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Research Article

Surgical Treatment Of Congenital Cholesteatoma of Middle Ear at Children

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Abstract

Objectives: Congenital cholesteatoma presents pathological lesion that develops behind intact membrane tympani. Objective of this paper is to present our experience in diagnostics and treatment of congenital cholesteatoma at children.

Material and Methods: Our study represents a retrospective review of 17 children who underwent surgery for congenital cholesteatoma from 2012. to 2019. at the ORL Department, Mother and Child Health Care Institute, Belgrade, Serbia. All patients had congenital cholesteatoma present on one ear. Diagnoses were made from between 3 months to one year after presentation (an average of 6.3 months).

Basic criteria in diagnosis of congenital cholesteatoma were presence of white mass behind the intact timpani membrane with no data on otorrhea, perforation of membrane timpani or surgical interventions. Staging of the cholesteatomas was based on the classification suggested by Potsic.

Results: Congenital cholesteatoma confined to the middle ear cavity was present in 16 (94%) patients, the majority of whom had involvement of multiple quadrants (9 patients or 57%), in 4 (25%) cases CC was localized to the posterior quadrants, in 2 (12%) to the anterior quadrants and in one (6%) case to the anterosuperior and posterosuperior quadrants. One case involved congenital cholesteatoma confined to the mastoid cavity. Preoperative contralateral middle ear disease was absent in 12 (70%) cases, whereas secretory otitis media (SOM) was identified in the contralateral ear in 3 (18%) cases and adhesive otitis in 2 (12%) cases. The following surgical interventions were performed: tympanoplasty type I in 3 (17%) patients with congenital cholesteatoma localized to the anterior quadrants, radical modified tympanomastoidectomy (canal wall up) and tympanoplasty type II in 5 (30%) patients, canal wall down mastoidectomy and tympanoplasty type III in 8 (47%) and simple (cortical) mastoidectomy in 1 (6%) patient (where the cholesteatoma was confined to the mastoid cavity). All patients were followed up 24 ± 6,4 months postoperatively. Recurrent cholesteatoma was diagnosed in 2 (12%) patients, one in a patient who had CWU and one in a patient who had CWD mastoidectomy performed. In both cases a small aplastic poorly pneumatized mastoid was encountered during surgery and both patients had concomitant contralateral middle ear disease (SOM and adhesive otitis media respectively).

Conclusions: Congenital cholesteatoma is characterized by insidious onset with predominantly ipsilateral CHL, which is why in many cases patients are erroneously treated for otitis media with effusion prior to establishment of correct diagnosis. Furthermore, the insidious nature of disease calls for a high index of suspicion in ORL specialists and indeed pediatricians performing newborn hearing screening and hearing tests in preschool children as CCs are most commonly diagnosed by that age. Disease stage is a significant risk factor for recurrence.

Keywords: Congenital Cholesteatoma; Stage; Surgical Procedures

Introduction

Congenital cholesteatoma represents a pathological lesion that develops behind an intact tympanic membrane. That is, a white granulated mass that develops behind an intact tympanic membrane without prior otorrhea, perforation of the tympanic membrane and/or surgery performed on the ipsilateral ear [1]. However, one cannot exclude the existence of otitis media with effusion in the diseased ear [2].

Congenital cholesteatoma represents 1 to 5% of cholesteatomas as reported by mainstream literature [2,3]. Early diagnosis is associated with better pediatrician and otorhinolaryngologist training and better equipment. Pediatric screening protocols are being applied in many countries in Europe consisting of audiometry, immittance testing and otoscopy [4].

The objective of this paper is to present our experience in the diagnosis and treatment of congenital cholesteatoma in children.

Materials And Methods

Our study represents a retrospective review of 17 children who underwent surgery for congenital cholesteatoma from 2012. to 2019. at the ORL Department, Mother and Child Health Care Institute, Belgrade, Serbia. Out of the 17 children, 10 (58%) were male and 7 (42%) females. All patients had congenital cholesteatoma present in one ear. Diagnoses were made from between 3 months to one year after presentation (an average of 6.3 months).

Basic criteria used for the diagnosis of congenital cholesteatoma were the presence of a white mass behind an intact tympanic membrane with no data on prior otorrhea, perforation of the tympanic membrane or surgical intervention.

