IgG-4 Related Retroperitoneal Fibrosis: A Rare Association with Riedel’s Thyroiditis

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Case Presentation

A 53-year-old male with history of RT previously treated with isthmectomy for compressive symptoms relief who presented with one week of severe localized lower abdominal and suprapubic pain. He denied any fever, gastrointestinal symptoms, genitourinary symptoms, or weight loss. He was initially diagnosed with acute prostatitis and was treated with ciprofloxacin without improvement of symptoms, which prompted him the second visit to the emergency room. The physical exam demonstrated a flat, soft abdomen with normal bowel sounds and no palpable masses, but with diffuse tenderness across the lower abdomen, especially in the right lower quadrant and suprapubic region.

The patient underwent a non-contrast CT scan of the abdomen and pelvis with findings significant for an extensive, predominantly right-sided retroperitoneal mass encircling the aorta, inferior vena cava and proximal right ureter producing severe obstructive uropathy of the right kidney with massive distention of the pyeocolical system and proximal ureter and marked loss of renal cortex (Figure 1).

The patient underwent right ureteral stent placement, which partially resolved hydrenephrosis and restored kidney function. The CT-guided biopsy of the retroperitoneal mass revealed fibro-inflammatory tissue without specific features (Figure 2). The immunohistochemistry staining was notable for IgG4 positive plasma cells (Figure 2) and CD68 positive histiocytes.

The patient was finally diagnosed with IgG4-related systemic fibrosclerosis. The additional lab testing showed normal LDH, uric acid and IgG4 levels. He started on high-dose prednisone at 60 mg daily. Throughout this time, he developed acute renal failure requiring additional stent placement by urology as well as refractory pain necessitating the use of narcotics.

Over the next few months, along with continuing decrease in the size of the retroperitoneal mass on the follow-up CT (Figure 3), his analgesic requirements began to decline. His renal function improved and he was able to taper prednisone to a lower dose.

Given the presence of an IgG4 positive plasma cell infiltrate, retroperitoneal fibrosis and history of RT, the diagnosis of IgG4-related retroperitoneal fibrosis in a patient with Riedel’s thyroiditis was made.

Case Discussion

IgG4-RD is an immune-mediated fibroinflammatory condition capable of affecting multiple organs. It is characterized by extensive fibrosis in various organs including the pancreato-hepato-biliary system, retroperitoneum, mesentery, aorta, salivary and lacrimal glands. Retroperitoneal fibrosis in IgG4-RD can present with poorly localized pain in the back or lower abdomen, leg edema, or hydrenephrosis from ureteral or prostate involvement.

Figure 1: Initial Imaging

Figure 2: Pathology

Figure 3: Follow-Up Imaging

Figure 4: ACR/ELAR 2019 IgG4-RD Criteria

- Presence of Inclusion criteria AND
- Absence of exclusion criteria AND
- Inclusion criteria ≥20
- Histopathologic findings
  - Lymphocytic infiltrate, IgG4/hpf, IgG4/IgG ratio
  - Serologic findings
  - Imaging findings

Conclusion

- Presence of inclusion, absence of exclusion, and inclusion criteria = 26
- Met criteria to diagnose IgG4-RD
- Steroid treatment lead to decreasing fibrosis in surrounding retroperitoneal organs
- Likely represents a rare case of IgG4-RD RF in RT patient

References

Thyrotoxic periodic paralysis (TPP) is a potentially fatal complication of hyperthyroidism and usually presents with acute muscle weakness and hypokalemia.

Diagnosis is confirmed by the presence of both hypokalemia and an increased level of T4 and T3.

Treatment includes correction of hypokalemia for the reversal of paralysis and restoration of euthyroid state for the prevention of future attacks.

A 33-year-old male presented to the ER for the second time in one day; with a complaint of weakness of bilateral lower and upper extremities of a few hours’ duration.

Diagnosed with Graves’ disease approximately one month prior and started on methimazole. Medication was discontinued after the development of a whole-body rash accompanied by severe itching and he was started on Benadryl.

The rash and itching continued over the long weekend, so the patient went to the ER where he was given epinephrine, sulfamerazine, and Benadryl for the rash and was sent home.

Other lab values including CBC, renal and liver function tests were within normal ranges.

Abnormal labs were as noted in figure 2; other lab values including TSH, free T3, and free T4 were elevated.

Event of potassium and phosphate following initial treatment.

Propenanol can alleviate TPP where potassium replacement, non-selective beta-blockade, and amending the underlying hyperthyroid state is definitive treatment of hyperthyroidism for the resolution of TPP.

A diagnosis of TPP during the initial presentation is often hindered and confused with the more common etiologies of hypokalemia and lower-extremity paralysis. It may last from a few hours to three days.

Patients are given K+ to quickly muscle recovery and shield from any cardiopulmonary complications.

Rebound hyperkalemia occurs in roughly 40% of patients, especially if they were administered >90 mEq of potassium chloride within the first 24 hours, due to the release of the intracellularly sequestered potassium and phosphate following initial treatment.

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Sarcoidosis Presenting as Obstructive Jaundice
Mikayla Forness, MSIIIa, Laura Nichols, MDb
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Case Presentation
- A 21-year-old male presented with acute epigastric pain and obstructive jaundice.
- CT of the abdomen and pelvis demonstrated a mass in the porta hepatis region, appearing to compress the common bile duct.
- Labs upon admission showed: alkaline phosphate 194, AST 102, ALT 252, total bilirubin 3.5, and direct bilirubin 2.8.
- Gastroenterology was consulted and performed upper esophageal ultrasound (EUS), which revealed three enlarged lymph nodes in the porta hepatis region surrounding and compressing the portal vein and distal common bile duct.
- Pathology from FNA revealed necrotizing granulomas with a mixed B- and T-lymphocyte population and negative AFB and GMS staining.
- Fungal serology and QuantiFERON were performed revealing only histoplasma M band, which was ultimately felt to be reactive due to prior infection.
- A chest CT was ordered due to concern for sarcoidosis and revealed a prominent left hilar lymph node as well as peribronchial capular micronodules at the posterior aspect of the left upper lobe, consistent with sarcoidosis. (Image 1)
- Pulmonology was consulted and diagnosed the patient with asymptomatic stage II pulmonary sarcoidosis, which was not an indication for initiation of steroid therapy.
- Biliary stent placement did lead to improvement of liver enzyme levels.
- Referral was placed to a tertiary care center for continued management and consideration of steroid treatment for the patient’s extrapulmonary lymphadenopathy and biliary stricture secondary to sarcoidosis.

