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Chronic otitis media with cholesteatoma: clinical presentation and surgical management

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ABSTRACT

Background: Cholesteatoma has long been a formidable adversary to the otologic surgeon. There is no single surgical treatment of choice for aural cholesteatoma. The extent of cholesteatoma, the amount of preoperative destruction, and mastoid pneumatization, guides the surgeon in choosing the type of operation for a particular ear. Intra-operative findings guide the surgeon to perform the required surgery despite thorough pre-operative work-up.
Methods: Prospective study on 42 patients who underwent surgery for cholesteatoma.
Results: Cholesteatoma most commonly affected males aged between 10-20 years. Also, the disease was noted to be more aggressive in the paediatric age group. Foul smelling ear discharge was the most common presenting symptom followed by hearing loss. 21% of patients presented with complications. Canal wall down mastoidectomy was done in patients with complications and in cases where adequate disease clearance could not be accomplished with the posterior canal wall intact. All patients in the study underwent ossicular reconstruction. This was owing to either the destruction of ossicles by the disease process or to achieve adequate disease clearance.
Conclusions: Disease eradication is still the main aim of the surgery, but, when it can be achieved keeping the posterior canal wall intact, the canal wall should always be left intact. With advances in technology and availability of angled endoscopes this should be given more consideration. However, in a patient with complications and patients who are not available for regular follow-up the canal wall down surgery is preferred.

Keywords: Cholesteatoma, ICW, CWD, Cholesteatoma complications, Residivism

INTRODUCTION

What differentiates a human being from other species is the ability to express ideas and concepts by means of language, the acquisition of which is dependent on normal hearing. Hearing is arguably one of the most important special senses for man.

Cholesteatoma is an erosive process causing destruction of the middle ear structures, leading to hearing impairment in a large majority of patients affected. The expansion of cholesteatoma may result in bone erosion of the ossicles, otic capsule, fallopian canal, tegmen tympani, and tegmen mastoideum.[1] The involvement of these structures that are in close proximity to the middle ear can cause irreversible sequelae and life-threatening complications.

Cholesteatoma are cyst-like, expansile lesions of the temporal bone lined by stratified squamous epithelium that contain desquamated keratin. They most frequently involve the middle ear and mastoid, but they may develop anywhere within the pneumatized portions of the temporal bone.[2]

Cruveilhier first described aural cholesteatoma as a "pearly tumor" of the temporal bone.[1] Cholesteatoma is a misnomer originally coined by German physiologist

Johannes Mueller in 1838 when he described a layered pearly tumor of fat, which was distinguished from other fat tumors by the biliary fat or cholesterin that is interspersed among the sheets of polyhedral cells.[2] Cholesteatomas do not contain fat, and they do not usually contain cholesterin. Nevertheless, the term remains, despite a more appropriate term suggested by Schuknecht: keratoma.[2]

Abramson et al has given a thorough definition of cholesteatoma "cholesteatoma is a three dimensional epidermal and connective tissue structure, usually in the form of a sac and frequently conforming to the architecture of the various spaces of the middle ear, attic and mastoid.[3] This structure has a capacity for progressive and independent growth at the expense of underlying bone & has a tendency to recur after removal"

The first scholarly treatise on mastoid surgery for suppurative disease was by Schwartze in 1873.[4] The procedure he described was a cortical mastoidectomy with limited exenteration of mastoid air cells. For acute and coalescent mastoiditis, which were prevalent in the preantibiotic era, this procedure proved remarkably efficacious. As one might expect, however, the simple mastoidectomy rarely cured chronic otitis media with cholesteatoma.

During the next 20 years, it became evident that creating an open cavity was necessary for these diseases, and in 1890, Zaufal described removing the superior and posterior canal wall, tympanic membrane, and lateral ossicular chain, a procedure now known as the radical mastoidectomy.[4] This procedure was modified by Bondy, who recognized that disease limited to the pars flaccida could simply be exteriorized, leaving the uninvolved middle ear alone.[4] His description of the "modified radical mastoidectomy" or "Bondy procedure" in 1910 represented one of the first reports addressing hearing function

Interest in hearing preservation and restoration gained further attention after Lempert introduced the fenestration operation in 1938, and Zollner and Wullstein described tympanoplasty techniques in the early 1950s.[4]

During the next decade (1960s), Jansen, Sheehy, and others extended these principles of restoring function and maintaining normal anatomy with the introduction of the intact canal wall mastoidectomy with facial recess approach.[4]

Several authors have described various techniques for eradication of cholesteatoma with hearing preservation ever since.[5,6]

Cholesteatoma whether congenital or acquired, can only be eradicated from the temporal bone by surgical resection. The goals of surgical management include the eradication of disease, restoration of hearing, and to the extent possible, maintenance or restoration of normal anatomic configuration, thus ensuring good quality of life.

