Endolymphatic sac tumour: a rare cause of recurrent vertigo

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ABSTRACT

Endolymphatic sac tumour occurring in a 32-year-old man presenting with Meniere's like symptoms of recurrent vertigo, hearing loss and tinnitus is described. Magnetic resonance imaging and computed tomography showed a vascular bone tumour centred over the retrolabyrinthine aspect of the temporal bone where the endolymphatic sac was located. Surgical excision via a translabyrinthine approach was performed. Endolymphatic sac tumours are rare papillary adenocarcinomas that arise from the endolympatic sac. It can be mistaken both on radiology and histology for other tumours such as paragangliomas, renal or papillary thyroid carcinoma metastases. Surgical excision is the treatment of choice but sacrifice of the auditory and facial nerve may be needed in advanced cases to achieve tumour clearance.

Keywords: ear tumour, endolymphatic sac tumour, Hippel-Lindau disease, Meniere's syndrome, papillary adenocarcinoma

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INTRODUCTION

A patient with an endolymphatic sac tumour of the temporal bone presenting with typical symptomatology of Meniere's disease is described. This case highlights the importance of screening to rule out underlying causes for recurrent vertigo, and also reinforces the notion that the endolymphatic sac plays a significant role in the pathophysiology of Meniere's disease.

CASE REPORT

A 32-year-old Chinese man presented in August 2004 with a two-year history of recurrent vertigo. Each episode of dizziness lasted for about an hour and was associated with nausea and vomiting. This was relieved with medications each time. He also reported reduced hearing and a high-pitched tinnitus in the left ear. He was previously seen at another hospital two years ago, and was diagnosed as and managed for vestibular neuronitis.

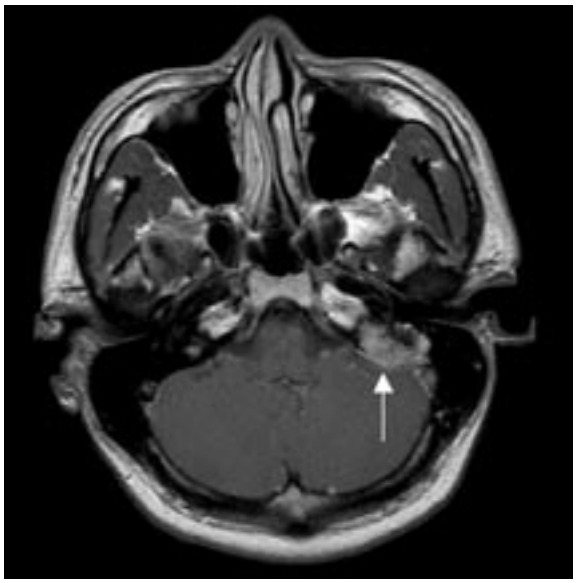

Fig. 1 Contrast-enhanced axial T1-W MR image shows heterogenous enhancement of the lesion located in the left retrolabyrinthine petrous temporal bone (arrow).

On otomicroscopical examination, both external ear canals were patent and tympanic membranes were intact and normal in appearance. There was no spontaneous or gaze-evoked nystagmus detected. Vestibular examination revealed a left vestibular hypofunction with impaired vestibulo-ocular reflexes. There were rapid head movements to the left, and a right beating post head-shake nystagmus. Positioning tests were negative for nystagmus. With the 512 Hz tuning fork, the Weber's test lateralised to the right. Air conduction was louder than bone conduction in both ears, suggesting a left-sided sensorineural hearing loss. Other cranial nerves examinations were intact. Stance and gait examination were normal. There were no cerebellar signs, and the rest of the otolaryngological examination was unremarkable.

