

Wall-Eyed Monocular Internuclear Ophthalmoplegia (WEMINO) with Contraversive Ocular Tilt Reaction

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Wall-eyed monocular internuclear ophthalmoplegia (WEMINO) with contraversive ocular tilt reaction has not been previously reported. A 71-year-old woman suddenly developed blurred vision. Examination revealed left internuclear ophthalmoplegia, left exotropia, right hypotropia, and rightward head tilt. Magnetic resonance imaging showed a tiny infarction at the area of the left medial longitudinal fasciculus in the upper pons. WEMINO with contraversive ocular tilt reaction may be caused by a paramedian pontine tegmental infarction that selectively involves the medial longitudinal fasciculus.

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Wall-eyed monocular internuclear ophthalmoplegia (WEMINO) is a rare variant of internuclear ophthalmoplegia (INO) and consists of INO with ipsilateral exotropia.¹ This distinct syndrome is a genuine prenuclear disorder and known to be associated with damage to the medial longitudinal fasciculus (MLF).² However, to our knowledge, WEMINO combined with the ocular tilt reaction (OTR) caused by a brainstem lesion has not been previously reported.

CASE REPORT

A 71-year-old woman with hypertension and Parkinson's disease suddenly developed blurred vision and exotropia of the left eye. Neurological examination

performed sixteen hours after onset revealed mild dysarthria and rightward head tilt.

She also had adduction paresis of the left eye, abducting nystagmus of the right eye, left exotropia in the primary position, a skew deviation with the right hypotropia, an excyclotorsion of the right eye and an incyclotorsion of the left eye, upbeat nystagmus, intermittent right exotropia on downward gaze, and deficiency of convergence of the left eye (Fig 1-A, 1-B). No ptosis was noted at any time. Her pupils were equal and responded promptly to light. These findings were consistent with WEMINO and rightward OTR. Other findings of the neurological examination were normal.

Diffusion-weighted brain MRI performed 18 hours after the symptom onset showed an acute infarction in the left upper paramedian pontine tegmentum adjacent to

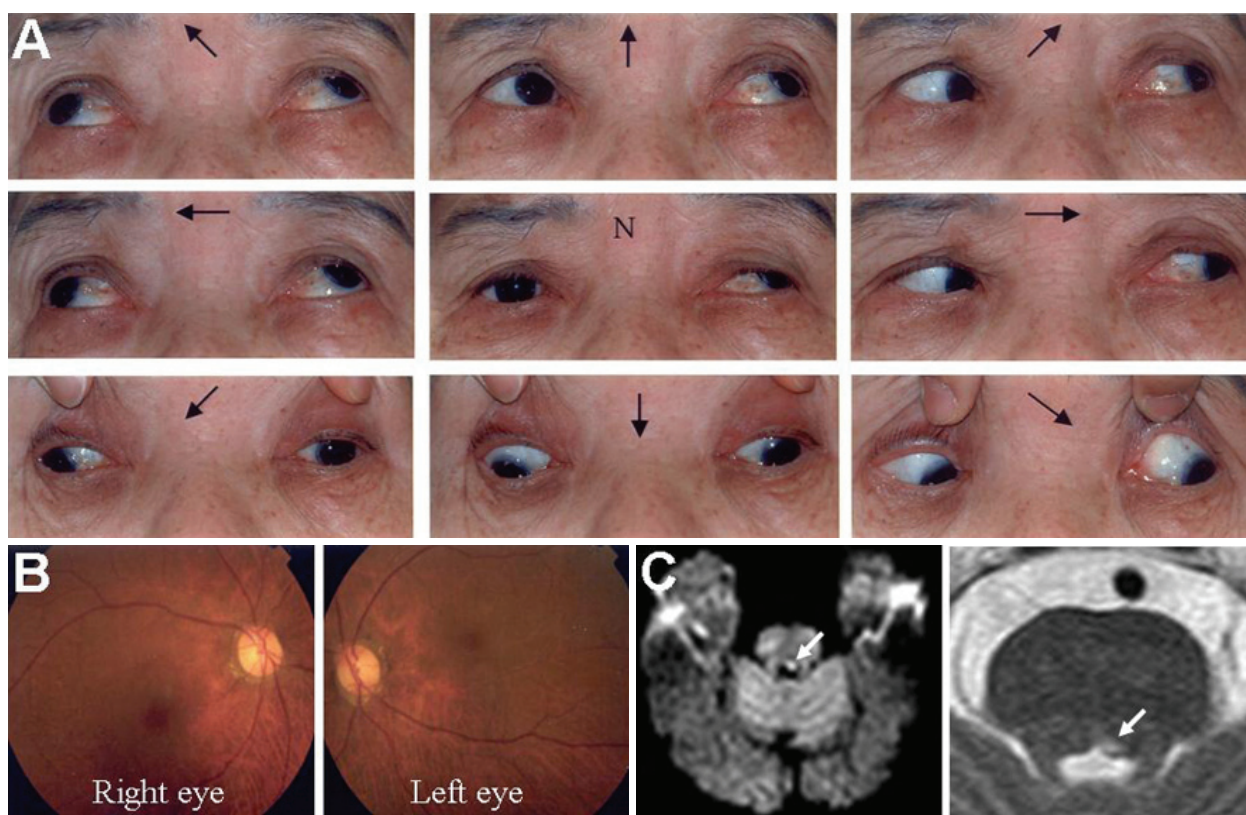


Figure 1. (A) Extraocular movements in 9 cardinal gazes. Left eye shows internuclear ophthalmoplegia with incomplete adduction, exotropia, and hypertropia. (B) Fundus photography. There are extorsion in the right eye and intorsion in the left eye. (C) Diffusion-weighted (performed 18 hours after the symptom onset) and T2-weighted MRIs (performed 9 days after onset) demonstrate a discrete infarction in the area of left MLF (arrows).

the fourth ventricle in the area of MLF. The follow-up MRI, performed 8 days later, revealed a tiny discrete lesion at the same area (Fig 1-C). This lesion showed high signal intensity on T2-weighted and fluid attenuated inversion recovery (FLAIR) MRI, and low signal intensity on apparent diffusion coefficient (ADC) map. Cerebral magnetic resonance angiography (MRA) results were normal. Over the next 30 days, the patient's skew deviation and exotropia improved gradually.

DISCUSSION

This is the first report of WEMINO combined with contraversive OTR due to a tiny lesion in the ipsilateral upper paramedian pontine tegmentum in the area of the MLF. There were no other neurologic signs except for

transient dysarthria.

WEMINO has rarely been reported.^{2,3} Of the four patients with WEMINO reported by Johnston and Sharpe,² only one had the responsible lesion confined to the pontine tegmentum in the area of the MLF, which was verified by neuropathologic examination. Iketa and Okamoto also reported a patient who had an infarct identified by MRI in the area of the MLF.³ However, these patients had neither skew deviation nor OTR.

OTR is common in patients with MLF involvement.¹ OTR is contraversive in patients with pontomesencephalic brainstem lesions involving the rostral MLF or the interstitial nucleus of Cajal, while it is ipsiversive in patients with medullary lesions.⁴ Therefore, contraversive OTR, observed in our patient, seems to be related to the lesion of the left pontine MLF rostral to the decussation.⁵

These findings should be differentiated from non-paralytic pontine exotropia, which is a syndrome of INO with contralateral exotropia. Contralateral exotropia in this condition is known to be due to secondary deviation induced by over-excitation of contralateral paramedian pontine reticular formation under fixation with the paretic eye.⁶ Although the mechanism of WEMINO remains unclear, the findings of our patient suggest that WEMINO is related to the MLF lesion.

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