Cholesteatoma has long been a formidable adversary to the otologic surgeon. There is no single surgical treatment of choice for aural cholesteatoma. The extent of cholesteatoma, the amount of preoperative destruction, mastoid pneumatization guides the surgeon in choosing the type of operation for a particular ear. Intra-operative findings guide the surgeon to perform the required surgery despite thorough pre-operative work-up.

The study highlights the intraoperative findings in cholesteatoma and how these guide in determining the required surgery.

METHODS

The patients attending the department of ENT and also patients referred from other departments at Vijayanagar Institute of Medical Sciences, Bellary formed the subjects for our study.

Inclusion criteria

Inclusion criteria were all patients of chronic otitis media with cholesteatoma; patients with recurrent or residual cholesteatoma (previously operated cases).

Exclusion criteria

Exclusion criteria were all patients with chronic otitis media– safe type (without cholesteatoma); patients with minimal retraction pockets, Tympanosclerosis, Adhesive otitis media

Method of collection of data

A written informed consent was taken from all patients included in the study. Detailed history-taking, thorough clinical examination was done for these patients. The data collected was entered into a specially designed case record form.

Duration of study

The study was conducted from December 2010 to March 2013.

Sample size

This study comprised of 42 patients who were admitted and who underwent surgical management in the Department of ENT, VIMS, Bellary.

Preoperative tests and evaluation

All the 42 patients who underwent surgical management for cholesteatoma in this study were, in their own

language, explained in detail about the nature of the disease, the anaesthetic procedure, the operative procedure and possible complications and consent was obtained from them.

In all the patients' thorough examination, routine investigations, relevant audiological and radiological tests were performed. Relevant investigations among the following were performed:

- Haematological– complete blood count
- Biochemical– blood sugar, blood urea, serum creatinine
- Pure tone audiometry
- Radiological- X-ray mastoids, HRCT Temporal bones

Data analysis

The data was analyzed using the IBM SPSS v24, with the data being represented as frequency and percentages and the significance determined using the Chi square test.

RESULTS

The study comprised 42 patients who underwent surgical management for cholesteatoma.

Table 1: Ear discharge characteristics.

Ear discharge	Frequency (N=39)	Percentage (%)
Type		
Continuous	15	38.4
Intermittent	24	61.6
Quantity		
Scanty	23	58.9
Profuse	16	41.1
Odour		
Foul smelling	39	100
Non foul smelling	00	00
Quality		
Mucoid	00	00
Mucopurulent	21	53.8
Purulent	15	38.4
Blood stained	03	07.8
Duration		
0–2 years	19	48.7
2–4 years	04	10.2
>4 years	16	41.1

Age and sex incidence

In our study 70% of the patients were aged less than 20years. Though cholesteatoma can present at any age, its occurrence reduces after the age of 20 years according to our study. Males aged between 10-20 years were most commonly affected. Also, the disease was noted to be more aggressive in the paediatric age group.

Table 2: Otoscopy vs Otomicroscopy findings in cholesteatoma.

Site	Otoscopy		Otomicroscopy	
	Frequency	%	Frequency	%
Pars flaccida	15	45.5	19	51.4
Pars tensa-marginal	11	33.3	11	29.7
Pars tensa-central	06	18.2	06	16.2
Both	01	3	01	2.7
Total	33	100	42	100

Presenting symptoms

Bilateral disease was noted in 26% of the study population. Literature suggests that 10% of cholesteatomas are bilateral.[7] Our study had a higher percentage of patients presenting with bilateral disease (26%) Otorrhea was the most common presenting symptom in our study noted in 93% of patients, followed by hard of hearing seen in 90% of the study population. Earache was seen in 36% and 10% of patients presented with post-aural swelling.

