



## Epithelial Myoepithelial Carcinoma of Maxillary Sinus —A diagnostic dilemma.

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### Abstract:

Epithelial – Myoepithelial Carcinoma (EMC) is a rare malignant salivary gland neoplasm that most commonly occurs in the Parotid gland, but can also arise in the Minor Salivary Glands. EMC of the maxillary sinus extremely rare. We describe here a case of a 74-year-old patient who presented with maxillary swelling for 4months and nasal discharge for 3 months. Computed Tomography Scan revealed an expansile soft tissue mass in the left maxillary sinus eroding all its walls. In View of high suspicion of malignancy, Left maxillectomy was done. Histopathological examination confirmed Epithelial Myoepithelial Carcinoma with Positive Reaction to CK, Vimentin, Smooth Muscle Actin (SMA) and S-100.

### Introduction

Epithelial-myoepithelial carcinoma (EMC), a rare neoplasm was first recognised in 1956 but was described in 1972 by Donath *et al.*<sup>1</sup> It has been reported under a variety of names including benign category like adenomyoepithelioma, clear cell adenoma, tubular solid adenoma,

monomorphic clear cell tumor, glycogen-rich adenoma and malignant when glycogen-rich adenocarcinoma and clear cell carcinoma ,tubular adenocarcinoma and Epithelial Myoepithelial carcinoma (when distinct biphasic tumor histology ) .<sup>2</sup>

It is most common in the parotid gland, representing about 1% of all salivary gland tumors. In addition, the major sites of involvement are the maxillary sinus, trachea, larynx, hypopharynx, and minor salivary glands, although it has also been reported in the mucoserous glands of the upper and lower aerodigestive tract<sup>1</sup>. Most of the reported cases of spindle cell and clear cell myoepithelioma have occurred in the parotid gland ,whereas most hyaline myoepitheliomas have been reported in minor salivary glands , particularly the Palate.<sup>2</sup> It is common in women in their fifth to eighth decades , however the reported age range being 8-103 years .<sup>1</sup>

EMC is a low grade malignant biphasic salivary-type tumor and the diagnosis is based on conventional light microscopy, confirmed by immune histochemical and ultrastructural investigation. Histologically, the tumor is composed of small tubules lined with two cell types: an outer layer of myoepithelial cells with clear cytoplasm surrounding an inner lining of eosinophilic cuboidal epithelial cells resembling intercalated ducts. An association has been described between epithelial –myoepithelial carcinoma and intercalated ducts hyperplasia and is suggested to be its precursor lesion because the tubular growth pattern of this tumor epitomizes this phenotype .<sup>2</sup> The clear cell type tumors are composed of small tubules lined by a single layer of small cuboidal cells surrounded by one or more layers of prominent clear cells<sup>1</sup>

Although the histological appearance of EMC is usually characteristic, there may be considerable morphological variation in the form of regressive changes ,Schwannoma like areas, Sebaceous differentiation, oncocytic change ,and a .double clear' appearance. Thus, the disease can overlap with other salivary-type tumors and differential diagnosis from many tumors of the salivary glands is necessary<sup>2</sup>. The morphological features cover a wide spectrum, ranging from purely epithelial aspects such as clear cell carcinoma to the appearance of a purely myoepithelial carcinoma. The recent oncocytic variant of EMC includes oncocytoma, oncocytic papillary cystadenoma and cystadenocarcinoma. <sup>1</sup>

There is a range of differentiation among these tumors, with both benign and malignant variants .The majority of cases with a hyaline cell morphology behave in benign fashion, although malignant may exist. Malignant forms of spindle and clear cells exist which are characterised by invasive properties and cytological atypia.<sup>3</sup> In the salivary gland, criteria that have been reported to be helpful in differentiating between benign and malignant myoepitheliomas include cytological atypia, mitotic activity and tumor infiltration into surrounding salivary

gland or other normal tissues. No specific mitotic-rate cut-off exists on which to base this distinction, although cytological atypia is not required for a diagnosis of myoepithelial carcinoma.<sup>4</sup>

