An Unexpected Cause of Upper Airway Obstruction After Extubation: An Illustrative Case of the Potential Value of Health Information Exchange

Anil Makam, Won Lee
Southwestern Medical Center, Dallas, Texas

LEARNING OBJECTIVES:
1. Recognize that myasthenia gravis can cause upper airway obstruction resulting in respiratory impairment and extubation failure. 2. Recognize the potential role of health information exchange in expediting diagnostic workups and reducing duplication of efforts.

CASE INFORMATION: A code blue was called for an 18-year-old African-American female with sickle cell anemia following a CT angiogram of the chest. She was emergently intubated for hypercapneic, hypoxic respiratory failure. She had been admitted 12 hours prior for sickle cell crisis with shortness of breath. The CT chest was remarkable for new consolidations in the right middle and left lower lobes, consistent with acute chest syndrome. After a single red blood cell exchange transfusion, she had minimal oxygen requirements and met adequate ventilator weaning parameters, such as rapid shallow breathing index and minute ventilation. Immediately following extubation she became hypoxic and tachypneic, with no stridor or audible airflow upon chest auscultation. She was re-intubated without difficulty. Although her neurological exam was limited by sedation, she had mild symmetric proximal muscle weakness with 4/5 strength, ptosis with sustained upward gaze, a negative inspiratory force of -22 cm H2O, and a forced vital capacity of 460 ml. A CT of her neck was unremarkable for extrinsic airway compression. A second attempt at extubation was performed with bronchoscopic assistance to evaluate for upper airway patency. As the tube was slowly retracted, the upper airway was observed to completely collapse at the level of the vocal cords. A diagnostic workup to evaluate for neuromuscular etiologies of upper airway obstruction revealed elevated anti-acetylcholine receptor antibodies and a significant decrement on a repetitive nerve stimulation study, confirming a diagnosis of myasthenia gravis. After instituting plasmapheresis, corticosteroids, and pyridostigmine, the patient made an uneventful recovery. Once extubated, she reported that she had already been diagnosed with myasthenia gravis, just 3 months prior at one of our neighboring teaching hospitals.

IMPLICATIONS/DISCUSSION: Upper airway obstruction due to bulbar predominant myasthenia gravis causing respiratory failure and asphyxia is infrequently reported in medical literature and to the best of our knowledge, this is the first documented case resulting in extubation failure. Although approximately 30% of patients with myasthenia develop some degree of respiratory impairment and 44% of intubated patients with myasthenic crisis in one case series experienced extubation failure, pulmonary complications typically are due to respiratory muscle weakness, inability to manage secretions, atelectasis, or infectious pneumonia. Despite favorable ventilator weaning parameters, myasthenic patients with significant bulbar muscle weakness are at risk for airway compromise when the stenting effect of an endotracheal tube is removed. When upper airway obstruction is suspected, direct visualization of the airway is essential to exclude more common etiologies, such as tracheal stenosis and laryngeal edema, and can confirm loss of airway patency after extubation. In the absence of structural abnormalities an evaluation for neuromuscular etiologies, including myasthenia gravis, should be pursued.

This case also highlights the potential value of health information exchange. The patient's perplexing and complicated clinical course, with repeated intubations and a prolonged intensive care stay, could have been preempted by access to her prior health records. However, the limited history and a family who was unaware of her recent diagnosis gave no indication that further relevant information was available. Knowledge of prior history, medications, and care plan at the point-of-care in real-time can circumvent diagnostic challenges, delays in initiating therapy, and duplication of health services—thus, improving outcomes and reducing costs.
An uncommon cause of sudden death and acute coronary syndrome (ACS) in the post partum period
Waridibo E Allison 1; Sudip Ghimere 1; Rupak Thapa 1; Eugenio Guzman 1. 1North Central Bronx Hospital, New York, New York. (Proposal ID # 11491)

LEARNING OBJECTIVES: 1. Recognize spontaneous coronary artery dissection (SCAD) as an infrequent and unusual cause of ACS in young patients with no risk factors. 2. Recognize SCAD as a diagnosis that must be considered particularly in young women with chest pain during pregnancy and the post partum period.

CASE INFORMATION:
A 33 year old woman of African origin, gravida 1 para 1, had sudden onset of severe central chest pain in OB/GYN clinic. She had undergone an uncomplicated cesarean section two weeks prior following a normal antenatal course. She had no known risk factors for coronary disease and did not smoke, drink alcohol or use recreational drugs.

She was immediately taken to the ER in a wheelchair and on arrival at triage collapsed and became unresponsive. The patient was found to be in ventricular fibrillation cardiac arrest and ACLS protocol was initiated. She received atropine, epinephrine, magnesium and amiodarone and was defibrillated with 120/200/200 joules. There was spontaneous recovery of circulation and the patient was intubated and ventilated. A transthoracic echocardiogram (TTE) post arrest showed normal left ventricle (LV) function, no wall motion abnormalities and no pericardial effusion.

