Case Report/Case Series

Persistent Spontaneous Nystagmus Following a Canalith Repositioning Procedure in Horizontal Semicircular Canal Benign Paroxysmal Positional Vertigo

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IMPORTANCE Nystagmus can occur spontaneously from multiple causes. Direction-changing positional nystagmus on the supine roll test is a characteristic clinical feature in horizontal semicircular canal benign paroxysmal positional vertigo. One of several mechanisms of spontaneous nystagmus is plugging of the otoconia, which has been described as a canalith jam.

OBSERVATIONS We evaluated a 52-year-old woman with a history of geotropic variant of horizontal semicircular canal benign paroxysmal positional vertigo on the right side who had been treated with a modified Lempert maneuver 3 months earlier. The patient had persistent spontaneous nystagmus, despite a positional change after the canalith repositioning procedure. A bithermal caloric test result demonstrated unilateral canal paresis on the right side. The following day, the patient's symptoms and nystagmus had subsided. On a repeated bithermal caloric test, a normal response was demonstrated on both sides.

CONCLUSIONS AND RELEVANCE To our knowledge, this is the first report of a case that shows on video persistent nystagmus findings consistent with a canalith jam. We discuss a possible mechanism underlying this phenomenon.

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Benign paroxysmal positional vertigo (BPPV) refers to a vestibular peripheral disease that features sudden episodes of short-lasting rotatory vertigo triggered by changes in the head position with respect to the gravitational vector. Although a small percentage of patients experience a persistent form, BPPV has often been described as a self-limiting disease.[1] The pathogenesis underlying the persistent form of BPPV is thought to differ from that generally believed to explain the canalithiasis[2] and cupulolithiasis[3,4] forms of BPPV.

Intractable cases of BPPV may be caused by a jamming of the otoconia within a canal or between the cupula and the adjacent ampulla wall. The canalith jam may create partial or complete obstruction within the canal, resulting in spontaneous nystagmus that persists irrespective of a change in head position.[5]

Herein, we describe a patient with horizontal semicircular canal BPPV (HSC-BPPV). The patient had persistent spontaneous nystagmus, despite a positional change even after a canalith repositioning procedure.

Report of a Case

The study was approved by the institutional review board of Myongji Hospital, Gyeonggi-Do, Korea. Written informed consent was obtained from the patient described herein. A 52-year-old woman visited our clinic as an outpatient, reporting an abrupt onset of dizziness and vomiting when getting out of bed. The patient had a history of geotropic variant of HSC-BPPV on the right side 3 months prior to the visit and had been treated with a modification of the maneuver described by Lempert and Tiel-Wilck.[6] The symptoms and positional nystagmus had resolved the day after receiving the therapeutic maneuver. A pure-tone audiogram, bithermal caloric test, and brain diffusion magnetic resonance imaging performed at the time of the previous episode had not shown any abnormal findings.

At the present visit, the Dix-Hallpike test and supine roll test induced geotropic direction-changing horizontal nystagmus, which was more severe on the left side. The severity of subjective dizziness was commensurate with the amplitude of nystagmus. Head-bending nystagmus (the head is bent 30° forward in the neutral sitting position) and lying-down nystagmus (the patient lies supine with the head flexed 30° forward) were not observed.[7] Spontaneous and gaze-evoked nystagmus were also not present. Considering the history of HSC-BPPV on the right side, a repeated right supine roll test was performed, which revealed right horizontal geotropic nystagmus with an amplitude similar to that seen during the left supine roll test. On a repeated head-bending nystagmus test,

