The Conundrum of Labyrinthitis Ossificans: An Etiology-Based Case Comparison and Review of Literature

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Abstract ▶

Labyrinthitis ossificans is the formation of pathological new bone within the membranous labyrinth of the inner ear due to various local and systemic pathologies. Most commonly it occurs as a sequelae of meningitis spreading to the labyrinth, from the subarachnoid space via the cochlear aqueduct and the internal auditory canal. We are comparing three different etiological presentations of labyrinthitis ossificans; namely, tympanogenic, meningitic, and traumatic, together with their management in the light of recent advances.

Keywords: Labyrinthitis, cochlear implantation, osteogenesis, labyrinthitis ossificans, case report

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Introduction

Labyrinthitis is the inflammation of the membranous labyrinth, commonly caused by infection. It can lead to the fibroblastic proliferation and ossification of the labyrinth, known as labyrinthitis ossificans (LO), resulting in irreversible profound sensory neural hearing loss (SNHL) (1). LO can be caused by tympanogenic, meningitic, or traumatic etiologies (2). Patients with labyrinthitis present with vertigo and hearing loss, which can be reversible if treated early (3). Age is a risk factor for LO. Rare causes of LO include Cogan's syndrome, otosyphilis, and meningitic LO. Comprehensive clinico-radiological analysis of LO based on etiology is limited, and the authors present three such rare cases.

Case Presentations

All three minor patients were included in the study after obtaining consent for publication from their parents.

Case 1

A 9-year-old boy presented with a history of left ear discharge for four years, which worsened in the last one year, becoming persistent and foul smelling. He also had gradually progressive hearing loss in the left ear, affecting his scholastic performance. On examination, there was purulent drainage from the left ear with granulation tissue in the posterosuperior quadrant of the pars tensa as well as in the pars flaccida region with scutum erosion. Pure tone audiometry showed profound SNHL on the left side. High-resolution computed tomography (HRCT) and

magnetic resonance imaging (MRI) showed ossicular erosion together with erosion of the bony walls in the middle ear and complete bony ossification in the apical turn of the cochlea and partial bony ossification in the middle and the basal turn of cochlea (Figures 1, 2). He underwent left canal wall down mastoidectomy, and bone-anchored hearing aid and bone conduction hearing aid were offered for the benefit of binaural hearing.

Case 2

A 6-year-old girl with a history of head injury and meningitis presented with impaired hearing for the past three years. An anterior cranial fossa cerebrospinal fluid (CSF) fistula was diagnosed as the culprit for meningitis and repaired by frontal craniectomy. Otological examination showed reduced absolute bone conduction bilaterally, and pure tone audiometry showed profound SNHL. HRCT and MRI of the inner ear showed bilateral cochlear ossification, and ossification of lateral semicircular canals, suggestive of LO as a sequalae to meningitis (Figures 3, 4). The parents were counselled about the prognosis of hearing loss and its impact
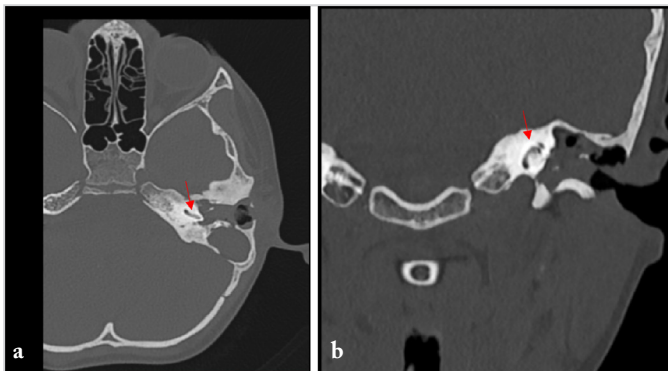

Figure 1. a) HRCT of temporal bones with axial cuts showing ossification within basal turn of left cochlea (red arrow), b) Coronal cut showing higher degree of ossification of middle turn and apical turn of left cochlea compared to basal turn (red arrow)
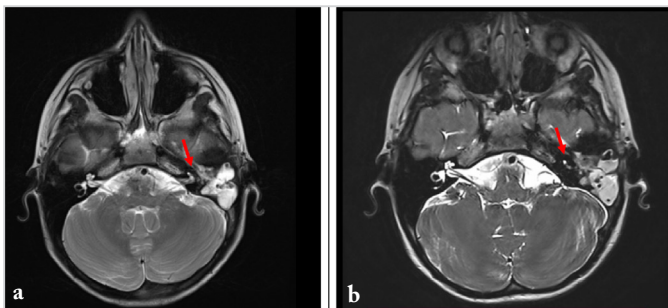
HRCT: High-resolution computed tomography

Figure 2. a, b) T2-weighted magnetic resonance images axial cuts showing the external auditory canal, the middle ear cavity, the aditus and mastoid air cells are filled with contents appearing T2 hyperintense with patchy restriction on diffusion, with loss of T2 hyperintensity in cochlear turns and semicircular canals on left side (red arrow)

on the child's development. The patient underwent cochlear implantation (CI) on the left side with the Veria technique, with drill-out procedure addressing partial ossification of basal turn of cochlea (Figure 5). The child recovered well, and CI switch-on was done on postoperative day 21, followed by rehabilitation therapy. Follow-up categories of auditory performance scores were promising.

