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Pediatric Cholesteatoma Aggressiveness; A Comparative Study of Predictive Factors and Recurrence Rate

Zeinab AlQudehy[1*], Abdulrahman Al-Abdulqader[1] and Hamzah Alshaikh[2]

[1]Department of Otorhinolaryngology, Dammam Medical Complex, Saudi Arabia

[2]Department of Otorhinolaryngology, King Fahd Military Medical Complex, Saudi Arabia

Research Article

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***Corresponding author:** Zeinab AlQudehy, Department of Otorhinolaryngology, Dammam Medical Complex, P.O. Box 508, Dhahran 31311, Saudi Aramco, Saudi Arabia, Tel: 00966504804838; Email: drzeinabent@gmail.com

Abstract

Objective: The optimal surgical treatment for pediatric cholesteatoma is controversial. Management decisions including intact canal wall versus open cavity techniques continue to be debated. In an attempt to clarify this issue, we conducted a retrospective analysis of our experience with cholesteatoma cases presenting in pediatric population.

Material and Method: Retrospective review was conducted on all children younger than 18 years of age and had cholesteatoma, between 2010 and 2013. Inclusion and exclusion criteria were specified. The children were divided into canal wall down (CWD) and canal wall up (CWU) mastoidectomies. The two groups were then compared regarding their age at presentation, clinical presentation, microscopic examination, radiological grading, intra operative findings, and postoperative outcomes, and compared with international recurrence rate figures.

Results: We had total of four CWD & seven CWU mastoidectomies. CWD children were an average of 6 years of age, and had more aggressive disease at presentation with attic erosion in 75% of the cases. Granulation tissues with eroded ossicles were present in almost all the cases. While, CWU children had a longer history of offensive ear discharge, and presented with retraction pocket in 43% of the cases. There was no significant difference in the recurrence rate in CWD and CWU groups (25% & 28.6% respectively).

Conclusion: Treatment of pediatric cholesteatoma should be individualized. The choice between CWU & CWD can be judged by several factors in patient history and through a thorough clinical ear examination. CWU procedure is an adequate surgical option for treating most acquired and congenital cholesteatomas, preventing disease recurrence, and maintaining good hearing outcomes, and CWD mastoidectomy chosen for patients with recurrent or more extensive disease.

Keywords: Pediatric Cholesteatoma; Aggressiveness; Canal Wall Up (CWU); Canal Wall Down (CWD)

Introduction

Pediatric cholesteatoma is a potentially dangerous disease affecting the quality of life of affected children. There are many challenges in diagnosing and treating cholesteatoma in pediatric age group with no universally accepted opinions about the best surgical options and its outcome. The primary goal of cholesteatoma surgery is complete eradication of disease with the secondary goal of hearing improvement post-operatively. Children with cholesteatoma have been shown to demonstrate more aggressive disease than adults with higher recurrence rates [1-7]. The two main surgical procedures used to deal with cholesteatoma are canal wall up (CWU) and canal wall down (CWD). CWU procedure is distinguished from CWD technique by the preservation of the posterior wall of the external auditory canal. While a number of surgeons prefer the CWD technique, others opt for CWU. Each of the two procedures had its own advantages and disadvantages. CWU in comparison to CWD has more rapid healing, allowance of water exposure during swimming, decreased postoperative aural care, and allow patients to use hearing aid in the postoperative period [8]. The main disadvantage of CWU is the technical difficulties with limited exposure of the epitympanum and sinus tympani typically leading to a higher residual and recurrence rate compared with CWD [1,9].

Predicting factors for aggressiveness of pediatric cholesteatoma are not yet well established, therefore, the best treating surgical options is not yet standardized. Regardless of techniques, recurrent cholesteatoma developing from postoperative tympanic membrane retraction and adhesions is still frequent problems encountered in 7-57% of the case [1,7,10-14]. Management of pediatric cholesteatoma remains to be debated. To our best knowledge, none of the previous studies addressed aggressive behavior of cholesteatoma in Middle East children. We performed a 3-year retrospective review of our experience. In an attempt to find predictive factors for aggressiveness of pediatric cholesteatoma. This will help treating surgeon to clarify the issue of the most appropriate management of pediatric cholesteatoma, therefore reduce the recurrence of the disease and eliminate the need for surgeon ear revisit.

