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A Huge Myoepithelial Carcinoma of Parotid Gland in 99 Years Old Patient

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Abstract

Myoepithelial carcinoma, also named malignant myoepithelioma or epithelial – myoepithelial carcinoma, is an unusual neoplasm arising from neoplastic myoepithelial cells. This tumor accounts for 0, 1–0, 45% of all salivary tumors; these neoplasms can involve both major and minor salivary glands.

Myoepithelial carcinomas generally occur in adults or elderly. The described cases in literature have an average age of 52, 5 years, with a range from 7 to 81 years.

This report shows a clinical case of a centenarian woman with a huge myoepithelial carcinoma of the right parotid gland.

Introduction

Myoepithelial tumors salivary glands are rare, comprising about 1–1.5% of all salivary gland neoplasms [1]. The most of myoepithelial neoplasms are benign; the malignant forms represent just 0, 1-0, 45% of all salivary gland tumors [2]. Approximately, 29-82% of the salivary gland myoepithelial carcinomas involve the parotid gland; nevetheless, these malignant lesions are also reported in palate and submandibular gland [2].

Myoepithelial carcinoma was first described by Donath et al. in 1972 [3,4]. Instead, myoepithelial carcinoma of the salivary gland was officially classified as a distinct clinicopathological state by World Health Organization (WHO) in 1991 [5,6].

Myoepithelial carcinoma is also named malignant myoepithelioma or epithelial-myoepithelial carcinoma. It corresponds to malignant variant of myoepithelioma [1].

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The described cases in literature have an average age of 52, 5 years, with a range from 7 to 81 years. No case of older patient is reported in international literature. Males accounted for 42, 6% and females accounted for 57, 4% [7,8].

Malignant transformation of a benign myoepithelioma can occur in recurrent cases. However, malignant myoepithelioma can also arise de novo. Malignant myoepitheliomas of parotid glands show as a slow growing and insidious swelling [9]. Diagnosis is usually based on clinic examination, radiological exam and histological analysis.

Microscopically, myoepithelial carcinoma is defined as a tumour composed of myoepithelial cells with an expansive, nodular, lobulated pattern featured by hypocellular and hypercellular zonal distribution. Its histological structure consisting of spindle, epithelioid, plasmacytoid or clear cytoplasmic features. A malignant myoepithelioma can be charachterized by one or a mixture of these cell types and a changeable stromal frame [10].

Histopathologically, the salivary gland tumours in which the ducts comprise less than 5% of the sections are classified as myoepitheliomas. Also, a tumor arising from myoepithelial cells does not present a chondroid or an osteoid formation [11].

Myoepithelial cells often express cytokeratins and vimentin markers in immunohistochemistry. Vimentin is reported to be positive in neoplastic myoepithelial cells and negative in normal myoepithelial cells [12]. Anyhow, myoepithelial cells express myoepithelial markers, such as SMA, p63 and calponin.

Cellular pleomorphism, high mitotic activity and necrosis are important cytological features. However, evidence of infiltrative and destructive growth, with perineural invasion, are necessary for the diagnosis and to distinguish myoepithelial carcinoma from benign myoepithelial tumors [3,10–13].

This report describes a case of a 99-year-old female affected by a huge malignant myoepithelioma of the right parotid gland.

Case report

A 99-years old female patient was referred for evaluation of a 20-year history enlarging mass over the right side of face at Maxillofacial Surgery Unit of San Camillo Hospital in Rome. Her medical history revealed a 30 years pharmacologically controlled hypertension and she never smoked cigarettes. The first presentation of the lesion was reported to date back to about 20 years before. The swelling was initially small in size and had progressively increased with time to attain the actual size. As clear, the elderly woman has been neglected over the years. Despite massive enlargement, the woman's family reported that previous medical consultations expressed disagreement with the surgery because of her age. After many years the family decided to turn to our hospital structure.

Extra-oral clinical examination highlighted a marked facial asymmetry caused by a huge and discomfort mass arising from right parotid gland (Figure 1 & 2). The skin overlying the surface of the mass was flushed with engorged vessels. On palpation, the swelling was solid in consistency, non pulsatile, and was not fixed to deep structures. The temperature over the swelling was normal.

