Intracochlear Schwannoma

Giuseppe Magliulo, M.D.,¹ Giovanna Colicchio, M.D.,¹
 Alla Francesca Romana, M.D.,¹ and Alessandro Stasolla, M.D.²

ABSTRACT

Intralabyrinthine schwannomas are benign, slow-growing tumors that originate from Schwann cells lining the terminal ends of the cochlear and vestibular nerves. Magnetic resonance imaging (MRI) with gadolinium is considered the best diagnostic tool for this disease. Kennedy et al proposed a classification system, based upon the MRI observations, that identifies seven different classes according to the site of the tumor: intravestibular, intracochlear, intravestibulocochlear, transmodiolar, transmacular, trans-otic, and tympanolabyrinthine. A case of a patient undergoing a 2-year follow-up with serial MRI and managed with a wait-and-see strategy is described. The rationale of the diagnosis and the different treatments of choice are discussed.

KEYWORDS: Intralabyrinthine schwannoma, MRI, surgery, follow-up

In the early 1990s, with the advent of gadolinium (Gd)-enhanced and heavily T2-weighted magnetic resonance imaging (MRI), an increasing number of intralabyrinthine schwannomas were detected during the investigation of hearing loss or vertigo. A few case reports and small series have described the imaging findings of intralabyrinthine schwannomas.¹⁻⁵ In the largest series, Tieleman et al⁵ reviewed 52 cases. An intralabyrinthine schwannoma usually presents with progressive hearing loss, and 75% of cases exhibit total deafness in the affected ear. Vertigo is less frequent, not exceeding 10% in the reported series. The vertigo may be pharmacologically intractable, requiring surgery.

CASE REPORT

A 41-year-old man presented with progressive, non-fluctuating asymmetric hearing loss that started at 35 years of age, with no vestibular disturbances. Pure-tone audiometry showed a profound hearing loss with 0% discrimination at speech audiometry. On MRI, heavily T2-weighted fast spin-echo (FSE) images in

the axial planes depicted a normal basal turn of the left cochlea. The posterior parts of both scalae of the second turn were occupied by a schwannoma. Normal fluid was seen in the anterior second turn of the scala tympani (Fig. 1). Grossly, the first turn was occupied by the lesion, although it opened somewhat at the apex. T1-weighted images and Gd-enhanced T1-weighted images in the axial planes showed a strongly enhancing schwannoma in the first and second cochlear turns (Fig. 2).

Wait-and-see management was planned with serial MRI, with follow-up at 6 months and then every year for 5 years, to assess the development of the tumor. At the 2-year follow-up, there was no evidence of tumor growth.

DISCUSSION

Intralabyrinthine schwannomas have variable presenting symptoms (unilateral hearing loss, vertigo, tinnitus, and ear fullness).¹⁻⁵ The literature reports no evidence of more frequent audiological impairment with progressive unilateral sensorineural hearing loss associated or not with vertigo. This latter usually occurs later in the course

¹Otorhinolaryngology, Audiology and Phoniatrics "G. Ferreri" Department, University La Sapienza of Rome, Italy; ²Radiology Department, Azienda Sanitaria San Camillo, Rome, Italy.

Address for correspondence and reprint requests: Giuseppe Magliulo, M.D., Via Gregorio VII n 80, 00165 Rome, Italy (e-mail: giuseppemagliuloor@yahoo.com).

Skull Base 2010;20:115-118. Copyright © 2010 by Thieme

Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1 (212) 584-4662.

Received: May 19, 2009. Accepted: June 14, 2009. Published online: August 12, 2009.

DOI: <http://dx.doi.org/10.1055/s-0029-1236166>.

ISSN 1531-5010.

Figure 1 Heavily T2-weighted fast spin-echo images in the axial planes (A, B, C). A normal basal turn of the left cochlea. A schwannoma occupies the posterior parts of both scalae of the second turn. Normal fluid in the scala tympani of the anterior second turn (arrow). The first turn is full, although it opens somewhat at the apex.

of the disease. The nonfluctuating hearing loss could differentiate the nature of the disease from other forms of hearing deteriorations, indicating the need of high-resolution techniques of imaging.

