Epithelial Myoepithelial Carcinoma of the Submandibular Salivary Gland

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Abstract

Epithelial myoepithelial carcinoma is a rare low grade malignant salivary gland neoplasm that most commonly occurs in the parotid gland but can also arise in minor salivary glands. We report a case of primary epithelial myoepithelial carcinoma of submandibular salivary gland in a 65 year old female who presented with left cheek swelling of six months duration. CT scan revealed a localized submandibular salivary gland tumour mass not involving surrounding tissues. Histological examination showed a mixture of ductal elements: cuboidal cells bordering small lumina surrounded by polygonal clear cells of myoepithelial component without any evidence of nuclear atypia or mitotic figures.

Introduction

Epithelial-myoadenoidal carcinoma (EMC), also known as adenomyoepithelioma, is an uncommon, low-grade epithelial neoplasm composed of variable proportions of ductal and large, clear-staining, differentiated myoepithelial cells. It comprises less than 1% of all epithelial salivary gland neoplasms.1 It is predominantly a tumour of the parotid gland. The mean age of patients is 60 years and about 55% of the patients are females.1

Localized swelling is commonly the only symptom, but occasionally patients experience facial weakness or pain. Overall, epithelial-myoadenoidal carcinoma is a low-grade carcinoma that recurs frequently, has a tendency to metastasize to periparotid and cervical lymph nodes, and occasionally results in distant metastasis and death.

Case Report

A 65 year old female came with complaints of left cheek swelling of 6 months duration. Medical examination revealed an enlarged, hard and non-painful swelling 7 x 5 x 4 cm in size. The patient underwent complete local excision.

We received a single tumour mass measuring 6 x 5 x 4 cm. The tumour was lobulated and grayish white in colour. Cut surface revealed an encapsulated, well circumscribed, lobulated tumour mass with central foci of chalky white deposits. A small cyst 1.5 cm in size containing haemorrhagic material was also present in the tumour mass.

Microscopic examination showed a well circumscribed, encapsulated tumour arranged in a lobular pattern. Each lobule comprised ductal and myoepithelial elements (Fig. 1). The ductal components showed eosinophilic cuboidal intercalated duct like cells bordering small lumina and were surrounded by myoepithelial component showing large polygonal clear cells (Figs. 2 and 3). Numerous psammoma bodies were also seen. There was no evidence of nuclear atypia or mitotic figures. Immunohistochemistry highlighted the biphasic nature of the tumour with myoepithelial cells being positive for P 63, Calponin and S-100.

Discussion

Epithelial-myoadenoidal carcinoma (EMC), also known as adenomyoepithelioma (AME) is low grade malignant salivary gland neoplasm composed of clear myoepithelial cells that surround epithelial lined ducts resembling intercalated ducts. This term was
coined by Donath et al. EMC is also known as adenomyoepithelioma, clear cell adenoma, tubular solid adenoma, monomorphic clear cell tumour, glycogen rich adenoma, glycogen rich adenocarcinoma and clear cell carcinoma. It is considered to be derived from intercalated ducts of salivary glands.²

The most common site is parotid gland, other sites being the remaining salivary glands, sinonasal mucoserous glands and minor salivary gland of the palate. Most tumours infiltrate adjacent normal salivary gland, adipose, muscular and bony tissues.

EMCs seem to arise in two different clinical settings: either de novo or in a recurrent pleomorphic adenoma. De novo EMCs arise in normal salivary gland, tend to be more aggressive and have a short clinical history. Recurrences may not develop or may occur as a single event within a short time interval, and generally metastases develop in the lungs.

Common clinical features are sudden and rapid tumour growth, superficial ulceration, bony destruction and nerve infiltration.

In our case, patient presented with de novo left submandibular swelling since 6 months without any metastasis.

Most EMCs show a characteristic nodular or multinodular growth pattern and classic biphasic tubular histology. The other variants are verocay like change, sebaceous differentiation, dedifferentiated EMC, oncocytic EMC, EMC ex pleomorphic adenoma, double-clear EMC and EMC with myoepithelial anaplasia.²

In routine histologic sections, the
morphologic variants of myoepithelial cells are clear, spindle, stellate, polygonal, angular, epithelioid and plasmacytoid.

Immunohistochemically, epithelial component is selectively well highlighted by all cytokeratins (CK) and epithelial membrane antigen (EMA). Myoepithelial component is demonstrated by S100, smooth muscle actin (SMA), p63 and vimentin. The newer markers like calponin (CALP), caldesmon (CALD), and smooth muscle myosin heavy chain may be useful tools for identifying myoepithelial cells when myoepithelial cell differentiation is not easily identified on routinely stained sections. In our case, immunohistochemistry was done to highlight the biphasic nature of the tumour. The myoepithelial cells were positive for p63, Calponin and S-100.

Low-grade stage I tumours are curable with surgery alone. Radiation therapy may be used for tumours in which resection involves a significant cosmetic or functional deficit or as an adjuvant to surgery when positive margins are present. Neutron-beam therapy is effective in the treatment of poor-prognosis malignant salivary gland tumours.

Patients with EMC showing marked cellular pleomorphism, tumour necrosis, angiolymphatic invasion and perineural invasion have a poor prognosis.

In conclusion, EMC is a rare low-grade malignancy with distinct histological appearance. It carries a low potential for lymph node or distant metastasis but relatively high tendency for local recurrences. The biologic behaviour also varies, depending on the site of involvement.

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References

BENEFITS OF TAXING SUGAR-SWEETENED BEVERAGES
Consumption of sugar-sweetened beverages has increased in recent decades; evidence suggests that consumption of these beverages contributes to obesity and adverse health outcomes. The authors discuss the potential public health and economic benefits of taxing sugar-sweetened beverages.