Unusual Monocular Pendular Nystagmus in Multiple Sclerosis

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Abstract: Two unusual cases of monocular pendular nystagmus in patients with multiple sclerosis are reported. One patient showed regular horizontal oscillations of the right eye in abduction, associated with right abduction paresis. The second patient had a similar abnormal eye movement of the left eye in adduction, with partial left internuclear ophthalmoplegia. Such eye position-dependent monocular pendular nystagmus provides new insights into pathogenic mechanism for acquired pendular nystagmus. Different mechanisms are discussed such as the combination of paresis and commonly accepted hypothesis of dysfunction of visual and/or motor feedback loops in the ocular motor neural network.

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The most frequently reported eye movement disorders in multiple sclerosis (MS) are internuclear ophthalmoplegia, isolated ocular motor palsy, gaze-evoked nystagmus, and pendular nystagmus (1). Pendular nystagmus is characterized by quasi-sinusoidal oscillations of the eyes along a horizontal, vertical, or torsional direction. Pendular nystagmus in MS may be asymmetrical and in this case, most frequently of greater amplitude in the eye with poorer vision (2). Strictly, monocular forms of pendular nystagmus in MS have been reported, either associated with chronic visual deficit following optic neuropathy (2,3) or observed during convergence (4). We report 2 patients with MS with monocular horizontal pendular nystagmus, which was specifically triggered in eccentric gaze.

CASE REPORTS

Case 1
A 40-year-old woman was evaluated with a 5-year history of relapsing-remitting MS. The first manifestation of the de- myelinating disease was a right sixth nerve paresis, and since then, she complained of diplopia in right gaze. Due to recurrent episodes of left lower limb paresthesia, she was treated with azathioprine. She was referred to the neuro-ophthalmology unit for daily episodes of paroxysmal oscillopsia in her right eye.

Her expanded disability status scale (EDSS) score was 2 (minimal disability). Visual acuity was 20/20 in each eye, with no relative afferent pupillary defect and normal fundi. Automated perimetry, static contrast sensitivity values, and scores for the Farnsworth-Munsell D-15 Hue test and Ishihara color plates for each eye were normal. Visual evoked potentials were normal bilaterally. Ocular motor examination showed right esotropia with abduction paresis of the right eye. Horizontal pendular nystagmus of the right eye was observed during right gaze. The nystagmus persisted as long as the eccentric gaze was maintained and ceased when the patient directed her eyes to primary position or left gaze. Pendular nystagmus was dampened at near fixation, convergence, and with a 4-diopter base-out prism placed in front of the right eye. Gabapentin (up to 900 mg/d for 2 months), carbamazepine (up to 600 mg/d for 2 months), or clonazepam (up to 1 mg/d for 2 months) yielded no benefit.

Case 2
A 42-year-old woman with a 5-year history of relapsing-remitting MS was referred to the neuro-ophthalmology unit...
for monocular oscillopsia. This occurred due to the development of pendular nystagmus in the left eye during adduction of that eye. There was no improvement following a course of systemic steroids. Two months later, the patient complained of decreased vision. Visual acuity was found to be 20/50 in each eye, and visual field testing disclosed centrocecal scotoma in the right eye and central scotoma in her left eye. Felt to have bilateral optic neuritis, a course of steroids was prescribed and interferon therapy was started.

Her most recent EDSS score was 1 (no disability). Visual acuity was 20/25, right eye, and 20/32, left eye. Static central visual field (Metrovision®, Péréniches, France) disclosed decreased macular threshold of the right eye and a mild global defect on the left. Static contrast sensitivity was subnormal for the left eye with 2 of the 6 tested spatial frequencies. Scores of Farnsworth-Munsell D-15 Hue test and Ishihara color plates test showed numerous errors for her left eye. Visual evoked potentials showed bilateral increase of latency with P100 value at 168 millisecond, right eye, and 176 millisecond, left eye. Ocular motor examination disclosed bilateral horizontal gaze-evoked nystagmus. In far right gaze, a monocular horizontal pendular nystagmus of her left adducting eye was observed, persisting as long as the adduction was maintained. Left adducting saccadic velocity was diminished, consistent with left internuclear ophthalmoplegia.

