When Paroxysmal Positioning Vertigo Isn’t Benign

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Abstract

The electronystagmographic auditory brainstem response and magnetic resonance imaging findings for a 33-year-old male with a 3.5 cm left vestibular schwannoma are presented. Of particular interest was the presence of an unusual positioning nystagmus following the Dix-Hallpike maneuver in the right head-hanging position. The patient demonstrated a nystagmus that was immediate in onset and not fatigueable upon repeated positioning. During positioning, the patient experienced a vertical bobbing sensation and dysphoria, but not rotational vertigo. Most importantly, the nystagmus had a predominant downbeating vertical component. The case illustrates the diagnostic significance of downbeating nystagmus elicited by the Dix-Hallpike maneuver.

Key Words: Benign paroxysmal positional vertigo (BPPV), electronystagmography, positional nystagmus

The clinical diagnosis of benign paroxysmal positioning vertigo (BPPV) is defined by signs elicited by the Dix-Hallpike maneuver that include nystagmus that is latent in onset (by several seconds) and fatigueable upon several repetitions of the maneuver and is accompanied by vertigo and nausea (but not vomiting) (Dix and Hallpike, 1952; Cawthorne et al, 1956; Hallpike, 1962). Additionally, this nystagmus, termed benign paroxysmal positioning nystagmus (BPPN) (Dix and Hallpike, 1952; Cawthorne et al, 1956), is usually purely rotatory, or horizontal-rotatory and reverses in direction when the patient is brought from a recumbent to a sitting position. The horizontal component beats toward the undermost ear. The vertical component is always upbeating in BPPN. The presence of BPPV usually denotes a self-limiting process that may require weeks or months to resolve. In a small number of cases, however, BPPV has been reported in association with acoustic neuroma, vertebrobasilar insufficiency, Meniere’s disease, luetic labyrinthitis, multiple sclerosis, polyarteritis nodosa, and systemic lupus (Watson et al, 1981; Watson and Terbrugge, 1982; Baloh et al, 1987). For this reason, it may be questioned whether BPPV is in all circumstances a sign of “benign,” self-limiting disease.

Nystagmus and/or vertigo elicited by the Dix-Hallpike maneuver have been described that do not fulfill the criteria for BPPV. The nystagmus is usually not latent in onset, nor is it usually accompanied by vertigo. Additionally, the nystagmus may occur in more than one head-hanging position and last for more than 1 minute. The nystagmus may not be fatigueable on repeated positioning. This nystagmus has been associated with central nervous system disease (Barber and Stockwell, 1980) and has been observed in patients with brainstem tumors (e.g., vestibular schwannoma [VS]), Arnold-Chiari malformation, lateral medullary infarction, multiple sclerosis, cerebellar tumor, and brainstem contusion (Watson et al, 1981).

Also, nystagmus elicited by the Dix-Hallpike maneuver may show many of the characteristics described above (no latent period, nonfatigueable) but, most notably, a downbeating vertical component.
component (Watson et al, 1981). These findings have been associated with disease in the posterior fossa affecting the cerebellar vermis and have been reported in patients with disease affecting the 4th ventricle (Watson et al, 1981). A case is presented herein of a patient with downbeating positional nystagmus following the Dix-Hallpike maneuver. This patient was found to have a large posterior fossa mass.

**CASE REPORT**

The patient was a 33-year-old male with a past history of episodic vertigo over a 4-year period. The attacks of vertigo were associated with nausea, vomiting, and dysequilibrium, but not aural symptoms. The patient also reported transient numbness in the distribution of the second division (V2) of the trigeminal nerve, episodic diplopia, and positioning vertigo, especially with his head hanging toward the right. Additionally, the patient complained of both bilateral tinnitus that was “ringing” in character and hearing loss on the left side. The patient first noticed the hearing loss when holding a telephone receiver over the affected ear.

His physical examination was remarkable for a decreased corneal reflex and a House-Brackmann grade II facial weakness on the left side. His gait was slow and wide based. Tandem walking was performed with great difficulty. A head-hanging right Hallpike’s maneuver elicited a downbeating nystagmus that persisted as long as the head was maintained in the offending position. The nystagmus was initiated without a latent period and was accompanied by a “bobbing” sensation and nausea and was not fatigable upon repeated positioning.

Audiometric testing revealed normal hearing for the right ear and mild, low- and high-frequency sensorineural hearing loss for the left ear. Word recognition ability (NU-6 lists) was excellent (96%) for the right ear and inordinately poor (66%) for the left ear in relation to the degree of hearing loss. Word recognition ability in the left ear worsened significantly when stimulus intensity was increased from 50 dB SL to 70 dB SL (66%-20%). Stapedial reflexes were absent with contralateral and ipsilateral stimulation presented to the left (involved) ear. Stapedial reflexes were obtained at normal intensity levels with the contralateral and ipsilateral stimuli presented to the right (uninvolved) ear.

As shown in Figure 1, auditory brainstem response (ABR) testing revealed normal wave I–wave V interwave intervals with stimuli presented to the right ear (AD). The wave V/wave I amplitude ratio was normal. Stimuli presented to the left ear (AS) elicited only wave I.

