Pearls & Oy-sters: Vertical Diplopia and Ocular Torsion

Peripheral vs Central Localization

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Abstract

We describe a case with torsional deviation of the eyes from a brainstem lesion. Torsional eye movement refers to changes in the position of the eyes in the roll plane around the visual axis. When the head is tilted laterally (that is, rolled toward the shoulder), the eyes roll in the opposite direction as part of the torsional vestibulo-ocular reflex known as the ocular counter-roll. Pathologies that affect the otolith-ocular pathway can lead to a torsional deviation of the eyes as part of the ocular tilt reaction (OTR) that also causes vertical deviation of the eyes (skew deviation) and head tilt. Lesions caudal to the pontomedullary junction (such as the labyrinth, eighth cranial nerve, or vestibular nucleus) result in an OTR with ipsiversive torsional deviation, whereas lesions rostral to the junction result in an OTR with contraversive torsional deviation. Furthermore, torsional deviation of the eyes in the OTR is conjugate (incyclotorsion in the higher eye and excyclotorsion in the lower eye), whereas torsional deviation from ocular palsy in peripheral lesions is disconjugate (e.g., exyclotorsion only in the higher eye with superior oblique palsy). Therefore, the pattern of torsional eye deviation can be helpful in localizing the lesion. Several techniques including fundus photography, double Maddox rod testing, optical coherence tomography, and video-oculography are used to measure torsional eye position.

Pearls

- Torsional deviation of the eyes is a component of ocular tilt reaction (OTR) in lesions affecting the otolith-ocular pathways.
- The OTR consists of a triad that also includes skew deviation and a compensatory head tilt.
- Caudal lesions (within the brainstem or labyrinth) can result in an ipsiversive torsional deviation of the eyes (the top poles roll toward the lesion side), whereas rostral brainstem lesions can result in a contraversive deviation of the eyes (the top poles roll away from the lesion side).

Oy-sters

- The torsional deviation of the eyes in OTR is conjugate (incyclotorsion in the higher eye and excyclotorsion in the lower eye).
- Torsional deviation in superior oblique palsy is nonconjugate (excyclotorsion only in the affected eye).

Case Report

A 30-year-old woman with a history of a CNS demyelinating disorder presented with constant vertical binocular diplopia and dizziness triggered by head movement or standing, which improved with sitting. Immediately before her encounter, there was worsening of symptoms, new vision changes, and right-hand weakness.

Her general neurologic examination was remarkable for right upper extremity weakness and dysmetria. The vestibular/ocular motor examination was notable for spontaneous upbeat nystagmus

