



REVIEW ON: ACOUSTIC NEUROMA

Rajas S. Sheth¹, Vaishnavi N. Nagapure², Mansi T. Rathod³ and Omkar A. Devade*⁴

^{1,2,3}Department of Pharmacology, AISSMS College of Pharmacy, Pune 01.

⁴Department of Pharmacology, YSPM YTC Faculty of Pharmacy, Satara.



*Corresponding Author: Omkar A. Devade

Department of Pharmacology, YSPM YTC Faculty of Pharmacy, Satara.

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ABSTRACTS

An acoustic neuroma is a noncancerous tumor that develops on the main nerve leading from the inner ear to the brain. This nerve is called the vestibular nerve. Branches of the nerve directly affect balance and hearing. Pressure from an acoustic neuroma can cause hearing loss, ringing in the ear and problems with balance. Another name for an acoustic neuroma is vestibular schwannoma. An acoustic neuroma develops from the Schwann cells covering the vestibular nerve. An acoustic neuroma is usually slow-growing. Rarely, it may grow quickly and become large enough to press against the brain and affect vital functions.

KEYWORDS: Acoustic Neuroma, Vestibular schwannoma, Treatment, Complication.

INTRODUCTION

Vestibular schwannoma (VS), acoustic neurinoma, vestibular neuroma, and acoustic neurofibroma are some of the various names for acoustic neuromas. The Schwann cells in the sheath encircling the vestibular and cochlear nerves are the source of these malignancies. An acoustic neuroma may develop extraaxially or intracranially. Acoustic neuromas typically lie in the

cerebellopontine angle, close to the vestibular and cochlear nerves, especially the vestibular nerve's inferior division. Meningiomas are tumors that can form anywhere in the brain and make up between 5% and 10% of tumors at the cerebellopontine angle (CPA). Bilateral auditory neuromas are usually associated with type 2 neurofibromatosis, however the vast majority are spontaneous.^[1-3]

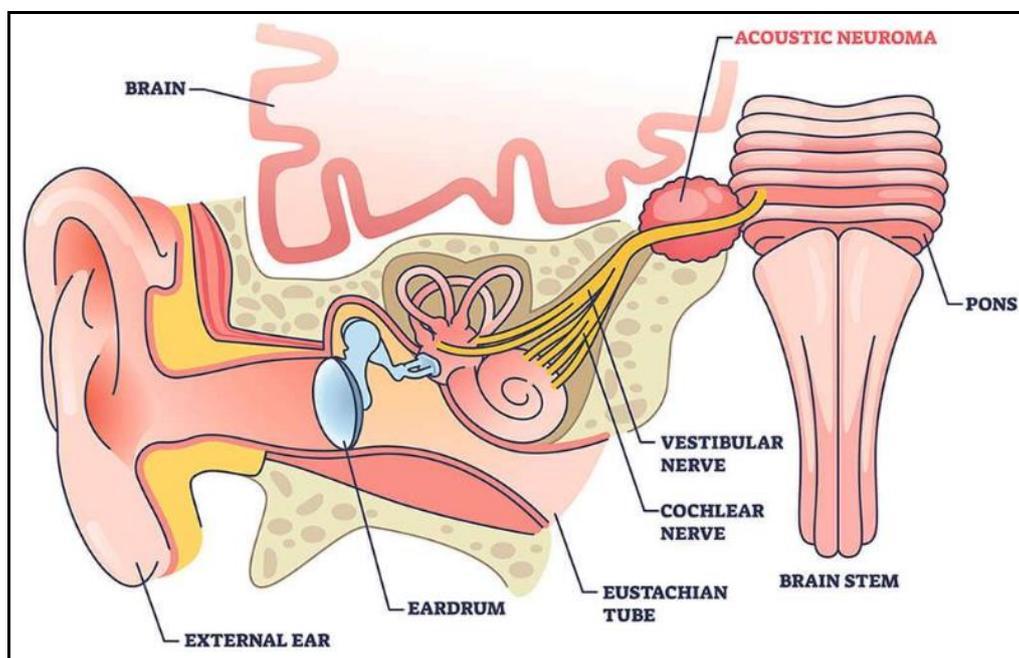


Fig. Acoustic neuroma.

