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Aggressive papillary middle ear tumour

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Abstract

A rare middle ear tumour is reported. The clinical presentation was similar to that of a glomus tumour but the pathology that of an aggressive papillary middle ear tumour. This is a recently recognized subgroup of middle ear glandular tumours. Clinical findings, imaging, pathology and treatment are presented.

Introduction

Glandular neoplasms of the middle ear and mastoid are uncommon and the terminology of primary middle ear cleft tumours is confused. Similar entities have been reported under different names. However, as more cases are described some well defined subgroups are emerging, such as middle ear adenoma (Derlacki and Barney, 1976; Hyams and Michaels, 1976), carcinoid and adenocarcinoma of both high or low grade.

Aggressive papillary middle ear tumour (Gaffey, 1988) is a rare subgroup which has low grade histological features but locally aggressive behaviour. Difficulties in clinical and histological diagnosis may be encountered. We report a case of aggressive papillary middle ear tumour that was diagnozed preoperatively as a glomus jugular tumour.

Case report

A 31-year-old female presented with a 12 month history of progressive left facial weakness and pulsatile tinnitus, with hearing loss present for 12 years.

There was a reddish-blue mass filling the lower half of the middle ear. The mass blanched with pressure and there was a moderate conductive hearing loss. The right ear was normal. There was a House grade IV left facial weakness (House, 1983) and a left palatal weakness. A pulsatile bruit was clearly heard over the left mastoid.

A CT scan (Fig. 1) showed a destructive lesion involving the left jugular foramen, mastoid air cells, internal auditory canal, and the vertical portion of the facial nerve. Digital Subtraction Angiography (DSA) (Fig. 2) showed a vascular tumour mass lying in the posterior fossa and petrous temporal bone. Feeding vessels arose from the occipital and maxillary arteries.

Operative findings were of an extensive, bright red, unencapsulated vascular tumour filling the lower half of the middle ear and invading bone around the vertical segment of the facial nerve. Medially it occupied the greater part of the petrous apex. It had invaded the sigmoid sinus as far as the junction with the tranverse sinus and filled the jugular bulb. The dura over the posterior face of the temporal bone was thickened by tumour infiltration and there was a moderate extension into the cerebellopontine angle. The facial nerve was free of tumour in the mid-portion of the horizontal segment and immediately below the stylo-mastoid foramen. Between these points it was completely invaded by tumour.

Sub-total removal of the temporal bone was performed, the facial nerve being sectioned at the mid-portion of the horizontal segment. Although the occipital artery was ligated there was copious bleeding as the temporal bone was infiltrated by vascular tumour in every nook and cranny. The divided facial nerve was repaired with a sural interposition graft. Tumour removal was clinically complete.

Post-operative recovery was complicated by a CSF leak due to resection of the posterior fossa dura. This settled after a ventriculo-peritoneal shunt procedure. She is very well, nine months later and facial function has improved significantly to House grade III.

Pathology

Routinely stained sections showed a papillary epithelial

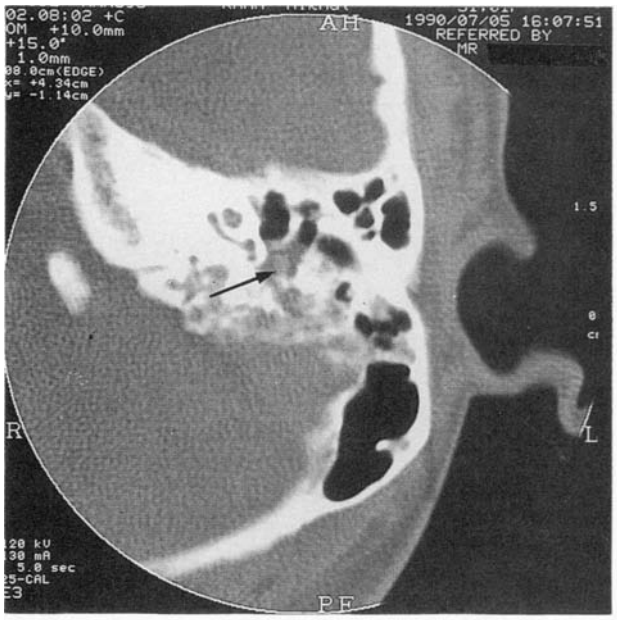

Fig. 1

CT Scan. Standard axial magnified view (bone window settings) showing extensive erosion of the left mastoid air cells and posterior surface of the temporal bone. All bone around the normal position of the vertical facial nerve has been eroded (arrow).

