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Rare Imaging Presentation In A Child With Clinical History Of Hearing Loss

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Abstract : *Hemifacial microsomia is a congenital deformity with deficiency of skeletal, soft tissue on one side of the face. It is a syndrome including hypoplastic mandibular ramus, ears, defects in facial nerve muscles and temporomandibular joint. The soft tissue malformations are present in the external ear. Underdevelopment of middle ear ossicles can be associated leading on to hearing loss with small external auditory meatus . we present a case of 6-year-old girl, who presented with history of loss of hearing with clinical facial asymmetry and congenital deformity of bilateral external ears*
Keywords - *Hemifacial microsomia, Hypoplasia, atresia, deformed external ear, facial asymmetry*

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I. Introduction

Hemifacial microsomia (HFM) was first described by German physician Carl Ferdinand Von Arlt in 1881. HFM comprises of unilateral microtia, malformation of the mandibular ramus, condyle with macrostomia [1]. HFM commonly affects unilaterally as compared to bilateral anomalies.
Using clinico radiological approach describing fundamental CT findings defining hemifacial microsomia and incorporating the OMENS modified classification system[2,3].

II. Case Report

6-year-old girl child clinically presented with ***history of loss of hearing , facial asymmetry and congenital deformity of bilateral external ears. She was Recently evaluated for*** deviation of angle of mouth to the left and asymmetry of left eye socket.

2.1 Imaging Findings:

Deformed hypoplastic right pinna seen . Atresia of right external auditory canal and atretic plate predominantly involving the cartilaginous part with hypopneumatized right mastoid air cells. Hypoplastic short process of incus. Right tympanic membrane is thinned out. Hypoplasia of tympanic plate of right temporal bone. Asymmetrically smaller left greater wing of sphenoid and zygomatic arch. Hypoplastic left pinna with normal left external auditory canal, normal left middle ear ossicles and normal pneumatization of mastoid air cells.

Based on OMENS classification system the it will be classified as:
Orbit – O2 -Abnormal orbital position (Inferior orbital displacement)
Mandible -M1 Left short ramus normal anatomical position of glenoid fossa and mild condylar hypoplasia.
Ear- Left E1 Bilateral external ear deformity seen as

Nerve- N0 Left
N1 Right Temporal and Zygomatic branch involvement of facial nerve
Soft Tissue -S1 Minimal soft tissue deficiency.
Microstomia- C0
No cleft.
Though the left side manefestations were predominant, External ear mal formationsand middle ear ossicular deformities were relatively significant in the right ear. As evidenced by
E2- Right Hypoplastic of anti helix absent inter tragic notch, lobule appears irregular and inferiorly placed
Short vertical tympanic segment of fascial nerve

III. Discussion

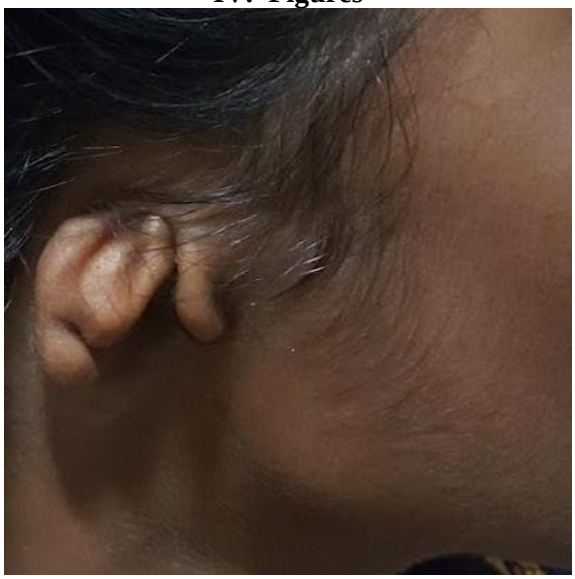
Hearing loss is commonly present in 50% of patients [4,5]. Facial nerve dysfunction is also equally mentioned in the literature [4,5]. The most common imaging findings are unilateral hypoplastic maxillary and temporal bones with or without hypoplastic zygomatic arch[6,7]. The facial midline chin are deviated to the affected side with one corner of the mouth situated higher than the other. In the present case, there was facial asymmetry, the left side of the face was underdeveloped with short zygomatic arch. The left ear was deformed with loss of hearing. The chin and midline were deviated to the left side with the corner of the mouth situated slightly higher on the right side compared to the left.

