Vertical “pseudospontaneous” nystagmus in a patient with posterior canal BPPV: case report

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A 34-year old men presented with 2-days history of quick episodes of vertigo caused by changes in the position of the head, mainly when turning over in the supine position. Video-Frenzel examination revealed a mild spontaneous downbeating nystagmus only with visual fixation removed that robustly increased in the bow test. The lean test and the Dix Hallpike test to the right side revealed an upbeating torsional nystagmus with the upper pole of the eye beating to the right ear. Then, the Epley maneuver was performed, given the clinical suspicion of right posterior canal BPPV, and he achieved immediate resolution of vertigo and nystagmus. Presumably, this is the first report in which a patient with posterior canal BPPV demonstrated a vertical ‘pseudospontaneous’ nystagmus.

Introduction

Benign paroxysmal positional vertigo (BPPV) is a very common cause of vertigo. The pathogenesis mechanism may be related to canalolithiasis, when otoliths are floating in the semicircular canals or cupulolithiasis, when attached to the cupula.

In most patients the posterior canal is impaired, succeeded by the lateral canal, while the anterior canal is rarely affected [1]. The posterior canal variant usually manifests with a vertical torsional nystagmus induced by the positional tests, such as the Dix Hallpike test.

The nystagmus observed in most cases of BPPV is usually typical and easy to differentiate from central diseases and other pathologies that may manifest with positional nystagmus. However, some patients may demonstrate atypical nystagmus patterns that may be misdiagnosed. Hence, knowing pathophysiology and anatomy of the semicircular canals is important to clarify the biomechanical etiology of the positional nystagmus induced by the action of gravity.

In order to explain the evoked nystagmus in patients with BPPV it is first necessary to consider the location of the otoliths in the starting position [2]. In posterior canal BPPV the debris are usually located in the most inferior part of the ampullary arm of this canal, next to the ampulla. When patients are submitted to the Dix Hallpike test to the affected side, the otoliths fall away from the ampulla, emerging in an ampullofugal excitatory endolymphatic flow. The posterior canal excitation activates the ipsilateral superior oblique and the contralateral inferior rectus muscles. The primary action of these muscles are respectively, incyclotorsion and depression of the eyes. Hence, the provoked nystagmus (fast phase) is geotropic vertical torsional with the vertical component upwards and the upper pole of the eye beating to the undermost ear.

Downbeating nystagmus is a clinical sign usually seen in central lesions such as cerebellar disorder. However, since BPPV is considered a biomechanical disorder, it could be observed induced by head position changes in relative to gravity. It has been established that both anterior canal BPPV and apogeotropic posterior canal BPPV are characterized by a vertical torsional nystagmus with the vertical component downwards and the upper pole of the eye beating to the affected and non-affected ear, respectively [3–5]. Indeed, differential diagnosis between these two variants remains a challenge [6]. Likewise, vertical nystagmus in the bow and lean tests [7] was
previously described in patients with posterior canal BPPV [8]. Otoliths located in the ampullary arm of the PC could move induced by gravity toward or away from the ampulla as the head is bent forward or extended in these tests.

A recent study proposed four possible locations of the otoconia in patients with posterior canal BPPV, closer or farther from the ampulla [8]. We presented a case, in which the otoconia could be located in an atypical position in the posterior semicircular canal far from the ampulla in the transition of the ampullary and non-ampullary arm. Presumably, this is the first report in which a patient with posterior canal BPPV demonstrated a vertical ‘pseudospontaneous’ nystagmus.

**Case report**

A 34-year old men presented with 2-days history of quick episodes of vertigo caused by changes in the position of the head, mainly when turning over in the supine position. He had no previous trauma, comitant neurotological disorders, hearing or vestibular complaints. Oculomotor Examination using a Video Frenzel goggle (Interacoustics, Middelfart, Denmark) revealed a mild spontaneous downbeating nystagmus only with visual fixation removed. He showed no gaze evoked nystagmus and his saccade and smooth pursuit testing was normal. The head impulse test was not performed at this time due to the clinical suspicion of BPPV. The bow test increased robustly the intensity of the downbeating nystagmus. The lean test, after a latency of 8s, revealed an upbeating torsional nystagmus directed the upper pole of the eye to the right ear. After that, the Dix Hallpike test to the right increased the intensity of the upbeating torsional nystagmus directed the upper pole of the eye to the right ear. Then, the Epley maneuver was performed at this time given the clinical hypothesis of BPPV. The second position of this maneuver (with the head turned 45° to the left), the upbeating torsional nystagmus maintained the same direction. In the third position of this maneuver (with the head turned 135° to the left side) he demonstrated a downbeating nystagmus. He achieved resolution of vertigo, spontaneous and positional nystagmus as measured 30 min after the repositioning maneuver.

On the seventh day, he returned to the clinic without symptoms or nystagmus. Video head impulse test was performed with normal gain responses of all 6 semicircular canals without refixation saccades. Brain MRI also was performed and no intracranial lesion was found.

**Discussion**

We reported here a case of a right posterior canal BPPV presented with a spontaneous downbeating nystagmus. This is a clinical sign usually seen in central lesions such as cerebellar disorder. In this case, we could consider it as a vertical ‘pseudospontaneous’ nystagmus. Horizontal ‘pseudospontaneous’ nystagmus was described previously in patients with lateral canal BPPV and it occurs due to a 30° inclination of this canal relative to the horizontal plane that allows the otoconia to move by the gravitational force [9]. Its differential diagnosis from spontaneous nystagmus is performed with the bow and lean test, in which the patient bows the head forward and leans the head backward in the sitting position [7,9]. The horizontal ‘pseudospontaneous’ nystagmus increases with the lean test and inverts its direction with the bow test [9], while a spontaneous nystagmus, (e.g. due to a vestibular neuritis), does not change its direction with head position changes in relative to gravity.

