Neuro-Behçet's Disease in a Middle Eastern Male: A Rare and Challenging Diagnosis

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Abstract:

The exact cause of Behçet's disease is unknown, but both genetic and environmental factors are believed to contribute. Its higher prevalence along the "Silk Route" and familial clustering suggest a genetic component, though it does not follow Mendelian inheritance. The strongest association is with HLA-B51/B5 carriers, who are at increased risk of developing the disease [9]

Hypersensitivity to Streptococcus sanguinis antigens, along with other infectious agents like Staphylococcus aureus, Herpes simplex virus type 1, and Prevotella species, has been linked to Behçet's disease [10][11].

Behçet's disease is an autoinflammatory vasculitis affecting arteries and veins of all sizes, without necrotising vasculitis or giant cell formation. Pulmonary and arterial aneurysms are distinctive features [1].

Unlike autoimmune diseases, Behçet's disease lacks specific autoantibodies, with cell-mediated immunity central to its pathogenesis. T-helper 1 activation boosts circulating T-lymphocytes, driving systemic symptoms, while elevated pro-inflammatory cytokines (IL-1, IL-12, IL-17, TNF) contribute to disease progression. Mucocutaneous lesions, such as oral aphthae, pustules, and erythema nodosum, result from neutrophil overactivation causing vascular injury [2].

Circulating immune complexes, anti-endothelial cell antibodies, and endothelial dysfunction further contribute to the disease process [3].

Synovial fluid in Behçet's disease typically shows a neutrophil-predominant leucocytosis, ranging from 300 to over 30,000 cells/mm³ [4]. The disease tends to follow a more severe course in males and younger individuals, with most cases occurring sporadically [5]