Imaging Findings in 3 Special Conditions of Behçet’s Disease

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Three special subclassifications of Behçet’s disease (BD), associated with the predominant lesion in the gastrointestinal (GI) tract, large vessels and central nervous system (CNS), are recognized; these are intestinal, angio- and neuro-Behçet’s disease, respectively. These three varieties are associated with high morbidity and mortality, and radiologic imaging plays a critical role in their detection. The purpose of this report is to describe the imaging findings of Behçet’s disease involving the GI tract, large vessels, and CNS and to discuss the differential diagnostic considerations.

Index word: Behçet’s disease

Behçet’s disease, originally described as a syndrome of aphthous stomatitis, genital ulceration, and uveitis, is recognized as a systemic disease with variable involvement of many organ systems (1). The disease affects mainly men aged 20–30 and is characterized by the exacerbation of symptoms and remission of unpredictable duration. Diagnosis of BD is based only on clinical findings, since no suitable pathognomic laboratory or histopathologic criteria exist. Current diagnostic criteria for BD consist of the presence of recurrent oral ulcerations plus two of the following conditions: recurrent genital ulceration, eye lesions, skin lesions, and a positive pathergy test. Minor criteria include subcutaneous or deep vein thrombophlebitis, epididymitis, arterial occlusion and/or aneurysm, family history, and gastrointestinal features (1).

Three special subclassifications, associated with the predominant lesion in the gastrointestinal (GI) tract, large vessels and central nervous system (CNS), are recognized; these are intestinal, angio- and neuro-Behçet’s disease, respectively. When confined to mucocutaneous manifestation, the disease has an indolent course, it presents with intestinal, vascular, and neurologic involvement, but the patient’s condition tend to deteriorate rapidly (1, 2). The early recognition of these special varieties of BD is important and radiologic imaging plays a critical role in their detection and appropriate management.

Intestinal Behçet’s disease

BD affects the GI tract in an estimated 10–15% of cases (2); pathologically, extensive, deeply penetrating ulceration is its hallmark. These ulcers are associated with perivascular lymphocytic infiltration and vasculitis, and tend to involve submucosa, muscle layer, and the entire intestinal wall, resulting in a deep and undermining ulcer similar to a benign gastric or duodenal ulcer.

The characteristic radiologic findings of intestinal BD are deep ovoid or geographic ulcers, with surrounding deformity, which tend to localize in the ileocecal region (Fig. 1) and wax and wane with medical treatment. They frequently perforate at multiple sites or bleed. Recurrence of the lesion near surgical anastomoses is also common; such a lesion tends to show the common radiographic features of the disease (Fig. 2). Less commonly, intestinal BD manifests as multiple shallow ulcers, innumerable aphthoid ulcers, or a longitudinal ulcer seen on double-contrast barium enema to involve any segment of the colon. Other segments of the small bowel (Fig. 3) or esophagus (Fig.
4) can be involved in BD. Although barium examination is the most useful method of diagnosing intestinal BD, CT frequently reveals nonspecific bowel wall thickening, with contrast enhancement, in the ileocecal region (Fig. 1. B). CT is helpful in evaluating the complications of intestinal BD, including bowel perforation or subsequent abscess formation, which may require surgical intervention.

Differential diagnosis of intestinal BD includes Crohn disease, tuberculous enterocolitis, ulcerative colitis, and other colitides. In the case of intestinal BD, ulcers tend to be larger and deeper than in Crohn disease or tuberculous enterocolitis, and may show a greater tendency to localize in the ileocecal region. The presence of a longitudinal ulcer of cobblestone appearance, and stricture are relatively uncommon in intestinal BD. Tuberculous enterocolitis is another important differential diagnosis, especially in endemic areas. Compare with intestinal BD, tuberculous enterocolitis tends to involve a long segment of the intestine, with severe mucosal irregularity and deformity of the ileocecal valve, and the cecum (3). The radiographic

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**Fig. 1.** Intestinal Behçet’s disease in a 34-year-old man who had oral ulcers, genital ulcers, and positive pathergy test.

