Approaching Acute Vertigo With Diplopia: A Rare Skew Deviation in Vestibular Neuritis

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Abstract

Evaluating the patient with acute constant vertigo or diplopia can be a daunting task for clinicians, who recognize that such symptoms can be the manifestation of potentially devastating disorders like stroke but may be uncomfortable eliciting and interpreting the key symptoms and subtle signs that distinguish dangerous from benign causes. We present a novel and highly instructive case of a patient with acute vertigo and binocular diplopia due to a large skew deviation due to vestibular neuritis. As the case unfolds, text and video commentary guide the clinician through the important elements of the history, bedside examination, and laboratory evaluation necessary for accurate diagnosis in the acute vestibular syndrome. We demonstrate how to interpret nystagmus and properly perform the head impulse test and test of skew deviation and discuss the pitfalls of overreliance on imaging when evaluating patients with acute vertigo.

CASE WITH COMMENTARY

Initial Symptoms

A 36-year-old woman with hyperlipidemia and occasional migraines without aura but no prior dizziness, vertigo, unsteadiness, or strabismus experienced her typical migraine with nausea and osmophobia. Hours into the episode, she developed progressive dizziness and unsteadiness without directional bias, requiring assistance walking. Although the headache improved with an over-the-counter analgesic, constant vertigo and vomiting soon followed.

Onset of vertigo during a migraine suggests a first episode of vestibular migraine as a possible etiology. Vestibular migraine is the most common cause of recurrent spontaneous (nonpositional) episodes of vertigo, and vestibular symptoms usually develop years after other migraine symptoms. Most episodes resolve within 72 hours.

Diplopia

The patient noted binocular vertical diplopia but no hearing loss, neck pain, dysarthria, dysphagia, facial weakness, limb incoordination, or sensory symptoms.

Possible lesion localization in patients with vertigo involves the vestibular periphery, brain stem, and cerebellum. Diplopia, dysphagia, dysarthria, or dysmetria (the “dangerous Ds”) point toward central localization, while hearing loss can have a central or peripheral cause. Patients with acute vertigo at the height of their symptoms are very distressed with nausea and vomiting and often ignore these associated symptoms. In this case, binocular vertical diplopia with an acute vestibular syndrome (AVS) is a red flag against peripheral and for central localization that requires further investigation.

Imaging

The patient was admitted to a local hospital after head computed tomography (CT) and head/neck CT angiography yielded normal findings in the emergency department. Head magnetic resonance imaging (MRI) with diffusion-weighted imaging (DWI), performed less than 24 hours from vertigo onset, also identified no abnormalities and no evidence of acute ischemia, demyelination, or vestibular nerve abnormality.

Magnetic resonance imaging with DWI and apparent diffusion coefficient is the criterion standard that defines an acute stroke. However, MRI performed too early in posterior fossa ischemia occasionally fails to detect restricted diffusion. This issue is particularly important in patients with vertigo, in whom overreliance on these early studies may cause the examiner to ignore signs of central
localization. Compelling central clinical signs (including the triad of ocular motor signs that will be described subsequently) require follow-up MRI, ideally obtained more than 48 hours after symptom onset, and careful clinical monitoring.

Nystagmus
Over 4 days, the patient was managed symptomatically with intravenous hydration and antiemetics. Vertigo and nausea resolved, although she could still induce brief dizziness with quick head turns. Gait and balance were improving. However, she was still squinting because of diplopia. She was discharged on day 5 with a treatment regimen of prednisone, 60 mg daily.

When evaluated in the otoneurology clinic the following day, results of a general neurologic examination were normal except for a moderately impaired, wide-based gait with tandem gait amination were normal except for a moderately.

Vestibulo-ocular Reflex Testing
A head impulse test (HIT) revealed catch-up saccades for the left horizontal and occasionally left anterior semicircular canals (SCCs) (Supplemental Video 2, available online at http://www.mcpiqojournal.org).

