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Case Report

Novel Use of Depth Gauge to Keep Cochlear Lumen Patent in Bilateral Sequential Cochlear Implantation

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Abstract

Cochlear otosclerosis is a progressive disease with otosclerotic foci in the cochlea making cochlear implantation (CI) a challenging task. Early implantation is necessary to achieve a favorable outcome. In our institute bilateral simultaneous CI is not permitted. In the case, we present a novel use of depth gauge as a spacer device to maintain the patency of cochlear lumen awaiting staged CI. Complete insertion of the electrode was achieved bilaterally with improvement in audition and speech perception. Depth gauge can be used as a spacer device for keeping cochlear lumen patent in patients with labyrinthitis ossificans where CI is delayed because of institutional policies or financial considerations to achieve complete insertion and good auditory outcomes.

Keywords: Cochlear Implantation; Cochlear Otosclerosis; Neural Response Telemetry (NRT)

Introduction

The role of cochlear implantation has expanded in recent times and is now has an established indication in cochlear malformations, cochleovestibular nerve deficiency including labyrinthine ossificans [1].

Otosclerosis is a process of continuous osteolysis and osteogenesis of otic and labyrinthine capsules. Once started it causes hearing loss which usually worsens over a period of time. Cochlear otosclerosis is a phenomenon whereby otosclerotic focus is in the otic capsule and affects cochlear endosteum causing mixed-type or sensorineural hearing loss [2]. House and Sheehy in 1961 defined advanced otosclerosis as hearing loss in air conduction (AC) threshold by 85dB with non-measurable bone conduction (BC) [3].

Cochlear otosclerosis is one of the causes for progressive labyrinthitis ossificans other than post-trauma and post meningitis. In cases of cochlear otosclerosis, radiographical imaging is by far the standard of choice to confirm the diagnosis [4].

Cochlear implantation in such cases becomes a challenge as ossification may impede electrode insertion and achieving ideal outcomes. Early implantation is therefore necessary before the lumen becomes completely obliterated [5].

We here report a case of bilateral far advanced otosclerosis who underwent sequential bilateral cochlear implantation with novel use of depth gauge to keep cochlear lumen patent while the patient was awaiting second side surgery.

Case Presentation

A male in mid 50's of Indian origin presented to our hospital with complaints of insidious onset gradually progressive hearing

loss in both ears for past 10 years. He had been using bilateral digital hearing aids for last 7 years. He had been having little or no benefit with hearing aids for the last 1 year and had started relying on lip reading to have conversation. He also had complaints of intermittent tinnitus. There was no history of otorrhoea, otalgia, vertigo, facial asymmetry. No history of trauma, exposure to loud noise or ototoxic medication. He had no family history of hearing loss.

General and otoscopic examination was normal. There was no evidence of Schwartze sign. Tuning fork examination was suggestive of bilateral sensorineural hearing loss (SNHL).

Based on clinical features, audiological workup and radiological findings the patient was diagnosed to have Bilateral Far Advanced Otosclerosis with Profound SNHL and poor SDS and Roteeveel grade 2C ossificans. Since the cochlear implantation programme at our centre is government aided and bilateral simultaneous cochlear implantation is not supported under it, patient was counselled to undergo sequential bilateral cochlear implantation. Since the sanction for second side sequential implantation could take upto 6 months, patient was planned for primary cochlear implant surgery on left due to better cochlear lumen patency and sequential cochlear implantation on right. Patient underwent left cochlear implantation with Cochlear nucleus CI 24RE (ST) with straight electrode array via transmastoid facial recess approach. Intraoperatively it was observed that round window niche was completely obliterated. An anterior cochleostomy was done. The scala tympani of basal turn was completely obliterated however, luminal patency and perilymph was visualized in the scala vestibuli. Perilymphatic oozer was encountered and was managed with hyperventilation, steroids and tissue seal around electrode. Complete insertion of electrode was achieved without resistance. Intraoperative impedance and Neural Response Telemetry (NRT) showed good response in all of the 22 electrodes. Following primary closure of wound and sterile dressing of left side, right side was approached. The intraoperative finding was similar on right side (Figure 4a and 4b). An anterior cochleostomy was done and patent lumen of scala vestibule was identified with perilymph. Complete insertion of test depth gauge (CochlearTM Nucleus® straight with basal diameter of 0.6 mm and apical diameter of 0.4mm respectively) was achieved with minimal resistance (Figure 5a and 5b). It was secured under periosteal flap and left in situ as a cochlear lumen spacer and wound was closed in layers.

