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Epithelial-Myoepithelial Carcinoma of the Palate on Pleomorphic Adenoma: Case Report and Review of the Literature: Experience of the Oncology-Radiotherapy Department of the Mohammed VI University Hospital, Marrakech

Dalila Sounny-Slitine[1*], Chahrazade ElAlouani[2], Sophia Berrada[3], Ouassima Elkadiri[1], Raja Ousalm[1], Mouna Darfaoui[4], Abdelhamid El Omrani[4], Mouna Khouchani[4]

[1]Resident, Radiation Oncology Department, Mohammed VI University Hospital, Marrakesh, Morocco
[2]Radiation Oncology Department, Mohammed VI University Hospital, Marrakesh, Morocco
[3]Anatomo-Pathologist Department, Mohammed VI University Hospital, Marrakesh, Morocco
[4]Professor, Radiation Oncology Department, Mohammed VI University Hospital, Marrakesh, Morocco

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***Corresponding author:** Dalila Sounny-Slitine
Resident, Radiation Oncology Department, Mohammed VI University Hospital, Marrakesh, Morocco

Abstract **Case Report**

Epithelial-myoepithelial carcinoma (EMC) is a rare malignant tumor of the salivary glands, mainly affecting the parotid gland. Its association with a pleomorphic adenoma is exceptional. We report the case of a 64-year-old Moroccan woman with a pleomorphic adenoma of the palate that had been evolving for several years. The recent increase in the mass associated with the appearance of homolateral cervical adenopathies led us to think of a malignant transformation. Final histological examination after resection of the mass showed an unexpected association of a pleomorphic adenoma and EMC. EMC is a low-grade malignant tumor. It can arise de novo or on a pleomorphic adenoma. Malignant transformation of the adenoma is suspected when the volume of the adenoma increases rapidly, with the appearance of cervical adenopathy. However, these clinical changes may herald the appearance of a distinct tumor. Despite its tendency to local recurrence and low metastatic potential, rare cases of EMC may exhibit aggressive behavior and distant metastases. Treatment consists mainly of complete surgical resection if possible, followed by radiotherapy to prevent local recurrence, and this is the treatment our patient received in this case.

Keywords: Pleomorphic adenoma, Epithelial-Myoepithelial carcinoma, Salivary gland.

INTRODUCTION

Tumors of the salivary gland are rare. They account for 1-4% of all head and neck tumors. Benign tumors are the most common, dominated by the pleomorphic adenoma, known as a mixed tumor due to its dual epithelial and mesenchymal component. Its extra-parotid localizations are rare [1], and it is characterized by a slow evolution and a potential for malignant degeneration observed in 3 to 14% of cases [2]. Malignant tumors are less frequent, and can develop de novo or from a pleomorphic adenoma. Among these tumors, epithelial myoepithelial carcinoma (EMC) remains very rare, accounting for less than 1% of salivary gland tumors [3]. It is characterized by tubular and solid growth pattern [4], The most common arising sites of myoepithelial carcinoma lie in the parotid gland [5], as well as the nasopharynx, paranasal sinus and nasal cavity of head-neck region [6, 7]. Myoepithelial carcinoma rarely occurs in the palate, this tumor has a slight female predilection and the mean age of patients at diagnosis is 60 years [8], and it's a low-grade malignancy with good prognosis, low recurrence rate and rare metastasis [9]. Here, we report a case of myoepithelial carcinoma on pleomorphic adenoma of the palate and we will develop the main clinical, histological and therapeutic characteristics of this clinical entity.

PATIENT ET OBSERVATION

The patient was a female of 64 years of age, non-smoker, non-drinker, with no history of any systemic disease or any drug history, with a surgical history of thyroidectomy under Levothyrox-based treatment. The history of his illness dated back 2 years, with the appearance of a slow-growing painless mass on

