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REVIEW ARTICLE

CT and MR imaging of Pediatric Temporal Bone: A Pictorial Review

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ABSTRACT:

Learning Objective- To describe imaging features of various temporal bone pathologies in pediatric population. **Background-** A wide range of conditions can effect pediatric population with significant morbidity. High resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) provide excellent details of the temporal bone anatomy. CT depicts minute details of osseous structures and MRI allows visualization of the fluid filled spaces and the vestibulococlear nerve. Together these modalities can detect various diseases effecting temporal bone and can also aid in surgical planning. **Procedure details-** We have illustrated HRCT and MRI findings in various congenital/ developmental, inflammatory, traumatic and neoplastic lesions of temporal bone. **Conclusion** - A vast spectrum of diseases can afflict the temporal bone in pediatric population. HRCT temporal bone is imaging modality of choice for evaluation of temporal bone. MRI is useful for inner ear pathology. Radiologist should be aware of imaging findings in various pathological entities. **Key words:** CT imaging, MR imaging, Pediatric Temporal Bone.

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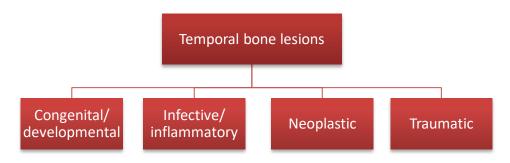
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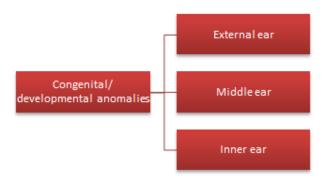
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BACKGROUND

- A wide range of conditions can effect pediatric population with significant morbidity.
- High resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) provide excellent details of the temporal bone anatomy.
- CT depicts minute details of osseous structures and MRI allows visualization of the fluid filled spaces and the vestibulocochlear nerve.
- Together these modalities can detect various diseases effecting temporal bone and can also aid in surgical planning.

A wide variety of pathologies can effect temporal bone. We have divided them into congenital/ developmental, infective/ inflammatory, neoplastic and traumatic pathologies





- As external and middle ear structures develop from first and second branchial arches, anomalies of external and middle ear are frequently associated with one another
- Development of inner ear occurs separately and concomitant developmental is unusual

ANOMALIES OF OUTER EAR

- Referred to as congenital aural dysplasias.
- Classified as first, second, and third degrees.
- In first- or second-degree dysplasias the EAC is commonly stenotic.
- In third degree EAC is atretic.
- EAC atresias can have the rudimentary or deformed pinna anywhere along a line connecting the angle of the mandible and the normal position of the EAC.

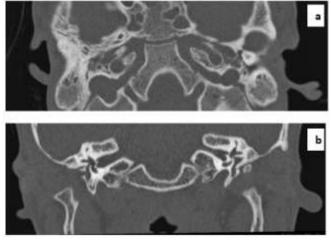


Fig.1: axial (a) and coronal (b) HRCT images shows complete b/l EAC atresia. Also note presence of small and deformed pinna

Anomalies of Middle Ear

- Anomalies of the ossicles and middle ear usually occur with EAC stenosis and atresia.
- Often associated with abnormalities in the course of facial nerve.
- May be associated with anomalies of carotid canal and jugular bulb
- These anomalies may also be found in association with a variety of syndromes, the more common of which are Goldenhar's and Treacher-Collins syndromes.

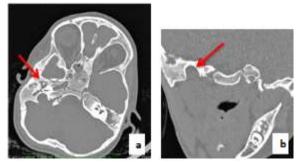


Fig. 2: Hypoplastic right middle ear cavity with dysplastic ossicular chain (depicted by arrow), associated with b/l EAC atresia (a). Also note the presence of high riding jugular bulb (arrow in b)



Fig. 3: Axial HRCT image depicting complete atresia of bilateral EAC and middle ear (a). Coronal HRCT image depicting anterolateral displacement of mastoid segment of facial nerve (arrow in b)

Treacher Collins Syndrome: Common feature Facial bone hypoplasia, involving the mandible and zygomatic complex in 75% of patients.

