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Bilateral Middle Ear Atresia with Right Sided Microtia

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Abstract

A dead female foetus presented normal left auricle and a small tag of tissue representing right auricle. CT scan revealed right sided absence of auricle and auditory canal and bilateral absence of middle ear. Foetal dissections presented right sided absence of musculocutaneous nerve with variation in the distribution of median nerve to muscles of arm.

Keywords: Anotia, microtia, middle ear atresia, absence of musculocutaneous nerve.

Introduction

The word Microtia (My-Kro-She-Uh) refers to a congenital abnormality in which auricle of external ear is small and deformed. Microtia and atresia of external auditory meatus usually occurs together[1]. The term Anotia (An-No-She-Uh) means congenital absence also called atresia of the auricle. Anotia/Microtia can occur unilaterally or bilaterally.

Case History: A female dead foetus (IUD) of 36 weeks gestation, 29.0 cms Crown-rump length (CRL), 33.0 cms Crown-heel length (CHL) and 2.5 Kg weight received by the department of Anatomy, SVMC, Tirupati from Govt. Maternity Hospital presented normal left auricle and a small tag of soft tissue representing right auricle (figure1).

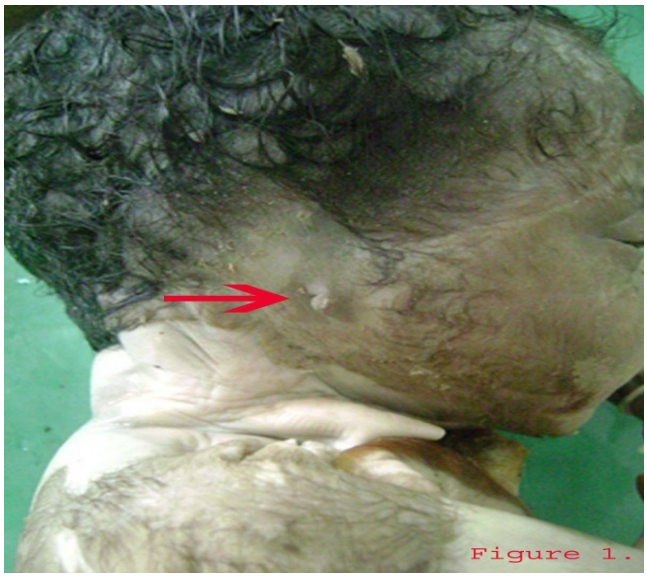

Figure-1
Foetus showing normal left auricle and a small tag of soft tissue representing right auricle

The foetus was subjected to C.T. scan of the head and temporal regions to determine anatomical defects of middle and internal ears. C.T scan revealed normal auricle and external auditory canal on left side and absence of auricle and auditory canal on right side (figure2). Middle ear cavity and ossicles were not visualized on both sides (figure3). Internal auditory canal, cochlea, semi circular canals, mastoid air cells and VII and VIII nerve complexes are normal on both sides. Temparo-mandibular joints were symmetrical on both sides.

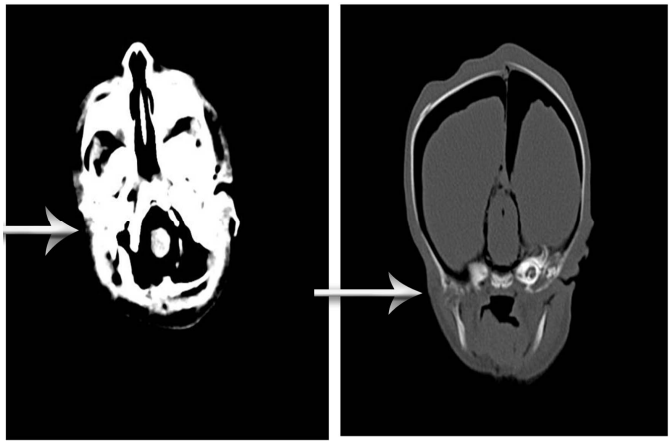

Figure-2
C.T. Scan of the head and temporal regions

After routine formalin preservation thorough dissection of the fetus was done to study the various regions and organs. Absence of Musculocutaneous nerve and variation in the distribution of median nerve to the muscles of arm (figure-4) were observed in right upper limb. On the right side lateral root of median nerve was found supplying the coracobrachialis muscle while biceps and brachialis were received innervation from median nerve in the absence of musculocutaneous nerve. On right side lateral cutaneous nerve of forearm was arising from median nerve in the middle of the arm.

