Paparella: Volume IV: Plastic and Reconstructive Surgery and Interrelated Disciplines

Section 2: Disciplines Closely Associated With Otolaryngology

Chapter 30: Imaging of the Temporal Bone

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The study of the temporal bone has always been a challenge to the radiologist. On the one hand, because of the different density of its bony components and the air and fluid spaces around and within them, the temporal bone lends itself to accurate visualization. On the other hand, the concentration in a small area of many important structures and the minuteness of those structures make the investigation quite difficult.

Several imaging techniques are available at present for the study of the temporal bone and of the neurovascular structures coursing through it: conventional radiography, computed tomography (CT), magnetic resonance (MR), and angiography.

Normal Radiographic Anatomy

A knowledge of normal radiographic anatomy is indispensable for the recognition and evaluation of pathologic conditions.

Conventional Radiography

Conventional radiography has the advantage of being a simple and inexpensive study that can be accomplished with standard radiographic equipment available in most departments. It has, however, the intrinsic defect of offering a picture that is the summation of a single plane of multiple structures located in different planes, so that the small structures under investigation are often more or less obscured.

Today, plain x-ray films are used only to evaluate mastoid pneumatization and to assess the position and integrity of cochlear implant electrodes. The integrity of implant wires cannot be established by tomographic techniques, since the wires are visualized in contiguous sections and their continuity cannot be demonstrated with certainty in any single section. Only three conventional radiographic projections are of practical interest: the lateral or Schüller's, the frontal or transorbital, and the oblique or Stenvers'. There are other projections of historical significance, but they have no useful contemporary application.

The Schüller or Runström projection is a lateral view of the mastoid obtained with the sagittal plane of the skull parallel to the film and with a 30-degree cephalo-caudad angulation of the x-ray beam. It allows an excellent visualization of the extent of the pneumatization of the mastoid, of the distribution and degree of aeration of the air cells, of the status of the trabecular pattern, and of the position of the vertical portion of the sigmoid sinus. Of the middle ear cavity, only the upper portion of the attic is usually visible, the remainder being obscured by the superimposition of the petrous pyramid.

The transorbital projection is obtained with the patient's back to the film in order to magnify the orbit and with the chin slightly flexed until the orbitomeatal line is perpendicular to the table top. In this view the petrous pyramid is clearly visualized through the radiolucency of the orbit. In particular, the internal auditory canal is outlined in its full length from the medial wall of the vestibule to the well-defined and smooth margin, concave medially, formed by the free margin of the posterior wall of the canal. In addition, the cochlea, vestibule, and semicircular canals are easily recognizable.

The Stenvers' projection is obtained with the patient facing the film with the head slightly flexed and rotated 45 degrees toward the side opposite to the side under examination. The x-ray beam is angulated 14 degrees caudad. Because of the rotation, the long axis of the petrous pyramid becomes parallel to the plane of the film, and the entire pyramid is well visualized, including its apex. The porus or medial opening of the internal auditory canal seen on face appears as an oval-shaped radiolucency open medially. Lateral to the porus the internal auditory canal is seen quite foreshortened because of the rotation. The remaining inner ear structures are usually recognizable, especially the posterior semicircular canal, which now lies in a plane parallel to the film.

Tomography

Computed tomography has replaced multidirectional tomography as the radiographic study for the assessment of the temporal bone. CT sections are easier to interpret because of the high contrast of the images and better recognition of soft tissue structures and pathology. In addition, CT allows, by the change of the window width and window level, assessment of various parameters of the image, including adjacent intracranial and extracranial structures and pathology.

Three prerequisites are necessary for the study of the temporal bone: high definition, thin sections, and multiple projections.

High Definition. In the late generation of the CT scanners the pixel has been reduced to 0.25 mm and the voxel to 0.1 cu mm.

This Sections. By narrowing of the collimation of the x-ray beam and of the aperture of each detector, the slice thickness has been reduced to 1.5 mm. For serial sections a 1-mm increment can be obtained by 0.5-mm overlapping of the sections.