In ICU the patient was titrated off Dobutamine, amiodarone was continued and she was put on a mild hypothermic protocol to minimize anoxic brain injury. The patient was additionally commenced on aspirin, clopidogrel, lisinopril and a heparin infusion. Serial troponin levels peaked at 12.73 on Day 2 after the cardiac arrest and repeat TTE on the same day showed markedly reduced LV systolic function with an ejection fraction of 30% and severe hypokinesis of the apical wall. Right ventricle function was also reduced. Electrocardiograms initially showed inferior-lateral ST depression and later anterior-lateral T wave inversion. Extubation occurred successfully on Day 3 after the cardiac arrest and the patient underwent coronary angiography on Day 6. This revealed spontaneous coronary artery dissection of the mid LAD artery causing 95% stenosis and apical akinesis. Two drug eluting stents were placed at the dissection site.

The patient made a good recovery and was discharged 9 days after the cardiac arrest.

IMPLICATIONS/DISCUSSION:
Prevalence of SCAD was recently estimated at 0.7% in an angiographic study. A 2010 review of the literature found 440 reported cases of which 70% were women. Pregnancy was associated with 26% of cases and of these 84% of cases occurred in the post partum period with 70 events occurring within 2 weeks of delivery. Approximately a fifth of all cases were diagnosed by post mortem examination and the majority of the remainder by coronary angiography. The patient in this case was fortunate to have collapsed in a hospital setting close to an ER. Mortality was even higher with pregnancy associated SCAD with sudden death or death within a few hours of symptom onset occurring in 40%.

Presentation is usually with typical symptoms of ACS including chest pain. Electrocardiogram may be normal initially. The exact etiology of SCAD is unclear but it may be influenced by eosinophilic infiltration and cystic medial necrosis, hormonal changes weakening the arterial wall and hemodynamic stress that occurs during the final trimester of pregnancy. Further research is needed to elucidate the processes involved in development of SCAD as this knowledge will assist in improving risk stratification.

There are currently no evidence based guidelines for medical management of this condition but pharmacological agents used include aspirin, clopidogrel, heparin, beta-blockers, and angiotensin-converting enzyme inhibitors. Thrombolytics may cause extension of the dissection and further narrowing of the true lumen and are not recommended. It has been shown that coronary intervention with stenting has superior
outcomes compared to conservative medical management of SCAD for both right and left coronary artery lesions.

Rapid diagnosis and subsequent appropriate treatment is essential to avert disastrous clinical sequelae and to achieve an excellent outcome in this condition that disproportionally affects young women.
Factor Five Leiden presenting with Small Vessel Ischemic Colitis George Lominadze¹; Kristina Chae¹; Darlene LeFrancois². ¹Montefiore Medical Center, Bronx, New York; ²Montefiore Medical Center, New York, New York. (Proposal ID # 10561)

LEARNING OBJECTIVES: 1. Recognize clinical manifestations of small vessel ischemic colitis. 2. Diagnose and manage ischemic colitis due to a hypercoagulable state.

CASE INFORMATION: 42 year old man presented with intermittent crampy left lower quadrant (LLQ) abdominal pain, tenesmus, pencil-thin stools alternating with bloody diarrhea, and fevers for 6 months. He had lost 56 lbs and felt fatigued. A colonoscopy 3 months earlier showed colitis of descending and sigmoid colon without pseudomembranes, and negative biopsy for granulomas or vasculitis. Empiric treatment with 5-ASA and metronidazole failed. Aside from thyroid hormone replacement, he took no medications. He had no toxic habits and his family history was unrevealing. Examination vitals were BP 134/97, HR 65, Tmax of 100.3, BMI XX. He preferred to lay still, with exquisite tenderness at the LLQ accompanied by localized guarding. Stool guaiac was positive and there was mild dependent edema. Laboratory testing was significant for CRP 24 and ESR 60. Celiac panel testing and ANCA studies were negative. Stool cultures and ova and parasite examinations were negative as was clostridium difficile testing. CT angiography showed mural thickening of the descending and rectosigmoid colon with infiltration of the mesenteric fat but no vessel thrombosis, although inferior mesenteric vein (IMV) could not be visualized. Colonoscopy revealed multiple descending and sigmoid colonic strictures and ulcers that microscopically displayed focal hyperplasia of the lamina propria and inflamed granulation tissue. A complete workup for hypercoagulable states was done, and genetic testing revealed a Factor V Leiden homozygous mutation. Antibiotics resulted in only transient improvement in symptoms and patient underwent a left-sided hemicolectomy. Pathologic examination of the specimen revealed colonic transmural necrosis with perforation, and fat necrosis of pericolic adipose tissue. Marked inflammation was associated with hemorrhagic changes thrombosis of submucosal and subserosal vessels. Patient is currently doing well on anticoagulation with warfarin.

