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Research Article

Mucocutaneous involvement in behçet's disease

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Abstract

Behçet's disease is a chronic inflammatory disease characterized by its clinical polymorphism associating mucocutaneous involvement to systemic manifestations. The mucocutaneous lesions are considered the hallmark of the disease, being the most common symptoms presenting at the onset of disease. Our objective was to determine the characteristics of this skin involvement during Behçet's disease. We conducted a descriptive study over a period of 30 years, having collected all patients with Behçet's disease. These were 98 patients. A male predominance was observed in our studied population with a Sex Ratio of 2.5. The mean age at diagnosis was 34 years. Mucocutaneous involvement was observed in all patients. Oral aphthosis was constant and genital ulcers, were observed in 81 cases. The other mucocutaneous manifestations were: pseudofolliculitis (61 cases), erythema nodosum (7 cases), skin ulcers (4 cases), acneiform lesions (2 cases), perianal ulcers (1 case), skin ulceration (1 case) and erythema multiforme. (1 case). All of our patients were treated with colchicine. Corticosteroids and non-steroidal anti-inflammatory drugs were each indicated in one case for resistant forms.

Keywords: behçet; oral aphtosis; genital ulcer; Pseudofolliculitis; pathergy test

Abbreviations: BD: Behçet's disease

Introduction

Behçet's disease (BD) is a chronic inflammatory disease of unknown etiology, characterized by its clinical polymorphism associating recurrent oral aphthosis, genital ulcers, and systemic manifestations mainly ocular, vascular and neurological. The mucocutaneous manifestations are fundamental and represent four criteria among the international criteria for Behçet's disease. Their frequency varies depending on the geographic origin.

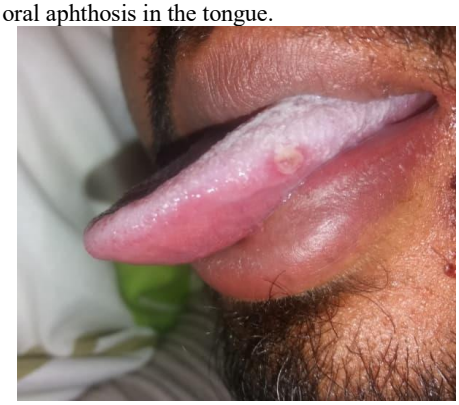
The aim of our work is to describe the main mucocutaneous manifestations during BD.

Materials and Methods

This was a single-center, retrospective and descriptive study, conducted over a period of 30 years (from January 1, 1990 to December 31, 2019). We have collected all the files of patients with BD, hospitalized or followed in the outpatient department of the Internal Medicine department of Mohamed Taher Maamouri hospital in Nabeul. The diagnosis was retained according to the international criteria of BD.

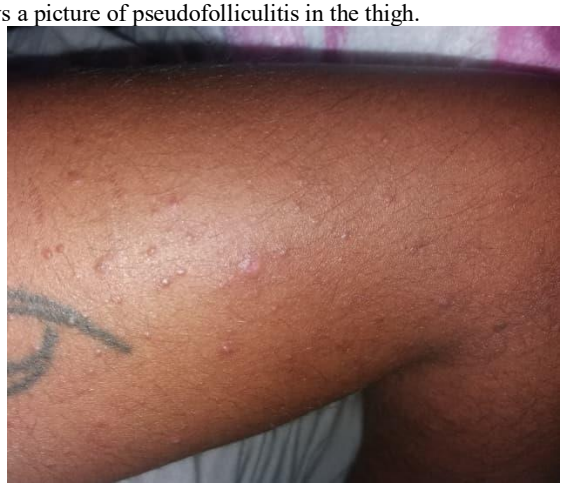
Results and Discussion

During the study period, we collected 98 cases of confirmed BD. The annual incidence of BD averaged 3.7 new patients per year. A male predominance was marked. These were 70 men and 28 women with a sex ratio of 2.5. The average age at diagnosis of BD was 34 years, ranging from 15 to 63 years. A family history of BD was found in five patients (5.1%). All of our patients had at least one mucocutaneous involvement. Oral aphthosis was present in all patients, whose different locations were at the level of: the internal surface of the lip (68%), the internal surface of the cheek (24%), the tongue (8 %) and gums (4%. Figure number 1 shows a picture of oral aphthosis in the tongue.

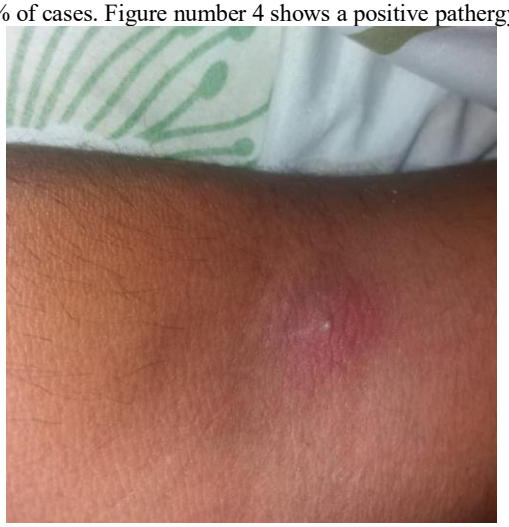


Genital ulcers were observed in 81 cases (82.7%). Genital aphthosis were noted in 63 patients (64.3%). Figure number 2 illustrates the image of genital aphthae in the scrotom.