Methods

This study was designed as a retrospective cohort study based on two surgeon's clinical practice at Dammam Medical Complex, Ministry of Health, Dammam, Eastern Province, Saudi Arabia. Eligible patients were identified from a prospectively kept surgical database, which include demographics, symptoms at presentation, otological examination, audiological findings, and performed surgical procedures along with intra operative detailed reports. Complications, and follow up information were extracted from individual patient's hospital medical records. Research ethics approval was granted by Dammam Medical Complex Research Ethical Approval Committee (DMC-RACNo. 0020). Eligible children were those younger than 18 years of age, who had clinical symptoms and signs suggestive of cholesteatoma, and underwent mastoidectomy between August 2010 and July 2013. Children were excluded if the indication for mastoidectomy was not suspicion of acquired cholesteatoma, or presence of congenital cholesteatma as judged by the clinical picture.

Data Collection

Review of the hospital medical records was then completed and crosschecked to ensure data accuracy. The exclusion criteria were then applied to give the final data for analysis.

From prospectively designed database

Variables of interest were collected prospectively by one pediatric otolaryngology surgeon (ZQ) and one otology surgeon (YN) in a special patient information sheet during the clinic visit, and operating room session, and included:

a) Age at time of initial presentation
b) Gender
c) Allergies
d) Ear affected
e) Ear discharge
f) Ear discharge characteristics, such as presence of offensive smell, or blood
g) Ear pain
h) Overall duration of symptoms
i) Previous ear trauma
j) Hearing loss
k) Recurrent upper respiratory tract infections
l) Tinnitus
m) Vertigo
n) Nystagmus
o) Facial Asymmetry
p) Findings at otologic examination, such as attic cholesteatoma, granulation tissue, tympanic membrane perforation, aural polyp, active ear

discharge, keratin flakes, or tympanic membrane retraction pocket.

q) Tuning fork tests
r) Fistula test
s) Facial nerve integrity
t) Basic audiology examination (PTA, and tympanogram)
u) Intraoperative findings including; mastoid pneumatization, presence of granulation tissues or keratin flakes, involvement of the ossicles, facial canal erosion, facial nerve dehiscence, lateral semicircular canal dehiscence, intracranial involvement, and if reconstruction done.

Hospital patient's medical record

Review of the hospital medical records was then completed and crosschecked to ensure data accuracy. The following variables were collected:

a) Complications (intraoperative & postoperative)
b) Evidence of recurrence of patient's initial symptoms/signs on follow up

Follow-up

All patients, included in this study, were followed up by the primary treating surgeon for a minimum of one-year duration after the mastoidectomy.

Statistical analysis

Simple descriptive statistics was used to interpret the results of this study.

Results

A total of 11 patients were included in our study. The average age of patients at initial presentation was11.6 years (range 5-18 years), with almost equal gender distribution. The children included in our study were having positive history of allergy in one fourth of the cases and was mainly allergic conjunctivitis and bronchial asthma. Right and left ears were equally affected. 10 of the included patients, 90% had ear discharge as their chief complaint at presentation, out of whom9 had offensive discharge. One quarter of patients had blood stained discharge and were having painful ears. The duration of patient symptoms were variable ranging from one month to 8 years, with average of 30 months. 90 % of patients had hearing loss at presentation. Two out of the 11 cases had history of bilateral myringotomy and ventilation tubes insertion.

From another view, a retrospective classification of the studied patients according to the surgical procedure was carried out. Children were divided into canal wall down (CWD) and canal wall up (CWU) mastoidectomies. The two groups were then compared according to their age at time of presentation, clinical presentation (Table 1), patient's examination (Table 2), intraoperative findings (Table 3) and performed surgical procedures with postoperative outcomes including recurrence rate (Table 4).

	CWD	CWU	COMMENT
Number of Cases	4	7	
***Average age of patients**	**6 years**	**16.3 years**	**Younger patients had CWD**
M:F Ratio	1 to 3	4 to 3	Apparently no differences
Allergies	1/4 (25%) allergic conjunctivitis	2/7 (28.5%) bronchial asthma	
Affected ears left: right	1 left and 3 right (25 % left & 75% right)	5 left and 2 right (71.4% left & 28.6% right)	
Ear discharge	Yes in 100%	Yes in 85.7%	
Offensive discharge	Positive in 100%	Positive in 83.3%	
Bloody discharge	Positive in 25% (1/4)	Positive in 28.6% (2/7)	
Otalgia	Positive 25% (1/4)	Positive 28.6% (2/7)	
***Average duration of symptoms**	**12.75 months (3-24 months)**	**39.86 months (1 month - 8 years)**	**Patients with longer duration of symptoms had CWD**
Hearing loss	Positive in 100%	Positive 85.7% (6/7)	Apparently no

