

Intralabyrinthine schwannoma shown by magnetic resonance imaging

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Abstract. Intralabyrinthine schwannomas are rare benign tumours which present with progressive or fluctuant audiovestibular symptoms and may mimic Menière's disease. The size and position of these lesions make preoperative diagnosis unusual and most are discovered incidentally at labyrinthectomy. A case is reported which was diagnosed on magnetic resonance imaging and confirmed at surgery.

Key words: Acoustic neuroma – Magnetic resonance imaging – Temporal bone tumours

The vestibular schwannoma (acoustic neuroma) is a benign tumour which usually arises from the superior vestibular nerve within the internal auditory meatus (IAM), although it may be found at any site along the nerve, including its distal branches [1, 2]. Tumours originating within the labyrinth are rare, even in large series [3–5]. In only one of the eight clinical reports in the world literature [6–14] was the tumour diagnosed preoperatively: Karlan et al. [7] were able to show a nonspecific abnormality (loss of normal radiolucency of the first cochlear turn) with temporal bone polytomography. We report the first intralabyrinthine schwannoma confidently diagnosed on magnetic resonance imaging (MRI) and confirmed at surgery.

Case report

A 22-year-old man presented complaining of left-sided hearing loss progressive over the previous five years and intermittent rotatory vertigo. There were no other otological symptoms and a full physical examination was normal. A pure tone audiogram showed profound sensorineural hearing loss on the left side with normal contralateral thresholds.

MRI scan with gadolinium DTPA demonstrated a small enhancing lesion within the left bony labyrinth, quite clearly separated from a normal internal auditory canal by a bony septum (Fig. 1).

At operation, a routine three-canal bony labyrinthectomy was carried out. A pinkish soft tissue mass was encountered occupying the anterior half of the vestibule, adjacent to the anterior crus and extending into the ampullated end of the posterior crus of the lateral semicircular canal (Fig. 2). It was easily removed in toto (Fig. 3). The medial wall of the vestibule (lateral end of the internal auditory meatus) was intact; the IAM was not opened. The patient made a complete recovery, with no further episodes of dizziness.

Histological examination of the tumour confirmed the diagnosis of schwannoma.

Discussion

From the limited number of reports describing this unusual tumour, it appears it usually causes symptoms similar to those of "burnt-out" Menière's disease. Despite the profound loss of cochlear and vestibular function, most patients continue to have severe episodic vertigo. Indeed, had they become asymptomatic, the labyrinthectomy at which the tumour was almost always a chance finding, would not have been performed. The standard battery of audiovestibular tests used in the diagnosis of the classical IAM schwannoma is usually unable to localise the intralabyrinthine lesion and the diagnosis has been dependent upon the clinician's decision to perform a labyrinthectomy for relief of vertigo [13]. In most cases, a transmastoid approach is adequate, although the surgeon must be prepared to open the internal auditory meatus (IAM) if there is evidence of medial extension [14].

With recent advances in neuroradiology, diagnosis of small vestibular schwannomas has been made possible, allowing the neuro-otologist to remove them at an early stage in the disease process, with a significant reduction in associated morbidity. MRI is now established as the investigation of choice in the detection of vestibular schwannomas [15], especially with contrast enhancement [16]. As MRI becomes more widely available and more patients are examined, many more of these primary intralabyrinthine tumours will presumably be diagnosed prior to surgery.

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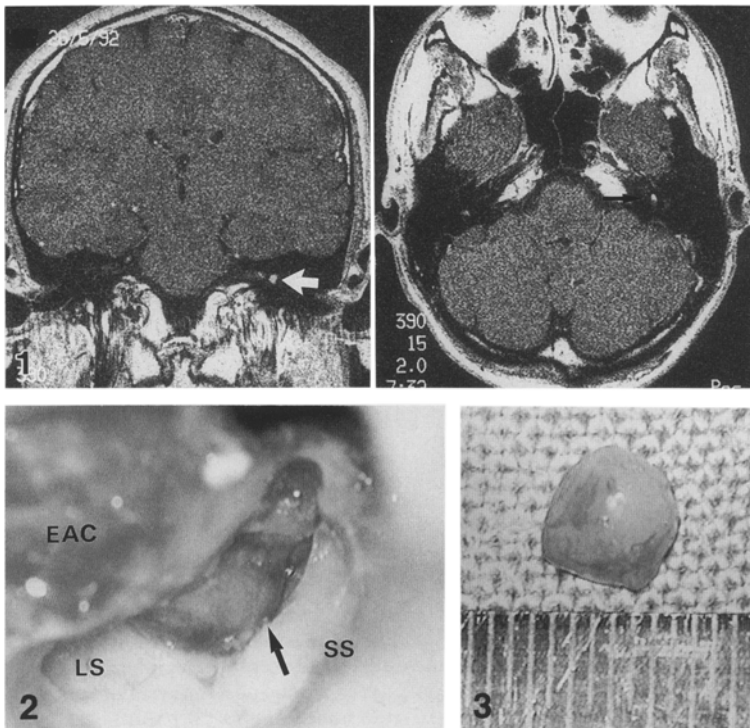


Fig. 1. Coronal and axial MRI with Gd-DTPA (T1-weighted spin-echo images). Discrete enhancing lesion within the left otic capsule (*arrow*)

Fig. 2. Operative view of the tumour (*arrow*) arising from the left labyrinthine vestibule. *LS* Lateral semi-circular canal *SS* Superior semicircular canal *EAC* External auditory canal

Fig. 3. Surgical specimen removed intact from the labyrinthine vestibule. Each gradation represents 1 mm

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