



Review Article

SALIVARY GLANDS EPITHELIAL-MYOEPITHELIAL CARCINOMA – UNRESOLVED CLANDESTINE

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ABSTRACT

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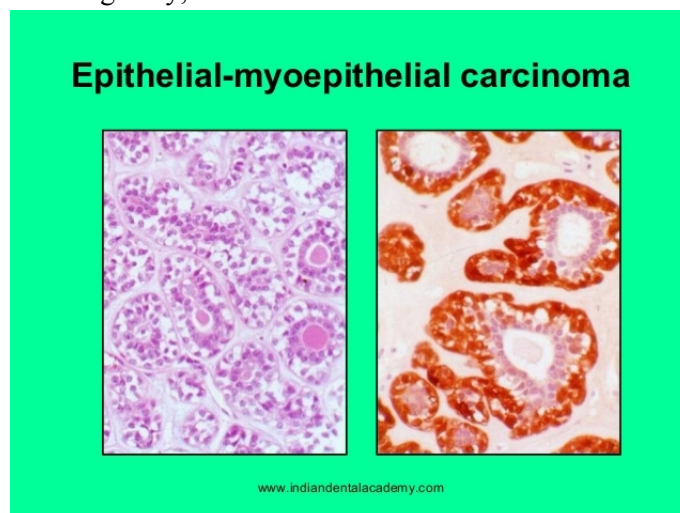
Epithelial- myoepithelial carcinoma (EMCa) is one of very rare carcinomas, firstly described tumor of salivary gland was by Donath ET in 1972. Approximately 1% of all salivary gland tumors is included in EMC. This tumor is a very low grade malignant tumor and the most common occurring sites in the body of this tumor are salivary gland and parotid gland but some uncommon location sites involve breast, nose, lung, trachea, bronchus and some more. Rare cases of malignant tumor of soft tissue were too presented in some cases. As the early symptoms noticed in the right forearm as a painless deep soft tissue mass and then later it raised without any remarkable symptoms like pain or distress. The one of the fatal salivary gland tumor, arose in a pleomorphic adenoma of the parotid gland. Firstly, the tumor was a pleomorphic adenoma with the epithelial and myoepithelial layers in the body. Later on, the tumor then occur again and may characterized by attacking on the nearby mandible. While examining histologically of this second recurrence it'll showing a harmful spindle cell neoplasm with different infiltrative growth pattern and a mitotic rate.

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INTRODUCTION:

Myoepitheliomas are those tumors that ascend from myoepithelial cells in the body and which shows lack ductal differentiation but both epithelial and smooth cell characteristics. EMCa, a low grade malignant salivary gland neoplasm which most frequently occurs in parotid gland and mainly arise in minor salivary glands too. In many cases of EM/Ca it is located in salivary glands, but in some cases, it occurs in some uncommon sites in the body such as breast, lachrymal gland, nose, trachea, bronchus and lung. This tumor shows different morphological structure and shows cytological divisions showing malignant changes in the body. This tumor act as an infiltrating cancer as it spread elsewhere in the layer of tissue in which it develops and grow into surrounding healthy tissue and spread to other parts of the body. Primary myoepithelial carcinoma salivary type tumor of the lung is rare. This tumor primarily occurs in the salivary glands, parotid glands and breast. Primarily this tumor of the lung arise from the sub mucosal layer in bronchial glands in the lower respiratory tract and shows similar morphologic features to those of their salivary glands counterparts.(1) EMCa is a very rare neoplasm, described by Donath et al in 1972. Histologically, EMCa is

categorized into two different cell arrangements; epithelial cells the inner layer and myoepithelial cells the outer layer. EMCa is always found clinically, and identified by the patient and observed by the professionals or the doctors.(2). The tumor in salivary glands is generally composed of different parts of two cells types: first inner layer of duct lining cells and outer layer of clear cells, and forms double layered duct like structures. In the year 1991, WHO accepted epithelial myoepithelial carcinoma (EMC) as a different entity and its subtype so it became part of the new classification system. Clinically, EMC mostly appears as a massive and slowly growing mass within the parotid gland. A more accurate definition of the disease can be achieved by histological immunohistochemically study. EMC is considered as a low grade large tumor that may mostly reappear locally after resection in about 20-50% of cases. This happens frequently because the capsule, which normally delimits the new formation may be incomplete. As far with this dedifferentiation of the cells of tumor has also been reported in some cases. This carcinomas are mainly treated by wide surgical elimination (excision), lymph node dissection and radiotherapy.(3)



Source: https://www.google.co.in/search?q=myoepithelial+carcinoma&rlz=1C1NHXL_enIN757IN757&source=Inms&tbn=isch&sa=X&ved=0ahUKEwjn2aLGnPLVAhVLMY8KHeJIAfUQ_AUICigB&biw=1366&bih=662#imgre=yZ2b9jaW2wi-9M:

EPIDEMIOLOGY:

Myoepithelial carcinoma, rare carcinomas with incidence reported of approximately 0.2% of all salivary gland tumors. However, some authors opposed that this tumor may not be a rare tumor as per lack of awareness and the diagnostic criteria may also be contribute to the small number of case which have been reported earlier of myoepithelial carcinoma.