### **Case report:**

A 74-year-old healthy man presented with swelling left maxilla since 4 months and nasal discharge since 3 months. At clinical examination, nasal cavity was normal. Computed Tomography Scan revealed an expansile soft tissue mass in the left maxillary sinus eroding all its walls. Maxillectomy specimen in multiple pieces was sent to the Department of Pathology, PGIMS Rohtak. On gross inspection, specimen showed portion of maxillary bone with growth measuring 8x6x5cm.also received in same container, multiple pieces of growth measuring together 7x6x1.5cm.Both surface of bones showed polypoidal growth. Sections were cut and prepared by the conventional routine, and stained with hematoxylin and eosin and by other appropriate methods.. Microscopically, the tumor was adenocarcinoma and infiltrating and destroying adjacent bone. Histologically, the specimens showed two cell types, an outer layer of myoepithelial cells and an inner layer of cuboidal eosinophilic duct-like cells. The cuboidal eosinophilic cells were surrounded by polygonal myoepithelial cells. The periepithelial stroma was partially hyalinized in some areas. Thus conforming the histology of EMC.

*Immunohistochemistry.* A panel of antibodies: cytokeratin (CK), S100 protein, Vimentin,  $\alpha$ -smooth muscle actin (SMA), Desmin, Synaptophysin, and Chromogranin was applied to further sections and appropriate positive and negative controls were employed. .

The ductal cells show strong reactivity for CK. EMA showed strong positivity of the cell membranes and cytoplasm. Vimentin and SMA was positive in myoepithelial cells.S100 was focally positive. In contrast Desmin, Synaptophysin and Chromogranin were negative.

### **Discussion**

EMC is a low-grade malignancy, only rarely have high-grade or dedifferentiated EMC cases been reported.

It has been observed that some morphologically low-grade myoepithelial carcinomas behave aggressively<sup>5</sup>. 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%. 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.<sup>6</sup>Thus, in the absence of frankly malignant cytomorphology, an invasive growth pattern is the single most useful criterion for establishing malignancy in salivary EMC. In the

present cases, no atypical mitoses were observed, but infiltrative margins features associated with malignant behaviour, were observed microscopically.<sup>5</sup>

It is rare tumor and a diagnostic dilemma. It should be considered in cases showing dual tumor cell population with clear cell change in histopathology. Common differential diagnosis includes metastatic renal cell carcinoma, acinic cell carcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, oncocytoma, and sebaceous carcinoma.<sup>7</sup> Renal cell carcinoma can be distinguished by presence of multiple vessels and CD10 positivity. Acinic cell carcinoma can be excluded by immunohistochemistry since it is negative for all markers. Mucoepidermoid carcinoma shows distinct mucoid areas. EMC must be distinguished from adenoid cystic carcinoma, and these two forms can be described together as the so-called “hybrid” carcinoma, when coexistent<sup>1</sup>. sebaceous carcinoma is positive for EMA and cam5.2. Oncocytoma can be distinguished by the presence of abundant eosinophilic granular cytoplasm.

In the present cases, the myoepithelial cells stained positively for SMA and Vimentin and the ductal cells stained for CK strongly and also weakly stained the myoepithelial components. There was a weak focal reaction for S100 protein. Thus conforming the diagnosis of EMC.

