Ir J Med Sci (2012) 181:131–134
DOI 10.1007/s11845-010-0572-5

CASE REPORT

Intracochlear schwannoma presenting as diffuse cochlear enhancement: diagnostic challenges of a rare cause of deafness

M. E. Miller · J. M. Moriarty · M. Linetsky · C. Lai · A. Ishiyama

Received: 10 April 2010 / Accepted: 3 September 2010 / Published online: 18 September 2010

Abstract Intracochlear schwannoma is a rare, treatable, cause of unilateral hearing loss. Due to the small size, position, and variable clinical and imaging features, diagnosis presents a significant challenge and is often delayed. We present a case of a patient with an intracochlear schwannoma presenting as a diffuse enhancement of the cochlea, mimicking an infectious or inflammatory process. The absence of focal nodularity in this lesion on multiple high-resolution MRI examinations led to a delay of over 3 years from the patient's initial presentation to surgical diagnosis. Clinical history and examination, imaging features, pathologic findings, and surgical management options are described.

Keywords Intracochlear schwannoma · Hearing loss · MRI · Labyrinthectomy

M. E. Miller · A. Ishiyama
Department of Head and Neck Surgery, David Geffen School of Medicine, UCLA, 10833 LeConte Avenue, Los Angeles, CA 90095, USA

J. M. Moriarty · M. Linetsky
Department of Radiological Sciences, David Geffen School of Medicine, UCLA, 10833 LeConte Avenue, Los Angeles, CA 90095, USA

C. Lai
Department of Pathology and Laboratory Medicine, David Geffen School of Medicine, UCLA, 10833 LeConte Avenue, Los Angeles, CA 90095, USA

J. M. Moriarty (✉)
Diagnostic Cardiovascular Imaging, David Geffen School of Medicine, UCLA, Peter V. Ueberroth Bldg, Suite 3371, 10945 LeConte Avenue, Los Angeles, CA 90095-7206, USA
e-mail: jmoriarty@mednet.ucla.edu

Introduction

Eighth cranial nerve schwannomas are benign tumors that are more likely to occur in the internal auditory canal (IAC) or in the cerebellopontine angle (CPA) cistern than within the cochlea. Although, most schwannomas are sporadic, they may be associated with Neurofibromatosis Type II [1]. Intracochlear schwannomas are a rare cause of unilateral hearing loss, with fewer than 80 cases reported in the literature. Within the labyrinth, intravestibular schwannomas are more common than intracochlear schwannomas [2].

Histologically, intracochlear schwannoma is identical to other schwannomas, apart from their location [3]. These lesions are composed of schwann cells and tend to occur in the modiolus and the basal turn of the cochlea along the path of spiral ganglion dendrites [4].

Typically, intracochlear schwannomas present as focal nodular enhancing lesions. We present a case of an intracochlear schwannoma demonstrating atypical imaging features on MRI, contributing to a delay in diagnosis and treatment. A high clinical index of suspicion is necessary for diagnosis and treatment.

Case report

A 33-year-old woman presented with 3 years of progressively worsening left-sided hearing loss, aural fullness, and intermittent vertigo. She denied tinnitus, ototoxic exposures, trauma, and past otitis media.

MRI of the internal auditory canals (IACs) performed on a 1.5 T scanner showed a diffuse enhancement of the left cochlea on T1 post-gadolinium images (Fig. 1). Axial 3D CISS images showed no evidence of a focal nodular lesion.

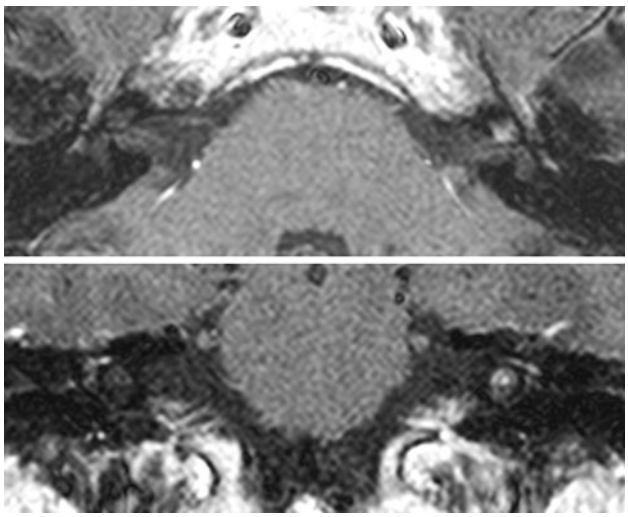

Fig. 1 Axial (*top*) and coronal (*bottom*) post-contrast T1-weighted images (obtained on a 1.5 T Siemens Sonata scanner) show diffuse enhancement of the left cochlea. There is no abnormal enhancement of the left internal auditory canal or vestibule

