

# Primary Intestinal Lymphangiectasia Causing Intussusception and Small Bowel Obstruction

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## ABSTRACT

Abdominal lymphangiectasia is a rare disease manifestation with a variety of anatomic locations and clinical presentations. The gastrointestinal tract may be affected, and lymphangiectasia originating in the wall of the intestine has rarely been described. We present a case of primary small bowel lymphangiectasia causing intussusception in a 30-year-old woman who presented with emesis and gastrointestinal bleeding. This case emphasizes the clinical presentation, diagnosis, and management in adults with abdominal lymphangiectasia. We highlight the importance of a high clinical suspicion for lymphangiectasia in an adult with acute abdomen to avoid catastrophic morbidity.

## INTRODUCTION

Lymphangiectasia is a benign malformation of the lymphatic system, characterized by focal or diffuse dilation of the mucosal, submucosal, and subserosal lymphatic vessels. Lymphangiomatosis refers to the presence of diffuse lymphangiectasia in an organ or across multiple organ systems.<sup>1</sup> Solitary lymphangiectasia rarely occurs in the abdomen, presenting predominantly in the mesentery with gastrointestinal bleeding and abdominal pain.<sup>1,2</sup> We report a case of primary intestinal lymphangiectasia presenting with intussusception and small bowel obstruction.

## CASE REPORT

A 30-year-old woman presented with nausea, vomiting, hematochezia, and severe abdominal pain for 2 days. She reported an extensive family history of inflammatory bowel disease (IBD). She denied dysphagia, fevers, weight loss, previous hematochezia, acid peptic disease, gastrointestinal reflux, or recent alcohol or drug use. Vital signs and the hemoglobin level were stable. Abdominal contrast-enhanced computed tomography (CECT) revealed pancolitis and a small bowel intussusception with proximal distention; no mass was seen (Figure 1). Obstructive symptoms relieved after 2 days of supportive management. Endoscopy performed afterward displayed a subepithelial small bowel lesion with a smooth mucosal surface causing luminal narrowing, suggesting an intussusception lead point in the distal jejunum with small bowel obstruction (Figure 2). Colonoscopic biopsy confirmed IBD with cryptitis and acute-on-chronic inflammation. Her inflammatory symptoms, likely related to underlying IBD, resolved with a course of mesalamine.

She underwent surgical resection of the small bowel mass seen on endoscopy. Exploratory laparotomy was unable to locate the mass on a standard bowel run. Intraoperative enteroscopy was performed through an enterotomy in the midjejunum, unveiling an enlarged fold with a protruding surface approximately 10 cm distal to the ligament of Treitz. The mass was resected with adequate margins, and the small bowel was closed without complication. Anatomic examination showed a 1.3 × 1.0 cm yellow-tan, polypoidal mass-like lesion. Histologic examination confirmed dilated lymphatic channels in the submucosa consistent with lymphangiectasia (Figure 3). She resolved clinically and was discharged in a stable condition.



**Figure 1.** Abdominal contrast-enhanced computed tomography showing a left-sided small bowel intussusception and moderate gastric distention.

## DISCUSSION

Lymphangiectasia is a rare, benign cystic lesion of endothelium-lined lymphatic spaces with surrounding stromal and connective tissue.<sup>3</sup> It most commonly occurs in children in the first few years of life and present in many anatomic locations.<sup>4</sup> The majority (>90%) occurs in the neck or axillary regions, whereas the remainder occurs in the lung, mediastinum, and abdomen.<sup>3</sup> Gastrointestinal lymphangiomas have been reported to occur with origin in the mesentery, but lymphatic malformation with primary small bowel origin has rarely been described.<sup>4-13</sup> To our knowledge, this is the first reported case of primary small bowel-originating lymphangioma causing intussusception.

