Neuroradiology (1998) 40: 684–689

J. F. Deux
K. Marsot-Dupuch
M. Ouayoun
P. Tran Ba Huy
J. M. Sterkers
B. Meyer
J. M. Tubiana

Slow-growing labyrinthine masses: contribution of MRI to diagnosis, follow-up and treatment

Received: 26 March 1997
Accepted: 27 January 1998

Work presented at SFR Congress, December 1996

J. F. Deux · K. Marsot-Dupuch (✉) · J. M. Tubiana
Service de Radiologie,
Hôpital Saint-Antoine, 184 rue du Faubourg Saint-Antoine, F-75012 Paris, France
Tel.: + 33-1-49.28.22.58
Fax: + 33-1-49.28.22.59

M. Ouayoun · B. Meyer
Service d'ORL, Hôpital Saint-Antoine,
184 rue du Faubourg Saint-Antoine,
F-75012 Paris, France

P. Tran Ba Huy
Service d'ORL, Hôpital Lariboisière,
2 rue Ambroise Paré, F-75010 Paris, France

J. M. Sterkers
4 Rue Michel Ange, F-75016 Paris, France

Abstract We report the use of MRI in the diagnosis, follow-up and therapeutic management of three cases of intralabyrinthine Schwannoma. The diagnosis was based on the history and initial and follow-up MRI findings. The main feature suggesting the diagnosis was a nodular intralabyrinthine mass of low signal intensity on T2-weighted images, and high or isointense signal on T1-weighted images (relative to cerebrospinal fluid), which showed contrast enhancement. Follow-up imaging showed growth of the tumour in one patient. One patient underwent surgery for severe tinnitus. To detect these lesions, MRI should be focussed on the inner ear, using thin-section T2-weighted and T1-weighted images before and after contrast medium. MRI allowed informed surgical planning.

Key words Ear, neoplasms · Temporal bone · Magnetic resonance imaging

Introduction

Patients with intralabyrinthine Schwannoma present with episodic vertigo, tinnitus and sensorineural hearing loss. However, they may have all or only some of these symptoms, Ménière's disease may be wrongly suggested. Until recently, these lesions were not detectable using conventional imaging techniques, and were discovered only at surgery or autopsy [1]. We present three patients with slow-growing labyrinthine masses to illustrate the role of MRI in diagnosis, follow-up, and management.

Materials and methods

Between 1991 and 1996, we examined 800 patients with sensorineural hearing loss, episodic vertigo or tinnitus, using MRI of the internal auditory meatus. Axial and coronal T1-weighted images were obtained before and after intravenous administration of 0.2 mmol/kg of gadolinium: 500/15/5 (repetition time/echo time/excitations); 3-mm-thick contiguous sections; matrix 512 × 256 half Fourier; field of view 250 mm. A constructive interference in steady state (CISS) sequence was also performed to investigate the inner ear: 20/8/1 and 50° flip angle; 1 mm section thickness; matrix 256 × 256; field of view 180 mm. The protocol was completed by computed tomography (CT) when abnormal signal in the labyrinth or abnormal morphology of the inner ear was observed.

Case reports

Case 1

A 68-year-old man had progressive worsening of right VIII nerve function over 23 years, starting with vertigo and tinnitus, which disappeared with the onset of sensorineural hearing loss at 2000 Hz. The patient became totally deaf in the right ear 16 years later. MRI revealed a labyrinthine mass affecting both the vestibule and the basal turn of the cochlea (Fig. 1). MRI was performed every 18 months; no growth of the lesion was observed after two follow-up examinations. The history suggested a lesion which was primarily vestibular and had extended towards the cochlea. The onset of hearing loss at 2000 Hz correlated with impairment of the basal turn of the cochlea. With the patient's agreement, no active treatment was given. Nevertheless, follow-up MRI is scheduled every 2 years.

Case 2

A 60-year-old woman had persistent, disabling tinnitus in the right ear for 18 years. About 6 months after the onset she had several attacks of vertigo. Although these attacks disappeared, ipsilateral sensorineural hearing loss appeared 6 years later, and tinnitus persisted. CT (Fig. 2 a) and MRI were thought to be normal. Some years later, MRI of the petrous bone showed a vestibular mass which gave slightly higher signal than the anterior labyrinth on T1-weighted images (Fig. 2 b). Marked contrast enhancement of the vestibule and the ampulla of the superior and lateral semicircular canals was observed (Fig. 2 c). Two years later, on T2-weighted images, the lesion was seen to give lower signal than cerebrospinal fluid (CSF); the signal of the posterior labyrinth had markedly decreased (Fig. 2 d, e). Due to the severity of the tinnitus, labyrinthectomy was performed. The MRI findings were confirmed at surgery: a tumour filled the vestibule and the ampulla of the lateral and superior semicircular canals, and extended to the basal turn of the cochlea. The remaining posterior membranous labyrinth was fibrotic. The tumour was easily removed in toto. Histological examination revealed a vestibular Schwannoma. Unfortunately, the patient's tinnitus was unchanged 6 months after removal of the lesion.