Discussion
- Diagnosis of sarcoidosis relies on three criteria:1
  - A compatible clinical and radiographic presentation
  - Pathologic evidence of noncaseating granulomas
  - Exclusion of other diseases with similar findings (e.g. malignancy, infection)
- Sarcoidosis is a granulomatous disease characterized by the formation of non-caseating granulomas with epithelioid giant cells in affected organs. The most commonly involved organs are the mediastinal lymphatic system (95-98%) and lungs (>90%) with presenting signs ranging from asymptomatic disease to shortness of breath, fatigue, fever and weight loss.2,3,4,5,6
- Sarcoidosis is relatively common with incidence of over 50% and can manifest in several ways, including mimicking primary biliary cirrhosis, primary sclerosing cholangitis, or extrahepatic biliary obstruction.1,2,3,4,6
- Cases presenting with obstructive jaundice due to extrapulmonary lymphadenopathy, such as the patient we present, have rarely been reported.
- In nearly two-thirds of patients, spontaneous remission may occur; therefore, treatment is not indicated with asymptomatic stage I or II sarcoidosis progressive cases.1,4,5,6 (Table 1)
- Our patient presented a therapeutic challenge given the obstructive jaundice even in the context of his asymptomatic pulmonary disease, which prompted referral for treatment consideration.

Table 1: Radiographic Staging of Sarcoidosis5

<table>
<thead>
<tr>
<th>Stage</th>
<th>Chest Radiography Results</th>
<th>Rates of Spontaneous Resolution</th>
<th>Suggested Treatment</th>
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<tbody>
<tr>
<td>0</td>
<td>Normal</td>
<td>–</td>
<td>None</td>
</tr>
<tr>
<td>I</td>
<td>Bilateral hilar lymphadenopathy</td>
<td>55% to 90%</td>
<td>None</td>
</tr>
<tr>
<td>II</td>
<td>Bilateral hilar lymphadenopathy and pulmonary infiltrates</td>
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</tr>
<tr>
<td>III</td>
<td>Pulmonary infiltrates without bilateral hilar lymphadenopathy</td>
<td>10% to 20%</td>
<td>Steroids or immunosuppressants</td>
</tr>
<tr>
<td>IV</td>
<td>Diffuse pulmonary infiltrates with fibrosis</td>
<td>0% to 5%</td>
<td>Steroids or immunosuppressants</td>
</tr>
</tbody>
</table>

Conclusions
Sarcoidosis can present asymptomatically in many cases and can affect many organ systems other than the lungs. As our case illustrates, it is important for clinicians to understand both the variety of presentations of sarcoidosis, including biliary obstruction, as well as the complexities of management depending on presenting symptoms.

References

Image 1: Chest CT with contrast showing prominent left hilar lymph node.
Porphyria Cutanea Tarda: A Rare Presentation of Hemochromatosis

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Case Presentation

A 61-year-old female with history of end-stage renal disease on dialysis presented with a three-week history of a blistering rash on the dorsum of her hands and face. She had previously undergone a biopsy of the lesions which demonstrated a pauci-inflammatory cell-poor subepidermal cleft. Immunofluorescence demonstrated thick homogenous deposition within the walls of superficial dermal vessels with IgG, IgA, and fibrinogen, a pattern that is seen in sun-damaged skin, but also in porphyria.

She was admitted, and on exam, was noted to have large bullae present on the dorsum of her hands and fingers, sparing her palms (Figures 1 and 2). She also had lesions involving her face. She was noted to have a dark pigmentation, significantly changed from the picture seen in her chart (Figure 3).

Serum porphyrins were found to be elevated and subsequent plasma porphyrin fractionation demonstrated elevated uroporphyrin, heptacarboxyprotoporphyrin, and hexacarboxyprotoporphyrin, supporting a diagnosis of porphyria cutanea tarda. This was further confirmed via urine porphyrin fractionation and genetic testing showing a monoallelic mutation in uroporphyrinogen decarboxylase (UROD).

At presentation, her skin pigmentation was darkened, and it was noted that ferritin obtained one-month prior was elevated. MRI of the liver subsequently demonstrated iron overload. She underwent genetic testing, which did not detect C282Y or H63D mutations, supporting a diagnosis of secondary hemochromatosis.

Diagnosis

Based on elevated serum porphyrins and urine porphyrins in conjunction with her monoallelic UROD mutation, the patient was diagnosed with porphyria cutanea tarda. This prompted further work-up, which demonstrated iron overload in the liver. Genetic testing for hereditary hemochromatosis was negative, allowing for diagnosis of secondary hemochromatosis.

Discussion

Porphyria cutanea tarda results from intrahepatic deficiency of uroporphyrinogen decarboxylase (UROD), which functions to convert uroporphyrinogen to coproporphyrinogen in the heme biosynthetic pathway [1].

Even with a UROD mutation, other susceptibility factors, such as alcohol abuse, estrogen use, HIV, or hemochromatosis, are required to express the phenotype [1-3]. One study found that 92% of patients presenting with PCT had at least 3 of these susceptibility factors present [4].

Hemochromatosis is a disorder of iron regulation, which may present with cirrhosis, arthralgias, skin hyperpigmentation, diabetes mellitus, cardiomyopathy, hypogonadism, hepatocellular carcinoma, or osteoporosis [5].

The relationship between PCT and hemochromatosis is due to the impact of iron overload on UROD function as UROD is inactivated through an iron-dependent process [8]. Additionally, clinical remission has been linked to therapeutic depletion of iron stores through phlebotomy [7].

Conclusions

Porphyria cutanea tarda as the initial presentation for hemochromatosis is a rare phenomenon and has been infrequently reported [8-10]. However, hereditary or secondary hemochromatosis should be suspected in any patient presenting with PCT.

