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Congenital Incus Anomaly: Any Role of HRCT As Single Diagnostic Tool?

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Abstract

We describe a rare case of isolated congenital incus anomaly without other otologic anomalies in this case report. We detected this condition in an adult who presented with long standing, non progressive unilateral reduced hearing. Isolated congenital incus anomaly was identified incidentally during her visit for other otological problem. We discussed regarding the case and the important role of HRCT imaging as a single tool for diagnosing the anomaly without surgical exploration.

Keywords: congenital, hearing loss, ossicle, ossicle anomaly, incus anomaly, HRCT.

Introduction

Ossicular anomalies with conductive deficit commonly associated with external auditory canal dysplasia. Stapes malformation is reported to be the most common ossicular anomaly. In this case report, we describe a rare case of isolated unilateral incus anomaly without other otologic manifestation which was found in a female patient who presented with long standing non progressive unilateral hearing loss.



Figure 1: prominent handle of malleus, body and long process of incus

Case report

We report a case of 38 years old female presented with long standing and non progressive hearing loss of left ear over 26 years. She only noticed the hearing loss when she was 12 years old. There was no history of otitis media or trauma. There is no significant antenatal, perinatal and birth history.

Examination revealed normal bilateral pinna and ear canal. Otoscopic examination of left ear showed a retracted tympanic membrane with prominent lateral process of malleus, body and long process of incus (Figure 1). There was no clinical evidence of cholesteatoma seen. Rinne's and Weber test consistent with left conductive hearing loss. Tympanometry test revealed a bilateral type A tympanogram (Figure 2) while her serial Pure Tone Audiogram revealed left conductive hearing loss with air bone gap of 30-50dB (Figure 3). The opposite ear has normal examination finding and normal hearing level.

