Clinical Characteristics of Posterior and Lateral Semicircular Canal Dehiscence

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Abstract

Keywords

- ▶ posterior semicircular canal dehiscence syndrome
- ▶ lateral semicircular canal dehiscence syndrome
- ▶ superior semicircular canal dehiscence syndrome
- ▶ vertigo
- ▶ autophony

The objective of this study was to evaluate the characteristic symptoms of and treatments for lateral semicircular canal dehiscence (LSCD) and posterior semicircular canal dehiscence (PSCD) and its proposed mechanism. A dehiscence acquired in any of the semicircular canals may evoke various auditory symptoms (autophony and inner ear conductive hearing loss) or vestibular symptoms (vertigo, the Tullio phenomenon, and Hennebert sign) by creating a “third mobile window” in the bone that enables aberrant communication between the inner ear and nearby structures. A PubMed search was performed using the keywords *lateral*, *posterior*, and *semicircular canal dehiscence* to identify all relevant cases. Our data suggest that PSCD, although clinically rare, is most likely associated with a high-riding jugular bulb and fibrous dysplasia. Patients may experience auditory manifestations that range from mild conductive to extensive sensorineural hearing loss. LSCD is usually associated with chronic otitis media with cholesteatoma.

Introduction

Semicircular canal dehiscence involving the superior canal was first described in 1998, and the less commonly identified lateral and posterior semicircular canal dehiscences were described soon thereafter.¹ A dehiscence acquired in any of the semicircular canals may evoke various auditory or vestibular symptoms by creating a “third mobile window” in the bone that enables aberrant communication between the inner ear and nearby structures.^{1–5} Superior semicircular canal dehiscence syndrome (SSCDS), posterior semicircular canal dehiscence syndrome, and lateral semicircular canal dehiscence syndrome may display similar symptoms but can be rooted in different causes.¹ Any of these bony dehiscences can result in the formation of a “third mobile window” into the inner ear and ensuing vestibular and/or auditory mani-

festations.^{6–8} In a series analysis of 700 temporal bone high-resolution multislice computed tomography (CT) examinations, Stimmer et al found semicircular canal dehiscence in 9.6% of temporal bones with 8% affecting the superior canal, 1.2% affecting the posterior canal, and 0.4% affecting the lateral canal.⁹

Possible auditory symptoms of canal dehiscence include autophony, aural fullness, hearing loss, and pulsatile tinnitus.⁶ Vestibular dysfunction including the Tullio phenomenon and chronic disequilibrium can also occur.^{1,6} However, incidental findings may occur in cases of asymptomatic patients following the exposure of the temporal bone during surgery^{10,11} or cross-sectional imaging of the temporal bone.¹² High-resolution CT scans of temporal bones prove valuable for diagnosis,^{1,13} and the axis of nystagmus helps indicate the

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dehiscent semicircular canal.^{13,14} Surgery aimed at resurfacing, plugging, and capping the bony erosion have each proven useful in alleviating symptoms in patients with vestibular and auditory manifestations (→Fig. 1).^{1,5}

Posterior semicircular canal dehiscence (PSCD), which is clinically rare,⁹ is associated with a high-riding jugular bulb^{1,15} and fibrous dysplasia.^{1,16} Lateral semicircular canal dehiscence (LSCD) is usually associated with chronic otitis media with cholesteatoma; ideal treatment strategies for managing LSCD secondary to cholesteatoma are debated due to hearing loss concerns.¹ Although some remain in favor of fully removing the cholesteatoma matrix, others maintain that preserving an undamaged thin layer of the matrix may prevent deafness that could otherwise result from labyrinthine exposure.¹

Chen et al proposed that because the superior and posterior semicircular canals are in closer contact to the overlying dura or dural venous sinuses of the middle and posterior fossas, they are more prone to growing dehiscent than the lateral canal that lacks direct contact with these structures.^{12,17} Appropriate ossification or thickening of the trilaminar bony layers may be precluded by the pressure and fluid changes of the dura and sinuses.^{12,17}

Fig. 1 The arrow demonstrates a middle fossa craniotomy exposing the semicircular canals. Once the dura is carefully retracted off the skull base, the dehiscence can then be repaired. However, with this surgical approach, a dehiscence in the portion of the temporal bone overlying the posterior canal may be overlooked because this site remains unexposed.

