Case Report

Management of otosclerosis with obliterated footplate and round window involvement

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ABSTRACT

Background: Otosclerosis with obliterated footplate describes a condition with an overgrowth of bone so that the oval window is almost indistinguishable from the surrounding area. The occurence of round window obliteration is uncommon. In such cases, surgery is more difficult to perform and the results are often unsatisfactory. **Purpose:** To find out the management option for obliterative otosclerosis. **Case report:** A 35-year-old-man with suspected bilateral otosclerosis. The patient underwent transcanal endoscopic stapes surgery of the left ear with large fenestra stapedotomy procedure. **Clinical question:** In adult patients with obliterative otosclerosis, does stapes surgery improve hearing function and reduce complication? **Review method:** Using PubMed, EBSCOhost, Scopus and Proquest to search for the evidence of randomised control trial (RCT), cohort and case-control studies. Critical appraisal was conducted using critical appraisal tools from Center for Evidence Based Medicine (CEBM), Oxford University and qFAITH for systematic review/meta-analysis. **Result:** The literature searching used eligibility criteria based on keywords from clinical question found one study regarding the evaluation of intra-operative factors in otoslecrosis surgery outcomes, which was relevant to our clinical question. **Conclusion:** Large fenestra stapedotomy or stapedectomy is the preferred surgical technique for the management of obliterative otosclerosis. The involvement of round window caused a sensorineural hearing loss (SNHL) component and has a worse prognosis of hearing outcome post operatively. A further study is required to assess the best management for obliterative otosclerosis.

Keywords: obliterative otosclerosis, stapedotomy, stapedectomy, round window involvement

ABSTRAK

Latar belakang: *Otosklerosis dengan obliterasi* footplate *menggambarkan suatu kondisi pertumbuhan tulang yang berlebihan sehingga tingkap lonjong hampir tidak dapat dibedakan dengan daerah sekitarnya. Obliterasi sampai ke tingkap bundar jarang terjadi. Hal tersebut menyebabkan operasi lebih sulit dilakukan dan hasilnya seringkali tidak memuaskan.* ***Tujuan:*** *Untuk mengetahui pilihan tatalaksana otosklerosis obliteratif.* ***Laporan kasus:*** *Laki-laki 35 tahun dengan suspek otosklerosis bilateral. Pasien menjalani operasi stapes telinga kiri dengan pendekatan endoskopi transkanal menggunakan prosedur stapedotomi fenestra besar.* ***Pertanyaan klinis:*** *Pada pasien dewasa dengan otosklerosis obliteratif, apakah operasi stapes dapat memulihkan fungsi pendengaran dan mengurangi komplikasi?* ***Telaah literatur:*** *Pencarian bukti melalui* database PubMed, EBSCOhost, Scopus *dan* Proquest*. Untuk uji coba kontrol acak, kohort dan studi kasus-kontrol, penilaian kritis dilakukan dengan menggunakan penilaian kritis dari CEBM,* Oxford University *dan qFAITH untuk tinjauan sistematis/meta-analisis.* ***Hasil:*** *Penelusuran literatur berdasarkan kriteria dari pertanyaan klinis, didapati satu artikel mengenai evaluasi faktor-faktor intraoperatif yang relevan dengan pertanyaan klinis.* ***Kesimpulan:*** *Stapedotomi fenestra besar atau stapedektomi adalah teknik bedah yang lebih dipilih untuk tatalaksana otosklerosis obliteratif. Keterlibatan tingkap bundar menyebabkan*

komponen gangguan pendengaran sensorineural, dan memiliki prognosis hasil pendengaran pasca operasi yang kurang baik. Dibutuhkan studi lebih lanjut untuk menilai tentang penatalaksanaan terbaik untuk kasus otosklerosis obliteratif.

Kata kunci: *otosklerosis obliteratif, obliterasi* footplate*, obliterasi tingkap bundar, stapedotomi*

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INTRODUCTION

Obliterative otosclerosis is a condition in which the stapes footplate is very thick, filling the oval window niche and invading the annular ligament. The incidence of obliterative otosclerosis ranged about 4-13% of all otosclerosis patients that were surgically treated. Gristwood quoted by De Souza et al.[1] reported a 12.5% incidence of obliterative otosclerosis. Ayache et al.[2], reported an incidence of 4.7% and noted that 50% chance that obliterative otosclerosis were bilateral findings. Obliterated footplates were more often found in young adults and teenagers with the mean age onset of 19 years. It was more common in male than female with a proportion of 1.48:1. Otosclerosis is an autosomal dominant hereditary disorder with approximately 40% disturbance of genes.[1-3]