Case 3

A 23-year-old man had sensoneurinal hearing loss in the right ear, at a 500 Hz notch. T1-weighted images showed a high-signal nodular lesion at the posterior border of the second turn of the cochlea, which enhanced with gadolinium (Fig. 3 a). On T2-weighted images, the lesion gave lower signal than CSF (Fig. 3 b). The lesion was judged to be in the vicinity of the affected frequency band. The basal turn and apex of the cochlea were clear. MRI 2 years later, during an acute episode of total deafness and severe vertigo, showed enlargement of the apical cochlear tumour. The vestibule and lateral semicircular canal seemed larger (Fig. 3 c).

CT at the same time showed no abnormality of the bony labyrinth. As the patient no longer had vertigo, he refused to undergo labyrinthectomy. One year later, imaging showed a larger tumour extending to the apical turn, with abnormal contrast enhancement of the labyrinthine segment of the facial nerve, not present on the first MRI. The posterior labyrinth still appeared normal. Clinical examination revealed complete loss of inner ear function with unilateral deafness and vestibular arreflexia on caloric testing.

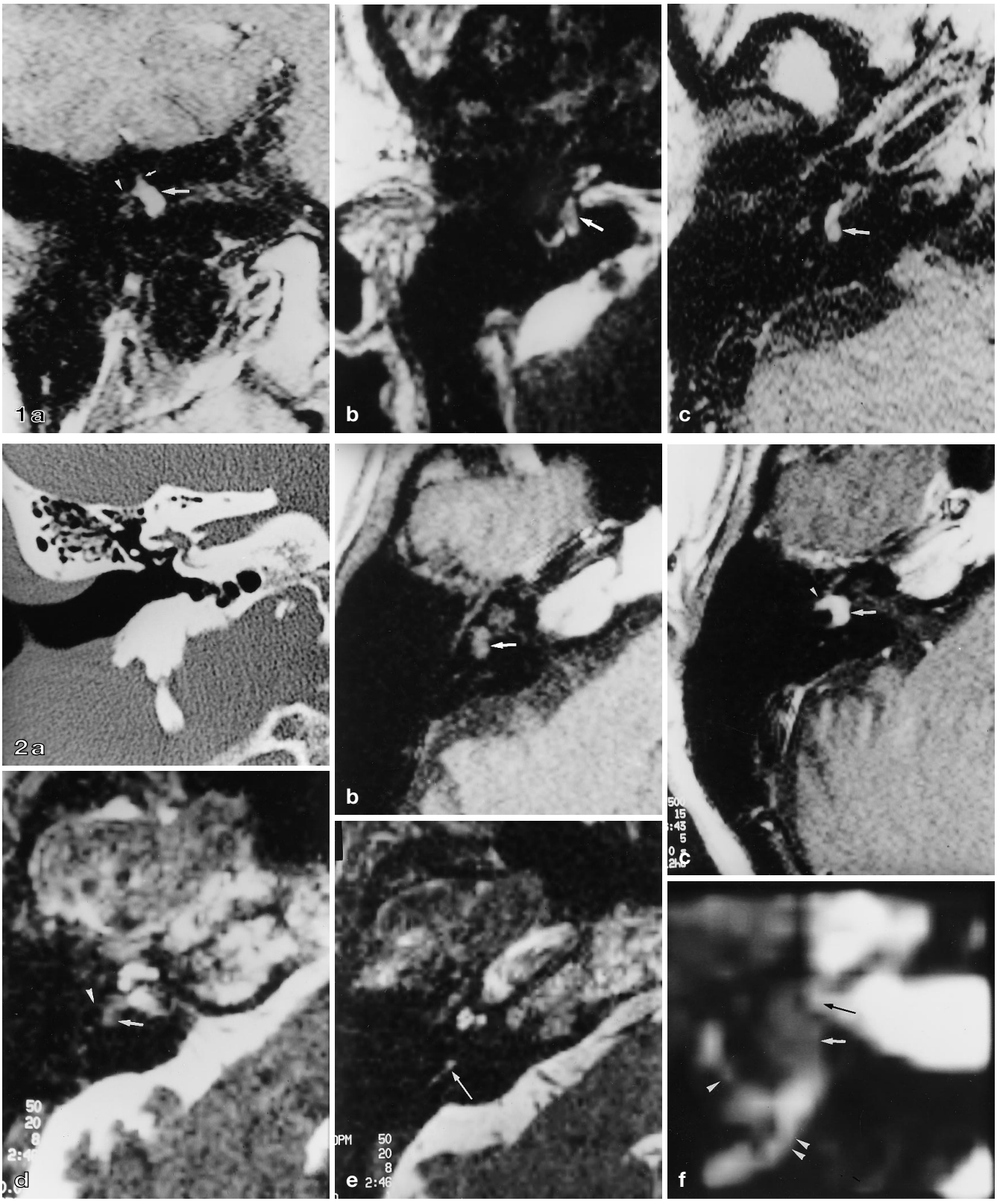
Discussion

Labyrinthine masses are usually cochleovestibular Schwannomas. Nevertheless, Huang [2] reported an intravestibular lipoma. There is another report of labyrinthine tumours in patients with metastatic meningoradiculitis [1].

Cochleovestibular Schwannomas are benign tumours resulting from the proliferation of Schwann cells. They typically arise in the internal auditory meatus (IAM) and invade the vestibular and, less often, the cochlear nerves [1]. Their estimated prevalence is 1 % in autopsy series [3, 4]. Schwannomas may arise within the labyrinthe: 1 case in 893 petrous bone examinations was described in an autopsy series (prevalence 0.1 %) [5]. The 0.4 % prevalence in our study was probably due to enrolment bias. Analysis of the 19 previously reported cases in the English-language