Minimally invasive endoscopic removal of primary inner ear schwannomas

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ABSTRACT
Primary Inner Ear Schwannomas (PIES) are rare benign tumors. Due to their slow growth, they are often missed during initial screening evaluation, and thus diagnosis is often delayed. Careful analysis of history with scrutiny of MRI should help to differentiate them from other pathologies resembling in symptoms. Here, we presented 4 cases diagnosed with PIES and clinical characteristics of them were analyzed. Furthermore, we elaborated a new surgical approach for the treatment of 4 PIES cases, resulting in complete tumor removal with no evidence of recurrence or residual growth on postoperative MRI follow up. No major short- or long-term complications were noted. Our case series emphasizes on treatment of PIES through a Minimal Invasive Endoscopic approach avoiding incisions and mastoid drilling thereby reducing operative time and surgical morbidity.

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Introduction
Intralabyrinthine Schwannoma (ILS) primarily arise from the Schwann cells of vestibular and cochlear nerves terminal nerve ends. Primary ILS are distinguished from Secondary ILS, the latter being developing from an extension of vestibular schwannoma (VS) through modiolus or macula. In primary ILS, Intracochlear schwannomas are more common than Intra vestibular schwannomas [1,2]. Kennedy et al. [3] in 2004 classified ILS into 7 subtypes based on the anatomical extent, however, this was later modified in 2013 by Van Abel et al. [1] renaming ILS to primary inner ear schwannomas (PIES) and adding 3 more subtypes (i) trans labyrinthine with tumors involving the vestibule, cochlea and IAC; (ii) ILS with extension into the CPA; and (iii) tumors not otherwise specified and to specify clear subsite categorization (Table 1).

PIES have an indolent growth pattern and before advanced imaging studies, they were most commonly an incidental finding during surgery for intractable vertigo or during autopsy [3]. Gadolinium-enhanced T1 weighted images with thinly sliced noncontrast T2 weighed imaging remains the gold standard for diagnosis [4]. Asymmetrical or Unilateral Sensorineural hearing loss [5,6] has been the most consistent and common finding in most of the literature accounting for about 95% of patient presentation, followed by tinnitus (70–94%), few presenting as pulsatile nature and next being vestibular symptoms ranging from acute attacks to positional vertigo and also at times mimicking Meniere’s like vertigo [7]. Since ILS have a rare incidence, no specific treatment protocols are available, however, studies and literature include conservative management, stereotactic radiotherapy, and chemical labyrinthectomy [8,9]. Surgical removal was seldom reported because the surgical approaches were too extensive. But depending on tumor characteristics and severity of patient symptoms such as handicapping vertigo, surgical treatment should be considered an option. Moreover, there were trials to remove these tumors through minimally invasive techniques such as endoscopic approaches [10]. Endoscopic ear surgery is gaining popularity and wider acceptance in diseases involving lateral skull base.

We present here few cases of PIES all presenting with progressive hearing loss over the years to non-serviceable hearing level and those with intractable vertigo. All these cases underwent minimally invasive endoscopic approach with successful outcomes and symptom-free intervals on regular follow up. The endoscopic approach offers several advantages over conventional...
approaches since bone drilling is less and provides better cosmetic results by avoiding external incisions.

**Case report**

**Patient 1**

A 41-year-old female presented with progressive left-sided hearing loss for 7 years to complete loss of hearing at her visit. She also complained of vertigo and tinnitus, not relieved by medications or repositioning maneuvers. Audiological examination revealed completely deaf in the left ear and absent WRS while the right side hearing was within normal limits. Videonystagmography revealed a significant 75% left-sided caloric paresis. Magnetic resonance imaging (MRI) revealed an enhancing lesion in the left vestibule extending to the left basal turn of cochlea (Figure 1(A)).

Because of intractable vertigo, she strongly wanted surgical removal and a minimally invasive modified Exclusive Endoscopic Transcanal Transpromontorial approach (mEETTA) was done [10]. Surgical procedures: 1) tympanomeatal flap elevation, all ossicles were removed, 2) facial nerve was demarcated from geniculate ganglion to proximal second genu, 3) transcanal endoscopic drilling of the promontory was done exposing basal turn filled with tumor and drilling continued inferiorly to expose the vestibule, 4) tumor was entirely removed, 5) promontory opening was plugged with soft tissue from tragal area, and 6) a tragal cartilage tympanoplasty was done (Supplement 1). The tumor was removed (Figure 1(B)) and histological diagnosis proved to be consistent with Intra labyrinthine schwannoma. The patient improved symptomatically and well-healed tympanic membrane was observed (Figure 1(C)) on regular follow up.

