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Small Bowel Hemolymphangioma Treated by Polypectomy in a Pediatric Patient with Cystic Fibrosis

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INTRODUCTION

Hemolymphangiomas are rare benign malformations that contain both vascular and lymphatic components (1). The lesions are congenital malformations that increase in size with age and have a predilection to involve the head, neck, and axilla (2,3). There are several reports of gastrointestinal (GI) lymphangiomas presenting with obstruction due to intussusception, GI bleeding, and abdominal pain (2–10). Standard management of these lesions is surgical resection (4,8–10). There are limited case reports of endoscopic management with sclerotherapy, double-balloon enteroscopy with polypectomy, and ablation with argon plasma coagulation (5–7).

We present a child who presented with chronic abdominal pain, constipation, and recurrent bowel obstructions who experienced symptom resolution following polypectomy removal of an ileal lymphovenous hemolymphangioma. Consent for publication was obtained from the patient's mother.

CASE REPORT

Our patient, a 15-year-old man with a history of cystic fibrosis (CF), exocrine pancreatic insufficiency, and CF-associated liver disease, presented with a 5-week history of intermittent spasmodic abdominal pain associated with increased frequency of nonbloody stools and worsened with stress. Physical examination was unremarkable. Initial laboratory investigation including complete blood count (CBC) with differential, erythrocyte sedimentation rate, c-reactive protein, electrolytes, amylase, lipase, and fecal occult blood was within normal limits. A serum liver panel showed a mild elevation in transaminases, consistent with his known CF-related liver disease. The initial impression was of functional abdominal pain and he was prescribed antispasmodic medications.

Due to refractory symptoms, he underwent esophagogastroduodenoscopy (EGD) and colonoscopy. Upper endoscopy was visually normal whereas colonoscopy showed mild scattered colonic exudates with normal terminal ileum (TI). Histology showed chronic active eosinophilic ileitis and colitis and normal upper intestinal biopsies. The patient was prescribed sulfasalazine and oral budesonide for mucosal eosinophilia.

The patient continued to experience symptoms despite medication adherence, requiring an increasingly complex medication regimen. He was admitted for presumed distal intestinal obstruction syndrome (DIOS), based on radiologic findings of organized contents in the distal small bowel, and was treated with polyethylene glycol via nasogastric tube and oral *N*-acetylcysteine.

At follow-up, he reported continued pain and underwent repeat EGD and colonoscopy, which showed resolution of colitis, but a protruding intraluminal mass with a smooth surface and normal mucosa was noted in the TI. Biopsy histology, including from the mass, was normal. It was unclear if this mass was present on initial colonoscopy, but not seen due to distance into the TI or if the mass was a new entity. Subsequent small bowel capsule endoscopy was normal and magnetic resonance enterography (MRE) noted thickening of the ileocecal valve but no discrete mass/polyp and no concern for external compression causing mucosal protrusion. Repeat CBC was normal. This was followed by readmission for recurrent bowel obstruction. Repeat endoscopy confirmed a nonocclusive mass in the TI but was otherwise visually normal (Fig. 1). Biopsy histology, including from the mass, returned with small bowel and colonic mucosal eosinophilia. Given normal biopsy histology, CBC, MRE, and small bowel capsule endoscopy it was felt the mass was benign. Moreover, since the mass did not appear to be causing luminal occlusion and biopsies did not show inflammation it was felt that recurrent bowel obstruction was secondary to DIOS and management tailored accordingly.

