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A Case of External Auditory Canal Atresia

Lipika Gupta[1], Archit Gupta[2]

[1]Department of Radiodiagnosis, NEIGRIHMS, Shillong; Email id - guptalipika94[at]gmail.com

[2] Assistant Professor, Department of Radiodiagnosis, SNMC, Agra, Uttar Pradesh; Email id - architgrmc[at]gmail.com
Corresponding author Email id: *architgrmc[at]gmail.com*

Abstract: ***This article presents a case study of a young boy diagnosed with External Auditory Canal Atresia (EACA), a rare congenital disorder. The patient exhibited conductive hearing loss and other associated abnormalities. High - resolution computed tomography (HRCT) was used for evaluation, revealing EAC atresia and other anomalies. The study underscores the importance of systematic evaluation of ear structures and the role of HRCT in diagnosing EACA.***

Keywords: Conductive hearing loss; External auditory canal atresia; High - resolution CT

1. Introduction

External auditory canal atresia (EACA) occurs in 1 in 10, 000–20, 000 births and is bilateral in approximately one third of the cases (1). Patients with EACA experience conductive hearing loss. Surgical restoration of conductive hearing can be achieved provided there is intact sensorineural hearing.

Malformations of the external ear are also known as congenital aural dysplasia's. Bilateral malformationsare possible and may result fromchromosomal aberrations, genetic disorders and exposure to teratogens. EAC atresia may be part of a congenital syndromes such as Goldenhar syndrome, Treacher Collins syndrome, or Pierre Robin sequence (2).

The external ear is an osseous - cartilaginous structure that extends from the auricle to the tympanic membrane. It is divided into two parts: the auricle (or pinna) and the external auditory canal (EAC). The auricle is derived from the first and second pharyngeal arches, and the external auditory canal is derived from the first pharyngeal cleft. The malleus and incus are derived from endochondral ossification of the cartilage of the first and second arches (3). Hence, abnormal development of the first and second arches results in auricular malformations, including microtia, external auditory canal stenosis, and ossicular dysplasia. The early identification of unilateral conductive hearing loss can be sometimes be difficult in the absence or presence of minor deformities of the external ear (4).

2. Case Report

We present the case of a 10 yearold male patient with right sided conductive hearing loss. Overall speech and perception skills were good. However, the hearing loss was progressive and patient was facing difficulties with language courses in school. There was no family history of similar problem. No associated disorders were present.

For further evaluation high - resolution computed tomography (HRCT) of the temporal bones was performed. Thin (0.6 mm) sections were acquired. HRCT showed right sided complete atresia of the external auditory canal (Figure 1). The ipsilateral auricle was also malformed (Figure 2a & b). The malleus was found to be fused with the atretic plate (Figure 3). The rightincudomallear articulation, stapes and inner ear structures were normal (Figure 4 - 7). Left auricle, external auditory canal, the middle and inner ear structures were normal. Based on the HRCT findings, the patient was diagnosed with right complete external auditory canal atresia.

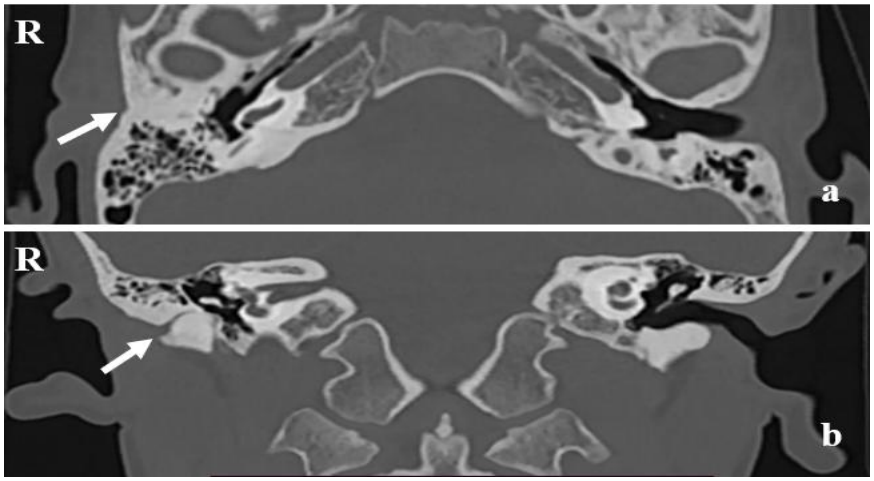

Figure 1: Axial (a) and coronal (b) sections of HRCT temporal bone showing right external auditory canal atresia (arrows)

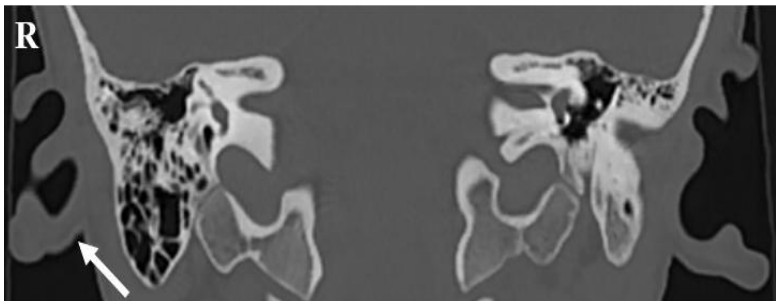

Figure 2a: Coronal thin section HRCT temporal bone showing malformed right auricle (arrow)

