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Serum Lipase levels carry prognostic value in Small Bowel Obstruction

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Abstract

Background: In small bowel obstruction (SBO), there is an interruption in the passage of intestinal contents through the gut lumen. Case reports have described elevated pancreatic enzymes in patients with small bowel obstruction initially misdiagnosed as having pancreatitis. However, no study addressed this issue systematically, or investigated prognostic value of this association.

Methods: Patients with SBO and at least one serum lipase level identified between September 2013 and September 2016 were evaluated. Demographic, laboratory and imaging information; need for surgery or intensive care unit and mortality were evaluated.

Results: Of 344 patients, 38 (11%) had elevated serum lipase. Patients with tomographic diagnosis of pancreatitis were excluded. Patients with elevated lipase had significantly higher levels of creatinine and lactic acid levels, length of stay, need for intensive care monitoring and death than patients with normal lipase levels. The need for surgical intervention was not significantly different between the two groups.

Conclusion: Small bowel obstruction should be considered in the differential diagnosis for elevated pancreatic enzymes. Abdominal imaging constitutes a useful resource to differentiate SBO from acute pancreatitis. In SBO, elevated serum lipase is a risk factor for higher serum creatinine and lactate, use of ICU, and mortality.

Keywords: Small Bowel Obstruction; Lipase; Pancreatic enzymes

Introduction

Small bowel obstruction (SBO) occurs when there is an interruption in the passage of intestinal contents through the gut lumen. Such interruptions can result from adhesions, hernias, tumors or inflammatory conditions such as Crohn's disease. Patients with SBO classically present with nausea, vomiting and abdominal pain along with constipation and/or obstipation. These symptoms lack specificity, and can also be present in acute pancreatitis. Therefore, radiological findings become essential in the differentiation of those conditions. In acute pancreatitis, the diagnosis can be based solely on clinical presentation and elevated serum lipase levels, while imaging studies are necessary when
alternative diagnoses are considered. In SBO the diagnosis is both clinical and radiologic. A diagnostic delay can lead to enteric necrosis, sepsis, and higher mortality.¹ Elevation of pancreatic enzymes in SBO has been reported in isolated cases.³,⁴ A retrospective review among patients who underwent surgery for small bowel obstruction showed elevation of serum lipase in half of the cohort.⁵ No study assesses the implications of such association.

The purpose of this study is to determine the frequency of serum lipase elevations among patients with SBO and whether this association carries prognostic value.

**Materials and Methods**

This study was performed at Lyndon Baines Johnson hospital, a large tertiary care center in Houston, Texas after obtaining Institutional Board Review approval. The electronic medical records of all adult patients diagnosed with small bowel obstruction between September 2013 and September 2016 were retrospectively reviewed. Patient consent requirements were waived.

*Inclusion criteria:*
We screened our electronic database for patients 18 years old and older, with ICD 9 and 10 codes for small bowel obstruction. Of those, we selected those whose diagnosis was confirmed by abdominal X ray or computed tomography (CT) using established guidelines.¹ Finally, we enrolled those with at least one serum lipase level measured during the day when the imaging studies were obtained ±24 hours.

Obstructive findings on abdominal X ray or CT abdomen/pelvis included dilated loops of proximal bowel with distal collapsed bowel and the presence of air-fluid levels, located within the duodenum, jejunum or ileum.

*Exclusion criteria:*
Patients who did not have a lipase level drawn during their hospital course, and those with large bowel obstruction or paralytic ileus were excluded.

We gathered demographic information (age and gender), laboratory results (serum lipase level, white blood cell count, serum creatinine, and lactic acid), imaging studies supportive of SBO, cause of the obstruction when known, need for operative management, length of hospital stay, need for intensive care unit (ICU) care, and mortality. Inconsistent documentation in the reviewed records limited our ability to calculate disease-severity scores such as SOFA or APACHE-2.

Serum creatinine normal range is 0.6-1.3 mg/dL. Lactic acid normal range is 0.4-2 mmol/L. The normal value of serum lipase in our hospital is 73-393 U/L. Any level above 393 U/L was considered as elevated.
Statistical Analysis:
Categorical variables were analyzed using the Fisher exact test, and discrete variables were analyzed using the Student t test for unpaired samples. A two-sided P < 0.05 was considered indicative of statistical significance.

Results

We included 344 patients. Baseline characteristics are described on table 1. The cause for SBO was known in 229 (67%) patients and included mostly adhesions (96 patients, 42%), strictures (65 patients, 28%), and tumors (54 patients, 24%)

Thirty-eight (11%) had elevated serum lipase levels (range 394 to 2740 U/L). Eleven (3%) had levels three times above normal or higher. Eight patients had also tomographic evidence of acute pancreatitis; these patients were excluded from the comparative analysis between patients with normal and elevated lipase presented in table 2. Patients with elevated serum lipase had more commonly concomitant elevations of serum lactate and serum creatinine. While elevated serum lipase did not predict a bigger need for surgical intervention, it did predict longer length of stay, higher use of ICU and higher mortality.

Discussion

We found that serum lipase elevation carries negative prognostic implications, including concomitant elevations of serum lactate and creatinine, more use of ICU and higher mortality. To our knowledge, this is the first study that assesses the prognostic value of this association.

Serum lipase level elevation was described before in patients with SBO requiring surgery after bariatric surgery. However, the article did not specifically address the influence of lipase levels on patient outcome. Moreover, our cohort included patients with any mechanical obstruction, regardless of whether they had gastric bypass surgery.

Decreased glomerular filtration rate under 50 ml/min is described as a cause of elevated serum pancreatic enzymes, though this association seems more evident for serum amylase than lipase. Furthermore, a combination of amylase response to secretin, and autopsy findings among patients with end-stage renal disease (ESRD), suggests that pancreatitis, found in up to 50% of those cases, is not responsible for all cases of hyperamylasemia in that complex cohort. In our cohort, after exclusion of patients with ESRD, we found that patients with elevated serum lipase more frequently had serum creatinine levels above 1.5 mg/dL. The range of creatinine elevation went from 1.5 to 8.4 mg/dL in the non-ESRD group. Only 4 of these patients had GFR barely above 50 ml/min when calculated using the MDRD equation.