the left side of the soft palate. There was no history of bleeding or tumors elsewhere in the body. On clinical examination, a sessile nodule measuring 4x3 cm was noted on left side of hard palate. The overlying mucosa was intact and normal in color. The lesion was soft in consistency, and in palpation it was nontender. She stated no history of pain, paresthesia, or dysphagia. As well, there was no evidence of any lymphadenopathy on palpation (Figure 1). A biopsy was performed (Figure 2), revealing a salivary tumor with no obvious signs of malignancy, in favor of a pleomorphic adenoma. Surgical treatment was indicated, with removal of the mass. The final anatomopathological result was surprising: it was an association of two perfectly distinct tumors: a pleomorphic adenoma and a myoepithelial epithelial carcinoma characterized by double-component carcinomatous proliferation (Figure 3, 4 and 5), with extra-adenomatous infiltration of 2mm, and healthy surgical resection limits. One month after surgery, clinical examination revealed an erythematous lesion with whitish soft palate, with no loss of substance, and no palpable cervical adenopathy. A maxillofacial surgical opinion was sought for possible revision surgery, and the response was that there was no indication for revision surgery. A facial CT scan revealed a mass on the left palate infiltrating the maxillary alveolar bone and the floor of the homolateral maxillary sinus and nasal cavities, and coming into contact with the mobile tongue with a suspicious tumour-like appearance (Figure 6). The patient was referred for bilateral lymph node dissection; histological results did not reveal right or left lymph node metastases. A facial MRI was ordered, revealing fibrous changes in the bony palate, with nodular formation of the mobile tongue lateralized to the left, which was suspicious and should be checked against the rest of the workup (Figure 7). The patient received radiotherapy on the oral cavity + palate + prophylactic lymph nodes at 54 Gy with boost to the tumour bed.

