PHILIPPINE JOURNAL OF OTOLARYNGOLOGY-HEAD AND NECK SURGERY VOL. 40 NO. 2 JULY – DECEMBER 2025

A Case of Transmodiolar Intralabyrinthine Schwannoma (ILS) in a 51-Year-Old Woman

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ABSTRACT

Objective: To highlight the clinical presentation, diagnostic dilemma and proposed management of a case of a transmodiolar intralabyrinthine schwannoma.

Methods:

Design: Case Report
Setting: Tertiary National University Hospital
Patient: One

Results: A 51-year-old woman consulted for a 12-year history of unilateral sensorineural hearing loss and progressively worsening vertigo. Workups revealed a hyperintense lesion approximating the entire right cochlea on Gadolinium-enhanced magnetic resonance imaging and profound hearing loss on the right on pure-tone audiometry, with no recordable response even at maximum intensity levels across all frequencies. Infectious and metastatic workups returned negative results, clinching the radiographic diagnosis of an intralabyrinthine schwannoma.

Conclusion: Intralabyrinthine schwannomas (ILS) are rare, benign neoplasms of the 8th cranial nerve, manifesting as progressive unilateral sensorineural hearing loss and worsening vertigo. These neoplasms are mainly detected by MRI; underdiagnosis and lack of standard treatment and diagnostic protocols result in their inaccurate diagnosis and delays in management.

Keywords: *intralabyrinthine; schwannoma; transmodiolar, intracochlear, vestibular, labyrinthine*

Intralabyrinthine schwannomas (ILS) are benign tumors of the 8th cranial nerve arising and located primarily within the membranous labyrinth.[1] They are rare lesions but the advent of improved imaging modalities now allows higher detection rates.[2,3,4] However, there is a lack of awareness of guidelines for their diagnosis and appropriate management has yet to be established. We report the case of a 51-year-old woman presenting with hearing loss and progressively worsening vertigo where imaging revealed an intracochlear lesion with extension into the internal auditory canal and discuss our diagnosis of an ILS.

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Disclosures: The authors signed a disclosure that there are no financial or other (including personal) relationships, intellectual passion, political or religious beliefs, and institutional affiliations that might lead to a conflict of interest.

Presented at the Philippine Society of Otolaryngology-Head and Neck Surgery Interesting Case Contest (2nd Place), Oriental Plaza Hotel, Legazpi City, Albay, May 29, 2025.

CASE REPORT

A 51-year-old woman consulted at our institution with a 12-year history of progressively worsening rotatory dizziness, associated with nausea, vomiting, and onset of unilateral hearing loss on the right. She initially consulted an otolaryngologist who ordered magnetic resonance imaging (MRI), suspecting an acoustic neuroma. However, upon review of initial imaging by a radiologist, no mass was supposedly appreciated. She was prescribed a course of oral steroids and betahistine 16mg/tab 3x a day which relieved her symptoms. She reported improvement of dizziness, claiming to only experience intermittent episodes that she was able to tolerate without intake of medications. She also reported decreased frequency of nausea, vomiting and bouts of headaches. During this time, however, the patient reported progression of her hearing loss, eventually resulting in total hearing loss of the right ear one year prior to initial consultation. Eventually, persistence of these symptoms and progressing severity of vertiginous attacks now unrelieved by betahistine intake prompted further consultation in our institution.

It is also important to note that during this time between consultations, she suffered two episodes of cerebrovascular infarcts in the right middle cerebral artery secondary to chronic uncontrolled hypertension, presenting as left-sided weakness. She was managed conservatively with antiplatelets, antihypertensives, and discharged with subtle left sided upper extremity weakness. She was also diagnosed with Stage IIIB cervical cancer, initially presenting as profuse vaginal bleeding for which she underwent pelvic external beam radiotherapy of 4500 cGy to the pelvic region, six cycles of Cisplatin therapy, and high dose-rate brachytherapy of 2800 cGy. All these were completed one year prior to her initial consultation at our outpatient department.

On our initial physical exam, tuning fork tests revealed sensorineural hearing loss. There was no appreciable sound perceived by the patient in her right ear with her left ear having normal hearing. Neurotologic examination revealed a positive right head impulse test, a positive Romberg test, and an abnormal tandem gait test.

