Case Report

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# Oral Manifestations of Behcet's Syndrome: Case Report

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**ABSTRACT**

Behcet's syndrome is an autoimmune disease of the seronegative vasculitic type, with a disease of vessels of variable caliber. It is mainly characterized by oral ulcers, genital ulcers and uveitis. The purpose of this case is to describe the oral manifestations and therapeutic behavior to be followed before a patient with Behcet syndrome. For our purpose, the case of a 20-year-old patient, female, black race, urban origin, who goes to the emergency departments of Maxillofacial Surgery of the body of guard of the Provincial Hospital Dr. Gustavo Aldereguía Lima in the province of Cienfuegos, referred by the specialist in Comprehensive General Stomatology who provided services in the area of health corresponding to the patient for presenting multiple ulcerated lesions throughout the oral cavity and lips. It was concluded that oral ulcers in Behcet's disease are the first clinical manifestation, which in most cases, lead the patient to seek medical help and are considered by countless authors as the cornerstone to reach the diagnosis of this disease. A total of 14 bibliographies were consulted.

**Keywords:** Behcet Syndrome; Oral Ulcers; Patergia

## Introduction

Systemic diseases may be preceded or spread, with oral manifestations. The lesions of the oral cavity are attended, for the most part, by Stomatology Specialists, so that knowledge of the oral manifestations of systemic diseases contributes to correct behavior in the face of these conditions that, finally, must be referred to the specialist in the attention of each one of them [1]. Behcet's syndrome is an autoimmune disease of the seronegative vasculitic type, with a disease of vessels of variable caliber [2]. Although the etiology of Behcet's Syndrome is unknown, there are numerous investigations that indicate the participation of genetic, infectious (viral and bacterial) and immunological factors. Among the most important are the association with the HLA genotype of the patients, the cross-reactivity with human peptides and the activation of the vascular endothelium [3]. This disease is more common and severe along the old "silk road", which extended from East Asia to the Mediterranean, Turkey's current territory. 80 to 370 cases are reported per 100,000 inhabitants in Turkey, while the prevalence ranges from 13.5 to 35 per 100,000 in Japan, Korea, China, Iran, Iraq and Saudi Arabia; being a more severe manifestation in Japan [4].

In relation to sex, a predominance of female sex in countries of America and male sex in Asia is described, mainly between the ages of 20 and 40 [5]. In Cuba there are no prevalence studies of this entity [3]. The first description made of this pathology arises may have been described by Hippocrates in his third book of endemic diseases [4]. In 1930, the Greek ophthalmologist Benediktos Adamantiades reported to a patient with arthritis, oral and genital ulcers, phlebitis, and iritis but it was not until 1936 that the Turkish dermatologist Hulsui Behcet determined the description

of this disease of more accurate way [6]. The clinical spectrum of this disease, characterized by a symptomatic triad (recurrent oral and genital ulcers with uveitis and hypotension) in addition to the fact that the histopathological study of the affected organs and the analytical data are nonspecific, makes its diagnosis based on the clinical method of description and grouping of signs and symptoms [7]. In 1990, the criteria for the diagnosis of this entity were established for the first time by the International Study Group for Behçet's Disease [8].

In 2006, new diagnostic criteria of the International Group for the study of Behcet's disease were established [9], which declared a major or mandatory criterion: recurrent oral ulcers and minor criteria: recurrent genital ulcers, eye lesions, skin lesions, vascular lesions and positive patergia test [10]. In 2013, the international criteria for the diagnosis of the disease were reviewed, as a result of the analysis they proposed to award 2 points to ocular lesions and oral and genital aphthous lesions and assign 1 point to skin lesions, of the Central Nervous System, vascular manifestations and the positive patergia test. According to these criteria, a patient with a score ≥ 4 points is classified as a Behcet disease [3]. At present it is known that it is not curable, but treatable [4]. The goals of therapy are to: suppress inflammation, reduce the frequency and severity of recurrences. To be effective, treatment must be implemented early [11]. Objective: Describe the oral manifestations and therapeutic behavior to be followed before a patient with Behcet syndrome.

## Case Presentation

Patient of 20 years of age, female sex, black race, urban origin, who comes to the emergency services of Maxillofacial Surgery of the body of guard of the Dr. Gustavo Aldereguía Lima Hospital in Cienfuegos, Cuba referred by the specialist in Stomatology General Comprehensive that provided services in the area of health corresponding to the patient for presenting multiple ulcerated lesions throughout the oral cavity and lips Figure 1.

