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A CASE STUDY ON BEHCET's DISEASE ASSOCIATED WITH CYCLOPHOSPHAMIDE-INDUCED CARDIAC FAILURE

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ABSTRACT

Behçet's disease (BD) is a rare, chronic inflammatory disorder characterized by recurrent oral and genital ulcers, skin lesions, and eye manifestations. This condition primarily affects young adults. Its exact cause remains unknown, though it is believed to be caused due to autoimmune and genetic factors. This case study presents a 32-year-old female admitted with fever, headaches, recurrent skin and oral ulcers, and eye complications including pseudophakia in the left eye and hemorrhage and detachment in the right. Additionally, she suffered from arthritis and abdominal aortitis. Blood tests showed positive pathergy and negative HLA-B51 antigen. She was diagnosed with Behcet's and treated with Adalimumab for 6 months, resulting in improved eye inflammation, alongside cyclophosphamide, steroids, and Rituximab. Behcet's disease presents diagnostic challenges due to its various symptoms, often leading to delays in diagnosis and treatment. Management focuses on symptom relief and prevention of complications.

Keywords: - Behçet's disease, Pseudophakia, Human Leukocyte -B51 Antigen, Uveitis.

INTRODUCTION

Behçet's disease (BD) is a long-term immune-mediated illness that can impact several organ systems. BD is distinguished by recurring oral and vaginal ulcers and often progresses in a relapsing-remitting pattern.

Although the exact cause of BD is still unknown, it is a complex chronic illness that affects people with a genetic predisposition such as Human Leukocyte Antigen (HLA-B51) that is brought on by environmental, microbiologic, and immunologic factors.

Patients who tested positive for HLA-B51 had increased rates of thrombophlebitis, oral ulceration, vaginal ulceration, and a family history of BD. Patients with HLA-B51 positivity had lower rates of erythema nodosum, papulopustular eruption, pathergy positivity, arthritis, and ocular involvement [(1)].

Disease criteria include three mouth ulcers within a year, as well as two of the following: reoccurring vaginal ulcers, ocular irritation, distinctive skin lesions, or a positive pathergy test. Inducing and maintaining remission, enhancing the quality of life, avoiding permanent damage, and preventing the disease from getting worse are the primary goals of treatment for patients with BD. Immunosuppressants including cytokine inhibitors and anti-tumor necrosis factor drugs are used in its symptomatic treatment. Oral corticosteroids are used to treat ocular symptoms [(2)].

Colchicine is used to treat mucocutaneous lesions that reoccur or to stop them from occurring in their initial stages. Anticoagulation therapy is a possible treatment for vascular involvement. The phosphodiesterase inhibitor apremilast has been used to treat oral ulcers [(3)].

Here, we describe the clinical findings of a patient with Behcet's disease with HLA B51 negative condition.

CASE STUDY

This case study features a 32-year-old female who was admitted to a tertiary care hospital with complaints of fever and intermittent headaches. She also had recurring skin and oral ulcers. The medical history revealed that she was diagnosed with Dyslipidaemia, Hypertension, Type -2 Diabetes Mellitus. After a thorough analysis of patient history, it was found that the oral and skin ulcers has persistently occurring to the patient since the past 1 year, also the patient had pseudophakia done in the left eye and vitreous hemorrhage and sub hyaloid hemorrhage with tractional retinal detachment at the macula in the right eye. These all symptoms lead to the provisional diagnosis of Behçet's disease. Later, the patient experienced progressive arthritis and abdominal aortitis. Blood tests revealed a positive pathergy test and a negative HLA-B51 antigen. Based on the symptoms and clinical manifestations, the patient was then diagnosed with Bechet's Disease. The pharmacotherapy initiated were, Inj. Adalimumab 11 doses given for a period of 6 months. Other medications were Inj. Cyclophosphamide 1 gm once weekly and steroids resulted in the clearing of the eye inflammatory changes and also started on Inj Rituximab 1gm were given for treatment. During the course of treatment wuth cyclophosphamide, the patient developed cardiac failure. Lab reports revealed the BNP level was found to be

12100 pg/ml. 2D- echo revealed the patient had Heart Failure with Preserved Ejection Fraction (HFpEF) due to the use of cyclophosphamide. Hence, cyclophosphamide was stopped. The symptoms of the patients improved on treatment and was discharged.

DISCUSSION

This case study describes Behcet's disease was diagnosed in a 32-year-old female patient. Behcet's disease (BD) is an enduring vasculitis characterized by recurrent episodes of unknown origin. Its impact spans across multiple organ systems due to its ability to affect arteries and veins of various sizes, leading to considerable morbidity and mortality. Commonly referred to as the 'Silk Road' disease, it is prevalent worldwide. Clinical manifestations encompass oral and genital ulcers, uveitis, and involvement of vascular, neurological, joint, renal, and gastrointestinal systems[(4)].