References

Bullous Fixed Drug Eruption Following Administration of the Recombinant Adjuvant Shingrix Vaccine
Hallie Anderson, MSIV; Laura Nichols, MD; and Tania Gonzalez Santiago, MD
University of North Dakota School of Medicine and Health Sciences and Sanford Health-Fargo

Case Presentation
A 51 year-old female with a past medical history significant for Crohn’s disease on infliximab, who presented to her primary care provider with a bullous rash on her left arm, axilla, and lateral chest wall (Figure 1) with associated subjective fever. Two days prior to presentation, she received her second dose of recombinant adjuvant Shingrix vaccine. She was not taking any new medications at the time, had not used any new topical products, and had not had a similar rash in the past. She was taking infliximab for her Crohn’s disease and was due for her next dose the following week. On physical exam the patient had diffuse erythema and swelling extending from the mid chest to axilla and down the upper arm with associated bullae, some of which had a central dusky appearance.

The patient was referred to dermatology and biopsy was performed. PCR from one of the bullae for HSV1, HSV2, and VZV was negative. Punch biopsy revealed an acute stratum corneum with epidermal necrosis leading to bullae formation along with a superficial and deep interstitial inflammation with numerous eosinophils and scattered neutrophils (Figure 2). The changes were consistent with a bullous fixed drug eruption in response to the vaccine. The patient was prescribed prednisone 40 mg daily for 5 days with subsequent taper and triamcinolone cream applied twice daily. Because results of the viral PCR were not available at the time, the patient was also given a prescription for valacyclovir 1000 mg three times a day for seven days.

Discussion
Dermatologic reactions have been reported immediately following vaccine administration in previous cases, but this is the first known case of a reaction to the recombinant Shingrix vaccine. The live Zostavax vaccine has been shown to cause fatal disseminated varicella zoster virus infections in both immunocompromised and immunocompetent patients, which is one of the factors that led to the development of an inactivated vaccine for the prevention of VZV reactivation. However, this case provides evidence that immunologically mediated reactions can occur with recombinant shingles vaccine administration despite their improved safety profile compared to the live vaccination. As in this case, patients with autoimmune disease may be at increased risk given the immune dysregulation associated with their underlying condition. Development of a bullous fixed drug eruption to medications and vaccinations is a rare complication that can appear similar to bullous pemphigoid, Stevens-Johnson Syndrome (SJS) and toxic epidermal necrolysis (TEN). The development of a bullous rash is likely due to a delayed-type hypersensitivity reaction, marked by T-cell hyperactivation. The reaction is generally self-limited and treated with discontinuation of the offending agent and corticosteroids.

Conclusion
This case highlights the possibility of an acute bullous reaction to the recombinant adjuvant Shingrix vaccine. The vaccine has been shown to be very effective at preventing VZV reactivation and postherpetic neuralgia and is well tolerated in most patients, so the benefits of receiving the vaccination outweigh the risks. Nonetheless, recognition of acute reactions associated with the vaccine such as the bullous drug eruption seen in this patient should be recognized and treated early. Additionally, clinicians should be aware of the potential for increased risk in patients with preexisting autoimmune conditions. Further investigation and post-marketing monitoring of the recombinant shingles vaccination should focus on delineating the frequency of these reactions and predisposing patient risk factors.

References
A rare presentation of seizure secondary to tricuspid valve endocarditis

Kristal Hudson & Dinesh Bande, MD

Learning Objectives

- Identify the neurological complications and their pathogenesis that can present in septic patients with infective endocarditis.
- Appreciate the role of brain imaging during the progression of neurologic symptoms and managing the risk of cranial septic embolic in infective endocarditis.

Case Presentation & Clinical Course

- 31-year old female with a significant history of IVDU presents to ED with chest pain and shortness of breath for four days after use of heroin and methamphetamine. Associated symptoms include arthralgias, weakness, and fatigue. Patient found tachypneic, hypoxic, and tachycardic. Labs revealed leukocytosis, thrombocytopenia, elevated creatinine, liver enzymes, and lactic acid. Blood cultures were drawn, and she was given Vancomycin and Zosyn empirically for severe sepsis.
- Chest CTA (Figure 1) revealed multiple nodular cavitating lesions throughout both lungs. TTE (Figure 2) showed moderate tricuspid regurgitation with mobile vegetations measuring 11 mm x 18 mm. Blood cultures positive for MSSA.
- Diagnosis of infective endocarditis (IE) was established.
- Patient continued to deteriorate to respiratory failure requiring sedation and intubation. After a month of intensive care, she was discharged to Vibra to continue ventilation weaning and IV Abx.
- 10-days later, patient presented after a rapid response involving seizure with head trauma. Brain MRI (Figure 3) showed diffuse leptomeningeal enhancement consistent with meningitis and cerebritis. LP showed no clear infection. EEG showed diffuse encephalopathy and no electrographic seizures. Antimicrobial therapy restarted for which she developed a rash and since experienced other complications and prolonged hospital course.

Discussion

- Mycotic aneurysms are arterial dilations caused by the infection spreading outwardly through the vessel wall. 
  - Mortality rate of mycotic aneurysm is dependent on unruptured (30%) and ruptured (80%) vessels.
- Patients with new neurologic symptoms undergo brain imaging (CT or MRI), especially during acute phase to thoroughly search for asymptomatic embolisms. 
  - MRI can show lesions not visible on CT, especially in IE.
  - A study showed that nearly half of asymptomatic patients with IE had early evidence of cerebral embolization via MRI.
- Early appropriate antibiotic therapy remains the cornerstone of IE management and decreases the risk of neurologic embolic events.
- Optimal timing of surgery for IE with cerebrovascular complications remains controversial.

Take Home points

- Neurological complications can arise early in disease course and could potentially be reduced with early aggressive medical and/or surgical management.
- The role of neuro imaging in the timing of surgery can aid in reducing neurologic comorbidities in IE patients but timing is still unclear.

