Spontaneous Nystagmus in Benign Paroxysmal Positional Vertigo: Is It A New Sign?

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Abstract- Benign Paroxysmal Positional Vertigo (BPPV) is a condition that indicates a benign inner ear disorder. It is generally believed that BPPV is due to the dislodged otoconial particles from otolith organs and unusual collection of them within any of semicircular canals or even in all three semicircular canals. Although the typical features of nystagmus in BPPV have been well-studied, very few studies (just four articles) have highlighted the presence of spontaneous nystagmus in BPPV recently. During the past 10 years, 2850 patients have been examined at the audiology unit of our department, and 254 patients have received diagnoses of BPPV but recently 2 patients presented with BPPV and spontaneous nystagmus, a new symptom that has been never observed in our clinical records. We herein describe this rare symptom in 2 case of BPPV. A 50-year-old woman with BPPV who showed an 18 degree spontaneous nystagmus treated with Epley maneuver and a 53-year-old man with 3 degree spontaneous nystagmus.

Keywords: Benign Paroxysmal Positional Vertigo; Horizontal canal BPPV; Spontaneous nystagmus

Introduction

BPPV is the most common cause of recurrent vertigo (1). To date, two theories are considered as main theories for benign positional vertigo pathophysiology: canalolithiasis and cupulolithiasis. In the first condition, detached otothlic particles from utricle enters semicircular canals and cause deflection of the cupula by inducing endolymph flow during head motion. These free-floating particles give rise to stimulation of hair cells and provoke vertigo. In cupulolithiasis theory, the otothlic particles becomes attached to the cupula, so specific gravity of cupula becomes heavier, thus responding to any change in gravitational position of the head (rather than angular acceleration)(2). However, canalolithiasis is currently accepted pathophysiology mechanism of typical BPPV (1,2). Although the exact etiology of BPPV is still unclear, it not only can be associated with some other disorders like head trauma, viral labyrinthitis, vertebrobasilar ischemia and vestibular neuritis but also can be a distinct dysfunction especially in elderly.³ However, most of all reported cases are idiopathic in nature (3,4).

BPPV have some general symptoms. The main symptom of BPPV is vertigo (spinning sensation) induced by a change in head position with respect to gravity. Episodic vertigo usually has short latency about 5-10 seconds (2), the brief duration from a few seconds to one minute and is fatigable (reduction in vertigo and nystagmus if positioning maneuvers is repeated). It can also be associated with nausea and vomiting. BPPV often resolves spontaneously within a few weeks or months and affects females more than males (5). It usually occurs when patients have a rapid head movement or performing a provocative position such as turning in the bed, standing up/lying down or bending over (1). The incident of idiopathic BPPV has been reported as 10.7-64 per 100000 population (5). BPPV can affect any of three semicircular canals, but posterior canal BPPV (PC-BPPV) has been said to account for 60-90% of all BPPV cases (6). In 1985 McClure described HC- BPPV that involves horizontal canal (7). A few years later in 1994, Steddin & Brandth presented BPPV of anterior canal that is a rare type of BPPV (8). HC-BPPV now appears to be more prevalent than was previously thought: it accounts for 17% of BPPV (9). There is also a mixed canal type that several canals are affected simultaneously (10).

Diagnosis of BPPV is based mainly on a history of characteristic positional vertigo along with the classical symptoms that stated previously. However type of nystagmus would distinguish three variant of this
disorder. In PC-BPPV, the Dix-Hallpike maneuver or the lateral head-trunk tilt are used to confirm the diagnosis of posterior canal involvement. In this condition, there is typically an up-beating nystagmus in uppermost eye and a geotropic torsional component beating toward the affected ear in the undermost eye (11). Geotropic torsional nystagmus in undermost eye with predominant down-beating component in uppermost eye during straight head hanging (SHH) or Dix-Hallpike maneuvers has been ascribed to AC-BPPV when there are no central pathologies (2,11). HC-BPPV is diagnosed by the supine roll test (the Pagnini-McClure maneuver) in which the head is turned by about 90° to each side while supine. Horizontal canal involvement manifest itself with horizontal nystagmus that would can be either geotropic or ageotropic (1). Spontaneous nystagmus is not included in classical symptoms of BPPV, but in recent reports, 66-76% of HC-BPPV patients exhibited spontaneous nystagmus (12,13).

