

Case Report

Right Incus Osteoma in a Child: A Differential Diagnosis of Middle Ear Malformations

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Cite this article as: Benoit C, Chebib E, Bloy O, Elmaleh M, Morcrette G, Van Den Abbeele T. Right incus osteoma in a child: A differential diagnosis of middle ear malformations. *J Int Adv Otol.* 2021;17(6):566-569.

Osteomas of the temporal bone, especially those involving the incus, are rare, unilateral, benign osseous tumors. The clinical presentation is usually isolated conductive hearing loss, and the diagnosis is confirmed by a temporal computed tomography scan. Osteomas of the incus represent a differential diagnosis of congenital middle ear malformations in children, which are the most frequent cause of conductive hearing loss with a normal eardrum in the pediatric population. In case of disabling symptomatology, surgery seems to be a safe way to recover normal hearing.

KEYWORDS: Conductive hearing loss, incus osteoma, middle ear, pediatric audiology

INTRODUCTION

Osteomas of the temporal bone are well-known, benign, bony tumors that usually arise from the external auditory canal or the mastoid. However, osteomas of the middle ear are exceptionally rare, with fewer than 40 cases reported in the literature, of which only 6 were incus osteomas.^{1,2} These tumors are usually unique and are not clearly associated with trauma, infection, or any other specific medical history. As a result, their pathogenesis remains controversial. The most common clinical presentation is a progressive conductive hearing loss. This case report describes the clinical presentation and management of a child who presented with unilateral conductive hearing loss due to an osteoma of the incus.

CASE REPORT

A 13-year-old girl was referred to our department for progressive right-sided hearing loss over several months and increasing difficulty in the classroom. No head trauma, familial otologic disease, or other Ear-Nose-Throat (ENT) pathologies (including otitis media) were reported, and the onset of symptoms was difficult to date. Pure tone audiometry (Figure 1A) showed mild conductive hearing loss in the right ear at 0.5, 1, 2, and 4 kHz (mean value at 30 dB with a 20-dB air–bone gap) and normal hearing on the left side. There was no tinnitus or balance disorders. The presence of a 25-dB Carhart notch at 2 kHz and abolition of the ipsilateral stapedius reflex suggested possible stapedovestibular ankylosis. The eardrums were normal on both sides.

A temporal bone high-resolution computed tomography (CT) scan revealed a uniform, deep osseous density (4 × 5 mm mass) seemingly attached to the body of the incus and concordant with an osteoma of the incus (Figure 2). No other abnormalities were found during this examination, in particular there was no footplate thickening.

Because of the patient's hearing complaints, we offered a choice between conventional hearing aids and surgical intervention, together with complete information concerning the operative risks and complications. A few months later, the family and the child decided in favor of surgery due to the growing impact of the deafness on her schooling. The surgical procedure, a right