References

A Rare Renal Complication in a Patient with Alagille Syndrome

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Introduction

- Alagille syndrome is an autosomal dominant disorder caused by a mutation of the Notch signaling pathway, most commonly in the JAG1 gene
- High degree of penetrance but variable expressivity
- Frequency of 1 in 30,000 live births
- Diagnosis is based on five "classical" criteria:
  1. Intrahepatic bile duct paucity
  2. Cardiac malformations (pulmonary artery stenosis)
  3. Skeletal involvement (butterfly vertebrae)
  4. Ocular anomalies (posterior embryotoxon)
  5. Distinctive facial features (prominent forehead, pointed chin, deep set eyes)

Case Description

- A 34-year-old male with a past medical history of Alagille syndrome consisting of secondary biliary cirrhosis, hyperlipidemia, hyperuricemia, and recurrent gout initially presented in 2011 with nephrotic range proteinuria and normal creatinine
- His serological workup was negative
- He underwent a renal biopsy which showed secondary FSGS with reduced glomeruli and sclerosis
- Lisinopril was started initially, but not tolerated due to dizziness
- He was switched to Losartan, but lost to follow up for a year
- He later presented with worsening creatinine and proteinuria and most seen recently in the follow up clinic with a creatinine of 3.07 mg/dL and 1.2 gm proteinuria
- The patient is currently being managed with Losartan for the proteinuria, allopurinol, and a low salt diet.
- He has been referred to Mayo clinic for a simultaneous kidney and liver transplant.

Pathology

- Figure 2. Renal biopsy of a 5-year-old girl with Alagille Syndrome. Trichrome staining shows FSGS in two glomeruli (dense, pale blue areas); arrows show podocytes with prominent red cytoplasmic vacuoles; inset shows an oil-red O stain which highlights lipid (red) deposited in a single capillary loop.

Discussion

- FSGS is rarely reported in the literature to be associated with Alagille syndrome
  – Thought to be related to high circulating lipids seen in this syndrome that deposit within the glomerulus and may contribute to podocyte injury
- The patient described has proteinuria, CKD stage 4, and FSGS which is felt to be related to his underlying Alagille syndrome
- Liver disease is a major cause of morbidity in Alagille syndrome, whereas cardiac and vascular involvement account for most of the mortality
- Since renal disease can impact liver transplantation, additional studies on renal involvement in Alagille syndrome are necessary to develop guidelines on evaluation and management

Take Home Points

1. A renal evaluation consisting of serum biochemistry, renal ultrasound, and urinalysis should be standard of care in Alagille Syndrome
2. Additional studies on renal involvement in Alagille syndrome are necessary as renal disease can impact liver transplantation
3. Studies have suggested that renal involvement be considered a sixth "classical" criterion for diagnosis

References

Epstein Barr Virus Infectious Mononucleosis complicated by profound lymphadenopathy, ruptured duodenal ulcer and sepsis

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Introduction

• Epstein Barr Virus (EBV) is a common illness causing infectious mononucleosis (IM).
• Worldwide, >90% of adults are infected with EBV. IM affects up to 70% of adolescents and adults, while infants and children are typically asymptomatic with primary infection.
• The most common clinical symptoms include fatigue, fever, pharyngitis, and cervical lymphadenopathy.
• Although most acute infections are mild and self-limiting, the complications vary widely.
• Few cases have reported EBV with concurrent gastric or duodenal ulcers, and ulcer perforation significantly increases the risk of severe complications.

Case Description

• A 39-year-old previously healthy woman presented with one-week history of pharyngitis, cervical lymphadenitis, fever, and fatigue. Monospot test was negative. She was sent home on antibiotics. With 3 doses, she failed to improve and developed nausea and diarrhea.
• Two days later, she was admitted for acute kidney injury due to dehydration. Serology was positive for EBV nuclear antigen, IgG and IgM. Cervical lymph node biopsy showed lymphoid hyperplasia typical of EBV without evidence of lymphoma.
• Abdominal imaging revealed splenomegaly and multiple enlarged lymph nodes consistent with EBV infection. She also had hepatic steatosis with elevated liver enzymes, intrahepatic biliary duct dilatation, and mild pancreatic duct dilatation.
• CT abdomen (Figure) shows prominent gastrohepatic nodes.

• In hospital, she developed worsening abdominal pain and signs of sepsis. Abdominal CT was ordered, revealing free fluid and pneumoperitoneum. Exploratory laparotomy discovered perforated duodenal ulcer which was repaired with a Graham patch. Hepatic artery lymph node biopsy found follicular hyperplasia consistent with EBV-associated lymphadenitis.
• Post-op, she developed acute respiratory failure, aspiration, and signs of sepsis. She was intubated and moved to the ICU.
• The patient’s symptoms gradually improved with intensive care. She was discharged home after a 25-day hospital course.

Discussion

• Well-known complications of EBV-IM include hepatosplenomegaly with elevated LFTs and risk of splenic rupture, lymphoproliferative disorders, and lymphomas. Mild hepatitis is not uncommon though steatosis was likely pre-existing in this case.
• Multiple cases reported lymphoepithelioma-like carcinoma or gastric adenocarcinoma contributed to EBV. Oral, skin or colonic ulcers are established complications in immunosuppression. However, only 3 cases reported benign gastroduodenal ulcers in immunosuppressed patients, and 4 cases reported gastric or duodenal ulcers in immunocompetent patients with EBV-IM.
• In one study, EBV serology was tested in 50 patients with peptic ulcer disease were tested for EBV-DNA. 35 of 50 ulcer samples had significantly higher EBV viral load versus normal gastric tissue.
• A systematic review cited 6 different studies describing EBV and coinfection with H. pylori. Their conclusions supported preliminary evidence that H. pylori may increase lytic activity of EBV.
• Non-steroidal anti-inflammatory drugs (NSAIDs) likely contributed to ulcer formation in this case as she took frequent doses prior to hospitalization for cervical lymphadenitis. NSAID use with EBV infection could render the gastric mucosa more susceptible to ulcer formation, but this requires further investigation which is challenging due to rare concurrence of these conditions.

Conclusion

• Most patients with EBV infections recover uneventfully. However, few patients may develop severe complications including sepsis and profound lymphadenopathy as demonstrated in this case.
Introduction

ACE inhibitor-induced angioedema occurs in only 0.7% of patients who take ACE inhibitors, but with over 40 million patients taking this medication it is a noteworthy side effect1. Most attention is paid to the life-threatening airway obstruction ACE inhibitors can cause, and intestinal angioedema is a less well-known manifestation. This can result in delay of diagnosis and several unnecessary interventions in patients who experience isolated angioedema of the gut.

