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

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Benign paroxysmal positional vertigo (BPPV) refers to a vestibular peripheral disease that features sudden episodic 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. The pathogenesis underlying the persistent form of BPPV is thought to differ from that generally believed to explain the canalithiasis and cupulolithiasis 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.

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 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.

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.
Nystagmus Following Canalith Repositioning

Case Report/Case Series Research

Spontaneous nystagmus can sometimes occur in HSC-BPPV, although direction-changing positional nystagmus on the supine roll test is a pathognomonic 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 endolympathic 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

Discussion

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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.

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 different diagnosis.

Conclusions

Continuous vertigo and spontaneous nystagmus 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.

ARTICLE INFORMATION

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