Epithelial-Myoepithelial Carcinoma of Minor Salivary Gland with Distant Metastasis: A Case Report

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Abstract
Myoepithelial carcinomas are rare tumors, making up only 1% of salivary gland neoplasms. They can form either independently or within an existing pleomorphic adenoma. We encountered a case concerning a 46-year-old woman who approached us with a previous occurrence of a solid mass in the hard palate that had been steadily increasing in size. Based on the results of the biopsy, it has been determined that the mass is an Epithelial-Myoepithelial Carcinoma of Minor Salivary Gland. The extension assessment revealed the existence of lung metastases. Chemotherapy was administered, resulting in stabilization of the process and the lung metastasis.

Index Terms: Myoepithelial Carcinoma, Minor Salivary Gland, Lung Metastasis.

1 INTRODUCTION
Myoepithelial carcinoma of the salivary gland is a rare malignancy accounting for 0.4% to 1% of all salivary gland tumors¹. It may develop de novo or may appear in a pre-existing pleomorphic adenoma². The parotid gland is typically affected, although it can also affect other salivary glands, either major or minor.

It was firstly described as a glycogen rich or clear cell adenoma because of the clear cell component. In 1972, Donath et al. introduced the term "epithelial-myoeplithelial carcinoma" in describing this entity. They described the clinicopathological features of a case of epithelial-myoeplithelial carcinoma of the parotid gland and emphasized the significance of being aware of this tumor in the differential diagnosis of biphasic tumors on fine needle aspiration cytology. They also noted that the myoepithelial component was an integral part of the tumor.

In 1991, the WHO recognized EMC as a distinct entity and subtype of salivary gland adenocarcinoma, and it became part of the new classification system⁴. It primarily affects older women, with the highest frequency occurring in the seventh decade.⁵ Less frequent is the finding of lymph node and hematogenous⁶. Wide surgical excision is the treatment of choice, and radiotherapy is currently recommended for postoperative treatment when a high risk of recurrence or metastasis exists. No standard chemotherapeutic drugs or regimens have yet been established for the treatment of individual types of salivary gland carcinomas.⁷
We present a new case of EMC and discuss the clinicopathological aspects, therapeutic approach of this rare tumor.

II CASE REPORT
A 46 years old woman presented to our institution with a 1-year history of a painless swelling in gingival palatine region, However, she had not sought treatment before this time. The past medical and family histories were noncontributory. there was no facial asymmetry or any sign of neuroparalysis or any other obvious abnormality. Oral examination revealed a mass measuring 5 cm that extended from the gingiva in the edentulous maxillary molar region to the palatal mucosa. There was no regional lymphadenopathy (Figure 1).

![Figure 1: clinical view](image_url)

craniofacial computed tomography (CT) revealed voluminous oblong mass of 50mm x 45mm extending from the region under the right pterygoid process to the right inner lateral mandibular region. craniofacial magnetic resonance imaging (MRI): Gingivo palatine tumoral process, poorly delineated, with irregular contours, intensely and homogeneously enhanced following gadolinium injection, measuring: 63x55x34mm. Anteriorly, it respects the gingiva opposite the teeth covering the maxillary arch (teeth 15-46). Superiorly, it infiltrates the posterior third of the hard palate, particularly on the right, and extends into the ipsilateral nasal cavity, reducing its patency. Posteriorly, it protrudes into the oropharynx, diminishing its lumen and reaches the long muscle of the left head, without a clear separation margin and without evident signs of infiltration. Inferiorly, it contacts the dorsum of the tongue, again without a clear separation margin and without evident signs of infiltration. Laterally, it infiltrates the parapharyngeal fat and makes contact with the bilateral medial pterygoid muscles, without a separation margin and without evident signs of infiltration. It infiltrates the maxillary dental arch near teeth 16 and 17 on the right and 27 on the left. Right lateral cervical lymph nodes with the largest in chain II A measuring 31x15mm. (Figure 2)
Palatal biopsy done at another center showed the presence of tumor proliferation of monotonous nodular architecture made of massives, and of cystic adenoid formation evolving into a myxoid or hyalin stroma accompanied by a range of central necrosis. Tumor cells have a poorly limited vacuolar clear cytoplasm, and a hyperchromatic rounded or oval angular nucleus. Mitotic activity is not observed. This proliferation infiltrates the mucous membrane and erodes the malpighial epithelium of the surface, in Immunohistochemical examination epithelial component is selectively well highlighted by Pancytokeratin (CK). Myoepithelial component is demonstrated by S100, SMA and p63, The Ki-67 labeling index was 20%. On the basis of these findings, the tumor was diagnosed as epithelial-myoeipithelial carcinoma of minor salivary gland (EMC).

