Benign Paroxysmal Positioning Vertigo with Indeterminate Cerebellar Lesion: Case Report

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Abstract

Of the numerous causes of dizziness, those that represent a life-threatening condition are rare. Physicians must guard against missing these rare but serious conditions while controlling the cost of the evaluation of patients who present with dizziness. This case study involving a 41-year-old female was written to illustrate the importance of systematic case history taking and of obtaining an ENG. The patient presented with classic symptoms of benign paroxysmal positioning vertigo (BPPV). The managing physician performed an MRI, which showed a cerebellar lesion. Results of a biopsy were negative. The patient's symptoms persisted, and she travelled to our clinic for further assessment. An ENG demonstrated a classic response to the Dix-Hallpike maneuvers, and a canalith repositioning maneuver was performed. The positioning dizziness resolved, and when contacted several months later, the patient stated she had remained asymptomatic.

Key Words: Benign paroxysmal positioning vertigo (BPPV), Dix-Hallpike maneuver, ENG, Epley maneuver

Benign paroxysmal positioning vertigo (BPPV) can be one of the easiest types of dizziness to detect and to alleviate. The dizziness is easily reproduced during the examination, with overt and specific signs. Barany (1921) first described the syndrome, which consists of a “burst” of dizziness accompanied by nystagmus, evoked exclusively by assuming specific head positions. Dix and Hallpike (1952) listed specific criteria to be met before applying this diagnosis. Their list included the following: (1) a history of dizziness occurring with head movements; (2) occurrence of dizziness accompanied by torsional nystagmus when the head was moved into the offending position; (3) response latency of 5 to 15 seconds; (4) a substantial diminution of the response within 30 seconds; (5) possible reversal of the nystagmus upon return to the sitting position from the offending position; and (6) reduction of response intensity upon repeat trials. Any patient whose responses do not meet these criteria should not be considered to have BPPV.

Using the recommended method of direct observation of eye movements during the Dix-Hallpike maneuver, the classic response is characterized by torsional nystagmus beating up and toward the undermost ear. Thus, head-hanging-right produces counter-clockwise rotary nystagmus and head-hanging-left produces clockwise rotary nystagmus. The response is accompanied by dizziness, which duplicates the patient's chief complaint. If the nystagmus is purely rotatory, it is not recordable with conventional electro-oculography, because the positively charged cornea and negatively charged retina do not deviate horizontally during the rotatory eye movement. In our experience, the nystagmus can often be recorded in either the vertical or the horizontal channel (or both), because it is rarely purely rotatory. We have, however, rarely evaluated anyone who became dizzy during the left or right head-hanging maneuvers who did not also have easily visualized nystagmus at the same time. Therefore, we routinely perform Dix-Hallpike maneuvers with eyes open and fixed, in a lighted room. If the patient's history is typical of BPPV, but the test result is negative (i.e., no dizziness and no nystagmus), we ask the patient if dizziness was expected with this type of movement. If the response is affirmative, we repeat the Dix-Hallpike maneuver, perhaps moving more quickly and waiting longer in the head-hanging position. Occasionally, a positive response is then evoked. If not, the patient may be in a period of spontaneous remission. It is then im-
portant to inquire about when the most recent symptoms occurred.

Many patients with BPPV present with a stereotypical description of brief, true vertigo that occurs only with quick movements. Characteristic situations when this might occur are turning in bed or reaching up to retrieve something from a high shelf. Some clinicians advocate performing the Dix-Hallpike maneuvers only if the patient has a history suggesting a position-related problem. In our experience, however, BPPV patients may report atypical types of dizziness or chronic imbalance as their major complaint. They may initially deny having any motion-related dizziness. After provoking a positive Dix-Hallpike response, during which the patient reports the typical "burst" of dizziness, the patients often comment that they do get this dizziness too, but that they are not troubled by it, since they have learned to avoid it. Other patients may report position-related dizziness, but may describe the dizziness in an atypical manner. Descriptions we have heard include "floating," "sinking," "dying," or just momentary lightheadedness. A physician or audiologist might be easily misled by such descriptions.

**ETIOLOGIC CONSIDERATIONS**

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An excellent review of BPPV was presented by Mohr (1986). He reviewed convincing evidence suggesting that the posterior semicircular canal is the specific site of dysfunction in most patients with BPPV. Surgery to destroy the function of the posterior semicircular canal, either by plugging the canal or by severing the posterior ampullary nerve, has been effective in eliminating BPPV. Parnes and McClure (1991) have described visualizing free-floating particles in the posterior semicircular canal during canal-plugging surgical procedures. The exact cause of BPPV, however, remains undetermined. Various investigators (Stahle and Terins, 1965; Herndon et al, 1976; Katsarkas and Kirkham, 1978; Baloh et al, 1987) have examined coexisting findings for groups of patients with BPPV. While the vast majority have no other known pathologies, some patients have a concurrent diagnosis of disturbances such as endolymphatic hydrops, acoustic neuroma, labyrinthitis, a history of head trauma, or surgical procedures. A few have concurrent central problems. Animal experiments have demonstrated paroxysmal positioning nystagmus subsequent to inducing cerebellar lesions (Fernandez et al., 1959). There is some question, however, as to whether the resultant nystagmus is rotatory, with the upper pole beating toward the undermost ear (Baloh et al, 1979b). Speculation that BPPV may have central causes appears to be based upon the coexistence of a lesion within the central nervous system, found at the same time the patient complained of positional vertigo. Thus, there is no strong evidence of any causal relationship between a central lesion and BPPV.

