Behcet's disease presenting as recurrent ocular oral and scrotal lesions in a young man

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Abstract
Behcets disease is a multisystem disorder which requires a careful constellation of symptoms for making a clinical diagnosis. We report a young man presented with reduced visual acuity to perception of light on left side with history of recurrent orogenial ulcers. Treatment with steroids azathioprine and colchicine helped in immediate recovery of mucocutaneous lesions with only a little improvement in vision. Recurrent posterior uveitis resulted in permanent visual loss.

Keywords: recurrent, oral, Behcet's, scrotal

Introduction

36 year old Airman was found to have reduced visual acuity (VA) on left eye to perception of light on a routine medical examination. He has had recurrent red eyes, scrotal ulcers over last four years which had being treated with topical steroids and antibiotics at outpatient clinics.

Ophthalmology examination revealed a clear cornea with quiet anterior chamber but, evidence of previous anterior uveitis by the presence of posterior psynachea and pigment deposits at anterior lens capsule. Posterior segment of left eye was remarkable with multi focal choroiditis and vasculitis (figure 1). There was epi-retinal membrane formation at macula with macula edema due to traction (figure 2). Optic disc was pale and there were no vitritis. Active oral ulcers (figure 3) scrotal lesions (figure 4) were observed along with a few partially healed acne form papular eruptions in lower limbs. Pathergy test was positive (formation of a sterile erythematous papule 2 mm or larger in 48 hours following a skin prick with a sharp sterile needle). Skin biopsy from an acne form lesion showed mild ortho keratosis and irregular acanthosis in epidermis. Dermis showed moderate perivascular lymphoplasmacytic infiltrate with focal neutrophilic infiltrations compatible with BD.

The diagnosis of Behcet's disease (BD) was made based on clinical picture; recurrent orogenital ulcers, uveitis, and positive pathergy test. Patient did not have renal, cardiac, pulmonary, gastrointestinal or neurological manifestations.

He was immediately commenced on topical prednisolone acetate 1% drops 5 hourly, oral prednisolone, colchicine and azathioprine. One week in to treatment, orogenital ulcers resolved. VA improved to hand movement and vasculitis resolved completely. Chronic macular edema and epi-retinal membrane hampered full recovery of VA.

Discussion

BD is a multi-organ systemic disorder with a remitting relapsing course. In the absence of a pathognomonic test, assemblage of clinical signs make the diagnosis. International Criteria for Behcet's Disease (ICBD) has been developed as a guide for diagnosis [1]. Our patient scored 8 points for oral genital apthosis, ocular and skin lesions, along with pathergy test. Score of 4 or more is classified as positive in ICBD criteria.

This patient has had recurrent uveitis over years which remained untreated. Recurrent posterior uveitis has resulted in lasting damage and lead to permanent visual loss. Bechet's ocular manifestations include recurrent granulomatous panuveitis and retinal vasculitis [2]. Presence of posterior uveitis has a prognostic value [2].

Treatment of BD based on the severity of the disease and the extension of multisystem involvement. Our patient had mucocutaneous and ocular symptoms. Colchicine is a preferred drug in mucocutaneous lesions [2]. Steroid eye drops, subconjunctival steroid injections or systemic steroids are used in acute ocular attacks depending the severity of inflammation.

Recommended treatment for posterior uveitis include azathioprine, cyclosporine-A, interferon-alpha or monoclonal anti-TNF antibodies [3]. Oral steroids are only recommended in combination with azathioprine or other systemic immunosuppressive [3]. We immediately commenced this patient on a combination of topical systemic steroids with colchicine and azathioprine. His steroid dose was gradually tapered over 3 months. He remained in remission for more than 12 months now.

