Case Report

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Intestinal obstruction secondary to infantile polyarteritis nodosa

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ABSTRACT

Polyarteritis Nodosa (PAN) is a rare systemic necrotising vasculitis of medium and small-sized arteries. Patients typically present with systemic symptoms. Obstructive intestinal symptoms are described but usually resolve with treatment of the underlying vascular disease. We report a case of a one year old boy with multiple ischemic small bowel strictures secondary to infantile PAN, who was treated with resection of the affected segments by single port laparoscopy.

Key words: Infantile polyarteritis nodosa, PAN, polyarteritis nodosa, small bowel obstruction, stricture

INTRODUCTION

Polyarteritis nodosa (PAN) is a systemic necrotising vasculitis that rarely leads to intestinal strictures. PAN typically affects medium- and small-sized arteries.[1] Patients usually present before 2 years of age with aneurysmal dilatation of coronary arteries or systemic symptoms such as involvement of kidneys, skin, joints, muscles, nerves, or the gastrointestinal (GI) tract.[2]

Kawasaki disease belongs to the same spectrum as PAN; it is an acute self-limited vasculitis that occurs predominately in infants and young children and was first described in 1967 by Kawasaki.[3] Infantile PAN and Kawasaki disease share certain clinical and pathological features and are considered by some to represent the same disease.[4]

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We report a case of ischemic small bowel strictures secondary to acute idiopathic vasculitis of medium and small vessels (or infantile PAN) in a 1-year-old boy treated surgically by single port laparoscopic small bowel resection and primary anastomosis.

CASE REPORT

A 12-month-old male presented with fevers, torsal rash, diffuse lymphadenopathy, eye redness, and progressive non-bilious vomiting. A weight loss of 1.5 kg within 4 months was recorded. At 9 months of age, the patient was diagnosed with Kawasaki disease at an outside institution and treated with intravenous immunoglobulin and oral Aspirin. Before admission, the patient had regular, formed bowel movements. On physical examination, no abdominal distension was noted. The initial echocardiogram showed right coronary artery aneurysm and left ventricular thrombus.

Gastric biopsies yielded acute gastritis with no other diagnostic abnormalities. Abdominal and pelvic ultrasound followed by computed tomography (CT) arteriogram were consistent with a partial small bowel obstruction and showed a markedly thickened appendix with mucosal hyper-enhancement, and a prominent vascular feeding pedicle, suggestive for vasculitis as a cause for appendiceal inflammation [Figure 1]. An upper GI (UGI) study with small bowel follow through confirmed delayed transit with high-grade small bowel obstruction related to stricture in the left lower quadrant.

The patient was taken to the operating room for a single port exploratory laparoscopy. One umbilical 11 mm port was placed to introduce the 10 mm Storz Hopkins telescope with inbuilt 5 mm working channel [Figure 2]. This device carries a 0-degree optic and allows visualization and manipulation of the intestines with soft bowel graspers. Two

small bowel strictures were found: 15 and 200 cm proximal to the ileocaecal valve. Both affected small bowel segments were exteriorised through the umbilical trocar site, resected extracorporally and primarily re-anastomosed in a hand sewn end-to-end anastomosis [Figure 3].

The pathology of the resected small bowel segments showed markedly narrowed and ulcerated small bowel segments with focal transmural necrosis and submucosal fibrosis consistent with a stricture after ischaemic type injury. No thrombosis of the blood vessels was found [Figure 4].

The patient recovered well from the operation and was discharged on the post-operative day 6 after. On follow-up visit 1 month later, he was tolerating oral diet well, had no more vomiting episodes and was gaining weight.

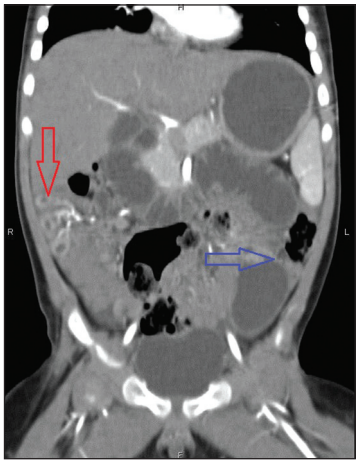


Figure 1: Abdominal computed tomography arteriogram. Coronal reformatted image shows marked enhancement of the appendiceal mucosa with dilated feeding vessels (red arrow) and area of abrupt calibre change at the distal small bowel in the left lower quadrant (blue arrow)

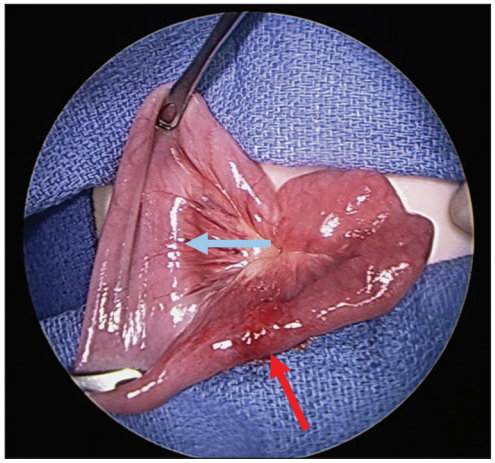


Figure 3: Exteriorised proximal small bowel stricture (red arrow) with dilated proximal small bowel (blue arrow)

DISCUSSION

The diagnosis 'systemic vasculitis' covers a wide spectrum of diseases that often overlap and not always present with a clear distinction.

