

Imaging Findings in 3 Special Conditions of Behçet's Disease¹

Jae Cheol Hwang, M.D., Hyun Kwon Ha, M.D.
Jeong Hyun Lee, M.D., Yong Ho Auh, M.D.

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 penetrative 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.

¹Department of Diagnostic Radiology, Asan Medical Center, University of Ulsan College of Medicine

Received June 19, 1997; Accepted November 10, 1997

Address reprint requests to: Jae Cheol Hwang, M.D., Department of Diagnostic Radiology Asan Medical Center University of Ulsan College of Medicine

388-1 Poongnap-dong, Songpa-ku, 138-040 Seoul

Tel. 82-2-224-4400 Fax. 82-2-476-4719

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

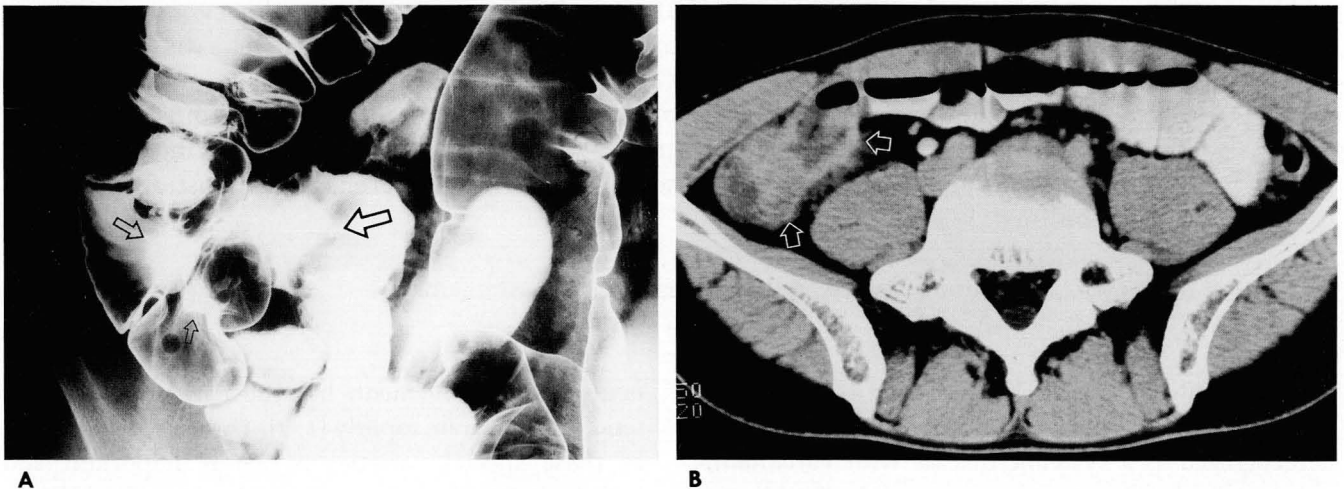


Fig. 1. Intestinal Behçet's disease in a 34-year-old man who had oral ulcers, genital ulcers, and positive pathergy test.
A. Double contrast radiograph shows three ovoid deep ulcers (arrows) in the terminal ileum and cecum, smooth folds radiating to edge of ulcer craters, and mild deformity in medial 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.

