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BEHÇET'S DISEASE

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Abstract:

Behçet's disease is a rare autoimmune disease that has multiple vague manifestations that affect multiple organ systems, such as the oral cavity, genitalia, eyes, nervous system, etc. Despite its widespread lesions, the disease is poorly understood and requires more extensive research about Behçet's.

Aim of the study: *to establish a better understanding of the symptoms, diagnostic methodology, and management of Behçet's disease.*

Materials and methods: *This review is a comprehensive search of PUBMED from the year 2015 to 2025.*

Conclusion: *Behçet's disease is a disorder that causes widespread, often vague symptoms that affect multiple organ systems of the body. A better understanding of the disease helps to reduce the recurrences along with the physical, psychological, and social well-being of the patient. Regular follow-up is essential to prevent any long-term complications of Behçet's.*

Keywords *Behçet's disease; autoimmune; aphthous ulcers; uveitis*

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INTRODUCTION:

Also known as Adamantiades-Behçet's syndrome, BD or Behçet's syndrome is an inflammatory that affects multiple organs, commonly including the mouth, genitals, skin, and eyes. Symptoms range from canker ulcers in the mouth, genital ulcers, and uveitis. Additionally, it can affect joints, blood vessels, the nervous system, and/ or the digestive tract.[1]

Studies report no specific gender predilection of Behçet's disease (BD), but the severe form of disease is seen more in males. The disease manifests commonly in the third decade of life and usually declines with age. BD can be seen in children, but not before the age of five years.[2] Early onset of the disease generally correlates with more severe manifestations and higher mortality.[3]

Behçet's disease is a rare disease in the USA and Western Europe.[1] It is more often seen in the eastern Mediterranean, the Middle East, and East Asia- along the ancient Silk Road, thus given the moniker of 'Silk Road Disease'. It is most prevalent in Turkey, with about 400 cases for every 100,000 individuals.[3]

The aetiology of BD is unknown but it is regarded as a genetic disorder wherein the appearance of lesions is triggered by external triggers, leading to an inflammatory response.[4] BD is seen commonly in families, but is otherwise seen sporadically. The most found genetic marker is HLA-B51, found in 60% of the patients. HLA-B15 and HLA-ERAP1 (endoplasmic reticulum aminopeptidases) are considered as susceptibility genes by the Genome-wide association studies (GWAS). Microbial exposure and cellular and humoral immunity leading to an inflammatory response play a role in susceptible individuals. These, along with responsiveness to immunosuppressant medication, contribute to the autoinflammatory-autoimmune nature of BD. [5] Pro-inflammatory cytokines are seen in surplus due to a dysregulated adaptive immune system, along with a disturbed Th1/Th2 balance, and a decrease in regulatory T cells.[4]

Signs and Symptoms

Behçet's is characterised by lesions that tend to disappear and spontaneously recur.[1] The disease is characterised by a triple symptom complex consisting of oral aphthous ulcers, genital sores, and ocular lesions. Acute inflammation manifests clinically as self-limiting, but relapsing episodes that have variable intensity that may produce a lasting sequelae. These attacks are unpredictable in frequency, duration, and severity, without distinguishable patterns.[3]

The most observed symptom is oral aphthosis, which is seen in about 90% of the patients.[6] The sores appear round to oval with erythematous borders, which can occur at any site in the mouth. The lesions can vary in depth, from shallow to deep, or in number, single or multiple lesions. The sores heal without scarring and typically last a few days-up to a week or more, but recur often. These lesions may precede other symptoms of BD by a few years.[1]

Similar lesions are seen on the genitalia of about 75% of the patients with BD. [3] The sores appear specifically on the scrotum and shaft of the penis in males and the vulva in females. Compared to mouth sores, the sores are round and painful, but can be larger and deeper in nature, and additionally, they tend to scar.[1] These genital ulcers do not affect fertility, but recurrences are painful and can limit the capacity for sexual intercourse. [5]

Ophthalmological manifestation is usually in the form of uveitis- posterior uveitis or anterior uveitis/iridocyclitis. Other inflammatory features include inflammation of the iris with pain, lacrimation, and the accumulation of pus (hypopyon iritis) can occur. Inflammation of the retina results in blurred vision, photophobia, and/or chorioretinitis.[1] It affects more commonly in younger individuals, more often in males than in females.[3] Estimated visual loss is approximately 10-20% at 5 years and is more common in younger males.[5]

Cutaneous lesions manifest as papulonodular lesions, erythema nodosum-like lesions, acneiform rashes, pseudofolliculitis, pyoderma gangenosum, and occasionally erythema multiforme-like rashes.[5] The nodules regress spontaneously, sometimes leaving faint scars or pigmentation. In certain individuals, cutaneous lesions might appear as acne-like eruptions or resemble hair follicles-pseudofolliculitis.[1]

