Kansas Journal of Medicine
Vol. 14, 2021; Supplement 1

Abstracts from the 2020 Annual Meeting and Virtual Conference
Kansas Chapter of the American College of Physicians

1. **Crystals Causing Confusion?: A Case of Crowned Dens Syndrome**
   Judges Award: Best Poster
   Christopher Williams, M.D., Zahra Rehman, M.D., Ghaith Noaiseh, M.D.
2. **Scrofula of the Abdomen**
   Chien Liu Co-Award: Best Infectious Disease Poster
   Maire Okoniewski, D.O., Kellie Wark, M.D., Rachel Weihe, M.D.
3. **The Effectiveness of a "Suppression Bundle" to Improve HIV Virologic Suppression in an Outpatient Infectious Disease Clinic**
   Chien Liu Co-Award: Best Infectious Disease Poster
   Victoria Poplin, M.D., Julia Katz, D.O., Guoqing Chen, M.D., Ph.D., MPH, Lisa Clough, M.D., Jessica Newman, D.O.
4. **When the Brain Diagnoses Your Cancer**
   Maisam Al Patty, M.D., Ammar Al-Obaidi, M.D., Vivar Cruz, M.D.
5. **Acute Cardiotoxicity Secondary to Immunotherapy**
   Ammar Al-Obaidi, M.D., Nathaniel Parker, D.O., Khalil Choucair, M.D., Jeremy Deutsch, M.D.
6. **Vascular Steal Syndrome Leading to Diffuse Cerebral Ischemia and Herniation in a Patient with Brain Arterio-Venous Malformations**
   Shandi Appier, M.D., Laxmi Dhakal, M.D., Mohinder R. Vindhyal, M.D., M.Ed.
7. **Primary Splenic Hemangiosarcoma: An Unusual Diagnosis with Astounding Presentation**
   Joseph Bennett, M.D., Miriam Baber, Natalia Roldan, M.D., Benjamin Powers, M.D., Janet Woodroof, M.D.
8. **VIPoma Empirically Treated with Octreotide**
   Charles Buess, M.D., Ryan Ford, M.D., Jeremy Deutsch, M.D.
9. **A Case of Extreme Hypernatremia**
   Eric Carrillo, M.D., Pharm.D., Mohinder R. Vindhyal, M.D., M.Ed.
10. **Acute Eosinophilic Pneumonia Due to Recent Medication Initiation**
    Jonathan Chandler, D.O., Laura Thomas, M.D.
11. **Severe Manifestations of Pernicious Anemia**
    Christopher Chapman, D.O., Mohammad Taha, M.D.
12. **On the Menu of Hospice Care Treatments is Costly HHT Treatment an Option?**
    Brianna Cline, D.O., Corey Braet, D.O.
13. **Dysarthria Associated with Lithium Toxicity: Case Report**
    Connor Ernst, M.D.
14. **Cause or Coincidence? Adult Onset Still's Disease and Celiac Disease Presenting Concurrently**
    Luke Frankl, M.D., Melissa Parkhurst, M.D.

15. **Hypokalemia - Common Yet with Multiple Possibilities**
    Chaitra Gopinath, M.D., Edgard Wehbe, M.D., George Elhomsy, M.D., Cyrus Munguti, M.D.
16. **Thyroid Type Papillary Carcinoma Arising in Mature Ovarian Teratoma**
    Ali Hamam, M.D., Daly Al-Hadeethi, M.D., Ammar Al-Obaidi, M.D., Georges Elhomsy, M.D., Kurt Rider, M.D.
17. **Primary Adrenal Insufficiency Secondary to Sarcoidosis**
    Corey Homan, M.D., Bakul Sangani, M.D.
18. **Intestinal Angiosarcoma: A Rare Cause of Upper Gastrointestinal Bleeding**
    Anthony Jackson, M.D., Luke Frankl, M.D., Mitesh Patel, M.D.
19. **An Unusual Case of an Elderly Male Newly Diagnosed with Collagenous Colitis and Celiac Disease**
    Luke Johnson, M.D., Chelsea Wuthnow, M.D., William Salyers, Jr., M.D., Paul A. Johnson, M.D.
20. **Short Telomere Related Pulmonary Fibrosis**
    Joe Johnstone, M.D., Andrea Covey, M.D.
21. **An Unusual Case of Urothelial Cell Carcinoma with Metastasis to the Pancreas**
    Katia El Jurdi, M.D., Ali Taleb, M.D., Khalil Choucair, M.D., William J. Salyers, M.D.
22. **Simultaneously Diagnosed Gastric Adenocarcinoma and Pernicious Anemia: A Classic Association**
    Syed Kamran, D.O., Mattias K. Dilling, Nathaniel A. Parker, D.O., Joel Alderson, D.O., Nathan D. Tofteland, M.D., Quoc V. Truong, M.D.
23. **Fahr's Syndrome: A Rare Cause of Status Epilepticus**
    Monica Khurana, M.D., Mohinder Vindyhal, M.D., M.Ed.
24. **An Uncommon Cause of Diabetic Ketoacidosis**
    Sean Lacy, D.O., Sushant Govindan M.D., M.Sc.
25. **Multiple Myeloma Associated with Dermatomyositis**
    Vijay Malladi, M.D., Donna Sweet, M.D.
26. **Brugada Syndrome: A Rare Case of Life-Threatening Dysrhythmias in a Young Lady**
    Job Mogire, M.D., Mohinder Vindhyal, M.D., M.Ed., Brent Duran, D.O., May Mohamad, M.D.
27. **Neuromyelitis Optica with Longitudinally Extensive Transverse Myelitis as a First Manifestation of Systemic Lupus Erythematosus**
    Maha Mohamad, M.D., Rami H Diab, M.D., May A. Mohamad, M.D., Timothy S. Shaver, M.D.
28. **Volunteering Saved My Life! A Case of Anomalous Right Coronary Artery Take Off**
    Grace Nassim, M.D., MPH, Patrick Ters, M.D.
29. **Suspected Anaphylaxis to Cosyntropin**
    Sonya Parashar, M.D., Corey Homan, M.D., David Naylor, M.D.
30. **Recurrent Dasatinib-Induced Chylothorax in Chronic Myeloid Leukemia: A Case Report**
    Jacob Ripp, D.O., Jessica Allen, Abdulraheem Yacoub, M.D.
31. **COVID-19 Related Acute Pancreatitis: A New Etiology?**
    Corey Braet, D.O., Ryan Ford, M.D., Paul Ndunda, M.D.

32. **Recurrent Pseudomonas Lung Infections in the Setting of Bronchiolitis Obliterans Following Allogeneic Stem-Cell Transplant**
    John Henry Carson, M.D., MBA
33. **Sickle Cell Disease and COVID 19**
    Karen Gichohi, M.D., Nicholas Ojile, M.D., Thomas Moore, M.D., Victor Bedros, MS-4
34. **A Rare Case of Human Monocytic Ehrlichiosis with Neurologic Presentation**
    Sophia Hitchcock, M.D., Luke Frankl, M.D., Katie Lester, M.D.
35. **Iatrogenic Cushing's Syndrome Due to Inhaled Glucocorticoids**
    Tyler Johnson, M.D., Lisa Clough, M.D.
36. **Cutaneous-Visceral Loxoscelism in an Adult with Bullae Dermatosis**
    Lorna Nguutu, M.D., Thomas Moore, M.D., Mohinder R. Vindhyal, M.D.
37. **COVID-19 and Heart Transplant: The Virus that Changed the Landscape of Medicine**
    Nikhil Parimi, M.D., Jane Titterington, M.D., Ph.D.
38. **Varicella Zoster Virus Meningitis in an Immunocompetent Adult Presenting as Isolated Intracranial Hypertension and Headaches**
    Pie Pichetsurnthorn, M.D., Pedro Vivar Cruz, M.D., Maha Assi, M.D., MPH
39. **Reducing Morbidity in Acanthamoeba Keratitis**
    Clarissa Smith, M.D., Nida Ashraf, MBBS, Kenneth Goins, M.D., Jessica Newman, D.O.
40. **A Case of Domestic Cholera Due to Lack of Personal Protective Equipment**
    Nora Strong, M.D., Maha Assi, M.D., MPH
41. **HIV Associated Non-Hodgkin Lymphoma: A Rare AIDS Defining Illness**
    Jordan Tichenor, D.O., MBA, Rachel Weihe, M.D.
42. **Encephalitis with Subdural Empyema Disguised as an Acute Stroke**
    Rhythm Vasudeva, M.D., Thomas Moore, M.D.
43. **When Treatment Becomes Disease: Disseminated BCG Infection after Intravesical Treatment for Bladder Cancer**
    Jerrica Werner, M.D., Lori Olson, M.D.
44. **Auscultated Blood Pressure Rounding Errors and Racial Disparities**
    Joshua Beyer, D.O., Robert Badgett, M.D., Hayrettin Okut, Ph.D.
45. **Tumor Mutation Burden, a Marker of Response to Immune Checkpoint Inhibitors: A Systematic Review and Meta-Analysis**
    Khalil Choucair, M.D., Hayrettin Okut, Ph.D., Ammar Al-Obaidi, M.D., Susan Morand, Laura Stanbery, Ph.D., K. James Kallail, Ph.D., John Nemunaitis, M.D.
46. **Resident and Patient Satisfaction with Bedside Rounds: Is There Room for Innovation?**
    Tanner Dean, D.O., Robert Badgett, M.D.
47. **Benefits of Aspirin for the Prevention of Esophageal Squamous Cell Carcinoma and Adenocarcinoma**
    Yasmine Hussein Agha, M.D., Sachin Srinivasan, M.D., Chelsea Wuthnow, M.D., Ali Taleb, M.D., Katia El Jurdi, M.D., Charles Buess, M.D., Nathan Tofteland, M.D., William Salyers, M.D.
48. **A Descriptive Study of IV Hydralazine Use in Adults with Hypertensive Urgency at an Academic Medical Center**
    Logan Kline, D.O., Shannon Stittsworth, Pharm.D., Abebe Abebe, M.D., Joanne Moore, RPh, Ethan Alexander, M.D.

49. **Disclosure of Unexpected Outcomes to Patients: Resident Attitudes and Practices Before and After Training Program**
    Nikki Miller, M.D., Leigh Eck, M.D., Becky Lowry, M.D., Marie Brubacher, M.D., Jessica Kalender-Rich, M.D.
50. **Assessing the Impact of a Diuresis Report on Electrolyte Management for Acute on Chronic Heart Failure**
    Dale Smith, M.D., Luke Nelson, M.D., Alex Robinson, D.O., Gong Weng, M.D., Laura Thomas, M.D., Heath Latham, M.D.
51. **Increasing Tdap Vaccination Rates in Internal Medicine Resident Clinic**
    Stephanie Slimmer, M.D., Grant Kirby, M.D., Robert Weidling, M.D., Raabia Nizamuddin, M.D., Maire Okoniewski, D.O., Joe Johnstone, M.D., Jamison Ryan, M.D., Autumn Brubaker, D.O.

Crystals Causing Confusion?: A Case of Crowned Dens Syndrome

Christopher Williams, M.D., Zahra Rehman, M.D., Ghaith Noaiseh, M.D.
KU School of Medicine-Kansas City

[Judges Award: Best Poster]

Introduction

Crowned dens syndrome is a rare manifestation of calcium pyrophosphate deposition disease (CPPD) resulting from acute inflammation due to calcium pyrophosphate crystal deposition near the odontoid process (dens) of the C2 vertebrae. This deposition forms a crown-like appearance around the dens on CT imaging. Clinical presentation can include fever, elevated inflammatory markers, neck pain, and, rarely, acute encephalopathy. The following case describes an atypical presentation of crowned dens syndrome involving acute encephalopathy in the setting of chronic CPPD.

Case Presentation

A 79-year-old African American male with a past medical history of mild cognitive impairment, osteoarthritis, and a family-reported diagnosis of rheumatoid arthritis not on active therapy presented to the hospital with a one-week history of progressive encephalopathy and inability to ambulate secondary to acute on chronic bilateral knee pain. Physical examination on admission revealed a somnolent patient oriented only to self and unable to provide a review of systems, bilateral knee effusions, and 3/5 strength in bilateral lower extremities. No neck stiffness was noted. The patient had a low-grade fever (99.3 degrees Fahrenheit) on admission. Admission laboratory studies were notable for ESR >130 and CRP 21.8 without leukocytosis. Urinalysis was unremarkable. Right knee arthrocentesis demonstrated WBC 8900 with 74% neutrophils, consistent with inflammatory arthropathy. No crystals or organisms were identified. Bilateral knee radiographs demonstrated bilateral menisci calcification consistent with CPPD arthropathy. The patient was initially started on empiric ceftriaxone therapy, but antibiotics were discontinued when infectious workup, including blood cultures and lumbar puncture, returned negative. The patient underwent non-contrast cervical spine CT which demonstrated calcific densities adjacent to the C2 odontoid process. Given the patient's altered mentation in the setting of a CPPD arthropathy flare and classic CT findings, a presumptive diagnosis of crowned dens syndrome was made. The patient was started on 20 mg IV methylprednisolone BID with significant improvement in mental status and arthralgias within 24-48 hours. Per chart review, patient was at his normal baseline at time of hospital discharge.

Conclusions

Few examples of crowned dens syndrome causing encephalopathy in the absence of neck stiffness or subjective complaints of neck pain exist in the literature. While the exact incidence is unclear, two studies found a 5% incidence of crowned dens syndrome in elderly patients presenting with acute neck pain based on radiographic findings. While uncommon, crowned dens syndrome should be considered in patients with a history of CPPD, evidenced by chondrocalcinosis on wrist, knee, or pelvic x-rays, presenting with encephalopathy in the setting of elevated inflammatory markers and negative infectious workup. Treatment of crowned dens syndrome includes corticosteroids and NSAIDs in patients without contraindications to NSAID use.

Scrofula of the Abdomen

Maire Okoniewski, D.O., Kellie Wark, M.D., Rachel Weihe, M.D.
KU School of Medicine-Kansas City

[Chien Liu Co-Award: Best Infectious Disease Poster]

Introduction

First described in 1843, tuberculous peritonitis (PT) is a rare entity even in high-endemic regions, where it makes up 0.1 – 0.7% of total tuberculosis (TB) cases. The insidious nature and varied presentations require a high clinical suspicion for diagnosis even in high-risk populations (2). We describe two cases of PT.

Case Presentation

Case 1 was a 32-year-old male from India, residing in the US for 8 years. Previously in excellent health, he presented with 6 months of intermittent epigastric pain, a 20-pound weight loss, occasional night sweats. No associated dyspnea, cough, hemoptysis, or constitutional symptoms. CT abdomen showed ascites, mesenteric lymphadenopathy and right iliac bone cortical irregularity. CT chest and spine were unremarkable. PET revealed multiple FDG-avid peritoneal nodularities and a hepatic lesion. Liver biopsy showed necrotizing granulomas; acid fast bacilli (AFB) and Grocott Methanamine silver (GMS) stains were negative. EGD duodenal biopsy resulted peptic duodenitis; colonoscopy was unremarkable. Ascitic fluid analysis was lymphocytic predominant (72%), adenosine deaminase (ADA) was normal (3.5 u/L). Fungal studies, M. tuberculosis complex (MTBC) PCR and AFB cultures were negative. After 10 months, open peritoneal biopsy was pursued. MTBC PCR was negative, but culture grew TB. He completed 2 months rifampin, isoniazid, pyrazinamide, and ethambutol, with planned additional 7-9 months of rifampin/isoniazid. Abdominal pain subsided with radiographic resolution of hepatic lesion, ascites, and improvement of ischial lesion by month 4.

Case 2 was a 42-year-old Vietnamese female residing in US for 23 years with annual visits to Vietnam and diagnosed with latent tuberculosis upon immigration. She completed a 9-month course of isoniazid. She had a history of 7 years of abdominal bloating, worsening for 2 years. CT showed ascites, peritoneal thickening, and enhancement with gastric enhancement. No thoracic pathology was noted. Ascitic fluid was lymphocytic (70%), ADA was normal (1.8 u/L). Bacterial, fungal, AFB cultures, and MTBC PCR were negative. Supraclavicular lymph node core biopsy revealed non-necrotizing granulomatous inflammation, with negative AFB, GMS stains, fungal, and AFB cultures. EGD revealed erosive gastropathy with negative biopsies. Colonoscopy was unremarkable. 6 months later, open peritoneal biopsy was done. Pathology showed necrotizing granulomas; negative MTBC PCR, AFB and GMS stains. AFB cultures positive 1-month later grew TB. Treatment was similar to regimen in Case 1. Eight-month CT showed resolution of ascites, peritoneal nodularities.

Conclusions

PT is divided into plastic and serous types, serous is more common and manifests with ascites and/or peritonitis. Plastic type presents as tender abdominal masses described as “doughy abdomen”. Symptoms include abdominal pain, fevers, night sweats, weight loss. It is diagnosed typically during surgery to resect masses or from acute abdomen presentation. In case series of patients with PT, diagnosis was suspected before death in 11 of 20. Ascitic fluid is lymphocytic predominant and exudative, AFB rarely positive, and cultures positive in 25% of cases. Ascitic ADA has high sensitivity and specificity but it proved unhelpful in both above cases.

The Effectiveness of a "Suppression Bundle" to Improve HIV Virologic Suppression in an Outpatient Infectious Disease Clinic

Victoria Poplin, M.D., Julia Katz, D.O., Guoqing Chen, M.D., Ph.D., MPH, Lisa Clough, M.D., Jessica Newman, D.O.
KU School of Medicine-Kansas City

[Chien Liu Co-Award: Best Infectious Disease Poster]

Introduction

As of 2018, approximately 37.9 million people worldwide were living with HIV, with nearly 1.1 million of those living in the US alone. Roughly 62% of the world's adult HIV-positive population was receiving combination antiretroviral therapy (cART). Combination ART is highly effective and leads to dramatic reduction in HIV-associated morbidity and mortality and leads to decreased HIV transmission when HIV is suppressed to undetectable levels. Despite the effectiveness of cART, only 49% of people with HIV in the United States in 2014 had suppressed viral loads. Strategies to improve HIV suppression are needed.