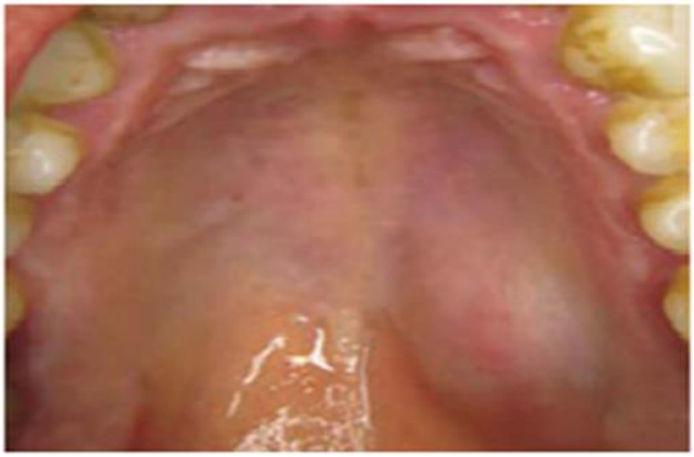

Figure 1: Swelling of the hard palate

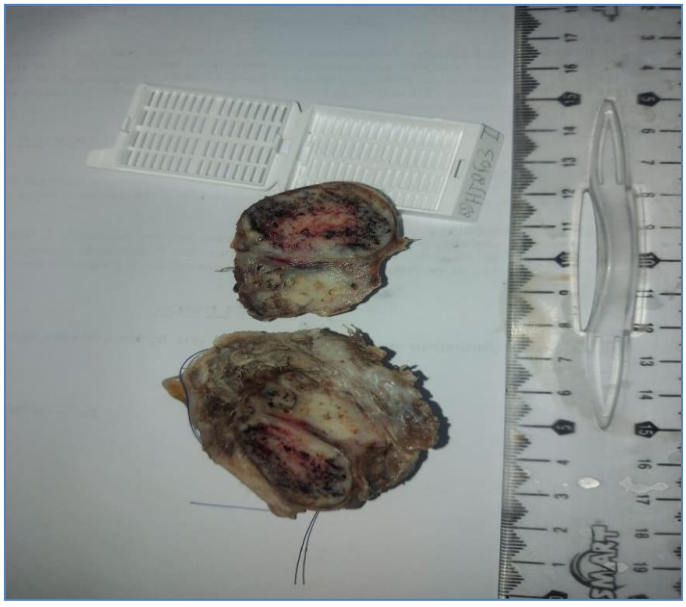

Figure 2: Biopsy fragments taken

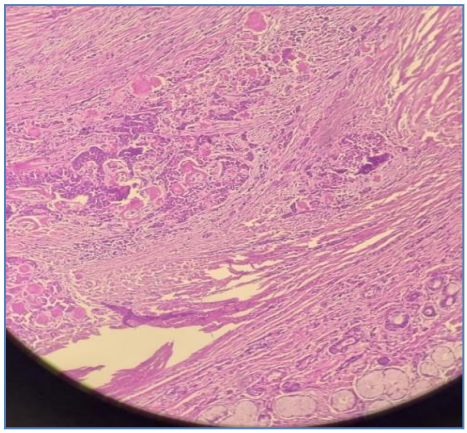

Figure 3: HE GX4 staining showing pleomorphic adenoma

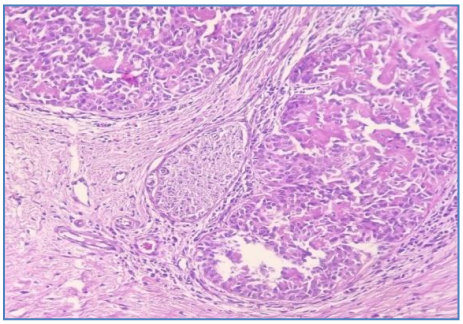

Figure 4: Epitheloid cells showing multinodular architecture (×40).

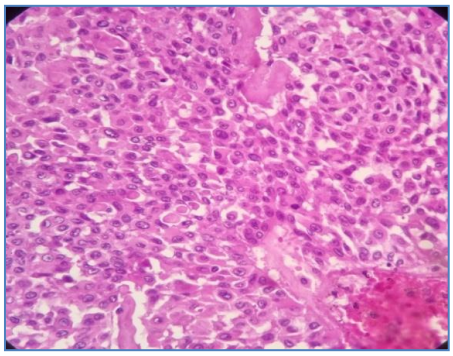

Figure 5: HE Gx10 staining carcinomatous proliferation with dual epithelial and myoepithelial components

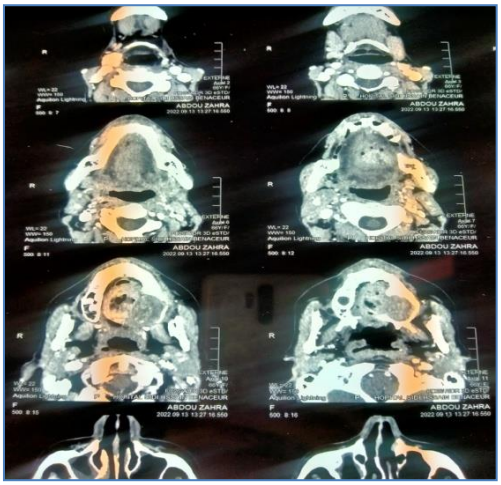

Figure 6: A facial CT scan revealed a mass on the left palate infiltrating the maxillary alveolar bone and the floor of the homolateral maxillary sinus and nasal cavities

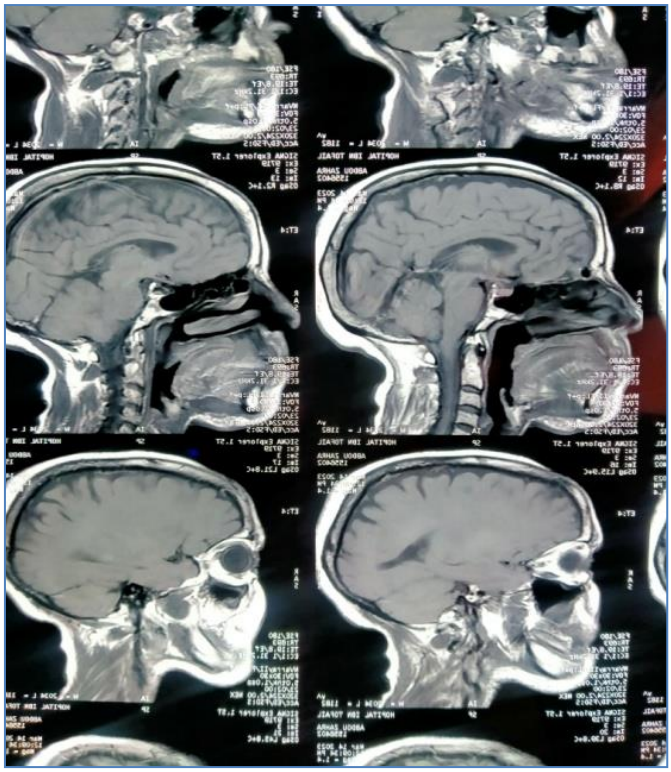

Figure 7: MRI sagittal section showing a nodular formation of the mobile tongue lateralized to the left