Repeat MRI revealed no enhancing lesion seen in both 7th-8th nerve complexes to suggest the presence of chronic neuroma/schwannoma. However, there was a noticeable enhancing lesion on T1 weighted imaging, approximating the entire right cochlea with extension to the internal auditory canal fundus on Gd-enhanced MRI. This lesion was also noted to exhibit signal loss on T2 weighted imaging, pointing toward a neoplastic or inflammatory etiology. Pure tone audiometry with speech testing (PTA-ST) revealed profound hearing loss on the right, with no recordable response even at maximum intensity levels across all frequencies. *(Figure 1)*

Due to her previously diagnosed cervical malignancy and the timing of onset of progression of vertigo severity, cerebrospinal fluid studies (CSF) were performed to rule out a metastatic cause of the intralabyrinthine lesion seen on MRI. However, CSF protein and glucose levels were normal, and CSF cytology did not reveal the presence of malignant cells.

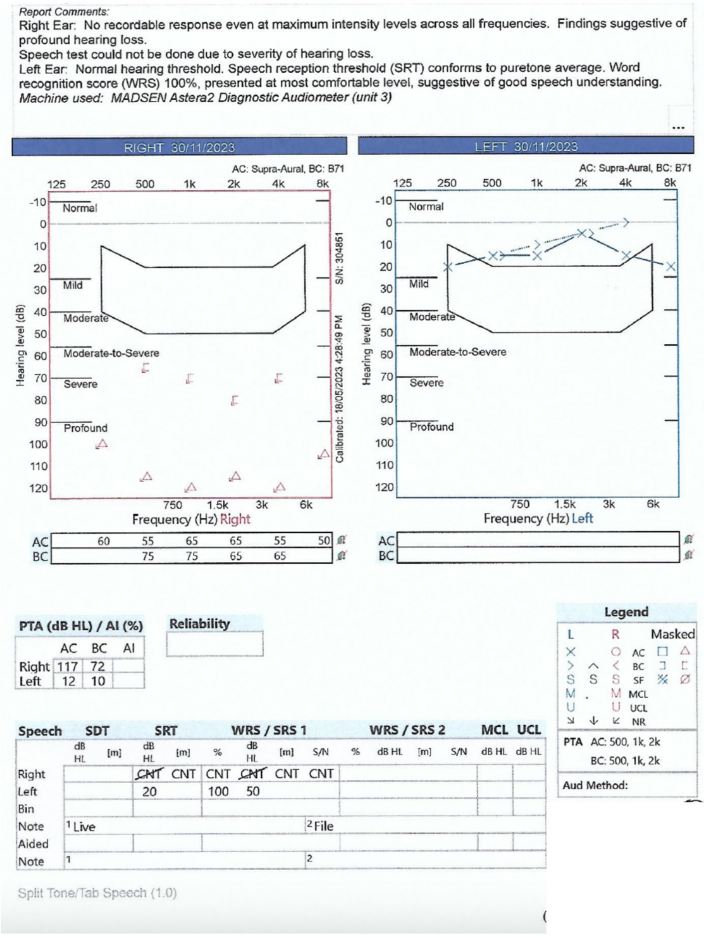

Figure 1. Puretone audiometry results showing no recordable response even at maximum intensity levels across all frequencies, suggestive of profound hearing loss in the right ear. The left ear showed a normal hearing threshold.

DISCUSSION

In this case, we highlight the important features of ILS to be able to accurately diagnose these lesions. We arrived at a diagnosis of ILS based on imaging findings after ruling out an infectious or metastatic disease process, as ILS is mainly detected by MRI.[3]

Mafee postulated that a unilateral hyperintense cochlea seen on contrast enhanced T1W imaging in a patient with progressive hearing loss and vestibular symptoms may be one of two things - labyrinthitis

