International Journal of Otorhinolaryngology and Head and Neck Surgery
Khavasi P et al. Int J Otorhinolaryngol Head Neck Surg. 2018 Jul;4(4):1017-1022
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Original Research Article

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20182705

Acquired cholesteatoma in children: presentation, complications and management

Prabhu Khavasi, Karra Bhargavi*, Santosh P. Malashetti, Yasha C.

Department of ENT, S. Nijalingappa Medical College and HSK Hospital, Navanagar, Bagalkot, Karnataka, India

Received: 11 April 2018
Revised: 25 May 2018
Accepted: 26 May 2018

***Correspondence:**
Dr. Karra Bhargavi,
E-mail: drkbhargavi28@gmail.com

ABSTRACT

Background: Acquired cholesteatoma in children is an aggressive disease due to its rapid growth and high recurrence rate. The objective of the study was to assess clinical features of cholesteatoma in children and evaluate our experience in the overall management of this disease.
Methods: This is a retrospective study of 20 children aged 6-15 years operated on for acquired middle ear cholesteatoma from June 2016 to December 2017. An analysis was made about the clinical and operative findings, surgical approaches and the overall management of complications. The data were then compared with the relevant and available literature.
Results: Majority of the children presented with complaints of otorrhoea (100%), decreased hearing (90%), otalgia (50%), complications (25%) and most of them (85%) were operated by canal wall down mastoidectomy technique (CWD).
Conclusions: Canal wall down mastoidectomy is the optimal management technique for adequate exposure and removal of cholesteatoma in paediatric population.

Keywords: Cholesteatoma, Cholesteatoma in children, Mastoidectomy

INTRODUCTION

Acquired cholesteatoma in children is an aggressive disease due to its rapid growth and high recurrence rate.[1,2] The most frequently encountered symptoms include otorrhoea, decreased hearing and ear ache with post auricular swelling, fistula, head ache and fever in case of complications. Surgical management aims to eradicate the disease process, prevent recurrences and preserve auditory function thus preventing learning difficulties.[3] The choice of adequate surgical technique, either canal wall down (CWD) or canal wall up mastoidectomy (CWU) must be determined for each individual case according to the extent of cholesteatoma.[2,4] The aim of this study is to assess the clinical features of cholesteatoma in children and evaluate our experience in the surgical management of this disease.

METHODS

We present a retrospective descriptive study from June 2016 to December 2017 reviewing a total of 20 children aged 6–15 years, operated on for acquired middle ear cholesteatoma in the department of ENT, SNMC & HSK Hospital, Bagalkot.

The data of all patients were reviewed containing the medical history, complete ENT examination including otoscopic and microscopic examination of the ear to evaluate the extension of the disease and the status of the

ossicular chain. Pre operatively, all patients underwent HRCT temporal bone and pure tone audiometry. Ophthalmological and neurological consultation done in complicated cases. Selection of the applied surgical technique (CWU or CWD mastoidectomy) was made according to the pre and intra operative assessment. CWD technique was performed, depending on anatomical conditions, when cholesteatoma was hardly fully controllable with a CWU technique. Results were tabulated using Microsoft excel version 2010.

Inclusion criteria

Inclusion criteria were all children in the age group of 1-15 years with CSOM - attico antral disease were included in the study; children willing for further surgical management were included.

Exclusion criteria

Exclusion criteria were children with ASOM were excluded from the study; children with intracranial complications not related to CSOM were excluded from the study.

Children not fit/not willing for surgery were not included in the study.

RESULTS

Middle ear surgery was done in 20 patients making a total number of 20 operated ears. For all cases primary surgery was done in our institution.

Mean age at the first operation was 10.5 years ranging from 6 to 15 years with a male predominance (sex ratio 3:2). Among the cases 6 had bilateral cholesteatoma but only one ear was operated. There was no history of eardrum trauma.

