Case Report: Behcet's Disease

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Case Report

Abstract: Reporting a young man aged 21 years diagnosed as Bechet's disease, who came with complaints of recurrent blurring of vision, painful buccal ulcers and skin lesions on both legs. Slit lamp examination revealed normal anterior segment, anterior vitreous haze. Fundus was showing vitritis, vasculitis. PositivePathergy test, presence of dermatographism and positive HLA-B51 confirmed the diagnosis.

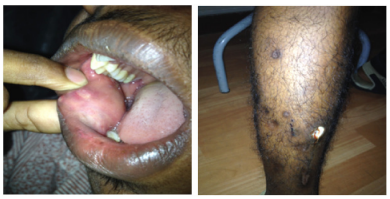
Keywords: Aphthous ulcer,Behcet'sdisease, dermatographism, HLA-B51, pathergy test, vitritis, vasculitis.

Introduction

Behcet's disease also called as behcet's syndrome,morbusbehcet's, adamantiades syndrome or silk road disease. It is named after a Turkish dermatologist Hulusi Behcet who recognized recurrent iridocyclitis with hypopyon, aphthous lesions in the mouth, and ulceration of the genitalia in two patients in 1937. It occurs more frequently in the Middle East and Japan, but in fact Behcet's disease is seen worldwide. Adamantiades Behcet's disease(ABD) is a chronic multisystem vasculitis characterized by mucocutaneous, articular, neurological, gastrointestinal and ophthalmological lesions. Ocular involvement is mainly represented by recurrent uveitis, especially posterior uveitis; however, iridocyclitis, retinal and choroidal vasculitis, optic neuritis and retinal vascular occlusion can also occur.

Case Report

A 21-year-old male patient presented with on and off blurring of vision in right eye during last 2 years. On examination, distant vision was 1 meter finger counting in right eye which was not improving by pin hole and 6/12 in left eye improving to 6/9 with pin hole, near vision was detected as N36 and N6 in right and left eye respectively. Slit lamp findings was normal anterior segment with quiet AC, no hypopion. Gonioscopy revealed grade Shaffer's 4 open angles. Intraocular pressure was normal in both eyes. Red colour saturation was decreased in right eye. On dilated fundus examination, right eye media was hazy due to grade 2 vitreous cells, left eye was normal. Ultrasound B-Scan showed attached retina in both eyes, with diffuse hyper echogenic pattern in the right eye suggesting of vitreous cells. On systemic evaluation of patient multiple nodular pyoderma gangrenosum ulcers noticed on ventral and dorsal surface of left leg. Also multiple, round, whitish aphthous ulcers in oral cavity were noticed, scrotal swelling was present, the provisional diagnosis of Behcet's was done and dermathological opinion was taken which confirmed the same. The patient's Pathergy test was positive (skin hypersensitivity to needle prick), dermatographism (firmly stroking the skin causes a urticarial wheal) was present. Laboratory investigations reported as ESR 50 mm/hr,HLA-B51 positive, suggesting a diagnosis of Behcet's disease.



Discussion

Behcet's disease is a multisystemic disorder commonly seen in young males. Nearly all patients present with some form of painful oral mucocutaneous ulcerations in the form of aphthous ulcers or non-scarring oral lesions. Painful genital ulcerations usually develop around the anus, vulva, or scrotum and cause scarring in 75% of the patients. Additionally, patients may present with erythema nodosum, cutaneous pustular vasculitis, and lesions similar to pyodermagangrenosum. Inflammatory eye disease can develop early in the disease course and lead to permanent vision loss in 20% of cases. Ocular involvement can be in the form of posterior uveitis, anterior uveitis, or retinal vasculitis. Anterior uveitis presents with painful eyes, conjunctival redness, hypopyon, and decreased visual acuity, while posterior uveitis presents with painless decreased visual acuity and visual field floaters. Arthralgia is seen in up to half of patients, and is usually a non-erosive poly or oligoarthritis primarily of the large joints of the lower extremities. The cause is not well-defined, but it is primarily characterized by auto-inflammation of the blood vessels. The primary mechanism of the damage is an overactive immune

system that seems to target the patient's own body. The involvement of a subset of T cells (Th17) seems to be important. A large number of serological studies show a linkage between the disease and HLA-B51. There are many criteria for diagnosis of behcet'sdisease, International Study Group Criteria for Behcet's disease is as follows:

Table 1: International Study Group Criteria for Behcet's Disease

Recurrent Oral Ulcerations	Minor aphthous, major aphtous or herpetiform ulceration observed by physician or patient, which recurred at least 3 times in one 12 month period .
Plus 2 of :	
Recurrent Genital Ulceration	Aphthous ulceration or scarring observed by physician or patient
Eye lesion	Anterior uveitis, posterior uveitis, or cells in vitreous on slit lamp examination or retinal vasculitis observed by ophthalmologist
Skin lesion	Erythema nodosum observed by physician or patient, pseudofolliculitis or papulopastular lesions, or acneform nodules observed by physician in postadolescent patients not on corticostreoid treatment
Positive pathergy test	Read by physician 24-48 hours.

Table 2: Diagnostic Criteria For Behçet's Disease (Proposed By The Behçet's Disease Research Committee of Japan)

Major Criteria	**Minor Criteria**
Recurrent Oral Aphthous Ulcers	Arthritis
Skin Lesions: **Erythema nodosum-like lesions **Folliculitis	Epididymitis
Genital Ulcers	Gastrointestinal Involvement
Ocular Disease: **Iridocyclitis with hypopyon **Posterior Uveitis with retinal vasculitis	Vascular Involevement: **Thrombophlebitis
	Neurologic Symptoms

Diagnosis

Complete	Presence of all major criteria
Incomplete	3 major criteria 2 major + 2 minor criteria ocular disease + 1 major criterion ocular disease + 2 minor criteria
Suspect	2 major criteria
Possible	1 major criterion

Conclusion

In the above said case though ocular manifestation was not severe, the patient was advised immunosuppressive and cytotoxic agent to avoid the ocular and other systemic complications. Behçet's disease is a chronic condition with remissions and exacerbations that "burn out" after approximately 10 years of activity. Ocular involvement is common and visual prognosis is poor if the patient is not treated properly. Behcet's disease is a rare condition in this part of the country. It is emphasized for a proper ocular and systemic evaluation and correct referral to other concerned specialities and regular follow up.

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