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내측연수증후군에서 발생한 나비넥타이, 상향 및 반시이소안진

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Bowtie, Upbeat and Hemi-seesaw Nystagmus in Medial Medullary Infarction

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A 20-year-old man with bilateral medial medullary infarction showed transition of bowtie and upbeat nystagmus into hemi-seesaw nystagmus. Follow-up MRI revealed near complete resolution of the right medullary lesion. This transition of nystagmus suggests that the upbeat nystagmus was generated by bilateral lesions in the ascending pathways from both anterior semicircular canals (SCC), and that the hemi-seesaw nystagmus was caused by damage to the pathway from the left anterior SCC.

Key Words: Bowtie Nystagmus, Hemi-seesaw Nystagmus, Medial Medullary Syndrome

Medial medullary infarction is characterized by contralateral paralysis of the arm and leg, contralateral loss of position and vibration sense, and an ipsilateral paralysis of the tongue. Rarely, upbeat nystagmus has been described. Bowtie nystagmus, which refers to oblique upbeat nystagmus with horizontal quick phases that alternate from right to left, has been reported in association with tobacco inhalation,[1] cerebellar disease[2], and brainstem anormalies.[3] However, the occurrence of upbeat and bow-tie nystagmus and their transition into hemi-seesaw nystagmus has not been reported in medial medullary infarction.

CASE REPORT

A 20-year-old man was admitted due to sudden loss of consciousness that had occurred on extending his neck after vigorous exercise. When he regained consciousness three to four minutes later, he noticed dysarthria, dysphagia, weakness in the right arm and leg, and oscillopsia. On admission, he had a blood pressure of 130/70 mmHg and a pulse rate of 90 beats per minute. He was alert, and he had a full range of extraocular movements. However, upbeat nystagmus of large amplitude was present in the primary position of gaze. Intermittently, the fast phases of nystagmus with a horizontal component that alternated from left to right occurred, which was consistent with bowtie nystagmus.[1-3] This nystagmus persisted without change, during up, down, and lateral gaze, and during convergence. He also

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showed right central type facial palsy and dysarthria. Both soft palate elevations were reduced bilaterally and no gag reflexes were elicited. He had a right hemiparesis of MRC grade II. Vibration and position senses were reduced in the right arm and leg, and both legs exhibited increased muscle tone. Deep tendon reflexes were increased and the Babinski sign was positive bilaterally. T2-weighted and diffusion-weighted MRI revealed a high intensity lesion in the bilateral rostral medulla (Fig. 1A). Cerebral angiography revealed areas of stenosis of the left vertebral artery at the C4-C1 segments (Fig. 1B). The imaging studies prompted a diagnosis of left vertebral artery dissection and bilateral medial medullary infarction. The patient was placed on anticoagulation. Three days after symptom onset, an examination disclosed a reduction in the amplitude of the upbeat nystagmus with minimal oblique quick phase. Seven days after symptom onset, he showed intorsional-upbeat nystagmus in the right eye and extorsional nystagmus in the left eye, which resembled a hemi-seesaw nystagmus. He also had a skew deviation with the hypotropic right eye. Head tilt was not evident. Follow-up MRI revealed near complete resolution of the right-sided lesion (Fig. 1C). Six months after symptom onset, the patient still showed a hemi-seesaw nystagmus, which decreased in amplitude and frequency markedly. Fundus examination revealed an extorsion ($22_{\circ}$) of the right eye and an intorsion ($8_{\circ}$) of the left eye (Fig. 1D).

DISCUSSION

Initially, our patient showed upbeat and bowtie nystagmus. The quick phases of the upbeat nystagmus were conjoined by horizontal saccades, giving rise to oblique quick phase that alternated from left to right. These trajectories created the pattern of bowtie nystagmus.[1-3] Although the mechanism of bowtie nystagmus remains to be elucidated, it has been reported to occur with upbeat nystagmus in association with tobacco inhalation or cerebellar disease.[2] The co-occurrence of bowtie, torsional, and seesaw nystagmus was also described in single patient with

