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Case Series

Middle Ear Congenital Cholesteatoma

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Abstract

Background: Although defined by strict criteria, congenital cholesteatoma can take several aspects.

The aim of this study is to describe the epidemiologic, clinical, and evolutive features of a series of patients managed for congenital cholesteatoma.

Methods: This is a retrospective study about nine patients managed for congenital middle ear cholesteatoma during 20 years. All the patients satisfied the criteria suggested by Levenson. Potsic's classification was used to define the stage. The evolution was appreciated on clinical, audiometric and radiological criteria.

Results: The mean age at diagnosis was 15 years [9-32 years]. Clinical examination revealed a whitish retrotympanic mass in 5 patients. Audiometry showed a conductive hearing loss in 7 patients with a mean threshold level of 56 db ± 14 db, an average air-bone gap of 45 db ± 8 db.

High-resolution temporal bone computed tomography showed cholesteatoma aspect in five patients.

All cases were classified in stage 3 and 4 of Potsic. An intact canal wall mastoidectomy was performed on 8 patients. Residual cholesteatoma was noticed in two cases on a mean follow-up of 5 years. A mean value of 28 db ± 18 db postoperative air-bone conduction gap was obtained in six patients with conductive hearing loss.

Conclusions: Congenital cholesteatoma is an underestimated diagnosis in our practice. Systematic screening by otoscopic examination at preschool age would be a pathway to earlier diagnosis and optimal management.

Keywords: Congenital cholesteatoma; Middle ear; Outcome

Abbreviations: ABG: Air Bone Gap; CHL: Conductive Hear Loss; db: Decibel; Hz: Hertz; HR CT: High Resolution Computed Tomography; MRI: Magnetic Resonating Imaging; CC: Congenital Cholesteatoma

Introduction

Since 1683, when cholesteatoma was first described by Du-Verney, many authors had suspected the congenital origin of cholesteatoma in some patients. It was only in 1965 that Derlacki and Clemis published a report of criteria distinguishing congenital middle ear cholesteatoma from acquired one. Later in 1989, Levenson modified these criteria and established the definition of Congenital Cholesteatoma (CC) that is at present widely accepted [1].

Congenital cholesteatoma accounts for approximately 2% to 5% of all cholesteatomas.

It is therefore a rare entity but currently better known because it is growingly reported in literature. Despite this, we notice that CC remains little known in our practice.

The purpose of this article is to report our experience in the management of 9 cases of congenital cholesteatoma.

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Methods

This study is a retrospective analysis of the medical dossiers of patients who underwent surgery for CC in our department, from January 1996 to December 2016.

The data collected were: epidemiologic, clinical, audiometric, radiologic and operative.

The patients satisfied the criteria suggested by Levenson for CC diagnostic [1]. We did not exclude patients with history of acute otitis media without otorrhea, and cases with diagnostic paracentesis.

Pre- and postoperative audiologic evaluations were performed on all patients. The mean hearing level was calculated from the average of threshold on the frequencies 500, 1000, and 2000 Hz.

Preoperative high-resolution temporal bone computed tomography (HR CT) was performed on seven patients.

CC was considered as an intraoperative diagnosis if it was not suspected on clinical nor radiological signs.

The surgeon's findings were recorded on the operative report, classifying the cholesteatoma into closed-type (cystic) or open type.

Middle ear quadrants affected by the disease, as well as ossicular chain involvement were precised on the operative report.

CC was classified according to Potsic staging system into:

- Stage 1, single quadrant: no ossicular involvement or mastoid extension.
- Stage 2, multiple quadrants: no ossicular involvement or mastoid extension.
- Stage 3, ossicular involvement: includes erosion of ossicles and surgical removal for eradication of disease; no mastoid extension.
- Stage 4: mastoid extension (regardless of findings elsewhere).

In this series, the attical involvement was considered as a stage 3: given the close relation of the cholesteatoma with the ossicles in this location, especially as in all the cases of extension to the attic, ossicular erosion was found.

Surgical technique and eventual ossiculoplasty have been specified.

The intraoperative data of a planned revision surgery (second look), or a functional time were also noted.

The evolution was assessed on clinical, audiometric and radiologic criteria.

