The Laryngoscope

Case Report

Persistent Positional Nystagmus: A Case of Superior Semicircular Canal Benign Paroxysmal Positional Vertigo?

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Involvement of the superior semicircular canal (SSC) in benign paroxysmal positional vertigo (BPPV) is rare. SSC BPPV is distinguished from the more common posterior semicircular canal (PSC) variant by the pattern of nystagmus triggered by the Dix-Hallpike position: down-beating torsional nystagmus in SSC BPPV versus up-beating torsional nystagmus in PSC BPPV. SSC BPPV may be readily treated at the bedside, which is a key component in excluding central causes of down-beating nystagmus. We present an unusual video case report believed to represent refractory SSC BPPV based on the pattern of nystagmus and the absence of any other central signs.

Key Words: Benign paroxysmal positional vertigo, semicircular canal, superior, anterior, nystagmus, cupulolithiasis.

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INTRODUCTION

Benign paroxysmal positional vertigo (BPPV) is the most common peripheral vestibular disorder. Patients complain of brief episodes of vertigo triggered by rapid motion and changes in head position. The pathologic basis for BPPV is the presence of otoconial debris from the utricular macula within a semicircular canal, where it may either float freely in the endolymph of the membranous canal (canalithiasis) or adhere to the cupula rendering it sensitive to gravity (cupulolithiasis). The posterior semicircular canal (PSC) is reported to account for 81% to 89% of cases, the horizontal canal in 8% to 17% of cases, and the superior canal in only 1% to 3% of cases.[1] Successful treatment often depends on correct identification of the involved canal, and is based on the pattern of nystagmus observed during provocative test maneuvers.

Additional Supporting Information may be found in the online version of this article.

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In this report, we present a video of unusual positional eye movements presumed to be due to the rare occurrence of cupulolithiasis involving the left superior semicircular canal (SSC). To our knowledge, it is one of the few videos of SSC BPPV in existence. A video of typical left PSC canalithiasis is also provided for comparison.

CASE REPORT

A 47-year-old woman presented in August 2010 complaining of daily episodes of vertigo triggered by reclining back in bed. The vertigo was particularly severe when lying on her left side, and resolved by rolling to her right side. It did not respond to Brandt-Daroff exercises. She denied any hearing loss, tinnitus, headache, neck pain, or focal neurologic symptoms. She reported a prior history of positional vertigo 6 years earlier that was successfully treated with a particle repositioning maneuver (PRM). Past medical, surgical, and family history were otherwise unremarkable. She did not take any medications with known central nervous system (CNS) or oculomotor side effects.

Examination in our clinic showed a healthy appearing woman. Speech was fluent without dysarthria. There was no spontaneous or gaze-evoked nystagmus in room light. Her tympanic membrane was intact, translucent, and mobile bilaterally. Examination of cranial nerves III to XII was normal. The ocular motor assessment revealed intact smooth pursuit and normal saccade velocity and accuracy. There was no dysdiadochokinesia, dysmetria, or tremors. Gait was steady and not ataxic.