Electronystagmographic testing (ENG) revealed normal ocular motility and an absence of spontaneous nystagmus. Positioning testing (Dix-Hallpike maneuver) in the head-hanging right position with eyes opened and with ocular fixation elicited a down-beating nystagmus with no latent period that persisted for the duration of the critical head positioning and was not fatigable upon repeated positioning (see Fig. 2). Alternate binaural bithermal caloric testing revealed a 93 percent unilateral weakness on the left side. Rotational testing (0.01 – 0.64 Hz, 50 deg/sec peak velocity) revealed abnormal phase leads occurring from 0.01 to 0.04 Hz. Gain and symmetry values for frequencies 0.01 Hz to 0.64 Hz were normal.

Magnetic resonance imaging (MRI) with gadolinium contrast revealed a mass filling the internal auditory canal, extending 3.5 cm into the left cerebellopontine angle on the left side (see Fig. 3 A, B). Additionally, there was an inhomogeneous component to this tumor near the brain stem, which at surgery was confirmed.
to be an intratumor cyst. The MRI scan showed extension of the mass caudally to the foramen magnum and rostrally to the tentorium incisura with evidence of compression and distortion of the left side of the brain stem and deformation of the 4th ventricle with flattening and distortion of the left anterolateral aspect. The lateral ventricles were not dilated. The preoperative diagnosis was vestibular schwannoma (VS), although a cystic meningioma could not be excluded.

The patient was counseled regarding the relative risks and benefits of translabyrinthine and suboccipital approaches to the removal of the tumor and chose the translabyrinthine approach in an attempt to best preserve facial nerve function. The histopathologic diagnosis was schwannoma. Postoperatively, at 6 months, the patient had a House-Brackmann grade II facial weakness on the left side and was free of positioning vertigo and nystagmus.

**DISCUSSION**

Interpreting results of positional nystagmus (i.e., nystagmus/vertigo elicited by change in head and body position) and positioning nystagmus (nystagmus/vertigo elicited by rapid movement of the head and body) subtests of the ENG test battery have been problematic for many clinicians because the presence of nystagmus does not necessarily represent evidence of disease. When results of these subtests indicate the presence of vestibular system disease, they may offer little or no localizing or lateralizing information. For instance, Barber and Wright (1973) reported that positional nystagmus occurred in a large proportion of asymptomatic

**Figure 2** Horizontal and vertical eye movement tracings obtained during Hallpike maneuver. Patient was tested with eyes open and fixating upon a target at approximately center gaze. Positioning of the patient in the head-hanging left position elicits an oblique nystagmus with a prominent and nonfatigueable downbeating vertical component.

**Figure 3** Transverse (A) and coronal (B) views of the VS. Notice the shifting of left brainstem structures to the right of midline. Also, notice the narrowing of the 4th ventricle.
normal subjects. Therefore, the identification of meaningful clinical findings (i.e., findings that point to a specific site of lesion) on positional and positioning testing is valuable.

It has been hypothesized by Schuknecht (1969) that the origin of BPPV is "free-floating" otoliths that have been dislodged from the otolith membrane due to head trauma or age-related degenerative processes (i.e., cupulolithiasis). The particles produce "push or pull" forces on the posterior semicircular canal cupula (resulting in increases and decreases in neural activity) when a patient is placed in an ear-dependent down position and then brought back to a sitting position. Experimental evidence supporting this hypothesis was offered by Schuknecht and Ruby (1973), who demonstrated histologic evidence of large basophilic deposits attached to the posterior semicircular canal cupulae of three patients diagnosed with BPPV. BPPV is usually a self-limiting disease process, and "liberatory" maneuvers have been developed to hasten the process of migration of the otolith out of the semicircular canal system (Semont et al, 1988).

The patient described in this report first presented with a complaint of vertigo evoked by changes in position. The Dix-Hallpike maneuver elicited a nystagmus with predominant downbeating vertical component. It is felt that the downbeating nystagmus occurred as a result of compression of structures controlling vertical gaze at the level of the 4th ventricle. Compression occurred due to the tumor mass coupled with the force exerted by the mass on the cerebellar vermis and caudal brainstem structures due to the movement associated with the Dix-Hallpike maneuver (Kattah et al, 1984). This patient was found to have a large (3.5 cm) VS. There have been few reports of downbeating nystagmus elicited by the Dix-Hallpike maneuver. Kattah et al (1984) presented a case report of a patient with a hematoma involving the lower and posterior cerebellar vermis. The patient presented with truncal ataxia, saccade dysmetria, and paroxysmal positioning nystagmus with a predominant downbeating component. Drainage of the hematoma resulted in a resolution of these findings.

This case highlights clearly the importance of distinguishing peripheral from central positioning nystagmus. The nystagmus demonstrated by the patient in the present report might be described by some as representing "nonclassical" BPPN. However, because this label may be used to describe any nystagmus or behavior (e.g., paroxysmal nystagmus without accompanying vertigo or nausea or vice versa), it is our feeling that this description has little diagnostic value. It is suggested that the phenomenon described in this report is caused by disease affecting the brain stem at the pontomedullary junction and should be referred to as central positioning nystagmus (CPN) (Brandt, 1993); the nystagmus accompanied by a perception of movement may be referred to as central positioning vertigo (CPV) (Brandt, 1993). As shown in this case report, the vertigo and associated nystagmus of CPV differs from BPPV in that CPN (1) does not decrease in magnitude on repeated positioning, (2) does not have a clear latency, (3) is of longer duration than BPPV (lasting longer than 30 seconds), (4) is unaffected by ocular fixation, and (5) typically is purely vertical with the fast phase beating downward.

REFERENCES


