INTERNUCLEAR OPHTHALMOPLEGIA ASSOCIATED WITH TRANSIENT TORSIONAL NYSTAGMUS

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Abstract: The association of internuclear ophthalmoplegia (INO) with torsional nystagmus is rare. We report the case of a 63-year-old man presenting with sudden onset of diplopia. On ocular examination, he had left INO and a torsional nystagmus with counter-clockwise quick-phases from the patient’s point of view in the primary position, ipsiversive to the side of the medial longitudinal fasciculus (MLF) lesion at the pontomesencephalic junction on magnetic resonance images. Neither skew deviation nor head tilting was noted at that time. Two days later, the torsional nystagmus disappeared, although the limitation of adduction in the left eye remained. We speculate that with sparing of the rostral interstitial nucleus of the MLF, a small lesion in the MLF may involve the ipsilateral vertical integrator, the interstitial nucleus of Cajal, producing an INO associated with a transient ipsiversive torsional nystagmus.

Case Report

A 63-year-old man presented with a sudden onset of diplopia in the right gaze 1 day before admission. He had no history of hypertension, diabetes, or hyperlipidemia. Ocular examination revealed that he had a left INO showing adduction paralysis of the left eye (Fig. 1A) and abducting nystagmus of the right eye in the right gaze. Convergence was normal. There was also a symmetrical and low-amplitude (< 10°) torsional nystagmus with counter-clockwise quick phases in the primary position from the patient’s point of view (Fig. 1B). The nystagmus was conjugate in both eyes without vertical dissociation, and the amplitude increased to the left gaze. The frequency was about 1 to 2 Hz. There was no vascular abnormality in the ocular fundi. Unsteady gait was also noted. He had no skew deviation and no head tilting. Two days after admission, the torsional nystagmus disappeared, but the INO remained unchanged.

Magnetic resonance (MR) images revealed a small lesion, probably an infarct, at the site of the left MLF at the pontomesencephalic level (Fig. 2). No lesion was seen at the site of the interstitial nucleus of Cajal (Fig. 3). Brainstem

Fig. 1. A) Left internuclear ophthalmoplegia (INO) showing limited adduction of the left eye, without impairment of other extraocular motility. B) A conjugate, low-amplitude (< 10°) torsional nystagmus with counter-clockwise quick phases (direction of large white arrow) in the primary position from the patient’s point of view, in association with left INO. C) Two months after onset, complete recovery of extraocular motility was noted.
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Fig. 3. Interstitial nucleus of Cajal (INC) is situated (white arrowhead) between the red nucleus (RN) (white arrow) and the superior colliculus (SC) (black arrow). There is no lesion at the INC on this magnetic resonance image. The rostral interstitial nucleus of the medial longitudinal fasciculus is not shown in this section; it is separated from the more caudal INC by tractus retrorflexus. MB = mammillary body; SN = substantia nigra.

Fig. 2. Axial T2-weighted magnetic resonance image at the level of the pons shows a high signal-intensity lesion at the site of the left medial longitudinal fasciculus (white arrow).

Discussion

INO results from an ipsilateral MLF lesion and is rarely associated with torsional nystagmus. Conjugate torsional movements that rotate the upper poles of the eyes toward the subject’s right (a clockwise direction from the subject’s point of view) are called clockwise movements, although they appear to be counter-clockwise to an observer. Conversely, conjugate movements that rotate the upper poles of the eyes toward the subject’s left are counter-clockwise. Torsional movements that rotate the upper poles of the eyes toward a certain side of the brain or toward the side of a brain lesion are called ipsiversive and ipsilesional, respectively [2].

Noseworthy et al postulated that torsional nystagmus was due to disruption of central vestibular connections [1]. Lopez et al suggested that torsional nystagmus resulted from lesions involving vestibular nuclei on the opposite side of the torsional nystagmus [3]. INO was not mentioned in either paper. In 1996, Dehaene et al reported four cases of INO with torsional nystagmus ipsilesional to the MLF lesion that inactivated the interstitial nucleus of Cajal [4]. The intact rostral interstitial nucleus of the MLF (riMLF) produced a corrective ipsiversive quick phase. Only two of the four cases had pure torsional nystagmus not combined with skew deviation and head-tilt, and in all their patients with INO associated with torsional nystagmus, as in our patient, the nystagmus disappeared within 3 days after its onset.

