

Otosclerosis and cochlear otosclerosis: A post mortem study on temporal bones

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Abstract. – We have chosen among many temporal bones of donors deceased individuals with concomitant otosclerosis, three particular cases, one with classic otosclerosis, another with cochlear otosclerosis with concomitant oval window ankylosis and another with cochlear otosclerosis without stapes fixation. The different histopathologic features are discussed and clinical and therapeutical guidelines are proposed.

Key Words:

Temporal bone, Histology, Conductive hearing loss, Cochlear, Genetics, Otitis media.

Introduction

Otosclerosis is a common focal lesion of the otic capsule of unknown aetiology, which is found principally in relation to the cochlea and footplate of the stapes. Otosclerotic deposits, not associated with hearing loss, are found in about 10% of all adult temporal bones at autopsy of white people¹. Otosclerosis usually affects both ears symmetrically. The lesion attacks mainly Caucasians and is said to be unusual in Negroes and Mongolian people. The disease process is probably confined to the temporal bone although some evidence of a general connective tissue disturbance has been put forward²; a similar bony change often appears, however, in a position similar to that of otosclerosis, in the generalized bone disease *osteogenesis imperfecta*³⁻⁵.

Materials and Methods

We have studied the histopathological changes in temporal bones of 3 deceased individuals, with concomitant chronic otitis media.

These patients were donors and agreed during their life to donate *post mortem* their temporal bones to the House Ear Institute Los Angeles CA, USA as a contribution to a better knowledge of temporal bone diseases.

We have removed the temporal bones in our usual way⁶.

Results

Gross appearances

In cases with prominent otosclerotic involvement of the otic capsule the lesion may be seen as a smooth prominence of the promontory. The stapes is sometimes fixed. The pink swelling of the otosclerotic focus may sometimes even be detected clinically through a particularly transparent tympanic membrane. In microsliced temporal bones showing otosclerosis the focus appears well demarcated and pink. Blood vessels are prominent and evenly distributed. X-rays show the well-defined lesion as a patch of mottled translucency.

Microscopic appearances

The histological characteristic of otosclerosis is the presence of *trabeculae* of new bone, mostly of the woven type. This contrasts with the well-developed bone under the outer periosteum, the endochondral middle layer and the endosteal layer of the otic capsule, a sharply demarcated edge between normal and otosclerotic bone being a prominent feature. The pathological bony tissue has a variable appearance, with areas of differing cellularity. In most places osteoblasts are very abundant within the woven bone. Osteoclasts may be present and are accompanied by evidence of bone resorption. Marrow spaces

contain prominent blood vessels and connective tissue. In a few places the bone may be more mature and less cellular, and even lamellar bone may be found. In these areas marrow spaces are small. Thus active (more cellular and more vascular) and inactive (less cellular and less vascular) otosclerotic foci may be recognized⁷.

The commonest site for the formation of otosclerotic foci is the bone anterior to the oval window. The *fissula ante fenestram*, a normally appearing slit connecting middle ear with vestibule, is present in the same region (Figure 1) but this anatomical relationship does not necessarily denote any developmental connection. Cartilaginous rests are also normal in this area and may be seen nearby.

Otosclerotic foci may also be seen in the bone near the round window membrane, in the inferior part of the cochlear capsule or in the bone around the semicircular canals.

Otosclerotic involvement of the stapes footplate leading to functional fixation of the stapes may occur in two ways:

- a) There may be actual participation by the stapes footplate in the formation of otosclerotic bone so that the otic capsular focus of pathological bone is continuous with the former. This is often described as an "invasion" of otosclerotic bone from the otic capsule to the footplate, but it is unlikely that extension takes place in such a tumour-like manner. It is more likely that a field change of new bone formation affects the footplate as well as the otic

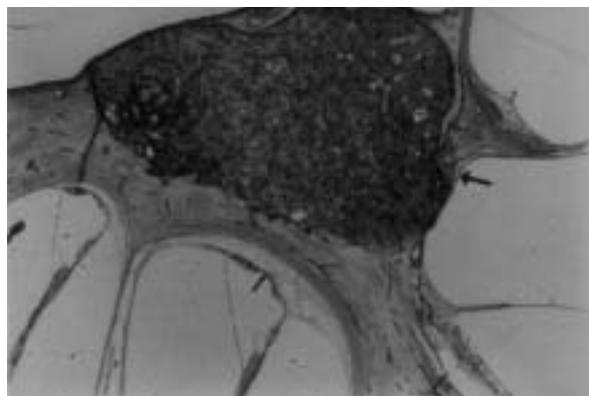


Figure 1. Oval window otosclerotic focus with ankylosis of the stapes (large arrow). Small arrow indicates spiral ligament of cochlea. $\times 35$

capsule, the fibrous annulus eventually undergoing obliteration. Involvement of the oval window takes place at any point of its circumference, or indeed around most of it. The process may also occasionally be associated with similar alterations in the lower parts of the stapedial crura.

- b) Frequently the footplate is not affected by the otosclerotic process, but the bone surrounding it proliferates to such an extent that the oval window is distorted and narrowed. Fibrous thickening of the fibrous annulus may be prominent. The otosclerotic focus may also encroach on the round window, narrowing it in the same fashion.

Otosclerotic bone frequently reaches the endosteum of the cochlear capsule. In some cases it may lead to a fibrous reaction deep to the spiral ligament. Overgrowth of otosclerotic bone may, rarely, cause distortion of the cochlear contours and even affect the modiolus and lead to spontaneous fractures of the *modiolar septa*⁸.