Mechanism

A semicircular canal dehiscence affects pressure transmission in the labyrinth.³ The bony dehiscence makes it possible for the canal to respond to sound and pressure stimuli through the creation of what has been dubbed a “third mobile window” into the inner ear.^{3,7} Heightened sound-induced umbo velocity within patients with SSCDS suggests decreased cochlear impedance.¹⁸ Studies conducted on a chinchilla model of the syndrome directly support this mechanism.^{3,18} Rosowski et al reported that the measured sound-induced velocity of the endolymph within the affected canals of chinchilla models of canal dehiscence revealed sound flow through the dehiscence, and cochlear potential measurements demonstrated heightened response to bone conducted sound.^{3,18} Furthermore, Hirvonen et al observed that after fenestration of the bone covering the semicircular canal, all afferents innervating the canal developed pressure sensitivity, although fewer than half of the other vestibular nerve afferents were rendered sensitive.¹⁹ These effects were consequently terminated once the fenestra was firmly sealed.¹⁹

For patients with posterior semicircular canal dehiscence, various stimuli that cause an inward or outward motion of the stapes footplate may trigger vestibular dysfunction including loud noises, positive and negative pressure in the external acoustic meatus, and the Valsalva maneuver or Müller maneuver.^{3,19,20} These stimuli also cause vestibular dysfunction in both lateral semicircular canal dehiscence and superior semicircular canal dehiscence. The evoked eye movements in these situations typically result in nystagmus whose velocity vectors are closely aligned with the anatomical axes of the affected canal.^{14,21}

Posterior Semicircular Canal Dehiscence

PSCD has been associated with a high-riding jugular bulb^{1,15} and fibrous dysplasia.^{3,16} Although the precise incidence of PSCD remains unknown,²² one temporal bone radiographic survey of patients with a high-riding jugular bulb found 4 of 112 cases to demonstrate PSCD.^{22,23} In the largest temporal bone multislice CT scan study to date, Erdogan et al examined the scans of 410 adult patients in a training hospital with symptoms unrelated to the inner ear, such as otitis media, cholesteatoma, and facial nerve palsy. The authors found five males to have PSCD (prevalence of 1.2%), three of whom demonstrated bilateral dehiscence.¹²

Patients with canal dehiscence have been found to present with both vestibular and auditory complaints,^{1,6} the latter of which ranges from mild conductive to extensive sensorineural hearing loss.⁶ The variation in possible auditory manifestations remains unexplained despite theories of the third-window mechanism regarding hearing loss.⁶ Gopen et al noted that a labyrinthine dehiscence should be suspected if bone-conduction responses are at -5 dBHL or better.⁶ In addition, vestibular-evoked myogenic potential (VEMP) testing commonly elicits a lowered threshold at an increased amplitude in patients with PSCD, a significantly different result from the VEMP testing responses in cases of ossicular

disease that are instead absent or present with elevated thresholds.⁶

Through an analysis of 507 temporal bone CT scans, Krombach et al found PSCD more commonly in patients with a history of vertigo than in their control group.^{10,20} However, further studies are required to verify this suggested causal link between PSCD and vertigo.¹⁰

Temporal bone CT scans, performed with the standard axial and coronal views in conjunction with VEMP responses, are diagnostically useful.⁶ It is crucial to identify the locations of any existing defects in all the semicircular canals with imaging prior to surgery.¹⁰ A dehiscence in the portion of the temporal bone overlying the posterior canal may be overlooked during SSCD surgery because this location remains unexposed during a middle fossa craniotomy (►Fig. 1).¹⁰ Surgical modifications would need to be made considering that the posterior semicircular canal faces the posterior fossa.¹⁰

