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A Review on Acoustic Neuroma

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ATHIRA PRASAD [1*], SOUMYA.R.V [2], PRASOBH.G.R [3]

1 Fifth Pharm D Students, Sree Krishna College Of Pharmacy And Research Centre Parassala, Trivandrum, Kerala, India*

2-Associate Professor, Department Of Pharmacy Practice, Sree Krishna College Of Pharmacy And Research Centre Parassala, Trivandrum, Kerala, India

3-Principal, Head of the department of pharmacy practice, Sree krishna college of pharmacy and research, centre, Trivandrum, Kerala, India

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ABSTRACT

Acoustic neuroma is also known as vestibular schwannomas which is a benign brain tumor, from 8th cranial nerve. Its non cancerous tumor in the main nerve leading from the inner ear to brain. There are several reports that explains discernable growth pattern for those tumors. However, their growth predictably maintain low, because of slow and indeterminate changes with time and the subsequent follow up reports do not exceed more than 3 years. The main clinical presentation is balance disorder or dizziness. Sensory symptoms include pins and needles or reduced sensation of touch. The common symptoms are hearing loss, rapid involuntary eye movement, or ringing in the ears. The main cause is linked to a problem with a gene on chromosome 22. In the normal case, this gene makes a tumor suppressor protein which will helps in control of growth of Schwann cells covering the nerves. Magnetic resonance (MRI) is one of the diagnosis which detects 1-2 millimeters sized tumor. The treatment includes stereotactic and radiation therapy.

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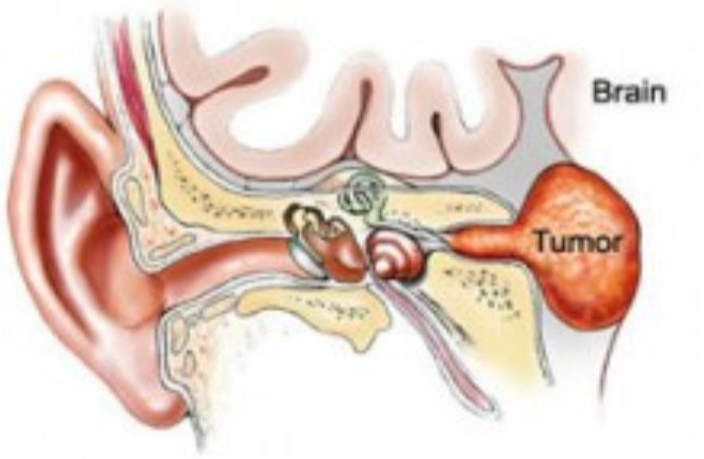
INTRODUCTION

Vestibular schwannomas are benign brain tumors that are supposed to arise from Schwann cells in the vestibular component of the 8th cranial nerve. These tumors have an occurrence of about 10–15/million a year. The major use of cranial imaging studies for diagnosis and management of several neurological condition lead to earlier detection of these tumors and may contribute to an incidence rate that is seen higher than earlier[1]. Early detection of small tumors results in a spectrum of size ranging from intracanalicular tumors to large extra metal extensions. The average age at diagnosis ranges between 46 and 58 years, with in younger patients often associated with neurofibromatosis type 2 (NF-2). The clinical symptoms varies depending on the size at diagnosis, presence of mass effect on the brain stem and whether there is any occlusion to cerebrospinal fluid (CSF) pathways[2]. The Schwann cells function is to protect and speed along balance and sound information to the brain. Without any specific reason, a mutation in the tumor suppressor gene, NF2, located on chromosome 22, leads to abnormal production of the cell protein called Merlin, and Schwann cells multiply to form a tumor. The origin of the tumor is mostly on the vestibular division of the nerve rather than the cochlear division, but hearing and balance will be affected as the tumor size increase. The vast majority of these VS (95%) are unilateral, in one ear only[3] They are called “sporadic” (it can be by-chance, non-hereditary). As it is non-cancerous, they can be harmful or may be life-threatening if they grow to put pressure on other cranial nerves and vital structures such as the brainstem. Variations in the mutation suggest the nature of the tumor’s development. The only environmental exposure that has been associated with the growth of a Vestibular schwannoma is therapeutic radiation exposure of the head. Unilateral/asymmetric hearing loss and/or tinnitus and loss of balance/dizziness are possibly early signs of a vestibular schwannoma[4]. Unfortunately, early diagnosis of the tumor may be difficult. Because the symptoms occur very often and may not appear in the beginning stages of growth. Magnetic resonance imaging (MRI) scans are crucial in the early diagnosis of a vestibular schwannoma and have a potential role in determining the location and size of a tumor. There are three options for treating a vestibular schwannoma, the first one is surgical removal, second is radiation, and the third is observation. Sometimes, the tumor is surgically extracted. The exact type of operation depends on the size of the tumor and the level of hearing in the affected ear[5]. If the tumor is small, hearing may be clear and the associated symptoms can be decreased by removing it to overcome the eventual effect on the hearing nerve. When the tumor grows bigger in size, surgical removal of the tumor lead to complication. The reason is

that the tumor may have damaged the nerves that play a crucial role incontrolling facial movement, hearing, and balance and may also have prominent affect on other nerves and certain primary structures of the brain.[6]



