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Myoepithelial Carcinoma of Intra-Oral Minor Salivary Glands Mimicking a Sarcoma - A Rare Case

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ABSTRACT

Background: Myoepithelial tumors are rare and account for less than 1% of all salivary gland tumors, of which majority are benign and only a few cases of malignant myoepitheliomas have been reported till date. A malignant myoepithelioma may arise *de novo* or develop within a preexisting pleomorphic adenoma or benign myoepithelioma.
Case report: We report a case of 60 year old male with complaints of pain and swelling over right side of mouth in upper jaw region. Intra oral examination revealed, a soft pedunculated lesion extending from the hard palate. Microscopically, the lesion was composed of fascicles of malignant spindle cells, mimicking a sarcoma. On IHC, tumor cells showed positivity for SMA, p63 and S-100 and final diagnosis of Myoepithelial carcinoma was reached.
Conclusion: The diagnosis of myoepithelial carcinoma due to its rarity and morphological heterogeneity can be challenging. Therefore a high index of suspicion and confirmation via immunohistochemical markers is essential.

Keywords: Myoepithelial carcinoma, minor salivary gland tumor, oral malignancy.

INTRODUCTION

Myoepithelial tumors are rare and account for less than 1% of all salivary gland tumors.[(1,2)] Benign myoepithelial tumors are mostly seen in extremities and head-neck region, while its malignant counterpart i.e. myoepithelial carcinoma occur in major salivary glands, most commonly parotid. Very few tumors have been reported to arise from minor salivary glands. Other sites of involvement are skin, nasal cavity, nasopharynx, larynx and lung and breast.[(2,3)]

Myoepithelial tumors are predominantly benign and only a few cases of malignant myoepitheliomas have been reported till date. They have been underrecognized in the past, but with the introduction of myoepithelial tumors as a separate entity in the World Health Organization's histological classification of salivary gland tumors 1991, its recognition has increased.[(4)] According to Armed Forces of Pathology (AFIP) database of salivary gland tumors, it accounts for less than 1% of malignant epithelial neoplasms.[(5)] 69 cases

of intraoral minor salivary gland myoepithelial carcinoma have been reported with only 8 cases involving the buccal mucosa and palate.[2]

Myoepithelial carcinoma may arise *de novo* or develop within a preexisting pleomorphic adenoma or benign myoepithelioma. The average age of presentation is about 55 years with no sex predilection. Usual presentation is as slow growing asymptomatic mass.

This article reports a very rare case of oral myoepithelial carcinoma arising from palate in an elderly male patient.

CASE REPORT

A 60-year old male presented to the oral and maxillofacial surgery clinic with the complaint of painful swelling over the upper right side of the roof of the mouth since past one month. The patient was a chronic tobacco chewer for past 40 years. On extra-oral examination, facial asymmetry was seen and mouth opening was reduced. Intraoral examination revealed, a soft pedunculated lesion measuring about 4 x3 cm over right buccal mucosa extending from hard palate. [Figure 1a] The mucosa overlying the swelling was inflamed and readily bled on manipulation. The maxillary right molar teeth were mobile. An orthopantomogram was advised which revealed no significant osseous changes. [Figure 1b]

Based on the clinical and radiographic findings provisional diagnosis of pyogenic granuloma was made. And the differential diagnosis included of peripheral giant cell granuloma, peripheral ossifying fibroma, sarcoma and lymphoma.