Pure tone audiogram showed a moderate left sensorineural hearing loss, and the history of

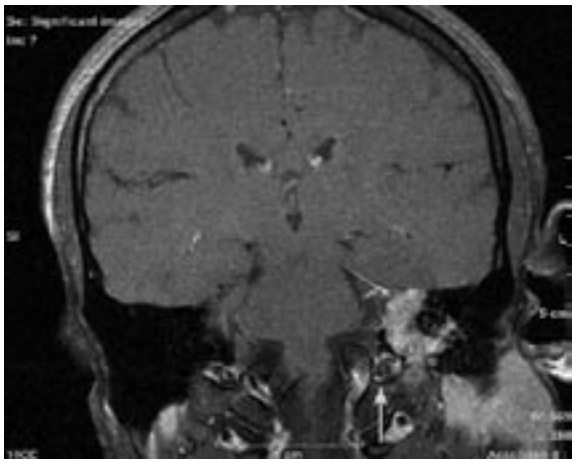

Fig. 2 Contrast-enhanced coronal T1-W MR image shows the relationship of the tumour with respect to the internal jugular vein (arrow).

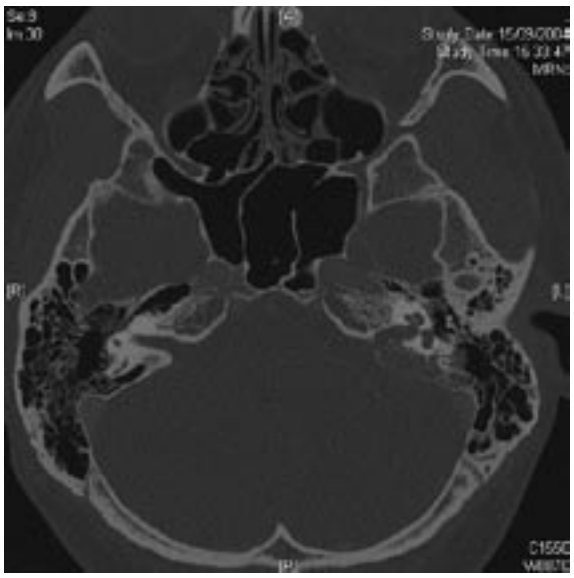

Fig. 3 Axial HRCT image shows a retrolabyrinthine soft tissue mass in the posteromedial aspect of the left temporal bone with bony erosion.

recurrent vertigo was clinically suggestive of Meniere's disease. Magnetic resonance (MR) imaging of the internal acoustic meatus showed a moderately well-defined mass (2.7 × 1.8 × 2.5 cm) arising from the left retrolabyrinthine petrous temporal bone. There was tumour extension into both the left posterior cranial fossa and the left internal acoustic canal. Hyperintense foci were also seen along the periphery. Heterogeneous enhancement of the lesion was seen with gadolinium and flow-void was noted within the lesion. Inferiorly, the lesion encroached the left jugular foramen and abutted the jugular bulb. There was no involvement of the middle ear (Figs. 1-2).

High-resolution computed tomography (HRCT) of the temporal bone was subsequently performed and confirmed the presence of the retrolabyrinthine soft tissue mass in the posteromedial aspect of the left temporal bone (Fig. 3). There was adjacent bony destruction, with scattered calcified foci in the soft tissue mass. The radiological findings suggested a vascular temporal bone tumour centred over the retrolabyrinthine aspect of the temporal bone where the endolymphatic sac is located. The tumour was locally invasive with extension seen into the internal acoustic canal, as well as medially into the posterior fossa. A provisional diagnosis of an endolymphatic sac tumour was made at this point.

The patient underwent surgical excision via a translabyrinthine approach by a combined neuro-otology and neurosurgical team. Preoperative angiogram with embolisation was performed one day prior to the surgery. The angiogram demonstrated a vascular tumour with feeding vessels from the left occipital and internal maxillary artery. Intra-operatively, the tumour was noted to be infiltrative and vascular. The disease was extensive, invading superiorly to the supralabyrinthine air cells, inferiorly to abut the jugular bulb, and medially breaching the dura and compressing onto the cerebellum. The tumour also extended into the internal auditory canal and was found to be adherent onto the facial nerve within the internal acoustic canal. The tumour was excised completely except for the part that was adherent on the facial nerve as the patient had insisted preoperatively on such a course of action to minimise facial nerve injury.

