

An Unusual Location of a Voluminous Epithelial–Myoepithelial Carcinoma in Nasal Cavities: A Case Report

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Abstract: Epithelial–myoepithelial carcinoma is an uncommon and low-grade malignant tumor, arisen most frequently in the salivary glands. It represents 1% of malignant tumors of salivary glands. The extraoral location is rarely reported in the literature, and nasal cavities represent one of the most unusual sites for these tumors. Originally was described in 1972 by Donath and al. It is essentially a tumor of older adults, with female predominance. Epithelial–myoepithelial carcinoma is a low-grade malignant tumor, because it has low rate of recurrence, and rare distant metastases. The treatment of choice is surgical resection with wide margins, and postoperative radiotherapy has been tried to prevent local recurrence. The prognosis of nasal cavities location is not well known, because the number of cases published is limited, but it seems to be good because in published cases, no recurrence or metastasis has been reported in this location. We report a case of a voluminous epithelial–myoepithelial carcinoma arisen in the left nasal cavity of a 59-year-old man, with review of literature. The patient underwent oncological surgery, with total excision of the tumor by lateral rhinotomy approach, and received adjuvant radiotherapy. He had neither recurrence nor metastasis, with a follow-up of 24 months after surgery. The purpose of this study is to describe the pathologic, therapeutic features, and prognosis of epithelial–myoepithelial carcinoma occurring in nasal cavities one of the most unusual locations, which is rarely reported in the literature.

Keywords: Voluminous epithelial-myoeplithelial carcinoma, Nasal cavity, surgical treatment, prognosis.

INTRODUCTION

Epithelial–myoepithelial carcinoma (EMC) is an uncommon malignant tumor which occurs typically in the salivary glands. Originally was described in 1972 by Donath and *et al.* [1-3], and was formally named by Seifert and Sobin in 1991 [4]. It represents 1% of malignant tumors of salivary glands [5], and most frequently arisen in the parotid gland (80% of cases) [6, 7, 4]. However, the extraoral location is rarely observed, a few cases arisen in extraoral mucoserous glands was reported in the literature [8], and nasal cavities represents one of the most unusual sites for this tumors [9]. EMC is a low-grade malignant tumor, because it has low rate of recurrence and rare distant metastases [4, 10]. The purpose of this study is to describe the pathologic, therapeutic features, and

prognosis of epithelial–myoepithelial carcinoma in nasal cavities.

CASE REPORT

We report a case of a 59-year-old man, without pathological antecedent, complaints of bilateral and progressive nasal obstruction with recurrent epistaxis and hyposmia since 14 month, associated with ipsilateral hypoacusis and diplopia. On rhinological examination, we observed a deformation of the nasal pyramid with a voluminous polyploid mass, occupying the totality of left nasal cavity and exceeding the left nostril (figure 1), with absence of the nasal flow of both sides. The remainder of the ENT examination revealed left sero-mucosal otitis, and no lymphadenopathy was founded in the neck.



Fig-1: Voluminous polypoid tumor, occupying the totality of left nasal cavity and exceeding the left nostril

The computed tomography scan revealed a voluminous and heterogeneous tumor in the left nasal cavity, with invasion into the ipsilateral maxillary sinus,

and erosion of the nasal bone, of the ethmoidal cells, and extending into the nasopharynx with invasion into the left orbit (figure 2).

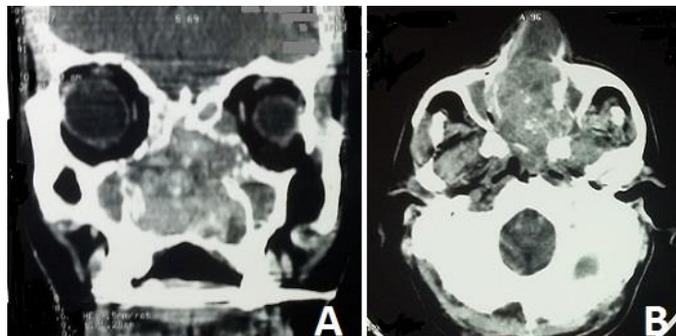


Fig-2: Coronal (A) and axial (B) images of CT scan, showing a heterogeneous mass occupying the nasal cavities with erosion of adjacent bones

Histopathological examination of the biopsy showed a mixture proliferation of Myoepithelial and ductal cuboidal epithelial cells. The Myoepithelial cells were predominant, with clear cytoplasm and vesicular nuclei. The immunohistochemically study revealed the cells reacting positively to cytokeratin (5,6, 7), to S-100

protein, to epithelial membrane antigen, to CD117, to vimentin and to actin.

