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Case Report

Giant Myoepithelial Carcinoma Ex-Pleomorphic Adenoma of the Parotid Gland: A Case Report

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Abstract

Myoepithelial carcinoma accounts for less than 1% of salivary gland malignant tumors It may develop de novo or, in approximately 50% of the cases, from a pleomorphic adenoma. Here we describe a rare case of 56 -year-old female, who presented with painful, rapidly enlarging and swelling mass located in the right parotid gland.

Keywords: *Myoepithelial Carcinoma; Pleomorphic Adenoma; Parotid Gland*

Introduction

Myoepithelial carcinoma accounts for less than 1% of salivary gland malignant tumors [1]. It may develop *de novo* or, in about half of the cases, from a pleomorphic adenoma [2]. In immunohistochemistry, they very often express smooth muscle actin (81.8% of cases) and the S100 protein (94.6% of cases), often vimentin (75% of cases) [3]. Here, we report a case of Myoepithelial carcinoma ex-pleomorphic adenoma of the parotid gland in a 56-year-old female.

Case Report

A 56-year-old female with a past history of a pleomorphic adenoma of the right parotid gland in whom the surgical indication of parotidectomy was retained but the patient was lost to follow-up. After an evolution of 8 years, the patient consulted our department due to rapid increase in the mass volume with swelling and unbearable pain, the patient accused a concomitant diplopia associated to right retro-orbital pain.

On physical examination, a firm non-tender fixed mass of (15 × 16 cm) was found on the right parotid gland extending to the retroparotid area, taking the ascending branch of the mandible as well as the mandibular angle and overflowing on the oropharynx with no ipsilateral facial paralysis (Figure 1).

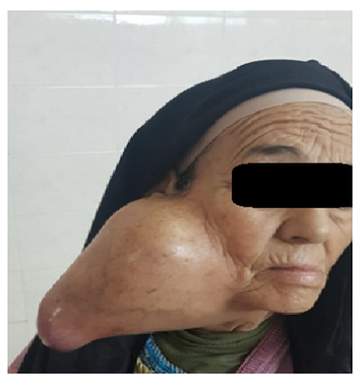

Figure 1: *Front view of the patient showing the mass of the right parotid gland extending to the mandibular and cervical areas.*

A computerized tomography (CT) scan of the head neck was indicative of a heterogeneous, moderately enhancing soft tissue mass of the right parotid gland with important necrosis areas and calcifications (red arrow) with invasion of parapharyngeal space (red star), the oropharynx (blue star) and a repression of vessels which remain permeable (blue arrow). No visible lysis of the ascending branch of the mandible (yellow star) was noticed.

The CT did also confirm an exophthalmia with a retro-orbital process having the same density as the parotid mass and infiltrating the right optic nerve (black arrow) (Figure 2).

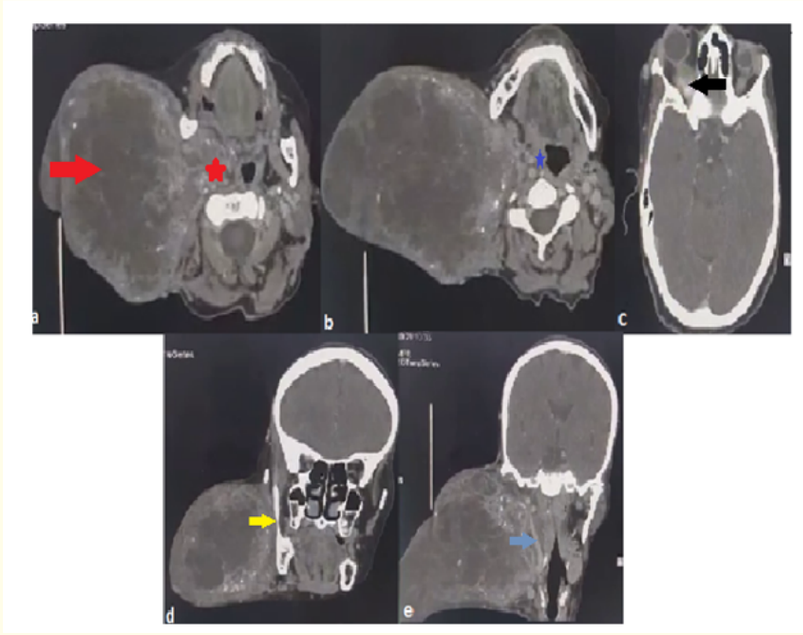

Figure 2: *Contrast enhanced axial (a, b, c) and coronal (d, e) soft tissue window CT showing the mass involving the right parotid gland with an infiltration of parapharyngeal space and an extension to the retro-orbital space.*

