancreatitis is a well-documented adverse reaction associated with TKIs with a rate of up to 6% with ponatinib. The etiology may be related to pancreatic ischemia or altered GI motility resulting in reflux of intestinal contents and subsequent parenchymal injury. Time to onset of pancreatitis varies and is not related to dosage. Most cases resolve within a week of cessation. Our patient took ponatinib for 6 months without evidence of efficacy. Three months after discontinuation, he developed acute pancreatitis. Other etiologies of pancreatitis, including alcohol, gallstone pancreatitis and hypertriglyceridemia were ruled out. The pathophysiology behind the episode of pancreatitis 3 months after cessation of TKI therapy is not clear but it can be postulated that the medication may have caused irreversible changes in the vascular supply of the pancreas making it more prone to injury.
Extensive Arterial and Venous Thrombi as a Presentation of Hypereosinophilic Syndrome with a Unique Complication of Hemolytic Anemia and Poor Response to Treatment

Mansi Shah, Matthew Deek (Co-author)

Idiopathic hypereosinophilic syndrome (HES) is defined as persistent eosinophilia with end organ damage in the absence of a neoplastic process or reactive eosinophilia. Major organ damage can occur due to eosinophil infiltration, which may manifest as fibrosis, thrombosis with or without thromboembolism, cutaneous or mucosal involvement, edema, and neurologic deficits. Amongst idiopathic HES is a lymphocytic variant caused by an aberrant T cell lymphocyte population that overproduces the cytokine interleukin-5. Some patients with the lymphocytic variant HES may eventually develop T-cell lymphoma. A previously healthy 46 year old man, who recently emigrated from Dominican Republic with a diagnosis of bilateral lower extremity DVT on warfarin therapy, presented with progressively worsening bilateral lower extremity pain, cyanosis of his right foot, and 18lb unintentional weight loss over three weeks. On exam, the patient was tachycardic with bilateral lower extremity edema, had dusky discoloration of the digits of hands and feet with weak peripheral pulses. No rash, lymphadenopathy, respiratory wheezes, masses or organomegaly were present. Initial diagnostic tests revealed severe eosinophilia (WBC, 30.4x10^3; eosinophils 20.7x10^3) and anemia (Hgb 9.6g/dL). Results of vascular studies confirmed with CT showed extensive arterial thrombi of the right upper, left upper, and right lower extremity and venous thrombi of the IVC, right peroneal, right posterior tibial, right popliteal, right femoral, right external iliac, left popliteal, left femoral, left external iliac, and hepatic vein. An extensive investigation was pursued and ruled out infectious etiologies including parasites, HIV, and hepatitis. His hospital course was further complicated by the development of hemolytic anemia for which he was treated with IVIG and required transfusion support. Subsequent hematological work up including bone marrow biopsy revealed a monoclonal T-cell population, and the patient was diagnosed with hypereosinophilic syndrome with a clonal T-cell mediated lymphoproliferative disorder (CD3-/CD4-/FIP1L1-PDGFRα-). He was treated with a trial of high dose corticosteroids and adequate control of eosinophilia was achieved. Due to his widespread arterial and venous thrombi, he was discharged on warfarin. Two weeks after discharge, the patient returned with a gangrenous right foot and was found to have refractory hypereosinophilia (WBC, 21x10^3; eosinophils 5.3x10^3) with recurrent thrombosis, which required a transmetatarsal amputation. Weekly methotrexate 20 mg/m2 IV and dexamethasone 40mg IV were initiated as treatment. Despite continued treatment with steroids and warfarin over the next two months, his hypereosinophilia persisted and symptoms of the disease continued to progress—eventually involving his fingers. This case represents a unique presentation of T-cell mediated HES with a lymphocytic variant. The extent of eosinophil-mediated venous and arterial thrombi is greater than typically found in the literature. Moreover, the presence of complications such as hemolytic anemia may be a marker for disease refractoriness and prognosis, and should be considered when determining treatment options.
Inter-hospital Transfers: How an Unstructured Process Impacts Patient Outcomes

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Background: Inter-hospital transfers are an important yet understudied subset in transitions of care, as they pose unique difficulties to health care providers. Timing of transfer, incompatible information systems, geographical distance, and patient acuity combine to create considerable obstacles to safe and efficient patient care. There is currently no data to suggest how the process of patient transfer between facilities impacts patient outcomes; this study aims to fill that gap. Methods: In this retrospective observational study, titled the Inter-Hospital Transfer study (IHTS), we used the Robert Wood Johnson University Hospital ICU tracker and patients’ medical records to evaluate consecutive patient transfers from outside hospitals (OSH) to RWJ three intensive care units (medical, surgical, and cardiac) during December 2011 to December 2012. We gathered data on the features of each patient’s transfer, and analyzed the information that came with these patients for completeness and relevance using a standardized scoring system with two independent reviewers (transfer score). Outcomes focused on adverse events, including in-hospital mortality, central line placement, transfusion, intubation, new vasopressor initiation, mortality, ICU readmission, initiation of renal replacement. We used multivariate logistic regression to identify predictors of high-risk inter-hospital transfers. Results: 328 patients were transferred to RWJ’s three ICUs from 37 OSH between December 2011 and December 2012. Summary statistics included average duration of stay at outside facility, delay of transfer, and percentage of patients that arrived during the night shift. On average, patients were admitted for 4.24 days to an OSH and took 9.6 hours to be transferred to RWJ once initial call was placed to transfer center. Important predictors of adverse events were critical labs on admission (p<0.001), MPM/ApacheII score (p<0.0001), MICU admission (vs SICU/CCU) (p=0.002). There was no correlation between severity of illness (MPM or APACHE), or other predictors of adverse outcomes and time to transfer. Patient arrival followed a bimodal distribution with peak arrival times at 6:00pm (within one hour of nursing and resident shift change) and at 2:00am. When controlling for MPM and transfer score, arrival at night (representing 50% of the transfer population) was associated with a significant increase in adverse events (p =0.006). Discussion: Patients are often transferred between facilities in unstable condition or at inopportune times, which is associated with an increase in adverse events. Unfortunately, patient transfers are not triaged solely based on severity of illness, but often on administrative issues such as bed and transport availability. Currently, there are no best practices identified for the coordination inter-hospital transfers. This study highlights some of the more challenging aspects of many inter-hospital transfers; a more systematic approach to these patients could improve patient outcomes.
Fever and Rash as Macrophage Activating Syndrome