Methods

Viral load data were collected from HIV-positive patients receiving care in the 24 months preceding the quality improvement project. Patients with a detectable viral load (> 40 copies of virus/mL) were included in the intervention group. A "Suppression Bundle" consisting of three to five interventions was selected for each patient by a team consisting of the patient's HIV physician, case manager, and an HIV pharmacist. Baseline demographics were collected. A three-month lead-in time was given to begin interventions. Viral load data were collected at the completion of a 9-month suppression bundle intervention period.

Results

Prior to the study initiation, there were 567 HIV-positive patients actively receiving care within the outpatient clinic. Of those, 89 patients had a measurable viral load (> 40 copies/mL). Of the 89 patients with a detectable viral load, 65 patients participated in the suppression bundles. At the completion of the 9-month intervention period, 51 of the 65 had a viral load < 200 copies/mL and 46 had a viral load < 40 copies/mL, demonstrating a substantial improvement with 70.1% of the previously non-suppressed patients having suppressed HIV viral loads.

Conclusions

Improving viral suppression in non-suppressed HIV positive patients is a complex issue. There are a multitude of factors that contribute to a non-suppressed viral load and/or medication non-adherence. By identifying patient risk factors, an individualized "suppression bundle" can be developed to improve HIV suppression.

When the Brain Diagnoses Your Cancer

Maisam Al Patty, M.D., Ammar Al-Obaidi, M.D., Vivar Cruz, M.D.
KU School of Medicine-Wichita

Introduction

Paraneoplastic cerebellum degeneration (PCD) is a rare non-metastatic complication of any type of cancer. PCD is caused by the immune system fighting primary systemic cancer rather than other mechanisms like metastases, infections, metabolic or side effects of cancer treatment. We present the fourth reported case of PCD attributed to papillary urothelial carcinoma as primary cancer.

Case Presentation

A 69-year-old male presented with a rapidly progressing gait disturbance, nausea, vomiting, headache, and double vision over two weeks period. He had a 50-pack year smoking history. Neurological exam was significant for dysarthric speech, nystagmus, significant ataxia during figure to toe and heel to shin. MRI brain was significant for leptomeningeal enhancement. Initial LP demonstrated atypical cells in the CSF concerning for metastatic disease. CSF paraneoplastic panel was positive for PCA-Tr antibody which strongly associated with Hodgkin lymphoma. Repeated LP showed no malignant cells and again was positive for PCA-Tr antibody. Additional pan CT was negative except for a mass in the left ureter concerning for urothelial cancer, suspected as the primary lesion. Cystoscopy and left ureteroscopy demonstrated a large papillary lesion; biopsy was taken with a finding of high grade papillary urothelial carcinoma. Additional work up with bone marrow biopsy and flow cytometry were insignificant. He received 5 days of IVIG and IV steroid which improved his neurological symptoms. He is following with the oncology clinic for urothelial cancer management options and the neurology clinic with a plan for IVIG and steroid, two doses monthly. Patient showed stable clinical picture with his current treatment.

Conclusions

In this case, MRI brain showed finding of cerebellum leptomeningeal enhancement, CSF didn't detect malignant cells, however, it showed Anti-Tr antibodies that appear to be highly associated with Hodgkin lymphoma. An extensive workup detects papillary urothelial carcinoma without any other evidence of lymphoma. These antibodies had been detected in the CSF only and not in serum, which is not unusual for PCD. Brain imaging mostly presents with normal cerebellum especially during the initial stage of the syndrome, diffuse atrophy can be seen in more advanced stages. In our patient, MRI showed cerebellum leptomeningeal enhancement which has less seen but reported before. Mainstay management of the PCD is focused on primary tumor treatment. Routinely, immune therapy has been tried to improve the neurological outcome; however, it not necessarily associated with recovery. PCD has overall poor prognosis, it varies according to the associated antibody and the underlying type of cancer. Better survival is seen with anti-Tr antibodies as compared to others. PCA-Tr antibody is one of the antibodies that predominantly associate with PCD, it usually specific for Hodgkin lymphoma, but can also be seen with papillary urothelial carcinoma.

Acute Cardiotoxicity Secondary to Immunotherapy

Ammar Al-Obaidi, M.D., Nathaniel Parker, D.O., Khalil Choucair, M.D., Jeremy Deutsch, M.D.
KU School of Medicine-Wichita

Introduction

Immune checkpoint inhibitors showed significant antitumor activity in multiple malignancies and have become essential oncology standard-of-care therapies. Despite their success, they are associated with a unique panel of side effects known as immune-related adverse events (irAEs). Classic irAEs involve the skin, GI system, endocrine organs, lungs, kidneys, joints, and liver. Myocardial involvement remains an extremely rare, yet fatal adverse event.

Case Presentation

A 52-year-old smoker female with COPD, recent diagnosis of EGFR/ALK/ROS1-negative, stage IV non-small cell lung cancer, was enrolled in an immunotherapy clinical trial with upfront nivolumab plus Ipilimumab. A pre-enrollment TTE was unremarkable with LVEF of 69%. Six-month follow-up PET/CT scan showed partial response without disease progression, so she remained on nivolumab and ipilimumab. One year later, she presented to the ED with acute hypoxic respiratory failure and required noninvasive positive pressure ventilation. Imaging suggested new interstitial edema, no pulmonary emboli, or pericardial effusion, but cardiomegaly was noted. ECG revealed mild sinus tachycardia. Initial serum laboratory testing was equivocal. TTE revealed a severely reduced LVEF of 15-20% with significant regional wall motion abnormalities, moderate pulmonary artery hypertension and dilated inferior vena cava (IVC). However, cavity size and wall thickness were normal with no valvular abnormalities. Left heart catheterization revealed normal coronaries without blockages. Bedside spirometry showed mixed ventilatory defect consistent with her COPD. It was determined that she had new-onset heart failure secondary to combination immunotherapy. High dose intravenous methylprednisolone started along with aggressive IV diuresis which improved her oxygen requirements. Follow-up TTE on hospital day 8 showed near-complete resolution of systolic function with LVEF of 45-50% without regional wall motion abnormalities or IVC dilation. She was dismissed on a 6-week slow oral steroid taper. A 3-month follow-up TTE showed LVEF of 50-55%.

Conclusions

Immunotherapy-induced myocarditis has been reported in few cases and it may result in poor outcomes if not recognized and managed. The precise pathophysiology is poorly understood but thought to be due to dysregulation of the auto-reactivity mechanisms that are usually maintained by immune checkpoints. Grade 2 irAEs are treated by withholding immunotherapy. Prednisone 0.5 mg/kg/day is usually given if symptoms persist for a week of withholding immunotherapy. Should symptoms be grade 1 or less, immunotherapy can be resumed. Severe or life-threatening cases are treated with high doses of corticosteroids with gradual tapering for at least a month when symptoms subside to grade 1 or less plus permanently discontinuing immunotherapy. This case highlighted the importance of pre-treatment cardiac screening despite the rarity of this condition due to its potentially fatal complications. It also signified the multidisciplinary team approach to manage the cardiac complications, particularly cardiac arrhythmias, and LV systolic dysfunction.

Vascular Steal Syndrome Leading to Diffuse Cerebral Ischemia and Herniation in a Patient with Brain Arterio-Venous Malformations

Shandi Appier, M.D., Laxmi Dhakal, M.D., Mohinder R. Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

Introduction

Arteriovenous malformations (AVMs) in the brain are an uncommon but potentially devastating occurrence of arterial to venous connections without an intervening capillary network. These AVMs are typically discovered after intracranial hemorrhage, seizures, or as an incidental finding. We present a case of brain AVMs that manifested as transient episodes of focal neurologic deficit consistent with vascular steal syndrome – the phenomenon in which arterial blood is shunted through low-resistance arteriovenous fistulae away from the higher-resistance capillary bed in the healthy brain, followed by rapid progression to diffuse cerebral ischemia and herniation.

Case Presentation

A 48-year-old woman initially presented to the Emergency Department due to an acute onset of slurred speech accompanied by left facial and right-hand paresthesia. Soon after admission, her neurologic symptoms had resolved. Computed Tomography (CT) angiography of the head/neck showed diffuse dural arterio-venous (AV) fistulas, which was confirmed on magnetic resonance imaging (MRI). A 4-vessel cerebral angiogram demonstrated extensive AV fistulas arising from the bilateral internal carotid and vertebral arteries. Following this study, the patient had an acute decrease in consciousness level with left upper extremity rigidity and later left visual neglect. A repeat CT angiography was negative for dissection, thrombus, or hemorrhage. A follow-up MRI showed a tiny focus of new ischemia in bilateral corona radiata. Her symptoms resolved over the next two days, and she was discharged home with antiepileptics and neurology follow-up.

One year later, the same patient returned to the hospital for a 2-day history of worsening headache with sudden onset of slurred speech and difficulty following directions. A CT head was negative for acute findings. Shortly after a magnetic resonance angiogram and venogram (MRA/MRV) was obtained, the patient acutely demonstrated decreased level of consciousness and vomiting. She was emergently intubated after an episode of aspiration with desaturation, and the imaging revealed diffuse bilateral ischemia and cerebral edema with descending cerebellar tonsillar and uncal herniation. The elevated opening pressure of 33 cm H2O was obtained on lumbar puncture (LP), and an external ventricular drain (EVD) was placed. Over the next seven days, the patient's intracranial pressures (ICP) and cerebral perfusion pressures (CPP) became increasingly difficult to manage, and she exhibited loss of primitive reflexes. Her status continued to decline until she passed away from cardiopulmonary arrest secondary to brain herniation.

Conclusions

Clinical symptoms of AVMs in the brain are routinely associated with intracranial hemorrhage and seizures. However, it is important to recognize vascular steal syndrome as a manifestation of brain AVMs, which presents with waxing and waning focal neurologic deficits. Furthermore, this condition can have massive cerebral ischemic consequences leading to cerebral edema and subsequent fatal herniation.

Primary Splenic Hemangiosarcoma: An Unusual Diagnosis with Astounding Presentation

Joseph Bennett, M.D., Miriam Baber, Natalia Roldan, M.D., Benjamin Powers, M.D., Janet Woodroof, M.D.
KU School of Medicine-Kansas City

Introduction

Primary Splenic Hemangiosarcoma (PSA) is one of the rarest and most aggressive tumors of vascular origin. The annual incidence is estimated around 1 in 5 million people. Presenting symptoms are often non-specific but may include fatigue, weight loss, weakness, abdominal pain. Due to the aggressive nature and indolent non-specific signs/symptoms, prognosis is often poor by time of diagnosis. Here, we present a rare case that offers insight into this unique entity.

Case Presentation

A 21-year-old female with medical history of celiac disease presented to the emergency department with complaint of lower back pain. Her symptoms began 6 months prior after falling off a horse; pain was progressive to the point of being unable to walk on the day of admission. On review of symptoms, she noted left upper quadrant pain, abdominal distention, night sweats, and weight loss. Initial exam was noteworthy for massive, tender splenomegaly to pelvic brim. Her labs were notable for hemoglobin of 9.3 g/dL, platelet count of 8 K/uL, LDH 462 U/L, and negative mono-spot. CT abdomen/pelvis showed marked splenomegaly with innumerable lytic osseous lesions. There was concern for an aggressive hematologic malignancy. A bone marrow aspiration/biopsy showed a vascular sarcoma. PET/CT revealed a hypermetabolic spleen and redemonstration of osseous lesions. After inter-institutional collaboration with Mayo Clinic, a diagnosis of high grade angiosarcoma was made, likely arising from the spleen. She was started on a regimen of gemcitabine plus docetaxel with G-CSF support. After 3 cycles, restaging PET scan showed responsive disease. Despite the improvement in PET imaging and platelet count, chemotherapy was not making her days more enjoyable, so she elected for home hospice. She died approximately 6 months after her last cycle of chemotherapy.

Conclusions

PSA is a rare and often confounding vascular malignancy. Due to its highly aggressive nature, the chance of metastases is extremely high, with most common locations being liver, lungs, lymph nodes, and bones. PSA has been associated with ionizing radiation, arsenic, vinyl chloride, and prior chemotherapy, but most cases occur without risk factors, as in our patient. In up to 30% of patients, presentation may involve a severe, life-threatening hemorrhagic shock due to spontaneous splenic rupture. Early detection and resection are integral components of management in hopes of preventing metastasis, but due to vagueness of symptoms/signs, this is not common to catch it early enough. Although rare, splenic angiosarcoma should be on the differential diagnosis of patients with massive splenomegaly and insidious constitutional symptoms.

VIPoma Empirically Treated with Octreotide

Charles Buess, M.D., Ryan Ford, M.D., Jeremy Deutsch, M.D.
KU School of Medicine-Wichita

Introduction

Pancreatic neuroendocrine tumors are rare neoplasms. Arising in the endocrine tissues of the pancreas, they can secrete a variety of peptide hormones, including gastrin, glucagon, insulin, and vasoactive intestinal peptide. Herein is a case of a patient with severe, chronic diarrhea who experienced refractory hypotension, renal failure requiring dialysis, and severe diarrhea. He was treated with empiric intravenous octreotide drip prior to definitive diagnosis and experienced rapid improvement of his symptoms. He later underwent successful laparoscopic distal pancreatectomy with splenectomy and complete tumor resection. VIP level subsequently returned drastically elevated.

Case Presentation

A 30-year-old male with past medical history of 6 months chronic diarrhea presented to the ED with worsening diarrhea and abdominal pain. CT scan showed a soft tissue mass in the anterior body of the pancreas measuring up to 5.6 cm. He had very frequent and large liquid bowel movements and quickly developed hypotension. GI panel and C-diff were negative. He was given upwards of 12L of IV fluid, however, he remained tachycardic in the 120s and hypotensive. He was placed on norepinephrine. He was given Loperamide to no avail. He soon became anuric, requiring two rounds of dialysis. Rectal tube was placed and showed nearly 6L output during a twelve hour shift alone. Empiric treatment for pancreatic neuroendocrine tumor was initiated with octreotide GTT 25 mcg/hr. The following morning, approximately 12 hours later, the patient showed significant improvement with decreased stool output down to 1L in a 12-hour period. His vitals rapidly improved, norepinephrine was discontinued, he began producing urine, and dialysis was discontinued. EUS was performed with FNA which showed an initial impression of a 6.5 cm x 5.1 cm neuroendocrine tumor that appeared to have invaded the splenoportal confluence and splenic artery/vein. He underwent successful laparoscopic distal pancreatectomy with splenectomy and solitary tumor resection. The patient's labs returned unremarkable for gastrinoma, glucagonoma, insulinoma, and carcinoid. Subsequent labs showed VIP 4360. Post-op pathology showed a 7 cm x 5.7 cm x 4.5 cm, grade 1, T3N0M0, Stage 2 neuroendocrine tumor of the pancreas with negative margins. There was no need for adjuvant therapy, and he will return for CT chest/abdomen/pelvis, VIP levels, CBC, Chromogranin A, Urine 5-HIAA every 4 months for continued surveillance.

Conclusions

This is a case where empiric octreotide therapy was administered prior to a definitive diagnosis of neuroendocrine tumor. Prior to confirmation via lab values and EUS, the only evidence supporting a diagnosis of neuroendocrine tumor was clinical course and CT abdomen findings. Although there was no definitive diagnosis when beginning treatment, there was a place for empiric octreotide therapy. There is great benefit to be had from further research regarding this subject matter.

A Case of Extreme Hypernatremia

Eric Carrillo, M.D., Pharm.D., Mohinder R. Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

Introduction

Hypernatremia is an increase in plasma sodium concentration above that of 145 mM. Hypernatremia is usually the result of a combined water and electrolyte deficit, with losses of H_2O in excess of that of sodium. Hypernatremia can result from iatrogenic administration of excess sodium, as after IV administration of hypertonic normal saline or sodium bicarbonate. Hypernatremia causes an increased osmolality in the extracellular fluid, which results in cellular shrinkage. Cells (brain cells in particular) accommodate chronic increases in extracellular fluid by activating membrane transporters that allow the intracellular accumulation of organic osmolytes which maintains an intracellular osmolality similar to that of the extracellular fluid. This cellular response, however, predispose patients with chronic hypernatremia to the development of cerebral edema with rapid correction of hypernatremia and osmolality mediated fluid shifts into and out of cells. We present a case of severe hypernatremia.

Case Presentation

A 77-year-old male with past medical history of CVA with chronic right-sided weakness, heart failure, and gastrostomy tube dependence presented to the emergency department for evaluation status post cardiac arrest. The patient had been found progressively less responsive by family prior to calling EMS. There was report of blood in patient's mouth and recently in his stool. On arrival, EMS noted he was confused but awake and alert. Enroute to hospital, heart rate and blood pressure declined to a critical degree and pulses were lost. CPR was initiated and ROSC was obtained after administration of a milligram of epinephrine. He was intubated in the field. Preliminary diagnosis was that of severe GI bleed due to the report of hematochezia and hypotension. Patient was profoundly anemic and transfused as appropriate. Preliminary basic metabolic profile revealed multiple electrolyte abnormalities, with the most pronounced being a sodium level of 193 mM. Immediate recheck of this lab value yielded results of 180 mM. These values correspond to a free water deficit of roughly between 8.5 and 9 liters in this patient. Over the next four days, serial basic metabolic panels were drawn to monitor sodium levels, as intravenous fluids of various sodium concentrations were given to slowly correct the extreme degree of hypernatremia. Over this period, the sodium concentration of the plasma was titrated down to 170 mM. At this point in time, the family opted for hospice care due to the patient's significant other comorbid conditions

Conclusions

This patient, who was gastrostomy tube dependent and struggled to communicate needs to caregivers, most likely suffered from increasingly chronic hypernatremia. In cases of extreme hypernatremia, clinicians need to be aware that prolonged correction of plasma sodium concentration is imperative to prevent cerebral edema.

