Proceedings from the 2007 Annual Meeting of the American College of Physicians, Wisconsin Chapter

INTRODUCTION

The Wisconsin Chapter of the American College of Physicians held its annual meeting in Wisconsin Dells, Wis, September 7-9, 2007. Internal Medicine residents from each of Wisconsin's 5 residency programs (Gundersen Lutheran Healthy System, Marshfield Clinic, the Medical College of Wisconsin, University of Wisconsin Hospital and Clinics, and University of Wisconsin Milwaukee Clinical Campus [Aurora Sinai Medical Center]) presented their research and/or unusual clinical experiences via posters and vignettes. Text versions of the research can be found below. The next Annual Meeting for the Chapter will be held September 12-14, 2008, at the Wilderness Resort in Wisconsin Dells, Wis.

PRESENTED POSTERS

Atraumatic Splenic Rupture

Vijay Aswani, MD, PhD, Maja Visekruna, MD; Marshfield Clinic, Marshfield, Wis

Case: Spontaneous splenic rupture is an extremely rare but life-threatening complication of Infectious Mononucleosis (IM) in young adults. A 25-year-old man presented to the emergency department (ED) with a 3-day history of fever and left-sided pleuritic chest pain. He denied any shortness of breath or recent trauma. Other than an oral temperature of 101.4ºF, his vitals signs were stable. His physical exam was significant for tenderness in the left lower chest wall on deep inspiration and tenderness to palpation in the left upper quadrant. Lab studies revealed leukopenia, thrombocytopenia, and lymphocytopenia. A mononucleosis test done in the ED was negative. D-dimer was elevated at 3.02. A computed tomography (CT) scan of the thorax and abdomen showed no evidence of pulmonary embolism but incidentally revealed an intraperitoneal hemorrhage into the abdomen and pelvis with an enlarged spleen. The patient was hospitalized for observation and remained hemodynamically stable. A CT scan repeated 4 days later revealed a splenic laceration. While Epstein-Barr Virus (EBV) IgG and IgM titers were negative, a polymerase chain reaction done 4 days after admission was positive. EBV IgM titers done 11 days following admission were positive. His fever, symptoms of pain and hematological abnormalities resolved. A CT done about a month later showed interval resolution of the hemoperitoneum and splenic laceration.

Discussion: Splenic rupture is a rare but known complication of IM. It is even more rare as the presenting sign of IM. The case illustrates the need for persistent clinical suspicion when investigating atypical presentations of common illnesses and keeping in mind the specificities and sensitivies of common screening tests.

Brain Drain

Vijay Bandhakavi, MD; Gundersen Lutheran Medical Center, La Crosse, Wis

Case: A 41-year-old woman presented with a 3-week history of occipital and bilateral temporal headaches following chiropractic manipulation of cervical spine for neck pain. The headache was relieved when she was recumbent but became worse when she stood up. She had no other systemic complaints. Her past medical history was significant for rheumatoid arthritis with ongoing Adalimumab therapy. Complete physical examination, including neurological examination, did not reveal any abnormal findings except for those of rheumatoid arthritis. A purified protein derivative test, Lyme serology, human immunodeficiency virus serology and VDRL serology were all negative. A CT scan of the head revealed small bilateral subdural hygromas. Magnetic resonance imaging (MRI) of the head revealed bilateral subdural effusions and diffuse enhancement of the dura, which was interpreted to be consistent with granulomatous meningitis. Multiple attempts to obtain cerebrospinal fluid (CSF), including eventually cisterna magna puncture, were unsuccessful despite appropriate positioning with fluoroscopy. MRI scan of the entire spine did not reveal any evidence of CSF leak. A right parietal burr hole was placed and subdural fluid was retrieved for further analysis. There was no evidence of mycobacterial, fungal, or bacterial infection on histopathology or culture. Histopathology was consistent with spontaneous intracranial hypotension. Her headaches resolved following treatment with lumbar epidural blood patch.

Discussion: We present a challenging case of spontaneous intracranial

hypotension in an immunocompromised patient with rheumatoid arthritis that radiographically mimicked granulomatous meningitis. Spontaneous intracranial hypotension is caused by single or multiple spinal CSF leaks. Women are affected more than men, with a peak around 40 years of age. In our patient, chiropractic manipulation of the cervical spine in combination with her underlying rheumatoid arthritis was felt to cause the CSF leak. An orthostatic headache is the typical manifestation. Associated symptoms include neck stiffness, tinnitus, photophobia, and nausea. Myelography is the study of choice to identify the spinal CSF leak. Typical MRI findings include subdural fluid collections and enhancement of the pachymeninges. Treatments include bed rest, epidural blood patching, and surgical CSF leak repair. Spontaneous intracranial hypotension is not rare but it remains underdiagnosed.

Neurofibromatosis Type I: A Case of Malignant Change

Tracy Brenner, MD; Aurora Sinai Medical Center, Milwaukee, Wis

Case: We present a 39-year-old man with a past medical history significant for Neurofibromatosis Type I (NF1), complaining of a 5-day history of progressive dyspnea on exertion and right-sided back pain. Pulmonary exam showed decreased breath sounds and dullness to percussion involving the entire right lung and at the left base. CT of the chest confirmed a large right pleural effusion and a complete collapse of the right lung. A thoracoscopy was performed that revealed multiple nodules blanketing the pleural space and a completely collapsed right lung in addition to a large effusion. A biopsy and subsequent histology revealed high-grade malignant peripheral fibromas with a confirmation of cell type as a sarcoma.

Two weeks after discharge the patient suffered recurrent, severe dyspnea. Emergency medical service found him to be in asystole. He was intubated and resuscitated and a chest CT was performed showing a complete opacification of the right hemithorax with significant mass effect, which appeared to be tumor burden on the left main stem bronchus, superior vena cava, and right atrium, along with a small right pleural effusion. Also seen was a pulmonary embolism in the left lower lobe. He died 2 days after admission.

Discussion: A recent international consensus statement on Malignant Peripheral Nerve Sheath Tumors (MPNSTs) in NF1 illuminated on the increased incidence of MPNSTs occurring with a significantly higher lifetime risk for the population diagnosed with NF1 versus the general population. MPNSTs are aggressive and potentially fatal soft tissue sarcomas that form in the outer layers of peripheral nerves, which may arise de novo, or from benign plexiform neurofibromas. MPNSTs are rare, with an expected incidence of 0.001% in the general population. However, about 40%-50% of MPNSTs are found in those individuals with a history of NF1. The lifetime risk of developing MPNST in patients with NF1 is documented to be as high as 10%-26%. To date, the only chance for survival is early detection and wide margin surgical resection. The goal of this discussion is to heighten awareness of MPNST in NF1 and thus, have a low threshold to work up any new symptoms.

When Dogs Are Not Man's Best Friend

Amberly Burger, MD; University of Wisconsin, Madison, Wis

Case: A 62-year-old immunocompetent woman presented with 2 days of constitutional symptoms followed by right upper quadrant (RUQ) abdominal pain radiating to the back. Blood cultures were obtained; she was started on broad spectrum antibiotics of ceftriaxone, ciprofloxacin, metronidazole, and ampicillin. Over the next 8 hours, the patient developed petechial rash of the trunk and extremities, purpura fulminans, and profound hypotension requiring 4 pressors. Early goal directed therapy was administered and she was transferred to the Intensive Care Unit at a tertiary care center. Due to RUQ tenderness, elevated liver function tests, and thickened gallbladder wall on ultrasound, digital vascular imaging was consulted and placed a percutaneous drain for control of the presumed source. Patient's illness progressed to septic shock with hypoxic respiratory and kidney failure. She was begun on a small dose of Recombinant Activated Protein C. Patient was given transfusions of blood products for disseminated intravascular coagulation. Patient developed multiple-organ dysfunction syndrome and died within 48 hours of presentation. Blood cultures from the original hospital grew out *Capnocytophaga canimorsus* and *Candida albicans* from her hospitalization in the tertiary care center. The patient owned 1 dog and had no history of dog bites or visible wounds on her body.

Discussion: Capnocytophaga canimorsus is well described in the literature of immunocompromised or asplenic hosts. In immunocompetent hosts it is associated with a known history of dog bite or workers at animal shelters. This disease course, although classic for *Capnocytophaga canimorsus* is rare in this population.

All Choked Up

Michael S. Harris; Medical College of Wisconsin, Milwaukee, Wis

Case: A 19-year-old female Division I swimmer with a history of essential hypertension presented with shortness of breath, chest tightness, and a choking sensation occurring primarily during exertion. She carried the diagnosis of exercise-induced asthma with a frequency of symptoms that placed her in the mild persistent category. Over the 4 years since this constellation of symptoms arose, she was treated unsuccessfully with a procession of bronchodilator and corticosteroid therapies. Management of her concomitant essential hyperten-

sion was complicated by frequent use of tachycardia-inducing beta-2-agonists and reluctance to initiate beta-1-blocker therapy in light of her reactive airway disease. Upon presentation, the patient's methylcholine bronchoprovocation tests were consistent with asthma. Flow volume loops, however, were suggestive not of small airway disease, but an intermittent variable extrathoracic obstructive process. Subsequent laryngoscopy with video stroboscopy revealed a paradoxical adduction of the true vocal cords during inspiration. With a correct diagnosis of Paradoxical Vocal Cord Dysfunction (PVCD), the patient began speech therapy, the mainstay of management for PVCD, and switched to a beta-1-blocker, bringing her blood pressures within normal limits.