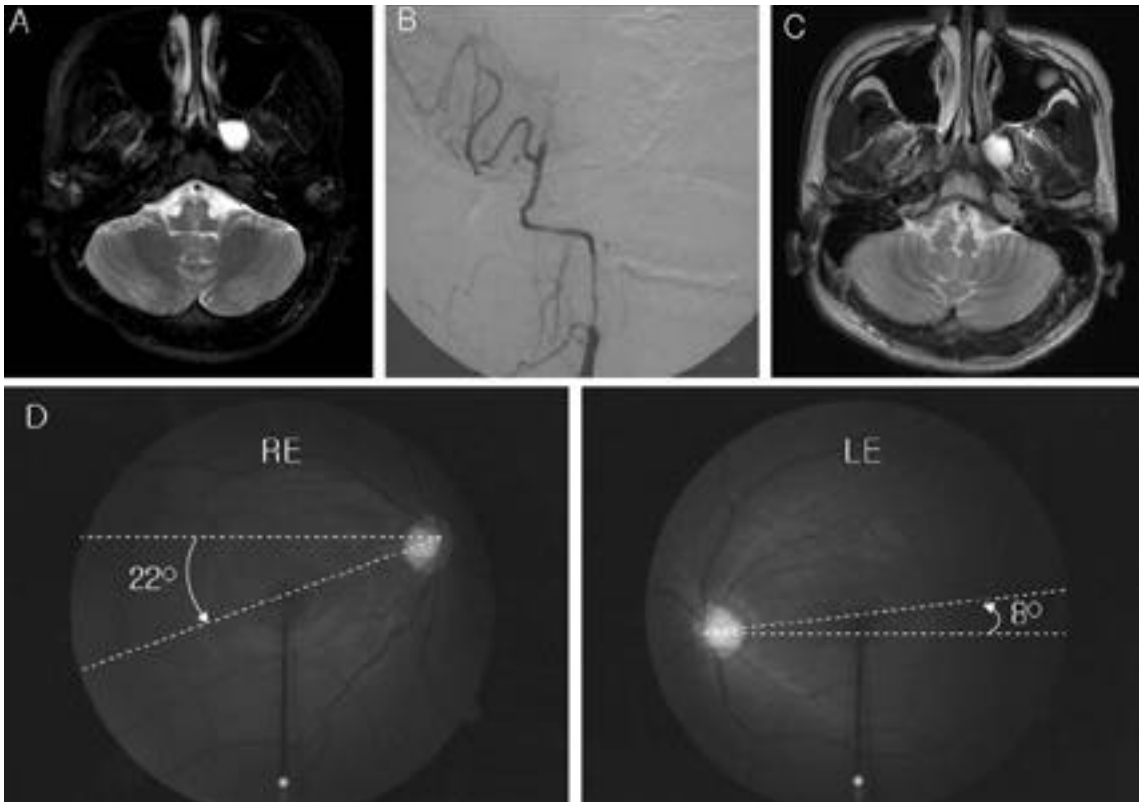

Figure 1. MRI and cerebral angiography of the patient. **A.** Initial T2-weighted axial image shows a high signal intensity lesion in bilateral rostral medulla. **B.** Cerebral angiography reveals stenotic left vertebral artery in the C4-C1 segments. **C.** Seven days after symptom onset, this follow-up MRI shows near complete resolution of the right-sided lesion. **D.** Fundus photos, taken six months after symptom onset, show an extorsion ($22_{\circ}$) of the right eye (RE) and an intorsion ($8_{\circ}$) of the left eye (LE).

syringomyelia, syringobulbia, and Arnold-Chiari malformation[3].

Upbeat nystagmus is usually described in lesions of the lower brainstem, ventral tegmentum, anterior vermis of the cerebellum, and the midbrain[4-6]. In cases of medial medullary infarction, the perihypoglossal nuclei, which consists of the nucleus prepositus hypoglossi, the nucleus of Roller, and the nucleus intercalatus were presumed to be involved.[5-7] The involvement of the ventral tegmental pathway for the upward vestibulo-ocular reflex (VOR) may also lead to a downward vestibular bias and consequent upbeat nystagmus[8]. The ventral tegmental pathway, with the appropriate connections to mediate the upward VOR, has been identified in cats.[9] Axons of this pathway project ventromedially along the medial border of the lateral lemniscus, ascend in the pontine tegmentum close to the medial lemniscus, and then abruptly turn dorsally, and ascend to the oculomotor nucleus. Indeed, in our patient, the lesion extended into the rostral part of the paramedian medulla, and the crossing pathways from the bilateral anterior semicircular canal, which mediate the upward VOR, may have been damaged, giving rise to upbeat nystagmus.