Multiple Projections. The CT study of the temporal bone should always include at least two projections. Use of a single projection may lead to serious mistakes since structures that are parallel to the plane of section are only partially or not at all visualized. For instance the floor of the external auditory canal and the tegmen cannot be evaluated in the axial sections. The basic projection is of course the axial, since it is the easiest to obtain. The patient lies supine on the table with the canthomeatal plane perpendicular to the table top. Because of the impossibility of showing the entire series of sections in this chapter, we have selected few representative images for this and the other projections. Direct coronal sections, either prone or supine, can be obtained in most patients by extending the patient's head and tilting the gantry. For the 20-degree coronal oblique projection the patient lies prone on the table. The patient's head is extended as for the coronal sections and then rotated 20 degrees

toward the side under examination. In this way the medial or labyrinthine wall of the middle ear cavity becomes perpendicular to the plane of section. This projection is particularly useful for study of the oval window. The direct lateral projection is instead extremely hard on the patient or impossible. Whenever it is necessary, images in this projection are obtained by computer reformatting. Reformatted images, however, are always suboptimal because of intrinsic distortion and volume averaging, and are further deteriorated by motion, which may occur not only during scanning but also during the longer interscan time.

A second series of scans performed in one or more projections following bolus and drip infusion of contrast material is mandatory whenever a vascular anomaly or an otogenic brain abscess is suspected, and in all tumors except for the osteoma. In these cases CT demonstrates not only the involvement of the temporal bone but also the presence and extension of the intra- or extracranial component of the lesion.

Recently, fast rotational (dynamic) CT scanning has been used for the diagnosis and differentiation of vascular masses in the temporal bone. A bolus of 30 to 45 mL of contrast material is injected rapidly into a vein of the arm, and six to 12 scans of a preselected section showing the mass under investigations are obtained during a 30-second period. The first image exposes the mass before the contrast arrives. Subsequent images demonstrate the enhancement of the mass during the scanning period. A graphic plot of the absorption changes is obtained by plotting the density of the mass measured in CT numbers on the X axis and the time one the Y axis. A vascular mass is characterized by a high peak (30 or more CT numbers) with rapid wash-in and wash-out phases. The time of peaking will allow differentiation of a vascular tumor from an aberrant artery or vein.

Magnetic Resonance (MR)

MR is an imaging modality capable of producing cross-sectional images like its predecessor, x-ray CT. Unlike CT, it does not use ionizing radiation, but rather an apparently safe interaction between protons or hydrogen nuclei, static magnetic fields, and radio-waves. Also unlike CT, tomographic images or proton resonance can be obtained in any plane.

The strength of the MR signal to be converted into imaging data depends on the concentration of the free hydrogen nuclei and on two magnetic relaxation times, T1 and T2, which are tissue specific.

Air, cortical bone, and calcifications contain very few free protons and therefore appear in the images as dark areas of no signal. Fat and body fluid are instead rich in free protons and produce signals of high intensity.

One of the characteristics of MR is the possibility of changing the appearance and therefore information of the images by changing the contribution of the T1 and T2 relaxation times. This is accomplished by varying the time between successive pulses (TR or repetition time) and the time the emitted signal or echo is measured after the pulse (TE or echo time).

Since cortical or nondiploic bone and air emit no signal, the normal mastoid, external auditory canal, and middle ear appear in the MR images as dark areas without pattern or structures within them. The petrous pyramid is equally dark except for a gray or white cast

of the inner ear structures and internal auditory canal produced by the fluid within their lumina.

Pathologic processes are demonstrated with MR whenever the hydrogen density and magnetic relaxation times of the pathologic tissues are different from the normal. Although MR is more sensitive than CT in the early identification of pathologic changes in the temporal bone, the exact location and extent of the abnormality and involvement of bony structures such as the ossicles, scutum, and labyrinthine capsule cannot be detected. For this reason, CT remains the study of choice to assess intratemporal bone pathology. If a lesion extends outside the confines of the temporal bone, MR usually defines intracranial and extracranial extent more precisely than CT.

Angiography

Angiography is seldom required for the diagnosis of vascular tumors or anomalies within or adjacent to the temporal bone.

However, arteriography is mandatory for identifying the feeding vessels of the lesion, usually a glomus tumor, whenever embolization or surgical ligation is contemplated. Subtraction is necessary to delineate the vascular mass, and feeding vessels that otherwise are obscured by the density of the surrounding temporal bones. The injection should be made in the common carotid to visualize both internal and external carotid circulation. A vertebral arteriogram may also be performed.

