CONGENITAL CHOLESTEATOMA WITH POSTERIOR CANAL WALL DESTRUCTION IN AN ADULT

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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 ottorhea 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 recurrency 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 choles- teatoma is often made at the time of surgery. Criteria diagnosis dor congenital cholesteatoma are if a white
mass is found behind an intact tympanic membrane (normal pars flaccida and pars tensa), and there is no prior history of otorrhea, tympanicmembrane perforation, or ear surgery.[3]

Treatment for congenital choles- teatoma is surgical removal of the cholesteatoma. Surgical approrach 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 usedbecause 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 downmastoidectomy.

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.

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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.

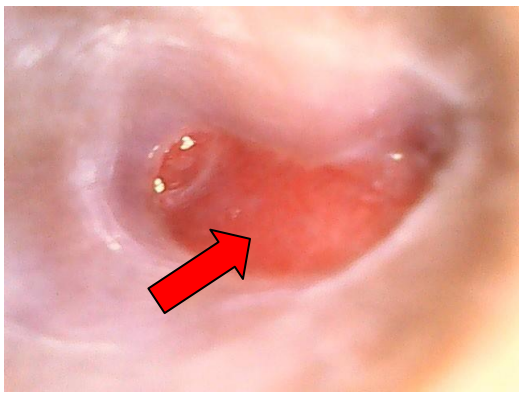

Figure 1. Right ear canal filled withgranulation tissue

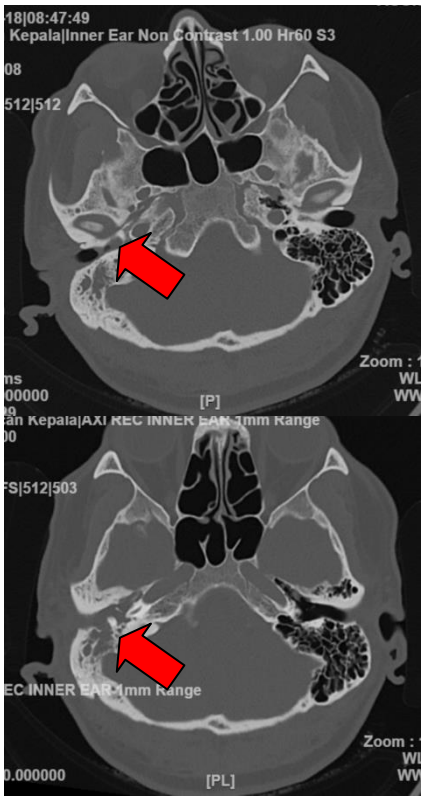

Figure 2. Head CT-scan axial view.

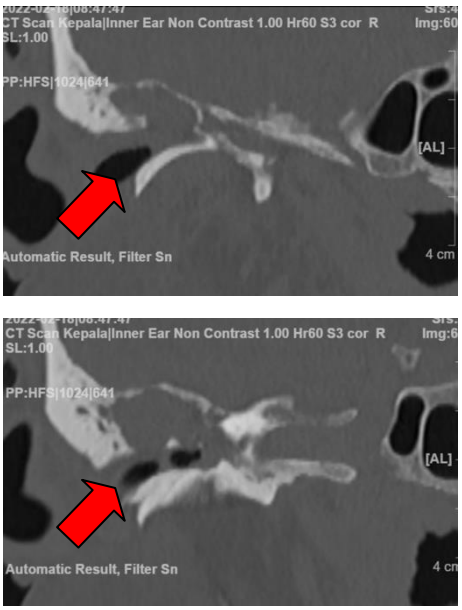

Figure 3. Head CT-scan coronalview.

