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Neurologic Involvement in Behcet Disease: Imaging Findings in 16 Patients

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Behcet disease is a rare multisystem immune-related vasculitis that is prevalent in Japan, the Middle East, and many Mediterranean countries. In our study population of 36 patients with Behcet disease, 16 (44%) had CNS involvement. CT was noncontributory, except in a single patient with dural sinus thrombosis. MR imaging in nine patients showed foci of high signal intensity on T2-weighted images within the brainstem, the basal ganglia, and the cerebral hemispheres in that order of frequency. Unlike the plaques associated with multiple sclerosis, Behcet lesions show no predilection for the periventricular regions.

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Neurologic manifestations of Behcet disease (neuro-Behcet) have been reported in 10-25% of patients. These are nonspecific and include loss of vision, diplopia, nystagmus, cranial nerve palsies, speech disorders, cerebellar signs, and cerebral and spinal sensory and motor disturbances. In general, three patterns of neurologic manifestations have been observed: a brainstem syndrome, a meningoencephalitic syndrome, and an organic confusion syndrome [1].

CT is of little diagnostic help in patients with neuro-Behcet disease. In most cases the CT findings are normal, equivocal, or nonspecific. In a few cases, CT may reveal small foci of low attenuation in the brainstem, basal ganglia, thalami, or cerebral hemispheres [2, 3]. These foci may show homogeneous or patchy enhancement following injection with contrast medium. The more chronic cases are associated with atrophy, especially of the brainstem [4, 5]. Rarely, the disease may manifest as a mass lesion simulating an intraaxial neoplasm [6].

There have been a few isolated reports of MR findings in neuro-Behcet disease [3, 6]. We describe our observations in 16 patients with this disorder and discuss the radiologic differential diagnosis.

Patients and Methods

During the past 10 years, 36 cases of Behcet disease were seen at a major tertiary referral center in the Kingdom of Saudi Arabia. The age and sex distribution of these patients are shown in Fig. 1. The mean age was 28.2 years. The youngest patient was a boy 15 years old, and the oldest, a woman of 54 years. There were 29 males and seven females; a sex ratio of 4:1 in favor of males. The frequency of disease manifestations is shown in Fig. 2. Involvement of the nervous system occurred in 16 patients (44%); of these, 10 (62.5%) had symptoms and signs referable to the brainstem, two (12.5%) presented with meningoencephalitis, two with pseudotumor cerebri, and two had spinal cord symptoms. The high rate of occurrence of neurologic manifestations is attributable to the referral pattern of our hospital. A total of 21 CT scans, nine MR scans, and four cerebral angiograms were performed on these patients; the type of tests depended on whether the patient was seen before or after the availability of MR in our institution, and up to six CT scans were obtained on some of the patients at different intervals.

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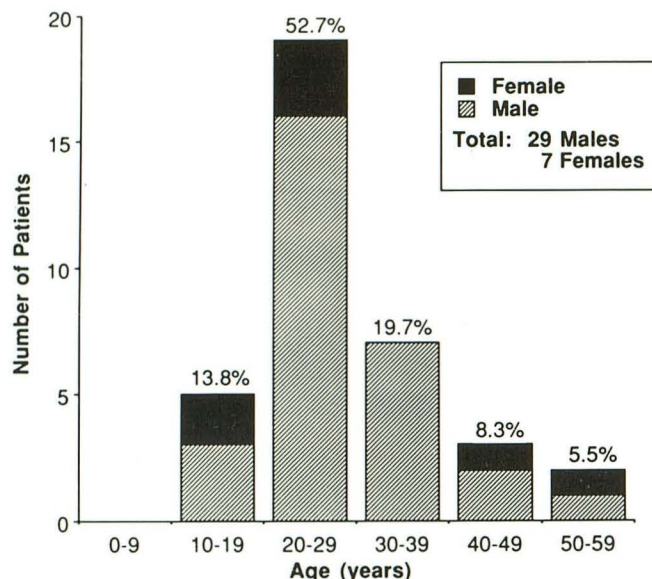


Fig. 1.—Age and sex distribution of 36 patients with Behcet disease who constituted study population.

MR studies were performed on a 1.5-T Vista scanner (Picker, Inc., Solon, OH). The scanning parameters were as follows: 600/20/2 (TR/TE excitations), 2000/40–80/2, and a 7-mm slice thickness. Both T1- and T2-weighted scans were obtained in the axial plane first, and this was supplemented by scans in the sagittal and/or coronal planes depending on the findings. The additional scans were obtained by using a T2-weighted sequence and a slice thickness of 4 mm.