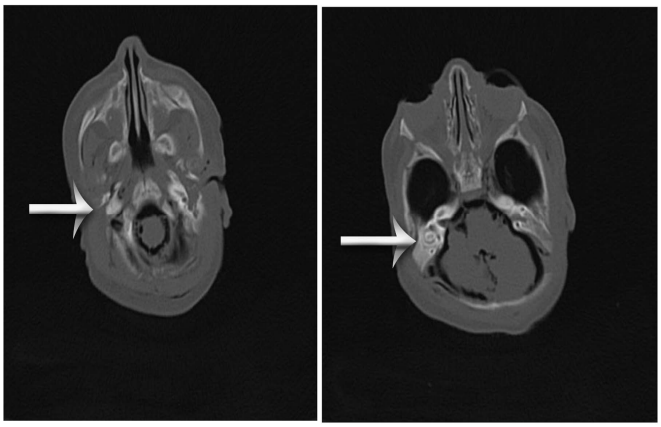

Figure-3
C.T. Scan showing Middle ear cavity and ossicles on both sides

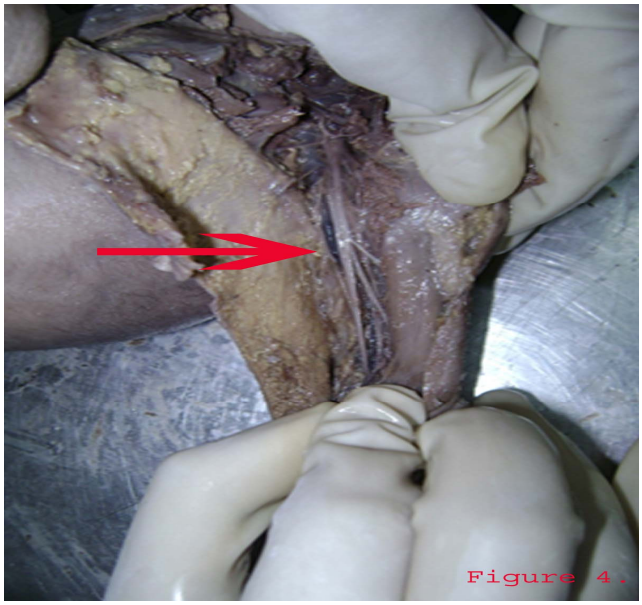

Figure-4
Showing Absence of Musculocutaneous nerve and variation in the distribution of median nerve to the muscles of arm.

Congenital malformations and agenesis of the ear can affect the external and middle ears sparing the internal ear. External ear develops from 6 mesenchymal auricular hillocks three on each of 1st and 2nd pharyngeal arches. These hillocks fuse to form the auricle. The external auditory meatus is derived from the 1st pharyngeal cleft between 1st and 2nd arches[2].

External and middle ear develop by 5th week of gestation from the posterior parts of 1st and 2nd branchial arches[3]. During 6th week of development the 1st hillock gives rise to tragus, the 2nd Hillock gives rise to Helical crus and the 3rd Hillock gives the Helix. During 12th week the 4th and 5th Hillocks become ante helix and 6th Hillock forms the anti tragus[4]. All these hillocks fuse during 20th week of development and form the adult auricle.

External auditory canal (EAC) develops between 1st and 2nd branchial arches. 1st branchial groove develops opposite corresponding 1st pharyngeal pouch. In the 2nd month this groove deepens medially into a funnel representing outer 1/3rd of the EAC known as primitive meatus. During 3rd month of development solid core of cells splits, at first pass deeply to form tympanic membrane and then extend outward to join primitive meatus to form medial 2/3rd of EAC[3.5]. Middle Ear develops during 4th -30th week dorsal aspect of 1st pharyngeal pouch develops laterally towards 1st branchial groove forming tubotympanic racess. Ossicles were develops as the Meckel's cartilage of 1st branchial arch gives rise to head of malleus, body and short process of incus and mandible. Richart's cartilage of 2nd branchial arch gives rise to short process of handle of malleus, long process of incus and stapes except vestibular portion of foot plate. Ossification of malleus and incus will takes place from 16th to 30th week, while the stapes ossifies during 18th to 21st week further remodelling through the 38th week[5].