Case Summary

A 38-year-old male with hypertension treated with lisinopril presented to the emergency room after two days of diffuse, debilitating abdominal pain. He was hypotensive and in lactic acidosis. Abdominal examination revealed diffuse rigidity and tenderness. CT was concerning for viscus perforation, so surgery was consulted. Exploratory laparotomy revealed an edematous small bowel without ischemia or perforation. The patient’s abdomen was left open and he spent one day in the ICU on pressors before re-exploration and closure. He stabilized and was discharged without a diagnosis, and his lisinopril was restarted. The next day, MRI enterogram showed significant improvement, confirming the diagnosis.

Discussion

This patient experienced symptoms for months before the source of his intestinal edema was identified. A literature review suggests that such a delay is not uncommon. Virtually all patients presented with abdominal pain which, as in this case, may mimic an acute abdomen. A case report about angioedema from acquired C1 esterase inhibitor deficiency stated that up to one-third of patients undergo exploratory laparotomy, appendectomy, or both during attacks2. Another study reported that 35% of patients with intestinal angioedema experienced longstanding abdominal pain before being diagnosed2.

If there’s suspicion for angioedema in a patient taking an ACE inhibitor, the medication should be discontinued until another etiology is determined. Imaging can support this decision, and CT or ultrasound findings include mesenteric edema, intestinal wall thickening, dilated bowel, and/or ascites. One article referred to this as a “stacked coin” appearance3. Most cases involve the small intestine, with the majority occurring in the jejunum4.

After cessation of the ACE inhibitor, angioedema should subside in 24-72 hours. If this condition is not identified and the medication continues, the angioedema will recur with increasing frequency and severity. Patients may still experience episodes for several months, but these should decrease in number and intensity over time2. If symptoms do not subside, alternative causes of angioedema such as hereditary angioedema and acquired C1 inhibitor deficiency should be explored.

Conclusion

This patient initially underwent an exploratory laparotomy, which confirmed what was seen on CT: gut edema. Given the acuity of his presentation (severe hypotension and lactic acidosis), a surgery may not have been avoided, but the additional two ED visits, hospitalization, and 4.5 months of symptoms may not have occurred had an etiology of ACE-inhibitor induced angioedema been considered. One should have a low index of suspicion when apparent cause of an acute abdomen is not clear after initial investigations in a patient taking an ACE inhibitor.

As of 10 months since lisinopril was discontinued, this patient has had no attacks requiring additional care.

References

Reactive Arthritis: An Unusual Presentation of Acute *Clostridium difficile* Colitis

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**Introduction**

- Reactive arthritis is a rare presentation of acute *Clostridium difficile* colitis and requires a high level of clinical suspicion to diagnose.
- Classically, reactive arthritis is associated with *Salmonella*, *Shigella*, *Campylobacter*, *Chlamydia*, and *Yersinia*.
- *Clostridium difficile* has only been cited as the etiology of reactive arthritis roughly 50 times in the literature thus far.

**Hospital Admission and Course**

- Two days later came to the emergency department with worsening severe polyarthralgia and diarrhea associated with chills and fatigue.
- Physical exam revealed extreme tenderness to palpation of joints without erythema or swelling.
- Denied oral antibiotic use within the past month.

**Initial Presentation to Urgent Care**

- 20-year-old Caucasian male with history of psoriasis presented to urgent care clinic with a 2 week history of polyarthralgia and a 3 week history of nonbloody diarrhea.
- Extensive workup showed normal rheumatoid factor and was negative for basic enteric panel, tick borne illnesses, and others.
- Sent home with prednisone and plan for follow up with rheumatology.

**Labs and Imaging**

**Figure 1.** CT showing abnormal cecal wall thickening and concern for inflammatory bowel disease.

**Figure 2.** Colonoscopy showing pseudomembranous colitis which prompted aspirate for *C. difficile*.

**Conclusions**

- Undiagnosed IBD can also present as *C. difficile* colitis, making the diagnosis of reactive arthritis difficult.
- Diagnosis was made with features of septic arthritis, a positive pathogen documentation and negative tissue biopsies of the colon ruling out other etiologies for diarrhea.
- Antibiotics coupled with NSAID therapy results in excellent prognosis of reactive arthritis.

**Significance**

- *C. difficile* colitis cases continue to rise with widespread antibiotic use and hospital acquired infection.
- Important to recognize *C. difficile* as an etiological agent of reactive arthritis, despite fewer number of case documentations.
- Clinicians should think of reactive arthritis when chief complain is severe polyarthralgia developing within 1-4 weeks of an enteric infection.

**References**


**Tables**

<table>
<thead>
<tr>
<th>Infectious agent testing</th>
<th>Urgent Care Facility</th>
<th>Hospital Admission</th>
<th>Reference</th>
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<tr>
<td>Basic Enteric Pathogen Panel</td>
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<tr>
<td>Toxicogenic <em>C. difficile</em> (PaLoc)</td>
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<tr>
<td>Giardia</td>
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- Aspirate returned positive for *C. difficile* toxin and vancomycin was started.
- At this point, still more likely that his polyarthralgia was due to undiagnosed inflammatory bowel disease (IBD) flare; however, tissue biopsy confirmed diagnosis of reactive arthritis in the setting of *C. difficile* colitis.
- No histopathological evidence of chronicity to suggest ulcerative colitis or Crohn’s as the etiology, ruling out IBD.
Rituximab: a rare cause of Progressive Multifactorial Leukoencephalopathy

Aishwarya Sharma MS1, Mounika Guduru, MD1, Dinesh Bande, MD1,2, Abhishek Matta, MD1,3

1 University of North Dakota School of Medicine and Health Sciences; 2 Creighton University Medical Center; 3 Sanford Health, Fargo ND

