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Radiology in Focus

Endolymphatic sac tumours

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Abstract
We present a case of a papillary tumour of the petrous bone. The established terminology for this rare neoplasm is endolymphatic sac tumour (ELST) but the true origin remains controversial. ELSTs are associated with von Hippel-Lindau disease. They are locally invasive, highly vascular and often require endovascular embolization prior to surgery. Both radiologically and histologically ELSTs are easily mistaken for other more common tumours such as paragangliomas and renal or papillary thyroid carcinoma metastases. This is important because local excision is curative.

Key words: Endolymphatic Sac; Petrous Bone; Diagnosis, Differential; Hippel-Lindau Disease; Therapy

Introduction

There are two distinct types of adenomatous tumour of the petrous bone, a papillary form and a non-papillary form known as middle-ear adenoma (MEA). Unlike MEAs, papillary tumours are hypervascular, locally invasive and tend to recur. However, they are slow growing, show little evidence of mitosis, nuclear anaplasia or pleomorphism, and do not metastasize. Consequently their malignant potential is ambiguous and they have been classified histologically by different authors as adenomas, low-grade adenocarcinomas, aggressive papillary tumours and invasive papillary cystadenomas.

The derivation of petrous bone papillary tumours is also controversial. Possible sites of origin include the epithelia of the middle ear and mastoid air cells, the endolymphatic sac and ectopic choroid plexus. Tumours were thought to arise from the middle ear until 1984 when Hassard described an extradural papillary lesion that was adherent to the endolymphatic sac.[1] In 1989 Heffner reviewed the light and electron microscopic and immunohistochemical features of 20 papillary-cystic tumours of the petrous bone, concluding they were low-grade adenocarcinomas likely to be of endolymphatic sac origin.[2]

The endolymphatic sac is derived from neuroectoderm so the discovery that these tumours express neuroectodermal epitopes such as S-100, glial fibrillary acidic protein and synaptophysin gave support to the theory. The term endolymphatic sac tumour (ELST) was first proposed by Li *et al.* in 1993[3] and around 60 cases have been reported since using this nomenclature,[4–6] although it has yet to receive universal acceptance because of the possibility that papillary tumours could also arise from the middle ear or mastoid.[7] Of greater clinical relevance is the need to differentiate ELSTs from other similar and more common tumours such as paragangliomas and renal and papillary thyroid carcinoma metastases.

Case report

A 68-year-old Maltese woman presented with four years of right sensorineural deafness and several months of progressive headache and right facial weakness. Apart from a dense lower motor neurone facial nerve palsy there were no neurological or other abnormalities on examination.

Computed tomography (CT) demonstrated a large heterogenously enhancing mass centred on the posterolateral aspect of the right petrous bone (Figures 1 and 2), eroding the jugular fossa inferiorly and extending laterally through the mastoid air cells to involve the external auditory canal and subcutaneous tissues of the pinna. Anteriorly it eroded the posterior semi-circular canal, internal auditory meatus and posterior part of the tympanic cavity. On magnetic resonance (MR) imaging it

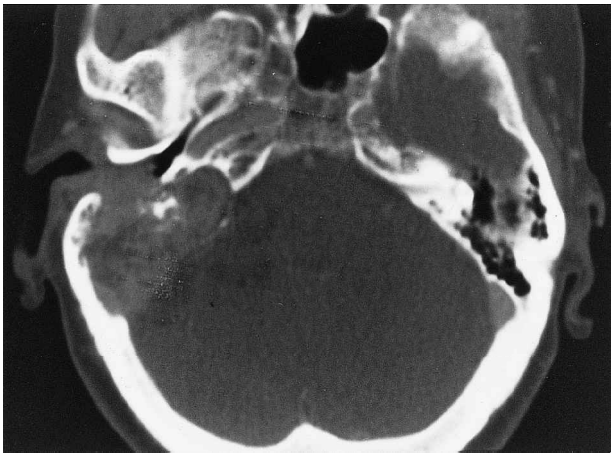

Fig. 1
Contrast enhanced axial CT petrous bones (bone windows).

