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BILATERAL MALLEUS ANKYLOSIS WITH STAPES FIXATION: A CASE REPORT

Authors: Zeesha Ahmad (1), Kriti Singh*(2)

Authors Affiliations: (1) Consultant E.N.T. Specialist (2) Senior Resident, Department of E.N.T.,

Charak Hospital and Research Center, Lucknow, Uttar Pradesh

ABSTRACT:

Introduction: We hereby report a case of bilateral malleus ankylosis with stapes fixation in a 9 year old female patient who presented with complain of bilateral hearing loss.

Clinically it is indicative of being a case of congenital conductive hearing loss, without vertigo or tinnitus, and with a normal otoscopic examination.

METHODS

HRCT Temporal bone is the main modality of visualization of ossicles in such cases. However the diagnosis is confirmed during surgery (exploratory tympanotomy).

The surgery involved right malleus head release with stapedotomy with malleus to vestibule prosthesis under general anaesthesia.

RESULT

The results were good with a gain of 21.66 Db. The patient was satisfied with no post operative complications.

DISCUSSION

The etiology of congenital malleus fixation includes predisposing anatomical and additional precipitating factors. The most

important predisposing factor seems to be a developmental anatomical anomaly of the epitympanic space which leaves the malleus in close contact with the walls.

CONCLUSION

Conductive hearing loss, unilateral or bilateral, needs a middle ear exploration. A pre-operative HRCT Temporal bone can be a good aid in identification of such cases. The diagnosis is however confirmed during surgery.

Key words: Malleus Ankylosis, Stapes Fixation, Ear Ossicles

INTRODUCTION

Osseous fixation of head of malleus was a little known entity until Mayer found it in 3 cases in year 1917(1). The advances in otologic surgeries, especially that of otosclerosis, attracted attention in this new area. Guilford pioneered its surgical treatment in 1961(2). Isolated malleus fixation was found a more common entity than malleus fixation with otosclerotic stapes fixation as per a study conducted by D. Katze and D. Plester in 1979(3) making this case report a unique one.

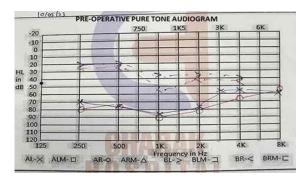
CASE REPORT

A 9 year old female attended ENT OPD with complains of bilateral decreased hearing which

was gradually progressive in nature. Otoscopic examination revealed bilateral intact tympanic membranes. There was no history of tinnitus, vertigo, ear discharge and allergies. There was no significant antenatal or postnatal history as given by parents.

The audiogram revealed a loss of 81 decibels(AC) in right ear and 78 decibels(AC) in left ear. Tympanometry was indicative of As type. HRCT Temporal bone revealed head of malleus completely fused to anterior epitympanum and slightly malformed incus in both ears.

PTA PRE OPERATIVE



HRCTTemporal Bone:

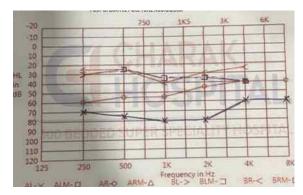




The patient was taken for right exploratory tympanotomy under general anaesthesia for right ear. Rosen's incision was given and tympanomeatal flap was elevated. Both malleus and stapes were found fixed. Incus was partially mobile but malformed. Atticotomy was done and malleus head was released from all bony ankylosis. Posterior overhang was drilled and stapes suprastructure was removed after cutting the stapedius tendon. Malleus to stapedotomy prosthesis was inserted. Soft tissue sealing of stapedotomy site was done. A tragal cartilage was used to reconstruct atticotomy site.

The post operative audiogram (performed 7 months later) revealed an improvement of 21.66 decibels.

POST-OPPTA:



DISCUSSION

The differential diagnosis of conductive hearing loss with intact tympanic membrane includes: middle ear anomalies, ossicular chain disruptions, generalized disorders of bone metabolism such as Paget disorder, osteogenesis imperfecta and so on (4).

Congenital etiology is mostly suspected in cases of decreased hearing based on the following findings (4):

- Absense of otologic history
- Healthy ears without inflammatory changes, with a normally developed pneumatic system
- The frequency of associations with other birth defects
- The existence of bilateral forms

.

In 1993 ,Teunissen and Cremers created a classification of minor malformations, based on the surgical approach, dividing them into four main groups: isolated stapes ankylosis, stapes ankylosis associated with other ossicular malformations, deformity of the ossicular chain with mobile stapes footplate and severe aplasia or dysplasia of oval or round window (5,6).

CLASS	MALFORMATIONS	%
1	Ankylosis or isolated congenital fixation of the stapes	30.6%
2	Stapes ankylosis associated with other malformations of the Ossicular chain: • Deformities of the incus and/or malleus, or aplasia of the long apophysis of the incus. • Bone fixation of the malleus and/or incus	38.1%
3	Congenital anomalies of the ossicular chain with mobile stapes Footplate: • Disruption of the ossicular chain • Epitympanic fixation • Tympanic fixation	21.6%
4	Congenital aplasia or severe dysplasia of the oval and round windows: • Aplasia • Dysplasia • Prolapse of facial nerve • persistence stapedial artery	9.7%

Charachon et al. proposed a classification of congenital middle ear anomalies based on their embryological origin⁽⁷⁾.

CLASS 0	Normal ossicular chain , almost normal	
	tympanic membrane but small atretic plate	
	around malleus handle	
CLASS 1	Fixation of the malleus head	
CLASS 2	Normal ossicular chain but fixation of the	
	footplate	
	2a: with abnormality of the facial nerve	
	2b: without abnormality of the facial nerve	
CLASS 3	Lack of a part of the ossicular chain with or	
	without abnormality of the stapes	
CLASS 4	Severe malformation of all the ossicular chain	

The current case seems to be most likely a case of congenital ossicular anomaly.

The etiology of congenital malleus fixation includes predisposing anatomical and additional precipitating factors. The most important predisposing factor seems to be a developmental anatomical anomaly of the epitympanic space which leaves the malleus in close contact with the walls(8,9,10,11). This was confirmed in a series where 68% of the patients had poorly pneumatized temporal bones(12). Developmental failures may also leave bony spiculae from the attic projecting in the direction of the ossicles. These bony spurs may also be due to post inflammatory new bone formation(12).

However in view of conductive hearing loss, the possibility of otosclerosis should be excluded as well to ensure proper treatment. Otosclerosis is the most common etiology of conductive hearing loss in 15–20 years old patients with intact tympanic membrane(13). A thorough examination of all ossicles is must during exploration of middle ear even in cases of otosclerosis to exclude any accompanying ossicular anomaly.

CONCLUSION

Conductive hearing loss, unilateral or bilateral, needs a middle ear exploration. It can be otosclerosis only or otosclerosis associated to malformations of middle ear area (ossicular chain anomalies, fixation of head of malleus). The diagnosis is confirmed during surgery. Proper palpation of all ear ossicles should be done even in cases of otosclerosis as quite a generous number of times stapedotomies do not result in proper hearing improvement due

to co-existing malleus ankylosis. A preoperative HRCT Temporal bone can be a good aid in identification of such cases.

DECLARATION

Conflict of Interest: There was no conflict of interest.

Ethical Approval: The study was approved by institutional ethics committee.

Consent: Written and informed consent were taken while doing the study. No human or animal was harmed in the study.

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*Corresponding Author:

Dr. Kriti Singh (MS)

Mailing address: senior resident, Tondan Marg, near Safed Masjid, Dubagga, Lucknow, Uttar Pradesh 226003

Mail: kritichoti123456@gmail.com

Contact Number-7905490822

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