Pearls & Oy-sters: Vestibular neuritis or not?

The significance of head tilt in a patient with rotatory vertigo

Carsten Finke, MD
Christoph J. Ploner, MD

Address correspondence and reprint requests to Dr. Carsten Finke, Department of Neurology, Charité-Universitätsmedizin Berlin, Campus Charité Mitte, Charitéplatz 1, D-10117 Berlin, Germany
carsten.finke@charite.de

CLINICAL PEARL The ocular tilt reaction (OTR) is a central vestibular disorder that is characterized by the triad of head tilt, skew deviation, and ocular torsion.

CLINICAL OY-STER As the OTR frequently shows an incomplete manifestation, a head tilt can be the only apparent clinical sign of significant brainstem disorders. Hence, even a slight head tilt should be thoroughly investigated.

CASE REPORT A 48-year-old man presented after sudden onset of a severe, persistent rotatory vertigo associated with nausea, vomiting, and postural imbalance. Examination revealed a horizontal-torsional spontaneous nystagmus with the quick phases beating to the right ear. The nystagmus was attenuated by fixation as examined by Frenzel goggles. Head impulse test was positive to the left. When attempting to walk and during Romberg testing he fell to his left side. Except for a head tilt to the right (see video on the *Neurology*® Web site at www.neurology.org), no other neurologic deficit was observed. The patient's medical history revealed no neurologic disorders. Quantitative caloric testing showed a canal paresis of the left vestibular organ. Taken together, the initial clinical presentation was highly suggestive of a left vestibular neuritis.

However, due to the head tilt, an extended neuroophthalmologic examination was performed that revealed a discrete but complete OTR to the right with head tilt, mild skew deviation (2° left hypertropia), ocular torsion (right eye excyclotropia 4°, left eye no cyclotropia), and a tilt of the subjective visual vertical to the right (right eye 7° to the right, left eye 4° to the right).

In contrast to the sparse clinical signs, cerebrospinal MRI showed multiple T2-hyperintense lesions of the right dorsolateral medulla oblongata, left mesencephalon, right medial cerebellar peduncle, corpus callosum, pericallosal white matter, and three lesions in the spinal cord. Reconstruction of the lesion of the dorsolateral medulla oblongata according to the atlas of Paxinos and Huang[1] revealed damage to the right spinal, medial, superior, and lateral vestibular nuclei. Reconstruction of the mesencephalic lesion showed involvement of the left interstitial nucleus of Cajal (INC) (figure). No further brainstem lesions were observed. No enhancement of the eighth cranial nerve, as occasionally described for vestibular neuritis,[2] was seen. CSF analysis revealed no pleocytosis and normal protein, glucose, and lactate, but oligoclonal bands in CSF only. Visual evoked potentials (VEP) showed a delayed P100 on the left.

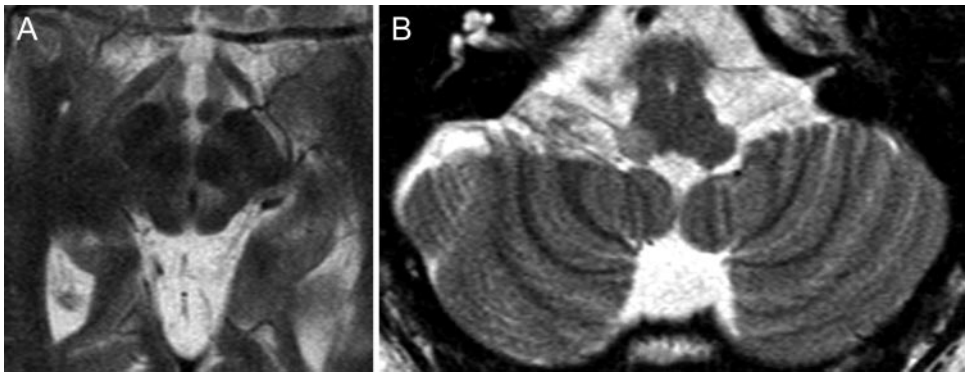
The cerebrospinal lesion pattern, the inflammatory CSF changes, and the abnormal VEP were suggestive of a demyelinating process. According to the revised McDonald Criteria, a clinically isolated syndrome (CIS) was diagnosed and treatment with IV steroids was initiated. On follow-up 4 weeks later, the patient had recovered from vertigo and nausea and had no nystagmus on examination. However, the head tilt to the right persisted. MRI showed no new lesions. Seven months later, the head tilt was still detectable, but neuroophthalmologic examination showed only a slight tilt of the subjective visual vertical to the right (both eyes 2°) and no skew deviation or ocular torsion. Cerebrospinal MRI showed no new lesions.

