Aphthous esophageal ulceration: A novel presentation of Crohn's disease?

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PL Beck, PK Bluestein, MA Andersen. Aphthous esophageal ulceration: A novel presentation of Crohn's disease? Can J Gastroenterol 1994;8(2):101-104. Crohn's disease (CD) commonly presents as involvement of the small bowel or colon – the esophagus is rarely involved. The authors describe the case of a 45-year-old woman who presented with odynophagia and was found to have aphthous ulcers of the esophagus. On questioning, she admitted to three to four episodes of nonbloody diarrhea, each lasting less than one week, over the past two years. The patient denied other symptoms of gastrointestinal, collagen-vascular, respiratory or cardiovascular disease. Viral, fungal and bacterial cultures of blood, stool and tissue biopsies were all negative. A small bowel enema showed inflammation of the distal ileum. Colonoscopy revealed patchy areas of inflammation of the colon and distal ileum. Biopsies from the esophagus, duodenum, terminal ileum and colon showed chronic inflammation, lymphoid aggregates, goblet cell hyperplasia (in the colon) and crypt abscesses. Giant cells, granulomas and fissures were not evident in any of the biopsies. The patient failed to respond to a three-week course of omeprazole, but her symptoms resolved within three days of starting prednisone. The exact cause of the esophageal ulcerations is unknown but most likely is CD of the esophagus.

Key Words: Crohn's disease, Esophageal ulcers

Ulcérations esophagiennes aphteuses: nouvelle présentation de la maladie de Crohn?

RÉSUMÉ : La maladie de Crohn se manifesterait généralement par une atteinte de l'intestin grêle ou du côlon, l'œsophage étant rarement affecté. Les auteurs décrivent le cas d'une femme de 45 ans qui s'est présentée avec l'odynophagie et chez qui des ulcères aphteux de l'œsophage ont été observés. À l'interrogatoire, elle dit avoir présenté à trois ou quatre occasions de la diarrhée non sanguinolente qui a duré moins d'une semaine, au cours des deux années écoulées. La patiente a déclaré n'avoir aucun autre symptôme de maladie digestive, vasculaire-col...
prior à l'œdème pharyngé, elle avait une légère diarrhée. Les examens sanguins étaient normaux. Elle a été d'accord pour se soumettre à des examens de biopsie, notamment de la peau et du rectum. Les résultats de l'examen histologique montraient une infiltration inflammatoire typique de la maladie de Crohn.

La patiente a été hospitalisée à cause de douleurs abdominales persistentes. Les examens diagnostiques ont révélé une perforation gastro-intestinale et un abcès abdominal. Les examens de biopsie ont montré des anomalies inflammatoires. Le patient a été hospitalisé à cause de douleurs abdominales persistantes. Les examens diagnostiques ont révélé une perforation gastro-intestinale et un abcès abdominal. Les examens de biopsie ont montré des anomalies inflammatoires.

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Esophageal ulcers and CD

Figure 1) Aphthous ulcerations of the esophagus noted on initial gastroscopy. The ulcers were present throughout the esophagus.

Figure 2) Histological appearance of one of the esophageal ulcers. The inflammatory cell infiltrate primarily consisted of macrophages, lymphocytes and fibroblasts. Both light and electron microscopy failed to identify an infectious agent in multiple biopsies from the esophagus, duodenum, ileum and colon.

Figure 3) Radiograph following a small bowel enema. Note the inflammatory changes in the distal ileum (arrows).

orally per day) was then started. Three days later, the patient was asymptomatic and returned to a full diet. The prednisone was tapered over a period of six weeks. Gastroscopy after six weeks of therapy showed no signs of inflammation or ulceration of the esophagus, stomach or proximal duodenum. She has been followed for two years and has been asymptomatic.

DISCUSSION

The exact etiology of the esophageal ulcers in this case is unknown. The lack of other symptoms and laboratory data to suggest an infectious or a collagen-vascular cause, the presence of diarrhea, and chronic inflammation of the duodenum, distal ileum and colon all point to CD. Esophagitis in Behçet’s disease is rare and is usually not due to aphthous ulcers (1). Of 26 reported cases of esophagitis in Behçet’s disease, 42% had ileocolonic ulcerations (1). The presented case did not have a history of rheumatological or eye problems, and denied skin rashes, and oral and genital ulcers. The diagnostic criteria of Behçet’s disease include: major criteria of buccal ulcerations, genital ulcerations, eye lesions and skin lesions; and minor criteria of arthritis, a positive family history, thrombophlebitis, and lesions involving the gastrointestinal, cardiovascular and neurological systems (21). A minimum of three major and two minor criteria are required for the diagnosis; thus, this case does not fulfil the diagnostic criteria (21). Further, there were no features to suggest infectious or collagen-vascular disorders.
Esophageal ulceration in UC occurs rarely and is usually associated with longstanding severe disease (5,6). In the present case, the findings on colonoscopy, barium enema and histological examination were not suggestive of UC.

Esophageal involvement in CD is usually associated with advanced ileocolonic disease and an increased prevalence of the extraintestinal manifestations of CD (10,15). As aforementioned, esophageal ulceration due to CD has been reported in two individuals who had no other evidence of CD on presentation with esophageal symptoms (16,19). Geboes et al (10) followed 500 patients with biopsy-proven CD and found esophageal ulcers in nine patients (1.8%), which probably was an underestimate of the involvement of the esophagus because less than 20% of the patients had upper endoscopy (10). In this same study, six of the nine patients with esophageal ulcers also had aphthoid oral lesions (10).

Giant cells and granulomas were not found in any of the biopsies in our case. However, in a review of 53 cases of esophageal CD reported from 1950-88, granulomas were found in only 12 cases, five of which required several sections (10,22).

CD involvement of the esophagus most commonly presents as strictures and fistulas which are thought to be due to longstanding esophageal involvement (10,16,23,24). Two patients with esophageal CD, when followed for six and 14 years, developed severe strictures and fistulas (16).

The presented patient’s odynophagia failed to improve on omeprazole but resolved within three days of starting 40 mg/day prednisone. Esophageal ulcerations in patients with biopsy-proven CD have been found to resolve completely and rapidly to 30 to 50 mg/day prednisone (10).

Esophageal ulcers usually occur late in the course of CD. However, in one of the two cases in which esophageal involvement was the presenting feature, four years lapsed from presentation to ileocolonic involvement (16).

CONCLUSIONS

In summary, esophageal involvement can be the presenting feature of CD and, if untreated, esophageal Crohn’s can lead to debilitating complications such as fistulation, perforation, and stricture. The symptom complex of odynophagia and dysphagia in Crohn’s with esophageal involvement appears to respond rapidly and completely to corticosteroids. There has not been a clearly documented case of recurring esophageal CD. Thus, it is essential to note that esophageal ulcers can be the presenting feature of CD or can occur at any time throughout the disease course, and that corticosteroids are very effective in treating this potentially debilitating manifestation of this disease.

REFERENCES
