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Olgu Sunumu / Case Report

Bilateral Incus Body Pneumatization

Bilateral İnkus Gövdesinde Pnömotizasyon

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Abstract

Embryological development of middle ear ossicles is a complex process and congenital abnormalities and variations are very rare. Therefore, adding new knowledge to this conditions is highly important. This case report describes a rare case of variation consisting of bilateral pneumatization in the incus body in a patient with left-sided mixed-type hearing loss. These incus variations were detected incidentally during the examination performed for an otological problem. This paper discusses the possible embryological mechanism for the occurrence of this rare incus variation and its significance.

Keywords: middle ear, variation, middle ear ossicles, incus, computed tomography

Özet

Orta kulak kemikçiklerinin embriyolojik gelişimi karmaşık bir süreçtir ve konjenital anormallikler ve varyasyonlar çok nadirdir. Dolayısıyla bu durumda yeni bilgilerin eklenmesi son derece önemlidir. Sol tarafta mikst tip işitme kaybı yakınması ile başvuran her iki inkus gövdesinde pnömotizasyon şeklinde nadir bir varyasyonu bulunan olgu sunulmaktadır. Bu inkus varyasyonu otolojik problem için yapılan muayenede tesadüfen tespit edildi. Bu yazı, bu nadir inkus varyasyonunun oluşumu için olası embriyolojik mekanizmayı ve önemini tartışmaktadır.

Anahtar Kelimeler: orta kulak, varyasyon, orta kulak kemikçikleri, inkus, bilgisayarlı tomografi

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1. Introduction

The malleus, incus and stapes are the smallest bones in the body. They are arranged in the middle ear in this order and ensure that the sound is transmitted mechanically from the outer ear to the inner ear. The incidence of ear malformations is approximately 1 per 15 000 in newborns1. Goldenhar syndrome, Treacher Collins syndrome, branchio-auto-renal syndrome, prenatal infection, and drug use during pregnancy are associated with malformation in the ossicles2. Isolated congenital ossicular malformations are even rarer1. Stapes malformation is reported to be the most common ossicular anomaly, while malformations including incus are among the rarest cases described3. In the postpartum period, cholesteatoma, chronic otitis media, and trauma may also cause ossicular deformity4.

In this report, we present a patient, who, rather than having a malformation or deformity, had pneumatization in the incus body, which can be defined as a normal developmental variation. In a recently published case report, a similar appearance located at the incus body was reported, however it was evaluated as pneumatization5. To our knowledge, the literature contains no other similar case. The aim of publishing this case report was to ensure that this appearance, whether defined as a pneumatization, is known as a variation and no invasive procedure is performed in patients with this variation.

2. Case report

A 27-year-old male patient presented to the hospital with progressive left-sided hearing loss that had started 18 months earlier. The patient had no other symptoms, such as otalgia and ear discharge. The otoscopic examination showed a normal ear canal and intact tympanic membrane. There was no history of ear disease, otologic surgery, significant head trauma, or family history of hearing loss. The audiogram revealed a left-sided moderate mixed-type hearing loss.

Further investigation was performed with high-resolution computed tomography (HRCT) of the temporal bone, which showed no otological pathology that would indicate the reason for the hearing loss. However, it demonstrated bilateral pneumatization of incus bodies (Figure). No personal information of the patient were used in the case report. Therefore no consent was obtained.

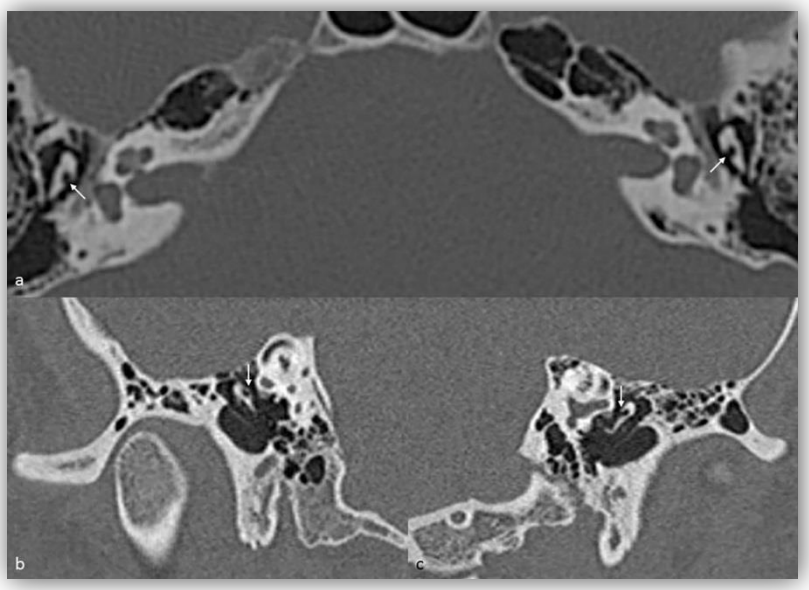

Figure. Axial (a) and coronal oblique (b and c) reconstructed high-resolution computed tomographic images showing bilateral cleft formation in the incus body (arrows).

3. Discussion

In a study in which the variations and clinical significance of middle ear ossicles were evaluated, the most frequent variation was found in the stapes followed by the malleus, and the least variation was observed in the incus. In the incus, the triangular shape of the long process accompanying the small body and a small notch in the short process were detected6. In a case report of Bhatt et al.5, pneumatization was observed in the incus body, as in the current case. The authors considered that this pneumatization detected in the incus was most likely a developmental variation5.

The embryological development of the middle ear is a complex process. The skeletal elements of the middle ear have been claimed to develop from the mesenchyme of the first two branchial arches. Although many different hypotheses have been postulated, in general it is well accepted that the first branchial arch forms the bodies of the malleus and incus above the neck of these ossicles, while the second branchial arch forms the parts of the ossicles below the neck of the malleus and the body of the incus, including the crura of the stapes, which grow to merge with the stapes footplate, the lenticular process and long process of the incus, and the manubrium of the malleus, which is induced separately. With further development, the ossicles first separate from the remainder of the arch cartilages, and then join to form the ossicular chain. The cartilaginous ossicles grow only throughout the first half of intrauterine life, and then ossify, each from a single center. The center for the incus appears at the 16th fetal week, the center for the malleus at the mid-16th fetal week, and that of the stapes at the 18th fetal week7.

While there is chondrogenesis in the ossicles, the entire middle ear is filled with soft tissue consisting of the mesenchyme derived from the neural crest and a single layer of endodermal epithelium that runs along the inner surface of the tympanic membrane and along the ventral areas of the soon-to-emerge middle ear cavity. This mesenchyme regresses and leaves the ossicles suspended in air8. It can be considered that the small airspaces in the primitive bone marrow cavities in the incus and malleus allow for the migration of the mesenchyme to the ossicles, thus leaving a small pneumatic air void after regression. Microscopic examination reveals pneumatization in 0.2% of the 1500 temporal bones in the body and long process of the incus5’9. This mechanism may explain the formation of the pneumatization seen in the incus. The observed variation being bilateral also supports the idea that it is a developmental variation.

A pneumatized incus, which was previously described only in one other case report, should not be treated incorrectly as a defect. If the middle ear is to be operated on for other purposes, the surgeon should be warned of the existence of this variation.

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