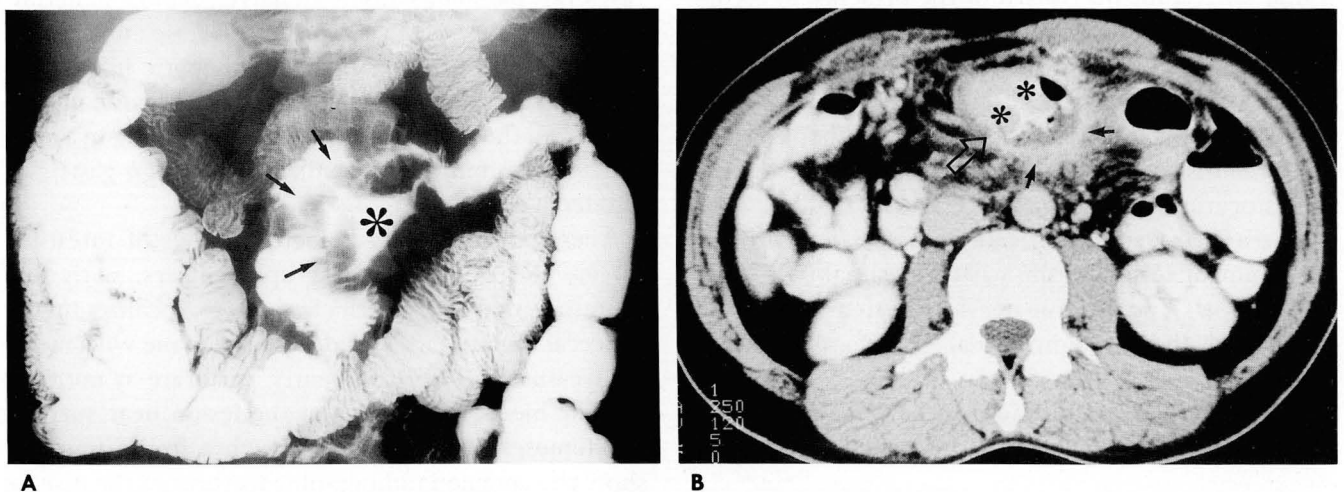


Fig. 2. Recurrent intestinal Behçet's disease at near the surgical anastomosis in a 49-year-old man with a history of right hemicolectomy 1 year ago due to the same disease at the ileocecal region.
A. Small bowel radiograph shows a large deep ulcer (asterisk) in the ileum proximal to ileocolic anastomosis 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.

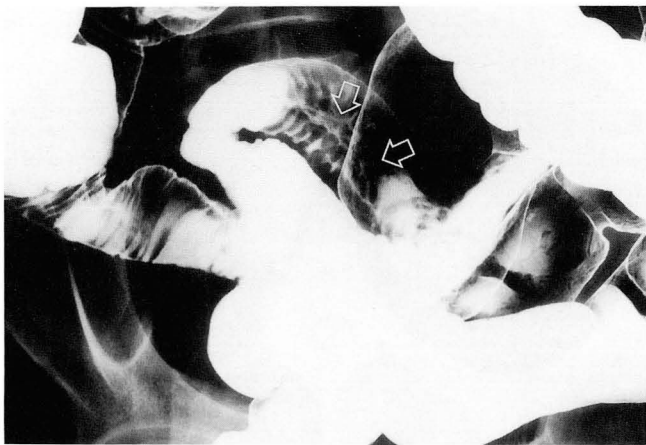


Fig. 3. Intestinal Behçet's disease involving the ileum in a 42-year-old man who had oral ulcer, genital ulcer, perianal ulcers, and positive pathergy test. Double contrast radiograph shows healed ulcer (arrows) with converging mucosal folds

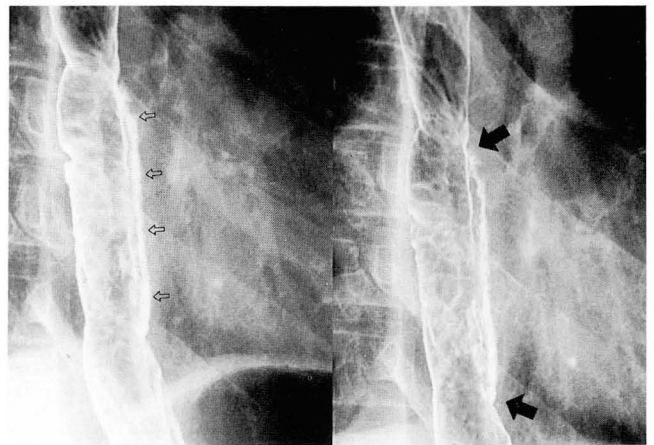
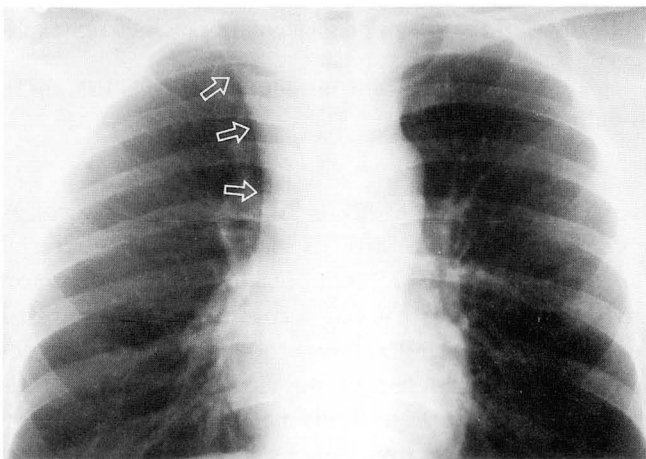