Auricular anomalies include absent EAC, middle ear malformations, and pinna deformities. Craniofacial defects in TCS are often bilateral and relatively symmetric.

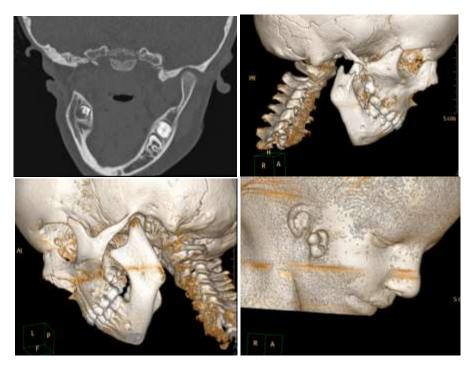


Fig. 4: Coronal HRCT and volume rendered images in a 3 years old child depicting bilateral mandibular hypoplasia, EAC atresia and microtia. Bilateral middle ear cavities were also hypoplastic.

Inner ear malformation	Imaging abnormalities
Complete labyrinthine aplasia	Complete absence of inner ear structures
Cochlear aplasia	Absent cochlea with normal or deformed vestibule and semicircular canals
Common cavity	Confluent cochlea and vestibule forming a cystic cavity with no internal
	architecture
Type I incomplete partition	Cystic cochleovestibular malformation with absence of modiolus; cystic
	vestibule present but separated from cochlea; figure-eight or snowman-like
	appearance
Cochlear hypoplasia	Small cochlear bud with less than one turn
Type II incomplete partition	Cochlea with normal basal turn and cystic apex; strong association with
	enlarged vestibular aqueduct

TABLE DEPICTING ANOMALIES OF THE INNER EAR ⁱ

ANOMALIES OF THE INNER EAR

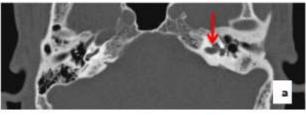
1. Complete Labyrinthine Aplasia

- Also known as Michel aplasia.
- Complete absence of inner ear structures.
- A narrow, atretic IAC is seen on high-resolution CT images.

2. Common Cavity-

Absence of the normal differentiation between the cochlea and vestibule.

CT and MR images show confluence of the cochlea and vestibule in a cystic cavity with no internal architecture (fig.5)



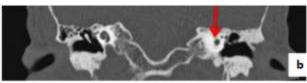


Fig. 5: Axial (a) and coronal (b) CT image shows confluence of the cochlea and vestibule in a cystic cavity with no internal architecture on left side s/o common cavity malformation .

(3) Cochlear Aplasia

- The cochlea fails to form.
- The other elements of the inner ear (i.e., the vestibule and SCCs) may be normal or malformed.

(4) Cochlear Hypoplasia

Cochlear hypoplasia displays a small rudimentary cochlear bud associated with a normal or malformed vestibule and SCCs.

(5) Type I Incomplete Partition

- Also known as cystic cochleovestibular malformation.
- The modiolus is entirely absent; the cochlea has a cystic appearance; and the vestibule is often dilated, forming a figure eight.
- The vestibular aqueduct is normal.
- The cribriform area between the cochlea and IAC is often defective, and all patients have a large IAC.

(6) Type II Incomplete Partition (Mondini deformity)

- Represents a developmental arrest in the 7th week of gestation.
- The cochlea consists of 1¹/₂ turns, and the interscalar septum and osseous spiral lamina are absent.
- The basal cochlear turn appears normal, but the middle and apical turns coalesce to form a cystic apex.
- Often associated with a large endolymphatic duct and sac and an enlarged vestibular aqueduct.
- At MR imaging, The interscalar septal defects and the absence of the osseous spiral lamina from the middle and apical turns are easily distinguished

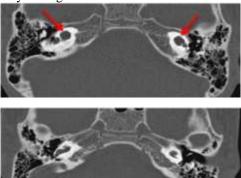


Fig.6: Axial CT image shows the absence of the modiolus from a cystic cochlear apex formed by coalescent apical and middle turns (arrows in a). Axial CT image obtained at a slightly lower level shows a normal basal turn of the cochlea (b).

