Acoustic neuromas, common benign tumors accounting for 10% of primary intracranial tumors, are the most common tumor in the cerebellopontine angle cistern. They almost always arise from Schwann cells of the vestibular portion of the vestibulocochlear nerve; therefore, vestibular schwannoma is the preferred term.

Patients usually present with unilateral hearing loss, often accompanied by tinnitus and vertigo. Sensorineural hearing loss is the result of extrinsic pressure from compression of the cochlear division of the eighth cranial nerve within the internal auditory canal by the tumor arising from the adjacent vestibular division. The accompanying facial nerve within the internal auditory canal is more tolerant of this local mass effect, and facial nerve symptoms such as ipsilateral facial paralysis are uncommon. Because this tumor grows slowly, symptoms evolve over months and years and the lesion can become quite large before it is detected. As the size of the tumor increases, it may extend into the posterior fossa. Symptoms related to increased intracranial pressure and cerebellar involvement such as ataxia may occur.

Treatment of vestibular schwannomas is aimed at removing the lesion or arresting its growth to prevent further neurologic impairment. Imaging plays a crucial role in both the detection and posttherapeutic management of these lesions.

Anatomy and Pathology
The anatomy of the facial and vestibulocochlear nerves must be understood to reliably evaluate and precisely localize...
pathology within the internal auditory canal and cerebellopontine angle. These nerves emerge from the brainstem at the pontomedullary junction and course through the cerebellopontine angle cistern. They enter the meatus, or porus acusticus, of the internal auditory canal as two separate structures. The shape of the facial nerve is tubular. Cross-sectional imaging viewed perpendicular to the long axis of the canal consistently shows the nerve in the anterosuperior quadrant of the internal auditory canal. The vestibular and cochlear components of the eighth cranial nerve usually enter the meatus as a single unit, and they divide into separate nerves within the internal auditory canal. The point of separation may vary among patients. The vestibulocochlear nerve assumes a crescent shape along the posterior and inferior aspects of the middle portion of the canal as the two components begin to divide.

Only in the lateralmost 3 to 4 mm of the internal auditory canal do the vestibular and cochlear nerves divide into separate identifiable structures. The cochlear division continues in the anterior-inferior quadrant of the canal beneath the facial nerve, and the vestibular nerves are located posterior to these two structures within the posterior half of the canal. At the fundus, the most lateral aspect of the canal, the vestibular nerve is separated into the superior and inferior divisions by the crista falciformis, a horizontal bony crest within the canal. The superior half of the canal is further divided into an anterior and a posterior compartment by a thin osseous septa referred to as Bill's bar. The facial nerve is located anterior to Bill's bar, and the superior division of the vestibular nerve courses in the posterior compartment. The four nerves classically described within the internal auditory canal are usually only delineated as individual nerves in the lateralmost portion of the canal.

The cranial nerves are unique because their nerve sheaths are formed by oligodendrocytes (glial cells) proximally as they exit the brain parenchyma. Within several millimeters the transition to myelin-forming Schwann cells occurs. For the vestibulocochlear nerve, this glial-Schwann cell junction occurs near the porus acusticus. Most schwannomas originate at this location, but they may occur anywhere along the course of the nerve myelinated by Schwann cells. Sensory nerves are more heavily myelinated than motor nerves. Therefore, these tumors tend to occur in association with sensory nerves such as the vestibular nerve. Trigeminal schwannomas are the second most common intracranial schwannoma accounting for about 5%, followed by facial schwannomas at a distant third. Histologically, schwannomas are composed of Antoni type A and type B tissues. Antoni type A tissue is composed of compact palisades of spindle cells while Antoni type B tissue is a loose myxoid stroma with few cells and regions of cyst formation. Most vestibular schwannomas consist of predominantly type A tissue, until their size increases and their cellularity decreases.

