Persistent Direction-changing Apogeotropic Horizontal Positional Vertigo is Not Always So Benign

Dear Editor,

A 65-year-old female presented in mid-October 2023 with a 2-week history of spontaneous vertigo, which got worse on lying down and aggravated on getting up from supine position. Oculomotor examination revealed a horizontal, left-beating spontaneous nystagmus with removal of fixation (by video-oculography infrared goggles). The horizontal head impulse test elicited a catch-up saccade on heaving the head to right. Tandem Romberg's test and tandem walking were mildly impaired, but there was no upper and lower limb dysmetria. The otoneurologic examination revealed normal vertical and horizontal saccadic and smooth pursuit eye movements.

The positional test results were as follows:

1. **Bow and lean test**: elicits left-beating nystagmus on the lean position and in the bow position, it disappears.
2. **Supine roll test**: elicits apogeotropic horizontal positional nystagmus on lateral head roll to the right and left. On the left lateral head roll direction reversal was conspicuously very late to occur. The sequence of oculomotor findings and positional tests carried out by the authors is accessible.[1]

Atypical clinical findings (spontaneous left-beating horizontal nystagmus, positive head impulse test, mild gait ataxia, disappearance of spontaneous left-beating nystagmus on the bow test, and a very late direction reversal of horizontal positional nystagmus on left lateral head roll) instigated us to order a contrast magnetic resonance imaging (MRI) of brain with dedicated cuts of the posterior fossa. MRI brain with contrast showed an enhancing lesion in the lateral wall of the fourth ventricle. The neuroimaging differentials considered were tuberculoma, sarcoidosis, toxoplasma, and cysticercosis [Figure 1]. A solitary infratentorial lesion hypointense on T1W, isointense on T2W, hyperintense on fluid attenuated inversion recovery (FLAIR) (representing perilesional edema) with nodular enhancement is most likely to be tuberculoma, and other possibilities are less likely. A definitive diagnosis could have been reached by biopsy. However, the patient refused to be investigated further and was lost to follow-up.

A nystagmus generated by head motion *per se* or a newly acquired head position in space relative to the gravity outlasting the head motion is termed positional nystagmus.[2] It has been classified according to its distinguishing attributes as follows:

1. Peripheral versus central depending on the site of the lesion
2. Paroxysmal versus persistent depending on temporal dispersion in time
3. A combination of temporal profile and directionality as follows:

Type 1: Persistent, direction changing;

Type 2: Persistent, direction fixed; and

Type 3: Transient.

It is customary to elaborate the nystagmus with characteristics like latency, duration, and fatigability. The diagnosis of a central positional syndrome is dependent on observing deviations in the diagnostic criteria of benign paroxysmal positional vertigo (BPPV) in any patient complaining of vertigo triggered by the change in position of head relative to the gravity.[3] Vestibulologists classify the persistent form of central positional syndrome as central positional nystagmus (CPN) and the transient form as central paroxysmal positional nystagmus (CPPN). Despite evolving colossally, the human vestibular system is often caught up in the dilemma of distinguishing between a tilt (angular acceleration) and translation (linear acceleration) that routinely occurs in many daily physical activities. In the diagnostic lateral head roll from the supine neutral position, the vestibular system senses angular acceleration (through horizontal semicircular canals) and linear acceleration (through utricle and saccule) and carries out the estimation of gravity direction by optimizing the ambiguities of inputs received from the peripheral vestibular system. This necessitates gravito-inertial acceleration (GIA), a vector summation of gravity and linear acceleration. GIA aligns the eye velocity generated by the angular vestibulo-ocular reflex (aVOR) and modulates the spatial orientation of aVOR. The velocity storage properties and somatogravic feedback loop of the vestibulocerebellum carry out aVOR modulation.[4]

Lesions in the nodulus–uvula of vestibulocerebellum result in apogeotropic CPN because of malfunctioning velocity-storage mechanism (VSM). During co-occurring tilt and translation, the otolith organs relay identical GIA signals, and a "tilt-estimator

