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Intralabyrinthine schwannoma

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Abstract
Intralabyrinthine schwannomas are rare tumours which present with symptoms similar to Menière's disease. Pre-operative diagnosis is rarely possible and most are found incidentally during labyrinthectomy for persistent vertigo. A further case of this tumour is reported together with a review of the literature.

Introduction

The acoustic schwannoma is a benign tumour which usually arises from the superior vestibular division of the eighth cranial nerve within the internal auditory meatus (IAM). However, it is well recognized that these tumours may arise from any site along the nerve, including its distal branches (Nager, 1969; Skinner, 1929). The labyrinth may be involved by lateral extension from the IAM, although true primary intralabyrinthine tumours have been reported in association with von Recklinghausen's disease (Gray, 1933; Scott, 1938; Nager, 1964) and as incidental findings during temporal bone studies, where the lesion seems to arise principally from the basal turn of the cochlea (Jorgensen, 1962; Gussen, 1971; Stewart *et al.*, 1975).

With the advent of more sophisticated imaging for early diagnosis of acoustic schwannomas and the development of the safe translabyrinthine surgical approach (House, 1970), clinical reports of intralabyrinthine tumours have appeared. Wanamaker described the first such case in 1972, suggesting that it arose from distal branches of the superior vestibular nerve

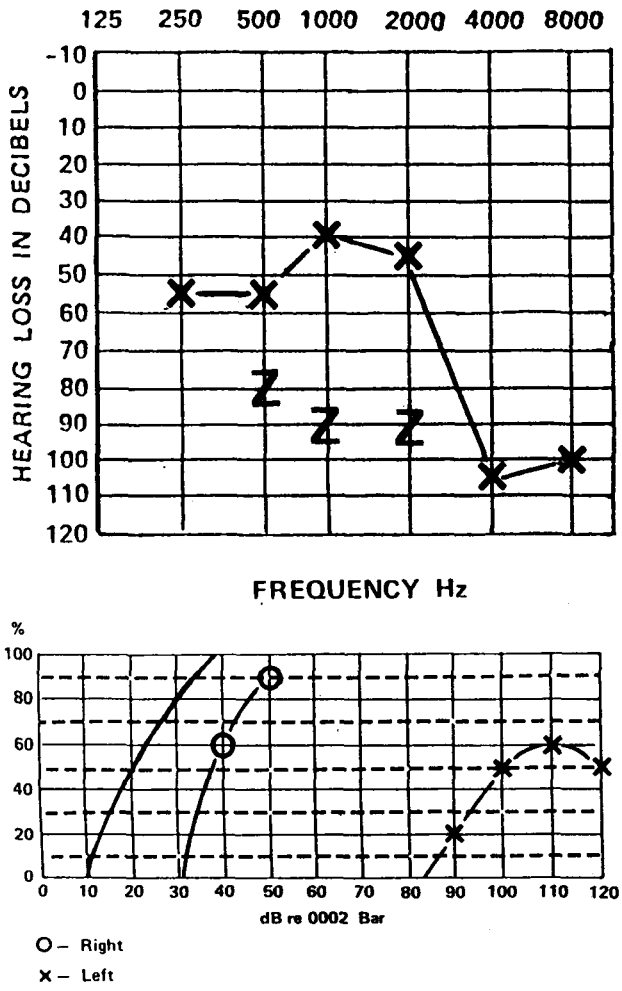

Fig. 1
Pre-operative pure-tone and speech audiograms.

