



AN UNUSUAL PRESENTATION OF EPITHELIAL - MYOEPITHELIAL CARCINOMA OF THE NASAL CAVITY

Gokcen Coskun Bal ¹, Guler Berkiten ², Muhlis Bal ³, Yavuz Atar ², Ziya Salturk ², Ilhan Topaloglu ⁴, Gulcin Kamali ⁵

¹ Department of Otorhinolaryngology, Mus Government Hospital, Mus, Turkey

² Department of Otorhinolaryngology, Okmeydani Training and Research Hospital, Istanbul, Turkey

³ Department of Otorhinolaryngology, Ataturk Government hospital, Sinop, Turkey

⁴ Department of Otorhinolaryngology, Yeditepe University Hospital, Istanbul, Turkey

⁵ Department of Pathology, Okmeydani Training and Research Hospital, Istanbul, Turkey

ABSTRACT

Myoepithelial carcinoma is a rare malignant epithelial neoplasm composed of variable proportions of salivary gland tumor. Most of cases which have been reported in the literature were located in salivary glands, except for a few cases occurring in unusual locations such as breast, lacrimal gland, nose, paranasal sinus, trachea, bronchus, and lung. We report a case 59-year-old female manifesting as nasal obstruction with left recurrent epistaxis and on nasal endoscopic examination was found to have a polypoid tumor in the left nasal cavity. The CT scan demonstrated a soft tissue mass without extension to the nasopharynx or bony destruction. The tumor was surgically treated with endoscopic excision successfully from the left nasal septum. Histopathologic examination revealed the tumor to consist of a mixture of a solid, tubular structures. Based on the histological and immunohistochemical findings, the tumor was diagnosed to be an myoepithelial carcinoma. The patient has had neither recurrence nor metastasis 8 months postoperatively. The purpose of this study was to describe myoepithelial carcinoma with nasal location, which is exceptionally reported in the literature.

Key Words : Myoepithelial carcinoma, nasal cavity, polypoid, tumor

INTRODUCTION

Epithelial-myoepithelial carcinoma(EMCs) was first reported by Donath (1), and was formally named by Seifert and Sabin in 1991 (2). EMCs are tumors arise in the salivary glands, predominantly the major salivary gland and the parotid gland. They are uncommon, approximately about 1% of all salivary gland tumors (3). However, they have rarely been observed in tissues other than the salivary glands, such as the breast (4), lacrimal gland (5), trachea (6), bronchus (7), lung (8) paranasal sinuses (9) and nasal cavity (10-14)

Nasal cavity one of the unusual site for these tumors. We here reported the case of EMC presented with left recurrent epistaxis and obstruction in the left nasal cavity as a polypoid tumor. EMC could easily be confused with other tumors histopathologically. Because of wide spectrum of morphological patterns of epithelial-myoepithelial carcinomas pathologist has to be familiar with these variations. Recurrence and metastasis rates of epithelial-myoepithelial carcinoma varied from respectively in different reports. EMCs have low rate of recurrence and rare distant metastases that's why EMCs are low-grade malignancy tumors

CASE REPORT

A 59-year-old female complaints of nasal obstruction and recurrent epistaxis of the left nostril for 10 months. On nasal endoscopic examination, a hemorrhagic polypoid mass based on the posterior end of the left nasal septum was observed. No lymph node was palpable in the neck. Computed tomography (CT) showed soft tissue density in the left nasal cavity (*Figure.1*). There is neither bony destruction nor invasion into the maxillary sinüs. The patient underwent endoscopic surgery under general anesthesia. Nasal cavity was observed then Killian incision is made, mucosa of the nasal septum were easily detached from the cartilage or bone. The septal cartilage and the perichondrium was intact on both sides. The tumor was excised by en bloc resection with a safety margin. The surface of the tumor looked like nasal mucosa. The tumor was elastic hard. The histologic diagnosis of a postoperative biopsy specimen from the nasal cavity was EMC. There was no evidence of recurrence after 2 years.

PATHOLOGICAL FINDINGS

The tumor which was connected to the mucosa of the septum was grayish and grossly size is $3 \times 2 \times 1$ cm. The specimen was fixed by two kinds of fixative, 10% formalin and 2.5% glutaraldehyde. Myoepithelial and ductal cells, were found, with the former being the predominant component. Myoepithelial cells appeared ovoid and spindle in shape and some had clear cytoplasm (*Figure.2*). The spindle cells were similar to fibroblast and smooth muscle cells in shape with eosinophilic fibril cytoplasm. The small ovoid myoepithelial cells with hyperchromatic nuclei. Though we had a clear - cut histological picture of EMC in post-operative microscopy with histochemistry (PAS and PAS with diastase), the diagnosis was reconfirmed with immunohistochemistry (CK, SMA, S-100 positive). Immunohistochemistry shows cytokeratin, S-100 protein positive in neoplastic myoepithelial cells and in some cases vimentin, actin and myosin. CK (cytokeratin) and SMA (smooth muscle actin) positivity highlighting the epithelial and myoepithelial component respectively while the positive S-100 protein highlighted both the epithelial and myoepithelial component.

DISCUSSION

EMC is a rare tumor which most commonly occurs in salivary glands and predominantly arise from the parotid gland and while a few in the submandibular gland and minor salivary glands (15,16). Tumors morphologically similar to salivary gland EMC have been reported in the breast (malignant adenomyoepithelioma), lacrimal gland, nose, paranasal sinus, trachea, bronchus and lung.