Neurofibromatosis type 2 (NF-2) is defined by bilateral acoustic neuromas. It is a neurocutaneous syndrome, or phacomatosis, that can be inherited in an autosomal dominant fashion or that occurs sporadically. In addition to the bilateral tumors of the internal auditory canal, NF-2 can be associated with multiple meningiomas, glial cell tumors, and other nerve sheath tumors. With the petrous bone. Because the tumor grows slowly, the bony canal may be remodeled and it may expand, particularly near the porus acusticus. This canalicular expansion is a late and unreliable radiologic sign because the width of the canal and degree of meatal flaring can vary considerably among normal patients. As the tumor enlarges, resulting areas of internal necrosis and hemorrhage can cause the formation of cysts, which appear as central regions of nonenhancement (Fig. 3). Large cerebellopontine angle tumors can cause significant mass effect and deformity of the brain stem and cerebellum. The ipsilateral cisterns widen and the contralateral cistern is compressed. Arachnoid cysts can develop when fluid is trapped because of the deformity and when adhesions form in the adjacent leptomeninges.

**T2-Weighted Imaging**

Magnetic resonance (MR) imaging is sensitive enough to define intracanalicular nerves for clinical purposes. Although contrast-enhanced T1-weighted imaging remains the most commonly employed method for evaluation, high-resolution T2-weighted imaging allows delineation of the individual nerves and evaluation of their relative sizes. When combined with knowledge of normal

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**Imaging**

**Tumor Characteristics**

Acoustic neuromas are well-circumscribed, enhancing lesions that typically arise near the porus acusticus. They are small when their location is intracanalicular. Intracanalicular schwannomas are ovoid or cylindrical-shaped and have a convex medial margin (Fig. 1). As the tumor grows, it often extends into the cerebellopontine angle cistern where it can expand unimpeded by the osseous confines of the internal auditory canal. This growth pattern creates an “ice cream on a cone” appearance with the cone representing the intracanalicular component (Fig. 2). The round cisternal component typically remains centered at the auditory meatus and forms acute angles

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**Figure 1. Axial T1-weighted image with contrast shows a cylindrical acoustic neuroma within the right internal auditory canal.**
anatomy, this information can also be used to detect tumor. Sagittal oblique imaging in a plane perpendicular to the long axis of the internal auditory canal provides a cross-sectional view of the canal and allows individual nerves to be depicted (Fig. 4). The high T2-weighted signal of the surrounding cerebrospinal fluid (CSF) provides excellent contrast between the relatively T2-weighted hypointensity of the nerves. T2-weighted fast spin-echo (FSE) MR imaging acquired on a 1.5 Tesla-strength magnet with thin-section thickness, a small field of view, and a large matrix (512 x 512) provides excellent anatomic detail. Direct two-dimensional (2D) acquisition in the axial, coronal, and sagittal oblique planes through the internal auditory canal is typical. Three-dimensional volume acquisition can be obtained as a single axial acquisition, and that data set can then be used for multiplanar reconstructed images. The high-resolution anatomic detail of these T2-weighted images not only detects nerve sheath tumors but also allows tumor volume and the degree of nerve involvement to be evaluated. The ability of high-resolution T2-weighted imaging to depict tumor involvement of individual nerves may provide prognostic information that can help preserve hearing in selected patients.15

Contrast-Enhanced T1-Weighted Imaging

Because acoustic neuromas enhance avidly, contrast-enhanced T1-weighted imaging continues to be used routinely to evaluate patients suspected of harboring tumor involving the internal auditory canal.10,16 Sections are typically 1.5 to 3.0 mm thick, allowing tumors as small as 0.05 cm³ to be detected.15 Because contrast enhancement provides the majority of the contrast distinction between tumor and normal brain parenchyma and, to some degree, between the surrounding CSF, the local anatomy and delineation of the nerves within the internal auditory canal are poorly seen on T1-weighted images. To reduce the cost associated with the administration of contrast material, high-resolution T2-weighted imaging has been advocated as an alternative to contrast-enhanced T1-weighted imaging for screening patients suspected of harboring a tumor involving the cerebellopontine angle or internal auditory canal.1,6 However, intravenous contrast administration is still necessary to evaluate other causes of hearing loss such as inflammatory, metastatic, or other idiopathic entities that cannot be detected reliably on noncontrast examinations.
Differential Considerations

