ACOUSTIC NEUROMA - BEWARE OF UNDUE USE OF CELL PHONES

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Summary

The acoustic neuroma is charactersied by include ipsilateral sensorineural hearing loss/deafness, disturbed sense of balance and altered gait, vertigo with associated nausea and vomiting, and pressure in the ear,along with tinnitus all of which can be attributed to the disruption of normal vestibulocochlear nerve function. Due to continous exposure of cellular phones causes acoustics approximately 3,000 cases are diagnosed each year in the United States with a prevalence of about 1 in 100,000 worldwide. This review disscussed about incidence,causes, Pathophysiology, clinical manifestation and different kinds of treatments of acoustic neuroma. Further this review suggesting that declination of execessive use of cellular phones and huge exposure of high frequency sounds.

Key words *:* Acoustic neuroma, Schwannoma, Cellular phones, Pathophysiology, Clinical manifestations, treatment.

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Introduction

The term "acoustic" is a misnomer, as the tumor rarely arises from the acoustic (or cochlear) division of the vestibulocochlear nerve. The term "neuroma" is also a misnomer, since it means "nerve tumor" but an acoustic neuroma is a Schwannoma. The correct medical term is vestibular schwannoma, because it involves the vestibular portion of the 8th cranial nerve [1] and it arises from Schwann cells, which are responsible for the myelin sheath in the peripheral nervous system. Approximately 3,000 cases are diagnosed each year in the United States with a prevalence of about 1 in 100,000 worldwide. It comprises 5-10% of all intracranial neoplasms in adults. Incidence peaks in the fifth and sixth decades and both sexes are affected equally.[2]

Incidence of Acoustic Neuromas

The incidence of acoustic neuromas is about one per 100,000 people. According to the National Institutes of Health, an estimated 2,000 to 3,000 new cases of acoustic neuromas are diagnosed in the United States each year [3]. Reported incidence may be rising, however, due to improved imaging technology that enables smaller tumors to be clearly visualized**.** Because of the slow growth rate of acoustic neuromas, a large number of cases never become clinically evident so that the actual number of cases may be much higher. Acoustic neuromas account for approximately:

- 6% of all intracranial tumors (tumors that occur in the brain)
- 30% of all brainstem tumors
- 85% of tumors in the cerebellopontine angle region

Although they can occur as early as age 7, most people with acoustic neuromas are diagnosed when they are between the ages of 30 and 60. The median age of diagnosis is 50. The incidence of acoustic neuroma is slightly higher among women (60%) than men (40%). Acoustic neuromas occur on one side (unilateral) in approximately 95% of patients unless they are associated with *neurofibromatosis-2*, in which case they are typically bilateral (occur on both sides). The overwhelming majority of acoustic neuromas are due to unknown causes. These tumors typically present later in life and are almost always unilateral. Acoustic neuromas can also be associated with a genetic condition called *neurofibromatosis-2*. [4]

Causes Acoustic Neuroma

Sporadic form

Acoustic neuroma occurs in two forms: a sporadic form and a form associated with an inherited syndrome. About 95% of all cases are sporadic. The cause of the sporadic form is unclear. Some small studies have found an association of acoustic neuromas with cellular phone use or prolonged exposure to loud noises, but other studies do not find this link (Christensen et al, 2004; Edwards et al, 2006; Edwards, 2007; Hardell, 2003; Lonn et al, 2004; Schoemaker et al, 2005; Schlehofer, 2007). There is not hard evidence supporting a link between environmental factors and acoustic neuromas.

Inherited syndrome (Neurofibromatosis type II)

The inherited syndrome called neurofibromatosis type II (NF2). NF2 is rare; there are only several thousand affected individuals in the entire United States, corresponding to about 1 in 40,000 individuals. Roughly 5% of patients with acoustic neuroma have NF2.[5,6]

Pathogenesis

Acoustic neuromas may occur sporadically, or in some cases occur as part of von Recklinhausen neurofibromatosis, in which case the neuroma may take on one of two forms.

- In ***Neurofibromatosis type I***, a schwannoma may sporadically involve the 8th nerve, usually in adult life, but may involve any other cranial nerve or the spinal root. Bilateral acoustic neuromas are rare in this type. [7]
- In ***Neurofibromatosis type II***, bilateral acoustic neuromas are the hallmark and typically present before the age of 21. These tumors tend to involve the entire extent of the nerve and show a strong autosomal dominant inheritance. Incidence is about 5 to 10%.