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Introduction: Macrophage activation syndrome (MAS), also known as Hemophagocytic lymphohistiocytosis, is a condition characterized by the activation and proliferation of T lymphocytes and macrophages, leading to widespread hemophagocytosis and cytokine overproduction. Clinically, it presents as pancytopenia, coagulopathy, neurologic symptoms, mucocutaneous findings, as well as hepatosplenomegaly. There are no validated diagnostic criteria for MAS, and although bone marrow biopsy shows hemophagocytosis, a negative result does not exclude the syndrome. A sample can be obtained from the spleen and lymph nodes as well. Macrophage activating syndrome has been related to numerous triggers, including viral infections. It is a life-threatening condition, and the reported mortality rates reach 20–30%. Case Description: A 27 year old Hispanic male with no past medical history presented to the hospital in July 2013 with the complaint of headaches for the past ten days. Physical exam was remarkable for epigastric tenderness and a diffuse maculopapular rash which developed shortly after taking an anxiety medication prescribed from Guatemala. The patient was febrile on presentation (102 F). He was admitted with a possible diagnosis of aseptic meningitis and an LP was performed which was negative. An extensive serology list was sent which included tick borne illnesses, HIV, vasculitis, and various other viral syndromes, all of which were subsequently negative. A skin biopsy was done to rule out DRESS syndrome. On day four of the admission, the patient's liver enzymes and pancytopenia worsened, and he became increasingly lethargic. Empiric antibiotics were not helpful in treating his fevers and lethargy. On hospital day five, low dose steroids were started and the patient became afebrile. His lethargy, pancytopenia, and transaminases only improved when switched to high dose pulse steroid therapy. Discussion: This case illustrates recognizing a rare but serious condition known as Macrophage Activation Syndrome (MAS). MAS should be suspected when a patient has a fever that is unresponsive to antibiotics, pancytopenia, low erythrocyte sedimentation rate (ESR), and high ferritin (>10,000 ug/l). Liver transaminases may be elevated as well. Although the exact pathogenesis remains unknown, it is our conjecture that this syndrome is a complication of a viral etiology. Early institution of high dose steroids can be life saving, even in the face of pancytopenia. We present this case to educate medical staff on this rare condition and to expedite its diagnosis in other patients.
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**Chest pain due to atherosclerotic coronary artery disease in a patient with single coronary artery**

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Coronary artery anomalies (CAA) are rare, and single coronary arteries (SCA) are even rarer occurring 0.024%-0.066% in the general population. We present a case of a 65-year-old lady with past medical history of hypertension and diabetes mellitus II, who presented with complaints of left sided chest pain with substernal tightness that radiated down her left arm and was relieved by sublingual nitroglycerine. This pain was accompanied by shortness of breath and palpitations. Initial ECG showed ST segment depression in lead II without ST segment elevation. The patient was started on nitroglycerin and heparin drip and was transferred to the coronary care unit for close monitoring until cardiac catheterization the following day. The patient was diagnosed, on cardiac catheterization as having a single coronary artery supplying the entire heart. The cardiac catheterization showed the patient did not have a right coronary artery (RCA). The blood supply to the heart originated entirely from the Left Main Coronary Artery (LMCA) which gave rise to Left anterior descending (LAD) and left circumflex (LCX) arteries. The LCX artery then continued to supply the right side of the heart. The cardiac catheterization also showed a 90% stenosis to the proximal D1 branch of the left anterior descending (LAD). After assessing for ischemia with Lexiscan stress test, a percutaneous coronary intervention (PCI) was performed to the D1 lesion. The patient was chest pain free upon discharge and was doing well at followup. We present this case as a rare instance of Single Coronary Artery Syndrome presenting as acute coronary syndrome.
A Case of Cerebral Venous Sinus Thrombosis in a young male with hypernatremia

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Cerebral venous sinus thrombosis is a rare condition that occurs approximately three to four times per one million population and about seven cases per one million children. The rarity of the condition does not lend itself to allow for frequent studies and investigations that allow for better interventions and therapeutics. A 29 year old male inmate with a past medical history of generalized anxiety disorder, depression, schizophrenia, cannabis abuse and malingering came to the emergency room from the prison because of poor appetite and increasing headaches and then eventually became increasingly confused, disoriented and drowsy and was admitted to the intensive care unit for altered mental status and generalized weakness. On physical examination the patient was lethargic and arousable, able to follow commands, localize to pain in all four extremities. The eyes tracked to voice but the patient demonstrated incoherent mumbling of words. The patients face was symmetric and exhibited no facial droop. The patient was able to move both upper and lower extremities and was able to squeeze hands bilaterally and the patient had a significantly stiff neck. The Reflexes demonstrated quick plantar stimulation with upgoing toes but rapidly became downgoing and withdrawing of the feet. Initial laboratory investigation revealed a sodium of 158 which eventually became 166 within 12 hours and a white blood cell count of 16.2 with 84% neutrophils. Coagulation profile showed an INR of 1.5, urinalysis and urine toxicology was negative, arterial blood gases showed did not show any hypoxia or acid base disorder. The initial chest radiograph was unremarkable and a CT head without contrast revealed findings suspicious for diffuse dural sinus thrombosis. An MRI of the brain with and without contrast showed the findings of major and superficial and deep dural venous sinus thrombosis and bilateral ischemia and/or infarction of the thalami. An MR venogram was used for confirmation of the previous imaging and revealed absene of flow within most of the superficial and deep major dural venous sinuses which supported the final diagnosis of superficial and deep dural sinus thrombosis. The patient was eventually transferred to a tertiary care facility for treatment.Cerebral venous sinus thrombosis is very rare and can develop with a gradual or sudden headache and its sequelae can include neurologic deficits, seizures, and behavioral changes and even coma and death. Even though hypernatremia can present with lethargy it is pertinent to exclude all causes of an alteration in mentation such that a rare cause cannot be missed as was demonstrated.
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Acute Inflammatory Demyelinating Polyneuropathy and Lyme Neuroborreliosis

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Introduction: Acute inflammatory demyelinating polyneuropathy is an antibody-mediated autoimmune syndrome of demyelination with or without axonal nerve loss. AIDP presents with distal ascending motor and/or sensory signs and symptoms, and areflexia. There is often an antecedent infection identified. While any infection can theoretically trigger AIDP, Borrelia burgdorferi is uncommon. We report a case of AIDP in a patient found to have central nerve system Lyme Disease (neuroborreliosis).

