J Am Acad Audiol 17:481–486 (2006)

Simultaneous Acute Superior Nerve Neurolabyrinthitis and Benign Paroxysmal Positional Vertigo

David A. Zapala*
Shane A. Shapiro†
Larry B. Lundy*
Deborah T. Leming‡

Abstract

An acutely vertiginous 47-year-old woman presented to the emergency department with simultaneous acute left neurolabyrinthitis and left posterior canal benign paroxysmal positional vertigo (BPPV). Gaze nystagmus from the neurolabyrinthitis hampered diagnosis of the BPPV. However, once the BPPV was identified and treated, the patient's subjective vertigo improved rapidly. Concomitant BPPV should not be overlooked when a diagnosis of acute neurolabyrinthitis is made in the emergency department.

Key Words: Benign paroxysmal positional vertigo, neurolabyrinthitis

Abbreviations: BPPV = benign paroxysmal positional vertigo

Sumario

Una mujer de 47 años de edad se presentó al Servicio de Emergencias con un cuadro de vértigo agudo, con una neurolaberintitis aguda izquierda y un vértigo posicional paroxístico benigno (BPPV) del canal posterior izquierdo, simultáneamente. El nistagmo de la mirada de la neurolaberintitis interfirió con el diagnóstico del BPPV. Sin embargo, una vez que el BPPV fue diagnosticado y tratado, el vértigo subjetivo de la paciente mejoró rápidamente. La presencia concomitante de un BPPV no debe pasarse por alto cuando se realiza un diagnóstico de neurolaberintitis aguda en el Servicio de Emergencias.

Palabras Clave: Vértigo posicional paroxístico benigno, neurolaberintitis

Abreviaturas: BPPV = Vértigo posicional paroxístico benigno

*Department of Otolaryngology–Head and Neck Surgery/Audiology, Mayo Clinic, Jacksonville, Florida; †Department of Family Medicine, Mayo Clinic, Jacksonville, Florida; ‡Department of Physical Medicine and Rehabilitation, Mayo Clinic, Jacksonville, Florida

David A. Zapala, PhD, Department of Otolaryngology–Head and Neck Surgery/Audiology, Mayo Clinic, 4500 San Pablo Road, Jacksonville, FL 32224; E-mail: zapala.david@mayo.edu

Two common causes of acute vertigo are neurolabyrinthitis (sometimes referred to as "sudden vestibular collapse," "viral labyrinthitis," or "vestibular neuronitis") and benign paroxysmal positional vertigo (BPPV) (Koelliker et al, 2001; Delaney, 2003). Both typically occur without accompanying otologic or neurologic symptoms. Appreciating the time course of vertiginous sensations and studying the effects of provocative movements on observable nystagmus can readily distinguish the two conditions. Acute neurolabyrinthitis may be caused by a viral infection localized to the vestibular labyrinth or eighth nerve roots or by local ischemia within the labyrinth. Clinically, these events are indistinguishable from each other and produce a sudden onset of severe vertigo that crescendos over the course of several days, with residual symptoms of motion sensitivity and disequilibrium that may persist for several weeks.

In contrast, BPPV can be characterized by transient episodes of vertigo, lasting several seconds, which are typically provoked by moving the head into certain provocative positions. BPPV is a recognized sequela of acute neurolabyrinthitis, often developing weeks, months, or even years after the initial labyrinthine crisis. The various forms of neurolabyrinthitis are thought to predispose the utricle to shed otoconia. Over time, otoconia debris (canaliths) find their way into specific semicircular canals, resulting in the symptoms of BPPV.

In this brief report, we describe a patient in whom BPPV was present simultaneously with acute neurolabyrinthitis. The importance of this observation is twofold. First, this case demonstrates that events that cause BPPV after neurolabyrinthitis may occur rapidly, exacerbating the vertiginous sensations produced by the latter. Second, distinguishing between vestibular induced eye movements that result from anterior canal hypoactivity (neurolabyrinthitis) and posterior canal hyperactivity (BPPV) can lead the clinician to detect both conditions when they occur simultaneously in an acute setting.

