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Case Report

Cochlear Implantation in Advanced Otosclerosis: Rising to the Challenges of Surgery and Decision making

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Abstract:

Introduction: *Cochlear implantation as well as stapedotomy followed by use of hearing aid are acceptable modes of surgically rehabilitating patients with far advanced otosclerosis. Surgical challenges of CI include difficulties associated with electrode insertion and facial nerve stimulation. Improvement in speech discrimination scores and overall satisfaction with stapedotomy and hearing aid use are reportedly poor in many patients, yet being a low cost procedure it may be used as initial management in a subset of patients.*

Case Report: *46 year old patient with diffuse confluent retrofenestral otoscerosis underwent cochlear implantation. He was mapped using behavioral thresholds as despite intracochlear electrode position no neural response was recordable per-operatively as well as in the post-operative period. Perimodiolar electrodes and sodium flouride therapy were used to overcome problems of FNS. 18 months post CI the patient has good audiologic outcomes (CAP 7) without any FNS.*

Conclusion*: High resolution computed tomography, air bone gap and speech discrimination scores are important in formulating treatment plan in patients with far advanced otosclerosis. Early cochlear implantation can be considered in patients with poor speech discrimination scores and extensive cochlear lesions. Facial nerve stimulation can be prevented by adequate pre-operative planning.*

Keywords*: otosclerosis; hearing loss, sensorineural; cochlear implantation; stapes surgery*

Introduction:

Otosclerosis (OS) is a hereditary disease in which the avascular enchondral bone of the otic capsule gets replaced by vascular spongy bone along with deposition of immature bone which lacks collagen. The area most commonly involved lies anterior to stapes footplate (fenestral otosclerosis) resulting in fixation of the footplate and clinically manifests as conductive hearing loss (CHL). Lesions involving the dense otic capsule (retrofenestral otosclerosis) can result in sensorineural hearing loss (SNHL)1. Far advanced otosclerosis (FAO) has been described as a condition where the air conduction (AC) thresholds are 85dB or above. In these patients the bone conduction (BC) thresholds are not measurable due to the limitation of clinical audiometers[2]. Speech discrimination scores (SDS) are markedly reduced in these patients.

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The treatment options that can be offered to patients with FAO include hearing aids, stapedotomy for closure of air bone gap (ABG) followed by use of hearing aids or cochlear implantation (CI)[3,4] Hearing aids do little to improve the SDS. Also both the surgical interventions can have unpredictable results. In the absence of guidelines for managing advanced OS, the surgeon often faces a dilemma in choosing the best treatment modality for optimum hearing outcome in these patients. We report a case of FAO with diffuse confluent retrofenestral otospongiotic lesions who underwent CI at our centre. The decision making, surgical challenges and the outcomes of CI are discussed.

Case report:

A 46 year old male presented with history of progressive hearing loss of 14 years duration. Early in the course of his disease he had been diagnosed to have OS with mixed hearing loss. He had not undergone any surgery for correction of air bone gap (ABG). He was a regular hearing aid user with good benefit but off late had developed inability to communicate at his workplace and in social gatherings resulting in loss of self-esteem. Being an office employee he also faced the risk of losing his profession and was thus concerned about his auditory rehabilitation. Pure tone audiometry (PTA) showed air conduction (AC) thresholds at 95-115dBHL on (R) and 110-120dBHL on (L) side. Bone conduction (BC) thresholds were 45 and 65 dBHL at 250Hz and 500Hz on (R) side. No BC thresholds were recordable at any other frequencies on either side. Otoacoustic emissions (OAE) were absent bilaterally and no auditory brain stem potentials were recordable with maximum stimulation. Rest of ENT evaluation was within normal limits. Aided responses with high gain hearing aids were recorded at 250, 500Hz on (R) side only. The aided speech discrimination scores (SDS) were 12% and 8% for (R) and (L) side respectively. High resolution CT (HRCT) scan of temporal bone showed bilateral Rotteveel grade 3 lesions (diffuse retro-fenestral confluent lesions) (Fig 1).

