Case Report

Progressive Supranuclear Palsy with Wall-Eyed Bilateral Internuclear Ophthalmoplegia Syndrome: Authors' Second Case

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Keywords

Progressive supranuclear palsy · Wall-eyed bilateral internuclear ophthalmoplegia syndrome · Medial longitudinal fasciculus · Paramedian pontine reticular formation

Abstract

Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) syndrome has previously been reported in only 2 patients with progressive supranuclear palsy (PSP). Herein, we report a third case of WEBINO syndrome with PSP. The patient was an 81-year-old man who had experienced gradually increasing gait disturbance and occasional falls since the age of 78 years. At 80 years of age, he presented with cognitive impairment, parkinsonism, and oculomotor abnormalities. The oculomotor abnormalities consisted of vertical gaze palsy and loss of eye convergence. Brain magnetic resonance imaging demonstrated marked atrophy of the midbrain. He was diagnosed with PSP. At the age of 81 years, he presented with alternating extropia in his forward gaze and adduction paresis and outward nystagmus of the abducted eye in his horizontal gaze, both of which were compatible with WEBINO syndrome. Previously, we reported the first case of PSP with WEBINO syndrome, and another group recently reported a second case. In light of the previous cases and the present case, WEBINO syndrome in PSP should not be considered extremely rare. Furthermore, WEBINO syndrome has not been reported in other neurodegenerative disorders, which suggests that it might be a useful and specific diagnostic finding in PSP.

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Introduction

Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) syndrome has mainly been reported in patients with cerebrovascular diseases [1] and multiple sclerosis [2]. In 2008, Matsumoto et al. [3] (the first author of this article) reported a case of progressive supranuclear palsy (PSP) that presented with WEBINO syndrome. The clinical findings in the first case and the previous literature indicate that WEBINO syndrome is a clinical manifestation of both bilateral medial longitudinal fasciculus (MLF) lesions and the overexcitation of each paramedian pontine reticular formation (PPRF). Bilateral MLF lesions are known in PSP as clinical and pathological findings. We thus proposed that particular attention should be paid to WEBINO syndrome in patients with PSP. In 2017, de Souza et al. [4] reported a PSP patient who presented with WEBINO syndrome. The characteristics of the oculomotor abnormalities in their case were almost the same as those in the first case, suggesting the possibility that WEBINO syndrome might not be extremely rare in PSP.

Here, we report another patient with PSP who presented with WEBINO syndrome. This is the third case of PSP with WEBINO syndrome in the literature to date (the authors' second case).

Case Report

The patient was an 81-year-old man. At the age of 78 years, he noticed postural instability when walking and he occasionally fell down. His gait disturbance and falls gradually grew worse. At the age of 80 years, he visited our hospital due to difficulty with walking. Neurological examinations showed cognitive impairment, parkinsonism, and oculomotor abnormalities. His cognitive impairment included slowing of cognition and frontal release signs. His parkinsonism consisted of masked face, bradykinesia, moderate rigidity of the neck, mild rigidity of both legs, and postural instability. His oculomotor abnormalities were vertical supranuclear palsy, including downward gaze limitation and loss of eye convergence. At this time, his eyes were in the normal middle position. Brain magnetic resonance imaging demonstrated severe atrophy of the midbrain and moderate atrophy of the frontotemporal lobes. Carbidopa-levodopa 300 mg per day was prescribed but was not effective. He was diagnosed with PSP according to the clinical criteria [5]. Thereafter, he became unable to walk without assistance. At the age of 81 years, he became bedridden due to parkinsonism and had difficulty eating due to dementia and dysphagia. He was admitted to our hospital for gastrostomy. His medical and family histories were unremarkable.

Neurological examinations again showed cognitive impairment, parkinsonism, and oculomotor abnormalities. The cognitive impairment and parkinsonism had become worse compared to the previous year, and the oculomotor abnormalities included new findings. In addition to vertical gaze palsy and loss of eye convergence, the oculomotor abnormalities included, in his right gaze, adduction paresis of the left eye and rightward nystagmus of the right eye, and, in his left gaze, adduction paresis of the right eye and leftward nystagmus of the left eye. In his forward gaze, his eyes showed alternating exotropia: only one eye fixated on the object in front of him and the other eye was abducted. He mainly gazed with his left eye but occasionally used his right eye. The exotropia was markedly relieved by Frenzel goggles. His oculocephalic reflex was preserved (Fig. 1). The alternating exotropia in his forward gaze and the adduction paresis and outward nystagmus of the abducted eye in his horizontal gaze were compatible with WEBINO syndrome.

Discussion

Here, we report an 81-year-old man with PSP presenting with WEBINO syndrome. His oculomotor abnormalities changed in parallel with the progression of PSP. When he visited our hospital, he presented with the oculomotor abnormalities commonly observed in PSP patients, namely vertical gaze palsy and loss of eye convergence [6]. One year later, he entered our hospital after presenting with additional oculomotor abnormalities compatible with WEBINO syndrome (i.e., alternating exotropia in his forward gaze and adduction paresis and outward nystagmus of the abducted eye in his horizontal gaze). This is the third case of PSP with WEBINO syndrome (the authors' second case).

WEBINO syndrome is considered to be a clinical manifestation of both bilateral MLF lesions and the overexcitation of each PPRF. Gonyea [1] (1974) reported 3 patients who clinically exhibited WEBINO syndrome. He concluded that this syndrome is caused by bilateral MLF lesions, because Spiller [7] (1924) previously described a patient presenting with similar oculomotor abnormalities who had bilateral MLF infarction but no oculomotor nuclear lesions in an autopsy study. Komiyama et al. [2] (1998) reported 2 patients with WEBINO syndrome. Since the exotropia was markedly diminished by Frenzel goggles, they speculated that the overexcitation of each PPRF contralateral to the adducted paretic eye is involved in the development or aggravation of the alternating exotropia.

Bilateral MLF lesions have been reported in PSP. Steele et al. [8] (1964) reported the involvement of MLF in their autopsy cases of PSP, which is the original literature on PSP. Masstaglia and Grainger [9] (1975) reported patients clinically diagnosed with PSP who presented with oculomotor abnormalities, including bilateral MLF syndrome. Other authors also reported bilateral MLF syndrome in patients who were clinically diagnosed with PSP [6, 10]. In short, bilateral MLF lesions are known as clinical and pathological findings in PSP.

The characteristics of oculomotor abnormalities in the present case are almost the same as those in the first and second cases [3, 4]. Therefore, we believe that the pathophysiology of WEBINO syndrome in PSP is compatible with the previous hypothesis, namely that it includes both bilateral MLF lesions and the overexcitation of each PPRF. Since we experienced a second case of PSP with WEBINO syndrome, WEBINO syndrome should not be considered extremely rare in PSP. It is of note that WEBINO syndrome has not been reported in other neurodegenerative disorders, which might suggest that it is a useful and specific diagnostic finding in PSP.

Statement of Ethics

The patient and his family provided oral informed consent for publication of this paper.

Disclosure Statement

The authors have no disclosures. The authors have no potential conflicts of interest.

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Fig. 1. Photographs of oculomotor abnormalities in the present case. **a** Upward gaze. **b** Right gaze. **c** Forward gaze. **d** Left gaze. **e** Downward gaze. **f** Frenzel goggles.