ORIGINAL ARTICLES

Temporal bone congenital anomalies and variatons

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Abstract

Background: We aimed to review the imaging findings of the most common congenital anomalies of the temporal bone and variations of the anatomic landmarks.

Material and methods: We retrospectively reviewed the 1,894 temporal bone computed tomography (CT) and 986 temporal bone magnetic resonance imaging (MRI) with regard to congenital anomaly and accompanying pathologies. Thin-section CT and thin-section T2-weighted gradient echo or fast spin-echo MRI was performed to identify malformations. The cases were admitted to our hospital with hearing loss, tinnitus, vertigo and non-specific infection complaints.

Results: We detected 22 cases with a temporal anomaly among 1,894 temporal CT and 986 temporal MRI images. We discussed the most common congenital anomalies of the temporal bone with imaging findings of our cases.

Conclusion: This clinical/imaging review illustrates and reviews congenital abnormalities of the temporal bone: external, middle, and inner ear anomalies, vascular malformations, and variations in high-resolution CT and MR images.

Keywords

Temporal bone, Congenital, Anomaly, Computed tomography, Magnetic resonance imaging

1 Introduction

Anatomic variations and anomalies of the temporal bone include abnormal dimension, contour and abnormal orientation of the structures. Functional impairment is the crucial distinction between anomaly and variations. In the literature, the largest temporal bone anomaly series is that reported by Valvassori *et al.* [1]. In that series, approximately 60% of temporal congenital anomalies demonstrated deformities of the external auditory canal, middle ear, or both structures [1]. Inner ear abnormalities accounted for 30% of the congenital defects, and the remaining 10% displayed mixed defects of the external, middle, and inner ear [1, 2]. We presented temporal anomalies and variations that we determined 22 cases among 2,880 patient's computed tomography (CT) and magnetic resonance imaging (MRI).

2 Material and method

We retrospectively reviewed 2,880 patients who performed temporal bone imaging (computed tomography and magnetic resonance imaging) over a 1.5-year period. We detected 22 cases with a temporal bone anomaly and variation among 2,880 patients. We reviewed anomalies and clinical complaints. We didn't consider the previous diagnosis of the patients, all anomaly cases had been included.

One thousand eight hundred ninety four (1,894) patients had high resolution temporal CT with 1 mm thick sections. Also, coronal, oblique, sagittal reformated images obtained. Nine thousand and eighty six (986) patients had temporal MRI. MRI was performed on 1.5 T system. MRI sequences are axial and coronal (reformation with 3D maximum intensity projection [MIP] reconstructions from axial images) 3D FIESTA (fast imaging enabling steady-state acquisition, TR: 6, TE: 1.7 FOV: 16 × 16, slice thickness: 1.0/-0.5, matrix: 384 × 320, NEX: 6), axial T1 weighted spin echo (TR: 600, TE: 20, slice thickness: 1), coronal T2 weighted turbo spin echo (TR: 3000, TE: 80, slice thickness: 1).

3 Results

Twelve female, 10 male (total 22, 0.7%) patients were detected with temporal bone anomaly/variation. The average age of the patients was 27 year-old (range: 12-51). Three of the patients were pediatric group. Also, there were not associated or another congenital anomaly in all age groups.

Eight patients presented with mild hearing loss, 5 patients had auricular and external acoustic canal anomalies and clinical diagnosis had previously been known. Nine of the patients had administered to the hospital with vertigo complaint.

Most of the patients (fourteen patients), had vestibular and semicircular canal anomaly or variation.

Four patients had external acoustic canal (EAC) or auricular anomaly (see Figure 1). Only one patient's congenital cholesteatoma was accompanying to other anomalies (see Figure 2). One patient had middle ear and also EAC anomaly, one patient internal acoustic canal variation, one patient had facial nerve variation. Six patients had middle ear ossicle anomalies alone or accompanying with other anomalies (see Figures 3-6). In our group, vestibular and semicircular canal anomalies/variations are most common type (see Figures 7-9). We detected three patients who had, simultanously, internal acoustic canal and facial nerve anomaly/variation (see Figures 10-12). And there were 2 patients with juguler vein anomaly/variation (see Figures 13, 14).