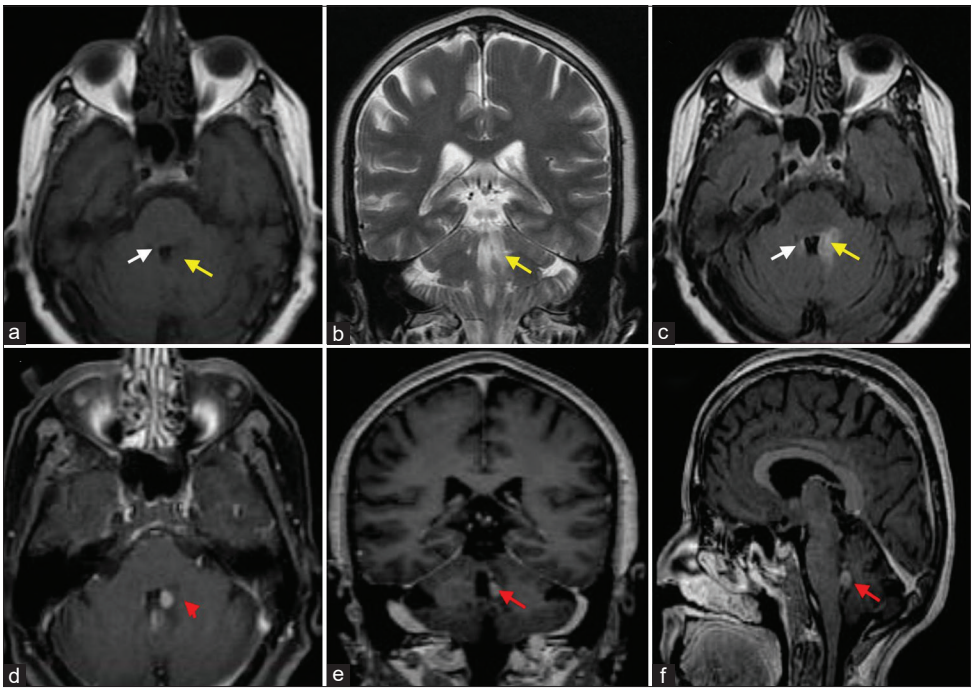

Figure 1: (a) Axial T1W shows a hypointense mass lesion in the left lateral wall of the fourth ventricle (yellow arrow) and it appears isointense on T2W sequence (b; yellow arrow) and hyperintense on FLAIR sequence (c; yellow arrow) adjacent to fourth ventricle (white arrow). T1W postcontrast (d; axial, e; coronal, f; sagittal) shows nodular enhancement of the lesion (red arrow)

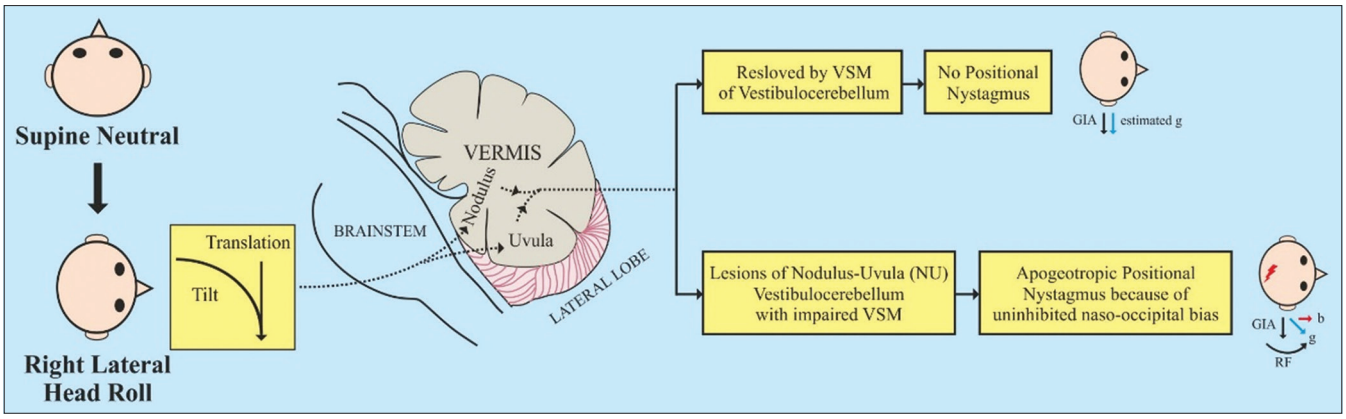

Figure 2: Tilt-translation ambiguity during right lateral head roll in healthy individuals versus those with Nodulus-Uvula (NU)-vestibulocerebellar lesion. b = bias, GIA = gravitoinertial acceleration, *g* = estimated gravity, RF = rotation feedback, VSM = velocity-storage mechanism

circuit" resolves this inherent ambiguity by combining the information from the semicircular canals about head rotation with otolith information about linear acceleration through VSM. Rotating normal subjects at a constant speed around an axis that is tilted away from the true vertical (off-vertical axis rotation) results in a sustained horizontal nystagmus. A malfunctioning tilt-estimator circuit resulting from lesions in the vestibulocerebellum makes estimates of the direction of gravity that are erroneously biased away from true vertical. If the bias is toward the nose, when the head is turned to the side while supine, there will be sustained horizontal positional nystagmus (apogeotropic type of CPN) because of an inappropriate feedback signal indicating that the head is rotating when it is not [Figure 2].[5,6]

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Conflicts of interest

There are no conflicts of interest.

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