DISCUSSION

EMC is a rare tumor [3]. It accounts for less than 1% of malignant tumors of the salivary glands. It preferentially affects the parotid gland, but localizations in the submandibular glands, minor salivary glands and even extraoral glands have also been described [10, 11] . EMC has long been considered a benign tumor. The first description of this tumor was made by Donneth *et al*., in 1972 [12]. CEM was included in the WHO classification of salivary tumours in 1991 as a low-grade malignant tumour. The rarity of this tumour is today underlined by several authors [13, 14]. MEC can arise de Novo or on a pleomorphic adenoma [13]. Transformation is suspected in the presence of a recent increase in size, the appearance of adenopathy, pain or facial paralysis. However, this clinical evolution may signal the appearance of a malignant tumour distinct from the adenoma on healthy parotid tissue. Our case report is a perfect illustration of this rare occurrence. The clinical presentation of EMC is nonspecific, consisting of a slowly growing, painless mass, with facial paralysis being exceptional. Women are most often affected between the ages of 60 and 70 [13]. Macroscopically , the neoplasm is considered a salivary gland of an epithelial-originating tumor with low malignant potential that shows an infiltrative tumor border with no capsule or incomplete capsules and extension into the adjacent salivary gland or other tissues, and microscopically , the cellular composition of myoepithelial carcinoma is complicated, but can include clear cells, spindle-shaped cells, plasmacytoid (hyaline) cells, and epithelioid cells, and it is usually a combination of 2 cell populations [15]. This dual component is confirmed by immunohistochemical studies [10]. Clinically, the presentation of ex-adenoma pleomorphic carcinomas can be similar to that of adenoma pleomorphic. The most frequent symptom is a glandular tumor mass. Around 30% of patients present with symptoms and signs consistent with malignancy: pain, cervical adenopathy and dysphagia [16]. Imaging data for pleomorphic exadenoma carcinoma are usually non-specific, and its differentiation from other malignant and benign salivary gland tumours is difficult [17]. From either the clinical or CT perspective, the tumor can be distinguished from most benign tumors of the salivary gland due to its indistinct lesion edge and infiltrating growth. However, it is similar to myoepithelioma, with inhomogeneous enhancement in imaging. Wang *et al*., noted that myoepithelioma typically has strong enhancement of nodules and a boundary in the arterial phase, and some cases also have a cystic and linear region with no enhancement observed in myoepithelial carcinoma. The enhancement amplitude of myoepithelioma being higher than that of myoepithelial carcinoma could be due to the latter's greater malignancy, which has more necrotic areas. The common manifestations of salivary gland malignant tumors are an unclear tumor boundary, invasion into surrounding structures, uneven density, and frequently observed cystic changes [17, 18]. Complete excision is the preferred treatment method for myoepithelioma [19]. For myoepithelial carcinoma, complete excision with tumor-free margin remains the first choice of treatment, in spite of the possibilities of local recurrence and distant metastasis. In selected cases, the surgery has been followed by adjuvant radiotherapy to avoid local recurrence [20]. The efficacy of

chemotherapy in the treatment is still unclear [21]. Recurrence is rare for benign myoepithelial tumors, while the overall prognosis of myoepithelial carcinoma is poor. The evolution of MEC is marked by the occurrence of local recurrence in 40% of cases, between 9 months and 20 years. Factors favoring recurrence include positive margins, lymph node involvement, tumor necrosis and myoepithelial anaplasia [2]. Several studies reported aggressive clinical behaviors for myoepithelial carcinoma, and the average metastatic rate was 47% and the mortality rate was 29% after a mean of 32 months. Distant metastases have also been reported, the most frequently described locations being the cervical lymph nodes, lung, kidney and brain. These metastases can occur very late, even after ten years of follow-up [13]. Recurrence and metastasis are more common in children than in adults even with a negative excision margin [22]. Therefore, Yu suggested myoepithelial carcinomas of the salivary gland should be classified as high-grade malignancies [23]. Five-year and 10-year overall survival rates are 80% and 72% respectively [12]. A long period of surveillance is therefore mandatory, even for tumors operated on at an early stage with complete surgical resection.

CONCLUSION

A pleomorphic adenoma of the salivary glands always gives rise to fears of malignant transformation, all the more so as it has a long evolutionary history. However, it is still possible for a malignant tumour to develop independently of the adenoma. Epithelial - Myoepithelial carcinoma, a rare tumor, can be a sign of this exceptional association. Although EMC is considered a low-grade malignant tumor, various studies have shown the occurrence of sometimes aggressive local recurrences and late metastases with a fatal outcome. Rigorous follow-up over several years is therefore recommended, even if the tumor is diagnosed at an early stage and completely resected.

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