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.

Introduction

Dehiscence of the semicircular canals is a well established otologic entity which presents with variable auditory and vestibular symptoms.

- Auditory dysfunction includes hearing loss, tinnitus, autophony, and feeling of fullness in the affected ear. The spectrum of hearing loss ranges from mild conductive hearing loss to profound sensorineural hearing loss. [1]
- Vestibular dysfunction includes chronic disequilibrium, Tullio phenomenon (sound-induced vertigo), and recurrent attacks of vertigo induced by changes in the middle ear pressure, i.e. abrupt noise, coughing or Valsalva maneuvers. [1]
- The diagnosis of canal dehiscence can be clinically challenging. Patients present with various auditory and/or vestibular symptoms, some of them may mimic other otologic entities, like otosclerosis and Ménière's disease. Patients may have a difficult time explaining their symptoms, further confounding the diagnosis. [2]
- High resolution computed tomography (HRCT) scan of temporal bone is crucial to confirm the diagnosis. However, it is not uncommon to misdiagnose dehiscence based on imaging alone because of the limitation of resolution and the fact that some dehiscence may be less than 0.1 mm.
- Although the clinical diagnoses and treatment of posterior semicircular canal dehiscence have been well described, there is a paucity of literature on possible etiologies. Developmental abnormalities, congenital defect, chronic otitis media with cholesteatoma, and high riding jugular bulb have been suggested as possible etiologies in previous reports. [2] To our knowledge, this is the first reported case of posterior semicircular canal dehiscence development following endolymphatic sac (ELS) surgery. This study was approved by the Institutional Review Board at University of California, Irvine.

Table 1. Speech audiometry results before and after endolymphatic sac surgery. Table will appear in only the online version of the journal.

<table>
<thead>
<tr>
<th></th>
<th>Pre-operation</th>
<th>Post-operation</th>
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<tbody>
<tr>
<td>Right</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>Left</td>
<td>35</td>
<td>105</td>
</tr>
<tr>
<td>Speech Reception Threshold (dB)</td>
<td>45</td>
<td>45</td>
</tr>
<tr>
<td>Level of Speech (dB)</td>
<td>70</td>
<td>45</td>
</tr>
<tr>
<td>Score (%)</td>
<td>100</td>
<td>100</td>
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</table>

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 semicircular canal, PSCD = location of posterior semicircular canal dehiscence, S = the shunt in the endolymphatic duct.

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 shunt surgery on the left ear 2 months before presentation by an outside neurootologist. 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 (Figure 1). The patient complained of vertigo sensation upon presentation. 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. 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 drift. 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).

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.

References


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