EPIDEMIOLOGY

Acoustic neuromas are benign tumors detected in 2,000 to 3,000 people per year, ie, it report an incidence of 1 per 100,000 per year. Acoustic neuromas (ANs) appear in approximately 6% of all intracranial tumors and have an incidence of 0.3 to 1 per 100 000 population per year. The use of thin-section gadolinium-enhanced magnetic resonance imaging (MRI) as the screening method for observation of retro cochlear disease has allowed detection of ever smaller tumors, many of which are associated with subtle, if any, symptoms[7-8]. This early diagnosis not only allows patients more therapy options but also potentially increase the outcome, mainly in the functional preservation of the auditory and facial nerves. However, a subset of the population is currently living with undiagnosed Acoustic neuroma and may be detected only after undergoing MRI for non neurotologicreasons.[10]

ETIOLOGIC FACTORS

The cause of acoustic neuromas can be associated with a problem of a gene on chromosome 22. Normally, this gene makes a tumor suppressor protein that helps control the growth of Schwann cells covering the nerves. Experts don’t know what causes this problem with the gene. In majority of the cases of acoustic neuroma, there is no known cause[9-11]This false gene is also inherited in neurofibromatosis type 2, which is a rare disorder that usually include the growth of tumors on the hearing and balance nerves on both sides of your head (bilateral vestibular schwannomas). There are two types of acoustic neuroma: a sporadic form and a

form associated with a syndrome called neurofibromatosis type II (NF2). NF2 is an inherited disorder specialized by the growth of noncancerous tumors in the nervous system [13]. Acoustic neuromas are the most common of these tumors and often occur in both ears at the age of 30.NF2 is a rare disorder. It accounts for only 5% of acoustic neuromas. This means the majority are the sporadic form. Doctors aren't sure what causes the sporadic form.[14]

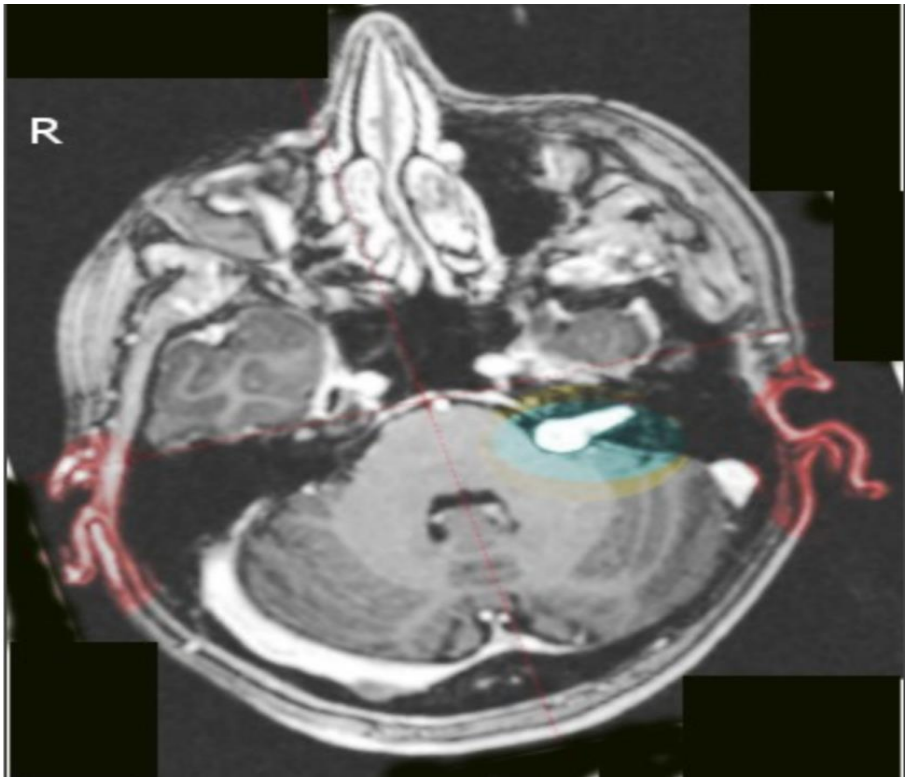
CLINICAL PRESENTATION

Signs and symptoms of acoustic neuroma are often easy to miss and may take more years to develop. They usually occur due to the tumor's effects on the hearing and balance nerves. Pressure from the tumor on nearby nerves controlling facial muscles and sensation (facial and trigeminal nerves), nearby blood vessels, or brain structures may also cause problems. As the tumor grows in size, it may cause more pronounced or severe signs and symptoms. [12-15]