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The interstitial nucleus of Cajal has extensive rostral and caudal connections, including receiving excitatory inputs from the vertical semicircular canals of the contralateral labyrinth via the MLF and descending projections to the vestibular nuclei [5]. In experiments, unilateral interstitial nucleus of Cajal electrical stimulation always produces ipsiversive torsional eye movement. In monkeys, temporary unilateral interstitial nucleus of Cajal inactivation can result in severe gaze-holding deficit in vertical eye movements, and primary-position torsional nystagmus with ipsilesional fast phases [2, 6].

Burst neurons in the riMLF are responsible for rapid eye movement generation in torsional and vertical directions [7, 8]. Each riMLF contains neurons that burst for upward and downward eye movements, but for torsional quick phases in only one direction. Thus, the left riMLF discharges for quick phases are directed counter-clockwise with respect to the subject [8]. Each riMLF projects predominantly to the ipsilateral oculomotor and trochlear nuclei; however, projections to motor neurons innervating the elevator muscles (superior rectus and inferior oblique) appear to be bilateral [8]. Electrical stimulation of unilateral riMLF produces conjugate ipsiversive torsional eye movements. If the riMLF is inactivated, a contralesional tonic torsional deviation and mild defect in vertical movement will be induced and presents skew deviation [2, 8]. For example, with a lesion of the left riMLF,
counter-clockwise torsional quick phases (extorsion of the left eye and intorsion of the right eye) are lost. In addition, there is a static, contralesional torsional deviation, with torsional nystagmus beating contralesionally [8].

Halmagyi et al proposed that jerk see-saw nystagmus is due to unilateral inactivation of the interstitial nucleus of Cajal, with sparing of the riMLF [2]. The same lesions can sometimes, however, result in torsional nystagmus beating toward the lesion side without a disjunctive vertical component, as in our patient. Either complete ocular tilt reaction (OTR) or skew torsion without head tilt, like the end phase of slow-phase torsional nystagmus, indicates a unilateral peripheral deficit of otolith input or a unilateral lesion of graviceptive brainstem pathways from the vestibular nuclei (crossing midline at the pontine level) to the interstitial nucleus of Cajal [9]. The torsional nystagmus associated with INO is sometimes dissociated and usually associated with a skew deviation [10]. In our patient, a small infarct lesion in the left MLF was detected on MR images. Such a small lesion may interrupt paramedian pontine reticular formation (PPRF) pulsing or inactivate the interstitial nucleus of Cajal, but we could not detect any lesion in the PPRF or interstitial nucleus of Cajal by MR studies, because the imaging sections were not thin enough or the lesion was too small to be detected. The findings of ipsiversive quick-phase torsional nystagmus without ocular tilt or skew deviation are compatible with features of inactivated interstitial nucleus of Cajal with intact riMLF. The findings suggest that a small MLF lesion may be responsible for inactivation of ipsilateral interstitial nucleus of Cajal with intact riMLF, resulting in a transient torsional nystagmus without skew deviation.

All skew deviations are contraversive (contralateral eye lowermost) with rostral pontomesencephalic lesions, and are associated with ocular torsion and tilt of subjective visual vertical toward the lowermost eye [11, 12]. OTR consists of head tilt, skew deviation, and binocular ocular torsion. INO with torsional nystagmus may or may not be associated with contraversive OTR [4, 10]. Our review of the literature suggests the following three observations: when pontomesencephalic lesions produce INO, it may be associated with skew deviation or OTR, which is always contraversive; torsional nystagmus associated with INO is almost always ipsiversive; and INO associated with OTR without torsional nystagmus may implicate a combined inactivation of the interstitial nucleus of Cajal and riMLF.

Transient ischemia of blood supply to the interstitial nucleus of Cajal or partial damage of innervation with regeneration may be the reason that torsional nystagmus can disappear quickly in patients with small MLF lesions. The association of INO with torsional nystagmus is rarely considered because the initial signs of transient torsional nystagmus typically go unrecognized, or patients visit a doctor long after the onset of symptoms.

References