Table 3: Ossicular status in cholesteatoma patients.

Ossicles	Frequency	Percentage (%)
M-I-S+	06	14.3
M-I-Ss-Sf+	11	26.2
M+I-S+	05	11.9
M+Il-S+	07	16.7
M+Il-Ss-Sf+	01	02.4
Mh-I-Ss-Sf+	02	04.8
Mh-Il-S+	03	07.1
M+I-Ss-Sf+	06	14.3
M+I+S+	01	02.4
Total	42	100

M-Malleus, I-Incus, S-Stapes, Ss-Stapes suprastructure, Sf-Stapes footplate, Il-Incus long process, Mh-Malleus handle.

The ear discharge was more frequently found to be foul smelling (100%), intermittent (62%), scanty (59%), and mucopurulent (54%) in character. It was occasionally blood stained (8%).

29% of the study population gave history of undergoing ear surgery in the past. Of these 6 patients (14%) had undergone ear surgery in the past and presented with recurrent disease, 3 patients had undergone ear surgery on the opposite ear, 3 patients had undergone incision and drainage of post-aural abscess. One of the patients who had undergone I and D was referred from neurosurgery following drainage of temporal lobe abscess.

Table 4: Ossiculoplasty material used for hearing reconstruction.

Ossiculoplasty material	Frequency	Percentage (%)
Autologous ossicles	11	26.2
Homologous ossicles	04	09.6
Synthetic	14	33.4
Homologous septal cartilage	12	28.6
Autologous conchal cartilage	01	02.4
Total	42	100

21% of patients presented with complications. 14% patients presented with extracranial complications and 12% of patients had intracranial complications with 5% presenting with both intra-temporal and intracranial complications. The most common extracranial complication was abscess noted in 12% patients, 4 patients presenting with post-aural subperiosteal abscess and 1 patient with Bezold's abscess. Facial palsy and labyrinthitis was noted in 1 patient (2.4%) each.

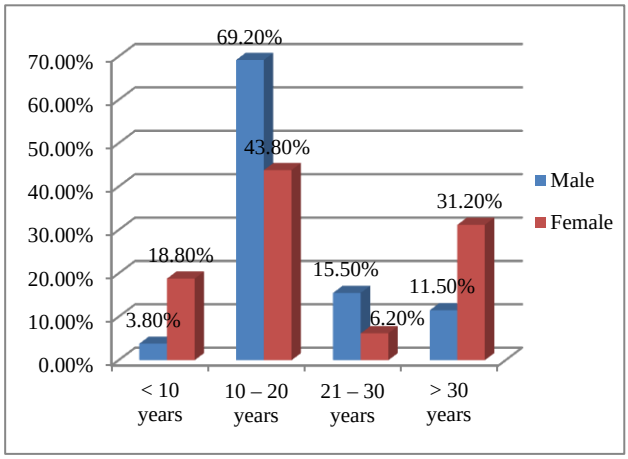


Figure 1: Age sex wise distribution of study subjects.

Figure 2: Patient with left post-auricular sub-periosteal abscess.

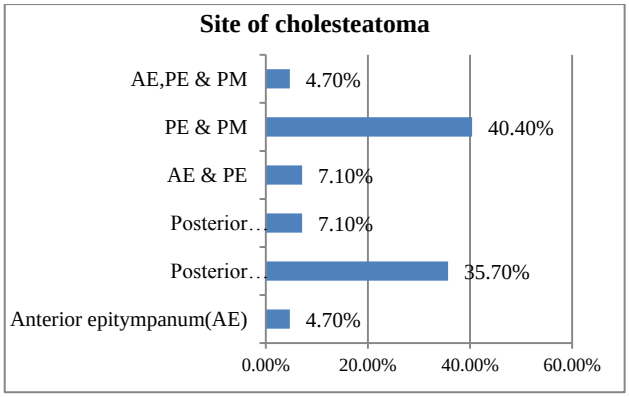
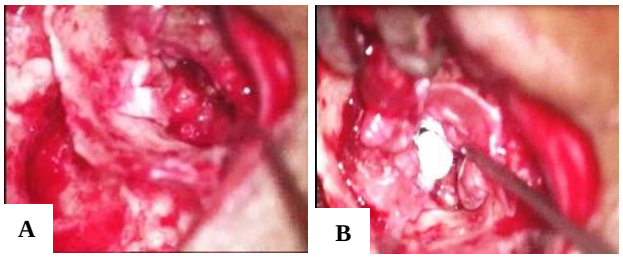


Figure 3: Site of cholesteatoma distribution.