Intralabyrinthine schwannomas may have slightly higher signal intensity than normal intralabyrinthine fluid on unenhanced T1-weighted images. On T2-weighted high-resolution images, they appear as hypointense lesions with sharp borders, replacing the high signal intensity fluid. T2-weighted high-resolution images can distinguish in which scala a lesion is situated. After Gd administration, they present as strongly enhancing, sharply circumscribed lesions on T1-weighted images.

Based on a series of 28 patients, Kennedy et al proposed a classification system that identifies seven different classes according to the site of the tumor: intravestibular, intracochlear, intravestibulocochlear, transmodiolar, transmacular, transotic, and tympanolabyrinthine.⁴ Our case was of the intracochlear type. This is one of the most common types of intralabyrinthine schwannoma and constitutes 28% of all reported cases. In our case, the tumor was confined to the first and second cochlear turns. T2-weighted FSE images showed that the scala tympani of the anterior second turn was tumor-free. The ability of MRI images to differentiate the involvement of a single scala from another is particularly useful because

Figure 2 T1-weighted (A) and gadolinium-enhanced T1-weighted (B, C) images in the axial planes. There is a strongly enhancing schwannoma in the first and second cochlear turns.

it provides essential information for clinical observation.

The most important differential diagnosis is labyrinthitis. In labyrinthitis, the enhancement is often pronounced, but less sharp; often the complete cochlea or vestibular system is enhanced, and the enhancement becomes weaker and eventually disappears on follow-up. In intralabyrinthine schwannomas, the enhancement is most often restricted to part of the cochlea or vestibular system; the enhancement persists, and the lesion stays the same size or grows. In subacute or chronic labyrinthitis, fibrosis or calcification can replace the fluid, but the edges of the regions with signal intensity loss are rarely sharp.

The signal intensity of the lesion on the unenhanced T1-weighted images may be slightly higher than

that of normal fluid. The lesion enhances strongly after intravenous Gd administration, with sharply delineated edges. A sharply delineated signal intensity loss is also seen for the high-intensity intralabyrinthine fluid on the heavily T2-weighted image.

Although surgery has been the treatment of choice in two-thirds of the reported series, Kennedy et al,⁴ in 2004, stated that the best method of managing intralabyrinthine schwannomas confined to the labyrinth is serial MRI. In our intracochlear schwannoma, we adopted this rationale, confirming that the rate of growth of this pathology is low. Our personal diagnostic protocol is to perform serial MRI at a fixed follow-up. The appearance of other symptoms, particularly vestibular disturbances, would indicate planned MRI for the suspicion of growth of the tumor.

Surgery is reserved for intralabyrinthine schwannomas that extend into the internal auditory canal, cerebellopontine angle, or middle ear. The appearance of vertiginous symptomatology also necessitates surgical removal.

CONCLUSIONS

1. Intralabyrinthine schwannomas are rarely diagnosed.
2. High-resolution MRI with Gd enhancement is the diagnostic method of choice.
3. The majority of the patients can be managed with a wait-and-see strategy, utilizing planned serial MRI.
4. Surgery should be reserved only for intralabyrinthine schwannomas affected by intractable vestibular disturbances and involving the internal auditory canal, cerebellopontine angle, or middle ear.

REFERENCES

1. Brogan M, Chakeres DW. Gd-DTPA-enhanced MR imaging of cochlear schwannoma. *AJNR Am J Neuroradiol* 1990; 11:407–408
2. Casselman JW, Kuhweide R, Ampe W, Meeus L, Steyaert L. Pathology of the membranous labyrinth: comparison of T1- and T2-weighted and gadolinium-enhanced spin-echo and 3DFT-CISS imaging. *AJNR Am J Neuroradiol* 1993; 14:59–69
3. Jackson LE, Hoffmann KK, Rosenberg SI. Intralabyrinthine schwannoma: subtle differentiating symptomatology. *Otolaryngol Head Neck Surg* 2003;129:439–440
4. Kennedy RJ, Shelton C, Salzman KL, Davidson HC, Harnsberger HR. Intralabyrinthine schwannomas: diagnosis, management, and a new classification system. *Otol Neurotol* 2004;25:160–167
5. Tieleman A, Casselman JW, Somers T, et al. Imaging of intralabyrinthine schwannomas: a retrospective study of 52 cases with emphasis on lesion growth. *AJNR Am J Neuroradiol* 2008; 29:898–905