Case 3

A 4-year-old girl with delayed speech and impaired hearing for two years following a head injury from a fall was evaluated. Bleeding from both ears was reported. Pure-tone audiometry and brainstem-evoked response audiometry confirmed profound SNHL. HRCT showed complete ossification of bilateral cochlea and the vestibule, and partial ossification of the semicircular canals. MRI showed complete loss of T2 hyperintensity in the cochlea and the vestibule, and partial loss in the semicircular canals (Figures 6, 7). After counselling the parents about the prognosis and the impact on the child's development, CI was done on the left side using the Veria technique. Drill-out procedure was used to address the ossification (Figure 8). However, post-implant
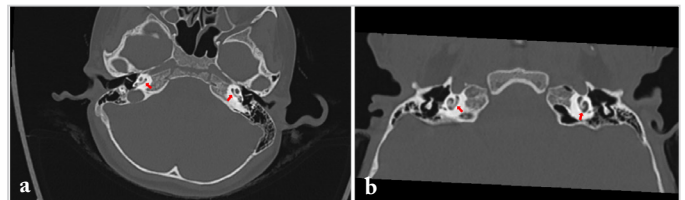

Figure 3. HRCT of temporal bones: a) axial cuts, and (b) coronal cuts showing mild ossification of basal turn and clear middle and apical turns bilaterally (red arrow)
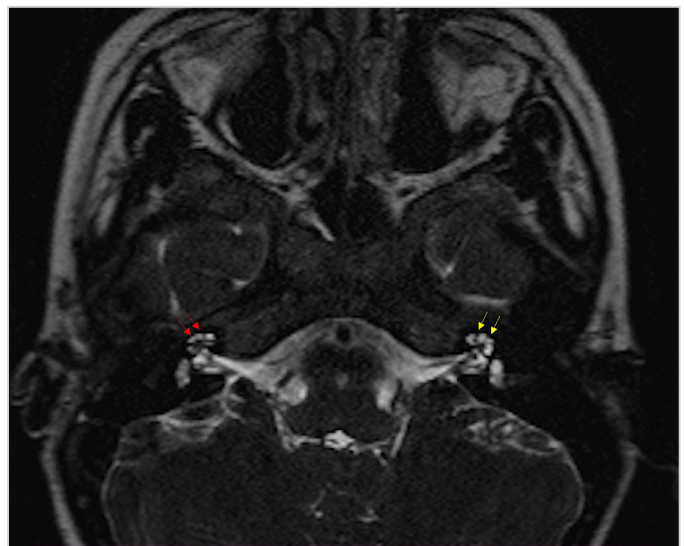
HRCT: High-resolution computed tomography

Figure 4. T2-weighted magnetic resonance images with axial cuts showing loss of hyperintensity in the basal turn of cochlea bilaterally (red arrow on the right and yellow arrow on the left)

responses were poor, and the child was planned for auditory brainstem implantation.

Discussion

Labyrinthitis, a rare complication of chronic otitis media, can be classified into different types (2, 3). Serous labyrinthitis is the best period for patients, as early diagnosis and treatment can reverse hearing loss and prevent complications like LO. Infective causes are more common than inflammatory, with the infection spreading via various pathways. LO is the end result of suppurative labyrinthitis, which can begin as early as three weeks after onset and progress up to nine months (1).

