CONGENITAL CHOLESTEATOMA WITH POSTERIOR CANAL WALL DESTRUCTION IN AN ADULT

Maximiliano Agustian Mahardhika*, Eka Putra Setiawan*

*Department of Otorhinolaryngology-Head and Neck Surgery, Faculty of Medicine UdayanaUniversity/ Prof Dr. I. G. N. G. Ngurah Hospital Denpasar

ABSTRACT

**Background:** Congenital cholesteatoma is a mass behind an intact tympanic membrane that develops from keratinizing squamous epithelium remnants in the temporal bone and has no prior perforation or history of otologic surgery. The most common symptom of congenital cholesteatoma is conductive hearing loss, which can occur in a number of locations inside the temporal bone. Surgical approach is needed to remove the diseases. **Purpose:** To discuss the best surgical approach for cholesteatoma. **Case report:** A 47-year-old male patient presented with hearing loss, and also long term otorrhea and otalgia. The diagnosis was congenital cholesteatoma with posterior canal wall destruction. Patient had been performed modified radical mastoidectomy. Patient showed improvement in symptoms and hearing after surgery. **Methods:** Method in this case report is evidence-based case report. Clinical question: Is modified radical mastoidectomy type of canal wall down mastoidectomy the best choice for treating and preventing recurrencry of cholesteatoma? To answer this question, we search the evidence from PubMed, Science Direct and Chocrane with the keywords: “cholesteatoma” AND “canal wall down mastoidectomy” AND “recurrence”. **Results:** From the searching method, we found 4 articles which was relevant and included to the critical appraisal step. **Conclusion:** Canal wall down mastoidectomy is the best management for cholesteatoma, which in this case patient has congenital cholesteatoma. This technique can also improve patient’s hearing.

**Keywords:** congenital cholesteatoma, cholesteatoma, modified radical mastoidectomy, canal wall down mastoidectomy.

INTRODUCTION

Congenital cholesteatoma by definition is a mass filled with keratinized squamous epithelial remnants located in the temporal bone, behind non perforated tympanicmembrane. The prevalence of congenital cholesteatoma is very rare, with a worldwide percentage of about 1-5% of all cholesteatoma cases. Congenital cholesteatoma is generally identified at the age of 6 months to 5 years. This causes the incidence of congenital cholesteatoma found in adult patients is less frequent.1,2

The pathophysiology of congenital cholesteatoma is still being debated by experts. The failure of epidermoid growth in the temporal bone is assumed to be the primary factor in congenital cholesteatoma development. In the third and fifth weeks of fetal development, there is a buildup of keratinized squamous epithelium during the development of the middle ear mucosa. The squamous cells which trapped in the middle ear space continue to grow forming cholesteatoma.3

Diagnosis of congenital cholesteatoma is often made at the time of surgery. Criteria diagnosis for congenital cholesteatoma are if a white mass is found behind an intact tympanic membrane (normal pars flacida and pars tensa), and there is no prior history of otorrhea, tympanic membrane perforation, or ear surgery.4

Treatment for congenital cholesteatoma is surgical removal of the cholesteatoma. Surgical approach for is determined by its stage or type. The operation can vary from tympano- mastoidectomy to canal wall down mastoidectomy. Canal wall down mastoidectomy is more frequent used because of its success rate.4

The purpose of this case report is to present a case of congenital cholesteatoma with posterior canal wall destruction in a 47-year-old male patient who underwent canal wall down mastoidectomy.

CASE REPORT

A 47-year-old man came to the Prof. Dr. I. G. N. G. Ngoerah Denpasar Hospital ORL-HNS’s clinic, with a complaint of a white-yellowish discharge from the right ear since 1 year ago. Complaints accompanied by pain and hearing impairment in the right ear since 1.5 years ago. There were no complaints of fever, headache, vertigo and facial paralysis. History of trauma, previously ear discharge and ear surgery were denied.
On physical examination in the right ear, the ear canal is narrow filled with secret and granulation tissue, the origin of the granulation is difficult to determine, tympanic membrane cannot be assessed. There is no abnormality in the left ear.

