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Cochlear implantation in a patient with osteogenesis imperfecta

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ABSTRACT

Osteogenesis imperfecta (OI) is a connective tissue disorder characterized by a deficit in the synthesis of type I collagen. Hearing loss affects 42–58% of OI patients and progresses to deafness in 35–60% of these patients. For OI patients, cochlear implantation (CI) is the only promising treatment option. However, literature on CI in patients with OI is relatively rare. After CI, speech perception is generally good. However, among patients with severe demineralization of the cochlea, most patients are reported to have complications of facial nerve stimulation (FNS), preventing some patients from using the cochlear implant on a daily basis. Here we report a successful CI using a Nucleus CI24 Contour Advance cochlear implant in a patient with OI. Although high-resolution computed tomography (HRCT) showed extensive demineralization of the cochlea, intracochlear electrodes were inserted properly. The use of a modiolus-hugging device and the advance off-stylet technique contributed to the successful implantation, with no complications such as FNS or misplacement of electrodes. Therefore, CI can be used for treating deaf patients with OI.

1. Introduction

Osteogenesis imperfecta (OI) is a connective tissue disorder characterized by a deficit in the synthesis of type I collagen [1]. OI was first described by van der Hoeve and de Kleyn in 1917 [2] and, therefore, is also known as van der Hoeve–de Kleyn syndrome. The disease is characterized by brittle bones, blue sclerae, defective dentition and hearing loss [3]. Progressive hearing loss has been reported, including conductive, sensorineural, or mixed types [4]. Conductive hearing loss may be the result of a fracture or localized dehiscence of the stapedial arch, distal atrophy of the long process of the incus, or fixation of the stapedial footplate [1]. Sensorineural hearing loss is caused by microfractures, hemorrhage, and encroachment of reparative vascular and fibrous tissue in and around the cochlea [1]. Previous studies have reported hearing loss in 42–58% of OI patients and profound deafness in 35–60% of OI patients [5–9]. Hearing loss usually begins in the late teens in OI patients. The sensorineural component appears and progresses gradually in the third decade, resulting in profound deafness by the end of the fourth to fifth decade [7]. Cochlear implantation (CI) is the only treatment option for profound sensorineural hearing loss. However, the scientific and medical literature on CI in patients with OI is relatively rare [5–10]. After CI, speech perception is generally good. However, most patients with severe demineralization of the cochlea are reported to have complications of facial nerve stimulation (FNS). Several cases of electrode mis-insertion have also been reported. Some patients with such complications give up daily use of the cochlear implant [7,11].

Here, we report a successful CI using a Nucleus CI24 Contour Advance (CA) cochlear implant in a patient with OI. Although high-resolution computed tomography (HRCT) showed extensive demineralization of the cochlea, intracochlear electrodes were properly inserted without any of the common complications.

2. Case presentation

A female patient had several episodes of bone fractures due to minor trauma from childhood. At 18-years of age, she began to complain of bilateral hearing loss. A clinical examination revealed blue sclerae with hearing loss, and the patient was diagnosed as OI based on the clinical criteria [12] at the age of 21 years. The patient had no family history of OI or hearing loss, except for her grandfather who had presbycusis. At the age of 27 years, the patient underwent an ossiculoplasty of the left ear that unfortunately resulted in deafness. Subsequently, she began to wear a hearing aid in the right ear. At the age of 52 years, the patient consulted our department when her hearing acuity in the right ear worsened. An otoscopy examination revealed normal tympanic membranes in both ears. A pure-tone audiogram demonstrated

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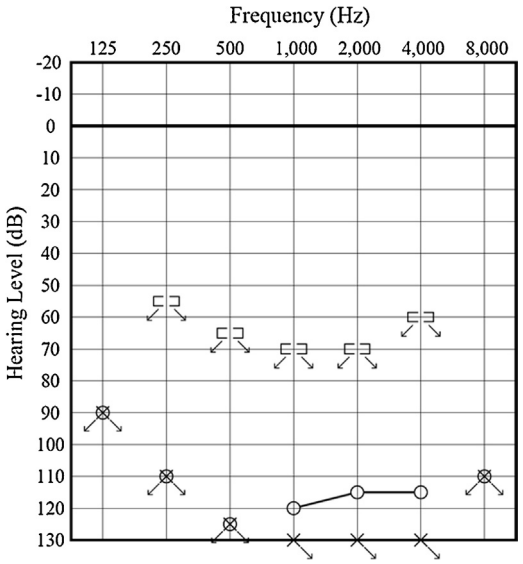

Fig. 1. Preoperative audiogram. A pure-tone audiogram demonstrated profound sensorineural hearing loss in both ears.

profound sensorineural hearing loss in both ears (Fig. 1). A speech discrimination test revealed no identification ability in both ears. Speech recognition scores with a hearing aid showed that only 10% of phonemes were recognized in the open set condition and 24% were recognized with verbal cues. The vestibular evoked myogenic potential (VEMP) was absent in both sides. A caloric test did not evoke nystagmus in either ear. A promontory stimulation test produced good auditory perception in both ears. HRCT revealed severe demineralization of the pericochlear and vestibular areas in both sides, and the outline of the cochlea was almost unrecognizable (Fig. 2). Magnetic resonance imaging (MRI) showed fluid intensity in the entire cavity of the right cochlea. However, fluid intensity in the scala tympani of the basal turn was decreased in the left cochlea (Fig. 3). The right and left cochlear nerves were well recognized on MRI.

