Original Article

Unsafe CSOM with Post-auricular Abscess in Patients with Microtia : Our Experience

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Abstract :

Microtia is a developmental anomaly of the external ear. It is often associated with congenital aural atresia and anomalies of the middle and inner ear. In such patients, owing to the structural deformity, the middle ear is poorly ventilated resulting in retraction pocket formation and eventually may give rise to squamosal chronic otitis media.

Case Outline :

Case 1 was a 7 year old boy with right sided microtia and complete aural atresia presenting with post-auricular abscess. Case 2 was a 17 year old female with bilateral microtia, left ear discharge with post-auricular abscess.

Conclusion :

Patients with developmental anomaly of external ear should be screened properly for early diagnosis of any underlying disease to prevent further complication.

Introduction :

Microtia is defined as the abnormal development of pinna resulting in a malformed auricle. The deformities caused could range from mild distortion of pinna to complete absence of auricle (anotia). However, other components of the external (acoustic meatus and tympanic membrane), middle, and inner ear are also frequently impacted, as are other craniofacial and extra cranial anomalies. Microtia is usually unilateral (77% to 93%) and right-sided (60%). The condition also occurs more frequently in males (2.5:1). Microtia's prevalence per 10,000 births in the United States ranges from 1.8 to 3.5 and worldwide from 0.4 to 8.3. The condition is more common among asians, pacific islanders, and hispanic individuals in the United States.[1]

Although differences in opinion regarding "normative values" for auricular position and protrusion exists, Tolleth provided some general guidelines on which surgical correction can be based.[2] With the head oriented vertically, the desired position of the auricle is approximately one-ear length posterior to the lateral orbital rim. The level of the brow defines the preferred position of the top of the ear, whereas the base of the columella marks the appropriate inferior extent of the lobule. The axis of the auricle should not lie in the vertical plane; rather, it should be rotated 15 to 20 degree in the posterior direction. A distance of 15 to 20mm between the scalp & the outer edge of the helix provides an esthetically pleasing degree of auricular protrusion.

Several grading systems for microtia exist, but the Marx classification is widely used. This grading system classifies the condition as follows:

- **Grade I :** the auricle is slightly smaller, by at least 2 standard deviations below the normal, but all subunits are present.
- **Grade II :** the auricle is smaller than usual, and subunits are severely underdeveloped or absent. The auricle's superior half is often less developed than the inferior half.
- **Grade III :** only a small piece of cartilage is present in the ear's superior remnant. The lobule is rotated anterosuperiorly. This

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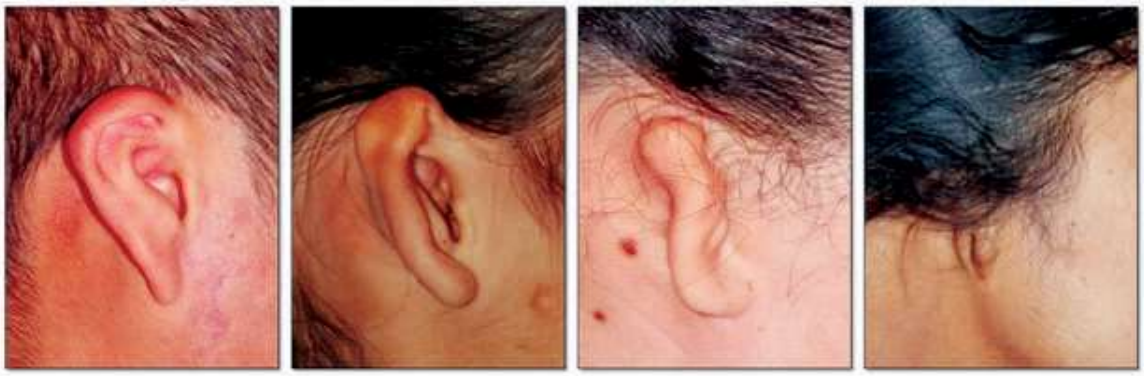
configuration is the most common, often colloquially called "peanut ear.

- **Grade IV :** the auricle and lobule are completely absent (anotia).

Marx Classification of Microtia



Grade I : Auricle small but all subunits present

Grade II : Auricle small and subunits under developed or absent

Grade III : Small cartilage remnant with anterosuperiorly rotated lobule

Grade IV : Anotia

Case 1 :

A 7 year old male child was brought to the outpatient department with complaints of right sided post auricular swelling and pain for 2 days. There was no history of nausea, vomiting, dizziness or vertigo. Hearing was markedly reduced in right ear.