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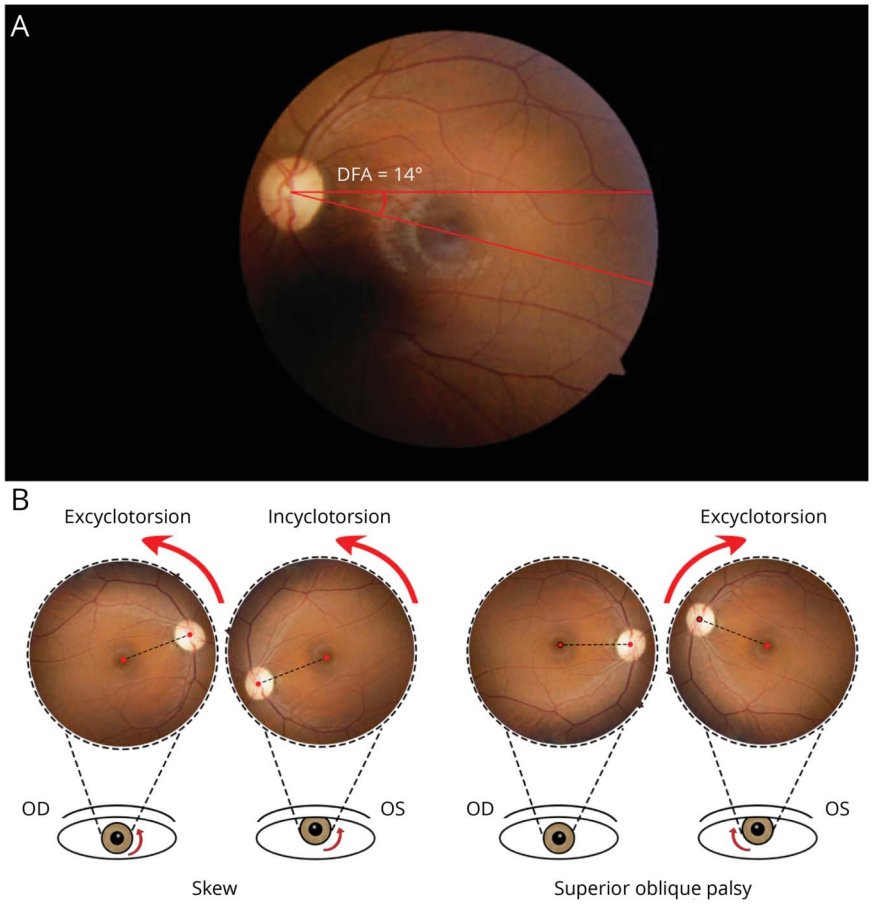
(UBN) and saccadic dysmetria. She also demonstrated torsional deviation of the eyes with right (OD) incyclotorsion and left (OS) excyclotorsion on fundus photography (Figure 1A). An MRI of the brain with gadolinium revealed enhancing lesions in the left middle cerebellar peduncle and pontomedullary junction. Lumbar puncture showed a mild pleocytosis (white blood cell count = 2), 2 CSF-specific oligoclonal bands, and normal glucose and protein levels. She underwent treatment with high-dose steroids with improvement of her symptoms; she was discharged with an outpatient follow-up.

Discussion

Torsional eye movement refers to changes in the position of the eyes in the roll plane around the visual axis.[1] When the head is tilted laterally (toward the shoulder), the eyes roll in the opposite direction as part of the torsional vestibulo-ocular reflex; this is called the ocular counter-roll (OCR).[1-3] During head tilt, the dynamic OCR is driven by inputs from the semicircular canals and otolith organs; it consists of a torsional nystagmus with slow phases away from and quick phases toward the direction of head tilt.[4] A sustained head tilt, however, results in a static OCR, which is typically 10%–25% of the head tilt.[1,2] This OCR response is mainly driven by otolith inputs through the projections from the labyrinths to the ocular motor nuclei (Figure 2A). Within this pathway, the graviceptive inputs reach the ipsilateral vestibular nucleus through the superior division of the vestibular nerve and the eighth cranial nerve.[5-7] The projections from the vestibular nucleus subsequently decussate at the pontomedullary junction and ascend through the contralateral medial longitudinal fasciculus (MLF) to innervate the ocular motor nuclei that drive the OCR response.[6-8] For example, with a leftward head tilt, the rightward OCR consists of (1) elevation and incyclotorsion of the left eye through activation of the superior oblique and superior rectus muscles and (2) depression and excyclotorsion of the right eye through activation of the inferior oblique and inferior rectus muscles.[5,8]

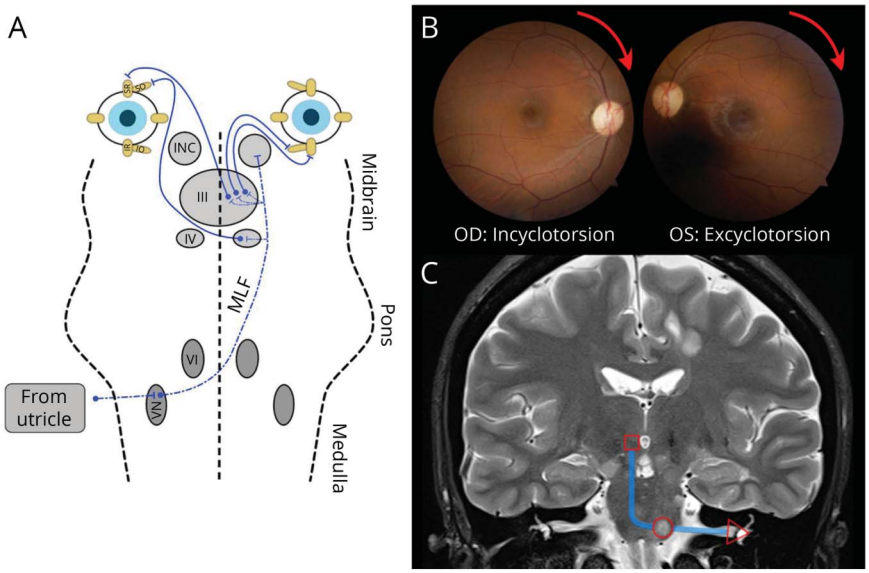
Pathologies that affect the otolith-ocular pathway can lead to torsional deviation of the eyes as part of the ocular tilt reaction (OTR).[5,8,9] The OTR also consists of a vertical misalignment of the eyes (skew deviation) with vertical diplopia and a lateral head tilt.[8] In such cases, torsional deviation presents with the top poles of the eyes rolled toward the side of the lower eye. As a sensitive clinical marker of vestibular tone imbalance, a complete or partial OTR can be

