Cholesteatoma Induced Labyrinthine Fistula: Is Aggressiveness in Removing Disease Justified?

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Abstract Issues of complete disease clearance and hearing preservation in cholesteatoma induced labyrinthine fistula cases has been discussed and updated in this article. Successful disease clearance and hearing preservation in a case of cholesteatoma induced isolated cochlear promontory fistula encouraged us to retrospectively analyse 13 more cases of cholesteatoma induced labyrinthine fistula who presented in emergency service as complicated chronic suppurative otitis media. Pre-operatively nine patients experienced vertigo, two had profound sensori neural hearing loss and radiology was suggestive of labyrinthine fistula in 12 patients. Lateral semicircular canal was involved in 13 cases. In all cases cholesteatoma matrix was completely removed from the fistula site irrespective of the fistula size and hearing status. Hearing was preserved in 11 out of 12 patients. Gentle and meticulous removal of the matrix and careful repair of labyrinthine fistula delivers significant hearing preservation rate along with a safe and dry ear which avoids a second look surgery.

Keywords Cholesteatoma · Labyrinthitis · Semicircular canals · Hearing preservation

Introduction

Chronic otitis media with cholesteatoma can lead to various complications owing to its bone eroding property. Among the spectrum of intratemporal complications, labyrinthine fistula is one of the common complications encountered in clinical practice [1]. Cholesteatoma induced fistula most commonly involves lateral semicircular canal (LSCC), but can involve other semicircular canals and rarely cochlea. The management of fistula has always been debated in the literature by various authors and is still ongoing. “To leave the matrix over fistula or to remove it completely” is the point of contention. The underlying concern is the preservation of the cochlear function. Conventionally, factors considered for decision making include extent of disease in radiology, preoperative hearing status in both ears and intra-operative fistula size. However, subtle disease induced changes are neither seen radiologically and nor manifest clinically. Such cases surprise the surgeon during intraoperative period and can be challenging. We encountered one such case where an isolated cochlear fistula without any other vestibular involvement amazed us during routine mastoid surgery. The cochlear function could be successfully preserved despite complete matrix removal from the fistula site. This case prompted us to retrospectively analyse 13 more incidences of labyrinthine fistula in complicated squamous otitis media cases. The purpose of study is two-fold, first to discuss management protocol for labyrinthine fistula and secondly to update the recent developments in the management of cholesteatoma induced fistulas.

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Materials and Methods

Index Case

A 10 year old female presented to us with complaints of left ear discharge for seven months with mild hearing loss, without any ringing sensation and giddiness. There was no history suggestive of any intra-temporal, extra-temporal or intracranial complications. Examination of left ear showed atelectatic tympanic membrane with granulations and cholesteatoma over postero-superior and postero-inferior quadrant. The posterior malleolar fold was destroyed and floor as well as posterior wall of bony canal adjacent to annulus was eroded resulting in widening of the bony canal. High resolution computed tomography (HRCT) of the temporal bone showed nondependent soft tissue density in the mesotympanum and epitympanum, partially eroded ossicles and subtle features of osteolysis over promontory as in Fig. 1. Pure tone audiogram showed average air conduction threshold of 35 dB and average bone conduction threshold of 10 dB in frequencies of 500, 1000, 2000 and 3000 Hz. Examination of right ear, nose and throat was normal.

Patient underwent canal-wall down modified radical mastoidectomy by postaural approach. Intra-operatively, cholesteatoma sac was seen in the antrum, aditus, epitympanum, mesotympanum and hypotympanum. The lateral semicircular canal and fallopian canal were intact. Incus was partially eroded and thus removed. Malleus head was sacrificed for disease clearance from anterior epitympanum. In mesotympanum, stapes suprastructure was absent and cholesteatoma matrix was seen over

