

Pathology in Focus

Epithelial-myoepithelial carcinoma of the parotid gland: a case report and review of the cytological and histological features

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Abstract

Epithelial-myoepithelial carcinoma (EMC) is a rare biphasic tumour of the salivary glands typically arising in the parotid. Fine needle aspiration cytology is widely used in the initial investigation of salivary gland swellings and whilst the cytological features of this tumour have been described they are not well recognized. This report describes the clinicopathological features of a case of epithelial-myoepithelial carcinoma of the parotid gland and highlights the importance of awareness of this tumour in the differential diagnosis of biphasic tumours on fine needle aspiration cytology.

Key words: Carcinoma; Epithelial Cells; Myoepitheloma; Salivary Gland Neoplasms

Case report

A 54-year-old woman presented with a two-year history of a painless lump in her left cheek that had only recently started to increase in size. On examination a smooth, firm 1 cm swelling was located at the anterior border of the right parotid gland. There was no associated facial weakness or cervical lymphadenopathy. Fine needle aspiration resulted in cellular smears containing many spherical globules of hyaline material admixed with cohesive sheets of relatively uniform epithelial cells including a background population of dispersed similar

appearing cells and stripped bare nuclei (Figure 1). A population of larger cells with clear amphophilic cytoplasm and small central nuclei was also identified (Figure 2). The cytological interpretation was that of a biphasic tumour and the differential diagnosis considered between that of a pleomorphic adenoma and adenoid cystic carcinoma. Computed tomography (CT) investigation showed an irregular enhancing mass in the left superficial lobe of the parotid with some deep lobe extension, but without associated lymphadenopathy. Chest X-ray, full blood count, urea, electrolytes and liver function tests were all normal. The tumour was excised via a superficial

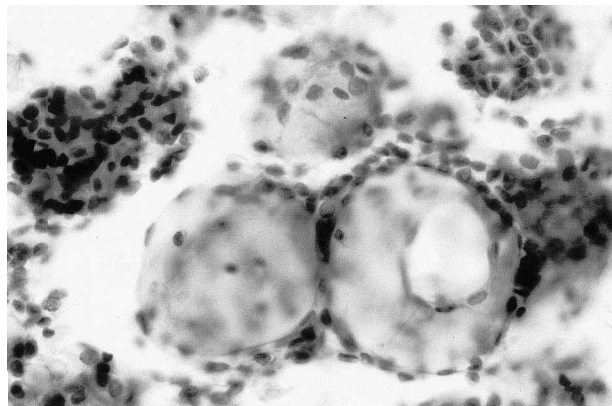


FIG. 1

Globules of hyaline basement membrane material surrounded by bland cuboidal epithelial cells (Pap; $\times 400$)

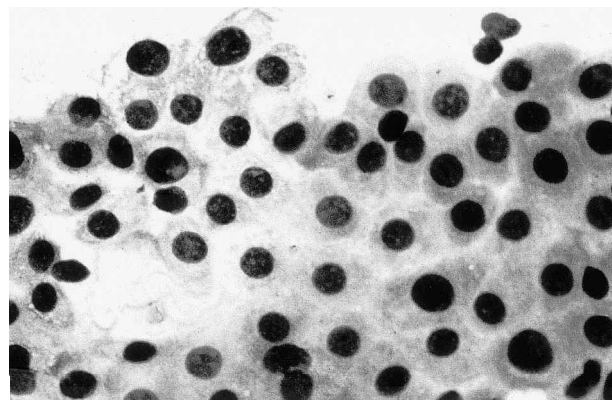


FIG. 2

Sheets of cells with amphophilic cytoplasm (MGG; $\times 400$)

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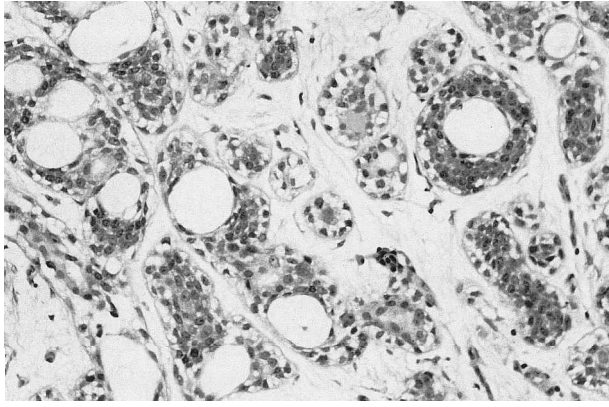