On ***physical examination***, the child hadright sided microtia with atresia of external auditory canal (probe could be passed for almost 3mm). External ear consisted of lobule, tragus and a small bulge of rudimentary cartilagenous tissue behind an atretic canal. However, lobule and tragus of the diseased ear were present almost at the same level compared to the other side. His left ear was completely normal. The patient presented with a globular post-auricular swelling on the right mastoid region, of size 2x2cm^2, fluctuant in nature, tender to touch with tense, inflamed and thinned out overlying skin which was on the verge of rupturing.

Past history of post-auricular abscess of right ear present, which ruptured and healed spontaneously without any treatment.

Blood investigations revealed raised leucocyte count (18.2x10^3/ml), predominantly polymorphs, with other parameters within normal limits.

Microscopic examination of the pus from post-auricular abscess showed gram positive cocci in cluster with no growth of pathogenic organism on culture.

Audiometry & tuning fork tests gave inconsistent results.

BERA : bc-abr suggestive of cochlear pathology in right ear. Ac-abr could not be done due to external ear pathology. Ac-abr of left ear suggestive of minimal hearing loss.

HRCT temporal bone showed right sided mastoiditis with retroauricular abscess. Otitis media and externa present. Tympanic membrane not visualized. Mild erosion of middle ear ossicles. Scutum eroded. Inner ear : cochlea, vestibule, semicircular canal, cochlear aqueduct, vestibular aqueduct, facial nerve canal, jugular fossa, carotid canal- normal.

Operative findings : In order to locate the antrum of the diseased ear the mastoid tip of that side was palpated and its position compared with that of the opposite side. Bilaterally the mastoid tip and the tragus were lying at the same level. The lobule was almost 2cm above and anterior to the mastoid tip. The external canal was probed and found to be blind measuring 3mm. A curvilinear incision was made passing over the post-auricular abscess 1cm above the mastoid tip running behind the lobule. After initial dissection the spine of Henle was identified which was rudimentary. Macewan's triangle was not well defined. Mastoid antrum & attic were hypoplastic. Cholesteatoma was seen in antrum, sinodural angle and attic goingfurther anteriorly into the eustachian tube opening. Eustachian tube orifice provided an important landmark to identify the otherwise malformed middle ear. Cholesteatoma sac was found lying on the bony canal overlying the vertical segment of the facial nerve. Dura was low lying. Incus and malleus were malformed & fused together. Stapes could not be identified. Canaloplasty and meatoplasty were done by pulling and suturing the tragus more anteriorly & lobule further inferiorly. A stent was kept to maintain the patency of the external canal.

Case 2 :

A 17 year old female presented witha left post auricular swelling with pain and left ear discharge for 5 days. She also complained of left sided facial asymmetry and decreased hearing from the same side since childhood. There was no complaint of nausea, vomiting, vertigo or tinnitus.

On ***physical examination***, the patient had bilateral microtia with a diffuse swelling over the left post-auricular area extending to the angle of mandible and lateral aspect of upper third of neck. The swelling was fluctuant and tender to touch. On left side, there were two cartilaginous spicules in place of the lobule lying inferiorly to the meatus and an isolated tag of soft tissue almost 2cm posterior to the external canal. The tragus was rudimentary. Left external auditory canal was stenosed and filled with foul smelling whitish discharge. The canal ran downward and posteriorly. Tympanic membrane could not be seen. Bilateral external auditory meatus were placed more posteriorly and at a lower level, almost 3cm below the level of the horizontal line drawn from the lateral canthus of eye on the temporal bone.

Past history of left sided post auricular abscess twice, 4 years and 8 years ago, for which the patient underwent incision and drainage.

Microscopic examination of pus from the abscess showed gram positive cocci but without growth of any organism.

Audiological evaluation :

Tuning fork test :

Pre-operative: Rinne's test was positive on right side with tuning forks of 256/512/1024hz but not perceived on the left. The Weber lateralized towards the left, whereas absolute bone conduction test was equivocal on right but could not be perceived on the left.

Post -operative : After surgery Rinne's test was found to be positive in both ears with Weber lateralizing to the left; absolute bone conduction was equivocal on each side.

Left ear - pure tone average was 55 dbhl - moderate conductive hearing loss. Bone conduction could not be done as the patient was having post-auricular pain.