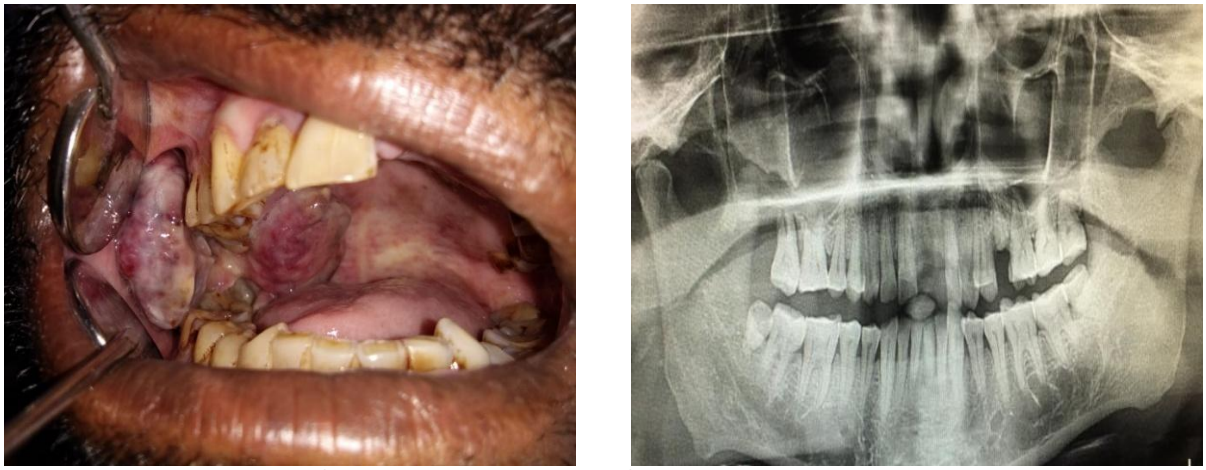

Figure 1: Photographs showing soft pedunculated lesion extending from buccal mucosa to the hard palate (a), orthopantomogram showing no significant bone involvement (b).

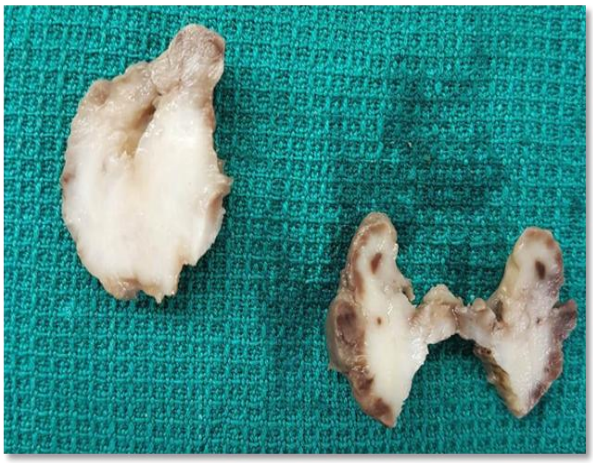

Figure2: Gross appearance of the lesion showing gray-white to gray-brown, firm tissue. The cut surface of lesion appeared fleshy.

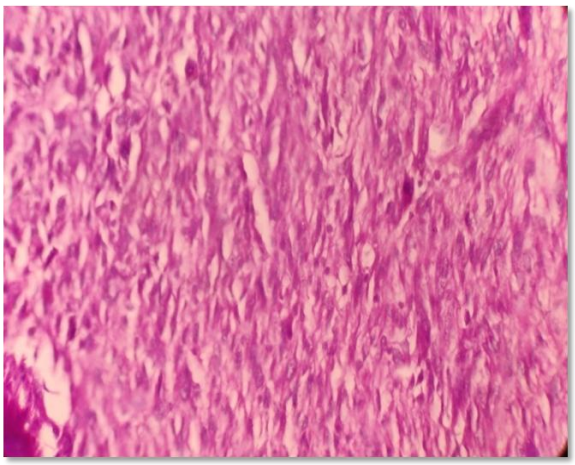

Figure 3: Photomicrograph showing spindle shaped neoplastic cells in fasciles with vesicular to hyerchromatic pleomorphic nuclei. (40X, H&E).

Excision biopsy of the lesion was done and the excised specimen was submitted for histopathological examination in our department.

The gross specimen was received in three pieces, which were irregular, grey white to grey brown, and soft to firm. The largest tissue piece measured 3 x 2.5 x 2.3 cm, which was partly covered with mucosa. Cut surface of all the three tissue pieces was grey white, fleshy and firm. [Figure 2]

Microscopically, the hematoxylin and eosin stained sections the tumor was composed of neoplastic spindle shaped cells arranged in bundles, fascicles and in haphazard manner. These cells showed highly pleomorphic enlarged vesicular to hyperchromatic nuclei with prominent nucleoli and moderate to scanty cytoplasm. At places, stroma appeared myxoid. Superficial part of the tumor showed ulceration with acute on chronic inflammation. The mitotic rate was 12-14/ 10 HPF with many atypical mitotic figures. [Figure3]

