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CASE REPORT | INFLAMMATORY BOWEL DISEASE

Fistulizing and Stricturing Esophageal Crohn's Disease Requiring Esophagectomy

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ABSTRACT

Esophageal Crohn's disease is uncommon and difficult to identify, especially in the adult population. In this study, we report a biologic-naïve patient with colonic Crohn's disease complicated by recurrent esophageal strictures despite many dilations, who presented to our center with recurrent aspiration pneumonia. He was found to have a tracheoesophageal fistula as the likely etiology. After multidisciplinary discussion, he underwent esophagectomy given the severity of his stenosis. The patient's surgical course was complicated by dysphonia and aspiration due to unilateral vocal cord paralysis, which resolved with vocal cord injection. This case highlights a severe manifestation of esophageal Crohn's disease.

KEYWORDS: Crohn's disease; esophagectomy

INTRODUCTION

Upper gastrointestinal tract Crohn's disease (CD-UGI) has a prevalence of 0.5%–4% in adults, compared with 30%–50% in children.[1,2] Adult prevalence is likely underestimated because—unlike in pediatrics—adults with inflammatory bowel disease (IBD) do not routinely undergo esophagogastroduodenoscopy (EGD) at the time of diagnostic colonoscopy. A study where EGDs were performed on adults with CD showed a prospective incidence of 16% for CD-UGI where 37% of study patients had UGI symptoms at the time of EGD.[3] Within CD-UGI, there is even less data on esophageal CD because symptoms overlap with other causes of erosive esophagitis and histologic findings are nonspecific. In this study, we present the case of a patient with fistulizing and stricturing CD affecting the esophagus and colon, who ultimately required esophagectomy after developing a tracheoesophageal fistula (TEF).

CASE REPORT

The patient is a 24-year-old man with fistulizing and stricturing Crohn's disease with esophageal, colonic, and perianal involvement complicated by esophageal strictures requiring multiple dilations, who presented to Stanford Hospital with recurrent aspiration pneumonia.

He was diagnosed with ulcerative pancolitis at the age of 16 years at an outside facility after having persistent diarrhea following campylobacter enterocolitis; he had an EGD at diagnosis, which was unremarkable. The patient was treated with mesalamine, then escalated to mercaptopurine (6-MP) and prednisone at the age of 18 years due to ongoing diarrhea. He also developed dysphagia at the age of 18 years. EGD showed an esophageal stricture requiring dilation, and biopsies showed nonspecific esophagitis. At that time, his diagnosis was changed to stricturing CD with esophageal and colonic involvement. He underwent ~40 dilations between ages 18–22 years; 1 dilation was complicated by an esophageal perforation, which was managed conservatively. At the age of 19 years, he stopped his 6-MP and prednisone due to nausea and vomiting, instead managing his CD with homeopathic therapies. His last preadmission colonoscopy was at the age of 21 years, which showed severe pancolitis, a tubular colon with scattered pseudopolyps, and a nondraining perianal fistula. Biopsies showed acute cryptitis, crypt abscesses, and crypt distortion throughout the colon and

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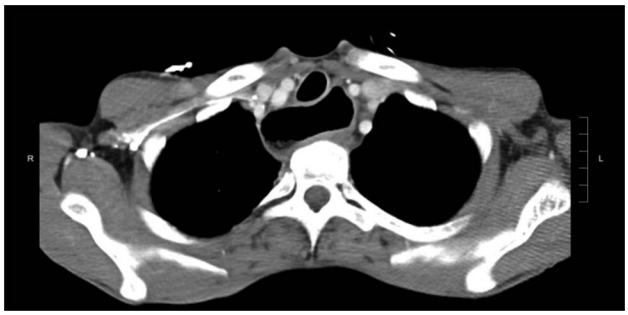

Figure 1. Axial slices of chest computed tomography showing a dilated upper esophagus (inferior to the trachea).

a normal terminal ileum. At the age of 22 years, magnetic resonance enterography showed normal small bowel and a tubular colon with loss of haustrations. He was subsequently lost to follow-up.

