

Intraosseous maxillary mucoepidermoid carcinoma: a rare case report

Carcinoma mucoepidermoide intraósseo maxilar: um raro relato de caso

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ABSTRACT

An unusual case of intraosseous mucoepidermoid carcinoma is reported in a 22-year-old female, located in the posterior maxilla region. The article summarizes the main characteristics of the disease, including clinical-pathologic characteristics, treatment and prognosis.

Key words: mucoepidermoid carcinoma; oral diagnosis; immunohistochemistry.

RESUMO

Relatamos um raro caso de carcinoma mucoepidermoide intraósseo localizado em região posterior da maxila em uma paciente de 22 anos de idade. O artigo resume as principais características do processo neoplásico, incluindo as características clinicopatológicas, o tratamento e o prognóstico.

Unitermos: carcinoma mucoepidermoide; diagnóstico oral; imuno-histoquímica.

RESUMEN

Reportamos un caso raro de carcinoma epidermoide intraósseo ubicado en región posterior de la maxila en una paciente de 22 años de edad. El artículo resume las principales características del proceso neoplásico, incluyendo las características clínico-patológicas, el tratamiento y el pronóstico.

Palabras clave: carcinoma mucoepidermoide; diagnóstico oral; inmunohistoquímica.

INTRODUCTION

Mucoepidermoid carcinoma (MEC) is the most frequently diagnosed malignancy in the salivary glands. This tumor is characterized by a variable biological behavior and is mainly found in the major salivary glands, especially the parotid gland. Involvement of the gnathic bones is extremely rare, with a higher incidence in the posterior mandible⁽¹⁻³⁾. MEC affects patients

between their second and seventh decades of life and has a slight female predilection. No racial predisposition has been described^(4,5).

Microscopically, MEC is characterized by the proliferation of three main cell types: epidermoid, mucous, and intermediate. Histopathological grading usually considers tumor cell arrangement, degree of differentiation, tumor cell anaplasia and the interrelationship of the three cell types to classify the tumor as low-, intermediate- or high-grade malignancy^(1, 3, 5, 6). Treatment ranges

from simple surgical excision in the case of low-grade tumors to broad excision associated with cervical lymph node drainage and adjuvant radiotherapy. Factors related to the prognosis of MEC include the size of the primary tumor at the time of diagnosis, histopathological grade, and the presence of metastases⁽⁴⁾.

In an attempt to increase our knowledge of the clinical and pathological features of intraosseous MEC, this study reports the case of a young patient with intraosseous MEC in the maxilla.

CASE REPORT

A 22-year-old white female patient was referred to the Oral Pathology Service of Universidade Federal do Rio Grande do Norte (UFRN) with the diagnosis of a dentigerous cyst four years ago. The patient underwent surgery for removal of tooth 28 and associated cystic enucleation.

Clinical examination showed the absence of facial alterations and cervical lymphadenopathies. Intraoral examination revealed a swelling in the left posterior maxilla and pain on palpation (**Figure 1**). Computed tomography (CT) showed the presence of a hypodense lesion in the area of tooth 28 expanding to the maxillary sinus, but no cortical bone destruction (**Figure 2**).

An incisional biopsy was performed and histopathological examination revealed a neoplasm mainly formed by solid islands of cells with epidermoid, intermediate and clear morphology, which exhibited different degrees of pleomorphism (**Figure 3**).

Histochemical staining with Alcian blue-periodic acid Schiff (PAS) and mucicarmine was positive, confirming the mucoid nature of the examined tissue (**Figure 4**). Immunohistochemical



FIGURE 1 – Intra-oral examination: expansive lesion in left posterior palate

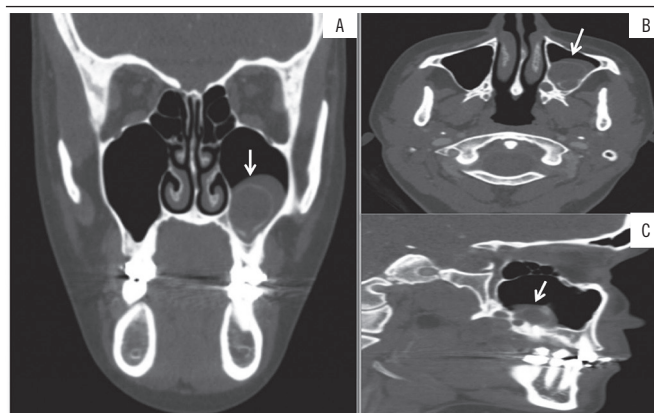


FIGURE 2 – Computed tomography
A) coronal section; B) axial section; C) sagittal section.

