Turkish Archives of Otolaryngology
Türk Otolarengoloji Arşivi
Turk Arch Otolaryngol 2014; 52: 121-5

Comparison of Primary and Secondary Cholesteatomas in Terms of Ossicular Destruction and Complications

Original Investigation

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Abstract

Objective: Cholesteatomas are divided into two groups by their origin: primary (attic) and secondary (tensa). It is highly probable that these two types differ by etio-pathogenetic mechanisms. In this study, we aimed to compare these groups in terms of complications and ossicle destruction.

Methods: Sixty-six consecutive patients, operated on between June 2012 and March 2014 in our department for cholesteatoma, were included in this study. The status of the ossicles was scored according to the Austin-Kartush classification between 0-7, and sub-groups were created according to the magnitude of the ossicle damage: primary-A (PrA), primary-B (PrB), secondary-A (SecA) and secondary-B (SecB).

Results: Thirty-eight patients had secondary cholesteatomas, and 28 patients had primary cholesteatomas. The average ages for primary and secondary patients were 38.4 and 42.6, respectively. All 5 patients under the age of 16 had primary cholesteatomas. Austin-Kartush score averages for the primary and secondary groups were not statistically different. However, when the PrA and SecA groups (patients with less ossicular damage) were compared statistically, the SecA group was found to have a significantly higher average score than the PrA group, which reflects less damage. Furthermore, the number of patients with complications and the number of complications were significantly higher in the primary group.

Conclusion: Primary and secondary cholesteatomas seem to differ from each other etio-pathogenetically. Secondary cholesteatomas are diagnosed with less ossicular damage compared to their primary counterparts, and this finding is attributed to the fact that primary cholesteatomas can not drain into the external ear canal, which leads to more destruction and complications.

Keywords: Cholesteatoma, primary, secondary, ossicles, Austin-Kartush classification

Introduction

Cholesteatoma was defined by Friedmann (1) in 1959 as a cystic structure covered with stratified squamous epithelium that leans over a fibrous stroma containing some elements from the original mucous membrane. It was also defined by Schucknecht (2) in 1974 as a pathology characterized with stratified squamous epithelium and its product keratin debris being trapped in the tympanomastoid compartment that was named as keratoma. Cholesteatomas are divided into two groups: congenital and acquired. Acquired cholesteatomas are also divided into two groups as attic and pars tensa cholesteatomas depending on the tympanic membrane (TM) or tympanum part from which they originate. Attic cholesteatomas are called primary acquired cholesteatomas (PACs), whereas tensa cholesteatomas are called secondary acquired cholesteatomas (SACs) (3, 4). According to Tos (5), SACs are divided into two groups depending on the part of the pars tensa they originate from; if SACs originate from the postero-superior part of the pars tensa, they are called "sinus cholesteatoma," and if they originate from the remaining part of the pars tensa, then they are called "tensa cholesteatoma." Cholesteatomas, in which attic and tensa are held together, are called "combined cholesteatoma" by some authors (6). Primary and secondary cholesteatomas also show differences in terms of the etiology and pathological changes they exhibit. In PAC, it is observed that the pars tensa is intact, and a deep retraction pocket (cholesteatoma sac) extending to attic, probably due to chronic negative pressure is present. On the other hand, in SAC, generally on top of chronic middle ear infection, recurrent acute attacks and polyps, granulation tissues, and extensive retraction sacs that pathologically involve the pars tensa are observed. Despite different pathological origins, there are no significant differences between these two groups in terms of complications and clinical outcomes in the advanced stages of the disease (7, 8). Although there are publications that compare ossiculoplasty results and complication rates of these two subgroups (6, 7, 9), the pattern and extent of damage that both groups cause directly on the ossicles is a less studied topic. In this study, it is

This study was presented at the 3rd National Otology and Neurootology Congress, 1-4 May 2014, Antalya.

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Received Date/Geliş Tarihi: 28.05.2014
Accepted Date/Kabul Tarihi: 13.10.2014

DOI:10.5152/tao.2014.624

aimed to compare PAC and SAC in terms of complications and their rates with the type and extent of the damage they cause. In order to assess the quantity and quality of the ossicular damage and select ossiculoplasty technique accordingly and make prognostic assessments, some classification and scoring systems have been developed. The most well-known and oldest system is the tympanoplasty types classified between type-I and type-V described by Wullstein, one of the founders of modern tympanoplasty (10) and associated ossicular damage classification. In 1972, Austin (11) thought that this classification was not prognostically sufficient and divided ossicular damage into four groups from A to D. In 1994, Kartush (12) revised this classification by adding stapes footplate fixation and incudomalleolar joint fixation parameters and presented the scoring system on which we based this study that numerically expresses ossicular damage.

