K. J. Lee: Essential Otolaryngology and Head and Neck Surgery (IIIrd Ed)

Chapter 8: Noninfections Diseases of the Ear

Histopathology of Otosclerosis

This bony disorder of the otic capsule assumes various histopathologic characteristics throughout its development. The early state is characterized by bone resorption and loose spongy bone widely interspersed with multiple vascular spaces. The more mature state, being denser, is characterized by fewer vascular spaces and by redeposition of bone. Otosclerotic bone can involve all three layers of the otic capsule (endosteum, endochondral, and periosteal) though it is said to begin at the endochondral layer. The involved endochondral layer lacks the typical "globuli ossei" (islands of ossified cartilage) of normal endochondral bone. Both osteoblastic and osteoclastic activities take place in otosclerosis. It is believed that the otosclerotic process starts with fingerlike invasion along blood vessels. On hematoxylin-eosin (HE) stain, this invasion appears bluish and is thus named the "blue mantle of Manasse". Histologic otosclerosis exists in about 8-10% of the white race; it is rarer among Orientals and blacks. The incidence among East Indians, however, is believed to be comparable to that of the white race. Clinical otosclerosis is estimated to account for 1% of conductive hearing loss in the white population.

The sites of predilection for otosclerotic involvement are:

1. Just anterior to the oval window (80-90% of the temporal bone with otosclerosis).

2. The border of the round window (30-50%).

The incidence of bilateral involvement is about 75-85% among temporal bones with otosclerosis. This disease shows a familial tendency; 60% of the patients with clinical otosclerosis have a positive family history. Some investigators believe it to be transmitted by a monohybrid autosomal-dominant gene with 25-40% penetrance; others believe that it may be transmitted by an autosomal-receissive gene. Among those with hearing loss, a positive family history can be obtained from 50-60%. The risk of an increasing hearing loss for any one pregnancy in a woman with stapedial otosclerosis is 1:4. If one parent has clinical otosclerosis, the children have a 20% chance of developing clinical otosclerosis.

Clinically, two eponyms are attached to otosclerosis: Schwartze's sign and Carhart's notch. *Schwartze's sign* is the reddening of the promontory seen through a translucent tympanic membrane. This supposedly represents the increase of vascular spaces in the periosteal layer of the promontory. Carhart's notch is a depression of the bone conduction threshold, greatest at 1000 or 2000 Hz. The Carhart's notch is eliminated after a successful stapedectomy, suggesting that it is not sensorineural, but rather is due to the impairment of bone conduction by a fixed footplate.

Malleus fixation caused by bone ankylosis between the head of the malleus and the tegment can give a conductive hearing loss similar to that of stapes fixation. The incidence of malleus ankylosis coupled with stapes fixation by otosclerosis has been estimated to be about 1.6% of the patients.

Paget's disease (osteitis deformans) is histopathologically very similar to otosclerosis. The main differences are:

1. Paget's disease has diffuse involvement whereas otosclerosis is limited to the temporal bone.

2. Otosclerosis involves all three layers of the otic capsule (endosteal, endochondral, and periosteal), whereas Paget's disease involves mainly the periosteal layer.

3. Osteitis deformans seldom involves the footplate or any part of the ossicles.

Osteogenesis imperfecta tarda is a systemic disease in which there is abnormal osteoblastic activity resulting in resorption of bone. This is an autosomal-dominant disease and is characterized by the patient with multiple fractures. Forty to sixty percent of these patients also have bluish-colored sclerae together with stapes fixation. This constellation of findings has been referred to as the van der Hoeve-de Kleyn syndrome.

Complications of Stapedectomy

Some of the complications of stapedectomy may indeed stem from inadequate preoperative evaluation and poor selection of the patients for surgery. To prevent unnecessary and avoidable complications, some of the suggestions by experienced otologists are as follows:

1. The minimum audiometric test requirements are pure tone bone- and air-conduction thresholds, speech reception levels, and speech-discrimination scores. The intelligent use of masking is highly important. The audiometric test results should always coincide with those of the tuning fork test. If they do not, do not proceed with surgery, but search for an explanation for the discrepancy. It may be due to a "shadow curve" from poor masking.

2. A sufficient conductive hearing loss should be confirmed by a negative Rinne test for at least two of the three speech frequencies.

3. A careful assessment of discrimination ability is essential. The ear that discriminates better should not be operated upon because of the risk of postoperative sensorineural deafness.

4. Stapedectomy when indicated should be performed in the worse hearing ear first. If a successful result is achieved, the operation on the second ear can be considered 12 months or more later.

5. This operation should never be done in an only-hearing ear.

6. The pros, and especially the cons of this operation should be carefully discussed with the patient before surgery. The patient should be advised of the chief risk of the operation, namely postoperative sensorineural deafness, and other possible complications.

7. Patients with otosclerosis rarely complain of vertigo. When there is vertigo and clinical evidence of coexisting labyrinthine hydrops (Ménière's disease), stapedectomy should

not be carried out for fear of a "dead ear". The distended saccule which may be in contact with the footplate, may de damaged during stapedectomy.

8. Stapedectomy should never be carried out in the presence of external otitis. The ear canal should be carefully examined the day before the operation and external otitis ruled out.

9. There is a higher incidence of failure in younger individuals because of the activity of the otosclerotic growth. This probably results from stimulation of the otosclerotic growth by stapedectomy.

10. Some otologists feel that stapedectomy should never be done in children because the risk of poststapedectomy sensorineural loss is high, especially if the preoperative bone conduction is poor. Bilateral infantile cases should be advised to wear a hearing aid until older.

11. The risk of prolonged postoperative vertigo is small but should be a consideration for some patients, i.e. professional athletes, high construction workers, roofers, and other individuals whose livelihood depends on special motor skills.

12. For patients engaged in frequent air travel, underwater sports, or other activities associated with unusual alterations in atmospheric pressure, Schuknecht suggests the use of a large fatty connective tissue graft and a shorter prosthesis.

13. Stapedectomy may result in worsening of speech discrimination. The elimination of stiffness by successful stapedectomy in a patient with a descending pattern of bone-conduction thresholds converts the pattern of air-conduction thresholds from flat to descending (paralleling the bone conduction) and results in a loss of speech discrimination. This discrimination is caused by the descending gradient of threshold sensitivity, not by morphologic changes in the inner ear. When this condition has been produced by the first stapedectomy, stapedectomy on the second ear should be discouraged.

14. Large exostoses of the external auditory canal may interfere with the surgical approach. In such cases, it is best to remove the exostoses first and to delay stapedectomy for several months.

Complications Encountered During Stapedectomy

Complications encountered during stapedectomy include tears of the tympanomeatal flap, dislocation of the incus, fracture of the long crus of the incus, cerebrospinal fluid leak, bleeding, vertigo, depressed footplate, and floating footplate. Exostoses, superiorly located jugular bulb, overhanging facial nerve, round window otosclerosis, persistent stapedial artery, malleus ankylosis, congenital anomalies of the stapes and incus, and obliterative otosclerosis also present problems during stapedectomy.

Tears of the Tympanomeatal Flap

A linear tear may require no repair. Most tears can be satisfactorily repaired with fat from the earlobe, temporalis fascia, or tragal perichondrium. A flat piece of tissue of suitable

size is introduced into the perforation with the graft on the medial surface. The margins of the perforation are approximated as closely as possible.

Dislocation of the Incus

1. Subluxation of the incus consists of a tear of the incudomaleal joint but with sufficient intact capsule to maintain the incus in its normal anatomic position. Although the long process of the incus will be excessively mobile, the operation may be completed and the functional result may be satisfactory.

2. Luxation of the incus is due to a complete disruption of the incudomalleal joint and demands that the incus be removed and a malleus-oval window prosthesis be utilized. Attempts to replace and maintain the incus in its original position usually are not successful. The incus may be accidentally dislocated during curetting of the bony annulus and during extraction of the stapes. The long process of the incus may be accidentally displaced when withdrawing hooked instruments from the oval window niche.