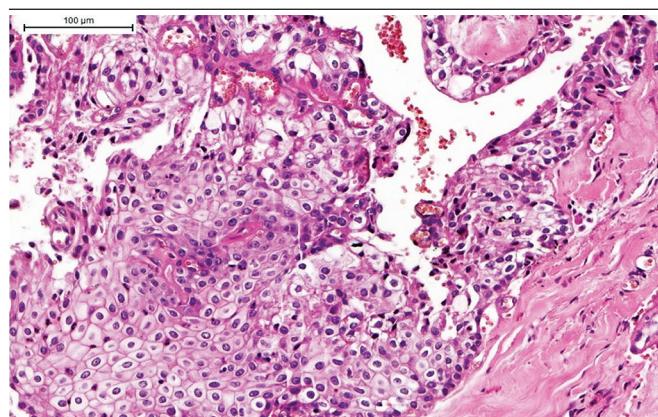


FIGURE 3 – Histological examination of the specimen revealed a neoplasm composed predominantly of nests of epidermoid cells with intermediate differentiation in a fibrous stroma

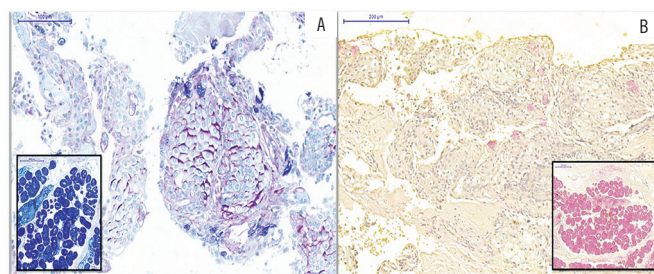


FIGURE 4 – Histochemistry
A) alcian blue-PAS; B) mucicarmine.
PAS: periodic acid Schiff.

staining was positive for pan-cytokeratin (AE1/AE3) and cytokeratin (CK) 7 and 19 (**Figure 5**) and negative for alpha-smooth muscle actin (α -SMA). Based on those results, the final diagnosis was MEC located in the left maxilla.

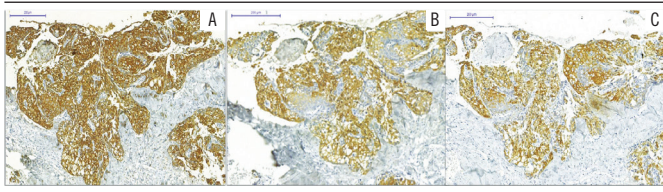


FIGURE 5 – Immunohistochemistry

A) AE1/AE3; B) CK7; C) CK19.

CK: cytokeratin.

Complete surgical resection of the tumor was performed and maxillary provisional obturator prosthesis was placed for rehabilitation (Figure 6). The patient is under clinical and radiographic follow-up and has been disease free for four years.

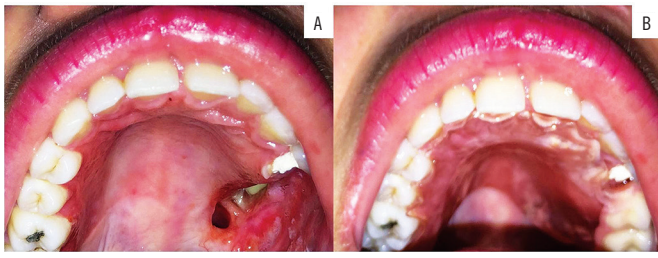


FIGURE 6 – The patient is under clinical and radiographic follow-up and has been disease free for two years

A) complete surgical resection of the tumor was performed; B) maxillary provisional obturator prosthesis was placed for rehabilitation.

DISCUSSION

MEC is the most common malignancy of the salivary glands, accounting for 12% to 30% of all malignant salivary gland tumors, and mainly affects the parotid gland^(7, 8). Although the clinical and histopathological features of MEC are well known, a differential diagnosis with cysts and benign or malignant tumors, especially those of odontogenic origin, is necessary when these tumors occur in gnathic bones. In this respect, histochemical and immunohistochemical markers are fundamental for an accurate diagnosis^(4, 9-11). In the present case, the initial diagnosis was a dentigerous cyst. Despite the clinical and radiographic suspicion of an odontogenic tumor, the morphological findings and the immunoexpression of pan-cytokeratin (AE1/AE3) and CKs 7 and 19, in addition to positive staining for Alcian blue-PAS and mucicarmine, confirmed the glandular nature of the tumor.

In view of the rarity of intraosseous MEC, few reports are available in the literature. The first case was described in the mandible of a female patient older than 60 years⁽¹²⁾. Intraosseous MEC shows a female predilection and mainly afflicts the posterior mandible. The tumor has been shown to affect different age groups, with most cases occurring between the fourth and fifth decades of life^(4, 10).

As shown in the **Table**, 10 cases of intraosseous MEC in the maxilla have been reported over the last 13 years^(11, 13-23). The tumor affected predominantly male patients and symptoms might be present, in agreement with the characteristics of the present case. Patients were, on average, in the fourth decade of life. However,

TABLE – Features of intraosseous mucoepidermoid carcinomas of the maxilla reported in the database PubMed/Medline (2005-2018)