Table 1. Austin –Kartush scoring

Risk Factor	Risk Value
The status of the ossicles	
M+ I+ S+	0
M+ S+	1
M+ S-	2
M- S+	3
M- S-	4
I/M Başı Fiksasyonu	2
Stapes Fiksasyonu	3

(Minimum score: 0, Maximum score: 7) [M: Malleus, I: incus, S: stapes (-): not existing/determined absent]

Methods

Cases

Patients with the diagnosis or provisional diagnosis of middle ear cholesteatoma and who were operated afterwards or patients on whom cholesteatoma tissue was discovered in the middle ear cavity during tympanoplasty between June 2012 and March 2014 were included in this study. Recurrent patients who were operated (once or multiple times) in another center, the patients whose cholesteatomas were too extensive to differentiate as primary or secondary, because had already ,and the patients whose retraction pockets which had not turned into cholesteatoma were excluded from the study. The first operation records were taken into consideration for the patients who had to be operated because of recurrence. A senior author either operated or supervised the operation.

Existing peripheral facial paralysis and labyrinthitis were preoperatively reported. While writing the patients' operation notes, the status of the ossicles, assessment of the middle ear cavity, boundaries of the cholesteatoma sac, status of the nerves (chorda tympani and facial nerves), (if any) detected complications [lateral semicircular canal (LSCC) erosion such as the mastoid cortex erosion], and other findings were recorded in detail in accordance with a standard template.

Cholesteatomas localizing to the pars flaccida or arising from defects over the posterior malleolar ligaments were grouped as primary and those caused by the pars tensa were grouped as secondary cholesteatomas. The patients' cholesteatoma types (primary or secondary) were preoperatively recorded, and the were confirmed or changed (if required) according to the intraoperative findings.

Evaluation parameters

Based on the Austin–Kartush score (Table 1), the magnitude of ossicle damage and other complications of the disease were retrospectively retrieved from the surgical records. While scoring the magnitude of the ossicular damage, for the malleus, the complete absence of the malleus or its non-functioning state (partial or complete absence of the manubrium and malleus head defect) are considered. According to this system, the minimum score is 0, and the maximum possible score is 7. After scoring, the patients were also grouped based on the magnitude of ossicle damage as "A" (Austin–Kartush score between 0 and 1) for patients without ossicular damage or with only incus damage and as "B" (Austin–Kartush score between 3 and 7 or more extensive and localized to more than one ossicles). As a result of this, we had the following four subgroups: primary-A (PrA), primary-B (PrB), secondary-A (SecA), and secondary-B (SecB). The number of patients divided into the PAC and SAC groups according to their cholesteatoma types; sex and average age of the patients in the PAC and SAC groups; Austin–Kartush scores in the PrA, SecA, PrB, and SecB groups; patients' age at disease onset in the PAC and SAC groups; and complication rates of the PAC and SAC groups are our statistical evaluation parameters to be compared.

Statistical Analysis

In order to evaluate the results obtained in this study, for statistical analysis, SPSS (Statistical Package for the Social Sciences ver. 10.0, SPSS Inc, Chicago, Illinois, USA) was used. Continuous variables were expressed as mean±standard deviation. Categorical variables, on the other hand, were expressed as percentages (%). Among the groups, parametric variables that show congruity with normal distribution were compared with Student's t-test and parametric variables that show incongruity with normal distribution were compared with Mann–Whitney U test. For the comparison of the categorical variables, chi-square and Fisher's exact test were used. In correlation evaluation, Pearson's correlation test was used. For all statistical evaluations, $p<0.05$ was considered significant.

Table 2. Cholesteatoma type comparisons—two patient groups were divided according to their ossicular damage. Group A: Austin–Kartush score is less than 2 (32 patients), Group B: Austin–Kartush score is 2 or more (34 patients)

	Primary	Secondary	p
Group A	9 (28.1%)	23 (71.9%)	0.02 (significant)
Group B	19 (55.9%)	15 (44.1%)	>0.05 (insignificant)

Table 3. Complications observed according to the types of cholesteatomas and rates according to the number of complications

Complications	Primary (%)	Secondary (%)
Subperiosteal Abscess	1 (3.57)	0
Facial Canal Defect	9 (32.14)	6 (15.78)
LSCC Defect	4 (14.28)	0
Other SCC Defects	2 (7.14)	0
Labyrinthitis	1 (3.47)	0
Facial Paralysis	0	1 (2.63)

(LSCC: lateral semicircular canal, SCC: semicircular canal)

Table 4. Cholesteatoma types—Comparison of the number of patients developing complications and the total number of complications

	Primary	Secondary	p
Number of patients developing complications	9	6	0.06
Number of complications	18	6	0.001

Results

Of the 66 patients included in the study, 28 patients were in the PAC group, whereas 38 were in the SAC group. Thirty-six of the patients were female, and 30 were male. Primary cholesteatoma in males and secondary cholesteatoma in females were statistically significantly higher. The average ages for primary and secondary cholesteatomas were 38.4(±17.0) and 42.6(±13.1) years, respectively. The average duration of the disease based on the patients' histories is found as 29 years 3 months (351 months), and no statistically significant correlation was found between the Austin–Kartush scores or the score groups and the reported duration of the disease. On the other hand, it was noteworthy that all five patients under the age of 16 were in the PAC group.