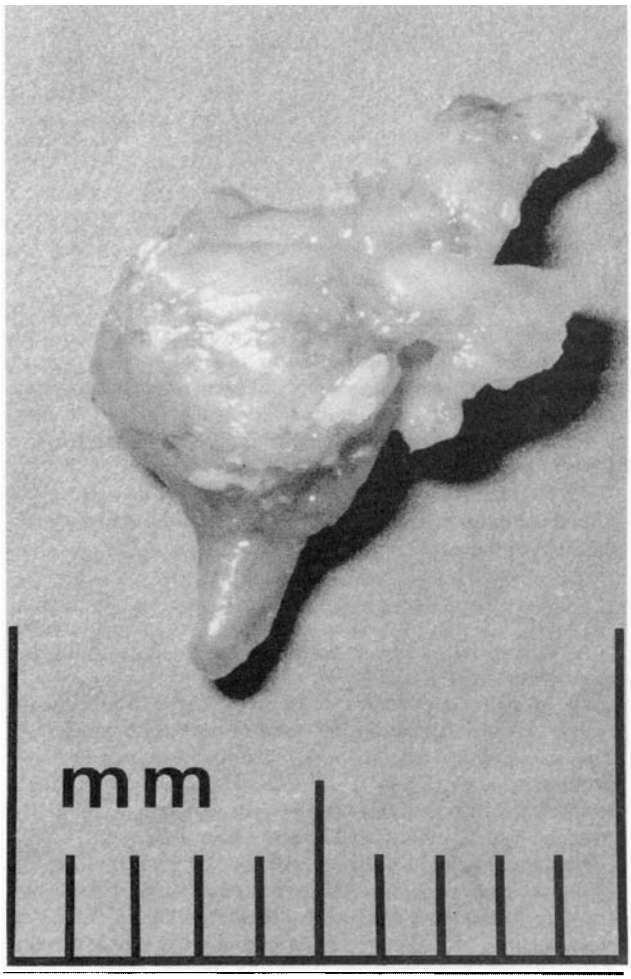

Fig. 2
Sugical specimen removed from the labyrinthine vestibule during transmastoid labyrinthectomy. Note the small extension which was located within the lateral semicircular canal.

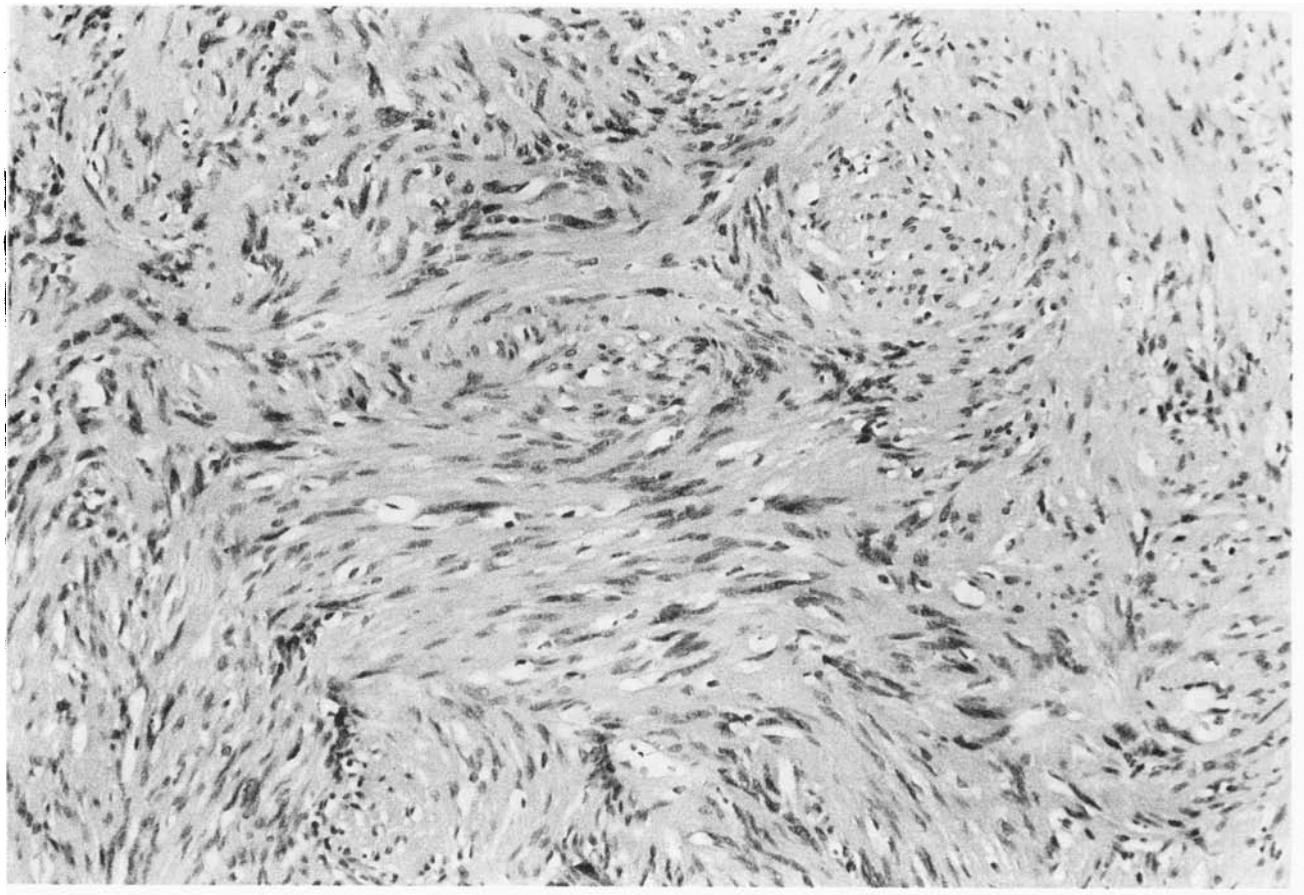

FIG. 3

Photomicrograph of tumour showing bundles of spindle cells with nuclear pallisading and occasional Verocay bodies. Appearances typical of an acoustic schwannoma (H and E × 42.5)

