Otosclerosis and Disorders of the Otic Capsule

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Introduction

Otosclerosis is a disease of bone confined to the otic capsule. Other systemic diseases however can mimic and even coexist with this condition. These conditions can be divided into:

- Systemic Bone Diseases:
  - Osteogenesis Imperfecta
  - Paget's disease
  - Osteopetrosis
  - Fibrous dysplasia
  - Craniotabular hyperostoses & dysplasias

- Metabolic Disorders;
  - Osteomalacias
  - Hypophosphataemic rickets
  - Osteoporosis
  - Hyperparathyroidism
  - Acromegaly

- Infective and Granulomatous Diseases;
  - Syphilis
  - Tuberculosis
  - Sarcoidosis
  - Histocytosis X

- Autoimmune Diseases;
  - Systemic lupus erythematosus
  - Cogan's syndrome
  - Relapsing polychondritis
  - Wegener's granulomatosis
  - Polyarteritis nodosa
  - Temporal arteritis
  - Behcet's disease

Normal Histology

The otic capsule is unique as it retains its fetal structure unlike other skeletal tissue. Histologically the otic capsule consists of three layers of bone: an outer periosteal, inner endosteal and middle endochondral layer. The endochondral layer consists of bone mixed with calcified cartilaginous cells which are otherwise resorbed by phagocytosis in other parts of the skeleton.

Calcification is usually complete by the age of one and there after there is very little remodeling. The adult otic capsule shows almost no osteoblastic or osteoclastic activity. Many different factors are needed to regulate remodeling which include growth factors,

cytokines, enzymes and free radicals. Failure to regulate these factors will lead to abnormal bone remodeling.

Otosclerosis

Otosclerosis is an autosomal dominant condition with variable penetrance. The incidence of the otosclerosis however seems to be in decline over the past thirty years. Some races including Asians, blacks and Native Americans have a very low incidence. A few authors have implicated measles virus in the etiology of otosclerosis. In one study Karosit and colleagues found measles RNA in 20 out of 34 footplates[4].

Histopathology
Bone remodeling in otosclerosis is confined to the otic capsule. There is disordered bone resorption and deposition with predilection for fissula ante fenestram. Fixation of the footplate therefore starts anteriorly and eventually involves the entire stapedial annular ligament. If it spreads in the opposite direction it will involve the cochlea and results in a sensorineural hearing loss.

Clinical presentation
The usual presentation is that of a progressive conductive hearing loss. A mixed or even a pure sensorineural loss (cochlear otosclerosis) might also occur.

Clinical Examination
This should include careful otoscopy which might reveal a red blush over promontory called Schwartze' sign. Pneumatic otoscopy and tuning fork tests (Weber and Rinne) are important to exclude other causes of hearing loss and confirm the audiometric results.

Investigations
Audiometry is the most important investigation. This usually reveals a conductive hearing loss. Tympanic membrane mobility is usually normal and acoustic reflexes are absent (although in the early stages the reflex might be diphasic).
With advanced cochlear otosclerosis there might be a severe sensorineural deficit with a conductive element which might not be obvious on the audiogram. Patients with an apparent sensorineural loss therefore might benefit from surgery.
High resolution CT scan might be able to detect areas of hypodensity in the otic capsule.

Medical Management
Fluorides in high doses (40mg/day) have been shown in few studies to stabilize hearing in a treated group (Bretlau et al).
Bisphosphonates have shown a promising result in osteoporosis and in one study Brookler and Tanyeri found Etidronate was able to diminish progression of otosclerotic foci as observed on CT scanning.
Hearing aids should be discussed with all patients as an alternative to surgery.

Surgical Management
Stapes surgery has evolved greatly over the years. Most surgeons now perform small fenestra stapedotomy. The senior author starts the fenestration with an Argon laser which is then enlarged with progressive dilators to 0.6mm allowing a 0.4 mm diameter prosthesis to be used.
In stapedectomy where the footplate is removed the oval window is often covered with perichondrium or a vein graft.
Other procedures performed in the past included fenestration of the lateral canal.
If the long process of incus is eroded or incus has to be removed to mobilize the malleus then a malleus to footplate prosthesis (malleolar vestibulopexy) may be used.

Variations Encountered at Surgery
- Persistent stapedial artery
- Round window obliteration
- Prominent promontory
- Facial nerve dehiscence with overhang
- Narrow oval window
- Gusher
- Floating footplate
- Depressed fragments
- Obliterative otosclerosis

Complications
- Altered taste occurs in about 20% of cases but almost all resolve with time.
- Infection, meticulous cleaning of the canal should reduce this risk.
- Vertigo, minor degrees are common and settle within few hours. If severe and persistent this might then suggest a perilymph fistula.
- Perilymph fistula; this will need re-exploration.
- Tinnitus
- Sensorineural hearing loss. In less than 1% of cases there is a severe or total sensorineural loss. The patient should be started on tapering high dose steroids.
- Facial palsy. A rare complication that may also be delayed in which case steroids should be used.
- Granuloma. This leads to gradual post operative hearing loss after an initial good result.
- Perforation of tympanic membrane which can be repaired easily at the same sitting.