Variability in sex distribution have shown of myoepithelial carcinoma for different series with generally similar age factor, though the reported series have not represented the old age group in all similar large series.

LOCATION:

Most of the cases of the myoepithelial carcinoma arise in the parotid gland which include (48% -75%), minor salivary glands and many reported sites include the palate, cheek, gum, nasal cavity, maxillary sinus, upper part of the pharynx, oral cavity, base of tongue and the submandibular gland.

TYPES OF CARCINOMA:

The Carcinoma can occur in many parts of the body. Some of the common types of carcinomas in the body include:

- Basal Cell Carcinoma (BCC): The cancerous cells develop in the basal cell layer of the skin or the lower part of the epidermis.
- Squamous Cell Carcinoma (SCC): The cancerous cells are developing from flat, squamous cells and which are the primary cell type making to the outermost layer of the skin, which is the epidermis.
- Renal Cell Carcinoma (RCC): The cancerous cells typically develop in the lining of very small tubes in the kidney, called tubules. Later time, these cells may grow into a mass and cause an obstruction.
- Ductal Carcinoma In Situ (DCIS): Cancerous cells are restricted within the lining of the milk ducts, and haven't spread through the duct walls into surrounding breast tissue.

- Ductal Carcinoma (IDC): Cancerous cells grow in the duct lining break through the wall of the duct and invade local breast tissue.

CLINICAL FEATURES:

Many of the patients with myoepithelial carcinoma observed with painless mass on their body which may be increased in size later. After observing some of these symptoms some authors have suggested that the tumor may remain small of a time period but later it starts growing rapidly.

Symptoms can vary depending on the site of tumor in the body. The average duration of symptoms before diagnosis varies from 3 months to 3 years.

CLINICAL PRESENTATION AND INTERVENTION:

Myoepitheliomas are the tumors which arise from myoepithelial cells completely. EMCa mostly occurs in salivary glands, in breast, skin, lung. Myoepithelial carcinoma is a large tumor which is equivalent to the Myoepitheliomas. While presenting immunohistochemically, myoepithelial carcinoma tumor cells shows not only the epithelial indications such as cytokeratin (recent nomenclature also called keratins), Epithelial Marker Antigen (EMA), but with these also shows indications like in calponin which is smooth muscle origin such as calponin (a calcium binding protein).

The criteria which is to be followed for myoepithelial differentiation of immunohistochemically is double positive for both. The prognosis of overall myoepithelial carcinoma is poor. Complete removal with tumor-free margin is always the preferred treatment, but radiotherapy and chemotherapy are always being the suggestive treatments as not always theses treatment works and can't be consider a curative treatment.

DIAGNOSIS:

Diagnosis of any patient with myoepithelial carcinoma begin with a detailed medical history and physical examination of an individual. While examining the neck and head tumor, the physical examination of the neck and head must

be complete. The entire head and neck must be checked for the further procedure as a diagnosis which may represent malignancies that could metastatic growth to parotid glands. The observation may include:

- Even benign tumors are usually firm but palpitation of the mass determines the degree of firmness but the malignancy denotes by the rock-hard mass.
- The external auditory canal must be visualized for tumor extension of the head and neck.

A diagnosis of EMC is made using the following tools:

- Complete evaluation of family's medical history, along with a thorough physical examination.
- Plain X-RAY of the part or the site of the infection.
- Ultra sound scan of the affected area of the body part.
- MRI scan of the part. Magnetic resonance imaging uses a magnetic field to create high quality pictures of that particular part of the body, such as tissues, muscles, nerves, and bones. Those high-quality pictures reveal the presence of the tumor.
- Fine needle aspiration biopsy (FNA) of the tumor

Diagnosis of EMC is based on predictable light microscopy and is confirmed by immunohistochemically and ultra-structural investigation. While histologically the tumor is characterized by well-defined ducts with two cell types first is an outer layer of cytoplasm surrounds an inner lining of eosinophilic cuboidal epithelial cells.(5)

FNAC (Fine Needle Aspiration Cytology):

- FNAC is a procedure used as the diagnosis of carcinomas which is used to investigate tumors or masses under the skin.
- FNAC is a type of biopsy procedure. The procedure of fine needle aspiration includes a

thin needle which is to be inserted into an area of abnormal appearing tissue or body fluid.

