Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 5: Skull Base

Chapter 54: Acoustic Neuroma

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Successful surgical extirpation of tumors of the statico-acoustic nerve was pioneered first by Harvey Cushing and later by Walter Dandy in the first half of this century. During this era tumors were invariably large at diagnosis and most patients had increased intracranial pressure and brain stem shift. Removal was accomplished using a suboccipital approach in the sitting position. Subtotal (intracapsular) removal was the rule. Surgical mortality was high, and postoperative facial paralysis was a regular sequela. Nonetheless, a great many patients were granted extended life spans of good quality.

In the 1960s the translabyrinthine approach (condemned by Cushing) was resurrected and refined by William House. Dramatic advances in surgical anesthesia and early diagnosis, the introduction of the surgical microscope, and later the implementation of a "team" approach have combined to markedly reduce morbidity and mortality. Perioperative death is uncommon, and preservation of the facial nerve is now the rule. Tumor removal is virtually always complete and the majority of patients fully regain their preoperative lifestyle with normal life expectancies. Conservation of hearing has become increasingly realistic in selected patients with good preoperative hearing.

Pathology and Pathophysiology

The term *acoustic neuroma* is misleading. The neoplasm does not arise fro neural tissue, but rather from the investing Schwann cell. The tumors are well encapsulated and entirely benign. The growth rate is generally slow and meticulous; the history will often reveal symptoms going back many years. The vestibular division of cranial nerve VIII is almost always the nerve of origin. Acoustic neuromas arising from the cochlear division do occur, but they are very uncommon. Signs and symptoms develop primarily as a consequence of pressure effects that result from increasing tumor size. Therefore, tumors that arise on that portion of the nerve within the internal auditory canal, where room to grow is very limited, will produce symptoms when they are relatively small. Tumors that arise on the medial portion of the nerve within the cerebellopontine angle may grow to large size without producing symptoms at all.

Hearing loss has always been regarded as the hallmark of the acoustic neuroma and, indeed, the majority of patients experience hearing changes as the first symptom. The most common presentation is unilateral, slowly progressive, high-frequency hearing loss. The patient may report that he or she has increasing difficulty when conversations are directed to the affected

ear or that he or she can no longer use the telephone in the affected ear. Virtually any pattern of hearing loss, however, may develop. Low-tone losses, flat losses, and mid-frequency losses are well documented. Sudden hearing loss is a frequent mode of presentation for acoustic neuroma. Eventual recovery of some or all hearing is quite common. Indeed, some patients will present with a fluctuating hearing loss suggestive of endolymphatic hydrops. Five per cent of acoustic neuromas are discovered in patients with normal hearing. Any unilateral change in hearing requires that acoustic neuroma be considered.

Marked reduction in word discrimination is a hallmark of retrocochlear pathology and always suggests acoustic neuroma. However, many patients will have relatively normal discrimination. About 10 per cent of patients will present with a chief complaint of tinnitus. No distinctive subjective features have been found. Routine audiometry may be normal or show hearing loss of which the patient is not aware.

Vertigo is an uncommon presenting complaint. Vestibular ablation is so slow that excellent compensation is achieved with few apparent symptoms. Most patients, if specifically questioned, will report minor disturbances of balance. Often they have thought of themselves as having become "clumsy" during recent years. Less commonly, patients will report episodes of rotary vertigo lasting several seconds with rapid position changes. Occasional patients report longer episodes of severe rotatory vertigo lasting minutes to hours and associated with marked visceral autonomic symptoms. Such patients may or may not have associated hearing loss, tinnitus, or aural pressure. Patients with all the classic features of endolymphatic hydrops are occasionally found to have acoustic neuromas and all patients with Ménière's disease must be evaluated for cerebellopontine angle lesions.

Fifty years ago, the diagnosis of acoustic neuroma was rarely, if ever, made in the absence of neurologic symptoms. Today it is uncommon to discover patients whose tumors have reached so large a size as to produce neurologic deficits. The most common neurologic deficits are changes in sensation within the distribution of cranial nerve V. Anesthesia of the floor of the external auditory canal is the earliest sign (Hitzelberger's sign). Later, hypesthesia and/or paresthesias of the face may occur. The motor division of cranial nerve V is apparently more resistant to pressure effects, and weakness of the muscles of mastication is rare. Deficits of the trigeminal nerve are much more common than deficits of the facial nerve, but occasionally patients will manifest preoperative facial nerve weakness. Considering the marked thinning, attenuation, and distortion seen in the facial nerve at surgery, it is truly remarkable that they function so well.