literature up to 1996 indicates that 10 tumours arose on the vestibular nerve fibres, 8 on the cochlear nerve fibres and the site of origin of one was unknown (Table 1) [6–18]. The sex ratio was 1 : 1. The mean age of the patients was 49 years (range 7–84 years). The patients were not known to have neurofibromatosis type 2, except one with bilateral lesions.

Cochlear Schwannomas tend to occur in the basal turn (scala tympani) and modiolus [3] a feature explained by the pathway of dendrites of the spiral ganglion in the modiolus. Growth in the scala tympani is thought to be due to the proximity of the basilar membrane, which is adjacent to the neurosensorial end area [19]. From the scala tympani, these tumours may then extend to the scala vestibuli [3]. To our knowledge, no case of isolated Schwannoma in the cochlear duct has been reported. Loss of spiral ganglion cells, especially in the basal turn, appears universal [3].

Vestibular Schwannomas may arise at any point along the vestibular nerve, including its distal branches [4] or at the end adjacent to the ampullae of the semicircular canals [5, 16]. Vestibular Schwannomas may grow to fill the vestibule and one or more of the semicircular canals. The labyrinth, middle ear and IAM are the main extension pathways for labyrinthine Schwannomas. Spread within the labyrinthe is progressive, with destruction of neurones and secondary atrophy of the ganglia. Within the labyrinth, cochlear and vestibular tumours progressively extend up to the region of the utriculosaccular valve or impair its function by causing distant fibrotic areas [11, 12, 17, 18]. Obstruction may induce endolymphatic hydrops [11, 12]. Other hypotheses proposed to explain intermittent endolymphatic hydrops include release of potassium ions or proteins secreted by the tumour [11], and damage to Reissner's membrane or the stria vascularis. Endolymphatic hydrops may cause intermittent vertigo or hearing loss without neoplastic invasion of the adjacent compartment [11, 12].