Although round window involvement is the second most common site after the oval window, it is still uncommon to get total obliteration of round window. If there was a round window obliteration, patient will have a component of sensorineural hearing loss which might occur once the round window niche is totally obliterated.[1]

Though there are some signs, symptoms and examination results that might lead to the diagnosis of otosclerosis, the definitive diagnosis of obliterative otosclerosis is usually made intra-operatively as a surprise finding. The management of obliterative otosclerosis comprises a great challenge for the otology surgeon because of the occurred pathology.[3,4]

This case report was presented to discuss the best operative techniques in the surgical management of the obliterated footplate and round window involvement.

CASE REPORT

A 35 years old male patient was referred to Otology outpatient clinic of the Department of Otorhinolaryngology Head and Neck Surgery, Cipto Mangunkusumo General Hospital with a chief complaint of fullness feeling in the left ear since one year previously. The patient also reported that he had a progressive hearing loss on the left ear since about three years, and intermittent tinnitus in both ears. The patient felt more comfortable using his right ear during phone calls. There was no history of ear discharge, dizziness nor facial asymmetry. No complaint of blocked nose nor itchy and nasal discharge.

From the physical examination, patient's general condition was good, compos mentis and the vital signs were within normal limits. Otomicroscopy examination of the right ear showed clear ear canal, no secrete nor cerumen, intact tympanic membrane with sclerotic plaque. The left ear showed clear ear canal, no secrete nor cerumen, intact tympanic membrane with sclerotic plaque. There was no abnormality on the nose, throat, and neck examination.

The result of tuning fork test, Rinne was negative in both ears in all frequencies (128, 256, 512, 1024, 2048 Hz), Schwabach were lengthened in both ears, and Weber showed lateralization to the left. Tympanometry examination result showed As type on both ears. Patient also had Eustachian tube canal dysfunction with no stapedial reflex in both ears.

Pure tone audiometry (PTA) was performed to the patient with result a mild conductive hearing loss (CHL) 30 dB with a Carhart notch in 2000Hz on the right ear; and a moderate CHL 53.75 dB with high frequency sensorineural hearing loss (SNHL) in 8000 Hz on the left ear. Patient also had undergone a speech audiometry procedure with result for the right ear, a speech recognition threshold (SRT) 50% in 43dB and speech discrimination score (SDS) 100% in 70 dB. On the left ear it was found that the SRT 50% was in 62 dB and SDS 100% in 80 dB. (Fig. 1)

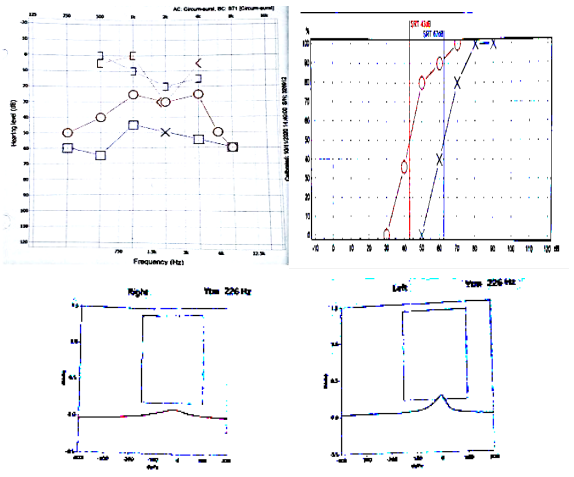

Figure 1. PTA, speech audiometry and tympanometry result before operation

The CT scan examination showed a lytic lesion on the right fissula ante fenestram, and on the left ear showed a chronic mastoiditis with a lytic lesion on the left fissula ante fenestram. (Fig. 2)

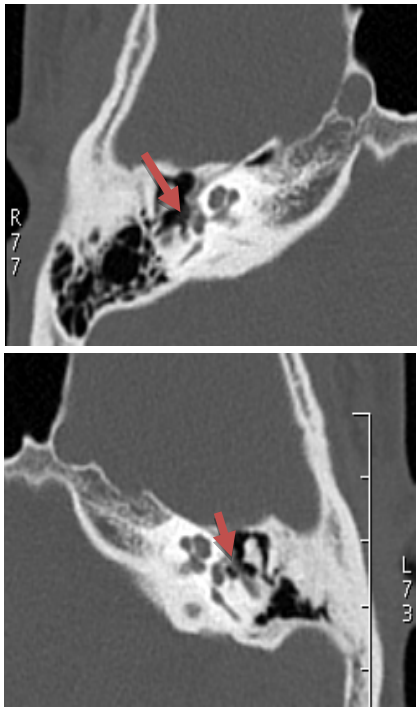

Figure 2. Red arrow shows a hypodense area in fissula ante fenestra

The patient was diagnosed with suspected bilateral otosclerosis. The plan was to perform a stapes surgery and insertion of prosthesis on the left ear. Surgery on the right ear would be performed around 3 months afterwards.