**DISCUSSION**

We describe 2 patients with MS complaining of chronic monocular oscillopsia brought about by an unusual form of monocular pendular nystagmus. Both patients presented some clinical features consistent with the common forms of pendular nystagmus observed in MS in terms of frequency (4–5 Hz), small amplitude (5), and for Case 2, in association with optic neuropathy (2–4,6). It may also be seen in the setting of normal optic nerve function (7,8) as in our Case 1. Our 2 cases are unusual in that their monocular pendular nystagmus was observed only in eccentric horizontal gaze. While monocular nystagmus of the abducting eye is observed with internuclear ophthalmoplegia in MS, it is of jerk and not pendular form. Monocular adduction pendular nystagmus can be observed in MS but has only been reported with convergence (4).

Explanations for pendular nystagmus in patients with MS involve abnormal delays in feedback loops that control eye stability. First, a role of a persistent delay of visual...
FIG. 2. Case 2. Horizontal eye position recording in primary gaze, eccentric right gaze, and during convergence. A monocular pendular nystagmus of the adducting left eye occurs in right gaze, and a binocular pendular nystagmus is triggered with convergence. The nystagmus is of 3.5 Hz frequency and 2° mean amplitude. Binocular gaze-evoked nystagmus is also seen on right gaze. Positive values: gaze right; negative values: gaze left.
feedback secondary to demyelination of the optic nerve has been proposed (2). This hypothesis is supported by the previous reports of large oscillations occurring in an eye having a severe optic neuropathy (2,4). This could explain the monocular nystagmus in our Case 2 but does not account for the nystagmus being triggered by eccentric gaze. Moreover, this proposal has been challenged by experimental data in patients showing persistence of the nystagmus in darkness (9), and in patients with MS, there is no change in the nystagmus even with prolongation in latency of visually guided eye movements (10). A second hypothesis involves a role of abnormal delay in the ocular motor feedback loops secondary to demyelination of central neurons. Instability in motor feedback could involve the ocular motor neural integrator, as suggested by transient suppression of the nystagmus following saccades (8). Our data did not allow us to evaluate the effect of saccades on pendular nystagmus. However, the theory of an unstable neural integrator could lead to triggering of nystagmus with eccentric gaze. The onset of nystagmus during convergence, such as observed in our Case 2, also supports the concept of instability in motor feedback loops involving the vergence system (4).

Finally, there may be a role for ocular motor paresis in precipitating monocular pendular nystagmus. In Case 1, demyelination involved the sixth nerve fascicle and in Case 2, the left medial longitudinal fasciculus. It is well known that peripheral motor paresis can trigger central adaptive changes (9). This consists of detecting visual errors due to ocular motor paresis and increasing the innervation to the paretic eye through central feedback loops (11). In our 2 patients, these adaptive changes took place in a neural network affected by demyelination, and instability in these feedback loops might have triggered monocular eye position–dependent pendular nystagmus.

REFERENCES


TABLE 1. Summary of eye movement recording for our 2 patients

<table>
<thead>
<tr>
<th></th>
<th>Case 1</th>
<th>Case 2</th>
<th>Normal Limits</th>
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<tbody>
<tr>
<td></td>
<td>Left Eye</td>
<td>Right Eye</td>
<td>Left Eye</td>
</tr>
<tr>
<td>Smooth pursuit</td>
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<td>Horizontal gain</td>
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<td>30°</td>
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<td>Vertical gain</td>
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<td>0.40</td>
<td>0.2–0.8</td>
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<tr>
<td>OFI 0.25 Hz (gain)</td>
<td>0.12</td>
<td>0.12</td>
<td>&lt;0.10</td>
</tr>
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OFFI, ocular fixation index; VOR, vestibulo-ocular reflex.