Diagnosis

Early diagnosis is crucial. Morbidity and mortality is directly proportional to tumor size. In occasional patients early diagnosis of small tumors may allow preservation of hearing. Moreover, the larger the tumor, the poorer the chances for preserving normal facial nerve function. Since tumors may grow to large size with minimal symptoms, it is imperative that the otologist maintain a very high index of suspicion. All unilateral otologic complaints require

evaluation for acoustic neuroma. This evaluation should always go beyond routine audiometry because 5 per cent of patients with acoustic neuromas will have normal pure-tone audiograms and discrimination scores. We feel that the minimum evaluation of any unilateral otologic complaint is pure-tone audiometry with word discrimination scores and auditory brain stem response (ABR) audiometry. The inclusion of stapedius reflex decay, electronystagmography (ENG), and plain films of the internal auditory canals is useful (Table 1). Abnormality of any study other than routine audiometry should be pursued with a complete radiographic evaluation. Adequate radiographic evaluation requires high-resolution imaging of the cerebellopontine angle and internal auditory canals. Computerized tomography (CT), when performed with the administration of intravenous contrast material, will reliably demonstrate tumors 2.0 cm in diameter or larger. The use of contrast enhancement is essential. Acoustic neuromas are isodense compared with normal brain, and we have seen many unenhanced studies that fail to demonstrate even very large tumors. Moreover, care must be taken not only to ensure that the contrast is administered in adequate dosage but also that there has been sufficient time for the contrast material to diffuse into the tumor prior to scanning. Those patients who have evidence of retrocochlear lesions on audiometric evaluations or who have other signs and symptoms suggestive of an acoustic neuroma, such as cranial nerve V or VII abnormalities or headache, should have MRI scanning with gadolinium.

When MRI scanning is not available or if patients are excluded because they have surgically placed ferrous prosthesis, aneurysm clips, or ferrous foreign bodies, then CT arteriography of the posterior fossa may be considered. A variety of contrast media can be placed into the subarachnoid space, and all will adequately demonstrate even small tumors of cranial nerves VIII. Pantopaque, metrizimide, and CO_2 have all been used, but air is the contrast medium of choice. A small bolus of air is injected into the subarachnoid space by lumbar puncture and allowed to rise into the basal cistern. Adjustments of the patient's position allow the air to be manipulated into the internal auditory canal and cerebellopontine angle. Cranial nerves VII and VIII can then be individually visualized. Even very small tumors arising from these nerves can be clearly identified.

Magnetic resonance imaging (MRI) is an excellent technique for demonstrating cerebellopontine angle lesions. It is very sensitive and will detect small lesions especially if paramagnetic contrast agents such as gadolinium are utilized. Acoustic neuromas that are only a few millimeters in diameter routinely can be detected by this technique. No ionizing radiation is required. The only disadvantage is that bony details of the temporal bone are not seen. The exact relationship of the tumor to the porus acusticus and lateral end of the internal auditory canal is not appreciated on magnetic resonance imaging (MRI). If necessary, fine-cut CT of the temporal bone may be obtained as a complement to MRI to define the details of the bony anatomy.

Table 1. Test Results in Acoustic Neuroma

Audiogram

Unilateral, high-frequency loss most frequent but any pattern of neurosensory loss is possible, including normal audiometric findings.

Word Discrimination

Classically, word discrimination is reduced out of proportion to pure-tone loss (ie, less than 50 per cent) but any pattern is possible including normal.

Auditory Brain-Stem Response (ABR)

Retrocochlear findings including *ipsilateral:* absolute wave V latency prolongation, I-V interpeak latency prolongation, III-V interpeak latency prolongation; I-III interpeak latency prolongation is the most common and usually the most pronounced finding. *Contralateral:* Wave V latency prolongation in large tumors with brain stem shift ("offside" abnormality). *Stapedius Reflex:* (Ipsilateral): (a) Increased latency of response; (b) "decay" of response (greater than 50 per cent return to base line in 10 seconds).

Electronystagmography (ENG)

1. Most common: Ipsilateral marked hypofunction or absent response.

2. Occasional: Positional nystagmus.

Film Evaluation of IAC (Stenvers, or transorbital view)

Enlargement of the IAC (ie, greater than 3 mm higher or greater than 2 mm shorter than the contralateral IAC).

