ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 03 | March - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

"MANAGEMENT OF A RARE CASE OF CONGENITAL CHOLESTEATOMA"

ENT

Dr. Sudhagar M E* MS (ENT), DNB, Senior Resident, Department Of ENT, Lady Harding Medical College And Hospital. *Corresponding Author.

Dr. Poornima MS (ENT), DNB, Senior Resident, Department Of ENT, Lady Harding Medical College And Hospital.

ABSTRACT

INTRODUCTION: Congenital Cholesteatoma incidence is increasing recently. Most of the times, it is an incidental finding. Sometimes patients can come with complaints of otalgia or hard of hearing. Examination reveals a white mass seen behind an intact tympanic membrane. Any previous history of ear discharge cannot rule out the possibility of Congenital Cholesteatoma.
DESIGN: Case Report
RESULT: We report a case of Congenital Cholesteatoma in the posterosuperior quadrant. Patient had a previous history of ear discharge but on examination there was an intact tympanic membrane with white mass behind it. CT scan helps in assessing the extent of the disease. Surgical removal is the treatment of choice. It depends on the extent of the disease.
CONCLUSION: Due to the variability in the clinical presentation, high index of suspicion is needed to diagnose. Early surgical excision should be done. In extensive diseases, canal wall down mastoidectomy may be needed. As these patients are mostly in pediatric age group, Intact Canal Wall Mastoidectomy. Can avoid mastoid cavity and its problems.

KEYWORDS

Congenital Cholesteatoma, Intact Canal Wall Mastoidectomy, Tympanoplasty, Tympanotomy.

INTRODUCTION

Cholesteatoma is a cystic sac like structure filled with keratin debris. It is classified as congenital and acquired types. Congenital cholesteatoma is a rare clinical presentation but its incidence is increased recently. 2 to 5 % of all cholestatoma is congenital[1]. This keratin filled cystic sac present from birth can expand and cause bone erosion. It arises from the epithelial cell rests or due to metaplasia of normal epithelium[2]. Congenital cholesteatoma can also be seen in ceerebellopontine angle or the middle cranial fossa[3]. In 1885, Luchae first described congenital cholesteatoma[4]. Derlacki and clemis in 1965 came up with the definition of congenital cholesteatoma[5]. It is defined as pearly white mass behind an intact tympanic membrane with no history of prior otorrhea, tympanic membrane perforation, or previous otologic procedures. Levenson later described in 1986, that previous otitis media cannot be used as to exclude congenital cholesteatoma as it is a very common infection in children[6].

Congenital cholesteatoma can arise anywhere in the middle ear or mastoid cavity but typical presentation is in the anterosuperior quadrant[7,8,9]. There is a variation in the age of presentation and the severity of the disease[7]. Conductive hearing loss, otalgia, tinnitus can be the presenting complaints or they can present with complications. Mostly congenital cholesteatoma is an incidental finding. Very rarely congenital cholesteatoma can expand and cause tympanic membrane perforation which makes it difficult to differentiate from acquired lesions. Early surgery should be planned to avoid complications. Surgery should be planned according to the extent of disease. Various approaches can be used. Transcanal tympanotomy and removal of the disease can be done with low rate of recurrence in initial stages. In this case report, we present a case of congenital cholesteatoma in posterosuperior quandrant and extending into the mastoid region which was managed by Intact Canal Wall Mastoidectomy.

CASE REPORT

9 year old boy presented with history of decreased hearing and pain in right ear on and off for 6 months. He had one episode of ear discharge 2 months back which was managed conservatively. There was no history of tinnitus and vertigo. On examination, there was pearly white mass which was seen behind the intact tympanic membrane in the posterosuperior quadrant. The other ear was normal. Tuning fork test was done. Rinne's test was negative on the right side and weber lateralized to the diseased ear. Pure tone audiometry showed mild conductive hearing loss in the right ear. High Resolution Computed Tomography of the temporal bone was done. It revealed soft tissue density in the epitympanic region extending to mastoid and the tympanic membrane found to be intact. There is no blunting of scutum seen (Figure 1).

