Mucoepidermoid carcinoma

Lester D.R. Thompson, MD

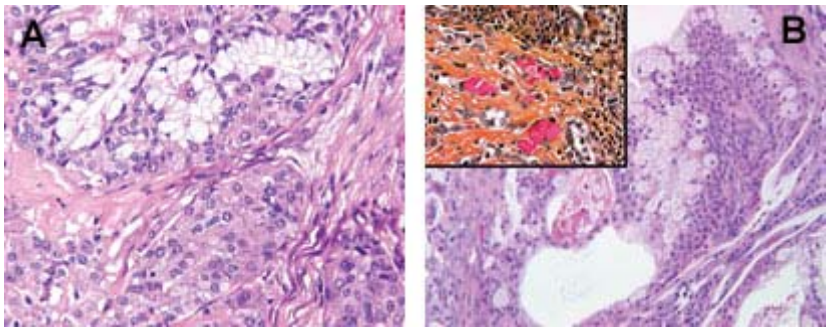

Figure. ***A:*** *A blending of intermediate, pavemented cells and goblet-type, mucus-filled cells is characteristic of an MEC.* ***B:*** *Cystic spaces are common in low-grade tumors. This MEC has an intermediate cell population and many mucocytes. Inset: An MEC demonstrates a strong reaction on a mucicarmine stain.*

Mucoepidermoid carcinoma (MEC) is the most common primary salivary gland malignancy, accounting for approximately 25% of all malignancies. More than half of these cases involve the major salivary glands, primarily the parotid glands. MEC can also involve a variety of other sites that have minor mucoserous glands. Women are more commonly affected than men (3:2), and the mean age at onset is in the 5th decade of life. MEC is also the most common salivary gland malignancy in children.

The tumor usually forms as a painless, fixed, slowly growing swelling of widely varying duration that sometimes goes through a phase of accelerated growth immediately before clinical presentation. Symptoms include tenderness, otorrhea, dysphagia, and trismus. Intraoral tumors are often bluish-red and fluctuant, and they may resemble mucoceles or vascular lesions. They occasionally invade the underlying bone.

MECs may be circumscribed and variably capsulated or infiltrative and fixed; the latter characteristics generally apply to higher-grade tumors. Areas of scarring are relatively common. Most tumors are smaller than 4 cm in diameter. Cysts of variable sizes are often present, and they usually contain brownish fluid. MEC cells form sheets, islands, duct-like structures, and cysts of various sizes. The cysts may be lined with intermediate, mucous, or epidermoid cells, and they are filled with mucus (figure, A). Papillary processes may extend into the cyst lumina, and this is occasionally a conspicuous feature.

The tumor is primarily made up of three cell types in widely varying proportions: intermediate, mucous, and epidermoid.

- The intermediate cells frequently predominate; their appearance ranges from small basal cells with scanty basophilic cytoplasm to larger and more oval cells with more abundant pale eosinophilic cytoplasm that appears to merge into epidermoid or mucous cells.
- Mucous cells (mucocytes) can occur singly or in clusters, and they have pale and sometimes foamy cytoplasm, a distinct cell boundary, and small, peripherally placed, compressed nuclei. Mucocytes often form the lining of cysts or duct-like structures (figure, B). Occasionally mucocytes are so scanty that they can be identified with confidence only by using stains such as mucicarmine (figure, B, inset).

- Epidermoid cells may be uncommon and focally distributed. They have abundant eosinophilic cytoplasm, but they rarely show keratin pearl formation or dyskeratosis. Oncocytic metaplasia is seen occasionally.

Higher-grade tumors show evidence of cytologic atypia, a high mitotic frequency, and areas of necrosis, and they are more likely to show neural invasion. Stromal hyalinization is common and sometimes extensive.

MECs exhibit remarkable variability in their clinical behavior. Several microscopic grading systems based on a numerical score have been advocated as a means of predicting outcome. These systems rely on subjective evaluations of the relative proportions of the various cell types, the degree of cellular atypia, mitotic frequency, presence of necrosis, and invasive characteristics.

MECs must be distinguished from necrotizing sialometaplasia, chronic sialadenitis, cystadenoma, cystadenocarcinoma, squamous cell carcinoma, epithelial-myoepithelial carcinoma, clear cell carcinoma (not otherwise specified), and metastatic tumors.

The prognosis is dependent on the clinical stage, site, grading, and adequacy of surgery. Survival is greater than 95% for low-grade tumors, and regional metastases are rare. The death rate increases to 45% for high-grade tumors. Tumors that transgress surgical margins have a very high recurrence rate, particularly high-grade tumors. Death is usually caused by uncontrolled locoregional disease and metastases to the lung, bone, and brain. Treatment is surgical excision with or without neck dissection. Radiotherapy is generally palliative in advanced tumors; it has little impact on prognosis.

Suggested reading

Brandwein MS, Ivanov K, Wallace DI, et al. Mucoepidermoid carcinoma: A clinicopathologic study of 80 patients with special reference to histological grading. Am J Surg Pathol 2001;25:835-45.
Goode RK, Auclair PL, Ellis GL. Mucoepidermoid carcinoma of the major salivary glands: Clinical and histopathologic analysis of 234 cases with evaluation of grading criteria. Cancer 1998;82:1217-24.
Goode RK, El-Naggar AK. Mucoepidermoid carcinoma. In: Barnes EL, Michael L, eds. Pathology and Genetics of Tumours of the Head and Neck. Kleihues P, Sobin LH, series eds. World Health Organization Classification of Tumours. Lyon, France: IARC Press, 2005:227-8.