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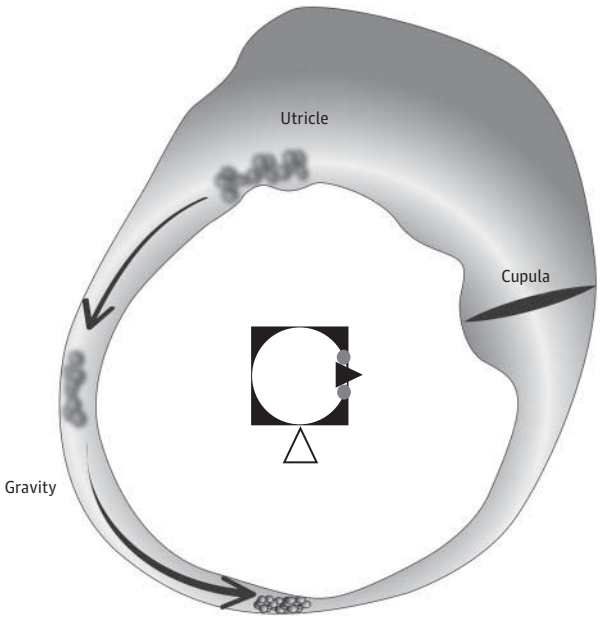
subtle right horizontal nystagmus was demonstrated (**Video 1**). However, a diagnosis of geotropic variant of HSC-BPPV on the left side was considered primarily because the patient reported more severe vertigo when lying on the left side and because of the greater amplitude of geotropic horizontal nystagmus seen during the left supine roll test at the initial positional testing. Accordingly, a modified Lempert maneuver had been performed on the left side for initial treatment.

At the next follow-up visit, the patient reported worsening of symptoms, despite the therapeutic maneuver. She had been lying primarily on the right side at home because the dizziness was alleviated in that position. A neurootologic examination revealed nonfatiguing spontaneous horizontal nystagmus beating toward the right side that persisted in all positions. Head shaking did not exacerbate the intensity of the right-beating nystagmus, and the head thrust test was negative (**Video 2**). The bithermal caloric test response demonstrated unilateral canal paresis on the right side. Results of brain magnetic resonance imaging performed for the differential diagnosis of a central pathologic condition were normal. The patient refused to be admitted for further evaluation and treatment and returned home for personal reasons.

Subsequently, the patient visited the emergency department at a local hospital because her symptoms had not been adequately controlled. She was treated with intravenous hydration only, which seemed to improve her symptoms. The following day, the patient returned to our clinic. Her symptoms had subsided, and no nystagmus was seen using Frenzel video goggles. On a repeated bithermal caloric test, a normal response was demonstrated on both sides.

Figure 1. Mechanism of Plugging of the Right Horizontal Semicircular Canal With the Patient Lying on the Right Side



The movement of the otoconia forms a plug (long curved arrows), resulting in complete impaction in the narrowest part of the canal, which may have been aggravated by the patient's position after physiotherapy. The white arrowhead indicates the affected ear, and the black arrowhead indicates the patient's nose.

Discussion

Spontaneous nystagmus can sometimes occur in HSC-BPPV, although direction-changing positional nystagmus on the supine roll test is a pathognomic clinical feature.[6] Generally, 2 mechanisms explain spontaneous nystagmus in HSC-BPPV. First, the HSC is anatomically tilted 30° upward relative to the horizontal plane in the neutral position. This natural inclination of the HSC can produce a slow falling of the otoconia in the utriculofugal direction caused by gravity. In this case, spontaneous nystagmus should easily be changed by forward or backward movement of the head.[8,9] Second, plugging of the endolymph with the otoconia may evoke permanent utriculofugal deflection of the cupula in the HSC, resulting in nonfatiguing spontaneous horizontal nystagmus. Under this condition, nystagmus may persist irrespective of the head position.[10]

We suspect that the nonfatiguing spontaneous nystagmus seen in this patient may have occurred because of a canalith jam, a condition in which the otoconia become plugged in the narrowest portion of the canal lumen. They form a closed space between the cupula and the plugged portion, which causes persistent cupular deviation that leads to directional fixed and nonfatiguing nystagmus without positional effects. The size of a single otoconia is 10 μm, meaning that it cannot fill the endolymphatic space of the semicircular canal, the inner diameter of which is 600 μm.[11,12] Nevertheless, according to a study by House and Honrubia,[12] if approximately 62 otoconia with dimensions of 10 μm form an agglomeration, it is possible to cause the canalolithiasis type of BPPV. Therefore, an agglomeration consisting of more than 62 otoconia, larger than the inner dimension of the semicircular canal, could theoretically occlude the narrowest portion of the semicircular canal. Other otoconial pathologic conditions may cause spontaneous nystagmus. According to a recent study,[13] the otoconia lodged at the basal portion of the crista can maintain the cupula in a deflected position regardless of the direction of gravity, causing a direction-fixed nystagmus similar to the circumstance resulting from a canalith jam. Cupulolithiasis is a more frequently encountered condition that causes prolonged nystagmus, but it results from the attachment of the otoconia to the cupula, which deflects the cupula according to the direction of gravity, producing direction-changing and fatiguing nystagmus.[8,9]