Figures 4 (A and B): Right ICW mastoidectomy with lateral attic wall reconstruction using autologous conchal cartilage and MS with PORP.

The most common intracranial complication was lateral sinus thrombophlebitis seen in 7% of patients. Meningitis and temporal lobe abscess were seen in 2.4% each of the study population at presentation. 78% of the patients who presented with complications were aged ≤19 years. 80% of the patients who presented with intracranial complications were aged ≤13 years. This clearly points to the aggressive nature of paediatric cholesteatoma.

Figure 5: Left CWD mastoidectomy with MP using TORP.

Otoscopic and otomicroscopic findings

The most common pathology noted in the tympanic membrane on otomicroscopy was perforation. Pars

flaccida was the most common site of perforation as noted in 51% of the study population followed by marginal perforations involving the postero-superior portion of the pars tensa in 30%. Central perforation was noted in 16% of the study population. In 3% both the pars tensa and flaccida were involved. Retraction of the tympanic membrane was noted in 12% and both perforation and retraction were noted in 24%. The perforation in the pars tensa was graded as large when it involved more than one quadrant and perforation in the pars flaccida was graded large when it involved more than 50% of the pars flaccida. 76% of the study population had large perforation and 24% had small perforation.

12% of patients having tympanic membrane perforations were falsely identified as having tympanic membrane retractions on otoscopy.

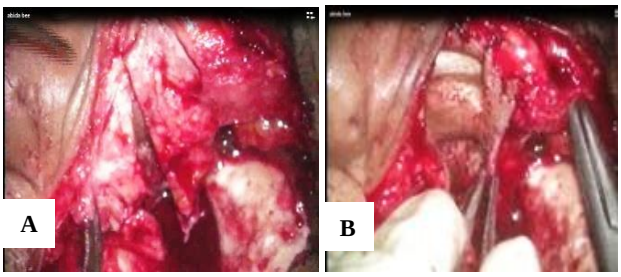


Figure 6 (A and B): Left ear meatoplasty done in CWD mastoidectomy.

Audiological evaluation

Preoperative pure tone audiometry was performed in all patients and pure tone average for air conduction and bone conduction was calculated for 0.5, 1, 2 kHz and the air-bone gap were determined.

Audiological evaluation of patients revealed 83% of the study population to have conductive hearing loss and 17% patients had mixed hearing loss. The pre-operative pure tone average for air conduction of the study population was 50 dB.

Radiological evaluation

All patients in the study population had x-ray mastoids Schuller's view done. Only 19% of the study population underwent high resolution computed tomography (HRCT) scanning of the temporal bones. This was largely limited to patients who presented with complications. In all cases with suspected intracranial complications Computed Tomography scan of the brain was also performed. The most consistent finding in HRCT temporal bones was the presence of soft tissue opacity in the middle ear and mastoids. Ossicles were noted to be eroded in all these scans. The second most common structure found to be eroded was the scutum. 50% of these scans showed the pattern of pneumatization of the temporal bones to be diploic, 37% showed a pneumatized pattern and 12% showed a sclerotic pattern of pneumatization.

Intraoperative findings

The most common ossicle eroded was the incus (98%) of which only the long process of incus was eroded in 26%. In 52% the malleus was noted to be eroded of which 12% patients had erosion of the malleus handle only. 48% of the patients had erosion of the stapes suprastructure.

The cholesteatoma was found to be involving the posterior epitympanum (PE) (88%) most commonly followed by the posterior mesotympanum (PM) (52%) and the anterior epitympanum (AE) (17%). The posterior epitympanum and posterior mesotympanum were together involved in 40% patients.

The cholesteatoma was associated with granulation tissue in most cases, together causing significant ossicular erosion. In only one patient there was limited cholesteatoma involving only the anterior epitympanum, and this patient had all ossicles intact, but required ossicular reconstruction in order to accomplish complete disease clearance. Cholesteatoma was considered limited when it was noted in the middle ear, aditus ad antrum and was considered extensive when it was noted to be infiltrating into the air cells. 52% patients had limited cholesteatoma whereas 45% had extensive cholesteatoma infiltrating into the air cells, making the disease clearance challenging.

