

A case of Behçet's disease with development of MS-like lesions in the CNS and spinal cord

Sirs,

Behçet's disease (BD) is a multisystem recurrent inflammatory disorder of unknown etiology. Neurological involvement in BD may clinically and radiologically mimic that in multiple sclerosis (MS) (1). Here, we describe a patient with BD who developed MS-like lesions in the CNS and spinal cord.

A 40-year-old Japanese male was referred from the Department of Neurology to our department owing to visual loss in the left eye in November 2010. The patient had a history of recurrent oral aphthous ulcer since he was a teenager and recurrent uveitis from 1994. In April 2008, myelitis with thoracic lesions was detected in magnetic resonance imaging (MRI). In October 2008, he complained of numbness in both legs, followed by numbness and paresthesia in his arms. Brain MRI showed multiple hemispherical white matter lesions close to the lateral ventricle, subcortical frontal lobe, and posterior limb of the internal capsule on T2-weighted images (Fig. 1A-C). Cervical spine MRI revealed a hyperintense lesion in the spinal cord (Fig. 1D-E) and thoracic spine MRI showed a hyperintense lesion at Th5, 7, and 9-11. Cerebrospinal fluid (CSF) analysis showed a clear, colorless fluid with a slightly elevated cell count of 7/μL (predominantly mononuclear cells), a protein level of 38 mg/dL, and a glucose level of 61 mg/dL. On subsequent examination, oligoclonal bands were positive and the IgG index was 1.18. The interleukin-6 level in CSF was not increased. Transverse myelitis due to MS was diagnosed and methylprednisolone i.v. 1 g/day for 3 days was initiated for myelitis. When oral prednisolone was decreased to 10 mg/day, the patient experienced acute painful visual loss in the left eye.

On presentation, his best-corrected visual acuity (BCVA) was 0.7 OD and 0.2 OS with a left relative afferent pupillary defect and cecocentral scotoma. Anti-AQP4 antibody was negative. Left optic neuritis was diagnosed and intravenous methylprednisolone (1 g/day) was administered. Following 3 courses of steroid pulse therapy, BCVA in the left eye improved to 0.8.

In November 2011, the patient had an attack of bilateral panuveitis with associated hypopyon, followed by 3 uveitis attacks with recurrent oral aphthous ulcer within 3 months. An examination showed BCVA of 0.9 OD and hand motion OS. Fluorescein angiography revealed retinal vasculitis with fern leaf hyperfluorescence. Genotyping of HLA genes indicated HLA-A*26:01,

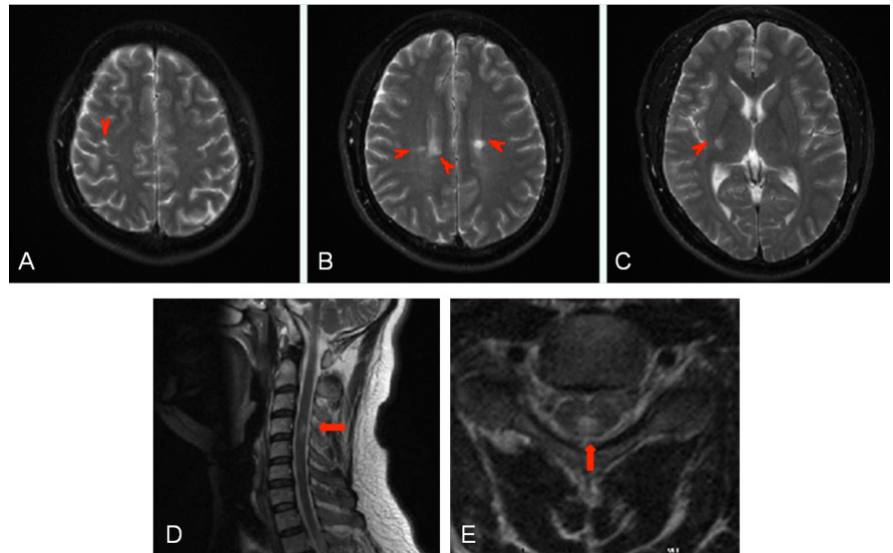


Fig. 1. Magnetic resonance imaging of the brain and cervical spine. The brain images showed multiple hyperintense MS-like plaques in the subcortical white matter (A), periventricular (B) and posterior limb of internal capsule (C). The cervical spine images showed the MS-like plaque was centered more posteriorly within the spinal cord (D) and extended to the periphery (E).

which is one of the primary susceptibility genes involved in development of BD in Japanese patients. BD was diagnosed and the patient was treated with a regimen of 0.5 mg colchicine twice daily. There has been no subsequent recurrence of Behçet's uveitis at 18-month follow up, although folliculitis and erythema nodosum-like lesions were seen in July 2012. Bilateral BCVA at the last examination was 1.0.

Neurological involvement in BD affects 5–50% of patients and can manifest as meningeal lesions, inflammatory lesions, and vascular changes (2–7). Therefore, the clinical course of CNS and spinal cord-affected patients with BD may clinically and radiologically mimic MS. Because our patient had neurological manifestations such as transverse myelitis and optic neuritis, and multiple hemispherical and spinal cord white matter lesions on MRI, a diagnosis of MS was made based on the McDonald criteria (8) before the onset of recurrent ocular attacks. Thus, differential diagnosis between neuro-BD and MS may be difficult if neurological manifestations develop before the ocular and systemic appearance fulfilling diagnostic criteria for BD (9).

In summary, this case indicates that clinicians should pay attention to the development of MS-like lesions in the CNS and spinal cord of a patient with BD.

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