Loss of Vision Alone May Result in Seesaw Nystagmus

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A patient is described who developed seesaw nystagmus (SSN) associated with progressive severe vision loss due to cone-rod dystrophy. She was otherwise neurologically normal, and findings on magnetic resonance imaging of the brain were normal, including optic chiasm, meso-diencephalic junction, and brainstem. The literature is reviewed on neurologically normal patients with SSN and ocular vision loss, and the hypothesis is presented that SSN may become manifest as a result of vision loss alone, even in patients with chiasmal lesions, without disruption of central ocular motility pathways.

Key Words: Seesaw nystagmus—Cone-rod dystrophy—Retina—Nystagmus—Blindness.

Seesaw nystagmus (SSN) is characterized by alternating elevation and intorsion of one eye and simultaneous depression and extorsion of the other eye. Pendular SSN is usually found in patients with bitemporal hemianopia (BTH) and suprasellar masses (1). The specific cause of pendular SSN in these patients remains unclear: secondary involvement of central otolithic/vestibular pathways at the diencephalic–mesencephalic junction is usually assumed, but has been poorly documented. Because nonchiasmal structures seem to be uninvolued in many cases, purely sensory causes of the nystagmus have been postulated (1). We report the case of a neurologically normal patient who developed pendular SSN in association with vision loss, with normal brain magnetic resonance image. This further supports the hypothesis that patients may develop pendular SSN due to vision loss alone, without lesions of the rostral brainstem.

CASE REPORT

The patient is a woman with progressive decrease in central vision that began at age 23. At age 27, best corrected visual acuity was 20/100 in the right eye and 20/70 in the left eye. Clinical course and the findings on funduscopic examination and electroretinography were felt to be consistent with autosomal recessive cone-rod retinal dystrophy. Eye movements and ocular alignment were reported to be normal.

The patient's central vision deteriorated over years, and visual fields never showed BTH. Ophthalmologic evaluations documented normal eye movements until age 37, when “vertical nystagmus” was first noted. There was no history of head trauma. The patient was first seen by us at age 40. Visual acuity was hand motion at 2 feet (0.6 m) in each eye (OU). Funduscopic examination findings were unchanged from prior descriptions. Visual fields showed only preservation of small islands of vision in the periphery OU.

Ocular motility examination showed typical pendular SSN such that the supraducting eye intorted as the infra­ducting eye extorted. Electro-oculography demonstrated that the vertical amplitude of the nystagmus increased in upgaze and decreased in lateral gaze and downgaze. There was no nystagmus in the dark (Fig. 1). Other eye movements and the results of the remainder of the neurologic examination were normal.

Standard magnetic resonance imaging of the brain was performed, including T1- and T2-weighted and balanced sequences. High-resolution images of 3.5-mm thickness with 1.0-mm intervals were also obtained of the chiasm, midbrain, and thalamus by using T1 weighting, with and without gadolinium. There were no abnormalities.

DISCUSSION

It is generally assumed that patients with BTH caused by chiasmal lesions who develop SSN must also have damage to the rostral brainstem (2). The case reported here provides evidence that patients with pendular SSN and ocular or chiasmal vision loss need not have any lesion of the meso-diencephalic junction.

There are two prior reports in the literature of apparently neurologically normal patients who acquired SSN in association with severe ocular vision loss (3,4). Skull radiographic findings were normal in each, and more sophisticated neuroimaging was not available. In fact, Rucker (4) presumed that his patient had an "inflammatory reaction . . . in the brain" simply based on the presenta­
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FIG. 1. Electro-oculography showing vertical eye movements in various positions of gaze and under different lighting conditions. Torsional eye movements were not recorded. Note that the nystagmus is pendular and dysconjugate (arrows indicate that when the right eye is up, the left eye is down, although not precisely 180° out of phase). The frequency of the nystagmus is -3.5 Hz. The amplitude of the nystagmus increases in upgaze (second panel), and is greatly diminished in downgaze (third panel). The nystagmus is absent in the dark (fourth panel). Electrodes were centered, one just below the eyebrow and the other at the margin of the lower lid of each eye. The recording technique used alternating current with a low-frequency cutoff of 0.5 Hz, time constant of 0.3, with 60 Hz filtered out. Tracings could not be calibrated, because of the patient’s blindness and continuous eye movements.

ence of the nystagmus. The clinical course and normal magnetic resonance image of our patient strongly suggest that there was no rostral brainstem lesion.

The etiology of pendular SSN in patients with ocular vision loss is uncertain, but may be the same as in patients with BTH. SSN may occur congenitally (5), and patients with acquired blindness may develop congenital and/or neurologically localizing nystagmus (6). Also, SSN caused by a suprasellar mass may appear only when fixation is interrupted (7). As such, certain patients with “underlying” SSN may demonstrate the nystagmus only when fixation is impaired or when binocular fusion is limited by BTH.

The disappearance of the SSN in our patient in the dark, as has been reported with BTH (1), attests to the vision dependence but complex etiology of pendular nystagmus in patients with vision loss. Acquired horizontal pendular nystagmus has been shown to be due not simply to a delay in visual feedback nor only to a disorder of central vestibular mechanisms, but is likely due to abnormalities of central feedback circuits (8). Along these lines, Nakada and Kwee have proposed that disruption at the chiasm of visual input destined for the inferior olive nucleus may cause synchronous oscillations of floccular Purkinje cells, resulting in SSN (1).

In summary, we report a case in which neuroimaging demonstrated a normal brain in a patient with pendular SSN due to vision loss. It therefore seems reasonable to classify SSN similarly to periodic alternating nystagmus, as proposed by Cross et al. (6). Pendular SSN may occur in one of three scenarios: with loss of vision, congenitally, or with evidence of meso-diencephalic disease. Future studies of patients with BTH, SSN, and suprasellar masses should be aimed at determining the true incidence of meso-diencephalic involvement, so that the specific etiology of the SSN in each case can be determined.

REFERENCES