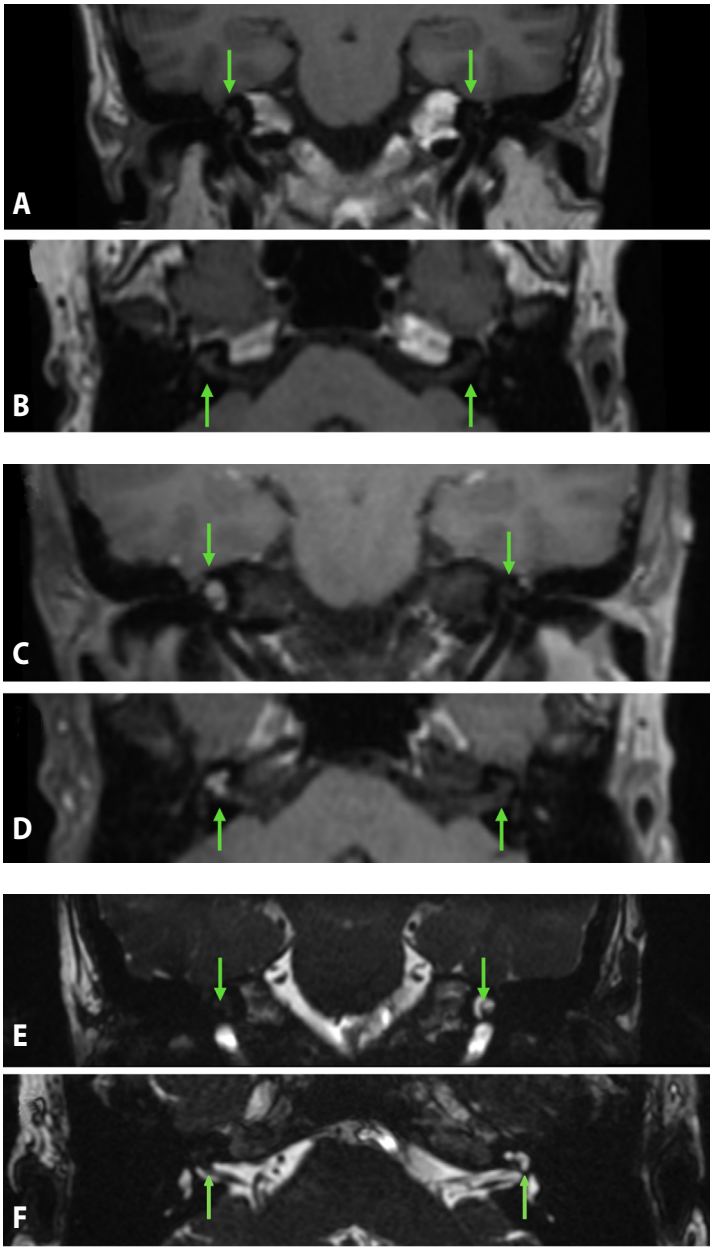

Figure 2. Gadolinium-enhanced MRI of the internal auditory canal, representative cuts with characteristic features of a transmodiolar ILS; arrows point to the cochlea on both sides. **A, B:** Non-contrast enhanced T1-weighted images (A: coronal, B: axial) showing the lesion is isointense compared to normal brain parenchyma. **C, D:** Contrast enhanced T1-weighted images (C: coronal, D: axial) demonstrating an appreciable hyperintense lesion in the right cochlea. **E, F:** T2-weighted images (E: coronal, F: axial) revealing an appreciable signal loss in the cochlea and portion of the distal IAC on the right, characteristic of an ILS.

or an ILS.[2] Due to the chronicity of our patient's symptoms and lack of infectious or viral symptomatology preceding the onset of her hearing loss and vertigo, an ILS was the more likely differential.

ILS are a benign subtype of vestibular schwannomas that are normally located in the internal auditory canal and cerebellopontine angle.[1] They originate from branches of the cochlear or vestibular nerves and can be located anywhere within the internal auditory canal (IAC) and cerebello-pontine angle (CPA).[1] These tumors may also present as a mass growing from the inner ear to the middle ear.[1] Transmodiolar ILS are a subtype of vestibular schwannomas wherein the mass is centered in the cochlea with extension into the IAC through the modiolus.[2] They are rare tumors with a US-based population study listing the incidence of ILS from 2011-2016 as only 1.1 per 100,000 person-years.[4]

ILS patients seem to receive a delayed diagnosis due to non-specific symptoms, radiologic misdiagnosis and a low case prevalence rate.[5] A retrospective multicenter study involving 12 European skull base surgery tertiary referral centers showed a mean diagnostic delay interval of 72.5 months from symptom onset.[6] However, due to technological advancements in MRI, ILS have become increasingly relevant when considering cochleovestibular disorders.[2] The gold standard for the diagnosis of these tumors remains to be MRI.[3,7] The normal inner ear has been described to have low-signal intensity on non-contrast T1-weighted imaging, similar to cerebrospinal fluid.[8] Hyperintense findings on non-contrast T1-weighted imaging point to a diagnosis of an intralabyrinthine hemorrhage or lipoma.[8] When viewed on non-contrast T1-weighted imaging, ILS may be seen as non-enhancing, isointense foci. *(Figures 2A and B)* When viewed using contrast enhanced T1-weighted imaging, ILS are typically seen as hyperintense lesions since normal inner ears would maintain a low signal intensity, similar to normal CSF.[8] *(Figures 2C and D)* On the other hand, the normal inner ear fluid presents as a bright, high intensity signal on T2-weighted imaging.[8] Conversely, an ILS would be visualized as a "filling defect" with a well-delineated signal loss. *(Figures 2E and F)*

In our case, representative cuts of our patient's MRI showed radiologic findings characteristic of ILS. *(Figure 2)* There was a contrast enhancing lesion seen over the right cochlea on T1 weighted imaging, with a signal loss over the right cochlea and IAC on T2 weighted imaging. Adequate visualization of the ILS as a cochlear mass with IAC extension clinched the diagnosis of a transmodiolar ILS.

