International Journal of Pediatric Otorhinolaryngology 78 (2014) 2205–2209

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Open vs closed type congenital cholesteatoma of the middle ear: Two distinct entities or two aspects of the same phenomenon?

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ARTICLE INFO

Article history:
Received 19 July 2014
Received in revised form 4 October 2014
Accepted 10 October 2014
Available online 22 October 2014

Keywords:
Congenital cholesteatoma
Middle ear
Surgery
Children
Residual disease

ABSTRACT

Objective: The clinical features and surgical results of "closed type" versus "open type" congenital cholesteatoma were compared in order to analyse the differences between the two forms; whether the morphology of the disease may have a role in the staging systems has been also evaluated.
Patients and methods: We reviewed retrospectively 95 patients (96 ears) who underwent surgery for congenital cholesteatoma over a 15-year period focusing on the clinical differences between open and closed type congenital cholesteatoma.
Results: Seventy-one patients (74%) had a closed-type and 25 (26%) an open type congenital cholesteatoma. Our study confirmed the higher prevalence of the closed type, as well as, a younger age at initial diagnosis compared with the open type congenital cholesteatoma. Other differences between the two forms were: modality of diagnosis (pathognomonic otoscopy in 100% of the closed type and in 40% of the open type), positive history for otitis media with effusion (51.4% in closed type vs 20% in open type), involvement of the tympanic membrane quadrants (anterior quadrants were more frequently involved in the closed forms, whereas posterior quadrants were more frequently involved in the open forms), disease extension and aggressiveness. A residual cholesteatoma was found in 6 out of the 71 patients (8.4%) with a closed type congenital cholesteatoma and in 10 out of the 25 patients (40%) with an open type congenital cholesteatoma. After adjusting for potential confounders, open-type congenital cholesteatoma was significantly associated with residual cholesteatoma compared to the closed-type (odds ratio [OR] 7.39, 95% confidence interval [CI] 1.10–49.77, $p = 0.03$).
Conclusion: This study confirmed that the open congenital cholesteatoma has global clinical features that are uniquely different from the classical closed form. These differences could reflect a distinct pathogenesis, but there is no proof of this to date. The classification of the congenital cholesteatoma could be further refined by adding the morphologic type of the disease.

1. Introduction

Congenital cholesteatoma (CC) is a relatively uncommon disease accounting for 2 to 5% of all cholesteatomas [1]. Diagnostic criteria for CC, proposed by Derlacki and Clemis [2] and successively modified by Levenson et al. [3] are: a white mass behind an intact eardrum; no history of tympanic membrane perforation, otorrhea or previous ear surgery. In spite of numerous studies, the pathogenesis of CC remains an area of controversy and some of the details are still subject of much debate. Among the different etiopathogenetic theories proposed in the literature, the most credited explanation for CC is that it originates from rests of epidermoid cells incorporated into the temporal bone during embryonic development [4,5]. In most cases, CC origins from the antero-superior part of the tympanic cavity and, over time, it grows with possible extension to the remainder of the tympanic cavity, attic, antrum and mastoid. However, it has been recognized that CC is also common in the postero-superior part of tympanic cavity, especially in Asian patients [6,7]. Histopatholgical and clinical studies demonstrated that CC can present as an epithelial cyst without exposure of keratin or can develop as flat keratinizing epithelium [8,9]. In the first circumstance, it is classified as "closed type" CC (CTCC), whereas in the second situation it is classified as "open type" CC (OTCC). Whether the two distinct types have a different etiopathogenetic origin or are a different evolution of the same phenomenon is uncertain. Some authors postulated that CC

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http://dx.doi.org/10.1016/j.ijporl.2014.10.014
0165-5876/

has a cystic origin and gradually evolves to an open form because of a rupture of the closed pearl [10]. Others proposed that in most cases CC is the open type and that middle ear inflammation contributes to the formation of CTCC [11]. Usually, OTCC is diagnosed at an older age, it may be more difficult to identify behind an intact tympanic membrane, and its surgical removal is more challenging compared with CTCC. Very few studies have specifically investigated the differences in residual disease rate between OTCC and CTCC [12]; in addition, histopathological typing has not been considered in the classification systems proposed in the literature. The aims of this study were to investigate clinical features and differences in residual disease rate between OTCC and CTCC, and to understand whether morphologic type may have a role in the classification systems or not.