Discussion

The term Microtia to a group of congenital external ear disorders and grouped them in to four different types depending on the degree of severity[3]. There were four grades by correlating the appearance of external ear with the development of the middle ear in patients with congenital aural atresia[6.7]. They are

Grade I: A slightly small ear with identifiable structures and presence of external ear canal though small in size. Grade II: A partial or hemi-ear with a closed off or stenotic external ear canal producing a conductive hearing loss. Grade III: Absence of the external ear with a small peanut vestige structure and an absence of the external ear canal and ear drum. Grade IV: Absence of the total ear or anotia.

Prevalence of the Anotia/Microtia in different population ranging from 0.63 to 8.7 per 10,000 live births and still births[7.8.9.10.11]. The prevalence of isolated cases, i.e., those live births and stillbirths with no other accompanying anomaly were less than that of non-isolated cases i.e. those associated with other defects/syndromes[9.10]. Isolated Microtia is most often unilateral. Majority of non-isolated cases were part of syndrome[1.12.13]. Syndromic forms and those associated with other major malformations are more common in bilateral Anotia/Microtia cases[11]. Rates of Anotia and Microtia vary according to maternal age, race/ethnicity, infant sex, plurality, birth weight, and gestational age[9.13]. Anotia and Microtia are less prevalent in whites and blacks than in Hispanics, native Americans and Asians. Among the two conditions Anotia is less prevalent than Microtia in whites[1.9.13].

Congenital malformations of external and middle ear can be part of genetic syndromes but in majority of instances they are isolated

and sporadic in occurrence[14]. More than 80% of the cases of Microtia or Anotia are unilateral[10.14]. Right ear is more often affected than the left[1,2,11,13]. The defects were less common among females and more common with multiple births, birth weight < 2500 g and gestational age <38 weeks[10]. Incidence of unilateral Anotia is approximately 1 in 8000 live births and bilateral cases are about 1 in 25,000 live births. If it is unilateral in presentation right sided Anotia is much more common (71%) than left side. It is more common in Japanese and Navaho American Indians. More common among the males (61%) while comparing with the females. During early foetal life (about 5th week) complex processes in development move the cells to correct position for the formation of ear. For unknown reasons this process is interrupted and results in Anotia. It may be due to lack of blood supply to the developing ear. It can also possibly be traced to mothers use of medications such as thalidomide and isoretinoin[11.14]. Foetal exposure to environmental toxins is also suspected as a cause in some instances other conditions that may be frequently associated with Anotia include, 18 q syndrome (Treachers Collins Syndrome)[14], Endif Rubella and other I.U infections, retinoic acid embryopathy, maternal diabetic embryopathy, The chance of recurrence in a future pregnancy is less than 6%. Common associated Conditions with Anotia these can occur independently or as a part of syndrome. 1st arch syndrome consists of congenital anomalies of the eyes, ears, palate and mandible. Also associated with the Occulo-Auriculo-Vertibral spectrum (OAV) involves facial renal vertebral anomalies[11]. Most common phenotype associated with Anotia is hemifascial microsomia, chromosomal abnormalities occurs in 6-16% of cases of Anotia, i.e. trisomy 21, trisomy 18, trisomy 13 and the deletion complexes 18q and 18p and 5p- Associated deformities[14].

The associated deformities are the percentised in the form of Branchial arch (36.5%), Facial nerve weakness (15.2%), Bony and soft tissue deformities (49.4%), Macrostomia (2.5%) Cleft lip and cleft palate (4.3%), Urogenital defects (4.0%), Cardio vascular malformations (2.5%) and miscellaneous Deformities (1.7%)[6].

Conclusion

A review of literature suggests increased incidence of right sided anotia that is more common in males. In the present case we observed right sided defect in a female foetus. Absence of musculocutaneous nerve and variation in the distribution of median nerve on right side are noted in the present case. In the literature 1.7% incidence of miscellaneous deformities were reported[15] in suspected cases. Determination of Cochlear function at the earliest is most important for consideration of starting early auditory amplification and effective auditory training (not later than 6 months) in a child. With unilateral atresia surgery is usually postponed until adult hood for adequate mastoid pneumanization. Pinna vs. Middle ear reconstruction prior to the other procedures and finally external ear by the autogenous reconstruction for pinna by the plastic surgeons and restoration of hearing by the ENT surgeons with the use of auditory amplification and training are the procedures to be undertaken.

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