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REVIEW ARTICLE

Benign positional vertigo and endolymphatic hydrops: what is the connection?

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Abstract

Background: Although benign paroxysmal positional vertigo and endolymphatic hydrops are considered to be distinct diagnoses, a minority of vertiginous patients exhibit features of both conditions. This coincidence has been reported previously in the literature, and is reviewed here in terms of possible aetiology.

Results and conclusion: A new hypothesis to account for both conditions is offered, implicating free-floating degenerating debris from the otolithic apparatus. It is postulated that the gelatinous/proteinaceous component may account for an osmotically induced hydrops, while the calcified fragments may induce positional vertigo.

Key words: Endolymphatic Hydrops; Benign Positional Vertigo

Introduction

While the textbook definitions of benign paroxysmal positional vertigo (BPPV) and endolymphatic hydrops are distinct, they are also somewhat restrictive. The definitions exclude patients with peripheral vertigo who do not neatly fit into either category, but who have a clinical picture that incorporates characteristics of both conditions. In fact, many patients with positional vertigo develop symptoms in multiple positions, and have a history of intermittent dizziness that is variable, and either fails to respond or recurs after an Epley manoeuvre has been performed. Similarly, some patients with endolymphatic hydrops may demonstrate episodic vertigo that is not only spontaneous but also positionally triggered.

Case report

The following three cases exemplify the clinical presentation of patients with characteristics of both BPPV and endolymphatic hydrops.

Case one

A 57-year-old woman presented with a 6-month history of spontaneous and positional vertigo. She had spontaneous episodes of an unprovoked sudden sensation of turning to her right. These episodes were followed by persistent dizziness, with nausea and vomiting for several hours, and right aural fullness. She also had recurrent positional dizziness on turning her head to the right. Clinical evaluation demonstrated right-beating nystagmus on Dix–Hallpike positioning with the head right and the head down. Caloric testing was normal. Electrocochleography was positive for the right ear (SP/AP ratio = 0.530) and negative for the left ear (SP/AP ratio = 0.312). The diagnosis of right BPPV and right endolymphatic hydrops was made.

Case two

A 51-year-old woman presented with a 1-year history of recurrent vertigo, with nausea and vomiting. The symptoms were worse with sudden head movements and on tilting her head back. Clinical evaluation revealed positive Dix–Hallpike test results on both left and right lateral and down positions. Calorics were normal, electrocochleography was positive for the right ear (SP/AP ratio = 0.608) and negative for the left ear (SP/AP ratio = 0.140). The diagnosis of bilateral BPPV and right endolymphatic hydrops was made.

Case three

A 67-year-old man presented with recurrent vertigo on turning his head to the left side. He was generally off balance with sudden movements, and on bending over. There was no spontaneous vertigo. He had a past history of left BPPV, which was relieved with the Epley manoeuvre five years ago. The condition returned two years ago, with positional vertigo on lying down and hyperextending of the neck, which again improved with repositioning. On examination, the Dix–Hallpike test result was positive on the left

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side. Calorics were normal, but electrocochleography was positive for the left ear (SP/AP ratio = 0.500) and negative for the right ear (SP/AP ratio = 0.313). The patient was diagnosed with BPPV and endolymphatic hydrops in the left ear.

Discussion

Is there a connection between BPPV and endolymphatic hydrops? A possible epidemiological link was noted by Mizukoshi *et al*.[1] In a study of two populations with BPPV in Japan, the authors concluded: 'Compared with other epidemiological features of Ménière's disease and sudden deafness with vertigo in other series, it appeared that the characteristic features of BPPV are epidemiologically similar to those of Ménière's disease, but different from those of sudden deafness'.

Statistical coincidence may account for a minority of cases, but the coincidence rate seems higher than chance would allow. Various estimates of the incidence in the general population suggest rates of 2.9–17 per 100 for BPPV and 15.3–46 per 100 000 for endolymphatic hydrops. Based on these statistics, coincidence should account for only about a 0.1 per cent overlap. However, Gross *et al*. reviewed 162 patients with Ménière's disease and reported that 5.6 per cent had coincident intractable BPPV.[2] Our personal review of 100 patients with complex and not clearly categorisable peripheral vertigo demonstrated that 10 per cent had both positional vertigo and hydrops as demonstrated on videonystagmography and electrocochleography. It would appear that the coincidence of the two conditions is significantly higher than might be statistically anticipated.

Post-traumatic degeneration following head injury may be one common causative factor. Ernst *et al*., in a study of patients with injury to the head or craniovertebral junction, found that 12 out of 63 cases (19 per cent) had endolymphatic hydrops.[3] In a review of dizziness following mild head injury, Hoffer *et al*. reported BPPV in 28 per cent of cases.[4]

Viral injury to the membranous labyrinth was postulated as a common cause by Gacek.[5] In a histological study, he noted that BPPV, vestibular neuronitis and Ménière's disease all show similar degeneration in the vestibular ganglion. Clinically, he found that herpes virus reactivation in the vestibular ganglion can cause BPPV, and 66 per cent of BPPV symptoms were relieved by antiviral medication.

Other pathological phenomena may also be involved. Karlberg *et al*. (2000) concluded that any inner-ear disease that detaches otoconia but does not destroy posterior semicircular canal function can cause BPPV.[6]

The otoconia were first implicated in BPPV in 1969, by Schuknecht, who identified a basophilic deposit embedded in the cupula of the posterior semicircular canal of a patient who suffered from BPPV.[7] He coined the phrase 'cupulolithiasis' for this condition. Converting the ampulla from a rotation sensor to a gravitational sensor appeared to account for the clinical findings of BPPV. Cupulolithiasis seemed to explain many features of classic BPPV, including single position, repeatability, latency and fatigability.

