BEHCET’S DISEASE

Symptoms and Treatment in Behcet's disease

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Behcet's is a chronic autoimmune disorder characterized by vasculitis, oral and genital ulcers, and uveitis, which can occur in patients with hepatitis C.

Symptoms in Behcet’s Disease

Behcet's disease is a chronic inflammatory autoimmune disorder that causes a constellation of different symptoms, including vasculitis, arthritis, bowel inflammation, meningitis, cranial nerve palsies, memory loss, impaired speech, memory loss impaired balance, and blisters and ulcerations in the mouth, eyes and genitals. Behcet's disease may progress to blindness, stroke, central nervous system impairment, spinal cord swelling, and intestinal complications.

Like most autoimmune disorders, Behcet's disease causes symptoms that wax and wane with periods of symptoms alternating with periods of remission.

Who is Affected?

Behcet's disease is thought to have a viral connection, as it is known to occur in patients with chronic hepatitis C and other viral conditions. Behcet's disease is most commonly seen in people from the "Old silk route" including Japan, China, Iran, Turkey, and Israel although it's known to occur worldwide and its incidence appears to be increasing in the United States. In Japan, Behcet's disease is one of the leading causes of blindness.

Persons with the immune system antigen HLA-B51 are predisposed to developing Behcet's although only about 5 percent of people with this gene are affected, suggesting that environmental triggers are an important factor.

As mentioned, many people with Behcet's develop this disorder following or in conjunction with a viral infection. Prior to the onset of Behcet's patients may report symptoms of malaise, loss of appetite, weight loss, generalized weakness, headache, body temperature changes, perspiration, swollen lymph glands, substernal and temporal pain, sore throat, muscle pain, and tonsillitis. Symptoms of Behcet's may develop suddenly or over a slow course lasting as long as 10 years from the initial symptoms.

Vasculitis

Vasculitis in Behcet's disease can affect blood vessels of all sizes and types. About 7-29 percent of patients with Behcet's disease develop vasculitis, and men are more likely to be affected than women. Veins and arteries in the following organs are most likely to be
affected: eyes, mouth, skin, lungs, joins, brain, genitals, and gastrointestinal tract. Inflammation may appear in front of the eye (anterior uveitis) or in the back of the eye (posterior uveitis) and both parts of the eye can be affected simultaneously. Anterior uveitis causes pain, blurred vision, light sensitivity, tearing and redness. Posterior uveitis is considered more dangerous and is more likely to cause blindness because of its predilection for the retina. Patients with Behcet's disease may develop pulmonary aneurysms or outpouchings of the blood vessels in their lungs that may lead to massive lung hemorrhage. Vascular lesions seen in Behcet's disease include arterial occlusions, venous occlusions, aneurysms, and varices.

**Blisters and Lesions**

Blisters in the mouth known as aphthous ulcers are similar to and indistinguishable from canker sores. In Behcet's disease, multiple lesions occur and are frequently large and painful. Ulcers and blisters can occur on the tongue, lips, and inside the cheek. Although symptoms in Behcet's disease are variable, most patients develop these mouth ulcerations. Pustular lesions resembling acne or folliculitis can appear anywhere on the body. Reddened lesions known as erythema nodosum are also commonly seen and may appear on the face, neck or arms. Blistering lesions may occur on the scrotum in males and genital ulcers may affect the vulva in females. The joint pain or arthralgia in Behcet's disease does not usually cause joint swelling.

**Nervous System Involvement**

One of the most dangerous complications to occur in Behcet's disease is central nervous system involvement. In this case, the white matter of the brain and brainstem is likely to be affected. Symptoms include headaches, confusion, strokes, personality changes and conditions of meningitis that are not associated with infection. Ocular involvement is the major cause of morbidity and the most dreaded complications because of its occasional rapid progression to blindness. Pregnant patients with Behcet's disease may notice an increase in symptoms, especially during the first trimester.

**Diagnosis**

Behcet's disease is diagnosed in persons with symptoms and signs of the disease who show signs of vasculitis on biopsy for which there are no other causes. Criteria for diagnosis can also include two of the following four symptoms: recurrent oral ulcerations, recurrent genital ulcerations, eye lesions (uveitis, retinal uveitis, iridocyclitis, chorioretinitis), skin lesions such as erythema nodosum or subcutaneous thrombophlebitis, or folliculitis and a positive pathergy test. In the pathergy test, the skin is pricked with a small sterile needle containing saline or dilute histamine. The formation of a small red bump with inflammation either immediately or within two days constitutes a positive result. In active Behcet's disease, levels of vascular endothelial growth factor, a proinflammatory cytokine, are significantly elevated and the erythrocyte sedimentation rate and C-reactive protein level are increased.
Treatment

There is no permanent cure for Behcet's disease and permanent remission has not been reported. Treatment for Behcet's disease is used to reduce symptoms and varies depending on the predominant symptoms. Medications such as corticosteroids and immunosuppressant agents such as azathioprine, chlorambucil and cyclosporine, are used to reduce inflammation and suppress the immune response. Oral gels and pastes such as Kenalog, Trental (Pentoxifylline) and colchicine are sometimes used to reduce ulcerations.

Thalidomide has been proven effective in patients with mucocutaneous (mucus membrane lesions) symptoms. Interferon-alpha, a drug used in hepatitis C can also offer benefits. Lifestyle changes that benefit Behcet's include a nutrient-rich diet, moderate exercise, proper rest, and an avoidance of environmental chemicals.

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