USA-Acquired Tungiasis: Late Bacterial Infection Complicating Remote Tunga penetrans Infestation

Mansee Desai BA1, Michael W Graham MD1,2, Nikole M Scalera MD1,2, Joseph P Myers MD1,2
1. Northeast Ohio Medical University, Rootstown, OH; 2. Department of Medicine, Summa Akron City Hospital, Akron, OH

Tungiasis is an infestation caused by Tunga penetrans usually acquired and diagnosed in South or Central America.1,6,7 We report a case acquired after travel to Florida and complicated by late secondary bacterial infection diagnosed in Ohio.

Clinical Course

41-year-old man without significant PMH was married 5 years ago on the beach at Sanibel Island, FL. Patient was horribly bitten by sand fleas over both lower extremities. The entire wedding party had similar bites, but the patient’s lesions took weeks to resolve, longer than anyone in the wedding party. His lesions finally healed, but he noticed one bitten area remained firm, raised and fleshy in color. In January 2018, the lesion began to protrude from the skin. The patient applied frankincense oil on the lesion resulting in erythema and skin breakdown. He pulled out two small calcific pieces from the open wound and the area became more erythematous and started draining purulent material.

Clinical Course (Cont’d)

- Patient was treated with TMP/SMX & cephalexin for cellulitis.
- Extraction of the calcific mass in his internist’s office was unsuccessful so patient was admitted to hospital.
- Vitals: T=99.3F, BP=150/87 HR:69bpm, RR:18/min
- PE: Negative except for shaking chills along with erythema and purulent drainage surrounding a protruding white calcific mass on the left lower leg
- Labs: WBC = 10,900 cells/mm³ with 78% granulocytes.
- Imaging: X-Ray of leg showed 1.5 cm-diameter ill-defined rounded calcified lesion superficially 15 cm above the level of the left ankle joint. (Figure 2.)
- Cultures: Methicillin-sensitive Staphylococcus aureus and a Proteus mirabilis resistant only to tetracycline
- Procedure: Patient had surgical removal at bedside (Figure 3.)
- Pathology: Calcified tissue measuring 2.0 x 1.4 x 1.0 cm
- Patient treated with IV vancomycin & ampicillin/sulbactam pending culture results; discharged on PO amoxicillin/clavulanate.
- The wound healed completely in 2 weeks.

Discussion/Conclusions

- Tungiasis is an ectoparasitic infection caused by penetration of the female sand flea.3,6,7
- The flea Tunga penetrans burrows into the epidermis of animal host. The flea then undergoes a complex 5-stage developmental sequence with varying degrees of host inflammatory response to each stage.3,4,5,6,7
- Tungiasis acquired in the United States is rare.2,7,8
- Secondary bacterial infection is common.2,5
- Primary care physicians should be aware of this rare but irritating ectoparasitic infestation.

References

Prostatic abscesses may present insidiously and are often misdiagnosed in anuric or oliguric patients without urinary symptoms. Treatment guidelines are not well established. Here we present a case of a patient found to have a large prostatic abscess in the absence of urinary symptoms that was likely undetected for months.

A 72-year-old man with insulin-dependent diabetes, ESRD on hemodialysis, and bladder outlet obstruction (performing weekly self catheterization) presented to the hospital with one month of subacute fatigue and one day of vague lower abdominal pain. Over the past six months he had a rising white blood cell count (WBC) to 24 K/uL without any other signs or symptoms of infection. Serial abnormal urinanalyses were regarded as expected colonization/contamination given his oliguria, and urine cultures were not sent.

Admission vitals were notable for a temperature of 100.8°F; exam was without abdominal or CVA tenderness. Laboratory Data

<table>
<thead>
<tr>
<th>Initial labs (Day 1)</th>
<th>Repeat labs (Day 2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBC: 39K/uL (2% bands)</td>
<td>WBC: 59.8 K/uL (4% bands)</td>
</tr>
<tr>
<td>C-reactive protein (CRP): 353 mg/L</td>
<td></td>
</tr>
<tr>
<td>Urinalysis: 3+ leukocyte-esterase, &gt;100 WBCs</td>
<td></td>
</tr>
</tbody>
</table>

The patient was started on broad spectrum antibiotics (vancomycin and piperacillin-tazobactam), however his WBC rose to 59.8 K/uL on day 2. At this point, a CT chest/abdomen/pelvis was obtained to evaluate for occult sources of infection, and it revealed a 5.3 x 4.7 x 4.5 cm prostatic abscess. Interventional radiology performed CT-guided percutaneous drain placement through the left buttocks. Cultures from the drain and initial urine sample grew MRSA, and antibiotics were narrowed to vancomycin with dialysis. Repeat imaging showed an interval decrease in abscess size and the drain was removed after output stopped on day three. His WBC and CRP had downtrended to 23.2 K/uL and 143 mg/L respectively and he was discharged on day five for at least two weeks of vancomycin pending clinical resolution.

Prostatic abscess is a rare complication of bacterial prostatitis and can be difficult to diagnose clinically. Risk factors include diabetes, immunosuppression, HIV, urinary tract instrumentation, indwelling catheters, and bladder outlet obstruction. Oliguric or anuric patients are at higher risk due to poor urinary washout, and frequent abnormal urinanalyses may be disregarded as expected asymptomatic bacteriuria. Patients may present with fever, leukocytosis, pyuria, dysuria, and prostatic or perineal tenderness. Transrectal ultrasound (TRUS) is the preferred imaging modality, though contrast enhanced CT may also be used, particularly when extension into the ischiorectal fossa or perineum is suspected. While there are no formal treatment guidelines, reported approaches typically combine broad-spectrum antibiotics with surgical drainage, either by TRUS-guided percutaneous needle drainage, transurethral drainage, or open deroofing.

While it is generally wise to regard the results of an abnormal urinanalysis in anuric or oliguric patients with caution, prostatic abscess should be considered in patients with underlying risk factors and unexplained leukocytosis without an obvious infectious source. Further investigation is required to standardize medical and surgical management of prostatic abscesses.

References

**A Rare Cause of a Rare Disorder (Cardiac Amyloidosis)**

Georgina Kolcun, M.S.¹, Lawrence Rice, M.D.¹,²

¹ College of Medicine, Texas A&M Health Science Center, College Station, Texas
² Department of Medicine, Weill Cornell Medical College, Houston Methodist Hospital, Houston, Texas

**Introduction**

- Cardiac amyloidosis generally complicates one of four processes: light-chain (AL) amyloidosis, hereditary mutant transthyretin amyloidosis, senile transthyretin amyloidosis, or inflammation-related secondary (AA) amyloidosis.
- AL amyloidosis represents abnormal depositions of light chain immunoglobulin fibril aggregates.
- This plasma cell dyscrasia is generally isolated or associated with multiple myeloma (MM). We are caring for a patient with a surprising underlying disorder.

**Case Presentation**

- A 67 year old man was admitted for increasing shortness of breath due to acute on chronic systolic heart failure.
- He had become unable to climb stairs due to dyspnea and was experiencing lower extremity swelling for four months.
- He had no bone pain, neuropathy, or visual changes.
- Home medications were carvedilol, lisinopril, furosemide, and 2+ bilateral pitting edema.
- Physical exam showed no lymphadenopathy, normal heart sounds, decreased breath sounds at the lung bases, no hepatosplenomegaly, and 2+ bilateral pitting edema.

**Diagnostic Studies**

- Additional laboratory tests revealed a normal troponin, hypoalbuminemia (3.3 g/dL), and markedly elevated BNP (1677 pg/mL) and beta-2 microglobulin (5.6 mg/L).
- Chest X-ray showed cardiomegaly and pleural effusion.
- Echocardiogram indicated moderate left ventricular hypertrophy and impaired systolic (EF 35-40%) and diastolic functions.
- Cardiac MRI found biventricular heart failure and evidence of infiltrative cardiomyopathy.
- Endomyocardial biopsy demonstrated AL (lambda-type) amyloid deposition (positive Congo red stain; lambda by mass spectroscopy).
- Bone marrow showed diffuse amyloid deposits and 28% lymphoplasmacytic lymphocytes on Congo Red stain.
- Flow cytometry demonstrated lambda light chain restricted plasma cells (1%) and a lambda light chain restricted B cell population (9%).

**Imaging and Pathology**

- **A) Chest X-ray**
  - Cardiomegaly and left sided pleural effusion
- **B) Echocardiogram**
  - Moderate left ventricular hypertrophy and impaired systolic (EF 35-40%) and diastolic functions
- **C) Cardiac MR**
  - Global subendocardial late gadolinium enhancements (LGE)
- **D) Cardiac Tissue Histopathology**
  - Congo red stain shows diffuse amyloid deposits
- **E) Cardiac Tissue Histopathology under Polarized Light**
  - Cardiac tissue shows apple green birefringence
- **F) Bone Marrow Histopathology**
  - Bone marrow shows lymphoid aggregates with lymphoplasmacytic cells
- **G) Bone Marrow Histopathology**
  - Congo red stain shows positive proteinaceous deposits
- **H) Bone Marrow Histopathology under Polarized Light**
  - Bone marrow shows apple green birefringence
- **I) Serum Protein Electrophoresis**
  - Albumin, Alpha-1, Alpha-2, Beta, Gamma

**Diagnostic Studies (cont.)**

- SPEP corresponded to a 1.4 g/dL IgM-lambda monoclonal paraprotein.
- Serum free light chains showed increased lambda (245 mg/L, normal <26 mg/L) and decreased kappa: lambda ratio (0.04, normal >0.26).
- PCR was positive for MYD88 L265P mutation, typical of light-chain (AL) amyloidosis.

**Hospital Course and Management**

- Before cardiac involvement, this patient had no symptoms of WM.
- Since prolonged survival (median ten years) can be seen with WM, this patient was judged a candidate for orthotopic heart transplant, and received a chemoimmunotherapy regimen of rituximab, bortezomib, and dexamethasone.
- Cardiac transplant was performed after a 3 month waiting period and resulted in a drop in IgM and lambda free light chains to 0.2 and 63 mg/L, respectively.

**Discussion and Conclusion**

- Cardiac amyloidosis has been seen with plasma cell dyscrasias such as monoclonal gammopathy of undetermined significance or WM.
- Whereas MM cells overproduce IgG, IgA, or light chains, WM cells overproduce IgM.
- WM underlies only 5% of AL amyloid cases,¹ with cardiomyopathy present in 44% of these cases.²
- This case highlights the challenge of recognizing an atypical presentation of a silent disease before life-threatening complications develop.
- It is important to perform a thorough workup to define the underlying cause of a disease, which may affect subsequent prognosis and treatment.

**References**

Learning Objectives

- Identify signs and symptoms of organophosphate toxicity
- Recognize the potential for toxicity from incorrect use of common pest control products in an urban setting

Case Presentation

A 50 year old man presented to the emergency room with shortness of breath and a non-productive cough. Onset was 20 minutes prior while driving his van on a hot summer’s day with the air conditioning turned on high. He described chest tightness, dyspnea, and inability to clearly visualize the road. He also was experiencing nausea and generalized weakness.

Current Medical Conditions: morbid obesity, dyslipidemia, obstructive sleep apnea, and poorly controlled hypertension.

Social Hx: worked in construction; roofing, cement. Tobacco: ½ ppd Alcohol: 6-8 beers/day for 30+yrs Drugs(-)

Vital signs: Temperature: 37.6 °C Respiratory rate: 20 bpm Pulse: 80 bpm Blood Pressure: 153/87mmHg BMI: 50 kg/m²

Physical exam remarkable for generalized distress, lacrimation, pupillary constriction, and modest bibasilar crackles.

Initial Differential Diagnosis:
- Pulmonary embolism
- Myocardial infarction
- Allergy-type 1 hypersensitivity
- COPD exacerbation
- Toxicity from drugs or environmental exposure

Presenting Laboratory Studies
- CBC with differential: within normal limits
- Basic metabolic panel: within normal limits
- ABG: unremarkable, saturating well
- D-dimer: 0.62 µg/mL

Final Diagnosis: Organophosphate Poisoning

Organophosphates bind to acetylcholinesterase (AChE), an enzyme responsible for the degradation of acetylcholine, rendering it permanently dysfunctional. This causes an inundation of acetylcholine within the synaptic cleft, notably causing a large array of muscarinic symptoms: diarrhea, urination, miosis, bradycardia, bronchorrhea, emesis, lacrimation, and salivation.

Immediate decontamination with a 10 minute shower and placement on supportive O₂ therapy.

Poison control was contacted; because the toxin had infiltrated the van’s ventilation. They recommended that the vehicle be destroyed.

The patient was discharged in stable condition two days later.

Discussion

Organophosphate (OP) poisoning is a diagnosis that is made on clinical grounds and is a major cause of mortality in several counties. Exposure is commonly agricultural.

Due to the different potential modes of exposure to this toxin, a level of vigilance and clinical astuteness must be implemented to quickly diagnose and treat OP poisoning.

Pivotal aspects in management include immediate assessment and management of disturbances in the airway, breathing, and circulation.

Resuscitation and intubation may be necessary due to the compromise of airway patency, breathing mechanics, and hemodynamic stability that moderate to severe OP toxicity causes. Management with atropine or pralidoxime is indicated in the presence of severe bronchorrhea.

While organophosphate poisoning is uncommon in urban areas of the USA, it would be prudent to know all of the classic signs and symptoms, as well as be aware of the several modes of exposure to this toxin.

References

Double Trouble: Simultaneous Cutaneous Lymphomas in an AIDS Patient
Albert Jang, BS1, Lucy Huo, BA1, Mahmoud Gaballa, MD2, Robert Hester, MD, MS3, Nan Chen, MD2, Tejo Musunuru, MD2, Mark Udden, MD2, Martha Mims, MD, PhD2
1School of Medicine, 2Section of Hematology/Oncology, 3Department of Internal Medicine, Baylor College of Medicine, Houston, Texas

Background
- Plasmablastic lymphoma (PBL) is a rare aggressive subtype of diffuse large B-cell lymphoma, making up ~2% of all HIV-related lymphomas
- NK/T-cell lymphoma (NKTL) is an uncommon destructive and necrotic lymphoma, rarely seen in the general population, including HIV patients

Learning Objectives
- Understand that patients with HIV may develop more than one type of lymphoma
- Recognize the need to biopsy a new or worsening cutaneous lesion in the setting of a diagnosed cutaneous malignancy

Case Presentation
- 41-year-old woman with HIV (CD4 154/mm3, undetectable viral load), cutaneous plasmablastic lymphoma (diagnosed 5 months prior, s/p 4 cycles of EPOCH chemotherapy), and adrenal insufficiency presented with syncp
- Two weeks prior, patient noticed a new lesion on her right anterior thigh, similar to how her other PBL leg lesions started
- Had 4 days of subjective fever and increasing right thigh pain
- Treating two separate but concurrent aggressive malignancies, rare case of an AIDS patient presenting with two aggressive cutaneous lymphomas nearly simultaneously, not previously seen in the literature
- Unclear whether these two malignancies had a common origin
- Factors possibly contributing to developing two types of rare lymphomas in this patient include HIV-immunocompromised state and EBV infection

Discussion
- Plasmablastic lymphoma and NK/T-cell lymphoma are both hard to diagnose, requiring immunohistochemical markers
- Epstein-Barr virus is present in ~70% of all PBL cases (~75% of HIV+ PBL cases) and 100% of NKTL cases
- Well-known that EBV invades B cells, but less is known how EBV invades NK and T cells
- Absence of EBV excludes diagnosis of NKTL
- Because of their rarity and poor prognoses, there is no standardized treatment regimen for either lymphoma
- For PBL, EPOCH and bortezomib have been shown to prolong survival
- For NKTL, regimens including L-asparaginase and platinum-based agents have shown encouraging results

Conclusions
- Rare case of an AIDS patient presenting with two aggressive cutaneous lymphomas simultaneously, not previously seen in the literature
- Treating two separate but concurrent aggressive malignancies, each with no standardized regimen, is extremely difficult
- Physicians should maintain a high index of suspicion when a new lesion appears different from the others, and obtaining a new biopsy if possible should be done quickly

References
Hunting Down the Zebras: Hemophagocytic lymphohistiocytosis secondary to EBV-driven classical Hodgkin lymphoma

Alessandra Petrillo, Marygrace Zetkull MD, Lori Ann Leslie MD, Chinwe Ogedegbe MD

Department of Medicine, Hackensack University Medical Center

St. George’s University School of Medicine

Introduction

• Secondary hemophagocytic lymphohistiocytosis (HLH), a rare and life-threatening condition is triggered by autoimmune diseases, infections, and malignancies.1
• Presentation of this condition is often a result of uncontrolled immune activation affecting macrophages and natural killer cells. Excessive cytokine secretion and organ infiltration by lymphocytes and histiocytes results in organ tissue damage, hepatosplenomegaly, and multisystem organ failure.2
• Macrophages phagocytize host blood cells and platelets resulting in cytopenias.1
• Epstein-Barr virus (EBV) has been the most consistently reported virus associated with HLH.4
• If HLH is left untreated, severe neutropenia results in death secondary to bacterial or fungal infections.3

Laboratory Studies

| WBC | 3.3 | 3.5-10.0 x 10⁹/L |
| RBC | 1.87 (L) | 4.5-5.0 x 10¹²/mL |
| Hemoglobin | 6.2 (L) | 12.5-15.5 g/dL |
| Hematocrit | 15.3 (L) | 38-46% |
| Platelets | 243 | 135-420 x 10⁹/mL |

| BUN | 18 | 8-22 mmol/L |
| Cr | 1 | 0.8-1.5 mg/dL |
| Sodium | 139 | 134-146 mmol/L |
| Potassium | 2.5 (L) | 3.5-5.2 mmol/L |
| Chloride | 98 | 95-108 mmol/L |
| Bicarbonate | 34 (H) | 24-32 mmol/L |
| Total protein | 4.4 (L) | 6.8-8.3 mmol/L |
| Albumin | 2.2 (L) | 3.5-5.0 mmol/L |
| T. bilirubin | 1.3 (H) | 0.6-1.0 mg/dL |
| AST | 30 | 8-43 U/L |
| ALT | 51 (H) | 14-45 U/L |

Table 1. Data of admission laboratory studies including complete blood count, metabolic panel, iron studies, and coagulation profile.

Case Presentation

Patient was a 54-year-old Dominican woman was admitted to a hospital for a four month history of relapsing fever, weight loss, and symptomatic anemia. She reported three months of right upper quadrant pain, constipation and vomiting. She was evaluated for symptomatic anemia at another hospital, where she had a colonoscopy/endoscopy and at her home country, Dominican Republic, where she received four blood transfusions. She was discharged with a diagnosis of liver infection and told to have a bone marrow biopsy when returning to United States.

Both adult T cell lymphoma and Hodgkin lymphoma were considered. Despite empiric utilization of dexamethasone and cyclophosphamide, on hospital day 13, patient suffered from cardiac arrest secondary to septic shock confirmed mixed bacteremia with resistant organisms. This patient had reactivation of EBV driven stage IV classical Hodgkin lymphoma accurately diagnosed after lymph node biopsy on autopsy.

On admission ER patient was anxious, ill appearing, tachycardic with flow murmur, abdominal distension, and periumbilical tenderness. Laboratory findings included: bilirubin, and elevated LDH. Patients whom are at high risk, develop PRES, which was incidentally found on this patient’s MRI.1

Therapeutic Management

• Adult HLH can be diagnosed if five of the eight clinical findings exist; fever >38.5°C, splenomegaly, peripheral blood cytopenias, absolute neutrophil count <1000/microL, hypertriglyceridemia, hypofibrinogenemia, hemophagocytosis in bone marrow, spleen, lymph node, or liver, low or absent NK activity, ferritin >500 ng/mL, or elevated soluble CD25 two standard deviations above age-adjusted laboratory specific norms.1
• Supportive evidence includes cerebral symptoms, transaminitis, elevated fibrinogen, and elevated LDH. Patients whom are at high risk, develop PRES, which was incidentally found on this patient’s MRI.1
• The patient had a fulminant case of HLH two weeks prior to death; therefore timing of diagnosis of both HLH and underlying cause were crucial.
• Patient was hospitalized three times prior to this definitive diagnosis leading to the delayed diagnosis of Hodgkin lymphoma. This delayed diagnosis was an effect of its atypical presentation; minimal clinical lymphadenopathy and rapid systemic decline.

Tips for the Internist:

• Similar to SLE and syphilis - always add to your differential
• Travel/ exposure history as well as family history key
• Obtain biopsies quickly, prior to steroids if possible
• Review case personally with pathologist
• Outcomes are poor overall, mostly due to delayed diagnosis/initiation of therapy

References

1. McClain, K.L. Etkin, O. Clinical features and diagnosis of hemophagocytic lymphohistiocytosis. UpToDate. 2017
Background

- Calciphylaxis is the result of arteriolar calcification and occlusion causing inadequate perfusion of subcutaneous and dermal tissue
- Most commonly found in patients with end-stage renal disease
- Non-uremic associations include: hyperparathyroidism, malignancy, alcoholic liver disease, connective tissue disease, obesity, female sex and treatment with medications including warfarin, corticosteroids, iron, calcium and vitamin D

This case illustrates the use of radiography to assess the degree of improvement in microvascular diameters by measuring involved vessel diameters

Case Report

HPI:
71-year-old Hispanic female presented with a five-month history of progressively worsening lower extremity wounds

PMH:
DM II, Atrial Fibrillation, Nonischemic Cardiomyopathy

Medications:
Prednisone, Apixaban, Insulin

Differentials:
Calciphylaxis vs Cellulitis Vs Vasculitis Vs DVT

Diagnoses:
ANA, ANCA, cryoglobulin negative
Hep C Ab positive, Hep C viral load negative
Lower Extremity Doppler negative for DVT

Treatment:
Sodium thiosulfate started at time of diagnosis, surgical debridement and negative pressure wound therapy

Figure 1. (A) Large, medial upper thigh, full-thickness necrotic ulcer covered with black necrotic eschar. More laterally, a deep smaller necrotic full thickness ulcer representing non-healed initial biopsy site both overlying a painful, indurated, purple, hard base. (B) Three month follow-up demonstrating resolution of the ulceration with improvement of underlying, non-tender induration

Figure 2. (A) Mammographic evaluation of the right proximal thigh demonstrating numerous calcified vessels in a mesh-like configuration within the subcutaneous tissues. Involved calcified vessels measured as small as 0.1 to 0.2 mm in diameter. (B) Three month follow-up imaging revealed a reduction of involved calcified small vessels with those involved measuring as small as 0.4 mm in diameter. Large arterial atherosclerotic-type calcifications persist in the femoral artery.

Discussion/Conclusion

NUC should be considered when evaluating suspicious skin lesions in patients with predisposing conditions even in the absence of ESRD

Radiologic imaging is a useful non-invasive diagnostic tool allowing for quicker diagnosis, and eliminating the risks associated with tissue biopsy

Mammography is superior to plain radiography in the evaluation of extremity soft tissues. Mammography allows for a more accurate determination of small calcifications

This is the first case to demonstrate improvement of the disease objectively by measuring the diameter of involved calcified vessels with radiography

Future studies may determine whether the change in size or number of involved calcified vessels throughout treatment will serve as a prognostic tool in the course of the disease

References


Figure 3. Biopsy specimen demonstrates acute cellulitis, fat necrosis and deep seated blood vessels with intimal proliferation and calcification of vessels of diameter 1-2 mm confirming calciphylaxis

Non-Uremic Calciphylaxis: Using Radiography to Aid in Diagnosis
Matthew Cornacchia BSc, Aishwarya Vyasa-Lahar MD, Jonathan Kandiah MD, Dave Munger MD, Emanuela Sangeorzan MD, Charles Halasz MD
Ross University School of Medicine; Norwalk Hospital affiliated with Yale School of Medicine
Lung Impedance: A Novel Technique to Assess the Clinical Status of Heart Failure Patients

Daniel Kapustin1, Kosalan Aklan1, Simcha Meisel2, Michael Kleiner Shochat2
1University of Toronto Faculty of Medicine, Toronto, Canada
2Hillel Yaffe Medical Center, Hadera, Rappaport School of Medicine, Technion, Haifa, Israel

Background
Readmissions for heart failure (HF) are a major healthcare burden. Using a new, non-invasive surface electrode device, we have demonstrated that lung impedance (LI), a marker of pulmonary congestion, is strongly related to patient outcomes in HF. The aim of this study was to assess correlations between both NT-proBNP and LI with New York Heart Association (NYHA) class in patients with HFrEF and HFpEF.