1a
b
c
2a
b
c
d
e
f

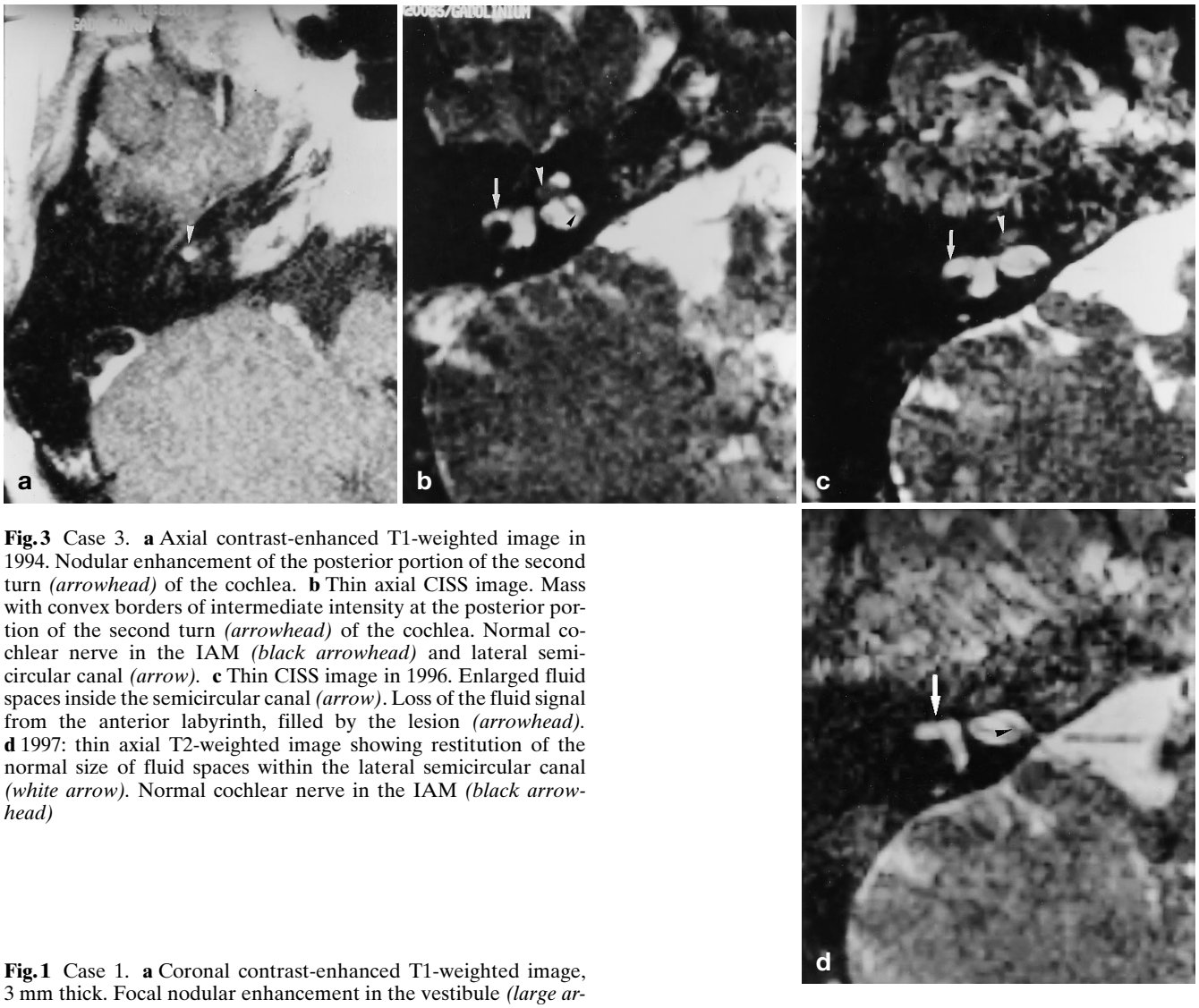


Fig. 3 Case 3. **a** Axial contrast-enhanced T1-weighted image in 1994. Nodular enhancement of the posterior portion of the second turn *(arrowhead)* of the cochlea. **b** Thin axial CISS image. Mass with convex borders of intermediate intensity at the posterior portion of the second turn *(arrowhead)* of the cochlea. Normal cochlear nerve in the IAM *(black arrowhead)* and lateral semicircular canal *(arrow)*. **c** Thin CISS image in 1996. Enlarged fluid spaces inside the semicircular canal *(arrow)*. Loss of the fluid signal from the anterior labyrinth, filled by the lesion *(arrowhead)*. **d** 1997: thin axial T2-weighted image showing restitution of the normal size of fluid spaces within the lateral semicircular canal *(white arrow)*. Normal cochlear nerve in the IAM *(black arrowhead)*

◀ **Fig. 1** Case 1. **a** Coronal contrast-enhanced T1-weighted image, 3 mm thick. Focal nodular enhancement in the vestibule *(large arrow)* and enhancement in the ampullae of the superior *(thin arrow)* and lateral *(arrowhead)* semicircular canals. **b** Thinsection T2-weighted axial image. Loss of signal of vestibular fluid *(arrow)*. **c** Axial contrast-enhanced T1-weighted image. Focal nodular enhancement of the basal turn of the cochlea due to spread of the tumour *(arrow)*

