CASE REPORT

Conversion from geotropic to apogeotropic direction changing positional nystagmus resulting in heavy cupula positional vertigo: case report

Antonia Elisa Lagos a,*, Phoebe Helena Ramos a, Karina Aracena-Carmona b, c, Iván Novoa c

a Pontificia Universidad Católica de Chile, Otolaryngology Department, Santiago, Chile
b Pontificia Universidad Católica de Chile, Otolaryngology Department, Otolaryngology Laboratory Technologist, Santiago, Chile
b Pontificia Universidad Católica de Chile, Faculty of Medicine, Health Sciences Department, Speech Therapy Career, Santiago, Chile

Received 27 September 2020; accepted 29 October 2020
Available online 29 November 2020

Introduction

Benign paroxysmal positional vertigo (BPPV) is the most common peripheral vestibular disorder, characterized by transient vertigo episodes triggered by head position changes.1 The most accepted theory for its pathophysiology is the detachment of otoconia from the utricular otolith macula into the semicircular canals (SCC). The horizontal SCC (HSCC) accounts for 5%–30% of all BPPV cases.1, 2 HSCC-BPPV can be confirmed with the supine roll test (SRT), where a paroxysmic positional nystagmus beating in the plane of the affected canal is evoked, with latency, fatigability, and generally lasting less than 1 min.3 When free otoliths or debris are suspended in the HSCC (canalithiasis), a geotropic or undermost ear direction changing positional nystagmus (DCPN) is observed. Conversely, when these particles are adhered to the cupula (cupulolithiasis), an apogeotropic (uppermost ear) DCPN is observed, with stronger intensity when the head is turned away from the affected ear in the SRT, with brief or no latency, and lasting more than 1 min.4,5

HSCC-BPPV is often a limited disease, nevertheless, there are cases of atypical HSCC-BPPV like heavy and light cupula, that present with common characteristics: persistent DCPN without latency or fatigability, symptoms lasting more than 1 min, and the presence of a null plane.6 The null plane is defined as the position where the nystagmus disappears when the head is rotated during the SRT, and can help identify the affected side in cupulopathy.7 These clinical findings can be explained by a different pathogenesis than for canalithiasis or cupulolithiasis. In heavy cupula this structure has an increased density compared to the endolymph, producing an ampullofugal deflection, with a persistent apogeotropic DCPN.8 In normal conditions the SCC are gravity independent, since the cupula and the endolymph’s gravity is the same, considering they have the same density.8 However, when the cupula’s density becomes lighter or heavier in comparison to the endolymph, its deflection renders it sensitive to gravity.8 Some authors describe heavy cupula to be caused by lighter surrounding endolymph, however, the change in the cupula’s density is the current accepted

* Corresponding author.
E-mail: aelagos@uc.cl (A.E. Lagos).

Peer Review under the responsibility of Associação Brasileira de Otorrinolaringologia e Cirurgia Cérvico-Facial.

https://doi.org/10.1016/j.bjorl.2020.10.016
1808-8694/© 2020 Associação Brasileira de Otorrinolaringologia e Cirurgia Cérvico-Facial. Published by Elsevier Editora Ltda. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
mechanism, where otolith debris transforms it into a gravity sensitive organ. Light and heavy cupula are challenging diagnosis, and their identification can be difficult in the clinical practice. Most of these patients have high spontaneous remission with no established treatment. This report aims to describe the case of a patient presenting with severe HSCC apogeotropic persistent DCPN.

Case description

A 47-year-old premenopausal female patient, with no comorbidities, attends the otolaryngology outpatient consult on July 27th 2019, due to episodic vertigo, triggered by head position changes, during the past week. Vertigo crises were specifically induced after standing from the bed, lasting seconds to minutes, with residual dizziness between crises. Vertigo crises also presented with nausea and vomiting, but no audiologic or neurologic symptoms. The patient recalls a similar clinical presentation two years earlier, however, in that opportunity symptoms were self-limited, and she did not undergo further study.

