A Brainstem Plus Sign in Neuro-Behçet Disease

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Picture 1.  (A-F) Axial FLAIR brain MRI revealed multiple hyperintense lesions involving bilateral pontomedullar junctions, right-sided midbrain, thalamus and gangliocapsular area. This finding of confluent brainstem lesions extending into unilateral thalamus and basal ganglia is so-called "brainstem plus" sign. (G) Lesions were isointense on T1-weighted image. (H) T1-weighted axial MRI with gadolinium injection depicted partial enhancement of the lesions. (I) Follow-up axial MRI with gadolinium injection at 1 month later showed marked improvement of the affected areas.

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A 21-year-old Dominican-Lebanese male experienced right hemicranial headache for a week followed by blurry vision and horizontal diplopia. Examination revealed visual acuity of 20/200 OD and 20/30 OS, bilateral abducens nerve palsy, right-sided ptosis and swelling of the right optic nerve head. MRI of the brain (Picture 1) disclosed a “brainstem plus” sign, a typical radiographic feature of neuro-Behçet disease (NBD) (1). Although there were no orogenital ulcers seen on the exam, the patient admitted having multiple recurrent painful oral ulcers since childhood and one scrotal ulcer recently. Lumbar puncture revealed normal opening pressure, mild to moderate pleocytoses (340 white blood cells /μL) and mildly increased protein levels (63 mg/dL).

After a 7-day course of methylprednisone, his visual acuity returned to 20/40 OD and 20/25 OS without residual ptosis or ophthalmoparesis.

Behçet disease (BD) is a multisystem vasculitis (1). Unlike uveitis, optic neuritis is an infrequent ocular manifestation of BD (2). Neurologic complications occur in 5 to 14% of the Middle Eastern BD patients but are less common in the North American BD patients (1). Patients usually develop NBD during their 20’s to 40’s (1). Despite its rarity, the patient’s ethnic background and the typical radiographic findings should prompt the clinicians to include NBD in the differential diagnosis of optic neuritis and demyelinating disease in the young.

References


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