Methods
This data is a secondary analysis of the data collected in the IMPEDANCE HF-reduced and IMPEDANCE HF-preserved trials. In both studies, patients were randomized to receive Lung Impedance-guided therapy or standard therapy for heart failure. The sample size used in our analysis included 285 patients with HFrEF and 67 patients with HFpEF. Patient data regarding NYHA class, NT-proBNP, and lung impedance (LI) was collected simultaneously during the initial study entrance visit, monthly outpatient clinic visits, and any hospitalizations for HF. Baseline LI for each patient was calculated upon entry to the study and was used to calculate a new parameter, the \( \Delta LIR = \left( \frac{\text{current LI}}{BLI} - 1 \right) \times 100\% \).

Results
Patient demographics are presented in table 1. A total of 615 measurements were obtained in the HFpEF group and 18,496 were obtained in the HFrEF group. Among HFrEF patients, increased NYHA class was associated with significantly increased serum NT Pro-BNP and significant decreases in measured \( \Delta LIR \) (Figure 1a and 1b). In the HFpEF group, patients presenting with NYHA class IV demonstrated increased serum NT-ProBNP, and patients with NYHA classes III and IV showed decreased \( \Delta LIR \) (Figure 2a and 2b). \( \Delta LIR \) showed greater correlation to NYHA class than NT-proBNP in both HFrEF (R² = 0.73, -0.31 respectively) and HFpEF patients (R² = -0.31, 0.1 respectively).

Conclusion
Our data demonstrates that NT pro-BNP and LI are efficacious biomarkers for monitoring patient functional status in HFrEF and HFpEF. Additional studies are necessary to further validate the sensitivity of these markers in HFpEF patients.

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>HFrEF</th>
<th>HFpEF</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>69.3 ± 11.7</td>
<td>74.7 ± 8.5*</td>
</tr>
<tr>
<td>Male (%)</td>
<td>86</td>
<td>47*</td>
</tr>
<tr>
<td>BMI (kg/m²)</td>
<td>29.1 ± 4.9</td>
<td>33.7 ± 5.2*</td>
</tr>
<tr>
<td>LVEF (%)</td>
<td>30.1 ± 7.0</td>
<td>58.8 ± 8.1*</td>
</tr>
<tr>
<td>Septum thickness (cm)</td>
<td>1.1 ± 0.2</td>
<td>1.3 ± 0.3*</td>
</tr>
<tr>
<td>LVPW (cm)</td>
<td>1.0 ± 0.2</td>
<td>1.1 ± 0.2*</td>
</tr>
<tr>
<td>LA area (cm²)</td>
<td>25.1 ± 5.9</td>
<td>23.6 ± 5.4</td>
</tr>
<tr>
<td>ΔLIR (%)</td>
<td>-24.6 ± 14.8</td>
<td>-19.9 ± 15.7*</td>
</tr>
</tbody>
</table>
Methylene Blue as a Rescue Therapy in the Treatment of Pressor-resistant Distributive Shock

Marc Incitti, MS, CCRN-CMC, CEN; Garrison Davis, BS, RRT; Tanya Meyers, BSN,CCRN; Michael Suarez, MS; Khaled Sorour, MD
1Geisinger Commonwealth School of Medicine, 2Signature Healthcare Brockton Hospital, 3Philadelphia College of Osteopathic Medicine
4Harvard Medical School

Introduction

Seen by most premedical students in the blue bottle experiment, methylene blue is not just a popular teaching prop to demonstrate reduction-oxidation reactions. As a purely synthetic medication, methylene blue has been used almost exclusively in the operating room and the emergency room for identification of urinary tract structures during surgery and as a reducing agent in cases of methemoglobinemia, respectively. Therapeutic use of methylene blue for circulatory shock has been studied in few small randomized controlled trials and case series with consistent reduction of pressor requirements in septic and anaphylactic shocks, but no mortality benefit had been documented. The efficacy of methylene blue for circulatory shock has not been evaluated in the intensive care unit. Methylene blue’s proposed mechanism during distributive shock is through the inhibition of both nitric oxide (NO) synthesis and cGMP synthesis. Recall that cGMP lowers intracellular calcium concentration in smooth muscle by shifting calcium ions into the sarcoplasmic reticulum. Nitric oxide can upregulate guanylyl cyclase, the enzyme responsible for converting GMP to cGMP. By blocking the action of both NO and guanylyl cyclase, the increased cytoplasmic calcium concentration promotes vasculature smooth muscle contraction. Another mechanism may be through the inhibition of catecholamine-O-methyltransferase which is responsible for the degradation of intrinsic and extrinsic catecholamines, thereby maintaining vascular tone.

Methods

We retrospectively evaluated patients with a diagnosis of septic shock who received intravenous methylene blue for recalcitrant hypotension. With a mean arterial pressure less than 65 mm Hg, hypotension secondary to distributive shock was resistant to vasopressors in the patients studied. Methylene blue was started at a bolus of 2 mg/kg IV bolus followed by 2 mg/kg/hr for 4 hours followed by 1 mg/kg/hr for 4 hours followed by 0.5 mg/kg/hr for 4 hours and then stopped. Statistical analysis was performed using the paired, two sample t-test, or Wilcoxon signed rank test where appropriate.

Results

We included eleven patients with a mean admitting APACHE II (“Acute Physiology And Chronic Health Evaluation II”) score of 24.8. Following one hour of methylene blue administration, there was a statistically significant increase in mean arterial pressures from 73.2 (± 11.9) mmHg to 84.5 (± 20.5) mmHg. (P<0.05)

Conclusion

Methylene blue may be useful as an adjunct therapy in patients with septic shock, resistant to standard treatment including vasopressors and adequate fluid resuscitation. The next frontier for methylene blue research should be in the intensive care unit. Larger patient samples, as well as controls, will be needed to further validate efficacy and determine if a prognostic benefit exists.
The Impact of Music on Nociceptive Processing

Jasmine Y. Gale, BS, BA1,2; Peter R. Chai, MD, MMS1,3; Guruprasad D. Jambaulikar, MBBS, MPH1; S. Wade Taylor PhD3,4; Rob R. Edwards PhD5; Edward W. Boyer, MD, PhD1,3; Kristin L. Schreiber MD, PhD5
Department of Emergency Medicine, Brigham and Women’s Hospital, Harvard Medical School1; Tufts University School of Medicine2; The Fenway Institute3; Boston University4; Department of Anesthesiology, Perioperative and Pain Medicine, Brigham and Women’s Hospital, Harvard Medical School5

BACKGROUND

• Music has been shown to decrease pain and modulate affect.
• The precise mechanism in which music tempers pain is unknown, but modulation of temporal summation of pain, anxiety, depression, and catastrophizing may underlie the analgesic efficacy of music.

AIMS

1. To assess the impact of a novel, music algorithm on nociceptive processing
2. To examine the impact of the music intervention on anxiety and situational catastrophizing

METHODS

• Observational study of healthy volunteers over the age of 18 without a self-reported history of chronic opioid use or neuropathy (n=60)
• Brief “bedside” Quantitative Sensory Testing (QST), comparing pain sensitivity in absence and presence of smartphone-based music intervention, including:
  1. Pressure pain threshold and tolerance performed on dorsal forearm (FA) and over trapezius bilaterally
  2. Repeated pinprick train (x10, 1 Hz) over dorsum of index and middle fingers bilaterally; Pain ratings (0-10) of 1st, 5th, and 10th stimuli, and 15 seconds after train (Painful After-Sensations (PAS)); Temporal Summation of Pain (TSP) calculated as difference between 10th and 1st pain rating
  3. Conditioned pain modulation (CPM): pressure pain threshold measured over non-dominant trapezius in absence and presence of dominant hand in an ice water bath while receiving pain pressure threshold testing on the contralateral trapezius
  • Psychosocial assessment (depression, anxiety, catastrophizing, somatization), using PROMIS short forms, Pain Catastrophizing Scale, and Brief Symptom Inventory-somatization subscale

RESULTS

• An increase in pressure pain thresholds in both the FA (p=0.007) and trapezius (p=0.002) was observed with music.
• A decrease in both amount of pinprick pain (p=0.001) and TSP (p=0.006) was observed with music.
• A decrease in CPM (p=0.001) was observed with music.
• No change was observed with anxiety or situational catastrophizing.
• The degree of pain reduction (pressure pain tolerance) with music was negatively correlated to catastrophizing and depression scores.

CONCLUSIONS

• Subjects exhibited a higher pain tolerance and reported lower pain scores during the music intervention, as compared to baseline, suggesting music may reduce pain sensitivity.
• Subjects reported less temporal summation of pain, suggesting the music may reduce facilitation of pain.
• CPM was significantly decreased, although this may be due to an overall decrease in pressure pain.
• There were no significant changes in anxiety or situational catastrophizing in response to music.
• Those with higher baseline catastrophizing and depression scores did not have as great an analgesic impact of music.
• Music may modulate nociception and is a potential adjunct in treating pain.

Change in Pressure Pain (Figure 1)

Figure 1: There was a significant increase in pressure pain thresholds in both the FA and the trapezius with music, as well as an increase in trapezius pressure pain tolerance with music (p=0.007, 0.002 Wilcoxon signed rank test). No significant change was seen with FA pressure pain tolerance. These measures were variable between individuals, but highly correlated within subject.

Change in Temporal Summation of Pain (Figure 3)

Figure 3: There was also a significant decrease in temporal summation of pain (TSP) to repeated pinprick probe stimulus in the music condition compared to baseline (p=0.038, Related Samples Wilcoxon signed rank test).

Change in Pain Tolerance with Music

Figure 5: There was a significant decrease in Pressure Pain Tolerance, [pain in presence-pain in absence of cold pressor/pain in absence of cold pressor] * 100) whether measured as a pain threshold or pain tolerance in the music condition (p=0.001, Wilcoxon signed rank test). Positive values represent a greater modulation of pressure pain by the cold pressor pain on contralateral hand.

Figure 6: There was a significant negative correlation with baseline depression t scores and pressure pain tolerance (p=0.038, Spearman’s Rho test).
Cancer of the urinary bladder (BC) ranks second in mortality and morbidity among the genitourinary cancers causing over 16,000 deaths annually. It is the most expensive cancer to treat from diagnosis to death, due in part to its intrinsic molecular heterogeneity that makes prognosis difficult, requirement of invasive procedures such as cystoscopy, and a high incidence of recurrence. Recent efforts to improve the diagnosis for BC are rooted in identifying molecular signatures of various BC subtypes based on the Tumor Cell Genome Atlas (TCGA) database and then personalizing treatment protocols. Urothelial Bladder Cancer arises from the urothelium, a multi-layered transitional epithelium lining the bladder lumen. Fully differentiated superficial cells overlie intermediate cells which have limited proliferative potential, and finally a basal layer composed of cuboidal cells resting on a basement membrane completes the urothelium. Thus, morphologically, BC can be divided into two molecular subtypes referred to as luminal and basal differing clinical sensitivities to therapy. Further, these epithelial layers express distinct cytoskeleton and cancer stem cell markers. Although treatment based on molecular signatures has the potential to be effective, a verification of their expression in strictly compartmentalized epithelial subtypes is not presently available. Furthermore, tumor cells with inherent genomic instability are unlikely to maintain a predictable molecular signature. We hypothesized that although the fidelity of molecular subtypes in BC may be less than optimal, expression of at least some molecules in the signature panel are likely to characterize either the basal or luminal compartment. The future implications of this project are the formation of a bed-side toolkit that can be used following a bladder tissue biopsy to cost-effectively evaluate the subset of cancer and provide an efficient disease prognosis.

**BACKGROUND**

The spectrum of UBC at presentation includes non-muscle invasive and muscle invasive disease. We analyzed genomic expression profiles in five bladder cancer cell lines (RT-4, 5637, T24, HT-1376, and 253J) ranging from a grade I cancer cell line (RT-4) to a grade IV cancer cell line (253J). Based on existing literature, we assigned these bladder cancers to one of two candidate intrinsic molecular subtypes: luminal and basal. Luminal tumors are characterized by expression profiles similar to intermediate/superficial layers of the epithelium and basal tumors correlate to the basal layer of the urothelium. Importantly, basal bladder cancers are more aggressive and lead to shorter survival times.

Through the use of RT-PCR, we analyzed genomic expression levels of eleven biomarkers (luminal: UPK, GATA-3, RAB 25, E-Cadherin; basal: CK-6, p63, CD44S, CD44V, CyclinB1, EGFR, and CD49). Particularly, for a cost-effective analysis, the expression profiles of only two markers (GATA-3 and CD44S) could be used to identify the molecular subtype of the bladder cancer to determine a first-look prognosis at the bedside.

To further our understanding of the differences between basal and luminal subtypes of bladder cancer, we used MITT Cell Viability Assays to test the effects of Gemcitabine and Cisplatin on five bladder cancer cell lines. Existing literature states that though basal bladder cancers are more aggressive, they are also more sensitive to chemotherapy initially compared to luminal bladder cancers. We hypothesized that the RT-4 and 5637 cell lines would be more resistant to treatment compared to the more aggressive T24, HT-1376, and 253J cell lines.

**RESULTS**

**Expression of Luminal and Basal Biomarkers in Bladder Cancer Cell Lines**

**CONCLUSIONS**

- Less aggressive bladder cancer cell lines (RT-4, 5637) showed higher expression profiles for luminal biomarkers (UPK, GATA-3, RAB 25, E-Cadherin).
- The more invasive cell lines (T24, HT-1376, 253J) showed a slight upregulation of basal target genes (CK-6, p63, CD44S, CD44V, CyclinB1, EGFR), but these markers were also modestly present in the less invasive cancer cell lines. For a cost-effective analysis, the expression profiles of only two markers: GATA-3 and CD44S could be used to identify the molecular subtype of bladder cancer.
- MITT Assays showed the significant cytotoxic effect of Gemcitabine especially but also Cisplatin on all bladder cancer cell lines. However, contrary to our hypothesis, basal cancers (253J, HT-1376, and T24) were not significantly more susceptible than luminal cancers when exposed to the chemotherapeutic agents.

**FUTURE EXPERIMENTS**

- Protein expression levels of the eleven biomarkers in the same five cell lines should be analyzed through the use of Western Blot Analysis to better validate the presence of luminal and basal biomarkers.
- The future implications of this project are the creation of a bedside tool-kit that can be used following a bladder tissue biopsy to cost-effectively evaluate the grade of the cancer to provide an efficient disease prognosis.

**FUNDING**

This work is supported from the Medical Scholar Program – MSP to Isha Dabke and a VA Merit Research Award (I01 BX003862-01A2) to Bal Lokeshwar.
OF ALL THE CAUSES OF BACK PAIN: ACUTE LYMPHOBlastic LEUKEMIA

PRESENTING WITH SPINAL CORD COMPRESSION

Kallie Kram BS1, Albert Jang BS1, Scott Berger BA1, Mahmoud Gaballa MD2, Marc Robinson MD3
1School of Medicine, Baylor College of Medicine, Houston, TX. 2Department of Hematology & Oncology, Baylor College of Medicine, Houston, TX. 3Department of Internal Medicine, Baylor College of Medicine, Houston, TX.

LEARNING OBJECTIVES
• Recognize acute lymphoblastic leukemia (ALL) as a rare cause of spinal cord compression (SCC)
• Understand that the appropriate initial treatment for SCC in ALL may be systemic chemotherapy alone rather than surgical decompression and/or radiation in SCC

History of Present Illness
38-year-old previously healthy man presented with acute on chronic midback pain
Began seven months prior, acutely worse over past four days
Associated with gait instability, saddle anesthesia, lower extremity weakness, and urinary retention

Medical and Social History
• No IV drug use, malignancy, or recent trauma

Review of Systems
• ++ gum bleeding, - fevers, chills, weight change

PHYSICAL EXAMINATION AND LABS
Vitals: Temp 98.4°F, BP 131/79, HR 68, RR 18, SpO2 99%
General: Alert, oriented male in no acute distress
HEENT: Petechiae on hard palate, mucosa moist
Neck: Left supravcavicular and cervical lymphadenopathy, nontender
CV: Regular rhythm, no murmurs, rubs or gallops
Respiratory: Lungs clear to auscultation bilaterally
Abdominal: Soft nontender, nondistended
Back: Point tenderness to T9-T12 vertebrae with associated parasternal tenderness
Extremities: 2+ pulses bilaterally with no edema
Skin: No rashes or wounds
Neurologic: CN II-XII grossly intact, 5/5 strength of hip flexion bilaterally, 4/5 in all other muscle groups, 3+ patellar reflexes bilaterally, 2+ biceps and triceps reflexes bilaterally, negative Babinski, fine touch sensation intact, normal rectal tone
CBC: Hemoglobin: 10.7 g/dL, Platelet Count: 13x10^9/µL

IMAGING AND PATHOLOGY

Chromosomal Testing
Philadelphia Chromosome Negative (Ph-)

FINAL DIAGNOSIS, MANAGEMENT, AND OUTCOME
Final Diagnosis: Philadelphia Negative B Cell Acute Lymphoblastic Leukemia

Treatment
• High dose dexamethasone was started to relieve the cord compression
• Started on high dose methotrexate, cytarabine, rituximab, and administered intrathecal methotrexate

Response
• At cycle 1 day 5, neurological symptoms further improved, with resolution of saddle anesthesia, normal reflexes, and normal urination
• After HyperCVAD cycle #1 parts A and B, bone marrow biopsy showed complete remission with no evidence of lymphoblastic leukemia

DISCUSSION
ALL is a malignant transformation and proliferation of lymphoid progenitor cells
• Bone marrow, blood, and extramedullary sites
• Nonspecific presentation: B symptoms, infection, easy bruising, fatigue
• Rapid initiation of systemic treatment warranted.
• Complete remission rate for adult ALL protocols is >90%, supporting that ALL is very chemosensitive in the first line setting.

Rarely presents spinal cord compression (SCC)
• More commonly due to solid tumors: breast, prostate, lung cancer
• Treat solid tumors with steroids followed by surgery and/or radiation

Key Points of This Case
• Unusual presentation with compression fractures and SCC without classic ALL symptoms
• Severe thrombocytopenia led to workup and early detection of acute leukemia
• Rapid initiation of systemic therapy adequately treated systemic and local disease
• Local treatment of a solid tumor with biopsy, surgery, or radiation would have delayed systemic treatment

CONCLUSION
• In patients presenting with spinal cord compression, ALL is rarely at the top of a differential diagnosis. However, suspicion should be raised in a patient with thrombocytopenia.
• Early diagnosis of ALL is key because standard initial management of SCC with local treatment modalities, such as radiation and neurosurgery, will delay systemic therapy with induction chemotherapy, which is optimal treatment for ALL.

REFERENCES
Requiring Discharge Summaries at Discharge to Improve Transitions to Rehabilitation Facilities

Bonnie Stedge MS3, Kristen Flint MS4, Natalie Giles MD, Julie Hollberg MD, Traci Leong PhD, Christopher O’Donnell MD, FHM
Emory University School of Medicine, Atlanta, GA 30326

Introduction

Readmission rates are an important quality metric for hospitals and marker of patient outcomes. In October 2017, subacute rehab centers (SARs) had to start reporting readmission rates and face potential penalties from Medicare based on those rates starting in October 2018. Thus, SARs will be closely tracking their readmission rates and now have vested financial interest to prevent readmissions. Decreasing readmission rates is a universal goal but can be a difficult issue to tackle. One of the many factors thought to influence readmission rates is communication during transitions of care. Discharge packets are an important component of this communication and include discharge summaries, medication lists, PCP information, and other components. Currently, Georgia medical guidelines do not necessitate a discharge summary transfer with the patient. There is conflicting and insufficient data on whether the presence of discharge summaries decreases readmission rates, especially related to readmissions from subacute rehab centers. Other important components of the discharge packet are often absent even if a discharge summary is present. Some of our previous work to assess SAR facilities’ needs has identified the frequent absence of follow-up appointments with subspecialists.

We aim to improve 30-day readmission rates of subacute rehab centers by 50% by mandating that discharge summaries accompany patients on arrival to subacute rehab by the end of 2018 for patients discharged from the hospital medicine service at Emory University Midtown.

Methods

1) Establish relationships with SAR facilities and develop auditing tool for data collection
2) Collect baseline readmission rates and conduct baseline discharge packet audits
3) Intervention: mandate that discharge summaries accompany patients upon discharge
4) Collect post-intervention readmission rates and conduct post-intervention discharge packet audits
5) Analyze pre- and post-intervention 30-day readmission rates

Results

39% relative risk reduction in 30-day readmission rates (p=0.0399, n=739)

Figure 2. Thirty-day readmission rates decreased after mandating that discharge summaries be sent with discharged patients. The total 30-day readmission rates from the three post-intervention months was 11.59%, the total 30-day readmission rate from the three post-intervention months was 7.02% with an absolute readmission risk reduction of 4.57% (95% CI 0.19-8.66%). The relative risk reduction was 39% (p<0.04, n=739).

Conclusions and Future Directions

Discharge summaries are critical modes of communication at hospital discharge. Following our mandate that the discharge summary accompany the patient at discharge, 30-day readmission rates for the hospital medicine service decreased from 11.6% to 7.0%. The relative risk reduction was 39% (p=0.04, n=739). To ensure that our mandate was successful in increasing the number of discharge summaries present upon discharge to SARs, we also measured the percentage of patients with discharge summaries. The percentage increased from 38% pre-mandate to 91% post-mandate (p<0.05). We also saw statistically significant increases in the number of discharge packets that included follow-up appointments and PCP information post-intervention.

Our study did have some limitations. We were limited to one hospital in metro Atlanta. Readmission rates from SARs were calculated using a data pull of ¼ of the total patient data from that timeframe (n=739). We did not control for temporal variation in readmission rates or other confounders; it is possible that readmission rates are at baseline higher in the winter months than the summer months.

Future directions include analyzing the other three quarters of data from the study time period to confirm that the trend of the results holds for a larger population. We will also continue to identify what information SARs want in discharge packets and what components of the discharge summary are most helpful.

Our end goal is to standardize the SAR discharge packet to improve patient care and decrease readmissions, possibly expanding this model to other institutions.

Overall, our mandate improved the quality and timeliness of discharge documentation for patients discharged to SARs. We saw a substantial decrease in 30-day readmission rates following our intervention. Good discharge documentation is a low cost and low effort intervention to prevent unnecessary readmission penalties.

References


The authors would like to acknowledge Jen Schuck and Mackenzie Moore for their help in connecting with SAR facilities.
A Rare Case of Pituitary Adenoma: The mysterious Crooke’s Cell Adenoma

Dina Jaber MS3, Jessica Padniewski MS4, Russell Pluhm MS4, Dr. Kartoumah MD, and Dr. Horani MD.
St. George’s University School of Medicine, A.T. Still University Kirksville College of Osteopathic Medicine, Mercy Gilbert Medical Center

Introduction: Cushing’s disease is defined by excess cortisol and can be caused by endocrinologically functional ACTH-producing pituitary adenomas [1]. Excess cortisol leads to a cytoplasmic accumulation of cytokeratin filaments and hyalinization which characterizes Crooke cell tumors, a rare (largest case series of 36) reported by George et al., variant of corticotroph adenoma[2].

