

Congenital Auditory Atresia and Malleus Incus Deformity

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ABSTRACT

External auditory canal atresia, or congenital auditory atresia, is a birth disease marked by underdevelopment of the external auditory canal. This condition frequently leads to abnormalities in the auricle and middle ear structures, and occasionally affects the inner ear structures. It is important to note that the structures of the middle-inner ear may be impacted, although infrequently, in individuals with atresia of the external auditory canal, as was the case with our patient. Furthermore, it is crucial to emphasize the importance of early diagnosis, as it can significantly enhance the patient's quality of life and expand their options for treatment.

Keywords: Congenital auditory atresia, external auditory canal atresia, incus, malleus

INTRODUCTION

External auditory canal atresia, also known as congenital auditory atresia, is a congenital defect that is characterized by hypoplasia of the external auditory canal and often causes deformities in the auricle and middle ear structures, and rarely in the inner ear structures. In this case, deformities in the middle ear structures, which are known to be rarely affected by congenital auditory atresia, are shown.

CASE PRESENTATION

Our 12-year-old male patient was known to have congenital auditory atresia on the right side. However, he did not have any auditory complaints until today. Hearing screening performed at school revealed unilateral conductive hearing loss. No abnormality was detected in the subsequent otolaryngological examinations and systemic and neurological examinations. Since temporal computed tomography (CT) can show the severity of the auditory atresia and the extent to which it affects the middle ear, inner ear structures, and ossicles, a temporal CT scan was performed to evaluate these regions.

In the temporal CT examination of our patient, it is observed that the right auricle is atresia and the right external auditory canal is hypoplastic (Figure 1-2). In addition to the previously known atresia of the auricle and external auditory canal, dysmorphic appearances were observed in the malleus and incus of the middle ear. It was observed that the malleus and incus were fused as a single bone. Left auricle, outer inner and middle ear structures are in natural appearance (Figure 3).

It was thought that auricular atresia, external auditory canal hypoplasia, and malleus-incusdysmorphic appearances that were understood to develop secondary to these, and the fusion structure of the malleus and incus as a single bone caused conductive hearing loss. Due to the fact that the fusion appearance of the malleus and incus has meaning in terms of postoperative results, the necessary information was provided by communicating with the otologic surgeon. Our patient was referred to the otorhinolaryngology clinic so that the malleus, incus, and stapes ossicles could be evaluated for surgical prosthesis. Surgical operations were performed in our patient who was referred to an otologic surgeon. In the following process, a 20 dB hearing gain was achieved in our patient.



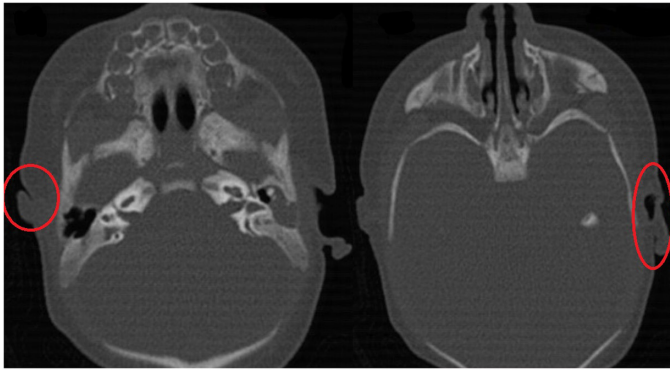


Figure 1. (A, B) While the normal auricle structure is seen on the left, the right auricle has an atresia appearance.