Staging of the cholesteatoma was based on the classification system suggested by Potsic., *et al*. [3] which is as follows: stage I, disease confined to a single quadrant; stage II, cholesteatoma in multiple quadrants without ossicular involvement or mastoid extension; stage III, ossicular involvement without mastoid extension; and stage IV, mastoid involvement. The type of CC was determined according to the morphological classification by McGill., *et al*. [5] that is, closed cyst (encapsulated cholesteatoma) or open infiltrative cyst (cholesteatoma matrix in direct contact with the middle ear mucosa).

Children were aged 3 to 14 years old, an average age of 6.6 years. Prior to surgery acoustic immittance testing was performed as well as pure-tone audiometry in children 5 years or older. CT of the temporal bone and in one case MRI was performed to confirm diagnosis preoperatively. All surgeries were performed by one surgeon. Surgical approach and technique were determined by the degree of cholesteatoma extension and ossicular chain destruction.

Patients were monitored at 3, 6-, 12-, 18- and 24-month intervals postoperatively at our center.

Descriptive statistics were used to describe the characteristics of patients - frequencies for categorical data, and the mean, standard deviation and quartiles for numerical variables. Numerical variables and their distribution among groups were visually assessed using boxplots and histograms. Due to the small sample size, we attempted correlating the ordinal variables with numerical ones using Spearman's nonparametric correlation, and after collapsing the levels into two groups we performed the t-test for comparison. All tests were two-tailed. $p<0.05$ was considered statistically significant. All analyses were conducted using the Statistical Package for the Social Sciences (IBM SPSS, version 21).

Results

Clinical symptoms of disease were unspecific. In fact, the only symptom reported was unilateral hearing loss in 14 children; six of these patients were erroneously treated for otitis media with effusion (OME) in other hospitals (primary and secondary health care centers). In 2 children incidental CT findings performed as part of headache workup led to diagnosis of congenital cholesteatoma, whereas in one child MRI performed as part of sensorineural hearing loss workup identified a congenital cholesteatoma.

Otomicroscopic findings showed that in one case the tympanic membrane appeared normal. In all other cases it was blurred with a visible white mass that was localized in the anterior quadrants in 7 patients, whereas in 9 patients it filled the whole middle ear. Audiology workup included tympanometry which in 16 cases showed a type B tracing in the diseased ear. One patient with isolated CC involvement of the mastoid recorded a type A tracing on tympanometry. Preoperative pure-tone audiometry was performed in 14 patients aged 5 years or older, demonstrating moderate conductive hearing loss with air-bone gaps ranging from 30 to 40 dB (36,7 ± 5,2 dB) with the exception of one patient who presented with bilateral sensorineural hearing loss most likely from synchronous SNHL of viral etiology.

Diagnosis was confirmed by temporal bone CT. One patient underwent MRI as part of sensorineural hearing loss workup; congenital cholesteatoma represented an incidental finding in this patient. The congenital cholesteatoma was not the cause of SNHL but was nevertheless treated operatively.

Staging of the cholesteatomas was based on the classification system suggested by Potsic., *et al*. [3], type II was diagnosed in 6 (35,5%) patients, type III in 6 (35,5%) and type IV in 5 (29%) patients (Table 1).

		Nr.	%
Otomycroscopic findings of tympani membrane	Correct	1	6
	Blurred in front quadrants	7	41
	Completely blurred	9	53
Audiological findings	Conductive partial deafness	16	94
	Senso-neural partial deafness	1	6
Stage of cholesteatoma	I	0	0
	II	6	35,5
	III	6	35,5
	IV	5	29
Location in tympanic cavity	ASQ +PSQ	1	6
	ASQ+AIQ	2	12
	PSQ+PIQ	4	25
	Multiple quadrant	9	57
Cholesteatoma morphology	Open	7	41
	Closed	10	59
Type of surgical intervention	Tympanoplasty tip I	3	17
	Canal wall up mastoidectomy +tympanoplasty	5	30
	Canal wall down mastoidectomy +tympanoplasty	8	47
	Mastoidectomy and posterior aticotomy	1	6

Table 1: Clinical symptoms, diagnostic findings and intraoperational results with children suffering congenital cholesteatoma.

ASQ (Anterior Superior Quadrant), AIQ (Anterior Inferior Qudrant), PSQ (Posterior Superior Quadrant), PIQ (Posterior Inferior Quadrant).

Open type CC was diagnosed in 7 (41%) patients, and closed type CC was diagnosed in 10 (59%) patients.

