Behçet’s Disease: a systemic case with recurrent oral ulcers

Evandro Barbosa dos Anjos 1 *
Edimilson Martins de Freitas 2
Daniella Reis Barbosa Martelli 2
Verônica Oliveira Dias 2
Renato Assis Machado 3
Jamile Pereira Dias dos Anjos 4
Hercílio Martelli-Júnior 2,3

Abstract:
Behçet’s disease is a chronic multisystem inflammatory disorder of unknown etiology and characterized mainly by recurrent oral ulcers, ocular involvement, genital ulcers, and skin lesions, presenting with remissions and exacerbations. Case report: The case report presents a patient with recurrent oral ulcers history and extracutaneous ulcers to 10 years who was diagnosed as having Behçet’s disease and was successfully treated. Discussion: Although complications involving the gastrointestinal, cardiovascular, renal, pulmonary, urological, artery, and central nervous system can be observed, some patients can remain asymptomatic for a long period after the diagnosis. Conclusion: This report emphasises that systemic diseases, including Behçet’s disease, can have variable presentations and can be frequently misdiagnosed.

Keywords: Vasculitis; Oral Ulcer; Skin Ulcer; Behçet Syndrome

1 Faculdade de Medicina, Universidade Estadual de Montes Claros, Unimontes, Montes Claros, Minas Gerais, Brazil.
2 Departamento de Patologia Oral da Faculdade de Odontologia da Universidade Estadual de Montes Claros, Unimontes, Montes Claros, Minas Gerais, Brazil.
3 Departamento de Diagnóstico Oral da Faculdade de Odontologia da Universidade Estadual de Campinas, Piracicaba, São Paulo, Brazil.
4 Faculdade de Medicina, Funorte, Montes Claros, Minas Gerais, Brazil.

Correspondence to:
Departamento de Saúde Mental e Saúde Pública, Universidade Estadual de Montes Claros.
Ruy Braga Avenue, Vila Mauricéia, CEP 39401-089, Montes Claros, Minas Gerais, Brazil.
E-mail: evandro.dosanjos@yahoo.com

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INTRODUCTION

Behçet’s disease is a multisystemic inflammatory disease of unknown etiology, which was described for the first time by the Turkish dermatologist Hulusi Behçet, in 1937, as a triad of recurrent aphthous oral ulcers, genital ulcers, and uveitis. Association with HLA-B51 polymorphisms as well as infectious agents has been confirmed in the disease susceptibility in different ethnic groups. At the beginning of the disease, the diagnosis is uncertain, as it presents a wide range of clinical manifestations and only after a long period of time can the full clinical picture be observed. In addition to the eyes, skin, and oral mucosa, the disease can also affect nearly all of the systems and organs, including the cardiovascular, gastrointestinal, renal, pulmonary, urological, and central nervous systems, as well as the joints.

Since neither the laboratory data nor the histopathological signs are truly pathognomonic in Behçet’s disease, the differential diagnosis depends on a careful evaluation of the clinical history and many times, only years after the disease’s first manifestation is one able to establish a definitive diagnosis. In the present report, we describe the case of Behçet’s disease in a young adult male with recurrent oral ulcers and extracutaneous ulcers.

CASE REPORT

A 22-year-old male patient was referred to the Oral Medicine Service - Unimontes, presenting a medical history of recurrent oral ulcers. These ulcers had been appearing for the last 10 years, with no significant resolution of this clinical picture. The painful lesions would last for approximately three months, disappearing without scars. Regarding anamnesis, the patient reported no appearance of extra-oral lesions. Upon performing an intra-oral exam, the patient presented two round ulcers in the right corner of his mouth, each of 1 cm at its widest diameter, with a yellow bottom and reports of spontaneous bleeding and pain (Figure 1A) and two ulcers with raised erythematous borders and a yellow bottom, which were also painful but showed no bleeding, on the left lateral border of the tongue. (Figure 1B). The laboratory blood test was performed for the evaluation of anemia and cellular immune abnormalities, and did not have alteration (Erythrocytes - 5,48 millions/mm³, Hemoglobin - 14.6 g/dl, Hematocrit - 45%, PCV - 82%, EVF - 27 pg, VPRC - 32 g/dl, RDW - 12.5%, Leukocytes - 5,700/mm³, Neutrophils - 60%, Eosinophils - 3%, Typical Lymphocytes - 33%, Monocytes - 4%, platelets - 214,000/mm³).

Despite denying any history or recent occurrence of an extra-oral lesion, the patient underwent a full dermatological exam, which detected a ulcer on the scrotum, with 1.5 cm in diameter, raised borders, and a yellow bottom (Figure 2A), as well as an ulcerated lesion in the region proximal to the left thigh, with 2 cm in diameter, with a raised erythematosus border and a bottom covered by a yellowish pseudomembrane (Figure 2B). The patient also presented papule-pustular erythematous lesions in the perinasal region (Figure 3).