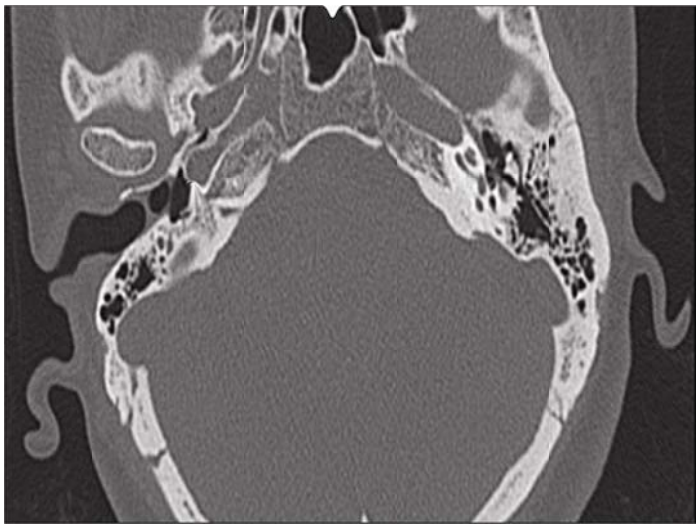

Figure 1. 43-year-old patient with fibrous septa in the right external auditory canal. Axial CT image shows thin, soft tissue bridge at right external auditory canal localization

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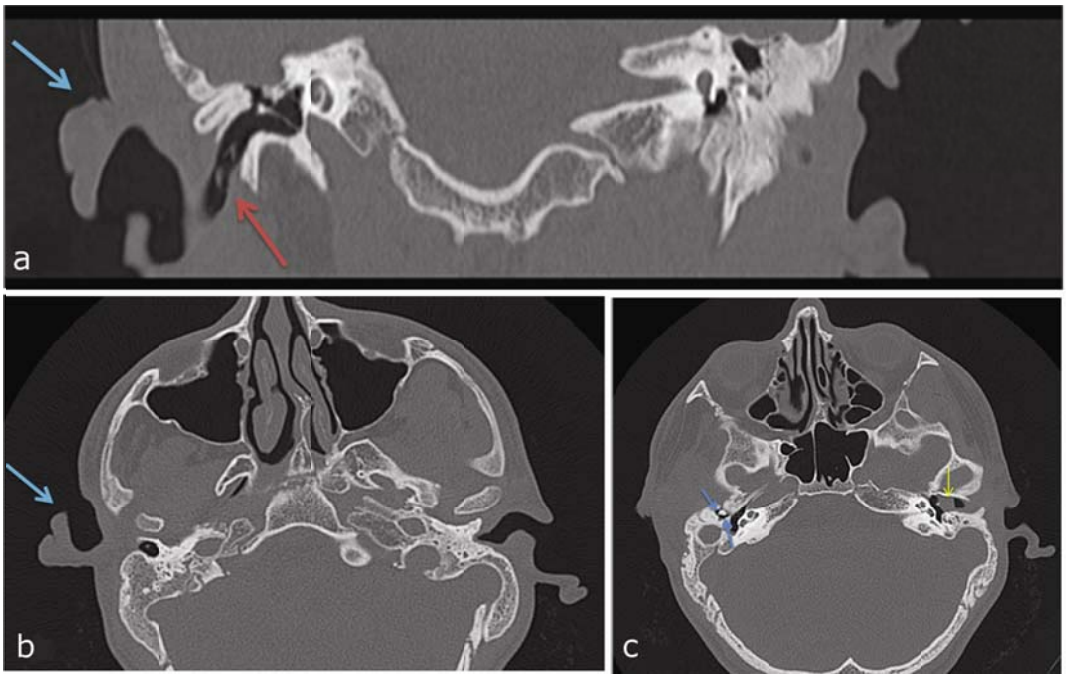

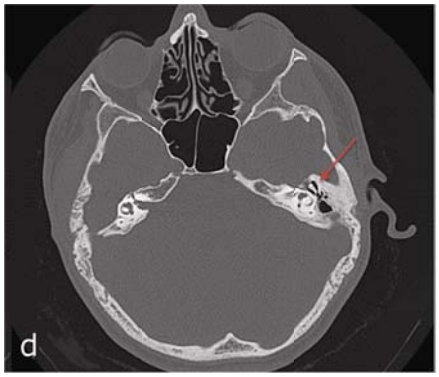

Figure 2. a: Coronal and b: Axial CT images, 15-year-old female with right minor microtia with narrow and short right external auditory canal (arrows). The left external acoustic canal and auricle is normal. Also, note, dehiscence of the tegmen tympani and hypoplasia of the mastoid air cells at right side; c: On the right, stapes aplasia and incus and malleus hypoplasia (arrows) and small middle ear cavity. Left congenital cholesteatoma at prussak space, lateral to the ossicular chain(arrow); d: with normal left ossicular chain(arrow) but hypoplasia of the mastoid air cells

