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Abstracts from the 2018 Annual Meeting
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1. **Rapid Progressive Dementia with Probable Sporadic Creutzfeldt-Jakob Disease**
   Sinan Khayyat, M.D., Daly Al-Hadeethi, M.D., Mohinder Vindyhal, M.D., M.Ed.
2. **When it isn't Sepsis: A Case Report of Hemophagocytic Lymphohistiocytosis**
   Kelsey Witherspoon, M.D., Mohinder R. Vindhyal, M.D., M.Ed.
3. **Not Nearly Enough Kleenex: A Life Threatening Vascular Tumor Complication**
   Ryan Morse, Jesse Richards, D.O., Matthew Swan, M.D.
4. **Sepsis-Induced Radiation Recall Dermatitis after Chemotherapy with Irinotecan and Bevacizumab**
   John Frey, D.O., Christopher Streiler, M.D., Matthew Swan, M.D.
5. **Macrophage Recruitment during Liver Repair is Attenuated and Delayed by Hyaluronan Synthesis Inhibition**
   Ben Harstine, Jennifer M. McCracken, Ph.D., Lei Jiang, Ph.D., Michele T. Pritchard, Ph.D.
6. **Once Bitten, Twice Shy: The Thick and Thin of Blood Parasites**
   Jamison Ryan, M.D., Lisa Clough, M.D.
7. **Lumbar Radiculopathy and Iliopsoas Abscess as Presenting Manifestations of Ruptured Appendicitis**
   David Rayan, Gillian Dryton, Anita Moudgal, B.A., M.Ed., Amanda Jobe, M.D., Radhika Gupta, MBBS, Bharata Lankachandra, M.D.
8. **Determination of Antimicrobial Prescribing Practices in an Integrated Health System Emergency Department**
   John Price, Sarah Sartain, M.D., Brooke Bitner, Pharm.D.
9. **Bone-Marrow Sarcoidosis in a Patient Presenting with Fever of Unknown Origin**
   Sanjay Parashar, Jesse Richards, D.O., Megan Krause, M.D.
10. **Babesiosis in an Immunocompromised Patient**
    Sonya Parashar, M.D., Rachel Weihe, M.D., Jessica Newman, D.O.
11. **BMP Signaling Regulates Support Cell Proliferation in the Regenerating Zebrafish Neuromast**
    Madeline St. Peter, Linjia Jiang, Ph.D., Tatjana Piotrowski, Ph.D.
12. **Acquired Factor VIII Inhibitor Related Retropharyngeal Hematoma: A Novel and Rare Differential for Acute Dysphagia and Odynophagia**
    Ryan Zwick, Jesse Richards, D.O., Ben Fangman, M.D., Abebe Abebe, M.D.
13. **Risk of Lead Dislodgement with Catheter Ablation in Patients with Recently Implanted Cardiac Implantable Electronic Device**
    Brendan Cokingtin, Mohammad-Ali Jazayeri, M.D., Kevin Cokingtin, Venkat Vuddanda, M.D, Courtney Jeffery, APRN-C, Donita Atkins, BSN
14. **Nelson's Syndrome: A Rare Treatment-Related Complication of Refractory Cushing's Syndrome**
    Anam Abbasi, M.D., Sachin Srinivasan, M.D., Georges Elhomsy, M.D.

15. **Transgender Health Medical Education Intervention and its Effects on Beliefs, Attitudes, Comfort, and Knowledge**
    Joseph Cherabie, M.D., Kari Nilsen, Ph.D., Sarah Houssayni, M.D.
16. **Effective Management of Obesity with Generic Combination of Phentermine/ Topiramate vs Qsymia®: Is it Possible?**
    Wajeeha Rizvi, M.D., Nicholas Ojile, M.D., Milan Bimali, Ph.D., K James Kallail, Ph.D., Georges Elhomsy, M.D.
17. **Polyarticular Calcium Pyrophosphate Deposition Disease Masquerading as Pseudoseptic Arthritis**
    Sarah El Chami, M.D., Timothy Shaver, M.D., Paul Ndunda, M.D.
18. **Elevated Procalcitonin with Atypical Presentation**
    Rachel Sigler, D.O., Kassem Hammoud, M.D.
19. **Polymicrobial Endocarditis**
    Joseph Borick, M.D., Thomas Moore, M.D.
20. **Ice Induced Cardiac and Systemic Thrombus**
    John Eliveha, M.D., Shravani Vindhyal, M.D., Mohinder Vindhyal, M.D., M.Ed.
21. **Non-Atherosclerotic Coronary Angina**
    Shravani Vindhyal, M.D., John Eliveha, M.D., V. Subbarao Boppana, M.D., Mohinder Vindhyal, M.D., M.Ed.
22. **Miliary TB in a Patient with End-Stage Liver Disease**
    Victoria Poplin, M.D., Brent Harbaugh, M.D., Nathan C. Bahr, M.D.
23. **Paenibacilli - Newly Identified Organism Concealed in an Old Genus**
    Tori Kunkel, Jessica Newman, D.O.
24. **Characteristics of Smokers among Patients in a Hepatology Clinic**
    Daly Al-Hadeethi, M.D., Victor Ankoma-Say, M.D.
25. **Computed Tomography Fractional Flow Reserve: An Appropriate Low-Risk Screening Tool for Coronary Disease**
    Alexander Robinson, D.O., Nicholas Isom, M.D., Chris Buckley, M.D., Thomas Rosamond, M.D.
26. **Use of Rituximab in Treatment of Recalcitrant Dermatitis Herpetiformis**
    Chisom Eze, M.D., Timothy Shaver, M.D.
27. **Acquired Methemoglobinenia and Sulfahemoglobinemia Secondary to Topical Lidocaine Use**
    Brandon Carlisle, M.D., Laura Thomas, M.D.
28. **Clinical Relationship between CVID and Inflammatory Arthritis**
    Reid Eggleston, Megan Krause, M.D.
29. **Long-Term Use of Dual Antiplatelets for the Secondary Prevention of Atherothrombotic Events in Patients with and without MI: Meta-Analysis of Randomized Controlled Trials**
    Sumaya Hammami, M.D., Amratash Malodiya, Ph.D., Sandra Weiss, M.D., Paul Kolm, Ph.D., William Weintraub, M.D., Zaher Fanari, M.D.
30. **VT Storm after Anti-Diarrheal Agent**
    Jihad Al-Khatib, Shravani Vindhyal, M.D., V. Subbarao Boppana, M.D., Mohinder Vindhyal, M.D., M.Ed.
31. **A Case of Red Herrings: Invasive Fungal Sinusitis**
    Kristen Constance, M.D., Nicholas Ojile, M.D., Brent Duran, D.O., Shelley Jones, M.D.

32. **A Nearly Deadly Skin Lesion: Subcutaneous Rhizopus Murcor in a New Diabetic**
    Carley Trentman, Jesse Richards, D.O., Melissa Taylor, M.D.
33. **The Effect of Pembrolizumab in Absence of Programmed Death 1 Receptor**
    Layth Al Attar, M.D., Phu Truong, M.D.
34. **An Often Unthought of Cause for High-Output Heart Failure**
    William Goodman, M.D., Mona B. Brake, M.D.
35. **Parathyroid Crisis in a Patient with a Benign Parathyroid Adenoma**
    Nicholas Ojile, M.D., Georges Elhomsy, M.D.
36. **Takotsubo Cardiomyopathy Leading to Left Ventricular Free Wall Rupture**
    John Fritzlen, M.D., Tarun Dalia, MBBS, Bashar Amr, M.D., Jared Kvapil, M.D.
37. **Extensive Deep Venous Thrombosis in a 31-Year-Old Female on Oral Contraceptives Diagnosed with May-Thurner Syndrome**
    Erik Calderon, M.D., Brent Duran, D.O., Stephanie Pankow, D.O.
38. **Not Your Usual Suspect: A Case of Pill-Induced Esophagitis Caused by Demeclocycline**
    Gong Weng, M.D., Matthew Swan, M.D., John A. Bonino, M.D.
39. **A Case of Intraparenchymal, Intraventricular, Subdural, and Subarachnoid Hemorrhage Following Catheter-Directed tPA for Extensive Iliac Deep Venous Thrombosis**
    Maha Mohamad, M.D., Rami H. Diab, M.D., John Peterson, D.O., Surayakumar Reddy, M.D., Nazih Moufarrij, M.D.
40. **Association between Electronic Cigarette Use and Coronary Heart Disease and Stroke**
    Job Mogire, M.D., Tabitha Muutu, M.D., Paul Ndunda, M.D.
41. **Blurred Lines: HLH Recognition Complicated by Infectious, Neoplastic, and Autoimmune Features**
    Clarissa Smith
42. **Percutaneous Repair of Mitral Valve Regurgitation Due to Post-Infarction Papillary Muscle Rupture Using Mitraclip Apparatus**
    Cyrus Munguti, M.D., Paul Ndunda, M.D., Bassem Chehab, M.D.
43. **The Problematic Nature of Fibromyalgia Diagnosis in the Community**
    Sachin Srinivasan, M.D., Eamon Maloney, M.D., Rami Diab, M.D., Brynn Wright, K. James Kallail, Ph.D., Michael Kennedy, M.D., Frederick Wolfe, M.D.
44. **A Rare Case of Prekallikrein Deficiency**
    Sammy Tayiem, M.D., Abdulraheem Yacoub, M.D.
45. **A Rare Case of Nonbacterial Thrombotic Endocarditis (NBTE) in a Young Patient with Acute Myelogenous Leukemia**
    Rami Atallah, M.D., Nicholas Ojile, M.D., Kamal Chamoun, M.D., Fredy Nehme, M.D., Mohinder Vindhyal, M.D., M.Ed.
46. **Leclercia Adecarboxylata in a Post-Fasciotomy Polymicrobial Infection**
    Andrew Weaver, M.D., Margaret E. Hagan, M.D.
47. **When Empiric is Imperative: Differentiating Encephalopathy from Encephalitis**
    Gretchen Beaver, Jesse Richards, D.O., Jessica Newman, D.O.
48. **Immunotherapy Induced Myocarditis**
    Garret Seiler, D.O., Aarati Keshary, M.D., Jeremy Deutsch, M.D.
49. **Cutaneous Leishmaniasis**
    Thanuja Neerukonda, Ryan Kubat, D.O., Fernando Merino, M.D.

50. **From Murmur to Amyloidosis: Stressing the Importance of Physical Exam**
    Ethan Hacker, M.D., Branden Comfort, M.D., MPH
51. **Congenital Renal Arteriovenous Malformation: A Rare but Treatable Cause of Hypertension**
    Nicholas Isom, M.D., Reza Massomi, M.D., Adam Alli, M.D., Kamal Gupta, M.D.
52. **Chronic Inflammatory Arthropathy in the Setting of Levamisole-Induced Vasculitis**
    Kyle Myers, D.O., Pooja Bhadbhade, D.O.
53. **Burning Fat 24/7: Dangers of the Ketogenic Diet when Taking a SGLT-2 Inhibitor**
    Sukhindervir Sandhu, Jesse Richards, D.O., Candice Rose, M.D.
54. **Gender Disparities in Secondary Preventive Measures for Atherosclerotic Cardiovascular Disease in Kansas**
    Roshni Jain, Tabitha Muutu, M.D., Paul Ndunda, M.D.
55. **Treatment of Refractory Acute Myeloid Leukemia with Isocitrate Dehydrogenase Inhibitor**
    Aarati Keshary, M.D., Jeremy M. Deutsch, M.D.
56. **Maintaining a High Suspicion for Ehrlichiosis in the Heartland**
    Mejalli Al-Kofahi, M.D., Usman Nazir, M.D.
57. **A Rare Cause of Anemia: 74-year-old Male with High-Grade Neuroendocrine Tumor of the Colon**
    Nicole Balmaceda, Mohammad Telfah, M.D., Mazin Al-Kasspooles, M.D., Hongyan Dai, M.D., Ph.D., Prakash Neupane, M.D., Joaquina Baranda, M.D.
58. **Sudden Cardiac Arrest in Late Adolescent with WPW Associated to Anabolic-Androgenic Steroid Abuse**
    Maher Bazzi, M.D., Bassem Chehab, M.D.
59. **Diffuse Bone Pain after Lung Transplant: Cystic Fibrosis Arthropathy or Voriconazole Induced Periostitis Deformans**
    Amna Batool, MBBS, Megan Krause, M.D.

Rapid Progressive Dementia with Probable Sporadic Creutzfeldt-Jakob Disease

Sinan Khayyat, M.D., Daly Al-Hadeethi, M.D., Mohinder Vindyhal, M.D., M.Ed.
KU School of Medicine-Wichita

[Judges Award: Best Poster]

Introduction

Prion diseases are rare neurodegenerative diseases which usually have long incubation periods and rapidly worsening neurological outcomes. Creutzfeldt-Jakob Disease (CJD) is a rare human prion disease with progressive, fatal encephalopathy characterized by dementia, cerebellar ataxia, and visual disturbances. CJD is caused by an abnormal protease-resistant isoform of prion protein (PrPSc), a misfolded version of the normal cellular isoform (PrPC). It is a rare disease with an incidence rate of approximately one case per million population per year with a worldwide distribution. We present an elderly male with Creutzfeldt-Jakob Disease (CJD).

Case Description

An 85-year-old man presented with confusion, aphasia, and episodic seizure-like activity in his right upper extremity and right eye twitching for two weeks duration. Neurological exam revealed stupor, aphasia, verbal non-responsiveness, generalized weakness, and lower extremity myoclonic movements. Pupils were constricted but reactive to light with no facial asymmetry. Cogwheel rigidity was noticed with only equivocal left Babinski. Workup for confusion showed generalized cerebral atrophy and chronic microvascular changes with scattered focal and confluent areas of nonspecific T2 bright signal in the periventricular and subcortical white matter but no acute events on MRI. EEG showed diffuse slowing with transient generalized epileptiform discharges for which the patient was started on anti-seizure medication. Lumbar puncture ruled out meningitis/encephalitis. CSF studies were negative for infectious and parasitic etiology. The CSF studies were also sent for 14-3-3 antigen test, which was elevated. Tau protein level was >4000 and Real-Time Quaking-Induced Conversion assay was also positive. The clinical diagnosis of CJD was given. The patient was discharged to hospice and passed away soon after that.

Conclusions

The definitive diagnosis of CJD is made with a brain tissue biopsy and western blot confirmation of protease-resistant PrP and presence of scrapie-associated fibrils. Criteria for the diagnosis of probable sporadic CJD in patients presenting with rapid progressive dementia are two of the following: myoclonus, visual or cerebellar signs, pyramidal/extrapyramidal signs, akinetic mutism, and a positive result of either a typical EEG with periodic sharp wave complexes and/or a positive 14-3-3 cerebrospinal fluid (CSF) assay. Our patient met criteria of probable sporadic CJD with the presence of progressive dementia, akinetic mutism, myoclonus, and an extrapyramidal sign of rigidity with positive 14-3-3 CSF assay. Our patient also tested positive on the Real Time Quaking-Induced Conversion assay which has high sensitivity and specificity for CJD disease. Prognosis with CJD has a rapid, deteriorating course invariably, with death generally within two years of symptom onset. No treatment has been proposed so far, and few medications help with symptoms. Early diagnosis also allows patients and their families to prepare for the expected disease course.

When it isn't Sepsis: A Case Report of Hemophagocytic Lymphohistiocytosis

Kelsy Witherspoon, M.D., Mohinder R. Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

[Chien Liu Award for Best Infectious Disease Poster]

Introduction

Hemophagocytic lymphohistiocytosis (HLH), a rare syndrome of immune dysregulation, can lead to clinical signs and symptoms of severe systemic inflammation, often mimicking more common presentations of illnesses. While primary HLH is an inherited disorder seen in childhood, secondary HLH presents in adults and results from infections, connective tissue disorders, or malignancies. HLH has a high mortality rate without rapid identification and intervention. Here, we present a case of secondary HLH in an elderly African-American male patient.

Case Presentation

A 69-year-old African-American man with a history of insulin-dependent diabetes, coronary artery disease, hypertension, chronic iron deficiency anemia, chronic heart failure, untreated Hepatitis C, and compensated cirrhosis presented with a three-month history of non-specific right upper quadrant and epigastric abdominal pain with diarrhea and a 20-pound weight loss. Admission labs were notable for leukopenia (3.0 K/cmm) and elevated lipase (1096; standard range 73-393 units). CT Abdomen/Pelvis showed non-specific findings of fluid and air-filled colon, suggestive of possible enteritis. GI Panel was negative for infectious diarrheal causes. The patient was soon discharged after being provided supportive care for suspected pancreatitis and viral enteritis. The patient returned after a week with acute onset high fevers (38.3 C - 39.4 C). Admission labs during the repeated admission showed pancytopenia with Hgb at 7.9 g/dL, a platelet count of 140 K/cmm, and severe neutropenia (ANC 322). Additional workup revealed the presence of hemophagocytosis with pancytopenia on peripheral smear and bone marrow biopsy. Follow-up labs also showed elevated ferritin of 13,869 ug/L and low fibrinogen 1.19 g/L in the face of persistent fevers (> 38.50 C). He was diagnosed with HLH based on the criteria and started on high dose dexamethasone and etoposide by Oncology.

Conclusions

A definitive diagnosis of HLH remains challenging, largely due to its variable clinical presentations and relative infrequency. The diagnosis is often delayed or mistaken for other etiologies. HLH can be diagnosed when patients have a gene mutation consistent with familial HLH or have at least five out of eight diagnostic criteria, as was the case with this patient.

Not Nearly Enough Kleenex: A Life Threatening Vascular Tumor Complication

Ryan Morse, Jesse Richards, D.O., Matthew Swan, M.D.
KU School of Medicine-Kansas City

Introduction

Kasabach-Merritt syndrome (KMS) is a rare disease that occurs when a vascular tumor causes a consumptive coagulopathy. The tumor traps platelets and clotting factors, leading to thrombocytopenia and a high risk of bleeding that may prove to be fatal. The rarity of this disorder creates a difficult clinical diagnosis, occurring in approximately 0.3% of hemangiomas.

Case Presentation

A 42-year-old female with a malignant hemangioendothelioma presented as a transfer with an uncontrolled nosebleed for the past five hours. The patient reported waking that morning from her sleep drenched in her own blood. EMS took her to the local hospital where her Hgb was recorded 5.7, platelets were 6,000, and a normal INR and fibrinogen. After bilateral nasal packing followed with multiple units of packed RBCs and platelets, hemostasis was unsuccessful and transfer to a tertiary institution was initiated. Upon arrival, she was seen by ENT and noted to be in severe distress with a 400-mL emesis of frank blood. Nares were packed bilaterally and large clots were observed on oral exam with blood running down the posterior oropharynx. Fluids, RBCs, cryoprecipitate, and platelets were given in an attempt to achieve hemostasis. Previous nasal packing was removed and replaced, without cessation of bleeding, thus insertion of an inflatable nasal balloon catheter was attempted next. The patient continued to bleed profusely with recurrent hemoptysis. An immediate rapid response was called and the patient was transferred to the ICU, where she was promptly intubated for airway protection.

With multiple failed packing attempts to cease bleeding, the patient was taken emergently to interventional radiology. Angiogram was performed with embolization of bilateral sphenopalatine arteries and control of bleeding was finally achieved. Imaging highlighted significant vascular tumor metastases in the posterior pharynx and skull base. With arterial embolization, the patient stabilized and was extubated a few days later. Nasal packing was removed and with the cessation of bleeding being complete, the patient was discharged.

Conclusions

A patient in the emergency room with epistaxis is not unusual, however, the differential for epistaxis can be quite broad and not always as straightforward as expected. This case highlighted a patient with a biopsy-proven malignant hemangioendothelioma, presenting with a rare syndrome. KMS occurs when the tumor consumes and actives platelets within blood vessels, causing microvasculature thrombosis and bleeding. In addition, the small vessel thrombi shear red blood cells and cause end-organ damage. Seen commonly in 60% of the population, uncomplicated epistaxis normally resolves with conservative measures. Refractory bleeding should heighten clinical suspicion that a common nosebleed could in fact be the presentation of a systemic bleeding diathesis. When KMS occurs, patients are at very high risk for bleeding, hemorrhagic shock, and death. Prompt evaluation with urgent intervention is life-saving.

Sepsis-Induced Radiation Recall Dermatitis after Chemotherapy with Irinotecan and Bevacizumab

John Frey, D.O., Christopher Streiler, M.D., Matthew Swan, M.D.
KU School of Medicine-Kansas City

Introduction

Radiation recall dermatitis (RRD) is an inflammatory reaction that occurs at previously irradiated sites. It is a rare phenomenon that typically occurs after the administration of certain drugs, days to years after radiation therapy. We report a case of sepsis-induced RRD in a patient receiving Irinotecan and Bevacizumab.

