Classification of Cholesteatoma According to Growth Patterns

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IMPORTANCE  Several classifications of cholesteatoma exist, but there are controversies about their clinical application.

OBJECTIVE  To classify cholesteatomas and describe the prevalence of the subtypes.

DESIGN, SETTING, AND PARTICIPANTS  A cross-sectional comparative study of 414 ears in 356 consecutive patients with middle ear cholesteatoma and no history of ear surgery treated at a tertiary hospital was conducted from March 8, 2000, to March 30, 2015. Data analysis was conducted from March 30, 2014, to March 30, 2015.

INTERVENTION  Otoendoscopy was conducted, and findings for both ears were recorded.

MAIN OUTCOMES AND MEASURES  Cholesteatoma growth patterns were classified as anterior epitympanic, posterior epitympanic, posterior mesotympanic, 2 routes (both the pars flaccida and the pars tensa are involved), and undetermined.

RESULTS  Among the 356 patients in this study, mean (SD) patient age was 33.23 (19.81) years (range, 4-82 years), and 125 patients (35.1%) were female. The right ear was identified as the affected ear in 220 patients (61.8%). A total of 272 (65.7%) ears were from adults. Of the 414 ears that underwent otoendoscopy, posterior epitympanic (142 [34.3%]) and posterior mesotympanic (140 [33.8%]) were the most frequent types of cholesteatoma observed, followed by undetermined (67 [16.2%]) and 2 routes (57 [13.8%]). Anterior epitympanic type was the least frequent (8 [1.9%]). Posterior epitympanic cholesteatoma was more prevalent in adults (111 [40.8%]), whereas posterior mesotympanic cholesteatoma was more frequent in children (43.0%) \((P < .001)\). Anterior epitympanic cholesteatoma was observed only in children.

CONCLUSIONS AND RELEVANCE  Classifying cholesteatomas according to the growth pattern (anterior epitympanic, posterior epitympanic, posterior mesotympanic, 2 routes, and undetermined) includes all existing types of cholesteatomas of the middle ear. In general, the prevalence of posterior epitympanic and posterior mesotympanic cholesteatoma were similar. Whereas anterior epitympanic and posterior mesotympanic cholesteatomas were more prevalent in children, posterior epitympanic cholesteatoma was more frequent in adults.

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Cholesteatoma is characterized by the accumulation of exfoliated keratin debris in the middle ear or other pneumatized areas of the temporal bone. Since cholesteatoma was first described, many classifications have been proposed. In general, cholesteatomas can be classified into 2 categories: congenital and acquired. Since congenital cholesteatomas are very rare, the existing classifications are applied to the acquired type. The classifications are based on otomicroscopic appearance, typical growth patterns, disease extension, surgical findings, and drum status as determined on otoscopic examination. However, there are still controversies about the clinical application of each of those classifications. Moreover, the real prevalence of each kind of cholesteatoma is still unknown. Although studies have systematically described the attic or posterior epitympanic cholesteatoma as the most frequent kind, a more recent study observed a greater prevalence of pars tensa cholesteatoma.

Several studies have found significant differences between cholesteatomas in children and in adults. Some authors have hypothesized that cholesteatomas in children may be more aggressive and can recur more frequently after surgery. To our knowledge, however, there has been no report as to whether cholesteatomas in children and adults are different according to prevalence of the typical growth patterns.

In this study, we aim to review all videotoscopic records of patients with cholesteatoma and classify them according to cholesteatoma growth patterns. The secondary objectives are to describe the prevalence of the cholesteatoma subtypes and to verify whether there are differences between children and adults in the prevalence of subtypes.

**Methods**

This prospective study from March 8, 2000, to March 30, 2015, included 414 ears of 356 consecutive outpatients treated at a tertiary hospital who met the inclusion criterion of the presence of cholesteatoma in at least 1 middle ear. The exclusion criteria were a previous history of any ear surgery except tympanostomy for ventilation tube placement and impossibility of cleaning the ear canal and performing videotoscopy for appropriate documentation. Detailed clinical history was obtained, and otologic examination was performed. Careful and detailed cleaning of the ear canal was performed before the examination. Fiberopticotoendoscopy in both ears was recorded sequentially.

The recorded images were independently reviewed by the same researcher (L.S.R). Cholesteatoma growth pattern was classified according to the Jackler classification:

1. Attic or posterior epitympanic (cholesteatoma confined to the pars flaccida; Figure 1A);
2. Tensa or posterior mesotympanic (cholesteatoma in the posterosuperior quadrant of the pars tensa; Figure 1B); and
3. Anterior epitympanic (cholesteatoma originates cranially and anteriorly to the malleus head; Figure 1C);
4. 2 Routes cholesteatoma (the pars flaccida and the pars tensa were both involved).

