POSTSTAPEDECTOMY LONG-TERM HEARING RESULTS

A SYSTEMATIC REVIEW

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Contents

List of Tables

List of Figures

Introduction

Definition of otosclerosis:

Otosclerosis is a disorder of the bony labyrinth and middle ear ossicles **(Schuknecht, 1974).** A distinction should be made between histological and clinical otosclerosis. Patients may have histological otosclerosis without any clinical symptoms **(Guil, 1944).**

Schuknecht and Kirchner, (1974) defined clinical otosclerosis as stapes fixation due to an otosclerotic lesion. Cochlear otosclerosis can mean the disease has replaced part of the endosteal layer of cochlear bone **(Schuknecht and Kirchner, 1974)** or it can be used to describe SNHL assumed to be caused by otosclerosis **(Balle and Linthicum, 1984**).

Otosclerosis is a disease of the otic capsule and middle ear ossicles. It is more common in Caucasian populations. Histological incidence in Caucasians varies between studies from 3.4% to 13% **(Declau et al, 2001).**

Embryology:

The maturation of the bony labyrinth plays a role in the pathogenesis of otosclerosis. The otic capsule arises from mesenchyme surrounding the otic vesicle at 4 weeks of embryologic development. At 8 weeks, the cartilaginous framework is begun. At 16 weeks, endochondral bony replacement of this framework begins in 14 identifiable centers. In some people, complete bony replacement does not occur and leaves cartilage in certain locations one of these regions, the fissula ante fenestram, is anterior to the oval window and is usually the last area of endochondral bone formation in the labyrinth **(Roland and Samy, 2006).**

According to temporal bone studies, this region is affected in 80% to 90% of patients with otosclerosis. Areas of predilection for otosclerotic lesions, such as the border of the round window the apical medial wall of the cochlea, the area posterior to the cochlear aqueduct, the region adjacent to the semicircular canals, and the stapes footplate itself (which is derived from

otic capsule, as opposed to the superstructure, which is a branchial arch derivative) **(Roland and Samy, 2006).**

Histology of otosclerosis:

In 1912, **Siebenmann**, using a microscope, found that patients with otosclerosis presented with spongiform foci in temporal bone. In these spongiform lesions, endochondral bone is resorbed by osteoclasts and osteolytic osteocytes. Lesions extend into the surrounding bone as lacunae.

These lacunas contain a vascular space rich in fibroblasts and osteoblasts. multinuclear osteoclasts are also present in the centre and osteolytic osteocytes at the advancing edges. As a result, the bone is disorganized, containing an enlarged marrow, where new immature bone is produced and again resorbed. After many remodeling cycles, sclerotic, highly mineralized bone is created. It has a mosaic-like appearance caused by irregular patterns of resorption and subsequent deposition of fatty tissue in marrow spaces. An otosclerotic focus typically contains areas of differing activity **(Declau et al, 2001).**

An otosclerotic focus is found only in the temporal bone and middle ear ossicles **(Wang et al, 1999).** Although otosclerosis may involve any area of temporal bone, the place of predilection is in the oval window region. Another common site is the round window area, which is involved in 30-40% of patients **(Nylen, 1949).**

Otosclerosis may also involve the middle ear ossicles a solid otosclerotic focus of stapes without involvement of the stapediovestibular joint was found in 12% of patients in (**Guil's, 1944).**

The histological characteristic of otosclerosis is the presence of trabeculae of new bone, mostly of the woven type with marked vascularity. This contrasts with the well-developed lamellar 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. In most places osteocytes are very abundant within the woven bone **(Michaels, 2006).**

The footplate of the stapes is often invaded by otosclerotic bone, and the lower end of the anterior crus of the stapes is sometimes invaded (Fig. 1). Otosclerotic bone sometimes reaches the endosteum of the cochlear capsule. In some cases it may lead to a fibrous reaction deep to the spiral ligament. These changes are probably the basis of the sensorineural hearing loss that is also occasionally found in cases of otosclerosis **(Michaels, 2006).**

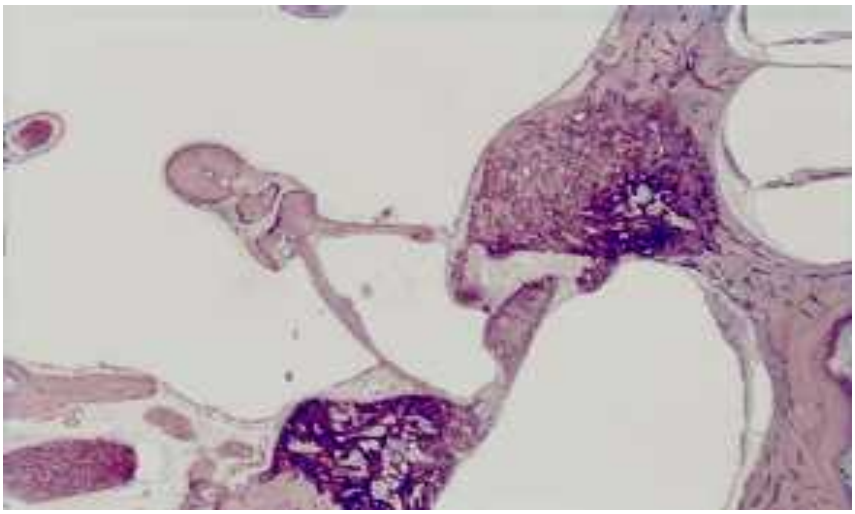

Figure1. Focus of otosclerosis involving both the anterior (*upper*) and posterior (*lower*) part of footplate of the stapes. The anterior focus has invaded onto the footplate and the anterior crus. This would have produced fixation of the stapes and its attendant