Case Presentation: 80 year old male with extensive past medical history including Hypertension, Hyperlipidemia, and Diabetes Mellitus presenting with ascending weakness beginning in his lower extremities and progressing to his hands and shoulders over 2 weeks. He had associated paresthesias in his lower extremities. Patient denied any urinary or fecal incontinence. Patient denied any recent viral or bacterial illness, and no sick contacts. General physical exam and vital signs were normal. Neurological exam showed 4/5 strength in the proximal lower extremities bilaterally, 5/5 strength in the distal lower extremities bilaterally, and 5/5 strength in the upper extremities bilaterally. No muscular atrophy was evident. Patient had normal tone. Deep tendon reflexes were absent. There was decreased proprioception, with decreased sensation in a "stocking and glove" distribution. The patient had unsteady gait. CT of the head showed no evidence of acute cranial abnormalities. CSF analysis showed an opening pressure of 10, RBC of 96, WBC of 1, total protein of 88, and glucose of 87. An electromyography demonstrated neuropathic findings with no evidence of axonal injury, most likely a demyelinating process. A nerve conduction study demonstrated evidence of a sensory polyneuropaty. Lyme Antibody was positive. A western blot was reactive at the IgG 58 KD Band and IgM 23 KD Band, confirming the findings. The patient was started on Prednisone and Doxycycline. He was followed up as an outpatient with neurology and clinically improved.
INTRODUCTION Retroodontoid pseudotumors are caused by inflammatory granulation or reactive soft tissue hypertrophy from chronic atlantoaxial subluxation of inflammatory or traumatic origin. CASE DESCRIPTION 86 years old Caucasian Priest presented to hospital after 2 episodes of falls in the chapels. He suddenly felt generalized weakness and could not get himself up. He was having gait dysfunction and has been using a cane for one year. He had new onset of neck pain and bilateral numbness over the palm for four months. There were no signs and symptoms of syncope. Physical examination showed normal vitals sign, no orthostatic hypotension nor neurocutaneous stigmata. Patient was awake, alert, oriented for time, place and person. His speech was fluent, coherent and relevant, without aphasia. Cranial nerves exam was normal. Sensory exam showed decreased pin prick sensation on bilateral feet. Motor strength was 4/5 on right upper extremity and bilateral lower extremities. Cervical spine exam was normal with no sign of cervical radiculopathy. Reflexes were 2+ bilaterally upper and lower extremities. An MRI of cervical spine showed spinal stenosis with spinal cord compression at the level of odontoid process caused by retro-odontoid pseudo-tumor which was about 12 mm in size causing chronic spinal cord myelomalacia with intact CSF flow. MRI of the whole spine demonstrated multilevel extensive spinal and foraminal stenosis along with spondylosis. This patient had multilevel spinal disease rising suspicion of diffuse idiopathic skeletal hyperostosis (Forestier’s Disease) as well. The management was beyond the scope of our community based hospital, so the patient was referred to tertiary care center for advanced management. DISCUSSION: Retro-odontoid pseudotumor is reactive soft tissue hypertrophy caused by chronic inflammatory granulation causing. A retro-odontoid pseudotumor is commonly associated with atlanto-axial subluxation. Rheumatoid arthritis is the most frequent cause of retro-odontoid pseudotumors. As per an article published in 2009, the spinal column is an extremely rare site for inflammatory pseudotumors; and only 16 cases have been reported. More common sites include lungs, orbits, gastrointestinal tract etc. Etiology can be associated with inflammatory conditions like Rheumatoid arthritis, infectious like TB, HIV, amebiasis; traumatic or mechanical stress related. Treatment may be surgical or radiation therapy depending upon overall presentation.
To Give or Not to Give? Biphosphonates in Primary Hyperparathyroidism

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A 32 year old African-American female presents complaining of right sided chest pain that began after a recent coughing episode. Initial imaging revealed multiple sub-acute rib fractures and humeral cystic lesions bilaterally. Her corrected Calcium was 11.7mg/dL, Phosphorus: 1.8mg/dL, iPTH: 655pg/mL, Alkaline Phosphatase: 863 U/L, serum 25-hydroxy Vitamin D: 79 pg/mL. Previous records revealed she initially refused surgical management. Pamidronate 60mg intravenously was initiated to decrease the hypercalcemia. Re-education of the risks and benefits of the proposed surgical intervention resulted in the patient agreeing to advance with the procedure. On postoperative day one, laboratory data revealed iPTH: 33pg/mL, corrected Calcium: 8.1mg/dL and Phosphorus: 2.5 mg/dL, suggesting Hungry Bone Syndrome (HBS). Intravenous Calcium Gluconate was administered. On day three, corrected Calcium dropped to 7.4mg/dL. Phosphorus levels remained low in spite of continuous intravenous replacement. On day 5 she presented muscle cramps, peri-oral numbness and demonstrated positive Chovstek"s and Trousseau"s Signs. Prolonged QTc intervals of 514 msec were recorded on the electrocardiogram. Despite intravenous and oral supplementary calcium for several days, refractory hypocalcemia persisted dictating the addition of Calcitriol. This improved her serum calcium levels and her clinical symptoms.
A 28 year old Puerto Rican woman with a history of Graves disease presented to the emergency department after a transient loss of consciousness, having been found unconscious in bed. She noted intermittent palpitations of 3 weeks duration, along with severe voice hoarseness and goiter. Initial evaluation revealed sinus tachycardia, labile blood pressure, severe anxiety, exophthalmus, an enlarged palpable thyroid gland, a serum thyroid stimulating hormone less than 0.02, and free T4 level greater than 7.77. Subsequently, tachycardia worsened and low grade fevers were noted, and a diagnosis of thyroid storm was made. Due to an iodine allergy, Lugol’s was not used; however, the patient initially improved with methimazole, metoprolol tartrate, and fluids. Subsequently, she began having generalized tonic-clonic seizures. Noncontrast CT and MRI of the brain were normal, and an electroencephalogram was performed. The EEG demonstrated bursts of high voltage sharp and slow wave activity. Her convulsions progressively worsened and became refractory despite intravenous benzodiazepines, and a diagnosis of status epilepticus was made. She was transferred to the intensive care unit, placed on continuous intravenous propofol infusion, and intubated for airway protection. Further investigation revealed recent sexual assault and the presence of herpetiform genital lesions; thus, a lumbar puncture was performed to rule out HSV encephalitis. Evaluation of cerebrospinal fluid was unremarkable. Her convulsions stopped once propofol was sufficiently titrated, and as her thyroid function tests normalized, she was weaned off of sedatives and successfully extubated.
Seton Hall- St. Francis Medical Center

**Intracystic papillary carcinoma (IPC) in association with Invasive ductal carcinoma – An extremely rare case of Male Breast Cancer (MBC)**

Nishith Mewada, Leslie Mechanic MD, Elizabeth Rosvold MD, Robert Moser MD, Karen J Kish MD

**Introduction:** IPC is a rare breast cancer accounting for 0.5-2% of all breast cancers which is seldom reported in males. **Case Report:** A 56 year old African American male was evaluated for enlarging left breast mass and intermittent bleeding from left nipple over past year after human bite. An antibiotics trial did not help. He is unmarried, denied smoking cigarettes, drinking alcohol or intravenous drug abuse. His sister died of breast cancer at 64 years age and brother had prostate cancer at the age of 51 years. He has two children. His children’s mother was diagnosed with breast cancer at the age of 41 years. Upon examination, he had 6x6 cm firm, non-tender mass in the left breast. The left nipple was inverted and had hemorrhagic discharge. Right breast exam was normal. There was no lymphadenopathy. Ultrasonography showed a poorly defined heterogeneous, hyperechoic, hypervascular mass beneath the nipple measuring 2.3x1.4 cm. Fine needle aspiration resulted in hemorrhagic fluid. Cytology revealed RBCs with hemosiderin laden macrophages and clumps of epithelial cells with atypia. CT chest demonstrated a large 6.9x4.9 cm complex mass measuring 18-30 Hounsfield units in the left sub-areolar region with solid and cystic components without regional lymphadenopathy. Ultrasound guided core biopsy of the solid component demonstrated E-cadherin positive papillary ductal proliferative lesion suspicious for ductal epithelial neoplasm. A collective decision was made to proceed with surgical excision. Sentinel lymph node biopsy were negative for malignancy. A left simple mastectomy was performed. Final diagnosis was pT1apN0M0, Stage IA multifocal Invasive Ductal Carcinoma arising in association with Intracystic Papillary Carcinoma. The largest tumor focus was 0.5 cm. Margins were free of carcinoma. The tumor was ER+/PR+, HER2 negative. He was recommended to receive Tamoxifen for 5 years and close medical and surgical surveillance. **Discussion:** MBC represents 1% of all breast cancer cases. In a review of 917 cases of IPC, only 3.5% cases were seen in male population. Nearly 22% cases have associated invasive ductal carcinoma. Overall mean age of diagnosis is 69.5 to 73.4 years. 89.6% are localized at the time of diagnosis. Most often a palpable complex hemorrhagic cyst is a presenting symptom. Cytology from fluid may not be conclusive. CEA level from the fluid more than 400ng/mL may suggest malignant disease. Around 75% of all MBC are hormone receptors positive. Although clear guidelines are not available, sentinel lymph node biopsy followed by mastectomy or breast conservative surgery is the standard approach. Radiation or Hormone therapy may not affect the outcome of the disease. IPC carries excellent prognosis. Absolute cumulative survival rates are 75% at 5 years and 60.6% at 10 years. Age adjusted relative cumulative survival rates are 93.9% at 5 years and 94.4% at 10 years.
Job Syndrome, Hyperimmunoglobulin E Syndrome (HIES): A case report and literature review.

