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Double challenge: cochlear implantation in the only hearing ear with progressive hearing loss following meningitis and vestibular dysfunction after implantation

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ABSTRACT

Objective: Vestibular dysfunction associated with cochlear implantation is rare. It is usually seen in patients with otosclerosis due to spread of electrical activity throughout the demineralized bone. A 17-year old female with progressive hearing loss 2 years after meningitis and vestibular dysfunction in the implanted ear is presented in this study.

Findings: The patient had mild hearing loss in the right ear and total hearing loss on the left side because of complete ossification of the cochlea following meningitis. She had to have cochlear implantation in the right ear because of progression of hearing loss. She had successful implantation but she experienced vestibular dysfunction following activation of cochlear electrodes. Closure of two electrodes caused disruption of auditory programming. Then the patient was subjected to long term vestibular rehabilitation program.

Conclusion: Timing for implantation before the completion of cochlear ossification is crucial not to miss the chance for hearing restoration. However, difficulties in hearing rehabilitation due to extensive ossification can be doubled by vestibular problems triggered by stimulation of the vestibular nerve by cochlear electrodes. Attempts to reduce the balance problem will complicate auditory programming. Vestibular rehabilitation for long term helps to carry on hearing progress.

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1. Introduction

Ossification of the cochlea is an important sequela of bacterial meningitis which usually begins at the basal cochlear helix and progressively occupies the whole labyrinth (Dodge et al., 1984). The internal auditory canal and the cochlear aqueduct are potential pathways for the spread of infection from meninges to the inner ear. Merchant and Gopen have reviewed histopathologic changes of the temporal bones from patients with bacterial meningitis (Merchant and Gopen, 1996). They have found that the sensory and neural structures of the inner ear were intact in majority of them. However, spiral ganglion cells were degenerated, indicating a retrocochlear site of hearing loss in addition to the cochlea which may explain why some patients may perform poorly after cochlear implantation. Children with cochlear ossification require higher level of stimulation of cochlear electrodes and they need frequent programming adjustments (Eshragi et al., 2004).

Alteration of normal fluid homeostatis of the inner ear, trauma in the vestibular sensorial structures, surgery-induced inflammation and particularly, spread of electrical stimulation along the spongy cochlea as seen in implanted otosclerosis patients are the possible causes of vestibular dysfunction following cochlear implantation (Hänsel et al., 2018). Rehabilitation of hearing loss in patients with meningitis is an interesting subject. We present case study of a patient with cochlear implantation in the only hearing ear due to progressive hearing loss following meningitis and vertigo after implantation.

2. Case report

A 17-year-old girl had pneumococcus meningitis two years ago. She had total hearing loss on the left side due to fast and complete ossification of the cochlea within days and moderate hearing loss

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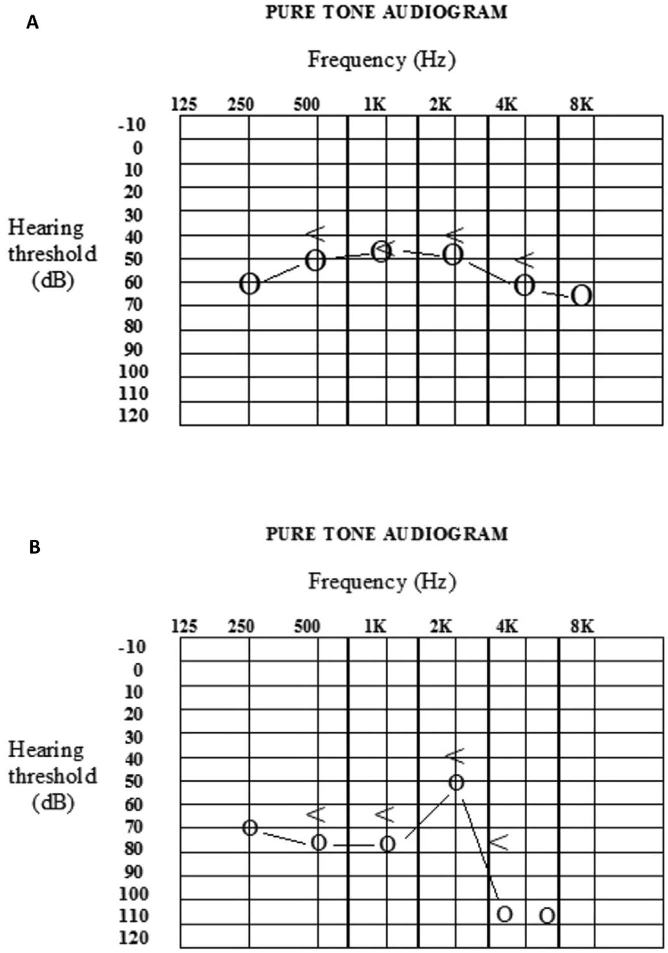

