

BEHCET DISEASE- A CLINICAL ENTITY REQUIRING A VERY HIGH GRADE OF CLINICAL SUSPICION

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ABSTRACT

INTRODUCTION

Behcet disease is an auto-inflammatory systemic vasculitis. It usually comprises a triad of recurrent oral and genital ulcerations, ocular manifestations.

CASE REPORT

A18 years old lady presented with recurrent oral ulceration since 1 year which was causing difficulty in chewing food and burning sensation in oral cavity. Out of high grade of clinical suspicion, due to the recurrent nature of ulcers and rapid resolution by steroids, HLA profiling was done, and HLA B5 came out to be positive and the patient was diagnosed as Behcet disease.

CONCLUSION

Behcet disease has a multi-systemic involvement. The diagnosis of Behcet disease requires a very high threshold of clinical suspicion to appropriately manage the disease.

KEY WORDS

Behcet Disease, HLA B5, Aphthous ulcer, Oral steroids

INTRODUCTION

Behcet's disease was described by Hulusi Behcet in 1937 as an inflammatory process of unknown etiology, characterized by recurrent aphthous ulcers, genital ulcers, uveitis and cutaneous

lesions. It is also associated with other less frequent systemic manifestations, such as gastrointestinal, central nervous system, vascular and joint infections¹. BD is mainly sporadic. The aetiology is suspected to be combination of genetic and environmental factors. HLA-B51 genetic marker is found in around 60% of BD patients.² The demonstration of an autoimmune genesis is given by the presence of antimucous autoantibodies, together with the association of the disease with the HLA configurations B5 and B51. The evolutionary history of the disease is marked initially by oral, followed by genital ulcers, ocular lesions skin disorders and arthritis³.

Skin lesions are present in forms of erythematous nodules, erythematous papules, vesicles, pustules, pyoderma, folliculitis and acneiform eruptions and are positive in the pathergy test, that is present in 40 and 80% of affected patients.² Posterior uveitis, retinal vasculitis, conjunctivitis, optic neuritis and retinal arthritis are frequent ocular manifestations, but anterior uveitis with hypopyon is the classical manifestation in the beginning of the Behcet's syndrome. The articular manifestations are present in over 50% of the patients and may precede, accompany or follow other manifestations of the Behcet's syndrome.

Neurological manifestations are rare due to vascular involvement at the location or because of peripheral polyneuropathy. The thrombosis of

small cerebral vessels, or large venous sinus manifested by endocranial hypertension.⁴ Vascular manifestations are presented as superficial thrombophlebitis, venous thromboses or as arteritis. These arteritis are manifested by occlusive-thrombotic and aneurismatic phenomena. Thus, they are responsible for heart attack or hemorrhagic phenomena in different organs.⁵ Gastrointestinal manifestations preferentially attack the intestines and esophagus, and are manifested through abdominal pain, diarrhea and occasionally by perforations. The main differential diagnosis of Behcet's syndrome are erythema multiforme or Stevens Johnson Syndrome and Crohn's disease, showing lesions in oral and genital mucosa. Sarcoidosis can present with erythema nodosum, uveitis, and arthralgia.⁶ The main objective of Behcet syndrome patient care is to treat in time the oral mucocutaneous lesions in order to hinder the progression of the disease and to prevent the irreversible organ involvement in particular during the active phase.⁷

International Study Group Criteria (1990)⁸

Criteria Features

- Recurrent oral ulcerations Minor/ major aphthous or herpetiform ulceration which recurred at least three times in 12-month period.
- Plus any two of the following:
- Recurrent genital ulceration Aphthous ulceration or scarring
- Ocular lesions Anterior/ posterior uveitis or vitreous cells or retinal vasculitis
- Dermatological manifestations Erythema nodosum, pseudofolliculitis or papulopustular lesions or acneiform nodules
- Positive pathergy test

CASE REPORT

We present a case report of a 18 years old lady, who was referred to our Institute, for the management of recurrent ulcers in the oral cavity. She was having oral ulceration since 1 year

duration. She also had difficulty while chewing food and burning sensation in oral cavity. Initially, she went to the local doctors where she was treated with topical mucosal analgesic ointments and oral multivitamins. Patient got no relief with these medications. She was given topical steroids with that she got temporarily relieved but then lesions eventually recurred. then she was referred to our institute in the Otolaryngology clinic, where a thorough clinical history and local and systemic examination were performed. There was no other systemic complaints. Bladder and bowel habits were normal. Patient's only major concern was recurrent multiple oral ulcers, which according to her was getting relieved with the application of topical steroids.

