Sensorineural Hearing Loss and the Diagnosis of Acoustic Neuroma

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Sensorineural hearing loss and tinnitus are gradually receiving more attention in medicine due to advances in diagnosis and treatment. Acoustic neuromas can now be detected when they are small, and early microsurgical removal results in the lowest overall morbidity. We examine the historical development of acoustic neuroma management, discuss current diagnosis and treatment, and present illustrative cases from our recent experience. Complaints of tinnitus and hearing loss, especially when unilateral, require appropriate medical evaluation. (Henry Ford Hosp Med J 1990;38:9-12)

In the past hearing loss and tinnitus were often regarded as minor nuisances not meriting medical attention. This neglectful attitude originated from the once common view that hearing loss was a hopeless or comical condition and was indicative of society’s prejudice against the aged and the hearing impaired. Moreover, little could be done for hearing-impaired individuals until the introduction of Lempert’s one-stage fenestration procedure just prior to World War II and the development and refinement of stapes surgery, tympanoplasty, and hearing aids in the first two decades after the war. Today hearing loss, like the loss of any sensory function, is more widely recognized as a medical symptom which requires evaluation and which does not in itself constitute a diagnosis.

Historical Perspective on Tumor Treatment

At the turn of the century, intracranial surgery uniformly resulted in fatality. Patients who did not die from hemorrhage succumbed to sepsis. Despite this grim prognosis, surgical treatment of acoustic neuromas was sometimes attempted when patients became bedridden by severe cerebellar ataxia, lapsing in and out of obtundation from brain stem compression and intracranial hypertension. If a patient presented with hearing loss, headache, and papilledema, the diagnosis of intracranial tumor was considered to be established. If the patient were also ataxic, the tumor was presumed to be in the posterior cranial fossa (1).

Boston neurosurgeon Harvey Cushing made significant early contributions to the diagnosis and treatment of acoustic neuromas. Cushing’s first successful treatment consisted of removing the suboccipital bone bilaterally, allowing for expansion of the intracranial contents. This procedure allowed most patients to become ambulatory and prolonged their life by a few months. Later, Cushing succeeded with the subtotal removal of tumors. Operative mortality was reduced to 25%, a great achievement at that time. Cushing’s careful accumulation of patient histories and physical findings led to the first detailed account of the clinical course and natural history of acoustic tumors. He described several surgical approaches to the cerebellopontine angle that are still in use today (2,3).

Cushing’s pupil, Walter Dandy, not as keen a clinical observer as Cushing though he had remarkable technical skill, was the first to carry out complete removal of tumors consistently. His lifetime operative mortality was 22.1%, and in one remarkable series toward the end of his career, operative mortality was 2.4% (4).

In 1949 the importance of the anterior inferior cerebellar artery as a vital structure was recognized (5). Sparing this structure reduced operative mortality further.

In 1964 William House, an otologist working with neurosurgeon William Hittsberger, published his series of 53 tumors removed through a transtemporal bone microsurgical approach, called the translabyrinthine approach (6). House and coworkers have since removed more than 2,700 tumors with operative mortality below 0.5%. Moreover, facial nerve function was preserved in the majority of cases and other neurologic deficits were rare (6,7).

These results were achieved because the facial nerve can be identified lateral to the tumor with the surgical microscope using secure bony landmarks. The nerve can then be followed medially to the tumor while the plane of separation between nerve and tumor is maintained. Manipulation of the brain stem and cerebellum is minimized, resulting in fewer intracranial complications. Patients are operated upon in the supine position, which avoids the rare complication of quadriplegia and the more common, potentially fatal, venous air embolism that can occur when the patient is operated upon in a sitting position (8).

Other surgical approaches continue to be useful, including the middle fossa approach described by House, which permits the removal of small tumors and preserves hearing. The combined approach, described by Cushing and popularized in the contemporary era by Glasscock (8), combines the advantages of the translabyrinthine and suboccipital approaches. Surgical ap-

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proaches and techniques continue to evolve. Most centers offer a team approach combining the skills of an otologist, neurosurgeon, neuroanesthesiologist, and neurophysiologist. The use of perioperative antibiotics, continuous arterial pressure monitoring, pulse oximetry, neuroanesthesia, intensive care units, and intraoperative facial nerve monitoring have improved the outcome of acoustic tumor surgery. Current operative mortality is in the range of 0.5% in most series.