Malformations of the Vestibule and Semicircular Canals.

- The development of the semicircular canals begins between the 6th and 8th weeks of gestation and is completed between the 19th and 22nd weeks.
- The superior semicircular canal develops first, and the lateral semicircular canal develops last.
- Malformation of the superior and posterior semicircular canals without involvement of the lateral canal is unusual.
- In extensive malformations, the vestibule is dilated and forms a common lumen with the lateral canal.
- Absence of all semicircular ducts is known to occur frequently in patients with CHARGE syndrome (a combination of coloboma, heart anomalies, choanal atresia, retardation of growth and development, and genital and ear anomalies)
- Isolated aplasia of the posterior semicircular duct has been described in patients with Waardenburg syndrome and Alagille syndrome.
- Vestibular malformations rarely occur in isolation. Commonly encountered anomalies include mild or globose dilatation of the vestibule with partial or complete assimilation of the semicircular canals into the vestibule

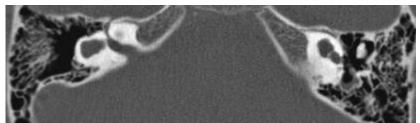


Fig. 7: Axial CT image depicting globose dilatation of the vestibule with partial or complete assimilation of the semicircular canals into the vestibule.

IAC and Cochlear Nerve Anomalies

- The normal diameter of the IAC ranges from 2 to 8 mm, with an average of 4 mm.
- An IAC with a diameter of less than 2 mm is described as stenotic.
- Three types of cochlear nerve anomalies have been described –
- Type 1 a stenotic IAC is seen with an absent eighth nerve
- Type 2 a common vestibulocochlear nerve is found, with hypoplasia or aplasia of its cochlear branch.
- If associated with other inner ear malformations it is termed type 2A
- And if occur in isolation it is termed type 2B

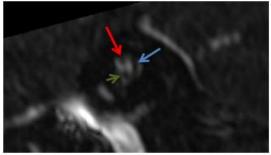


Fig. 8a: A normal facial nerve (Red arrow) and a normal-sized cochlear branch (*olive green arrow*) and dividing common vestibular branch (*blue arrow*) of the VCN can be seen.

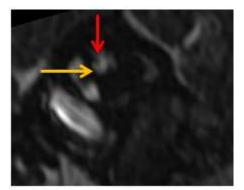


Fig. 8b: Sagittal MR image shows absence of the cochlear nerve (yellow arrow). Note presence of normal facial nerve in superior compartment (red arrow)

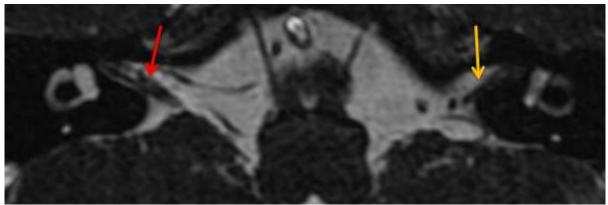


Fig. 9: Axial MR image shows absent left VIII nerve (yellow arrow). Normal VII and VIII nerve seen on right side (Red arrow).

Inflammatory Disease of Temporal Bone

External Ear -

(1) Acute External Otitis

- Most common external ear infection.
- Patients present with auricular discomfort, swelling, and pain.
- Bacterial disease (*Pseudomonas aeruginosa, Staphylococcus aureus*) is most common, followed by fungal disease, which is typically seen in diabetics.
- Imaging has a limited role except when the infection is refractory to conventional therapy.

(2) Chronic External Otitis:

- Result in a fibrous plug at the medial margin of the bony external auditory canal termed as (medial canal fibrosis)
- CT demonstrates crescentic soft tissue abutting the TM.

Middle Ear

(1) Acute otitis media - Acute otitis media is primarily a disease of infants and young children.

- Caused by bacteria such as Streptococcus species or Haemophilus influenza.
- On imaging opacities are seen in the middle ear and mastoid, with possible fluid levels.
- There is preservation of the mastoid trabeculae and the overlying cortex.ⁱⁱ

(2) Chronic otitis media- Chronic inflammation of the middle ear is known as chronic otitis media, or chronic otomastoiditis if there is mastoid involvement.