Case Presentation

A 35-year-old female was diagnosed with metastatic rectal adenocarcinoma and was initially treated with Capecitabine and concurrent radiation to the abdomen, pelvis, and lower extremities, for a total dose of 50.4 Gy. Due to progression of her disease she was treated with FOLFOX and Bevacizumab, followed by Capecitabine and Bevacizumab, then Irinotecan and Bevacizumab.

Seven months after initiating chemotherapy with Irinotecan and Bevacizumab, the patient presented to the hospital for a one-day history of fever, headache, back pain, abdominal pain, painful rash, and diarrhea following an epidural steroid injection the week before. Vital signs were significant for fever and tachycardia. The patient's exam was remarkable for photophobia, tenderness to palpation in the suprapubic region, as well as the thoracic and lumbar spine. Furthermore, there were maculopapular erythematous blanching patchy lesions located on the anterior aspect of both lower extremities from the hip to knee. Labs were remarkable for leukocytosis and lactic acidosis. The patient was admitted for sepsis and empirically started on Vancomycin and Piperacillin/Tazobactam. Given her symptoms, there was concern for development of spinal abscess and/or meningitis. MRI of the T/L-spine was obtained which was negative for spinal abscess. A lumbar puncture was performed with studies suggestive of viral meningitis, cultures were negative. In the following days, her blood cultures were positive for Streptococcus agalactiae, and her stool sample was positive for Clostridium difficile. Her antibiotic regimen was changed to Cefazolin and oral Vancomycin.

While the patient's systemic symptoms began to improve with antibiotic administration, the erythema on the patient's legs continued to progress. It ultimately became sharply demarcated, and when prompted, the patient stated the erythema was localized to the area of her prior radiation treatments. Upon further questioning regarding her history of radiation dermatitis, she stated the pain was the exact same as when she received radiation. Given the nature and description of the lesions, the patient was diagnosed with RRD. Since she had received 14 cycles of Irinotecan and Bevacizumab without complication, and development of the dermatitis coincided with onset of her systemic symptoms, her RRD was determined to be secondary to sepsis.

Conclusions

While RRD is typically attributed to the administration of certain drugs, this case provides evidence that environmental factors may also trigger RRD. To our knowledge, this is the first report of sepsis as a trigger for RRD. Future work is needed to uncover the underlying pathophysiology of RRD.

Macrophage Recruitment during Liver Repair is Attenuated and Delayed by Hyaluronan Synthesis Inhibition

Ben Harstine, Jennifer M. McCracken, Ph.D., Lei Jiang, Ph.D., Michele T. Pritchard, Ph.D.
KU School of Medicine-Kansas City

Introduction

The liver possesses a remarkable capacity to repair following injury. The difference between regenerative and non-regenerative wound healing is orchestrated, in part, by the recruitment of macrophages (møs) and their sequential development of different wound-repair phenotypes. Hyaluronan (HA), an ECM component, is synthesized and degraded during repair of extrahepatic tissues. The role of HA in liver repair is unknown. The purpose of this study was to evaluate the role of hyaluronan in mø phenotypic change that occurred throughout liver wound repair.

Methods

Wild type mice fed either a standard diet or a hyaluronan synthesis inhibiting diet containing 4-methylumbelliferone (4-MU) were given one dose of carbon tetrachloride (CCl_4) to cause acute liver injury. Markers of injury, regenerative wound healing, mø phenotype, and ECM composition were evaluated.

Results

HA was induced by CCl_4-induced liver injury. 4-MU feeding did not alter CCl_4-induced liver injury or synthesis of the macrophage recruiting chemokine, Ccl2 (MCP1). However, injury-induced HA synthesis was inhibited in 4-MU fed mice relative to mice fed the control diet. Moreover, hepatocyte proliferation was suppressed 75% in 4-MU fed mice following injury and cyclin mRNA transcripts demonstrated a proliferative delay. After acute injury, mø recruitment, as assessed using QPCR for macrophage-specific markers, was elevated at 72h and 120h post-CCl_4 in mice fed the control diet, indicating recruitment of peripheral møs. However, these same parameters were suppressed in 4-MU fed mice. Furthermore, fibrosis resolution after chronic CCl_4 was delayed in 4-MU-fed mice.

Conclusions

HA is a prominent feature of regenerating livers and is crucial for mø recruitment during this process. HA is also crucial for promoting normal hepatocyte proliferation following injury, perhaps through the recruitment, phenotypic change, and subsequent activity of møs.

Once Bitten, Twice Shy: The Thick and Thin of Blood Parasites

Jamison Ryan, M.D., Lisa Clough, M.D.
KU School of Medicine-Kansas City

Introduction

Malaria is caused by blood parasites of the genus Plasmodium and is transmitted to humans by infected mosquitoes. Malaria is rarely seen in the U.S., but due to its severity, it should be routinely considered in febrile patients with travel to known endemic regions. Symptoms are often non-specific, which means a high degree of clinical suspicion should be maintained, particularly in patients with fever of unknown origin and an appropriate travel history.

Case Presentation

A 48-year-old female was transferred to our facility with a five-day history of fever and headache. She was originally from Zambia and had recently traveled to the Democratic Republic of Congo (DRC) and Liberia. She reported having a mosquito bite while in the DRC but no fevers at that time. She arrived in the U.S. two days prior to fever onset. The first day she noted fevers and headache. She presented to an outside hospital where a broad infectious workup was performed including tests for bacterial and viral pathogens as well as thick and thin smears for malaria. All tests were negative. She returned to the clinic three days later with similar symptoms and was admitted with fever and tachycardia. Infectious workup, including a second malaria smear, was again negative. She was subsequently transferred to our facility for additional workup. She denied other infectious symptoms including cough, nausea, vomiting, dysuria, hematuria, hematochezia, myalgias, arthralgias, fatigue, or night sweats. She stated the fevers and headache occurred daily. She denied new sexual contacts or drug use. She had no significant past medical history other than a remote history of malaria 25 years ago. Physical exam was remarkable for a febrile, uncomfortable appearing, middle-aged woman. Labs were notable for anemia, lactic acidosis, and elevated ESR/CRP. Blood and urine cultures and RVP were negative. A rapid antigen test was positive for P. falciparum. A third malaria smear confirmed the diagnosis and demonstrated 4.3% parasitemia. The patient was started on Atovaquone-Proguanil based on CDC recommendations. She responded rapidly to therapy and subsequent testing for the presence of parasite was negative.

Conclusions

The malaria antigen test can provide a rapid diagnosis and ensure the quick delivery of appropriate treatment. It is particularly useful for laboratory technicians who are not familiar with performing malaria smears. Thick and thin smears are useful for confirming the detection of the parasite, species identification, and quantification of parasite load. Negative smears can be seen early in the infection, particularly in non-immune individuals as symptoms can manifest at very low levels of parasitemia. Smears should be repeated every 12-24 hours for three sets before effectively ruling out malaria. Treatment should be based on identified species, geographic related resistance patterns, and clinical severity.

Lumbar Radiculopathy and Iliopsoas Abscess as Presenting Manifestations of Ruptured Appendicitis

David Rayan, Gillian Dryton, Anita Moudgal, B.A., M.Ed., Amanda Jobe, M.D., Radhika Gupta, MBBS, Bharata Lankachandra, M.D.
KU School of Medicine-Kansas City

Introduction

Appendicitis has been called 'the great mimicker' due to variability in clinical presentation and in orientation of the appendix itself. We report a case of a middle-aged male who had an unusual presentation of appendicitis resulting in a delay in diagnosis and further medical complications.

Case Presentation

Patient is a 58-year-old male with a history of chronic back pain and cecal tubulovillous adenoma who presented to the ED with chronic lower back pain with associated right-sided radicular symptoms for one month, fevers for one week, and areas of swelling in his right lower back and buttocks for two days. He had been following with a pain specialist for presumed lumbar radiculopathy. An MRI was eventually obtained which showed concern for an iliopsoas abscess, so he was directed to the ED for further evaluation. On presentation, a CT scan of the abdomen and pelvis and later an MRI of the pelvis were obtained. Imaging demonstrated a large right iliopsoas abscess with erosion into the right iliac crest and a right gluteal abscess, likely the result of a ruptured retrocecal appendix. The patient was started on Vancomycin and Zosyn, and General Surgery was consulted who recommended IR drainage. IR placed drains into both abscesses and cultures from the iliopsoas abscess were positive for Bacteroides fragilis and Fusobacterium nucleatum. He was treated with a long course of antibiotics. Due to prior history of tubulovillous adenoma, repeat colonoscopy was performed which demonstrated an ileocecal mass with pathology consistent with invasive adenocarcinoma. He later underwent right hemicolectomy with operative report notable for a cecal mass, large mesenteric lymph nodes, and thickened and dilated appendix with dense adhesions of the ileocecal junction and appendix to the retroperitoneum. Patient established with Oncology and is receiving chemotherapy with FOLFOX.

Conclusions

Classically, appendicitis is described as presenting with periumbilical pain radiating to the lower right quadrant. However, when the appendix is located retrocecal, this may not be the case. In retrocecal appendicitis, inflammation tends to evolve slowly and abdominal pain may be minimal due to coverage of the appendix by the cecum. When pain is present, it often presents in a posterolateral distribution, which increases the risk of complications such as a retroperitoneal abscess as seen in this case. Abscesses have been reported to occur in 3.8% of individuals with appendicitis. Diagnostically, ultrasound has been the first-line imaging modality for assessing appendicitis. However, CT is often required to assess complex cases. Studies have shown that compared to ultrasound, CT has improved sensitivity and specificity. Given the patient's unconventional presenting symptoms, analysis of imaging in line with physical examination findings was critical to establish the etiology of symptoms, diagnosis, and guide management.

Determination of Antimicrobial Prescribing Practices in an Integrated Health System Emergency Department

John Price, Sarah Sartain, M.D., Brooke Bitner, Pharm.D.
KU School of Medicine-Kansas City

Introduction

Healthcare improvement is constant. Communication gaps can exist between institutions and the communities served. Antibiotic stewardships play a key role in minimizing antibiotic usage and preventing resistance in hospitals and associated clinics by systematically reviewing patients' data. Antibiotic stewardships utilize interprofessional collaborative practices including professionals from medicine, pharmacy, nursing, social services, and clinical laboratory scientists to proactively identify potential problems. A tertiary care, integrated health system's Emergency Department (ED) wants to identify antimicrobials prescribed into the outpatient community. The primary goal is determining the number of oral antimicrobial prescribed to adult outpatients (18+) within a 5-14 day treatment window. Secondary goals include the diagnosis, and non-capsule/tablet antimicrobial prescriptions to non-adults, or outside the treatment window.

Methods

This quality improvement study is a pilot, prospective evaluation. Patients are identified via the report: "ED Recently Discharged and NOT admitted Patients" over 30 days in the summer of 2017. Other project inclusions are prescribed antimicrobial(s) and "discharged from ED". Exclusions to this project include: "admission to the hospital" and antimicrobial hypersensitivity.

Results

Total antimicrobials prescribed over thirty days is 653. Adults prescribed oral antimicrobials are 467 (15.6 daily). Patients are diagnosed with infections including: urinary tract, cellulitis, soft tissue injury, abscess, upper respiratory, dental caries, gastrointestinal, sexually transmitted, otitis, pneumonia, viral, pyelonephritis, tick-borne, fungal, Bell's Palsy, and sepsis. The number of non-adult, non-oral, and outside window treatment antimicrobials are 186 (6.2 daily).

Conclusions

With 67,000 patients annually, or 184 patients daily, treated in the ED, approximately 11.9% receive an antimicrobial treatment on discharge. These antimicrobials treat 16 different forms of infection. Limitations and potential future studies include seasonal variability, whether the patient obtained prescription, and complied with treatment.

Bone-Marrow Sarcoidosis in a Patient Presenting with Fever of Unknown Origin

Sanjay Parashar, Jesse Richards, D.O., Megan Krause, M.D.
KU School of Medicine-Kansas City

Introduction

Sarcoidosis is an idiopathic granulomatous disease commonly affecting the lungs and thoracic lymphatics. Bone marrow involvement is infrequent in sarcoidosis and may be associated with anemia and leukopenia. This case report is unique in that sarcoidosis was diagnosed from a bone marrow biopsy in the absence of other locations of organ involvement during the workup of a fever of unknown origin (FUO).

Case Presentation

A 57-year-old white male with a history of hyperlipidemia, cholecystectomy, lumbar fusion of L4-S1, and recurrent fevers of 102˚F for several months presented with fatigue, myalgia, weight loss, and chills. On admission, he had a leukocytosis of 14,000, elevated ESR of 116, elevated CRP of 21, elevated ferritin of 509, and low hemoglobin of 10.9. Blood cultures returned negative while a PET scan showed enlarged hypermetabolic periportal lymph nodes with concern of metastatic disease. Endoscopic ultrasound, fine needle aspirate, and flow cytometry of the lymph nodes found no obvious lesions and showed no evidence of malignancy. He was referred to orthopedic surgery due to recurrent back pain and his spine implant was removed due to concern of infection. Samples of the bone showed bone necrosis with granulation tissue, fibrosis, and mild chronic inflammation with negative cultures. The patient was discharged but returned less than a month later, presenting to the ED with chest pain. Troponin and EKG were negative, however, a chest x-ray showed bilateral opacifications with concerns of pneumonia and the patient was admitted. A CTA showed bilateral upper lobe alveolar opacities and bronchoscopy revealed no malignant cells or alveolar hemorrhage. A positive ANA was noted along with a positive PR-3, however, c-ANCA was negative and the PR-3 was trending downwards. Additionally, ACE and Vitamin D 1,25 levels were normal. The patient was discharged on multiple course of antibiotics that were unsuccessful in controlling his fever. A bone marrow biopsy, ordered due to persistent anemia and thrombocytopenia, revealed hypercellular bone marrow (80% for age) and several non-caseating granulomata. A diagnosis of sarcoidosis was made, and the patient was placed on prednisone 40 mg/day with improvement of symptoms and continual follow up.

Conclusions

Extrapulmonary bone sarcoidosis is a rare finding. Granuloma formation within bone marrow may cause the release of pro-inflammatory cytokines that may cause recurrent fevers, and, like in this case, present as a fever of unknown origin. Similarly, the bone marrow remodeling that may occur during sarcoidosis may manifest as myalgia and fatigue due to impaired hematopoiesis. Several studies have postulated that bone marrow involvement may be more likely in sarcoidosis patients who present with abnormal hematological parameters. While statistically low in likelihood, sarcoidosis should be considered in patients who have fever of unknown origin, especially once an exhaustive infectious and oncological work-up has taken place.

Babesiosis in an Immunocompromised Patient

Sonya Parashar, M.D., Rachel Weihe, M.D., Jessica Newman, D.O.
KU School of Medicine-Kansas City

Introduction

In the United States, Babesia microti's reservoirs include white-footed mice, vole, and deer. The vector is the Ixodes scapularis nymph and is commonly seen in spring and summer in the Northeast and upper Midwest United States. Risk factors for severe disease include: age > 50 years old, asplenia, HIV, malignancy, and immunosuppressive therapy exposure, namely with TNF-α inhibitors and exposure to rituximab. This case presents a compelling presentation of babesiosis and extended treatment course in an immunocompromised patient.

Case Presentation

A 62-year-old man with a history of chronic lymphocytic leukemia presented with a three-week history of daily episodes of 103˚F fever and abdominal pain. He previously completed five cycles of chemotherapy with bendamustine and rituximab. During his last cycle of chemotherapy, he developed increasing left upper quadrant pain, fatigue, and anorexia and had progressive splenomegaly. He underwent laparoscopic splenectomy and pathology showed extramedullary hematopoiesis with no malignancy. Following splenectomy, he developed fever with progressive leukopenia (WBC 2.4 with ANC 1800) and was treated with cefepime and vancomycin without improvement. Extensive infectious workup was unrevealing though ultrasound suggested possible cholecystitis and a percutaneous drain was placed with no growth from gallbladder fluid. Antibiotics were broadened to meropenem, tobramycin, and amphotericin B without defervescence. He developed septic shock and was transferred to our institution. A thorough exposure history was taken, noting he lived on a wooded property outside Lawrence, Kansas and had a lake home in Wisconsin (last visited nine months prior). He often trapped raccoons on his property. Laboratory revealed hemolytic anemia, with hemoglobin 6.1 grams, haptoglobin < 30, LDH 3871, total bilirubin of 9.7, and AST of 296. A peripheral blood smear revealed 50% parasitemia and Babesia microti PCR was later positive. The patient was treated with two exchange transfusions reducing parasitemia to less than 5% resulting in rapid improvement and was started on azithromycin and atovaquone for a total of six weeks with repeat negative smears. Three months after therapy, he had recurrent fever with low levels of parasitemia. This resolved after a six-month course of atovaquone-proguanil and azithromycin.

Conclusions

This patient's advanced age, asplenia, malignancy, and treatment with bendamustine and rituximab were risk factors for severe babesiosis and contributed to his extended treatment course. Studies have recently discovered that ongoing antimicrobial exposure in relapsed cases result in acquired resistance to both atovaquone and azithromycin. Case reports have shown success with achieving clinical cure with the use of atovaquone-proguanil and azithromycin over a six-month period in such cases of acquired resistance, which proved to be effective for this patient. Though raccoons may carry babesia, the PCR for babesia microti suggested he was more likely exposed in Wisconsin with disease resting in a dormant stage until his splenectomy.

BMP Signaling Regulates Support Cell Proliferation in the Regenerating Zebrafish Neuromast

Madeline St. Peter, Linjia Jiang, Ph.D., Tatjana Piotrowski, Ph.D.
KU School of Medicine-Kansas City

Introduction

Deafness is a debilitating condition that is often caused by sensory hair cell (HC) death in the inner ear. Mammals do not have the capacity to regenerate HCs, therefore, cell death due to noise exposure, aging, chemotherapeutic drugs, or antibiotics can result in permanent hearing loss. In contrast, non-mammalian vertebrates such as the zebrafish can robustly regenerate HCs after injury. Zebrafish HCs are located inside neuromasts, which are small, circular organs that span both sides of the fish. HCs in the neuromast are surrounded by multiple sub-populations of support cells (SCs), each with different proliferative capacities. Following HC death, some will divide and differentiate into new HCs, some will self-renew to replenish the SC pool, and some will remain quiescent. While the molecular cues responsible for the differentiation of SCs into HCs start to be well understood, the genetic mechanisms responsible for SC self-renewal remain unexplored. In zebrafish, the bone morphogenic protein (BMP) pathway has been implicated as a key regulator of cell proliferation during both embryonic development and regeneration of some mature tissue types. We thus hypothesize that BMP signaling regulates SC proliferation in the neuromast both in homeostasis and during regeneration.

Methods

In order to test our hypothesis, we overexpress a BMP ligand using a transgenic heat shock line (tg:hsbmp2b) and show that activation of the BMP pathway thwarts SC self-renewal following drug-induced HC death.

Conclusions

Using an in situ hybridization screen, we are currently identifying targets of the BMP pathway to better understand how BMP signaling controls SC proliferation in the neuromast.

Acquired Factor VIII Inhibitor Related Retropharyngeal Hematoma: A Novel and Rare Differential for Acute Dysphagia and Odynophagia

Ryan Zwick, Jesse Richards, D.O., Ben Fangman, M.D., Abebe Abebe, M.D.
KU School of Medicine-Kansas City

Introduction

Retropharyngeal hematoma is a serious complication of acquired factor VIII inhibitor that has not been previously described in literature. Due to the possibility of acute decompensation in these patients due to airway compromise, timely diagnosis and treatment is vital.

Case Presentation

A 56-year-old man with past medical history of spinocerebellar ataxia presented to the Emergency Department with odynophagia, dyspnea, and facial swelling. He first noticed a sore throat two days prior and decided to come to the ED when he could no longer swallow liquids. In the ED, diffuse ecchymosis across his upper extremities was noted. Initial labs revealed elevated WBC (18.8 K/µL), decreased hemoglobin (7.5 gm/dL), normal INR 1.1, but prolonged aPTT that did not correct with a 1:1 mixing study. Due to concern for an impending upper airway compromise, direct laryngoscopy was performed which revealed a large posterior pharyngeal wall mass with 80% obstruction of the larynx. Urgent CT revealed odontogenic infection and soft tissue thickening in the pharynx and larynx likely secondary to infection rather than tumor or hemorrhage. Broad spectrum antibiotics were started, and he was admitted to the Medical Intensive Care Unit for airway monitoring.