Nishith Mewada, Mohammad Ali Ursani MD, Sylvia Sasinowska MD, Asim Hussain MD, Nigahus Karabulut, MD

Introduction Hyperimmunoglobulin E Syndrome (HIES), also described as Job Syndrome, is a rare immunodeficiency syndrome that is classically known to present with the triad of eczema, recurrent lung and skin infections, and significantly elevated IgE level (>2000 IU/ml). Clinical Vignette A 23 year old Hispanic male with history of premature birth, life threatening otolaryngeal and sinus infections postpartum, developmental delay, congenital deafness and mutism, asthma, bullous emphysema, and spontaneous pneumothorax requiring pleurodesis in adolescence was brought to the emergency department for evaluation of pleuritic chest pain, cough and low grade fever. He had normal WBC count with 22% eosinophils. Upon workup was diagnosed to have 7.2 cm cavitory lung lesion, bronchiectasis and ground glass opacities. After ruling out Tuberculosis, he was treated with Amoxicillin clavulanate. No certain etiology could be established. In 3 months, he was readmitted to the hospital with the same complaints and presentation. Given Eosinophilia, IgE levels were checked and were 13405 IU/L. He also had dry skin and eczematous lesions over elbows and upper back, deep set eyes, prominent forehead and scoliosis (12 degree). He consistently had eosinophilic response to the infection. A suspicion of Job Syndrome was raised. He underwent resection of the lesion which upon culture grew Aspergillus. He was treated with Amphotericin and Vancomycin and discharged successfully from the hospital. Patient’s clinical presentation is definitely suggestive of Job Syndrome. We are working in conjunction with the NIH for STAT3 mutation analysis. Discussion HIES is a rare, probably underdiagnosed disorder. In addition to the triad, is also described with a broader array of clinical characteristics, such as deep set eyes, prominent forehead, and coarse skin, hyper-extensibility, scoliosis (>10 degree), fragility fractures, CNS abnormalities, Craniosynostosis, dental abnormalities and arterial aneurysms. The actual incidence is unknown. Autosomal dominant HIES is a missense or in-frame deletion of the STAT3 gene. STAT3 is a protein involved in signal transduction processes such as angiogenesis, wound healing, and immunity. Autosomal recessive HIES also presents with elevated IgE levels, eczema and recurrent infections but does not encompass the musculoskeletal and dental manifestations. The classic manifestation in early infancy is a papulo-pustular rash, typically caused by Staphylococcal aureus. Recurrent pneumonia and bronchiectasis also develop within the first few years of life with Staphylococcus aureus, Streptococcus pneumoniae and Hemophilus influenzae being the common organism. It predisposes to opportunistic infections with Aspergillus fumigatus and Pneumocystis jiroveci. Death in adulthood is usually related to the complicated infections. Later in life, HIES increases the risk of Hodgkin’s and Non-Hodgkin’s lymphoma. Thus far, management is based on symptomology. Prophylaxis can be considered for recurrent infections. Topical agents can control eczema. Immune-modulators and hematopoietic cell transplantation have not yet demonstrated long-term benefits.
Seton Hall- St. Francis Medical Center

The association between chest pain control and clinical outcomes in patients with acute coronary syndrome

Eric Osgood, Muddassir, S. Tariq, S. Mewada, N. Thomas, KK. Sarwar, M

Introduction Aggressive treatment of chest pain is considered pivotal in acute coronary syndrome (ACS); the putative physiology being myocardial demand reduction by minimizing sympathetic amines. However, analyses of the impact of chest pain severity on clinical outcomes have never been published.

Methods We conducted a chart-based retrospective case control pilot study investigating the association between chest pain and adverse outcomes in adults under 80 years old with ACS. Patients with angina admitted for ACS who underwent cardiac catheterization without prior intervention or bypass were included. Subjects with previous cardiac or renal disease were excluded. Composite outcome of re-infarction, heart failure/cardiomyopathy, ventricular arrhythmia, or mortality comprised Cases. Controls had none of these outcomes. ROC analysis defined good versus bad pain control, while one-way-ANOVA and Mann-Whitney-U testing determined magnitude and significance of pain reduction. Between-group differences in pain control were analyzed via Fisher''s Exact testing. Angiographic findings as well as pertinent baseline characteristics were also compared between groups using these methods.

Results Seven hundred charts were screened, and thirty eligible subjects were identified: 20 Cases, 10 Controls. Elimination of chest pain occurred in 93% of subjects (95% Controls, 90% Cases). Good versus bad pain control dichotomized subjects optimally based on whether chest pain was eliminated (0/10) within 12 hours [AUC 0.75; 95% CI 0.54-0.96; p=0.03]. Mean pain intensities remained higher for Cases at all time intervals, reaching significance at hour 12 [0.0/10 vs. 3.2/10; F=13.0, p=0.001; Mann-WhitneyU=50, WilcoxonW=260, 2-tailed p=0.001, 1-tailed p=0.028]. Five Cases and zero Controls experienced bad pain control [50%, 0%; Fisher p=0.002]. Presence of obstructive coronary lesions was higher among Cases but without statistical significance [66% vs. 69%, p=0.41]. There were no statistically significant differences between groups for gender, body mass index, age, smoking history, glomerular filtration rate, or in prevalence of diabetes mellitus, hypertension, or hyperlipidemia. Conclusion Adverse clinical outcomes were associated with poorer antecedent pain control during initial treatment, which were not explained by discrepancy in intraluminal disease severity or in underlying medical comorbidity, implicating analgesia in ACS as an independent outcome predictor. To our knowledge, this is the first study to date examining whether adequacy of analgesic treatment of angina in ACS impacts clinical outcomes. This study is limited by its small sample size and retrospective design.
An Atypical Presentation of a Cerebral Colloid Cyst

Sahana Ramanujam, Syna Jose DO; Ralph Meloro MD; Salman Muddassir MD; Sara Wallach MD

Introduction Cerebral colloid cysts are benign congenital lesions classically found in the rostral aspect of the third ventricle near the foramen of Monroe. These cysts are rare and represent less than 2% of all benign tumors. Acute hydrocephalus, brain herniation, and even death has shown to result from this “benign” lesion. The quintessential presentation of positional headaches, memory loss and gait disturbances normally present in those patients who have already developed normal pressure hydrocephalus. Slow onset of symptoms may be reflective of the slow growing process of the cysts. Early detection leading to early intervention typically results in an excellent prognosis.

