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Multidimensional staging system for pediatric acquired cholesteatoma: A 30-year verification data☆

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ABSTRACT

Objective: This article presents a points-based prognostic prediction model for pediatric acquired cholesteatoma, incorporating the multidimensional factors that contribute to surgical failure.
Methods: This study included 132 ears with acquired cholesteatoma from 128 children (≤18 years) identified between 1982 and 2012. Each case was scored for the extent of the cholesteatoma, history of grommet insertion, age of the patient, ossicular destruction, and otorrhea. The patients were classified as stage I, II, or III. We compared differences between stages regarding the cumulative rates of recidivism and linear trends in these rates.
Results: Among stage I cases, the rate of cumulative recidivism was 0%; however, among stage 2 cases, this increased with time, eventually leveling off at 15.7% after 18 years of follow-up. The same was observed among stage III cases, which leveled off at 34.1% after 17 years of follow-up. In the second half of the cohort and the entire cohort, differences in the cumulative recidivism curves reached statistical significance, as did the linear trends (all $p < 0.05$).
Conclusions: Our findings demonstrate the efficacy of the proposed multidimensional staging system in linking the severity of cholesteatoma to outcomes, thereby enabling the stratification of patients according to prognosis in order to identify children at risk of recidivism.

1. Introduction

Acquired cholesteatoma is a locally invasive disease, which commonly causes damage to the important structures in the middle ear cleft if left untreated [1]. Staging the severity of cholesteatoma prior to surgery provides a comprehensible reference with which clinicians and patients can discuss the status of the disease and how best to proceed. At present, surgeons must develop their own definitions by which to rate the severity of cholesteatoma. Without a staging system based on commonly accepted standards, these descriptions of cholesteatoma can lead to confusion. A staging system would allow surgeons to compare the surgical outcomes in various studies according to common criteria and provide a useful tool with which to obtain a reliable prognosis.

A panel discussion on the classification and staging of cholesteatoma was held at the 9th International Conference on Cholesteatoma and Ear Surgery in Nagasaki, Japan on June 5, 2012 [2]. Analysis of participant responses revealed fundamental disagreements with regard to the severity and progression of the disease, underscoring the need to establish a consensus regarding the nomenclature used to describe the disease. Panelists at the conference proposed the establishment of a standardized system for the staging of cholesteatoma, similar to the TNM system used to define the stages in cancers. Such a staging system should incorporate relevant clinical features, offer a precise means of assessing the severity and progression of the disease, and possess demonstrable prognostic value with regard to the long-term outcomes of cholesteatoma. The research gaps related to the

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staging system was expected to be an important issue in the next conference in 2016, the 10th International Conference in Cholesteatoma and Ear Surgery in Edinburgh, Scotland, United Kingdom.

A number of multidimensional staging systems have been proposed, including "SOC" (1999) [3], "JOS" (2008) [4], "ATM" (2009) [5], and "TMC" (2012) [6]. At present, the prognostic value of these staging systems has yet to be validated and no single system has gained wide (i.e., international) acceptance. The senior author of the current study, Lien [7], described a multidimensional points-based system for CAO (cholesteatoma, atelectasis, and ossicle) staging [7,8]. The accumulation of experience and clinical data over the intervening 30 years led to the development of the simplified version of this model presented in this study.

Compared with adult-acquired cholesteatomas, the pediatric-acquired variant is more aggressive and more highly proliferative [9,10]. The aggressive behavior of the disease has been shown to contribute to an increase in the rate of recidivism, ranging from 2 to 10 times greater than that found in adults [9,11]. As a result, children are at a higher risk of revision surgery. These clinical differences are the primary factors differentiating pediatric-acquired cholesteatoma from its adult counterpart. This study evaluated the revised version of the CAO staging system using a pediatric data set. Our aim was to assess the prognostic value of the proposed staging system with regard to the long-term outcomes of pediatric-acquired cholesteatoma.

