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Neuro radiology

Brain stem type neuro-Behçet's syndrome

Correlation of enhanced CT scans and MRI during the acute and chronic stage of the illness

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Summary. Brain stem type neuro-Behçet's syndrome was studied with enhanced CT and MRI during the acute and chronic stage of the illness. During the acute stage, brain CT revealed a low density lesion in the brain stem extending from the lower pons up to the midbrain ventrolaterally with marginal enhancement effect. T2-weighted image showed a high signal intensity lesion in the brain stem which mainly involved the basis ponti, tegmentum, tectum and cerebral peduncle. During the chronic stage, the lesion became low in signal intensity with T2-weighted image and reduced in its size without enhancement in brain CT. The prolonged relaxation time of the lesions was gradually normalized with steroid treatment. Sequential brain CT with enhancement and MRI study with T1- and T2-weighted images were useful to detect the lesions and to evaluate the activity in the neuro-Behçet's syndrome.

Key words: Neuro-Behçet's syndrome - Behçet's syndrome - Brain stem type neuro-Behçet's syndrome - Magnetic resonance imaging

Neurological involvement is estimated at 10–20% in Behçet's syndrome and these cases have been designated as neuro-Behçet's syndrome [1]. Neuropathological investigations revealed that the basic nature of this syndrome is recurrent meningoencephalitis or encephalomyelitis with or without the brain stem involvement [2, 3]. This neurological syndrome is usually classified into the following three types according to Pallis and Fudge [4]. (1) Brain stem syndrome, (2) Meningomyelitic syndrome, (3) Organic confusional syndrome or dementia. The recent development of neurological techniques has made it possible to demonstrate lesions in this syndrome. In this report we describe two cases of brain stem type neuro-Behçet's syndrome, analyze the findings by sequential brain CT and MRI, and correlate these radiological changes with brain stem neurological signs.

Case report

Case 1

A 41-year-old house wife was admitted on December 7, 1983 because of headache, fever, diplopia and progressive left side weakness over 5 days duration. She had a history of recurrent stomatitis and genital ulcers over the past several years. On January 17,

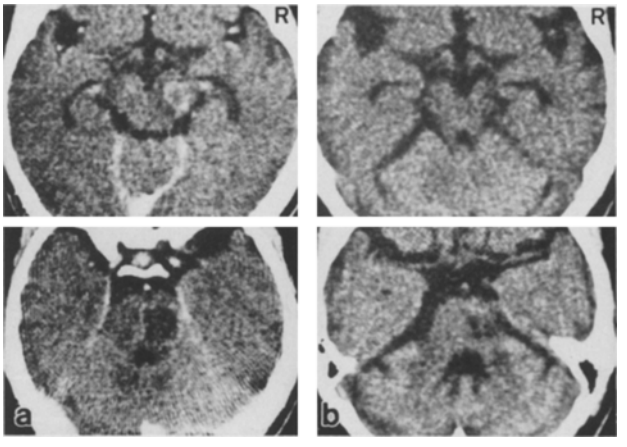

Fig. 1 a, b. Enhanced CT scans of case 1. The left row (**a**) shows enhanced CT 10 days after onset of clinical symptoms. The right row (**b**) shows enhanced CT about 11 months after onset of clinical symptom. **a** A unilateral low density area with marginal enhancement effect is noted in the brain stem (lower pontine base-midbrain cerebral peduncle) and the pontine swelling is significant. The structures involved in this case are pontine base, pontine tegmentum and cerebral peduncle in the right side. **b** A small low density area is seen in the right brain stem (lower pons up to cerebral peduncle). There was no definite enhancement around the low density area. The brain stem is atrophic with widened cisterns

