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Pathology in Focus

Epithelial-myoepithelial carcinoma of the parotid gland. An unusual cause of ear canal stenosis

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Abstract
Epithelial-myoepithelial carcinoma (EMC) accounts for approximately one per cent of salivary gland tumours. This tumour is gaining wider recognition following inclusion into the WHO histological classification of salivary gland tumours in 1990. Salivary gland tumours characteristically present with an enlarging mass. We describe an unusual presentation of a salivary gland tumour with stenosis of the external ear canal in the absence of a palpable mass. EMC usually arises from the salivary glands but isolated cases have been described arising primarily from the paranasal sinuses, trachea and lacrimal gland. The management of this tumour is still evolving with surgical excision being the main-stay of treatment. The efficacy of radiotherapy has not yet been established but high local recurrence rates despite apparently adequate excision and the possibility of a multicentric origin of the tumour may herald an increasing role for radiotherapy in the future. We stress the importance of awareness of adjacent structures when considering the cause of cartilaginous canal stenosis.

Key words: Salivary gland neoplasms; Ear canal; Carcinoma

Case report

A 55-year-old lady presented in February 1994 with right-sided otalgia and deafness. She was otherwise asymptomatic and had no notable past otological history. Examination revealed stenosis of the right external ear canal occluding the tympanic membrane. The left ear looked normal. No masses were palpable in the head or neck. Pure tone audiometry showed moderate high frequency conductive hearing loss on the right side and normal hearing thresholds on the left. A high resolution CT scan of the temporal bones demonstrated a soft tissue mass in the external ear canal. The middle and inner ears appeared normal (Figure 1).

Examination of the ear under anaesthesia showed stenosis along the full length of the cartilaginous canal. The bony canal and tympanic membrane appeared normal. The patient was subsequently admitted for meatoplasty but on this occasion a soft tissue mass was found bulging into the anterior wall of the external ear canal. The mass was dissected out and appeared to be arising from the parotid region. Histological examination revealed this to be an EMC of salivary gland origin.

A repeat high resolution CT scan suggested that there was a mass related to the parotid gland but it was difficult to differentiate from parotid tissue. Exploration of the parotid gland was undertaken. A fibrous mass was found extending from the posterior part of the parotid gland to the mastoid origin of the sternocleidomastoid muscle. There did not appear to be any involvement of the external ear canal. A superficial parotidectomy was carried out with excision of sternocleidomastoid fibres underlying the gland and the periosteum over mastoid process in continuity. Surprisingly, histology of the excised tissue showed no evidence of residual tumour. It was assumed that excision had been complete during the initial biopsy as multiple intra-operative frozen sections of the external canal wall

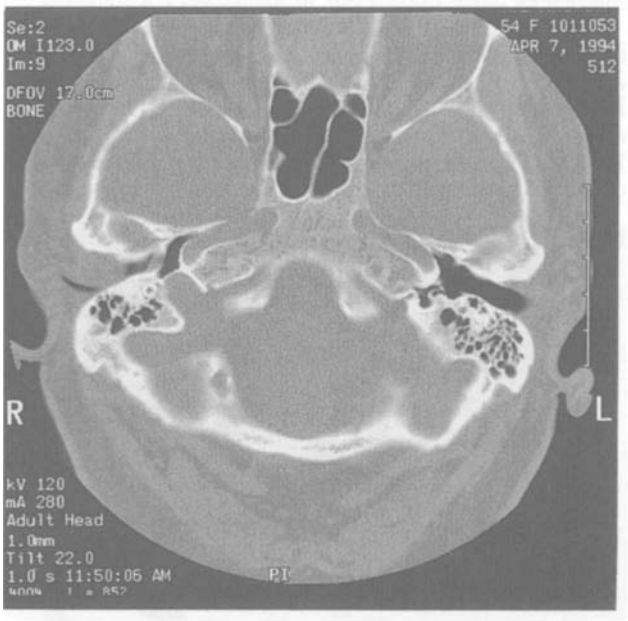

FIG. 1
CT scan of temporal bones showing localized soft tissue mass in the right external auditory canal.

