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Case Report

A rare case of an aggressive submandibular gland cancer revealed by a parietal-pleural metastasis: Case report and literature review

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Abstract:

Around 10% of all salivary gland tumors are located in the sub-mandibular gland, with a high rate of malignant tumors. Mucoepidermoid carcinoma "MCE" represent 13 % of all sub-mandibular gland tumor and is reported to have an aggressive behavior and the most difficult to treat.

The purpose of this article is to report a case of 60-year-old patient diagnosed with a high grade mucoepidermoid carcinoma of sub-mandibular gland revealed by a parietal-pleural metastasis in whom a surgical, radiotherapy and chemotherapy treatment has been adapted, and by a literature review we will describe the clinical, histological, therapeutic features and the prognostic factors of this rare and aggressive histological type of sub-mandibular gland.

Keywords: submandibular gland; metastasis; mucoepidermoid carcinoma; case report

Introduction

Salivary gland carcinomas represent less than 6% of malignant tumors of head and neck [1].

Parotid is the most common localization of these tumors among the major salivary glands, and palate is the most frequent site for minor salivary glands tumors.

Around 10% of all salivary gland tumors are located in the sub-mandibular gland, with a high rate of malignant tumors.

72% of submandibular gland tumors were diagnosed as adenoid cystic carcinoma, while mucoepidermoid carcinoma "MEC" represented 13% [2].

Complete surgical resection is the gold standard for MEC. The protocols for adjuvant radiotherapy or chemotherapy are still matter of discussion.

We report a case of aggressive high grade MEC of sub mandibular gland reveled by a

parietal-pleural metastasis.

Case report:

we report a case of a 60-year-old patient, chronic smoker of 40 packs a year, with no particular medical history, referred by a thoracic surgeon to our department for management of a right submandibular mass. The patient initially consulted for a right thoracic pain evolving for 1 month in a context of altered general condition, in whom imaging revealed a parietal-pleural lytic process of the anterior arch of the first rib of neoplastic appearance and undetermined primary or secondary origin (Figure 1).

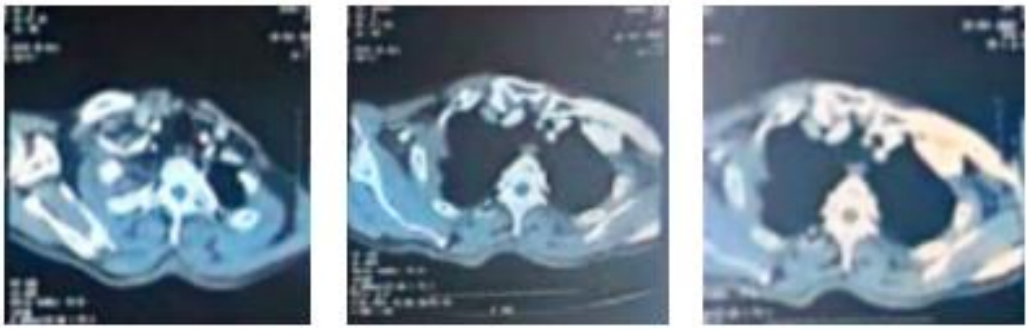

Figure 1: Cervicothoracic CT scan in axial section showing a parietal-pleural lytic process of the anterior arch of the first rib.

For the past 2 months, the patient had also presented a painless right submandibular mass that was progressively enlarging, and which presented on clinical examination as a painless mass of the right submandibular gland, of stony consistency, not very mobile to the deep plane, approximately 5.5 cm long, with normal skin and no cervical adenopathies associated (Figure 2).

