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Cochlear Implantation in a Patient with Advanced Otosclerosis

Kye Chun Park[1], Dae Bo Shim[1], In Seok Moon[2] and Mee Hyun Song[1]

[1]*Department of Otorhinolaryngology, Kwandong University College of Medicine, Myongji Hospital, Goyang; and*
[2]*Department of Otorhinolaryngology, Yonsei University College of Medicine, Seoul, Korea*

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관동대학교 의과대학 명지병원 이비인후과학교실,[1] 연세대학교 의과대학 이비인후과학교실[2]

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Address for correspondence
Mee Hyun Song, MD, PhD
Department of Otorhinolaryngology,
Kwandong University
College of Medicine,
Myongji Hospital,
55 Hwasu-ro 14beon-gil,
Deogyang-gu, Goyang 412-270,
Korea
Tel +82-31-810-5406
Fax +82-31-969-0500
E-mail meehyun924@hanmail.net

The incidence of otosclerosis, especially the retrofenestral advanced type, is relatively low in Koreans compared to that of the Western population. A case is reported in which cochlear implantation was performed in a patient with advanced otosclerosis presenting with mixed profound hearing loss on one side and pure sensorineural hearing loss on the other side. Intraoperative or postoperative complications of cochlear implantation commonly encountered in patients with otosclerosis did not occur in our patient and successful auditory outcome could be achieved. The results are reported with the review of literature, and clinical considerations regarding cochlear implantation in otosclerosis are discussed.
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Key Words Cochlear implantation · Hearing loss · Otosclerosis.

Introduction

Otosclerosis is a primary disorder of the bony labyrinth and stapes known to affect only humans, leading to progressive conductive and sensorineural hearing loss. The hearing loss in otosclerosis is most commonly presented as conductive or mixed type but sensorineural hearing loss can result from otosclerosis in approximately 10%.[1)] Otosclerosis with retrofenestral involvement can result in far-advanced otosclerosis (FAO) referred to conditions with air-conduction thresholds in excess of 85 dB and nonrecordable or fragmentary bone conduction, or very far-advanced otosclerosis (VFAO) meaning that both bone and air conduction thresholds are not measurable.[2,3)] Stapedectomy or stapedotomy with adjunctive hearing aid use has been performed in FAO and VFAO with favorable outcome in some cases, but the auditory outcome has been variable and limited.[4)] With the technical and surgical advancements, patients with retrofenestral otosclerosis showing severe to profound hearing loss are increasingly being treated with cochlear implantation (CI) which has shown successful outcome, although there are factors that need to be considered concerning surgical complications or postoperative programming.[5-7)]

Here, we report a case of CI performed in a patient with otosclerosis presenting with profound mixed hearing loss on one side and pure sensorineural hearing loss on the other side. The results are reported along with review of literature and discussion on factors that need consideration in performing CI in otosclerosis.

Case

A 52-year-old female visited our clinic complaining of tin-

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nitus and hearing impairment on both sides that had started several years ago. The hearing loss first began 8 years ago on the left side and the hearing on the right side started to deteriorate 6 years ago. She had no other medical disease or any family history of hearing loss. She had been working at a noisy sewing factory for 20 years and was using a hearing aid on the right ear for 6 years. On physical examination, the tympanic membrane was intact on both sides without any evidence of Schwartze sign. Pure tone audiometry demonstrated sensorineural hearing loss of 71 dB HL on the right side and profound mixed hearing loss on the left side (Fig. 1). The speech discrimination score was 32% on the right and 0% on the left. Since a large air-bone gap of 40 dB HL was detected on the left side, high resolution temporal bone CT was performed. On axial images, low density was identified at fissula ante fenestram and around the vestibule bilaterally, and double ring effect of the cochlea was seen on both sides (Fig. 2A). The patient was diagnosed with bilateral retrofenestral otosclerosis with fenestral involvement.[1)]

Considering that the patient gained limited benefit with hearing aids, cochlear implantation was performed. On preoperative speech evaluation, the aided open-set sentence score in auditory-only condition was 37%. The categories of auditory performances (CAP) scores were 1 under unaided condition and 4 using a hearing aid on the right side. The patient did not complain of dizziness or unsteadiness, and the bithermal caloric test revealed normal responses bilaterally while vestibular evoked myogenic potential could not be detected on the right side. Brain magnetic resonance imaging (MRI) was performed for the evaluation of cochlear nerve integrity and patency of the cochlea. The cochleovestibular nerve was normally identified within the internal auditory canal on the parasagittal view and there was no sign of obliteration of the cochlear turns on MRI (Fig. 2B and C).

