

Malignant myoepithelioma of palate

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Abstract

Malignant myoepithelioma is a rare salivary gland neoplasm, which accounts for less than 2% of all the salivary gland carcinomas. Majority of cases have been reported in parotid, and only 8 cases of involvement of the hard palate have been reported in the literature so far. Hereby, a case of painless, ulcerated palatal mass of 2 years of duration reported. A diagnosis of malignant plasmacytoid myoepithelioma was made with the aid of immunohistochemical analysis, and wide surgical excision was considered keeping in mind the biological behavior of the tumor.

Keywords: Malignant myoepithelioma, plasmacytoid variant, salivary gland neoplasms

Introduction

Salivary gland tumors account for up to 6.5% of all neoplasms of the head and neck region and around 21% to 46% of these tumors are malignant.^[1] Among these, myoepitheliomas are rare tumors of myoepithelial differentiation, which account for 1.5% of all the salivary gland tumors. Its malignant counterpart, myoepithelial carcinoma or malignant myoepithelioma (MME), is much rarer and comprise less than 2% of all the salivary gland carcinomas.^[2] Myoepithelial cells are difficult to recognize in routine hematoxylin and eosin (H and E) stained sections and do not exhibit consistent immunohistochemical phenotype.^[1] Increasing number of salivary gland tumors have been studied in recent past, and MME may not be as rare as has been suggested before in the literature due to their recent recognition as a separate entity.^[1-3]

Case Report

A 50-year-old female reported with the complaint of a painless mass in the hard palate for the last 2 years, which progressively increased to the present size over the period of time. On examination, there was a smooth, bulging,

6 cm × 5 cm in size sessile mass in the left palatal region crossing the midline with areas of ulcerations [Figure 1]. The swelling was soft to firm, non-tender, and negative on aspiration. Standard occlusal radiograph of the maxilla revealed no erosion of the underlying bone [Figure 2]. After appropriate medical history, routine hemogram, and informed patient consent, incisional biopsy was performed from the representative area under local anesthesia, and further histopathological examination was performed.

Histopathological section stained with routine hematoxylin and eosin under the scanner view revealed intact stratified squamous surface epithelium with a cluster of small duct-like spaces lined by small deeply staining cells just beneath the epithelium. Deeper part of the section revealed pleomorphic large round cells with scanty eosinophilic cytoplasm and eccentrically placed nuclei in sheets along with areas of hyperchromatic spindle cells as shown in [Figure 3a]. Under higher magnification (40×), solid sheets of large, highly pleomorphic cells with hyaline-like cytoplasm, eccentrically placed nuclei, and scattered nucleoli were observed. Frequent mitotic figures were encountered. The stroma was scanty and avascular and showed occasional eosinophilic structures with fine needle-like processes radiating outwards as shown in [Figure 3b].

Further collagenous nature of the spherules was confirmed with Von Geison's stain, which stained red as shown in Figure 3b inset. Immunohistochemical analysis showed immunoreactivity for cytokeratin 5, 6, alpha smooth muscle actin, calponin as shown in [Figure 4a-d] but negative to epithelial membrane antigen (EMA), and Ki 67, respectively. So, the present case suitably fits into the criteria set by WHO 2005 guidelines set for malignant myoepithelioma, according to which, immunoreactivity for cytokeratin and at least one of the myoepithelial markers, including smooth muscle actin, GFAP, CD10, calponin, and smooth muscle myosin heavy chain, is required for diagnosis.^[2]

Differential diagnosis includes pleomorphic adenoma ex-carcinoma and metastatic melanoma^[4]

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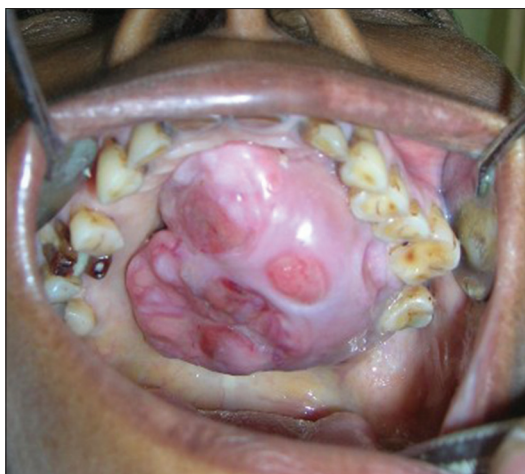