Results

Of the 21 CT scans, only two showed equivocal findings suggestive of brainstem atrophy. Of the four cerebral angiograms, two showed conclusive evidence of dural venous sinus thrombosis. The findings on MR were abnormal in nine pa-

tients in whom there was clinical evidence of CNS involvement. All showed abnormal high-signal-intensity foci on T2-weighted images. On the T1-weighted images (600/20), only two lesions 3–5 mm in size were seen as areas of low signal intensity, but smaller lesions that were evident on T2 were not apparent on T1. On the long TR dual-echo pulse sequence, the lesions were seen on both the early and late echoes. Although the late echo (2000/80) gave more tissue contrast, small lesions in the vicinity of the CSF spaces were detected more readily on the early T2 (2000/40) proton-density images. On these, Behcet lesions had higher signal intensity than CSF. Selected examples of the lesions are shown in Figures 3 and 4. The distribution is summarized in Table 1. Following treatment with corticosteroids alone or in combination with immunosuppressive drugs, the lesions decreased in size; in some patients, they disappeared (Fig. 5). Dural venous sinus thrombosis occurred in two patients. The findings on MR were definite in one of these cases and probable in the other (Fig. 6). MR studies in five additional patients with no neurologic deficit were normal.

Discussion

Behcet disease was first described by Hulusi Behcet (1889–1948) of Istanbul University. In 1937 Behcet published his observations on two patients with a triple-symptom complex of recurrent oral and genital ulcerations and ocular inflammation, which he attributed to viral infection [7]. During the 50 years or so that followed Behcet's description, two important facts came to light. First, Behcet disease is not a viral infection, but a multisystem immune-related vasculitis characterized by periods of remission and exacerbation with an unpredictable prognosis. Second, involvement of other systems may occur concomitant with, but more often some months or years following, a complete or incomplete triple-symptom complex. This may manifest as erythema nodosum, polyarthritides, thrombophlebitis, arterial occlusions, pulmonary infar-