Embryology

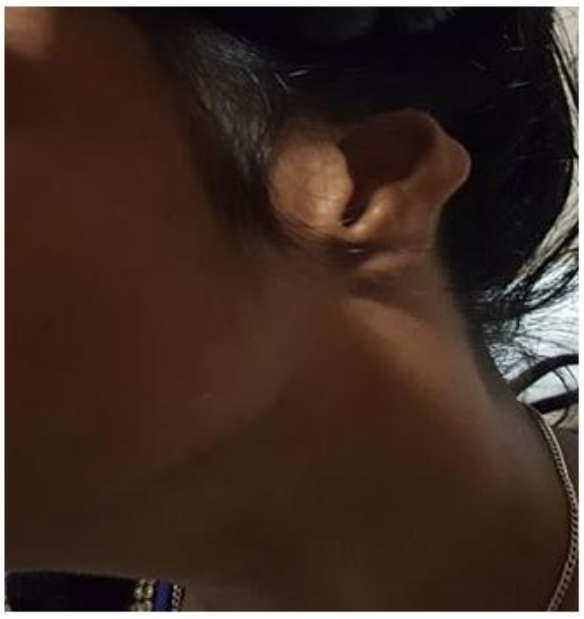
First and second pharyngeal arches are commonly affected in these patients. During the development the neural crest cells migrate to the first pharyngeal arch from the posterior mesencephalic fold which forms skeletal maxillo-mandibular component later. Damage or disruption of the migrated neural crest cells result in HFM.

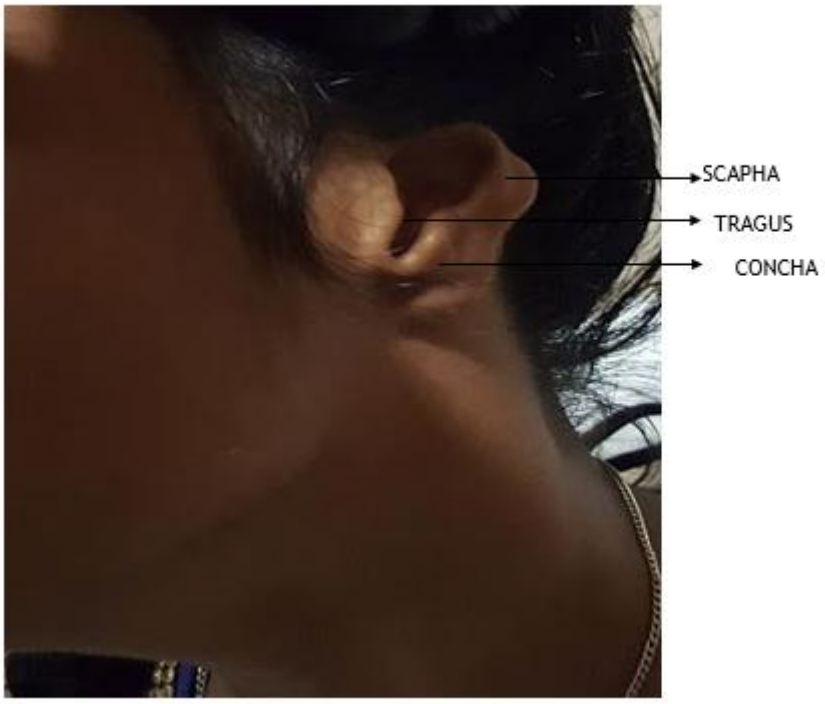
HFM is common unilaterally.