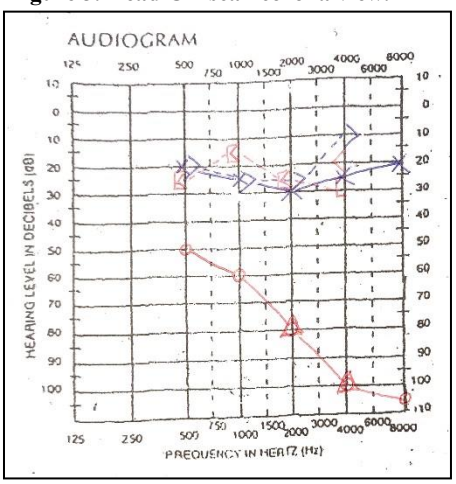

Figure 4. Audiogram pre operative.

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 plannedto 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. Tendays 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.

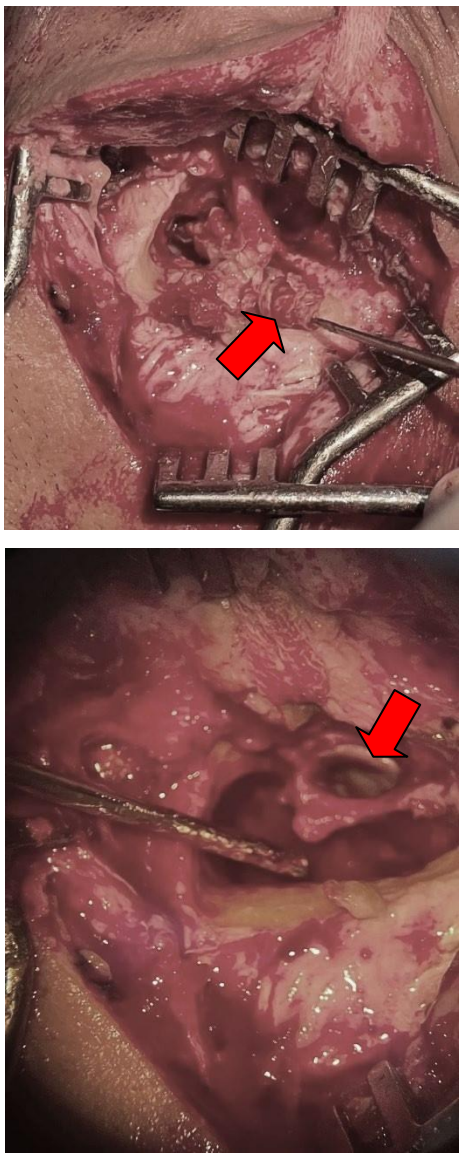

Figure 5. Cholesteatoma and intacttympanic membrane are seen

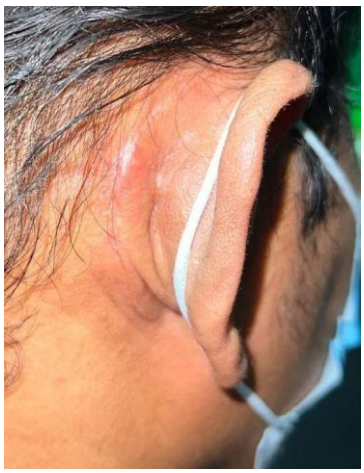

Figure 6. Post operative wound.

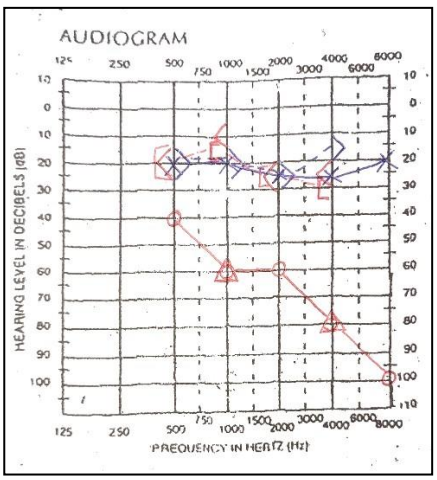

Figure 7. Audiogram post operative.CLINICAL

QUESTION

Is modified radical mastoidectomytype of canal wall down mastoidectomy the best choice for treating and preventing recurrency of cholesteatoma?