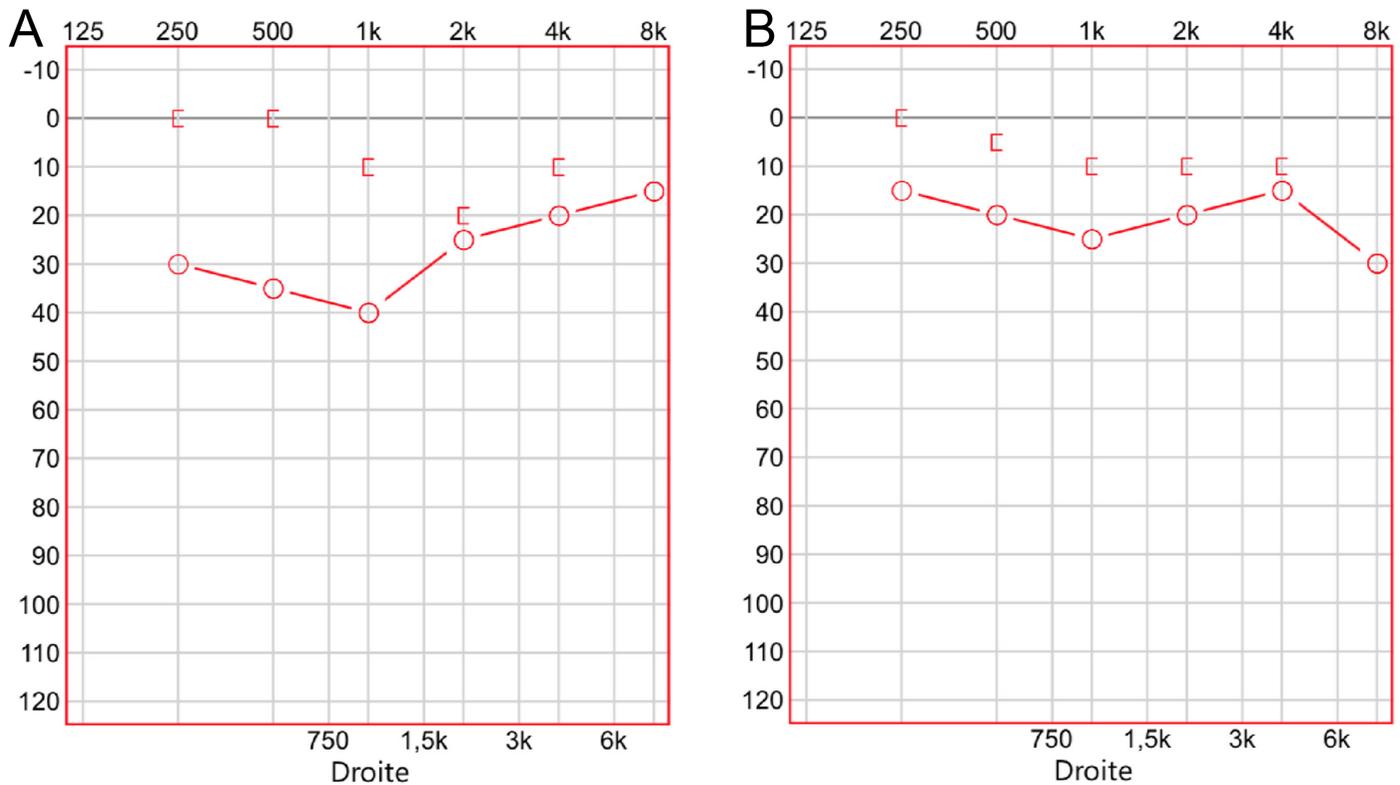


Figure 1. (A) Preoperative pure tone audiometry (dB on the left, Hz on the top). (B) Postoperative pure tone audiometry (dB on the left, Hz on the top).

tympantomastoidectomy, was performed under general anesthesia. We found a complete ossicular blockade due to the osteoma and its fixation to the incus body and the surrounding mastoid bone (Figure 3). The incudostapedial joint was then fully disarticulated in order to prevent the high risk of inner ear trauma during dissection of the osteoma. The osteoma was then carefully mobilized from the bony edges of the fossa incudis and gently detached from the incus body. The incus was then found almost intact, removed, sculpted “ex corpore,” and fitted between the malleus and the capitulum of the stapes. This procedure led to a satisfactory improvement in air conduction (+15 dB at 0.5-1 kHz and +5 dB at 2-4 kHz), a mean +10 dB closure of the air–bone gap and complete reduction of the Carhart notch (Figure 1B). Histopathological analysis of the tumor and hematoxylin–eosin staining showed a very thick Haversian cortex-type bone structure with numerous concentric apposition lines and small

osteocytes and, therefore, allowed the diagnosis of osteoma to be made with certainty and to rule out the differential diagnoses such as reactive new bone formation or ligament ossification of ossicles (Figure 4).