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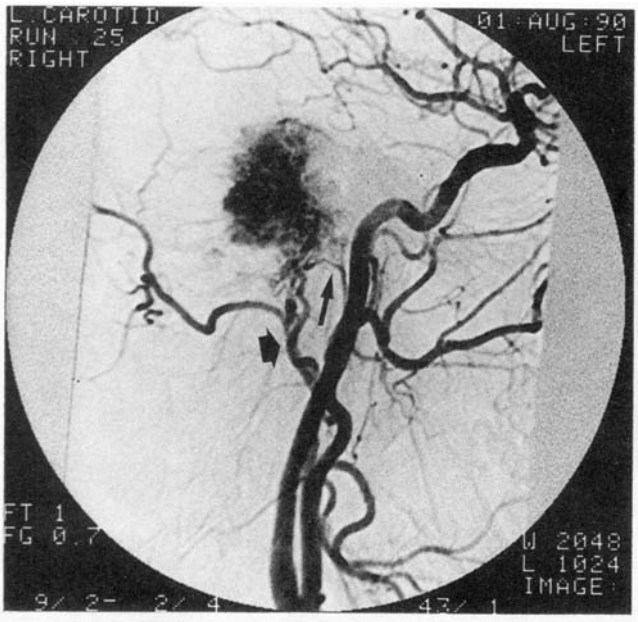

FIG.2

Digital Subtraction Angiogram showing very vascular left temporal bone tumour with large feeding vessels derived from the occipital artery (large arrowhead) and smaller vessels from the origin of the maxillary artery (small arrow).

tumour that invaded bone. The papillae were covered by a single layer of columnar cells with apical nuclei showing no atypia and no mitoses. The cores of the papillae contained numerous capillary blood vessels. The cells had eosinophilic, vacuolated or clear cytoplasm, containing glycogen but no mucin. There was some extracellular material resembling thyroid colloid (Fig. 3). These features led to a diagnosis of aggressive papillary middle ear tumour (APMET).

Immunoperoxidase stains (Hsu and Fangen, 1981) were positive with antibodies against epithelial membrane antigen, cytokeratin, S-100 protein, neuron specific enolase and vimentin. Glial fibrillary acidic protein was focally positive. Synaptophysin, chromogranin, lysozyme and thyroglobulin were negative.

Electron microscopic study showed short microvilli and subluminal junctional complexes. Adjacent cells were linked by desmosomes and the tonofilament bundles inserting into them had an unusual round shape and an electron dense amorphous appearance. Throughout the cytoplasm were numerous large round, oval and sausage shaped tonofilament bundles. There was also occasional lysosomes and lipid droplets (Fig. 4).

Discussion

Vascular middle ear tumours, characteristically glomus tumour, present with a long history of pulsatile tinnitus and hearing loss with facial paralysis being a relatively late event (Cheesman, 1987). An aggressive papillary middle ear tumour is very rare. The clinical picture and imaging studies led to a provisional diagnosis of glomus tumour, which was questioned at the time of surgery because

(1) the tumour was soft and friable;
(2) its colour was brigher red than that seen in glomus tumours;
(3) it eroded the posterior fossa dural plate near the endolymphatic sac rather than the jugular bulb.