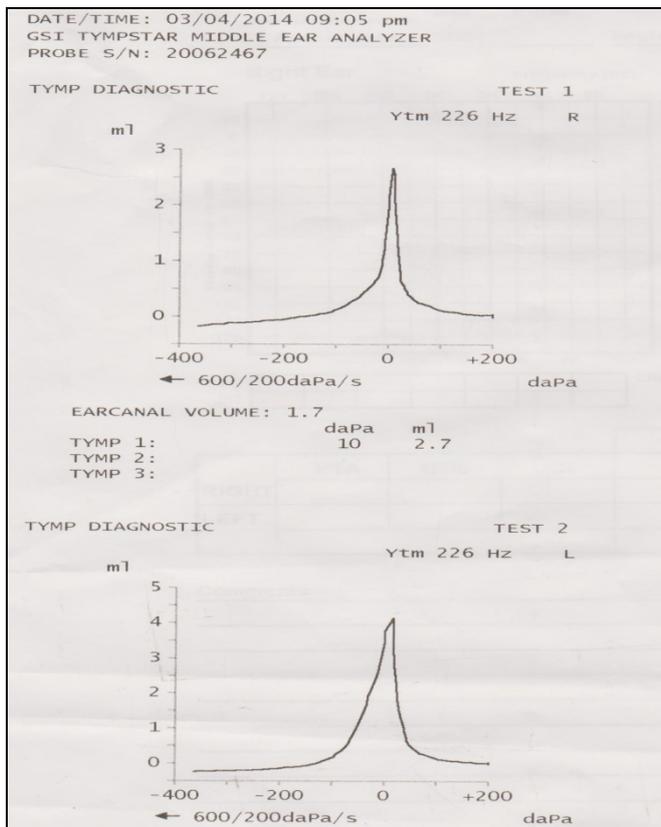


Figure 2: Tympanogram: Bilateral Type A

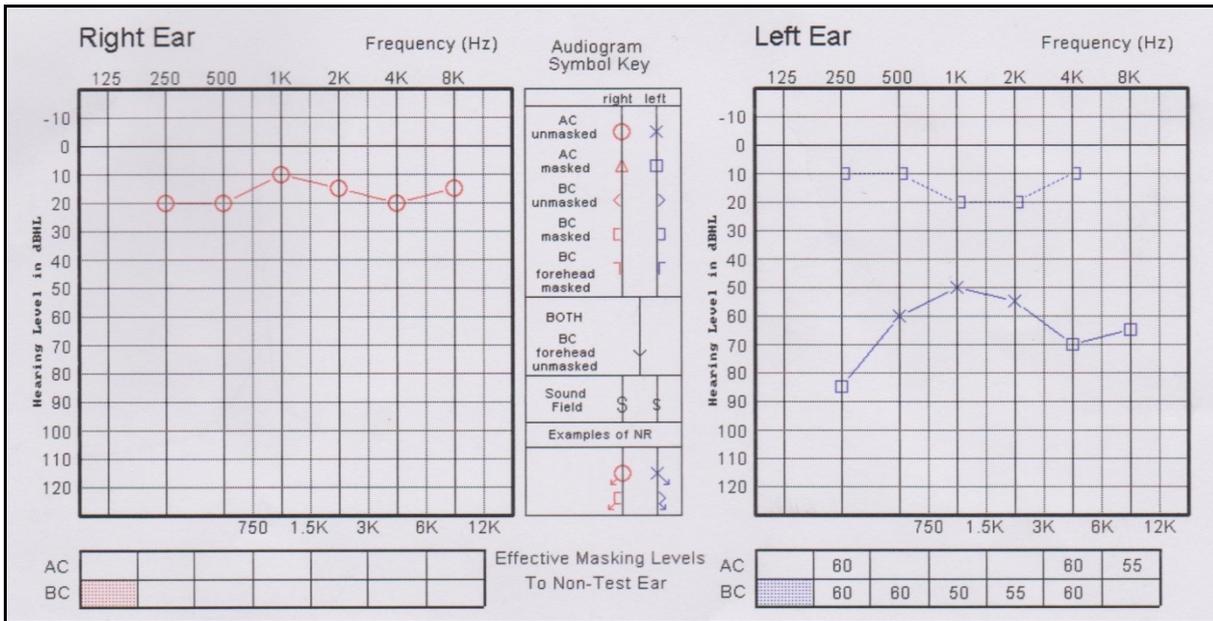


Figure 3: Pure Tone Audiogram showed left conductive hearing loss with air-bone gap (ABG) of 30-50dB

Further investigation was performed with High Resolution Computed Tomography (HRCT) of temporal which showed a dislocated left incudo-malleolar and incudo-stapedial joint with evidence of incus anomaly of left ear. Stapes and malleus appeared normal characteristically. There was no evidence of canal or middle ear cholesteatoma. The HRCT finding could explain her clinical symptoms, clinical finding and audiological examination result.

Hearing level was monitored in the past 6 years and it remained the same. She was offered surgical ossicles reconstruction with prosthesis, which has the possibility to have a good outcome. However, patient refused surgery and opted for conservative treatment.

Discussion

Congenital ossicular anomalies resulting in conductive deficit commonly associated with external auditory canal dysplasia while it is less common without any external ear anomalies. Stapes malformation is reported to be the most common ossicular anomaly. A unilateral absence of incus without other otologic manifestation is a rare entity [1]. Study by Swartz and Faerber in Medical College and Hospital of Pennsylvania, 4 out of 8 patients with isolated congenital ossicular deformity cases, the anomaly was bilateral. Stapes is the most common ossicle involved and was abnormal in all cases of isolated deformity. Numerous anomalies are possible and bilaterality is common [5]. The conductive hearing loss with absence of pinna and external auditory canal anomaly can also be caused by other conditions such as osteogenesis imperfecta, branchio-otorenal syndrome, orofaciogigital type II syndrome and Treachers Collins Syndrome [4].

A non-progressive and conductive hearing loss with a normal tympanic membrane, without history of trauma or middle ear infection, is highly suggestive of a congenital ossicular anomaly. In this case, her late presentation is likely due to her hearing disability being masked by good hearing of the opposite ear until it was detected incidentally. In ossicular disruption, PTA (Pure Tone Audiometry) usually demonstrates a large air bone gap and tympanometry may demonstrate an axis deviation curve (type Ad) [2, 3]. In our case her serial PTA corresponds well with the expected finding except her tympanometry showed normal type A bilaterally.