Recurrent URTI	All negative	All negative	differences
Tinnitus	All negative	All negative	
Vertigo	All negative	All negative	
Nystagmus	All negative	All negative	
Facial Asymmetry	All negative	All negative	
Past Surgical History BMAT	Positive 25% (1/4)	Positive 14.3% (1/7)	

Table 1: Demorgraphic & Symptomatic Characteristics of Both Groups (Cwd & Cwu Groups).
• Significant variable upon observation

	CWD (4)	CWU (7)	COMMENT
Findings at Microscopic Ear Examination			
Attic cholesteatoma	25% (1/4)	14.3% (1/7)	Apparently no differences
Granulation tissue	25% (1/4)	14.3% (1/7)	
TM perforation	25% (1/4)	28.6% (2/7)	
***Aural polyp**	**50% (2/4)**	**0%**	**Patient with one or more of those signs had CWD**
***Ear discharge**	**50% (2/4)**	**0%**	
***Keratin flakes**	**50% (2/4)**	**0%**	
***TM retraction pocket**	**25% (1/4)**	**42.9% (3/7)**	
Un-affected ear TM retraction	25% (1/4)	28.6% (2/7)	Apparently no differences
Nasal Examination			
DNS	25% (1/4)	0%	Apparently no differences
HIT	25% (1/4 HIT)	14.3% (1/7)	
Throat: Tonsil Enlargement	75% (3/4)	0%	
Tuning Fork Test			
Rinnie Test in The Affected	Negative: 100% (4/4)	Negative 71.4% (5/7) Positive 28.6% (2/7)	Apparently no differences
Weber Test	Lateralized to the affected ear: 100% (4/4)	Lateralized to affected ear: 71.4% (5/7) Centralized: 28.6% (2/7)	
Fistula Test Positive	0%	0%	
Facial Nerve Paralysis	0%	0%	
Audiological Examination			
PTA	100% (4/4) moderate CHL	71.4% (5/7) moderate CHL, 28.6% (2/7) mild- moderate CHL	Apparently no differences
Tympanogram	25% (1/4) type B 75% (3/4) type A with absent stabedial reflex	28.6% (2/7) type B	

Table 2: Patient's Clinical Examination Characteristics of Both Groups (Cwd & Cwu Groups).
• Significant variable upon observation

	CWD (4)	CWU (7)	COMMENT
*Pneumatization of the mastoid	100% (4/4) sclerotic	57.1% (4/7) sclerotic	Patients with sclerotic mastoids had CWD
*Presence of granulationtissues	100% (4/4) - 50% (2/4) sinus tympani - 50% (2/4) epitympanum - 25% (1/4) mesotympanum - 50% (2/4) medial to the ossicles	57.1% (4/7) 0% 0% 0% 100% (4/4) lateral to the ossicles	Patients with granulation tissues in the middle ear had CWD
Keratin flakes	100% (4/4)	100% (7/7)	Apparently no differences
*Ossicules engulfed by the granulation tissue	100% (4/4)	57.1% (4/7)	Patients with ossicular involvement had CWD
*Incus erosion + IS joint affection	75% (3/4)	57.1% (4/7)	
*Stapes suprastructure erosion	75% (3/4)	28.6% (2/7)	
Facial canal erosion	0%	0%	Apparently no differences
Facial nerve dehiscence	0%	0%	
LSCC dehiscence	0%	0%	
Intracranial involvement	0%	0%	

Table 3: Intraoperative Ear Findings Characteristics of Both Groups (Cwd & Cwu Groups).
• Significant variable upon observation

	CWD (4)	CWU (7)	COMMENT
Performed surgical procedures	- 100% (4/4) CWD + Conchomeatoplasty	- 71.4% (5/7)atticoantrostomy - 28.6% (2/5) retrograde atticotomy - 42.6% (3/5) combined approach	Variable types of CWU procedures were performed
Reconstruction done	- 25% (1/4)EAC + attic cartilage reconstruction -50% (2/4)tympanoplasty	-28.6% (2/7) tympanoplasty	Apparently no differences
Intraoperative & postoperative complications	0%	0%	
Follow up average & range	20.5 months (12-30 months)	26.3 months (12-35 months)	
*Recurrence Rate	25% (1/4)	28.6% (2/7)	Almost equal recurrence rate in bot CWD & CWU

Table 4: Performed Surgical Procedures and Outcome Measures Characteristics of Both Groups (Cwd & Cwu Groups).
• Significant variable upon observation