IMPLICATIONS/DISCUSSION: Factor V Leiden (FVL) is an autosomal dominant mutation that can lead to a hypercoagulable state through activated protein C resistance. 3-7% of the general population is heterozygous, while 0.06 to 0.25% is homozygous. Heterozygocity increases the risk of venous thrombosis around 5 fold, while homozygocity can increase the risk 20-30 fold. Most cases of thrombosis occur in patients in their twenties. To our knowledge this is the first report of ischemic colitis presenting as the initial thrombotic event that led to diagnosis of homozygous FVL mutation. There are two case reports of heterozygous FVL mutation presenting with ischemic colitis, one in a 24 year old woman and another in a 73 year old man. Two small studies assessed the prevalence of hereditary thrombotic risk factors in patients with ischemic colitis, with one study finding that 22% of 36 patients with colon ischemia had FVL heterozygous mutation, while in another study one out of 18 patients did. Our patient had small mesenteric vessel thrombosis (MVT), similar to the 73 year old patient with FVL heterozygous mutation, suggesting that MVT associated with FVL may not be readily detectable by CT angiography. In addition, our patient’s thrombosis was in the distribution of the IMV, with superior mesenteric vein (SMV) patent. According to literature, mesenteric venous thrombosis accounts for 5 to 15 percent of all mesenteric ischemic events and usually involves the superior mesenteric vein. The involvement of IMV rather than SMV likely made the prompt diagnosis difficult, as IMV drains a much smaller section of the GI tract than SMV, leading to less severe symptoms. Nonspecific findings on colonoscopy and the lack of identifiable risk factors for ischemic colitis in this patient undoubtedly contributed to the 6 month delay in his diagnosis. Clinicians should consider studies for hypercoagulable states in patients with ischemic colitis so as not to miss this important and treatable condition.
Recurrent common iliac venous thrombosis Julie Kim 1; Abigail Deyo 2; Kurt Pfeifer3. 1Medical College of Wisconsin Affiliated Hospitals, New Berlin, Wisconsin; 2Medical College of Wisconsin, Milwaukee, Wisconsin; 3Medical College of Wisconsin Affiliated Hospitals, Milwaukee, Wisconsin. (Proposal ID # 11908)

LEARNING OBJECTIVES:
1. Identify patients at high risk for antiphospholipid antibody syndrome (APS) and describe appropriate treatment for acute venous thromboembolism in patient with APS
2. Recognize that lupus coagulant antibodies can falsely prolong INR levels

CASE INFORMATION: A 28-year-old woman with Hashimoto's thyroiditis presented with a one-day history of acute onset of left thigh pain, chest discomfort, left lower rib pain with inspiration and mild dyspnea. Physical exam revealed tachypnea and an erythematous, warm, and swollen left thigh. Her medications were oral contraceptives, but she denied smoking or a family history of clotting disorders. Left lower extremity ultrasound confirmed thrombosis of the left common iliac vein, and ventilation-perfusion scan showed high probability for bilateral multi-subsegmental pulmonary emboli. She was started on warfarin and bridged with dalteparin until her INR was therapeutic. She was discharged home but returned with progressive thigh pain. Left lower extremity ultrasound revealed worsening thrombosis with clot now extending from the left common iliac vein to popliteal vein. She underwent catheter-directed thrombolysis and was restarted on intravenous unfractionated heparin. Venogram revealed no structural cause, such as May-Thurner syndrome, for her recurrent thrombosis. Thrombophilia work-up revealed minimally elevated lupus anticoagulant, partial thromboplastin time and dilute Russell's viper venom time. She was discharged home on therapeutic-dose dalteparin. Four days later, she returned with onset of new left calf pain. Ultrasound showed a patchy distribution of thrombosis from her femoral vein to her popliteal vein. Repeat catheter-directed thrombolysis and continuous heparin and tissue plaminogen activator were administered, and ultrasound evaluation afterwards showed near resolution of her clot. She was again discharged on dalteparin with hematology follow-up. Antibody screening four weeks later was significant for positive lupus anticoagulant and elevated PTT and DRVVT. She has remained on dalteparin without further recurrence of venous thromboembolism (VTE).