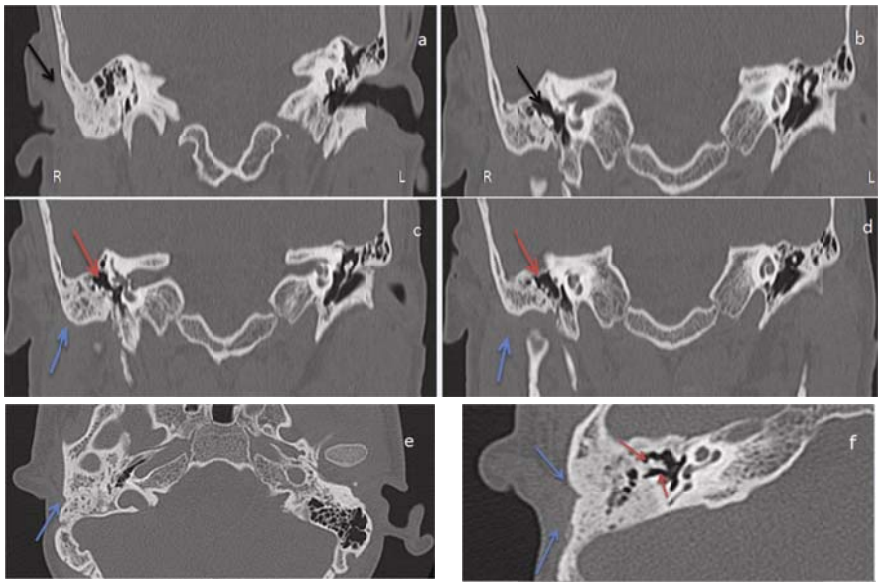

Figure 3. 17-year-old male. (a-d): consecutive coronal; (e-f): axial CT images. Microtia and atresia of the external auditory canal (EAC) (a-f, arrows), aplasia of the stapes, deformed appereance and hypoplasia of the malleus and incus (b-f, arrows) and small middle ear cavity; accompanying middle ear ossicle anomalies to EAC atresia. Left ear structures are normal

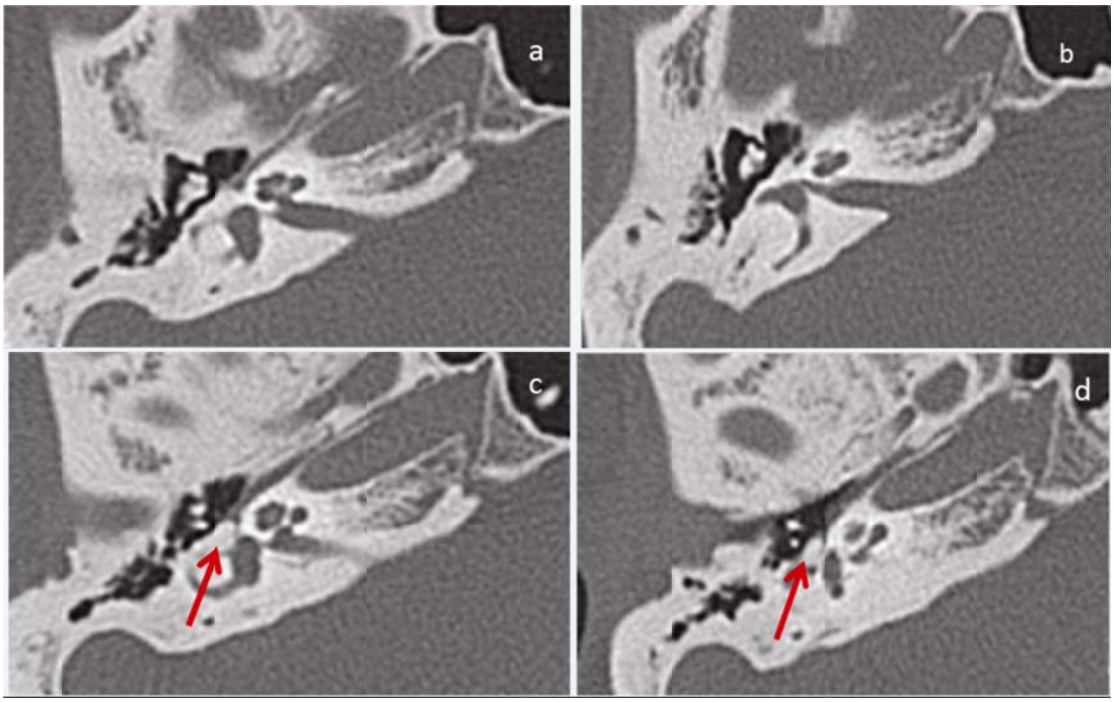

Figure 4. Axial CT images in a 14-year-old boy with conductive type hearing loss on right side. (a-b): Incus, malleus and incudomalleolar joint is normal; (c-d): Congenital absence of the oval window and the stapes on CT scan. The oval window is completely obliterated with bone (arrows). The lateral semicircular canal is normal. The facial nerve is aberrant passed in the middle ear. The stapes is agenetic. Note that the nerve is medial in location