Transcanal endoscopic stapes surgery of the left ear was executed on May 5th, 2021. Patient was in supine position on the operating table under general anesthesia. The left ear was evaluated using endoscopy. Infiltration of ear canal using adrenaline 1:100,000. Circular incision was performed using a round knife 4-5 mm lateral to tympanic annulus from 12 to 6 o'clock. First, undermining the tympano-meatal flap using a round knife to the lower layer of the tympanic annulus, and then identifying the chorda tympany and malleus. The attachment of the chorda to the tympanic membrane was released. The flap was put back to original position, and the bleeding was controlled with adrenaline cotton pack.

Stapes fixation test was performed by moving the head of the stapes and it was found firmly fixated. Next, the incudostapedial

tendon was separated using 90-degree needle. Following the procedure, the stapedial tendon was cut using Belluci scissor. The stapes superstructure was then down-fractured into inferior direction so that the stapes footplate could be visualized. After that, scutum removal was performed using microdrill for better stapes footplate visualization. The footplate was found thick without any bluish appearance.

The distance between the stapes footplate and the medial surface of the *processus longus incus* was measured. Piston chausse was used as a prosthesis. Its length was cut to size 3.5 mm as necessitate. At first, an attempt to perform stapedotomy was utilizing a perforator to make a small hole, but it failed because the footplate was thick and very hard. Finally, stapedotomy was successfully performed using diamond burr 0.6 mm at 5000 rpm. The prosthesis was then placed in the stapedotomy hole and hooked to the *processus longus incus*. (Fig.3) The movement of the prosthesis was checked, but there was no light reflex movement on the round window. Then the oval window area was covered with about 1 ml of patient's venous blood, and the tympano-meatal flap was replaced. The ear canal was packed using gel and unabsorbable foam without antibiotic. After surgery the patient was educated not to blow his nose, no straining, and no lifting heavy objects.

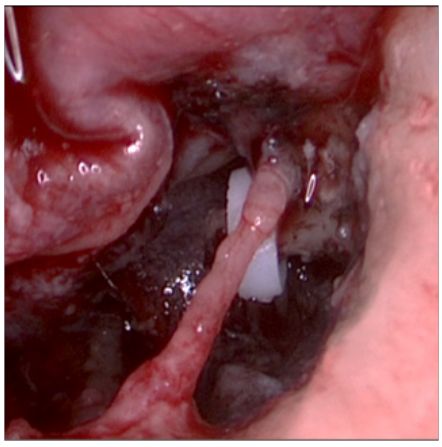

Figure 3. Piston placement in the stapedotomy hole

On the 1st day post-surgery, tuning fork examination showed lateralization towards the operated ear. The patient did not suffer vertigo nor taste disturbance. One week post surgery, patient came to Otology outpatient clinic. Patient had no complaint and the unabsorbable foam was than removed from the ear canal. Afterwards, the patient underwent whisper test and said that his hearing was better than before surgery.

Three weeks post-surgery, the patient came to Otology clinic without any complaint and underwent hearing examination. The result showed an improvement in PTA result to mild CHL (37.5 dB) with an SNHL component at 8000 Hz on the left ear; and moderate degree CHL on the right ear (41.25 dB) with an SNHL component at 8000 Hz. Speech audiometry was performed with SRT 50% reached at 45 dB while SDS 100% reached at 60 dB on both ears. (Fig. 4)