Case Description

• A 79-year-old woman with a known history of rheumatoid arthritis on long-term prednisone (5mg daily) and rituximab (1000mg every six months), presented to the emergency room with multiple falls, gait difficulty, slurred speech, and confusion.
• On hospital admission, her physical examination revealed right spastic hemiparesis (a known residual from her old cerebrovascular insult), axial and appendicular ataxia, slurred dysarthria, and vertical nystagmus.
• She was negative for HIV, HbsAg, and Anti-HCV.
• Computed tomography (CT) scan of the brain showed right pontine and right cerebellum hypodense irregular lesion without significant surrounding edema or mass effect and left parietal cortical and subcortical encephalomalacia, likely sequel are of an old vascular insult (Figure 1).
• Magnetic resonance imaging (MRI) with gadolinium of the brain and posterior fossa revealed the right pontine lesion (at the brachial pontis) as involving the right cerebellar white matter. The lesion was isointense on T1 and hyper intense on T2 and FLAIR sequences. The outer borders of the right pontocerebellar lesion were more defined than the inner border and there was minimal surrounding edema, with no mass effect on the adjacent fourth ventricle.
• The lesions showed diffusion restriction on diffusion-weighted images (DWI) and apparent diffusion coefficient (ADC) maps. The contrast study showed heterogenous leading edge enhancement at the right pontocerebellar lesion (Figure 2). A smaller T2 hyper intense lesion was also noted in the left middle cerebellar peduncle, extending to the left cerebellar hemisphere with heterogenous enhancement.
• These radiological features, along with the clinical features in a rituximab-treated immunocompromised patient are highly suggestive of Progressive multifocal leukoencephalopathy (PML).
• Treatment options were discussed with the patient, However, the patient and her family decided to adopt a palliative approach, and the patient was discharged from the hospital to a nursing home where she died within a month.

Discussion

• PML is a rare fatal central nervous system disorder characterized by infection-induced demyelination of white matter due to the opportunistic reactivating of John Cunningham (JC) polyomavirus in an immunocompromised patient [1,2].
• JC-virus primary infection often occurs during childhood or early adolescence but the virus stays dormant in body tissues for long-periods in immunocompetent individuals. Several studies in the literature reported that up to 50% of adults may be seropositive for JC-virus [3].
• However, an intact immune system suppresses viral activation. In an immunocompromised state, the virus becomes reactivated, resulting in disease [4].
• Immunocompromised states such as HIV infection, leukemias, lymphomas, and different malignancies are well-known to be associated with PML. Several reported cases in the literature show a strong association of PML with some immunosuppressants such as natalizumab, fingolimod, dimethyl fumarate, and rituximab [4-6].
• Rituximab is an anti-CD20 monoclonal antibody used in the treatment of many lymphoproliferative conditions and many immunomeditated diseases such as non-Hodgkin’s lymphoma, neuromyelitis optica, psoriasis, and rheumatoid arthritis [7] via various mechanisms of action such as antibody-dependent cytotoxicity, cell-mediated cytotoxicity, apoptosis, and direct sensitization of cells to chemotherapy [8].

Conclusion

• As PML can be fatal, patients receiving rituximab should be meticulously monitored.
• Regular follow-up of CD4b is essential, and abrupt discontinuation of rituximab is fundamental to reduce mortality if PML is suspected.
• Although there is no definitive FDA approved medication for PML to date, plasma exchange to remove circulating drug as well as stopping the offending drug represent the treatment of choice for these cases.

Figure 1: Brain CT axial (a) and coronal (b) sections showing a right cerebellar hypodense lesion with no significant surrounding edema or mass effect and left cortical and subcortical parietal encephalomalacia (on coronal section).

Figure 2: MRI brain with contrast showing an irregular right pontine isointense lesion on T1 (a), a hyper intense signal on T2 (c) and FLAIR (d) sequences, with the extension of the lesion to the right cerebellar hemisphere showing minimal edema adjacent to fourth ventricle. The lesion shows restriction on DWI (e) and ADC map (f), and heterogenous enhancement at the margins (g, h). Another T2 hyper intense smaller lesion is demonstrated at the left middle cerebellar peduncle and left cerebellar hemisphere (c) and heterogenous enhancement (h).

References

Infectious Complications of Recreational Urethral Sounding with Retained Foreign Body

Aishwarya Sharma MSI1, Dubert Guerrero, MD2; 1 University of North Dakota School of Medicine and Health Sciences; 2 Sanford Health, Fargo ND

Case Description

- A 62-year-old male was admitted for evaluation and management of 3 weeks of progressive back pain radiating to his legs with a history of nicotine and amphetamine abuse, hypertension, mood disorder, and neuropathic pain.
- Physical examination including neurologic assessment was unremarkable.
- White blood cell count was normal at 8.6 K/ul but inflammatory markers erythrocyte sedimentation rate and C-reactive protein were significantly elevated at 94 mm/hr and 110 mg/L respectively.
- Magnetic resonance imaging (MRI) of the back revealed abnormal marrow signal and enhancement in the T12 and L1 vertebral bodies centered at the T12-L1 disc space likely secondary to diskitis. There is corresponding abnormal paraspinal edema and enhancement with a probable 1.7 cm intramuscular abscess in the left psos muscle (Figure 1).
- The abscess was aspirated and grew Staphylococcus epidermidis. He was placed on intravenous cefazolin.
- He also complained of dysuria and hematuria. Further history revealed frequent urinary tract infections with S. epidermidis for which he received different courses of oral antibiotics without relief.
- A computed tomography urogram was done for persistent urinary symptoms and identified a tubular 1.5cm diameter peripherally calcified 10-12cm structure with tapered distal ends and intermediate internal attenuation coiled in the urinary bladder (Figure 2).
- After careful history, he admitted his girlfriend inserted a sex toy shaped like a fishing worm into his urethra few months back, but he did not remember if it was removed.
- He underwent cystoscopy and open cystolithotripsy in which the foreign body was extracted (Figure 3). The patient was discharged to skilled nursing facility to complete IV antibiotics for 6 weeks followed by oral cephalexin for additional 6 weeks.
- Repeat MRI showed destruction in the intervertebral disc between T12 and L1 and paraspinal soft tissue enhancement. He eventually required a T12 & L1 corpectomy and posterior instrumented fusion of T9-L3.

