

# Behcet disease - A nonvenereal cause of genital ulceration

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## Abstract

Behcet's disease (BD) is a chronic, relapsing, inflammatory vascular disease with no diagnostic or pathognomonic test. Here, we present a case of 26-year-old male with a complaint of recurrent oral, genital, and cutaneous lesions. The diagnosis of BD was confirmed on the basis of revised the International Criteria for BD. The case was treated satisfactorily with systemic corticosteroid in the tapering dose and oral Dapsone.

**Key words:** Behcet's disease, dapsone, international criteria for behcet's disease

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## INTRODUCTION

Behcet's disease (BD), also called Adamantiades-Behçet is a chronic, recurrent, multisystem inflammatory disorder of unknown etiology, characterized by triad of oral ulcers, genital ulcers, and ocular lesions.<sup>[1]</sup> This association was probably first recognized by Hippocrates but is named after Turkish dermatologist Hulusi Behçet, who in 1937 first described symptom complex of recurrent oral ulcers, genital ulcers, and uveitis.<sup>[2]</sup> It also involves vascular, intestinal, articular, urogenital, and neurologic systems<sup>[1]</sup> and is also known to cause erectile dysfunction.<sup>[3]</sup>

The disease is worldwide in distribution, more common in populations with a higher prevalence of human leukocyte antigen (HLA) B5 and its split, HLA B51, in the Mediterranean basin, Middle and the Far East. HLA B5 prevalence in north Indians is high.<sup>[1]</sup> Familial aggregation of BD has also been reported mainly from Japan and Turkey.<sup>[4]</sup>

Its exact etiology is unknown, both genetic and environmental factors are thought to play a role in its pathogenesis. Association with HLA-B51 and hyper-reactivity against streptococcal antigen suggest Th1 type of autoimmune disease.

The diagnosis is primarily based on the International Criteria for BD (ICBD), derived from an evidence-based protocol [Table 1].<sup>[5]</sup>

## CASE REPORT

A 26-year-old male presented with multiple oro-genital ulcers, skin lesions over face and legs on and off for 1–1.5 years; increased for the past 2 weeks. In the past, he was treated for uveitis; at that time, there was no history of mucocutaneous lesions.

On examination, multiple aphthous ulcers were seen over the inner aspect of the upper lip, dorsum of the tongue, posterior aspect of bilateral buccal mucosa, the shaft of the penis [Figures 1, 2 and 3a]. Few erythema nodosum-like lesions were seen bilaterally over the shin. Erythema was seen over the right periorbital area [Figure 3b and c].

Ophthalmic examination was normal. All other routine investigations, including complete blood count, renal, and liver function tests, were within the normal limits. S. HIV Antibody test and Venereal Disease Research Laboratory

(VDRL) test were nonreactive, X-ray chest was normal. The pathergy test was negative. The biopsy was not taken as the patient was not inclined to the same.

Based on history and clinical features diagnosis of Behcet's was considered. As per the Revised ICBD, score of our patient was five (oral aphthosis-2, genital aphthosis-2, and skin lesions-1), confirming the diagnosis of BD.

He was advised to take tablet dapsone 100 mg and oral prednisolone 40 mg, along with a course of antibiotics and supportive treatment. On 3 weeks follow-up, oral and genital lesions showed regression while few lesions over lower limbs were still persistent. After 6 weeks, complete resolution of all the lesions was observed. Oral prednisolone was tapered over a period of 6 weeks and tablet dapsone 100 mg was continued for 3 months to maintain remission. The patient was regularly followed up for a period of 6 months, and there was no recurrence of lesions after discontinuation of dapsone therapy.

## DISCUSSION

BD is an inflammatory vascular disease, affecting vessels of all sizes.<sup>[3]</sup> BD usually affects young adults between 20 and 40 years of age. Our patient was 26-year-old. The mean age of disease onset in a study conducted by Singal *et al.*,<sup>[1]</sup> in India was 27.4 years, lower than that reported by Pande *et al.*,<sup>[6]</sup> 33.1 years.