In addition to bipolar aphthosis, other mucocutaneous manifestations were noted in 66 cases (67.3%). Pseudofolliculitis was observed in 61 cases (62.2%), located in the back, thighs, trunk and face. Figure number 3 shows a picture of pseudofolliculitis in the thigh.



The other mucocutaneous manifestations were erythema nodosum in 7 cases, cutaneous aphthosis in 4 cases, acneiform lesions in 2 cases, perianal aphthosis, skin ulceration and erythema multiforme in one case respectively. Skin hypersensibility was demonstrated by the skin pathergy test in 76% of cases. Figure number 4 shows a positive pathergy test.



All of our patients were treated with colchicine. Nonsteroidal anti-inflammatory drugs were indicated in one case of colchicine-resistant erythema nodosum. Topical corticosteroids were started in a patient with genital ulcers resistant to colchicine.

The most common manifestation is mucocutaneous involvement in BD. Our study confirms this finding. The presence of specific skin signs is a precious help for a definite diagnosis. Histologically, the lesions are frequently perivascular with proeminent infiltrates of neutrophils and/or lymphocytes. Among the international classification criteria for BD, four criteria are dermatological (oral aphthosis, genital aphthosis, and positive pathergy test and skin involvement) [1]. Skin manifestations may precede or occur concomitantly with other systemic manifestations. But can also occur after the systemic manifestations which makes the diagnosis of BD difficult, explaining the important diagnostic delays.

Oral ulcers are observed in 92 to 100% of cases [2, 3]. Their evolution is marked by disappearance without scars but recurrences are frequent [2]. The entire oral cavity may be concerned (inner face of the cheeks, inner face of the lips, tongue, palate, and pharynx).

Genital ulcers, reported in 60% to 87% of cases [4–6], are lesions very suggestive of BM which leave depigmented scars allowing retrospective diagnosis of the disease. However, relapses are less common compared to mouth ulcers.

Aphthae can also be found throughout the digestive tract from the mouth to the anal mucosa [7]. Cutaneous aphthosis is rare and occurs between 0.9% and 6% of cases [3, 8, and 9]. It is the most characteristic skin lesion in BD and usually leaves scars. Its seat is variable [10].

Pseudofolliculitis is the most common skin lesion in BD [11]. It is seen in 31 to 66% of cases [3, 5, 9, and 12]. It is a papule not centered by a hair, located mainly at the back, the front of the thighs, the face and the lower limbs [13]. Erythema nodosum is very painful and occurs mainly in the lower limbs and disappears within a few days. It is seen in a third of cases of BD according to the literature [5, 6, and 14].

Acneiform papulopustular lesions can be seen during BD in 0.4 to 8% of cases [8, 14], erythema multiforme [8], vascular purpura [11], leg ulcer [6], pyoderma gangrenosum [15], and sweet syndrome [15] have also been described in BD.

Skin hypersensibility at the injection sites is common during BD. It is explored by the pathergy test, described for the first time in 1937 [11], resulting in a popular lesion, which becomes papulo-vesicular then papulo-pustular. This skin hypersensitivity is one of the most specific signs of BD. Several authors therefore give it great diagnostic value, which justifies its inclusion among the classification criteria for BD [1]. The positivity of this test varies between 45% and 70% of cases according to the different series published [3–5, 12]. The dispersion of the results can be explained by the ethnic origin of the patients, the heterogeneity of the techniques for carrying out the pathergy test between the teams and the taking of colchicine or anti-inflammatory drugs before performing this test.

When there is no systemic lesions requiring oral corticosteroids or immunosuppressive therapy, the treatment of dermatological lesions is based on colchicine and topical corticosteroids, possibly thalidomide and dapsone for refractory lesions.

Conclusion

The main clinical manifestation of the Behçet's disease is mucocutaneous involvement. It is polymorphic but it is fundamental to find for a diagnosis of certainty.

Oral and genital aphthae are the main clinical dermatologic manifestations. Cutaneous lesions mainly include pseudofolliculitis, erythema nodosum and pyoderma gangrenosum. Histologically, these lesions are essentially perivascular with proeminent infiltrates of neutrophils and/or lymphocytes. Hypersensibility to needle pricks is explored by the skin pathergy test. But the frequency of its positivity

varies depending on the country. In the absence of systematic lesions justiciable of aggressive treatment, the treatment of dermatological lesions is based on colchicine.

Conflict of Interests

The authors declare that they have no conflict of interests regarding the publication of this paper.

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