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Behçet's Syndrome: Diagnosis of Exclusion

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Abstract

Behçet's syndrome is a rare immune-mediated disease often presents with mucous membrane ulceration and ocular problems. Nearly all patients with Behçet's syndrome present with some form of painful oral mucocutaneous ulcerations in the form of aphthous ulcers. The cause is not well-defined; the primary mechanism of the damage is autoimmune. Treatment is aimed at easing the symptoms, reducing inflammation, and controlling the immune system. Here we present a case of Behçet's syndrome with 2 week follow up.

Keywords: Behçet's Syndrome; Autoimmune Disorder; Oral Ulcers; Patehrgy Reaction.

Behçet's Syndrome - Diagnosis of Exclusion

Behçet's syndrome is a rare immune-mediated disease often presents with mucous membrane ulceration and ocular problems. In 1937 Turkish dermatologist Hulusi Behçet, who first described the triple-symptom complex of recurrent oral aphthous ulcers, genital ulcers, and uveitis [1]. As a systemic disease, it can also involve visceral organs such as the gastrointestinal tract, pulmonary, musculoskeletal, cardiovascular and neurological systems [1,2].

Nearly all patients with Behçet's syndrome present with some form of painful oral mucocutaneous ulcerations in the form of aphthous ulcers. The sores usually heal in one to three weeks. Painful genital ulcerations usually develop around the anus, vulva, or scrotum. Inflammation in blood vessels (veins and arteries) may occur in Behcet's disease, causing redness, pain, and swelling in the arms or legs [2,3,4].

The cause is not well-defined, the primary mechanism of the damage is autoimmune. For a patient to be diagnosed with Behçet's disease, the patient must have oral (aphthous) ulcers (any shape, size, or number at least 3 times in any 12 months period) along with 2 out of the following 4 hallmark symptoms [1,4]:

- Eye inflammation (iritis, uveitis, retinal vasculitis,)
- Genital ulcers (anal ulcers and spots in the genital region and swollen testicles or epididymitis in men)
- Pathergy reaction (papule >2 mm dia. 24-48 hrs or more after needle-prick).
- Skin lesions (papulo-pustules, folliculitis, erythema nodosum, acne)

The diagnosis can sometimes be reached by pathologic examination of the affected areas. A large number of serological studies show a linkage between the disease and HLA-B51 [5]. Complications of Behcet's syndrome depend on your signs and symptoms. For instance, untreated uveitis can lead to decreased vision or even blindness.

Treatment is aimed at easing the symptoms, reducing inflammation, and controlling the immune system. High-dose corticosteroid therapy is often used for severe disease manifestations.

Topical antifungal therapy is also advised.

People with oral, genital and eye signs and

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symptoms of Behcet's disease need to visit doctor regularly to prevent complications. Here we present a case of Behçet's syndrome with 2 week follow up.

