Intralabyrinthine Schwannomas

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KEYWORDS

• Intralabyrinthine schwannoma • Vestibular schwannoma • Acoustic neuroma • Inner ear • Cochlea • Vestibule

KEY POINTS

- Most patients with intralabyrinthine schwannomas (ILS) will present with unilateral, severe sensorineural hearing loss as the primary indication to obtain an MRI.
- The differential diagnosis is predominately radiographic because hearing loss and vestibular symptoms are common to both ILS and other inner ear lesions.
- Interval MRI scanning and observation is currently performed for most patients with ILS, especially those without vestibular complaints.
- Surgical removal of ILS can be safely performed, and surgery may be increasingly indicated if cochlear implantation demonstrates effectiveness for unilateral hearing loss rehabilitation.

INTRODUCTION

Vestibular schwannomas are benign, slow-growing tumors that most commonly arise from the vestibular division of the eighth cranial nerve.[1,2] They account for approximately 80% to 90% of tumors within the cerebellopontine angle (CPA) and approximately 6% of all intracranial tumors.[3] Although there is ongoing debate over the specifics of tumor origin, most would agree that they arise from Schwann cells within the internal auditory canal (IAC). These progenitor Schwann cells are also found beyond the confines of the IAC in the more distal inner ear labyrinth.[4] Tumors arising in this area are far less common than the typical schwannoma involving the IAC. First described by Mayer[5] in 1917, the most recent systematic review of the literature revealed 234 intralabyrinthine schwannomas (ILS), which have now been described.[6]

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Before the advent of MRI, these tumors were found exclusively at the time of autopsy or as an unexpected discovery during neurotologic surgery for another reason, such as labyrinthectomy for Ménière disease.[7] Because of their rarity and presentation overlap with Ménière disease, diagnosis can be delayed. Visualizing these tumors is also a challenge because the MRI quality and sequences used are critical in avoiding a missed diagnosis.

ANATOMY AND PATHOGENESIS

Anatomy

The otic capsule is a dense section of bone within the petrous pyramid that houses the inner ear organs of hearing and balance. Collectively termed the membranous labyrinth, this includes the cochlea, semicircular canals, utricle, saccule, and endolymphatic duct and sac. The radiographic and surgical bony space that contains the utricle and saccule is called the vestibule. The bony labyrinth, containing perilymph, surrounds the more interior membranous portion, which contains endolymph. The neuroepithelium, which is responsible for this sensory system, is contained within the membranous portion.

The cochlea spirals 2 and a half times around a cone-shaped structure of porous bone called the modiolus, with the apex of the cone corresponding to the apex of the cochlea. The basilar membrane essentially separates the cochlea into a double-lumen tube made up of the scala tympani and scala vestibuli (Fig. 1). The scala tympani communicates with the middle ear near its basal turn via the round window. The scala tympani also connects to the posterior fossa via a potential space called the cochlear aqueduct, which, if abnormally enlarged, can be an open connection

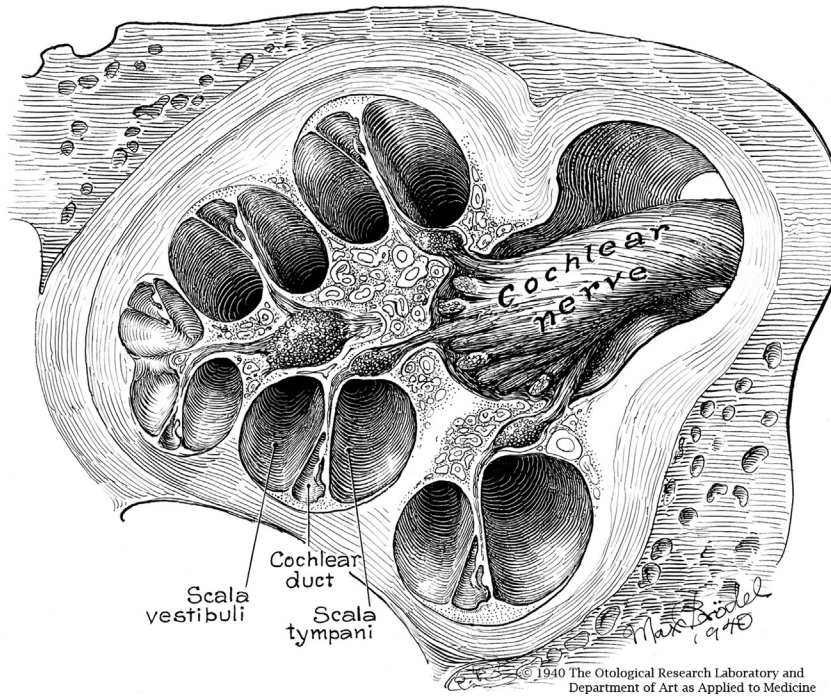

Fig. 1. Anatomy of the cochlea and cochlear scalae. (Original illustration #9987 in the Walters Collection of the Max Brödel Archives. Department of Art as Applied to Medicine, The Johns Hopkins University School of Medicine, Baltimore, MD, USA. Used with permission.)