At the age of 24 years, he presented to our institution with shortness of breath secondary to aspiration pneumonia—his fourth episode in a year. Gastroenterology was consulted due to concern for esophageal pathology causing aspiration. On history, he reported 2–3 formed, nonbloody bowel movements per day and denied dysphagia or odynophagia. He was tolerating a plant-based diet. His body mass index was 13 kg/m^2, and examination was notable for a benign abdomen. His albumin was 2.8. Computed tomography of the chest showed a markedly dilated air and debris filled upper esophagus measuring up to 5.7 cm with layering intraluminal fluid (Figure 1). EGD showed a loss of clear esophageal lumen with multiple false lumen outpouchings (Figure 2). There was no endoscopic evidence of inflammation. During the EGD, the camera became clouded with the patient's breathing, and the end tidal carbon dioxide reading increased with insufflation of the outpouchings, raising concern for a TEF. Pathology showed squamous mucosa with no significant histopathological abnormality. A barium esophagogram showed tracheal esophageal fistula originating from a suspected upper/mid esophageal stricture (Figure 3).

Given the severity of the esophageal findings, it called into question whether the patient had CD with esophageal and

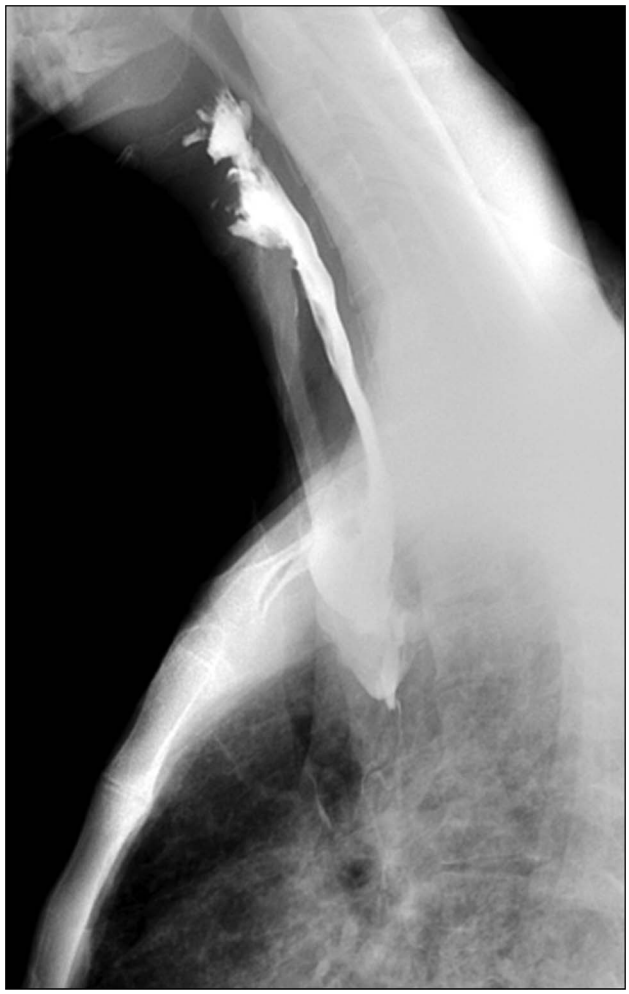

Figure 3. Barium esophagram showing a tracheoesophageal fistula.

colonic involvement, or ulcerative pancolitis with a non-IBD reason for the TEF. With regards to alternative diagnoses for his TEF, we considered his prior recurrent endoscopic interventions, caustic (alkali) ingestion, or infections, such as tuberculosis. On history, the patient had not had a caustic ingestion, and on testing, he had an indeterminate QuantiFERON Gold with 3 cultures negative for acid fast bacilli. In this setting, we felt that his

