

Lecture No. 5

Behcet's Disease

Behcet's disease is a **rare, chronic, lifelong disorder** that involves **inflammation of blood vessels** throughout the body. It is a **form of vasculitis** that can lead to **ulceration** and other **lesions**, slightly affecting **more men than woman** characterized by a triad of **symptoms**, including aphthous **ulceration of the oral mucous membranes and genitalia and uveitis**. **Rare manifestations** include **oligoarthritis** of the **lower extremities**, **vasculitis of the pulmonary vessels, cerebrovascular** symptoms. **The etiology** of the Behcet's disease is **unknown**. It is believed to be partly **genetic**, an association with **HLA-B51** has been found.

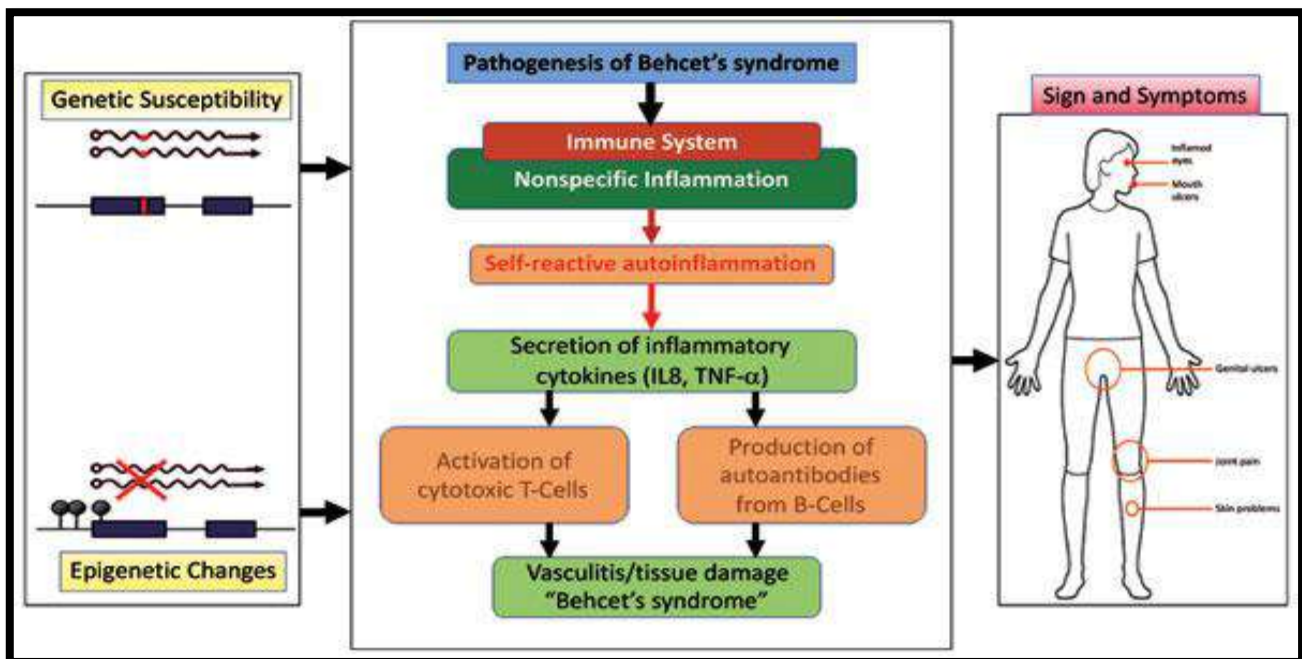


Figure: Pathogenicity of Behcet's disease

Risk factors

- ❖ **Age.** Behcet's disease most commonly affects **men and women in their 20s, 30s and 40s**, though children and older adults also can develop the condition. When the condition occurs at an **earlier age**, it tends to be **more severe**.
- ❖ **Geography.** Although the disease occurs worldwide, people from countries in the **Middle East and Asia**, including **Turkey, Iran, Iraq, Japan and China**, are more likely to develop Behcet's.
- ❖ **Sex.** While Behcet's disease occurs in **both men and women**, the disease is usually **more severe in men**.
- ❖ **Genes.** Having certain genes **HLA-B51** is associated with a higher risk of developing Behcet's.

Clinical features

- ❖ **Mouth:** Painful mouth sores, identical to canker sores, are the most common sign of Behcet's disease. Sores begin as raised, round lesions in the mouth that quickly turn into painful ulcers. The sores heal usually in seven to 21 days, though they do recur.
- ❖ **Skin:** Skin lesions may occur in people with Behcet's disease. Skin problems can vary. Some people may develop acne-like sores on their bodies. Others may develop nodules on the lower legs.
- ❖ **Genitals:** People with Behcet's disease may develop sores on their genitals. The sores most commonly occur on the scrotum or the vulva.
- ❖ **Eyes:** Behcet's disease may cause inflammation in the eye — a condition called uveitis. In people with Behcet's disease, uveitis causes redness, pain and blurred vision in one or both eyes.
- ❖ **Joints:** Joint swelling and pain most commonly affect the knee in people with Behcet's disease.
- ❖ **Vascular system:** Inflammation in veins and large arteries may occur in Behcet's disease, causing redness, pain and swelling in the arms or legs when a blood clot results.
- ❖ **Digestive system:** Behcet's disease may cause abdominal pain, diarrhea or bleeding.
- ❖ **Brain:** Inflammation in the brain and nervous system that leads to headache, fever, disorientation, poor balance or stroke.

Diagnosis

There is no diagnostic test and the diagnosis is entirely clinical. Laboratory findings are nonspecific and reflect the inflammatory state, C-reactive protein levels, erythrocyte sedimentation rate (ESR), leukocyte count, complement components, and acute-phase reactants may all be elevated during an acute attack. Internationally accepted diagnosis criteria have been published recently.

International Clinical Criteria for Behcet's Disease

An international group of physicians has established a set of guidelines to aid in the classification of Behcet's patients. The International Clinical Criteria for Behcet's Disease classification states patients must present with:

- Recurrent oral ulcerations (aphthous or herpetiform) at least three times in one year.
- Additionally, patients must present any two of the following:
 - Recurrent genital ulcerations.
 - Eye lesions (uveitis or retinal vasculitis) observed by an ophthalmologist.
 - Skin lesions (a variety of rashes or acne-like sores) may be caused by Behcet's disease. Positive
 - Positive pathergy test, your doctor inserts a sterile needle into your skin and then examines the area two days later. If the pathergy test is positive, a small