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Case Report

Congenital Agenesis of the Stapes: A Rare Cause of Conductive Hearing Loss

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Abstract

Introduction: Congenital agenesis of the stapes is a rare cause of conductive hearing loss. Computed tomography plays an essential role in the positive diagnosis and in the assessment of associated malformations.

Case Report: We report the case of left stapes agenesis revealed by congenital conductive hearing loss in a 3-year-old girl.

Discussion: Congenital agenesis of the stapes is a rare congenital malformation. It is often associated with malformations of the facial nerve canal and the ossicular chain. Computed tomography allows a detailed assessment of the inner ear and middle ear, providing a non-invasive alternative to exploratory tympanotomy. Treatment is based on bone-conduction hearing devices or surgical reconstruction.

Conclusion: Computed tomography plays a key role in the diagnosis of stapes agenesis and associated malformations, allowing better therapeutic planning.

Keywords: Congenital Agenesis; Computed Tomography

Introduction

Congenital agenesis of the stapes is a rare congenital malformation revealed by conductive hearing loss. It is often associated with abnormalities of the facial nerve canal and the ossicular chain. Computed tomography plays a crucial role in the positive diagnosis of stapes agenesis and associated malformations [1-3].

We report the case of left stapes agenesis revealed by conductive hearing loss.

Clinical Observation

A 3-year-old girl presented with her parents for evaluation of left-sided hearing loss present since birth. Clinical examination showed normal external auditory canals and tympanic membranes. Auditory evoked potentials demonstrated left-sided deafness.

Computed tomography of the temporal bones revealed left stapes agenesis (Figures 1 and 2).

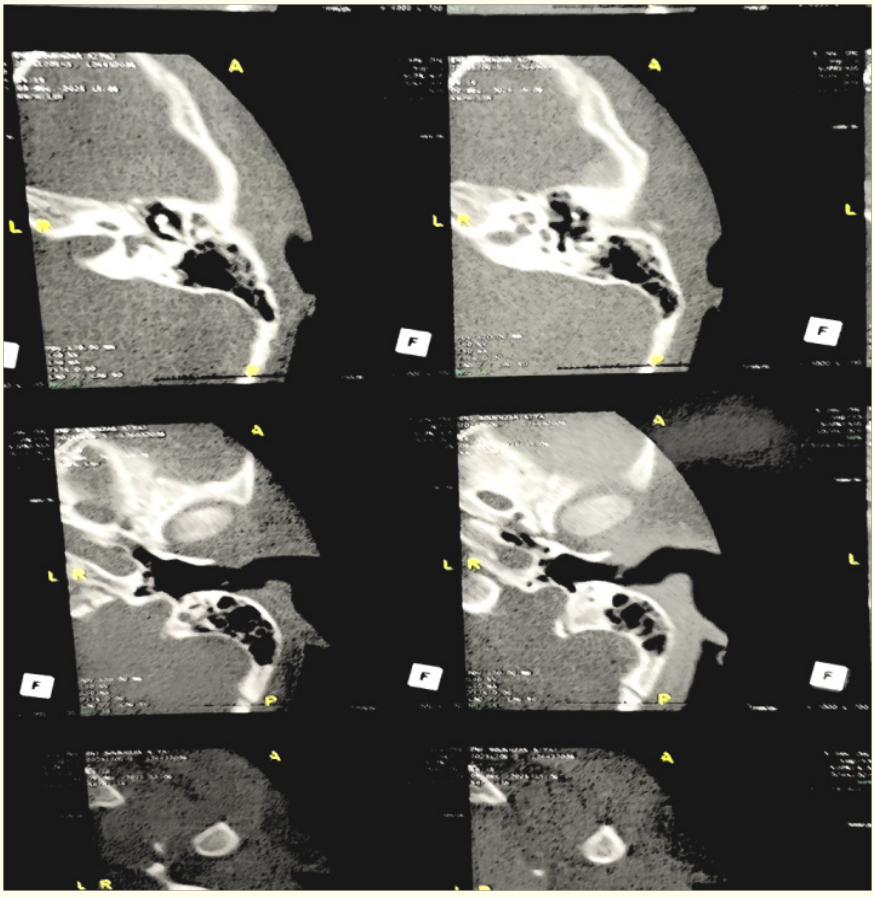

Figure 1: Computed tomography of the left temporal bone. Axial reconstruction showing agenesis of the stapes.