Congenital cholesteatoma confined to the middle ear cavity was present in 16 (94%) patients, the majority of whom had involvement of multiple quadrants (9 patients or 57%), in 4 (25%) cases CC was localized to the posterior quadrants, in 2 (12%) to the anterior quadrants and in one (6%) case to the anterosuperior and posterosuperior quadrants. One case involved congenital cholesteatoma confined to the mastoid cavity.

Preoperative contralateral middle ear disease was absent in 12 (70%) cases, whereas secretory otitis media (SOM) was identified in the contralateral ear in 3 (18%) cases and adhesive otitis in 2 (12%) cases.

The following surgical interventions were performed: tympanoplasty type I in 3 (17%) patients with congenital cholesteatoma localized to the anterior quadrants, radical modified tympan mastoidectomy (canal wall up) and tympanoplasty type II in 5 (30%) patients, canal wall down mastoidectomy and tympanoplasty type III in 8 (47%) and simple (cortical) mastoidectomy in 1 (6%) patient (where the cholesteatoma was confined to the mastoid cavity).

Ossicular chain involvement was reported in 11 (64%) patients, with predominant involvement of the incus (partial destruction in 7 and total destruction in 4), whereas stapes involvement was recorded in 4 patients.

There was no extracranial or intracranial extension of disease.

Pure-tone audiometry was performed 3 months postoperatively. Improved hearing thresholds were established at 3 months postoperatively in all patients with the exception of one child with preexisting synchronous SNHL. Closure of the air-bone gap was reported in all patients even those who underwent CWD mastoidectomy who had postoperative air-bone gaps of between 22 and 28 dB, whereas patients who underwent CWU mastoidectomy recorded postoperative air-bone gaps of between 15 and 20 dB. The average values of the air-bone gap were 24,4 ± 4,2 dB (Table 2).

Intraoperatively small aplastic poorly pneumatized mastoids were encountered in 5 (30%) patients whereas well pneumatized mastoids were seen in 12 (70%) patients.

		Nr.	%
Air-bone GAP before surgical procedures (total 14 patients)	> 20 dB 20-30 dB >30 dB	1 5 8	6 37 57
Air-bone GAP after surgical procedures (total 14 patients)	>20 dB 20-30 dB	4 10	27 73

Table 2: Air-bone GAP before and after surgical procedures.

All patients were followed up 24 ± 6,4 months postoperatively. Recurrent cholesteatoma was diagnosed in 2 (12%) patients, one in a patient who had CWU and one in a patient who had CWD mastoidectomy performed. In both cases a small aplastic poorly pneumatized mastoid was encountered during surgery and both patients had concomitant contralateral middle ear disease (SOM and adhesive otitis media respectively).

Mean patient age was non-significantly smaller when the cholesteatoma was confined to the middle ear cavity as opposed to extension into the mastoid cavity (4.5 vs 7.3 yrs). Duration of symptoms was significantly shorter in patients without extension (4.25 vs. 7 months). Due to the large number of surgical techniques and small dataset, we could not assess the impact of individual surgical techniques but we found no statistically significant differences in hearing improvement for open vs. closed surgical techniques.

Discussion

The etiology and pathogenesis of congenital cholesteatoma is not well known. The most common theory was presented by Teed in the year 1936 [6]. One proposed theory is that congenital cholesteatoma develops from epidermoid formation within the middle ear cavity up to 33 weeks gestation. Lee., *et al.* [7] performed the resection of 211 temporal fetus bones in 1998, and found the presence of epidermoid formation in 88 (42%), whereas in 18% epidermoid formation was present in both ears.

There are other theories on the origin and development of congenital cholesteatoma. Reudi [8] believes congenital cholesteatoma develops as a consequence of inflammatory damage of the tympanic membrane. This damage leads to micro-perforations in the tympanic membrane through which the basal layer of squamous epithelium enters the cavum tympani forming congenital cholesteatoma.

Sade [9] in their metaplastic theory, stated that inflammatory insult of the epithelial layer of middle ear causes metaplastic transformation into keratin producing squamous epithelium. Therefore, congenital cholesteatoma forms in the absence of tympanic membrane perforations.

Aimi [10] suggests an epithelium migration theory. He believes that congenital cholesteatoma originates from ectodermal cells of the outer ear canal that migrate by accident through the tympanic isthmus into the middle ear.