Despite the use of hearing aid on the right side, the patient's hearing had worsened progressively and the patient had great difficulty in everyday communication even with lip-reading. Since the patient had been using hearing aids for 6 years on the right side exhibiting residual hearing, we decided to perform CI on the left side showing profound hearing loss despite longer duration of hearing loss. Intraoperatively, hypervascularization was observed on the promontory and around the round window niche. When cochleostomy was performed anteroinferior to the round window, the bone was very brittle. The patient was implanted with Nucleus CI 512 device with perimodiolar type electrode. Complete insertion of electrode was achieved without resistance and no perilymphatic gusher was encountered. Intraoperative neural response telemetry (NRT) revealed good response in all electrodes. On postop-

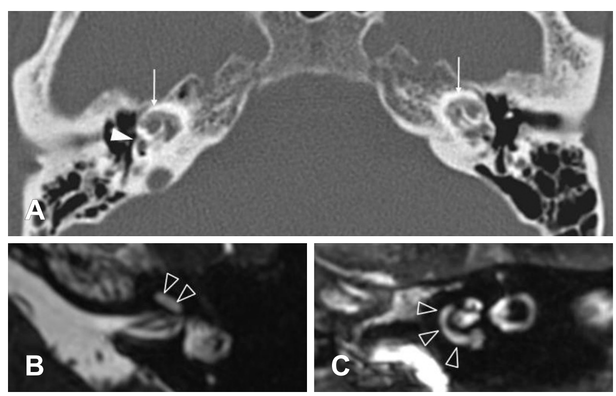

Fig. 2. Radiologic findings of the temporal bone. Axial images of temporal bone CT demonstrated double ring effect of the cochlea (white arrows) and hypodensity at fissula ante fenestram (white arrowhead) on both sides (A). On MRI, patency of the cochlear basal turn of the left side was identified on axial (empty arrowheads in B) and parasagittal (empty arrowheads in C) views (B and C).

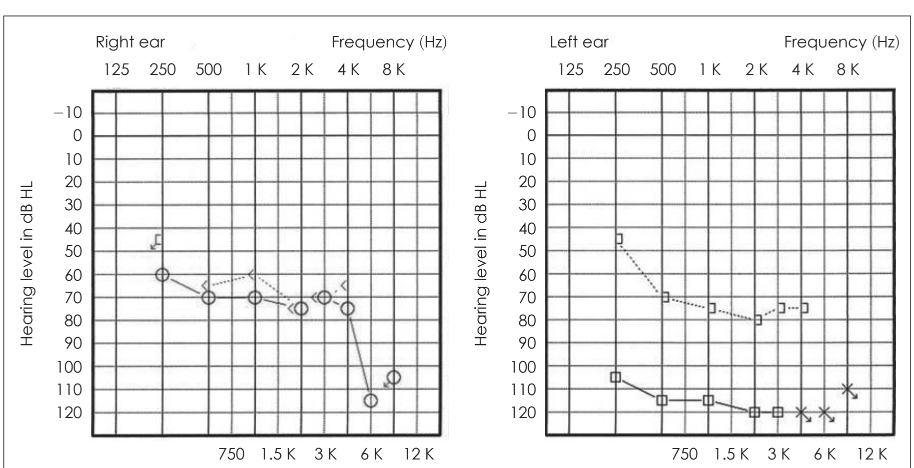

Fig. 1. Preoperative pure tone audiometry. Right side exhibited sensorineural hearing loss of 71 dB HL and left side showed profound mixed hearing loss.

erative transorbital X-ray, proper positioning of the electrode within the cochlea could be confirmed (Fig. 3). Pathologic analysis of a specimen taken from the bone near the round window niche using micro cup forceps demonstrated perivascular bone resorption and basophilic spongiosis as well as acidophilic sclerotic lesions with accumulation of otosclerotic bone (Fig. 4).