Retrograde jugular venography is rarely used for the diagnosis of a high jugular bulb or of a glomus jugulare tumor. The study is done by percutaneous puncture of the vein with a Seldinger needle. The stylet is withdrawn and a guide wire advanced through the needle lumen into the internal jugular vein to the bony roof of the jugular fossa. The needle is then removed and a radiopaque polyethylene catheter threaded over the guide wire, which is in turn removed.

Pathologic Conditions

The major categories of pathologic conditions involving the ear are congenital malformations, traumatic effects, inflammatory processes, neoplasms, and otodystrophies. Cholesteatomas and tympanosclerosis will be included under inflammatory processes since they usually occur in association with, or as a complication of, these disorders.

It is of great advantage to the otolaryngologist to know the nature and extent of the pathologic process in deciding whether corrective surgery may be attempted. In addition, whenever surgery can be performed, imaging may furnish important information leading to the selection of the most suitable surgical approach.

Congenital Malformations

Conventional radiography is of limited value in diagnosing congenital malformations, except in the evaluation of the degree of development of the mastoid, because of the frequent absence of the normal radiolucencies of the mastoid air cells, external auditory canal, and

middle ear cavity; the presence of a dense atretic block obscuring the superimposed structures; and finally the distortion in axis and location of some of the structures. All these factors make the interpretation of a conventional radiogram extremely difficult or impossible. CT should be performed in at least two projections, axial and coronal, with sections 1 or 1.5 mm apart. Lateral sections may be required for the assessment of the vertical or mastoid segment of the facial canal and of the vestibular aqueduct.

Our records include a series of 800 cases of congenital malformations of the ear classified as follows: 60 per cent of patients with anomalies of the sound conducting system, 30 per cent with anomalies of the inner ear structures, and the remaining 10 per cent representing a group of patients with combined external, middle, and inner ear anomalies.

Malformations of the Sound Conducting System. The impossibility of a direct otoscopic examination in most of these conditions resulting from a complete or partial atresia of the external auditory canal makes the CT study more essential than in any other pathologic process. In addition, this group is unquestionably the most important of the congenital anomalies, not only because it is the most frequently encountered but above all because a functional surgical repair can often be successfully performed. The branchial origin of the middle ear cavity and ossicles explains the frequent association of abnormalities of other structures of the same embryologic derivation; a typical example of this association is mandibulofacial dysostosis, also known as the Treacher Collins or Franceschetti syndrome.

A proper CT study can provide the surgeon with the following information, which is of basic importance in deciding whether and how to perform corrective surgery:

1. The degree and type of abnormality of the tympanic bone, which may range from a minor deformity of the external auditory canal to a complete agenesis.

2. The degree and localization of development of the mastoid cells and mastoid antrum.

3. The position of the sigmoid sinus and jugular bulb. It is not uncommon to demonstrate a deep jugular fossa protruding from below into the hypotympanic or tympanic cavity. Usually the jugular bulb remains covered by a thin shell of bone, but occasionally it may bulge without covering bone in the hypotympanum.

4. The degree of development of the middle ear cavity and ossicular chain.

5. The status of the labyrinthine windows.

6. The route of the facial nerve canal. A more anterior position than usual of the vertical or mastoid portion of the facial nerve canal is quite common in cases of aplasia or hypoplasia of the external auditory canal and middle ear cavity. In less frequent, but not rare, cases, the third portion of the facial nerve canal may be grossly ectopic and run horizontally outward.

7. The relationship of the meninges to the mastoid and petrous ridge. A low-lying dura is frequently encountered as the middle cranial fossa deepens to form a large groove lateral

to the labyrinth. Occasionally a dehiscence may be demonstrated in the tegmen with a soft tissue mass protruding into the middle ear cavity from above, indicative of a meningocele or meningoencephalocele.

Malformations of the Inner Ear Structures. Congenital sensorineural deafness and vestibular loss may be the result of abnormalities involving the membranous portion of the labyrinth only (and therefore not radiographically demonstrable) or of anomalies involving both the membranous labyrinth and the otic capsule (and therefore roentgenographically visible). Congenital anomalies of the inner ear structures range from complete agenesis or aplasia to hypoplasia of one or more of the structures.

The most severe anomaly of the otic capsule is the Michel type of deformity, which is characterized by a hypoplastic petrous pyramid and a complete or almost complete lack of development of the inner ear structures. A single labyrinthine cavity of varying size is often observed, which occupies the space normally taken by the vestibule, cochlea, and semicircular canals.