2. Materials and methods

A retrospective study was performed by analyzing our database for patients operated on for CC in a 15-year period and followed up for at least 5 years after their last operation. Although this study has a retrospective nature, it is based on a database that has been accomplished prospectively, permitting reliable clinical and surgical comments. The study has been conducted according to the principles expressed in the Declaration of Helsinki. An informed written consent has been obtained from all patients. All cases satisfied the criteria of having an intact tympanic membrane with no history of tympanic membrane perforation, otorrhea or previous ear surgery. Patients with a history of otitis media with effusion (OME) were not excluded. All patients underwent preoperative audiologic evaluation and high resolution computed tomography of the temporal bone. Preoperative characteristics analyzed included: age and presenting symptoms at the time of initial diagnosis, sex, side, otoscopic findings, status of the controlateral ear, and history of OME. Intraoperative findings included: location, extension, and morphologic type of the disease, ossicular chain status, presence of labyrinthine fistula, fallopian canal erosion or bony defect of the middle cranial fossa, and surgical technique involved. CC was considered as closed type when it presented as an epithelial cyst without exposure of keratin (Fig. 1), and as open type when it developed as flat keratinizing epithelium without formation of an epithelial cyst (Fig. 2). CC was staged by using the classification system advocated by Potsic et al. [13] (Table 1). The incidence of residual cholesteatoma was evaluated in both closed and open form.

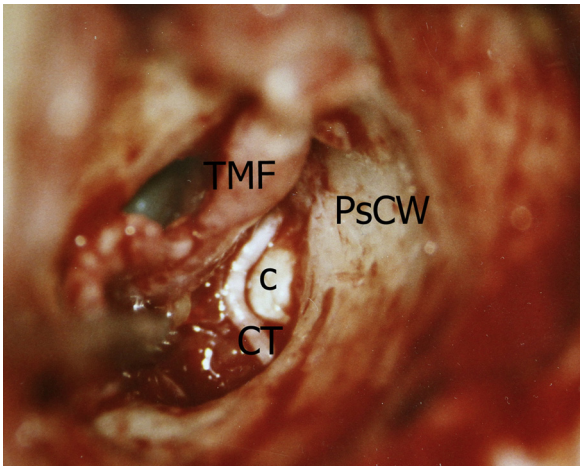

Fig. 1. Closed type congenital cholesteatoma. Intraoperative view of a left ear. The cholesteatoma (c) is located in the postero-superior portion of the tympanic cavity. PsCW: postero-superior canal wall; CT: chorda tympani; TMF: tympanomeatal flap.

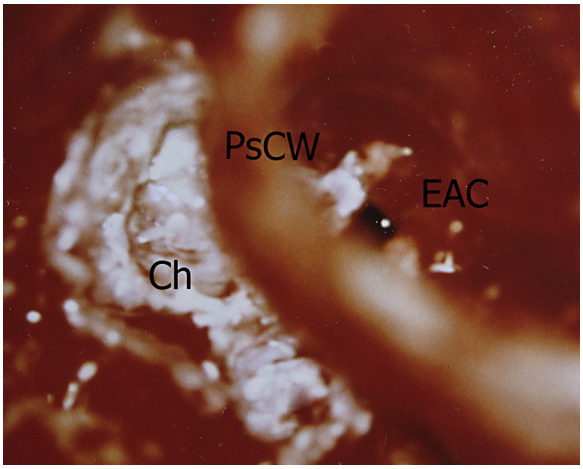

Fig. 2. Open type congenital cholesteatoma. Intraoperative view of a right ear. The cholesteatomatous matrix (Ch) extends from the posterior mesotympanum to the mastoid cavity. PsCW: postero-superior canal wall; EAC: external auditory canal.

The surgical approach was single stage transcanal or post-auricular tympanoplasty for stage 1 and 2 cholesteatomas. A preplanned staged canal wall up tympanomastoidectomy was perfomed in all stage 3 and 4 cholesteatomas. In order to prevent secondary retraction pockets after surgery for CC, we reconstructed all defects of the postero-superior canal wall using bone patè, as previously described [14]; in addition, in order to decrease the chance of residual disease, in all cases we used Mesna, a mucolytic agent capable to break disulfur bonds and to make the cholesteatoma removal easier [15–17].

In patients who underwent single stage surgery (stage 1 and 2 CCs), the diagnosis of residual cholesteatoma was based on postoperative otoscopy.

For descriptive purpose, baseline characteristics of the two groups were compared using a χ^2 test and ANOVA model for categorical and continuous variables, respectively. Bivariate tests were used: CHI-square for dichotomic variables was used to test the significance of categorical covariates. Parametric (ANOVA) and non-parametric tests (Kruskal–Wallis, median test and *U*-test of Mann–Whitney) were used to test the significance of continuous covariates. Logistical regression analysis was used to compare residual disease rate between the two forms, after adjusting for age, sex and other covariates. Statistical analysis was carried out using "SAS 8.2 version" statistical software. The results with a value of $p < 0.05$ were considered significant.

3. Results

Ninety-six CC were identified in 95 patients; there was one bilateral case. In line with published literature, there was a high preponderance for males (66%) and no predilection for either side (53% right and 47% left). Mean age at initial diagnosis was 7.7 years (range, 3–18 years) with a significant difference ($p < 0.0001$)

Table 1
Potsic et al. [13] staging system.

Stage	Description
1	Single-quadrant disease without ossicular involvement or mastoid extension
2	Disease involving multiple quadrants without ossicular involvement or mastoid extension
3	Ossicular involvement, defined as erosion of ossicles or surgical removal for eradication of disease
4	Disease with any mastoid extension (regardless of finding elsewhere)