Table 2 Main differential diagnoses

Organ/system and specific feature	Histology and investigations	Differentials
Skin		
Erythema nodosum-like lesions	More common in female than male patients Inflammatory perivascular infiltrate can be seen, unlike in other types of erythema nodosum They present as either lobular or mixed septal and lobular panniculitis	Other causes of panniculitis
Papulopustular lesions	Sterile pustules, pseudofolliculitis, acne-like lesions	Acne vulgaris Folliculitis Furuncles
Neutrophilic dermatoses	Increased neutrophilic infiltrate seen (pyoderma gangrenosum is a diagnosis of exclusion with no specific histological features)	Idiopathic Sweet syndrome and pyoderma gangrenosum
Mucosal ulceration		
Aphthous oral ulcer (recurrent aphthous stomatitis)	Recurrent and painful ulcers affecting the mucosal surface of the lips, soft palate, buccal mucosa and tongue. These may be minor or major (heal with scarring)	Idiopathic recurrent aphthous ulcers Aphthous ulcers associated with inflammatory bowel disease
Aphthous genital ulcer	Recurrent and painful ulcers; the labia minora and majora are the most commonly affected sites in female patients The scrotum and perianal area are the most commonly affected sites in male patients	Idiopathic recurrent aphthous ulcers Aphthous ulcers associated with inflammatory bowel disease
Ocular		
Anterior uveitis	Granulomatous vs. nongranulomatous keratic precipitates The bulk of inflammation is seen anterior to the crystalline lens	Idiopathic uveitis Chronic anterior uveitis: anterior uveitis is not a classic defining ocular finding in Behçets, but can be supportive
Intermediate uveitis	The bulk of inflammation is seen behind the crystalline lens and anterior to the retina Significant inflammation can be seen within the vitreous in the context of an inflammatory retinal vein occlusion	Intermediate uveitis is not a classic defining ocular finding in Behçets, but can be supportive
Posterior uveitis	The bulk of inflammation is within the retina or choroid Inflammatory retinal vein occlusions resemble a traditional retinal vein occlusion but can be multifocal and occur in the presence of intraocular inflammation. While retinal vein occlusions occur commonly in the general population, they are typically a sign of cardiovascular disease and occur in the older population Retinal infiltrates may occur within the inner retina and cause irreversible retinal destruction. Infiltrates may be confused with an infectious aetiology. Common intraocular infections should be excluded including toxoplasmosis, tuberculosis and syphilis	Both inflammatory retinal vein occlusions and retinal infiltrates are a common ocular finding in Behçets
Scleritis and episcleritis	Focal or diffuse redness of the sclera or episclera are usually associated with pain or discomfort Other inflammatory causes should be excluded. Both infections and inflammatory diseases of collagen can cause scleritis or episcleritis	While scleritis and episcleritis are reported in Behçets, they are not commonly thought of as disease-defining events
Neurological		
Brainstem presentation	Neuro-Behçets can be differentiated from its mimics by a combination of characteristic clinical and paraclinical neurological findings in addition to the associated systemic features	Multiple sclerosis, stroke affecting younger people (< 45 years of age), intracranial hypertension, meningoencephalitis and myelitis
Myelitis	Spinal cord intrinsic signal change	Multiple sclerosis
Meningoencephalitis	CSF pleocytosis (neutrophils and lymphocytes) and raised CSF protein, usually absent oligoclonal bands	Infectious or other inflammatory causes of meningoencephalitis
Venous sinus thrombosis	Venous drainage obstruction on venography	Other neuroinflammatory conditions, antiphospholipid syndrome
IIH (secondary IIH)	Raised CSF pressure	Idiopathic or primary IIH
Vascular		
Arterial aneurysm	Abnormal imaging: computed tomography angiography, MRI angiography, conventional angiography or ultrasound, showing aneurysms Echocardiography	Takayasu arteritis Atherosclerosis Treponemal infection Ehlers–Danlos syndrome Marfan syndrome Turner syndrome Familial thoracic aortic aneurysms
Deep-vein thrombosis	Abnormal duplex venous ultrasonography with obstruction Venous drainage obstruction on venography Elevated D-dimer Factor V Leiden	Prothrombotic clotting disorder Prolonged inactivity Chemotherapy Pregnancy Obesity

(*Continued*)