Fig. 2 Case 2. **a** Thin coronal CT through oval window. Normal vestibule and lateral semicircular canal. **b** Axial contrast-enhanced T1-weighted image. Sightly increased vestibular signal *(arrow)* relative to the anterior labyrinth. **c** Axial contrast-enhanced T1-weighted image. Enhancement of the vestibule *(arrow)* and ampulla of the lateral semicircular canal *(arrowhead)*. **d** Thinsection axial T2-weighted CISS image. Intermediate signal from vestibular mass *(arrow)*, with signal loss in the endo- and perilymphatic fluid of the lateral semicircular canal *(arrowhead)*. **e** Section 2 mm lower. High signal in the posterior semicircular canal *(arrow)*. **f** CISS image. Vestibular mass with convex borders *(white arrow)* extending to the ampulla of the lateral semicircular canal. Loss of signal in the superior *(white arrowhead)* and posterior *(double arrowheads)* semicircular canals. Normal high signal from the vestibular nerve canal *(black arrow)* and anterior labyrinth

The tumour may extend through the round or oval window into the middle ear [14]. Extension toward the oval window may lead to fixation of the stapes and cause conductive hearing loss [17]. The differential diagnosis of an isolated soft tissue mass in the middle ear located near the oval or round window is wide; MRI should therefore be performed before surgical exploration.

Labyrinthine Schwannoma may extend through the porus acusticus toward the IAM. Cochlear tumours occur along the cochlear nerve in the inferoanterior segment, and vestibular tumours along the upper or lower vestibular nerves [20]. Isolated tumours occurring simultaneously in the labyrinth and the acusticovestibular nerve have been reported in neurofibromatosis type 2 [17]; MRI clearly showed a separation between the labyrinthine tumour and invasion of the acusticovestibular nerve [21].

Table 1 Review of 22 patients with intralabyrinthine tumours

Reference	Age (years)/sex	Hearing loss (Hz)	Vertigo	Tinnitus	Diagnosis		Origin	Spread
					Initial	Final		
18	58 F	Global	–	+	?	Schwannoma[a]	Basal turn of cochlea	Modiolus
17	23 M	Global	+	–	?	Schwannoma[c]	Internal auditory meatus	–
		Global[b]	–	+	?	Schwannoma	Vestibule, internal auditory meatus	Oval window
15	40 M	+[d]	–	+	Ménière's disease	Schwannoma	Vestibule	–
16	38 F	4–8000	+	+	Deafness	Schwannoma[c]	Vestibule	Basal turn
14	70 F	+	+	–	Ménière's disease	Schwannoma[c]	Cochlea, vestibule	–
5	67 F	?	?	?	?	Schwannoma[a]	Ampulla of lateral semicircular canal	–
13	73 M	4–8000	+	–	Ménière's disease	Schwannoma[c]	Vestibule	Basal turn
11	20 M	4000	+	+	Ménière's disease	Schwannoma[c]	Vestibule	Basal turn
	38F	Global	+	+	Ménière's disease	Schwannoma[c]	Basal turn of cochlea	–
12	51 M	1–2000	+	+	Ménière's disease	Schwannoma[a]	Basal turn of cochlea	Modiolus
3	74 M	2–4000	+	+	?	Schwannoma[a]	Basal turn of cochlea	–
	84 F	?	–	–	?	Schwannoma[a]	Vestibule	–
10	54 M	?	–	–	?	Schwannoma[a]	Basal turn of cochlea	–
9	57 F	+	+	+	Ménière's disease	Schwannoma[c]	Ampulla of superior semicircular canal	Vestibule
8	41 M	+	+	+	Ménière's disease	Schwannoma[c]	Vestibule	–
7	36 F	+	+	+	Tumour	Schwannoma[c]	First turn of cochlea	Basal turn
4	69 M	Global	–	+	?	Schwannoma[c]	Cochlea	Vestibule, Round window
	7 F	+	–	–	Cholesteatoma	Schwannoma[c]	Cochlea	Round window, middle ear
6	22 M	+	+	–	Tumour	Schwannoma[c]	Vestibule	Ampulla
This	23 M	500	–	–	Tumour	?	Second turn of chochlea	
report	60 F	Global	+	+	Tumour	Schwannoma[c]	Vestibule	
	68 M	Global	+	+	Tumour	?	Vestibule, basal turn of cochlea	

[a] Autopsy confirmation
[b] Bilateral tumours
[c] Surgical confirmation
[d] Range not stated

In our experience, the notch of the sensorineural hearing loss correlated with the site of the tumour: high-frequency loss with tumours in the basal turn, and low-frequency deficits with tumours in the second turn and apex. It is difficult to establish a correlation between the site of the tumour and hearing loss from the literature. As the cochlear tumour grows, hearing is lost over all frequencies.