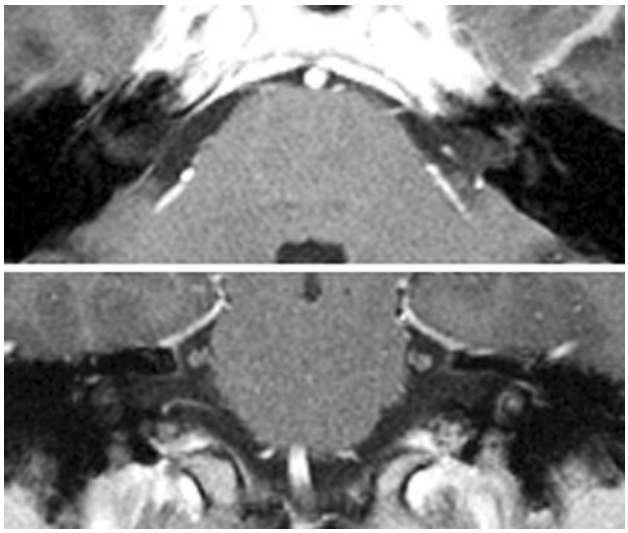

Fig. 2 Axial (*top*) and coronal (*bottom*) post-contrast images obtained on a 3 T Siemens Verio scanner continue to show faint diffuse enhancement of all turns of the left cochlea

The differential diagnosis at that time included viral, bacterial, and autoimmune phenomena. Short-term follow-up MRIs showed a qualitative decrease in the degree of enhancement, and the acute process causing these MRI changes was thought to have improved.

The patient presented 3 years later with worsening symptoms. Repeat MRI, this time performed on a 3 T scanner, again showed smooth diffuse enhancement of all three half-turns of the cochlea (Fig. 2). Steady-state imaging continued to show no evidence of a focal nodular lesion (Fig. 3). The patient's neurotologic exam was within normal limits. Audiologic examination revealed normal hearing in the right ear and moderate-to-severe sensorineural hearing loss in the left ear with a speech reception threshold of 45 dB and word discrimination score (WRS) of 44%. Electronystagmography demonstrated a left-sided significant 40% caloric paresis. Repeated audiologic exam several months later demonstrated stable hearing thresholds, but a worsening WRS of 8%. Due to the lack of serviceable hearing and intractable vertigo, the patient underwent left transmastoid labyrinthectomy with a bone-anchored hearing aid (BAHA) placement.

At surgery, a mass (dimensions 0.4 × 0.3 × 0.1 cm) was found to be confined to the labyrinth and was removed as part of the labyrinthectomy. On pathology, a schwannoma was found arising from within the cochlea. The lesion consisted of schwann cells appearing as spindle cell proliferation, and the S100 marker was positive on immunohistochemical staining. Verocay bodies, which consist of a group of fusiform cells arranged in whorls or palisades, were also evident (Fig. 4).

Discussion

Intracochlear schwannoma was first described in 1917 [5], using archival human temporal bone specimens. This tumor often presents with non-specific audiovestibular symptoms. Hearing loss is the most common presenting symptom. Although the majority of patients present with progressive hearing loss, as in our case, 31% of patients with intralabyrinthine schwannomas have sudden sensorineural hearing loss [1]. Other symptoms include tinnitus, vertigo, imbalance, and aural fullness. Diagnosis of intracochlear schwannoma is difficult and often delayed because these presenting symptoms are shared with other otologic diseases, particularly Meniere's disease. In one series of 19 patients, the mean delay from the onset of symptoms to diagnosis was 11 years [6].

Although auditory brainstem response (ABR) testing is highly sensitive for schwannomas larger than 1 cm, it is inferior to gadolinium-enhanced MRI, which is the gold standard for diagnosis [7]. In one study of intralabyrinthine schwannomas, MRI was diagnostic in 96% of patients (one patient had an incidental finding of intracochlear schwannoma at cochlear implant surgery) [1].

Current MRI evaluation of the inner ear includes axial and coronal thin-section gadolinium-enhanced sequences to detect the presence of an enhancing lesion as well as a steady-state free precession sequence (SSFP, also known as FIESTA or CISS) for improved spatial resolution. Although MRI is useful in making the diagnosis, it too, is limited by tumor size. Small intralabyrinthine lesions may be particularly difficult to see on MRI, and may be seen only in retrospect [8].

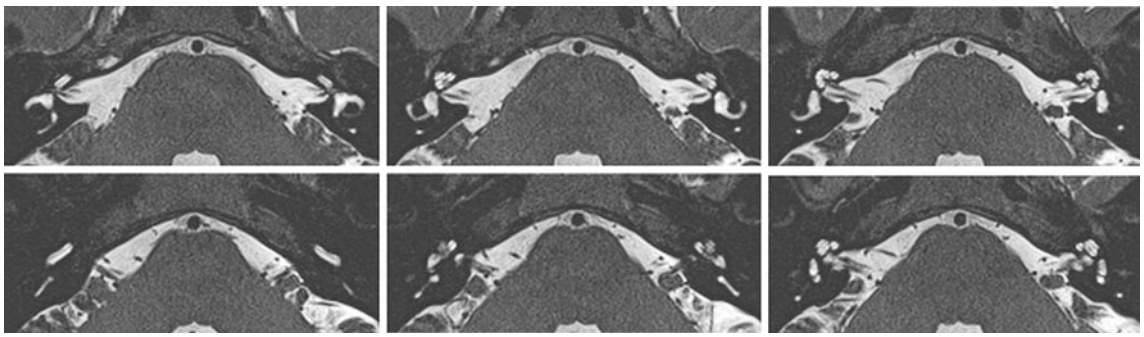

Fig. 3 Multiple axial steady-state free precession (SSFP) images obtained on a 3 T Siemens Verio scanner show normal cochlear anatomy. There is no evidence of a focal nodular lesion

