Perilymphatic Fistula

14

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14.1 Introduction

Perilymphatic fistula is an abnormal connection between the labyrinth and structures surrounding it [1]. It is a condition that can present with a variety of non-specific symptoms; it is considered a diagnostic challenge. A proper history and physical examination is a key element during workup of those patients. Treatment options vary depending on the severity and response to medical therapy and conservative measures.

14.2 Clinical Manifestations

It has a wide range of symptoms:

1. Hearing loss [2]
   (a) It is the most common symptom.
   (b) It can show high frequency, low frequency, or even a flat sensorineural hearing loss.
   (c) It tends to fluctuate.
   (d) It can sometimes present with conductive hearing loss.
2. Vertigo [2]
   (a) It is the most common reason for seeking medical advice.
   (b) It can resemble Benign paroxysmal positional vertigo (BPPV) or Meniere's disease.
   (c) It can mimic disequilibrium in certain occasions. For example, patients may present with disequilibrium upon exposure to loud voices (Tullio's phenomenon) or by increasing Cerebrospinal fluid (CSF) pressure (during lifting or blowing of the nose).
3. Tinnitus [3]
   (a) Other symptoms of perilymphatic fistula usually accompany tinnitus.

14.3 Workup

- Diagnosis of the perilymphatic fistula is usually tricky, but it is mainly based on history, physical examination findings, and exclusion of other diagnoses.
- Some tests that can be used are as follows:
  – Audiometry: it can show high frequency, low frequency, or flat sensorineural hearing loss. A low-frequency conductive hearing

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loss can be seen in superior semicircular canal dehiscence. It is recommended to do the test in lateral decubitus while the affected ear is up [4, 5].

– Fistula test: the presence of nystagmus upon applying pressure on the external auditory canal is considered a positive fistula test. Hennebert sign is the presence of a positive fistula test without any middle ear pathology [5].
– Measuring of postural sway upon applying pressure on the external auditory canal [5].
– Electronystagmography/videonystagmography: It can show unilateral weakness. It can also be used to increase the sensitivity of a fistula test [6].
– Some other methods were utilized to help in diagnosis, like measuring Beta 2 transferrin and using fluorescein to detect perilymphatic fistula. However, their utilization has been questioned and still in doubt [7, 8].
– Exploration: it is considered unreliable for spontaneous perilymphatic fistula. However, its reliability is increased in the presence of a temporal bone fracture or stapedectomy surgery in a patient's history [3].

14.4 Management

- Conservative: Bed rest, head elevation, and avoid straining [3].
- Surgical management in case of failure to respond to conservative therapy. It consists of patching the otic capsule. Some surgeons patch the oval and round window regardless of the intraoperative findings as the detection rate of the perilymphatic fistula is quite low. Fat, perichondrium, fascia, and temporalis muscle can be used for patching [3, 9].

14.5 Etiologic Causes

Look at Table 14.1.

Table 14.1 Location and the probable etiologic causes of perilymphatic fistula

Otic capsule	Oval window	Round window
• Cholesteatoma • Temporal bone fracture • Superior semicircular canal dehiscence	• Barotrauma • Head trauma • Stapedectomy • Acoustic trauma	• Barotrauma • Mondini dysplasia

Fig. 14.1 Traumatic rupture of the tympanic membrane caused by barotrauma

14.5.1 Barotrauma (Fig. 14.1)

- Pressure changes can cause damage to the inner ear by three mechanisms:
  – Alternobaric trauma: it occurs in cases of increased pressure or asymmetrical pressure of the middle ear. It usually occurs more during the ascent of the divers or ascent of the planes. Conditions that affect the functions of the eustachian tube (URTI, sinusitis) can increase individual's susceptibility to these kinds of trauma [10].
  – Atmospheric barotrauma: it occurs in cases of the increase of the air pressure, which can cause middle ear and inner ear damage. It usually gives long-lasting symptoms as compared to the alternobaric trauma [10].

– Inner ear decompression sickness: it occurs due to the formation of bubbles in the labyrinth or its blood supply. It happens in deep water divers who use mixed gases like oxy-helium [3].