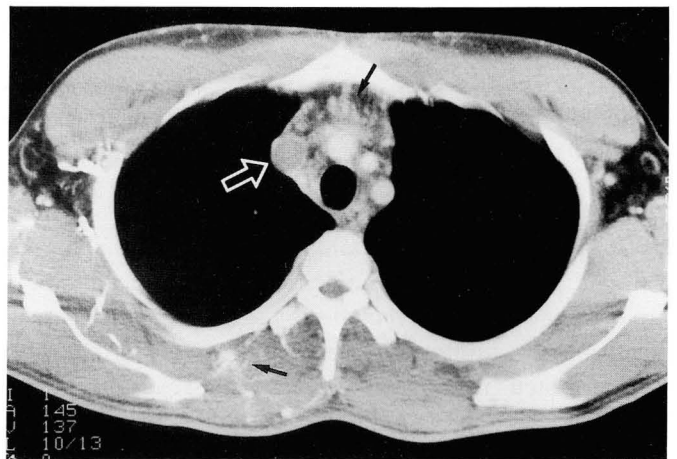
Fig. 4. Intestinal Behçet's disease involving the esophagus in 33-year-old man who had oral ulcers, genital ulcers, skin lesion, and uveitis. Double contrast radiograph shows longitudinal ulcer (open arrows) during healing and retraction of esophageal wall (solid arrows).



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.



A



B

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.