On arrival to the MICU, repeat laryngoscopy was interpreted as pharyngeal hematoma. Coagulation factor assay was performed which revealed decreased factor VIII. Mixing study did not correct aPTT that was compatible with an acquired factor VIII inhibitor. Based on this finding, he was started on prednisolone and cyclophosphamide. Gradual improvement in aPTT followed with eventual resolution of the hematoma. He was then discharged with follow up.

Conclusions

This case serves to remind that a retropharyngeal hematoma is an important consideration in the differential diagnosis of acute odynophagia and dysphagia. Although rare in incidence, a retropharyngeal hematoma can compromise the airway making its recognition and management urgent. This case also introduces acute odynophagia and dysphagia as presenting symptoms of acquired factor VIII inhibitor. Acquired factor VIII inhibitor is a rare disorder that typically presents with catastrophic bleeding episodes later in life. Unlike congenital factor VIII inhibitor which commonly presents with hemarthrosis, acquired factor VIII inhibitor presents with purpura or soft tissue bleeding including retropharyngeal hematoma. Due to its diagnostic challenge, acquired factor VIII inhibitor carries significant morbidity and mortality (mortality rate up to 22%).

Risk of Lead Dislodgement with Catheter Ablation in Patients with Recently Implanted Cardiac Implantable Electronic Device

Brendan Cokingtin, Mohammad-Ali Jazayeri, M.D., Kevin Cokingtin, Venkat Vuddanda, M.D, Courtney Jeffery, APRN-C, Donita Atkins, BSN
KU School of Medicine-Kansas City

Introduction

Patients with cardiac implantable electronic devices (CIEDs) frequently have arrhythmias warranting catheter ablation (CA). The risk of lead dislodgement with CA in patients with CIED implant within one year is uncertain. We retrospectively evaluated patients undergoing CA of arrhythmias at an academic medical center with CIED implant, lead revision, or upgrade within the preceding year.

Methods

We assessed rates of CIED-related mechanical complications and infection over six-months follow-up. Pre-/post-ablation device interrogations were reviewed for interval changes and parameters compared with the Wilcoxon Signed-Rank test.

Results

Among 1810 CIED patients undergoing CA from 2012-17, 170 had CA within 12 months of CIED implant, lead revision, or upgrade. Median age was 69 years, and 68% were men. Implantable cardioverter defibrillators and cardiac resynchronization therapy devices were each present in nearly 25% of patients. Median time to CA was 138 days; 50 (29%) and 12 (7%) had CA within 90 and 30 days of CIED procedures, respectively. One atrial lead required revision after dislodgement during atrial fibrillation CA 190 days after CIED upgrade. No CIED infections, other mechanical complications, or clinically significant changes in device function were observed.

Conclusions

CA of arrhythmias in patients with CIEDs appears safe with a very low risk of lead dislodgement, even in those with device implant/upgrade 90 days or less pre-ablation.

Nelson's Syndrome:
A Rare Treatment-Related Complication of Refractory Cushing's Syndrome

Anam Abbasi, M.D., Sachin Srinivasan, M.D., Georges Elhomsy, M.D.
KU School of Medicine-Wichita

Introduction

Nelson's syndrome is a rare neuroendocrine condition described as a complication of bilateral adrenalectomies undertaken for refractory Cushing's syndrome. It is seen in about 8-47% of patients after the procedure. Nelson's syndrome refers to a spectrum of symptoms and signs arising from an adrenocorticotropic (ACTH)-secreting pituitary macroadenoma after therapeutic bilateral adrenalectomy. The observed spectrum of clinical features relates to the local effects of the tumor on surrounding structures, the secondary loss of other pituitary hormones, and the effects of the high serum concentrations of ACTH on the skin.

Case Presentation

A 25-year-old female with a past medical history of hypertension, hyperlipidemia, and type 2 diabetes presented with 150-pound weight gain in two years associated with fatigue, increased appetite, polyuria, polydipsia, constipation, amenorrhea, decreased libido, constipation, bruising, headache, and somnolence. She was obese and had hirsutism, severe acne, acanthosis nigricans, abdominal stretch marks without characteristic purple striation, and elevated BP. Labs showed a normal thyroid function test, IGF1, and prolactin, elevated morning cortisol level, and a positive dexamethasone suppression test. 24-hour free urinary cortisol was high. Elevated corticotropin, midnight salivary cortisol, and ACTH confirmed Cushing's syndrome. Pituitary MRI revealed a hypo-intense adenoma on the left side of sella, close to the left internal carotid artery. She was recommended for resection of a pituitary adenoma but deferred it to preserve fertility. CT chest, abdomen, and pelvis ruled out sources of ectopic ACTH production. Treatment proceeded with bilateral adrenalectomy for persistent Cushing's syndrome followed by supplementation with fludrocortisone, dexamethasone, and tapering prednisone. At one-month post-adrenalectomy, she had lost 40 pounds but reported persistent headaches, fatigue, polyuria, and polydipsia. Her ACTH level remained elevated. MRI brain showed an interval increase in size of the pituitary adenoma without mass effect on optic chiasm. This macroadenoma combined with her symptoms and an even higher post-surgical ACTH favored the diagnosis of Nelson's syndrome. She underwent gamma-knife radiation therapy for the pituitary adenoma without any complications.

Conclusions

Predictive risk factors of Nelson's syndrome include existing pituitary macro-adenomas, lack of prophylactic radiation at the time of adrenalectomy, duration of Cushing's disease, rapid rise of ACTH in the first year after surgery, and high urinary cortisol levels. Pathophysiology involves uninhibited corticotropin releasing activity leading to increased ACTH, propiomelanocotrin, and by-products. This leads to increased ACTH, tumor progression with compression on cranial nerves optic pathways leading to hyperpigmentation, oculomotor nerve palsies, hemianopsia, and possibly blindness. Treatment options include observation, surgery, radiation, and/or pharmacotherapy. Given the significant morbidity associated with this condition, prompt detection and treatment are necessary to prevent further devastating complications.

Transgender Health Medical Education Intervention and its Effects on Beliefs, Attitudes, Comfort, and Knowledge

Joseph Cherabie, M.D., Kari Nilsen, Ph.D., Sarah Houssayni, M.D.
KU School of Medicine-Wichita

Introduction

Transgender health disparities have been well documented in the literature in recent years, as have the lack of transgender health issues in medical education programs across the country. Currently, there is little to no transgender health medical education within the University of Kansas School of Medicine-Wichita.

Methods

A prospective study was conducted with an hour-long didactic lecture on transgender health being given to faculty, medical students, and residents at the University of Kansas School of Medicine-Wichita. The didactic lecture included educational information and presentations by transgender persons. A pre-intervention and post-intervention survey was given to assess attitudes, comfort level, knowledge, and beliefs regarding the treatment of transgendered persons and associated health concerns, as well as a 90-day post-intervention survey assessing the same variables. The question of what attendees planned to do differently as a result of the intervention was asked.

Results

The intervention provided a significant positive increase in attitudes, comfort levels, and knowledge with respect to transgender health issues between the pre- and post-intervention surveys, however, did not provide a significant positive increase in beliefs on transgender health issues. There was no significant change in attitude, comfort levels, knowledge, or beliefs from the post-survey after 90 days. Four categories of what attendees planned to do differently as a result of the intervention also were identified.

Conclusions

A didactic lecture on transgender health issues can positively change attitudes, comfort levels, and knowledge on transgender health issues significantly with the changes sustaining after 90 days. Beliefs tend to be much harder to change.

Effective Management of Obesity with Generic Combination of Phentermine/Topiramate vs Qsymia®: Is it Possible?

Wajeeha Rizvi, M.D., Nicholas Ojile, M.D., Milan Bimali, Ph.D., K James Kallail, Ph.D., Georges Elhomsy, M.D.
KU School of Medicine-Wichita

Introduction

Obesity is a significant health concern in the developed world, with nearly 40% of the worldwide population estimated to be overweight and 13% meeting World Health Organization criteria for obesity in 2016. Medical management is needed in patients who are unable to attain ideal weight loss with conservative methods and lifestyle modification. In this study, we assessed if the less-expensive generic combination of phentermine and topiramate is an effective alternative weight loss medication.

Methods

A retrospective, cross sectional, observational study was conducted on all eligible patients at a university endocrinology clinic to compare the weight changes among users of Qsymia® versus those who took a combination of generic phentermine and topiramate. Medical records were abstracted for relevant data including weight, vital signs, and relevant lab data.

Results

Seventy-three eligible patients were analyzed (17 who took Qsymia® and 56 who took phentermine/topiramate). The number of weight measurements recorded per patient ranged from 1 to 10. There were no statistically significant differences in age, sex, and race/ethnicity. The distribution of A1c, LDL, HDL, TSH, T4, and duration of drug use were comparable across the two groups. Generalized estimation equation-based modeling estimates revealed no significant difference in weight measurements between the two groups in the unadjusted as well as each of the three adjusted models. Modeling on median imputed data revealed similar findings.

Conclusions

A generic combination of phentermine and topiramate provided similar weight loss results as the more expensive alternative, Qsymia®. This observational study warrants the need for a larger study to verify that the less expensive medication can be provided for weight loss.

Polyarticular Calcium Pyrophosphate Deposition Disease Masquerading as Pseudoseptic Arthritis

Sarah El Chami, M.D., Timothy Shaver, M.D., Paul Ndunda, M.D.
KU School of Medicine-Wichita

Introduction

Calcium pyrophosphate dihydrate (CPPD) disease is the second most common crystal-induced arthropathy. The acute form of the disorder, pseudogout, occurs in one fourth of patients with CPPD. Pseudogout has a heterogenous presentation that can mimic other causes of severe joint inflammation including rheumatoid arthritis, osteoarthritis, and septic arthritis. Our case of polyarticular pseudogout presented as pseudo-septic arthritis.

Case Presentation

A 64-year-old male with a medical history of recurrent mantle cell lymphoma, COPD, and type 2 diabetes presented for acute left shoulder pain. Six weeks prior, the patient was admitted for right wrist pain and swelling. Irrigation and drainage of the right wrist revealed purulent synovial fluid; gram stain showed many WBCs with no organisms; and there was no growth on synovial fluid or blood cultures. The patient was diagnosed with presumed septic arthritis and was discharged on a six-week course of IV antibiotics.

After taking antibiotics, the patient presented with left shoulder pain after lifting a heavy object. The patient was started empirically on IV antibiotics for possible septic arthritis. Vital signs were T 37.3^{o} C, HR 113 bpm, and BP 144/86 mmHg. Examination of the left shoulder revealed anterior tenderness with limited abduction and flexion. White blood count was 13,600 cells/µl with 83% neutrophils and ESR and CRP were elevated at 44 mm/hr and 22.9 mg/l, respectively. Aspiration of the left shoulder revealed 71,000 RBCs, 18,540 WBCs, and calcium pyrophosphate crystals. Gram stain showed no organisms and viral, bacterial, and fungal cultures revealed no growth. Later, the patient developed left wrist and right ankle pain and swelling. Examination revealed bilateral diminished range of motion with extensive synovitis/ tenosynovitis, warmth, and tenderness. Synovitis was present in right and left metacarpo-phalangeal diffusely, with tenderness over right 2nd-5th metacarpophalangeal, 2nd-3rd proximal interphalangeal joints, left 2nd-5th metacarpophalangeal joints, and 2nd-4th proximal phalangeal joints. Feet were negative for synovitis but positive for tenderness in bilateral 2nd-3rd metatarsophalangeal joints. Aspirate of the left wrist was dry and radiographs demonstrated radiocarpal narrowing with chondrocalcinosis in the triangular fibrocartilage distribution. The patient was diagnosed with pseudogout. Antibiotics were discontinued. The patient was started on colchicine and steroids, which led to immediate and dramatic improvement.

Conclusions

Pseudoseptic arthritis describes a clinically expressive septic joint with purulent aspirate when no microorganisms can be isolated. It can be caused by several inflammatory conditions and is indistinguishable from true septic arthritis. Fourteen percent of patients diagnosed with septic arthritis that had negative bacteriological results subsequently developed rheumatic disease. When no microorganism is identified, the diagnosis of septic arthritis should remain presumptive and further evaluation should screen for other diseases, especially those of rheumatologic origin.

Elevated Procalcitonin with Atypical Presentation

Rachel Sigler, D.O., Kassem Hammoud, M.D.
KU School of Medicine-Kansas City

Introduction

Procalcitonin is a marker of bacterial infection, commonly used to distinguish community acquired pneumonia due to bacterial causes versus viral etiologies. We report a case of elevated procalcitonin in a patient with autoimmune disease and anakinra withdrawal.

Case Presentation

A 22-year-old male with systemic juvenile idiopathic rheumatoid arthritis presented to an outside hospital for evaluation of muscle and joint pain. He was treated with anakinra previously, but ran out of his medications five days prior. At the outside hospital, he reported a cough and had chest radiograph with concern for left lower lobe pneumonia. He was started on levofloxacin and transferred to KU for rheumatology evaluation. At the time of his presentation, his WBC count was 32000, with 13% bands, and a procalcitonin of 5. A CT chest demonstrated a small subpleural consolidation. Rheumatology started plaquenil and prednisone 5 mg for one week. Patient spiked a fever of 100.4 on hospital day six and ID was consulted. Blood cultures at outside hospital were negative, as were cultures obtained on admission. Vancomycin was started as patient continued to have nightly febrile episodes. On hospital day 10, procalcitonin was 28 and continued to rise over the next two days, peaking at 42 in spite of broad-spectrum antimicrobial coverage with linezolid, meropenem, and liposomal amphotericin B. Extensive infectious work-up was negative. Patient underwent bronchoscopy, which revealed rhinovirus infection. Anakinra was resumed and his antimicrobials were stopped. The procalcitonin and his WBC count rapidly decreased and his fever resolved by the next day.

Conclusions

Procalcitonin is produced by neuroendocrine cells in the thyroid and lungs, most of which is converted to calcitonin. It is encoded by gene CLAC-1 on chromosome 11. Bacterial infections increase CALC-1 expression on parenchymal tissue, which leads to procalcitonin that is not converted to calcitonin peripherally. Viral infections have less effect on procalcitonin levels due to interferon gamma expression. Infections associated with TNF alpha release, such as gram-negative infections and malaria, have the highest procalcitonin levels. Infections that trigger different inflammatory pathways, such as Lyme disease and mycobacterial infections, do not increase procalcitonin levels. In this case, procalcitonin was falsely elevated, likely due to anakinra withdrawal. His symptoms started within days of stopping the medication, which is not typical for autoimmune disease flair. Although procalcitonin has shown clinical utility in distinguishing bacterial infections from viral, it is important to remember its limitations in the hospital setting. Procalcitonin can be helpful in cases of lower respiratory tract infections and deciding when to de-escalate or discontinue antibiotics.

Polymicrobial Endocarditis

Joseph Borick, M.D., Thomas Moore, M.D.
KU School of Medicine-Wichita

Introduction

Polymicrobial endocarditis (PE) is becoming more common and can occur in 1-7% of patients with endocarditis. It disproportionately effects intravenous drug users (IVDU) and is more likely to have a fatal outcome. The patient's history can provide essential clues to guide the provider in decision making. Analysis of the choice of illicit drug and method of injection can hint to the most likely organism and may necessitate fungal cultures.

Case Presentation

A 28-year-old female with history significant for IVDU and pregnancy (24 weeks) presented with three weeks of hip pain followed by progressive leg weakness, shortness of air, hemoptysis, and fevers. She used methamphetamine, hydromorphone and marijuana. When she used IV drugs, she often used tap water for dilution and shared needles.

Hospital labs were significant for anemia, thrombocytopenia and leukocytosis. A CTA chest was performed and showed innumerable thick walled cavitary lesions consistent with septic emboli. She was admitted to the ICU and started on Vancomycin and Piperacillin-Tazobactam. Blood cultures were drawn and were positive in 2/2 vials for methicillin-sensitive Staphylococcus aureus. Piperacillin-Tazobactam was discontinued and Vancomycin was changed to Nafcillin. ECHO was significant for tricuspid valve vegetation. Fungal cultures eventually grew Aspergillus fumigatus. She was started on Amphotericin and switched to Voriconazole after risks to infant were discussed. Repeat fungal cultures on hospital day 10 also were positive for Aspergillus fumigatus. On hospital day 11, she went into pulseless electrical activity and a perimortem c-section was performed at bedside. Her daughter was delivered and transferred to the NICU. There was return of spontaneous circulation but she was made comfort care after MRI showed severe hypoxic ischemic injury and she passed away on hospital day 18. Three months later, her daughter was discharged from the NICU with her grandmother.

Conclusions

While Staphylococcus aureus is most common organism in IVDU, oral flora is associated with patients who lick their needles/skin. Candida is associated with patients who use brown heroin dissolved in lemon juice. Pseudomonas is associated with using pentazocine-tripelennamine and Aspergillus is associated with dissolving drugs in tap water. Aspergillus is found in 6-55% of water samples. Of the above, Aspergillus is unique as blood cultures are rarely positive, necessitating fungal cultures. Delayed or mistaken diagnosis and extra cardiac manifestations occur in the majority of cases of endocarditis with Aspergillus. Diagnosis of Aspergillus endocarditis requires a high clinical index of suspicion and mortality rates approach 80%. In the appropriate patients, fungal cultures should be obtained even when blood cultures are positive for other organisms, such as Staphylococcus aureus, as polymicrobial infections can occur. Our case manifests the necessity in obtaining fungal cultures in IVDU who utilize tap water in their drug use process.

Ice Induced Cardiac and Systemic Thrombus

John Eliveha, M.D., Shravani Vindhyal, M.D., Mohinder Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

Introduction

Methamphetamines (ICE) are sympathomimetic amines that increase synaptic concentration of serotonin, noradrenaline, and dopamine, creating hallucinogenic, euphoric, and stimulant effects. Cardiovascular complications are the second leading cause of death in methamphetamine users. The cardiac complications of methamphetamines are hypothesized from a variety of mechanisms such as hypertension, tachycardia, and myocardial toxicity with cellular death, fibrosis, and contraction band necrosis. We present a case of ventricular thrombus with systemic emboli in a patient with dilated cardiomyopathy after methamphetamine use.

Case Presentation

A 24-year-old female with history of AIDS, non-ischemic cardiomyopathy, and polysubstance abuse presented with acute onset lower extremity pain. The patient had snorted methamphetamine overnight. She woke up with severe lower extremity pain and inability to move her lower limbs. Her vital signs showed tachycardia, tachypnea, and hypotension. Examination was positive for tender lower extremities with no palpable dorsalis pedis, posterior tibial, and popliteal pulses bilaterally. Initial lab tests were significant for lactic acidosis, acute kidney injury, non-specific ST-T wave changes, elevated troponin, and urine drug screen positive for methamphetamine and marijuana. Arterial and venous duplex of the lower extremities revealed no blood flow. CT angiogram showed large segment aortic occlusion (4 cm) just beyond the renal arteries. Transthoracic echocardiogram showed an echo dense mass in the left ventricle with defined margins that were distinct from the endocardium and seen throughout systole and diastole showing left ventricular thrombus.

Conclusions

Factors causing ventricular and systemic thrombus in patients following methamphetamine abuse involve the size of myocardial infarct and extent of systolic dysfunction. Usually patients with decreased ejection fraction are at a higher risk for thrombus after unrecognized myocardial infarction. The formation of thrombus occurs within 24 hours post myocardial infarction. With the increasing prevalence of methamphetamines use, primary care physicians should be aware of complications such as cardiac and systemic thrombus.

Non-Atherosclerotic Coronary Angina

Shravani Vindhyal, M.D., John Eliveha, M.D., V. Subbarao Boppana, M.D., Mohinder Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

Introduction

Coronary artery fistula (CAF) is an abnormal communication between an epicardial coronary artery and a cardiac chamber or a major vessel (vena cava, subpulmonary veins, pulmonary artery, mediastinal vessels, or coronary sinus) bypassing the capillary bed. CAFs are mostly solitary but at times there could be multiple micro fistulas. CAF incidence is 0.2% to 1.2%, but the incidence of CAFs is on the rise over the last decade due to the wide use of echocardiography and angiography. CAFs are reported to arise more from the right coronary system than the left, and more than 90% drain into the venous circulation. We present a case of CAF in a male presenting with chest pain.