Fracture of the Long Process of the Incus

This complication is rare. If the fracture occurs near the tip of the long process, the wire prosthesis may be placed on the stump. If the stump is too short, a malleus oval window prosthesis should be used. When the lenticular process is particularly long, and close to the promontory, it is advisable to fracture and shorten it to prevent a fibrous adhesion to the promontory.

Cerebrospinal Fluid Leak

In about 1:300 cases, opening of an oval window reslts in a sudden profuse flor of clear fluid. Those ears apparently have large patent cochlear aqueducts through which cerebrospinal fluid enters the inner ear. The fluid traverses the scala tympani to the helicotrema and then passes through the scala vestibuli to the vestibule to reach the oval window. In these cases, the fat graft technique is preferred and the patient is maintained in a head-elevated position for several days post-operatively. Cerebrospinal fluid flow is more common in cases of congenitally fixed stapes.

Bleeding

All bleeding from the ear canal should be controlled before the middle ear is opened. Bleeding from the mucous membrane of the oval window usually subsides spontaneously, but if necessary, may be controlled by pressure applied with a cotton pledget or Gelfoam soaked with epinephrine. Occasionally there are large vascular channels in the otosclerotic bone which may bleed during footplate removal. Thus bleeding into the vestibule may be unavoidable. Most otologists prefer to leave the clot in the vestibule untouched to complete the stapedectomy. A blood clot in the vestibule does not compromise the end result.

Vertigo

Vertigo occurring during the stapedectomy operation is either due to mechanical stimulation of the vestibular sense organs or to cold caloric stimulation. One of the advantages of using local anesthesia is to permit this symptom to be monitored during surgery.

The aspiration of perilymph and its replacement with air results in displacement and collapse of the vestibular labyrinth and is associated with vertigo. Manipulations within the vestibule may cause vertigo. No instrumentation should be performed with the vestibule, and any manoeuver which creates vertigo should be avoided. A fragment of footplate or a "broken" instrument which has fallen into the vestibule should be left alone. Attempts to remove it are followed by a high incidence of sensorineural hearing loss.

The introduction of a prosthesis which is too long may cause vertigo. The proper length should be determined by measurement. Before a prosthesis is tightened to the incus it should be determined if vertigo is elicited by an inward movement of the prosthesis. This is particularly important when fitting a piston prosthesis.

When perilymph has been removed, the utriculus may assume an abnormally superior position in the vestibule. In this position it will be free from possible contact with a prosthesis during surgical procedure, but it will return to its normal position subsequently, as perilymph is reformed. This constitutes an important reason for avoiding loss of perilymph.

The pooling of an anesthetic agent in the round window niche may lead to its resorption into the inner ear and result in vertigo and nystagmus. During the injection procedure, care should be used to avoid allowing the anesthetic agent to enter the middle ear.

Depressed Footplate

Every effort should be made to avoid a depressed footplate. A severely depressed footplate or footplate fragment should be left in the vestibule and a Gelfoam wire or tissue wire prosthesis introduced in the usual manner. These patients usually experience postoperative vertigo for several weeks and unsteadiness for many months.

Floating Footplate

The two conditions which lead to a floating footplate are previous stapes mobilization and minimal stapes fixation. When these conditions exist, a small opening should be created in the footplate before removal of the superstructure. This opening should be large enough to admit a 0.3 mm hook. If in spite of adequate precautions the floating footplate occurs, its removal must be effected through an inferior margin burr hole. A 0.5 mm sharp cutting burr is used to make a notch in the inferior margin of the oval window.

Complications Following Stapedectomy

Postoperative complications following stapedectomy include acute otitis media, suppurative labyrinthitis and meningitis, vertigo, reparative granuloma, perilymph fistula, facial paralysis, fluctuating conductive hearing loss, persistent perforation of the tympanic

membrane, taste disturbance and dry mouth, postoperative fibrosis, incus necrosis, and delayed sudden deafness.

Acute Otitis Media

To prevent this complication, many otologists prescribe antibiotics. Acute infections should be treated intensively with the appropriate antibiotics as determined by culture and sensitivity studies.

Suppurative Labyrinthitis and Meningitis

The most serious complication of stapedectomy is suppurative labyrinthitis leading to meningitis. Although it is extremely rare, several deaths from this complication have occurred. Presumably the bacterial invasion occurs through a thin membrane or fistula of the oval window. There is a higher incidence of meningitis with a polyethylene strut than with other tissue-wire prosthesis. Patients having had stapedectomy should be counseled about the importance of prompt treatment of respiratory infections when they are associated with ear discomfort.

Postoperative Vertigo

Vertigo may occur immediately following stapedectomy, or its onset may be delayed. Immediate postoperative vertigo is a result of perilymph loss, direct surgical trauma, or postoperative serous labyrinthitis, and it usually subsides within a few days. When vertigo or a sensation of unsteadiness persists for longer than a few days, a search for the cause should be made. Possible causes are: depressed footplate, reparative granuloma, excessively long prosthesis, and an oval window fistula. The long prosthesis is readily detected by eliciting the fistula response with the pneumatic otoscope. The exact position of the metallic prosthesis may be demonstrated by polytomography. Surgical intervention may be required not only for the comfort of the patient, but also to preserve inner ear function. Positional vertigo of the benign paroxysmal type (cupulolithasis) may follow stapes surgery and probably is due to injury to the utricle with release of otoconia. This type of vertigo is usually self-limiting, but it may persist for several months or years.

Reparative Granuloma

Granuloma of the posterior mesotympanum occurs as a complication of stapedectomy in approximately one or two out of 100 cases. The condition usually becomes manifest between the seventh and fifteenth postoperative days. Progressive sensorineural hearing loss after an initial hearing gain in the earliest and most consistent symptom. Vertigo, tinnitus, and sensation of fullness in the ear may be present. Reparative granulomas are associated with serous labyrinthitis in the early stages and serofibrinous labyrinthitis in the later stages when the inner ear damage becomes permanent. (Kaufman, R. S. and Schuknecht).

Examination reveals an edematous thickened tympanic membrane with redness in its posterior half. Audiometric tests show a combined sensorineural and conductive hearing loss worse in the high frequencies and with marked decrease in speech discrimination. Emergency surgical intervention is imperative in these patients. Complete removal of granuloma within

the first two poststapedectomy weeks may prevent permanent sensorineural hearing loss.

Perilymph Fistula (Oval Window Fistula)

The incidence of oval window fistula following a stapedectomy varies from 0.3% in 1772 patients to 2.5% in 1784 patients. The symptoms of perilymph fistula are similar to those of endolymphatic hydrops. A sudden decrease in hearing often with vertigo and unsteadiness, a sense of fullness in the affected ear, and sometimes roaring tinnitus, are common symptoms. Gradual progressive deterioration, but with fluctuation, is the rule in perilymph fistula.

The symptoms of oval window fistula may be evident within a few days or weeks following stapedectomy, or may be delayed for weeks or months. The delayed type of fistula is especially common after insertion of a polyethylene tube or Teflon prosthesis. The incidence is higher with a wire-Gelfoam prosthesis than with a wire-tissue prosthesis.

The treatment of perilymph fistula is unsatisfactory in most cases as far as hearing is concerned, but satisfactory for elimination of vertigo and risk of meningitis. Removal of the entire membrane from the oval window and replacing it with a wire-tissue prosthesis usually will stop the leakage of perilymph and the fluctuating symptoms, including vertigo. Early recognition and treatment may enable recovery of hearing to the previous good level.

Facial Nerve Paralysis

Immediate facial paralysis usually results from the local anesthetic reaching the facial nerve. Recovery of facial function is complete in 2-4 hours. If immediate paralysis persists, surgical trauma is likely and the nerve should be explored and decompressed within 24 hours. Direct instrument injury to the facial nerve is rare, but extreme caution should be used to prevent this complication. Approximately 50-60% of patients have dehiscence of the inferior aspect of the facial canal. Facial paralysis that begins several days after operation may be assumed to be the result of edema of the facial nerve.

