

Epithelial-Myoepithelial Carcinoma of the Parotid Gland

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Abstract

Epithelial-myoepithelial carcinoma (EMC) is an unusual malignant salivary gland tumor. Little is known about the clinical and biological behavior of EMC. There is no consensus for its treatment. It is considered as a low grade malignant tumor. Whatever, it presents a potential of local recurrence and possibility of metastases. We report a 76-year-old patient who presented an epithelial-myoepithelial carcinoma of parotid gland, treated successfully with wide local resection and adjuvant radiotherapy with no signs of recurrence for 3 years.

Keywords: Epithelial-myoepithelial carcinoma, parotid gland, surgery.

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INTRODUCTION

Epithelial-myoepithelial carcinoma (EMC) is a rare malignant tumor accounting about 1% for all salivary gland tumors [3]. It arises most commonly in the parotid gland, but it has also been described in the submandibular gland, minor salivary gland and palate [4]. EMC is composed by a dual cells population: central duct lining cells surrounded by clear cells of myoepithelial origin. In 1972, Donath and al introduce term of epithelial myoepithelial carcinoma [1]. The World Health organization (WHO) recognized EMC as a distinct entity in 1991 [2]. EMC is considered to be a low grade malignant tumor with high potential of recurrence and possibility of metastases. Treatment is mainly surgical [2].

We report a case about a 76-year-old patient treated in our department for an epithelial-moyepitheial carcinoma of parotid gland with no recurrence on 3 years' follow-up period.

CASE REPORT

We present the case of a 76-year-old patient; followed for high blood pressure; admitted in our university hospital for a mass over the left parotid gland. This painless mass had been slowly enlarging for two years. At the physical examination, it appeared as a

bulky mass of 6 cm. It had a firm consistency, was fixed to the skin and was not painful on palpation. There was a facial paralysis stage V of House-Brackmann classification of facial function. The oral cavity examination was normal. There were no palpable cervical lymph nodes on both sides.

A preoperative cervical computed tomography (CT) showed a left superficial parotid lobe tumor. It was well a limited, hypodense, heterogeneous, multi-lobed mass with irregular contours. It was containing micro-calcifications, and increasing heterogeneously after injection of contrast product. There were no cervical lymph nodes on CT either (Figure-1).

Our patient was classified T₄N₀M₀, then the treatment was as follow: a left exofacial parotidectomy was performed without lymph nodes dissection. The extemporaneous examination was inconclusive.

In the final pathology report, two components of the tumor cells including the luminal ductal inner epithelial cells and peripheral outer myoepithelial cells were revealed (Figure-4). The results of biphasic differentiation were confirmed by immunohistochemical staining: positive for S-100 and smooth muscle actin in the surrounding myoepithelial cells, and intense positive for epithelial markers, such

as cytokeratin-7 (CK7) and CD99 in the luminal tumor cells. They are also negative for CD45, CD20, CD3, CD10, bc16, MuM1. The Ki-67 (proliferation index) was less than 1% (Figure-5). Based on these results, the mass was diagnosed as an epithelial-myoepithelial

carcinoma of the parotid gland. Postoperatively, the patient was treated with adjuvant radiotherapy

The follow-up was satisfying for 3 years without signs of recurrence.

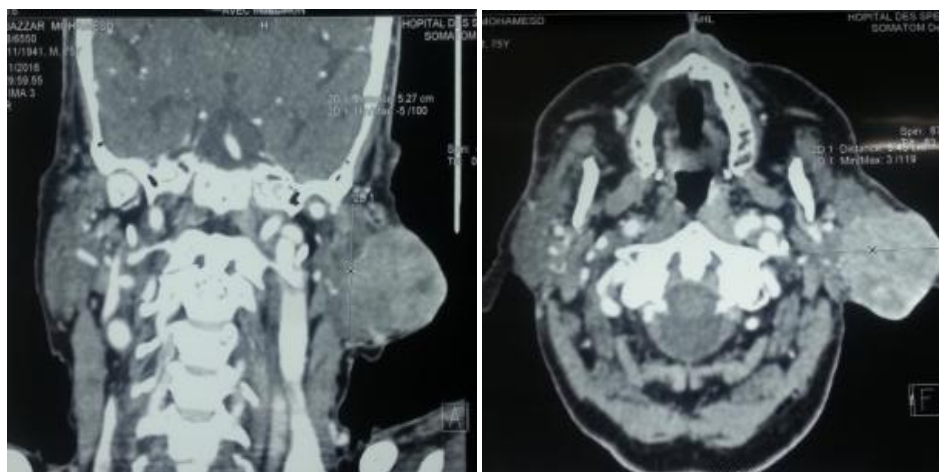


Fig-1: A computed tomography scan shows a non-homogeneously enhancing mass in the left parotid gland