Case Report

82 year old male presented with three week history of dizziness, headaches, and mild short-term memory loss. Initial workup was suggestive of hyperglycemia due to non-compliance. However, a cerebral colloid cyst was identified during a routine non-contrast CT scan of the head. Magnetic Resonance Imaging confirmed the presence of the ovoid mass in the anterior third ventricle with no evidence of obstructive hydrocephalus. It was decided to hold any invasive therapy until such time that the colloid cyst produced life-limiting effects. Patient was directed toward medication compliance, supportive measures including symptomatic treatment with non-steroidal anti-inflammatory agents as needed for headaches and modest lifestyle adjustments in diet, daily physical activity and dizziness precautions. Follow up examination showed improved glycemic control, no increase in headaches or episodes of dizziness and slight subjective reduction in short-term memory loss. There will be no invasive procedures unless symptoms change which suggest increased colloid cyst size possibly leading to life threatening effects.

Discussion

Colloid cysts are benign intracranial congenital lesions, representing less than 2% of all benign tumors. Patients typically complain of positional headaches and gait disturbances. Slow onset of symptoms may be reflective of slow growing process of the cysts. Early detection leading to early intervention typically result in an excellent prognosis.
Isolated Oculomotor Nerve Palsy: A Rare Presentation of Pituitary Adenoma

Sahana Ramanujam, Ramesh Adhikari, MBBS, MS; Kendal Thomas, MD; Samir Undavia, MD; Sara Wallach, MD.

Introduction Pituitary adenomas comprise of 10-25% of all intracranial tumors. Vision loss, endocrine dysfunction and headaches are the most common initial presentations. Sudden ophthalmoplegic deficits have been observed in patients with more aggressive tumors and usually indicate ominous complications such as acute pituitary apoplexy. Cranial nerve palsies accompany visual field defects as the tumor invades the cavernous sinus. In these instances, typically cranial nerves three, four, five [1st, and 2nd divisions], and six can be involved to varying degrees. Rarely have solitary oculomotor nerve paresis been the presenting symptom. Case Report: A 72 year old African American female with multiple co-morbidities, including Ramsey Hunt Syndrome, presented with a two week history of progressively worsening headaches, left eye pain and ptosis. Magnetic resonance imaging of the brain showed a pituitary adenoma impinging on the optic chiasm with invasion into the cavernous sinus. After undergoing a transsphenoidal transseptal hypophysectomy and resection of pituitary adenoma, patient had complete resolution of presenting complaints. Discussion: Pituitary adenomas typically present with vision and/or endocrine deficiencies. Oculomotor nerve palsy is a relatively rare symptom and typically indicates development of pituitary apoplexy. However, in rare cases, isolated third nerve palsy may represent a benign process. Early detection and treatment may reverse neurological impairments in patients with pituitary adenomas.
INTRODUCTION: Prostate Cancer is the second most common cancer in men in the world. It is more common in African American than Caucasian men in USA. 95% of prostate cancers are adenocarcinoma. This case report describes the rare presentation of metastatic adenocarcinoma of prostate with anemia and congestive heart failure. CASE REPORT: Our patient is a 65 year old African American male with no past medical history. He had never seen a primary care doctor or visited an emergency room in the past. He was admitted to our hospital with chief complaint of shortness of breath. Progressive worsening of shortness of breath made him seek medical attention. In addition, patient reported having bleeding per rectum for 4 days with bilateral swelling of legs, scrotum, penis and upper abdomen without any urinary symptoms. Physical examination revealed jugular venous distension, bilateral pitting edema of lower abdomen, penis, scrotum and legs with palpable bilateral inguinal lymph nodes. Irregular palpable nodular prostate was noted on rectal exam. Patient labs showed iron deficiency anemia with low hemoglobin (4.8 gm/dl), elevated alkaline phosphatase (784 Units/liter), elevated BNP (30005 pG/ml). Chest x-ray was significant for bilateral pleural effusions with pulmonary edema. On CT abdomen and pelvis, severe diffuse sclerotic osseous metastatic disease were present, as well as bilateral hydronephrosis with retroperitoneal adenopathy. Patient had high PSA which when quantified was 67370 ng/ml. Prostate biopsy confirmed adenocarcinoma of prostate, Gleason Score 8, Stage IV (T4N1M1b). Patient currently is being treated with Bicalutamide at the rehabilitation facility. He has most recent follow up PSA is 1114 ng/ml. CONCLUSION: Prostate cancer is a disease of elderly males. Incidence escalates dramatically with increasing age. Clinical symptoms are hard to differentiate from benign prostate hypertrophy. Diagnosis is confirmed by biopsy. The American Joint Committee on Cancer has designated staging of prostate cancer using TNM classification. Treatment options depend on the stage of cancer.
Peripheral primitive neuroectodermal tumour of the pleura in an adult female

Gwenalyn Garcia, Jumana Chatiwala, MD

INTRODUCTION    Peripheral primitive neuroectodermal tumors (pPNET) are small round blue cell neoplasms belonging to the Ewing sarcoma family of tumors (ESFT). They occur more commonly in childhood and are rare in adults. We report a rare case of pPNET of the pleura in a 55-year-old female with a demonstrated translocation involving the EWSR1 gene.

CLINICAL CASE    A 55-year-old Taiwanese female presented with exertional dyspnea, left posterior chest pain, and dry cough of three weeks duration. Her past medical history was significant for carcinoma in-situ of the left breast treated with lumpectomy and radiation. She had initially been evaluated seven months prior for similar complaints; a CT chest done at the time showed a 6.6 cm pleural mass along the left upper lobe. However, the patient was lost to follow-up. Repeat CT chest showed increase in size of the mass to 8.5 cm in diameter. It also showed a second soft tissue mass in the left major fissure, mediastinal lymphadenopathy, and loculated pleural effusions along the left upper and lower lobes.

The patient underwent CT-guided biopsy of the mass, which showed small round blue cells consistent with pPNET/Ewing sarcoma. The tumor cells were positive for CD 99 and BCL-2. Fluorescent in-situ hybridization (FISH) was positive for a translocation involving the EWSR1 gene in 88.5% of cells. The patient underwent video-assisted thorascopic surgery with drainage of pleural effusion, which resulted in improvement of her dyspnea. A bone scan showed local spread to the left 6th rib. The patient was started on a regimen of vincristine, adriamycin, and cyclophosphamide, alternating with ifosfamide and etoposide. She tolerated her first cycle of chemotherapy well.