Owing to the requirement for obtaining a histopathological diagnosis, incisional biopsy was performed under local anesthesia. Histopathological examination showed a poorly differentiated tumor process requiring phenotyping immunostaining to support the diagnosis. The immunohistochemistry showed in tumor cells, positive CK7, positive CK19. At the level of a tumor contingent a positive P63, positive PS100 and positive AML. Were found also in tumor cells positive EMA and positive ACE. On the basis of these findings, a final diagnosis of carcinoma ex-pleomorphic adenoma of the parotid gland was established (Figure 3).

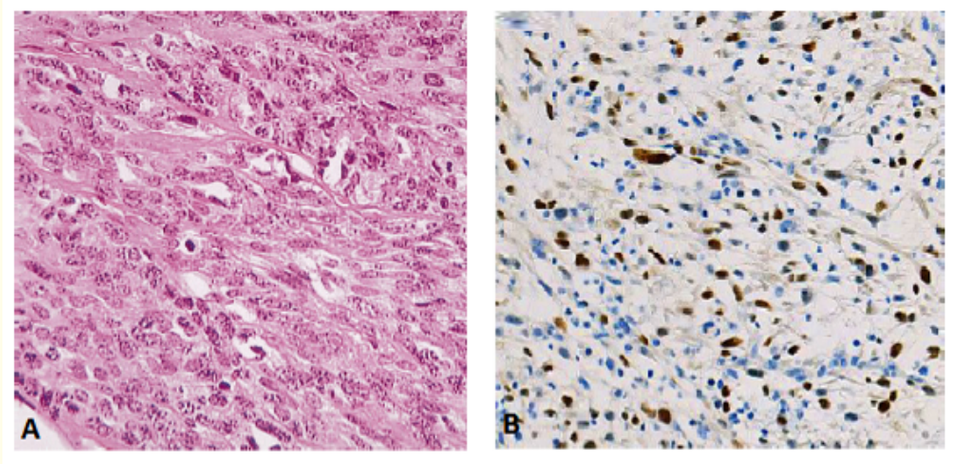

Figure 3: *Proliferation of cells with ovoid/elongated nuclei, with existence of figures of mitosis hematoxylin eosin x400 (A) spindle-shaped tumor cells express p63, in favor of the myoepithelial origin of the proliferation (B).*

It was decided to complete with a brain and orbital MRI in order to assess the process which is infiltrating the right optic nerve and responsible of exophthalmia. Being claustrophobic, in addition to the difficulty in lying down for a long time, the MRI was stopped several times and subsequently refused by the patient which refused also any surgical procedure and left our establishment against medical advice.

Discussion

Myoepithelial carcinoma accounts for less than 1% of salivary gland malignant tumors. It may develop *de novo* or, in approximately 50% of the cases, from a pleomorphic adenoma [1].

The pleomorphic adenoma is the most common tumor salivary glands. Although benign, she presents a potential for recurrence and malignant transformation [2].

The once widely used term malignant mixed tumor, groups together three types:

- Carcinoma ex-pleomorphic adenoma.
- Carcinosarcoma or true mixed malignant tumor.
- The metastasizing pleomorphic adenoma [3].

The carcinoma ex-adenoma pleomorphic is the most common.

Its frequency is variable according to the series. It represents approximately 12% of malignant salivary gland tumors and occur develops in about 6% of pleomorphic adenomas [4].

By reviewing a series of 518 epithelial tumors of the salivary glands among which 282 pleomorphic adenomas, found 48 carcinomas ex-pleomorphic adenomas mainly classified as undifferentiated carcinomas or adenocarcinomas, but no cases of myoepithelial carcinoma has been reported in this series.

Lewis., *et al*. reported 73 carcinomas pleomorphic ex-adenomas among which, only two myoepithelial carcinomas. We believe that myoepithelial carcinoma is certainly rare but many cases reported, in particular in old series, of carcinomas undifferentiated or sarcomatoid carcinoma could correspond to myoepithelial carcinomas [5].