IMPLICATIONS/DISCUSSION: Recurrent thrombosis in the setting of therapeutic anticoagulation presents a life-threatening diagnostic dilemma. Possible etiologies include May-Thurner Syndrome, malignancy, warfarin failure, and antiphospholipid antibody syndrome. Warfarin failure can occur secondary to the presence of malignancy or due to variability in the depression of various clotting factors. Specifically, factor II is considered to be most important for warfarin's clinical antithrombotic efficacy, but least well represented in the INR. Furthermore, in 10% of patients with lupus anticoagulant, INR may be falsely elevated. Thus, in patients presenting with VTE in the setting of lupus anticoagulant, a factor II level can confirm adequate anticoagulation, especially prior to labeling a patient as a warfarin failure. Individuals presenting with new onset venous or arterial thrombosis in unusual locations should be screened for APS, a rare thrombophilic disorder most commonly observed in females of childbearing age. APS is diagnosed using the Sapporo criteria, which require a thrombosis or pregnancy loss in addition to repeatedly positive lab testing (lupus anticoagulant or antiphospholipid serologies), as tested at least 12 weeks apart. APS patients presenting with venous thrombosis can be managed as outpatients on warfarin with INR goal between 2-3 and should be considered candidates for long-term anticoagulation.
Lower Extremity Numbness in a Patient with Acute Arterial Thrombosis Secondary to Heparin Induced Thrombocytopenia Ryan Wesley Nall 1; Ryan Wesley Nall1. 1Beth Israel Deaconess Medical Center Internal Medicine, Boston, Massachusetts. (Proposal ID # 12262)

LEARNING OBJECTIVES:
1. Review the criteria used to determine the pre-test probability of heparin induced thrombocytopenia (HIT)
2. Review the role of serotonin release assay, platelet aggregation assay and the solid phase ELISA for heparin-dependent antibodies

CASE INFORMATION: 88 year old female status post left knee replacement presents from rehab with a one day history of ascending weakness and numbness in the lower extremity. Numbness started in her right toes and ascended to the level of the ankle over five minutes. A similar sensation developed moments later in the left foot. Associated symptoms included weakness with dorsiflexion of the feet and pain in the region of numbness. Patient denied other weakness or numbness, headache, vision changes, recent trauma, febrile illness or vaccination. Medications at rehab include Percocet, Lovastatin, and Fragmin. Examination revealed a temperature of 101, pulse of 110, and blood pressure of 130/80. Neurological examination of the lower extremity revealed profound weakness with dorsiflexion of the feet bilaterally, normal sensation to light touch, temperature, with decreased sensation to pin prick bilaterally. Patellar reflexes were brisk while achilles were absent bilaterally. The upper extremity neurological examination was normal. Cranial nerves II-XII were intact. Lower extremity was found to be cool to touch with absent dorsalis pedis, popliteal and femoral pulses bilaterally. Laboratory data revealed hematocrit of 30.2, white blood cell count of 9.6, platelets of 55. INR, PTT, renal function, liver function, and electrolytes were normal. Given the low platelet count and concern for HIT, all heparin products were held, heparin induced antibodies were measured and Argatroban was initiated. CTA of the chest, abdomen, pelvis, and lower extremity was performed and showed thrombus in the aorta, with flow down both iliac arteries, occlusion of the common femoral artery on the right, and popliteal artery on the left. Vascular surgery performed a thrombectomy of the left and right lower extremity thrombi. Heparin dependent antibodies later returned positive with an optical density of 2.895. The patient had a full recovery of sensation and function of her lower extremity.

IMPLICATIONS/DISCUSSION: Heparin induced thrombocytopenia (HIT) can often be a challenging diagnosis. One studied method of calculating the pretest probability of HIT is the 4T score. This score is based upon 1) the degree of thrombocytopenia and percent decline in platelet count 2) the timing of platelet decline to heparin exposure 3) thrombosis and 4) presence of other causes of thrombocytopenia. Based on this score patients fall into low, intermediate, and high probability for HIT. Testing for HIT should be performed on those with intermediate and high pretest probability of HIT. Diagnostic tests include: the serotonin release assay (SRA), platelet aggregation assay and the solid phase ELISA. The gold standard diagnostic test for HIT is the SRA with both specificity and sensitivity greater than 95%. However, this test is costly, technically difficult and results often take as long as a week to be reported. The solid phase ELISA determines the presence of heparin-dependent antibodies and results are reported much more rapidly than the functional assays (i.e. SRA, platelet aggregation assay). The ELISA has a sensitivity of >97 percent giving it an excellent negative predictive value. A positive test is often difficult to interpret as the test has a low specificity (74 to 86 percent). Warkentin et al showed that the strength of the ELISA result measured as an optical density can be helpful in predicting the likelihood of HIT. An optical density (OD) greater than 2.0 is associated with a positive serotonin release assay in 89 to 100 percent of patients. While an OD of 1.4 to <2.0 had a positive serotonin release assay of only 19 to 46 percent. Because the functional assay results are delayed the diagnosis of HIT should be considered established in patients with intermediate or high pretest probability and positive heparin induced antibodies. Optical density can be used to predict the likelihood of positive SRA and guide clinical decision making in a timely manner.