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. 1a, b. Enhanced CT scans of case I. 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.
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/mm³). 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 polymorphonuclear cells, 110 mononuclear cells/mm³) 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 methylprednisolone 1000 mg/day for 3 days was given, followed by dexamethasone 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 prednisolone 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-