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Chronic Progressive Neuro-Behçet's Disease: Magnetic Resonance Spectroscopy and Apparent Diffusion Coefficient Findings

Kronik Progresif Nöro-Behçet Hastalığı: Manyetik Rezonans Spektroskopi ve Görünür Difüzyon Katsayısı Bulguları

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Dear Editor,

A man aged 43 years with Behçet's disease (BD) presented with speech disorder, imbalance, and involuntary laughing and crying episodes. He had a history of recurrent oral aphtous ulcerations, genital ulcers, uveitis, and deep vein thrombosis, and he had been using colchicine until 5 years ago. A neurologic examination revealed severe cerebellar dysarthria, minimal loss of muscular power in the upper and lower extremities, spasticity, hyperactive deep tendon reflexes, positive bilateral Babinski signs, loss of abdominal skin reflex on the left, ataxic gait, and impaired tandem gait. Brain magnetic resonance imaging demonstrated atrophy, which was marked in the pons and cerebellum, and mild in the tegmentum of mesencephalon and supratentorial structures, and there was dilatation of the prepontine and cerebellopontine cisterns, and ventricular dilatation (Figure 1). Magnetic resonance spectroscopy demonstrated a normal spectral view at the corona radiata level and elevated choline at the pons level; choline/N-acetyl aspartate (NAA) ratio was 1.01 (Figure 2A, 2B). Diffusion-weighted evaluations detected increased apparent diffusion coefficient (ADC) secondary to atrophy (Figure 3). The findings were interpreted as chronic progressive neuro-BD (NBD) and methotrexate was initiated.

Central nervous system involvement in BD is known as NBD and it is one of the most severe complications, which significantly affects quality of life. Chronic progressive NBD, which is characterized by neurologic impairment and is usually resistant to immunosuppressive treatments, is present in 30%

