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Case Report

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Juvenile Otosclerosis: A Case Report

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ABSTRACT

Juvenile otosclerosis is an uncommon but important cause of progressive conductive hearing loss in children and adolescents. Its diagnosis can be challenging because of its rarity, subtle radiologic findings, and similarities to other middle ear pathologies. We present the case of a 12-year-old girl who developed progressive right-sided hearing loss accompanied by intermittent tinnitus, without a history of recurrent otitis media or speech delay. Clinical, audiological, and high-resolution CT evaluations confirmed right-sided Type 1A otospongiosis. As her audiometric thresholds remained stable, a conservative management plan with hearing aids was chosen following shared decision-making with the patient and her family. This case highlights the value of thorough clinical and imaging assessments in diagnosing juvenile otosclerosis and emphasizes that conservative management can be appropriate for selected pediatric patients. Differentiation from congenital stapes fixation is essential given the differing treatment implications. Overall, juvenile otosclerosis should be considered in the differential diagnosis of pediatric conductive hearing loss, and individualized, multidisciplinary management remains key to optimizing outcomes.

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Introduction

Juvenile Otosclerosis (JO) is a rare but important cause of progressive conductive hearing loss (CHL) in children and adolescents, characterized by abnormal bone remodeling of the otic capsule, most commonly around the stapes footplate. Despite being well-documented in adults, its diagnosis in the pediatric population remains challenging due to its low incidence, overlapping clinical features with other causes of CHL, and often subtle radiological findings. The condition may be mistaken for congenital stapes fixation or other ossicular anomalies, making accurate diagnosis critical for effective management. While CT imaging is considered the gold standard for evaluating middle and inner ear structures, the role of preoperative imaging in diagnosing juvenile otosclerosis has not been thoroughly explored. Moreover, the optimal management of JO whether through hearing amplification or stapes surgery remains controversial, primarily due to concerns over surgical risks and variability in outcomes. This case report aims to contribute to the limited literature by presenting our experience with juvenile otosclerosis, supported by radiologic findings and audiological outcomes of a 12 years old patient

Case Report

A 12-year-old female patient, born to second-degree consanguineous parents, presented with a history of progressive right-sided hearing loss evolving over the past four years. The hearing impairment was accompanied by intermittent tinnitus, without any associated vertigo, facial asymmetry, language delay, or speech articulation issues. There was no relevant past medical history apart from an adenoidectomy performed at the age of five.

On clinical examination, the patient was alert, with normal breathing and no dysmorphic features. Nasal breathing was present and symetrical. Psychomotor and language development were appropriate for age. Otologic examination revealed normally developed and symmetrically positioned auricles. Otoscopic evaluation showed clear external auditory canals and intact, healthy tympanic membranes bilaterally. Tuning fork tests revealed a positive Rinne test on both sides and a Weber test lateralized to the left ear, indicating sensorineural hearing loss on the right.

Rhinological and nasopharyngeal evaluation demonstrated residual adenoid tissue at the roof of the nasopharynx, with patent tubal orifices. The remainder of the ENT examination, including vestibular, orobuccal, and cervical assessment, was unremarkable.

Pure-tone audiometry confirmed right-sided sensorineural hearing loss of moderate to severe degree, with a bone conduction average of 55 dB and an associated mild to moderate conductive component demonstrated by an air-bone gap of approximately 17 dB (Figure 1; A).

Impedancemetric evaluation demonstrated normal middle ear pressure and compliance bilaterally, with preserved stapedial reflexes on the left but absent on the right. Tympanometric values were as follows: for the right ear (OD), compliance 0.56 ml, volume 0.82 ml, pressure –14 daPa, and gradient 75 daPa; for the left ear (OG), compliance 0.62 ml, volume 0.77 ml, pressure 4 daPa, and gradient 36 daPa (Figure 2; B).