A subsequent study of 556 temporal bones by Moriarty *et al*. identified multiple cases of basophilic cupular deposits involving all 3 semicircular canals, suggesting that these deposits are more prevalent, and may be responsible for multipositional vertigo.[8]

However, cupulolithiasis did not adequately account for intermittent vertigo, coincidental positional and non-positional vertigo, and coincidental vestibular weakness. Furthermore, the concept of BPPV caused by a basophilic deposit embedded in the gelatinous matrix of the cupula does not explain the high success rate of the Epley manoeuvre.

In 1992, a clinical finding by Parnes and McClure fundamentally changed otological thinking about BPPV.[9] On opening the posterior semicircular canal in patients with BPPV, they found loose particles, floating freely within the endolymphatic fluid. They postulated that BPPV was not a result of cupulolithiasis, but rather of hydrodynamic drag exerted by debris on the endolymph, due to gravitational pull.

A subsequent, larger study by Welling *et al*. confirmed the presence of free-floating particles in patients with BPPV, but not in asymptomatic patients.[10] These authors attributed the source of the material to degenerating otoconia, and concluded that free-floating endolymphatic debris is responsible for positional vertigo. More recently, Kao *et al*. confirmed with scanning electron microscopy that debris in the posterior semicircular canal of patients with BPPV consists of fragments of otolithic membrane with embedded otoconia.[11] However, Kveton and Kashgarian also found debris in the posterior semicircular canal of patients who were undergoing translabyrinthine removal of an acoustic neuroma.[12] Electron microscopic study suggested that the debris was a mineral and protein material. The significance of this paper is that these patients did not have vertigo. As the surgical studies[9,11] only investigated the labyrinths of patients with BPPV and who were undergoing posterior canal occlusion, they (for obvious reasons) lacked a control group for comparison.

Degenerating otolith apparatus appears to be a prime potential source of both calcified and proteinaceous material. In addition to the otoliths, which are calcified protein, the otolith apparatus also includes anchoring proteins in the gelatinous layer, and a subcupular meshwork, in addition to hair cells and supporting cells.

But can endolymphatic debris also cause hydrops? Waltner and Raymond sampled endolymph during an endolymphatic sac shunt procedure, and demonstrated that, in at least one Ménière's patient, endolymph had twice the protein content of cerebrospinal fluid.[13] In 1976, Dohlman postulated increased colloid pressure exerted by endolymphatic cellular debris, and

suggested that hydrops might be due to the increased water binding capacity of proteinaceous debris.[14] He developed an experimental model to investigate osmotic hydrops by injecting mannitol into the endolymphatic space.[15] Johnsson *et al*. studied hydrops associated with cochlear otosclerosis, and concluded that endolymphatic hydrops in this condition was probably due to 'altered composition and density of the endolymph'.[16] It would seem that, at least in some cases of endolymphatic hydrops, increased osmotic pressure within the membranous labyrinth may play a role.

If cellular debris is causally implicated in both positional vertigo and endolymphatic hydrops, what might the linking mechanism be? Gross *et al*. proposed that Ménière's disease may predispose patients to intractable BPPV, stating 'Hydropically induced damage to the maculae of the utricle and saccule, or partial obstruction of the membranous labyrinth may be possible mechanisms that explain the coincidence of Ménière's disease and BPPV'.[2]

We would like to offer a different hypothesis, based on four observations in the medical literature. First, the coincidence of BPPV and endolymphatic hydrops seems to be significantly greater than statistical coincidence. Second, in at least some cases, the endolymphatic compartment appears to contain free-floating debris, comprising protein and mineral particles of varying size, weight and consistency. Third, endolymphatic debris may be asymptomatic, or be associated with BPPV. Fourth, conditions resulting in increased colloid pressure in the endolymphatic compartment may predispose to endolymphatic hydrops.

Conclusion

We propose that, depending on the physical characteristics of its components, endolymphatic debris may be asymptomatic, or may cause either or both positional vertigo and hydrops. The cellular and proteinaceous components may generate colloidal osmoactivity, while the calcified elements may be subject to gravitational pull. The amount, location and composition of endolymphatic debris could explain the coincidence of positional vertigo and osmotically induced hydrops in some patients.

This hypothesis offers one possible mechanism to link the two conditions, and may explain the coincidence of BPPV and endolymphatic hydrops. It does not account for every aspect of the clinical picture in complex cases of peripheral vertigo; other factors, such as regional sludging of debris in the semicircular canals, may also play a role. Certainly, the dynamic concept of temporary endolymphatic traffic jams caused by impacted sludge more elegantly addresses such findings as intermittent and multipositional vertigo, a good initial response to the Epley manoeuvre, and unpredictable subsequent recurrence, as well as the spontaneous resolution of positional vertigo.

Clearly, more studies are needed if we are to arrive at a unifying theory such as suggested by Phillips and Prinsley, who, in 2009, went so far as to propose that 'the three distinct syndromes of vertigo (labyrinthitis, Ménière's disease and BPPV) which arise from a malfunction of the vestibular labyrinth are in fact a spectrum of disorders all resulting from the presence of free-floating particles within the vestibular fluid chambers'.[17]

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