Myoepithelial Carcinoma: A Rare Case Report with Review

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ABSTRACT
Myoepithelial carcinoma is a rare neoplasm of salivary glands that account for < 1% of all salivary gland tumors. It composed entirely of myoepithelial cells that exhibit a dual epithelial and smooth muscle phenotype. Major salivary glands mainly parotid gland is most common site of involvement. It also occurs in other head and neck region such as palate, nasopharynx, paranasal sinuses, nasal cavity and larynx. MECs most commonly seen in third to fifth decade of life, with equal sex predilection. They are characterized by distinct morphologic heterogeneity and an infiltrative growth pattern into adjacent tissues. Here, we report a case of a 50 year old female with myoepithelial carcinoma of palate.

INTRODUCTION
Myoepithelial carcinoma (MC), also known as malignant myoepithelioma, is a rare neoplasm of salivary gland that was first described in 1975 by Stromeyer et al. MC was defined as a solitary pathological diagnosis in 1991 by the World Health Organization (WHO), and its definition was updated in 2005.1,2 Myoepithelioma are tumors arising from myoepithelial cells lacking ductal differentiation which exhibit both epithelial and smooth muscle cell characteristics. Benign myoepithelial tumors were seen mostly in extremities and head-neck region, while malignant counterparts mostly occur in the salivary gland, parotid and breast tissues.3 MC is rare, representing about 0.4% to 0.6% of all salivary gland tumors and 1.2% to 1.5% of carcinomas. The average age of presentation is about 55 years (range 14-86 years), with equal sex predilection. MC arises predominantly from the major salivary glands, either de novo or as a carcinoma arising from a preexisting pleomorphic adenoma or myoepithelioma.2, 5, 6 Histologically, morphological heterogeneity is the hallmark in MC, and the majority of neoplasms display more than one cell type. Immunohistochemically, these tumors are mostly positive for vimentin, S-100 protein, cytokeratin and α-smooth muscle actin (α-SMA), and certain new myoepithelial markers have been reported to have high expression levels in MC, such as calponin (CALP), p27, p53, CAM and epithelial membrane antigen, amongst others.1,7 The currently accepted diagnostic criteria for myoepithelial carcinoma are exclusive myoepithelial differentiation (morphologic and immunohistochemical) and clear-cut tumor infiltration into adjacent salivary gland or other tissues.4 The mainstay of treatment for myoepithelial carcinoma is surgical, including wide excision with free margins, with or without nodal dissection.
Adjuvant radiotherapy and postoperative chemotherapy do not seem to improve their prognosis significantly. The role of radiation in the treatment of malignant myoepithelioma is still controversial.\(^1\)\(^-\)\(^4\)

**CASE REPORT**

A 50 year old female patient visited our hospital with chief complaint of swelling on maxillary left palatal aspect since 14 years. Initially it was small and gradually increased in size to present size. She was a known case of valvular heart disease and had undergone surgery before 5 year. On extraoral examination, no abnormality was detected [figure 1(a)]. On intraoral examination, single well defined unilateral dome shaped swelling of size 2*2 cm present on left palatal aspect extending anteroposteriorly from distal of 23 to distal of 27 and mediolaterally form midline of palate to palatal gingival margins with normal overlying mucosa. On palpation it was non tender, soft and fluctuant, compressible and attached to the underlying structures. No bleeding, pus discharge, displacement of adjacent teeth noted [figure 1(b)]. Electric pulp vitality test shows no teeth with delayed response. Provisional diagnosis of pleomorphic adenoma was considered and for confirmation various investigations were carried out. IOPA, Maxillary occlusal radiograph and Orthopantomograph showed soft tissue shadow of swelling superimposed over left maxillary posterior teeth with no evident bony changes present except crestal bone loss. No definitive etiologic factors have been identified but, p53 accumulated mutations have been reported.\(^1\)

However, the reported case was present in 40 year old female with involvement of palatal minor salivary gland was rare. Histologically, MC are classified into spindle, plasmacytoid, epithelioid, clear cell, and mixed cell subtypes, according to the predominant displacement of adjacent teeth, no root resorption was noted [figure 2(a-b), 3]. CT scan revealed presence of 25*18*18 mm sized mildly enhancing soft tissue lesion with internal calcific foci is noted on left side of palate. Lesion causes suspicious erosion of adjacent hard palate. Lesion abuts adjacent soft palate also. FNAC aspirate was bright red colored [figure 4] and showed smears of scant cellularity consists of occasional clusters of benign epithelial cells. On the basis of clinical findings & investigations provisional diagnosis of pleomorphic adenoma was considered. To confirm the diagnosis biopsy and immunohistochemistry was advised. H & E section shows predominantly spindle cells, clear cells with abundant hyaline type material with glandular component and calcification. Possibility of malignant salivary gland tumor was made. IHC was positive for AE-1, P-63, and vimentin and negative for CKit (CD 117), CD 10 [figure 5 (a)-(d)]. Final diagnosis of myoepithelial carcinoma of minor salivary gland of palate was considered. Patient was treated with wide local excision along with lower left partial maxillectomy under GA. On four months follow up patient was normal with no recurrence noted.

**DISCUSSION**

Myoepithelial carcinomas (or malignant myoepithelioma) are a rare group of tumors which are characterized by myoepithelial differentiation.\(^4\)\(^,\)\(^6\) MECs most commonly seen in third to fifth decade of life, with equal sex predilection. The more prevalent site is the major salivary glands. There is no
cell type (>75% of the cells); however, when 2 or more cell types predominate, the tumor is classified as mixed. Presented case showed predominantly spindle cells, clear cells with abundant hyaline type material with glandular component and calcification. The malignant nature of the lesion, distinguishing primarily on its destructive, infiltrative growth pattern. Mitotic figures and necrosis, which are not typically seen in myoepithelioma, are also helpful in supporting the diagnosis. 

Confirmation of myoepithelial differentiation was performed with epithelial and myogenic markers. Presented case showed positive AE-1, P-63, vimentin markers and negative C Kit (CD 117), CD 10 markers which was suggestive of the same. For myoepithelial carcinoma, complete excision with tumor-free margin remains the first choice of treatment, in spite of the possibilities of local recurrence and distant metastasis. 

Our case also underwent complete excision with tumor free margins with partial left maxillectomy. The clinical prognosis and biologic behavior of MECs are not well characterized. The overall prognosis of myoepithelial carcinoma is poor. Several studies reported aggressive clinical behaviors for myoepithelial carcinoma, and the average metastatic rate was 47% and the mortality rate was 29% after a mean of 32 months. Recurrence and metastasis are more common in children than in adult even with a negative excision margin. 

In the presented case on follow up for 4 months did not show any further complain or signs of recurrence suggestive of better prognosis.

CONCLUSION
The myoepithelial carcinoma of palate is a rare case in head and neck region. It requires sound knowledge of clinicopathological and immunohistochemical features for its accurate diagnosis & better prognosis.

(a) 
(b)

Figure 1 extraoral frontal profile shows no abnormality, intraoral view shows single well define palatal swelling on left side.
Figure 2 IOPA of 24, 25, 26, 27 region shows superimposed soft tissue shadow on molar teeth, maxillary occlusal shows no bony architectural changes

Figure 3 orthopantomogram shows superimposed soft tissue shadow with crestal bone loss interproximal to 25 & 26
Figure 4 FNAC aspiration shows bright red colored aspiration

(a) vimentin positive

(b) p63 positive
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