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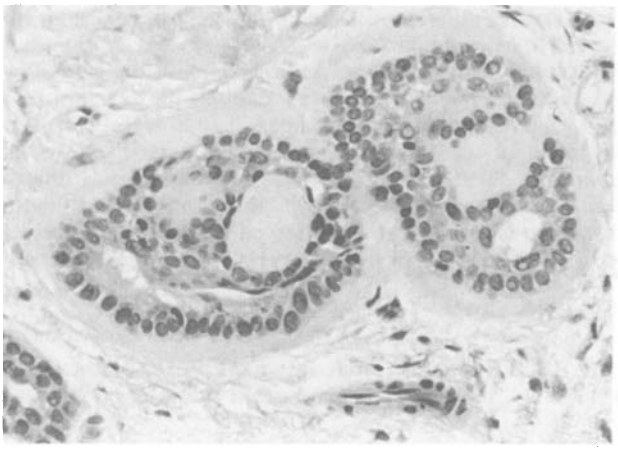

FIG. 2
Ducts with two cell layers (H & E; × 20).

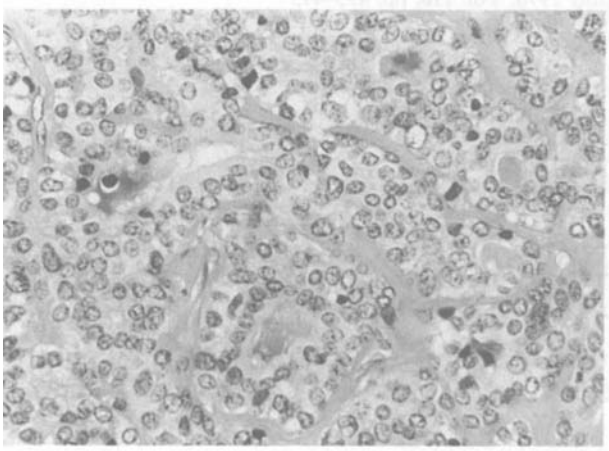

FIG. 3
Deposition of hyaline basement membrane material (H & E; × 20).

had shown no evidence of tumour at the resection margins. Facial nerve function was normal following the operation.

Pathology

The initial biopsy consisted of a mass of tissue measuring up to 1.8 cm in diameter. The tissue consisted of a tumour mass formed of duct-like structures with an inner layer of epithelial cells and an outer layer of clear cells (Figure 2). Focally there was deposition of hyaline basement membrane material (Figure 3). Immunohistochemically the inner cells expressed low molecular weight cytokeratin (Figure 4) while the outer cells stained positively with a pan-cytokeratin antibody and for S100 protein consistent with a myoepithelial phenotype (Figure 5). A diagnosis of epithelial-myoepithelial carcinoma of presumed salivary gland origin was made.

Discussion

EMC is a rare tumour accounting for less than one per cent of salivary gland neoplasms (Seifert *et al.*, 1990). Although not recognized in the WHO International Histological Classification of Tumours until 1990 (Seifert *et al.*, 1990), the tumour was first described in 1972 by Donath *et al.* and it is likely that similar tumours had been described previously by different names.

Patients are mostly female in their fifth to eighth decades (Fonseca and Soares, 1993) although ages ranging from eight to 103 years have been described (Simpson *et al.*, 1991; Morinaga *et al.*, 1992). The tumour usually occurs in the major salivary glands and in particular the parotid. It may occur in the minor salivary glands of the mouth and has also been described in the maxillary sinus (Luna *et al.*, 1987), trachea (Horinouchi *et al.*, 1993) and lacrimal gland (Ostrowski *et al.*, 1993). Presentation is usually with a progressively enlarging lump over a period of months to years. Features typifying malignancy such as pain and facial nerve palsy occur rarely. Stenosis of the external ear canal as the initial manifestation of the tumour has not previously been described.

Histologically the tumour is characterized by well-defined tubules with two types of cells. An inner lining of eosinophilic cuboidal epithelial cells is surrounded by an outer mantle of larger myoepithelial cells with clear cytoplasm. The clear cells characteristically contain glycogen but not mucin (Batsakis *et al.*, 1992). The cellular constituents are surrounded by a mucoid or mucohyaline stroma. Perineural invasion and necrosis may be seen but nuclear pleomorphism and mitoses are rare. The tumour may exhibit the classical biphasic pattern or there may be varying degrees of clear cell, epithelial cell or stromal predominance within each tumour. Immunohistochemistry is useful in distinguishing the two ductal layers. Myo-

