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Case Report

Epimyoepithelial carcinoma – an uncommon salivary gland tumour

M.D.S. Lokuhetty [1], I.V. Premathilake [2], C. Amarasinghe [3]

Abstract:
Epithelial myoepithelial carcinoma is a rare salivary tumour of low malignant potential arising mainly in the parotid gland. It has a classical biphasic histological appearance comprising small tubules and glandular lumina surrounded by clear myoepithelial cells. Ancillary tests are not necessary for diagnosis. We report a case of epithelial myoepithelial carcinoma arising in a 56-year old male and discuss its differential diagnosis.

Introduction

Epithelial myoepithelial carcinoma (EMC) of the salivary gland is an uncommon salivary gland neoplasm. It is composed predominantly of well formed glandular elements surrounded by myoepithelial cells, although a spectrum of morphological changes have been described (1). It has been known by a variety of benign names in early literature before its malignant potential was identified (2,3,4,5,6,7). Donath and co-workers introduced the term epithelial myoepithelial carcinoma in German literature in 1972 (8). Corio et al introduced this term to English literature in 1982 (9).

Case report

A 56 year old male presented with a mass in the left cheek of two years duration. The lump had gradually increased in size. The overlying skin had ulcerated approximately 6 months before presentation. The mass was surgically excised together with the overlying skin.

The specimen consisted of a skin covered well-circumscribed mass measuring 35mm in greatest dimension. The surface was nodular with three small ulcers ranging in size from 20 to 60mm. The cut surface was whitish gray with a vague lobulated appearance. A small cyst measuring 8mm in diameter containing straw colored fluid was present. Necrotic and hemorrhagic areas were absent. Normal salivary gland tissue was not identified (figure 1).

Microscopically the tumour was well circumscribed with a lobulated growth pattern. It had a predominantly organoid biphasic pattern with small duct lumina lined by cuboidal cells surrounded by sheets of clear polygonal myoepithelial cells. Duct lumina contained eosinophilic protinaceous material. Pleomorphism and mitotic activity were absent in both the epithelial and myoepithelial components (Figures 2,3 and 4). A spindled myoepithelial areas was present . There was invasion of salivary gland tissue. The nerves and blood vessels were not involved.

Discussion

Epithelial and myoepithelial carcinoma is an uncommon tumor in the salivary gland accounting for 1.1% of all epithelial salivary gland neoplasms (1). Several other studies indicate an average incidence of about 1% (5,10,11,12,13). In a study of 511 salivary epithelial neoplasms done in Sri Lanka, only 2 cases (0.39%) of EMC were identified (14).

EMC is predominantly a tumour of major salivary glands (1) involving mainly the parotid gland, as was seen in this patient. EMC is also been reported in the submandibular gland and intra oral minor salivary glands (1,15).

[1] *Department of Pathology , Faculty of Medicine, Colombo.*
[2,3] *Department of Pathology, Cancer Institute, Maharagama.*
Correspondence: *M.D.S. Lokuhetty, Department of Pathology, Faculty of Medicine, Kynsey road, Colombo 8.*
Telephone: 695300 – 343 E-mail: dillokuhetty@hotmail .com

Figure 1. Macroscopic tumour

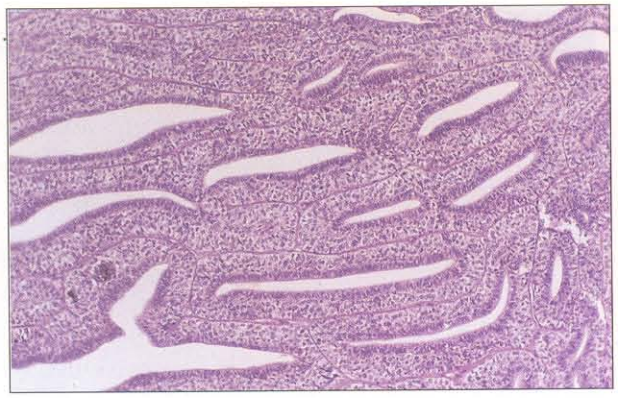

Figure 2. Epmyoepithelial carcinoma

EMC is also very rarely seen in other organs, containing salivary type seromucinous glands including the breast, sweat glands, bronchi and the maxillary antrum (16).