Despite patient-reported response to medication, he had repeat admissions for abdominal pain and bowel obstruction with inconclusive radiology findings prompting repeat colonoscopy for mass

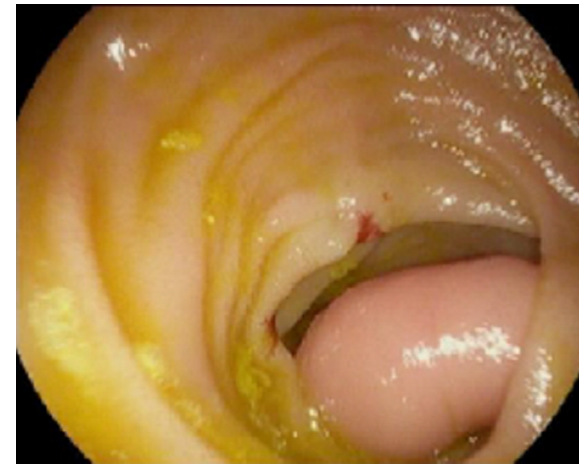

FIGURE 1. Polypoid lesion in ileocecal valve as seen on third colonoscopy.

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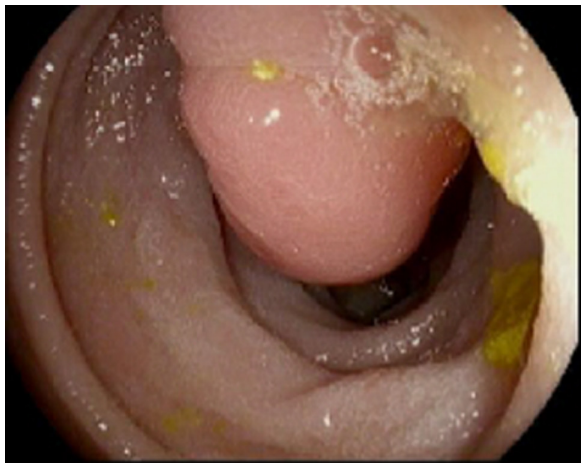

FIGURE 2. Polypoid lesion at time of resection.

excision. The lesion (Fig. 2) located approximately 5 cm beyond the ileocecal valve measured 2 cm × 1.4 cm × 1.3 cm and had a broad but identifiable stalk and smooth surface. It was snared, cauterized, and removed uneventfully. Histopathology of the mass showed multiple variable-sized vessels in the submucosa with immunohistochemical markers positive for CD31 (endothelial-specific staining) and D2-40 (lymphatic-specific staining), consistent with a venolymphatic hemolymphangioma (Fig. 3). Following polypectomy, the patient's GI symptoms completely resolved. He was able to discontinue all related medications and continues to be symptom free at 6 months follow-up.

DISCUSSION

Our patient presented with abdominal pain, constipation, and recurrent obstruction from a terminal ileal polyp masquerading as DIOS.

Lymphangiomas are benign lesions of expanding lymphatic space which is typically diagnosed in childhood as an enlarging mass affecting the head, neck, or thorax (2). GI hemolymphangiomas, a subtype of lymphangioma, once thought to be extremely rare are increasingly recognized during endoscopic procedures (1,3,10). While lymphangiomas are often diagnosed in childhood, GI hemolymphangiomas are more commonly diagnosed in adulthood (4–10). A retrospective review of one institution's experience of all GI vascular malformations and hemangiolymphangiomas over a 22-year period identified 12 hemangiolymphangiomas – all diagnosed in adult patients (ages 36-89 years) (1). A second review which identified cases of intestinal lymphangioma or lymphangiectasia diagnosed over a 40-year period identified a total of 36 lymphangiomas of which only 12 were in pediatric patients. Moreover, of the 12 identified, only 4 were intraluminal with the remaining 8 either in the mesentery or retroperitoneum (3).

Intraluminal hemolymphangiomas are most commonly found in the small bowel and can range from diminutive, incidentally found polyps to large masses resulting in intraluminal obstruction (1,3). They have a varied presentation, likely related to size and location of the lesion. In adults, there are several case reports of patients presenting with persistent GI bleed or bowel obstruction due to intussusception (4–7,9,10). Of the 12 pediatric lymphangiomas identified in the retrospective review by Lawless et al., 50% presented with acute abdominal pain, 25% with bowel obstruction, and the remaining with a palpable abdominal mass and/or increasing abdominal girth (3).