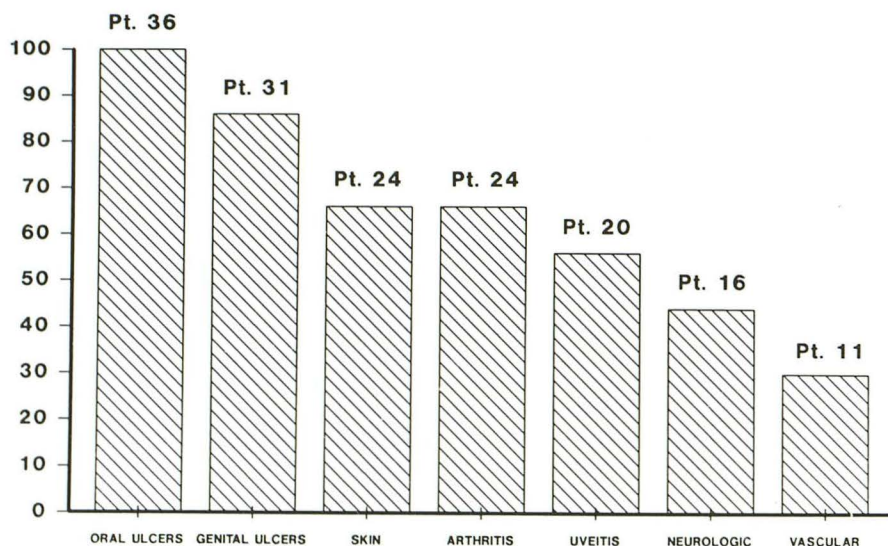
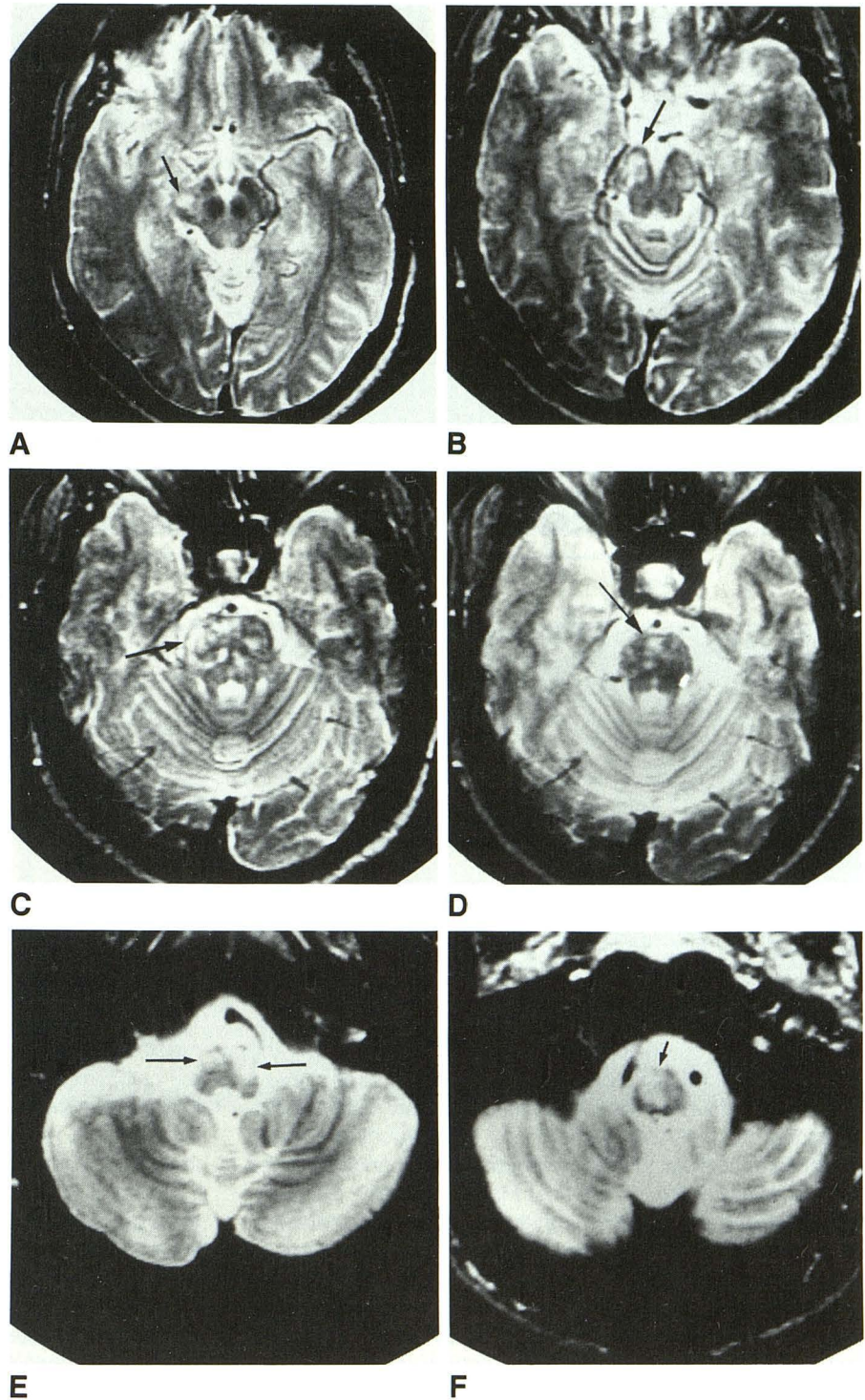


Fig. 2.—Clinical manifestations of 36 patients with Behcet disease who constituted study population.

Fig. 3.—A–F, Axial T2-weighted MR images (2000/80) from different patients showing multiple Behcet foci (arrows) within midbrain (A and B), pons (C and D), and medulla (E and F). Note predilection of lesions for ventral portion of brainstem.



tion, ulcerative colitis, portal hypertension, or neurologic, cardiac, or renal complications. One or more systems may be involved in the same patient [7, 8].

The majority of patients with Behcet disease are young adults, but the disease has been reported in neonates [9] and in subjects over 70 years of age. It is two to four times more

common in males. A few familial cases have been recorded [7, 8].

Although Behcet disease is generally rare, it is prevalent in Japan and many Mediterranean and Middle Eastern countries [7–11]. Its incidence in these regions varies from place to place, but in only a few localities does it exceed 10 cases per

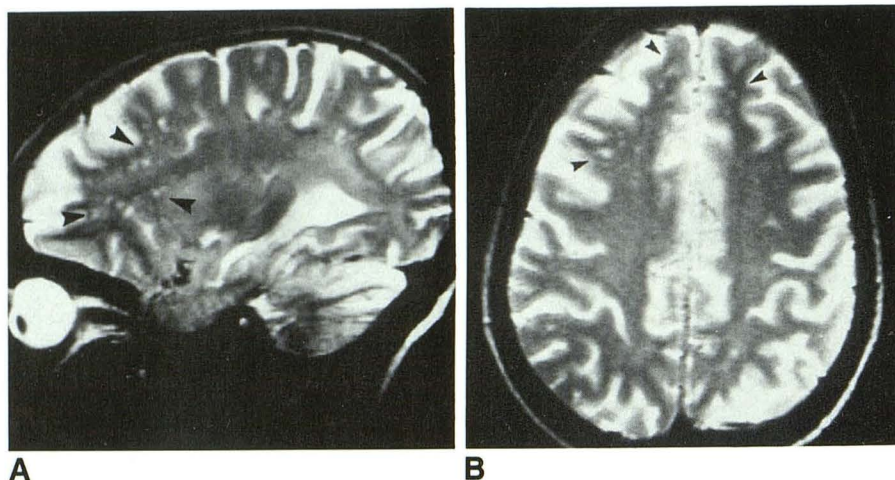


Fig. 4.—A and B, T2-weighted MR images (2000/80) showing Behcet lesions within cerebral hemispheres.