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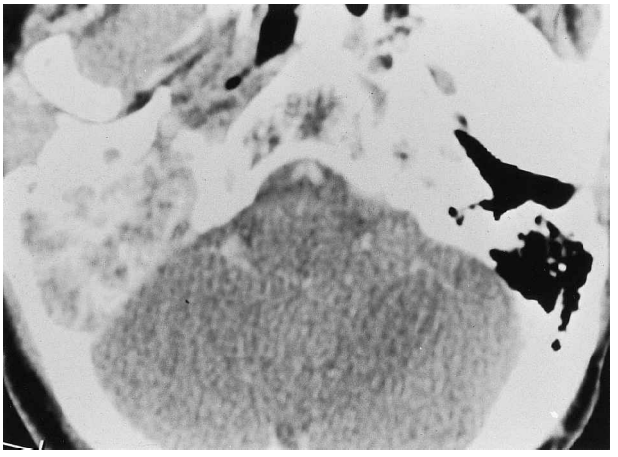

FIG. 2
Contrast enhanced axial CT petrous bones (soft tissue windows).

was heterogenous on both T1- and T2-weighted sequences with evidence flow voids and areas of high and low T1- and T2-weighted signal suggestive of blood products (Figures 3 and 4). The mass indented the cerebellum but the dura was intact. Cerebral angiography confirmed the hypervascular nature of the tumour, which was supplied predominantly by branches of the ascending pharyngeal and occipital arteries (Figure 5).

A radiological diagnosis of jugular paraganglioma was made. Pre-operative embolization of several feeding vessels arising from the ascending pharyngeal and occipital arteries was performed using 150–250 micrometer PVA particles (Figure 6) before transferring the patient directly to the operating theatre. The tumour was resected using a combined right occipital craniectomy and translabyrinthine approach. The patient remains well at two year follow-up.

Histopathological examination showed a complex papillary architecture with areas of haemorrhage and haemosiderin and scattered inflammatory cells. Epithelial cells were low columnar to cuboidal with eosinophilic cytoplasm and small ovoid nuclei. No nuclear pleomorphism or mitotic figures were seen.

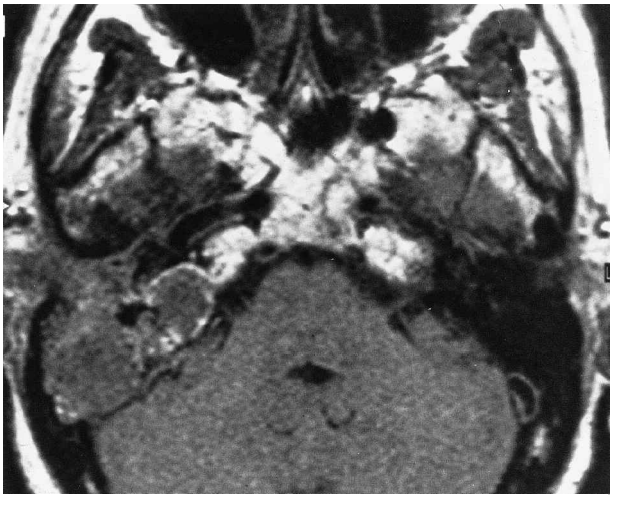

FIG. 3
Axial T1W TSE petrous bones (unenhanced).

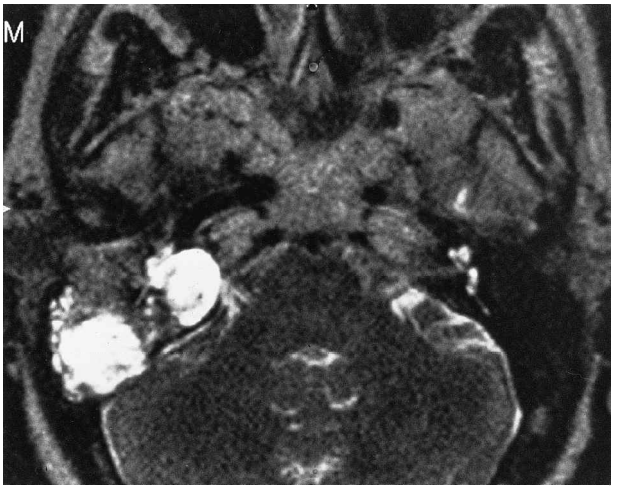

FIG. 4
T2W axial FSE petrous bones.