Children with bacterial meningitis have a 5% chance of profound hearing loss, with up to 80% developing some degree of ossification (3). Infection spreads from the subarachnoid space to the labyrinth via the cochlear aqueduct, with LO occurring first and being worst where the aqueduct enters the labyrinth. A central nervous system etiology of LO typically presents bilaterally, unlike tympanogenic. Infection via the cochlear aqueduct causes intense inflammation and ossification at the basal turn of
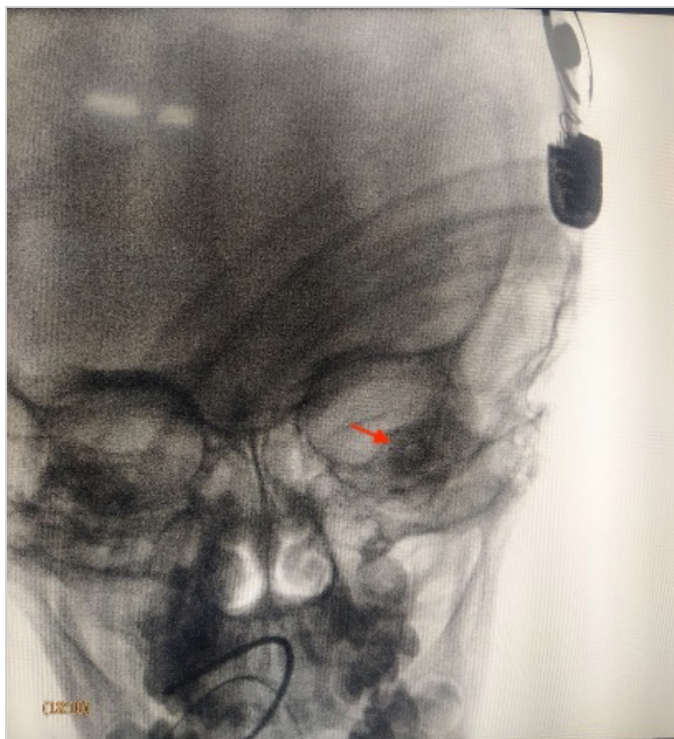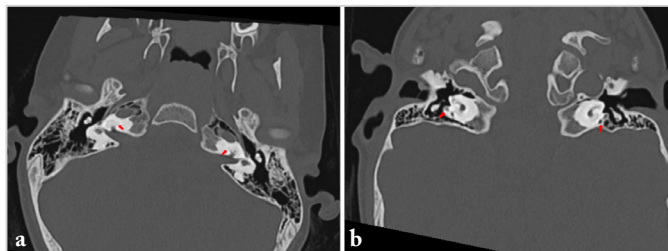

Figure 6. HRCT of temporal bones: a) axial cuts, and b) coronal cuts showing complete ossification of bilateral cochlea, and mild ossification of semicircular canals (red arrow)
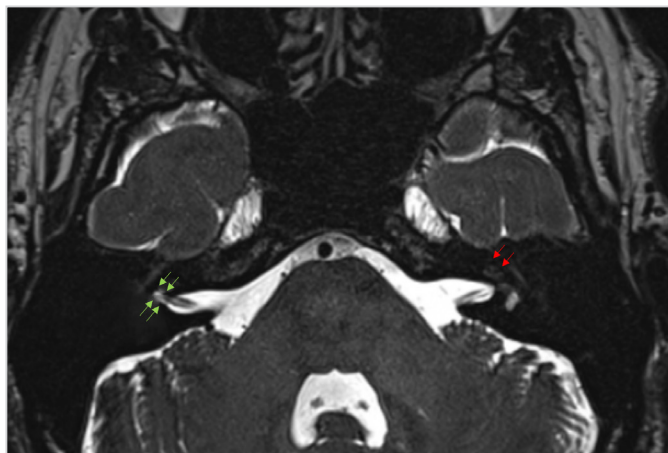
HRCT: High-resolution computed tomography

Figure 7. T2-weighted magnetic resonance images with axial cuts showing loss of hyperintensity in all turns of the cochlea bilaterally with mild hyperintensity in the vestibule and the semicircular canal (green arrows on the right and red arrows on the left)

Figure 5. Postoperative X-ray of skull with trans-orbital view showing the position of electrode

Figure 8. Intra-operative images: a) drill-out procedure (yellow arrow), b) electrode inserted after the drill-out (red arrow), c) insertion test device to assess the length of the electrode (green arrow)

the cochlea, while infection via the internal auditory canal (IAC) leads to more pronounced ossification in the first and second turns according to Aralaşmak et al. (4).

Tympanogenic LO is caused by middle ear pathologies and results in bone formation in the inner ear. Inflammation from the middle ear can spread to the inner ear through preformed pathways, membranes, or channels. The basal turn of the cochlea and lateral semicircular canal are most commonly affected. The disease progresses from inflammation to fibrosis and ossification. Typically, cholesteatoma in the middle ear serves as the nidus of infection. Lin et al. (5) found an incidence of 2% in a retrospective study of 195 patients who underwent mastoidectomy, with four patients testing positive for tympanogenic LO.

LO has three pathological stages: Acute (normal CT, labyrinthine enhancement on MRI), fibrous (loss of fluid signal intensity on T2-weighted MRI; normal CT), and ossification (membranous labyrinth replaced by bone). HRCT is commonly used to classify patients into mild, moderate, or severe stages for management purposes. Buch et al. (6) conducted a radiology study and found that patients with prior temporal bone surgery had significant ossification patterns. The semicircular canals were commonly affected in all etiologies, but no characteristic mineralization pattern was identified in any pathology.