The traditional management of GI hemolymphangiomas has been surgical resection. In adults, this is being superseded by endoscopic resection which has not yet been described in pediatrics (5–7).

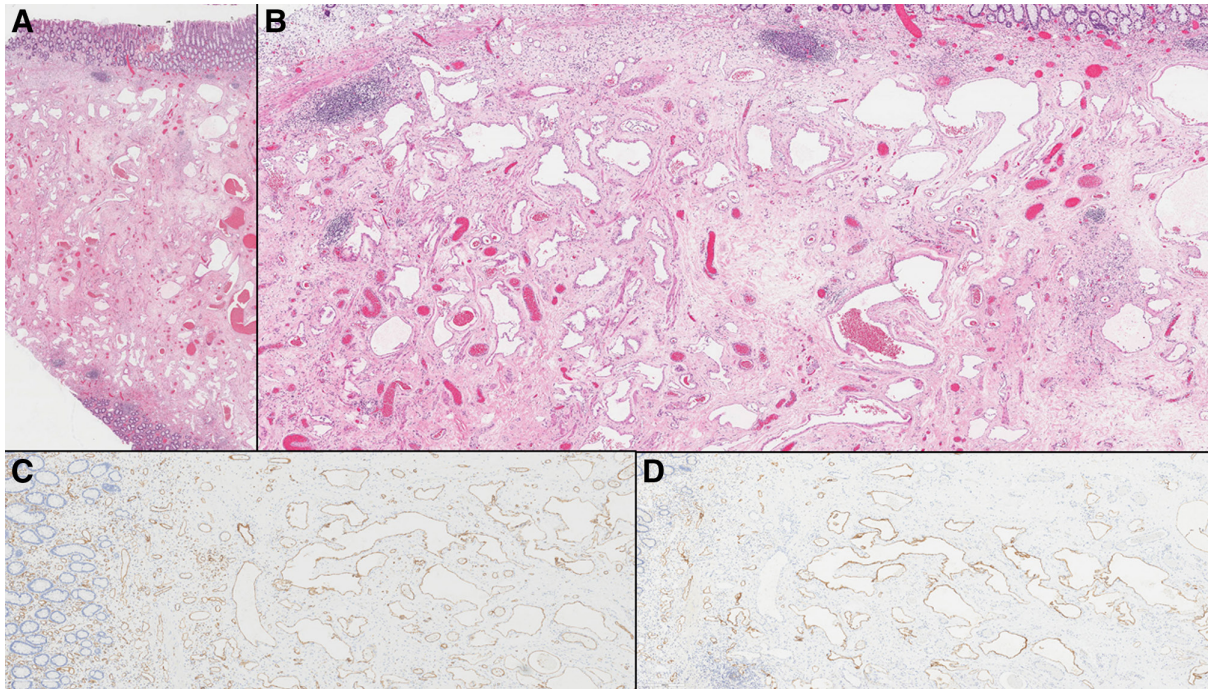

FIGURE 3. Histopathology of polypoid lesion. A) (H&E 4X) and B) (H&E 10X). Submucosal proliferation of the multiple, variable-sized and irregularly-shaped vessels with patchy extension into the lamina propria. C) Endothelium of all vessels is positive for immunohistochemical marker CD31 (10X). D) Endothelium of some vessels (lymphatics) is positive for immunohistochemical marker D2-40 (10X).

Although endoscopic polypectomy does carry inherent risks, including bowel perforation, bleeding, and incomplete resection, the endoscopic approach is less invasive and associated with decreased time for patient recovery compared to surgical bowel resection. Therapeutic endoscopic intervention has been described in patients with chronic GI bleeding (5–7), but successful management of an obstructive-symptomatic hemolymphangioma with polypectomy in a child has not been reported. Our patient underscores the feasibility of polypectomy as a safe and noninvasive approach that may prove curative in children.

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