Because there was a long period of auditory deprivation of the left ear and the MRI suggested partial occlusion in the basal turn, we decided to perform CI for the right ear. At the age of 54 years, the patient underwent surgery in the right ear to implant the Nucleus CI24R Contour Advance device. A mastoidectomy and a posterior tympanotomy were performed uneventfully. The foramen obturatum and the oval window were obliterated, and the round window was barely identified by the new bone formation of the promontory. We performed chocleostomy using the location of stapes as a landmark. The bone of the cochlear capsule was spongiotic and fragile; however, a cochleostomy was easily performed and the scala tympani was identified. All of the 22 electrodes were inserted successfully using the advanced off-stylet (AOS) technique. Postoperative neural response telemetry (NRT) showed good responses in all electrodes without FNS. Postoperative radiography and HRCT revealed the fully inserted electrodes inside the cochlea (Fig. 4). All of the electrodes showed normal impedance at first stimulation, and

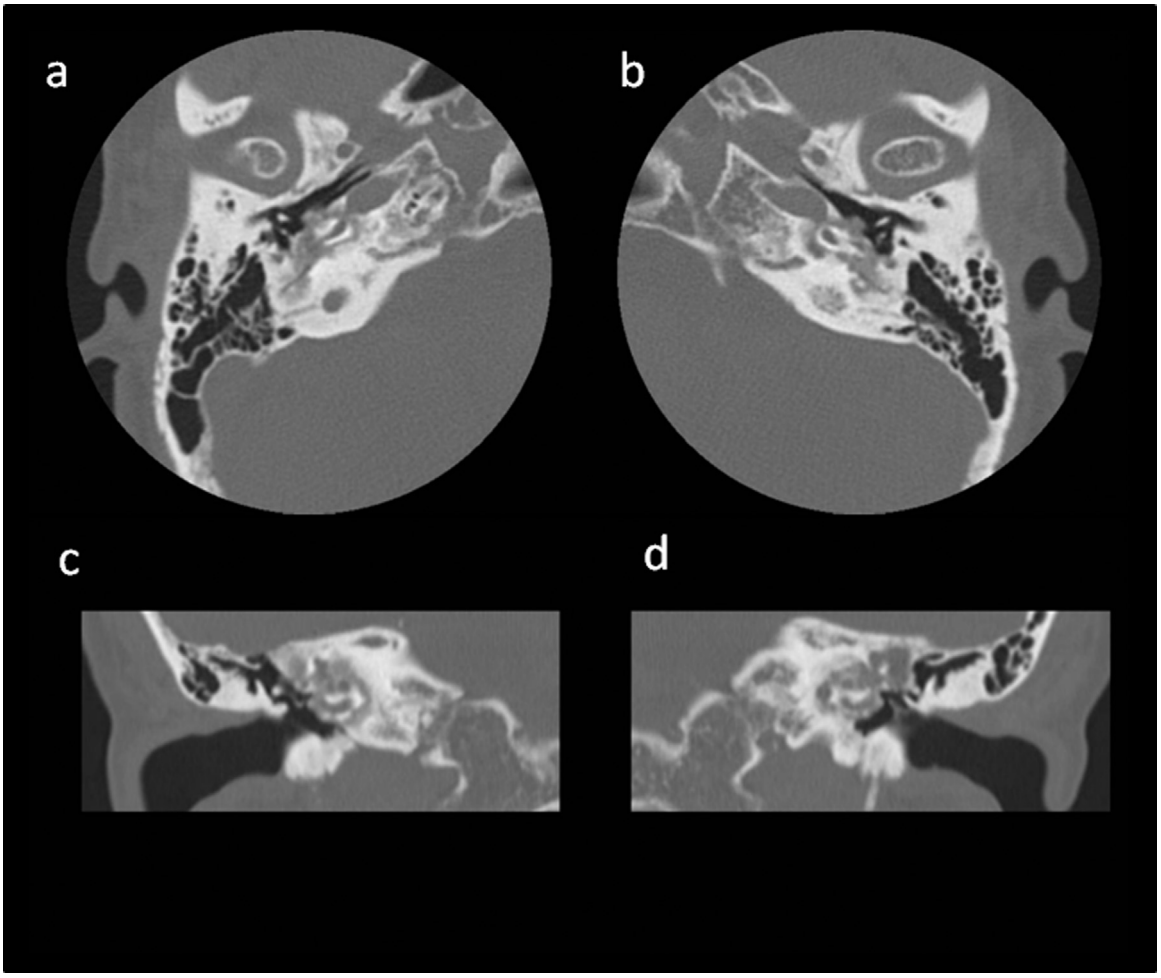

Fig. 2. Preoperative CT image of the cochlea. High resolution computed tomography (HRCT) revealed significant demineralization of the right and left pericochlear and vestibular areas, and the structures of the cochleae were almost unrecognizable.