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Clinical Records

Osseo-integration in Paget's disease: the bone-anchored hearing aid in the rehabilitation of Pagetic deafness

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Abstract
The first report of a patient with gross Paget's disease and progressive hearing loss who successfully underwent hearing rehabilitation with an osseo-integrated hearing aid is presented. The otological manifestations of Paget's disease and the principles of osseo-integration are discussed. The use of a bone-anchored hearing aid (BAHA) in selected patients with Paget's disease can provide useful amplification and hearing rehabilitation.

Key words: Osseointegration; Paget's Disease; Hearing Aids

Introduction

Hearing loss in Paget's disease is found in 30–50 per cent of patients with skull involvement.[1,2] The most frequently encountered type of hearing loss is a mixed deafness which is progressive and often symmetrical.[3]

Conventional aiding generally provides effective rehabilitation of hearing in these patients, however, there are limitations. In the following section we discuss a case of a patient with gross Paget's disease who derived minimal benefit from conventional aiding and was successfully treated with a bone-anchored hearing aid (BAHA). This is, to the best of our knowledge, the first such report in the literature. A brief review of relevant literature is also presented.

Case report

A 72-year-old man with a known history of Paget's disease and progressive hearing loss was referred to the otolaryngology department. His Paget's disease had been diagnosed some 30 years previously. He had been using bilateral in-the-ear body-worn aids, but found them increasingly ineffective as his hearing loss progressed. His problems were compounded by bilateral chronic otitis externa.

Clinical examination revealed an elderly male with the classical features of Paget's disease; kyphosis of the spine, shortened neck, bowing of the tibia, enlarged skull vault with collapse of the back and sides of the skull and marked leontiasis ossea (protrusion of the upper jaw) (Figure 1). He had bilateral otitis externa with gross narrowing of the external auditory meati.

Pure tone audiometry demonstrated a bilateral mixed hearing loss with bone conduction thresholds between 40–90 dB and a conductive loss varying between 60 dB in the lower frequencies and 30 dB in the higher frequencies

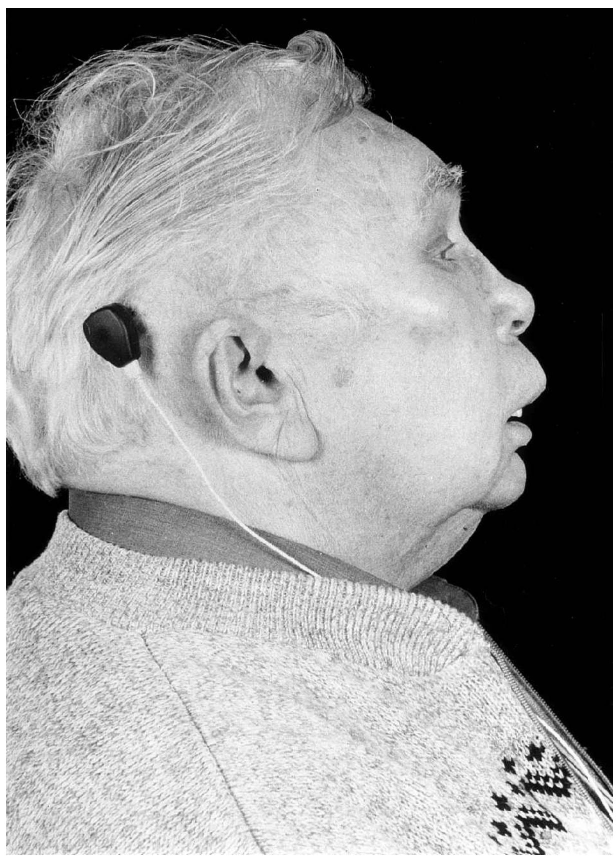

Fig. 1
Lateral profile of patient. Note the protrusion of the upper jaw (leontiasis ossea) and BAHA in-situ.

