Intralabyrinthine Schwannomas: Disease Presentation, Tumor Management, and Hearing Rehabilitation

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Abstract

Keywords

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- dizziness
- tinnitus
- hearing loss
- auditory rehabilitation
- inner ear masses

Intralabyrinthine schwannomas (ILS) are rare tumors that frequently cause sensorineural hearing loss. The development and increased use of magnetic resonance imaging in recent years have facilitated the diagnosis of these tumors that present with otherwise nondiscriminant symptoms such as tinnitus, vertigo, and hearing loss. The following is a review of the presentation, pathophysiology, imaging, and treatment with a focused discussion on auditory rehabilitation options of ILS.

Introduction

Primary intralabyrinthine schwannomas (ILS) are tumors of the terminal schwann cells of the cochlear or vestibular nerves and develop within the cochlea, vestibule, and/or semicircular canals. Mayer first described them in 1917 based on autopsy findings.^{1,2} Primary ILS are distinguished from secondary ILS, which develop from extension of a vestibular schwannoma (VS) through the modiolus or macula. ILS are mainly diagnosed on imaging, though a few reports have described incidental diagnoses during labyrinthectomy.^{3–5}

They are rare tumors, with only ~526 described in the literature to date and present with many of the same symptoms of other pathologic processes such as vertigo, dizziness,

tinnitus, and hearing loss.^{6,7} With the advent of magnetic resonance imaging (MRI) and improvements in imaging technology over time, diagnosis has become more prevalent.^{8–13}

These tumors can present an interesting therapeutic challenge given their benign nature, location, and symptomatology. In the following sections, we will review the biology, imaging characteristics, classification, and treatment options in patients with ILS.

Biology and Presentation of Intralabyrinthine/Intracochlear Schwannomas

ILS are thought to arise from the schwann cells surrounding the terminal branches of the cochlear and/or vestibular nerves inside the membranous labyrinth.¹⁴ They can be

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sporadic and isolated or they can present in neurofibromatosis type 2 (NF2) patients, the former being more common.^{2,15} Histopathologically, these tumors are identical to vestibular nerve schwannomas originating in the internal auditory canal (IAC) or cerebellopontine angle (CPA). They have Antoni A and B morphologic patterns and stain positive for S-100 immunoperoxidase.¹

In the largest series of ILS studied to date, 110 cases from 12 tertiary care referral centers were pooled. Of note, 87.3% of the patients were diagnosed after 2000. The average age was 53.9 years, with 47.3% male and 52.7% female. The average delay between the emergence of symptoms and diagnosis was 72.5 months.² No tumor was discovered incidentally.

The vast majority of patients (94.5%) experienced hearing loss at the time of diagnosis. Hearing loss in various studies has been described as progressive, sudden, or fluctuating.^{7,16–18} Hearing loss is mainly sensorineural; however, mixed hearing loss has been described and is thought to be from inner ear conductive loss caused by blockage of inner ear fluid movement by tumor.^{11,19} Tinnitus is the next most common presenting symptom with studies showing a range of 69.1 to 95.8% of patients with tinnitus, 10.5% being pulsatile tinnitus.^{1,2} Vestibular symptoms were present in 59.1%, ranging from dizziness, positional vertigo, or Meniere's like vertigo attacks. When present, dizziness can often be more acute and intense than the dizziness that occurs with CPA VS, and at times the vertigo can be intractable.^{2,16,18} Possible proposed explanations for this are more direct tumor contact with the vestibular epithelium in ILS or a type of hydrops may be occurring from tumor obstruction.¹⁶ Very rarely, patients can present with facial palsy and hemifacial spasm, with a few cases noted in the literature.^{16,20,21}

The majority of patients presented with 1995 American Academy of Otolaryngology—Head and Neck Surgery (AAO-HNS) class D hearing loss, 42.7%. A smaller number, 32.7%, of patients presented with class C hearing, and 21.8% were completely deaf. Only 8.2% patients were class A and 16.4% were class B. Among the patients with measurable hearing, the majority, 57%, had a flat audiogram configuration.² Abnormal vestibular testing has been found in ~65.7 to 77.8% of patients using a variety of vestibular tests.²