Discussion: PVCD is a condition described by William Osler in 1902 in which the glottic space narrows during inspiration rather than demonstrating the normal vocal cord abduction to maximize inspiratory volume. Wheezing, shortness of breath, dyspnea, and stridor are common features of PVCD, and as such, it often masquerades as asthma. A number of etiologies of PVCD have been identified including neurologic, irritant-induced, and somatization/conversion disorder. The most common form, however, is exercise-associated PVCD. The condition has a striking predilection for young, competitive, athletic women, and it is often misdiagnosed as exercise-induced asthma, the treatments for which it is recalcitrant. The average time from presentation to correct diagnosis is 4½ years. In any patient with a history of poorly controlled or atypical asthma, particularly of an exercise-induced variety, PVCD should be considered in the differential diagnosis. Correct identification of PVCD based on subtleties in clinical presentation and volume-flow loops is important for early initiation of appropriate management and avoidance of untoward outcomes associated with unnecessary and ineffectual asthma treatments.

Severe Rhabdomyolysis and Death in a Man with Idiopathic Epilepsy

Aymen M. Khogali, MD; Aurora Sinai Medical Center, Milwaukee, Wis

Case: A 45-year-old African American man with human immunodeficiency virus/acquired immune deficiency syndrome (HIV/AIDS) and idiopathic epilepsy was admitted with severe metabolic acidosis and rhabdomyolysis in context of a 30-minute episode of witnessed seizure. He remained anuric, hypotensive, and hypoxic despite resuscitation with intravenous fluids and pressors. A striking increase in creatine phosphokinase (CPK) (265,000 u/L) and a phosphate of 17.7 mg/dl were noted along with hyperkalemia (7.5 mEq/L) that could not be managed with extra-corporeal therapy as he remained hemodynamically unstable. An autopsy was refused.

Discussion: Non-traumatic rhabdomyolysis has been reported in medical literature due to multiple etiologies. We were unable to find a series of cases in which severe rhabdomyolysis was associated with prolonged seizure.

An Octopus Trap

Gabrielle LeMarbre, MD; University of Wisconsin, Madison, Wis

Case: A 65-year-old woman presented with 1-day history of mild chest pressure. She was visiting her husband in the hospital when she became diaphoretic, nauseated, and short of breath. She experienced palpitations and her chest pressure increased to 6/10. On presentation, her troponin-I was elevated to 5.6. Her electrocardiogram (ECG) showed T-wave inversions in anterolateral leads suggestive of ischemia. Her coronary angiography was unremarkable. Her vetriculogram illustrated apical ballooning and decreased left ventricular function with an ejection fraction of 40%. Follow-up echocardiogram (ECHO) 2 days later demonstrated reduced ventricular function with apical ballooning, supporting a diagnosis of Takotsubo cardiomyopathy.

Discussion: In 1990, Japanese clinicians diagnosed a cardiomyopathy in which the left ventricle resembled the octopus trapping pot used by Japanese fisherman, known as a takotsubo. Takotsubo has been described in the United States population with increasing prevalence. In 1 series, 2.2 percent of patients presenting with ST-segment elevation acute coronary syndrome were diagnosed with Takotsubo. In 1 meta-analysis, 88.8% of patients were women, typically with a preceding psychological or physical stressor. Over 93% of patients were older than 50. Presentation mimics acute coronary syndrome. ECG in the disease can show ST-segment elevations or T-wave inversions. Troponin-I is typically mildly elevated, but does not predict the ventricular function or recovery. For diagnosis, ventriculography or echocardiography must show apical ballooning and akinesis without evidence of significant coronary artery stenosis or spasm. Patients commonly have reduced ejection fraction (20%-49%) that improves within days to weeks. Prognosis is favorable with an in-hospital mortality of 1.1%, but heart failure complicates the outcome in 17.7% of patients. One study suggests that treatment of Takotsubo with beta-blockers, aspirin, ace-inhibitors, and calcium-channel blockers does not improve the ejection fraction in patents at discharge or at 30-day follow-up. This lack of therapeutic benefit emphasizes the importance of prompt diagnosis and appropriate management.

Congratulations! It's a Neutrophil!

Swapna Narayana, MD; Gundersen Lutheran Medical Center, La Crosse, Wis

Case: A 27-year-old woman, 15 weeks primigravida, presented with a 3-day history of low-grade fevers, headache, joint pains, mouth sores, and malaise. She also had developed purple, vesicular skin lesions on her torso, back, and extremities. The skin lesions were initially noticed on

the abdomen and gradually spread towards the extremities. A recent trans-abdominal ultrasound revealed an age appropriate singleton pregnancy. She had a history of multiple sexual partners. She also had a history of multiple outbreaks of genital herpes simplex virus infections in the past. She had not taken any new medications. On physical examination, she had tender, purple, asymmetric, vesicular skin lesions involving the torso, back, and extremities. The physical examination was otherwise normal. Routine blood tests were significant for an elevated white count (14,000) and neutrophilia (90%). A comprehensive metabolic panel, including liver function tests, was normal. An erythrocyte sedimentation rate was 32 mm/hr. An exhaustive workup for infectious diseases including Tzanck smear, gram stain, fungal smear, viral culture and human immunodeficiency virus test was negative. In addition, CSF studies, blood cultures, urine cultures, VDRL, and tests for gonorrhea and chlamydia were all negative. She was empirically started on ceftriaxone and acyclovir. However, the patient's rash and pain clinically worsened despite this treatment. A punch biopsy of the skin lesions was then performed and revealed a superficial and deep, dense inflammatory infiltrate of neutrophils with focal spongiosis of upper dermis. No vasculitic changes were noted. This combination of perivascular neutrophilia with the absence of vasculitis or vascular thrombosis led to the diagnosis of Sweet's syndrome. She was then initiated on systemic steroids, and the skin lesions completely resolved after a week. Systemic steroids were tapered off after 6 weeks.

Discussion: Acute febrile neutrophilic dermatosis, or Sweet's syndrome, was described in 1964 by RD Sweet. It is a rare skin disorder that occurs mostly in women, 30-50 years of age. The importance of correctly identifying this disease lies in its association with other diseases. Pregnancy-induced Sweet's syndrome occurs in 2% of the cases and usually occurs during the first or second trimester. Diseases associated with Sweet's syndrome include inflammatory bowel disease, malignancy, and infections of the upper respiratory tract and the gastrointestinal tract. Treatment of Sweet's syndrome is with oral steroids and usually carries a good prognosis if the disease is not associated with malignancy.

More Than Weakness

Vipulkumar Rana, MD, Maria Delgado, Kurt Pfeifer, MD, FACP; Medical College of Wisconsin, Milwaukee, Wis

Case: A 47-year-old man with no past medical history presented with a 4-day history of proximal lower extremity weakness. Additionally, he reported a 25-pound weight loss over the past 4 months and a 2-month history of generalized weakness, fatigue, dizziness, and headaches. He was admitted to the neurology service, and evaluation revealed non-focal, 4 of 5, proximal, give-way weakness, with normal muscle bulk and tone. Electromyogram and head CT were also within normal limits, and infectious work-up was unremarkable. The patient was discharged with the diagnosis of fatigue of unknown etiology, only to return 5 days later with worsening of his previous symptoms, inability to bear weight, and mild diffuse abdominal pain. On initial evaluation, he had normal vitals signs and basic laboratory studies, and the symptoms were attributed to a psychological cause. However, after further work-up, adrenocorticotropic hormone stimulation test was performed and the diagnosis of adrenal insufficiency was made. Glucocorticoid and mineralocorticoid replacement was started with marked improvement of all symptoms.

Discussion: Adrenal insufficiency frequently presents with an insidious onset of weakness and fatigue that can dramatically progress to the point of disability. While a common presentation includes weakness, fatigue, and weight loss as seen in this patient, many physicians dismiss the diagnosis in a patient without hypotension or electrolyte abnormality. It is a common misconception among physicians that a patient cannot have adrenal insufficiency without hypotension and/or electrolyte abnormalities. Maintaining a high index of suspicion for the diagnosis of adrenal insufficiency in the absence of these findings may lead to earlier diagnosis, faster patient recovery, and avoidance of unnecessary testing and patient stress.

Unusual Etiology of Lung and Liver Nodules in a Smoker

Jason Ricco, BS, Anna Haemel, MD, David Feldstein, MD; University of Wisconsin, Madison, Wis

Case: A 60-year-old woman with a 30-pack-per-year smoking history presented to the ED with progressive fatigue, dyspnea on exertion, and mild hemoptysis. She also complained of anorexia, a 15-pound weight loss, small joint arthralgias, and decreased sensation in her lower extremities, all developing over 1 month. Examination revealed cachexia, crackles at the left lung base, and decreased light touch in the lower extremities. Initial laboratories included mild anemia and creatinine 1.9. Urinalysis showed proteinuria, hematuria, and granular casts. CT revealed numerous bilateral pulmonary nodules, some with cavitation, and several low attenuation liver lesions not amenable to biopsy. Biopsy of the most prominent lung lesion showed histiocytes and scattered giant cells with a necrotic background. Renal biopsy showed pauci-immune crescenteric glomerulonephritis and C-Anti-Neutrophil Cytoplasmic Antibody (ANCA) came back positive. The initiation of cyclophosphamide and prednisone resulted in improvement of her systemic complaints and kidney function.

Discussion: This patient was initially thought to have metastatic cancer due to her smoking history, pulmonary nodules and liver lesions consis-

tent with metastases. Her arthralgias and paresthesias raised the question of paraneoplastic syndrome. The granular casts on initial urinalysis misled the team to believe she had acute tubular necrosis. However, the unresponsiveness of her acute kidney injury to fluid resuscitation led to a unifying diagnosis of Wegener's granulomatosis.

Wegener's classically presents as granulomatous vasculitis of the small vessels of the respiratory tract with accompanying glomerulonephritis. The extent of disseminated vasculitis is variable. However, case reports of hepatic involvement are limited. Differential diagnosis of the nonspecific low attenuation liver lesions seen on CT includes metastases, abscesses, or granulomas. Although this patient's liver function remains within normal limits, the liver lesions may represent granulomas. This case exemplifies the diverse presentation of Wegener's and the high index of suspicion imperative in any patient with pulmonary nodules and renal dysfunction.