During the physical examination there was no spontaneous nystagmus, a geotropic fatigable horizontal DCPN was found during the SRT, and a horizontal fatigable nystagmus to the left was found during the Dix-Hallpike maneuver to...
Three days later, a videoystagmography (VNG) showed a spontaneous horizontal nystagmus beating to the right (with reduced intensity during eye fixation). Oculomotor tests were normal. Positional tests with VNG showed an apogeotropic DCPN during the SRT, without latency and more than 4 min duration, of greater intensity when the head turned to the right (Fig. 1A). The Bow and Lean Test showed a horizontal right-beating nystagmus (bow), and then left-beating nystagmus (lean) (Fig. 1B). The caloric test showed a symmetric response. With these results, a HSCC-BPPV was diagnosed, possibly from the left HSCC. Consequently, different repositioning maneuvers were performed in 6 occasions in a 23 day period (first Zuma e Maia; second Gufoni and Zuma e Maia; third Zuma e Maia; finally Zuma e Maia with vibration in 3 different days), yet no improvement was recorded.

Given the failure with repositioning maneuvers and the persistent positional nystagmus, a null plane was directly sought, identifying it 20° to the left (Fig. 2). This confirmed an atypical HSCC-BPPV: left heavy cupula. An MRI was indicated, ruling out central lesions, and serum 25-hydroxy-vitamin-D was measured, noting a severe deficiency (5.6 ng/mL). A cholecalciferol megadose was prescribed (50,000 UI weekly, during 6-weeks), and subsequently the Zuma e Maia maneuver was performed on 2 occasions immediately after a repeated head-shaking maneuver. Two
and three months after the diagnosis, a slight decrease in the nystagmus was observed (Fig. 3A and B). 25-hydroxyvitamin-D was measured 2-months after the megadose, reaching 66.3 ng/mL. The patient persisted with occasional slight dizziness, accentuated during head position changes, but being able to undergo normal life activities. She was evaluated with a VNG 4 months after the diagnosis, identifying only a mild horizontal apogeotropic DCPN (Fig. 3C). Seven months later, the positional nystagmus completely resolved (Fig. 3D). The patient gave written informed consent, and this manuscript was reviewed and approved by the scientific ethics committee of Pontificia Universidad Católica de Chile (ID: 200630014).

**Discussion**

We present a patient with initial geotropic fatigable DCPN, that later changed to apogeotropic persistent DCPN with a null plane at 20° to the left, and initial spontaneous nystagmus beating to the right. The transition from geotropic fatigable DCPN to apogeotropic persistent DCPN could be explained by heavy debris initially in the HSCC, increasing the endolymph density, that later either (1) attaches to the cupula making it heavier, or (2) is followed by an endolymph homeostatic overcompensation, producing a relative increase in the cupula’s density. The presence of a spontaneous nystagmus can be seen in both canalithiasis and cupulolithiasis, defined as pseudospontaneous nystagmus.

This can be observed in an upright head position where a 30° angle inclination between the HSCC and the horizontal gravity plane places the ampulla at a higher position than the rest of the canal. This causes deflection of the ampulla away from the utricle in heavy cupula cases, resulting in nystagmus beating towards the healthy ear. Differential diagnosis should be made with cupulolithiasis in the presence of apogeotropic nystagmus, and canalithiasis in the presence of geotropic nystagmus (Tables 1 and 2). Central lesions must be ruled out in patients presenting with cupulopathy, given the persistent character of the DCPN, which can also occur in central lesions.

The torpid evolution with no or little response to repositioning maneuvers during 2–3 months, and complete resolution after 7 months is noteworthy. BPPV is usually treated with particle repositioning maneuvers, nevertheless, in atypical BPPV these maneuvers may not be as efficient, hence vestibular suppressants should be prescribed. Most studies state there are no effective treatment options. Some authors advocate repositioning maneuvers, although spontaneous remission within first weeks is frequent, while more severe cases have been described to last 2 months. On the other hand, Tang et al. report 16 patients with heavy cupula diagnosis, treated with the Barbecue maneuver, or Gufoi and then Barbecue maneuvers. Treatment failure was seen in 3 patients at 7 days and only 1 patient after 30 days. However, these patients may have presented spontaneous resolution.

**Table 1** Differential diagnosis findings during examination.

| Diagnosis         | Supine roll test | Bow and lean test                  |
|-------------------|------------------|-------------------------------------|
|                   | Direction        | Intensity | Latency | Duration | Null plane | Flexion | Extension |
| Canalithiasis     | Geotropic        | Greater intensity on the affected side | Without latency | Less than 1 min | No | Towards the affected side | Towards the healthy side |
| Light cupula      | Geotropic        | Greater intensity on the affected side | With latency | More than 1 min | Yes | Towards the affected side | Towards the healthy side |
| Cupulolithiasis   | Apogeotropic     | Greater intensity on the healthy side | Without latency | Less than 1 min | No | Towards the healthy side | Towards the affected side |
| Heavy Cupula      | Apogeotropic     | Greater intensity on the healthy side | Without latency | More than 1 min | Yes | Towards the healthy side | Towards the affected side |