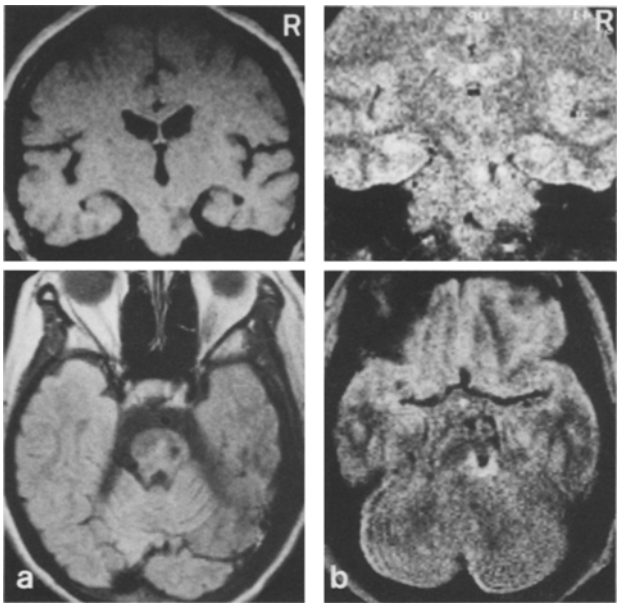

Fig. 2a, b. MRI of case 1 17 months after the onset of clinical symptoms. The left row (**a**) shows T1-weighted images (TR = 600 ms, TE = 35 ms). The right row (**b**) shows T2-weighted images (TR = 1600 ms, TE = 70 ms). **a** A low signal lesion is seen in the right pons. **b** A low signal lesion with marginal high signal area is noted in the right pons, suggesting a liquefying necrosis with surrounding gliosis

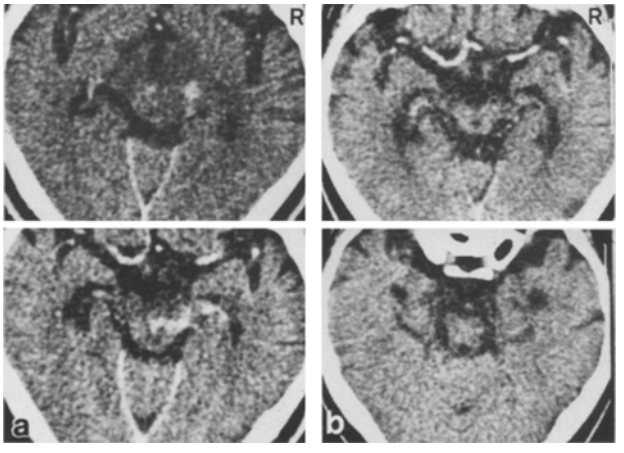

Fig. 3a, b. Enhanced CT scans of case 2. The left row (**a**) shows enhanced CT 5 days after onset of clinical symptoms. The right row (**b**) shows enhanced CT about 4 months after onset of clinical symptoms. **a** A low density area with irregular marginal enhanced effect is noted in the upper pons, midbrain and thalamus. Swelling of low density area is noted in the midbrain and thalamus and third ventricle is compressed by the right side thalamus. Involved structures are cerebral peduncle, midbrain tectum, periaqueductal gray matter, and hypothalamus. **b** Small low density areas are seen in the midbrain cerebral peduncle. There was a marked atrophy of the brain stem from the upper pons to midbrain

1983, she had sudden onset of acute uveitis of the right eye and erythema nodosum in the lower extremities. She was diagnosed as having Behçet's syndrome by the clinical findings and skin biopsy. She had been suffering from recurrent arthritis with fever since April 1983. Family history was non-contributory. Physical examinations on admission revealed a low grade fever (37.2° C), multiple scars in the oral cavity and swollen knee joints. There was neither genital ulcers nor skin lesions.

Neurological examination revealed that she was alert and oriented well in time, place, and person. She was dysarthric with slow speech. Mild nuchal rigidity was noted. The visual acuity was 20/20 in both eyes and her discs were clear. The pupils were equal and promptly reactive to light. The vertical eye movement was limited with up-beating nystagmus on upward gaze. She had a horizontal gaze palsy to the right. In an attempt to look to the left, her right eye did not adduct with full abduction of the left eye. She had a left central facial palsy. Hearing was intact bilaterally. In addition to the left hemiparesis, ipsilateral sensory disturbance involved facial area for superficial and deep sensation. The left side deep tendon reflexes were more brisk than the right side. Babinski's sign was positive on the left side and equivocal on the right side. There were no cerebellar signs.

Laboratory data. The ESR was 104 mm/h. CRP was +6. There was mild leukocytosis (11570/mm^3). Serum C3 was 134 mg/dl, and C4 was 78.9 mg/dl. Serum fibrinogen was 498 mg/dl. LE test and RA factor were negative and serum TPHA and FTA-ABS test were negative. Lumbar puncture revealed marked pleocytosis (165 polynuclear cells, 110 mononuclear cells/mm^3) and CSF protein was 45.5 mg/dl and myelin basic protein was elevated to 29 mg/ml. The cultures of CSF were negative. Viral titers of the serum and cerebrospinal fluid were unrevealing.