An enhancing lesion within the internal auditory canal typically represents a vestibular schwannoma. Other lesions, however, can occur in this location and should be excluded. Although rare, a schwannoma arising from the facial nerve can appear identical to a vestibular schwannoma on contrast-enhanced T1-weighted imaging. The distinction between the two lesions can affect treatment and is reliably made by imaging if enhancement extends into the labyrinthine segment of the facial nerve canal within the temporal bone (Fig. 5). Meningeal processes such as meningiomas, inflammation (meningitis, sarcoid), or metastatic disease can also demonstrate intracanalicular enhancement and mimic vestibular schwannomas as the leptomeninges and dura extend into the internal auditory canal (Fig. 6).

In terms of cisternal vestibular schwannomas, meningiomas within the cerebellopontine angle cistern are probably the most common differential consideration. Both can appear as large, round, densely enhancing, extraaxial masses causing significant local mass effect. Meningiomas typically arise adjacent to the porus acusticus but sometimes extend into the internal auditory meatus itself. The presence of a “dural tail” or an obtuse angle with respect to the petrous bone can also help differentiate a meningioma from a schwannoma (Fig. 7). Other common cerebellopontine angle masses such as epidermoids and arachnoid cysts are easily distinguished by their hyperintense T2-weighted signal characteristics and lack of enhancement (Fig. 8). On postcontrast T1-weighted imaging, an intracanalicular lipoma can appear as an ovoid hyperintense mass that can be differentiated from a schwannoma on precontrast T1-weighted images (Fig. 9).
Posttherapeutic Imaging

The treatment options of vestibular schwannoma include observation, surgical resection, and stereotactic radiosurgery. Clinical observation may be reserved for older patients using periodic MR imaging to monitor growth of the tumor. Imaging also plays an important role in the follow-up of patients undergoing surgery and stereotactic radiosurgery.

Surgery

Postcontrast T1-weighted MR imaging is crucial during postoperative management. The pattern of enhancement within the internal auditory canal can help differentiate expected normal postoperative appearance from residual tumor. The rate of inadvertent subtotal resection is only 0.5 to 1.0%. Sometimes, however, small foci of tumor are intentionally left to preserve facial nerve function. These small foci of tumor usually enhance in a nodular pattern. Some form of postoperative enhancement within the internal auditory canal is common, if not uniformly present. Linear enhancement is almost always a normal finding associated with an uncomplicated acoustic neuroma resection. Nodular or mass-like enhancement within the internal auditory canal raises the possibility of residual tumor and should be followed. In the absence of radiation therapy, any enhancement pattern, including nodular and mass-like, that remains stable or decreases in size or degree of enhancement on follow-up examinations is unlikely to represent residual tumor.

The type of surgical approach must be considered when evaluating images of a patient who has undergone resection of an acoustic neuroma. Three surgical approaches are commonly used to access the cerebellopontine angle and internal auditory canal: the retrosigmoid, translabyrinthine, and middle fossa craniotomy. The retrosigmoid suboccipital approach can preserve hearing in appropriately selected patients because the labyrinth (cochlea, vestibule and semicircular canals) is left intact. The posterior wall of the internal auditory canal is drilled to expose the intracanalicular contents. However, the lateralmost aspect of the posterior wall is left intact to avoid damage to the posterior semicircular canal (Fig. 10). The fundus of the canal cannot be visualized during surgery and is therefore associated with the highest risk of unintentional tumor residual. After this type of approach, nodular or mass-like enhancement in the fundus merits close follow-up.