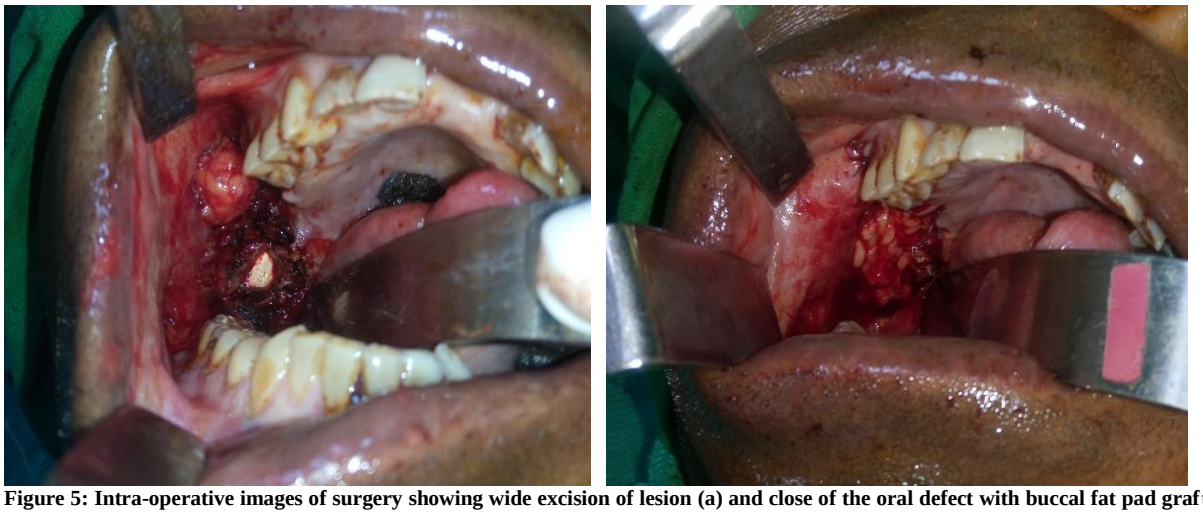

Figure 5: Intra-operative images of surgery showing wide excision of lesion (a) and close of the oral defect with buccal fat pad graft (b).

Based on histological findings diagnosis of High grade malignant tumor with features favoring Pleomorphic Sarcoma was made.

On Immunohistochemistry, tumor cells showed positivity for SMA, P63, S 100 and immunonegativity for cytokeratin, EMA, CD31, CD34. The above findings were suggestive of sacromatoid carcinoma, of myoepithelial origin. Based on the histopathological findings and immunohistochemistry study final diagnosis of myoepithelial carcinoma was made.

Patient was kept on periodic follow-up. Subsequently after 5 months, patient came with recurrence of a large tumor involving Right upper alveolus, buccal mucosa and hard palate.

Computed Tomography showed a heterogeneously enhancing soft tissue density mass lesion involving soft palate and extending to the hard palate. Wide excision of the lesion along with right side neck node dissection was carried out. [Figure 5]

Grossly, the excised specimen showed a tumor which was gray-white to gray-brown, exophytic mass measuring 4.2 x 4.2 x 1.7 cm, involving the right lower gingiva, gingivo-buccal sulcus and buccal mucosa. Also, a large polypoidal tumor mass measuring 7.5 x 5 x 3.3 cm was received from hard palate along with attached the bony plate. [Figure 6]

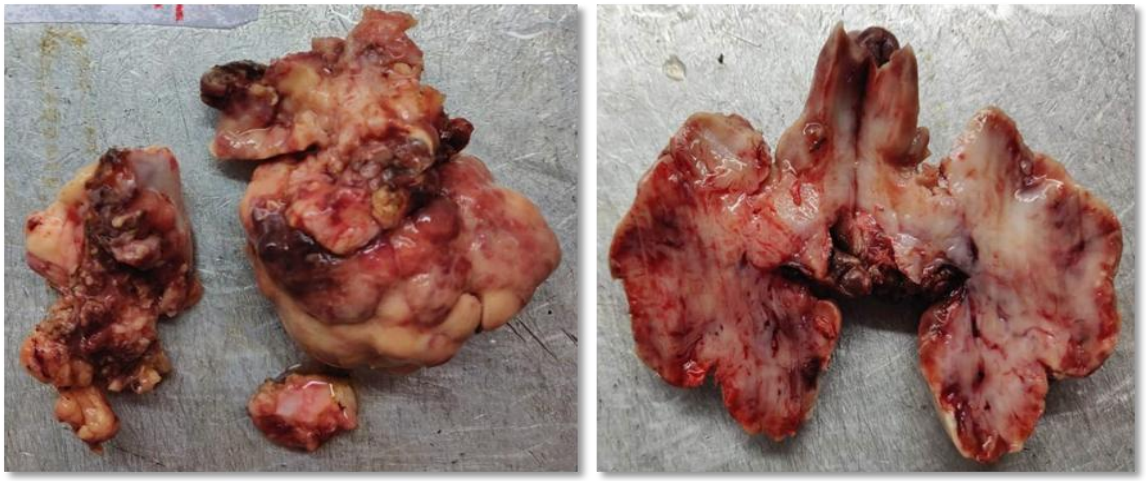

Figure 6: Exophytic gray white tumor involving right lower gingiva, gingivo-buccal sulcus and buccal mucosa. Cut surface is gray white to gray brown and fleshy.

