Case Report
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Central Maxillary Mucoepidermoid Carcinoma: A Case Report

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Objectives Salivary gland tumors account for 3%-4% of all head and neck cancers, with mucoepidermoid carcinoma (MEC) being the most common type of salivary gland malignancy observed in adults. Intraosseous MEC, which originates in the jaws, is exceedingly rare. It is predominantly found in the posterior part of mandible and typically affects individuals aged 30-50, with a slight female predilection.

Case: A 50-year-old female patient presented with a chief complaint of mild swelling on the left side of palate. The patient did not report any difficulties in swallowing, chewing, or breathing. There was no history of pain, discharge, or bleeding associated with the swelling. During the physical examination, the patient appeared to be in good health. Intra-oral examination revealed a well-circumscribed, pale pink, oval, smooth, sessile, non-tender, and non-mobile nodule measuring 1.0 * 0.6 cm^2 in size. Radiographic findings showed an expansile radiolucent lesion with thinning and perforation of the buccal and palatal cortices, as well as an elevation in the maxillary sinus floor.

Conclusion A rare case of central MEC (CMEC) in the maxilla was presented in this article. The clinical presentation of CMEC in the maxilla can pose challenges in the initial diagnosis, as it may resemble benign lesions, as observed in this case. Therefore, caution is advised in diagnosing CMEC, regardless of the lesion's location, in order to establish a timely and accurate diagnosis and initiate appropriate treatment, even if the clinical features do not strongly indicate malignancy.

Keywords Salivary glands; Carcinoma; Mucoepidermoid, maxilla

Introduction

Salivary gland tumors account for 3%-4% of all head and neck cancers [1], with mucoepidermoid carcinoma (MEC) being the most common type of salivary gland malignancy seen in adults. Stewart et al. described MEC as a tumor containing both epidermal and mucus-secreting components [2], typically originating from the palatal glands. [3-7] While approximately 60% of MECs develop in the major salivary glands, particularly the parotid gland, they frequently affect the minor glands of the palate. [8]

Intraosseous MEC arising in the jaws is exceedingly rare, representing only 2%-3% of all MEC cases. [9, 10] It is predominantly found in the posterior part of mandible and generally occurs in individuals aged 30-50, with a slight female predilection. [11] In 50% of cases, intraosseous MECs are associated with an odontogenic cyst or an impacted tooth. [12] The potential origins of central MEC (CMEC) involve the epithelial lining of an odontogenic cyst, the epithelial rests of the dental lamina, entrapped salivary gland tissue during embryonic development, salivary choristoma, iatrogenically entrapped salivary tissue, or maxillary sinus epithelium. [9] The chromosomal translocation of t(11;19) (q14-21;p12-13) is the most frequent genetic aberration associated with MEC, occurring in 30%-50% of cases. [13, 14]

Although there are no specific guidelines for the treatment of this neoplasm, radical resection with adequate oncologic margins is the recommended treatment modality. [15]

Case Report

A 50-year-old female patient was referred to the Department of Oral and Maxillofacial Pathology, School of Dentistry, Shahid Beheshti University of Medical Sciences, Tehran, Iran, with the chief complaint of swelling in her left palate that had been present for several months. The patient did not report any difficulties in swallowing, chewing, or breathing. There was no history of pain, discharge, or bleeding associated with the swelling. The patient did not have any adverse oral habits. During the physical examination, the patient appeared to be in good health. There were no similar complaints in immediate or distant relatives based on the family history. Intra-oral examination revealed a well-circumscribed, pale pink, oval, smooth, sessile, non-tender, non-mobile nodule measuring 1.0 * 0.6 cm in size. The overlying mucosa was non-ulcerated. Radiographically, a well-defined unilocular radiolucent lesion on the left side of the maxilla, which elevated the floor of the maxillary sinus, was observed (Figure 1).

Based on the clinical examination, radiographic appearance, and patient history, the differential diagnoses included radicular cyst and residual cyst. To obtain a precise diagnosis, an incisional biopsy was performed and sent to the pathology department of Shahid Beheshti University of Medical Sciences.

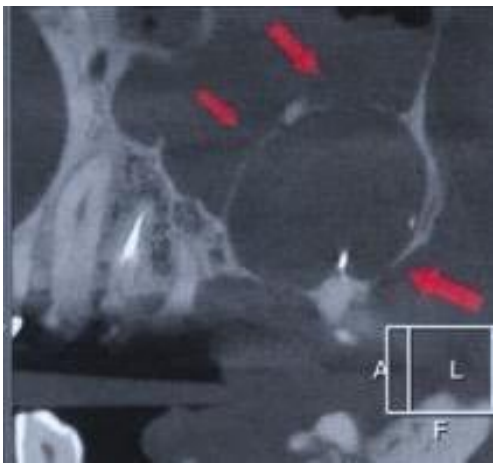

Figure 1- Radiological findings: An expansile radiolucent lesion with thinning and perforation of buccal and palatal cortex and elevation of maxillary sinus floor.

