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Behçet's Disease - A Rare Form of Small Vessel Vasculitis

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Abstract

Behçet's disease is a rare vasculitis of unknown aetiology though there is a strong genetic association with HLA-B51 characteristically targets small arteries and venules. Multiple deep oral ulcer and genital ulcer is a common presentation. Ocular involvement is also commonly occurs in the form of anterior or posterior uveitis or retinal vasculitis. Neurological manifestation occurs in the form of brainstem or pyramidal tract symptom. Thrombosis can occur but renal involvement is unlikely. Our patient Mrs X, 47 year old has been suffering from recurrent episode of oral ulcer, genital ulcer, redness of eye and dysphagia for the last 12 years. T here was relapse and remission of the symptoms. Her Ophthalmoscopic examination was normal. Upper GI endoscopy revealed multiple esophageal ulcers. ANA, Anti CCP, cANCA, pANCA, HSV 1&2- IgM, CMV- IgM, TPHA, Anti -HIV 1&2 were negative.

She was treated with oral steroid and azathioprine and all the lesions resolved completely.

Key words: *BS, oral ulcer, genital ulcer, eye lesion*

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Introduction

Described in 1937 by the Turkish dermatologist Hulusi Behçet as a triad that associated uveitis with oral and genital aphthosis, it had been known since the fifth century BC, and its description can be found in the Hippocratic third book of endemic diseases. Since then, other manifestations have been described and added besides new aspects of epidemiology, immunopathogenesis, and treatment[1].

Behçet's syndrome is a multisystem disorder presenting with recurrent oral and genital ulcerations as well as ocular involvement. The diagnosis is clinical and The syndrome affects young males and females from the Mediterranean region, the Middle East, and the Far East, suggesting a link with the ancient Silk Route. Males and females are affected equally, but males often have more severe disease. Blacks are very infrequently affected. The main pathologic lesion is systemic perivasculitis with early neutrophil infiltration and endothelial swelling. Vasculitis resulting in occlusion of the vessels supplying the optic nerve may be the cause of acute optic neuropathy and progressive optic atrophy in Behçet's disease[3]. Oral and genital ulceration can be managed with a combination of colchicine and topical glucocorticoid preparations. Colchicine can be effective for erythema nodosum and arthralgia. Glucocorticoids and immuno-suppressants such as azathioprine are indicated for uveitis, recurrent venous thrombosis and neurological disease.

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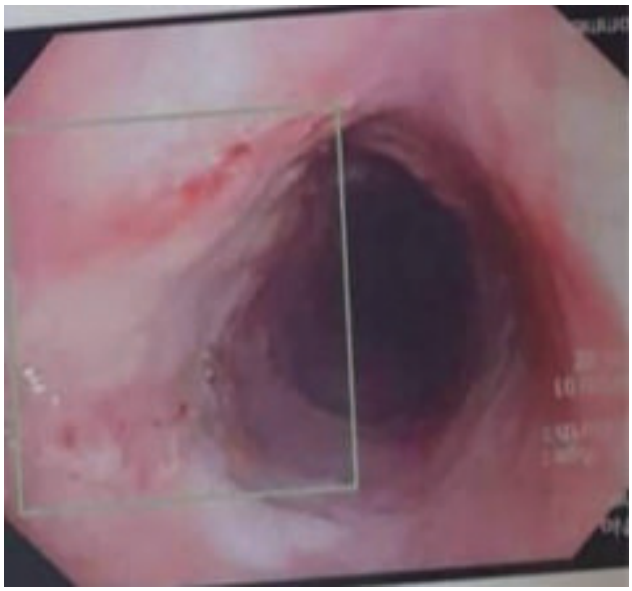
Case Summary

