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Low-Grade Cribriform Cystadenocarcinoma of the Parotid Gland

This is the case of a 44-year-old woman with a one-year history of a left pre-auricular mass. The surgical specimen is a 5 centimeter diameter tan-brown irregularly-shaped tissue whose cut surfaces are brown with cystic spaces. Microscopic sections show cystic and dilated ductal spaces lined by cells forming irregular, variably-sized secondary spaces. These spaces are arranged in a cribriform pattern that is reminiscent of breast ductal hyperplasia. (Figure 1) The ductal cells lining the spaces are small, multilayered, and generally bland. The superficial cells show apocrine-type cytoplasmic snouting. There is no significant nuclear atypia or mitotic activity noted. Necrosis is also absent. (Figure 2) Based on these features, we signed the case as a low-grade cribriform cystadenocarcinoma (LGCCC).

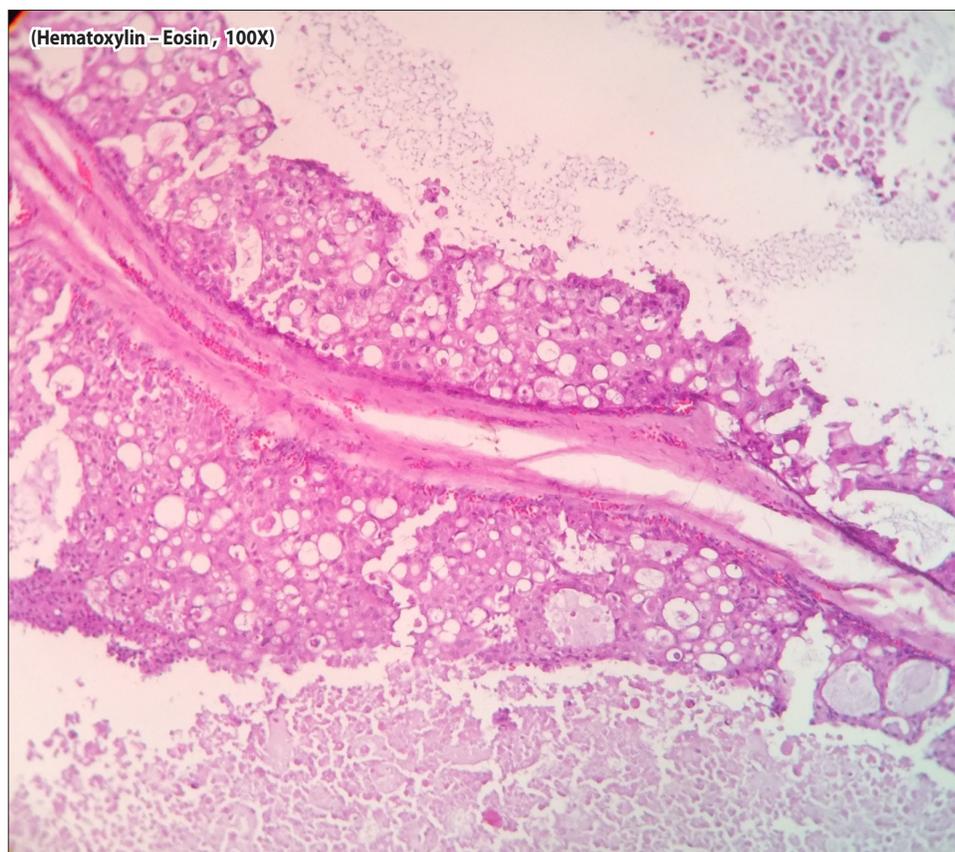


Figure 1. Dilated ductal spaces with cribriform structures reminiscent of breast ductal hyperplasia (Hematoxylin-eosin, 100X magnification)

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Figure 2. Bland ductal cells forming cribriform spaces. Superficial cells display apocrine-type snouting (arrows) (Hematoxylin-eosin, 400X magnification).

LGCCC is an uncommon tumor presenting primarily as cystic parotid masses in elderly females. The histologic hallmark of this tumor is its morphologic resemblance to the spectrum of breast lesions ranging from ductal hyperplasia to low-grade ductal carcinoma-in-situ.¹⁻⁴

Microscopic sections show an unencapsulated tumor consisting of single or multiple cysts lined by proliferated small and bland ductal cells with fine chromatin and small nucleoli. Within the cystic spaces, the cells are often arranged in a cribriform pattern with anastomosing intracystic micropapillae lining the cavity. Many superficial cells show apocrine-type secretions. Thus, the over-all appearance is quite comparable to breast lesions that are termed atypical ductal hyperplasia and low-grade ductal carcinoma-in-situ.¹⁻⁴ Focal invasion into the surrounding tissue can be seen. Perineural or vascular invasion however is typically absent. Cellular pleomorphism and mitoses are also usually absent and necrosis is rare. Occasional tumors however may demonstrate a transition to an intermediate or high-grade cytology with the appearance of scattered mitoses and focal necrosis.^{1,2}

This tumor needs to be distinguished from a conventional cystadenocarcinoma. The latter is a more frankly invasive tumor with smaller duct-like structures that infiltrate into the salivary parenchyma and surrounding connective tissue. A papillary-cystic variant of acinic cell carcinoma will have areas of acinar differentiation and a greater degree of epithelial proliferation.^{1,5} A high-grade salivary duct carcinoma will have a high-grade cytology with more frequent necrosis,

mitoses, and pleomorphism.⁶ Special stains that help in the differential diagnosis include Periodic Acid-Schiff (PAS) stain with Diastase digestion (diastase-resistant cytoplasmic granules in an acinic cell carcinoma), and S100 (strong diffuse positivity in LGCCC).¹

LGCCC is treated by complete surgical excision. Although there are only a few reported cases with follow-up, to our knowledge, none, to date, have recurred.¹⁻⁴

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