The device was switched-on at postoperative one month, and NRT demonstrated good responses in the basal, middle, and apical electrodes. No facial twitching was observed during electrical stimulation. The sound field test using the cochlear implant device demonstrated pure tone threshold of 20 dB HL 3 months after switch-on. On speech evaluation, monosyllabic and disyllable word discrimination scores reached 80% and 85%, respectively, 10 months after switch-on (Fig. 5). The open-set sentence score improved from 49% preoperatively to 86% 5 months after switch-on and this improvement has been maintained until the last evaluation performed 16 months after switch-on (Fig. 5). The auditory performance improved dramatically to reach CAP 7, 10 months after switch-on and telephone conversation was possible using the cochlear implant alone.

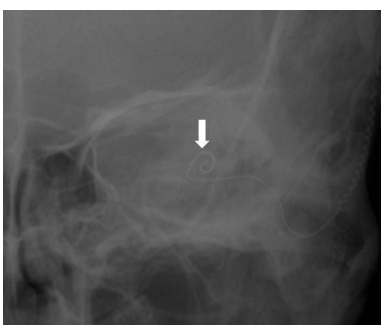

Fig. 3. Postoperative transorbital X-ray. Imaging performed immediately after cochlear implantation revealed a fully inserted electrode (white arrow) in the cochlea on the left side.

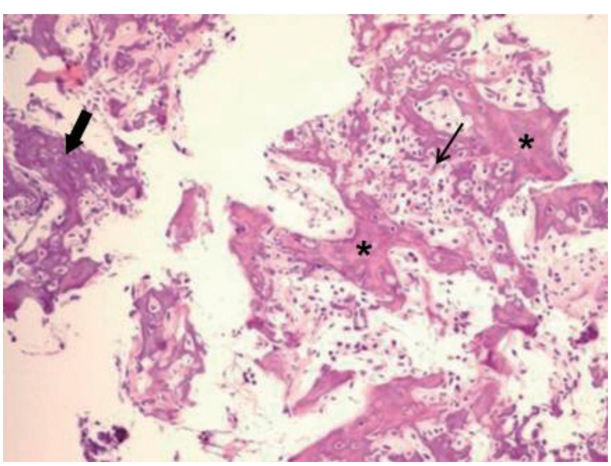

Fig. 4. Pathologic findings of otosclerotic bone (H&E staining, ×40). Early active otospongiotic regions with basophilic staining were seen (thick arrow). Hypervascularization and resorption of bone with enlargement of perivascular space and spongiosis were also observed in the same specimen (thin arrow). Other regions demonstrated acidophilic sclerotic lesions and accumulation of otosclerotic bone distorting the striation of collagen (asterisks).

Discussion

This report presents a case of successful CI without any intraoperative or postoperative complications performed in a patient with advanced otosclerosis. Auditory rehabilitation in otosclerosis can be achieved by various approaches including hearing aids, stapes surgery, or CI. Stapes surgery and hearing aid amplification might be initially considered in patients who still have residual hearing and measurable bone conduction since stapedotomy is a safe, simple, and low-cost procedure. Nonetheless, the hearing outcome of stapes surgery can be unpredictable in advanced otosclerosis and deterioration of hearing may occur with time.[4,8] Calmels, et al.[8] reported significantly better speech performance in patients with FAO who underwent CI compared to those receiving stapedotomy alone, and there are many studies reporting successful auditory and surgical outcome of CI in otosclerosis.[1,5,6,9] The patient in this report had been using a hearing aid on the right side showing severe sensorineural hearing loss which had worsened progressively to the level that the patient experienced communication difficulty in everyday life. Although there was measurable bone conduction hearing on the left side, CI on the left side was decided rather than trial of stapes surgery consider-

