Otosclerosis

- Otosclerosis is a primary disease of the otic capsule and the ossicles. It is characterized by a progressive hearing loss that may be conductive, sensorineural, or mixed. Histologically, it is seen as abnormal resorption and then deposition of bone in the labyrinthine capsule and middle ear.

History

- Valsalva was the first to describe stapes fixation as a cause of hearing loss in 1704 when he performed an autopsy of a known deaf patient.

Histopathology

- Otosclerosis is a pleomorphic bone dyscrasia. It may fix the stapes by a tiny bridge of abnormal bone or may totally obliterate it. It may spread to the cochlea to produce either a sensorineural loss of varying degree or deafness. Lesions may be sclerotic, spongiotic, or fibrous but usually compose all three histologic types. This diversity of size and anatomic distribution coincides with the variable types of hearing loss, from a mild conductive deficit to a total loss.
- The smallest otosclerotic lesions usually appear adjacent to the fissula ante fenestram, just anterior to the oval window in 80% of cases Schwartze’s sign that is occasionally seen on otoscopic exam. Osteocytes begin resorbing bone in perivascular spaces, which is replaced by cellular, fibrous connective tissue. Within this connective tissue, reticular cells and fibroblasts become osteoblasts, which lay down immature bone with a woven, disordered pattern of collagen fibers. It stains blue with H & E, thus forming “blue mantles of Manasse.”
- During the late, or sclerotic phase, the new bone is resorbed and replaced with osseous tissue containing many collagen fibers and little ground substance. The osseous tissue stains red with H & E. The large areas of vascular, cellular connective tissue are replaced with new bone, and the lesion becomes quiescent.
- Within each temporal bone containing otosclerosis, lesions can be found in each of these phases although the overall histologic status seems to be fairly uniform. The lesions begin in endochondral bone, but the endosteal and periosteal layers eventually become involved as well.

Etiology

- The etiology of otosclerosis remains unknown as of yet. Recent work has postulated infection by the measles virus as the initial insult and eventual cause of otosclerosis. There are other unproven but logical supports for this etiology as the incidence of otosclerosis has markedly decreased as immunization practices have improved. Also women seem to be susceptible to more severe and even fatal bouts with the measles virus and they have an increased incidence of otosclerosis as well.

History & Physical Examination

- The patient with otosclerosis typically presents with a history of slowly progressive hearing loss that is often bilateral and asymmetric. The hearing loss usually presents between the ages of 15 and 45. It occurs approximately twice as often in women than in men. In 70% of patients there is a family history of deafness. Younger patients often have a more rapid progression of the hearing loss. It is often rapidly progressive during pregnancy and in women on estrogen therapy.
- These patients often speak with a low-volume because they hear their own voices by bone conduction. Tinnitus is a common presenting complaint in patients with otosclerosis and occurs in approximately 75% of patients. Many patients (approximately 25-30%) have some type of associated vestibular symptoms. These are attributed to lesions of the horizontal canal and are usually not severe. These symptoms must be carefully differentiated from those of Meniere’s disease as hydrops is a contraindication to stapes surgery.
• Physical examination of the ear is usually normal in patients with otosclerosis but it is important in ruling out other diagnoses that may present with similar symptoms. Occasionally, a reddish blush may be seen over the promontory and in the oval window niche area. This is due to the rich vascular supply of an immature otospongiotic focus of bone and is termed Schwartz’s sign. It is only identified in approximately 10% of patients with otosclerosis.

• Tuning forks with the Rinne and Weber tests are an important part of the physical exam and should utilize the 256, 512, and 1024 Hz frequencies. The Weber test can help identify patients with a more advanced conductive hearing loss and can determine which ear has suffered the greatest insult.

• Your differential diagnosis of the conductive hearing loss with a relatively normal tympanic membrane should include tympanosclerosis, middle ear effusion, incus or malleus fixation, ossicular discontinuity, congenital footplate fixation, and cholesteatoma. Systemic diseases that can masquerade as otosclerosis include Paget’s disease and osteogenesis imperfecta. Paget’s disease may fix the stapes but more often, it crowds the ossicles in the epitympanum, partially fixing the ossicular chain. Cochlear involvement is a late finding. Elevation of the serum alkaline phosphatase level and involvement of other skeletal bones are often seen. Osteogenesis imperfecta (van der Hoeve syndrome) should be suspected in the presence of blue sclera, progressive conductive hearing loss, and multiple fractures. Their hearing loss is secondary to stapes fixation and these patients can have stapes surgery with results similar to those with otosclerosis.

