Epithelial-myoepithelial Carcinoma, uncommon Tumour with a rare presentation, importance of Immunohistochemistry for the Dignosis

Mayed M. Radi,[1] Hossam A. Makki,[1] Issam Al-Bozom[2]
[1]Otolaryngology Department, Hamad Medical Corporation, [2]Department of Laboratory Medicine and Pathology, Hamad Medical Corporation, Qatar.

Correspondence to: Mayed M. Radi, Email: mayedradi@yahoo.com

Epithelial myoepithelial carcinomas (EMC) are rare carcinomas of the salivary glands. We report here the 6th case of this tumor which arises at the nasal cavity. In case of any histopathological report shows up a pleomorphic adenoma in an unusual site, we are highly recommended the use of immunohistochemistry assay as an important toll for the Diagnosis of this tumor.

Keywords: ***Epithelial myoepithelial carcinoma (EMC), Nose & Maxillary sinus, Immunohistochemistry, Smooth Muscle Actin (SMA), S- 100 protein, Salivary gland .***

INTRODUCTION

Epithelial myoepithelial carcinomas (EMC) are rare low grade carcinomas of salivary glands, they are composed of varying portions of ductal cells and clear myoepithelial cells. They constitute 1.1% of all epithelial salivary gland tumors reviewed by the AFIP[(1)] (Armed Forces Institute of Pathology, Washington DC). This is usually a tumor of the parotid gland,[(4)] rarely the tumor can be found in other sites. The small number of reported cases of (EMC) in the nose & Para nasal sinuses makes the clinical management & prognosis prediction more difficult. There are only 5 cases reported in literatures for EMC in the nose and sinuses.[(5,6,7,8,10)] We are presenting here the 6th Case.

Because of the difficulty to differentiate between (EMC) & pleomorphic adenoma by using ordinary histopathological microscopically examination we recommend doing Immunohistochemistry tests whenever we find a pleomorphic adenoma at the nose & Para nasal sinuses.

CASE REPORT

A 30 years old Indian male patient presented to our hospital with nasal obstruction mainly on the left side for the past 18 months. His condition started before 3 years when he starts to complain from nasal obstruction at the left side of the nose. On 2003, partial maxillectomy was done to the patient in (India). Excision of the mass taken place, the mass was filling the nose & the left maxillary sinus.

The histopathological diagnosis was polymorphic adenoma.

Eighteen months later he started to develop a progressive left nasal obstruction, anosmia and very slight bleeding if he tries to touch the mass.

On examination the mass is red, firm, with smooth surface filling the whole left nostril. The nasal septum was intact,

also the skin over the sinus, the orbit and the teeth all are intact.

C.T. scan confirmed the findings (Fig. 2) the mass partially filling the left maxillary sinus & the left nasal cavity.

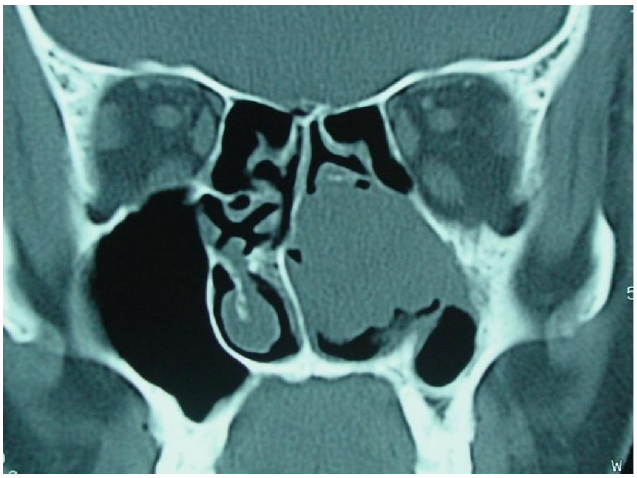

Fig 2. CT scan, mid-nasal coronal section showing Left lobulated mass filling most of the nasal cavity.

Biopsy of the nasal mass done & the result was epithelial myoepithelial carcinoma of the left nasal cavity.

When we informed the patient about the diagnosis & we discussed with him the management plane (operative management) he decided to go back to his home country (India) to complete his treatment over there.