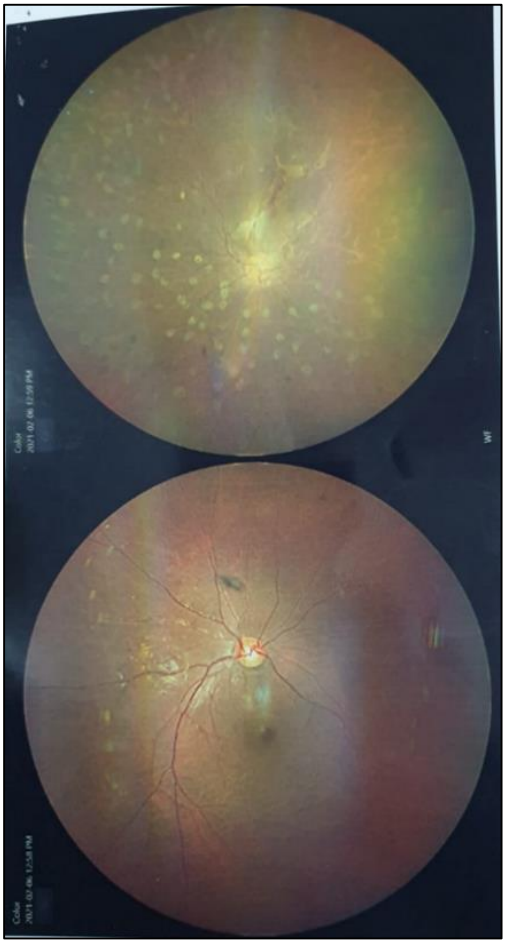

Fig 1: Multifocal choroidits with pale optic disc and evidence of old retinal vasculitis

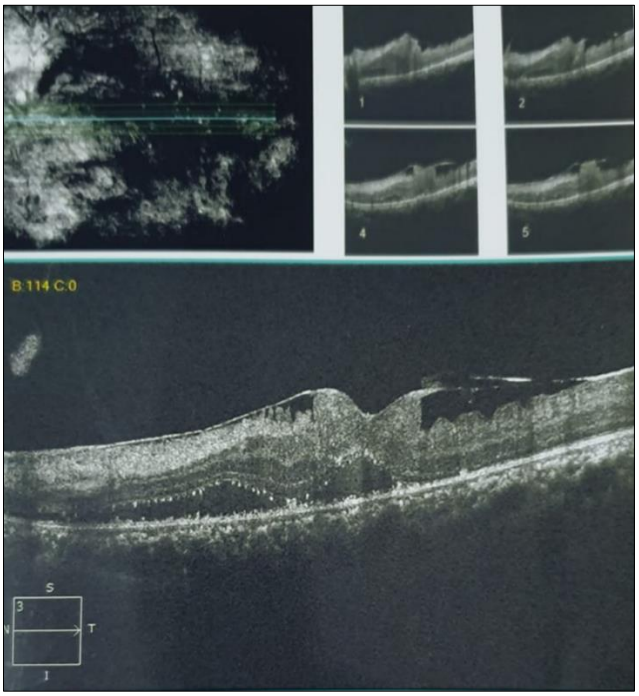

Fig 2: Ocular coherence tomography of macula which shows EPI retinal membrane causing macular tension and Oedema

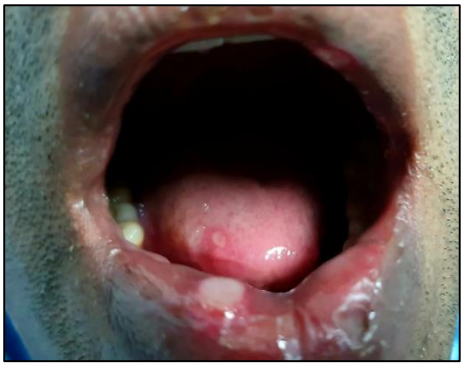

Fig 3: Multiple apthous ulcers of varying sizes and ages.

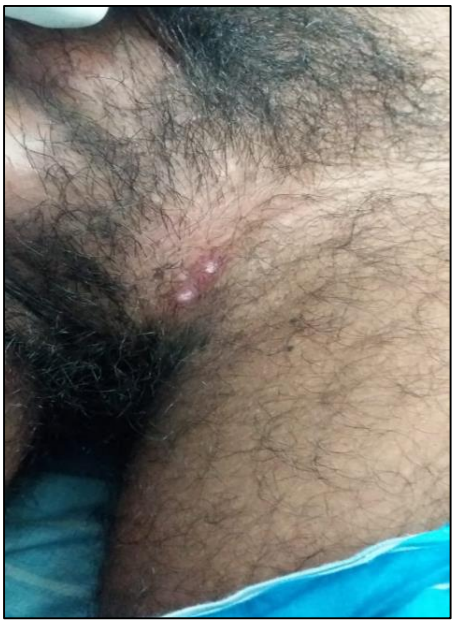

Fig 4: Apthous like ulcers next to the scrotal skin in the groins

Conclusion

Delay in making the correct diagnosis missed early therapeutic intervention in this patient. Meticulous clinical evaluation and a multi-disciplinary approach are essential for early diagnosis of BD.

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