Myoepithelial carcinoma arising in a benign myoepithelioma of the palate. Case report and literature review

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The study hasn't been presented anywhere.

Abstract

Myoepithelial carcinomas are tumors arising from myoepithelial cells mainly or exclusively, they showed varied cell types and patterns leading to a wide range of differential diagnoses. Immunohistochemical analysis helped to determine the diagnosis. Recognition of myoepithelial carcinoma is clinically significant because, compared to its benign counterpart (myoepithelioma), It has increased frequency of local recurrences and metastases, which warrants close clinical follow-up. The aim of this case report was to present a rare neoplasm, myoepithelial carcinoma arising from a benign myoepithelioma of the palate, and to review its diagnostic criteria, pathologic and clinical characteristics, treatment options and prognosis.

Background

Myoepithelial tumors of the salivary gland including myoepitheliomas (benign) and myoepithelial carcinomas (malignant) are a rare group of tumors. Although myoepitheliomas were first described as early as 1943 by Sheldon [1], the best description of myoepithelial tumors was given in the landmark articles by Dardick et al in 1989 [2] and Dardick in 1995 [3]. Myoepithelial tumors have also been included as a separate entity in the second edition of the World Health Organization's histological classification of salivary gland tumors (1991) [4-5]. Herein we present a case of malignant myoepithelial tumor araising in a benign myoepithelioma of the palate.

Observation

A 76 years old man consulted the clinic with chief complaint of a painful swelling in the hard palate which does not response to neither symptomatic nor antibiotic treatment. He underwent a biopsy of palate lesion which shows a benign myoepithelioma. A complete resection of the tumor was performed without any adjuvant treatment.

Nineteen years after, he presented to our institution with a history of recurrence of palate tissue mass that progresses since 3 years. The oral examination and the panendoscopy showed a great mass arising from the soft palate and packed the hole of oral cavity with invasion of: hard palate, tonsillar fossa, lateral wall of the oropharynx, and nasopharynx which was obstructed **(Fig.1)**. A facial computed tomography (CT) confirmed the local tumor invasion, without any lymph nodes involvement **(Fig.2)**.

An urgent tracheotomy was performed after the installation of an acute dyspnea. A biopsy was performed, the tumor showed proliferation of a double component: plasmocytoïd-cells showing eccentric nucleus and pale eosinophilic cytoplasm, they also were weakly cohesive and were arranged in sheets or trabeculae **(Fig.3)**; and spindle cells with centrally placed elongated nuclei, the mitosis was rare, there was no necrosis.

A fascicular arrangement of tumor cells was noted. These tumor-cells were disseminated in a variable amount of hyaline stroma. A tumor infiltration in the normal salivary glands and the adjacent adipose was found. Immunohistochemical stains were performed: the epithelial component was positive to keratine marker, the smooth muscle component positive for Protein S 100 (PS100), vimentine and calponin **(Fig.4 and 5)**.

Despite the absence of frank histological criteria of malignancy (atypia, mitosis and necrosis), the invasive character referred to radioclinical and histological data (massive infiltration of salivary glands), lead to the diagnosis of low grade myoepithelial carcinoma araising in a benign myoepithelioma of the soft palate. The resection of tumor has been impossible due to the massive infiltration of the hole oral cavity and pharyngeal space, and in front of the advanced patient age, impaired karnofsky scale and nutritional condition, chemotherapy was not indicated in the first intension. We decided then to process by radiotherapy to reduce the tumor seize. He is actually under treatment.

Discussion

Myoepithelial tumors arise from myoepithelial cells that surround acini and ducts of salivary glands, these cells exhibit both epithelial and smooth muscle cell characteristics [6, 7].

These tumors commonly occur in major salivary glands but can arise in the submandibular, sublingual and rarely in minor salivary glands of the oral cavity [8-11]. In a large Indian series of 51 cases of myoepithelial carcinoma, tumors were located essentially in the parotid gland (29.4%), palate (29.4%), oral mucosa (13.7%), nasal cavity (9.8 %), and maxilla, lower alveolus and tongue (16%) [5].

Myoepithelial carcinoma can appear de novo (77 %) or develop in a pre-existing benign tumor (23%). The time between the onset of benign tumor and the occurrence of myoepithelial carcinoma is variable; it is 10 years in our case.

Our patient is 76 years old, however, cases reported in the literature are of an age between 14 and 70, most patients were in their third to fifth decade of life; for the majority of them, the primary complain was a painless mass. Usually, myoepithelial carcinoma had a localized presentation at initial diagnosis with mean tumor seize of 4 cm [5]. Our case is different from other cases of the literature by the importance of local and regional extension of primitive tumor.

At histological study, the tumor cells showed a wide morphologic variation: epithelioïd
(29%), plasmocytoïd (14%), spindle (12%), stellate (16%) or mixed (24%). High-grade tumors showed nuclear pleomorphism and/or large areas of necrosis and mitosis [5]. The tumour cell may form solid and sheet-like formations, trabecular and reticular patterns, but they can also be dissociated, often within plentiful myxoïd or hyaline stroma [12].