The histopathological diagnosis was aggressive papillary

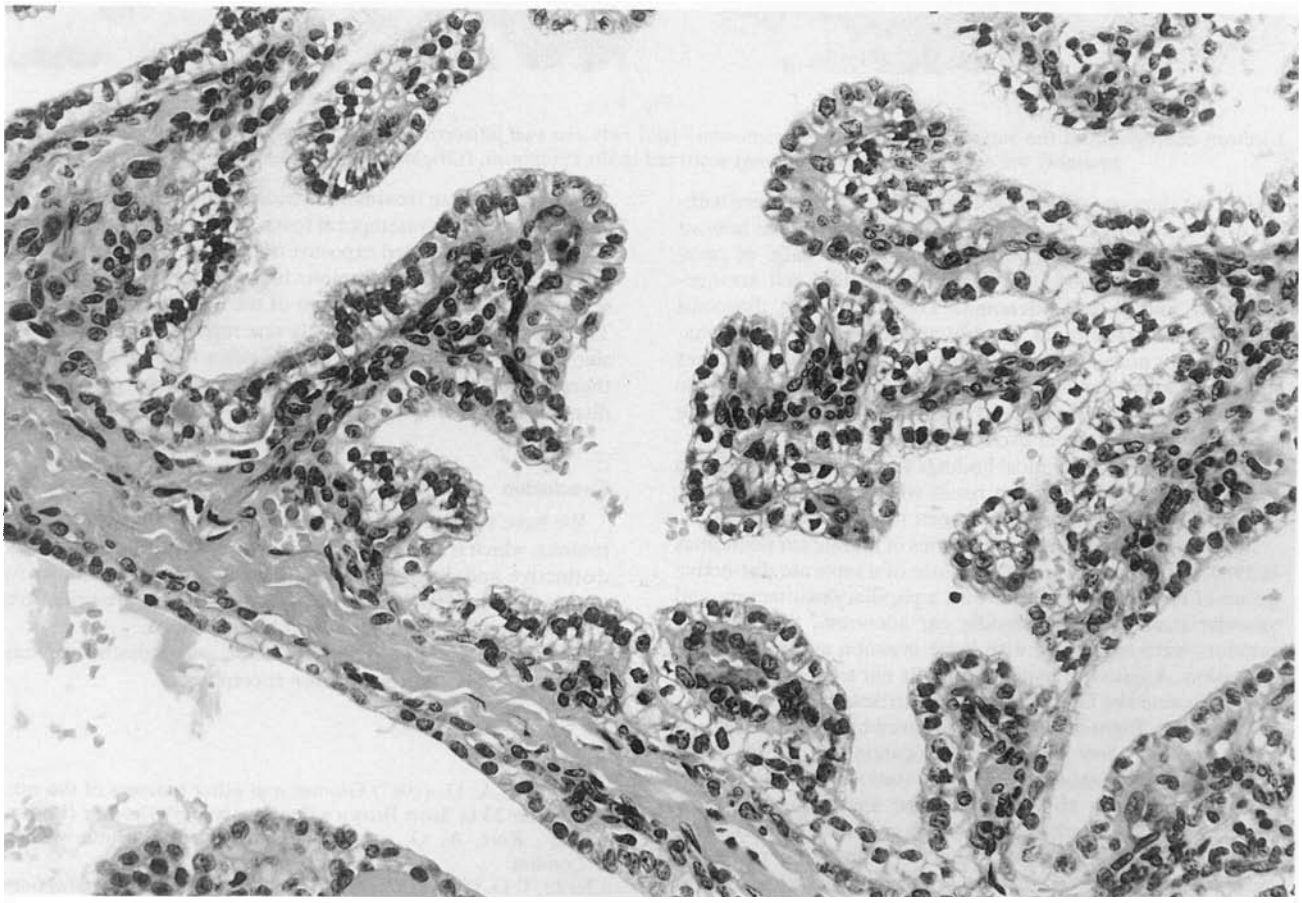

FIG. 3

Photomicrograph of tumour showing papillary architecture and clear cytoplasm. (Haematoxylin and eosin, original magnification ×20)