Population prevalence estimates for PAN ranges from 2 to 33/million.[5] The pathogenetic mechanisms in PAN are poorly understood. Thickening of the inflamed vessel walls and intimal proliferation can cause luminal narrowing, reducing the blood flow with subsequent tissue ischemia. The resulting tissue damage or infarction can cause variant clinical manifestations.

Abdominal pain is the most common and an early symptom in patients with mesenteric arteritis.[6] Ischaemia due to vasculitis affects the small intestine more commonly than other areas of the GI tract. Only a subset of patients suffers from predominant mesenteric arterial involvement with little or no clinical evidence of extraintestinal vasculitis.[6]

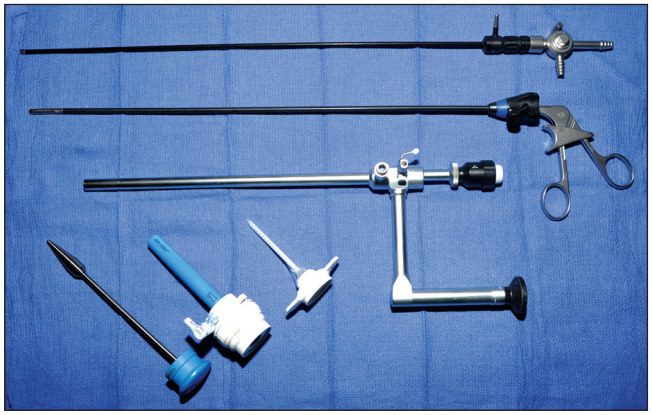


Figure 2: Single port instrument: Storz Hopkins 10 mm with inbuilt 5 mm working channel

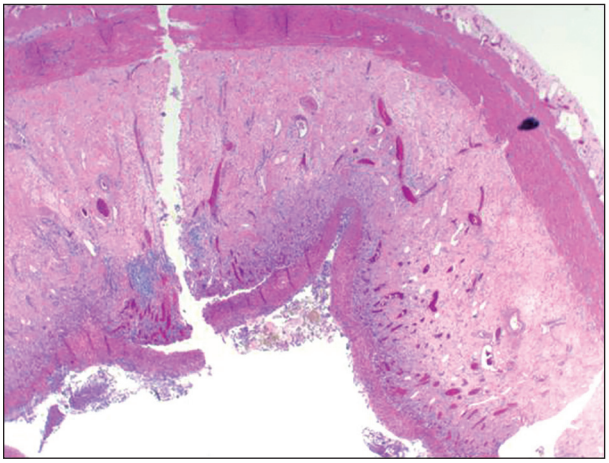


Figure 4: Pathology of the small bowel wall (H and E, ×10). Submucosal fibrosis and a serosal fibro-inflammatory reaction

There are few reports in the literature of vasculitis leading to intestinal obstruction. Most cases have been managed non-operatively. The scarce reports of ischemic strictures caused by vasculitis that required surgical resection have been associated with Kawasaki disease and Henoch-Schonlein purpura.[7,8] Our review of the current literature resulted in only two reported cases with clinically relevant intra intestinal manifestations of PAN: One adult patient with ischemic colon, treated non-operatively and one paediatric patient with jejunal stricture, who underwent bowel resection via laparotomy.[9,10] There are no cases in the literature of intestinal obstruction associated with infantile PAN.

Radiologic findings in various types of vasculitis often overlap considerably and therefore have limited value in making a specific diagnosis. Nevertheless, the possibility of vasculitis should be considered whenever mesenteric ischaemic changes occur in young patients, are noted at unusual sites (e.g., stomach, duodenum, and rectum), have a tendency to concomitantly involve the small and large intestine, and are associated with genitourinary involvement.

The UGI showed a delayed intestinal transit of contrast with high-grade small bowel obstruction in the right lower quadrant. CT arteriogram replaced the conventional angiogram and showed signs of active vasculitis in vessels arising from the superior mesenteric artery, feeding a hyperaemic appendix. The latter is consistent with on-going vasculitis, whereas we interpreted the strictures as post-ischemic sequelae of chronic focal small bowel vasculitis. This was consistent with the pathology. Although no definite active vasculitis or intravascular thrombus was identified in these sections, the pathological findings were consistent with an ischemic-type injury secondary to vasculitis.

Single port laparoscopy using the 10 mm Storz Hopkins telescope with inbuilt 5 mm working channel offers a safe, economical, and very valuable tool for exploration and treatment of intestinal processes. Compared to an initial exploratory laparotomy, the advantages of this technique lie in its very small umbilical skin incision with excellent cosmetic results and quicker recovery.

This report describes the unique finding of two small bowel strictures caused by a systemic small and medium vessel vasculitis, diagnosed by ultrasound and CT arteriography in a 12-monthold-child. Compared to the more invasive conventional angiogram, ultrasound and CT arteriography provided sufficient diagnostic evidence to diagnose the small bowel obstruction and vasculitis. Single port laparoscopy was shown to be safe and effective in the treatment of this condition with a virtually scarless cosmetic result.

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