The usual tumor in the adult presents as a solitary tumor, originating in the nerve. It usually arises from the vestibular portion of the 8th nerve, just within the internal auditory canal. As the tumor grows, it usually extends into the posterior fossa to occupy the angle between the cerebellum and the pons (cerebellopontine angle). Because of its position, it may also compress the 5th, 7th, and less often, the 9th and 10th cranial nerves. Later, it may compress the pons and lateral medulla, causing obstruction of the cerebrospinal fluid and increased intracranial pressure. Schwannomas can occur in relation to other cranial nerves or spinal nerve roots, resulting in radiculopathy or spinal cord compression. Trigeminal neuromas are the second most common form of schwannomas involving cranial nerves. Schwannomas of other cranial nerves are very rare.

Pathphysiology

The vast majority of acoustic neuromas develop from the Schwann cell investment of the vestibular portion of the vestibulocochlear nerve. Less than 5% arise from the cochlear nerve. The superior and inferior vestibular nerves appear to be the nerves of origin with about equal frequency. Overall, 3 separate growth patterns can be distinguished within acoustic tumors, as follows: (1) no growth or very slow growth, (2) slow growth (ie, 0.2 cm/y on imaging studies), and (3) fast growth (ie, $\geq$1.0 cm/y on imaging studies). Although most acoustic neuromas grow slowly, some grow quite quickly and can double in volume within 6 months to a year. Although some tumors adhere to one or another of these growth patterns, others appear to alternate between periods of no or slow growth and rapid growth. Tumors that have undergone cystic degeneration (presumably because they have outgrown their blood supply) are sometimes capable of relatively rapid expansion because of enlargement of their cystic component. Because acoustic tumors arise from the investing Schwann cell, tumor growth generally compresses vestibular fibers on the surface. Destruction of vestibular fibers is slow; consequently, many patients experience little or no disequilibrium or vertigo.[8] Once the tumor has grown sufficiently large to fill the internal auditory canal, it may continue growth either by expanding bone or by extending into the cerebellopontine angle. Growth within the cerebellopontine angle is generally spherical.

Acoustic tumors, like other space-occupying lesions, produce symptoms by any of 4 recognizable mechanisms:

(1) compression or distortion of the spinal fluid spaces,

(2) displacement of the brain stem,

(3) compression of vessels producing ischemia or infarction, or

(4) compression and/or attenuation of nerves.

Because the cerebellopontine angle is relatively empty, tumors can continue to grow until they reach 3-4 cm in size before they contact important structures. Growth is often sufficiently slow that the facial nerve can accommodate to the stretching imposed by tumor growth without clinically apparent deterioration of function.

Tumors that arise within the internal auditory canal may produce relatively early symptoms in the form of hearing loss or vestibular disturbance by compressing the cochlear nerve, vestibular nerve, or labyrinthine artery against the bony walls of the internal auditory canal. As the tumor approaches 2.0 cm in diameter, it begins to compress the lateral surface of the brain stem. Further growth can occur only by compressing or displacing the brain stem toward the contralateral side. Tumors greater than 4 cm often extend sufficiently far anteriorly to compress the trigeminal nerve and produce facial hypesthesia. As the tumor continues to grow beyond 4 cm, progressive effacement of the cerebral aqueduct and fourth ventricle occurs with eventual development of hydrocephalus.[9]

Symptoms of Acoustic Neuroma

Hearing loss is the most frequent symptom, occurring in more than 95% of patients. About 90% present with a one-sided, slowly progressive hearing impairment Figure 2.
A high-frequency sensorineural pattern is the most common type, occurring in approximately two-thirds of patients. In the remaining third, the next most common observation is hearing loss at low frequency (which would be more typical of Meniere's disease). Even less commonly, some have the "cookie bite" pattern (suggestive of congenital hearing loss). A sudden hearing loss occurs in about 25% of patients with acoustic neuroma. However, because acoustic neuroma is a rare condition, sudden hearing loss attributable to an acoustic tumor occurs in only 1 to 5% of patients with sudden hearing loss as there are many more common causes (Daniels et al, 2000). Hearing can be completely normal in about 11% of patients (Morrison and Sterkers, 1996). Tinnitus is very common in acoustic neuroma, and is usually unilateral and confined to the affected ear.In spite of the usual origin of acoustics in the vestibular nerve (Komatsuzaki and Tsunoda, 2001), vertigo (spinning) is not common, occurring in only about 20% of persons with acoustic neuroma. Vertigo is more common with smaller tumors than larger ones. Unsteadiness is much more prevalent than vertigo, and approximately 70% of patients with large tumors have this symptom. Cerebellar symptoms (that is, poor coordination of the arms and legs) are unusual. Facial sensory disturbances occur only in large tumors (about 50% of those greater than 2 cm in size). The facial sensory disturbance may respond to carbamazepine medication for neuralgia. Facial weakness is uncommon. Facial twitching, also known as facial synkinesis or hemifacial spasm, occurs in about 10% of patients. Headache prior to surgery occurs in roughly 40% of those with large tumors. [10,11]