**Patient 2**

A 40-year-old male presented with unilateral hearing loss of right side, progressive in nature for 4 years. Four years before, his initial hearing test revealed mild hearing loss only in high frequencies and there was no tumorous condition in MRI. But his hearing

**Table 1.** Classification of Intralabyrinthine Schwannoma by Kennedy et al. [3].

| Subtypes                  | Anatomical Location          |
|---------------------------|-------------------------------|
| Intravestibular (IV)      | Vestibule and/or SCC          |
| Intracocharal (IC)        | Cochlea                       |
| Intravestibulocochlear (IVC) | Vestibule and/or SCC + Cochlea |
| Transmodiolar (TMOD)      | Vestibule and/or SCC + IAC    |
| Transmacular              | Vestibule and/or SCC + cochlea + IAC |
| Transotic (TO)            | Vestibule and/or SCC + cochlea + middle ear |
| Tympanolabyrinthine (TML) | Vestibule and/or SCC + cochlea + Middle ear |

SCC: semicircular canals; IAC: internal auditory canal; CPA: cerebellopontine angle.
worsened progressively and reached to deaf in four years along with intractable vertigo not subsiding on medications. Videonystagmography revealed 100% canal paresis on the right side. MRI revealed a mass of $0.8 \times 0.5$ cm involving the right vestibule and a diagnosis of intravestibular schwannoma was obtained (Figure 2(A)). Considering an obvious increase in the size of tumor during follow-ups and the presence of intractable vertigo, the patient opted for surgical removal. Using the endoscopic approach, the tumor was removed and dummy electrode (Nucleus 422) was inserted for cochlear implantation at a later stage considering his financial constraints (Figure 2(B,C), Supplement 2). The patient resolved completely from vertiginous symptoms and is on follow up with a plan for future replacement with a cochlear implant as and when the patient desires.

**Patients 3 and 4**

Our last two patients in the series presented with left-sided progressive hearing difficulty and developing intractable vertigo during serial follow-ups. Both patients were evaluated initially showing nontumorous state, however audiological parameters worsened from Class C to Class D (AAO HNS Grading) with WRS score of 5% for the former and <50% for the latter during serial follow-ups. Serial MRI showed tumor evidence involving the vestibule in patient 3 (Figure 3(A)) and cochlear involvement in patient 4 (Figure 3(C)). Both the patients opted for surgical removal considering the persistence of vertiginous symptoms affecting routine activities not resolved to any medical management. Endoscopic approach was advocated for both and is on regular follow up with no evidence of tumor residue or recurrence (Table 2).

**Discussion**

The presentation of PIES can be sporadic and isolated or can present in Neurofibromatosis type (NF2), the former being more common [7]. Hearing loss is the most common symptom, as observed in our case series, and can range from sudden, progressive to fluctuating variants [5,12]. Pathophysiology of hearing loss in Intracochlear tumors could be due to compression...
of the organ of Corti, or cochlear nerve and its vascular supply, however for intravestibular tumors only a few theories have been stated. One possible mechanism depicted was compression of ductus reuniens and saccule leading to endolymphatic hydrops [12]. Other symptoms of PIES include vertigo, tinnitus, and aural fullness. One of the probable delay in diagnosis has been stated as misdiagnosis with Meniere’s disease because of the resemblance of symptoms [3], others being slow growth of the tumor and very small lesion not being detected in MRI, also subtle symptoms may not always prompt for MRI Examination and often they are diagnosed later [7]. Currently, MRI with three-dimensional fast spin-echo T2 weighted imaging generating images of 0.8–1.0 mm can be used effectively as a screening test and also to assess tumor growth and stability. Recent studies have shown that filling defect in T2 can be substantiated by post-contrast T1 not only adding cost-effectiveness but also reducing gadolinium exposure to patients [14]. Also, MRI along with proper history evaluation can exclude other differentials such as labyrinthitis, labyrinthitis ossificans, inner ear hemorrhage, and lipoma. However, it is sometimes difficult to differentiate between actual residual tumor and connective tissue enhancement in contrast MRI during serial follow ups after surgery.

