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Acquired monocular nystagmus in chiasmal glioma—a video-oculographic study

Michael C. Brodsky, MD,[a,b] and Laura A. Torrado, MD[a]

We report the case of a 9-year-old girl with chiasmal glioma and longstanding monocular nystagmus, in whom video-oculography showed a disconjugate binocular nystagmus indistinguishable from spasmus nutans. The "acquired monocular nystagmus" associated with chiasmal glioma is actually a binocular nystagmus that is indistinguishable from spasmus nutans, except that it lacks the associated head nodding and torticollis. Superimposed visual loss in one eye predisposes to acquired monocular nystagmus and explains the absence of the other components of the spasmus nutans triad.

Spasmus nutans is a benign, self-limiting clinical entity characterized by an asymmetric nystagmus, head nodding, and torticollis.[1] It generally begins between 6 and 12 months of age and resolves over months to years.[1] The nystagmus is of low amplitude and high frequency, and is asymmetrical in the two eyes. Most cases are idiopathic, but some children are found to have congenital retinal dystrophies.[2] It is the critical association with chiasmal glioma that dictates clinical management.[3-6] In most cases, chiasmal glioma is signaled by associated findings, such as a relative afferent pupillary defect, optic atrophy or disk swelling, large head size, café au lait spots, and coexistent neurological dysfunction.[4] Rarely, however, these associated neurological abnormalities are absent.

In some children, chiasmal glioma can present as acquired monocular nystagmus.[5] Farmer and Hoyt[5] described 10 children with monocular nystagmus in infancy or early childhood. Neuroimaging revealed that 6 of these patients had chiasmal glioma, whereas 4 had no tumor and were diagnosed as having spasmus nutans. Acquired monocular nystagmus can also accompany unilateral congenital visual loss and resolve following successful treatment of associated amblyopia.[6]

To our knowledge, eye movement recordings have not been performed in children with acquired monocular nystagmus due to chiasmal glioma. Pathogenetically, these high-resolution measurements are critical, because there is no known mechanism by which the human ocular motor pathways can generate a purely monocular nystagmus. To elucidate the true nature of this condition, we obtained video-oculography for a 9-year-old girl with chiasmal glioma who had longstanding isolated monocular nystagmus.

Case Report

A 9-year-old girl with chiasmal glioma had been followed at the Mayo Clinic Department of Ophthalmology for a monocular nystagmus of the left eye that was first noted at 6 years of age. She had no stigmata of neurofibromatosis 1, no family history of nystagmus, and was neurodevelopmentally normal.

On ophthalmologic examination, visual acuity was 20/20 in the right eye and 20/250 in the left eye (reduced from 20/60 at age 6). She had a normal pupillary response to light in the right eye and a sluggish pupillary response to light, with a 2+ relative afferent pupillary defect, in the left eye. A pendular vertical nystagmus of moderate amplitude and frequency was noted in the left eye only (Video 1, available at jaapos.org). SensoMotoric instruments (SMI) video-oculography showed a disconjugate binocular pendular nystagmus of low amplitude and high frequency in the right eye and high amplitude and low frequency in the left eye (Figure 1). Retinoscopy showed a mildly hyperopic refractive error (+1.00 sphere) in both eyes. Retinal examination disclosed small optic disks, with a bilateral band atrophy that was worse in the left eye. Humphrey 30-2 visual field testing disclosed bitemporal hemianopia.

Magnetic resonance imaging revealed enlargement of a homogeneously enhancing mass involving the optic chiasm, with extension along the intracranial optic nerves bilaterally (Figure 2). The superior aspect of the tumor was larger on the left, corresponding to the more severe visual loss and optic atrophy in the left eye. The tumor exerted a local downward mass effect on the pituitary gland, but no extension to the hypothalamus was found, endocrinologic testing disclosed no abnormalities.