Case: A 42-year-old Caucasian male, with a history of obesity, HTN, DM2, and refractory hypokalemia presented to the hospital as a direct admit with symptoms of headache, eye pressure, pedal edema, and weight gain for 4 weeks. Despite interventions, his symptoms didn’t improve and suspicion for Cushing’s syndrome increased. The patient had a cushingoid appearance with positive parinaud sign (limited upward eye deviation) both suggestive of pituitary mass. CT head revealed a 1.2cm right pituitary macroadenoma with remodeling of sella turcica. Random cortisol 39.6 and ACTH 140. Low and high dose dexamethasone suppression test failed to suppress cortisol levels which suggested ectopic hypercortisolism. The patient underwent endoscopic endonasal transsphenoidal (EET) resection of pituitary macroadenoma with limited visualization secondary to cavernous sinus bleeding. Initially postoperative cortisol levels decreased but then started to rise increasing suspicion for residual sellar tumor. The patient underwent repeat EET resection after which cortisol levels dropped and reached a nadir of 3.0 with ACTH 13. Pathology results revealed Crooke cell variant adenoma.

Discussion: Crooke’s cell tumors are a rare variant of pituitary adenoma and are important to distinguish from other adenomas due to their unique cellular behavior. The cause of hyalinization within these tumors is unknown. It is also unclear as to why this hylanization results in increased ACTH production as hyalinization typically results in loss of function[3]. These tumors are also very aggressive and prone to recurrence even after resection. Some patients with cushing’s syndrome and macroadenoma may fail high dose dexamethasone suppression testing and behave as ectopic cushing’s syndrome[4]. If patients with evidence of pituitary tumor fail dexamethasone suppression test it is important for clinicians to maintain suspicion of aggressive tumor variants such as Crooke’s tumor which require close follow up and management including possible radiation therapy[5]. In conclusion, this case highlights a cushing’s syndrome caused by a rare variant of pituitary adenoma, a Crooke’s variant tumor.
Case
A 36-year-old man with human immunodeficiency virus (HIV) on antiretroviral therapy presented with an anterior mediastinal mass on computed tomography (CT). A biopsy was not concerning for a malignant or infectious process, and the mass was monitored with routine imaging. Two years later, he presented with neck weakness, diplopia, and dysphagia concerning for myasthenia gravis. Medical workup found a positive anti-acetylcholine receptor antibody, and a biopsy of his mediastinal mass was consistent with thymoma. CT imaging demonstrated implants along the right hemi-diaphragm consistent with pleural metastases. He received a partial course of radiation or chemotherapy.

Four years after initial presentation, he returned with chronic diarrhea and failure to thrive. His stool was not concerning for a malignant or infectious process, and the mass was monitored with routine imaging. Two years later, he presented with neck weakness, diplopia, and dysphagia concerning for myasthenia gravis. Medical workup found a positive anti-acetylcholine receptor antibody, and a biopsy of his mediastinal mass was consistent with thymoma. CT imaging demonstrated implants along the right hemi-diaphragm consistent with pleural metastases. He received a partial course of radiation or chemotherapy.

Physical Exam
Pulse: 93 (05/25/18 0709) BP: 105/69 Temp: 36.6 °C (97.9 °F) Temp src: Oral Respiratory Rate: 16 Height: 6' Weight: 56.7 kg (125 lb 1.6 oz) SpO2: 92 % O2 Device: None (Room air) BMI (Calculated): 17

General: chronically ill-appearing, cachectic, in discomfort but not distressed, weak cough
HEENT: oral mucosa dry with leukoplakia, conjunctivae normal, non-icteric sclera, supple neck
Cardiovascular: regular rate and rhythm, 2/6 systolic ejection murmur, no rales or gurgles
Pulmonary: decreased breath sounds in bases, scant rales
Abdomen: soft, thin, non-tender, non-distended, no guarding or rebound tenderness
Musculoskeletal: diffuse decrease in muscle bulk
Neuro: alert and oriented x3, cranial nerves grossly intact
Skin: warmpink plaques with serpiginous borders and scaly, violaceous edges scattered on the face, arms, and legs and confluent on the chest, abdomen, and back
Lymph nodes: no lymphadenopathy

Labs and Biopsies

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>138 g/dL</td>
</tr>
<tr>
<td>Platelets</td>
<td>95 ×10^9/L</td>
</tr>
<tr>
<td>Neutrophils</td>
<td>4.1 ×10^9/L</td>
</tr>
<tr>
<td>Lymphocytes</td>
<td>33 ×10^9/L</td>
</tr>
<tr>
<td>Monocytes</td>
<td>0.69 ×10^9/L</td>
</tr>
<tr>
<td>WBC</td>
<td>166 ×10^9/L</td>
</tr>
<tr>
<td>Red Blood Cells</td>
<td>11.99 ×10^12/L</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>41.5 %</td>
</tr>
<tr>
<td>Mean Corpuscular Volume</td>
<td>362 fL</td>
</tr>
<tr>
<td>AST</td>
<td>22 U/L</td>
</tr>
<tr>
<td>ALT</td>
<td>14 U/L</td>
</tr>
<tr>
<td>ALP</td>
<td>166 U/L</td>
</tr>
<tr>
<td>T. bili</td>
<td>0.5 mg/dL</td>
</tr>
<tr>
<td>Alb</td>
<td>2.6 g/dL</td>
</tr>
<tr>
<td>INR</td>
<td>1.2</td>
</tr>
<tr>
<td>PT</td>
<td>14.1 s</td>
</tr>
<tr>
<td>CD4</td>
<td>605 cells/mm³</td>
</tr>
<tr>
<td>Viral load</td>
<td>undetectable</td>
</tr>
<tr>
<td>ANA titer</td>
<td>1:2560</td>
</tr>
<tr>
<td>Anti-SSA</td>
<td>negative</td>
</tr>
<tr>
<td>Anti-SSB</td>
<td>negative</td>
</tr>
<tr>
<td>Anti-Smith</td>
<td>negative</td>
</tr>
<tr>
<td>Anti-SCL-70</td>
<td>negative</td>
</tr>
<tr>
<td>Anti-jo-1</td>
<td>negative</td>
</tr>
<tr>
<td>Anti-RNP</td>
<td>negative</td>
</tr>
<tr>
<td>Calcium channel binding antibody</td>
<td>negative</td>
</tr>
</tbody>
</table>

Skin biopsy: vacuolar interface dermatitis with acanthosis and numerous epidermal necrotic keratinocytes. Direct immunofluorescence to evaluate for paraneoplastic pemphigus was negative for IgG, IgA, IgM, and C3 deposits.

Esophagogastroduodenoscopy: gastric oxyntic and antral mucosa with chronic active gastritis

Colonoscopy: active colitis with prominent apoptosis, scattered neutrophilic cryptitis, and eosinophilia

Discussion
Paraneoplastic syndromes result from tumor secretion of peptides or hormones or immune cross-reactivity. They most commonly affect the endocrine, neurologic, dermatologic, and rheumatologic systems and may manifest prior to diagnosis of malignancy or with an established malignancy. An estimated 8% of individuals with cancer have a paraneoplastic syndrome. This patient presented with a diffuse rash and chronic diarrhea in the setting of HIV and thymoma. Biopsies demonstrated dermatitis, gastritis, and colitis, consistent with multi-organ autoimmune syndrome.

Thymoma-associated multi-organ autoimmunity (TAMA) is a rare paraneoplastic syndrome that mimics graft-versus-host disease (GVHD). It can affect the liver, skin, and gastrointestinal tract and histologically resembles GVHD. Physicians should maintain a high index of suspicion for TAMA in patients with cutaneous eruptions, chronic diarrhea, and/or abnormal liver enzymes in the setting of thymoma. More broadly, physicians should consider paraneoplastic syndromes to explain a constellation of seemingly disjointed symptoms in the setting of malignancy or prior to a diagnosis.

References
Hemoperitoneum is a condition in which blood accumulates in the peritoneal cavity. Clinical manifestations vary based on the degree of internal bleeding. In the unstable patient, prompt diagnosis and intervention are imperative for improving outcomes and mortality.

We present a case of hemoperitoneum in an unstable patient diagnosed efficiently by bedside ultrasound (US). This diagnosis may have otherwise been delayed if pursued by other imaging modalities.

Introduction

Hemoperitoneum is a condition in which blood accumulates in the peritoneal cavity. Clinical manifestations vary based on the degree of internal bleeding. In the unstable patient, prompt diagnosis and intervention are imperative for improving outcomes and mortality.

We present a case of hemoperitoneum in an unstable patient diagnosed efficiently by bedside ultrasound (US). This diagnosis may have otherwise been delayed if pursued by other imaging modalities.

Bedside US findings

The fluid demonstrated the presence of “hematocrit” sign, which is a layering effect with progressively increasing echogenicity in the gravity dependent regions of the abdominal cavity. This is caused by the accumulation of echogenic particles such as coagulated blood and cellular debris in such areas. Furthermore, the presence of swirling echogenic particles within the fluid, known as “plankton” sign, and mobile fibrinous strands indicate the presence of ongoing coagulation further indicating the presence of intraperitoneal blood.

US Images

Case Presentation

A 77-year-old male presents to our institution for evaluation of encephalopathy for 2 days duration. In the emergency room the patient was lethargic and initial workup revealed acute liver failure. As part of ongoing workup, a core biopsy of the right hepatic lobe was performed by interventional radiology under US guidance.

One day post-procedure, the patient was noted to have sudden onset hypotension. Physical exam was notable for a tensely distended and tender abdomen. Blood pressure showed a precipitous decline from 94/66 to 68/48 within 15 minutes. These findings were notable to be in association with a 3.7 g/dl decrease in hemoglobin. Patient was subsequently transfused several units of blood.

At this time, a bedside US was performed to guide resuscitative efforts and revealed a large amount of complex intraperitoneal fluid consistent with hemoperitoneum.

Case cont’d

Despite interventions, the patient suffered cardiac arrest. Based on the above US findings, massive transfusion protocol was initiated and emergent decompressive paracentesis was performed with 1L of frank blood drained. Return of spontaneous circulation was obtained within 10 minutes.

Once stabilized the patient was sent for angiography and embolization of the right hepatic artery.

Discussion

Bedside US allowed physicians to make a rapid diagnosis of hemoperitoneum and abdominal compartment syndrome, allowing swift and goal directed intervention to stabilize the patient. In prior studies, US has demonstrated sensitivity of 81.8% and 93.9% in the identification of hemoperitoneum.

Although CT scan is often the common imaging modality for the diagnosis of hemoperitoneum, US offers real-time and portable imaging to adjunct the clinical exam in detection of hemoperitoneum in unstable patients without significant delay or transport.

REFERENCES

Hashimoto Encephalopathy mimicking Bells palsy as an initial presentation
Ricci Kalayanamitra BS, Cheren Elangovan MD, Justin Lowe PA-C, Rohit Jain MD, Raymond Reichwein MD
Penn State Milton S. Hershey Medical Center

Introduction
Hashimoto encephalopathy (HE) is a rare syndrome associated with Hashimoto thyroiditis. HE mostly affects middle-aged women and often appears to be due to autoimmune vasculitis or an immune complex deposition that disrupts the cerebral microvasculature. This immunologic phenomenon has been observed on brain biopsy with a lymphocytic infiltration around small arterioles and venules. Common presentation of HE is diffuse hyperreflexia and other pyramidal tract signs (85%), followed by generalized tonic-clonic seizures (67%), myoclonus or tremor (38%), and status epilepticus (12%). The presence of antithyroid antibodies is essential for the diagnosis of HE.

Here we present a case report of Hashimoto encephalopathy presenting with acute focal neurologic symptoms mimicking TIA and stroke.

Case
A previously healthy 57-year-old female with past medical history significant only for borderline diabetes on metformin, presented to the emergency room after 3 days of right facial droop. The patient noticed the facial droop after accidentally bumping her head on a staircase 3 days prior. She denied any other symptoms.

On arrival her blood pressure was 240s/120s and blood glucose was 159, so she was given multiple doses of labetalol and metoprolol, which brought them down to 190s/100s and 118, respectively. She underwent MRI scan of the brain, which revealed restricted diffusion concerning for strokes in the pons and corona radiata. Microbleeds within the left thalamus and left pons, along with old infarcts of the right medulla and right corona radiata were also seen.

Serial MRI(brain) revealed scattered multiple acute progressive subcortical hyperintensities, not typical for demyelinating disease, such as multiple sclerosis. Intracranial MRA did not reveal evidence for infection. She was felt to have active CNS autoimmune disease, most likely related to significantly elevated anti-tPO antibodies. CSF showed elevated IgG of 1710mg/dL. She was discharged on methotrexate and pioglitazone.

An ultrasound of head and neck was done 2 months later and it was impressionistic for Hashimoto’s thyroiditis, evidenced by findings of diffusely enlarged thyroid gland at 7.5cm with heterogeneous parenchymal echogenicity. There are subcentimeter isoechoic to hyperechoic nodules within the isthmus.

Discussion
Hashimoto encephalopathy is a rare syndrome of Hashimoto thyroiditis that may mimic stroke. Diagnosis requires high index of suspicion. Findings on brain imaging are widely variable, including multiple ischemic infarcts, hemorrhagic lesions, non-specific white matter lesions, meningeal enhancement, and gadolinium enhanced T1 lesions among others. Elevated anti-thyroid peroxidase antibody or anti-thyroglobulin antibody, along with response to corticosteroids, are required for definitive diagnosis. Because of this response to immunosuppressive therapy, it is especially important to thoroughly exclude infection.

Conclusion
Hashimoto encephalopathy is a rare disorder which necessitates investigation for elevated anti-thyroid antibodies. The rapid progression and association with both ischemic and hemorrhagic events may be mistaken for CNS vasculitis. This case underscores the importance of checking for anti-thyroid antibodies in rapidly progressive CNS vascular conditions.

References:
Leukocytosis & elevated lactate – is it always sepsis?
Shadi Ahmadmehrabi, Juan Barbastefano MD
Cleveland Clinic Lerner College of Medicine

**Case Presentation**

- **ED**
- Lactate 3.5 mmol/L WBC 33.8k
- CT: 6.2 cm right upper lobe mass, increased in size from prior, no evidence of chest wall and bony invasion
- CT: negative for PE, did not show infiltrate or obstruction

**Imaging & Pathology**

- Fig 1. Contributing factors to elevated lactate and leukocytosis in the presented patient
- Fig 2. CT demonstrating apical mass with evidence of chest wall invasion
- Fig 3. Pathology specimen from technically difficult thoracotomy revealing spindle cell carcinoma with extensive areas of necrosis

**Discussion**

- Acutely, patient w/ high tumor burden was treated with empiric IV antibiotics for presumed sepsis
- Though pt met SIRS criteria (HR, RR, WBC), no source of infection was apparent on H&P; vitals and labs were stable throughout admission
- Similar presentation at prior admission, DC’d when infection was ruled out
- WBC>40k in non-hematologic cancer patients is associated w/ large tumor burden, higher DVT/PE risk, poor clinical outcomes; uncommonly due to infxn1
- Elevated lactic acid (≥ 1.4 mmol/L) also a negative prognostic factor in metastatic lung cancer8

**Key Points**

- Recognition of lab abnormalities associated with high tumor burden is important for institution of appropriate testing and management strategy
- Tumor progression, especially NSCLC, can present with leukocytosis >40k, which is uncommonly due to infection in non-hematological cancer patients
- Leukocytosis is a negative prognostic indicator
- Non-ischemic/hyperperfusion causes of elevated lactate include:
  - Alcoholism
  - - Liver dysfunction
  - - Thiamine deficiency
  - Malignancy
  - - Paraneoplastic
  - - - Tumor necrosis
  - Steroids

**References**

"You don’t have to be dying to do comfort measures": Patients’ and Physicians’ Perceptions of Inpatient Attire

Christy Lucas, BS¹ and Cheryl Dellasega, CRNP/MS, PhD, MFA²
¹Penn State College of Medicine, Hershey, PA; ²Penn State College of Medicine, Department of Humanities, Hershey, PA

Introduction

Patient gowns have become an unquestioned expectation of “what patients wear” in the hospital. Even with a shift toward patient-centered care and evidence-based medicine, changes to the gown have been neglected due to tradition and lack of research.

Design flaws have even become elements of humor in the medical field:
- “See-more Hiney,” the “inventor” of the patient gown
- UPMC’s two Super Bowl LIII ads depicting a construction worker and businessman in a patient gown moaning co-workers

In spite of cheeky humor, patient gowns have a profound psychosocial impact on patients as individuals. Upon admission to a hospital, a person partakes in the transformative medical ritual of gowning, becoming “the patient” with a one-size-fits-none gown that diminishes identity, individuality, and agency.

Patients are vulnerable to loss of dignity in hospitals, but what is less known is that wellbeing can be impacted by wearing the current gown. Providers can also react negatively to seeing patients in standard hospital attire.

Objective

The purpose of this study was to explore how patients and physicians perceive the function and impact of the patient gown.

Methods

Patients (5 women, 5 men) and physicians with an inpatient and outpatient practice (6 women, 4 men) at an academic medical center participated in standardized 1:1 interviews that explored the impact of gowning.

Perceptions about health status, meaning of the gown, reasons for its use, and barriers to change were asked. Interview transcripts were analyzed using an iterative process to identify themes within and between participants. Themes were grouped into separate categories for each group and consensus on gowning reached.

Participants sketched their “ideal” patient attire (Figure 1).

Results

Select quotations from patients and physicians can be viewed with the QR code shown below using the camera app of your mobile device.

Patient Themes

No choice but to wear a gown

Strategies for wearing the gown

Gowns are physically uncomfortable
- Restricting, not practical for toileting, “one-size-fits-none,” exposing

Gowns have a negative psychological impact
- Reinforce dependence and perpetuate a loss of identity, self-esteem, choice, humanity, and dignity
- Gowns show that the hospital doesn’t care

Physician Themes

Biases associated with patient attire
- Patients in gowns = “sicker,” resigned, and dependent
- Patients in clothing = “healthier,” motivated, and hopeful
  - Perhaps even a “frequent flier” or drug-seeking

Physician autonomy

Results Cont’d

Physicians’ distress at seeing patients in gowns they themselves viewed negatively

Medical myths perpetuate the “required” gown
- Auscultating under the gown on every patient
- MRSA/infectious control
- Wires for EKG
- Gowns are cleaner
- Gowns make the physical exam easier

Common Threads

Negative first impressions of the gown
- Visually unappealing
- Makes patients vulnerable

Purpose of the gown is for physician’s ease of access
- MRSA/infectious control
- Need for choice in patient attire (e.g., sizing, color)
- Wish to wear one’s own clothing while admitted to be more comfortable

Therapeutic role of the gown

Discussion and Future Directions

Studies emphasizing redesign efforts are vital to improving the patient gown.

Patients and physicians believe gowns need upgraded and that doing so could enhance wellbeing. Still, myths about gowns’ use persist, clouding the negative psychosocial impact and standing as a barrier to change in patient attire.

Participants highlighted the need for an evidence-based patient gown re-design and sketched proposed modifications. These are being translated into a colorful and comfortable patient-guided, provider-approved design.

References


UPMC Superbowl LIII TV Commercials (2019), "Are You Covered?" (below to view)
Specific Neuroanatomical Abnormalities Identified in Cyclic Vomiting Syndrome Based on Brain MRI Analysis

Jake Wilson1, Hugo Sandoval2, Marisol Ramirez2, Roshni Mandania1, Osagie Usen1, Jose Gavito-Higuera2, Carola Mullins2, Irene Sarosiek3, Tyler Davis4, Richard W. McCallum3

1Paul L. Foster School of Medicine, Texas Tech University Health Sciences Center El Paso, TX; 2Department of Radiology, Texas Tech University Health Sciences Center, El Paso, TX; 3Department of Internal Medicine, Texas Tech University Health Sciences Center, El Paso, TX; 4Department of Psychological Sciences, Texas Tech University, Lubbock, TX

Introduction

Cyclic vomiting syndrome (CVS) in adults is a disorder characterized by recurrent abrupt bouts of nausea, vomiting, and abdominal pain separated by variable symptom-free periods that may last from days to months. Major etiological factors established by previous studies include migraine, stress, and marijuana usage. A dysregulation of central nervous system (CNS) pathways and neuroendocrine mediators may play a role in pathophysiology. Despite the potential role for CNS contributions to CVS, literature regarding CNS neuroanatomical differences that occur with CVS is limited. The current study investigated whether there are CNS neuroanatomical differences in acutely symptomatic CVS patients during relapse compared to controls using brain magnetic resonance imaging (MRI).

Methods

23 CVS patients were enrolled and 20 healthy controls were matched based on age and gender (Table 1). CVS patients were scanned during an acutely symptomatic episode. Risk factors identified in CVS patients included 14 with migraines, 14 with high levels of stress, and 6 with all 3 risk factors. High-resolution anatomical Magnetization-Prepared Rapid Gradient-Echo1 (MP-RAGE) images were obtained in each participant. MP-RAGE is a radiology sequence for structural brain imaging that captures high tissue contrast and provides high spatial resolution. Images were preprocessed using standard pipelines in FreeSurfer2 and tested for differences in cortical thickness and subcortical volume between CVS subjects and healthy controls. Surface-based results were thresholded and corrected for multiple comparisons using Monte Carlo simulation.

Table 1: Subject Demographics

<table>
<thead>
<tr>
<th>Group</th>
<th>N(%)</th>
<th>Mean Age (Range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CVS</td>
<td>23</td>
<td>32 (19-59)</td>
</tr>
<tr>
<td>Non-Marijuana Use CVS</td>
<td>13</td>
<td>35 (22-59)</td>
</tr>
<tr>
<td>Male</td>
<td>4 (31)</td>
<td>41 (23-59)</td>
</tr>
<tr>
<td>Marijuana Use CVS</td>
<td>10</td>
<td>30 (19-46)</td>
</tr>
<tr>
<td>Female</td>
<td>9 (69)</td>
<td>33 (22-46)</td>
</tr>
<tr>
<td>Male</td>
<td>5 (50)</td>
<td>28 (24-36)</td>
</tr>
<tr>
<td>Healthy Controls</td>
<td>20</td>
<td>32 (18-53)</td>
</tr>
<tr>
<td>Female</td>
<td>12 (60)</td>
<td>31 (18-44)</td>
</tr>
<tr>
<td>Male</td>
<td>8 (40)</td>
<td>34 (19-53)</td>
</tr>
</tbody>
</table>

Results

Surface-based cortical thickness analysis (Figure 1) revealed that CVS patients had significantly lower cortical thickness in bilateral precuneus (left: p=.001; right: p=.0002), left superior frontal gyrus (p=.008), and right precentral gyrus (p=.0084) than healthy controls. No cortical differences were found between marijuana use (MU) and non-marijuana use (non-MU) subgroups of CVS patients. Subcortical results (Figure 2) identified reduced cortical volumes in non-MU CVS patients in bilateral hippocampus (left: p=.002; right: p=.001) and amygdala (left: p=.023; right: p=.038). The MU subgroup had significantly greater cortical volumes in bilateral putamen (left: p=.05; right: p=.03) and pallidum (left: p=.004; right: p=.01) relative to non-MU patients.

Discussion

Based on brain MRI analysis, our major conclusions were that symptomatic CVS patients had cortical thinning relative to healthy controls in the precuneus and caudal regions of prefrontal/pretector cortex. Non-marijuana CVS subgroup had significantly lower subcortical volume measures in the amygdala and hippocampus compared to healthy controls. Marijuana subgroup were similar to controls and had greater basal ganglion cortical volumes consistent with increased dopaminergic stimulation. The precuneus is involved in visuo-spatial, vestibular processing and self-awareness3, which suggests a potential sensory-integration or vestibular basis for CVS. The amygdala and hippocampus are involved in processing pain, emotion, and stress which are relevant to CVS risk factors. Interestingly, we identified significant difference in the marijuana subgroup which may differentiate marijuana associated CVS from other etiologies. Our results provide possible novel brain MRI evidence for a CNS neuroanatomical basis of CVS with potential implications for targeting future therapy. Future studies to analyze CNS involvement in CVS patients should focus on identifying differences between neuroanatomy during an acutely symptomatic phase and periods of remission.

