Cochlear Implantation in a Patient with Osteopetrosis

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Abstract

Osteopetrosis is a rare and heterogeneous group of hereditary skeletal dysplasia syndromes. Manifestations of this condition are common in the temporal bone, particularly with regards to hearing loss. We present the case of a 41 year old male with a well-documented history of osteopetrosis and longstanding, profound, bilateral sensorineural hearing loss who underwent chochlear implantation.

Background

Osteopetrosis is a heterogeneous family of rare genetic disorders characterized by increased bone density due to defective osteoclast functioning.[1] Sclerotic bone formation in the temporal bone can lead to manifestations of hearing loss in multiple ways. For example, compression of the cochlear blood supply or compression of the cochlear nerve within the narrowed IAC, can lead to a sensorineural loss, while fixation of the ossicular chain and obliteration of the round and oval windows can lead to a conductive loss.[3]

Patient and Case Description

A 41 year-old Caucasian male a well-documented history of osteopetrosis presented with longstanding, progressive, bilateral sensorineural hearing loss. He had used bilateral hearing aids since the age of five with progressively less benefit over time. On physical exam, he had a skull shape consistent with osteopetrosis, bilateral narrowing of the external auditory canal, and unremarkable tympanic membranes. Vestibular function and all other cranial nerves were intact.

Audiometry showed a profound SNHL in both ears. Speech in noise testing in the best aided condition (AZ-Bio) demonstrated 0% speech understanding in each ear and bilaterally. High resolution CT of the temporal bone was notable for extensive thickening of the calvarium and temporal bone with bilateral narrowing of the internal auditory canal (IAC). MRI with and without gadolinium confirmed the CT findings and demonstrated adequate intracochlear fluid signal bilaterally.
The cochleovestibular nerves on either side were poorly visualized in the narrowed IACs (Figures 1 and 2).

The patient was counseled extensively on the possible risks and outcomes of cochlear implantation, including lack of improvement in speech perception post-operatively as well as risks associated with obliteration of the external auditory canal. A transcanal approach to the cochlea with a blind sac of the external auditory canal was performed. Intraoperatively, the entire medial wall of the mesotympanum was found to be ossified and the round window was not visualized. A cochleostomy was performed in the basal turn and a Cochlear Corporation Contour Advance electrode was fully inserted. Intra-cochlear placement was confirmed by an intra-operative X-ray and intraoperative neural response telemetry detected an auditory response to electrical stimulation. Post-operative high resolution CT scan further confirmed correct placement and verified full insertion of the patient's electrode (Figures 3 and 4). The patient tolerated the procedure well with postoperative complications. He demonstrated significant improvement in his access to speech sounds with an average aided threshold of 20 dB in his implanted ear at his two month follow-up visit. with

Figure Legend

1) Pre-operative axial CT scan showing diffuse bony sclerosis and thickening of the temporal bone around the otherwise normal appearing bilateral cochlea and labyrinth
2) Pre-operative T2 MRI demonstrating an intact but poorly defined and notably small cochleovestibular nerve (white arrow)
3) Post-operative axial CT scan showing CI electrode positioned within the right cochlea (black arrow)
4) Post-operative coronal CT scan, again showing CI electrode within the right cochlea (black arrow), as well as its connecting wire traveling through the blind sac external ear canal (red arrow)