Audiology

• The key test in the diagnosis of otosclerosis remains the audiogram. The audiological manifestations of otosclerosis are as distinctive as they are varied. Characteristic abnormalities appear on tympanometry, acoustic reflexes, and the pure-tone audiogram. The most typical case is the presence of a normal, type A tympanogram. As the disease progresses the height of the peak decreases and you can eventually obtain a type As tympanogram.

• Acoustic reflex morphology is a very sensitive indicator of otosclerosis. Advancing stapedial fixation affects both the ipsilateral and contralateral acoustic reflexes.

• Pure tone audiometry is always a part of the assessment and a gradually progressive low-frequency conductive hearing loss is first seen. As the sclerosis worsens and the footplate becomes fixed, a mass effect is added to the audiogram. This results in a stabilization of low-frequency thresholds, with worsening in the high frequencies and a gradual widening of the air-bone gap. The configuration changes from upward sloping to flat. Complete stapes fixation will give a maximum conductive hearing loss of 60 to 65 dB HL as long as there is not progression of the disease to involve the cochlea. Usually though, there is development of cochlear otosclerosis, and the loss becomes mixed or sensorineural. High frequencies are more severely affected at this point.

• The audiometric hallmark of stapes fixation is the Carhart notch, characterized by elevation of bone conduction thresholds of approximately 5 dB at 500 Hz, 10 dB at 1,000 Hz, 15 dB at 2,000 Hz, and 5 dB at 4,000 Hz. It is important to realize that the Carhart notch is a mechanical artifact, not a true representation of cochlear reserve. Commonly, one can obtain “overclosure” of the air-bone gap after stapedectomy with postoperative air-conduction thresholds lower than preoperative bone-conduction thresholds.

Management

A) Hearing aids

• All patients with otosclerosis should be offered the option of amplification during preoperative counseling. Patients refusing surgery or who cannot tolerate surgery should be considered for amplification. Patients with stapedial otosclerosis usually have excellent discrimination and hearing aids may provide effective treatment. Patients with advanced otosclerosis and mixed hearing losses in the severe range can obtain serviceable hearing with hearing aids after stapedectomy.
B) Medical treatment

- Medical treatment is controversial and consists of fluoride therapy, vitamin D, and calcium supplements. This should be considered for any poor surgical candidates, patients who opt against surgery, those who are suspected of having cochlear otosclerosis, or patients with vestibular symptoms due to otosclerosis. Several well conducted studies have been performed which verify the efficacy of fluoride therapy. It is believed that the fluoride ion replaces the hydroxyl ion forming a more stable fluorapatite complex that resists breakdown by osteoclasts. Shambaugh recommends 60 mg of sodium fluoride per day to obtain the maximum bone calcifying effect. The drug is well tolerated with minimal side effects. Evaluation of efficacy can be based on the disappearance of Schwartze’s sign, stabilization or improvement in hearing, and improvement in the CT appearance of the otic capsule. In addition, sodium fluoride can be used after successful stapedectomy to prevent progression of sensorineural hearing loss.

C) Stapedectomy

- Stapedectomy is the mainstay of treatment for otosclerosis. It was the first successful microsurgical operation and played an important part in the development and advancement of otology. ”
- The objectives of stapedectomy are: (1) to open the oval window for sound entry into the labyrinth; (2) to reconstruct a conductive bridge between the incus and the labyrinth; and (3) to accomplish these goals as efficiently and physiologically as possible for long-term hearing without complications.
- When performing stapes surgery there is always a fenestration made into the vestibule, the variable is the size. The techniques vary from total stapedectomy to partial stapedectomy to stapedotomy. Recently there has been a shift to stapedotomy, as one can gain reportedly equal or improved hearing with fewer complications.

Meniere’s disease

Introduction

- Meniere’s disease --also known as idiopathic hydrops-- is a disease process characterized by vertigo, fluctuant sensorineural hearing loss, tinnitus, and aural fullness.

History

- Meniere’s disease derives its name from Prosper Meniere, a French physician from the 19th Century. At the time, vertigo and several other neurological symptoms were believed to occur secondary to overfilling of blood vessels in the head. The role of the inner ear in balance was unknown. Seizures, headaches, and vertigo were considered part of “apoplectiform cerebral congestion.