A. Double contrast radiograph shows three ovoid deep ulcers (arrows) in the terminal ileum and cecum, smooth folds radiating to edge of ulcer craters, and mild deformity in medial aspect of cecum.

B. Contrast enhanced CT scan at the level of ileocecal valve shows irregular wall thickening of terminal ileum and cecum (arrows), and adjacent inflammation with mesenteric stranding.

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**Fig. 2.** Recurrent intestinal Behçet’s disease at near the surgical anastomosis in a 49-year-old man with a history of right hemicolecction 1 year ago due to the same disease at the ileocecal region.

A. Small bowel radiograph shows a large deep ulcer (asterisk) in the ileum proximal to ileocolic anastomosis and thickened surrounding mucosal folds (arrows).

B. Contrast enhanced CT scan at the level of ileocolic anastomosis (open arrow) shows extensive bowel wall thickening of ileum (asterisks) and transverse colon (solid arrows) with marked surrounding inflammatory infiltration.
Fig. 3. Intestinal Behçet’s disease involving the ileum in a 42-year-old man who had oral ulcer, genital ulcer, perianal ulcers, and positive pathergy test. Double contrast radiograph shows healed ulcer (arrows) with converging mucosal folds.

Fig. 4. Intestinal Behçet’s disease involving the esophagus in 33-year-old man who had oral ulcers, genital ulcers, skin lesion, and uveitis. Double contrast radiograph shows longitudinal ulcer (open arrows) during healing and retraction of esophageal wall (solid arrows).

Fig. 5. Angio- Behçet’s disease involving large deep vein in a 32-year-old man who had oral ulcers, genital ulcers, recurrent deep vein thrombosis, and positive pathergy test. Contrast enhanced CT scan at the level of thyroid cartilage shows nonopacification of central portion (asterisk) of right internal jugular vein with peripheral rim enhancement (solid arrows), surrounding soft tissue edema, and multiple collateral veins. Note also obliteration of left internal jugular vein (open arrow), which is suggestive of chronic thrombosis.

Fig. 6. Thrombosis of the superior vena cava in a 27-year-old man who had oral ulcers, positive pathergy test, skin lesion, and multiple joint pain.
A. Posteroanterior chest radiograph shows smooth mediastinal widening (arrows).
B. Contrast enhanced CT scan shows nonopacification of superior vena cava with peripheral rim enhancement (open arrow), increased attenuation of mediastinal fat, which is suggestive of edema, and multiple collateral vessels (solid arrows) in mediastinum and chest wall.
Fig. 7. Aneurysms of the right and left pulmonary arteries in a 38-year-old man with massive hemoptysis. He had a history of recurrent oral ulcers, genital ulcers, and deep vein thrombosis in right lower extremity.

A. Initial posteroanterior chest radiograph shows a mass (large arrow) in the right upper lung and a nodule (small arrows) in the left upper lung.

B. Contrast enhanced CT scan shows a large aneurysm (asterisk) with enhanced patent lumen.

C. Intravenous digital subtraction angiogram shows a aneurysm (arrows) of the posterior segmental branch of right pulmonary artery.

D. Follow-up posteroanterior chest radiograph obtained 1 year later shows multiple coils introduced for embolization in both upper lung with complete regression of aneurysms of both pulmonary arteries.

findings of intestinal tuberculosis are very similar to those of Crohn disease and differentiation from intestinal BD is difficult. The loss of hausturations and the presence of multiple shallow ulcers in the affected segment of the colon, characteristic findings of ulcerative colitis are seldom demonstrated radiographically in intestinal BD. The radiographic findings are, however, similar and differentiation of intestinal BD from these diseases may be difficult or impossible. For differential diagnosis, a careful investigation of the clinical findings of BD is very helpful.

Angio-Behçet’s disease

BD with the involvement of large or medium-sized vessels, namely angio-BD, is divided into three groups: venous occlusion, arterial occlusion, and arterial aneurysm formation. Venous lesions are more common than arterial, and are noted in approximately one-third of all patients with BD. The pathogenesis of venous occlusion is considered to be vasculitis with superimposed thrombosis, and this may not be prevented by anticoagulant therapy. Venous lesions manifested as superficial or deep vein thrombophlebitis involving primarily the legs, less often the arms; migrating thrombophlebitis may also be seen, though this is rare. In the case of caval occlusion, the process usually begins adjacent to the affected large vein.