Much emphasis has been placed on the presence of lesions known as "blue mantles" in otosclerosis. At the centre of each such lesion is a small blood vessel and the mantle is represented by a deposit of new bone around the blood vessel which, since it is haematoxyphil, is described as being blue. Schuknecht⁹ warns against confusing blue mantles, which are actually formations of bone, with the thin blue-staining membranes normally found on the inner surfaces of *lacunae* in cartilage and *canaliculi* in bone and in the walls of bony vascular channels. He calls these membranes "Grenzschleiden" (boundary partitions). Lindsay¹⁰ observed blue mantles in almost all cases of diffusely distributed otosclerosis, but also in some cases of localized otosclerosis and in a few cases without the usual type of otosclerosis. They are present usually in the bony capsule around the semicircular canals¹⁰. The pathological process represented by the designation blue mantle would seem to be similar to the conventional form of otosclerosis and, indeed, areas of blue mantle formation may be present within the otosclerotic focus at the classical sites.

Hearing loss in relation to otosclerotic foci

By far the commonest form of hearing loss

in otosclerosis can be accounted for by oval window fixation and is conductive in type. Depression of sound waves derived from air conduction also occurs following obstruction of the round window by otosclerosis. The possibility of sensorineural hearing loss by otosclerotic involvement of the cochlea has been much discussed. It is feasible that a biochemical change may take place in the labyrinth from the proximity of the otosclerotic focus, which could lead to a disturbance of cochlear function.

In some cases the otosclerotic lesion may spread along the cochlea, surrounding it. This condition is clinically evident with a sensorineural hearing loss instead of a transmissive one and is confirmed by a HRCT scan. The oval window may be involved or not (Figures 2 and 3).

Discussion

The history is critical in the diagnosis of otosclerosis. The typical symptoms of otosclerotic deafness are a gradually increasing unilateral or bilateral transmissive hearing loss, most frequently occurring between the end of the second and fifth decades. In two thirds of the patients, there is a family history of deafness with autosomal dominant pattern of transmission with 40% of penetrance. It oc-

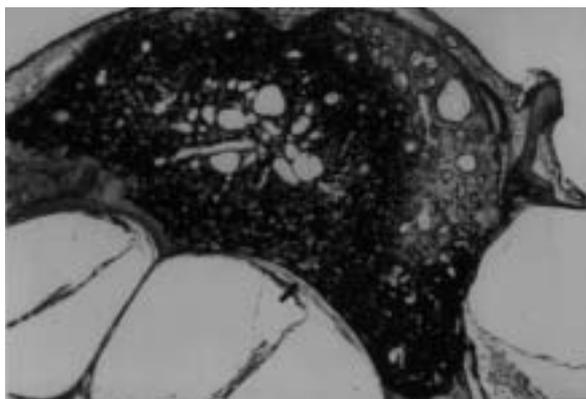


Figure 2. Cochlear otosclerosis. Note acellular hyalinization of spiral ligament adjacent to otospongiotic portion of the lesion (arrow). Also note the difference between the active otospongiotic lesion, which stains darkly, and the inactive otosclerotic lesion, which has stained lightly. Normal bone is visible on the left-hand side of the picture. $\times 35$

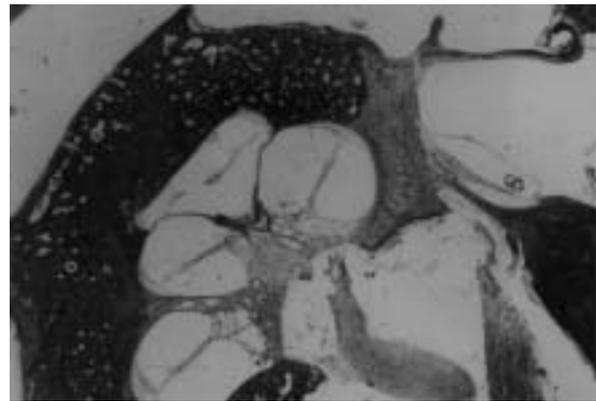


Figure 3. Coclear otosclerosis without stapes fixation. Otosclerosis surrounds the cochlea, but there is none in the oval window. $\times 15$

curs two times more frequently in women than in men and also is more common in whites than in other races. The progression of otosclerosis seems to be more rapid in younger patients. The hearing loss is also often rapidly progressive during pregnancy and in women on estrogen therapy.

Stapedioplasty is indicated when the stapes is fixed: absence of stapedial reflexes and air-bone gap > 20 dB for low frequencies.

It is also important to remember that in a certain subset of patients, profound bilateral sensorineural hearing loss may be caused by far advanced otosclerosis. It is important that these patients are identified, because often their hearing can be improved through surgical correction so that serviceable aided hearing is possible¹¹. The diagnosis of far-advanced otosclerosis is made from the history and can be confirmed by the presence of cochlear otosclerosis on the CT scan. A positive family history with gradual progressive hearing loss starting in early adult life, paracusia noted in an earlier stage of hearing loss, present or previous use of a hearing aid and measurable air-bone gap on previous audiogram are highly suggestive of far-advanced otosclerosis (Figure 2).

If a patient is believed to have far-advanced otosclerosis, and also the oval window is interested by the otosclerotic lesion, the stapedioplasty operation is useful to provide audible hearing aid¹²⁻¹⁴. If a patient has profound sensorineural loss for other reasons, cochlear implantation may be the procedure of choice^{15,16}.

Contraindications to stapes surgery in-

clude cochlear otosclerosis with no involvement of the oval window (Figure 3), poor speech discrimination or a history of vertigo in recent months, because either can indicate the probability of endolymphatic hydrops, with the risk of further cochlear loss should the labyrinth be opened. Otomicroscopic evidence of outer or middle ear infection requires postponement of surgery until the infection is successfully treated. Operation on a patient's only useful hearing ear should be avoided because of the small but definite and unpredictable risk of a permanent cochlear loss.

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