Incus long process dysplasia – a clinical case

Cinical Case

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Abstract

Middle ear malformations are rare events, occurring in approximately 1 in 15,000 live births.1–3 They are generally associated with other head and neck anomalies and may also be part of syndromic malformations. Isolated malformations of the middle ear are therefore even rarer and pose a diagnostic challenge4, especially given the normal appearance of the auricle, external auditory canal, and tympanic membrane.

In this article, we describe the clinical case of a 19-year-old female patient with a history of untreated hearing loss since childhood. Otolaryngological examination revealed no abnormalities, prompting further audiometric evaluation. Pure tone audiometry demonstrated a left-sided conductive hearing loss with an air-bone gap (ABG) of 54 dB and a type A tympanogram. Computed tomography (CT) was reported as normal. An exploratory tympanotomy was therefore performed, revealing an isolated dysplasia of the long process of the incus. A type II ossiculoplasty (Portmann classification)14 was performed using autologous cortical mastoid bone and tragal cartilage grafts. The postoperative audiogram showed a reduction in the average ABG to 18.75 dB.

Keywords: Dysplasia; Incus; Malformation

Introduction

Middle ear malformations (MEMs) are rare, occurring in one in 15,000 live births[1–3]. They can be classified as major or minor[6]. MEMs are considered major when they are associated with abnormalities of the tympanic membrane, external auditory canal, or auricle; and minor when they occur in isolation[7,8]. They may also be associated with other congenital anomalies of the head and neck or may be part of a broader malformation syndrome.

Minor MEMs are rarer and more difficult to diagnose due to the absence of externally visible abnormalities. According to the literature, the time to diagnosis for minor MEMs often exceeds six years[4,5].

Case report

A 19-year-old woman, born in Brazil and residing in Portugal for over five years, presented for an otorhinolaryngology (ORL) evaluation due to left-sided hearing loss, which was reportedly present "since birth." She had no personal or family history of hearing loss, head trauma, or use of ototoxic medications. She revealed that she was evaluated by an ORL specialist during childhood and diagnosed with otitis media with effusion. The patient denied having undergone any surgical intervention. Otoscopy revealed no deformities of the auricles or abnormalities of the external auditory canals or tympanic membranes bilaterally. Pure-tone and speech audiometry showed conductive hearing loss on the left side, with an average air-bone gap (ABG) of 54 dB, speech recognition threshold (SRT) of 70 dB, and 100% speech discrimination at 100 dB. The tympanogram was type A, according to the Jerger classification. On the right side, hearing was within normal limits, with an SRT of 15 dB, 100% speech discrimination at 45 dB, and type A tympanogram (Jerger classification). Computed tomography (CT) of the ears (Figure 1) revealed no anatomical abnormalities. Exploratory tympanotomy was subsequently performed and revealed isolated dysplasia of the long process of the incus (Figure 2). Subsequently, a type II ossiculoplasty (Portmann classification)[14] was