A less severe anomaly of the labyrinthine capsule is the Mondini type of deformity, which is characterized by an abnormal development of the cochlea and is often associated with an abnormality of the vestibular aqueduct, vestibule, and semicircular canals. The cochlea may be hypoplastic or of normal size, but the bony partition between the cochlear coils is hypoplastic or absent, giving the appearance of an empty cochlea. The vestibular aqueduct is often shortened and dilated. The vestibule appears larger than normal, and there is often dilatation of the ampullated portions of the horizontal and superior semicircular canals.

A dilated and shortened vestibular aqueduct, similar in appearance to that of the Mondini deformity, may often be present without other radiographic abnormalities.

A common anomaly of the otic capsule seen tomographically is an abnormal development of the horizontal semicircular canal. The horizontal canal may be shortened and dilated, or may appear as a single lateral outpouching of the vestibule without the bony core around which the canal would normally loop. Such an isolated anomaly of the horizontal canal may occur with normal cochlear and vestibular function.

Hypoplasia or agenesis of the internal auditory canal rarely occurs as an isolated malformation but is usually associated with other anomalies of the otic capsule.

In approximately 20 per cent of ears with congenital anomalies of the otic capsule, the cochlear aqueduct may be abnormally dilated. Dilatation of the cochlear aqueduct is occasionally the only anomaly radiographically visible in patients with congenital sensorineural hearing loss. It is conceivable that malformations of the membranous labyrinth coexist in such cases, but they are not demonstrable by present imaging techniques.

The recognition by CT of inner ear anomalies has allowed the diagnosis during life of conditions that in the past were recognized only at autopsy. In addition, evaluation of the type and extent of the malformation is necessary whenever a cochlear implant is contemplated.

Vascular Anomalies. Whenever a vascular anomaly is suspected on the basis of the clinical findings or the preinfusion CT examination, a repeat study after bolus and drip injection of contrast material is mandatory. Dynamic CT scanning of a single preselected plane or sequential with table incrementation is extremely useful in these patients. This technique has been described previously. The most important vascular anomalies are:

1. An anterior position of the vertical segment of the sigmoid sinus.

2. A high jugular bulb projecting into the hypo- or mesotympanum.

3. An ectopic course of the internal carotid artery within the middle ear cavity.

Traumatic Effects

The petrous pyramid, mastoid, and tympanic bone are unquestionably much more often involved in a fracture following trauma to the skull than is demonstrated by standard radiographic examinations. These structures form, in fact, the most common site of fractures of the base of the skull. The demonstration of a fracture is important for the therapeutic approach and from the medicolegal aspect.

One should be aware that whereas fractures with separation or displacement of the fragments can be easily demonstrated by conventional radiography or CT, small fracture lines without displacement and separation of the fragments can be detected only by CT if the plane of the fracture lies in, or close to, the direction of the x-ray beam. A fracture line may disappear at a certain level only to reappear a few millimeters distant. This apparent gap is not due to interruption of the fracture but rather to the fact that the plane of the fracture line changes course and becomes invisible in some tomographic sections.

One or more of the three following clinical findings are present in patients referred for radiographic studies:

1. Cerebrospinal fluid otorrhea, usually due to a fracture extending from the superior wall of the external auditory canal to the floor of the middle cranial fossa or to a fracture of the tegmen, when a tear of the tympanic membrane is present.

2. Paralysis of the facial nerve, due to the involvement of the facial nerve with consequent simple compression or complete tear of the nerve. The CT findings vary from a complete disruption of a segment of the facial canal to transection of the canal by a fracture line and finally to separation of a fragment of the canal wall. In two of our cases the body of the incus was luxated and propelled with its short process into the facial canal just distal to the pyramidal turn. The facial canal may be injured either by transverse fracture or by the more common longitudinal fracture of the petrous bone. Contrary to the common belief and to the traditional data available in the literature, our case records indicate that the most common site of involvement of the facial canal in a longitudinal fracture is at or about the geniculate ganglion. In these instances the longitudinal fracture (after crossing the mastoid, the external auditory canal, and the attic) extends into the petrous pyramid anterior to the labyrinth and catches the facial canal in the region of the superficial and poorly protected anterior genu.