Etiology

Neurofibromatosis type 2, which is brought on by a mutation on chromosome 22q12.2 at the locus of the neurofibromin 2 gene, which codes for the merlin protein, has been linked to bilateral acoustic neuromas. Research has revealed a mutation that predisposes someone to acquire an auditory neuroma. Furthermore, radiation exposure might make this illness more likely to manifest.^[4]

Although there have been worries about the radiation from mobile phones, numerous studies have not been able to prove a link between the radiation and the development of vestibular schwannomas.^[5]

Epidemiology

About 8% of all brain tumors with clinical manifestations are Schwannomas. The majority of acoustic neuromas are sporadic and unilateral. Bilateral acoustic neuromas, which make up less than 5% of all schwannomas, are influenced by genetic factors. Acoustic neuromas are typically identified in people between the ages of four and six.^[6] On the other hand, those with neurofibromatosis type 2 (NF II) typically manifest earlier in life, with the third decade of life marking the peak occurrence. Children can develop auditory schwannomas, albeit they are uncommon. Pregnancy can make symptoms worse, and there is a small female predilection.^[6] Despite neurofibromatosis type 1 (NF I) being more common, the hereditary form of auditory neuroma is more frequently linked to NF II are autosomal dominant, with genetic defects localized to chromosome 17 and chromosome 22, respectively.^[7]

Histopathology^[8]

The Obersteiner-Redlich zone, which marks the transition between glial and Schwann cells, is the location of origin for acoustic neuromas. Acoustic neuromas usually have a rubbery-firm consistency and a grayish-pale appearance when viewed in gross anatomy. They have a distinct capsule and can exhibit different levels of vascularity. Tumors can cause nerve fibers to be stretched and displaced, which can modify the appearance of the capsule in noticeable ways.^[9]

When an auditory neuroma is sectioned, the delicately trabeculated pattern on the sliced surface has a hard, pale gray look. Larger tumors may show signs of calcification points, bleeding, xanthomatous alterations, and cystic degeneration. These alterations contribute to the enormous tumors' varied appearance in terms of consistency and color. The internal auditory artery, which splits off into multiple small vessels on the tumor's surface, is the main source of the tumor's blood supply. Larger tumors might receive blood flow from tiny branches of nearby pontine and cerebellar arteries.

The tumor's cellular makeup is revealed by light microscopy analysis, showing spindle cells with elongated nuclei and fibrillary cytoplasm.^[10]

History and Physical^[11]

The compression of surrounding structures, such as cranial nerve VIII (the vestibulocochlear nerve), nearby cranial nerves, the cerebellum, the brainstem, and the ensuing rise in intracranial pressure (ICP), is the primary source of the signs and symptoms of acoustic neuroma. The majority of people with acoustic neuromas experience unilateral hearing loss as a result of damage to the auditory nerve or disruption of its blood supply. Tinnitus, impaired word interpretation, vertigo, headaches, and numbness are additional clinical characteristics. The mass inside the CPA will eventually crush the brainstem as the acoustic neuroma grows. Abnormalities in gait may arise from this compression of the brainstem. The clinical characteristics of an auditory neuroma are summarized as follows.

Involvement of Cranial Nerve VIII

The most prevalent and early sign of an acoustic neuroma is hearing loss. The impairment usually manifests as high-frequency retro-cochlear sensorineural hearing loss that progresses slowly. The hearing loss may go undetected or be mistaken for other conditions because of its gradual onset. To evaluate hearing impairment during a physical examination, a number of tests can be performed. Among these tests are the following ones: Speech discrimination exams assess a person's comprehension of speech. Using tuning forks with a broad frequency range Finding out if a hearing loss is bilateral or unilateral might be assisted by the Weber test. The Rinne test measures sound transmission via air and bone. Another common symptom of an acoustic neuroma is tinnitus, which is the sensation of ringing or buzzing sounds in the ears. One may experience tinnitus as a continuous or sporadic sensation. Vestibular symptoms, such as nystagmus and head instability, are caused by involvement of the vestibular part of the eighth cranial nerve (vestibulocochlear nerve).^[12]