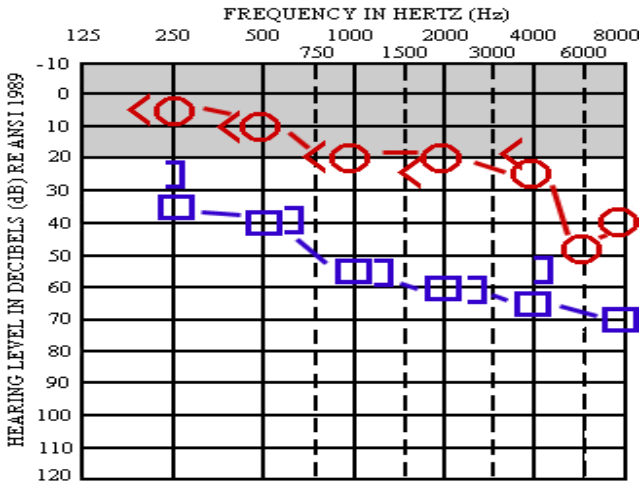


Figure 2: typical audiogram (hearing test) in a patient with an acoustic neuroma

Clinical manifestations

The earliest symptoms of acoustic neuromas include ipsilateral sensorineural hearing loss/deafness, disturbed sense of balance and altered gait, vertigo with associated nausea and vomiting, and pressure in the ear, all of which can be attributed to the disruption of normal vestibulocochlear nerve function. Additionally more than 80% of patients have reported tinnitus (most often unilateral high-pitched ringing, sometimes a machinery-like roaring or hissing sound, like a steam kettle). Large tumors that compress the adjacent brainstem may affect other local cranial nerves. Involvement of the nearby facial nerve (CN VII) may lead to ipsilateral *facial weakness*, sensory impairment, impairment of glandular secretions and loss of taste sensation in the anterior two-thirds of the tongue; involvement of the trigeminal nerve (CN V) may lead to loss of sensation in the involved side's face and mouth. The glossopharyngeal and vagus nerves are uncommonly involved, but their involvement may lead to altered gag or swallowing reflexes.Larger tumors may lead to increased intracranial pressure, with its associated symptoms such as headache, vomiting, and altered consciousness.

Different diagnostic techniques [12,13,14]

Conventional audiometry is the most useful diagnostic test for acoustic neuroma. The most common abnormality is an asymmetrical high-frequency sensorineural hearing loss. However, recall that only about 1 in 1,000 patients with hearing asymmetry have acoustics. It has been estimated that 5% of persons with sensorineural hearing loss have acoustics (Daniels et al, 2000), but this estimate is suspect as it would imply a much higher prevalence of acoustic neuromas than are commonly accepted.

Auditory brainstem response

When abnormal with a progressively worsening pattern, audiometry usually leads to further testing such as ABR (auditory brainstem response) and gadolinium enhanced MRI, which establishes the diagnosis. ABR testing is less sensitive than MRI, but it is considerably less expensive. A new technique, called summated ABR, which is essentially several ABRs compared over time, may provide better sensitivity.

MRI (Magnetic resonance image**),** On MRI, acoustic neuromas are frequently uniformly enhanced, dense, and expand the internal auditory meatus. A fast spin-echo T2 variant of MRI is very sensitive to acoustics, and in some clinical settings, can be done fairly inexpensively.

Electronystagmography (ENG testing) is frequently abnormal, and about 50% of all tumors are associated with unilateral loss of calorics. Nevertheless, ENG is not a reasonable diagnostic test because it is not specific.

Rotatory chair testing is less sensitive than caloric testing. Posturography is insensitive to acoustic neuroma.

Various treatements of acoustic neuroma

About 25% of all acoustic neuromas are treated with medical management consisting of a periodic monitoring of the patient's neurological status, serial imaging studies, and the use of hearing aids when appropriate. There are four distinct treatment options:

- Medical treatment or “wait and see” (conservative management)
- Surgery
- Gamma-knife procedure
- Cochlear implantation

Medical Treatment

About 25% of all acoustic neuromas are treated with medical management. Medical management consists of periodic monitoring of the patient’s neurological status, use of hearing aids when appropriate, and serial imaging studies.. Once a tumor is diagnosed, a repeat scan is obtained at six months and then at yearly intervals (Perry et al, 2001). This treatment has its own risks. Even when the tumor is not growing on MRI, there is a risk of losing useful hearing in this situation, making the individual no longer a candidate for hearing preservation type surgery. Approximately 20% of acoustics treated with medical management ultimately require surgery or radiotherapy (Yoshimoto, 2005; Yamakami, 2003). Oral prednisone may improve hearing in patients undergoing medical management with acute hearing loss (Aronzon et al, 2003). This route is common among patients over 70 years old. Records suggest that about 45% of acoustic neuromata do not grow detectably over the 3–5 years of observation.