Behcet's disease (BD), also known as Adamantiades-Behcet, manifests as a systemic inflammatory condition marked by recurrent oral aphthae, genital ulcers, cutaneous symptoms, and uveitis. While the precise cause of BD remains elusive, both genetic and environmental factors are believed to contribute to its development. BD is often categorized as a Th1-type autoimmune disorder due to its association with HLA-B51 and heightened reactivity to streptococcal antigens. The prevalence of HLA B5 is notably high among North Indians. Despite this, reports of BD from India are limited, with only two significant studies from North India: one conducted by the rheumatology department, documenting 58 cases over 16 years, and another recent retrospective study from a uveitis clinic, documenting 53 cases over 17 years.

The prevalence of Behcet's disease varies across different populations, with rates ranging from 80 to 370 cases per 100,000 individuals in Turkey, 10 cases per 100,000 in Japan, and 0.6 cases per 100,000 in Yorkshire. While European cases are commonly reported, they are not limited to the migrant population. Behcet's disease shares MHC class I associations with conditions like ankylosing spondylitis and psoriatic arthropathy. HLA-B51 stands out as the most strongly associated genetic factor with BD. However, it only explains a fraction of the genetic risk, even in familial cases, suggesting the existence of other yet undiscovered genetic factors[(5)].

In Behcet's disease, there is no specific biological test available for diagnosis. Instead, international classification criteria have been established with a sensitivity of 85% and specificity of 96%. While markers like erythrocyte sedimentation rate, CRP, and other acute phase reactants are often not elevated during acute phases or relapses, they do not reliably correlate with disease activity. A cutaneous biopsy, typically performed via intradermal injection with physiological saline solution, may reveal vasculitis with immune complex deposition.

Treatment primarily involves symptom management through the use of steroids and immunomodulatory therapy, with efficacy contingent upon prompt initiation, patient compliance, and treatment duration. Immunomodulatory drugs have demonstrated effectiveness but are typically prescribed alongside corticosteroids initially due to their delayed onset of action. While Ciclosporin has shown efficacy in treating uveitis, its prescription may be limited by the risk of secondary nephropathy. Recent studies have reported the efficacy of infliximab in severe cases of BD uveitis. Additionally, anecdotal reports suggest some efficacy of

Disulone, Sucralfate, and Pentoxifylline. Patient education, adherence to treatment, regular follow-up, and timely therapeutic intervention contribute to improved prognosis, similar to other chronic diseases. BD significantly increases morbidity and mortality, primarily due to uveitis with the potential for visual loss and neurologic involvement[6].

CONCLUSION

Since the involvement of key organs is so common, appropriate care and routine follow-up are necessary for Behçet's disease. Its multisystemic nature necessitates collaboration with pertinent specialties, including internal medicine, ophthalmology, dermatology, neurology, dentistry, and rheumatology, to improve patient outcomes. Although a permanent cure has not yet been found, patients must continue with their treatment plan and be fully educated about the condition's relapsing and remitting nature.

REFERENCES

1. Erdem Sultanoğlu T, Eröz R, Ataoğlu S. Evaluation of HLA-B51 frequency and its relationship with clinical findings in patients with Behçet's disease: 4-year analysis in a single center. Egypt Rheumatol Rehabil. 2023;50(1):15. doi:10.1186/s43166-023-00181-1.

2. Hatemi G, Seyahi E, Fresko I, Talarico R, Uçar D, Hamuryudan V. One year in review 2021: Behçet's syndrome. Clinical and Experimental Rheumatology. 2021;39(5):3-13. doi:10.55563/clinexprheumatol/lnvc9k.

3. Mahoney JM, Millonig K. Behcet's Disease. Journal of the American Podiatric Medical Association. 2017;107(5):454-456. doi:10.7547/16-087.

4. Emmi G, Bettiol A, Hatemi G, Prisco D. Behçet's syndrome. The Lancet. 2024;403(10431):1093-1108. doi:10.1016/S0140-6736(23)02629-6.

5. Singal A, Chhabra N, Pandhi D, Rohatgi J. Behcet′s disease in India: A dermatological perspective. Indian J Dermatol Venereol Leprol. 2013;79(2):199. doi:10.4103/0378-6323.107636.

6. Alibaz-Oner F, Direskeneli H. Update on the Diagnosis of Behçet's Disease. Diagnostics. 2022;13(1):41. doi:10.3390/diagnostics13010041.