 DOI: 10.5958/2394-6792.2016.00137.X

Central mucoepidermoid carcinoma of the maxilla – case report

Dhanalakshmi Subramaniam[1,*], Sudha Venkatesh[2], Sofia Chandrababulu[3]

[1]Assistant Professor, [2]Professor, [3]PG Student, Madras Medical College, Chennai

***Corresponding Author:**
Email: dhanadinesh25@gmail.com

Abstract

Central mucoepidermoid carcinoma is very rare constituting 2–4% of all mucoepidermoid carcinomas. Its location in the maxilla is even rarer. The occurrence of this common tumor at the rare site poses a diagnostic challenge to the clinicians and pathologists. Here we report one such rare interesting case of mucoepidermoid carcinoma in a 30 year old female developing in the maxilla with involvement of adjacent vital structures.

Keywords: Mucoepidermoid, Maxilla, Carcinoma, Salivary gland, Mucin

Introduction

Central mucoepidermoid carcinoma are extremely rare tumors, but well known entity affecting the jaw bones representing about 2 to 4% of all mucoepidermoid carcinomas. They are histologically low-grade cancers, usually affecting the mandible. There are very few case reports available in literature on central mucoepidermoid carcinoma of the maxilla. One such rare case is reported here.

Case Report

A 30 year old female presented with complaints of pain and swelling on left side of face for the past 6 months. Initially it started as a small swelling then progressed to present size. On local examination a diffuse hard swelling involving the left side of face extending from infraorbital rim to zygomatic buttress, medially upto ala of nose, laterally to zygomatic arch was made out. Overlying skin was smooth and no secondary changes was made out.

On oral examination a well-defined hard mass was noted involving the left maxilla measuring about 4 x4 cm extending from left premolar to maxillary tuberosity posteriorly and medially upto the mid palatal suture.

CT paranasal sinuses showed an expansile and lytic lesion involving left maxilla with bony erosion and extension into the nasal cavity and into the adjacent soft tissues.

Based on the clinical and radiological findings a provisional diagnosis of ameloblastoma was made. An incisional biopsy was done which revealed the presence of mucus cells, intermediate cells and epidermoid cells arranged in sheets and strands along with few cystic spaces in a connective tissue stroma. The diagnosis of mucoepidermoid carcinoma was made. Resection of the maxilla with roof of nasal cavity for clearance was done.

Received maxillectomy specimen measuring 7x6x5 cm with two teeth. External surface was congested. Cut surface showed a partially encapsulated irregular mass measuring 5x4x2cm, which was predominantly cystic. Some cysts were filled with brownish material and focal areas showed mucin filled cysts admixed with solid grey white areas.(Fig. 1) Roof of nasal cavity clearance specimen showed two grey brown soft tissue fragments each measuring 1.5 x 1 x 0.5 and all were embedded.

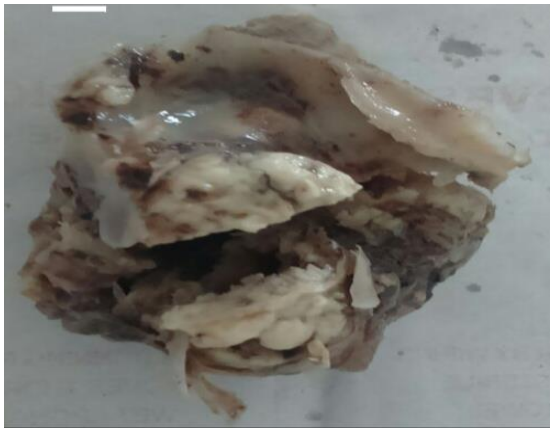

Fig. 1: Gross picture showing grey white mass with solid and cystic component

Microscopic examination showed a neoplasm composed of cystic spaces of varying sizes lined by mucous cells and intermediate cells and contained mucinous material. Nests and strands of squamous cells were seen forming the epidermoid component (Fig. 2). Clear cells were also seen. (Fig. 3) Mucinous material was extra vasating into the surrounding stroma with dense inflammatory cell infiltrate along with areas of calcification.

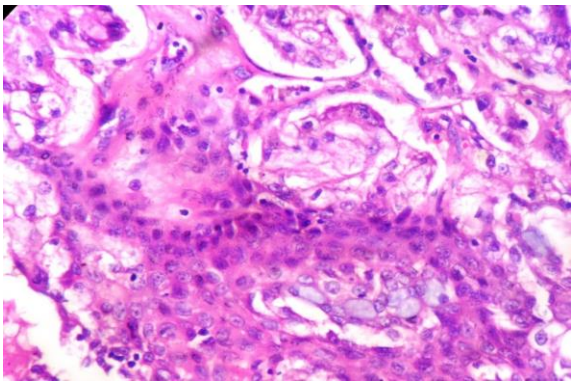

Fig. 2: H&E photomicrograph showing squamous cells & mucous cells

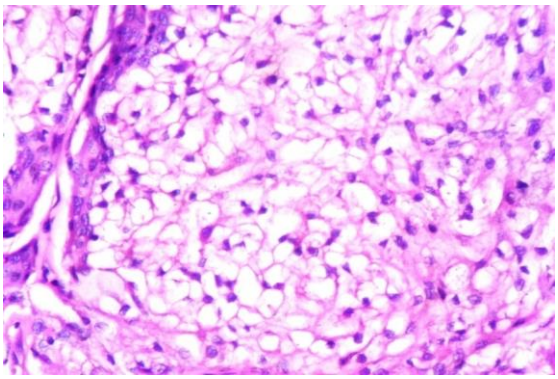

Fig. 3: H&E photomicrograph showing clear cell component

Roof of the nasal cavity also showed the presence of tumor. Also seen were 2 lymph nodes with features of reactive hyperplasia.