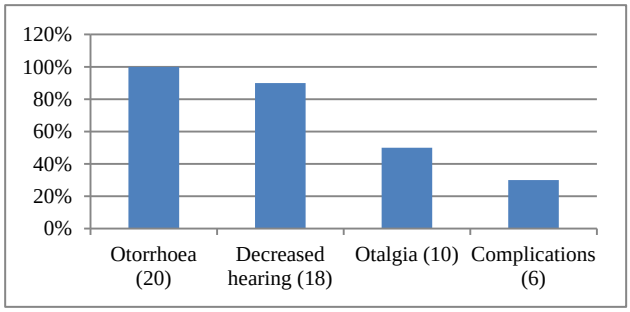

Figure 1: Distribution of cases (%) according to the frequency of presenting complaints.

The main symptoms reported by our patients were chronic otorrhoea (100%), hearing loss (90%) and otalgia (50%) (Figure 1). Five children presented with a complication revealing the disease: acute mastoiditis (3 cases), facial palsy and labyrinthitis (1 case) and meningitis (1 case).

Preoperative otoscopy revealed pars flaccida retraction in 35%, attic perforation in 20%, aural polyp in 20%, cholesteatoma in 15%, and pars tensa perforation in 10% (Table 3).

Abnormalities in the contralateral ear were observed in 8 ears (40%): attic retraction (4 cases), tympanic perforation (3 cases) and both attic retraction and tympanic perforation in 1 case. Nasal endoscopy showed a non-obstructive septal deviation in 5 cases and adenoid vegetations in 7 cases.

Preoperative audiometry revealed a mean air conductive hearing loss of 45 dB in 19 patients. A mixed hearing loss was found in 1 patient with a mean ABG of 30 dB.

On CT scan, cholesteatoma extended to the epitympanum in 12 cases (60%), to the mastoid in 19 cases (95%) and to the window region in 7 cases (35%). Ossicular chain was complete in 50% and eroded in 50%. The incus was the ossicle most frequently lysed followed by the malleus and the stapes. CT also revealed complications including a facial canal lysis in 2 cases, a labyrinthine fistula in 1 case and meningitis in another.

Intraoperatively sites involved by cholesteatoma are detailed in Table 5. Ossicular chain was disturbed in 17 cases (85%). The ossicle most commonly destroyed was found to be incus (80%), followed by the malleus (75%) and the stapes (30%).

All the patients underwent single stage operation. CWD mastoidectomy is the preferred operative technique in our centre, performed in 85% of the cases (n=17). It was advocated to the following findings: extension of cholesteatoma into the attic (10 cases), to the retrotympanum (6 cases) and meningeal protrusion (1 case). Considering these anatomical conditions, cholesteatoma was found to be hardly controllable with a CWU technique.

CWU technique was performed in 3 cases (15%). It was indicated for patients with a large well pneumatized mastoid and a well-controlled disease. It was combined with a mastoidectomy. One child had a large mesotympanic cholesteatoma extending to the protympanum, responsible for a labyrinthine fistula of lateral semicircular canal. A CWD mastoidectomy helped to control the disease, remove the matrix completely and seal the fistula.

Ossicular reconstruction was possible in 15% of the cases. Ossicles were removed in 25% of the cases (incus in 15% and head of malleus in 10%). Reconstruction was carried out by means of columellization with shaped autologous ossicles or cartilage. There were no cases of sensorineural hearing loss or facial nerve injury after surgery in our series.

Table 1: Distribution of children according to age and sex.

Age Group	Gender				Total	
	Male		Female			
	Number	%	Number	%	Number	%
6-10 years	7	35	3	15	10	50
11-15 years	6	30	4	20	10	50

Table 2: Distribution of children according to the frequency of presenting complaints.

Sl. No	Variables	Frequency
1.	Otorrhoea	20
2.	Decreased hearing	18
3.	Otalgia	10
4.	**Complications:**	
	Swelling behind the pinna	3
	Fever, headache, vomiting	1
	Facial weakness	1
	Vertigo	1

Table 3: Distribution of cases according to otoscopy findings.

Otoscopy findings	Number	Percentage (%)
Retraction pocket in postero-superior quadrant	7	35
Perforation		
Attic	4	20
Anterior	2	10
Polyp	4	20
Cholesteatoma	3	15

Table 4: Distribution of cases according to cholesteatoma extension on HRCT temporal bone.