Post op X ray modified Stenvers view done on post op day 2 revealed all electrodes to be intracochlear. However there was a minimal kinking of depth guaze on right side as evident on the Xray (Figure 6). Cone beam CT further confirmed the intracochlear placement of electrodes of the implant and the test electrodes. The device was switched-on at postoperative day 14 and NRT demonstrated good responses in the basal, middle, and apical electrodes.

Facial twitching was observed during electrical stimulation on electrodes no.1,2,3,20,21,22. Facial nerve stimulation was managed by reprogramming and manual deactivation of the electrodes.

In the post operative period speech therapy was given and there was progressive improvement in audition and speech understanding. 3 monthly evaluation of speech and aided audiometry was done which was within speech banana.

After approval for second side sequential CI, patient was planned for a second side (right side) sequential cochlear implantation 6 months after first surgery. Intra operatively the depth guaze was found in situ and lumen of scala vestibule was patent with presence of perilymph. The depth gauge was replaced with complete insertion of Cochlear nucleus CI 24 RE (ST) straight electrode array without any resistance. Perilymphatic oozer was encountered and was managed with hyperventilation, steroids and tissue seal around electrode. Intraoperative impedance and Neural Response Telemetry (NRT) showed good response in all of the 22 electrodes. Post op X ray modified Stenvers view done on post op day 2 revealed all electrodes to be intracochlear with no kinking or tip roll over (Figure 7). The device was switched-on at postoperative day 14 and good response was obtained in all electrodes. Facial nerve stimulation was observed during switch on of electrodes no. 6,11,12,13,14,15,19 in the form of facial spasms. Facial nerve stimulation was managed by reprogramming and manual deactivation of the electrodes.

In the follow up period, patient was given continuous speech therapy and post operative speech and audiological evaluation was done after 3 months which revealed a good response in both ears.

Investigations

Pure tone audiometry (PTA) demonstrated bilateral profound SNHL (Figure 1). Aided audiometry suggested no benefit with

hearing aids. The speech discrimination score (SDS) (unaided and aided) was 0% in both ears.

Figure 1: Pure tone audiometry (PTA) demonstrates bilateral profound SNHL.

High resolution computed tomography (HRCT) temporal bone revealed bilateral fenestral and retrofenestral otosclerotic foci with double ring effect of the cochlea (Halo sign). There was bilateral narrowing of basal turn of cochlea by otosclerotic foci suggestive of partial ossification (Rotteveel Grade 2C) (Figure 2).

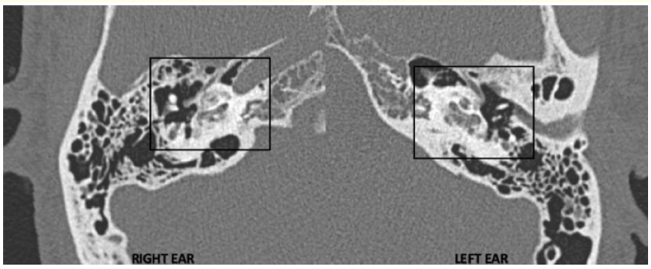

Figure 2: High resolution computed tomography (HRCT) temporal bone reveals bilateral Halo sign. (Rotteveel Grade 2C).

Magnetic Resonance Imaging (MRI) of brain and inner ear was performed for the evaluation of patency of the cochlea. Bilateral presence of cochlear nerve confirmed. Three-dimensional (3D) constructive interference in steady state images showed there was bilateral obliteration of scala tympani of basal turn of cochlea more on right as compared to left with scala vestibuli patent bilaterally (Figure 3).

