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Vision loss in behcets disease

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Abstract
Behçet's disease (BD) is a chronic systemic inflammatory disorder affecting venules. A 25-year-old male was referred to the Ophthalmology clinic with two-week history of loss of vision in both eyes. On examination there were oral and genital ulcers along with fundus findings. Diagnosis of Behcet's disease was made following investigations. He was advised vitreoretinal surgery, topical steroids, systemic corticosteroids and cyclophosphamide. This case report suggests that high index of suspicion helps timely diagnosis and early treatment can prevent blindness.

Keywords: vasculitis, ulcer, autoimmune, inflammation

Introduction
Behcet's disease is a systemic vasculitis of unknown etiology characteristically affecting venules. Onset is typically in young adults with recurrent oral and genital ulceration, uveitis, skin manifestations, arthritis, neurological involvement, and a tendency to thrombosis.

Case Report
A 25-year-old male was referred to the Ophthalmology clinic with four weeks history of gradual, painless loss of vision in both eyes. On physical examination patient was calm, conscious, oriented, with pulse rate of 80 per min and blood pressure of 130/80 mm Hg. On further examination of the individual revealed presence of large oral ulcer with a hemorrhagic base, on the uvula and posterior most part of soft palate. Genital examination also showed multiple ulcers on penile shaft, scrotum and surrounding part of upper thighs. On ophthalmologic examination extraocular motility was full, without diplopia and extra ocular muscle limitation. Pupils were normal sized, reactive to light; with no relative afferent pupillary defect. Iris was normal colored with normal pattern. Intraocular pressure was 14 mm Hg in right eye and 16 mm Hg in the left. Conjunctiva was normal without any congestion. Cornea was clear. On slit lamp examination there were few pigmented keratic precipitates in both eyes without presence of pus cells in the anterior chamber (hypopyon). Funduscopic evaluation revealed extra retinal fibro vascular proliferation inducing tractional retinal detachment in his right eye (Panel A) and arterial attenuation along with occlusion and perivascular sheathing along the inferotemporal vascular arcade in left eye (Panel B). Fundus fluorescein angiography was performed which also showed previous healed lesions of chorioretinitis in peripheral retina (Panel C). Pathergy test came out be positive. Serology testing showed HLA-B51 positive. There was past history of similar oral ulcers 3 times in same year.

He was diagnosed of having Behcet's disease according to International classification criteria of Behcet's disease (Table 1) [1]. He was advised vitreoretinal surgery in right eye, topical steroids, systemic corticosteroids and cyclophosphamide. Patient refused surgery. After 6 weeks, there was only perception of light in right eye and 20/40 VA in left eye. Skin lesions improved however (Panel F)

Results and Discussion
Behçet's disease (BD) is a chronic systemic inflammatory disorder at the crossroad between auto- immune and autoinflammatory syndromes [2]. The male to female ratio is 7:1 in symptomatic forms, but women predominate over men in studies were less symptomatic forms are systematically looked for and included [3]. Symptoms include mucocutaneous lesions, hallmark of BD. Oral ulcers are unique or multiple. The typical lesion is round with a sharp, erythematous and elevated border, surface is covered with a yellowish pseudo membrane. Genital aphthae occur in 60 to 65% of cases and are very suggesting of the diagnosis of BD. They are localized in men on the scrotum, less frequently on the penis or in the urethra and in women on the vulva and vagina where they can be disseminated and painful or totally indolent. They are morphologically similar to the oral ulcers but usually larger and deeper. The lesions leave scars in 50% of cases allowing retrospective diagnosis. The pathergy test, a 2mm papule induced 24 to 48 hours after a cutaneous prick, is one of the 4 criteria of classification, but its sensitivity is low with disposable needle and cutaneous disinfection [4]. Ocular involvement is frequent and severe, often bilateral, compromising rapidly the visual function [5, 6]. It includes anterior uveitis, cataract, glaucoma, posterior segment involvement with vasculitis, vitritis, retinitis, panuveitis, retinal edema, cystoid macular degeneration, venous or arterial occlusion, disc edema and retinal detachment. Fluorescein angiography show capillary

Dilatation, vascular obstruction and area of leak. With intensive therapy and careful care, the prognosis is good with only 2% of de novo severe visual loss at 6 years of follow-up [4]. The eye involvement may also be associated with neurological manifestations: cranial nerves palsy, optic neuropathy or papilledema with benign intracranial hypertension.

Vascular manifestations include venous thrombosis occurs in 30% of cases. Deep vein thrombosis can be seen especially on big vessels: iliofemoral, superior or inferior vena cava or on unusual localization such as dural sinus thrombosis (headache, papilledema, intracranial hypertension), hepatic veins (Budd-Chiari syndrome) or inferior vena cava with pulmonary aneurysms (Hughes- Stovin syndrome) [7]. Other manifestations includes arthritis and neurological involvement.

There are no diagnostic investigations for the disease the diagnosis relying solely on the clinical picture.(Table 1) Several studies have confirmed a strong association with HLA-B51 particularly in patients from Japan [8]. (B51 and DRw52), Mediterranean, and Middle Eastern countries[9] whereas this is not mirrored in patients from Northern Europe and England [10].