Drugs acting on Acoustic Neuroma

Many types of treatment have been proposed, including different approaches like

a) Cognitive therapies,

b) Electric and magnetic brain stimulation,

c) Herb therapies and drugs.

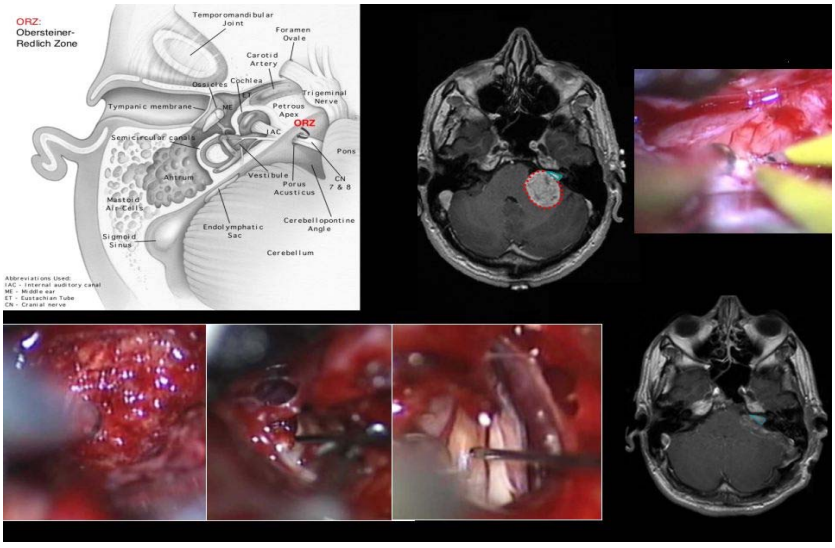
Many drugs have been tried for tinnitus, most of them with weak results. Methodological weakness and difficulties to measure tinnitus may account for some failures. In the last years, drug therapies have been focused in drugs that act in brain neurotransmitters, like Glutamate , GABA , serotonin , acetylcholine and dopamine.

Glutamate has shown some results with a double action mechanism (potencializes GABA transmission and blocks NMDA). Acting at the serotoninergic system could be of value at tinnitus treatment. Serotonine has been associated with hyperactivity and blood levels were found to be higher in tinnitus patients. Blockage of 5-HT receptors could be helpful for some patients. Drug therapy could be optimized if one could target the drug for the imbalanced neurotransmitter in a specific patient. Many methods are promising for this, like the MRI spectroscopy and the use of electrophysiological methods (electrocochleography and acoustic otoemissions).[15,16]

Preliminary results from the study "Tinnitus treatment with Dopamin guided by electrocochleography and acoustic otoemissions" seem to confirm the idea that electrophysiological methods could help in rational tinnitus drug therapy: a dopamine agonist that acts at D2 Dopamine receptors of neural terminations at the inner ear, tinnitus improvement (measured with the Tinnitus Handicap Inventory and a Visual-Analog Scale) was statistically correlated with the findings of a double-peak and enlargement of the action potential (CAP duration of 2,4 ms or more). This may be due to hyperactivity at the neural termination, which may be intrinsic and/or related to Efferent System dysfunction.[17]

Surgery

About half of all acoustic neuromas are presently treated with surgery. In most instances, surgical removal of the tumor is the preferred option because it prevents potentially fatal complications of tumor growth. Surgery may enable preservation of hearing.



Radiation therapy

Radiation therapy is done in a variety of ways, but mainly by three methods: gamma knife radiosurgery or fractionated stereotactic radiotherapy, with a linear accelerator (linac), or proton therapy. [18] In the gamma knife approach, 201 beams of gamma radiation are focused on the tumor in a single session. The damage to the tumor at the convergence point may cause it to stop growing but usually does not cause it to shrink in the long term. It may cause short-term shrinkage due to necrosis in the tumor. The damage may be to the tumor cells and/or to the tumor vasculature.