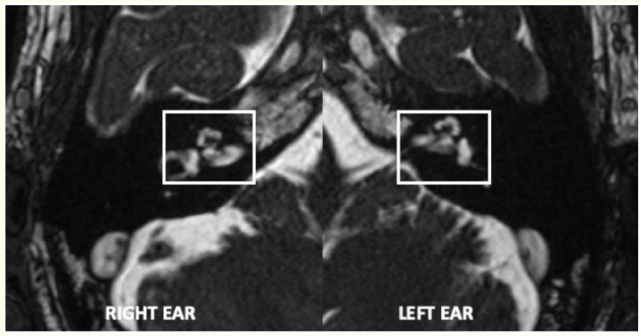

Figure 3: MRI images shows bilateral obliteration of scala tympani of basal turn of cochlea more on right as compared to left with scala vestibuli patent bilaterally.

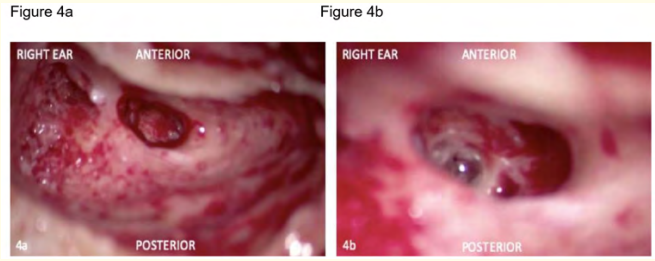

Figure 4: a: Intraoperative image shows obliteration of Round window niche. b. Intraoperative image shows cochleostomy with perilymphatic ooze.

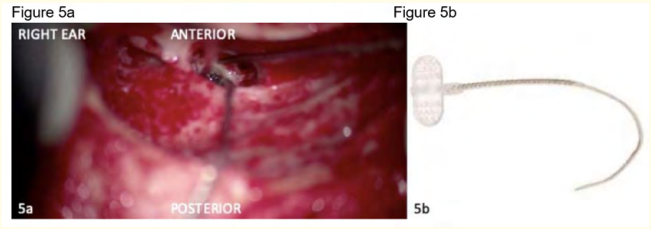

Figure 5: a. Intraoperative image shows insertion of depth guaze. b. CochlearTM Nucleus® depth guaze.e.

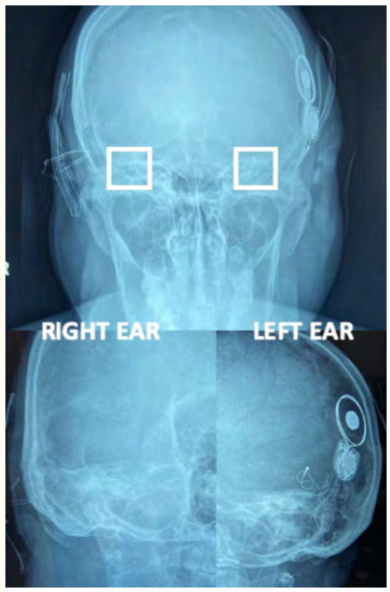

Figure 6: X ray modified Stenvers view shows all electrodes to be intracochlear with minimal kinking of depth guaze on right side.

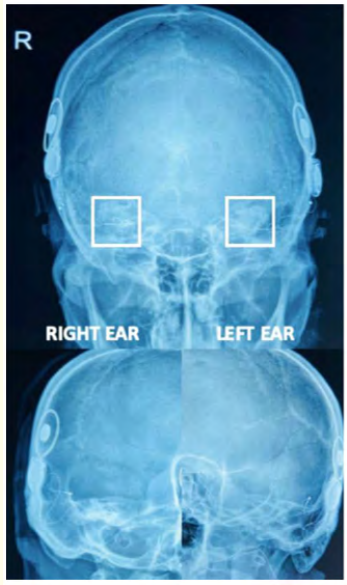

Figure 7: X ray modified Stenvers shows all electrodes to be intracochlear with no kinking or tip roll over.

Outcome and follow-up

The post operative period was uneventful. Patient was followed up for 3 months for mapping and monitoring outcomes. There was improvement in the auditory performance and speech discrimination. Patient was able to do conversation without lip reading using the cochlear implant by 3 months.