Final histology of the tumour was consistent with the diagnosis of endolymphatic sac tumour. Epithelial cells were arranged in papillary, follicular and nested patterns. The papillae were short and blunt in appearance. The nuclei were round to oval and in many cells the cytoplasm was voluminous, vacuolated and clear. There was no evidence of necrosis and mitotic figures were rare. No sheets of high-grade pleomorphic cells were identified. The tumour cells stained positive for CK AE 1/3 and CAM5.2 and were negative for synaptophysin. The postoperative recovery was uneventful. There was no facial nerve palsy (House-Brackmann grade 1), and he was discharged after one week. Postoperative MR imaging done six months later did not reveal any tumour recurrence. He was scheduled for a repeat scan after another year.

DISCUSSION

Endolymphatic sac tumours (ELSTs) are uncommon neoplasms of the temporal bone. Previously classified as aggressive papillary middle ear tumours, it has recently been recognised as a distinct

clinicopathological entity. It was first described by Heffner[1] who identified a series of middle ear lesions and proposed the endolymphatic sac as the site of origin. To our knowledge, this is the first case to be reported in Singapore.

ELSTs appear to be more common in women[2,3]. Most patients present in their forties and fifties[2-5]. Most cases are sporadic, though approximately 15% of patients have von Hippel-Lindau (VHL) disease[2,6]. Our patient was counselled preoperatively to undergo genetic and clinical tests to rule out VHL, but he refused. Von Hippel-Lindau disease is an autosomal dominant disorder characterised by tumours of the central nervous system (cerebellar haemangioblastoma, choroids plexus papilloma, and retinal angioma), renal cell carcinoma, renal and pancreatic cysts, pheochromocytoma, and papillary cystadenomas of the epididymis[4]. 11% of patients with VHL disease develop ELSTs, and these may occur bilaterally[5,7]. Hearing screening and serial imaging to exclude ELSTs are essential in these patients as early diagnosis can result in successful hearing preservation[8].

Symptoms are often non-specific and patients often present with audiovestibular symptoms such as sensorineural hearing loss, tinnitus or vertigo. Some patients may also present with Meniere's like symptoms, which was the initial working diagnosis for our patient[7]. A possible mechanism for the similarity in presentation can be attributed to endolymphatic duct obstruction with secondary hydrops as a result of unchecked ELST growth[7]. This highlights the importance of screening all patients with unilateral auditory symptoms for retrocochlear pathology[3]. Facial nerve palsy is often a late feature[3,4]. Trigeminal, lower cranial nerves (IX, X) palsies or cerebellar involvement may be present if the tumour has extended to the cerebellopontine angle or jugular foramen.

ELSTs are slow-growing tumours that are hypervascular and locally invasive with bony destruction. Metastases have not been reported[4,5]. Local recurrence occurs in cases of incomplete excision though it may take years to develop[2]. Macroscopically, they appear reddish or bluish and hypervascular. They are generally soft, but may contain parts of bone[9]. Microscopically, two main growth patterns exist and the tumour may show variable distribution of each pattern. The first, a papillary form, consist of papillary processes embedded in sheets of dense fibrous tissue. These papillary processes are lined by a single layer of epithelial cells, which are low columnar to cuboidal.

The second form shows a glandular arrangement with more flattened cells enclosing round spaces containing proteinaceous material, which is similar to thyroid follicles. In both patterns, the epithelial cells are often clear and occasionally eosionophilic. The cells also have small centrally-placed, ovoid nuclei. Pleomorphism is minimal, mitotic activity and necroses are also very rare[5,9]. The endolymphatic sac is derived from neuroectoderm and the detection of immunohistochemical markers such as glial fibrillary acid protein and vimentin, which are neuroectodermal epitopes, further provides evidence that the endolymphatic sac epithelium is the site of origin[10].