HRCT finding	Number	Percentage (%)
Epitympanum	12	60
Mesotympanum	20	100
Mesotympanum + Mastoid	19	95
Mesotympanum + Window region	7	35

Table 5: Distribution of cases according to intraoperative findings.

Cholesteatoma extension	Number	Percentage
Attic + Mesotympanum	11	55
Retrotympanum/Stapes/Window region	6	30
Attic + Mastoid	11	55
Attic + Mesotympanum + Mastoid	6	30

Table 6: Distribution of cases according to complications.

Complication	Number	Percentage (%)
Intra temporal		
Post auricular abscess	3	15
Post aural fistula	1	5
Labyrinthitis	1	5
Facial palsy	1	5
Intra cranial		
Meningitis	1	5

DISCUSSION

Retraction pocket of the tympanic membrane, resulting from Eustachian tube dysfunction is the most common cause of acquired cholesteatoma in children.[5] These pockets usually develop in pars flaccida or upper posterior region of the pars tensa.[5,6] The mean age at onset varies between 9 and 13 years.[3,7]

Many authors believe that cholesteatoma is more aggressive in children than in adults. Bujia analyzed the expression of MIBI (a monoclonal antibody marker of cell proliferation) in a child and an adult cholesteatoma and found a higher proliferation rate in children.[8] Mobeen also confirmed a significantly higher proliferative rate of the cholesteatoma matrix in this group.[9] In addition, Dornelles showed that pediatric specimens expressed higher levels of matrix metalloproteinases and exhibited an exaggerated inflammatory profile.[10]

The most common presenting symptom is otorrhea.[11] Physical findings associated with cholesteatoma include abnormal appearance of the tympanic membrane, otorrhoea, aural polyp, and hearing loss. The diagnosis is made on otoscopic examination and often requires visualization of the ear with a binocular microscope. A crust overlying the posterior superior quadrant of the tympanic membrane should raise the suspicion of underlying cholesteatoma. The presence of a defect in the tympanic membrane (i.e., perforation or retraction) associated with pearly white squamous debris is diagnostic of secondary or acquired cholesteatoma; however, the physical findings may be more subtle, with a bland retraction pocket or aural polyp. Shallow retraction pockets are pathologic and should be followed carefully.[12]

Hearing assessment should be performed on all children with cholesteatoma. Age-appropriate audiologic evaluation should be performed after treatment of any acute infection. Ideally, ear-specific air and bone conduction thresholds should be obtained.[12]

Temporal bone imaging may be helpful in delineating the extent of the lesion. High-resolution CT scans of the temporal bone, in both the axial and coronal projections, provide anatomic information regarding the status of the ossicles, involvement of the labyrinth, course of the facial nerve, and size of the mass. The middle ear should be as healthy as possible at the time of the scan because fluid is not distinguishable from soft tissue. One of the characteristic radiological changes associated with acquired cholesteatoma is erosion of the scutum (i.e., the postero-superior lateral wall of the middle ear space) which is best seen in the coronal view.[13] This finding, when present, is highly suggestive of cholesteatoma. MRI is indicated for initial assessment of extension, in case of meningeal contact and also during follow up when CT scan is in doubt.[14]

As pediatric cholesteatoma often involves the entire mastoid and mesotympanum, surgery is more difficult than in adults and results are considered to be poorer.[5] It aims to eradicate the disease, preserve or improve hearing and prevent recurrence or residual disease.[3] The two main types of tympanomastoidectomy that are used are canal-wall up (CWU) and canal wall down (CWD). The canal wall here refers to the posterior wall of bony EAC that separates EAC from the mastoid. CWU mastoidectomy preserves the normal anatomy of the canal so that postoperative care is simple. This surgical approach necessitates complete removal of all squamous epithelium from the middle ear and mastoid. Communication between the middle ear and mastoid can be established through the antrum and can be augmented by opening the facial recess when required. Children undergoing CWU mastoidectomy usually requires a "second-look" procedure to remove the recurrent or persistent disease.