In this study, we described 2 patients, diagnosed with BPPV and had this new symptom of spontaneous nystagmus.

Case Report

Case 1

A 50-year-old woman was referred to us with complain of vertigo, disequilibrium, nausea and vomiting lasting for 1 year. Vertigo had decreased her quality of life significantly, so she was experiencing a long-term stress. She had been evaluated by a Gastroenterologist for vomiting and used some pills to prevent it, but there was no change in her symptoms. We performed Videonystagmography (VNG) to detect if any vestibular lesion exists. Results were presented as follow:

On first examination, an 18 degree Spontaneous nystagmus was seen (Figure 1). Interestingly it disappeared with tilting of head and fixation. There is no nystagmus in gaze testing. In saccade test, latency was 450 ms. Tracking was saccadic in both right and left side. Optokinetnic nystagmus was normal and symmetric. Right Dix-Hallpike positioning testing provoked vertigo with rotatory nystagmus in the right eye and upbeating nystagmus in the left eye. Similarly, in left positioning maneuver, there was a rotatory nystagmus in the left eye and an upbeating nystagmus in the right eye. In both side positioning, vertigo and nystagmus was latent and fatigable. In positional testing, there was a 6 degree right-beating nystagmus (ageotropic) in supine head right position. In supine head left position also, a 7 degree right-beating nystagmus (ageotropic) was seen. In rolling maneuver in both right and left head turning, there was an ageotropic nystagmus and vertigo, more severe in left head turning. In caloric testing, left ear hypoactivity was seen. We suspected to PC-BPPV in both ears, and horizontal canal cupuluthiasis in the right ear, so Home-Epley maneuver, three times a day for two weeks was recommended.

After 2-week home-Epley maneuver

She was evaluated again after 2-week Home-Epley maneuver. Vertigo, disequilibrium, nausea and stress had reduced significantly; spontaneous nystagmus had reduced to a 6 degree right-beating nystagmus (Figure 2). Left Dix-Hallpike maneuver was negative, but positioning to right evoked a right-beating nystagmus without vertigo. Saccade and smooth pursuit testing was normal.

Figure 1. Spontaneous nystagmus testing at first examination demonstrating an 18 degree spontaneous nystagmus.

Figure 2. Spontaneous nystagmus testing after 2-week Home-Epley maneuver.
After 4-week home-Epley maneuver

A further 2-week Home-Epley maneuver was recommended. Spontaneous nystagmus had been disappeared completely (Figure 3). Dix-Hallpike maneuver was negative in both side positioning, and she had no symptoms of vertigo, nausea, vomiting and stress. All symptoms had been recovered.

Case 2

A 53-year-old man with 2-year vertigo lasting for a few minutes was presented to us. He had complains of continuous dizziness, nausea and vomiting. Vertigo was provoked when he was bending over, rolling on the bed and looking up.

Results of VNG

There was a 3 degree right beating spontaneous nystagmus that disappeared when the head was bent 30 degree forward. There is no nystagmus in gaze testing. Saccade, optokinetic and smooth pursuit was normal. In right Dix-Hallpike maneuver, a rotatory nystagmus beating toward the right eye and an upbeating nystagmus with 30 second latency was observed. There was also a fatigable vertigo. Left Dix-Hallpike was negative. In rolling maneuver, ageotropic nystagmus with vertigo was seen in both head turning. It persisted for 60 second, and it was more severe in left positioning direction. Caloric test revealed unilateral weakness and direction preponderance but suppressed with fixation. Results indicated a right posterior and horizontal canal BPPV. Epley maneuver was performed, and 2 week Home-Epley maneuver was recommended.

