Myoepithelial carcinoma of buccal mucosa: A rare tumor

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Abstract

Myoepithelial carcinoma is a rare neoplasm of salivary glands that account for < 1% of all salivary gland tumors. The most common sites of involvement are major salivary glands mainly parotid gland. Intraorally, it can arise from minor salivary glands; palate is the most common site of occurrence. It also occurs in nasopharynx, paranasal sinuses, nasal cavity and larynx in head and neck region. Myoepithelial tumors were first described in 1943. Their malignant variant, myoepithelial carcinoma, was first reported by Stromeyer et al., in 1975, characterized by distinct morphologic heterogeneity and an infiltrative growth pattern into adjacent tissues. Here, we report a rare case of a 55-year-old female with myoepithelial carcinoma of buccal mucosa. It was also rare because of unusual location of tumor. Our patient was treated with wide local resection and remained free of disease for 15 months.

Keywords: Buccal mucosa, minor salivary gland, myoepithelial carcinoma

Introduction

Myoepithelial carcinoma is the malignant counterpart of myoepithelioma, tumor cells exclusively manifest myoepithelial differentiation.\(^1,2\) According to the World Health Organization (WHO) classification, myoepithelial carcinoma is referred to those lesions which are composed almost exclusively of tumor cells with myoepithelial differentiation. It is a rare neoplasm that has not been included in most reported surveys of salivary gland neoplasia. This category of tumor was added to the classification of salivary gland tumor as a distinct clinicopathological entity by the WHO in 1991.\(^3\) In the Armed Forces Institute of Pathology database of salivary gland neoplasms, it constitutes <1% of malignant epithelial neoplasms.\(^4\) In reported series, the majority of myoepithelial carcinomas occur in the parotid gland,\(^5\) with fewer tumors in the submandibular and minor salivary glands. Among the reported cases, the parotid gland is involved in about two-thirds of the cases. Intraorally only 69 cases affecting minor salivary glands have been reported in the English literature.\(^6\) Similar to benign myoepitheliomas, the palate is the most common intraoral site of involvement.\(^7,8\) Men and women are almost equally affected. The average age of patient at presentation is over 50 years and the tumor usually appears as an asymptomatic mass that slowly increases in size.\(^9\) Other than salivary glands, this tumor also occurs in the breast, skin, nasal cavity, nasopharynx, larynx and lung.\(^10-12\) We herein report a rare case of myoepithelial carcinoma of a 55-year-old female at buccal mucosa, a rare location. To the best of our knowledge, there are only eight cases of myoepithelial carcinoma of buccal mucosa are reported in the literature.

Case Report

A 55-year-old female patient presented in the Department of Oral and Maxillofacial Surgery, Punjab Government Dental College and Hospital, Amritsar complaining of swelling in the back region of right cheek. The history dates back to 4 months when patient felt a small, asymptomatic swelling in back region of right cheek which was spontaneous with no history of trauma or functional impairment, which gradually increased in size. Patient got extracted her two right upper posterior teeth from some private practitioner and was on antibiotic and anti-inflammatory medication, but got no relief. Therefore, patient was referred in our institution for diagnosis and management.

On extraoral examination, there was no obvious facial asymmetry or swelling evident, no cervical lymph node was palpable. Intraoral examination revealed an outgrowth involving buccal mucosa of right posterior region extending up to the right maxillary tuberosity region. Outgrowth was approximately 3.0 ×  1.5 cm in size, cylindrical in shape, sessile, pink in color, smooth in surface with few bosselated areas secondary to occlusal trauma [Figure 1a]. On palpation, temperature of growth was not raised, firm in consistency, nontender, ill-defined margins with fixation to underlying structure. No sign of fluctuation was evident. Extraoral examination revealed no obvious lymph nodes or other metastatic spread.
radiography showed no bony involvement. Magnetic resonance imaging of face and neck was performed, showed well-defined mass measured 2.2 × 1.3 × 1.0 cm in size on right side of buccal mucosa lying anterior to the masseter muscle and lateral to the buccinator muscle [Figure 1b].

Patient was advised fine-needle aspiration cytology (FNAC) of the lesion. FNAC report was suggestive of pleomorphic adenoma with epithelial predominance. No mitotic activity was seen, suggested for histopathological examination to further evaluate. Basal cell adenoma, solid variant of adenoid cystic or mucoepidermoid tumor were also kept in differential diagnosis.

