

Familial Behcet's Disease Involving Four Members in a Family - Maximum Numbers in a Family Published in Indian Literature till Date

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Abstract

There are very few reports of familial behcet's from India. We report here familial Behcet's disease from India in two brothers, one sister, and their father. They had recurrent orogenital ulcers. Pathergy test was positive in all. This is the maximum no of members involved in a family published from India.

Keywords: Oral ulcers; Behcet's disease; Genital ulcers; Pathergy test

Introduction

Behcet's disease is an inflammatory disease having unknown origin presenting with recurrent oral and genital ulcers and ocular involvement. Its prevalence is highest along silk route. Familial aggregation of the disease has been reported mainly from Turkey and Japan. We here report two brothers, one sister and their father with recurrent orogenital ulcers due to Behcet's disease.

Case Report

56 year male is suffering from recurrent orogenital ulcers for last 12 years. He is married to 52 years unrelated female (non-consanguineous marriage). She has no features of behcets disease. They have 5 children. 32 year male first in birth order has no features of behcet's at present. 30 year female second in birth order is having recurrent orogenital ulcers. 27 male third in birth order is having recurrent orogenital ulcers for last 5 years. He was treated for uvetis left eye with azathioprine 2 years before. 26 female fourth in birth order, has no features of behcet's at present. 24 male fifth in birth order is having recurrent orogenital ulcers for last 2 years. His slit lamp examination revealed uvetis of right eye. All the involved members of the family were subjected to pathergy test and found to be positive in all. All four involved patients were fulfilling international study group criteria for behcets disease. All had recurrent oral ulcers (more than 3 times in a year) and recurrent genital ulcers with positive pathergy test sufficient to diagnose behcets disease. In addition third and fifth were also having uveitis, in 3rd sibling managed with azathioprine and in 5th being managed with topical steroids and azathioprine. None had arthritis, pulmonary, cardiac or renal involvement. Blood samples have been taken for genetic testing from all involved and uninvolved persons for hlab51 and hla-dr1, hla-dqw1 respectively. Episodes of mucocutaneous manifestations respond to colchicine.

Discussion

Behcet's disease is a systemic autoimmune vasculopathy manifesting usually by recurrent orogenital ulcerations. The disease also involves eyes. Genital ulcers heal by leaving behind scars. Pathergy test an

excessive response to needle prick is a unique manifestation of Behcet's disease. Besides skin vasculitic lesions have been seen in eyes, CNS, G1 system, bones, kidneys and large blood vessels. Although pathogenesis of BD remains unknown-heredity, immunologic factors, infectious agents and inflammatory mediators are likely contributors.

The ISG for Behcet's disease has developed diagnostic criteria which require the presence of oral ulcerations plus any two of the following: genital ulcers, typical defined eye lesions, typical defined skin lesions or a positive pathergy test [1]. In our case all patients had positive pathergy test, unusual with Indian population involved by Behcet's disease. Majority of familial cases are from Turkey [2] and Japan [3]. From India a few instances of familial occurrence are mentioned, however detailed clinical features except in two cases mother and her daughter were mentioned. In addition to above two brothers in a family are also reported. Behcet's disease has been known to be strongly associated with HLA B51, one of the split antigens of HLA-B5 [4]. Recent studies have suggested that there is no association between factor V laiden mutation and DVT in Behcet's disease [5], while ICA-1 genes [6] and MEFV mutation [7] may contribute to the vascular inflammatory changes associated with Behcet's disease. Thus for mother, daughter in one paper [8], two brothers in the next paper [9], and two brothers, one sister and their father in our paper suggest familial Behcet's along with sporadic familial cases rare in India may not be actually so.

We believe heightened awareness of this condition among clinicians is likely to generate and depict the actual magnitude of the controllable disease. Treatment of BD should be based on degree of systemic involvement.

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