DISCUSSION

To our knowledge, this case report presents one of the first cases of incudal osteoma in the pediatric population. Although osteomas are the most common benign tumors of the temporal bone, very few occur in the middle ear. Most are located on the promontory, with the incus being the second localization in the 6 cases described.^{1,2} However, ossicular osteomas are often asymptomatic and probably underdiagnosed.³ The etiopathogenesis remains unclear; however, a congenital or genetic origin is suspected in pediatric cases as suggested by the possible association with congenital cholesteatoma or familial cases.¹

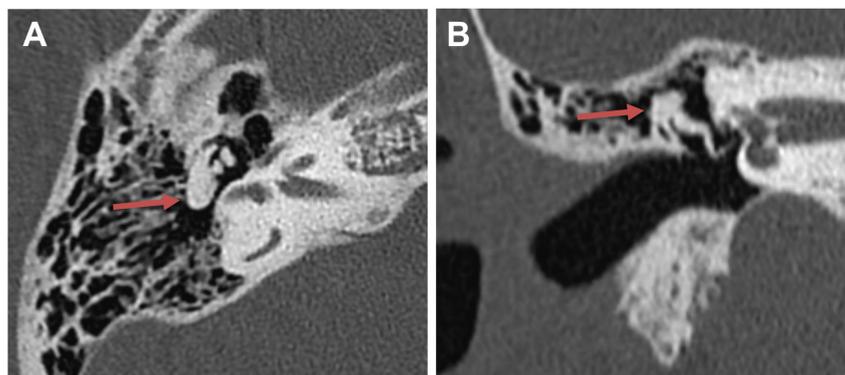


Figure 2. Right temporal bone computed tomography scan highlighting the osteoma attached to the head of the incus and the epitympanum. (A) Axial section and (B) coronal section. Note the integrity of the stapes and footplate. Arrows indicate the osteoma.

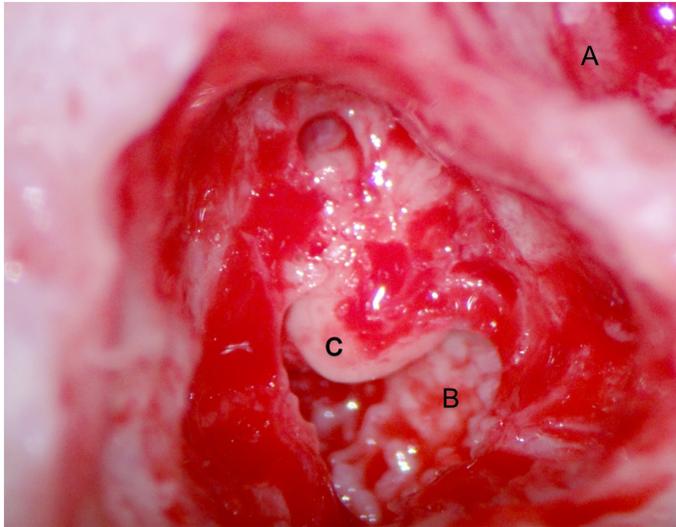


Figure 3. Intraoperative findings: (A) external auditory canal; (B) right tympanomastoidectomy; (C) incus body osteoma.

In the pediatric population, middle ear osteomas are very rare and difficult to detect. In their review, Yoon et al¹ reported 34 cases with a mean age of 28 years, with 12 patients under the age of 18, of which only 3 osteomas originated from the ossicles.¹ In pediatric cases presenting with conductive hearing loss associated with a normal eardrum and no history of infections, the primary diagnosis is usually a congenital middle ear malformation that occurs in 1 out of every 15,000 births.⁴ Other hypotheses could be made, such as inflammatory ossicular chain blockade, juvenile otosclerosis, ossicular disjunction, or middle ear osteoma. Diagnosis is complicated by the frequent medical history of recurrent otitis media or chronic effusion in children, which can lead to hearing loss. Moreover, most osteomas are single-sided and slow-growing lesions that do not lead to delayed language acquisition neither to an early hearing test.