When the cholesteatoma types and Austin–Kartush scores were matched, the average score was 2.25 (±1.35) for the PAC group and 1.76 (±1.65) for the SAC group. This difference was not statistically significant. However, when the subgroups were compared, in the SecA group, the Austin–Kartush score was statistically significantly higher than that of the PrA group [23 patients (71.9%) vs.9 patients (28.1%); p-value=0.002].

In contrast, when the PrB and SecB subgroups that demonstrated a higher level of ossicular destruction (with 3–7Austin–Kartush scores) were compared, a statistically significant difference could not be found [19 patients (55.9%) vs.15 patients (44.1%)] (Table 2).

When the cholesteatoma complications were studied, nine complications in the primary group and six in the secondary group were observed in a total of 15 patients. Except for one patient who had extratemporal complications (subperiosteal abscess), all other complications observed in the rest of the patients were intratemporal (facial paralysis, labyrinthitis, fallopian canal defect, LSCC defect, other semicircular canal defects) (Table 3). When the number of patients who had complications was compared, no statistically significant difference was observed between the PAC and SAC groups. However, when facial canal erosion (dehiscence), the most common complication, was excluded, the number of patients with complications in the PAC group was significantly higher (p=0.001). Some patients had more than one complication, and when the numbers were considered, 18 complications in the PAC group 18 and 6 in the SAC group were observed. This means that a statistically significantly greater number of complications were present in the PAC group (p=0.001) (Table 4).

Discussion

The fact that primary cholesteatoma causes more complications was also confirmed in our study (7, 13). The results of our study show that the SAC group patients were diagnosed at a stage when there was less ossicular damage compared with that in the PAC group patients.

In cholesteatoma, cortical bone and otic capsule erosion and ossicular erosion that cause complications are connected to the direct pressure necrosis of cholesteatoma and enzymatic resorption created by secreted metalloproteinases (14, 15). In their study, Dornelles et al. (16) could not find a correlation between patients' age and perimatrix inflammation intensity or granulation tissue thickness and assessed ossicular damage magnitude. Additionally, In our study, no correlation was found between patients' age and reported duration of the disease and ossicular damage magnitude either.

In both primary and secondary cholesteatomas, the most commonly affected and observed conductive component of the hearing loss or the most common reason causing air–bone gap is incus damage (6, 8, 14). The reason for that is the hanging position of the long arm of the incus and its relatively weaker vascularization which makes it extremely sensitive to pressure effect, including trauma, and it being the most exposed ossicle to cholesteatoma tissue in both types of cholesteatomas (17).

In a study where intraoperative videos of primary (attic) cholesteatomas were retrospectively watched, it was observed that in the majority of cases, "isthmus blockage" is present and that the epitympanum and mezotympanum are largely separated from each other through the tensor tympani plica (18). Thus, cholesteatoma sacs are forced to grow toward the aditus and attic and move downward towards the posterior tympanum along the long arm of the incus. Concordantly, compared with tensa cholesteatomas with wider perforation–retraction, primary cholesteatomas can excrete much less cholesteatoma lamellae and inflammatory products from the pars flaccida defect that can be likened to homologous "narrow bottleneck." This also allows the cholesteatoma to reach outside the mastoid bone boundaries more rapidly and cause ossicular destruction and complications. It was reported that primary attic cholesteatomas cause 2.3 times more complications than SACs (7). In our patients, if the facial canal opening was left aside, in the tympanic segment of 10–11%, dehiscence may be present in ears operated for other reasons (19, 20). In PAC, a statistically significantly higher complication rate was found. The most important finding we obtained in our study is that patients with SAC who have less ossicular damage are diagnosed and can be operated at an earlier stage. The role of the frequent and recurrent attacks that start during childhood in the etiopathogenesis of SACs, which can be controlled by periodic treatments, or chronic otitis media turning into a cholesteatoma in more advanced stages may be the reasons for SACs causing less complications and ossicular damage (8). PAC, on the other hand, follows a more aggressive and destructive course and causes more ossicular destruction and complications.

Conclusion

Primary and secondary cholesteatomas differ from each other in terms of pathogenesis and probably etiology and clinical course. In primary cholesteatomas, more ossicular destruction and complications are observed compared with those of their secondary counterparts, and the most important reason for this situation is that unlike secondary cholesteatomas, primary cholesteatomas cannot sufficiently drain into the anterior tympanum and external ear canal.This factor seems to be the reason for higher prevalence of PACs in pediatric age group in which it also follows a more aggressive and destructive course, and for leading more complications.

Ethics Committee Approval: Ethics committee aproval was not obtained due to the retrospective nature of this study.

Informed Consent: Written informed consent was obtained from patients who participated in this study.
Peer-review: Externally peer-reviewed.

Author Contributions: Concept - N.Y., S.K.; Design - N.Y., S.K.; Supervision - N.Y., S.K.; Materials - N.Y., S.K.; Data Collection and/or Processing - Z.Z.S.; Analysis and/or Interpretation - N.Y., C.K.; Literature Review - Z.Z.S., C.K.; Writing - N.Y.; Critical Review - N.Y., C.K.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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