

Review Article

Vestibular schwannoma: anatomical, medical and surgical perspective

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ABSTRACT

The term "acoustic" is a misnomer, as the tumor rarely arises from the acoustic (or cochlear) division of the vestibulocochlear nerve. The correct medical term is vestibular schwannoma, because it involves the vestibular portion of the 8th cranial nerve. They are benign, rather rare tumors. They expand in size and grow larger; they can push against the brain. While the tumor does not actually invade the brain, the pressure of the tumor can displace brain tissue.

Keywords: Acoustic neuroma, Benign, Merlin, Schwannoma, Vestibular, Cochlear

MAIN POINTS

1. Acoustic neuromas are a rare cause of unilateral hearing loss, dizziness, as well as rarely other symptoms related to the brain.
2. The best tests to diagnose acoustic neuroma are audiometry (hearing testing) and MRI scanning of the head with gadolinium contrast.
3. Half of all acoustic neuromas are treated by surgery, about a quarter with radiation (this is increasing), and about a quarter are watched.
4. No matter what method of treatment is used, hearing preservation is very unlikely.

INTRODUCTION

The internal auditory canal (IAC) is 10–17 mm in length and contains the facial, vestibular and cochlear nerves, surrounding blood vessels and a common dural sheath.¹

An acoustic neuroma is a benign tumor involving cells of the myelin sheath that surrounds the vestibulocochlear nerve (eighth cranial nerve). Acoustic neuromas are often called vestibular schwannomas because they are tumors that arise from the myelin sheath that surrounds the vestibular nerve. Acoustic neuromas are considered

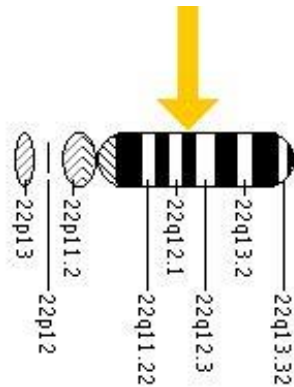
benign (non-cancerous) tumors since they do not spread to other parts of the body. They can occur anywhere along the vestibular nerve but are most likely to occur where the vestibulocochlear nerve passes through the tiny bony canal that connects the brain and the inner ear. A unilateral tumor is a tumor arising from one nerve and a bilateral tumor arises from both vestibular nerves. Unilateral acoustic neuromas usually occur spontaneously (by chance). Bilateral acoustic neuromas occur as part of a hereditary condition called Neurofibromatosis Type 2 (NF2).

They are typically benign and slow growing, but can cause symptoms through mass effect and pressure on local structures, eventually becoming life-threatening. Considering the possibility can enable earlier diagnosis, increase management options and may decrease morbidity.

Acoustic Neuroma represents 6-10% of primary intracranial tumours but is the most common form of cerebellopontine angle (CPA) tumour. Sporadic tumours are unilateral and account for 95% of cases, whilst those associated with neurofibromatosis are bilateral and account for the other 5%.

GENETICS

The NF2 gene (Schwannomerlin, Schwannomin, MERL_HUMAN, merlin Moesin-Ezrin-Radixin-Like Protein) is located on the long (q) arm of chromosome 22 at position 12.2.



More precisely, the NF2 gene is located from base pair 28,329,564 to base pair 28,424,586 on chromosome 22.

The NF2 gene provides instructions for the production of a protein called merlin, also known as schwannomin. This protein is made in the nervous system, particularly in specialized cells that wrap around and insulate nerves (Schwann cells).

Merlin is believed to play a role in controlling cell shape, cell movement, and communication between cells. To carry out these tasks, merlin associates with the intern Acoustic neuroma, Benign, Mer Acoustic neuroma, Benign, Merlin, Schwannoma, Vestibular, Cochlear lin, Schwannoma, Vestibular, Cochlear al framework that supports the cell (the cytoskeleton). Merlin also functions as a tumor suppressor protein, which prevents cells from growing and dividing too fast or in an uncontrolled way. More than 200 mutations in the NF2 gene have been identified in people with neurofibromatosis type 2. Research suggests that the loss of merlin allows cells, especially Schwann cells, to multiply too frequently and form noncancerous tumors. The most common tumors in neurofibromatosis type 2 are vestibular schwannomas, which develop along the

nerve that carries information from the inner ear to the brain. Other tumors affecting the nervous system also occur in people with this condition.

Embryological considerations: Certain structural peculiarities of the acoustic nerve ostensibly predispose to neoplastic transformation, for unlike other cranial and spinal nerves, glia rather than Schwann cells frequent its proximal 8- to 12-mm segment. Only upon entry through the porus acusticus does the nerve acquire a vestment of Schwann cells and assume the true characteristics of a peripheral nerve. It is principally at this interface, between a stroma of oligodendroglia and Schwann cells that neoplastic transformation occurs. Interestingly, the propensity for neoplasia is also evidenced at a comparable location in fetal rats exposed transplacentally to nitrosourea.