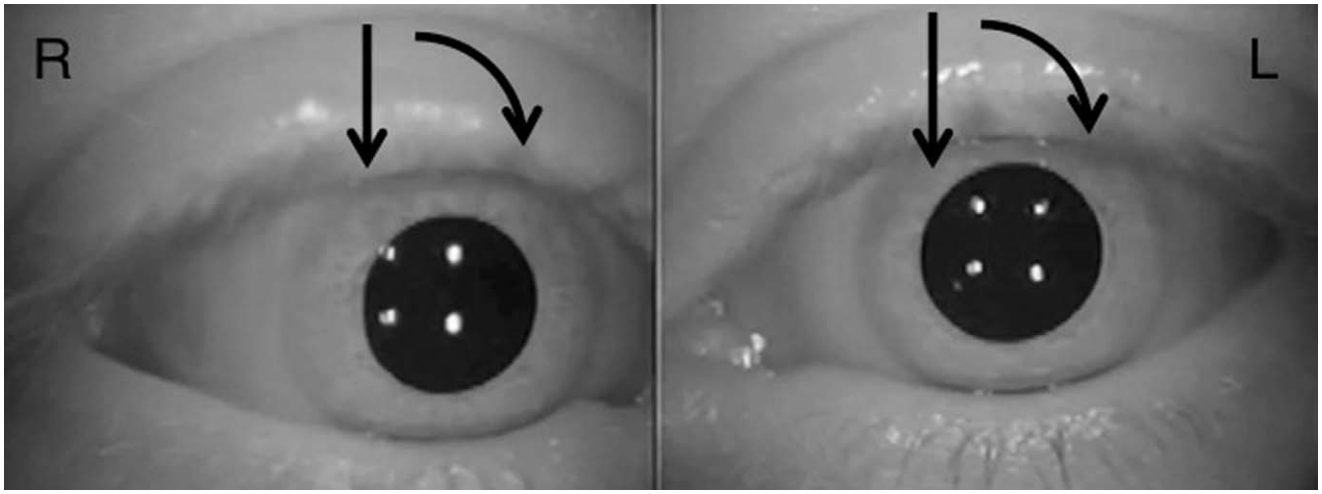

Fig. 1. Nystagmus associated with left superior semicircular canal BPPV as seen under video-oculography goggles. The video where the nystagmus can be viewed is available online. Placing our patient in the left Dix-Hallpike position caused a persistent down-beating torsional nystagmus, with the torsional component beating toward the dependent ear. The nystagmus was of immediate onset and persisted as long as the patient remained in the provocative position. This is consistent with cupulolithiasis. Note that the left eye has more of a vertical component and the right eye has a more of a torsional component. R = right; L = left.

Dix-Hallpike and positional testing were conducted using video-oculography goggles to suppress visual fixation. In the sitting position, there was no nystagmus in primary, horizontal, or vertical gaze. Left Dix-Hallpike testing was positive for immediate onset of down-beating torsional nystagmus, with the torsional component beating toward the dependent ear. The torsional component was more prominent in the right eye and the vertical component more prominent in the left eye (Fig. 1 and online Video 1). It persisted as long as the patient remained in the provocative position, and she was persistently symptomatic. The right Dix-Hallpike test triggered this same pattern of nystagmus, although it was less in velocity and duration. An audiogram showed normal hearing acuity bilaterally. Magnetic resonance imaging (MRI) of the brain and internal auditory canals was negative for retrocochlear, cerebellar, or craniocervical junction pathology.

BPPV of the left SSC was suspected. Cupulolithiasis was the most likely mechanism because the nystagmus did not exhibit latency to onset or fatigue on repeated positional tests. A series of directed liberatory and PRMs were performed with and without mastoid vibration over 2 months, without success. Additional Brandt-Daroff exercises were prescribed, but the patient reported limited compliance due to nausea during the exercises. Central pathology was unlikely given her clinical history, lack of clinical signs, or imaging features to suggest CNS pathology, a prior history of BPPV responding to particle repositioning, and the observed pattern of nystagmus. She was offered surgical plugging of the left SSC, but declined. She continues to experience positional