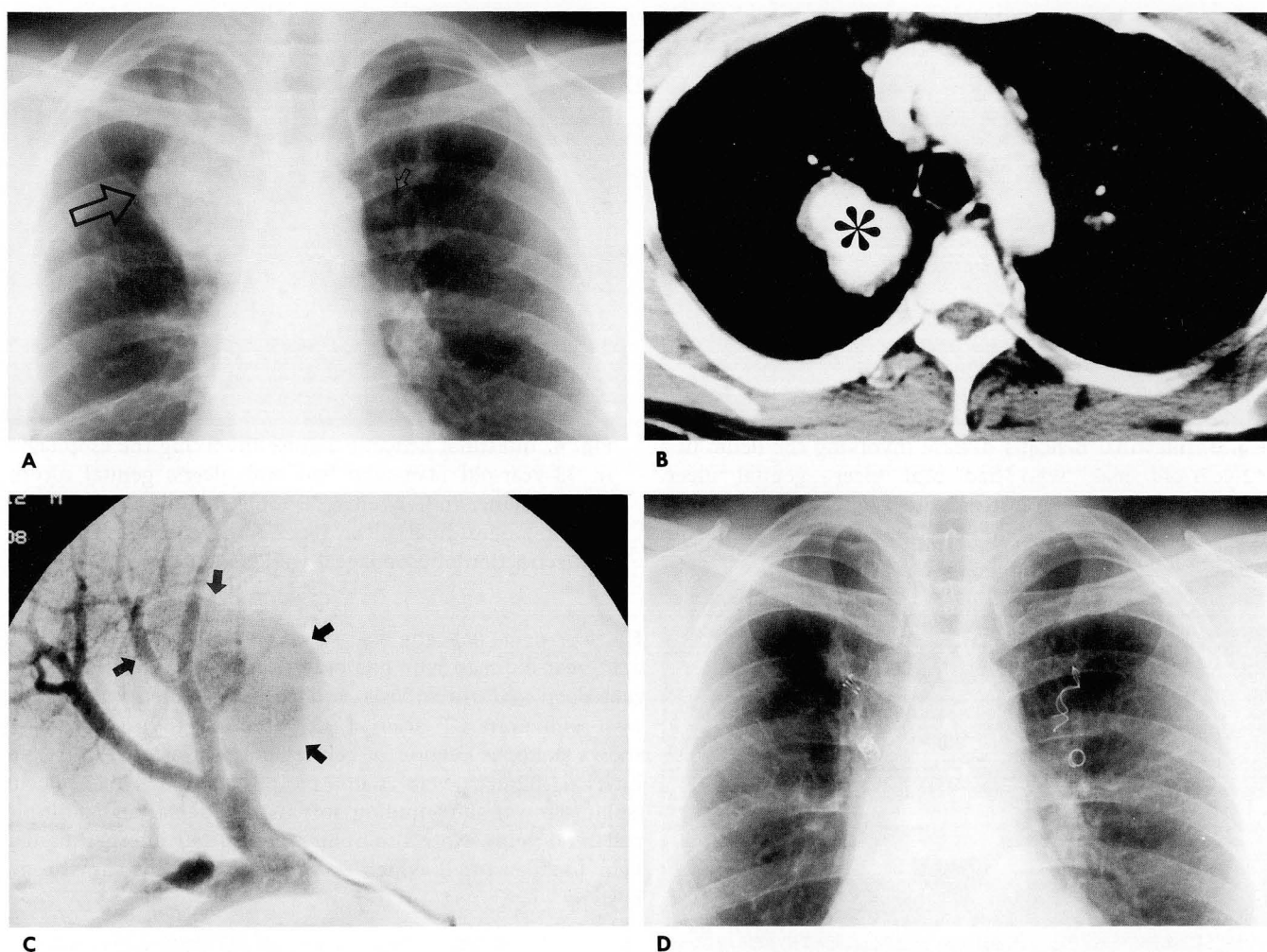


Fig. 7. Aneurysms of the right and left pulmonary arteries in a 38-year-old man with massive hemoptysis. He had a history of recurrent oral ulcers, genital ulcers, and deep vein thrombosis in right lower extremity.
A. Initial posteroanterior chest radiograph shows a mass (large arrow) in the right upper 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 an 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 haustrations 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-

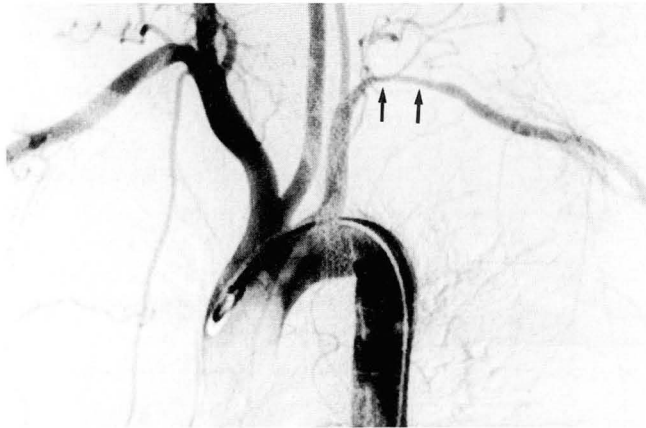


Fig. 8. Angio-behçet's disease involving the left subclavian artery in a 24-year-old woman who had oral ulcers, uveitis, genital ulcers, and positive pathergy test. Digital subtraction arch aortogram shows segmental luminal narrowing of the subclavian artery (arrows). This finding is very similar to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

