House Syndrome (Fixed Malleus Head): A Diagnostic Pitfall to Consider Before Stapes Surgery

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Abstract

Background: Conductive or mixed hearing loss with a normal tympanic membrane is most often attributed to otosclerosis. However, primary malleus fixation—also known as House or Goodhill syndrome—is a rare differential diagnosis that can mimic stapes fixation and fundamentally alters surgical management [1–4].

Case presentation: A 67-year-old man presented with long-standing, bilateral hypoacusis, more pronounced on the right side. Otoscopy was normal. Audiometry showed bilateral mixed hearing loss with a mean air–bone gap (ABG) of about 30 dB, without a Carhart notch. The initial high-resolution temporal bone CT (HRCT) appeared normal. Endoscopic middle-ear exploration was undertaken for presumed otosclerosis. Intraoperatively, the handle of the malleus was immobile, and stapes palpation did not show the typical fixity of otosclerosis. The procedure was halted for diagnostic reassessment. Re-evaluation of the HRCT revealed ossification of the malleus suspensory ligament, confirming House syndrome [2,5]. A revision transcanal endoscopic ossiculoplasty was then performed, including uncudo-stapedial disarticulation, section of the incus long process, and insertion of a partial ossicular replacement prosthesis (PORP) between the malleus and stapes head. Postoperative recovery was uneventful, with partial ABG closure and significant functional improvement.

Conclusion: House syndrome must be included in the differential diagnosis of conductive hearing loss with a normal tympanic membrane. A meticulous HRCT review focusing on the ossicular ligaments is crucial to avoid unnecessary stapes surgery and to guide appropriate ossiculoplasty.

Keywords: House syndrome; Goodhill syndrome; malleus fixation; fixed malleus head; conductive hearing loss; HRCT; ossiculoplasty; PORP; stapes surgery differential

Introduction

Conductive hearing loss with an intact tympanic membrane is frequently due to otosclerosis; however, other ossicular pathologies may mimic the same audiometric pattern [1,2]. Among them, primary malleus fixation—historically described by Goodhill and later by House—represents a rare but important entity, since its management differs completely from standard stapes surgery [3,4]. High-resolution CT (HRCT) allows visualization of ossified malleal ligaments and other subtle ossicular abnormalities, but accurate detection requires experience and targeted multiplanar reconstruction [5–7]. This case highlights how unrecognized malleus fixation can mislead diagnosis and how proper imaging and intraoperative vigilance prevent unnecessary stapes surgery.

Case Presentation

A 67-year-old male, without any relevant medical history, presented with bilateral progressive hearing loss, worse on the right side. Otoscopy showed intact, normal tympanic membranes. Audiometry confirmed bilateral mixed hearing loss with a mean air–bone gap of approximately 30 dB on the right side and no Carhart notch, which is often absent in non-stapes fixations [1,2]. A high-resolution CT scan of the temporal bones was interpreted as normal, without fenestral otospongiosis. An endoscopic middle-ear exploration was performed for presumed otosclerosis. Intraoperatively, the handle of the malleus was immobile; mobilization did not move the incus, and the stapes appeared mobile—findings incompatible with stapes fixation. The procedure was therefore suspended to allow further evaluation.

A re-reading of the HRCT showed ossification of the anterior malleal ligament, consistent with House syndrome (primary malleus fixation) [2,5]. After informed consent, a revision endoscopic transcanal ossiculoplasty was performed, including uncudo-stapedial disarticulation, section of the incus long process, and placement of a PORP between the malleus and stapes head. Postoperative evolution was simple, with partial closure of the air–bone gap and a clear subjective improvement in hearing.

Discussion

House and Goodhill first described primary malleus fixation as ossification of the anterior or superior malleal ligaments leading to immobility of the malleus head despite a normal middle ear [3,4]. The incidence ranges between 0.4% and 1.6% of cases initially diagnosed as otosclerosis [2,8]. Preoperative differentiation between otosclerosis and ossicular fixation can be difficult based solely on audiometry. HRCT plays a central role in detecting ossified malleal ligaments and excluding fenestral otospongiosis, tympanosclerosis, or ossicular discontinuity [5–7,9]. Thin-slice (≤ 0.6 mm) imaging with coronal and oblique reconstructions enhances detection, and radiology–otology collaboration is essential to preventing misdiagnosis [9,10].

When diagnosed intraoperatively, stapes surgery should not proceed. Surgical options include drilling or laser removal of the ossified ligament or ossiculoplasty with a PORP between the malleus and stapes [1,11]. Successful outcomes depend on accurate diagnosis and adequate restoration of ossicular mobility.

Conclusion

Primary malleus fixation (House syndrome) is a rare but crucial differential diagnosis in patients with conductive hearing loss and a normal tympanic membrane. Thorough preoperative imaging and careful intraoperative assessment are key to avoiding unnecessary stapes surgery and to achieving optimal hearing outcomes.

Ethical Considerations and Acknowledgments

Written informed consent was obtained from the patient for publication of this case report.

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