Clinically, this tumor manifests as a slowly growing, painless mass. The radiological appearance of this tumor is variable, which can rarely be cystic, like the right parotid localization in our case, or solid, like the left parotid localization in our case [3].

On histopathological examination, the main characteristic of this tumor is the presence of dual composition. On the one hand, there is myoepithelial cells, organized in spans or lobules and characterized by a clear cytoplasm dyed purple by PAS staining and an irregular large nucleus. However, the positivity of PAS is not specific to CEM since it is also observed in acinar carcinoma, in which there is no myoepithelial contingent. In immunohistochemistry, they very often express smooth muscle actin (81.8% of cases) and the S100 protein (94.6% of cases), often vimentin (75% of cases) and, to a variable extent, GFAP (40, 0% of cases) [6]. On the other hand, it is associated with an epithelial part, sometimes very minor, organized in tubes or rows, the cells of which have a smaller and more regular nucleus, without atypia and an acidophilic cytoplasm. Mitoses are rare. These luminal epithelial cells consistently express cytokeratin (pancytokeratin KL1, cytokeratin 7), EMA, and CEA. Clear myoepithelial cells organize around acidophilic cells, together forming nodules well bounded by hyaline fibrosis. Finally, this tumor shows areas of focal infiltration and/or perinervous sheaths on the periphery [7].

When the myoepithelial nature of a tumor proliferation is confirmed, distinguish a myoepithelioma from a Low-grade myoepithelial carcinoma can be difficult. For Savera., *et al*. [8] the only sufficient criterion is infiltration tumor of adjacent tissues. Currently, this criterion seems to be accepted by the majority of authors [2,6,7]. The case that we report was considered to be carcinoma to a proliferation of ovoid or elongated cells without particular arrangement and above all without epithelial differentiation subsidiary. The cells were monomorphic without atypia notable and mitotic activity was estimated at five mitoses by ten consecutive microscopic fields examined at high magnification [8].

Indeed, myoepithelial carcinomas considered histologically low-grade (minimal atypia and rare mitosis) have rapidly metastasized, while others of high rank histologic, did not present with recurrence or metastasis after a fairly long follow-up [8]. The origin of the myoepithelial carcinoma could influence its evolution. Di Palma and Guzzo [8] consider that myoepithelial carcinoma *de novo* would be more aggressive than myoepithelial carcinoma ex-pleomorphic adenoma, but other studies have not confirm this hypothesis [7,8]. Unfortunately, we were unable to follow the evolution of this patient since she refused any therapeutic intervention and was subsequently lost to follow-up.

Conclusion

Invasive carcinomas developing in pleomorphic adenoma are known to be highly aggressive neoplasms but myoepithelial carcinomas ex-pleomorphic adenoma seem to have a better prognosis.

Bibliography

1. Barnes L., *et al*. "Pathology and genetics of head and neck tumors". Lyon: IARC Press (2005).

2. Seethala RR., *et al*. "Epithelial-myoepithelial carcinoma: a review of the clinicopathologic spectrum and immunophenotypic characteristics in 61 tumors of the salivary glands and upper aerodigestive tract". *The American Journal of Surgical Pathology* 31 (2007): 44-57.

3. Nikitakis NG., *et al*. "Immunohistochemical expression of cytokeratins 7 and 20 in malignant salivary gland tumors". *Modern Pathology* 17 (2004): 407-415.

4. Yu GY., *et al*. "Multiple primary tumours of the parotid gland". *International Journal of Oral and Maxillofacial Surgery* 33 (2004): 531-534.

5. Nagao K., *et al*. "Salivary gland malignant myoepithelioma: a clinicopathologic and immunohistochemical study of ten cases". *Cancer* 83 (1998): 1292-1299.

6. Chhieng DC and Paulino AF. "Cytology of myoepithelial carcinoma of the salivary gland". *Cancer* 96 (2002): 32-36.

7. Suba ZS., *et al*. "Malignant myoepithelioma. Clinicopathological and immunohistochemical characteristics". *International Journal of Oral and Maxillofacial Surgery* 32 (2003): 339-341.

8. Di Palma S and Guzzo M. "Malignant myoepithelioma of salivary glands: clinicopathological features of ten cases". *Virchows Archiv A Pathological Anatomy and Histology* 423 (1993): 389-396.

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