

# Rare esophageal ulcers related to Behçet disease

## A case report

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### Abstract

**Rationale:** The fundamental pathogenesis of Behçet disease (BD) is still unclear and controversial. Many cases of oral aphthous ulcers and genital ulcers related to BD are reported; nevertheless, idiopathic giant esophageal ulcers related to BD are rare. A rare case for esophageal ulcers related to BD is presented.

**Patient concerns:** In China, BD is represented with esophageal involvement which is called esophageal BD (EBD).

**Diagnoses:** A 56-year-old man diagnosed to the Gastroenterology Department of Integrated Traditional Chinese and Western Medicine Hospital, for multiple discrete, elliptical esophageal ulcers related to BD.

**Interventions:** The esophageal ulcers were treated with corticosteroid treatment for 12 weeks.

**Outcome:** The esophageal ulcers were cured.

**Lessons:** Our report might give further strength to avoiding the erroneous diagnosis or missed diagnosis for EBD, which is different from esophageal carcinoma, esophageal tuberculosis and esophageal Crohns disease.

**Abbreviations:** ANCA = antineutrophil cytoplasmic antibody, BD = Behçet disease, EBD = esophageal BD, UGIE = upper gastrointestinal endoscopy.

**Keywords:** corticosteroid, endoscopy, esophageal behçet disease, esophageal carcinoma, esophageal ulcer, rare

## 1. Introduction

Behçet disease (BD) is unusual, but the prevalence of BD is so high in these countries such as China, Korea, Japan, and Turkey, along the ancient Silk Road from East Asia to Mediterranean, and the lower prevalence of BD is in the United States and Northern European countries.<sup>[1-4]</sup> The oral aphthous ulcers, genital ulcers, and intestinal ulcers related to BD have been reported<sup>[5-8]</sup>; however, esophageal ulcer compatible with BD is rarely reported. The idiopathic esophageal ulcers lesion compatible with BD is still without a definite diagnostic criteria<sup>[8]</sup>; we present a report that the patient successfully is treated with prednisone therapy.

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The authors report no conflicts of interest.

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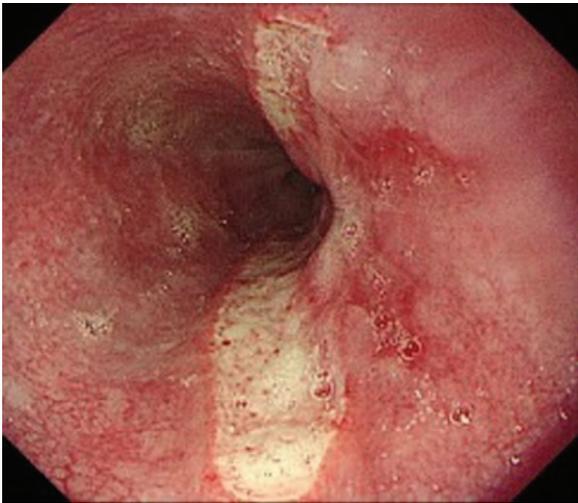
## 2. Case report

A 56-year-old man who presented with a 1-month history of dysphagia and odynophagia was taken to Tianjin Hospital of Integrated Traditional Chinese and Western Medicine (Tianjin, China). The patient lost weight for 10 kg. The patient had no other digestive syndromes containing vomiting, acid reflux, or heartburn. Furthermore, there were no positive results by routine laboratory tests but exception of erythrocyte sedimentation rate (ESR), 50 mm/h and C-reactive protein (CRP), 11.3 mg/dL. The upper gastrointestinal endoscopy (UGIE) examination showed first, a 6.0 × 2.0-cm, longitudinal, elliptical esophageal ulcer with irregular border was located 32 cm from the incisors; another 6.0 × 3.0 cm, longitudinal, elliptical esophageal ulcer with irregular border existed nearby. Around the mucosa of esophageal ulcer, congestion and edema were discovered (Fig. 1). Second, narrow band imaging (NBI) showed the appearances of two less stained areas in the center of both of ulcers (Fig. 2). We suspected that the ulcers were malignant lesions through observed results.

The patient had little sign of improving in symptoms after 2 weeks with treatment containing lansoprazole (60 mg daily) (intravenously). At the same time, the outcome of endoscopic biopsy revealed part of squamous epithelium diagnosed with low-grade intraepithelial neoplasia and chronic inflammatory reaction, but tumor cell was not discovered; all results showed that esophageal ulcers were not esophageal carcinoma (Fig. 3).

After our discussion, we considered that the appearance of longitudinal ulcers probably belonged to esophageal Crohn disease,<sup>[7]</sup> so we took him for colonoscopy, which revealed no obvious findings. We asked for a detailed medical history once again, and then the patient said he had recurrent oral aphthous ulcers.