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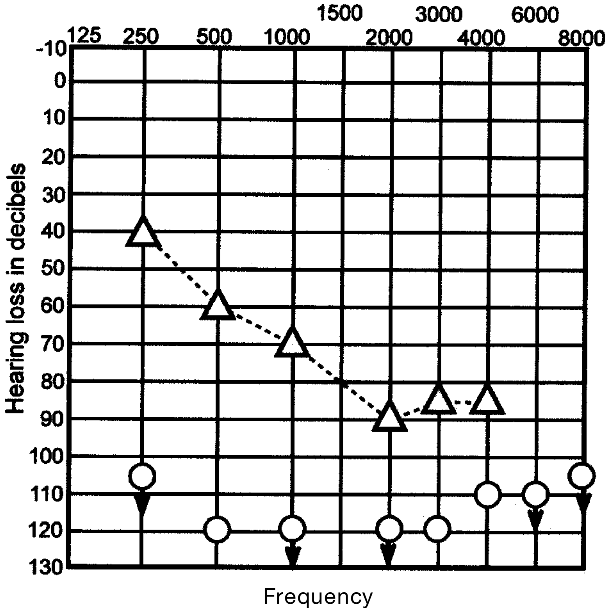

FIG. 2
Pre-operative audiogram.

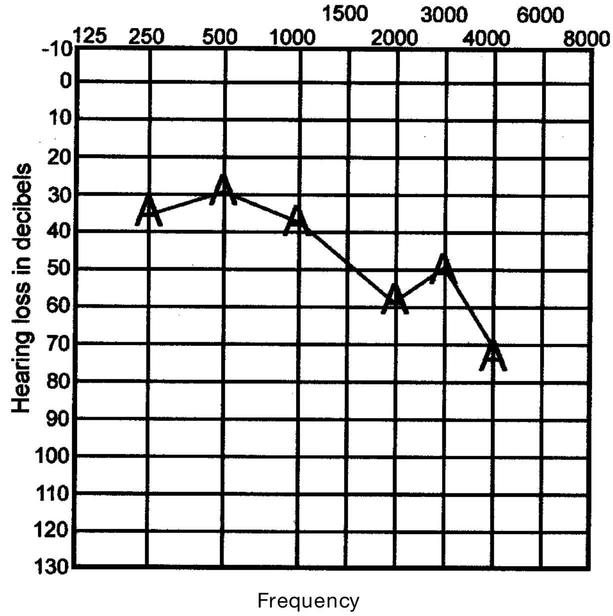

FIG. 4
Post-operative audiogram. A: Aided hearing thresholds.

(Figure 2). Computerized tomography (CT) of the skull revealed changes consistent with gross Paget's disease (Figure 3).

He was assessed for a BAHA and subsequently a 4 mm titanium fixture was inserted into the right temporal bone. The bone was found to be particularly soft and vascular so the procedure was performed in two stages with the abutment being fitted after three months.

The patient was fitted with a HC220 Superbass BAHA one month after the second stage procedure, that was used consecutively with bilateral body-worn aids. An immediate improvement in his hearing was noted with aided hearing thresholds varying between 30–60 dB (Figure 4).

Discussion

The clinical and pathological features of Paget's disease (or osteitis deformans) were described by Sir James Paget in 1877.[4] The condition is characterized by spreading osteolytic and osteoblastic changes, most frequently affecting the pelvis, lumbar spine, skull, femur and tibia.[2]

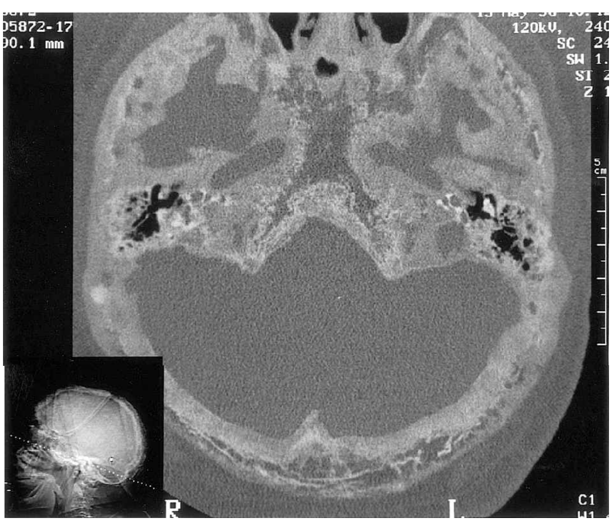

FIG. 3
Axial CT scan demonstrating gross Paget's disease affecting the whole skull base and encroaching upon the labyrinthine capsule bilaterally.