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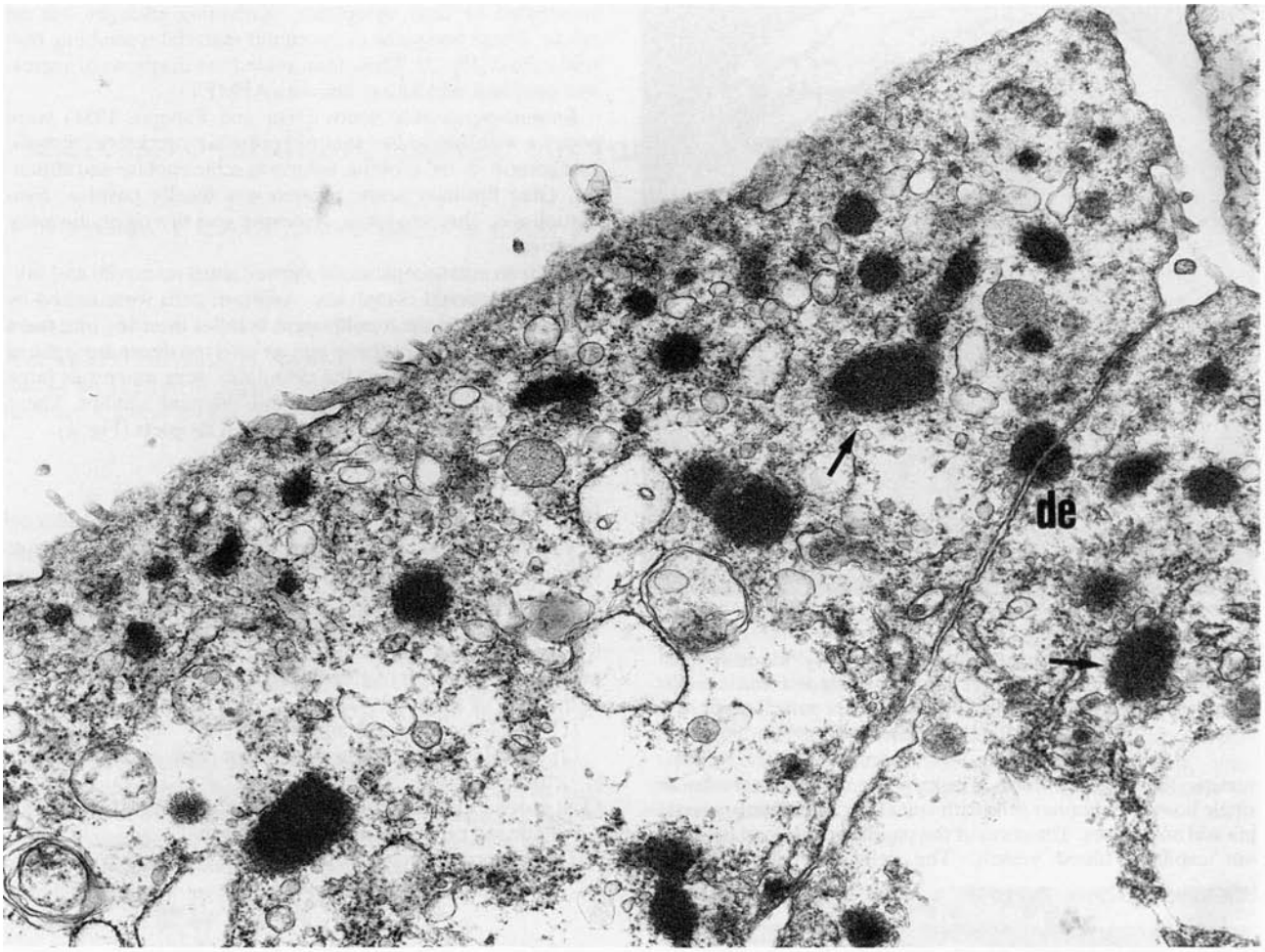

FIG. 4

Electron micrograph of the surface of a papilla. A desmosome (de) between two adjacent cells has dense tonofilament bundles which resemble the other dense bodies (arrows) scattered in the cytoplasm. (Original magnification ×14000)

middle ear tumour (APMET). The papillary architecture without cytological atypia was typical of descriptions of this tumour (Gaffey *et al.*, 1988) and quite unlike the 'cell balls' of paraganglioma or the solid and glandular cuboidal cell arrangements of middle ear adenoma. The differential diagnosis included metastatic renal, thyroid and lung adenocarcinoma, but there was no evidence of primary tumour elsewhere. Other similar tumours include primary temporal bone high grade adenocarcinoma and malignant papillary meningioma, but the lack of cytological atypia excluded these.