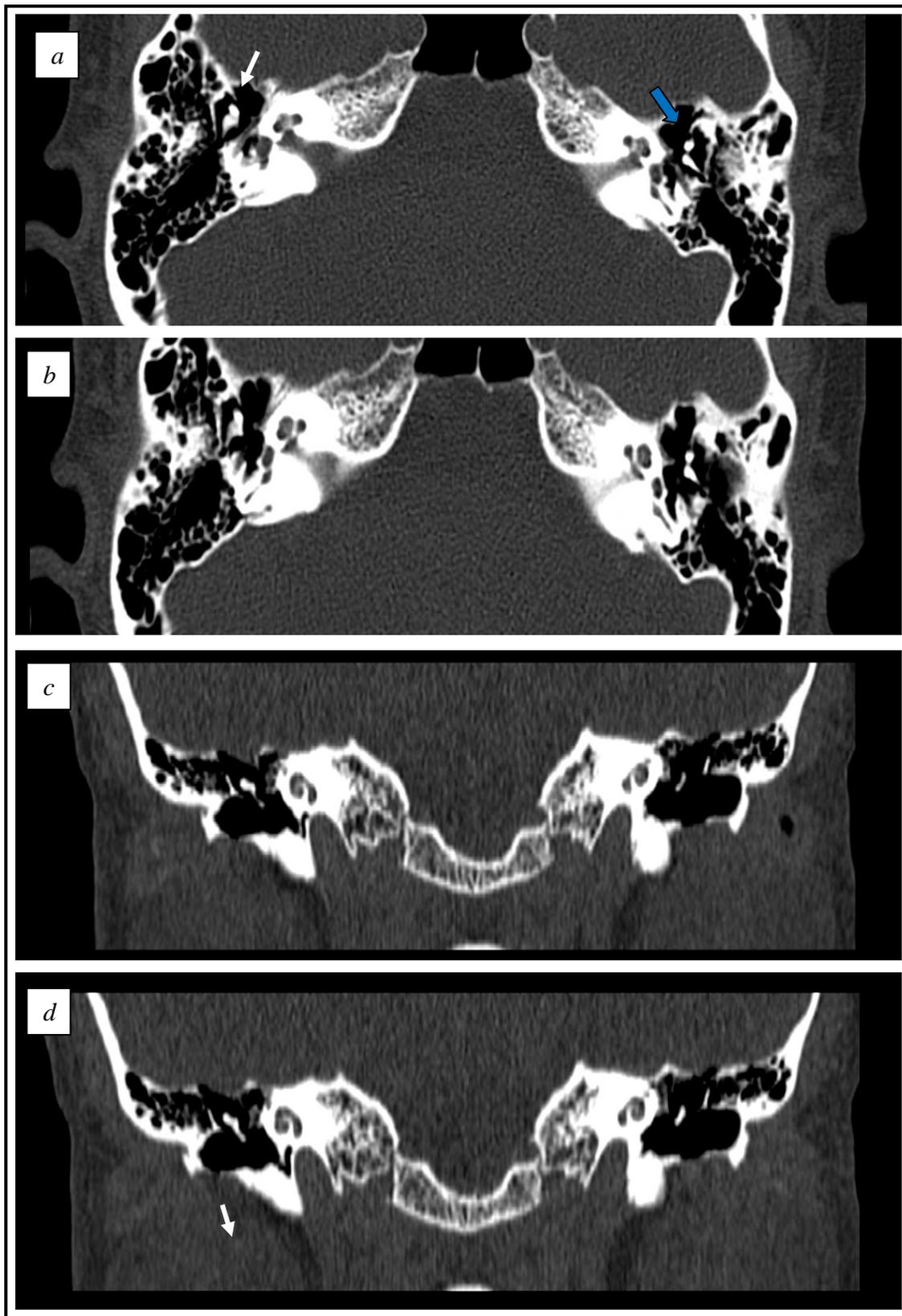


Figure 3: a and b are axial views of HRCT temporal of the patient. In a, 'ice cream' cone configuration (white arrow) is seen in right temporal represent normal head of malleus and incus while on the left temporal, there is separation of malleus and incus. The incus is seen dislocated and displaced (blue arrow). This finding can be compared with the normal incus in right temporal .c and d are coronal views of HRCT temporal. At this plane, right head of malleus and long process of incus seen in its normal anatomical position (arrow). Left long process of incus more prominent in this plane and head of malleus not visualized likely they are detached and long process of incus is displaced posteriorly.

