Some Considerations about Middle Ear Cholesteatoma

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Abstract

Although acquired cholesteatoma is widely studied disease, there are still some key points that remain controversial. In this article we intend to highlight important aspects of this intriguing disease that we had recently studied. Clinical classification based on cholesteatoma growth patterns, study the pathogenesis by the analysis of the contralateral ear and hearing impairment associated with cholesteatoma are the main topics that we will discuss.

ABBREVIATIONS

CLE: Contralateral Ear; TM: Tympanic Membrane; PT: Pars Tensa; PF: Pars Flaccida; BC: Bone Conduction; AC: Air Conduction; PTA: Pure Tone Averages; SNHL: Sensorineural Hearing Loss; COM: Chronic Otitis Media; ABG: Air-Bone Gap

INTRODUCTION

Cholesteatoma is the most aggressive spectrum of chronic otitis media. It is a progressive disease that causes hearing loss and recurrent otorrhea. Some patients can also experience clinical complications such as perilymphatic fistulae, facial palsy, and central nervous system infections.

Although a few centuries have passed since its first description by Duverney in 1689 [1], the pathogenesis of acquired middle ear cholesteatoma is still debated. Moreover, its classification and clinical repercussions are also object of several studies.

The objectives of the present paper are to review some important aspects of this intriguing disease.

CLASSIFICATION

Cholesteatoma is characterized by the accumulation of exfoliated keratin debris in the middle ear or other pneumatized areas of the temporal bone [2]. Since cholesteatoma was first described, many classifications have been proposed. In principle, it is accepted that cholesteatomas can be classified into two categories: congenital and acquired [3]. As congenital cholesteatomas are very rare, the existent classifications are applied to the acquired type. The classifications are based on otomicroscopic appearance [4], typical growth patterns [5], disease extension [6], surgical findings [2], and otoscopic drum status [7]. However, there are still controversies about the clinical application of each of those classifications. Moreover, the real prevalence of each form of cholesteatoma is still unknown. Although studies have systematically pointed the attic or posterior epitympanic cholesteatoma as the most frequent [5,8], a more recent study observed a greater prevalence of pars tensa cholesteatoma [7].

In our previous study, we classified the cholesteatomas by analyses of the otoendoscopies and then we described the subtype’s prevalence. We included 414 ears of 356 consecutive patients with middle ear cholesteatoma with no history of surgery. The recorded images were independently reviewed by the same researcher. Cholesteatoma growth pattern was classified following Jackler classification [5]:

- Attic or posterior epitympanic: when the cholesteatoma is confined to the pars flaccida;
- Tensa or posterior mesotympanic: when the cholesteatoma arises in the posterosuperior quadrant of the pars tensa;
- Anterior epitympanic: when the cholesteatoma originates cranially and anteriorly to the malleus head.

Posterior epitympanic (34.3%) and posterior mesotympanic (33.8%) were the most frequent types of cholesteatoma observed. Anterior epitympanic type was the least frequent (2%). However, 30% of the cholesteatomas could not be classified as posterior epitympanic, posterior mesotympanic or anterior epitympanic. We observed that in 13.8% both the pars flaccida and the pars tensa are involved, so we called them two routes cholesteatomas. Finally, in 16.2% no precise growth pattern could be identified by videotoscopy. We classified them as undetermined cholesteatomas.
Posterior epitympanic cholesteatoma was more prevalent in adults (41.9%), whereas posterior mesotympanic cholesteatoma was more frequent in children (43.7%, p < 0.0001). Anterior epitympanic cholesteatoma was only observed in children.

In conclusion, classifying cholesteatomas according to growth pattern into anterior epitympanic, posterior epitympanic, posterior mesotympanic, two routes, and undetermined includes all existing types of middle ear cholesteatoma. In general, the prevalence of posterior epitympanic and posterior mesotympanic cholesteatoma were similar. Whereas anterior epitympanic and the posterior mesotympanic cholesteatomas were more prevalent in children, the posterior epitympanic was more frequent in adults.

Pathogenesis

The pathogenesis of acquired middle ear cholesteatoma is still controversial. At present, the four main theories are as follows: metaplasia (transformation of the inflamed middle ear mucosa into keratinized squamous epithelium); migration (ingrowth of the squamous epithelium through a pre-existing peripheral perforation); invagination (progressive retraction of the tympanic membrane [TM] due to chronic dysfunction of the eustachian tube); and papillary proliferation (infection leading to proliferation of epithelial cones in the basal layers of the pars tensa [PT] or pars flaccida [PF]) [4]. These theories mainly originated from clinical observations and experimental studies [9] since well-designed cohorts are difficult to perform because cholesteatoma is an infrequent disease and needs several years to develop.

Since 2008, we have been indirectly studying the pathogenesis of chronic otitis media by examining the contralateral ear (CLE) [10]. Our observations have systematically showed a high prevalence of alterations in the CLE in clinical [10], histopathological [11], radiological [12] and functional [13] studies. Moreover, our results demonstrated that the frequency of alterations in the CLE was even higher in patients with COM with cholesteatoma [10]. Tympanic membrane retraction and cholesteatoma in the CLE tended to be common in patients with cholesteatoma in the main ear regardless of the growth pattern. Costa et al. stressed the importance of studying the diseased ears in pairs to somehow understand the dynamic pathological process at presentation. Therefore, the maxim “you will be in my shoes tomorrow”, used by those authors to emphasize that the ears should be analyzed as an intrinsically related pair and not as an isolated unit. In doing so, frequently the most affected ear might, somehow, predict the future status of the contralateral side. So, we also believe that nowadays the analysis of the CLE is the best way to study cholesteatoma pathogenesis in humans.

