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Case Report

Multidisciplinary Approach in Management of Behcet's Disease

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Abstract

Behcet's disease (BD), also known as Silk Route disease, is a recurrent systemic inflammatory disorder characterized by oral aphthous ulcers, skin lesions, ocular lesions, and genital ulcerations. Despite multiple studies on the etiopathogenesis of the disease, its exact mechanisms remain undefined. Behçet's disease is most common in Turkey, the Mediterranean basin and Middle and Far East. Though symptoms usually begin to appear between the ages of 20 and 30, anyone can develop this disease at any age. It affects both men and women equally. Out of the 17 criteria proposed for the diagnosis of BD, the most commonly used criteria are the International Study Group (ISG) on BD criteria and the "International Criteria of BD" (ICBD) criteria. We will discuss a case of a 24-year-old male who presented with Fever, Papulonodular trunk lesions, Mucocutaneous lesion – aphthous ulcer, Genital ulcer – over glans, Ciliary conjunctival congestion. On evaluation the patient had acute non-granulomatous anterior uveitis, and along with that he fulfilled all the major criteria for Bechet's disease. HLA-B51 was also found positive. He was managed with a course of oral as well as topical steroid along with immunomodulator and supportive drugs and the patient recovered well.

Keywords: Behcet's disease; Human Leucocyte Antigen; Population

Abbreviations

BD: Behcets Disease; ISG: International Study Group; ICBD: International Criteria of Behcets Disease; HLA: Human Leucocyte Antigen; OPD: Out Patient Department; FUO: Fever of Unknown Origin; SLE: Systemic Lupus Erythematosis; ANA: Anti-Nuclear Antibody

Introduction

Behçet's disease is a rare inflammatory disorder characterized by the triad of recurrent aphthous ulcers, genital ulcers, and eye involvement [1,2]. It has also been described as a variable vessel vasculitis by the 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides [3]. Inflammation usually happens when your immune system is fighting an infection as a result of which your immune system attacks your blood vessels instead. The disease has the ability to involve blood vessels of nearly all sizes and types, ranging from small arteries to large ones, and involving veins too. This can cause problems in many parts of the body, including the eyes. This condition is chronic but has a waxing and waning curve.

An important risk factor for this disease is the presence of HLA-B51 gene. However, it must be emphasized that presence of the gene is not enough to cause Behcet's, that is, many people possess the gene, but relatively few develop the disease.

Even though the presence of HLA-B51 is defined as the predisposing factor for the disease, familial cases constitute only 5%. Therefore, the belief is that other factors play equally important roles, among which infections and environmental exposures require a larger degree of assessment.

The prevalence of BD is variable globally, with Turkey having the highest prevalence, with 370 cases/100,000 population [4]. It usually presents in the third to fourth decade of life, and it is unusual for the disease to occur during adolescence or after 40 years of age. In India, the mean age of presentation ranges from 23 years to 33 years [5-7]. Globally, the disease affects males predominantly; however, in India, the data are variable, with two studies showing male predominance, and one study demonstrated female preponderance [7].

About 3 out of 4 people with BD have symptoms that afflict their eyes. It may cause either anterior uveitis or posterior uveitis, and sometimes both. Anterior uveitis results in pain, blurring of vision, photophobia, tearing, or redness of the eye. Posterior uveitis is considered to be a greater threat to vision because while it damages the retina, it often presents with fewer symptoms. Severe eye disease that can cause blindness is more common in the Middle East and Japan than elsewhere.

Our case reported to the emergency department with complaints of fever and generalized rash. The rash was a mucocutaneous lesion with multiple aphthous ulcers over the tongue, soft palate, and post-pharyngeal wall along with papulonodular trunk lesions. The genital ulcer was over glans penis. Both eyes had acute non-granulomatous ant uveitis.

Materials and Methods

The patient was subjected to a detailed general examination by the physician. The patient was referred to Ophthalmology OPD for his complaints of redness, watering and mild blurring of vision in both eyes. His comprehensive eye examination included scrutinizing distant and near visual acuity in both eyes which were 6/9 and N8 (unaided) respectively.

Further his anterior segment evaluation under magnification using slit lamp biomicroscopy revealed ciliary congestion with few fine keratic precipitates along with minimal cells and flare. Posterior segment evaluation was carried out using an indirect ophthalmoscope with 20D Lens and over slit lamp biomicroscopy using 90D lens which was found to be clear.

Hence, a diagnosis of non-granulomatous anterior uveitis in both eyes was established and subsequently managed with a course of topical steroids and mydriatics. Because of the classical triad of Oro-genital ulcers, skin involvement, bilateral uveitis, a suspicion rose for BD.

He tested positive for human leukocyte antigen (HLA) B51; however, the pathergy test was negative. Hence, a diagnosis of BD was settled in accordance with the criteria proposed by International Criteria For Behcet's Disease and International Study Group Criteria.

The case was managed successfully with a course of oral steroids and colchicine along with topical steroid and mydriatic. His vision improved; redness reduced and other systemic as well as local complaints were resolved in due course of time and he went back fit and fine.