Clinical course. The right sided one-and-a-half syndrome persisted with intermittent high fever (39–40° C). The pulse therapy of methylprednisolon 1000 mg/day for 3 days was given, followed by dexamethason therapy (20 mg/day for 4 days, 16 mg/day for 4 days, 12 mg/day for 4 days, and 8 mg/day for 4 days) and a small dose of prednisolon therapy thereafter. After corticosteroid therapies febrile episodes disappeared within 2 days. The right gaze palsy and MLF syndrome were improved and CSF abnormalities were normalized gradually over 3 weeks.

Brain CT. On the 7th hospital day (acute stage), enhanced CT scans revealed a swollen brain stem and a low density area with peripheral enhancement from the right lower basis ponti which extended ros-

trally up to the right cerebral peduncle (Fig. 1 a). The low density area in the brain stem was concentrically reduced in its size with steroid therapy and the swelling of the brain stem improved within 2 months. About 11 months after admission (chronic stage), brain CT scans showed a mild atrophy of the brain stem and a smaller low density lesion in the right brain stem from the basis ponti up to the cerebral peduncle rostrally. The peripheral ring enhancement of the lesion was noted by the 10 months after the onset, but thereafter there was no definite marginal enhancement effect around the low density area (Fig. 1 b).

MRI. 18 months after the onset (chronic stage), T1- and T2-weighted images (TR = 600 ms, TE = 35 ms and TR = 1600 ms, TE = 70 ms) showed a small low signal area in the white matter of the right pons (Fig. 2). T1 value of the lesion was 1257.2 ms (the mean ± SD of normal white matter: 722 ± 90.8 ms). T2 value of the same lesion was 85.9 ms (the mean ± SD of normal white matter: 77.0 ± 5.3 ms). T1 and T2 values of the peripheral area were more prolonged than the central area. And there were no significant changes of these values and neurological findings during one year after admission (Fig. 5 a).

Case 2

First admission. A 31-year-old woman was admitted on June 23, 1984 with progressive right sided weakness. There was a history of recurrent oral and genital ulcers since 21 years of her age. She was diagnosed as having Behçet's syndrome in January 1974 by skin biopsy.

Neurological examinations on admission disclosed a disoriented lady with dementia. There were no meningeal signs. She had a right sided hemiparesis and hemisensory loss with facial involvement. Deep tendon reflexes were exaggerated on the right with Babinski's sign. Her jaw jerk and finger jerks were exaggerated bilaterally with right sided preponderance.

Lumbar puncture revealed a moderate pleocytosis (12 polynuclear cells and 9 mononuclear cells/mm^3) with normal protein and sugar. Brain CT revealed a small low density area in the posterior limb of internal capsule without enhancement. These neurological findings improved with prednisolon therapy of 50 mg/day except for her dementia. Four months after discharge, her neurological disability progressed and she was sent to a local nursing home.

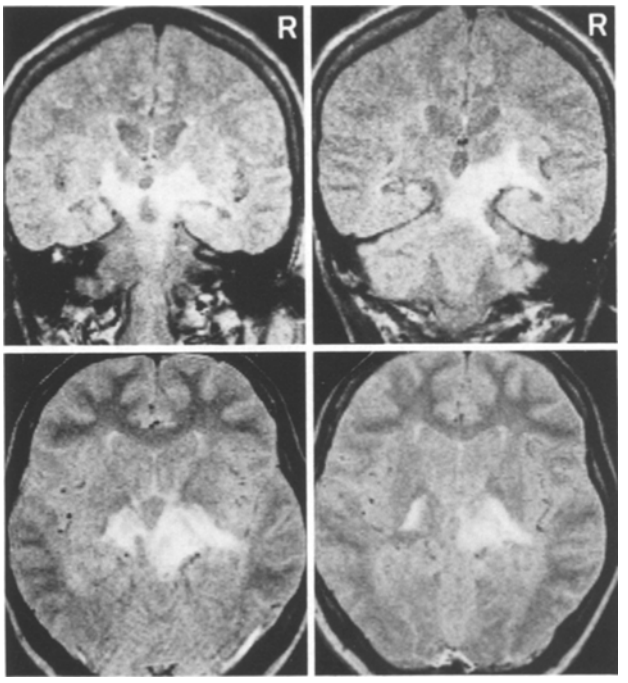

Fig. 4. T2-weighted MRI of case 2, 5 days after the onset of clinical symptoms. An extensive high signal area is seen in the brain stem, bilaterally involving upper pons, cerebral peduncles, thalamus and posterior limbs of internal capsule with right sided predominancy