Data were analysed using Excel software. We calculated simple frequencies and percentages for qualitative variables. We calculated averages; medians and standard deviations for the quantitative variables

Since it is a retrospective study, no consent to participate was needed.

Patient's files were analyzed respecting anonymity.

Results

Nine patients were included, the sex ratio was 1.2. The mean age of patients at time of diagnosis was 15 years, ranging from 9 to 32 years.

Eight of our patients complained of hearing loss, which was the most frequent symptom. In the 32-year-old patient, hearing loss was an audiometric discovery during a pre-employment test.

At micro-otoscopy, a whitish area behind an intact tympanic membrane was observed in 5 patients, two of them had a myringotomy confirming the suspected diagnosis. Table 1 summarizes epidemiologic and clinical data.

Table 1: Epidemiologic and clinical data.

	Sex	Age at diagnosis	Hearing loss	Side	Micro-otoscopy	Myringotomy
Case 1	M	11	yes	R	WRTM	no
Case 2	F	9	yes	L	NTM	no
Case 3	M	20	yes	R	NTM	no
Case 4	F	13	yes	R	WRTM	no
Case 5	F	15	yes	R	WRTM	no
Case 6	M	10	yes	L	WRTM	yes
Case 7	M	32	yes	R	NTM	no
Case 8	F	13	yes	L	NTM	no
Case 9	M	9	yes	L	WRTM	yes

M: Male; F: Female; R: Right; L: Left; NTM: Normal Tympanic Membrane; WRTM: Whitish Retro-Tympanic Mass

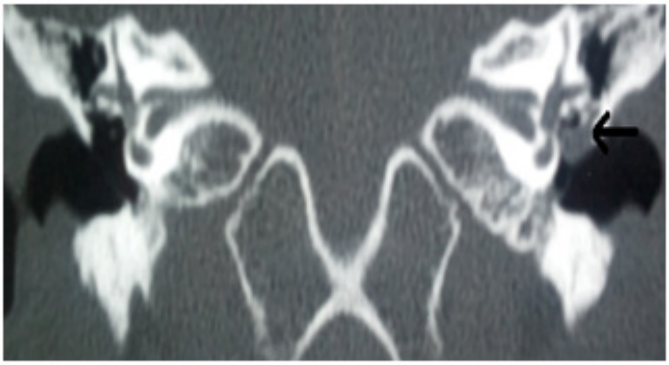

Figure 1: Coronal cut of CT scan showing a left congenital cystic cholesteatoma located in the posterior superior quadrant in relation with the incudo-malar joint which seems eroded (black arrow).

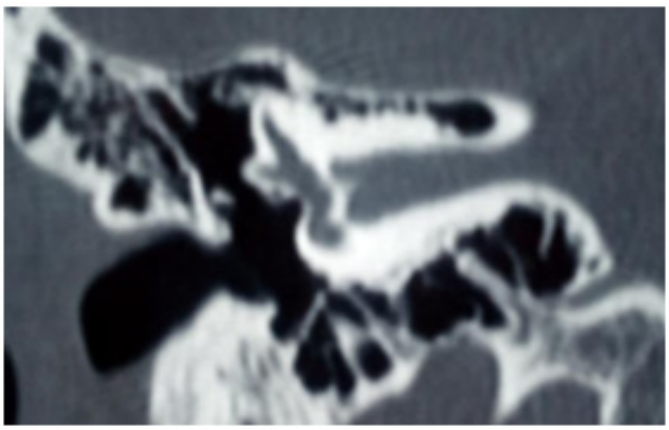

Figure 2: Coronal cut of CT scan of an open type right congenital cholesteatoma showing lysis of the long process of the incus and stapes and no evidence of classic cystic cholesteatoma mass (case 4, diagnosed per operatively by transcanalar approach).

Table 2: Surgical findings and procedure.

