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Posterior Semicircular Canal Dehiscence Following Endolymphatic Sac Surgery

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Abstract

Posterior semicircular canal dehiscence is a rare otologic entity which presents with third window signs and symptoms. Petrous apex cholesteatoma, fibrous dysplasia, high riding jugular bulb, and eosinophilic granuloma have been reported to be associated with posterior semicircular canal dehiscence. Here we report a case of development of posterior semicircular canal dehiscence following an endolymphatic sac surgery for the first time.

Keywords

dehiscence; endolymphatic sac surgery; posterior semicircular canal; endolymphatic shunt

Introduction

Dehiscence of the semicircular canals is a well established otologic entity which presents with variable auditory and vestibular symptoms including conductive hearing loss, autophony, chronic imbalance, and third window signs and symptoms (i.e., vertigo and torsional nystagmus induced by loud noise or pressure).¹ Although the clinical diagnoses and treatment of posterior semicircular canal (PSC) dehiscence have been well described, there is a paucity of literature on possible etiologies. Petrous apex cholesteatoma, fibrous dysplasia, high riding jugular bulb, and eosinophilic granuloma have been reported to be associated with PSC dehiscence.^{2,3} To our knowledge, this is the first reported case of PSC dehiscence development following endolymphatic sac (ELS) surgery. This study was approved by the Institutional Review Board at University of California, Irvine.

Case Report

In April 2008 a 54-year-old man presented to our clinic with a complaint of intractable vertigo and loss of hearing in left ear. He had a 5 years history of severe episodic vertigo, fluctuating hearing loss, fullness and continuous tinnitus of the left ear. Based on a diagnosis of left Ménière's disease, he had undergone endolymphatic sac shunt surgery on the left ear 2 months before presentation by an outside neurotologist. Otologic examination revealed normal-appearing external auditory canals and normal tympanic membranes. Audiometric testing revealed profound sensorineural hearing loss of the left ear and significant increase in hearing thresholds at all frequencies compared to his audiometric findings prior to surgery

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(Figure 1). The patient complained of vertigo sensation upon presentation of loud sounds to his left ear during audiometry. The Speech Detection Threshold (SDT) was 5 dB on the right side. Testing the left ear, he could not distinguish words but detected speech at 105 dB (Table 1). He reported vertigo sensation when hearing speech at that level.

Videonystagmography (VNG) including bithermal caloric testing with both cool and warm air revealed minimal vestibular response on the left side. (Cold air = -2 deg/s, Warm air = 2 deg/s). Pressure on the tragus to produce positive pressure in the ear canal caused a low velocity nystagmus with downward quick phases. Release of pressure caused a counter-clockwise nystagmus. A Baha device was implanted and vestibular rehabilitation therapy was started. In 2009 he presented with aural fullness, hyperacusis and new vertigo attack exacerbations precipitated by sharp loud noise, pressure on left ear, nose blowing, and Valsalva maneuver. Temporal bone computed tomography (CT) imaging revealed an iatrogenic posterior semicircular canal dehiscence (Figure 2). Three courses of intratympanic gentamicin were administered and vestibulo-ocular reflex therapy was continued. The patient has done very well with rehabilitation and has not required any further therapy.

Discussion

Our patient exhibits several characteristics of a third window abnormality. First, he suffered from a Tullio phenomenon despite having minimal hearing. The patient described a rotatory vertigo with sound and pressure. Second, the patient had a rotatory nystagmus on tragal pumping seen on VNG. The amplitude of this nystagmus was lower than that of a posterior canal dehiscence without a previously performed labyrinthotomy. Finally, the patient did not have any chronic disequilibrium as would be expected in a chronic labyrinthitis.

To our knowledge, this is the first reported case of development of PSC dehiscence following ELS surgery. The PSC injury is possible when performing this surgery as the ELS generally lies inferior and posterior to the PSC. Oftentimes, the posterior fossa dura has to be retracted behind the PSC to visualize the ELS. It is unclear whether the surgeon who performed the surgery inadvertently entered the PSC or if the PSC erosion was caused by the ELS-subarachnoid shunt. Regardless of etiology, the patient suffered from a loss of hearing and most vestibular function.

Although our patient had severe episodic vertigo, the caloric testing revealed minimal vestibular response at the left side. None of the semicircular canal dehiscence cases reported by Minor et al. had abnormal results of caloric tests.¹ In a temporal bone survey, Carey et al. reported one case of semicircular canal dehiscence with no ice-water caloric responses, indicating bilateral labyrinthine hypofunction. But their patient had no Tullio phenomenon or Hennebert's sign.⁴ In another retrospective study, Krombach et al. reported complete missing caloric test response only in 2 out of 44 cases of semicircular canal dehiscence.⁵ Interestingly, despite minimal caloric function and severe to profound hearing loss, the patient had signs and symptoms of PSC dehiscence. The findings on video-oculography on sound stimulation and pressure stimulation were milder compared to other patients with superior semicircular canal or PSC dehiscence. We speculate that in our patient the significantly reduced caloric function could be due to injury to the membranous labyrinth or a resultant labyrinthitis. Since the caloric testing mainly tests the lateral semicircular canal function, some function can be expected in the posterior semicircular canal and this patient continued to have Tullio phenomenon and Hennebert's sign, though to a milder degree compared to superior or posterior canal dehiscence patients we have seen with normal caloric function.

Our patient presented with a complete sensorineural hearing loss following endolymphatic sac surgery which is a well known complication of this procedure.⁶ However, the mechanism of hearing impairment is unclear. Although this can be due to inadvertent labyrinthotomy during surgery, it also could be explained by perilymphatic fistula formation and posterior semicircular canal dehiscence.

Conclusion

We report a case of iatrogenic posterior canal dehiscence following endolymphatic shunt placement. The patient demonstrated Tullio phenomenon and Hennebert's sign. Patients who develop sensorineural hearing loss following endolymphatic sac surgery with delayed onset symptoms of a third window (or perilymph fistula) should be evaluated with imaging. The posterior canal dehiscence in these patients can be treated with high dose gentamicin (if non-usable hearing present) or posterior canal occlusion.

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Figure 1.
Pure tone audiometry before and after endolymphatic sac surgery in the right and left ear.

Figure 2.
High-resolution computed tomography scan of the left temporal bone, axial projection, four consecutive sections. C = cochlea, ICA = internal carotid artery, PSC = posterior semi circular canal, PSCD = location of posterior semicircular canal dehiscence, S = the shunt in the endolymphatic duct.

Speech audiometry results before and after endolymphatic sac shunt surgery. Table will appear in only the online version of the journal.

Table 1

	Pre-operation		Post-operation	
	Right	Left	Right	Left
Speech Reception Threshold/Speech Awareness Threshold (dB)	5	35	10	105
Speech Discrimination Score	45	70	45	0
	100	84	100	0