Facial nerve presented a minimal stupor of right marginalis mandibular branch, probably caused by stretching due to the huge mass weight; so, the right lip appeared distorted towards the healthy side. Physical examination of the neck shows no cervical lymphadenopathy. Eye movements were normal on examination. Intraoral clinical examination had no significant findings; the patient was edentulous.

A clinical diagnosis of benign or malignant tumor of the right parotid gland was taken into consideration. Unfortunately, we could not carry out any radiographic investigation nor an incisional biopsy because patient was no collaborative. So, the preoperative evaluation was based only on anamnesis and clinical examination.

Patient underwent a collegial anesthesiological videat and a presurgical geriatric evaluation. So, after obtaining the informed consent of the patient and of her relatives, we planned the surgical operation.

The patient underwent total parotidectomy with preservation of the facial nerve. Neck dissection was not performed because of advanced age of patient and negative clinical examination. The peripheral nerve branches were identified and preserved following a retrograde approach (Figure 3 & 4). Excision was complete (Figure 5) and the mass was sent to the histology department (Figure 6). Postoperatively facial nerve function was the same as in preoperative.

Histologic examination revealed a myoephitelial carcinoma. The weight of the excised mass was 5.2 kilograms.

Microscopically, it was composed enterily by myoepithelial cells, with an expansive, nodular, lobulated, pattern and hypocellular and hypercellular zonal distribution, with abundant hyaline stroma. It can be grouped in spindle cell pattern (Figure7&8).

The patient recovered from the surgery without sistemic complications or surgical ones and her facial nerve was functioning well. The patient was periodically followed up for 6 months and no recurrences or complications were observed during this period (Figure 9&10).

This is the first case of a centenarian patient underwent to surgical operaton of myepithelioma excision described in the literature.

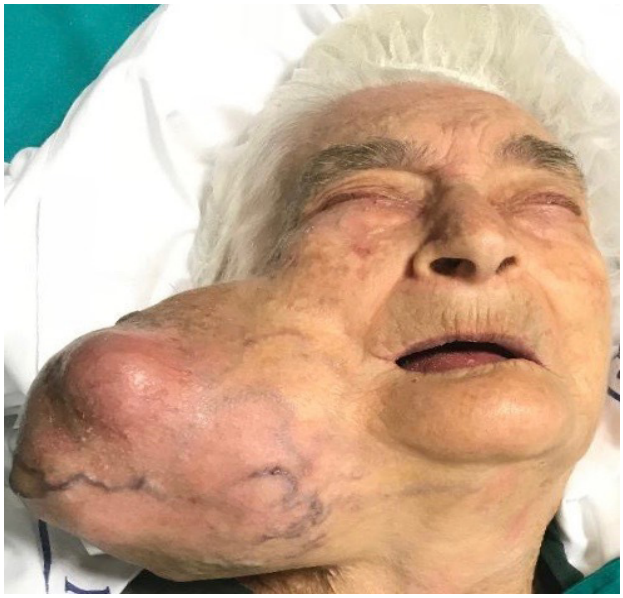

Figure 1: Right parotd gland mass in frontal view.

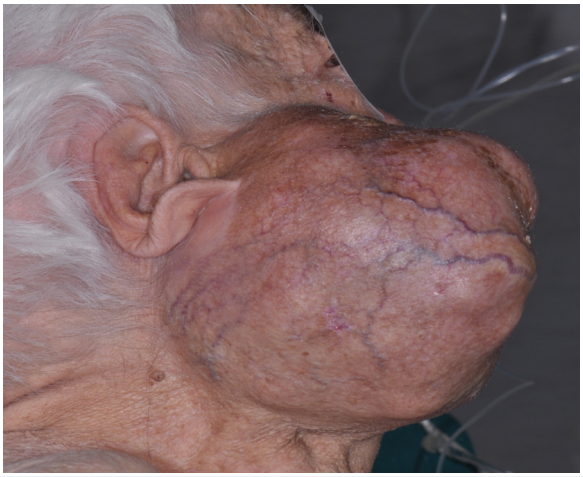

Figure 2: Right parotid gland mass in lateral view.