Fig-2: A per-operative Tumor mass (6cm) in the left parotid with involvement of skin



Fig-3: Macroscopic appearance of the resected tumor. The cut surface revealed a homogenous multilobated and gray-white appearance

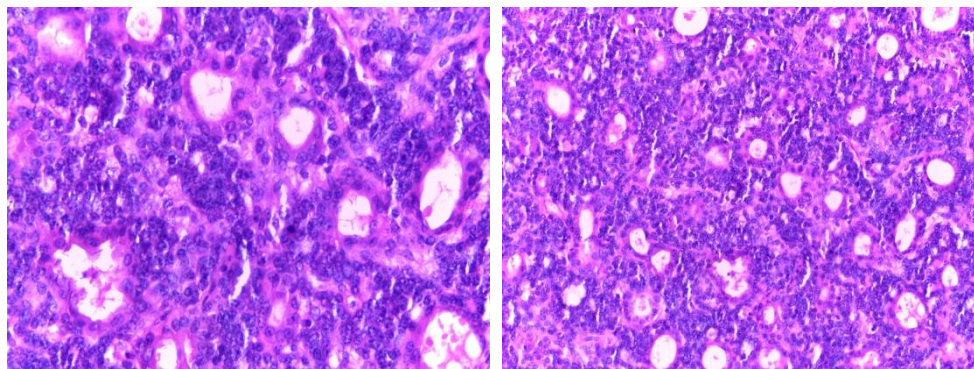


Fig-4: High-power microscopic observation of the hematoxylin-eosin–stained specimen shows a glandular structure consisting of inner layer (eosinophilic cuboidal cells) and outer layer (clear cells)

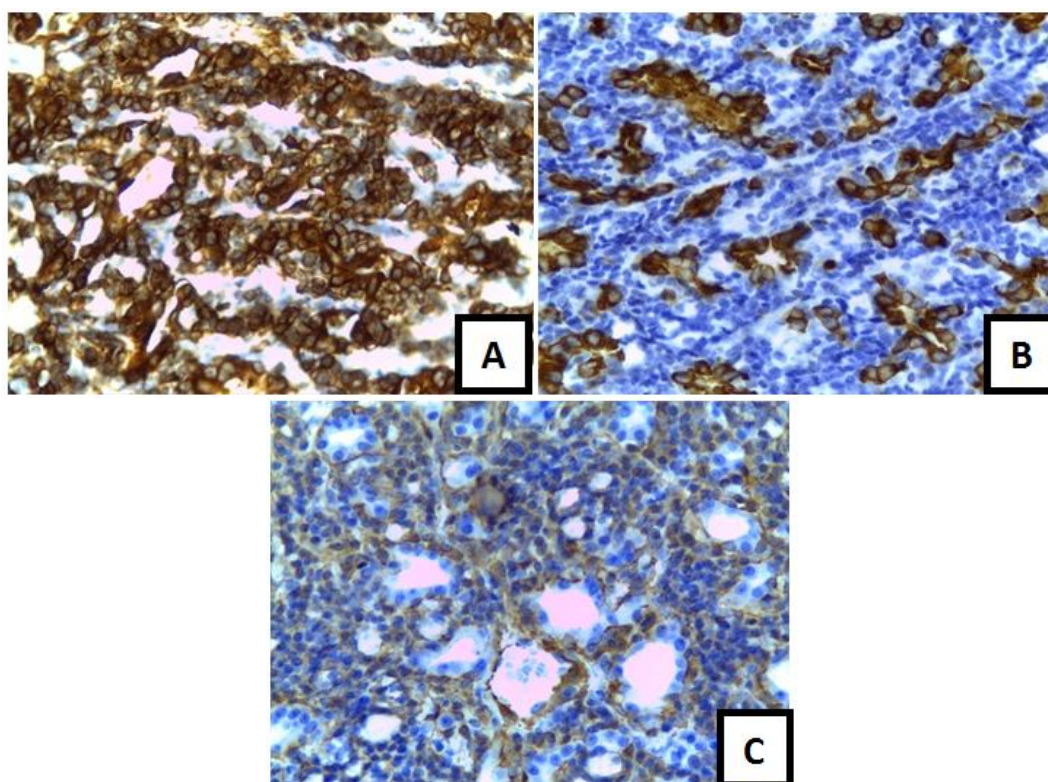


Fig-5: Immunohistochemical staining of sections: The inner-layer cells are positive for cytokeratin AE1/AE3 (A) and CK7 (B) as epithelial markers. The outer-layer cells are positive for actin (C) as a myoepithelial marker

DISCUSSION

Epithelial myoepithelial carcinoma (EMC) is a rare low grade malignant tumor, firstly described in 1972 by Donath *et al.*, [1]. It was recognized as a distinct entity by the World Health Organization in 1991 [2, 3].

