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CASE REPORT

ISOLATED CONGENITAL CHOLESTEATOMA OF THE MASTOID PROCESS: A CASE REPORT

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ABSTRACT: Congenital Cholesteatomas have historically been considered a rare disorder, an incidence ranging from 4 to 24%. The most common sites of presentation on physical examination are Anterior superior and posterior superior quadrants of the tympanic membrane; conductive hearing loss is the most common presenting symptom. There is a lack of uniformity of reporting and classifying congenital cholesteatomas. Recidivism of the lesion appears more commonly with posterior superior congenital cholesteatomas. Treatment of Congenital Cholesteatoma is still surgical. We are here presenting a case of isolated congenital cholesteatoma in Mastoid process treated with modified radical mastoidectomy and tympanoplasty, patient followed up to one year no recurrent was found, hearing also improved, heightened awareness and early diagnosis of congenital cholesteatoma is imperative, early treatment decreases the extent of the disease and reduces the risk of recidivism and complications.

KEYWORDS: Cholesteatoma, epidermoid cyst, petrous apex, keratin debris, cholesterol granuloma, mastoidectomy, congenital.

INTRODUCTION: Congenital cholesteatoma arises from the embryonic epidermal crest, is a benign disease with slow progressive growth that destroys neighboring structures, it is considered an epidermal cyst originating from the remnants of squamous keratinized epithelium, the disease may appear in several regions of the temporal bone such as in the middle ear (most frequent site) as well as in the petrous apex, cerebellopontine angle, external acoustic meatus and mastoid process, congenital cholesteatoma of the mastoid process is the rarest form of presentation in the temporal bone, only 2 to 4% of cholesteatoma presenting to pediatric otologist are congenital in origin. It is well accepted that congenital cholesteatoma behave in a more aggressive manner than the acquired form, keratin filled cysts that grow medial to the tympanic membrane, result either from birth abnormality, trauma or metaplasia, and if they fulfill the following criteria they are considered to be congenital:

- Mass medical to the tympanic membrane.
- Normal tympanic membrane.
- No previous history of ear discharge perforation or ear surgery.

CASE REPORT: A 40 years old female patient, presented to our OPD with complaint of pain in the left ear since 02 years and minimal decrease in hearing, there was no ear discharge, vertigo, tinnitus or facial asymmetry, on otoscopic examination of the left ear revealed. (Fig No.1)

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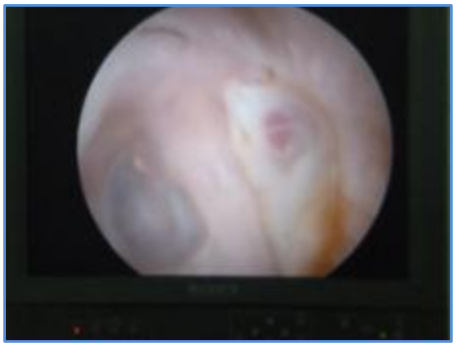

Fig. 1: Endoscopic Picture

An intact, dull tympanic membrane and granulations seen in the posterior superior wall, the tuning fork test revealed a conductive hearing loss on the left side, pure tone audiogram shows moderate conductive hearing loss with an air bone gap of 25dB, normal hearing on right side. High resolution computed tomography (HRCT) of the temporal bone was performed and shows soft tissue density in left mastoid air cells. (Fig No.2 & 3)

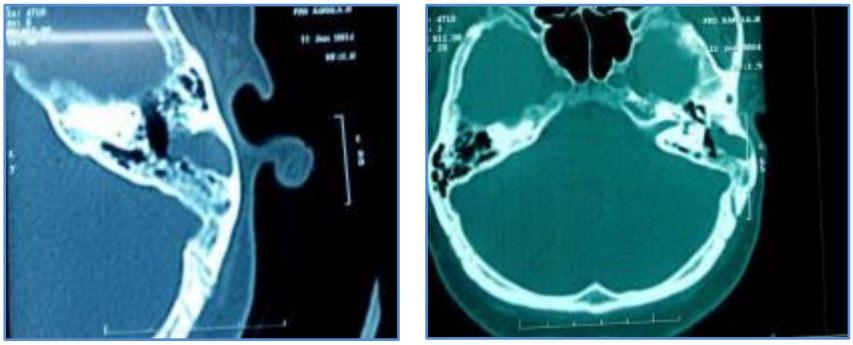

Fig. 2 & 3: CT Scan Photos

After all necessary investigations she was subjected to elective mastoid exploration under general anesthesia, tympanic membrane intact few granulations at posterior superior wall, incus was eroded, cholesteatoma sac present in the tip of the mastoid modified radical mastoidectomy was done with clearance of all the cholesteatoma. (Fig No.4 & 5)