Though we couldn’t calculate disease-severity indices in our cohort, we believe the most likely explanation for elevated lipase in our cohort is pancreatic ischemia. A supportive
factor is the concomitantly elevated serum lactate. A study showed elevated serum lipase in 40% of severely ill patients admitted to the ICU (5% with diagnosis of acute pancreatitis). This finding was associated with longer mechanical ventilation, longer ICU length of stay, but not higher mortality.\textsuperscript{11} Visceral hypoperfusion and pancreatic ischemia are the proposed underlying mechanisms in conditions like SBO, where hypovolemia is present.\textsuperscript{1,12,13}

Another hypothesis explaining the elevation of pancreatic enzymes comes from studies including patients with bariatric surgery. It is thought that the lipase elevation results from increased intraluminal back pressure caused by the obstruction. This can lead to a reflux of intestinal content into pancreatic and biliary ducts that subsequently activates pancreatic zymogens. This concept is referred to as reflux pancreatitis, and in the referred study, it did not carry increase mortality or risk of pancreatic necrosis.\textsuperscript{5}

We found no difference in the need for surgical intervention patients with normal or elevated serum lipase levels. However, factors such as the nature and chronicity of the obstruction (acute vs subacute vs chronic and proximal vs distal) were not taken into account and further studies may be needed before this conclusion is made.

Conclusion

Small bowel obstruction should be considered in the differential diagnosis for elevated pancreatic enzymes. Abdominal imaging constitutes a useful resource to differentiate SBO from acute pancreatitis. In patients with SBO, elevated serum lipase is a risk factor for higher serum creatinine and lactate, use of ICU, and mortality.

References


Table 1. Patients’ baseline characteristics (n=344).

<table>
<thead>
<tr>
<th></th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>56 ± 15 years</td>
</tr>
<tr>
<td>Gender (male)</td>
<td>173 (50%)</td>
</tr>
<tr>
<td>Imaging supportive of pancreatitis</td>
<td>8 (2%)</td>
</tr>
<tr>
<td>Surgical resolution of SBO</td>
<td>141 (41%)</td>
</tr>
<tr>
<td>Known cause of SBO</td>
<td>229 (67%)</td>
</tr>
<tr>
<td>Serum creatinine &gt;1.5 mg/dL</td>
<td>73 (21%)</td>
</tr>
<tr>
<td>Patients with end-stage renal disease</td>
<td>7 (2%)</td>
</tr>
<tr>
<td>Patients with WBC&gt;15,000/µl</td>
<td>72 (21%)</td>
</tr>
<tr>
<td>Patients with lactate&gt;2 mmol/L</td>
<td>110 (32%)</td>
</tr>
<tr>
<td>Length of hospital stay (in days)</td>
<td>8.5 ± 8</td>
</tr>
<tr>
<td>Intensive care unit use</td>
<td>26 (8%)</td>
</tr>
<tr>
<td>Death during the same admission</td>
<td>14 (4%)</td>
</tr>
</tbody>
</table>

SBO: small bowel obstruction; WBC: white blood cells.
Table 2. Comparison of patients with SBO and normal or elevated serum lipase.

<table>
<thead>
<tr>
<th></th>
<th>With elevated lipase (n=34)</th>
<th>With normal lipase (n=302)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (mean ± SD) (in years)</td>
<td>53±15</td>
<td>54±15</td>
<td>0.76</td>
</tr>
<tr>
<td>Gender (Male)</td>
<td>20 (59%)</td>
<td>148 (49%)</td>
<td>0.37</td>
</tr>
<tr>
<td>Serum creatinine&gt;1.5 m/dL*</td>
<td>17 (50%)</td>
<td>55 (18%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Serum lactate&gt;2 mmol/L **</td>
<td>16 (47%)</td>
<td>89 (29%)</td>
<td>0.009</td>
</tr>
<tr>
<td>WBC count&gt;15,000/μL</td>
<td>7 (21%)</td>
<td>62 (21%)</td>
<td>1.00</td>
</tr>
<tr>
<td>Need for surgery</td>
<td>12 (35%)</td>
<td>127 (42%)</td>
<td>0.47</td>
</tr>
<tr>
<td>Length of stay (in days)</td>
<td>17±17</td>
<td>8±7</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Need for ICU care</td>
<td>11 (32%)</td>
<td>15 (5%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>In-hospital mortality</td>
<td>7 (21%)</td>
<td>7 (2%)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

* Patients with ESRD were excluded from this calculation
** 48 patients did not have a lactic acid level drawn
WBC: white blood cells; ICU: intensive care unit
Noncompaction Cardiomyopathy associated with Polycystic Kidney Disease: coincidence or genetic association?

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Abstract

Left ventricular noncompaction is a rare abnormality of cardiac development, which displays a broad spectrum of genetic penetrance, symptomatic severity and association with multiple congenital diseases. We describe a 35-year-old man who presented with a stroke associated with left ventricular dysfunction and chronic kidney disease.

Introduction

Autosomal dominant polycystic kidney (ADPKD) disease is a common hereditary disorder, characterized by the formation of multiple cysts in the kidneys, liver and other organs. The most common cardiovascular disorders associated with ADPKD include valvular abnormalities and aneurysm. We describe a 35-year-old man who presented with stroke and ADPKD. The patient was diagnosed with impaired left ventricular function resulting from noncompaction of the left ventricle (LVNC). There are few case reports in the literature that associate ADPKD and various cardiomyopathies, including LVNC, are all reported to involve mutations in sarcomere genes, suggesting a possible link between the conditions.

Case Presentation

A 35-year-old male with past medical history of hypertension, presented with sudden onset of left side weakness. Physical exam was positive for left side hemiparesis, left facial droop and aphasia. A magnetic resonance of the brain (MRI) showed right MCA ischemic stroke. The patient received intravenous tissue plasminogen activator (tPA) and stroke work up was started. A transthoracic echocardiogram (TTE) was performed reporting sinusoids and trabeculae two times the size of the underlying myocardium of the left ventricle, severely dilated left ventricle, global hypokinesis with an ejection fraction 20-25% and multiple liver cysts. Initial labs were remarkable for an increased creatinine of 2.0 mg/dl. The abdominal ultrasound was consistent with polycystic kidney disease and cardiac magnetic resonance (CMR) findings consistent with left ventricle noncompaction cardiomyopathy (Fig 1). Patient’s focal deficit improved during his hospital course and he finally was discharged home with optimal medical therapy.
Figure 1. Cardiac magnetic resonance image shows noncompaction of the left ventricular myocardium (arrows) and, in the enlarged left kidney and liver with multiple cystic changes (arrowheads).