The large delay between the emergence of symptoms and diagnosis can be attributed to four main reasons per Dubernard et al 2014: (1) very small lesions can escape detection on imaging, (2) conventional diagnostic algorithms for sensorineural hearing loss (SNHL) and vertigo may not include high-resolution MRI, (3) symptoms can be subtle and vestibular problems are rarely acute and are frequently compensated, and (4) growth seems to be slow.² There have been some reports of patient presentations that have led to a misdiagnosis of Meniere's disease which could also contribute to a delay; however, patients that present with this symptomatology are the minority.¹¹

Studies have found that both ILS that are located in the cochlea and those that are located in the vestibule/semicircular canals can cause SNHL, though intravestibular lesions cause hearing loss at a lesser rate.² Intracochlear (IC) tumors are presumed to cause SNHL by either direct compression of

the organ of Corti and/or direct compression of the cochlear nerve or its vascular supply.^{14,22} There are a few theories regarding how intravestibular tumors cause hearing loss. One theory is through compression of the ductus reunions and the saccule which leads to endolymphatic hydrops. Histopathologic temporal bone studies have shown structural changes to the cochlea such as distention and collapse of Reissner's membrane.¹⁶

Another hypothesis is that both IC and intravestibular tumors may release toxins or potassium ions, thereby changing the metabolic status of the inner ear fluid; however, these metabolites have never been isolated.¹⁶ Biochemical degradation of inner ear fluid has been documented in cases of VS with hearing loss.^{14,23} Histopathologic studies have shown a combination of loss of hair cells, loss of peripheral processes (dendrites), reduction in spiral ganglion cells, and reduction in the stria vascularis. Pure impedance of fluid by mass effect of tumor also remains a possibility for causing hearing loss.⁶

In a study by Santos et al, 17 of 24 patients had more than one MRI for comparison of tumor growth.¹ The duration between the MRIs ranged from 6.3 to 101.7 months. Only 4 of 17 (23.5%) patients showed tumor progression to an additional location within the labyrinth or the IAC and the time to progression was, 6.8, 6.9, 22.8, and 96 months, respectively.¹ Tieleman et al, however, raises a question as to whether the studies investigating ILS tumor growth were specific to ILS or included some IAC VS with intralabyrinthine extension. In their study, they focused only on lesions that were in the membranous labyrinth at diagnosis and found that 59% of their 27 patients with follow-up imaging showed growth to adjacent structures in the inner ear.²⁴ Van Abel et al also noted tumor growth in 50/96 (52%) of patients with follow-up imaging.⁶

Imaging

ILS is mainly detected by MRI. Characteristics of ILS on MRI show focal enhancement on T1 after gadolinium contrast and hypointensity (filling defect) on heavily T2-weighted sequences in the same location (→ Fig. 1).^{13,16,24,25}

Recent imaging protocols described for ILS or VS screening include MRI with a 1.5T magnet and high-resolution three-dimensional (3D) fast spin echo or gradient echo T2-weighted imaging using thin cuts such as 0.8 to 1.0 mm.²⁶ Historically, a post-contrast T1-weighted image at the same time as the without non-contrast T2 was the gold standard in diagnosis of VS; however, recent studies evaluating cost-effectiveness have suggested that with further improvements in imaging technology, thinly sliced non-contrast T2 weighted imaging is sufficient for ruling out ILS, VS, and other CPA tumors. These studies support obtaining a post-contrast T1 scan only if there is a filling defect on T2, thereby reducing gadolinium exposure, acquisition time, and cost.²⁷ Similarly, thin slice heavily T2-weighted imaging is sufficient to evaluate for tumor stability or growth.