With only a handful of case reports described in the literature for PSCD,^{6,23–28} Gopen et al (2010) described one of the first large series of symptomatic patients, including 12 adults and children, with PSCD.^{1,6} The patients were aged 2 to 67 years, with one bilateral case. In 70% of cases in which the dehiscence was not introduced iatrogenically, PSCD was found in association with a high-riding jugular bulb. VEMP testing results showed elevated amplitude and lowered threshold. In contrast with SSCD, this series showed a tendency for PSCD to exist in the right rather than left ear, a trend potentially explained by the higher proportion of patients with right dominant jugular venous drainage. In addition, evidence of SSCD in two of these patients along with PSCD may suggest an underlying bone development abnormality

that could help explain general canal dehiscence. One patient showed bilateral SSCD and right PSCD; another patient demonstrated SSCD in one ear and PSCD in the other.¹

Lateral Semicircular Canal Dehiscence

Idiopathic LSCD has rarely been reported.²⁹ The condition more commonly presents as an acquired lesion from cholesteatoma^{1,29} and canal wall down mastoidectomy.²⁹ Various factors have been ascribed to the bony defect including pressure from the jugular bulb, superior petrosal sinus, or brain on naturally thin bone that may erode the bone.³⁰ Krombach et al reported only two patients with LSCD, and both dehiscences stemmed from corrosion due to cholesteatoma.¹⁰ For proper diagnosis of LSCD, axial and coronal images in the planes parallel and perpendicular to of the lateral canal should be thoroughly examined (►Fig. 2).^{13,31} SSCD and LSCD may share similar symptoms of vertigo and autophony.³¹

Fistula formation may follow the erosion of the dense otic capsule bone surrounding labyrinthine structures,¹ and the most often form in the lateral canal in cases of chronic otitis media.^{1,32} Chronic otitis media with cholesteatoma and chronic granulomatous otitis media without cholesteatoma have been found to cause labyrinthine fistula formation.^{1,33} A fistula formed secondary to a cholesteatoma can be determined with a perilymph fistula test where a pressure applied to the external auditory canal induces an ipsilateral nystagmus, whereas the release of the pressure induces a contralateral nystagmus.³⁴ However, contention exists over the comparative benefits of removing the cholesteatoma matrix completely or leaving a thin layer to prevent possible deafness

Fig. 2 (A) Defect evident in coronal image reformed through the lateral semicircular canal plane [perpendicular]. (B) Intact bony cover shown in axial image reformed through the lateral semicircular canal plane [parallel]. (C) Coronal and (D) Axial: Intact bony cover displayed in images of the left lateral semicircular canal (reprinted with permission from reference 13).

resulting from labyrinthine exposure.¹ Postoperative hearing preservation rates in studies that removed the cholesteatoma matrix and studies that left it intact were found to be 84% and 83%, respectively.^{1,35} In a later study, Chen et al treated lateral canal fistulas with occlusion in 22 patients after fully removing the cholesteatoma; two of these patients underwent 5 to 15 dB hearing loss postoperatively, whereas the other 20 experienced stable or enhanced hearing.³⁶

Conclusion

A relatively recently discovered syndrome of the inner ear, canal dehiscence may induce vestibular symptoms such as vertigo, the Tullio phenomenon, and Hennebert sign, and auditory issues such as autophony and inner ear conductive hearing loss. Surgical resurfacing, plugging, or capping of the dehiscence through either the transmastoid approach or a craniotomy has been reportedly successful in alleviating these symptoms. CT, Valsalva maneuvers, VEMP, and particular types of auditory testing have proven useful in the confirmation and further diagnosis of a suspected dehiscence.

LSCD and PSCD are clinically rarer forms of canal dehiscence than SSCD, but the precise incidences remain unknown. PSCD has been associated with a high-riding jugular bulb and fibrous dysplasia and LSCD with cholesteatoma and canal wall down mastoidectomy.

Although current treatments tend to have good outcomes and may restore patient quality of life, true understanding of the causes and ideal management for canal dehiscence is limited by the scarcity of the literature.

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