# BEHCET'S SYNDROME
## A Case Report

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**Summary**

A patient with classical features of Behcet's Syndrome is presented. The case which initially had ophthalmological manifestations posed great difficulty in diagnosis until the genital and oral ulcerations appeared. Systemic manifestations in the form of arthritis and hepatomegaly appeared still later. Neurological involvement set in as a terminal event.

Behcet's Syndrome is a progressive multisystem disease characterized by triad of recurrent ulcerations of oral mucosa, external genitalia and recurrent uveitis that is frequently complicated by hypopyon. It is not necessary that all three manifestations should be present together and one may precede the other by variable period. Many other manifestations have been added to the original triad and involving various systems of body. These include various skin lesions, arthritis, orchitis, epididymitis, venous and arterial thrombosis, pericarditis and various lesions of the central nervous system and gastrointestinal tract. Usually this syndrome runs a benign course except recurrent uveitis, which almost invariably leads to blindness[1]. The life-threatening manifestations are rare and include central nervous system[2] and gastrointestinal[3] lesions.

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### Case Report

A twenty two years old male reported in November 1976 with signs and symptoms suggestive of anterior uveitis. It deteriorated inspite of treatment, resulting in panuveitis in about six months' time. Cataract also had developed in both eyes. Two months later patient developed febrile polyarthritis which started with the left knee joint. Systemic corticosteroids were of no benefit. In another two months' time, cutaneous manifestations viz lesions at the site of injections, punched out ulceration on scrotum and gluteal region had appeared. These ulcerations healed with scar formation. Mucosal ulcerations involving the tongue, eyes and palatal mucosa (Fig. 1) followed suit. Arthritis got worse. Hepatomegaly also developed. Secondary glaucoma complicated panuveitis and patient lost the vision. Pupils were immobile due to synechiae. In September 1977 he developed neck rigidity and left facial palsy and soon became comatose and died.

### Investigations

Hemoglobin, total and differential white blood cell count, on admission

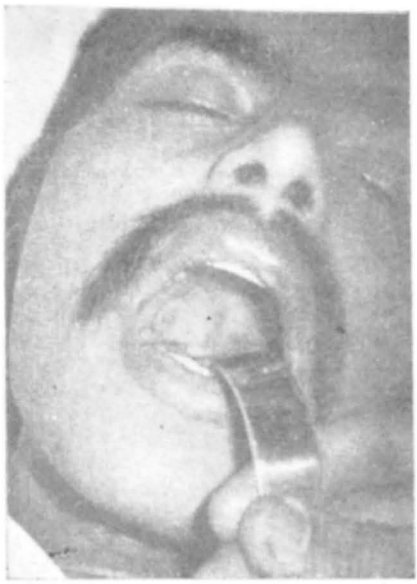

**Fig.** Necrotic lesion in buccal mucosa

were within normal limits. ESR was 75 mm. in first hour. Platelet count remained low throughout, the highest reading was 141500 cells/c.mm. Rose-Waller test was positive 128 dilution. LE cell phenomenon was negative. Urine and blood culture remained sterile. Skiagram of chest did not show any abnormality. X-ray of knee joint showed evidence of mild osteoarthritis. CSF done in September '77 revealed leucocytosis, increase in protein (200 mg/100 ml) mainly due to globulin. The haemoglobin level dropped to 5 gms% towards the terminal stage of the disease. Biopsy of the oral ulcer showed complete loss of epithelium and a dermal infiltrate consisting of lymphocytes, plasma cells and few polymorphs.

As the course of the disease was fulminant, and the patient moribund, immunological and other investigations were not done.

**Treatment**

Systemic and oral corticosteroids, local instillations of cortisol for the eye, systemic broad spectrum antibiotics, blood transfusion and gammaglobulin injections were given. However, patient steadily deteriorated and died.

**Discussion**

Berlin[4] who reviewed Behcet's cases adopted a diagnostic criteria that included only two out of the three cardinal signs viz iritis, genital / oral ulcer, and one of the other recognised signs[5] like increased susceptibility to pyoderma, thrombophlebitis, arthropathy, neurological and visceral involvement like hepatomegaly. The present case developed ocular manifestations which became chronic and progressive. Later many other features included among major and / or minor criteria[6] manifested. Neurological manifestation with CNS involvement and CSF abnormalities set in as the terminal event.

The appearance of pustular lesions at injection sites gave a clue to diagnosis in this case.

The cause of Behcet's syndrome is unknown but there is evidence to suggest some abnormalities in immune mechanism. Complement consumption by classical pathways may occur in Behcet's syndrome since low levels of C2, C4 and C3 have been found before an attack of uveitis. William et al[8] suggested that immune complexes are more common with neuro-ocular type of Behcet's syndrome than with mucocutaneous type. Immune complexes were also associated with active disease. These support the hypothesis that formation of immune complexes is an important step in the pathogenesis of Behcet's syndrome.

This case is being presented on account of its rarity.

**References**

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