A primary weakness in the evaluation and diagnosis of these patients is that the definitive criteria set forth by Dix and Hallpike have not been strictly or rigorously applied by all investigators (Mohr, 1986). Another potential weakness is that the examiner who evaluates the patient makes the determination of these criteria based upon subjective observations. The subjectivity of these judgments may cause variation from examiner to examiner, based on their experience in determining the direction of the fast phase of nystagmus and on patient cooperation at the time of the test. Such problems must be kept in mind when reviewing records for a given patient or when evaluating literature on BPPV.

In our caseload, between 5 and 10 percent of the patients have a final diagnosis of BPPV, based upon a classic Dix-Hallpike response. Other facilities report prevalences of 10 percent (Drachman and Hart, 1972) to 17.5 percent (Barber, 1984). In view of the frequency of occurrence and the wide variety of descriptions of the problem, the Dix-Hallpike maneuver is best used as a standard test in the assessment of the dizzy patient. The procedure typically takes less than 5 minutes to perform, and could be performed as part of a routine clinical assessment, as it requires no special equipment other than an examining table.

Even though the demonstration of BPPV with the Dix-Hallpike maneuver is straightforward for an experienced examiner, it is not uncommon that patients who have complained of positional dizziness have been misdiagnosed or have remained undiagnosed. Such outcomes occur in part because patients seek medical help for their dizziness from physicians within a variety of different specialties. Unfortunately, physicians vary widely in their knowledge about dizziness and its assessment. Some physicians may not have easy access to objective tests of vestibular function. Further, individuals performing these tests (including audiologists) have a wide range of skill in performing and interpreting the tests accurately.
PATIENT PRESENTATION

In March of 1992, a 41-year-old female came to our clinic after unsuccessful and unusual evaluation and treatment for dizziness elsewhere. She stated that in the spring or summer of 1988, she had experienced brief episodes of vertigo, as well as chronic, on-feet imbalance, which disappeared in about 1 month without treatment. In May of 1990, she experienced a recurrence of the symptoms. Her dizzy spells had never lasted more than a few moments and had always been precipitated by motions such as rolling over in bed to the left and getting out of bed in the morning. She stated that her symptoms had not varied from 1990 to the time of our evaluation. She consulted a physician in her home town, who ordered a magnetic resonance image (MRI) and an audiogram, but no vestibular tests (Fig. 1). Audiometry indicated normal hearing, bilaterally. Contralateral acoustic reflexes were reported as normal.

MRIs were done in July of 1990 and in March, May, and November of 1991 (Fig. 2). All MRIs demonstrated a lesion in the right cerebellum just posterior and lateral to the fourth ventricle with no mass effect, and there was no enhancement with gadolinium. She underwent a stereotaxic biopsy of the lesion, which showed normal cerebellar tissue. Following the biopsy, her speech was slurred, and she noticed difficulty with her coordination. Her motion-related dizzy spells persisted.

Neurologic evaluation within our clinic demonstrated normal mental status, language, blood pressure, and cranial nerve function. She had a slight dysarthria, and her muscle stretch reflexes were present. She also showed a terminal tremor and reduction of alternating motion rates in the right upper extremity. Brainstem auditory evoked potentials were normal. It was concluded by the managing neurologist that the cerebellar lesion probably represented a congenital cyst, which would continue to produce no symptoms.

The patient was referred to our department for vestibular evaluation. Electronystagmography (ENG) and computerized dynamic posturography tests were conducted (Fig. 3). The warm water caloric irrigations demonstrated strong and symmetric responses (right, 67 degrees/sec, and left, 55 degrees/sec). Gaze testing, visual pursuit, fixation suppression, and ocular saccades were all well within normal limits (Fig. 4A–C). There was some mild, direction-fixed, right-beating positional nystagmus with eyes closed and alerting in the supine, head-left, and head-hanging positions (Fig. 5A–C). The computerized dynamic posturography
test was normal for both the motor and sensory portions of the test (Fig. 6A, B).