3. Hearing loss. Conductive hearing loss is the result of disruption of the ossicular chain most often encountered in fracture involving the attic. The most common type of dislocation involves the incus, whose fixation by ligaments is looser than that of the malleus. A rarer interruption of the ossicular chain is due to fracture of the crura of the stapes or separation at the incudostapedial joint. The diagnosis of this type of interruption can be made radiographically by direct visualization of the fragments or by detection of an abnormal rotation of the long process of the incus that has lost its normal relationship to the oval window.

Fractures involving the labyrinth, large enough to be radiographically demonstrable, usually produce a complete nerve or vestibular loss, or both. Longitudinal fractures of the petrous pyramid may involve one or more inner ear structures, but often skip the labyrinth by running just in front or back of it. Transverse fractures of the petrous pyramid may occur at any level, although they tend to follow the plane of least resistance. This plane usually runs from the dome of the jugular fossa, to the petrous ridge medial to the arcuate eminence, and across the vestibule and basilar turn of the cochlea.

Inflammatory Processes and Cholesteatomas

Acute Mastoiditis. This usually occurs in well-pneumatized temporal bones in patients with acute suppurative otitis media. This typical radiographic finding consists of a diffuse haziness or clouding of the mastoid air cells. In the initial stage of the process the trabecular pattern is intact. However, it appears less clear than usual because of the lack of the normal airborne interface, as a result of the edema of the mucosa or collection of fluid in the air cells, or both. A similar involvement is, of course, present in the air cells that have developed in the petrosa. Whenever the infection is not arrested by proper therapy, necrosis of the cell walls develops, which may lead to the formation of abscesses. A conventional lateral view such as the Schüller projection is usually satisfactory for the study of the mastoid, but CT sections should be added whenever further information concerning the middle ear cavity and ossicular chain is requested. In the differential diagnosis one should remember that reticuloendothelioses (in particular, eosinophilic granuloma) produce changes that are quite similar to acute mastoiditis and petrositis with areas of coalescence. However, the clinical course and the otoscopic findings are usually helpful in the differential diagnosis.

Chronic Mastoiditis. This is the result of long-standing or recurrent infectious processes. The typical radiographic findings consist of diffuse sclerosis of the trabecular pattern and diffuse clouding of the air cells and, if no perforation is present, of the middle ear cavity. Because of the thickening of the trabeculae, the air cells become constricted at first and later are completely obliterated. The lumen of the residual air cells, the mastoid antrum, and the middle ear cavity are usually filled with granulation tissue and therefore appear cloudy. Erosion of the long process of the incus is not an unusual finding.

Tympanosclerotic plaques are occasionally demonstrated as irregular calcifications in the middle ear cavity and attic. Ankylosis of the ossicles, especially the head of the malleus, is best diagnosed in axial CT sections by the filling in of the space between the anterior aspect of the head of the malleus and the attic wall.

Cholesteatoma. Cholesteatomas may be congenital or acquired. Congenital cholesteatomas arise from epithelial rests within or adjacent to the temporal bone. Acquired cholesteatomas originate in the middle ear and extend into the mastoid and occasionally into the petrous pyramid. There is another distinct form of cholesteatoma that arises in the external auditory canal.

Acquired Cholesteatoma. Microscopic otoscopy usually provides a qualitative diagnosis of cholesteatoma but given no concept of the size of the lesion. The main purpose of the radiographic study is therefore to determine the degree and extent of the pathology of the cholesteatoma. This can be accomplished best by tomography. The pathognomonic findings of cholesteatomas are (1) erosion of the anterior portion of the lateral wall of the attic and of the anterior tympanic spine in cases of attic perforation, and (2) erosion of the posterior canal wall and of the posterior portion of the lateral wall of the attic in cases of posterosuperior marginal perforation. Erosion of the ossicular chain, widening of the aditus, and enlargement of the mastoid antrum by destruction of the adjacent trabecular pattern with formation of a well-defined cavity are other typical findings as the lesion progresses. If the cholesteatoma lies free in the middle ear or mastoid and is not surrounded by granulation tissue or serous fluid, it is possible to visualize the actual cholesteatoma sac as a soft tissue mass. However, when the cholesteatoma is surrounded by granulation tissue, it is difficult to discern radiographically where the actual cholesteatoma ends and the granulation tissue begins, since both soft tissues have similar radiographic density.