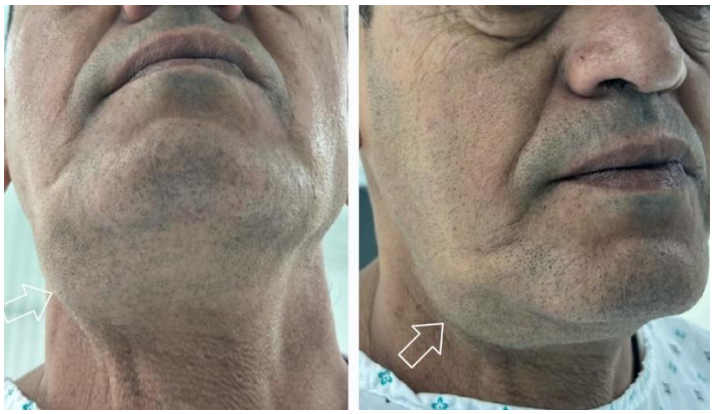

Figure 2: Images of the patient showing the right submandibular mass.

An extension work-up was performed:

Cervico-facial MRI revealed a well-limited lesional process with irregular contours at the expense of the right sub-mandibular gland, isosignal T2, discrete hyposignal T1, not fading in FAT SAT without associated cervical adenopathy or facing mandibular bone lysis (Figure 3,4).

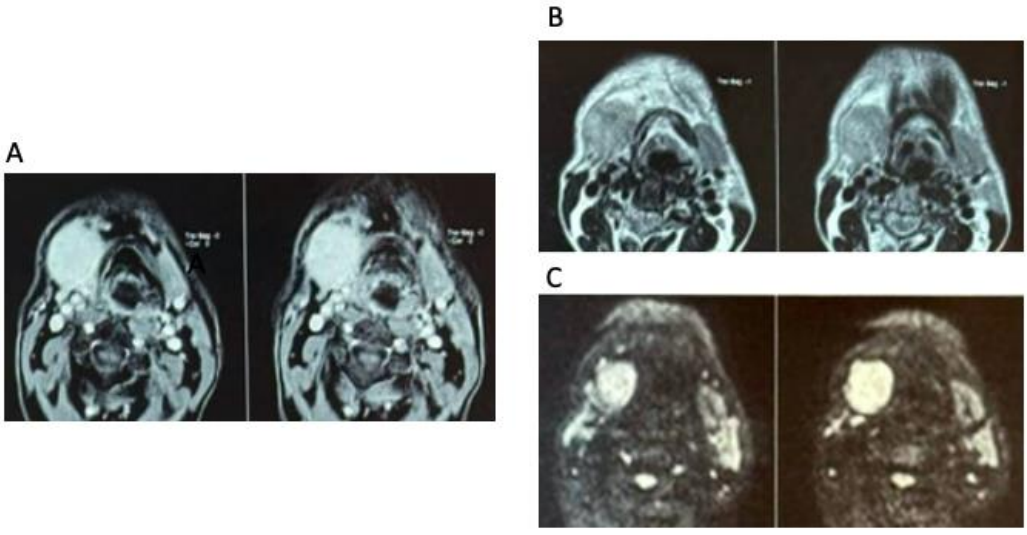

Figure 3: IMR in axial section showing the submandibular gland tumor.