Other lesions that can have enhancement of the inner ear on T1 post-contrast MRI are labyrinthitis, labyrinthitis ossificans, hemorrhage, or lipoma. Acute labyrinthitis can be

Fig. 1 A right intravestibular schwannoma (white arrow) causing profound sensorineural hearing loss shows focal enhancement on post-gadolinium T1-weighted images.

differentiated by the T2-weighted MRI study which will show no filling defect; labyrinthitis also usually has much more diffuse enhancement.²⁸ Labyrinthitis ossificans can show enhancement of the inner ear during the fibro-osseous phase and will have decreased T2 signal intensity also in the inner ear; however, the key differentiating factor in these cases is the clinical history of prior meningitis or suppurative otomastoiditis. A computed tomography temporal bone may also show bony encroachment on the membranous labyrinth.^{28–30} Inner ear hemorrhage or lipoma has high signal intensity without enhancement on the pre-contrast T1-weighted imaging differentiating them from lesions consistent with ILS.^{28,31}

Classification

In 2004, Kennedy et al classified ILS into seven groups based on location within the inner ear:¹¹ (i) IC schwannomas which are seen as focally enhancing masses within the cochlea alone; (ii) intravestibular (IV) schwannomas are located in the vestibule and may or may not involve the semicircular canals; (iii) transmacular (TMA) schwannomas are also located in the vestibule but extend out through the macula cribrosa into the fundus of the IAC; (iv) transmodiolar (TMO) schwannomas have tumor within the inner ear that extends through the modulus into the fundus of the IAC; (v) transotic (TO) schwannomas are those that involve the cochlea, vestibule, IAC, and middle ear; (vi) intravestibulocochlear (IVC) schwannomas involve the vestibule and the cochlea; and (vii) tympanolabyrinthine schwannomas involve the middle ear, vestibule and cochlea.¹¹ In 2013, Van Abel et al modified the original Kennedy classification to include three more classes: (viii) translabyrinthine with tumors involving the vestibule, cochlea and IAC; (ix) ILS with extension into the CPA; and (x) tumors not otherwise specified.⁶

Various studies have shown that the IC schwannomas are more common than the other types, representing 50% or greater of cases.^{6,7} Intravestibular types and ILS that extend outside of the labyrinth make up ~19% and mixed vestibulocochlear types comprise 11.8%. TO cases are the least common, with 2 of 45 (4.4%) cases reported in one study.¹³

Treatment

In general, patients with ILS and serviceable hearing are treated with observation since the lesion is benign, a significant number shows no growth, and surgical removal leads to complete hearing loss in the operated ear.³² Only one case in the literature describes a case of a schwannoma in the lateral and superior semicircular canals that had a surprising improvement in hearing after excision of tumor.¹⁷ In cases of intractable vertigo or tinnitus or further extension of tumor to the CPA even with serviceable hearing, intervention with surgical resection or stereotactic radiation may be more strongly considered.^{11,32} Intractable vertigo can also initially be treated with intratympanic gentamycin; however, this is not always effective. Previous cases report on the resolution of vestibular symptoms after surgical removal of ILS; four patients who underwent surgery for vertigo had complete resolution of their vertigo in the 2004 study by Kennedy et al and two patients had complete resolution of vertigo postoperatively in the 2003 study by Neff et al.^{11,32}

Given the rarity of ILS, there are limited studies comparing treatment algorithms. Gosselin et al performed a meta-analysis on the symptomatic evolution of patients following observation or surgical excision; 66 patients were included with a mean follow-up of 2.4 years.³³ Their study exhibited significant selection bias given those patients that had intractable vertigo or tinnitus preferentially underwent surgical excision. The authors reported that 50% of the surgical excision cases showed an improvement in tinnitus, 42.9% had unchanged tinnitus, and 7.14% had worsened tinnitus. None of the observation group patients had improved tinnitus over time, 87.1% reported no change, and 12.9% noted worsening. Vertigo symptoms were improved in 100% of surgical cases, whereas only 22.2% of observation cases reported improvement. Hearing loss was noted to progress in 27.3% of observation patients and 70.5% had stable hearing loss. All operated patients, as expected, had anacusis. The authors concluded that surgical excision has a low incidence of complications,^{16,34} and that significant improvement in vertigo and dizziness, as well as tinnitus though to a lesser degree, could be expected following surgical excision in patients with nonserviceable hearing.³³

Patients who elect observation can still be counseled on auditory rehabilitative options such as hearing aids, osseointegrated implants, and cochlear implants.¹ Currently, there is no set standard for MRI intervals for patients who have elected observation—however, a schedule similar to that used for VS seems reasonable. It has been suggested that post-contrast MRI may not be necessary in these follow-up scans since tumor progression may be adequately assessed by the high-resolution, thin slice 3D T2-weighted images.³⁵

Excluding the previously mentioned indications for surgical resection, there is some debate on when to intervene for patients with nonserviceable hearing and a growing tumor that does not yet extend into the middle ear, IAC, or CPA. Arguments have been made for intervening early before extension into compartments that risk a poorer long-term outcome. However, there are some reports of lesions being followed with some initial small growth but eventual stability in the size of the lesion on further follow-up imaging. Generating a treatment plan should take into account the location of the lesion, hearing status, symptoms, tumor growth, age, and patient's wishes.