CWD or modified radical mastoidectomy results in a mastoid cavity which communicates with the external meatus. This approach affords improved visualization of the antrum and epitympanum with the intent of managing the cholesteatoma with a single surgical procedure. Part of the cholesteatoma matrix can be left to epithelialize the cavity. The disadvantage of this approach is that the cavity tends to be large in children, requires a close surveillance, often for a lifetime, to remove accumulated debris if any. A meatoplasty, which means creating an enlarged opening in the outer ear, is performed at the same time to facilitate postoperative care. The complications of ear surgery include hearing loss, facial nerve palsy, cerebrospinal fluid leak, and tympanic membrane perforation, in addition to the general risks of bleeding and infection. The most important risk is that of recurrent or persistent cholesteatoma in children, regardless of the initial surgical approach.[15,16] Significant predictors for recurrence include involvement of the oval window niche, sinus tympani, and ossicular erosion.[15-17] Although some authors prefer routine CWD mastoidectomy for management of childhood cholesteatoma, most authors believe that the surgical approach must be tailored to the patients' disease.[18]

On the other hand, CWD mastoidectomy is reserved for large cholesteatoma that cannot be adequately removed by CWU technique, children with a small mastoid, limited air cells or meningeal prolapse and patients with poor follow up.[1,3,9,19,20] It is associated with a lower recurrence rate, varying from 13% to 17%.[7,17,20-22] However, factors that should be kept in mind for choosing the best surgical technique in pediatric cholesteatoma include: intraoperative view of disease extension, mastoid pneumatisation, mucosal and ossicular chain conditions, which can be only supposed with the preoperative radiological scans, and Eustachian tube function.

In our study, CWD mastoidectomy was performed in 14 (70%) patients and CWU mastoidectomy in 2 (10%) patients. 2 children (10%) with post auricular abscess were subjected to incision and drainage and then taken up for CWD mastoidectomy. One child with post aural abscess and fistula underwent excision of the fistulous tract and then managed accordingly. One child with intra cranial complication i.e., meningitis was treated with IV antibiotics and then subjected to CWD technique.

Intra operatively, we found a cholesteatoma invading the window region (2 cases), sinus plate (3 cases), dural plate (2 cases), facial canal (2 cases) and automastoidectomy noted in 4 cases. Ossicular chain was disturbed in 13 (65%) patients. The ossicle most commonly destroyed was the incus (80%), followed by malleus (75%) and the stapes (30%). These findings were correlated with the study done by Tos.[23]

CONCLUSION

Eradication of cholesteatoma and restoration of hearing function are more difficult to obtain in children than in adults. The balance between these two goals is related to the incidence of recidivism, the degree of ossicular damage and the experimental evidence that this disease exhibits a more aggressive behaviour than in adults. Fortunately, intratemporal and intracranial complications, such as inner ear fistula, facial nerve paralysis and epidural or intracerebral abscess, are rare in children. Early diagnosis, allowing early treatment, is a key element of the prognosis. In this small series of children mostly operated by CWD technique, results obtained were in line with those reported in the literature i.e., prevention of recurrences are considered to be better with CWD mastoidectomy. These children need to be followed closely by the otolaryngologist. In light of the above mentioned data, an individualised approach is required for the treatment of paediatric cholesteatoma, and the choice of surgical technique should depend on the anatomical, biological, radiological and social factors.

Limitations

As this was a retrospective study, patients could not be followed up post operatively and hence difficult to comment upon the incidence of recurrence and residual cholesteatoma in children.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