Fig. 1 High resolution computer tomography images of temporal bone in coronal plane showing nondependent soft tissue density in epitympanum and aditus with widening of annulus and adjacent canal wall. Osteolytic destruction of promontory can be appreciated

promontory, round window and going deep into sinus tympani. On gentle palpation with blunt instrument, fistula was noticed underneath epithelium and it seemed to involve promontory and round window niche region. Radiology was again reviewed intraoperatively and doubtful areas of osteolysis in the region of promontory corresponded with clinical finding of fistula over promontory and round window niche. The cholesteatoma matrix was gently elevated and the cochlear fistula of approximately 4×3 mm size exposing the membranous labyrinth was became apparent as in Fig. 2. Perilymph leak was noticed which was repaired in multiple layers using temporalis muscle and fascia after complete disease clearance. So a final diagnosis of cholesteatoma induced isolated cochlear promontory fistula (Dornhoffer stage IIb) was made [2]. Patient had uneventful post-operative recovery without any giddiness and at follow up after three months patient had dry ear with moderate conductive hearing loss.

Remaining Cases

This index case prompted us to analyse other cases of labyrinthine fistula due to cholesteatoma. We retrospectively reviewed 81 cases of complicated chronic suppurative otitis media (CSOM) operated in emergency between 2011 and mid 2015 in a tertiary care referral centre. In only 63 cases, the details of disease and surgery could be retrieved. Out of these 63 cases, 13 patients had intra-operative labyrinthine fistula. These cases were thoroughly analysed for pre-operative complaint of vertigo, other symptoms and signs, radiological finding of labyrinthine fistula, intra-operative fistula site, size and grading, surgical details as well as post-operative hearing status.

Fig. 2 Intraoperative picture showing fistula over cochlear promontory and round window niche

Table 1 Summary of all patients with cholesteatoma induced labyrinthine fistula

Case no.	Labyrinthine fistula (site, size and Dornhofer grade)	Other associated complications	Pre-operative hearing loss (CHL), mild	Vertigo	Fistula test	HRCT finding of labyrinthine fistula	Surgery	Matrix removal status	Post operative hearing
1	Promontory and round window niche, 4 × 3 mm, IIb	None	Conductive hearing loss (CHL), mild	No	Negative	Subtle osteolysis over promontory	Canal wall down mastoidectomy (CWDM)	Completely removed	CHL (moderately severe)
2	LSCC, PSCC, Utricle, Sacculle, extensive involvement, III	None	Mixed hearing loss (moderately severe)	Yes	Positive	Yes	Radical mastoidectomy (RM)	Completely removed	SNHL (profound)
3	Dome LSCC, 4 × 5 mm, IIa	Meningitis, sub periosteal mastoid abscess	CHL (moderate)	No	Negative	Yes	CWDM	Completely removed	CHL (moderately severe)
4	Dome LSCC, 3 × 2 mm, IIa	None	Mixed HL (moderate)	Yes	Negative	Yes	CWDM	Completely removed	Mixed HL (moderate)
5	Dome LSCC, 2 × 1 mm, IIa	Facial palsy	CHL (mild)	Yes	Negative	Yes	CWDM, facial nerve decompression	Completely removed	CHL (moderate)
6	Dome LSCC, 2 × 2 mm, IIa	None	CHL (mild)	Yes	Negative	Yes	CWDM	Completely removed	CHL (moderately severe)
7	Dome LSCC, 2 × 2 mm, IIa	None	Mixed HL (mild)	Yes	Negative	Yes	CWDM	Completely removed	Mixed HL (mild)
8	Dome LSCC <2 mm, IIa	None	CHL (mild)	Yes	Negative	Yes	CWDM	Completely removed	CHL (mild)
9	Dome LSCC 4 × 2 mm, IIa	None	CHL (Severe)	Yes	Negative	Yes	CWDM	Completely removed	CHL (moderate)
10	Dome LSCC, 3 × 3 mm, IIa	Facial palsy	SNHL (profound)	No	Negative	Yes	CWDM, facial nerve decompression	Completely removed	SNHL (profound)
11	LSCC, <2 mm, IIa	Facial palsy, sub periosteal mastoid abscess	SNHL (profound)	No	Negative	No	RM, facial nerve decompression	Completely removed	SNHL (profound)
12	Dome LSCC, <2 mm, IIa	Facial palsy	CHL (moderate)	No	Negative	No	CWDM, facial nerve decompression	Completely removed	CHL (moderately severe)
13	Dome LSCC, 4 × 5 mm, IIa	None	CHL (mild)	Yes	Negative	Yes	CWDM	Completely removed	CHL (moderate)
14	Dome LSCC, <2 mm, IIa	None	Mixed HL (mild)	Yes	Negative	Yes	CWDM	Completely removed	Mixed HL (mild)

