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Original Research Article

Management of advanced cholesteatoma: Madras ENT Research Foundation experience

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ABSTRACT

Background: Cholesteatoma is a common condition encountered by the otolaryngologist in the Indian subcontinent. Due to absence of pain in most patients, they may have advanced disease at presentation. Lack of awareness, long distance between the patient's home and the treatment centre are also reasons for late presentation. Surgical management is the mainstay of treatment. The aim of this study was to retrospectively analyze the number of patients with advanced cholesteatoma, the extent of disease, and associated complications.
Methods: Fifty one patients out of a total of 1132 patients with cholesteatoma presented with advanced disease in a tertiary referral centre between January 2010 to January 2020. The surgical issues in the management of the disease and the outcomes were studied.
Results: Fifty one patients presented with extensive cholesteatoma in a tertiary referral centre. There were 33 males and 18 females in the study. Of fifty one patients, pediatric cholesteatoma was seen in 6 patients. Hearing loss was the most common presentation. Facial palsy, labyrinthine fistula, dural involvement, internal auditory meatus involvement was noted in some patients. All patients underwent surgical management with good outcomes.
Conclusions: Advanced cholesteatoma is a dreaded disease that can result in various complications. Involvement of the facial canal, labyrinth, cochlea, dura, internal auditory meatus, internal carotid artery, jugular bulb, and sigmoid sinus involvement in the presence of extensive disease should be identified before surgery. Appropriate treatment with complete disease clearance can result in good outcomes.

Keywords: Advanced cholesteatoma, Imaging, Complications

INTRODUCTION

Cholesteatomas very often exist in a non-aggressive state, remaining undetected for years before potentially dangerous manifestations occur.[1] The goal of cholesteatoma surgery is the complete removal of squamous epithelium to minimize the risk of recurrence. Bone resorption is a mechanism that may explain the increase in osteolysis associated with cholesteatoma.[1] A canal wall down procedure is usually performed in presence of extensive disease. A thorough pre-operative evaluation is essential for appropriate management of advanced cholesteatoma. Cholesteatoma matrix must be removed completely to avoid recurrence.[2] Long term follow-up is mandatory. Pediatric cholesteatoma demonstrates aggressive growth with greater extension and higher rates of residual and recurrent disease compared with adults, due to anatomic and physiologic differences. The aim of this study was to retrospectively analyze the number of patients with advanced

cholesteatoma, the clinical presentation, disease extension with imaging, and associated complications such as hearing loss, facial paralysis, labyrinthine fistula, dural involvement, etc. The surgical issues in the management of the disease and the outcomes were studied.

METHODS

This retrospective study consisted of fifty one patients who presented with advanced cholesteatoma to Madras ENT Research Foundation, a tertiary referral centre in South India from January 2010 to January 2020. The inclusion criteria included all patients with extensive disease and presence of complication. The exclusion criteria included patients with early disease and lack of complications. All patients underwent ENT examination, otomicroscopy, audiological evaluation and radiological evaluation (Figures 1-3). CT scans helped confirm the extent of disease, presence of labyrinthine fistula, involvement of the fallopian canal, tegmen, cochlea, internal auditory meatus, and internal carotid artery. Magnetic resonance imaging (MRI) was useful in patients with dural inflammation, sigmoid sinus thrombosis and in suspected intracranial involvement. All patients underwent surgical management of the disease and associated complication, if any. In all patients a canal wall down procedure was done for complete disease clearance. Ossicular reconstruction was done in all patients except in nine patients who had profound hearing loss. Facial nerve decompression was done in four patients with facial palsy. Patients with labyrinthine fistula (Figure 4) had complete disease clearance. The fistula was located in the lateral semicircular canal in all patients. At the end of the procedure, matrix covering the fistula was removed under high magnification, and sealed with temporalis fascia. Patients with cholesteatoma extending into the cochlea underwent complete disease removal. Patients with cholesteatoma in the internal auditory meatus had profound hearing loss and underwent a translabyrinthine approach for disease clearance (Figure 5 and 6). Cerebrospinal fluid (CSF) leak was controlled by packing with fat, temporalis fascia and fibrin glue. In patients with recurrent cholesteatoma and fungus cerebri (Figure 7), the herniating brain tissue was coagulated with bipolar diathermy, CSF gusher was controlled by using fat, fascia and fibrin glue. In patients with jugular bulb or sigmoid sinus involvement, matrix was dissected completely. One patient had cholesteatoma involving the internal carotid artery; matrix was dissected off without any adverse event.

