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Upbeat-torsional nystagmus and contralateral fourth nerve palsy due to unilateral dorsal ponto-mesencephalic lesion

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Abstract

The central projections of the anterior semicircular canals are thought to be conveyed from the vestibular nuclei to the ocular motor nuclei in the midbrain by three distinct brainstem pathways: the medial longitudinal fasciculus, crossing ventral tegmental tract, and brachium conjunctivum. There is controversy as to whether upbeat nystagmus could result from lesions involving each of these pathways. We report a 52-year-old man who presented with a contralesional fourth nerve palsy and primary position upbeat-torsional nystagmus due to a small unilateral dorsal ponto-mesencephalic lymphomatous deposit. We postulate that the upbeat-torsional nystagmus was caused by involvement of the brachium conjunctivum, which lies adjacent to the fourth nerve fascicles at the dorsal ponto-mesencephalic junction, but we cannot exclude involvement of the crossing ventral tegmental tract. Our observations suggest that, in humans, excitatory upward-torsional eye movement signals from the anterior semicircular canals could be partly conveyed to the midbrain by the brachium conjunctivum.

Keywords

Upbeat Nystagmus; Torsional Nystagmus; Fourth Nerve Palsy; Pons; Mesencephalon; Brachium Conjunctivum

Introduction

Fourth nerve palsy is rarely caused by lesions involving the fourth nerve nucleus or fascicles in the brainstem.¹ When it does occur, the palsy is contralesional, as the nerve decussates after it emerges from the brainstem. The palsy may be isolated or associated with other brainstem signs, such as primary-position upbeat nystagmus.² We report a patient who presented with a contralesional fourth nerve palsy and primary-position upbeat-torsional nystagmus due to a small unilateral lesion at the dorsal ponto-mesencephalic junction.

Case Report

A 52-year-old man presented with a 1-week history of diplopia and oscillopsia. The diplopia was vertical-torsional and increased by both leftward gaze and rightward head tilt. The oscillopsia was vertical and increased by upward gaze. He had a past history of diffuse large B-cell lymphoma, with no known central nervous system involvement, and had been in remission for 1 year following an autologous stem cell transplant.

Examination of his visual system and pupils was normal. Eye movement examination revealed primary-position nystagmus, with upward and clockwise quick phases from the patient's point of view. The nystagmus was predominantly vertical in leftward gaze and predominantly torsional in rightward gaze. It increased with upward gaze, but was not modulated by convergence or changes in head position relative to gravity. There was also a right hypertropia that increased with leftward gaze and rightward head tilt, consistent with a right fourth nerve palsy. There was no internuclear ophthalmoplegia and neurologic examination was otherwise unremarkable.

Magnetic resonance imaging of brain with gadolinium showed a discrete enhancing lesion at the ponto-mesencephalic junction, adjacent to the cerebral aqueduct, on the left (fig. 1A). Increased signal in the adjacent brainstem tegmentum, consistent with vasogenic edema, was noted on the T2-weighted imaging (fig. 1B). CSF protein level was increased (164mg/dL). CSF microscopy revealed an increased white cell count with atypical lymphocytes, but no organisms on Gram stain, consistent with a central nervous system relapse of lymphoma.

Within days of starting treatment with high-dose corticosteroids, intrathecal methotrexate, and systemic chemotherapy, there was improvement in the ocular motor deficit and reduction in both the size of the lesion and extent of edema on imaging. By the time of discharge, 1 month later, he was asymptomatic and had no deficit.

Discussion

Primary-position upbeat nystagmus is thought to result from disruption of central pathways conveying compensatory upward eye movement signals from the vestibular nuclei to the ocular motor nuclei in the midbrain. When it is modulated by vergence and changes in head position relative to gravity, disruption of central otolithic projections is the likely cause.³ In our patient, convergence and changes in head position had little effect on the nystagmus, suggesting that it was caused by disruption of central semicircular canal projections. Indeed, the direction of nystagmus slow phases implied that disruption of left anterior canal projections was the cause.

While horizontal and posterior canal projections are conveyed to the ocular motor nuclei in the midbrain solely by the MLF, anterior canal projections are conveyed by the crossing ventral tegmental tract (CVTT)^{4,5} and brachium conjunctivum (BC),⁶⁻⁸ as well as the MLF.^{9,10} There is controversy as to whether primary-position upbeat nystagmus could result from lesions involving each of these pathways and it has been postulated that many cases might arise due to involvement of the CVTT rather than the BC.^{9,11} While the exact location of the CVTT is not known in humans, primary-position upbeat nystagmus has been observed with ventral ponto-mesencephalic lesions,⁹ where the CVTT is known to be located in the cat.^{4,5} If the CVTT is located ventrally at the ponto-mesencephalic junction in humans, it should be intact in our patient. Indeed, our patient's lesion is likely to have involved the left BC, although the CVTT could have been affected by vasogenic edema in the tegmentum (fig. 1). It is also possible that the MLF could have been disrupted in our patient, although there was no clinical evidence of internuclear ophthalmoplegia to support this notion.

The BC has been implicated in prior cases of primary-position upbeat nystagmus, although the lesion often involved other adjacent structures.¹²⁻¹⁴ One patient with a contralesional fourth nerve palsy and primary-position upbeat nystagmus also had a small unilateral dorsal ponto-mesencephalic lesion.² As in our patient, that patient's fourth nerve palsy resulted from involvement of the fourth nerve fascicles prior to their exit from the brainstem and the authors postulated that the nystagmus likely resulted from involvement of the BC. Projections from the vestibular nuclei to the midbrain have been found in the BC in prior

anatomical studies in rabbit,^{6,7} cat,⁸ and primate,¹⁵ although they were not observed in a recent primate study.¹⁶ In that study, projections were seen only in the CVTT, which followed a parallel, yet more ventral, course to the BC in the brainstem.¹⁶ Although a lesion of the CVTT cannot be excluded, given the findings of anatomical studies combined with our observation that a unilateral dorsal ponto-mesencephalic lesion produced nystagmus in the plane of the ipsilesional anterior canal, we postulate that excitatory upward-torsional eye movement signals from the anterior semicircular canals could be partly conveyed to the midbrain by the BC.

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Figure 1.

(A) Axial T1-weighted MRI with gadolinium contrast shows an enhancing unilateral dorsal ponto-mesencephalic lesion on the left. (B) Increased signal in the adjacent brainstem tegmentum, consistent with vasogenic edema, is seen on the T2-weighted MRI.