Right ear - bone conduction threshold withinnormal limit.

Tympanometry, Brainstem Evoked Response Audiometryand Otoacoustic emission could not be tested due to active discharge.

HRCT temporal bone : homogenous mildly enhancing soft tissue density seen infiltrating

the left external and middle ear cavities with erosion of posterior & inferior bony wall of external canal, scutum & long process of incus and stapes. It was seen completely filling epi, meso and hypo tympanum. It is also extended to Prussack's space, aditus and mastoid air cells with erosion and opacification. It was seen eroding the lateral semicircular canal with dehiscence of facial canal. Evidence of heterogenous enhancing thickwalled collection was noted in left posteroinferior auricular region. It extended inferiorly upto mid thyroid level and superiorly communicated with the left external ear cavity.

Operative findings : probing revealed a stenosed external canal running downward and posteriorly. An initial small incision of 2cm was given just below and almost 1.5cm posterior to the cartilaginous remnant away from the mastoid tip to drain the abscess. The incision was extended upward and anteriorly running between the soft tissue tag and the external canal, to 1.5cm above the meatus. Spine of Henle was rudimentary. Drilling revealed sclerosed mastoid antrum. Granulation tissue was present in aditus, attic and middle ear. Malleus and incus were malformed and fused; stapes head and stapedius tendon were visualised. Round window was found placed anteriorly. Lateral semicircular canal was eroded. Bony canal of the vertical part of the facial nerve was partly dehiscent. Abscess found to extend from canal inferiorly. Canaloplasty was done. The soft tissue tag was pulled and sutured superior to the meatus. Meatoplasty was maintained by keeping a stent.

Discussion :

The external auricle or pinna develops from a series of small cartilaginous tubercles or hillocks. Hillock 1-3 comes from 1st or mandibular arch & 4-6 comes from 2nd or hyoid arch. According to Park[3], hillock 1 produces the anterior portion of the ear lobule, hillock 2 tragus and hillock 3 the ascending helix. Of the 2nd arch hillocks, 4 and 5 produces antihelix and helix, with 6 contributing to the posterior lobule. By the end of fifth week, five branchial arches are discernable. In a 38 day old embryo, six hillocks have developed in the mesenchymal tissue of the first (mandibular) and second (hyoid) arch and a process of fusion produces a primitive ear in the 50 day old embryo. Both **case 1 & 2** presented with microtia, case 1 unilateral whereas case 2 bilateral. Malformation, such as anotia and microtia, are likely to be caused by the disturbance of development at seven or eight weeks gestational age. In both the cases tragus & lobule (1st arch structures) though present, 2nd arch anomaly was more pronounced. The ear initially forms in the neck region and moves upward onto the head by week 10. In **case 2** this migration was affected & bilateral ears were low set.

Case1 had atresia of right external auditory canal whereas **case 2** presented with stenosis of bilateral canal. At 28 weeks, a core of ectoderm canalizes from medial to lateral and eventually breaks through to communicate with the conchal depression. Failure of canalisation or more likely lack of ectodermal migration can lead to atresia of external auditory meatus and partial canalisation leads to meatal stenosis (diameter of the canal less than 4mm).

In both the cases malleus and incus were malformed and fused. The head of the malleus and body and short process of the incus develops from Meckel's cartilage (first arch derivative), whereas, the manubrium of the malleus, long process of the incus and stapes suprastructure arise from Reichert's cartilage (second arch derivative). The process begins at 4 weeks and adult shape, size and ossification is present by 25 weeks.

The full-sized outline of membranous labyrinth is formed by 25 weeks of gestation. The Superior

Semicircular canal starts to develop at 35 days. Cochlea is also formed by 25 weeks. The Organ of Corti starts developing as a single block of heaped up ectodermal cells at about 11 weeks. Within this mass develop inner & outer hair cells and then specialized supporting cells.[4] Recent studies using high resolution computed tomography suggest a higher rate of inner ear congenital anomalies affecting between 10 and 47 percent of patient with atresia. BERA of **case 1** is suggestive of cochlear pathology in right ear. Audiological evaluation in **case 2** revealed moderate conductive hearing loss.

Pathology :

Developmental anomaly of external ear is usually associated with malformed middle ear which compromises the ventilation of middle ear cavity via eustachian tube. This causes retraction of tympanic membrane that marks the beginning of any disease process. Because of the stenosed or atretic canal there is inadequate drainage and patients usually present late with complications like subperiosteal abscess.