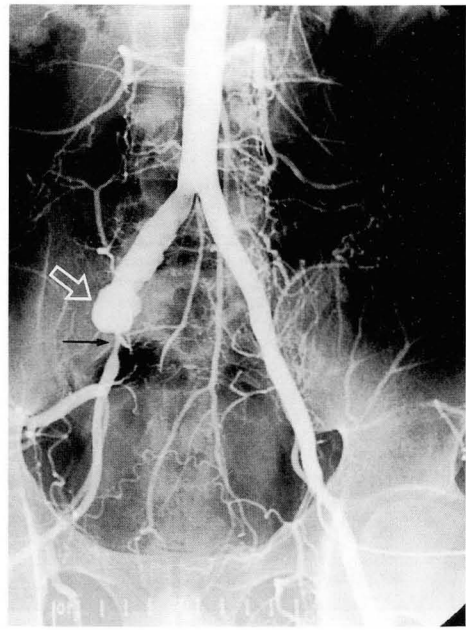


Fig. 9. Angio-Behçet's disease involving the right common iliac artery (RCIA) in a 24-year-old man who had genital ulcers, skin lesion, and epididymitis. Distal abdominal aortogram shows complete occlusion of right external iliac artery, irregular luminal dilatation of RCIA, and an aneurysm (open arrow)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

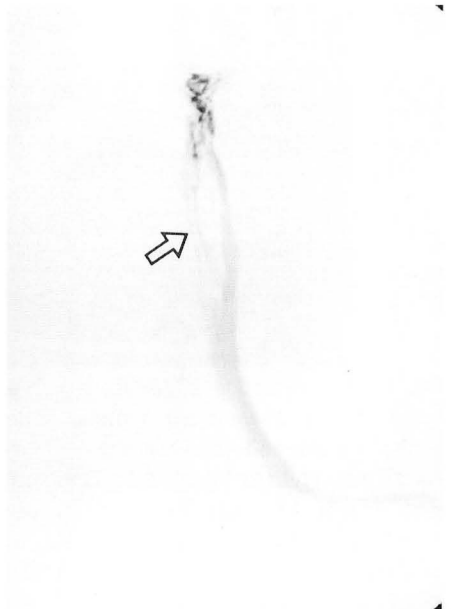
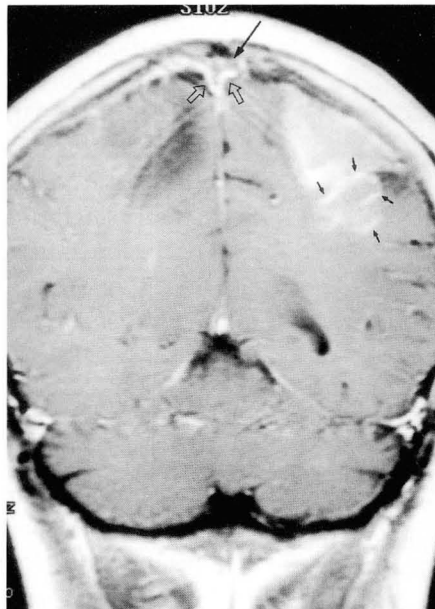
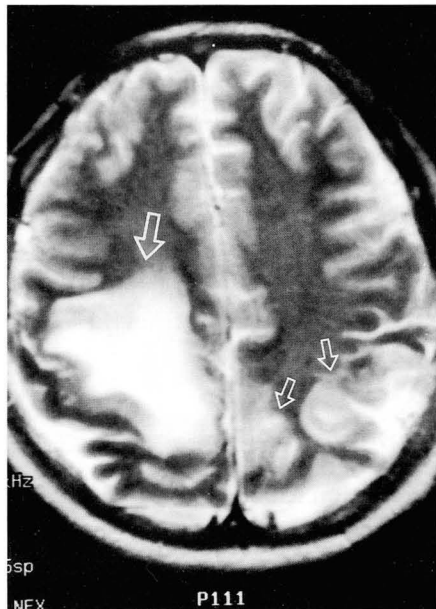
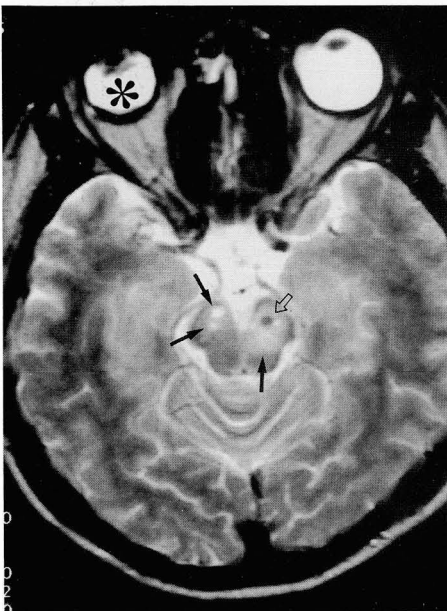