![Figure 1. Right ear canal filled with granulation tissue](image1)

**Figure 1. Right ear canal filled with granulation tissue**

![Figure 2. Head CT-scan axial view](image2)

**Figure 2. Head CT-scan axial view.**

**Figure 3. Head CT-scan coronalview.**

**Figure 4. Audiogram pre operative.**

On examination with 512 Hertz tune fork, conductive hearing loss was found in the right ear. Audiometric examination revealed that the right ear had severe hearing loss with hearing threshold of 73.5 dB, while the left ear had normal hearing with hearing threshold of 25 dB. From head CT-scan showed a right cholesteatoma that caused destruction of the
anteroposterior wall of the right mastoid bone, right tympanic tegmen, right scutum, right Prussak space and right hearing ossicle.

The patient was diagnosed with suspect malignant type of chronic suppurative otitis media (CSOM), with a differential diagnosis of congenital cholesteatoma. The patient was planned to undergo a modified radical mastoidectomy (Bondy procedure) under general anesthesia. The patient underwent surgery, at the time of the operation it was found that the mastoid cortex was destroyed by cholesteatoma which filled the mastoid cavity, the posterior wall of the ear canal was partially destroyed, granulation tissue came out through the wall of the ear canal which destroyed the ear canal, the tympanic membrane was intact and the malleus ossicles were still remaining. There were no perioperative or post operative complications. Post operative diagnosis was congenital cholesteatoma post radical modified mastoidectomy (Bondy procedure).

Two days after surgery, the fixation bandage and drain were removed, the surgical wound was well maintained, then the patient was discharged. Ten days after surgery, stitches and tampons in the right ear canal were removed. Then the patient control routinely every week for the first 1 month. Three months after the operation, the patient returned to control, there were no complaints, then an audiometric examination was performed and the results were found moderate to severe conductive hearing loss (hearing threshold at 60 dB) in the right ear, with normal hearing in the left ear.

Figure 5. Cholesteatoma and intact tympanic membrane are seen
The selection of literatures was based on inclusion criteria and exclusion criteria. The inclusion criteria were: 1) cholesteatoma. 2) cholesteatoma treated by surgery. 3) thorough follow-up for recurrence. The exclusion criteria were studies that inconsistent with the study design and congenital cholesteatoma management not by surgery.

RESULT

The literature search obtained after screening and searching full texts which were released in the last 10 years and relevant with the topics.

Piras et al., conducted a study with 236 patients with cholesteatoma. Surgical canal wall down tympano- mastoidectomy were performed on 134 patients. The average follow-up period was 100 months. According to the study, there are decreased rates of residual and recurrent cholesteatoma (7.6% and 2.3%) in patients who received canal wall down tympanomastoidectomy. There was also air bone gap improvement on post operative audiometry.4

METHOD

A literature search was conducted through Pubmed, Science Direct and Chocran with keywords “cholesteatoma” AND “canal wall down mastoidectomy” AND “recurrence”. 
### Table 1. Description of Eligible Journal Article

