Case In Point: Perilymphatic Fistula

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By David Tetzlaff, MD [1] and Deepak M. Kamat, MD, PhD [2]

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Her parents reported that she was following them when they entered their garage to get a ball. When the girl did not appear in the garage, her parents went to look for her and found her lying next to a tree. When they attempted to get her to stand up, she complained of dizziness and then vomited. Her parents noted that her eyes were moving back and forth rapidly. She was continent, alert, and interactive during the entire episode. All extremities moved equally and there were no signs of trauma, abrasions, or bleeding.

The child remembered that she felt better lying down than sitting or standing. She recalled her parents coming for her but could not remember how she fell.

In the ED, the patient's vital signs were normal. However, the ED staff noted horizontal nystagmus and also dried blood in the child's left ear canal. CT scans of the head were reported as normal. Serum electrolyte levels and results of a complete blood cell count were within normal limits. The patient was referred to Children's Hospital of Michigan because of continued nausea and nystagmus. In our ED, the patient was alert, oriented, and interactive. We also noticed horizontal nystagmus and the blood in the left ear canal. There were no other abnormal physical findings. An otolaryngology consult was obtained: the blood in the ear canal was thought to be secondary to external trauma. An audiogram was performed; results showed moderate to severe conductive hearing loss on the left. Tympanography findings were within normal limits. Hearing in the right ear was normal.

CT scans of the temporal bone showed a left perilymphatic leak and displacement of the stapes bone from the oval window into the vestibule. The patient was taken to the operating room. Surgeons found that the tympanic membrane had sealed over a twig that protruded from the middle ear into the oval window. The twig had dislocated the stapes, which could not be found. A patch was placed over the oval window.

The patient was treated with intravenous ampicillin/sulbactam (Unasyn). Her nystagmus and nausea diminished after 2 days and she was discharged home on oral therapy with amoxicillin/clavulanate (Augmentin).

Discussion

We initially considered temporal bone fracture (because of the child's fall), seizures, CNS tumor or infection, and labyrinthitis. Seizure was unlikely, however, because the patient had no history of seizure-like activity. A mass lesion was a concern because of the nystagmus and nausea, but was ruled out after CT scans of the head proved to be normal. Infection was excluded after the child remained afebrile and had a normal white blood cell count. Labyrinthitis was unlikely because the patient had no recent history of viral illness. Trauma was highest on our differential list because of the blood in the child's left ear canal.

Vertigo and nausea after trauma may be caused by temporal bone fracture, damage to the inner ear, or perilymphatic fistula. Temporal bone fractures may be transverse or longitudinal. Transverse fractures are less common and usually manifest with vertigo, severe sensorineural hearing loss, and hemotympanum. Longitudinal fractures present with conductive hearing loss, bloody otorrhea, and loss of consciousness. A temporal bone CT is the recommended test for evaluating the inner ear. In our patient, the CT scans showed both an inner ear problem and a perilymphatic fistula, but not the foreign body.

A perilymphatic fistula may result from any congenital or traumatic disruption of the labyrinth that allows perilymph to leak into the middle ear (Table). House and colleagues estimated the incidence at less than 1 per 1000 outpatient otolaryngology visits. However, the literature contains mostly case reports and case series conducted over long periods--often involving patients with perilymphatic fistulas misdiagnosed as Ménière disease or idiopathic sudden hearing loss.

Page 1 of 2
Seltzer and McCabe found that in all age groups, 50% of perilymphatic fistulas were the result of surgery or trauma. In a series by Goto and colleagues, 49% of cases resulted from trauma or exertion and 51% from undetermined causes. Look for neoplasm or cholesteatoma if the history does not suggest a congenital, surgical, or traumatic cause.

**Diagnosis and Treatment**

The history and physical examination are the most important tools in diagnosing perilymphatic fistula--especially in cases involving trauma. Symptoms include unilateral hearing loss, vertigo, dizziness, and/or tinnitus. Goto and colleagues found hearing loss in 93%, vertigo or dizziness in 91%, and tinnitus in 76% of their cases. These symptoms can be persistent or intermittent: none is pathognomonic of perilymphatic fistula. Goto's series (like others) also found that most perilymphatic fistula leaks occurred through the oval window; this was followed by leaks through the round window. Leaks through both windows were least common.

Various studies--such as the B-2 transferring detection assay, fistula test, jugular venous compression, fluorescein injection into cerebral fluid, and positional testing--have been suggested as methods for diagnosing perilymphatic fistula. However, none is reliable enough to make a definitive diagnosis. As noted, temporal bone CT is the radiologic test of choice to evaluate the inner and medial ear structures. All affected patients should under-go audiometry to evaluate for hearing loss. Treatment of non-penetrating perilymphatic fistula is either conservative (with bed rest) or surgical. In penetrating and persistent perilymphatic fistula, treatment involves exploratory tympanotomy with a connective tissue graft and use of fibrin glue.

In one series of 36 patients, hearing improved in 12% but remained unchanged or became worse in 88% of cases. Seltzer and McCabe reported preservation or improved hearing in 73% of their patients, while Goto and colleagues reported that 20% of their patients recovered. The recovery of hearing can be difficult to predict: the chance of recovery appears to be worse in trauma-induced perilymphatic fistula. However, vertigo and dizziness resolve in most patients.

The most worrisome complication is meningitis. CT scans of the inner ear are indicated for all children with recurrent meningitis without an obvious cause to exclude inner ear malformation. If perilymphatic fistula symptoms return, recurrent leakage is most likely present; re-exploration of the middle ear may be necessary.

**References:**


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