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Review Article

Radiology Section

Intralabyrinthine Schwannomas: Review of Anatomy, Pathology, Clinical Features from an Imaging Perspective

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INTRODUCTION

Intralabyrinthine schwannomas (ILSs) are primary neoplasms of the vestibulocochlear nerve that arise de novo in the cochlea, vestibule and the semi-circular canal, or in a combination of these structures [1,2]. It represents a rare entity with a prevalence of 0.1% to 1% in autopsy reviews [3]. ILSs have also been shown to cross into both the middle ear and the internal auditory canal [4]. Prior to high-resolution MRI of the inner ear, ILS was commonly diagnosed as an incidental finding during labyrinthectomy or autopsy, with the first case being reported in 1917 [5]. In 1972, Wanamaker discovered a case of ILS at the time of labyrinthectomy in a patient who was presumed to have Menier's disease [6]. In the same year, Karlan reported the first patient with the identification of ILS by using computed tomography [7]. It was not until 1994, that the utility of contrast enhanced MRI was realized in the diagnosis of ILS. With the advent of high-resolution T2 weighted imaging with gadolinium contrast-enhanced T1 weighted imaging, ILSs are being diagnosed in patients as incidental findings, as well as on dedicated imaging in the workup of patients with senorineural hearing loss and vertigo [4,8,9]. A recent retrospective review has shown that up to 10% of all the vestibulocochlear schwannomas are intralabryinthine, and that they are most frequently found in the cochlea. However, this may slightly overestimate the actual prevalence, as the study took place a referral centre [10]. Additionally, this same study also demonstrated that these lesions can grow over time, often crossing into anatomically distinct portions of the inner ear, as well as into the internal auditory canal. The management of ILS typically consists of serial observations. Surgical excision is recommended for intractable vertigo, for the extension of an ILS into the cerebellopontine angle cistern, or for any evidence of tumour growth in a patient who is fit for surgery [11,12]. Thus, a working knowledge of the anatomy of the cochlea, vestibule and internal auditory canal are critically important when these neoplasms are being evaluated. While ILS does have a typical appearance on MRI with intravenous gadolinium contrast injection, the mimics to ILS do often present on MR imaging. In the present review, we will discuss the ILSs from an imaging perspective, particularly correlating them with their anatomical classifications and clinical features.

IMAGING PROTOCOLS

MR images the endolymph and the perilymph within the membranous labyrinth via high-resolution T2 weighted images, as opposed to CT, which images the bony labyrinth with high-resolution axial images. Most of these sequences are based on either a constructive interference in the steady state (CISS) or on the fast spin-echo (FSE) technique. Each technique has inherent shortcomings when it images the small structures of the membranous labyrinth. The balanced- steady state free precession sequences which are often used, suffer from a susceptibility artifact at the air-bone and the bone–soft tissue interfaces. The artifact is accentuated at the skull base and with an imaging at 3T. This artifact can be mitigated with CISS, which sums the successive phase-cycled acquisitions which are reconstructed with a maximum intensity projection [13]. However, the interface related artifacts remain problematic with the very small field of views which is required for temporal bone imaging and minute portions of the membranous labyrinth. The FSE sequences are limited by image blurring, which is secondary to a higher noise to signal ratio, from the shorter imaging times which is accentuated at 3T [13].

The 3D variable flip-angle FSE technique has been recently described as a technique which reduces both the susceptibility artifacts and image blurring [14]. In contrast to the standard FSE, where the flip angle remains constant (eg, 180°), the flip angle of the refocusing pulses in 3D VFA FSE is varied to achieve a desired image contrast. This technique allows longer echo trains and it reduces the change in the signal intensity through the echo train. This results in a higher signal to noise ratio, allowing high-resolution single slab isotropic acquisition. The image blurring is also reduced by the consistent echo-to-echo signal intensity which is particularly problematic in tissues with a relatively short T2. Finally, the smaller flip angles allow this technique to be utilized at 3T with a reduced radiofrequency heating, as can be measured by the specific absorption rate (SAR) as compared to the standard FSE technique [15].