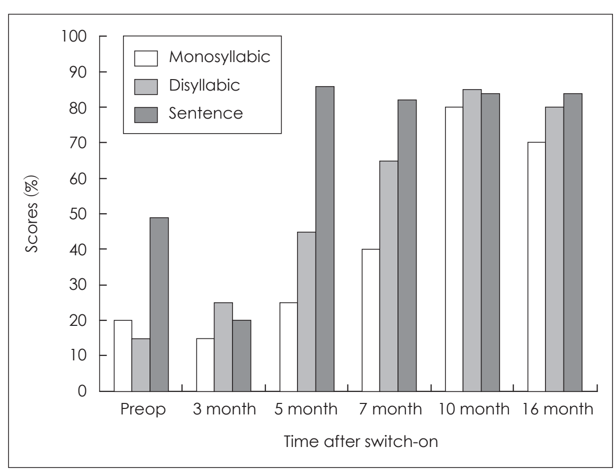

Fig. 5. Postoperative speech performance after cochlear implantation. Monosyllabic and disyllabic word scores gradually improved up to 10 months after switch-on and the open-set sentence scores showed a significant improvement in a shorter period following switch-on.

ing the decreased benefit of hearing aids on the right side and the profound air conduction level with very low speech discrimination score on the left side. As reported by Matterson, et al.,[9] the longer duration of deafness on the left side, which may have a negative effect on CI outcome, did not seem to influence postoperative auditory performance in our patient.

There are several complicating factors that need consideration in performing CI in otosclerosis. There is a greater chance of partial insertion or misplacement of the electrode array because of the severely thinned and distorted otic capsule in advanced stages of otosclerosis.[1] Also, facial nerve stimulation that may require deactivation of the electrodes or revision surgery possibly leading to deterioration of auditory performance, is reported to occur in up to 50% of patients with otosclerosis receiving CI.[1,6,10] The higher incidence of facial nerve stimulation after CI in otosclerosis is thought to be related to the increased conductivity of electrical current through the soft, remodeled bone especially in the region where the labyrinthine segment of the facial nerve crosses the superior segment of the basal turn of the cochlea.[11] Considering the increased risk of facial nerve stimulation in otosclerosis, a perimodiolar electrode was used in our patient since its contacts face toward the modiolus and the position of the electrode is more distant from the outer wall of the cochlea releasing less current flow through the otic capsule.[5,9] Another surgical factor to be considered in otosclerosis is the obstruction or narrowing of the cochlear lumen due to new bone formation.[12] We had confirmed the patency of the cochlear basal turn on temporal bone CT and brain MRI preoperatively and full insertion of the electrode was achieved in our patient. The electrical thresholds and maximal comfort levels have been reported to increase with time after CI in otosclerotic patients, but this was not seen in our patient during the postoperative programming sessions.[7]

Both conductive and sensorineural component of hearing loss can result from otosclerosis, but pure sensorineural hearing loss, as seen in this case, is not a typical finding. Sensorineural hearing loss in otosclerosis can be caused by the lytic enzymes released from the otosclerotic foci leading to the degeneration of the organ of Corti, and has also been related to spiral ligament hyalinization and stria vascularis atrophy.[13] In our case, although the degree and type of hearing loss differed considerably in each ear, the radiologic findings were almost similar in both ears suggesting that local factors other than the disease severity demonstrated on imaging could influence the auditory manifestations in otosclerosis.

The incidence of otosclerosis, especially retrofenestral advanced otosclerosis, is relatively low in the Korean population compared to the western population. Only two case reports of CI performed in otosclerosis have been reported to date.[14,15] It seems that CI is an excellent choice for auditory rehabilitation in patients with severe to profound hearing loss caused by otosclerosis. Our case adds to the experience of CI in Koreans with otosclerosis and the successful outcome presented here is expected to provide valuable information in the clinical management of patients with otosclerosis in Korea.

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