In our case, oral and genital ulcers were the presenting features. Lesions suggestive of erythema nodosum were present over lower legs with mild erythema and swelling

**Table 1: International criteria for Behcet's disease**

Symptom	Points
Ocular lesions (recurrent)	2
Oral aphthosis (recurrent)	2
Genital aphthosis (recurrent)	2
Skin lesions (recurrent)	1
Central nervous system	1
Vascular manifestations	1
Positive pathergy test	1

Behçet disease score:  $\geq 4$  indicate Behçet disease

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**Figure 1: Aphthous ulcer over the inner aspect of the upper lip. Pretreatment (a), posttreatment (b)**



**Figure 2: Ulcer over the shaft of the penis. Pretreatment (a), posttreatment (b)**



**Figure 3: Oral aphthous ulcer (a), erythema nodosum-like lesions over bilateral shins (b), erythema over the peri-orbital region (c)**

over the right eyelid. In a study conducted by Singal *et al.*, oral ulcers were most common finding seen in 100% of cases, followed by genital and cutaneous lesions seen in 93.1% of cases, among the cutaneous findings erythema nodosum was most common. In a study conducted in the uveitis clinic by Sachdev *et al.*,<sup>[7]</sup> ocular involvement was documented in 92.4% cases and oral ulcers in 88.7%. The occurrence of ocular lesions in BD varies from 50% to 85% and is one of the major causes of morbidity, classical features being acute anterior uveitis.<sup>[1]</sup>

In our case, the pathergy test was negative. Singal *et al.* and Pande *et al.* reported positive pathergy in 31% and 8.6%, respectively. The sensitivity of the pathergy test is reported low in Indian series, nonetheless, high specificity maintains its importance as a diagnostic criterion.

Kharkar *et al.*<sup>[8]</sup> reported a case of 32-year-old HIV-positive female presented with recurrent orogenital and cutaneous lesions for 1 year, differentials of BD, recurrent herpes simplex, and secondary syphilis were considered, positive pathergy test, and histopathological examination confirms the diagnosis of BD. This case highlights both clinical resemblances to secondary syphilis and biological false-positive VDRL test in a setting of HIV infection in a patient with BD, creating diagnostic dilemma.

Kaul *et al.*<sup>[3]</sup> reported a case of BD with erectile dysfunction and depression without any significant drug history, neurological, or local vascular involvement. Depression should be kept in mind as a treatable cause of erectile dysfunction in BD.

Our patient was treated satisfactorily with oral dapsone and systemic corticosteroid in tapering dosage.

Dapsone is used off label for BD with variable efficacy.<sup>[2]</sup> Sharquie<sup>[9]</sup> and Convit *et al.*<sup>[10]</sup> have found good results using dapsone in BD. Singal *et al.* treated 16 cases with Colchicine for 3–12 months with no relapse and seven cases with dapsone for 3–6 months with relapse in two cases. The withdrawal of dapsone resulting in a relapse and the reintroduction controlling the condition suggested the active role of dapsone in BD.<sup>[11]</sup> The role of dapsone in BD is worth evaluating in a large number of cases.

Other immunosuppressive agents such as methotrexate, azathioprine, cyclophosphamide, and chlorambucil can also be used. If these agents fail, tumor necrosis factor inhibitors such as infliximab or etanercept should be considered as the next line of therapy. Recent therapeutic options include tacrolimus, cyclosporine, and interferon- $\alpha$ -2a.<sup>[2]</sup>

## CONCLUSION

BD is an important nonvenereal cause of genital

ulceration. A high index of suspicion in a patient with mucocutaneous lesions may result in the early diagnosis, management, and prevention of complications. A detailed ophthalmologic evaluation in all patients is recommended. In neutrophilic dermatosis such as BD, dapsone offers an efficacious, safe, and economical option.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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