Discussion

LVNC is a rare disease with a prevalence of 0.014% in the adult population. This pathology results from the intrauterine arrest of compaction of the loosely interwoven meshwork of the fetal myocardial primordium. Prominent left ventricular (LV) trabeculae, deep intertrabecular recesses, and a thin compacted layer is part of the main characteristics. LVNC may occur as an isolated trait or may be associated with autosomal dominant, X-linked, or
mitochondrial genetic mutations.\textsuperscript{1,2} Several genes have been associated, such as sarcomeric or cytoskeletal protein genes, including mutations in the Notch1 pathway and TAZ protein, that lead to dysregulated remodeling of cardiolipin.\textsuperscript{3}

The classical clinical presentation is a triad of heart failure, arrhythmias, and embolic events from mural thrombi. The subendocardial hypoperfusion restricted filling due to prominent trabeculae, and microcirculatory dysfunction is the main for ventricular dysfunction and arrhythmogenesis.\textsuperscript{2} The gold standard for diagnosis is cardiac imaging, and the most common imaging modalities include echocardiography and cardiac magnetic resonance (CMR). Most used criteria are the: Peterson's CMR criteria (non-compacted/compacted ratio > 2.3 in diastole on long axis images) and Jenni's echocardiographic criteria (non-compacted/compacted ratio > 2.0 at end-systole on short-axis images).\textsuperscript{2}

The management of LVNC is similar to other cardiomyopathies and includes appropriate heart failure treatments for left ventricular systolic dysfunction, appropriate management of arrhythmias, and long-term prophylactic anticoagulation for all patients regardless of whether they have experienced thromboembolic complications (prevalence of 21-25\%) and irrespective of the degree of left ventricular dysfunction.\textsuperscript{1,2}

On the other hand, we have ADPKD, which is a common disorder, responsible for 6-10\% of end stage renal diseases cases in the US. ADPKD is caused by mutations of either PKD1 (polycystin-1 on chromosome 16) or PKD2 (polycystin-2 on chromosome 4), most common being the first one. Those proteins are involved in adhesive protein-cell-matrix interactions, through the JAK-STAT pathway.\textsuperscript{4}

LVNC has been associated with multiple congenital diseases such as Barth syndrome, but only 3 cases in the literature have been associated with ADPKD, one of them within the same family. Akhtari et al. concluded no association between those two pathologies after they reviewed 126 CMR of patients with known ADPKD (but not cardiomyopathy) and found that none of the CMR met criteria for LVNC.\textsuperscript{5,6} Up to date, there are no studies overarching genetic causes between these two pathologies.

Occasionally LVNC is underreported, due to the fact that not all patients with ADPKD undergo cardiac imaging to exclude LVNC. CMR imaging or echocardiography could be useful in establishing the extent of association between LVNC and ADPKD. Autosomal polycystic kidney disease usually presents as a regional disease that is treated conservatively. However, high suspicion of associated cardiomyopathy might be warranted to prevent further life-threatening complications such as sudden cardiac death due to arrhythmias and thromboembolic events, by incorporating targeted cardioprotective therapy in early stages of the disease.
Conclusion

Left ventricular noncompaction is a rare abnormality of cardiac development, which displays a broad spectrum of genetic penetrance and symptomatic severity. The frequency of LVNC in association with PCKD might well be underreported; it certainly deserves further study. CMR imaging (gold standard) or echocardiography could be useful in establishing the extent of association between LVNC and ADPKD. Furthermore, overarching genetic causes should be also considered. Establishing association between these two diseases might affect clinical management and prevent further life threatening complications, such as sudden death and thromboembolic events.

References

Antiphospholipid Syndrome with Simultaneous Thrombotic and Hemorrhagic Manifestations Following Anticoagulation Therapy

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Abstract

Case Background: We present a case of a 43-year-old female with systemic lupus erythematosus, recently diagnosed with secondary antiphospholipid syndrome (APS) and acute ischemic strokes. She was started on anticoagulation and achieved therapeutic INR. Five days later, she presented complaining of worsening right calf pain and swelling, and was found to have acute drop in hemoglobin with a supratherapeutic INR. A hematoma within the soleus muscle was evident on computed tomography (CT) scan, while a Duplex ultrasound with Doppler revealed a concurrent new occlusive deep venous thrombosis of the right popliteal vein. Despite holding anticoagulation, her neurological status rapidly declined and a CT head without contrast evidenced hemorrhagic conversion of her previous ischemic lesions resulting in significant brainstem compression, upward herniation and acute hydrocephalus. Emergent decompressive suboccipital craniectomy and evacuation of the bilateral cerebellar hematomas was performed; unfortunately, the patient deceased soon after.

Discussion: APS is characterized by the occurrence of vascular thrombosis in the presence of antiphospholipid antibodies. In these patients, hemorrhage is a less frequent complication than thrombosis. Furthermore, simultaneous bleeding and thrombosis is a rare event, usually reported in the setting of catastrophic APS or thrombocytopenia, none of which our patient had.

Key words: Antiphospholipid syndrome, Systemic lupus erythematosus, Anticoagulation, Thrombosis, and Intracranial bleeding.

Introduction
Antiphospholipid syndrome (APS) is a multisystem autoimmune disease characterized by arterial or venous thrombosis and/or pregnancy morbidity due to antiphospholipid antibodies (aPL). The mainstay of treatment includes antithrombotic agents, with a target international normalized ratio (INR) of 2.0-3.0.\(^1,2\) Major bleeding is unusual and occurs in 10\% of patients with APS, moreover simultaneous bleeding and thrombosis in APS is a rare event.\(^3\)

**Case**

A 43-year-old female with history of systemic lupus erythematosus (SLE), diagnosed over 10 years ago, off treatment, acute ischemic strokes involving multiple vascular territories (left frontal lobe, right occipital lobe and both cerebellar hemispheres) two weeks prior to presentation. At that time, studies were positive for lupus anticoagulant, IgM anti-cardiolipin, and IgM anti-beta2-glycoprotein; therefore, the diagnosis of secondary APS was made. She was started on anticoagulation with enoxaparin and warfarin and achieved an INR of 2.0 at the time of discharge. Workup for CNS vasculitis including lumbar puncture during the admission was negative. She was discharged to an inpatient rehabilitation facility, where Aspirin 325 mg daily was added to her regimen. Her other medications included hydroxychloroquine 200 mg, prednisone 30 mg and mycophenolate 500 mg BID.