Figure 1
computed tomography (CT) scan of the left ear - coronal view

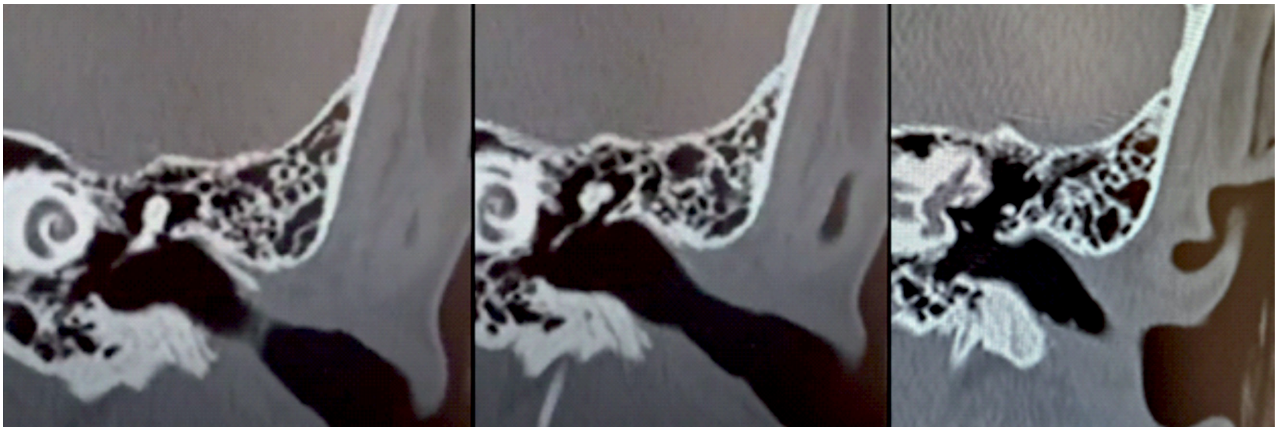
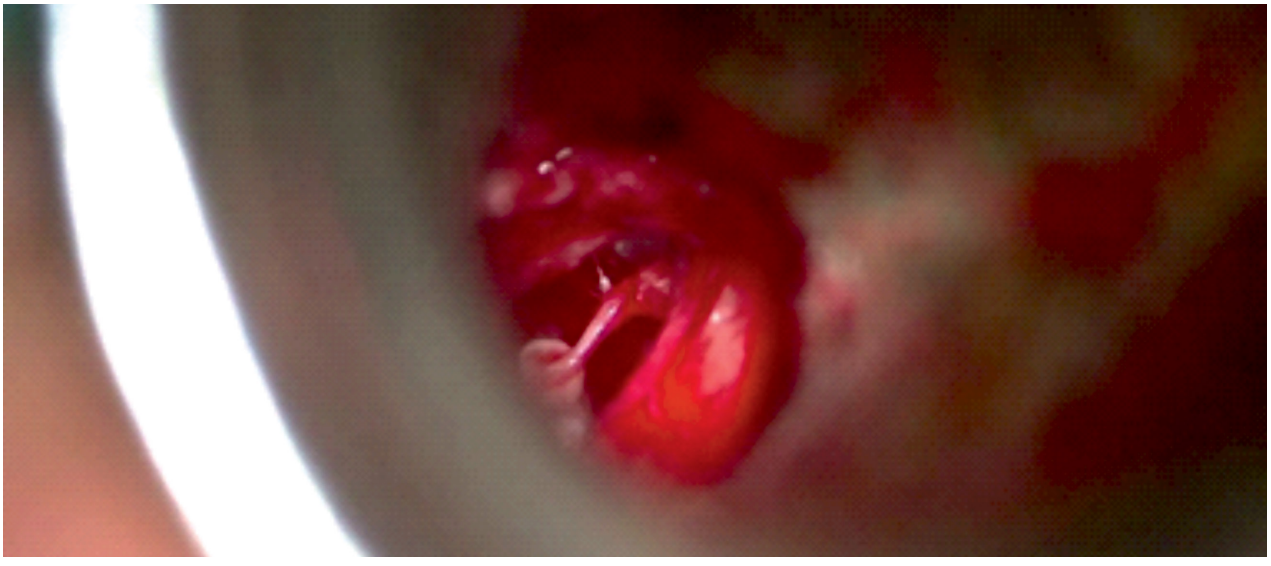


Figure 2
Dysplasia of the long process of the incus



performed using an autologous graft of the mastoid cortex and tragal cartilage, interposed between the stapes head and the tympanic membrane. The postoperative period was uneventful. The patient reported a subjective improvement in hearing immediately after removal of the ear packing seven days after surgery. Audiometry performed two months postoperatively demonstrated an ABG reduction to 18.75 db on the left side (figure 3).

Discussion

Isolated (minor) congenital MEMs account for less than 1% of all causes of conductive hearing loss in children[9]. Unlike major malformations, the lack of observable anatomical abnormalities on physical examination often leads to substantial diagnostic, with considerable implications for hearing, as well as for psychosocial, cognitive, and educational development.

These isolated malformations were practically unknown until they were described by Hough et al. in the late 1950s[10]. The ossicular chain develops between the fifth and seventh week of gestation from the fusion of Meckel's cartilage (derived from the first branchial arch, and gives rise to the malleus and body of the incus) and Reichert's cartilage (originates the second branchial arch, and gives rise to the long process of the incus and stapes superstructure)[11]. The stapes footplate develops from the otic capsule.

Based on an understanding of embryology and surgical management of congenital middle ear anomalies, Cremers and Teunissen proposed a classification system for MEMs in 1993, categorizing them into four types[12] (Table 1). Although the frequency of each type varies across different studies, type IV malformations are considered the rarest. In the case described above, the patient was considered to have type IIIa MEM.

The diagnosis of isolated MEMs requires additional diagnostic workup, including

Figure 3
Pure-tone audiogram
Left – Preoperative audiogram with mean air-bone gap (ABG) of 54 dB
Left – Postoperative audiogram with mean ABG of 18.75 dB

