Recurrent “light cupula” occurring alternately on both sides

Chang-Hee Kim¹, Jung Eun Shin¹
Department of Otorhinolaryngology-Head and Neck Surgery, Konkuk University Medical Center, Konkuk University School of Medicine

Correspondence to: Chang-Hee Kim, MD, PhD
Department of Otorhinolaryngology-Head and Neck Surgery
Konkuk University Medical Center, Konkuk University School of Medicine
120-1 Neungdong-ro (Hwayang-dong), Gwangjin-gu
Seoul, Republic of Korea, 143-729
Telephone: +82-2-2030-7666
Fax: +82-2-2030-5299
Email: ryomachang@naver.com

Jung Eun Shin’s email address: gracejshin@hanmail.net
Abstract

Background: The “light cupula” is a condition wherein the cupula of the semicircular canal has a lower specific gravity than its surrounding endolymph. It is characterized by a persistent geotropic direction-changing positional nystagmus in the supine head-roll test, and the identification of a null plane with slight head-turning to either side.

Case presentation: This study describes a case of recurring light cupula that occurred alternately on both sides. At the first episode, a null plane was identified on the right side, and the nystagmus was stronger in the left head-roll than in the right, which led to the diagnosis of a light cupula on the left side. At the second episode, the nystagmus was more intense in the right head-roll than the left, and a null plane was identified on the left side, leading to the diagnosis of a light cupula on the right side.

Conclusion: This is the first case report of recurring light cupula alternately involving both sides. Although the pathophysiology is not entirely understood yet, the light cupula should be considered as one of causes of recurrent positional vertigo.

Keywords: Direction-changing positional nystagmus, Light cupula, Head-roll test, Positional vertigo
Background

The condition of “light cupula”, characterized by a cupula with a lower specific gravity than its surrounding endolymph, has been introduced as an emerging concept accounting for positional vertigo [1-4]. In cases where the light cupula involves the horizontal semicircular canal (hSCC), a persistent geotropic direction-changing positional nystagmus (DCPN) without latency or fatigability is typically observed in the supine head-roll test. A null plane, in which the nystagmus disappears, can be identified when the patient’s head is slightly turned to the right or left side while the patient is in the supine position. In this report, we will describe a case of recurrent light cupula occurring alternately on both sides with an interval of 2 months.

Case presentation

A previously healthy 38-year-old man who suddenly developed positional vertigo was referred to our hospital. He did not complain of audiological symptoms or headaches, and reported no previous history of vertigo or neurological disorders including migraine. The otoscopic examination revealed a normal tympanic membrane. Further examination yielded a very weak left-beating spontaneous nystagmus when the patient was in the sitting position, and the head impulse test revealed no catch-up saccade. Additionally, there were no focal neurological deficits found. Examinations of pure tone audiometry revealed normal hearing on both sides. The patient’s eye movement was examined at various head positions and was recorded using a goggle equipped with an infrared camera (SLMED, Seoul, Korea), thus facilitating the “light cupula” diagnosis. A right-beating nystagmus was observed when the patient bowed his head at 90°. While in the supine position, a horizontal nystagmus beating toward the left side was persistently observed, and the maximal slow-phase velocity (SPV) was 33°/s (Fig 1A, Supp. Video 1). When the patient’s head was turned to the right (Fig. 1B, Supp. Video 2) or left (Fig. 1C, Supp. Video 3) at 90° in the supine position, a geotropic DCPN continued persistently as long as the position was maintained. The intensity of the nystagmus was stronger with head-roll to the left (maximal SPV = 53°/s) than to the right (maximal SPV = 10°/s). A null plane, at which the nystagmus ceases and the direction of nystagmus changes, was identified when the patient turned his head slightly to the right (25~30°) while in the supine position, leading to a diagnosis of a light cupula on the left side. The diagnostic criteria for the light cupula are the presence of a persistent geotropic DCPN on the supine head-roll test and the identification of a null plane [1]. Magnetic resonance imaging of the brain revealed no abnormal finding.

The patient was prescribed vestibular suppressants for symptomatic relief, and the positional vertigo and nystagmus disappeared within 1 week.
Two months later, the patient revisited our clinic with complaints of severe positional vertigo akin to the previously experienced episode of vertigo. Thorough neurological examinations revealed no focal neurologic signs, and the patient’s hearing was normal on both sides. A weak spontaneous nystagmus beating toward the right side was observed when the patient was in the sitting position, and a weak left-beating nystagmus was observed when the patient bowed his head at 90°. In the supine position, a right-beating nystagmus was continuously observed, and the maximal SPV was 19°/s (Fig 2A, Supp. Video 4). When the patient’s head was turned to the right (Fig. 2B, Supp. Video 5) or left (Fig. 2C, Supp. Video 6) at 90° in the supine position, a persistent geotropic DCPN was observed. The intensity of the nystagmus was stronger with a head-roll to the right (maximal SPV = 38°/s) than to the left (maximal SPV = 10°/s). A null plane was identified when the patient turned his head slightly to the left (25~30°) in the supine position, which led to the diagnosis of a light cupula on the right side.

Discussion

Our patient suffered from recurring positional vertigo, which is, as far as we know, the first report of recurring light cupula alternately involving both sides. There are two points that may be addressed from this observation: (1) the orientation of the hSCC cupular axis with regard to the gravitational vector, (2) the pathophysiology of the light cupula.