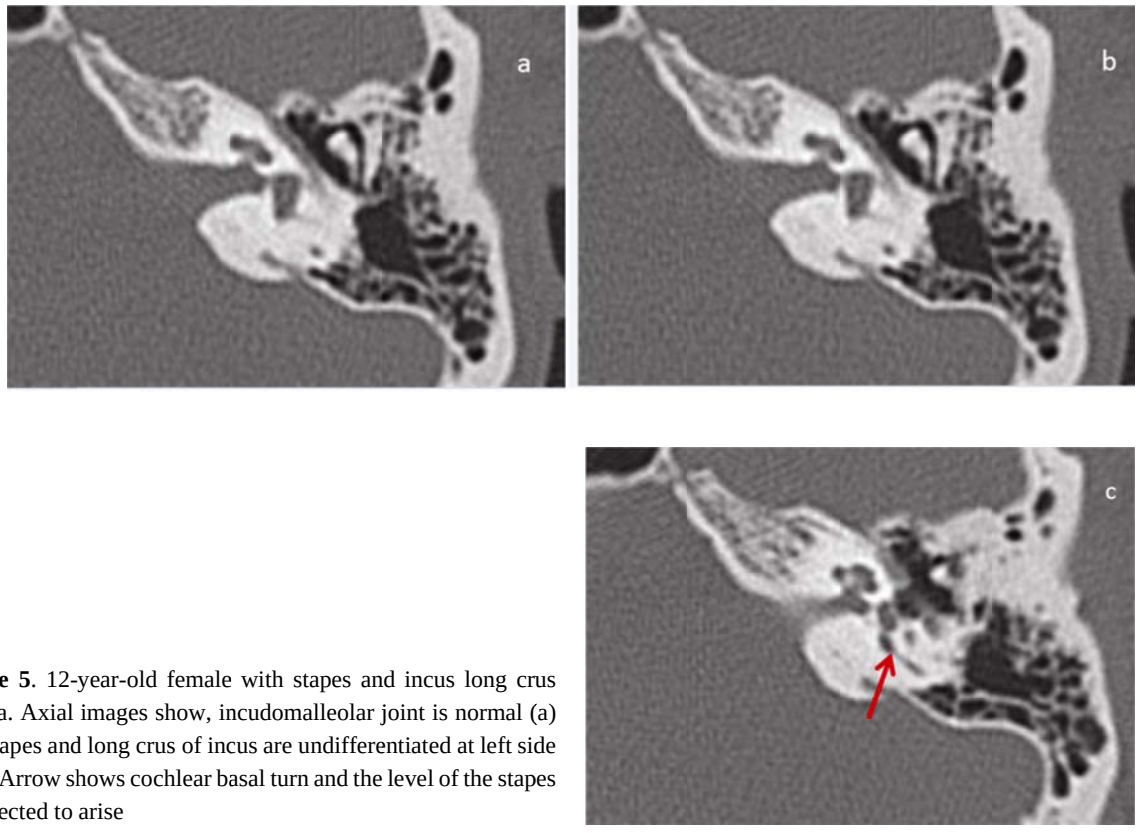

Figure 5. 12-year-old female with stapes and incus long crus aplasia. Axial images show, incudomalleolar joint is normal (a) but, stapes and long crus of incus are undifferentiated at left side (b-c). Arrow shows cochlear basal turn and the level of the stapes is expected to arise