Fluctuating Conductive Hearing Loss

Fluctuating conductive hearing loss is the result of loose linkage of either end of the prosthesis. A faulty linkage characterized by intermittent contact with the oval window membrane causes large fluctuations in hearing and is corrected by introducing a longer prosthesis. Loose linkage on the incus is characterized by small fluctuations in hearing, and is corrected by either tightening the wire loop or by removing the prosthesis and replacing it with a properly tightened wire hook.

Although hearing impairment with perilymph fistula is predominantly of the sensorineural type, a fluctuating or persistent conductive loss may be caused by perilymph fistula. This is considered to be due to the "backsplash" of a leaking vestibular perilymph space.

Perforation of Tympanic Membrane

A small postoperative perforation will usually heal spontaneously. Large perforation due to surgical trauma or postoperative infection require later myringoplasty.

Taste Disturbance and Dry Mouth

Stretching, tearing, or cutting the chorda tympani nerve can cause loss of taste on that side of the tongue and dryness of the mouth. Dryness of the mouth may be a particularly disagreeable poststapedectomy complication if both nerves are damaged. Some otologists feel that the patient has fewer symptoms when the chorda tympani nerve is cut than when it is overstretched.

The disturbance of taste, dryness of the mouth, and numbness of the anterior part of the tongue after trauma or section of the chorda tympani nerve, begin to lessen in a month or two and usually disappear after 3 or 4 months; but they may persist longer or can be permanent.

Postoperative Fibrosis

A fibrous band from the incus to the promontory is a common cause for incomplete closure of the bone-air gap. The incidence of this complication can be minimized by avoiding surgical trauma to the mucosa of the promontory. Corrective surgery includes removal of the fibrous band and introduction of a Teflon or Silastic disc.

Incus Necrosis

Pressure necrosis of the tip of the long process of the incus appears to be caused by direct surgical trauma or by the irritation of the prosthesis. The incidence of this complication is highest with polyethylene tube prostheses in which case the resorption is due to osteitis initiated by foreign body reaction. A loosely crimped wire also can produce a low-grade reaction and local bone resorption. For this reason the loop of the prosthesis must be secured tightly to the incus.

Delayed Sudden Sensorineural Hearing Loss

This is a rare complication of stapedectomy and may be caused by alterations in atmospheric pressure such as are experienced in an airplane or elevator. It is most common in patients who have had a polyethylene tube prosthesis inserted and is usually accompanied by severe vertigo and profound hearing loss. Presumably in these cases the prosthesis has been displaced into the vestibule.

Sudden profound sensorineural hearing loss, without vertigo and without obvious provocation, may occur some months or years following stapedectomy, particularly after revision procedures.

Recent Studies on Stapedectomy

Small-Fenestra Stapedectomy

1. A small-fenestra stapedectomy is based on the rationale of creating a more effective acoustic mechanical transmission system, while reducing potential labyrinthine disturbance. The surgical technique for the small-fenestra stapedectomy described by Bailey at al, includes the creation of the fenestra and the use of a McGee stainless steel piston prosthesis with loose areolar tissue around the piston. Postoperative results are compared in a series of 100 cases, 50 having the small-fenestra technique SFT) and 50 having a partial or total footplate removal procedure. Vestibular results demonstrate a noticeable reduction in postoperative complaints of balance disorders in the SFT patients. Hearing results, when compared between the two groups, show a statistically significant advantage for the SFT patients in postoperative high-frequency threshold sensitivity and speech discrimination scores. The advantages of lowered risk, based on reduced trauma to, and contamination of, the labyrinth, as well as improved high-frequency hearing sensitivity and speech discrimination, support the procedure according to Bailey et al.

2. McGee analyzed the results of 280 consecutive stapedectomies. One hundred forty-one (141) operations were done using the "total stapedectomy" technique, and 139 were done using the "small-fenestra" technique. The auditory acuity of the two groups was compared with reference to conductivew and sensorineural performance. The data supported the author's original impression that the small-fenestra technique is the treatment of choice in the surgical management of clinical otosclerosis.

3. According to B. Farrior, the small-hole stapedectomy with a bare piston is contraindicated when there are excessive fluid pressure changes in the inner ear and when there are excessive air pressure changes in the middle ear. The small-hole stapedectomy is the operation of choice in thick fooplate or obliterative otosclerosis which requires the use of the hand or power drill to create an opening in the oval window. When a small-hole stapedectomy is attempted and there is a tilting of a portion of the footplate, the footplate should be removed and the window sealed with a graft.

Complications of a small-hole stapedectomy are rare but consist of fistulas, a tented piston, a dislocated piston, or a partial closure with a tilted footplate. Complications of a small-hole stapedectomy may be reduced by placing a thin tissue graft over the foot of the piston, a socked piston, or a capped piston.

Total Stapedectomy

The concept of total footplate removal in stapedectomy is recommended by Robinson as the preferred technique, providing that the surgery is atraumatic. Only that part of the footplate which can be removed easily should be removed; in 4671 consecutive stapedectomies a total footplate removal could be accomplished in 74%. Total and partial footplate removal necessitates a connective tissue graft to seal the oval window and must be coordinated with a suitable prosthesis whcih is efficient, functional, and self-centering. The successful results between total and partial footplate removal are not statistically significant when employing the criteria of air-bone gap closure to within 10 dB, but when comparing the

complete air-bone gap closure rate there is a statistically signigicant difference with total footplate removal providing a more favorable hearing result.

Revision Stapedectomy

1. Sheehy et al reviewed the records of 258 revision stapedectomy operations performed during an 8-year period.

Displacement of the prosthesis to the inferior edge of the window was the most common cause of failure (41%) and occurred predominantly in wire-Gelfoam pad cases. An oval window fistula, a short prosthesis, or bony closure of the window were causes of failure in 9%. Incus necrosis was the cause of failure in 5%.

Less than 50% of the operations resulted in postoperative conductive deficit of 10 dB or less. The results were better in incus bypass procedures in revisions of patients in which a tissue graft was used over the oval window, and in revisions of ears initially operated on elsewhere. Severe sensorineural hearing impairment was the result in 7% of the operations and half of these impairments resulted in dead ears. The majority of these adverse results followed a repeat drill out of obliterative otosclerosis, or followed reopening of the oval window in patients with a postoperative inner ear problem other than a fistula.

They concluded that (a) revision stapedectomy is a less satisfactory procedure than primary stapedectomy; (b) there is rarely an indication for a repeat drill out of obliterative otosclerosis; and (c) the oval window membrane usually should not be disturbed in revision stapedectomy in a patient with inner ear symptoms unless there is a fistula.

2. In a study by Crabtree et al of 35 patients who underwent revision stapedectomy, revision stapes surgery improved hearing to a satisfactory level in fewer than half of the patients. The risk of sensorineural hearing loss is greater in revision surgery particularly if a sensorineural loss followed primary surgery.

Stapedectomy in Congenital Anomaly

Stapedectomy in a patient with congenital malformation of the middle and external ear should be done with caution. If there is a footplate, one should make the smallest possible diagnostic tap to rule out a profuse flow of cerebrospinal fluid. If none occurs, then one might consider stapedectomy and reconstruction.

Laser Stapedectomy

Argon laser stapedectomy was developed by Perkins. Fenestration of the stapes footplate with an argon laser microscope appears to be an advantageous method of restoring hearing in otosclerotic patients. The use of the laser, which gives increased precision and control to the surgeon, appears to be translate into a lower risk of negative results, less postoperative balance disturbance and better hearing when compared with current methods of management. This should mean shorter average hospitalization, less time away from work when analyzed from an economy viewpoint, and better hearing when viewed from the qulity-of-life standpoint.