The diagnosis myoepithelial tumor can be helped by immunohistochemical (IHC) analysis that shows high expression of epithelial markers such as cytokeratin, epithelial membrane antigen (EMA), S-100 protein, and markers of smooth muscle origin such as smooth muscle actin and calponin on the tumor cells of myoepitheliomas. Current IHC criteria for the confirmation of myoepithelial differentiation are double positivity for both cytokeratins (pan CK or preferentially basal type CK) and one or more myoepithelial immunomarkers (S-100, calponin, p63, GFAP, maspin, actins, and a variety of myogenic markers) [13,14,15]. However, it must be noted that these markers are not always positively expressed in the tumor cells and that negative staining does not necessarily exclude myoepithelial differentiation [3]. IHC findings in our study are consistent with those of previous reports. In our cases, tumors positivity for both cytokeratins and myoepithelial markers (S-100 and Calponin) confirms the diagnosis of myoepithelial carcinoma.

Indications of malignancy are based on features such as nuclear atypia, high mitotic rate and infiltrative growth into adjacent tissues. Currently, benign and malignant myoepitheliomas are differentiated by mitotic count, presence of invasive growth, cellular polymorphism, tumour necrosis, or their combination. Destructive growth and infiltrative character distinguishes myoepithélial carcinoma from benign myoepithelial tumors [12]. In our case, despite the absence of frank histological criteria of malignancy (atypia, mitosis and necrosis), the invasive character referred to radioclinical and histological data (massive infiltration of salivary glands), lead to the diagnosis of low grade myoepithelial carcinoma. The differential diagnosis of myoepithelial carcinoma includes a wide range of neoplasms, depending on the predominant cell type. It is sometimes difficult to differentiate myoepithelial carcinoma showing epithelioid morphologic characteristics from other salivary gland neoplasms showing myoepithelial differentiation, especially adenoid cystic carcinoma, polymorphous low-grade carcinoma [16]. In tumors with clear-cell morphologic characteristics, the differential diagnosis includes hyalinizing clear-cell carcinoma, epimyoepithelial carcinoma, and metastatic renal cell carcinoma [17]. Melanoma, high-grade lymphoma, or plasmacytoma must be ruled out when the tumor shows plasmacytoid differentiation. With spindle cell morphologic characteristics, the most common differentials diagnosis are sarcomatoid squamous carcinoma, spindle cell melanoma, and sarcoma [5, 16].

Given their rarity and only recent recognition, there is no consensus on the optimal treatment. Complete excision is the preferred treatment method for myoepithelioma. For myoepithelial carcinoma, complete excision with tumor-free margin with or without nodal dissection remains the first choice of treatment, in spite of the possibilities of local recurrence and distant metastasis [7, 16, 18]. Local radiation therapy and chemotherapy can be needed for myoepithelial carcinoma particularly in palliative situations. There is a case reported by Ibrahim and al [19] consisted in myoepithelioma of the hypopharynx and larynx with lymphatic invasion and liver metastasis treated by chemotherapy and palliative laryngeal radiotherapy with 3 month of follow-up, the patient was died of disease. There is a single published case report of a patient with metastatic myoepithelial carcinoma of the vulva who showed a complete response to chemotherapy with carboplatin/paclitaxel [20].

Compared with myoepitheliomas, myoepithelial carcinoma shows highly aggressiveness and high rate of recurrence even after adequate therapy [7, 21]. The most common site of metastasis is lung, followed by lymph nodes, bone and soft tissue with an average rate of 47%. Interestingly, this pattern of spread has features of both carcinomas (lymph nodes) and sarcomas (lung) [22].

Recurrence and metastasis are more common in children than in adult even with a negative excision margin [1]. Therefore, Yu suggested myoepithelial carcinomas of the salivary gland should be classified as high-grade malignancies [2].

In the Indian study [5] of 51 cases of myoepithelial carcinoma main prognostic factors for local recurrence were stellate, clear, and spindle cell types; large tumor size; and perineural or bone invasion. Similarly, a high incidence of metastasis was noted with the presence of positive margins, large areas of necrosis, high mitotic count (> 4 per 10 hpf), Ki-67 labeling index of 4% to 10%, nuclear atypia, and spindle cell morphologic characteristics. There is no difference in clinical behaviour of "de novo" myoepithelial carcinomas and of those arising in pleomorphic adenomas and benign myoepitheliomas [12].

Conclusion

We report a rare case of myoepithelial carcinoma arising in a benign myoepithelioma of the soft palate. Myoepithelial carcinomas showed varied cell types and patterns leading to a wide range of differential diagnoses. Immunohistochemical analysis helped to determine the diagnosis. Recognition of myoepithelial carcinoma is clinically significant because, compared to its benign counterpart (myoepithelioma), myoepithelial carcinoma has increased frequency of local recurrences and metastases, which warrants specific treatment and close clinical follow-up. Overall, the prognosis of a myoepithelial carcinoma is poor. However, a better clinical outcome can be expected if proper management and suitable operations are performed for patients.

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