Case Report

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Magnetic Resonance Imaging and High-Resolution Computed Tomography in the Otospongiotic Phase of Otosclerosis

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Key Words
MRI · High-resolution CT · Petrous bone · Otosclerosis · Otospongiosis

Abstract
Otosclerosis very often leads to severe hearing loss in a chronic progressive manner. In the first phase of the disease, otospongiosis causes an inflammatory osteolytic process in the osseous labyrinthine capsule. In the cases reported here, this osteolytic process was pronounced in the osseous capsule of the cochlea. High-resolution CT and MRI showed the precise localization and stage of this inflammatory-osteolytic process in the petrous bone, while scintigraphy confirmed the diagnosis.

Introduction

Otosclerosis is the cause of about 2% of all cases of severe hearing loss between the ages of 30 and 59 years. Otosclerotic lesions are found in about every eighth histologically examined petrous bone in women and about every fifteenth in men [1–3]. Hormonal differences might be the reason for this difference, but clear racial distinctions are also found.

It is assumed that every tenth case of otosclerosis with histologic manifestations leads to clinical deficits (conductive or mixed hearing loss) [4].

Regarding the etiology of otosclerosis, there are a number of hypotheses, genetic and metabolic factors have been discussed, but many arguments have been found for an organotropic measles virus infection in the labyrinth causing changes in superficial antigens of mesenchymal cells, which in turn could lead to an immunologic response. The result might be a chronic inflammation with an initial osteolytic phase called otospongiosis. In the further course of the disease, otosclerosis in a narrow sense must be interpreted as a kind of postinflammatory scarring [5, 6].

Case Reports

Case 1

A 29-year-old woman presented to the ENT Department with bilateral chronic progressive hearing loss over several years. Audiometry showed bilateral mixed, but predominantly sensineural, hearing loss; no vestibular signs were found.

MRI examination of the head was performed, focussed on the posterior fossa and temporal bone. T2-weighted images showed an amorphous increase in signal intensity in both temporal bones in the region of the labyrinth. In the same area on T1-weighted scans, a hyperintense signal was also found, and strong enhancement was seen after administration of gadolinium-DTPA. The cerebellopontine angle cistern and internal auditory canal showed no pathologic findings, and no signs of inflammatory disease were found in mastoid cells (fig. 1–3).

On high-resolution CT of the temporal bone, demineralization of the pericochlear osseous capsule was found; the cochlea itself was poorly defined.

Similar lesions of a lesser degree were depicted around the vestibule but not in the region of the semicircular canals.

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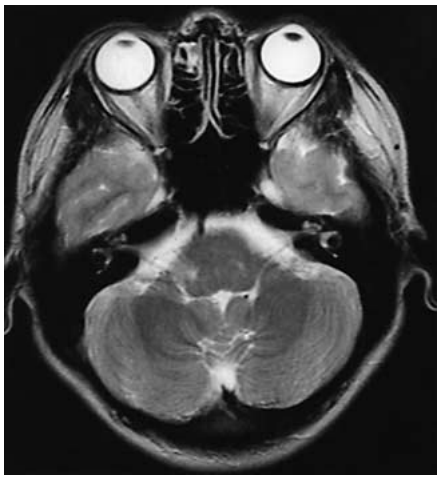


Fig. 1. Axial T2-weighted MRI scan (TR 3692, TE 96) depicts increased signal intensity in the osseous labyrinthine capsule of both temporal bones.

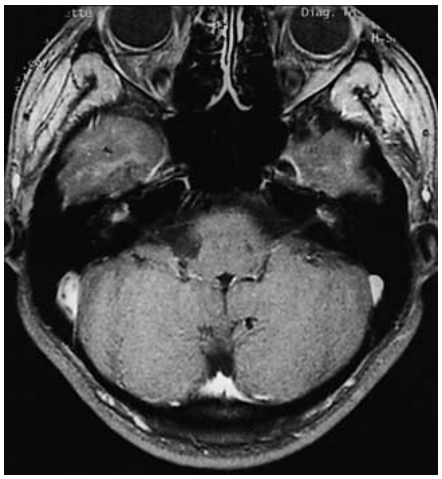


Fig. 2. Axial T1-weighted MRI scan (TR 286, TE 18) after intravenous gadolinium-DTPA shows irregular enhancement in the temporal bone around the cochlea on both sides.

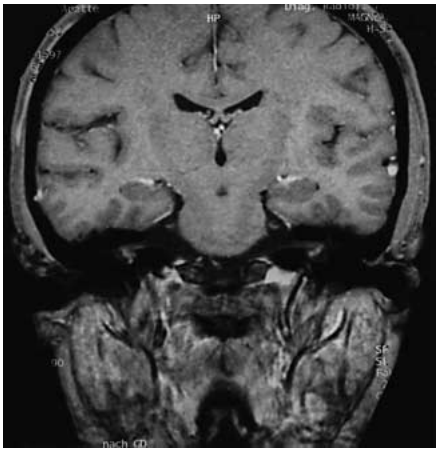


Fig. 3. Postcontrast coronal T1-weighted MRI scan (TR 286, TE 18) confirms the finding of pericochlear enhancement.