Possible histological differentials include metastatic renal cell or follicular thyroid carcinoma[4] and middle ear adenomas (MEA). Clear cell carcinoma of renal origin is also highly vascular but shows nuclear atypia and is more commonly tubular than papillary. Thyroid-like areas in ELSTs can also suggest metastatic follicular thyroid carcinoma. In the latter, thyroglobulin stains will be positive and clear cell papillary areas are not seen[4,5,9]. ELST and MEA share similar histological features, but MEA are restricted to the middle ear and normally do not erode the bone, which allows them to be distinguished radiologically.

Any patient with unilateral audiovestibular symptoms should be advised to have imaging studies to rule out the possibility of a retrocochlear lesion, such as a cerebellopontine angle tumour. This is even more so when there is progressive hearing loss, as was suggested in our patient's history. The imaging recommendations for endolymphatic sac tumour include both HRCT of the temporal bone and contrast-enhanced MR imaging[5,11]. The HRCT of the temporal bone complements MR imaging evaluation by better delineating bony erosion along the posterior surface of the temporal bone. MR angiography and MR venography may also help in defining its vascular relationship.

HRCT of the temporal bone reveals a destructive, retrolabyrinthine mass centred in the pre-sigmoid, posterior surface of the petrous pyramid. The posterior wall of the temporal bone is eroded, and in larger lesions (more than 3 cm), such as in this case, can also involve the jugular foramen wall and the posterior wall of the internal auditory canal. It may spread to the middle ear through the inner ear. Central spiculated calcifications within the tumour matrix are almost always present and are important diagnostic clues[11-13]. The tumour enhances heterogeneously with contrast.

MR imaging reveals hyperintense foci (due to

blood products or colloids) within the tumour on T1-weighted sequences[11,12]. 80% of ELSTs have these foci of increased signal intensity[11,13]. This high signal appear along tumour margins when it is less than 3 cm in size but is found within the tumour matrix when it is more than 3 cm[13]. Flow voids (focal low signal areas) may be present when the tumour is more than 2 cm in size. On T2-weighted images, the tumour shows heterogeneity, with the high signal secondary to protein and old haemorrhage, while the low signal represents bone fragments or calcification. MR images also shows heterogeneous enhancement due to solid components that enhance strongly after gadolinium administration[14].

Angiography shows a vascular tumour with feeding vessels most commonly arising from the ascending pharyngeal artery and occipital artery. In larger tumours (more than 3 cm), contributions may also arise from the internal carotid artery or the posterior circulation[13]. ELSTs may be confused radiologically with glomus jugulare paraganglioma, which originate from the jugular foramen and spreads upwards and laterally into the middle ear[2]. For paragangliomas, HRCT shows an isodense soft tissue mass and intense enhancement with contrast, mass effect and permeative bone changes. On MR imaging, paragangliomas will have an intermediate signal on unenhanced T1-weighted images, with moderate to intense enhancement after gadolinium administration. A characteristic salt and pepper pattern is also seen on T2-weighted images. Angiography shows dense tumour staining. Other important radiological differential diagnoses include cholesterol granuloma of the petrous apex, schwannoma of jugular foramen and meningioma of jugular foramen-cerebellopontine angle cistern. It is possible to differentiate these lesions from ELSTs radiologically, based on their locations and their characteristic imaging properties.

Early diagnosis and management is important as it improves surgical outcomes. Complete surgical resection with wide resection is the treatment of choice and results in cure[3,4]. Prognosis is good if complete surgical resection is achieved. Incomplete resection is associated with a high rate of local recurrence though this may be late as the tumour is slow-growing[9]. Larger lesions will benefit from preoperative angiography with embolisation.

Facial nerve sacrifice and grafting may be required in cases with preoperative facial nerve palsy or direct involvement at surgery[4]. Hearing preservation is possible with small tumours, hence the need for early diagnosis[8], but this should take second place to tumour clearance in the management of these tumours. Radiotherapy is not routinely given, as ELSTs appear to be resistant to radiation[3,5]. It should only be considered in patients unfit for surgical resection or for those lesions deemed inoperable.

In summary, ELSTs are rare, papillary tumours of the temporal bone that arise from the endolymphatic sac. Early diagnosis and treatment are important as it leads to an improved outcome. Surgical resection is the primary modality of treatment and offers excellent tumour control.

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