Stapedectomy - Postmortem Findings

1. In reviewing histologic specimens of 16 temporal bones of 13 subjects who had stapedectomy operations, Schuknecht and Jones conclude:

Prostheses which overlap the margins of the oval windows result in an incomplete closure of the air-bone gaps. This is primarily a complication of methods which employ total removal of the footplate, and is caused by either improper placement or subsequent migration of the prosthesis. This problem can be minimized by careful centering of the prosthesis in the oval window or by the employment of partial stapedectomy in association with a piston prosthesis method in which the piston extends the vestibule.

b. Fibrous adhesions are a common consequence of trauma to the mucous membrane of the middle ear. Fibrous bands which extend between the lenticular process and promontory may cause incomplete closure of the air-bone gape. On the other hand, fibrous tissue which extends between the oval window and incus in association with the prosthesis appears to have little effect on sound transmission. The occurrence of fibrous adhesions can probably be minimized by avoiding trauma to the mucous membrane, particularly in the area of the promontory.

c. Gelfoam implantation following total stapedectomy promotes the formation of thin oval window membranes. However, even the thinnest membrane is at least as thick as the round window membrane. There is not evidence to suggest that the thin membrane is an inadequate seal for the oval window.

d. Thin membranes frequently show cupping (bulging out of the oval window), as a consequence of which the medial ends of the prostheses extend through the membranes. This condition appears to have no adverse effect on sound transmission.

e. Most inci show small areas of cortical bone resorption at the site of contact with the prostheses. However, the process appears to be self-limiting.

2. Two patients who developed sensorineural deafness following stapedectomy were studied by Schuknecht and Mendoza. A postmorted histologic study of one patient showed severe cochlear degeneration, presumably caused by necrosis of the fat graft. The study of the other patient showed that the prosthesis impaled the saccule, causing collapse of its wall with subsequent cochlear endolymphatic hydrops. An atraumatic surgical technique is strongly suggested.

3. Surgeons who perform stapedectomy for otosclerosis should be cognizant of the relevant surgical pathology to better assess their techniques and adopt methods which will optimize functional results and minimize complications.

Traumatic Perforation

Traumatic perforation can be caused by blast injury, welding injury, a force striking the auricle thereby occluding the external auditory canal (i.e. a slap with the open palm), and by a penetrating injury (i.e. Q-tip injury). There are many ways of treating these perforations

and the results are, to a large extent, comparable. Most otologists would agree that, without infection, 85-90% of all traumatic tympanic perforations heal spontaneously. However, whenever vertigo is associated with the injury, dislocation of the stapes is suggested, and hence surgical exploration should be performed as soon as possible. Among the above mentioned causes, a penetrating injury gives the highest incidence of stapes dislocation.

A small traumatic perforation without ossicular dislocation with no foreign body or squamous epithelium embedded in the mesotympanum can be treated expectantly. When no infection is evident antibiotic drops are not necessary. The use of 10% trichloroacetic acid and a paper patch (or Gelfoam) has been practiced by some otologists.

In the case of a large perforation without ossicular dislocation but with infolding of the edges of the perforation, the mesotympanum should be cleaned of foreign debris and the edges unfurled over a piece of adipose connective tissue or over pieces of Gelfoam. A tympanoplasty packing should be applied over this repair. In adults or in cooperative older children, this minor procedure can be performed under local anesthesia.

Whenever there is doubt as to the possibility of ossicular dislocation with a traumatic perforation, exploratory tympanotomy and tympanoplasty should be considered.

Sudden Deafness

Sudden deafness is defined as a sensorineural deafness that becomes instantly apparent, or one that rapidly develops over a period of hours or a few days. The hearing loss is often noticed on awakening in the morning, or during any of the days' activities such as those involving physical or emotional strain or even while at rest. Some patients do not complain about a hearing loss but may complain of stuffiness or blocked feeling in the ear or tinnitus. The hearing loss may range from mild to total and is typically unilateral although it may be bilateral. There may be an accompanying dizziness or vertigo (50%) but this is usually mild and typically improves over a period of a few days. Males and females appear to be equally affected.

Etiology

1. Frequently cited predisposing factors include changes in the physical environment such as altitude and other forms of change in atmospheric pressure, allergic manifestations, use of alcohol, emotional disturbances of the patient, physical exertion, diabetes, arteriosclerosis, pregnancy, use of a contraceptive drug, stress of surgery, and general anesthesia.

2. Specific etiologies that are well-documented are limited to viral agents. The sites of the viral-induced pathologic changes are the cochlea ("viral and endolymphatic labyrinthitis") and the components of the eight cranial nerve ("viral neuronitis and ganglionitis"). The viruses of mumps, measles, influenza, and adenoviruses may cause sudden deafness of the viral endolymphatic labyrinthitis type. The viruses of herpes zoster are the sole agents that have been shown to produce viral neuronitis and ganglionitis. The presence of active upper respiratory infection is noted in 25% of the patients at the onset of sudden deafness.

3. Vasospasm, thrombosis, embolism, hemorrhage into the inner ear, hypercoagulation, and sludging of blood frequently are considered as the most common causes of sudden deafness, but the evidence is lacking.

4. Cerebellopontine angle tumors (retrocochlear tumor) have been demonstrated to produce sudden deafness.

5. Simmons theorized a break in Reisner's membrane as a cause of sudden deafness resulting from sudden pressure changes (getting out of bed, sneezing, coughing, bending, performing a Valsalva maneuver, or scuba diving).

6. Goodhill recognized round window fistula as a cause of sudden deafness. Many reports of labyrinthine window ruptures followed. The anterior part of the oval window is the most likely area of rupture. The findings of positional nystagmus, a positive Romberg's or fistula sign with a sensorineural loss makes the diagnosis of an inner ear window rupture most probable. Tympanotomy is necessary to identify and close the fistula.

7. Morimitsu reported a new theory and therapy for sudden deafness. He treated 60 cases of sudden deafness with Urografin, injecting 1 mL intravenously the first day, then 2 mL injections daily until hearing reached the maximum recovering point.

Twenty-two of those cases have had a complete recovery. Eight have had a "remarkable recovery, meaning that there has been a hearing gain of more than 30 dB in the average threshold of 250, 500, 1000, 2000, and 4000 Hz". Three cases have shown slight improvement with average hearing gains between 10-30 dB. The remaining 27 cases have shown either no change or hearing threshold changes within 10 dB.

He noted that there was no improvement with Urografin in patients with vertigo. He also noted that recovery was almost complete in all test frequencies even in the patient treated 43 days after onset. Therefore, he believed that the lesion is probably not in the hair cells "which easily develop irreversible changes", but is a functional change in the cochlea. Based on the pharmacology of Urografin, Morimitsu has deduced that dysfunction occurs in the stria vascularis.

It is well established that both stria vascularis and the renal tubular epithelium are damaged equally by ototoxic antibiotics and the diuretics, furosemide and ethacrynic acid. Both diuretics deactivate the sodium-potassium ATPase in the stria vascularis, decreasing the endocochlear DC potential and cochlear microphonics, and the stria vascularis generates endocochlear DC potential.

If one assume that the hearing loss of sudden deafness is caused by a depression of endocochlear DC potential by whatever cause, then it is easy to understand why the hearing loss occurs suddenly, recovers quickly, and also very completely.

It is Morimitsu's theory that in sudden deafness of this type the blood-cochlear barrier is broken down at the area of the stria vascularis and that the molecular weight and character of Urografin is such that a little of it leaks into the circulation to fill the broken membrane pores and reactivate the sodium pump to produce again normal DC potentials and also normal

endolymph.

8. Wetmore and Abramson reported three cases of reversible sensorineural hearing loss associated with bullous myringitis.

9. Unilateral sudden sensorineural hearing loss after open heart surgery was reported by Plasses et al. All seven cases were male. The most likely cause is particulate microemboli generated by a cardiopulmonary bypass.

Classification

Sudden deafness may be classified from the etiological standpoint into two groups:

1. Localized lesions of the temporal bone:

a. Acoustic neuroma.

b. Cerebellopontine angle tumor.

c. Oval or round window fistula.

d. Aneurysm of anterior inferior cerebellar artery.

2. Systemic diseases involving the temporal bone:

a. Viral infections which are cochleapathic.

b. Accelerated coagulation.

c. Hyperviscosity.

1) Polycythemia vera.

2) Macroglobulinemia.

d. Arteriosclerosis secondary to:

1) Aging.

2) Hypertension.

3) Diabetes.

4) Hyperlipidemia.

e. Collagen diseases.

f. Multiple sclerosis, syphilis, and many others.