Differential Diagnosis

A number of lesions of the cerebellopontine angle will present with unilateral otologic complaints. It will not always be possible to distinguish these preoperatively from acoustic neuromas. However, if the lesions are anatomically suited to removal through the temporal bone little is lost in failing to make a preoperative determination of histology unless preoperative hearing is good. The likelihood of sparing hearing in patients with good preoperative aural acuity is greater with lesions other than acoustic neuroma.

Eighty per cent of tumors presenting in the cerebellopontine angle will be acoustic neuromas. Of the remaining 20 per cent, the vast majority will be meningiomas arising from dural fibroblasts of the basal meninges in the posterior fossa. About one-fourth of these tumors

will invade the temporal bone directly and extend extracranially into the neck or infratemporal fossa. Extracranial extension makes the diagnosis of acoustic neuroma unlikely and all primary cerebellopontine angle tumors with spread outside the cranium should be considered to be meningiomas. Those tumors limited to the cerebellopontine angle should be suspected of being meningiomas if they are eccentrically placed over the porus acusticus or if hearing is normal. Preoperative identification may be impossible and it is fortunate that the surgical approaches used for removal of acoustic neuromas are quite suitable for removal of these meningiomas.

Congenital cholesteatomas of the petrous apex make up most of the remaining cerebellopontine angle tumors encountered. These are keratinizing epidermoid inclusion cysts arising from congenital epithelial cell rests. Because of their very slow rate of growth, these lesions may develop to enormous size without producing symptoms. They are markedly hypodense compared to surrounding brain, and they are easily identified from CT scans. Although the usually presenting complaint is retrocochlear hearing loss, cranial nerve V and VII abnormalities are much more common in cholesteatomas of the petrous apex than in acoustic neuromas.

A variety of other benign lesions are uncommon and encountered only sporadically. These include arachnoid cysts, A-V malformations, hemangiomas, and lipomas. Malignant lesions are rare, but malignant schwannomas, gliomas, and malignant meningiomas are occasionally encountered.

Management

Patients with acoustic neuromas are best managed by a team consisting of a neurotologist and neurosurgeon. The neurotologist's expertise in the surgical anatomy of the temporal bone provides ready access to the tumor by the transtemporal and middle fossa approaches. He or she may play a crucial role in achieve complete tumor removal via suboccipital approaches by assuring access to the lateral portions of the internal auditory canal. The neurotologist's intimate knowledge of the three-dimensional anatomy of the facial nerve permits reliable identification of the facial nerve in the lateral-most portion of the internal auditory canal, where it is undistorted by tumor growth. This specialist's expertise in facial nerve surgery may be required for those patients in whom preservation of the facial nerve is not possible. Immediate re-anastomosis, cable grafting, or facial-hypoglossal anastomosis may be indicated. The neurosurgeon provides familiarity with brain stem anatomy, detailed knowledge of the vasculature of the posterior fossa, and experience in identifying the root entry zones of the cranial nerves. Both specialists have a large experience in microsurgery, which both utilize in all areas of their practice. Through use of a team approach, one can offer the patient an individualized treatment plan utilizing any of the four available surgical procedures singly or in combination.

Preoperative evaluation requires careful scrutiny of previously obtained radiographic studies. The presence of preoperative hydrocephalus should be noted, and thought should be give to placement of a ventricular shunt at the beginning of the operative procedure. Radiographs should be carefully examined for the presence of a contralateral or additional tumor as their

presence may alter management. Tumor size should be assessed generally, but specific note should be made of anterior extension and possible involvement of the vertebral or basilar arteries. The superior limit of the mass should be defined, and the amount of distortion of the tentorium assessed. The inferior extent should be carefully appreciated, and possible involvement of cranial nerves IX through XII should be noted. All of this information, considered in the light of preoperative hearing levels and pre-existing neurologic deficits, is used to plan the surgical approach and provide detailed preoperative counseling.

If postoperative swallowing difficulties due to involvement of cranial nerves IX through XII are anticipated, or if facial nerve exercises are to be used postoperatively, then preoperative evaluation by the paramedical personnel involved should be considered. Preoperative evaluation by an internist is essential, as it may forestall future problems and will familiarize the internist with the patient should postoperative consultation be required.