The overall incidence of Paget's disease is three per cent, however, this figure steadily increases to 10 per cent by the eighth decade.[5] The clinical onset of the disease is uncommon before the fourth decade, it affects men four times more often than women. Most cases are thought to be sporadic, but in 15 per cent of cases there is an autosomal dominant inheritance pattern.[1]

The exact aetiology of Paget's disease is unknown. The detection of nuclear viral inclusions in osteoclasts suggests a viral aetiology, prompting some authors to hypothesize that the disease may result from a slow viral infection of osteoclasts.[6]

The histopathological changes result from alternating waves of osteoclastic and osteoblastic activity producing haphazard bone resorption and deposition of structurally weak, demineralized cancellous bone. Bone turnover is increased 20 fold or more in diseased areas, unaffected bone is normal and remains normal. Initially bone resorption dominates and produces lytic lesions, subsequently the marrow spaces are filled with fibrovascular tissue that later become sclerotic.[2] The ultimate outcome is bone which is soft, with a tendency to fracture and deformity.

Skull involvement in Paget's disease occurs in 65–70 per cent of advanced polyostotic cases.[7] Hemifacial spasm occurs in seven per cent of patients and trigeminal neuralgia in six per cent of patients, the aetiology is thought to be secondary to neural foraminal encroachment.[8] Otological manifestations include tinnitus and vertigo which are seen in 32 per cent and 36 per cent of patients respectively.[9] There have been no reports in the literature of facial nerve dysfunction due to Paget's disease.

The pattern of hearing loss in Paget's disease is predominately mixed. The conductive loss is pronounced in the lower frequencies and the sensorineural loss in the higher frequencies. There is, therefore, a progressively decreasing magnitude of conductive loss from low to high frequencies.[3] Baraka[10] found that the average rate of

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TABLE I
OTOLOGICAL INDICATIONS

(1) Congenital:	Conductive/mixed loss especially with external ear canal atresia and malformations making the fitting of conventional behind the ear aids impossible
(2) CSOM:	Bilateral radical mastoidectomy cavities, persistent discharge, problems associated with conventional aiding, constant discharge with ear mould and feed-back in radical mastoidectomy cavities
(3) Otitis externa:	In those requiring conventional aiding
(4) Otosclerosis:	In patients unwilling, unable or unsuitable for stapedectomy and unable or unwilling to wear conventional aids

hearing loss in Paget's disease was 2 dB per annum higher than the expected 0.5 dB per annum in a normal matched group.

The exact cause of hearing loss in Paget's disease remains unclear. A number of pathological findings have been reported in the literature in an attempt to explain the hearing loss. In conductive losses they are: external auditory canal stenosis, tympanic membrane abnormalities, tympanic cavity fibrosis and ossification, incus or malleus fixation, stapes fixation and round window niche obliteration. In sensorineural hearing loss; hair cell depopulation, arteriovenous shunts, otic capsule microfractures, internal auditory canal (IAC) stenoses, microneuromata and acoustico-facial bundle elongation have been implicated.[3]

Khertarpal and Schuknecht,[3] in one series, examined 26 Pagetic temporal bones. They could not detect any consistent findings to explain conductive or sensorineural hearing loss in Pagetic patients. They concluded that the hearing loss is caused by changes in bone density, mass and form that serve to dampen the finely tuned motion mechanics of the middle and inner ear. Proops *et al.*[11] found in their series that the effects of Paget's disease on the middle-ear structures was more variable than its extension into the otic capsule. Monsell *et al.*[12] studied the relationship between hearing loss and IAC diameter in Paget's patients, but no significant relationship was found. They concluded that the hearing loss in associated with-intact auditory nerve function and suggest a cochlear site of lesion. This was confirmed in another study revealing no evidence of auditory nerve dysfunction and a cochlear site of lesion. Loss of bone mineral density in the cochlear capsule was found to be associated with both high-tone hearing loss and a low-tone air-bone gap.[13]

The treatment of symptomatic Paget's disease is almost exclusively medical. However, its effect on hearing loss is variable. Calcitonin acts by inactivating osteoclasts and thus leads to a reduction in bone resorption. Its use for hearing loss in Paget's disease has had variable success in the past.[14–16] Sodium etidronate and other biphoshonates inhibit calcium deposition and appear to have selective cytotoxicity for osteoclasts. Combined calcitonin and sodium etidronate therapy has been shown to stabilize and even reverse hearing loss in two patients with Paget's disease.[17] Overall the use of these agents in the treatment of hearing loss is debatable, some of which are not devoid of risk and associated with significant side-effects.