Northrop [11] believes that squamous epithelial cells are transferred by amniotic fluid in utero into the middle ear and represent a potential source of development of congenital cholesteatoma.

Tos [4] supports an epithelial formation theory. He believes that the tympanic membrane retracts by sticking to the handle of the maleus, neck of the maleus or long process of the incus. His assumptions are supported by frequent retractions of the pars tensa and pars flaccida or Shrapnell's membrane found in childhood. These retractions are presumably caused by Eustachian tube dysfunction and the presence of otitis media with effusion, which has a prevalence of 75% in children during the first year of life. Due to these considerable pressure changes within the cavum tympani certain cells of the tympani membrane are lost, leading to cholesteatoma development.

Insidious onset and unspecific symptoms make diagnosis of congenital cholesteatoma difficult. All our patients presented with unilateral congenital cholesteatoma, though Darouset., *et al*. [12] described cases of bilateral congenital cholesteatoma. The only reported symptom was congenital hearing loss ranging from moderate to severe. Incidental findings on CT or MRI may lead to diagnosis of congenital cholesteatoma as in the case of 3 of our patients who presented with headache (in 2 patients) and sudden sensorineural hearing loss (in 1 patient). Other described symptoms such as elevated body temperature, facial nerve paralysis and vertigo are only signs of cholesteatoma expansion into nearby bony structures [12]. Otogenic complications were not described in our patients.

Otomicroscopy helps to identify signs of congenital cholesteatoma. Our research confirmed that whenever we had cholesteatoma present in the cavum tympani, the tympanic

membrane was blurred, with a noticeable white mass behind it. Nevertheless, some studies claim preoperative otomicroscopy having positive predictive values of between 29 and 45% as established by definitive intraoperative findings.

CT is not wholly sensitive or specific but is a valuable diagnostic aid. Some authors state that CT cannot accurately determine cholesteatoma extension. Congenital cholesteatoma expansion can lead to Eustachian tube dysfunction and subsequent OME which may mask CC identification and impede diagnosis [14]. In these cases, mandatory pressure equalizing tube (PET) insertion may be required to resolve OME prior to reliable imaging. Although diffusion-weighted imaging (DWI) may help with differentiating effusion from cholesteatoma, authors do not recommend the use of general anesthesia in children in order to obtain preoperative MRI unless a concern for petrous apex or intracranial involvement is present [15].

Tympanometry and pure-tone audiometry are useful in congenital cholesteatoma workup. With larger CCs type B tracings are obtained on tympanometry whereas pure-tone audiometry shows variable degree conductive hearing loss (CHL). With smaller CCs confined to only two quadrants of tympanic membrane, type A tracings on tympanometry are possible [16].

Some authors point to an association between CC and other congenital anomalies such as first branchial cleft anomalies, branchiootorenal syndrome (BOR), congenital heart disease and pyloric stenosis [12,17].

Congenital cholesteatoma requires operative management. As with acquired cholesteatoma, surgical goals include first and foremost complete removal of the cholesteatoma (and corollary impact on residual disease), ossicular chain reconstruction with hearing restitution and normal anatomy preservation with an aim to prevent recurrent disease. Type of applied surgical procedure depends on the extent of cholesteatoma.

Localization of congenital cholesteatoma can be variable. CC confined to the anterosuperior quadrant develops within an area delineated by the neck and handle of the malleus laterally, and tensor tympani tendon and processus cochleariformis medially. Cholesteatoma does not adhere to the medial surface of the tympanic membrane [2]. Another commonly encountered site is the posterosuperior quadrant of the mesotympanum. CC extension in this site commonly leads to involvement of the long process of the incus and stapes suprastructure and/or spread into the attic via the tympanic isthmus. The latter site is less frequently encountered than the former [4].

Hao J., *et al*. [18] showed the most frequent site affected by CC to be the posterosuperior quadrant of the mesotympanum in 74.07%. They hypothesized that the majority of congenital cholesteatomas arise within the anterosuperior quadrant and subsequently migrate to the posterosuperior quadrant eroding the ossicular chain with eventual extension into the mastoid. In our study CC most frequently involved multiple quadrants with the exception of cholesteatoma confined to the posterosuperior quadrant in 4 cases.

Surgical approach and procedure depend on CC site and extension. The transcanal approach enabled by endoscopy ensures adequate exposure of the majority of CCs confined to the middle ear cavity. Mastoid involvement suggested by CT imaging mandates a postauricular approach with simple (cortical) mastoidectomy or CWU or CWD mastoidectomy and these options need to be discussed with the parent or caregiver [15].

