

## Clear cell Odontogenic carcinoma of Maxilla: A Rare Case Report

Dr.Surej Kumar LK<sup>1</sup>, Dr.Sherin.A.Khalem<sup>2</sup>, Dr. Nikhil Mathew Kurien<sup>3</sup>

### Abstract:-

Clear cell odontogenic carcinoma is not very common in oral cavity. Many cases had been reported in tumours affecting kidney but very few cases had been reported in mandible or maxilla. Literature review reveals only 9 cases had been reported in maxilla since now. In 1985, Hansen et al, reported a locally aggressive odontogenic neoplasm. The clear cell Odontogenic tumour is classified as a benign but locally invasive odontogenic tumour in the current World Health Organization classification for Odontogenic tumours. Our case is one of the rare entities which have one of the largest dimensions among the clear cell odontogenic tumour variant among the 45 cases of head and neck region had been reported so far.

**Keywords:** Odontogenic carcinoma; Odontogenic neoplasm, hemi maxillectomy, invasive Odontogenic tumour

<sup>1</sup>Prof & Head, Department of Oral & Maxillo-facial Surgery <sup>2</sup>Sr Lecturer, <sup>3</sup>Reader

PMS College Of Dental Science & Research, Vattappara, Trivandrum.

Corresponding Author mail: [surejkumarlk@gmail.com](mailto:surejkumarlk@gmail.com)

### Introduction

In 1992 WHO defined CLEAR CELL ODONTOGENIC TUMOUR as “A benign but locally invasive neoplasm originating from odontogenic epithelium and characterized by sheets and islands of uniform, vacuolated and clear cells”

Clear cells are found in many different tumours and usually result from fixation artifacts; intracellular storage of various substances such as glycogen,

mucin, or lipid; or paucity of organelles<sup>1</sup>. In the maxillofacial area, clear cell tumours usually are salivary<sup>2</sup> or odontogenic in origin, although occasionally metastatic tumours need to be considered<sup>3</sup>. Odontogenic neoplasms composed entirely or predominantly of clear cells are exceptionally rare<sup>4</sup>. E.g: include the clear cell variants of calcifying epithelial odontogenic tumour and of ameloblastoma and clear cell odontogenic carcinoma. The latter has been shown to

exhibit aggressive behavior in terms of recurrence or metastases to regional nodes and distant sites. Consequently, terms such as *clear cell odontogenic tumour* and *clear cell ameloblastoma* seem inappropriate to identify this rare entity, and most authors now agree that these tumours should be called *clear cell odontogenic carcinomas*, even if showing occasional ameloblastoma-like histologic patterns<sup>2,3</sup>. A search of the English literature, using clear cell and odontogenic tumour, odontogenic carcinoma, and ameloblastoma as key words, identified unequivocal cases of clear cell odontogenic carcinoma. Owing to the limited numbers of such tumours reported so far, only incomplete data are available on their clinical, immunohistochemical, and prognostic features. The present study was aimed at reporting a case of clear cell odontogenic carcinoma (CCOC) of maxilla, focusing on its morphologic, immunophenotypic, differential diagnostic features and mode of surgery. Also, in view of the attenuated malignancy demonstrated by the tumours reported herein, the therapeutic approaches and the prognosis of CCOCs are discussed.

### CASE REPORT

A female patient of age 50 years came with a history of painful swelling in the

left posterior region of the upper jaw for the past 6 months. Swelling - slowly enlarged and reached the present size. Pain was moderate, intermittent and localized in the left side of the maxilla. On **inspection** a diffuse swelling was present which was roughly oval in shape. The skin appeared normal. Obliteration of buccal and palatal vestibule was present. (Fig 1)

**On palpation** lobulated swelling of approximate size 3 X 4cm which was firm, tender and not mobile. The surface was irregular and nodular. Surface was irregular and nodular. The swelling has started growing slowly to the present size measuring about 5.5 cm anterioposteriorly and 4.5 cm superioinferiorly. The skin overlying the swelling was stressed and the borders were ill defined. On intraoral examination the swelling was extending from 21 – 28 area with obliteration of the palate. The patient was subjected to routine blood investigation, ECG chest radiograph. After the complete physical examination no abnormality was noticed, so as a result fnac was done which was negative.



Fig 1: Swelling in the right side of the mandible

Biopsy was done and the tissue specimen consisted of large and irregular sheets or cords of neoplastic cells in a richly cellular, collagenous stroma. The neoplastic cells were cuboidal or polyhedral, with centrally placed, rounded nuclei. Some of the neoplastic clusters showed a peripheral rim of cells with abundant eosinophilic cytoplasm and centrally located clear cells.