Discussion

Anatomy

The endolymphatic sac plays a role in the autoregulation of endolymphatic pressure and ion balance. The endolymphatic duct communicates between the saccular and utricular ducts and the endolymphatic sac. The endolymphatic sac consists of a proximal partially intraosseous pars rugosa and an extraosseous saccular portion which extends beyond the vestibular aqueduct sheathed between two layers of dura.

Epidemiology

ELSTs are so rare it is difficult to draw accurate conclusions about their epidemiology from the literature. They appear to be more common in women. The mean presenting age is approximately 45 years although there have been cases in

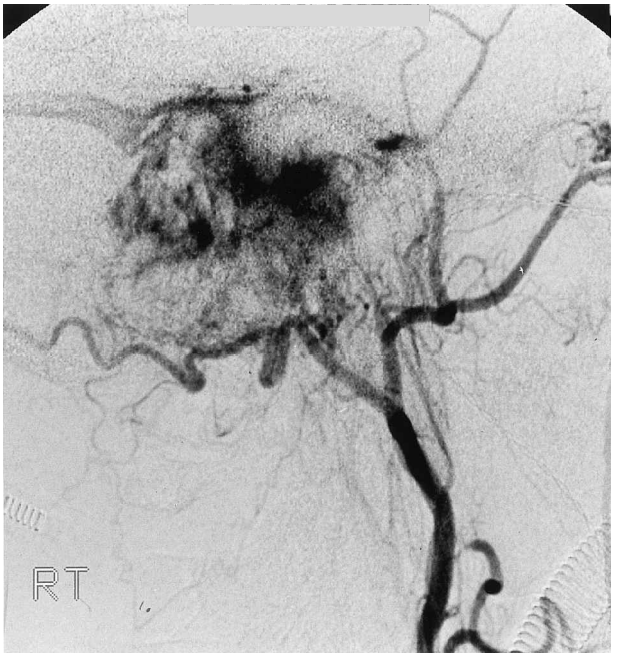

FIG. 5
Right external carotid artery angiogram, pre-embolization.

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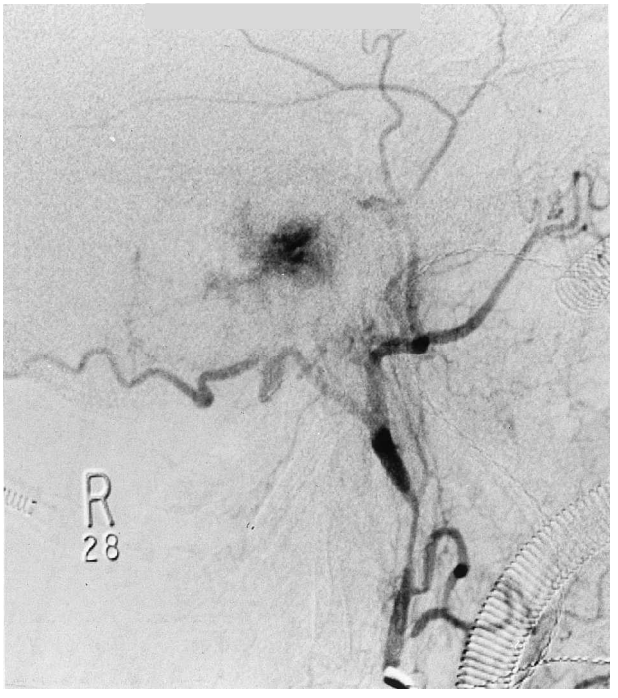

FIG. 6
Right external carotid artery angiogram, post-embolization.

both teenagers and septuagenarians.[4] Approximately 15 per cent of patients have von Hippel-Lindau (VHL) disease and most of the few reported bilateral cases are associated with the condition. The incidence of ELST in VHL has yet to be established but many authors now advocate audiometric screening in these patients.[8–10]

- **A patient with a right sensorineural deafness, facial weakness and headache was found to have a papillary tumour of the temporal bone that was embolized and excised**
- **The origin of such lesions is not known but is thought to be the endolymphatic sac. Such tumours are associated with von Hippel-Lindau disease and may be confused, radiologically and histologically, with paragangliomas and metastatic deposits**
- **The differential diagnosis is important as excision of papillary tumours is curative**