Otomicroscopy was done at all follow-up visits to diagnose recurrence. In two patients who had granulations in the mastoid cavity, diffusion weighted MRI helped to diagnose recurrence. (Figure 8). For the statistical analysis, paired t test was used to study the outcome of surgery. Follow up after surgery ranged from 2 months to 10 years and the mean follow-up period was 69 months.

Figure 1: CT scan showing cholesteatoma with labyrinthine fistula.

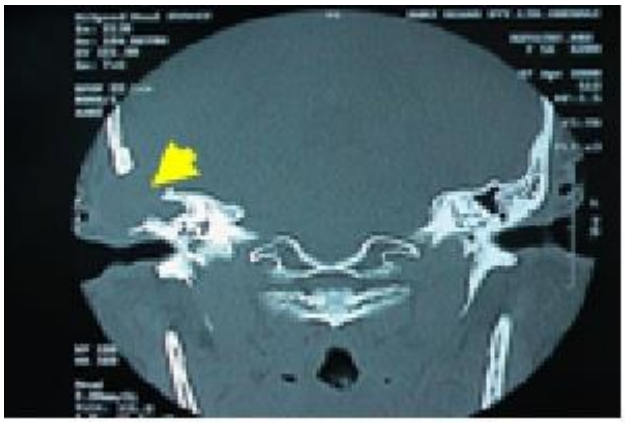

Figure 2: Recurrent cholesteatoma right side with extradural abscess.

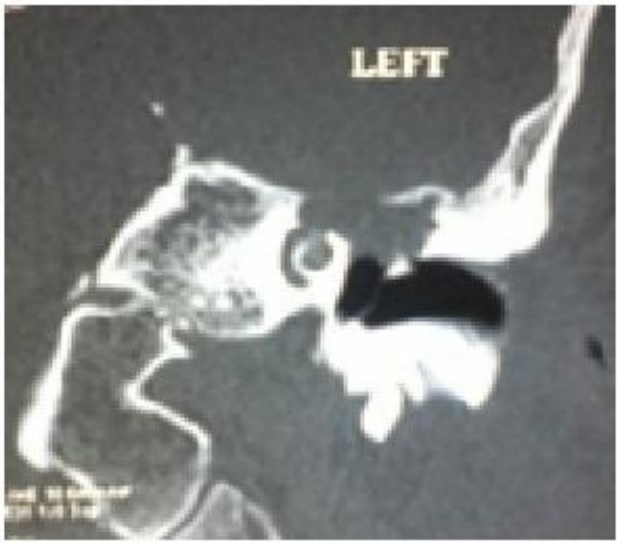

Figure 3: Cholesteatoma with tegmen erosion.

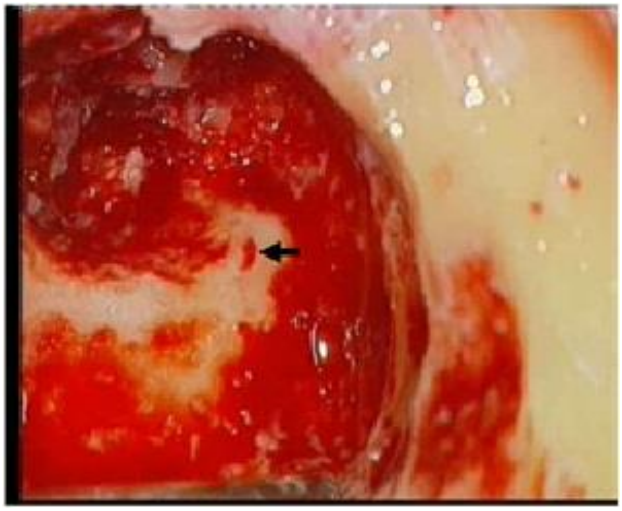

Figure 4: Labyrinthine fistula in a patient with cholesteatoma.