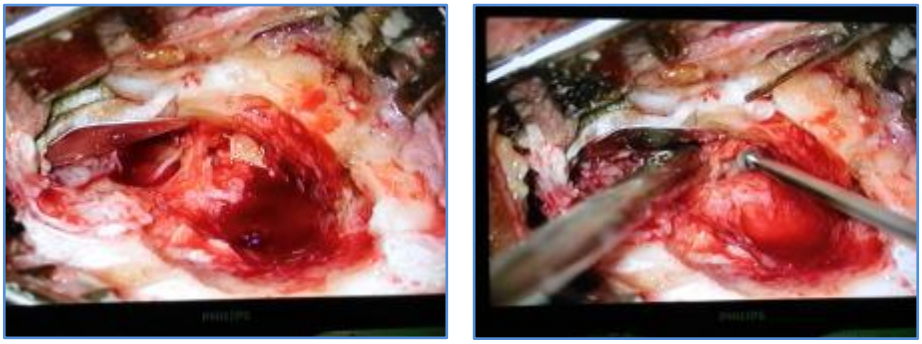

Fig. 4 & 5: Intra operative photos

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Cartilage graft kept in between the malleus and head of the stapes, temporalis fascia graft placed over the ossicles, ribbon gauze soaked with BIPP packing was inserted, kept for 2 weeks, wound was closed in layers after wide meatoplasty, mastoid bandage for 05 days, suture removal done on 7th day, wound was healthy patient was followed for 12months, no more ear discharge and considerable improvement in hearing also seen, the results of the histological analysis confirmed cholesteatoma. (Fig No.6 & 7)

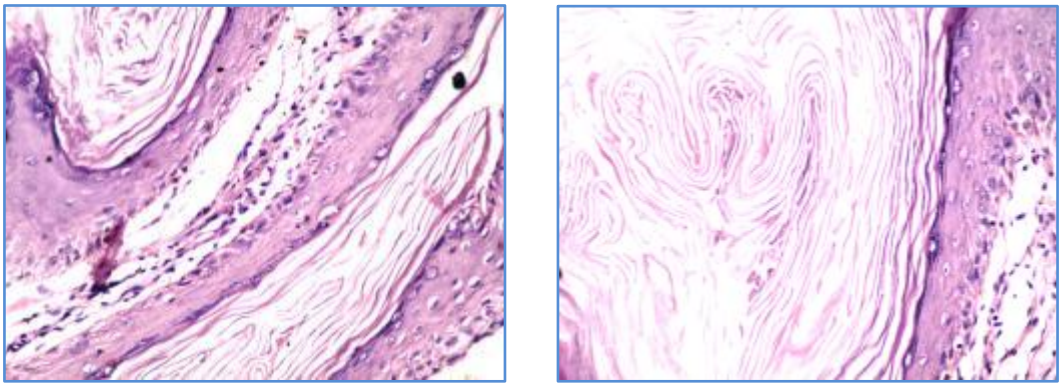

Fig. 6 & 7: Cholesteatoma Photos

DISCUSSION: Congenital cholesteatoma is a rare entity, arising from aberrant epithelial remnants left at the time of closure of the neural groove between the third and fifth week of fetal life,[1] it is also called epidermoid cyst or primary cholesteatoma, is lined with stratified squamous epithelium and filled with debris (keratin and cholesterol crystals) originating from the progressive desquamation of the epithelium, congenital and acquired cholesteatoma histologic finding are similar, thus absence of previous otologic disease and normal tympanic membrane are necessary to consider a cholesteatoma to be congenital.[2]

In 1953, house HP[3] was the first to describe a cholesteatoma behind a intact membrane, while in 1991 proctor B[4] reported that congenital cholesteatoma originated from the same ectoderm which forms a primitive notochord and that embryonic cell remnants of this ectodermic structure can occur in any cranial bone, It is well accepted that congenital cholesteatoma behave in a more aggressive manner than the acquired form the most common presenting complaint is hearing reduction as the disease progress a patient may present with vertigo, facial nerve palsy or intracranial abscess, early diagnosis decreases the size of congenial cholesteatoma and subsequently decreases the incidence of complications, early detection is possible in a routine audiological or otological screening.