**Table 2** Differential diagnosis according to direction of nystagmus.

| Differential diagnosis | Apogeotropic persistent DCPN | Geotropic persistent DCPN |
|------------------------|------------------------------|---------------------------|
| Pseudo nystagmus       | Cupulolithiasis              | Canalithiasis             |
|                        | Heavy cupula                | Light cupula              |
|                        | Nystagmus beats towards the healthy side | Nystagmus beats towards the affected side |
The accurate localization of the affected side is fundamental for improved treatment outcomes. In heavy cupula, this can be identified by the nystagmus’ intensity, the Bow and Lean test, and the null plane position\(^\text{8}\) (Table 1). The presence of null points in patients with persistent DCPN may indicate a gravity sensitive cupulopathy,\(^\text{6}\) and the position varies greatly, with a standard deviation of 22.4° in heavy cupula.\(^\text{4}\) In the present case, multiple repeated repositioning maneuvers failed to resolve the vertigo and nystagmus, and 2 months after the diagnosis, only a reduction in the nystagmus had been achieved. After this, no more repositioning maneuvers were performed, and the patient evolved positively with a reduction in symptoms intensity.

Additionally, the patient’s serum 25-hydroxy-vitamin-D levels were normalized as symptoms decreased. Many studies have tried to establish the relation between vitamin D deficiency and BPPV. To date, an association has been demonstrated between vitamin D deficiency and more severe symptoms, a longer duration, a lower success rate of repositioning maneuvers and a higher recurrence rate.\(^\text{5}\) This could explain the treatment failure in the present case. On the other hand, studies have proven that patients with canalithiasis have lower 25-hydroxy-vitamin-D levels than patients with cupulolithiasis.\(^\text{10}\) In the present cupulopathy case, the patient had a severe vitamin D deficiency. More studies are required to clearly establish the difference between vitamin D levels in canalithiasis and cupulolithiasis, and to determine the possible pathophysiology for this finding. It is unclear whether this patient’s symptoms resolved due to natural history, and/or the cholecalciferol megadose.

Conclusions

BPPV is a frequent cause of vertigo, yet its atypical forms may present a defying diagnosis and treatment. When faced with an atypical BPPV with failure to treatment, a null plane must be sought, and central lesions must be ruled out. The affected side should be accurately localized in order to improve the treatment outcomes. This patient presented a challenging treatment and her symptoms resolved after 7 months.

Funding

This study did not require funding.

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Wang H, Yao Q, Li Z, Yu D, Shi H. Characteristics of positional nystagmus in patients with horizontal canal canalolithiasis or cupulopathy. J Neurol. 2019;266:2475–80.
2. Tang X, Huang Q, Chen L, Liu P, Feng T, Ou Y, et al. Clinical findings in patients with persistent positional nystagmus: the designation of ‘heavy and light cupula’. Front Neurol. 2019;10:326.
3. von Brevern M, Berholon P, Brandt T, Fife T, Imai T, Nuti D, et al. Benign paroxysmal positional vertigo: diagnostic criteria. J Vestib Res. 2015;25:105–17.
4. Ichijo H. Neutral position of persistent direction-changing positional nystagmus. Eur Arch Otorhinolaryngol. 2015;273:311–6.
5. Kim CH, Pham NC. Density difference between perilymph and endolymph: a new hypothesis for light cupula phenomenon. Med Hypotheses. 2019;123:55–9.
6. Kim C, Shin JE, Kim YW. A new method for evaluating lateral semicircular canal cupulopathy. Laryngoscope. 2015;125:1921–5.
7. Ichijo H. Caloric testing in patients with heavy or light cupula of the lateral semicircular canal. Laryngoscope Invest Otolaryngol. 2016;1:163–8.
8. Shin JE, Jeong K, Ahn SH, Kim C. Conversion between geotropic and apogeotropic persistent direction-changing positional nystagmus. Acta Otolaryngol. 2015;135:1238–44.
9. Yetiser S. Review of the pathology underlying benign paroxysmal positional vertigo. J Int Med Res. 2019, 300060519892370.
10. Nakada T, Sugiuara S, Uchida Y, Suzuki H, Teranishi M, Sone M. Difference in serum levels of vitamin D between canalolithiasis and cupulolithiasis of the horizontal semicircular canal in benign paroxysmal positional vertigo. Front Neurol. 2019;10:176.