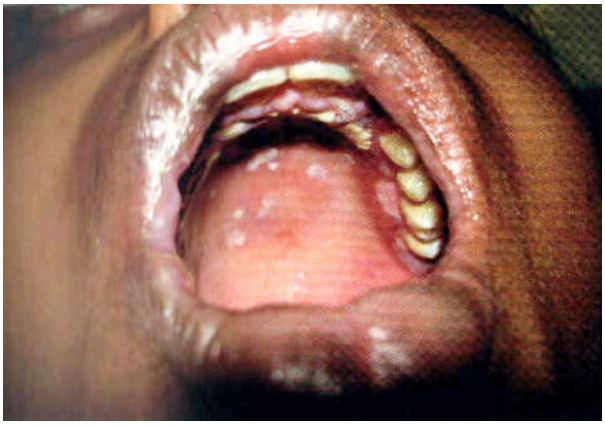

Fig. 1: Multiple ulcers on palate

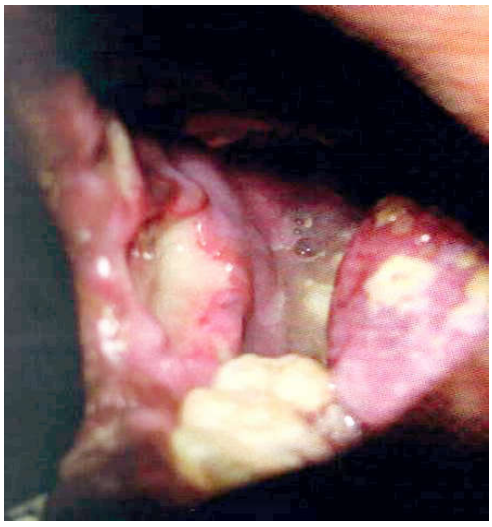

Fig. 2: Major aphthous ulcer ulcers on left buccal muccosa

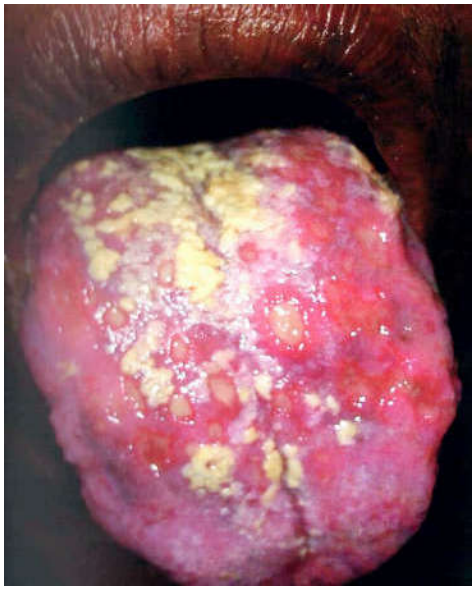

Fig. 3: Candidiosis on tongue

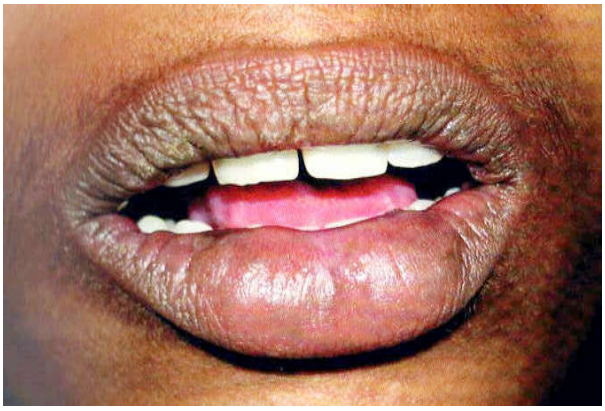

Fig. 4: Healing lesions over lip & left buccal mucosa (1 week)

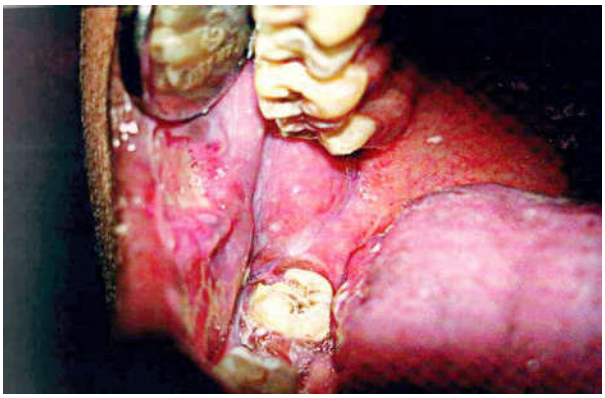

Fig. 5: Healing lesions over lip & left buccal mucosa (1 week)

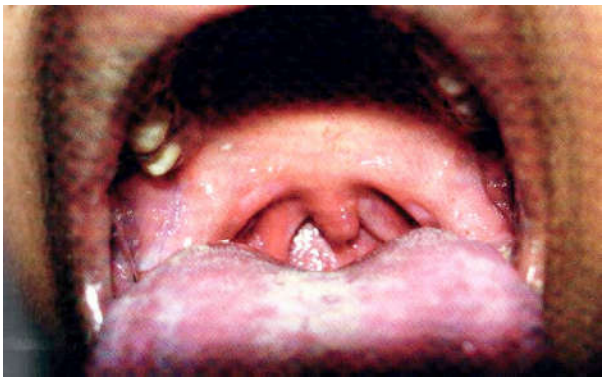

Fig. 6: Healing lesions over palate (1 week)

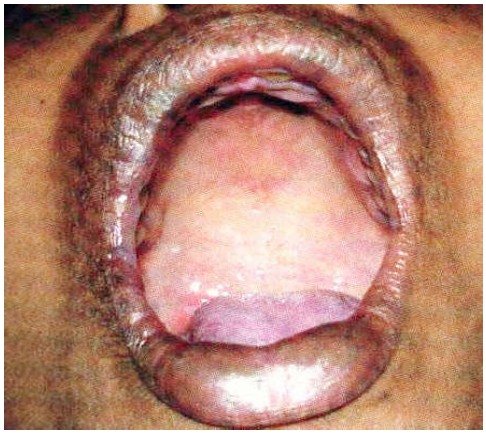

Fig. 7: Healing lesions over palate (1 week)