Congenital Cholesteatoma. Congenital cholesteatomas histologically are epidermoid tumors originating from embryonic epidermoid rests located anywhere in the temporal bone or adjacent epidural and meningeal spaces.

The clinical symptomatology of congenital cholesteatoma depends on the site and size of the lesion.

Middle Ear Cholesteatoma. Middle ear cholesteatomas appear as whitish globular masses lying medial to an intact tympanic membrane. There is usually no history of inflammatory ear disease. Occasionally there is an associated serous otitis media.

The CT study shows a well-defined soft tissue mass within the middle ear. If the cholesteatoma involves the entire middle ear space or if there is an accompanying serous otitis media, the entire tympanic cavity appears cloudy. The cholesteatoma mass may erode portions of the ossicular chain.

The inferior margin of the lateral epitympanic wall, which is typically eroded in acquired cholesteatoma, is intact in congenital lesions. The medial aspect of the lateral epitympanic wall is often eroded from within when the congenital lesion extends into the epitympanum.

Petrous Pyramid Cholesteatoma. Findings depend on whether the cholesteatoma arises from within the petrous apex or from the adjacent epidural or meningeal spaces.

When the cholesteatoma arises from within the petrous apex, CT shows an expansile, cystic lesion in the apex. The involved area of the pyramid is expanded and the superior

petrous ridge is usually elevated and thinned out. As the lesion expands, the internal auditory canal and the labyrinth become eroded.

Cholesteatomas arising from the epidural or meningeal spaces on the superior aspect of the pyramid cause a scooped-out defect of the adjacent aspect of the pyramid. The defect is caused by erosion of the pyramid from without, and there is no bony rim as in lesions arising from within the pyramid.

A CT study with infusion shows no enhancement of the mass except for its capsule. By CT it is difficult to differentiate a congenital cholesteatoma of the petrous apex from a cholesterol granuloma cyst that occurs in extensively pneumatized petrous pyramids. The two lesions can be differentiated by MR since congenital cholesteatomas produce a signal of medium intensity in the T1 images and high intensity in T2, whereas cholesterol granulomas have a similar high signal in both T1 and T2 sequences. In addition, areas of void are observed in cholesterol granulomas produced by deposits of hemosiderin.

Cholesteatoma of the External Auditory Canal. There are two types of this condition. The first, keratosis obliterans is caused by osteomas, stenosis of the canal, or hard mass of cerumen. Blockage of the external canal for a long period permits epithelial debris to accumulate in the canal and enlarge the bony contour of the external canal. The second type is called invasive keratitis and is characterized by localized accumulations of desquamated debris that occur on the floor of the bony canal. When the external canal cholesteatoma is large and reaches the annulus, the lesion erodes into the middle ear and attic.

Neoplastic Conditions

The most common of the benign tumors is certainly the osteoma, which can be found not only in the external auditory canal but often in the middle ear cavity, especially in the attic. The use of radiography in osteomas is usually limited to tumors obstructing the external auditory canal in order to determine whether any other disease is present behind the obstruction.

Malignant tumors of the ear are rare; among them, carcinoma occurs most frequently. The carcinoma is usually squamous cell type, originating in the external auditory canal and then extending into the middle ear, mastoid, and petrosa. The typical radiographic finding consists of destruction of the outline of the external auditory canal and middle ear, with a moth-eaten appearance of the mastoid as a result of the tendency of the neoplasm to infiltrate rather than erode bone.

Two lesions deserve special attention not only because of their relative frequency but, above all, because of the fundamental role played by imaging techniques in their diagnosis: glomus jugulare tumors and acoustic neuromas. Both tumors are usually histologically benign but often follow a malignant clinical course because of the large destruction produced in the base of the skull, and the involvement of the adjacent cranial nerves and central nervous system by the growing tumor mass.