Second admission. She was transfered to our service from the nursing home for further evaluation and treatment of low grade fever, dysphagia and emaciation on January 13, 1986.

Neurological examinations revealed a drowsy, withdrawn lady with decorticate posture. Mild nuchal rigidity was noted. The right pupil was 5 mm and the left was 4 mm in diameter. The light reaction was absent on the right and sluggish on the left. Optic atrophy was noted on the right side. Gaze to the right was full, to the left, the left eye was abducted with coarse nystagmus and the right eye was fixed with no adduction. Incomplete upward gaze palsy was seen. There was quadriparesis with marked bilateral rigospasticity and Babinski's signs. Exaggerated jaw jerk and finger jerks were seen bilaterally.

Cerebrospinal fluid examinations revealed moderate pleocytosis (15 polynuclear cells and 1 mononuclear cell/mm^3) with normal sugar and protein.

Clinical course. Inspite of extensive steroid therapy with methylprednisolon, there was no remarkable improvement in her neurological condition except for her returned pupillary light reaction.

Brain CT. On the second admission, enhanced CT demonstrated the involvement of the right cerebral

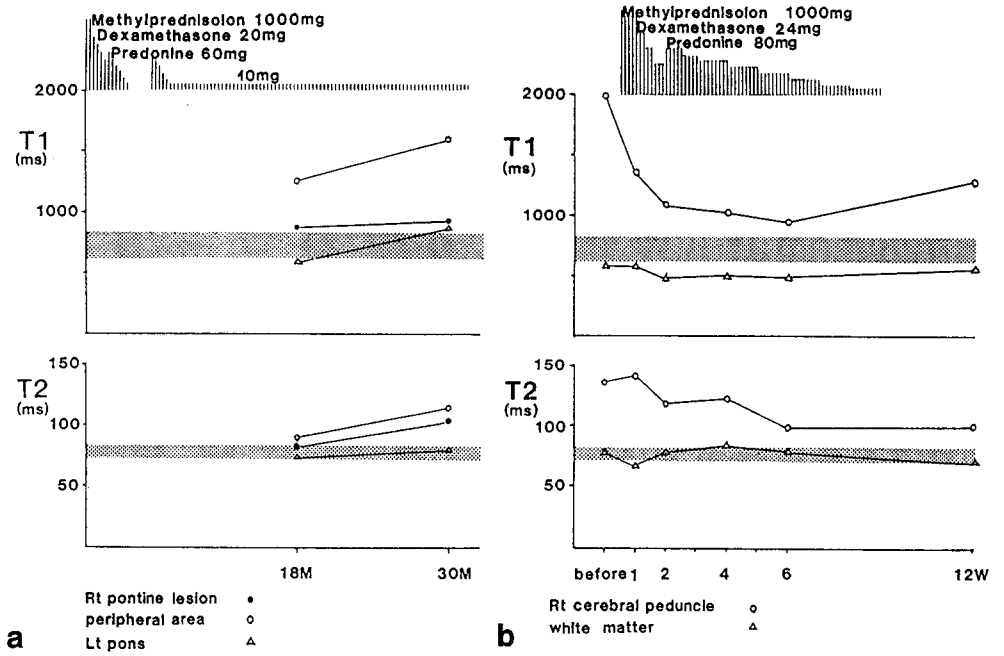

Fig. 5 a, b. Sequential changes of relaxation times. **a** Case 1 (chronic stage): T1 and T2 values are mildly prolonged in the peripheral area. **b** Case 2 (acute stage): T1 and T2 values are moderately prolonged initially and then gradually decreased by steroid therapy

peduncle, tectum, periaqueductal gray matter in the midbrain, thalamus and posterior limb of internal capsule. Irregular marginal enhancements were seen in the cerebral peduncle, tectum and thalamus (Fig. 3 a).