Arthralgia is commonly observed in BD. It is indistinguishable compared to other forms of inflammatory arthritis, but is non-erosive and non-deforming. Sometimes a predominant feature can be enthesial inflammation, with fibromyalgia being the common cause of pain.[5] The pain can range from mild to severe, with affected joints including knees, wrists, elbows, and ankles. Long-term damage to the affected joints is considered rare.[1]

About 10-20% Behçet's patients involve the central nervous system (CNS)- known as Neuro-Behçet's.[1] The regions affected are the brain stem, basal ganglia, thalamus, cerebellum, and internal capsule. The neurological symptoms follow five years after the onset of non-neurological symptoms of BD. It is characterised as motor dysfunction, cranial nerve

palsy, and cognitive impairment. [7] Recurring inflammatory attacks can result in neurological damage.[1]

Vasculitis in BD commonly affects the venous vascular system vis-à-vis to the arterial vascular system. Arterial lesions cause aneurysms, ulcerations, thrombosis, and stenosis. Venous lesions are characterised by venous thrombosis and thrombophlebitis; these thrombi rarely embolise and remain adherent to the vessels.[5] In rare cases, these thrombi can travel to the lungs- pulmonary emboli, causing chest pain, coughing, difficult or dyspnea, and hemoptysis.[1]

Diagnosis

Like other rheumatic diseases, BD has no characteristic tests that can be utilised to diagnose the disease. No elevation of ESR and CRP is seen in every attack. Other criteria such as mean platelet volume (MPV), the neutrophil to lymphocyte ratio (NLR), and platelet to lymphocyte ratio (PLR) are utilised to measure the inflammatory states in BD. [6] The International Criteria for Behçet's disease, given in 2006, allot points to each symptom, with three points or more indicating Behçet's.[5]

Table 1: International Criteria for Behçet's disease (2006) [5]

SYMPTOMS	SCORE
Genital aphthous	Two points
Ocular lesions	Two points
Oral aphthous	One point
Skin lesions	One point
Vascular lesions	One point
Pathergy	One point

Thus, clinical diagnosis is essential for BD with the exclusion of differential diagnoses. The various investigations that can be employed include: **Routine investigations-** full blood count, liver and kidney profile, inflammatory markers, urine analysis, chest x-rays, coeliac screen, stool sample, autoimmune screen, and mouth and genital swab and culture and **specific investigations** – biopsies, doppler studies, CT scan, MRI scan, immunofluorescence, electrocardiogram, echocardiogram, endoscopy, and cultures.[5]

Pathergy tests are used for the diagnosis of mucocutaneous lesions, which is based on a hypersensitivity reaction after a needle puncture leading to the development of erythematous papules or pustules around the injection site 24-48 hours after the intradermal puncture, which is considered a positive test. Approximately 50% of the patients in the Middle and Near East show a positive pathergy test.[8]

Differential Diagnosis

It is essential to consider the various differential diagnoses before establishing a diagnosis for Behçet's disease. The various conditions that must be considered are nutritional deficiencies, such as B vitamins, iron, zinc, folate, and Stevens-Johnson syndrome, which affect the mucous membrane. [2] Sweet syndrome is a skin disorder that is characterised by red eruptions on arms, legs, neck, and face, which are painful, along with a general sense of malaise. Reactive arthritis is a similar condition that includes arthritis, inflammation of the urinary tract, and conjunctivitis. [1] Oral and genital ulcers are also seen in MAGIC (mouth and genital ulcers with inflamed cartilage) syndrome, cyclical neutropenia, or as an adverse drug effect.[5]

Management

As BD causes multiple attacks and progresses through multiple remissions. The treatment is symptom-specific. The mucocutaneous manifestations are amenable to local treatment and do not necessitate aggressive therapy. Arthritis treatment is dependent on attacks and remissions, and NSAIDs and colchicine are commonly employed.[6][1] For more aggressive and refractory cases, azathioprine and anti-TNF agents can be employed. As corticosteroids do not prevent recurrences, immunosuppressive agents are employed.[1]

Oral and genital sores can be treated using topical application of corticosteroid preparations. Local anaesthetics such as lidocaine and diphenhydramine can be used to relieve local pain. Apremilast is an FDA-approved drug for Behçet's patients with recurrent oral ulcerations.[1] Maintenance of oral hygiene and avoidance of sodium lauryl sulphate-containing toothpaste is essential.[5]

For ophthalmological manifestations, treatment involves corticosteroid eye drops to relieve pain and a combination with immunosuppressant agents for aggressive or refractory cases.[1] Cutaneous lesions require local application of steroids and oral antibiotics- lymecycline. Erythematous nodosum-like lesions respond to colchicine and corticosteroids.[5] For neuro-Behçet's, high-dose corticosteroids, cyclophosphamide, and TNF-alpha inhibitors are employed. [7]

CONCLUSION:

Behçet's disease is a disorder that causes wide-spread, often vague symptoms that affect multiple organ systems of the body. A better understanding of the disease helps to reduce the recurrences along with the physical, psychological, and social wellbeing of the patient. Regular follow up is essential to prevent any long-term complications of Behçet's.

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