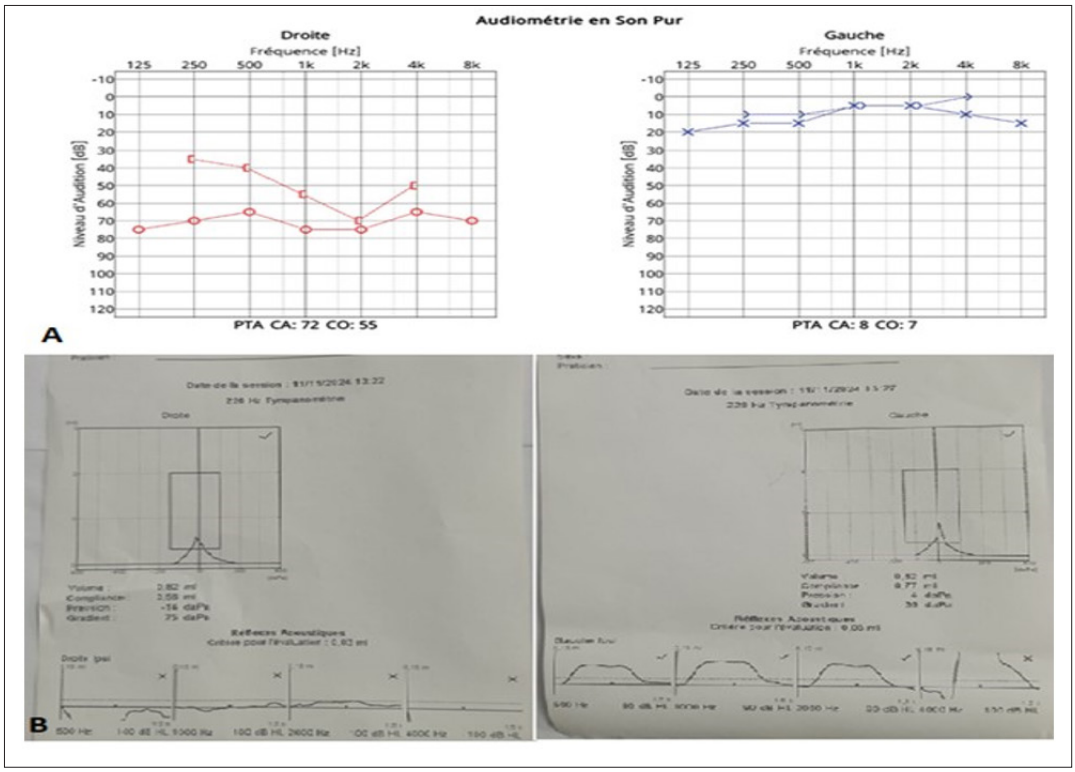

Figure 1: A: The audiogram reveals a mixed hearing loss in the right ear, with a moderate to severe sensorineural component (bone conduction average of 55 dB) combined with a mild to moderate conductive component (air-bone gap of approximately 17 dB), B: Impedancemetric results showing middle ear pressure and compliance bilaterally, with preserved stapedial reflexes on the left but absent on the right

High-resolution computed tomography (CT) of the temporal bones identified Type 1A otospongiosis on the right side and bilateral dehiscence of the tegmen tympani (Figure 2). Magnetic resonance imaging (MRI) of the brain, internal auditory canals, labyrinth, and cerebellopontine angles showed no abnormalities.

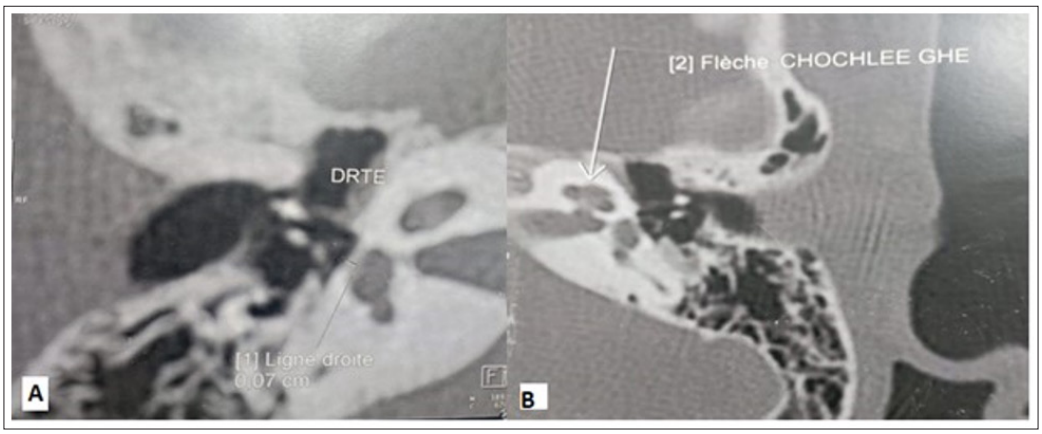

Figure 2: Axial view of Temporal bone CT of the right (A) and left ears (B) showing stage IA otosclerosis on the right A: The audiogram reveals a mixed hearing loss in the right ear, with a moderate to severe sensorineural component (bone conduction average of 55 dB) combined with a mild to moderate conductive component (air-bone gap of approximately 17 dB), B: Impedancemetric results showing middle ear pressure and compliance bilaterally, with preserved stapedial reflexes on the left but absent on the rightside.

Serological testing for Cytomegalovirus (CMV) infection revealed a negative qualitative IgM antibody index (0.04 E/S) and a positive IgG titer of 19.00 UA/mL, consistent with past CMV exposure without evidence of recent or active infection.

After discussion of treatement with the patient and her parents, hearing aids was trialed and found to be well tolerated, offering a non-invasive and effective alternative. With regular audiologic surveillance.