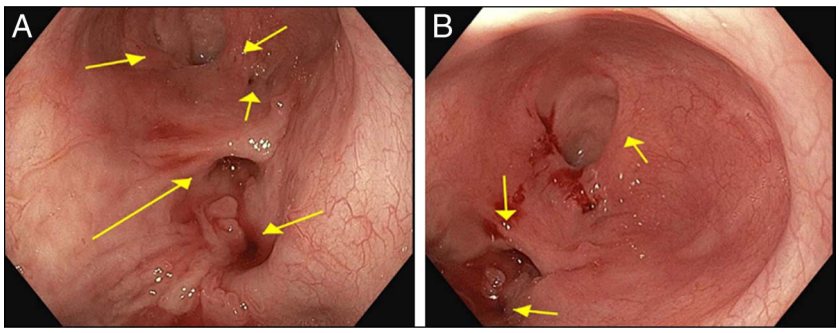

Figure 2. (A and B) Loss of clear esophageal lumen with multiple outpouchings seen on esophagogastroduodenoscopy. Arrows are pointing to the outpouchings seen during the esophagogastroduodenoscopy.

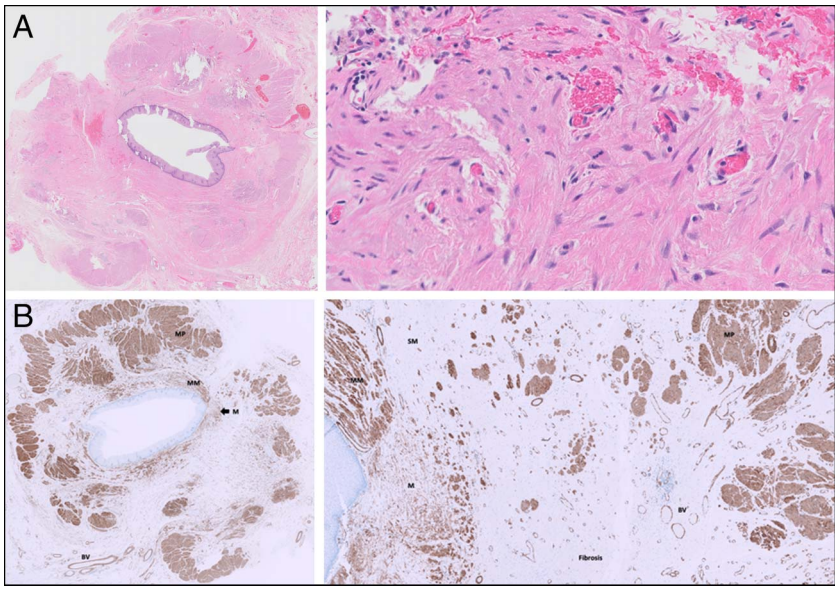

Figure 4. (A) On the left is an H&E stain at 5× magnification—esophagus involved by Crohn's disease showing extensive scarring of muscularis mucosae, submucosa, and muscularis propria with narrowing of lumen. On the right is an H&E at 400× magnification—nearly all cells in field represent myofibroblasts with mitotic figure in center. (B) On the left is an actin smooth muscle antibody (1A4) immunohistochemistry at 5× magnification showing disrupted MM and MP and with separation of muscle bundles by fibrosis (which do not stain). Blood vessels and a cluster of residual M also stain. On the right is an actin smooth muscle antibody (1A4) immunohistochemistry at 50× magnification showing extensive fibrosis (white background throughout picture) involving MM, MP, and submucosa. A cluster of residual M remains. H&E, hematoxylin and eosin; M, myofibroblasts; MM, muscularis mucosae; MP, muscularis propria.

presentation was consistent with CD with esophageal and colonic involvement.

Following these diagnostic studies, a multidisciplinary discussion was held between gastroenterology, IBD, therapeutic endoscopy, and thoracic surgery. The IBD team felt that while immunologic therapy may help his colonic disease, it would be unlikely to heal a tracheoesophageal fistula.[4–6] In addition, there was concern about starting him on immunosuppression given recurrent necrotizing pneumonias. The therapeutic endoscopy team felt that further dilation or stent placement would not be beneficial. Therefore, the decision was made for him to undergo nutritional optimization followed by esophagectomy. A surgical jejunal feeding tube (J-tube) was placed to address his severe protein calorie malnutrition. After several months of enteral feeding through the J-tube, the patient underwent a modified McKeown esophagectomy.

