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Stapes Agenesis and Oval Window Atresia with Anomalous Facial Nerve Path: Case Report

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Abstract

Introduction: Middle ear malformations are considered the main causes of hearing loss, and both the origins and the correlation of middle ear structures are not yet fully understood. **Objectives and Methods:** description of a tomographic study of a case of bilateral hearing loss. **Results and Discussions:** The study shows the absence of stapes with bilateral oval window atresia, associated with facial nerves with anomalous paths, medialized and positioned adjacent to the expected site for the oval windows. **Conclusion:** This study demonstrated a possible relationship between the embryogenesis of the stapes and its positioning in the proper formation of the oval window, as well as the correlation of these structures with the anatomical path of the facial nerve through the topography of the middle ear.

Keywords

Middle Ear, Hearing Loss, Stapes, Oval Window, Facial Nerve, Computed Tomography

1. Introduction

The sensitive balance that makes hearing possible depends on the smallest and lightest bone in the human body, the stapes, and its relationship with the oval window, which is responsible for connecting the middle ear to the inner ear. Even minimal changes in these structures can result in conductive hearing loss, which is one of the most common forms of hearing loss [1].

The stapes' movement is limited by the stapedius muscle, which is innervated by a branch of the facial nerve [2]. When exposed to excessive sound stimulation,

the muscle contracts and limits the ossicle's movement, thus protecting adjacent structures and the organ of Corti from potential trauma, in addition to preventing hyperacusis.

Although their origins are not yet fully understood, it is known that these structures have different embryological origins, with the stapes and facial nerve originating from the second brachial arch and the oval window from the otic capsule [3]. It is noted that the development of the oval window depends on stimuli originating from the stapes, and the concomitant absence of these structures has been noted in several studies [4], most frequently observing its correlation with syndromes such as Klippel-Feil, Crouzon, or Pfeiffer syndrome [5] [6].

Minimal changes in this arrangement will cause changes in the individual's hearing, and, as seen, the existence of these structures and their positions depend on each other. The triad of stapes agenesis with oval window atresia and facial nerve presenting an anomalous course is a rare and underreported condition, and is analyzed in this study using CT images (the main method for analyzing these structures [7] [8]).

2. Objectives and Methods

To describe the Computed Tomography imaging findings of a rare case of absence of stapes with bilateral oval window atresia, associated with facial nerves with anomalous paths, medialized and positioned adjacent to the expected site for the oval windows.

This study aims solely to document the tomographic images, without focusing on the patient's previous clinical history, which was not available.

The images were analyzed without prior knowledge of middle ear syndromes or anomalies, and without considering otoscopy or audiometry results (which were not available). The evaluation was performed as a routine study to rule out structural causes that could cause bilateral hearing loss.

3. Results and Discussion

This study documents the images obtained in the mastoid study of a 28-year-old male patient who is investigating the cause of his bilateral hearing loss over the last fifteen years.

In the analysis of the external ear, no local changes or malformations were observed, nor in adjacent regions (e.g, **Figure 1**).

The middle ear examination revealed atresia of the oval window, with only a small, rudimentary formation in its usual topography, and a thickening of the bony plate adjacent to the round window. Furthermore, the stapes was not characterized, and there were no changes in the other ossicles (malleus and incus) on the right, indicating isolated agenesis of this structure, while hypoplasia of the long process of the incus was noted on the left (e.g, **Figures 2-4**).

In the topography studied, a medialization of the facial nerves was observed in their horizontal portions bilaterally (e.g, **Figure 5** and **Figure 6**), with duplication

only on the right (e.g, Figure 2 and Figure 3). The medialized course of the facial nerve, more precisely between the long branch of the incus and the atretic oval window (e.g, Figure 6), may represent an additional challenge for the surgical intervention in this region and could contraindicate certain approaches.

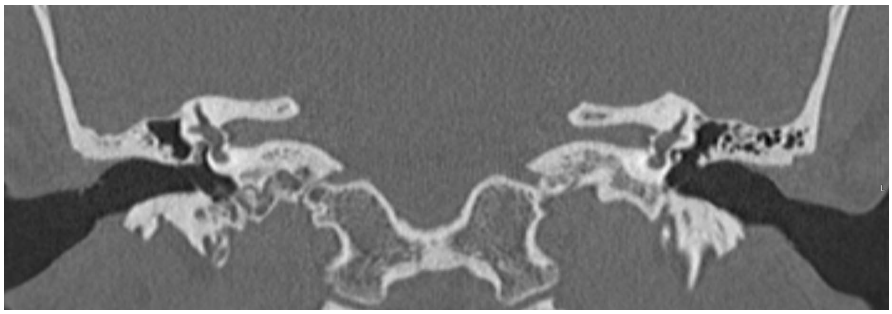

Figure 1. The bilateral coronal image of the mastoid study showed no evidence of changes in the external ear and adjacent structures.