A, Sagittal image shows clusters of Behcet foci within frontal lobe (arrowheads).

B, Axial section above the level of lateral ventricles shows that, unlike the plaques that occur with multiple sclerosis, Behcet lesions (arrowheads) have no predilection for periventricular area.

TABLE 1: Distribution of Behcet Lesions Within the Brain in Nine Patients

Case No.	Brainstem ^a			Thalami	Corpus Striatum		Cerebral Cortex				Cerebellum
	Medulla	Pons	Midbrain		Caudate	Lentiform	Frontal	Parietal	Temporal	Occipital	
1	+	+	+	—	—	—	—	—	—	—	—
2	+	—	—	—	—	—	—	—	+	—	—
3	—	—	+	+	—	—	—	—	—	—	—
4	—	—	—	—	+	—	—	—	—	—	—
5	+	+	—	—	—	—	—	+	—	—	—
6	—	+	—	—	—	—	+	+	—	—	—
7	+	+	+	—	—	+	—	—	—	—	—
8	—	+	+	—	—	—	—	—	—	—	—
9	Dural venous thrombosis										

^a Note frequent involvement of the brainstem.

100,000 of the population [8]. It is a less well known entity in Europe and North America [7, 8].

Despite the lapse of more than half a century since the original description of Behcet disease, it remains an enigmatic condition with regard to origin, geographic distribution, and clinical course. Pathologic studies on autopsy cases are few and came principally from Japan. The basic brain lesion, according to Japanese researchers, is a chronic relapsing inflammatory cellular infiltration around venules and capillaries and occasionally around arteries. Macroscopically, these appear as foci of brain softening, which affect the brainstem more frequently than elsewhere. As the lesions become more chronic, there is excessive gliosis, atrophy, and in some cases thickening and fibrosis of the meninges [8]. It is believed that the inflammatory reaction is brought about by human leukocyte antigens generated by many factors, which might be environmental, genetic, or otherwise. Venous thrombosis occurs in approximately one third of patients with Behcet disease [12]. The vena cava and the portal vein are the most common sites, and involvement of the dural venous sinuses is rare. Venous thrombosis may be precipitated in some patients by high fibrinogen levels, but in many cases the exact mechanism is unknown [12].

The MR findings in neuro-Behcet disease consist of small foci of high signal intensity on T2-weighted images, which are iso- or hypointense relative to brain on T1-weighted images.

The lesions may be circular, linear, crescentic, or irregular, and occur most frequently within the brainstem, especially around the cerebral peduncles and in the pons. The thalamus and basal ganglia are second in frequency and similar foci may be seen in the cerebral hemispheres, the spinal cord, or the cerebellum. They most likely represent inflammatory cellular infiltration, demyelination, and edema, which may diminish in size in response to corticosteroids alone or in combination with immunosuppressive drugs.

Cerebral angiography of patients with neuro-Behcet disease usually shows no evidence of arteritis and most abnormalities are attributed to venous thrombosis. However, at least one case has been recorded in which cerebral angiography revealed widespread areas of stenosis and ectasia of the anterior middle cerebral arteries [12].

In the absence of a clear history of genital or oral ulcers, the diagnosis of neuro-Behcet disease could be extremely difficult. From a radiologic viewpoint, the differential diagnosis includes four main entities: multiple sclerosis, brainstem infarction, rhomboencephalitis, and dilated perivascular spaces.

Multiple sclerosis and neuro-Behcet disease have many features in common. They are both disseminated, demyelinating, and inflammatory diseases of unknown origin, but clear-cut cases can be differentiated on the basis of clinical history and serum and CSF findings [14]. Multiple sclerosis is a disease of white matter, and these plaques occur more

Fig. 5.—T2-weighted MR images (2000/80) showing response of Behcet lesions to corticosteroids.

A, Axial image shows marked involvement of midbrain in region of left cerebral peduncle and thalamus (arrow).

B, Corresponding section after 3 weeks of treatment shows almost complete resolution of the lesion.