Acute Eosinophilic Pneumonia Due to Recent Medication Initiation

Jonathan Chandler, D.O., Laura Thomas, M.D.
KU School of Medicine-Kansas City

Introduction

Drug reaction with eosinophilia and systemic symptoms (DRESS) is a difficult diagnosis to make with its variable presentations. We report a case of a young female that presented with respiratory failure, and ultimately thought to have DRESS with Acute Interstitial Nephritis (AIN) and Acute Eosinophilic Pneumonia.

Case Presentation

A 20-year-old female with a history of recurrent UTIs and depression, recently started on escitalopram and topiramate, presented to the emergency room with fevers and dyspnea. She rapidly developed worsening hypoxemia necessitating intubation, started on broad spectrum antibiotics for septic shock, and was transferred to another hospital. On presentation to the subsequent hospital, she underwent a large infectious workup, as family had reported the patient had recently gone camping. Workup initially yielded a positive Babesia Microti PCR. Her antibiotics were tailored to include quinine and clindamycin and was found to have < 1% involvement of babesia on peripheral smear with no evidence of hemolysis. Throughout her hospitalization, she developed progressive peripheral eosinophilia. Due to worsening oxygenation and development of ARDS, the patient was transferred to our institution. On arrival, she was found to have an acute kidney injury (AKI), likely from acute interstitial nephritis (AIN), peripheral eosinophilia (30%), a fine maculopapular rash, and underwent bronchoscopy revealing significant eosinophilia (41%). After discussion with Infectious Disease, Nephrology, Dermatology, and Hematology, a decision was made to initiate corticosteroids and empiric Ivermectin. Two days after presentation to our institution, we were informed the initial PCR for Babesia microti was misreported as positive. Antimicrobials were subsequently weaned, as infectious workup was unyielding and bone marrow biopsy was negative for malignancy or HLH. It was ultimately decided that the patient's presentation was DRESS, from escitalopram or topiramate that had been started two weeks prior to initial presentation. The patient slowly improved and completed a five-month steroid taper.

Conclusions

Drug reaction with eosinophilia and systemic symptoms (DRESS) is difficult to diagnose due to its varied clinical presentation and diverse laboratory abnormalities. With its wide range of presentations, many cases go undiagnosed limiting our knowledge of its true incidence. While our patient did meet diagnostic criteria for DRESS by RegiSCAR and Japanese consensus groups, the patient's treatment course was initially anchored to the incorrectly reported Babesia Microti PCR. Additionally, our patient's lack of initial liver enzyme elevation led to an initial lower suspicion for DRESS. While elevation of liver enzymes is extremely common, it only occurs in up to 70% of cases. On presentation to our institution, the patients AIN and eosinophilic lung disease led to initiation of steroids, both of which are known to be associated with DRESS. DRESS has a wide array of presentations and overlap with other clinical entities making it a difficult clinical dilemma for physicians.

Severe Manifestations of Pernicious Anemia

Christopher Chapman, D.O., Mohammad Taha, M.D.
KU School of Medicine-Kansas City

Introduction

Pernicious anemia results from any process that interferes with the intestinal absorption of cobalamin, or Vitamin B12. Generally insidious in onset, classic presentations include macrocytic anemia, glossitis, and ataxia of the lower extremities. Common neurologic findings stem from demyelination of the posterior columns of the spinal cord, termed subacute combined degeneration (SACD). Rarely, in the case of profound cobalamin deficiency, patients may present with severe anemia that mimics hematological malignancies and with neurological findings that extend beyond the central nervous system. This case report details a patient with such a presentation.

Case Presentation

The patient is a 43-year-old African American male without significant past medical history presenting for evaluation after he fell while attempting to get out of bed that morning. He noted the fall was preceded by several weeks of fatigue and leg weakness with painful muscle spasms. He initially denied difficulties voiding but later had bowel incontinence, urinary retention, and impotence. Exam showed mucosal pallor, decreased muscle bulk in lower extremities, increased reflexes at the patellar and Achilles tendons, and absent vibration sense below the knees. Laboratory evaluation revealed pancytopenia with hemoglobin of 4 with MCV 139.4, white cell count of 3000, and platelet count of 31000. Peripheral smear demonstrated no nuclear or membrane abnormalities to suggest autoimmune hemolytic anemia. Absolute reticulocyte count was low. Hematology recommended a bone marrow biopsy, which showed hypercellular marrow with decreased erythrocyte and megakaryocyte production. Serum cobalamin concentration was < 50pg/mL. Intrinsic factor blocking antibodies were identified in the serum. Neurology recommended MRI of cervical, thoracic, and lumbar spine revealing diffuse symmetric T2 hyperintensity of anterior, lateral, and posterior columns in the cervical and thoracic region. The patient was transfused two units of packed red cells and started on intramuscular cobalamin with improvement in his cell counts throughout his hospital course.

Conclusions

It is estimated that 1-2% of adults aged 40-59 years in the U.S. have cobalamin deficiency due to pernicious anemia and the prevalence increases with age. Our case demonstrates several rare characteristics of this disease process including pancytopenia, anterior cord involvement, and autonomic dysfunction. The severity of presentations of pernicious anemia varies widely depending on the degree of cobalamin deficiency. The presence of intrinsic factor antibodies is 100% specific for pernicious anemia though prior administration of cobalamin may give a false positive result. SACD can affect the entire nervous system with symptoms ranging beyond what would be seen in classic posterior column distribution affecting the lower extremities. Neurological recovery is variable even with adequate treatment. Patients with confirmed antibody mediated pernicious anemia require lifelong parenteral cobalamin.

On the Menu of Hospice Care Treatments is Costly HHT Treatment an Option?

Brianna Cline, D.O., Corey Braet, D.O.
KU School of Medicine-Wichita

Introduction

Hereditary Hemorrhagic Telangiectasia, or HHT, is a rare autosomal dominant disorder. Patients present with telangiectasias and arteriovenous malformations, which often lead to life-threatening bleeding and anemia. Although the disease is rare, there are treatments, including the costly medication bevacizumab, which can alleviate bleeding and symptomatic anemia in these patients. Subsequently the issue of whether the drug may be continued when these patients pursue palliative care or hospice may arise.

Case Presentation

A 53-year-old male with a past medical history significant for HHT and alcoholic cirrhosis with a history of variceal bleeding was admitted to the hospital for complaint of shortness of breath, increased abdominal girth, decreased urine output, and black stools. On admission, labs were significant for a hemoglobin of 4.6 g/dl. The patient had been placed on bevacizumab treatment for HHT in the past and had good clinical response for approximately nine months. After three months off bevacizumab treatment, the patient began to exhibit dark stools and symptomatic anemia. His oncologist resumed bevacizumab treatments approximately one month prior to this admission. A gastroenterology consult ruled out repeat variceal bleeding. The patient's hemoglobin remained stable after two units of packed red blood cells were transfused, and his melena resolved. However, the patient's kidney function declined, and he was diagnosed with hepatorenal syndrome. On day six of admission, the patient was transferred to the ICU for decompensation of respiratory status. He was diagnosed with hepatopulmonary syndrome, which has a very poor prognosis. A goals of care discussion with the patient revealed that he wished to pursue home hospice care, however, the patient was concerned about continuing his bevacizumab treatment for HHT. This led to a very important discussion regarding the continuation of costly treatments of rare conditions in patients pursuing hospice care.

Conclusions

When transitioning to hospice care, it is often difficult to determine what treatments are appropriate to continue. In this patient, hepatopulmonary syndrome served as the hospice diagnosis, and because HHT was not the principle diagnosis, bevacizumab treatment was continued. Bevacizumab costs around $100,000 per year. When a patient is admitted to hospice, the hospice service acquires the financial responsibility for the patient's treatment, so often, companies are disinclined to continue this kind of treatment. However, when a patient is having significant symptom relief from the treatment, and the condition being treated is not the primary diagnosis for hospice admission, it is reasonable to continue.

Dysarthria Associated with Lithium Toxicity: Case Report

Connor Ernst, M.D.
KU School of Medicine-Wichita

Introduction

Lithium has widely been considered the mainstay of treatment for acute and chronic bipolar disorder for the last half century. Optimal management of lithium requires strict monitoring and titration due to its narrow therapeutic index and significant side-effect profile. Speech disturbances are recognized as a relatively rare effect of elevated lithium levels. We present a case of lithium toxicity manifesting primarily with neurologic and cognitive deficits.

Case Presentation

A 36-year-old female with a past medical history of bipolar disorder, pseudoseizures, unspecified personality disorder, hypertension, and hypothyroidism presented to the emergency department due to subacute, progressively worsening dysarthria and cerebellar ataxia with occasional vomiting and diarrhea. Upon further history, the patient's home lithium dose was increased by 50% four months prior to admission by her primary care provider with minimal follow-up monitoring. On examination, her speech was characterized by increased effort and fluency deficits. Comprehension, reading, and writing were all spared. Additionally, one episode of unilateral upper extremity spasm was observed and her gait was unsteady. Lithium level on admission was twice the upper limit of normal. Kidney function was mildly depressed. Imaging of her brain was unchanged as compared to three years prior. She was observed in the hospital with rapid improvement in symptoms which corresponded with decreased lithium levels.

Conclusions

Identifying signs of lithium toxicity is challenging. Typical lithium intoxication involves predominantly gastrointestinal side effects, but neurologic deficits may become apparent when chronic. Neurologic sequelae vary and include tremors, ataxia, encephalopathy, dysarthria, and can progress to coma without intervention. This case demonstrates a subacute presentation of lithium toxicity that was promptly identified and treated before the development of irreversible damage. Often with severe lithium intoxication, hemodialysis is indicated. Regular serum lithium levels are a vital measurement to titrate therapy, not only when re-dosing, but also to gauge physiologic clearance.

Cause or Coincidence?
Adult Onset Still's Disease and Celiac Disease Presenting Concurrently

Luke Frankl, M.D., Melissa Parkhurst, M.D.
KU School of Medicine-Kansas City

Introduction

Adult Onset Still's Disease (AOSD) is a rare systemic inflammatory syndrome presenting with fever, rash, and arthralgias. The quotidian fever, often the first symptom, is stereotypically high-spiking (> 39C) and lasts < 4h. Arthralgias typically affect wrists, knees and ankles and initially are mild and transient. Long-standing disease can progress to destructive polyarthritis. The rash presents as evanescent salmon-pink erythema predominantly affecting proximal limbs and trunk. Histopathology reveals a mixed inflammatory infiltrate. Other common signs/ symptoms include myalgias, pharyngitis, liver inflammation, splenomegaly, and lymphadenopathy. We present a case of AOSD with extra symptoms of diarrhea and weight loss.

Case Presentation

A 28-year-old man presented with one month of low-grade, undulating fever, 20 lb weight loss, pharyngitis, diaphoresis, chills, progressive pruritic rash, arthralgias, and diarrhea. He had multiple well-demarcated, salmon-colored, urticarial plaques on his trunk, upper and lower extremities, feet, and palms. He had an erythematous oropharynx without tonsillar exudates. There was no arthritis, cardiac or pulmonary anomalies, or palpable hepato-splenomegaly. Labs revealed ANC 7.5, Abs Eos 0.72, ESR 32, CRP 8.1, AST 162, ALT 98, ANA titer 1:160, LDH 485, ferritin > 7500, triglycerides 115, TTG IgA > 250. Infectious disease workup was unrevealing aside from prior EBV infection. Exhaustive rheumatologic serologies workup was negative indicating low likelihood for autoimmune disease including lupus, RA, scleroderma and multiple vasculitides. Skin biopsy consistent with urticarial eruption. PET scan positive for diffuse hypermetabolic lymphadenopathy and bone marrow. Bone marrow biopsy with non-specific inflammatory changes without findings of lymphoproliferative disorder. Inguinal lymph node biopsy revealed benign lymphoid tissue. Endoscopy with normal appearing mucosa of upper and lower GI tract. Duodenal biopsies revealed marked villous blunting and increased intraepithelial lymphocytes. He was diagnosed with Adult Onset Still's Disease and Celiac Disease. He was treated with gluten free diet and prednisone with significant improvement in his symptoms.

Conclusions

AOSD is a clinical diagnosis with no definitive diagnostic test. However, several diagnostic criteria have been proposed for AOSD with sensitivity as high as 93.5%. These criteria account for the necessary exclusion of potential mimickers which include infectious, neoplastic, and other autoimmune conditions prior to diagnosis of AOSD. Pathogenesis of AOSD includes aberrant immune system activation that induces the release of multiple pro-inflammatory cytokines including IL-1, IL-6, IL-17 and IL-18. Like most autoimmune disease, AOSD pathogenesis is likely related to multiple genetic and environmental factors. Multiple infectious diseases and malignancies have been postulated as inciting events, yet no specific cause and effect pattern has been revealed. The above case provides an example of celiac disease presenting concomitantly with AOSD and possibly representing the inciting event for the systemic inflammatory response. Moving forward, clinicians should have a low threshold to screen for celiac disease in patients being worked up for suspected AOSD.

Hypokalemia - Common Yet with Multiple Possibilities

Chaitra Gopinath, M.D., Edgard Wehbe, M.D., George Elhomsy, M.D., Cyrus Munguti, M.D.
KU School of Medicine-Wichita

Introduction

Hypokalemia is a common entity in hospitalized patients and severe hypokalemia, defined as potassium less than 2.5 mEq/L can be life threatening. Though diuretic use and loss of potassium through GI tract are most common causes of hypokalemia; evident from history and physical exam, diagnosis of other causes of hypokalemia can be challenging. We present a case of severe hypokalemia who presented with altered sensorium.

Case Presentation

A 58-year-old, white, female was brought to the hospital after her husband noticed that she had been increasingly confused for 3 days with poor oral intake. She had a past medical history significant for schizophrenia, had not seen a primary care physician for years and was not on any medications. Initial exam revealed she was febrile, tachycardic, hypotensive, confused and had an abscess over right axilla with easily expressible thick exudate. Laboratory exam revealed leukocytosis, profound hypokalemia, and an ABG showed metabolic alkalosis. CT brain showed no acute changes. Patient was treated with fluids, broad spectrum intravenous antibiotics and potassium replaced. Patient improved hemodynamically but continued to have profound hypokalemia requiring daily replacement. Further evaluation for hypokalemia was done and urine K+ level obtained, which was indicative of renal K+ loss. On exam patient was found have mild hirsutism, acne, truncal obesity which though noted, were not clinically correlated during the initial encounter for an acute presentation. Serum renin, aldosterone and cortisol ordered. CT abdomen performed which revealed a 10cm adrenal mass over the left adrenal gland with CT features suggestive of adrenal myelolipoma. Serum cortisol, renin and aldosterone returned as normal. However due to a high clinical suspicion of Cushing's low dose dexamethasone suppression test, 24-hour urine cortisol, serum ACTH levels, DHEAS were obtained. Plasma metanephrine was obtained to rule out pheochromocytoma. Patient had a serum cortisol above 5 after a low dose dexamethasone suppression highly suggestive of Cushing's. 24-hr urinary cortisol was the upper limit of normal range. ACTH was 30th percentile. Patient was recommended surgery due to the size of the adrenal mass and work up suggestive of Cushing's. Patient underwent surgery and pathology of the specimen confirmed myelolipoma.

Conclusions

Hypokalemia is a common entity encountered by Internists in both in-patient and out-patient setting. Though the common causes are easy to ascertain, other etiologies like an underlying endocrine disorder may be missed due to subtle clinical features. A high clinical suspicion is essential to consider a broad differential, requiring a multidisciplinary, stepwise approach to diagnose these rarer causes of hypokalemia.

Thyroid Type Papillary Carcinoma Arising in Mature Ovarian Teratoma

Ali Hamam, M.D., Daly Al-Hadeethi, M.D., Ammar Al-Obaidi, M.D., Georges Elhomsy, M.D., Kurt Rider, M.D.
KU School of Medicine-Wichita

Introduction

Mature teratoma represents more than 95% of all ovarian teratomas and about 20% of ovarian tumors. Most mature teratomas are benign and unilateral. Malignant transformation occurs in 0.2 % to 2% of mature cystic teratomas and it represents 2.9 % of all malignant ovarian germ cell tumors. The risk factors to develop malignant neoplasm in a mature cystic teratoma include age greater than 45 years, tumor diameter greater than 10 cm, rapid growth, and imaging finding (e.g., low resistance intratumor flow on Doppler).

Case Presentation

The patient was a 57-year-old female accidentally found to have large right pelvic mass measuring 9.1 x 13.9 x 13.4 cm after a workup for kidney stones. She had a past surgical history of tubal ligation and breast lumpectomy. Her father had a history of liver cancer but no family history of breast or ovarian cancer. CT abdomen and pelvis showed a large complex mass of the right adnexa; the lesion measured 14 x 9 cm. A total laparoscopic hysterectomy with bilateral salpingo-oophorectomy was done 4 months prior, with the specimen removed without rupture. Pathology showed papillary thyroid carcinoma arising in mature teratoma with strong uniform staining for TIF-1 and lack staining for thyroglobulin. The endocrinology service was consulted for the follow-up and treatment plan for this rare type of cancer. Lab results showed normal TSH and Free T4 and thyroid ultrasound showed the presence of multinodular goiter with the largest nodule on the right measures up to 1.3 cm and an ill-defined sub centimetric nodule in the right lobe. FNA biopsy of the right thyroid nodule showed benign hyperplastic/adenomatoid nodule, so serial neck ultrasound needed to check for any new changes that could alter the treatment plan.