Many different techniques for surgical resection have been described. An approach that is tailored to the lesion and allows for access to complete tumor removal is preferred. Santos et al describe removing these tumors with a traditional translabyrinthine-TO craniotomy with abdominal fat grafting and blind sac closure of the external auditory canal (EAC). This extensive type of approach may be necessary for IVC lesions, TO lesions, or TMO lesions depending on extension. Other approaches for smaller lesions include transmastoid or transcanal labyrinthectomy for IV lesions and TO approach for IC lesions.

Endoscopic approaches have also been used for ILS. Marchioni et al describe the transcanal transpromontorial endoscopic approach (TTEA) and the enlarged transcanal transpromontorial endoscopic approach (ETTA) which are applied when the tumor extends to the CPA. In their series, six out of eight patients underwent TTEA, five were IC schwannomas, and one was a IVC schwannoma. The TTEA is performed via the EAC, after raising a tympanomeatal flap. The ossicles are removed including the stapes, and the basal, middle, and apical turns of the cochlea are opened giving access to the tumor. When further dissection was needed for the ETTA approach, a blind sac of the EAC was performed.^{18,36}

Hearing Rehabilitation

Hearing status in the ipsilateral and contralateral ear as well as auditory rehabilitation options are among several important factors that may influence treatment. The type of surgical approach does not matter for auditory rehabilitation options such as the CROS hearing aid or bone conduction devices. However, another rehabilitation option that is gaining favor is cochlear implantation (CI).

Traditionally, CI is done only in cases of significant bilateral hearing loss; however, there is growing literature that supports the benefits of CI in patients with single-sided deafness (SSD).³⁷⁻⁴¹ The majority of ILS patients present with SSD or AAO-HNS Class C or D hearing in the ipsilateral ear only.^{1,2,17} In these situations, if the patient is undergoing surgical removal for tumor growth or intractable vertigo, CI for auditory rehabilitation could be offered. Given the intralabyrinthine location of these tumors and the need for manipulation of the inner ear for removal, CI in cases thus far has either been performed concomitantly with tumor removal or without tumor excision. Less commonly, a dummy electrode carrier (insertion test device, off label use) has been inserted and left in place to keep the cochlea patent for future replacement with a CI if the patient desired.⁴² When considering CI in this population,

there are a few things to consider surgically. CIs activate the distal auditory nerve branches, and ILS are thought to arise from the schwann cells at the distal aspect of these nerves. Therefore, damage to these nerves during tumor removal would result in poor CI performance outcomes. In IV and TMA tumors, where the cochlea is not involved, a simple translabyrinthine approach allows excellent access to the tumor for removal without much concern for damaging structures important for CI performance and a CI can be placed in the standard fashion (► Fig. 2).

In all the other presentations of ILS, however, the cochlea as well as other structures are involved by tumor. The goal in these cases is to remove the tumor in its entirety, while maintaining the anatomic structure of the cochlea and the spiral ganglion cells intact. Many of the previously described approaches destroy the cochlea for IC tumors, thereby removing the option for CI. Complete tumor removal with preservation of distal auditory nerve structures is likely not possible in TMO tumors. Previous reports have described placing the cochlear implant through tumor in these cases.⁴³ Initial outcomes in these patients have been very promising with 8 out of 10 patients achieving median consonant-nucleus-consonant scores of 50% and median AzBio scores of 73%. Postoperative surveillance was also shown to be possible in these patients with the CI magnet in place.⁴³