Author	Year	Age	Sex	Symptoms	Radiographic features	Clinical diagnostic hypothesis	Treatment
Namin <i>et al.</i> ⁽¹³⁾	2005	11	F	Pain	Radiolucent multilocular	Infectious lesion	Left hemimaxillectomy
Raut, Khedkar ⁽¹⁴⁾	2009	40	M	Pain	Radiolucent unilocular	OC, nasopalatine duct cyst	Right maxillectomy with neck dissection
Sherin <i>et al.</i> ⁽¹⁵⁾	2011	29	F	Asymptomatic	Radiolucent multilocular	Fibroosseous lesion, benign salivary gland tumor and vascular lesion	Radical resection and adjuvant radiotherapy
Takano <i>et al.</i> ⁽¹⁶⁾	2012	18	M	Asymptomatic	Radiolucent multilocular	OC	Right maxillectomy with neck dissection
Lakouichmi <i>et al.</i> ⁽¹⁷⁾	2013	42	F	Pain	Radiolucent multilocular	Cystic lesion	NI
Chan <i>et al.</i> ⁽¹⁸⁾	2013	51	F	Asymptomatic	Multilocular/mixed	NI	NI
Chan <i>et al.</i> ⁽¹⁸⁾	2013	42	M	Asymptomatic	Radiolucent multilocular	NI	NI
Rathore <i>et al.</i> ⁽¹⁹⁾	2014	18	M	Pain	Radiolucent unilocular	Cystic lesion	Right maxillectomy with neck dissection
Suresh <i>et al.</i> ⁽²⁰⁾	2014	52	M	Asymptomatic	Multilocular/mixed	Osteosarcoma and metastatic tumor	NI
Nallamilli <i>et al.</i> ⁽¹¹⁾	2015	36	M	Pain	Radiolucent unilocular	OT	NI
Martins <i>et al.</i> ⁽²¹⁾	2016	17	M	Asymptomatic	Hypodense image with well-defined edges involving dental apex of the elements 13, 14, and 15	OC	Right maxillectomy with neck dissection
Purohit <i>et al.</i> ⁽²²⁾	2016	28	M	Asymptomatic	Mixed radiopaque-radiolucent lesion	OC	Left hemimaxillectomy
Razavi <i>et al.</i> ⁽²³⁾	2017	43	F	NI	Radiolucent unilocular	OC and adenomatoid odontogenic tumor	Left hemimaxillectomy
Present case	2018	22	F	Pain	Radiolucent unilocular	OC	Left hemimaxillectomy

F: female; M: male; OT: odontogenic tumor; OC: odontogenic cyst; NI: not informed.

the present case was uncommonly diagnosed in a younger patient (22 years), which agrees with some of the studies reported in the Table, while most cases were diagnosed in older patients.

The main symptoms of intraosseous MEC are swelling, pain, trismus and paresthesia, depending on the size of the tumor^(10, 20). In the present study, the patient had a painful swelling in the posterior maxilla. Computed tomography detected a hypodense, unilocular lesion expanding to the cortical bones of the maxillary sinus, but no bone destruction was observed. These imaging findings led to the clinical diagnosis of odontogenic lesions, considering their higher incidence in this region^(1, 4, 24, 25). However, an incisional biopsy is important in the case of osteolytic lesions for histopathological, histological and immunohistochemical analyses that will establish the correct diagnosis.

In addition to the usual histopathological features, the definitive diagnosis of intraosseous MEC is made based on positive staining for mucicarmine, Alcian blue-PAS and cytokeratins (which confirm glandular differentiation), as well as the absence of cortical bone destruction and clinical and histopathological exclusion of metastases and/or odontogenic lesions^(4, 19). In the present case, the final diagnosis was established based on these criteria.

Clinical staging of the tumor is performed according to the tumor-lymph node-metastasis (TNM) system to guide adequate treatment in each case⁽²⁰⁻²⁴⁾. In the present study, the patient was classified as stage II, showing no regional metastases. The treatment of choice was surgical resection of the posterior part of the maxilla and left maxillary sinus. According to the literature, the recurrence rate of stage I and II tumors ranges from 13% to 15%⁽²⁵⁻²⁷⁾. The present patient has been disease-free for two years,

a fact confirming the good prognosis of low-grade, early-stage tumors.

The main goal of palatal obturator prostheses is the rehabilitation of patients with oronasal or oro-sinusal communications⁽²⁶⁾. The objectives of prosthetic rehabilitation in patients with oro-sinusal communications after surgical resection include remodeling and reconstruction of the palatine contour and the restoration of esthetics, mastication, deglutition, respiration and phonation. In the present case, rehabilitation consisted of the placement of a palatine obturator prosthesis. If no tumor recurrence occurs after five years of follow-up, reconstruction using bone and soft tissue grafts will be performed^(26, 27).

Within the context of cancer therapy, surgical resection is without doubt the main objective of treatment. However, the rehabilitation of mutilated patients is a matter of concern since the concept of health includes the physical, psychological and social well-being of an individual. Thus, oral-maxillofacial mutilation after antineoplastic treatment needs to be minimized to restore patients' health and to permit their dignified integration into society after treatment.

CONCLUSION

Although rare, intraosseous MEC should be included in the differential diagnosis of proliferative and osteolytic lesions of the gnathic bones even when the clinical or radiological findings do not suggest malignancy. Early diagnosis, adequate treatment and systematic follow-up by a multiprofessional team are important for a favorable prognosis.

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