Conclusions

Papillary carcinoma is the most common type of thyroid carcinoma to occur in mature teratoma and it is diagnosed based on the histopathological criteria and guidelines for primary thyroid gland disease. Malignant metastasis occurs in about 5% to 23% of cases, mostly intra-abdomen, but blood-borne metastasis can occur. The treatment of thyroid carcinoma derived from ovarian mature teratoma remains controversial as it is still rare in occurrence. In previous reports, about 55% of cases will have unilateral salpingo-oophorectomy; total hysterectomy with bilateral salpingo-oophorectomy done in the remainder. The decision for treatment plan will be affected by the peritoneal dissemination, patient age, and desire to preserve fertility. Strict follow-up needed in this rare type of cancer as there is little evidence on its prognosis and no data on recurrence.

Primary Adrenal Insufficiency Secondary to Sarcoidosis

Corey Homan, M.D., Bakul Sangani, M.D.
KU School of Medicine-Kansas City

Introduction

Primary Adrenal Insufficiency secondary to Sarcoidosis is a rare complication. This case report details the presentation and diagnosis in a 50-year-old male with known history of sarcoidosis.

Case Presentation

A 50-year-old male with history of sarcoidosis and hypertension and family history of gastric cancer was admitted to the hospital for nausea, vomiting, and hypotension for three days. He also reported that over the last one year he had an unintentional weight loss of over 50 pounds. Patient had undergone no previous work-up since 2015, which was when he underwent lung biopsy diagnosing sarcoidosis, and was not on long term therapy for this condition; however, the patient reported he had intermittently received steroid bursts for sarcoid flares with last course being in January, 2019. During his hospitalization, patient underwent work-up for malignancy with CT Chest significant for innumerable pulmonary nodules with some associated with volume loss consistent with sarcoid and CT Abdomen significant for enlargement of both adrenal glands. A morning cortisol level was obtained, which was 0.8, and STIM test positive for adrenal insufficiency with elevated ACTH 664. Endocrinology was consulted and further work-up was obtained including Adrenal antibody, which was negative, and the work-up was consistent with Sarcoidosis Induced Adrenal Insufficiency.

The patient was started on stress dose steroids during his admission and his nausea, vomiting, and hypotension all improved. His sodium, which was 126 on admission, also improved to 133 during his hospitalization. Prior to discharge, the patient was transitioned to 20/10 mg of hydrocortisone and started on methotrexate for sarcoidosis per Pulmonary recommendations. On follow-up as outpatient, patient reported continued resolution of symptoms and weight gain of 30+ pounds.

Conclusions

Sarcoidosis most frequently involves the lungs but is actually a multisystem granulomatous disorder that is characterized by noncaseating granulomas in the involved organs. The most common organs involved are the lungs, skin, lymph nodes, eye, and heart; however, it has also been documented to rarely involve the adrenal gland. While adrenal insufficiency in sarcoidosis is typically autoimmune in nature, previous case studies have demonstrated it can also be due to sarcoid infiltration of the organ.

Intestinal Angiosarcoma: A Rare Cause of Upper Gastrointestinal Bleeding

Anthony Jackson, M.D., Luke Frankl, M.D., Mitesh Patel, M.D.
KU School of Medicine-Kansas City

Introduction

Angiosarcoma is an aggressive sub-type of soft-tissue sarcomas arising from endothelial cells of vascular or lymphatic origin. They account for about 2% of soft tissue sarcomas and can arise from any soft-tissue structure or viscera. Angiosarcomas mostly spread hematogenously with the lungs being the primary site of metastasis. Other common sites of metastasis include liver, bone, soft-tissue, and lymph nodes. We report a rare metastasis of angiosarcoma to the upper GI tract.

Case Presentation

A 55-year-old female presented with a history of vertebral angiosarcoma and three months of anemia requiring multiple blood transfusions. She had dyspnea on exertion, lightheadedness, and intermittent melenic stools. Vital signs were within normal limits and exam was unremarkable. Upper endoscopy three months earlier showed multiple 2 mm mucosal papules in stomach and duodenal bulb. Gastric biopsies at that time were consistent with hyperplastic polyps with hemorrhage in the lamina propria. Repeat upper endoscopy revealed erythematous gastric mucosa and innumerable erythematous lesions, some with central erosions, from the duodenal bulb to the jejunum. Biopsies were taken and revealed duodenal angiosarcoma. Due to continued blood loss, she underwent small bowel enteroscopy and video capsule endoscopy which revealed non-bleeding nodular hypervascular lesions extending from the duodenum to the jejunum. In an attempt to control bleeding, Octreotide drip and aminocaproic acid were started. She received 14 units of pRBC over the course of 26 hospital days. She died 48 days after the initial diagnostic EGD.

Conclusions

Intestinal angiosarcoma is exceedingly rare. A thorough review of the available literature in 2017 revealed only 66 case reports. Sixty-four of the cases were adequately staged with 49 representing primary intestinal angiosarcoma while the remaining 15 were metastatic disease. Presenting symptoms included 42 with GI bleeding, 31 with abdominal pain, 21 with distension, 20 with anemia, and 15 with nausea and vomiting. The median survival time for all cases was 30 weeks with a range from 1 to 208 weeks. The median survival time for patients with GI bleeding was 8.5 weeks vs 50.4 weeks in patients without GI bleeding.

There is an association between radiation exposure and intestinal angiosarcoma. Between 25-36% of cases of intestinal angiosarcoma were associated with prior radiation therapy. Interestingly, angiosarcomas associated with radiation were significantly less likely to be associated with gastrointestinal bleeding.

The rarity of the condition makes it exceedingly difficult to diagnose. The increase in survival seen in patients presenting prior to developing overt gastrointestinal bleeding proves the value of diagnosing this condition at the earliest stage possible. This case especially highlights the importance of taking biopsies of multiple lesions in order to accurately diagnose this condition. A high index of suspicion should be maintained for patients presenting with anemia and previous radiation or known angiosarcoma.

An Unusual Case of an Elderly Male Newly Diagnosed with Collagenous Colitis and Celiac Disease

Luke Johnson, M.D., Chelsea Wuthnow, M.D., William Salyers, Jr., M.D.,
Paul A. Johnson, M.D.
KU School of Medicine-Wichita

Introduction

Celiac disease is an immune mediated disease of the small intestine triggered by the ingestion of gluten, often leading to chronic diarrhea. The incidence and prevalence of celiac disease have increased nearly 5-fold in the past 40 years, with the highest prevalence seen in children and females. Microscopic colitis (MC) is another possible cause of chronic diarrhea that is also seen more often in females. MC is comprised of lymphocytic or collagenous colitis and is described as a chronic inflammatory disease of the colon. Although both diseases have an affinity for females, they should both still be considered when evaluating an elderly male that presents with chronic diarrhea. We present a unique case of a new concurrent diagnosis of celiac disease and collagenous colitis in an elderly male.

Case Presentation

An 88-year-old male presented to his primary care physician for evaluation of multiple daily loose stools of 5 month's duration. At that time, he was advised to try loperamide, a high fiber diet, and to return to the clinic if there was no improvement. After several weeks with no resolution of symptoms, laboratory work-up as well as tests for Clostridioides difficile and ova and parasites were negative. He was referred to a gastroenterologist who recommended endoscopic evaluation. Colonoscopy showed normal appearance of colonic mucosa, however, random biopsies were taken throughout the colon. These biopsies revealed a thickened subepithelial collagen band, favoring microscopic colitis with granulomatous inflammation, consistent with collagenous colitis. As part of the work-up for chronic diarrhea, a celiac panel was obtained. Anti-transglutaminase IgA was positive. The patient underwent EGD with biopsies that confirmed duodenal villous blunting consistent with celiac disease. He was advised to follow a gluten-free diet and was started on bismuth subsalicylate. The patient's diarrhea subsequently resolved.

Conclusions

Chronic diarrhea can be defined as increased frequency of defecation or increased stool weight for more than four weeks. Up to 5% of patients who present with chronic diarrhea are diagnosed with celiac disease and up to 15% of patients who undergo colonoscopy for evaluation of chronic diarrhea are diagnosed with MC. Both diseases show a female predominance with a ratio of 1.5:1 found in celiac disease and 9:1 in MC. They also have an association, as the presence of celiac disease carries a 70-fold increased risk of MC. This association is not well understood but both diseases can have similar clinical as well as histologic presentations. Concurrent celiac disease and MC are associated with advanced age, more severe villous atrophy, and often requires corticosteroid therapy.

Short Telomere Related Pulmonary Fibrosis

Joe Johnstone, M.D., Andrea Covey, M.D.
KU School of Medicine-Kansas City

Introduction

Dyskeratosis Congenita is a rare genetic disorder caused by premature telomere shortening. The classical presentation is abnormal pigmentation, nail dystrophy, and oral leukoplakia, in the setting of bone marrow failure. It is an increasingly recognized etiology of pulmonary fibrosis previously thought to be idiopathic. This report describes a novel mutation in Telomerase Reverse Transcriptase (TERT) leading to pancytopenia and pulmonary fibrosis.

Case Presentation

A 44-year-old man with a 30 pack-year smoking history presented to pulmonary clinic with a three-year history of progressive dyspnea on exertion, dry cough, and macrocytic pancytopenia. His pancytopenia evaluation included bone marrow biopsies, flow cytometry, micronutrient and heavy metal concentrations, viral serologies, and serum protein electrophoresis without establishing an etiology. His symptoms began insidiously and progressed to dyspnea with climbing one flight stairs. His cough was dry, non-paroxysmal, and exacerbated by exertion. Exam was notable for a grey-haired mildly obese nearly edentulous man with bibasilar crackles, digital clubbing, and hallux nails. High Resolution CT Chest displayed peripheral subpleural reticular changes, scattered ground glass opacities, and apical honeycombing. Pulmonary function testing demonstrated severe restrictive lung disease.

Genetic sequencing discovered a novel TERT variant with a premature stop codon. Telomere length measurement was below the first percentile, diagnostic of short telomere syndrome. He began danazol, followed by pirfenidone. His course was complicated by avascular necrosis leading to a hip fracture. His pulmonary function progressively declined requiring supplemental oxygen with ambulation. His recovery from his hip fracture was complicated by a global pandemic and he was enrolled in home pulmonary rehab through a grant funded by the VHA. He is undergoing evaluation for single donor lung and bone marrow transplant.

Conclusions

Dyskaratosis Congenita should be considered as a case of pulmonary fibrosis in patients with early graying or bone marrow failure. Telomerase Reverse Transcriptase is the central protein in the telomerase complex. Eleven genes across the process of telomere replication and maintenance have been linked to pathological shortening. TERT mutations are highly associated with aggressive and rapidly progressive pulmonary fibrosis, estimated to cause 1-23% of sporadic and 15-37% of familial cases. Heritance can be autosomal dominant or recessive, and anticipation has been described. The androgen, danazol, can lengthen telomeres; however, progressive pancytopenia eventually requires hematopoietic stem cell transplantation. Although this is well described in the literature, treatment of pulmonary failure is considerably more complicated. Patients with TERT-pulmonary fibrosis undergo supportive care and typically receive the antifibrotics, nintedanib or pirfenidone. Disease progression is typical and lung transplantation remains the only definitive treatment. Dyskeratosis Congenita poses unique challenges in treatment with suppressive therapy after transplantation as well. Single donor dual bone marrow-lung transplant immensely complicates this process yet offers hope in an otherwise devastating and fatal disease.

An Unusual Case of Urothelial Cell Carcinoma with Metastasis to the Pancreas

Katia El Jurdi, M.D., Ali Taleb, M.D., Khalil Choucair, M.D., William J. Salyers, M.D.
KU School of Medicine-Wichita

Introduction

Metastasis to the pancreas is far less common than primary pancreatic tumors with bladder cancer metastasis involving the pancreas. Here we report a case of urothelial cell carcinoma (UC) with metastasis to the pancreas, diagnosed via upper endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA).

Case Presentation

A 59-year-old man with metastatic UC was referred to the gastroenterology service for symptoms of biliary obstruction. Metastatic sites included the spine, sternum, prostate, adrenal glands, and retroperitoneum. He was previously treated with neo-adjuvant chemotherapy, Trans-Urethral Resection of Bladder Tumor (TURBT), open radical cystoprostatectomy, dual chimney ileal neo-bladder insertion, radiotherapy, and immunotherapy. Patient had subsequently developed lymph node metastasis in the porta hepatis region with resultant extrinsic compression on the common bile duct requiring biliary stent placement. Over the next few months, the patient underwent CT chest and abdomen for cancer re-staging. Imaging revealed a suspicious appearance of the left lateral aspect of the bladder and a low-density lesion in the pancreas concerning for pancreatic malignancy versus pseudocyst or low-density lymph nodes. Due to recurrent abdominal complaints, MRI abdomen was ordered and revealed a hypo-enhancing mass in the pancreatic tail, with possible involvement of the splenic artery and vein. Patient subsequently underwent an EUS-FNA that demonstrated a 50.1 mm x 42.2 mm irregular, hypoechoic mass with poorly defined borders in the pancreatic tail. Endosonography was suggestive of invasion into the splenic artery and celiac trunk, and celiac plexus block was performed for pain control. The remainder of the pancreas was unremarkable. Two peripancreatic lymph nodes were seen which appeared malignant, the largest measuring 13.6 mm x 8.3 mm. Biopsies from pancreatic tail mass showed malignant cells with a papillary, glandular pattern. Immunohistochemical (IHC) staining panel showed no mutation of KRAS G12D or G13D. Immunostaining was positive for CK7, CK20, GATA-3, P63, and negative for CK5/6, NKX3.1. No microsatellite instability was found. Diagnosis was consistent with high-grade carcinoma favoring bladder cancer origin. Results were communicated with the oncologist for initiation of timely and appropriately targeted treatment.

Conclusions

This is a rare case of metastatic UC to the pancreas. Although rarely reported, it is vital to consider pancreatic metastasis as a plausible diagnosis in a patient with a pancreatic mass and a primary bladder tumor. EUS-FNA with IHC staining of biopsy specimen, is pivotal in distinguishing secondary neoplasms that may otherwise present as primary solitary pancreatic lesions.

Simultaneously Diagnosed Gastric Adenocarcinoma and Pernicious Anemia: A Classic Association

Syed Kamran, D.O., Mattias K. Dilling, Nathaniel A. Parker, D.O., Joel Alderson, D.O., Nathan D. Tofteland, M.D., Quoc V. Truong, M.D.
KU School of Medicine-Wichita

Introduction

Individuals with pernicious anemia (PA) have increased risk for gastric cancer. The pathogenesis of gastric cancer arising from PA is due to chronic inflammation with extensive atrophy of the gastric mucosa, leading to increased risk of progression to gastric neoplastic lesions. This report presents the uncommon, but classic association, of PA with primary gastric cancer.

Case Presentation

A 61-year-old Hispanic female presented to the emergency department with epigastric pain. Her chief complaints were associated with heartburn, decreased appetite, and weakness that had progressively worsened for three weeks, along with a 60-pound unintentional weight loss over the past six months. Her family history was negative for any gastrointestinal diseases or malignancies. Vital signs and detailed physical examination were non-revealing. Laboratory analysis was notable for a significant anemia, as well as a low vitamin B12 and iron deficiency. She was urgently managed with a restrictive transfusion strategy by one unit of leuko-reduced packed red blood cells, which improved her symptoms and hemoglobin. Abdominopelvic imaging was obtained by computerized tomography (CT) scans with contrast, which were primarily equivocal. Subsequently, she underwent esophagogastroduodenoscopy for further evaluation. Gastric findings included an extensive, deep ulcer noted to involve the entirety of the incisura and pre-pyloric area, as well as extended along the lesser curvature. Due to the tumor's size, gross appearance, ulcerations, and bleeding erosions, a malignant process was suspected.

Based on the suspicions for an underlying malignant gastric neoplasm and the presence of a vitamin B12 deficiency, serologic testing for PA was obtained. Laboratory testing was positive for anti-IF confirming the diagnosis of PA. Microscopically, antral mucosa demonstrated mild chronic gastritis. Histopathology revealed a diffuse-type, invasive poorly differentiated adenocarcinoma. She underwent a positron emission tomography scan for staging. No obvious metastatic disease was present. She was started on intravenous vitamin B12 and iron replacement and slowly improved. She was dismissed from the hospital and established care with a local oncologist for chemotherapy using the FLOT regimen (oxaliplatin, leucovorin, docetaxel, fluorouracil). However, the patient did not tolerate chemotherapy well and only completed cycle. She stopped all therapies and was transitioned to hospice care. Two weeks after stopping chemotherapy, the patient expired.

Conclusions

Primary gastric cancer remains one of the most prevalent malignancies worldwide and is associated with poor outcomes. This is likely due to patients remaining asymptomatic until late-stage progression. Early detection of gastric cancer is paramount. PA is uncommonly seen in clinical practice but remains a classic risk factor for the development of primary gastric cancer. Identifying pertinent physical exam features and pairing them with lab findings of a vitamin B12 deficiency can be crucial steps in uncovering the gastric cancer early.

Fahr's Syndrome: A Rare Cause of Status Epilepticus

Monica Khurana, M.D., Mohinder Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

Introduction

Fahr's disease refers to idiopathic bilateral and symmetric intracranial calcification of the basal ganglia and cerebellar nucleus. It is likely sporadic or inherited in an autosomal dominant pattern. In contrast to the primary form, Fahr's syndrome occurs secondary to various infectious, endocrine, or metabolic diseases. The syndrome typically manifests as seizures, parkinsonism, vertigo, dysarthria, cognitive impairment, and behavioral changes. We present a case of Fahr's syndrome associated with secondary hyperparathyroidism presenting with status epilepticus.

