CASE REPORT

Rare and unusual case of polyarteritis nodosa involving the gastrointestinal tract leading to bowel gangrene

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SUMMARY

Polyarteritis nodosa (PAN) is a systemic necrotising vasculitis preferentially targeting medium-sized arteries and not related with glomerulonephritis or small vessel involvement. Clinical manifestations of PAN are multisystem. The gastrointestinal, renal, cardiac, musculoskeletal, skin and central nervous systems may be involved. The aetiology remains unknown, and the ensuing vasculitis may lead to aneurysm formation and thrombosis in any organs of the body with resultant ischaemia. PAN of the intestines is a relatively common manifestation of this disease but rarely causes bowel ischaemia resulting in necrosis. Here we report a case of a young Chinese patient who presented with an acute abdomen requiring surgery and made good recovery post operatively. He remains free of symptoms while on steroid therapy.

BACKGROUND

The diagnosis of polyarteritis nodosa (PAN) is a challenge and is frequently missed in the early stage. This is mainly attributed to the diversity of symptoms and the non-existence of any specific biochemical or haematological tests. As a consequence, PAN with gastrointestinal involvement carries a poor prognosis and is significantly associated with increased mortality. Medical treatment with immunosuppressant therapy remains the mainstay of management. An acute abdomen resulting from PAN consist of an array of pathological occurrence's processes, for example haemorrhage, ischaemia and necrosis of organs will more often require emergent surgical treatment.

CASE PRESENTATION

A 39-year-old Chinese male presented to the emergency department with a 3-days history of right iliac fossa pain. Physical examination revealed tenderness and localised guarding on palpation of the affected region. There was no history of per rectal bleeding or any constitutional symptoms. Examination for rebound tenderness was positive. He has no significant medical history. He had a low-grade fever. In view of the history, examination and haematological findings, a provisional diagnosis of acute appendicitis was made. The patient was subsequently prepared for laparoscopic appendicectomy.

INVESTIGATIONS

Initial investigations revealed mild leucocytosis and other biochemical blood tests were within normal range.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis was acute appendicitis, acute diverticulitis or less likely perforated right-sided colon tumour.

TREATMENT

On laparoscopic examination, it was noted, however, that the appearance of the appendix did not concur with the physical examination findings. Instead, the caecum appeared to be gangrenous and this pathological change has extended to the proximal part of the ascending colon and transverse colon ([figure 1](#)). The rest of the small bowel and colon appeared healthy. A decision to proceed with an extended right hemicolectomy was taken and executed in the usual manner via open surgery. A double barrel stoma was fashioned in an area of least tension. The patient made good recovery post-operatively and was discharged on day 5 postoperatively. On further inspection, the gross specimen showed multiple areas of infarction interspersed by areas of hyperaemia. The proximal and distal borders were of healthy normal bowel ([figure 2](#)).

OUTCOME AND FOLLOW-UP

Subsequently, the patient was managed together with rheumatologists who continued with immunological and hepatitis screening. The tests turned to be negative. Microscopically, the specimen showed enlarged medium-sized blood vessels with markedly thickened walls. The blood vessels that were dilated

Figure 1 A laparoscopic view of the gangrenous segment of the ascending colon.

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Figure 2 The colon is partially cut open revealing multiple areas of infarction surrounded by areas of hyperaemia.

and congested were seen and some showed evidence of vasculitis with infiltration of neutrophils. Some vessels show atheroma with calcification and a few had total occlusion by calcium deposits. Osseous metaplasia was noted in the vessels with the formation of bony trabeculae within. Elastic Von Gieson stain was used to show loss of elastic fibres in the blood vessels, consistent with vascular disease. The final pathological diagnosis was vasculitis secondary to polyarteritis nodosa (PAN) (figure 3). The angiographic findings in our patient did not reveal any of the common abnormalities found in PAN (figure 4). Nonetheless, he was still given a high dose immunosuppressant (oral cyclophosphamide; 2 mg/kg/day) and has revealed good response and remains in remission. He now is free of further attacks and having good quality of life.

DISCUSSION

PAN is a primary systemic necrotising vasculitis predominantly targeting medium sized arteries defined as the main visceral arteries and their associated branches.¹ It may be idiopathic or triggered by specific agents. PAN may be a systemic disease or confined to single organs or systems.² The annual incidence of PAN currently ranges from 0 to 1.6 cases/million residents in European countries and its prevalence is ~31 cases/million.^{3 4}

PAN affects patients of any age, gender or ethnic background. It is initially considered as an immune-complex-related disease based on the formation of necrotising arteritis in animal models of immune-complex-mediated injury.⁵ Characteristic arteriography lesions in PAN are arterial saccular or fusiform microaneurysms (1.5 mm in diameter), which usually co-exist with stenotic lesions predominant in the kidneys, mesenteric and hepatic arterial branches.⁶ Owing to the ubiquity of blood

Figure 3 Medium-sized vessels, mainly arteries, show evidence of vasculitis with marked infiltration with neutrophils. (Hematoxylin and Eosin stain under 10× magnifications).

