
CASE REPORT

Intralabyrinthine Schwannoma: an Uncommon but Underdiagnosed Entity

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ABSTRACT

Intralabyrinthine schwannoma is an uncommon tumour arising from the vestibulocochlear nerve within the cochlear, vestibule, or semicircular canals. Intralabyrinthine schwannomas are classified according to the location, and 'intracochlear' is the commonest subtype. Patients frequently present with progressive hearing loss. Imbalance or vertigo are also common presentations. On magnetic resonance imaging, the tumour will be shown as a filling defect of the labyrinth on T2-weighted images. Intense contrast enhancement is also evident. Intralabyrinthine schwannoma was believed to be under-reported in the past, but with recent advancements in magnetic resonance imaging technology, sensitivity for the detection of this tumour has increased. Radiologists should be aware of intralabyrinthine schwannoma when screening patients with hearing or balance disorders.

Key Words: Hearing loss; Neuroma, acoustic; Vertigo; Vestibule, labyrinth

中文摘要

迷路內神經鞘瘤：一種罕見的但屬診斷不足的病症

吳業軒、盧承迅

迷路內神經鞘瘤是起源於耳蝸、前庭或半規管內的前庭蝸神經的一種罕見腫瘤。根據病變部位分類，「耳蝸內」是最常見的迷路內神經鞘瘤亞型。患者一般會出現進行性聽力受損，而平衡失調或眩暈亦是常見的臨床表現。腫瘤在磁共振T2加權像上顯示為迷路充盈缺損，亦會有明顯的對比增強。以往迷路內神經鞘瘤病例被認為報導不足，但隨着磁共振成像技術近來的進步，對此腫瘤的檢測靈敏度有所增加。放射科醫生遇到聽力受損或平衡失調的病人時，應意識到可能是患有迷路內神經鞘瘤。

INTRODUCTION

Vestibular schwannoma is the most common neoplasm of the internal auditory canal (IAC) and cerebellopontine angle. Schwannoma is a benign tumour of the nerve sheath, most often arising from the vestibular division

of the vestibulocochlear nerve. Intralabyrinthine schwannomas (ILSs) are defined as tumours arising primarily from the terminal vestibulocochlear nerve within the membranous labyrinth in the cochlear, vestibule, or semicircular canals.

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Submitted: 30 Jul 2012; Accepted: 11 Sep 2012.

ILS has been considered a rare lesion in the past. In 1917, the first ILS was found during autopsy.¹ During the 1970s, ILSs were found during labyrinthectomy performed because of intractable vertigo or Menière's disease.² In 1990, the first cases of ILS were described on magnetic resonance imaging (MRI).³ Since then, due to rapidly increasing use of MRI, together with improving imaging techniques and resolution, an increasing number of ILSs have been reported. ILSs are now considered a pathology that is much more common than had been previously thought.

CASE REPORT

A 38-year-old woman presented to the Department of Otorhinolaryngology, United Christian Hospital, Hong Kong, in January 2011. She had had vertigo for 6 months, which was associated with tinnitus in the left ear. The vertigo was position-associated and became more severe when the patient turned left. She

reported sudden hearing loss in her left ear 1 week earlier. Otoscope examination was normal, as were the neurological examination and balance test. Pure tone audiometry detected left-side low-tone sensorineural hearing loss with a threshold of 50 decibels. She was followed up as an outpatient for 1 year, and the symptoms persisted but remained stable. The provisional diagnosis was Menière's disease.

She then underwent gadolinium-enhanced MRI using axial and coronal slices in spin-echo T1- and T2-weighted sequences. A mass was identified, confined at the vestibule in T2-weighted sequence, as a filling defect replacing the normal high-signal fluid density (Figure 1). On T1-weighted images, the lesion had slightly higher signal intensity than the normal intralabyrinthine fluid on unenhanced images. Homogeneous and intense enhancement was observed after administration of gadolinium contrast (Figure 2). There was no mass