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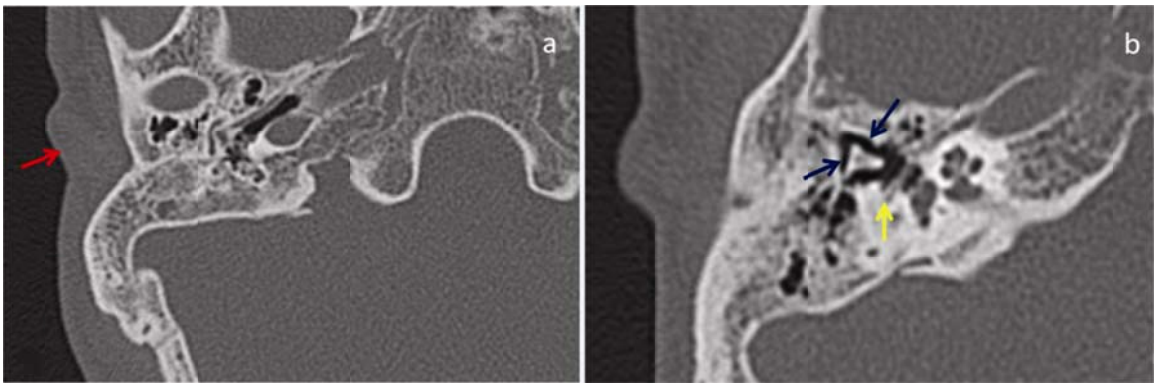

Figure 6. a: 12-year-old girl with profound right sided congenital conductive type hearing loss (Major microtia). Axial CT scan shows right external auditory canal bone aplasia with hypoplasia of auricula (arrow); b: The volume of the middle ear is decreased (arrows). The incudomalleolar joint is absent, the second portion of the facial nerve shows that the free passage of the middle ear (arrow)

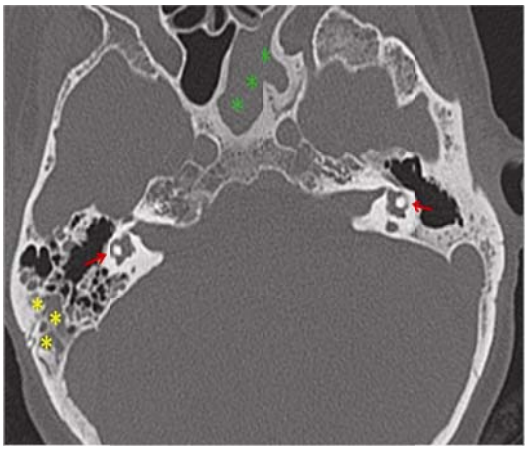

Figure 7. Axial CT scan image in a 26-year-old patient with bilateral hypoplasia of the lateral semicircular canal. Lateral semicircular canals are seen petite (arrows). There are observed insidentally right mastoiditis (markings) and left chronic sphenoid sinusitis (markings) in this image

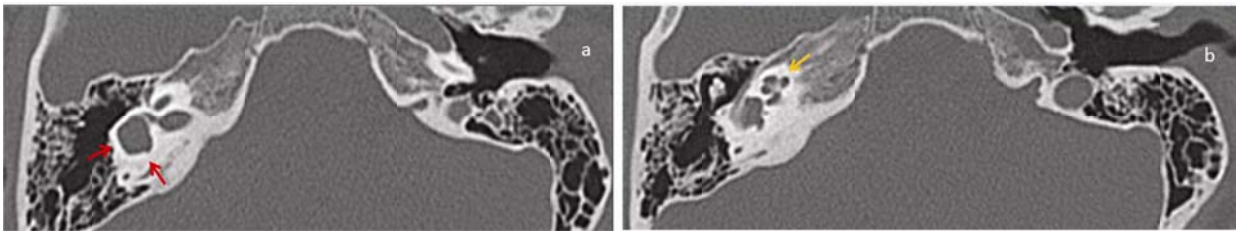

Figure 8. Axial CT images (a-b) of right inner ear shows saccular enlargement of vestibule and no separation of the lateral semicircular canal with the vestibule (arrows). Also note cochlea is normal

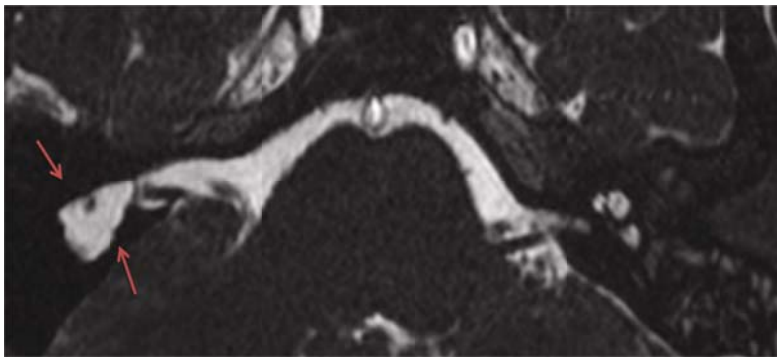

Figure 9. 19-year-old girl with common cavity on right side (arrows). Axial 3D FIESTA images of the right ear shows cochlea and the vestibule can not be distinguished from each other. Note left inner ear is normal

