Case Report

Intracochlear Schwannoma

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Abstract Intracochlear schwannoma may cause progressive or sudden sensorineural hearing loss. Radiologic findings can be overlooked especially when attention is focused on the internal auditory canal. Otolaryngologists should be familiar with the diagnosis and treatment of this important but uncommon lesion.

Keywords intracochlear schwannoma; intralabyrinthine schwannoma; intravestibular schwannoma; magnetic resonance imaging; hearing loss; tinnitus; vertigo; tumor growth; acoustic neuroma

1. Introduction

Acoustic schwannomas are intracranial tumors that arise from the Schwann cell sheath investing either the vestibular or cochlear nerve typically at the cerebellopontine angle or within the internal auditory canal. Intralabyrinthine schwannomas (ILSs) can involve the cochlea, vestibule, semicircular canals or a combination of these structures. ILSs are rare and difficult to diagnose. ILSs are rare and difficult to diagnose [2,4,5,6,8,9,10,11,12,14,15,17,18,20,21,22,23,24,25,26,27,28,29,30,31,32,33,34,35,36,37,38,39,40,41,43,44,45,46,47,48,49] and intracochlear schwannomas (ICSs) are even more rare [3,13]. Consequently, there is limited information in the literature concerning their epidemiology and management.

2. Case report

A 60-year-old female developed sudden decreased hearing and pulsatile tinnitus in the right ear three years prior to our evaluation. Initial pure tone audiometry and speech discrimination score revealed asymmetric sensorineural hearing loss, slightly worse in the right (Figure 1).

An MRI with gadolinium revealed a normal examination of the brain and internal auditory canals. However, enhancement of the basal turn of the cochlea was read as suggestive of labyrinthitis. The patient was diagnosed subsequently with autoimmune hearing loss on the basis of blood tests (HLA typing and Western blot for 68–70 kDa antigen) and started on high dose prednisone and methotrexate. Hearing loss persisted and the tinnitus progressed. Repeat pure tone audiometry and speech discrimination scores six months later were worse (Figure 2). A repeat MRI with gadolinium of the brain with attention to the IACs showed increased enhancement of the right cochlea suspicious for cochlear schwannoma. At that point, the patient was referred to our office. Presentation included fluctuating tinnitus characterized as “tunnel-like” and progressive hearing loss. Otoscopic examination was normal. Hitselberger’s sign was positive on the right, and she had diplacusis (relatively flat [lower pitch] on the right).

Electronystagmography, facial electroneuronography, and laboratory testing (glucose, cholesterol, triglycerides, FTA-abs, Lyme titer, thyroid function tests and collagen vascular tests) were normal except for repeat 68–70 kDa antigen and HLA typing. A new MRI showed an abnormal focus of enhancement within the middle turn of the right cochlea measuring 5 mm × 2 mm (Figure 3).

Options were reviewed, and the patient elected observation. Over the following 6 months, monthly pure tone audiograms showed progressive deterioration on the left. The patient elected to have the tumor excised because of growth of the lesion, progressive hearing loss, tinnitus, and imbalance.

A right extracranial translabyrinthine approach was used (Figure 4), and a bone anchored hearing aid was implanted simultaneously. The patient did well post-operatively. Histopathology confirmed the diagnosis of schwannoma.

3. Discussion

ILSs are rare tumors of neural origin found within the cochlea, semicircular canals, vestibule, or any combination of these structures. ILSs arise from Schwann cells of the cochlear, superior and inferior vestibular nerves [28,30]. ICSs are a subcategory of ILSs [3]. ILSs are considered by most to be a separate entity from vestibular schwannomas (acoustic neuromas), as ILSs usually do not involve the internal auditory canal (IAC) or cerebellopontine angle (CPA) [13,16,31,42]. While it is common for vestibular...
schwannomas to invade the labyrinth, it is uncommon (but not unprecedented) for ILSs to invade the IAC or brainstem. Unfortunately, some authors have categorized all schwannomas involving the cochlea as ICS, regardless of the origin [3]. This confusion has led to what we believe is an overestimate of the incidence of ICS.