Phenytoin-Induced Dress Syndrome

Matthew A. Schmidt, MD, Kurt Pfeifer, MD, Amandeep Gill, MD; Medical College of Wisconsin, Milwaukee, Wis

Case: A 50-year-old man presented with nausea, vomiting, and abdominal pain after recently being admitted to an outside hospital for hypertensive emergency, hemorrhagic stroke, and seizure. He was treated in the intensive care unit, and started on phenytoin 900 mg 3 times daily for seizure prophylaxis. Following inpatient rehabilitation at the outside hospital, he was discharged home and then admitted to our hospital 1 day later. Initially, he was treated for suspected partial small bowel obstruction given radiographic findings of dilated loops of small bowel. The next day the patient developed a maculopapular rash on his chest and back, and phenytoin was discontinued as it was the suspected cause. His rash spread to his limbs, palms, soles and head, covering his entire body. At this time, he was also noted to have eosinophilia, and the diagnosis of Drug Reaction, Eosinophilia, and Systemic Symptoms (DRESS) was made. Following several doses of intravenous corticosteroids, his symptoms improved markedly, and his rash began to resolve. Levitiracetam was started in place of phenytoin for seizure prophylaxis, and he was discharged home on a tapering dose of steroids with dermatology follow-up.

Discussion: DRESS syndrome is associated with several antiepileptics including phenytoin, carbamazepine, and phenobarbital. The incidence with phenytoin is approximately 1 in 1000 to 1 in 10,000, and the hypersensitivity reaction usually occurs 2-6 weeks after the drug is first introduced. Patients typically have fever, rash, lymphadenopathy, and hepatitis, and less commonly interstitial nephritis, pulmonary involvement, and eosinophilia. When the viscera are involved, it can become potentially life-threatening. Standard treatment is discontinuation of the offending agent, intravenous hydration, antihistamines, and possibly corticosteroids, depending on the severity of reaction.

Hyperosmolar Therapy: The Good, the Bad, and the Ugly

Jayanth Vedre, MD, Jasjyot Nanra, MD, Anupama Inaganti, MD, Narayana, S. Murali, MD; Marshfield Clinic, Marshfield, Wis

Case: Mannitol, an osmotic diuretic, occupies a preeminent position in the therapeutic armamentarium of intracranial hypertension and acute angle closure glaucoma. Albeit uncommon, like all good therapies, this therapy too is not impervious to malevolent side effects. We report an 82-year-old man who presented with acute onset, painful right red eye with loss of vision, and elevated intraocular pressure with a subchoroidal hemorrhage. He was administered high dose intravenous mannitol in the inpatient setting. Baseline creatinine of 1.0 mg/dL (estimated glomerular filtration rate[GFR] 87.4ml/min) and sodium of 136 mmol/L. On day 3 of his treatment, he was noted to have mild hyponatremia (130 mmol/L) and lisinopril treatment was initiated to address hypertension (systolic blood pressure 170-180 mmHg). On day 4, he was slightly somnolent, profoundly bradycardic (27 bpm) with an ECG revealing a very slow ventricular response, peaked T waves, and was immediately addressed by temporary pacing. His laboratory investigations revealed marked hyponatremia (115 mmol/L), hyperkalemia with acute kidney injury that rapidly progressed to severe oliguric renal failure (creatinine, 2.6 to 4.5 mg/dL, eGFR 13.6 ml/min) over the next 12 hours. Measured serum osmolarity was 334 mOs/kg; calculated osmolality was 255, with an osmolar gap of 79. Mannitol and Lisinopril were immediately discontinued. He underwent Continuous Renal Peplacement Therapy (CRRT) to lower the serum levels of mannitol. CRRT was discontinued on Day 5, when measured serum osmolality was 309 mOs/kg. His urine output improved remarkably in less than 24 hours with prompt recovery of renal function. Cardiac rhythm returned to baseline and pacing was discontinued.

Discussion: This is the second such reported case in the English medical literature of acute reversible oliguric renal failure related to use of high-dose mannitol and Angiotensin Receptor Antagonists (ATRA). It illustrates not only the entire spectrum of complications of mannitol therapy ranging from severe hyperosmolar hyponatremia, acute oliguric renal failure, to atrioventricular block, but also showcases clinical prudence of avoiding ATRAs with hyperosmolar therapy on sound physiological principles. High dose mannitol incites intense vasoconstriction of afferent arterioles; decreasing GFR, which is compounded by concomitant ATRA-induced efferent arteriolar dilatation, further diminishing GFR leading to acute renal failure.

DISPLAYED POSTERS

Misleading Presentation of a Pulmonary Artery Arcoma

Muhammad Bakr Ghbeis, MD, William G. Hocking, MD; Marshfield Clinic, Marshfield, Wis

Case: We evaluated a 48-year-old woman who presented with increasing exertional breathlessness over 4 weeks. She had a persistent nagging dry cough 5 weeks earlier, following a slowly resolving cold. She reported right calf pain, 2 weeks earlier, for 2 to 3 days. She saw her primary care provider, who performed a chest CT scan, indicated by an elevated D-Dimer. The scan was interpreted as a "massive pulmonary arterial embolus." Subsequently, she was admitted to our critical care unit. There was no evidence of hemodynamic instability. A physical examination revealed no abnormalities except for obesity (body mass index of 41) and superficial varicosities in the lower extremities. The patient was started on anticoagulation therapy with unfractionated heparin and warfarin. She was discharged 6 days later, with instructions to continue low-molecular-weight heparin and warfarin.

One day after discharge, the patient returned to the ED for increasing breathlessness. A repeat CT scan was interpreted as persistent massive clot in the pulmonary arterial tree. There was no evidence of hemodynamic instability. Bilateral lower extremity venous Doppler ultrasonography showed no evidence of thrombosis. The abdominal and pelvic CT scan showed no evidence of malignancy or thrombosis. Despite active anticoagulation for 14 days, a repeat CT scan demonstrated no improvement, and the possibility of alternative diagnoses were considered. Positron emission tomography (PET)/CT imaging showed no evidence of hyper-metabolic lesion in neck, chest, abdomen or pelvis.

The patient underwent a thoracotomy via a midline sternotomy. A mass was found arising in main pulmonary artery, extending to both left and right pulmonary arteries. The pulmonary artery wall and the anterior pulmonary valve leaflet were grossly invaded by the tumor. A subtotal resection was performed with reconstruction of the pulmonary valve. Pathology showed a high-grade intimal sarcoma. The patient is recovering from surgery. Chemotherapy followed by radiation is planned.

Discussion: Pulmonary artery sarcomas are rare neoplasms and often cause symptoms suggestive of recurrent pulmonary emboli. A diagnosis of pulmonary artery sarcoma is virtually never considered initially. In patients with presumed thromboembolic disease, certain clinical and imaging characteristics may suggest the alternative diagnosis of pulmonary artery sarcoma.

Malaria

Mark Gaulke, MD, Matthew Hall, MD; Marshfield Clinic, Marshfield, Wis

Case: A 20-year-old man with no significant past medical history was transferred to our facility for management of malaria. He had presented 4 days prior to another facility with fever (103°F). He complained of having fever, headache, nausea, fatigue, and abdominal pain but denied weight loss, jaundice, diarrhea, or cough. He was treated overnight with intravenous fluids and doxycycline for a presumed tick borne illness (Lyme or Anaplasmosis). He returned within 24 hours of discharge with continued fever (106°F) and experiencing hallucinations. After 3 blood smears at the outside institution, a blood smear was found to have organisms consistent with malaria. On further questioning, the patient was noted to have traveled to New Guinea and Australia 2 months earlier with a traveling companion known to have contracted malaria. Prior to transfer, his platelet count was known to be anemic (11.5), thrombocytopenic (67,000) and to have splenomegaly. The patient was treated with 650 mg oral Quinine sulfate every 8 hours along with the doxycycline that he was already taking (100 mg twice a day). Upon discharge he was started on 15 mg Primaquine for 15 days. His follow up was scheduled 3 weeks later during the most likely time of reoccurrence. If needed treatment could be restarted. Otherwise the patient was to restrict activity until spleen size normalized. The patient was also educated to watch for relapses that are most likely the first 4 months and very unlikely after 1 year.

Discussion: Malaria is caused by *Plasmodia falciparum, P. ovale, P. malariae,* and *P. vivax. P. falciparum* is the most likely to kill due its unique vasooclusive characteristic. Symptoms begin 1-4 weeks after infection during the erythrocytic stage. *P. vivax* infect only early reticulocytes, approximately 2% of red blood cells and no microvascular sequestration occurs.

A Case of Cryptogenic Stroke in the Setting of a Patent Foramen Ovale (PFO) and Factor V Leiden Heterozygosity

Esayas Gebreyesus, MD; Aurora Sinai Medical Center, Milwaukee, Wis

Case: A 27-year-old man presented to our ED with an acute onset of vertical double vision and oscillopsia. Patient was driving when he suddenly experienced this. His past medical history includes history of 3 different eye muscle surgeries for correction of a head shift to the left, eyes right at the age of 9 or 10; gun shot wounds; recent history being dragged 4 blocks by a moving vehicle with scalp laceration and hand injury; and a back pain secondary to a motor vehicle accident a year ago. There is no past history of thromboembolic events. Upon admission, he had the following vital signs: temperature=99.1°F, rest rate=16/min, blood pressure=140/97mm/Hg, heart rate=82/min and pulse oximetry of 97% at room air. Physical examination showed a patch over the left eye. Upon removal of patch, patient had coarse horizontal nystagmus increased greatly in gaze left. Also on gaze right, he had gaze palsy to the right, which didn't improve on dolls-

head, and developed right hypotropia. Up gaze was impaired more than down gaze. Pupils were equally reactive. Visual acuity without correction was 20/25 each eye separately. Tonometry by applantation was 13/15. Fundoscopy examination was benign. Other motor/sensory examination was normal. Electrolytes, complete blood cell count, cardiac enzymes, and blood glucose were normal. Urine tox screening was negative. Coagulation workup revealed heterozygocity for Factor V Leiden (FVL) and low-normal AT 3 level. Magnetic resonance angiography of the head showed 2 areas of acute ischemia in the left thalamus and left brainstem. Transesophageal Echocardiography showed a Patent Foramen Ovale (PFO) and right-to-left shunt (RLS) on Valsalva maneuver. Patient was discharged to inpatient rehab with anti-platelets agents and his PFO was later closed.