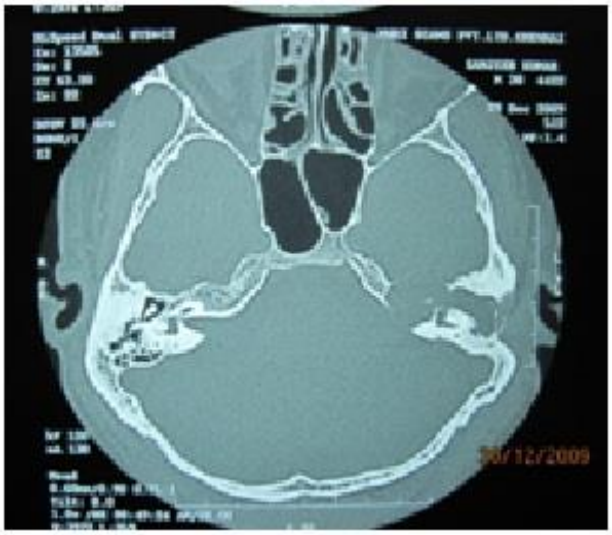

Figure 5: CT scan cholesteatoma involving the left internal auditory meatus.

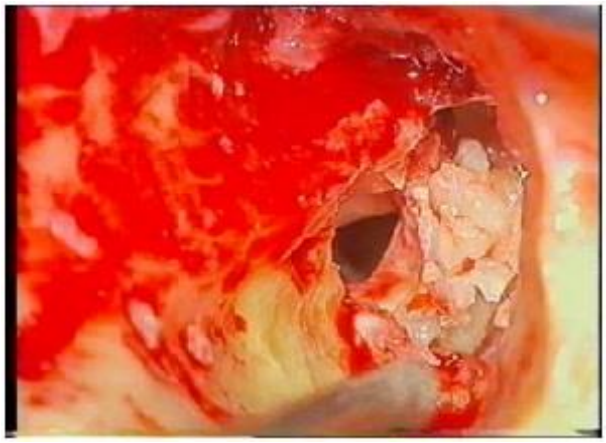

Figure 6: Intra-operative picture showing translabyrinthine approach for removal of internal auditory meatus cholesteatoma.

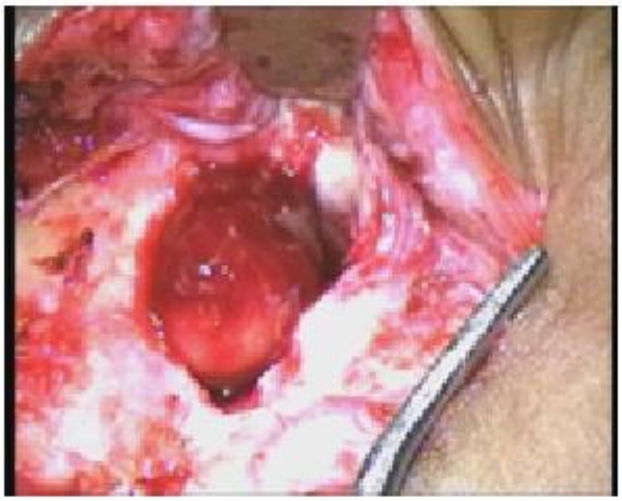

Figure 7: Cholesteatoma and fungus cerebri seen at revision surgery.

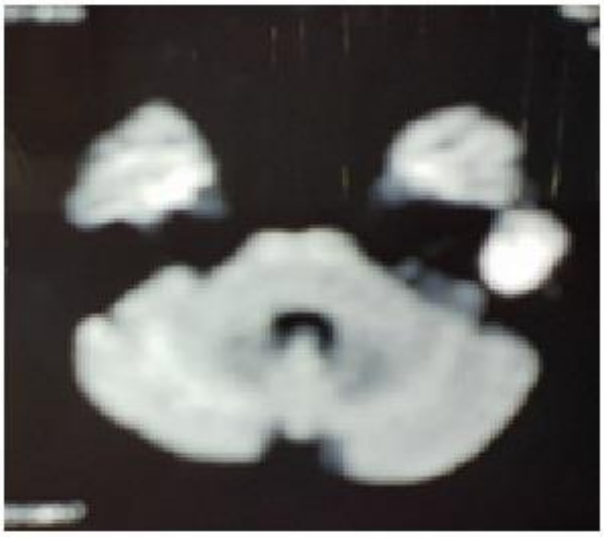

Figure 8: Non-echo planar diffusion-weighted MRI showing recurrent cholesteatoma.

