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Original Research Article

Clinical aspects and surgical outcomes of pediatric cholesteatoma in a tertiary care teaching hospital

Vijay Gupta[1], Navid Mirza[2]*, Arindam Gupta[3]

Department of ENT, [1]Government District Hospital, Rajouri, [2]Care-N-Cure Hospital, Jammu, Jammu and Kashmir, India
[3]SGT Medical College Hospital & Research Institute, Gurugram, Haryana, India

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***Correspondence:**
Dr. Navid Mirza,
E-mail: drnavidmirza@gmail.com

ABSTRACT

Background: Cholesteatoma in children is customarily viewed as a separate and special entity as it presents the otolaryngologist with a verity of diagnostic and therapeutic challenges. The study was planned with objective of studying the clinical aspects and surgical outcomes of pediatric cholesteatoma.
Methods: The patients younger than 15 years and diagnosed as congenital cholesteatoma were included in the present study. All the patients underwent thorough clinical examination, laboratory and radiological investigations. Depending on the stage of disease, surgical procedure was decided. All patients were followed-up for at least 12 months after surgery.
Results: Total 46 children congenital cholesteatoma (CC) was included in the study. The mean age of diagnosis was 7.08±5.64 years. All the cases have unilateral ear involvement. 14 patients were asymptomatic and CC were found incidental. Patients with CC represented with more commonly with hearing impairment. The CC was most commonly found in the anterosuperior quadrant. All 46 patients with CC underwent operative procedure with transcanal myringotomy was commonest surgical approach. During the 12 month follow-up, 28.86% of patients presented with recurrence of disease.
Conclusions: Despite of improved understanding of this disorder with better surgical techniques and skill, high recurrence rate is still major challenge for successful management of CC.

Keywords: Cholesteatoma, Congenital cholesteatoma, Clinical profile, Surgical interventions

INTRODUCTION

In 1838, Johannes Müller was the first to coin the unhappy term of 'cholesteatoma', which is so much deeply rooted in otological nomenclature that in spite of its being misleading is still used to overcome the conflicting views of various successive authors.[1] Although, cholesteatoma is not a neoplastic lesion, yet it is nonetheless insidious and potentially dangerous disease.[2] Histologically, it is an epidermoid cyst characterised by a benign a mass of keratinised squamous epithelium, peri-matrix and keratin, usually found in the tympanic cavity, mastoid or subepithelial connective tissue, which demonstrates destructive and inflammatory properties.[3]

The etiopathogenesis of cholesteatoma is still controversial.[4] On histopathological examination, cholesteatomas characterized by accumulations of desquamating keratinized epithelium within the middle ear or other pneumatized portion of the temporal bone.[5] Based on the origin, cholesteatoma can be classified into two types: (i) congenital cholesteatoma; and (ii) acquired cholesteatoma.[6] Congenital cholesteatoma is mainly seen in pediatric population and usually presents with an intact tympanic membrane, while the acquired form is usually

seen in adults and is typically associated with a defect in the tympanic membrane.[6] Congenital cholesteatoma characterized by white mass that forms prior to birth behind an intact tympanic membrane and has no history of otitis media or previous otologic procedures.[7]

As such, cholesteatoma is uncommon with incidence of acquired cholesteatoma ranges from 9-12.6 cases per year per 100,000 adults and in children 3-15 cases per year per 100,000 children.[8-10] According to geographical studies regarding cholesteatoma, there are higher rates of cholesteatoma in less developed countries.[11] In pediatric cholesteatoma, the symptoms may vary depending on the extent and the location of lesions from asymptomatic to the conductive hearing loss, labyrinthitis, facial palsy, sensorineural hearing loss, intracranial complications, etc.[12] The goals of treatment of pediatric cholesteatoma include complete removal of the lesion; improvement and preservation of hearing; and prevention of recurrence. Surgical intervention is mainstay for management of pediatric cholesteatoma to achieve these goals. The surgical interventions include tympanoplasty, canal wall up mastoidectomy, and canal wall down mastoidectomy. Again, the type and approach of surgical intervention is determined by the location, type, and stage of the lesion as well as by the surgeon's preference and experience.[13]

Cholesteatoma in children is customarily viewed as a separate and special entity as it presents the otolaryngologist with a verity of diagnostic and therapeutic challenges. So, this was planned with objective of studying the clinical aspects and surgical outcomes of pediatric cholesteatoma.

METHODS

This was a prospective study of was conducted in the department of otorhinolaryngology of Govt. medical college, Jammu. The study was approved from Institutional Ethics Committee. The study was carried out between August 2004 to December 2005. The patients younger than 15 years and diagnosed as congenital cholesteatoma in the department of otorhinolaryngology were included in the present study.

For the diagnosis of congenital cholesteatoma, criteria suggested by Levenson et al were applied in the present study.[14] The criteria include, (i) a white-coloured mass present in the middle ear cavity with intact tympanic membrane; (ii) normal pars flaccida and pars tensa; (iii) no past history of otorrhea and perforation; (iv) no past history of otological surgery; (v) the occulusion of the external canal, intramembranous cholesteatoma and giant cholesteatoma were excluded; and (vi) no preclusion of the past history of otitis media.