Discussion: PFO is a frequent remnant of embryological development with clinical importance in thromboembolism, paradoxical embolism, and stroke. A meta-analysis of 9 studies showed a significantly higher prevalence of PFO in cryptogenic stroke (55.7%) as compared with stroke of known etiology (17.1%), giving a significant Odds Ratio of 6.0. Harvey et al made their first study of young patients with cryptogenic stroke and found evidence of RLS in 73% of cases. Some studies found FVL mutation in 7% of the patients with cryptogenic brain infarction and PFO, but in only 1% of the controls. Some studies estimate a 3-fold increase in the risk of thromboembolism with factor V heterozygosity.

Conclusion: A constellation of FVL heterozygosity and PFO with RLS on TEE could be an adequate explanation of this young patient with arterial embolism/cryptogenic stroke.

Painless, Acute Aortic Dissection Presenting as Seizures

Naga Grandhe, MD, Ravi Mareedu, MD, Kenneth P Madden, MD, Steven Gilbert, MD; Marshfield Clinic, Marshfield, Wis

Case: An apparently healthy 53-year-old man presented with recurrent episodes of witnessed seizures at 3 AM and was intubated for airway protection. Past medical history and social history was unremarkable except for reflux disease and occasional low back pain. Physical examination revealed stable vital signs but he was non-responsive to verbal commands. His neurological exam showed symmetrical reflexes and responsiveness to pain in all extremities except for the left lower extremity. Initial laboratory evaluation was unremarkable. CT of the head and cerebrospinal fluid analysis was unremarkable. Patient was loaded with phenytoin in the interim. Electronenceophalogram showed diffuse encephalopathy. MRI of the head showed multiple ischemic areas predominant on the right side. Stroke and infective endocarditis were considered in the differential at this time. CT abdomen done for evaluation of infectious source revealed aortic dissection extending from the aortic root through descending aorta and terminating at infra renal aorta. A 2-D ECHO confirmed the dissection and did not show any valvular vegetations. Carotid Dopplers showed a complete occlusion of the right carotids and mild stenosis on left side. A cardiothoracic surgeon evaluated the patient and recommended medical management because of the severe cerebral encephalopathy. As there was no improvement in neurological status after 72 hours of observation, patient was transferred to palliative care services per family wishes. Patient expired after withdrawing support. Autopsy did not show any evidence of infective endocarditis.

Discussion: Around 5%-15% of aortic dissections present without pain as per previous studies. Neurologic manifestations as a presenting symptom of acute aortic dissection without chest pain are uncommon. There have been case reports of seizures as one of the neurological manifestations of acute aortic dissection in literature. To our knowledge, there are no case reports of painless acute aortic dissection presenting as seizures. In our case, the occurrence of ischemic stroke could have precipitated the seizures.

Presentation of painless aortic dissection in an individual in their early 50s with no prior history of systemic diseases is also a unique feature in our case. Because of the absence of pain, the diagnosis of painless aortic dissection is usually delayed, as in our case. Even though isolated neurological manifestations are rare presentations of acute aortic dissection, physicians should always suspect this condition as it is associated with high mortality.

Pure Coincidence or Serendipitous Causality: Splenic Lymphoma and Hepatitis C Virus

Anupama Inaganti, MD, Jayanth Vedre, MD, Narayana S. Murali, MD; Marshfield Clinic, Marshfield, Wis

Background: Chronic Hepatitis C Virus (HCV) infection is the most common etiology of chronic viral hepatitis in the United States, afflicting 4 million in the United States alone. While everyday extra-hepatic manifestations of HCV are familiar to most physicians, the intriguing association with splenic lymphoma is poorly recognized largely due to its rarity. However, it is clinically important as it has provided valuable insight into lymphoma genesis and therapy.

Case: A 56-year-old woman presented with acute severe abdominal pain. Her history included chronic HCV genotype 1a, a recent bout of relapsing auricular polychondritis, coronary artery bypass surgery, stroke and end-stage renal disease on hemodialysis attributed to unusually rapid deterioration of diabetic nephropathy. Exam was notable for severe left hypogastric tenderness and massive splenomegaly with normal bowel sounds. CT revealed splenic enlargement to the pelvic brim, greater than 21 cm pole to pole (splenomegaly=10 cm), with a 3.5 cm decreased attenua-

tion consistent with splenic infarction and regional adenopathy. Doppler ultrasound revealed normal flow through hepatic vasculature without evidence of portal hypertension. A peripheral blood-flow cytometry was consistent with a B-cell lymphoproliferative disorder and a bone marrow biopsy was normal. HCV quantitative polymerase chain reaction titers were above the upper analytical limit of positivity. The diagnosis of splenic lymphoma was confirmed on excision biopsy, which revealed cyclin D1 positive mantle cell splenic lymphoma.

Discussion: Splenomegaly in the setting of HCV should trigger workup for splenic lymphoma—the clinical immediacy of such recognition is particularly relevant given recent compelling therapeutic evidence of remission with relatively benign antiviral therapy. The unusually rapid deterioration of biopsy proven Diabetic Nephropathy in the setting of well-controlled blood sugars (hemoglogin A1C less than 6) highlights not only the association of HCV to diabetes but also its documented role in accelerating diabetic kidney disease. Relapsing polychondritis in HCV, while very unusual, has been reported to have remitted with ribavirin and pegylated interferon therapy therapy and has been linked to the propensity of HCV to trigger or exacerbate autoimmune disorders.

The Clot is Just the Beginning of the Plot!

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Case: A 43-year-old woman was admitted with sepsis secondary to chronic non-healing ulcers. She had a history of profound diabetic peripheral vascular disease and underwent a below-the-knee amputation during her hospitalization. Three days later, she developed confusion and thrombosis of her central venous lines. Her past history was also notable for end-stage renal disease with a long history of hemodialysis and recurrent thromboses of her dialysis lines. A complete physical exam, at that point, was notable for a stump wound eschar along with palpable purpura involving both the lower extremities and the abdomen. She also had prominent swelling over her face and both upper extremities. Lab studies revealed an elevated parathyroid hormone at 91 pg/ml, an elevated thyroid stimulating hormone at 10.9 μ/ml, and an elevated ionized calcium at 1.4 mmol/L. She also had low platelet at 63 K/uL, low Protein C and S levels and activity at 24% and 21% respectively, an elevated prothrombin time (PT) at >8.0 and partial thromboplastin time (PTT) at >240 sec and abnormal PTT mixing study. A venous ultrasound of the upper extremities and neck revealed bilateral internal jugular vein thromboses.

Further evaluation for hypercoagulable states revealed the patient had lupus anticoagulant. A skin biopsy was performed and revealed epidermal and dermal necrosis associated with calcifications in the walls of the subcutaneous small blood vessels and neutrophilic vasculitis in the medium size blood vessels. This was felt to be consistent with calciphylaxis with a secondary autoimmune vasculitic process. Despite treatment, she eventually progressed into severe disseminated intravascular coagulation and sepsis along with multiorgan system failure and died within a month of her initial presentation.

Discussion: There have been several well-documented cases of lupus anticoagulant and calciphylaxis in patients with end-stage renal disease on hemodialysis. Having both of these conditions simultaneously poses a serious challenge for medical management. Calciphylaxis is a serious skin disorder characterized by small vessel calcification and necrosis in the dermis or subcutaneous fat. Ischemic changes often progress to necrotic ulcers that become superinfected with a resultant 60%-80% mortality. The incidence of calciphylaxis is only 1% in the dialysis population. However, there were multiple risk factors for calciphylaxis present in this patient, including an elevated ionized calcium, obesity, significant weight loss, low serum albumin, female sex, low protein C and S, multiple renal dialysis catheterizations, warfarin therapy, and calcium with Vitamin D supplementation.

Conclusion: This case points out the need for high clinical suspicion for the serious but preventable complications in dialysis patients and the awareness of various factors influencing management in this subset of patients.

Helicobacter Pylori Seropositivity in Patients with Both Negative Rapid Urease Test (CLO) and No Histopathological Evidence of Hp

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Purpose: Gastric biopsies are obtained routinely during esophogastroduodenoscopy (EGD) for campylobacter-like organism test (CLO) and histology; however, questions have been raised about a decreased diagnostic yield of CLO and histology in detecting this organism compared to the original published reports for several reasons, including a gradual decrease in *Helicobacter pylori* (Hp) infection over the past decade and increase in the use of acid-reducing medications. The aim of this study was to investigate the role of Hp serology in patients with negative CLO and no histological evidence of Hp.

Methods: Seven hundred seventy-six consecutive patients (age 18-95) who underwent a CLO test at a tertiary care center in the Midwest between July 2005-December 2006 were identified. Fifteen patients were excluded due to lack of CLO test verification and 5 due to lack of availability of EGD report. We reviewed the CLO results, EGD reports, available histology, and aspirin/nonsteroidal anti-imflammatory drug (NSAID) use. Serology was then compared to CLO and histology in the detection of Hp in patients with and without ulcers and erosions detected on EGD. The results were analyzed with the bino-

mial distribution using serology test specificity of 92% provided by the manufacturer (Meridian).