Case Presentation

A 38-year-old female with type 1 diabetes mellitus, stage IV chronic kidney disease, and hypothyroidism presented to the Emergency Department (ED) for status epilepticus. The physical exam was benign except a left-sided tongue laceration likely from the seizure episode. The patient required intubation for airway protection in the ED and later transferred to the intensive care unit for medical management. Computer tomography of the brain in the ED looking for possible bleed revealed dense calcifications in the basal ganglia and dentate nuclei. Further investigation of seizure etiology revealed the patient to be in a hyperosmolar hyperglycemic state. The patient was managed on an insulin drip, lorazepam, and levetiracetam, which controlled the seizures. Further workup of the basal ganglia calcification with magnetic resonance imaging demonstrated marked T1 hyperintensity within the caudate, putamen, and dentate nuclei suggestive of Fahr's disease/syndrome. The patient's seizures were resolved and she was discharged home on vitamin D supplementation. Further workup on an outpatient basis revealed secondary hyperparathyroidism (due to kidney disease and vitamin D deficiency), which is the likely cause of her calcifications in the basal ganglia confirming Fahr's syndrome. The patient had been admitted multiple times since then for seizures and derangements in glucose with concern for neurocognitive decline as a barrier to adherence to her anti-diabetic medication regimen.

Conclusions

Management and treatment of Fahr's syndrome focus on symptomatic relief and eradication of causative factors. Although rare, providers should maintain a high index of suspicion for Fahr's syndrome in patients with a seizure disorder or progressive neuropsychiatric disturbances. It is essential to perform neuroimaging and laboratory investigations for the recognition of possible contributing factors. Timely recognition allows for appropriate treatment that can slow the progression or prevent clinical manifestations.

An Uncommon Cause of Diabetic Ketoacidosis

Sean Lacy, D.O., Sushant Govindan M.D., M.Sc.
KU School of Medicine-Kansas City

Introduction

Ketosis prone diabetes (KPD) is an uncommon subset of diabetes syndromes that presents as intermittent diabetic ketoacidosis (DKA) requiring temporary insulin treatment. It is characterized by severe beta cell dysfunction and variable clinical course.

Case Presentation

A 40-year-old African American male with a past medical history of type 2 diabetes mellitus (DM) and hypertension presented to the emergency department after routine labs showed hyperglycemia and metabolic acidosis. His DM was previously treated with metformin, but the patient had run out of metformin months prior to presentation. He reported increased thirst, increased urination, and weight loss. He reported a viral illness 3 months ago, but no current infectious symptoms or ischemic symptoms. His vital signs were relatively unremarkable. His initial metabolic profile showed hypokalemia (3.1), CO2 14, Cr 1.84, Glucose 504, Beta hydroxybutyrate 9.92, and A1c > 16.9. He was started on the Yale Protocol for DKA. His anion gap normalized the following day with an insulin drip requirement of 7.5 units/hour. Further work up showed GAD and islet cell antibodies were negative. He had a fasting C peptide of 0.26. Endocrinology was consulted and he was transitioned to insulin glargine 60 units daily and insulin aspart 20 units with meals. He was discharged home in stable condition with primary care and endocrinology follow-up. At two-week follow-up, his insulin aspart requirement had decreased to 15 units, with plans to continue further titration and eventual discontinuation.

Conclusions

In this case, we describe a rare subset of diabetes: ketosis-prone diabetes. This condition presents with intermittent beta cell failure resulting in DKA. In KPD, patients in DKA will typically require intermittent insulin therapy until beta cell function can be restored. It is important to distinguish this from late onset type 1 diabetes mellitus as this will require lifelong insulin. In KPD, 70% of patients will recover beta cell function within 10 weeks of presentation. This subset of diabetes was first categorized in Nigeria in the 1960s and is more common in people of African or Asian descent.

KPD has been further categorized into four subgroups based on the presence of autoantibodies and presence or absence of B cell function, known as the AB classification. In our patient, he lacked autoantibodies and beta cell function as evident by fasting C-peptide < 1.0. Recognition of this etiology of DKA is important because KPD will likely have resolution of insulin requirements while adult onset type 1 diabetes will require lifelong insulin therapy. Importantly, inpatient teams must plan for insulin titration and eventual discontinuation during transitions of care.

Multiple Myeloma Associated with Dermatomyositis

Vijay Malladi, M.D., Donna Sweet, M.D.
KU School of Medicine-Wichita

Introduction

Multiple myeloma (MM) is characterized by the neoplastic proliferation of plasma cell clones that produce monoclonal immunoglobulin. Dermatomyositis (DM), an autoimmune disease, carries a higher risk of cancer than that of the general population. The concurrence of DM and MM is rare and has not been fully investigated. The mechanism underlying the association between idiopathic inflammatory myopathies and malignancies remains unclear. DM with calcinosis cutis as seen in our patient was successfully treated with IVIG.

Case Presentation

A 62-year-old white male presented with a history of hepatitis C and multiple myeloma. Skin biopsy showed evidence of calcinosis. If there is an underlying autoimmune etiology, then the consideration would be dermatomyositis versus less likely systemic sclerosis (no Raynaud's phenomenon and no skin thickening on the exam). Labs revealed ANCA negative, APS negative, and ANA/RF negative. A slight elevation in cyclic citrullinated peptide at 22 with a normal value of less than 19 was not significant. Myositis panel was negative. Muscle markers were normal. Colchicine prescribed for 6 months without any improvement of the rash. Minocycline was taken for 3 months without improvement. He takes diltiazem and developed a more diffuse rash and muscle weakness likely secondary to the diltiazem. The more diffuse erythematous rash has resolved but he continues to have severe ulcers. He has complications of severe calcinosis cutis.

The patient was seen at Mayo Clinic. A muscle biopsy of the right deltoid showed necrosis of the muscle tissues with regenerating fibers and collections of mononuclear cells at perimysial sites. There was perifascicular atrophy. Chemical studies showed focal capillary depletion and scattered capillaries displayed membrane attack complex deposition. This was consistent with complement-mediated micro vasculopathy. He was diagnosed with DM with muscle involvement. He was continued on IVIG replacement for DM with calcinosis cutis. For multiple myeloma, he was initially on bortezomib, lenalidomide, and dexamethasone then transitioned to bortezomib maintenance.

Conclusions

Dermatomyositis is an autoimmune disease associated with Multiple Myeloma. Inflammation in the setting of autoimmunity may serve as a trigger for MM. A common genetic susceptibility for developing both DM and MM might also exist. There is a need for further exploring the link between MM and DM.

Brugada Syndrome: A Rare Case of Life-Threatening Dysrhythmias in a Young Lady

Job Mogire, M.D., Mohinder Vindhyal, M.D., M.Ed., Brent Duran, D.O., May Mohamad, M.D.
KU School of Medicine-Wichita

Introduction

Brugada syndrome is a hereditary cause of arrhythmogenic disease. Its diagnosis requires a high index of suspicion, thorough history, and keen electrocardiographic evaluation. Young people presenting with syncope plus a family history of sudden death without structural heart disease should trigger an evaluation for rare causes, including Brugada syndrome.

Clinical Presentation

A 26-year-old Caucasian female with 5-year-history of recurrent syncope, and family history of sudden deaths without structural heart disease, presented after an unprovoked syncopal episode. Symptoms occurred while she was seated, playing with her two children. She experienced sudden dizziness and lost consciousness, with no recollection of events that followed. Her husband witnessed jerking of upper limbs for 5 seconds before the loss of consciousness; no other jerking movements were noted. She did not bite her tongue, froth/drool at the mouth, or soil her clothes. When she came around 30 seconds later, she was not post-ictal. Enroute to the hospital, paramedics recorded intermittent episodes of wide complex tachycardia interspersed with significant bradycardia. Telemetry and intravenous Amiodarone drip were initiated on arrival. Dizzy spells accompanied by tachycardia alternating with bradycardia (rates as low as 20) persisted. CT chest with contrast ruled out PE despite elevated D-dimers, EEG ruled out seizures, Telemetry showed intermittent Torsade-like tracings, and serial EKGs showed persistently long QTc intervals, right bundle branch block, and persistent ST-elevation. Labs remained unremarkable except for a minimal elevation in troponin. 2D Echo showed an ejection fraction of 30%. Coronary angiography was negative for obstructive lesions. Brugada syndrome type 1 with a high risk for transition to fatal Torsade du pointe was suspected. The patient received a temporary intravenous pacemaker, followed by a permanent pacemaker with an automatic implantable cardioverter-defibrillator (AICD), and discharged on oral Amiodarone for outpatient follow-up. Per prior history, the patient first experienced similar syncopal episodes during pregnancy and delivery, and subsequently, she had at least two episodes a year, but none as severe. Two of her siblings had died of sudden cardiac arrest; genetic workup had not been pursued.

Conclusions

Since the discovery and electrocardiographic characterization of Brugada syndrome, Brugada is often misdiagnosed, underdiagnosed, or undiagnosed. Nevertheless, treatment for Brugada syndrome has undergone significant changes in the last three decades, from the initial enthusiasm of ‘prophylactically’ inserting defibrillators in asymptomatic patients to the current practice of treating with defibrillators only high-risk cases. Though most patients with Brugada syndrome remain asymptomatic most of their life, the minority of patients who develop arrhythmias face a significant risk for cardiac arrest, often as the presenting symptom. As our case shows, Brugada can present with recurrent syncope. Though often challenging, early diagnosis in symptomatic patients can be lifesaving.

Neuromyelitis Optica with Longitudinally Extensive Transverse Myelitis as a First Manifestation of Systemic Lupus Erythematosus

Maha Mohamad, M.D., Rami H Diab, M.D., May A. Mohamad, M.D., Timothy S. Shaver, M.D
KU School of Medicine-Wichita

Introduction

Neuromyelitis optica spectrum disorder (NMOSD) is an autoimmune syndrome characterized by inflammatory lesions of the optic nerve and spinal cord potentially presenting as transverse myelitis (TM) or more specifically, longitudinally extensive transverse myelitis (LETM). Such patients have seropositivity for NMO-IgG/AQP4-Ab which has been reported in association with a variety of autoimmune diseases, most notably systemic lupus erythematosus (SLE) and Sjogren's disease.

Case Presentation

A 31-year-old female presented to the ED with two months of back pain, paresthesia, and lower extremity weakness precipitating a fall. On arrival, CT of the thoracic and lumbar spine revealed mild right L4-L5 and L5-S1 disc bulges without central or foraminal canal stenosis, yet she was found to have new urinary retention draining 1300 cc of urine. A cervical, thoracic, and lumbar spine MRI with contrast showed an extensive longitudinal segment with T2 brightness extending from C2 to T10 suspicious for LETM. Lumbar puncture revealed elevated WBC 148 (H), protein 144 (H), and IgG 13.6 (H), with normal oligoclonal bands. She was started on high-dose methylprednisolone followed by 5 days of IVIG. Her anti-DS DNA Ab returned 80 (H) in association with malar erythema suggestive of SLE with CNS involvement and cyclophosphamide/mesna was started. Finally, an NMO IgG Ab returned 3123.1 (H) suggesting NMOSD, prompting addition of Rituximab. Patient did well on the therapeutic regimen. However, she was lost to follow-up in our clinic.

Conclusions

NMOSD can present as a first manifestation of autoimmune disease. Findings of LETM on MRI should prompt serologic testing for NMO. Findings of positive NMO-IgG/AQP4-Ab should trigger evaluation for NMOSD's most common comorbid autoimmune disorders, namely SLE and Sjogren's disease.

Volunteering Saved My Life! A Case of Anomalous Right Coronary Artery Take Off

Grace Nassim, M.D., MPH, Patrick Ters, M.D.
KU School of Medicine-Wichita

Introduction

Around 80% of coronary artery abnormalities perceived during catheterization are benign incidental findings. However, ectopic coronary origin from the pulmonary artery or opposite aortic sinus, single coronary artery, or large coronary fistulae are serious anomalies resulting in angina pectoris, myocardial infarction, heart failure, arrhythmias, and even sudden cardiac death. We report a case report of a surgically repaired anomalous right coronary artery in a patient presenting with an abnormal EKG strip.

Case Presentation

A 54-year-old white male with no significant past medical history, presented to his primary care physician with an abnormal EKG strip. He worked as an EMT and, during his first aid course, he volunteered for an EKG strip which showed ST depressions. A repeat EKG at his primary office was normal. The patient was sent for an outpatient cardiology work-up. Upon further investigation, the patient stated a positive family history of coronary artery disease in his father. He reported tiredness and exertional dyspnea. A stress test demonstrated a large area of reversible ischemia within the right coronary artery distribution. A coronary angiogram demonstrated anomalous right coronary originating from the left sinus of Valsalva. There was no evidence of any obstructive atherosclerotic coronary artery disease. A cardiac CT scan subsequently was obtained confirming this diagnosis with a large, dominant, anomalous right coronary artery originating from the left coronary cusp and coursing between the aorta and pulmonary artery. The patient was referred for a vein graft bypass to a 1.5 mm Posterior Descending Artery. He tolerated the procedure well and was discharged on aspirin, a statin, and a beta-blocker.

Conclusions

The incidence of anomalous origin of the right coronary artery arising from the left coronary cusp that courses between the great vessels varies between 0.026% and 0.250%. The initial presenting symptom may be sudden cardiac death, because the course of the anomalous right coronary artery between the pulmonary artery and aorta presents a risk for the artery to be compressed during exertion. This is the first known case to reveal another presenting chief complain for an underlying anomalous right coronary artery. It is crucial to document such presentations among physicians for early recognition and treatment.

Suspected Anaphylaxis to Cosyntropin

Sonya Parashar, M.D., Corey Homan, M.D., David Naylor, M.D.
KU School of Medicine-Kansas City

Introduction

Anaphylaxis to cosyntropin is a very rare complication. This case report details the anaphylactic reaction to cosyntropin in a 64-year-old male with history of multiple sclerosis.

Case Presentation

A 64-year-old male with a history of multiple sclerosis (MS) was admitted to the hospital for acute diarrhea and failure to thrive. He reported that over 2 years he had an unintentional weight loss of over 100 pounds. He had previously undergone an extensive evaluation, including malignancy screenings, which did not reveal the cause of his failure to thrive. The patient reported that he had been on and off steroids several times in his life for MS exacerbations. A morning cortisol level was obtained and came back low at 1.9 mcg/dl, which raised concern for secondary adrenal insufficiency. He was scheduled for an adrenocorticotropic (ACTH) stimulation test. About 90 minutes after completing the stimulation test, he started complaining of sudden onset shortness of breath, nausea, and new rash on his chest. His initial vitals were stable except for new onset tachycardia as well hypoxemia. At the bedside, the patient was noticeably uncomfortable with increasing oxygen requirement and wheezing on exam. A new urticarial rash was present on his chest. He also reported nausea and subsequently vomited several times. He did not receive any new medications other than cosyntropin on that day.

The patient received intramuscular epinephrine 0.3 mg, methylprednisolone 125 mg IV, famotidine 20 mg IV, Benadryl 25 mg IV, and Compazine 10 mg IV. Tryptase was ordered about 90 minutes after the event and was within normal limits. The patient reported feeling subjectively better and his vitals remained stable. Due to his persistent tachycardia, an EKG was obtained and showed ST depression in the inferior leads with no troponin elevation. He underwent a stress test over the next 48 hours which was read as low risk. Cosyntropin was listed as an allergy on this patient's chart. He was started on treatment for secondary adrenal insufficiency with prednisone and gained about 20 pounds over the next 6 months.

Conclusions

Anaphylaxis is a clinical diagnosis, which requires sudden onset of mucocutaneous manifestation and either gastrointestinal symptoms, pulmonary symptoms, or hypotension. There is only one previously documented case of anaphylaxis to cosyntropin in 1987 in a patient with MS. ACTH is a short-chained polypeptide, which likely drives its immunogenicity. While this is an extremely rare side effect of cosyntropin, it is a potentially life-threatening side effect and must be treated emergently.

Recurrent Dasatinib-Induced Chylothorax in Chronic Myeloid Leukemia: A Case Report

Jacob Ripp, D.O., Jessica Allen, Abdulraheem Yacoub, M.D.
KU School of Medicine-Kansas City

Introduction

Tyrosine kinase inhibitors (TKIs) are a breakthrough medication class in the era of targeted therapy for chronic myeloid leukemia (CML), acute lymphoblastic leukemia (ALL), and more recently GI stromal cell tumors. Dasatinib is a second generation TKI approved in 2006 for Philadelphia chromosome-positive (Ph+) CML and AML. In all TKIs, fluid retention is a known and relatively common side effect. However, there are currently only eight case reports that describe dasatinib-induced chylothorax. This case report discusses another such case and discusses possible mechanisms behind dasatinib-induced chylothorax as well as management strategies.

Case Presentation

The patient is a 57-year-old male. He has a past medical history of chronic myeloid leukemia with major molecular response (MMR) on dasatinib 140mg once daily dosing and presented to the hospital with a 10-day history of progressive body aches, low-grade fevers, cough, and weight gain. On exam, he was found to have diminished breath sounds in the bilateral bases of his lungs. A chest x-ray and CT imaging revealed moderate right and small left pleural effusions. Subsequent pleural fluid studies from a diagnostic thoracentesis revealed 3000 white blood cells with 76% lymphocytes, glucose 76, protein 4.1, LDH 100, triglycerides 1,163, bilirubin 1.4, lipase 43, cholesterol 71, amylase 36, albumin 2.9, pH 7.51. These findings are consistent with a chylothorax. His dasatinib was subsequently discontinued. One month later, he again required another therapeutic thoracentesis that was also consistent with a chylothorax. Given the absence of other, more common causes of chylothorax, it was determined that dasatinib was the ultimate culprit. He was then started on a low fat diet and cholestyramine. He was a candidate for treatment free remission and has been off of dasatinib with controlled disease for three months.

Conclusions

While rare, this case represents a possible complication of tyrosine kinase inhibitor therapy, including dasatinib. The vast majority of chylothoraces are caused by trauma or an underlying malignancy, namely, lymphoma. The mechanism involves any disruption of the lymphatic vasculature, but typically involves obstruction (malignancy) or tear (surgery or trauma). Medication-induced chylothoraces are extremely rare, and the precise mechanism of dasatinib-induced chylothorax is currently unknown. However, there may be evidence of dasatinib-induced inhibition of normal T-lymphocytes that support lymphatic vasculature. Other suggested mechanisms include further kinase inhibition by dasatinib, including PDGFR-β, which supports lymphangiogenesis and regulates interstitial fluid pressure. Successful management of dasatinib-induced chylothorax typically includes thoracentesis, diuresis, corticosteroid therapy, and/or dose reduction or discontinuation of dasatinib depending on the severity. Dose reductions or switching to an alternative TKI should be done with careful molecular monitoring to maintain MMR. Supportive treatment, including a low-fat diet and/or cholestyramine to prevent worsening of chylothoraces, are important if they are recurrent.