Therefore, we performed further immunological tests including antinuclear antibodies, anti-extractable nuclear antigen antibodies, immunoglobulin A, immunoglobulin G, and



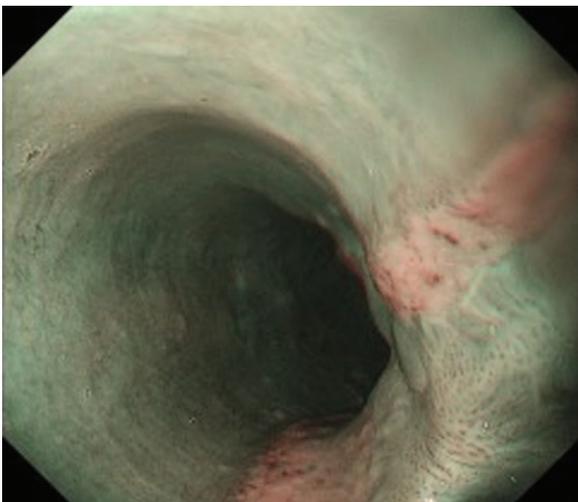
**Figure 1.** Upper gastrointestinal endoscopy showing a 6.0 × 2.0 cm, longitudinal, elliptical esophageal ulcer with irregular border located 32 cm from the incisors; another 6.0 × 3.0 cm, longitudinal, elliptical esophageal ulcer with irregular border existed nearby.

antineutrophil cytoplasmic antibody (ANCA). We found only ANCA was positive.

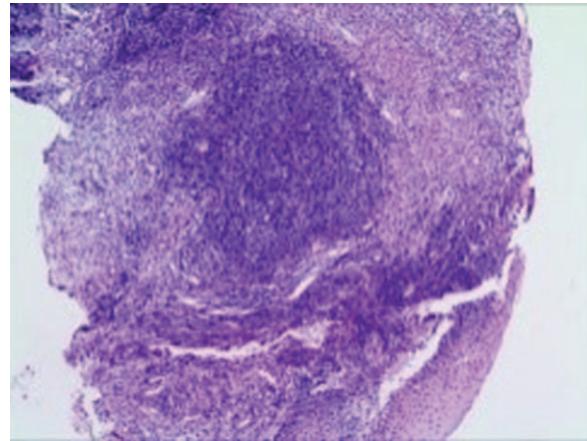
Nevertheless, the patient had no manifestations including genital ulcers, dermatitis contusiformis in the limbs, and uveal infusion. A pathology test of the skin was negative. We considered that the patient was diagnosed as incomplete-type BD. The patient was treated with prednisone (60 g) for oral administration based on empirical therapy. The patient had great sign of improving in symptoms after 4 days and symptoms completely disappeared after 1 month. We asked the patient to decrease the dose of prednisone to 5 mg daily and maintained for 2 weeks. Three months later, the esophageal ulcers were completely cured after reviewing the UGIE.

### 3. Discussion

BD is a chronic, recurrent multisystem inflammatory disorder that usually is characterized by recurrent oral aphthous ulcers,



**Figure 2.** Narrow band imaging showing 2 less-stained areas in the center of both the ulcers.



**Figure 3.** Endoscopic biopsy of ulcers with low-grade intraepithelial neoplasia, chronic inflammatory reaction (H&E 4×). An ethics committee: Tianjin Hospital of Integrated Traditional Chinese and Western Medicine's Ethics Committee approved the study. Patient consent was given.

uveal infusion, genital ulcers, and cutaneous manifestations.<sup>[1,9,10]</sup> BD patients can also have symptoms including the gastrointestinal lesions, the central nervous system, vascular lesions, articular manifestations, pulmonary systems, cardiovascular systems, and renal systems.<sup>[11]</sup> However, the fundamental pathogenesis of BD is not still proven, and at present, there was no unified diagnostic criteria including serology or diagnostic tool for BD.<sup>[5]</sup> Although intestinal BD (IBD) is reported<sup>[7]</sup>, EBD is rare reported. Although idiopathic large esophageal ulcer is often associated with esophageal carcinoma, our report is different from other reports because of the patient's benign disease process. Furthermore, the present report shows the effectiveness of prednisone therapy, which contributes to improvement in symptoms and curing ulcer is very significant.<sup>[1]</sup>

On the basis of our report, we might have a discussion on EBD and take notice of 4 aspects, which show first, we should establish the unified diagnostic criteria for BD including typical characteristic manifestations and a standard for laboratory findings such as platelet to lymphocyte ratio and anti-CTDP1 antibody.<sup>[12,13]</sup> Second, although there were no typical features for UGIE of EBD, we should pay high attention to a single or multiple, longitudinal, and elliptical ulcers, which belong to the patients who have recurrent oral aphthous ulcers, uveal infusion, genital ulcers. We should inquire detailed history of the patient and clearly know EBD is different from other diseases including esophageal carcinoma, esophageal tuberculosis, and esophageal Crohn disease. Third, treatment can be controversial and we should rationally choose a sensitive drug for EBD to relieve the symptoms. Moreover, we should pay more attention to any adverse effect of corticosteroid. According to the latest report, anti-tumor necrosis factor-alpha is a good therapeutic option.<sup>[14]</sup> Fourth, we need to pay more attention to the recurrence rate of EBD, and then, a long-term follow up will be desirable to observe whether the patient will have uveal infusion, genital ulcers, and erythema nodosum. And reviewing the UGIE is necessary for us so that we can observe whether the endoscopic biopsy has other results.

In conclusion, we need a large number of clinical cases reported and trial data to establish the diagnostic criteria for EBD. Also the pathogenesis, diagnosis, and treatment of EBD remain a challenge for our clinicians.

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