Systemic Bone Diseases

Osteogenesis Imperfecta
This is a hereditary disorder of collagen synthesis. The defect is in the cross linkage of collagen fibers. Four types have been identified with type 2 being the most severe form and fatal in utero. Other types lead to multiple fractures in childhood and are often

mistaken for child abuse. The frequency of these fractures are reduced as affected individuals reach puberty.
Typical features include spontaneous fractures, blue sclerae, hypermobile joints, yellow stained teeth and hearing loss. X-rays usually show a mottled appearance with areas of demineralization and sclerosis.

Otological manifestation
Osteogenesis imperfecta can mimic otosclerosis with a progressive conductive or mixed hearing loss. Pre-operatively there might be footplate fixation, a missing long process of the incus or a hypermobile tympanic membrane.
If the preoperative diagnosis is made then a hearing aid should be the treatment of choice. In a few patients with a large air-bone gap stapedotomy might be considered keeping in mind that there is a higher rate of long process fracture and floating footplate.

Paget's disease

This condition is characterized by spreading lytic and sclerotic bony changes most often affecting skull, pelvis, lumbar spine, femur and tibia. The etiology of this disease is unknown. Recent studies however have suggested a paramyxovirus infection as a possible etiologic factor.
Patients typically suffer from bone pains, neurological deficits due to narrowing of skull foramina, hearing loss and vertigo.

Otological manifestation
Pagetoid involvement of the otic capsule can result in a progressive sensorineural or mixed hearing loss. Other otological features include occasional vertigo and pulsatile tinnitus.
The mainstay of treatment is a hearing aid. Other beneficial medical treatments for the general condition include Calcitonin injections and Bisphosphonates which seem to inhibit both osteoblastic and osteoclastic activity.

Fibrous Dysplasia

This is a disorder involving fibro-osseous tissue of unknown etiology. Three different types have been identified. Type 1 only involves a single bone, type 2 more than one bone and type 3 demonstrates systemic involvement of extra skeletal tissue (McCune-Albright Syndrome).
In fibrous dysplasia normal bone is replaced with a mixture of fibrous tissue and abnormal immature woven bone. In the skull the sphenoid and the frontal bones are most frequently affected.

Otological manifestation
Involvement of the temporal bone can lead to progressive hearing loss, bony occlusion of the external auditory canal and localized swelling.
Treatment is conservative unless there is a need to open an obstructed external canal.

Osteopetrosis
Also known as marble bone disease this is an inherited autosomal dominant condition with failure to reabsorb cartilage with immature bone formation. The result is bone with a thick cortex and narrow medullary cavity.

Otological manifestation
Otological features include conductive hearing loss and occasionally recurrent facial nerve palsy which might require surgical decompression.

Genetic Craniotabular Hyperostoses
These hereditary conditions with bony overgrowth include;
- Van Buchem's (Autosomal Recessive)
- Sclerosteosis (Autosomal Recessive)
- Congenital hyperphosphatasia (Autosomal Recessive)
- Progressive diaphyseal dysplasia (Autosomal Dominant)

Otological manifestation
Overgrowth of the temporal bone will lead to progressive conductive deafness and facial palsy. Surgical decompression of the nerve will be required in some cases.

Dietary & Metabolic Disorders

Osteomalacias (Vitamin D Deficiency)
Due to low levels of vitamin D there is usually reduced calcium and phosphate levels in the blood.

Otological manifestation
Progressive sensorineural deafness is seen in these patients. Ikeda et al (1987) showed that a normal serum calcium level is important for hair cell function.
Treatment therefore consists of vitamin D and calcium supplements. Despite treatment there is usually no significant improvement in hearing. Of note vitamin D toxicity can also lead to hearing loss from excessive tympanic membrane calcification.

Osteoporosis
In this condition there is a reduction in total bone mass due to increase loss of bone matrix. There might be an association with sensorineural hearing loss but this has not been established in the literature. Further studies are required.

Hyperparathyroidism
High levels of parathyroid hormone can be primary due to parathyroid gland adenomas or secondary due to chronic renal failure. High level of parathyroid hormone will in turn result in high serum calcium and low serum phosphate levels.

Otological manifestation
A mixed hearing loss is possible but rare.

Acromegaly
Excessive growth hormone levels will lead to overgrowth of bone. Unusual enlargement of hands, feet, skull and mandibles are seen.

Otological manifestation
Although this condition often involves the temporal bone, involvement of the otic capsule is rare.

Infective and Granulomatous Diseases

Syphilis
Treponema Pallidum infection can be congenital or acquired. Both congenital and acquired syphilis have been known to involve the inner ear.
In suspected cases where there is sudden bilateral hearing loss or fluctuating loss with vertigo laboratory tests for syphilis should be performed. Of these tests the fluorescent traponemal antibody absorption test (FTA-ABS) is the most sensitive. The venereal disease research laboratory test (VDRL) is however still widely used. In suspected cases of neurosyphilis cerebrospinal fluid should be obtained for testing.
A positive test does not necessarily mean that the patient has the disease as these tests remain positive even after effective treatment. False positive tests are not infrequently identified in connective tissue disorders from time to time.

