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Histopathology of the human inner ear in Cogan's syndrome with cochlear implantation

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Abstract

Cogan's syndrome is a rare disorder characterized by nonsyphilitic interstitial keratitis and audiovestibular symptoms. Profound sensorineural hearing loss has been reported in approximately half of patients with Cogan's syndrome resulting in candidacy for cochlear implantation in some patients. The current study is the first report of the histopathology of the temporal bones from a patient with Cogan's syndrome who during life underwent bilateral cochlear implantation. Preoperative MRI revealed tissue with high density in the basal turns of both cochleae and both vestibular systems consistent with fibrous tissue due to labyrinthitis. Histopathology demonstrated fibrous tissue and new bone formation within the cochlea and vestibular apparatus, worse on the right. Severe degeneration of the vestibular end organs and new bone formation in the labyrinth were seen more on the right than the left. Although severe bilateral degeneration of the spiral ganglion neurons was seen, especially on the right, the postoperative word discrimination score was between 50 and 60 % bilaterally. Impedance measures were generally higher in the right ear, possibly related to more fibrous tissue and new bone found in the scala tympani on the right side.

Keywords

Histopathology; Cogan's syndrome; sensorineural hearing loss; cochlear implantation; impedance; vasculitis; human

Introduction

Cogan's syndrome is a rare disorder characterized by inflammatory eye disease and audiovestibular symptoms thought to be of autoimmune etiology [Kessel et al., 2014; Jung et al., 2016]. It may cause bilateral sudden or rapidly progressive sensorineural hearing loss,

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Disclosure statement

The authors have no conflicts of interest to disclose.

and despite immunosuppressive therapy, profound hearing loss has been reported in 52 % of patients [Gluth et al., 2006]. Patients with bilateral profound sensorineural hearing loss may become candidates for cochlear implantation. Some clinical reports of patients with Cogan's syndrome who have undergone cochlear implantation describe wound healing problems postoperatively [Wang et al., 1990; Kontorinis et al., 2010]. In addition, of six patients who sustained deafness due to Cogan's syndrome, partial obliteration or neo-ossification of the cochlea was encountered during cochlear implantation in all cases, which required an alteration of the standard surgical technique [Aschendorff et al., 2004]. Fibrous obliteration of the scala tympani was found as early as eight weeks after onset of complete deafness [Aschendorff et al., 2004]. Although deterioration of auditory performance after cochlear implantation has been described [Bovo et al., 2011], presumably secondary to an ongoing inflammatory response due to Cogan's syndrome, stable postoperative hearing has been reported with follow-up of one year [Pasanisi et al., 2003], two years [Wang et al., 2010] and five years [Bacciu et al., 2015] with good to excellent word and sentence recognition scores suggesting that cochlear implantation provides excellent and stable hearing rehabilitation with long term follow-up in most patients.

Although there have been several reports of the histopathology of the inner ear in Cogan's syndrome [Fisher and Hellstrom, 1961; Wolff et al., 1965; Zechner, 1980; Rarey et al., 1986; Schuknecht and Nadol, 1994; Ishii et al., 1995; Jung et al., 2016] to date there has been no description of the histopathology of the inner ear following cochlear implantation in Cogan's syndrome. The temporal bones of a 64-year old patient who suffered from a bilateral onset of rapidly progressive and profound sensorineural hearing loss due to Cogan's syndrome and who underwent bilateral cochlear implantation 19 months prior to death have been obtained by the Otopathology Laboratory of the Massachusetts Eye and Ear Infirmary.

Materials and Methods

Temporal Bone Acquisition

Temporal bone specimens were obtained in compliance with Human Subject Assurance No. FWA00006221 approved by the Institutional Review Board of the Massachusetts Eye and Ear Infirmary.

Histological techniques

The temporal bones were removed 22 hours following death and were fixed in 10 % buffered formaldehyde. After CT scanning, the temporal bone specimens were decalcified using ethylene diamine tetracetic acid. Following decalcification, the electrode array was removed from the cochlea on the right side. On the left, the electrode array was inadvertently removed before CT scanning could be done. The specimens were then dehydrated in graded alcohols and embedded in celloidin. Sectioning was done in the horizontal (axial) plane at a thickness of 20 μ m. Every tenth section was stained with hematoxylin and eosin, and mounted on glass slides for subsequent two-dimensional reconstruction [Guild, 1921; Schuknecht, 1993].