Common signs and symptoms of acoustic neuroma include:

- Hearing loss, that may slowly worsen over months to years — although in rare cases sudden — and occurring on only one side or more severe on one side.
- Ringing (tinnitus) in the affected ear
- Taste changes
- Unsteadiness or loss of balance
- Dizziness (vertigo)
- Facial numbness and weakness or loss of muscle movement
- Headaches, clumsy gait, and mental confusion

It's must to see the doctor if experiencing these symptoms. Symptoms like clumsiness and mental confusion can signal a serious problem that requires urgent treatment. In some rare situations, an acoustic neuroma may grow large enough to compress the brainstem and may be life-threatening.[16]



RISK FACTORS

The one and only one risk factor for acoustic neuroma is having a parent with the genetic disorder neurofibromatosis 2 (NF2). Most of these tumors appear suddenly. They often occur in people with no family history of the disease. [17]

Scientists even now don't understand why some people get these tumors. Some risk factors might include:

- Loud noises
- A parathyroid neuroma, which is a benign tumor of the thyroid.
- Exposure to decreased levels of radiation during childhood

The risk of acoustic neuroma is lower among ever and current cigarette smokers than among never smokers, while there was no significant relationship for ex-smokers. However, no significant Relationship were found between acoustic neuroma and history of any allergic diseases, such as asthma, eczema, and seasonal rhinitis. [18]

UNILATERAL AND BILATERAL SCHWANNOMA

Unilateral vestibular schwannomas affect only one ear. They occur fairly often approximately 8 percent of all tumors inside the skull; and one out of nearly every 100,000 individuals per year reported with vestibular schwannoma. Symptoms can develop at any age but especially occur between the ages of 30 and 60 years. Most unilateral vestibular schwannomas are not hereditary and appear sporadically. A rough estimate of approximately one out of every 100,000 individuals per year develops a vestibular schwannoma.[14-17]

Bilateral vestibular schwannomas affect both hearing nerves and are mainly associated with a genetic disorder called neurofibromatosis type 2 (NF2). Half of affected individuals have inherited the disorder from an affected parent and half of them seem to have a mutation for the first time in their family. Each child of an suspected parent has a 50 percent chance of inheriting the disorder. Unlike those with a unilateral vestibular schwannoma, individuals with NF2 especially develop symptoms in their teens or early adulthood. Moreover, patients with NF2 usually develop multiple brain and spinal cord associated tumors. They also can develop tumors of the nerves important for swallowing, speech, eye and facial movement, and facial sensation[8]. Finding the best treatment of the vestibular schwannomas as well as the additional nerve, brain, and spinal cord tumors is more complicated than deciding how to manage a unilateral vestibular schwannoma. Further research is needed to evaluate the best treatment for individuals with NF2.

Scientists truly imagine that both unilateral and bilateral vestibular schwannomas form following the damage of the mechanism of a gene on chromosome 22. Scientists blindly believe that this particular gene on chromosome 22 produces a protein that controls the growth of Schwann cells. When this gene malfunctions, Schwann cell growth is uncontrolled, resulting in a tumor[11]Scientists also think that this gene may help control the growth of other types of tumors. In NF2 patients, the false gene on chromosome 22 is inherited. For individuals with unilateral vestibular schwannoma, however, some scientists concluded that this gene somehow loses its ability to function accurately.[14-16]

DIAGNOSIS

- Hearing test (audiometry): It's a test for hearing function that measures how well the patient hears sounds and speech, is especially the first test conducted to detect acoustic neuroma. The patient listens to sounds and speech by wearing earphones attached to a

machine which records responses and measures hearing function. The audiogram may show enhanced "pure tone average" (PTA), enhanced "speech reception threshold" (SRT) and lowered "speech discrimination" (SD).

- Brainstem auditory evoked response (BAER): This test is conducted in some patients to provide essential information on brain wave activity as a response to clicks or tones. The patient Focused and listened to those sounds at all times while wearing electrodes on the scalp and earlobes and earphones. The electrodes pick up and record the brain's response to these sounds.[15]

- Scans of the head: If other tests show that the patient may have acoustic neuroma, magnetic resonance imaging (MRI) is used as a major tool to confirm the diagnosis. MRI uses magnetic fields and radio waves, rather than x-rays, and computers to create complete pictures of the brain. It shows visual "slices" of the brain that can be added to create a three-dimensional picture of the tumor. A contrast dye is injected into the patient. If an acoustic neuroma is present, the tumor will soak up more dye than normal brain tissue and appear clearly on the scan. The MRI especially shows a densely "enhancing" (bright) tumor in the internal auditory canal.[16-18]

TREATMENT

These tumors can often be removed fully with surgery. Moreover, many are so small that they do not need spontaneous treatment. The management for acoustic neuromas depends on the size of the tumor and the patient's age, general health and preferences, and may involve surgery, radiosurgery and sometimes observation.[19]

Surgeons have developed various types of craniotomy to remove acoustic neuromas. Surgical craniotomy make use of a sub occipital, translabyrinthine approach or middle fossa approach.