DISCUSSION    Ewing sarcoma and pPNET are small round blue cell tumours comprising the ESFT. Ewing sarcoma is the second most common malignant bone tumor in children, while pPNET are soft tissue tumors postulated to arise from neural crest cells. On immunohistochemical staining, over 90% of ESFT are positive for CD 99 and FLI-1. Ninety-five percent of ESFT express a characteristic translocation involving the EWSR1 gene on chromosome 22. A review of the English literature revealed 3 cases of adult primary pleural pPNET. Cytogenetic testing for translocations involving EWSR1 was performed in one case, while a diagnosis was made based on immunohistochemistry in the remaining cases. Ours represents the second case of adult primary pleural pPNET with a demonstrated ESFT translocation.
A Fatal Case of MSSA Gangrenous Cholecystitis in an Intravenous Drug User

Gwenalyn Garcia

INTRODUCTION Staphylococcus aureus is an uncommon pathogen in acute cholecystitis. We present a 50-year-old male intravenous drug abuser brought to the emergency department for fever, jaundice, and confusion. The patient deteriorated within a few hours of presentation and subsequently expired. An abdominal ultrasound read postmortem revealed gangrenous acute cholecystitis. Blood cultures grew methicillin-sensitive S. aureus (MSSA). CASE PRESENTATION A 50-year-old Caucasian male with a history of intravenous heroin use was brought to the emergency department for confusion of 10 hours duration. The patient also had a 4-day history of high grade fever and generalized weakness. On presentation, the patient was jaundiced and moaning unintelligibly. His blood pressure was 146/94, heart rate 128 beats/minute, respiratory rate 37 breaths/minute, and rectal temperature 101.6°F. His abdomen was mildly distended, but there was no tenderness or guarding. Skin examination revealed jaundice and a diffuse fine ecchymotic rash. Laboratory studies revealed a total bilirubin elevation of 11.3 mg/dL, with a direct bilirubin of 8.2 mg/dL. A CT abdomen showed a distended gallbladder. The patient was intubated for respiratory distress. He was given IV normal saline and started on piperacillin-tazobactam. The patient developed septic shock within hours of presentation and expired despite resuscitation. An abdominal ultrasound performed previously was read postmortem as cholelithiasis with gangrenous acute cholecystitis. Blood cultures grew MSSA. DISCUSSION S. aureus is an uncommon pathogen in acute cholecystitis. A review of English literature from 2003 to 2012 yielded 9 reported cases of S. aureus cholecystitis in adults. Our case adds to the literature recognizing S. aureus as a pathogen in acute cholecystitis. In our patient, infection of the gallbladder likely occurred via hematogenous seeding given his history of intravenous drug use. Although gallbladder cultures were not obtained, the patient’s bacteremia and rapid decline make S. aureus the likely culprit of disease.
Personality changes as the initial presentation of a rare tumor, Esthesioneuroblastoma: A case report

Norlalak Jiramethee, Rampal, Upamanyu

Esthesioneuroblastoma (ENB), also known as olfactory neuroblastoma, is a rare malignant tumor of the head and neck, which most often arises from the olfactory neuroepithelium in the superior nasal cavity at the anterior skull base. The tumor accounts for 1% to 6% of all intranasal cancers, which constitute less than 1% of all human malignancy. Several treatment approaches have been described in literature, but prospective treatment studies are absent given the tumor’s rarity and pattern of recurrence that requires an extended posttreatment observation period. I present here a case of 47 years old male, who was brought into the hospital after exhibiting strange behavior started 2 weeks prior to this hospitalization. Patient’s parents called psychiatric center to evaluate the patient for increasing abnormal behavior, including refusal to eat, drink or getting out of his room, and was found covered with urine and feces. Physical examination revealed unkempt male patient, who was disoriented to time. Right eye was proptotic. Examination of the right nostril revealed a foul-smelling exophytic mass completely occluding the right nostril, displacing nasal septum to the left. CT head showed expansile heterogeneous mass lesion along the midline of the anterior cranial fossa extending into the ethmoid sinuses and into the nasal cavity with associated osseous erosive changes. The tumor is approximately 5.5 x 4.6 cm in its greatest axial dimension. Patient also developed profound hyponatremia with laboratory work up was most consistent with SIADH. Because of tumor involvement in both intranasally and intracranially, ENT and neurosurgery consultation were obtained. Patient underwent combined ENT and neurosurgery procedure, craniofacial exenteration. Pathology result was consistent with esthesioneuroblastoma. Patient did remarkably well after the procedure and was discharged 10 days after the surgery. Patient received chemotherapy with cisplatin and etoposide during the same hospitalization with excellent tumor response. He later also received a radiation therapy and 2 more cycles of cisplatin and etoposide chemotherapy. Patient did well after the treatment. Although there was residual disease, but it was inactive and size remained stable after follow up MRI 1 year after last cycle of chemotherapy. Patient had permanent loss of olfactory sensation. His abnormal behavior has resolved, and he has returned to work 1 year after initial diagnosis. He will continue to follow up with the oncologist at our hospital’s cancer center given that this tumor has extended time to local and regional recurrence up to >10 years. This case offers a treatment approach with combined modality included surgery, radiation and adjuvant chemotherapy of a rare tumor that yielded a considerably successful result after one year follow-up.
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TYPE III HEREDITARY ANGIOEDEMA IN A MALE

Vidya Kollu, Ashok Movva, Scott Howard

Hereditary angioedema (HAE) is a disorder characterized by recurrent episodes of severe swelling (angioedema). The following description is a rare presentation of Type III HAE in a male. A 48-year-old African American male with no known allergies presented with sudden onset of non-pruritic upper lip swelling. In the past he had similar history of five spontaneous episodes for twelve years. There is no known family history of angioedema. On examination vital signs were stable with no evidence of airway compromise, upper lip was markedly swollen with no clear demarcation between swelling and surrounding skin. Treatment was initiated with high-dose corticosteroid and antihistamine in the emergency department. X-ray neck was suggestive of prevertebral soft tissue swelling. Initial laboratory workup was normal. A clinical diagnosis of Hereditary angioedema was made after careful evaluation and ruling out all other possible conditions. At this point steroids and antihistamines were discontinued as they were not proven to be of any benefit as per the literature reviewed. Significant clinical improvement was seen in 2-3 days with marked reduction of the lip swelling. His complement 3, 4 levels and C1 esterase inhibitor level came back normal and a final diagnosis of HAE type III was made. Type I is the most common, accounting for 85 percent of cases. Type II occurs in 15 percent of cases, and type III is very rare. Features that distinguish type III from types I and II include: presence of normal C1 inhibitor activity levels, average later age of onset, predominance of facial swelling as compared to swellings of the extremities, less frequent symptoms and sex bias. Affected individuals are predominantly women, where it appears to be correlated with conditions of high estrogen levels—for example, pregnancy or the use of oral contraceptives. Though men can be affected, typically at a later age of onset and with less frequency and severity of attacks. Minor trauma or stress may trigger an attack, but swelling often occurs without a known trigger. Mutations in the F12 gene result in the production of factor XII with increased activity, more bradykinin is generated which leads to episodes of swelling, are associated with some cases of hereditary angioedema type III. The diagnosis "HAE with coagulation factor XII gene mutation" requires the corresponding demonstration of the mutation. Until now there is no further laboratory test which could confirm the diagnosis "HAE type III". Recognition and management of hereditary angioedema is required to prevent a potentially fatal outcome. In conclusion, this case illustrates the potential for hereditary angioedema in men in spite of normal C1 esterase inhibitor levels and that unnecessary medications not proven to be of any benefit can be avoided with watchful monitoring.
PSEUDOXANTHOMA ELASTICUM IN SICKLE CELL DISEASE

