An Unusual Case of Gastrointesetinal Symptoms Leading to a Diagnosis of Polyarteritis Nodosa

by Anita Mittal, Marie L. Borum and Anthony C. Venbrux

INTRODUCTION

Classic Polyarteritis Nodosa, or c-PAN, is a type of systemic vasculitis characterized by necrotizing inflammatory lesions affecting predominately medium and small muscular arteries, usually involving the nerves, gastrointestinal tract, skin, heart, and nonglomerular renal vessels. The inflammatory lesions occur preferentially at vessel bifurcations. It results in microaneurysm formation, thrombosis, aneurysmal rupture with hemorrhage, and organ infarction.

The following case is about a woman with progressive arthritis of unknown etiology who developed upper gastrointestinal bleeding. This case is interesting because it was her gastrointestinal symptoms that led to a thorough GI evaluation and subsequent diagnosis of classic polyarteritis nodosa. In addition, the gastrointestinal findings that this patient displayed are not usually seen, occurring less than 10% of the time.

CASE REPORT

A 77-year-old Caucasian female with multiple medical problems, including a history of progressive arthritis complicated with skin ulcerations, was readmitted to the ICU with sepsis, abdominal pain, and respiratory distress therefore requiring intubation. She had been discharged from the hospital just five days prior to this admission for a small bowel obstruction and perforated duodenal ulcer, followed by a partial gastrectomy and J-tube placement. A GI consult was obtained to assist in management of the patient.

Her evaluation on this admission initially included laboratory studies that demonstrated evidence of anemia. The Hct = 29.2, Hb = 9.6, and the WBC = 5.5. A lactate was also drawn and found to be initially elevated. The patient then developed an upper gastrointestinal bleed. An EGD showed erythema, edema, small erosions in the proximal jejunum, and the previously placed J-tube eroding through the mucosal lining.

Radiographic studies and labs for P-ANCA and C-ANCA were then ordered to further evaluate the cause of these findings. The labs were normal (P-ANCA < 21; C-ANCA <21). However, an abdominal radiograph revealed dilated loops of bowel, and a gallbladder with fluid and a thickened wall. These findings, combined with the elevated lactate level were concerning for possible ischemia, and an angiogram was subsequently performed. The results showed small microaneurysms in the superior mesenteric artery and renal arteries, thus leading to a diagnosis of c-PAN (Figures 1, 2).

Prior to this angiogram and admission, the patient was being treated presumptively with Cytoxan and prednisone for suspected vasculitis based on clinical symptoms. During this admission, Azathioprine 50 mg

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A CASE TO REMEMBER

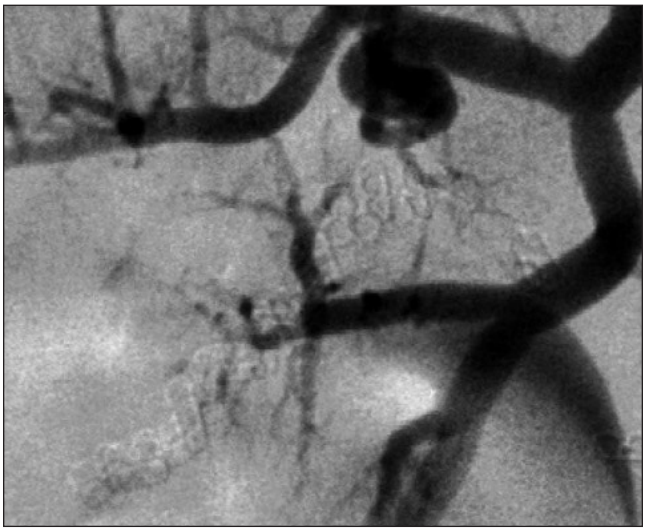

Figure 1.

daily, and prednisone 40 mg daily were started, and currently the patient shows no signs of active vasculitis.

DISCUSSION

This particular case is unusual because our patient's progressive arthritis and upper gastrointestinal bleeding led to the work up that established a diagnosis of polyarteritis nodosa. Gastrointestinal manifestations can include abdominal pain (in up to 25% of patients with c-PAN), gastrointestinal bleeding (7%), peritonitis (4%), intestinal infarction (2%), pancreatitis (2%), duodenal ulcer (2%), and cholecystitis (1%). Other rarer gastrointestinal manifestations include ischemic hepatitis, gastritis, and esophagitis.

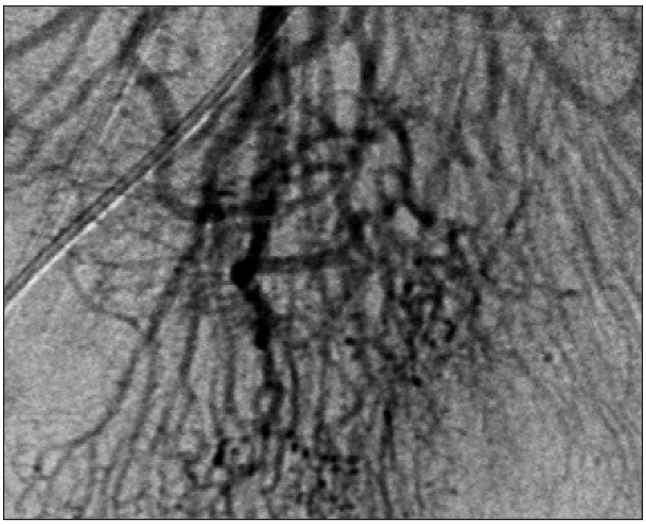

Figure 2.

Classic PAN is diagnosed by biopsy or arteriography while keeping clinical symptoms in mind, but not by clinical presentation alone. Our patient was being presumptively treated for vasculitis before this admission based on clinical presentation, and had not yet undergone any studies such as biopsy or arteriography to obtain a specific diagnosis. Arteriography is most often performed to study the visceral and renal circulation, in which the diagnosis is suggested by the presence of microaneurysms, stenoses, or a beaded pattern brought about by sequential areas of arterial narrowing and dilation. On this admission, the aforementioned picture was seen in the angiogram of our patient, and therefore these findings combined with the patient's clinical symptoms were thought to be consistent with a diagnosis of c-PAN.

This case is also interesting because gastrointestinal involvement is typically regarded as an indicator of poor prognosis, yet our patient survived. In addition, aneurysms found in the kidney, the liver, and the mesenteric arteries are associated with more severe and extensive disease. When left untreated, the 5-year survival rate of PAN is 13%. However, corticosteroid treatment improves the 5-year survival rate to 50-60%. When steroid treatment is combined with other immunosuppressants, such as in the case of our patient, the 5-year survival rate may increase to greater than 80%. Prompt steroid and immunosuppressant therapy probably contributed to her survival, even though she had poor prognostic indicators and severe, extensive disease. ■

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