Background:

Skin does not belong in the middle ear or mastoid cavity. When keratinizing squamous epithelium (the skin that normally grows on the outside of the ear-drum and lines the ear canal) gets into these air spaces, it can form a progressively enlarging and destructive cystic lesion called a cholesteatoma.[1] Cholesteatoma often starts as a pocket of retracted tympanic membrane, pulled into the middle ear space by negative middle ear pressure. Less commonly it is associated with tympanic membrane perforation or repair (known as tympanoplasty or myringoplasty) and is occasionally a congenital lesion. It commonly causes hearing loss and an unpleasantly discharging ear. Rarely, extensive cholesteatoma can extend through the roof of the temporal bone towards the brain, and associated infection may cause intracranial abscesses.[2]

Most cholesteatoma can be successfully treated by surgery, and the extent and effectiveness of surgery is strongly correlated with the size of the cholesteatoma at presentation.[3] The goals of surgery for cholesteatoma in children are: firstly, to completely eradicate the disease, thus preventing complications; secondly, to make the ear dry and manageable; and thirdly, to optimize hearing.[4] Surgical treatment includes repair of the tympanic membrane using similar tympanoplasty techniques to those required for management of more simple tympanic membrane perforation. The management of cholesteatoma in childhood presents particular problems, partly because it is reputed to behave more aggressively, with extensive disease found more frequently in children than adults, with higher rates of residual and recurrent disease after surgery.[5]

There is limited understanding of the etiological factors associated with cholesteatoma and their relationship to the severity of cholesteatoma found at the time of surgery, particularly in the pediatric population.[6] Understanding is confounded by the use of different surgical techniques: it is difficult to determine whether a risk factor or a surgical technique is the larger determinant of outcome.

Purpose:

The purpose of this study is to retrospectively examine the records of children treated for cholesteatoma, tympanic membrane retraction and tympanic membrane perforation in our department. We will explore the existence of any trends or clear associations between pre-existing factors and the severity of cholesteatoma / middle ear disease, as determined by the anatomical extent at the time of surgery, the presence of complicating factors, and the level of hearing impairment. The factors of interest include age, sex, pre-existing medical conditions associated with ear disease (e.g. cleft palate), genetic factors (e.g. Trisomy 21, connexin mutations), site of origin of the cholesteatoma, any history of previous ear surgery or other surgery (e.g. ventilation tubes, tonsillectomy) radiological appearance and outcome (including hearing and recurrence of cholesteatoma) according to type of surgical treatment.

Methods

This study will include around 1000 consecutive children who have been surgically treated for cholesteatoma up to 27 August 2016. The records and scans of these children will be retrospectively reviewed to obtain basic demographic information, and previous medical history which may be etiologically related to subsequent severity of cholesteatoma. Information categorizing the severity of their cholesteatoma at the time of surgery and follow up will also be collected using a validated grading system [7], and we will analyze the data to look for any associations between these characteristics and the severity of cholesteatoma. Details of surgery will also be collected to assess the impact of surgery on outcome in comparison with disease severity. This will include success of tympanic membrane closure with different surgical techniques.

Analysis

Children will be sub-divided into groups based on the severity of their cholesteatoma / retraction / perforation, as determined by the extent, stage of ossicular erosion, presence of complications, and stage. The potential etiological factors will then be compared between the groups, using a combination of non-parametric statistical tests (Fisher's exact test and Mann Whitney U-test), and logistic regression analysis, using SPSS version 14.0; SPSS Inc, Chicago, Illinois), to look for statistically significant differences between the groups.

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