As such, according to the International Criteria for Behçet’s Disease (ICBD), the diagnosis was of Behçet’s disease. The patient was submitted to an ophthalmological exam, which showed no changes. This exam also presented no gastrointestinal, cardiovascular, renal, pulmonary, urological, artery, or central nervous system changes. The patient was prescribed Colchicine, 1 mg/day, which improved clinical picture, given that, in the first 30 days, the patient presented two crises of oral and genital ulcers, which lasted a shorter time than usual. Upon increasing the dose to 2 mg/day, the patient presented important progress in the clinical picture, and has now gone two months without any oral lesions (Figure 4).

DISCUSSION

Behçet’s disease is a variable vessel vasculitis no predominant type of vessel involved that can affect vessels of any size (small, medium, and large) and type (arteries, veins, and capillaries). Different hypotheses to its etiology have been demonstrated, but the most probable is that of an inflammatory reaction set off by infectious agents. Professor Hulusi Behçet was one of the first physicians asserting the role of an infectious agent, when was found a part of the herpes simplex virus (HSV)-1 genome in mononuclear cells of patients with BD. Others studies showed several atypical streptococcal species in the oral flora of patients with BD and a particular strain, S. sanguinis, has been shown to enable the KTH-1 cells to secrete the pro-inflammatory mediators of interleukin (IL)-6, IL-8 and tumor necrosis factor (TNF)-α. The HLA-B51 gene also considered to play important roles in the development of BD, because it is thought to play a role in neutrophil activation. However, the presence only of HLA-B51 gene is not sufficient to explain the symptoms observed in BD and,
Figure 1. A: Ulcers in the labial mucosa of the right corner of mouth  
B: Two ulcerated lesions in left lateral border of the tongue

Figure 2. A: Ulcerated lesion in the scrotum  
B: Ulcerated lesion in the proximal region of the left thigh

Figure 3. Papule-pustular erythematous lesions in the perinasal region  
(Pseudofolliculitis)

Figure 4. Left lateral border of the tongue after the use of colchicine
The genital ulcers, when initially appearing as Behçet's disease, are ranked second, affecting 80% to 90% of the patients. Clinically speaking, they resemble the oral ulcers but are larger and deeper, and have more irregular borders, leaving incurable scars. In men, these ulcers generally occur on the scrotum, in addition to the body of the penis and the glans, while in women these more commonly appear on the small and big lips. In both genders, these ulcers attack the perineal, perianal, and groin regions. The genital ulcers are generally asymptomatic, so much so that the patient often forgets to report them. Thus, anamnesis is geared toward this matter, and the physical exam of the patient's genital organs must be routine practice in cases of oral ulcers.²,³,⁵

The papule-pustular, or acne-like, lesions can occur anywhere and are morphologically similar to pimples, or acne vulgaris. These lesions are the most common cutaneous manifestations, and their distribution is more diffuse than acne vulgaris, affecting the face, limbs, trunk, and buttocks.¹,² Extracutaneous ulcers are uncommon (3%), but when they occur, they are recurrent and primarily affect the inner portion of the thigh, the inguinal region, and the axillary region, and less commonly the neck, thorax, inframammary fold, lower limbs, and interdigital area of the feet. They can be cured, leaving a scar, and are most common in children with Behçet's disease.³

Treatment has become much more efficient in recent years due to advances in the understanding of the pathogenesis this systemic vasculitis and the availability of an ample spectrum of therapeutic agents.¹,³ According to the EULAR (European League Against Rheumatism) the drugs commonly used in the treatment of autoimmune disease is determined by which organ(s) is (are) affected and the extension and severity of the involvement. In addition, new agents such as the anti-TNF-alpha monoclonal antibody infliximab (IFX) have been used to treat in resistant cases with mucocutaneous involvement.⁴ In this case, was prescribed initially 1mg/day of Colchicine, which improved clinical aspect, but in the first 30 days the patient presented two crises of oral and genital ulcers. So, the dose of 2 mg/day was prescribed and a satisfactory clinical improvement was obtained. Colchicine is a important resources and it is associated with the significant remission of the disease, especially when considering the lesions in the oral mucosa, genital ulcers, and pseudofolliculitis.⁵ Promising results have been reported when applying doses of 0.5 to 2.0 mg/day of Colchicine.⁵

Therefore, faced with the clinical picture of recurrent oral ulcers, one should take Behçet's disease into consideration as a diagnostic hypothesis, highlighting the importance of being able to recognize the diagnostic criteria of the disease, even by dentists, as well as the importance of skin analysis regardless of what the patient actually reports.

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