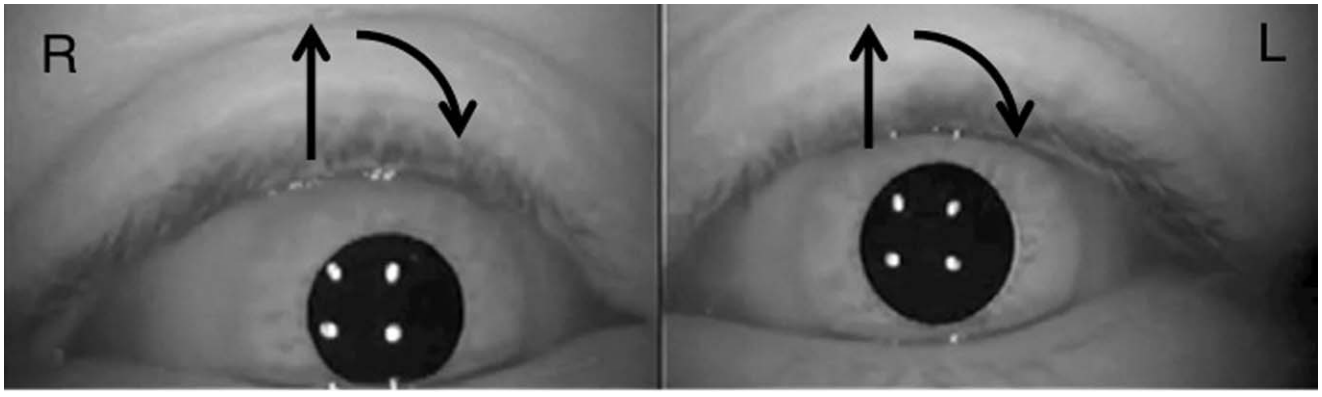

Fig. 2. Example of typical nystagmus seen with left posterior semicircular canal BPPV, canalithiasis variant, under video-oculography goggles. The video where the nystagmus can be viewed is available online. The key characteristic that differentiates posterior from superior canal BPPV is the direction of the vertical component of the nystagmus. With involvement of the posterior canal, the vertical component is up-beating. This video depicts left posterior canal BPPV in a different patient, which manifests as up-beating torsional nystagmus during Dix-Hallpike testing. The nystagmus started after a brief delay, and has a crescendo-decrescendo pattern of intensity, which is consistent with canalithiasis. Also note that the left eye has more of a torsional component and the right eye has more of a vertical component. R = right; L = left.

vertigo with lying on her left side, nearly 6 months after presenting to our clinic.

DISCUSSION

SSC BPPV is rare because the anatomic orientation of the canal does not favor retention of otoconial debris. The posterior arm of the canal descends directly into the common crus, facilitating clearance of any debris back to the vestibule. Because there are so few published reports of SSC BPPV, it is less defined than the posterior and horizontal canal variants.

Controversy exists regarding the best test to diagnose SSC BPPV; ipsilateral, contralateral, and bilateral Dix-Hallpike testing have all been reported to trigger the nystagmus.[2–4] There is greater consensus regarding the pattern of nystagmus seen with SSC BPPV, which should align in the plane of the affected canal in accordance with Ewald's first law. Ampullofugal deflection of the cupula from either cupulolithiasis or canalithiasis would excite superior canal afferents, causing a down-beating torsional nystagmus. With SSC BPPV, the torsional component is usually more pronounced in the contralateral eye and the vertical component in the ipsilateral eye.[5] Nystagmus associated with PSC BPPV has key differences: the vertical component is up-beating and more pronounced in the contralateral eye, whereas the torsional component is more prominent in the ipsilateral eye (Fig. 2 and online Video 2).

Because the nystagmus associated with SSC BPPV has a strong down-beating component, care must be taken to exclude CNS pathology particularly when the patient's nystagmus is refractory to particle repositioning therapy, as was the case for this patient. The differential diagnosis for positional down-beating nystagmus includes Arnold-Chiari malformation, cerebellar degeneration, multiple system atrophy, multiple sclerosis, and medication side effects. The largest series published on positional down-beating nystagmus found that the majority of patients with unequivocal CNS pathology exhibited a purely down-beating nystagmus (i.e., without a torsional component) and had coexisting gait, speech, or autonomic dysfunction.[6] Because our patient did not have any other central signs and no structural abnormality on MRI, the distinct characteristics of the observed nystagmus suggested SSC BPPV as the most likely diagnosis.

It is unclear why our patient's BPPV did not respond to liberatory maneuvers and PRMs. However, this is not particularly surprising because BPPV secondary to cupulolithiasis is often less responsive to conservative treatment measures than canalithiasis.[7] This is presumably because otoconial debris must be freed from the cupula before repositioning maneuvers can be effective. We suspect that this patient has otoconial debris firmly adherent to the cupula, which were not mobilized despite extensive efforts in vestibular rehabilitation therapy. Surgical plugging of the SSC for BPPV refractory to medical therapy is not well described, but there is one published success using a transmastoid approach to treat persistent SSC canalithiasis.[8] Canal plugging in this case could have provided definitive evidence to support the clinical diagnosis.

CONCLUSION

SSC BPPV is exceedingly rare. It can be easily confused with the more common PSC counterpart because patients present with similar symptoms and both forms are diagnosed with the Dix-Hallpike test. Careful attention to the precise pattern of nystagmus elicited by the Dix-Hallpike test should allow the clinician to distinguish between these two variants. Because the nystagmus associated with SSC BPPV has a strong vertical component, care must be taken to exclude central pathology.

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