Systemic Evaluation and Management

The evaluation and management of these patients with sudden deafness must proceed in a systematic fashion.

1. Otologic Examination.

Sudden deafness may be of a conductive or sensorineural type. These can be differentiated by otoscopic examination, tuning fork tests, and audiometric tests. When the tympanic membrane is normal, sensorineural loss is most likely present. Wax in the external auditory canal and serous or acute otitis media may often present as sudden conductive deafness.

Vestibular function tests should include spontaneous nystagmus, positional test, Rhomberg's gain, caloric test, and electronystagmography when indicated.

2. Audiogram.

The pure tone audiogram with air and bone conduction will confirm the clinical diagnosis of sensorineural loss. Cochlear or retrocochlear (eight nerve and cochlear nuclei) losses can be differentiated by special audiometric tests which include alternate binaural loudness balance (ABLB), short increment sensitivity index (SISI), discrimination score, tone decay, and Bekesy audiometry.

3. Coagulation Studies.

The sudden nature of sudden deafness suggests a sudden vascular occlusion which could arise from thrombosis of the cochlear vessels. Yet, many of the patients are young, without any evidence of arteriosclerosis, diabetes mellitus, hypertension, hyperlipidaemia, and other vascular diseases. Therefore, the possibility of a thrombosis arising from a hypercoagulated state is considered by Jaffe and multiple tests have been used to assess the presence of accelerated coagulation. About 40% of the patients have an increase in prothrombin consumption, a test which has proved to be the most sensitive indicator of accelerated coagulation. When the coagulation is normal, only about 25% recover. Other hematologic studies include CBC, platelet count, prothrombin time, and partial thromboplastin time (PTT). When the coagulation study is normal, proceed to number 5.

4. Viral Studies.

About 25% of patients with sudden deafness have an antecedent upper respiratory infection. The documentation of an acute infection should include virus isolation from swabs of the nasopharynx and a fourfold or higher rise of antibody titer when comparing the acute and convalescent sera. Specimens for virus studies should be obtained as early as possible and within 21 days of the onset of deafness. Specimen sources include whole clotted blood, stool, washings from throat or nasopharynx, cerebrospinal fluid, and fluid from the middle ear. Jaffe and Maassab, and others, identified adenovirus types I and III, Mycoplasma pneumoniae, and parainfluenza. About 60% of the patients with viral infections will recover spontaneously. No specific antiviral therapy exists.

5. Exploration of Middle Ear.

When the coagulation studies are normal and when there is a history of otologic trauma or physical exertion prior to the onset of sudden deafness, an exploration of the middle ear is indicated to rule out a fistula of the oval window or round window leakage of perilymph from the inner ear. A plug of Gelfoam or fat may be used to seal the fistula.

6. Glycerin Test.

A new application of the glycerin test in the diagnosis of posttraumatic perilymph fistula was described by Lehrer et al. Temporary disappearance of abnormal responses to the fistula and Quix tests and improvement in the hearing occurred. The glycerin test was useful in confirming the diagnosis of posttraumatic fistula in 13 patients in whom fistulas were found at tympanotomy.

7. Medical Therapy.

When coagulation studies indicate accelerated coagulation, one should proceed with the medical therapy.

a. Ambulatory care: Vasodilating drugs such as nylidrin (Arlidin), nicotinic acid, and nicotinyl alcohol (Roniacol) have been used. Suga and Snow showed that nicotinic acid, even in massive doses, has no measurable effect on cochlear blood flow.

b. Hospitalization should include: (1) Bed rest; (2) Intravenous histamine given as 2.75 mg in 500 mL normal saline over 30 min daily for 3 days with careful monitoring of blood pressure and pulse every 5 minutes; (3) low-molecular-weight dextran (10%) given intravenously, 500 mL every 12 hours for 3 days; (4) heparin given to maintain a clotting time of two to three times normal.

c. Anticoagulants, steroids, and Urografin may be effective in some cases.

d. For idiopathic sudden sensorineural deafness, Meyerhoff suggests:

1) Hospitalization and bed rest.

2) Heparin 5000-10.000 units subcutaneously every 12 hours.

3) 40 units of adrenocorticotropic hormone (ACTH) IM each morning.

4) Low-molecular-weight dextran (10%) IV in 500 mL quantities over a 4-hour period; repeated every 12 hours.

5) Papaverine-hydrochloride (smooth muscle relaxant) orally 150 mg every 12 hours.

6) The above regimen is continued for 5 days. The progress is monitored with daily audiograms. If no improvement is seen, the patient is evaluated neuro-otologically. If the possibility of a perilymph fistula leak exists, tympanotomy is performed.

8. X-Ray Studies of Internal Auditory Canal.

When the audiometric studies indicate a retrocochlear loss, radiographic examination of the internal auditory canal should be performed (see Chap. 24).

9. Posterior Fossa Myelogram.

If the retrocochlear hearing loss does not recover and if the hearing loss is mild or moderate, one should perform a posterior fossa myelogram. If the posterior fossa myelogram is abnormal, acoustic neuroma is suggested and removal indicated. If the posterior fossa myelogram is normal the patient must be followed every 6 months with repeat audiograms, repeat posterior fossa myelogram, or repeat diagnostic labyrinthotomy.

10. Perilymph Tap (Diagnostic Labyrinthotomy).

Small acoustic neuromas produce an elevated protein content of the perilymph of the inner ear while cerebrospinal fluid protein levels remain normal. Because the diagnostic labyrinthotomy may produce a worsening of the hearing loss, it should not be used to evaluate a mild or moderate hearing loss.

For diagnostic labyrinthotomy, under local anesthesia, a tympanomeatal flap is created and the middle ear entered. The footplate of the stapes is identified, cleaned of mucosa, and all bleeding controlled with Adrenalin. Then a hole is made in the footplate and a capillary tube is placed into the vestibule of the inner ear and perilymph obtained. Analysis of the fluid will be diagnostic of an acoustic neuroma if the protein content is 1000 mg% or greater.

11. Removal of Acoustic Neuroma.

Three major approaches to the internal auditory canal are available: (a) translabyrinthine approach, (b) middle cranial fossa approach, and (c) combined suboccipital and translabyrinthine approach (see following section: Benign Tumors of the Ear).

The translabyrinthine approach destroys hearing so it is used only if severe or total deafness is present or if the discrimination is so poor as to produce a nonfunctioning ear. The middle cranial fossa approach is somewhat more complicated by intracranial complications but it is possible in some cases to remove the tumor, while preserving the hearing and facial nerve function.

Prognosis and Rehabilitation

1. Approximately one-third of patients have a return of normal hearing, one-third are left with a 40-60 dB speech reception threshold, and one-third have total loss of useful hearing.

2. Spontaneous recovery to normal hearing is more likely to occur if the deafness is not associated with severe vertigo and if the deafness initially is not total.

3. Once recovery of hearing begins, it usually takes place rapidly in a matter of a few

days. The longer the delay between the onset of deafness and the onset of recovery, the worse the prognosis for complete recovery.

4. Children who do not recover spontaneously from a unilateral sudden deafness should have preferential seating in school. Adults should be advised of the availability of a CROS (contralateral routing of signals) hearing aid.

5. Those patients who do not recover serviceable hearing from bilateral sudden deafness should have speech reading and auditory training. A hearing aid should be tried and used when appropriate.

6. When sudden deafness develops in an only hearing ear, exploration of the middle ear is indicated to rule out a hidden oval or round window fistula.

7. Priapism (a painful penile erection) as an unusual complication of heparin therapy for sudden deafness is reported by Clark et al.

Therapeutic Protocols

Therapeutic protocols

Therapeutic protocols suggested by the National Registry for Idiopathic Sudden Deafness include:

Method A:

1. Bed rest for 3 days with barbiturate or phenothiazine sedation.

2. Atropine (only if begun within 4 hours of the onset of deafness): 0.75 mg IM or in 250 mL of 5% dextrose in water IC as a single dose.