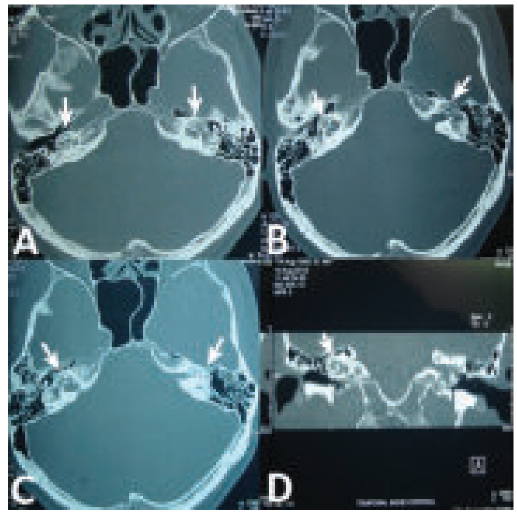

Figure 1: High resolution computed tomogram showing bilateral diffuse confluent spongiotic lesions in cochlea (arrows), in axial section (A, B & C), and in coronal section (D).

Considering his poor SDS with high gain hearing aids he was considered as a potential candidate for CI with caution considering the confluent otospongiotic lesions. Prior to surgery he underwent magnetic resonance imaging (MRI) in which reduced cochlear fluid signal of the basal turn could be identified bilaterally. There was no medical contraindication for surgery and his neuro-psychological assessment was normal. He and his family were counseled about the possible risks, expected outcomes, the need for frequent mapping sessions and auditory therapy in the post-operative period. In particular he was counseled about the possibility of incomplete electrode insertion, misplacement of electrode and the anticipated high stimulus levels required for auditory stimulation which could result in facial nerve stimulation (FNS). Three months prior to surgery the patient was started on flouride therapy along with calcium and vitamin D3 supplementation.

He underwent (R) CI under general anesthesia using the standard transmastoid facial recess approach. Extended RW cochleostomy was done. Soft granular bone was found in the initial part of the lumen of basal turn which was removed with a fine pick. Complete insertion of Nucleus freedom contour advance electrode (CI24RE(CA)) could be achieved. Electrode impedances were within normal limits. On neural response telemetry (NRT) no evoked compound action potentials (ECAP) were recorded in any of the electrodes per-operatively. X ray modified Stenvers projection done in post-operative period confirmed the intracochlear electrode position (Fig 2). The patient was discharged on the third post-operative day on oral antibiotics.

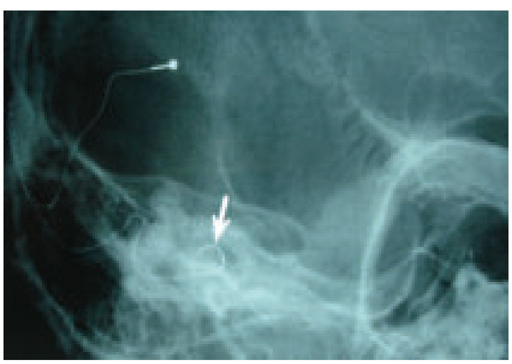

Figure 2: *X Ray (modified Stenver's projection) showing intracochlear electrode position (arrow).*

The implant switch on with processor CP 920 was done after four weeks using Nucleus fitting software. Behavioral threshold (T) and comfortable (C) levels were obtained at all the electrodes (Fig 3) using default parameters.

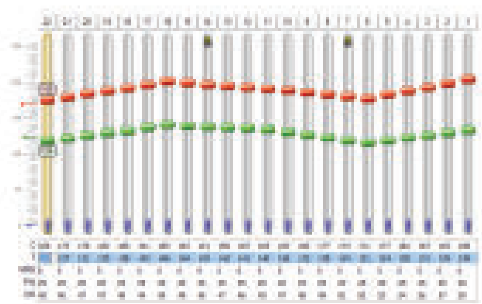

Figure 3: *Map of patient one year after cochlear implantation showing threshold and comfort levels.*

There was no non auditory stimulation in the form of facial twitch. In subsequent mappings there has been no requirement to decrease the stimulus levels or switch off the active electrodes. The patient continues to have good auditory perception with good dynamic range. He has been on follow up for last 18 months and continues to be on flouride therapy, however till date we have been unable to obtain NRT by both manual and auto mode. His SDS in quiet are 94% and category of auditory performance (CAP) score is seven. He is able to converse on telephone and feels socially and professionally rehabilitated.

Discussion:

Retrofenestral lesions have been reported in 10% patients with otosclerosis and can result in SNHL[1]. The enzymes released by the focus are toxic to Organ of Corti, also atrophy of stria vascularis and hyalinization of the spiral ligament, hair cell and ganglion cell loss contribute to the hearing loss. Hearing aids alone are not well accepted in patients with FAO. The surgical options for hearing rehablitation include stapedotomy and CI.