Five days after discharge, she was readmitted for worsening right calf pain and swelling. Her hemoglobin was 5.9 g/dL (decreased from 8.9 g/dL prior to discharge), her INR was 3.3 and PTT was 41.7 seconds. A computed tomography (CT) of her lower extremity evidenced a large hematoma within the soleus muscle. A Duplex ultrasound with Doppler revealed a concurrent new occlusive deep venous thrombosis of the right popliteal vein. Anticoagulation was held but no vitamin K or fresh frozen plasma (FFP) were given. She was alert and answering questions appropriately, but 24 hours later her neurological status rapidly declined. In a matter of minutes, she became unresponsive, tachycardic and apneic and was transferred to the intensive care unit followed by intubation for airway protection. A CT of the head without contrast showed hemorrhagic conversion of her previous posterior fossa ischemic lesions, with significant brainstem compression, upward herniation and acute hydrocephalus. Packed red blood cells and FFP were given and she underwent emergent ventriculostomy, and then, decompressive suboccipital craniectomy with evacuation of the bilateral cerebellar hematomas. Her neurological status progressively declined, and the patient deceased soon after.
Figure 1. Panel A: Initial computed tomography scan of the head without contrast showing multiple areas of hypodensity in multiple vascular territories, including both cerebellar hemispheres without hemorrhagic conversion. Panel B: Large acute cerebellar hematoma in the region of the prior bilateral posterior circulation infarcts.

Figure 2. Simultaneous thrombosis and bleeding of the right leg. Panels A and B: Right popliteal vein is not compressible (A) and has no color Doppler flow (B), confirming the Diagnosis of an occlusive deep venous thrombosis. Panels C and D: computed tomography scan showing a large (12.6 x 5 x 4.3 cm) hematoma within the soleus muscle.
Discussion

Around 40% of patients with SLE have aPL, but less than 40% of them will develop a thrombotic event. Major bleeding is unusual, and occurs in 10% of patients with APS due to capillaritis, microthrombosis, antiprothrombin antibodies, thrombocytopenia, and/or excessive antithrombotic therapy. In most cases reported in the literature, this happened in the setting of catastrophic APS or thrombocytopenia. Catastrophic antiphospholipid syndrome (CAPS) represents the end of the spectrum of APS, characterized by widespread small vessel thrombosis resulting in prompt multi-organ failure. While a concern for CAPS existed in this patient, she did not meet the diagnostic criteria. Furthermore, she had a rethrombosis despite being on anti-platelet and anticoagulation therapy. Recurrences are fairly common and have been reported to be present in 9.1 cases per 100 patient-years. Noteworthy, most recurrences take place in the same vascular bed as the original thrombosis. Even though most experts recommend a target INR of 2.0-3.0, other authors have suggested a higher intensity regimen (INR 3.0-4.0), which has not proven to be more effective in preventing re-thrombosis but is often associated with greater bleeding rates (threefold increased risk for minor bleedings).

We suspect that the INR of 3.3 on admission was one of the factors that likely contributed to our patient’s fatal bleeding. Although anticoagulation was held, she did not initially receive reversal agents (vitamin K and/or FFP) due to a concomitant new thrombotic event. Once the patient acutely decompensated, with concerns for intracranial bleeding, both interventions were given. Management of bleeding in APS is extremely difficult due to baseline increased risk of thrombosis. This is even more problematic in the light of an acute simultaneous thrombotic event. Decision to stop anticoagulation must be made based on each patient’s risks and benefits. Most of the times, clinicians face the dilemma of starting anticoagulation, which may worsen the bleeding versus holding anticoagulation, which may cause further thrombosis. Unfortunately, the form of anticoagulation (heparin, warfarin, etc.), as well as the dose and time to restart them remains unclear and are usually based on clinical expertise. More studies are needed in order to establish a safer anticoagulation regimen for these patients.

References

Pancreatic Mucinous Adenocarcinoma Mimicking Walled-off Necrosis

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Abstract

Malignant cystic tumors of the pancreas are a rare entity. We present a case of a 69 year-old man with a large mucinous adenocarcinoma of the pancreas that presented with initial imaging studies suggesting walled-off pancreatic necrosis. On cyst biopsy, the patient’s malignancy was confirmed. Sensitivity of radiological imaging in differentiating cystic pancreatic lesions is limited. Cyst histology and, often biopsy of the cyst wall, is necessary for diagnosis and treatment.

Introduction

Mucin-producing cystic neoplasms of the pancreas is well-recognized entity, diagnosed in approximately 10%-20% of resected pancreatectomy specimens. Most of the lesions displayed low-grade dysplasia and involve primarily the main pancreatic duct with or without extension to the branches. Mucinous neoplasms can be further sub-classified into main-duct type (20%), mixed-type (40%) and branch-duct type (40%).

Case

A 69-year-old Hispanic man with past medical history of diabetes type 2, hypertension, dementia and gallstone pancreatitis presented to our institution with a 2-month history of worsening epigastric abdominal pain. The pain was intermittent, 7/10 in intensity, radiating to the back and associated with nausea and 20-pound weight loss. Physical examination was remarkable for epigastric abdominal tenderness. Initial laboratories studies showed alkaline phosphatase 2061 U/L, AST 519 U/L, ALT 990 U/L, total bilirubin 1.9 mg/dl, lipase 37 U/L, CA 19-9 339 U/ml. Abdominal imaging with computerized tomography (CT) revealed necrotizing pancreatitis with described walled-off necrosis. Due to an elevated CA 19-9 level, patient underwent endoscopic ultrasound-guided biopsy with fine needle aspiration. Fluid collected from the cyst was viscous and final pathology report was significant for Mucinous Adenocarcinoma (MADC) of the pancreas with signed cell features.
Figure 1. Magnetic Resonance Imaging (MRI) abdomen and pelvis. Large complex collection replaces the entire pancreas compatible with sequela of necrotizing pancreatitis with walled off pancreatic necrosis.
Discussion

Mucinous cystic neoplasms (MCNs) of the pancreas are comparatively rare. The differential diagnosis of cystic lesions of the pancreas is rather broad; including benign (mucinous cystadenomas and intraductal papillary mucinous adenomas), inflammatory (pseudocyst and retention cyst), infectious (primary hydatid disease) or malignant (MADC and cystic lymph node metastasis). Imaging modalities can be useful in differentiating the etiology.