The HIT is necessary to identify the precise lesion site. The examiner rotates the patient’s head quickly by a small amount toward the side to be tested, observing whether the patient can effectively use the VOR to maintain visual fixation on a target or whether a “catch-up” saccade is required to re-fixate the target once the head stops. A normal HIT result in a patient with AVS verifies the integrity of the 3-neuron VOR arc and thus suggests central localization, as is seen with posterior inferior cerebellar artery strokes. In contrast, lesions of the vestibular nerve or labyrinth impair the VOR and produce an abnormal HIT result. In this case, unidirectional horizontal nystagmus beating away from the side with an abnormal HIT result points to a peripheral vestibulopathy, most commonly vestibular neuritis (VN). However, the possibility of an anterior inferior cerebellar artery (AICA) stroke remains. The internal auditory artery, a branch of the AICA, supplies the inner ear. Accordingly, AICA infarcts may involve the labyrinth and present like an acute peripheral vestibulopathy, typically accompanied by hearing loss from cochlear infarction. Thus, the bedside examination should include assessment for acute hearing loss. So far in this case, with peripheral-appearing nystagmus and HIT result and normal hearing, the next step is to test for skew deviation.

Skew Deviation
The patient had a prominent leftward head tilt and right hypertropia, preferentially fixating with the left eye, as demonstrated with cover testing (Supplemental Video 3, available online at http://www.mcpiqojournal.org). Maddox rod testing revealed the hypertropia to be comitant,
and results of a Bielschowsky 3-step test did not suggest a trochlear nerve palsy.

In normal individuals, tilting the head laterally activates the utricle driving the otolith-ocular reflex, a mechanism to maintain a gravitationally upright visual axis by generating a compensatory ocular counterroll in the opposite direction. The resulting torsional eye movements are conjugate, whereas the vertical movements are slightly disconjugate.

Under pathologic conditions involving the peripheral labyrinth or central utricular-ocular pathways, the resulting internal misperception of verticality may produce a partial or full ocular tilt reaction (OTR) in an attempt to realign the patient with the erroneously computed vertical. The OTR is a triad of head tilt, ocular torsion in the same direction as the head tilt, and skew deviation with the lower eye on the side of the head tilt (Figure 1). Skew is a prenuclear vertical ocular misalignment that is usually comitant (the degree of misalignment does not change in different gaze positions as it does with an extracocular muscle weakness from an acute cranial nerve palsy) and represents an "otolithic pathway" cause of diplopia. In a patient with an AVS, skew is the most likely cause of vertical diplopia.

The magnitude of skew deviation and head tilt are important. Small-amplitude skew detected by cover testing may rarely occur in VN, but a large skew deviation in a patient with AVS, such as the one seen in this patient, is much more frequent in acute stroke and represents a red flag requiring further investigation.

**Clinical Diagnosis**

Because of the large persisting skew deviation, a second head MRI was performed on day 8. The results were also normal, and acute left VN was diagnosed.

**Vestibular Testing**

Within 2 weeks of symptom onset, diplopia, dizziness, and unsteadiness completely resolved. Quick head movements would sometimes cause the patient’s vision to take a moment to catch up. Subsequent vestibular evaluation confirmed a left peripheral vestibulopathy. Warm caloric reflex testing produced 61°/s right-beating nystagmus when irrigating the right ear but no response from the left ear. Video HIT of the horizontal SCC revealed a gain of 0.87 on the right but 0.51 on the left, with overt and covert catch-up saccades (Figure 2). Gain was also reduced for the left anterior but not the posterior canal. Cervical vestibular evoked myogenic potentials (VEMPs) were normal and symmetric. Ocular VEMPs of 97 dB nHL were present bilaterally at 500 Hz, but the P1 to N1 amplitude on the left was less than half that on the right. Audiometry yielded normal results.
From the labyrinth, the superior vestibular nerve (SVN), the most common target of VN, carries afferent fibers from the horizontal and anterior SCCs, the utricle and small portion of the saccule. The inferior vestibular nerve carries afferents from the posterior SCC and the saccule. In this case, vestibular testing with calorics and video HIT confirmed impairment of the SVN afferents, with decreased function of the horizontal and anterior canals. The inferior vestibular nerve carries afferents from the posterior SCC and the saccule. In this case, vestibular testing with calorics and video HIT confirmed impairment of the SVN afferents, with decreased function of the horizontal and anterior canals. In contrast, the posterior canal function via the inferior vestibular nerve was spared. The role of video-oculography recording of the HIT cannot be overemphasized, as it provides both multicanal VOR assessment and quantification of VOR gain and catch-up saccades.

Results of the ocular VEMP, a test of utricular function, were abnormal on the left, while results of the cervical VEMP, a test of saccular function, were normal. Such utricular pathway dysfunction could both explain the patient’s OTR and support localization to the SVN. Normal audiographic results are expected in VN and argue against Ménière disease or bacterial labyrinthitis. Most strokes do not cause hearing loss, although AICA strokes may cause cochlear infarction.