2. Materials and methods

2.1. Study design and patients

Between 1982 and 2012, a total of 154 ears (in children ≤18 years) with acquired cholesteatoma underwent primary surgery at the Taipei Veterans General Hospital in Taiwan. This study excluded cases with previous mastoidectomy for cholesteatoma or non-cholesteatomatous chronic otitis media ($n = 22$). We limited our analysis to the most recent medical records of patients who fulfilled the study criteria. We also collected data related to sex, the side of the head on which the ear was located, age at time of surgery, imaging studies, preoperative ear status, intraoperative findings, surgical technique, recidivistic disease, and history of grommet insertion. This retrospective study was approved by the Institutional Review Board of Taipei Veterans General Hospital.

2.2. Revised CAO staging system (Table 1)

The original CAO staging system involved an assessment regarding the extent of cholesteatoma (C), atelectasis degree (A) and ossicle involvement (O) at the time of surgery [7,8]. In the revised staging model, we included two additional factors of clinical importance: age and otorrhea. The designation "A" in CAO refers to two crucial factors: (1) the "aeration" status of the middle ear cleft, and (2) "age" of the patient. Not having undergone grommet insertion is an indicator of favorable aeration status in the middle ear cleft. This study set a minimum cut-off point of 7 years of age, based on the fact that children above this age have improved Eustachian tube function [12]. The designation "O" in CAO refers to two crucial factors: (1) "ossicular destruction" by the cholesteatoma, and (2) "otorrhea." Each case was scored as follows: C (1–4 points), A (1–2 points) and O (1–2 points) according to the degree of severity. Thus, the combined C, A, and O scores ranged between 5 and 12 points. A score of 5 or 6 represents early stage cholesteatoma (stage I), 7–9 represents a moderate stage of the disease (stage II), and 10–12 represents an advanced stage (stage III). The designation of disease stage in this study involved data from clinical examinations, imaging results, and clinical–surgical correlations.

2.3. Surgical techniques

Most of the patients in the study cohort underwent one-stage canal wall down (CWD) mastoidectomy. In the early period of this study (1982–1988), the primary technique used in our institution was modified radical mastoidectomy (MRM). Starting in the mid-1980s, we gradually revised the technique due to the problems associated with the resulting postoperative cavity with regard to the patients' quality of life. The CWD technique was modified by performing retrograde mastoidectomy in accordance with the extent of the disease. We also sought to eliminate the open cavity through the use of autologous cartilage for reconstruction. We refer to this technique as "tailor-made tympanomastoidectomy with cartilage reconstruction (TTCR)" [13]. Previous studies have reported that the strategy of disease-tracking with cavity reduction is a less invasive option capable of eliminating the problems associated with the open cavity [11,14,15], particularly for children who are unable to undergo regular aural toilet in an otolaryngologic clinic [11,16]. In the late period of this study (1988–2012), most of the patients underwent the CWD-based TTCR technique. In some of the cases, intact canal wall (ICW) mastoidectomy was also applied. The specific techniques used were determined by the surgeons based on the particulars of each case.

2.4. Follow-up protocol

Regular follow-up examinations were performed every 3–6 months throughout the first 5 years, and annually thereafter. Computed tomography (CT) was performed in the patients with suspected cholesteatoma recidivism, as indicated by clinical symptoms and otologic examination (e.g., an unsafe retraction pocket with invisible bottom). Since 2007, diffusion-weighted magnetic resonance imaging has also been used to augment CT

Table 1
Revised CAO staging system of cholesteatomas.

Score	C	A		O	
	Cholesteatoma extent	Aeration	Age	Ossicular destruction	Otorrhea
1	Middle ear	No VTI history	≥7 years	No	No
2	Attic	VTI history	<7 years	Yes	Yes
3	Antrum				
4	Mastoid				

Stage	Sum of score of each variable
I	5–6
II	7–9
III	10–12

VTI, ventilation tube insertion.

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