3. Procaine: 0.2% in 250 mL of 5% dextrose in water IV twice daily for 3 days.

4. Diphenhydramine (Benadryl): 50 mg 4 times daily orally or by injection until the hearing has stabilized for at least 3 weeks.

5. Nylidrin (Arlidin): 6 mg orally four times daily until the hearing has stabilized for at least 3 weeks.

6. Ascorbic acid (vitamin C): 1000 mg in each IV of procaine.

Method B:

1. Bed rest for 3 days.

2. Histamine phosphate: 2.75 mg in 250 mL of 5% dextrose in water IC daily for 3 days. Adjust the rate to produce a flush bot not a headache. Do not run at the same time as dextran as this seems to produce a severe headache.

3. Dextran: 10% (Rheomacrodex), 500 mL every 12 hours for 3 days.

4. Nicotinic acid in a flushing dose (50-300 mg) before meals and at bedtime, until hearing has stabilized for at least 3 weeks. This is begun in the office when the patient is first seen.

Method C:

1. Hospitalization for at least 3 days.

2. Heparin: 200 mg (20.000 units) every 12 hours IM, IV, or subcutaneously to keep the Lee White clotting time between 15-20 minutes which is two to three times normal.

3. Warfarin (Coumadin) is begun on the secon day and then the heparin is discontinued when the prothrombin time is two and a half to three times normal. Anticoagulation is continued for 4 weeks. Medical consultation should be obtained to manage the heparin and Coumadin anticoagulation.

4. Procaine: 0.2% in 250 mL of 5% dextrose in water IV twice daily for 3 days.

Method D: Prednisone: 10 mg three times daily for 10 days, then in reducing amounts to nil over 10 days.

Method E: Exploratory tympanotomy to seal a perforated round or oval window.

Method F:

1. Bed rest for 3 days or until hearing and/or vertigo is stable.

2. No medication: It is possible that the previously recommended medications have no effect or even an adverse effect on the prognosis. This group would serve as a control.

Benign Tumors of the Ear

External Ear

Benign tumors of the auricle and the external auditory canal include angioma (capillary hemangioma, cavernous hemangioma and lymphangioma), dermoid tumors, cylindroma, melanoma, Winkler's disease, osteoma, exostosis, adenoma, ceruminoma, chondroma, lipoma, xanthoma, myoma, myxoma, mixed tumors of the salivary gland type, and keratosis obturans.

Osteoma

Osteoma (cancellous) occurs in the auditory canal as a single large pedunculated tumor near the lateral end of the bony portion on one side only. It arises from the region of either the tympanosquamous suture or the tympanomastoid suture. Hearing loss and discomfort are common symptoms.

Treatment. The treatment is surgical removal when symptomatic.

Exostosis

Exostoses (dense ivory compact bone) are the most common tumors of the external auditory canal. These are usually bilateral and asymptomatic unlewss accompanied by accumulation of debris against the tympanic membranes resulting in infection or obstruction. The causative factor most likely responsible is prolonged and repeated stimulation of the external auditory canal by cold water. Exostoses most commonly are seen in saltwater swimmers. Other factors which might play a role include chronic irritation from infection, eczema, and trauma.

Treatment. Treatment consists of surgical excision when symptomatic. Attempts to remove exostoses with a hammer and gauge may produce fractures in the surrounding bone, possibly resulting in facial palsy. Removal should be done with a small cutting burr. In some cases exostoses can be removed through a speculum, raising a flap of meatal skin by an incision external to the exostoses. In others, an endaural or postauricular incision is necessary.

Cerumenoma

This uncommon tumor is an adenoma of sweat gland origin and presents as a smooth intraverted polypoid swelling in the outer end of the meatus. This tumor may become malignant.

Treatment. Local recurrence after excision is common. Wide excision including a margin of healthy skin is advised.

Winkler's Disease

This is a rare, painful nodular growth of unknown origin occurring on top of the helix and mostly in men. It consists of tiny arteriovenous anastomosis with many nerve endings similar to a glomus body. The small nodule is painfully tender, preventing some patients from sleeping on the ear.

Treatment: Surgical excision or injection of cortisone for the relief of pain is the usual treatment.

Keratosis Obturans

This rare condition also is called cholesteatoma of the external auditory canal, and is characterized by an accumulation of large plugs of desquamating squamous epithelium (cholesteatoma) deep within the external auditory canal. It is often associated with chronic pulmonary disease, sinusitis, and bronchiectasis. Pain is the common presenting symptom and results from erosion of the bony canal (destructive or invasive keratitis). Hearing loss is usual.

Etiology. The cause is unknown, but probably is due to faulty migration of squamous epithelial cells from the surface of the tympanic membrane and adjacent canal.

Treatment. This consists of periodic removal of accumulated debris. General anesthesia may be required.

Middle Ear

Bengin tumors of the middle ear and mastoid include glomus jugulare tumor, osteoma, neurinoma of the seventh and eight cranial nerves, intratympanic meningioma, glioma, cylindroma, dermoid cysts, hemangioma, and acoustic neuroma.

Glomus Jugulare Tumor

This tumor is also called: chemodectoma, nonchromaffin paraganglioma, carotid body tumor, tympanic body tumor, and glomus tympanicum. Glomus jugulare tumor arises from the glomus bodies located in the adventitia of the dome of the jugular bulb or along branches of the tympanic plexus.

Pathology. Glomus jugulare tumors usually arise in the hypotympanum at the site of the entrance of Jacobson's nerve in the adventitia of the jugular bulb. In many instances they arise on the promontory. The incidence is five times more frequent in women than in men. The tumors consist of vascular sinuses supplied by the ascending pharyngeal artery which enters the tympanum along with Jacobsen's nerve. These tumors grow slowly but are progressively destructive by invasion into the surrounding structures. Instances of multicentric origin and association with carotid body tumors have been reported. The tumors occasionally metastasize to the lungs and cervical nodes.

Clinical Features. The earliest symptom is a pulsating tinnitus which is synchronous with the pulse. The hearing loss follows as the tumor enlarges. As it invades the tympanic membrane, spontaneous bleeding and discharge due to secondary infection will occur. Isolated facial paralysis frequently is present. Invasion along the course of the jugular wall results in multiple involvement of the ninth, tenth, eleventh, and twelfth cranial nerves. Invasion of the cochlea and petrous tip is a late occurence resulting in sensorineural hearing loss and (rarely) paralysis of the fifth and sixth cranial nerves.

Diagnosis. Examination in the early stages may reveal a reddish swelling behind the tympanic membrane which with magnification may be seen to pulsate. If the drumhead is dull and the tumor small, this appearance may be confused with Schwartze's sign in otosclerosis. As the tumor enlarges, it causes the inferior portion of the tympanic membrane to bulge and finally burst revealing a smooth large red polypoid mass which bleeds very easily (and often massively) on manipulation. Application of pressure with the pneumatic otoscope causes the tumor to increase in pulsation. As the pneumatic pressure is raised to exceed systolic pressure, a sudden blanching occurs (Brown's pulsation sign). Radiographic examination is of little value in early cases but as the tumor advances will reveal the extent of bony destruction. In advanced tumors, external carotid angiography, polytomography, and retrograde jugularography are helpful in delineating the extent of tumor. The diagnosis is confirmed by biopsy which must be done carefully in the hospital because of the severe hemorrhage which may occur. With an intact tympanic membrane the biopsy is best done through a tympanotomy approach and combined with total excision.

Classification. Rosenwasser classified glomus tumors into three groups: group I include those cases in whom the tympanic membrane is intact and the lesion is small and confined to the middle ear; group II include those cases in whom the middle ear, aditus, antrum, and mastoid bone appear to be involved, and group III include those cases in whom there is a wide spread extension, at times with intracranial involvement.

Alford and Gilford classified the tumor into five stages:

Stage O: The earliest manifestation of a glomus tumor. The patient complains of hearing loss and/or pulsating tinnitus. There will be normal hearing or a conductive hearing loss. The drum head is intact but discolored. Radiographs will be normal.

Stage I: Aural discharge due to involvement of the tympanic membrane by the tumor is noted. Radiographs show clouding of the middle ear but no bone erosion. There is no cranial nerve involvement.