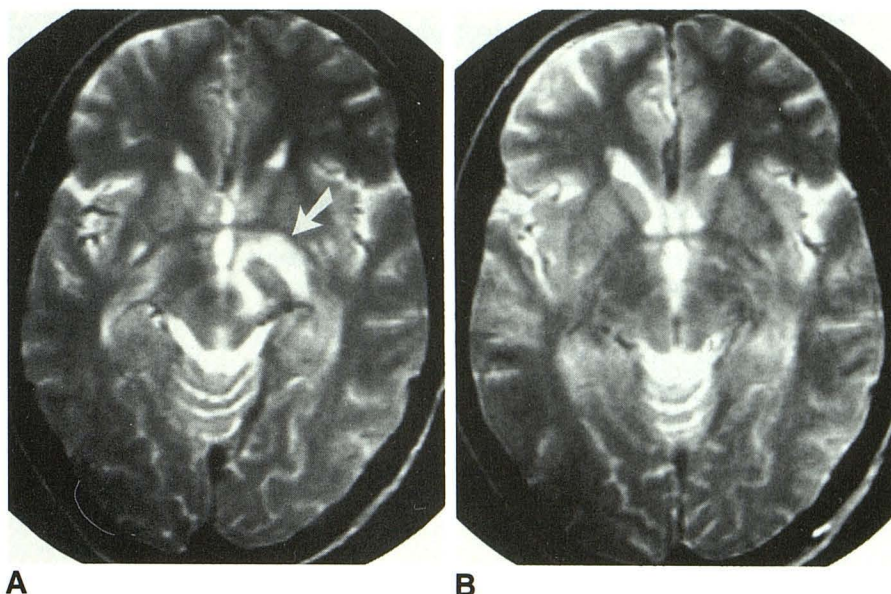
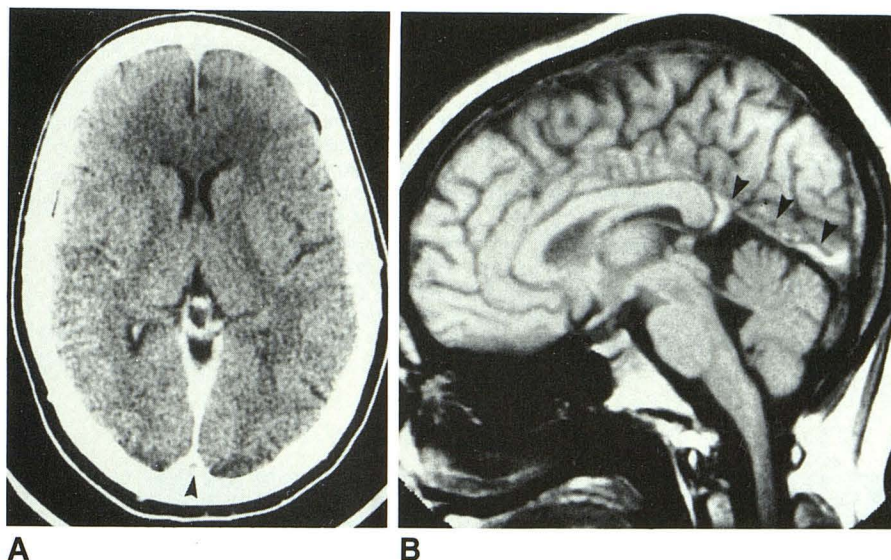


Fig. 6.—CT and MR images showing dural venous sinus thrombosis in Behcet disease.

A, Axial contrast-enhanced CT scan shows filling defect within confluence of dural sinuses (arrowhead).

B, Sagittal T1-weighted MR image (600/20) shows high signal intensity of clotted blood within vein of Galen and straight sinus (arrowheads).



frequently in the periventricular regions. Behcet lesions, on the other hand, may involve the white and gray matter, and are much more common within the brainstem.

Lacunar infarction from arteriosclerotic disease occurs in an older group of patients with evidence of cerebrovascular disease elsewhere. To date, sizable brain infarction due to occlusion of major cerebral arteries has not been reported in Behcet disease.

Rhombencephalitis is a rare entity in which there is acute inflammation, which for no known reason is almost always limited to the brainstem. It appears as areas of high signal intensity on T2-weighted images and can be differentiated from Behcet disease by the clinical history and the CSF findings [15, 16].

Dilatation of perivascular spaces is an anatomic variant, which may be seen in the brainstem as punctate high-density

foci in the substantia nigra and the cerebral peduncles. These may be unilateral or bilateral and they most likely represent perivascular spaces around penetrating branches of the collicular arteries [17].

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