We had initially speculated that the diagnosis of the patient described herein was a geotropic variant of HSC-BPPV on the right side because she had been treated for that diagnosis 3 months earlier and because a relapse had occurred on the right side as posterior semicircular canal BPPV after the initial attack. However, the positional test elicited more vigorous nystagmus and symptoms on the left side, so the diagnosis was changed to geotropic variant of HSC-BPPV on the left side based on the second law by Ewald.[14] When the patient's right-sided HSC-BPPV had been misdiagnosed and treated with a modified Lempert maneuver on the left side, this procedure may have caused the migration of the otoconia toward a

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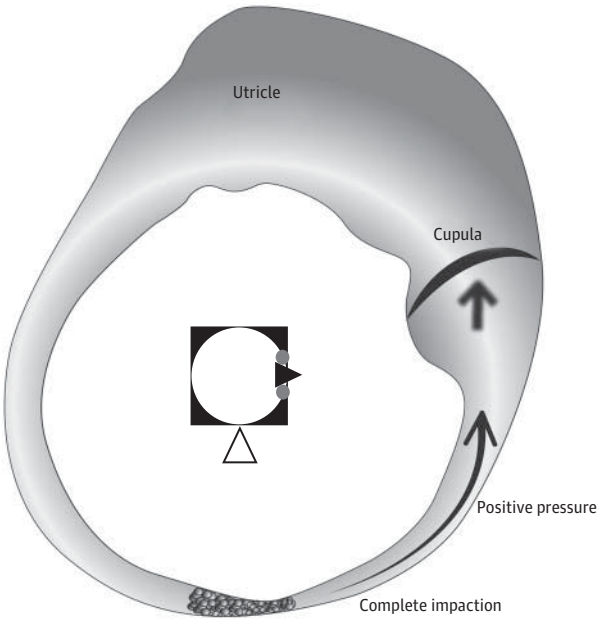
Figure 2. Possible Mechanism of Persistent Spontaneous Nystagmus With a Canalith Jam



Complete impaction may result in a positive endolymph pressure (long curved arrow) that could induce prolonged utriculopetal deviation of the cupula (short straight arrow). The white arrowhead indicates the affected ear, and the black arrowhead indicates the patient's nose.

more dependent position corresponding to the anterior arm of the right HSC. Furthermore, maintaining a right-sided lying position may have accentuated the migration of the otoconia. The additional movement of the otoconia, which remained in the utricle or the posterior arm of the HSC, could have formed an otoconial plug that induced complete impaction in the narrowest portion of the HSC (**Figure 1**). This could have induced a positive endolymph pressure to cause prolonged utriculopetal deviation of the cupula, leading to continuous excitatory stimuli (**Figure 2**). Based on these mechanisms, we believe that the continuous nystagmus and symptoms seen in our patient may have been the result of this continuous excitatory stimulus.

No specific lesion was demonstrated on brain magnetic resonance imaging, so a central pathologic condition is unlikely to be the underlying cause in this case. Acute vestibulopathy was excluded because the results of the head-shaking test and the head thrust test were negative. Also, the patient's symptoms and the bithermal caloric test response had normalized without any special treatment after only one day. The improvement of symptoms by simple hydration in the emergency department may have been due to spontaneous remission during the time spent there and not due to the hydration therapy.[15]

Conclusions

To our knowledge, no other case of nystagmus consistent with a canalith jam observed using Frenzel video goggles has been reported in the literature. Considering the clinical course of the patient described herein, misdiagnosis of HSC-BPPV can lead to the application of an improper maneuver in which the head is rotated toward the affected side starting from the healthy side, occasionally causing a canalith jam. The persistent spontaneous nystagmus seen in all positions may be indirect proof of the existence of a positive endolymphatic pressure caused by an improper maneuver. Therefore, physicians should pay special attention when localizing the site of HSC-BPPV. In addition, when a canalith jam is suspected, they should perform detailed examinations that are relevant to the differential diagnosis.

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Study concept and design: All authors.
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