Antoni A tissue is a compact, well-organized structure made up of long, bipolar cells arranged in an interlaced pattern. Antoni's fibers and nuclei A sample may have a characteristic pattern, sometimes taking the shape of a spiral framework like the appearance seen in meningiomas. Antoni B is typified by a less ordered cell arrangement and is frequently seen in big acoustic neuromas. Cells in Antoni B tissue are distributed irregularly and frequently gather near blood arteries, necrosis, previous bleeding, and cystic abnormalities. This tissue type also exhibits variable degrees of lymphocytic infiltration. The tissue type Antoni B is typically observed in bigger tumors and is believed to be the result of ischemia. The relative proportions of Antoni A and Antoni B tissue types determine the consistency of the acoustic neuroma. Although these changes are benign and malignant transformation is rare, nuclear pleomorphism is a typical feature of schwannomas. Mitotic figures, which are indicative of cell division, are relatively rare. Necrosis, if present, is attributed to poor

blood supply rather than rapid tumor growth. Edema, formation of micro or macrocysts, xanthomatous alteration, and areas of calcification are considered degenerative changes within the tumor tissue. In electron microscopy (EM), the characteristic basement membrane of Schwann cells can be observed. Additionally, wide-spaced collagen is often present, indicating abnormalities in collagen fiber arrangement.^[13]

Evaluation

Computed tomography scans or contrast-enhanced magnetic resonance imaging (MRI) are used to diagnose acoustic neuromas. Contrast compounds, such as gadolinium, help make tumors easier to see and provide details about the features of the tumor and its relationship to surrounding structures. In these experiments, contrast is also necessary to see tiny tumors. Audiometric tests are frequently required to evaluate a patient's hearing ability if they have a hearing impairment. Although the test known as auditory brainstem evoked response is helpful, it is not frequently employed as the main method of screening for acoustic tumors because of its inability to accurately identify tiny cancers.^[9-11] The "trumpeted internal acoustic meatus sign," which is characterized by an enlargement or widening of the orifice, can be caused by an intracanalicular component in the porus acusticus. On the other hand, if the tumor grows into the extrameatal space, it may cause an identifiable feature called the "ice cream cone" sign. On T1-weighted MRI images, the majority of acoustic neuromas seem hypo- to isointense, whereas on T2-weighted images, they exhibit heterogeneously hyperintensity. Furthermore, when a contrast agent like gadolinium is administered, these tumors frequently show notable contrast enhancement. Because fast spin-echo MRI is noninvasive and inexpensive, it could be useful as a screening test for acoustic neuromas.^[14]

Treatment / Management^[15-17]

There are various approaches of treating acoustic neuromas.

The following are possible therapeutic options:

- **Observation:** The best course of action is to track the tumor's progress over time if it is tiny and not causing any noticeable symptoms. The elderly patient with several comorbidities is also taken into consideration for this technique.
- **Stereotactic radiotherapy:** In this course of treatment, the tumor cells are destroyed by high-energy radiation. Cyberknife, often known as Gamma Knife, is a popular and accurate radiation therapy tool.
- **Surgery:** For bigger tumors or those causing symptoms, surgical tumor removal may be required.

The following are the primary surgical methods used to treat acoustic neuromas

1. Retrosigmoid approach: This method entails cutting the tumor out of the base of the skull through an aperture. This method offers good access to the

tumor and cranial nerves in this area. It is possible to approach a tumor of any size and maybe keep hearing.

2. Middle cranial fossa approach: With this technique, the tumor is accessible to the surgeon via the base of the skull's middle cranial fossa. Tumors with minimal cisternal components and a prominent intracanalicular component are most suited for this method. In most situations, hearing is retained. The primary drawback is the requirement for temporal lobe retraction, which increases the risk of venous infarction and postoperative seizures in the event that the Labbe vein is injured.
3. Translabyrinthine: This procedure is most frequently performed on patients who have large tumors and are deaf or hard of hearing. It entails taking out the tumor through the inner ear. This method's primary benefit is its ability to protect and expose the facial nerve at an early age. Furthermore, there is no need to retract the cerebellum. However, there is restricted access to the lower portions of the foramen magnum and the jugular foramen.
4. Within six to twelve months following surgery, an MRI is necessary to record the amount of tumor removed and create a baseline image. Anterior inferior cerebral artery injury, bleeding, cerebellar trauma, facial paralysis, hearing loss (the most common), and hydrocephalus are among the most frequent surgical consequences