In our previous study, only about one-third of the CLEs in patients with cholesteatoma in the main ear were considered normal. Moderate-to-severe TM retraction and cholesteatoma were undoubtedly the most prevalent pathological changes. Analyzing only those with alterations in CLE we observed that 95.8% of the patients presented retraction or signs of previous retraction (outside-in perforations) or progression of these retractions (cholesteatoma) in the CLE. Interestingly, our results show that there is a strong correlation between growth patterns of cholesteatomas in the main ear and location of TM retractions in the CLE. Therefore it seems plausible to infer that these retractions represent the earlier phases of cholesteatoma formation in the main ear. We still don’t know if the TM retraction per se is enough to cholesteatoma formation. We believe that other factors that can disrupt the stability of the retraction are essential. Sudhoff and Tos [4], after observing the retraction of both the PT and the PF in some children, proposed a four-step concept for the pathogenesis of cholesteatoma that combines the retraction and proliferation theories: (i) the retraction pocket stage; (ii) proliferation of the retraction pocket, subdivided into cone formation and cone fusion; (iii) expansion of cholesteatoma; and (iv) bone resorption. Although bone erosion can occur earlier in the development of TM membrane retraction without...
cholesteatoma formation, the inflammatory process may be involved in both sustenance of negative pressure in the middle ear and progression of TM membrane retraction to cholesteatoma. Jackler et al. on the other hand proposed the theory of mucosal pouch which is based upon the premise that the squamous pouch is drawn inward by the interaction of opposing motile surfaces of middle ear mucosa [14]. The only point of convergence of these theories is that TM retractions were almost universally implied in the first stages of cholesteatoma formation.

Hearing loss

Hearing loss is one of the more disturbing symptoms in patients with cholesteatoma. In our previous study, we found that hypoacusis and otorrhea were the main complaints of most patients, and 87% had otorrhea at the time of evaluation. Complaints of hearing loss were confirmed by audiometry, which showed that the vast majority of patients had conductive hearing loss with larger air-bone gap of 20 dB in speech recognition area [15].

We also demonstrated that, compared with posterior epitympanic cholesteatomas, posterior mesotympanic cholesteatomas had greater AGB thresholds at 500 Hz, 2000 Hz, and ABG PTA, which correspond to the speech reception frequencies. The two growth patterns, however, were very similar with regard to the other audiometric parameters. In children, no audiometric differences were found between the two groups, whereas, in adults, compared with posterior epitympanic cholesteatomas, posterior mesotympanic cholesteatomas showed higher AGB thresholds at 500 Hz to 3000 Hz and higher ABG PTA, in addition to higher AC thresholds at several frequencies [16].

When we studied the sensorineural hearing loss in cholesteatoma, we found that the cholesteatoma ear was associated with greater BC thresholds than the CLE. The presence of cholesteatoma in the middle ear was associated with greater BC thresholds at all frequencies tested, when compared with the normal CLE. These BC differences were observed both in children and adults, and independently of cholesteatoma growth pattern. The frequency of labyrinthine fistula in our sample was low, and thus it may have little influence in the global sensorineural hearing impairment associated with cholesteatoma. The correlation between the air bone gap media in the ear with cholesteatoma and the difference in bone conduction thresholds between both ears was direct and moderate. In other words, when more damaged the tympanossicular system is, greater the sensorineural deficit will be [17].

An understanding of the different types, behavioral patterns, and hearing impairment caused by cholesteatomas will enable us to improve the prognosis of this disease by optimizing its treatment and improving surgical techniques for both complete removal of the pathological tissue and successful reconstruction of the damaged ossicles. We also believe that a better understanding of the sensorineural damage associated with cholesteatoma cannot be overstressed. It is quite obvious that all these changes are time-related, so, in our opinion, it is very important to monitor these patients very closely in order to decide for a surgical intervention during the earlier phases of the disease. In doing so, we may abort the natural history of cholesteatoma and its deleterious consequences. To define with precision the surgical timing for carrying out such an intervention is still a matter of great debate, but it seems plausible that during the early phases surgeries may be less aggressive and residual function is maintained. The preservation of an adequate inner ear function is of paramount importance especially nowadays with the development of new prosthesis and the advent of bone anchored hearing aids and active middle ear implants specifically indicated for patients with COM. This knowledge may assist the development or improvement of new technologies, and may be of even more benefit with regard to BC than our demonstration that SNHL may be present in most patients with cholesteatoma, and that this damage can be clinically relevant in several cases.

DISCUSSION & CONCLUSION

The classification of cholesteatomas according to growth patterns is the most embracing one since is based on pathogenesis and can explain the different aspects in progression and hearing impairment. The contralateral ear is the best method to study the pathogenesis of cholesteatoma in humans. Our studies have shown that the tympanic membrane retractions could be implied in the first stages of cholesteatoma formation in most of our cases. The hearing loss is an important factor to be approached before and during the surgery.

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