EMC accounts about 1% of all salivary gland tumors. It arises most commonly in the parotid gland (60 to 83%) [3, 4] followed by the submandibular gland (9.8 to 13.01%) [5, 4] and occasionally occurs in minor salivary gland and palate [5].

The peak of incidence appears in the sixth and seventh decades, with a female predominance (57.2 to 62.2%) [3, 4].

Clinical presentation of EMC is similar to other parotid gland tumors. It may be presented by an asymptomatic painless slow growing mass suddenly increasing in size. Pain and facial paralysis can be associated. Radiological appearance (computed tomography (CT) and magnetic resonance imaging (MRI)) of this tumor is non-specific [6, 7].

According to literature, the majority of the patients had T1 (21.8 to 38.35%) or T2 (21.6 to 35.33%) tumors. The nodal status of 3.76 to 4.2% of patients was N+. The rate of distant metastases at diagnosis is about 2.6% [4, 5].

Table: Patient tumor (T), nodal (N), and metastasis (M) characteristics:

| | Vazquez 2015 [4] | Gore 2018 [5] |
|------------------|------------------|---------------|
| T classification | | |
| T1 | 38.35% | 21.8% |
| T2 | 32.33% | 21.6% |
| T3 | 16.54% | 9.8% |
| T4 | 5.26% | 0.4% |
| Tx | 7.52% | 6.0% |
| N classification | | |
| N0 | 90.23% | 57.5% |
| N1 | 3.76% | 3.8% |
| N2 | - | 0.4% |
| N3 | - | 0.2% |
| Nx | 6.02% | 4.7% |
| M classification | | |
| M0 | 93.98% | 61.1% |
| M1 | - | 2.6% |
| Mx | 6.02% | 5.2% |

Histopathologically, EMC is characterized by a glandular structure consisting of a double layer of inner and outer layers: the inner layer consists of intercalated duct epithelium-like cells; whereas the outer layer consists of clear neoplastic myoepithelial cells often predominate in number. [8]

The morphologic appearance of the neoplasm may mimic other clear cell salivary tumors as: clear cell carcinoma, acinar cell carcinoma, mucoepidermoid carcinoma, metastatic renal cell carcinoma, myoepithelial carcinoma, clear cell oncocytoma and pleomorphic adenoma. [8]

The diagnosis can be determined by immunohistochemical findings, because only inner-layer cells are positive for epithelial markers (EMA+, Keratin+) pancytokeratine KL1 and only outer-layer cells are positive for myoepithelial markers (S100 protein +, Smooth muscle actin +, vimentin) [8].

Until now, there is no consensus about the treatment of EMC. Whatever, surgery appears to be the primary treatment of the EMC of parotid gland. Either exofacial or total parotidectomy is indicated to obtain a complete resection with negative margins even it is possible [3-5].

Based on nodal status, surgery was including neck dissections in 53.33% of the patients with N0 diseases, 80% with N+ and 37.5% with Nx [4].

Adjuvant radiotherapy is also required. Its survival benefit still unclear, but it may be effective in preventing local recurrence [4]. The efficacy of adjuvant chemotherapy has not been established [9].

Indeed, the low-grade malignancy of this tumor, the recurrence rate was 36.3% (12/33 patient), with a median disease-free survival DFS of 11.34 years

[3]. Overall disease-specific survival was about 91.3%, 90.2% and 80.7% at 60, 120 and 180 months [4].

Different studies showed that overall survival was significantly affected by TNM stage (presence of nodal/distant metastases), surgical versus nonsurgical treatment, race and age at diagnosis [5].

CONCLUSION

Epithelial myoepithelial carcinoma is an unusual salivary gland tumor. The usual treatment is wide surgical resection with negative soft-tissue margins. The role of radiation and chemotherapy is still controversial. EMC is a low-grade malignant tumor with a potential of local recurrence and possibility of metastases. Therefore, more studies are needed to better understand the role of neck dissection and adjuvant therapy to affect survival and recurrence.

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