Interestingly, seven days after symptom onset, the upbeat and bowtie nystagmus were replaced by mixed torsional-vertical nystagmus, concurrent with near complete resolution of the right-sided lesion in the follow-up MRI. His right eye was hypotropic and the quick phase of the nystagmus was upbeating and intorsional in the right eye and mainly extorsional in the left eye, resembling half-cycles of seesaw nystagmus with corrective fast phases (jerky hemi-seesaw nystagmus). Although hemi-seesaw nystagmus usually occurs in unilateral meso-diencephalic lesions, lesions of the medulla can produce mixed torsional-vertical nystagmus, which can be indistinguishable from meso-diencephalic hemi-seesaw nystagmus. Hemi-seesaw and mixed torsional-

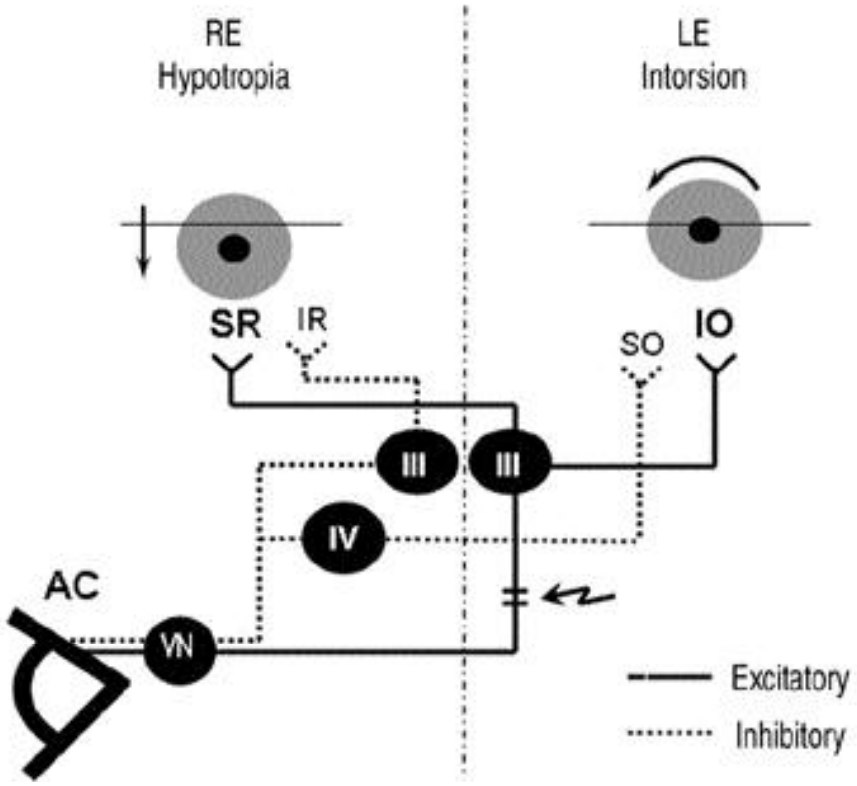

Figure 2. Hypothetical explanation of hemi-seesaw nystagmus in medial medullary lesion. Schematic representation of the three neuron vestibulo-ocular reflex arc between the anterior semicircular canal and the extraocular muscles. An excitatory ascending pathway is linked to the ipsilateral superior rectus and the contralateral inferior oblique muscles. An inhibitory ascending pathway is linked to the ipsilateral inferior rectus and the contralateral superior oblique muscle. Damage to the excitatory ascending pathway in the left paramedian medulla (angled arrow) causes primarily a downward deviation of the right eye, and intorsion of the left eye. The resulting corrective quick phases of nystagmus would be mostly upbeating in the right eye and extorsional in the left eye, which is consistent with the nystagmus observed in our patient. RE: right eye, LE: left eye, SR: superior rectus, IR: inferior rectus, SO: superior oblique, IO: inferior oblique, III: oculomotor nucleus, IV: trochlear nucleus, VN: vestibular nucleus, AC: anterior semicircular canal

vertical nystagmus may be explained by a central imbalance in the input from the vertical SCCs.[10] We propose that the initial upbeating nystagmus was caused by bilateral damage to the decussating or ascending pathways from bilateral anterior SCCs, and that the transition into hemi-seesaw nystagmus was induced by resolution of the right-sided lesion, leading to involvement of the pathway only from the right anterior SCC after decussation. Thus, the resulting ocular deviation would be downward and extorsional in the right eye and mainly intorsional in the left eye, in combination with the corrective quick phases of nystagmus in the opposite directions (Fig. 2). The torsional deviation of both eyes, observed during the follow-up examination, also supports this postulation.

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