An Unusual Inaugural Manifestation of Behçet's Disease

Uma Manifestação Inaugural Incomum de Doença de Behçet

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A healthy 11-year-old boy with a family history of a father, sister, and cousin with recurrent oral aphthous lesions was admitted to the emergency department with sudden onset of solid dysphagia three days before. He denied retrosternal pain, heartburn, vomiting, odynophagia, fever, difficulty breathing or recent medication. Physical examination was normal. An upper gastrointestinal endoscopy (UGE) was performed, which showed an ulcerated esophageal mucosa in its entirety (Fig. 1). Biopsies were taken. He was hospitalized with a diagnosis of esophagitis of unknown etiology and started on acyclovir. On the third day of hospitalization, he presented oral ulcers (Fig. 2). Following the recommendation of rheumatology, he was treated with colchicine with significant clinical improvement. Ophthalmological examination showed no signs of uveitis. Histology revealed esophagitis with rare eosinophils and PCR testing for herpes family viruses was negative. The blood tests results were: normal blood count; CRP 1.43 mg/dL; ANAs positive (1/320, dense granular cytoplasmic pattern); ENAs, ANCAs and HLA-B51 typing negative. An extensive investigation was carried out and the following etiologies were also excluded: immunodeficiencies, celiac disease and inflammatory bowel disease. On the fifth day of colchicine/tenth day of acyclovir, a second UGE showed regression of esophageal ulcers (Fig. 3). He completed 14 days of acyclovir and colchicine was used during one month. A year later he presented acneiform lesions on the back and limbs and presented again oral ulcers, which resolved with the reintroduction of colchicine.

Several diagnostic criteria have been published for BD. The pediatric BD (PEDBD) criteria, published in 2015, focused on pediatric BD, while the others mainly based on adult studies and are not validated for children.[1] According to these criteria, the disease is diagnosed with the presence of three of the following findings: oral aphthosis, genital aphthosis, cutaneous manifestations, thrombosis, ocular manifestations and neurological manifestations.[1]

Our patient has only 2 criteria. However, in more than 80% of BD patients, the disease is not complete before the age of 16 years. Thus, children do not fulfil some BD diagnostic criteria for a longer time than adults, although they have the disease.[1]

This is an unusual case of Behçet's disease (BD), where ulcerative esophagitis was the inaugural manifestation. The clinical

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evolution with the presence of oral ulcers, the good response to colchicine, the appearance of acneiform lesions, the family history of recurrent oral aphthous lesions and the exclusion of other possible diagnoses reinforce BD as the diagnosis in this case.

BD is a recurrent, multisystem and inflammatory disorder that affects all sizes of vessels.[2] Gastrointestinal involvement is more common in children than in adults.[3,4] The frequency of gastrointestinal involvement in pediatric BD ranged from 4.8% to 56.5% in various reports.[2] Although mucosal lesions may occur in any part of the digestive track, the ileocecal region is most frequently involved.[3,4] Esophageal involvement is uncommon.[5] The diagnosis and treatments of the disease in children remain challenging because of its rarity and diverse presentations. The main goal of the treatment is to control the inflammatory process and prevent recurrences.[2]

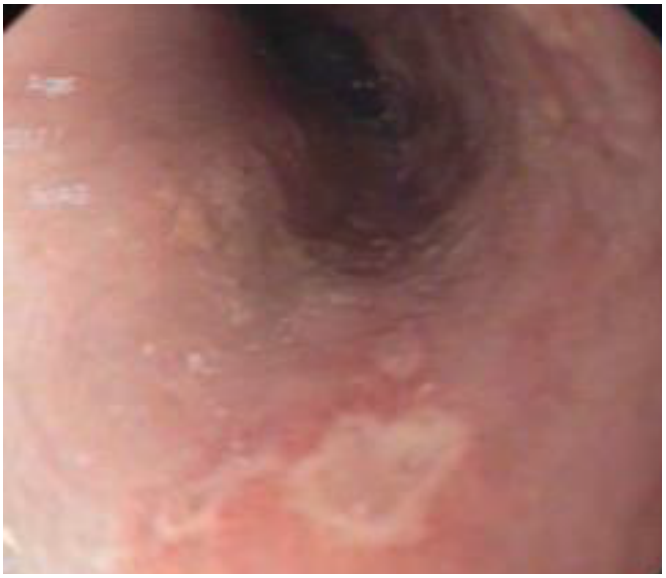

Figure 1. First upper gastrointestinal endoscopy showing an ulcerated esophageal mucosa.

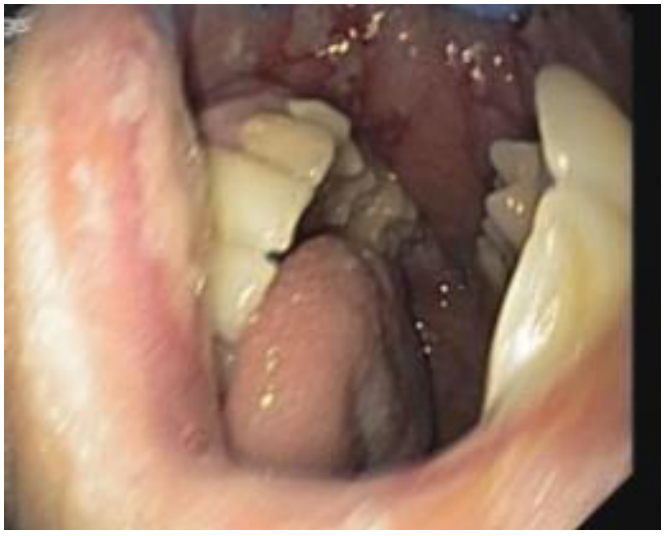

Figure 2. Oral ulcers.

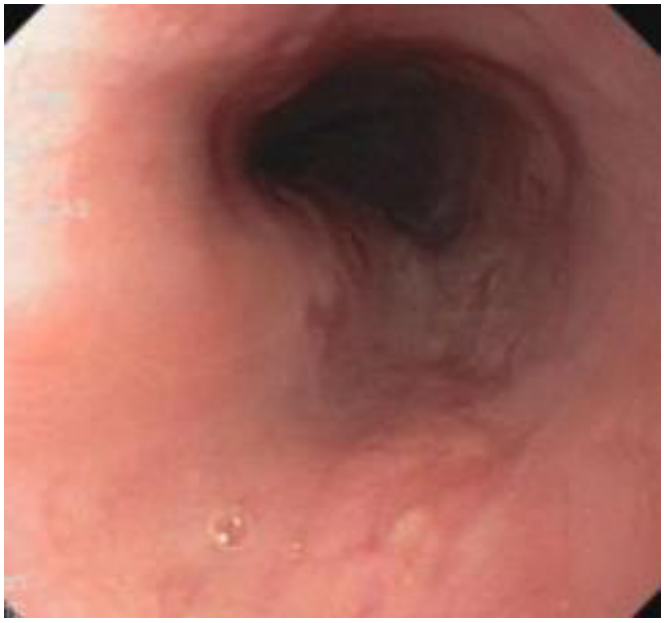

Figure 3. Second upper gastrointestinal endoscopy showing healing of ulcerated esophageal mucosa.

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SD: Data collection, bibliographical search and drafting of the article

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