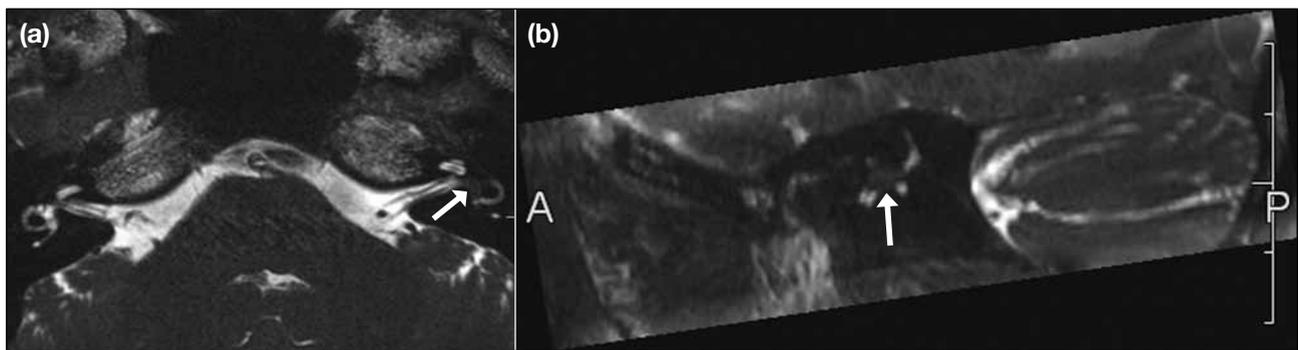


Figure 1. T2-weighted magnetic resonance images of the vestibule: (a) axial and (b) oblique sagittal images show a hypointense filling defect confined to the left vestibule (arrows) replacing the normal intense intralabyrinthine fluid signal, representing the intravestibular intralabyrinthine schwannoma. There is no extension into the internal auditory canal.

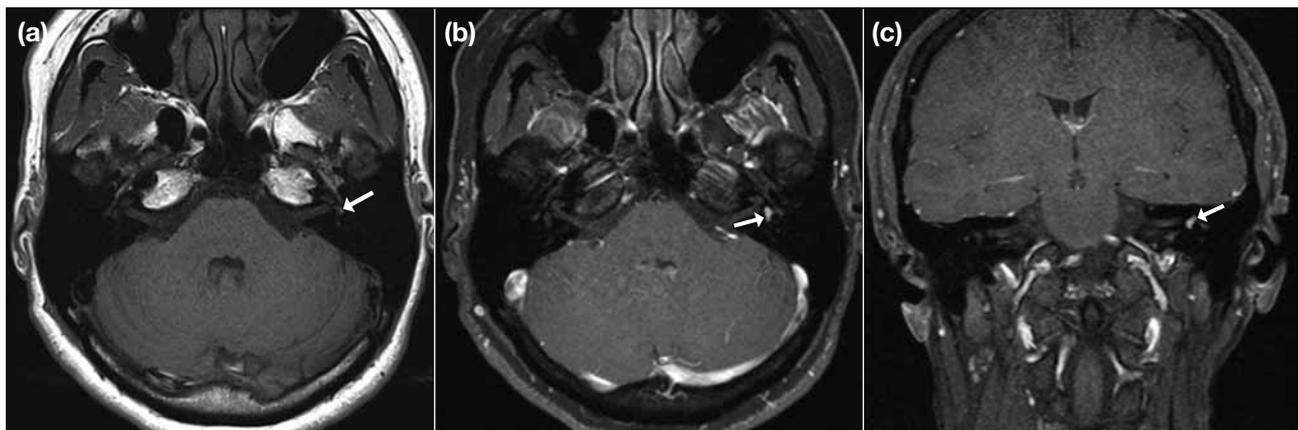


Figure 2. T1-weighted magnetic resonance images of the vestibule: (a) axial non-enhanced image shows a vague left intravestibular lesion with slightly higher signal intensity than the normal intralabyrinthine fluid (arrow); (b) enhanced axial and (c) enhanced coronal images show homogeneous and intense enhancement of the lesion (arrows), representing the intralabyrinthine schwannoma.

formation over the IAC and the cerebellopontine angle. This enhancing lesion was suggestive of ILS of the vestibule. It was decided to monitor the lesion by serial annual MRI evaluation.