Discussion

Gottlob and colleagues[7] used eye movement recording to define the electro-oculographic waveform of spasmus nutans as a disconjugate binocular pendular nystagmus with the two eyes oscillating out of phase. In our patient with a longstanding chiasmal glioma, video-oculography showed the isolated monocular nystagmus to be a subclinical binocular nystagmus that conformed to the waveform

Author affiliations: Departments of [a]Ophthalmology and [b]Neurology, Mayo Clinic, Rochester, Minnesota
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Correspondence: Michael C. Brodsky, MD, Mayo Clinic 200 First St SW, Rochester, MN, 55905 (email: Brodsky.michael@mayo.edu).
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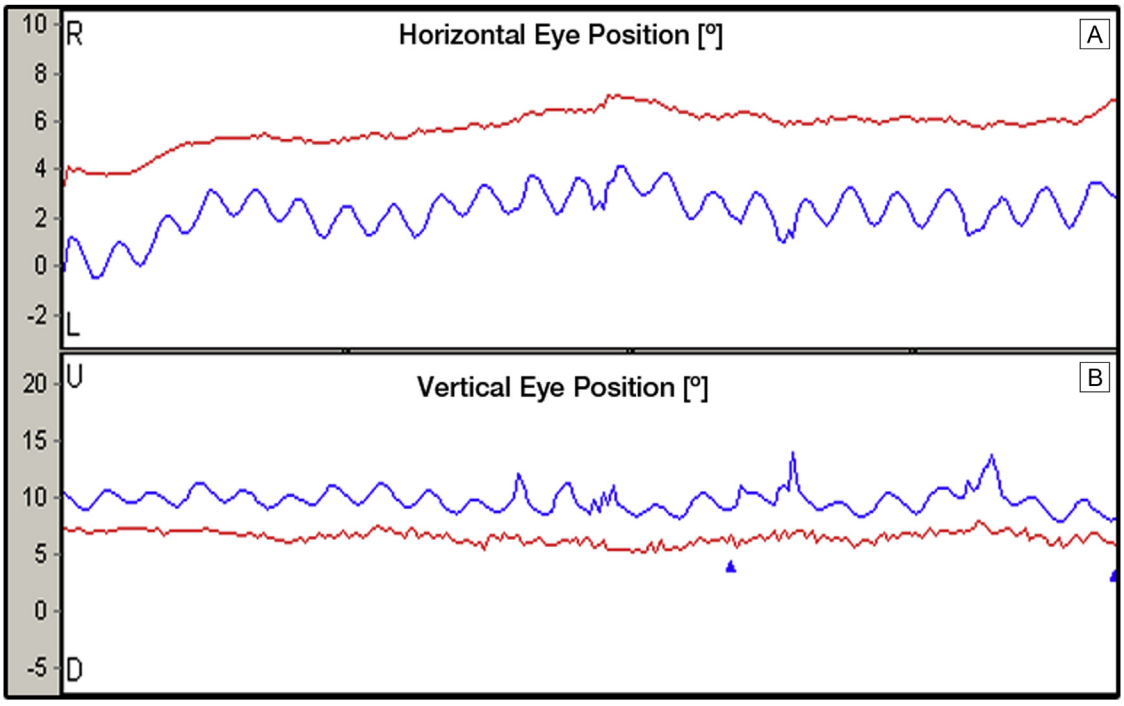

FIG 1. Video-oculography showing disconjugate pendular horizontal (A) and vertical (B) nystagmus of greater amplitude but lower frequency in the left eye (blue line) than in the right eye (red line). The vertical axis indicates degrees of movement. *D*, down; *L*, left; *R*, right; *U*, up.