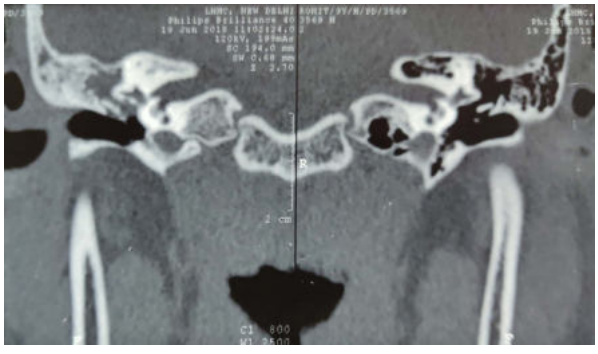

Figure 1. Soft tissue density in middle ear with intact tympanic membrane. No blunting of scutum seen.

Patient underwent mastoid exploration. First exploratory tympanotomy was done. Once tympanomeatal flap was elevated pearly white mass was found in the posterosuperior quadrant (Figure 2).

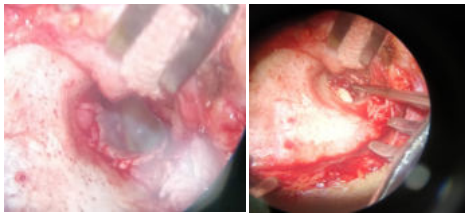

Figure 2. Intact tympanic membrane with White mass seen in posterosuperior quadrant

Intact Canal Wall Mastoidectomy was done and cholesteatoma sac completely removed from attic, antrum and the posterosuperior quadrant of pars tensa (Figure 3).

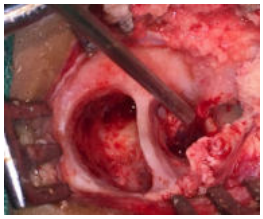

Figure 3. Intact Canal Wall Mastoidectomy and excision of cholesteatoma.

Ossicular chain was found to be intact and tympanoplasty was completed. Postoperative period was uneventful. Patient was discharged on postoperative day one and there were no complications. Patient is under close watch for any recurrence.

DISCUSSION

Congenital cholesteatoma is a rare clinical presentation. But its incidence is on the rise because of the awareness and high clinical suspicion. Most common age of presentation is 5-7 years. Kojima[10] found that the mean age of presentation was 13.3 years. Male patients are commonly affected than females. The diagnosis of congenital cholesteatoma depends on the clinical finding of a white mass behind intact tympanic membrane. According to Derlacki and Clemis there should be no previous history of otorrhea or tympanic membrane perforation or previous ear procedures. Levenson added that previous ear discharge cannot rule out congenital cholesteatoma as otitis media is a very common problem in this age group. In this case, we found that the patient had an episode of ear discharge which was managed with topical and oral antibiotics.

Origin of congenital cholesteatoma still remains controversial. Teed in 1936, proposed the epithelial cell rest theory, which is most commonly accepted[11]. This was supported by the study of Michael[12]. Anterosuperior quadrant is most common site. It can also be seen in posterosuperior quadrant[13,14]. Parisier et al[15] suggested the possibility of migration of epidermoid cyst to posterosuperior region. Posterior quadrant cholesteatoma can attain larger size that sometime it can cause disruption of tympanic membrane. This can lead to difficulty in making a diagnosis of congenital cholesteatoma. Sade et al supported the theory of squamous metaplasia of middle ear mucosa[16]. Cholesteatoma expands due to the accumulation of keratin debris within the cyst. It can extend into the mastoid antrum and cause bony erosion due to the enzymatic process by cytokine mediated inflammation[17]. Ossicular erosion can occur especially in the posterosuperior quadrant congenital cholesteatoma causing conductive hearing loss. Early diagnosis and surgery can prevent the ossicular erosion. In this case the cholesteatoma was seen over the ossicles but the continuity was still maintained.

Patient might present with complaints of hearing loss and ear discomfort on and off. Kojima et al suggested that 60.3% of their patients complained of hearing loss and 17.5% of the patients underwent otologic examination for previous ear discharge episode as in this case[10]. Tagaki concluded that early stage congenital cholesteatoma were diagnosed incidentally and late stage patients had hearing loss[18]. Diagnosis can be made on clinical grounds. Pure tone audiometry will reveal the conductive hearing loss. CT scan of the temporal bone can help in assessing the extent of the disease and complications if any.