14.5.2 Acoustic Trauma

- Explosive blast produces sound more than 200 dB, which can cause tympanic membrane (TM) perforation and disruption of the ossicles. Inner ear injury can happen, especially if conductive mechanisms are not disrupted [1].

14.5.3 Trauma

- It can happen in cases of head trauma or penetrating injury with or without temporal bone fracture. However, it is rare to have a perilymphatic fistula after trauma in normal temporal bone anatomy. Look at Fig. 14.2 [10, 11].

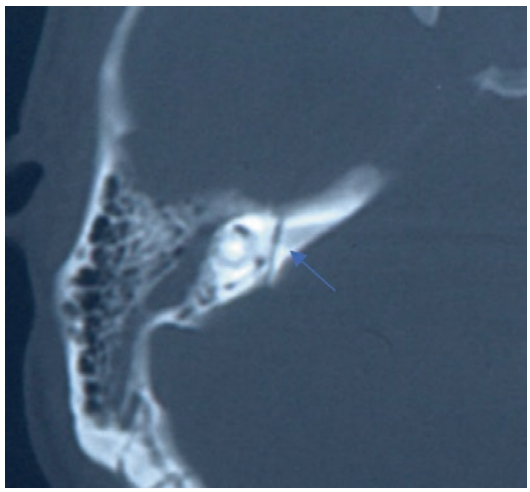

Fig. 14.2 Axial CT scan of left temporal transverse fracture (arrow) (Image Reprinted with permission from Elsevier: Elsevier books, Skull base imaging, Bert De Foer, Abdellatif Bali, Anja Bernaerts, Joost van Dinther, Erwin Offeciers, Jan W. Casselman, Jan 1, 2018)

14.5.4 Stapedectomy

- Perilymphatic fistula can happen after stapedectomy. It can be a primary or secondary one.
  – Primary: if it presents at the end of the procedure.
  – Secondary: if it manifests months or years after the procedure.
- It is more common in stapedectomy than stapedotomy.
- It is more common in revision surgeries.
- The use of gelatin sponge to seal the stapedectomy presents the highest risk of developing the fistula [10].

14.5.5 Superior Semicircular Canal Dehiscence

- Prevalence: It can be seen in 4–8% in high-resolution CT [2].
- Pathogenesis: It is often multifactorial (congenital and acquired factors) [10].
- Symptoms: Patients can have only vestibular symptoms, only auditory symptoms, or a combination of both [10, 12]. They often present with hearing loss, autophony (they can hear their voice), pulsatile tinnitus, and hearing the movement of their eyes (due to increased bone conduction). They can present with the Tullio phenomenon (vertigo and imbalance after exposure to loud voices). They might complain of vertigo and imbalance after conditions that increase Cerebrospinal fluid (CSF) pressure. Pure tone audiometry shows conductive hearing loss (CHL) with a larger air-bone (AB) gap at low frequencies [10].
- Diagnosis: High-resolution CT scan. Dehiscence can be seen in coronal view, Stenvers view (in the plane of orthogonal), or Poschl view (in the place of the SSC) [13]. CT usually overestimates the size of the dehiscence. To improve the specificity, then 0.5 mm CT scan should be reformatted to the Poschl plane [14] Fig. 14.3.

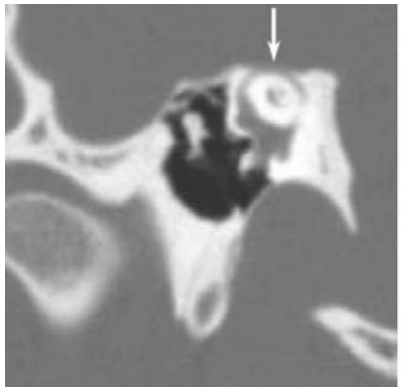
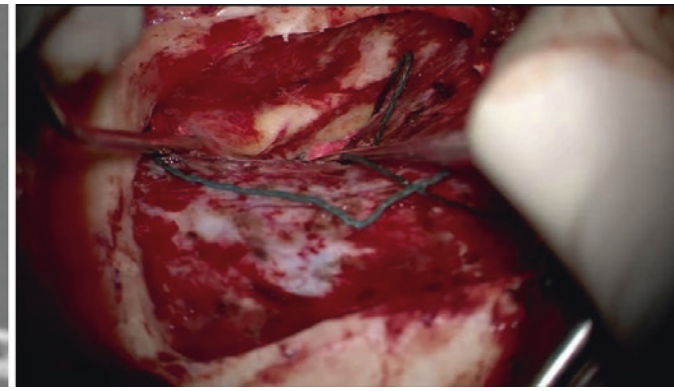