About 4 months after the admission, brain CT showed atrophy of the brain stem from the upper pons up to the midbrain and multiple low density areas in the right cerebral peduncle and the medial thalamus (Fig. 3 b). These marginal enhanced effects disappeared within one month and the swelling of brain stem disappeared within 2 months.

MRI. On the second admission, T2-weighted image (TR = 1600 ms, TE = 90 ms) revealed a high signal area extending from the upper pons to the diencephalon, clearly demonstrating the widespread involvement of the brain stem, both cerebral peduncles, superior cerebellar peduncle, midbrain tectum, and posterior limb of internal capsule with right sided preponderance in the coronal section image (Fig. 4). The high signal lesion with T2-weighted image reduced in its size during steroid treatment. The relaxation time of the right cerebral peduncle was prolonged moderately during the acute stage and was normalized gradually (Fig. 5 b). No definite abnormal signals were detected with T1-weighted image (TR = 600 ms, TE = 45 ms) during the acute stage. But about 4 months after the second admission, a small low signal lesion was detected with T1- and T2-weighted image in the midbrain. The relaxation time of the lesion was more prolonged than the value of normal white matter after treatment.

Discussion

Brain stem type neuro-Behçet's syndrome with acute and subacute brain stem lesion was sequentially evaluated by enhanced CT scans and MRI. In our study, brain stem lesions were well documented by MRI as well as enhanced CT scans during the acute and chronic stage of the illness. Although there have been several reports of neuro-Behçet's syndrome with neuroradiological investigations previously [5–7], it was difficult to detect or evaluate the brain stem lesion with serial CT examination. MRI, especially T2-weighted images, have been known to be useful to detect active lesions of the brain stem in demyelinating [8, 9] or ischemic disorders [10], as well as in neuro-Behçet's syndrome. Recently MRI has proved to be a sensitive method for detecting lesions, and to be important for the early detection of neuro-Behçet's syndrome [11–14]. In our study, coronal T2-weighted MRI revealed more extensive lesions than enhanced CT scans in the acute stage of the illness. MRI is much superior to CT in the posterior fossa because of the absence of bone artifact.

Sequential enhanced CT scans demonstrated various characteristics of this syndrome. In the acute stage, there was swelling and irregular low density associated with transient marginal enhancement in the brain stem and diencephalon. During the chronic stage, there was atrophy of brain stem, reduced density without enhancement and the enlarged third and fourth ventricles due to the surrounding atrophy. Enhancement of the low density

area was seen only during the acute stage or exacerbation, and the effect lasted for several months. The duration of enhancement is probably related to the severity of the inflammatory process in each case.

The MRI images demonstrated several characteristics of this syndrome with sequential examinations. In the acute stage, they showed a rather extensive high signal lesion in the brain stem with T2-weighted images, T1 and T2 values were both prolonged. In the chronic stage, there was a low signal lesion in the T2 weighted image. MRI shows more widespread lesions than CT. The most affected structures in our cases were distributed over the pontine base, pontine tegmentum, cerebral peduncle, superior cerebellar peduncle and posterior limb of internal capsule. This rostro-caudal extension of the lesion in the one side of the brain stem can not be explained by arterial occlusion. In the acute stage of the illness, the active lesions were detected by enhanced CT and T2-weighted MR images. The low density area with mass effect and marginal enhancement by CT scans and a high signal area in T2 images can be explained by the increased free water content at the site of the lesions, secondary to variable degrees of blood-brain-barrier damage from the disease. From the hitherto published neurological findings in this disease [2, 3], multifocal necrotizing lesions with marked inflammatory cell reactions are frequently noted with nonspecific inflammatory changes of vessels in the brain stem, as well as the meninges and cerebrum. The inflammation of vessels, particularly the venous system has been recognized as one of the main features of this disease. So the unilateral lesion, which extended from the lower pons up to the mesencephalon, seen in our cases, could be explained by a disturbance of the venous outflow from the brain stem.

It is suggested that the diagnostic sensitivity of T2-weighted images for brain stem type neuro-Behçet's syndrome is quite high and sequential MRI studies with T1- and T2-weighted images are useful for the detection of lesions in the brain stem, and to evaluate the activity or evolution of lesions in this disease.

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