There are three treatment options currently accepted for PIES: i) wait and watch policy using serial MRI scans, ii) stereotactic radiotherapy, and iii) surgical excision. Ideally, any plan generated should take into account the tumor location and growth, patient symptoms, age of the patient, and comorbid illness and patient wishes. Those tumors with serviceable hearing without intractable vertigo, wait, and watch can be employed since they are benign tumors with slow growth [8]. In patients with intractable vertigo and non serviceable hearing, chemical labyrinthectomy using gentamycin is an alternative intial procedure considering low cost and low risk [8]. There is currently lack of literature or consensus for indications of stereotactic radiotherapy except in cases those unfit for surgery like the elderly [12]. Surgical removal is also not the 1st line indication, but in those patients with tumor growth over serial follow scans contributing to non-serviceable hearing and intractable vertigo, surgical removal seems to be promising [3,8]. Our series comprised of similar presentations who were initially under serial follow-ups, but tumor growth with progression of
symptoms made them opt for surgical management over others.

In the past, the surgical options for PIES only include transmastoid, translabyrinthine, or transotic approach depending on the location and extent of the tumor. Most of these approaches require excessive intervention to normal anatomical structures. Recently, microscopic transcanal approaches were reported to reduce morbidities from previous excessive approaches [15].

Moreover, endoscopic application to ear surgery and lateral skull base surgery facilitated this minimal invasive concept. Marchioni et al. [10] reported on 8 patients of ILS with unserviceable hearing undergoing total endoscopic removal without any significant complications and on follow up. Our series are similar but confined to PIES. Total transcanal endoscopic approach to PIES has advantages: minimal intervention, reducing operative time, reducing postoperative morbidities, providing better illumination/magnification, and minimizing blind spots for preventing incomplete resection [10,16]. Limitations of surgical approach would include facial nerve injury, CSF leakage, and thermal damage due to excessive heat generated from endoscope. Our series did not have any complications though limitation of our study was fewer number of cases. Thermal damage was minimised through frequent saline irrigation and a trained assistant can help with limitation of single handedness of an endoscopic approach.

Our series of four patients during follow ups had no residual/recurrent tumor growth, completely resolved from vertigo possibly due to abolition of atypical peripheral signals after tumor resection, and improvement in tinnitus among two of whom had severe grade pre op (Table 3).

One among our series underwent simultaneous surgical removal and electrode dummy insertion for future implantation. Financial constraints prevented real implantation in the initial stage. Carlson et al. [17] described Cochlear Implant insertion in 3 cases of PIES without removing the tumor, however, in a few cases stylet was lately deployed to overcome the resistance of insertion through the tumor and got promising initial outcomes. Postoperative surveillance of tumor also can be possible with a magnet in place. Fear of a tumor within prioritized our patient for tumor removal and subsequent dummy implantation. Therefore, Endoscopic Transcanal Transpromontorial Approach provides a viable option for tumor removal and auditory rehabilitation simultaneously improving the quality of life for patients.
Conclusion
PIES are rare tumors. Its location, growth, patient symptoms, and comorbid illness all should be taken into account in treatment planning. According to the author’s opinion, minimally invasive endoscopic surgical removal is a promising option in properly selected candidates.

Informed Consent
Written and informed consent was obtained from the patient regarding the use of his clinical findings and reports of the investigations that were conducted.

Disclosure statement
The authors involved in this study declare no conflicts of interest.

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Table 3. Follow up.

| Patients | Pre op HB grading | Post op HB grading | MRI follow up (months) | Follow up status | Symptoms during follow up |
|----------|-------------------|--------------------|------------------------|-----------------|--------------------------|
| Case 1   | I                 | I                  | 39                     | No tumor        | Tinnitus initially but improved |
| Case 2   | I                 | I                  | 26                     | No tumor        | None                     |
| Case 3   | I                 | I                  | 14                     | No tumor        | None                     |
| Case 4   | I                 | I                  | 16                     | No tumor        | Tinnitus immediate post op but improved |

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