Chest computed tomography (CT) study showed multiple bilateral parenchymal nodules. T4bN2bM1 Although the patient underwent chemotherapy using cisplatin Adriamycin for three cures with clinical stabilization of the process than the patient was blind.

III DISCUSSION:
Epithelial-myoeipithelial carcinoma (EMC) is a rare type of tumor that affects the salivary glands, making up only about 1% of all cases. EMC was first identified as a separate pathological diagnosis in the World Health Organization's classification of salivary gland tumors in 1991. It mainly impacts people in their sixties and seventies, with a slightly higher occurrence in females. Our case is one of the rarest described, arising in a younger patient (46 years old). The rarity of EMC poses significant challenges in both diagnosis and management. These tumors predominantly arise from the parotid gland, However, they can also originate from minor salivary glands located in various anatomical sites such as the palate, buccal mucosa, and base of the tongue. The presentation in the minor salivary glands, as observed in this case involving the gingival palatine region, is particularly rare and may lead to delays in recognition and appropriate intervention. EMCs seem to arise in two different clinical settings: either de novo or in a recurrent pleomorphic adenoma. De novo EMCs arise in normal salivary gland, tend to be more aggressive and have a short clinical history. Our patient presented with de novo palatal swelling since 1 year with lung metastasis.
Most EMCs have a typical biphasic tubular histology and a nodular or multinodular development pattern. The other variations include double-clear EMC, EMC with myoepithelial anaplasia, dedifferentiated EMC, oncocytic EMC, EMC ex pleomorphic adenoma, and sebaceous differentiation. In routine histologic sections, the morphologic variants of myoepithelial cells are clear, spindle, stellate, polygonal, angular, epithelioid and plasmacytoid. Immunohistochemically, epithelial component is selectively well highlighted by all cytokeratins (CK) and epithelial membrane antigen (EMA). Myoepithelial component is demonstrated by S100, smooth muscle actin (SMA), p63 and vimentin. The markers like calponin (CALP), caldesmon (CALD), and smooth muscle myosin heavy chain may be useful tools for identifying myoepithelial cells when myoepithelial cell differentiation is not easily identified on routinely stained sections. In our case, immunohistochemistry was done to highlight the biphasic nature of the tumor, the epithelial component were positive for Pancytokeratin (CK) and The myoepithelial cells were positive for p63, S-100 and SMA. Patients with EMC showing marked cellular pleomorphism, tumor necrosis; angiolymphatic invasion and perineural invasion have a poor prognosis. Epithelial-myoeopithelial cancers are commonly treated with surgical excision to achieve R0 resection. Up to 25% of patients experience lymph node metastases, although kidney, lung, and brain metastases are less common. Approximately 50% of patients with EMCs experience local recurrence. Patients' 5- and 10-year survival rates are approximately 87% and 67.5%, respectively. Treatment options such as chemotherapy and radiotherapy may be considered for patients with advanced disease, positive surgical margin, or surgically unresectable disease. However, there is limited research available on the effectiveness of these therapies. At the moment, there is no established treatment for metastatic myoepithelial carcinoma. Treatment options such as surgical resection of metastases, radiotherapy, and chemotherapy with dacarbazine have been documented in medical literature. DCF (also called TPF) chemotherapy is a regimen combining the CF regimen and a Taxane and has been used for induction chemotherapy for locally advanced or unresectable squamous cell carcinomas of the head and neck. Because improved overall survival has been documented, the TPF regimen has recently been commonly used. In our case, the patient received 3 cycles of cisplatin Adriamycin with stabilization of the process.

**IV CONCLUSION**

This article described a rare case of EMC in the minor salivary. The rarity of EMC makes diagnosis still challenging. There is no consensus on the optimal management of the minor salivary glands EMCs. The best. Longer-term cohort studies would help prevent late diagnosis and offer a better knowledge of how they evolve and how best to be managed. It is consequently necessary to establish clear, uniform guidelines for the treatment of these individuals.

**V REFERENCES**

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