Discussion

- Foreign bodies localized to the urogenital tract represent a relatively rare pathology and their presence may predispose to infection, calculus formation, and bladder outlet obstruction, bacteremia and abscesses [1].
- Most patients are hesitant to admit urethral sounding or the act of inserting an object for autoerotic, psychiatric, therapeutic, or any other reasons [2]. Patients present when symptoms develop or there are complications.
- Hematuria may occur from trauma due to self-manipulation or rough objects that injure the bladder wall.
- Foreign bodies in the urinary tract increase the risk of urinary tract infection [1]. Such infections are often recurrent as a result of bacteria persistence within or on the foreign body and sometimes because of poor drainage.
- Bacteremia and risk of endotoxemia should always be borne in mind and risk may increase in the process of extraction of the foreign body.
- Complications such as calculus formation have been widely reported in association with migrated intruterine contraceptive devices and surgical needles [3,4].
- Radiologic evaluation is necessary to determine the exact size, number, and nature of the foreign body. Ultrasonography is usually able to localize the foreign body to the bladder and determine the exact size and number but is unable to evaluate the exact nature.
- Cystoscopy confirms the diagnosis, and some foreign bodies are successfully removed during the process [5]. Endoscopic and minimally invasive techniques should be encouraged.
- Our patient was treated for recurrent urinary tract infections of the same pathogen for over 6 months before he eventually ended up with bacteremia and deep-seated infectious complication of disksitis that was managed both medically and surgically.

Conclusion

- The presence of a foreign body in the urinary bladder from urethral sounding is rare and therefore requires a high index of suspicion for diagnosis.
- Presentation is sometimes delayed with complications that may be life-threatening.
- It should be considered in patients with recurrent urinary tract infection and poor response to antibiotic therapy. Such patients should have radiologic evaluation in order to facilitate diagnosis and treatment.

References

Introduction

- Systemic lupus erythematosus (SLE) – “the great imitator”
  - frequently associated with overlap syndromes with other rheumatologic illnesses such as rheumatoid arthritis
- Overlap with ankylosing spondylitis (AS) rarely reported
- We present a case of a patient with a known diagnosis of SLE who presented with low back pain and was ultimately diagnosed with an SLE-AS overlap syndrome

Case Report

- A 48-year-old female with a history of SLE controlled on azathioprine presented with subacute intermittent low back pain radiating to her groin and left flank pain
- Saw improvement with chiropractic intervention but experienced subsequent worsening of her pain
- No red flag symptoms
  - weakness, numbness, tingling, bowel or bladder dysfunction
- Left flank tenderness to palpation
- Given pain on palpation and lack of red flag symptoms, it was felt the most likely cause of her pain was muscle strain
- Concern for a renal stone with radiation of flank pain to the groin, thus CT kidney stone protocol and UA were performed
- UA was negative
- CT was negative for renal stones but showed chronic sclerotic and erosive changes on either side of both SI joints with typical sequela of sacroiliitis and a chronic left-sided pars interarticularis defect at L5 (Figure 1 and 2)
- Discussed with her rheumatologist given sacroiliitis on imaging
- At rheumatology follow up, she reported intermittent low back pain since her 20s in a pattern of stiffness at night with improvement with movement
- Diagnosed with ankylosing spondylitis in addition to her SLE
  - Based on inflammatory back pain in the presence of SI joint inflammation on imaging
- HLA-B27 measurement was not required for diagnosis
- Pain was treated successfully with celecoxib

Discussion

- Concomitant SLE and AS rarely reported in the literature
- SLE can cause a polyarticular synovitis, yet abnormalities within synovial lined sacroiliac joints are reported in only a few cases
- Diagnosis of AS includes radiological changes in the sacroiliac joints and joint pain >3 months that improves with exercise
- Still unknown if genetic factors like HLA-B27 with DR3 and/or DR5 play a critical role or whether the sacroiliitis is simply an independent and infrequent manifestation of SLE
- Genetic testing showing HLA-B27+ not necessary for diagnosis
- AS is an uncommon (0.3% of all cases) cause of low back pain
- Our case illustrates the importance of considering inflammatory back pain in the differential diagnosis, particularly in patients with positive histories and rheumatologic conditions like SLE

References

Introduction

- Central Retinal Artery Occlusion (CRAO) constitutes an ophthalmic emergency
- CRAO most commonly occurs through embolic occlusion of the central retinal artery from atherosclerotic plaques embolizing from the internal carotid artery
- Retinal ischemia can cause an acute, painless vision loss
- CRAO is a relatively rare condition, with an incidence of 1:2 per 100,000, 1-2% of which are bilateral cases
- Conservative management includes anterior chamber paracentesis, CO and oxygen inhalation therapy, ocular massage, topical beta-blockers, and IV acetazolamide

Case Report

- A 66-year-old female, with a past medical history of hypertension, dyslipidemia, and 50-pack-year smoking history presented to the ED with three to five hours of transient, painless, monocular vision loss in her right eye
- Since symptom onset, she had experienced several episodes of vision loss, with intermittent return of normal vision
- On physical exam
  - she had a blood pressure of 223/105
  - Clonus was noted, but vision was intact in both eyes
  - No corneal abrasions or ulcers were seen
- Intraocular pressure was measured at 20 mmHg, bedside ultrasound did not show any obvious retinal detachment; and slit lamp exam of the anterior eye, lid, and orbit did not reveal any abnormal findings other than cataracts in her right eye
- At that point, inflammatory markers were tested, and a CTA Head and Neck was performed to rule out temporal arteritis and stroke, respectively
- While these tests were being run, she had another episode of vision loss, which coincided approximately three-and-a-half hours after her last-known well time
- Angiography took place approximately 4 hours and 39 minutes from the last known well time
- No stenosis was seen at the origin of the right internal carotid artery and its intracranial course was within normal
- Selective microangiography was then performed, which also showed decreased flow in the right central retinal artery, with significantly delayed choroidal blush
- We injected 5 mg of intra-arterial tPA over 15 minutes
- Following injection, normal flow was seen in the central retinal artery, with a normal choroidal blush (Figure 1)
- On follow-up no visual abnormalities were seen

Discussion

- The clinical findings of CRAO are often easily recognized making the diagnosis straightforward
- While visual improvement can spontaneously occur following CRAO, improvement largely depends upon both the duration and type of CRAO
- Additionally, recent discussion has focused on the length of retinal survival time as a crucial factor in prognosis
- Furthermore, much has been written about the diagnosis itself, however, uncertainty and disagreement exist in management and treatment
- Non-arteritic transient CRAO (transient monocular blindness) accounts for 15–17% of CRAOs, which is what was seen in this patient
- Interestingly, while this patient did have history that would suggest atherosclerotic disease, she did not have significant stenosis of her right carotid or ophthalmic arteries, when viewed during cerebral angiography, which would explain her symptoms
- Hence, work-up was underway for hypercoagulable states and cardio-embolic origins
- Of note, as of October 2020, no abnormalities were seen via loop recorder or on any of her hypercoagulable labs

Figure 1. Angiogram of CRAO. Before (delayed choroidal blush) and After tPA (restored choroidal blush.)
Maggot Wound Therapy Associated with *Wohlfahrtiimonas chitiniclastica* Blood Infection: Case Report and Review of Literature.

