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Paget's disease and cochlear implantation

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Abstract

Paget's disease of bone is a common disorder of unresolved etiology characterized by excessive bone resorption followed by excessive bone formation. If the skull is affected this may result in hearing loss and eventually develop into profound deafness. To date, no cases of cochlear implantation in patients with Paget's disease have been reported.

The authors present a case of radiographically confirmed Paget's disease of the skull in a 77-year-old man with a 20-year history of progressive bilateral sensorineural hearing loss who underwent cochlear implantation. A successful insertion of the Nucleus 24 Contour electrode array was achieved without surgical and post-operative complications.

At the 10 months' postoperative evaluation, the patient had gained useful open-set speech perception. In quiet conditions, his performance scores on the word and sentence recognition tests were 100 and 98 per cent, respectively. In the presence of noise (at +10 dB. signal-to-noise ratio), his performance scores on the word and sentence recognition tests were 96 and 94 per cent, respectively.

Key words: Cochlear implants; Osteitis Deformans

Introduction

Paget's disease (osteitis deformans) is a common bone disorder of unresolved etiology characterized by excessive bone resorption by osteoclasts followed by an increase in new and abnormal bone formation by osteoblasts. If the skull is affected, particularly the temporal bones, then hearing loss may result and eventually develop into profound deafness. The prevalence of hearing loss has been estimated to range from 30 to 50 per cent in all patients evaluated with Paget's disease of the skull.[1–3] Hearing loss in Paget's disease of the skull may be conductive, sensorineural or mixed.

Conductive hearing loss can be rehabilitated by means of otologic surgery or conventional hearing aids; the latter can also be helpful in the presence of a mild to severe sensorineural hearing loss (SNHL) but they are inappropriate if bilateral profound SNHL is present. In deafened patients in whom no benefits are obtained from conventional hearing aids, cochlear implantation is a highly effective technique in restoring hearing abilities.

Searching through the published literature using the Medline database, no information was available as to the feasibility and outcomes of cochlear implantation in subjects with Paget's disease. To the best of our knowledge, this is the first description of cochlear implantation in a patient with Paget's disease of bone.

Case report

A 77-year-old man presented with a 20-year history of progressive bilateral hearing loss associated with tinnitus, strongly motivated to verify if his deafness could be rehabilitated by means of a cochlear implant. The age of onset of hearing loss was 54 years, with a deterioration to right profound SNHL at the age of 60 years and bilateral profound SNHL at the age of 75 years. He had been fitted with a powerful postauricular hearing aid in the left ear at 75 years, but the rehabilitative effects had been weak.

At the time of admission, his medical history included medically controlled essential hypertension. He was otherwise well, reporting no other signs or symptoms. Otoscopic examination revealed a normal external auditory canal and a normal tympanic membrane in both ears. The remainder of the clinical and neurologic examinations was normal. Pure tone audiometry displayed a total deafness in the right side and a left-sided profound SNHL with a pure tone average (PTA) of 110 dB. Laboratory data disclosed a raised serum alkaline phosphatase (ALP) concentration (1035 IU/l; normal range 80–130 IU/l) and normal serum calcium and phosphate concentrations. None of his other blood levels were abnormal and electrocardiography showed no abnormal findings.

A computed tomography (CT) scan of the skull was performed, documenting pagetic involvement of the skull base and temporal bones with marked narrowing of the right internal auditory canal compared with slight narrowing on the opposite side (Figure 1). Demineralization of both otic capsules was also present. The anatomy of the middle ear was normal bilaterally. A magnetic resonance imaging (MRI) scan was undertaken to check the patency of the cochlear compartments and to

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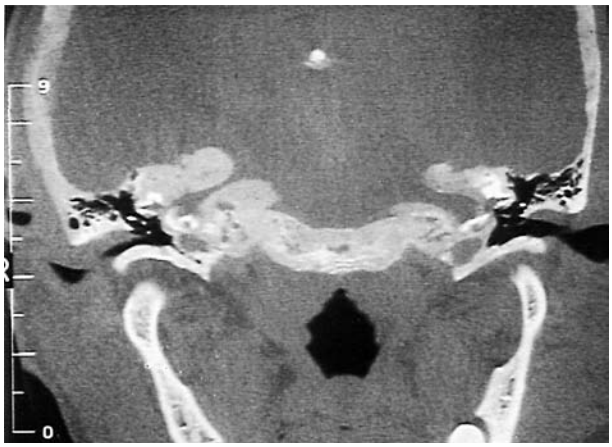

FIG. 1

Coronal CT section. Diffuse cotton wool appearance at the cranial base bilaterally with involvement of both temporal bones. Narrowing of the right internal auditory canal.

evaluate the neuroanatomy of the internal auditory canals (Figure 2). The elevated ALP concentration associated with the radiological findings confirmed the diagnosis of Paget's disease.