With this a diagnosis of low grade mucoepidermoid carcinoma with involvement of the roof of nasal cavity was made.

Discussion

Central mucoepidermoid carcinoma was first reported by Lepp in the mandible of a 60 year old female in 1939[1]. There are only less than 200 cases reported in literature[2]. Various theories have been proposed on the origin of the tumor and is thought to be due to one of the following: 1. Metaplasia of the odontogenic cyst epithelium, 2. Entrapped salivary tissues or minor salivary glands during development, 3 from the Maxillary sinus epithelium, 4. Odontogenic remnants of the dental lamina[3].

Central mucoepidermoid carcinoma occurs more commonly in the mandible than the maxilla. It has predilection for females twice than that of males and is found to occur in all age groups with majority occurring in fourth and fifth decades.

Clinically the patient presents with pain, trismus and parasthesias. The pain sometimes radiating to the teeth, palate, face, nose misleading the clinician to focus on neural, dental or maxillary sinus problems.

Radiologically they usually present as multilocular cyst like radiolucent lesion with very few cases also showing radioopacities in it. Considering the location and radiological features, the differential diagnosis of ameloblastic carcinoma, CEOT, clear cell odontogenic carcinoma should be considered.

Brookstone and Huvos 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[4]

So radiography plays a vital role in categorizing the central lesions of the jaw

World Health Organization classification for the histopathological typing of the Odontogenic tumours[5]

Table 1 : Classification of Primary Intraosseous Carcinoma
Type 1: PIOC ex odontogenic cyst
Type 2A: Malignant ameloblastoma
Type 2B: Ameloblastic carcinoma arising de novo, exameloblastoma or exodontogenic cyst
Type 3: PIOC arising de novo
a. Keratinising type
b. Non-keratinising type
Type 4: Intraosseous mucoepidermoid carcinoma

Our case falls under type 4. The commonly accepted criteria for diagnosis was proposed by Alexander et al[6] and modified by Brow and Waldron[7] to consider it as a central lesion which includes radiographic evidence of lytic lesion in the bone, histological confirmation, evidence of intracellular mucin, exclusion of other primary tumor or an odontogenic tumor.

Our case showed presence of calcification which could be dystrophic due to the amorphous eosinophic material secreted by the intermediate cells and could be an evidence for long standing lesion. Though our tumor was of low grade due to extensive involvement, radical resection was considered as the treatment of choice.

Most of the central mucoepidermoid carcinomas reported in literature are of low grade and carries a favourable prognosis. But maxillary cases are found to have worse prognosis due to extension into vital structures. Eventhough they are low grade tumors, treatment includes wide local resection[8], enbloc resection or hemi mandibulectomy, as conservative line of management favour recurrences. When regional nodes are involved, neck dissection is done[9]. Adjuvant radiotherapy is useful in high grade tumors and in cases with positive surgical margins.

Conclusion

Central mucoepidermoid carcinoma of the maxilla is a rare entity and is usually misdiagnosed as an odontogenic tumor. The difficulty in histopathological diagnosis could be due to the occurrence of this common tumor in a rarest site. Hence a thorough knowledge about the clinical, advanced imaging techniques, and histopathology will not only help in confirming the diagnosis but also to evaluate the extent of the lesion which has an impact on the treatment and prognosis. Extension into vital structures like base of skull carries a grave prognosis. Central MEC must have a long term follow up for recurrence and regional metastasis.

References

1. Namin AK, Moshref M, Shahoon H, Mashhadi A, Khojasteh A. An intraosseous mucoepidermoid carcinoma of the maxilla in a teenager: A case report and review of literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2005;100:93-6.
2. Diana Bell, Carol Lewis, Adel K E1- Naggar, Randal S. Weber Primary intraosseous mucoepidermoid carcinoma of the jaw: Reappraisal of the MD Anderson Cancer Centre experience. Head and Neck Vol 38 issue S1, version of Record online 28 August 2015.
3. Eversole LR, Sabes WR, Rovin S. Aggressive growth and neoplastic potential of odontogenic cysts with special reference to central mucoepidermoid and mucoepidermoid carcinomas. Cancer 1975;35:270 82.
4. Brookstone MS and Huvos AG Central salivary gland tumors of the maxilla and mandible: a clinicopathologic study of 11 cases with an analysis of the literature. *J Oral MaxillofacSurg*1992,50:229-236.
5. Pindborg JJ, Kramer IRH, Torloni H. Histologic typing of odontogenic tumors, jaw cysts and allied lesions. Geneva: World Health Organization, 1971, pp35–36.
6. Alexander RW, Dupuis RH and Holton H Central mucoepidermoid tumor (carcinoma) of the mandible. *J Oral Surg*1974,32:541-547.
7. Browand BC and Waldron CA central mucoepidermoid tumors of the jaws. *Oral Surg Oral Med Oral Pathol*1975,40:631-643.
8. Martinez Madrigal F, Pineda-Daboin K, Casiraghi O, Luna MA. Salivary gland tumors of the mandible. Ann Diag Pathol 2000:4:347–353.
9. Simon D, Somanathan T, Ramdas K, Pandey M. Central mucoepidermoid carcinoma of mandible: A case report and review of literature. World J SurgOncol 2003;1:1.