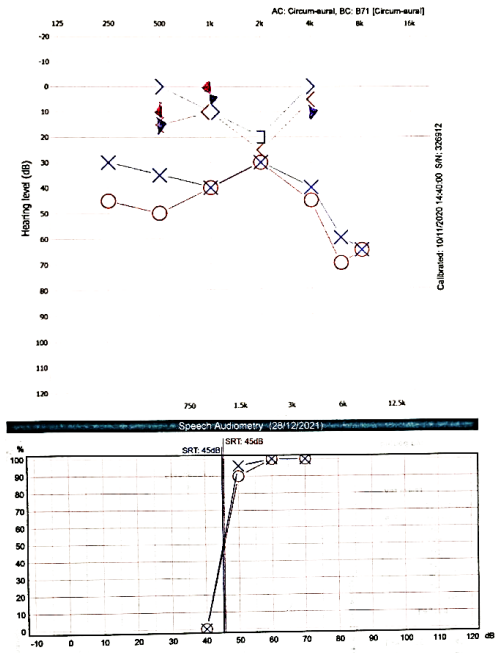

Figure 4. PTA and speech audiometry 3 weeks post-surgery

Four weeks post-surgery, PTA test was again performed with result a mild CHL on the left ear (27.5dB) with a mean air bone gap (ABG) 16.25 dB on the left ear. (Fig.5)

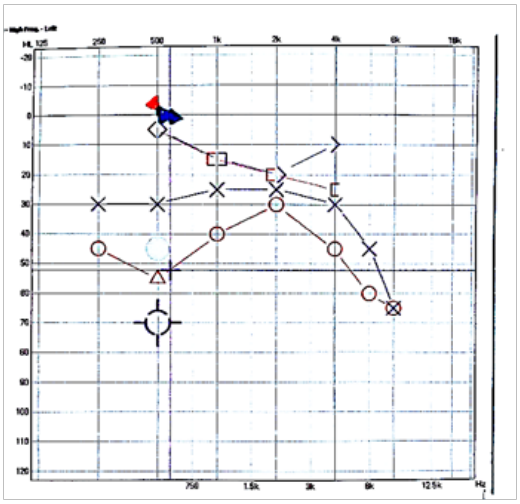

Figure 5. PTA 4 weeks after surgery

CLINICAL QUESTION

Based on the case presented above, we wanted to identify the management options in patients with obliterative otosclerosis with the following clinical question: In adults with obliterative otosclerosis, does stapes surgery improve hearing function and reduce complication?

REVIEW METHOD

The literature searching was conducted through PubMed, EBSCOhost, Scopus, and Proquest used keywords (otosclerosis) OR (otosclerosis obliterative) OR (obliterated footplate) OR (advance otosclerosis) AND (stapedotomy) OR (stapedectomy) OR (microsurgery) OR (stapes surgery).

The selection of articles was made by using inclusion criteria as follows: **1)** systematic review/meta-analysis of randomized control trials (RCTs), cohort or case control study**,** primary studies of RCT, cohort or case control; **2)** study population were adult patients with obliterative otosclerosis; **3)** intervention by stapes surgery; **4)** the outcome was hearing improvement and reduced complication.

Non-English articles were excluded.

RESULT

The literature searching used eligibility criteria based on keywords from clinical questions, found one article which was relevant to our clinical question.

DISCUSSION

Our patient was diagnosed with suspected bilateral otosclerosis from the history taking, PTA examination showed a Cahart notch, and CT scan showed a lytic lesion in bilateral fissula ante fenestram. The definitive diagnosis of obliterative otosclerosis was made intra-operative. A study by Genc et al.[5] found that a large air-bone gap in patients with conductive hearing loss might be a sign of diffuse obliterative otosclerosis.

In this case report, the patient had SNHL component in 8000 Hz from the PTA examination before surgery. During the surgery we found a total obliterated footplate and after we inserted the Teflon-piston prosthesis, we tried to test for piston mobilization but there was no sign of light reflex around the round window. This finding showed a probability of round window involvement. In our case, the patient perceived a better postoperative hearing outcomes at the lower frequencies. This might possibly be due to the fact that the higher frequencies are usually more affected by the disease process.[1,5]

According to Quaranta et al.[6], patient with obliterative otosclerosis had a better post-operative ABG and PTA than patient with peripheral rim or diffuse otosclerosis. In our case the mean ABG and PTA 4 weeks after operation were also showing a good result with a mean ABG of 16.25 dB compared to 45 dB before surgery, despite that there was no improvement in the hearing level at higher frequencies (8000Hz).

Other factors that might be an important determinant for post-operative hearing outcome was the size of prosthesis inserted

to the footplate. According to Quaranta et al.[4], there were no significant difference in terms of mean post-operative ABG and HFBC between 0.4 mm vs 0.6 mm piston diameter, but the 0.6 mm piston had a better ABG values at low frequencies. In our case, even we used a 0.6 mm diameter Teflon piston, it showed a good result in lower frequencies. Wegner et al.[7] found that larger diameter prosthesis will increase round window velocities and higher perilymphatic volume displacement. The post-operative hearing outcomes were also influenced by the pre-operative threshold, cochlear function, anatomical conditions, and also the experience of the surgeon.