Figure 1 Comparing Ocular Torsion in Central vs Peripheral Lesions



(A) Fundus photograph of this patient's left eye demonstrating the estimation of the DFA to determine the degree of ocular torsion. (B) Illustration of ocular misalignment and resulting ocular and fundus torsion seen in a skew deviation and a superior oblique palsy. In a fourth nerve palsy, the higher eye is excyclortorted, whereas the fellow eye has no torsion (no OCR). However, in a skew deviation, the higher eye is inclyclotorted, whereas the fellow eye is excyclortorted—producing an OCR. DFA = disc-foveal angle; OCR = ocular counter-roll; OD = oculus dextrus; OS = oculus sinister.

Figure 2 Understanding the Utriculo-Ocular Motor Pathway Through Multiple Modalities



(A) Basic anatomy of the utriculo-ocular motor pathway, starting with fibers from the utricle to ocular motor nuclei and interstitial nucleus of Cajal. Reused with permission by the creator Daniel R. Gold, DO (Johns Hopkins School of Medicine), from the Neuro-ophthalmology Virtual Education Library: NOVEL (online) available at: novel.utah.edu/Gold/. Adaptations are themselves works protected by copyright. To publish this adaptation, authorization must be obtained both from the owner of the copyright in the original work and from the owner of copyright in the translation or adaptation. (B) Fundus photographs demonstrating a left OCR (left eye excyclotorsion and right eye incyclotorsion) from a left medullary lesion as estimated by the DFA method. (C) A coronal FLAIR sequence on MRI highlighting the schematic graviceptive pathway (blue line) from the utricle (red triangle) to the INC (red square) and the left pontomedullary lesion (red circle) that produced the patient's left OCR. DFA = disc-foveal angle; FLAIR = fluid-attenuated inversion recovery; III = oculomotor nuclei; INC = interstitial nucleus of Cajal; IO = inferior oblique; IR = inferior rectus; IV = trochlear nuclei; MLF = medial longitudinal fasciculus; OCR = ocular counter-roll; OD = oculus dextrus; OS = oculus sinister; SO = superior oblique; SR = superior rectus; VI = abducens nuclei; VN = vestibular nuclei.

seen with lesions affecting the otolith-ocular pathway from the labyrinths to different levels of the brainstem.[5,7-9] With central lesions, OTR persists longer and thus can be valuable for their localization, especially in patients with other minor or rapidly improving neurologic deficits. In particular, lesions caudal to the decussation of MLF fibers within the pontomedullary junction, such as the labyrinth, vestibular nerve, or vestibular nucleus will result in an *ipsiversive* OTR.[4,5] For example, patients with Wallenberg syndrome involving the vestibular nuclei may present with an ispiversive torsional deviation of the eyes (the top poles rolled toward the lesion side). In such cases, the skew deviation may emerge with the lower eye on the side of the lesion, along with a compensatory head tilt toward the lesion side. By contrast, because projections in the MLF from the vestibular nuclei in the medulla cross the midline before ascending to the ocular motor nuclei and the midbrain, skew deviation with lesions in the rostral pons or midbrain is usually associated with an ipsilateral hypertropia (elevated eye on the side of the lesion) and contralateral head tilt. These patients show a *contraversive* torsional deviation of the eyes (the top poles roll away from the lesion side).