The middle fossa temporal approach also preserves hearing while allowing complete exposure of the fundus. This approach is usually reserved for patients with small tumors and near-normal hearing. Intense linear enhancement along the roof of the internal auditory canal is an expected postoperative finding because fascial grafts are typically used to close the bony defect (Fig. 11).

A translabyrinthine approach is reserved for large tumors when hearing
preservation is no longer a concern because residual hearing has been destroyed. The mastoid, vestibule, and posterior wall of the internal auditory canal are resected to obtain complete exposure of the internal auditory canal, including the fundus. The cochlea is preserved. Surgical fat grafts are used to fill the mastoidectomy cavity to prevent CSF leaks. The T1-weighted hyperintense fat graft necessitates the use of fat-saturation pulse sequences to evaluate the surgical bed and remaining internal auditory canal for underlying enhancement (Fig. 12). It is also important to recognize that the fat graft typically involutes with time and can become infiltrated with enhancing fibrous scar tissue.

Hyperintense precontrast T1-weighted signal within the labyrinth, particularly within the cochlea, can also be a normal postoperative finding. The hyperintensity is likely related to blood products introduced into the membranous labyrinth by traction on or transection of the vestibulocochlear nerve during surgery.19 Postoperative enhancement of the labyrinth is probably related to local irritants such as blood products. It may be difficult to delineate labyrinthine enhancement if enough blood products are present to result in a hyperintense signal on precontrast images. Postoperative MR imaging is also valuable for detecting other surgical complications such as infarcts, hemorrhage, parenchymal edema, and dural sinus thrombosis.

Stereotactic Radiosurgery

The rate of tumor control with radiation therapy ranges between 81 to 96%.8,11 Only a small percentage of tumors continues to enlarge. MR imaging can show different patterns of tumor behavior, which can help predict the long-term effects of treatment. As many as 40% of treated tumors undergo a period of temporary enlargement after Gamma knife radiosurgery.11 In these cases, the size of the tumor initially enlarges and peaks at one year. Sustained regression follows in the second year.
To determine if acoustic neuromas have stabilized or regressed, the lesions must be imaged during the first 2 to 3 years after radiation therapy.

The enhancement pattern of schwannomas also changes after radiosurgery. Most tumors lose enhancement centrally during the first several months (Fig. 13). This change is usually transient, and central enhancement recovers within about a year. Some studies indicate that this transient loss of enhancement is a good prognostic indicator of tumor regression.\(^7\) MR imaging also can be used perioperatively to evaluate for signal changes in the adjacent parenchyma. T2-weighted hyperintense signal changes can occur in the adjacent cerebellar peduncle and pons. These signal changes sometimes persist on long-term follow-up but do not significantly correlate with neurological deficits.\(^8\)

Summary

Acoustic neuromas, or vestibular schwannomas, are commonly encountered benign lesions that can result in profound hearing loss. Early detection and treatment can prevent progression of hearing loss and other neurologic deficits. MR imaging remains the most sensitive method of detection.

References


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Figure 11. (A) Coronal pre- and (B) postcontrast-enhanced T1-weighted images after a middle fossa approach. The precontrast image shows the temporal craniotomy and fat graft used to close the roof of the internal auditory canal. Postcontrast imaging with fat saturation shows only minimal linear enhancement within the lateral aspect of the auditory canal.

Figure 12. (A) Axial precontrast T1-weighted image after translabyrinthine resection. The fat graft within the mastoidectomy defect causes an extensive T1-weighted hyperintense signal that could mask foci of pathologic enhancement. (B) Postcontrast image with fat saturation allows peripheral linear enhancement within the surgical bed to be detected. Although the left cochlea is preserved with this surgical approach (B), hearing is not.

Figure 13. (A) A vestibular schwannoma before stereotactic radiotherapy enhances homogenously. (B) Eight months after treatment, the size of the tumor is unchanged while enhancement has decreased centrally. Such a change in enhancement is typically transient.