Discussion

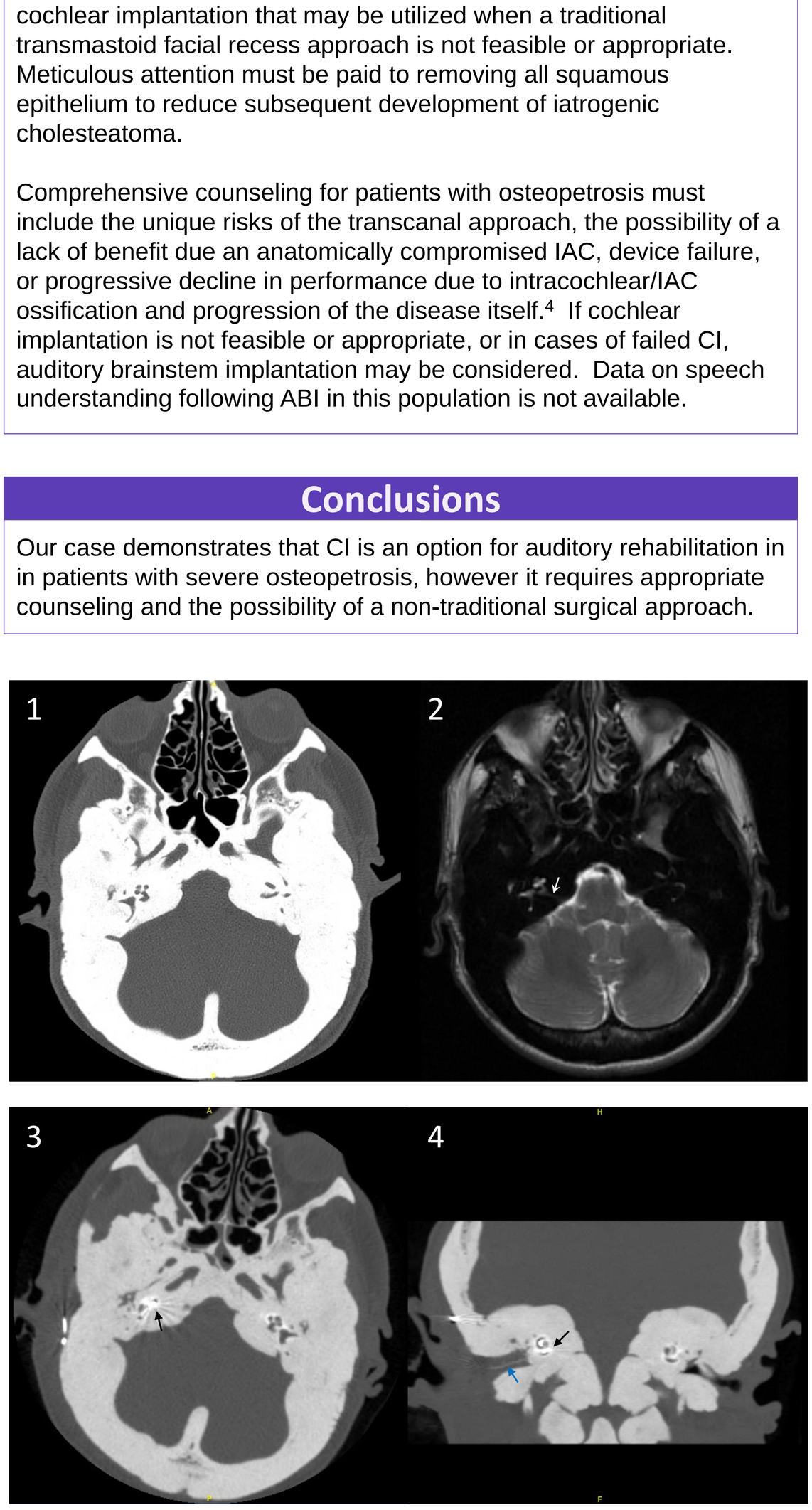
To our knowledge, this is only the second documented case of CI for the treatment of sensorineural hearing loss in a patient with osteopetrosis.[4] Noninvasive treatment with traditional amplification is the usual initial intervention for hearing loss due to osteopetrosis. Ossiculoplasty to treat the conductive component may be done, but is often technically difficult due to increased density of the middle ear osseous structures.[2] When the patient has bilateral severe SNHL, CI is discussed, bearing in mind that the possibility of treatment failure due to compression of the cochlear nerve within the IAC must be considered. However, indirect signs of a functional cochlear nerve can be inferred, as in the case of normally functioning facial and vestibular nerves.[3] Promontory stimulation testing, if available, can also be considered to asses auditory response to electrical stimulation.

Transcanal approach with blind sac is a non-traditional method of cochlear implantation that may be utilized when a traditional transmastoid facial recess approach is not feasible or appropriate. Meticulous attention must be paid to removing all squamous epithelium to reduce subsequent development of iatrogenic cholesteatoma.

Comprehensive counseling for patients with osteopetrosis must include the unique risks of the transcanal approach, the possibility of a lack of benefit due an anatomically compromised IAC, device failure, or progressive decline in performance due to intracochlear/IAC ossification and progression of the disease itself.[4] If cochlear implantation is not feasible or appropriate, or in cases of failed CI, auditory brainstem implantation may be considered. Data on speech understanding following ABI in this population is not available.

Conclusions

Our case demonstrates that CI is an option for auditory rehabilitation in in patients with severe osteopetrosis, however it requires appropriate counseling and the possibility of a non-traditional surgical approach.



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