FIG. 3

Tumour nodule composed of duct-like structures lined by a two cell population (H & E; ×200)

parotidectomy approach. At the time of surgery the tumour was found to be surrounding the buccal branch of the facial nerve and was separated from it leaving the nerve intact. The specimen was received as multiple pieces of firm lobulated tan tissue measuring in aggregate 70 × 55 × 30 mm. Microscopic examination revealed well-circumscribed, non-encapsulated nodules of tumour composed of duct-like structures lined by cyokeratin positive cells with oncocytic cytoplasm surrounded by S100 positive myoepithelial cells with clear cytoplasm (Figure 3). Solid sheets of these clear cells were also present. The appearances were those of an epithelial-myoeplithelial carcinoma. As the tumour was received piecemeal, completeness of excision could not be confirmed. A staging bone scan and CT of the neck and chest were all negative. The patient was referred for radiotherapy and went on to receive 30 Gy over 25 fractions. There was no evidence of disease at nine months follow-up.

Discussion

Approximately one per cent of salivary gland tumours are epithelial-myoeplithelial carcinomas (EMC).^{1–4} Although they have a predilection for the parotid gland they may arise in minor salivary glands^{2,4,5} and rarely in extra-oral sites such as the paranasal sinuses,^{4,5} pharynx⁴ and bronchus.⁶ Whilst it is a tumour predominantly of elderly women, with a peak incidence in the seventh decade, cases have been reported over an age range of eight to 103 years.^{3,7}

Clinical presentation is not specific and is usually that of a long-standing and progressively enlarging painless mass. Facial nerve palsy occurs only rarely.⁸ Although the tumour may appear well-circumscribed at surgery it is not encapsulated and may be multinodular. Microscopically on histological examination the tumour is biphasic and characterized, as in this case, by tubules lined by an inner layer of cyokeratin-positive bland cuboidal epithelial cells surrounded by an outer layer of S100 positive myoepithelial cells. Characteristic extracellular globules of hyaline basement membrane material are present.^{9,10} Typically the tumour nodules have an infiltrative margin and perineural and vascular invasion are present. In keeping with other biphasic tumours the diagnosis may be difficult histologically and other salivary gland tumours such as pleomorphic adenoma, adenoid cystic carcinoma and also tumours with a predominant clear cell population such as clear cell carcinoma and sebaceous carcinoma should be considered before rendering the diagnosis. In

addition, metastatic carcinoma, in particular renal cell carcinoma should be considered as the two tumours may appear similar microscopically.

Fine needle aspiration cytology is a baseline investigative tool in the assessment of patients with salivary gland swellings. Whilst it is an accurate method of distinguishing neoplastic from non-neoplastic lesions it may not be possible to always accurately predict a specific tumour type due to the overlapping spectrum of cytological appearances found in a wide variety of salivary gland neoplasms. This can result in a differential diagnosis given in the cytology report in those tumours not characteristic of a specific entity.

There are few reports on the cytological features of EMC^{11–16} and in only one of these was the diagnosis considered prior to surgery.¹¹ A combination of three-dimensional well-defined cell clusters, often surrounded by homogenous acellular material, clear cytoplasm in the peripheral cells and fragments of acellular hyaline non-fibrillary material have been suggested as being sufficiently characteristic to suggest the diagnosis on aspirates.¹² However, as highlighted by this case, the presence of a biphasic tumour population with hyaline globules prompted the more common differential diagnosis of pleomorphic adenoma and adenoid cystic carcinoma, underscoring the poor recognition that EMC has received in the cytology literature. EMC is generally regarded as a low grade malignancy. However, in a series of 22 cases 41 per cent of tumours recurred and death was attributable to the tumour in 40 per cent of cases suggesting a more aggressive behaviour than previously considered.³

Histological features that may predict a worse prognosis include solid growth pattern, nuclear atypia, DNA aneuploidy and a high proliferative activity.⁹ There are few reports on the effectiveness of different treatment modalities, although wide surgical excision is considered the first choice of treatment.¹⁷ Adjuvant radiotherapy may be of use in preventing local recurrence.^{3,18} The role of chemotherapy is uncertain.

In summary, EMC is a rare salivary gland tumour with a distinctive histological appearance that should be considered in the differential diagnosis of biphasic tumours on fine needle aspiration cytology.

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Dr H. Deere takes responsibility for the integrity of the content of the paper.

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