

단독 상방 안진으로 재발한 다발성 경화증

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임의성 · 신병수

Relapse of Multiple Sclerosis with Isolated Upbeat Nystagmus

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Multiple sclerosis is an autoimmune demyelinating disorder of the nervous system. The ocular manifestation includes optic neuritis, internuclear ophthalmoplegia and nystagmus. Upbeat nystagmus is a rare manifestation of multiple sclerosis. We report a patient with relapsing multiple sclerosis who presented with upbeat nystagmus from a circumscribed lesion in the caudal medulla.

Key Words: Upbeat nystagmus, Multiple sclerosis, Relapse

Multiple sclerosis is an autoimmune demyelinating disorder of the nervous system. The ocular manifestation includes optic neuritis, internuclear ophthalmoplegia and nystagmus which result in diplopia, oscillopsia, blurred vision, loss of stereopsis, and reading fatigue.¹ Upbeat nystagmus (UBN) is a rare neurologic sign of relapsing multiple sclerosis with lesions affecting the pontomesencephalic junction, the rostral medulla, the caudal pons, or the cerebellum.^{2,3} We report on a patient with isolated UBN as a manifesting sign of relapsing multiple sclerosis.

Case

A 22-year-old woman was admitted to the hos-

pital with one day history of vertigo and vertical oscillopsia. She had suffered from clinically definitive multiple sclerosis with a relapsing and remitting course during the past five years, which had developed motor weakness, sensory disturbance, bladder dysfunction and optic neuritis. General physical examination were unremarkable and her mental status was intact. She showed upbeat nystagmus in the primary position. The nystagmus was unchanged by positional change, fixation removal, or convergence. Ocular motor ranges were full. Other neurologic examination was normal except for left optic nerve lesion due to previous optic neuritis. Normal laboratory studies included complete blood counts, blood chemistry, urinalysis and cerebrospinal fluid.

Computerized infrared video-oculography showed upbeat nystagmus with a constant velocity decay of the slow phases in the primary position (Fig. 1). Brain MRI revealed high signal intensity in the paramedian area of the caudal medulla on axial and sagittal T2-weighted images (Fig. 2).

Intravenous methylprednisolone was given with

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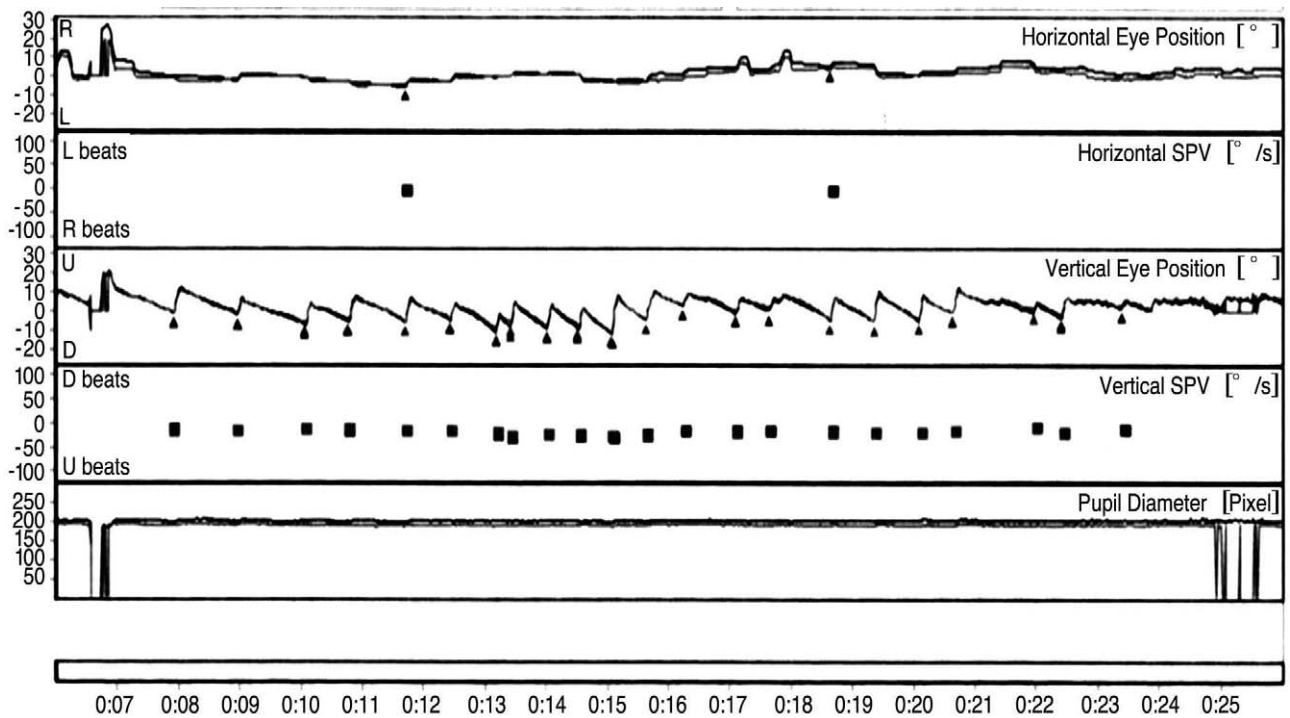


Figure 1. Video-oculography shows upbeat nystagmus in the primary position of gaze.