Histopathological report:

The tumor cells are arranged in rounded organoid clusters, nests, and trabeculae (Fig. 1a) .The tumor is composed of a biphasic cell population: myoepithelial cells and ductal cells. The myoepithelial cells are characterized by their large size, polygonal shape, and clear cytoplasm. The ductal elements are composed of intercalated duct-like cells that border some lumens. They are cuboidal, with eosinophilic cytoplasm and uniform rounded nuclei (Fig. 1b). There is no significant necrosis, pleomorphism, apoptosis, vascular embolization or perineural infiltration.

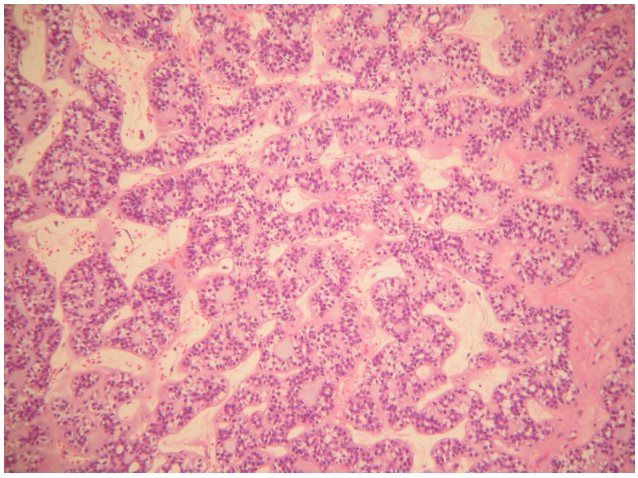

Fig 1a. Low power microscopic view shows the organoid pattern of the Tumor (Hematoxylin &Eosin x100).

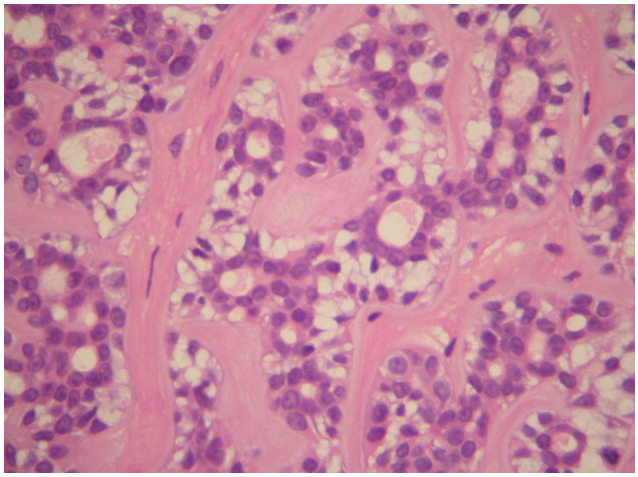

Fig 1b. High power shows the biphasic nature of the tumor (i.e. inner Epithelial cells and outer clear myoepithelial cells) (Hematoxylin &Eosin x400).

The mitotic rate is 1/10HPF. Immunoperoxidase stains highlighted the biphasic nature of the tumor, the luminal ductal epithelial cells are positive with cytokeratin cocktails (MNF-116, and CK AE1/AE3) (Fig. 1c), and the peripheral myoepithelial cells are positive with S-100 protein and Smooth Muscle Actin (SMA) (Fig. 1d).

The diagnosis is epithelial myoepithelial carcinoma of the left nasal cavity.

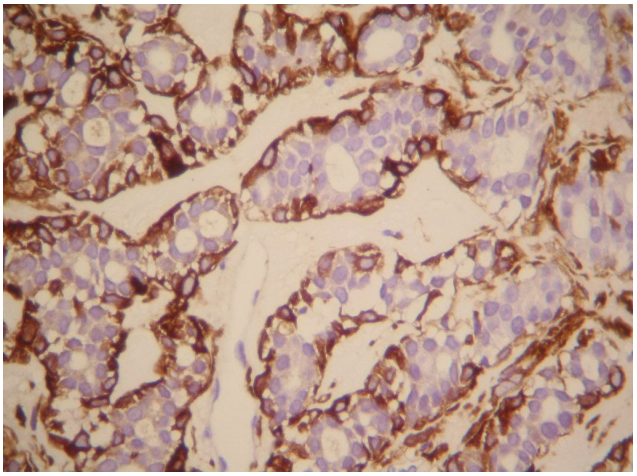

Fig 1c. Immunoperoxidase stain: Cytokeratin cocktail (MNF-116) showing The inner epithelial cells, while outer myoepithelial cells are not Taking the stain (x400).

