DIAPHRAGM DISEASE OF THE SMALL BOWEL PRESENTING WITH INTUSSUSCEPTION

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Introduction:

The use of NSAIDs is very common, especially in those with chronic pain and rheumatological disorders. NSAIDs have been known to have adverse effects on many organ systems including the gastrointestinal tract, renal system, and cardiovascular system. Diaphragm disease of the small bowel is a rare disorder caused by chronic NSAID use, in which strictures form throughout the small intestine, occasionally resulting in bowel obstruction. Given the rarity of the disease as the cause for small bowel obstruction, diaphragm disease is difficult to diagnose without histopathological analysis. Physicians who are familiar with the symptoms of this disease and its potential complications are at an advantage for correctly identifying the disorder and performing the necessary steps for treatment.

Case Presentation:

A 73-year-old female with history of anemia and chronic back pain (for which she used NSAIDs frequently) presented with nausea, vomiting, abdominal pain, diarrhea, and weight loss. She was initially diagnosed with collagenous colitis and underwent multiple endoscopies over several years. On EGD, she had mid-esophageal, pyloric and duodenal strictures with ulcerations that were dilated. Recent colonoscopy showed an ascending colon ulcer and chronic active ileitis concerning for Crohn's disease. She began treatment for Crohn's disease, but her symptoms persisted. She then underwent video capsule endoscopy, which demonstrated numerous small bowel ulcers and inflammation consistent with Crohn's; however, the PillCam did not reach the colon. After nearly a month without evacuation of the PillCam, CT enterography was performed and showed intussusception with possible small bowel mass. She continued to have abdominal pain, nausea, and vomiting over the next few months. Despite having bowel movements, she did not pass the PillCam.

The decision was made to proceed with diagnostic laparoscopy with intraoperative push endoscopy through the planned enterotomies. A wound protector was used to extracorporealize the bowel (Figure 1). Enterotomies were made and push endoscopy was performed until the area of intussusception and PillCam was palpated and visualized. The intussusceptum contained a 10 cm mass. Another enterotomy was made distal to the mass and push endoscopy was continued to the cecum with removal of the PillCam (Figure 6) and one polyp. A total of 14 strictures were visualized (Figures 2-5). The small bowel was resected to include the entirety of the strictures, the area of intussusception and the presumed mass. A functional side-to-side anastomosis was created, and the patient did well in the postoperative period. Pathologically, the specimen was identified as a large inflammatory small bowel fibroid polyp with associated mucosa ulceration, serositis and serosal adhesions consistent with intestinal intussusception and multiple diaphragm disease lesions. The patient was educated on cessation of NSAID use to prevent recurrence of stricture formation.



Figure 1. Intraoperative photo of intussusception

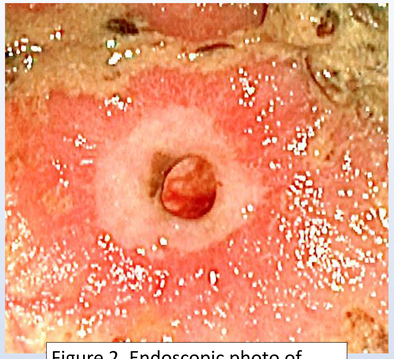


Figure 2. Endoscopic photo of small bowel diaphragm stricture



Figure 3. Endoscopic photo of ulcerated diaphragm stricture

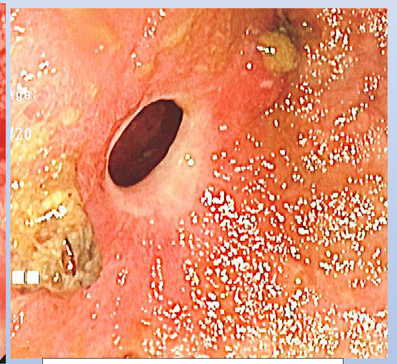


Figure 4. Endoscopic photo of small bowel diaphragm stricture

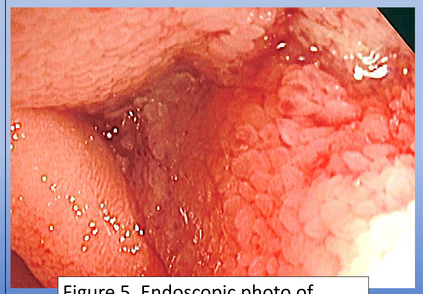


Figure 5. Endoscopic photo of intussuscepted diaphragm stricture



Figure 6. VCE retrieved intraoperatively

Discussion:

Small bowel diaphragm disease is a rare cause of small bowel obstruction that is often misdiagnosed due to the rarity of the condition. The appearance of the small bowel lumen as concentric rings of fibrotic tissue helps define the term 'diaphragm' disease [6,8], and the diagnosis is made via history, gross appearance and pathological examination. Stricturing of the small bowel is seen in a variety of conditions [1], thus the diagnosis of diaphragm disease requires exclusion of all other causes of stricture formation. Risk factors include advanced age, chronic NSAID use [5,6], and female gender [4]. Many cases had an average use of NSAIDs for at least 3-5 years, though strictures have developed in as little as 2 months of NSAID usage [2,4]. Of chronic NSAID users, it is suspected that about 2% will develop small bowel diaphragm disease [6,7,9]. Presenting symptoms include chronic abdominal pain, iron deficiency anemia, weight loss, and constipation. If symptoms develop more acutely, imaging often shows findings concerning for adhesions or a mass causing small bowel obstruction [5]. Intussusception has not been explicitly discussed in literature, as in this unique case. Intraoperatively, strictures can be misdiagnosed due to the rarity of the disease. Without its consideration as a differential diagnosis, physicians may not make the appropriate decisions in treatment.

The effect of chronic NSAID use on the small bowel was first discovered histologically in 1988 [5,6]. Characteristic findings include thickening of the muscularis mucosa, fibrous change to the lamina propria, eosinophilic mucosa, and ulceration of the diaphragm [1]. Diaphragm disease can be distinguished histologically from inflammatory bowel disease by its typical limitation to the mucosa and submucosa, without full thickness disruption [2]. It can cause thickening of the muscularis mucosa without penetrating the serosa [2,10]. It is believed that NSAIDs cause increased susceptibility of the small bowel mucosa to the inflammatory responses of bacteria and digestive enzymes by inhibiting prostaglandins and hindering the intestinal villous microcirculation [5,7,8]. This results in inflammatory circumferential injury to the submucosa, forming ulcerations which scar and stricture lumen when healed [5].

Balloon-assisted enteroscopy, video capsule endoscopy, CT and MRI enterography can show nonspecific strictures [11]. MRI enterography can provide better evaluation and assessment for strictures than CT imaging. If the diagnosis is made early, surgery can sometimes be avoided, and patients can be treated with endoscopic balloon dilatation of the small bowel, in addition to cessation of NSAID use [6,7]. Diagnostic laparoscopy may reveal the point of obstruction, but laparotomy facilitates palpation and tactile appreciation of the strictures, which may be multiple [7,10]. Intraoperative enteroscopy is recommended if diaphragm disease is suspected as it allows for intraluminal assessment of further diseased segments of the small bowel that should be resected to prevent recurrence of symptoms [7]. Stricturoplasty of the affected areas of the small bowel is often undertaken due to misdiagnosis or when the extent of small bowel involvement makes resection unfavorable [1]. Resection remains the optimal treatment in most situations as it is the most definitive management to prevent recurrence and confirm the diagnosis.

Conclusion:

Histopathological analysis of the intestinal lumen of the small bowel is often necessary to correctly diagnosis diaphragm disease. Chronic NSAID use appears to cause the stricturing that is often missed on diagnostic laparoscopy but palpated on laparotomy and visualized during enteroscopy. The presenting symptoms are nonspecific for diaphragm disease, but relate to the partial or intermittent small bowel obstruction. Surgical resection is often undertaken when patients present acutely with obstructive symptoms, nonetheless stricturoplasty and balloon dilatation have been shown to be effective treatment options when combined with NSAID cessation. This case demonstrates that physicians must be mindful of this disorder and the wide range of effects that NSAIDs can have on the bowel to avoid unnecessary surgery in patients with diaphragm disease.

References:

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