The first case of an ICS was reported by Mayer in 1917 [28]. However, other authors have cited Nager as the first to discover an ICS in 1917 upon autopsy of a deaf patient [30]. Most early reports of ILS were based upon autopsy or surgery for severe vertigo [46]. Many patients were thought to have had Meniere’s disease. Accurate diagnosis has been much more common since the advent of high resolution MRI [18, 16]. Today, with MRI we can make a diagnosis of ICS at a lesion size of $< 2$ mm [18].

ILSs have been reported in 94 patients, dating back to Mayer [28] in 1917 [2, 4, 5, 6, 9, 10, 13, 14, 15, 17, 18, 20, 21, 22, 23, 24, 25, 26, 27, 29, 31, 32, 33, 34, 35, 36, 37, 38, 39, 40, 41, 43, 44, 45, 46, 48]. Roughly 80 cases of ICS have been reported worldwide, but we believe this figure to be too high. Upon review of the literature, Kennedy et al. have found only 26 cases of ICS using a classification model described in their paper [25]. We believe that this is a more accurate assessment of ICS incidence.
ILSs cannot be differentiated from vestibular schwannomas histopathologically. Both tumors may contain Antoni A and B components [38, 48]. Theories regarding this lesion have evolved from initial beliefs that these tumors originated from the Schwann-glial cell junction (Obersteiner-Redlich zone) to more recent evidence suggesting that they arise from the myelinated portion of the nerve, lateral to the transition zone [13, 25, 34, 38].

Table 1: Presenting symptoms of Intralabyrinthine schwannomas (n = 55).

<table>
<thead>
<tr>
<th>Symptom</th>
<th>No. of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing loss</td>
<td>48 (87)</td>
</tr>
<tr>
<td>Progressive</td>
<td>33 (69)</td>
</tr>
<tr>
<td>Sudden</td>
<td>12 (25)</td>
</tr>
<tr>
<td>Fluctuating</td>
<td>3 (6)</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>31 (65)</td>
</tr>
<tr>
<td>Vertigo</td>
<td>14 (29)</td>
</tr>
<tr>
<td>Aural fullness*</td>
<td>2 (4)</td>
</tr>
</tbody>
</table>

*Aural fullness not accounted for in Grayeli paper. Statistics used from [1, 9, 18].

The clinical manifestations of ILS have been documented well. These tumors typically produce sensorineural hearing loss, tinnitus, vertigo, disequilibrium, and aural fullness. Progressive hearing loss is the most common presenting symptom of ILS (Table 1) in general, as well as for the ICS subset (Table 2). Profound-to-severe sensorineural hearing loss associated with poor speech discrimination scores is found on initial presentation in a majority of cases. Tinnitus often is associated with hearing loss at the time of initial presentation [12, 30, 31]. Misdiagnosis as Meniere’s disease is common. Mixed or conductive hearing loss without middle ear pathology also has been found in patients with ILS [9, 14, 24, 42]. Some might suspect that if the tumor were more confined to the vestibular system or cochlear system, then the patient would present with more predominant vertigo or hearing loss, respectively; but there is no evidence to support this presumption.
## Table 2: Presenting symptoms of Intracochlear schwannomas (n = 22).

<table>
<thead>
<tr>
<th>Symptom</th>
<th>No. of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing loss</td>
<td>15 (68)</td>
</tr>
<tr>
<td>Progressive</td>
<td>12 (80)</td>
</tr>
<tr>
<td>Sudden</td>
<td>3 (20)</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>6 (27)</td>
</tr>
<tr>
<td>Dizziness/vertigo</td>
<td>4 (18)</td>
</tr>
<tr>
<td>Facial paresis</td>
<td>1 (5)</td>
</tr>
<tr>
<td>Aural fullness</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

Statistics used from [12,18].

Little information is available on diagnostic tests other than pure tone audiometry (PTA) for patients with ICS. Otologists commonly include auditory brainstem response (ABR), electronystagmography (ENG), and otoacoustic emission (OAE) testing in their initial evaluation of patients with the above presenting symptoms. Contrary to ABR findings in Schwannomas of the IAC, ICSs typically do not show abnormalities consistent with a retrocochlear disorder. If the ABR in a known case of ICS begins to show retrocochlear signs, involvement of the IAC must be considered [25]. While most authors agree that ILS will reveal reduced vestibular response on ENG as seen by Neff et al. [31], no authors noted ENG differences between ICSs and other ILSs versus Schwannomas of the IAC and brainstem.