Results: A total of 756 patients were studied: 441 (58.3%) males and 315 (41.7%) females (mean age: males, 59.86 and females, 61.93 years). Most patients were caucasian (669; 88.5%) with more outpatients (411; 54.4%) than inpatients (345; 45.6%). CLO was positive in 52 (6.88%) patients and negative in 704 (93.12%) patients. Hp serology was available in 91 patients, of which 16 had it done more than 1 year prior to CLO. In the remaining 75 patients, no gastric biopsies were done on 25 patients. Of these 50, 49 patients had negative CLO and histopathology, but 9 (18.4%) had positive Hp serology. Exclusion of patients with Hp serology done >4 weeks before or after the CLO identified 26 patients of which 6 had positive Hp serology with this proportion (6/26) being significant, greater than the false positive rate ($p<0.05$).

Conclusion: CLO is positive in a small percentage of patients undergoing EGD. Negative CLO and histology may not be sufficient to exclude infection with Hp. Prospective studies assessing the role of Hp stool antigen testing and polymerase chain reaction for Hp16S ribosomal DNA on gastric tissue are warranted in this patient population.

PFO with Right to Left Shunt as a Cause of Hypoxia

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Case: A 58-year-old woman presented with a 9-month history of shortness of breath and a 1-month history of bilateral lower extremity pedal edema with baseline oxygen (O_2) saturation in the 60s to 70s. She exhibited central cyanosis and elevated jugular vein distension (JVD). The patient's O_2 saturation measured 66% on 2 liters of O_2 and increased to 73% on 5 liters of O_2. (ECHO) showed evidence of severely enlarged right atrium and right ventricle, depressed right ventricular systolic function, normal left ventricular systolic function, continuous flow to left atrium from an unknown source and pulmonary artery trunk (PA) pressure of around 40-45 mmHg. Transesophageal ECHO (TEE) showed marked right heart enlargement and right to left shunting across a probe patent foramen ovale (PFO). Catheterization confirmed presence of low femoral artery saturation (82%) with normal left atrial saturation (100%). PA pressure was moderately elevated at 40/20/28 mmHg with normal capillary wedge pressure at 8 mm Hg. To evaluate for tolerance of right heart after closure, temporary occlusion of the PFO with the sizing balloon was attempted, and after the patient was found to be stable, a 23 mm CardioSeal device was subsequently placed in the PFO without complications. The patient's O_2 saturation was ranging between 85%-93% on 5-6 liters of oxygen in the first 24 hours. At 1 month follow-up the patient showed significant improvement in functional status with O_2 saturations of 82% on room air. She continued to be stable at 6 months post procedure.

Discussion: Hypoxia secondary to right to left shunt (without Eisenmengers physiology or elevated Pulmonary Artery pressures) is an uncommon presentation. Initial diagnosis via transthoracic ECHO requires detection of a shunt with either color Doppler or agitated saline contrast with or without Valsalva maneuver. The agitated saline contrast study with TEE and the Valsalva maneuver is the gold standard test for detection of PFOs. It is sometimes simple to find the trigger for right to left shunt, but in patients such as ours in whom the PA pressures were not significantly elevated, there is no easily-identifiable single cause. Multiple theories have been postulated to explain severe shunting that can lead to hypoxia. Causes could be transient elevation of right atrial pressure in each cardiac cycle, or the flow of blood from the inferior vena cava preferentially towards the PFO (and inter-atrial septum), similar to the circulatory pattern in the fetus, or decreased compliance of the right ventricle in comparison to the left ventricle. Mechanical closure is clearly indicated in significantly hypoxic patients.

Lyme Meningitis

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Case: A 59-year-old woman presented with 1-week history of nonspecific complaints of fever, fatigue, and generalized weakness. There was no history of any headaches, neck stiffness, photophobia, dizziness, or changes in vision or hearing. She had slight nausea but denied any history of arthralgias, skin changes, or rashes. Patient was from rural Georgia and enjoyed gardening and being outdoors. She was visiting Milwaukee to see her children. She also denied any history of tick bite. Her physical examination was unremarkable, except for temperature of 103ºF. No evidence of any skin rashes. Complete blood cell count, blood cultures, urine analysis, and chest X-ray was unremarkable. Echocardiography was unremarkable. Patient continued to have high-grade fevers 103ºF-105ºF and had some episodic confusion with picking at clothes and medical equipment. Lumbar puncture and MRI of the brain were performed. CSF demonstrated white blood cell count 265 with 10% neutrophils, 81% lymphocytes, glucose 73 mg/dL and protein of 110 mg/dL. Gram stain and culture, viral cultures were negative. Enterovirus and herpes simplex virus in CSF were not detectable. Serum Lyme IgM antibody was positive but Lyme IgG antibody was negative. Diagnosis of Lyme meningitis and mild encephalopathy was established. Patient was treated with ceftriaxone 2 grams intravenously with resolution of fever and improvement of symptoms.

Discussion: Lyme disease is a multisystem infectious disease caused by tick-borne spirochete *Borrelia burgdorferi.* Clinical manifestations most often involve the skin, joints, nervous system, and heart. Extracutaneous manifestations are <10% of cases. When Lyme borreliosis affects the nervous system, it typically presents with (1) all or part of a

triad-meningitis, cranial neuritis, and radiculoneurirtis; (2) parenchymal inflammation of the brain or spinal cord; (3) mild radiculoneuropathy or (4) encephalopathy with or without evidence of brain infection. Lyme meningitis is a manifestation of the early-disseminated Lyme disease (2-10 weeks after tick bite). CSF has a lymphocytic pleocytosis, elevated protein, and normal glucose level. Diagnosis of Lyme disease requires confirmation by Western blot analysis after detection of a positive enzyme-linked immuosorbent assay. For adult with early Lyme disease and the acute meningitis, the use of ceftriaxone 2 g per day intravenously for 14 day (range, 10-28 days) is recommended.

Ca 19-9, a Pancreatic Tumor Marker?

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Case: A 61-year-old woman was brought to the ED for nausea, vomiting, and dizziness. She denied any abdominal pain, fever, diarrhea or hematemesis. She had no history of smoking or alcohol use. She reported a weight loss of around 18 pounds in last 2 months. Past medical history included diabetes mellitus, hypertension, and mild mental retardation. Physical examination revealed hemodynamically stable female with jaundice. Abdomen was non-tender with no peritoneal signs. Rest of the exam was normal. Initial laboratory evaluation revealed leukocytosis with bandemia, conjugated hyperbilirubinemia, and acute renal failure (presumed to be from biliary sepsis). Ultrasound of abdomen was suggestive of intra and extra hepatic biliary dilation. The gallbladder was non-tender, long and tortuous with debris and with very slight wall thickening. Acute renal failure precluded a CT examination. Due to the high suspicion of pancreatic-biliary malignancy, CA 19-9 level was checked and was found to be significantly elevated (8211). Further evaluation with endocopic retrograde cholangiopancreatography showed common bile duct stone and sphincterotomy was done. After her renal failure resolved, CT abdomen was done and did not suggest any malignancy. Colonoscopy and repeat liver function tests done during follow up were normal.

Discussion: CA 19-9 is considered a useful tumor marker for pancreatic cancer, and it has sensitivity of 70%-90% and specificity of around 90%. Higher levels of CA 19-9 (more than 1000) are associated with surgically unresectable cancer. Our case provides evidence that significantly elevated CA 19-9 level can be in benign biliary, pancreatic, and liver diseases. The non-malignant conditions associated with high Ca 19-9 reported are alcoholic liver disease, primary sclerosing cholangitis, primary biliary cirrhosis, hepatitis, acute cholangitis, and gallstones.

Familial Hypokalemic Periodic Paralysis

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Case: A 41-year-old woman was admitted with sudden onset of generalized muscle aches and weakness after recent sinus surgery. She had similar episodes in the past. Family history was unobtainable as patient was adopted. Examination was significant for generalized weakness with muscle tenderness. She had elevated muscle enzymes and slightly low potassium (3.2 mEq/L). A neurologist, psychiatrist and a rheumatologist reevaluated the patient. Head and spinal MRI along with antinuclear antibody, C3, C4, C-reactive protein, and Rheumatoid Factor were unremarkable. Patient continued to suffer similar episodes along with myoclonus involving the left upper extremity mostly accompanying asthma exacerbations. Due to uncertain diagnosis, patient was given trial of acetazolamide and muscle biopsy was performed, revealing muscle fibers with rimed vacuoles, some atrophic fibers and no inflammatory infiltrates. Biopsy along with clinical improvement confirmed hypokalemic periodic paralysis.

Discussion: Hypokalemic periodic paralysis is an uncommon but life threatening clinical syndrome. Most cases are familial. This disease usually presents in early childhood but may also present in the third decade. Paralysis results from ion channel mutations or channelopathies. Familial forms are associated with muscle calcium, potassium, or sodium channels defects. The mechanism during paralytic attacks is a transient membrane depolarization that inactivates the sodium channels along with K+ shift into the muscle cells. Severe attacks are usually precipitated in the morning, with strenuous exercise or with large carbohydrate diet. Weakness may occur with minimal hypokalemia. Creatine phosphokinase rises during episodes.

One of the most informative diagnostic tests is an exercise test. Compound Muscle Action Potential (CMAP) is usually measured during and after exercise. After a brief increase in CMAP amplitude, decrease of more than 40% is considered abnormal. This test is 98% sensitive.

Recent studies have shown that measuring Trans-tubular concentration gradient (TTKG) and potassium-creatinine ratio (Polymerace chain reaction) distinguishes primary from secondary causes of periodic paralysis. Most characteristic biopsy finding is the presence of vacuoles in muscle fibers and tubular aggregates. During attacks, the main objective is to normalize serum potassium levels. Oral potassium is preferable to intravenous. Continuous ECG monitoring and serum potassium measurements are mandatory. For prophylaxis, Acetazolamide is administered. Low sodium and low carbohydrate diet decreases the frequency of attacks.