Torsional eye position can be quantified using techniques such as fundus photography, optical coherence tomography (OCT), video-oculography–based OCR (vOCR) measurement, and double Maddox rod testing.[2,3,10] Fundus photography and OCT are useful tools for the evaluation of torsional deviation. In these methods, the angle between the optic disc and fovea, known as the disc-center-fovea angle (DFA), is calculated for each eye, such as for our patient (Figure 1A).[10] The vOCR is useful for the evaluation of the torsional VOR with lateral head tilt (i.e., OCR), and a diminished vOCR response can indicate the loss of otolith-ocular function.[2,3] The pattern of torsional eye deviation can also help differentiate peripheral ocular motor disorders from central lesions affecting the vestibulo-ocular pathways (Figure 1B). While torsional deviation in OTR is conjugate (incyclotorsion in the higher eye and excyclotorsion in the lower eye), torsional deviation in the superior oblique palsy is disconjugate (excyclotorsion only in the higher eye). Therefore, the relative direction of the torsion in the elevated eye—intorsion with OTR and extorsion with a superior oblique palsy—can be helpful in making the diagnosis.[11] Such torsional deviation can be examined at the bedside by using the double Maddox rod test, in which 2 different color cylinders (e.g., red and white) are held in front of each eye while the patient is looking at a light source. The cylinder converts the light source to a line, and therefore, when held vertically, the line seen with that eye is used to evaluate torsional disparity between both eyes.

In general, lesions causing a pathologic OTR are classically localized to the posterior fossa as depicted earlier, with differential including stroke, demyelination, trauma, etc.[6,7] It is also important to note that in the evaluation of patients with the acute vestibular syndrome,[12] a pathologic OTR can be present in central lesions and acutely in peripheral vestibulopathies.[4,9]

While in the past there has been minimal research focused on the diagnostic/localization value of ocular torsion, there are several studies showing that skew deviation can help differentiate central and peripheral lesions.[5] When there is vertical misalignment of the eyes causing diplopia, the diagnostic

challenge is to differentiate between a fourth nerve palsy and a skew deviation. In skew deviation, the degree of misalignment is relatively independent of where the eyes are pointed, that is, the deviation is conjugate. By contrast, the hallmark of a superior oblique palsy is that the degree of misalignment changes with the direction of gaze, that is, the deviation is nonconjugate. The affected eye is higher, and the vertical misalignment is greatest with the affected eye adducted and depressed, or when the head is tilted toward the side of the higher eye (toward the side of the lesion). This is the basis for the 3-step Parks-Bielschowsky test used to identify a superior oblique palsy.[13] In addition, the relative direction of the torsional deviation (using methods described earlier) is helpful in distinguishing between peripheral ocular nerve palsy and central pathologies—intorsion in the elevated eye with skew (and extorsion of the hypotropic eye) but extorsion in the elevated eye with a superior oblique or trochlear nerve palsy.[13]

In our patient, the presence of UBN and saccadic dysmetria along with OTR are compatible with an ispiversive torsional eye deviation from a lesion in the left pontomedullary junction (Figure 2C). Saccadic dysmetria can be caused by lesions extending to the pontine reticular formation or cerebellar pathways.[14] UBN can occur with the involvement of the brainstem and cerebellar network responsible for gaze holding, and in such cases, specific waveforms of the nystagmus can offer insight into specific localization.[15] For instance, UBN with a constant velocity may localize to the pons or caudal medulla, or UBN with a velocity-increasing slow phase may localize to the paramedian tract within the rostral medulla.[15]

In conclusion, in this study, we describe a patient with a conjugate, leftward torsional deviation of the eyes (i.e., right eye incyclotorsion and left eye excyclotorsion) consistent with OTR (Figure 2B). Because OTR can be caused by caudal and rostral brainstem lesions, this finding is not enough by itself to accurately localize lesions; for example, a leftward OTR could be from a left pontine or a right midbrain lesion. Therefore, other ocular motor or neurologic findings are imperative to assist with the localization in conjunction with OTR.

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