REPORT OF A CASE

A 47-year-old woman arrived at the emergency department complaining of sudden onset of nausea, vomiting, and intense vertigo. Head movement exacerbated these symptoms. On examination, she was a well-developed, well-nourished woman in obvious discomfort with her eyes closed. She was afebrile. Initial vital signs revealed blood pressure of 138/73 mm Hg, heart rate of 72 beats per minute, respiratory rate of 20 breaths per minute, and oxygen saturation of 100%. She demonstrated what was described as "a very bad, fatigable, horizontal nystagmus bilaterally, with a positive Dix-Hallpike test." Findings on the remainder of the physical examination were unremarkable. The neurologic examination was limited by the severity of her nausea and vertigo. Laboratory and head computed tomographic studies were unremarkable. She was hydrated, given promethazine (for nausea) and diazepam (for suppression of vertigo), and admitted to the hospital overnight.

The following morning, she was evaluated by the attending physician who reported strong horizontal nystagmus with the eyes opened and a positive Dix-Hallpike maneuver to her right. Later that morning, she was reevaluated by a resident physician who noted a positive Dix-Hallpike maneuver to her left. She was discharged with a diagnosis of "acute labyrinthitis and BPPV." She was given a prescription for meclizine (for suppression of vertigo) and referred to the audiology department for canalith repositioning (Koelliker et al, 2001; Parnes et al, 2003; Baloh, 2004).

On presentation for canalith repositioning, she remained in distress, although her nausea had improved. She rated her vertigo as a "10" on a 10-point scale. A brief bedside examination (following Walker and Zee, 2000) revealed no focal neurologic signs. (All eye movements in this article are described relative to the patient's perspective.) A right-beating gaze nystagmus was present in the midline position, enhanced on rightward gaze and suppressed with visual fixation (Alexander second-degree nystagmus [Leigh and Zee, 1999]).[1] In addition, on rightward gaze, a brisk, persistent clockwise torsional component could be appreciated. Head thrusts were consistent with a left horizontal semicircular canal hypofunction (Baloh, 2004).

The underlying gaze nystagmus was observed on Dix-Hallpike testing to the patient's right. As the patient directed her eyes toward the examiner's head, the eyes

moved to the right of center gaze, revealing the right horizontal and clockwise torsional gaze nystagmus. As expected, this nystagmus was persistent. More importantly, the nystagmus disappeared when her gaze was directed to the left of midline and reappeared with her gaze redirected to the right.

Dix-Hallpike maneuvers to the patient's left provoked a robust, transient counterclockwise nystagmus that diminished over repeated trials, suggesting left posterior semicircular canal BPPV. Two canalith-repositioning maneuvers resolved the BPPV symptoms. Subjectively, the patient felt much better. She was instructed to stay upright for the next 24 hours and to avoid lying on her left side for the next six days. Although the canalith repositioning was successful, the Alexander nystagmus remained clearly evident in room light.

On ten-day follow-up, the patient noted that her vertiginous symptoms had greatly diminished immediately after canalith repositioning ("1" on a 10-point scale). Head turning, lying down, or rolling over in bed were no longer provocative, and her residual imbalance was gradually resolving. On examination, eye movements were intact, and no spontaneous nystagmus was evidenced in room light. There remained a right-beating spontaneous nystagmus with visual fixation removed. BPPV-provoking maneuvers were negative. Neuro-otologic and vestibular laboratory tests (normal audiologic evaluation, 51% left caloric weakness, preserved vestibular evoked myogenic potentials) confirmed the diagnosis of neurolabyrinthitis involving the left superior vestibular nerve.

DISCUSSION

BPPV is characterized by severe, transient vertiginous sensations provoked by moving into specific head positions (as in the Dix-Hallpike or Nylan-Bárány maneuver). Movement-provoked torsional nystagmus is typically fleeting but is readily detectable in room light with careful observation. Importantly, between provocations, the patient may feel unsteady and nauseous but typically does not experience true vertigo. Two of many excellent reviews of BPPV can be found in Parnes et al (2003) and Furman and Cass (1999).