IV. Figures



Deformed small right pinna

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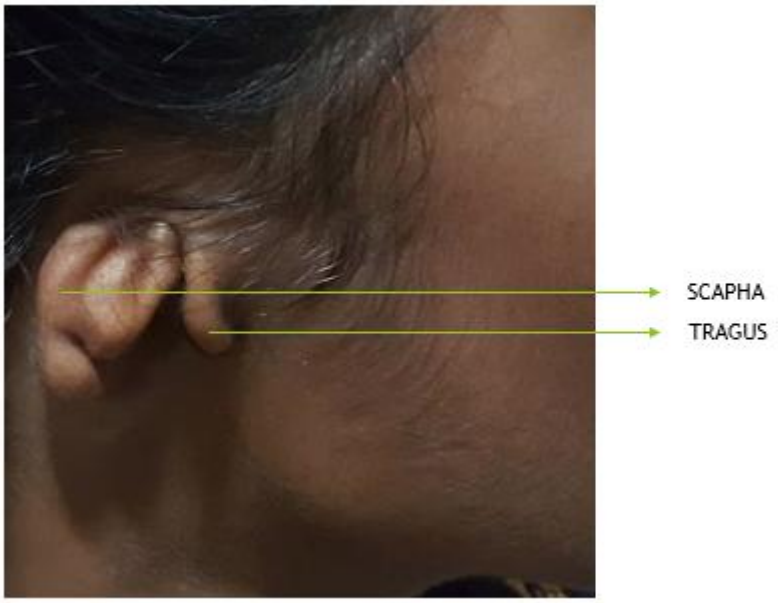
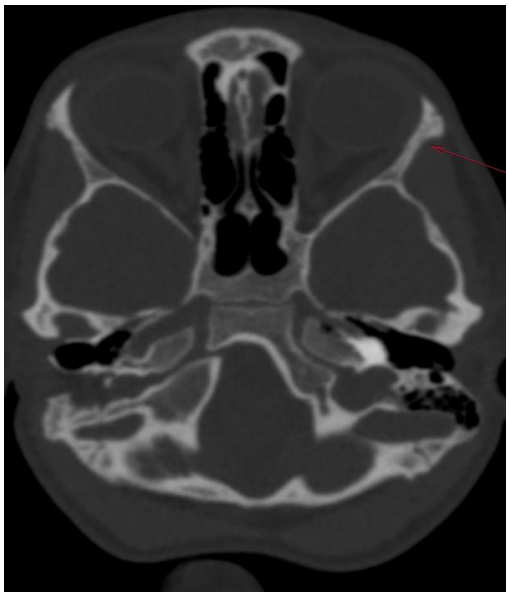
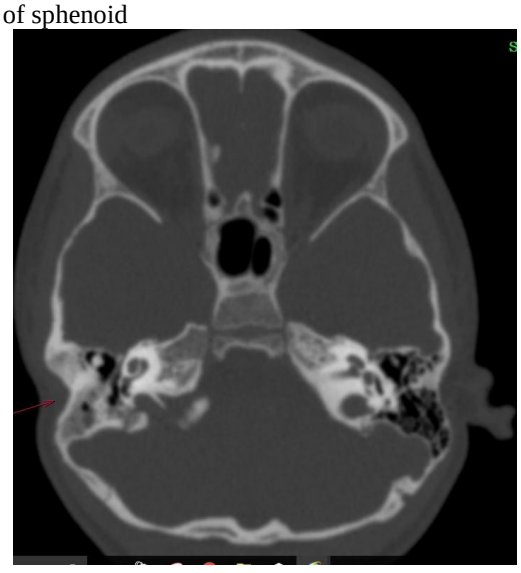
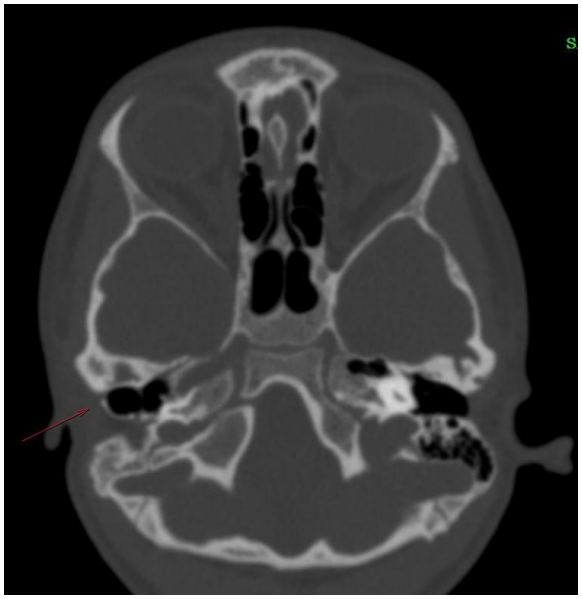




