CASE REPORT

Bilateral Epithelial-Myoepithelial Carcinoma: An Extraordinary Tumor of the Paranasal Sinuses

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ABSTRACT

A 54-year-old woman presented with a 6-month history of swelling on the right side of her face. On physical examination she was found to have a huge mass in the right nasal cavity. Magnetic resonance images of the paranasal sinuses revealed a soft tissue mass in the right maxillary sinus and a mass was also seen in the left maxillary sinus. On histopathologic examination, the tumor had a lobular structure with infiltrating margins. Two cell types, an outer layer of myoepithelial and an inner layer of duct-like cells, were found. On immunohistochemical examination, myoepithelial cells stained positively for calponin, p63, GFAP, S-100 protein, α -smooth muscle actin, cytokeratin-14. The tumors were resected completely and no recurrence or metastasis was found 30 months after surgery. We describe here an unusual case of epithelial-myoepithelial carcinoma (EMC) arising from the paranasal sinuses. This is the first case report in the literature describing bilateral EMC in the maxillary sinuses.

KEYWORDS: Epithelial-myoepithelial, paranasal sinus, bilateral, carcinoma

Epithelial-myoepithelial carcinoma (EMC) of intercalated duct origin is an uncommon clear cell carcinoma of the salivary glands, primarily arising from the parotid gland, though isolated cases have been described from the maxillary sinus, trachea, larynx, hypopharynx, and lacrimal gland.¹⁻³

EMC originating from the paranasal sinus is very rare. Herein, we report the first case of bilateral EMC arising in the maxillary sinuses.

CASE REPORT

A 54-year-old woman came to our department in December 2004 with the complaint of a 6-month history of swelling on the right side of her face. She also reported progressively increasing pain in the previous 2 months. On ear, nose, and throat examination, a fixed, firm mass of 3 × 3 cm was found in the right maxillary sinus area. Nasal endoscopic

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evaluation revealed a mass originating from the right lateral nasal wall and extending to the hard palate and posterior end of the right inferior turbinate in the nasal cavity, while the left nasal cavity was normal. No cervical lymphadenopathy was found. Magnetic resonance images (MRI) of the paranasal sinuses showed a soft tissue mass in the right maxillary sinus extending to the right nasal cavity and hard palate, with bony destruction of the posteromedial wall of the maxillary sinus and inferior orbital wall. A mass was also seen filling the left maxillary sinus, with bony destruction of its posterior wall, extending to the hard palate and narrowing the nasal cavity (Fig. 1). Surgery was planned only for the right mass due to its obstruction of the nasal cavity and the swelling of the right maxillary sinus area; the mass in the left sinus was not obstructing the nasal cavity and the patient had no complaint regarding the left side.

The patient underwent a right modified maxillectomy including a palatal excision under general anesthesia. Tumors were found to be two discrete masses according the intraoperative evaluation. The right tumor was resected completely and histologic diagnosis was EMC. A second operation

was planned for the left mass because of this histological result. The patient underwent left subtotal maxillectomy 2 weeks after the first operation and histologic diagnosis was EMC again. The patient's postoperative course was unremarkable. Follow-up at 30 months revealed no evidence of disease.

Pathologic Findings

The first material, surgically resected from the right side, measured $6.5 \times 5.5 \times 3.5$ cm in diameter. The cut surface showed well circumscribed gray-white multiple nodules, with the largest measuring 3 cm. The second material, surgically resected from the left side, measured $4.5 \times 4 \times 2$ cm in diameter. The cut surface showed gray-white color, without necrosis or hemorrhage. Microscopically, both tumors were multinodular and infiltrated adjacent tissues (Fig. 2). Two cell types, an outer layer of myoepithelial and an inner layer of cuboidal eosinophilic duct-like cells, were found. Cuboidal eosinophilic cells were surrounded by polygonal myoepithelial cells (Fig. 3). The periepithelial stroma was hyalinized. Immunohistochemically, myoepithelial cells

Figure 1 (A) Coronal and (B) axial magnetic resonance images of the paranasal sinuses, showing a tumor mass in the right maxillary sinus extending to the right nasal cavity and also tumor mass in the left maxillary sinus.

Figure 2 Microscopic view of epithelial-myoepithelial carcinoma, multinodular growth pattern with infiltrating margins in adjacent tissues (hematoxylin and eosin $\times 100$).

stained positively for calponin, p63, GFAP, S-100 protein, α -smooth muscle actin, cytokeratin-14 (Fig. 4). Ductal cells stained for cytokeratin AE1/AE3 and also these keratin cocktails stained myoepithelial components weakly. The Ki-67 proliferation index was low.

DISCUSSION

EMC is a rare neoplasm, first described in 1972 by Donath et al, which most commonly occurs in salivary glands and comprises about 1% of all salivary gland tumors.⁴⁻⁶ Most cases of EMC arise in the parotid gland; a few arise in the submandibular gland and minor salivary glands. Patients are mostly female in their fifth to eighth

decades, ranging from 8 to 103 years.^{7,8} EMC located in the maxillary sinus appears to be very unusual. There have been few studies reported about paranasal sinus EMC.⁹⁻¹³ Seethala and associates showed that most cases of EMC occurred in the parotid gland (62.1%), sinonasal mucoserous glands (10.3%), palate (8.6%), and submandibular gland (8.6%).¹³ To our knowledge, this is the first case report in the literature about bilateral EMC in the maxillary sinuses.

Diagnosis of EMC is based on conventional light microscopy and is confirmed by immunohistochemical and ultrastructural investigation. Histologically, the tumor is characterized by well-defined tubules with two cell types: an outer layer of myoepithelial cells with clear cytoplasm surrounds an inner lining of eosinophilic cuboidal epithelial cells.¹⁴ Perineural invasion and infiltrating margins

Figure 3 Cuboidal cells surrounded by myoepithelial cells (hematoxylin and eosin $\times 400$).

are characteristically present in most cases of EMC.¹⁴ In our case, myoepithelial cells appeared to be polygonal-shaped and stained positively for calponin, p63, GFAP, S-100 protein, α -smooth muscle actin and cytokeratin 14.

EMC is considered to be a low-grade malignancy. In keeping with its low-grade malignant nature, the recurrence rate of EMC is 35 to 50% and the metastatic rate is 8.1 to 25%.¹⁵ Seethala and colleagues found that the recurrence rate of EMC was 36.3%; survival rates were 93.5% and 81.8% for 5 and 10 years, respectively.¹³ Our patient has shown no sign of recurrence or metastasis during the 30 months of follow-up. EMC is characterized by a relatively good prognosis, in contrast to salivary duct carcinoma.⁷ The only morphological feature found to correlate with the prognosis was the presence of nuclear atypia in more than 20% of the tumor cases.¹⁵

There is no consensus regarding the optimal treatment of this neoplasm, largely due to its rarity. Wide surgical excision with a clear margin is the treatment of choice because of the tumor's tendency to infiltrate locally.⁸ If the tumor is more than 4 cm in diameter, combined radiotherapy and surgery has been recommended. Radiotherapy may be of benefit in preventing local recurrence. The effect of chemotherapy is uncertain in this neoplasm.⁸

SUMMARY

In conclusion, localization of EMC in bilateral maxillary sinuses is unusual. The patient we present is the first case described in the world literature of bilateral EMC arising in the maxillary sinuses.

Figure 4 Myoepithelial cells stained positively for calponin (calponin $\times$ 400).

Close and prolonged follow-up is recommended despite its low-grade malignancy.

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