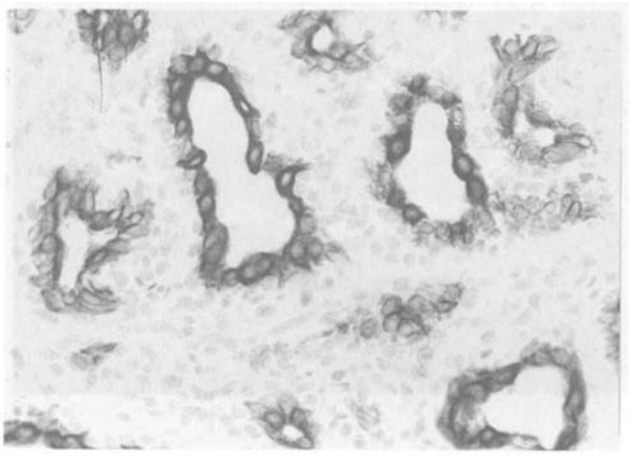

FIG. 4
Inner epithelial cells staining for low molecular weight cytokeratin (× 20).

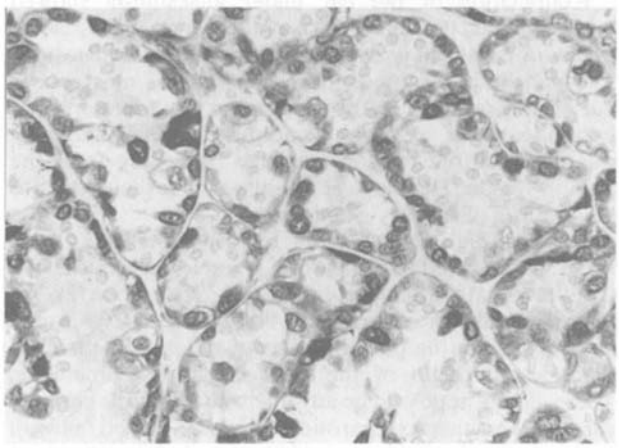

FIG. 5
Outer myoepithelial cells staining for S100 protein (× 20).

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epithelial cells are best identified using S100 protein. Epithelial cells are identified by cytokeratin reactivity.

The differential diagnosis of epithelial myoepithelial carcinoma and, in particular, the clear cell predominant type is that of clear cell tumours of salivary glands. Such tumours include acinic cell carcinoma, adenoid cystic carcinoma, sebaceous carcinoma and metastatic renal carcinoma. The biphasic pattern distinguishes EMC from these tumours. Additional features include characteristic secretory granules in acinic cell carcinoma and the presence of lipid droplets in the clear cells of sebaceous carcinoma. Abdominal imaging is sometimes necessary to exclude metastatic renal carcinoma. EMC and adenoid cystic carcinoma may represent part of a morphological spectrum of tumours arising from the intercalated ducts of salivary glands.

Reports evaluating different treatment modalities for EMC are scanty although surgical excision is considered the treatment of choice. Wide excision margins have been recommended as the tumour has a high rate of local recurrence which may relate to the locally infiltrative growth pattern. Recently the possibility of multiple tumour foci within the gland has been suggested (Di Palma, 1994) and may help to explain high recurrence rates despite apparently adequate surgical excision (Corio *et al.*, 1982; Luna *et al.*, 1985). Adjuvant radiotherapy may be of benefit in preventing local recurrence (Corio *et al.*, 1982; Simpson *et al.*, 1991) however its efficacy to date is largely based on anecdotal reports. The role of chemotherapy has not been evaluated.

In Luna's series of 35 cases local recurrences occurred in up to 37 per cent following surgical excision (Luna *et al.*, 1987). Lymph node metastases occurred in 17 per cent of cases and deaths attributable to the tumour in six per cent. Tumour size may be a predictor of local recurrence. Corio *et al.* (1982) and Hamper *et al.* (1989) found higher recurrence rates in tumours larger than 3 and 4 cm respectively. Until recently no relationship between histological features and tumour behaviour has been identified (Hamper *et al.*, 1989; Collina *et al.*, 1991). Fonseca and Soares (1993), however, found that nuclear atypia in more than 20 per cent of cells was related to poor prognosis and suggested the use of DNA ploidy as a prognostic tool. The tumour has been considered to be of relatively low-grade malignant potential (Luna *et al.*, 1987; Simpson *et al.*, 1991; Noel and Brozna, 1992) although in Fonseca and Soares's recent series of 22 patients, deaths attributable to the tumour occurred in 40 per cent of cases (Fonseca and Soares, 1993). Death due to metastatic disease may occur many years after initial diagnosis and has been recorded after an interval of up to 27 years (Luna *et al.*, 1987).