Treatment must be tailored to the organ involvement and degree of severity of the individual patient. Combinations of drugs in an attempt to control the various clinical manifestations. It includes Colchicine (1-2 mg/day), Azathioprine (2.5 mg/kg/day), Methotrexate (7.5 mg once a week orally), Tacrolimus, Cyclophosphamide (orally 2 mg/kg/day or intravenously 750 to 1g/m^2 every 4 weeks), Thalidomide etc. Alpha interferon (2a or 2b) is also efficient, especially in case of severe and/or resistant uveitis [11].

Behçet's disease runs a chronic course with unpredictable exacerbations and remissions whose frequency and severity diminish with time BD significantly increases morbidity and mortality. The leading causes of morbidity in BD are the uveitis with the potential threat of visual loss and neurologic involvement. Main causes of death included major vessel disease (mainly arterial aneurysm and Budd-Chiari syndrome) cancer and central nervous system involvement and sepsis.

Tables and Figures

International Classification Criteria of Behcet's Disease [1]

In the absence of other clinical explanations, patients must have:

1. Recurrent oral ulceration (aphthous or herpetiform) observed by the physician or patient recurring at least three times in a 12-month period;

And two of the following

2. Recurrent genital ulceration.
3. Eye lesions: anterior uveitis, posterior uveitis, cells in the vitreous by slit lamp examination or retinal vasculitis observed by an ophthalmologist.
4. Skin lesions: erythema nodosum, pseudo folliculitis, papulo pustular lesions or acneiform nodules in post adolescent patients not on corticosteroids.
5. Pathergy, read by a physician at 24–48 hours. (Sensibility 85% - Specificity 96%)

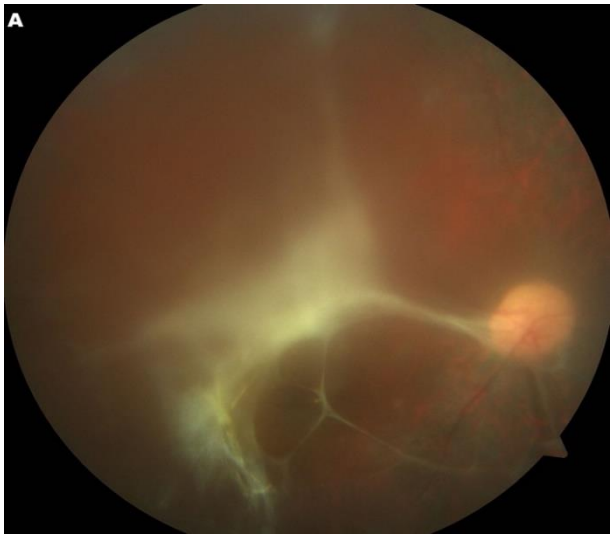

Fig A: Extra Retinal Fibro Vascular Proliferation (RT Eye)

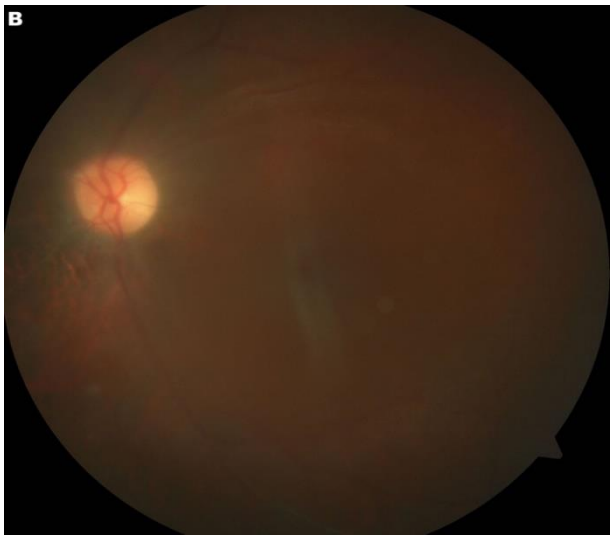

Fig B: Arterial Attenuation along with Occlusion and Perivascular sheathing (LT Eye)

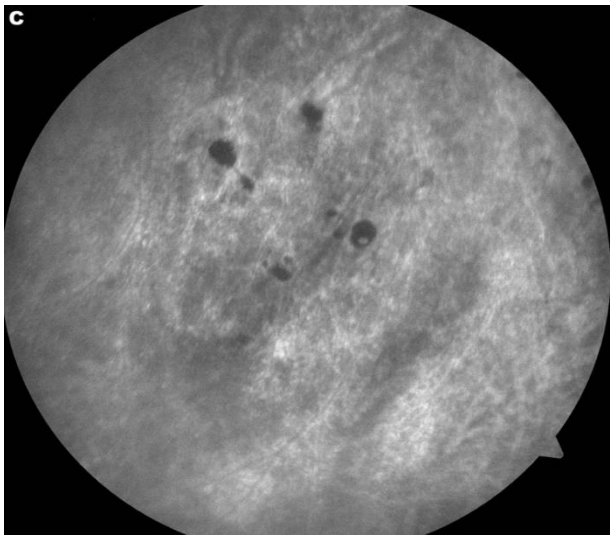

Fig C: Healed lesions of Chorioretinitis in peripheral Retina

Conclusions

Vision loss is a rare presentation in Behcet's disease if managed timely. Heightened physician awareness is important for early ophthalmological examination along with detection and prompt treatment to prevent irreversible visual loss.

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