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IMAGE | ESOPHAGUS

Esophageal Crohn's Disease Unresponsive to Glucocorticoids

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CASE REPORT

A 24-year-old woman presented with a 1-week history of odynophagia and epigastric abdominal pain. One month before presentation, the patient was diagnosed with severe ileocolonic Crohn's disease (CD) and started on glucocorticoids (prednisone 40 mg per oral daily). Esophagogastroduodenoscopy revealed numerous esophageal "punched-out" ulcerations and erosions throughout the distal and mid esophagus (Figure 1). Biopsy revealed chronic inflammation with lymphoplasmacytic infiltration of the squamous mucosa (Figure 2). Immunohistochemical stains for cytomegalovirus and herpes simplex virus were negative. Esophageal CD was highly suspected. Infliximab 5 mg/kg intravenous every 8 weeks and azathioprine 50 mg per oral daily were started and glucocorticoids were gradually tapered. Upper gastrointestinal symptoms resolved 2 months after beginning the combination therapy. Repeat esophagogastroduodenoscopy at 6 months revealed esophageal ulcer healing with residual scarring (Figure 3).

Esophageal CD is a rare entity, with an estimated prevalence of 0.3%–10% in adults with CD.[1] The most common presenting symptoms are odynophagia, dysphagia, heartburn, and chest pain.[2] Esophageal CD is rarely the initial disease presentation and rarely presents concomitantly with intestinal CD. The median interval between the diagnosis of intestinal and esophageal CD is approximately 1–3 years.[1]

A retrospective review of 20 patients with esophageal CD by Decker et al found that luminal disease activity occurred most commonly in the distal esophagus alone (80%), followed by the distal and mid esophagus (15%) and pan esophagus (5%).[3] Common

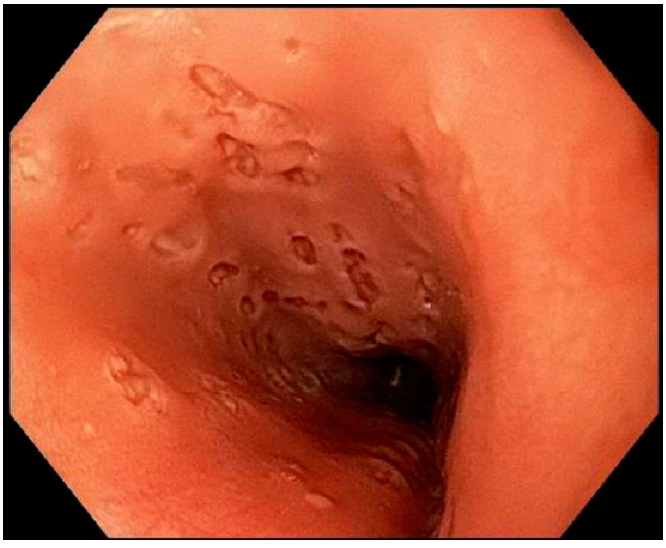

Figure 1. Numerous punched-out ulcers and erosions in the esophagus.

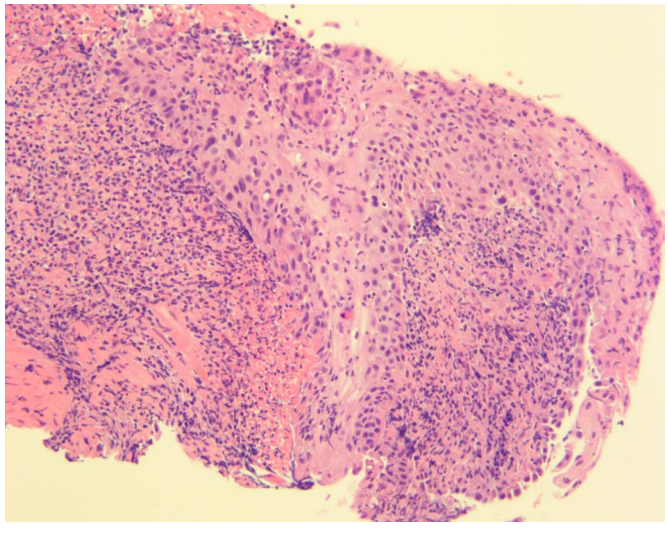

Figure 2. Lymphocytes and plasma cells in the submucosa of the esophagus.

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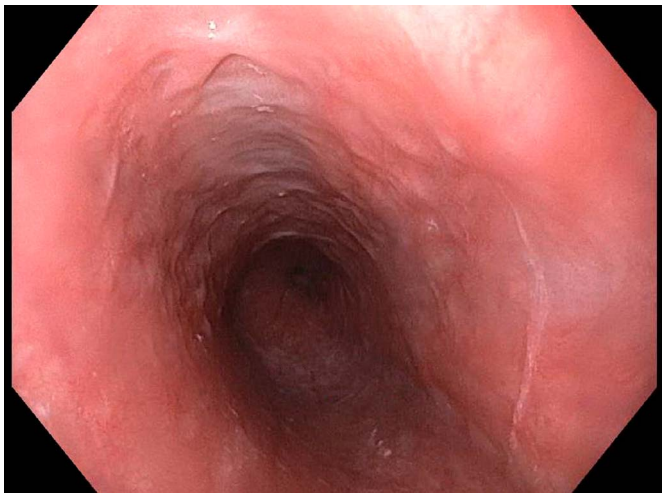

Figure 3. Posttreatment endoscopy showing residual scarring after healing of ulcers.

endoscopic findings include aphthous ulcers, serpiginous ulcers, nodules, pseudopolyps, skip lesions, and, in severe cases, strictures and fistulae.[1] Esophageal CD can be classified into 3 types: inflammatory, structuring, and fistulizing.[6] Chronic inflammation is the most common histologic finding, and non-caseating granulomas are seen in only 7%–9% of cases. A combination of clinical, endoscopic, and pathologic findings is required to make the diagnosis.[4]

There are no treatment guidelines specific for esophageal CD. Small retrospective studies have shown that esophageal CD responds to steroids with complete resolution within 2–4 weeks of therapy.[5] In patients who become steroid dependent or respond inadequately to steroids, immunomodulators have shown to produce a good clinical response. There have been a few case reports in which biologics like infliximab and adalimumab were used to achieve clinical remission in esophageal CD.[2] Treatment of esophageal CD may need to be tailored individually with dilation for strictures, antibiotics for fistulae, etc., in addition to medical therapy. There are not many articles highlighting the importance of combination therapy for esophageal CD, which makes our case unique. In a retrospective observational study, De Felice et al reported using combination therapy in patients with esophageal CD.[6] They recommended early aggressive combination therapy in esophageal CD. We conclude that early combination therapy should be strongly considered in proximal CD.

DISCLOSURES

Author contributions: T. Kochar reviewed the literature and wrote and edited the manuscript. M. Krafft wrote and edited the manuscript. S. Gayam reviewed the literature, edited the manuscript, and is the article guarantor.

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