It is appropriate to observe certain patients rather than operate upon them. Principally, these will be elderly individuals or those with serous medical problems. If observation is elected, however, close follow-up is essential, with repeated radiographic evaluation every 6 months until the rate of tumor growth has been well established. We have seen a number of patients whose tumors doubled in size over a six-month period. Other individuals for whom observation may be most appropriate are those with either bilateral tumors or tumors in an only-hearing ear. In such cases, hearing levels and the ABR should be carefully and frequently reassessed, along with repeated radiographs.

Surgical Procedures

Middle Fossa Approach

The middle fossa approach is used for small tumors when hearing conservation is attempted. It is useful when the tumor extends 2.5 cm or less out of the porus acusticus. Access to the posterior fossa, even after division of the superior petrosal sinus at the tentorial margin, is insufficient to remove larger tumors. Patients with marked expansion of the internal auditory canal are less likely to have a good hearing result but do not present increased technical difficulty. The principal disadvantage of this procedure is increased risk to cranial nerve VII. The facial nerve will be found lying over both the tumor mass and the cochlear nerve. The tumor must therefore be removed from beneath cranial nerve VII, increasing the risk of injury. The procedure is also considerably more difficult in patients over 50 years of age. Elevation of dura from the underlying bone in elderly patients produces marked bleeding that is quite difficult to control.

Technique

The procedure is performed with the patient in a supine lateral position. A microphone is placed in the external auditory canal and electrodes are placed into the scalp and ear lobe for continuous monitoring of the brain stem response audiogram. Fully half the head is shaved,

prepped, and draped. The surgeon performs the procedure seated directly above the patient's head. The zygomatic process is carefully marked with a skin marking pen. The incision must not be carried below the zygomatic arch or damage to the frontozygomatic portions of the facial nerve will occur. The vertical incision at right angles to the zygomatic arch is drawn 1 cm anterior to the root of the zygoma as determined by palpation. The incision is extended directly cephalad to the vertex of the skull. The incision must be straight. If it angles posteriorly or anteriorly even a few degrees, exposure will not be adequate.

It is paramount that the incision be meticulously planned as exposure will be inadequate if the incision is even 1 to 2 cm off. The planned incision is injected with Xylocaine with epinephrine to limit bleeding. It is incised sharply down to the temporalis fascia. The temporalis fascia itself is next incised sharply as well. The electrocautery should be avoided on the fascia because it induces contraction of the tissue, making closure more difficult. Care should be exercised in preserving the temporalis fascia, as accurate fascia closure will significantly diminish postoperative cerebrospinal fluid leak, edema, and hematoma formation. The temporalis muscle is now incised down to the pericranium. A portion of the muscle is removed and set aside for later use as a plug for the internal auditory canal (IAC). The pericranium is incised, and the pericranium and temporalis muscle are elevated from the underlying skull.

A middle fossa craniotomy is performed by removing a portion of the temporal squamosa. The bone flap should be placed exactly 1 cm behind the root of the zygoma, which can now be clearly identified. The anterior margin should be placed 2 cm anterior to the root of the zygoma. The superior limit should be placed either 3 cm above the zygomatic arch or at the level of the squamoparietal suture - whichever is greater. The inferior margin of the bone flap must be at the level of the zygomatic arch itself. The bone flap is outlined with the surgical bur and removed from the underlying dura with a blunt periosteal elevator. The dura is elevated from the underlying superior surface of the temporal bone. Pesky bleeding may be quite profuse, but time should be taken to create a completely dry field. Elevation is carried anteriorly to the foramen spinosum and middle meningeal artery, posteriorly just past the arcuate eminence and medially to the tentorial margin of the petrous apex. After dural elevation is complete, the House-Urban retractor is positioned into the wound.

The diamond bur is now used to expose the internal auditory canal. First the air-cells over the middle ear are opened until the head of the malleus can be identified. The facial nerve can be seen in its tympanic course at its junction with the geniculate ganglion. The arcuate eminence and greater superficial petrosal nerve should now be identified. By noting these three landmarks, the labyrinthine portion of the facial nerve can be identified and bone removed from over it. Tolerances are minimal here, as often only a few millimeters separates the posterior cochlea from the anterior portion of the superior semicircular canal.