References


Acknowledgement

This study was funded in part by the President’s Collaborative Research Initiative Award from Texas Tech University Health Sciences Center El Paso.
Chlamydia pneumoniae is an obligate intracellular bacterium that causes respiratory infections in adults and children (1). There is evidence for an association between atypical bacterial pathogens (C. pneumoniae, M. pneumoniae) and asthma pathogenesis, as well as production of immunoglobulin (Ig) E responses in vitro (2,3). Previous studies in our laboratory demonstrated the presence of anti-C. pneumoniae IgE antibodies (Abs) by immunoblotting in children with culture confirmed C. pneumoniae infection (pneumonia and asthma) who were wheezing (2). We sought to determine whether past C. pneumoniae infections trigger production of C. pneumoniae-specific IgE Abs in adult subjects with and without asthma, who had positive C. pneumoniae-IgG titers.

Table 1. Detection of total serum IgE levels. Total serum IgE levels were similar in asthmatics compared with non-asthmatic subjects (186±159 vs. 170±142; P=0.720) (Figure 1).

Table 2. Detection of specific IgE C. pneumoniae antibody. However, C. pneumoniae IgE Ab levels were significantly higher in asthmatic patients compared with non-asthmatic subjects (1.015±0.305 vs. 0.39±0.340; P<0.001) (Figure 2). No significant association was found between total serum IgE levels and C. pneumoniae IgE Ab levels (R= -0.004, P=0.981).

Figure 1. Detection of total serum IgE levels (P=0.720). Asthma (N=22), non-asthma (N=22).

Figure 2. Detection of specific IgE C. pneumoniae antibody (P=0.001). Asthma (N=22), non-asthma (N=22).

References
Prosthetic Joint Infection: An Extremely Rare Complication of Intravesicular BCG Therapy.

Michael Storandt MSIIa, Avish Nagpal MD, MPHb

aUniversity of North Dakota School of Medicine & Health Sciences, Grand Forks ND; bDivision of Infectious Disease, Sanford Health, Fargo ND

Case Presentation

A 66-year-old male was seen in clinic due to concerns of tuberculosis of the right hip. He had a history of urothelial bladder carcinoma, which was treated via transurethral resection of the tumor two years prior to presentation. Following resection, he received intravesicular instillations of Mycobacterium bovis Bacillus Calmette-Guerin (BCG) for a total of 6 doses. He reported significant shaking and chills following each instillation, but the symptoms were self-limited and resolved within 24 hours on each occasion.

A few months later, he began to develop slowly worsening pain over his right hip, which had been replaced with an artificial prosthesis (Figure 1) 27 years prior due to secondary osteoarthritis resulting from pelvic fracture sustained in a motor-vehicle accident as a teenager. He was evaluated by an orthopedic surgeon and due to the concern of mechanical failure of the prosthesis, it was recommended he undergo a surgical revision of his prosthesis. At the time of surgery, joint effusion was noted and synovial fluid was sent for bacterial and mycobacterial culture.

Immediately postop, the patient developed shaking and chills similar to those experienced during BCG treatment. He also developed erythema around the site of the incision. For suspicion of Lyme disease due to his residence in an endemic area. After 3 weeks of incubation, the mycobacterial cultures of synovial fluid obtained at the time of surgery grew an acid-fast bacillus. A nucleic acid probe identified the isolate as Mycobacterium bovis.1

Diagnosis

Based on this clinical and microbiological data, the patient was diagnosed with a prosthetic joint infection secondary to Mycobacterium bovis Bacillus Calmette-Guerin. He was started on treatment with isoniazid, rifampin and ethambutol which he continues to tolerate well.

Discussion

BCG was first developed in the early 1900’s as a vaccination for tuberculosis, but it was soon discovered that it exhibited anti-tumor properties. The most successful and lasting of these therapies is utilization of BCG for superficial bladder carcinomas.2 While its exact mechanism has not been determined, BCG is known to elicit a cell-mediated immune response consisting of T cells and activated macrophages, which in turn, is thought to target cancer cells within the bladder.2 BCG instillation can cause local complications, the most common being cystitis, followed by fever and granulomatous prostatitis, but systemic complications such as sepsis, pneumonitis, or granulomatous hepatitis are rare, occurring in less than 1% of cases.3

BCG prosthetic joint infection is even more rare, with only a few cases being previously reported. These cases are difficult to detect as symptoms are mild and subacute, and in some cases, they may not manifest for years following BCG treatment.4 Treatment of BCG prosthetic joint infection has not been well established and is primarily based on previous case reports. M. bovis is largely resistant to pyrazinamide, and therefore, treatment regimens do not include this drug.2 Based on previous case reports, it seems that treatment should consist of prosthetics replacement in addition to some combination of first-line anti-mycobacterial agents including isoniazid, rifampin, and ethambutol.5-7

Table 1: Past cases of prosthetic joint infection secondary to BCG therapy

<table>
<thead>
<tr>
<th>Case</th>
<th>Age,Sex</th>
<th>Orthopedic Procedure</th>
<th>Age of Arthroplasty</th>
<th>Presentation of Symptoms Following BCG (Time)</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Our Patient</td>
<td>66, M</td>
<td>Right, THR</td>
<td>27 years</td>
<td>&lt;1 Year</td>
<td>THR, INH/RIF/ETM (currently undergoing treatment)</td>
<td>Currently undergoing treatment, well-tolerated</td>
</tr>
<tr>
<td>Gomez et al.</td>
<td>82, M</td>
<td>Right, THR</td>
<td>10 years</td>
<td>2 Years</td>
<td>THR, INH/RIF for 1 year</td>
<td>No symptoms after year of treatment</td>
</tr>
<tr>
<td>Reigstad and Siewers5</td>
<td>86, M</td>
<td>Left, THR</td>
<td>10 years</td>
<td>During Treatment</td>
<td>THR, INH/RIF/ can be for 6 months, INH for 1 year</td>
<td>No symptoms 30 months post-operation</td>
</tr>
<tr>
<td>Segal and Krauss</td>
<td>76, M</td>
<td>Left, THR</td>
<td>18 years (revision 12 years prior)</td>
<td>4 Years</td>
<td>THR, INH/RIF/ETM for 1 year</td>
<td>No infection 30 months post-operation</td>
</tr>
<tr>
<td>Guerra et al.7</td>
<td>66, M</td>
<td>Right, THR</td>
<td>6 years</td>
<td>20 Months</td>
<td>THR, INH/RIF for 6 months</td>
<td>3 months post-treatment: AFB culture negative, bone biopsy revealed AFB</td>
</tr>
</tbody>
</table>

Abbreviations: total hip replacement (THR), acid fast bacilli (AFB), isoniazid (INH), rifampin (RIF), ethambutol (ETM), and pyrazinamide (PZA)

Conclusion

Intravesicular instillation of Mycobacterium bovis Bacillus Calmette-Guerin, is a primary adjunctive treatment for bladder carcinomas, but can, in rare cases, result in systemic complications. Prosthetic joint infection has been infrequently reported as one such complication (Table 1). As there have been very few reported cases, a treatment protocol for this condition has not been established, although it seems intuitive to replace the prosthesis and treat with isoniazid, rifampin, and ethambutol for at least six to twelve months.

Our case highlights the challenges associated with diagnosis and management of this rare complication of a commonly used therapy.

References

Implementation of a coordinated plan for chronic opioid therapy in the primary care setting

Jennifer Woodard, MSIII1; Leigh Cervino, P32; Stephanie C. Blease3; Christopher D. Kearney, MD3; Kathryn A. Walker, PharmD, BCPS, CPE2,3
1. University of Maryland School of Medicine; 2. University of Maryland School of Pharmacy; 3. MedStar Health

Abstract

Background: Deaths from opioid overdose have increased dramatically in the past decade. Half of all dispensed opioid prescriptions are written by primary care clinicians. To improve management of non-cancer chronic pain in adults in the primary care setting, the Centers for Disease Control (CDC) developed a clinical practice guideline in 2016. The aim of this study was to assess documentation practices of primary care clinicians (PCPs) before and after the implementation of a coordinated plan for chronic opioid therapy (COT) at 18 outpatient clinics.

Methods: This retrospective pre-post study included 18 primary care sites (9 control, 9 intervention) before and after the intervention. Twenty COT patients from each site (defined as receiving opioids ≥ 3 of 6 months) were randomly selected for inclusion. One visit for each patient was selected for review during the study timeframes for baseline (5/16-10/16) and post-implementation (11/16-5/17) periods. The data coded from the chart included 28 criteria based on CDC guidelines and included calculating morphine milligram equivalents (MME).

Results: A total of 599 patient charts were reviewed (baseline: 172 control, 166 intervention; post-intervention: 140 control, 121 intervention). There was no significant difference in average MME at baseline or post-intervention. Four encounters included a physician-documented MME. There were increases in the intervention clinics in the frequency of documentation of pain scores and functional assessments.

Conclusions: The implementation of CDC standards in the intervention clinics did not show a significant impact on physician documentation practices in the first 6 month phase. Given the small sample sizes, only large effects would have been detectable; smaller effects may have occurred undetected. Clinician education and improved integration of clinical decision support tools within the electronic medical record is needed to improve adherence to the new CDC guidelines for COT.

Objective

- Assess documentation of PCPs before and after implementing a coordinated plan for chronic opioid therapy in outpatient clinics.

Methods

- Retrospective chart review of one randomly selected patient encounter with PCP during time frame.
- 29 elements in chart review (MME calculation, PDMP check, UDT order, treatment agreement, etc.)

Results

Demographics (n=608)

<table>
<thead>
<tr>
<th></th>
<th>Control</th>
<th>Demonstration</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>173</td>
<td>166</td>
</tr>
<tr>
<td>Age (avg yrs)</td>
<td>63</td>
<td>64</td>
</tr>
<tr>
<td>Female</td>
<td>57%</td>
<td>65%</td>
</tr>
<tr>
<td>Race</td>
<td>AA: 44%</td>
<td>AA: 34%</td>
</tr>
<tr>
<td></td>
<td>W: 45%</td>
<td>W: 57%</td>
</tr>
<tr>
<td></td>
<td>Other: 12%</td>
<td>Other: 9%</td>
</tr>
</tbody>
</table>

Baseline Follow-up

<table>
<thead>
<tr>
<th></th>
<th>Control</th>
<th>Demo</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>140</td>
<td>129</td>
</tr>
<tr>
<td>Age (avg yrs)</td>
<td>61</td>
<td>65</td>
</tr>
<tr>
<td>Female</td>
<td>77%</td>
<td>62%</td>
</tr>
<tr>
<td>Race</td>
<td>AA: 61%</td>
<td>AA: 48%</td>
</tr>
<tr>
<td></td>
<td>W: 38%</td>
<td>W: 47%</td>
</tr>
<tr>
<td></td>
<td>Other: 1%</td>
<td>Other: 5%</td>
</tr>
</tbody>
</table>

Follow-up*: One of the demonstration site clinics closed during follow-up.

Signed Treatment Agreements

<table>
<thead>
<tr>
<th></th>
<th>Baseline</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control sites</td>
<td>17 (10%)</td>
<td>17 (12%)</td>
</tr>
<tr>
<td>Demo sites</td>
<td>21 (13%)</td>
<td>27 (21%)</td>
</tr>
</tbody>
</table>

Average MME Prescribed

<table>
<thead>
<tr>
<th></th>
<th>Baseline</th>
<th>Follow-up</th>
<th>Change</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control sites</td>
<td>66</td>
<td>71</td>
<td>+5 (7.6%)</td>
</tr>
<tr>
<td>Demo sites</td>
<td>68</td>
<td>70</td>
<td>+2 (2.9%)</td>
</tr>
</tbody>
</table>

Discussion

- Provider documentation for COT needs to improve significantly to comply with current CDC guidelines.
- Providers using the EMR directed decision support tool were more likely to meet standards.
- Limited use of support tool indicates need for better provider education and implementation into current EMR workflow.

Disclosures: All authors of this presentation report nothing to disclose concerning possible financial or personal relationships with commercial entities that may have a direct or indirect interest in the subject matter of this presentation.

Contact information:
Jennifer Woodard, University of Maryland School of Medicine
Jennifer.Woodard@som.umaryland.edu
655 W Baltimore St
Baltimore, MD USA 21201
Background

- Cardiovascular events are the commonest cause of morbidity and mortality in kidney transplant patients (KTxPs).
- Endothelial dysfunction and consequent atherosclerosis may play a role in this.
- There is a poor understanding of how endothelial function changes with time in KTxPs and effect of novel biomarkers are unknown.
- This pilot study investigated changes in endothelial function over time, in stable kidney transplant patients (KTxPs) compared to controls.

Methods

- Brachial artery flow-mediated dilation (FMD) and nitroglycerin-mediated dilation (NMD) were assessed in 18 KTxPs and 17 controls at baseline and 3-6 months after.
- Blood biomarkers were assessed using Luminex technology multiplex assay.
- All subjects were recruited after written consent and all measurements were done in our vascular laboratory under standard conditions.

Results

- The median time since transplantation was 86 months (interquartile range 123 months).
- Comparing KTxPs to controls:
  - There were more dyslipidemics (10 vs. 3; \( P=0.02 \)).
  - eGFR was lower (67.61 ± 20.25 vs. 97.59 ± 15.59; \( P<0.01 \)).
  - There was no difference in age (51.28 ± 13.29 vs. 45.82 ± 10.85; \( P=0.19 \)), body mass index (25.56 ± 5.18 vs. 24.59 ± 2.59; \( P=0.49 \)), diabetes status (3 vs. 0; \( P=0.08 \)), systolic blood pressure (131.94 ± 11.79 vs. 125.53 ± 12.39 mmHg; \( P=0.13 \)), diastolic blood pressure (82.17 ± 9.22 vs. 77.24 ± 7.61 mmHg; \( P=0.10 \)) or vitamin D (57.56 ± 25.21 vs. 43.65 ± 22.03 nmol/L; \( P=0.09 \)).
  - Fibroblast growth factor 23 (FGF-23) was higher (145.91 ±176.79 vs. 35.78 ± 58.32 pg/ml; \( P=0.02 \)).
  - Matrix metalloproteinase 2 (MMP-2) was numerically higher (744.38 ± 578.26 vs. 552.93 ± 363.32 pg/ml; \( P=0.25 \)), but statistically not significant.
  - Baseline FGF-23 correlated with MMP-2 (\( r = 0.52 \); \( P=0.03 \)).
  - No significant difference existed in vascular markers at baseline: FMD (4.34 ± 3.45 vs. 4.63 ± 3.02 %; \( P=0.79 \)), NMD (15.15 ± 6.08 vs. 16.00 ± 5.47 %; \( P=0.67 \)).
  - Markers did not change in controls upon follow-up.
  - In KTxPs, FMD decreased (-1.52 ± 2.74 %; \( P=0.03 \)).

Conclusions

- Endothelial dysfunction worsened in stable KTxPs upon long-term follow-up.
- FGF-23 and MMP-2 may contribute to elevated cardiovascular risk by adversely affecting endothelial function in KTxPs.
**Systemic Lupus Erythematosus: A pancytopenic and pancreatic presentation**

B Ahmed MS4, O Chaudhary MS4, M Mustafa MS4, F Siddique MS4, S Shah MS4, B Okwesili MS4

American University of Antigua College of Medicine, Coolidge, Antigua

---

**Introduction**

- Systemic Lupus Erythematosus (SLE) is a multisystem disease leading to various presentation of symptoms.
- Commonly SLE may present with malar rash, photosensitivity, oral ulcers, serositis, arthritis, nephritis, endocarditis, or hematologic abnormalities.
- As a rare initial presentation of SLE, acute pancreatitis presents as generalized flare-ups in cases of patients previously diagnosed with SLE.
- Here we report a rare case of acute pancreatitis and pancytopenia as the initial presentation of Systematic Lupus Erythematosus.

**Case History**

- A 15 year old female patient presented with chief complaints of abdominal pain, nausea, vomiting and non bloody diarrhea.
- The onset of abdominal pain began 4 days prior to admission, localizing to the right upper quadrant on day 2. Associated symptoms consisted of nonbloody diarrhea and nonbloody/nonbilious vomit for 4 days. Social history was significant for recent travel to Western Europe and Michigan. No contributory past medical history or past surgical history. Patient was on no medications and denies any drug allergies.
- General physical examination revealed a fine lacy rash on all four extremities. Vitals were T 37.9, HR 104, RR 18, BP 99/58. CBC revealed pancytopenia with a platelet count of 115 (150-400 x103/μL), hemoglobin 8.5 (12-15 g/dL) and white blood cell 1.5 (4.5-13 x103/μL). Lipase 489 u/L and Lactate Dehydrogenase 2392 u/L.
- Patient was admitted with diagnosis of pancreatitis with pancytopenia and supportive treatment with investigative studies was initiated.

---

**Laboratory Findings**

<table>
<thead>
<tr>
<th></th>
<th>Lipase</th>
<th>WBC</th>
<th>RBC</th>
<th>Platelet</th>
</tr>
</thead>
<tbody>
<tr>
<td>Day 1</td>
<td>489</td>
<td>1.5</td>
<td>3.62</td>
<td>117</td>
</tr>
<tr>
<td>Day 3</td>
<td>940</td>
<td>1.4</td>
<td>3.63</td>
<td>133</td>
</tr>
<tr>
<td>Day 5</td>
<td>1432</td>
<td>1.6</td>
<td>3.48</td>
<td>132</td>
</tr>
<tr>
<td>Day 7</td>
<td>3167</td>
<td>1.5</td>
<td>3.7</td>
<td>116</td>
</tr>
<tr>
<td>Day 9</td>
<td>5590</td>
<td>4.6</td>
<td>3.4</td>
<td>70</td>
</tr>
<tr>
<td>Day 11</td>
<td>&gt;10000</td>
<td>6.6</td>
<td>3.18</td>
<td>111</td>
</tr>
</tbody>
</table>

---

**Clinical Course**

- Laboratory test results indicated positive IgG and IgM antibodies to Brucella, Rickettsia, Ebstein Barr Virus and Parvovirus, complicating the diagnosis. Patient started on doxycycline empirically. Bone marrow biopsy returned negative.
- Day 4, Erythrocyte sedimentation rate was 74 mm/hr, positive antinuclear antibody (ANA) 1/640, positive anti-dsDNA antibody 1/320, positive anti-Smith antibody, negative rheumatoid factor, C3 <40mg/dL, Lipase 2784 u/L. Following lab results and course, the diagnosis of Systemic Lupus Erythematosus was made and steroids were initiated.
- Lipase continued to rise to 8,136 u/L which prompted an MRCP; this showed extensive changes of pancreatitis with upper abdominal fluid, no organized collection or dilation of ducts (Figure 1.1 and 1.2). Also noted was extensive edema of the gallbladder wall without filling defect.
- Lipase began trending down on day 2 of steroids. CBC corrected to within normal limits.

---

**Conclusion**

- Pancreatitis is a rare complication of SLE and only occurs in 2% - 8.2% of patients with SLE.
- A 2010 article published in the Journal of Rheumatology stated acute pancreatitis should be suspected in any SLE patient with abdominal pain and treated with steroids or azathioprine.
- Although rare, physicians should be aware of SLE as an etiology of pancreatitis, when treatment has not lead to the resolution of symptoms and all other etiologies are ruled out.

---

**References**

Cavitary Lung Lesions in a Patient with Positive IGRA and PR3-ANCA are not Always due to TB or GPA: A Case Report of Right-Sided Infective Endocarditis

Toshiro Goto MS1, Hidetaka Yanagi MD, PhD, FACPM, Masayuki Oki MD, PhD2, Hideki Ozawa MD, PhD2
1) Tokai University School of Medicine, Kanagawa, Japan. 2) Tokai University Hospital, Kanagawa, Japan.

Introduction

- Right-sided infective endocarditis (RSIE) may lack peripheral signs and can present with migratory pulmonary infiltrates.
- IE has also been associated with antineutrophil cytoplasmic antibody (ANCA), that could lead to misdiagnosis as granulomatosis with polyangiitis (GPA).

Discussion

- RSIE accounts for approximately 10% of the total IE cases.
- Risk factors: IV drug users; central venous catheters; implanted cardioverter defibrillators; underlying right-sided cardiac anomalies.
- T-valve involvement is common at about 90% of all RSIE.
- Common causative organisms: Staphylococcus aureus, coagulase negative staphylococcus, and streptococcus (pneumococci is associated with alcoholism).
- Enterococcus faecalis accounts for 6.6% of all RSIE cases.
- PR3-ANCA and GPA
- Cytoplasmic and PR3-ANCA for GPA shows sensitivity of 66-92%, specificity of 98-99%.

References

2. Ortiz C et al. Medicine 2014;93:e137
5. Rheumatology 2003;42:223-229

Take Home Points

- We need to keep a high index of suspicion for RSIE in a case of recurrent fever and multiple infiltrates which may be cavitary and increasing in number, even if there are no risk factors.
- For patients with positive vasculitis presenting with positive ANCA, blood culture is one of the most important tests to avoid a diagnostic error seen in this case.
HZO AND ABDUCENS NERVE Palsy

Abstract

Herpes Zoster ophthalmicus is a localized disease, caused by Varicella zoster, characterized by unilateral or hemifacial vesicular eruptions with pre- eruptive erythema, or localized scalp tenderness. There are very few reports focusing on ocular motor palsy / paresis in patients of HZO. We report a case of Abducens nerve / lateral rectus palsy with Herpes zoster. Abducens nerve palsy developed in the post-eruptive phase without any other cause. Various theories for the sixth nerve involvement have been postulated like independent motor neuritis itself, associated intracavernous radiculo-meningitis or perivascular myositis, but nothing very conclusive.

In management, early quick treatment is the cornerstone to limit HZO related morbidity. Adopting vaccination for age 50 and above and all immunocompromised will further reduce occurrence of this disease and related morbidity complications.

Keywords: Herpes Zoster, Ophthalmicus, sixth nerve palsy

Introduction

Herpes Zoster Ophthalmicus (HZO) is the involvement of the ophthalmic division of the trigeminal nerve and represents 10–20% of all cases of herpes zoster.1-4 The acute course is usually benign, however serious ocular complications have been documented, when associated with nasociliary branch/corneal involvement. Apart from compromised immunity, severity is related to age, older being affected more severely than younger.1-4 We report a rare case of Herpes Zoster Ophthalmicus with horizontal diplopia because of lateral rectus palsy or sixth cranial nerve lesion.

Case Report

A 50 year old male presented with extensive facial swelling and excruciating pain. He had a history of fever, lethargy, scalp tenderness for 2-3 days for which he was taking treatment from a physician.

On ocular examination, patient had extensive localized vesicular eruptions limited to left half of his face with bilateral periorbital oedema, extensive lid oedema of both eyes, left-sided pre-aorticus and sub-mandibular lymphadenopathy with extensive neck oedema.

Patient was unable to open his eyes. The vesicular eruptions were distributed over the ophthalmic division of the fifth cranial nerve on the left side, sparing tip of nose. There was bilateral mechanical ptosis but lifting the lid manually revealed no motor deficit or diplopia. There was moderate conjunctival congestion without any corneal involvement.

Seeing the clinical picture, patient was put on medical management and further investigation after processing informed consents. The patient was non-diabetic, with normal Renal and Liver function. Total cell count revealed an increase i.e. 12000/cumm with a neutrophil dominance. There was a history of headache and fever. A Medical and Dermatology reference was sought and patient underwent CSF examination, which revealed higher values of IgM antibodies. Other findings of CSF were insignificant. Patient was prescribed oral Acyclovir 800 mg 5 times a day, Ibuprofen 400 mg twice a day. Pantoprazole 40 mg once a day, oral Multivitamins, Cynocobalamin and topical Acyclovir 3% eye ointment 6 times a day, Mofoxacin 0.3% eye drops 6 times a day. Lubricant eye drops and Acyclovir 5% skin ointment for skin lesions.

After one week, when the patient reported for follow up he was almost cured of the fresh vesicular eruption (phasing out stage), lid oedema and generalized sickness, but he complained of diplopia in primary position.