conductive hearing loss. Notice that the otosclerotic foci are more darkly staining and vascular than the adjacent normal bone. Quoted from **(Michaels and Hellquist, 2001).**

Epidemiology:

Otosclerosis is transmitted in an autosomal dominant fashion with incomplete penetrance (25% to 40%). The degree of penetrance is related to the distribution of lesions in the otic capsule. Some lesions are not located where they can cause clinical symptoms. About 10% of Caucasians have histological findings of otosclerosis. However, of those with histological changes, only 12% have clinical symptoms; thus, overall, this represents about 1% of the Caucasian population. In all races, when one ear is affected, the contra lateral ear shows histological involvement 80% of the time **(Roland Samy, 2006).**

In Norway, the overall female: male ratio was 3:2 with variation from 1:1 to 2:1 between different areas of norway (Hall 1974). In a recent study of 64112

patients in Germany between 1993 and 2004, the overall distribution was 1.6:1 (female: male) **(Arnold et al, 2007).**

The age at which symptoms become apparent is variable due to the insidious progression of hearing loss, but hearing loss often begins between the ages of 15 and 45 years. The average age at presentation is 33 years. About 60% of patients with clinical otosclerosis report a family history of this condition. Otosclerosis has been reported to advance more rapidly in females than males **(Roland and Samy, 2006).**

Aetiology of otosclerosis

Epidemiological studies have shown a genetic pattern of autosomal dominant inheritance with reduced penetrance. There are also environmental factors affecting the manifestation of otosclerosis. The interaction between these factors is very poorly understood. Recent gene expression analysis of human otosclerotic stapedial footplates showed 110 genes that

were expressed differently compared with controls. These genes showed multiple pathways that could lead to bone remodeling, for example interleukin signalling, inflammation, p53 signalling, apoptosis, epidermal growth factor receptor signalling and an oxidative stress response **(Ealy et al, 2008).** Thus, otosclerosis appears to have multiple aetiologies.

Genetic inheritance:

In epidemiological studies, a familial component has been well established. The majority of epidemiological studies on families with otosclerosis suggest autosomal dominant inheritance with approximately 40% reduced penetrance but the causative genes associated with these loci have not been identified, and the molecular process is unknow **(Markou and Goudakos, 2009).**

Therefore, it is not known whether genetic factors influence the histological manifestation of the disease, the progression to clinical disease, or both. One gene associated with otosclerosis is COLA1, the error of which leads to osteogenesis imperfecta (type I).

Histological similarities exist between these two entities, and some authors have suggested that otosclerosis is a local manifestation of osteogenesis imperfecta (type I) **(McKenna et al, 2002).** Although a familial link is well established, 40-50% of clinical cases are sporadic **(Markou and Goudakos, 2008).**

Viral aetiology:

The best established environmental factor connected to the otosclerosis is measles virus **(Arnold et al, 2007).** A specific virus has not been isolated, but measles virus ribonucleic acid (RNA) has been detected within the otosclerotic focus of the stapes footplate. In a study of 116 patients with stapedectomy, **Karosi et al, (2007)** observed measles virus RNA in all footplates with a histological focus. Similarly, the expression of a virus binding receptor (CD46) was increased substantially in footplates with a histological focus. Measles virus RNA has been thought to be evidence of the presence of a vital virus because without active viral replication, RNA rapidly disintegrates. Epidemiological reports of a decreased incidence of otosclerosis after a

measles vaccination programme also support the presence of a viral aetiology **(Arnold et al, 2007).**

Endocrinological factors:

Pregnancy has been suggested to cause onset or progression of otosclerosis. Abnormal parathyroid hormone function is another factor postulated to be involved in the aetiopathogenesis of otosclerosis. **Grayelin et al, (1999)** showed that expression and function of parathyroid hormone receptors in otosclerotic stapes footplates were lower than in bones collected from the external ear canal. However, in the same article, he concluded that this is probably not the triggering event in pathogenesis, but a consequence of abnormal regulation of bone matrix protein metabolism caused by another aetiological factor like measles infection. Similarly, glucocorticoids and the renin-angiotensin-aldosterone system have altering effects on bone remodeling of otosclerotic bone in vitro studies, but these are more likely to be epiphenomena of the disease **(Imauchi et al, 2008).**