- The sample is to be collected during fine needle aspiration which can be helpful in ruling out the condition and the stage or type of the tumor or cancer.
- A fine needle aspiration procedure is most often done on swellings or lumps located just under the skin.

A lump may be felt during a doctor's examination. Or it may be discovered on an imaging test such as:

- CT scan
- Mammogram (the process of using low-energy X-rays)
- Ultrasound

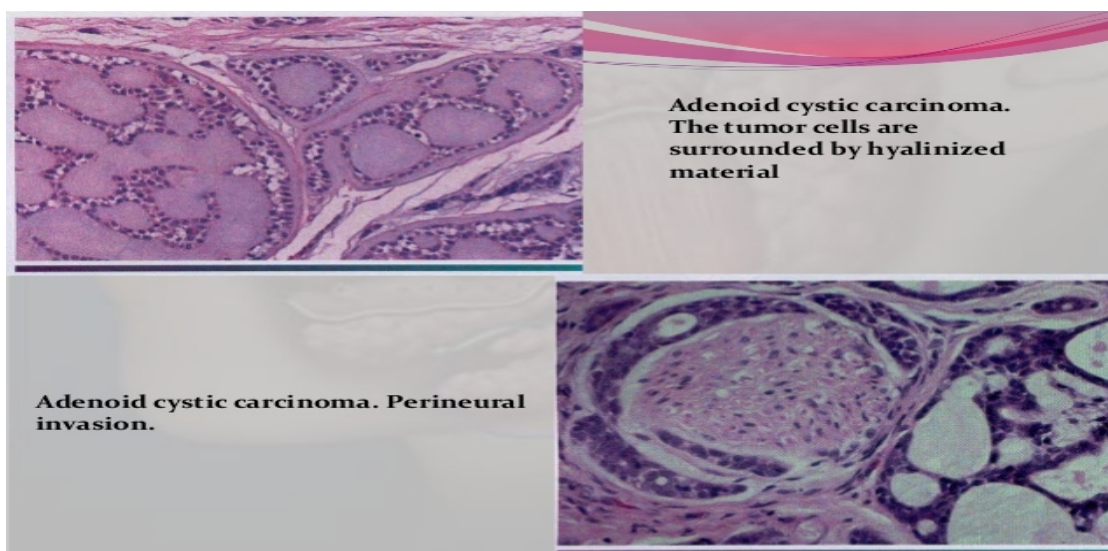
Doctors may recommend FNA for areas such as:

- Cysts (body fluid lumps)
- Nodules or masses (solid lumps)
- Enlarged lymph nodes

Without biopsy, it's usually difficult for a doctor or a professional to confirm what these abnormal areas contain or it may be a threat to the individual health or not. The other way of diagnosing the tumor is imaging test. Imaging test may also discover abnormal spots deeper inside the body while processing.

IMAGING STUDIES:

Imaging studies as the diagnostic procedure may be helpful in staging and for surgical planning. Medical ultrasound which is called sonography. Sonography may be very useful sometimes, as benign lesions are of lower density and have smaller caliber blood vessels. Computer tomography (CT) scanning and magnetic resonance imaging scanning can be valuable for evaluation of parotid malignancies. Ct scan provides better detail of the surrounding tissues of the site of the tumor, whereas MRI demonstrates the mass in greater contrast than a CT scan.



Source: <https://image.slidesharecdn.com/salivaryglandtumors23-5-2016160521060753/95/salivary-gland-tumors-23-52016-21-638.jpg?cb=1463810921>

TREATMENT:

To eliminate the main tumor from the body from the specific body part with the proper surgery by the professionals comes in the beginning of the treatment for myoepithelial carcinoma. Apart from the surgery radiation therapies can be used to reduce the chance that the tumor could return in the same location. While treating EMCa there are limited evidence about the usefulness of chemotherapy as there are no standard treatment guidelines.