In contrast to BPPV, acute neurolabyrinthitis presents as a near-continuous, severe vertiginous sensation that may be minimized by avoiding head movement of any kind. Acutely, spontaneous nystagmus may be appreciable with the patient sitting quietly, gazing straight ahead in normal room light. The nystagmus may increase or diminish with changes in gaze, but the horizontal component always beats in the same direction. Importantly, the nystagmus increases in intensity when visual fixation is removed. These symptoms reflect a sudden insult and resulting loss of vestibular tone in affected peripheral vestibular structures. Three excellent review articles can be found in Baloh (1998), Strupp and Arbusow (2001), and Tusa (2003).

Superior nerve neurolabyrinthitis is a subtype of neurolabyrinthitis in which labyrinthine dysfunction follows the distribution of the superior vestibular nerve. The underlying etiology may be a superior nerve neuronitis (Aw et al, 2001; Murofushi et al, 2003) or anterior vestibular artery occlusion (Hemenway and Lindsay, 1956; Schuknecht, 1969). In either case, the superior and horizontal semicircular canals and utricle are targeted. The inferior vestibular nerve, which innervates the posterior semicircular canal and saccule, is largely spared. This is demonstrated in our case by the persisting VEMP on the involved side. Because BPPV typically develops in the posterior semicircular canal, it is possible for it to develop simultaneously with acute superior nerve neuronitis. However, to our knowledge, no such case has been reported in the literature. Moreover, clinicians who are confronted with an acutely ill patient may not always check for BPPV when neurolabyrinthitis is clearly present.

In this case, confusion in identifying the side of BPPV involvement likely occurred as a result of the persistent torsional gaze nystagmus component (Fig. 1). Many audiologists and residents are taught to "bury the iris" when checking medial and lateral rectus integrity. It is not uncommon to see normal endpoint nystagmus during this check. However, in an acutely vertiginous patient, it is helpful to evaluate nystagmus when the eccentric gaze is much less than 40°. This can be accomplished by directing the patient's gaze laterally, to a point where the iris does not touch the lateral canthi. Persistent nystagmus is not normally seen in