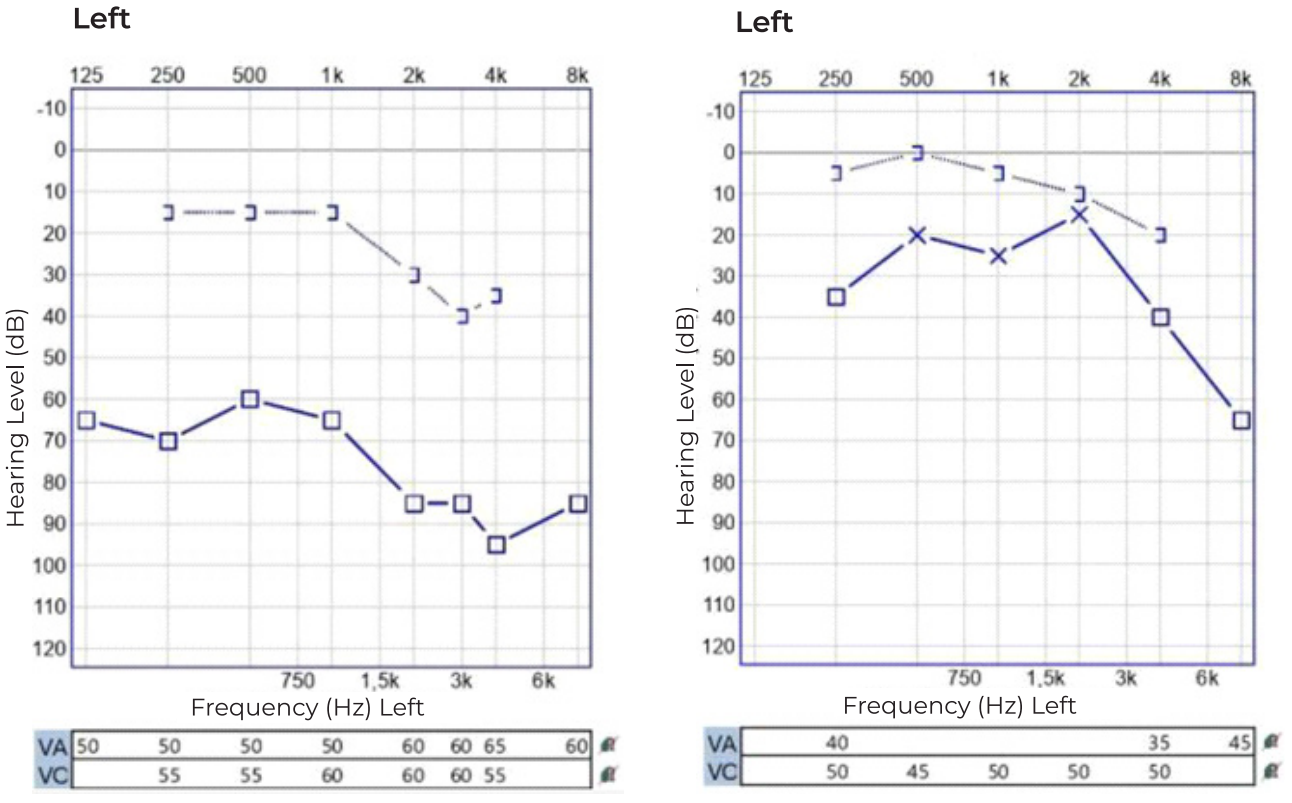


Table 1
Cremers and Teunissen classification of middle ear malformations (1993)

Cremers and Teunissen classification	
Type I	Isolated ankylosis of the stapes footplate
Type II	Stapes ankylosis associated with ossicular chain malformation
Type III	Ossicular chain malformation (discontinuity or fixation) with a mobile footplate
IIIa	Ossicular chain discontinuity
IIIb	Ossicular chain fixation
Type IV	Oval and/or round window aplasia or dysplasia with or without facial nerve anomaly

audiometric evaluation, which normally shows conductive or mixed hearing loss, and imaging. High-resolution CT is the modality of choice, as it provides adequate anatomical detail of the osseous structures. However, despite the recent advances in imaging resolution, CT may still miss some subtle malformations. In a 2019 study, Zhang et al. observed that in their series of 145 ears with minor MEMs, 37.9% were classified as normal on CT, and the diagnosis was ultimately confirmed through exploratory tympanotomy[4], as occurred in our case.

The treatment of MEMs depend on their classification. This classification has been widely implemented because it helps to predict the most appropriate treatment approach for each type. For type I MEMs, stapedotomy is the first-line treatment; for type III, tympanoplasty with ossiculoplasty is usually indicated; and for type IV, fenestration with piston placement may be the most viable surgical option, when surgery is indicated[6]. Treatment of type II MEMs depends on the individual anatomy, but one possible option is malleus-to-stapedotomy reconstruction. For all MEM types, viable alternatives include conventional hearing aids, osseointegrated implants, and contralateral routing of signals (CROS)/bilateral contralateral routing of signals (biCROS) systems, which may be considered in patients who decline surgery[6, 13]. In the case reported here, we opted for type II reconstruction (Portmann classification)[14], and used an interposition graft made of autologous mastoid cortical bone and tragal cartilage.

Figure 4
QR code for a short explanatory video of the clinical case

Conclusion

Isolated MEMs are rare and associated with a mean diagnostic delay of more than six years[4,5]. Definitive diagnosis may be achieved through high-resolution CT or exploratory tympanotomy.

Despite their low prevalence, MEMs should be considered in the differential diagnosis of conductive hearing loss with normal otoscopy, especially in younger patients.

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

Data Confidentiality

The authors declare having followed the protocols used at their working center regarding patient data publication.

Protection of humans and animals

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and the 2013 Helsinki Declaration of The World Medical Association.

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Availability of scientific data

There are no datasets available, or publicity related to this work.

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