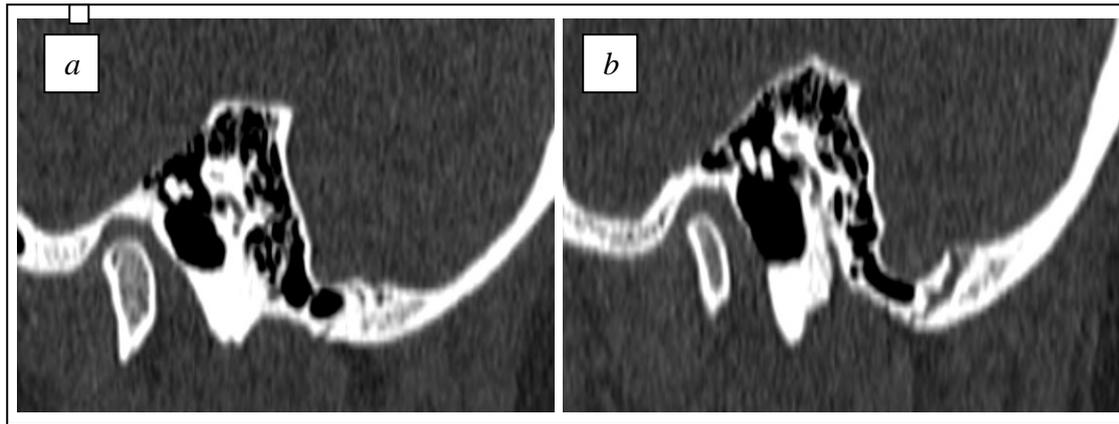


Figure 4: a : sagittal view of right temporal and b is sagittal view of left temporal, showing separation and dislocation of incus and malleus

Embryological development of middle ear is otherwise a complex process. The skeletal elements of the middle ear develop from the mesenchyme of the first two branchial arches. Although there are many hypothesis postulated differently, in general it was well accepted that middle ear develops primarily from first and second branchial arches and that the ossicles are derived from various contribution by Meckel's cartilage, Reichert's cartilage and the otic capsule. The malleus and incus are formed mainly by endochondral ossification. Their development starts as a condensation of concentric cells in the caudal extremity of Meckel's cartilage. This extends perpendicular to the main axis of Meckel's cartilage towards the otic capsule. This condensation then separates into two parallel components that remain connected dorsally but separate ventrally. The most caudally located will form the incus and the most rostral is the malleus [3]. In details, Meckel's cartilage was solely responsible for the head and neck of malleus and the body and short process of incus, while Reichert's cartilage was responsible for the handle of the malleus, the long process of incus, the crura and lateral stapedial footplate while the otic capsule was responsible for the medial stapedial footplate [4]. Therefore, the complexity of the development makes the concomitant anomalies are not uncommon.

Proper and timely diagnosis is essential to the management of these patients. A complete history, proper examination and hearing assessment could lead to the correct diagnosis. Imaging technique plays a vital role in detecting an ossicular anomaly. To date, High Resolution Computed Tomography (HRCT) is the most accurate diagnostic tool for identifying congenital ossicular anomaly pre-operatively. Often, preoperative HRCT of the temporal bone prior to exploratory tympanotomy provide guidance for otologist before revealing them intraoperatively. The major objectives of HRCT in such cases include the identification of the type of anomaly and determination of surgical correctibility. In this case, we highlight the role of HRCT as a single tool in diagnosing congenital ossicular anomaly. From HRCT, there was separation and dislocation of the incudomalleolar joint and incudostapedial joint. The incus appears abnormal. It was displaced posteriorly. The long and lenticular process of the incus was displaced inferiorly. (Figure 3, 4)

A thorough discussion on exploratory tympanotomy and ossicular reconstructive techniques is beyond the scope of this article. Exploratory tympanotomy and ossiculoplasty is helpful to confirm the diagnosis and produce a functional sound transformer mechanism to improve hearing in such cases. In **conclusion**, HRCT is a reliable diagnostic tool for ossicular anomaly and diagnosis could be obtained without surgery.

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