Lymphangiectasia is thought to originate from a congenital or acquired malformation of lymphatic vessels, forming a cystic area of dilated vascular spaces.<sup>7,8</sup> Severity can range from solitary masses (ie, lymphangioma). Our patient had focal jejunal lymphangiectasia with sparing of the surrounding mucosa. Although most occur congenitally, they can be acquired because of abdominal trauma, inflammation, radiation, or abdominal surgery.<sup>14,15</sup> Our patient did present with pancolitis because of previously undiagnosed IBD. Although the etiology in our case is

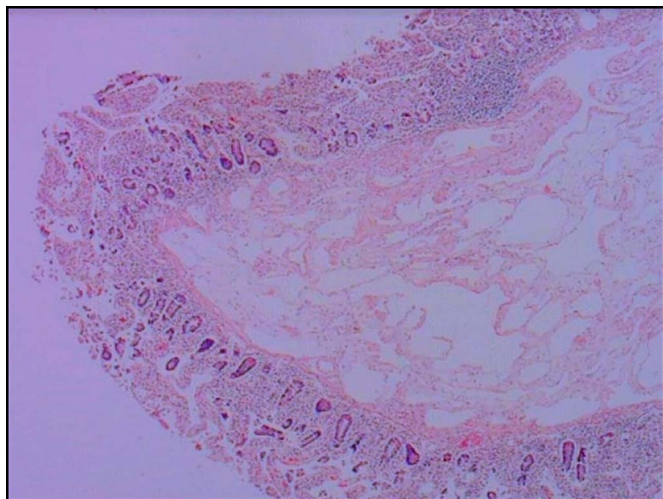


**Figure 2.** Capsule endoscopy revealing a protruding lesion in the jejunum, approximately 10 cm distal to the ligament of Treitz.

unclear, ongoing bowel inflammation may have triggered the formation of the lymphangiectasia, which resulted in this acute clinical presentation.

Signs and symptoms of this lesion vary. The most severe variant, primary intestinal lymphangiomatosis, is characterized by diffuse involvement of the small bowel lymphatics, resulting in a protein-losing enteropathy.<sup>16</sup> Reported cases describe complications of mesenteric volvulus and perforation.<sup>7,8,17</sup> Our patient presented with intussusception and upstream bowel distention, emesis, and gastrointestinal bleeding. Only 5% of intussusceptions occur in adults, commonly because of adhesions, metastatic or primary neoplasm, feeding tubes, or idiopathic etiology.<sup>18</sup> It is thought to occur from a proximal bowel segment “telescoping” into a distal bowel segment using a “lead point.” Our patient’s lymphangiectasia may have acted as a lead point in the distal jejunum to induce intussusception.

Lymphangiectasia may be evaluated with several imaging modalities. Ultrasound can reveal a uniloculated or multiloculated, anechoic cystic mass with possible wall contrast enhancement on CECT.<sup>19</sup> However, cystic lesion on imaging is not diagnostic of lymphangiectasia because its appearance may mimic organ cysts, carcinomas, and abscess.<sup>19</sup> A mass was not seen on CECT for our patient, likely for several reasons. First, the lymphangioma was small and found to be embedded in the submucosa. Second, any visible luminal mass on CECT would have been hidden in the bulk of the intussusception. Therefore, endoscopic evaluation is likely necessary if symptomatic lymphangiectasia is suspected.



**Figure 3.** Microscopic analysis of the resected lesion revealing a region of dilated lymphatic channels limited to the intestinal submucosa. There was no mucosal involvement or invasion.

Surgical resection is effective in relieving symptoms with little disease recurrence.<sup>10</sup> Underlying gastrointestinal disorders should be adequately treated to promote recovery and minimize inflammation. Our patient did not report return of any symptoms at 2 follow-up visits.

In conclusion, primary intestinal lymphangiectasia is a rare lesion that can predispose to small bowel intussusception with resulting obstruction. Intussusception in an adult should raise clinical suspicion for primary lymphangiectasia; appropriate clinical management with supportive care and surgical resection may avoid the grave consequences of peritonitis and vascular compromise.

## DISCLOSURES

**Author contributions:** All authors contributed equally to this manuscript. Y. Zafar is the article guarantor.

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Informed consent was obtained for this case report.

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