COVID-19 Related Acute Pancreatitis: A New Etiology?

Corey Braet, D.O., Ryan Ford, M.D., Paul Ndunda, M.D.
KU School of Medicine-Wichita

Introduction

Our understanding of the virus that caused the global COVID-19 pandemic, severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), is constantly evolving as is the revelation of the myriad of other symptoms and conditions that result from infection. Many infections may be asymptomatic with the most common symptoms being cough, fever, myalgia, headache, and dyspnea; however, it also has a major impact on multiple organ systems throughout the body. An acute pancreatitis diagnosis requires two of three criteria: typical abdominal pain, enzyme elevation three times or greater than the upper limit of normal, and characteristic findings on imaging. The diagnosis of pancreatitis can easily be made but determining the etiology may be more complex as it includes: mechanical, toxic, metabolic, drugs, trauma/procedures, and infections. Only a few case reports have associated SARS-CoV-2 infection and acute pancreatitis and these cases all involved obese patients less than 60 years of age. Herein, we report a patient case of COVID-19 who presented with acute pancreatitis without other risk factors or noted causes.

Case Presentation

A 78-year-old African American female with a normal BMI and history of hypertension, psoriatic arthritis, and GERD presented with nausea, vomiting, fever, and sharp abdominal pain that radiated to the back. The patient's lipase was 398 U/L (normal 8-78 U/L) and she was diagnosed with acute pancreatitis, with no previous history of the condition. Due to presence of gastrointestinal symptoms and fever, she was tested for SARS-CoV-2 by PCR and was positive. No other etiologies for pancreatitis were discovered during the admission. She underwent computed tomography of the abdomen and pelvis with contrast which was negative for any acute pathology with normal gallbladder, biliary tract, and pancreas. A complete abdominal ultrasound was also without noted pathology. Additionally, ethanol, urine drug screen, serum calcium, and serum triglycerides were negative or normal. She was not taking any new medications or on any medications with great potential for causing acute pancreatitis. Both the patient's COVID-19 and pancreatitis were mild and quickly improved with supportive treatment, the patient discharged home after 5 nights.

Conclusions

Pancreatitis has many known etiologies, including different viral infections like mumps, hepatitis B, cytomegalovirus, HSV, and HIV. Additionally, it is known that the pathogenesis of COVID-19 is partly mediated by binding to the angiotensin-converting enzyme-2 receptor on host cells. These receptors are highly expressed in the pancreas and the pancreatic islets, with reports of the infection inducing acute diabetes. Although direct evidence of viral pancreatitis from SARS-CoV-2 is difficult to demonstrate, the lack of other causes and the association noted above points to this very real possibility.

Recurrent Pseudomonas Lung Infections in the Setting of Bronchiolitis Obliterans Following Allogeneic Stem-Cell Transplant

John Henry Carson, M.D., MBA
KU School of Medicine-Wichita

Introduction

Graft-versus-host disease (GVHD) is a known complication following lung transplant. Lung manifestations of GVHD destroy small airways leading to bronchiolitis obliterans (BO), which historically, has had poor prognosis. Reported sequelae of BO includes recurrent lung infection. Pseudomonas has been recorded as a notable and worrisome offender for lung infection in patients developing BO after lung transplant. Less commonly, hematopoietic cell transplant has been associated with BO. This case report discusses an atypical presentation of recurrent Pseudomonas lung infection in a patient following allogeneic stem cell transplant (Allo-SCT).

Case Presentation

A 69-year-old woman presented to the Emergency Department with altered mental status and respiratory failure. Her past medical history was relevant for bronchiolitis obliterans (BO) related to Allo-SCT and GVHD. From 2017-2020, the patient was subjected to 12 hospital admissions with the driver of her decompensation being characterized by recurrent lung infections, particularly by Pseudomonas. Her treatment for chronic GVHD and BO included chronic immunosuppression (prednisone) and gene-expression modulators (Ruxolitinib, Ibrutinib), both of which potentiated her risk for infections. She was admitted to intensive care, where treatment with broad-spectrum antibiotic coverage and non-invasive positive-pressure ventilation were initiated. She responded well to intervention and was transferred to the general medical floor on hospital day two where she continued to convalesce until her discharge on hospital day five.

Conclusions

While some cases have reported concomitant Pseudomonas colonization and infection in the setting of BO after lung transplant, this case is relatively more unique in that it highlights an uncommon presentation of Pseudomonas infection, one that complicates BO after stem cell transplant rather than lung transplant. Infection is a leading cause of death in patients following Allo-SCT and may exacerbate alveolar damage in patients with BO. Pseudomonas is of particular concern as it is notoriously difficult to eradicate after colonization. In general, a high index of suspicion regarding infection is likely appropriate when surveilling patients following Allo-SCT and may help to facilitate early recognition and treatment. Interventions such as bronchoalveolar lavage (BAL) may help in early recognition of pathogen, and chronic antibiotic prophylaxis may be warranted in those patients who demonstrate recurring infection.

Sickle Cell Disease and COVID 19

Karen Gichohi, M.D., Nicholas Ojile, M.D., Thomas Moore, M.D., Victor Bedros, MS-4
KU School of Medicine-Wichita

Introduction

Respiratory viruses cause more severe complications in people with sickle cell disease (SCD). Patients with sickle cell had poor outcomes during the H1N1 pandemic. They do not fare well in flu season either. Since SARS-COV-2 is a respiratory virus that causes severe pneumonia, infection could lead to more adverse outcome in this patient population due to underlying chronic inflammation, compromised immune system, increased risk of thrombosis, and a shared propensity for both diseases to cause acute lung injury. COVID 19 affects more African Americans which may impose a higher risk in patients with SCD. We present a COVID 19 positive patient with history of SCD who was treated successfully with convalescent plasma with basic respiratory support.

Case Presentation

A 53-year-old patient with history of SCD, chronic Afib, and hypothyroidism presented to the ED with watery stool, fever, and fatigue. He was in septic shock with AKI and positive for COVID 19. He was admitted to the ICU in sickle cell crisis manifesting as acute severe hemolytic anemia. He was not significantly short of breath and required between 2-3l oxygen by NC. Chest x-ray showed bibasilar patchy pulmonary opacity. COVID 19 inflammatory markers were markedly elevated. On hospital day 2, he received convalescent plasma, plasma exchange transfusion, and dexamethasone, but not remdesvir. He was on pressure support, IV fluids, and broad-spectrum antibiotics for septic shock. Anticoagulation was initiated on hospital day 3 for increased thrombotic risk from both SCD and COVID 19. ICU stay was protracted by Afib with rapid ventricular response, septic shock, and NSTEMI. He was transferred from the ICU on hospital day 6 on room air. His presenting complaints resolved. He was discharged home on hospital day 8 on apixaban and dexamethasone.

Conclusions

Though our patient had a remarkable inpatient course, the respiratory component was mild and his predominant symptoms were gastrointestinal. His chest x-ray findings were mild compared to the findings in majority of COVID 19 pneumonia patients. Abnormally high plasma values of proinflammatory cytokines such as IL-1, IL-6, and TNF-alpha have been reported in SCD patients at steady state. SARS-CoV S protein induces up-regulation of Il-1, IL-6, and TNF-alpha. This similar pathophysiology could explain the low impact of the cytokine storm, reported as a complicating mechanism of COVID 19, in SCD patients, whose bodies are already accustomed to a chronic inflammatory state. Rapid diagnosis and intervention including plasma exchange and blood transfusion can result in excellent recovery. The background chronic inflammatory, hemolytic, and anemic state in SCD might have a favorable influence in protecting this patient population from fatal COVID 19 infection. More research and case reports across the globe are required.

A Rare Case of Human Monocytic Ehrlichiosis with Neurologic Presentation

Sophia Hitchcock, M.D., Luke Frankl, M.D., Katie Lester, M.D.
KU School of Medicine-Kansas City

Introduction

Human Monocytic Ehrlichiosis has a clinical presentation and laboratory profile that mimics a number of diseases. The decision to initiate treatment should not be delayed by awaiting confirmatory testing.

Case Presentation

A 71-year-old male presented with one month of falls and acute confusion. Review of systems was otherwise unremarkable. Physical exam revealed disorientation to self, place, and time. Neurologic exam significant for dysarthria, dysmetria, and ataxia. Throughout admission, laboratory workup was significant for Hgb 9.8, WBC 1.45, Plt 28, ESR 85, CRP 11.9, LDH 205, Haptoglobin 225, ferritin 1021, TIBC 156, B12 1121, AdamsTS13 95% (68-163), negative West Nile IgM, ANA, CMV IgM, and EBV IgM. Peripheral smear showed occasional, scattered schistocytes. CSF analysis revealed 7 NRBC's w/87% lymphocytes, protein 56.5, glucose 101, positive oligoclonal bands, negative enterovirus, HSV, lymphoma panel, gram stain, culture. On imaging, MRI brain was without acute intracranial findings. After failed response to empiric treatment for autoimmune encephalitis, further history was obtained. It was noted that he was a farmer in Southern Missouri and was empirically started on Doxycycline to cover tick-borne illness. His mental status rapidly improved. Lab later confirmed that the patient indeed had Ehrlichiosis Chaffeensis, positive for IgG and IgM.

Conclusions

Human monocytic ehrlichiosis is an infection caused by Ehrlichia Chaffeensis, which is endemic to the Mid-Atlantic, Southeastern, and South-Central United States. The principal vector is the lone-star tick, and white tail deer are the principal animal reserve. The most common symptoms reported are malaise, myalgias, headache, chills, nausea, vomiting, and cough. Also, a macular, maculopapular, or petechial rash can occur. Neurologic symptoms, including mental status change, can be present in up to 20% of cases. Common laboratory findings include leukopenia, thrombocytopenia, anemia, and elevated liver enzymes. In neurologic cases, CSF most commonly shows lymphocytic pleocytosis and elevated protein levels. To confirm infection, the preferred diagnostic test is indirect fluorescent antibody test. Due to these nonspecific signs and symptoms, Ehrlichiosis often mimics other tick-borne or viral illness, community-acquired pneumonia, thrombotic thrombocytopenic purpura, or hematologic malignancy.

The decision to initiate treatment should be made based on a presumptive diagnosis. Relying on definitive testing could delay treatment and put the patient at risk of complications, including death. Standard treatment is a 5-7 day course of Doxycycline. Rapid improvement in symptoms should be seen. The above case was a unique presentation of Ehrlichiosis, as the patient presented with ataxia, altered mental status, and pancytopenia. In conclusion, physicians should rely on clinical presentation, laboratory evidence, and exposure history to initiate treatment of Ehrlichiosis.

Iatrogenic Cushing's Syndrome Due to Inhaled Glucocorticoids

Tyler Johnson, M.D., Lisa Clough, M.D.
KU School of Medicine-Kansas City

Case Presentation

A 60-year-old female with a history of COPD, lung adenocarcinoma, and well-controlled HIV, presented to infectious disease clinic complaining of a 15-pound weight gain and swelling in her face over the last 6 months along with fatigue and increased facial hair. A previous provider thought she may have been suffering from lipodystrophy due to her HIV medications, but no changes were made at that time. At the time of presentation, she had been taking elvitegravir/cobicistat/emtricitabine/tenofovir alafenamide for 3 years for treatment of her HIV and fluticasone/salmeterol for COPD, which had been started approximately 6 months prior to presentation. On exam, she had rounded facies, increase in cervical fat pad, increased abdominal girth, and enlarged breasts. Due to the known interaction between certain HIV medications and steroids, she was evaluated for iatrogenic Cushing's Syndrome. A random cortisol level was 0.6 mcg/dL (reference range 5-20) and a serum ACTH level was < 5 pg/mL (reference range 7.2-63), suggesting a suppressed HPA axis. She was referred to endocrinology and a confirmatory 24-hour urine free cortisol level was < 1 ug/24 hours (reference range 3-50). The patient was transitioned to bictegravir/emtricitbine/tenofovir alafenamide to minimize interactions and was given a short course of hydrocortisone to prevent adrenal crisis along with serial lab monitoring to ensure HPA axis recovery.

Conclusions

Cobicistat, similar to ritonavir, is a CYP3A inhibitor used to boost the effect of antiretrovirals by slowing their metabolism. Fluticasone and many other steroids are also metabolized by CYP3A and when combined with potent CYP3A inhibitors, local low dose steroids can begin to have systemic consequences. The interaction between boosting agents and inhaled corticosteroids is well documented; however, this may go overlooked and not all physicians are aware of this interaction. It is important to recognize these interactions as inhaled, intranasal, and topical steroids are very commonly prescribed and are often thought by many to have very low risk of systemic effects. Not only does this interaction put patients at risk for significant morbidity from systemic effects of glucocorticoids such as weight gain, hyperglycemia, osteoporosis, and immunosuppression, but also for potentially life-threatening adrenal crisis when the medications are abruptly discontinued.

Cutaneous-Visceral Loxoscelism in an Adult with Bullae Dermatosis

Lorna Nguutu, M.D., Thomas Moore, M.D., Mohinder R. Vindhyal, M.D.
KU School of Medicine-Wichita

Introduction

Loxoscelism refers to the clinical presentation that results from a brown recluse spider bite. In the US, these spiders naturally inhabit the Southern and Central regions. The typical presentation to a recluse bite is a localized cutaneous inflammation with eventual necrosis. Rarely in adults, a systemic reaction will occur manifesting as hemolytic anemia, renal failure, rhabdomyolysis, and disseminated intravascular coagulopathy with increased fatality. There are currently no validated venom assays or treatment guidelines. We present a case of an adult female with an atypical presentation of systemic loxoscelism.

Case Presentation

A 50-year-old Hispanic female without prior medical history presented with a worsening rash. The initial onset was three days prior, after she had a spider bite. The site became progressively dark. Over two days, she developed fever and a widespread, pruritic rash across her body. The symptoms did not improve on Diphenhydramine or Naproxen. She denied any other medication usage, known allergies, or other pertinent history.

Upon evaluation, the patient was hemodynamically unstable with hypotension and tachycardia. Labs showed increased inflammatory markers. At the bite site, she had a 4 cm, black patch with marginal hypopigmentation. She also had a widespread, diffusely tender erythematous and edematous rash across her arms, legs, abdomen, and back along with tense bullae. Her chest had pustular skin eruptions. There was no facial or mucosal involvement. Based on the hospital sepsis protocol, she was given intravenous fluids and started on Vancomycin, Ceftriaxone, and Clindamycin. Steroids were started for the inflammation.

The following day, the steroids were stopped and Dapsone was added based on some studies that showed benefit for dermonecrotic loxoscelism. On day four, the hospital course was complicated by increased bullae, mild transaminitis, respiratory failure, and hemolytic anemia. She required three units of blood and oxygen via nasal canula. A chest x-ray showed pulmonary edema and she was started on diuretics. The dapsone was stopped due to its known risk of anemia in certain populations.

A skin biopsy on a bullae site was obtained and showed neutrophilic infiltration, although nonspecific, was consistent with a recent insect bite. On day seven, the bite site showed central necrosis. There was slight improvement in the generalized rash. The antibiotics were discontinued, and she was weaned off oxygen. She was discharged on a short prednisone taper.

Conclusions

This case highlighted some potential challenges of diagnosing and managing systemic loxoscelism. Without venom assays, a presumptive diagnosis is made based on a known spider bite with correlating symptoms. Subsequent treatment often follows a sepsis protocol without clear evidence behind antibiotics. The patient presentation of persistent bullae is also atypical. Knowledge of the endemic regions may help in making a diagnosis alongside supportive management.

COVID-19 and Heart Transplant: The Virus that Changed the Landscape of Medicine

Nikhil Parimi, M.D., Jane Titterington, M.D., Ph.D.
KU School of Medicine-Kansas City

Introduction

We illustrate a case of a 36-year-old female with end stage cardiomyopathy who was found to be SARS-CoV-2 positive and subsequently underwent an orthotopic heart transplant.

Case Presentation

A 36-year-old female presented with over ten years of heart failure symptoms. Echocardiogram demonstrated severely diminished ejection fraction of 20% and the patient was diagnosed with non-compaction cardiomyopathy. She was being managed with home inotrope while being evaluated by the transplant committee. The patient was notified by the committee of an available donor heart. She was admitted the following day to the hospital and pre-procedurally screened for COVID-19, which resulted positive. Upon further questioning, patient reported mild respiratory symptoms in early April with positive SARS-CoV-2 PCR at that time. Infectious disease recommended to delay proceeding with heart transplant due to concern of reactivation of the virus with post-procedural immunosuppression regimen until after a 14-day quarantine with follow-up PCR testing. Her repeat PCR test was negative, and she was notified of an available donor heart shortly after with subsequent successful transplant. Her immune suppression induction consisted of high dose steroids and Simulect, and she was then maintained on steroids, mycophenolate, and tacrolimus. Her initial endomyocardial biopsy one week after transplant demonstrated moderate to severe (2R-3R) cellular rejection. The patient was started on IV Solumedrol and subsequently discharged on a steroid taper. Subsequent biopsy the following week showed resolving (1R) cellular rejection.

Conclusions

This is the first established case of a Coronavirus positive patient that received an orthotopic heart transplant at our institution. Given the novel nature of this virus, there has only been one case series investigating adult heart transplant recipients on chronic immunosuppression who subsequently developed COVID-19 post-transplant. The mortality rate of these 28 patients was found to be 25%, while none of the 28 patients experienced overt graft rejection, despite reduction of immunosuppression. To our knowledge, there is a paucity of information regarding COVID-19 patients who then underwent heart transplant. As demonstrated by this case, numerous challenges were encountered including safety of cardiac transplant with COVID positive PCR, timeframe of re-initiation on the transplant list, selection of induction and chronic immunosuppression regimen post-transplant, and effects of the virus on allograft rejection.