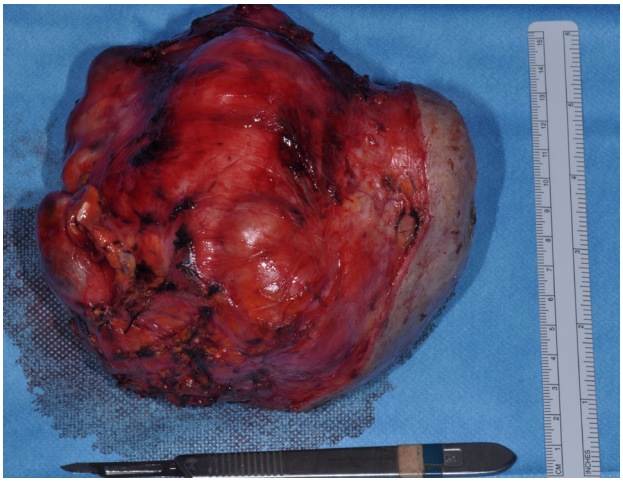

Figure 6: Tumor removed.

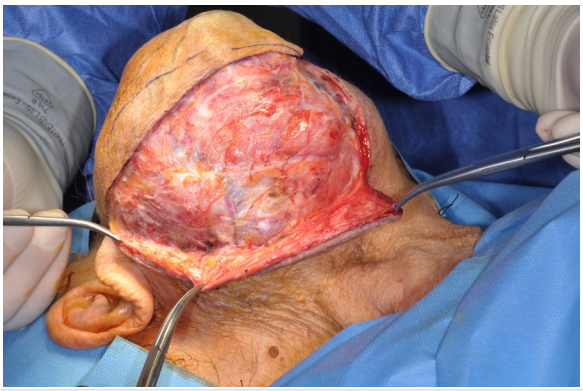

Figure 3: Intraoperative image of surgical approach.

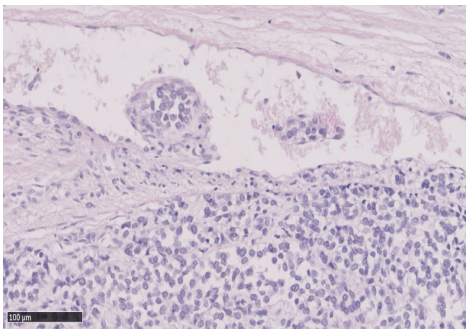

Figure 7: Microscopic image.

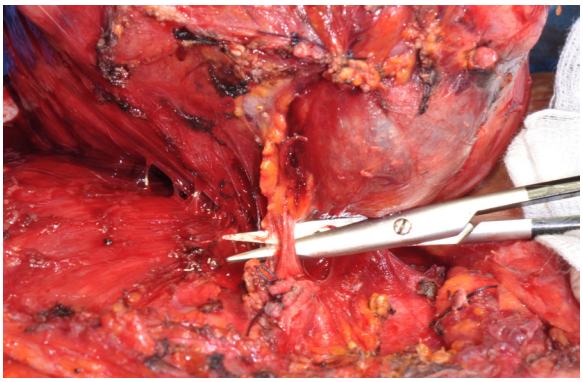

Figure 4: Intraoperative image of facial nerve preservation.

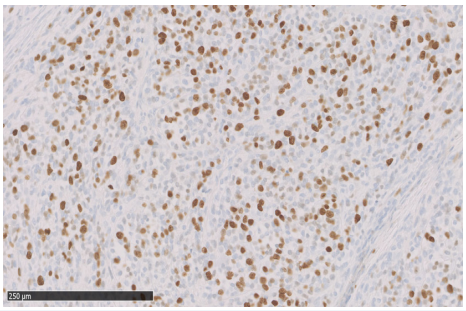

Figure 8: Microscopic image.