**Outcome**

Examination 2 months after onset revealed steady visual fixation, although weak right-beating nystagmus was elicited by removing fixation with video-oculography and could be intensified following horizontal headshaking and with mastoid vibration. The HIT continued to show catch-up saccades for the left horizontal canal. There was a slight leftward head tilt. Alternate cover testing revealed no vertical ocular misalignment, and prism testing identified less than 1 diopter of right hyperdeviation. Gait and station...
were normal. Video HIT showed improved left horizontal canal gain to 0.64, but left warm caloric response was still absent.

Normal MRIs, vestibular evaluation, and the clinical course confirmed the diagnosis of VN despite the red flag of a pronounced head tilt and large skew deviation. Through vestibular compensation, the patient’s symptoms resolved except with quick head movements that challenge the high-frequency VOR. When examined long after acute vertigo has resolved, provocative maneuvers such as headshaking and vibration, with visual fixation blocked, may bring out diagnostically important nystagmus effectively suppressed with visual fixation.

**DISCUSSION**

This case illustrates the frequent diagnostic challenges in patients with vertigo. The best approach starts with characterizing the vestibular syndrome by considering the timing and triggers and performing a targeted examination. Although benign paroxysmal positional vertigo (BPPV) is the most common vestibular disorder, it causes brief (<1 minute) episodes of vertigo and nystagmus triggered by position changes or positional testing. In this case, spontaneous nystagmus in the upright position with constant symptoms makes BPPV highly unlikely, although the horizontal canal BPPV variant can occasionally cause “pseudospontaneous” nystagmus that should be suspected if the nystagmus reverses direction when the head is pitched forward 60 degrees. The first attack of an episodic disorder such as vestibular migraine was initially possible (and could produce nystagmus that appears central or peripheral) but became unlikely because vestibular symptoms and signs persisted for days.

In patients with AVS who lack obvious central nervous system features, a test for 3 ocular motor signs—head impulse, nystagmus, and test of skew (HINTS)—is highly sensitive for distinguishing central causes such as stroke from peripheral causes such as VN.  A normal HIT result or the presence of spontaneous nystagmus that is direction-changing, vertical, or purely torsional are unequivocal central signs. Skew deviation on cover testing is also very suggestive of a central lesion (Table). A large skew such as that detected in this case is rare in

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**TABLE. HINTS to INFARCT**

| INFARCT (or central if any of these symptoms are present) | INFARCT (or central if any of these symptoms are present) |
|---|---|
| Head Impulse | Head Impulse |
| Nystagmus characteristics | Nystagmus characteristics |
| Head impulse negative (no catch-up saccades) | Acute hearing loss is a red flag for possible anterior inferior cerebellar artery stroke even when other signs point toward a peripheral cause |
| Fast-phase alternating (direction-changing or purely vertical or torsional) | Severe truncal and gait ataxia (inability to sit or stand without holding on) is a red flag for a central cause |
| Relocation on Cover Testing (demonstrating vertical misalignment) | Other findings |
| Test of Skew | Bedside hearing test |
| Positive for potential central cause | Acute hearing loss is a red flag for possible anterior inferior cerebellar artery stroke even when other signs point toward a peripheral cause |
| Degree of Imbalance | Degree of Imbalance |
| No skew deviation (no vertical deviation) on alternate cover testing | no skew deviation (no vertical deviation) on alternate cover testing |
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SKEW IN VESTIBULAR NEURITIS

Patients with acute constant vertigo and diplopia present a diagnostic challenge that requires a careful bedside examination to distinguish benign from potentially life-threatening causes. Imaging with CT has little value, and even MRI with DWI may miss small strokes affecting the otolith-ocular pathways. Ongoing research aims to improve diagnostic accuracy by nonexpert clinicians through both medical education and use of new technologies.

CONCLUSION

Patients with acute constant vertigo and diplopia present a diagnostic challenge that requires a careful bedside examination to distinguish benign from potentially life-threatening causes. Imaging with CT has little value, and even MRI with DWI may miss small strokes if performed too soon. When overt central neurologic signs are absent, the clinician must perform and correctly interpret results of the HIT, characterize the nystagmus, and test for skew deviation as well as assess for acute hearing loss (the HINTS Plus examination). Although this examination battery has very high sensitivity and specificity in AVS in expert hands, this case illustrates how on rare occasions a large skew deviation can result from a peripheral vestibulopathy affecting the otolith-ocular pathways. Ongoing research aims to improve diagnostic accuracy by nonexpert clinicians through both medical education and use of new technologies.