Microscopically EMC has a characteristic organoid biphasic appearance with ducts lined by cuboidal epithelial cells surrounded by clear myoepithelial cells as seen in this case. A rim of PAS positive basement membrane material of variable thickness surrounds the outer myoepithelial cells. Clear myoepithelial cells are usually more numerous than the cuboidal duct epithelial cells. In some areas myoepithelial cells form sheets and may even appear spindly. The cuboidal epithelial cells may also form sheets, discrete tumour nests or an organoid pattern. Cystic areas have been observed with epithelial papillary proliferations projecting in to these cystic spaces. There is no cytologic atypia and mitotic activity is sparse. Although the tumours appear well circumscribed grossly, microscopic examination frequently reveals infiltration of adjacent tissues (1). Anaplastic transformation is also documented in EMC (17).

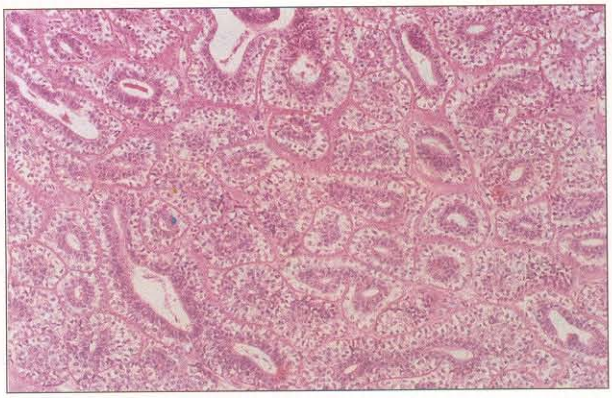

Figure 3. Epmyoepithelial carcinoma

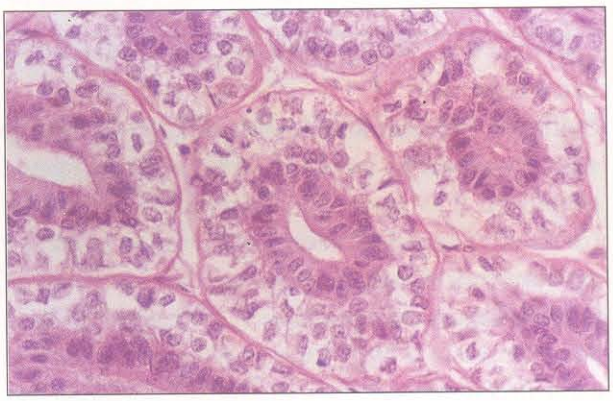

Figure 4. Epmyoepithelial carcinoma

On most occasions routine microscopic appearance of this tumor is diagnostic. Other ancillary diagnostic tests (immunohistochemistry and ultrastructural evaluation) are not routinely done, although imunohistochemical evaluation will show cytokeratin positivity in epithelial cells and positive smooth muscle actin in clear myoepithelial cells (1).The differential diagnosis of EMC comprising predominantly of clear cells includes mucoepidermoid carcinoma, acinic cell adenocarcinoma, sebaceous carcinoma, oncocytoma, clear all adenocarcinoma of minor salivary glands and metastatic renal cell carcinoma. Unlike in these tumours EMC will show the classic biphasic appearance at least focally.

When EMC is composed predominantly of spindled myoepithelial cells the differential diagnosis includes myoepithelioma, neurofibroma, leiomyoma and haemangiopericytoma. Mixed tumors, adenoid cystic carcinoma and polymorphous low grade adenocarcinoma are also composed of both luminal (ductal) and abluminal (myoepithelial) cells.

EMC does not have the characteristic myxochondroid stroma observed in mixed tumors. The cribriform pattern of growth seen in adenoid cystic carcinoma is not a feature of EMC. The cells in adenoid cystic carcinoma are also smaller and have more hyperchromatic, irregular and angulated nuclei. In polymorphus low grade adenocarcinoma clear cells form only a minor component and a biphasic pattern is not seen. Polymorphus low grade adenocarcinoma is also a rare tumor in the parotid gland unlike EMC (1).

EMC is described as a low grade malignancy with frequent recurrences and metastases to peri parotid and cervical lymph nodes. Distant metastases and death are reported as rare events (1,9,10,12,13,17). Most recurrences manifest within five years of resection of the primary tumor (1). Distant metastasis has been reported in sites such as the lung, kidneys and brain (1). Most investigators have not found any histological features associated with a poor prognosis. However Fonseca and Soares reported nuclear atypia involving more than 20% of tumour cells as indicating a poorer prognosis (18).

There are no definite treatment recommendations for EMC. Surgical treatment comprising partial or complete parotidectomy with or with out radiation therapy are the widely practiced treatment options.

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