Case Report

Acute Pancreatitis with Pseudocyst Formation in a Patient with Polyarteritis Nodosa

ERNEST SURESH, WENDY BEADLES, PHILIP WELSBY, and RAASHID LUQMANI

ABSTRACT. Polyarteritis nodosa (PAN) is a term that includes patients with necrotizing inflammation of medium sized arteries, and excludes those with microscopic vessel involvement. Its manifestations are protean and include constitutional symptoms such as fever, malaise, weight loss, myalgia, peripheral neuropathy, rash, and gut and renal involvement. Although gastrointestinal manifestations have been noted in up to a third of patients with PAN, clinical presentation with pancreatic involvement has been reported only rarely. We describe a patient with PAN who developed acute pancreatitis with pseudocyst formation as well as infarcts in the spleen and liver. (J Rheumatol 2005;32:386–8)

Key Indexing Terms:
ACUTE PANCREATITIS
SPLENIC INFARCT
HEPATIC INFARCT
PANCREATIC PSEUDOCYST
POLYARTERITIS NODOSA

The term polyarteritis nodosa (PAN) is now restricted to patients with necrotizing inflammation of medium sized arteries, and excludes those with microscopic vessel involvement. Its manifestations are protean and include constitutional symptoms such as fever, malaise, weight loss, myalgia, peripheral neuropathy, rash, and gut and renal involvement. Although gastrointestinal manifestations have been noted in up to a third of patients with PAN, clinical presentation with pancreatic involvement has been reported only rarely. We report a patient with PAN who developed acute pancreatitis with pseudocyst formation as well as infarcts in the spleen and liver.

CASE REPORT

A 62-year-old man presented in January 2004 with a one-month history of fever, rigors, myalgia, weight loss of about 7 kg, and a rash over both legs. Systemic enquiry was otherwise entirely negative. His medical history included a 6-year history of recurrent episodes of fever, arthralgia, and rash similar to the current presentation. These episodes usually lasted for a few weeks each time and resolved without treatment. He had also been diagnosed with benign monoclonal IgG gammopathy 4 years previously (maximum paraprotein level of 11 g/dl). He drank less than 6 units of alcohol weekly. He was not taking any regular medications at the time of admission.

His temperature was 38°C on admission and there were a few faint macular lesions on his calves. The rest of the physical examination, including urine dipstick, was unremarkable. Initial investigations revealed the following: hemoglobin 13.7 g/dl, white cell count 5.7 × 10^9/l, platelet count 369 × 10^9/l, Westergren erythrocyte sedimentation rate 80 mm at 1 h, C-reactive protein 869 mg/l, urea 4.3 mmol/l, creatinine 66 µmol/l, alanine aminotransferase 40 units/l, alkaline phosphatase 100 units/l, and creatine kinase 30 units/l. Chest radiograph was within normal limits. Serial blood cultures were sterile. Serology for hepatitis B and C, human immunodeficiency virus, Brucellosis, meningococcus, parvovirus and Lyme disease was negative. Tests for anti-nuclear antibody, antibodies to extractable nuclear antigens, and anti-neutrophil cytoplasmic antibodies were all negative.

Five days after admission, he developed generalized abdominal pain associated with vomiting. On examination, his abdomen was diffusely tender but otherwise unremarkable. His amylase was normal. Plain radiograph of his abdomen revealed large bowel dilatation and edematous bowel. Gastroscopy showed a fundal lesion of uncertain etiology, while colonoscopy revealed patchy areas of inflammation. Gastric and colonic biopsies showed only non-specific changes. Computed tomographic (CT) scan of his abdomen and pelvis revealed no abnormalities.