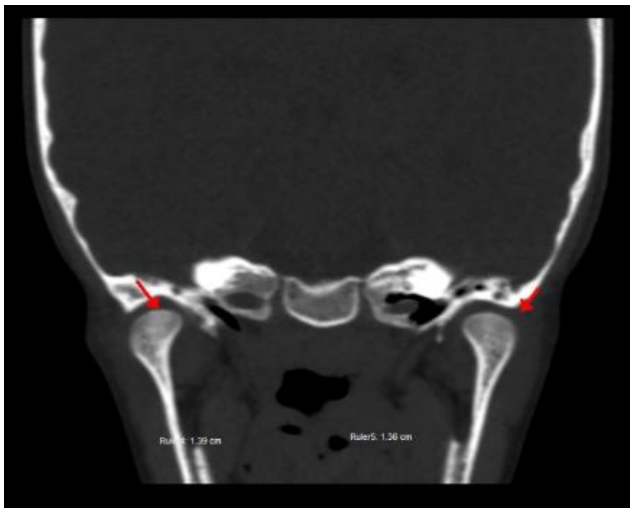
Hypoplasia of left greater wing of sphenoid

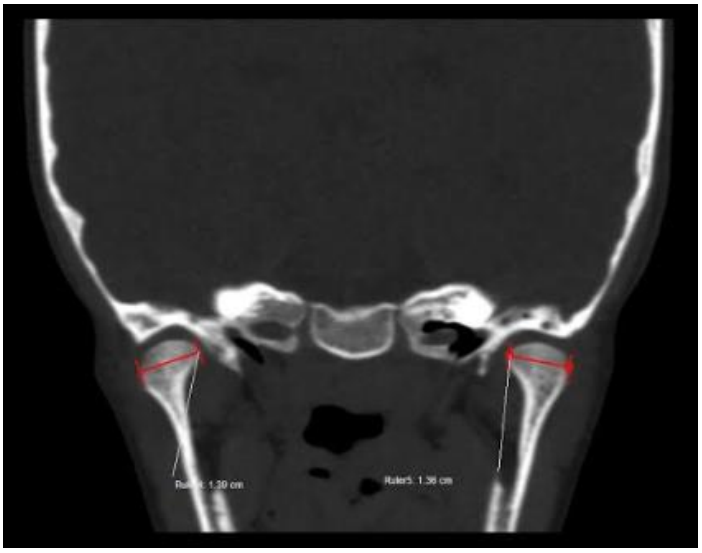


Agenesis of right mastoid air cells



Right External Auditory Canal atresia





Mild hypoplasia of condyle of left half of mandible

V. Conclusion

The hemifacial microsomia is a rare anomaly with features of unilateral hypoplasia of the craniofacial skeleton and its overlying soft tissue. OMENS classification helps to categorise the patients with diverse imaging and clinical manifestations.

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