Figure 4 Mesenteric angiography showing no significant bleeding or microaneurysm of the inferior mesenteric artery and its associated branches.

vessels, the occlusion or rupture of the inflamed arteries may produce tissue ischaemia or haemorrhage in a various organs or systems. In our case, the gastrointestinal system was affected due to mesenteric ischaemia and the resultant colonic infarction. Gastrointestinal involvement is seen in 14–65% of patients with PAN with the gallbladder and small bowel as the most frequent sites of involvement, therefore making our case a rare example of colonic involvement of PAN. Other systems that may be involved include the renal, cardiac, central nervous and musculoskeletal systems. As a consequence, the clinical manifestations of PAN are very broad that includes non-specific constitutional indicators such as malaise, weight loss, fever, arthralgia and myalgia.⁶ The peripheral nervous system and the skin are the most frequently involved areas. Other major reported gastrointestinal complications include perforation, haemorrhage, ulceration and obstruction. Interestingly, our patient denies of any prodromal symptoms preceding his presentation to us. It is indeed a rare case of isolated gangrenous colon caused by a systemic disease.

There are no laboratory tests specific for PAN. The histopathology demonstration of vascular inflammation in medium-sized or small arteries is fundamental to sustain the diagnosis of vasculitis as well as excluding other disorders.⁷ Visceral angiography may be carried out in circumstances in which PAN is highly suspected and histological diagnosis of vasculitis cannot be achieved, or in patients predominantly undergoing symptoms reminiscent of abdominal, renal or cardiac involvement. Our patient underwent emergency laparotomy with a provisional diagnosis of acute appendicitis; therefore, there was a very low index of suspicion for any vasculitic pathology at that point. The diagnosis of PAN was established postoperatively upon histopathological examination, thus making visceral angiography mandatory to further investigate the extent of disease.

Characteristic arteriography lesions in PAN are arterial saccular or fusiform microaneurysms (1.5 mm in diameter), which

usually co-exist with stenotic lesions predominantly in the kidneys, mesenteric and hepatic arterial branches.⁶ The angiographic findings in our patient did not reveal any of the common abnormalities found in PAN. Furthermore, the immunological markers (ie, anti-neutrophil cytoplasmic antibodies, anti-nuclear antibodies, rheumatoid factors), hepatitis screening and urinalysis test were also negative. The path to establishing diagnosis was far from straightforward with our patient. Even with the conflicting results, PAN was still suspected and can be qualified on the basis of high probability. To date, there are no specific diagnostic criteria for PAN. The rheumatologists decided to pursue therapeutic trial with immunosuppressant therapy with which the patient responded well over time.

PAN has been classically considered as a monophasic disease with a relapse rate inferior to 10%. The level of evidence supporting therapeutic decisions in PAN is low. Current therapeutic methods consider treating mild forms of primary PAN with corticosteroids only. In life-threatening situations or rapidly progressive disease, therapy with intravenous methylprednisolone pulses (1000 mg/day for 3 days) may be clinically useful. In the presence of critical organ involvement, immunosuppressants are given in addition to prednisolone.⁸ Currently, our patient is on a daily maintenance dose of 10 mg of oral prednisolone.

PAN with gastrointestinal involvement and acute abdomen carries a poor prognosis and is significantly associated with an increased mortality if not diagnosed and treated early. The 6-month and 5-year survival rates are 60% and 46% correspondingly.⁹ However, prognosis has markedly improved in the past few decades largely owing to improved management with

prompt surgical intervention if indicated, and the combined use of immunosuppressive therapy. Even though the mainstay of treatment of PAN is medical, surgery may be required for some disease complications, such as perforation, rupture, ischaemia or haemorrhage of the gastrointestinal tract or kidneys. The surgical removal of the involved organ is in this case curative¹⁰

Contributors All authors from surgery departments of various universities around the country cooperated and contributed to this case report. AVJ and NQF are responsible for reporting, coordinating and planning besides the data acquisition. RFJ and ADZ monitored, finalised and edited the case report and data to be published.

Competing interests None declared.

Patient consent Obtained.

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Learning points

Polyarteritis nodosa of the intestines is a common manifestation of the disease, but it should not be taken lightly as it can lead to bowel gangrene. The multidisciplinary approach significantly improves patients' long-term survival.

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