Results

Including the index case the important relevant details of all the 14 cases are shown in Table 1. 35.7% ($n = 5/14$) cases had other complications other than labyrinthine fistula. Pre-operatively 57% ($n = 8/14$) patients had normal bone conduction (BC), 28.5% ($n = 4/14$) patients had preserved BC and 14% ($n = 2/14$) had profound sensori neural hearing loss (SNHL). 64% ($n = 9/14$) patients had giddiness at presentation. Fistula test was positive in only one (7%) patient. All patients had pre-operative HRCT of temporal bone. In 86% ($n = 12/14$) patients labyrinthine fistula could be appreciated in HRCT. Canal wall down (CWD) mastoidectomy was done in 86% ($n = 12/14$) of patients and Radical mastoidectomy in two patients. Out of these two patients, one had very extensive cholesteatoma involving LSCC, Posterior SCC, utricle and saccule, though he had mixed HL. The other patient had preoperative profound SNHL. Cholesteatoma matrix was removed completely in all the patients. LSCC was the most common site (93%, $n = 13/14$), cochlear promontory was involved in one (7%, $n = 1/14$) and PSSC along with vestibule was additionally involved in one case. As per Dornhoffer staging, 86% ($n = 12/14$) had stage IIa fistula, 7% ($n = 1/14$) had stage IIb fistula and 7% ($n = 1/14$) had stage III fistula. Post operatively the bone conduction was same as pre-operative value in 92% ($n = 11/12$) cases.

Discussion

Labyrinthine fistulas constitute around 4–12% of complications due to cholesteatoma. In a large meta-analysis, the affected site of fistula was LSCC in 87% of cases, promontory in 8% cases, SSCC in 6% cases, and PSSC in 2% of cases [3]. Our data is slightly different from this data which may be attributed to our small number of cases. Cochlear fistula which is quite rare, is mostly associated with concomitant fistula in other areas of labyrinth [4]. In the index case, we encountered a rare clinical scenario, in which cholesteatoma induced isolated cochlear fistula was noted and was managed successfully in terms of complete disease clearance and hearing preservation.

The paediatric patient in the index case had a short history of ear discharge and the disease was apparently limited on clinical evaluation. However intraoperatively, disease induced promontory fistula was identified, reflecting the aggressiveness of cholesteatoma in paediatric patients. This aggressiveness is attributed to cellular mastoid, higher proliferation rate of the keratinocytes and higher collagenase activity of the cholesteatoma matrix in children [5].

In general, among patients with labyrinthine fistula, history of vertigo is present in 64% of cases and fistula test may be positive in up to 50% patients. Profound SNHL is noted in 15% patients [3] and radiological finding suggestive of fistula would be seen in up to 97% of cases [6]. Our findings are quite similar to that described in the literature. The rate of radiological finding of labyrinthine fistula was lower in our series as some of the HRCT which the patients brought along at presentation were not of good quality.

In the index case as well as four other cases (35.7%, $n = 5$), there were no clinical signs and symptoms of vestibular involvement, despite the disease eroding bony labyrinth. This could be explained by intactness of the membranous labyrinth and shielding of the inner ear fluid from middle ear pressure or sound waves by cholesteatoma matrix itself. Most of such patients would have normal hearing, unless there is migration of inflammatory mediators or toxins into inner ear fluid. Another mechanism described in literature for the absence of vestibular symptoms in fistula cases is the presence of utriculo-endolymphatic valve (membrane limitans) and the connective tissue in the perilymphatic system which provides relative independence to the labyrinthine fluid of cochlea from that of the posterior labyrinth [7]. Nevertheless, the preoperative radiology has good sensitivity to detect labyrinthine fistula even in asymptomatic patients as in this case, and thus needs thorough review by operating surgeon before proceeding with surgical management.