THE IMAGING ANATOMY OF THE LABYRINTH AND THE CLASSIFICATION OF ILS

The classification of ILS is primarily based upon which anatomic portion of the membranous labyrinth is occupied by the lesion. This classification is shown in [Table/Fig-1] [4]. In order to properly evaluate and characterize ILS, a basic working knowledge of the membranous labyrinth is required. The membranous labyrinth which is enclosed in a bony labyrinth within the petrous portion of the temporal bone, is a continuous structure which consists of the cochlea and the vestibule. The membranous labyrinth contains endolymph and it is surrounded by perilymph. The perilymph can be formed from an ultrafiltrate of blood or cerebrospinal fluid. The vestibule can then be further divided into the utricle, saccule, and the three semicircular canals (horizontal, superior and posterior). The utricle lies in the elliptical recess of the medial wall of the vestibule and it contains five openings form the semicircular canals, as the superior and the posterior canals share a common crus. The vestibule communicates with the fundus of the internal auditory canal via the macula cribrosa [23]. The cochlea contains two and a half turns with an opening into the fundus of the internal auditory canal, which is known as the cochlear aperture via the modiolus. The cochlea communicates with the vestibule via the ductus reuniens to the cochlear recess of the vestibule. The cochlear nerve communicates directly with the internal auditory canal through the cochlear niche. The inner structure of the cochlea can be further divided into three spiral chambers: the scala tympani, scala vestibule, and the scala media. The cochlear nerve travels in closer proximity to the scala tympani, in the region where it leaves the osseus spiral lamina which is called the habenula perforate, which runs towards the organ of Corti. The anterior saccular port of the vestibule has an anatomic connection with the scala vestibule. The scala tympani ends at the round window and it does not have a connection with the saccule [23]. Recent studies have demonstrated in exquisite detail, the ability to visualize the anatomy of the inner ear at both 3T and 9.4T [15]. ILSs can extend from one seemingly anatomically distinct portion of the middle ear to another, or even the IAC or the middle ear cavity. This is possible, given the anatomic communications between the distinct parts. The previously mentioned routes provide potential routes of spread as the ILS grows. An understanding of these anatomic connections can help in clarifying the classification system of ILS and in identifying the predictable growth patterns.

THE IMAGING FEATURES OF ILS

ILSs have a typical imaging appearance on gadolinium contrast-enhanced MRI. The typical appearance is a T2 hypointense, sharply circumscribed filling defect which is seen on the high-resolution T2 weighted images, which displaces the normal high T2 signal from the intralabryinthine endolymphatic and perilymphatic fluids. The lesion also demonstrates enhancement on the post-contrast T1 weighted images [3,16]. The region of enhancement on the T1-weighted images should correlate with the T2 filling defect on the high-resolution images to ensure that the enhancement does in fact represent a space-occupying lesion rather than an infectious, inflammatory or a haemorrhagic aetiology [10]. As has been discussed later, the sharply circumscribed margins set apart ILS from other intralabryinthine entities. In the same retrospective case series, they found an increased, unenhanced T1-weighted signal within a predominate number of lesions. However, the level of the increased, unenhanced T1-weighted signal depends on the size of the tumour, with the smaller tumours demonstrating no significant abnormal signs due to the volume averaging. An increase in the T1 signal is nonspecific; however, it can be used to further confirm the diagnosis. It is important to note the extent of the enhancement, as it relates to the normal anatomy of the IAC, cochlea, labyrinth and the middle ear cavity. Imaging in both the axial and the coronal planes can help in delineating the extent of the tumour, as shown in [Table/Fig-1].