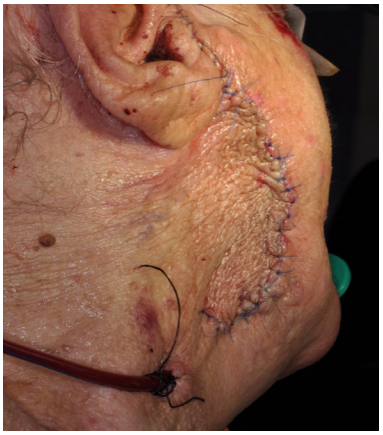

Figure 5: Plastic tissue after tumor removal.

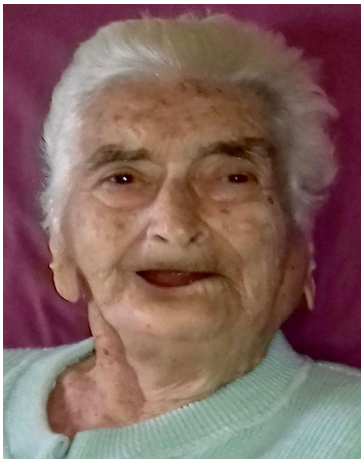

Figure 9: Postoperative image in frontal view.

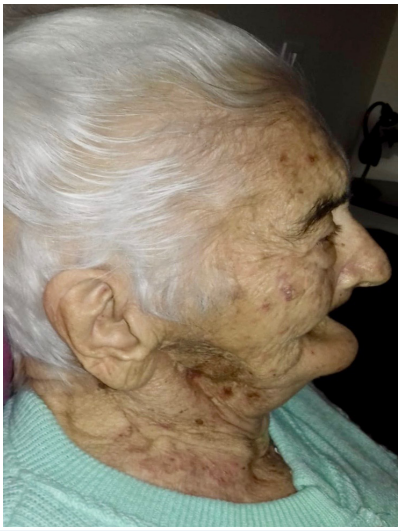

Figure 10: Postoperative image in lateral view.

Discussion

Parotid gland neoplasms represent 2-3% of head and neck tumors and 0.6% of all tumors in the body. Pleomorphic adenoma is the most frequent salivary glands benign tumor (70-80% of benign parotid glands tumors), followed by Warthin tumor. Malignant tumors are comparatively rare; mucoepidermoid carcinoma is the most represented in this category [14].

Myoepithelial cells of the salivary gland were first described by Zimmerman in 1898. In 1943, Sheldon first classified myoepitheliomas as tumors of salivary gland in a review of 57 mixed tumors of the salivary glands [15]. Myoepithelial carcinoma was first described by Donath et al. in 1972. World Health Organization (WHO) recognized myoepithelioma and myoepithelial carcinoma as histologically distinct entities in 1991 [3,5,6,12].

Myoepithelial carcinoma, also named malignant myoepithelioma or epithelial-myoepithelial carcinoma, is an unusual malignant tumor arising from neoplastic myoepithelial cells localized between the basement membrane and the basal plasma membrane of acinar cells. They are characterized by various cellular elements including smooth muscle actin, myosin, and intermediate laments [14,16].

Malignant myoepitheliomas only account for approximately 0, 1-0, 45% of all salivary neoplasms [2].

The most frequent location of malignant myoepithelioma of the head and neck is the parotid gland (29-82%). Rare cases of palate and submandibular gland malignant myoepitheliomas have been described. Otherwise, neoplastic myoepithelial cells can be found in salivary gland and other exocrine glands, except pancreas [17,18].

Microscopic anatomy of myoepitheliomas provides four subtypes:

- Spindle cell pattern: It is the most frequent type (65%) and it is characterized by a proliferation of spindle-shaped cells with eosinophilic cytoplasm and interlacing fascicles;
- Plasmacytoid pattern: It represents 20% of cases and it consists of round cells and hyaline cytoplasm with abundant eosinophilic cytoplasm;
- Epithelioid: It is based on nests or cords of polygonal cells, that are characterized by centrally located nuclei and a variable amount of eosinophilic cytoplasm;
- Clear: Its feature is a cell structure with polygonal cells and abundant optically clear cytoplasm, containing glycogen or missing mucin [15].