Two weeks later, Dix-Hallpike and rolling maneuver was negative and spontaneous nystagmus had been disappeared.

During the past 10 years, 2850 patients have been examined at the audiology unit of our department, and 254 patients have received diagnoses of BPPV, but only 2 patients presented BPPV and spontaneous nystagmus recently.

Discussion

We have recently observed spontaneous nystagmus in 2 patients with BPPV. Although a spontaneous nystagmus is not included in classical signs of this disorder, recently there have been some reports of spontaneous nystagmus in BPPV; a not well- documented sign. It’s reported that 66-76% of HC-BPPV patients exhibited spontaneous nystagmus (13,14). In 2001,, Beren (14) reported spontaneous nystagmus in a 25 year-old woman with head injury. At the first step, the right PC- BPPV was diagnosed and revealed after a single Epley maneuver. A few days later, Dix-Hallpike positional testing was negative but in roll testing, the right HC-BPPV was diagnosed, and Head-Shaking maneuver was recommended. On following day examination, there was a spontaneous vertigo and nystagmus that did not alter with positioning maneuver. Spontaneous nystagmus was predominantly horizontal, away from the affected ear and slow phase velocity of 11.4. There was also a small vertical & torsional component with slow phase velocity of 3.2 and 2.9 respectively. In 2002 Bisdroff (15) presented 2 cases with history of positional vertigo that manifested spontaneous nystagmus while sitting, lying down and in the supine position with either ear downwards. There was a permanent apogeotropic nystagmus too. In pitch, the nystagmus stopped when the head was slightly tilted forwards, and the direction was reversed when the face was turned downwards. The spontaneous nystagmus was similar to the sitting position. A null position for the nystagmus in the supine position was found when the head was slightly turned to one side. In 2008, Asperrla (13) termed spontaneous nystagmus as a pseudo-spontaneous nystagmus. He examined 293 patients with horizontal canal BPPV and observed spontaneous nystagmus in 222 cases. He described spontaneous nystagmus as a long lasting non-paroxysmal nystagmus that is caused by involuntary horizontal rotation of head because this rotation causes slow floating of otoliths. In head pitch test when patients bent the head 30 degree forward, pseudo-spontaneous nystagmus disappeared, and when bent it 60 degree forward, its direction reversed. It was different from null position of what Bisdroff had observed. In 2010 De Stefano et al. (16) examined data of 412 patients with
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idiopathic BPPV in a retrospective study. 70 patients had HC-BPPV and spontaneous nystagmus that it disappeared by bending the head, so he didn’t accept the theory of Beren for spontaneous nystagmus. He also remarked that it doesn’t occur only at apogeotropic type. He proposed that spontaneous nystagmus is based on natural inclination of horizontal canal with respect to the horizontal plane. Von Beren believed that the likely mechanism of spontaneous nystagmus in BPPV is plugging of the Horizontal canal by otocinia, negative pressure on the cupula and thus permanent utriculofugal deflection of horizontal canal cupula. In our opinion when spontaneous nystagmus exist, symptoms like vomiting and nausea maybe increase and self-limited recovery of BPPV decrease. This new sign can be helpful in diagnosis of BPPV types and therefore the right treatment. We were unable to confirm the Von Beren theory because it appears that plugging of horizontal canal, induce spontaneous nystagmus in any condition, but in our case it disappeared with head tilting. We accept De Stefano and Asperrla's idea that believe spontaneous nystagmus in BPPV is because of natural anatomical position of horizontal canal which is inclined 30º backwards from the horizontal plane. The gravitational force may affect the otolithic particles inside the canal or the heavy cupula, even when in the upright sitting position. For the same reason, spontaneous nystagmus disappears when the patient’s head is bent forwards by about 30º in our and previous cases. In this position, since the horizontal canal is aligned with respect to the earth horizontal plane, the effect of gravity is negated.

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References