	Surgery type	Extension: Number of quadrants	Ossicular erosion	Ossicular removal	Ossicular reconstruction	Revision surgery	Residual disease
Case 1	CWUM	2+mst	In+St	Ml head	Cart Col	Planned 2nd look	Antral pearl
Case 2	CWUM	2	In	In	Cart Col	_	
Case 3	CWUM	1	In+St	In	Cart Col	Planned 2nd look	
Case 4	CWUM	2	In+St	In	TORP	_	
Case 5	CWUM	2+mst	Ml+In+St	Ml head+In	Oss Col	Residual on imaging	Atrial pearl
Case 6	CWUM	1+mst	None	Ml + In	Cart Col	_	
Case 7	CWUM	1	In+St	In	_	_	
Case 8	CWDM	2	In+St	_	Oss Col	Planned 2nd look	
Case 9	CWUM	4+mst	In+St	_	_	_	

CWUM: Canal Wall up Mastoidectomy; CWDM: Canal Wall Down Mastoidectomy; mst: Mastoid; In: Incus; Ml: Maleus; St: Stapes; Cart Col: Cartilage Collumel; Oss Col: Ossicular Collumel

Table 3: Audiometric outcome.

	Preoperative threshold (dB)	Preoperative air- bone gap (dB)	Stapes conservation	Ossicular reconstruction	Postoperative threshold (dB)	Follow up duration (months)
Case 1	67	30	_	Yes	55	192
Case 2	45	40	Yes	Yes	55	36
Case 3	71	52	Yes	Yes	70	72
Case 4	67	50	_	Yes	20	4
Case 5	70	50	_	Yes	20	48
Case 6	30	30	Yes	Yes	20	48
Case 7	37	50	_	_	30	2
Case 8	75	45	_	Yes	65	18
Case 9	50	47	_	_	33	120

Pure tone audiometry revealed in the affected ear, a Conductive Hearing Loss (CHL) in 7 patients with a mean threshold level of 56 db ± 14 db, and an average air-bone gap (ABG) of 45 db ± 8 db. Mixed hearing loss was found in two patients (including located lesions to the oval window in one case) with a mean threshold level of 70 ± 6 db, an average ABG of 37.5 ± 10 db.

HR CT was performed on 7 patients. It showed a feature of cystic cholesteatoma in five patients (Figure 1). In the other two cases, only ossicular erosion had been identified and diagnosed as an ossicular malformation (Figure 2).

Two patients had no imaging, the first one had a CHL with an intact ear drum and was proposed for a middle ear exploration in 1998 (case 8), the other one had a diagnostic myringotomy (case 6).

Six patients had preoperative diagnosis of CC, and the 3 others had intraoperative diagnosis. In fact, 3 patients had at first, a transcanal approach for suspected ossicular chain malformation and cholesteatoma was an intraoperative finding. It was an open type CC in two of them.

Eight of our patients underwent canal wall up mastoidectomy, only one needed for cholesteatoma removal, a canal wall down mastoidectomy.

In 7 patients, cholesteatoma was classified stage 3 of Potsic, in the remaining 2 cases it was classified stage 4. Only one patient had intact ossicular chain but mastoid involvement (case 6).

Ossicular reconstruction was performed at primary surgery in 5 patients and at revision surgery in two others.

Residual disease has been diagnosed in two patients. In one patient, discovered on a planned second look (1 year post operatively), it was an antral pearl. In the second patient, residual disease was revealed by MRI, supported by CT scan (Figure 3).

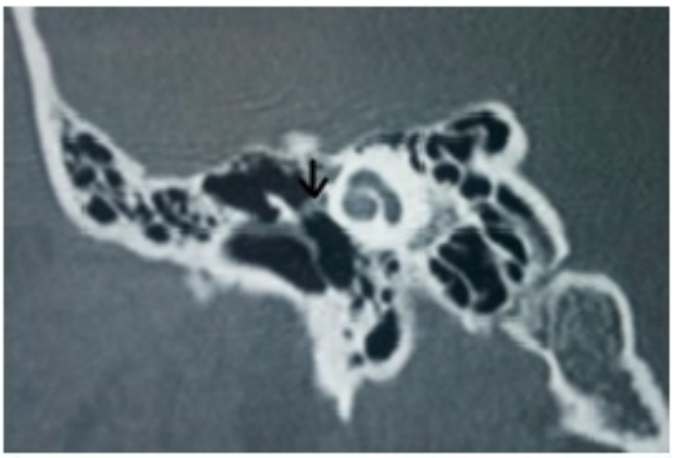

Figure 3: Coronal cut of CT scan showing right residual cholesteatoma pearl (black arrow) located internally to the malleus neck (case 5).

