Neuro-Behçet's disease presenting with WEBINO syndrome

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Internuclear ophthalmoplegia (INO) is caused by various lesions of the brainstem with a horizontal gaze center, especially in the medial longitudinal fasciculus (MLF). Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) syndrome, is characterized by bilateral adduction deficits and large-angle exotropia. WEBINO has been reported in association with inflammatory, toxic, degenerative, infectious, traumatic, demyelinating, and neoplastic conditions. However, so far, there have been no reports of WEBINO syndrome caused by Behçet's disease. Herein, we report a case of Neuro-Behçet's disease with WEBINO syndrome.

A 20-year-old Korean woman visited our hospital with a sudden onset of binocular diplopia a week ago. The corrected visual acuity was 20/20 in both eyes, and 45 prism diopter exotropia at distance and near was observed. There was adduction limitation in both eyes with dissociated abducting nystagmus (Fig. 1A). There was no fundus torsion. She had a history of being treated for oral ulcer and uveitis. She denied any history of trauma and reported no other symptoms, including neurologic symptoms.

In the laboratory tests, blood count, renal and liver functions tests, coagulation panel, erythrocyte sedimentation rate, C-reactive protein, and angiotensin-converting enzyme were within the normal limits. The HLA-B51 was positive, and the pathergy test was positive. Tests for infection markers were all negative. The chest radiograph showed no abnormalities. Other autoimmune antibodies are all negative and thyroid function tests showed no specific findings. Brain magnetic resonance imaging showed T2/FLAIR high signal intensity lesions in the midbrain and pons, accompanied by changes suggestive of vasculitis (Fig 1B).

Intravenous administration of steroids was performed for 3 days at a dose of 1 g/day under the diagnosis of WEBINO syndrome associated with probable Neuro-Behçet's. After treatment, the adduction limitation was improved and the angle of exodeviation decreased. At the time of the final follow-up, 3 years later, T2/FLAIR high signal intensity lesion in the brain magnetic resonance imaging remained in the form of a subtle high signal lesion, and there was no adduction limitation of both eyes presenting orthotropia without diplopia (Fig 1C).

We demonstrated that Neuro-Behçet's disease presenting with WEBINO syndrome has a favorable outcome. The prognosis of INO is different depending on the cause, but the prognosis is relatively good for inflammatory and demyelinated diseases, and the prognosis is poor when accompanied by neurological symptoms. As in this case, treatment of the underlying disease has priority.

WEBINO is a rare variant of internuclear ophthalmoplegia associated with bilateral lesions of the MLF. The mechanism of exotropia in WEBINO syndrome is not clearly known, but it is caused by various lesions from the
pons to the midbrain. So far, there are not many reports about the prognosis of WEBINO.

Behcet's disease is a multisystem disease with features of mucocutaneous, ocular, intestinal, articular, vascular, and neurologic involvement. The most common ophthalmological abnormalities caused by Behcet's disease are uveitis. Alghamdi A et al. reviewed patients diagnosed with Neuro-Behçet's disease, and neuro-ophthalmological manifestations represented 13% of Neuro-Behçet's disease. They reported that 8 patients (27.5%) had diplopia due to ocular motor nerve palsies. There was no case presenting INO. In this case, the patient presented with diplopia and positive for pathergy test and HLA-51 with specific findings in brain MRI. After steroid treatment, symptoms improved without complications.

MRI findings of neurobehçet's disease usually present the lesions appearing to be the same signal or low signal intensity on T1-weighted image and, T2-weighted image or FLAIR image lesions of high signal intensity are seen. It is a finding associated with vasculitic changes in Behcet's disease, and the most often locations are known as the brain stem, basal nucleus, and cortex. In this case, high signal intensity lesions in the midbrain and pons were noted. The horizontal gaze center was involved, so the patient presented bilateral adduction limitation. Therefore, we could diagnose the patient with probable Neuro-Behçet's disease according to the 2014 International consensus recommendation criteria.

In conclusion, we report a case of WEBINO syndrome as a neuro-ophthalmologic manifestation of Behcet’s disease. Exotropia due to adduction limitation, considered with WEBINO syndrome, can occur in neuro-Behcet's disease. Since the prognosis is favorable, patients with sudden diplopia with eye movement limitation should be investigated thoroughly and performed timely treatment.

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Figure legend

Fig. 1. Patient presented exotropia with adduction limitation on both eyes at the initial visit (A). Brain MRI revealed high signal intensity lesions (yellow arrow) in the midbrain and pons (B). The adduction limitation on both eyes was improved after intravenous steroid treatment (C).