Glomus Tumors. Glomus tumors arising in the tympanic cavity (glomus tympanicum) produce a soft tissue mass, usually well outlined in CT sections, in the lower portion of the

middle ear cavity, with possible thinning and destruction of the inferior wall of the hypotympanum. In such cases the differential diagnosis between a glomus and an ectopic or a high jugular bulb protruding into the middle ear cavity may be difficult without a jugular venogram or a CT dynamic study. The computer-generated density-time curve will demonstrate an early or arterial high peak (30 CT numbers or more) for a glomus tumor, but a delayed or venous high peak for a high jugular bulb. Glomus jugulare tumors arising in the jugular fossa also extend into the middle ear cavity through a destruction of its floor. In these cases the erosion of the cortical outline of the jugular fossa, the undermining of the petrous pyramid, and the involvement of the adjacent portion of the occipital bone (including the hypoglossal canal) are typical CT findings leading to the diagnosis. Large tumors protrude extradurally in the posterior cranial fossa and inferiorly below the base of the skull along the jugular vein. These extensions are better demonstrated by MR images where the tumor appears in both T1- and T2-weighted images as a mass of medium signal intensity containing several small areas of signal void produced by blood vessels. The signal intensity of the glomus is easily differentiated from the surrounding intra- and extracranial structures. In addition, MR allows determination of displacement, encroachment, narrowing, or obstruction of the jugular vein and internal carotid artery, since these large vessels are well visualized without the need for invasive vascular procedures. A carotid arteriogram should also be performed before embolization of the lesion or ligation of the feeding vessels.

Acoustic Neuromas. Acoustic neuromas account for approximately 10 per cent of unilateral sensorineural hearing loss and vestibular loss of unknown origin. They normally originate within the internal auditory canal and therefore produce osseous changes detectable by a proper radiographic study. The battery of radiographic tests for the diagnosis of acoustic neuroma can be divided into screening and diagnostic tests.

Screening Tests. Whenever conventional radiography is used, two projections are indispensable for study of the internal auditory canal: the transorbital view, which shows the canal in its full length, and the Stenvers' view for the study of the opening or porus of the canal. A more precise study of the internal auditory canal can, of course, be obtained by CT. Both sides should always be examined for comparison purposes. In fact, although there are slight variations in size and shape of the internal auditory canals of any one person, these variations are small compared with the difference between various individuals.

The following should always be examined to detect changes indicative of an acoustic neuroma (all figures have been corrected for magnification):

1. *Vertical diameter.* This normally ranges between 2 and 10 mm, with an average of 4.5 mm. An enlargement of 1 to 2 mm of any portion of the internal auditory canal under investigation in comparison with the corresponding segment of the opposite side should be considered questionable, and an enlargement of 2 mm or more definitely abnormal.

2. *Length of the posterior wall.* This normally ranges between 4 and 12 mm, with an average of 8 mm. Shortening of the posterior wall of one canal by 2 to 3 mm should be considered questionable, and shortening by 3 mm or more definitely abnormal.

3. *Crista falciformis.* This structure divides the canal into two compartments but is always located at or above the midpoint of the vertical diameter of the internal auditory canal.

A reverse of this ratio or an asymmetry by at least 2 mm in the position of the crista is strongly suggestive of an intracanalicular mass.

Diagnostic Tests. Acoustic neuroma is not visualized in plain CT scans since the tumor is isodense to the surrounding brain and is not surrounded by edema. Following infusion of iodinated contrast material the mass enhances and becomes visible. Intracanalicular lesion and cisternal masses smaller than 0.8 cm usually are not visualized by the infusion technique, and a CT pneumocisternogram is indicated. This examination requires a spinal puncture for the injection in the subarachnoid space of air, CO_2, or O_2. By proper positioning, the gas is moved into the cerebellopontine angle under investigation and several thin sections are obtained. In normal cases the gas fills the cerebellopontine cistern and the internal auditory canal outlining the seventh and eight cranial nerves. If a small tumor is present the gas outlines the localized swelling of the nerve, and if the tumor is larger the convex medial aspect of the mass obstructing the canal.

At present MR has become the study of choice for the diagnosis of acoustic neuromas without exposing the patient to ionizing radiation and without the necessity for spinal puncture. In the plain study the tumor appears in T1 images brighter than cerebrospinal fluid and isointense to gray matter. In T2 sections neuromas are brighter than brain but isointense to cerebrospinal fluid. Large tumors are easily identified in both sequences, but small lesions can only be detected in T1 images, since in the T2 they are obscured by the isointense cerebrospinal fluid. A further improvement in diagnosis of small tumors is achieved by injecting intravenously paramagnetic agents. Gadolinium-DPTA concentrates in the tumor and produces a shortening of the T1 relaxation time, with a consequent marked increase of the MR signal in T1 images. Tumors as small as 1 mm can be diagnosed by this technique.