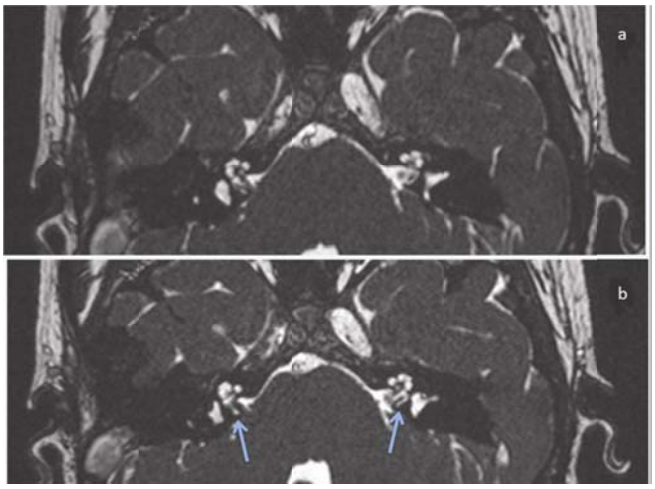

Figure 10. 19-year-old female, without any clinical finding, a and b, consecutive axial T2-weighted FIESTA images; bilateral small and short internal acoustic canal (arrows). VII and VIII. cranial nerves are normal. Also there was not inner, middle and external ear anomalies

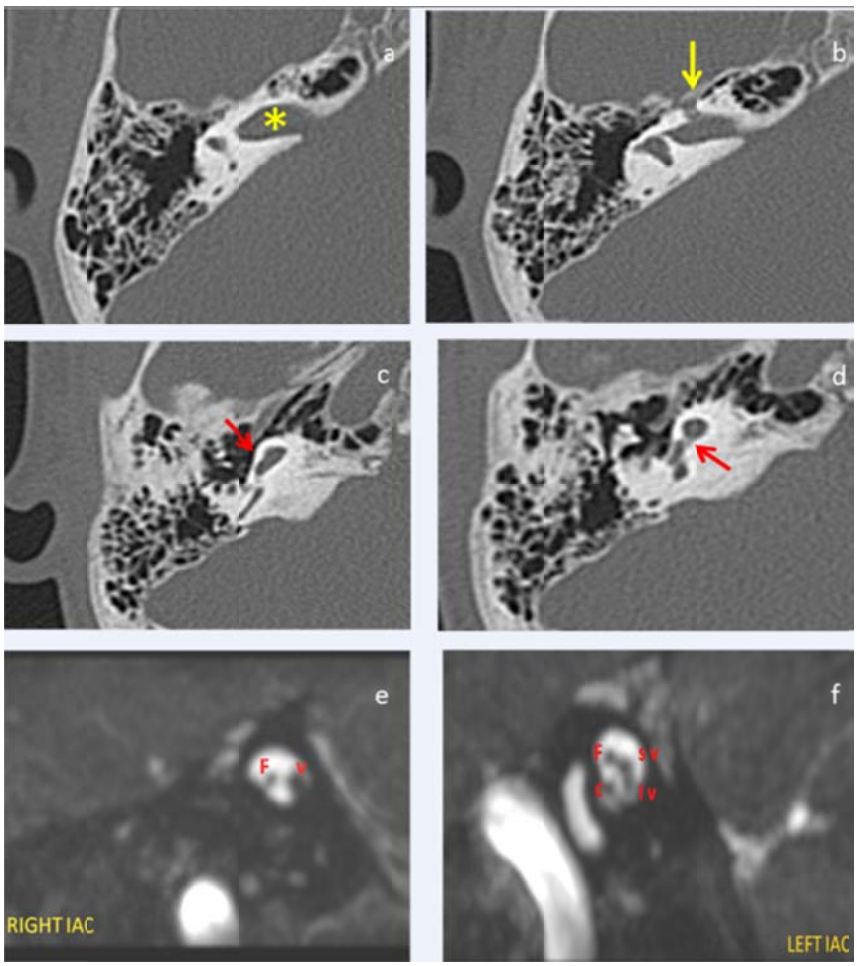

Figure 11. a-d: Sequential axial CT images in a 17-year-old woman with sensorineural hearing loss on right side. Images shows cohlear hypoplasia (arrows: b, c, d) with normal internal acustic canal and facial nerve (marking and arrow: a, b); b: Further MR imaging of the same patient, Coronal T2-weighted three-dimensional gradient echo sequence shows abscence of the right cohlear nerve. There are seen normal facial nerve (F) and conjoint vestibuler nerve (V) on right side (e). Observed in all the nerves as normally on the left side (F-facial nerve, SV- superior vestibular nerve, IV-inferior vestibular nerve, C-cohlear nerve: f). (*MR images were obtained at the level of internal acoustic canal.)