Surgical pathology showed marked mural fibrosis and severe stricture with myofibroblasts in the esophagus (Figure 4). There was no inflammation in the excised esophagus or stomach. Intestinal fibrosis progresses from extracellular matrix deposition in the submucosa to hyperplasia of the muscularis mucosa to extracellular matrix deposition in the muscularis mucosa.[7] Therefore, the absence of inflammation does not rule out active Crohn's disease. In addition, 11 lymph nodes were resected during the esophagectomy without significant abnormality. His immediate postoperative course was complicated by an anastomotic leak in the cervical esophagus and aspiration and dysphonia due to unilateral vocal cord paralysis, which resolved with carboxymethylcellulose gel injection. At a visit with thoracic surgery 2 months postoperatively, the patient was continuing with enteral nutrition through the J-tube to allow the anastomotic leak to heal.

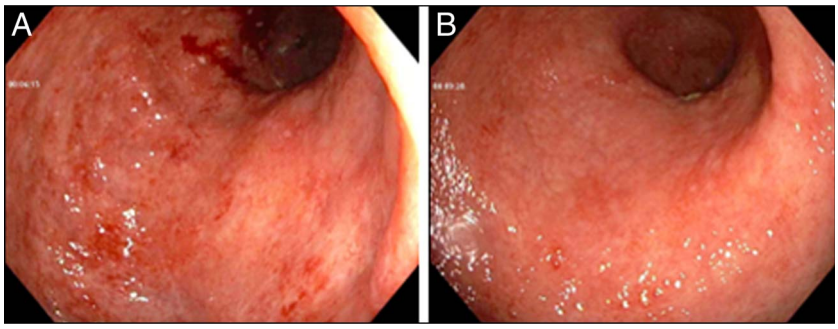

Figure 5. (A and B) Colonoscopy performed 4 months after initiating enteral nutrition, showing ulcerations throughout the right-sided and left-sided colon, respectively.

His colonic CD, which had been dormant on a plant-based diet, unfortunately reactivated with enteral nutrition. After surgery, he underwent colonoscopy, which showed pancolitis with biopsies demonstrating moderately active colitis (Figure 5). Once the anastomotic leak resolves, plans have been made to initiate an advanced immune therapy.

DISCUSSION

This case describes a patient with CD who required esophagectomy due to severe stricturing of the esophagus, leading to TEF formation. Prior endoscopic findings of esophageal CD included erythema, erosions, and ulcerations most commonly, with strictures and fistulas appearing more rarely. Prior pathologic findings showed lymphocyte and plasma cell predominance; intraepithelial cells or noncaseating granulomas were rare. Prior treatments for fistulizing and stricturing esophageal CD include a combination of biologic therapies, proton-pump inhibitors, and serial dilations and/or endoscopic stenting.[8] Patient outcomes between endoscopic stenting and esophagectomy have not been studied. Recurrence rates after esophagectomy are unknown, given the paucity of patients who undergo the surgery for esophageal Crohn's disease.[5,9,10] We report a markedly severe presentation of esophageal Crohn's disease with stricturing and TEF requiring esophagectomy. Given the low incidence of esophageal Crohn's disease, this case contributes to the literature in terms of endoscopic and pathologic presentation, as well as surgical management options, in patients with fistulizing and stricturing esophageal Crohn's disease.

DISCLOSURES

Author contributions: C. Dimopoulos-Verma: Data acquisition and interpretation, drafting the work, and final approval of version to be published. A. Ott: Data acquisition, reviewing the work, and final approval of version to be published. A. Yeoh: Data acquisition and interpretation, reviewing the work, and final approval of version to be published. M. Barakat: Data acquisition and interpretation, reviewing the work, and final approval of version to be published. D. Bingham: Data acquisition and interpretation. K. Keyashian: Data acquisition and interpretation, reviewing the work, and final approval of version to be published. G. Barber: Data acquisition and interpretation, reviewing the work, and final approval of version to be published and is the article guarantor.

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Informed consent was obtained for this case report.

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