Mycotic Aneurysm in a Patient With Infective Endocarditis

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Case: A 63-year-old man initially presented with a 2-month history of headaches, generalized malaise, decreased appetite, sweats, fevers, and weight loss. He had a history of

colon cancer with successful resection a number of years ago, a hemorrhagic stroke in 1973, and an ischemic stroke in 2003. In addition, his past medical history was significant for subacute bacterial endocarditis, for which he had failed to keep several follow-up appointments. A complete exam was performed and revealed no new neurological deficits. Initial studies revealed an elevated erythrocyte sedimentation rate of 49, white blood cell count elevation at 15,400 and hematuria. A CT scan of the head was performed and revealed an acute right hemorrhagic stroke. He then underwent cerebral angiography, which showed a new, lobulated irregular aneurysm of the distal right internal carotid artery, which was 9 mm in size. A left internal maxillary artery aneurysm was also seen. Blood cultures eventually grew *Streptococcus bovis.* Therefore, he underwent an ECHO, which demonstrated bileaflet mitral valve prolapse with posterior leaflet rupture and vegetations on mitral valve, consistent with endocarditis. He also developed acute renal failure, which was felt to be related to his infective endocarditis. It was determined that he had mycotic aneurysms, which were nonoperative in nature and therefore, medical management was initiated. However, the patient decompensated hemodynamically and required transfer to the intensive care unit for stabilization. Over the next several days, despite vigorous medical management, he had 2 grand mal seizures and a sudden loss of consciousness. A CT scan of the head was again performed and demonstrated an extensive new right basal ganglia and right frontal lobe intraparenchymal hemorrhage. At this point the family decided on comfort care and he died the same day.

Discussion: Mycotic aneurysms develop due to an infection in the arterial wall. They are extremely rare. One important predisposing factor is bacterial endocarditis. In fact, only about 2%-3% of intracranial aneurysms are mycotic in origin. Signs and symptoms of mycotic aneurysms may often be misleading during the early stages, resulting in misdiagnosis and delay in treatment. Mortality is greater than 90% in untreated patients. Subacute bacterial endocarditis can cause membranoproliferative glomerulonephritis from immune complex deposition. This patient had history of colon cancer with bacteremia due to *Streptococcus bovis.* This resulted in bacterial endocarditis, which eventually led to complications including mycotic aneurysms and acute renal failure.

Juvenile Rheumatoid Arthritis in an Adult?

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Case: Juvenile Rheumatoid Arthritis (JRA) is the most common form of persistent arthritis in children. It is also seen in adults, where it is known as Adult Onset Still's Disease. A 36-year-old man presented to his physician's office with episodic high fevers, weight loss, a sore throat, jaw pain, a stiff neck, and pleuritic pain. He did have rheumatic fever at age 17. At hospital admission he also had a non-migratory, persistent joint pain and swelling and intermittent nonpruritic macular rash. Imaging showed pleural effusion, pericarditis, and abdominal lymphadenopathy. Labs revealed leukocytosis, thrombocytosis, anemia, extreme elevated CRP, elevated anti-streptolysin o, and anti-Dnase B titers. Skin biopsy suggested erythema marginatum—seen in rheumatic fever and Still's disease. He was treated with indomethacin. He presented 3 weeks later with persistent thrombocytosis and joint flare. A tapering dose of prednisone was added to the indomethacin. Two months later, he improved significantly—showing weight gain, improved energy levels, no rash for several months, no joint flare, and a normal complete blood cell count, C-reactive protein, kidney and liver function on lab results. A year later the indomethacin was stopped and he did not have another flare.

Discussion: Features of this case that suggest a diagnosis of adult-onset JRA are the extreme leukocytosis and thrombocytosis, quotidian fevers, persistent joint pain lasting for more than a month, and the Still's rash. Additionally, the onset of the arthritis coexistent with a sore throat, pleural pericarditis, and extreme weight loss are also typical of this disease. This case illustrates the importance of considering JRA in the differential diagnosis of a multisystemic rheumatologic disorder in adults.

ORAL VIGNETTES

Hemolytic-Uremic Syndrome Complicated by Pulmonary Emboli and Heparin-Induced Thrombocytopenia

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Case: A 58-year-old man with no significant past medical history presented with acute kidney injury. Over the last 2 weeks, he had experienced symptoms consistent with a viral upper respiratory infection. Several days prior to admission, he developed low-grade fevers, malaise, nausea, watery stools, and oliguria. On admission, laboratory analysis revealed acute kidney injury with a blood urea nitrogen (BUN) of 127 and creatinine of 8.2, microangiopathic hemolytic anemia with a negative direct Coombs, thrombocytopenia with a platelet count of 98,000, and fibrinolysis. These findings, in combination with mild mental status changes, supported the diagnosis of hemolytic-uremic syndrome. Thrombotic thrombocytopenia purpura and antiphospholipid antibody syndrome were deemed less likely when his a disintegrin-like and metalloproteinase with thrombospondin (ADAMTS) 13 activity, lupus anticoagulant, and anticardiolipin antibodies returned within normal limits. The patient improved on hemodialysis and daily plasmapheresis. On day 6 of hospitalization, the patient became acutely hypoxemic and had a syncopal episode. Ventilation/Perfusion scan showed bilateral pulmonary emboli. Given his hemodynamic stability and normal echocardiogram, the patient was treated conservatively with heparin. He had not received prophylactic anticoagulation out of concern that it could exacerbate his preexisting thrombocytopenia.

Four days later, the patient's platelets dropped from 230,000 to 146,000. This was highly suspicious for heparin-induced thrombocytopenia (HIT), given his improving creatinine, decreasing lactic acid dehydrogenase and lack of schistocytes on peripheral blood smear. Heparin was discontinued and lepirudin started, as HIT can be a prothrombotic state. An enzyme-linked immunosorbent assay for HIT antibodies was positive, and a 14C-serotonin release assay subsequently confirmed HIT. The patient was bridged to warfarin when his platelets recovered.

Discussion: This case illustrates the relatively unusual occurrence of hemolytic-uremic syndrome in an adult patient, as well as the complexities of managing anticoagulation in such patients.

It's Not Just a Sore Throat—Lemierre's Syndrome

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Case: A 22-year-old previously healthy microbiology student currently doing research on *Haemophilus aphrophilus, Actinobacillus actinomycetemcomitans, Cardiobacterium hominis, Eikenella corrodens, and Kingella kingae (HACEK)* organisms came to the ED with complaints of a sore throat for 2 weeks. He was seen 1 week earlier at which time a rapid strep test and Monospot test were negative. He was given oral Ampicillin and sent home. He developed a rash resulting in a switch to Azithromycin the next day. He subsequently developed fevers, chills, increasing cough with pleuritic chest pain, worsening sore throat, dizziness, fatigue, and generalized malaise. A complete evaluation revealed tachycardia, fever, an erythematous oropharynx, and anterior cervical chain lymphadenopathy. Laboratory tests revealed an elevated white count, a left lower lobe infiltrate and positive Monospot test. He was admitted with possible community acquired Methicillin-Resistant *Staphylococcus aureus* and he was placed on Linezolid and levofloxacin intravenously. However, he rapidly deteriorated with acute respiratory failure requiring intubation. Chest CT showed worsening infiltrates bilaterally with cavitations and bilateral pleural effusions. Blood cultures grew anaerobic gram-negative rods and metronidazole was added to his antibiotic regimen. A CT of his neck and chest was then obtained to look for mediastinitis or neck abscess. It showed a questionable abscess on right side of neck. This was thought to be an inflammatory mass as interventional radiology was not able to drain any fluid. Due to bradycardia and pauses, a transthoracic echocardiogram was performed that was normal. His blood cultures were positive for *Fusobacterium necrophorum* and his antibiotics were changed to ertapenem. From this point he rapidly recovered with extubation a few days later and eventually was discharged home.

Discussion: Lemierre described Lemierre's syndrome as post anginal septicemia. It is an extremely rare, life-threatening septic thrombophlebitis of the jugular vein associated with anaerobic sepsis. It may arise from inflammatory lesions of oropharynx, otitis media, mastoiditis, genitourinary tract, or after gastrointestinal surgery. Immunocompromised patients are especially susceptible, but it is usually seen in young previously healthy patients with recent pharyngitis. Usual complications are posterior pharyngeal space infections with abscess formation, jugular vein phlebothrombosis, septic pulmonary emboli, and respiratory failure. Throat swabs, blood cultures, and imaging modalities like ultrasound and CT should guide in diagnosis. Early empiric treatment with good anaerobic coverage helps prevent fatal complications. Usually, 2 weeks to 3 months of antibiotic treatment is recommended. Surgical treatment is indicated in certain cases.

These Old Bones in My Young Body!

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Case: A 35-year-old Hispanic woman presented to the ED after waking with a swollen, erythematous right lower extremity associated with non-radiating pain and inability to bear weight. She was evaluated 1 week prior at another hospital for non-traumatic right lower fibular and tibial fracture for which the patient was treated with foot immobilization and referral to orthopedic surgery. Her past medical history was significant for diabetes mellitus type II, non-traumatic fractured femur and osteoporosis. On physical examination, vital signs were stable. The right lower extremity was swollen, tender to touch with erythema around the ankle up to the mid shaft region, decreased range of motion, inability to evert or invert the right foot secondary to pain. There was evidence of dentingenous imperfecta and onycholysis with a missing fingernail on the third left finger. Complete blood cell count and cytidine monophosphate were normal except for elevated alkaline phosphatase. X-ray showed evidence of fractured mid-shaft fibula and tibia and fracture of the third, fourth and fifth toes on the left foot. Osteoporosis of the hips bilaterally was seen. The limb was stabilized and patient was taken to surgery.