The clinical presentation comprised a left peripheral vestibular syndrome suggestive of vestibular neuritis and a right central vestibular syndrome manifesting as OTR. The latter is characterized by the triad of head tilt, skew deviation, and ocular torsion.[3] Additionally, a tilt of subjective visual vertical (SVV) is observed. The head is tilted toward the lower eye. Ocular torsion manifests as incyclotropia of the upper eye and excyclotropia of the lower eye, i.e., the upper poles of the eyes rotate toward the lower ear. Pathogenetic substrate is a vestibular tone imbalance caused by unilateral lesions of the graviceptive vestibular pathways that run from the otoliths and the vertical semicircular canals to the ocular motor nuclei and the rostral integration centers for

Supplemental data at
www.neurology.org

From the Department of Neurology, Charité-Universitätsmedizin Berlin, Germany.
Disclosure: The authors report no disclosures.

Figure Cerebral MRI revealing (A) a left mesencephalic lesion affecting the interstitial nucleus of Cajal and (B) T2-hyperintense lesions of the right dorsolateral medulla oblongata affecting the spinal, medial, superior, and lateral vestibular nuclei



vertical and torsional eye movements (INC and riMLF). They also provide input to vestibular thalamic nuclei and cortical areas involved in perception of verticality.[4] Altogether, lesions of these projections lead to the clinical syndrome involving ocular motor (ocular torsion, skew deviation), perceptual (SVV tilt), and postural (head tilt) dysfunction. As the graviceptive vestibular pathways cross at a lower pontine level, direction of the ocular tilt reaction holds a clinically relevant topographic value: with peripheral and pontomedullary lesions below the crossing all tilt effects (ocular motor, perceptual, and postural) are ipsiversive, i.e., the ipsilateral eye is undermost. With pontomesencephalic lesions all tilt effects are contraversive, i.e., the contralateral eye is undermost. Hence, the OTR to the right in our patient cannot be explained by a left-sided peripheral vestibular syndrome but must rather be attributed to the lesion of the vestibular nuclei in the right dorsolateral medulla or to the lesion of the INC in the left mesencephalon. As pontomedullary lesions typically cause a disconjugated ocular torsion and pontomesencephalic lesions, a conjugated ocular torsion,[5] damage to the vestibular nuclei appears to be the cause of OTR in our patient.

The two vestibular syndromes that differ in localization (peripheral vs central) and side (right vs left) could point to separate pathogenetic mechanisms, i.e., an inflammatory brainstem lesion and an incidental left peripheral vestibular neuritis. Alternatively, the peripheral vestibular syndrome may have resulted from a MRI-negative plaque at the root entry zone of the left eighth nerve causing a vestibular pseudoneuritis.[6] Such a strategically localized lesion causes a vestibular syndrome mimicking vestibular neuritis in clinical presentation and calorimetric testing. However, there is no means to reliably differentiate between these two possibilities.

Recently, the reliability of clinical examination in the differentiation of vestibular neuritis from vestibular pseudoneuritis was investigated.[7] It was shown that single clinical signs provide only limited sensitivity and specificity, except for skew deviation that indicated a vestibular pseudoneuritis with a high specificity. In our case, the key for the diagnosis of a central vestibular syndrome in addition to the obvious peripheral vestibular syndrome lay in recognizing the head tilt to the right side as an incompatible component of a left peripheral vestibular syndrome. Recognizing this incompatibility and consequently assuming an additional vestibular disease called for further investigations that finally led to the correct diagnosis.

ACKNOWLEDGMENT

The authors thank Dr. Juri Katchanov for recording the patient video.

REFERENCES

1. Atlas of the Human Brainstem. San Diego: Academic Press; 1995.
2. Brodsky MC, Donahue SP, Vaphiades M, Brandt T. Skew deviation revisited. Surv Ophthalmol 2006;51:105–128.
3. Rabinovitch HE, Sharpe JA, Sylvester TO. The ocular tilt reaction: a paroxysmal dyskinesia associated with elliptical nystagmus. Arch Ophthalmol 1977;95:1395–1398.
4. Brandt T, Dieterich M. Vestibular syndromes in the roll plane: topographic diagnosis from brainstem to cortex. Ann Neurol 1994;36:337–347.
5. Brandt T, Dieterich M. Skew deviation with ocular torsion: a vestibular brainstem sign of topographic diagnostic value. Ann Neurol 1993;33:528–534.
6. Thomke F, Hopf HC. Pontine lesions mimicking acute peripheral vestibulopathy. J Neurol Neurosurg Psychiatry 1999;66:340–349.
7. Cnyrim CD, Newman-Toker D, Karch C, Brandt T, Strupp M. Bedside differentiation of vestibular neuritis from central "vestibular pseudoneuritis." J Neurol Neurosurg Psychiatry 2008;79:458–460.

Pearls & Oy-sters: Vestibular neuritis or not?: The significance of head tilt in a patient with rotatory vertigo

Carsten Finke and Christoph J. Ploner

Neurology 2009;72;e101-e102
DOI 10.1212/WNL.0b013e3181a60a44