Case Presentation

A 40-year-old Chinese descent male presented to the ER complaining of chest pain. The pain was characterized as heavy, retrosternal, and non-radiating associated with dizziness, nausea, and palpitations. The pain started while lifting a heavy object in the kitchen and was relieved gradually with rest. Similar episodes of chest pain for the past several years limited his physical activity but he did not seek medical attention due to lack of medical insurance. Physical exam was positive for a continuous grade II/VI murmur at left sternal border, otherwise, lungs were clear, jugular venous pressure was normal, and no pedal edema was noted. An exercise stress test was terminated prematurely at seven minutes as he developed dizziness, ST-segment depression in the lateral leads, non-sustained monomorphic ventricular tachycardia, and a transient left bundle branch block.

Cardiac catheterization showed insignificant coronary artery disease but revealed a fistula leading from the left coronary artery near the bifurcation to the main pulmonary artery. Attempts to coil the fistula via the pulmonary circulation proved unsuccessful. The fistula was isolated and ligated close to the left anterior descending artery. Transesophageal echocardiography was used to confirm the flow to the pulmonary artery before and after ligation. This resulted in patient recovery without any complications and he remained asymptomatic.

Conclusions

Patients with CAF often remain asymptomatic. Development of symptoms depends on the severity of the left to right shunt and the degree of volume overload produced by the fistula. Coronary angiography remains the gold standard for diagnosis. Transthoracic echocardiography combined with Doppler color flow imaging, transesophageal echocardiography, magnetic resonance imaging, and contrast-enhanced multi-detector tomography can be used as an adjunct to coronary angiography. The usual indications for treatment include the proximal location of the fistula, a single drain site, extra-anatomic termination of the fistula away from normal coronaries, older patient age, and absence of concomitant cardiac disorders requiring surgical intervention.

Miliary TB in a Patient with End-Stage Liver Disease

Victoria Poplin, M.D., Brent Harbaugh, M.D., Nathan C. Bahr, M.D.
KU School of Medicine-Kansas City

Introduction

Miliary tuberculosis (TB) is a rare form of TB that results from massive lymphohematogenous dissemination. We present a case of fatal military TB in a patient with end-stage liver disease (ESLD).

Case Presentation

A 52-year-old male with a history of ESLD due to alcohol abuse and type 2 diabetes mellitus presented to the emergency department due to multiple falls and confusion. He was recently discharged after having been admitted for pneumonia and culture-negative spontaneous bacterial peritonitis. On admission, a CT chest showed tiny nodular opacities in all five lobes with upper lung predominance and some more prominent patchy alveolar opacities at the lung apices and an irregular lytic lesion in the anterior and inferior aspects of the T9 vertebral body. He was also noted to have severe thrombocytopenia and altered mental status. The patient was admitted and placed in airborne isolation due to concern for possible TB. An interferon gamma release assay was obtained and later returned positive. Shortly into his hospital stay, he was started on empiric TB treatment with isoniazid, ethambutol, rifampin, and pyrazinamide. After discussion with outside TB specialists, isoniazid and pyrazinamide were discontinued and daily levofloxacin and three times weekly amikacin were started in setting of his known ESLD. A bone marrow biopsy showed multiple granulomas, but special stains were negative for acid fast and fungal organisms. Approximately three weeks into his stay, his peritoneal culture from previous hospitalization returned positive mycobacterium tuberculosis (MTB) complex and a few days later one sputum sample grew MTB complex as well. Lumbar puncture was not able to be obtained due to thrombocytopenia although his platelet count did eventually improve after TB therapy initiation. By this time, the diagnosis was confirmed. Unfortunately, he went on to develop acute hypoxic respiratory failure, worsening mental status and acute renal failure, which ultimately lead to a fatal arrhythmia.

Conclusions

In 2016, approximately 10.4 million people fell ill with TB. Miliary TB is a rare form of disseminated TB that affects up to 2% of individuals and carries up to a 33% of mortality. Its diagnosis is often delayed as presenting symptoms are non-specific and varies depending on the sites involved. The most common symptoms include fever, cough, weight loss, and anorexia. The diagnosis of miliary TB involves obtaining tissue/fluid of the sites involved and sending for AFB smear and culture. Treatment of military TB has yet to be standardized. Currently, the IDSA recommends six months duration for drug susceptibly miliary TB, however, this must be extending if there is bone, joint, or meningeal involvement. We suspected both bone and meningeal involvement in this patient. Therapy choice was further complicated by his liver disease, thrombocytopenia, and eventual renal failure.

Paenibacilli - Newly Identified Organism Concealed in an Old Genus

Tori Kunkel, Jessica Newman, D.O.
KU School of Medicine-Kansas City

Introduction

Paenibacillus species are gram-positive bacteria originally classified as Bacillus species until the establishment of a new genus in 1993. These organisms typically reside in soil and have been identified in diverse environments including honeybee colonies, ocean sediment, and fresh water. Organisms of the Paenibacillus genus were not known to cause human disease until recently, when scattered case reports of human infections, including bacteremia, osteomyelitis, and endocarditis caused by several species began appearing in medical literature.

Case Presentation

A 23-year-old female with a history of neurogenic bladder status post bladder stimulator placement and ulcerative colitis status post colectomy presented to the emergency department with fevers, chills, nausea, and discomfort in the left upper extremity at the site of a central line. The patient had recently been diagnosed with bacteremia secondary to a gram-positive rod at an outside institution. A peripherally inserted central catheter was placed, follow-up blood cultures were negative, and she was discharged on intravenous daptomycin 6 mg/kg/day. Final identification of the gram-positive rod was pending. On physical examination at our institution, the patient was afebrile at 37.6° C, with pulse of 90 bpm, and a normal blood pressure. A complete blood count revealed a WBC of 6.8 K/UL with 88% neutrophils. Blood cultures were obtained and the central line was removed. An ultrasound of the left upper extremity demonstrated a near-completely occlusive thrombus in the left axillary brachial and basilic veins, which was treated with enoxaparin. Finalized blood cultures from the previous hospital stay later identified Paenibacillus jamilae/polymyxa, and blood cultures at our hospital revealed the same isolate. The isolate was resistant to penicillin but susceptible to clindamycin, trimethoprim/ sulfamethoxazole, and vancomycin. An attempt to ascertain the source of infection could not be confirmed. The patient denied use of intravenous drugs or injection of a foreign substance into the venous access site but was concerned about potential exposure from soil in a horse arena she frequented. The bladder stimulator generator site was non-tender, and patient declined removal. Hospital treatment consisted of intravenous vancomycin and oral trimethoprim/ sulfamethoxazole. She was discharged on oral linezolid to complete a 14-day course of therapy.

Conclusions

Paenibacilli are a newly classified genus of bacteria that were originally categorized as Bacillus species. Paenibacilli have been implicated in human infection and have been isolated from urine, cerebrospinal fluid, and blood cultures. Paenibacillus polymyxa specifically was previously implicated as a causative agent for bacteremia in an elderly patient with cerebral infarction who weeded her garden daily. Current methods in clinical microbiology laboratories may not be sufficient for identification and more complex analysis may be required for species identification. Clinical isolates may be intermediate to penicillin G and quinolones but susceptible to trimethoprim/sulfamethoxazole, aminoglycosides, vancomycin and linezolid.

Characteristics of Smokers among Patients in a Hepatology Clinic

Daly Al-Hadeethi, M.D., Victor Ankoma-Say, M.D.
KU School of Medicine-Wichita

Introduction

The 2014 Report of the Surgeon General, The Health Consequences of Smoking-50 Years of Progress, marks an anniversary since the first Surgeon General's report on smoking and health. In the 1964 and 2004 Surgeon General's reports, cigarette smoking was associated with diseases affecting multiple organs of the body. The most recent report highlighted that the number of diseases and other adverse health effects caused by smoking and secondhand smoke have increased and that the disease risks are greater than presented in previous reports, including liver cancer and colorectal cancer.

Smoking rates for adults have decreased from about 43% in 1965 to about 18% in 2012. Despite significant progress since the first Surgeon General's report, smoking remains the single largest cause of preventable disease and death in the United States, with more than 40 million Americans dependent on tobacco smoking. Although the rate of tobacco smoking has declined steadily, more needs to be done to prevent and stop smoking. To promote smoking cessation and assist tobacco users to quit, smokers visiting health care settings are advised routinely about tobacco cessation and are reminded about future consequences of continuing smoking. Providing smoking cessation resources to patients in both general and specialty settings would improve the efforts and help them to quit. This study described characteristics of smokers in a hepatology clinic and provided smoking cessation resources to them according to current guidelines.

Methods

Data were collected over a period of seven months among patients (age > 18) who attended The Liver Associates of Texas clinic and completed the voluntary survey based on the Fagerstrom Test for nicotine dependence. Patients who screened positive for smoking were provided with both hard copies and on-line patient educational resources to help them to quit.

Frequency distributions on age, gender, ethnicity, race, BMI, education, diagnosis of interest, and the presence or absence of cirrhosis in the study population were performed. Data were analyzed using SPSS version 21.

Results

From 51 patients screened for smoking, 44 were interested in quitting. All 44 were provided with both copy and online educational resources. Four decided to try quitting without any intervention. Forty decided to quit using pharmacotherapy. Nine used Chantix®, three used Bupropion, and the remaining 28 used nicotine replacement therapy (12 prescribed patches, 10 prescribed gums, and six prescribed lozenges). From 44, 41 (93%) quit after three to six months follow-up without relapse.

Conclusions

Most participants were interested in smoking cessation. These results suggested that smoking cessation resources (both hard copy and online educational resources) should be available to hepatology patients as part of routine care.

Computed Tomography Fractional Flow Reserve:
An Appropriate Low-Risk Screening Tool for Coronary Disease

Alexander Robinson, D.O., Nicholas Isom, M.D., Chris Buckley, M.D.,
Thomas Rosamond, M.D.
KU School of Medicine-Kansas City

Introduction

Left heart catheterization for direct visualization of coronary vessels has been common practice for many years. The decision to perform percutaneous coronary intervention (PCI) is often based upon the observed percent stenosis in each vessel, and vessels with 70% or greater stenosis often have intervention performed. Over the last decade, fractional flow reserve (FFR) has gained traction in determining if a lesion is hemodynamically significant. FFR uses direct measurement of pressure and flow to determine if the stenosis is truly causing significant ischemia, thus giving a more approachable and objective measurement to assist with making the decision to intervene. More recently, computed topography fractional flow reserve (CT FFR) imaging has allowed physicians to obtain an FFR value without requiring an invasive left heart catheterization. As this is a relatively new technique, there is limited data comparing CT FFR with direct visualization of left heart catheterization.

Methods

Seventy-one patients received CT FFR during their care at Kansas University. Of those 71 patients, 19 patients had a diagnostic left heart catheterization as part of an ischemic workup. Seven of those patients had to be excluded due to misalignment, motion artifact, or previous stents obscuring the results. The 12 remaining patients had their CT FFRs compared with their catheterization results. An FFR result of less than 0.8 was considered to be hemodynamically significant, while a stenotic lesion of 70% or more was also determined to be significant.

Results

Using the guidelines noted previously for determining significant lesions, five of the patients were found to have FFR values that were less than 0.8. Three of those patients were found to have significant stenosis on catheterization. The remaining seven patients receiving CT scans all had non-significant FFR values. All seven of those patients had negative left heart catheterizations as well. For purposes of screening or diagnostics, CT FFR was found to have a sensitivity of 100% and specificity of 77.8% when compared to gold standard left heart catheterization.

Conclusions

Fractional flow reserve is a technique that has been gaining attention to help distinguish whether a stenotic lesion has hemodynamic significance. With this small cohort of patients, CT FFR was found to be a potentially useful screening tool for stenotic lesions requiring PCI. This could help prevent unnecessary left heart catheterizations which carry risks of bleeding and infection.

Use of Rituximab in Treatment of Recalcitrant Dermatitis Herpetiformis

Chisom Eze, M.D., Timothy Shaver, M.D.
KU School of Medicine-Wichita

Introduction

Dermatitis herpetiformis (DH) is a cutaneous-intestinal disorder caused by hypersensitivity to gluten whose treatment has mostly centered around gluten free diet, dapsone, and various immunomodulators. There has been recent evidence, however, supporting the use of Rituximab in the treatment of recalcitrant DH. This systematic review compiled and assessed evidence regarding: a) the mode of action, b) different treatment protocols, c) clinical responses, d) safety profile and future directions in B-cell-targeted therapy in treatment of DH.

Our case involves an 88-year-old male with biopsy proven celiac disease, recalcitrant DH, diffuse osteoarthritis, and prosthetic joint infection (on chronic Minocycline) in whom GI manifestations of disease had been well controlled with gluten free diet for over 10 years. However, diffuse bullous skin manifestations involving the trunk and proximal upper extremities have persisted despite Dapsone therapy which was aborted once patient developed methemoglobinemia. He then presented to the rheumatology clinic personally requesting initiation of therapy with Rituximab following online research regarding its use.

Methods

A systematic database search according to the PRISMA statement was conducted. Reference lists were searched for cases of recalcitrant DH and autoimmune bullous skin diseases treated with Rituxan on PubMed, Google Scholar, and EBSCO Health.

Results

The primary search yielded five articles. Four articles highlighted the use of Rituxan in the treatment of autoimmune bullous disease with an emphasis on pemphigoid and epidermolysis bullosa acquisita, as they were considered the more treatment unresponsive disorders. Mode of action was well described by one article as binding of Rituximab to CD20-expressing B lymphocytes leading to destruction of the target cells by a combination of antibody-mediated and complement-mediated cytotoxicity and apoptosis. One article provided treatment protocols with Rituxan specifically highlighting its use for DH administered at a dose of 375 mg/m(2) IV in weekly intervals for four consecutive weeks (lymphoma protocol) in addition to the standard immunosuppressive treatment. Clinical response was highlighted in two articles which reported use of Rituxan in more than 160 patients with autoimmune bullous dermatosis, with more than 95% benefiting from the treatment, however, these data did not elaborate on which specific diseases. One seminal case report detailed clinical and serological remission with normalization of both IgA Transglutaminase G2 and G3 antibody levels in an 80-year-old male with recalcitrant DH treated with Rituxan. Three articles described common adverse events of Rituxan including infections, lower limb deep vein thrombosis, PE, hypogammaglobulinemia, and neutropenia, with an overall mortality of 4%.

Conclusions

Our patient began four weekly infusions following the lymphoma protocol. The first successfully treated patient achieved complete clinical/serological remission and remained symptom-free up to 18 months. We anticipate a similar result for our patient. Additional research is needed to include Rituximab within the treatment algorithm for DH.

Acquired Methemoglobinenia and Sulfahemoglobinemia Secondary to Topical Lidocaine Use

Brandon Carlisle, M.D., Laura Thomas, M.D.
KU School of Medicine-Kansas City

Introduction

Methemoglobinemia and sulfahemoglobinemia are hemoglobinopathies which result in hypoxia from decreased oxygen binding due to iron oxidation. Medications commonly cause acquired methemoglobinemia and sulfahemoglobinemia. In this case, we present a 75-year-old male admitted to the medical intensive care unit (MICU) for hypoxemia of unknown etiology. The unique aspects of this case seem like it was a topical medication that caused this instead of oral medications.

Case Presentation

A 75-year-old with a history of coronary artery disease, end-stage renal disease requiring hemodialysis, presented from an outpatient urological procedure with acute hypoxic respiratory failure. On admission, he was oxygenating at 83% on room air and required 10 liters of oxygen via facemask despite being asymptomatic during this period. His exam was notable for perioral cyanosis and oxygenation improved when pressure was applied to his AV fistula. Initial laboratory work showed normocytic anemia, no electrolyte abnormalities or anion gap, BNP of 84, and negative troponins. Of note, his admission ABG showed a pH of 7.45, CO2 of 39, PaO2 of 186, and Bicarb of 27. He had imaging that was obtained that was unremarkable. Throughout his admission, he had low saturations on the pulse oximeter, but always had high PaO2 levels. An initial methemoglobin was negative. Initially, isosorbide mononitrate was thought to be the causative agent due to an elevated methemoglobin level, but the patient remained hypoxic at his follow-up appointment after discontinuation. His outpatient PFT's were unremarkable. A sulfahemoglobin and repeat methemoglobin levels were checked at his outpatient follow-up, which resulted in 3% and 2%, respectively. Medications were reassessed. The patient remarked that he applies lidocaine jelly on his AV fistula on days he receives dialysis, leading to the diagnosis.

Conclusions

Methemoglobin and sulfahemoglobin cause hypoxia by oxidation of iron in heme preventing oxygen binding. If a patient presents with a low SpO2 and a High PaO2, consider obtaining a methemoglobin level and sulfahemoglobin level. These causative medications include sulfonamides, dapsone, nitrates, benzocaine, or lidocaine. Patients typically have symptoms with methemoglobin since it shifts the oxygen dissociation curve left and patients with sulfahemoglobinemia typically have no symptoms since the oxygen dissociation curve moves to the right. After the diagnosis has been made, removing the offending agent typically resolves the hemoglobinopathy. If the patient is symptomatic and has Meth-Hb above 20%, methylene blue is indicated. There is no specific treatment for sulfahemoglobinemia except removal of the offending agent and close monitoring. Although methemoglobinemia and sulfahemoglobinemia are rare side effects of topical lidocaine, clinicians should suspect this in a patient with resistant hypoxia.

Clinical Relationship between CVID and Inflammatory Arthritis

Reid Eggleston, Megan Krause, M.D.
KU School of Medicine-Kansas City

Introduction

Common variable immune deficiency (CVID) is the most widespread form of primary immune deficiency. Diagnostically, CVID is marked by low serum IgG levels (<400 mg/dL) as well as concurrent IgA and/or IgM deficiency. Meanwhile, it has been reported that patients with CVID have higher incidence of autoimmune conditions. The objective of this project was to assess the impact of arthritis in a cohort of patients with CVID and to assess the pattern of arthritis involvement.

Methods

A retrospective chart review was performed at University of Kansas Medical Center. All individuals above the age of 18 between 1/1/2007-12/31/2016 with CVID and selective antibody deficiency with normal immunoglobulins (SADNI) were identified first by billing diagnosis then confirmed by chart review as determined by the treating immunologist. The chart was reviewed for rheumatologic evaluation for history of joint symptoms and data regarding arthritis including clinical assessment, laboratory testing, and radiographic results. Treatment strategies utilized for joint pain indication were recorded.

Results

One hundred, seventy patients were identified in the cohort with CVID and SADNI. In this cohort, 31 (18%) had an additional rheumatologic diagnosis and 19 (11%) patients had comorbid inflammatory arthritis confirmed by a rheumatologist. In the patient population investigated, the average age at CVID diagnosis was 48. One hundred, twenty (71%) of these patients were female and 163 (96%) were white. Of the 31 patients with a rheumatologic diagnosis, 10 had Sjögren's syndrome, six had rheumatoid arthritis, three had psoriatic arthritis, and three had systemic lupus erythematosus. Small joints of the hands tended to be the most commonly affected joint regardless of rheumatologic diagnosis with 14 patients having inflammatory PIP joint pain and 13 patients having inflammatory MCP joint pain. Other common sites of joint pain included the shoulder, knee, and wrist. Nine of the 19 patients (47%) identified with inflammatory joint pain were found to have erosive disease radiologically. Treatment of joint pain consisted of both traditional DMARD therapy and even newer-line monoclonal antibody regimens. Eighty-five percent of patients were treated with hydroxychloroquine and 50% were treated by TNF inhibitors.

Conclusions

Understanding the clinical pattern of arthritis that individuals with CVID experience will help inform future study and treatment strategies for clinicians.

Long-Term Use of Dual Antiplatelets for the Secondary Prevention of Atherothrombotic Events in Patients with and without MI: Meta-Analysis of Randomized Controlled Trials

Sumaya Hammami, M.D., Amratash Malodiya, Ph.D., Sandra Weiss, M.D., Paul Kolm, Ph.D., William Weintraub, M.D., Zaher Fanari, M.D.
KU School of Medicine-Wichita

Introduction

The potential benefit of long-term dual antiplatelet therapy (DAPT) for secondary prevention of atherothrombotic events is unclear. Data from different randomized controlled trials (RCT) using different agents in different subgroups showed inconsistent results. The goal of this study was to evaluate the efficacy and safety of long term DAPT for secondary prevention in patients with coronary artery disease with and without prior myocardial infarction (MI).

Methods

We performed a systematic review and meta-analysis from seven RCTs that tested different prolonged durations of DAPT for secondary prevention. Long term DAPT arm was defined as those receiving DAPT for more than 12 months. The long-term aspirin arm was defined as those receiving either long-term aspirin monotherapy or DAPT for six months or less.