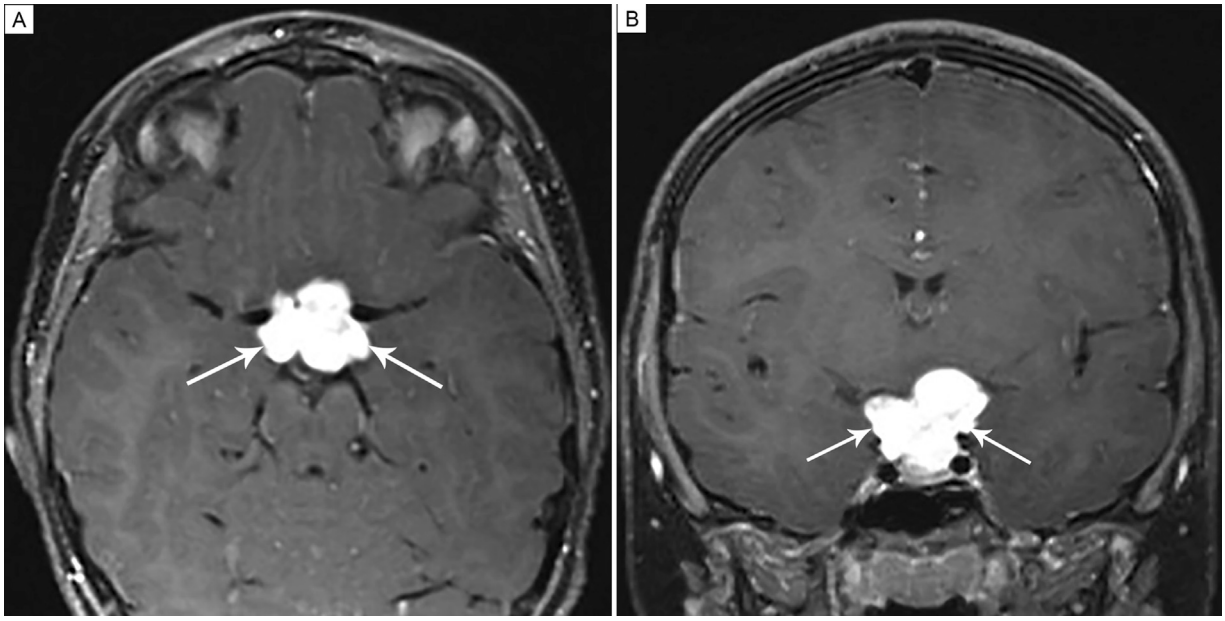

FIG 2. Gadolinium-enhanced T1-weighted magnetic resonance imaging (A, axial; B, coronal) showing chiasmal glioma with superior extension on the left side: the arrows denote the lateral margin of the tumor.

of spasmus nutans: unlike in spasmus nutans, however, our child had no head nodding or torticollis under 6 years of observation.

We attribute our child's acquired monocular nystagmus to superimposed monocular visual loss in the eye with nystagmus, which could explain the delayed onset of the nystagmus at age 6. In the series by Farmer and Hoyt,[5] 4 of the 6 children with monocular nystagmus due to chiasmal glioma also had decreased visual acuity and optic atrophy in the eye with the nystagmus. The finding that reversible monocular nystagmus can rarely accompany amblyopia further suggests that monocular visual loss

may be a contributing factor.[6] In the setting of chiasmal glioma, the development of monocular nystagmus may require a "two hit" scenario of both chiasmal glioma and superimposed visual acuity loss in one eye. The clinical phenotype of monocular nystagmus may arise from asymmetric chiasmal involvement of crossed fibers from one eye and both crossed and uncrossed fibers from the other eye. This mechanism would explain why monocular nystagmus in this age group is so often a harbinger of chiasmal glioma. The superimposed loss of visual acuity loss in one eye also explains the absence of head nodding and torticollis in children with acquired monocular nystagmus, because these latter components of the spasmus nutans triad have been found to be compensatory for binocular vision.[8] The documented association of spasmus nutans with amblyopia could similarly explain the less common presentation of monocular nystagmus in infants with "isolated" spasmus nutans.

Spasmus nutans may resolve clinically, even in children with chiasmal glioma.[9] However, some degree of residual nystagmus often remains detectable on eye movement recording following clinical resolution.[10] This discordance reflects the higher sensitivity of eye movement recording for detecting subclinical nystagmus. In both spasmus nutans and chiasmal glioma with monocular nystagmus, eye movement recording discloses the subclinical component of the binocular oscillation.

We conclude that the "acquired monocular nystagmus" of chiasmal glioma may actually be a highly asymmetrical disconjugate nystagmus that conforms to the waveform of spasmus nutans. In the child with chiasmal glioma, this binocular nystagmus can manifest as acquired monocular nystagmus in the eye with more severe visual loss.

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