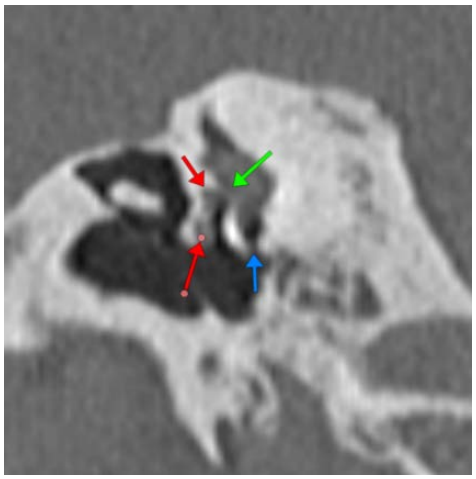

Figure 2. The right ear, coronal section, shows atresia of the oval window (green arrow) with agenesis of the stapes and duplicated facial nerve (red arrows), showing superior and inferior divisions with an anomalous path. Anatomical round window (blue arrow).

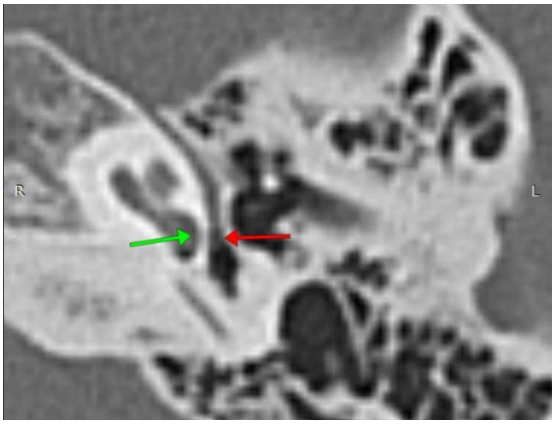

Figure 3. Left ear, axial section. Green arrow indicating the atretic oval window. Red arrow indicating the path of the facial nerve without bifurcation.

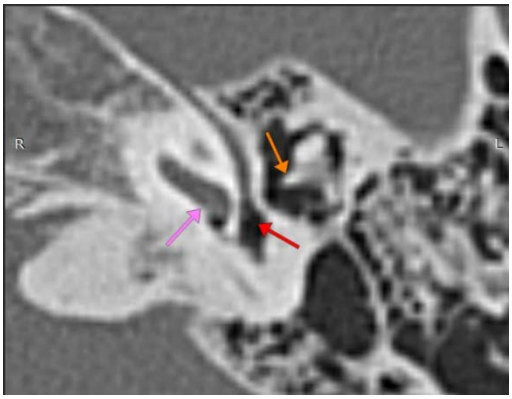

Figure 4. Axial section of the left ear. The hypoplastic long process of the incus is indicated by the orange arrow. The red arrow indicates the path of the medialized facial nerve. The pink arrow indicates the basal turn of the cochlea.

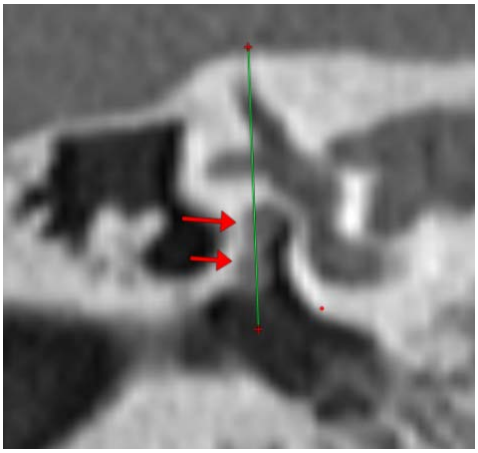

Figure 5. Right ear, coronal section, demonstrating medialization of the duplicated facial nerve (indicated by arrows). Vertical line delimiting the usual limits of the facial nerve canal, which should be positioned lateral to this vertical line at the level of the superior semicircular canal.

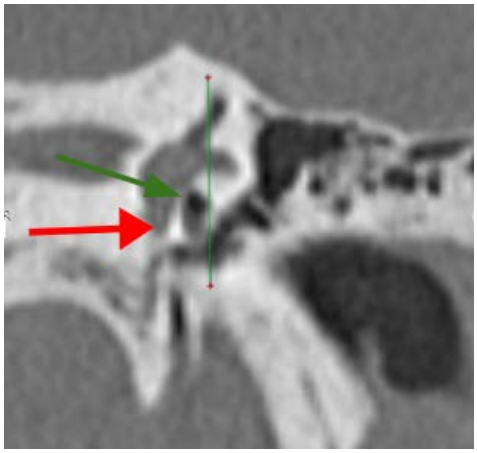

Figure 6. Left ear, coronal section. Green line demonstrating medialization of the facial nerve path (red arrow) in relation to the level of the superior semicircular canal and again the atretic oval window (green arrow).

4. Conclusion

Although structural alterations in the ear are one of the most common causes of hearing loss, the origin of the structures responsible for conducting sound stimuli is still not fully understood. This case highlights the possible relationship between the stapes and its positioning in the proper formation of the oval window, as well as the correlation of these structures with the proper/usual path of the facial nerve in the middle ear. Further studies are needed to clarify the cause-and-effect relationship between these malformations.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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