To the best of our knowledge, there is still no gold standard treatment algorithm or reliable management plan for ILS due to the rarity of these tumors.[5] This has caused otolaryngologists to manage these patients on a case-to-case basis.[5] Kennedy *et al.* proposed that repeated MRI and close radiologic follow-up is the best option for smaller ILS seen in patients with smaller tumors.[9] However, this treatment approach may result in progressive enlargement of the mass and eventual extension into the IAC may result in progression of the patient's symptoms, a more complicated surgical course and less favorable patient outcomes.[5]

Pharmacological labyrinthectomy with intratympanic gentamicin injection has also been studied to be valuable in addressing vestibular symptoms in ILS patients undergoing radiological surveillance.[10]

Aminoglycoside administration in an ILS patient non-desirous of surgery showed significant improvement of intractable vertigo.[10]

Surgical excision of ILS remains the best option for definitive symptomatic management. However, surgery is only indicated for intractable vertigo or evidence of tumor growth leaving the membranous labyrinth into the IAC or middle ear.[11] Other indications for surgery include profound hearing loss (defined as PTA >50 dB), intolerable tinnitus, radiologically enlarging lesions, and large schwannomas with CPA extension.[11] Surgical options for the resection of ILS include labyrinthectomy, tumor resection via tympanostomy and cochleostomy, cochleoectomy, and transpromontorial endoscopic removal.[11] Plontke *et al.* note that the procedure to be done is dictated by the location of the tumor.[11] Labyrinthectomy with simultaneous cochlear implantation is recommended for tumors in the vestibular part of the inner ear. Tumors in the cochlea may be removed by total or subtotal cochleoectomy with or without second stage cochlear implantation.[11] Lastly, tumors with growth into the internal auditory canal and cerebellopontine angle can be addressed by endoscopic transcanal or transpromontorial approach.[5] The transpromontorial endoscopic approach is a recently developed surgical option for the management of ILS that offers lower morbidity.[5] A retrospective case review by Marchioni *et al*.[5] describes this technique, which allows for the complete removal of the tumor directly through the auditory canal, using the promontory region as a means to access the lesion. This approach is minimally invasive and can access the cochlea without wide skin incisions or mastoid drilling. Dural manipulation and exposure is also not required with this surgery, reducing morbidity and hospital stay. It also allows the surgeon to continuously monitor the facial nerve during the surgery, lowering the risk for iatrogenic facial nerve injury. The study involved eight ILS patients who underwent surgery using the transpromontorial approach described the advantages of the transpromontorial endoscopic approach.[5] In their study, the patients presented no major short- or long-term complications, showing good facial nerve function postoperatively.[5] They concluded that this surgical approach has the potential to become the appropriate treatment of choice for ILS, given that it is minimally invasive and reduces the risk of postoperative complications.[5]

Recently, Curatoli *et al.* suggested cochlear implantation as an option for hearing rehabilitation postoperatively if the modiolus and spiral lamina are preserved.[6] Unfortunately, our patient has a poor prognosis for recovery of hearing due to the involvement of the modiolus and the fundal portion of the cochlear nerve.

To the best of our knowledge, based on a search of HERDIN Plus, the Western Pacific Region Index Medicus (WPRIM), the Directory of Open Access Journals (DOAJ), and MEDLINE (PubMed and PubMed Central), using the search terms "intralabyrinthine," "vestibular," "schwannoma," "case report," and "Philippines," ours is the only published report of ILS in the Philippines to date, limiting data more tailored to our local setting.

In summary, ILS are rare, benign neoplasms of the 8th cranial nerve, most commonly manifesting as progressive unilateral sensorineural hearing loss with progressively worsening vertigo. A lack of disease awareness leading to misdiagnosis poses a problem when managing these cases, ultimately causing the lack of standard treatment protocols.

We recommend that clinicians be made more aware of the clinical presentation and radiologic features of ILS in order to accurately diagnose patients with this condition in a timely manner. Increased experience in managing such cases can contribute to the formation of treatment guidelines to achieve the best possible patient outcomes.

ACKNOWLEDGEMENTS

We would like to express our sincere appreciation to Dr. Angelo Augusto M. Sumalde for his valuable participation in the clinical management and for his assistance in imaging interpretation for this case report. His expertise significantly contributed to the successful completion of this work.

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