Re-examination at this stage revealed abduction deficit in the left eye.

Discussion

Herpes zoster ophthalmicus is a sensory neuron disease involving the fifth cranial nerve where the virus stays dormant and waits for the opportunity to attack.5

The most common cranial nerve palsy affecting the ocular motility is the sixth nerve while the most commonly involved motor nerve in herpes disease is the third nerve.6

The mechanism by which these motor neurons are involved is still not clear but many theories have been put forward. It has been postulated by Brown et al that motor neuritis is independent of the inflammation of any ganglion.6

Edgerton postulated that the involvement of the second, third, fourth, fifth cranial nerves is attributable to cavernous sinus inflammation or intracavernous radiculo-meningitis.7

Muscle ischaemia remains a strong possibility as a combination of orbital nerve and muscle inflammation. Hence Abducens nerve palsy may be caused by localized orbital myositis or lymphocytic cranial motor neuropathy. All these patients of HZO must be added with Neurotropic drugs in the beginning and must be watched for any association of motor deficit during their follow up course.

Conclusion

There are numerous probabilities, either working separately or simultaneously which are responsible for motor nerve deficits along with HZO. These patients warrant close follow up for any motor deficits along with Neurotropic drugs. If we adopt vaccination for individuals above 50 year of age and the immunocompromised, this will further reduce HZO occurrence and its related morbidity complications.

References

Care Coordination: Scalable Student-led Strategies
To Address Social Determinants of Health in the Greater Camden Region
Jessica W. Oribabor1, Dr. Anjali Desai2, and Dr. Behjath Jafry2
1Cooper Medical School of Rowan University, 2Cooper University Hospital Department of Internal Medicine, Camden, NJ

Since inception of the Department of Health and Human Services’ Healthy People 2020 initiative, there has been a strong emphasis to understand social determinants of health that negatively impact clinical outcomes and promote local partnerships to address these barriers to care. Student-run free clinics (SRFC) serve a unique role to not only offer primary care services to the medically uninsured, who are more vulnerable to such barriers, but also be a local community partner.

The Cooper Rowan Clinic (CRC) in Camden, NJ offers this assistance through Care Coordination (CC), a student-led social work (SW) program. As the program’s popularity grew, CC faced a large administrative burden due to its paper-based records and was without a formal process to track patient outcomes over time.

**Primary Objectives**
- Implement a novel social work model based on a free, scalable IT interface in REDCap.
- Create a centralized online database to facilitate the evaluation of patient social needs, like an Electronic Health Record.
- Eliminate program redundancies: improve timeliness, paperwork legibility, & productivity.
- Gain new insight into the patient population.

**Why REDCap?**
- Server-based software (PHI not stored on devices)
- Free, multi-user web application
- HIPAA Compliant
- Real-time analytics
- Modifiable templates to facilitate survey updates and connecting platform (Figure 1A)

**Methods**
1. Evaluated previous SW model's weaknesses and gaps in workflow to formulate REDCap database structure. (Figure 2.1)
3. From Jan 2018-Aug 2018, all CRC patients received the new paper CC intake survey (Figure 1A) before each clinical visit.
4. Volunteers recorded responses & recommendations into REDCap (Figure 1B) at the end of the clinical encounter via HP 210 G2 tablets or personal device.
5. Collectively forms created a centralized database of all patient self-reported social needs & resources distributed.

**Care Coordination Surveys & Database Structure**

**Figure 1. Comparison of the Care Coordination paper survey (1A. Left), and expanded digital version in REDCap (1B. Right) which automatically prompts the user to suggested local resources & services.**

**How to Implement:**
1. Interview members of the patient community to gauge unique needs.
2. Coordinate with hospital’s inpatient Social Work Dept for more resources and perform site visits.
3. Create paper based survey based on research and ensure all Q's have consistent plan of action, i.e. flyer or phone number, address, etc.
4. Program survey into REDCap.
5. Pilot 50-70 patients.
6. Train volunteers on database management and survey administration.
7. Advertise social work program launch at student clinic and incorporate into workflow.

**Who Do We Serve?**

<table>
<thead>
<tr>
<th>N = 291 patients*</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Care Coordination Visits</td>
<td>500</td>
</tr>
<tr>
<td>Mean Age (SD)</td>
<td>47.5 (+/- 12.5)</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>128</td>
</tr>
<tr>
<td>Female</td>
<td>163</td>
</tr>
<tr>
<td>Ethnicity</td>
<td></td>
</tr>
<tr>
<td>Hispanic</td>
<td>209</td>
</tr>
<tr>
<td>African-American</td>
<td>39</td>
</tr>
<tr>
<td>Caucasian, Asian, Other</td>
<td>41</td>
</tr>
<tr>
<td>City</td>
<td></td>
</tr>
<tr>
<td>Camden Residents</td>
<td>182</td>
</tr>
<tr>
<td>Non-Camden Residents</td>
<td>108</td>
</tr>
<tr>
<td>Top Reported Social Needs*</td>
<td></td>
</tr>
<tr>
<td>Medication Affordability, Dental Care, Health Insurance Enrollment, &amp; Transportation</td>
<td></td>
</tr>
</tbody>
</table>

* Surveys in the "No Date Visit Arm" and patients without records in EPIC were excluded from analysis.
** Reported at initial visit

| Table 1. Demographic Data from Jan to Aug 2018

**Conclusion**
- Innovative SRFC social work services are necessary to provide comprehensive care to vulnerable and uninsured patient populations. REDCap is a free, and scalable IT. solution to track and analyze social determinants of health that disproportionately affect these patient populations. The findings from this project proposed to demonstrate an efficient model of student-led social work care and guidelines for the implementation of similar programs at various SRFC nationwide.
- Lessons Learned / Future Goals
  - Increased volunteer participation with personal laptop use compared to HP tablets (dropping hazard, incompatibility w/ REDCap App).
  - Database facilitated continuity of care between different volunteer shifts each week.
  - Discovered the most frequently recommended resources/services and initiated early interventions based on patient feedback.
  - Plan to map and stratify reported social needs & resource utility using entire patient database.

**References**

Acknowledgements: The support for this project by Cooper Medical School of Rowan University, Cooper Research Institute, and Rowan Library Services is gratefully acknowledged.
Wernicke’s Encephalopathy Presenting as Sensorineural Hearing Loss

Benjamin Bryant, BA1, David Hewson, MD2, Paul Ehrlichman Jr, MD2, Christopher Chu, MD2, John Sanders III, MD, MPH3

Wake Forest School of Medicine, Winston-Salem, NC1
Department of Internal Medicine, Wake Forest Baptist Medical Center, Winston-Salem, NC2
Department of Infectious Disease, Wake Forest Baptist Medical Center, Winston-Salem, NC3

Introduction:
- Wernicke’s Encephalopathy (WE) is a known complication following bariatric surgery (1).
- Although it is known that patients with WE often do not present with the classic triad of ophthalmoplegia, confusion, and ataxia, bilateral sensorineural hearing loss (SNHL) is a rare presenting symptom and its incidence is unknown. The mechanism of its pathogenesis is thought to be due to involvement of the ascending afferent auditory fibers in the brainstem (2).
- Unfortunately, the diagnosis of WE is often missed in its early stages (3, 4) due to the variability in presentation as well as the fact that thiamine levels can take days to result.

Learning Objectives:
- Investigate an atypical presentation of Wernicke’s Encephalopathy (WE)
- Discuss how a low thiamine level is the only specific laboratory marker for WE

Conclusions:
- Elevated inflammatory markers in the setting of WE can be misleading, as they are nonspecific and do not necessarily suggest an autoimmune etiology.
- Besides a low thiamine level, no laboratory markers are specific for the diagnosis of WE (5).
- For at-risk patients presenting with neurologic complaints, one should have a high index of suspicion for WE and start early thiamine supplementation to prevent irreversible neurodegeneration (6).

References:

Clinical Vignette:
- 22-year-old African-American female presents 2 months status post sleeve gastrectomy with 10 days of decreased oral intake, bilateral lower extremity weakness, and hearing loss.
- She was seen 3 days prior at outside hospital where she was treated for dehydration.
- She was given no diagnosis, nor did symptoms resolve.

Hospital course:
- Initial thiamine level resulted low at 17 nmol/L.
- Thiamine supplementation resulted in gradual symptom improvement, and patient was discharged.

Day 0
- Initial laboratory workup showed fasting ketosis
- Thiamine level was drawn

Day 1
- EGD showed no evidence of stricture
- Nutritional supplementation was started
- Audiometric testing showed bilateral SNHL

Day 2
- Autoimmune workup: CRP of 44.9 mg/L, ESR of 83 mm/hr, and positive rheumatoid factor
- Empiric steroids were started

Day 3
- MRI Brain: bilateral mamillary body edema and enlargement
- Empiric steroids were stopped

Day 0
- Initial laboratory workup showed fasting ketosis
- Thiamine level was drawn

Day 1
- EGD showed no evidence of stricture
- Nutritional supplementation was started
- Audiometric testing showed bilateral SNHL

Day 2
- Autoimmune workup: CRP of 44.9 mg/L, ESR of 83 mm/hr, and positive rheumatoid factor
- Empiric steroids were started

Day 3
- MRI Brain: bilateral mamillary body edema and enlargement
- Empiric steroids were stopped

Day 3
- MRI Brain: bilateral mamillary body edema and enlargement
- Empiric steroids were stopped
Introduction

There are approximately 20 million college students in the United States, and despite medical issues ranging from sniffles to sexually transmitted diseases, these students still have to attend class, complete homework, and study for mid-terms.

Numerous studies are noticing a shift in the way millennials access medical care, choosing Acute Care Clinics or “Dr. Google”, in comparison to older generations who prefer Primary Care Providers.

Current research doesn’t specifically focus on the reasons behind why college students avoid seeking medical care.

When students avoid healthcare, it not only affects the individual student, but may also lead to the spread of communicable diseases in the classroom and throughout campus.

This study was an exploratory analysis to identify and understand the factors resulting in healthcare avoidance in college students.

Methods

Participants:

- 270 Boise State University student respondents
- Mean age: 19.3 years old, SD of 2.5 years, Range from 18 - 33 years old.
- Research approved by the Institutional Review Board of Boise State University.

Materials & Procedure:

- 84 question computer based anonymous survey
- Statistical tests were run using SPSS version 23

Apprehension in Students Covered by Family Health Insurance:

- Significantly more likely to self-treat the symptoms of sadness and depression
- 70% of students covered under parent’s insurance would avoid seeking professional mental care when suffering from overwhelming stress
- Significantly less likely to seek care from a mental health counselor for symptoms of sadness and depression
- Significantly more likely to worry about hidden fees and additional costs outside the coverage of their parents’ insurance plan
- Significantly less apprehensive of medical care in general, than uninsured students

Apprehension in Female College Students:

- Significantly more worried about their parents asking the details of their recent medical visit
- Significantly more likely to avoid care due to the embarrassment of being seen walking into the clinic by someone they know
- Significantly more likely to attend class when suffering from a sudden illness instead of seeking medical care
- Significantly more likely to ignore the symptoms of debilitating stress
- Significantly more likely to look up symptoms online before visiting the doctor

Apprehension in Male College Students:

- Significantly more likely to use online resources to diagnose and treat a suspected sexually transmitted infection, instead of seeking care from a physician
- Significantly more likely to self-treat the symptoms of stress, sadness, and depression
- Significantly more likely to choose medical care over attending class or completing homework assignments
- Less likely to worry about their parents asking the details of their checkup

Discussion

Students are suffering in silence, and not seeking care for elevated levels of stress, panic attacks and depression.

Our research suggests this could be the result of underlying apprehensions from being on a parents’ health insurance policy, or may choose to avoid care due to stigma.

The previous findings of millennials preferring “Dr. Google” and self-treatment was mirrored in our findings.

Although students covered by family insurance were less apprehensive of professional care in general, female students like to check their symptoms online before seeing the doctor, and male students prefer online resources and self-treatment when they suspect a sexually transmitted infection.

Recommendations

- Campus health centers can highlight their student-focused confidentiality and prompt checkup times
- Medical clinics near college campuses can advertise sliding-fee scales and the enhanced confidentiality of paying out-of-pocket for services
- College newsletters can highlight the benefits of seeking care from healthcare professionals

References

Outpatient Parenteral Antibiotic Therapy (OPAT) Experience at a South Carolina Referral Hospital

Coursen J, Schrank C, Schrank J, Roth P
Greenville Health System

Introduction
Outpatient parenteral antimicrobial therapy (OPAT) allows patients to receive intravenous antibiotics for the treatment of infectious diseases outside of the inpatient hospital setting. First described as a treatment option in the United States in 1974, studies have since established OPAT as an acceptable treatment alternative to reduce healthcare expenditure, decrease hospital length of stay, and increase patient satisfaction. Despite its benefits to both the physician and patient, OPAT continues to pose significant risks, including treatment failure, hospital readmissions, adverse drug events, and catheter-related complications. Many studies have contributed their outcomes to the body of OPAT literature, but rates of complications and treatment success have varied markedly. By retrospectively reviewing our database of OPAT patients at Greenville Health System, we hope to gain a better understanding of the factors influencing outcomes. This will enable us to optimize the OPAT experience going forward for our patient population at an academic tertiary center in rural South Carolina.

Aim
To identify the factors associated with successful treatment outcomes and also with adverse events, to include:
- Readmission rates
- Adverse Events
  - Catheter related events
  - Medication side effects
- Need for retreatment

Methods
- Retrospective chart review of 336 consecutive patients who were treated with OPAT under the direct guidance of the Greenville Health System’s Infectious Disease Division between 01/01/17 and 06/30/17. Therapy was initiated in both inpatient and outpatient setting
- Demographic information collected included age, sex, and insurance coverage
- Clinical criteria
  - Indication
  - Choice and duration of parental antimicrobial therapy
  - Concomitant usage of oral antibiotics
  - Location of OPAT (home vs infusion center)
- Outcome variables
  - Successful completion of planned course of antibiotics
  - Missed doses (number and frequency)
  - Need for hospital (re)admission
  - Catheter related infections
  - Therapy related side effects
  - Medication changes and reason for change
  - Documented C. difficile infection
  - Relapse of infection at same site with same organism within 6 months following completion of OPAT
  - Death
- Statistical Analysis performed using Chi-squared Regression Model

Results

Clinical Indications for Outpatient Parenteral Antibiotics

- 336 individual patients received OPAT during the 6 month period
- Average Age 59.5 years
- 57% Male; 43% Female
- Insurance Coverage: 46% Medicare; 25% Private; 14% Medicaid; 15% None/other
- Bacteremia (25.4%); Osteomyelitis (14.9%) and Diabetic Foot Infections were the most common indications for OPAT (see chart)
- MSSA was the most common organism treated
- Completion of planned course: 94%
- Average duration of treatment: 4.78 weeks
- 11% of patients required a change in medication during the course of OPAT; the most common reason was Nausea/Malaise (26%) and AKI (26%)
- Hospital readmission rate was 8.7%
- Home Infusion was significantly more likely to result in hospital readmission compared to receiving OPAT at an infusion center (P<0.02)

Antibiotics Usage Among 336 Patient Receiving OPAT

- % of OPAT
- Ertapenem/Meropenem
- Other Cephalosporin
- Vancomycin
- Other
- Daptomycin
- Continuous penicillin
- Other

Discussion
Our retrospective review found similar outcomes regarding hospital readmission rates and similar rates of adverse events requiring medication change as prior studies. A novel finding in our study was the increased risk of hospital readmission in patients who received their therapy at home as opposed to receiving it at an infusion center. This difference may be related to factors such as administration technique, compliance, or more severe illness. We should further investigate to see if the risk holds true for a larger population. If so, then the different factors associated with home infusion versus an infusion center should be analysed to identify those which increase the risk of hospital readmission.

References
A Rare Presentation of Aberrant Papillary Thyroid Cancer

Mir Inzamam Ali1 and Sasmith Menakuru2
1. RAK Medical College, UAE 2. Naryana Medical College, India

Abstract

To describe extra thyroid tissue masses in the neck, especially lateral to the jugular vein, Albers in 1829 coined the term "lateral aberrant thyroid". However, this term was only limited to thyroid tissue found on the lateral side of the neck and did not consider the possibility of presence of thyroid tissue at any other site. Defects in embryogenesis during its usual pre-tracheal position in the neck have resulted in lateral aberrant thyroid. A berrant site describes but is not limited to: (a) lying lateral to the jugular vein, (b) along the thyroglossal tract, (c) intrathoracic/mediastinal with no connection to the main thyroid body (d) found in teratoid tumors at a distance from the thyroid [1-3]. "Struma ovarii" is a term describing ovarian teratoma containing a large amount of thyroid tissue which was biologically and microscopically similar to normal thyroid [4]. Primary malignant transformation of ectopic thyroid tissue is considered rare. It is generally more common in females with an average incidence of 1 per 300,000 cases [4]. Pericarditis, on the other hand, may be caused by any malignant tumor that has metastasized via lymphatics or blood or through direct extension into the pericardial sac causing irritation and inflammation [5]. Awareness of this condition is needed to prevent unwarranted complications. Hereby we present a case of a mediastinal thyroid papillary carcinoma without evidence of cervical thyroid gland involvement.

Case Presentation

A 21-year-old woman presented to the emergency department with chest pain and shortness of breath of three days duration. The pain was pressure-like in sensation, initially 3/4-10 in intensity on severity scale and gradually progressed up to a 7/10 intensity on the day of presentation to the ER. The pain radiated to her right shoulder down to the right arm as well as her mid-back during inspiration. The pain was relieved with naproxen and did not occur at rest. No exacerbating factors were noted other than deep inspiration. She also noticed a gradual decrease in exercise tolerance down to two blocks walking distance. She denied any fevers, chills, nausea, vomiting, diarrhea, constipation, palpitations, abdominal pain, lumps, night sweats or leg swelling. She also denied any recent colds, infections, recent contact with illnesses or travel. Review of other systems was negative. She had no relevant past medical or family history. Her last menstrual period was 18 days ago, and is not sexually active. She has never smoked or used illicit drugs and only drinks alcohol occasionally.

On physical examination she was afebrile, her blood pressure was 120/80, pulse 110 bpm, respiratory rate 18/min, and her oxygen saturation on room air was 99%. On inspection of chest and back, no abnormality was detected. No palpable thrills. On auscultation, normal S1 and S2 sounds were heard with no murmurs or additional sounds. Chest auscultation revealed normal breath sounds with no wheezes or crepitations. Abdomen was lax and non-tender without any palpable masses. No other notable findings on physical examination.

Initial Investigations and Workup

ECG showed tachycardia, PR depressions and diffuse ST-elevation. Labs showed microcytic anemia of 9.8 g/dl with an MCV of 62 fl, WBC of 12.1 K/cul with a differential leukocyte count of 44% trypsin negative, and coagulation profile was within normal limits. ESR was 42 mm/hr. Urinalysis revealed positive leukocyte esterase without pus or RBCs. On chest Xray, a central shadow suggestive of mediastinal mass was seen thereby a CT angiography with contrast of chest was requested. The CT showed a 5.1 cm right anterior mediastinal mass with 1.7 cm right paratracheal lymph node mass. Transthoracic Echocardiogram was done, it was free with no evidence of pericardial effusion with an EF of 77%.

Analysis and Further Workup

The presence of a lymph node mass raised suspicion of lymphoma. Further investigations were done to rule out different possibilities. Alpha Fetoprotein (AFP) was 1.1 ng/ml, beta HCG was less than 1.2 mIU/ml, LDH was 472 U/l and TSH was 3.56 mIU/l. A provisional diagnosis of pericardial secondary to the mediastinal mass was made, and the patient was admitted and consequently started on coldkine 0.6 mg b.i.d and ibuprofen 600 mg q.i.d. Patient reported initial improvement in treatment in the hospital just after one day. Her chest pain decreased. Cardiothoracic surgery was consulted for biopsy. Lymph node fine needle aspiration biopsy (FNAB) was planned through CT guidance due to the close proximity to the heart and great vessels. FNAB of the lymph nodes was inconclusive. Thereby, CT guided core biopsy of the mediastinal mass was scheduled and done. Four 18 gauge core biopsy specimens were obtained. Pathology specimens obtained were placed both in formalin and in Roswell Park Memorial Institute medium (RPMI) for culture and flow cytometry. Procedure was performed with no complications.

Treatment

The patient was initially treated with 0.6 mg omeprazole b.i.d along with ibuprofen 600 mg q.i.d for her acute pericarditis. Symptoms improved within a day and she was subsequently discharged to await biopsy results. As per biopsy results diagnosing papillary cell carcinoma of an aberrant thyroid, surgery was planned by the cardiothoracic surgeons to avoid further complications. She underwent sternotomy for complete resection of the aberrant thyroid with lymph node dissection. The primary thyroid gland was not operated on as there was no indication.

Summary

Malignant transformation of a mediastinal aberrant thyroid is a rare condition. In our case, a 21-year-old female patient presented with chest pain, investigations had revealed a papillary carcinoma of an aberrant mediastinal thyroid mass. ECG showed sinus tachycardia, PR depression and ST segment elevations suggesting acute pericarditis. CT-guided core biopsy of the mediastinal mass confirmed presence of malignant thyroid tissue leading to the diagnosis of papillary carcinoma. The commonest type of thyroid cancer. Cases of idiopathic pericarditis raise the suspicion of malignancy which should be included in the differential diagnosis. Thorough work up of patients with initial presentation of acute pericarditis should be done to exclude malignancy. This case also sheds a light on the possibility of intrathoracic thyroid carcinoma presenting with local symptoms (pericarditis). Prompt and early diagnosis of such cases should be warranted to avoid metastatic complications.

Key Points

Pericarditis can be a marker of malignancy and any case with an initially unknown cause should be investigated further. Biopsy of all masses in the anterior mediastinum must be stressed upon and further treatment should be of utmost importance. Carcinoma of an aberrant thyroid can occur without involvement or symptoms of primary thyroid gland cancer.
Acute onset of focal neurological deficits correlated to a single vascular distribution reflexively necessitates stroke assessment to identify ischemia or hemorrhage. Patients that present within 24 hours of symptoms are defined as having a transient ischemic attack if no acute pathology is noted by CT or MRI. To further stratify risk, further imaging of the carotid arteries and assessment of comorbidities such as hypertension, hyperlipidemia, and diabetes is performed and prophylactic medications like statins are initiated. However, recurrent symptoms without underlying unaddressed comorbidities raises suspicion for an alternative etiology. A Simple Partial Status Epilepticus (SPSE) is a seizure with an electrical surge affecting a focal area producing typical symptoms without impairment of consciousness. We describe the case of an 82-year-old male with multiple CVA comorbidities that presented for recurrent ephemerol neurological deficits within the same vascular distribution for 5 years presumed to be TIAs.

CASE

HOSPITAL COURSE:
The patient was extensively worked up for similar symptoms for the last 5 years with CT, CTA, MRI, MRA, and Ultrasound of the carotid arteries, and only once did the MRI show remote lacunar infarcts in the bilateral basal ganglia and right pons. As with previous admissions, the stroke protocol was initiated and the patient had a CT scan and preventative CVA reinfarction therapy was initiated. Once the CT scan revealed no abnormalities, MRA and MRI of the brain were ordered and revealed no acute ischemia or infarct. Patient was initiated on Cardizem for suspected cerebral vasospasm; however, he returned to the Emergency Department with recurrent symptoms. Once again, the stroke protocol was initiated and the findings were not significant for acute pathology. However, based on his focal neurological symptoms he was started on and responded well to Keppra without recurrence of symptoms.