DISCUSSION

ILS is an uncommon disease of the inner ear. To date, its prevalence is still uncertain. The figure quoted in a study in the 1970s was 0.1%, which was a single case found among 893 petrous bone autopsies.⁴ A study in 1998 estimated the prevalence to be 0.4%, based on three cases of ILS found by MRI in 800 patients with Menière-like symptoms.⁵ However, it is apparent that with recent advances in MRI technology with increased image resolution, sensitivity of detection of ILS has increased. More cases have been reported, and prevalence figures have started to rise. A large series of 52 cases by Tieleman et al⁶ reported a prevalence of 10.8%. However, the study was conducted in a centre specialising in temporal bone imaging, which may reflect a selection bias.

The most frequent presenting symptom is unilateral hearing loss, which is almost a universal symptom in ILS. The loss of hearing is usually progressive, with a smaller number of patients presenting with sudden hearing loss. This patient also presented with vertigo. In a series of 28 patients reported by Kennedy et al,⁷ 46% had balance disorders, but only 25% had typical vertigo crises. Tumours that are confined to the vestibule are more likely to cause vertigo or symptoms similar to Menière's disease.⁸

ILSs are classified according to the location. 'Intracochlear' ILS is defined as a tumour that is confined to the cochlear turns, and is the commonest subtype. 'Intravestibular' ILS refers to a tumour confined to the vestibule with or without involvement of the semicircular canals. This patient had this subtype of ILS. If the tumour fills both the cochlea

and vestibule, it is an 'intravestibulocochlear' ILS. 'Transmodiolar' ILS is defined as a tumour extending through the modiolus from the cochlear into the IAC. 'Transmacular' ILS is defined as a tumour extending through the macula cribrosa from the vestibule into the IAC. 'Tympanolabyrinthine' ILS refers to a tumour extending from the vestibule or cochlear into the middle ear. Finally, if either the vestibule or cochlear or both, together with the middle ear and the IAC are all involved, the tumour will be described as a 'transotic' ILS. The above definitions are summarised in the Table.

MRI with contrast is the modality of choice for the diagnosis of ILS. On T2-weighted images, ILS appears as a sharp focal filling defect replacing the normal high-signal intensity intralabyrinthine fluid. The lesion is usually more difficult to identify in T1-weighted unenhanced images, although some prior reports concluded that a higher signal intensity than the intralabyrinthine fluid may give a clue to the diagnosis.⁶ Nevertheless, the lesion will be shown as an intense homogeneously enhancing mass on gadolinium administration.

The most important differential diagnosis for ILS is labyrinthitis. On MRI, labyrinthitis commonly enhances diffusely, involving the complete cochlear and / or vestibular system. Even if focal, their borders are often blurred. No focal filling defects or loss of signal will be seen in T2-weighted images. The enhancement will also eventually disappear on follow-up imaging.

Labyrinthitis ossificans, during its fibro-osseous phase, can also show T2 hyperintensity and contrast enhancement. Differentiation is possible by computed tomography, where bony encroachment on the membranous labyrinth would be a feature.⁹ Moreover, a previous history of meningitis or suppurative otomastoiditis will point towards this diagnosis. Haemorrhage and lipoma are differential

Table. Classification of intralabyrinthine schwannoma based on location.

Classification	Cochlear	Vestibule ± SCC	Cochlear or vestibule	IAC	Middle ear
Intracochlear	✓				
Intravestibular		✓			
Intravestibulocochlear	✓	✓			
Transmodiolar	✓			✓	
Transmacular		✓		✓	
Tympanolabyrinthine			✓		✓
Transotic			✓	✓	✓

Abbreviations: SCC = semi-circular canals; IAC = internal auditory canal.

diagnoses, and can be readily differentiated on T1-weighted images, appearing as hyperintense areas without contrast enhancement.¹⁰

Serial MRI monitoring of the tumour will usually be adequate for most ILSs. The proposed frequency is 1 year after first detection, followed by 2-year intervals if the first follow-up MRI shows a static tumour size.⁷ Since surgical resection of the intralabyrinthine tumour will sacrifice ipsilateral hearing, surgery is only indicated for patients with intractable vertigo, tumour extension to the cerebellopontine angle, or evidence of tumour growth on serial follow-up imaging.¹¹

CONCLUSION

ILS is an uncommon, but previously underdiagnosed tumour of the inner ear. With recent advancements in MRI technology, detection sensitivity is now much increased, allowing identification of smaller ILSs on routine MRI. Radiologists should be aware of this tumour when screening patients with hearing or balance disorders.

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