"SPONTANEOUS" PERILYMPH FISTULA: A CASE REPORT

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A surgically documented case of barotrauma-induced perilymph fistula is presented in this case report. A brief review of the literature on this entity confirms the difficulty of making a definitive preoperative diagnosis in most instances. Clinical, audiometric, radiologic, and intraoperative findings are presented, and the classic presumed mechanisms for this uncommon cause of sudden sensorineural hearing loss are discussed. The presence of intact evoked otoacoustic emissions in an ear demonstrating a severe cochlear-type loss was considered helpful in narrowing the differential diagnosis in this case, and may suggest a productive avenue for future study.

KEY WORDS — distortion-product otoacoustic emissions, perilymph fistula, sudden sensorineural hearing loss.

INTRODUCTION

Perilymph fistula is defined as a congenital or acquired otologic disorder resulting from an abnormal communication between the fluid-containing space around the membranous labyrinth and the middle ear.1 The initial symptoms are variable, but typically consist of the sudden onset of unilateral sensorineural hearing loss that may be fluctuating or progressive, tinnitus, and dizziness, often in the form of persistent dysequilibrium punctuated by episodic motion-induced vertigo. The onset of symptoms may be associated with trauma, whether iatrogenic, an external blunt or penetrating force, or barotrauma related to altitude changes or extreme exertion.2 A classification system for perilymph fistulas is presented in the Table.1

The first reported cases of perilymph fistula occurred as postoperative complications associated with the use of polyethylene strut or wire-Gelfoam prostheses in stapedectomy surgery.3 Fee4 reported 3 cases of perilymph leak from the oval window after head trauma; relief from vertigo and fluctuating sensorineural hearing loss were achieved by exploratory tympanotomy and repair of the leak. The first instances of “spontaneous” perilymph leak were cited by Stroud and Calceterr,# who postulated that increased intracranial pressure was the etiologic event that produced the fistula. In 1971, Goodhill6 proposed his classic theory of implosive (Valsalva-induced) and explosive (increased intracranial pressure) forces leading to membranous rupture and perilymph fistula, illustrated diagrammatically in Fig 1.

Initial excitement at the possibility of identifying a surgically treatable cause of sensorineural hearing loss resulted in reports of large series of middle ear explorations in the 1980s and 1990s, with fistula confirmation rates ranging from 11%7 to 89%.8 Variable results for positive and negative explorations resulted in a backlash against the procedure and its proponents. Predictably, both extremes have subsided and are now replaced by a stance of rational evaluation, wherein most ear surgeons believe that spontaneous perilymph fistula is a rare occurrence, but one that must be considered as a potentially treatable cause of hearing loss, especially when the onset is

### CLASSIFICATION OF PERILYMPH FISTULAS

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<th>Congenital</th>
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<tr>
<td>Mondini dysplasia</td>
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<td>Pendred syndrome</td>
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<td>Klippel-Feil syndrome</td>
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<tr>
<td>Without radiologic abnormalities</td>
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<tr>
<td>Microfissures at Hyrtl’s fissure and fissula ante fenestram</td>
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<tr>
<td>Acquired</td>
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<td>Iatrogenic</td>
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<td>Stapedectomy</td>
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<td>Neuro-otologic surgery</td>
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<td>Chronic ear surgery</td>
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<td>Physical injury</td>
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<td>Head trauma</td>
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<td>Temporal bone fracture</td>
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<td>Penetrating ear wound</td>
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<td>Barotrauma</td>
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<td>Erosion</td>
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<td>Cholesteatoma</td>
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<tr>
<td>Chronic tympanomastoiditis</td>
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<td>Neoplasm</td>
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Modified from Roman et al.1

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subject end with the authors encouraging the development of better diagnostic tests for the preoperative identification of perilymph fistulas.

The following report describes the clinical, diagnostic, and operative findings in a case of surgically confirmed perilymph fistula that underwent a complete restoration of hearing after repair. We discuss the specific audiometric and clinical findings that were considered helpful in establishing the diagnosis.

CASE REPORT

A 38-year-old man noted the sudden onset of hearing loss in the right ear, accompanied by a low-tone tinnitus and imbalance with sudden motion 1 week before seeking medical attention. He denied ear pain, drainage, whirling vertigo, and nausea, but had noticed significant noise sensitivity in that ear. There were no specific antecedent events, although he had begun a weight lifting program at the time of the onset of the hearing loss.

His medical history was unremarkable. There was no history of recent significant head trauma, noise exposure, or ototoxic medications. His family and social history were noncontributory. He was employed as a musician and music teacher (percussion) and complained of difficulty in judging at music contests due to the hearing loss and noise sensitivity.

On physical examination, the ear had a normal appearance by pneumatic otomicroscopy, and the fistula sign was negative. No other abnormalities were noted.

His audiogram (Fig 2) showed a severe low- and middle-frequency sensorineural hearing loss, to 80 dB from 250 to 1,000 Hz. Speech discrimination was 0%, the tympanogram was type A, and the stapedius reflex was absent on the right, but distortion-product otoacoustic emissions (DPOAEs; Fig 3) were present and robust, demonstrating intact outer hair cell function. The Stenger test was negative.

Further laboratory tests included a complete blood count with differential, the erythrocyte sedimentation rate, a blood chemistry profile, serum protein electrophoresis, a fluorescent antinuclear antibody test, and serologic tests for autoimmune inner ear disease. All were negative.