On CT imaging, MADC tends to appear as round, externally smooth, near-water-density cystic lesion. In contrast, walled-off pancreatic necrosis, is more commonly found as a cystic lesion around or within the pancreas, with areas of non-enhancing tissue representing necrosis, surrounded by a wall.2,3

One retrospective study that evaluated the histological features of pancreatic cystic lesions, reported that 1 out of 7 cases of MCNs had an invasive carcinoma component within it.4 Histopathological examination remains the goal standard for diagnosis of these lesions, especially in the setting of concerning features such as advanced age and high levels of tumor markers, such as in our patient.

Accurate diagnosis is essential, as treatment depends on the tumor histology. When localized to the pancreas alone, noninvasive and invasive MADC have a favorable prognosis after surgical resection.5 However, patients with unresectable disease have been reported to have a 2-year survival of approximately 12%; therefore, cognizance of the malignant potential of cystic lesions can aid in early diagnosis, ultimately leading to improved survival outcomes.

References

Phaeohyphomycosis

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Abstract

Background: Phaeohyphomycosis is a chronic infectious condition caused by opportunistic melanized fungi. These infections are seen particularly in immunocompromised patients, especially following organ transplant, and can spread systemically without prompt treatment.

Case Report: Here, we discuss a case of an 84-year-old man with B. mackinnonii cutaneous phaeohyphomycosis after a brief course of oral prednisone, presenting as a pruritic, scaly rash on the forearm. A skin biopsy confirmed the diagnosis, and the patient was treated with itraconazole with rapid resolution of symptoms.

Discussion: Until recently, Biatriospora mackinnonii had only been implicated in two reports of phaeohyphomycosis in renal transplant patients. We explore risk factors for this infection in this otherwise immune competent patient.

Introduction

While disseminated phaeohyphomycosis occasionally affects the central nervous system and the lungs, subcutaneous phaeohyphomycosis is the most commonly reported form of the disease, and is characterized by an array of physical findings including papulonodules, cysts, abscesses, pyogranulomas, non-healing ulcers, and verrucous, hyperkeratotic and ulcerated plaques.1 These infections are seen particularly in immunocompromised patients, especially those on immunosuppressive therapy following organ transplant. Infection typically occurs as a result of traumatic inoculation of dematiaceous fungi into the subcutaneous tissue. Here, we report a case of cutaneous phaeohyphomycosis caused by Biatriospora mackinnonii in an immunocompetent patient.

Case

An 84-year-old man presented to a primary care clinic with a skin lesion located on the right dorsolateral forearm. There was no history of trauma or recent infection. The patient was retired, had no outdoor hobbies, no new medications or personal care products, and no contacts with similar findings. At his presenting visit, the patient was found to have been on a two-month prednisone taper from 60 mg to 5 mg for pulmonary symptoms. The lesion appeared two weeks earlier as a small erythematous patch on the right dorsolateral forearm, which progressed into a painful, scaly 15 cm x 8 cm plaque that expressed purulent material (Figures 1A & 1B). The first evoked diagnosis was cutaneous candidiasis and the patient was treated with Ketoconazole 2% topical cream and Fluconazole 200 mg
tablets for 14 days without improvement. A punch biopsy was performed and specimen underwent Hematoxylin and Eosin and Gomori methenamine silver staining for microorganisms. Microscopic examination revealed granulomatous dermal inflammation and multiple pigmented hyphal forms (Figure 2). Direct examination in 20% KOH showed melanized branching hyphae with rare pigmented yeast forms. Histologic examination of a second punch biopsy after Gomori methenamine silver staining revealed pigmented fungal forms with septate hyphae, confirming the diagnosis of phaeohyphomycosis (Figure 3). The mold was identified as *Biatriospora mackinnonii* on culture. The patient was then treated with Itraconazole 200 mg twice daily and saturated solution of potassium iodide 10 drops three times daily with instructions to increase by 1 drop per day as tolerated. Over the following two weeks, the patient showed remarkable response, with full resolution twelve weeks after the therapy started.

Figure 1A. Erythematous, 15 cm x 8 cm plaque on dorsolateral forearm, characteristic of cutaneous phaeohyphomycosis.

Figure 1B. Lesion visualized under dermatoscopy.

Figure 2. Punch biopsy of plaque revealing dermal inflammation and pigmented fungal forms (H&E, original magnification x200).
Figure 3. Second punch biopsy indicating septate hyphae within the dermis (GMS, original magnification x400).
Discussion

Phaeohyphomycosis is an umbrella term for a heterogeneous group of opportunistic fungal infections. Increasingly biodiverse species are being identified as causative pathogens in both immunocompromised and immunocompetent individuals. It is important to
accurately speciate offending pathogens, as different species may have tropism for
different organs and varying susceptibilities to antifungals.\textsuperscript{2} To our knowledge, \textit{B}
\textit{mackinnonii} has only been implicated in 2 reported cases of phaeohyphomycosis.\textsuperscript{2,3} This is
the first reported case of \textit{B mackinnonii} phaeohyphomycosis in a non-transplant patient.
Until recently, many reported cases of phaeohyphomycosis did not identify the causative
species because the criterion for diagnosis is based solely on the histologic finding of
dematiaceous mycelia. Thus, the true prevalence of this and other implicated species may
be understated based on scarcity and unreliability of strain identification methods.\textsuperscript{3}

We believe this immunocompetent patient suffered this unusual infection due to the
presence of two risk factors: corticosteroid use and advanced age.

Corticosteroids like prednisone temporarily blunt the immune system and are a potential
risk factor for the development of opportunistic infections.\textsuperscript{4} A large meta-analysis of 71
clinical trials found that a daily dose of \(\geq 10\) mg or a cumulative dose of \(\geq 700\) mg of
prednisone was associated with significantly higher infection rates when compared with
control subjects.\textsuperscript{5} While the exact cumulative dose in our patient was not known, the
schedule does appear to have led to a total dose eclipsing 700 mg. That being said, this
particular species has only been implicated in transplant patients up until this point. Given
the brevity of the prednisone treatment and the low dose of 5 mg/day at presentation, we
believe that the exploration of other possible risk factors, like the patient’s advanced age, is
warranted.