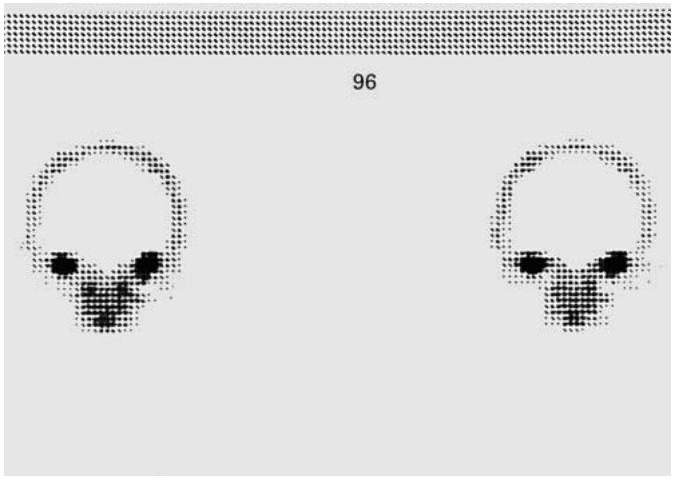
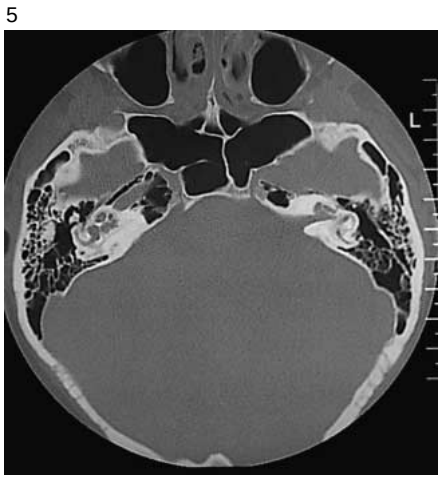


Fig. 4. SPECT with 800 MBq of ^{99m}Tc-MDP shows intense radioisotope uptake in both temporal bones in the region of the labyrinthine capsule.



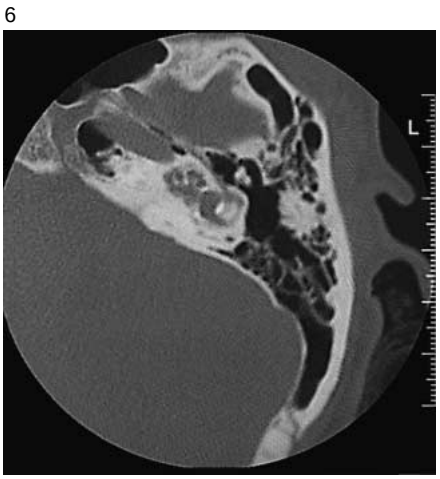


Fig. 5. High-resolution CT scan of both petrous bones shows demineralization in both labyrinthine capsules with irregular margins of the cochlea and to a smaller extent around the vestibule.
Fig. 6. Magnified high-resolution CT scan of the left petrous bone clearly depicts pericochlear demineralization.

Case 2

A 53-year-old female patient first came to the ENT clinic a few years ago with severe bilateral sensineural hearing loss near to deafness. There was a long history of different surgical interventions in both middle ears because of otosclerosis; the last operation was a stapes prosthesis on both sides. A few years after this operation, rapid progressive sensineural hearing loss began. High-resolution CT showed diffuse cochlear otospongiosis; scintigraphy of the petrous bone confirmed this diagnosis by demonstrating foci of high activity in the region of the osseous labyrinthine capsule (fig. 4).

High-dose prednisolone therapy led to an improvement of sensineural hearing, but recurrent episodes of disturbed inner ear function meanwhile caused complete deafness. Cochlea implant surgery is planned.

Discussion

In the otospongiotic stage of otosclerosis, osteolysis occurs in the osseous labyrinth, and the margins of the capsule appear more and more irregular on high-resolution CT scans, culminating in some cases in extensive demineralization of the whole labyrinthine capsule [1].

Because of this progressive decalcification, contrast between the cochlea itself and the bony structures of the otic capsule is lost (fig. 5, 6].

In the subsequent phase of sclerosis, new ossification leads to more or less pronounced thickening of the labyrinth's capsule, so histologically an irregular pattern of condensation can be seen. Simultaneous appearance of otosclerotic and otospongiotic foci is possible.

The present cases demonstrate typical constellations of CT and MRI findings in otospongiosis of the labyrinth's capsule [7–9]. The center of these otospongiotic changes lies in the region of the cochlea [5, 10, 11]. Lack of sclerotic changes shows that otospongiosis, considered as an early stage, obviously can persist for a long time without any sclerosis. Histologic appearance with growing capillary vessels, osteoclastic activity and progressive demineralization [5] finds its correlation in the osteolytic zones of the osseous labyrinth with production of new immature bone [12, 13].

Only anecdotal reports [14] were found on the phenomenon of pericochlear enhancement after administration of contrast medium. In the present cases, MRI examination showed distinct enhancement around the cochlea, well documented in the axial and coronal plane. The pathophysiologic basis of this enhancement is an active inflammatory process; lymphocytes as well as specific antibodies are detected in otospongiotic foci [5].

These inflammatory characteristics with active osteolysis and consecutive hypermetabolism make it possible to show otospongiotic foci also with scintigraphy of the temporal bone [5]. In differential diagnosis, primary bone diseases like osteogenesis imperfecta and, to a lesser degree, fibrous dysplasia and Paget's disease must be considered [15], but these diseases mostly show a plurifocal pattern; unspecific inflammatory effects ('labyrinthitis ossificans') should also be considered [16].

We conclude that high-resolution CT and particularly contrast-enhanced thin-section MRI of the posterior fossa and temporal bone are reliable instruments in the diagnosis of otospongiosis.

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