Fig. 10. Central venous thrombosis involving the superior sagittal sinus (SSS) with bilateral cerebral venous infarction in a 40-year-old woman who had oral ulcers, genital ulcers, uveitis, and positive pathergy test.

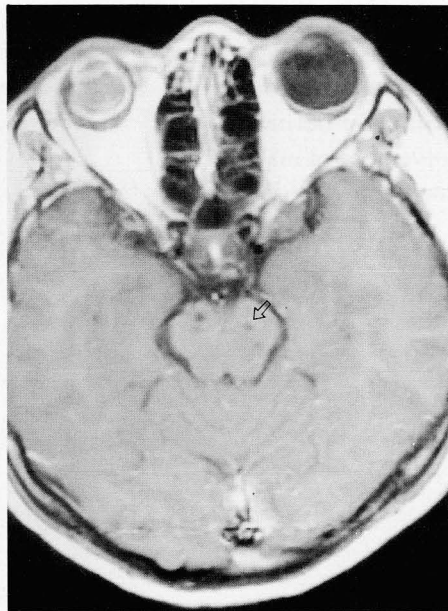
A. Axial T2-weighted (3500/96) image shows high signal intensity in bilateral parietal areas (arrows). The hyperintensities are not confined to arterial territories.

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.



A



B

Fig. 11. Typical neuro-Behçet's disease involving the central nervous system in a 35-year-old man with right side visual loss due to chronic recurrent uveitis.

A. Axial T2-weighted (3800/90) image shows multiple foci of high signal intensity (solid arrows) and a small hypointense intensity (open arrow) in ventral aspect of mid-brain. Note also the small and deformed right globe that is phthisis bulbi (asterisk).

B. Corresponding contrast enhanced T1-weighted (540/14) image shows foci of subtle nodular enhancement and a focus of hypointense (open arrow). The hypointense lesion is suggestive of focal hemorrhage.

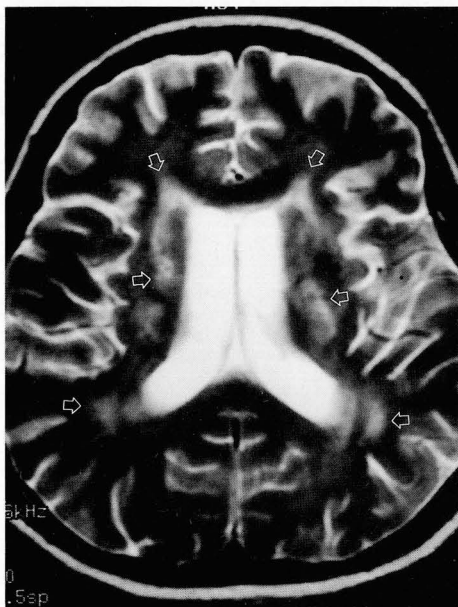


Fig. 12. Neuro-Behçet's disease involving central nervous system in a 27-year-old woman who had oral ulcers, genital ulcers, and skin lesions.

Axial T2-weighted (3500/96) image shows multiple foci of high signal intensity (arrows) in the bilateral periventricular white matters and mild atrophy of periventricular white matters with ventricular dilatation.

tinal widening, or mediastinal, or lung mass, corresponding to occlusion of the superior vena cava (Fig. 6), great vessel aneurysm, or aneurysm of the pulmonary artery.

