Case Report

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Myoepithelial Carcinoma: A Rare Entity

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ABSTRACT

Background: Myoepithelial Carcinoma (MC) (malignant myoepithelioma) is a rare salivary gland tumor composed of entirely of myoepithelial cells that exhibits a dual epithelial and smooth muscle phenotype. We report a case of soft tissue myoepithelial carcinoma in 49 years old female patient which presented as unilateral mass on the hard palate. Histological examination revealed multi-lobular growth pattern of epitheloid cells with marked atypia and frequent mitosis. Immunohistochemistry was reactive for p63, CD 10, Ae1 and S100. Rarity in oral cavity among all the other salivary neoplasm (0.2%) and heterogeneity in cellular morphology and architectural patterns may lead to misdiagnosis of extra glandular myoepithelial carcinoma. Careful and meticulous observation of both histopathological and Immunohistochemical features are mandatory for correct diagnosis of such entities.

Keywords: Myoepithelial carcinoma, oral cavity, hard palate, immunohistochemistry.

INTRODUCTION

Myoepithelial carcinoma (MC) is defined by the World Health Organization (WHO) classification in 1991 as a rare low-grade malignant neoplasm and account for less than 1% of salivary glands tumors.[3] It is a rare neoplasm that was first described in 1975 by Stromeyer et al. Though the name was coined by Donath et al in 1972, it was likely recognized as early as 1956 and reported under variety of names, such as adenomyoepithelioma, clear cell adenoma, and tubular solid adenoma[2]. This tumor arises most frequently (80%) in the parotid gland, but lesions have also been reported in submandibular glands (10%) and minor salivary glands (1%)[1].The diagnosis is confirmed by immuno- histochemical and ultrastructure investigations[3]. Approximately half of salivary gland MC arises in association with a benign mixed tumor or myoepithelioma[3]. Histologically, morphological heterogeneity is the hallmark in MC, and the majority of neoplasms display more than one cell type[3]. Immunohistochemically, these tumors are mostly positive for vimentin, S-100protein, cytokeratinand 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, CAM5.2 and epithelial membrane antigen, amongst others[3].

CASE REPORT

A 49 years old female patient was referred to Darshan Dental College, Udaipur in Rajasthan, for diagnosis and treatment of a lesion in hard palate. She had noticed the lesion about 1 year back and complained that the lesion was “growing and difficulty in breathing”. On extra-oral examination

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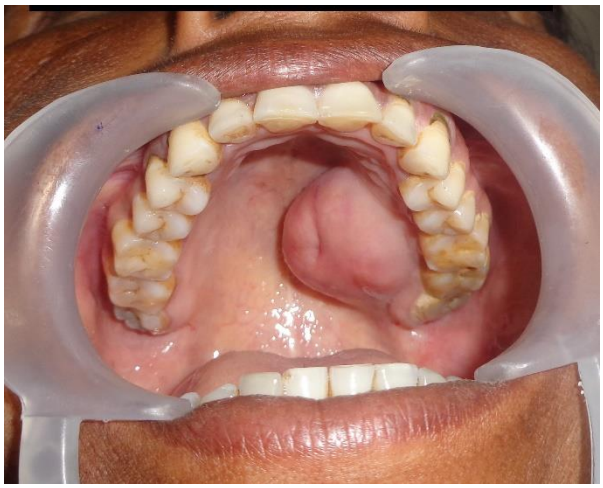

Fig 1: Tumoral lesion observed in hard palate region. Overlying mucosa is normal in colour and lesion is well circumscribed.

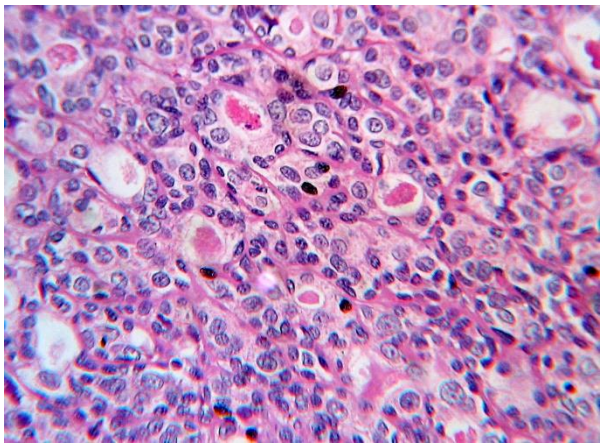

Fig 2: Histopathological picture of MC, Duct like arrangement of cells is seen and a raw cuboidal darkly stained cells form a lumen, often containing PAS-positive material. This is covered by one or more layer of myoepithelial cells covered with pale or clear cytoplasm.