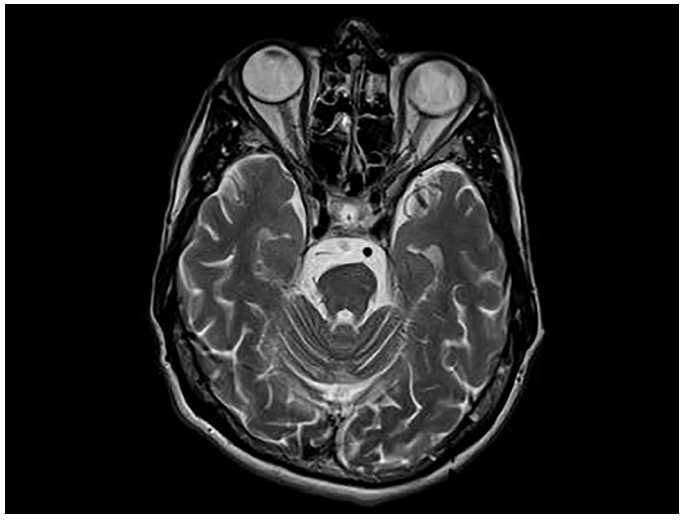

Figure 1. Brainstem atrophy

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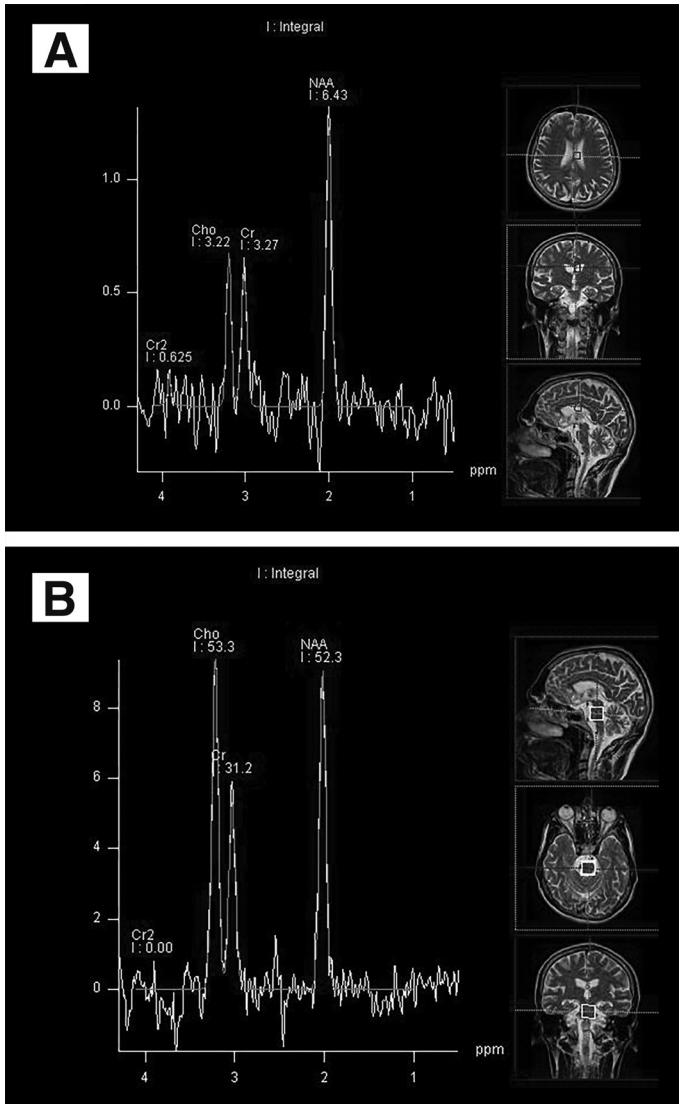

Figure 2. Normal spectral appearance at the corona radiata level (A); increased choline at pons level (B), choline/N-acetyl aspartate ratio 1.01
NAA: N-acetyl aspartate, Cho: Choline, Cr: Creatine

of patients with NBD (1). The diagnosis of chronic progressive NBD is made using radiologic findings that indicate brainstem atrophy, especially in the mesencephalon tegmentum and pons, and persistant elevation of interleukin-6 levels in cerebrospinal fluid (1,2). Other reported radiologic abnormalities include ADC increase in normal-appearing white matter, which indicate chronic progressive inflammation, and decreased NAA to creatine (NAA/Cr) ratio in the pons and cerebral white matter in magnetic resonance spectroscopy, which indicate extensive neuronal damage in cerebral white matter (3,4). There are reports that suggested that methotrexate and infliximab may be useful for the treatment of chronic progressive NBD (5).

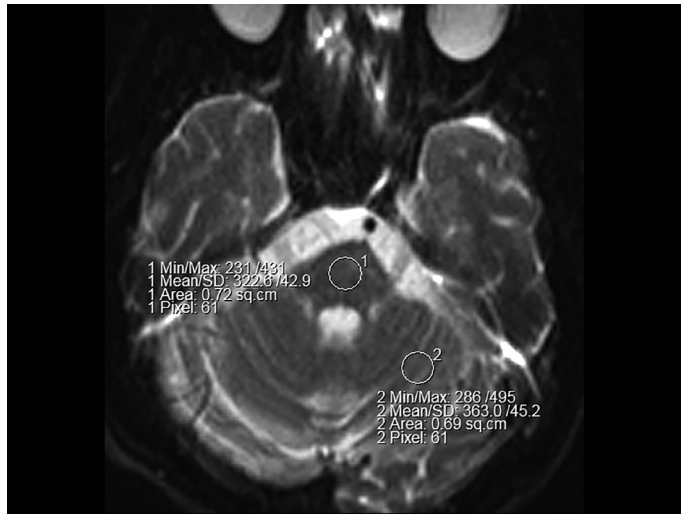

Figure 3. Increased apparent diffusion coefficient secondary to atrophy in diffusion-weighted imaging
SD: Standard deviation

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Internally peer-reviewed.

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