Other initial reports on CI in patients with ILS show promising results regarding auditory rehabilitation as well.^{35,43-46} Plontke et al described 12 cases of ILS in which five patients underwent CI at the time of their tumor removal. Two other cases had a dummy electrode placed at the time of surgical removal for possible replacement with a CI in the future. Three of the patients had IV schwannomas and underwent labyrinthectomy and standard cochlear implant insertion. Two of the patients had IC tumors, one had a small lesion at the basal turn that could be removed via extended cochleostomy, and CI insertion was performed through this opening after tumor removal. The second had more extensive involvement of the cochlea and underwent a subtotal cochleostomy with CI insertion and reconstruction of the lateral cochlear wall. In the former, a cochlear CI24RE (CA) was used and in the latter a cochlear CI 512 (CA) was used. In this group, as with CI outcomes in the general population, there was variability in outcomes at 12 months post-surgery. Word recognition scores at 6 and 12 months were close to 100% using the Freiburg multisyllables test for all of the IV tumor cases and for the IC case that was limited to the basal turn. Using the Freiburg monosyllables test, the basal turn IC schwannoma case scored in the low 90% and the others scored in the 30 to 55% range. The poorest outcome was in the IC schwannoma case that underwent subtotal cochleostomy. Six months data in this case showed only an ~30% correct in the Freiburg multisyllables and 0% in the monosyllables.³⁵ These results show that CI is a viable option for many ILS cases. However, preservation of the cochlea and neuronal targets for the CI will likely prove to be important in CI outcomes for these patients.

When discussing auditory rehabilitative options with a patient and CI, device choice should take into account a few considerations in ILS cases in addition to patient preference,

Fig. 2 (A) Left-sided intravestibular schwannoma in a patient with nonserviceable hearing and intractable vertigo. The patient underwent a left-sided transmastoid labyrinthectomy. (B) The tumor can be seen within the vestibule. (C and D) Following tumor removal (C, arrow), the undisturbed patent cochlear duct (D, arrowheads) and cochlear modiolus (D, arrow) can be visualized using rigid endoscopy.

including (1) future surveillance needs of the patient, (2) the possible need to place the implant through tumor, and (3) possible advantages of a pre-curved electrode. With regard to surveillance, the majority of ILS cases are sporadic and though solitary lesions are less likely to have clinically relevant recurrence, they will require some imaging surveillance. In cases of NF2 and cases where the CI is placed through tumor, placement of the receiver-stimulator to minimize artifact and surveil the ipsilateral CPA and IAC becomes important (► **Fig. 3**).^{43,47} Some CI manufacturers also produce devices with improved MRI compatibility, including magnet systems that align within the MRIs magnetic field that may also factor into device choice.

If the implant is placed through tumor, the use of a styleted electrode with delayed release of the stylet has been reported to assist insertion of the electrode past the mass.⁴³ And lastly, precurved modular hugging electrodes have been described to be beneficial in cases of partial or subtotal cochleoectomy.⁴²

Similar to CPA VS, radiosurgery is also an option for the treatment of ILS. This option appears to be uncommonly considered however, because the majority of the ILS are easily accessible for full tumor removal without having to perform an intracranial approach.³⁵ There is also the potential added benefit of performing a cochlear implant at the time of tumor excision.

Fig. 3 In this neurofibromatosis type 2 patient observing a small left intracanalicular schwannoma after resection of a right intracochlear schwannoma and cochlear implantation with a magnet in place, the contralateral lesion (white arrow) is well seen on coronal, T1-weighted post-gadolinium images (A) and with device artifact still allowing views of both internal auditory canals and cerebellopontine angles on heavily T2-weighted images (B).

Conclusion

ILS are rare tumors that are frequently diagnosed years after first symptom onset. This prolonged diagnostic delay points to the possible need for more awareness of this pathology and inclusion in the differential diagnosis of patients that present with SNHL and/or vertigo. Appropriate imaging protocols for screening and careful scrutiny of the imaging must also be stressed as these lesions can be small and easily missed.

Once diagnosed, careful consideration of lesion size, location, symptoms, hearing status, and patient desires must be weighed to form an optimal and tailored therapeutic treatment plan. CI, either with schwannoma left in situ or at the time of tumor excision, is a feasible option for hearing rehabilitation in these cases with promising auditory performance.

Conflict of Interest

None.

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