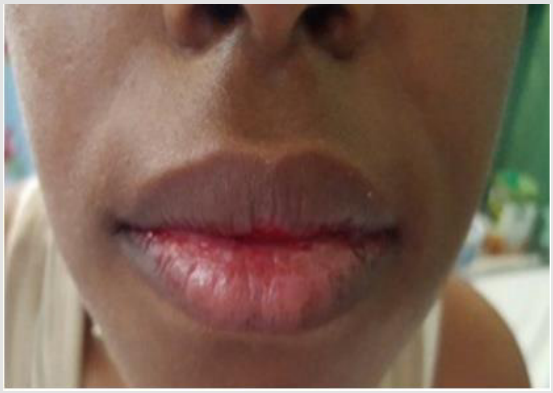

**Figure 1:** Ulcerated lesions on lips.

### Anamnesis

During the interrogation, the patient was referred to as inflammation of the entire oral cavity, pain during food intake and difficulty swallowing (dysphagia), burning sensation and burning of the mouth, halitosis, recurrent episodes (7 to 8 times a year) ulcerated bleeding lesions, which previously healed spontaneously until their total disappearance but during the last episode the lesions persist. It also refers to the presence of genital ulcers, skin rash in the frontal region and persistent joint pain.

### Physical Exam

Multiple ulcers were detected in the oral cavity of approximately 1cm and more, covered by a yellowish-green pseudo membrane and with fetidity due to the over-added infection of the lesions, with irregular, undefined, elevated, rounded, hyperemic edges, bleeding to the minimum stimulus, painful on palpation, located in the region of the oropharynx, tongue, palate, gum, floor of mouth, mucosa of cheeks and lips Figure 2a. In addition, irregular and rough surface was detected upon palpation of the frontal cutaneous region Figure 2b and in the left axillary and subaxillary region Figure 2c. The case of the patient was consulted with the Internal Medicine services for a better treatment of genital lesions and joint pain.

### Complementary Exams

The following complements were indicated:

**a)** Blood count

**b)** Leukogram

**c)** Blood chemistry

**d)** Hepatic Profile

**e)** HIV serology

**f)** Rheumatoid Factor

**g)** C-reactive protein

**h)** LE cells

**i)** Immunoglobulins Serum

**j)** Patergia test

**k)** Surface antigen

**l)** Hepatitis C antibody

The results of the complementary examinations were within the normal parameters except the Leukogram in which there was a considerable increase in segmented leukocytes (neutrophils) with a value of 80%, which showed that the over-added infection of the lesions was bacterial. During the patergia test, a small sterile needle was inserted into the skin of the forearm, causing a small red papule to appear at the needle insertion site one or two days after the test was performed, which indicated hyperreactivity of the immune system to minor trauma or damage Figure 3. It is not 100% specific, only a proportion of patients with Behcet syndrome have a positive response.


**Figure 2:**
(a) yellowish-green pseudo membrane located in the tongue and oropharynx;
(b) Skin rash in the frontal region;
(c) Irregular and rough surface in the left axillary and subaxillary region.

