BEHÇET’S DISEASE: A CASE REPORT


*1Post Graduate, Department of Dermatology, Andhra Medical College, King George Hospital.
2Associate Professor, Department of Dermatology, Andhra Medical College, King George Hospital.
3Assistant Professor, Department of Dermatology, Andhra Medical College, King George Hospital.*

**ABSTRACT**

Behçet’s disease (BD) is a chronic, recurrent, multi-system inflammatory disorder of unknown aetiology, characterised by the triad of oral ulcers, genital ulcers and cutaneous lesions. This association was probably first recognised by Hippocrates, but bears the name of Behçet after his detailed description of this illness in 1937. We are reporting a rare case of Behçet’s disease limited to skin with positive pathergy and showing good response to colchicine.

**KEYWORDS**

Behçet’s, Pathergy, Colchicine.


**INTRODUCTION**

Behçet’s disease (BD) is a chronic, recurrent, multi-system inflammatory disorder of unknown aetiology, characterised by the triad of oral ulcers, genital ulcers and cutaneous lesions. The disease is now recognised to be a multi-system disorder involving mucocutaneous (MC), ocular, intestinal, articular, vascular, urogenital and neurologic systems. The diagnosis of Behçet’s disease is based on the identification of its typical clinical features as per the diagnostic criteria laid down by the International Study Group (ISG). We present a case report of this rare disease & its response to treatment with colchicine.

**CASE HISTORY**

A 32-year-old male patient presented with the chief complaints of painful ulcers in the mouth, genitalia, axillae since 2 months. The present illness started 4 years ago as vesicles & pustules on erythematous skin which ruptured themselves in a span of 4-5 days forming erosions which later ulcerated followed by healing with hyperpigmentation.

Similar lesions appeared over genitalia, oral cavity and axillae in the present episode. Similar episodes 4-5/yr. since 4 years which were slow to heal even with medication. History of formation of ulcers following trauma was present. Patient denies extra marital exposures. On examination multiple ulcers were present on genitalia, oral mucosa, trunk and upper limbs. Pathergy was positive. 2 ulcers were present on the scrotum & ventral aspect of penis (Kissing ulcer) of size approximately 1*2 cm, the ulcers were oval in shape with well-defined edges & floor covered with greyish slough, they were painful. No discharge was present on the ulcer. No discharge from the urethra, Testes were normal, Peri-anal area was normal. In the oral cavity a single ulcer of size approx. 0.5 cm was found on the lateral margin of the tongue, with well-defined borders & clean base & is painful. All the investigations were in normal limits. Ophthalmological examination was normal. SKIN BIOPSY showed stratified squamous epithelium with focal papillary process, acanthosis & elongated rete ridges. Sub epithelial stroma shows dense inflammatory infiltrate composed of lymphocytes, plasma cells, neutrophils. Vessels of all caliber in the stroma show perivascular inflammatory infiltrate with some of them showing transmural inflammation. Some vessels show fibrinoid necrosis. Possibility of Behçet’s is considered correlating with clinical features. We diagnosed the case as Behçet’s syndrome as it satisfied the criteria.

Financial or Other Competing Interest: None.

Submission 11-03-2016, Peer Review 25-05-2016,
Acceptance 30-05-2016, Published 20-06-2016.

**Corresponding Author:**

Dr. K. R. Harsha Vardhan,
Post Graduate,
Department of Dermatology,
KGH, Vizag-530002.
E-mail: harsha.prince99@gmail.com
Recurrent Oral Aphtha

Axillary Lesions

Pathergy Positive

Positive Pathergy

Genital Ulcers after treatment

Oral Ulcers after treatment
DISCUSSION
Behcet’s Disease
Synonym Adamantiaides-Behcet’s disease.

Definition
It is a multisystem disease that is defined by the presence of oral aphthae, genital lesions, posterior uveitis, and cutaneous vascular or meningocerebral involvement. It can be associated with HLA-B51.1 There is a Th1-predominant immune reaction.