adjacent to the utricle or saccule. Since then, seven further cases have been documented and a clearer picture of the clinical features of this unusual lesion has emerged. As most patients present with a long history of cochlear deficit together with persistent vertigo, resembling atypical Menière's disease, the tumour is often found incidentally during destructive labyrinthectomy.

An interesting case is presented here of a primary intravestibular schwannoma which was encountered during transmastoid labyrinthectomy.

Case report

A 44-year-old male was admitted for a left transmastoid labyrinthectomy.

He presented eleven years earlier to the ENT department with a two year history of intermittent positional vertigo. At that time, physical examination, full audiometry, serology and pretrous tomography were all normal, although there was a reduced caloric response on the left side. His symptoms settled over the next 12 months and he was lost to follow-up.

Nine years later, he returned with a gradual onset of left tinnitus, deafness, occasional otalgia and disabling vertigo. Investigation confirmed a left canal paresis, unilateral recruiting sensorineural deafness, normal stapedial reflex thresholds and poor speech discrimination (Fig. 1). Brain Stem Evoked Responses were inconsistent on that side and computed tomography showed no abnormality. A diagnosis of Menière's disease was made.

At operation, a pink soft tissue mass was found within the vestibule, extending into the lumen of the left lateral and superior semicircular canals with no apparent cochlear involvement. This was easily removed and no abnormality was found on surgical exposure of the IAM (Fig. 2). Histological sections of the lesion showed typical features of an acoustic schwannoma (Fig. 3). The patient made an uneventful post-operative recovery and two years later is symptom-free and in good general health.

Discussion

Historically, the first case of an intralabyrinthine schwannoma was described by Mayer (1917) in a patient with von Recklinghausen's disease. Since then, a number of temporal bone studies have demonstrated such lesions in patients without this disease. Unlike acoustic schwannomas in the IAM, which usually arise from the superior vestibular nerve, those of the labyrinth seem to have a predilection for the cochlear division, often arising from the basal turn of the cochlea (Leonard and Talbot, 1970; Gussen, 1971; Hoshino and Ishii; 1972, Stewart *et al.*, 1975).

Wanamaker and Karlan (1972) both described intralabyrinthine schwannomas found unexpectedly at labyrinthectomy, whilst Storrs (1974) encountered a similar lesion extending into the middle ear. Further cases have been reported by other authors (Weymuller, 1975; De Lozier *et al.*, 1979; Miyamoto *et al.*, 1980; Vernick *et al.*, 1984; Huang, 1986) and in most of these, the patients presented a history suggestive of Menière's disease with unilateral hearing loss, tinnitus and disabling vertigo. Common features have been a reduced or absent caloric

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response, poor speech discrimination and inconsistent or normal Brain Stem Evoked Responses. In only one case (Karlan, 1972) was preoperative radiological localisation possible and most lesions were found incidentally during transcanal labyrinthectomy (Wanamaker, 1972; Weymuller, 1975; De Lozier *et al.*, 1979); transmastoid labyrinthectomy (Miyamoto *et al.*, 1980; Huang, 1986) or middle ear exploration (Storrs, 1974).

The case we have described filled the labyrinthine vestibule and extended into the semicircular canals with no surgical evidence of IAM involvement. The initial symptom of positional vertigo with normal hearing, suggests an origin from the region of the utricle, which contrasts with most other reported cases. Subsequent cochlear deficit may have resulted from an alteration in endolymphatic flow. As the clinical features may suggest atypical Menière's disease with an absent or reduced caloric response and poor speech discrimination, a high index of suspicion is required in all such patients. Although the standard battery of pre-operative tests for the early diagnosis of IAM schwannomas fails to localise these intralabyrinthine tumours, high resolution CT or magnetic resonance imaging of the cochlea may be helpful in suspicious cases.

We feel that a transmastoid approach to the labyrinth is more than adequate for these lesions as it allows complete tumour removal, provides access to the IAM and relieves the patient's sysmptoms.

Conclusions

The intralabyrinthine acoustic schwannoma is a rare lesion which mimics atypical Menière's disease and defies attempts at pre-operative diagnosis. Its removal during transmastoid labyrinthectomy should be followed by exposure of the internal auditory canal.

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