Fig. 14.3 Coronal CT scan of left IAM demonstrating left superior semicircular canal dehiscence, and next image shows SCC intraoperatively

- VEMP: it has high sensitivity and specificity (around 90%). Patients with SSC dehiscence will have a lower threshold in the affected ear for both air and bone conduction [1].
- Treatment: if patients have mild symptoms, then conservative measures should be taken. Insertion of ventilation tubes can relieve pressure symptoms. If symptoms are severe, then surgical correction should be considered [10].
- Surgical Approaches [10].
  - Middle cranial fossa craniotomy: gives a good exposure. Disadvantages are risks of craniotomy, more extended hospital stay, and temporal bone retraction.
  - Endoscopic middle cranial fossa: it has less hospital stay but still has risks of craniotomy.
  - Transmastoid: avoid craniotomy risks but do not have direct exposure to the SSC.

14.5.6 Mondini Malformation [2] (Fig. 14.4)

- It is the most common malformation of the cochlea.
- The cochlea has only 1.5 turns.
- The modiolus is hypoplastic and lacking the interscalar septum.
- Patients usually have variable auditory functions depending on the organ of Corti formation and the number of neurons.
- Now it is subtyped into three types: *Type 1*, entire modiolus and interscalar septa are lacking (it has a cystic appearance), *Type 2*, normal base turn with a cystic apex (Mondini type), and *Type 3*, where the modiolus is deficient (it has partial septation on the interscalar at the periphery of the cochlea).

14.5.7 Congenital Perilymphatic Fistula

- It is an abnormal communication between the inner ear and the middle ear. It is associated with micro-fissures around the round window or oval windows and dysplasia of labyrinthine or internal auditory meatus. It is usually suspected if a child presents with progressive or fluctuating sensorineural hearing loss (SNHL) [10, 15].

14.5.8 Perilymphatic Fistula in Children

- It can be seen in temporal bone anomalies and children who have severe or profound sensorineural hearing loss (SNHL). It can present with recurrent meningitis or Cerebrospinal fluid (CSF) behind the TM [10].

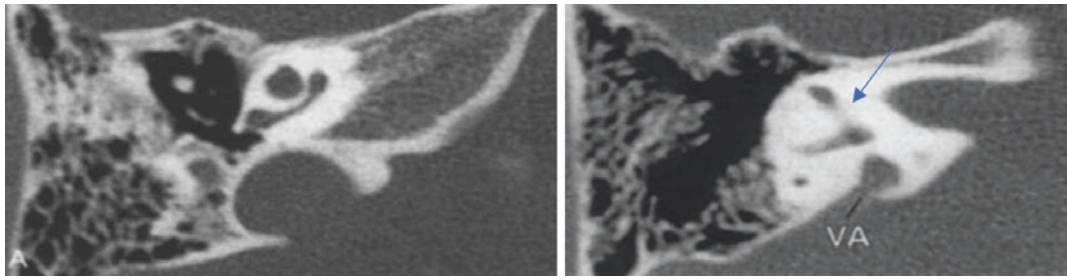

Fig. 14.4 Axial HRCT of the right temporal bone showing type II incomplete partition of the cochlea (Mondini malformation) with failure of segmentation of the middle and apical turns (arrow) associated with enlarged vestibular aqueduct (VA)

Take-Home Messages

- Perilymphatic fistula is a diagnostic challenge.
- Different images and diagnostic modalities should be used judiciously not to overwhelm the patient with unnecessary tests.
- Consider conservative management first.

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