- Important sequelae of chronic otitis media that may be seen at imaging include middle ear effusion, granulation tissue, cholesterol granuloma, and cholesteatoma.
- Granulation tissue is a sequela of inflammation of the middle ear and mastoid.
- It encases middle ear structures but does not destroy or displace them, and there is no mass effect.

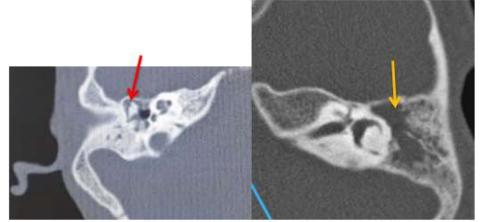


Fig. 10: Axial CT images depicting soft tissue attenuation contents in middle ear cavity (red arrow), mastoid antrum (yellow arrow) and mastoid air cells

Cholesteatoma -

Cholesteatoma is seen as a rounded expansile lobulated lesion in the Prussak space eroding the scutum, with medial displacement and erosion of the ossicles.ⁱⁱⁱ

Cholesteatoma - Soft tissue opacification in the attic, aditus and can reach mastoid air cells with blunting / erosion of the scutum and ossicular erosions.^{iv}



Fig. 11: Soft tissue attenuation seen in Prussack, s space with blunting of scutum (Red arrow) s/o cholesteatoma

Complications:

(1) **Tegmen tympani erosion** – Erosion of roof of middle ear (tegmen tympani) is a common complication of cholesteatoma.

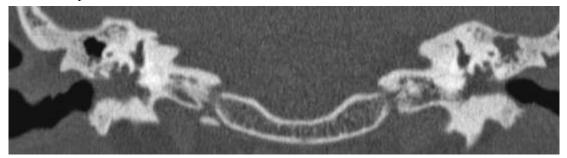


Fig. 13 Coronal HRCT image showing erosion of tegmen tympani on left side (Red arrow)

(2) Petrous Apicitis:

- Petrous apicitis occurs in the setting of a pneumatized petrous apex.
- It is characterized by septal and cortical destruction, osteitis, and adjacent meningeal inflammation.^v
- Because of the close proximity of the fifth and sixth nerves to the petrous apex, patients can develop sixth nerve palsy and deep retro-orbital pain in the V1 distribution in association with otomastoiditis and petrous apicitis (termed as Gradinego syndrome).
- The MR imaging findings of petrous apicitis are high signal intensity on T2-weighted images, low signal intensity on T1-weighted images, and contrast enhancement in a pneumatized anterior petrous apex. There may be associated enhancement of the adjacent dura mater and cranial nerves due to meningitis

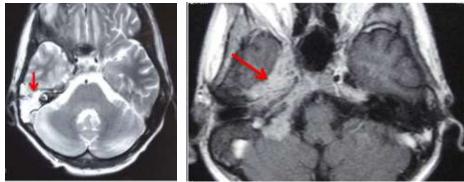


Fig. 14: T2 weighted images showing high SI in mastoid air cells s/o mastoidits (arrow in a). Gadoliniumenhanced T1-weighted MR image shows diffuse enhancement in rt anterior petrous apex, and clivus (b) s/o petrous apicits.

(3) Labyrinthitis ossificans –

- Ossification of the membranous labyrinth develops as the final result of many inflammatory processes, for example, meningitis, blood-borne septic emboli, middle ear infection, and cholesteatoma.
- On CT High-density bone deposition within the membranous labyrinth.
- On MRI T2: may show low-intensity foci within high signal fluid of inner ear.^{vi}

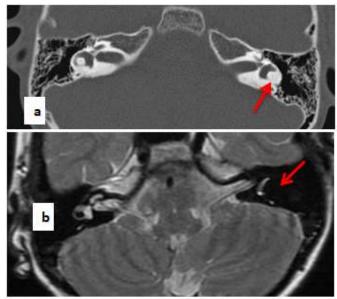


Fig. 15: a) Axial HRCT image shows ossification of left lateral semicircular canal(red arrow). b) Axial T2 MR image shows loss of bright Signal in left semicircular canal

Trauma

- Fractures of the temporal bone are associated with head injuries.
- Acute signs like bleeding or acute facial paralysis.
- Temporal bone fractures can be classified as longitudinal or transverse.

