**Neuro-Behçet’s Disease Manifesting as a Neoplasm-Like Lesion**

—Case Report—

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**Abstract**

A 50-year-old man presented with neuro-Behçet’s disease (NBD) manifesting as a large neoplasm-like lesion affecting the brainstem, basal ganglia, and white matter of the cerebral hemisphere. He had no history of disease except for psychoneurosis. On admission, neurological examination found left hemiparesis and dysarthria. Magnetic resonance (MR) imaging showed multiple small ring-like enhancement in the basal ganglia, brainstem, and deep white matter. Biopsy of the mass was performed. Histological examination revealed invasion of inflammatory cells in the white matter, especially around the blood vessels. After the brain biopsy, the patient developed oral aphthae, genital ulcers, and skin eruptions, which are indicative of Behçet’s disease. MR imaging after three courses of steroid pulse therapy revealed that the edematous lesion had become smaller with minimum midline shift. NBD should be considered in the differential diagnosis of lesions with multiple ring-like enhancement extending from the basal ganglia to the brainstem, because dermatological manifestations are sometimes obscured during periods of remission.

Key words: neuro-Behçet’s disease, magnetic resonance imaging, steroid pulse therapy

**Introduction**

Behçet’s disease is a multisystemic, recurrent, inflammatory disorder affecting the eyes, skin, mucosa, joints, vascular system (mainly the veins), lungs, gastrointestinal tract, and nervous system. The syndrome manifests as recurrent oral aphthae, skin symptoms such as nodular erythematous eruptions, genital ulcerations, and ocular symptoms such as uveitis. Neuro-Behçet’s disease (NBD), which comprises 10–20% of all cases of Behçet’s disease, predominantly manifests as central nervous system symptoms, often involving the brainstem, basal ganglia, and the white matter of the cerebrum. However, NBD occasionally manifests as a neoplasm-like lesion with significant mass effect. We describe a case of NBD with a large intracranial mass.

**Case Report**

A 50-year-old man with a 2-week history of left dysstasia and dysbasia was referred to our hospital. He had been treated for psychoneurosis (euphoria) since the age of 28 years. One year prior to admission, he experienced lumbar pain and mild weakness of the left leg, which gradually recovered.

On admission, he had mild impetigo on his body. He was alert. Neurological examination found left hemiparesis including the face, a positive left Babinski reflex, dysarthria, and normal sensation. Laboratory examinations found negative reaction for human leukocyte antigen-B51, levels of sugar, chlorine, immunoglobulin M (IgM), and IgD within the normal ranges, and no oligoclonal band. The leucocyte count was 15,700 10⁶/l and the C-reactive protein level was 3.21 mg/dl. Serum IgG and IgA, CH₅₀, antistreptolysin O, erythrocyte sedimentation rate, and serum copper measurements were all within the normal limits. Other hematological examinations revealed no abnormalities. Computed
Fig. 1 Magnetic resonance images on admission showing a mass-like lesion around right basal ganglia and thalamus. (A) Axial T₁-weighted image, (B) axial T₂-weighted image, (C) axial T₁-weighted image with gadolinium.

Fig. 2 Photomicrograph showing inflammatory infiltration of lymphocytes and small numbers of neutrophils in the Virchow-Robin spaces. Hematoxylin and eosin stain, original magnification ×400.

tomography (CT) showed a low-density area with significant mass effect in the right basal ganglia and thalamus with ring-like enhancement following injection of contrast medium. Magnetic resonance (MR) imaging showed an area of hypointensity on the T₁-weighted image, and an area of mixed intensity (mostly hyperintensity with some iso-hypointensity) on the T₂-weighted image. T₁-weighted MR imaging with gadolinium showed multiple small ring-like enhancement with marked perifocal edema (Fig. 1). Single photon emission CT using thallium showed a slight hot spot within the lesion.

A biopsy of the mass was taken from both the non-enhanced and enhanced areas. Histological examination of both areas revealed large numbers of chronic inflammatory cells mainly in the Virchow-Robin spaces (Fig. 2). Immunohistochemical staining for kappa and lambda light chains and MB-1 showed that the infiltrating lymphocytes were polyclonal. Subsequent lumbar puncture showed an opening pressure of 15 cmH₂O. Examination of the cerebrospinal fluid found 14 leukocytes/dl, chloride level of 125 mmol/l, sugar level of 53 mg/dl, protein level of 70 mg/dl, IgG of 9.0 mg/dl, and IgA of 0.7 mg/dl. Following the biopsy, oral aphthae, genital ulcers, and a skin eruption gradually appeared. The diagnosis was NBD on the basis of the clinical symptoms and histological findings.

The patient was treated with three courses of steroid pulse therapy separated by 2-week intervals. Each course consisted of injection of methylprednisolone sodium succinate 1000 mg/day for 3 days. Then, dexamethasone sodium phosphate was given orally with a dose of 2.0 mg/day for one day, 1.0 mg/day for one day, and 0.5 mg/day for one day. After the therapy, he could walk with a stick because of amelioration of the lower limb paresis, but the upper limb paresis did not improve. MR imaging 3 months after the therapy revealed that the edematous lesion had become smaller, and the midline shift had decreased (Fig. 3). The skin lesions had a recrudescent course. Examination of radiological images 2 years after the biopsy revealed that the brain edema had resolved and the mass had not recurred.

Discussion

A definitive diagnosis can be made in most cases of
NBD on the basis of the clinical symptoms. However, the dermatological symptoms that are characteristic of Behçet’s disease were probably in remission in our patient at admission. In addition, our patient had a large intracranial lesion with significant mass effect. The typical MR imaging appearance of NBD is areas of hypointensity on T₁-weighted imaging and hyperintensity on T₂-weighted imaging. Several types of infectious diseases such as tuberculoma or brain abscess are characterized by areas of hypointensity on T₂-weighted MR imaging. However, NBD lesions have no characteristic appearance on T₂-weighted imaging. NBD lesions tend to be confined to the basal ganglia and upper brainstem. The very extensive lesions seem to be the most important and typical of acute NBD. Gadolinium enhancement may be indicative of isolated or multiple demyelinated inflammatory lesions in NBD. In our case, MR imaging showed multiple small ring-like enhancement in the basal ganglia, brainstem, and cerebral white matter. This pattern of enhancement is rarely seen in neoplastic lesions. The large lesion possibly resulted from the confluence of several isolated lesions.

Corticosteroids (pulsed methylprednisolone) are usually used for the treatment of NBD, followed by cyclophosphamide (pulsed) and azathioprine. Drug therapy achieved a dramatic improvement in 83% of patients, and a mild improvement in others. However, only about 30% of the patients had a single episode whereas 37% of patients had a chronic progressive course. A recent review of 200 patients with NBD suggested that relapse during steroid tapering was associated with a poorer prognosis.

In conclusion, NBD should be considered in the differential diagnosis of a large lesion extending from the basal ganglia to the brainstem with multiple ring-like enhancement, because dermatological manifestations are sometimes absent during remission. Definite and early diagnosis of NBD is important, because diffuse involvement of the pyramidal tract may result in a poor response to steroid therapy.

References

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