It was a cholesteatoma pearl located between the inner side of the malleus neck and the ossicular reconstruction. Table 2 summarizes surgical findings and procedure. Post operative microotoscopy showed intact reconstructed tympanic membranes in all patients.

Two patients were lost to follow up after 2 and 4 months. The mean follow up was 5 years. Six patients with CHL presented an average of 28 db ± 18 db postoperative ABG, while one patient had a deterioration of hearing loss of 10 db.

In the two patients who had mixed hearing loss, a postoperative hearing gain of 10 db was found in one patient and a deterioration of hearing loss of 14 db in the other. Table 3 summarizes audiometric outcomes according to ossicular status.

Discussion

CC is commonly an expanding cystic mass with a keratinizing squamous epithelium located medially to intact membrana

tympana. It is supposed to be present at birth, but is generally diagnosed in early childhood in patients with no prior history of otorrhea, perforation, or previous ear surgery [2].

A history of effusion or previous periods of acute otitis media does not exclude congenital cholesteatoma.

Even though this definition is the recent one taken literally from the last consensus paper on cholesteatoma, definition criteria for CC still match Levenson's one [1].

The clinical presentation of CC depends on location and extent of lesion. It can be distinguished by:

- a whitish mass behind an intact tympanic membrane
- a hearing loss when it spreads through all the middle ear or erodes the ossicles.
- pain (extremely rare), and/or CT/MRI findings [2].

There is a subset of CC originating in the anterior superior quadrant of the middle ear. This location was the most typical in many studies [1,3].

But many CC spared this quadrant, and were located in the posterior superior one. In several more recent series, the CC was more frequently located in the posterior superior quadrant of the middle ear [4,5].

The CC of the middle ear can therefore originate from the anterior posterior-superior quadrants and then gradually extends to the rest of the middle ear cavities.

In our series, over 21 years of practice, only 9 cases of congenital cholesteatoma were diagnosed and treated. This is relatively low compared to other published series.

In fact, congenital cholesteatoma diagnosis could be underestimated in favor of acquired cholesteatoma. Indeed, the otorrhea that some authors consider the ultimate stage of evolution of congenital cholesteatoma, essentially in the young child, could be considered as an acquired cholesteatoma when the patient is seen late, especially by an inexperienced doctor.

The median age at diagnosis was 15 years ranging from 9 to 32 years in our series. This average age is older than reported in European series (6.5 years) and also American series (approximately 4 years old) [6-9]. Younger age reported in American series could be explained by the systematic otological examination via otoscopy for CC screening before getting the appropriate age to undergo audiometric exploration. Potsic reported the most frequent clinical presentation, to be a white middle ear mass (82%) [3].

The relatively old age at diagnosis in this series could be explained by several factors. Firstly, unilateral hearing loss is difficult to note in children. Many factors related to cholesteatoma can also explain it: because of the scarcity of presenting symptoms, the anterior superiorly located CC could avoid detection for a long time before posterior extension which would cause hearing loss [3,9,10].

Hearing impairment was the main complaint in this series as in several published series [6,8,9,11].

At micro-otoscopy, the ear drum appeared to be intact in 4 patients. Actually, a conductive hearing loss, on a normal tympanic membrane, especially if it is unilateral; highly suggests CC diagnosis. Then imaging data play a significant role [8,9,12].

Hearing impairment is rarely noticed when the lesions involve the anterior superior quadrant; It is rather observed in case of posterior involvement to the ossicles or to the eustachian tube with secondary effusions [8,12,13].

Cholesteatoma that develops in the posterior superior quadrant is considered as a hearing loss provider from the outset, given its proximity to ossicles [10].

HR CT showed in this study a feature arguing for congenital cholesteatoma in only five patients. In the other two cases, only ossicular erosion had been identified and cholesteatoma was identified intraoperatively. It was an open type CC in the two cases.