Results

Clinical history

This 64-year old male suffered from a bilateral sudden and rapidly progressive sensorineural hearing loss starting approximately two years before death in the left ear and involving the right ear two days later. His hearing loss was accompanied by severe vertigo. His previous otologic history was unremarkable other than left facial weakness which resolved spontaneously one month prior to his hearing loss. He had been treated by an ophthalmologist for a “recurrent idiopathic orbital inflammatory syndrome” prior to onset of audiovestibular symptoms and which showed some responsiveness to oral steroids with which he had been treated for six months. His otolaryngologic examination was unremarkable other than hearing loss. An audiogram done at the age of 62 showed anacusis in the left ear and a severe to profound sensorineural hearing loss with 0 % speech discrimination and a puretone average of 90 dB in the right ear (Figure 1A, B). The patient was hospitalized for evaluation and treatment of his rapidly progressive bilateral hearing loss and vertigo. Magnetic resonance imaging (MRI) demonstrated the presence of hyperintense lesions in T1-weighted images in the basal turn of the cochlea on both sides, the vestibule on the right side, and in the ampulla of the superior semicircular canal on the left side, which were consistent with labyrinthitis in both inner ears (Figure 2A–D). Lumbar puncture and examination of the cerebrospinal fluid were negative. The C-reactive protein (CRP) was 242.6 mg/dl (normal range, 0–3 mg/dL), and the rheumatoid factor was less than 30 IU/mL (normal range, less than 30 IU/mL). The TREP-Sure™ enzyme-linked immunosorbent assay (ELISA) for syphilis was negative. Testing for antineutrophil cytoplasmic antibodies (ANCA) was negative. The erythrocyte sedimentation rate (ESR) was 95 mm/hour (normal range, 0–11 mm/hour). The diagnosis of Cogan’s syndrome was made, and he was treated with high dose systemic steroids with improvement in his vestibular symptoms but not in the hearing loss. At the age of 63, he underwent bilateral cochlear implantation using Advanced Bionics Hi-Res 90K receiver stimulator with HiFocus Helix electrodes (perimodiolar). Intraoperatively upon opening the cochleostomy in the left ear, the contents were described as “sclerotic and gelatinous”. However a full insertion of the electrode was accomplished using an off-stylet approach with an insertion tool, and facilitated by the use of an injection of sodium hyaluronate into the cochlea as a lubricant. The difficulties encountered on the left were not present in the subsequent right cochlear implantation. A full insertion was achieved in both ears.

The last available postoperative audiologic testing was done one year following bilateral cochlear implantation and six months prior to death. The sound awareness threshold in both ears was approximately 30 dB. The postoperative last-recorded word recognition score (CNC (consonant-nucleus-consonant) word list) was 50 % on the left side, 60 % on the right side, and 56 % with bilateral stimulation.

Impedance measures

Mean impedance measures ($\pm$ SD) of the electrode of both ears recorded in the last three sessions (157, 248, and 393 days after initial activation) are shown in Figure 3. Impedance

measures in the right ear were approximately 5 k Ω all along the electrode array, and generally higher than those in the left ear.

Past medical history and autopsy findings

The past medical history included ulcerative colitis for the last 40 years of his life. The patient died of acute hemorrhagic pancreatitis with necrosis. Additional autopsy findings included ascites, cardiomegaly, fungal cystitis, and rectal adenocarcinoma. Neuropathologic autopsy revealed mild gliosis and prominent stroma in the optic nerves and mild gliosis of CNVIII thought to be consistent with the diagnosis of Cogan's syndrome. The temporal lobes, pons, and medulla were normal.

Histopathologic findings

Cochleae—Both cochleae possessed 2-1/2 turns by two-dimensional reconstruction. The length of the cochlear duct on the right was 36.2 mm and 34.9 mm on the left. There was severe to profound degeneration of inner and outer hair cells throughout the both cochleae (Figure 1A, B). There was endolymphatic hydrops in all turns of the right ear, whereas there was collapse of Reissner's membrane in all turns of the left ear. The tectorial membrane of the right ear was rolled up, detached, and encapsulated.

Cochlear implants—The length of the implant electrode was 18.9 mm on the right and 23.5 mm on the left. The track of the electrode was completely within the scala tympani in the right ear, whereas the electrode track on the left migrated from the scala tympani into the scala vestibuli at millimeters 10 to 12 from the round window with fracture of the osseous spiral lamina (Figure 4). Loose fibrous tissue filled much of the scala tympani and scala vestibuli along the electrode track in both ears (Figure 4), more on the right. There was a fibrous sheath around the electrode array in both cochleae (Figure 4, 5). New bone formation was seen in the scala vestibuli and spiral ligament in the right ear (Figure 5A), and in the scala tympani, scala vestibuli, and spiral ligament in the left (Figure 5B).