METHOD

A literature search was conducted through Pubmed, Science Direct and Chocrane with keywords “choles-teatoma” AND “canal wall down mastoidectomy” AND “recurrence”.

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 yearsand 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]

Table 1. Description of Eliglible Journal Article

	Piras et al (2021)	Chamoli et al (2018)	Wilkie et al (2019)	Pareschi et al (2019)
Research question of thestudy	To evaluate long-term effects ofCWUT and CWDT within pediatric population with cholesteatoma, with a focus on hearing outcomes and cholesteatoma recurrence rates.	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.	To compare disease recidivism rates between canal wall up mastoidectomy and a canalwall down with obliteration technique.	To evaluate the long- term morphological andfunctional results of thecanal wall down tympanoplasty for the treatment of tympano- mastoid cholesteatoma, as well as the prognostic variables.
Study design	Cohort study	Cohort study	Cohort study	Cohort study
Participants	236 patients	100 patients	98 patients	895 patients
Intervention	canal wall-up or a canal wall- down tympanomastoidectomy	Inside out mastoidectomy (canal wall-down tympanomastoidectomy)	canal wall up mastoidectomy and a canalwall down with obliteration technique	canal wall down tympanoplasty
Outcome measurements	Rates of postoperative mean Air-Bone Gap (ABG), recurrent cholesteatoma and residual cholesteatoma.	Rates of residual cholesteatoma andmean Air Bone Gap (ABG) after surgical.	Recidivism rate and post-operative ear discharge rate.	Rate of recidivism andair pure-tone-average.
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 ($p<0.01$). Also95% of patients had a dry, self- cleaning cavity.	When compared to a canalwall up approach, canal wall down mastoidectomywith obliteration offers a lower rate of recidivism and post-operative ear discharge.	Recidivism rates weregreater 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 tympano-mastoidectomy 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 cavitycleaning.[5]

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.[6]

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 in36,4% of patients.[7]

DISCUSSION

Congenital cholesteatoma isdiagnosed based on the patient's medical history, physical examination, and supporting investigations. We report acase of a 47-year-old man which
complained discharge from the ear canal, hearing loss and otalgia on his right ear. From physical examinationon the right ear, we found narrowingear canal filled with granulation tissue and secret, also tympanic membrane cannot be assessed. From examinationwith tune fork and audiometric
examination showed that the right earhad conductive hearing loss. From head CT-scan showed a right cholesteatoma. Congenital cholesteatoma originates from epithelial remnants trapped in thetemporal bone during embryogenesis. Congenital cholesteatoma cases are relatively rare cases where congenital cholesteatoma has progressive growth and generally appears at a later agewith a preponderance in males. Goh et al., reported the results of a retrospective study from 1999 to 2008there were 5 casesof congenitalcholesteatoma or 12,2% with an agerange of 5-18 years, where there weremore on men, namely 3 men and 2women.[8] While research by Park et al.,from 1995 to 2006 obtained 35 cases of middle ear congenital cholesteatomawith ages ranging from 1 to 13 yearsold and a male to female ratio of 3.4:1.[9]Kojima et al reported 48 cases congenital choles-teatoma with ages ranging from 2 to 62 years and a mean of 16,7 years.[10]

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.[11] 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.[9] 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.[9]

A CT-scan is needed to determinethe 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 cholesteatomacan 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.[12,13] Congenital cholesteatoma in themastoid is often found in the mastoid cavity, same to this case where cholesteatoma in mastoid cavity was found in 47-years-old patient.

Having the cholesteatoma surgically removed is the gold standard for treating congenital cholesteatoma.[5-7] 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 requirementfor 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.[5-7]

Surgery can range from tympanomastoidectomy to radical mastoidectomy. Potsic 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.[14] Park et al., performed surgery on 17 cases of stage 4 cholesteatoma, performing canal wall up tympanomastoidectomy in 10 cases
(59%) and canal wall downtympanomastoidectomy in 7 cases (41%).[9] 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.[5,6,15]

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 (hearingthreshold 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 bonegap 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.

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