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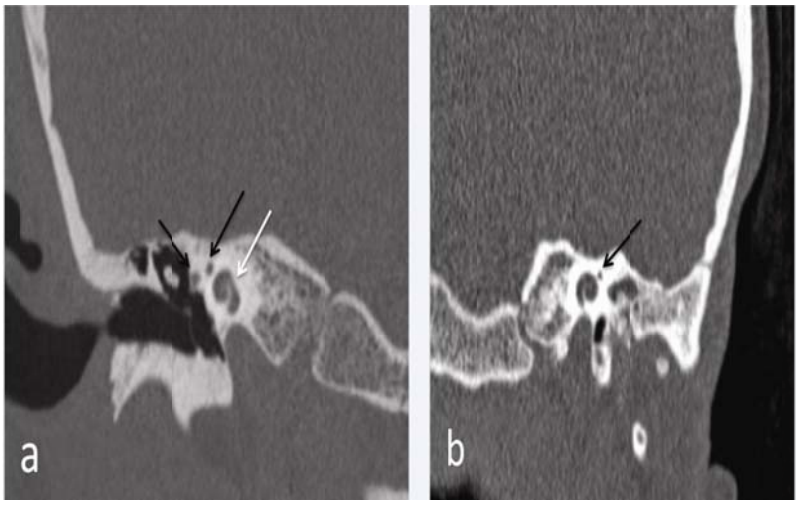

Figure 12. a: The labyrinthic and tympanic segments of the facial nerve (black arrows) have normal appearance at coronal CT image. Also, cohlea is normal (white arrow). The labyrinthic and tympanic segments of the normal facial nerve and cohlea show "eyes of the snake" appearance at coronal CT images; b: The tympanic segment of the facial nerve is hypoplastic. Also, the labyrinthic segment of the facial nerve is small according to normal side (arrow). The accompanying pathologies of the middle ear and ossicles were detected at other images

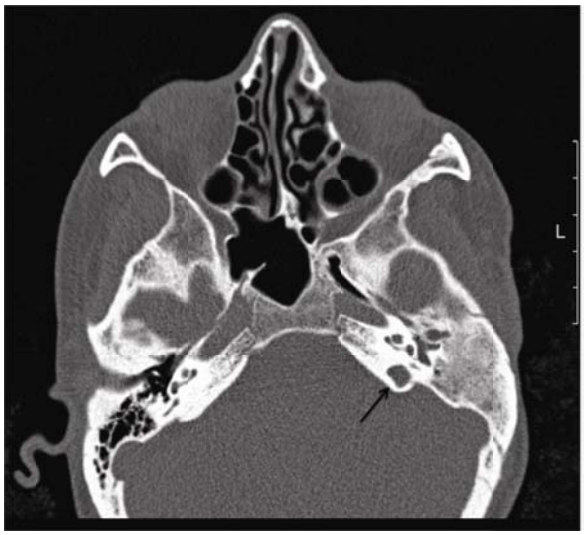

Figure 13. Axial CT image; the high jugular bulb (arrow) is seen that positioned higher from the floor of the tympanic cavity at left side. Also, there is hypoplasia of the mastoid cellulares and middle ear anomalies

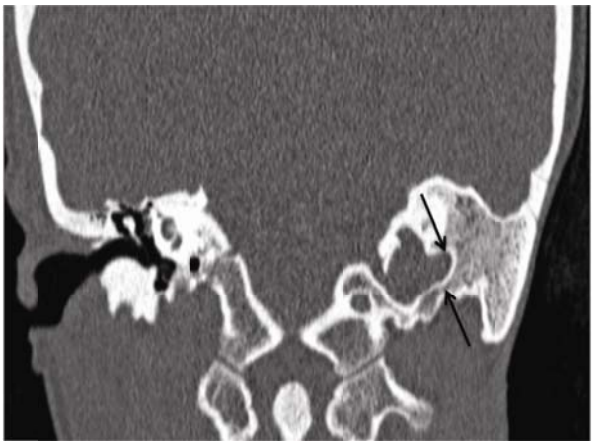

Figure 14. Coronal CT image; the jugular diverticulum (black arrows) is seen as smooth outpouching component of the jugular bulb that extends superiorly and posteriorly

4 Discussion

Microtia is characterized by a small, abnormally shaped auricle. It is usually accompanied by a narrow, blocked or absent external ear canal. In microtia, often a small tissue tag and a small pit are present in place of the normal auricle and external canal [2-5].