Next, the superior semicircular canal is "blue-lined" so that the posterior medial exposure of the IAC can be performed. Maximal exposure here is crucial and requires working as close to the superior semicircular canal (SSC) as possible. With the key landmarks identified bone can be removed from the length of the IAC. A full 180 degrees of exposure is essential. The dura

over the IAC must remain intact during bone removal, as the facial nerve lies immediately below the dura and is often both thinned and pressed against the dura by the underlying bone.

After bony removal is complete, the dura is opened posteriorly as far from the anticipated position of the facial nerve as possible. The facial nerve should now be identified and separated from the underlying tumor. If the tumor is small, it should be separated from the underlying cochlear nerve as gently as possible. If, at any point during the procedure, manipulation of the tumor or nerves produces marked alteration of the ABR audiogram, then manipulation is immediately stopped. Dissection should hug the tumor capsule as closely as is feasible in order to avoid inadvertent disruption of blood supply to the cochlea. If the tumor is large, debulking using the CO_2 laser is performed in order to allow tumor removal with less manipulation of the underlying nerves and vessels. It is paramount that the contents of the IAC be disturbed as little as possible. It must be remembered that retention of an intact cochlear nerve is a necessary but not sufficient condition for hearing preservation. The vascular supply of the membranous labyrinth must also be preserved. Superior and inferior vestibular nerves are sacrificed and removed with the tumor. If the tumor extends medial to the tentorial margin the superior petrosal sinus may be divided between clips, the tentorial margin incised, and limited exposure to the posterior fossa achieved.

After meticulous hemostasis had been obtained, the IAC is covered with the previously harvested temporalis muscle and the House-Urban retraction is removed. The temporal lobe is now allowed to re-expand. The previously removed bone flap is sutured back into position, using 3-0 silk sutures through small holes drilled into the bone plate and inferior temporal squama. The temporalis muscle is then closed over the bone flap, followed by careful closure of the temporalis fascia and skin. A large pressure dressing is applied to limit postoperative edema and hematoma formation. The dressing is left in place for 5 days.

Results

Tumor removal should be complete in all cases. Damage to the facial nerve is uncommon but is more frequent than for tumors of similar size removed by the translabyrinthine approach. Despite anatomic preservation of the cochlear nerve, useful postoperative hearing will be retained in only 50 per cent of patients. This disappointing result is no doubt a consequence of inadvertent disruption of the blood supply to the membranous labyrinth.

Wound infection will occur episodically and may be treated by wound opening, local care, and appropriate antibiotics. The antibiotics should be selected from the results of wound cultures. Occasionally, hematomas will form and will require evacuation. Cerebrospinal fluid leaks will occur in 5 to 10 per cent and will usually require re-operation if leakage is through the nose. If leakage is through the wound, a short period of treatment using reinforcing skin sutures and pressure dressings may be undertaken if the patient is closely observed for early signs of meningitis. The smallest change in mental status, postoperative fever, or increasing headaches requires prompt lumbar puncture in any postcraniotomy patient. Daily lumbar puncture or continuous spinal drainage should be performed if the cerebrospinal fluid leak is managed

nonoperatively.

The Suboccipital Approach

The suboccipital approach should be considered in those patients who are candidates for hearing conservation when the tumor is 1.5 cm or greater in size. In general, these will be patients whose pure-tone averages are better than 50 dB and whose discrimination scores are better than 50 per cent in the affected ear. Occasional patients with hearing levels worse than this may be considered for hearing conservation procedures, if the contralateral ear is seriously diseased. Tight wedging of the tumor into the lateral end of the IAC and marked expansion of the IAC are relative contraindications, because the changes of salvaging useful hearing are significantly reduced in these circumstances.

Technique

The procedure is performed with the patient in the supine position and the surgeon seated to the side of the patient's head, as in the translabyrinthine approach. A 4- to 5-cm postauricular flap is elevated. A simple mastoidectomy is performed, with care being taken to leave the bony labyrinth intact. The facial recess is not opened, and the ossicles are not disturbed. The sigmoid sinus is identified and skeletonized, including 1 to 2 cm of the distal lateral sinus. Three to five centimeters of bone are now removed from the occipital bone behind the sigmoid sinus. Bone removal should be carried as far inferiorly as possible. An area of dura about 4 by 4 cm posterior to the sigmoid sinus will have not been exposed. A cruciate incision is made in the dura and carried to the margins of the craniotomy defect. The dural flaps are reflected and held with dural hooks. The arachnoid around the lateral cerebellar hemisphere is carefully dissected in an anterior direction, and the lateral cerebellar cistern is entered.

