

Familial Behçet's disease

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ABSTRACT

There are very few reports of Behçet's disease from India. Familial aggregation of Behçet's disease has been reported with restricted geographical distribution. We report here familial Behçet's disease from India in two brothers aged 30 and 32 years. Both patients had recurrent oral and genital ulcers for approximately five years. They also had arthralgias on and off along with fever. Pathergy test was positive in both cases. Their younger brother and a sister had recurrent oral aphthous ulcers.

Key words: Aphthous ulcers, Behçet's disease, Pathergy test, Vasculitis

INTRODUCTION

Behçet's disease is an inflammatory disease of unknown origin presenting with recurrent oral and genital ulcers and ocular involvement. Its prevalence is highest in countries of the eastern Mediterranean, middle East and the eastern Asian rim. Familial aggregation of the disease has been reported mainly from Turkey^[1] and Japan.^[2] We describe two brothers aged 30 and 32 years with recurrent oral and genital aphthous-like ulcers due to Behçet's disease.

CASE REPORT

Two brothers aged 30 and 32 years, from Sasaram district, Bihar, presented with recurrent oral and genital aphthous-like ulcers since the last five years. They also had arthralgias on and off for one year and 4½ years respectively, along with mild grade fever. Their younger brother and a sister also had recurrent oral aphthous ulcers. There was no history of extramarital sexual contact in either case.

Blood investigations revealed mild leucocytosis and raised erythrocyte sedimentation rate (32 and 40 mm in the first hour, respectively). Tzanck smear did not show features

of herpes infection. Pathergy test was done by oblique insertion of a 20-gauge needle intradermally under sterile conditions. Papules formed at the injection sites in both cases at 48 hours. The papules were punch biopsied and histopathological examination showed leukocytoclastic vasculitis. Ophthalmological examination was normal.

Diagnosis of Behçet's disease was made according to the diagnostic criteria developed by the International Study Group for Behçet's Disease.^[3] Patients were prescribed topical triamcinolone in orabase for oral lesions, clobetasone butyrate cream for genital lesions and dapsone 100 mg thrice daily orally. The ulcers healed in about four weeks but new ulcers appeared after a month despite continued treatment. After the relapse, dapsone was stopped and oral prednisolone (40 mg/day) was added. In about two weeks, the ulcers healed completely and prednisolone was tapered to 40 mg on alternate days. No new ulcers have developed in two months' follow-up.

DISCUSSION

Behçet's disease is a systemic autoimmune vasculopathy manifesting usually by recurrent oro-genital aphthous-

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like ulcerations and eye findings.^[1] There are no specific manifestations or special diagnostic tests for the disease. Aphthous oral ulcers are usually the first and most characteristic clinical feature. Genital ulcers resemble oral ulcers and may be single or multiple. Pathergy, an excessive skin response to trauma, is a unique manifestation of Behçet's disease reflecting neutrophil hyper-reactivity. Besides skin, vasculitic lesions can be seen in eyes, central nervous system, gastrointestinal system, bones, kidneys and large blood vessels.

The diagnosis of Behçet's disease depends on the recognition of a number of typical clinical findings as well as exclusion of common mucosal ulcerative disorders. The International Study Group for Behçet's disease has developed diagnostic criteria which require the presence of oral ulceration plus any two of the following: genital ulceration, typical defined eye lesion, typical defined skin lesion or a positive pathergy test.^[3] These criteria are most commonly used to diagnose the disease. It has been reported that the pathergy test is rarely positive in Indian patients with Behçet's disease.^[4] However, both of the present cases tested positive for pathergy.

Behçet's disease occurs primarily in young adults with the mean age of onset being 25-30 years. The highest prevalence of Behçet's disease is seen in Japan, Korea, China, Iran and Turkey.^[5] The prevalence for a population of 100,000 is 13.4 in Japan, 14 in China, 16.7 in Iran, 20 in Saudi Arabia, 80 to 370 in Turkey and much less in other countries. There are only a few reports of Behçet's disease from India.^[4] This may be explained by two possibilities, either the disease is really uncommon in India or it is simply under-diagnosed and under-reported. Most cases of Behçet's disease are sporadic and the parents of patients are usually unaffected^[1] although a familial aggregation of Behçet's disease has been reported.

A peculiar geographical distribution and a familial aggregation have been regarded as evidence supporting a genetic influence on the pathogenesis of Behçet's disease.^[1] However, the rarity of Behçet's disease in Japanese immigrants to Hawaii shows the greater importance of environmental factors^[5] rather than genetic factors. A higher incidence of familial aggregation was noted in juvenile patients with Behçet's disease.^[6] Majority of reports of familial cases are from Turkey^[1] and Japan.^[2] In a paper on Behçet's disease from India,^[4] a few instances of familial occurrence were mentioned. However, detailed clinical features of these cases, except in two cases—a mother and her daughter, were not described. From this paper, it appeared that oral ulcers in family members were also considered to be cases of familial occurrence.

In studies from abroad^[7] but not from India,^[4] Behçet's disease has been known to be strongly associated with human leukocyte antigen (HLA) B51, one of the split antigens of HLA-B5. Recent studies have suggested that there is no association between factor V Leiden mutation and deep vein thrombosis in Behçet's disease,^[8] while ICAM-1 genes^[9] and familial Mediterranean fever gene (MEFV)^[10] mutation may contribute to the vascular inflammatory changes associated with Behçet's disease. Behçet's disease is considered to be rare in India.^[4] The two brothers reported here presented with recurrent oral and genital aphthous-like ulcers. They also had periodic arthralgias. Pathergy test was positive in both cases. Also, their one younger brother and a sister have recurrent oral aphthous ulcers. This paper suggests that Behçet's disease in India including its familial occurrence may not be as rare as is generally believed. Heightened awareness of this condition among the clinicians is likely to generate more data on Behçet's disease in India.

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