**Figure 1. Cholesteatoma Growth Pattern**

A. Posterior epitympanic cholesteatoma
B. Posterior mesotympanic cholesteatoma
C. Anterior epitympanic cholesteatoma
D. 2 Routes cholesteatoma

A. Posterior epitympanic cholesteatoma is confined to the pars flaccida. B. Posterior mesotympanic cholesteatoma arises in the posterosuperior quadrant of the pars tensa. C. Anterior epitympanic cholesteatoma originates cranially and anteriorly to the malleus head. D. 2 Routes cholesteatoma (the pars flaccida and the pars tensa were both involved).
3. Anterior epitympanic (cholesteatoma originating cranially and anteriorly to the malleus head; Figure 1C).

For comparisons, the study population was divided into the pediatric group, comprising patients aged 18 years, 11 months, and 30 days (United Nations Convention on the Rights of the Child, 1989) or younger, and the adult group, with patients 19 years or older.

The procedures followed were in accordance with the ethical standards of the responsible institutional committee on human experimentation and with the Declaration of Helsinki. The Hospital de Clínicas de Porto Alegre Research Ethics Committee approved this study. All participating patients, or their parents or guardians when the participant was a child, provided written informed consent before their inclusion in the study.

Data analysis was conducted from March 30, 2014, to March 30, 2015. Statistical analysis using the \( \chi^2 \) test and Fisher exact test was performed using SPSS, version 20.0 (SPSS, Inc). Statistical significance was set at \( P < .05 \).

Results

Mean (SD) patient age was 33.23 (19.81) years (range, 4-82 years) and 125 patients (35.1%) were female. The right ear was identified as the affected ear in 220 patients (61.8%). A total of 272 (65.7%) ears were from adults.

The prevalence of cholesteatoma growth patterns in the study population is shown in Figure 2. Posteriorepitympanic (142 ears [34.3%]) and posterior mesotympanic (140 [33.8%]) were the most frequent cholesteatomas observed. Anteriorepitympanic type was the least frequent (8 [1.9%]).

However, 124 of the cholesteatomas (30.0%) could not be classified as posterior epitympanic, posterior mesotympanic, or anterior epitympanic. We observed that, in 57 ears (13.8%), both the pars flaccida and the pars tensa were involved, so we called them 2 routes cholesteatoma (Figure 1D). Finally, in 67 ears (16.2%), no precise growth pattern could be identified by videotoscopy. We classified these cholesteatomas as undetermined.

In terms of sex, the prevalence of children and adults were similar (males, 49 [38.3%] vs 118 [51.8%]; \( P = .10 \)) and affected ear (right ear affected, 75 [32.4%] vs 145 [53.5%]; \( P = .92 \)). However, in terms of cholesteatoma growth patterns, posterior epitympanic cholesteatoma was more prevalent in adults (111 [40.8%]), whereas anterior epitympanic (7 [4.9%]) and posterior mesotympanic cholesteatoma (61 [43.0%]) were more prevalent in children (\( P < .001 \)). In fact, anterior epitympanic cholesteatoma occurred only in children. The prevalence of 2 routes and undetermined cholesteatoma was similar between both groups (Figure 3).

Discussion

Acquired cholesteatomas are classically divided into attic, originating from the Shrapnell membrane, and sinus, originating from the posterosuperior retraction of the pars tensa. Later, Tos proposed a widely accepted classification that further subdivided the pars tensa cholesteatoma into sinus cholesteatoma, with primarily posterior retraction and posterior pathologic findings, and tensa retraction cholesteatoma, with primary retraction of the entire pars tensa and primary pathologic findings in the anterior, inferior, and posterior parts of the tympanic cavity.

Comparing the Tos and Jackler classifications, posterior epitympanic cholesteatoma is the equivalent of attic cholesteatoma. According to Jackler, this route is the most common and passes through the superior incudal space, which lies lateral to the incus body, and then traverses the aditus ad antrum to enter the mastoid. Posterior epitympanic cholesteatoma usually reaches the middle ear by descending through the floor of the lateral epitympanic space (Prussak space) into...
the posterior pouch of von Troeltsch. In our study, we observed a similar prevalence of 34.3% for posterior epitympanic and 33.8% for posterior mesotympanic cholesteatomas. These findings are in agreement with those of a previous study that found a prevalence of 45% for pars tensa and 41% for attic cholesteatomas.6

When we analyze the pars tensa cholesteatoma, however, the classifications diverge. While Jackler4 considered only posterior mesotympanic cholesteatoma that originated from the posterior portion of the pars tensa retraction, Tös7 sub-divided the pars tensa cholesteatoma into sinus, which is the equivalent to posterior mesotympanic, and tensa. We agree with the posterior mesotympanic concept because it refers to a typical route of extension in which, in contrast to posterior epitympanic, the mastoid progression of cholesteatoma typically passes medial to the malleus and incus, and the sinus tympani and facial recess are commonly involved. When the entire area of the middle ear is affected by the cholesteatoma, however, it is difficult to determine precisely whether the disease is the result of a complete pars tensa atelectasis (tensa cholesteatoma) or if it is a posterior mesotympanic or even a posterior epitympanic cholesteatoma that has advanced to other compartments. For such reasons, we preferred to classify unknown cases as undetermined.