Based on the clinical presentation and FNAC report diagnosis of pleomorphic adenoma of right buccal mucosa was made with a differential diagnosis of adenoma, solid variant of adenoid cystic and mucoepidermoid tumor. The wide excision of the lesion was performed under local anesthesia and wound was covered with a biodegradable collagen membrane dressing [Figure 1c]. The specimen [Figure 1d] was fixed in formalin and sent for histopathological examination. Microscopic examination showed a malignant tumor mostly composed of cells arranged in nests and lobules, separated by thin fibrovascular septae in a background of fibromyxoid stroma. The tumor cells had clear to pale eosinophilic cytoplasm, pleomorphic nuclei with prominent nucleoli and brisk mitoses [Figure 2a-c]. Hyaline basal laminar material was also seen at places. Occasional squamous islands and comedo-type necrosis were also noted. The tumor infiltrated into adjacent muscle. Immunohistochemical reactions were carried out and positive reactions obtained for pan-cytokeratin [Figure 3a], vimentin [Figure 3b], p63 [Figure 3c] and S-100 [Figure 3d]. The Ki-67 [Figure 3e] index was high (>10%). On the basis of histopathological and immunohistochemical results it was diagnosed as myoepithelial carcinoma of buccal mucosa.

As the wide local resection of the lesion had already been performed, no further treatment was done except a chest radiograph, to rule out any distant metastasis, which was reported as normal. Patient was kept on regular follow-up. Patient remained free of disease for 15 months and still on follow-up [Figure 4].

**Discussion**

Myoepithelial cells are ectodermally derived contractile cells, routinely identified in many normal tissues with a secretory function such as major and minor salivary glands, lacrimal glands, sweat glands, breasts, and the prostate. These cells are one of the most frequent components of salivary gland tumors. Salivary gland neoplasms that frequently contain myoepithelial cells are pleomorphic adenoma, adenoid cystic carcinoma and epithelial-myoeplithelial carcinoma of intercalated duct origin. Neoplasms composed exclusively of myoepithelial cells are uncommon accounting for <1% of all salivary gland tumors. Most of these tumors are located in the parotid gland, while others occur in the sub-mandibular gland or in the accessory glands of the oral cavity (hard and soft palate, lip, cheek, tongue, floor of the mouth, gingiva, retromolar area). They sometimes arise from the glands of the respiratory tract (nasal cavity, nasopharynx, larynx, and lung).

Many of the reported tumors have been described as developing in pleomorphic adenomas or myoepitheliomas i.e., myoepithelial carcinoma ex pleomorphic adenoma. Patients are generally over 50 years of age, with both sexes being equally affected. Sheldon was the first to identify myoepithelial salivary gland tumor as a distinct neoplastic entity. Myoepithelial carcinoma was first reported by Stromeyer et al., in 1975. Most of the 100 reported cases have been described within the last decade. The 27 and 25 cases reported by Yu et al., and Saveria et al., respectively are the largest series reported in English literature. Hornick and Fletcher have reported 101 myoepithelial tumors of soft-tissue that were not associated with salivary gland tissue. Tortoledo et al., have reported 37 carcinoma ex pleomorphic adenoma in their study, out of which only three cases were myoepithelial carcinoma. Young et al., have reported a myoepithelial carcinoma of the cheek in a patient with familial adenomatous polyposis. Kane and Bagwan presented a retrograde study of 51 cases of myoepithelial carcinoma of the salivary glands in a tertiary cancer center. Out of 51, 36 cases showed minor salivary glands involvement and only 15 cases affected major salivary glands.

Clinically, most myoepithelial carcinomas are painless masses. Tenderness, pain, hoarseness, dysphagia, and weight loss are experienced by a few few patients. Some patients with myoepithelial carcinoma ex pleomorphic adenoma have reported a sudden rapid increase in size of mass that had been present for many years. It may be 2-20 cm in largest dimension, nonencapsulated, nodular and soft to firm. Microscopically the neoplastic cell population in myoepithelial carcinoma exclusively manifests myoepithelial differentiation. However, in myoepithelial carcinomas that develop in pleomorphic adenomas (carcinomas ex pleomorphic adenoma), the pleomorphic adenoma portion of the neoplasm manifests ductal differentiation. Squamous differentiation is one of the manifestations of neoplastic myoepithelial cells and is acceptable as a minor component in myoepithelial carcinomas. The cellular configurations in myoepithelial carcinoma are the same as those in benign myoepithelioma such as spindle, plasmacytoid, epithelioid, clear and mixed cells. The epithelioid cell type is the most common and purely clear cell myoepithelial carcinomas are rare and hard to distinguish from other types of clear cell neoplasms.