Discussion: Osteogenesis Imperfecta (OI) is an autosomal dominant mutation (sometime sporadic) that causes brittle bones, which are susceptible to fractures. The clinical range extends from mild symptoms to perinatal lethal disease (Type I-Type IV). The mutation occurs in the Type I collagen protein fibers, which are found in tendons, sclera, bones, organ capsules, fascia, dermis, and meninges. Two pro-alpha 1, and one pro-alpha 2 chains form this left-handed helical structure. Both chains have a primary structure of uninterrupted repeats of the tripeptide composed of glycine (Gly), proline (Pro), and hydroxyproline (Hyp), in that order. When Gly is substituted by another protein, OI occurs. Heterozygous mutations affecting the primary collagen structure cause moderate and severe osteogenesis imperfecta, whereas decreased collagen production causes mild osteogenesis imperfecta. The prevalence of OI is estimated to be 1 per 20,000 live births, the mild form is underdiagnosed, and the actual

prevalence may be higher. The diagnosis is mainly clinical, with a history of repeat non-traumatic fractures, osteoporosis in teens or early twenties usually exacerbated by childbirth in women, and a history of easy bruising during childhood. Confirmation is with elevated alkaline phosphatase, imaging, and genetic testing. This is a genetic disorder and there is no cure. The mainstay treatment is surgery with lifestyle modifications. Fractures heal normally in approximately 85% of patients.

Prevention is Always Better Than Cure!

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Case: A 78-year-old woman presented with a 48-hour history of progressive dysphagia, neck pain, and difficulty opening her mouth. Five days before presentation she accidentally stepped on a rusty nail and sustained a puncture wound extending to 1.5 cm into the plantar aspect of her right foot. She didn't seek medical attention until the day of admission. Past medical history is significant only for remote history of gout. She has not received primary medical care or tetanus immunization for many years.

Physical exam revealed an obese female in moderate distress with blood pressure=218/96, temperature=98.9ºF, heart rate=87, and Oxygen saturations of 95% on room air. She had inability to open her mouth beyond 3 cm, trismus, and risus sardonicus were noted. Neck exam revealed markedly restricted range of motion, tenderness to palpation, and trapezius spasm. Cardiopulmonary examination was significant only for II/VI aortic systolic murmur. Remaining neurological exam was normal. Initial labs showed normal cell counts with elevated creatine kinase and minimally elevated Troponin. Chest X-ray showed mild cardiomegaly and electrocardiogram with left ventricular hypertrophy. CT scan of soft tissues of neck showed no evidence of retropharyngeal abscess or cervical inflammatory process. Fiberoptic laryngoscopy showed no obvious airway compromise or edema. After initial clinical diagnosis of tetanus, patient was given 2000 IU of human tetanus immunoglobulin, started on metronidazole, diazepam, tetanus toxoid series, and admitted to ICU for airway monitoring. She developed hypoxemic respiratory failure 10 hours later and underwent emergent tracheostomy. After prolonged intensive care unit and hospital stay of 46 days, patient improved gradually and was discharged home after rehabilitation.

Discussion: Tetanus continues to be a major medical problem in the developing countries. Tetanus immunization dramatically reduced annual incidence in the United States. However, unvaccinated or inadequately vaccinated adults and intravenous drug abuser continue to be at risk.

Primary Hepatic Lymphoma With Hepatitis C

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Case: A 41-year-old woman with a history of chronic hepatitis C without cirrhosis presented with a 2-week history of right-sided abdominal pain, nausea, vomiting, and 25-pound weight loss. Abdominal CT revealed multiple hypo-attenuating lesions in the liver suggestive of malignancy. Alpha-fetoprotein level was 8.7. No other site of a primary tumor could be found. A diagnosis of multifocal hepatocellular carcinoma was entertained; however an ultrasound-guided biopsy of the liver was consistent with diffuse large B-cell lymphoma. Serologic studies for human immunodeficiency virus were negative. Staging PET scan did not show any evidence of extra-hepatic disease confirming a diagnosis of primary hepatic lymphoma (PHL). Chemotherapy with rituximab, cyclophosphamide, adriamycin, vincristine, and prednisone (R-CHOP) was started, and she has since completed 3 cycles with a good clinical response.

Discussion: Non Hodgkin's lymphoma (NHL) is the fifth most common cancer in the United States. Primary hepatic lymphoma, however, is extremely rare, accounting for <0.05% of all NHL. This has been described more frequently in the setting of hepatitis C virus, HIV/acquired immune deficiency syndrome (AIDS) and other immunocompromised states. Available literature on the incidence of PHL is scant; some authorities believe that the incidence may be on the rise with the rising epidemic of HIV and hepatitis C. Diffuse large B-cell lymphoma is the most common subtype. Lymphomagenesis related to chronic hepatitis C continues to be an area of intense study. Treatment options include surgery, chemotherapy, and radiation, depending on the extent of the disease and histological subtype. Chronic hepatitis C infection confers a 20-fold increased risk of HCC; the risk of NHL is increased more modestly (20%-30%). The clinical and radiological features of PHL may overlap with those of HCC with biopsy being the only distinguishing test. While the prognosis of HCC is extremely poor with expected 6 month survival of <10%, the prognosis of PHL is quite favorable (70% 5 year survival with treatment). Therefore, pathologic confirmation of the diagnosis is crucial since treatment and prognosis differs considerably.

Migratory Poyarthralgia—Is it a Harbinger of Serious Systemic Illness?

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Case: A 37-year-old previously healthy woman was admitted from the rheumatology clinic with a 10-week history of migratory polyarthralgia, hemoptysis, dyspnea, morning stiffness, and bilateral earache and discharge. There was no associated erythema or joint swelling. She had minimal constitutional symptoms of low grade fever and fatigue. She had a strong family history of rheumatoid arthritis and Hashimoto's thyroiditis. She was evaluated 3 times in the ED

prior to this when migratory arthralgia was her only symptom with minor earache in the latter weeks. Her joint imaging was normal. Initial labwork in the ED showed mild anemia and microscopic hematuria with mildly elevated inflammatory markers. She received symptomatic treatment with opioid pain medications, empiric doxycycline therapy for possible Lyme's disease, and ciprofloxacin eardrops for earache. Examination on admission revealed crackles in her right lung base and dried secretions in both ear canals. Significant lab results on admission included hemoglobin 10.3 g/dl, sedimentation rate 120, C-reactive protein 22.9, and microscopic hematuria with red cell casts and proteinuria. Chest CT showed multiple cavitating lesions. Urgent renal biopsy showed necrotizing crescentic glomerulonephritis. She was diagnosed with Wegener's granulomatosis (WG) and was started immediately on cyclophosphamide and methylprednisone to which she responded very well. Antineutrophil cytoplasmic antibody (cANCA), especially the anti-proteinase 3 antibody, was elevated.

Discussion: WG is a necrotizing granulomatous vasculitis affecting small blood vessels with classic predilection for otolaryngeal and renopulmonary systems with a severe complication profile and high mortality. The American College of Rheumatology criteria for diagnosis include oral or nasal inflammation, abnormal chest radiograph (nodules, cavitary lesions, infiltrates), abnormal urinary sediment, and granulomatous inflammation on biopsy of an artery. Interestingly, musculoskeletal signs and symptoms are not part of the diagnostic criteria and yet joint symptoms are reported in more than 65% of the cases and can be inaugural in more than 20% of cases. Arthralgia is the most common symptom. Frank poly- and oligoarthritis, mostly nondeforming and nonerosive, have been reported in 25%-30% of cases. Effusion is rare and usually shows nonspecific inflammatory fluid. Migratory pattern is noted in about 5% of cases. Rheumatoid factor and antinuclear antibody can frequently be positive. Recognizing the atypical presentations of multisystem disorders is important, as these can often be a harbinger of potentially serious systemic illness. Looking diligently for further clues earlier in the illness can prevent potentially life-threatening complications.

Out of Africa

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Case: A 62-year-old man presented with a 4-day history of pain, swelling, and erythema of the left leg, which was accompanied by fever, chills, malaise, diffuse myalgia, and headache. This illness began just prior to returning to the United States after hunting deer and impala in rural South Africa. He had no specific vaccinations or prophylaxis prior to travel. Physical exam was significant for temperature of 101.8°F, bilateral inguinal lymphadenopathy, and a black eschar at the base of the left first metatarsophalangeal joint with associated streaking erythema, warmth, and tenderness extending proximally to the knee. Initial laboratory testing revealed a normal complete blood cell count, but his C-reactive protien was elevated to 5.9 mg/dL and erythrocyte sedimentation rate to 47. Routine blood cultures and malaria smears were negative. Given the characteristic appearance of the eschar, or tache noir, a diagnosis of African tick bite fever was made. After treatment with doxycycline, he rapidly improved. Serum IgG for *Rickettsia africae* was negative likely due to the acuity of the infection.

Discussion: With increasing international travel, it becomes important to recognize diseases endemic to various regions. The incidence of African tick bite fever (ABTF) is estimated in up to 5.3% of travelers from sub-Saharan Africa and the eastern Caribbean. Infection with *R. africae* occurs via cattle ticks of the Amblyomma genus. ABTF commonly presents as an influenza-like illness accompanied by fever, regional lymphadenopathy, and an inoculation eschar, or tache noir. Risk factors include travel in endemic areas during the rainy season and contact with wild animals. Laboratory diagnosis consists of serologic tests, which become positive approximately 3 weeks after the onset of symptoms, as well as serum or tissue polymerase chain reaction and immunohistochemistry. The treatment of choice is doxycycline. Travelers should be counseled on prevention via application of DEET-containing products.