Aging is a complex process that affects every organ system. The immune system gradually
deteriorates with natural age advancement, a process often referred to as
immunosenescence. Chronic involution of the thymus gland is one of the primary
contributing factors to this decline. Recent studies suggest that this is indeed an active
process. As the body ages, cytokines induce atrophy of the thymopoietic space.\textsuperscript{6} The
thymus is responsible for the production of naive T cells, which are vitally important in the
mechanism of cell-mediated immunity. This arm of the immune system is the primary
defense against fungal infections, so it reasonably follows that thymic atrophy could have
predisposed our patient and others with his degree of immunosenescence to these
pathogens without any significant external immune suppressors.\textsuperscript{7}

Perhaps in part due to its rarity, no treatment guideline currently exists for
phaeohyphomycosis. Our patient was treated with itraconazole for two weeks, and rapidly
experienced near-complete remission. A 19-patient case series of phaeohyphomycosis
indicated a nearly 50\% success rate with itraconazole therapy, including several patients
who had already failed therapy with other azoles and amphotericin.\textsuperscript{8}

New microbiological techniques may increase our ability to recognize \textit{B mackinnonii}
infection, therefore understanding further its virulence as well as broader opportunities for
treatment.
References


Balanitis Circumscripta Plasmacellularis Associated with Acyclovir Resistant-HSV2 in an Immunocompromised Host

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Abstract

Balanitis Circumscripta Plasmacellularis is an idiopathic and benign condition that can mimic the appearance of multiple infectious diseases, leading to misdiagnosis and invasive or unnecessary procedures. We present a case of balanitis circumscripta plasmacellularis in a host with HSV-2.

Introduction

Balanitis Circumscripta Plasmacellularis is a cutaneous benign condition characterized histologically by plasma cell infiltration of the dermis leading to epidermal atrophy. The lesions are often associated with chronic infections and trauma, commonly manifesting in the middle age or elderly and uncircumcised adult. There is scarce data on the incidence and prevalence of Balanitis Circumscripta, mostly derived from case series of patients with penile lesions. Despite being a benign condition, rarely it can present in association with malignancies. Balanitis Circumscripta should be taken into consideration in the differential diagnosis of patients presenting with refractory diseases, such as herpes simplex virus (HSV) and human papillomavirus (HPV).

Case

A 50 year-old male with past medical history of Human Immunodeficiency Virus (HIV) and recurrent genital herpes simplex type 2 (HSV-2), presented with 1-month history of worsening genital ulcers associated with bleeding. The patient had been on antiretroviral therapy for over 17 years and was also on acyclovir for recurrent HSV-2 for the past 4-months with partial improvement of symptoms. Initial laboratory data reported a CD4 of 336/ul and viral load < 20 copies/mL. Physical examination was remarkable for erythematous plaques on the ventral aspect of the penis associated with areas of bleeding surrounded by small perilesional ulcers. Biopsy and HSV-2 polymerase chain reaction (PCR) were performed, reporting Balanitis Circumscripta Plasmacellularis and HSV-2, respectively. Patient was subsequently started on cidofovir for acyclovir-resistant disease. Two weeks after initiation of treatment and post circumcision, the patient’s symptoms improved with minimal residual rash and decreased pain.
Discussion

Balanitis Circumscripta Plasmacellularis is an idiopathic, benign condition that usually presents as a solitary, persistent plaque on the glans of uncircumcised, middle-aged or elderly men. Histopathology examination with skin biopsy is the goal standard for diagnosis. Atrophy of the epidermis, band-like infiltrate of plasma cells in the upper dermis, absence of atypia and erythroplasia are the main characteristic histopathologic findings. Its etiology and pathogenesis remain unclear but plasma cell infiltration is likely a result of non-specific inflammatory response to an unknown exogenous agent, such as chronic infections with non-tuberculous Mycobacterium and HPV. Multiple skin disorders can mimic the appearance of this condition leading to misdiagnosis and invasive or unnecessary procedures. Among the differential diagnosis candidiasis, lichen planus, syphilis, HSV, erythroplasia of Queyrat and Bowen’s disease are the most likely to be confused with Balanitis Circumscripta. The diagnosis should be suspected particularly in those with lesions that do not respond to empiric therapy and in those who have chronic and/or refractory sexually transmitted diseases. The clinical course as mentioned above is chronic and often does not respond to topical treatment alone. In refractory cases surgical management with circumcision can be curative and may even be considered as initial therapy.

References

Pyogenic Liver Abscess: an insidious manifestation caused by Streptococcus

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Abstract:

Background: Pyogenic liver abscess, a potentially life-threatening disease, has undergone significant changes in epidemiology, management, and mortality over the past several decades. This case presents the insidious manifestation of a pyogenic liver abscess in a 72-year-old male.

Case Report: 72-year-old male who presented to the ED for fever, malaise, weakness, poor appetite, and abdominal pain for 3 weeks. He denies any nausea, vomiting, diarrhea, or hematochezia. He continued to have persistent abdominal pain described as stabbing and constant. Patient endorses fever, night sweats, 6 lbs weight loss in last month.

Discussion: What was interesting regarding this case was the absence of the classic manifestations of liver abscess. Due to the patient’s history of liver cirrhosis relative immunosuppression may be the cause. Clinicians should be vigilant in order to establish the right diagnosis and give the correct treatment. The absence of fever, leukocytosis or liver function abnormalities can conceal the presence of pyogenic liver abscess. Further evaluation with blood and abscess culture are crucial.