Microtia is the developmental anomaly of ear due to failure of fusion of hillocks developing from 1st & 2nd pharyngeal arches. The normal size of the auricle at birth is 66% of the length & 76% of the width of an adult ear. By the age of six, the auricle has attained 90% of adult proportion.[5] A study by Becker & Tos[6] showed the incidence of atresia of external auditory canal to be 0.5 per 1,00,000 population, incidence of membranous atresia being twenty times less than the solid form. Congenital aural stenosis as compared to aural atresia, carries a much greater risk of cholesteatoma. Jahrsdoerfer & Cole[7] reviewed 600 cases of major congenital ear malformation. Fifty patients (54 ears) were found to have aural stenosis. The most significant finding was that in children of 12 years or older with a meatus narrower than 2mm, 91% develop cholesteatoma.

Conclusion :

Microtia may occur in isolation or be associated with other congenital anomalies. Family history & proper antenatal screening helps us in anticipating such occurrences.

Hearing should be assessed early in an infant with microtia or aural atresia. Auditory brainstem response testing should be performed, especially in young children. A moderate to severe 50 to 65 db conductive hearing loss can result from unilateral aural atresia, although 10% to 15% may have simultaneous sensorineural hearing loss. Testing of the non-atretic ear is necessary, as clinicians should not assume hearing is normal on that side.[8] Patients having squamosal chronic otitis media with microtia presents late and usually with complications. A computed tomography scan of the temporal bone is necessary to see the extent of the disease, grade aural atresia and assess candidacy for repair.

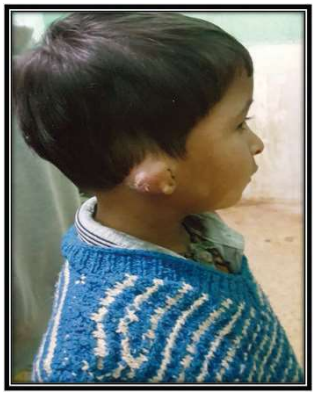
Management of microtia withsquamosal chronic otitis media requires a multi-disciplinary approach. Otologists take care of removing the unsafe disease and restore hearing as far as possible, plastic surgeons perform surgical reconstruction to improve ear appearance and function. Audiologists assess hearing function, provide rehabilitative services, and monitor auditory progress. Speech pathologists evaluate and treat speech and language disorders. Anaplastologists fabricate custom prostheses, collaborating closely with patients and the surgical team to create lifelike alternatives when surgical reconstruction is not suitable or preferred. Psychosocial support and counseling to address any emotional or social challenges associated with the condition.

Lastly, patients with microtia and their families must be educated regarding the treatment options available. Long-term follow-up with various specialists can help minimize the condition's physical and psychosocial tolls on the patient.

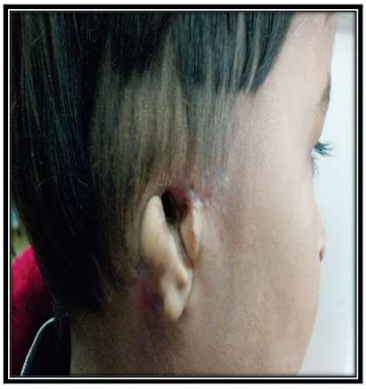
Discussion regarding the timing of interventions is crucial. Surgical intervention for comorbid conditions, such as aural atresia, must be considered.

CASE: 1

PRE-OPERATION

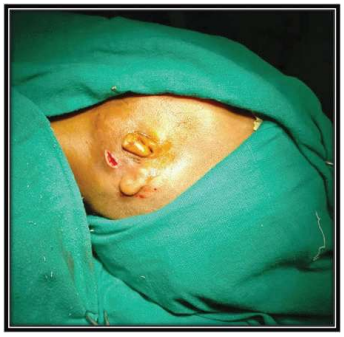


POST-OPERATION

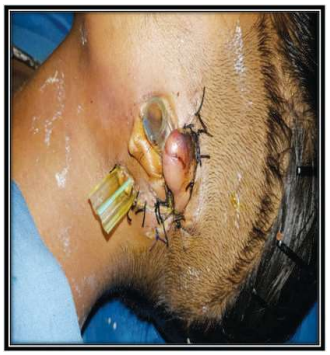


CASE: 2

PRE-OPERATION



POST-OPERATION



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