Cochlear neurons—There was degeneration of the cochlear neurons of the both ears, worse in the right. The corrected spiral ganglion cell count on the left ear for segment I was 1,714 cells; segment II was 5,936 cells; segment III was 1,306 cells, and segment IV was 925 cells. The total corrected spiral ganglion cell count was 9,881 on the left which represented 43% of normal for age (Figure 1A). The corrected cell count on the right for segment I was 911 cells; segment II was 2,428 cells; segment III was 870 cells, and segment IV was 1,115 cells. The total corrected spiral ganglion cell count was 5,324 on the right which represented 23 % of normal for age (Figure 1B).

Vestibular system—In the right ear, the ampullae of the superior and lateral semicircular canal were totally degenerated (Figure 6A), and the ampulla of the posterior semicircular canal showed evidence of atrophy. On the left, the ampullae of the superior, lateral, and posterior semicircular canals showed moderate to complete degeneration (Figure 6B). The maculae sacculi and utriculi showed moderate to total degeneration on the right, and moderate to mild degeneration on the left. There was endolymphatic hydrops affecting the saccule on the right (Figure 7), whereas no evidence of hydrops was found on the left. There

was new bone formation in the superior, lateral, and posterior semicircular canals and crus commune in both ears (Figure 8A, B).

Discussion

Characteristics of Cogan's syndrome

Cogan's syndrome is a rare disorder characterized by nonsyphilitic interstitial keratitis and audiovestibular symptoms. The association of these symptoms was first described in 1934 [Morgan and Baumgartner, 1934], and Cogan's syndrome was named after David Cogan, who described an additional four cases [Cogan, 1945]. Systemic vasculitis including aortitis may occur during the course of Cogan's syndrome [Gluth et al., 2006; Kaya et al., 2015]. Aortitis with aortic insufficiency, which may lead to congestive heart failure has been reported to occur in about 10 % of patients with Cogan's syndrome [Grasland et al., 2004; Gluth et al., 2006; Kessel et al., 2014]. Cogan's syndrome is considered to be associated with inflammatory bowel disease (IBD) such as ulcerative colitis (UC) and Crohn's disease (CD) [Scharl et al., 2010; Vavricka et al., 2015], and UC was described in the medical history of the current case. The etiology of Cogan's syndrome has been assumed to be an autoimmune disorder with vasculitis [Kessel et al., 2014; Jung et al., 2016].

Two types of Cogan's syndrome have been described; i.e. typical and atypical types [Kessel et al., 2014; Jung et al., 2016]. Typical Cogan's syndrome consists of non-syphilitic interstitial keratitis and audiovestibular symptoms similar to those of Ménière's syndrome. Atypical Cogan's syndrome is characterized by other ocular inflammation with or without interstitial keratitis in association with audiovestibular symptoms unlike those of Ménière's syndrome [Kessel et al., 2014]. Systemic symptoms such as rheumatoid arthritis are much more frequent in atypical Cogan's syndrome than in the typical variant [Kessel et al., 2014]. The current case is consistent with typical Cogan's syndrome given the lack of systemic symptoms.

Treatment of Cogan's syndrome

Treatment of Cogan's syndrome depends on severity and extensiveness [Kessel et al., 2014; Espinoza and Prost, 2015]. Topical corticosteroids are chosen in cases with mild ocular inflammation, and systemic corticosteroids in cases with severe ocular inflammation, audiovestibular symptoms, and systemic vasculitis [Kessel et al., 2014; Espinoza and Prost, 2015]. At the onset of audiovestibular symptoms, earlier treatment of systemic corticosteroids is preferable. Haynes et al. [1980] has reported that 55 % of patients treated with systemic steroids within two weeks showed hearing improvement, whereas only 8 % of patients treated after two weeks showed hearing improvement. However, Gluth et al. [2006] has reported that about half of patients suffered bilateral profound sensorineural hearing loss despite treatment.

Cochlear implantation in Cogan's syndrome

Cochlear implantation for patients with Cogan's syndrome has been reported in several studies [Pasanisi et al., 2003; Aschendorff et al., 2004; Kontorinis et al., 2010; Wang et al., 2010; Bovo et al., 2011; Bacciu et al., 2015]. Generally, postoperative hearing outcomes