Results

The use of long term DAPT was associated with a significant decrease in composite of death, MI, and stroke (6.08% vs. 6.71%; Odds Ratio OR=0.86 [0.78-0.94]; P=0.001). The reduction was mainly driven by a reduction in MI (2.77% vs. 3.16%; OR=0.75 [0.58-0.96]; P=0.03), but not in death, cardiac death, or stroke. This reduction of death, MI, and stroke was mainly noticed in patients with prior MI (6.32% vs. 7.28%; OR=0.86 [0.79-0.94]; P <0.001). The reduction was not seen in patients who underwent drug-eluting stent with no prior MI (4.66% vs. 5.03%; OR=0.87 [0.80-0.94]; P=0.38). The reduction in death, MI, and stroke was prominent with prasugrel (3.10% vs. 5.90%; OR=0.53 [0.37-0.74]; P<0.001) and ticagrelor (6.95% vs. 5.72%; OR=0.84 [0.75-0.93]; P=0.001). Long-term use of DAPT was associated with significant increase in major bleeding (1.47% vs. 0.88%; OR=1.65 [1.23-2.21]; P=0.001).

Conclusions

Long-term use of DAPT for secondary prevention is associated with lower risk of death, MI, and stroke, and beneficial in patients with prior MI, but it is associated with increased risk of bleeding. This reduction is not seen in post PCI patients with no prior MI. Prolonging DAPT requires careful assessment of the trade-off between ischemic and bleeding complications and should probably be reserved for patients with higher risk for atherothrombotic events.

VT Storm after Anti-Diarrheal Agent

Jihad Al-Khatib, Shravani Vindhyal, M.D., V. Subbarao Boppana, M.D., Mohinder Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

Introduction

Loperamide is an effective anti-diarrheal agent which is available over the counter. Loperamide is a synthetic opiate acing on mu-opiate receptors in the gastrointestinal tract to slow intestinal peristalsis and increase rectal tone. Loperamide does not cross the blood-brain barrier at therapeutic doses, hence it does not cause central nervous system effects such as euphoria similar to opiates. We report a case of recurrent ventricular tachycardia (VT) storm in a patient taking large recreational doses of loperamide.

Case Presentation

A 45-year-old female patient presented to the hospital after multiple episodes of dizziness, shortness of breath, and palpitations. Vital signs revealed a heart rate of more than 200 beats per minute and blood pressure was unrecordable. The rest of the physical examination was normal. Thorough history from the patient revealed opioid addiction and use of loperamide at 400 mg per day (maximum dose is 16mg/day). A 12-lead EKG demonstrated polymorphic ventricular tachycardia with a heart rate of 220 beats per minute. The patient was started on amiodarone and lidocaine drips, externally defibrillated 16 times, and underwent emergent cardiac catheterization which showed normal coronary arteries. She remained conscious through most of the VT storm. Transthoracic echocardiogram showed normal ejection fraction and no valvular abnormality. A temporary venous pacemaker was placed for ventricular tachycardia pacing and a heart rate of 100 beats per minute was achieved. Serum electrolytes and urine toxicology screen were normal. Cardiac MRI did not reveal any abnormality. Eventually, a temporary pacer was removed successfully, and the patient subsequently remained in sinus rhythm with normal QT interval without antiarrhythmic agents.

Conclusions

VT is a potentially fatal arrhythmia and is usually a manifestation of a serious heart condition such as ischemia, cardiomyopathy, myocarditis, or structural heart disease. Repeated VT episodes requiring cardioversion/defibrillation or appropriate implantable cardioverter defibrillator therapy are referred to as VT storm. Loperamide is known to interact with opiate receptors in the intestine and slows down peristalsis. Loperamide lacks the typical euphoric opiate effects when administered at recommended doses; euphoric effects can be elicited at higher-than-recommended doses. Loperamide acts on L-type calcium and potassium channels. Blocking of the sodium and potassium channels causes an increase in action potential duration resulting in prolonged QT and increased QRS duration leading to polymorphic VT. The uniqueness of our case is that our patient not only experienced one episode of VT but also had sustained and recurrent polymorphic VT with loperamide. Abrupt resolution of the VT, normalization of QT, and lack of the need for an anti-arrhythmic after discontinuation of loperamide in our patient makes us believe a causal relationship, but the pathogenesis remains unclear.

A Case of Red Herrings: Invasive Fungal Sinusitis

Kristen Constance, M.D., Nicholas Ojile, M.D., Brent Duran, D.O., Shelley Jones, M.D.
KU School of Medicine-Wichita

Introduction

Acute invasive fungal sinusitis (AIFS) is an aggressive and often fatal condition characterized by involvement of the paranasal sinuses with local extension to vital organs. It is a rare condition but should be suspected in a patient with immunocompromised status, recent surgical intervention, or uncontrolled diabetes mellitus.

Case Presentation

A 63-year-old Hispanic male with uncontrolled diabetes mellitus 2 was admitted for right facial droop. He had been admitted eight days prior for acute vision loss of the right eye attributed to central retinal artery occlusion and a third cranial nerve palsy. A CT of the head at that time was negative for hemorrhagic stroke and MRI of the brain was normal. He was started on high dose steroids for concern of temporal arteritis and biopsy was taken (biopsy later found to be negative).

On readmission, the patient had a severely elevated blood glucose at 700 mg/dL with a white blood cell count of 30 x 103/uL; however, he was afebrile and the leukocytosis was initially attributed to steroids. Neurological exam revealed multiple cranial nerve palsies of the right 3rd, 5th, 7th, and 12th nerves. MRI showed sphenoid sinusitis and no evidence for acute ischemia; MRV was negative. Cerebrospinal fluid analysis was unremarkable, and fluid culture and gram stain were negative. The patient was started on broad spectrum antibiotics as well as antiviral/antifungal therapy with acyclovir and amphotericin B. Facial palsies progressed to involve the right 2nd and 9th nerves. Infectious disease was consulted for sinusitis on day four of admission. The patient's hard palate was black. Repeat MRI showed infarction of intraocular muscles and necrosis of the posterior nasal turbinates. Urgent biopsy by ENT confirmed invasive fungal disease, and the patient was immediately transferred to university level hospital for emergent surgery. Family opted for comfort care at this facility, and the patient died.

Conclusions

This case illustrated the high mortality rate associated with AIFS, and the importance of maintaining a high degree of clinical suspicion for diagnosis. Although this patient had multiple confounders in his case (central retinal artery occlusion, steroids masking a possible immunologic reaction), he represented a highly at-risk patient population. Early recognition of AIFS with immediate initiation of antifungal treatment and surgical intervention is critical for improved prognosis in this highly fatal condition.

A Nearly Deadly Skin Lesion: Subcutaneous Rhizopus Murcor in a New Diabetic

Carley Trentman, Jesse Richards, D.O., Melissa Taylor, M.D.
KU School of Medicine-Kansas City

Introduction

Mucormycosis are fungi ubiquitously found in soil and rotting vegetation. These fungi release spores into the air, a common contamination in clinical laboratories. Mucormycosis infections typically occur in patients with hematologic malignancy or diabetes. The prevalence of mucormycosis varies significantly, from an estimated 0.014% of diabetics in India, to 1/80th of that in other developed countries. Rhizopus, a species of mucormycosis, is a deadly infection with mortality rates approaching 100% if disseminated or persistent neutropenia is present. Generally, sinus is the most common presentation with cutaneous involvement being rare. Here, we examine cutaneous mucormycosis in a man who presented in a hyperosmolar hyperglycemic state found to subsequently have diabetes.

Case Presentation

A 63-year-old Caucasian man with a history of basal and squamous cell carcinoma presented with a 5-cm wide necrotic lesion on the back of his neck. The lesion was present for several weeks, blackening two days before admission. The patient denied trauma to the area but was found to have poor living conditions. In the last four months, the patient lost 45 pounds with accompanying polydipsia, polyuria, and bilateral paresthesias over his toes. He reported a history of anxiety and depression and self-medicated with daily marijuana and 2-ppd of tobacco. Upon presentation, his glucose was markedly elevated (1,015); his A1C was >18.5%. CT revealed a density collection inferior to the occiput extending to the subcutaneous tissues. A pathology specimen was obtained. The patient was diagnosed with hyperosmolar nonketotic hyperglycemia secondary to newly diagnosed type II diabetes and started on insulin therapy. Vancomycin and Zosyn were initiated while culture and gram stain results pended. His diabetes remained controlled after admission. The culture returned positive for Rhizopus species consistent with mucormycosis. The patient was sent for surgical debridement. IV amphotericin B and IV isavuconazonium were initiated and a wound vacuum was placed. Once stable, he was discharged on four weeks of oral posaconazole.

Conclusions

While cutaneous mucormycosis is rare in immunocompetent hosts, this case illustrates when cutaneous involvement in a previously unknown diabetic was diagnosed and treated. Cutaneous involvement of Rhizopus is a recent discovery, with the first case reported from an ulcer in 2005. In general, the skin barrier represents a host defense against cutaneous mucormycosis, and the agents responsible are typically incapable of penetrating intact skin. Typically, traumatic disruption of the skin such as burns, maceration of the skin, introduction of contaminated soil, or surgical dressings/adhesives are the source. However, this patient denied any trauma to the area which illustrates a very atypical presentation. Overall, this patient's clinical course improved markedly. This case demonstrates the importance of early intervention of debridement of necrotic Rhizopus infection and glycemic control.

The Effect of Pembrolizumab in Absence of Programmed Death 1 Receptor

Layth Al Attar, M.D., Phu Truong, M.D.
KU School of Medicine-Wichita

Introduction

Tumor therapy has evolved greatly since the discovery of immunotherapies. New therapies permit a targeted approach with less side effects. Immunotherapies target specific components recognized on tumor cells, such as Programmed Death-1 (PD-1). Overexpression of PD-1 markers in solid gastrointestinal tumors is a consequence of deficiency in DNA mismatch repair mechanism. Pembrolizumab is an example of immunotherapy targeting PD-1. Pembrolizumab binding of PD-1 helps the immune system recognize tumor cells and initiate destruction cascade. We present a case of a PD-1 negative appendiceal tumor responding to pembrolizumab.

Case Presentation

A 63-year-old female presented in 2012 for increased abdominal girth associated with nausea, vomiting, constipation, and unintentional weight loss of 10 pounds. The patient was diagnosed with primary appendiceal adenocarcinoma based on pathology evaluation post-surgical resection of an abdominal mass. Genetic testing revealed a negative result for KRAS of wild-type, absence of microsatellite instability, and negative PDL-1 testing. The patient received different regimens of chemotherapy to manage the initial tumor and recurrence but failed due to side effects. She was started on pembrolizumab, although tested negative for PDL-1. After the first dose, she reported resolution of her abdominal pain and experiencing regular bowel movements. CEA was 95 ng/mL 10 days after the first dose. The test was repeated monthly to monitor pembrolizumab benefit. Results showed a downward trend to 80 then 43 ng/mL. CT scan was repeated two months after initiation of pembrolizumab. It showed an enlargement from previous left supraclavicular adenopathy, stable retroperitoneal periaortic lymphadenopathy, and decreased size of hepatic metastasis. The patient continued on this medication with notable clinical improvement. She completed 10 doses over a period of six months. A repeat CT scan showed increased calcification in relatively similar left supraclavicular lymphadenopathy and retroperitoneal adenopathy. Hepatic metastasis lesion was similar in size from previous scans. A more recent CEA test showed CEA dropping to 1.9 ng/mL.

Conclusions

To the best of our knowledge, this is the first report of a gastrointestinal tumor that is PDL-1 negative and without microsatellite instability showing impressive response to pembrolizumab. This case revealed a potential benefit of pembrolizumab. In normal conditions, the PD-1/PD-L1 pathway downregulates the immune response. Activation of this immune checkpoint inhibitor pathway by tumor cells allows for their evasion of an antigen-specific T-cell immunologic response. PD-L1 overexpression has been correlated with deficiency in DNA mismatch repair. Mismatch repair is a cellular defense mechanism that recognizes and repairs mismatch of bases during DNA replication and recombination. POLE gene mutation and tumor mutational burden are suggested additional tests to MSI testing aiming to improve patient selection.

An Often Unthought of Cause for High-Output Heart Failure

William Goodman, M.D., Mona B. Brake, M.D.
KU School of Medicine-Wichita

Introduction

High-output heart failure is an uncommon cause of heart failure (HF). High-output HF patients present with tachycardia, wide pulse pressure, hyperkinetic precordium, and jugular vein distention (JVD). A relatively infrequent, and less often thought of cause of high-output HF, is an AV-fistula (AVF) used for hemodialysis and is more common in upper arm fistulas as compared to forearm fistulas, and is usually seen when the blood flow exceeds 2 L/min. There is increasing evidence to support AVF associated heart failure. A cohort study looked at the incidence and main determinants of CHF in patients with stage 4-5 pre-dialysis CKD and found patients with mainly upper arm fistulas had a 9.5 times greater risk of developing acute decompensated CHF (OR=9.54, $P<0.0001$). A retrospective study found that 25.7% of patients with AVF post kidney transplant required fistula closure due to symptoms associated with high-output cardiac failure.

Case Presentation

A 53-year-old male presented with a past medical history of diabetes, hypertension, and a renal transplant 18 years ago for obstructive uropathy. He developed end stage renal disease due to transplant nephropathy and had a brachial artery to cephalic vein fistula created three years ago and was started on hemodialysis a few months after AVF creation. In the summer of 2017, he received his second kidney transplant. In May 2018, he presented to the emergency department with complaint of shortness of air, sharp left upper quadrant abdominal and left-sided chest pain of two days duration. Echocardiogram, troponin, and lipase were negative. Computed tomography of the abdomen showed no acute abnormalities. Chest x-ray revealed cardiomegaly and pulmonary venous congestion with a small left sided pleural effusion. Lab was significant for a slightly elevated BNP of 180 pg/mL. He was tachycardic (pulse 100) and tachypneic (rate 26). He had a grade 5/6 pistol shot systolic murmur best heard at apex. No JVD or edema were noted. Echocardiogram showed normal left ventricular systolic function with EF 55-60%, dilated IVC, and PAP 35 mmHg. Ultrasound of his AVF showed brachial artery flow at 5.3 L/min and cephalic vein flow of 4.4 L/min. He was treated with furosemide and had a rapid improvement of his symptoms. Chest x-ray done the following day showed resolution of the pulmonary venous congestion and the left pleural effusion. He was referred for surgical banding and discharged home.

Conclusions

AVF formation has proven survival advantages in patients starting chronic hemodialysis. Upper arm AVFs are more likely to cause high-output HF. Blood flow in AVFs should be monitored and banding considered in high-flow fistulas. Ligation of AVF maybe be considered in kidney transplant recipients with good graft function.

Parathyroid Crisis in a Patient with a Benign Parathyroid Adenoma

Nicholas Ojile, M.D., Georges Elhomsy, M.D.
KU School of Medicine-Wichita

Introduction

Primary hyperparathyroidism occurs in 0.1-0.3% of the general population with most cases in the seventh decade of life. Primary hyperparathyroidism is the result of inappropriately elevated parathyroid hormone (PTH) secretion causing hypercalcemia, with underlying pathologies including parathyroid adenoma and parathyroid hyperplasia; less than 1% of cases are due to parathyroid carcinoma. Patients can present in a parathyroid crisis with significantly elevated serum calcium and PTH levels, a rare complication of hyperparathyroidism.

Case Presentation

A 72-year-old Caucasian female was admitted for hypercalcemia. Family members reported that over the past several days the patient had become more confused, lethargic, and weak and she fell the day of her admission. Initial laboratory evaluation showed serum calcium of 14.7 mg/dl (normal 8.5-10.1 mg/dl) and phosphorus of 2.5 mg/dl (normal 2.5-4.9 mg/dl). A serum intact PTH was 1351 pg/ml (normal 14-85 pg/ml). Her renal function was normal. The patient was volume resuscitated with crystalloid fluid and subsequently treated with intravenous furosemide. She was also given 4 mg of zoledronic acid. A Sestamibi scan was suggestive of a parathyroid adenoma, and her serum calcium normalized prior to parathyroidectomy. A right inferior parathyroid gland mass was resected and was consistent with a parathyroid adenoma on pathology. The first fragment measured 4 x 2 x 1.3 cm and had a mass of 3.175 g; the second measured 3 x 1.4 x 0.9 cm with a mass of 1.286 g. Postoperatively, the patient developed hypocalcemia and required calcium supplementation.

Conclusions

Parathyroid crisis is a rare complication of primary hyperparathyroidism, occurring in 1.6% to 6% of primary hyperparathyroidism cases. Patients usually present with markedly elevated PTH levels and a calcium greater than 14 mg/dl and symptoms of hypercalcemia. The laboratory evaluation in a patient presenting with hyperparathyroid crisis from an adenoma can be confused with parathyroid malignancy as both can present with markedly elevated PTH. These patients are at high risk for multi-organ decompensation due to the severe hypercalcemia including oliguria, coma, and cardiac arrhythmias or even cardiac arrest if the calcium levels are not decreased prior to surgery with fluid resuscitation and loop diuretics. Parathyroidectomy is the definitive treatment for parathyroid crisis.

Takotsubo Cardiomyopathy Leading to Left Ventricular Free Wall Rupture

John Fritzlen, M.D., Tarun Dalia, MBBS, Bashar Amr, M.D., Jared Kvapil, M.D.
KU School of Medicine-Kansas City

Introduction

Takotsubo cardiomyopathy (TTC) is a rare but recognized cardiomyopathy presenting typically in post-menopausal women. The prevalence of TTC was reported in 0.02% of nationwide hospitalizations with an annual recurrence rate of 1.5% and an inpatient mortality rate of 4.5%. While TTC is usually reversible and resolves without complication, it may present with systemic embolism, life threatening arrhythmias, left ventricular outflow obstruction, and cardiogenic shock. We present an exceptional case of TTC leading to left ventricular wall rupture. To the best of our knowledge, only 19 cases have been reported so far showing this very rare outcome.

Case Presentation

A 75-year-old woman with past medical history of coronary artery disease, atrial fibrillation, hypertension, hyperlipidemia, and systemic lupus erythematosus initially presented to her local emergency department with dyspnea and chest pain. Upon evaluation in the emergency department, her systolic blood pressure was 230 mmHg and troponin was 0.26 ng/mL. She was transferred to our hospital for management of non-ST-segment elevation myocardial infarction (NSTEMI). Transthoracic echocardiogram revealed severe basal hypertrophy, moderately reduced ejection fraction, and Takotsubo appearance of the left ventricle involving the mid to apical portions. Repeat troponin was elevated at 6.83 ng/mL. The cardiac catheterization lab was activated immediately. En route, she became unresponsive and went into ventricular tachycardia. ACLS protocol was initiated. She was profoundly hypotensive and had a significant drop in hemoglobin from 14.2 mg/dL to 6.1 mg/dL. Resuscitation efforts were performed for approximately 50 minutes but were ultimately unsuccessful. Although angiography was unable to be performed, autopsy revealed blood in the left chest cavity, grossly patent coronary arteries, and a slit-like rupture approximately 1.0 cm x 0.8 cm along the anterior wall at 1.5 cm from the base of the heart with large areas of infarction in the left ventricle.

Conclusions

The criteria for the diagnosis of TTC include transient left ventricular systolic dysfunction, elevation of cardiac troponin, and electrocardiographic abnormalities in the absence of coronary obstruction. In this case, the echocardiogram findings were suggestive of TTC and autopsy showed non-obstructive coronary arteries confirming the diagnosis. The pathogenesis of TTC is incompletely understood but thought to be caused by a catecholamine surge often related to a physical or emotional stressor. Characteristics of patients with cardiac rupture in TCC include female gender, older age group, higher systolic and diastolic blood pressure, greater frequency of ST elevations in inferior leads, low ejection fraction, and higher left ventricular peak systolic pressure. Though the wall motion abnormalities in TTC typically resolve in days to weeks from onset, as described in our case, it can become a life-threatening condition in the acute phase. Hence, we recommend close monitoring of TTC patients for the first few days.

Extensive Deep Venous Thrombosis in a 31-Year-Old Female on Oral Contraceptives Diagnosed with May-Thurner Syndrome

Erik Calderon, M.D., Brent Duran, D.O., Stephanie Pankow, D.O.

Introduction

May-Thurner Syndrome is a rare but important diagnosis in a small percentage of new deep venous thrombosis diagnoses. The prevalence is unknown but approximated to be 2-5% of the causes of symptomatic deep venous thromboses. Risk factors include female gender, post-partum, and oral contraceptives. It is essential that providers are aware of the high rate of recurrence in patients with this diagnosis and act to decrease other modifiable risk factors such as oral contraceptives.