Drug Induced Sweet's Syndrome in Assosciation with Polychondritis and Multiple Autoantibodies

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Case: We present a case of drug-induced Sweet's Syndrome (SS) with polychondritis and multiple autoantibodies in an 86-year-old woman. She presented with a 5-day history of fever; conjunctivitis; painful tense vesicles on her ears, nose, and back; and painful digital lesions. She has a history of bipolar disorder and hypertension and was on multiple medications. Patient was on lithium for years, which was stopped 10 days prior to admission because of lack of efficacy and carbamazepine was started. She had been on hydralazine just over 1 year; dose was increased 8 months prior. Physical exam revealed fever, blepharoconjunctivitis, tense vesicles and bullae on the nose, superior aspect of the ears, and upper back, erosions on the post auricular crease, chondritis involving ear and nose, and oral ulcers. Hemorrhagic papulovesicles and bullae were noted on the distal finger pads and lateral fingers. Laboratory studies revealed elevated erythrocyte sedimentation rate and C-reactive protein, azotemia, microscopic hematuria, and proteinuria, antinuclear antibody positive (1:640), p-ANCA positive, MpoAb 200, Pr3 48.5, antihistone antibody positive, type II collagen antibody positive (47.6), lupus anticoagulant positive. Skin biopsies from the back and finger were consistent with neu-

trophilic dermatosis. A chest X-ray showed no infiltrates. Hydralazine and carbamazepine were discontinued. Cutaneous lesions and renal insufficiency rapidly improved on systemic corticosteroids.

Discussion: Sweet's syndrome (SS) is an acute febrile neutrophilic dermatosis. Association with infections, autoimmune diseases, inflammatory bowel disease, malignancy, and drugs are reported. There are few case reports of association between SS and relapsing polychondritis. Our patient had biopsy proven SS, clinical diagnosis of polychondritis supported by positive anti collagen antibodies, and multiple autoantibodies. She was on 3 drugs that have been associated with drug-induced SS, drug-induced lupus, and p-ANCA positive vasculitis.

The Lethal Giant

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Case: A 34-year-old man presented with an acute onset of dyspnea, orthopnea and paroxysmal nocturnal dyspnea. He had no history of diabetes, hyperlipidemia, or coronary artery disease. Past medical history was notable for ulcerative colitis, primary sclerosing cholangitis, alcohol abuse, and smoking. Medications included flagyl, ibuprofen, and bupropion. A complete physical exam revealed hypotension, tachycardia, tachypnea, S3 gallop, bibasilar lung crackles, and mild hepatomegaly. A chest X-ray revealed marked pulmonary congestion, and ECG revealed low voltage QRS with a left posterior hemiblock, ST elevation from V1 to V6. Troponin-I was elevated to 2.09. With the initial impression of cardiogenic shock from acute myocardial infarction he was started on oxygen, heparin, pressors, and diuretics. However, a coronary angiogram was essentially normal. ECHO revealed EF of 31% with global left ventricle dysfunction, which later dropped to 15% on day 5 despite intra-aortic balloon pump. The transplant team was then contacted. However the patient went into ventricular tachycardia, and despite all efforts of resuscitation he remained unresponsive and died shortly thereafter. The autopsy revealed diffuse myocardial necrosis, inflammatory infiltrate with multinucleated giant cells consistent with the diagnosis of giant cell myocarditis.

Discussion: Giant cell myocarditis was first described in 1905. It is a rare but fatal myocarditis occurring in relatively young healthy adults. Etiology is unknown, but 20% of cases had autoimmune diseases. Evidence suggests potential autoimmune pathogenesis involving CD4 T lymphocytes. This disease frequently presents as acute heart failure. Endomyocardial biopsy is the standard diagnostic test with an 85% sensitivity. There is an average survival of 5.5 months, which is prolonged from 12.3 months with combined immunosuppressive treatment. Definitive treatment is heart transplant. Post transplant survival was noted to be roughly similar to patients receiving transplant for other etiologies. However, post transplant care involves close follow up as recurrence has been noted.

Conclusion: It is important to consider the possibility of giant cell myocarditis in otherwise healthy individuals who present with new onset, rapidly progressing left ventricular failure, especially in those with associated refractory ventricular tachycardias. Prompt pathological diagnosis will allow early pursuit of transplant and combined immunosuppressive therapy that can potentially improve survival.

Rapid Renal Ruins

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Case: A 57-year-old woman with a history of degloving extremity injuries and multiple reconstructive surgeries 6 years ago presented with bilateral leg swelling. The previous year she had several admissions for lower extremity cellulitis and was treated with ertapenem, amoxicillin-clavulanate, and cefazolin. Now her leg was mildly erythematous and edematous, and intravenous naficillin was started for presumptive cellulitis and furosemide for diuresis. Within 17 hours of the first doses, her creatinine more than doubled. She remained afebrile and hemodynamically stable. She had no prior renal problems and did not develop rashes, arthralgias, flank pain, malaise, anorexia, oliguria, or nausea. The medications were held, and a prerenal etiology was considered because her fractional excretion of urea was <35%. Despite intravenous fluids, her blood urea nitrogen (BUN) and creatinine increased every hour. No plasma or urine eosinophilia was found, and renal ultrasound was normal. Urinalysis showed no white blood cells/casts but had protein and blood. Proteinuria and hematuria in the setting of acute renal failure suggested glomerular disease; however, complement and antinuclear antibody levels were normal. Renal biopsy was performed to further elucidate the cause and revealed acute tubulointerstitial nephritis (AIN). Intravenous methylprednisolone was administered for 2 days and then switched to oral prednisone. Her creatinine and BUN returned to normal 2 weeks later.

Discussion: This case illustrates an atypical presentation of AIN. Although AIN usually manifests over several days, with average time to clinical development being 2 weeks after drug administration, her creatinine doubled within a few hours of receiving naficillin. Also, classic symptoms and signs such as rash, fever, arthralgias, urine and serum eosinophilia, and urine white blood cell casts were absent. No evidence has clearly demonstrated that corticosteroids are effective in AIN. Most patients respond to discontinuation of offending medications, but cyclophosphamide has been considered in refractory cases and dialysis for severe electrolyte and metabolic disturbances. Our case demonstrates the need to maintain a high suspicion for AIN even when the clinical picture is not entirely consistent with this common cause of renal failure.

Casting With Plastic!

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Case: A 60-year-old woman was evaluated for shortness of breath following repeat coronary artery bypass graft (CABG) surgery. She had CABG in 2002 with repeat in 2005 utilizing the right internal mammary artery. The dyspnea began shortly after hospital discharge in 2005. She also described a cough productive of firm, rubbery debris in the shape of some portion of the bronchial tree. She has a remote history of smoking and no prior lung disease. CT of the chest was normal. Pulmonary function testing demonstrated mild obstruction with good bronchodilator response. She underwent bronchoscopy with multiple bronchial casts extracted. Pathology demonstrated composition of blood, fibrin and mucin. Cytology and cultures were negative for infection and malignancy. Further work up was negative for cytomegalovirus, Aspergillosis, IgE, ANCA and angiotensin-converting enzyme. Hypersensitivity panels were negative as well. Initial treatment included inhaled corticosteroids and bronchodilators. As the disease progressed, she required courses of oral prednisone and was started on home oxygen and flutter valve therapy.

Over the course of 3 years, she has had multiple bronchoscopies for airway clearance. Her cast production has been increasing in frequency and severity, which has significantly affected her lifestyle. She produces 10-15 samples per day. The largest cast was 9.0 x 6.5 x 1.2 cm, taking her 13 hours to cough out. Treatment to date has been guided by case reports as the disease is rare and seen almost entirely in children. Azithromycin, as an anti-inflammatory agent, and dornase alfa, a rhDNase to reduce mucous viscosity, have given the most benefit to date.

Discussion: Plastic bronchitis is a rare disease most commonly seen in the pediatric population following the Fontan procedure as part of congenital heart defect repair. It is characterized by the formation of endobronchial casts thought to be formed secondary to damage of the lymphatic system. It has been reported to occur also in patients with allergic and inflammatory diseases including asthma, cystic fibrosis, pulmonary infections, and acute chest syndrome in sickle cell disease. These casts can cause pulmonary symptoms of wheezing, coughing, or respiratory distress. Diagnosis is made after expectoration of bronchial casts or by bronchoscopy. Multiple therapies have been trialed such as steroids, antibiotics, pulmonary vasodilators, and thoracic duct ligation with varied results.

Steroids for Parasites? A Case of Undifferentiated Hypereosinophilia

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Case: A 29-year-old woman with a history of asthma presented to the hospital with progressive shortness of breath over several days. In addition to her respiratory symptoms, she described fatigue associated with a 20-pound unintentional weight loss. She also complained of pain and weakness in her right leg and left arm. She had a history of childhood asthma that recurred around the age of 21. Her symptoms were well controlled with a fluticasone/salmeterol inhaler, her only medication. She emigrated from India approximately 4 years prior and had returned there within the past year. She followed a healthy lifestyle and her family history was significant for coronary artery disease and diabetes. Pertinent exam findings included hypotension (92/67 mm Hg), tachycardia (100-120 bpm), right eyelid droop, bibasilar pulmonary crackles, normal S1 and S2 cardiac sounds with presence of S3, decreased muscle strength in her left arm and right leg, normal reflexes, and a macular rash on her lower extremities. Initial lab work was remarkable for a leukocytosis to 14,200 with an eosinophilic predominance of 5970. Her hemoglobin was normal at 11.7 g/dL and platelets normal at 322,000. Her chemistries were within normal limits including a blood urea nitrogen (BUN) of 13 and creatinine of 0.7. Her erythrocyte sedimentation rate was elevated at 50. A comprehensive work up for infectious diseases including parasites was negative. A chest CT revealed bilateral pleural effusions and patchy pulmonary infiltrates. An ECG was obtained which showed Q waves in leads V1-V4. Cardiac catheterization was without coronary artery disease. A transthoracic ECHO demonstrated severely reduced left ventricular function with an estimated ejection fraction of 20%. A myocardial biopsy similarly showed eosinophilic infiltrate.

Discussion: The constellation of her symptoms including asthma prodrome, eosinophilia, elevated inflammatory markers, cardiac infiltrate resulting in heart failure and mononeuritis multiplex suggested the diagnosis of Churg-Strauss Syndrome (CSS).