have been reported to be good to excellent, as was true in the current case. However, intraoperative and postoperative complications such as partial obliteration of the cochlea [Pasanisi et al., 2003; Aschendorff et al., 2004; Kontorinis et al., 2010], flap complications including exposure of the implant [Wang et al., 1990; Kontorinis et al., 2010], and postoperative deterioration of auditory performance [Bovo et al., 2011] have been reported. In the current case, although the preoperative MRI scan showed hyperintense lesions within the basal turn of both cochleae, full insertion of the electrode was achieved on both sides. Preoperative MRI scan may help to predict the difficulty encountered during insertion of the implant electrode with enhancement of the cochlea or focal loss of signal predicting active labyrinthitis or labyrinthitis ossificans, respectively. Aschendorff et al. [2004] have reported that fibrous obliteration may occur as early as eight weeks after onset of deafness. Bovo et al. [2011] have reported two cases of postoperative deterioration of auditory performance of the cochlear implant. In one case, deterioration of speech perception was attributed to progressive cochlear ossification. In the other case, an abrupt deterioration of loudness perception required an increase in electrical stimulation. Generally, new bone formation is seen in the cochlea after cochlear implantation for deafness of various etiologies [Li et al., 2007; Somdas et al., 2007; Seyyedi and Nadol, 2014; Kamakura and Nadol, 2016], as well as in the cochleae deafened by Cogan's syndrome without cochlear implantation [Fisher and Hellstrom, 1961; Wolff et al., 1965; Zechner, 1980; Rarey et al., 1986; Schuknecht and Nadol, 1994]. Postoperative word recognition scores have been reported to be negatively correlated with the % volume of new bone formation within the cochlea after implantation [Kamakura and Nadol, 2016].

Impedance of the implant electrode

Post-implantation impedance measures were measured in both ears in the current study. The impedance in the right ear was approximately 5 k Ω all along the electrode array and generally higher than measures in the left ear (Figure 3). Fibrous tissue and new bone filled the scala tympani and scala vestibuli all along the electrode track in the right ear (Figure 5A). The degree of fibrous tissue and new bone was less in the left ear (Figure 5B). These findings are consistent with the study of Ni et al. [1992] who have demonstrated that electrode impedance measures were positively correlated with the degree of tissue response within the scala tympani in the kitten.

Conclusions

The histopathology of the inner ear in Cogan's syndrome with bilateral cochlear implantation has been reported herein. Despite severe degeneration of the spiral ganglion cells, the patient showed good postoperative speech discrimination in both ears. There was a considerable deposition of fibrous tissue and new bone in both ears, more on the right, which may correlate with the higher impedance measures on the right. The degenerative changes in the vestibular system in both ears can logically be attributed to the inflammatory process in Cogan's syndrome rather than a consequence of cochlear implantation.

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Figure 1. Audiogram and Cytocochleogram of the right (A) and left (B) ears. Missing cytological elements are shown in black. Audiogram of the patient obtained at the age of 62 (seven months prior to cochlear implantation). Speech discrimination testing was not possible on the left ear, and on the right, the CNC (consonant-nucleus-consonant) score was 0 %. IHC indicates inner hair cells; OHC, outer hair cells.

Figure 2.

MRI imaging of both temporal bones performed 18 days before cochlear implantation. A: T1-weighted image. A hyperintense lesion was seen in the basal turn of both cochleae (arrows). B: post-contrast T1-weighted image. An enhancing lesion was seen in the basal turn of the right cochlea (arrow). C: post-contrast T1-weighted image. An enhancing lesion was seen in the vestibule of the right ear (arrow). D: T1-weighted image. A hyperintense lesion was seen in the ampulla of the left superior semicircular canal (arrow).

Figure 3. Postoperative impedance measures (mean and standard deviation of last three measures) in both ears. Impedance measures in the right ear (red) were generally higher than in the left ear (blue).

Figure 4.
Photomicrograph of the basal turn of the left cochlea. The electrode track (E) was in the scala vestibuli, and a fracture-dislocation of the osseous spiral lamina (arrow) had occurred. There was deposition of fibrous tissue (FT) and new bone (NB) near the electrode.

Figure 5. Photomicrographs of both cochleae. A: midmodiolar section of right cochlea. New bone formation (NB) was seen in the scala vestibuli and spiral ligament. The electrode track (E) was within the scala tympani in the lower basal turn, and very close to the modiolus in the upper basal turn. B: midmodiolar section of left cochlea: New bone formation (NB) was seen in the scala tympani, scala vestibuli, and spiral ligament, and fibrous tissue (FT) was seen incompletely filling the scala tympani.

Figure 6.

Photomicrographs of both ampullae of the lateral semicircular canals. A: On the right, the lateral ampulla was totally degenerated (arrow), and the endolymphatic space was filled with loose fibrous tissue. B: On the left, the neuroepithelium of the ampulla was partially intact (arrow). The endolymphatic compartment was otherwise normal.

Figure 7.
Photomicrograph of the right saccule (S). Endolymphatic hydrops was seen (arrow), and the perilymphatic compartment was filled with loose fibrous tissue.

Figure 8. Photomicrograph of the vestibular apparatus of both ears. A: In the right ear, new bone formation (NB) was seen in the lateral (LSCC) and posterior (PSCC) semicircular canals, and fibrous tissue (FT) nearly filled the vestibule. B: in the left ear, new bone formation (NB) was seen in the lateral (LSCC) and posterior (PSCC) semicircular canals.