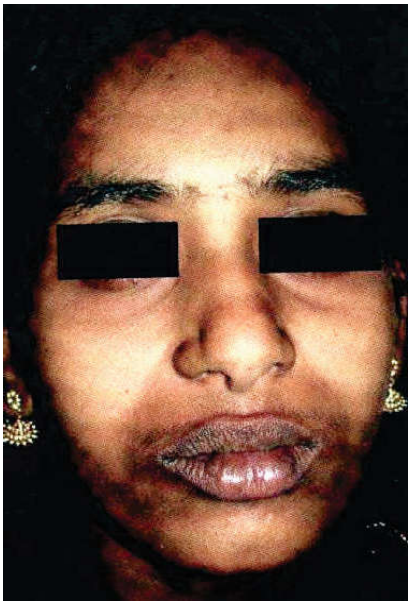

Fig. 8: Healed lesions (2 week)

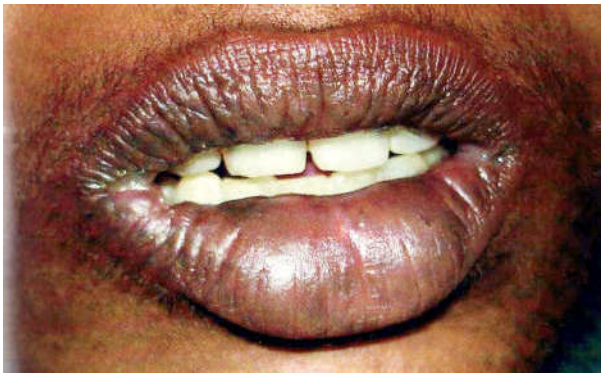

Fig. 9: Healed lesions on lips (2 week)

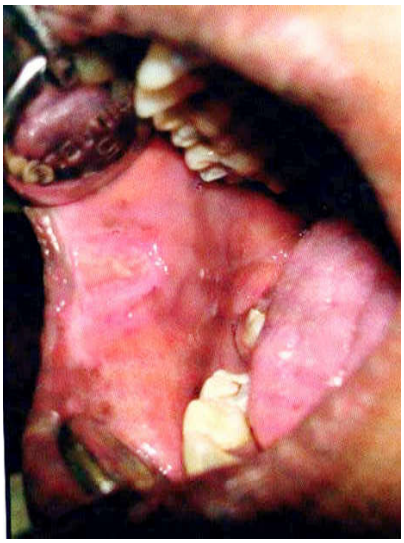

Fig. 10: Healed lesions on buccal mucosa (2 week)

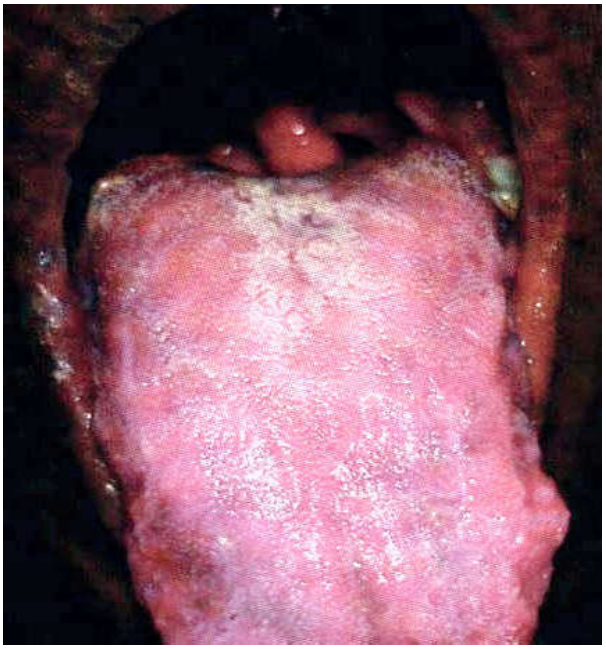

Fig. 11: Healed lesions (2 week)

Key Messages

The diagnosis can sometimes be reached by pathologic examination of the affected areas. A large number of serological studies show a linkage between the disease and HLA-B51.

Conflict of Interest: nil

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