Classification	Space that is occupied by the lesion
Intravestibular	Within the vestibule of the inner ear
Intracochlear	Within the cochlea
Vestibulocochlear	Involving both the vestibule and the cochlea
Transmodiolar	Crossing the modiolus from the cochlea to the fundus of the IAC
Transmacular	Crossing from vestibule to fundus of IAC
Transotic	Crossing the inner ear from the fundus of the IAC to middle ear

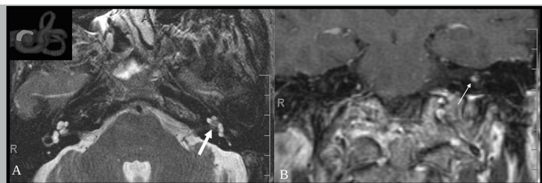
[Table/Fig-1]: Classification help in delineating the extent of the tumour

ILSs can range from simple to complex forms. The simplest form of ILS is intracochlear schwannoma. [Table/Fig-2] demonstrates an intracochlear schwannoma as an enhancing lesion in the basal turn of the cochlea, with a corresponding T2 filling defect on the high-resolution axial images. [Table/Fig-3] shows a variant of an intracochlear schwannoma, affecting only the middle turn of the cochlea. Tielman et al., [10] reported in their series, that eighty percent of the ILSs were confined to the cochlea. Of these, half of them were situated near the transition between the basal and the second turn. Based on the high-resolution T2-weighted imaging, the lesions were further characterized to always involve the scala tympani with some demonstrating extension to the scala vestibuli, which concurred with the above mentioned anatomy.

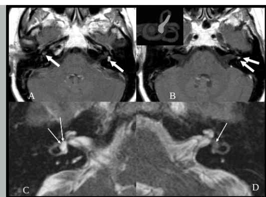
The utility of post-contrast imaging in the two planes is demonstrated in [Table/Fig-4]. The axial T1 post-contrast image easily demonstrates the enhancement of the right vestibule, but the coronal T1-weighted images clearly show the enhancement extending into the superior semi-circular canals. Teilman at al. reported the vestibular involvement to be far less frequent and to represent about 13.5% in their series. They noted that most of the intravestibular schwannomas typically involved the vestibule and the superior SCC. Exclusive involvement of either remaining SCC in addition to the vestibule is much less common.

[Table/Fig-5] demonstrates a vestibulocochlear schwannoma with abnormal enhancement, which involved the cochlea and extended into the superior and the lateral SCCs. A typical growth pattern for a vestibulocochlear schwannoma is having its origin in the cochlea within the scala tympani with an extension into the scala vestibuli. Once the intracochlear space has been completely occupied, the disease extends via the perilymph communication from the scala vestibule to the anterior saccular part of the vestibule. Vestibulocochlear schwannomas have a tendency to involve the superior SCC, because their spread in the vestibule involves the anterior most portion which is nearer to the opening of the superior SCC [10]. This pattern of growth also explains the extension of

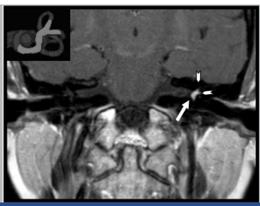
[Table/Fig-2]: Patient with left sensorineural hearing loss. Axial T2-wt images, (A) right side showing normal fluid signal in the basal turn of cochlea (arrow) where as (B) crescenteric hypointense lesion along the basal turn of left cochlea partially replacing the normal fluid signal and (C) post-contrast axial T1-wt image showing enhancement of the intracochlear schwannoma.



[Table/Fig-3]: 66 years old male with progressive left-sided hearing loss. Axial T2 wt image (A) shows a filling defect in the middle turn of the left cochlea. Coronal post-contrast T1 wt image (B) shows enhancement in the area corresponding to the filling defect.



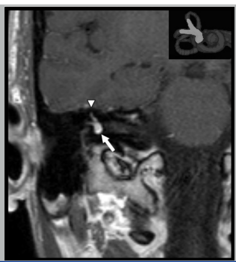
[Table/Fig-4]: Patient with bilateral tinnitus, greater on the left side. (A, B) Post-contrast T1-wt axial images show enhancing lesion involving the left vestibule (A, arrow) extending into the superior semicircular canal (B, double arrow). Small enhancing lesion also seen in the right cochlea (A, arrowhead). (C) Normal fluid signal in the vestibule on right side (double thin arrows) and (D) replacement of normal fluid signal in the left vestibule (single thin arrow) corresponding to enhancing lesion seen in (A, B).