The pathophysiology of light cupula is still unclear. The attachment of light debris to the cupula has been suggested as a cause of light cupula [2], but the light debris has not been identified yet. Others have proposed that the increase in the specific gravity of the endolymph may contribute to light cupula [1, 5], which was further supported by recent findings suggesting that light cupula can be accompanied by sudden sensorineural hearing loss ipsilaterally [6], and that the condition of light cupula may involve all 3 SCCs on the same side [7].

In previous studies, the cupular axis of hSCC has been described as running medial to lateral in direction, and the angle between the sagittal plane and the cupular axis was variable, ranging from 11° to 58° [1, 2, 5, 8]. In this condition, the side of the null plane corresponds to the side of light cupula, and is suggested to be the most important finding for the determination of the affected side [1]. In contrast, other sources of literature have suggested that the cupular axis of the hSCC is approximately parallel to the anterior SCC [3, 9], indicating that a null plane may be identified when the patient’s head is slightly rolled to the contralesional side when the patient is in the supine position, as observed in our patient. At the first episode of vertigo, which was caused by a light cupula in the left hSCC, a null plane was identified on the right side, and the nystagmus was stronger in the left
head-roll than in the right. A right-beating nystagmus was observed in the bowing position, and a left-beating nystagmus was observed when the patient was placed in the supine position. At the second episode, persistent geotropic DCPN was more intense in the right head-roll than in the left, and a null plane was identified on the left side.

Because the incidence of light cupula in patients showing geotropic DCPN was as high as 14% [1], meticulous investigation of the duration and latency of positional nystagmus during supine head-roll test is essential for differential diagnosis between light cupula and hSCC canalolithiasis. Though it has been reported that the affected side of light cupula can be determined as the same side with a null plane, a null plane may be found on the opposite side of the light cupula as in the present case.

**Conclusion**

This is the first case report of recurring light cupula alternately involving both sides. Although the pathophysiology of the light cupula is still not clear and the treatment modality for this disease needs further investigation, the light cupula should be ruled out in patients with recurrent positional vertigo particularly when the patient shows geotropic DCPN.

**Consent**

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

**Abbreviations**

DCPN: Direction-changing positional nystagmus, hSCC: Horizontal semicircular canal, SPV: slow-phase velocity.

**Competing interests**

The authors declare that we have no competing interests or financial disclosures.

**Author’s contributions**
CHK interpreted the case, and drafted and submitted the final manuscript. JES made substantial contribution in interpreting the case, and drafted the part of the discussion. All authors have revised the manuscript critically and approved the final manuscript.

Acknowledgements

This study was supported by Basic Science Research Program through the National Research Foundation of Korea (NRF) funded by the Ministry of Education, Science and Technology (2012R1A1A2044883).
References


Fig. 1 The instance of light cupula observed in the left LSCC. The patient’s head and left LSCC were viewed from the top of the patient’s head. (A) In the supine position, a left-beating nystagmus (maximal SPV = 33°/s) was persistently observed due to an utriculopetal deflection of the cupula. (B) When the patient’s head was turned to the right while in the supine position, the left LSCC was inhibited because of an utriculofugal deflection of the cupula resulting in a persistent right-beating nystagmus (maximal SPV = 10°/s). (C) The cupula of the left LSCC was deflected utriculopetally when the head was turned to the left side, which caused a persistent left-beating nystagmus (maximal SPV = 53°/s). Note that the intensity of the nystagmus is stronger in left-head rolling than in right head-rolling. LSCC, lateral semicircular canal; SPV, slow-phase velocity.
Fig. 2 The incidence of light cupula observed in the right LSCC. The patient’s head and the magnified right LSCC were viewed from the top of the patient’s head. (A) In the supine position, a left-beating nystagmus (maximal SPV = 19°/s) was persistently observed due to an utriculopetal deflection of the cupula. (B) When the patient’s head was turned to the right side, the right LSCC was activated because of the utriculopetal deflection of the cupula resulting in a persistent right-beating nystagmus (maximal SPV = 38°/s). (C) When the head was turned to the left side, the cupula of the right LSCC was deflected utriculofugally, which caused a persistent left-beating nystagmus (maximal SPV = 9°/s). Note that the intensity of the nystagmus is stronger in right-head rolling than in left head-rolling. LSCC, lateral semicircular canal; SPV, slow-phase velocity.
Figure 1

A. Supine  
B. Right head-roll  
C. Left head-roll
Figure 2

A  Supine  

B  Right head-roll

C  Left head-roll

deg

sec

deg

sec

deg

sec
Additional files provided with this submission:

Additional file 1: Supplemental video 1.avi, 804K
http://www.biomedcentral.com/imedia/1381693072144629/supp1.avi
Additional file 2: Supplemental video 2.avi, 896K
http://www.biomedcentral.com/imedia/7853283661446298/supp2.avi
Additional file 3: Supplemental video 3.avi, 930K
http://www.biomedcentral.com/imedia/7611212931446298/supp3.avi
Additional file 4: Supplemental video 4.avi, 828K
http://www.biomedcentral.com/imedia/1115955004144629/supp4.avi
Additional file 5: Supplemental video 5.avi, 776K
http://www.biomedcentral.com/imedia/5550472101446298/supp5.avi
Additional file 6: Supplemental video 6.avi, 638K
http://www.biomedcentral.com/imedia/1450432810144629/supp6.avi