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Pathology of labyrinthine ossification

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Abstract

Ossification of the inner ear is the result of multifactorial pathogeneses, such as infection or malignant infiltration, and otosclerosis. Ossification of the inner ear spaces is a well documented sequela of suppurative labyrinthitis.

In this study of human temporal bones, sections from 14 patients (28 temporal bones) were studied. In addition to the osseous tissue within the inner ear, findings included neoplasms, otosclerosis, otitis media, trauma, and Fabry's disease. We have attempted to correlate these conditions and their influence on the formation of osseous tissue within the spaces of the inner ear. Tympanogenic infection and vascular compromise were found to play an important role in ossification. The scala tympani of the basal turn of the cochlea was frequently the site involved.

Introduction

Pathological ossification of spaces in the membranous labyrinth is a response to processes destructive to the membranous labyrinth or the endosteum of the otic capsule. The diagnosis has primarily been made histologically. Recently, however, multidirectional tomograms and computerized tomographic (CT) scans have made it possible to identify and diagnose this condition radiologically (Hoffmann *et al.*, 1979; Becker *et al.*, 1984; Swartz *et al.*, 1985). Ossification within the cochlea and labyrinth is most often the result of infections that may reach the inner ear by haematogenous, tympanic, or meningeal routes. Other aetiological factors such as otosclerosis, tumours, allergy, and trauma have been postulated to cause ossification within the inner ear. The exact mechanism by which this occurs is not always clear.

Ossification in the inner ear has currently become clinically relevant in those patients who are candidates for cochlear implants. Reports in the literature stress the need for detection of the ossification by use of complex motion tomography and CT-scanning (Hoffmann *et al.*, 1979; Becker *et al.*, 1984). In this study we have kept this added clinical relevance in view and tried to determine the significance that ossification in the inner ear would have in patients contemplating cochlear implants. Our series evaluates human temporal bone sections in which ossification was detected. Implications regarding origin of the ossification and its effect on the anatomical and functional status of the inner ear are analysed and discussed.

Methods and materials

Microscopic examination of sections from 1,383 human temporal bones was performed to detect those with pathological ossification within the inner ear. Sections of the temporal bone 18 to 20 microns thick were cut horizontally from superior to inferior and every tenth section retained for examination and stained with haematoxylin and eosin.

Results

Twenty subjects were observed to display evidence of ossification within the inner ear. Of these, only the 14 patients whose left and right temporal bones were available were included in this study. There were eight men and six women, ranging in age from 29 to 82 years with a mean age of 60 years. Bilateral evidence of ossification was seen in five subjects (36 per cent) and unilateral in nine (64 per cent).

The causes of ossification in the patients from our study are listed in Table I. Many of these patients showed evidence of multifactorial causes responsible for ossification; however after review of history and temporal bone sections, they were listed under the cause deemed likely to have initiated the process. Factors responsible for ossification were chronic otitis media (Fig. 1a, b) in three subjects (21 per cent), otosclerosis (Fig. 2) in three (21 per cent), trauma with chronic otitis media in two (14 per cent), trauma in one (7 per cent), neoplasms (Fig. 3a, b) in four (29 per cent) and angio-

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TABLE I
AETIOLOGIES AND SITES OF OSSIFICATION WITHIN THE INNER EAR IN TEMPORAL BONES FROM 14 PATIENTS

Age	Sex	Etiology	Ossification	Site of Ossification
50	M	Bilateral chronic otitis media (COM)	Bilateral	LEFT: ST SV, T1, vest, ES. RIGHT: SV, T1, vest
73	F	Scar Fever and COM	Unilateral	LEFT: cochlea, vest, sup post-lat SCC (completely obliterated)
82	M	Mumps, Measles, COM	Unilateral	LEFT: sup SCC
68	M	Trauma and COM	Bilateral	RIGHT: post SCC LEFT: ES
65	F	Trauma and COM	Unilateral	RIGHT: lat SCC
42	F	Trauma	Unilateral	RIGHT: lat SCC
70	M	Acoustic neuroma and surgical trauma	Unilateral	RIGHT: both limbs of post SCC
75	M	Metastases	Unilateral	RIGHT: lat SCC
49	F	Leukemia	Bilateral	RIGHT: sup-post-lat SCC LEFT: ST, SV, T1, Lat SCC
29	M	Leukemia	Bilateral	RIGHT: ST, T1, post SCC LEFT: ST, T1, vest
77	F	Otosclerosis	Unilateral	RIGHT: ST, SV, T1
77	M	Otosclerosis	Unilateral	RIGHT: ST, T1
78	F	Otosclerosis	Bilateral	RIGHT: ST, SV, T1 LEFT: ST, SV, T1, ES
48	M	Fabry's disease	Unilateral	RIGHT: sup SC

ST = scala tympani, SV = stria vascularis, T1 = first cochlear turn (Basal turn of the cochlea), vest = vestibule, ES = endolymphatic sac. sup = superior, post = posterior, lat = lateral, SCC = semicircular canals.

keratoma corporis diffusum (Fabry's disease) in one (7 per cent).