Stage II: Facial paralysis now present, and there is sensorineural hearing loss. Radiographs may show enlargement of the jugular forament but no bone erosion.

Stage III: Involvement of the jugular foramen with paralysis of the ninth, tenth, eleventh, and/or seventh cranial nerve. Radiographic evidence of erosion of the petrous bone and enlargement of the jugular foramen is noted.

Stage IV: Intracranial extension producing papilledema, extensive involvement of the petrous bone, and paralysis of the third, fourth, fifth, sixth, and seventh cranial nerves.

Treatment. Early cases may be cured by surgical excision but advanced cases should be treated with palliative radiation. The treatment may be summarized as follows:

Group I: Tumor involving the middle ear only (glomus tympanicum). Excision via:

1. Tympanotomy.

2. Endaural hypotympanotomy of Shambaugh.

3. Postauricular facial recess approach.

Group II: Tumor extending into the attic or mastoid: Radical mastoidectomy and hypotympanic resection (with ligation of ascending pharyngeal artery or external carotid artery) followed by radiation when removal was incomplete.

Group III: Tumor involving the jugular forament with cranial nerve paralysis:

1. Partial excision with radiation.

2. Combined radical mastoid and hypotympanic neck dissection.

3. Temporal bone resection with dissection and ligation of lateral sinus and jugular

vein.

4. Modified infratemporal fossa approach.

Radiation often is used especially when complete removal is not possible. Tumor doses of 2400 to 6000 rad are given over a period of 2-4 weeks. Palliative benefit from radiotherapy is probably due to reduction in vascularity as the tumor cells are not considered to be radiosensitive.

Internal Ear

Acoustic Neuroma

Acoustic neuroma accounts for 78% of all tumors of the cerebellopontine angle. It is found in about 8% of all intracranial tumors and constitutes even a greater percentage of all posterior fossa lesions. The tumor most often becomes symptomatic between the ages of 30-40 years. Routine autopsies have revealed a 2.4% incidence of asymptomatic acoustic neuroma. It is more common in females in a ratio of 3:2.

Pathology. Acoustic neuroma is a benign encapsulated tumor arising from the sheath of Schwann (neurilemma) of the eight nerve. The usual site of lesion is the vestibular portion of the nerve in the region of Scarpa's ganglion. The incidence of involvement of vestibular and auditory nerves is a 2:1 ratio. The eight nerve loses the neurilemmal sheath at the porus acousticus, and hence it is unlikely that the tumor would arise proximal to the porus. It usually begins in the internal auditory canal, slowly enlarges within the canal and with some degree of erosion extends toward the cerebellopontine angle. It is usually unilateral, but bilateral lesions may be noted as in the case of von Recklinghausen's disease. The size of the tumor may reach 5.0 cm although the more common size outside the canal is about 2.5-3.0 cm.

Histologically, the tumor is characterized by streams of elongated spindle cells, with the elongated nuclei often arrayed in a palisade pattern. Tumors in which there is a thick concentration of cells are called Antoni type A, whereas those in which the cells are loose are called Antoni type B.

Clinical Features

They include:

1. The earliest symptoms are tinnitus and unilateral progressive hearing impairment. In the early stages of acoustic neuroma, dizziness also is an extremely common complaint, appearing in the form of unsteadiness in about 83% of the patients.

2. Vertigo is not common initially, but may become a more prominent symptom with continued growth of the tumor.

3. Other early complaints are a sensation of prickling and itching, and pain in the ear.

4. Late manifestation of the disease develop from great pressure upon the auditory canal and extension of tumor into the posterior cranial fossa. The sensory part of the fifth nerve may become involved first, producing unilateral numbness of the face. The motor part of the seventh nerve may be affected, causing facial weakness. Eventually the cerebellum may be disturbed, producing slurring of speech, ataxia, and incoordination of one or both upper extremities. With continued growth the tumor can obstruct the flow of cerebrospinal fluid, creating an internal hydrocephalus. Headache, nausea, vomiting, and dullness of mental faculties may accompany these complications.

Diagnosis. Any patient suspected of acoustic neuroma should undergo a complete audiologic, vestibular, and neurologic evaluation.

Audiometric Examination. The audiometric examination indicates that:

1. All the patients have sensorineural hearing loss. Approximately 64% have a high-tone loss, but others can present with a flat-type curve.

2. Impairment of speech discrimination is much greater than would be expected from pure tone loss. Discrimination scores of 0-30% occur in over half of the patients.

3. The short increments sensitivity index (SISI score) is in the region of 0-35% with retrocochlear lesions.

4. Approximately 50-60% of the patients with acoustin tumors show type III and type IV Bekesy tracings.

Brain Stem Evoked Response Audiometry. Recent studies with the early part of the evoked response, presumably from the brain stem, indicate that pressure on the auditory nerve from tumors can cause an increase in latency or the elimination of early response waves. The waves found as a result of the click stimulus are best compared by evaluating the P-5 wave latency from two ears.

The between-the-ears (intra-auricular) latency is usually less than 0.2 msec, and is found to be 0.4 msec or greater for acoustic tumor cases. In 21/35 acoustic neuroma Selters and Brackmann reported that the P wave was completely absent. The intra-auricular latency difference was positive in 91% of these cases and measured from 0.4-3.2 msec.

In four patients the intra-auricular difference was less than 0.2 msec, but one of these patients had bilateral acoustic tumors. It was proposed that in the other three cases, intra-auricular latency was normal because of insufficient pressure on the cochlear nerve.

Although false-positive findings may occur in patients with severe (greater than 75 dB) hearing losses, the study is a most valuable noninvasive test to be added to the battery of tests used in the detection of acoustic neuromas when the hearing threshold is less than 70 dB.

Vestibular Examination. Diminished or absent response to caloric testing is an important and early sign of acoustic tumor. About 96% of patients with acoustic neuroma have an abnormal caloric response. Electronystagmography may show spontaneous nystagmus

away from the side of the lesion. In some patients positional nystagmus may be noted. Vertical nystagmus may suggest posterior fossa involvement.

Neurologic examination. With an increase in intracranial pressure there may be blurred optic discs, impairment of occular motor function, diminished sensation of the face, and facial weakness. Hitselberger and House noted hypesthesia of the posterior wall of the external auditory canal (Hitselberger's sign). Lacrimation, taste, and blink reflexes should be tested. A complete neurological examination should be a must in all suspected cases of acoustic neuroma.

Radiologic Evaluation (also see Chap. 24 Radiology)

1. The radiologic examination for an acoustic tumor usually includes conventional films, polytomography, computed tomography, posterior fossa myelogram, and arteriography.

2. The x-rays that are most valuable in evaluation of the internal auditory canal are Stenvers, transorbital, Towne's, and submentovertical projections. These views demonstrate enlargement of the canal or erosion of the petrous portion of the temporal bone in about 85% of the patients.

3. Computed tomography provides an excellent screening technique permitting diagnosis of acoustic neuromas over 2 cm in size when the examination is carried out with contrast enhancement. Since acoustic neuromas have tissue densities equal to those of the surrounding brain, enhancement with contrast is necessary to produce increases in density and well-circumscribed margins. All tumors over 2 cm in diameter usually are shown with contrast enhancement. The 20% false-negative studies may occur in patients with tumor sizes less than 2 cm.

4. When the screening studies such as conventional film and polytomography are positive and agree with the clinical and audiometric examinations, a CT scan then should be performed. If the latter is negative, Pantopaque myelography or air-CT should follow.

5. The decision to proceed to an invasive radiologic study for a small tumor recently has been based on results from brain stem evoked audiometry (BSEA). When BSEA is negative together with other audiometric studies, then invasive radiologic studies are not indicated.

6. A positive BSEA test indicating a retrocochlear lesion and combined with a negative CT scan should be followed by posterior fossa myelography or cisternography combined with CT.

7. It has become clear that the conventional CT scan cannot detect acoustic tumors in the cerebellopontine angle (CPA) that are less than 1.5 cm in size, or tumors that lie wholly within the internal auditory canal (IAC).