As cerebrospinal fluid is aspirated from the cistern, exposure of the cerebellopontine angle will begin and the tumor can be visualized. A self-retaining retractor can now be inserted, and the cerebellar hemisphere gently retracted medially until the medial margin of the tumor and/or brain stem can be seen. Large tumors may require resection of the lateral third of the cerebellar hemisphere in order to develop adequate exposure. Removal of cerebellar tissue is preferable to forceful retraction, which could interrupt vascular flow.

Once the tumor has been well exposed, debulking using the CO_2 laser may begin. Debulking is continued until the root entry zones of cranial nerves VII and VIII can be seen. The tumor capsule is then removed from the medial portions of the facial and statoacoustic nerves. The posterior wall of the IAC is removed. As dissection of the IAC proceeds laterally from the porus acusticus, great care must be taken to avoid inadvertent penetration of the vestibule and posterior semicircular canals. It is usually not possible to fully expose the lateral-most portion of the IAC. As dissection of the tumor capsule proceeds laterally from the brain stem, the cochlear nerve moves from an inferior position to an anterior position within the IAC, and the vestibular nerves move from superior to posterior.

The vestibular nerve, as the nerve of origin of the tumor, will be sacrificed. Great care must be taken during the procedure to interfere as little as possible with the blood supply to the cochlea. Hemostasis must be meticulous. The cruciate dural incision is closed with 4-0 silk sutures and the exposed mastoid air-cells "waxed" off. The postauricular flap is closed in a routine fashion.

Results

Reports of success in conserving hearing vary rather broadly but about one-third of patients retain useful hearing postoperatively. Central nervous system complications related to cerebellar edema or compromise of blood flow to the brain stem occur occasionally. Review of the literature shows salvage of the facial nerve to be somewhat less with the suboccipital approach compared to the translabyrinthine approach. Postoperative convalescence is prolonged with the suboccipital approach.

Translabyrinthine Approach

The translabyrinthine approach to the cerebellopontine angle is the one most frequently employed by the authors. It is the most direct approach to the cerebellopontine angle and has to date yielded the highest salvage rate of the facial nerve. It provides the best exposure of the lateral end of the IAC which allows the surgeon to remove all tumor with great confidence. Complete exposure of the lateral IAC also allows identification of the facial nerve in its labyrinthine segment where it is least distorted by tumor.

Technique

The procedure is performed with the patient in the supine lateral position. The abdomen is prepped and draped, and about 30 cubic centimeters of fat are removed and set aside for the use in wound closure. The fat is kept in an antibiotic solution until ready for use.

A postauricular flap extending 3 to 4 cm posterior to the postauricular fold is designated and injected with 1 per cent Xylocaine with epinephrine, 1:100.000. The flap is incised down to include the periosteum and then elevated until the posterior wall of the external auditory canal is reached. Care must be taken to avoid inadvertent elevation or laceration of the skin of the external auditory canal. A complete, simple mastoidectomy is now performed. The mastoidectomy should be well saucerized and carried posteriorly just past the posterior limit of the sigmoid sinus. Through a standard facial recess approach, the incudostapedial joint is identified and the incus is removed. The incus is preserved in 70 per cent ethyl alcohol and kept in the bone bank for later use as a homograft in ossicular reconstruction.

When the mesotympanic orifice of the eustachian tube is clearly visualized, 3 to 4 pieces of sterilized Proplast (3 to 4 mm long and about 1 mm in diameter) are inserted into it in order to occlude it. The eustachian tube should be filled with care taken to avoid pushing pieces through the eustachian tube into the nasopharynx. Proplast should not be allowed to protrude into

the middle ear, in order to prevent eventual extrusion through the tympanic membrane. The use of Proplast has all but eliminated postoperative cerebrospinal fluid rhinorrhea.

The facial nerve should now be identified from the geniculate ganglion to the stylomastoid foramen. A thin covering of bone is left over the nerve. Dissection is continued inferiorly, following the sigmoid sinus into the retrofacial air-cells until the jugular bulb is encountered. The semicircular canals should be well defined. The bone over the middle fossa dura and sigmoid sinus is removed. The bipolar cautery may be applied to the exposed dura and will cause shrinkage and increase exposure. A large, protruding, anteriorly placed sigmoid sinus may thereby be significantly reduced in size and render an almost impossible situation very manageable.