The surgical management of cochlear fistula (as in the index case) is no different from that of semicircular canal fistula and the surgical principles are essentially the same. The controversies in surgical management of cholesteatoma induced fistula mostly revolve around removal of the matrix overlying the fistula and lowering of the posterior canal wall. Over the decades, the concepts are evolving and recently there has been a shift of consensus towards complete removal of cholesteatoma matrix from the fistula, as shown in Table 2. Studies have shown that risk of labyrinthine damage due removal of epithelium lining the fistula is very less compared to the long term risk of labyrinthitis due to residual epithelium over fistula which continues to produce collagenase [3]. Chang Hyun in his review of 23 cases with fistula, showed that not only did the fistula size, but also location of fistula, number of fistula and even preserving posterior canal wall had no statistically significant bearing on hearing preservation rate [8]. Moreover, complete removal of disease enables good and adequate grafting of the fistula so as to protect the labyrinth from caloric effect after lowering the canal wall, in addition to providing a dry ear to the patient. In our study 92% of cases had preserved or improved hearing post-operatively which

Table 2 Different authors' work on labyrinthine fistula and their results

References	Cases	Operative details	Results
Ostri et al. [11]	20 cases	Removal of the cholesteatoma matrix, covering of the fistula with fascia and bone in all cases	76% of the patients achieved a hearing improvement and one had anacusis
Vanden Abeele [12]	18 cases	Total removal of the epithelium and two-stage closed technique was followed in all	Hearing preserved or improved in all cases except one in which labyrinthectomy was needed due to extensive disease
Portier et al. [13]	22 cases	Matrix removed in 91% of cases, whereas it was left in the course of a canal down procedure in 9%	With a follow-up of 5.7 years, hearing remained unchanged in 80% of patients
Chen et al. [14]	22 cases	All patients were treated by completely removing the cholesteatoma matrix followed by semicircular canal occlusion	No surgery-related hearing detriment and four of them demonstrated hearing improvement
Meyer et al. [15]	42 cases	Matrix was completely removed from the fistula and immediately covered by autogenous material in all patients	Hearing was preserved or improved in 76% of the patients

is comparable to that of most of the current studies where cholesteatoma matrix was completely removed.

Presently, the consensus on the surgical management of labyrinthine fistula is complete removal of epithelium from fistula site (irrespective of size and location) and grafting thoroughly with soft tissue. The aggressiveness in removing “complete epithelium over fistula” is justified in terms of giving a dry and safe ear, preventing long term complications and avoiding revision surgery. However, caution is indispensable regarding intraoperative gentle manoeuvring and meticulous disease removal. Avoidance of direct suctioning on the fistula, prevention of entry of blood/irritants into labyrinth and intraoperative use of corticosteroid has shown to preserve cochlear function and reduce incidence of post-operative vertigo [9, 10].

Regarding the decision on preservation of posterior canal wall in fistula cases, factors to be considered include hearing status in ipsilateral and contralateral ears, presence/absence of other complications, patient's vocational activities and availability of patient for follow up along with surgeon's preferences and expertise [8]. Although, canal wall is lowered in majority of the cases, one can also preserve canal wall in selected cases with limited disease, preserved hearing, absence of other complications and good patient compliance, where second look surgery is to be planned after 6–12 months. We lower the canal wall in all extensive and/or complicated cases of cholesteatoma as the patient compliance is not good in our scenario.

Conclusion

Isolated cochlear fistula due to cholesteatoma is a rare clinical scenario. No similar case has been reported in the English literature to the best of our knowledge. Radiology is a sensitive diagnostic tool for cholesteatoma induced labyrinthine fistula even in patients with no vestibular symptoms. The present recommendation in management of

labyrinthine fistula, backed by the literature is to perform complete removal of epithelium meticulously with grafting of fistulous site. Such an approach has shown to improve the post-operative outcome in terms of disease free ear along with preserved cochlear reserve and prevent long term complications.

Compliance with Ethical Standards

Conflict of interest None.

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