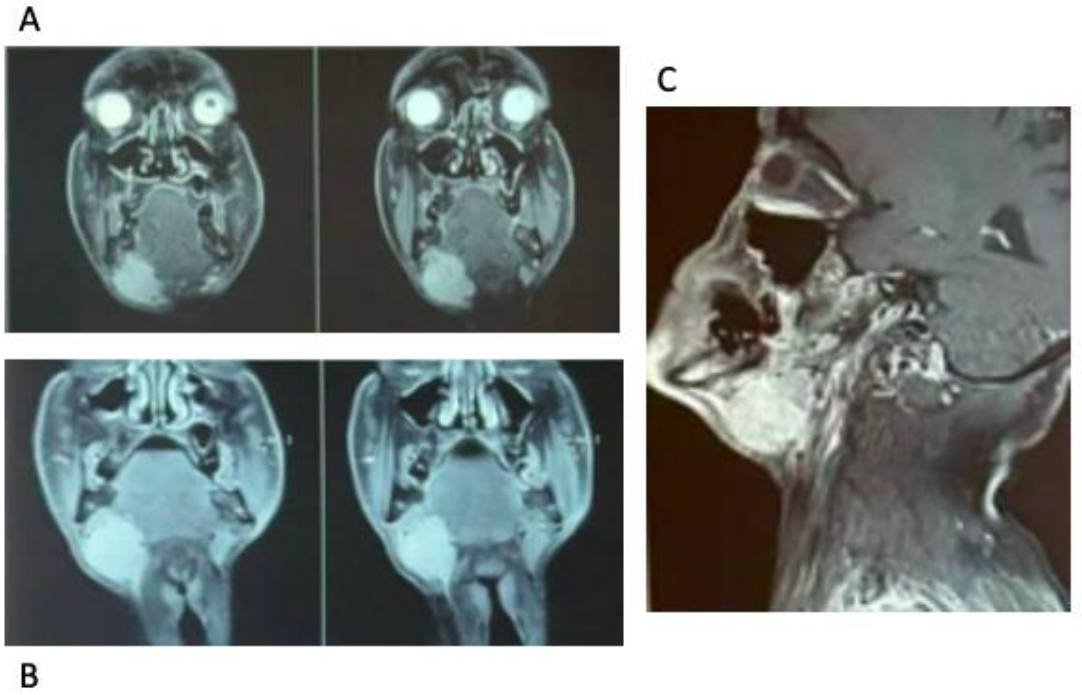

Figure 4: IMR in coronal and sagittal sections showing the submandibular gland tumor.

The patient underwent a right submandibulecto my under general anesthesia (Figure 5),

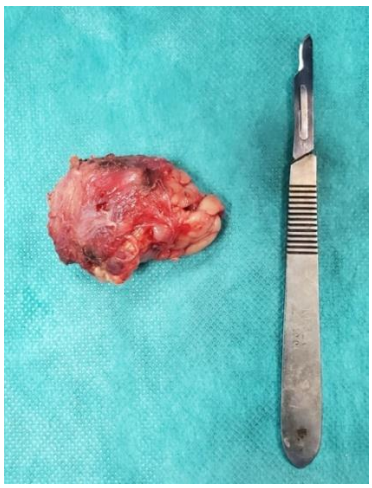

Figure 5: Macroscopic appearance of MCE of submandibular gland tumor.

the pathology and immunohistochemistry examinations revealed a diagnosis of a high-grade mucoepidermoid carcinoma. The multidisciplinary consultation meetings "RCP" decision was to perform lymph node dissection: classified as T4N1M1, radiotherapy and chemotherapy were completed.

Discussion:

Salivary gland carcinomas represent less than 6% of malignant tumors of head and neck [1].

Parotid is the most common localization of these tumors among the major salivary glands, and palate is the most frequent site for minor salivary glands tumors.

They form a heterogeneous group of tumors, with 24 subtypes according to the latest *World Health Organization* "WHO" classification [1]. Around 10% of all salivary gland tumors are located in the sub-mandibular gland, with a high rate of malignant tumors.

Mucoepidermoid carcinoma accounts for between 3% and 10% of salivary tumors and 29% of malignant salivary gland tumors. The

most common location is parotid (56.8%) followed by oral cavity (14%), oropharynx (9.5%), submandibular gland (6.8%), nasal mucosa (6.1%) and sublingual gland (1.3%) [3].

72% of submandibular gland tumors were diagnosed as adenoid cystic carcinoma, while mucoepidermoid carcinoma "MEC" represented 13% [2].

The prevalence is higher among women aged between 50 and 60 years.

MEC commonly presents as a painless swelling with pressure; however, symptoms vary with tumor size, the extension of the tumor and presence of lymph nodes or distant metastasis [4].