RESULTS

Fifty one patients presented with advanced cholesteatoma in Madras ENT Research Foundation, a tertiary referral centre in South India from January 2010 to January 2020. The age range was 6 years to 68 years (mean 43 years). There were 33 males and 18 females in the study. Six patients were in the pediatric age group. Hearing loss was the commonest symptom seen in 39 patients; 9 patients had profound sensorineural hearing loss on presentation (Figure 9). Four patients presented with facial palsy (grade VI in 3 patients). The sites of involvement in advanced disease and complications are mentioned in Figure 10. In all patients a canal wall down procedure was done for complete disease clearance. In 42 patients, the surgery was a primary procedure and in 9 patients, the surgery was a revision procedure; primary procedure was done elsewhere in these patients. No surgical

complications were noted. The outcomes of surgical management were good and patients were on an average follow-up of 69 months. In 49 patients canal wall down mastoidectomy was effective in disease clearance ($p<0.05$). Two patients had recurrent disease and became disease free after revision surgery. Hearing improvement was noted in seventy percent of patients. None of the patients had worsening of hearing after the procedure.

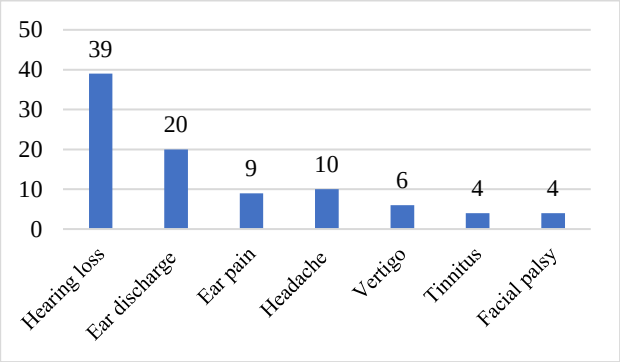

Figure 9: Clinical presentation of patients with advanced cholesteatoma.

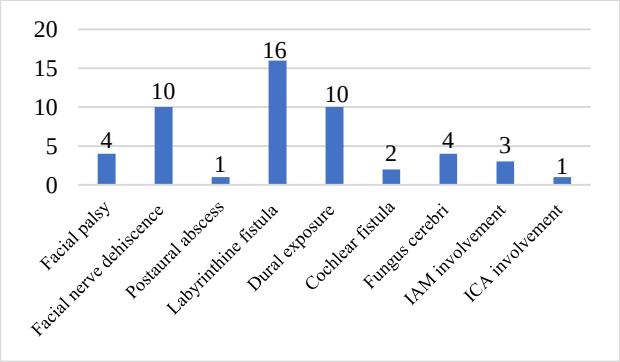

Figure 10: Sites of involvement and complications in patients with advanced cholesteatoma.

Vertigo was reported by 6 patients prior to the surgery. Post-surgery, none of the patients experienced vertigo ($p<0.05$). Tinnitus was present in 4 patients pre-operatively. Post-surgery, there was resolution of tinnitus in all patients ($p<0.05$). The p value as shown above is significant. Facial palsy was the presenting feature in four cases, of which three patients presented with grade VI palsy. In one patient the paralysis recovered completely and in three patients, facial function improved to House – Brackmann grade III after facial nerve decompression.