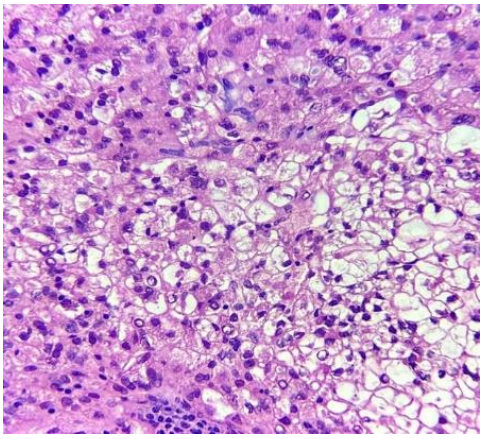

Figure 2- Histopathological findings: Clear cells, mucous cell, epithelial cells, and intermediate cells (×400 magnification, H&E)

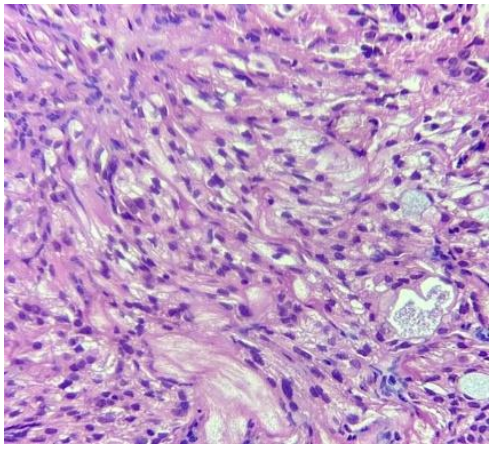

Figure 3- Histopathological findings: Mucous cell, epithelial cells and intermediate cells arranged in sheets within hyalinized stroma (×400 magnification, H&E)

The final microscopic examination revealed an intraosseous neoplasm composed of epithelial, mucous, intermediate, and clear cells arranged in nests, sheets, and islands within a fibrous to hyalinized stroma. Chronic inflammatory cells, cholesterol clefts, foreign body giant cells, psammoma body calcification, normal salivary glands, adipose tissue, and extensive hemorrhage were evident (Figures 2 and 3).

Informed consent was obtained from the patient before publishing her radiograph and histopathological view.

Discussion

Mucoepidermoid carcinoma is the most prevalent type (20%) of malignant salivary gland tumors. [16] A literature review by de Souza et al. [17] revealed that CMEC was slightly more common in women (51.7%) than in men (48.3%), with a mean age of 46.51 years, primarily occurring in the fifth to seventh decades of life. The patient discussed in this case report aligned with the age range and gender preference identified in the study by de Souza et al. Additionally, de Souza et al. found that CMECs are more frequently located in the mandible, affecting it in about two-thirds of cases. [17] However, what distinguished this case report was the unusual location of the lesion in the maxilla.

According to Brookstone et al., swelling and pain with trismus, paresthesia, and tooth mobility are common clinical symptoms of CMEC. [18] In contrast to these findings, the patient in this case report presented only asymptomatic swelling.

Since the clinical presentation of CMEC in the maxilla can mimic benign conditions like odontogenic cysts and tumors, its rare location initially led us away from suspecting MEC.

Reports indicate that 12% of CMEC cases involve metastases, commonly affecting local lymph nodes, lung, and brain. [19] In rare instances, metastasis may occur in the ipsilateral clavicle. [20]

Over the years, numerous hypotheses have been suggested to explain the pathogenesis of MEC, which remains elusive. [21]

The etiology of maxillary CMEC is still debated, with several theories proposed, including: [22]

1. Neoplastic transformation of ectopic salivary gland tissue within the jaw bones.
2. Transformation of the epithelial lining of the maxillary sinus.
3. Origin from odontogenic cysts.

However, there is a lack of direct evidence supporting these pathogeneses in the literature. Although these are considered likely risks for CMEC, the direct evidence for such pathogenesis has not been well-documented in previous reports.

Brookstone et al. [18] proposed a three-tier grading system for intraosseous MEC, aiding in prognosis determination:

- Grade 1: Lesions showing no cortical plate expansion or rupture (best prognosis).
- Grade 2: Lesions causing cortical expansion but not rupture.
- Grade 3: Lesions presenting cortical perforation, periosteal breakdown, or nodal spread (poorest prognosis).

The patient in this case report was classified as grade 3.

For radiological evaluation, panoramic views, cone-beam computed tomography (CBCT), and thoracic CT are recommended [23], as CMEC can easily be confused with other intraosseous or odontogenic pathologies [24, 25] such as radicular cyst, paradental cyst, calcifying epithelial odontogenic cyst, benign odontogenic keratocystic tumor, dentigerous cyst, ameloblastic fibroma, odontogenic myxoma, adenoid cystic carcinoma, intraosseous squamous cell carcinoma, and metastatic tumors to the jaws from lung, kidney, or prostate cancer.

Histopathologic grading by Goode et al. [26] categorizes MEC into low, intermediate, and high grades. According to de Souza et al., the most common grade observed in their study was low grade [17], which aligns with the patient described in the present study.

It is crucial to differentiate the glandular odontogenic cyst (GOC) and CMEC due to their clinical and histopathological similarities but differing treatment approaches and prognoses. To distinguish these two entities, fluorescence in situ hybridization (FISH) for MAML2 gene rearrangement can be a valuable diagnostic tool. [27] The presence of MAML2 rearrangement is indicative of MEC and can significantly aid in ensuring the correct diagnosis and guiding appropriate treatment strategies. [28] Indeed, it is worth noting that despite its diagnostic utility, approximately 50% of CMECs do not exhibit it. [29] In this case, CMEC was established based on the solid nature of the lesion, infiltrative growth pattern, and invasion of adjacent stroma. These histopathological features play a key role in confirming the nature of the tumor and differentiating it from a GOC.

Conclusion

A rare case of central MEC (CMEC) in the maxilla was presented. The clinical presentation of CMEC in the maxilla can pose challenges in the initial diagnosis, as it may resemble benign lesions, as observed in the present case. Therefore, caution is advised in diagnosing CMEC, regardless of the lesion's location, in order to establish a timely and accurate diagnosis and initiate appropriate treatment, even if the clinical features do not strongly indicate malignancy.

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