

## **DIAGNOSIS OF BEHÇET'S SYNDROME IN DENTISTRY: CASE REPORT**

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*Ana Luiza Bergonzi Rocha*

Grande Dourados University Center  
Dourados-MS

<https://orcid.org/0000-0001-9748-5726>

*Thais Sumie Nozu Imada Pivetta*

Grande Dourados University Center  
Dourados-MS

<http://lattes.cnpq.br/6744609491181367>

*Tainara Bielecki Yamanaka*

Grande Dourados University Center  
Dourados-MS

<http://lattes.cnpq.br/5398424459501920>

*João Pedro Gasparin Tadano*

Grande Dourados University Center  
Dourados-MS

<http://lattes.cnpq.br/3611551172076504>

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**Abstract:** Behçet's Syndrome (BS) is a multisystem inflammatory disease with mucosal involvement. Its cause is unknown and there is no specific test for the diagnosis of the disease. Lesions in the oral mucosa are usually the first signs that the disease manifests, so it is extremely important for Dental Surgeons to be aware of this syndrome. We report here the case of a patient with Behçet Syndrome, who recently presented oral ulcers characteristic of the disease, with the aim of emphasizing oral lesions to assist in the early diagnosis of the disease through Dental Surgeons.

**Keywords:** Inflammatory disease; Ulcers; Oral injuries; Dental surgeon.

## INTRODUCTION

Behçet's Syndrome (BS) is a systemic inflammatory disease with mucosal inflammation and can be classified as systemic vasculitis, since there is the possibility of reaching arteries and veins of any organ. Its main manifestations involve recurrent oral ulcers, genital ulcers, uveitis and skin lesions. Neurological and gastrointestinal involvement are more serious manifestations (FORTE-VILLA, 2019). According to Forte-Villa (2019) the cause of the syndrome is still unknown, however, studies show that immunological, bacterial or viral factors are suggestive. Ohno (1982), made the first association of Behçet Syndrome with the Human Leukocyte Antigen (HLA-B51) and from that, several studies found a relationship between the syndrome and the allele in the countries of the "Silk Road" between Asia and Eastern Mediterranean, where the prevalence of (HLA-B1) is relatively high compared to other parts of the world. However, according to Neves et al. (2006), the relationship between the HLA allele and the syndrome has not been proven.

It is a disease of young adults, aged between

25 and 30 years, however, cases in neonates have already been reported. According to Tolentino et al. (2018) there is no specific exam or test for BS, the diagnosis is made based on clinical criteria. A detailed clinical history of the patient is essential to exclude other conditions with similar characteristics. Treatment is mainly symptomatic, and may include corticosteroids with or without immunosuppressants for more severe cases. Carneiro et al. (2005) report that Behçet's disease is of total interest to the Dental Surgeon (CD), because it involves the oral mucosa with episodes of recurrent and painful ulcers that can prevent oral hygiene, facilitating the establishment and aggravating gingivitis. Thus, the CD can be the first professional to initiate investigations to carry out an early diagnosis. Given the above, due to the disease being little known in the dental environment, the objective of the research is to emphasize the oral manifestations of BS, showing its characteristics through a case report about the disease, and to say about the diagnosis through the Dental Surgeon and the treatment that it can offer.

## CASE DESCRIPTION

An interview was carried out with the patient of the case, due to this the research was sent to the ethics committee of the institution, it was analyzed and approved. A 46-year-old female patient with BS diagnosed 14 years ago presented the first symptoms of the disease in 2007 with difficult-to-heal oral ulcers, accompanied by genital ulcers, reddish lesions on the hands and lower legs. In the same year, she presented fainting and joint pain, but in 2008, after several consultations with specialist doctors and dentists, a rheumatologist concluded the probable diagnosis as SB, associating oral ulcers with the other symptoms.

Most people who have systemic

autoimmune rheumatic diseases have a positive antinuclear factor (ANA) on the complete blood count. To close the diagnosis, blood, ophthalmological and neurological tests were performed, in addition to biopsies of the lesions. The patient had a positive ANA, which related to the other symptoms, the diagnosis was closed for Neuro Behçet, when Behçet's Syndrome affects the nervous system.

