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Case Study

Behçet Diseases: A Puzzling Case of Recurrent Aphthous Ulcers – A Comprehensive Report

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I N F O

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A B S T R A C T

Behçet's illness is an unidentified multisystem autoimmune ailment that frequently causes oral and genital ulcers and ophthalmic and cutaneous damage as well as indications for arthritis and may also affect the gastrointestinal tract and the central nervous system. In this research, we focus on the clinical characteristics and management of a case with recurrent aphthous ulcer having Behcet syndrome.

Keywords: Bahcet, Recurrent Aphthous Ulcers (RAUs), Recurrent Aphthous Stomatitis (RAS), Genital Ulcers

Introduction

Hulusi Behçet (1889-1948), a Turkish dermatologist, initially identified Behçet disease (BD) in 1937 as "recurrent oral aphthous ulcers as well as genital ulcers and 'hypopyon-uveitis".[1] Behcet's disease is a systemic autoinflammatory vasculitis with no recognized cause. It is distinguished by mucocutaneous signs, such as recurrent oral and vaginal ulcerations, ocular manifestations, particularly recurrent regressive uveitis, and systemic vasculitis affecting arteries and veins of all sizes. It is sometimes referred to as malignant aphthosis and Behcet's syndrome.[2]

Recurrent aphthous ulcers (RAUs) are idiopathic intraoral ulcerative diseases that appear in otherwise healthy people. An alternative term for RAU is recurrent aphthous stomatitis (RAS). Simple aphthosis and complicated aphthosis are the two subtypes of RAS. In the absence of BD, complex aphthosis is characterised as the presence of three or more oral or genital aphthae that are nearly always present. The most prevalent and frequently the first clinical signs of BD are oral and vaginal ulcers.[4]

Therefore, it is crucial to take BD into account while making a differential diagnosis and monitoring individuals who have recurring aphthous ulcers.

Case Report

A 21-year-old man from Rajkot, Gujarat, presented with fever with chills and joint pain for 2 days, and he has recently experienced two bouts of recurrent aphthous ulcers. He was on a mixed diet and had no significant family history. The past medical history reported was rheumatoid arthritis since for 1 year with an increased RA factor – 26 IU/mL as well as positive CRP test – 35 mg/dL. The patient was treated with Tab. Methotrexate – 7.5 mg twice a week with NSAIDs. He has no social history. On examination he was conscious and oriented.

Diagnosis

The presence of Bahcet's diseases is confirmed by a positive skin pathergy test (SPT) with an increased Erythrocyte Sedimentation Rate (ESR) – 18.5 mm/h.

USG Abdomen suggests mildly enlarged liver with size of 16.2 cm, and Gall bladders distended with no ascites. His serology report shows high CRP levels (>0.6 mg/dl) and the ECG shows sinus tachycardia.

Treatment

During hospitalisation he was treated with Inj. Ceftriaxone – 1.5 gm twice a day, Inj. Diclofenac 75 mg IM STAT, Inj. Paracetamol 1 gm IV 8 hourly, Tab. Prednisolone 2.5 mg 1-0-1, Tab. Azathioprine 50 mg 1-0-1 with Tab. Colchicine 0.6 mg 1-1-1 for two weeks throughout the first stage of the therapy.

Discussion

Behçet's disease (BD) is an uncommon form of systemic vasculitis that manifests as eye lesions, genital ulcers, oral aphthous ulcers, and other generalized symptoms. The historic route for trade called the "Silk Road" connects eastern Asia with the Mediterranean region, and it is along this path that Eurasian people are most likely to develop BD. The origins of BD are unknown; however, it is thought to be brought on by an autoimmune reaction in those with a genetic predisposition that is brought on via an infection or environmental factor. The greatest risk factor for BD in regions throughout the Old Silk Route has been the MHC locus, or HLA-B51 allele, on chromosome 6p. Streptococcus and herpes simplex virus-1 have been proposed as potential environmental causes of BD.[5] The core of BD pathogenesis is currently thought to be T cell homeostasis disturbance, particularly Th1 and Th17 growth and impaired control by Tregs. The histology reveals vasculitis that affects all sizes of vessels, including arteries and veins. The systemic vasculitis known as BD features fibrinoid necrosis, endothelial cell oedema, and substantial neutrophil infiltration. Only clinical criteria can support the diagnosis of BD, hence other diseases must be ruled out according to clinical presentation. No pathognomonic laboratory evidence of BD has been found. Blindness and systemic involvement can both be deadly consequences of this uncommon illness. Major vascular disease and involvement of the central nervous system are the main causes of mortality.[6]

In comparison to female patients, male individuals with Behçet's illness have a larger hereditary risk. The risk inside the HLA region is substantially responsible for this genetic difference, which is predominantly obtained from our Turkish cohort. These findings imply that the illness presentation of men and women having Behçet's disease may differ due to hereditary reasons.[7]

It might be challenging to make a Behçet's syndrome diagnosis. The sets of categorization criteria are useful for research, but a clinical judgement still has to be made when making a diagnosis. The ISG criteria need the presence of oral ulceration in order to make a diagnosis.[8]

A quicker, more precise diagnosis & well-integrated treatment plans are made possible by managing BD. The mainstay of treatment is corticosteroids. Other medications that are utilized for induction and/or maintenance therapy include colchicine, AZA, ciclosporin-A, cyclophosphamide, IFN alpha, & tumour necrosis factor alpha inhibitors. Despite the growing usage of biologic medicines, however are still gaps in supply. Studies comparing some therapy approaches head-to-head are necessary (for instance, TNF inhibitors vs. IFN alpha in uveitis). Future pattern of care for BS may be altered by novel therapeutics under development.[9]

Conclusion

Bachet Diseases (BD) is a relapsing, chronic vasculitis that has the potential to have a severe impact on every bodily system and result in significant morbidity and death. Various signs and a lack of reliable biomarkers cause a delay in diagnosis. Numerous biomarkers have been developed, but none have been shown to be useful or trustworthy for use in clinical practice for illness diagnosis, monitoring, or activity evaluation.

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