Characteristically, acoustic schwannomas arise in the vestibular division, predictably in that short segment of passage through the porus acusticus or the internal auditory canal. This canal is approximately 2 cm. (0.8 inches) long and it is here that acoustic neuromas originate from the sheath surrounding the eighth nerve. The youthful neoplasm compresses the cochlear division and may also obstruct the labyrinthine blood vessels that supply the organ of Corti and the vestibular end organs. Although the origin of the tumor is typically within the vestibular division, the earliest symptoms generally do not reflect this localization but rather the compression of the auditory component. Tinnitus is the most common initial symptom and in approximately 25 percent of cases, is forthwith accompanied by vertigo

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. The increased pressure in the posterior fossa and the disfigurement of the foramina of Luschka and of Magendie dispose to obstruction of cerebrospinal fluid (CSF) flow and the development of hydrocephalus.

As the intruder enters the cerebellopontine angle, it acquires a vestment of arachnoid. This delicate membrane may remain distinct while becoming fibrotic, but more often merges with the fibroblastic tumor and imparts an appearance of encapsulation. On occasion a vestment of arachnoid about the tumor creates a cul-de-sac that fills with clear fluid, presumably CSF. Such cysts may add significantly to the dimensions of the mass and augment compression.

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.

SIGNS AND SYMPTOMS

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 a unilateral high-pitched ringing, sometimes a machinery-like roaring or hissing sound, like a steam kettle) as well as Headache.

Headache

- Upon awakening in the morning
- Wakes patient from sleep
- Worse when lying down, reclining position
- Worse when standing up
- Worse when coughing, sneezing, straining, lifting (Valsalva maneuver)
- With nausea or vomiting

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, and impairment of glandular secretions; involvement of the trigeminal nerve (CN V) may lead to loss of taste and 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.

Even larger tumors may lead to increased intracranial pressure, with its associated symptoms such as headache, vomiting, and altered consciousness.

CONFIRMED RISK FACTORS INCLUDE

Neurofibromatosis: 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.¹

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%.

1. High dose ionising radiation (Children who received radiation for benign conditions of their head and neck, for example, to decrease the size of

their tonsils and adenoids, were at increased risk of developing AN much later in life² but the medical use of low dose ionising radiation such as used in imaging has not been established as a risk.²

2. Hay fever has been suggested as a risk factor although not confirmed.³
3. Occupational noise exposure⁴ has not been shown to be a risk factor although some other epidemiological studies have suggested a link. Similarly risk due to radiofrequency exposure from mobile phone use remains contentious: the large Interphone case-control study suggested no increased risk in the first decade of use but the risk of longer term use is unknown.⁵

Among cellular telephone users, it was more common that the tumor appeared on the contralateral side of the head than that of the ear used during phone calls.⁶

There is public concern that use of mobile phones could increase the risk of brain tumours. If such an effect exists, acoustic neuroma would be of particular concern because of the proximity of the acoustic nerve to the handset.⁷

Acoustic neurinomas occur more often in women than men with an approximate ratio of 3: 2. Interestingly, the same ratio obtains with meningiomas.

S-100 appears to be a useful marker for identifying neoplasms derived from Schwann cells and melanocytes. Acoustic neuroma, Benign, Merlin, Schwannoma, Vestibular, Cochlear cytes.

MICROSCOPIC HISTOLOGY

Upon microscopic examination, the acoustic neurinoma presents two distinctive architectural patterns, designated Antoni A and Antoni B. Both are created by spindle cells with elongated nuclei and fibrillated cytoplasm, predominantly those of Schwann cells. The two tissue patterns differ in cellular weave and density.

Antoni A tissue is compact, with a prominence of interwoven fascicles. Antoni B tissue is porous and less structured. The cells are dispersed randomly about blood vessels, microcysts, collections of xanthomatous cells and sites of previous hemorrhage. Lymphocytes attest to antecedent degenerative events within Antoni B tissue. The degree of nuclear pleomorphism varies considerably among acoustic neurinomas as well as between different areas within the same tumor. This pleomorphism often contributes a random population of large, bizarre nuclei that taunt the pathologist with thoughts of anaplasia: however, fortunately, malignant transformation is of a rarity that permits individual case reports. Mitotic figures are most infrequent. Necrosis is commonly present but most often testifies to the meagerness of native blood vessels and their compression by tumor expansion within a restricted compartment.