The Dix-Hallpike maneuver demonstrated a classic response in the head-hanging-left position. The response consisted of paroxysmal, clockwise nystagmus, which began approximately 5 seconds after assuming the position. The response was accompanied by the patient's report that she was dizzy and that the sensation duplicated the spells she had been having. The response was much weaker on the second trial. There were no other positions tested that provoked the patient's symptoms of dizziness. Based on these observations, a diagnosis of benign paroxysmal positioning vertigo was made, and a maneuver to relieve the symptoms was performed. Today, 7 months later, she reports periodic slurring of her speech, but her position-related vertigo has completely resolved.

The maneuver we used is called the canalith repositioning procedure, first reported by Epley in 1980. A more refined procedure was described by Epley (1992) as follows (Fig. 7):

1. The patient is seated on a table so that when laid back, his/her head will extend off the end 45 degrees toward the affected ear. In this position, it is essential to wait a minimum of 30 seconds for the delayed response. (We have seen responses delayed as much as 40–50 seconds for some patients.)

2. Following cessation of the nystagmus, the head is rotated 90 degrees (45 degrees off center, away from the affected ear). The patient is kept in this position for a duration equaling the delay in the response plus the duration of the nystagmus (T sec = delay + duration).
3. The patient’s head and body are then rotated 135 degrees from the supine position (the patient rolls over onto the shoulder opposite the affected ear), and that position is maintained for another like time period, i.e., T sec.

4. Maintaining the same head position, the patient is then raised slowly to a sitting position and held there for T sec.

5. The patient’s head is then turned forward and down 20 degrees for T sec.

The patients are asked to visually fixate in each position. These five steps are repeated until there is no nystagmus or subjective dizziness in any of the positions.

Written instructions are given to the patients and discussed with them after completing the maneuver. These instructions include keeping the head as erect as possible for 48 hours, avoiding excessive head movements (especially in the vertical plane), and not lying with the affected side down for 9 days. Complete instructions are found in the Appendix. When these instructions have been completed for the specified length of time, the patients are asked to return to normal activities without movement restrictions.

**SUMMARY**

A patient having a classic history and clinical findings for BPPV was reviewed. Vestibular testing was bypassed initially in favor of MRIs and led to neurosurgery for what appeared to be a benign, congenital cerebellar cyst. Further testing at our facility demonstrated results consistent with classic BPPV.
Figure 5A Recording demonstrating right-beating positional nystagmus obtained with eyes closed and alerting in the supine position.

Figure 5B Recording demonstrating right-beating positional nystagmus obtained with eyes closed and alerting in the head-left position.

Figure 5C Recording demonstrating right-beating positional nystagmus obtained with eyes closed and alerting in the head-hanging position.
Positional vertigo was satisfactorily resolved with a canalith repositioning procedure. This observation implies an unlikely relationship between the patient’s symptoms and the cerebellar lesion. This case illuminates the importance of ENG with Dix-Hallpike testing. It should be emphasized that the treatment procedures described here should be applied only after the presence of BPPV has been confirmed. This particular form of patient management should not preclude comprehensive medical assessment to rule out any coexisting, treatable pathology.

REFERENCES


Figure 7  "Positions for the canalith repositioning procedure (CRP), targeting left posterior semicircular canal (PSC). Dark figure, side view; boxes, operator’s exposed view of left labyrinth, showing gravitating canaliths. Semicircular canals are labeled. S (Start), patient is seated, operator behind, oscillator applied. 1, Head is placed over the end of the table, 45 degrees to the left (canaliths gravitate to center of PSC). 2, While head is kept tilted downward, it is rotated to 45 degrees right (canaliths reach common crus). 3, Head and body are rotated until facing downward 135 degrees from supine position (canaliths traverse common crus). 4, While head is kept turned right, patient is brought to sitting position (canaliths enter utricle). 5, Head is turned forward, chin down 20 degrees. General: Pause at each position until induced nystagmus approaches termination, or T a (delay + duration) if no nystagmus. Keep repeating entire series (1 through 5) until no nystagmus any position pg. 401." (Reprinted from Epley J. [1992]. The canalith repositioning procedure: for treatment of benign paroxysmal positional vertigo. Otolaryngol Head Neck Surg 107:401-404. With permission from Dr. John Epley and Otolaryngology—Head and Neck Surgery.)

1Note: Oscillator was not used while carrying out maneuver for patient reviewed.

APPENDIX

Patient instructions for the Epley Maneuver:

1. For the next 2 days and nights (48 hours), you should keep your head completely vertical. To sleep, you might sit in a recliner chair, but do not lie all the way back; just far enough to support your head. You might devise a support for your head by purchasing a neck brace or by pinning two pillows next to your head.
2. Next, please avoid any head movements upward or downward for the next week and try to move your head and body as a unit without excessive turning of your neck.
3. Do not sleep on the side that generates your dizziness for an additional week. You might pin a pillow to that shoulder to keep you from rolling over on it during the night. For example: if your right ear down is causing the problem, then sleep on your left side or stomach.