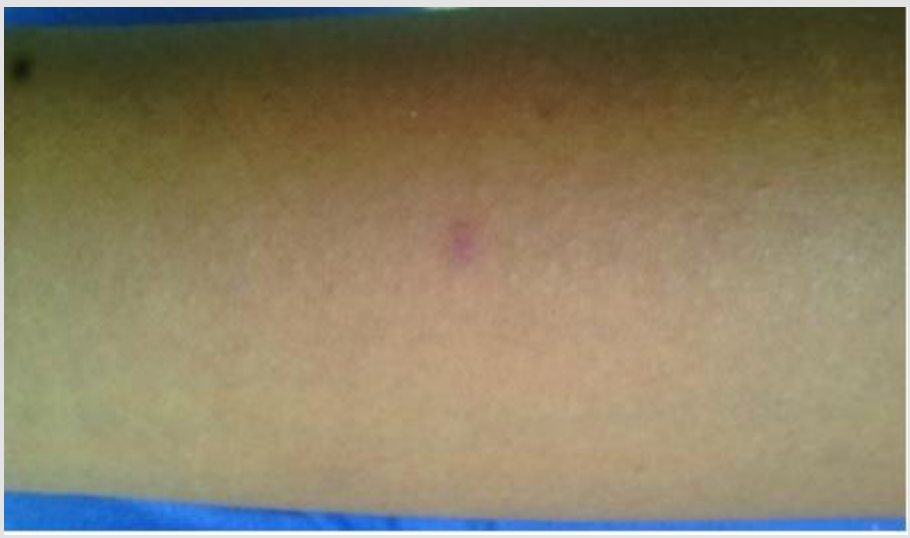

**Figure 3:** Red papule on forearm skin two days after the patergia test.

### Diagnosis

With these results, some possible diagnoses were ruled out: Human Immunodeficiency Syndrome, Rheumatoid Arthritis, Hepatitis B and C, Herpes Simplex type 1, Recurrent Aphthous Stomatitis, Parvovirus, Discoid Lupus Erythematosus. Finally, the patient was diagnosed with Behcet Syndrome with a scientific basis in the criteria for the diagnosis of this syndrome of 2013, complying with the mandatory or major requirement: recurrent oral ulcers in oral mucosa with a minimum of 3 episodes during a year (2 points ) and with three minor criteria: genital ulcers (2 points), erythema nodosum (1 point) in this case in the left frontal, axillary and subaxillary region and positive patergia test (1 point). According to this current classification criterion it is only necessary to accumulate a score ≥ 4 points, in this case the patient added a total of 6 points.

### Treatment

As part of the Stomatological treatment, the patient was indicated a series of hygienic dietary measures such as: eliminating or reducing acidic foods from the diet (lemon, orange, tangerine, pineapple, guava, tomato, natural yogurt); avoid ingesting foods with artificial condiments and at elevated temperatures and instead use natural condiments and ingesting food and drinks that are refreshing but not gaseous; Eliminate local irritants such as coffee, cigar and alcohol. In addition, local topical anesthetics such as 0.2% lidocaine were indicated half an hour before eating food to relieve the pain caused by chewing and swallowing in these cases. In this case, the indication of mouthwash with saline solution or 20% chamomile tincture was very effective, 20 drops in 200 milliliters of boiled water 3 times a day for 7 days, the use of chamomile was preferred as part of the natural medicine treatment and traditional for its properties: anti-inflammatory, analgesic, antibacterial, antiulcer, antiviral and antifungal.

The Internal Medicine services treated the patient with a prednisone in a 20-milligram bulb intravenously every 8 hours, then gradually reduced to a maintenance dose of 10 milligrams orally in tablets, this corticosteroid was very effective in the treatment of inflammatory component of oral, genital and arthropathic lesions of the patient and as an immunomodulator. In addition, colchicine (0.5 mg) was given one tablet every 8 hours; methotrexate (2.5 mg) four tablets in weekly dose; folic acid (5 mg) one tablet daily, except on the day of Metrotexate, all in order to control the rheumatologic component. Currently, the patient is compensated for the disease.

## Discussion

Systemic diseases are those morbid processes that affect more than one organic system. The etiopathogenesis of many of these diseases is still not completely clear, but it is well known that, in large part of them, inflammatory processes and immune system disorders that give rise to the various manifestations are involved. The majority of patients with recurrent oral aphthous ulcers have no other involvement, but in others the presence of chronic aphthous stomatitis lesions is associated with systemic processes [1]. Behcet's syndrome is a very rare autoimmune disease condition in these latitudes [4]. The most accepted concept so far defines it as: in a chronic, multisystemic, recurrent inflammatory process whose main alteration lies in a vasculitis that involves the arteries and veins of any caliber. It is characterized in the clinic by presenting patients with inflammation of the mucous membranes, translated by oral and genital ulcers; in addition to uveitis; digestive symptoms, skin lesions, arthritis and occasional neurological intake.

The etiology of Behcet's disease is unknown and although the majority of cases are sporadic, some studies support the possible genetic origin with evidence of autosomal recessive inheritance as it is reported that in 1 in 10 families there is another member with the disease or other autoimmune diseases in first-degree relatives such as hypothyroidism, scleroderma, discoid lupus erythematosus and juvenile idiopathic arthritis. Another of the arguments described in defense of this genesis is the increased risk of suffering from Behcet's disease when it is associated with the presence of the HLA-B51 main histocompatibility system antigen [12]. It is believed as for many autoimmune or auto-inflammatory syndromes, that certain infectious factors (in particular, Streptococcus antigens) and / or environmental factors are capable of triggering symptoms in individuals with certain genetic variants [13]. In some investigations, the herpes simplex virus type 1 and parvovirus B 19, among the triggers of Behcet's syndrome, other research reports that in this syndrome there is an alteration in the number and activation of lymphocytes, so the CD4 + / CD8 + index inversion has been observed.