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<td>To evaluate long-term effects of CWUT and CWDT within pediatric population with cholesteatoma, with a focus on hearing outcomes and cholesteatoma recurrence rates.</td>
<td>To investigate the effectiveness of inside-out treatment in totally eliminating cholesteatoma from the middle ear and mastoid, maintaining hearing, and improving quality of life following mastoidectomy in terms of recurrent discharge, wax, and granulations.</td>
<td>To compare disease recidivism rates between canal wall up mastoidectomy and a canal wall down with obliteration technique.</td>
<td>To evaluate the long-term morphological and functional results of the canal wall down tympanoplasty for the treatment of tympano-mastoid cholesteatoma, as well as the prognostic variables.</td>
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<tr>
<th>Study design</th>
<th>Cohort study</th>
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<tr>
<td>Participants</td>
<td>236 patients</td>
<td>100 patients</td>
<td>98 patients</td>
<td>895 patients</td>
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<td>Intervention</td>
<td>canal wall-up or a canal wall-down tympanomastoidectomy</td>
<td>Inside out mastoidectomy (canal wall-down tympanomastoidectomy)</td>
<td>canal wall up mastoidectomy and a canal wall down with obliteration technique</td>
<td>canal wall down tympanoplasty</td>
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<td>Outcome measurements</td>
<td>Rates of postoperative mean Air-Bone Gap (ABG), recurrent cholesteatoma and residual cholesteatoma.</td>
<td>Rates of residual cholesteatoma and mean Air Bone Gap (ABG) after surgical.</td>
<td>Recidivism rate and post-operative ear discharge rate.</td>
<td>Rate of recidivism and air pure-tone-average.</td>
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| Results | The CWDT procedure offers a definitive surgical therapy, with minimal residual and recurrence rates and equivalent audiological outcomes to the CWUT approach. | At the conclusion of the research period, none of the patients still had residual disease. After surgery, there was a noticeable improvement in the mean air conduction (p<0.01) and the mean air bone gap (p<0.01). Also, 95% of patients had a dry, self-cleaning cavity. | When compared to a canal wall up approach, canal wall down mastoidectomy with obliteration offers a lower rate of recidivism and post-operative ear discharge. | Recidivism rates were greater in pediatric patients, at 7.7%. In 36.4% of patients, air pure-tone-average (aPTA) 30 dB was obtained. |
Chamoli et al., investigate 100 cholesteatoma patients who had canal wall down tympanomastoidectomy surgery. They found 100% success rate after surgery in terms of residual disease and air bone gap improvement. On routine follow-up, 95% of the patients had dry self-cleaning cavities, with only 5% patients require continuous cavity cleaning. According to Wilkie et al., compared to the canal wall up surgery, disease recidivism rates were reduced in patients who underwent canal wall down mastoidectomy with mastoid obliteration. Clinical examination and/or radiological imaging were used to confirm the clinical suspicion of recidivism.

Pareschi et al., found that recidivism from patient who underwent canal wall down mastoidectomy was higher in pediatric patients than the adults. Postoperatively, hearing function improvement was also achieved in 36.4% of patients.

**DISCUSSION**

Congenital cholesteatoma is diagnosed based on the patient's medical history, physical examination, and supporting investigations. We report acase of a 47-year-old man who complained of discharge from the ear canal, hearing loss and otalgia on his right ear. From physical examination of the right ear, we found narrow ear canal filled with granulation tissue and secret, also tympanic membrane cannot be assessed. From examination with tune fork and audiometric examination showed that the right ear had conductive hearing loss. From head CT-scan showed a right cholesteatoma. Congenital cholesteatoma originates from epithelial remnants trapped in the temporal bone during embryogenesis. Congenital cholesteatoma cases are relatively rare cases where congenital cholesteatoma has progressive growth and generally appears at a later age with a preponderance in males. Goh et al., reported the results of a retrospective study from 1999 to 2006 there were 5 cases of congenital cholesteatoma or 12.2% with an age range of 5–18 years, where there were more men, namely 3 men and 2 women. While research by Park et al., from 1995 to 2006 obtained 35 cases of middle ear congenital cholesteatoma with ages ranging from 1 to 13 years old and a male to female ratio of 3:4:1. Kojima et al., reported 48 cases congenital cholesteatoma with ages ranging from 2 to 62 years and a mean of 16.7 years.