Gamma Knife

This is a method of irradiating the tumor, invented by Lars Leksell in 1971. Gamma knife stereotactic radio surgery has become more prevalent recently as it has been demonstrated to be safe and effective in the control of acoustic neuromas (Likhterov, 2007). Gamma knife does not generally make tumors go away — Figure 3 is actually that of a patient who had gamma knife surgery several years prior. Instead, gamma knife radiation shrinks the tumor and prevents future growth in most patients. Patients are best followed with periodic MRI scans for the remainder of their lives. The recurrence rate of the tumor is about 3% after surgery, and 14% after gamma knife, but of course, this figure will vary with the surgeon and the gamma knife protocol.[19]

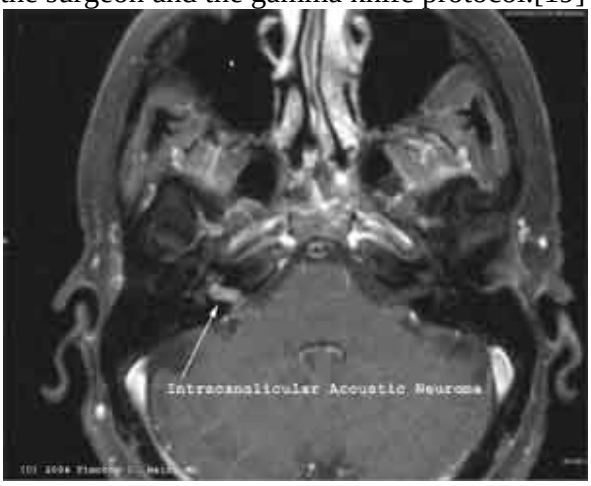


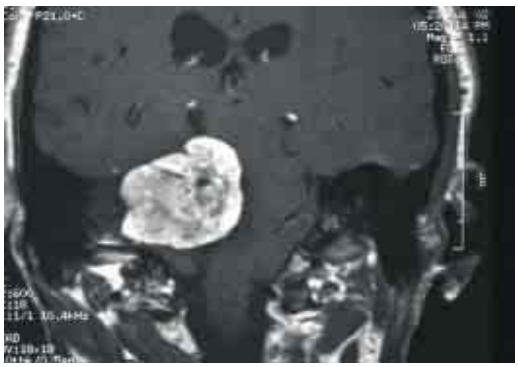
Figure 3: MRI scan of the brain showing an acoustic neuroma (the white spot on the left side of the picture).

Stereotactic radiotherapy

Radiation other than gamma rays can also be used to treat acoustic neuroma. It is similar to gamma knife in its overall features. Long term hearing preservation is very rare in persons with Stereotactic radiotherapy (6.7% according to Lin et al, 2005). We see no particular reason to seek out Stereotactic radiotherapy rather than gamma knife. The chance of recurrent tumor using current dose regimens is roughly 5-10%. Tumor growth is rare in patients who remain stable 6-7 years post therapy.

Proton therapy

A machine uses a beam of protons to kill the tumour and a cyclotron is used to generate the beam. Theoretically, this is preferable to the x-rays used by the linac and gamma knife machines as the protons can, in theory, be stopped before they exit the tumor, thus reducing damage to normal tissue. However, to date, very few people have been treated by this method and the results known are not statistically good.[20]



Very large acoustic neuroma (coronal view-large white blob)

Cochlear implantation

Very rarely, a person with acoustic neuroma might desire a cochlear implant. This might occur if an acoustic tumor is present in the only hearing ear, or after surgery to remove bilateral acoustic neuromas. Belal (2001) reported that cochlear implantation is possible only if there is an intact cochlear nerve (as shown by a positive response to promontory stimulation), and if the implantation is done at the time of acoustic tumor removal, before the cochlea ossifies (turns to bone).

Conclusion

The term "neuroma" means "nerve tumor" but an acoustic neuroma is a Schwannoma. Because it involves the vestibular portion of the 8th cranial nerve and it arises from Schwann cells, which are responsible for the myelin sheath in the peripheral nervous system. Scientific studies showed cellular phone use or prolonged exposure to loud noises, are giving preliminary evidence (Christensen et al, 2004; Edwards et al, 2006; Edwards, 2007; Hardell, 2003; Lonn et al, 2004; Schoemaker et al, 2005; Schlehofer, 2007). There is not hard evidence supporting a link between environmental factors and acoustic neuromas. There is no optimized surgical or treatment regimn was established. Moreover the major risk factors for causing acoustic neuroma was causing cellular phones and more exposure of high frequency sounds.This reviewe suggests that to avoid more expose to cellular phones and high frequency sounds to prevent acoustic neuroma.

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