

Case Report

Central Mucoepidermoid Carcinoma: A Case Report

Bindiya PK, Samreen Taufiq, Selvamani M

Abstract

Central mucoepidermoid carcinoma is a rare tumour, but a well-known entity affecting the bones of the jaws. It comprises 2 - 4% of all mucoepidermoid carcinomas. Occurrence in maxilla is rarely reported. This case report presents a central mucoepidermoid carcinoma in the maxilla of a 17-year-old female patient.

Keywords: Central Mucoepidermoid Carcinoma; Maxilla; Malignant Salivary Gland Neoplasm; Mucus Cells.

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Introduction

Mucoepidermoid carcinoma (MEC) is the most common malignant salivary gland neoplasm. It has the second highest frequency of occurrence among all salivary gland tumors.¹ It is the commonest salivary gland neoplasm in adults as well as in children and comprises of mucus secreting cells, epidermoid cells and intermediate cells in varying combinations forming cysts and solid islands.² They are histologically low grade and usually affects the mandible. Occurrence in maxilla is rarely reported.³

Most of the reported cases of this lesion are asymptomatic and appear as unilocular or multilocular radiolucency found during routine dental examinations.¹ Atypical salivary gland neoplasms arising within the jaws as primary central bony lesions are extremely rare. They represent 2-4.3% of all MECs reported.⁴⁻⁶

Case Report

A 17-year-old female patient reported with a chief complaint of pain and swelling on the right side of face since three months. History of present illness revealed that the swelling initially was small which gradually has enlarged to the present size and was associated with occasional mild pain. Patient gave a history of trauma to the anterior teeth region on the right side of the face, three years back. Upon extra oral examination, a solitary swelling on the right side of face involving the zygomatic region, extending from ala of nose to the zygomatic arch anteroposteriorly and from occlusal line of premolars to infra orbital margin supero-inferiorly was identified.(Figure 1) The swelling measured approximately three centimetres in diameter and was almost oval

in shape with a rough texture of the overlying skin. On palpation, all the inspectory findings were confirmed along with tenderness, firm consistency and absence of any kind of discharge. The swelling appeared to be fixed to the underlying structures. Intraorally there was obliteration of the upper right buccal vestibule and teeth #14 and #15 appeared to be non-vital upon vitality test.



Figure 1: The clinical picture showing swelling on the right side of the face.

Orthopantomograph showed an unilocular radiolucency with ragged borders and thick sclerotic margins extending from periapical area of tooth #14 to #16 till the lower border of the maxillary sinus.(Figure 2) The 3D reconstructed computed tomography (CT) image showed the destruction of the bone in the maxillary region.(Figure 3) Thus on the basis of overall clinical and radiological findings, a provisional diagnosis of odontogenic tumour of right maxilla was agreed.

On histopathological examination, hematoxylin and eosin stained sections revealed tumour mass containing connective tissue stroma interspersed with numerous mucin containing cystic spaces lined by mucinous cells.(Figure 4) Nests and cords of epidermoid and intermediate cells, interspersed with mucous cells (Figure 5) and, sheets of clear cells were also evident.(Figure 6) Based on the clinical, radiographic and histopathological features, the lesion was diagnosed as mucoepidermoid carcinoma.



Figure 2: The Orthopantomograph showing the unilocular radiolucency with ragged borders

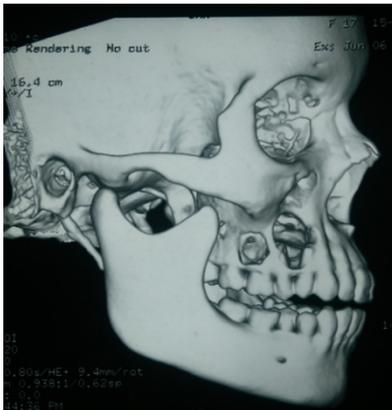


Figure 3: The 3D reconstructed CT image showing the destruction of the bone in the maxilla.

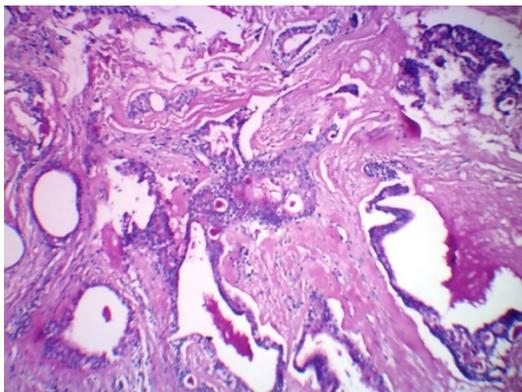


Figure 4: The photomicrographs of H & E stained sections shows tumour islands with

multiple cysts containing mucin and nests and cords of tumour cells (10x).