Treatment of epithelial myoepithelial carcinoma surgery is always being the first

choice, which include the complete resection or removal of the part of the body by the surgery. After the surgical procedure, both light microscope assessment by routine H&E (Hematoxylin and Eosin) staining and immunohistochemically analysis by using specific antibodies were also carried out to confirm the pathological diagnosis of myoepithelial carcinoma. Afterwards focal external beam radiation therapy is to be performed on the patients in the target regions of the tumor cite of the body part.(4)

There are some drugs which can be used as a medicinal drug in the chemotherapy treatment or any other drug involvement treatment of myoepithelial carcinoma. Here some are the following examples:

S.No.	Brand Name	Generic Name	Mechanism of Action
1	Ametycine Mito-medac Mitogel Mitosol Mutamycin	Mitomycin	DNA inhibitor
2	Adriacin Adriamycin Adriblastina Doxophos Resmycin rubex	Doxorubicin hydrochloride	DNA inhibitor
3	Altoprev Artein Liposcler Lovarlord Mevacor Mevinacor Nergadn Rextat Altacor	Lovastatin	HMG Co Reductase Inhibitor
4	Etopol Etosid lastet Vepesid Vepeside	Etoposide	DNA Intercalator Topoisomerase II Alpha Inhibitor

DISCUSSION:

Myoepitheliomas are those tumors that arise from myoepithelial cells in the body and which shows both epithelial and flat cell features but absence of ductal variation. EMCa, mostly occur in parotid gland which is a low grade malignant salivary gland neoplasm and mainly rise in minor salivary glands too. In many reports of EMCa it is located in salivary glands, but in some cases, it occurs in some uncommon sites in the body such as lung, bronchus, trachea, nose, lachrymal gland and breast. This tumor shows different morphological structure and shows cytological divisions showing malignant changes in the body. This tumor act as an infiltrating cancer as it supper out there in the layer of tissue in which it develops and propagate into adjacent healthy tissue and spread to other body parts (7).

In the cases of parotid EMC all the descriptions mainly discovered gradually defined counters and shows some development of the nodules, signifying a malignant tumor. The procedure concluded that the radiological estimation of the injury/lesion only will be insufficient and misrepresentative in the analysis of salivary gland tumors, which is applicable exclusively in the reports of low grade malignant tumor such as EMC. So in the cases like this the FNAC of that particular injury/lesion will also be recommendable(8) Many of the patients with myoepithelial carcinoma observed with painless mass on their body which may be increased in size later. After observing some of these symptoms few biographers have proposed that the tumor might persist small of a time era but later it starts rising promptly (9). Primary myoepithelial carcinoma salivary type tumor of the lung is infrequent. This tumor predominantly occurs in the salivary glands, parotid glands and breast. Primarily this tumor of the lung ascends from the sub mucosal layer in bronchial glands in the lower respiratory tract and shows similar morphologic structures to those of their salivary glands complements(10)

CONCLUSION:

Myoepithelial carcinoma mainly rises from the salivary glands, the Parotid or the breast but

they might rise in soft tissue infrequently and most frequently arise in lower and upper limbs which occurs correspondingly in males and females. The majorly myoepithelial carcinoma of the lung is extremely exceptional only few patients are being described yearly. Myoepitheliomas are those tumors which arise from myoepithelial cells primarily or entirely. Myoepithelial cells are generally situated in the basal lamina of ducts of salivary glands, breast and sweat gland of the skin and between the epithelial cells. It must have been detected in some cases that some morphologically low positioned myoepithelial carcinomas destructively. Thus in the non-appearance of freely malignant cytomorphology, an aggressive development arrangement in the particular utmost beneficial condition for forming malignancy in salivary EMCa. Myoepithelial carcinoma of the lung is a very infrequent tumor, it shows merely myoepithelial differentiation lacking the formation of ducts on the site of tumor. Mostly while examining it usually shows certainty with Myoepitheliomas, myoepithelial carcinoma and epithelial myoepithelial carcinoma of the lung. As the epithelial myoepithelial carcinoma and myoepithelial carcinoma of the salivary gland are renewed by the occurrence or nonappearance of ductal cells, their respiratory complements should be distinguished also. EMCa constitute very small percentage of all salivary gland tumors. At present the recognized analytical criteria for myoepithelial carcinoma are limited myoepithelial differentiation morphologically and immunohistochemically and the surgery of removing the malignant tumor part or the tissue. Calculation of offensive is generally challenging in tumors which arise with in minor salivary glands which involves suitable selection of the tumor multitude tissue interfacing and which might not be involved in the biopsy specimen. Most of the studies have established that 75% of the myoepithelial carcinoma ascend in the major salivary glands and partial rise in ancestor injuries but somehow the consequences in existing study is

fairly different from the earlier study as minor salivary gland participation was more than the major salivary glands participation. At times it turns out to be very challenging to recognize the established compassionate constituent surrounded by the tumor, especially when the tumor is low grade tumor. The pre-existing compassionate tumor of myoepithelial carcinoma can be assumed if nearby is an elongated history of compassionate parotid tumor which may have the antiquity of prompt development or various reappearances in a pre-existing adenoma through or lacking lymph node metastasis

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