As the number of COVID-19 patients requiring heart transplant inevitably rises, we hope to have a better understanding between viral infection and its effect on cardiac transplant morbidity and mortality through published case reports or research studies.

Varicella Zoster Virus Meningitis in an Immunocompetent Adult Presenting as Isolated Intracranial Hypertension and Headaches

Pie Pichetsurnthorn, M.D., Pedro Vivar Cruz, M.D., Maha Assi, M.D., MPH
KU School of Medicine-Wichita

Introduction

Varicella Zoster Virus (VZV) meningitis is well described in immunocompromised adults but is rare among immunocompetent. Presenting signs and symptoms include neurological deficits and vesicular exanthems. We present a rare case of VZV meningitis in an immunocompetent patient mimicking idiopathic intracranial hypertension.

Case Presentation

A 59-year-old female with chronic migraines, obesity, and hypertension presented to the neurology clinic with longstanding headaches of increasing severity and frequency of one-month duration not relieved with abortive and prophylactic therapy. Physical exam was unremarkable except for papilledema. No abnormal skin findings were noted. MRI of the brain and MRV of the head demonstrated no intracranial mass, abnormal enhancement, or sinus thrombosis. Lumbar puncture was remarkable for elevated opening pressure (30 cm H2O) with immediate improvement of headache. Cerebral spinal fluid (CSF) analysis showed an elevated protein (79 mg/dL), normal cell count, and VZV detected by polymerase chain reaction. The patient denied history of chicken pox or zoster but recalled having had the chicken pox vaccine. She was admitted to the hospital and was started on intravenous (IV) acyclovir 600 mg every 8 hours with marked improvement in headache. She was discharged with planned 14 days total IV therapy duration. Follow-up after completion of antiviral therapy revealed complete resolution of headache.

Conclusions

Isolated VZV meningitis in an immunocompetent adult is rare. This case is unique as VZV meningitis presented as idiopathic intracranial hypertension without neurological deficits or exanthem, features commonly reported in previous case studies. In addition, the lack of pleocytosis in the CSF did not suggest meningitis, therefore the clinician might not be inclined to pursue PCR testing. However, the abnormally elevated protein in the context of persistent headache can be suggestive of aseptic meningitis and should prompt CSF PCR testing. Making a diagnosis of VZV meningitis is paramount for symptomatic relief as it responds best to full duration IV antiviral therapy, a distinct aspect not seen in treatments of other viral or aseptic meningitis infections in the absence of encephalitis. Providers should consider VZV meningitis when presented with new refractory headaches with increased intracranial pressure in immunocompetent adults even without a typical exanthem.

Reducing Morbidity in Acanthamoeba Keratitis

Clarissa Smith, M.D., Nida Ashraf, MBBS, Kenneth Goins, M.D., Jessica Newman, D.O.
KU School of Medicine-Kansas City

Introduction

Acanthamoeba are free-living protozoa found pervasively in water and soil which cause infections of the central nervous system, skin, and eye. Amebic keratitis (AK) is a sight-threatening infection that is associated with soft contact lenses due to corneal microtrauma and improper cleaning. Although AK infections are rare, they cause significant morbidity including vision loss due to diagnostic and therapeutic challenges.

Case Presentation

A previously healthy 38-year-old woman was referred to ophthalmology clinic for 3 months of left-sided pruritis and photosensitivity associated with headaches and declining visual acuity. Social history was significant for use of soft contact lenses and contact with cattle and cats. Physical examination was significant for left-sided conjunctival injection, decreased visual acuity, and corneal ulceration. Confocal microscopy revealed multiple double-walled cysts. Nucleic acid amplification testing of corneal scrapings was positive for Acanthamoeba. The patient's inflammation and ulceration worsened despite topical chlorhexidine, voriconazole, moxifloxacin, and atropine with oral valacyclovir and fluconazole. Due to disease progression, she underwent keratoplasty with corneal transplant. Pathology revealed severe inflammation with numerous cysts and trophozoites. Postoperatively, an aggressive regimen of oral miltefosine, voriconazole, and trimethoprim/sulfamethoxazole was added. The patient continued to improve and remains free of new or recurrent pathology.

Conclusions

AK is associated with profound vision loss and morbidity. The pathogen has two main phases in its life cycle: The trophozoite phase where it attaches and invades epithelial surfaces and the cystic phase which provides resiliency in suboptimal conditions. Miltefosine is a drug approved for treatment of Leishmaniasis that has shown efficacy against both the trophozoite and cystic forms of acanthamoeba by disrupting phospholipids and cytochrome c.

Diagnosis is often delayed due to overlap of symptoms with more common etiologies as well as the well-documented frequency of co-infection. Diagnosis is complicated further by barriers to diagnostic testing such as the need for expensive equipment and a highly-trained specialist associated with confocal microscopy. PCR and culture are also both sensitive but depend on collection techniques and require invasive procedures for specimen acquisition. Two or more methodologies are often performed concurrently due to the necessity for early identification. Reliable diagnosis and early treatment of AK is crucial to improve outcomes and preserve sight.

A Case of Domestic Cholera Due to Lack of Personal Protective Equipment

Nora Strong, M.D., Maha Assi, M.D., MPH
KU School of Medicine-Wichita

Introduction

Cholera is an acute secretory diarrheal illness caused by Vibrio cholerae with a potentially devastating mortality rate in untreated severe disease. It is primarily encountered in developing nations with poor sanitation and wastewater treatment, where it continues to cause outbreaks. It is very rarely encountered in resource-rich areas and is primarily associated with travel from endemic regions. This case is presented to alert clinicians to the possibility of domestic cases of cholera, a potentially fatal diarrheal illness, acquired through the fecal-oral route from animals.

Case Presentation

A 70-year-old male from the rural Midwest presented with a severe acute diarrheal illness. He reported fatigue, weakness, decreased oral intake, and multiple liquid stools for 2-3 days, without vomiting. The patient and his family denied any history of recent travel and any recreational water contact, including rivers, lakes, or ponds. Laboratory evaluation revealed acute kidney injury with hyperkalemia which improved with intravenous fluid resuscitation and medical management. Stool polymerase chain reaction (PCR) testing revealed Vibrio cholerae and enteropathogenic Escherichia coli. He was treated with 1 gram of azithromycin and recovered quickly. Upon further questioning, the patient reported that he worked at a feedlot spraying out cattle pens with no face protection from aerosols or splashes. He was instructed on the importance of proper use of personal protective equipment when handling biological waste and the state health department was informed of the pathogen.

Conclusions

Although cholera is mostly found outside the United States (US), it can also be encountered domestically. Although most cases in the US are imported through travel to endemic areas, it can also present as a zoonosis and constitute a workplace hazard in the agricultural setting with exposure to animal waste. This case highlights the importance of following safety measures based on Occupational Safety and Health Administration (OSHA) guidelines. Cholera should remain in the differential of acute diarrheal illness due to the potential severity of untreated disease and propensity for widespread. Proper treatment dramatically decreases morbidity and mortality.

HIV Associated Non-Hodgkin Lymphoma: A Rare AIDS Defining Illness

Jordan Tichenor, D.O., MBA, Rachel Weihe, M.D.
KU School of Medicine-Kansas City

Case Presentation

A 59-year-old Kenyan male presented with two weeks of nontender swelling on the left side of his neck and night sweats. His medical history included a new diagnosis of HIV/AIDS two months prior, complicated by disseminated cryptococcosis with meningitis. His viral load at initial presentation was greater than one million copies with a CD4 count of 70 (2%). He completed induction therapy with two weeks of amphotericin and flucytosine, with transition to fluconazole for consolidation therapy at discharge. Antiretroviral therapy (ART) was deferred initially to avoid immune reconstitution inflammatory syndrome, and he started Symtuza (Darunavir-Cobicistat-Emtricitabine-Tenofovir) two weeks prior to this hospital presentation.

During this hospital stay, an ultrasound of his neck showed pathologic left cervical lymphadenopathy. Computed tomography (CT) of the chest revealed a 1.9 centimeter right lower lobe pulmonary nodule, with mild mediastinal and right hilar lymphadenopathy. A few days later he underwent an IR guided biopsy of a cervical lymph node, with pathology and flow cytometry consistent with B-cell lymphoma.

He was discharged and re-admitted shortly thereafter for a DVT and pulmonary emboli, with new consolidations in his right upper and lower lobes on CT. He underwent a left inguinal lymph node biopsy with pathology showing Epstein-Barr Virus (EBV) positive diffuse large B-cell lymphoma. PET scan was remarkable for multiple hypermetabolic cervical, thoracic, and abdominopelvic lymph nodes, with focal uptake in the spleen, and the right lung masses. Bone marrow biopsy with flow cytometry was consistent with lymphoma. His Symtuza was changed to Biktarvy (Bictegravir-Emtricitabine-Tenofovir) to avoid drug interactions with planned chemotherapy. He was discharged to home to continue fluconazole (for cryptococcal meningitis maintenance therapy) and prophylactic trimethoprim-sulfamethoxazole. HIV viral load at discharge was less than 40, CD4 count 130 (4.3%). He was started on chemotherapy with intrathecal Methotrexate, but unfortunately expired soon after.

Conclusions

HIV associated non-Hodgkin lymphoma is an AIDS-defining illness, and rates of occurrence have declined with initiation of ART therapy. Diffuse large B cell lymphoma is the most common non-Hodgkin lymphoma seen with AIDS. Co-existing EBV or HHV-8 infection is common. Risk factors include CD4 count less than 100, high HIV viral loads, family history, and lack of ART. Patients will often present with B-symptoms (fever, night sweats, weight loss), cough, pancytopenia, increased LDH. Diagnosis often requires a biopsy. Treatment consists of ART along with chemotherapy agents.

Despite the increased availability of anti-retroviral therapy, AIDS associated lymphoma remains an aggressive disease that should be included in the differential diagnosis in patients presenting with B-symptoms and low CD4 counts.

Encephalitis with Subdural Empyema Disguised as an Acute Stroke

Rhythm Vasudeva, M.D., Thomas Moore, M.D.
KU School of Medicine-Wichita

Introduction

The NIH Stroke Scale (NIHSS) is a validated tool to assess neurological status when suspecting stroke. Elements on the scale like dysarthria and altered mentation, however, may overlap with other intracranial processes. Patients with underlying motor deficits and risk factors for stroke may further bias its interpretation. We present a case that mimicked features of a stroke, but found to have an underlying infection explaining the presentation.

Case Presentation

A 56-year-old male presented to the hospital after his wife found him confused with bilateral knee abrasions. An initial evaluation of the patient revealed dysarthria, confusion, and a NIHSS of 22, suggestive of severe stroke. Initial workup was significant for leukocytosis with a neutrophil predominance and no acute intracranial hemorrhage. The patient was admitted to the stroke unit with a diagnosis of acute encephalopathy secondary to a suspected stroke. The patient's medical history was significant for hypertension, dyslipidemia, atrial fibrillation with a Watchman™ device in-situ, chronic occlusion of left middle cerebral artery, history of multiple strokes with baseline right-sided weakness, and chronic sinusitis. His home medications included amiodarone, apixaban, aspirin, dabigatran, carvedilol, atorvastatin, and fluticasone nasal spray.

Upon admission, the patient was monitored on telemetry with frequent NIHSS scoring and permissive hypertension. MRI brain was done and sequelae from previous strokes were seen with a non-specific bright signal within the right hippocampus. Crusted vesicular lesions were noted along the C6-7 dermatomal distribution of the left arm. A lumbar puncture with CSF analysis revealed pleocytosis and elevated protein. IV acyclovir was started upon diagnosis of viral encephalitis after Varicella-Zoster virus was detected on the meningoencephalitis panel. A repeat MRI brain on day 3 of admission showed a new focal area of subdural fluid along the left frontal region and an increased signal in the left frontal lobe. This lesion evolved into a multiloculated fluid collection suggestive of an empyema on a follow-up MRI the following day. Further imaging showed sinus inflammatory changes with bony rarefaction in the walls of the left frontal sinus with a suspicious connection to the intracranial space. Patient's MRSA nares surveillance was negative from a recent admission. IV ceftriaxone and metronidazole were subsequently added to the treatment regimen.

The patient underwent nasal endoscopic surgery per otolaryngology's recommendation. Cultures of the left frontal purulence did not grow any organisms. The patient's clinical condition improved, and the patient was discharged to complete 21 days of IV antimicrobial therapy with respective follow-up visits.

Conclusions

Differential diagnoses, such as an infectious process in this scenario, should be considered when evaluating a patient with features of stroke, especially when significant risk factors for stroke are prominent. Potential clues may present on history, physical examination, and initial labs if carefully sought.

When Treatment Becomes Disease:
Disseminated BCG Infection after Intravesical Treatment for Bladder Cancer

Jerrica Werner, M.D., Lori Olson, M.D.
KU School of Medicine-Kansas City

Case Presentation

A 64-year-old man was admitted for severe sepsis thought secondary to an urinary tract infection related to complications of extravasation and necrosis of intravesical mitomycin-C requiring a urethrectomy due to urethral necrosis and perineal urethrostomy two months prior to admission. While he improved to transition out of the ICU, he and his wife shared a concerning story of weight loss, nausea and anorexia, recurrent fevers, subjective chills, and global failure to thrive for several months. He again fevered in the hospital, and a broad diagnostic work-up ensued evaluating for malignancy and infection. Pancytopenia and a quickly expanding infrarenal aortic pseudoaneurysm were further clues, and a diagnostic gastric biopsy revealed noncaseating granulomas. The patient was started on isoniazid, rifampin, and ethambutol for presumed disseminated Bacillus Calmette-Guérin (BCG) infection related to intravesical BCG therapy he had received two years earlier. Steroids were added when a renal biopsy confirmed active inflammation along with granulomas.

Conclusions

BCG is a live attenuated strain of Mycobacterium bovis, developed as a vaccine in the early 20th century against Tuberculosis. Much later, in the 1980s, BCG replaced cystectomy as the treatment of choice for Carcinoma in situ of the bladder as an immunomodulator by inducing a localized immune response leading to cell death. Over thirty years later, BCG is a continued proven primary treatment or adjunct for various types of bladder cancer. Intravesical BCG is generally well tolerated; however, infectious manifestation can occur both in early-onset infection as well as in a delayed-onset infection. Early onset is more often a systemic disease process whereas the delayed onset tends to be a localized illness. With treatment, the patient had significant improvement in fatigue, has been able to regain weight, and has had resolution of fevers and chills. His cell counts, kidney function, and pseudoaneurysm have all stabilized.

This case illustrates the potential for infectious complications after intravesical BCG therapy for bladder cancer and the value of a complete history when encountering fever of unknown origin. Although uncommon, systemic infection is a known side effect of this therapy; clinicians should maintain a degree of suspicion for disseminated BCG infection in patients as the etiology for systemic disease even years after its use in intravesical therapy. Early recognition is essential as initiation of anti-tuberculous treatment is critical to minimize further systemic and life-threatening complications.

Auscultated Blood Pressure Rounding Errors and Racial Disparities

Joshua Beyer, D.O., Robert Badgett, M.D., Hayrettin Okut, Ph.D.
KU School of Medicine-Wichita

Introduction

Auscultated blood pressure values are routinely performed and providers rely on accurate data when prescribing anti-hypertensive medications. There is a well-established tendency for humans to round blood pressure readings to the nearest zero or five terminal digit, and existing data from the KUSM-W Adult Medicine Clinic supports this observation. To elucidate if this human rounding bias contributes to racial health care disparities, we studied blood pressure recordings in patients to search for possible asymmetries in blood pressure distributions among self-reported White vs. non-White patients.

Methods

The charts of clinic patients were reviewed retrospectively between April, 2014 and April, 2020. In accordance with JNC 8 hypertension guidelines, blood pressure readings were evaluated that centered around specific zero-end digit cutoffs of 140 mmHg systolic and 90 mmHg diastolic. Ethnicity of patients was based on self-report. Bias in measurement was tested by: 1) measuring the distribution of diastolic and systolic pressures among White patients, 2) measuring the same for non-White patients, 3) creating a predicted non-White distribution of pressures based on the measured distribution of White values, and 4) testing the goodness of fit of the measured Non-white pressures to the predicted non-White values using chi-square.

Results

Data included 15,166 blood pressure measurements in the KUSM-W Adult Medicine Clinic. Of these, 2726 diastolic values were between 85 and 95 mm Hg and of these 758 also had systolic pressures between 135 and 145 mm Hg. Thus, 361 non-White and 397 White systolic values between 136 and 144 mm Hg and 1229 non-White and 1498 White diastolic values between 86 and 94 mm Hg were included. For systolic pressures of non-White patients, no significant difference was found between measured and predicted values (chi-squared = 3.302, df = 4, p-value = 0.509). For diastolic pressures of non-White patients, a significant difference was found between the measured and predicted values (chi-squared = 16.68, df = 4, p-value = 0.002). For examination of the diastolic distribution of non-White patients, the frequency of rounding up to a value of 90 mmHg was calculated to occur in 5.70% of patients.

Conclusions

The data supported a statistically significant tendency for auscultated diastolic blood pressure recordings of non-White patients to be rounded up to 90 mmHg in approximately 6% of patients whose diastolic pressure was 86 to 89 mm Hg. This is congruent with our hypothesis of racial asymmetry. Automated blood pressure devices more accurately correlate to ambulatory blood pressure readings and would reduce human bias and help to ensure equal care for patients regardless of race.