Myoepithelioma cells have been usually shown to be positive for cytokeratins as SMA, calponin, p63, CK 5/6, Cam 5.2, Ki-67, AE1/AE3, CK-7, and CK-14 and vimentin markers in immunohistochemistry. Vimentin is negative in normale myoepithelial cells and positive in neoplastic ones. Myoepithelial cells often express GFAP (glial fibrillary acidic protein) and S100 protein [19].

Myoepithelial carcinomas generally occur in adults. The average ave is 52,5 years, with a range of 7-81 years: no case of older patient is reported in international literature. These tumors are prevalent in women (57,4%) [7,8,20].

Malignant myoepitheliomas are low-grade malignant neoplasms and they are characterized by an insidious and slow growing swelling. Other symptoms can be pain (10-29%), facial palsy (10-15%), lingual or lip numbness (5-8%) or dysphagia (1-3%). Conversely to benign form, it can invade the adjiacent structures because is locally invasive. These tumors are chacacterized as low-grade malignant and they have a high recurrence rate (35-40%); nevertheless, the mortality is quite low. Myoepithelial carcinomas can arise de novo; however, they may occur from previously benign myoepitheliomas. In the largest clinical series, 75% of the malignant myoepitheliomas arose de novo and 25% originated from a previously benign tumour [2].

Myoepithelial carcinoma comprises approximately 10% of all myoepithelial neoplasms [9].

Otherwise, the rate of distant metastases seems to be high in some studies, but other works reporting a locally aggressive progression with low rate of distant metastases.

If clinically possible, the diagnosis is usually made with a combination of clinic examination, radiologic investigation and histological analysis.

Fine-needle aspiration (FNA is a minimally invasive procedure, but often in myoepithelial tumors it is not conclusive because it can often mistakenly testify for a benign form [3]. So, immunohistochemistry is very important in diagnosis.

The CT and MR imaging findings of myoepithelioma are not specific. Myoepitheliomas are represented by well-circumscribed, unilocular or multicoluar areas, typical of solid tumors. Most of these tumors are located in the superficial lobe of the parotid gland and they exhibited homogeneous signal intensity or density on either the pre-contrast or contrast-enhanced MRI and CT scan [21,22].

Because of rarity of myoepithelial carcinoma there is not a standard treatment guideline. Surgical approach is the most used treatment, and it is the mainstay in the management of myoepithelial carcinoma. Where possible, the aim should be to achieve widely clear surgical margins in view of the tumour's propensity to local recurrence and aggressive behaviour. Instead, some of the reported cases described the use of adjuvant radiotherapy [16,20,23].

Neck dissection should be considered in cases of lymph node positivity along with radiotherapy and/or chemotherapy in patients with positive surgical margins or surgically unresectable disease; nevertheless, there was not many studies of these therapies [2,13,24,25].

However, the risk of neck node involvement is not well defined because to the wide number of variants of myoepithelial carcinoma. From published data, nodal involvement rate ranges between 0 and 41 % [2,26,27]. Chen et al. reported a rare case of a patient with a primary base of tongue epithelial-myoepithelial carcinoma with neck metastases and multiple lung nodules.

In cases of local recurrence following superficial parotidectomy, total parotidectomy and radical neck dissection are appropriate. The use of palliative chemotherapy has been described in unresectable recurrences with no effectiveness. Super selective intraarterial infusion of high dose of Cisplatin along with radiotherapy is also reported after several recurrences, with a complete response after 7 months [30].

Conclusion

Although myoepithelial carcinoma of the parotid gland is a rare salivary gland tumor, it should always be a part of the differential diagnosis [28,29]. Treatment of malignant myoepitheliomas consist of complete surgical removal, nevertheless it is difficult to draw conclusions about the risk of nodal involvement from published data. The authors consider this case as unique both for the rarity of the tumor, and especially for the centenary age of the patient.

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