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Epithelial-Myoepithelial Carcinoma of the Parotid Gland

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Summary: We describe an unusual salivary neoplasm, an epithelial-myoepithelial carcinoma (EMEC) of the parotid gland, that occurred in a 64-year-old man. A CT scan showed a fairly well defined heterogeneous lesion with smooth margins and slight enhancement. MR images showed a lesion with intermediate T1-weighted signal intensity and a relatively high T2-weighted signal intensity. Although the imaging characteristics of EMEC on both CT and MR studies are nonspecific, clinicians and radiologists must be aware of its high local recurrence rate, which has been reported to approach 50% in some series, and thus the need for periodic postoperative imaging to detect early recurrence.

Index terms: Carcinoma; Salivary glands, neoplasms

Epithelial-myoepithelial carcinoma (EMEC) of the salivary glands was described by Donath et al in 1972 (1). It is a rare tumor, accounting for less than 1% of all salivary gland neoplasms, that arises most commonly in the parotid gland but has also been described in the submandibular gland and in the minor salivary glands (2, 3). There is a female predominance, with a peak occurrence in the seventh decade (4). EMEC is a low-grade malignant tumor that may commonly recur locally after resection. Distant metastasis rarely occurs (4–6). Sonographic findings of EMEC have been described (7). This article describes the cross-sectional imaging characteristics of a case of EMEC.

Case Report

A 64-year-old man had painless parotid swelling for 2 years. His medical and surgical history was noncontributory. On physical examination, a 4-cm fixed, slightly tender mass was palpated in the left parotid gland. There was no palpable cervical adenopathy, and facial nerve function was intact.

A computed tomographic (CT) scan showed a nonhomogeneously enhancing mass within the deep portion of the left parotid gland extending through the stylomandibular tunnel. The lesion protruded into and displaced the parapharyngeal space fat. There was no gross invasion of the parapharyngeal space fat planes and there was a smooth interface with the remainder of the parotid gland (Fig 1A). There was no evidence of pathologic adenopathy. A magnetic resonance (MR) image showed that the mass had an intermediate T1-weighted signal intensity, as compared with skeletal muscle, and high, slightly nonhomogeneous T2-weighted signal intensity (Fig 1B and C). The tumor's interface with the parotid gland appeared more clearly defined than on the CT scan, and there was no invasion of the adjacent fat planes. The margins of the lesion with the adjacent fat planes were smooth and fairly well defined. The posterior margin of the lesion was more sharply delineated by MR imaging than by CT.

A total parotidectomy was performed. Pathologic examination showed a uniform pattern of salivary ductular epithelium surrounded by clear cells of myoepithelial origin (Fig 1D). The background was hyalinized. The tumor was of uniform morphology without other elements to suggest pleomorphic adenoma. Some limited infiltration into surrounding parotid tissue was appreciated, but the tumor remained confined to the parotid gland. The diagnosis of EMEC was made. Over the first postoperative year, the patient has done well, without evidence of either local recurrence or metastases.

Discussion

This unusual salivary tumor, so descriptively named by Donath et al in 1972, was included in the World Health Organization classification of salivary tumors in 1991. This tumor occurs in older persons (sixth decade and beyond), and has a slight (55%) female predominance. Statistics from 57 patients seen at the Armed Forces Institute of Pathology reveal that the parotid gland is the most commonly involved (75%), followed by the submandibular gland (12%) and palate (7%) (8). They present as bulky, bosselated, slow-growing tumors that may become large.
EMEC tends to grow in a bulky lobulated fashion, with necrosis and hyalinization of large tumor nodules. The tumor has a distinctive histopathologic pattern with a proliferation of ductular structures. The ducts may be seen in cross section or longitudinally, and they may be densely packed together or separated by abundant dense hyaline material. The inner cells of these ductules constitute the epithelial component of EMEC. These mildly to moderately pleomorphic cells have irregular ovoid shapes, may overlap and have prominent nucleoli and fine chromatin. Mitotic figures are not common. The outer cell layer that surrounds the ductules is the clear cell myoepithelial component of EMEC. The nuclei are smaller than those of the epithelial cells, with a definitely condensed and triangular appearance. The cells vary from being “naked nuclei” to having abundant clear cytoplasm. The clear areas are glycogen-positive and should stain for periodic acid Schiff and be sensitive to diastase. This case was further unusual in that there were cystic and papillary components.

The pathologic differential diagnosis includes other clear cell salivary tumors such as clear cell carcinoma, mucoepidermoid carcinoma, acinar cell carcinoma, sebaceous carcinoma, metastatic renal cell or (rarely) balloon cell melanoma, and clear cell oncocytoma. Pleomorphic adenoma and canalicular adenoma should also be added to this differential diagnosis, as they also form ductal structures. EMEC most resembles clear cell carcinoma, a related tumor composed almost entirely of myoepithelial clear cells. The ductal elements separate these two tumors.

As EMEC histologically recapitulates the intercalated duct/myoepithelial unit, it would be tempting to theorize that these cells give rise to this neoplasm, consistent with the reserve cell theory of salivary tumorigenesis. This theory postulates that the basal reserve cells of the excretory ducts and the intercalated duct myoepithelial units are the stem cells from which all salivary tumors are derived. However, recent evidence has shown that rat salivary gland will proliferate and regenerate after injury, and that
the proliferating cells are not confined to one cell type. Even end-differentiated cells such as acinar cells have the ability to reenter the cell cycle. Thus, the pathogenesis and derivation of salivary gland tumors are even more diverse than previously thought (9, 10).

Clinically, EMEC usually appears as a bulky, slow-growing mass within the parotid gland, ranging in size from 2 to 12 cm (5). Local spread to lymph nodes has been reported (4). The usual treatment is wide surgical resection, including adjacent lymph nodes; however, there is a high reported rate of local recurrence, approaching 50% (11–13). Therefore, adequate resection with negative soft-tissue margins is the minimum recommended and necessary therapy. Recurrences, sometimes multiple, have been seen from 9 months to 28 years after initial resection (3). Resection of recurrences usually results in a good prognosis, with less than 10% of patients dying as a result of this tumor (7). Distant metastases to kidney, lung, and brain have also been reported, and are usually fatal (4).

In one study, EMEC was characterized sonographically as a cystic mass with smooth walls and posterior acoustic enhancement (7). In that report, the tumor was followed up for a period of 4 years without significant change in its size or appearance. The findings in this case of EMEC indicate that the CT and MR appearances of EMEC are nonspecific, and that EMEC cannot be differentiated from more common parotid neoplasms on the basis of its imaging characteristics.

In conclusion, the CT and MR findings of a case of EMEC of the parotid gland are presented. Because the imaging findings of this rare tumor appear to be nonspecific, the initial role of the radiologist is the preoperative identification and localization of the mass. However, once the diagnosis of EMEC is made, both the clinician and the radiologist must be vigilant regarding periodic postoperative imaging because of the potentially high rate of local recurrence. Early identification of a recurrence of EMEC appears to be important in that repeated resections have yielded an excellent prognosis.

Acknowledgments

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References