Tumor Diagnosis

Improvements in surgical techniques have been matched by improvements in diagnostic techniques. Audiology has developed rapidly as a clinical discipline since the 1940s. It is now possible to measure hearing pure tone air and bone conduction thresholds, speech reception thresholds, and speech discrimination. The introduction of the measurement of acoustic impedance has added the ability to determine the threshold and decay properties of the acoustic reflex (9). Auditory brain stem response (ABR) testing determines the integrity of the auditory nerve. The ABR is an auditory-evoked response and the most cost-effective screening test available for acoustic neuroma and other disorders of the auditory nerve (10). Its main limitation is its dependence on intact hearing. Test subjects must have hearing at the 60 to 70 dB level or better in the high frequency range of 3,000 to 4,000 Hz; otherwise the recording may be uninterpretable. The sensitivity of the test depends on the magnitude of the normal range, which is defined statistically and is generally in the range of 98%. For this reason, ABR is best used in the context of the full neurotologic evaluation.

Imaging techniques were once limited to skull radiography that showed the outline of the internal auditory canals (IACs). The IAC is usually widened in medium to large acoustic neuromas. Complex motion tomography combined with the introduction of radiographic contrast material into the subarachnoid space allowed tumor detection and estimation of tumor size and boundaries. Computed tomography (CT) with intravenous radiographic contrast and air contrast in the cerebellopontine angle represented an advance that made early routine detection of even small tumors possible for the first time. Magnetic resonance imaging (MRI) with gadolinium contrast has become the “gold standard” diagnostic test for acoustic neuroma. This method is able to detect tumors as small as 2 to 3 mm and is the most accurate in determining tumor boundaries at any size. The history of acoustic neuroma diagnosis and the history of treatment are linked; as treatment has improved, the imperative for early diagnosis has grown.

Clinical Characteristics

Histologically, acoustic neuromas are benign tumors of the Schwann cell sheath of the eighth cranial nerve. They grow slowly but continuously and, if untreated, are eventually fatal in most cases. Regardless of the surgical technique used, risks and complications are fewer if tumors are removed when small. Now that early tumor detection is possible and the advantages of early diagnosis are well appreciated, there is a compelling need to pursue this diagnosis.

Cushing showed that the earliest sign of acoustic neuroma is hearing loss. Today asymmetric hearing loss is still recognized as the most prevalent sign on presentation, although patients may present with other symptoms (11).

Hearing Loss

Sensorineural hearing loss is prevalent in approximately 10% of the population, with the risk increasing to 20% by age 65 years. While the hearing loss rate of 2 to 3 dB/year after age 65 years may be a part of the normal aging process (presbycusis), an asymmetric sensorineural hearing loss at any age should be considered the result of an acoustic neuroma until proved otherwise. Moreover, tumors may present with dysequilibrium, numbness of the face, aural fullness, tinnitus, or aural pain. They may be detected as incidental findings in asymptomatic patients. The incidence of acoustic neuroma has been estimated to be 0.8 to 1.0 per 100,000 population per year.

Case Presentations

Case 1

This healthy 16-year-old boy was sitting on his front porch when he noticed a ringing sound in his left ear and suddenly lost all hearing in the ear. He had no vestibular symptoms. He sought medical attention and was referred to an otolaryngologist. Physical examination revealed the ears, head, and neck to be normal. Audiometric evaluation revealed a profound sensorineural hearing loss on the left. CT demonstrated a soft tissue mass filling the internal auditory canal and extending 1 cm medial to the porous acusticus. He was referred to Henry Ford Hospital.

A 1 cm tumor was removed via the translabyrinthine route in a combined neurotologic and neurosurgical procedure. Facial nerve function was normal postoperatively. Histopathologic examination of the tumor specimen established the diagnosis of schwannoma (acoustic neuroma). An organized hematoma was found in the tumor specimen. The patient’s recovery was uneventful.

Comments

This case illustrates the occurrence of acoustic neuroma presenting as sudden sensorineural hearing loss. The presence of an old hematoma suggests that the sudden loss of hearing may have been caused by sudden bleeding into the tumor. It has been established that acoustic neuroma may present as sudden hearing loss and that hearing may recover even in the presence of a tumor (12).