EMCs are extremely rare tumors in the nasal cavity. The ages of EMC cases ranged from 22 to 70 years, and the ratio of male to female was 1:3. The tumors arose from the nasal septum or the inferior nasal turbinate and paranasal sinuses. In one case, a tumor has invaded into the maxillary sinus at the time of the initial diagnosis, in another case tumor has invaded contralateral nasal cavity (Medial maxillectomy and radiation therapy done), 7 months follow up revealed extensive bone metastases, the post-operative courses of other cases were uneventful with no recurrence during follow-up periods of 7–40 months.

Because of variable appearance and the varied histological characteristic of EMC differential diagnosis is wide in the nasal cavity. Therefore, the careful examination of serial tumor sections is required for the definite diagnosis of EMC (17,18). The myoepithelial cells in epithelial-myoepithelial carcinoma appear ovoid to spindle shaped and exhibit clear cytoplasm, while the ductal cells are usually cuboid in shape. Correct diagnosis can be done by finding ductal cell differentiation and duct-like structure that exhibit myoepithelial differentiation. Immunohistochemically, the myoepithelial cells react positively with a panel of antibodies including S-100 protein, smooth muscle actin, vimentin and low molecular weight cytokeratin (19,20).

There is no consensus regarding the optimal treatment of this neoplasms, largely due to its rarity. In the literature, EMC in the nasal cavity were treated by tumor excision or in few cases treated by endoscopic excision (10-14) . Radiotherapy has been tried to prevent local recurrence in some reports (21,22), but up to date its role is uncertain because of inadequate statistical analysis due to the small number of cases so far investigated. On the other hand, there are no reports dealing with chemotherapy in treatment of EMCs except the case report of Pierard et al. (23) which reported that chemotherapy might allow the stabilization of pulmonary metastases of EMC of the submandibular gland. As a result, the efficacy of adjuvant chemotherapy therefore remains uncertain at present. We can say that the treatment of choice for EMC is complete resection with wide margins.

Intervals between diagnosis to recurrences, and distant metastases in the salivary glands for EMCs, Cho et al. (24) have reported 5 years (range, 1–19 years) for recurrence and 15 years (range, 4–20 years) for metastasis. Therefore, a much longer follow-up period seems to be required for the evaluation of the prognosis of EMCs in the nasal cavity. Fortunately, our patient has shown no sign of recurrence or metastasis during the 8 months since the operation. Although this tumor has been described as a low-grade malignancy, close and prolonged follow-up is recommended.

Although sinonasal epithelial-myoepithelial carcinoma is extremely rare, its occurrence is still possible therefore the head and neck surgeon and the surgical pathologist should be aware of this entity.

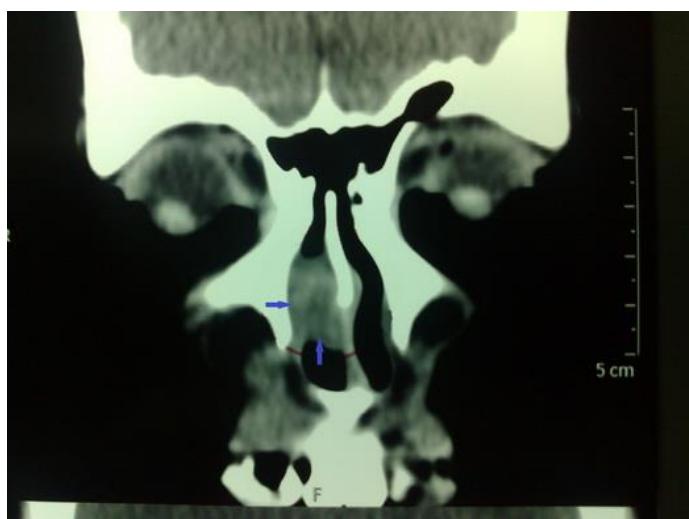


Figure.1 Coronal image of CT- soft tissue density in the left nasal cavity.

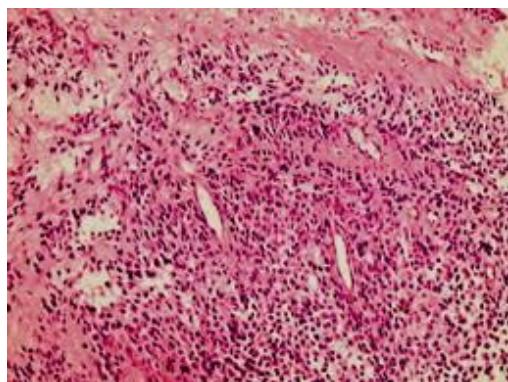


Figure.2 Microscopic picture of Epithelial - Myoepithelial Carcinoma, The myoepithelial component is represented by the cells having clear cytoplasm (Hematoxyline-eosine stain)

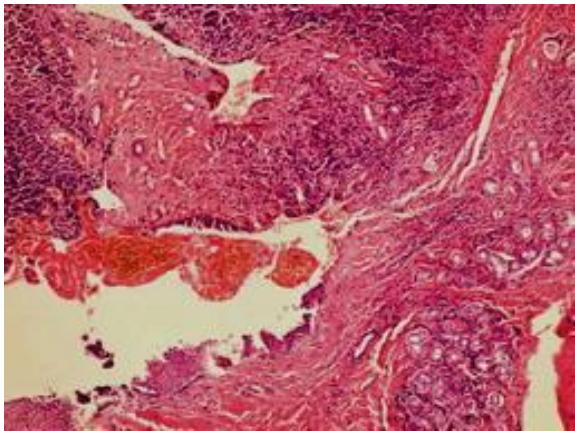


Figure.3 Ovoid myoepithelial cells, Tumor cells are composed of two types of cells, i.e., duct-forming cells and cells surrounding these cells

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