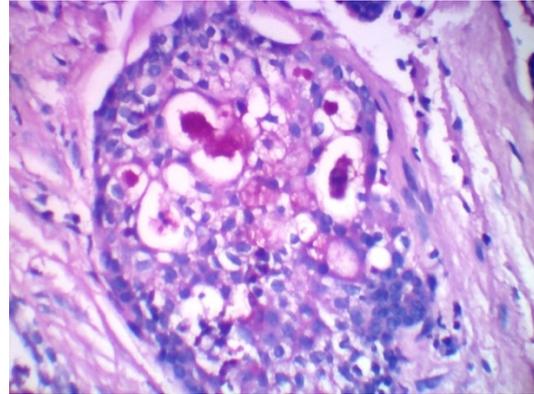


Figure 5: The photomicrographs of H & E stained section shows nest of tumour cells made of epithelial and intermediate cells and interspersed with mucous cells (100x)

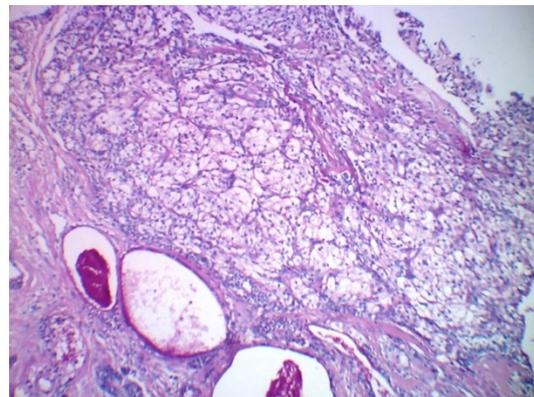


Figure 6: The photomicrographs of H & E stained section shows sheets of clear cells (40x)

Discussion

Loos in 1913 described Intraosseous carcinoma arising in the jaw bones as a central epidermoid carcinoma.⁷ The term "primary intraosseous carcinoma" (PIOC) was coined by Pindborg in the first edition of the World Health Organization classification for the histopathological typing of the odontogenic tumors.⁸

- Type 1 PIOC ex odontogenic cyst
- Type 2a Malignant ameloblastoma
- Type 2b Ameloblastic carcinoma arising de novo, ex ameloblastoma or ex odontogenic cyst
- Type 3 PIOC arising de novo: (a) keratinizing type (b) non-keratinizing type
- Type 4 Intraosseous MEC

Kochaji et al. published the criteria for diagnosis of intraosseous MEC^{6,9,10}

- Presence of an intact cortical plate

- Radiological evidence of bone destruction
- Histological confirmation
- Positive mucin staining
- Absence of primary lesion in the salivary gland
- Exclusion of an odontogenic tumour.

Histopathological criteria for MEC diagnosis as per Seifert¹¹, Sobin¹² and Auclair et al¹³ the tumours are graded for malignancy as follows:

- Low Grade: Highly differentiated neoplasia with a predominance of macro and micro cysts. Presence of intermediate and mucin-producing cells.
- Intermediate Grade: Predominance of intermediate cells and a few cysts. Presence of mucin-producing cells and islands of epidermoid cells.
- High Grade: Poorly differentiated neoplasia with predominance of intermediate and epidermoid cells in solid blocks. Mucin-producing cells are present.

Various possibilities for the origin of MEC are^{14,15}

- Entrapment of the retro-molar mucous glands within the mandible, which later undergo neoplastic transformation.
- Embryonic remnants of the submandibular and sublingual glands trapped within the mandible during development.
- Neoplastic transformation and invasion from the lining of the maxillary sinus.
- Neoplastic transformation of the mucous secreting cells from the epithelial lining of the dentigerous cyst associated with impacted third molars.

Possibilities for origin in the present case could be due to neoplastic transformation and invasion from the lining of the maxillary sinus and neoplastic transformation of entrapped minor salivary glands within the maxilla. Age of incidence is 1st to 7th decade of life. In adults it occurs mostly in females and more often in the posterior mandible.⁴ Radiographic examination is important for categorizing it as a central jaw lesion and to know the extent of the lesion with involvement or encroachment of adjacent vital structures and hence concluding the diagnosis.

Brook stone and Huvos⁹ suggested a three-grade classification for intraosseous MEC.

- Grade 1: Without expansion and rupture of cortical plate.
- Grade 2: With expansion but without rupture of cortical plate.
- Grade 3: With rupture of cortical plates or presence of regional metastasis.

Though panoramic radiography is a simple and cost effective imaging modality for jaw lesions, lesions involving maxilla require a CT imaging. CT gives a better information about the extent of the lesion, which is particularly needed in cases involving the maxilla because involvement of floor of the orbit, nasal fossa, maxillary sinus, etc., will change the treatment and prognosis of such cases in a major way. Management of central intraosseous MEC is through surgery, which includes curettage, enucleation, marsupialization and wide local excision.^{3,16,17} As a rule, even the low-grade MEC's should be managed by wide local resection, en-bloc resection, hemi-mandibulectomy or hemi-maxillectomy.^{9,18,19} Neck dissection is usually part of the treatment in cases where metastasis to the cervical nodes is suspected.¹⁹ Radiotherapy is recommended for high-grade MEC cases.^{19,20} These tumours usually have a good overall prognosis, but central MEC cases should be followed-up for a longer period due to the possibility of late recurrence or regional metastasis. Death may occur if there is extension into vital structures such as the base of the brain.^{12,15}

Conclusion

Central MEC of the maxilla is a rare lesion. Modern investigation methods like CT can accurately identify the extent of the lesion and thereby a correct treatment can be given to the patient. Long term follow-up is very important in these cases to identify recurrences at the earliest.

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