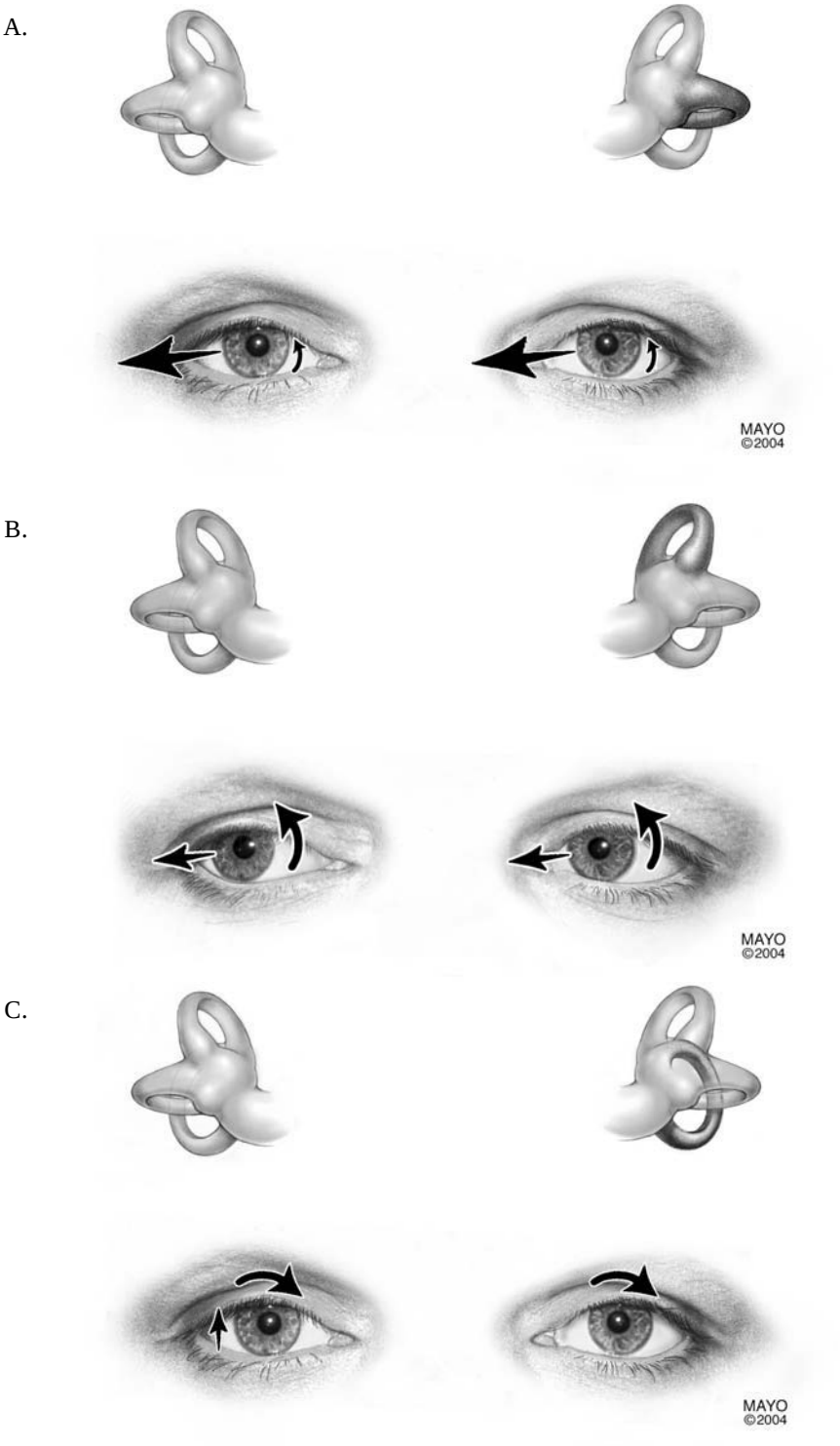

Figure 1. Nystagmus observed in acute left superior and horizontal canal hypofunction (superior vestibular nerve neurolabyrinthitis) and left inferior canal hyperfunction (benign paroxysmal positional vertigo [BPPV]). When the patient was sitting, there was a persisting right-beating and clockwise torsional gaze nystagmus evidenced with the eyes centered or with gaze directed to the right. This was compatible with a loss of tone in the left horizontal and superior semicircular canals (*A* and *B*). During the left Dix-Hallpike maneuver, a brisk counterclockwise torsional nystagmus was elicited as a result of transient, BPPV-induced left posterior canal hyperactivity (*C*). During the right Dix-Hallpike maneuver, the gaze nystagmus was mistaken for right BPPV. However, this was the same as the nystagmus observed in the sitting position, and it persisted as long as gaze was directed to the right. Recognizing the transient torsional nystagmus that beat in the opposite direction on the left Dix-Hallpike confirmed the left posterior canal BPPV. (By permission of Mayo Foundation for Medical Education and Research.)

this position. Any persistent nystagmus observed when the eyes are thus positioned is likely a pathologic spontaneous nystagmus that may also be observed during the Dix-Hallpike maneuver. Should this be seen, care must be taken to assess the persistence of observable nystagmus while controlling the direction of gaze during the Dix-Hallpike test.

Horizontal or torsional nystagmus could be appreciated with rightward gaze well short of 40° in this case. This nystagmus was observable without maneuvering the patient into a provocative position (Dix-Hallpike) and persisted as long as lateral gaze was held (Fig. 1B). The attending physician likely observed this gaze nystagmus and interpreted it as a positive right Dix-Hallpike test.

The Dix-Hallpike maneuver was negative to the left at least once. This is not surprising because BPPV symptoms wax and wane, diminishing with repeated head movements (adaptation and fatigue effects). Clinically, when provocative testing does not substantiate a strong history of BPPV, rechecking after the patient has been sitting or lying still for several minutes may reveal the condition.

The nystagmus provoked by the Dix-Hallpike maneuver to the left was consistent with posterior canal BPPV. The direction of the torsional nystagmus changed while the patient was sitting, from a clockwise nystagmus (stemming from left superior canal hypofunction) to a counterclockwise nystagmus (stemming from BPPV-induced transient left posterior canal hyperfunction) (Fig. 1B, C). Subjectively, this provoked severe vertigo and nausea. The patient aggressively guarded against moving her head into this provocative position. However, detection of BPPV ultimately led to rapid symptom improvement after canalith repositioning. We wonder how many times this condition might be missed for fear of provoking uncomfortable symptoms in patients with obvious neurolabyrinthitis.