HR CT is the preferred imaging method, when CC is suspected. This imaging has a high sensitivity, due to its excellent spatial resolution. Nevertheless, its specificity is lower, especially in case of a mass lesion, which may correspond to granulation tissue, secretion, cholesterol granuloma, or neoplasm [14,15]. Ossicular erosion typically associated with acquired cholesteatoma, is sometimes absent in the congenital cholesteatoma making its diagnosis less obvious [14].

HR CT can be useful for analyzing CC extension beyond what is noticed on micro-otoscopy [12].

As in acquired cholesteatoma, in congenital one, MRI contribution is due essentially to its ability to study labyrinthine involvement and intracranial space [15].

Still, MRI's most important value is detecting postoperative residual or recurrent cholesteatoma and avoiding systematic second look [15].

In this series, the attical involvement was considered as a stage 3, especially as in all these cases, ossicular erosion was noticed.

This leads us to highlight a possible inaccuracy in the classification of Potsic.

The classification proposed by Nelson in 2002, not adopted in our study, is more explicit regarding the attic involvement [13]:

Type 1: mesotympanum with no incus or stapes erosion.

Type 2: mesotympanum or attic with ossicular erosion, but no mastoid extension.

Type 3: mesotympanum with mastoid extension.

In this series, all patients were classified stage 3 or 4 of Potsic's classification. With Nelson's Classification, they would be classified in stage 2 or 3, since stage 1 in Nelson's classification represents stage 1 or 2 of Potsic's one.

For some authors, age appeared to be correlated to disease extension. They found that extensive cholesteatoma increased significantly with age. Even though it remains unclear if the cystic mass grows and changes by rupture into the diffuse form or if new extensive lesion tends to develop with aging [16].

In terms of surgery, most authors consider the conservative technique, via canal wall up tympanoplasty, the gold standard [9,11].

Congenital cholesteatoma does not tend to recur as frequently as acquired one. In fact, inflammation is minimal in the

CC and temporal bone pneumatization is often good. So that recurrence occurs only in cases of important pathologic changes of the mucosa [8,11].

Several publications suggest removal of the entire portion of the tympanic membrane having close contact with cholesteatoma, and perform tympanic membrane transplantation as often as possible [8,9].

Several authors correlate the functional results with the initial state of the ossicular chain, especially stapes: the results tend to be better in case of intact stapes. Benhammaou reported a postoperative ABG of 15 to 20 db in case of stapes preserved and 30 to 40 db in case of destructed stapes [8].

Aimi reported a residual cholesteatoma level of about 50%. Aimi attributes this high rate, on one hand, to extensions to difficult accessing areas (regions of the windows, sinus tympani, epitympanic recess), and on the other hand to the open type CC, where the limits between matrix and normal mucosa is unclear [8,11].

Nelson reported a recurrence rate of 0% in the case of middle ear involvement without ossicular lysis (excepted the manubrium of the malleus), 34% in the case of postero-superior quadrant or attic involvement and 56% in case of mastoid involvement [13].

Lazard and al considered that CC extending to more than one quadrant, is as likely to develop residual disease as acquired cholesteatoma [6]. They also considered the extension to the eustachian tube and a non-reconstructed atticotomy as risk factors to secondary retraction pockets. Hence, they recommend tympanic membrane reinforcement and systematic reconstruction of scutum in CC extended to more than two quadrants [6].

For Stapleton, the condition of the cholesteatoma matrix determines the surgical procedure and the prognosis. Ossicles should not be removed if matrix is intact. In case of matrix rupture, and if the cholesteatoma is abutting or surrounding the incus, or internal to the malleus and incus, or covering the stapes, or eroding the ossicles; then the involved ossicle(s) should be removed [17].

Finally, for Potsic, mastoid extension and/or ossicular involvement are the predictor factors of residual disease [7].

Our sample in too small and all cases are stage 3 and 4, since diagnosis is late in our practice. Therefore, we have no possibility of analyzing outcome according to extension in our study.

Conclusion

With the low frequency given, congenital cholesteatoma seems to be under diagnosed in our practice. In view of the advanced age at the time of diagnosis and absence of early stages, congenital cholesteatoma is diagnosed late in our practice.

Congenital cholesteatoma is not as rare as believed; whenever suspected, CT scan can be of great help to diagnosis.

Author Statements

Competing Interest

The authors declare no competing interest.

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