The syndrome is usually more serious and frequent in men [3], however the case presented corresponds to a female patient. The white skin color shows a predominance, although there is a great miscegenation in Cuba [12], this ailment is uncommon in the black population but when it appears they present greater complications and worse prognosis [14]. Over the years different criteria have been used to diagnose the disease, the International Study Group for Behçet's Disease in 1990 considers the existence of recurrent oral ulceration with at least two of the following clinical manifestations: recurrent genital ulceration, ocular involvement, involvement cutaneous or positive patergia test to make the diagnosis [8]. In 2006 the International Group declares it necessary to present: Mandatory Criteria or major criteria: recurrent oral ulcers (minor canker sores, major canker sores or herpetiform ulcers, in oral mucosa with a minimum of 3 episodes for a year). Minor criteria: recurrent genital ulcers (ulcers or aphthous scars in genital areas observed by the doctor or the patient) (2 points). Eye lesions (anterior or posterior uveitis, or the presence of vitreous

cells in the examination with slit lamp or vasculitis retinal diagnosis by an ophthalmologist) (1point).

Skin lesions (erythema nodosum, folliculitis, papulopustular lesions, acneiform nodules, observed by the doctor in post-adolescent patients not treated with corticosteroids) (2 points). Patergia test: positive (cutaneous hypersensitivity characterized by the appearance of a sterile pustule, 24 to 48 h after needle puncture, observed by a doctor) (1 point). Vascular lesions (arterial, venous thrombosis or aneurysms) (1 point). The diagnosis is made with the mandatory criterion plus 3 points [10]. In 2013, an international group, composed of representatives from 27 countries, reviewed the international criteria, according to these criteria, a patient with a score ≥ 4 points is classified as a Behcet disease [3]. The presence of recurrent, painful, variable-sized canker sores that heal in 1-3 weeks usually without scarring in most cases are the first manifestation, as occurred in this case. The genital canker sores present in 72-94% of cases are morphologically similar to oral ones, usually heal at 2-4 weeks, but they do leave a scar [7]. There are no typical laboratory alterations of Behcet Syndrome. Erythrosedimentation and C-reactive protein levels are often moderately high but do not correlate well with disease activity, in this case these values were kept within normal parameters.

Acute phase reactants may be elevated, mainly in patients with vasculitis of large vessels. Serum immunoglobulins are sometimes elevated and immunocomplex levels may be elevated, but autoantibodies such as rheumatoid factor, antinuclear antibodies, anticardiolipin and neutrophil antithoplasm (ANCA) are negative [3]. The patergia test is not 100% specific, only a proportion of patients with Behcet Syndrome have a positive response and the positivity in patients with the syndrome vary in different geographical areas (60-70% in Turkey and Japan, but rare in America and Europe) [11], however, the case presented is of a patient born in Cuba who is part of the American continent in which the test was positive, not coinciding with world statistics, where positivity is rare in the Americas. The differential diagnosis should be made with: Reiter's disease, Steven-Johnson syndrome, aphthous stomatitis, recurrent Mollaret meningitis, Whipple's disease, multiple sclerosis, Harada's disease, inflammatory bowel diseases, sarcoidosis and ankylosing spondylitis. The diagnosis is based on detailed medical history and long-term clinical observation. There is no way to predict whether a patient with recurrent oral ulcerations will develop the disease even if frequent outbreaks constitute an alarm sign, as confirmed in this case. The choice of treatment depends on the clinical manifestations and their severity.

An early diagnosis followed by a treatment with corticosteroids, medication used in this patient, could prevent the fearful neurological complications in the opinion of several authors, although more recent studies report highly positive results with immunosuppressants [7]. It is described that all patients should use colchicine and steroids, with Metrotexate being the disease-modifying drug most used in the treatment of this [5]. The majority of patients diagnosed with Behcet syndrome have a productive life. The symptoms are controlled with a healthy diet, rest, physical exercises and drug treatment and the prognosis varies according to the affected organs [7]. Behcet's disease has a chronic course with exacerbations and unpredictable remissions, the frequency and severity of which may decrease over time. After the first five years, the disease acquires a stable course or towards improvement; after the fourth decade, clinical severity is decreased, with longer intervals of recurrence between exacerbations. The prognosis is good, unless there is commitment of vital organs. The presentation at the age of early onset (before age 25) has been associated with more severe manifestations of the disease and increased mortality [13].

## Conclusion

While it is true that multiple systemic diseases present with aphthous oral manifestations, oral ulcers in Behcet's disease are the first clinical manifestation, which in most cases, lead the patient to seek medical help and constitute for countless authors the cornerstone to reach the diagnosis of this disease, so it is considered vital that the Comprehensive General Stomatologist putting into practice the knowledge acquired about the syndrome, consider it a possible differential diagnosis more before a patient with recurrent oral ulcerations. The timely management of a patient with an unusual and exceptional pathology can have an optimal evolution and reduction of complications in the quality of life. It is recommended to expand coverage and health education to be able to detect these cases in the first instance, thus avoiding underdiagnosis.

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