DISCUSSION

Congenital auditory atresia is a congenital ear deformity involving hypoplasia of the external auditory canal. Most often malformation includes, to varying degrees, the eardrum, ossicles, and middle ear cavity. Although associated abnormalities of the auricle are common, the inner-middle ear development of these cases is mostly normal.¹

Malleus was found to be the most affected ossicle in patients whose middle ear structures were affected. The malleus and incus usually appear as a single, fused

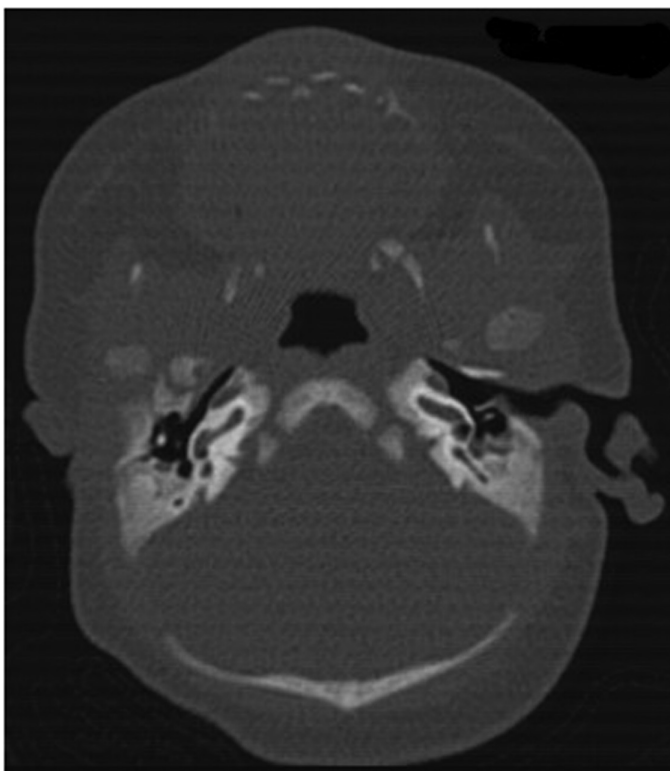


Figure 2. While the left external and middle ear canal appears normal, the right external ear canal and middle ear are atresia.

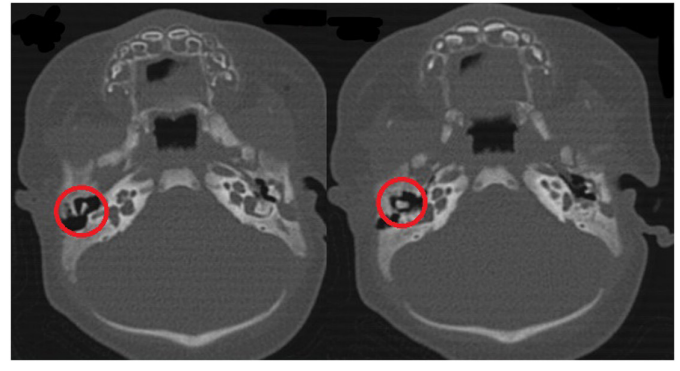


Figure 3. (A, B): Middle ear ossicles with a dysmorphic appearance, malleus and incus with a fused appearance are observed.

bone structure. It is known that the stapes and the includostapedial joint are less frequently affected.² The definition that the malleus and incus in the middle ear fuse and appear as a single bone in patients with congenital auditory atresia has implications for the otologic surgeon and possible postoperative hearing outcomes.³

This malformation may be associated with other congenital anomalies and may manifest in different syndromes, but mostly occurs sporadically. It has been reported that the disease is associated with hydrocephalus, posterior cranial hypoplasia, hemifacial microsomia, cleft palate, and genitourinary abnormalities. Congenital auditory atresia can be part of different syndromal abnormalities such as Treacher Collins, Goldenhar, Crouzon, Mobius, Klippel-Feil, Fanconi, DiGeorge, and Pierre Robin syndromes.^{3,4}

The incidence of congenital auditory atresia is approximately 1 in 10 000 to 20 000 live births.⁵ Unilateral atresia is 3-5 times more common than bilateral atresia. Men are affected more often than women, and the right ear is more frequently involved in unilateral cases.³⁻⁶

Although it was known that the external auditory canal atresia was congenital in our patient, it was understood that the middle ear structures were also affected by the process when the patient was 12 years old. It should be kept in mind that the middle-inner ear structures may be affected, albeit rarely, in patients with external auditory canal atresia, as in our patient. In addition, awareness should be raised that early diagnosis will increase the patient's comfort of life and treatment opportunities.

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