The cellular density varies from tumor to tumor and within individual tumors. Neoplastic myoepithelial cells are capable...
of producing prodigious amounts of basal lamina in some tumors, which appears as eosinophilic hyaline material. Both nodular and diffuse patterns of growth occur, but the nodular pattern is more common. Tumor cells are arranged as sheets, nests, or cords; the latter has been referred to as the reticular pattern. Perineural and vascular invasion are evident in many myoepithelial carcinomas. While marked cellular and nuclear pleomorphism, high number of mitotic figures and necrosis are indicative of malignancy. Myoepithelial carcinoma is distinguished from myoepithelioma on the basis of infiltrative growth, cytologic abnormalities, or both. In our case, marked mitosis and infiltrative growth was seen.

Immunohistochemically myoepithelial cells in their normal environment, i.e., in a gland not involved by neoplasm or inflammation, express CK14, smooth muscle actin,
muscle-specific actin, calponin, smooth muscle myosin, and p63. Non-neoplastic myoepithelial cells in an area adjacent to neoplasm or in an area of inflammation often also express S-100 protein and glial fibrillary acidic protein, neoplastic myoepithelial cells variably express immunoreactivity to these antigens.[19,22] S-100 protein is often a sensitive marker for neoplastic myoepithelium,[17] but is not specific for myoepithelial carcinoma since normal and neoplastic intercalated duct cells also sometimes express S-100 protein. P63 also appears to be a sensitive marker for myoepithelial differentiation, but normal and neoplastic ductal and epidermoid basal cells also express p63. Similarly, vimentin is a sensitive but not specific marker in myoepithelial carcinoma.[17] Calponin was a more sensitive myoepithelial marker in the series reported by Savera et al.[5] It is also found that the proliferation index derived from immunostaining for Ki-67 could differentiate between benign and malignant tumors, a proliferation index above 10% indicated malignancy.[17,22] Same proliferation index was used in our case, which was high (> 10%), confirmed the malignant nature of tumor.

Treatment and prognosis

Complete excision is the preferred treatment method for myoepithelioma. For myoepithelial carcinoma, complete excision with tumor-free margin remains the first choice of treatment,[6,17,18,22] in spite of the possibilities of local recurrence and distant metastasis to lung, kidney and brain.[2,20] Adjunctive 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.[32] In a study by Stromeyer et al., they found that the tumor is not sensitive to radiation, while Takeda reported a good clinical response to radiation.[9] Chemotherapy was unsuccessful in controlling extensive diseases in four cases, but may have been helpful in another.[4]

Different studies regarding recurrence and prognosis of myoepithelial carcinoma provided variable data. In a study of 25 patients published by Savera et al.,[10] 10 patients developed recurrences, mostly multiple. Follow-up of 17 patients out of these 25 showed that 47% (eight patients) developed metastasis and 29% (five patients) died of disease after a mean of 32 months. Two patients were alive with disease (19 and 49 months). 59% (10 patients) were without any evidence of disease after a mean of 42.2 months. In another study, Pontes et al.[6] reviewed 69 cases previously reported in English literature, concluded that 72.5% did not present recurrences or metastasis. However, recurrence was observed in 8.7% of cases and metastasis in 5.8% of cases, especially to the cervical lymph nodes and the lungs. A total of 4.3% of patients affected, died from the disease and in 8.7% the follow-up features were not specified. As it is a rare neoplasm, the clinical parameters and pathological factors for determining the prognosis of patients affected by myoepithelial carcinoma are not well-established, although cellular pleomorphism, p53 over-expression, extensive invasion to surrounding tissues, perineural permeation, and high cellular proliferation may be correlated with a poorer clinical outcome.[6,22]

Conclusion

We reported a rare case of myoepithelial carcinoma of buccal mucosa. The case is rare also because of unusual location of the tumor. This report also provides the summary of the previous literature about myoepithelial carcinoma. Sometime diagnosis of myoepithelial carcinoma is difficult only by histopathological examination so role of immunohistochemistry in diagnosis should be considered as elucidated in present case. The concept, histological feature, immunohistochemical features, genetic changes, treatment options, and prognosis of myoepithelial carcinoma should be further discussed to uncover more cases.

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