Acquired stenosis of the cartilaginous ear canal is not uncommon and can occur as a result of infection, trauma or neoplasia involving the external ear canal. The only effective treatment of non-neoplastic causes is meatoplasty. Parotid tumours are a recognized cause of external canal stenosis but stenosis as the sole presenting symptom of a parotid tumour in the absence of a palpable mass has not previously been described. We stress the importance of awareness of adjacent structures when considering the cause of cartilaginous canal stenosis.

References

Batsakis, J. G., El-Naggar, A. K., Luna, M. A. (1992) Epithelial-myoepithelial carcinoma of salivary glands. *Annals of Otology, Rhinology and Laryngology* **101:** 540–542.

Collina, G., Gale, N., Visona, A., Betts, C. M., Cenacchi, V., Eusebi, V. (1991) Epithelial-myoepithelial carcinoma of the parotid gland: a clinico-pathologic and immunohistochemical study of seven cases. *Tumori* **77:** 257–263.

Corio, R. L., Sciubba, J. J., Brannon, R. B., Batsakis, J. G. (1982) Epithelial-myoepithelial carcinoma of intercalated duct origin: a clinicopathologic and ultrastructural assessment of sixteen cases. *Oral Surgery, Oral Medicine, Oral Pathology* **53:** 280–287.

Di Palma, S. (1994) Epithelial-myoepithelial carcinoma with co-existing multifocal intercalated duct hyperplasia of the parotid gland. *Histopathology* **25:** 494–496.

Donath, K., Seifert, G., Schmitz, R. (1972) Zur diagnose und ultrastruktur des tubulären speichgangcarcinoms. Epithelial-myoepitheliales schaltstrückcarcinom. *Virchows Archiv A Pathological Anatomy and Histopathology* **356:** 16–31.

Fonseca, I., Soares, J. (1993) Epithelial-myoepithelial carcinoma of the salivary glands. A study of 22 cases. *Virchows Archives A. Pathological Anatomy and Histopathology* **422:** 389–396.

Hamper, K., Brugmann, M., Koppermann, R., Caselitz, J., Arps, H., Askensten, U., Auer, G., Seifert, G. (1989) Epithelial-myoepithelial duct carcinoma of salivary glands: a follow-up and cytophotometric study of 21 cases. *Journal of Oral Pathology and Medicine* **18:** 299–304.

Horinouchi, H., Ishihara, T., Kawamura, M., Kato, R., Kikuchi, K., Kobayashi, K., Maenaka, Y., Torikata, C. (1993) Epithelial myoepithelial tumour of the tracheal gland. *Journal of Clinical Pathology* **46:** 185–187.

Luna, M. A., Ordonez, N. G., Mackay, B., Batsakis, J. G., Guillamondegui, O. (1985) Salivary epithelial-myoepithelial carcinomas of intercalated ducts: a clinical, electron microscopic and immunocytochemical study. *Oral Surgery, Oral Medicine, Oral Pathology* **59:** 482–490.

Luna, M. A., Batsakis, J. G., Ordonez, N. G., Mackay, B., Tortoledo, M. E. (1987) Salivary gland adenocarcinomas. A clinicopathologic analysis of three distinct types. *Seminars in Diagnostic Pathology* **4;** 117–135.

Morinaga, S., Hashimoto, S., Tezuka, F. (1992) Epithelial-myoepithelial carcinoma of the parotid gland in a child. *Acta Pathologica Japonica* **42:** 358–363.

Noel, S., Brozna, J. P. (1992) Epithelial-myoepithelial carcinoma of salivary gland with metastasis to lung: report of a case and review of the literature. *Head and Neck* **14:** 401–406.

Ostrowski, M. L., Font, R. L., Halpern, J., Nicolitz, E., Barnes, R. (1993) Clear cell epithelial-myoepithelial carcinoma arising in pleomorphic adenoma of the lacrimal gland. *Ophthalmology* **101:** 925–930.

Seifert, G., Brocheriou, C., Cardesa, A., Eveson, J. W. (1990) WHO international histological classification of tumours. Tentative histological classification of salivary gland tumours. *Pathology, Research and Practice* **186:** 555–581.

Simpson, R. H. W., Clarke, T. J., Sarsfield, P. T. L., Gluckman, P. G. C. (1991) Epithelial-myoepithelial carcinoma of salivary glands. *Journal of Clinical Pathology* **44:** 419–423.

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