In the same way, in our case, we could call it a vertical ‘pseudospontaneous’ nystagmus as it is due to a biomechanical etiology induced by the action of gravity. In this patient, the otoconia could be located in an atypical position in the posterior semicircular canal far from the ampulla in the transition of the ampullary and non-ampullary arm (Figure 1). This inclination relative to the horizontal plane allows the otoconia to slowly float in the direction of the ampulla provoking an ampullopetal inhibitory flow and resulting in a mild nystagmus with a vertical downbeating component. In the bow test, the otoliths will also move in the direction of the ampulla, provoking an ampullopetal inhibitory flow and resulting in a nystagmus with a vertical downbeating component (Figure 1). On the other hand, otoconia will fall away from the ampulla in the lean and Dix Hallpike tests, provoking an ampullofugal excitatory endolymphatic flow, resulting in a nystagmus with a vertical upbeating component (Figure 1). Indeed, vertical nystagmus in the bow and lean tests was previously described in patients with posterior canal BPPV [8].

In our clinical practice, we have performed the strategy of the minimum stimulus [10] for diagnosis of all types of BPPV [11]. First, we check whether there is ‘pseudospontaneous’ nystagmus. Then we perform the bow and lean test. If there is a horizontal nystagmus, we may proceed with the Seated Supine
Positioning Test succeeded by the McClure-Pagnini test, given the clinical suspicion of lateral canal BPPV. On the other hand, if the patient did not demonstrate any nystagmus or if a vertical nystagmus is observed, we may continue with the Dix Hallpike test. In that way, this case report reinforces the relevance of the strategy of the minimum stimulus for diagnosis of BPPV.

The mild ‘pseudospontaneous’ downbeating nystagmus could only be identified with visual fixation removed. Therefore, it is essential to suppress visual fixation when examining patients with vestibular complaints. This also reinforces the peripheral etiology of this nystagmus.

Moreover, we have previously exposed the importance of examining the pattern of the nystagmus induced by each step of the repositioning maneuver in patients with BPPV [11,12]. According to this hypothesis, we may assume where the debris are located, clarify its paths in the direction of the utricle and confirm the diagnosis. In the second position of the Epley maneuver, the nystagmus maintained the same direction as the otolith were still flowing away from the ampulla. In the third position, the downbeating nystagmus observed was previously called liberatory nystagmus and it means that the otoliths are entering in the common crus in the way back to the utricle [13]. In this case, the pattern of the nystagmus observed in the second and third position of the Epley maneuver confirmed the diagnosis of right posterior canal BPPV. Besides, the patient achieved resolution of symptoms, spontaneous and positional nystagmus after the repositioning maneuver.

We may consider a few differential diagnosis in cases of spontaneous or positional downbeating nystagmus. Downbeating nystagmus that occurs spontaneously or following a canalith repositioning maneuver was previously related to a complete or incomplete canalith jam [6,14–17]. A complete canalith jam may be developed by an otoconial mass that blocks a possible stenosis or a narrow portion of the canal, resulting in a blockage of the endolymphatic flow and a persistent ampullopetal displacement of the posterior canal cupula. Consequently, a spontaneous downbeating nystagmus regardless of head position may be demonstrated. In cases of incomplete canalith jam the otoliths may be partially entrapped in the non-ampullary arm of the posterior canal close to the common crus, decreasing the endolymphatic flow in this canal. Thus, the cupula may respond to a low frequency stimulus, provoked by the otoconia displacement but it may not be activated to a high frequency stimulus.

**Figure 1.** Possible movement of the otoliths based on an atypical location in the right semicircular posterior canal in the upright position; bow, lean and Dix Hallpike tests. C: cupula; G: gravity.
such as the head impulse test [16,17]. Therefore, the video head impulse test could be a useful tool for detecting canal involvement in BPPV, mainly in patients presented with downbeating nystagmus [15,16]. Canalith jam in the posterior canal should be considered in cases of spontaneous or positional downbeating nystagmus, presented with an isolated posterior canal hypofunction recovered after a successful repositioning maneuver [14]. In our case we did not proposed a canalith jam as the main clinical hypothesis due to the fact that the nystagmus changed its direction during the bow and lean test. Besides, the patient achieved resolution of nystagmus after the repositioning maneuver.

Another diagnosis to be considered in cases of spontaneous downbeating nystagmus is the inferior vestibular neuritis. In this case, an isolated posterior canal hypofunction in the video head impulse test and an asymmetric cervical vestibular-evoked myogenic potential could elucidate the diagnosis [18,19]. However, in patients with inferior vestibular neuritis the downbeating nystagmus does not change its direction by head position changes in relative to gravity and the posterior canal hypofunction does not recover after the repositioning maneuver.

**Informed consent**

This is a case report conducted in accordance with the Declaration of Helsinki and an informed consent was obtained from the patient.

**Disclosure statement**

RC is a paid speaker of Grunenthal, Abbott and UCB Pharmaceutical. He received free devices for testing from Natus and Interacoustic. BR has received speaker’s honoria from Natus. The remaining authors have no conflict of interests to declare.

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