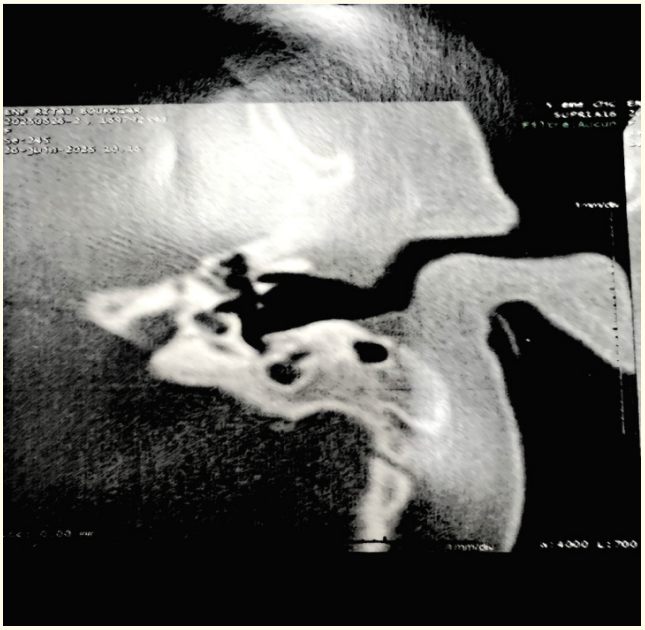

Figure 2: Computed tomography of the left temporal bone. Coronal reconstruction showing agenesis of the stapes.

The patient was eligible for surgical treatment given the absence of malformation of the oval window. The proposed treatment was placement of a piston via an endaural approach under general anesthesia (Figure 3).

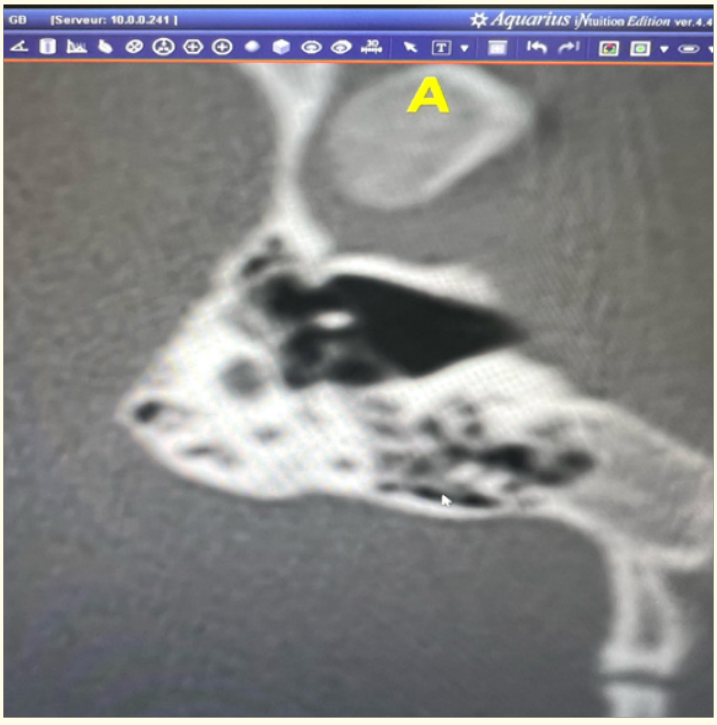

Figure 3: Postoperative axial CT scan of the left temporal bone demonstrating a piston in place.