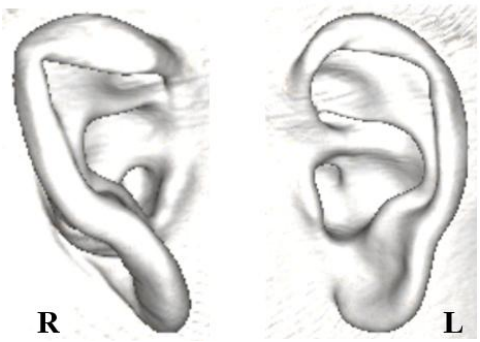

Figure 2b: Reconstructed VR images showing malformed right auricle

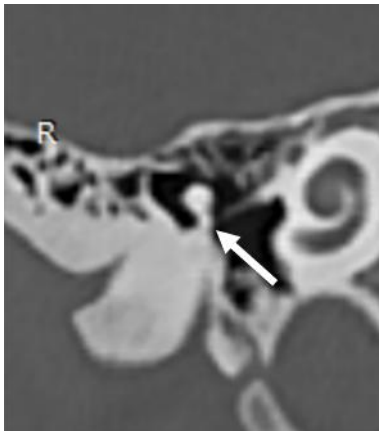

Figure 3: Coronal thin section HRCT temporal bone showing malleus fused with the atretic plate (arrow)

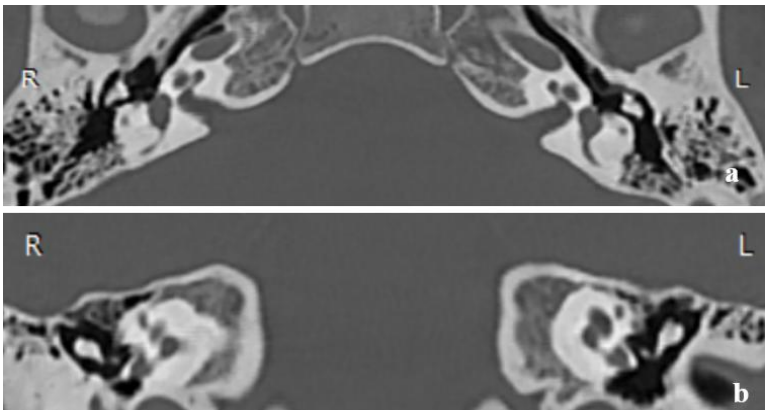

Figure 4: Axial (a) and coronal (b) thin section HRCT temporal bone showing normal incudomallear articulation in both middle ears

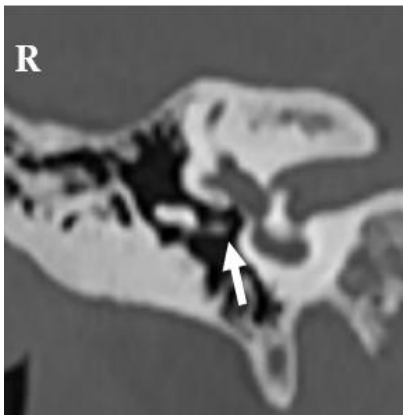

Figure 5: Coronal thin section HRCT temporal bone showing normal right stapes (arrow)

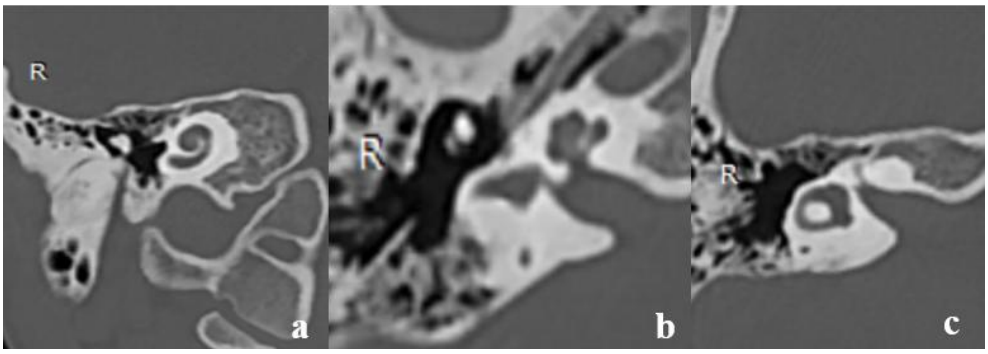

Figure 6: Coronal (a) and axial (b, c) thin section HRCT temporal bone showing normal formation of the right inner ear structures and internal acoustic meatus

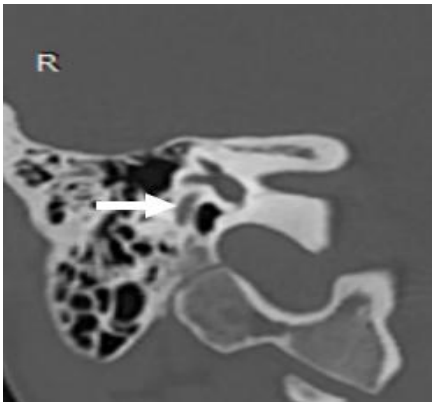

Figure 7: Coronal thin section HRCT showing normal mastoidsegment of the right facial nerve (arrow)

3. Conclusion

Patients presenting with unilateral conductive hearing loss should be evaluated for EAC stenosis and atresia with or without characteristic ossicular malformations. Anomalies of the pinna are commonly present in this group of patients (5). Systematic evaluation of the external ear, middle ear and inner ear structures is suggested with the help of imaging. Temporal bone HRCT findings are used to select suitable candidates for surgery, to plan the surgery, and to predict the postoperative hearing outcome.

Conflicts of interest: The authors declare that they have no conflicts of interest

References

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