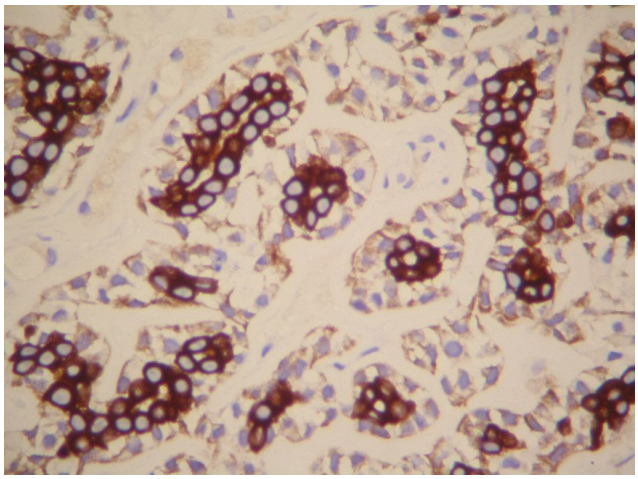

Fig 1d. Immunoperoxidase stain: Smooth muscle actin (SMA) showing Opposite staining to MNF-116)(i.e. inner epithelial cells are negative While outer myoepithelial cells are positive) (x400).

Literature review:

CASES	Age	Sex	Nationality	Site of origin	Immunohistochemical Positive Exam	Reference
1	56	male	Japanese	Nasal Septum	Myoepithelial & ductal cells (positive)	Harada 1996
2	61	female	Chinese	Nose	*	Jin 1999
3	65	female	Japan	Maxillary sinus	SMA	Sunami 1999
4	22	male	Korea	Nasal cavity	S-100 protein + SMA	Lee 2000
5	26	female	Korea	Maxillary sinus	*	Woo 2004
6	33	male	Indian (work in Qatar)	Nose and maxillary sinus	S-100 protein + SMA	Present case

* No data available.

DISCUSSION

EMC is typically a tumor of the major salivary glands, with the parotid and submandibular glands accounting for 75% and 10-12% of all cases, respectively, rare sites are nose & Para nasal sinuses,[5-8,10] Most patients tend to present with complaints of painless, enlarging mass that may have been present from weeks to months. Some patients may present with more ominous signs of malignancy such as pain or facial paralysis. Since these tend to be slow growing, low-grade malignancies, most patients will present with more benign symptoms.

Epithelial-Myoepithelial carcinoma is a challenging diagnosis particularly when it occurs outside the major salivary glands, and more challenging if an incisional biopsy is encountered. The differential diagnosis includes but not limited to: pleomorphic adenoma, monomorphic adenoma, polymorphous low grade adenocarcinoma, adenoid cystic carcinoma, myoepithelioma and others. The important findings that might differentiate EMC from

others are the peculiar microscopic features with emphasis on the biphasic nature of the tumor (epithelial and myoepithelial components) which can be easily illustrated by immunohistochemistry.

With immunohistochemistry, the ductal cells are strongly positive for cytokeratin and negative for S-100 protein and smooth muscle actin (SMA). In contrast, the clear, myoepithelial-like cells are often strongly positive for S-100 and variably positive for SMA.

Prognosis: There is a 30-50% local recurrence rate.[2,9] Metastases to local lymph nodes are seen in 18% of cases and distal metastases to lung, kidney and brain in 8% of cases, resulting in a similar mortality rate. Recurrence and metastases may occur more than 20 years after first presentation. Nuclear atypia in more than 20% of cells[9] and aneuploidy[9] may indicate a worse prognosis.

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