Vidya Kollu, Rajasingam Jayasingam, Vanessa Canonigo, Fred Aleskerov

Pseudoxanthoma elasticum (PXE) is a connective tissue disorder primarily affecting elastic fibers of the skin, eyes and heart. The association between certain hemoglobinopathies and elastic tissue abnormalities has been known since the late 1950s. We report a patient with sickle cell disease and PXE. A 30-year-old African American male with a history of sickle cell disease, had features consistent with pseudoxanthoma elasticum, such as hyper-extensile redundant skin folds in the neck lasting for 10 years. A skin biopsy taken from a papule showed dermal fragmentation of elastic fibres stained with elastin. There was mild cardiomegaly on chest-X-ray and a trace mitral regurgitation on echocardiogram. The complete clinical spectrum of classical PXE can be present in patients with sickle cell disease and β-thalassemia. PXE-like syndrome is acquired, age-dependent, with a generally late onset, occurring during the second decade of life and increases in frequency with age in a milder form. The high prevalence of the findings implicates the elastic tissue injury as one of the main co-morbid abnormalities encountered in the sickling syndromes. Free radical-mediated oxidative damage to elastic tissue secondary to hemolysis is implicated. In these patients a number of complications, sometimes serious, have been recognized to be related to ocular and vascular elastic tissue defects. As patients with sickle cell disease are living longer, it is likely that more cases of the PXE-like syndrome will be seen. Because several organ systems are involved, medical specialists should be aware of this phenomenon. Routine follow-up of these patients should include careful skin inspection and annual retinal examination. In conclusion, this case illustrates the need for careful evaluation of possible co-morbid conditions in patients with sickle cell disease.
Pericardial tamponade, a rare complication of amyloid heart disease.

Vidya Kollu, Patel Hiten

Background: Restrictive cardiomyopathy from amyloid deposition within the myocardium is a well-described complication; however, an amyloid deposit in pericardium with subsequent pericardial tamponade has rarely been described. We present a case of a patient with both, myocardial & pericardial involvement in primary amyloidosis. Case: 73-year old male with history of hypertension, non-obstructive CAD on recent angiogram, presented with worsening dyspnea on exertion, orthopnea and bilateral leg swelling. Clinically patient was in florid heart failure with elevated blood pressure and physical exam showed distended neck veins, feeble heart sounds, scattered lung rales and 2+ pedal edema. EKG showed non-specific T wave inversion in lateral leads. Cardiac enzymes negative, BNP 892 pg/ml. Chest-X-ray showed increased cardiac silhouette suggestive of pericardial effusion and bilateral pleural effusion. Pt was treated for hypertensive emergency with nitroglycerine and lasix. Bedside Echocardiogram revealed left ventricular (LV) systolic dysfunction (5 months ago LVEF was 55%), severe LV hypertrophy, pericardial effusion with right atrial collapse but no right ventricular collapse on diastole and near tamponade. Emergent pericardial window was placed and patient’s symptoms improved markedly. Repeat Echo showed improved, hyperdynamic LV function, also features suggestive of restrictive cardiomyopathy with sparking quality of LV myocardium. Lasix was discontinued. Serum protein electrophoresis showed M spike and immunofixation revealed IgG lambda monoclonal band. Fat pad biopsy revealed small vessels with amyloid deposition and on Congo red stain, an apple green birefringence was seen on polarization. Pericardial biopsy showed fibrocollagenous deposition negative for malignancy and pericardial fluid was also negative for malignancy. Discussion: Cardiac amyloidosis is more common in men (esp. African Americans) than in women and is rare before age of 40yrs. It is usually seen in primary amyloidosis (plasma cell dyscrasias; as seen in our patient) and can manifests as restrictive cardiomyopathy, systolic heart failure, orthostatic hypotension, or conduction system disease; with most common presentation being CHF. EKG most often reveals low QRS voltage and bundle branch block. Echo shows increased ventricular wall thickness with distinctive sparkling and granular texture, small intracavitary chambers, enlarged atria, and thickened interatrial septum. Amyloid deposits in the pericardium occasionally result in a pericardial effusion, but cardiac tamponade is rare. Because of increased stiffness of the atrial and ventricular walls & high filling pressures in both ventricles, the classical echocardiographic signs of cardiac tamponade, such as right atrial and right ventricular compression, may be absent (as seen in our patient) so one should entertain a high degree of suspicion in a patient with a moderate or large pericardial effusion associated with amyloid heart disease and heart failure. Management includes emergent pericardial window and judicious use of diuretics to avoid systemic hypotension. In conclusion, early recognition of this complication and prompt initiation of therapy is critical.
Introduction: Wilson’s disease (WD) also known as "Hepato-lenticular disease" is an autosomal recessive disorder of copper excretion and can clinically present with hepatic, neurological as well as psychiatric manifestations which makes the diagnosis challenging in most of the cases. We present a case whose atypical presentation led to unmasking of his underlying WD. Case presentation: We present a 32 years old male patient, recently diagnosed with type II diabetes mellitus, who presented with pain and swelling of his right lower extremity. On detailed history, the patient mentioned having 3 month history of intention tremors, sialorrhea, hypophonia, progressive micrographia and family history of 2nd degree relative having WD. Besides having tender swollen right calf, he was found to have advanced intension tremors, cogwheel rigidity in the upper extremities, as well as olfactory dysfunction. His laboratory investigations showed D-dimer of 4335 units, elevated PT (16.5 sec) and INR (1.8), low albumin level (3.0 gm/dl) with normal liver enzymes. Right popliteal acute deep venous thrombosis (DVT) was confirmed by Doppler US. MRI brain showed cortical and cerebellar atrophy. MRI abdomen showed liver cirrhosis, mild splenomegaly and small esophageal varices. Extensive workup proved the diagnosis of WD by low serum copper level of 60 mcg/dl, low serum ceruloplasmin level of 15 mg/dl and high 24 hours urine copper of 211 mcg. Liver biopsy showed macrovascular steatosis less than 5%, grade 3 portal and periportal chronic inflammation, grade 1-2 lobular inflammation, and stage 2 portal fibrosis with intact architecture and negative iron stain. Chemical quantification of copper in the liver biopsy was 219 ug/g drywt. Discussion: WD is caused by mutation of ATP7B gene on long arm of chromosome 13 which codes for a protein responsible for incorporating excess copper into ceruloplasmin to be excreted from the body. Subsequently, this leads to accumulation of copper in several organs mainly the liver, brain and cornea, and inevitably leads to progressive liver and neurological dysfunction [1]. Hepatic manifestations range from asymptomatic biochemical abnormalities up to acute hepatitis or chronic hepatitis and cirrhosis. Neurological manifestations include dysarthria, dystonia, tremors or Parkinson’s like manifestations [2]. Less commonly, WD may present with cerebellar manifestations. Our patient had manifestations of cerebellar involvement as well as Parkinsonian features. This case presents a unique initial presentation of WD in the form of unprovoked DVT secondary to the hypercoagulable state complicating chronic liver disease. As far as we know, DVT has not been reported to be one of the presenting complications of WD especially in patients without advanced liver cirrhosis. References: 1-Purchase R. The treatment of Wilson’s disease, a rare genetic disorder of copper metabolism. SciProg. 2013;96:19-32. 2-Lorincz MT. Neurologic Wilson’s disease. Ann N Y Acad Sci. 2010 Jan;1184:173-87.
Cardiogenic shock secondary to acute chest syndrome in a patient with sickle cell disease. Case report.