Obliteration of round window has a sensorineural component while the obliteration of oval window has a conductive element. Hearing loss only occurs when round window niche is totally obliterated. Even a small opening is thought to be adequate for vibratory movement of the cochlear fluids between the round and oval windows. The obliterated round window should not be drilled out, as it might lead to profound SNHL. If a round window obliteration occurs, some surgeons consider a sodium fluoride therapy. Obliteration of round window has a worse prognosis of surgical hearing outcome compare to the oval window involvement.[3,7,8]

The management of obliterative otosclerosis is stapes surgery. From other two descriptive studies, the management of obliterative otosclerosis was large fenestra stapedotomy or total stapedectomy, whether using a microdrill or laser. This procedure was supposed to prevent mobilization of the temporalis muscle and re-obliteration of the vestibule niche.[8,9]

The administration of sodium fluoride was found to be useful in active phase to slow down the progression of obliteration footplate. Sodium fluoride can be given post operatively in patient with obliterative footplate. Stapedectomy or stapedotomy with large fenestra are the preferred technique for obliterative otosclerosis.[3]

Stapedotomy is a procedure to make a fenestration on the stapes footplate, while stapedectomy is a procedure that removes the entire stapes footplate. The stapedotomy procedure can be performed using a manual micro instrument (perforator), microdrill, and also laser. After the hole was made, prosthesis should would be placed inside the hole. In obliterative otosclerosis, a small fenestra usually could not be made using a manual perforator. The pathology that caused bone remodeling in the stapes footplate through oval window, makes it hard to perforate the stapes footplate.[3,9]

The microdrill is a sophisticated and safe instrument used for footplate fenestration during stapedotomy. The microdrill is a tiny electric motor with little diamond burrs ranging in size from 0.5 to 1 mm; it has low noise intensity and torque, making it a safe instrument for footplate fenestration when used for a few seconds without inflicting acoustic stress. Most surgeons choose the microdrill for a variety of reasons, including its versatility in all situations, such as obliterative otosclerosis or a narrow oval window. During the process, the rotatory action of the drill removes bone dust and prevents it from entering the vestibule.[10,11]

The microdrill may also be used to drill a thick stapes posterior crura. In obliterative otosclerosis, a large stapedotomy opening is the procedure of choice in these patients. The size of the opening should be large enough to prevent a lateral contact of the piston with the surrounding bone. Surgery requires saucerization of the bone in the oval window niche with a 1 mm diamond burr drill to convert the posterior part of the obliterative footplate to a blue lined thin shelf bone cover of the vestibule. Complete fenestration is then accomplished with a 0.7 mm diamond burr. At times, these thick footplates cannot be fractured and come out in one large piece, leaving a seemingly extremely wide oval window opening. In such

cases, the window is to be sealed completely with perichondrium and the prostheses placed over the perichondrium. Lasers have added a new dimension of safety and convenience to surgery on an obliterated footplate. There is a risk of reclosure if only a small channel is created in the foci on a large piece of bone, such as a small fenestra stapedectomy. [9,10]

From Ayache et al.[2] study, it was found that the comparison of the hearing results with stapedectomy (4 cases) or stapedotomy (4 cases) on the latest audiograms showed no statistically significant difference between the two procedures, using Chi-square and Fisher exact tests (p>0.5 for the air-bone gap closure and variations in threshold of the air-conduction at 8 kHz and bone-conduction at 4 kHz).

Amplifications by using hearing aid can be an alternative to surgery for patient with otosclerosis and should be asked to the patients as an option before surgery. Hearing aids are risk free compare to the stapes surgery. For very advance otosclerosis with cochlear involvement and profound SNHL, cochlear implant (CI) can give a benefit. Burmeister et al.[11] stated that the cochlear implant had proven to be effective in the treatment of patients with SNHL, including those with cochlear otosclerosis.

Until now there is no systematic review or RCT about the management of obliterative otosclerosis. This might be due to the low incidence of obliterative otosclerosis. In some literature, the preferred management for otosclerosis obliterative is large fenestra stapedotomy using microdrill or laser. The stapedotomy must be big enough to prevent the reclosure of the fenestra in the footplate. Administration of sodium fluoride may be beneficial to patient with otosclerosis obliterative after the surgery.

Large fenestra stapedotomy or stapedectomy is the preferred surgical technique for the management of obliterative otosclerosis.

Further research still needs to be implemented to study the best management for otosclerosis obliterative.

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