Wall-Eyed Bilateral Internuclear Ophthalmoplegia in a Patient with Progressive Supranuclear Palsy

Munetaka Ushio, MD, Shinichi Iwasaki, MD, Yasuhiro Chihara, MD, and Toshihisa Murofushi, MD

Abstract: Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) is a rare disorder consisting of a bilateral adduction deficit and primary gaze position exotropia. Associated with bilateral medial longitudinal fasciculus lesions, it has been mostly reported in patients with multiple sclerosis and brainstem stroke. A 72-year-old man with characteristic clinical features of progressive supranuclear palsy (PSP) later developed WEBINO. Brain MRI revealed atrophy of the midbrain tegmentum. Caloric irrigation revealed intact horizontal eye movements in both eyes. We believe this to be the first report of WEBINO in PSP. The preservation of vestibulo-ocular horizontal eye movements supports the notion that the WEBINO in this condition was caused by a supranuclear rather than a nuclear lesion and suggests the possibility that even in other causes of WEBINO, the lesion is supranuclear and not in the medial rectus subnucleus as is often suggested.

Case Report

A 72-year-old man was referred to our clinic with complaints of diplopia and gait disturbance. He had noted mild dysarthria and resting tremor in the right upper extremity for 11 years. Parkinson disease had been diagnosed earlier, and he had undergone levodopa therapy. However, it had proved ineffective. For 7 years, he had noted a frozen gait, mild disequilibrium, difficulty in writing, and mild recent memory disturbance. Amantadine hydrochloride therapy had been effective in alleviating the resting tremor. A disorder of postural reflexes, dysarthria, and dementia gradually developed thereafter. For 1 year, he had noted diplopia on lateral gaze bilaterally. Bilateral internuclear ophthalmoplegia with primary gaze position exotropia was identified. Past medical and family histories were noncontributory.

On our examination, the patient displayed mild dementia (15/30 on Mini Mental State Examination). Visual acuity and fields were normal in both eyes. Pupils were of normal size, equal in diameter, and normally reactive to light. The magnitude of the exotropia of the right eye (right eye fixating) in primary gaze position was 50 prism diopters; the magnitude of the exotropia of the left eye (right eye fixating) was 55 prism diopters using the Krimsky test. The patient showed an adduction deficit on lateral gazing bilaterally. Vertical eye movements were severely limited. Convergence was absent (Fig. 1).

Distal and proximal wasting and weakness were not observed in any of the four extremities. Although he displayed a mask-like facies, frozen gait, and nuchal rigidity, he was able to stand and walk with aids. Deep tendon reflexes were normal except at the patella, where they were

The most common causes of internuclear ophthalmoplegia and its WEBINO variant are demyelination and stroke (1,3-5). Internuclear ophthalmoplegia has also been reported rarely in patients with progressive supranuclear palsy (PSP) (6-8), but to the best of our knowledge, its WEBINO variant has not been reported in PSP. We report such a case.
absent. Extensor plantar reflexes were seen bilaterally. No loss of sensation was evident.

Brain MRI revealed atrophy of the midbrain tegmentum (Fig. 2). The pons, cerebellum, and basal ganglia showed no evidence of atrophy.

Spontaneous, positional, and positioning nystagmus were not seen with an infrared charge-coupled device (CCD) camera. On electronystagmography (AC-coupled; time constant of original recording = 3 seconds, time constant of eye velocity = 30 milliseconds, upper frequency limit = 10 Hz), he showed smooth pursuit in the horizontal plane. Precise calibration was precluded by the ophthalmoplegia, but an estimate of ductional amplitudes could be attained by clinical observation. Neither eye was able to adduct past the midline with saccades or smooth pursuit, but oculocephalic maneuvers during attempted fixation of an object in light elicited a full range of horizontal eye movements (Fig. 3A). On lateral gaze, dissociated nystagmus on abduction was observed bilaterally (Fig. 3B). Although vertical eye movements, including saccades and smooth pursuit, were severely limited, oculocephalic maneuvers during attempted fixation of an object in light elicited a full range of vertical eye movements (Fig. 3C). On caloric tests using ice water, each eye adducted and abducted to the full horizontal range contralateral to the

**FIG. 1.** Cardinal gaze positions: primary position, lateral (←, →), upward (↑), downward (↓), and convergence (→ ↔). Exotropia is evident in the primary position. Adduction deficits are evident on lateral gaze bilaterally. Upward and downward eye movements are severely reduced. Convergence is absent.

**FIG. 2.** T2 sagittal brain MRI reveals atrophy of midbrain tegmentum (arrow).
in response to clicks at 95 dB normal hearing level (9), showed normal responses on both sides. Normal ocular counter-rolling was observed under the infrared CCD camera. Other cranial nerves functioned normally by clinical examination.

On the basis of these findings, PSP with WEBINO was diagnosed. Gait disturbance and ophthalmoplegia have displayed gradual deterioration despite medication.

**DISCUSSION**

Our patient displayed the typical features of PSP and was distinctive only in having a WEBINO, which has not been previously reported in this condition. As the lesions of PSP characteristically involve the pontine and midbrain tegmentum and those of WEBINO involve the same regions (1,10), the appearance of WEBINO in PSP is not surprising. Brain MRI for this patient did reveal atrophic changes in the midbrain tegmentum, a finding previously described in WEBINO (11).

In this patient, horizontal and vertical eye movements, including saccades and smooth pursuit, were limited, whereas oculocephalic and caloric maneuvers elicited full range of movements. These findings suggest that the vestibulo-ocular reflex is intact and that the ophthalmoplegia in this patient probably originated from a supranuclear lesion (12). McGettrick et al (1) have suggested that an oculomotor nucleus lesion accounts for the exotropia in patients with WEBINO, but Gonyea (3) reported that the oculomotor nucleus was not involved. The findings in our patient favor Gonyea's observation.

This patient also showed loss of convergence, but this finding does not necessarily support the postulate that the exotropia of WEBINO is due to an oculomotor nuclear lesion (14). Impaired convergence is common in older adults. In our patient, the cause of the convergence disorder is unclear.

We believe that the WEBINO in this patient was caused by extension of the PSP lesions into the midbrain tegmentum. The preservation of vestibulo-ocular movements supports the notion that the WEBINO was caused by a supranuclear rather than a nuclear lesion and suggests that WEBINO in demyelination and stroke may also be of supranuclear origin.

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**REFERENCES**


