Progressive Supranuclear Palsy With Walleyed Bilateral Internuclear Ophthalmoplegia Syndrome

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Background: Walleyed bilateral internuclear ophthalmoplegia (WEBINO) syndrome has mainly been reported in patients with cerebrovascular diseases and multiple sclerosis, but has never been described in patients with neurodegenerative diseases.

Objective: To describe a patient with progressive supranuclear palsy (PSP) who presented with WEBINO syndrome.

Design: Case report and review of literature.

Setting: A university hospital.

Patient: A 72-year-old man began to display akinesia, freezing of gait, postural instability, mild rigidity of the neck, and vertical supranuclear palsy, including downward gaze limitation, at 66 years of age. At 68 years, he started to develop diplopia. At 70 years, he had bilateral medial longitudinal fasciculus syndrome. Later, his eye positions gradually showed alternating exotropia.

Results: A diagnosis of probable PSP was made based on the National Institute of Neurological Disorders and Stroke and the Society for Progressive Supranuclear Palsy criteria. He showed alternating exotropia in the forward gaze, and adduction paresis and monocular nystagmus of the abducted eye in the horizontal gaze, 2 clinical symptoms of WEBINO syndrome.

Conclusion: This is the first reported case of a patient with PSP presenting with WEBINO syndrome. Because bilateral medial longitudinal fasciculus lesions are commonly observed in PSP as clinical and pathological findings, particular attention should be given to WEBINO syndrome in patients with PSP.

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REPORT OF A CASE

The patient was a 72-year-old man who showed walking hesitation and was prone to falling at 61 years of age. At 66 years, akinnesia, freezing of gait, postural instability, mild rigidity of the neck, and vertical supranuclear palsy, including downward gaze limitation, were observed. Carbipodailevoda, pergolide, and droxydopa were prescribed, but their effects were not obvious. At 68 years, he started to develop diplopia. At 70 years, his adducted eye remained at the mid position and his abducted eye showed monocular nystagmus in the lateral gaze, bilaterally. Later, his eye positions gradually showed alternating exotropia. At 72 years, he was admitted to the University of Tokyo Hospital for evaluation of his gait disturbance and oculomotor abnormalities. His medical and family histories were unremarkable.

Neurological examination revealed cognitive impairment, parkinsonism, oculomotor abnormalities, and apraxia of eyelid opening. His cognitive impairment included apathy, marked slowing of cognition, decreased verbal fluency, and frontal release signs. His parkinsonism was characterized by akinnesia, freezing of gait, postural instability, moderate rigidity of the neck, and mild rigidity of both legs. His oculomotor abnormalities were as follows: (1) In the vertical gaze, his upward
gaze was markedly restricted and his downward gaze was also moderately restricted. (2) His eye convergence was disturbed. (3) When he was asked to fix his eyes on an object in front of him, 1 of his eyes fixated at the mid position, whereas his other eye deviated outward with accompanying abduction nystagmus. (4) His exotropia alternated between both eyes, and the condition was remarkably relieved by wearing Frenzel goggles. (5) In his horizontal gaze, adduction paresis and monocular nystagmus of the abducted eye were bilaterally observed. (6) His oculocephalic reflex was preserved (Figure).

Brain magnetic resonance imaging showed marked atrophy of the midbrain tegmentum, dilatation of the third ventricle, and slight brain atrophy of his frontal and temporal lobes on T1-weighted images, but no abnormal signals were observed on T2-weighted images.

A diagnosis of probable PSP was made, based on the National Institute of Neurological Disorders and Stroke and the Society for Progressive Supranuclear Palsy criteria.\(^3\) In addition, the observed alternating exotropia in the forward gaze and the adduction paresis and monocular nystagmus of the abducted eye in the horizontal gaze were indicative of WEBINO syndrome.

**COMMENT**

Here, we describe a 72-year-old man with PSP presenting with WEBINO syndrome. His oculomotor abnormalities progressed parallel to the progression of PSP. He showed alternating exotropia in addition to vertical gaze palsy and loss of eye convergence, 2 symptoms commonly observed in PSP patients.\(^4\) His adduction paresis and monocular nystagmus of the abducted eye with intact oculocephalic reflex suggest internuclear ophthalmoplegia. Because internuclear ophthalmoplegia is a disturbance of the medial longitudinal fasciculus (MLF), part of his oculomotor abnormalities are considered to be caused by bilateral MLF lesions.\(^5\)

Gonyea\(^1\) reviewed the literature concerning internuclear ophthalmoplegia and reported 3 cases of WEBINO syndrome that are associated with alternating exotropia. He concluded that this syndrome is caused by bilateral MLF lesions, because Spiller\(^6\) had previously described a patient with pathologically bilateral MLF infarction but no oculomotor nuclear lesions. Komiyama et al\(^2\) described the following observation of 2 patients with WEBINO syndrome: When the patients were asked to look at objects using both eyes, 1 eye of each patient fixated on the objects and the other eye became abducted, with accompanying monocular nystagmus. When visual fixation was deprived by the use of Frenzel goggles, exotropia was remarkably diminished. They speculated that the overexcitation of each contralateral paramedian pontine reticular formation in the abducted paretic eye was involved in the development or aggravation of the alternating exotropia, indicating that WEBINO syndrome is caused by both bilateral MLF lesions and the overexcitation of each paramedian pontine reticular formation.

Steele et al,\(^7\) in their original report on the neuropathological findings of PSP, described the involvement of the MLF. Mastaglia and Grainger\(^8\) reported patients clinically diagnosed as having PSP who presented with oculomotor abnormalities including bilateral MLF syndrome.

We conclude that WEBINO syndrome with alternating exotropia in our patient is a clinical manifestation of both bilateral MLF lesions and the overexcitation of each paramedian pontine reticular formation in PSP. Because bilateral MLF lesions are commonly observed in PSP as clini-
and pathological findings, particular attention should be given to WEBINO syndrome in patients with PSP.

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REFERENCES


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