It is not unusual for multiple cholesteatoma sacs to occur in the same ear involving 2 or even all of the typical routes.4 Passing through the anterior tympanic isthmus is the main route of drainage and aeration of the attic rooms, which can be occluded by mucosal edema, thick mucus plugs, or even retraction of the posterior portion of the pars tensa, ultimately resulting in ventilation problems in the epitympanum.18–20 Black and Gutteridge6 reported a prevalence of 14% for combined attic and pars tensa cholesteatoma, which we named 2 routes. In our study, the prevalence of 2 routes cholesteatoma was 13.8%.

The classification of cholesteatomas into congenital and acquired is useful since it separates 2 types of cholesteatoma with distinct pathogenesis and biological behavior. However, sometimes it is difficult to clinically determine if the cholesteatoma is congenital or acquired. First, congenital cholesteatoma is rare, accounting for approximately 4% of childhood cholesteatomas and 2% to 5% of all cholesteatomas.21,22 Second, the classic definition by Derlacki and Clemis23 of congenital cholesteatoma as a pearly mass medial to an intact tympanic membrane, a normal pars tensa and pars flaccida, and no history of otorhea, tympanic membrane perforation, or previous otoplastic procedures has been questioned mainly because children with congenital cholesteatoma can have middle ear infections and associated complications.24 In our study, we found a prevalence of 1.9% of anterior epitympanic cholesteatoma. This cholesteatoma growth pattern was described by Jackler4 as an epitympanic retraction that forms anterior to the malleus head following the embryologic course of either the saccus anticus or the anterior sacculus of saccus medius. In our study, all of the cases of anterior epitympanic cholesteatoma were observed in children. Moreover, another analysis of these patients showed that all of the contralateral ears were healthy.25 For these reasons, we hypothesize that this cholesteatoma growth pattern may be congenital, even with pars flaccida retraction and history of otorhea. The most prevalent location of congenital cholesteatoma has been reported as lying within the anterosuperior quadrant,26 and the most accepted theory about its formation, the epithelial rest theory, is based on the presence of epidermoid residues in the anterosuperior quadrant of the fetal middle ear.27 The development of cholesteatoma in this specific position can lead to obliteration of the anterior portion of the tympanic isthmus, which in healthy ears is an open structure with no fold.28 This obliteration can cause pars flaccida retraction, or the cholesteatoma can simply expand superiorly and erode the attic.

In addition to our finding that anterior epitympanic cholesteatoma occurred only in children, we also observed that pediatric patients had a higher prevalence of posterior mesotympanic cholesteatoma, while adults had a greater prevalence of posterior epitympanic cholesteatoma. Chronic otitis media with effusion is frequent in children and is caused mainly by eustachian tube dysfunction. If poor middle ear ventilation persists, it is possible that some cases of chronic otitis media with effusion progress to moderate and severe posterior pars tensa retraction and then, in the absence of mechanisms to stop the process, to bone erosion and keratin accumulation. Although the pars flaccida is more fragile and thus more susceptible to retraction, the progression of the cholesteatoma seems to depend on a preexistent, perhaps congenital, alteration of the attic ventilation routes and a longer time for disease development when compared with posterior mesotympanic cholesteatoma. These findings could be the reasons why, even if both pathologic processes are initiated in childhood, pars tensa tympanic membrane retractions can progress more easily than attic retractions to formation of cholesteatoma, occurring earlier in the patient’s life.

Conclusions

After review of cholesteatoma classifications, it seems that the classification proposed by Jackler4 is the most encompassing. However, the classification of cholesteatomas, based on both videotoscopy and disease pathogenesis, into anterior epitympanic, posterior epitympanic, posterior mesotympanic, 2 routes, and undetermined, includes all of the existing types of cholesteatoma of the middle ear. In our study population, the prevalence of posterior epitympanic and posterior mesotympanic cholesteatoma were similar. Anterior epitympanic and posterior mesotympanic cholesteatomas were more prevalent in children, whereas posterior epitympanic cholesteatoma was more frequent in adults.
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