In their histopathological study, Kaya et al. (7) found that LO was mainly observed in the scala tympani of the basal cochlear turn. They also discovered that increased endolymphatic hydrops led to a significant reduction in the spiral ganglion cell population, the outer and inner hair cells in all turns of the cochlea. Additionally, their study revealed differential atrophy of the stria vascularis and the spiral ligament, with the former being greater in all turns of the cochlea and the latter being greater in the basal and middle turns. Merchant et al. (2), in their book, similarly noted that the number of spiral ganglion cells decreased with increasing ossification and duration of deafness.

Tympanogenic LO is commonly found in aggressive cholesteatomas like pediatric cholesteatoma with labyrinth erosion and congenital cholesteatoma in the IAC or the petrous apex. Mesotympanic cholesteatoma cases can also extend to the IAC without LO via the supra-labyrinthine route, requiring extensive treatments like subtotal petrosectomy, translabyrinthine or middle cranial fossa approaches with radical mastoidectomy. In case 1, disease clearance was achieved through modified radical mastoidectomy since cholesteatoma was confined to the mesotympanum and the epitympanum.

Temporal bone trauma is an infrequent cause of LO and is classified into otic capsule sparing and otic capsule violating types (4, 8). The latter type is linked with SNHL, vestibular dysfunction, CSF leak, and peri-lymphatic fistula. However, there is currently no evidence to establish a correlation between otic capsule violating fractures and LO.

This article does not provide a detailed discussion on LO management, but prelingual children with LO are a significant subgroup of CI candidates. The classic drill-out procedure, introduced by Balkany et al. (9), involves drilling a 6–8 mm distance to allow 4–8 electrodes for CI. The compressed electrode array (Form series - MEDEL) and double/split array are commonly used. While the ideal number of spiral ganglion population for serviceable hearing is 3500, and histopathological studies show a decrease in spiral ganglion number, there is no evidence linking spiral ganglion population with auditory-verbal outcomes in CI patients, as supported by a large meta-analysis by Cheng and Svirsky (10). For patients with failed CI in LO, the auditory brainstem implant is an alternative option, although its efficacy remains uncertain and is generally poorer than that of CI.

Michel deformity, otosclerosis, and labyrinthine schwannoma are among the differential diagnoses for LO. Cochlear aplasia or hypoplasia, also known as Michel deformity, has a different labyrinth contour that is altered or absent. Otosclerosis, which causes osteodystrophy of the otic capsule, is classified into fenestral and retro-fenestral (cochlear) types. Although cochlear otosclerosis can radiologically mimic LO, the clinical presentation includes mixed hearing loss and differs in histopathological profile. Table 1 provides a comparison of all three etiologies of LO.

Conclusion

Patients with LO who have profound SNHL can be difficult to diagnose, as routine imaging modalities such as HRCT may not detect early stages of the condition. Therefore, it is recommended that centers with ample resources consider using MRI. Despite the different etiologies of LO, the pattern of mineralization is consistent among them. With recent advances, the drill-out procedure and CI are recommended for bilateral LO, especially in prelingual children.

Table 1. Etiology based comparison of labyrinthitis ossificans (LO)

Features	Meningitic LO	Tympanogenic LO	Traumatic LO
Clinical presentation	Children, Bilateral involvement, Preceding history suggestive of suppurative labyrinthitis.	No specific age group, Associated with squamosal type chronic otitis media, Vestibular symptoms are usually not evident in its history.	Unilateral > bilateral, Bimodal age groups - children and young adults, associated with otic capsule violating fracture.
Imaging features	CT: Irregularities/obliteration/sclerosis of labyrinth. MRI: T2 may show loss of normal fluid signal in the inner ear. T1 with contrast enhancement on MRI is seen in the early acute/subacute stage when there is inflammation-related hypervascularity. These features are more pronounced at the basal turn of the cochlea especially in the scala tympani where it meets the cochlear aqueduct. First and middle cochlear turn involvement with sparing of basal turn suggest spread via the IAC.	Same CT/MR features, with occasional erosive characteristics due to cholesteatoma. Lateral semicircular canal is more commonly involved.	Same as other three, enchondral layer demonstrates fibrous changes.

CT: Computed tomography, MRI: Magnetic resonance imaging, IAC: Internal auditory canal

Main Points

- Labyrinthitis ossificans is a rare but serious complication that can result from severe inner ear infection or inflammation which leads to the abnormal calcification or ossification of the delicate structures of the labyrinth.
- It can cause a range of symptoms, including hearing loss, dizziness, and tinnitus.
- Diagnosis typically involves imaging studies, such as a computed tomography scan or magnetic resonance imaging, to visualize the inner ear and identify any calcifications or ossifications.
- Treatment options for labyrinthitis ossificans are limited and can depend on the severity of symptoms. In some cases, hearing aids or cochlear implants may be recommended to improve hearing.

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