

Cochlear Implantation in a Patient with Granulomatosis with Polyangiitis

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ABSTRACT **Background:** Granulomatosis with polyangiitis (GPA) otologic manifestations include conductive and sensorineural hearing loss (HL). Vasculitis is assumed to be the primary cause of otologic manifestations. Deaf patients and patients with HL who do not benefit from hearing aids can benefit from cochlear implants (CI). There are currently no specific guidelines for treatment of patients with GPA suited for CI.

Objectives: To assess whether patients who are deaf due to GPA are good candidates for CI and if prior surgical or medical treatment of the inflammation are needed.

Methods: A case report is presented.

Results: A 71-year-old female patient with GPA and bilateral profound HL underwent CI. Prior to CI, preparation consisted of audiological evaluations by an otolaryngologist and a rheumatologist, followed by a course of prednisone and methotrexate for middle ear and nasal inflammations. CI was performed with no complications. The speech reception threshold and the monosyllabic word discrimination score after surgery were 25 dBHL and 75%, respectively.

Conclusions: Inflammation due to GPA can be controlled medically with immunosuppressive medications without subtotal petrosectomy, as in chronic suppurative otitis media. Satisfactory audiological results can be expected.

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KEY WORDS: cochlear implant, chronic otitis media, granulomatosis with polyangiitis (GPA), petrosectomy

nerve palsy [2]. Vasculitis is assumed to be the primary cause of otologic manifestations, although hemorrhage, changes in vessel diameter, and lymphocyte infiltration may be causes [3]. Therapy for GPA includes a combination of corticosteroids and immunosuppressive (steroid sparing) medications [1,4].

Deaf patients and patients with HL who do not benefit from hearing aids can benefit from cochlear implantations (CI). Temporal bone and chronic ear diseases may complicate CI surgery in several ways, such as bleeding from granulation tissue, adhesions, and ossification. Patients without GPA but with infected chronic suppurative otitis media possibly need a preliminary subtotal petrosectomy to control infection before the CI is performed [5]. We present a case of a patient presenting with GPA who underwent CI.

PATIENTS AND METHODS

A 71-year-old female, diagnosed with GPA for 2 years presented to our clinic with bilateral profound HL. In her left ear the HL was mainly sensorineural, with a speech reception threshold (SRT) of 85 dBHL. In her right ear she had no functional hearing. The SRT was non-responsive (NR) with and without a hearing aid. Her pure-tone average was NR without a hearing aid and improved to 90 dBHL with a hearing aid. She had no speech perception for monosyllabic or bi-syllabic words in the right ear [Table 1]. Her tympanogram was B in both ears with bilateral effusion upon otoscopy. She also presented with tinnitus in the right ear. When additional medical history was taken, she reported a sudden HL in her right ear in January 2012, which was treated with systemic and salvage intratympanic steroids with no improvement. Other manifestations were a saddle nose deformity, yellow nasal crusts with a septal perforation, pulmonary and cardiac symptoms with positive anticardiolipin antibodies, positive myeloperoxidase (16.7 U/ml), antiproteinase-3 levels of 2.17 U/ml, and an elevated sedimentation rate (62 mm/hour). A brain and internal auditory canal MRI was normal.

She was using a hearing aid on the left ear but found it dif-

Granulomatosis with polyangiitis (GPA) is an immunologically mediated systemic disease of unknown etiology that is characterized by inflammatory reaction patterns (necrosis, granulomatous inflammation, and vasculitis), which occur in the upper and lower respiratory tracts and kidneys. GPA is an autoimmune small-vessel vasculitis that is included in the group of anti-neutrophil cytoplasmic antibody (ANCA)-associated small-vessel vasculitides [1]. GPA otologic manifestations include conductive HL through inflammation of the mucosa and middle-ear fluid accumulation, sensorineural HL, and facial

Table 1. Speech perception tests, prior and post-cochlear implant

	Prior to cochlear implant		Post cochlear implant	
	Unaided	Aided (cochlear implant)	Unaided	Aided (cochlear implant)
Pure tone average	NR	90 dB	NR	25 dB
Speech reception threshold	NR	NR	NR	25 dB
Bi-syllable speech perception	NT	NT	NT	quiet 80% noise 65%
Monosyllabic speech perception	NT		NT	quiet 75% noise 50%

dB = decibel, NR = non-responsive, NT = not tested

difficult to hear and understand speech. The option of CI for the right ear was presented to the patient and she consented. Prior to CI, a ventilation tube was inserted into the right ear, removing thick fluid from the middle ear. Treatment with prednisone 20 mg and methotrexate 12.5 mg was given. A follow-up visit after 3 months demonstrated that the nasal crusts and the effusion from the right ventilation tube subsided. Because of the response to medical treatment, we decided not to perform a subtotal petrosectomy. A temporal bone computed tomography (CT) demonstrated that the middle ear cleft was bilaterally filled with fluid but there were no bony abnormalities. The prednisone and methotrexate doses were lowered to 10 mg and 2.5 mg respectively.

RESULTS

The CI (Mid-Scala, Advanced Bionics, Switzerland) surgery used the facial recess approach. There was some necrotic material in the mastoid cavity, but otherwise the surgery was uneventful with no complications. Histologic examination of the mastoid mucosa demonstrated granulation tissue with no evidence of a granuloma. The audiological evaluation of the patient included sound-field detection thresholds and speech perception tests. When administered to the implanted ear alone, the recipient achieved an SRT of 25 dBHL and a flat curve of sound-field detection threshold to warble tones at 25 dBHL. We used the Hebrew Arthur Boothroyd monosyllabic speech perception test administered in monitored live voice, using a presentation level of 50 dBHL. In quiet conditions she achieved a score of 75%,

and with the addition of 40 dBHL speech noise (+10 dB speech noise ratio) her performance score was 50%. Her bi-syllable speech perception in quiet and noise conditions showed a score of 80% and 65%, respectively [Table 1]. A month after surgery, steroid treatment was tapered down and treatment consisted of steroid sparing medications alone. These results were stable for 12- and 24-months follow-up.

DISCUSSION

There is little information regarding CI in GPA, with six reported cases of CI performed in patients with GPA. Two cases underwent subtotal petrosectomy and had good post-CI results, four did not undergo subtotal petrosectomy, of which one patient had good results and three had poor results. Our case highlights several points. CI can be effective in hearing rehabilitation of patients with GPA. Unlike chronic ear disease, which will usually be treated surgically before CI is performed, GPA can be treated medically. A course of prednisone with immunosuppressive therapy should be administered before surgery and steroids tapered down soon after surgery. A subtotal petrosectomy should possibly be reserved for patients in whom inflammation is severe and cannot be managed by medication. Surgical complications were not documented, and good audiological results were achieved.

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**The opposite of a correct statement is a false statement.
But the opposite of a profound truth may well be another profound truth.**

Niels Bohr (1885–1962), Danish physicist who made foundational contributions to understanding atomic structure and quantum theory, for which he received the Nobel Prize in Physics in 1922