Frequently the cholesteatoma was associated with the presence of granulation tissue which by itself could be implicated as the cause for destruction of the middle ear structures.

In the patient with facial nerve palsy House Brackmann grade III, cholesteatoma was noted around the second genu and adjoining horizontal and vertical portion of the facial nerve. Also, in this patient there were defect noted in the tegmen mastoideum and the sinus plate, though the patient did not have any intracranial complications.

In 3 patients who presented with lateral sinus thrombophlebitis, erosion of the sinus plate with granulation tissue adjoining the sigmoid sinus was noted. In one of these patients there was frank pus oozing from this region.

In 2 patients who presented with intracranial complications of meningitis and brain abscess large defects in the tegmen mastoidea were noted.

In one patient who presented with post-aural abscess, incidentally limited erosion of the lateral semicircular canal was noted intraoperatively but patient had no features suggestive of labyrinthitis.

Surgical management

All patients in the study population underwent mastoidectomy with tympanoplasty at a single stage via the post-aural approach. 36 (86%) patients underwent primary surgery for cholesteatoma. Of these, 24 (67%) underwent intact canal wall (ICW) mastoidectomy and 12 (33%) patients underwent canal wall down (CWD) mastoidectomy.

Canal wall down mastoidectomy was done in patients with complications and in cases where adequate disease clearance could not be accomplished with the posterior canal wall intact.

14% of patients had revision surgery. Of these 6 patients, 5 had undergone canal wall down mastoidectomy in the past for cholesteatoma and presented with recurrence and 1 patient had undergone cortical mastoidectomy for cholesteatoma. In the patient who underwent cortical mastoidectomy in the past there was significant presence of cholesteatoma noted at surgery. In the 5 patients who underwent canal wall down procedure in the past 4 had cholesteatoma and in 1 patient there was no cholesteatoma noted. 3 patients had limited cholesteatoma and 1 had extensive cholesteatoma among the 4 patients who underwent revision canal wall down mastoidectomy. These patients had irregular cavities with inadequate meatoplasty causing symptoms.

All patients in the study underwent ossicular reconstruction in the form of myringoplatinopexy (MP) or myringostapedopexy (MS). This was owing to either the destruction of ossicles by the disease process or to achieve adequate disease clearance. Myringoplatinopexy was done in 22 (52%) patients, 20 of these patients had destruction of the stapes suprastructure and in 2 patients myringoplatinopexy was performed to accomplish adequate disease removal. 20 (48%) of the patients in the study population underwent myringostapedopexy.

The most common material used for ossicular reconstruction was synthetic materials which were used for reconstruction in 33%. Total ossicular reconstruction prosthesis was used for MP and partial ossicular reconstruction prosthesis was used for MS followed by homologous septal cartilage (29%), which was used for reconstruction in both MS and MP. Autologous ossicles were used for reconstruction in 26% of patients and homologous ossicles were used in 10%. Malleus was used for reconstruction in myringoplatinopexy and incus was used for reconstruction in myringostapedopexy.

Temporalis fascia graft was used for reconstruction of the tympanic membrane defect by underlay technique.

All patients underwent satisfactory removal of the disease (mastoidectomy) and hearing reconstruction (tympanoplasty) in a single stage surgery.

DISCUSSION

In a CWD mastoidectomy, the bony tympanic annulus and much of the ear canal is removed, and the tympanic membrane graft is placed onto the facial ridge and medial attic wall. Thus, significantly reducing the size of the residual middle ear space. However, as long as this air space is ≥0.5 cc, the resultant loss of sound transmission should be less than 10dB.[8] Since the average volume of the tympanic cavity is 0.5–1cc, a CWD procedure should not cause any acoustic detriment if the middle ear is well ventilated.[9] Indeed, clinical studies comparing the acoustic results of CWD vs. ICW mastoidectomy have shown no significant difference.[10,11]