Clinical symptoms in this patient in the form of discharge from the ear canal and hearing loss. Based on research conducted by Steward et al., found that 73% patient complaint hearing loss and 55% patient with otorrhea. According to Park et al., individuals most frequently seek medical attention due to hearing loss. The percentage obtained was 29% for complaints of hearing loss, 20% otalgia, 11.4% tinnitus, and 8.5% ear fullness. Otorrhea occurs due to cholesteatoma that destroys the posterior wall of the ear canal so that granulation tissue is formed. Cholesteatoma can become infected, causing symptoms such as CSOM. The preoperative diagnosis was suspected to be malignant type of CSOM on the right ear because in the course of the disease there was granulation tissue in the right ear canal which resulted in discharge of secretions since 1 year ago. The tympanic membrane is difficult to assess until surgery. Congenital cholesteatoma is often diagnosed late because it is often asymptomatic at first. Usually patients come when there are already complications that difficult to distinguish from malignant types of CSOM. Research by Park et al., obtained as many as 17 cases (63%) of 27 cases of congenital cholesteatoma, patients diagnosed with congenital cholesteatoma after the lesion extended to the mastoid. A CT-scan is needed to determine the location of the cholesteatoma and its extent. Cholesteatoma will be seen as a hypodense picture with clear boundaries and bone destruction in the form of erosion of the tegmen and sigmoid sinus. Based by Derlacki and Clemis criteria which were later modified by Levenson stated that the diagnosis of congenital cholesteatoma can be made if there is a cholesteatoma with intact tympanic membrane, and no history of otorrhea or previous ear surgery. Based on the pathogenesis of congenital cholesteatoma according to epithelial Rest theory, cholesteatoma in the mastoid can occur due to trapped epithelial remnants in the third and sixth weeks of fetal life, when closure of the neural cleft occurs. Congenital cholesteatoma in the mastoid is often found in the mastoid cavity, same to this case where cholesteatoma in mastoid cavity was found in 47-year-old patient.

Having the cholesteatoma surgically removed is the gold standard for treating congenital cholesteatoma. In this patient, canal wall down mastoidectomy was performed because the lesion destroyed the mastoid and ossicles as well as the posterior wall of the ear canal, with no perforation on tympanic membrane and no requirement for ossicular reconstruction. Surgery in cases of congenital cholesteatoma is very varied. The choice of the type of surgery is adjusted to the stage, site and type of cholesteatoma.

Surgery can range from tympanomastoidectomy to radical mastoidectomy. Potsie et al., divided cholesteatoma into 4 stages and related to the choice of surgery, namely stage 1: involving one quadrant not affecting the ossicles and mastoid, stage 2: multiple quadrants not affecting the ossicles and mastoid, stage 3: affecting the ossicles not affecting the mastoid, and stage 4: has involved the mastoid. The surgery is adjusted according to the stage. At stages 1 and 2 tympanomastoidectomy can be performed. Stage 3 involves an extended tympanomastoidectomy but sometimes requires canal wall up mastoidectomy. At stage 4, a canal wall down mastoidectomy was performed. Park et al., performed surgery on 17 cases of stage 4 cholesteatoma, performing canal wall up tympanomastoidectomy in 10 cases (59%) and canal wall down tympanomastoidectomy in 7 cases (41%). Research conducted by Inokuchi et al., obtained 23 cases of congenital cholesteatoma. The most common surgical procedure is tympanomastoidectomy. As many as 57% of cases underwent canal wall down procedure and 9% of cases underwent canal wall up procedure. The advantage of canal wall down is a lower recurrence and residual rate.

Audiometric examination before surgery revealed a severe degree of conductive hearing loss (hearing
threshold at 73.5 dB). It might happen because a granulation tissue buildup in the external ear canal may have caused obstruction or there may have been an ossicular discontinuity. Right ear moderate-severe hearing loss (hearing threshold at 60 dB) was discovered on postoperative audiometric testing. This patient had an improvement of 13.5 dB. This is in line with research done by Park et al., patients who have undergone radical mastoidectomy surgery also shown improvement on their hearing thresholds.\(^9\) Other studies also showed that canal wall down mastoidectomy can improve air bone gap in audiometry examination post operatively.\(^5^7\) After surgery done, patients need to be monitored regularly for long term.\(^5\) In the early postoperative period, the surgical cavity's process of epithelialization must be carefully monitored. Granulation tissue can inhibit the epithelialization process. Local antibiotics can inhibit infection and granulation tissue formation. In long term, the patient should come regularly to clean the cavity from desquamation of the epithelium and cerumen. Besides that, it is also necessary to pay attention to complications that might arise such as repeated infections, formation of cholesteatoma or presence of residual cholesteatoma.

REFERENCES

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