Behcet Disease

Behcet disease is a systemic vasculitis affecting arteries and veins of all sizes. It occurs most frequently in young adults from Mediterranean countries like Turkey and eastern Asian countries like Japan and is associated with HLA-B51 positivity. This disease is characterized by vasculitis of all sized vessels. In the lung, a highly specific pulmonary artery aneurysm may be evident. Other clinical features include recurrent and painful oral ulcers, erythema nodosum, uveitis, and thrombosis. Diagnosis is established with the help of a positive pathergy test. Management strategies include steroids and other immunosuppressants.



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Characteristics

Mediterranean and East Asian Populations
Turkey-flag and Japan-flag
Behcet disease is most common in patients from the Mediterranean region and eastern Asia, especially in Turkey and Japan. It is also referred to as the Silk Road disease. It usually presents in young adults, usually between 30 and 40 years of age. It is rare for it to present for the first time in those over 50 years old or before puberty.

HLA-B51
HuLA-hoop B51-bomber
Behcet disease is strongly associated with HLA-B51, suggesting a genetic component. Other diseases associated with this serotype include mucocutaneous lymph node syndrome and rubella.

Clinical Features

Vasculitis
Vessels-on-fire
Behcet disease is characterized by vasculitis affecting vessels of all sizes. It is thought to be due to an autoimmune reaction. Infectious triggers and molecular mimicry have also been proposed.

Pulmonary Artery Aneurysm
Lung Archer with Bulging-aneurysm
Vascular involvement is an important cause of morbidity and mortality in Behcet disease. This systemic vasculitis can lead to pulmonary artery aneurysms, which carry significant mortality. Pulmonary artery aneurysms usually involve the large proximal branches of the pulmonary arteries. Hemoptysis is the most common presenting symptom. Other symptoms include cough, dyspnea, fever, and pleuritic pain.

Recurrent Painful Ulcers
Recurring-clock Pain-bolts and Ulcer-volcano
The most common and classic clinical finding in Behcet disease is recurrent painful oral (aphthous) ulcers. This is typically the presenting symptom. Ulcers usually last 1-4 weeks and resolve spontaneously. Patients may also present with recurrent genital ulcerations, most commonly on the vulva in females and on the scrotum in males. These ulcers may be single or multiple and they are similar to the oral aphthous ulcers.

Erythema Nodosum
Nodosaur with Red Bumps on Legs
Patients with Behcet disease may also present skin lesions such as erythema nodosum, which is defined as inflammation of the subcutaneous fat typically on the anterior shins. Other skin lesions that may be found include papulopustular lesions, pyoderma gangrenosum, and acneiform eruptions.

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Uveitis

UV-sunglasses

Another clinical finding in Behcet disease are eye lesions. Patients may present with uveitis, keratitis, and retinal vasculitis. The eye disease is typically bilateral and usually presents after the onset of the ulcers.

Thrombosis

Trombone

Due to the vasculopathy caused by the inflammatory process in the vessels, patients with Behcet disease may present with thrombosis. Thrombosis may occur in the large veins and lead to deep vein thrombosis or Budd-Chiari syndrome, or may present with arterial thrombosis.

Diagnosis

Positive Pathergy Test

Positive Patch-ghee

A positive pathergy test aids in the diagnosis of Behcet disease. A positive test is defined as exaggerated skin ulceration with minor trauma such as the one produced with a needlestick. It is positive when an erythematous papule or pustule appears after 24-48 hours of a needle prick.

Management

Steroids

Steroid-stairs

Topical corticosteroids are indicated for oral and genital ulcers. Topical lidocaine can be added as a symptomatic treatment for pain. Systemic corticosteroids are also indicated in case of eye involvement, such as uveitis, and vasculopathy.

Immunosuppressants

Moon-suppressed

Immunosuppressive medications in addition to systemic glucocorticoids are indicated for the management of systematic involvement in Behcet disease. For instance, posterior uveitis requires management with intensive immunosuppression to avoid vision loss. The immunosuppressive regimen chosen depends on the severity and clinical involvement.

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