DISCUSSION

Cholesteatoma is a cyst-like mass lined with stratified squamous epithelium, usually keratinizing, and filled with desquamating debris.[3] Cholesteatoma activates osteoclasts and will erode through bone.[4] Cholesteatoma is a potentially serious disease which can lead to life-threatening complications.[5] Cholesteatoma displays enzymatic keratin debris with uninhibited growth and risk of ossicular, mastoid, and temporal bone invasion when left untreated.[6] The annual incidence of acquired cholesteatoma ranges from 9 to 12.6 cases per 100,000 adults and from 3 to 15 cases per 100,000 children. A male predominance of 1.4:1 in cholesteatoma incidence has been reported in literature.[1] Advanced cholesteatoma is not infrequently seen in otolaryngology practice.[7] Cholesteatoma-induced bone destruction may be a multifactorial process, involving biomechanical (i.e., pressure), biochemical (i.e., osteolytic enzymes), and cellular (i.e., osteoclasts) factors (13Y16).[8] The proximity of the middle ear cleft and mastoid air cells to the intratemporal and intracranial compartments increases the risk of infectious complications.[7]

The presenting signs and symptoms associated with cholesteatoma depend on the extent of disease. Signs and symptoms of more advanced disease include sensorineural hearing loss, which implies involvement of the cochlea, headache, dizziness, facial nerve paralysis, altered mental status, etc.[3] In a series by Osma et al, 78% of subjects who had complications secondary to chronic otitis media were found to have cholesteatoma. Labyrinthine fistula is the most common complication and is reported in approximately 7% of cases. Patients who have erosion of the labyrinth present with vertigo and a positive fistula test on examination. Due to its location near the antrum, the horizontal semicircular canal is the most commonly involved, and accounts for approximately 90% of these fistulae. Although the horizontal canal is usually involved, fistulae can occur in both the superior canal and posterior canal, and in the cochlea. Cochlear fistulae are associated with a much higher incidence of sensorineural hearing loss.[7]

Cholesteatoma in children is more aggressive than in adults, is more extensive, causes greater ossicular chain pathology, has higher recurrence rates and is more difficult to eradicate.[9] Extensive mastoid pneumatization in children provides adequate space, which allows cholesteatoma to spread, exacerbating the aggressive behavior of pediatric cholesteatoma. The immature development of the Eustachian tube is believed to be associated with the rate of disease recidivism in children, which is 2 to 10 times higher than that of adults. Children with craniofacial syndromes are predisposed to develop cholesteatoma. Approximately 0.9–5.9% of children with a cleft palate develop primary acquired cholesteatoma.[1]

Preoperative assessment should include radiographic imaging to focus on the extent of disease.[6] High-resolution computed tomography of the temporal bone and MRI are the imaging techniques of choice. They are important for assessing the extent of disease, to look for complications and for planning the surgical approach.[2] High-resolution computed tomography of the temporal bone has high sensitivity but poor specificity in the case of a mass lesion because it may correspond to granulation tissue, secretion, cholesterol granuloma, or neoplasm.

Presence of bony erosions indicates cholesteatoma. MR imaging with the conventional sequences (T1WI, T2WI, post contrast T1WI) provides additional information. Cholesteatoma is usually hypointense or isointense on T1WI and hyperintense on T2WI. Non-echo planar diffusion-weighted (non-EPI DW) and DWI propeller sequences have improved diagnostic sensitivity and specificity for even small (5 mm) cholesteatomas.[10]

Surgical management is the mainstay of management.[6] The primary surgical goals are eradication of disease and the creation of a dry middle ear; maintenance or restoration of hearing is a secondary goal.[11] Cholesteatoma matrix must be removed completely to avoid recurrence. The canal wall down procedure consists of a modified radical mastoidectomy and results generally in a lower rate of recurrence.[12]

Dehiscence of the facial nerve is common in patients with cholesteatoma, and the surgeon should avoid damage to the facial nerve.[8] Dehiscence is most frequently observed in the tympanic portion of the facial nerve near the oval window.[4] Selesnick et al reported that the incidence rate of facial nerve dehiscence in cholesteatoma surgery in 67 cases was 33%.[13] The presence of a semicircular canal fistula increases the risk of dehiscence of the facial nerve by about 4.7 times.[14] Chronic otitis media causing facial nerve paralysis is most likely due to cholesteatoma. The onset of paralysis is either acute or gradual.[3] Facial nerve paralysis occurs rarely with middle ear cholesteatomas.[10] Prevalence of facial palsy is 0.04 to 0.16 per cent of patients with cholesteatoma.[15] Facial palsy can be present in 20%–64% of extensive cholesteatoma cases, half of which are higher grades.[10] The incidence of facial palsy in petrous apex cholesteatoma is reported to be between 50 to 81 per cent.[15] In presence of facial paralysis, early surgery to remove disease and preserve facial function is essential. Cawthorne stated that nerve sheath incision is not necessary for incomplete paralysis but should be performed for complete paralysis.[3] Patients undergoing surgery >2 months after the onset of facial paralysis and patients with facial palsy associated with petrosal cholesteatoma have poor outcomes.[3]