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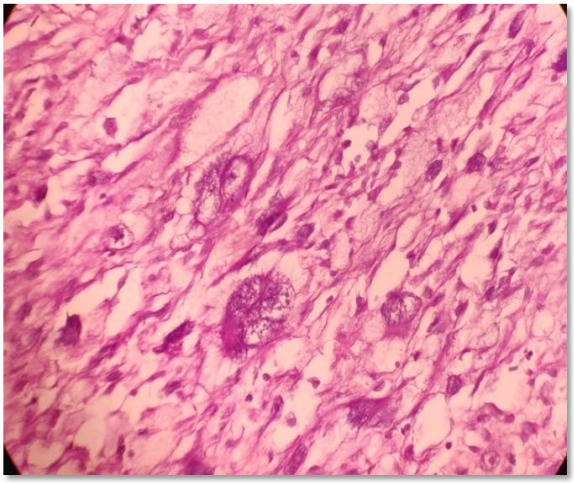

Figure 7: Pleomorphic neoplastic cells with bizarre nuclei & many tumor giant cells (100X H&E).

With recurrence, the tumor appeared more pleomorphic, bizarre with many tumor giant cells. The tumor was seen invading the underlying muscle, minor salivary glands and underlying maxillary bone. No lymph node involvement was seen microscopically. [Figure7]

The patient was subsequently kept on periodic recall and is tumor free at 12 months follow-up.

DISCUSSION

Myoepithelial carcinoma is a malignant salivary gland neoplasm in which tumor cells demonstrate cytologic differentiation toward myoepithelial cells and lack ductal or acinar differentiation. These tumor cells show wide range of morphological variation, comprising of spindle, plasmacytoid (hyaline), epithelioid, stellate and clear cell subtypes. The epithelioid cell type is the most common type. Combinations of these cell types may be present within the same tumor. These tumor cells are present in myxoid or hyalinized matrix.[1] Stroma is generally absent in tumors with spindle cell pattern.[5]

Immunohistochemically, markers of myoepithelial differentiation such as vimentin, S-100 protein, calponin, glial fibrillary acidic protein and smooth muscle actin, and cytokeratins, are diagnostic. P63 is a useful marker of myoepithelial cells in salivary gland neoplasms.[1,6,7] While Prasad et. al. in their study confirm Smooth Muscle Actin (SMA) and calponin to be specific myoepithelial markers in salivary gland tumors. Although not specific, S-100 protein is often a sensitive marker for neoplastic myoepithelium. Our case exhibited immunopositivity for P63, SMA and S100.

The currently accepted diagnostic criteria for myoepithelial carcinoma are predominant myoepithelial differentiation (morphological and immunohistochemical) and distinct malignant features(nuclear atypia, tumor necrosis, tumor infiltration, more than seven mitoses per 10 HPFs or a Ki-67 labeling index of more than 10%).[8,9] Similarly, the present case showed high mitotic rate and myoepithelial differentiation.

The differential diagnosis of a malignant myoepithelioma depends on the predominant cell type. With spindle cell morphological features, the differentials include sacromatoid squamous carcinoma, spindle cell melanoma, schwannoma and malignant peripheral nerve sheath tumor.

Immunohistochemical staining is helpful in differentiating these lesions.

Distinction between a malignant myoepithelioma and spindle cell or poorly differentiated squamous cell carcinoma can be difficult, especially in oral lesions. In our case Pleomorphic Sarcoma was a differential diagnosis. However, with Immunohistochemistry a definitive diagnosis of myoepithelial carcinoma was made.

Prognosis of myoepithelial carcinoma is variable. There are no definite histological features that correlate clearly with their clinical and biological behavior.

Therefore, wide surgical excision is the most appropriate treatment modality available.

Therapeutic neck dissection is indicated when there are clinically or radiologically apparent metastases in the cervical lymph nodes.

CONCLUSION

The diagnosis of myoepithelial carcinoma due to its rarity and morphological heterogeneity can be challenging. Therefore a high index of suspicion and confirmation with a number of immunohistochemical markers, especially in tumors with pure spindle-cell or plasmacytoid morphologic characteristics with rhabdoid differentiation are essential.

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