A Carhart notch at 2 kHz was clearly observed in our case, suggesting a footplate fixation, such as in otosclerosis or congenital stapes malformation. However, Kashio et al⁵ outlined that this notch is not specific to a footplate fixation and can also be seen in cases of incudostapedial joint detachment and malleus or incus fixation.^{5,6} Thin bone window sections (axial and/or coronal) on a temporal bone CT scan without injection easily confirm an incus osteoma diagnosis and eliminate

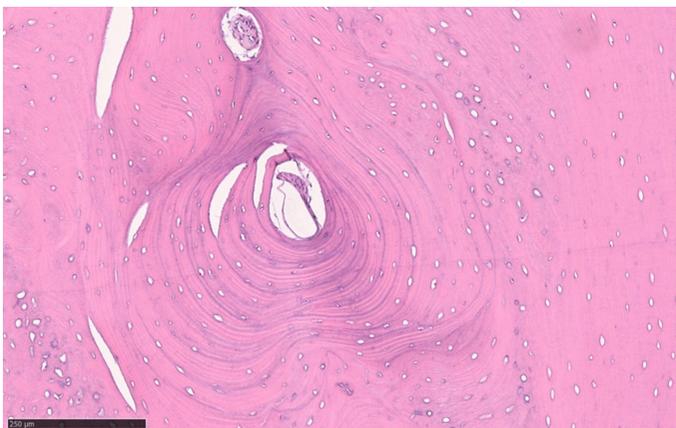


Figure 4. Hematoxylin and eosin staining of the incus osteoma.

other hypotheses, such as footplate anomalies. This highlights the importance of performing a temporal bone CT scan prior to any otologic surgery in case of hearing loss, as suggested in several studies.⁷

Osteomas of the middle ear are benign and usually slow-growing tumors, allowing simple watchful waiting in cases with minor complaints. Because of our patient's audiological complaints and her refusal to wear hearing aids, surgery was performed and it proved to be efficient. Our experience of surgery for congenital middle ear malformations⁴ led us to expect, in this case, a good functional result, due to the absence of inflammatory lesions and tympanic perforation, as well as the integrity on CT scan of the malleus, stapes, and footplate. The ossiculoplasty outcome parameter staging score was calculated at 1 suggesting a low risk of complications and a good functional prognosis.⁸

However, there is no specific surgical procedure recommended for the treatment of middle ear osteomas. After resection of the osteoma, the shape and thickness of the residual incus were sufficient for an autograft reconstruction stable over time. It avoided the placement of a prosthesis and therefore a foreign body, even if these prostheses are very well tolerated.⁴ It is certain that if the transposition had seemed not enough strength, we would have chosen a partial ossicular chain reconstruction prosthesis.

The choice of surgical procedure depends on the surgeon's experience and on local intraoperative conditions. The auditory results of these different surgical techniques are almost equivalent as reported by Quesnel et al⁴ concerning congenital middle ear anomalies.

CONCLUSION

Although exceptional, osteomas of the ossicular chain should be considered in children presenting with unilateral conductive hearing loss and a normal eardrum. In this situation, physicians should perform a systematic temporal bone high-resolution CT scan in order to make the appropriate diagnosis and to manage treatment and follow-up. Even if the risks of middle ear surgery are minor, the surgical removal of osteomas and ossicular reconstruction can induce sensorineural hearing loss; therefore, surgical treatment should be reserved only for disabling symptomatology, and hearing aids should be first proposed in case of mild hearing loss.

Informed Consent: Written informed consent was obtained from the patients who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – C.B., E.C., T.V.D.A.; Design – C.B., T.V.D.A.; Supervision – M.E., G.M., T.V.D.A.; Materials – G.M., M.E.; Data Collection and/or Processing – C.B., T.V.D.A.; Analysis and/or Interpretation – C.B., G.M., M.E., T.V.D.A.; Literature Search – C.B., T.V.D.A.; Writing – C.B., T.V.D.A.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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