Otodystrophies

We include two types of processes under the classification of otodystrophies: (1) a localized disease of the ear, otosclerosis; and (2) a group of diffuse processes such as Paget's disease, osteogenesis imperfecta, and fibrous dysplasia, in which the involvement of the ear could be one of the pathologic manifestations.

Both groups of diseases may produce (1) conductive hearing loss by involvement of the ossicular chain, and especially by the ankylosis of the stapes in the oval window; (2) sensorineural hearing loss by involvement of the inner ear structures; or (3) a mixed type of hearing loss by combination of the two types of involvement.

Fenestral Otosclerosis. Axial and 20-degree coronal oblique CT sections at 1-mm increments should be obtained.

The oval window is best seen in the 20-degree coronal oblique sections as a well-defined bony dehiscence in the lateral wall of the vestibule below the ampullated limb of the horizontal semicircular canal. The round window is instead more satisfactorily visualized in the axial sections through the lower portion of the promontory. The window appears as a gap or indentation in the contour of the posterior aspect of the basal turn.

The CT findings in fenestral otosclerosis vary with the severity and extent of the process. The changes range from loss of definition due to demineralization of the margin of the window, to narrowing and finally to complete obliteration of the oval window opening and niche. CT is particularly helpful in determining the cause of post-stapedectomy vertigo and hearing loss. It may disclose protrusion of the prosthesis into the vestibule, separation of the lateral end of the prosthesis from incus, dislocation of the medial end of the strut from the oval window, and reobliteration of the window with fixation or dislocation of the prosthesis.

Cochlear Otosclerosis. Otosclerotic foci arise in the enchondral layer of the capsule. In an active focus there is a loose and irregular network of bony trabeculae with numerous blood vessels, osteoblasts, and osteoclasts. In a mature focus there is a dense type of bone that is relatively avascular and acellular. These foci may progressively enlarge and extent to the periosteal and endosteal layers of the labyrinthine capsule. Active otosclerotic foci appear as single, multiple, or confluent areas of demineralization in the thickness of the capsule. Mature foci are recognizable only if their density is different from that of the normal otic capsule. Localized or diffuse areas of thickening and scalloping of the capsule are observed and are due to apposition of new otosclerotic bone.

The importance of CT in otosclerosis can be summarized as follows:

1. As a diagnostic test in questionable cases.

2. For evaluation of the degree, type, and extent of the process.

3. For selection of the side for corrective surgery.

4. For demonstration of possible abnormalities of the structures adjacent to the oval window, with special attention to the course of the facial nerve canal.

5. For evaluation of the postsurgical status, especially in relation to two points, i.e. recurrence of the process and position of the strut or prosthesis when radiopaque material is used.

6. For demonstration in patients with sensorineural deafness or foci of otosclerosis in the cochlea and in other areas of the otic capsule.

In Paget's disease the haversian bone of the pars petrosa is affected first, and because of the demineralization of this bone, the cochlear capsule and the ossicles at first become more prominent than usual. Then the otic capsule itself becomes involved, producing an irregular and fuzzy appearance and, finally, a complete disappearance of the outline of the inner ear structures. The progression of the involvement follows a path from the apex to the base of the petrous pyramid. The internal auditory canal is involved first, followed by the cochlea and the vestibular system. Narrowing of the oval window is often demonstrated.

Osteogenesis imperfecta involves the labyrinthine capsule, producing changes that are indistinguishable from the changes of otosclerosis, although usually more widespread to the entire labyrinthine capsule. However, the appearance of the long bones, which are abnormally

thin and have multiple fractures often leading to a gross distortion, and the blue color of the sclerae are unmistakable findings leading to the diagnosis.

The involvement of the skull by fibrous dysplasia is quite characteristic. Whereas the involvement of the calvarium and mandible consists of expansion of the affected portion by cystic lesions, the changes in the base of the skull, including the temporal bone, are almost always of the proliferative type. The affected petrous pyramid becomes extremely dense and thick with consequent asymmetry between the two sides. The outline of the labyrinthine capsule first becomes poorly distinguishable from the surrounding bone and finally may disappear as the lumen of the inner ear structures becomes partially or totally obliterated.

Involvement of the otic capsule, similar to the types described, has also been found in other rare conditions such as osteopetrosis or Albers-Schönberg disease, cleidocranial dysostosis, Hurler's syndrome or dysostosis multiplex, and Pyle's syndrome or craniometaphyseal dysplasia.