Therefore, only small CCs confined to readily accessible areas of the middle ear may be removed using a transcanal approach. Extensive disease may mandate more aggressive surgery such as modified radical (CWD) mastoidectomy with removal of involved ossicles [19].

Canal wall up mastoidectomy is preferable to CWD mastoidectomy and should be performed when possible. However, it is not advisable to perform CWU mastoidectomy in certain situations, such as with extensive disease, a low-lying dura, anteriorly placed sigmoid sinus, or destruction of the bony ear canal, for which CWD may be better suited. Type I tympanoplasty was performed in patients with CC confined to the anterior quadrants (2 patients) and CC confined to the anterosuperior and posterosuperior quadrants, without ossicular chain involvement. The remaining 5 cases mandated CWU mastoidectomy along with type II tympanoplasty.

Extensive disease was found in 11 cases with concomitant involvement of the incus and stapes where type III or IV tympanoplasty was necessary (64%). Extracranial or intracranial disease extension was not found. CWD mastoidectomy using a

postauricular approach was performed in 8 (47%) patients. As per staging proposed by Postic., *et al*. these extensive cholesteatomas belonged to stage III and IV disease, and were commensurately managed using CWD mastoidectomy.

Incidence of ossicular chain involvement was 64%. The incus was most frequently involved (in 11 cases) followed by the stapes (in 4 cases). Other authors reported similar rates of ossicular chain involvement in the range of 65 to 76% [12,20].

In one case congenital cholesteatoma was confined to the mastoid which supports the assumption that CCs can develop in isolated parts of the temporal bone other than the middle ear cavity. Presentation of congenital cholesteatoma confined to the mastoid may be insidious allowing considerable disease extension and serious complications [21].

Incidental findings may lead to diagnosis as shown by our study.

Studies have shown poorer mastoid pneumatization in children with CC when compared to children without middle ear disease when adjusted for age, whereas lower degrees of mastoid pneumatization are found in the ipsilateral ear when compared to the contralateral unaffected ear in patients with congenital cholesteatoma [22]. In our study 30% of patients had poorly pneumatized mastoids.

Postoperative functional results depend on the type of applied ossicular chain reconstruction, condition of the tympanic membrane and degree of mastoid pneumatization [23]. As expected, superior functional results are achieved using a closed surgical technique. The average preoperative air-bone gap was 36,7 dB, whereas the average postoperative air-bone gap was 24.4 dB and ranged from 15 to 20 dB with CWU mastoidectomy as opposed to 22 to 28 dB with CWD mastoidectomy. Other authors reported worse thresholds with more extensive disease (Postic stage III and IV). ParkJ., *et al*. [19], reported postoperative air-bone gaps of 17.7 dB in stage I disease and 33.8 dB in stage III disease.

Our patients were monitored for two years postoperatively, and recurrent or residual disease was recorded in one case after CWU mastoidectomy and another after CWD mastoidectomy, a total of 12%; both patients presented with stage IV disease with signs of ETD in the contralateral ear and poorly pneumatized mastoids. Other authors report different rates of recidivism depending on localization and extension of disease and applied surgical technique. Postic., *et al*. reported recurrent cholesteatoma rates of 14% with disease confined to one quandrant, 33% with extension into 2 or more quadrants and up to 67% with mastoid involvement [24].

Hao J., *et al*. [18] reported recurrent cholesteatoma in 12.64% of children with CC. Other authors [25] reported recurrent or residual disease in 24% of patients. Higher rates of cholesteatoma recurrence are recorded with greater disease extension involving more than one quadrant (46%) as opposed to disease confined to one quadrant (15%). Congenital cholesteatoma extension measuring ≥4 mm was significantly associated with a greater likelihood of recurrent or residual cholesteatoma.

Conclusion

In conclusion, congenital cholesteatoma is characterized by insidious onset with predominantly ipsilateral CHL, which is why in many cases patients are erroneously treated for otitis media with effusion prior to establishment of correct diagnosis. Furthermore, the insidious nature of disease calls for a high index of suspicion in ORL specialists and indeed pediatricians performing newborn hearing screening and hearing tests in preschool children as CCs are most commonly diagnosed by that age. Disease stage is a significant risk factor for recurrence.

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