Figure 1: Intra-oral photograph showing palatal mass with areas of ulcerations

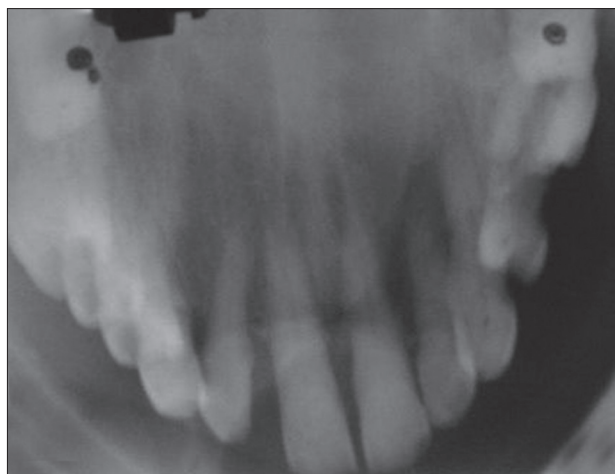


Figure 2: Standard occlusal radiograph of maxilla showing no bone loss

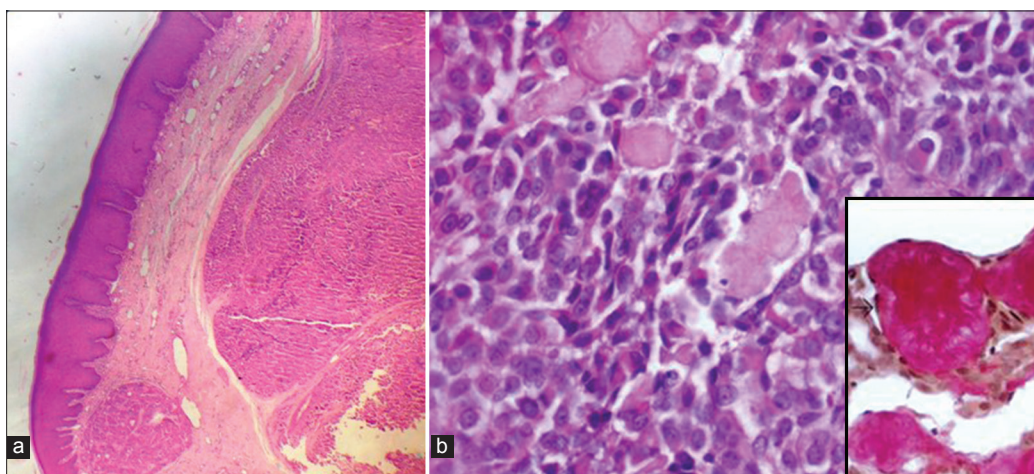


Figure 3: (a) H&E (10 × view). (b) Collagenous spherules showing fine needle-like radiating structures. (40 × view). Inset: Under Von Geison's stain

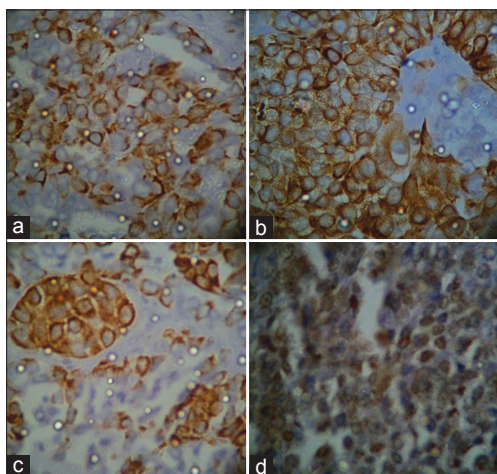


Figure 4: (a) Cytokeratin 5 (40 × view). (b) Cytokeratin 6 (40 × view). (c) Alpha Smooth Muscle Actin (40 × view). (d) Focal immunopositivity for Calponin (40 × view). (e) Negative for Epithelial Membrane Antigen (40× view)

After the diagnosis, the patient was referred to the department of oncosurgery where wide surgical excision of the tumor mass was performed. Post-operative histopathological diagnosis confirmed the initial diagnosis. No other adjuvant radiotherapy or chemotherapy was delivered. Patient reported with no signs of local or regional recurrence 6 months post-operatively.