Discussion

Congenital agenesis of the stapes is a rare malformation responsible for conductive hearing loss, with no progressive character and no history of otitis. Agenesis may occur in isolation or be associated with Turner syndrome or CHARGE syndrome (coloboma, heart defect, choanal atresia, retarded growth and development, genital hypoplasia, ear anomalies/deafness) [4]. The most widely accepted theory is anterior displacement during development of the facial nerve, which interposes between the otic capsule and the stapes blastema during the 5th and 6th weeks of gestation, preventing the contact required for oval window development. The consequence is oval window agenesis and stapes malformation [3].

Computed tomography constitutes a non-invasive alternative to exploratory tympanotomy by allowing detailed evaluation of the inner ear and middle ear, including the ossicular chain, with good correlation with surgical findings. CT therefore plays a crucial preoperative role in therapeutic decision-making [4].

Exploration of the middle ear remains the gold standard for evaluating middle ear malformations and for confirming CT findings to verify the feasibility of surgical treatment [4].

On computed tomography, stapes agenesis appears as complete absence resulting in ossicular chain discontinuity. The oval window may be normal, absent, or dysplastic. Several ossicular or facial nerve canal anomalies may be associated [1]:

- **Stapes:** Posterior displacement toward the pyramidal eminence, absence of the head, neck, and crura, absence of the footplate, rudimentary appearance, loop-shaped, amorphous, or incorporated into the facial nerve canal;
- **Incus:** Fixation to the epitympanic wall, absence of the long process and lenticular process, shortening or curvature of the long process, incudomalleolar fusion;
- **Facial nerve canal:** Total or partial overhang of the oval window, dehiscence, partial absence, bifurcation, hypoplasia, and persistence of the stapedial artery. Coronal reconstructions better identify facial nerve canal and oval window anomalies [1].

Treatment is mainly based on bone-conduction hearing devices, transcutaneous or percutaneous, or surgical reconstruction. The latter consists of performing a vestibulotomy above the presumed site of the oval window, followed by placement of a piston. However, reconstruction is contraindicated in cases of low position of the tympanic segment of the facial nerve covering the oval window, total absence of the oval window, or absence of the round window. In addition, prosthesis placement requires an intact incus with an appropriate distance between the incus and the vestibulotomy site [4].

Conclusion

Congenital stapes agenesis is a rare malformation. Computed tomography plays a key role in its diagnosis and in the identification of associated middle ear malformations, allowing selection of patients eligible for surgery. Bone-conduction hearing devices constitute a good alternative to surgery when it is contraindicated.

Protection of Human and Animal Subjects

The authors declare that the conduct of the research described in this article is in accordance with the revised 2013 Declaration of Helsinki of the World Medical Association concerning experimentation on patients or subjects, as well as with European Directive 2010/63/EU concerning animal experimentation.

Informed Consent and Data Confidentiality

The authors declare that the article contains no personal data that could identify the patient or subject. The authors declare that written informed consent was obtained from the patients and/or subjects referred to in this article. The authors also declare that all personal details of the patient(s) and/or volunteers have been removed.

Declaration of Conflicts of Interest

The authors declare that they have no conflicts of interest.

Bibliography

1. Thomeer H., *et al*. "Congenital oval or round window anomaly with or without abnormal facial nerve course: surgical results for 15 ears". *Otology and Neurotology* 33 (2012): 779-784.

2. Teunissen EB and Cremers WR. "Classification of congenital middle ear anomalies. Report on 144 ears". *Annals of Otology, Rhinology and Laryngology* 102 (1993): 606-612.

3. Zeifer B., *et al*. "Congenital absence of the oval window: radiologic diagnosis and associated anomalies". *AJNR American Journal of Neuroradiology* 21 (2000): 322-327.

4. Khoo HW., *et al*. "High Resolution Computed Tomography (HRCT) Imaging Findings of Oval Window Atresia with Surgical Correlation". *Annals Academy of Medicine, Singapore* 49 (2020): 346-353.