Surgery is the treatment of choice for congenital cholesteatoma. Main goal is to eradicate the disease and establish healthy aerated ear. It depends on the location of cholesteatoma and involvement of ossicles. In children canal wall up procedure is preferred. Tympanotomy can be used to remove small anterosuperior quadrant cholesteatoma. Large posterosuperior cholesteatoma can involve ossicles and mastoid exploration will be needed. In this case, CT suggested involvement of mastoid region. Intact Canal Wall Mastoidectomy was done. Cholesteatoma was seen in mastoid antrum. Disease was removed and ventilation between attic and antrum was established with intact posterior canal wall. The prognosis depends on the extent of the disease, age of the patient and involvement of ossicles.

CONCLUSION

Surgery is the ideal treatment option for cholesteatoma. Early diagnosis of congenital cholesteatoma can prevent extensive disease and complications. High degree of clinical suspicion is needed to make early diagnosis. Incidence of congenital cholesteatoma is increased due to the awareness about the disease. Anterosuperior quadrant is the most common location. Disease in posterosuperior quadrant can have a large extension with ossicular destruction. CT scan should be considered to look for the extent of the disease. Surgery should clear the cholesteatoma with restoration of normal ventilation of the ear and improvement in hearing. Canal wall up technique should be considered in paediatric patients. If disease clearance is inadequate then canal wall down technique can be preferred. Long term follow up is needed in these patients.

REFERENCES

1. Paparella M, Rybak L (1978) Congenital cholesteatoma. Otolaryngol Clin North Am 11:113–120.
2. Bennett M, Warren F, Jackson GC, Kaylie D (2006) Congenital cholesteatoma: Theories, facts, and 53 patients. Otolaryngol Clin North Am 39(6):1081–1094.
3. Robert Y, Carcasset S, Rocourt N, et al.: Congenital cholesteatoma of the temporal bone: MR findings and comparison with CT. Am J Neuroradiol 1995, 16:755–761.
4. Karmody CS, Byhatt SV, Blevins N, et al.: The origin of congenital cholesteatoma. Am J Otol 1998, 19:292–297.
5. Derlacki EL, Clemis GD: Congenital cholesteatoma of the middle ear and mastoid. Ann Otol Rhino Laryngol 1965, 74:706–727.
6. Levenson MJ, Parisier SC, Chute P: et al. A review of twenty congenital cholesteatomas of the middle ear in children. Otolaryngol Head Neck Surg 1986,15:169–174.
7. Potsic WP, Samadi DS, Marsh RR, et al. A staging system for congenital cholesteatoma. Arch Otolaryngol Head Neck Surg 2002; 128:1009–1012.
8. Lee TS, Liang JN, Michaels L, et al. The epidermoid formation and its affinity to congenital cholesteatoma. Clin Otolaryngol Allied Sci 1998; 23:449–454.
9. Lee JH, Hong SJ, Park CH, et al. Congenital cholesteatoma of mastoid origin. J Laryngol Otol 2007; 121:e20.
10. Kojima H, Tanaka Y, Shiwa M, et al. Congenital cholesteatoma clinical features and surgical results. Am J Otolaryngol. 2006;27(5):299–305.
11. Teed RW: Cholesteatoma verum tympani: its relationship to the first epibranchial placode. Arch Otolaryngol 1936, 24:455–474.
12. Michaels L. Origin of congenital cholesteatoma from a normally occurring epidermoid rest in the developing middle ear. Int J Pediatr Otorhinolaryngol. 1988;15:51-56.
13. Tos M: A new pathogenesis of mesotympanic (congenital) cholesteatoma. Laryngoscope 2000, 110:1890–1897.
14. Zappia JJ, Weit RJ: Congenital cholesteatoma. Arch Otolaryngol Head Neck Surg 1995, 121:19–22.
15. Parisier SC, Levenson MJ, Edelstein DR, Bindra GS, Han JC, Jolitsky JN. Management of congenital pediatric cholesteatomas. Am J Otol. 1989;10:121-123.
16. Sade J, Babiacki A, Pincus G. The metaplastic and congenital origin of cholesteatoma. Acta Otolaryngol. 1983;96:119-129.
17. Seaman MT, Cliff A (2006) Pathophysiology of cholesteatoma. Megerian Otolaryngol Clin North Am 39(6):1143–1159.
18. Tagaki T, Gyo K, Hakuba N, et al. Clinical features, presenting symptoms, and surgical results of congenital cholesteatoma based on Potsic's staging system. *Acta Otolaryngol*. 2014;134(5):462–7.