Post bilateral CI, patient reached a peak CAP score of 7 within 3 months of implantation. Monosyllabic and disyllable word discrimination scores reached 100% and 80% respectively at 3 months post switch-on of CI. Aided audiometry of the patient was found near normal (Figure 8).

Discussion

Far advanced otosclerosis or cochlear otosclerosis is presence of otosclerotic foci in the cochlear endosteum causing obliteration of the cochlear lumen and SNHL. This process is gradually progressive in nature and demands early intervention [1,2].

Cochlear implantation in cases of ossification is not only surgically challenging but also imposes chances of complications

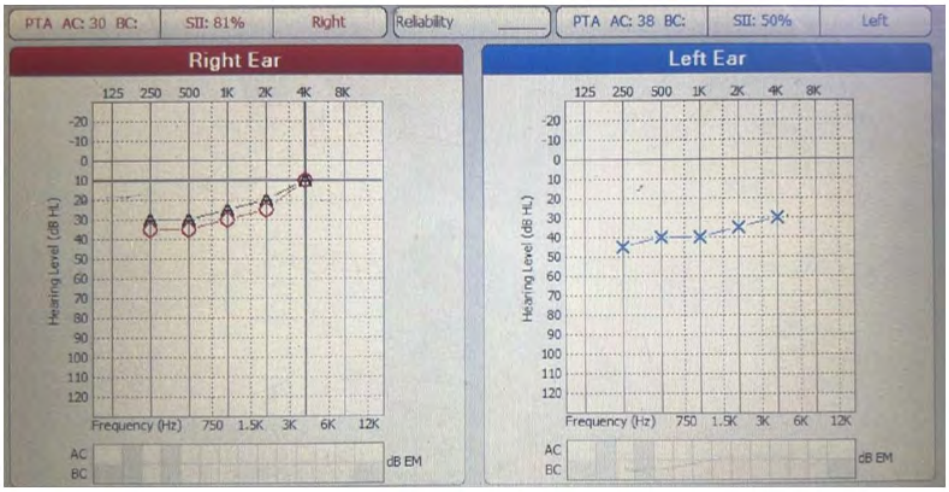

Figure 8: Aided audiometry post bilateral cochlear implantation of the patient shows bilateral hearing within normal threshold.

like facial nerve stimulation. Patients post implantation may have limited auditory benefits in these cases [1]. Labyrinthitis ossificans can occur in cases of meningitis (within 72hrs to 04 weeks), trauma and cochlear otosclerosis which causes progressive narrowing of lumen until complete obliteration occurs [2,5]. Both our cases were of progressive cochlear otosclerosis presenting with hearing loss not benefitting with hearing aids.

Evaluation of the patient requires thorough otologic, audiologic and radiographic investigations. Merkus., *et al*. had published an evidence based article on decision making in advanced otosclerosis where patient is considered for hearing aid, stapedotomy, cochlear implantation based on audiological findings and grade of labyrinthine ossificans based on HRCT temporal bone [3,6]. Rotteveel and colleagues developed a grading system for extent of labyrinthine ossificans based on HRCT of temporal bone which aids the surgeon in planning cochlear implantation [4,7]. In both the present reported cases, the indication of cochlear implantation was profound hearing loss with poor SDS on audiological evaluation and Rotteveel grade 2C labyrinthine ossificans.

Depth guage is used routinely and a standard technique at CI centres in cases of labyrinthitis ossificans to determine cochlear lumen patency and depth of luminal patency intraoperatively including in cochlear otosclerosis [4]. To the best of our knowledge this is the first reported use of depth gauge as a cochlear lumen spacer device to keep it patent in patient with far advanced otosclerosis with labyrinthine ossificans awaiting cochlear impantation.

Conclusion

- Depth guage has been used as a spacer device while waiting for sequential cochlear implantation.
- This technique is unique and beneficial when CI is delayed because of institutional policies or financial considerations to achieve complete insertion and good auditory outcomes.
- Post-operative good auditory outcomes suggest that this technique is a boon for far advanced otosclerosis.

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