A 47 years old, married Muslim female, normotensive, nondiabetic hailing from Kaunia, Rangpur presented with recurrent episode of oral ulcer, genital ulcer, redness of eye and dysphagia for the last 12 yrs. There was relapse and remission of the symptoms. The oral ulcers were painful, persist for 1-2 wks and then subside without leaving any scar. She also complained of painful genital ulcer. The patient complained of redness of eyes with minimal eye ache but there was no visual difficulty. She also complained of dysphagia to both solid and liquid which was intermittent and not associated with regurgitation, heart burn, chest pain, nocturnal cough or weight loss. There was no history of joint pain, skin rash, cough, hemoptysis, headache, vomiting,

focal neurological deficit, urethral discharge, history of travelling abroad or multiple sexual exposure. Her physical examination revealed oral and genital ulcers, redness of eyes with normal fundoscopy. On investigation, her complete blood count revealed normal total count with 9% eosinophil, Hb level was 10.5 g/dl, ESR was 42 mm in 1st hour and RBS was 5.20 mmol/l. Her Chest x-ray was normal.

Upper GI endoscopy revealed multiple esophageal ulcer.
Oesophageal ulcer in Behcet's disease



ANA, Anti CCP, cANCA, pANCA, HSV 1&2- IgM, CMV-IgM, TPHA, Anti HIV- 1&2 were negative.

She was treated with oral steroid and azathioprine and all the lesions resolved completely.

(Phtograph taken with permission)
Complete resolution of oral and eye lesion after treatment

Discussiopn

This is a vasculitis of unknown aetiology that characteristically targets small arteries and venules. It is rare in Western Europe but more common in 'Silk Route' countries, around the Mediterranean and in Japan, where there is a strong genetic association with HLA-B51. In the Silk Road countries, Behçet's disease is more common in men than in women. However, in Western Europe and the U.S., the trend is reversed: more women have BD than men. Symptoms typically develop when patients are in their 20s and 30s; however, the disease has been seen in all ages, from infants to the elderly. The incidence is around 1 in 10,000 people in silk road countries and 1 in 20,000 in USA[4]. Oral ulcers are invariably present. The ulcers are usually painful, are shallow or deep with a central yellowish necrotic base, appear singly or in crops, and are located anywhere in the oral cavity. The ulcers persist for 1–2 weeks and subside without leaving scars. The genital ulcers are less common but more specific, are painful, do not affect the glans penis or urethra, and produce scrotal scars. The usual skin lesions are erythema nodosum or acneiform lesions, but migratory thrombophlebitis and vasculitis also occur. Nonspecific skin inflammatory reactivity to any scratches or intradermal saline injection (pathergy test) is a common and specific manifestation. Eye involvement with scarring and bilateral panuveitis is the most dreaded complication, since it occasionally progresses rapidly to blindness. In addition to iritis, posterior uveitis, retinal vessel occlusions, and optic neuritis can be seen in some patients with the syndrome. Vasculitis resulting in occlusion of the vessels supplying the optic nerve may be the cause of acute optic neuropathy and progressive optic atrophy in Behçet's disease[3]. Nondeforming arthritis or arthralgias are seen in 50% of patients and affect the knees and ankles[2]. Neurological involvement occurs in 5% and mainly involves the brainstem, although the meninges, hemispheres and cord can also be affected, causing pyramidal signs, cranial nerve lesions, brainstem symptoms or hemiparesis[2]. CNS involvement in Behçet's may lead to intracranial hypertension most commonly due to dural venous sinus thrombosis and subsequent secondary optic atrophy[5]. Recurrent thromboses also occur. Renal involvement is extremely rare.

Pulmonary artery vasculitis presenting with dyspnea, cough, chest pain, hemoptysis, and infiltrates on chest roentgenograms has been reported in 5% of patients and should be differentiated from thromboembolic disease since it warrants anti-inflammatory and not thrombolytic therapy. Laboratory findings are mainly nonspecific indices of inflammation, such as leukocytosis and elevated erythrocyte sedimentation rate, as well as C-reactive protein levels.

Conclusion

Behçet's disease is a small vessel vasculitis of unknown aetiology and it can be treated effectively with glucocorticoid and immunosuppresants. As it has a dreadful complication like blindness and it has effective treatment with expected outcome, so it should be considered as an important differential when we are dealing a patient with oral, genital ulcer with skin and eye lesion.

Conflict of interest- None

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