Keywords: Liver Abscess, Abdominal Pain, Group A Streptococcus

Introduction

Pyogenic liver abscess was initially reported in the writings of Hippocrates, who based prognosis of his patients on the type of fluid recovered from the abscess.\(^1\) In 1938, Debacke and Ochner described the treatment and mortality of patients with pyogenic liver abscess and recommended surgical treatment as the primary treatment modality.\(^2\) At that time, pyogenic liver abscess occurred primarily as a complication of acute appendicitis, predominately in young men, and was associated with a high mortality rate.\(^3\) Based on the recommendations of Debacke and Ochner and no viable alternatives, surgery remained the preferred therapy until the 1990s when percutaneous drainage was shown to be a safer and less invasive alternative.\(^4\) Since that time, mortality rates have decreased substantially with recent studies reporting rates between 11%-31%.\(^3\) The most common cause reportedly has shifted to biliary disease.\(^3\) Despite improving trends in mortality, pyogenic hepatic abscesses are challenging conditions due to an unspecific patient presentation, manifold differential diagnosis and no common consensus on the best therapeutic approach with respect to antibiotic treatment alone versus addition of drainage. An interesting case of an afebrile liver abscess is hereby presented caused by Group A Streptococcus without liver function abnormalities, fever or leukocytosis.
Case Report

A 72-year-old Hispanic male with a past medical history significant for diabetes mellitus type 2, alcoholic cirrhosis, pancreatitis and pancreatic pseudocyst presented to the emergency department with a three-week history of fever, malaise, weakness, poor appetite, and abdominal pain. The patient described the abdominal pain as stabbing and constant without any alleviating or aggravating factors primarily in the epigastric region. Past surgical history was significant for an uncomplicated cholecystectomy ten years ago. The patient reported no nausea, vomiting, constipation, diarrhea, or hematochezia. In addition, the patient reported one week of night sweats and an unintentional six-pound weight loss over the last month. The patient had no significant family history, no known allergies, and lived at home with his wife and son. He denied recent travel. The patient reported drinking alcohol several years ago but had since quit and denied smoking or illicit substance abuse.

The patient was found to be afebrile and normotensive without tachycardia. Physical exam demonstrated an abdomen that was soft, mildly distended and tender to palpation in the epigastric region. No hepatosplenomegaly was noted and there were no palpable masses. Laboratory examination including complete blood count, liver function tests, and electrolytes were within normal limits. Chest radiograph demonstrated no acute cardiopulmonary abnormality.

Due to the patient’s history of pancreatitis computed tomography with contrast was obtained which demonstrated numerous hypodense lesions throughout the liver, with the largest measuring 3.6 cm in the left hepatic lobe (figure 1). There was a hyper-dense mass at the pancreatic neck measuring 4.1 cm with adjacent peripancreatic lymph nodes. Fluid tracks were noted around the pancreas and liver. Gastroenterology was consulted and an EUS with FNA biopsy was obtained of one of the representative lesions due to concern for possible metastatic disease. Pathology of the liver lesion revealed findings consistent with a pyogenic liver abscess. Cultures of the lesion grew Group A streptococcus, and blood cultures grew the same species. Due to the size and number of abscess on computed tomography surgical or percutaneous debridement was deferred. The patient was administered intravenous ceftriaxone and metronidazole.

The patient’s clinical status rapidly improved though he continued to have intermittent low grade fevers for the next three days. Repeat blood cultures were negative. Transesophageal echocardiogram was negative for vegetations. Infectious disease was consulted and due to the size of the various cysts they recommended that the patient received a two week course of intravenous ceftriaxone. The patient had a PICC line placed and was discharged home on day five of hospital admission with a plan to follow up in clinic. The patient was then transitioned to oral levofloxacin and metronidazole for an additional two weeks of antibiotics. Repeat computed tomography at four weeks post discharge demonstrated resolution of the liver lesions.
Figure 1. Abdominal CT scan with intravenous contrast demonstrates numerous hypodense lesions throughout the liver, with the largest measuring 3.6 cm in the left hepatic lobe. Imaging findings were primarily suggestive of an abscess. Another possibility would be metastatic tumors with central necrosis.

**Discussion**

Liver abscesses are the most common type of visceral abscess, accounting for forty eight percent in recent reports. Various pathogens have been reported as the primary cause, but are usually poly-microbial, consisting of a mixture of enteric facultative and anaerobic pathogens. Diagnosis is made from the patient’s history, clinical examination and imaging followed by aspiration and culture of the abscess. Typical clinical manifestations of liver abscess include fever (90%) and abdominal pain (50–75%). In addition, white blood cells (WBCs) and liver function tests are usually increased.

According to the current literature review, however, a minority of patients may present without fever (13.9–31%). In addition, a small subset have a normal WBC count (23–24.6%) and liver function tests (13–29.5%) at the time of diagnosis. In this case, the patient was admitted afebrile with normal liver enzymes and no leukocytosis. The reason this patient failed to present with typical findings of liver abscess is unknown but the patients history of liver cirrhosis may be postulated to be the cause.
Liver abscesses are the cause of major complications including sepsis and multi-organ dysfunction and can lead to death if left untreated. Antibiotics and appropriate drainage are the standard of practice. Typically patients present with a solitary lesion making drainage more practical. Definitive evidence based guidelines have yet to be developed on the treatment of patients who present with numerous small liver abscesses as they are often not good candidates for percutaneous or surgical drainage.

Microorganisms associated with liver abscesses include most commonly gram-negative enteric bacteria (Escherichia coli, K. pneumoniae, Pseudomonas, and Proteus), gram-positive aerobes (Streptococcus milleri, Enterococcus, Staphylococcus aureus, Staphylococcus epidermidis and Streptococci sp.), anaerobic organisms (Bacteroides sp. and Fusobacterium), actinomyces, Candida albicans, Salmonella typhi, Brucella melitensis or other protozoa (Entamoeba histolytica and Echinococcus granulosus). The microbe isolated from the liver abscess is related to its origin. Since the liver receives blood from both systemic and portal circulation, the liver is more prone to infections from biliary and gastrointestinal tract bacteria.

In this case, the abscess and blood cultures isolated a bacterium frequently found in the oral cavity and part of the skin normal flora. Group A streptococcus (GAS) is a group of gram positive bacteria that are aero-tolerant, no motile and non-spore forming cocci. It is one of the most common kinds of microorganism causing a range of human infections with the most common of which is pharyngitis in children. This group of micro-organism’s is also prone to developing liver abscesses due to liver tropism and the hydrolytic enzymes such as intermedilysin. Speculation about the source of the infection in this case included origination from the oral cavity, as the patient exhibited poor oral hygiene with the presence of dental plaques but origin remains a mystery.

**Conclusion**

What was interesting regarding this case was the absence of the classic manifestations of liver abscess. Due to the patient’s history of liver cirrhosis relative immunosuppression may be the cause. Clinicians should be vigilant in order to establish the right diagnosis and give the correct treatment. The absence of fever, leukocytosis or liver function abnormalities can conceal the presence of pyogenic liver abscess. Further evaluation with blood and abscess culture are crucial.