DISCUSSION:

- Cerebrovascular disease is the leading cause of aphasia; however, aphasia can also occur secondary to traumatic, epileptic or neurodegenerative conditions.
- Multiple imaging modalities were used to exclude stroke, which revealed multiple remote lacunar infarcts in the bilateral basal ganglia and right pons 1 year prior and no imaging since or before those findings.
- A 24-hour EEG was unable to capture abnormal wave forms while the patient was admitted and treated with Keppra.
- Rosenbaum et. al2 proposed a diagnostic criteria for epileptic aphasia that was subsequently modified by Grimes and Guberman4:
  i. The patient has language production during the seizure.
  ii. Language production shows aphasic features.
  iii. Consciousness is preserved.
  iv. The seizures are correlated with the aphasia, as documented by EEG monitoring and behavioral testing.
  v. The aphasia resolves, or nearly so, concurrent with successful treatment of the seizures.
- Because the patient’s presentation was concerning for CVA, the stroke protocol takes precedence over workup for seizure disorder. Unfortunately therefore, an EEG was not performed at the time of his symptoms. However, his marked response to Levetiracetam and an absence of recurrence of symptoms strongly correlate to an underlying seizure disorder.
- Regardless of historical context, patients with acute focal neurological deficit localized to an arterial distribution require a stroke assessment. While prior imaging and patient history may guide clinical decision-making regarding aggressiveness in risk stratification and further imaging, initial evaluation with non-contrast CT or MRI for intracranial pathology and hypercoagulability workup are imperative. Unfortunately, in a patient with recurrent intermittent symptoms, that leads to excessive radiation, bloodwork, and medication readjustments to rule out and prevent strokes while the true etiology remains elusive. In patients confirmed to not be experiencing CVA or TIA in a recurrent setting, a trial of antiepileptic medications may uncover a partial status epilepticus.

REFERENCES:
**What’s at the Heart of this Ischemic Stroke?**

Anna E Berry, BS, Lauren Nicholas Herrera, MD, Basant Arya, MD  
Baylor College of Medicine and Baylor St. Luke’s Medical Center, Houston, TX

**LEARNING OBJECTIVES**
- Review presentation and cardiac workup of ischemic stroke
- Describe echocardiographic and pathological findings associated with papillary fibroelastoma

**HISTORY OF PRESENT ILLNESS**
- Pt. is a 65 y/o woman with a history of hyperlipidemia on simvastatin and hypertension controlled by losartan-HCTZ who presented with sudden onset left arm weakness, left leg weakness, and dysarthria
- Had intermittent, self-resolving palpitations, but never formally diagnosed with an arrhythmia
- Denies chest pain, lightheadedness, history of stroke, and family history of clotting disorders
- Pt. is a 65 y/o woman with a history of hyperlipidemia on simvastatin and hypertension

**PHYSICAL EXAMINATION**
- T 98.0°F HR 69 BP 128/75 RR 24 SaO2: 96%  
- Cardiovascular: Normal S1, S2. Regular rate and rhythm. No murmurs, rubs, gallops.
- Neurologic: Strength: 4+/5 in the hip flexors, knee flexors, and knee extensors. Sensation: Decreased to left leg and face as compared to right.
- Cranial Nerves: Trigeminal nerve (V) with decreased sensation to light touch in the left V1-V3 distribution

**TESTS/LABS/IMAGING**
- MRI Brain: Restricted punctate and patchy ischemic changes in the right MCA territory.
- Electrocardiogram (ECG): Regular rate and rhythm without ST changes
- Transthoracic and Transesophageal Echocardiogram: 1cm x 1cm x 1.3cm large, mobile echodensity that is homogenous and well-circumscribed with a visible stalk attached to the base of the left coronary cusp of the aortic valve. No patent foramen ovale
- CT Head negative
- NIHSS 15
- CT A** with possible right M2 partial occlusion
- NIHSS 2
- NIHSS 9
- CT Ao VVG: Central fibrous core strongly positive for elastic fibers
- H&E Stain: Papillary configuration with a fibrous core, endothelial lining, and surrounding myxoid-like material at the periphery of each papillae
- Verhoeff–Van Gieson (VVG) Stain: Central fibrous core strongly positive for elastic fibers
- Alcian Blue Stain: Cortical fibers strongly positive for elastic fibers

**FINAL DIAGNOSIS**
Cardiac Papillary Fibroelastoma of the Aortic Valve

**MANAGEMENT & FOLLOW UP**
- Initiation of heparin drip
- Minimally Invasive Surgical Removal of Papillary Fibroelastoma without Valve Intervention
- Recovered uneventfully and was discharged home on postoperative day 3 with 3 week cardiology follow-up

**DISCUSSION**
- Embolic cardiac phenomena are responsible for 15-30% of ischemic strokes.
- The incidence of primary cardiac neoplasms is low (0.02%), with cardiac papillary fibroelastomas (CPF) being the second most common primary cardiac tumor.
- Most CPFs are asymptomatic and discovered incidentally.
- Symptomatic CPFs most commonly manifest with embolic phenomena to the cerebral, systemic, or coronary circulations. In rare cases, they can present with heart failure or sudden death.
- Tumor mobility is an independent predictor of CPF-related death or nonfatal embolization.
- Echocardiographic findings of CPFs include an intracardiac mass that is speckled with echolucencies and have a stippled pattern near the edges.
- Pathology findings demonstrate an avascular papilloma with a single layer of endocardial cells covering a papillary surface with a matrix comprised of proteoglycans and elastic fibers.
- Surgery is the first-line treatment for symptomatic or highly mobile CPFs, as it has shown to be highly effective, safe, and curative. If the patient is not a surgical candidate, they can be managed with oral anticoagulants.

**CONCLUSIONS**
- Though rare, ischemic sequelae from embolic cardiac tumors such as CPFs should be considered in the differential diagnosis of ischemic stroke.
- Initial cardiac workup includes ECG and TTE. If a cardiac neoplasm is detected, it may be further characterized by transesophageal echocardiogram and cardiac MRI.
- If symptomatic or highly mobile, gold standard treatment is surgical removal of the papillary fibroelastoma. If the valve is compromised, repair or replacement of the valve may be necessary.

**REFERENCES**

*National Institutes of Health Stroke Scale  
**Computed tomography angiogram*
**INTRODUCTION**

A patient presenting with an unknown etiology of anemia is a commonly encountered scenario in internal medicine. A patient presented to the University of Texas Medical Branch with an unusual case of a severe macrocytic anemia with evidence of hemolysis.

**CASE PRESENTATION**

History of Present Illness:
- 35-year-old female with no significant past medical history presented to the hospital with dyspnea on exertion and dizziness.
- Four month history of fatigue, jaundice, 12 pound weight loss
- Non-vegetarian, vegan, or restrictive diet
- Four month history of fatigue, jaundice, 12 pound weight loss

**Differential Diagnoses**
- Vitamin B12 or folate deficiency due to impairment of nucleic metabolism, including pernicious anemia
- Reticulocytosis secondary to hemolysis, liver disease, hypothyroidism, or alcohol use

**Diagnostic Work-Up**

Turning Point in The Case:
- After her blood smear demonstrated numerous hypersegmented neutrophils, the diagnosis of vitamin B12 deficiency was suspected.

**Diagnostic Imaging:**
- Chest x-ray and abdominal x-ray normal
- Head, ears, eyes, nose, and throat: scleral and sublingual icterus
- Cardiac: normal S1 and S2, normal rate and rhythm
- Lungs: normal breath sounds bilaterally
- Abdomen: normal bowel sounds, non-palpable liver or spleen, non-tender, non-distended
- Neurological: cranial nerves II through XII grossly intact; extremities, normal proprioception and vibratory sensation
- Muscle strength 4 out of 5 in all four extremities, normal proprioception and vibratory sensation
- Spleen, non-tender, non-distended

**Analysis of Laboratory Data**

- HGB: 6.7 (L) 8.1 (L) 9.1 (L)
- MCHC: 33.6
- RDW-CV: 26.5 (H) 18.6 (H)
- MCV: 109.2 (H)
- RDW-SD: 124.6 (H)
- PLT: 136 (L)
- WBC: 5.24
- AST (SGOT): 157 (H)
- ALK PHOS: 59
- T PROTEIN: 9.1 (H)
- CALCIUM: 8.9
- TIBC: 100.0 (H)
- HCT: 13.1 (LL)
- TSH: 0.83
- VIT B12: <159 (L)
- FOLATE: >20.0 (H)
- TPO Ab: >1,000.0 (H)
- FREE T4: 0.83

**DISCUSSION**

- Typically, vitamin B12 deficiency can present with a megaloblastic anemia, often accompanied by neurological symptoms, pancytopenia, or jaundice.
- One cause of vitamin B12 deficiency is pernicious anemia.
- Pernicious anemia, an autoimmune disorder, is caused by antibodies targeting intrinsic factor or gastric parietal cells.
- Pernicious anemia is associated with other autoimmune disorders, including hypothyroidism.
- Parietal cells release intrinsic factor, which binds to vitamin B12 and is required for absorption in the terminal ileum.
- In this case, the her vitamin B12 deficiency secondary to pernicious anemia was clouded by a hemolytic picture.
- The hemolysis is due increased fragility of erythrocytes and accumulation of homocysteine causing damage to important cellular mechanisms, resulting in increased hemolysis.
- While this patient received blood transfusions for a hemoglobin of 4.4 g/dL, transfusions are not usually necessary and vitamin B12 supplementation often corrects the anemia promptly.
- Early recognition of this diagnosis will shorten hospital courses with rapid symptomatic improvement and avoid unnecessary blood transfusions with its associated risks.
- It emphasizes the importance of understanding that a severe vitamin B12 deficiency can present as hemolytic anemia.

**REFERENCES**


**ACKNOWLEDGEMENTS**

John P. McGovern Academy of Oslerian Medicine for travel arrangements, Karen Szauter, MD for her mentoring and support, Jack Alperin, MD, for his mentoring and help with hematology photos, and Bilal Mohammad Bilal, MD, for his mentorship and support.
**Introduction**

- Dermatomyositis (DM) is a systemic inflammatory disease that affects skin, muscle and lung.
- Auto-antibodies against anti-melanoma differentiation-associated protein 5 (MDA5) are linked with the unfavorable lung variant of DM which presents with rapidly progressive Interstitial Lung Disease (RP-ILD), more severe dermatosis, and mild myopathy.1

**Case Presentation**

**HPI:**
A 44-year-old male with no past medical history presents with worsening fatigue, dyspnea, and pain in his fingers, hands, wrists, and shoulders over the past 3-4 months.

**Physical exam:**
Palms with multiple punctate erythematous lesions as well as bush-like violaceous capillary loops on proximal nail folds. Pulmonary exam revealed bilateral inspiratory fine crackles.

**Labs:**

**Clinical Course:**
He developed worsening dyspnea and hypoxia. His extended myositis panel from the prior week resulted with positive MDA5. He was treated with cyclophosphamide, mycophenolic acid, and pulse dose steroids. Unfortunately his respiratory status continued to decline and he passed away two weeks later.

**Radiology**

- Figure 1: Chest CT with patchy ground-glass opacities at the periphery and lung bases with mild cylindrical bronchiectasis, consistent with a nonspecific interstitial pneumonia pattern.
- Figure 2: Repeat CT chest 14 days later with interval development of bilateral fine reticulations with worsening ground-glass opacities and mosaic attenuation with areas of air trapping.

**Discussion**

- The Bohan and Peter criteria for diagnosis of DM:4
  - Clinical evidence of myopathy and dermatosis
  - Elevated lab levels of inflammatory auto-antibodies
  - Microscopic evidence of inflammation on muscle biopsy
  - Electromyographic data
- Identification of the specific auto-antibody is crucial as each antibody confers a different natural history and mortality.
- Specifically, anti-MDAS antibodies are linked with rapidly progressive lung disease with a high mortality.

**Summary and Conclusions**

- Anti-MDAS antibody-positive DM predicts the presence of RP-ILD and is associated with very poor prognosis with ninety day mortality as high as 66.7%.5
- Due to rapid deterioration, early imaging with high resolution CT, diagnosis, and aggressive therapy are extremely important.
- In some cases, it is possible for pulmonary findings to precede the typical joint, skin, and muscular symptoms associated with autoimmune diseases.

**Expected Exam and Pathology Findings**

- Figure 3: Photograph of erythematous lesions on palmar aspect of patient with MDA5-positive dermatomyositis.2
- Figure 4: Muscle biopsy in a patient with DM showing inflammatory infiltrate at perivascular site. 4

**References**

**INTRODUCTION**

Dermatomyositis is amongst the idiopathic inflammatory myopathies that manifest with underlying muscular weakness, elevated serum muscle enzymes, and photosensitive dermatitis. We describe a case of longstanding, undiagnosed dermatomyositis complicated by diagnostic delay related to the patient’s lack of health insurance.

**Case:** A 29-year-old female presented to a Federally Qualified Health Center Rheumatology clinic (FQHC-Rheum) for evaluation of facial swelling, myalgia, and photodermatitis. In the preceding two years, she presented multiple times to emergency departments and acute-care clinics. Systemic corticosteroids controlled the symptoms, however recurrence was noted with discontinuation of each course. After presenting to a student-run, dermatology clinic, the patient was instructed to undergo evaluation by a rheumatologist. She presented to the FQHC-Rheum clinic on prednisone 20mg daily with a widespread erythematous, maculopapular rash. She exhibited normal muscle strength (5/5) in the proximal extremities and neck flexors/extensors. Laboratory analysis revealed minimally elevated ALT and AST concentrations, normal creatine kinase (CK) concentration (131 U/L; reference: 26 – 192) and a negative anti-nuclear antibody (ANA). Prednisone was rapidly tapered. Within two weeks, the dermatitis, generalized swelling and myalgia recurred. Muscle strength and CK (138 U/L) remained normal, prednisone was resumed, and symptoms improved. An autoimmune connective tissue disease was suspected, however further evaluation was halted due to the patient’s financial status. Two months after initial presentation to the FQHC-Rheum, the patient returned with moderate weakness and worsening photodermatitis. Despite normal CK levels (81 U/L), muscle strength testing revealed proximal hip flexor weakness (3+/5), and the patient was diagnosed with dermatomyositis. She elected to forego an extremity MRI, electromyography, muscle biopsy, and screening CT scans due to prohibitive costs. The patient subsequently obtained Medicaid health insurance, and a thigh MRI confirmed the diagnosis of dermatomyositis nearly 36 months after symptom onset.

**Discussion:** Patients with dermatomyositis often endure long diagnostic delays. An uninsured status further limits the prompt and accurate diagnosis of rheumatic disease [1]. Typically, the diagnosis of dermatomyositis is based upon a clinical presentation of muscle weakness, elevated serum muscle enzymes, electromyography findings, MRI results and characteristic muscle histopathology [2]. Uninsured patients are more likely to refuse or postpone diagnostic evaluation and choose not to fill necessary prescriptions due to cost [3].

**Conclusion:** A lack of health insurance had detrimental effects on the timing and quality of care provided to our patient with dermatomyositis. Initially, she had limited access to specialty care and chose not to pay for cost prohibitive procedures and imaging studies. A recent member survey of the American College of Rheumatology revealed the most common practice-related ethical issues in Rheumatology involve the cost of expensive treatments and the care of uninsured/underinsured patients [4]. These issues proved to be at the forefront of this patient’s care.

**RESULTS**

**Serum Muscle Enzymes**

- Creatine kinase (Ref: 143 U/L)
- Aldolase (Ref: 8.1 U/L)
- AST (Reference < 29 U/L)

**Aminotransferases**

- ALT (Reference < 29 U/L)
- AST (Reference < 29 U/L)

**DISCUSSION**

- Hypomyopathic DM typically manifests with features of muscle inflammatory activity, muscle biopsy, EMG, or imaging without signs or symptoms of weakness [8]
- 32% of DM patients have cancer (9% age-sex/matched controls)
- Commonly ovarian, breast, lung & gastric carcinoma, and NHL [9]
- Economic, geographical, and psychosocial factors may limit treatment options/availability [9]
- Amongst insured patients, the average time to DM diagnosis is 17 months [7]
- Evaluation may include laboratory testing, imaging (MRI, US, PET & HRCT), EMG, muscle biopsy, and skin biopsy
- Systemic corticosteroids are mainstay of initial therapy and conventional/biologic immunosuppressants are used for maintenance
- The uninsured status of our patient contributed to a significant delay in diagnosis and effective treatment (36 mos.), consistent with published CDC data [10]
- Rheumatology specialty care reduces disability & pain, and improves access to care in patients with rheumatic disease [11]
- Patients with Medicaid face unique challenges obtaining specialty care:
  - Rheumatologist shortage, lack of Medicaid acceptance, travel times, low physician reimbursement, excessive paperwork / administrative burden, longer wait times, referral patterns to low volume hospitals
- Lack of specialty care leads to higher utilization of EDs, acute care clinics, and free clinic services due to poorly controlled disease, more co-morbidities, lack of social support & increased disease complexity [11]
- Increasing access to rheumatology may be accomplished via traveling clinics, teleconsulting, direct care, use of midlevel providers, and funding of fellowship training programs [12, 13, 14, 15, 16]
- The following may improve access to specialty care in uninsured/underinsured patients:
  - Immediate access rheumatology clinics to shorten wait times [12]
  - Use of contrast enhanced ultrasound in place of the gold standard MRI to detect myositis [17]
- Specialty care embedded into community health centers (e.g. FQHC)

**REFERENCES**

2. Kaiser Family Foundation. Available at: http://dx.doi.org.uiwtx.idm.oclc.org/10.2217/ijr.11.78
INTRODUCTION

Angioedema is characterized by a localized, noninflammatory, non-pruritic, and well-demarcated, nonpitting swelling that occurs as large erythematosus areas in the skin and subcutaneous tissue. It involves areas such as the lips, tongue, face, glottis, oropharynx, periorbit, intestines, and genitals. There are two types of angioedema: mast cell-mediated and bradykinin-induced. Angiotensin Converting Enzyme inhibitor (ACEi) associated angioedema is considered a bradykinin induced angioedema and its prevalence ranges from 0.1% to 0.7% of those on ACE-inhibitors. While this adverse effect is often recognized when a patient is on an ACEi, repetitive episodes of angioedema after discontinuation are relatively uncommon.

CASE PRESENTATION

Initial Presentation: This is an 80 year old male with a history of hypertension and has been on Benazepril daily for over ten years who presented to the walk in clinic earlier in the week with complaints of mild lip swelling. He was instructed to stop the ACEi. Despite stopping Benazepril and internal improvement, he presented with facial and neck soft tissue edema and dysarthria. He was intubated for airway protection. A CT of the neck was performed showing pharyngeal thickening with soft tissue edema, no abscesses were seen. Solu-Medrol and Benadryl therapy were initiated along with one unit of fresh frozen plasma (FFP). The patient’s angioedema improved overnight and he was extubated the next morning. His symptoms completely resolved and the patient was discharged home.

ED Visit #2 – 11 days after stopping ACEi: The patient presented to the Emergency Department (ED) with lip and cheek swelling accompanied by dyspnea. Symptoms started in the morning when he woke up. He received high dose steroids, epinephrine, and two units of FFP in the ED and was admitted for further evaluation. Complement C4, C1q, and C1 esterase inhibitor levels were checked and were within normal limits. The patient’s symptoms completely resolved the next morning. He was prescribed an additional three days of prednisone and an EpiPen to use as needed.

ED Visit #3 – 48 days after stopping ACEi: The patient presented to ED with tongue and lip edema. The patient used an EpiPen at home which began to resolve his symptoms. He was kept for four hours in observation and discharged with a six day prescription of Prednisone.

ED Visit #4 – 54 days after stopping ACEi: The patient developed upper lip edema and presented to the ED. The swelling began on the left side and progressed to both. He denied dyspnea, dysarthria, or dysphagia. He was treated with Solu-Medrol.

The swelling rapidly improved and was discharged home from the ED. He was discharged with two EpiPens and a three day course of prednisone.

DISCUSSION

Angioedema is a rare adverse effect of ACE-inhibitors that can be life threatening. It has a higher incidence in women and a five-fold higher incidence among African Americans. Other risk factors include older than the age of 65, smoking, NSAID use, seasonal allergies, obesity, history of a transplant, and a history of ACEi associated cough. Two-thirds of cases occur within the first three months of starting an ACEi. However, like in this case, it can occur years after starting the medication. The longest case reported is after 23 years.1 Patients who experience angioedema from ACE-inhibitors should permanently discontinue all ACE-inhibitors and switch to a different antihypertensive medication. Small studies have shown that there is a low cross reactivity (<10%) for patients who switch to an Angiotensin Receptor Blocker (ARB). This cross reactivity suggests there may be undiscovered common pathways between ACE-inhibitors and ARBs that contribute to angioedema.4 In this case, the patient was switched to a calcium channel blocker. Despite stopping the ACEi, studies have shown up to 46% percent have recurrences of angioedema. Within this group, 88% of patients had recurrences in the first month the ACEi was discontinued. The recurrences can continue for six months or more.5

Diagnosis is made clinically based on the presentation of angioedema without itching or urticaria currently taking an ACEi. Important laboratory tests to obtain include CBC, CMP, CRP, ESR, and levels of the complement protein C4. Decreased levels of C4 should prompt further evaluation for hereditary or acquired C1 inhibitor deficiency. C4 and C1 inhibitor levels will be normal in ACEi induced angioedema.

ED Visit #5 – 72 days after stopping ACEi: One month later, the patient presented with asymmetric upper lip swelling that slowly progressed across both sides along with upper respiratory symptoms. He denied shortness of breath, stridor, or dysphagia. The patient was admitted and treated with two units of FFP and Solu-Medrol. His symptoms resolved and was discharged the next day.

The patient has had no further episodes since this last admission and is doing well.

REFERENCES

6. Fosshag, OMS-IV, Pacific Northwest University of Health Sciences, Yakima, WA
7. Sargsyan, MD, FACP, Benefis Health System, Great Falls, MT
What is the quality of online information on Irritable Bowel Syndrome (IBS)?
Nabeel M. Akhtar¹, MD; Louis J. Levine⁶, BS; Lori-Ann K. Glasgow⁵, BA; Elizabeth A. Thompson¹, PA-C; Sanam Razeghi¹, MD; Ann Ouyang¹, MD
1. Division of Gastroenterology and Hepatology, Department of Medicine, Penn State Milton S. Hershey Medical Center, Hershey, PA
2. Penn State College of Medicine, Hershey, PA

Introduction
Irritable Bowel Syndrome (IBS) patients frequently have difficulty understanding the disease process due to its complexity and lack of definitive testing. This may lead patients to rely on web-based information, but online resources have not been adequately studied for overall quality. The aim of this study was to identify popular IBS online resources and to assess their validity, readability, and content.

Methods
50 adult non-patients, non-healthcare workers were asked to search “irritable bowel syndrome” on an online search engine and to report the first page of results. Each website was ranked based on frequency of appearance, and the top 11 websites were individually scored for validity, readability, and content by three investigators.

• Validity was measured by the DISCERN and HON tools.
• Readability was measured via Flesch Reading Ease and Flesch-Kincaid Grade Level scales.
• Content was measured by a rubric, developed by three IBS specialists, consisting of terms pertaining to diagnosis, etiology, signs and symptoms, and treatment for IBS (maximum score 66).

The Atlantic and MedlinePlus were not scored because they were an editorial and a reference guide, respectively. For validity, four sites were certified by HON as indicated by a visible emblem.

No source scored a 5 on DISCERN primarily due to lack of details of support and information.

Results
Subject demographics: 25 were < 30 years old; 3 were 30-50; 22 were > 50. 7 resided outside the U.S. 7 had previously searched IBS.