SIGNS AND TESTS

The health care provider may diagnose an acoustic neuroma based on the history, neurological examination or testing of the patient. The results of a physical examination are often unremarkable, except for the following signs:

1. Facial drooping on one side
2. Unsteady walk
3. Drooling

If the tumor is large, there may additionally be signs of increased pressure on the brain (increased intracranial pressure), including the following:

1. Nausea and vomiting
2. Unilateral (one side only) dilated pupil -- see eyes, pupils different size
3. Sleepiness

The most useful (i.e., sensitive and specific) test to identify acoustic neuromas is an Gadolinium enhanced MRI of the head. Other useful tests used to diagnose acoustic neuroma and to differentiate it from other causes of dizziness or vertigo include:

Audiology (a test for hearing) The most common abnormality is an asymmetrical high-frequency sensorineural hearing loss.

Caloric stimulation (a test for vertigo).

Electronystagmography (a test of equilibrium and balance) After routine auditory tests reveal loss of hearing and speech discrimination ("I can hear a sound in that ear, but I can't understand what is being said."), an auditory brainstem response test (ABR) may be done.

Brainstem auditory evoked response (BAER, a test of hearing and brainstem function) Acoustic neuroma, Benign, Merlin, Schwannoma, Vestibular, Cochlear.

TREATMENT

Surgery

Goals of surgical treatment are removal of the tumor and prevention of facial paralysis. Preservation of hearing is more difficult. If a tumor is removed when it is very small, hearing may be preserved. Any hearing that is lost prior to surgery will not be regained. Large tumors usually result in total loss of hearing on the affected side.

Large tumors may also compress nerves important for facial movement and sensation. These tumors can typically be safely removed, but the surgery often results in paralysis of some facial muscles.

Extremely large tumors may additionally compress the brainstem, threatening other cranial nerves and preventing the normal flow of cerebrospinal fluid. This can lead to a build-up of fluid in the head (hydrocephalus) which can cause potentially life-threatening increased intracranial pressure. Goals of surgery in these cases are treatment of the hydrocephalus and decompression of the brainstem. Common surgical approaches to Acoustic Neuroma

Translabyrinthine (through the inner ear). Hearing loss is expected and inevitable. Not appropriate for very large tumors.

Retro sigmoid or sub occipital (through the skull behind the ear). Retraction of the cerebellum (part of the brain) is necessary. Headaches are common after this approach.

Stereotactic radio surgery

The goal of radiation therapy is to slow or stop the tumor growth, not to cure or remove the tumor. Stereotactic radio surgery targets tumour with a single large dose of radiation using convergent beams of high energy x-rays, gamma-rays ('gamma knife radiosurgery') or charged particles and is an alternative, emergent treatment. Other potential longer-term risks associated with stereotactic radiosurgery and stereotactic radiotherapy include:

- Radiation-induced brain necrosis⁸
- Radiation-related cranial nerve injury
- Malignant change (for example, to a glioblastoma multiforme)⁹

Salvage surgery for neuromas post-radiotherapy is technically more difficult than primary surgery

Radiosurgery is often performed in elderly or sick patients who are unable to tolerate brain surgery.

Sometimes during brain surgery to treat acoustic neuromas, not all of the tumor can be safely removed, and some residual tumor must be left behind. Radiosurgery is often used post-operatively to treat residual tumor in these cases.

Radiosurgery is only appropriate for small tumors, so that radiation damage to surrounding tissues can be minimized.

Like brain surgery, radiosurgery can sometimes result in facial paralysis or loss of hearing. Resection is indicated for patients with larger tumors which have caused major neurological deficits from brain compression. Surgeons perform stereotactic radiosurgery for small or medium-sized tumors with the goals of preserved neurological function and prevention of tumor growth. The long-term outcomes of radiosurgery, particularly with gamma knife technique, have proven its role in the primary or adjuvant management of this tumor. Fractionated radiotherapy has

been suggested as an alternative for selected patients with larger tumors for whom microsurgery may not be feasible or for some patients in an attempt to preserve cranial nerve function. Most such centers do not offer conformal radiosurgery. Patients with neurofibromatosis type 2 pose specific challenges, particularly in regard to preservation of hearing and other cranial nerve function.¹⁰

OBSERVATION

Since these tumors usually grow very slowly, small tumors that have minimal or no symptoms (asymptomatic) can be safely observed with regular MRI scans and left untreated unless they grow dangerously.

Very often elderly patients will die of other natural causes before small, slow growing tumors become symptomatic.

EXPECTATIONS (PROGNOSIS)

Acoustic neuromas are benign and noncancerous. They do not spread (metastasize) to other body systems, but they may continue to grow and compress vital structures within the skull.

COMPLICATIONS

Brain surgery results in complete removal of the tumor in greater than 95% of cases.

About 95% of patients with small tumors will have no permanent facial Acoustic neuroma, Benign, Merlin, Schwannoma, Vestibular, Cochlear I paralysis following surgery. However, roughly two-thirds of patients with large tumors will have some permanent facial weakness following surgery.

Approximately one half of patients with small tumors will retain useful hearing in the affected ear following surgery.

There may be delayed radiation effects following radiosurgery, including nerve damage, loss of hearing, and facial paralysis.

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