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CASE REPORT

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Cecal vascular malformation mimicking appendicitis in a child

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Abstract A 5-year-old girl presented with abdominal pain suggestive of appendicitis. Intraoperatively, a solid cecal mass was identified along with mesenteric adenopathy. A right hemicolectomy was performed. Pathologic examination revealed a vascular malformation with evidence of recent hemorrhage.

Keywords Vascular malformation · Colon · Pediatric · Abdominal pain · Appendicitis

Introduction

Vascular malformations of the gastrointestinal tract are rare findings in children [1, 2]. They usually present with painless lower gastrointestinal bleeding. We present a case of a cecal vascular malformation in a child who presented with acute abdominal pain but without gastrointestinal bleeding.

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Case report

A 5-year-old Indian girl presented to our emergency room with right lower quadrant abdominal pain for 2 days, which was associated with fever, anorexia, nausea, and vomiting. She did not have any diarrhea, melena, or hematochezia. Her past medical history was unremarkable. Physical exam showed a temperature of 38.4°C and a soft, nondistended abdomen with tenderness and guarding in the right lower quadrant. Laboratory work-up revealed a white blood cell count of 11,800 with 75% neutrophils and normal hemoglobin.

The patient was presumptively diagnosed with acute appendicitis and taken to the operating room for appendectomy. Under anesthesia, a mobile mass was palpable in the right lower quadrant. Intraoperatively, a normal appendix was found. A solid mass in the wall of the cecum was identified, located in proximity to the ileocecal valve and measuring approximately 2 cm. The mass was adherent to an inflamed omental cake. There were multiple enlarged lymph nodes in the ileocecal mesentery. The remainder of the bowel appeared grossly normal. The surgical team felt that these findings were suspicious for a localized abdominal lymphoma, and an oncologic right hemicolectomy was performed with a primary ileocolic anastomosis. The adherent omentum was also removed. The patient recovered uneventfully.

Histopathologic examination demonstrated a vascular malformation of the colonic serosa that extended through the muscularis propria into the submucosa with evidence of acute focal submucosal hemorrhage, ulceration, and edema. The malformation consisted of thick-walled arterioles and arteries as well as dilated veins (Fig. 1). Twenty-two resected lymph nodes all demonstrated simple lymphoid hyperplasia without evidence of neoplasm, granuloma, or lymphoma.

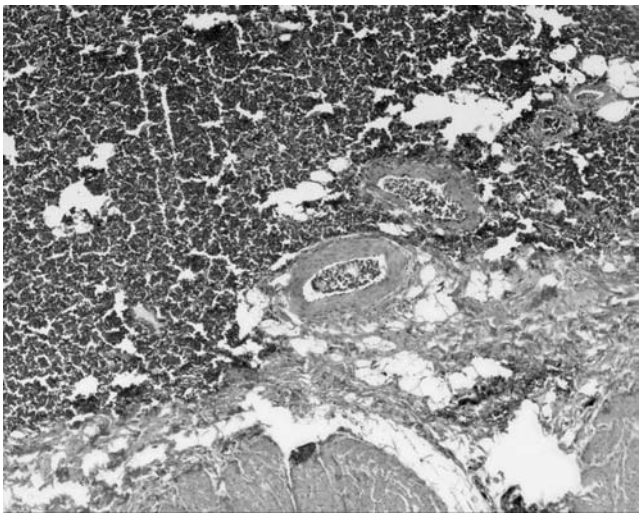

Fig. 1 Representative section of the vascular malformation showing thick-walled arterioles and dilated veins in the subserosa extending to the muscularis. Hemorrhage is seen adjacent to these structures

Discussion

Vascular malformation of the gastrointestinal tract is a rare finding in children [1–4]. Fremond et al. [2] reported nine patients treated over a 20-year period in a large Canadian children's hospital. De la Torre et al. [4] more recently reported on 18 patients with colonic lesions treated over a 12-year period in a Mexican referral center. The signs and symptoms usually begin in infancy or early childhood. Although lower gastrointestinal bleeding is the hallmark symptom of these lesions, manifestations can also include obstruction (10%), diarrhea (10%), ascites (10%), pain (5%), emesis (5%), intussusception (5%), and protein-losing enteropathy (5%) [3, 5]. Colonic lesions can lead to massive bleeding [4].

Diagnosis of vascular malformations of the gastrointestinal tract can be very challenging. A high degree of suspicion is required. These lesions may involve only a small segment of intestine and can escape detection despite repeated investigations [4, 6]. Endoscopy is an essential modality often used for initial diagnostic assessment and staging, especially when bleeding is the presenting symptom [7]. Some lesions have a pathognomonic appearance on endoscopy. Others are less clearly identifiable and require additional investigation. Colonoscopy is successful only 50% of the time in identifying colonic vascular malformations [4]. Selective mesenteric angiography detected all colonic malformations in the largest reported series [4]. It can demonstrate the characteristic appearance of the lesions as well as potentially embolize the feeding vessel [8]. Because these lesions are typically submucosal, operative identification may not be possible, and accurate preoperative localization is essential [2, 7]. These lesions can be treated by resection of the involved segment of intestine (the mostly common modality), endoscopic banding or sclerosis, percutaneous or intraoperative sclerosis, embolization, or laser photoablation [2, 3]. Lesions that did not result in significant bleeding have been simply observed [2].

Vascular anomalies are more common in the colon [1–5]. Unlike their adult distribution, which is mainly right-sided, pediatric lesions are more common in the left colon and rectum [4]. The main microscopic finding consists of vascular ectasia in the mucosa and submucosa, with no involvement of the muscularis propria. The earliest lesions consist of numerous dilated, distorted, thin-walled vessels, mostly lined only by endothelium and, less frequently, by a small amount of smooth muscle.

Our case is unique in several aspects. The malformation affected the serosal rather than the more typical mucosal aspect of the intestine. Gastrointestinal bleeding was therefore not seen. Instead, the acute hemorrhage caused a significant inflammatory reaction with lymphoid hyperplasia and omental adhesions. This inflammatory reaction was responsible for the fever and clinical picture mimicking appendicitis. The gross appearance in the operating room was highly suspicious for an abdominal lymphoma. Because total resection still constitutes optimal treatment of a localized lymphoma of the bowel and mesentery [9, 10], we performed a resection of the lesion along with its mesenteric basin. The final diagnosis was made by the pathologist. To our knowledge, this is the first reported case of an intestinal vascular malformation presenting in this manner.

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