A CWD procedure also results in the creation of a large air space lateral to the ear drum, i.e., the air space within the mastoid bowl including the EAC.[8] This mastoid bowl and ear canal space generates resonances which can influence middle-ear sound transmission favourably or unfavorably.[12] The structure-function relationships between the size and shape of the mastoid cavity, and cavity resonances have not been well defined. An improved understanding of this issue may help oto-surgeons to configure mastoid cavities in ways that are acoustically beneficial.[8]

Following an initial surgery, regardless of whether an ICW or CWD technique is used, both residual and recurrent disease- recidivism are possible. Residual disease is defined as persistence of cholesteatoma following an incomplete removal. Recurrent cholesteatoma is a newly developed cholesteatoma secondary to a retraction pocket into the mesotympanum or the epitympanum/ antrum. Canal wall defects, created by the original disease process or by the surgeon during removal of the initial cholesteatoma, predispose to retraction pockets. Recurrent cholesteatoma is primarily a concern with ICW mastoid surgery. To minimize these problems adequate surgical expertise is required. All our patients who underwent ICW mastoidectomy had reconstruction of the lateral attic defect with autologous cartilage and soft tissues as required. ICW and CWD mastoidectomy each have inherent limitations and advantages that involve ease of disease removal, incidence of recurrent/residual disease, and the extent of postoperative care. For cholesteatomas involving the middle ear, epitympanum, and mastoid, the canal wall partially obscures the disease. Maintaining the canal wall, therefore, necessitates more surgical manipulations (e.g., facial recess approach and alternately working on the external canal and epitympanic sides of the canal wall) and the requirement of oto-endoscopy and usually more operative time, especially for removing disease in the epitympanum and around the stapes. Cholesteatoma that has extended into the sinus tympani and adjacent recesses may be difficult to remove, regardless of whether the canal wall is left intact or removed.

Because disease exposure is more difficult when the canal wall is left intact, the incidence of inadvertently leaving a focus of squamous epithelium is increased. Maintenance of the canal wall also predisposes to the possibility of retraction pocket formation. Thus, both residual and recurrent cholesteatoma are more common following ICW versus CWD mastoidectomy. As a result, the decision to maintain the canal wall may necessitate more surgical procedures than a CWD technique in order to ensure disease eradication.

Detection of residual disease after surgery is easier with a CWD mastoidectomy. When the canal wall has been removed, only disease in the mesotympanum, hidden by the tympanic membrane, may be difficult to see. An exception to this condition would be cases in which tissue flaps and/or bone pate have been used to partially obliterate the mastoid cavity. With an ICW, the mastoid and epitympanum are not accessible to postoperative inspection. Planned second-stage surgery may mitigate postoperative concern in ICW mastoid surgery.

The principal advantages of the ICW technique are the more rapid healing and, most importantly, the obviation of many long-term postoperative concerns. Epithelialization of the mastoid cavity in CWD cases may be a slow process (taking months) and certain areas may require special attention to promote healing. In our study most patients (90%) had satisfactory healing of the mastoid cavity at 6 months follow-up but required special attention and extra care. Once healing is complete, the mastoid bowl may require periodic cleaning because of irregular contours and the inability of desquamated epithelium to migrate effectively to the meatus. This condition predisposes to mastoid bowl infection (especially if water exposure has occurred). By contrast, healing of the ICW mastoidectomy is usually rapid, periodic ear cleanings are not necessary, the incidence of external ear infections is not increased, and no limitations of water activities are imposed. In addition, an ICW approach provides more options for a hearing aid (e.g., canal type), if required, and its use is usually more trouble free (e.g., involving fewer canal infections).

CONCLUSION

Disease eradication is still the main aim of the surgery, but, when it can be achieved keeping the posterior canal wall intact, the canal wall should always be left intact. With the technical advances and availability of angled scopes which have marched into the oto-surgeons kitty the aim of a cholesteatoma surgery should be to provide the patient a good quality of life post-surgery, and this is best achieved by leaving the posterior canal wall intact. However, in a patient with complications and patients who are not available for regular follow-up the canal wall down surgery may be preferred. Patients presenting with central perforation may have underlying cholesteatoma as seen in the study. Also, patients with cholesteatoma will require ossicular reconstruction owing to either destroyed ossicles due to disease process or owing to obtaining complete clearance of the disease process, and, patients need to be counselled regarding the hearing outcomes.

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