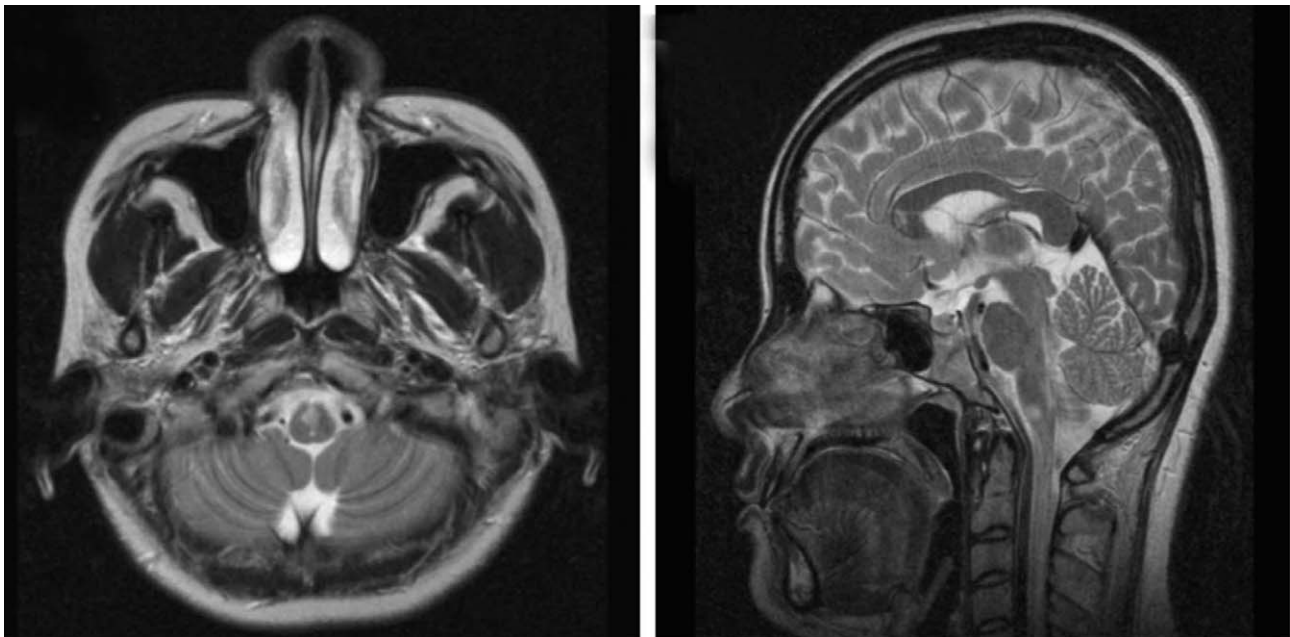


Figure 2. Axial and sagittal T2-weighted MRIs show a high signal intensity lesion in the paramedian area of the caudal medulla.

a dose of 1g per day for 3 days, which was followed by oral prednisolone 60 mg daily in a tapering schedule. The patient left the hospital 5 days after her admission. We tried gabapentin (1200 mg/day) for control the upbeat nystagmus, and there was partial response.

Discussion

UBN is characterized by a downward drift of the eyes when the patient attempts to maintain the primary position of gaze. The drift is interrupted by saccadic movements which correct eye position back towards the center and give rise to the

upward beating appearance of the nystagmus.² Few cases of upbeat nystagmus were reported with low-grade astrocytoma, cystic tumor, extensive demyelination, pontine hemorrhage associated with lesion of the anterior cerebellar vermis, perihypoglossal and inferior olivary nuclei of the medulla, pontine tegmentum, brachium conjunctivum, midbrain, and brainstem diffusely.⁴⁻⁶

Medullary lesions invariably involve the perihypoglossal nucleus and adjacent medial vestibular nucleus, nucleus intercalates (NI), and ventral tegmentum, which contain projections from vestibular nuclei that receive inputs from the anterior semicircular canals.⁷⁻¹⁰ The perihypoglossal nucleus consists of three small subnuclei, the nucleus prepositus hypoglossi (NPH), the nucleus of Roller (NR), and nucleus intercalates.⁸ The NI, the most caudal of the perihypoglossal nuclei, has strong reciprocal connections with the NPH. It receives afferents from the medial and inferior vestibular nuclei and has numerous projections including those to the cerebellum and the ocular motor nuclei.¹⁰

The pathophysiology of UBN is not well known. Since most lesions were located inferiorly to the NPH in the posterior paramedian part of the medulla, it has at times been suggested that the NI, lying just caudally to the NPH, could be involved.^{8,10,11} Pierrot-Deseilligny et al. suggested that no obvious link with UBN can be found if the NPH or NI circuitry is considered.¹² By contrast, the NR appears to be a better candidate to play a role in upward vestibular eye movements. This small nucleus is located at the same caudal medullary levels as the NI, lying slightly anteriorly and medially to the superior part of this nucleus. Therefore, the NR was probably also damaged in most, if not all, of the caudal medullary lesions resulting in UBN.¹²

The caudal medulla may receive a collateral branch from the superior vestibular nucleus and project to the flocculus via a probably inhibitory pathway. UBN in the caudal medullary lesion may be ascribed to an impairment of this inhibitory pathway. The result would be disinhibition of the

inhibitory flocculovestibular neurons and overinhibition of the ventral tegmental tract with a slow downward deviation of the eye.¹²

In our case, symmetrical demyelinating lesion was found in the lower medulla around the IN and NR. We propose that UBN can be a manifestation of relapsing multiple sclerosis.

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