REFERENCES

1. Drahy A, De Barros A, Lerosey Y, Choussy O, Dehesdin D, Marie JP. Acquired cholesteatoma in children: strategies and medium-term results. Eur Ann Otorhinolaryngol Head Neck Dis. 2012;129:225-9.
2. Morita Y, Yamamoto Y, Oshima S, Takahashi K, Takahashi S. Acquired cholesteatoma in children: clinical features and surgical outcome. Auris Nasus Larynx. 2014;41:417-21.
3. Schraff S, Strasnick B. Pediatric cholesteatoma: a retrospective review. Int J Pediatr Otorhinolaryngol. 2006;70:385-93.
4. Gocmen H, Kilic R, Ozdek A, Kizilkaya Z, Safak M, Samim E. Surgical treatment of cholesteatoma in children. Int J Paediatr Otorhinolaryngol. 2003;67:867-72.
5. De Corso E, Marchese MR, Scarano E, Paludetti G. Aural acquired cholesteatoma in children: surgical findings, recurrence and functional results. Int J Paediatr Otorhinolaryngol. 2006;7:1269-73.
6. James Adrian L. Endoscopic middle ear surgery in children. Otolaryngol Clin North Am. 2013;46:233-44.
7. Brown JS. A ten year statistical follow-up of 1142 consecutive cases of cholesteatoma: the closed vs. the open technique. Laryngoscope. 1982;92:390-6.
8. Bujia J, Holly A, Antoli-Candela F, Tapia MG, Kastenbauer E. Immunobiological peculiarities of cholesteatoma in children: quantification of epithelial proliferation by MIBI. Laryngoscope. 1996;106:865-8.
9. Shirazi Mobeen A, Muzaffar Kamil, Leonetti John P, Marzo Sam. Surgical treatment of pediatric cholesteatomas. Laryngoscope. 2006;116:1603-07.
10. Dornelles Cde C, da Costa SS, Meurer L, Rosito LP, da Silva AR, Alves SL. Comparison of acquired cholesteatoma between pediatric and adult patients. Eur Arch Otorhinolaryngol. 2009;266:1553-61.
11. Wetmore RF, Konkle DF, Potsic WP, Handler SD. Cholesteatoma in the pediatric patient. Int J Pediatr Otorhinolaryngol. 1987;14:101-12.
12. Kathleen CY. Pediatric clinics of North America. 1996;43:1245-52.
13. Leighton SE, Robson AK, Anslow P, Milford CA. The role of CT imaging in the management of chronic suppurative otitis media. Clin Otolaryngol. 1993;18:23-9.
14. Nevoux J, Lenoir M, Roger G, Denoyelle F, DucouLePointe H, Garabedian N. Childhood cholesteatoma. Eur Ann Otorhinolaryngol Head Neck Dis. 2010;127:143-50.
15. Rosenfeld RM, Moura RL, Bluestone CD: Predictors of residual-recurrent cholesteatoma in children. Arch Otolaryngol Head Neck Surg. 1992;118:384-91.
16. Stern SJ, Fazekas-May M. Cholesteatoma in the pediatric population: Prognostic indicators for surgical decision making. Laryngoscope. 1992;102:1349-52.
17. Vartiainen E. Factors associated with recurrence of cholesteatoma. J Laryngol Otol. 1995;109:590-2.
18. Mills RP, Padgham ND. Management of childhood cholesteatoma. J Laryngol Otol. 1991;105:343-5.

19. Park KT, Song JJ, Moon SJ, Lee JH, Chang SO, Oh SH. Choice of approach for revision surgery in cases with recurring chronic otitis media with cholesteatoma after the canal wall up procedure. Auris Nasus Larynx. 2011;38:190-5.
20. Osborn AJ, Papsin BC, James AL. Clinical indication for canal-wall down mastoidectomy in a pediatric population. Otolaryngol Head Neck Surg. 2012;147:316-22.
21. Sun J, Sun J, Hu Y, Lv Q, Wang Y, Li X, et al. Canal-wall down mastoidectomy with mastoid obliteration for pediatric cholesteatoma. Acta Oto Laryngol. 2010;130:259-62.
22. Hajri H, Mannoubi S, Ben Mansour Mh, Kaffel N, Marrakchi M, Kooli H, et al. Le cholesteatome de I'enfant. J Tun ORL. 2005;15:5-8.
23. Tos M. Pathology of the ossicular chain in various chronic middle ear diseases. J Laryngol Otol. 1979;93:769-80.

Cite this article as: Khavasi P, Bhargavi K, Malashetti SP, Yasha C. Acquired cholesteatoma in children: presentation, complications and management. Int J Otorhinolaryngol Head Neck Surg 2018;4:1017-22.