

CLEAR CELL MYOEPIHELIAL CARCINOMA – A CASE REPORT

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ABSTRACT

Myoepithelial carcinoma is a very rare neoplasm that comprise only 2% of all salivary gland carcinoma. The variable morphologic appearance of myoepithelial carcinoma leads to a wide differential diagnosis, including primary salivary gland tumors and metastatic tumors. We report a case of clear cell variant of myoepithelial carcinoma which is extremely rare and only very few cases are reported. A male patient reported with swelling on his left neck below the lower border of mandible, with slight difficulty in swallowing and there was deviation of tongue. Surgical excision was done and on histological examination and on immunohistochemistry reported as myoepithelial carcinoma.

KEYWORDS : Myoepithelial carcinoma, Salivary glands, Clear cells.

INTRODUCTION:

Salivary gland tumors pose a great challenges in diagnosis because of their varied morphology and unpredictable behavior. Myoepithelial carcinoma account for 0.4 to 0.6% of all salivary gland neoplasms and was introduced by Stromeyer et al in 1975.¹ Myoepithelial carcinoma has a diagnostic challenge as it mimics benign neoplasms clinically and many clear cell lesions histologically thus mandating the use of immunohistochemical markers such as S 100, alpha Smooth muscle actin, Cytokeratins, EMA, Calponin and vimentin to confirm the diagnosis.²

Case Report

A 70 year old male patient reported with swelling on his left neck since 2 months that was present below the lower border of mandible, slow growing and measured about 10cm X 5cm. Swelling was soft in consistency and was not fixed to underlying structures. Tongue was deviated towards left and had difficulty in swallowing. (Figure 1)



Figure 1 Clinical picture showing swelling in his left neck

CT scan revealed left enlarged submandibular gland with multinodular cystic lesion showing hyperdensity. (Figure 2)

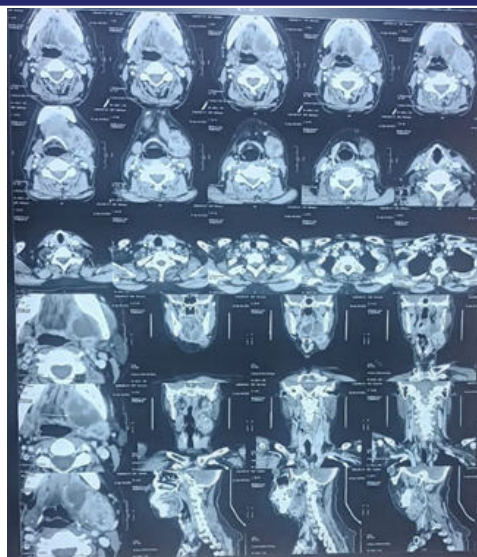


Figure 2 CT scan - left enlarged submandibular gland with multinodular cystic lesion

Lesion was excised and sent for histopathology analysis. Department of Oral Pathology received a single bit of rectangular soft tissue specimen, brownish in color, firm in consistency with regular borders and surface that measured about 8.0cm X 6.0cm X 4.0cm in its greatest dimension.

Histopathologic examination revealed well encapsulated tumor. Tumor islands arranged in solid nests, cords along with ductal and cribriform pattern, periphery of tumor islands showed columnar cells while cells in the centre were pleomorphic having scanty to clear cytoplasm with hyperchromatic nucleus and occasional mitosis. Tumor islands were surrounded by hyalinising stroma. Areas of necrosis and few calcifications were evident. Fibrous capsule also showed normal acini along with few tumor islands, blood vessel and nerves. Perivascular presence of tumor cells was observed. With the above histopathological features myoepithelial carcinoma, hyalinising clear cell carcinoma of salivary gland, and metastatic tumor from other organ were considered. (Figure 3)

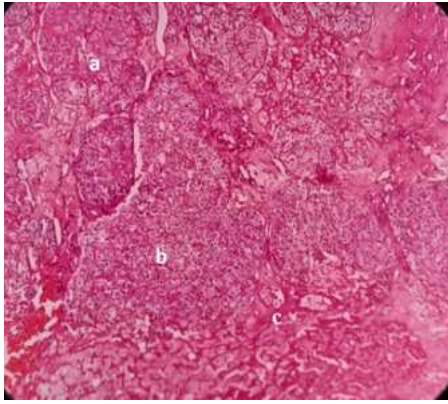


Figure 3: Histopathologic picture-

- a. Tumor islands arranged in solid nests and cords
- b. Clear cells
- c. Hyalinizing stroma