It is generally agreed that the results of reconstructive middle-ear surgery in Paget's disease are unsatisfactory.[18] Davies[9] reviewed the outcome of stapedectomies in patients with conductive hearing loss and found disappointing persistent air-bone gaps with variable patient benefit. This may be explained by the fact that there is no consistent defect responsible for the conductive loss, also the presence of a co-existing sensorineural hearing loss often mitigates against a successful outcome.

Modern hearing aids are capable of excellent amplification and can provide good rehabilitation for hearing loss patients. However, hearing augmentation in the setting of chronic otitis externa together with severe mixed hearing loss as in our case can be challenging. The benefit obtained is often minimal because conventional hearing aids lack sufficient fidelity, comfort and positional stability to effectively address the communication needs of such patients. These limitations can be overcome by the use of a bone conductor anchored to the temporal bone by osseointegration.

Osseo-integration implants were first introduced by Brånemark in 1965 for dental implants.[19] The use of titanium implants in the temporal bone for the attachment of BAHAs was developed by Tjëllström in Sweden in the late 1970s.[20] Sound transmission by direct bone conduction is achieved by using a skin-penetrating coupling from an osseo-integrated implant in the temporal bone to an impedance matched transducer that the patient can apply and remove at will.[21] The end product of osseo-integration is implant-bone anchorage. The absence of interposed soft tissues gives better quality sound, requires less energy and offers much greater comfort.[22]

Several definitions of osseo-integration have been proposed since Brånemark first coined the term in 1977. A recent definition states 'an implant is said to be biomechanically osseo-integrated if there is no progressive relative motion of living bone and implant under functional levels and types of loading for the entire life of the patient'.[23] Albrektsson *et al.*[24] identified the following important pre-requisites for osseo-integration; implant material, implant design, implant finish, status of bone, surgical technique and implant-loading conditions.

Bone morphology, bone quality and other local conditions of the recipient site have been recognized as factors affecting implant stability, position and successful osseo-integration.[25,26] Healing and bone formation at the implant-tissue interface may be prolonged in tissues where there is a more open trabecular network.[27] In cases of poor bone quality, the two stage procedure with fitting of the titanium abutment at a later date is advocated.[28]

There is no available data on osseo-integration in Pagetic bone. In view of the rapid bone turnover in diseased areas one would assume that osseo-integration would be unpredictable and sub-optimal. This poses a dilemma for the surgeon and patient. However, our patient responded well to surgery, the titanium fixture did osseo-integrate and at review showed no sign of loosening or instability.

The otological and audiological indications for the BAHA are shown in Tables I and II respectively.[29] There are few contra-indications to providing a patient with a BAHA. It is important that the patient understands

TABLE II
AUDIOLOGICAL INDICATIONS

(1) Average BC thresholds <40 dB = ear level BAHA (HC200/HC300)
(2) Average BC thresholds <60 dB = bodyworn superbass BAHA (HC220)
(3) Speech discrimination >60%
(4) Realistic expectations
(5) Suitable domestic support

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the concept of the BAHA. Patients with psychiatric disease, immature personality and a history of drug abuse have a high failure rate.[30] The patient should not have a poor cochlear reserve, for the body-worn HC220 the bone conduction thresholds should not exceed 60 dB.

Our patient was within the otological indications for a BAHA but did not strictly fulfill the audiological criteria. However, he was fitted with a conventional bone conduction hearing aid during his assessment. He noted a definite improvement in sound quality and so it was thought that a BAHA would be beneficial.

The patient had been using body-worn air conduction aids for many years but now found the level of amplification inadequate. He also complained of troublesome chronic otitis externa, that was exacerbated by the use of conventional aids. As a result he found himself socially isolated and increasingly withdrawn.

Cochlear implantation was deemed unlikely to be beneficial due to the continuing Pagetic activity in the otic capsule and the surgical difficulties posed. It was thought in view of the above history that the most appropriate management option was the use of a BAHA.

He underwent successful surgery with the fixture osseo-integrating. Improvement in hearing thresholds were noticed immediately on using the BAHA. His hearing did deteriorate further a few years later and a more powerful Megabass processor was used to increase amplification with a satisfactory outcome. We believe this is the first case of a patient with Paget's disease to be fitted with an osseo-integrated hearing aid. The use of a BAHA in carefully selected motivated patients with Paget's disease can provide useful amplification and rehabilitation.

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