- Keyhole Brain Surgery (Retro-sigmoid craniotomy)
- Translabyrinthine craniotomy

Complication for the surgery of large tumor is by the probable damage to hearing, balance, and facial nerves. Alternative treatment option is radiosurgery, which prominently and most carefully focusing on radiation to decrease the size or limit the huge growth of the tumor. People with small acoustic neuroma, surgery and radiation therapy are equally successful. Doctors may also suggest radiation therapy to treat older patients.[20-21]

- Radiosurgery

Radiosurgery management can be classified as single or fractionated (multiple smaller treatments other than one large one). Fractionated stereotactic radiosurgery (FSR) occur very often with high rates of control with preservation of hearing and preservation of facial strength. Surgery for acoustic neuromas may include removing all or part of the tumor. There are three main surgical methods for removing an acoustic neuroma:[21-24]

Translabyrinthine, which involves developing an incision behind the ear and taking off the bone behind the ear and some of the middle ear. This procedure is usually done for tumors larger than 3 centimeters. The benefit of this method is that it may allow the surgeon to see an important cranial nerve (the facial nerve) clearly before removing the tumor. The demerit of this technique is that it results in permanent hearing loss.

Retro sigmoid/sub-occipital is clearly defined as the exposing of the back of the tumor by opening the skull near the back of the head. This approach can be used for removing tumors of any size and offers the possibility of preserving hearing.[20-24]

Middle fossa, which involves removing a small piece of bone above the ear canal to access and remove small tumors positioned to the internal auditory canal, the narrow passageway from the brain to the middle and inner ear. Using this approach may enable surgeons to maintain a patient's hearing.

- Observation

In some slow-growing acoustic neuromas observation with slower management may be successful. This is a widely used treatment option for elderly or infirm patients with low onset of symptoms where the risks of treatment may be greater and where the tumor size does not increase during their lifespan. Observation is also called watchful waiting. Because acoustic neuromas are not cancerous and grow slowly, sudden treatment may not be necessary. Often doctors observe the tumor with periodic MRI scans and will suggest other treatment if the tumor size increase a lot or causes life threatening symptoms.[26]

- Radiation therapy

Radiation therapy is suggested in some cases for acoustic neuromas. Radiation therapy for this condition is usually given in one of two ways:

- Single fraction stereotactic radiosurgery (SRS), in which hundreds of small beams of radiation are focused at the tumor in a single shot.

- Multi-session fractionated stereotactic radiotherapy (FRS), which delivers smaller doses of radiation per day, usually over several weeks. Early studies recommend multi-session therapy may preserve hearing better than SRS.

After surgical treatment for acoustic neuroma, some patients suffer from hearing loss, leakage of cerebrospinal fluid, nerves damage in the face and other problems.[27-29]

- Surgical treatment for related problems

The bone-anchored hearing aid (Baha) speech processor is a hearing aid that doctors surgically implant under the skin close to the ear. The device grows into the skull bone and increases the bone's natural transmission of sound. The Baha speech processor transmits sound vibrations within the skull and inner ear that activate the nerves of the inner ear, allowing the patient to hear. We may suggest regular hearing aid instead of a Baha speech processor for some patients. Patients with neurofibromatosis 2 often develop acoustic neuromas in both ears. This results in life threatening hearing problems or deafness in both ears. Cochlear implants can provide a sense of sound to these patients and help them understand speech. The device must include an external earpiece and a device that the doctors have implanted via surgery to the skin near the ear. Cochlearim plants mechanism is by bypassing the destroyed portions of the ear and directly activating the hearing nerve. They create signals from the auditory nerve to the brain, which identifies the signals as sound. Leakage of Cerebrospinal fluid is a widely reported problem after surgical removal of an acoustic neuroma. Leaks are caused by a hole or tear in the dura, which is a membrane that envelopes the brain. Doctors carefully observes the patients for leakage of Cerebrospinal fluid, and if it occurs, they can conduct a procedure to stop the hole that is leaking cerebrospinal fluid.[29]

- Repairing facial nerve damage

If the patient's facial nerves are damaged by the acoustic neuroma, plastic surgeons may conduct procedures to reserve and restore the movement in the face.[30]

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