Clinical features

ELSTs are slow growing locally invasive tumours that follow two patterns of progression. Most are believed to arise from the pars rugosa and preferentially extend into the petrous bone. These patients typically present with long-standing sensorineural deafness. Facial nerve palsy is a late feature. Tumours arising from the extraosseous portion preferentially extend into the jugular foramen and cerebellopontine angle causing lower cranial nerve palsies but cerebellar signs are rare. Local recurrence occurs but may take years to develop. ELSTs have never been shown to metastasize.

Radiology

CT typically shows a retrolabyrinthine mass. Small tumours may cause no bony abnormality other than widening of the vestibular aqueduct but as they expand there is progressive 'moth-eaten' bone erosion into the supra- and infralabyrinthinem and mastoidotympanic regions, often associated with reactive new bone. The cochlea and middle-ear cavity are usually spared. ELSTs enhance intensely, may be necrotic or cystic and usually have central spiculated calcification. On MR imaging ELSTs are heterogenous in signal with heterogenous enhancement. Multiple high signal intensity foci on both T1- and T2-weighted images indicate the presence of methaemaglobin, blood-filled or proteinaceous cysts and cholesterol clefts. Scattered low signal foci represent haemosiderin due to repeated parenchymal bleeding. Such blood products are unusual in other temporal bone lesions. ELSTs are hypervascular, usually supplied by dural branches of the ascending pharyngeal and stylomastoid arteries although larger tumours may acquire a supply from the intrapetrous internal carotid artery and posterior circulation.[4–6]

Histopathology

Macroscopically ELSTs are highly vascular, friable and polypoid. Microscopically they are papillary adenocystic tumours with fibrous stroma and often with thyroid follicle-like glandular structures. The epithelial cells are cuboidal and low columnar with eosinophillic or vacuolated cytoplasm and a subepithelial vascularity typical of a papillary cystadenomatous tumour. Siderophages, cholesterol clefts and inflammatory cells are frequently found. Despite little mitotic activity and only slight cellular polymorphism, ELSTs are classified as low-grade adenocarcinomas because of their clinical course. They stain positively for periodic acid-Schiff-reactive material and keratin and may express neurone-related epitopes such as synaptophysin, S-100, glial fibrillary acidic protein and leu-7, reflecting the neuroectodermal origin of the endolymphatic sac.[11]

Differential diagnosis

ELSTs are most commonly misdiagnosed radiologically for jugulotympanic paragangliomas although the differential includes vascular metastasis, aggressive meningiomas, chondrosarcomas, haemangiopericytomas and plasmacytomas.[4–6] The radiological similarity of ELSTs to these more common tumours is paralleled by a histological resemblance to paragangliomas, thyroid and renal cell carcinomas, papillary meningiomas and choroid plexus papillomas that have similar immunohistochemical patterns. Differentiating ELSTs from middle-ear tumours is relatively easy because the mastoid air-cells usually remain pneumatized. Middle-ear adenomas are hypovascular and do not erode bone. Adenocarcinomas cause otalgia and otorrhoea that are unusual features in ELSTs.

Management

Early diagnosis is essential, as cure is virtually impossible in advanced tumours. The treatment of choice is complete excision with wide margins and pre-operative angiography and embolization is advisable.[6] A retrolabyrinthine approach may be considered if hearing is preserved, but if there is tumour extension into the labyrinth a translabyrinthine or transcochlear approach may be necessary. Heffner *et al.* reported a 90 per cent cure rate with total excision but the recurrence rate following partial resection appears to be high.[2] No long-term follow-up studies are available yet and published series are too small to determine the controversial role of post-operative radiotherapy.

Conclusion

We describe a case of a papillary tumour of the petrous bone. The established terminology for this rare neoplasm is ELST but the true origin remains controversial. ELSTs are associated with VHL. They are locally invasive, highly vascular and often require endovascular embolization prior to surgery. Both radiologically and histologically they are easily mistaken for more common tumours such as paragangliomas and renal or papillary thyroid carcinoma metastases. This is important because complete excision is curative.

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