In the event of clinical suspicion of malignancy, ultrasound supplemented by a CT scan or MRI are the examinations of choice for assessing local extension and the presence

of adenopathy [5]. Some authors defend the value of ultrasound, especially in differentiating between benign and malignant nature. Images suggestive of malignancy are heterogeneous, poorly limited and more extensive, sometimes with associated adenopathy [6].

Surgical biopsy is to avoid in view of the risk of swarming. Extemporaneous examination on frozen sections may be indispensable.

The confirmation diagnosis is histological, MEC are composed of epidermoid cells, mucinous cells, and a variable proportion of intermediate cells. Several grading systems have been proposed, from low to high grade. The one adopted by the WHO is based on 5 criteria to define 3 grades of malignancy (cystic component less than less than 20%, nerve invasion, necrosis, at least at least 4 mitoses per 10 fields at high magnification, presence of anaplastic contingent).

Low-grade MEC: Mucosecretory cells predominate (> 50%). The squamous cells are well differentiated, and intermediate cells are rare. Their nuclei are regular and mitoses are rare.

High-grade MEC: less than 10% of cells are mucosecreting, squamous or intermediate cells predominate. Marked mitoses and nuclear irregularities.

Intermediate-grade MEC: tumors with intermediate characteristics between the two preceding types.

The grade of differentiation plays a crucial role in the long-term outcomes of oncology patients.

Submandibular gland MECs have been reported to have an aggressive behavior and are the most difficult to treat, since their location and nearby important structures (marginal mandibular nerve, hypoglossal nerve and lingual nerve), most of the time do not allow resection with disease-free margins [7].

Surgical management and the protocols for adjuvant radiotherapy or chemotherapy depend on different factors including perineural or named nerve invasion, positive margins, high-grade tumors, extra glandular spread, positive lymph nodes and locally recurrent disease.

Salivary gland cancers are classified into 4 stages:

Stage I: Noninvasive tumors with no spread to lymph nodes and no distant metastasis

Stage II: An invasive tumor with no spread to lymph nodes and no distant metastasis

Stage III: Smaller tumors (< 4 cm) that have spread to regional lymph nodes but no signs of metastasis.

Stage IVa: Any invasive tumors with either no lymph node involvement or spread to only a single same-sided lymph node, but no metastasis.

Stage IVb: Any cancer, with more extensive spread to lymph nodes but no metastasis.

Stage IVc: Any cancer with distant metastasis.

The therapeutic management depends on each stage.

Stage I: Surgery, +/- radiotherapy (If it's high grade on biopsy or if cancer could not be removed completely).

Stage II: Surgery (may include removal of lymph nodes). +/- radiotherapy (If it's high grade on biopsy or if cancer could not be removed completely).

Stage III: Extensive surgery (removing the submandibular gland, nearby tissues and all lymph nodes in your neck on the same side) +/- radiotherapy +/- chemotherapy.

Stage IV: Extensive surgery + radiotherapy +/- chemotherapy +/- clinical trial of newer treatments

Most of the time Submandibular gland MECs do not allow resection with disease-free margins due to the difficulty of surgery and nearby important structures [7], and tend to recur of metastasize [2]. The current problem is to be able to determine the best therapeutic strategy in order to obtain the best compromise between recovery and sequel.

The prognosis is uncertain, with 25% recurrence and 10% metastases.

The evolution of these tumors is slow (recurrence after 7 to 10 years).

Low-grade carcinomas have a 5-year survival rate of 95%, compared with 40% for high-grade carcinomas. A translocation t(11 ;19)(q21 ;p13) is present in 70% of MECs, generating a

MECT1-MAML2 fusion transcript, responsible for alteration of the

Notch pathway [8]. The presence of this translocation (never identified in high-grade carcinomas), is thought to have a good prognosis.

Conclusion:

Mucoepidermoid carcinoma of submandibular gland is a very rare and aggressive malignant tumor, which makes it difficult to manage and challenging to develop a treatment guideline. Although a complete surgical resection is the gold standard, various treatment options such as chemotherapy, radiotherapy, and immunotherapy have been investigated.

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