Specific triggers found were Streptococcus infection & parvo virus B19. It typically affects young adults, but can occur in children, our patient was a middle aged adult.5

There are very few reports of Behcet’s disease from India.6 This may be explained by two possibilities, either the disease is really uncommon in India or it is simply under-diagnosed and under-reported. Only 2 major studies from north India exist. From south India only a single case series of 12 cases that occurred over a period of 10 years is present.

Clinical Features
Mucocutaneous lesions are the hallmark. The most common presenting features are oral aphthae followed by genital aphthae. The skin manifestations are pseudo-folliculitis and erythema nodosum like lesions. Pathergy phenomenon is present. Eye manifestations are potentially dangerous as they may cause blindness.7 The most common lesion being posterior uveitis. Other systems involved are vascular gastrointestinal neurological pulmonary genital urinary & lymphatic systems.

Our patient present with the oral genital & skin ulcers correlating with the previous studies & case reports.

In a study conducted by Archana Singal Namrata Chhabra et al oral lesions were most common followed by genital lesions with pathergy positive in less than one third of the patients, with eye changes in less than one fifth of patients.

In a case report by Satish Chand Neeraj Srivastava1 two members of the same family presented with oral & genital ulcers & arthralgias

In our case report the patient presented with both oral & genital ulcers simultaneously with positive pathergy so it is comparable to previous studies. Our patient had no eye changes. Histopathology correlated with previous studies

HISTOPATHOLOGY
Three Main Types of Reaction in Behcet’s Disease Namely
1. Vascular.
2. Extravascular with or without vascopathy.
3. A suppurative or mixed suppurative/granulomatous folliculitis.
4. Our patient had histology similar to vascular type with fibrinoid necrosis.

DIAGNOSIS
International Study Group criteria for the diagnosis of Behcet's disease were used for diagnosis in BD there is no relevant biological test for diagnosis. The erythrocyte sedimentation rate, CRP and other acute phase reactants are seldom elevated during the acute phase and/or relapses of BD but are not well correlated with disease activity. Abnormalities of fibrinolysis, elevated factor VIII, immune circulating complexes and cryoglobulinemia have been occasionally reported. Leucocytosis is frequently encountered. The positivity of HLA B 51 allele is of no diagnostic value. Cutaneous biopsy of intradermal injection with physiologic saline solution may demonstrate vasculitis with immune complexes deposit.

TREATMENT
Due to the lack of an etiologic agent, the treatment is symptomatic without consensus.

The goals are the functional recovery of visceral involvement (eye, CNS) and prevention of relapse(s).

The risks of BD are an increased mortality especially in case of arterial involvement, and a high morbidity due to the cumulative sequel of ocular and neurological involvement

Oral or genital ulcers may be treated symptomatically with topical or intralesional corticosteroids, topical sacralute gel, topical tacrolimus, viscosid lidocaine (Lignocaine). Nicotine patches, Colchicine (0.6 mg two to three times daily), Dapsone, Dapsone in combination with colchicines, Systemic corticosteroids, azathioprine, methotrexate, cyclosporine, Cytokines such as interferon-α are useful (Alone or with azathioprine), thalidomide, intravenous immunoglobulin (IVG) with aspirin, and granulocyte and monocyte adsorption apheresis, anti-TNF agents such as infliximab or etanercept.

Novel treatments under investigation are induction of tolerance using oral administration of the 336–351 sequence of HSP60 linked to recombinant cholera B toxin A subunit.

We treated our patient with T. colchicine 0.5 mg bid and had an excellent response in two weeks without any recurrences in follow up till now.

Therefore, we are presenting this case on account of its rarity and moreover the disease is limited to skin without any internal systemic involvement which is very rare and had an excellent response to colchicine.

PROGNOSIS-BD
Significantly increases morbidity and mortality. The leading causes of morbidity in BD are the uveitis with the potential threat of visual loss and neurologic involvement

Main causes of death included major vessel disease (Mainly arterial aneurysm and Budd-Chiari syndrome) (43.9%), cancer and malignant hemopathy (14.6%), and central nervous system involvement and sepsis (12.2%).

The mortality rate at 1 and 5 years was of 1.2% and 3.3%, respectively.11

REFERENCES