Fig. 1. (a). Audiometry of the patient a few weeks after meningitis shows 45 dB hearing loss on the right side. (b). Two years later, hearing deteriorates to 65 dB hearing loss.

on the right side. She was doing well with hearing aid in the right ear over a year. She was followed on regular basis with MRI and audiometry. However, her hearing on the right side gradually decreased from 45 dB to 65 dB in two years (Fig. 1a and b). Un-aided speech discrimination score decreased from 73% to 32% and the patient was unable to manage with hearing aid. Computed tomography of the temporal bone demonstrated that the cochlea was normal on the right side and the left cochlea was completely ossified (Fig. 2a and b). T2-weighted magnetic resonance imaging demonstrated no cochlear patency on the left and normal appearance on the right side (Fig. 3 a, and b). It was possible to electrically stimulate the left ear. The patient was a good lip-reader. Alternatives for better hearing rehabilitation was discussed with the parents and she had cochlear implantation in the only hearing right ear (Fig. 4a and b, c and d). Electrode array was implanted without any difficulty and the patient was able to respond across a wide frequency range after activation of the electrodes. Sound perception, discrimination and the ability to detect speech at a close-to-normal hearing intensity in noisy and silent environment was good at six months. Disyllabic word score was 80% with lip-reading. However, postoperative balance problem complicated the hearing rehabilitation. Electrode re-programming with closure of two electrodes resulted with decreased hearing performance. Patient was later subjected to medical therapy and long-term vestibular rehabilitation with some relief. An informed consent was obtained from the patient.

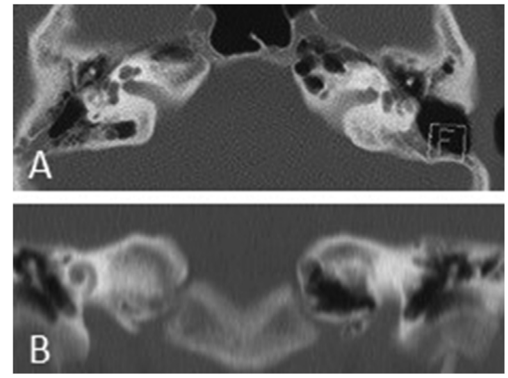

Fig. 2. Axial (a) and coronal (b) sections of the temporal bone computed tomography demonstrates no cochlea with severe ossification on the left side and normal cochlea on the left side.

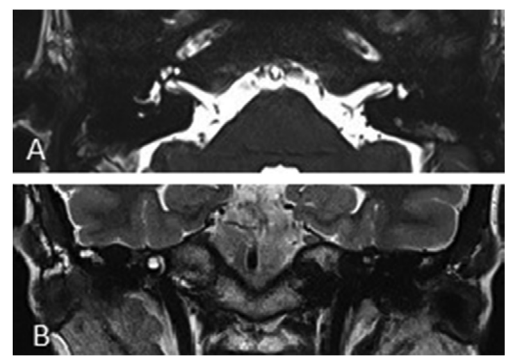

Fig. 3. T2-weighted temporal bone MRI demonstrates cochlea with normal patency on the right side on axial (a) and coronal (b) sections. Note complete ossification of the left cochlea.

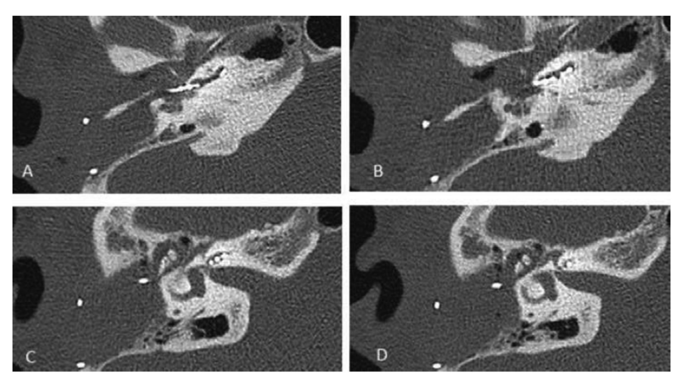

Fig. 4. Serial axial sections of the temporal bone demonstrate complete insertion of the electrode (a, b, c and d).

