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BEHCET'S DISEASE

Dermatology
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ABSTRACT

Behcet's disease (BD) is a chronic, inflammatory and relapsing vascular disease. Here, we present a case of 23 year old male adult with complaints of recurrent painful oral lesions, cutaneous lesions and diminition of vision. The diagnosis of BD was confirmed by international criteria for BD. The case was treated satisfactorily with systemic corticosteroids and colchicine.

KEYWORDS

Behcet's diseases, panuveitis,

INTRODUCTION

Behcet's disease was first described in 1937 by Hulusi Behcet who described patients with oral and genital ulcers, erythema nodosum and uveitis[1,2]. Later on other clinical features were added to the spectrum. Adamantiads-Behcets disease is a chronic, recurrent, multisystem inflammatory disorder which consists of triad of oral ulcers, genital ulcers, ocular lesions (oculo-oral-genital syndrome)[3]. The etiology is unknown but it is an autoinflammatory systemic vasculitis. It is characterized by mucocutaneous manifestations, ocular manifestations and systemic vasculitis involving veins and arteries of all sizes.

The etiology is unknown, while both genetic and environmental factors play a role in its pathogenesis[4]. Among environmental factors microbial exposure and cellular and humoral immunity play a pathogenic role[5].

The diagnosis is based on international criteria for BD (ICBD)[6]. (TABLE 1)

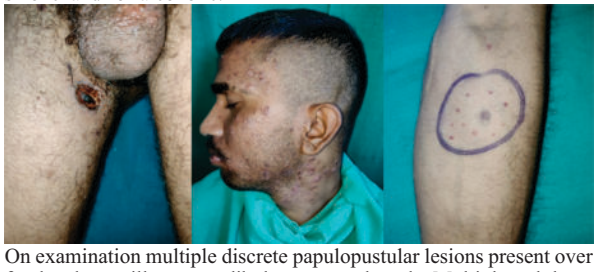
Table 1 : International Criteria For BD

Symptom	Score
Oral ulcer	2
Genital ulcer	2
Ocular manifestations	2
Skin manifestations	1
Vascular manifestations	1
Central Nervous System manifestations	1
Positive pathergy test	1

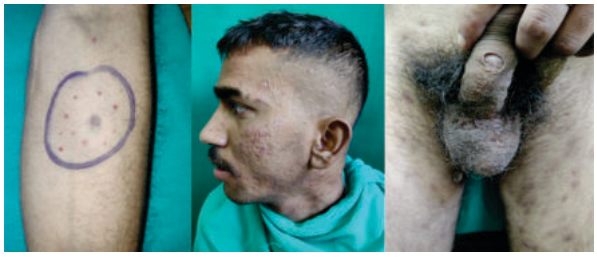
Case Report

A 23 year old male presented with dimintion of vision for the past 9 months, recurrent painful oral ulcers on and off for the past 3-4 years, recurrent genital ulcers for the past 2 years, cutaneous lesions in the form of papulopustular and acneform type for the past 2 years. The patient had redness of eyes and lacrimation for the past 2 years, headache and joint pains in the large joints for the past 2 years.

There was no family history of similar complaints. Patient was non smoker and non alcoholic.



On examination multiple discrete papulopustular lesions present over forehead, maxillary, mandibular area and neck. Multiple aphthous lesions were present over inner aspect of lower lip, angle of mouth and tip of tongue. Single punched out ulcer approximately 3cm×2cm present in the ventral aspect of shaft of penis with well defined borders and having serous discharge. An erosion was present on the scrotum of size approximately 3.5cm×2cm with sero-sanguinous discharge. Single ulcer was present over the inner aspect of right thigh with well-defined borders and reddish colored crusting present over it.



Pathergy test was positive after 48 hrs. Ophthalmic examination showed panuveitis. Routine investigations such as renal function test and liver function test were done which were with in normal limits and complete blood examination showed neutrophillia (89%). HIV antibody test and Veneral Disease Research Laboratory (VDRL) test were non reactive, X-ray chest was normal.

Based on the history and clinical examination diagnosis of behcets disease was considered. The diagnosis was based on revised ICBD criteria, as the patient met the score of eight points (recurrent oral aphthosis -2, recurrent genital aphthosis-2, ocular lesion -2, skin lesions -1and pathergy test positivity -1).

Patient was managed on oral tablet prednisolone 60 mg, tab colchicine 0.5 mg OD, eye drops atropine 1% BD, timolol 0.5 % BD along with supportive treatment. After treatment of three weeks, oral genital and cutaneous lesions showed regression and after 6 weeks complete resolution of genital and oral lesions was there. Oral prednisiolone was tapered over the period of 6 weeks and tablet colchicines was continued for 3 months to maintain remission.

DISCUSSION

Behcets disease is a systemic inflammatory disease. It presents with recurrent oral aphthosis, genital ulcerations, cutaneous lesions and systemic manifestations . Although the exact etiology is unknown, both genetic and environmental factors play a role in the pathogenesis[7]. There is association of BD with HLA B51 suggesting the role of th1 type autoimmunity. BD usually affects young adults between 20 to 40 years of age[8].

In our case oral ulcers along with eye involvement was the presenting feature. Cutaneous lesions in the form of papulopustular lesions were present over face and neck. In a study conducted by singal et al[9], oral ulcers were most common finding followed by genital; and cutaneous lesions. Ocular involvement was seen in 92.4% cases and oral ulcers in 88.7% cases in a study conducted by Sachdev et al[10]. The ocular lesions in BD varies from 50-85% and the major classical lesion being acute anterior uveitis[10].

On our case pathergy test was positive. Singal et al[9] also reported

positive pathergy test in 31% of patients. The high specificity of pathergy test maintains its value in diagnostic criterion.

Our patient was treated satisfactorily with colchicine and systemic corticosteroid in tapering dose. Singal [9] et al treated their 16 cases of BD with colchicines for 3-12 months without any relapse. Other immusuppresants such as azathioprine, methotrexate, chlorambucil, cyclosporine and cyclophosphamide can be used. Other agents such as infliximab, etanercept and tacrolimus can be considered as second line of therapy.

CONCLUSION

BD is an important cause of recurrent oral and genital ulcerations. A detailed ophthalmologic evaluation in all the patient is recommended as it can cause panuveitis. A high index of suspicion in a patient with mucocutaneous lesions can relult in early diagnosis and management of the patients.

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