HRCT scan is of immense value in identifying these osteolytic lesions preoperatively. The lesions can be classified based on their location and type using Rotteveel's radiologic classification as: fenestral (Grade 1), retrofenestral double ring or halo effect in cochlea (Grade 2A), narrow basal turn (Grade 2B), Grade 2C when both are present, Grade 3 when diffuse confluent retrofenestral lesions are present[1]. Thus a fair estimation of the surgical difficulties and outcomes can be made and the patient counseled accordingly.

Stapedotomy corrects the ABG making the hearing thresholds aidable but has little effect on the poor discrimination scores which these patients have. One of the most feared complication of stapedotomy in extensive OS is increase in SNHL resulting in a dead ear[5].. CI offers a very good alternative surgical option in FAO. Excellent auditory outcomes with CI have been reported by various authors[6,7].

Coexisting osteogenetic and osteolytic lesions make CI surgery challenging in FAO. Ossification of the RW and scala tympani has been reported and may require drilling for identification of a patent lumen [1, 5, 8]. Placing an electrode in presence of ossification is difficult though not contraindicated [9]. Partial electrode insertion has been reported in presence of ossification but audiologic outcomes in cases where complete insertion has been achieved are not compromised [5,8]. Otospongiosis can also result in formation of a false path in the cochlea resulting in misplacement of electrode into the osteolytic cavity[5]. FNS is a known complication and has been reported in around 14 -38% cases [1, 10]. Spongiotic bone lesions may increase the conductivity leading to FNS at the initial switch on itself [1,6,11]. Erosion of the thin lateral cochlear wall due to physical pressure by the straight array can also directly stimulate the facial nerve[10]. The incidence is more in cases of grade 3 lesions and with the use of non modiolar hugging electrodes[12]. Natural progression of disease may necessitate the current levels to be increased for auditory perception resulting in FNS during subsequent programming sessions.

FNS can be managed by lowering the current amplitudes, switching off the offending electrodes or changing the programming strategies[13,14]. The electrodes in the superior part of basal turn (mid array contacts) lie in close proximity to labyrinthine and meatal segment of facial nerve and may cause stimulation if the density of intervening bone is reduced[11]. Switching off these electrodes may affect the implant performance and compromise the audiologic outcomes. Other strategies to manage FNS include use of perimodiolar electrodes and flouride therapy [13,15]. Troublesome FNS may require reimplantation into scala vestibuli which is further away from facial nerve or explantation[10]. Intractable tinnitus due to increased current requirement may also necessitate explantation in these patients[7].

Better results and greater overall satisfaction has been reported with CI when compared to stapedotomy and use of hearing aids [3,5]. However, measurable BC thresholds and improvement in SDS have also been reported with stapedotomy [3,9]. Thus stapedotomy being a relatively low cost procedure may still be considered as an initial management in FAO in some patients. Calmels et al have reported their preference for initial stapedotomy in FAO cases[3]. In their protocol CI is reserved only for those patients in whom there is no improvement in thresholds or SDS three months post stapedotomy or when the satisfaction levels are low.

Merkus et al have proposed an algorithm for effective management of patients with FAO. According to them SDS, HRCT findings and ABG should guide the decision making in selecting patients for appropriate therapy. They advocate early CI in patients with SDS less than 30 or extensive cochlear involvement on HRCT (Rotteveel grade 2C and 3). Patients with SDS better than 30%, early HRCT lesions and a measurable ABG of at least 30 dB can be managed with stapedotomy alone or followed by hearing aid [5].

Our patient had very poor SDS and extensive spongiotic bone lesions along with ossification of basal turn of the cochlea. Considering this he was offered CI as the initial choice of rehabilitation. We contemplated FNS, misplacement or difficult insertion of array. However, with careful surgical planning we were able to circumvent these problems. We adopted two important strategies to reduce the FNS. Perimodiolar array was used to limit the spread of current and secondly flouride therapy to promote recalcification of the otospongiotic lesions was started three months prior to surgery which is being continued till date. 18 months after surgery the implant is performing well and the patient continues to have good speech discrimination.

Conclusion:

Management of patients with FAO is challenging. There are no clear guidelines on the management of patients with FAO thus it is imperative that each case should be evaluated individually. Important factors to be considered prior to deciding the type of surgical management include presence of an AB gap, SDS and radiologic staging of disease. Surgical rehabilitation in the form of CI itself poses challenges. Surgical outcomes can be improved by using a perimodiolar array and addition of fluoride therapy to limit the spread of current.

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