Pearls & Oy-sters: Paroxysmal ocular tilt reaction

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In skew deviation, one eye is higher than the other, causing vertical diplopia. Skew deviation is a pre-nuclear disorder resulting from imbalance in otolith input to the oculomotor system, produced by unilateral lesions anywhere from the otoliths to the interstitial nucleus of Cajal in the rostral midbrain,¹ and as such is one component of the ocular tilt reaction.^{2,3} We report a woman with a history of fluctuating diplopia after resection of a left thalamic angioma. She was shown to have intermittent paroxysmal ocular tilt reaction episodes superimposed on a tonic ocular tilt reaction.

CASE REPORT A 64-year-old woman initially presented at age 55 with malaise, headache, and imbalance, leading to the discovery on neuroimaging of a vascular malformation in the left thalamus. Following resection, she had persistent vertical diplopia on downgaze, and episodes several times a day of vertical diplopia in other gaze positions, associated with a sense of twisting of her eyes. For 9 years she had been on both gabapentin and carbamazepine, with uncertain effect: she reported some modest reduction in episodes with increasing the dose of gabapentin from 600 mg/day to 1,200 mg/day. Visual acuity, tests of color vision with pseudo-isochromatic plates, visual fields by confrontation, pupils, and fundus examinations had normal results. Palpebral fissures were symmetric. Ductions were full in all directions. Fixation was steady. Pursuit and cancellation of the vestibulo-ocular reflex were normal. Saccades were rapid and accurate. Cover testing showed that she was orthophoric in primary position (i.e., eyes aligned in straight-ahead gaze) but had a small 3-prism diopter left hypertropia in downgaze. Maddox wing testing showed a small 1-degree excycloptropia (top of the eyes rotated away from each other). She had a right head tilt. The remainder of the neurologic examination had normal results. During her examination, she developed episodes of conjugate counterclockwise torsion (i.e., the upper pole of both eyes tilted toward her left shoulder) with right hypertropia, lasting 1 to 3 seconds, at variable intervals (see video).

MRI showed a large cavernous hemangioma in the left mesodiencephalic junction (figure).

DISCUSSION Ocular tilt reaction consists of skew deviation, ocular torsion, head tilt, and deviation of

the subjective visual vertical, all tilted toward the lower (hypotropic) eye,^{2,3} with the side of the tilt named for the side of the hypotropic eye. It can be produced experimentally by stimulation of the utricular nerve,⁴ and represents a normal otolithic-ocular response to lateral displacement of the linear acceleration vector, as happens when one travels around a curve in a vehicle. The pathway for the otolithic-ocular response projects from the vestibular end-organ to the vestibular nuclei in the medulla and on to the interstitial nucleus of Cajal in the midbrain. This pathway decussates in the pons: hence static ocular tilt reactions from hypofunction are ipsiversive (lower eye on the side of the lesion) with peripheral vestibular and pontomedullary lesions and contraversive with pontomesencephalic lesions.¹ The direction of the deviation produced by paroxysmal hyperfunction in this pathway (i.e., irritative lesions), on the other hand, is in the opposite direction.^{5,6} Our patient, who has a lesion in the vicinity of the left interstitial nucleus of Cajal, illustrates this point. She has a tonic right head tilt and mild left hypertropic skew deviation, which represents a partial ocular tilt reaction—partial in that there is no significant torsion at baseline—that is contraversive with respect to the lesion (head is tilted away from the side of the lesion). Superimposed on this, she develops paroxysmal episodes of left ocular tilt reaction, with a right hypertropic skew deviation and counterclockwise torsion.

Paroxysmal ocular tilt reaction is distinctly rare. One patient with multiple sclerosis had pendular nystagmus and a paroxysmal ocular tilt reaction that improved with carbamazepine.⁵ Another patient with a lesion in the vicinity of the left interstitial nucleus of Cajal presented with right hypertropia, conjugate torsion, left head tilt, and nystagmoid eye movements.⁶ Three patients were described with episodic ocular torsion and skew deviation due to mesodiencephalic lesions, in whom the conjugate ocular

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(<http://www.neuroophthalmology.ca/cases/case33.html>). The video was adapted with permission from the Web site of the Canadian Neuro-Ophthalmology Group (<http://www.neuroophthalmology.ca/>).

Figure T2-weighted axial and coronal images showing hypointense lesion of the left mesencephalon, in the vicinity of the interstitial nucleus of Cajal

torsion was initiated by a torsional fast eye movement.⁷ Similar to our case, one of these patients had a cavernoma in the right mesodiencephalic region: there was a baseline ipsilateral hypertropic skew deviation with episodes of clockwise ocular torsion and dystonic movements in the contralateral limbs. Such vascular lesions may be particularly likely to produce both tonic hypofunction and transient hyperfunction.

CLINICAL PEARLS

- Ocular tilt reaction is a prenuclear disorder due to imbalance in otolithic pathways anywhere from the vestibular organ to the midbrain.
- Ocular tilt reaction is the triad of 1) skew deviation, a prenuclear vertical misalignment of the eyes, 2) head tilt toward the hypotropic eye, and 3) ocular torsion. If all three components are present, ocular tilt reaction is complete. Partial forms also exist.
- Ocular torsion in ocular tilt reaction usually consists of incyclotorsion of the upper eye (the upper pole of the eye is rotated nasally) with excyclotorsion of the lower eye (the

upper pole of the eye is rotated temporally); thus, the tops of both eyes are rotated toward the lower ear. (In contrast, IV nerve palsy, the most common cause of vertical misalignment, involves excyclotorsion of the upper eye.)

- As the pathway for the otolithic-ocular response decussates in the pons, lesions of the lower pons/medulla cause an ipsiversive (ipsilateral eye is hypotropic) ocular tilt reaction, whereas lesions of the upper pons/midbrain cause a contraversive one.
- Whereas most cases of ocular tilt reaction are tonic and due to a decrease in tonic neural activity, paroxysmal ocular tilt reaction is due to intermittent unilateral hyperfunction, with tilt in the direction opposite to that of tonic ocular tilt reactions.

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