Discussion

Juvenile otosclerosis (JO) is an uncommon but clinically important cause of progressive conductive hearing loss (CHL) in children and adolescents. Its estimated prevalence is relatively low, with otosclerotic foci identified in only 0.6% of children under five and up to 4% of those aged 5–18 years [1]. Because of its rarity, the diagnosis of JO is often delayed or misattributed to more common pediatric conditions such as otitis media, congenital stapes fixation (CSF), or ossicular chain anomalies [2,3]. Our case, involving a 12-year-old girl, illustrates the importance of a thorough clinical and radiologic evaluation in establishing the diagnosis and guiding a tailored management plan. In this instance, a non-surgical, conservative approach was chosen based on clinical stability, imaging findings, and shared decision-making with the patient and her family.

Differentiating JO from CSF is essential, as both conditions may present similarly on audiological testing but have different natural histories and implications for treatment. CSF typically presents earlier in life, often before the age of five, and tends to remain stable over time. In contrast, JO generally appears in late childhood or adolescence and is often progressive. Additionally, JO may have a positive family history, present in up to 53% of cases [4,5]. In our patient, the age of onset, absence of a history of chronic otitis media, and family background supported the diagnosis of JO. Serial audiological assessments showed stable thresholds, further reinforcing the decision to observe rather than operate.

High-resolution computed tomography (HRCT) is a critical tool in the evaluation of JO. Although its sensitivity is lower in children than in adults due to the subtlety of early lesions CT remains essential for confirming the diagnosis, excluding alternative causes of CHL, and identifying any anatomical variations that might increase the risk of complications if surgery is pursued [6]. In recent years high- resolution cone beam CT scans have been used to acess middle and inner ear with significantly lower effective radiation dose than traditional CT scans [7]. In our case, HRCT revealed findings consistent with otosclerosis, including hypodensity near the fissula ante fenestram, which are typical radiologic hallmarks of the condition [8,9]. The absence of high-risk anatomical features, such as enlarged vestibular aqueducts or other congenital malformations, supported the safety of a conservative approach, should surgery be reconsidered in the future.

Although stapedotomy is widely regarded as the standard treatment for JO with significant hearing loss, its role in pediatric patients remains debated. Advocates for early surgical intervention argue that delaying surgery risks progression to more advanced or obliterative otosclerosis, which may complicate future operative management [10]. Outcomes from pediatric stapedotomy have been encouraging in selected cases, with studies reporting postoperative air-bone gap (ABG) closure within 10 dB in up to 80–90% of cases [11-14]. However, surgical risks including sensorineural hearing loss (SNHL), perilymph fistula, and delayed complications remain a concern, particularly in younger or less compliant patients [15,16]. Moreover, younger children may face challenges with postoperative care, including adherence to physical activity restrictions and recognition of early complications.

In our case, the decision to avoid surgery at this stage was based on several key clinical considerations. The patient's hearing had remained stable across multiple audiograms, and there was no evidence of bilateral disease or severe functional impairment. She exhibited no speech or language delay, and her academic and social development were unaffected. Given these factors, the potential risks of surgical intervention were deemed to outweigh the immediate benefits. Instead, hearing amplification was trialed and found to be well tolerated, offering a non-invasive and effective alternative that preserved the option of surgery in the future if needed.

Involving the patient and her family in the decision-making process was a fundamental aspect of care. While the patient was not yet of legal age to consent independently, her understanding of the condition and engagement in discussions were prioritized. Recent literature emphasizes the value of shared decision-making, especially as children approach adolescence and can actively participate in treatment planning. This collaborative approach fosters trust, ensures that family values and preferences are respected, and supports long-term adherence to monitoring and care recommendations [17].

Finally, it is important to acknowledge the current limitations in the evidence base for JO management. Much of the literature consists of retrospective studies with limited follow-up and variable diagnostic criteria. Furthermore, long-term outcomes of both surgical and non-surgical strategies remain poorly defined. Our case contributes to a growing recognition that conservative management is a valid approach for selected pediatric patients with JO, particularly when hearing is stable and quality of life is unaffected. Continued audiologic surveillance is essential, both to track disease progression and to identify the appropriate timing for intervention, should it become necessary.

Conclusion

This case highlights that individualized, conservative management is a reasonable and effective approach in pediatric patients with juvenile otosclerosis who present with stable hearing and minimal functional impairment. While stapedotomy remains a valuable treatment option for progressive or severe cases, the potential risks of surgery and challenges related to postoperative care in children necessitate careful patient selection. Comprehensive audiological monitoring, detailed imaging, and shared decision-making with patients and their families are essential components of optimal care. Further prospective studies with long-term follow-up are needed to better understand the natural history of juvenile otosclerosis and to establish evidence-based guidelines for its management in children.

Informed Consent

Informed written consent for publication of clinical details and accompanying images was obtained from the patient's legally authorized representative.

Declaration of Conflicting Interests

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