Discussion

Ossification occurring within the inner ear is most often the sequela of an inflammatory response. Becker *et al.* (1984) have described infection as the most common cause of inflammation within the inner ear. The route of the infection can be either meningeal, haematogenous or tympanic in origin.

Meningitis has been identified as a cause of infection within the inner ear. The infection travels from the subarachnoid spaces via the cochlear aqueduct (Bhimani *et al.*, 1984) and internal acoustic meatus (Paparella and Sugiura, 1967; Sugiura and Paparella, 1967) into the inner ear, where it results in suppurative labyrinthitis.

Although ossification can take place following meningeal infections in childhood (Altmann and Waltner,

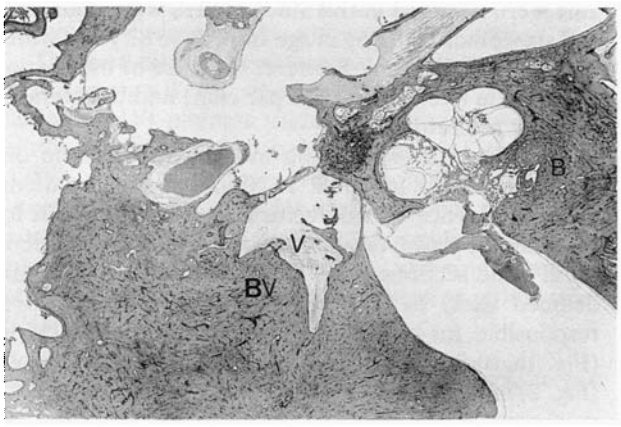

FIG. 1a

Section of left temporal bone of 68-year-old woman who had had scarlet fever as well as chronic otitis media. The anterior cochlear basal turn is almost obliterated with new bone. Ossification (B) is seen in the scala vestibuli of the posterior cochlear basal turn. All semicircular canals are completely obliterated. New bone (BV) occupies almost the entire vestibule (V). A small otosclerotic focus is seen just below the processus cochleariformis, anterior to the stapedial footplate. H&E stain ×60.

FIG. 1b

Section of right temporal bone from the same patient, demonstrating a virtually normal inner ear. H&E stain ×60.

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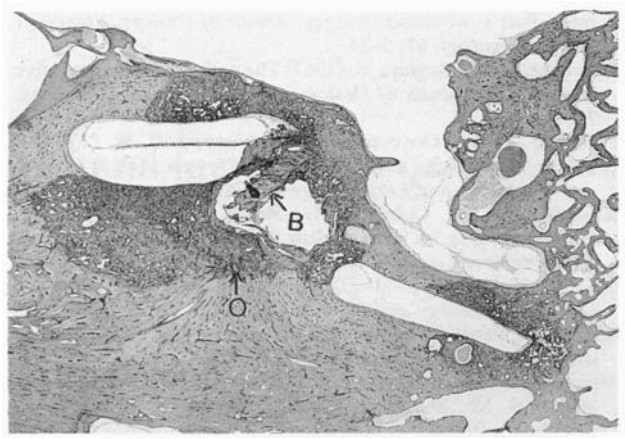

FIG. 2

Section of left temporal bone from a patient with evidence of massive otosclerosis (O). The region of the round window is completely obliterated by otosclerotic bone (B). Bone seen within the scala tympani is not otosclerotic. H&E stain ×60.

1944), the age-range in this study was 29 to 82 years, with a mean age of 60, with none of the patients having a history of meningitis.

Five patients were found to display evidence of tympanogenic infection. Two had evidence of surgical trauma that most likely provided the infection direct access to the inner ear. However, in two patients the otic capsule was found to be intact. Two patients (3 temporal bones) with tympanogenic infections demonstrated ossification in the basal turn of the cochlea near the membrane of the round window. In one patient, suffering from chronic otitis media, the stapedial footplate was eroded by the disease.