Arterial involvement in BD occurs at the late stage of the disease, usually in young men, and is seen in only about 5–10% of all cases. The artery most often affected

is the aorta, followed by the pulmonary, femoral, subclavian, and popliteal (4). Although nearly every major artery can be affected by BD, a large artery is most commonly involved, and a visceral artery, rarely. Involvement of pulmonary artery involvement is a unique manifestation of BD, and is to date the only vasculitis known to lead to aneurysm of this artery. Aneurysm formation, especially in the pulmonary artery (Fig. 7), is a serious condition associated with fatal hemorrhage, and may be a leading cause of death in young BD patients. The pathogenesis of arterial aneurysm is thought to be obliterative endarteritis of the vasa vasorum, with resulting dilatation and aneurysm formation. The arteriographic findings of occlusion may be confused with those of pulseless disease (Fig. 8) or Buerger disease, though the associated aneurysm often helps to distinguish BD from these diseases (Fig. 9).

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 meningoencephalitis (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.

References

1. International Study Group for Behçet's disease. Criteria for diagnosis of Behçet's disease. *Lancet* 1990; 335: 1078-1080
2. Rosenberger A, Adler OB, Haim S. Radiologic aspects of Behçet's disease. *Radiology* 1982; 144: 261-264
3. Kim JC, Kim YG, Kim SJ, Choi BI, Park JH. Radiologic study on differential diagnosis of intestinal tuberculosis and intestinal Behçet disease. *J Korean Radiol Soc* 1986; 22: 111-118
4. Numan F, Islak C, Berkmen T, Tuzun H, cokyuksel O. Behçet disease: pulmonary arterial involvement in 15 cases. *Radiology* 1994; 192: 465-468
5. Wechsler B, Dell'Isola B, Vidaihet M, et al. MRI in 31 patients with Behçet's disease and neurologic involvement: prospective study with clinical correlation. *J Neurol Neurosurg Psychiatry* 1993; 56: 793-798

대한방사선의학회지 1998; 38: 33-39

Behçet병의 3 특수 상황에서의 영상 소견¹

¹울산대학교 의과대학 서울중앙병원 진단방사선과학교실

황재철 · 하현권 · 이정현 · 오용호

Behçet병은 주로 침범하는 장기에 따라 분류할 수 있는데, 위장관계, 큰 혈관, 그리고 중추신경계를 침범하는 경우 각각 위장관-, 혈관-, 그리고 신경- Behçet병으로 불리어진다. 이러한 3가지의 Behçet병은 높은 유병율과 사망율과 많은 관련이 있으며 방사선학적 영상이 이러한 상황을 발견하는데 중요한 역할을 한다. 이 임상 화보의 목적은 위장관계, 큰 혈관, 그리고 중추신경계를 침범하는 Behçet병의 영상을 보여주고 감별 진단에 대하여 토의하는데 있다.

1998년도 대한방사선의학회 중요행사 일정 안내(Ⅲ)

대 회 명	일 정		제출처/개최장소
	내 용	마감일 / 일정	
신경·두경부 방사선과학 연구회	학술월례모임	98. 1. 19(월) 18:30 -	서울대학병원
		2. 16(월) "	강남성모병원
		4. 20(월) "	서울중앙병원
		5. 18(월) "	신촌세브란스
		7. 20(월) "	삼성의료원
		9. 21(월) "	강남성모병원
		10. 19(월) "	서울대학병원
		11. 16(월) "	서울중앙병원
	Subspecialty Imaging Conference	98. 3. 18(수) 18:00 -	삼성의료원
	제 12 회 학술대회(예정)	98. 6. 27(토) 09:00 -	고려대학병원
	Subspecialty Imaging Conference	98. 12. 16(수) 18:00	삼성의료원
근골격방사선과학 연구회	Symposium	98. 5. 9(토)	삼성의료원
유방방사선과학 연구회	Symposium	98. 3. 22(일)	미정