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Introduction: Sickle cell disease (SCD) is an inherited disorder that results from the substitution of a valine for glutamic acid as the sixth amino acid of the beta globin chain, producing a hemoglobin tetramer (Hgb S) that is poorly soluble when deoxygenated[1]. Vasoocclusive phenomena and hemolysis are the clinical hallmarks of SCD. We present an unfortunate case that died from severe sickle cell crises.

Case presentation: We present a 29 years old male with SCD (Hgb SS) and history of recurrent vasoocclusive crises including previous episodes of acute chest syndrome (ACS) complicated with mild to moderate pulmonary hypertension, who presented by bilateral extremity pain as well as lower back pain. On presentation he had significant scleral icterus, right upper quadrant tenderness and lower back spinal tenderness. He had Hgb of 7.3 gm/dl, WBC of 14.8 k/ul, AST of 510 u/l, ALT of 278 u/l, Total bilirubin of 17.7 mg/dl and direct bilirubin of 9.3 mg/dl. Later same day, patient developed fever of 39.1°C, chest pain with elevation of troponin levels, worsening of liver enzymes, development of new segmental pulmonary infiltrate and shock. Stat echocardiography showed marked right ventricular dilatation with severe pulmonary hypertension (PAP of 62 mmHg compared to 42 mmHg in 2011). In spite of aggressive hydration and while preparing for exchange transfusion by A positive, E and Kell antigen negative blood type, the patient deteriorated within few hours and expired. Autopsy of the lungs and liver showed microvasculature occlusion by sickled red blood cells, pulmonary hypertension and diffuse patchy pulmonary edema. Autopsy of the heart showed dilated cardiomyopathy with right ventricular myocardium measuring 0.2 cm in thickness.

Discussion: Approximately 50 percent of patients with SCD especially those with Hgb SS will have an episode of ACS in their life [2]. ACS is diagnosed by radiographic evidence of a new segmental pulmonary infiltrate, and at least one of the following: (1) Temperature ≥38.5°C, (2) more than 2 percent decrease in O2 saturation from documented steady-state value on room air, (3) PaO2 <60 mmHg, (4) Tachypnea, (5) Intercostal retractions, nasal flaring or use of accessory muscles of respiration, (6) Chest pain, (7) Cough, (8) Wheezing or (9) Rales [3]. Our patient’s ACS was complicated with severe pulmonary hypertension, acute right ventricular failure and cardiogenic shock. Our patient also had acute hepatic sickle crisis, which has been observed in approximately 10 percent of patients with SCD. Patients usually present with acute right upper quadrant pain, nausea, low grade fever, tender hepatomegaly, and jaundice with elevated liver enzymes and serum bilirubin levels [4].
Rare Case of Immune Reconstitution Inflammatory Syndrome Presenting as Sarcoidosis in Acquired Immunodeficiency Syndrome patient on Highly Active Antiretroviral Therapy

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Rare Case of Immune Reconstitution Inflammatory Syndrome Presenting as Sarcoidosis in Acquired Immunodeficiency Syndrome patient on Highly Active Antiretroviral Therapy. Priyasha Srivastava, Julius Salamera, Christian Chiavetta, Department of Medicine, Seton Hall School of Health and Sciences-Trinitas Regional Medical Center, NJ. Immune Reconstitution Inflammatory Syndrome (IRIS) describes a collection of inflammatory disorders associated with paradoxical worsening of infectious or inflammatory processes upon initiation of Highly Active Antiretroviral Therapy (HAART). In patients with Acquired Immunodeficiency Syndrome (AIDS), worsening of pre-existing infections is seen more commonly as a manifestation of IRIS, while autoimmune or granulomatous conditions are rare. A 54 year old African male was recently diagnosed with HIV and Hepatitis C virus co-infection when he presented with progressive weight loss, anorexia, multiple pulmonary nodules, and hepatosplenomegaly. Further diagnostic tests revealed normocytic anemia, elevated alkaline phosphatase, hypercalcemia, and transaminitis. The HIV viral load PCR was 5100720 copies/ml with a CD4 count of 56 cells/uL. Blood cultures, acid fast bacilli (AFB) blood cultures, serum cryptococcal antigen, and urine histoplasma antigen were negative. A liver biopsy revealed non-caseating granulomata without evidence of malignancy, and AFB as well as fungal cultures were negative. A bronchoscopic examination disclosed no evidence of AFB on multiple smears. He had received treatment for presumed disseminated mycobacterial infection (Mycobacterium tuberculosis versus Mycobacterium avium intracellulare) with isoniazid, rifabutin, ethambutol, pyrazinamide, and azithromycin along with trimethoprim-sulfa for prophylaxis against Pneumocystis jiroveci pneumonia. HAART was initiated 8 weeks after starting the above regimen. The patient was re-admitted four months later with altered mental status, worsening hypercalcemia, acute kidney injury, mediastinal adenopathy, persistent diffuse pulmonary nodules, and multiple splenic lesions. Intravenous fluids, corticosteroids, and calcitonin were administered. A CT-guided biopsy of splenic lesions was compatible with non-caseating granulomata, and fungal as well as AFB cultures were negative. Multiple determinations of serum angiotensin converting enzyme (ACE) were consistently abnormal at greater than 100. A repeat CD4 count was 337 cells/uL and viral load reduced to 821 copies/ml. Hyperparathyroidism and multiple myeloma were ruled out. He was discharged on maintenance corticosteroids with titration based on his symptomatology, serum ionized calcium, and radiographic parameters. Outpatient follow-up shows a stable patient with improved renal function, stable electrolytes, improved HIV parameters, although he developed steroid-induced hyperglycemia. A case of Sarcoidosis involving the lungs and gastrointestinal tract presenting as IRIS is rarely seen in AIDS. Whether such associations represent a causal or coincidental finding is still unproven. Prior to introduction of HAART, progressive Sarcoidosis and advanced HIV infection were considered to be divergent diseases because CD4 cells were believed to be essential in granuloma formation. However, occurrence of Sarcoidosis immediately following HAART, typically after 12 months of therapy, suggests that immune reconstitution may trigger the reactivation of preexisting Sarcoidosis.