For clinical staging of the congenital cholesteatoma, the recommendations of Potsic et al was applied.[15] The different staging include, (i) stage I: single quadrant involvement: no ossicular involvement or mastoid extension; (ii) stage II: multiple quadrant involvement: no ossicular involvement or mastoid extension; (iii) stage III: ossicular involvement: includes erosion of ossicles and surgical removal for eradication of disease; no mastoid involvement; and (iv) stage IV: mastoid extension (regardless of findings elsewhere).

For classification of the congenital cholesteatoma for type, classification of McGill et al was used, which include, (i) closed cyst (encapsulated cholesteatoma); (ii) open infiltrative cyst (cholesteatoma matrix in direct contact with the middle ear mucosa).[16] For classification of the congenital cholesteatoma for location, position of the handle of malleus was taken into consideration, it was divided into the lesion in the anterior and posterior area.

All the patients underwent thorough clinical examination, laboratory and radiological investigations. Depending on the stage of disease, surgical procedure was decided which included, tympanoplasty, epitympanoplasty, canal wall up mastoidectomy, canal wall down mastoidectomy, and ossiculoplasty (if needed). All patients were followed-up for at least 12 months after surgery.

All data obtained from the patients were recorded in case record form. The results were presented as frequency, proportions, mean and standard deviations. P value of less than 0.05 was regarded as statistically significant difference.

RESULTS

Total 46 children under the age of 15 years with paediatric/congenital cholesteatoma (CC) were included in the study. The common age group represented and diagnosed having CC was 6–10 years with mean age of 7.08±5.64 years. The males (26, 56.52%) were more affected as compared females (20, 43.48%). All the cases have unilateral ear involvement with right ear (28, 60.87%) more involved. Out of 46 patients, 14 (30.43%) patients were asymptomatic and CC were found incidental. With symptoms, patients with CC represented with hearing impairment (16, 34.78%); otalgia (5, 10.87%); facial palsy (1, 2.17%); tinnitus (6, 13.04%); and ear fullness (4, 8.70%) (Table 1).

According to Table 2, closed type of CC was found in 25 (54.35%) patients while open type in 21 (45.65%) patients. The CC was most commonly found in the anterosuperior quadrant (17, 36.96%). Multiple quadrants were involved in 12 (26.09%) patients. On classifying the CC based on the disease stage, 17 (36.96%) patients had stage I CC followed by stage III (12, 26.09%), stage IV (9, 19.57%) and stage II (8, 17.39%). As shown in Table 3, disease stage I and II more diagnosed in age group of 0–5 years, while disease stage III and IV in age groups of 6–10 years and 11–15 years, respectively.

All 46 patients with CC underwent operative procedure with transcanal myringotomy was performed in 17 (36.96%) patients followed by canal wall up mastoidectomy (14, 30.43%); canal wall down mastoidectomy (8, 17.39%); and tympanoplasty (7, 15.22%) (Table 4). During the 12-month follow-up, 13 (28.86%) patients presented with recurrence of disease with all disease stages had similar rate of recurrence (Table 5).

Table 1: Characteristics of patients with congenital cholesteatoma (n=46).

Variables	N	%
Age (years)		
0–5	15	32.61
6–10	22	47.83
11–15	9	19.57
Gender		
Male	26	56.52
Female	20	43.48
Involved Ear		
Right	28	60.87
Left	18	39.13
Presence of symptoms		
Asymptomatic	14	30.43
Symptomatic	32	69.57
Type of symptoms		
Hearing impairment	16	34.78
Otalgia	5	10.87
Facial palsy	1	2.17
Tinnitus	6	13.04
Ear fullness	4	8.70

Table 2: Type, stage and location of congenital cholesteatoma (n=46).

Variables	N	%
Type of mass		
Closed	25	54.35
Open	21	45.65
Disease stage		
I	17	36.96
II	8	17.39
III	12	26.09
IV	9	19.57
Location		
AS quadrant	17	36.96
PS quadrant	4	8.70
AI quadrant	1	2.17
PI quadrant	1	2.17
AS+PS quadrant	6	13.04
AS+PI quadrant	3	6.52
AS+AI quadrant	2	4.35
Multiple quadrants	12	26.09

Table 3: Distribution of the patients according to disease stage and age.

Disease stage	0-5 years		6-10 years		11-15 years		Total	
	N	%	N	%	N	%	N	%
I	10	55.56	5	31.25	2	16.67	17	36.96
II	4	22.22	2	12.50	2	16.67	8	17.39
III	3	16.67	7	43.75	2	16.67	12	26.09
IV	1	5.56	2	12.50	6	50	9	19.57
Total	18	100	16	100	12	100	46	100

Table 4: Distribution of the patients according to type of surgery (n=46).

Type of surgery	N	%
Transcanal myringotomy	17	36.96
Tympanoplasty	7	15.22
Canal wall up mastoidectomy	14	30.43
Canal wall down mastoidectomy	8	17.39

Table 5: Distribution of the patients according to disease stage and recurrence after surgery.