Table 1. Results of validity (HON, DISCERN), readability (Flesch Reading Ease, Flesch-Kincaid Grade Level), and content (Content Score) for the most popular online searches for Irritable Bowel Syndrome

<table>
<thead>
<tr>
<th>Average Online Search Order</th>
<th>HON Certification (present/absent)</th>
<th>DISCERN Mean ± SD (max. 5)</th>
<th>Flesch Reading Ease (max. 121.23)</th>
<th>Flesch-Kincaid Grade Level</th>
<th>Content Score Mean ± SD (max. 66)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Wikipedia</td>
<td>Absent</td>
<td>3.3 ± 0.5</td>
<td>13.9</td>
<td>16.0</td>
<td>51 ± 3</td>
</tr>
<tr>
<td>2. Med Nurses</td>
<td>Present</td>
<td>3.7 ± 0.5</td>
<td>38.6</td>
<td>12.0</td>
<td>62 ± 1.2</td>
</tr>
<tr>
<td>3. WebMD</td>
<td>Present</td>
<td>3.3 ± 0.5</td>
<td>63.1</td>
<td>7.9</td>
<td>40.7 ± 0.6</td>
</tr>
<tr>
<td>4. Mayo Clinic</td>
<td>Present</td>
<td>3.7 ± 0.5</td>
<td>51.0</td>
<td>9.2</td>
<td>45 ± 3.5</td>
</tr>
<tr>
<td>5. MedlinePlus N/A</td>
<td>Absent</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>6. NIDDK</td>
<td>Absent</td>
<td>3.3 ± 0.5</td>
<td>63.0</td>
<td>7.7</td>
<td>51 ± 3</td>
</tr>
<tr>
<td>7. Emedicine Health</td>
<td>Present</td>
<td>4.1</td>
<td>41.7</td>
<td>11.3</td>
<td>48 ± 3.5</td>
</tr>
<tr>
<td>8. Dr. Axe</td>
<td>Absent</td>
<td>3.3 ± 0.2</td>
<td>45.1</td>
<td>12.3</td>
<td>34 ± 1.5</td>
</tr>
<tr>
<td>9. The Atlantic N/A</td>
<td>Absent</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>10. Health.com</td>
<td>Absent</td>
<td>3</td>
<td>40.8</td>
<td>13.0</td>
<td>25 ± 2.9</td>
</tr>
<tr>
<td>11. FASCRS</td>
<td>Absent</td>
<td>1.7 ± 0.2</td>
<td>47.2</td>
<td>10.6</td>
<td>40 ± 1.6</td>
</tr>
</tbody>
</table>

Table 2. Readability and Content scores of most popular online searches for Irritable Bowel Syndrome, sorted based on Readability

<table>
<thead>
<tr>
<th>Flesch Reading Ease Score</th>
<th>Source</th>
<th>Content (max 66)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;70 : Easy</td>
<td>NIDDK</td>
<td>51 ± 3</td>
</tr>
<tr>
<td>60-69: Standard</td>
<td>WebMD</td>
<td>40.7 ± 0.6</td>
</tr>
<tr>
<td>50-59: Fairly difficult</td>
<td>Mayo Clinic</td>
<td>45 ± 3.5</td>
</tr>
<tr>
<td>30-49: Difficult</td>
<td>FASCRS</td>
<td>40 ± 1.6</td>
</tr>
<tr>
<td>Dr. Axe</td>
<td>Emedicine Health</td>
<td>48 ± 3.5</td>
</tr>
<tr>
<td>Health.com</td>
<td>25 ± 2.9</td>
<td></td>
</tr>
<tr>
<td>Medicinenet</td>
<td>52 ± 1.2</td>
<td></td>
</tr>
</tbody>
</table>

• WebMD and NIDDK scored > 60 on the Flesch Reading Ease Score; Mayo Clinic scored 51 (Fairly difficult); All other sites scored as difficult or confusing < 49. Only WebMD and NIDDK were written at an appropriate grade level for patient resources (6-8th Grade).
• For content, 5 sites scored > 45; 3 sites scored 30-45; 1 site scored < 30.
• There was no correlation between readability and content (correlation coefficient 0.13).

Discussion
Most online resources that appear on a search for IBS have a poor readability score, particularly Wikipedia. The top site for both readability and content was NIDDK. These data suggest that healthcare providers should take an active role in directing patients to online resources on IBS.
Emergency Medicine Palliative Care Access (EMPallA): Preliminary Data from a Multi-Center Randomized Controlled Trial

Abigail M. Schmucker, BA1, Deborah J. Shim, BS2, Corita R. Grudzen, MD, MSHS3, Jeanne Cho, MPH3, Keith Goldfeld, DrPH4, The EMPallA Investigators

1Sidney Kimmel Medical College at Thomas Jefferson University, Philadelphia, PA; 2Augusta University Medical College of Georgia, Augusta, GA; 3New York University, Ronald O. Perelman Department of Emergency Medicine, New York, NY; 4New York University School of Medicine, Department of Population Health, New York, NY

Introduction

- The World Health Organization defines palliative care as "an approach that improves the quality of life of patients and their families facing the problems associated with life-threatening illness".
- Palliative care can be delivered alongside life-prolonging care and has been shown to improve symptoms and quality of end-of-life care across a range of illnesses.
- Palliative care also decreases costs by reducing unnecessary hospitalizations, diagnostic and treatment interventions, and avoidable intensive and emergency department (ED) care.

Objectives

- The aims of this study are to compare nurse-led telephonic case management to facilitated, outpatient specialty palliative care for older adults with serious, life-limiting illnesses: 1. Quality of life 2. Healthcare utilization (ED revisits, hospital admissions, hospice use) 3. Patient loneliness and symptom burden 4. Caregiver strain and quality of life
- This poster reports preliminary recruitment progress, demographic information, and quality of life data for the study cohort

Methods

- Pragmatic, two-arm, multi-center, randomized controlled trial
- 1350 older adults aged 50+ years, and 675 of their informal caregivers from the EDs of 9 diverse sites will be enrolled over 3 years
- Patients randomized to either arm receive palliative care that addresses their needs within the eight domains of palliative care
- Both arms have standardized assessment, referrals, and criteria for follow-up visits or phone calls

Results

- From April 16, 2018 to December 31, 2018, 354 patients were eligible
- Patients randomized to either arm receive palliative care that addresses their needs within the eight domains of palliative care
- Both arms have standardized assessment, referrals, and criteria for follow-up visits or phone calls

Discussion

- The EMPallA study is recruiting a gender-balanced, racially-diverse cohort of older adults with a range of serious illnesses
- Average baseline FACT-G T-scores are below the general population means of 50 by more than a clinically-meaningful difference (5 points), suggesting this cohort of older adults has the potential to benefit from palliative care
- When trial enrollment and 6-month follow up is complete, comparative impact of palliative care interventions on quality of life can be assessed

Study Registration and Disclaimer

- The EMPallA study is registered at ClinicalTrials.gov (NCT03836694)
- This work was (partially) supported through a Patient-Centered Outcomes Research Institute (PCORI) Award (PLC-1606-39306) and a Medical Student Training in Aging Research (MSTAR) grant from the National Institute on Aging (Abigail Schmucker)
- All authors listed on this poster consent to its publication

References


4. Block randomization at patient level, ratio of 1:1, stratified by ED site and disease

5. FACT-G raw scores were rescaled into T-scores based on a general US population sample, standardized with mean of 50 and standard deviation of 10

6. FACT-G subscale and total T-scores means and standard deviations were reported; total T-scores were summarized in a histogram

7. Study participants were a racially-diverse, predominantly white, non-Hispanic (90%) cohort of older adults with a range of serious illnesses suggesting this cohort of older adults has the potential to benefit from palliative care

8. When trial enrollment and 6-month follow up is complete, comparative impact of palliative care interventions on quality of life can be assessed

9. All authors listed on this poster consent to its publication

10. The EMPallA study is registered at ClinicalTrials.gov (NCT03836694)

Table 1: Patient Demographics

<table>
<thead>
<tr>
<th>Age (years), mean (SD)</th>
<th>68 (10)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender (female), N (%)</td>
<td>99 (56%)</td>
</tr>
<tr>
<td>Ethnicity (Hispanic), N (%)</td>
<td>20 (11%)</td>
</tr>
<tr>
<td>Race (white), N (%)</td>
<td>91 (51%)</td>
</tr>
<tr>
<td>Born in United States, N (%)</td>
<td>155 (88%)</td>
</tr>
</tbody>
</table>

Table 2: Patient Inclusions

<table>
<thead>
<tr>
<th>Advanced Cancer</th>
<th>79 (45%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart Failure</td>
<td>44 (25%)</td>
</tr>
<tr>
<td>End Stage Renal Disease</td>
<td>29 (16%)</td>
</tr>
<tr>
<td>Chronic Obstructive Pulmonary Disease</td>
<td>47 (27%)</td>
</tr>
</tbody>
</table>

Table 3: Baseline Quality of Life Scores

| FACT-G Quality of Life T-Score, mean (SD)* | Physical Well-Being | 37 (11) |
|--------------------------------------------|---------------------|
|                                            | Social/Family Well-Being | 51 (10) |
|                                            | Emotional Well-Being | 41 (13) |
|                                            | Functional Well-Being | 42 (10) |
|                                            | Total Score | 41 (11) |

*Relative to US general population, standardized with mean of 50 and SD of 10
INTRODUCTION
Obstructive Sleep Apnea Hypopnea Syndrome (OSAHS) is characterized by the limitation of the passage of air by functional and/or anatomical alterations, which compromise the quality of life, becoming a public health problem that generates high costs in health due to its association with cardiovascular risk (hypertension, diabetes mellitus and obesity). Different tools have been developed to detect the risk of presenting OSAHS (Berlin, STOP-BANG, Epworth Sleepiness Scale). Its diagnosis must be made by polysomnogram (PSG), considered the gold standard.

RESULTS
A statistically significant association was found between the lower probability of having AHI ≥15 and the perception of having abrupt reduction or loss of muscle strength (OR 0.35; P 0.29; CI 0.13-0.94) and lucid dreams while awake (OR 0.39; P 0.036; CI 0.16-0.94). We observed a higher probability of having AHI≥15 in patients with mento-sternal distance >15cm (71.18; OR 2.47; P 0.09; CI 1.25-4.9), fall asleep while sitting and inactive in a place (61.83%; OR 1.62; P 0.003; CI 1.18-2.23), fall asleep as passengers in a car during one hour of journey (59.83%; OR 1.49; P 0.004; CI 1.13 -1.96), fall asleep while talking to someone (65.15%; OR 1.87; P 0.007; CI 1.19-2.94). EPWORTH score >14 (71.18%; OR 2.47; P 0.023; CI 1.13-5.39), sleep apnea (74.09%; OR 2.86; P 0.002; CI 1.45-5.63), daytime fatigue (73.26%; OR 2.74; P 0.003; CI 1.39-5.38), STOP BANG ≥5 (68.65; OR 2.19; P 0.019; IC 1.13-4.26), REM sleep ≥46 minutes (25.92%; OR 0.35; P 0.003; CI 0.18-0.69) and awake saturation ≤91% (67.21%; OR 1.05; P 0.043; IC 1.02-4.1).

CONCLUSION
A lower relationship with other sleep disturbances was observed in patients with OSAHS with AHI≥15, who also showed higher drowsiness, daytime fatigue and presence of apneas with a better performance on the STOP-BANG scale in the diagnosis. A shorter duration of REM sleep and lower saturation than 91% were seen to be remarkable characteristics that should be taken into account for the severity of OSAHS.
Background

In 2018, a series of laws aimed toward curtailing the opioid crisis started to be implemented in the State of Michigan. They are summarized as follows:

Prescribers (such as physicians, physician assistants, nurse practitioners, etc.) must have a bona fide relationship with their patients prior to prescribing opioids for chronic pain, barring certain exceptions.

Prescribers must also receive informed consent from their patients, both minors and adults, prior to prescribing opioids. Prescribers will be required to document informed consent on a form created by the Michigan Department of Human and Health Services.

Prescribers must query the Michigan Automated Prescription System (MAPS) before prescribing a controlled substance, which include opioids, to their patients.

Prescribers are no longer allowed to prescribe greater than a 7-day supply for acute pain.

Prescribers who treat a patient for an opioid overdose are mandated to provide the patient with information on addiction treatment.

Methods

After review from the local Institutional Review Board, an anonymous 11-question survey was distributed electronically via an online software program (RedCap) to prescribers in the State of Michigan through medical society memberships and affiliated healthcare systems. In addition to a series of questions regarding their knowledge, preparation, and perceptions of the new statewide opioid prescribing laws, prescribers were asked if opioid prescribing was part of their regular medical practice. 173 prescribers completed this survey between July 9 and August 15, 2018.

Results

Survey highlights are presented below:

In your view, what level of impact has the opioid crisis had on your practice (or the healthcare setting you work) and/or patient population?

![Survey Results](image)

How much of an impact do you believe the new statewide opioid prescribing laws will have on reducing opioid prescribing?

![Survey Results](image)

Further results:

- While 84.8% indicated being at least somewhat prepared to comply with the opioid prescribing laws, only 16.4% reported feeling extremely prepared.
- Despite 58.1% of prescribers reporting not receiving any training in their workplace regarding the new laws, 94.2% believed that the new laws will reduce opioid prescribing.
- 70.9% reported that the consequences for not complying with the prescribing laws were not appropriate. Results from an optional commentary section in the survey indicate the respondents felt the consequences are overly punitive.

Prescriber Commentary

At the conclusion of the survey, prescribers were given an opportunity to comment on the new prescribing laws. Here are a few samples:

- “The consequences of failure to comply with these laws (however unintentional) adds to the fear, frustration, administrative burden, cost of care, and clinician burnout that have plagued our profession. A more collaborative and educational approach to these laws would have been appreciated and welcomed.”

- “Makes it more difficult to actually prescribe pain meds to people who are in pain and really need it like cancer patients.”

- “The law adds a great deal of bureaucracy and red tape to providing a prescription for a patient. In some ways it is good to make it more difficult to prescribe a narcotic to decrease the amount in the system, in other ways it makes all patients negatively affected because of how much work is required to arrange a single prescription that a lot of offices have to spread staff thin to stay in compliance and to continue to care for regularly scheduled visits.”

Conclusions

The ability of the new statewide opioid prescribing laws to reduce opioid prescriptions, and therefore opioid addiction and overdoses, relies on prescribers’ awareness of the laws and the time afforded for implementation.

Based on the results and subjective comments obtained from this survey, engaging prescribers prior to mandated requirements may lead to more satisfactory outcomes. Ultimately, the emphasis of the new laws is to improve patient outcomes through effective coordination and management when and where controlled substances are prescribed.

References


INTRODUCTION

Hyperbilirubinemia is defined as elevated bilirubin in the blood.

Two types:
• Unconjugated - Bilirubin overproduction, impaired uptake of bilirubin, defects in conjugation
• Conjugated – intrinsic liver dysfunction

We describe a case of mixed hyperbilirubinemia likely due to a rare genetic cholestatic liver disorder: Benign Recurrent Intrahepatic Cholestasis (BRIC)

CASE PRESENTATION

24 year old Korean man presented with two weeks of jaundice and pruritus. He endorsed multiple similar self-resolving episodes of jaundice and pruritus starting in childhood.

Family history of “liver disease” (mother)

Physical examination was notable for:
• Scleral icterus
• Jaundiced skin
• Diffuse excoriations

Notable Lab Values On Presentation

<table>
<thead>
<tr>
<th>Test</th>
<th>Patient’s Value</th>
<th>Normal Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Bilirubin (mg/dL)</td>
<td>16.7</td>
<td>0.3-1.3</td>
</tr>
<tr>
<td>Direct Bilirubin (mg/dL)</td>
<td>9.1</td>
<td>0.0-0.2</td>
</tr>
<tr>
<td>Alkaline Phosphatase (U/L)</td>
<td>133</td>
<td>35-115</td>
</tr>
<tr>
<td>Bile Acid (µmol/L)</td>
<td>100</td>
<td>0-60</td>
</tr>
<tr>
<td>Gamma Glutamyl Transferase (U/L)</td>
<td>52</td>
<td>0-65</td>
</tr>
</tbody>
</table>

H&E stained slides of liver core biopsy.
• 100x: A low power view shows multiple regions of hepatic parenchyma with cholestasis (boxes). Masson’s trichrome stain (inset, 400x) shows an expected degree of perivascular collagen deposition (stained blue). However, perisinusoidal/pericellular fibrosis is also evident, which may represent underlying, chronic injury.

H&E stained slides of liver core biopsy.
• 400x: The cholestasis is noted to involve perivenular parenchyma (Zone 3) and is seen in hepatocytes (arrow) as well as canaliculi (arrowhead). The cholestasis appears bland with minimal hepatocyte injury and no significant lobular inflammation or steatosis.

HOSPITAL COURSE

During his hospitalization, the patient underwent the following workup:
• Abdominal Ultrasound: 0.9 cm hemangioma, otherwise normal
• AMA negative, ANA negative, Myeloperoxidase antibody negative
• MRCP: no biliary abnormality
• Transjugular liver biopsy was done

Symptom management: He was started on ursodiol and cholestyramine with relief of his pruritus.

Following discharge: The patient was discharged with hepatology follow-up pending biopsy and genetic testing results showing:
• Biopsy showed perivenular bland cholestasis and perisinusoidal/pericellular fibrosis.
• Genetic testing was notable for genetic variants of uncertain significance (test did not cover gene mapping for BRIC).

DISCUSSION

Alternative differential diagnoses:
• Hemolytic (normal LDH, lactoglobulin, and reticulocyte count)
• Infectious etiology (normal transaminases, negative hepatitis panel)
• Autoimmune pathology, like PSC and PBC (negative AMA/ANA, MPO, MRCP)

BRIC is an autosomal recessive disorder characterized by recurrent episodes of cholestasis. It is clinically diagnosed by:
1. At least two episodes of jaundice associated with severe pruritus and asymptomatic intervals lasting for several months/ years
2. Absence of any known etiology for cholestasis
3. Signs of cholestasis indicated by laboratory tests contrasted with normal MRCP/ERCP
4. Bile plugs within the ducts as seen by liver histology

Genetic mutations:
• BRIC1: ATP8B1 on chromosome 18q21
• BRIC2: ABCB11 on chromosome 2q24

Diagnosis of BRIC and other inherited liver diseases is important to:
• Guide management of symptoms
• Decrease the rate of cirrhosis development
• Reduce possibility of malignancy
• Aid in family planning

BRIC Management:
• No known cure
• Symptomatic management with ursodiol, cholestyramine, rifampin

CONCLUSION

In addition to more common causes of hyperbilirubinemia, genetic causes of hyperbilirubinemia should be considered in a patient with recurrent symptoms of jaundice and pruritus.

REFERENCES

Mothball Ingestion as a Manifestation of Pica, leading to Paradichlorobenzene CNS Toxicity

Joon Yau Leong¹, Margarita Gianniosis¹, Ann Lee¹, Elwin Tham¹, Saman Zafar², Yan Zhang²

¹Sidney Kimmel Medical College, Thomas Jefferson University; ²Department of Neurology, Albert Einstein Medical Center Philadelphia

INTRODUCTION

- Pica, a psychiatric disorder presenting with ingestion of non-nutritious substances, is commonly associated with iron deficiency anemia.
- A high index of suspicion for unusual toxin exposure aids in the diagnosis of pica patients presenting with unexplained neurodegenerative features.
- Paradichlorobenzene (PDCB) has largely replaced naphthalene as the primary component in mothballs.
- **Objective:** We present a case of PDCB CNS toxicity predominantly affecting the middle cerebellar peduncles (MCP) on MRI and provide a list of differential diagnoses with similar MRI features.

CASE PRESENTATION

1) History Taking
   - 47 y/o F presents with recurrent falls and declining cognition over the past year.
   - Prior to this, she was fully independent with her ADLs but is now wheelchair-bound due to unsteady gait and frequent falls.
   - PMH: menorrhagia, iron deficiency anemia
   - No PSH, FH, Meds/Allergies
2) Physical Examination
   - General: Noted to have a unidentified chemical odor in patient room
   - CNS: Bilateral dysmetria on finger-nose testing, ataxic gait, symmetrical 4/5 weakness on all 4 extremities but normal sensation
   - Psych: Childlike affect, emotional lability, poor short term recall
3) Laboratory Findings
   - CBC – Hemoglobin low at 7.8 g/dL
   - Iron studies (serum ferritin, TIBC and iron saturation) – Confirmed the diagnosis of iron deficiency anemia

IMAGING STUDIES

These T2 FLAIR images show amorphous, relatively symmetric ill-defined regions of T2 hyperintense signal within the supratentorial white matter, corpus callosum and bilateral middle cerebellar peduncles (MCP) without mass effect. There was no restricted diffusion or enhancement on other sequences.

Figure 1: MRI brain with and without contrast (arrow pointing to T2 hyperintense signals within the middle cerebellar peduncles)

FURTHER INVESTIGATIONS

1) Collateral history
   - Patient’s daughter revealed that the patient has been ingesting mothballs for the past 5 years
   - Over the past year, quantity of mothballs ingested have increased from 1/day to 6/day
   - Patient states that she is “addicted to the smell” and “it helps relieve stress”
   - She denies feelings of euphoria or any visual/auditory hallucinations
2) Other investigations
   - Genetic Fragile X PCR – negative
   - Serum PDCB levels – elevated at 15 mcg/mL

DISCUSSION / DIFFERENTIAL DIAGNOSES

1) **Spinocerebellar Ataxia (SCA)**
   - Patients brain imaging lacks findings of atrophy on the pons and cerebellum
2) **Multiple system atrophy – cerebellar type (MSA-C)**
   - Patient lacks the classical features of autonomic dysfunction and parkinsonism
3) **Fragile X-associated tremor/ataxia syndrome (FXTAS)**
   - Although more common in men, female carriers often present with infertility or premature ovarian failure
4) **Bilateral anterior inferior cerebellar artery infarction**
   - Patient lacks risk factors nor did she exhibit signs and symptoms of a stroke
5) **Hypoglycemic coma**
   - Excluded based on clinical presentation and labs
6) **Toxin (PDCB) ingestion**
   - Collateral history of pica and mothball ingestion, presence of chemical odor in patient room
   - Elevated serum PDCB levels, bilateral and symmetrical changes on brain imaging

CONCLUSIONS

- Toxic-metabolic insults to the CNS generally leads to symmetrical changes on MRI, as opposed to other etiologies.
- Iron deficiency anemia leads to pica, which can lead to toxin ingestion. Seeking out a history of unusual ingestions in a patient with pica can be immensely helpful in diagnosis.
- Mothball ingestion may be a manifestation of pica, and presents with neurological findings such as cognitive deficits and ataxia.
- Previous case reports of PDCB neurotoxicity have reported diffuse or symmetrical changes on neuroimaging.
- To our knowledge, there has not been any reported cases with PDCB toxicity predominantly involving the MCPs.

REFERENCES

Minnesota Physicians’ Familiarity and Use of Provider Order for Life Sustaining Treatments (POLST)

Karly Boll1; Paul Blake1; Beret Fitzgerald1; Bruce Gregoire1; Jack Inglis1; Dylan McCreary1; Lisa Skarbakka1; Becca Branum JD2

1: University of Minnesota Medical School  2: Minnesota Medical Association

September 2018

Introduction

What is the POLST?

Provider Orders for Life Sustaining Treatment (POLST) is a medical order for patients who are likely to die from a terminal condition within a year. It allows emergency medical providers to follow the patients’ wishes in the event of an emergency medical event.

Why is POLST important?

As the Minnesota populace ages, increasingly patients will live with chronic conditions. Many will experience emergency situations where they can not express their care preferences. POLST offers a standardized, accessible, and transportable form to advocate for patients’ desires.

What were our research goals?

Describe the familiarity and use of POLST by Minnesota physicians, and the patterns of use by specialty and region.

Provide insight to the MMA about physicians’ use of POLST to increase use of the POLST and decrease misunderstandings about POLST.

Methods

This project utilized a cross-sectional survey of Minnesota physicians on the MMA’s email list. The online survey was a 16 question multiple choice survey which employed skip logic. 6,526 physicians were contacted using a repeated email request. 656 physicians completed the survey.

Participants

Among the participants there were 157 physicians specialized in family medicine (22%), 137 in internal (20%), 91 in surgery (13%), 49 in emergency (7%), 42 in pediatrics (6%), 19 in hospice and palliative care (3%), and 203 in other specialties (29%).

363 physicians worked in the Twin Cities metropolitan and 293 worked in the surrounding regions of Minnesota.