There are several theories on the patho physiology of congenital cholesteatoma of the middle ear include presence of congenital cell rest, metaplasia of middle ear epithelium and papillary in growth through an intact tympanic membrane, research reports depict that the most common location are anterior superior quadrant followed by posterior superior quadrant of the tympanic membrane, koltia et al. (2002) reported that most cholesteatoma start out as a matrix enclosed spherical keratin pearl in the anterior superior quadrant accordingly, the pearl then grows like a round inflated balloon. Anterior growth is towards the Eustachian tube, inferiorly towards the hypotympanum and posteriorly towards the handle of the malleus.

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Posterior growth may extend further, involving the incudostapedial joint and the stapes suprastructure, as well as up towards the incudomalleolar joint, around the incus, and into the attic. The growth of congenital cholesteatoma may progress from the attic into the antrum and then into the mastoid and follows the pattern of the enlargement of other categories of middle ear cholesteatomas, the only studies found in the literature similar to the present case were conducted by Luntz M,[5] Mevio E,[6] in 2002. Derlacki & Clemis JD. [7]

Congenital cholesteatoma is a persistent disease, once the diagnosis is made the standard treatment is to surgically remove the growth,[8] the challenge of cholesteatoma surgery is to permanently remove the cholesteatoma whilst retaining or reconstructing the normal function of the structures housed within the temporal bone,[9] the general objective of cholesteatoma surgery has two parts, it is both directed against the under lying pathology and directed towards maintaining the normal functions of the temporal bone these aims are conflicting and this makes cholesteatoma surgery extremely challenging the variation in technique in a surgery results from each surgeons judgment whether to retain or remove certain structures housed within the temporal bone in order to facilitate the removal of cholesteatoma, this typically involves some form of mastoidectomy[10] which may or may not involve removing the posterior ear canal well and the ossicles, clearly preservation and restoration of ear function at the same time as total removal of cholesteatoma requires a high level of surgical expertise, if the disease is difficult to remove so that there is an increased risk of residual disease, then removal of involved ossicles in order to fully clear cholesteatoma has generally been regarded as necessary and reasonable, it is important that the patient attend periodic follow up checkups, because even after careful microscopic surgical removal, cholesteatoma may recur such recurrence arise many years or even decades after treatment.

CONCLUSION: Early detection of congenital cholesteatoma is crucial as it may limit the size of retro tympanic and reduce the possible risks and complications from surgery as well as from the disease itself, a high index of suspicion in a patient with reduced hearing and an intact tympanic membrane may help in the diagnosis of congenital cholesteatoma, in our case the congenital cholesteatoma was presented in mastoid process, which is the rarest presentation in Temporal bone after surgery no recurrence was found.

REFERENCES:

1. McGill T J, Merchant S, Healy G B, Friedman E M. Congenital cholesteatoma of the middle ear in children: A clinical and histopathological report. Laryngoscope. 101: 606-613.
2. Levenson M J, Parisier S C, Chute P, Wenig S, Jarbe C. A review of twenty congenital cholesteatoma of the middle ear in children. Otolaryngol Head Neck Surg. 15: 169-174.
3. House HP. An apparent primary cholesteatoma: case report. Laryngoscope. 1953, 63 (8): 712-3.
4. Proctor B. Chronic otitis media in mastoiditis. In: Paparella MM, Shumrick DA (eds). Otolaryngology. Vol.2: The Ear. 3rd ed. Philadelphia: WB Saunders; 1991 p. 1366-8.
5. Luntz M, Telischi F, Bowen B, Ress B, Balkany T. Imaging case study of the month. Congenital cholesteatoma isolated to the mastoid. Ann Otol Rhino Laryngol. 1997; 106 (7 Pt 1): 608-10.
6. Mevio E, Gorini E, Sbrocca M, Artesi L, Lenzi A, Lecce S, et al. Congenital cholesteatoma of the mastoid region. Otolaryngol Head Neck Surg.2002; 127 (4): 346-8.
7. Derlacki EL, Clemis JD. Congenital cholesteatoma of the middle ear and mastoid. Trans Am Otol Soc.1965; 53: 208-31.

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8. Rizer F M, Luxford W M. The management of congenital cholesteatoma: surgical result of 42 cases. 1998. Laryngoscope. 98: 254-256.
9. Hammel J P. Congenital cholesteatoma: classification, management and outcomes. Arch Otolaryngol Head Neck Surg. 128: 810-814.
10. Rashad U, Hawthorne M, Kumar U, Welsh A. Unusual cases of congenital cholesteatoma of the ear. J Laryngol Otol.1999; 113 (1): 52-4.

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