

Epithelial-myoepithelial carcinoma

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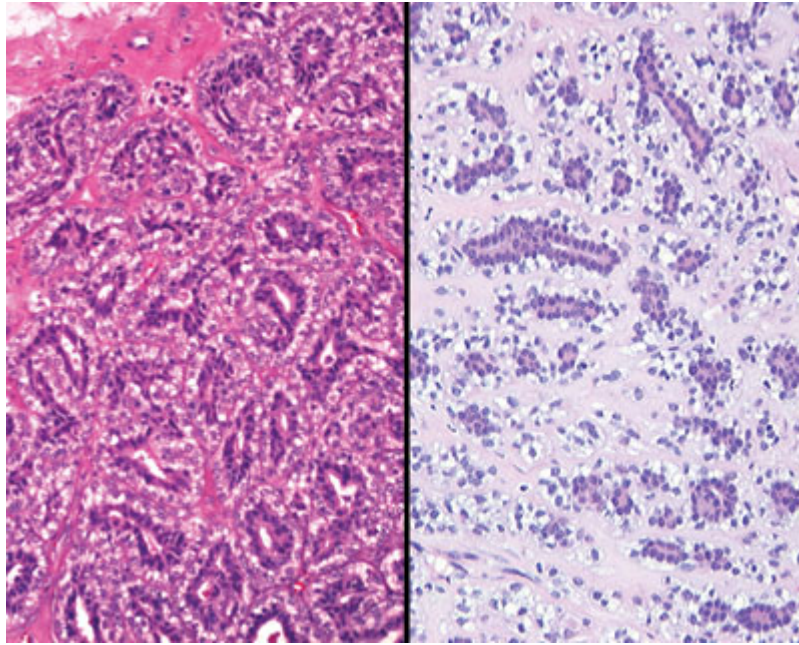


Figure 1. Images show the gland-like and duct-like architecture of the biphasic tumor cell population in EMC.

Epithelial-myoepithelial carcinoma (EMC) is an uncommon salivary gland malignancy, representing 1% of all salivary gland tumors. Women are more frequently affected (2:1). The incidence of EMC peaks between the ages of 50 and 60 years, although the age range is broad. The parotid gland is most frequently affected (60%), but both major and minor salivary glands can be involved. Patients typically present with a painless, slowly growing mass; occasionally there is rapid growth, pain, or facial weakness.

In the major salivary glands, EMC characteristically appears as a circumscribed mass with expansile borders. Minor gland tumors may be multinodular and poorly circumscribed, and they may exhibit hemorrhagic and necrotic areas. The tumor cells are arranged in lobules, sheets, or discrete nests that are separated by an eosinophilic, hyalinized, basement-membrane-like material (figure 1). Perineural and vascular invasion can be present, and bone invasion may occur.

EMC is defined as a malignant tumor made up of a variable ratio of two cell types, typically arranged in duct-like structures (figure 1). The biphasic tumor is formed by a peripheral, outer layer of myoepithelial cells that surrounds a central, inner layer of ductal epithelial cells. The myoepithelial cells are large, with clear-staining cytoplasm. The inner epithelial cells are cuboidal, with eosinophilic cytoplasm and uniform round nuclei (figure 2). Cytologic atypia is mild or absent in most tumors, but occasionally cellular and nuclear pleomorphism and increased mitotic figures are present. The duct- or gland-like lumens may contain eosinophilic proteinaceous material (which is reactive to periodic acid-Schiff staining). Myoepithelial immunohistochemical markers (e.g., smooth-muscle actin, calponin, and p63) highlight the clear myoepithelial cells, and the ductal epithelial cells react with cytokeratins.

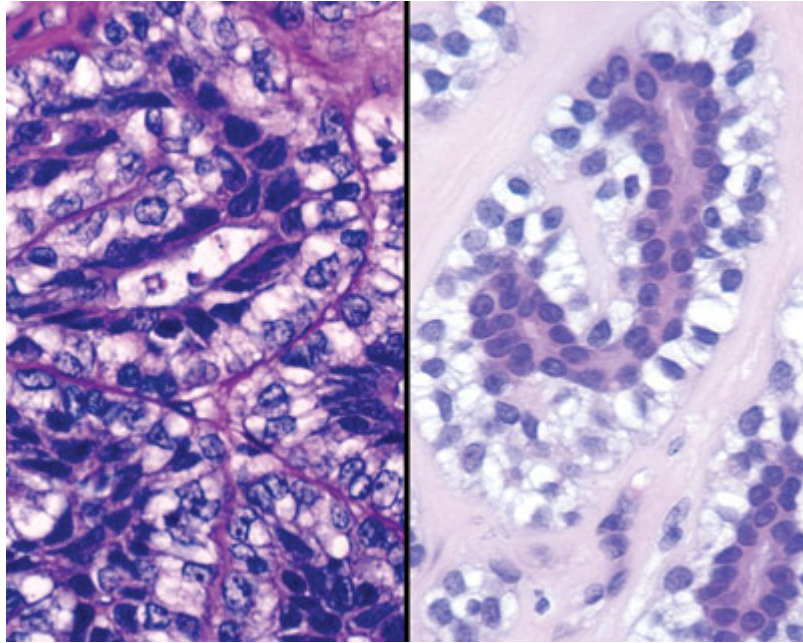


Figure 2. Eosinophilic cuboidal duct-like cells form lumens and are surrounded by larger, polygonal, clear myoepithelial cells.

EMC must be distinguished from primary salivary gland neoplasms that contain clear cells (e.g., pleomorphic adenoma, myoepithelioma, oncocytoma, mucoepidermoid carcinoma, and clear cell carcinoma) and metastatic clear cell tumors (e.g., renal cell carcinoma and thyroid carcinoma).

Overall, the prognosis is good—80% survival at 5 years following complete surgical excision. Recurrences have been reported in as many as 40% of patients, usually within 5 years of the initial surgery. Metastasis occurs in about 15% of patients, usually in the cervical lymph nodes, lung, liver, and kidney. Fewer than 10% of patients die from disease complications. Large tumor size and rapid growth are associated with a worse prognosis.

Suggested reading

Fonseca I, Soares J. Epithelial-myoepithelial carcinoma. In: Barnes L, Eveson JW, Reichart P, Sidransky D, eds. Pathology and Genetics of Head and Neck Tumours. Kleihues P, Sobin LH, series eds. World Health Organization Classification of Tumours. Lyon, France: IARC Press; 2005:225-6.
 Fonseca I, Soares J. Epithelial-myoepithelial carcinoma of the salivary glands. A study of 22 cases. Virchows Arch A Pathol Anat Histopathol 1993;422:389-96.