Case 2

This 38-year-old woman presented to her otolaryngologist with a three-year history of intermittent mild dysequilibrium and a nine-month history of mild left hearing loss. Physical examination was unremarkable. Audiometric evaluation revealed mild left sensorineural hearing loss and normal acoustic reflexes. An auditory-evoked potential study revealed abnormality on the left side. CT showed mild widening of the left internal auditory canal. A faint shadow suggested the possibility of a mass in the cerebellopontine angle. The patient was referred to Henry Ford Hospital.

MRI with gadolinium enhancement demonstrated an enhancing mass extending from the fundus of the internal auditory canal to the brain stem. The tumor was removed via the translabyrinthine route in a combined neurotologic and neurosurgical procedure. Facial nerve function was normal postoperatively. Histopathologic examination confirmed the diagnosis of acoustic neuroma. The patient’s recovery was uneventful.
Comments
This case illustrates the presentation of acoustic neuroma with mild hearing loss. Hearing preservation was not attempted because the tumor filled the internal auditory canal and extended to its fundus. CT only suggested the presence of a tumor, which was confirmed by MRI with gadolinium enhancement.

Case 3
This 26-year-old man presented with a one-year history of fluctuating hearing loss and aural fullness without vestibular symptoms. Audiometric evaluation showed a mild right sensorineural hearing loss. Subsequent evaluations demonstrated normal hearing. MRI with gadolinium enhancement showed a tumor in the internal auditory canal, extending 0.5 cm toward the brain stem. The acoustic neuroma was removed through the suboccipital approach with electrophysiologic monitoring of both auditory and facial nerves. Intraoperatively, the tumor was noted to be tightly adherent to the auditory nerve without a clear plane of separation. Postoperative hearing remained normal through 1,000 Hz but dropped precipitously at higher frequencies (Figure). Postoperative speech discrimination was 60% and facial nerve function was normal.

Comments
This case illustrates that mild hearing loss usually associated with relatively small tumors may fluctuate and could even present as normal. Even a history of hearing loss should be pursued, despite initially negative findings. Hearing preservation is possible in some cases.

Case 4
This 60-year-old woman was followed for several years because of hearing loss and episodic vertigo which were thought to be caused by Meniere’s disease. She sought a second opinion at Henry Ford Hospital. Audiometric evaluation showed severe bilateral sensorineural hearing loss, with hearing better on the right side. CT showed a 3 cm tumor in the right cerebellopontine angle. The acoustic neuroma was removed through the translabyrinthine approach. Postoperatively facial nerve function was normal, and the patient’s recovery was uneventful. She was referred for auditory rehabilitation.

Comments
This case illustrates that acoustic neuromas can mimic classical Meniere’s disease. Whereas this patient had episodic vertigo of the Meniere type, case 3 showed fluctuating hearing in the manner of early Meniere’s disease. Acoustic neuroma should be excluded in patients with episodic vertigo, tinnitus, and hearing loss before that clinical syndrome is accepted as diagnostic of Meniere’s disease.

Discussion
We are in a new era in tumor management and in the management of sensorineural hearing loss. Tumors can be diagnosed when small, and operative complications are fewer if surgery is performed when tumors are small (13). Tumors present with common, seemingly minor symptoms, such as sensorineural hearing loss. Whereas tumors are rare, sensorineural hearing loss is common.

While it might be desirable for everyone with mild ear symptoms to undergo MRI, health care resources are limited and scans are expensive. Unfortunately, we have no uniformly accepted standard method to detect the rare acoustic tumors among the common cases of sensorineural hearing loss. Development of such a standard would need to take into account a monetary estimate of the “cost” of the morbidity added by delay in diagnosis. The costs of tests vary between institutions as does the rate of surgical complications. Tumor growth rates are unpredictably variable across individuals and over time for the same individual.
We recommend that patients be screened with air, bone, speech, and impedance audiometry. Patients with asymmetric hearing, asymmetric speech discrimination, or afferent limb acoustic reflex abnormalities should be evaluated further (9,14). We believe that the possibility of acoustic neuroma should be considered in all patients with asymmetric sensorineural hearing loss or with other persistent unilateral or asymmetric ear symptoms. A less aggressive approach can be taken in asymptomatic patients who are permanently debilitated or over age 65 years.

References