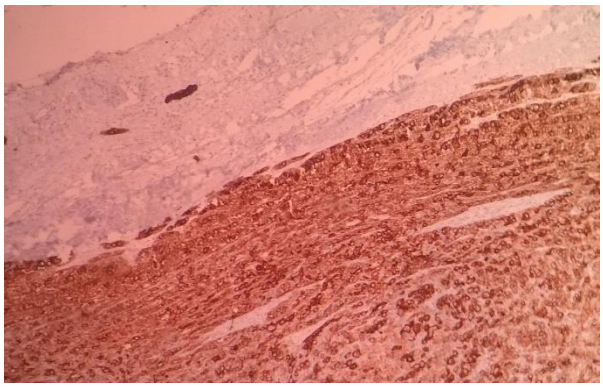

Fig 3: ae1 marker is immunoreactive for myoepithelial cells.

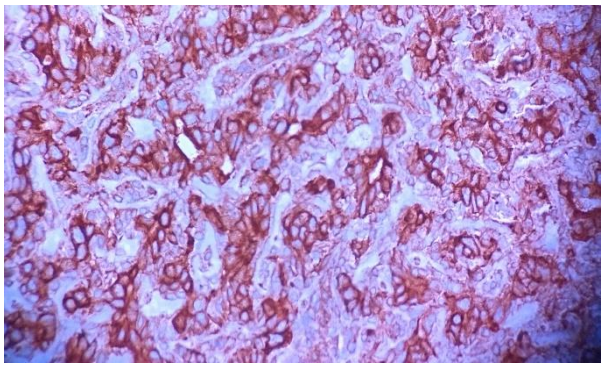

Fig 4: p63 is positive showing brown colored myoepithelial cells.

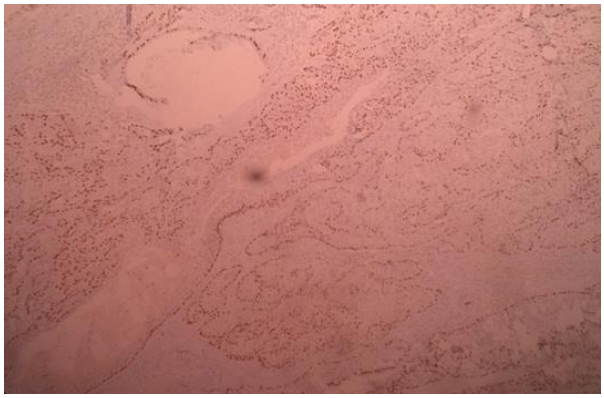

Fig 5: Vimentin is positive and negative in epithelium.

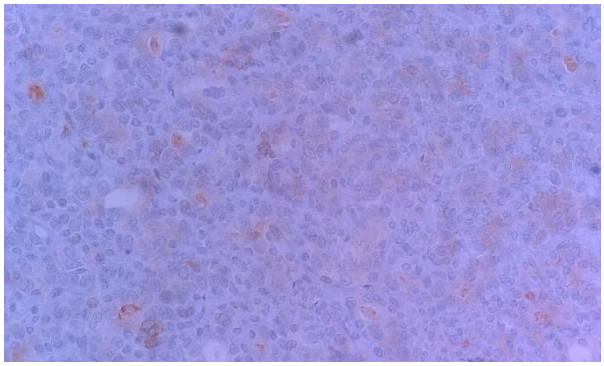

Fig 6: Brown coloured myoepithelial cells are showing positive immunoreactivity for s100.

no alterations were observed. Intra-oral examination showed fibrous tumoral lesion in hard palate, having regular surface, which was erythematous and asymptomatic (Fig. 1).

Gross Appearance

The lesion was 3.5 cm x 3 cm in size, roughly round in shape and overlying mucosa was pink in colour. The initial clinical diagnosis was

Mucoepidermoid Carcinoma and Adenoid Cystic Carcinoma. A panoramic radiograph was advised, in which there was no bone involvement. In incisional biopsy the histological diagnosis was adenocarcinoma not otherwise specified.