Transient-evoked otoacoustic emissions were absent. Auditory brainstem responses could not be detected at the highest levels of stimulation. Vestibular testing did not indicate any sign of peripheral or central vestibular impairment. Electrical promontory stimulation did not elicit auditory sensations in the right ear, although it showed good electric thresholds, dynamic range and temporal decoding capabilities in the left ear. Reliable electric auditory brainstem responses (EABRs) were also obtained using a transtympanic promontory electrode in the left ear.

Neuropsychological examination did not reveal disorders concerning intelligence or cognitive abilities; furthermore, the patient was realistic in his expectations and verified strong motivation and a normal psycho-social and family-related situation.

Since the selection program did not reveal any contraindication, we decided to implant the patient. The left ear was implanted with the Nucleus 24 Contour cochlear implant (Cochlear Corporation, Melbourne, Australia) in March 2003. The surgical procedure did not present any complication and a full insertion of the electrode array into the scala tympani was achieved. Intraoperative EABRs confirmed proper implant function and effective stimulus delivery. The postoperative course was uneventful. At the first postoperative control, 4 weeks after surgery, the wound was well healed and no signs of flap infection or necrosis were present. At the time of writing, with a follow-up of 10 months, no local or systemic complications had occurred. At 4 weeks postoperative, the patient received the Esprit 3G speech processor (Cochlear Corporation, Melbourne, Australia) programmed in the Spectral peak cooling (SPEAK) strategy with monopolar 1+2 mode.

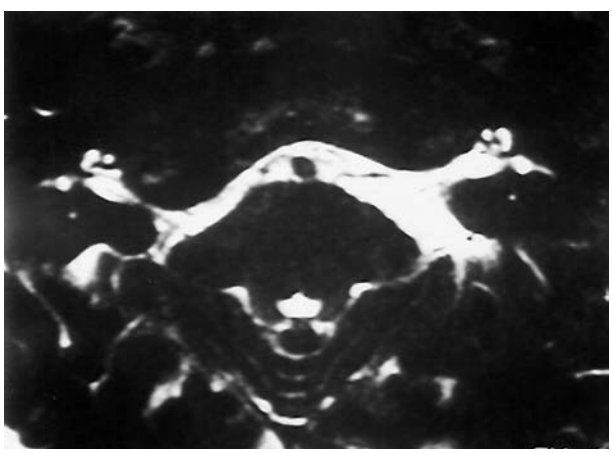

FIG. 2

Axial MRI showing an atrophic acousticofacial bundle on the right side.

Each stimulus had a pulse rate of 250 pulses per sec and a pulse duration of 25 μsec per phase. All the electrodes were activated without nonauditory, facial or pain sensation. At activation, values of electrode impedance ranged between 6.22 and 12.23 kΩ. Threshold levels (T levels) were set between 138 and 187 CL. Comfortable levels (C levels) were set between 169 and 211 CL.

Six months after initial stimulation, the patient continued to use the Esprit 3G speech processor programmed in the SPEAK strategy with monopolar 1+2 mode. Pulse duration was increased to 37 μsec per phase. Values of electrode impedance ranged between 4.67 and 9.71 kΩ. The T levels ranged between 167 and 202 CL and the C levels ranged between 201 and 221 CL. Increased electrical stimulation levels did not produce facial nerve stimulation and all the electrodes continued to be active. Therefore, we referred the patient to a rheumatology service and treatment with biphosphonate (risedronate) was started.

Speech perception was evaluated using word and everyday sentence speech recognition tests. An Italian version of the North-western University Phonetically Balanced Word List and the Central Institute for the Deaf Everyday Sentence List was used to measure speech perception benefits. The speech materials were presented in a hearing-only condition using a monitored live-voice through the sound field at an intensity level of 70 dB. sound pressure level (SPL). The patient was assessed with both materials in quiet conditions and in the presence of background noise at a signal-to-noise ratio (SNR) of + 10 dB.

Preoperatively, the word and everyday sentence recognition scores were 0 per cent.

In quiet conditions, the patient's performance scores were 83 per cent, 94 per cent and 100 per cent on the word recognition test at 1, 3 and 10 months after implantation, respectively. Everyday sentence scores at these time points were 75 per cent, 95 per cent and 98 per cent, respectively.

At +10dB. SNR, the patient's performance scores were 71 per cent, 86 per cent and 96 per cent on the word recognition test at 1, 3 and 10 months after implantation, respectively. At these time points, his everyday sentence scores were 60 per cent, 90 per cent and 94 per cent, respectively.