Paparella *et al.* (1980) have documented changes that occur in the inner ear as a result of toxins from otitis media that pass through the round window membrane. In nine temporal bones of patients other than those of otitis media in our study, fluid and granulation tissue were found in the round window niche and the round window membrane was found to be thickened. Ossification occurred within the basal turn of the cochlea, indicating the possibility that the round window membrane was the portal of entry.

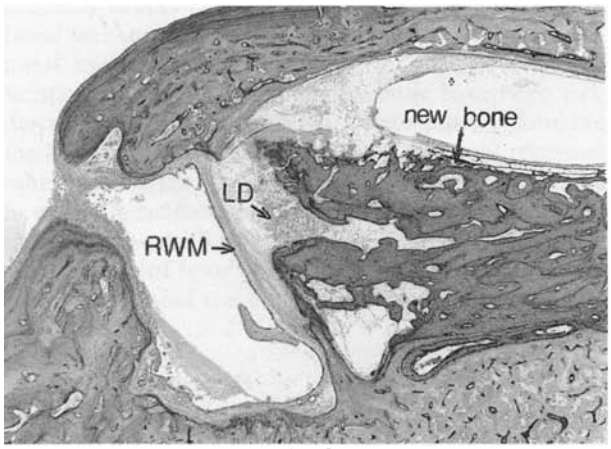

FIG. 3a

Section from the right temporal bone of a patient who had suffered from leukaemia. Ossification (new bone) and the leukaemic deposits (LD) can be seen in the region of the scala tympani near the round window. The round window membrane (RWM) is thickened. H&E stain ×90.

It has been postulated by some investigators (Suga and Lindsay, 1977) that in pathological conditions such as otosclerosis and tumours, ossification is caused by invasion of the inner ear. In three cases of extensive otosclerosis in our study, despite the nearly symmetrical involvement of the otic capsule, only one case demonstrated bilateral evidence of ossification; in all cases the endosteum was involved. It is now of clinical relevance in those patients about to receive a cochlear implant. This is especially true with intracochlear implants, as ossification may obstruct the proper positioning of the electrodes and impair successful stimulation.

Radiographic imaging is important in determining which side is affected and can determine which side is suitable for a prosthesis. In our study, bones from five patients showed evidence of bilateral ossification. Rosenberg *et al.* (1987) have encountered such cases of bilateral ossification and brought them to be not entirely suitable for cochlear implants. Ibrahim and Linthicum (1980) have found dense ossification a contraindication for these prostheses, but they do not exclude those cases in which mild ossification is present. The present study shows that when ossification occurred in the cochlea it was accompanied by degenerative changes in the stria vascularis, the organ of Corti and endolymphatic hydrops. Such patients would therefore fail to be candidates for a cochlear implant.

Conclusions

(1) Ossification within the inner ear spaces was found to occur as a result of multiple factors working through various mechanisms. Infection and compromised vascularity were thought to explain much of the origin of formation of bone in the inner ear.

(2) The endolymphatic spaces were most often spared and formation of bone occurred in the perilymphatic spaces.

(3) The region of the scala tympani near the round window was the most common site for ossification.

(4) The stria vascularis, organ of Corti and ganglion cells were found to demonstrate marked evidence of degeneration.

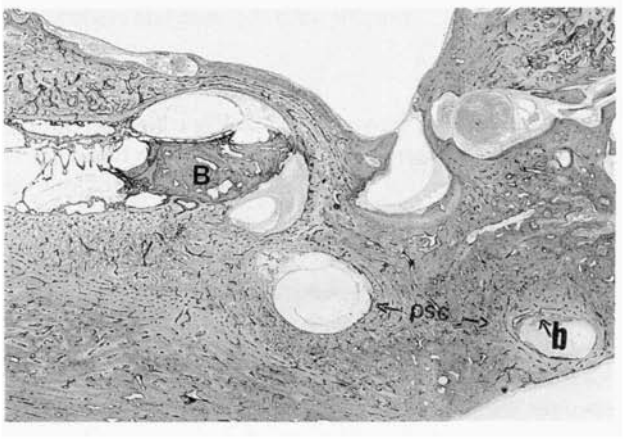

FIG. 3b

Section from the left temporal bone from the same patient, with evidence of bone (B) in the scala tympani at the basal turn of the cochlea and bone (b) in the posterior semicircular canal (psc). H&E stain ×60.

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