Histopathological Features

The patient underwent subsequent excisional biopsy. Microscopically, observed by hematoxylin-eosin staining the presence of duct-like structures with an inner layer of epithelial cells layer of clear myoepithelial cells which are forming solid nests and myxoid islets. Duct like arrangement of cells was also seen. Cuboidal darkly stained cells formed a lumen, at places containing PAS-positive material which was covered by one or more layer of myoepithelial cells covered with pale cytoplasm. (Figure 2)

Immunohistochemical Evaluation

Immunohistochemical staining was carried out with the streptavidin-biotin method. Immunoreactivity for Ae1, p63, and S100 were observed in epithelial cells. Ae1 is immunoreactive for myoepithelial cells; brown colour is showing immunoreactivity for the malignant myoepithelial cells. (Figure 3a) strands of myoepithelial cells are showing positive reactivity in p63 marker. (Figure 3b), vimentin is also immunoreactive for the malignant myoepithelial cells and is nonreactive in epithelium. (Figure 3c) and brown coloured myoepithelial cells are showing positive immunoreactivity for s100. (Figure 3d) Immunohistochemical examination revealed malignant myoepithelial cells which were immunoreactive for above markers and thus the diagnosis myoepithelial carcinoma was confirmed.

Management

In our case surgical excision was done with the margin all around the tumor. No recurrences or metastasis were found 12 months after the surgery. The adjunctive radiation therapy is sometimes used, and in this case, the patient was submitted 33 sessions of radiotherapy and observed 6 months follow-up after the radiotherapy without any distant metastasis.

DISCUSSION

Epithelial-myoepithelial carcinoma, recognized by WHO in 1991, is a rare malignant neoplasm in the salivary glands, comprising approximately 1% of all salivary gland tumors[1]. The case related was localized in the soft palate, a rare area of onset, where it showed histologic characteristics of a low-grade behavior of aggressiveness. Rarely, high-grade or dedifferentiated EMC has been described[2, 3]. These tumors arise in the salivary glands, predominantly the major salivary gland and the parotid gland. However, they have rarely been observed in tissues other than the salivary glands, such as the breast, lacrimal gland, lung and nasal cavity[1].

Microscopically on histological examination the tumor is biphasic and characterized, as in this case, by tubules lined by an inner layer of cytokeratin-positive bland cuboidal epithelial cells surrounded by an outer layer of S100 positive myoepithelial cells. Characteristic extracellular globules of hyaline basement membrane material are present[7]. Typically the tumor nodules have an infiltrative margin and perineural and vascular invasion are present. In keeping with other biphasic tumors the diagnosis maybe difficult histologically and other salivary gland tumors such as pleomorphic adenoma, adenoid cystic carcinoma and also tumors with a predominant clear cell population such as clear cell carcinoma and sebaceous carcinoma should be considered before rendering the diagnosis[7]. Fine needle aspiration cytology is a baseline investigative tool in the assessment of patients with salivary gland swellings. Whilst it is an accurate method of distinguishing neoplastic from non-neoplastic lesions it may not be possible to always accurately predict a specific tumor type due to the over-lapping spectrum of cytological appearances found in a wide variety of salivary gland neoplasms[7]. Four cell types are identified in myoepithelial salivary gland neoplasms: namely, plasmacytoid, spindle, epithelioid and clear cell types. Of these, the clear cell type is the least frequently encountered[7].

According to the literature, the treatment is varied and the main method is surgery with a safety margin and if necessary, subsequent using of radiotherapy and chemotherapy. In some cases,

treatment may include enlargement of the incision and neck dissection with lymph node emptying. In the present clinical case the initial treatment was a local surgical incision with safety margin, even without prior confirmation of a malignant lesion of salivary gland. Since it was confirmed the malignant nature of the lesion, the patient was referred to an oncology service[1]. In conclusion, it is noteworthy; therefore, the importance of critical and reflective evaluation by the dentist as well as diagnostic and prognostic value of histopathology and immunohistochemistry, especially for cases of malignant tumors that do not exhibit clinical features that may characterize such condition[1].

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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