Noninvasive methods such as ultrasound with
Doppler, CT (Fig. 5), or MR angiography have more recently become the diagnostic tool of choice for deep vein thrombosis. Lower extremity venography, however, remains an important diagnostic tool for defining calf vein thrombi. In the case of involvement of major thoracic vessels, chest radiographs may show medias-

Fig. 8. Angio-behçet’s disease involving the left subclavian artery in a 24-year-old woman who had oral ulcers, uveitis, genital ulcers, and positive pathergy test. Digital subtraction arch aortogram shows segmental luminal narrowing of the subclavian artery (arrows). This finding is very similar to that of pulseless disease.

Fig. 9. Angio-Behçet’s disease involving the right common iliac artery (RCIA) in a 24-year-old man who had genital ulcers, skin lesion, and epididymitis. Distal abdominal aortogram shows complete occlusion of right external iliac artery, irregular luminal dilatation of RCIA, and an aneurysm (open arrow) at bifurcation of RCIA. Note also prominent right internal iliac artery with focal stenosis (solid arrow) at immediately distal to bifurcation of RCIA.

Fig. 10. Central venous thrombosis involving the superior sagittal sinus (SSS) with bilateral cerebral venous infarction in a 40-year-old woman who had oral ulcers, genital ulcers, uveitis, and positive pathergy test. 
A. Axial T2-weighted (3500/96) image shows high signal intensity in bilateral parietal areas (arrows). The hyperintensities are not confined to arterial territories.
B. Contrast enhanced T1-weighted coronal image shows isointense signal (long arrow) within the lumen of SSS with peripheral enhancement (open arrows), which is suggestive of thrombus. Note also the enhancement of cortical ribbon (small arrows) in left high parietal area.
C. Venogram of SSS shows tubular filling defect (arrow) in the lumen of SSS.
Neuro-Behçet’s disease

Neurologic manifestation in BD is observed in 10–15% of patients and the prognosis has generally been poor (2). Neurologic involvement can be divided into three categories: cerebral venous thrombosis (CVT), CNS involvement, and recurrent meningoecephalitis (5). CVT is observed in one-third of patients with neurologic manifestations and the clinical features and imaging findings are similar to those of a patient with CVT of other origin. Isolated headache is the usual presenting symptoms. Conventional angiography is still required for diagnosis but MRI has been shown to be a safe and reliable method for the diag-
nosis of dural sinus thrombosis. MRI shows direct signs of thrombosis, with or without associated cerebral venous infarction (Fig. 10). The important MRI finding of dural sinus occlusion is abnormal signal intensity, representing thrombus within the affected dural sinus instead of signal void. The signal intensity of thrombus varies with clot age.

CNS involvement is a severe and devastating manifestation of BD. The basic lesion is a chronic, relapsing, inflammatory, cellular infiltration around venules and capillaries and occasionally around arteries. Radiologic findings generally correlate well with neurologic symptoms and may be reversible in response to medical treatment. For the detection of brain lesions in BD, MRI is more sensitive than CT. The characteristic MRI findings consist of multiple small foci of high signal intensity, seen on a T2-weighted image; these are iso or hypointense relative to gray matter on T1-weighted image, and show variable enhancement (Fig. 11). The lesions occur most frequently within the brain stem, thalami, and basal ganglia, but similar foci may be seen in the cerebral hemisphere (Fig. 12), cerebellum, or spinal cord. Rarely, the disease manifests as a mass lesion simulating an intra-axial neoplasm.

The MRI findings are not specific to BD and can be observed in other type of vasculitis of the CNS, such as systemic lupus erythematosus (SLE), though in SLE, the brain stem is rarely involved. Multiple sclerosis should be included in differential diagnosis, since some features which favor BD include the involvement of basal ganglia and thalami, the absence of periventricular predominance, and the involvement of ventral pons. Other differential diagnoses of neuro-BD include brain stem infarction, rhomboencephalitis, and dilated perivascular spaces.

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