

19 - 376 Behçet Syndrome

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Behçet Syndrome PART 11 Immune-Mediated, Inflammatory, and Rheumatologic Disorders Behçet syndrome is a systemic vasculitis, first described by Hulusi Behçet, a Turkish dermatologist. It can present with oral and genital ulcers, skin lesions, uveitis, arthritis, major arterial and venous vessel disease, and gastrointestinal and neurologic manifestations. These manifestations can be present in various combinations and sequences over time. Patients are most commonly from the Middle East, the Mediterranean region, and the Far East; it is most prevalent in Turkey, with a prevalence of 1 in 250 adults. It is relatively rare before the late teens and after age 50. Males and females are equally affected; however, males frequently have more severe disease and poorer outcomes. Some manifestations may show regional differences; for example, gastrointestinal involvement, rare in Turkey, is more common in Japan and is seen in ~30% of patients in the United States. ■

■ **DIAGNOSIS** Behçet syndrome is diagnosed clinically. There are no specific laboratory, imaging, or histologic features that can help in the diagnosis of a patient with suggestive symptoms. However, these can be used in ruling out conditions that may mimic Behçet syndrome and the diagnosis is based on a combination of clinical features in the setting of ruling out other potential causes. In this regard, some patients may require months to years to develop the array of symptoms that would lead to a definitive diagnosis, although a tentative diagnosis may be made well before. The most commonly used and best performing diagnostic criteria are the International Study Group (ISG) criteria (sensitivity ~95%, specificity ~96%); patients need to have recurrent oral ulcers plus two of the following four clinical manifestations: recurrent genital ulcers, skin lesions, eye lesions, or a positive pathergy test (Table 376-1). Additional clinical manifestations may involve various organ systems, including the gastrointestinal, vascular, pulmonary, and central nervous systems. Up to 50–60% of patients, depending on where they are from, can be positive for HLA B*51; however, it is not used as a diagnostic test because it is also found in around 20% of the normal population. ■ ■ **PATHOGENESIS** The pathogenesis and etiology of Behçet syndrome are unknown. Family studies show a possible genetic predisposition, and increased inflammation and immunologic mechanisms play a role. Both innate and adaptive immune systems may be involved. Unlike other autoimmune diseases, however, Behçet syndrome is not typically associated with autoantibodies, Raynaud's phenomenon, Sjögren's syndrome, thrombocytopenia, hemolytic anemia, sun hypersensitivity, serosal involvement, or an increased risk for other autoimmune diseases. On the other hand, features that separate it from autoinflammatory conditions include tendency to abate with time, absence of mutations

TABLE 376-1 International Study Group Criteria for the Diagnosis of Behçet Syndrome

CRITERIA	FREQUENCY
Oral ulcers	~98%
At least 3 times in a 12-month period	Plus 2 out of 4 from below:
Recurrent genital ulcers	~80%
Usually scarring	Skin lesions ~80%
Erythema	

nodosum, pseudofolliculitis, papulopustular or acneiform nodules (postadolescent, not receiving corticosteroids) Eye lesions ~50% Anterior or posterior uveitis, cells in vitreous or retinal vasculitis Pathergy ~50% Evaluated in 24–48 h, after dermal insertion of a 20-gauge needle

associated with autoinflammatory diseases, and higher prevalence than typical autoinflammatory diseases such as familial Mediterranean fever (Chap. 381). There is neutrophil hyperreactivity; however, it is not clear whether this is primary or secondary to cytokine-directed activation. There is also evidence from retrospective patient cohort analyses that there may be different clusters of disease presentation; for example, acne lesions are more commonly seen with arthritis and associated with enthesitis, and each of these clusters may have a different pathogenesis. ■

■ **CLINICAL PRESENTATION** The most common symptoms are associated with mucocutaneous tissues. Oral ulcers are seen in virtually all patients and are commonly the first manifestation (Fig. 376-1). Commonly, like ordinary canker sores, they are usually multiple. They last around 10 days but recur unless treated. Only the uncommon, major ulcers tend to scar. Beneficial effects of dental and periodontal therapies suggest that decreased oral health is associated with disease severity. Genital ulcers are the most specific lesions, most commonly occurring on the scrotum or labia (Fig. 376-1). They are larger and deeper and take longer to heal than oral ulcers and tend to form scars. Acne-like or papulopustular lesions are indistinguishable from acne vulgaris in appearance and pathology. They are seen both at the usual acne sites as well as at uncommon sites such as lower extremities. Other skin findings are the nodular lesions, which are of two types: erythema nodosum lesions due to panniculitis and superficial vein thromboses. Superficial thrombophlebitis often occurs in men and is associated with deep-vein thrombosis; it should trigger workup for other vascular involvement, including pulmonary artery aneurysms. Pathergy reaction is a nonspecific hyperreactivity of the skin to trauma. Typically, a papule or pustule forms in 24–48 h after a A B

FIGURE 376-1 Clinical findings in Behçet syndrome. A. Behçet oral ulcer. B. Behçet scrotal ulcer.

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