Disease stage	Total patients	Recurrence	
		N	%
I	17	4	23.53
II	8	2	25.00
III	12	4	33.33
IV	9	3	33.33
Total	46	13	28.26

DISCUSSION

In the present study, total 46 children under the age of 15 years with paediatric/congenital cholesteatoma (CC) were included in the study. The mean age of diagnosis of CC was 7.08±5.64 years. The mean±SD age of children with CC was 5.6±2.8 years in the study done by Nelson et al.[17] The study done by Cho et al also shown similar mean age of 6.1 years.[13] Though this condition is congenital, often the diagnosis is delayed because CC may grow for years without causing signs or symptoms and, having grown without early detection.[18]

In the present study, all the cases have unilateral ear involvement with right ear more involved. In a study done by Cho et al, all 93 patients had unilateral ear involvement.[13] Particular attention must be paid to the status of the ears of children to complain about symptoms associated with cholesteatoma, especially when presentations are subtle or unilateral.[19]

Out of 46 patients, 14 (30.43%) patients were asymptomatic and CC were found incidental in the

present study. 16 (45.00%) cases were discovered asymptomatic in a study done by Park, et al.[12] CC is usually asymptomatic in the initial stages of growth.[20] Cholesteatomas often exist in a nonaggressive state and asymptomatic, remaining undetected for years before potentially dangerous presentations manifest.[21]

According to symptomatology, patients with CC represented with more commonly with hearing impairment followed by tinnitus, otalgia, ear fullness, and facial palsy in this study. In a study done by Park et al, hearing impairment (29.0%) was most prevalent, followed by otalgia (20.0%), tinnitus (11.4%), and ear fullness (8.5%).[12] The literatures also suggest that patients with CC may be asymptomatic or present as a conductive type of hearing loss. Other presentations may include otalgia, vertigo and facial palsy. The later symptoms indicate the progression of the disease in terms of erosion into the semi-circular canals or the Fallopian canal, respectively.[20]

Regarding the location of CC, in a study by Park et al, out of total 35 cases, CC were limited to the anterior area in 17.1% of cases, posterior area in 14.2% of cases, and in 68.5% of cases, the anterior and posterior area were involved simultaneously with CC.[12] Levenson et al. have reported that it is located primarily in the anterior area of the tympanic cavity.[14] The anterior and posterior area were involved simultaneously with CC in 45.65% of cases while CC were limited to the anterior area in 43.48% of cases in the present study.

On classifying the CC based on the disease stage, 17 (36.96%) patients had stage I CC followed by stage III (12, 26.09%), stage IV (9, 19.57%) and stage II (8, 17.39%). As shown in table 3, disease stage I and II more diagnosed in age group of 0–5 years, while disease stage III and IV in age groups of 6–10 years and 11–15 years, respectively. Cho et al reported patients with stage I CC accounted for the highest proportion of the cases until the age of 5, while all of the patients with CC discovered after the age of 7 years had stage III or IV disease.[13]

The surgical method for CC is determined by the location, type, and stage of the lesion as well as by the surgeon's preference and experience.[13] All 46 patients with CC underwent operative procedure with transcanal myringotomy was performed in 17 (36.96%) patients followed by canal wall up mastoidectomy (30.43%); canal wall down mastoidectomy (17.39%); and tympanoplasty (5.22%). According to the study report of Nelson et al, the stage 1 can be managed satisfactorily by tympanoplasty, the stage 2 may require extended tympanoplasty with canal wall up mastoidectomy and ossiculoplasty. Stage 3 can be managed similar to type 2 cases, with occasional requirement of canal wall down mastoidectomy.[17]

During the 12-month follow-up, 28.86% patients presented with recurrence of disease with all disease stages had similar rate of recurrence in the present study. Cho et al reported somewhat similar rate of recurrence of CC.[13] As our patients were fallowed-up for very short-duration–12 months only, we cannot really comment upon the recurrent or residual of cholesteatoma since it is too short a period for any conclusion. Deciding an appropriate duration for follow-up following cholesteatoma surgery is a challenging issue and it is difficult to resolve in the near future.[21]

Paediatric cholesteatoma poses many challenges to the otolaryngologists. Firstly, the otologist must make a correct and early diagnosis. Secondly, the otologist must provide a disease-free ear that will remain stable throughout adulthood and can be easily followed, up. Thirdly, a serviceable hearing level, which for the development of normal language and communication skills should be achieved. Fourthly, the otologist should educate the patient and the family regarding the nature of the disease; the need for long-term follow-up; and the possibility of recurrence of the disease, further radiographic studies, reconstructive surgery, and aural rehabilitation. The diagnosis and management of cholesteatoma in children is often difficult. In general, many children are diagnosed incorrectly because of a paucity of symptoms, their uncooperativeness, and their small tortuous ear canals which makes examination difficult. Despite the correct diagnosis and proper operative procedure, there are higher rates of recurrence.

CONCLUSION

In conclusion, this study provides a comprehensive summary of clinical representation and management of CC. Despite of improved understanding of this disorder with better surgical techniques and skill, high recurrence rate is still major challenge for successful management of CC. Further research should be directed toward early diagnosis and surgical intervention.

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