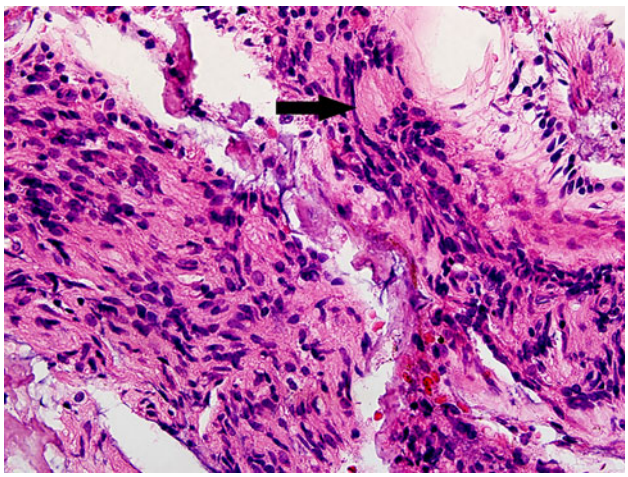

Fig. 4 Histopathology of the labyrinthine lesion consistent with schwannoma, appearing as spindle cell proliferation. The arrow indicates a Verocay body, which consists of a group of fusiform cells arranged in whorls or palisades

MRI is usually ordered when asymmetric hearing loss raises the question of a possible acoustic neuroma. However, the expensive use of frequent MRIs to diagnose intracochlear schwannoma is a concern, especially as many initial screening scans are negative. Some authors have suggested more stringent criteria for obtaining MRIs to diminish these costs [9].

Once diagnosed, not all the patients with intracochlear schwannomas require surgery; the decision for treatment depends on size and extent of tumor as well as patient symptoms [7]. Large tumors with extra-labyrinthine extension usually undergo surgical treatment or stereotactic-guided radiotherapy. In patients with small tumors and serviceable hearing, conservative management with repeat MRIs to follow tumor growth may be appropriate [10].

When patients have small tumors without useful hearing and when vestibular symptoms indicate the need for treatment, as in our case, surgery is planned. Both translabyrinthine and transotic approaches have been described; the transotic approach is superior for more extensive tumors [7]. Because of the risks of surgery, some authors advocate conservative management whenever possible: in his series from 1992 to 2002, Kennedy describes only 30% of patients with intralabyrinthine schwannomas requiring operative management [1].

Conclusion

We present an unusual appearance of an intracochlear schwannoma. Although the vast majority of these tumors are seen as focal nodular enhancing masses on MRI, smooth diffuse enhancement of the cochlea does not exclude the presence of a schwannoma. In patients with a prolonged clinical presentation atypical for an inflammatory or infectious lesion, diffuse cochlear enhancement should not exclude the possibility of an intracochlear schwannoma.

Conflict of interest None.

References

1. O'Keefe LJ, Camilleri AE, Gillespie JE et al (1997) Primary tumors of the vestibule and inner ear. J Laryngol Otol 111:709–714
2. Kennedy RJ, Shelton C, Salzman KL et al (2004) Intralabyrinthine schwannomas: diagnosis, management, and a new classification system. Otol Neurotol 25(2):160–167
3. Soler YC, Fayad JN (2007) Intracochlear schwannomas. Otol Neurotol 28(1):142
4. Deux JF, Marsot-Dupach K, Ouayoun M et al (1998) Slow-growing labyrinthine masses: contribution of MRI to diagnosis, follow-up and treatment. Neuroradiology 40(10):684–689
5. Mayer O (1917) Ein fall von multiplen tumoren in den endausbreitungen des akustikus. Z Ohrenheilkd 75:95–113

6. Grayeli AB, Fond C, Kalamarides M et al (2007) Diagnosis and management of intracochlear schwannomas. Otol Neurotol 28(7):951–957
7. Zappia JJ, O'Connor CA, Wiet RJ, Dinces EA (1997) Rethinking the use of auditory brainstem response in acoustic neuroma screening. Laryngoscope 107(10):1388–1392
8. Di Lella F, Dispenza F, De Stefano A, Falcioni M, Sanna M (2007) Management of intralabyrinthine schwannomas. Auris Nasus Larynx 34(4):459–463
9. Saliba I, Martineau G, Chagnon M (2009) Asymmetric hearing loss: rule 3, 000 for screening vestibular schwannoma. Otol Neurotol 30(4):515–521
10. Bozorg Grayeli A, Kalamarides M, Ferrary E et al (2005) Conservative management versus surgery in small vestibular schwannomas. Acta Otolaryngol 125(10):1063–1068