In summary, acute superior nerve neurolabyrinthitis and BPPV can develop simultaneously. Table 1 presents the key features by which these two conditions can be recognized when they co-occur. Distinguishing between transient position-induced versus persistent gaze-induced torsional nystagmus can lead to the detection and treatment of co-occurring BPPV, with corresponding improvement in vertiginous symptoms.

NOTE

1. We follow the definition offered by Barber and Stockwell [1980] for "spontaneous nystagmus." They define "spontaneous nystagmus" as a nystagmus that is direction fixed and beating with about the same intensity in all head positions with the eyes closed [p. 145]. Some authors may refer to gaze nystagmus as "spontaneous" because it is present without provocation in the eyes' center position. We prefer to refer to nystagmus that is present with the eyes opened and fixated as a "gaze nystagmus" and then describe its behavior to avoid ambiguity.

Table 1. Distinguishing between BPPV and Superior Nerve Neurolabyrinthitis-Induced Eye Movements

Characteristic	Posterior canal benign paroxysmal positional vertigo	Acute superior nerve neurolabyrinthitis
Nystagmus description	Torsional and up-beating, in the plane of the dependent posterior canal, upper pole of the eye rolls toward the floor (counterclockwise torsional nystagmus for left posterior canal BPPV)	Horizontal and torsional, horizontal component beats away from the affected ear, torsional component rolls toward the contralateral posterior canal (ipsilateral anterior canal hypofunction)
Nystagmus provocations	Dix-Hallpike maneuver	Horizontal component follows Alexander's law (nystagmus is greater with gaze in the direction of the quick phase)
Nystagmus time course	Transient, with delayed onset, crescendo, and fatigue	Persists as long as provocative eye position is maintained

REFERENCES

Aw ST, Fetter M, Cremer PD, Karlberg M, Halmagyi GM. (2001) Individual semicircular canal function in superior and inferior vestibular neuritis. *Neurology* 57:768–774.

Baloh RW. (1998) Vertigo. *Lancet* 352:1841–1846.

Baloh RW. (2004) Differentiating between peripheral and central causes of vertigo. *J Neurol Sci* 221:3.

Barber HO, Stockwell CW. (1980) *Manual of Electronystagmography.* 2nd ed. St. Louis: CV Mosby Company.

Delaney KA. (2003) Bedside diagnosis of vertigo: value of the history and neurological examination. *Acad Emerg Med* 10:1388–1395.

Furman JM, Cass SP. (1999) Benign paroxysmal positional vertigo. *N Engl J Med* 341:1590–1596.

Hemenway WG, Lindsay JR. (1956) Postural vertigo due to unilateral sudden partial loss of vestibular function. *Ann Otol Rhinol Laryngol* 65:692–706.

Koelliker P, Summers RL, Hawkins B. (2001) Benign paroxysmal positional vertigo: diagnosis and treatment in the emergency department: a review of the literature and discussion of canalith-repositioning maneuvers. *Ann Emerg Med* 37:392–398.

Leigh RJ, Zee DS. (1999) *The Neurology of Eye Movements.* 3rd ed. New York: Oxford University Press.

Murofushi T, Monobe H, Ochiai A, Ozeki H. (2003) The site of lesion in "vestibular neuritis": study by galvanic VEMP. *Neurology* 61:417–418.

Parnes LS, Agrawal SK, Atlas J. (2003) Diagnosis and management of benign paroxysmal positional vertigo (BPPV). *CMAJ* 169:681–693.

Schuknecht HF. (1969) Cupulolithiasis. *Arch Otolaryngol* 90:765–778.

Strupp M, Arbusow V. (2001) Acute vestibulopathy. *Curr Opin Neurol* 14:11–20.

Tusa RJ. (2003) Dizziness. *Med Clin North Am* 87:609–641.

Walker MF, Zee DS. (2000) Bedside vestibular examination. *Otolaryngol Clin North Am* 33:495–506.