Case Presentation

A 31-year-old female, who was three months post-partum following a C-section, presented to the hospital with left lower extremity pain and swelling of one-day duration. She had started oral contraceptives three weeks prior to presentation. On physical exam, she had left lower extremity edema and palpable dorsalis pedis pulses. There was no warmth or tenderness to palpation. Ultrasound showed extensive thrombus throughout the left lower extremity venous system involving the popliteal to the common femoral veins with extension into the external iliac vein. There was no personal or family history of hypercoagulability. She was given a dose of therapeutic enoxaparin at that time and admitted for further management. A heparin drip was initiated while bridging to warfarin. Due to the extensive nature of her thrombus, she received catheter-directed TPA treatment to her left lower extremity and was found to have a chronically occluded inferior vena cava with a large clot burden. At that time, she was diagnosed with May-Thurner Syndrome. The patient decided to leave before bridging was complete. She left the hospital on therapeutic enoxaparin with arrangements to have outpatient stent placement performed by interventional radiology. Upon discharge, the oral contraceptives were discontinued and lower extremity stockings were placed. She likely will require long-term anticoagulation to prevent any further thrombus formations.

Conclusions

May-Thurner Syndrome, also known as iliac vein compression syndrome, is an extrinsic venous compression by the arterial system against bony structures in the iliocaval venous territory. The most common variant occurs when the left iliac vein is compressed between the right common iliac artery and the L5 vertebrae. Many times, a patient will be asymptomatic when there is only a partial obstruction, but it can progress to more symptomatic extensive deep vein thrombosis as was seen in this patient. Although uncommon, it is important to differentiate May-Thurner Syndrome as a cause of deep venous thrombosis as the treatment modalities and long term anticoagulation may differ.

Not Your Usual Suspect: A Case of Pill-Induced Esophagitis Caused by Demeclocycline

Gong Weng, M.D., Matthew Swan, M.D., John A. Bonino, M.D.
KU School of Medicine-Kansas City

Introduction

Pill-induced esophagitis is a well-recognized phenomenon. Most commonly reported cases are linked to tetracycline and its variant doxycycline. This case is unique in that demeclocycline, a semi-synthetic tetracycline derivative, is the causative agent. In a search of FDA adverse event reporting system and WHO adverse drug reactions database, only one similar case of esophagitis attributable to demeclocycline was found.

Case Presentation

A 59-year-old Caucasian woman presented to outpatient endoscopy lab for evaluation of odynophagia. She had a medical history of small cell lung cancer (SCLC) complicated by syndrome of inappropriate anti-diuretic hormone secretion (SIADH). This was being managed with demeclocycline. The patient was a former smoker and denied alcohol and NSAID use. Her physical exam was without notable abnormalities. On endoscopic evaluation, the patient was found to have a circumferential, ulcerated, necrotic, and fungating mass approximately 17 centimeters from the incisors. This appearance was suspicious for malignancy, and biopsies were obtained. Microscopic examination of the specimen demonstrated fibrinopurulent exudate and abundant foreign material, but no malignant cells. The patient was admitted to the oncology service for further evaluation, and underwent PEG placement for nutritional support. Given the high suspicion for underlying malignant etiology of the esophageal lesion, two subsequent EGDs were performed. Both sets of subsequent biopsies were also negative for malignancy. Upon review of the patient's medications, it was felt that patient was likely suffering from pill-induced esophagitis related to demeclocycline. Her demeclocycline was stopped, and the patient was started on a proton-pump inhibitor. Due to severe hyponatremia, a decision was made to restart the demeclocycline but to administer via her PEG tube. On subsequent follow-up in clinic, she reported complete resolution of her odynophagia. Due to her overall prognosis, in the setting of advanced lung cancer and other comorbidities, a repeat elective EGD was not scheduled.

Conclusions

This patient presented with a common complaint of odynophagia. Her presentation was obscured by a history of SCLC overshadowing a less frequently associated cause of pill-induced esophagitis, demeclocycline. Earlier recognition of demeclocycline's association with pill-induced esophagitis may have negated the need for multiple repeat endoscopies in the presence of non-diagnostic biopsy results. This case highlights the need to consider pill-induced esophagitis in the differential of individuals using demeclocyline who present with esophageal symptoms.

A Case of Intraparenchymal, Intraventricular, Subdural, and Subarachnoid Hemorrhage Following Catheter-Directed tPA for Extensive Iliac Deep Venous Thrombosis

Maha Mohamad, M.D., Rami H. Diab, M.D., John Peterson, D.O., Surayakumar Reddy, M.D., Nazih Moufarrij, M.D.
KU School of Medicine-Wichita

Introduction

Untreated lower extremity deep venous thrombosis (DVT) may progress to post-traumatic syndrome, pulmonary emboli, and death. Anticoagulation remains the hallmark of treatment, however, thrombolysis is deemed appropriate in patients with massive proximal DVT with low risk of bleeding and good functional status. Importantly, the use of low dose tPA with catheter-directed therapy as compared to higher dose tPA with systemic therapy has not been shown to reduce bleeding complications. As of yet, there remains no clinically validated tool to estimate the risk of bleeding in patients with DVT receiving catheter-directed thrombolysis.

Case Presentation

A 64-year-old female with atrial fibrillation (not previously anticoagulated), fibromyalgia, and osteoarthritis was admitted with severe left lower extremity pain of a few days duration. Lower extremity ultrasound revealed extensive veno-occlusive disease and confirmatory venography demonstrated filling defect extending from the mid-left external iliac vein into the entirety of the common iliac vein. Catheter-directed tPA was offered by interventional radiology. The patient's HAS-BLED score was 2 (hypertension, age > 65). An infusion catheter was placed in the iliac vein, primed with 4 mg of tPA and left to instill tPA at 1 mg/hour with heparin instilled through the indwelling sheath.

The following day, the patient complained of a progressive headache. CT head without contrast revealed intraparenchymal hemorrhage in the right occipital-temporal region measuring 2.0 x 1.0 cm with subdural and subarachnoid hemorrhage overlying the parasagittal frontal lobes. The patient was started on Gabapentin for headache, hypertonic saline for cytotoxic brain edema, and physical therapy for deconditioning. Neurosurgery recommended medical therapy. The patient's headache acutely worsened the following day prompting another CT head showing an increase in size and density of both temporal-occipital lobe and left frontal lobe hemorrhages now measuring 3 x 6 cm and 1.6 x 2.6 cm respectively, each with focal mass effect. A new intraventricular hemorrhage also was seen in the dependent portion of the occipital lobes. Follow-up CT head showed evolution of intraparenchymal hematomas to 6.5 x 3.2 cm with slight worsening of edema and mass effect. The patient gradually improved with tighter blood pressure control and physical therapy. Repeat CT head showed reduction in edema of parenchymal hematomas and minimal subarachnoid blood, and the patient was discharged to inpatient rehab with follow-up.

Conclusions

Regardless of the dose or route of administration, tPA is associated with bleeding risk even in the absence of absolute contraindications. In the absence of a validated bleeding risk assessment tool for such scenarios, clinical acumen and a high index of suspicion are quintessential for timely management of potentially debilitating bleeding complications should they arise.

Association between Electronic Cigarette Use and Coronary Heart Disease and Stroke

Job Mogire, M.D., Tabitha Muutu, M.D., Paul Ndunda, M.D.
KU School of Medicine-Wichita

Introduction

In 2016, 3.2% of US adults and 11.3% of high school students reported using electronic cigarettes (e-cigarettes) in the preceding 30 days. Its use among young people increased by 900% between 2011 and 2015. Though CHD and stroke are leading causes of death in the US (nearly 500,000 deaths in 2015), the association between e-cigarette use and coronary heart disease (CHD) and stroke remains unknown. This study explores the association between e-cigarette use and CHD and stroke.

Methods

The study is a cross-sectional analysis of the 2016 Behavioral Risk Factor Surveillance System (BRFSS), an annual chronic disease risk factor survey conducted by the CDC. The sample included 66,795 respondents reporting regular use of e-cigarettes. The control group was the 343,856 respondents that reported never using e-cigarettes. Odds ratios were calculated using logistic regression analysis. Among the covariates tested, age, sex, smoking status, diabetes, exercise, and Body Mass Index (BMI) categories showed significant effects on the model and were adjusted for in the outcomes. Since this was a telephone-administered questionnaire, fatal events were not included.

Results

Overall 21% of BRFSS respondents reported ever using e-cigarettes, with 54.7% of the users being female ($P<0.0001$). Compared with non-users, e-cigarette users had a lower mean age (44 vs 57 years [$P<0.0001$]), lower mean BMI (27.7 vs 28.1 [$P<0.0001$]) and a lower rate of diabetes (9.8% vs 12.1% [$P<0.0001$]). They, however, had higher rates of cigarette smoking (78.7% vs 37.4% [$P<0.0001$]). Compared with non-users, e-cigarette users had higher odds of myocardial infarction (adjusted Odds Ratio [OR] 1.59 [1.53-1.66] $P<0.0001$), angina or coronary heart disease (OR 1.4 [1.35-1.46] $P<0.0001$), and stroke (OR 1.71 [1.64-1.8] $P<0.0001$). There was an interaction between cigarette smoking and e-cigarette use for all three outcomes ($P<0.05$).

Conclusions

E-cigarette use is associated with higher rates of non-fatal myocardial infarction, non-fatal angina/coronary heart disease, and non-fatal stroke. However, there is a need for cohort studies to establish causation and study fatal events.

Blurred Lines:
HLH Recognition Complicated by Infectious, Neoplastic, and Autoimmune Features

Clarissa Smith
KU School of Medicine-Kansas City

Introduction

Hemophagocytic lymphohistiocytosis (HLH) is an uncommon pathophysiological entity in which the body indiscriminately autophagocytoses hematopoietic cells and their precursors within tissues such as liver, bone marrow, spleen, and lymph nodes. The Histiocyte Society's diagnostic criteria for HLH is met with five of the following: fever, splenomegaly, two-lineage cytopenia, hypertriglyceridemia, hypofibrinogenemia, elevated ferritin, hemophagocytosis, low or absent natural killer cells, or elevated CD25. Unfortunately, many cases of HLH are still diagnosed at autopsy, due to these vague clinical indicators in combination with the rapidity of irreversible hemostasis disruption.

Case Presentation

A 76-year-old male was admitted in transfer for acute onset mental status changes with elevated transaminases, thrombocytopenia, ferritin > 40,000µg/L, and 2/4 Systemic Inflammatory Response Syndrome (SIRS) criteria. Past medical and family history were bland, but the patient reported incurring recent insect bites while doing yardwork around his rural home. Consultations with infectious disease and hematology were requested right away and empiric doxycycline for tickborne illness and dexamethasone for HLH was initiated while additional testing was performed. An extensive infectious workup was positive only for Epstein Barr Virus (EBV) viremia. Due to its known correlation with HLH, Etoposide was started according to HLH-2004 recommendations. The patient's condition continued to decline with worsening pancytopenia, a new dialysis requirement, loss of airway protection, and DIC. The patient awoke with acute left-sided weakness, and MRI demonstrated multiple scattered infarcts. Due to the patient's continued decline, Etoposide was subsequently discontinued. He then developed a pulmonary infiltrate, hypotension, as well as bradycardia, and ultimately died from cardiac arrest. Autopsy was performed with family consent and revealed disseminated hyalohyphomycosis due to the immunocompromise induced by HLH triggered by EBV viremia.

Conclusions

While familial forms of HLH have been described, it more frequently arises secondarily to states of systemic inflammation such as malignancy, autoimmunity, or infection. Ongoing investigations demonstrate positive correlation with the exuberant production of macrophage-activating cytokines such as interferon-gamma and tumor necrosis factor-alpha. Many diverse viral, protozoal, fungal, bacterial, and even plasmodium pathogens and opportunistic infections have been implicated in HLH. Of these, EBV is the most common and also has the highest lethality rate. Testing for underlying pathogens should be guided by the patient's medical and social history. Despite increased awareness and recognition of the syndrome, 50% of patients die due to multi-organ failure or infectious complications. The rarity of this syndrome also limits trials to create and improve therapeutic options. Current treatment recommendations are based on a combination of immunotherapy and chemotherapy outlined in the HLH-2004 study, but early recognition of this condition continues to be the most effective means of decreasing associated morbidity and mortality.

Percutaneous Repair of Mitral Valve Regurgitation Due to Post-Infarction Papillary Muscle Rupture Using Mitraclip Apparatus

Cyrus Munguti, M.D., Paul Ndunda, M.D., Bassem Chehab, M.D.
KU School of Medicine-Wichita

Introduction

Post-infarction papillary muscle rupture carries high morbidity and mortality. While surgical repair is gold standard, percutaneous repair is now feasible especially in high surgical risk patients. We present a case of successful percutaneous repair of mitral valve regurgitation from a ruptured papillary muscle.

Case Presentation

A 66-year-old male patient presented to the emergency department with sudden onset chest discomfort and exertional dyspnea. He described a left-sided, dull discomfort accompanied by exertional dyspnea. He had had a recent sternotomy and was not very active at baseline. He had a past medical history of coronary artery bypass (CABG) for coronary artery disease, open aortic valve replacement followed by a trans-femoral aortic valve replacement (TAVR). On examination, he had a systolic murmur at the apex radiating to the axilla and bilateral edema on the ankles. A transthoracic echocardiogram (TTE) revealed mitral regurgitation that was not previously noted. A trans-esophageal echocardiogram (TEE) showed flail anterior leaflet at A1 segment with torrential mitral regurgitation. He declined a third sternotomy but was agreeable to MitraClip repair. He consented to the procedure.

An intraoperative TEE revealed the papillary muscle at the base of the anterior mitral leaflet mainly at the A1 segment, with significant flail and prolapsing leaflet and severe mitral regurgitation. Trans-septal puncture was done and then an E-clip sheath was inserted in the left atrium and heparin was given per protocol. A first clip was deployed in at the medial part of the A1 segment, capturing the base of the papillary muscle in the A2 segment. Then a second clip was deployed lateral to the first at the level of A1-P1, decreasing mitral regurgitation to trace with a mean gradient of 5 mmHg. The system was removed. Perclose was deployed with excellent hemostasis.

Conclusions

Trans-catheter mitral valve repair using the MitraClip (Abbot) apparatus is mostly done for chronic mitral regurgitation due to valve degeneration or functional chronic conditions. Percutaneous repair using MitraClip offers benefits to high risk patients including less destruction of surrounding tissue associated with surgery, sudden return in pre-morbid cardiac output following the repair by reducing the left atrial and ventricular pressures hence increased contractility and reversal of cardiogenic shock. Our patient had complex anatomy following open aortic valve replacement, TAVR valve in situ, and CABG but still managed to get good valve coaptation. A TTE done on the 30-day follow-up showed minimal mitral regurgitation.

In select cases, percutaneous repair of ruptured papillary muscle is possible and should be considered where surgical intervention is not an option.

The Problematic Nature of Fibromyalgia Diagnosis in the Community

Sachin Srinivasan, M.D., Eamon Maloney, M.D., Rami Diab, M.D., Brynn Wright, K. James Kallail, Ph.D., Michael Kennedy, M.D., Frederick Wolfe, M.D.
KU School of Medicine-Wichita

Introduction

Fibromyalgia is a common, if contested diagnosis. A physician's diagnosis is required for access to treatment and disability status. Recently, two studies have suggested that clinical diagnosis is inaccurate and does not reflect current scientific definitions, but this hypothesis has not been tested formally in the community. We tested whether fibromyalgia was diagnosed accurately in the community, whether diagnosis was biased by sex, and examined the effect of physician and criteria-based diagnosis on treatment.

Methods

We surveyed 3276 consecutive adult patients attending one of 26 primary care practices in Kansas using a completely de-identified self-report questionnaire that contained the 2016 modification of the American College of Rheumatology diagnostic criteria to determine current fibromyalgia status (CritFM). We also determined by self-report whether the patient had a previous or current physician's diagnosis of fibromyalgia (MDFM), the level of fibromyalgia symptom severity as measured by the polysymptomatic distress scale (PSD), time from MDFM diagnosis, and the use of current and past FDA approved pharmacotherapy. Questionnaires were dispensed and collected prior to physician visits by second year medical students during a Summer Training Option in Rural Medicine (STORM).

Results

The prevalence of physician (MDFM) and criteria (CritFM) diagnosed fibromyalgia in primary care was 5.3% (95% CI 4.4%, 6.1%) and 5.5% (95% CI 4.8%, 6.4%), respectively. However, only 34.5% with MDFM met fibromyalgia 2016 criteria (CritFM), and only 33.1% with CritFM also had MDFM. The kappa statistic for agreement beyond chance between physician diagnosis and diagnostic criteria was 0.330 (slight agreement). The mean pattern standard deviation score was 13.2 and 18.4 in MDFM and CritFM, and generalized pain was present in 42% and 100%, respectively. Diagnostic agreement increased slightly with duration of MDFM. The odds ratio for a patient being a women compared to being a man was 4.0 (95% CI 2.5, 6.2) for MDFM and 1.9 (95% CI 1.4, 2.8) for CritFM, $p = 0.004$. Current treatment and ever treatment was related to MDFM but not to CritFM. Only 4.1% of patients with CritFM but not a physician diagnosis ever received the drug treatment. By contrast, 70.7% of patients with a MDFM were treated.

Conclusions

There is little agreement between MDFM and CritFM (kappa= 0.330). Only 1/3 of MDFM satisfy fibromyalgia criteria, and only 1/3 of patients who meet criteria have a clinical diagnosis of fibromyalgia. Physician diagnosis compared with CritFM is biased and more likely in women. Fibromyalgia treatment is rare in CritFM (4.1%), but very common in MDFM (70.7%). Overall diagnosis of fibromyalgia by physicians appears idiosyncratic and unrelated to FM criteria. There appears to be no common definition of fibromyalgia in the community.

A Rare Case of Prekallikrein Deficiency

Sammy Tayiem, M.D., Abdulraheem Yacoub, M.D.
KU School of Medicine-Kansas City

Introduction

Prekallikrein (PK) deficiency is an extremely rare autosomal recessive disease. First described in 1965, PK deficiency was originally discovered in four siblings with an asymptomatic and prolonged aPTT. It has since only been reported around 80 times in the literature. We hereby report a patient with confirmed PK deficiency.

Case Presentation

A 41-year-old African-American female without a significant past medical history was referred to our Hematology and Oncology clinic due to a prolonged aPTT (100.7 seconds) on her pre-operative workup prior to a hepatic hemangioma resection. She denied any personal or family history of abnormal bruising, bleeding, or thrombotic phenomena. The patient has had two C-sections and a hernia surgery without hemorrhagic complications. Physical exam was unremarkable.

Hematological workup confirmed a prolonged aPTT with full correction on mixing study, normal PT/INR, normal thrombin time, normal von Willebrand Factor profile, and normal platelet function assay. The patient had normal clotting factor levels including XII, XI, IX, VIII, X, V, II, and a normal HMW kininogen. Additional testing confirmed a severe PK deficiency (< 5%). A confirmatory assay at a reference lab confirmed the results. She underwent the planned resection of hepatic hemangioma without special precautions and had no abnormal bleeding. The PK deficiency and prolonged aPTT persisted after resection of the hemangioma.

Conclusions

PK is a single-chain protein that is converted to its active form, kallikrein, by activated factor XII. Activated kallikrein may further activate factor XII, promote the activation of the fibrinolytic system, and cleave kininogen to release bradykinin. PK deficiency can either be inherited or acquired in patients with hepatic disease or disseminated intravascular coagulation. The deficiency has been localized to mutations in the KLKB1 gene on chromosome 4. PK deficiency has been reported in nearly all races, however, it is seen more frequently in African-American patients. The true prevalence of PK deficiency is unknown as most affected people are usually asymptomatic. PK deficiency can be identified by an isolated prolonged aPTT and normal PT, in which the aPTT corrects to normal in mixing studies. It is then finally confirmed with a specific PK assay.

Identification of such a rare deficiency is clinically significant in that a prolonged aPTT will repeatedly prompt an extensive workup that may unnecessarily delay treatments or procedures. Despite a prolonged aPTT, there is no increased risk of bleeding due to compensation by the extrinsic pathway. PK deficiency may, however, be associated with thrombotic phenomena due to defective activation of the fibrinolytic system, as well as hypertension secondary to decreased bradykinin levels; demonstrating the far-reaching and complicated effects of prekallikrein.