Prior to beginning the labyrinthectomy, the patient's identification bracelet, chart, audiogram, and CT scan are all checked to ensure that the correct side is being operated on. The labyrinthectomy is carried out in a systematic fashion.

The horizontal canal is first opened and its lateral portion removed. The posterior and superior canals are opened next and followed medially to the crus communis. The crus communis is then followed into the vestibule. Caution must be exercised not to remove the macula cribrosa superior of the superior semicircular canal, where nerve roots of the superior vestibular nerve enter the membranous labyrinth through tiny bony fenestra to enervate the macula of the superior semicircular canal. This is a crucial landmark. After widely opening the vestibule, all but a thin sheet of bone should be removed from the tympanic facial nerve inferiorly and posteriorly, to provide wide access to the lateral portion of the IAC. The posterior fossa dura should be exposed anterior to the sigmoid sinus until the dura that invests the contents of the IAC is encountered. Bone from the middle fossa dura should be removed until the superior IAC is exposed. All of the bone over the jugular bulb should be removed to expose the inferior IAC. It is paramount that at least a full 180 degrees of the IAC be exposed.

A high jugular bulb can usually be shrunken with bipolar cautery or compressed with cottonoid pledgets to increase exposure. It is important that flow through the sigmoid sinus and jugular bulb be maintained. Occlusion of a dominant sigmoid sinus may result in severe postoperative cerebral edema or cortical venous infarction. Inadvertent bleeding from the sigmoid sinus or jugular bulb can be controlled by patching the hole with a piece of Gelfoam and maintaining flow within the sinus. A wet piece of Gelfoam is placed over the laceration and immediately covered with a cottonoid pledget, which holds it in place. Continuous suction on the pledget is maintained with a large Frazier suction tube for 3 to 4 minutes until bleeding ceases. The pledget can be removed and the Gelfoam left as a semipermanent "patch". The bone over the IAC itself should now be rendered paper-thin and removed with a small right-angle pick. Attention is now turned to the macula cribrosa superior, and the remaining bone is removed.

The terminal ends of the superior vestibular nerve are seen and help to identify it. Using a blunt right-angle hook Bill's bar is palpated and the hook is passed anterior to it into the labyrinthine portion of the facial canal. Thus, the facial nerve is definitively identified in its most constant anatomic relationship, undistorted by tumor growth. The dura of the IAC is now opened

inferiorly and removed from the tumor.

It is useful at this point, prior to beginning debulking of the tumor in the cerebellopontine angle, to establish whether the facial nerve has been displaced anteriorly, as is usual, or posteriorly or superiorly by the tumor mass. The posterior fossa dura is then opened anterior to the sigmoid sinus. Care is taken not to injure the underlying vasculature. The CO_2 laser is used to debulk the tumor mass, which is isolated from the brain stem by gently lifting the capsule away from the brain stem and placing cottonoid pledgets between the tumor and the brain. Laser debulking should proceed cautiously so as not to inadvertently penetrate the capsule from the inside out, thereby accidentally damaging the facial nerve or other important neural or vascular structures.

As separation from the brain stem proceeds, the root entry zone of cranial nerves VII and VIII is identified, and cranial nerve VIII is sacrificed. If there is difficulty in determining which nerve is which, an intraoperative nerve stimulator may be helpful. The tumor capsule is separated from the facial nerve, often a difficult task. The nerve will be markedly "splayed out" and will frequently be five to six times its normal width and one- to two-tenths of its normal thickness. From time to time, it simply will not be possible to remove all the tumor and leave the nerve intact. In those cases, the nerve should be sacrificed in order to ensure complete tumor removal. Immediate reconstruction, either by primary anastomosis achieved after re-routing the nerve to gain length, or by cable grafting, should be undertaken. In cases in which nerve repair is not possible, hypoglossal-facial nerve anastomosis should be performed 7 to 10 days postoperatively.

When tumor removal is complete, the wound is irrigated, and meticulous hemostasis is achieved. The patient's posterior fossa should be carefully observed for bleeding during and after Valsalva maneuvers performed by the anesthetist. With hemostasis assured, a small piece of muscle is taken from the exposed temporalis muscle and is used to obliterate the middle ear space. The previously taken fat is cut into strips and 0.5 cm to 1.0 cm of the end of one of the strips is placed into the cerebellopontine angle. The remaining strips are used to obliterate the mastoidectomy cavity and seal the wound. The wound is closed in three layers. Meticulous fascial closure is essential in minimizing postoperative cerebrospinal fluid leaks. Subcutaneous closure is followed by skin closure with surgical staples. A standard mastoidectomy dressing is then applied. The patient is observed in the recovery room overnight with hourly neurologic checks and is transferred to the floor the next morning.