EAC malformations may be observed as atresia, stenosis, or a duplication anomaly (cyst, sinus, or fistula). Additionally, there may be a soft-tissue plug or a bony plate localized at the tympanic membrane (TM). The incidence of primary and secondary cholesteatomas or epidermoids increases in EAC malformations [2, 3].

Middle ear ossicular deformities can be associated with EAC malformations. Additionally, the air content of the middle ear may be reduced [2, 6].

Aplasia or hypoplasia of the tympanic part or mastoid process of the temporal bone, mandibular condyle dysplasia, rotation or elevation of the carotid canal, hypoplasia/aplasia of the internal carotid artery, aplasia of the oval and round windows, and an anomalous course of the facial nerve may be observed [2, 6].

Stapes and incus are the most frequently malformed or absent ossicles. Stapes footplate fixation is the most common isolated congenital ossicular anomaly, which is usually bilateral. Stapes anomalies include aplasia, hypoplasia or fetal form, absence of the head and crura, fusion of the head to the promontory, and footplate fixation [2, 6-8].

Incus anomalies include aplasia, fusion of the short process to the lateral semicircular canal, shortening or malformation of the long process, absent or fibrous union of the incudostapedial joint, and isolated fixation of the incus to th scutum [2, 6 , 7-12].

The most common inner ear anomaly is malformation of the lateral semicircular canal. The malformed canals may be short, wide, or narrow [2]. In severe malformations, a dilated vestibule may form a common lumen, a condition known as lateral semicircular duct-vestibule dysplasia or common utriculosaccular-lateral semicircular duct cavity [2, 11-13]. Common cavity anomaly is seen as a large cystic cavity with no inner ear structure. The semicircular canals may be normal or malformed [2].

Incomplete partition and dilatational defects (including Mondini's dysplasia) of the inner ear.

There is a small cochlea with incomplete partitioning of the interscalar septum. The interscalar septal defects and absence of the osseous spiral lamina can be visualized on MRI. Additionally, dilatation of the vestibular aqueduct may be present. The vestibule and semicircular canals may be normal or malformed. This malformation was first described by Mondini [2, 12-17]. We didn't determine Mondini dysplasia among our patients.

The internal acoustic canal (IAC) may also be atretic or may have a bony septum. The bony margins of the stenotic/atretic canal can be demonstrated by CT. The bony deformity may compress the vestibulocochlear nerves (VCNs) [2, 12, 18, 19].

Hypoplasia or aplasia of the vestibulocochlear nerves and facial nerve can be identified by MRI. Three types of vestibulocochlear nerve hypoplasia and aplasia can be distinguished, as follows: type 1 displays stenotic IAC and VCN aplasia; type 2A involves a common VCN with aplasia/hypoplasia of the cochlear branch and labyrinthine malformation; type 2B shows normal labyrinthine structures [2, 12, 18]. Also, facial nerve hypoplasia may be seen with middle ear anomalies. Additionally, isolated congenital cochlear nerve hypoplasia has been reported [2, 12, 17-19].

Duplication of the facial nerve, also referred to as a bifid facial nerve, is another anomaly.

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Anterior migration of the facial nerve has also been described with cochlear malformations. Identification of the position of the facial nerve is a crucial part of preoperative planning [2,12, 18, 19].

The dome of the jugular bulb, when positioned higher from the floor of the tympanic cavity, is called a high jugular bulb [2]. A high lateral jugular bulb may obstruct the round window. A high medial jugular bulb may compress the distal portion of the vestibular aqueduct. A relationship exists between pneumatization of the mastoid air cells with a high jugular bulb [2, 20].

A jugular diverticulum (JD) is a very rare anomaly characterized by smooth outpouching of the jugular bulb that extends superiorly, medially, or posteriorly [2, 20].

4 Conclusion

Recognizing congenital abnormalities of the temporal bone guides management of these conditions by clinicians. We aimed to review and illustrate anomalies of the temporal bone and outer, middle, and inner ear structures, as well as vascular and nerve anomalies, with or without hearing loss on high-resolution CT and MR images.

Conflict of interest disclosure

The authors declare no conflicts of interest.

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