Discussion

Myoepithelial cells are ectodermally-derived contractile cells, routinely identified in many normal tissues with secretory functions such as salivary glands, lacrimal glands, breast, and prostate.^[5] Salivary gland tumors with myoepithelial cell participation include pleomorphic adenoma, myoepithelioma, basal cell adenoma, adenoid cystic carcinoma, polymorphous low-grade adenocarcinoma, epithelial myoepithelial carcinoma, and carcinoma ex-pleomorphic adenoma.^[3]

The first case of malignant myoepithelioma was described by

Stromeyer *et al.* in 1975 in the parotid.^[4,6,7] It was first defined by Ellis GL in 1991^[4] and appeared for the first time in the WHO classification as a distinct clinicopathological entity in same year.^[6,7] Since then, large number of cases have been reported in the parotid gland. Its involvement of hard palate is extremely rare, and there are only 8 cases reported in the world with short-term follow-up.^[4]

The mean age of patients at the time of presentation is 55 years (range 14-86 years) with equal gender predilection. 75% of the cases arise in the parotid, but they also occur in the sub-mandibular and other minor salivary glands.^[2,3]

Malignant myoepitheliomas are usually unencapsulated, but may be well-defined with nodular surfaces and the tumor size ranging from 2 to 10 cm.^[2,3] It has been reported that MME may arise *de novo*, but at least 50% develop in the pre-existing pleomorphic adenoma or benign myoepithelioma.^[2,3] The tumor is locally destructive, and majority of patients report with the complaint of painless mass.

The microscopic architecture is often multinodular with infiltration into adjacent structures. The nodules comprise of solid sheet-like or trabecular or reticular patterns, but they can be dissociated, often with plentiful myxoid or hyaline material and occasional central necrosis. The range of cell types reflects that seen in its benign counterpart i.e. spindled, stellate, epithelioid, plasmacytoid (hyaline) or occasionally, vacuolated with signet ring-like appearance.^[2] Rarely, malignant myoepithelioma contains duct-like lumina, usually with non-luminal cell differentiation of the lining cells. Usually, most malignant myoepithelioma are less monomorphic than benign myoepitheliomas. High mitotic activity (3 to 51 per 10 × high power fields) and atypical forms are observed frequently.^[3]

Various immunohistologic studies have revealed variable expressions to S-100, vimentin, broad-spectrum cytokeratin and other myoepithelial markers, including S100, SMA, GFAP, CD10, calponin, maspin, and SMMHC (smooth muscle myosin heavy chain).^[2,3,7-9]

Tumor can involve the adjacent bone. Regional and distant metastases are uncommon, but may occur late in the course of disease.^[2,10] Wang Z had reported secretion of matrix-degrading proteinases, as well as proteinase inhibitors appears to be associated with demonstrated inhibition of angiogenesis and thus anti-invasive effect. Perineural invasion is seen in 44% and vascular invasion in 16% of cases.^[3]

Electron microscopy studies have confirmed both epithelial and smooth muscle differentiation with small desmosomes and few actin filaments. Longitudinally-oriented 6-8 nm fine cytoplasmic microfilaments with focal dense bodies, pinocytotic vesicles, hemidesmosomes, and intermediate filaments have been observed.^[3,11]

Comparative genomic hybridization has revealed infrequent abnormalities, and only 5 have manifested chromosome 8 alterations.^[2] Young J *et al.*, in 2002, reported a case of malignant myoepithelioma with biallelic inactivation of the tumor suppressor gene APC in a patient with familial adenomatous polyposis.^[12,13] But, no distinct cytogenetic aberrations have been reported.

Wide surgical excision is the preferred treatment.^[6] The prognosis of malignant myoepithelioma is variable, but the reports suggest that approximately one-third of patients die of the disease, another third have residual tumor, and remaining third are disease-free.^[1,2] There is no reported difference in the clinical behavior of “*de novo*” myoepithelial carcinomas and those arising in pleomorphic adenoma and benign myoepithelioma.^[2] To conclude, we hereby report an additional case of malignant myoepithelioma of palate, which was successfully treated with wide surgical excision, avoiding overzealous treatment.

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