8. Posterior fossa myelography with Pantopaque remains the best diagnostic procedure for the small acoustic tumor (90-100% accuracy).

Laboratory Examination. In acoustic neuroma:

1. Cerebrospinal fluid examination shows elevated protein more often than in any other intracranial tumor.

2. A radioisotope brain scan may show a high incidence (85%) of positive scan.

3. Diagnostic labyrinthotomy: Small acoustic neuromas produce an elevated content of the perilymph of the inner ear in the presence of a normal content of the cerebrospinal fluid. Through a tympanotomy approach, a small hole is made in the footplate and a capillary tube is placed into the vestibule and the perilymph is obtained. A protein content of 1000 mg% or higher is diagnostic of an acoustic neuroma.

Differential Diagnosis. The following conditions may be confused with acoustic neuroma:

1. Endolymphatic hydrops is most easily and frequently confused with acoustic neuroma. Clinical history, further radiologic and laboratory tests will help to differentiate these lesions.

2. Cystic arachnoiditis of the cerebellopontine angle from previous acute or chronic otitis media producing otitic hydrocephalus may stimulate an acoustic neuroma. The history of otorrhea is important.

3. Meningioma arising from the posterior surface of the petrous pyramid produces the angle syndrome. Involvement of other cranial nerves generally occur earlier in meningioma, and loss of hearing and vestibular response occur later than in acoustic neuroma. The spinal fluid protein is generally not elevated in meningioma. Radiologic studies show hyperostosis, calcification, or destruction of the petrous pyramid, or an increased vascularity on angiography.

4. Congenital cholesteatoma of the petrous pyramid may produce the angle syndrome. Facial nerve paralysis occurs earlier than in acoustic neuroma and the x-ray changes are characteristic.

5. Multiple sclerosis may stimulate an angle tumor except for its characteristic remissions. Spinal fluid protein is not greatly elevated in multiple sclerosis.

Surgical Treatment. A classification of patients and a system of management on the surgical removal of acoustic tumors were suggested by Pulec et al and summarized as shown in Table 8-1.

Table 8-1. Classification and Surgical Management of Acoustic Neuromas

Site of Tumor
==> Associated Conditions
==> Management

Intracanalicular tumor (up to 8 mm diameter)
==> Some hearing
==> Removal via middle cranial fossa approach (facial nerve preserved and hearing can be saved in 60% of cases)

Intracanalicular tumor
==> No hearing
==> Translabyrinthine approach

Medium-sized tumor (2.5 cm in diameter)
==> With or without fifth nerve sign but no increased intracranial pressure, no papilledema, no cerebellar or long tract signs
==> Translabyrinthine approach

Large tumor (2.5 cm or more in diameter)
==> Increased intracranial pressure, fifth nerve signs, papilledema, cerebellar and long tract signs, headache, depressed mental ability
==> Suboccipital decompression with removal of occipital bone from the midline to the sigmoid sinus; removal of arch of the atlas, incision of the atlantooccipital ligament without opening the dura, followed in 5-7 days by translabyrinthine approach for removal of tumor.

Bilateral medium or large tumor
==> Useful hearing
==> Retrolabyrinthine approach with preservation of labyrinth and endolymphatic sac and removal of major portion of tumor leaving only small part over the sevent and eight nerves and internal auditory artery. Purpose: (1) preserve hearing, (2) relieve the life-threatening tumor mass. Repeat surgery when and if indicated.

Small, medium, or large tumor
==> High-risk case with disabling symptoms of vertigo, nausea, and ataxia
==> Translabyrinthine approach: to accomplish labyrinthectomy and brief subtotal removal of tumor.

The argon laser has been used routinely for acoustic tumor surgery by Glasscock et al. It has been used to vaporize and cut tissue and for photocoagulation in conjunction with a variety of surgical approaches to the cerebellopontine angle.

Tumor-Like Conditions of the Temporal Bone

Histiocytosis X (Reticuloendotheliosis)

The characteristics of the three clinical syndrome or variants of histiocytosis X (all involving the skin, skeleton, and reticuloendothelial system) may be summarized as follows.

Letterer-Siwe disease. This is a rare and rapidly fatal form of acute disseminated histiocytosis occurring in children before the age of 2 years, and characterized by fever, splenomegaly, hepatomegaly, lymphadenopathy, skeletal lesions, purplish skin rash, and anemia.

Hand-Schüller-Christian disease. This is a less severe and more chronic form in children and young adults, characterized by exophthalmos and diabetes insipidus from involvement of the sphenoid bone. When the temporal bone is affected, it may involve the mastoid cortex, the external auditory canal, labyrinth, the facial nerve, and the jugular foramen.

The characteristic histologic feature is the presence of lipoid-filled histiocytes (foam cells).

Treatment: Irradiation. The mortality is 30%.

Eosinophilic Granuloma. This is a less acute condition occuring in children and young adults, characterized by osteolytic lesions in one or several bone areas and a predilection for the frontal or temporal bone.

Otologic manifestations include swelling over the mastoid, granulations in the external auditory canal, otorrhea, deafness, and facial paralysis.

Histologically, the lesion presents two types of cells: (1) large, pale mononuclear histiocytes with mitotic figures, and (2) eosinophils.

Treatment is: (1) surgical excision or curettage of individual lesions; (2) radiotherapy for recurrences and inaccessible lesions. Useful for relief of pain.

Fibrous Dysplasia (Osteitis Fibrosa Cystica)

Fibrous dysplasia may involve a single bone (monostotic type), or, less often, several bones (polyostotic type). Fibrous dysplasia of bone usually first becomes manifest during childhood or early adult life and the lesions grow slowly. The polyostotic type characteristically involves the long bones and rarely the skull. The monostotic type may occur in the long bones, facial bones, or membranous bones. Occasionally the polyostotic type of fibrous dysplasia occurs in a form known as Albright's syndrome which is characterized by multiple involvement of the long bones, pigmentation of the skin, and precocious puberty in females.

Fibrous dysplasia of the temporal bone manifests itself as a painless swelling in the

region of the mastoid and the external auditory canal. Conductive hearing loss due to occlusion of the external auditory canal may be the only symptom.

Histologically, fibrous dysplasia is characterized by replacement of marrow with fibrous tissue containing spicules of bone undergoing resorption and formation. Active osteoblastic and osteoclastic activity usually is present and islands of cartilage may be seen.

Fibrous dysplasia has a female sex predominance in a ratio of 3:1. The onset is usually in childhood, and the lesions, when multiple, are often unilateral.

Differential diagnoses include hyperparathyroidism, Ollier's enchondromatosis, von Recklinghausen's disease, Paget's disease, Hand-Schüller-Christian disease, and other bone tumors. The history of painless swelling associated with the characteristic radiographic appearance (a typical loss of cellular structure and increased radiolucency due to replacement of osseous substance by fibrous tissue), and biopsy are adequate to differentiate fibrous dysplasia from these conditions.

Treatment of fibrous dysplasia is surgical excision. Radiotherapy appears to have a predisposing propensity to malignant degeneration and is considered as contraindicated for treatment of fibrous dysplasia.

Presbyacusis

This can be classified under the following subheadings:

Sensory

1. Atrophy of the organ of Corti and the auditory nerve in the basal end of the cochlea. Characterized by abrupt high-frequency loss.

2. Begins at middle age and is slowly progressive.

Neural

1. Loss of ganglion cells and degeneration of nerve fibers.

2. Significant disability in discrimination of speech.

3. Occurs late in life.

Metabolic

1. Stria atrophy.

2. Good discrimination.

3. Flat audiometric curve.

Mechanical

1. Descending audiometric curve. The basal turn is most involved.

2. Questionable stiffening of basilar membrane.

Miscellaneous

1. Seventy-five percent of ganglion cells can be missing and yet pure tone thresholds are maintained.

2. Loss of spiral ganglion cells does not necessarily produce hair cell degeneration.

3. Loss of hair cells with normal supporting cells does not produce spiral ganglion cell degeneration.

4. The stria vascuularis is the source of +80 mv DC potential of the scala media.