Discussion

Pediatric cholesteatoma is considered to be more aggressive in clinical behavior than the adult ones by many authors [2,4,5,15-17], but there is still a lot of debate on this topic. The main goals of cholesteatoma surgery are to; eradicate the disease, preserve and/or improve patient's hearing, and prevent of residual and/or

recurrent disease. A review of the literatures addressing the follow up period, after acquired cholesteatoma surgery, identified various predictors of disease recurrence. Most of recurrences were observed in younger patients aged less than 8 years [18-20] with strong relationship between the young age of patients at presentation and the disease aggressiveness. In our study, patients with more aggressive cholesteatoma, who required CWD procedures, were those who were younger at time of start of their symptoms while the patients with less aggressive disease and requiring CWU were older by almost ten years.

In regards to the average duration of patients symptoms, Palva et al. [16] reported that average duration of symptoms in children with cholesteatoma was 5.8 years, our participants had their symptoms lasted for more than three years in patients required CWU techniques, and about one year in patients required more aggressive operation, i.e. CWD. Opposite to our study, Belachdi et al. had linked symptoms longer than two years with higher risk of recurrence of pediatric cholesteatoma [18]. Ossicular chain involvement at the time of presentation was associated with more extensive disease as seen by Shirazi and colleagues [21], and found that ossicular chain involvement was a significant predictor of patients with a high risk for recurrent disease and therefore more aggressive surgical disease eradication procedures became a necessity as shown in our study. Stanger up and colleagues had found that the age of the patient, presence of ear discharge, ossicular chain resorption, and preoperative Eustachian tube dysfunction were the main predictors for pediatric cholesteatoma [22]. Aggressive cholesteatoma was associated with presence of posterior middle ear invasion, ossicular erosion, and discharging ears preoperatively as found by Vartiainen [23].

In our current study, the findings of more aggressive disease was found in younger children, who had shorter duration of the disease, and found in examination to have active persistent ear discharge, with keratin flakes and retraction pockets. Intraoperative findings of granulation tissues in sinus tympani, epitympanum, and medial to the ossicles, along with ossicular erosion were predictors of aggressive disease and in favor of CWD procedures to eradicate the disease adequately. Using these predictors on regular bases can lead to reduce cholesteatoma recurrence rate and reduce the incidence of ear revisit. In our study, recidivism occurred with comparable rates in CWD and CWU groups 25% and 28.6% respectively. These results are compared with the previously published studies (Table 5).

The main limitations of our study are that being retrospective in nature and did not address the radiological feature as predictors for cholesteatoma aggressiveness. In addition to this, the number of the patient included in the study owing to the fact that it was carried out by data collected from two surgeons only over a relatively short study period in comparison to the literatures (Table 5).

Author	Study Duration/Study Population	N. of Patients	N. of CWU	N. of CWD	Overall RR (%)	CWU RR (%)	CWD RR (%)
Glasscock et al. [2]	6 years/ adult + children	144	142	2	46	-	-
Brown [10]	10 years/ adult + children	98	62	36	34	35	30
Charachon & Gratacap [11]	15 years/ children (3-15 yrs.)	136	99	37	42	45	38
Sanna et al. [12]	Children	148	144	4	40	-	-
Parisier et al. [13]	Children	165	62	103	10	15	14
Dodson et al. [1]	11 years/ children (10 m – 18 yrs.)	58	41	17	36	41	12
Darrouzet et al. [14]	10 years/ children	210	189	21	31	29	43
Scott et al. [24]	11 years/ children	278	221	57	16	17	12

Shirazi et al. [21]	16 years/ children	106	-	-	7	8	6
CURRENT STUDY	3 years/ children	11	7	4	27.3	28.6	25

Table 5: A comparison between the previously reported studies and our current study.
N: Number; RR: Recurrence Rate

Conclusion

Cholesteatoma is more common to be an extensive disease in pediatric population in comparison to that in adults, with higher incidence of residual and recurrent disease. Although it is controversial, it is largely accepted that pediatric cholesteatoma necessitate a more advanced form of treatment. There are many available surgical options to treat cholesteatoma, and the outcome of the surgical management can be determined mainly by the rate of recidivism. Some of the several factors contributing to the final choice of technique are age of patients, duration of symptoms, extent of the disease, anatomical variation, structures affected, and surgeon's choice were highlighted in this study and should be taken into consideration preoperatively and intra operatively to reduce the risk of the disease recurrence.

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