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I would like to thank the patient for his consent to publish the case and related pictures.

**References**

Progression and Resolution of Varicella Pneumonia on Serial Chest Radiographs

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Abstract

We present a patient with varicella pneumonitis and the chest imaging throughout the course of disease. Chest radiographic changes of Varicella pneumonia typically consist of poorly-defined nodules. The patient’s image is presented to describe the rate of respiratory compromise and subsequent improvement after treatment with intravenous acyclovir and controversial use of corticosteroids.

Case Presentation

The patient is a 37-year-old woman with no significant medical history who presented to the emergency department with rash and shortness of breath after the appearance of a significant pustular rash three days prior. On initial evaluation, she reported mild dyspnea and the initial chest radiograph (day 1) was obtained. Patient was discharged home from the ED but returned the following day after clinical decline. She was admitted due to progression of the rash and diffuse pulmonary rales. Chest imaging was concerning for Varicella Pneumonia (day 2), and she was initiated on intravenous acyclovir and corticosteroids. On the evening of admission, patient had escalating oxygen requirements, progressing from stable vital signs on room air to requiring 6L nasal cannula within 12 hours. Chest x-ray on day 3 was notable for increasing interstitial infiltrates bilaterally with diffuse nodularity, characteristic of varicella pneumonitis. Patient was subsequently transferred to the ICU due to respiratory decline. By the following morning (day 4), the patient’s oxygen requirement had progressed to high velocity nasal insufflation at 40 L/minute with 80% oxygen saturation. She showed clinical improvement in both her respiratory and dermatologic conditions on day 5 and was weaned to 8L high flow nasal cannula. During her hospitalization, a skin biopsy was performed, confirming the diagnosis of Varicella. Patient did not recall prior episode of Varicella nor immunization.

Figure 1. Sequential Progression of Radiographic Changes in a Patient with Confirmed Varicella
Figure 1. (cont.) Sequential Progression of Radiographic Changes in a Patient with Confirmed Varicella

Discussion

Varicella pneumonia typically presents one to six days after the appearance of the characteristic rash with symptoms of tachypnea, dyspnea, or chest tightness.\(^1\) Initial physical symptoms and radiographic signs are poor predictors for subsequent need for artificial ventilation.\(^1\) Regardless, varicella pneumonia may progress rapidly to fulminant respiratory failure despite maximum conventional support, including IV acyclovir and IV corticosteroids.\(^1\) Corticosteroids are used adjunctively and have been shown to reduce hospital and ICU length of stay but without mortality benefit.\(^2\) Many arguments have been
made for the use of extracorporeal membrane oxygenation/life support in these patients with studies showing up to 60% survival.¹

Triebwasser et al. ³ were the first to report the radiographic changes in varicella pneumonia with seven adult cases at their institution and a review of another 246 previously reported cases. Of the seven cases, some were not intentionally imaged with the intent to identify nodules. While the radiographic changes and severity of disease varied among the cases, one patient exhibited classical bilateral nodular infiltrates on radiographic exam and required intermittent positive pressure ventilation. Many subsequent cases with these findings have since been documented.

Chest radiographic changes of Varicella pneumonia typically consist of many 5-10 mm poorly-defined nodules, rarely associated with hilar lymphadenopathy and pleural effusion.⁴ These lesions may resolve within one week of the disappearance of the skin lesions, but can persist for many months.⁴ Additionally, the lesions may calcify and persist as numerous random and well-defined 2-3 mm dense calcifications.⁴

Due to the high mortality of disseminated varicella zoster infection, sometimes documented up to 50%, it is important to quickly identify risk factors for pulmonary involvement for optimal intervention. Chest radiograph with such lesions may be critical in establishing the correct route of treatment and care for the adult with varicella zoster.

References

Systemic Lupus Erythematosis is a systemic disease imparting effects on many organ systems including the spleen. A varied array of presentations involving the spleen including splenomegaly, infarction and splenic rupture have been described. However, there have been few reports of incidentally found diffuse splenic calcifications in lupus patients. An underlying association with patterns of splenic calcifications and connective tissue disorders remains to be clarified.

Introduction

Splenic calcifications have been reported in several systemic disease states including rheumatoid arthritis, systemic sclerosis, sickle cell anemia, histoplasmosis, tuberculosis, brucellosis, and pneumocystis jiroveci. Patterns have been described as “punctate”, “centrally calcified abscesses”, and “singular” although no reports mention any pattern specific to a disease. In reports of SLE and calcifications they were noted as “diffuse” thereby a potential indicator of disease or at least a novel association.

Case Presentation

A 69 year-old female with known systemic lupus presented to Texas Medical Center with refractory respiratory failure owing to lupus pneumonitis requiring Venous-Venous Extracorporeal Mechanical Oxygenation. On plain film imaging of her abdomen for nasogastric tube insertion, a peculiar finding was noted. The spleen appeared diffusely and densely populated with calcified nodules, outlining the entire organ. No extra-splenic calcifications were noted. Her spleen did not appear enlarged on exam or imaging and no infection was detected on admission. Her lupus presentation was characterized by acute renal failure from anti-dsDNA positive lupus nephritis requiring continuous dialysis, normal levels of complement, an ARDS-like pneumonitis, coagulopathy and pancytopenia. Her course was further complicated by coagulopathy and arterial vessel injury during tracheostomy placement. She died later on in the course from brain hemorrhage.
Figure 1.

Discussion

SLE has effects on many organ systems including the spleen. Splenomegaly, splenic infarction as well as spontaneous splenic rupture have been described. Few reports describe, “diffuse calcifications” on radiographic imaging without any association with functional state of the spleen. Other disease entities also may involve the spleen via calcifications seen on imaging however no other disease states have characteristic diffuse patterns of calcification as seen in this image, yet few small studies with SLE associated calcifications do show a strikingly similar pattern. Although no pathophysiological explanation has been fully characterized, one report noted gross autopsy exam findings of evenly distributed spherical nodules 1-3mm in diameter.\(^2\) Our patient presented with an active lupus flair characterized by lupus nephritis, pneumonitis, coagulopathy and cytopenias. Infectious workup was negative including blood, urine and respiratory cultures. Clinical history, lab data and radiographic imaging ruled out other known causes of splenic calcifications.

References