Considering myoepithelial carcinoma being the most common tumor having clear cell differentiation among the other clear cell tumors, blocks and slides were referred for ck 5/6 and calponin immunohistochemical test. With cytokeratin 5 & 6 positivity and calponin positivity finally the case was reported as Myoepithelial carcinoma (Figure 4)

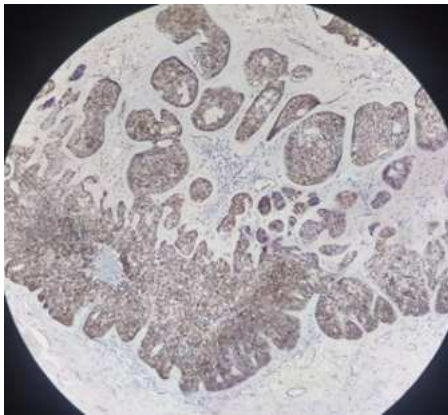


Figure 4: Immunohistochemistry image- tumor cells showing calponin positivity

DISCUSSION:

Myoepithelial carcinoma (MC) is defined as a malignant neoplasm of salivary gland with exclusively myoepithelial differentiation. It is an extremely rare salivary gland tumor which comprises of less than 1% of all salivary gland tumors.

According to World Health Organization (WHO) classification 1991, Myoepithelial carcinoma has been included as a distinct clinicopathological entity.³

It occurs in both major and minor salivary glands. The most common sites are the parotid gland, submandibular gland, and hard palate. Rarely they are also reported in buccal mucosa, alveolar ridge, retromolar trigone, lips, lung and sinonasal tract.⁴

Myoepithelial carcinoma is histologically a challenging entity.

These are a diverse group of tumor cell types seen in this neoplasm in which most cases display multiple cell types such as epithelioid, clear cell, plasmacytoid, spindled, and mixed.^{4,5}

Myoepithelial carcinoma include a rare subtype of clear cell tumors.⁴

Myoepithelial cells often display a uniform monotonous cytology, reflecting clonal growth of malignant tumor in which the tumor may exhibit solid, trabecular or reticular patterns with myxoid stroma or hyalinized stroma.

Regardless of cell morphology, the tumor displays multinodular architecture with a lobulated uniform cell proliferation with expansile invasive borders. The cellular arrangement of hypercellular peripheral rim with hypocellular center is the most commonly seen feature. Hypocellular center may show necrosis, myxoid stroma or hyalinized stroma⁵

Di Palma and Guzzo reported that this tumor is a low grade tumor if it arises from the pleomorphic adenoma or benign myoepitheliomas, whereas denovo tumors are seen to be more aggressive and have a higher risk of distant metastasis.^{6,7}

The differential diagnosis of clear cell myoepithelial carcinoma includes all other tumors that contain clear cells such as hyalinizing clear cell carcinoma, epithelial myoepithelial carcinoma, acinic cells carcinoma, mucoepidermoid carcinoma and metastatic renal cell carcinoma.⁶

Immunohistochemical criteria for confirmation of myoepithelial carcinoma are positivity for cytokeratin (basal cell cytokeratin preferentially or even pan cytokeratin). Tumor is positive for one or more of myoepithelial markers which include S100, calponin, p63, glial fibrillary acidic protein, and actins. These tumors consistently express cytokeratins, epithelial membrane antigen, vimentin, S100 protein and calponin⁷

Vimentin is reported to be positive in neoplastic myoepithelial cell whereas normal myoepithelial cells will not express vimentin.⁸

Few studies confirm that alpha smooth muscle actin, smooth muscle myosin heavy chains and calponin are specific myoepithelial markers for salivary gland tumors.⁷

The preferred treatment of choice is Surgery where initially, Radical surgery is suggested with sufficient normal tissue margins to prevent the recurrence and facial nerve is to be sacrificed. Preservation of the facial nerve to be tried only when parotid gland tumor is small and is situated far away from the nerve. Careful resection of lymph nodes surrounding parotid and submandibular gland should be done. When the tumor is large and when it occurs in parotid and submandibular gland, elective neck dissection is considered.

Radiotherapy is not sensitive for myoepithelial carcinoma. Postoperative chemotherapy may also be used to prevent blood stream metastasis.⁹

CONCLUSION:

As many tumors in the head and neck area shows clear cell differentiation, careful observation and differential diagnosis helps us in narrowing down with the diagnosis, also supplementing with the application of IHC marker can aid us in confirmation of the lesions.

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