Otological manifestation
In congenital syphilis treponemal labyrinthitis is sometimes seen while in secondary and tertiary syphilis a meningo-labyrinthitis usually occurs. In the late stages gummatous osteitis might involve the otic capsule. This will lead to secondary endolymphatic hydrops. There is often progressive sensorineural hearing loss associated with vertigo. Other sequele include facial and ocular palsies.
Treatment is with high dose penicillin. In some patients steroid therapy might improve hearing although there is no definite scientific evidence for this.

Tuberculosis
This condition usually affects the middle ear but can involve the otic capsule.

Otological manifestation
In chronic suppurative otitis media one should think of tuberculosis if there are atypical features such as excessive pale granulation tissue, sensorineural hearing loss or multiple perforations of the tympanic membrane.

Treatment should include surgery for chronic ear disease with antituberculous medication.

Sarcoidosis

This condition most often involves the lungs, liver, spleen, skin, eyes and lymph nodes. It can however rarely involve the temporal bone. The etiology of this non-caseous granulomatous disease is still unknown.

Otological manifestation

Hearing loss can be severe or even total. Vestibular symptoms are not infrequent. The facial nerve can also be involved.
Treatment is with steroids and immunosuppressive agents.

Histocytosis X

Pathologically histiocytosis X is an inflammatory reticuloendotheliosis involving the organs of the reticuloendothelial system i.e. skin, lungs, bone marrow, lymph nodes, liver, spleen and the central nervous system. There is an accumulation of histocytes in these organs.

Otological manifestation

The otic capsule can be involved leading to sensorineural deafness. Treatment is with systemic steroids.

Autoimmune inner ear diseases

Systemic Lupus Erythematosus

This condition is known to be an autoimmune mediated multisystem disorder with immune complex deposition. General features include a high erythrocyte sedimentation rate, low hemoglobin and positive anti-nuclear antibody factor.

Otological manifestation

This condition has been associated with sudden bilateral sensorineural hearing loss. Hamblin et al (1982) described immune complex deposition in the stria vascularis.

Cogan’s Syndrome

This condition causes fluctuant aggressive damage to cochlea and vestibule. It is characterized by a non syphilitic interstitial keratitis of the eyes.

Otological manifestation

This includes secondary hydrops with fluctuant vestibular function and sensorineural hearing loss.
Treatment is with steroids and immunosuppressive agents. Plasma exchange might be indicated in some cases.

Relapsing Polychondritis

As its name suggest there is an inflammatory reaction involving cartilage.

Otological manifestation

Auricles are affected most commonly. Sensorineural hearing loss can occur perhaps from autoimmune complexes. A conductive hearing loss can be due to the eustachian tube involvement.

Wegener's Granulomatosis

Pathologically Wegener's granulomatosis is an idiopathic inflammatory disorder associated with necrotizing granulomas and vasculitis most often involving the kidneys and lungs. The diagnostic test is c-ANCA which is highly specific for this condition.

Otological manifestation

There might be granulomatous tissue in the middle ear cavity. Treatment is with steroids and immunosuppressive agents.

Polyarteritis Nodosa

This condition another type of vasculitis usually involving visceral circulation.

Otological manifestation

Sudden bilateral sensorineural hearing loss can develop. Treatment is again with steroids and immunosuppressive drugs.

Temporal Arteritis

This is due to vasculitis involving the extra cranial arteries and if untreated can result in blindness due to involvement of the ophthalmic artery.

Otological manifestation

Otological complications are probably due to involvement of internal auditory artery leading to progressive hearing loss and vertigo. Treatment is with systemic steroids.

Behcet's Disease

This is a systemic vasculitic disorder often presents with uveitis and orogenital ulceration.

Otological manifestation

Most commonly cause a bilateral sensorineural hearing loss. Treatment is with immunosuppressive agents.

References

1. Brookes GB and Booth JB (1997) Diseases of the temporal bone. In: *Scott-Brown's Otolaryngology*, sixth edn, edited by J B Booth, A G Kerr, Vol. 3. London: Butterworth's. pp. 15/1-15/52.

2. House JW (1999) Otosclerosis. In: *Otolaryngology—Head & Neck Surgery*, Third edn, edited by, C W Cummings and others: Mosby. Chapter160.

3. Vrabec JT and Coker NJ: Stapes surgery in the United States. *Otology & Neurotology* 2004; 25:465–469.

4. Karosit et al. Measles virus prevalence in otosclerotic stapes footplate samples. *Otology & Neurotology* 2004; 25:451–456.

5. House HP et al: Stapedectomy versus Stapedotomy, comparison of results with long term follow up. *The Laryngoscope* 2002; 112:2046-2050.

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