A Rare Case of Nonbacterial Thrombotic Endocarditis (NBTE) in a Young Patient with Acute Myelogenous Leukemia

Rami Atallah, M.D., Nicholas Ojile, M.D., Kamal Chamoun, M.D., Fredy Nehme, M.D., Mohinder Vindhyal, M.D., M.Ed.
KU School of Medicine-Wichita

Introduction

Nonbacterial thrombotic endocarditis (NBTE), formerly known as marantic endocarditis, is a rare condition characterized by deposition of sterile thrombi on heart valves with the absence of bacterial growth on blood cultures. NBTE is clinically significant as it increases the risk of systemic embolic events including stroke. Microscopically, NBTE results from agglutinated platelets mixed with strands of fibrin. NBTE is most commonly associated with advanced malignancy and various autoimmune conditions. In this report, we present a case of NBTE in a 26-year-old woman presenting with AML.

Case Presentation

A 26-year-old female with a history of recurrent pancreatitis presented to her primary care physician with abdominal pain, cough, dyspnea, and fatigue. Outpatient lab work showed a hemoglobin of 4 g/dl for which she was admitted to our institution for further evaluation. On admission, her platelets were 56x10^9/L, WBC 14x10^9/L with 40% peripheral blasts. Bone marrow biopsy confirmed the diagnosis of acute myelomonocytic leukemia. The patient was then started on induction chemotherapy with daunorubicin and cytarabine. An initial transthoracic echocardiogram revealed an ejection fraction of 55% with no valvular abnormalities. After starting chemotherapy, the patient’s respiratory status rapidly worsened requiring intubation. The patient's blood counts continued to drop requiring multiple blood and platelet transfusions. Blood cultures were repeatedly negative. A week after being intubated, the patient developed multi-organ failure including brain and kidneys. The patient subsequently suffered from left leg ischemia caused by an arterial clot in the deep femoral artery. Family discussion led to comfort care decision. Advance supportive care was withdrawn, and the patient died soon after. The autopsy revealed fibrin vegetations on the aortic and the tricuspid valve consistent with NBTE as well as massive non-hemorrhagic infarct on the left frontal, parietal, and the occipital lobes. Multiple infarcts were evident in spleen and both kidneys. Pulmonary hemorrhage was also noted.

Conclusions

NBTE is a rare condition, associated with very high mortality and morbidity rates. It is uncommonly reported in patients with acute myeloid leukemia and is often a postmortem diagnosis. The most likely cause of death is multiple infarcts seen in the brain and various organs. Diagnosis of this condition is always challenging. Physicians should always suspect NBTE in patients presenting with multiple infarcts with a history of malignancy or autoimmune disease.

Leclercia Adecarboxylata in a Post-Fasciotomy Polymicrobial Infection

Andrew Weaver, M.D., Margaret E. Hagan, M.D.
KU School of Medicine-Wichita

Introduction

Leclercia adecarboxylata is a rare Gram-negative bacillus that previously belonged to the Escherichia genus. It has been implicated in traumatic wounds, nosocomial pneumonias, and bloodstream infections including central line-associated bloodstream infections. Some strains have shown antibiotic resistance including extended spectrum beta-lactamase. The following case demonstrated Leclercia adecarboxylata as part of a lower extremity polymicrobial infection that developed in a patient following compartment syndrome, fasciotomy, and surgical debridement.

Case Presentation

A 31-year-old male was admitted for management of a suspected infected fasciotomy wound. One month prior, he had sustained a tibia fracture that was surgically repaired; he subsequently developed compartment syndrome that necessitated fasciotomy and surgical debridement of necrotic muscle. The patient was previously healthy with no chronic medical problems, no home medications, no allergies, and a benign family history. Surgical history included back surgery and appendectomy in addition to his recent surgeries. He was a daily smoker. Review of systems at admission was negative. Vital signs were notable for tachycardia. Physical examination revealed a large left leg wound with granulation tissue and an odor. He had mild leukocytosis as well as low hemoglobin and serum albumin. Empiric intravenous vancomycin and piperacillin-tazobactam were initiated. Culture of the wound bed grew pan-susceptible Leclercia adecarboxylata, Group G Streptococcus, and Corynebacterium species. Surgical culture from a repeat debridement grew Enterobacter cloacae, Enterococcus faecalis, and Candida parapsilosis. The infection improved with piperacillin-tazobactam, oral fluconazole, and local vancomycin-tobramycin beads followed by a prolonged course of intravenous ertapenem and oral fluconazole at home.

Conclusions

Leclercia adecarboxylata is an uncommon but important pathogen in both mono- and polymicrobial infections. Clinicians should consider this entity particularly when managing post-trauma and nosocomial infections.

When Empiric is Imperative: Differentiating Encephalopathy from Encephalitis

Gretchen Beaver, Jesse Richards, D.O., Jessica Newman, D.O.
KU School of Medicine-Kansas City

Introduction

A differential diagnosis of altered mental status is expansive, especially when the patient is non-English speaking and multiple comorbidities are present. Knowing where to start is daunting; determining if the patient has encephalopathy or encephalitis is paramount as a delay in antimicrobials is devastating. This case highlights how immediate empiric therapy for common causes of encephalitis can have a profound impact on outcome when a history cannot be obtained.

Case Presentation

A 71-year-old, non-English speaking, Southeast Asian woman presented with two-days of lethargy and one-day of altered mental status. Her past medical history included metastatic breast cancer, systolic and diastolic heart failure, and stage 3 chronic kidney disease. She was hospitalized the month prior for pneumonitis with a prednisone taper and remained on 40 mg daily. Obtaining medical history was limited. Family accounts noted impaired cognition at baseline and deconditioning from the recent hospitalization. She was afebrile and hemodynamically stable. On examination, she was lethargic and only responsive to pain. Additional pertinent findings included scleral icterus and abdominal distention without fluid wave. She had no meningismus or rash.

A complete blood count was notable for stable chronic pancytopenia. Pertinent lab results included a baseline creatinine of 2.04, total bilirubin of 3.9 (increased from 2.2), an unremarkable urinalysis, and a normal lactate. Computed tomography of the abdomen/pelvis demonstrated a mildly nodular liver, suggestive of cirrhosis and mesenteric congestion. Lumbar puncture was delayed due to pending coagulopathy studies and CNS imaging (no acute findings), so empiric intravenous ceftriaxone, vancomycin, ampicillin, and acyclovir were initiated. Cerebrospinal fluid demonstrated RBC 1400, WBC count of 70 (53% lymphocytes), protein 64, and normal glucose. The pleiocytosis confirmed the clinical diagnosis of encephalitis. No organisms were seen on gram stain and cultures remained negative. HSV-2 PCR was positive with antibiotic therapy de-escalated and prednisone tapered. Within one week of treatment, her mental status improved. By week two, she was at baseline per family and stable for discharge.

Conclusions

While HSV-2 encephalitis was low on the initial differential, failure to promptly initiate antimicrobial therapy may have led to irreversible neurologic disability or death. Studies have shown that time of onset of medical treatment impacts resultant outcome with treatment initiation within 1.8 days correlating to good outcome and 4.0 days correlating to poor outcome. HSV-1 encephalitis is generally uncommon (incidence 1.2/100,000) and HSV-2 encephalitis even more rare. As treatment with IV acyclovir has decreased mortality from ~70% to ~20%, there should be no delay in empiric therapy. If encephalitis is on the differential (especially in this patient on prolonged steroid therapy) and lumbar puncture is delayed, consider prompt empiric IV antimicrobials including acyclovir. The diagnostic standard, HSV PCR remains positive for the first few days of therapy.

Immunotherapy Induced Myocarditis

Garret Seiler, D.O., Aarati Keshary, M.D., Jeremy Deutsch, M.D.
KU School of Medicine-Wichita

Introduction

Tumor cells evade the host immune system by overexpressing PD-1 and CTLA-4 surface proteins. Monoclonal antibodies targeting these receptors have revolutionized cancer treatment. Combinations of these immunotherapies have received FDA approval as salvage therapy or first line therapy in select tumors. Autoimmune inflammatory reactions are well-recognized side effects of these medications. The skin, the gastrointestinal system, the thyroid gland, and the liver are the most commonly affected organs. Less commonly, the lungs, muscles, or brain may be affected. Signs of these side effects merit prompt, high-dose corticosteroids and cessation of the offending agent.

Case Presentation

A 50-year-old female presented to the hospital with shortness of breath and left-sided chest pain. She had a past medical history of Stage IV non-small cell lung adenocarcinoma (EGFR, ALK, ROS-1 Negative). She had been placed on Ipilimumab and Nivolumab 13 months prior to the current visit as part of a clinical trial and had experienced a good response without tumor progression or significant side effects. Her current evaluation was significant for an elevated troponin of 0.21 ng/mL as well as a new intra-ventricular conduction delay without ST segment elevation on EKG. Vitals revealed heart rate of 106 beats per minute, respiratory rate of 17, blood pressure of 95/73 mmHg, and oxygen saturation of 100% on room air. AP view chest x-ray was suspicious for pulmonary edema. Brain natriuretic peptide and creatinine phosphokinase were unremarkable. Over 12 hours, her troponin trended up to 2.9 ng/mL. A trans-thoracic echocardiogram revealed diffuse akinesis and reduced ejection fraction of 15%. A heart catheterization showed no evidence of coronary artery disease. Immunotherapy-induced myocarditis was suspected. High dose corticosteroid therapy was initiated. By the fourth day of hospitalization, her troponin trended down to normal levels and her symptoms began to abate. On hospital day eight, a repeat trans-thoracic echocardiogram showed improvement of her ejection fraction to 45%. EKG no longer showed intra-ventricular conduction delay. Serum PCR studies for adenovirus, enterovirus, and Epstein-Barr virus were negative.

Conclusions

Autoimmune inflammatory reactions are well recognized side effects of PD-1, PDL-1, and CTLA-4 inhibitors. Increased T-cell activation following the unmasking of the tumor cells to the immune system leads to bystander tissue damage. In 2016, two case reports of Ipilimumab and Nivolumab induced fulminant myocarditis were described. Both cases resulted in rapid mortality despite high-dose corticosteroid therapy. This case of immunotherapy induced myocarditis shared similarities but also contained key differences from the previous cases: the timing of onset, degree of cardiac injury, and therapeutic response to steroid therapy. As their indications increase, myocarditis is increasingly recognized, as well as the deadly side effect of immunotherapies. Prompt recognition may improve mortality outcomes.

Cutaneous Leishmaniasis

Thanuja Neerukonda, Ryan Kubat, D.O., Fernando Merino, M.D.
KU School of Medicine-Kansas City

Introduction

Leishmaniasis is a spectrum of diseases caused by species of Leishmania protozoa and transmitted by sandfly vectors. The most common clinical manifestation is cutaneous leishmaniasis with approximately 0.7-1.2 million new cases per year, primarily in the Middle East and Central and South America. We report a case of complicated cutaneous leishmaniasis diagnosed in an immigrant from Honduras and a unique treatment with excisional therapy only.

Case Presentation

A 22-year-old male with no past medical history was referred to the University of Kansas Infectious Diseases clinic with a left facial lesion. He first noted a small “pimple” about six months prior. The lesion began growing in size and developed some surrounding erythema. He was evaluated by a physician and prescribed a course of oral antibiotics without improvement. The lesion grew for three months and developed a necrotic center, after which it stopped growing and kept the same appearance. It was not associated with any pain or drainage and he did not have any fever or systemic symptoms. Examination revealed a 2x3 centimeter left pre-auricular skin lesion with well-defined borders and a large central necrotic eschar. There were no mucosal lesions or lymphadenopathy. CBC and CMP were normal. Further history revealed the patient had emigrated from Honduras three months prior to onset of symptoms. He traveled for two weeks, sleeping in the open air, and recalling many insect bites during his trip. He crossed the border in Texas and spent three weeks in a detention center in Louisiana prior to moving to Kansas City.

A biopsy obtained by the referring physician revealed abundant intracellular organisms with acute and histiocytic inflammation, most consistent with leishmaniasis. After consulting with the CDC and plastic surgery, the decision was made to excise the lesion for definitive diagnosis along with performing a skin flap for optimal cosmetic outcome. Surgical pathology was again consistent with leishmaniasis. A sample was submitted to the CDC for PCR testing and DNA sequencing, confirming infection with Leishmania mexicana. Pharmacologic treatment with liposomal amphotericin B was attempted, however, he reported chest tightness, dyspnea, and flushing within the first minute of infusion. He developed the same symptoms despite pre-medicating with diphenhydramine and acetaminophen while slowing the infusion rate. Other pharmacologic options were discussed, but the patient declined further treatment. He continues to be seen for surveillance; he has a hypertrophic scar but no evidence of recurrent infection.

Conclusions

Recognition and diagnosis of cutaneous leishmaniasis is important for determining disease severity and initiating treatment if indicated. Although only a few cases of transmission have been reported within the United States in Texas and Oklahoma, it is an emerging infection to be aware of in travelers and migrants from endemic areas.

From Murmur to Amyloidosis: Stressing the Importance of Physical Exam

Ethan Hacker, M.D., Branden Comfort, M.D., MPH
KU School of Medicine-Kansas City

Introduction

AL amyloid cardiomyopathy is a rare, but serious disease caused by abnormal deposition of amyloid protein within the heart leading to heart failure with significant mortality. Here, we present a case of cardiac amyloidosis diagnosed after unusual exam findings.

Case Presentation

A 53-year-old female with past medical history of breast cancer who had undergone mastectomy and radiation, depression, hypertension, and chronic pancreatitis presented to her primary care physician with complaint of enlarging neck lump over the past two months. This had been accompanied by gagging, difficulty with swallowing food, and weight loss of seven pounds. She described no history of stridor, but stated that with exertion she became more short of breath. On examination, she had an enlarged thyroid gland without nodularity, moderate tachycardia, and a 3/6 holosystolic murmur. She also had 1+ pitting edema in her lower extremities that had been developing over the same time period as the rest of her symptoms. A CT scan of her neck showed a substantially enlarged thyroid with evidence of compression on the esophagus. The patient was referred to ENT and plans were made for thyroidectomy. Prior to her thyroidectomy, an echocardiogram was obtained for her new systolic murmur which showed obstructive hypertrophy with diastolic dysfunction. Her surgery was delayed and she was transitioned from a diuretic to a beta blocker in order to avoid decreased preload that could lead to further obstructive cardiac physiology. Cardiology referral was placed, and she underwent evaluation with cardiac MRI and stress testing. After being cleared by cardiology, she underwent thyroidectomy and was found to have significant deposition of amyloid on surgical pathology. Her cardiac MRI showed a prominent basal interventricular septum, as well as diffuse infiltration of myocardium consistent with amyloid cardiomyopathy. Serum free light chain analysis revealed an elevated free lambda light chain, and a significantly decreased kappa/lambda light chain ratio. SPEP revealed a monoclonal spike of beta/gamma immunoglobulins. Fat pad biopsy confirmed AL amyloidosis on Congo red staining and bone marrow biopsy revealed 5% monoclonal plasma cells. The patient was diagnosed with multiple myeloma and initiated on cyclophosphamide, bortezomib, and dexamethasone chemotherapy.

Conclusions

This case highlights the importance of physical exam findings and the heterogeneous array of diagnoses possible from relatively common findings. In this case, findings of a murmur and goiter led ultimately to diagnoses of multiple myeloma and cardiac AL amyloidosis. Had clinical evaluation of her murmur not been undertaken, she would have remained on dangerous pre-load depleting diuretic therapy with obstructive cardiomyopathy during surgical resection of her thyroid. While her AL amyloid cardiomyopathy caries a poor long term prognosis, early recognition and treatment likely led to significantly improved quality and quantity of life.

Congenital Renal Arteriovenous Malformation: A Rare but Treatable Cause of Hypertension

Nicholas Isom, M.D., Reza Massomi, M.D., Adam Alli, M.D., Kamal Gupta, M.D.
KU School of Medicine-Kansas City

Introduction

Congenital renal vascular anomalies are rare yet important potential causes of significant morbidity. They have been classified into three categories: cirsoid, angiomatous, or aneurysmal. These classifications are based on the size, location, and number of vessels involved. This case illustrates the diagnosis and unique treatment of this rare etiology of secondary hypertension.

Case Presentation

A 29-year-old Caucasian female was seen in the hypertension clinic as a referral from the high-risk obstetric clinic for management of hypertension (HTN). She was contemplating pregnancy. She had been diagnosed with HTN at age 19 and had been on antihypertensive medications (was on Methyldopa 500 mg BID) with uncontrolled HTN. She had an extensive secondary cause work-up that was reported as normal (ruling out Pheochromocytoma, hyperaldosteronism, Cushing's, and thyroid disorders). Renal artery duplex done elsewhere was reported as moderately increased left renal artery peak systolic velocity but normal renal to aortic ratio (ARR) not suggestive of renal artery stenosis. Our review of the images showed that the peak proximal left renal artery velocity was 225 cm/sec with ARR of 1.3. However, the diastolic flow was especially prominent in the proximal left renal artery as compared to the right side and color Doppler US image of the left kidney showed a large diameter vascular structure at the hilum that had not been interrogated with spectral Doppler. An MRA was done that showed a large left renal upper pole arteriovenous malformation (AVM) with associated vascular shunting and early opacification of the left renal vein. Since the patient had no history of trauma or surgery, this was considered likely to be a congenital AVM. Selective arteriograms identified a single origin from the proximal renal artery and this was closed successfully with coil embolization. Fortunately, the arterial branch involved was only perfusing a small part of the renal parenchyma and thus there was no significant renal injury. The patient's BP normalized within a few days and she came off her antihypertensive medications. At six-month follow-up, she remained normotensive off all medications and was discharged from the hypertension and high-risk obstetric clinics.

Conclusions

Aneurysmal malformations, such as this one, have a single (and dilated) feeding and draining vessel. The prevalence of renal AVMs is estimated at less than 0.04% making them rare causes of secondary hypertension. However, they respond well to embolization with resolution of hypertension in 59% of patients treated. This case illustrates that a careful review of duplex waveforms beyond just peak velocity and ratios is important to identify uncommon pathology.

Chronic Inflammatory Arthropathy in the Setting of Levamisole-Induced Vasculitis

Kyle Myers, D.O., Pooja Bhadbhade, D.O.
KU School of Medicine-Kansas City

Introduction

Levamisole, an anti-helminthic drug, is a common adulterant of cocaine with well-described auto-immune phenomena, most notably complications of ANCA-associated vasculitides. Though arthralgia is a relatively common symptom of levamisole-induced vasculitis, the specific association of levamisole toxicity and inflammatory arthropathy is less well-documented. This case describes the incidental findings suggestive of chronic, deforming, erosive arthropathy in the setting of a patient with inflammatory eye disease secondary to suspected levamisole-induced vasculitis.

Case Presentation

A 46-year-old woman with remote history of levamisole-induced skin necrosis and chronic cocaine use presented with right eye foreign-body sensation, eye pain, redness, and decreased vision that began abruptly four days prior to admission. She had associated fever, myalgia, night sweats, joint pain, swelling, and stiffness in her hands, elbows, and knees. She had history of laboratory-proven levamisole-induced skin necrosis six years prior from cocaine abuse. Her exam showed findings suggestive of chronic inflammatory arthropathy and a round ulcerative right ankle lesion. Ophthalmologic exam demonstrated anterior uveitis, posterior scleritis, dacryoadenitis, and ocular hypertension. Notable admission labs were cocaine on urine drug screen, elevated erythrocyte sedimentation rate, and C-reactive protein, positive anti-nuclear antibody with a titer of 1:160, positive p-ANCA with a titer of 1:1280, and elevated anti-myeloperoxidase at 0.7. Notably, WBC count was normal, urinalysis was without hematuria or proteinuria, and c-ANCA, PR3, complement 3 and complement 4 as well as complete extractable nuclear antigen panel were negative. Infectious workup was negative. Plain films of the hands showed findings consistent with inflammatory arthropathy. CT chest was without significant findings. Lacrimal gland biopsy demonstrated dacryoadenitis with foci of chronic inflammatory cells and biopsy of her right ankle lesion showed non-specific inflammation. She was treated with topical eye drops, acetazolamide, anterior chamber tap, and systemic steroids with improvement in her ocular symptoms and intraocular pressure. She was extensively counseled on stopping cocaine use.