The two most common workplace settings where in outpatient settings (205; 31%) and a combination of inpatient and outpatients settings (190; 29%).

Rates of familiarity, use of, and signing of POLST forms

Among the physicians who had used the POLST, many signed the form themselves, showing active participation in its use.

A significantly higher proportion of family physicians were actively involved in using POLST forms

Perceived problems with POLST forms

We asked physicians about benefits and problems they perceived with the current POLST form. Problems listed were (1) confusing or difficult to complete, (2) inadequate space on the form to document wishes, (3) inadequate reimbursement for services related to the POLST, (4) patients or family are unaware of the form, (5) patients or family don’t understand the purpose of the form, (6) time consuming to complete. The problems were grouped by form, reimbursement, or patient/family related.

Most physicians (74%) and family physicians (74%) endorsed all benefits.

Discussion

Findings

We found that family physicians were more familiar with POLST and more likely to use and sign POLST forms than other physicians.

Physicians responded that the most significant barrier to using the POLST was lack of knowledge and understanding of the POLST among patients and families.

The most frequent situation prompting a POLST conversation was initiation by the patient or perceived need by the physician.

Implications

We found that specialists were less familiar with the POLST than primary care physicians, suggesting that patients receiving care from these specialists may not have access to the POLST. We determined that statewide patient-focused education about POLST would provide the greatest benefit to physicians who use the POLST in their practice for end-of-life discussions.

Acknowledgements

Thank you to the MMA, the MMA’s POLST Steering Committee, the University of Minnesota Medical School, Dr. Katie Freeman, and Dr. David Satin for their support and guidance on this project.
Timeline

**Jan 2017**
- **Diagnosis:** Female presents with uncharacteristic weight loss of 40lbs, memory problems, inability to focus at school.
- **Evaluation:** Brain comets at MRI; FDG PET reveals hypometabolism in temporal lobes and basal ganglia.
- **Psychiatric:** Methylphenidate with bipolar disorder, onset with depression.
- **Diagnostic:** Quetiapine, topiramate, lithium stopped; lorazepam taper.

**Feb 2017**
- **Psychiatric:** Depression with elements of bipolar disorder.
- **Gyn-Onc:** No evidence of neoplasm.
- **CT abd/pelvis, chest:** Negative.
- **LP:** CSF for Mayo encephalopathy panel, Lyme.
- **Serum/Labs:** ANA, dsDNA, ANCA, ENA, SSA/SSB, TPO, Anti-GBM.
- **30 min EEG:** R hemispheric dysfunction with diffuse encephalopathy.
- **Brain MRI:** MRI brain March 2018; T2 FLAIR abnormalities in the medial temporal lobes and basal ganglia.

**Mar 2018**
- **Brain MRI:** Abnormal signal and enhancement in the basal ganglia aspect of the brain compared to prior scans.
- **CFF: 11 bilateral bands, left > right.
- **MRI:** Symmetry abnormal, hemispheric lesions noted, requiring f-b scan.
- **Diagnosis:** Unspecific diagnosis, further study needed.
- **Psychiatric:** Advanced to consider psychiatric test other suicide attempt.
- **Diagnosis:** Manic depression.
- **Neuromuscular:** Valproate, lamotrigine, and clonazepam.
- **Neuroimmunologist:** Permits institutional recommendation.
- **Hospital:** Transfer to outside psychiatric unit.
- **Diagnosis:** Depression with elements of bipolar disorder.

**May 2018**
- **Neuroimmunologist:** Admits patient to University Hospital to expedite hospital course.
- **Hospital Course:**
  - **Neuromuscular:** Admits patient to University Hospital to expedite hospital course.
  - **Diagnosis:** Seizure activity, absence of seizures.
  - **Brain MRI:** 35% hippocampal lesions.
  - **EEG:** Infrequent epileptic activity, but frequent slow, disorganized activity.
  - **CSF:** Lymphocytic pleocytosis, oligoclonal banding
- **Labs & Imaging:**
  - **Seizures:** May occur at any time during the disease.
  - **Speech:** Dysfunction, dyskinesias, memory deficits, autonomic instability, decrease in level of consciousness.

**July 2018**
- **Hospital:** Patients admitted to University Hospital for evaluation with trial of IV steroids vs IVIG for diagnosis of NMDA encephalitis.
- **Hospital Course:**
  - **Neuromuscular:** Admits patient to University Hospital to expedite hospital course.
  - **Diagnosis:** Seizure activity, absence of seizures.
  - **Brain MRI:** 35% hippocampal lesions.
  - **EEG:** Infrequent epileptic activity, but frequent slow, disorganized activity.
  - **CSF:** Lymphocytic pleocytosis, oligoclonal banding

**Outlook:**
- **Diagnostic:** Manic depression.
- **Neuromuscular:** Valproate, lamotrigine, and clonazepam.
- **Neuroimmunologist:** Permits institutional recommendation.
- **Psychiatric:** Advanced to consider psychiatric test other suicide attempt.
- **Diagnosis:** Manic depression.
- **Neuromuscular:** Valproate, lamotrigine, and clonazepam.
- **Neuroimmunologist:** Permits institutional recommendation.

**Conclusion**
This case illustrates the arduous journey of a young girl whose diagnosis and treatment were delayed for over a year, leading to permanent scarring in her brain and emotional trauma to her family.

This is a difficult diagnosis to make because anti-NMDA receptor encephalitis can masquerade as a psychiatric illness, is relatively new, and has no textbook definition for the clinical diagnosis.

Physicians should avoid “anchoring,” and watch out for “availability bias.” After this patient’s case was labeled as a psychiatric case during her initial hospital presentation, there was no consideration for neuropsychiatric or neurological causes for over a year. Although this disorder is rare, it presented with red flags within the first six months. Recognition and suspicion of “zebras” by providers is critical in diagnosing appropriate diagnostic procedures and initiating therapy for these patients.

References

**Hospital Course**
- **Neuromuscular:** Admits patient to University Hospital for expedite hospital course with trial of IV steroids vs IVIG for diagnosis of encephalitis and encephalomyelitis.
- **Brain MRI:** 30 min EEG: B hemispheric dysfunction with diffuse encephalopathy.
- **Serum/Labs:** ANA, dsDNA, ANCA, ENA, SSA/SSB, TPO, Anti-GBM.
- **Lipid- and CSF:** For Mayo encephalopathy panel, Lyme.
- **EEG:** Electrocardiogram, positive oligoclonal bands, elevated IgG.
- **CSF:** Lymphocytic pleocytosis, oligoclonal banding

**Proposed Diagnostic Criteria [4]**
1. **Rapid onset** (less than 3 months) of at least four of the six following major groups of symptoms:
   - Abnormal (psychiatric) behavior or cognitive dysfunction
   - Speech dysfunction (pressured speech, verbal reduction, mutism)
   - Abnormal (psychiatric) behavior or cognitive dysfunction
   - Seizures
   - Movement disorder, dyskinesias, or rigidity/abnormal postures
   - Decreased level of consciousness

2. At least one of the following laboratory study results:
   - Serum/Labs: Antibodies against NR1 or NR2 subunits of the NMDA receptor
   - Serum/Labs: 38% of patients have underlying neoplasm
   - Serum/Labs: 94% ovarian teratomas

3. Reasonable exclusion of other disorders
   - Seizures may occur at any time during the disease
   - Speech dysfunction, dyskinesias, memory deficits, autonomic instability, decreased in level of consciousness
   - Malignancy or autoimmune disease

**Disease Information**
**Anti-NMDA Receptor Encephalitis**
- **Discorded in 2007**
- **Mothers commonly present NR1 or NR2 subunits of the NMDA receptor**
- **38% of patients have underlying neoplasm**
- **94% ovarian teratomas**
- **Presentation commonly presents ages 12 to 45 years**
- **40% of all cases are in children 18 years**
- **Female predominance 4:1**
- **38% of patients have underlying neoplasm**
- **Antibodies against NR1 or NR2 subunits of the NMDA receptor**
- **Discorded in 2007**
- **Anti-NMDA Receptor Encephalitis**
- **American Journal of Neuroradiology (4), 391–404.**
- **http://doi.org/10.1002/ana.21050**
- **Lancet Neurology**
- **http://doi.org/10.1016/S1474-4422(12)70130-1**
- **http://doi.org/10.1002/ana.23576**
- **Lancet Neurology**
- **http://doi.org/10.1016/S1474-4422(15)00419-7**
- **Annals of Neurology**
- **http://doi.org/10.1002/ana.21756**
- **Annals of Neurology**
- **http://doi.org/10.1002/ana.21756**
- **American Journal of Neuroradiology**
- **http://doi.org/10.3174/ajnr.A5593**
Human adipose tissue was obtained from two patients undergoing panniculectomies and Human Adipose-Derived Mesenchymal Stem Cells (hADSCs) were purchased from American Type Culture Collection (ATCC). Cells from these sources were induced into a neural lineage in complete growth media with four different concentrations of IBMX and dbcAMP was obtained, a 12-day time course was completed to assess for differences in the expression levels of IBMX/1.0mM dbcAMP, while ATCC hADSCs show little fluorescence at 3 days and 6 days post-induction. Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Patient-derived and ATCC cells showed expression of S100B by 9 hours with a decreased intensity of S100B starting at 6 days. Additionally, patient-derived cells induced with IBMX and dbcAMP were slightly positive for GFAP, but not ATCC cells, starting at 3 days and 6 days (Figure 2).

Conclusions

With the differences in expression of the neural cell markers S100B and GFAP observed between patient-derived and commercially available hADSCs, this data suggests that patient-derived hADSCs are a better source to study the differentiation of stem cells into neural-like cells compared to ATCC cells. Future work will include additional patient-derived hADSCs to confirm the results of this study.

References

Incidence and Outcome of Bacteremia during Acute Graft-Versus-Host Disease Involving the Gastrointestinal Tract following Hematopoietic Stem Cell Transplant

Christian Lindner¹, Jenna Petersen MD², Morgan Hakki MD¹

¹ Oregon Health & Science University

Background

- Acute graft-versus-host disease (aGVHD) involving the gastrointestinal (GI) tract is a complication of allologeneic hematopoietic stem cell transplant (HSCT).
- The risk of bacteremic events after onset of aGVHD may be increased due to both mucosal disruption of the GI tract and augmented immune suppression required for aGVHD treatment.
- Data pertaining to the incidence and outcome of bacteremic events in this specific setting is not well reported.

Research Objectives

- To describe the incidence of bacteremic events, causative organisms, and infection attributable mortality in HSCT recipients with GI tract aGVHD.

Methods

Design:
- Retrospective study of adult allogeneic HSCT recipients with grades 2-4 GI aGVHD occurring between 2009-2017 at Oregon Health and Science University (OHSU).
- Patient characteristics including grade and date of onset of GI aGVHD, age, sex, underlying diagnosis, date of death (if applicable), and source of cells used for HSCT were obtained from OHSU’s HSCT registry.
- Bacteremic events and the causative organisms were found by review of the electronic medical record (EMR).
- Infection-attributable mortality was defined as death within 7 days of a bacteremic event.

Inclusion Criteria:
- aGVHD of the GI tract Grade 2-4
- Bacteremia occurring within 30 days onset of aGVHD
- Positive blood cultures included only if performed due to clinical suspicion of infection

Exclusion Criteria:
- Bacteremia occurring >30 days of aGVHD onset
- Positive blood culture performed as surveillance blood culture

Statistical methods:
- p-values obtained using fishers exact t-test
- Kaplan-meier survival curve created using SPSS, overall comparison using Log Rank test

Results

Table 1: Patient characteristics

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Patients</td>
<td>159</td>
</tr>
<tr>
<td>GVHD Grade</td>
<td></td>
</tr>
<tr>
<td>Grade 2</td>
<td>56 (35.2)</td>
</tr>
<tr>
<td>Grade 3</td>
<td>45 (28.3)</td>
</tr>
<tr>
<td>Grade 4</td>
<td>58 (36.5)</td>
</tr>
<tr>
<td>Age</td>
<td></td>
</tr>
<tr>
<td>Median, Range</td>
<td>58.5, 60 (21-81)</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>102 (64.4)</td>
</tr>
<tr>
<td>Female</td>
<td>57 (35.6)</td>
</tr>
<tr>
<td>Diagnosis before HSCT</td>
<td></td>
</tr>
<tr>
<td>AML</td>
<td>62 (39)</td>
</tr>
<tr>
<td>MDS</td>
<td>33 (20.8)</td>
</tr>
<tr>
<td>Other</td>
<td>65 (42.0)</td>
</tr>
<tr>
<td>Source of stem cells</td>
<td></td>
</tr>
<tr>
<td>Peripheral stem cells</td>
<td>151 (95)</td>
</tr>
<tr>
<td>Bone marrow</td>
<td>5 (3.1)</td>
</tr>
<tr>
<td>Cord Blood</td>
<td>3 (1.9)</td>
</tr>
</tbody>
</table>

Table 2: Microbiology of bacteremic events

<table>
<thead>
<tr>
<th>Cultured organism</th>
<th>N</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gram positive: (n=23)</td>
<td></td>
</tr>
<tr>
<td>Methicillin-susceptible Staphylococcus aureus</td>
<td>7</td>
</tr>
<tr>
<td>Methicillin-resistant Staphylococcus aureus</td>
<td>3</td>
</tr>
<tr>
<td>Enterococcus faecium</td>
<td>3</td>
</tr>
<tr>
<td>Coagulase-Negative Staphylococci</td>
<td>2</td>
</tr>
<tr>
<td>Streptococcus species</td>
<td>2</td>
</tr>
<tr>
<td>Enterococcus faecalis</td>
<td>2</td>
</tr>
<tr>
<td>Streptococcus bovis</td>
<td>1</td>
</tr>
<tr>
<td>Anaerobic gram-positive rod</td>
<td>1</td>
</tr>
<tr>
<td>Vaocmycin-resistant Enterococci</td>
<td>1</td>
</tr>
<tr>
<td>Group B streptococal</td>
<td>1</td>
</tr>
<tr>
<td>Gram negative: (n=21)</td>
<td></td>
</tr>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>7</td>
</tr>
<tr>
<td>Escherichia coli</td>
<td>5</td>
</tr>
<tr>
<td>Klebsiella pneumonia</td>
<td>4</td>
</tr>
<tr>
<td>Bacteroides fragilis</td>
<td>3</td>
</tr>
<tr>
<td>Neisseria species</td>
<td>1</td>
</tr>
<tr>
<td>Fusobacterium nucleatum</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 3: Infection attributable mortality

<table>
<thead>
<tr>
<th>GVHD Grade</th>
<th>Grade 2</th>
<th>Grade 3</th>
<th>Grade 4</th>
<th>Overall</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bacteremic (N)</td>
<td>6</td>
<td>7</td>
<td>32</td>
<td>45</td>
</tr>
<tr>
<td>Death within 7 Days (N)</td>
<td>2</td>
<td>0</td>
<td>6</td>
<td>8</td>
</tr>
<tr>
<td>% dead within 7 days</td>
<td>25</td>
<td>0</td>
<td>15.8</td>
<td>15</td>
</tr>
</tbody>
</table>

Figure 1: Bacteremic events by grade GVHD

Discussion

- Bacteremic events are common in HSCT recipients with aGVHD involving the gastrointestinal tract.
- Patients with grade 4 GVHD appear to be at higher risk for bacteremia compared to patient with grade 2 or 3 GVHD.
- Both gram positive and negative organisms contribute to bacteremia in GVHD about equally.
- Bacteremic events result in significant infection-attributable mortality without a significant difference between grades.
- Bacteremic patients have a significantly reduced survival at 60 days compared to non-bacteremic patients.
The use of Microbial Flow Cytometry to analyze the Intestinal Microbiota

Stauffer P1, Klick M1, Martin T1, Diamond S2, Rosenbaum JT1,3,4, Asquith M1

1) Division of Arthritis and Rheumatic Disease, Dept. of Medicine, OHSU, Portland OR; 2) Division of Gastroenterology and Hepatology, OHSU; 3) Casey Eye Institute and Dept. of Ophthalmology, OHSU; 4) Legacy Devers Eye Institute, Portland, OR.

INTRODUCTION

The role of the intestinal microbiota in the pathogenesis of autoimmune disorders such as Ankylosing Spondylitis (AS) has been widely studied through methods that analyze gross microbial community dysbiosis between healthy controls. Current methods however may fail to detect moderate changes to the intestinal microbiota, for instance in individuals that harbor disease susceptibility genes but have yet to manifest with clinical symptoms.

In this study we used flow cytometry and a novel analysis workflow, Quantitative Microbial Profiling (QMP), in a cohort of patients with AS to improve upon standard microbiome analysis techniques that rely on relative microbial profiling (RMP). Furthermore we interrogated the composition and host immune response of the gut microbiome using IgA-SEQ, a method in which we flow sort and 16S rRNA sequence IgA coated bacteria. The latter method was performed in otherwise healthy individuals which harbor the major AS susceptibility gene HLA-B27.

METHODS

Facies of healthy controls (n = 23) and AS patients (n = 24) were subjected to QMP through the combination of microbial flow cytometry and 16s rRNA sequencing. To uncover mechanisms through which major genetic risk factor HLA-B27 might contribute to dysbiosis irrespective of disease state we subsequently compared the total frequency of IgA coated bacteria in feces of healthy controls and those who express the HLA-B27 gene.

RESULTS

Quantitative microbial profiling reveals significant variation among total microbial load not accounted for by relative microbial analysis. Genus-level fecal microbiome composition of study cohort participants (n = 23). Relative microbiota profiles deduced from standard microbiome sequencing protocols. The latter method was performed in otherwise healthy individuals which harbor the major AS susceptibility gene HLA-B27.

CONCLUSIONS

- In this study we show that quantitative microbiota profiling of the intestinal microbiota identifies novel dysbiotic changes exemplified by a five-fold decrease in fecal microbial load in AS patients relative to healthy controls.
- Quantitative microbial profiling reveals a significant increase in Roseburia colonization in AS patients not revealed by RMP alone.
- Flow-based methods reveal HLA-B27 expression drives significant changes to both the magnitude and specificity of the microbiota-specific mucosal IgA response.
- The observation of these events in healthy individuals indicate these B27-dependent events may be a predisposing event in AS pathogenesis rather than merely secondary to disease.

REFERENCES

An Uncommon Cause of an Uncommon Syndrome: A Case of Non-Sodium Dependent Osmotic Demyelination

A. Murtagh, MSIV; K. Moseman, MSIV; A. Mutasher, MD; H. Cale, MD

University of Nevada, Reno – School of Medicine – Internal Medicine Residency

Introduction

Osmotic demyelination syndrome (ODS), formerly known as central pontine myelinolysis, is traditionally associated with overly rapid correction of severe hyponatremia or relative hyponatremia after correction of a hyperosmolar hyperglycemic state.

Herein, we describe a rare presentation of ODS in the absence of sodium derangement, caused by osmotic shifts in serum glucose in a patient with uncontrolled type 2 diabetes mellitus.

Case

59 M presents to the emergency dept. with dysarthria x 2 days.
- Bilateral lower extremity weakness L>R and associated unsteadiness with ambulation, progressive x 1 month.
- Denies dysphagia, vertigo, lightheadedness, tinnitus.
- Chronic diabetic neuropathy. Denies dysphagia noted on esophagram.

Review of Systems:
- Dysuria, urgency, polyuria x 1 week.

PMHx:
- Diabetes mellitus, type II, poorly controlled, insulin requiring
- Hypertension, dyslipidemia
- Home glucometer revealed blood sugars ranging from 75 to >650 mmol/L
- Current, everyday smoker, 24 pack years
- Previous heavy drinker, quit 3 months prior
- Family history of type II diabetes in brother. No family history of stroke or cardiovascular disease.

Clinical Findings

Vital signs: BP 136/65 | HR 68 | T 98.1 | 96% RA | BMI 26.2

Neurologic exam:
- Diminished sensation on entire L side of the face. Otherwise cranial nerves II-XII intact.
- 4/5 strength with flexion and extension of the L hip & knee.
- Dysmetria and dysdiadochokinesias of upper extremities, L>R.
- Difficulty sitting upright with a tendency to lean L.
- Diminished sensation to fine touch & proprioception in bilateral feet.
- Dysphagia noted on esophagram.

Labs:
- A1c 17.5% | Glucose 315 | eGFR 48
- Urinalysis: glucose 3+ | ketones neg. | LE/nitrites neg.
- Urine drug screen: neg.
- Otherwise CBC & CMP WNL (Na 136-138). B-hydroxybutyrate 0.11.

Imaging:
- Head CT: no evidence of infarction, hemorrhage, or mass effect.

Carotid duplex, echo: within normal limits.

MRI:
- T2: Hyperintensity noted in the pons diffusely (Figures 2 & Figure 3) with normal appearing midbrain & medulla (Figure 1 & Figure 4, respectively).
- Diffusion weighted: Signal intensity in the pons consistent with osmotic demyelination (Figure 5)

Management/Outcome

The patient was treated for hyperglycemia with insulin and volume repletion per hyperglycemia protocol. He passed his swallow study on day 2 and experienced gradual resolution of his dysarthria and LE weakness over the course of 2 weeks.

Pathophysiology

The proposed mechanism for non-sodium dependent ODS is rapid and drastic increases in serum glucose resulting in hyperosmolarity. When the introduction of extracellular hypertonicity outpaces a cell’s ability to regulate its osmolality, water is drawn out of oligodendrocytes causing cell shrinkage and death.

Prognosis

Outcomes vary from full recovery to near-normal level of function to death. Neurology symptoms generally improve over weeks to months. Patients may be left with residual impairments and outcomes worsen with repeated episodes of hyperglycemia.

Discussion

This uncommon cause of an already uncommon disease process is important for clinicians to be aware of, so they may consider it in their differential when a patient presents with atypical neurological deficits.

Non-sodium dependent ODS may manifest with subacute symptoms, prompting patients to present in the outpatient setting, where neuroimaging is not always readily available. Furthermore, because deficits do not typically follow the classic patterns seen in an acute cerebrovascular accidents of specific vessels, physicians may delay referring patients for neuroimaging, postponing a definitive diagnosis.

References

Health Behavior Outcomes of Patients Utilizing the Fresh Produce Program

Lawrence Greenblatt, MD, Sarah Armstrong, MD
Christelle Tan, Jackée Okoli, Julian Xie, Peter Callejo-Black, Spencer Chang, Janice Wong, Elana Horwitz, Willis Wong
Duke Outpatient Clinic, Durham, NC

Introduction

Obesity, diabetes, and heart disease occur at a higher frequency in low-income communities due to difficulty affording and accessing healthy foods. It is important that clinics address these upstream factors in order to keep their patients healthy. The Fresh Produce Program (FPP) was launched by 5 medical students in August 2017 and provides packages of fresh produce and education materials to patients experiencing food insecurity.

Methods

Patients were referred to the program by healthcare providers. They were surveyed at baseline, one month, and six month intervals when they came into clinic to pick up their bag of food. Surveys asked about demographics, food security, food intake, cooking habits, and produce bag utilization.

The following objective data was obtained via EMR: height, weight, HbA1c, and date of most recent visit.

Pre/post-T-tests were used to analyze data on patients who received 5 or more bags of produce.

Results

Since its inception, 318 patients were referred to the program. The FPP has distributed 493 bags of produce to 187 patients over the course of 25 distribution days.

Our population served was 69% female, 77% Black/African American, 82% earning less than $25,000 in the past year, and 78% with a high school degree or less.

Survey Results

Since joining the program:
- 70% of patients reported eating more vegetables
- 76% of patients reported feeling healthier
- 90% of patients reported cooking more
- 59% of patients had improvements in food security score

No statistically significant changes in the following:
- Pre/post servings of vegetables patients report eating, meals cooked at home, subjective health ratings, difference in pre/post intervention food security scores, change in BMI or HbA1c at 6 months or 12 months

Discussion

This study is limited by a small sample size, but the results show that consistent distribution of produce has the potential to improve health behaviors among patients experiencing food insecurity, while also reducing the burden of food insecurity.

References