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. Commonly, like ordinary cancer 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. 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 needle prick. It is rather unique for Behçet syndrome and is part of the ISG diagnostic criteria.

Arthralgia or arthritis is seen in about half of patients; it is usually a mono- or oligoarthritis in the lower extremities and does not usually cause deformity or erosions.

Eye involvement is seen in half of all patients and in ~70% of males. It is most commonly a bilateral panuveitis. A hypopyon, seen in ~10% of patients with eye disease, is an intense inflammation in the anterior chamber and is quite specific for Behçet syndrome.