Discussion

Paget's disease of bone is the second most common metabolic bone disorder in the elderly, after osteoporosis. Paget's disease was first described by Sir James Paget in 1877 and is characterized by enhanced osteoclastic bone resorption followed by an excessive deposition of dense, disorganized and ineffectively mineralized bone matrix.[4,5] The prevalence of Paget's disease varies geographically. In western Europe, the prevalence has been estimated to be 3 per cent among individuals over the age of 40.[6,7] The disease can affect one bone (monostotic) or multiple bones (polyostotic). The most common sites for

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involvement are the pelvis, femur, lumbar spine, tibia, skull, humerus and forearm.[8] Following the pelvis, the skull is the second most commonly affected site.

The etiology of the disease remains unknown, although both genetic and environmental factors have been implicated. An autosomal dominant mode of inheritance was suggested by Morales-Piga *et al*. in 1995.[9] Recent studies point to genetic heterogeneity of the disorder. Susceptibility loci for Paget's disease have been mapped to chromosome 6q21-3 (PDB1), 18q21-q22 (PDB2), 5q35-qter (PDB3) and 5q31 (PDB4).[10] A viral etiology is suggested by the presence of paramixovirus-like inclusions in pagetic osteoclasts.[11,12] Other potential etiological factors include autoimmune, connective tissue and vascular disorders.

Typically, most patients affected by the disease are asymptomatic; only 10 per cent of patients are symptomatic. The most frequent complaints referable to the disease are bone pain, skeletal deformities and fractures. Sarcomatous degeneration in Paget's disease occurs in less than 1 per cent of patients.[8] Complaints of patients with skull involvement may include hearing loss, tinnitus, vertigo and a variety of cranial nerve compression syndromes.

Paget's disease is most commonly an incidental finding during an unrelated radiological or biochemical investigation. Alkaline phosphatase serum concentration analysis and the use of plain radiography remain the most frequent investigations to establish the diagnosis of Paget's disease. Computed tomography and MRI scans provide useful information for assessing the complications of Paget's disease, such as arthritis, fracture, neoplastic degeneration, spinal involvement and neurologic compromise.

Involvement of the temporal bones may result in a conductive, sensorineural or mixed hearing loss. The cause of hearing impairment associated with Paget's disease of the skull remains unclear. Possible causes of conductive hearing loss include external auditory canal stenosis, tympanic cavity fibrosis and ossification, incus or malleus fixation, stapes fixation and round window obliteration.[13] The mechanism of sensorineural hearing loss may be related to loss of bone mineral density in the cochlear capsule,[14] hair cell damage[15] and progressive internal auditory canal stenosis with compression of the eighth cranial nerve.[16]

In our patient, the cause of hearing loss may be related to both the loss of bone mineral density in the cochlear capsule (replaced by pagetic bone) and the progressive narrowing of the internal auditory canals, with subsequent compressive cochlear neuropathy.

The most widely used treatment for Paget's disease is inhibition of bone resorption using the newer bisphosphonates such as alendronate and risedronate.[17] The efficacy of antipagetic therapy on the development or progression of deafness has not been established. However, most authors recommend bisphosphonate therapy in symptomatic patients and in those asymptomatic patients who are likely to develop complications due to the site of the disease.[8,17] In our patient the diagnosis of Paget's disease was made in the late phase of the disease when profound hearing loss was already present. The patient had never received antipagetic treatment before cochlear implantation. Six months after his initial activation, he has required a slight increase in current to elicit auditory responses. This may indicate a progression of the disease, with pagetic activity in the otic capsule. Antipagetic therapy with biphosphonate was then started.

Ten months after implant activation, the patient developed excellent open-set speech recognition skills and was also able to converse on the telephone without a code. Describing the benefits received from the 10-month use of his cochlear implant, our patient reported feeling more independent, more socially involved and even less lonely. Although significant conclusions cannot be formulated from a single case, the results obtained in the reported patient are encouraging. Cochlear implantation allowed this patient to regain social hearing and dramatically improved his quality of life, increasing his self-confidence, independence and social integration.

- **Paget's disease results in resorption and new bone formation, which may cause hearing loss if the skull is affected**
- **This is the first report of successful cochlear implantation in a patient with this disease**

Conclusion

Paget's disease of bone is characterized by enhanced osteoclastic bone resorption followed by an excessive deposition of dense, disorganized and ineffectively mineralized bone matrix. The most common sites for involvement are the pelvis, femur, lumbar spine, tibia, skull, humerus and forearm. Following the pelvis, the skull is the second most commonly affected site. If the skull is affected this may result in hearing loss and eventually develop into profound deafness.

To our knowledge, cochlear implantation in Paget's disease has not been previously reported. We report a male patient with a 20-year history of progressive bilateral sensorineural hearing loss and radiographically confirmed Paget's disease of the skull who was succesfully implanted with the Nucleus 24 Contour cochlear implant.

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