[Table/Fig-5]: 52 years old female presented with left sensorineural hearing loss. Coronal T1-wt image showing enhancement of within the left vestibule extending into the cochlea (arrow) as well as into the semicircular canals (arrowheads).

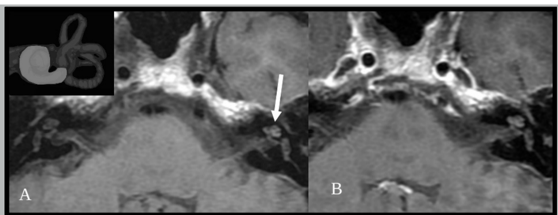
the schwannomas which originate in the vestibule, which will first involve the scala vestibule prior to their extension into the scala tympani. An understanding of this growth pattern can increase the reader's sensitivity for detecting their growth and extension. [Table/Fig-6] demonstrates another example of vestibulocochlear schwannomas with involvement of the superior and the horizontal SCCs.

[Table/Fig-7] demonstrates a transmodiolar schwannoma with a T1-weighted image which shows enhancement in the IAC and extension into the middle turn of the cochlea.

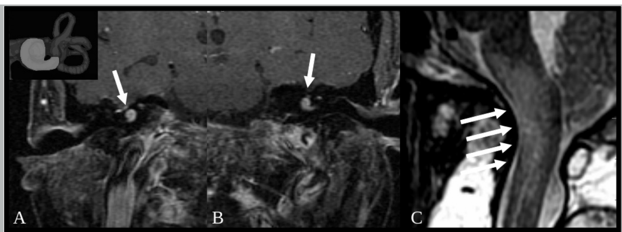


[Table/Fig-6]: 54 years old male presented with sudden right sensorineural hearing loss. Coronal T1-wt image showing enhancement of within the right vestibule (arrow) extending into the semicircular canals (arrowheads.

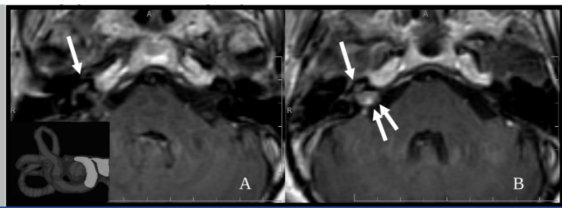
[Table/Fig-7]: 62 years old female with a history of neurofibromatosis type 1. Axial T1-wt image showing enhancement in the IAC (arrow) extending into the middle turn of the cochlea (arrowhead), a transmodiolar schwannoma. Additional meningioma is also seen in the right posterior fossa.



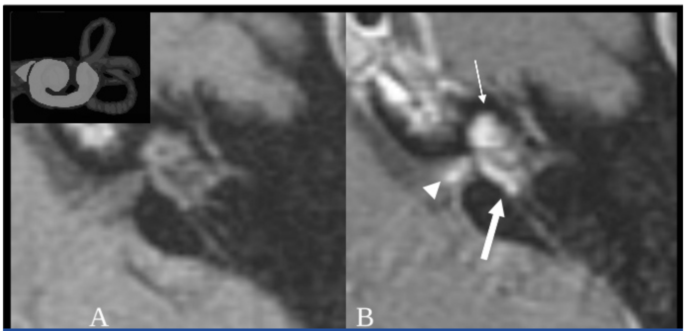
[Table/Fig-8]: 60 years old female presented with a sudden history of left hearing loss and roaring sensation. Axial pre contrast T1 wt images (A) show increased signal in the left cochlea as compared to the right. No enhancement is seen on the post contrast images (B). These findings are consistent with hemorrhagic products from hemorrhagic labyrinthitis.