The signs and symptoms of BS are not always active, the disease goes into remission and the patient can be symptom-free for up to a year, and with that it can be controlled with corticosteroids. When the disease is active, with constant fainting and more frequent canker sores, the protocol suggested by the rheumatologist was to perform pulse therapy, which consists of the intravenous application of cyclophosphamide and methylprednisolone, and for oral ulcers, laser therapy associated with topical application of

triamcinolone, which were the first treatments to be performed on the patient. After 4 years of diagnosis, the BS went into remission in the patient and medications were started to control the disease with prelone 40ml/day, colchicine 5ml/day, azathioprine 40ml/day, and omcilon for canker sores.

The patient's last episode in which the disease was active was monitored, she had been without any type of sign or symptom for more than 10 months. At the beginning of the episode, it can be noted that the first sign was the return of oral ulcers, followed by severe joint pain, headaches, genital ulcers, uveitis and skin lesions in the region of the hands and legs. There were reddish lesions in the upper lip region (Figures 1 and 2), and oval ulcers with a yellow center on the cheek mucosa (Figure 3) and palate (Figure 4). The patient did not seek clinical care this time.



Figure 1- Oral lesion on the upper lip.



Figure 2- Oral lesion on the upper lip.



Figure 3- Oral ulcer on the buccal mucosa.



Figure 4- Oral ulcer on the palate.

Source: Prepared by the author.

## DISCUSSION

BS was initially characterized by the triad of recurrent aphthous stomatitis, genital ulceration and recurrent uveitis, but over the course of 60 years, other associations with the disease began to appear, such as joint, vascular, gastrointestinal, cardiopulmonary and neurological problems. In a study carried out, it is shown that aphthous ulcerations are the first manifestations of the disease in 86% of the cases, followed by genital ulcerations usually accompanied by oral aphthas in 7% of the cases (GÜLER et al. 1997).

In the present case, oral ulcers are very recurrent and were the first manifestations of the disease in the patient, which led the Rheumatologist to investigate a possible diagnosis for Behçet Syndrome. These oral ulcers are usually painful and recurrent, round or oval, measuring from 2 to 10 mm in diameter, superficial or deep with a yellowish necrotic center, and can be found anywhere in the oral cavity (FORTE-VILLA, 2019) and heal without scarring. in about 10 days.

According to Yazici, (1991) BS is more common between the third and fourth decade of life, but it can also be seen after 50 years of age or in childhood. In the case reported, the patient was diagnosed at 33 years of age. Diagnosis is usually late because its manifestations are nonspecific and can be insidious. It can be distinguished by its combination of recurrent symptoms with spontaneous remissions and multi-organ involvement.

International criteria for diagnosis include recurrent oral ulcers, three times in a year, plus two of the main signs, such as genital ulcers, eye lesions, skin lesions, and positive pathergy test in the absence of any other clinical explanation. Laboratory tests such as blood count, ESR or C-reactive protein, serum albumin and total protein levels are also performed. The results are nonspecific,

but characteristic of inflammatory diseases. A differential diagnosis is also made to exclude some diseases such as Reactive Arthritis, Systemic Lupus Erythematosus, Crohn's Disease, Ulcerative Colitis, Ankylosing Spondylitis, Periodic Fever Syndrome and Herpes Simplex infection (FORTE,-VILLA 2019).

Topical local anesthetic preparations [2% lidocaine gel] may alleviate minor symptoms and local steroid preparations that adhere to the oral mucosa (eg, oral triamcinolone acetonide) applied locally several times a day give symptomatic relief and likely shorten the duration of ulceration of the cold sore. Some patients also note the development of a red mucosal papule prior to acute ulceration at the same site. Local application of corticosteroids will reduce the severity and duration of ulceration (Yazici & Barnes 1991).

Some studies suggest other prophylactic measures composed of biological mechanisms exerted in each phase of oral mucositis, such as the use of low-level laser (SANDOVAL et al. 2003). Laser therapy has biological effects and acts in the abolition of pain and in the modulating action of inflammation (FIGUEIREDO et al. 2013).

## CONCLUSION

The knowledge of Behçet Syndrome by the dental surgeon is extremely important, because in most cases the oral manifestations are the first to appear. Therefore, the CD must know how to identify these lesions and relate them to other signs and symptoms of the syndrome so that there is an early diagnosis and a correct referral of the patient. In addition to the importance of diagnosis, treatment of lesions that are extremely painful must be performed, for a better quality of life for the patient.

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