Labyrinthine Schwannomas do not cause bone destruction [3]; CT does therefore not contribute to their diagnosis except when they extend into the middle ear. As standard MRI studies without thin T2- and T1-weighted sections may appear normal, detection of labyrinthine tumours requires high-resolution imaging of the inner ear, including both T2-weighted, and T1-weighted images before and after gadolinium. CISS or fast spin-echo (FSE) sequences susceptible to changes in the signal from endo- and perilymphatic fluid give thin-section T2-weighted images with adequate spatial resolution.

Diagnosis is based upon a localized soft tissue intralabyrinthine mass, of low signal intensity on T2-weighted images and of moderate or high signal intensity on T1-weighted images, enhancing after gadolinium.

References

1. Schuknecht HF (1993) Pathology of the ear. 2nd edn Lea and Febiger, Boston, pp 460–473
2. Huang TS (1989) Primary intravestibular lipoma. Ann Otol Rhinol Laryngol 98: 393–395
3. Babin RW, Harker LA (1980) Intralabyrinthine acoustic neurinomas. Otolaryngol Head Neck Surg 88: 455–461
4. Mafee MF, Lachenauer CS, Kumar A, Arnold PM, Buckingham RA, Valvassori (1990) CT and MR imaging of intralabyrinthine Schwannoma: report of two cases and review of the literature. Radiology 174: 395–400
5. Stewart TJ, Liland J, Schuknecht HF (1975) Occult schwannomas of the vestibular nerve. Arch Otolaryngol 101: 91–95
6. Saeed SR, Birzgalis AR, Ramsden RT (1994) Intralabyrinthine schwannoma shown by magnetic resonance imaging. Neuroradiology 36: 63–64
7. Brogan M, Chakeres DW (1990) Gd-DTPA-enhancement MR imaging of cochlear schwannoma. AJNR 11: 407–408
8. Huang TS (1986) Primary intralabyrinthine schwannoma. Ann Otol Rhinol Laryngol 95: 190–192
9. Vernick DM, Graham MD, McClatchey KD (1984) Intralabyrinthine schwannoma. Laryngoscope 94: 1241–1243
10. Johnsson LG, Kingsley TC (1981) Asymptomatic intracochlear neurinoma. Arch Otolaryngol 107: 377–381
11. De Lozier HL, Gacek RR, Dana ST (1979) Intralabyrinthine schwannoma. Ann Otol 88: 187–191
12. Gussen R (1971) Intramodiolar acoustic neurinoma. Laryngoscope 81: 1979–1984
13. Weymuller EA (1975) Unsuspected intravestibular schwannoma. Arch Otolaryngol 101: 630–632
14. Storrs LA (1974) Acoustic neurinomas presenting as middle ear tumours. Laryngoscope 84: 1175–1180
15. Wanamaker HH (1972) Acoustic neuroma: primary arising in the vestibule. Laryngoscope 82: 1040–1044
16. Karlan MS, Basek M, Potter GB (1972) Intracochlear neurilemmoma. Arch Otolaryngol 96: 573–575
17. Hallpike CS (1963) Stapes fixation by an intralabyrinthine “seedling” neurofibroma as a cause of conductive deafness in a case of von Recklinghausen’s disease. Acta Otolaryngol (Suppl) 183: 62–65
18. Jorgensen MB (1962) Intracochlear neurinoma. Acta Otolaryngol 54: 227–232
19. Daniels DL, Swartz JD, Harnsberger HR, Ulmer JL, Shaffer KA (1996) Anatomic moment. Hearing I: the cochlea. AJNR 17: 1237–1241
20. Brizon J, Castain J (1953) Les feuillets d’anatomie, ostéologie de la tête. Fascicule X. 1ère partie. edn Maloine, Paris, p. 50
21. Mafee MF, Dada S, Villanueva E, Camras LR, Schmelka DR (1996) MR imaging in evaluation of intralabyrinthine pathology. Radiology 201: 457