Case Report

Acute Ischemic Enteritis due to Polyarteritis Nodosa: A rare case report

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Abstract

Polyarteritis nodosa is a form of vasculitis that affects several organs. Gastrointestinal involvement is frequent, but cases in which the gastrointestinal tract is the only site of disease are rare. In this paper, we report a case of a 40-year-old patient with polyarteritis nodosa restricted to small part of the small intestine, which underwent resection and anastomosis. Laparotomy was performed despite normal radiological findings. He underwent resection of the infracted segment. Histopathological examination confirmed the diagnosis. Thus, a pathological diagnosis of ischaemic infarction due to polyarteritis Nodosa (PAN) was made. The patient had no other signs of chronic vasculitis and for this reason surgical treatment resolved the clinical symptoms.

Keywords: polyarteritis nodosa, gastrointestinal vasculitis, ileal necrosis

INTRODUCTION

Polyarteritis nodosa (PAN), initially described by Kussmaul and Maier, is a systemic vasculitis that affects medium-sized muscular arteries. Abdominal complications, due to mesenteric vasculitis, observed in up to 50% of patients with polyarteritis nodosa, are especially difficult to diagnose because the symptoms often mimic those of more common diseases (Cengiz et al., 2010).

Polyarteritis nodosa (PAN) is a rare form of systemic vasculitis that predominantly affects male (M/F: 1.5/1) and mostly occurs in the 4th decade (Shirai et al., 2013). In rare cases, PAN is associated with hepatitis C infection (Seja et al., 2006). However, acute surgical abdomen associated with the intestines as the first manifestation of PAN is a rare event. Although prognosis has significantly improved owing to immunosuppression, PAN may still have a fatal course when gastrointestinal complications occur.

Case report

A 40-year-old male, presented to the emergency department with severe abdominal pain and vomiting for the last two days. His medical history was remarkable for lower respiratory tract infection. On physical examination, he was ill and the entire abdomen was tender on palpation. The laboratory tests were as follows: white blood cells (WBC), 19700/mm³; hemoglobin (Hb), 10.9 g/dl; platelets, 385000/mm³; and erythrocyte sedimentation rate (ESR), 82/mm. Blood for Hepatitis B surface antigen (HBsAg) and urine for porphyrins tested negative. A total serum protein was 57 g/l and serum albumin 12 g/l. The serum bilirubin level was 135 umol/l; platelets, 385000/mm³; and erythrocyte sedimentation rate (ESR), 82/mm. Blood for Hepatitis B surface antigen (HBsAg) and urine for porphyrins tested negative. A total serum protein was 57 g/l and serum albumin 12 g/l. The serum bilirubin level was 135 umol/l, AST 52 IU/l, ALT 58 IU/l, and alkaline phosphatase 210 IU/l. The blood urea level was 26.7 mmol/l, serum creatinine was 1.91 mol/l. Serum electrolytes and amylase levels were normal.
Figure 1. Segment of small bowel showed intestinal ischaemia and infarction.

Figure 2. Intense inflammation of submucosal medium-sized vessel walls with circumferential or eccentric/segmental fibrinoid necrosis (HE, original magnification x40)

Abdominal computerized tomography (CT) scan revealed normal mesenteric vasculature. Since abdominal pain was intensified and the examination was consistent with acute abdomen despite appropriate antibiotic treatment, laparotomy was performed, which showed distended intestinal loops and segmental ischemia (10 cm length) about 60 cm from the ileocecal valve. He underwent resection of infarcted segment. The stomach, large bowel, liver and spleen on inspection looked normal.

Grossly, the segment of resected small bowel measured 10 cms in length. Externally, it appeared dark with a diffuse grayish brown exudate on the serosal aspect (Figure 1). Microscopically, the ileal segment showed patchy ischaemic ulcerations and transmural necrosis. Besides vascular ectasia and congestion, medium sized submucosal arteries showed endothelial proliferation, occasional luminal thrombi and distinct fibrinoid necrosis in the media, focal to patchy destruction of internal elastic lamina, sparse giant cell reaction and a varying proportion of chronic inflammatory cells mainly eosinophils, infiltrating the adventitia (Figure 2). Similar pathology was seen involving the branches of mesenteric arterial arcades producing distinct aneurysms. Thus, a pathological diagnosis of ischaemic infarction due to polyarteritis Nodosa (PAN) was made.

DISCUSSION

Gastrointestinal involvement in PAN is a common cause of morbidity and mortality. This patient illustrates the uncommon complication of small bowel infarction. In PAN, gastrointestinal involvement is noted in 30-50% of cases (Lefrou et al., 2003). In our case the first indication of the disease was evident on histology. Our patient presented with abdominal pain which is common in two thirds of cases with PAN. Gut infarction is reported in 10% of patients with PAN (De Golovine et al. 2008). Early
diagnosis of PAN with gastrointestinal involvement is difficult. Acquisition of tissue for histological diagnosis is essential. Once PAN is established, joint medical and surgical management is essential to prevent the relapse and complications such as perforation, haemorrhage or obstruction. Immunosuppression with steroids and cyclophosphamide is the mainstay of treatment particularly for the patients with gastrointestinal involvement. Abdominal CT scan is able to show the intestinal involvement and the most common radiological signs include: symmetrical bowel thickening, target signs and vascular engorgement (Ha et al., 2000).

Selective angiography of the mesenteric vessels is regarded as the gold standard for arterial occlusion. Although of only moderate sensitivity, it is specific for Polyarteritis nodosa when medium size vessel aneurysms are demonstrated. Angiography presents 80% true positivity in patients with polyarteritis Nodosa (Ebert et al., 2008). However, in the absence of clinical information, it is difficult to determine whether the signs seen on diagnostic procedures are caused by vasculitis. In our patient the disease was confined only to the small bowel. The treatment of intestinal vasculitis is challenging. Generally, a surgical approach is rarely adopted in patients with systemic vasculitis and is indicated in the case of complications. In our case, because of isolated disease, the surgical procedure resolved the clinical symptoms and no additional therapies were required. Antibiotics may be essential to treat polymicrobial infections. Prognosis of untreated patients has been improved by use of steroids with 48% survival at 5 years. The combination of cyclophosphamide with prednisolone results in 96% remission (Ahn et al., 2009).

CONCLUSION

Polyarteritis Nodosa is a rare cause of ischaemic enteritis and, in isolated forms, the pathological report is essential for the right diagnosis.

REFERENCES