Conclusions

Her presentation was thought secondary to either levamisole-induced vasculitis or idiopathic ANCA-associated vasculitis. Given the pattern of her autoimmune labs and history of levamisole-induced skin necrosis, levamisole was favored as the likely etiology of her inflammatory eye disease. Levamisole has many known immunological effects, including the induction of antibodies against various antigens resulting in a multitude of systemic complications. If levamisole can induce such manifestations as necrotic skin lesions, vasculitis, glomerulonephritis, and pulmonary hemorrhage, it is possible that over time it could also lead to an erosive arthritis, as in this patient. This case documents the possible relationship between levamisole and inflammatory arthropathy. Recognizing this correlation would have unique implications on treatment and prevention of associated systemic manifestations.

Burning Fat 24/7: Dangers of the Ketogenic Diet When Taking a SGLT-2 Inhibitor

Sukhindervir Sandhu, Jesse Richards, D.O., Candice Rose, M.D.
KU School of Medicine-Kansas City

Introduction

There are several pharmacologic treatments available to treat type 2 diabetes mellitus (T2DM). Sodium-glucose co-transporter 2 (SGLT-2) inhibitors are one of the commonly indicated medications because of their weight loss, cardiovascular benefits, and low risk of hypoglycemia. However, the purpose of this case is to recognize when popular diets can complicate use of these medications.

Case Presentation

A 38-year-old male with past medical history of T2DM, tobacco use, and pulmonary embolism presented to the emergency department with crushing chest pain that radiated down his left arm, with intermittent chest pain the last several weeks. The only medications he took were metformin and canagliflozin, the latter of which he started less than a week prior. Social history revealed that the patient was on a ketogenic diet, restricting carbohydrate intake while increasing fat consumption. His vitals and physical exam were benign, with the exception that the chest pain was reproducible on palpation of the anterior chest wall. He had normal troponins and myocardial perfusion. His complete metabolic panel indicated a low sodium of 131 mmol/L, elevated serum glucose of 127 mg/dL, decreased bicarbonate of 12 mmol/L, serum anion gap of 17, and was otherwise within normal limits. His lactic acid was normal. However, urinalysis indicated 3+ glucose, 2+ ketones, and 1+ protein. He had elevated beta-hydroxybutyrate at 9.3 mmol/L (normal < 0.3) even though his serum glucose levels remained below 200 mg/dL for most of his two-day hospital course. Finally, an arterial blood gas of 7.21/25/112 indicated anion gap metabolic acidosis with appropriate respiratory compensation. His history and labs were suggestive of euglycemic diabetic ketoacidosis (DKA). He was successfully treated with an insulin drip coupled with D5, and later D10 drips and frequent potassium replacement that resolved his chest pain while decreasing his serum beta-hydroxybutyrate and serum anion gap.

Conclusions

This case suggests practitioners should be cautious in prescribing SGLT-2 inhibitors for type 2 diabetics on the frequently encountered ketogenic diet. SGLT-2 inhibitors decrease reabsorption of glucose at the renal tubules while promoting fatty acid oxidation and ketone bodies formation as an alternative energy source. The ketogenic diet promotes ketosis without significant changes in blood pH, plasma bicarbonate, or blood glucose in comparison to DKA because of the intentional restriction of carbohydrate intake with increased consumption of dietary fats. It is recognized that the harmless ketosis caused by SGLT-2 inhibitors can become harmful DKA when coupled with factors like postoperative status or poor nutrition, especially given that this patient tolerated SGLT-2 inhibitors for six months without issue before starting the diet. Further research is needed on the topic, and in the interim mindfulness of euglycemic DKA is critical in this population.

Gender Disparities in Secondary Preventive Measures for Atherosclerotic Cardiovascular Disease in Kansas

Roshni Jain, Tabitha Muutu, M.D., Paul Ndunda, M.D.
KU School of Medicine-Wichita

Introduction

In Kansas, cardiovascular disease (CVD) is the leading cause of death. Heart disease and stroke caused 6,988 deaths in 2015, making Kansas' death rate from CVD the 23rd highest in the country. More than 47% of heart attack survivors and 30% of stroke survivors suffer recurrent events, which highlights the need for data on secondary preventive care. There is evidence from national data of gender disparities in CVD morbidity, mortality, and preventive medication use. However, data are lacking on the disparities in secondary prevention in Kansas. The objective of this study was to assess the gender differences in lifestyle secondary preventive measures in patients from Kansas with CVD.

Methods

This study is an analysis of the 2015 Behavioral Risk Factor Surveillance System (BRFSS), a chronic disease and behavioral risk factor survey conducted by the CDC. The sample included 2,686 subjects from Kansas with a history of coronary heart disease and stroke. The demographic characteristics were analyzed using chi square and 2-sample t-test. The secondary preventive measures tested included: exercise, diet, smoking cessation attempt, alcohol intake, body mass index, and use of blood pressure medications in hypertensive patients. Gender disparities were analyzed using logistic regression analysis. Among the covariates tested age, race, education, and income showed significant effects on the model and were adjusted for in the outcomes.

Results

Women comprised 48.7% of the CVD cohort with no significant differences in race proportions between genders. The median ages of the study subjects were 68 and 72 years for men and women, respectively. Women were less likely to have ever smoked (47.5% vs 66.8% OR 0.44 (0.35, 0.55) $p < 0.0001$) and more likely to have made a smoking cessation attempt (70.7% vs 54.7% OR 2.35 (1.33, 4.15) $p = 0.003$). There was no difference in the odds of being current smokers (17.1% vs 17.8% OR 1.17 (0.96, 1.49) $p = 0.213$). Women were less likely to meet the AHA aerobic and muscle strengthening exercise guidelines (11.9% vs 17.4% OR 0.71 (052, 0.95) $p = 0.023$), but they were more likely to eat ≥ 4 servings of vegetables per day (OR 1.66 (1.06, 2.60) $p = 0.026$) and ≥ 4 servings of fruit per day (OR 1.83 (1.05, 3.17) $p < 0.032$). Women were more likely to have a BMI < 25 (1.79 (1.47, 2.18) $p = 0.0001$), and hypertensive women were more likely to be on antihypertensive medications (OR 1.636 (1.033, 2.591) $p = 0.036$). There was no difference in heavy alcohol use (OR 0.813 (0.472, 1.399) $p = 0.45$).

Conclusions

Patients with CVD in Kansas do not meet AHA lifestyle modification recommendations, and there are significant gender disparities in adherence to the studied CVD secondary preventive measures.

Treatment of Refractory Acute Myeloid Leukemia with Isocitrate Dehydrogenase Inhibitor

Aarati Keshary, M.D., Jeremy M. Deutsch, M.D.
KU School of Medicine-Wichita

Introduction

Acute myeloid leukemia (AML) is a disorder of poorly differentiated myeloid precursors that accumulate in the bone marrow and interfere with the production of normal cells. The overall prognosis with a five year survival rate is 22%. This prognosis can be affected by the presence or absence of certain mutations. The Isocitrate Dehydrogenase (IDH) gene is involved in cellular metabolism and in epigenetic regulation in the Krebs cycle. Mutations in IDH1 or IDH2 are detected in approximately 20% of patients with AML after patients undergo additional genotyping. These mutations catalyze alpha-ketoglutarate leading to DNA hypermethylation, altered gene expression, and blocking of hematopoietic differentiation of myeloid progenitor cells due to overproduction of 2-hydroxyglutarate, an oncometabolite, promoting leukemogenesis. A drug has been developed for the treatment of IDH-2 and IDH-1 mutations and is undergoing clinical trials. We present a case of refractory AML with IDH-1 mutation that achieved remission with an IDH-2 mutation inhibitor, Enasidenib.

Case Presentation

A 53-year-old female with a history significant for hypothyroidism was seen for evaluation of pancytopenia. Bone marrow (BM) biopsy confirmed AML with more than 90% blast cells. She was admitted for initiation of induction chemotherapy with idarubicin and cytarabine. Following the first induction, a repeat BM biopsy showed refractory AML. She underwent a second induction with Mitoxantrone and VP16. Repeat BM biopsy showed hypocellular marrow with 18-20% residual AML. She developed a diffuse erythematous rash that was thought to be due to Cytarabine. Given that the patient failed two induction therapies, she was approved for Enasidenib. The rash continued to worsen with ulceration. Biopsy revealed septate fungal hyphae and blood culture was positive for Fusarium species. Infectious disease recommended treatment with ambisome, vancomycin, and meropenem. The patient achieved complete remission on Enasidenib as evidenced by repeat BM which showed less than 5% blast cells. She developed complications such as acute renal failure, elevated liver enzymes, and neutropenic fever which resolved. She was discharge on day 61 with three-month remission.

Discussion

AML with IDH1 and IDH2 mutations are rare with IDH1 mutation being rarer. However, prognosis based on mutation type in AML remains controversial. Many cases have been refractory to a traditional chemotherapy regimen. Enasidenib was developed specifically for the treatment of refractory and relapsing AML cases such as ours. It is a selective targeted inhibitor of the mutant IDH2 enzyme that is approved for the treatment of refractory or relapsing AML with IDH2 mutations. However, we present a case of AML with IDH1 mutation that showed complete remission on Enasidenib. Further investigation is needed to assess the efficacy of Enasidenib as part of a first line regimen for the treatment of AML with IDH mutations.

Maintaining a High Suspicion for Ehrlichiosis in the Heartland

Mejalli Al-Kofahi, M.D., Usman Nazir, M.D.
KU School of Medicine-Kansas City

Introduction

Ehrlichiosis is a rare tick-borne illness with increasing incidence. In 2016, there were 1,377 cases, 50% of which reported from four states that included Missouri. Clinical presentation is non-specific and difficult to distinguish from other etiologies. We present a case with concurrent enteropathogenic Escherichia coli (EPEC) infection confounding the clinical picture leading to further evaluation to diagnose Ehrlichiosis.

Case Presentation

A 43-year-old male with a past medical history of colon cancer and ulcerative colitis status post hemi-colectomy presented with a one-week history of myalgias, fevers, nausea, vomiting, confusion, watery diarrhea, and ten-pound weight loss after a recent diagnosis of EPEC. He had frequent tick bites removed regularly by his wife. He lived in a ranch close to Joplin, Missouri raising livestock. Pertinent positives on physical exam included tachycardia, hypotension, injected bilateral conjunctiva, and diaphoretic appearance with erythema over his abdomen. Laboratory work showed hemoglobin 12.3 g/dl, platelet count 114 K/UL, lactate dehydrogenase (LDH) 1084 U/L, haptoglobin 219 mg/dl, fibrinogen 146 mg/dl, creatinine 7.68 mg/dl, aspartate aminotransferase (AST) 329 U/L, alanine aminotransferase (ALT) 191 U/L, and alkaline phosphatase 168 U/L. Computer tomography (CT) of head, abdomen, and pelvis was positive for gallbladder wall thickening.

The patient started on vancomycin, meropenem, and doxycycline, and aggressively resuscitated without requiring vasopressors. Peripheral smear showed one schistocyte per high power field, and ADAMTS13 was 58% (normal range > 70%). Acute hepatitis panel, fungitell, blood, and urine cultures, serum shiga toxin, urine eosinophilic stain, and serum antibodies against Leptospira, Brucella, and Babesia were all negative. Ehrlichia IgG titer was 1:256 with a reference range of < 1:64. Serum PCR of Anaplasma phagocytophilum, Ehrlichia chaffeensis, Ehrlichia ewingii/canis, and Ehrlicia muris like were all negative. Diagnosis of Ehrlichiosis was made and symptoms completely resolved within five days. He was discharged to complete a total of 14 days of doxycycline.

Conclusions

Missouri and Kansas are listed by the CDC as endemic areas for E. chaffeensis, which is one of the most common etiologies of Ehrlichiosis. Thus, internists in those states should keep Ehrlichiosis on the differential diagnoses with patients who have non-specific flu-like illnesses, and unexplained thrombocytopenia, leukopenia, elevated liver enzymes, and LDH, especially in patients who have history of tick bites. Our case was confounded by the recently diagnosed EPEC that could explain the severe diarrhea and AKI, but not the high-grade fevers and other lab abnormalities. Diagnosis of Ehrlichiosis should be based on clinical suspicion as the sensitivity of serum PCR rapidly declines with treatment. Antibody titer against E. Chaffeensis antigen could assist in the diagnosis if a four-fold increase is seen within the first week of symptoms, or four fold decrease is seen within two to four weeks after treatment. First line treatment is doxycycline that should be started prior to diagnostic confirmation.

A Rare Cause of Anemia:
74-year-old Male with High-Grade Neuroendocrine Tumor of the Colon

Nicole Balmaceda, Mohammad Telfah, M.D., Mazin Al-Kasspooles, M.D., Hongyan Dai, M.D., Ph.D., Prakash Neupane, M.D., Joaquina Baranda, M.D.
KU School of Medicine-Kansas City

Introduction

Colorectal cancer is the third most common cause of malignancies in both men and women, with the overwhelming majority being adenocarcinoma. An exceptionally rare histology type of colon cancer is high-grade large cell neuroendocrine tumors. Highly aggressive, these rare malignancies present challenges in treatment.

Case Presentation

A 74-year-old male presented to his primary care physician with weight loss, poor appetite, fatigue, and diffuse abdominal pain in 2018. Review of systems was negative for nausea, vomiting, change in bowel movement, blood in the stools, shortness of breath, or flushing. Past medical history includes hypertension and chronic kidney disease. On examination, he was pale but had no organomegaly. His hemoglobin was 8 gm/dL and creatinine was elevated. Bone marrow biopsy and urine studies were consistent with monoclonal gammopathy of undetermined significance. Five months later, the patient underwent colonoscopy which showed a large mass within the proximal transverse colon at the hepatic flexure. Biopsy of the colonic mass showed poorly differentiated invasive adenocarcinoma. Because of renal insufficiency, a CT with contrast could not be performed. Therefore, PET/CT imaging was done which demonstrated a hypermetabolic colonic mass with a maximum SUV of 25.9, compatible with primary malignancy, hypermetabolic mesenteric lymphadenopathy consistent with nodal metastatic disease, and multiple hypermetabolic mediastinal, hilar, and periportal lymph nodes of indeterminate significance. The patient underwent laparoscopic extended right hemicolectomy. Intraoperatively, suspicious-appearing enlarged central lymph nodes were noted. Surgical pathology revealed high-grade large cell neuroendocrine tumor with proliferative index of over 90%. He was initially staged as III T3N2aM0. Because of this unexpected pathology associated with rapid progression, a baseline PET/CT was done, and systemic therapy planned. There were lung lesions too small to characterize, but no definite evidence of metastatic disease in the chest. However, this showed abdominal lymphadenopathy consistent with metastatic disease. Liver function tests, serum CEA, and urine 5-HIAA/CP were within normal limits. Chromagranin A was elevated at 315. The patient was started on carboplatin and VP-16 three weeks post-operatively with plans to complete four to six cycles of this regimen.

Conclusions

Neuroendocrine tumors account for only 0.4% of all colorectal neoplasms. Current literature on optimal treatment strategies for large cell neuroendocrine tumors of the colon is sparse. Given the histologic similarities, these colonic malignancies are managed with the same regimens used in small cell lung cancer. However, emerging data suggest that poorly differentiated neuroendocrine tumors are a distinct disease entity from small cell lung cancer. A more thorough investigation of large-cell neuroendocrine tumors of the colon is warranted.

Sudden Cardiac Arrest in Late Adolescent with WPW Associated to Anabolic-Androgenic Steroid Abuse

Maher Bazzi, M.D., Bassem Chehab, M.D.
KU School of Medicine-Wichita

Introduction

Anabolic-androgenic steroids (AAS) have been implicated in multiple metabolic and cardiac abnormalities that can lead to disastrous events even in young healthy individuals. Reported here is a case of a late adolescent recently diagnosed with Wolff-Parkinson-White (WPW) syndrome presenting with sudden cardiac arrest following ventricular fibrillation in the setting of AAS abuse. This case highlighted the metabolic and imaging findings that may have contributed to the near-fatal presentation.

Case Presentation

A 19-year-old previously healthy male with recent diagnosis of WPW presented to an outside ED following loss of consciousness and cardiac arrest witnessed by his girlfriend. She initiated CPR for 10 minutes prior to EMS arrival. In the ED, he was found to be in ventricular fibrillation and ACLS protocol was performed, receiving two doses of epinephrine and a single shock prior to return of spontaneous circulation, at which point hypothermia protocol was initiated. He was intubated and sedated in the ED, then transferred to ICU. ABGs on admission showed mixed metabolic and respiratory acidosis. He was found to be in shock, requiring initiation of norepinephrine and vasopressin. Echocardiogram showed severely hypokinetic left ventricle with ejection fraction of 20-25%. Left heart catherization was performed and showed a co-dominant system with no significant coronary disease. Ventriculography showed a 15-20% ejection fraction. An intra-aortic balloon pump (IABP) was placed due to cardiogenic shock to help with pressure support. His troponin on admission was 1.06 and peaked within hours at 13.15. On day two, pressors were discontinued. IABP was removed by day three and paralytics were stopped. The patient was extubated and started on NIPPV for fluid overload. On day four, chest x-ray showed worsening pulmonary congestion and was started on diuretics. On day five, he was switched to high-flow nasal cannula at 6L with good urine output. He was eating, awake, and comfortable; the patient's only complaint was short term memory loss. On day six, a cardiologist performed an EP study and showed left lateral pathway which was ablated. Repeat echocardiography on day seven showed concentric thickening with remodeling and estimated ejection fraction of 40-45%. Due to improved function, ICD placement was not indicated. On day eight, the patient was discharged home on aspirin and metoprolol, without need for oxygen.

Conclusions

This case illustrates the harmful effects of AAS with metabolic and structural changes including dyslipidemia and hypertrophic cardiomyopathy, which increases the risk for fatal arrhythmias. Previous literature is scarce on connecting AAS to sudden cardiac arrest, mostly related to arrhythmias and rarely related to ischemic heart disease. Awareness for AAS abuse with its associated short- and long-term effects should always be discussed, not only with professional athletes and bodybuilders, but also with young individuals that use steroids for aesthetic purposes.

Diffuse Bone Pain after Lung Transplant: Cystic Fibrosis Arthropathy or Voriconazole Induced Periostitis Deformans

Amna Batool, MBBS, Megan Krause, M.D.
KU School of Medicine-Kansas City

Introduction

Voriconazole is a fluoride-containing anti-fungal medication commonly prescribed to organ transplant recipients. Periostitis deformans is one of its uncommon side effects characterized by diffuse bone pain, elevated alkaline phosphatase, dense, irregular, bulky periosteal new bone formation on radiographs/CT, and increased uptake on Tc-99m MDP bone scan.

Case Presentation

A 34-year-old Caucasian female with cystic fibrosis status post lung transplant presented for evaluation of joint pain. She had developed polyarticular pain along with stiffness and limitations in range of motion three months after the transplant. There was no associated history of morning stiffness. Prior to transplant, she had a diagnosis of cystic fibrosis arthropathy which had responded well to hydroxychloroquine which had been restarted without any improvement. Physical exam was remarkable for significantly limited range of motion of the neck and other joints due to pain without any synovitis. Of note, she was on voriconazole for heavy growth of trichosporon in sputum. Laboratory work showed alkaline phosphatase elevated at 161 U/L, ANA elevated to 1:320, however, other serologies were within normal limits. Creatine kinase was 35 U/L. White blood cell count was normal. Rheumatoid factor was negative. Joint survey did not demonstrate any erosions. There was evidence of cortical thickness of the left medial femur without any history of trauma to that area. Given lack of improvement of symptoms with hydroxychloroquine, normal serologies, lack of inflammatory symptoms, and findings on radiological imaging, voriconazole-related periostitis was suspected and this medication was discontinued. She reported improvement of symptoms soon after discontinuation of the medication. Her alkaline phosphatase levels also normalized after stopping the medication.

Conclusions

Voriconazole-induced periostitis is commonly seen in immunosuppressed post-transplant patients especially lung transplant patients on chronic voriconazole therapy. Most common side effects of voriconazole are rash and elevated transaminases. Voriconazole is like fluconazole, however, it contains a fluorinated pyrimidine which is thought to be the etiology of this condition. Fluoride stimulates bone formation by activating osteoblasts. It has been reported that patients on at least six months or longer of chronic voriconazole therapy have elevated fluoride levels. Patients commonly present with diffuse severe bone pain, universally elevated alkaline phosphatase levels and dense, irregular, bulky periosteal new bone formation on radiographs/CT. It is also associated with increased uptake on Tc-99m MDP bone scan. Other differentials of diffuse periostitis include hypertrophic osteoarthropathy, thyroid acropachy, and other medications (vitamin A, prostaglandin). Treatment consists of discontinuation which leads to prompt improvement of symptoms. It is an uncommon condition, however, a thorough history and physical exam is very helpful in including or excluding most of these diagnoses and expediting a timely referral.