Tumor Mutation Burden, a Marker of Response to Immune Checkpoint Inhibitors: A Systematic Review and Meta-Analysis

Khalil Choucair, M.D., Hayrettin Okut, Ph.D., Ammar Al-Obaidi, M.D., Susan Morand, Laura Stanbery, Ph.D., K. James Kallail, Ph.D., John Nemunaitis, M.D.
KU School of Medicine-Wichita

Introduction

Immune Checkpoint Inhibitors (ICIs) have revolutionized cancer therapy. Despite durable response rates not all patients respond as effectively to ICIs. Thus, a strong need exists to explore biomarkers that contribute to proper patient selection. Tumor mutation burden (TMB) is an emerging genomic biomarker that quantifies the total number of neo-antigens present in a tumor specimen. Therefore, a systematic review and meta-analysis were conducted to explore the association between TMB and outcomes of ICIs across several solid tumors.

Methods

PubMed, Embase, Web of Science, and Cochrane databases (to December 2019) were searched for eligible studies. Studies reporting 1-year overall survival (1-YR OS), median progression-free survival, and median overall survival were included in the qualitative analysis. The primary endpoint of the meta-analysis was 1-YR OS and the pooled hazard ratios (HR) and 95% confidence interval (CI) for 1-YR OS were performed using random effect model.

Results

Sixteen studies, including 3,787 patients, met eligibility criteria: most patients had non-small cell lung carcinoma (NSCLC; n=2,200). Patients received ICIs as 1st line therapy (n=1,081) or > 1st line (n=1,421). The line of therapy was not reported in all studies. Studies involved PD-1/PD-L1 inhibitors monotherapy (n=9), anti-CTLA-4 (n=1), or both (n=3), and various ICIs (n=3). In the meta-analysis, 156 patients from 10 studies reporting 1-YR OS were included. Comparing high (n=631) to low TMB (n=1,525), the pooled HR for 1-YR OS was -0.81 [95% CI: -1.63 to 0.02; $P < 0.01$] in patients with high TMB. Subgroup analysis revealed consistently higher 1-YR OS in patients with high TMB vs. low TMB regardless of histology (NSCLC: 3 studies; HR: -0.47 [95% CI: -0.75; -0.20]; SCLC: 2 studies; HR: -0.68 [95% CI: -1.48; 0.12]; pool of diverse solid tumors: 2 studies; HR: -0.40 [95% CI: -1.49; 0.68]). Similarly, patients with high TMB had significantly longer 1-YR OS regardless of sequencing methods (Whole Exome: 5 studies; HR: -0.49; [95% CI: -0.96; -0.02] or Next Generation: 5 studies; HR: -1.14; [95% CI: -2.82; 0.53]. 1-YR OS was consistently higher in patient with high TMB with nivolumab monotherapy (2 studies; HR: -0.32 [95% CI: -0.66; 0.02], any anti-PD1/L1 monotherapy (7 studies; HR: -0.90 [95% CI: -2.08; 0.28], and combination therapy (n=2 studies; HR: -1.01 [95% CI: -01.37; -0.66]). This effect persisted in 1st line (3 studies; HR: -0.42 [95% CI: -0.77; -0.07] or later lines of therapy (>1 line; 5 studies; HR: -1.23 [95% CI: -2.90; 0.43].

Conclusions

TMB may be a tumor-agnostic biomarker of response to ICIs and survival regardless of the histological subtype, the line of therapy, sequencing method, or ICI therapy. Primary physicians' understanding of the significance of such tumor-agnostic biomarkers is essential to facilitate informed discussions with their cancer patients.

Resident and Patient Satisfaction with Bedside Rounds: Is There Room for Innovation?

Tanner Dean, D.O., Robert Badgett, M.D.
KU School of Medicine-Wichita

Introduction

COVID-19 has forced academic medical teams to reexamine processes that had previously been unquestioned. As social distancing, personal protective equipment uses, and restricting patient-family-provider interactions become the new norm, the culture of team rounding needs an evidenced-based backing to either continue at the patient's bedside or innovate to less exposed means. Our objective was a systematic review of relevant trials that compared bedside to non-beside rounding for the outcomes of resident and patient satisfaction.

Methods

We sought trials with a mix of Boolean searching and citation pearling. Results were converted to standardized mean differences (SMD) for random effect meta-analysis. We interpret the SMD as 0.2 = small; 0.5 = medium; 0.8 large. Initial Boolean searching revealed 1,374 relevant articles. Abstracts were then reviewed to identify articles comparing bedside vs. non-bedside rounding styles and ten trials were identified.

Results

Ten trials were identified (8 though OVID search and 2 through citation pearling). Six trials reported patient preference or satisfaction while four trials reported resident satisfaction or preference. One trial reported both resident and patient satisfaction. The settings of the trials were: Seven internal medicine wards, one family medicine ward, and one labor and delivery ward. The setting was a statistically significant modulated of both outcomes. Patient satisfaction was increased in two studies, the single study conducted in obstetrics (SMD = 1.18; CI: -2.06 to -0.03; large effect) and only one of the internal medicine studies. Resident satisfaction almost researched significance (SMD = 1.65; CI: -3.36 to 0.06) in the same trial in obstetrics.

Conclusions

These preliminary search results add two additional studies to a previous meta-analysis and suggest question the need for medical teams to routinely round at the patient's bedside in settings other than labor and delivery.

Benefits of Aspirin for the Prevention of Esophageal Squamous Cell Carcinoma and Adenocarcinoma

Yasmine Hussein Agha, M.D., Sachin Srinivasan, M.D., Chelsea Wuthnow, M.D., Ali Taleb, M.D., Katia El Jurdi, M.D., Charles Buess, M.D., Nathan Tofteland, M.D., William Salyers, M.D.
KU School of Medicine-Wichita

Introduction

Esophageal cancer has consistently been among the ten most common cancers in the United States. Adenocarcinoma (EAC) and squamous cell cancer (SCC) make up 90% of all malignant esophageal tumors. Tobacco and alcohol consumption have been recognized as leading risk factors for SCC, while Barrett's esophagus and gastroesophageal reflux are associated with an increased risk of progression to EAC. While attempting to control these modifiable triggers, the incidence of the squamous cell subtype has steadily decreased over the last decade; however, the incidence of EAC has continued to rise. Multiple studies have evaluated the role of cyclooxygenase inhibitors in the development of cancer, especially esophageal tumors. The aim of this meta-analysis is to evaluate the potential benefits of aspirin as a chemoprotective agent against esophageal cancer.

Methods

We performed an electronic search of PUBMED databases for studies that reported data on esophageal cancer occurrences (all, adenocarcinoma (EAC) and squamous cell carcinoma (ESCC) and aspirin use from inception to January 1, 2020. Case reports, case series, editorials and review articles were excluded. Primary outcome included pooled odds of all esophageal cancer occurrence in persons who were on aspirin compared to those who were not. Secondary outcomes included the pooled odds of EAC and ESCC occurrence in persons who were on aspirin compared to those not taking it. We also performed a meta-regression for the cancer incidence (all, EAC and ESCC) based on duration of follow-up. Meta-analysis outcomes, heterogeneity (I2), and meta-regression (for effect of covariates) were derived by statistical software R and open meta-analyst.

Results

A total of 11 studies with 34,779 patients with a mean age of 62.3 ± 5.6 years and a mean follow-up duration of 116 months (range 24-264 months) were included for analysis. There were 6 studies that reported data on incidence of overall esophageal cancer, 7 studies reporting incidence of EAC, and 4 studies reporting of incidence of ESCC in patients who were on aspirin. Odds of esophageal cancer were significantly lower in individuals who were taking aspirin compared to those who were not (OR 0.54; 95% CI 0.33-0.88; I^2=82%). We also noted significantly lower odds of both EAC (OR 0.60; 95% CI 0.44-0.82; I^2=69 %) and ESCC (OR 0.59; 95% CI 0.46-0.78; I^2=79%) in patients who were taking aspirin compared to those who were not. Meta-regression did not demonstrate any significance of duration of follow-up to the incidence of all esophageal cancer (p=0.38), EAC (p=0.29), or ESCC (p=0.45). We did not assess for publication bias since there were fewer than 10 studies in all outcome evaluations.

Conclusions

The use of daily aspirin was associated with decreased incidence of all esophageal cancer, EAC, and ESCC.

A Descriptive Study of IV Hydralazine Use in Adults with Hypertensive Urgency at an Academic Medical Center

Logan Kline, D.O., Shannon Stittsworth, Pharm.D., Abebe Abebe, M.D., Joanne Moore, RPh, Ethan Alexander, M.D.
KU School of Medicine-Kansas City

Introduction

Hydralazine is a potent vasodilator approved for second line therapy to treat hypertensive emergency, defined as a blood pressure (BP) ≥ 180 mmHg systolic and or ≥ 120 mmHg diastolic with signs of end organ damage. IV hydralazine is often used to treat hypertensive urgency at the University of Kansas Health System (TUKHS), which is defined with the same BP parameters but without signs of end organ damage. There are no guidelines or consensus on the treatment of hypertensive urgency, in fact studies argue that it is widely overtreated leading to hypotension and reflex tachycardia. The study purpose was to assess the utilization of IV Hydralazine for hypertensive urgency.

Methods

A single-center retrospective chart review was conducted for IV hydralazine doses ordered PRN or once and administered between August 1, 2018 and July 31, 2019. Non-critical care patients 18 years and older were included if they received one or more doses of IV hydralazine. Patients who were pregnant, had a history of stroke or heart failure, used hydralazine prior to admission, or with a diagnosis of hypertensive emergency were excluded. Data collected included demographics, hydralazine dose and frequency, vitals one hour before and after administration, unit and service during administration, and past medical history. The results summarize the frequency of IV hydralazine orders, administration, and resulting blood pressure and heart rate.

Results

3,706 administered doses were extracted. 10 mg every 6 hours was the most common dose and frequency ordered. Of the 1,148 patients who met inclusion criteria, 40.8% carried a diagnosis of hypertension prior to hospitalization. 36.1% of doses were given to patients with a BP > 180 mmHg systolic and/or > 120 mmHg diastolic. 16% of doses were administered without a recorded BP one-hour prior. Of the patients who did have a BP recorded prior to administration, 39% did not have a BP recorded within one hour after administration. Approximately 25% of patients developed reflex tachycardia following administration.

Conclusions

63.9% of patients receiving IV hydralazine did not meet BP parameters for hypertensive urgency, leading us to ask why these patients received this medication. Additionally, most patients were not monitored appropriately, either not having their BP checked one hour prior to and/or after administration. This has profound safety implications. Currently, the enterprise medication safety committee is reassessing monitoring parameters for IV hydralazine. Planned future directions include a subset analysis that will dive deeper into the reasons why these patients were given IV hydralazine, in hopes to formulate criteria for use policy related to this medication.

Disclosure of Unexpected Outcomes to Patients: Resident Attitudes and Practices Before and After Training Program

Nikki Miller, M.D., Leigh Eck, M.D., Becky Lowry, M.D., Marie Brubacher, M.D., Jessica Kalender-Rich, M.D.
KU School of Medicine-Kansas City

Introduction

Unexpected outcomes and errors are inevitable, but difficult to disclose and report. Research tells us that patients want disclosure when errors occur and that resident physicians are inclined to disclose these errors. Despite these concordant attitudes, only a minority of errors are either disclosed or reported. It is important that our physicians are trained in reporting and disclosing medical errors and unexpected outcomes to patients and their families.

Methods

PGY2 and PGY3 internal medicine residents were enrolled in the Disclosure of Unexpected Outcomes Training Program. The program consisted of a pre-work packet, pre-survey, standardized patient (SP) experience, and two post-surveys. The first post-survey was administered immediately after the SP experience and the second post-survey was administered 3 to 6 months after the SP experience.

Results

Responses were received from 50 (100%) participants from the pre-survey, 25 (50%) from the first post-survey, and 11 (22%) from the second post-survey. In the pre-survey, almost all residents responded that they would feel an obligation to disclose an error to a patient (58% strongly agree and 40% agree). 78% of residents reported disclosing an actual minor error (resulting in prolonged treatment or discomfort), while 44% of residents acknowledged that they made a minor error and had not disclosed it to the patient. 2% of residents made a major error (resulting in disability or death) and disclosed it to the patient, while 0% of residents made a major error and had not disclosed it to the patient. Only 47.8% of residents had participated in disclosing an unexpected outcome. All residents that participated in the first post-survey responded that they would feel an obligation to disclose an error to a patient (64% strongly agree and 36% agree). However, only 76% of residents reported disclosing a minor error, while 36% of residents acknowledged that they made a minor error and had not disclosed it to the patient. No residents reported making a major error. All residents that participated in the second post-survey responded that they would feel an obligation to disclose an error to a patient (81.8% strongly agree and 18.2% agree). 73% of residents reported disclosing an actual minor error, while 36% of residents acknowledged that they made a minor error and had not disclosed it to the patient. No residents reported making a major error. 90.9% of residents had participated in disclosing an unexpected outcome after the SP experience.

Conclusions

It appears that there is a difference between resident physicians' attitudes and practices regarding disclosing unexpected outcomes. It was shown that residents more strongly agreed that they would feel obligated to disclose an error after the training program. Actual participation in disclosing an unexpected outcome increased after the SP event.

Assessing the Impact of a Diuresis Report on Electrolyte Management for Acute on Chronic Heart Failure

Dale Smith, M.D., Luke Nelson, M.D., Alex Robinson, D.O., Gong Weng, M.D., Laura Thomas, M.D., Heath Latham, M.D.
KU School of Medicine-Kansas City

Introduction

Multiple large studies of heart failure patients show that hypokalemia is associated with increased short-term mortality. Due to large time-gaps between data points, managing potassium is difficult in this cohort of outpatients. To reduce the risk of hypokalemia post-discharge, we developed a comprehensive diuresis report in the University of Kansas Health System's inpatient electronic health record. The diuresis tracking report includes weight, electrolytes, intake, output, diuretic dose, and kidney function. We aimed to improve potassium management for recently discharged heart failure patients by providing clearer data for the inpatient teams, and consequently better electrolyte control in the outpatient setting.

Methods

After obtaining IRB-approval for this quality improvement (QI) project, we used retrospective chart review to measure diuresis and electrolyte management for patients hospitalized with acute on chronic heart failure for three months before and after the introduction of the diuresis report. To reduce provider variability, we chose to narrow our QI project to those patients discharged by specialty nurse practitioners on the heart failure service at the University of Kansas Health System. In addition, we performed a survey to measure provider satisfaction.

Results

The number of hypokalemic events at first follow-up were reduced after employing the diuresis report. Of the 23 patients discharged before implementation of the diuresis report, 3 recorded a potassium less than or equal to 3.5 mmol/L. Due to the impacts of COVID-19, only 12 patients were discharged after implementation of the diuresis report, with no hypokalemic events. It is unclear if the reduced number of hypokalemic events was related to the diuresis report versus an overall reduced number of admissions related to COVID-19 to the heart failure service which allowed more time for NPs to focus on individual patient care. However, provider satisfaction improved after use of the report, with all survey participants reporting they were satisfied with how they tracked diuresis, as opposed to only 20% before the report. Providers also used fewer reports to manage diuretics; using between 2-5 before the report, and between 1-2 after.

Conclusions

The diuresis report improved provider satisfaction and may have contributed to improved inpatient electrolyte management for patients hospitalized with acute on chronic heart failure by specialty nurse practitioners, though the results could have been affected by the impact of the COVID-19 pandemic on heart failure admissions.

Increasing Tdap Vaccination Rates in Internal Medicine Resident Clinic

Stephanie Slimmer, M.D., Grant Kirby, M.D., Robert Weidling, M.D.,
Raabia Nizamuddin, M.D., Maire Okoniewski, D.O., Joe Johnstone, M.D., Jamison Ryan, M.D.,
Autumn Brubaker, D.O.
KU School of Medicine-Kansas City

Introduction

The CDC recommends the Tdap vaccination at least once in adulthood followed by booster Td every ten years. There are tremendous cost saving implications related to this vaccine preventable illness, which accounts for a significant cost and burden to the health care system. The Tdap vaccination is generally well tolerated with limited contraindications. It can be given with other vaccinations and provides adequate immunologic protection. The internal medicine resident patient population was identified to have a lower Tdap vaccination rate when compared to faculty. Our aim was to improve Tdap/Td vaccination rates from 57% to 64% in non-Medicare patients ages 18-64 at Kansas University Medical Center General Medicine clinic over a three-month period by implementing a standing order system for nursing and administration of Tdap vaccination.

Methods

A subset of residents in the internal medicine clinic were selected for implementation of a new protocolized system of Tdap administration. This patient population included non-Medicare adults aged 18-64 presenting to internal medicine clinic. A fishbone analysis was used to create a better understanding of the existing protocols used by clinic and staff prior to administering a Tdap vaccination. The first PDSA cycle included implementing a standing order protocol for the nurses to administer the vaccine. This standing order allowed nurses to give the vaccine at the beginning of the visit after following a protocol of chart reviewing to determine the need for the vaccine. This was implemented after in-person education with nurses and emails informing the residents of the protocol were completed. Based on feedback obtained through written surveys after one week, nurse buy-in needed increased and familiarity with protocol for residents needed improved. Prior to the second PDSA cycle, further intervention included nursing huddle attendance to discuss protocol and completing in-person education to the residents.

Results

An increase of 4.45% in vaccination compliance rates was seen in an internal medicine resident clinic cohort after two PDSA cycles of implementing a standing order protocol for Tdap vaccination administration. Prior to intervention, the internal medicine resident clinic cohort Tdap vaccination rate was 57.44%. After implementing the standing order, the same resident cohort percentage had increased to 61.89%, a change of 4.45%. Medicare patients were excluded from this study as Medicare insurance does not cover the Tdap vaccination.

Conclusions

By implementation of a new protocol for administration of the Tdap administration, we were able to successfully improve vaccination rates near faculty average. A systems-based understanding of vaccination administration was gained, allowing both resident and nursing input in order to implement an effective protocol and improve compliance rates. Broader implementation to the entire resident clinic can be used to improve vaccination rates across all internal medicine resident clinic patients.