Presentation

- Vertigo classically occurs in discrete attacks that last three hours, but may vary in duration from twenty minutes to twenty-four hours. Patients occasionally describe an aura--similar to migraine aura--which occurs before the onset of their vertigo. In contrast, other patients describe rapid and violent onset of their vertigo attacks which results in a fall. These “drop attacks” can cause traumatic injury. Hearing loss is sensorineural and usually unilateral. The side of the hearing loss is the same as the side as the vestibular weakness that causes the vertigo. Similar to the episodic nature of the vertigo attacks, patients often describe a fluctuating course of their hearing loss. The vertigo attacks and episodes of hearing loss often occur concurrently. Tinnitus is variable in pitch but is often described by patients as a buzzing sound. Patients also complain of a feeling of ear fullness which may feel like the ear is stopped up. Early in the disease process, patients do not complain of all symptoms simultaneously. Frequently, vertigo will occur first followed by hearing loss after several months. The course of the disease is highly variable. Patients may experience clusters of frequent vertigo attacks followed by long periods of remission.
Pathophysiology of Hydrops

- Endolymph is produced in the stria vascularis by dark cells of the vestibular labyrinth. In endolymphatic hydrops, an overaccumulation of endolymph results in encroachment of the perilymphatic space. The mechanism is officially unknown but remains controversial. Hydrops could occur from inadequate absorption in the endolymphatic sac or by constriction of the endolymphatic duct.

Possible Mechanisms for ELH

- Auto-immune disease via the production of antibodies is a possible by which endolymphatic hydrops occurs. Unlike other auto-immune diseases of the ear, Meniere’s disease patients show no white blood cell infiltration or evidence of cellular destruction. Viral causes are also possible. A sub-clinical viral infection could cause a delayed-onset hydrops. Finally, neuro-vascular mechanism similar to migraine could be responsible. Additionally, endolymphatic hydrops occurs from several known mechanisms such as trauma, acute otitis media, labyrinthitis, and congenital inner ear deformity. It is not clear why only a small subset of these patients develop Meniere’s disease.

Schuknecht Theory

- Rupture of the membranous labyrinth is thought to occur frequently in menieres because of the increased pressure within the scala media. The Schuknecht theory is a prominent theory that postulates that ruptures in the membranous labyrinth allow leakage of potassium-rich endolymph into the perilymph. The potassium is then exposed to CNVIII and the surrounding hair cells. Depolarization of the nerve cells occurs resulting in their inactivation. The final result is decreased hearing and vestibular function.

Making the Diagnosis

- History, physical exam, and audiogram are standard in evaluating complaints involving vertigo and hearing loss. In addition to these basic tools, there are several other diagnostic modalities that may be used to aid in the diagnosis of Meniere’s disease. ENG (electronystagmography) can localize the involved ear experiencing vestibular weakness.
- Significant reduction in caloric response is found in 48-73% of patients with Meniere’s patients. ECoG (electrocochleography) - measures evoked potentials that are created in the normal chain of events during hearing. Endolymphatic hydrops changes the ratio of these potentials in a characteristic way that can be measured to aid in the diagnosis.
- VEMPs (vestibular evoked myogenic potential) is a measurement of a type of neural impulse created when a person hears a sound. This impulse is altered in patients with ELH. MRI is frequently ordered to rule out an accoustic schwamoma since many of these patients will have unilateral SNHL. Blood tests may be used to rule out auto-immune inner ear disease.
- ECoG measures evoked potentials generated in the cochlea and auditory nerve as part of normal hearing physiology. SP (summative potential) occurs during depolarization of the hair cells. AP (action potential) is generated by the summed response of numerous auditory nerve fibers firing simultaneously. Both potentials are measured within 3-4 milliseconds after presentation of a stimulus to the ear. The SP/AP ratio in normal hearing patients has a characteristic ratio. The SP/AP ratio becomes elevated in hydrops (greater than 0.4). The mechanism for this increased ratio has been been fully explained but may occur from mechanical biasing of vibration of the organ of corti from the endolymphatic hydrops.
Treatment

- Initial medical management includes low salt diet, diuretics, avoidance of triggers (alcohol), and vasodilators. Symptomatic management control during acute attacks may be improved with antivertigenous, anti-emetics, sedatives, and anti-depressants. Despite widespread use of salt restriction and diuretics as the first-line treatment for Meniere’s, neither treatment modality has been evaluated in a double-blind placebo controlled study.

- Intra-tympanic steroid injections are a frequently used if conservative treatment fails. This is a reasonable option since steroids are unlikely to result in further hearing loss.

- Endolymphatic sac surgery (ESS) was described first in 1926 by Portmann, but its efficacy remains controversial today.

- Intra-tympanic gentimicin (aka chemical labyrinthectomy) was first tried in 1970s, but came into wide-use in the 1990’s. Gentimicin is a selectively vestibulotoxic aminoglycoside which induces apoptosis in vestibular dark cells. This reduces or eliminates peripheral vestibular function. The cochleotoxic effects are variable and hearing deterioration occurs in 13-35% of patients.

- Vestibular nerve section (VNS) via a retrosigmoid approach was first described by Dandy in 1930’s. Today multiple possible approaches include translabyrinthine, retrolabyrinthing, retrosigmoid, middle fossa, and combined.

- VNS is preferable when the patient has serviceable hearing. If hearing is poor, however, labyrinthectomy may be selected.