On clinical intraoral examination, multiple ulcers were observed on the edge and dorsum surface of the tongue, lips and bilateral buccal mucosa. It was measuring approximately 0.5–1.2 cm in diameter, with the presence of an erythematous halo and whitish margin. Dental scaling was



present.

Figure 1: Oral examination of patients shows multiple aphthous ulcer over the labial mucosa and dental scaling of the lower incisors and canine teeth

Considering the recurrent nature of the illness and resolution of symptoms by aid of steroids and on taking a detailed history we found out she had a past history of genital ulceration which got relieved eventually. These findings corroborative of the autoimmune nature of disease.



Figure 2: Multiple aphthous ulcer seen on the dorsum of tongue along with depapillated regions.

All the routine blood investigations were performed. And out of suspicion of Autoimmune disease, ANA, C-RP and HLA profile were performed. Her C-RP was elevated and HLA B5 came out to be positive. Diagnosis of Behcet Disease was made and accordingly, the treatment was prescribed with topical Triamcinolone 0.1% as well as oral steroidsie. Methyl Prednisolone in the dosage of 1mg/day/kg body weight, given in divided dosage. She responded drastically well and the nature of disease of patient was explained which helped to alleviate her anxiety.

Clinically, there was significant improvement in the symptoms and signs of disease.

DISCUSSION

Behcet disease is a multisystemic disease of unknown etiology. It is usually characterized by triad of oral ulcers, genital ulcers, and eye inflammation⁹. In our case, the patient presents with predominantly oral ulcers and genital ulcer. It commonly occurs in third decade of life. Men are more frequently affected than women¹⁰. In our case study, patient is 18 years female.

Oral and genital ulcers are the hallmarks of the disease, seen in up to 97% and 60–90% of patients, respectively in the study by Krause et al. The oral manifestation includes benign recurrent aphthous stomatitis. These typical aphthous ulcers generally vary in size, measuring from 2 to 10 mm in diameter, and present with a yellowish necrotic base, raised edges and a diffuse erythematous area¹¹. Ocular lesions are present

in 30–70% of the cases¹². They can manifest as photophobia, followed by uveitis and conjunctivitis.

The clinical details encountered in our patient are recurrent aphthous oral ulcers and genital ulcers. There was no ocular complaints by the patient.

The management of this condition involves a multi-disciplinary approach as Behcet Disease has varied clinical manifestations. The diagnosis of Behcet disease can be made clinically and can be difficult due to the lack of any pathognomic laboratory finding. When the patient has multi-systemic involvement then imaging studies can be beneficial which includes X-rays and arthrocentesis to assess arthritis, CT-scan to assess for bleeding, thrombosis, and ischemia, angiography to look for aneurysms, which can be a fatal complication.

The main objective of patient having Behcet syndrome is to treat the oral mucocutaneous lesions effectively to avoid the progression of the disease and also to prevent the irreversible organ involvement in particular during the active phase¹³. The treatment strategy is decided based on organ involvement, severity, and prognostic factors.

Currently treatment varies according to severity of manifestations, starting with simply informing, explaining and reassuring the patient. For oral manifestations in patients which present complex aphthous ulcers, patient can be started with topical colchicine and dapsone. In more severe cases, a combination of corticosteroids with immunosuppressors, such as cyclosporin, azathioprine, cyclophosphamide, interferon-alfa 2a or chlorambucil, can be used; however, the need for multidisciplinary treatment is required because this is a systemic infirmity. In our case report, patient got a tremendous response to oral as well as topical steroids.

CONCLUSION

In Behcet Disease, a multi-disciplinary approach is required for patient's management. The diagnosis of Behcet Disease requires a very high threshold of clinical suspicion to appropriately manage the disease.

In case of patient with history of recurrent oral ulceration, a physician should always refer patient to an otorhinolaryngologist for diagnosis and further management.

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