SUPPLEMENTAL ONLINE MATERIAL

Supplemental material can be found online at http://www.mcpiqojournal.org. Supplemental material attached to journal articles has not been edited, and the authors take responsibility for the accuracy of all data.

Abbreviations and Acronyms: AICA = anterior inferior cerebellar artery; AVS = acute vestibular syndrome; BPPV = benign paroxysmal positional vertigo; CT = computed tomography; DWI = diffusion-weighted imaging; HINTS = head impulse, nystagmus, and test of skew; HIT = head impulse test; MRI = magnetic resonance imaging; OTR = ocular tilt reaction; SCC = semicircular canal; SVN = superior vestibular nerve; VEMP = vestibular evoked myogenic potential; VN = vestibular neuritis; VOR = vestibulo-ocular reflex

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REFERENCES

1. Hotson JR, Baloh RW. Acute vestibular syndrome. N Engl J Med. 1998;339(10):680-685.
2. Brodsky MC, Donahue SP, Vaphiades M, Brandt T. Skew deviation revisited. Surv Ophthalmol. 2006;51(2):105-128.
3. Brandt T, Dieterich M. Pathological eye-head coordination in roll: tonic ocular tilt reaction in mesencephalic and medullary lesions. Brain. 1987;110(pt 3):649-666.
4. Westheimer G, Blair SM. The ocular tilt reaction—a brainstem oculomotor routine. Invest Ophthalmol. 1975;14(1):833-839.
5. Shin BS, Oh SY, Kim JS, et al. Cervical and ocular vestibular-evoked myogenic potentials in acute vestibular neuritis. Clin Neuropathol. 2012;31(2):369-375.
6. Edelow JA, Gurley KL, Newman-Toker DE. A new diagnostic approach to the adult patient with acute dizziness. J Emerg Med. 2018;54(4):469-483.
7. Kattah JC, Talkad AV, Wang DZ, Hsieh YH, Newman-Toker DE. HINTS to diagnose stroke in the acute vestibular syndrome: three-step bedside oculomotor examination more sensitive than early MRI diffusion-weighted imaging. Stroke. 2009;40(11):3504-3510.

8. Newman-Toker DE, Kerber KA, Hsieh YH, et al. HINTS outperforms ABCD2 to screen for stroke in acute continuous vertigo and dizziness. Acad Emerg Med. 2013;20(10):986-996.

9. Safran AB, Vibert D, Issoua D, Häusler R. Skew deviation after vestibular neuritis. Am J Ophthalmol. 1994;118(2):238-245.

10. Vibert D, Häusler R, Safran AB, Koerner F. Diplopia from skew deviation in unilateral peripheral vestibular lesions. Acta Otolaryngol. 1996;116(2):170-176.

11. Lee H, Kim JS, Chung EJ, et al. Infarction in the territory of anterior inferior cerebellar artery: spectrum of audiovestibular loss. Stroke. 2009;40(12):3745-3751.

12. Saber Tehrani AS, Kattah JC, Mantokoudis G, et al. Small strokes causing severe vertigo: frequency of false-negative MRIs and non-lacunar mechanisms. Neurology. 2014;83(2):169-173.

13. Stanton VA, Hsieh YH, Camargo CA Jr, et al. Overreliance on symptom quality in diagnosing dizziness: results of a multicenter survey of emergency physicians. Mayo Clin Proc. 2007;82(11):1319-1328.

14. Royl G, Pioner C, Leithner C. Dizziness in the emergency room: diagnoses and misdiagnoses. Eur Neurol. 2011;66(5):256-263.

15. Saber Tehrani AS, Kattah JC, Kerber KA, et al. Diagnosing stroke in acute dizziness and vertigo: pitfalls and pearls. Stroke. 2018;49(3):788-795.

16. Acute Video-oculography for Vertigo in Emergency Rooms for Rapid Triage (AVERT). ClinicalTrials.gov website. https://clinicaltrials.gov/ct2/show/NCT02483429. Posted June 29, 2015. Updated December 18, 2019. Accessed February 4, 2020.