The patient could not tolerate magnetic resonance imaging of the cerebellopontine angles, either closed or open, but a computed tomography scan of the temporal bones with and without contrast was negative for acoustic neuroma or any other anatomic abnormality.

A diagnosis of sudden sensorineural hearing loss in the right ear was made, and the possibility of a...
perilymph fistula was considered because of the temporal association with weight lifting.

Initially, conservative therapy was undertaken with rest; avoidance of nicotine, caffeine, excessive salt, and loud noise; and a tapering 12-day course of prednisone starting at 60 mg/d.

At the time of follow-up, the patient reported no real improvement in his symptoms, and had continued hearing loss, noise intolerance, and tinnitus. The audiogram now showed a change to a middle- and high-frequency sensorineural hearing loss (Fig 4), again with intact DPOAEs. Because of ongoing symptoms in spite of conservative therapy and nonservice-
Fig 4. Later preoperative audiogram.

able hearing that prevented the patient from performing his work, surgical exploration was recommended.

The next day, the patient was taken to the operating room, and a right exploratory tympanotomy was performed. A pooling of clear fluid in the oval window was noted after repeated gentle aspiration and prolonged observation, and a presumed oval window fistula was repaired with postauricular areolar tissue. No other anatomic abnormalities were noted.

One month later at a follow-up visit, the patient reported a marked improvement in his hearing, and no longer noticed any tinnitus or noise intolerance. His audiogram had returned to normal (Fig 5). He was able to resume his work as a musician and music teacher.

**DISCUSSION**

In a survey of 167 otologists performed in 1990, Hughes et al. found that the feature considered most important in the history of a patient with a possible perilymph fistula was antecedent trauma or barotrauma. They also agreed that the diagnosis is based on the total clinical picture, including the history, physical examination, and diagnostic tests. In our case, the salient features that suggested the possibility of a perilymph fistula were the onset of hearing loss, tinnitus, and vertigo in association with the Valsalva maneuver (weight lifting), the fluctuating nature of the hearing loss, and the preservation of robust otoacoustic emissions in the presence of a severe cochlear-type hearing loss. Exploratory tympanotomy for repair of the suspected fistula was considered after serviceable hearing was not obtained after a trial of conservative therapy.

The anatomic basis for perilymph fistula resides in the exquisitely compartmentalized cerebrospinal fluid (CSF)–perilymph–endolymph system. Delicate intralabyrinthine and extralabyrinthine membranes separate the hydrostatically loaded fluids of the inner ear from their intimate connections to CSF pressure gradients, the intracranial vascular systems, and the rheological relationships of the tibotympanic apparatus and middle ear space. Goodhill's pioneering work regarding the explosive force of increased intracranial pressure transmitted by the cochlear aqueduct or internal auditory canal to the perilymphatic space has been borne out in several animal and human experiments. Beentjes determined that an abrupt change in CSF pressure resulted in an equal, but attenuated, change in perilymph pressure in cats, and using human subjects, Sakikawa et al. found that nose blowing resulted in a CSF pressure increase of up to 500 mm Hg during diagnostic lumbar puncture.

An interesting aspect of this case is the preservation of evoked otoacoustic emissions (OAEs) in the presence of a severe sensorineural hearing loss. Dis-
tortion-product otoacoustic emissions are an epiphenomenon of the biomechanical properties of healthy outer hair cells. Briefly, the ciliated sensory receptor cells for hearing undergo a shearing effect induced by contact with the tectorial membrane during the traveling wave’s differential displacement of the basilar membrane. This shearing process generates a receptor potential that creates lateral wall electromotility in the rod-like outer hair cells. It is hypothesized that actin- and myosin-like motor molecules perform this contractile and lengthening function, modulating basilar membrane motion in response to various sound levels. The psychoacoustic by-product of this activity is the exquisite sensitivity of the auditory thresholds, and the dynamic range, frequency selectivity, and temporal resolution of human hearing.14

The equipment used for DPOAE testing in this case was the Starkey DP2000 Distortion Product Otoacoustic Emission Measurement System. Details of the testing procedure will not be discussed, but the test result (“DP-grain”) indicated that outer hair cell damage was not present in the frequency range of the audiometrically documented hearing loss, and thus the injury sustained was not the result of outer hair cell dysfunction, but some other insult to the cochlea.

Experimentally induced sensorineural hearing loss from a variety of causes, including ototoxicity,15 ischemia,16 noise trauma,17 viral infection,18 and endolymphatic hydrops,19 results in a loss of evoked OAEs in laboratory animals. These electrophysiological data were often correlated with histologic data showing destruction of the outer hair cells in these types of cochlear insults. Experimentally induced perilymph fistulas demonstrate various results with respect to preservation of evoked OAEs, but in some reports these and other electrophysiological functions such as the endocochlear direct current potential were preserved even with the induction of a pneumolabyrinth.20-22

In human subjects, the presence of evoked OAEs has been evaluated during the early and recovery stages of idiopathic sudden sensorineural hearing loss. Several investigators have noted the presence of intact OAE responses in patients with sudden hearing loss, even when audiometric thresholds exceeded 40 dB, and observe a positive correlation between recordable OAEs and eventual recovery of hearing.23,24

The preservation of OAEs in surgically documented cases of perilymph fistula has not been studied in a systematic manner. Whether this particular test, in conjunction with other clinical, audiometric, and radiologic information, will prove reliable in discerning an actual “site of lesion” in patients with sudden hearing loss cannot be determined from a single case report. In this instance, it did seem helpful in narrowing the differential diagnosis and was supportive of intervention that led to a successful clinical outcome.

REFERENCES


