



Case Report

A Sequela of Gastrointestinal Perforation Due to Small-Medium Vessel Vasculitis

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Abstract

Background: Systemic vasculitis resulting in inflammation of vessel walls may progress to ischemic damage of major dependent organs, including the gastrointestinal system. Polyarteritis nodosa (PAN) is a vasculitis that is known to present with necrotizing vasculitis in small-medium arteries, leading to life-threatening injury and surgical emergencies. We report the case of a young patient who presented with an acute abdomen and perforation of the small intestine confirmed to be caused by a small-medium vessel vasculitis with PAN as the leading diagnosis.

Case Presentation: A 27-year-old African American male with a past medical history of Stage 4 CKD and uncontrolled hypertension presented to the ED complaining of generalized abdominal and bilateral flank pain. He was found to have pneumoperitoneum on abdominal imaging requiring an emergent exploratory laparotomy found to have multiple microperforations of the small bowel requiring 110 cm of small bowel resection with primary anastomosis.

Conclusion: This case reports an unusual presentation of advanced vasculitis, possibly polyarteritis nodosa that resulted in acute abdomen requiring emergency bowel resection. Patients with multisystem involvement of vasculitis necessitate comprehensive work up for early diagnosis and management of the underlying etiology to improve outcomes and avoid significant morbidity/mortality.

Case Presentation

A 27-year-old African American male presented to the emergency room for acute on chronic generalized abdominal and bilateral flank pain and non-bloody, non-bilious emesis. Past medical history is significant for poorly controlled hypertension and associated chronic kidney disease. Patient had previously received dialysis but self-discontinued and was non-compliant with medications. He also reported a 20-pound weight loss over the past three months, attributed to his chronic abdominal pain and poor oral intake. Physical exam revealed a young man in moderate distress. He was afebrile in hypertensive crisis with systolic blood pressure in the 200s and acute kidney injury on laboratory results. The abdominal exam was notable for diffuse tenderness to palpation in all quadrants without peritonitis.

Initial x-ray imaging showed dilated loops of small bowel with air fluid levels without free air. Given the patient's presentation of bowel obstruction, patient was initially managed nonoperatively with NGT decompression, bowel rest and serial abdominal exams. However, he remained in the emergency department, his abdominal exam acute worsened prompting a repeat x-ray which showed subdiaphragmatic free air (Figure 1). An emergent exploratory laparotomy was performed with findings of purulent ascites upon entry and multiple punctate microperforations of the small bowel with inflammation of the distal branches of the mesenteric vessels (Figure 2). The decision was made to resect 110cm of small bowel, and given the patient's hemodynamic stability and minimal feculent peritonitis, a primary anastomosis was completed without complications. He was transferred to the ICU on ventilatory support and continued on broad spectrum antibiotics

as well as strict blood pressure control. Patient was extubated on POD2 and his postop course remained uncomplicated from the surgical perspective. Further inpatient work-up with cardiology, nephrology, rheumatology, and endocrinology was recommended for a definitive diagnosis prior to discharge; however, the patient left against medical advice on POD9 with ongoing noncompliance to oral hypertensive medications. Patient did not return for his postoperative follow up appointment and upon multiple attempted contacts to the patient, a patient's family informed the team that the patient had presented to an outside hospital with massive hematemesis in hemorrhagic shock ultimately resulting in his death. Unfortunately, medical records from the outside hospital was unable to be obtained for confirmation of the encounter.



Figure 1: XR abdomen AP upright showing multiple dilated loops of small bowel with air-fluid levels. There is subdiaphragmatic free air.



Figure 2: Photo of dissected bowel, measuring ~110cm. Areas of bowel perforation are evident.

Subsequently, pathology of the resected small bowel revealed findings consistent with small and medium vessel vasculitis, including occlusion of arteries with intimal thickening and transmural inflammation with fibrinoid necrosis. Serology was negative for SPEP, UPEP, ANA, ANCA, PR3, MPO, anti-GBM, and anti-PLA2R antibodies. Upon review of his rheumatology follow up documentation, no additional diagnostic work up was being pursued given negative serology tests.

Discussion

Several differentials were considered in this patient who presented with an acute abdomen found to have spontaneous multifocal small-bowel perforations in the setting of malignant hypertension and CKD. No IgA deposits on a recent colonic resection biopsy and lack of IgG4 plasma cells and anti-GBM antibodies suggested that IgA vasculitis, IgG4 related diseases, and Goodpasture's were unlikely causes. Our working differential was possibly polyarteritis nodosa (PAN), a small-medium vessel vasculitis that affects renal and visceral vasculature, resulting in tubulointerstitial nephritis and progressive kidney failure. In PAN, vascular lesions are typically segmental and occur in branch points with absent giant cells and granulomas, which is reflected in the patient's small bowel pathology [1]. The ACR criteria for diagnosing PAN are: 1) mandatory histopathological evidence of necrotizing vasculitis in medium- or small-sized arteries or angiographic abnormality; 2) plus one of the following: skin involvement, myalgia or muscle tenderness, hypertension, peripheral neuropathy, or renal involvement [2]. Our patient presented with histopathological evidence of vasculitis and related organ involvement. However, without a comprehensive workup, the definitive connection between his underlying medical history and bowel perforation presentation remains unanswered.

An estimated 50-70% of patients with PAN present with gastrointestinal involvement [3]. In one study reviewing 54 patients with polyarteritis nodosa, 24 cases (44%) were found to have gastrointestinal lesions, with 13 of them (54%) developing acute abdomen, and a higher mortality rate in the group that received surgery (23% compared to 9% in the conservative treatment group) [4]. According to the revised criteria Five-Factor Score (FFS) criteria for evaluation of patients with vasculitis, there are four factors associated with increased mortality in PAN: 1) age >65 years, 2) cardiac symptoms, 3) gastrointestinal involvement, and 4) renal insufficiency (plasma creatinine >1.7 mg/dL). Our patient met 3 of the 4 criteria, indicating a 40% 5-year mortality ($p < 0.0001$) [5]. The poor prognosis of patients with possible undiagnosed PAN presenting with acute abdomen warrants a complete and thorough vasculitis workup with appropriate management in patients with suggestive clinical profiles. This case presents a possible rare incidence of an exacerbation of PAN in a

young, African American male under the age of 30 with his poor outcomes compounded by medication noncompliance. In cases of acute abdomen with evident organ involvement, and thorough differential including underlying small-medium vasculitis should be considered, even in younger patients.

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