One week after the onset of abdominal pain, he developed mononeuropathy multiplex that progressively resulted in right ulnar sensory and motor impairment, right median motor impairment, and left ulnar motor impairment. His macular rash became more prominent at this stage and a biopsy of the rash showed features consistent with leukocytoclastic vasculitis. A presumptive diagnosis of PAN was made and it was planned to treat him with intravenous pulses of methylprednisolone and cyclophosphamide. Treatment with cyclophosphamide however had to be deferred as he developed septic shock due to Escherichia coli soon thereafter, and had to be admitted to the intensive care unit. He was therefore given only one intravenous pulse of methylprednisolone and subsequently continued daily hydrocortisone until the infection resolved.

Recurrent epigastric pain continued, associated now with elevation of serum amylase. An abdominal CT scan was repeated and this showed extensive peripheral areas of low attenuation throughout the spleen typical of splenic infarcts, multiple cystic areas in the pancreas suggestive of pancreatic infarcts, and multiple liver lesions that were also consistent with infarcts (Figure 1). A CT scan one month later showed progression of changes with extensive pancreatic pseudocyst formation extending into the peritoneum (Figure 2). Clinical symptoms and amylase elevation gradually
resolved with conservative treatment. CT scan of his abdomen at this stage showed regression in the size of the pancreatic pseudocyst. After receiving 6 intravenous pulses of cyclophosphamide and methylprednisolone, he was switched to azathioprine and his vasculitis was in remission on followup one month later.

DISCUSSION
Several management difficulties were encountered with this patient. First, the differential diagnosis on presentation was broad and the priority was to exclude infectious causes. There was no indication of vasculitis at this stage. Second, pancreatitis could not be diagnosed initially, as the clinical picture was not typical and serum amylase was normal. The underlying cause of his abdominal pain remained unclear, as the results of other investigations were unhelpful. Third, although the later development of mononeuritis multiplex and vasculitis on skin biopsy enabled a presumptive diagnosis of systemic necrotizing vasculitis, treatment with cyclophosphamide had to be delayed as gram-negative septicemia had developed. All these factors probably resulted in progression of vasculitis and led to the pathogonomic development of infarcts in his pancreas, spleen, and liver.

PAN was considered the most likely diagnosis as there was evidence of involvement of medium sized vessels (e.g., mononeuritis multiplex) and sparing of microscopic vessels (e.g., absence of glomerulonephritis), although definite histological evidence of PAN was not obtained from any infarcted organs. Celiac axis angiography was considered but the patient was too ill to undergo this procedure. Moreover, the radiologist felt that the abdominal CT scan revealed a lot of vascular calcification in keeping with severe atherosclerosis and therefore that it would be difficult to distinguish between thrombosis secondary to vasculitis and thrombosis secondary to atherosclerosis. In retrospect, we feel that a celiac axis angiogram might have given us useful information.

In 230 cases with PAN examined post mortem, involvement of pancreatic vessels occurred in 50% and pancreatic infarction in 6%. As this study predated the 1993 Chapel Hill Conference (at which the classification system of vasculitic disorders was standardized), it is possible that a proportion of these cases might not have had currently categorized PAN. Presentation with acute pancreatitis has been reported only rarely in PAN and the diagnosis was usually established at laparotomy or autopsy. Death was common in cases reported before the 1980s. Complications of pancreatitis reported in PAN include pancreatic insufficiency with malabsorption, infected necrosis, and formation of pseudo-pancreatic cyst. Splenic and hepatic infarcts are rare. The coincidence of simultaneous infarcts in the pancreas, spleen, and liver (celiac axis triad) in PAN has been reported only once. Acute pancreatitis has also been reported in Wegener’s granulomatosis, Henoch-Schonlein purpura, Kawasaki disease, Behçet’s disease, and as a side effect of drugs such as corticosteroids and azathioprine used to treat the vasculitis. In our patient, the onset of pancreatitis predated the use of these medications.

In conclusion, acute pancreatitis is a rare cause of abdominal pain in PAN. Serum amylase may be normal in the early stages. CT scanning and/or celiac axis angiography are useful investigations. Although there are no controlled trials, treatment with cyclophosphamide should be commenced to improve prognosis. An early surgical opinion should be sought in all patients.
REFERENCES