The patient underwent oncological surgery with total excision of the tumor by lateral rhinology approach, and received adjuvant radiotherapy. He had neither recurrence nor metastasis after operation with a follow-up of 24 months (figure 3) after surgery.



Fig-3: 24 months after surgery, showing nasal cavities without recurrence of the tumor

DISCUSSION

Epithelial myoepithelial carcinoma [11] is a rare and low-grade malignant neoplasm, which most frequently occurs in salivary glands and predominantly arise in the parotid gland [6, 7, 4]. It composed of ductal epithelial and myoepithelial, which derive epithelial and myoepithelial cells of the intermediate glandular canals [10].

It is rarely observed in tissues other than the salivary glands, a few cases occurring in unusual locations were reported in the literature, such as breast [12], lacrimal gland [13], nasal cavity [14, 15], paranasal sinus [16], trachea [17], bronchus [18], and lung [19]. The occurrence in the nasal cavity is exceptional [9]. EMC is essentially a tumor of older adults occurring after the age of 70 [6, 7], with female predominance, and the ratio of male to female was 1/3 [5, 20]. Clinically, often it presents by nasal obstruction and recurrent epistaxis [9, 15], with polypoid tumor on endoscopic examination.

The features of tumor on computed tomography scan and MRI are nonspecific, but it can be locally aggressive with a variable degree of bone erosion. The diagnosis of the EMC is confirmed by histopathological examination with immunohistochemical study. It is characterized by the presence of ductal structures, which are lined by an inner layer of cuboidal epithelial-type cells, and an outer layer of myoepithelial cells with clear or eosinophilic cytoplasm and eccentric vesicular nuclei [11, 21]. The epithelial cells are strongly positive with cytokeratin stains and negative for myoepithelial markers. The myoepithelial cells are usually only weakly positive for cytokeratin but stain strongly with myoepithelial markers (S-100, smooth muscle actin, vimentin, p63, HHF35, and calponin) [21].

The differential diagnosis of EMC in the nasal cavity is wide, because of varied histological characteristic [22]. It includes mucoepidermoid carcinoma, and other tumors containing clear cells, such as clear cell myoepithelioma, or acinic cell carcinoma [23].

In the literature there is no consensus regarding the optimal treatment of EMC in the nasal cavities. The treatment of choice for EMC is complete surgical resection with wide margins [24], by lateral rhinotomy or by endoscopic excision in few cases [15, 14].

Postoperative radiotherapy has been tried to prevent local recurrence [23, 25], and it is indicated for tumors larger than 4 cm [23, 25], but its role is uncertain because of unadequate statistical analysis due to the small number of cases investigated. The chemotherapy might allow the stabilization of metastases of EMC [26].

The prognosis of the EMC of nasal cavities is not well known, because the number of cases published is limited, but in published cases, no recurrence or metastasis has been reported in this location. In the salivary glands location, the local recurrence rates for EMC range from 30% to 50% [27, 4, 10]. The regional lymph node metastases occur in 18% of patients, and distant metastases in 7-25% [28-30] in different reports. The overall 5-year survival rate is 80%. Our patient has shown no sign of recurrence or metastasis after surgery, with a follow-up of 24 months.

CONCLUSION

Epithelial myoepithelial carcinoma is a rare low-grade malignant tumor, most frequently arisen in the salivary glands. The sinonasal location remains extremely rare, but its occurrence is still possible. The prognosis of EMC in nasal cavities seems to be good, because in our case report and in published cases, no recurrence or metastasis has been reported in this location, But a much longer follow-up period is required for the evaluation of its prognosis. Although this tumor has been described as a low-grade malignancy, it should be treated aggressively, because it can be locally invasive and because it can recur, and give rise to distant metastases in extranasal location.

Contributions of the authors

All the authors contributed to the realization of this work.

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