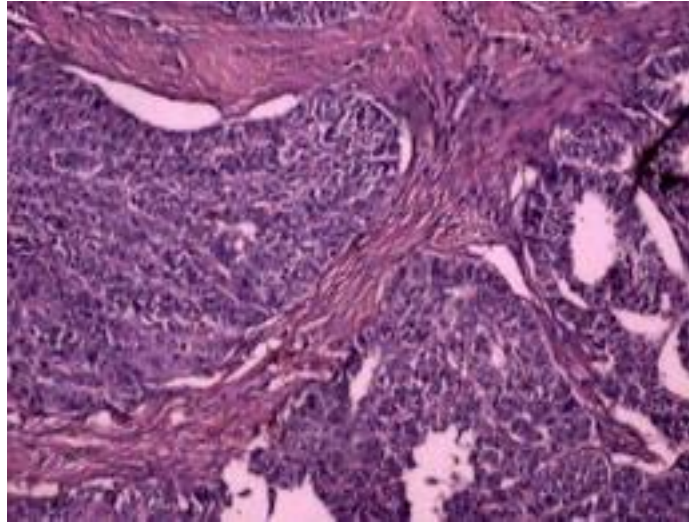
### **Treatment.**

There is no consensus regarding the optimal treatment of this neoplasm, largely due to its rarity. Close and prolonged follow up is required for this tumor because of low grade behaviour. However, wide surgical excision with a clear margin is the treatment of choice because of the tumor's tendency to infiltrate locally. If the tumor is more than 4 cm in diameter, combined radiotherapy and surgery have been recommended. Radiotherapy may be of benefit in preventing local recurrence. The effect of chemotherapy is uncertain in this neoplasm<sup>8</sup>.

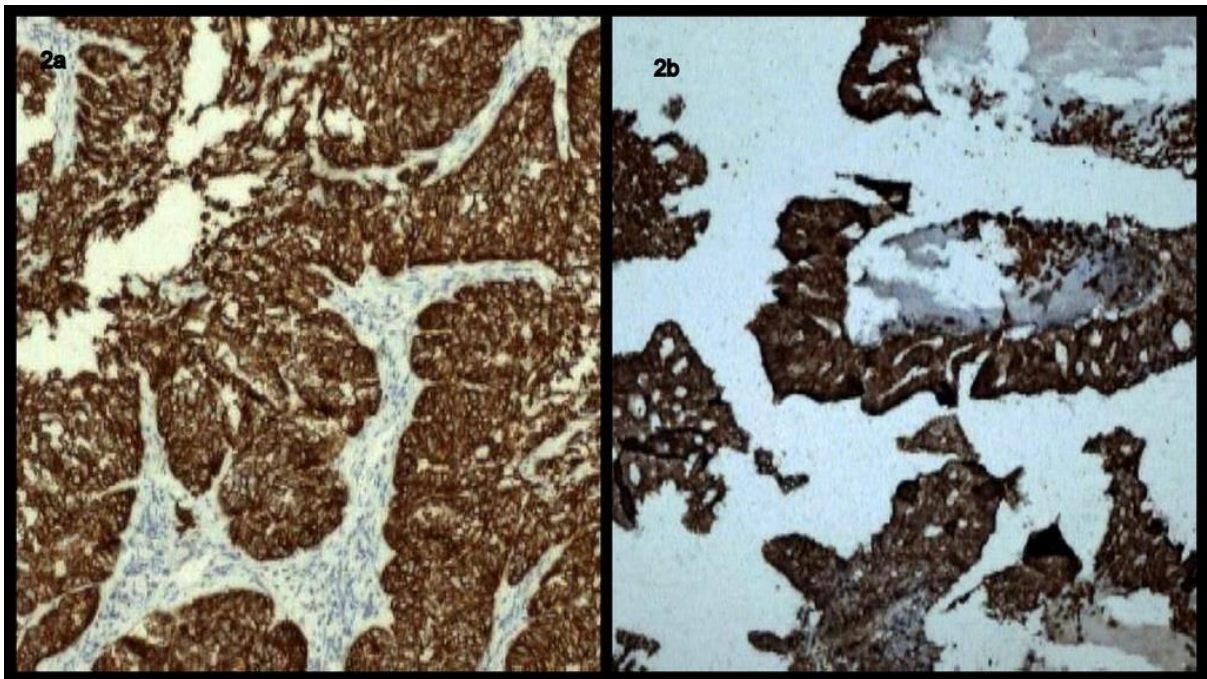
### **Conclusion**

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EMC is a rare neoplasm and maxillary bone involvement is even rarer. But despite the lack of cytological atypia (prominent nucleoli, vesicular or coarse chromatin, pleomorphism), the immunohistochemical profile, morphology of the lesion and the local infiltrative and invasive nature of the tumor cells enabled us to better define these tumors and made it possible to reach a diagnosis of EMC.



**Fig 1:** showing outer layer of polygonal myoepithelial cells and an inner layer of cuboidal eosinophilic duct-like cells



**Fig 2:** showing Ck positivity of ductal cells and vimentin positivity of myoepithelial cells.

## References

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