Labyrinthine fistula secondary to cholesteatoma: a video demonstration

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DESCRIPTION

Labyrinthine fistula is a defect of the bony labyrinth of the petrous portion of the temporal bone. This can proceed to involve the membranous labyrinth. Causes include surgery, trauma, barotrauma, cholesteatoma and even noise exposure. This results in vertigo, hearing loss and tinnitus. The fistula test is a bedside clinical examination which transmits pressure through the ear canal and through the fistula to the labyrinth resulting in vertigo and nystagmus. There is a conjugate deviation of the eyes away from the stimulated side with a corrective fast phase towards that side. In a lateral semicircular canal fistula, the nystagmus (fast phase) is accordingly horizontal and towards the affected ear. This can be elicited with digital tragal pressure or a pneumatic speculum. The pressure generated transmits to the inner ear, resulting in stimulation of the vestibular-ocular reflex and therefore the eyes move away but they attempt to revert to their original position.

We present the case of a 35-year-old man who previously underwent tympanoplasty and canal wall down mastoidectomy 13 years ago. He presented to the emergency department with a 6-week history of severe right-sided otalgia and mastoid pain associated with otorrhoea, debilitating vertigo and pulsatile tinnitus.

Clinical examination revealed an unsteady gait, Romberg’s positive sign and a positive fistula. His facial nerve was intact and there were no features of meningism. Postauricular cellulitis changes were also present. His pure tone audiogram indicated mixed hearing loss. Intravenous antibiotic therapy was commenced and CT imaging was arranged which identified a large, recurrent cholesteatoma filling the right mastoidectomy cavity with a lateral semicircular canal fistula and erosion of the tegmen.

Subsequent MRI confirmed recurrent cholesteatoma (approximately 1.5 cm × 2 cm in the transverse and anteroposterior dimension, respectively) and erosion of the tegmen mastoideum and contiguous mastoid cavity demonstrating the facial canal and left semicircular canal (LSCC) fistula.

Figure 1  Non-Echo-Planar Diffusion-Weighted Imaging MRI with high signal confirming the sizeable cholesteatoma (see arrow).

Figure 2  Axial T2 weighted imaging demonstrating a large cholesteatoma abutting the right lateral semicircular canal (see arrow).

Figure 3  Intraoperative view of canal wall down mastoid cavity demonstrating the facial canal and left semicircular canal (LSCC) fistula.
tegmen tympani resulting in elevation of the dura overlying this region (figure 1). The cholesteatoma approximated the lateral genu of the lateral semicircular canal and had eroded parts of the tympanic and mastoid segments of the facial canal (figure 2).

The patient was listed for theatre 3 weeks later for a right revision mastoidectomy with obliteration. Findings included a large keratin filling cavity with loss of the posterior canal wall and bone exposure over the left semicircular canal. Once the cholesteatoma and necrotic bone were removed, the defect was reconstructed using temporalis fascia and autologous bone graft (figure 3).

Postoperatively, the patient had a gradual reduction of his dizziness which took 4 months to resolve. His ear pain and otorrhoea resolved completely with improved hearing (figures 4 and 5). He remains clinically well and undergoes regular follow-up.

The video clip demonstration of a positive fistula test (video 1) provides practitioners in both primary and secondary as well as prospective physicians and allied healthcare professionals with a useful educational tool for this sign which indicates bony erosion. In the context of a painful, discharging ear, urgent referral to an otology specialist is required.

Learning points

► Recurrent cholesteatoma can occur several years after initial surgery.
► Ear, nose and throat examination should always include otomicroscopy to ensure an adequate view and clearance of keratin for preventing progression.
► A positive fistula test is a sign of a large erosive cholesteatoma in the context of otalgia and otorrhoea and is an important clinical test to diagnose advanced disease and surgical urgency.

Contributors

As author (JM), I would like to thank the support of our co-authors (DS, RG and AA) at The Royal London Hospital for their concerted efforts towards producing this paper, both through data collection and patient record acquisition. The guidance throughout this project has been greatly appreciated.

Funding

The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests

None declared.

Patient consent for publication

Obtained.

Provenance and peer review

Not commissioned; externally peer reviewed.

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REFERENCES
