

Behçet Syndrome, Clinical Presentation and Risk Factors

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ABSTRACT

Behçet's syndrome, a systemic vasculitis, manifests across multiple organ systems, presenting challenges in diagnosis and management. This chronic condition, characterized by recurrent oral and genital ulcers, cutaneous lesions, headaches, and ocular symptoms, poses complexities in its clinical evolution. The elusive etiology, potentially rooted in autoimmune reactions triggered by infection or environmental factors, complicates diagnostic endeavors. Diagnosis relies on evolving clinical criteria, necessitating the exclusion of other diseases, with imaging aiding in localizing symptoms. The differential diagnosis involves careful distinction from conditions with similar presentations. Ongoing research explores the intricate interplay of genetic factors, immunology, and environmental influences. A comprehensive understanding holds promise for refined diagnostics and targeted interventions, improving outcomes for those affected.

KEYWORDS: Behçet's syndrome, systemic vasculitis, autoimmune reaction, clinical criteria, diagnostic challenges, recurrent ulcers, cutaneous lesions, ocular manifestations, differential diagnosis, genetic factors, immunology, environmental influences, targeted interventions.

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INTRODUCTION

Behçet's syndrome is a complex systemic vasculitis that intricately involves various organ systems, including the epidermal, mucocutaneous, vascular, ophthalmologic, gastrointestinal, pulmonary, and central nervous systems. This chronic condition manifests through recurrent episodes of symptoms, with the most common being oral ulcers, genital ulcers, and uveitis. In certain cases, Behçet's syndrome can present more severely, involving complications like aneurysms and neural issues.

A distinctive aspect of Behçet's syndrome is the evolving nature of its clinical criteria, which may appear over time. The diagnostic process is not solely reliant on isolated symptoms; rather, it entails identifying a constellation of clinical criteria that may occur concurrently or in succession. Importantly, the diagnosis is confirmed by the careful exclusion of other potential diseases that might mimic Behçet's syndrome.

The therapeutic approach to Behçet's syndrome revolves around alleviating symptoms, often employing corticosteroids and immunosuppressive drugs. However, the administration of these medications is not without its challenges, as complications such as leukopenia and compromised immune function may arise, heightening the risk of infections.

Prognostic outcomes in Behçet's syndrome exhibit variability, contingent upon the specific organs involved and the frequency of symptomatic episodes. Navigating the diagnostic landscape of Behçet's syndrome requires an awareness of potential pitfalls, including the fact that symptoms may not manifest simultaneously. This temporal disjunction from symptom onset to diagnosis necessitates a comprehensive and time-sensitive diagnostic approach.

Furthermore, individuals at an elevated risk for Behçet's syndrome are often from geographic regions with a heightened prevalence of tuberculosis. The use of immunosuppressive drugs in the management of Behçet's syndrome poses an additional concern, as it can increase the risk of active tuberculosis in individuals with latent infections. This underscores the importance of a nuanced and geographically informed perspective in the comprehensive care and monitoring of individuals with Behçet's syndrome.

CLINICAL PRESENTATION AND RISK FACTORS

Behçet's syndrome, recognized as a complex systemic vasculitis, exhibits a spectrum of symptoms that intricately involve various organ systems. Its clinical presentation unfolds in an episodic manner, with different manifestations emerging at distinct intervals. The diagnostic journey for

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Behçet's syndrome is often prolonged, requiring months or even years to witness the evolution of a constellation of clinical features that align with established criteria.

Central to Behçet's syndrome are the recurrent aphthous oral ulcers, a hallmark feature that manifests more than three times a year and persists for 10 to 14 days. Intriguingly, these ulcers may precede the onset of other symptoms by considerable intervals. Alongside these, painful genital ulcers are pervasive, affecting a substantial percentage of cases (70%-90%). The dermatological landscape is further enriched with recurring cutaneous lesions, often marked by pathergy, and headaches, reported in over 80% of cases. Ocular involvement is a common facet, encompassing blurred vision and, occasionally, eye tearing or redness, with these symptoms typically surfacing within a few years of other manifestations.

The clinical narrative of Behçet's syndrome extends to encompass abdominal pain reminiscent of irritable bowel syndrome, intermittent arthritis prevalent in 40%-60% of cases, and a pervasive sense of fatigue. Within the clinical tapestry, familial connections may unravel a history tethered to Behçet's syndrome or related inflammatory conditions, adding a layer of complexity to the diagnostic landscape.

A meticulous clinical examination unveils nuanced features such as conjunctival injection, indicative of uveitis, and subsequent ocular manifestations like cataracts, synechiae, and band keratopathy. Oral ulcers, variably located within buccal mucosa, inner lip, tongue, hard and soft palate mucosa, and pharynx, coexist with potential indicators of meningeal inflammation, such as neck stiffness. Genital ulcers, cutaneous lesions, and sporadic focal neurologic abnormalities further contribute to the intricate clinical mosaic.

Despite the advancement in understanding Behçet's syndrome, its etiology remains elusive. The prevailing hypothesis suggests an autoimmune reaction triggered by infection or environmental stimuli, with a backdrop of predisposing genetic factors. Notably, the HLA-B51/B5 allele exhibits a robust association with the syndrome, and certain ethnic groups, particularly those from the Middle East and Southeast Asia, bear a higher prevalence.

The diagnostic process for Behçet's syndrome relies on the intricate weaving of clinical criteria over time, necessitating the exclusion of alternative diseases. While no specific diagnostic tests are recommended, the International Team for the Revision of the International Criteria for Behçet's Disease has devised a point system to aid in diagnosis. Imaging assumes a pivotal role in delineating neural, gastrointestinal, and large vessel involvement in cases where symptoms localize.

The labyrinth of differential diagnosis involves careful discernment to distinguish Behçet's syndrome from conditions like aphthous stomatitis, HSV infection, acne vulgaris, Crohn's disease, ulcerative colitis, vasculitis, and psoriatic arthritis. This requires a nuanced consideration of

the distinctive characteristics and locations of symptoms, adding layers of complexity to the diagnostic odyssey.

CONCLUSION

In conclusion, Behçet's syndrome stands as a complex and multifaceted systemic vasculitis, impacting diverse organ systems with a spectrum of episodic symptoms. Its diagnosis is intricate, often unfolding over months or years as a constellation of clinical features evolves. Recurrent oral and genital ulcers, cutaneous lesions, headaches, ocular manifestations, and systemic symptoms contribute to the intricate clinical tapestry. The elusive etiology hints at an autoimmune reaction triggered by infection or environmental factors, underscored by genetic predisposition, with the HLA-B51/B5 allele playing a significant role.

Navigating the diagnostic landscape requires a meticulous weaving of evolving clinical criteria and the exclusion of alternative diseases. The absence of specific diagnostic tests emphasizes the reliance on a comprehensive clinical evaluation, supported by imaging when symptoms localize. The differential diagnosis adds further complexity, necessitating careful discernment to distinguish Behçet's syndrome from various conditions with overlapping features. As research progresses, a deeper understanding of Behçet's syndrome may pave the way for more targeted diagnostic approaches and effective treatment strategies. The intricate interplay of genetic, immunologic, and environmental factors remains a focal point for ongoing investigations, offering hope for improved outcomes and quality of life for individuals grappling with this enigmatic disorder.

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