

Case Report

A rare case of a spindle handle and malleus bar without deformities of the external ear: a case report and literature review

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Received: 31 March 2026

Revised: 30 April 2026

Accepted: 18 September 2026

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ABSTRACT

Congenital fixation of the malleus and incus is an uncommon cause of conductive hearing loss. The malleus bar, characterized by a bony bridge from the malleus neck to the posterior annulus, presents diagnostic challenges preoperatively due to its subtle radiological features. A 9-year-old boy visited our hospital with hearing loss as his main complaint with autism spectrum disorder. The left tympanic membrane lacked identifiable structures such as the short process or umbilicus, and the malleus appeared rod-shaped, oriented anteriorly. Intraoperative findings confirmed a bony cord extending from the posterior wall of the tympanic cavity to the malleus, with the chorda tympani nerve running parallel. The malleus exhibited restricted mobility due to the malleus bar, which was easily severed, leading to enhanced ossicular mobility and hearing improvement. Histopathological analysis confirmed the bony composition of the excised tissue. Postoperative audiometry indicated a reduction in hearing loss, although a residual air-bone gap persisted. This report presents a rare case involving a spindle-shaped malleus handle and malleus bar in the middle ear, occurring without external ear deformities, emphasizing the importance of detailed anatomical assessment.

Keywords: Middle ear malformation, Conductive hearing loss, Spindle handle, Malleus bar

INTRODUCTION

Congenital anomalies of the middle ear have been systematically classified by researchers such as Funasaka and Teunissen et al, providing a structured framework for understanding various developmental malformations.^{1,2} However, the malleus bar, first described by Nomura et al, is notably absent from these classifications. The malleus bar is characterized by a bony bridge extending from the neck of the malleus to the posterior annulus. Nomura et al identified several clinical and anatomical features associated with this anomaly, including a flat tympanic membrane with an absent umbo and a white, cord-like structure between the malleus handle and the posterior

wall of the tympanic cavity, visible through the tympanic membrane.³ These features are essential for accurate preoperative diagnosis and surgical planning. Despite their clinical significance, literature describing characteristic tympanic membrane findings remains limited. This report presents a rare case involving a spindle-shaped malleus handle and malleus bar in the middle ear, occurring without external ear deformities.

CASE REPORT

A nine-year-old male patient with a documented history of autism spectrum disorder was referred for otological evaluation due to pre-existing unilateral hearing

impairment on the left side. The patient's facial skeletal structure appeared normal, with no external auditory canal stenosis or malformations of the external ear observed bilaterally.

Examination of the tympanic membranes revealed a normal right tympanic membrane. Conversely, the left tympanic membrane lacked identifiable structures such as the short process or umbilicus, and the malleus appeared rod-shaped, oriented anteriorly. A white, cord-like structure was visualized extending posteriorly from the malleus (Figure 1). Audiometric testing showed a mild conductive hearing loss on the right side (10 dB in average hearing level of 0.5, 1.0, 2.0, 4.0 kHz) and a moderate conductive loss on the left (37.5 dB in average hearing level of 0.5, 1.0, 2.0, 4.0 kHz), with a maximum air-bone gap of 30 dB on the affected side (Figure 2a). Tympanometry results showed a type C curve on the right and a type of Cs (C shallow) curve on the left, suggesting Eustachian tube dysfunction or middle ear pathology (Figure 3). A temporal bone CT scans interpreted by a radiologist proved no significant malformations of the middle or inner ear structures, with well-aerated tympanic and mastoid cavities bilaterally.

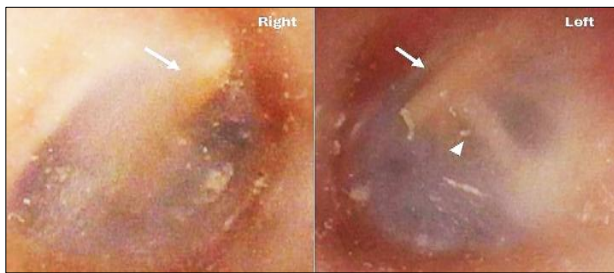


Figure 1: Otoscopic findings.

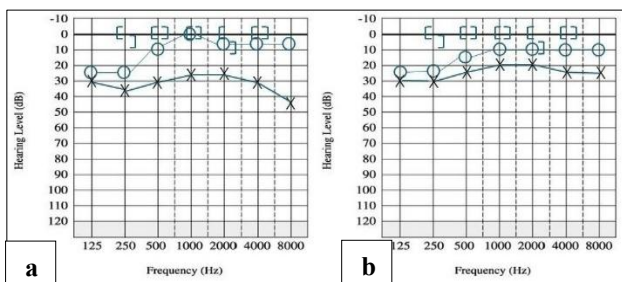


Figure 2: (a) Pre- and (b) post-operative audiograms.

The surgical procedure was performed via the external auditory canal. Upon elevating the tympanomeatal flap, a bony cord was identified extending from the posterior wall of the tympanic cavity to the malleus (Figure 4a), with the chorda tympani nerve running parallel to this structure (Figure 4b). The superstructure of the stapes was intact and mobile, and the incudomalleolar and incudostapedial joints appeared to be normal. However, mobility of the malleus was restricted, consistent with a diagnosis of malleus bar. Palpation indicated that the attachment of the malleus bar to the malleus was relatively soft and could be

easily severed with malleus nippers. The attachment to the posterior wall was dissected using a chisel, resulting in improved mobility of the malleus and incus. Histopathological examination of the excised bony structure confirmed the presence of bone tissue (Figure 5).