(1) Transverse fractures:

- Perpendicular to the long axis of the petrous pyramid
- labyrinthine in lateral variety
- Sensorineural hearing loss
- medial and lateral subtypes.

(2) Longitudinal fractures

- · Parallel to the long axis of the petrous pyramid
- Most often extralabyrinthine
- Conductive hearing loss is quite common

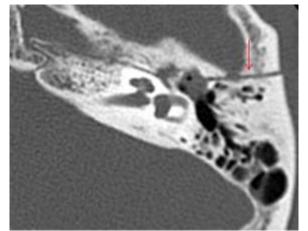


Fig. 16: Longitudinal temporal bone fracture on Left side with soft tissue in middle ear and mastoid with involvement of tympanic segment of facial nerve.

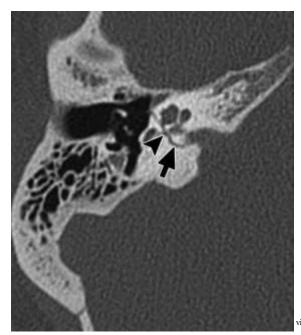


Fig. 16 b: The transverse fracture is oriented perpendicular to the petrous pyramid (arrow, b) and traverses the basal turn of the cochlea (arrowhead, b).

Tumor

- Rhabdomyosarcoma is the most common soft tissue malignancy found in children less than 5 years of age.
- Lesions originating in the middle ear are initially manifested as chronic otitis media.
- Temporal bone rhabdomyosarcomas usually show aggressive osseous destruction with obliteration of the normal land marks of the base of the skull.

MR - they are minimally hyperintense to muscle on T1-weighted images and markedly hyperintense to muscle on T2-weighted sequences.^{viii}

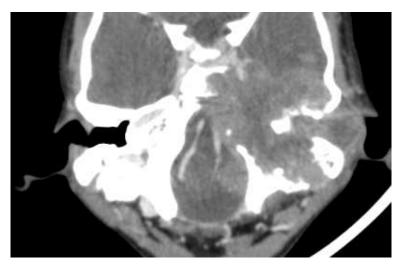


Fig. 17: Contrast enhanced axial CT image showing enhancing soft tissue mass in the region of left temporal bone causing its osseos destruction and loss of normal anatomical landmarks. FNAC from the lesion came out to be rhabdomyosarcoma

Anatomical Variants

- (1) High Riding Jugular Bulb
 - A "high-riding jugular bulb" is defined as an extension of the most cephalad portion of the jugular bulb superior to the floor of the IAC.^{ix}

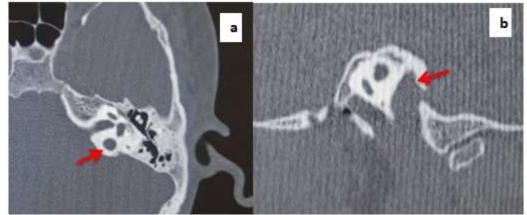


Fig. 18: Axial and Sagittal CT images showing high riding Jugular bulb (arrows in a and b)

(2) Bulging sigmoid sinus

The sigmoid sinus can protrude into the posterior mastoid. It can be accidentally lacerated during a mastoidectomy and therefore should be mentioned in the radiological report when present.

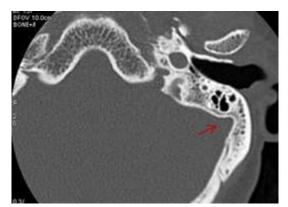


Fig.19: Axial image depicting bulging sigmoid sinus.

Conclusion

- A vast spectrum of diseases can afflict the temporal bone in pediatric population.
- HRCT temporal bone is imaging modality of choice for evaluation of temporal bone especially middle ear structures.
- MRI is particularly useful for inner ear pathology.
- Radiologist should be aware of imaging findings in various pathological entities.

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