Results

Tumor removal should be complete. Very rarely, in exceptional circumstances, a small amount of tumor is deliberately left attached to the facial nerve in order to spare it. Mortality is less than 1 per cent. The facial nerve can be anatomically preserved in most cases, but preservability is clearly a function of tumor size. In 5 to 10 per cent of patients there will be a cerebrospinal fluid leak from the postauricular wound and 1 to 2 per cent of patients will develop a postoperative meningitis secondary to that cerebrospinal fluid leak. Also, 1 to 2 per cent of patients, usually patients with tumors larger than 3.5 cm, will have postoperative hydrocephalus.

In many cases it is temporary and can be managed with prolonged steroid therapy, but some patients will require ventricular peritoneal shunting.

The majority of patients will have postoperative facial nerve paralysis, but almost all will have complete or nearly complete recovery. Eye care in those patients with nonfunctioning nerves must be aggressive. Instillation of artificial tears every 1 to 2 hours, the use of ophthalmic ointment each night, and application of a protective eye "bubble" during sleep are routine. The liberal, early use of temporary lateral tarsorrhaphy is very helpful. The effectiveness of "facial nerve" exercises is questionable but not harmful, and many patients are enthusiastic about using them.

Combined Retrosigmoid Translabyrinthine Approach

The combined approach is useful in excising large tumors (usually greater than 3.5 cm) and those with marked posterior extension when the translabyrinthine approach provides insufficient exposure to the posterior fossa. This approach affords wide exposure of the posterior fossa and lateral IAC. Complete tumor removal can be accomplished in a single stage with a single incision. With good access to the lateral IAC and Bill's bar, chances of preserving the facial nerve are maximized.

Technique

A large postauricular flap is designed, often extending 6 to 8 cm posterior to the postauricular fold. The flap is elevated and the translabyrinthine dissection is completed. Bone is removed 3 to 4 cm posterior to the sigmoid sinus. The facial nerve is identified in the lateral portion of the IAC. A U-shaped flap is made in the retrosigmoid posterior fossa dura. It is based posteriorly.

The CO_2 laser is used to debulk the tumor and, as debulking proceeds, the tumor is separated from the cerebellum using moist cottonoid pledgets. Dissection and debulking are continued in this manner until the tumor capsule can be delivered anteriorly under the sigmoid sinus. Dissection is completed as in the translabyrinthine approach. Closure is the same, with the addition of water-tight closure of the retrosigmoid dura flap.

Displacement and distortion of significant vascular and neural structures is considerably more likely with the large tumors that require the combined approach. A very intimate knowledge of the anatomy of the posterior fossa is essential to avoid damaging the anterior inferior cerebellar artery or other major vessels. The usual price exacted for disruption of these vessels is the patient's demise. Involvement of cranial nerves IX through XII inferiorly or cranial nerves III through VI superiorly is common and every effort should be expended to preserve them.

Results

Tumor removal should be complete. Mortality is about 1 per cent but postoperative morbidity is increased. Recovery may be slow, and postoperative hydrocephalus may require placement of a ventricular peritoneal shunt in the postoperative period if this was not accomplished preoperatively. About 5 to 10 per cent of patients will have transient dysfunction of cranial nerves other than VII and VIII.

Long-term results are excellent and parallel those of removal of smaller tumors, except in the area of the facial nerve. Preservability of the facial nerve drops dramatically as tumor size exceeds 4.0 cm. Barely 50 per cent of the nerves can be spared in these large tumors. Fortunately, immediate repair or early VII-XII anastomosis has provided satisfactory recovery for the majority of patients whose nerves have been sacrificed.

Summary

Early diagnosis of acoustic neuroma results in decreased morbidity and mortality. Early symptomatology may be subtle and easily escape detection. Therefore, all unilateral otologic symptoms require evaluation for acoustic neuroma. Four approaches to tumor removal are available to the team managing these patients. Rigid commitment to a single approach denies the patient the opportunity to have therapy individualized to meet his or her unique needs and circumstances.