As radiologic imaging technology has improved, so too has the ability to diagnose ICS. The first documented diagnosis of ICS by radiologic imaging was made by Karlan et al. [24]. This diagnosis was again recognized by Mafee et al. in 1990. They described widening of the basal turn of the cochlea, with erosion of the promontory [27]. Following reports by Doyle and Brackmann and Saeed et al., MRI was utilized routinely to diagnose these lesions [9, 36]. Currently, the “gold standard” test for all acoustic neuromas, including ILS, is MRI with gadolinium [8], although other techniques have been proposed [19]. With improvements in MRI, ICS can now be identified as small as 2 mm in size utilizing T1/T2-weighted views [18]. However, two cases of intralabyrinthine tumors without conclusive evidence on MRI have been described in the literature [49].

ICSs are seen on high-resolution T2-weighted MRI as an area of filling defect confined within the cochlea. In T1-weighted scans, the lesion within the cochlea is focally enhancing with moderate or high signal intensity [1,7,8]. The scala tympani is involved more frequently or more extensively than the scala vestibuli [2].

In order to avoid a missed diagnosis of ICS, it is necessary to pay particular attention to abnormal lesions within the cochlea and to repeat MR imaging if suspicious lesions are noted. With early diagnosis, it should be possible to follow many of these tumors in patients with serviceable hearing with repeat imaging. This permits hearing preservation for as long as possible and may allow us to learn more about the natural history of ICS and to consider the efficacy of early use of nonsurgical treatment modalities.

Management of ILS varies widely depending on the location of the tumor as well as the patient’s clinical presentation. We suggest, as others have, that if serviceable hearing is present with moderate-to-absent vestibular symptoms, observation usually is indicated with serial audiograms and MRI scans [5,12,31]. Our protocol entails an MRI with gadolinium every six months for the first two years to assess growth. If no growth is identified, yearly MRI is performed for years 3 to 5. After five years, we repeat the MRI every two years. However, audiograms are performed at least every six months, and sooner if symptoms worsen. MRI may be repeated sooner if hearing worsens or other symptoms change. Patients with nonserviceable hearing and intractable vertigo are candidates for surgical tumor excision, although continued observation with serial MRIs is an acceptable option. In addition, surgery may be indicated when ICSs show evidence of extension into the IAC [3,9,31]. As these tumors encroach on the IAC, the risk of injury to the facial nerve may increase either through direct extension or increased surgical risk. Chemical labyrinthectomy has been discussed as a viable option for treating symptoms of ICS [31]. This treatment is reserved for the rare patient who exhibits serviceable hearing and intractable vertigo. If a patient is not responsive to conservative treatment of vertigo, early excision should be considered, as these lesions typically lead to severe hearing impairment. Stereotactic radiotherapy is a management option for ILSs although its use for these lesions has not been explored extensively, and the risk of hearing loss from radiosurgery is unknown.

When surgical excision is indicated in ICSs, two approaches have been discussed. A translabyrinthine approach has been reported as effective in several studies [9,14,31]. However, some authors believe this approach may limit surgical exposure and increase risk of incomplete removal of tumor extension in the basal turn of the cochlea [3]. A transotic approach may allow for adequate exposure of labyrinthine structures, IAC, and the CPA. The literature does not provide statistically a significant evidence to suggest that one approach is superior to the other.

### 4. Conclusion
ICS is a rare, benign cause of asymmetric sensorineural hearing loss. Typical presentation includes hearing loss, vertigo, and tinnitus. MRI with gadolinium is the best test to diagnose these tumors. Management of ICS should be individualized based on the patient’s presentation and needs.

### References
1. American Academy of Otolaryngology-Head and Neck Surgery Foundation, Inc., Committee on Hearing and Equilibrium guidelines for the evaluation of hearing preservation in acoustic