Labyrinthine fistula continues to be one of the most common complications of cholesteatoma with a frequency varying approximately from 4% to 12%.[16] Sensorineural hearing loss can occur. Computed tomography is a good diagnostic investigation for lateral semicircular canal fistula.[4] Collagenase activity in the matrix and the pressure exerted on the bone contribute to bone erosion. In nearly 90% of the patients, the fistula is located in the horizontal semicircular canal. Canal-wall-down surgery with removal of the matrix, and sealing of the fistula is recommended.[16] Cholesteatoma over a fistula in the labyrinth is removed at the end of surgery. Suctioning over the fistula must be avoided. After the matrix is removed, the fistula can be closed with temporalis fascia, temporalis fascia and fibrin glue, or temporalis fascia and bone pate.[4] A 35% rate of profound deafness has been reported with fistulae involving the promontory, compared with a 3% rate of deafness with fistulae involving the semicircular canal. The use of corticosteroids at the time of cholesteatoma removal from fistulae may protect hearing.[7]

Dural exposure of the mastoid tegmen can occur with cholesteatoma.[17] The matrix may be densely adherent to the dura of the middle and posterior fossae and is one of the major reasons for recurrences. Using bipolar coagulation to devitalize the epithelium is the safest way to ensure complete clearance. There is a risk of opening the dura and causing an intra-operative CSF leak which can be managed by use of muscle plugs and obliteration of the cavity.[2] When the dura is involved, the matrix should be removed very carefully. The subarachnoid space should not get exposed to cholesteatoma or chemical meningitis can ensue. At revision surgery, particular care should be taken especially in the presence of fungus cerebri.

In presence of intracranial complications, combined neurosurgical and otological clearance is vital. Jugular bulb, sigmoid sinus involvement, Internal carotid artery involvement should be identified before surgery.[2] The sigmoid sinus and the jugular bulb are difficult areas from which to remove cholesteatoma because of their fragile walls, and the lower cranial nerves are at risk of injury.[2] Infection of the petrous apex is dangerous because of its proximity to the middle and posterior cranial fossae.[7] The Sanna classification for petrous bone cholesteatomas (PBCs) includes supralabyrinthine, infralabyrinthine, massive, infralabyrinthine-apical, and apical depending on both the location and the extent of the lesion.[2] The extension of PBCs into the clivus, sphenoid sinus, or nasopharynx is rare, but can be extremely difficult to treat. These extensions require a combination of an infratemporal fossa approach type B with a transotic approach or modified transcochlear approach type A.[2]

In our study, several challenging situations were encountered and dealt with successfully. A thorough pre-operative imaging study proved vital in assessing the spread of cholesteatoma. Long-term follow-up is necessary after surgical treatment.[5] Diffusion weighted MRI has a role in detecting recurrence. Non-EPI DW-MRI sequences are superior to EPI sequences in identifying recurrent or residual cholesteatoma.[1]

Improvement in understanding the pathogenesis of cholesteatoma may lead to new treatment strategies. Stem cells residing in cholesteatoma tissue may contribute to disease progression. Cells expressing the “stemness” markers Nestin and S100B have been detected in middle ear cholesteatoma and auditory canal skin. Middle ear cholesteatoma-derived stem cells have been shown to display an enhanced susceptibility to inflammatory stimuli, and this suggests a possible contribution to the inflammatory environment in cholesteatoma tissue.[18]

CONCLUSION

Cholesteatoma has the potential to grow relentlessly and destroy neighboring structures. Advanced cholesteatoma with facial nerve, skull base, labyrinth involvement, etc is not uncommon in the Indian scenario. Complete eradication of disease must be attempted. Long-term clinical and radiological follow-up if needed is mandatory to detect recurrence of cholesteatoma.

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