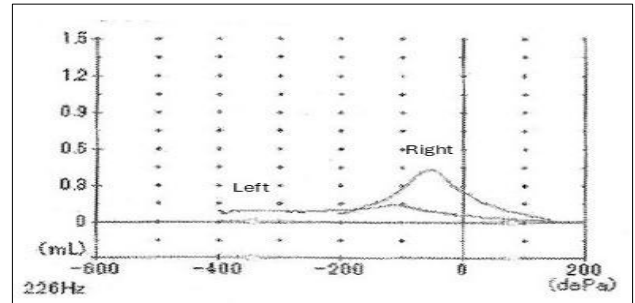


Figure 3: Tympanograms. Tympanometry showed a type C curve on the right and a type of Cs curve on the left.

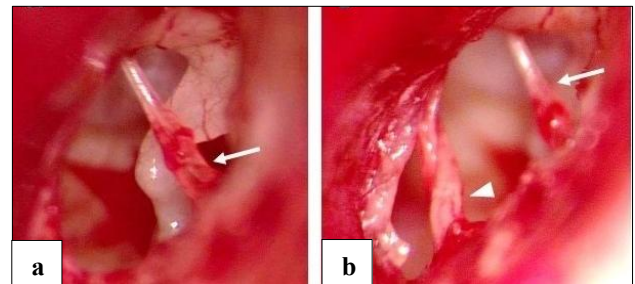


Figure 4: Intraoperative findings (a) a bony cord was identified extending from the posterior wall of the tympanic cavity to the malleus (arrow), and (b) with the chorda tympani nerve running parallel to this structure (arrowhead).



Figure 5: Pathological finding of the removed bar.

At six months post-surgery, audiometric evaluation demonstrated an improvement in hearing thresholds exhibiting a hearing level of 22.5 dB in the average hearing level of 0.5, 1.0, 2.0, and 4.0 kHz (Figure 2b). Despite this improvement, a slight air-bone gap persisted, indicating residual conductive components. The patient, along with his parents, gave written consent to undergo surgical intervention. They expressed satisfaction with the subsequent improvement in auditory function after the operation.

The left tympanic membrane lacked identifiable structures such as the short process or umbilicus, and the malleus appeared rod-shaped, oriented anteriorly (arrow) and normal malleus in the right ear. A white, cord-like structure was visualized extending posteriorly from the malleus (arrowhead).

DISCUSSION

Nomura et al reported two cases featuring a spindle handle of the malleus and four cases with a malleus bar among a cohort of 54 patients presenting with narrow auditory canal stenosis. These morphological features-the spindle handle and malleus bar-rarely co-occur, with no cases exhibiting both anomalies simultaneously. The spindle handle is characterized by a rod-shaped, anteriorly directed malleus, often associated with a flat tympanic membrane lacking an umbo. Malformations involving the long crus of the incus and absence of the lenticular process are also observed in these cases. These findings underscore the importance of detailed radiological and surgical assessments in patients with external auditory canal stenosis to identify associated ossicular anomalies, which can significantly impact clinical management and surgical planning.^{3,4}

The malleus bar is often associated with specific clinical and morphological characteristics, including a narrowed external auditory canal, a flat tympanic membrane lacking a short process, and an indistinct handle of the malleus on the tympanic membrane surface. Recent case reports have identified instances of congenital malleus bars occurring independently of congenital aural stenosis or atresia, suggesting variability in presentation.⁶ These features are significant in the assessment of middle ear pathology and can influence surgical planning and outcomes. Notably, there have been no documented cases describing the presence of thin, rod-shaped trabeculae extending from the posterior aspect of the malleus neck, indicating a potential area for further investigation into the morphological spectrum of this condition. Understanding the detailed anatomy and variations of the malleus bar is essential for otologic surgeons and radiologists involved in diagnosing and managing middle ear conditions.^{5,6}

Diagnostic imaging techniques, especially high-resolution thin-slice computed tomography (CT) scans, have proven effective in detecting the presence of a malleus bar, thereby facilitating precise surgical planning and intervention.⁷ However, these features may not be detectable in all cases, as exemplified by the present case.

Surgical management typically involves removal of the malleus bar, which often results in significant hearing improvement; however, minor residual air-bone gaps may persist postoperatively in the present case.^{3,5,6} It is crucial to consider minor ossicular fixation or additional malformations, as these can influence surgical strategy and prognosis.⁸ When the malleus bar is notably thick,

meticulous surgical technique is essential to prevent inner ear injury.^{9, 10} In such cases, a safer approach may involve removal of the ossicular chain followed by reconstruction, with the decision to preserve or reconstruct the ossicular chain based on specific anatomical features and ossicular integrity.

CONCLUSION

This report presents a rare case involving a spindle-shaped malleus handle and malleus bar in the middle ear, occurring without external ear deformities, emphasizing the importance of detailed anatomical assessment.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Aoki M, Yamaguchi C, Obara N, Suzuki K, Isono Y, Mori H. A rare case of a spindle handle and malleus bar without deformities of the external ear: a case report and literature review. *Int J Otorhinolaryngol Head Neck Surg* 2026;12:811-3.