Peter Bueide\(^1\), Jeff Hunt, DO\(^1\), Dubert Guerrero, MD\(^1\)

\(^1\)Sanford Health; \(^2\)University of North Dakota School of Medicine and Health Sciences

**Introduction**

- Maggot therapy was made popular during the First World War and is still occasionally used to treat infections.
- *Wohlfahrtiimonas chitiniclastica* is an aerobic, nonmotile, gram-negative rod first isolated in 2008 from *Wohlfahrtia magnifica* maggot.
- 23 previous human cases of *W. chitiniclastica* have been reported so far.
- We add this case of *W. chitiniclastica* bacteremia which is the first to be associated with maggot therapy.
- He was discharged on levofloxacin.
- He underwent left facial, head and neck excision.
- Review of previous human cases of *W. chitiniclastica* including this case report is summarized in Table 1.

**Case Report**

- 70-year-old male with a PMH of stage I large B cell non-Hodgkin lymphoma diagnosed in 2012 treated with 3 cycles of R-CHOP now in remission.
- Noticed a slow growing ulcerating mass in the left temple for the past year.
- Sought alternative medicine therapies with a naturopathic provider with no improvement.
- Attempted maggot therapy.
- Mechanical fall resulted in an emergency department visit at an outside facility.
- Noted large ulcerative and infiltrative soft tissue lesion in the left temple with evidence of secondary infection and parasitic infection (Figure 1).
- Subsequently admitted to Sanford Medical Center for further evaluation and management and was started on empiric antibiotics.
- Tissue biopsy revealed invasive squamous cell carcinoma.
- Secondary bacterial infection of the wound:
  - Local cultures: *Staphylococcus aureus* and *Proteus mirabilis*.
  - Blood cultures: *Wohlfahrtiimonas chitiniclastica*.

**Figure 1.** Myiasis of the left temple by an unidentified species of maggot (left) and appearance of the wound following debridement in the emergency room (right).

**Discussion**

- Previous human cases of *W. chitiniclastica* have occurred globally.
- This is the first reported case of *W. chitiniclastica* bacteremia associated with medical maggot therapy.
- Maggots have been demonstrated to secrete substances, defensins, that may result in the beneficial antimicrobial properties of maggot therapy.
- Bacteria of the maggot microbiome may still be a source of infection.
- Sterilization precautions are typically followed throughout the processing and utilization of maggots for wound debridement.
- *Wohlfahrtiimonas chitiniclastica* was first isolated in *Wohlfahrtia magnifica* but has been identified in the gut of *L. sericata, Musca domestica,* and *Hermetia illucens*.
- *L. Sericata* is commonly used for medicinal purposes in maggot therapy.
- Chronic wounds, myiasis, and underlying medical and social conditions impacting personal sanitation are risk factors for infection.
- A total of 6 out of 24 patients were deceased.

**Conclusions**

- This is the first case of *W. chitiniclastica* bacteremia associated with medical maggot therapy.
- Review of literature suggested chronic wounds and myiasis as well as medical and social conditions are risk factors for the infection.
- Infection associated mortality is up to 25%.

**Acknowledgements**

We would like to thanks the staff at Sanford Health for help throughout this process.

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**Table 1.** Demographic information and outcomes data associated with reported Cases of *W. chitiniclastica* reported globally.
Tale of a Twisted PICC Line in Pandemic Times
Madeline DeFrance, MS4; Siddharth Singhal, MD; Yuri Nakasato, MD

Introduction
Peripherally inserted central catheter (PICC) lines are commonly used for safe and prolonged administration of drugs and fluids in hospitalized patients. One complication of PICC lines is malposition, with a reported incidence of 5-31%. Further complications of malposition are patient discomfort and loss of line usage. Due to these possible complications, malpositioned lines are usually repositioned or removed and replaced [1]. We present a case of a malpositioned PICC line that was unable to be repositioned and continued to be used for IV antibiotics and lab draws.

Case Report
- 94-year-old male admitted to the hospital with delirium, fever, and hypotension
- Diagnosed with a urinary tract infection (UTI) and found to be COVID-19 positive
- Significantly agitated and combative due to acute delirium with a history of Alzheimer's
- Placed on IV antibiotics for UTI
- Patient was confused and pulled out IV on hospital day three
- PICC line was inserted for antibiotics and lab draws
- No reported complications and PICC line easily drew back blood and flushed
- Chest x-ray following procedure showed PICC line curved back on itself with the tip in the left axillary vein (Figure 1)
- PICC line continued to function well for IV fluids and lab draws and did not appear to be causing any discomfort

Case Report Cont.
- Continued low grade fevers after eleven days of antibiotics
- Transferred to hospice due to ongoing fevers and COVID infection
- Discussion regarding the best process for removal of the coiled PICC line since the patient was hospitalized at a smaller facility without a vascular surgeon
- Interventional radiology from an outside facility was consulted and felt that the PICC line should come out easily with no need for transfer
- General surgery removed the PICC line successfully
- No further complications related to the PICC line removal
- Patient passed away after 7 days in hospice

Discussion
Tip position of a PICC line is critically important to avoid complications, such as thrombosis and catheter failure. It is standard practice to check the position of the tip after every placement of a PICC line and reposition if necessary. The commonality between many case reports is that malpositioned PICC lines were repositioned to avoid complications. We report a case where the malpositioned PICC line was unable to be repositioned due to severe patient agitation and complications of the COVID-19 pandemic. The PICC line was able to be used safely to deliver antibiotics for an 11-day period. Additionally, the coiled PICC line was able to be removed safely by a general surgeon without the assistance of specialty vascular care. This case suggests that as a last resort, it is possible for PICC lines to be safely used in a malpositioned state and could provide guidance in similar rare situations.

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References

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