Differential Diagnosis

Acoustic neuromas account for around 80% to 90% of CPA lesions. Differential diagnoses of an acoustic neuroma include the following:

- Meningioma (5%-10% of CPA lesions)
- Ectodermal inclusion tumors
- Epidermoid (5%-7% of CPA lesions)
- Dermoid
- Metastases
- Neuroma from cranial nerves other than cranial nerve VIII:
 - Trigeminal neuroma.
 - Facial nerve neuroma
 - Neurinoma of the lowest 4 cranial nerves (IX, X, XI, XII)
- Extensions of nearby lesions in the CPA:
 - Dolichobasilar ectasia
 - Aneurysm
 - Cholesterol granuloma (distinct from epidermoid)
 - Neurenteric cyst
 - Arachnoid cyst
 - Brainstem or cerebellar glioma
 - Pituitary adenoma
 - Craniopharyngioma
 - Chordoma and tumors of the skull base
 - Fourth ventricle tumors (ependymoma, medulloblastoma)
 - Choroids plexus papilloma from the fourth ventricle through foramen of Luschka

- Glomus jugulare tumor
- Tumors of the temporal bone

Complications^[12-14]

Most complications are related to the surgical procedure and include the following:

- Injury to the anterior or posterior inferior cerebellar arteries
- Neurological injury
- Brain herniation
- Brain hemorrhage
- Injury to the cerebellum
- Facial paralysis
- Hearing loss

After surgery, tinnitus is a common problem in at least 10% to 20% of patients. The recurrence rate after excision is less than 5%. Facial nerve paralysis has been shown to occur in about 15% to 30% of patients, but most make a complete recovery over time. Hearing loss occurs in more than 50% of patients and may not improve. Residual hearing loss has a significant impact on the quality of life.

Postoperative and Rehabilitation Care^[15-19] Rehabilitating

The residual vertigo and disequilibrium that both surgical and nonsurgical patients frequently experience can be managed with activities focused on adaptation, substitution, and habituation. The following exercises are among them, but they're not the only ones: Exercises for gaze stability and adaptation Habituation is the practice of repeatedly doing movements that cause symptoms. Alternate: active eye movements between targets or visual control Patients who have had surgery for an acoustic neuroma may develop facial palsy as a side effect. Rehabilitation techniques, however, can help patients restore function by addressing these problems. The two methods that can be used are motor imagery and mirror therapy. Programs for vestibular rehabilitation should be tailored to the unique impairments that each patient has because research has shown that a focused approach is more effective for these people than.

Pearls and Other Issues^[20]

Early diagnosis and detection of acoustic neuromas can enhance treatment results and protect vital capabilities including hearing and facial nerve function. There are several possible treatment choices, including as radiation therapy, surgery, and observation. Comprehensive care and optimal treatment techniques can be provided by a multidisciplinary team consisting of neurosurgeons, otolaryngologists, radiation oncologists, and rehabilitation specialists. following an acoustic neuroma is surgically removed, recurrence is rare, but it is important to note that tinnitus may get worse in some people following the procedure. For many people, facial paralysis and hearing loss may go better with time. For both technical and prognosis reasons, it is imperative to

differentiate between an acoustic schwannoma and a meningioma prior to surgery.

Enhancing Healthcare Team Outcomes^[12-14]

In clinical practice, acoustic neuromas are not often occurring tumors. On the other hand, they frequently exhibit unilateral hearing loss. So, when making a differential diagnosis, medical professionals should take the lesion into account. An interdisciplinary team of radiation oncologists, neurosurgeries, otolaryngologists (ENTs), hearing and rehabilitation specialists, and others is most suited to address the tumor after it has been discovered.

Tinnitus is a prevalent post-operative issue in at least 10-20% of patients. After excision, the recurrence rate is less than 5%. 15% to 30% of individuals have been reported to experience facial nerve paralysis; nevertheless, the majority eventually recover fully. In excess of 50% of patients experience hearing loss, which may not get better. The quality of life is greatly impacted by residual hearing loss.

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