3. Discussion

Bilateral hearing loss associated with labyrinthine ossification following meningitis presents diagnostic and surgical challenges to the cochlear implantation. Incomplete insertion is frequently experienced. Drill-out technique using short and condensed electrodes for patients with partial ossification has been described (Gantz et al., 1988). If cochlear obliteration is restricted to the basal turn, electrode insertion is possible with apical cochleostomy or the implants with double arrays can be used (Montandon et al., 1994). However, these options were not valid for the left ear with complete cochlear obliteration. On the other hand, she had good response to electrical stimulation. Placing a dummy electrode in the left cochlea to keep a space at the very early weeks of meningitis before completion of the labyrinthine ossification would increase the choices of hearing rehabilitation for future planning. Two years after meningitis, progression of hearing loss in the right ear urged the patient to seek a better way for hearing rehabilitation. Brainstem implantation in the left ear and to continue to wear hearing aid in the right ear for a while was an option to review. Yet, she still had some benefits from hearing aid in the right ear even though there was worsening over 2 years. On the other hand, cochlear implantation in the right ear was another option not to miss before hearing has been completely lost. The patient was followed with computerized tomography and MRI of the temporal bone on regular basis. There was no change in terms of ossification in the right ear despite gradual worsening of hearing.

Fluctuation of hearing, delayed or progressive hearing loss following meningitis are interesting and rare incidents. Hearing loss years later following meningitis in the presented patient could be due to intracochlear fibrosis which is not quite evident on CT scanning. Javarajan and Rangan have reported 2 interesting cases with worsening of hearing 17 years after meningitis in one case and 3 years after in the other one (Javarajan and Rangan, 1999). Fluctuating of hearing could be associated with endolymphatic sac destruction. Silkes and Chabot have reported an 11-year documented fluctuating but downward progression of hearing loss following meningitis (Silkes and Chabot, 1985). Durisin et al. have reviewed computed tomographic findings of 126 patients with profound hearing loss due to meningitis and have reported increased rate of osteoneogenesis over time after meningitis (Durisin et al., 2010). Animal studies indicate that the sequence of events starts with inflammation that progresses to fibrosis and the ossification of the cochlea can continue over years (Brodie et al., 1998). Inflammatory cell infiltration to the labyrinth following meningitis on animal studies demonstrate that the process is not abrupt but progressive (Kesser et al., 1999). Computed tomography is useful to show bony structure of the cochlea. But, its sensitivity for the detection of cochlear ossification is poor and it will not assure whether osteoneogenesis is still ongoing. T2 weighted MR images are superior in the evaluation of the cochlear patency (Kopelovich et al., 2011).

Cochlear implantation is generally a safe and reliable procedure. Vestibular end-organ dysfunction following cochlear implantation has been reported before (Ito, 1998). This is usually due to spread of electrical activity through the bone which is actually experienced more often in patients with otosclerosis. Vestibular nerve stimulation after cochlear implantation following meningitis has been reported as well (Cushing et al., 2013). The underlying cause is unclear. On the other hand, anatomical studies indicate neural anastomosis between the cochlear and inferior vestibular nerves although its functionality is obscure (Ozdogmus et al., 2004). This connection presents a potential pathway for electrical transmission. Treatment is generally based on medication and rehabilitation. Review of electrode stimulation in some instances could demonstrate how it is related with unexpected vestibular neural firing. This is really very disturbing condition. Often times, it can be cured with re-programming of the electrodes. However, this may present an additional challenge. Closure of some troublesome electrodes will compromise auditory performance when the hearing has to be provided by a limited number of electrodes. In such condition, decisive commitment of the patient to the vestibular rehabilitation program has utmost importance.

In conclusion, the case indicates that the patients with hearing loss following meningitis should be monitored closely for many years. Early measures for hearing restoration are important since poor outcomes are reported to be associated with severity of ossification, partial insertion of the array and duration of deafness. When there is complete ossification of the cochlea on both sides and promontory test did not reveal any cochlear activity, auditory brainstem implant is indicated although results are not always promising as cochlear implantation (Malerbi et al., 2018). Timing for cochlear implantation is crucial before complete ossification to avoid delay for proper rehabilitation. Vestibular dysfunction following cochlear implantation is rare but is challenging for auditory programming in patients with hearing loss due to meningitis. Some electrodes have to be switched off in severe cases. However, large case studies indicate that medication and vestibular compensation particularly in younger patients are helpful to overcome the threat (Hänsel et al., 2018).

Declaration of competing interest

The authors declare that they have no conflict of interest. This study has no grant or funding.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.joto.2019.11.002.

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