[Table/Fig-9]: Patient with metaastatic breast cancer status post stereotactic radiation therapy presenting with bilateral sensorineural hearing loss and unsteadiness. Post contrast coronal images (A & B) show enhancement of the bilateral cochlea consistent with radiation-induced labyrinthitis (arrow). T2 sagittal image (C) shows well-defined hyperintense signal involving the brainstem and at the cervicomedullary junction (short arrows) representing radiation injury conforming to the radiation portal.



[Table/Fig-10]: Patient with right hearing loss, tinnitus and imbalance. Pre (A) and post contrast (B) T1 wt MR showed an enhancing lesion involving right cochlea and vestibule (single arrows) and extending into IAC (double arrow), mimicking a transmodiolar transmacular schwannoma, however, surgical resection revealed an undifferentiated carcinoma metastases from a previously undiagnosed lung cancer.



[Table/Fig-11]: Patient with tinnitus and subjective hearing loss in the left ear. Pre (A) and post contrast (B) axial T1 wt images show an enhancing lesion involving the cochlea (thick arrow) , vestibule (thin arrow) and fundus of the IAC (arrowhead). The patient underwent surgical resection, which showed meningioma. Used with Permission. Aho, TR et. Al. Intralabyrinthine Meningioma. American Journal of Neuroradiology 24:1642-1645, September 2003.

MIMICS OF ILS

Not all enhancing lesions within the membranous labyrinth represent ILS. Unfortunately, the symptoms of the mimics of ILS are similar, which include hearing loss and vertigo. The imaging characteristics as well as the patient history can be used to differentiate some of the mimics from ILS, but surgical excision and pathological studies are often needed to make the final diagnosis.

Labyrinthitis is a non-specific term for the inflammation of the membranous labyrinth. The common aetiologies include spontaneous haemorrhage and bacterial and viral infections [11]. Haemorrhagic labyrinthitis demonstrates an abnormal high T1 signal with a normal increased T2 signal, without contrast enhancement [17].The patients with haemorrhagic labyrinthitis often present with a sudden hearing loss or a roaring or "whooshing" sensation. [Table/Fig-8] shows haemorrhagic labyrinthitis as an increased T1 signal without an enhancement or a space occupying lesion on the T2 weighted images. Haemorrhagic labyrinthitis can also be a sequela of radiation therapy. An increased T2 signal in the brain, in the radiation port, can be seen along with the typical findings of haemorrhagic labyrinthitis, as has been demonstrated in [Table/Fig-9]. Viral labyrinthitis will show a normal, non-contrast T1 signal without contrast, but an enhancement of the labyrinth without a T2 filling defect [11].

The other obvious aetiologies of the mimics for ILS include enhancing masses that are not primary to the inner ear. Metastases to the inner ear have previously been reported, including lung carcinoma [18]. The enhancing lesion in [Table/Fig-10] was originally suspected to be a transmodiolar transmacular ILS. However, on biopsy, the pathology returned as undifferentiated adenocarcinoma. Upon additional imaging of the chest, abdomen and the pelvis, an adenocarcinoma of the lung was identified. Other metastases have been reported to involve the internal auditory canal, which include metastatic rhabdomyosarcoma, malignant melanoma, and carcinoma of the breast [19, 20, 21].

[Table/Fig-11] demonstrates a rare case of an intralabrynthine meningioma which has features that are entirely indistinguishable from those of a schwanomma. The images were reproduced with the author's permission [22]. The T1 weighted images show an enhancing lesion which involved the cochlea (thick arrow), vestibule (thin arrow) and the fundus of the IAC [23].

CONCLUSIONS

ILS is a rare primary tumour of the inner ear that typically presents with sensorineural hearing loss with or without vertigo. These lesions are typically followed with serial imaging exams to ensure their stability. A careful examination of the patient's history, as well as inspection of the post-contrast T1 weighted images can lead the reader to the diagnosis of ILS. Care should be taken to verify a T2 filling defect on the high-resolution images, to ensure that one is not diagnosing one of the common mimics as a benign aetiology such as ILS.

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