Results and Discussion

In 1930's, a Turkish dermatologist named Hulusi Behcet, observed a triad of oral ulcers, genital lesions, and recurrent eye inflammation, and became the first physician to describe the disease in modern times. But Dr. Hulusi Behcet was indeed not the first one to identify this condition. In the year 1922 Planner and Remenovsky had too described a case which was similar to Behcets disease. Finally in 1947, the medical world acknowledged Behcet's observations as a new disease entity, and Dr. Miescher propounded that the new disease be named "Morbus Behcet" [8].

Dr Hulusi Behçet accredited Dr. Adamantiades' work in his original publication, and Behçet's syndrome is sometimes referred to as the "Adamantiades–Behçet syndrome" [9]. Over these seven decades 17 sets of diagnostic criteria have been proposed [10] and out of these 17, the most commonly used criteria to diagnose BD are The Japan criteria (1972), The O'Duffy criteria (1974), The International Study Group (ISG) on BD criteria (1990/1992), The Dilsen criteria (1986), and the "International Criteria of BD" (ICBD) criteria (2006) [10].

Of all these myriad, ISG and ICBD are the most commonly used. Out of the two, ISG has better specificity, and ICBD has better sensitivity. ICBD utilize six norms: oral ulcers, genital ulcers, skin involvement, eye involvement, vascular manifestations, and neurological involvement. The presence of genital ulcers and eye lesions get two points each. The other four items get one point each. A positive pathergy test gets additional one point. A patient has to get four or more points to be diagnosed/classified as having Behcets. Our patient had oral ulcers (2 point), eye involvement (2 points), and skin involvement (2 point), with a total behcets score of six and hence diagnosed to have BD.

The most commonly used criteria do not include HLA-B51 positivity. Its positivity is associated with a relative inflation in the prevalence of genital ulcers, ocular or skin manifestations, and a 30% relative abatement in gastrointestinal tract involvement prevalence. HLA-B51 positivity is seen in 50%-72% of the BD patients [11]. Our patient also tested positive for HLA-B51 and had bilateral non-granulomatous anterior uveitis along with skin and genital manifestation.

A positive pathergy phenomenon is defined as the formation of erythematous nodule or pustule more than 2 mm in diameter on the skin following a prick by a sterile needle [2]. It is commonly associated with Behcets Disease, Sweet's syndrome, Inflammatory bowel disease, and spondyloarthropathies [12]. We performed pathergy test in our patient by pricking the skin (3 mm deep) over the right forearm's ventral aspect with the help of a 21G blunt hypodermic needle. It was negative at the end of 48 hrs. In India, the pathergy test was found positive in 14.8% of the patients in a study by Pande., *et al*. [5] and in 31% of the patients in a study by Singal., *et al*. [7] Pathergy phenomenon is part of the ICBD criteria, and a positive test is accorded one point. However, it is not mandatory for BD's diagnosis, as was the case with our patient.

Our patient initially presented as a case of fever with rash. In a study conducted by Mir., *et al*. infection was described as the most common cause of fever, followed by noninfectious inflammatory diseases and malignancies [13]. In our patient following the potentially diagnostic clues of oro-genital ulcers, and bilateral uveitis, we considered the diagnosis of BD. Few case reports have reported FUO as the initial presentation of BD as seen in our case too [14,15] other differentials were also kept into consideration in our patient like inflammatory bowel disease, SLE, reactive arthritis, and herpetic infections which can mimic BD and shall be ruled out first. SLE can have a very similar presentation to BD and can involve all the organs involved in BD in a similar fashion. However, inflammatory thrombi are not usually seen in SLE, and the SLE-specific autoantibodies can help differentiate these two conditions [14]. Our patient was also evaluated for SLE; however, it was ruled out by negative ANA test.

The most common skin manifestations in Behcet's are papulopustular lesions, erythema nodosum lesions, thrombophlebitis, and varied cutaneous and vasculitic lesions. The papulopustular lesions are clinically similar to lesions of acne vulgaris, and they are seen in patients with positive pathergy reaction and joint involvement. Our patient did not have a positive pathergy test or joint involvement. But he had papulopustular eruptions on trunk.

Ocular findings generally occur within the first 2–4 years of the disease. In 80% of the patients, the manifestations are bilateral [15]. Anterior uveitis is usually the only initial ocular manifestation in patients with Behcets and it can occur as an isolated finding in about 10% of the patients. Ocular Behcets may presents as iridocyclitis with or without hypopyon, vitritis, retinitis, occlusive retinal vasculitis, and cystoid macular edema (CME). Band keratopathy, glaucoma, vitreoretinal hemorrhage, posterior vitreous detachment, macular degeneration, epiretinal membrane, vein occlusion, and phthisis of the eye may also be observed as complications of ocular Behcets [16]. Although ocular Behcets is characterized by an explosive acute hypopyon uveitis, a more common presentation is iridocyclitis without hypopyon, which is seen in two-thirds of the cases [17-19]. Our patient too presented with bilateral acute anterior non-granulomatous uveitis without hypopyon and without any posterior segment involvement.

Conclusion

By far we have gathered enough knowledge about Behcets disease in terms of diagnosing and treating the disease. The knowledge propounds that a multidisciplinary approach should be followed to manage the disease. From the ophthalmologist point of view, I would like to evince that no red eye should be trivialized, and treatment be erred rather one should try to excavate the cause as many a times it is embarkation of an ongoing masquerading systemic malady.

Conflict of Interest

There is no financial interest, or any conflict of interest exists.

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