Our immunohistochemical findings agree with Gaffey and to these we add a focal positive result with glial fibrillary acidic protein. We believe this has not been reported previously.

Mills and Fechner, reporting a series of middle ear adenomas in 1984, first pointed out the existence of a separate distinctive group of middle ear tumours with a papillary achitecture and vascular stroma. Unlike middle ear adenoma, the papillary tumours were aggressive with bone invasion and intracranial extension. Aggressive papillary middle ear tumour (APMET) is a term coined by Gaffey *et al.* (1988) to describe these distinctive tumours. Eight of nine cases reviewed by Gaffey were originally called low grade adenocarcinoma. Six were pre-operatively diagnozed as glomus tumour as was our case. APMET lies in a clinicopathological spectrum of disease between middle ear adenoma and high grade adenocarcinoma.

The origin of APMET is generally considered to be the modified respiratory epithelium of the middle ear cleft. Most cases have been centred in the middle ear, but some appear to commence within the mastoid air cells and petrous temporal bone (Clarke, 1989; Gaffey, 1989).

The appropriate treatment is wide surgical resection, in this case the type A infratemporal fossa approach of Fisch (1982). This operation included exposure of the upper cranial nerves in the neck, control of the tumour blood supply, subtotal petrosectomy and complete resection of the intracranial extension. This type of approach is the only one reported to cure aggressive papillary middle ear tumours (Gaffey *et al.*, 1988). Radiotherapy has been used as adjunctive therapy without discernable effect.

Conclusion

We have reported a case of aggressive papillary middle ear tumour, which is locally invasive, slowly growing, histologically distinctive and does not metastasize. It is clinically similar to paraganglioma requiring the same careful pre-operative assessment of tumour extent and blood supply.

Treatment is wide surgical resection and radiotherapy has little role to play when resection is complete.

References

Cheesman, A. D. (1987) Glomus and other tumours of the ear. Chapter 23 in Scott Brown's Otolaryngology. The Ear (Booth, J. B., Kerr, A. G., eds.) 5th edition. Vol. 3. Butterworths: London.

Clarke, T. D. (1989) Letter to the editor. *American Journal of Surgical Pathology*, **13(11):** 985–986.

Derlacki, E. L., Barney, P. L. (1976) Adenomatous tumours of the middle ear and mastoid. *Laryngoscope*, **86:** 1123–1135.

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Fisch, U. (1982) Infratemporal fossa approach for glomus tumours of the temporal bone. *Annals of Otology, Rhinology and Laryngology,* **91:** 474–479.
Gaffey, M. J., Mills, S. E., Fechner, R. E., Intermann, S. R., Wick, M. R. (1988) Aggressive papillary middle-ear tumour. A clinicopathological entity distinct from middle-ear adenoma. *American Journal of Surgical Pathology,* **12(10):** 790–797.
Gaffey, M. J. (1989) Reply to Clarke . . . Letters to the editor. *American Journal of Surgical Pathology,* **13(11):** 986–987.
House, J. W. (1983) Facial nerve grading systems. *Laryngoscope,* **93:** 1056–1069.
Hsu, S. M., Fanger, H. Use of avidin-biotin peroxidase complex (ABC) in immunoperoxidase techniques. *Journal of Histochemistry and Cytochemistry,* **29:** 577–580.
Hyams, V. J., Michaels, L. (1976) Benign adenomatous neoplasms (adenoma) of the middle ear. *Clinical Otolaryngology,* **1:** 17–26.
Mills, S. E., Fechner, E. (1984) Middle ear adenoma. *American Journal of Surgical Pathology,* **8:** 677–685.

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