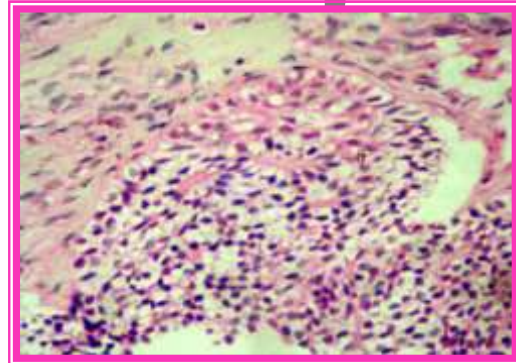
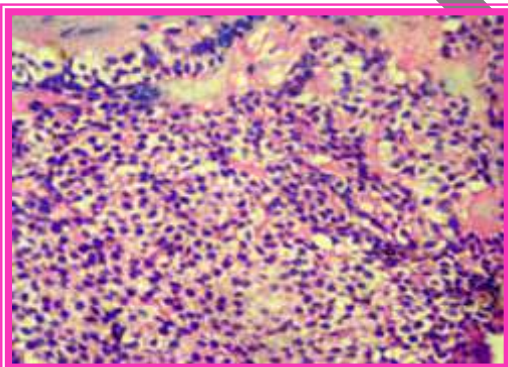


Fig 2: The peripheral cells of each islands

Fig 3: Few island show stellate type of central cells & show hyperchromatic nuclei where as

the reverse of polarity. Central cells show clear cytoplasm and oval to stellate nuclei.

**Surgical procedure:-**

Under nasotracheal intubation GA was administered. Extended Weber Fergusson incision was placed. Hemi maxillectomy was done giving 1cm safety margin(Fig 4). Tumour removed in toto. The surgical defect was covered using acrylic stent and split skin graft.

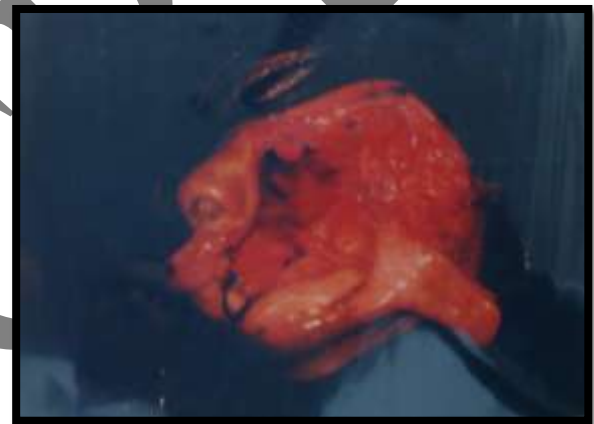


Figure 4: Exposure of tumour

This is disease free after 3 years until last follow up. (Figure 5)

Figure 5: Follow up Profile



← = after surgery

**Discussion:-**

Clear cell odontogenic carcinoma is a rare neoplasm first described by Hansen et al (1985)<sup>1</sup>. It is locally invasive and destructive; lymph node and pulmonary metastases reported by Bang et al. Clear cell normally presents with mild pain or tenderness or loosening of teeth. Lesion usually presents with expansion of the jaw and ragged area of radiolucency. First case was reported in Maxilla. COC is having aggressive behaviour with a predilection for local recurrence, evidence of distant, metastasis, and histological distinct features; these tumours are now considered as malignant<sup>2</sup>. Long term surveillance (including chest –radiography) is necessary because of the potential for local – regional recurrence and/or late metastasis spread. It is noteworthy that ultrasound evaluations of the liver, kidneys and spleen did not reveal any metastatic lesions.

Tumours of clear cell component in the head and neck region could originate from odontogenic epithelium and salivary glands or even as metastasis from distant locations like kidneys. Even additional specimen staining of mucin is recommended to rule out – mucoepidermoid carcinoma. Furthermore

clear cell odontogenic carcinoma can be distinguished from the clear cell variant of calcifying epithelial odontogenic tumour lacks calcification and amyloid deposits<sup>4</sup>. CCOC is rare it has been difficult to identify risk factors for recurrence. Experience with other tumours suggests that factors such as soft tissue involvement, size of lesion, location and nodal or distant metastasis should be considered when developing treatment strategies. Lymph node – metastasis on initial presentation was infrequent, whereas nodal involvement was markedly increased in recurrent disease. Distant metastasis spread – occurred in 15% (Six of 40). Half of them were involving the lung. None of the distant metastasis had locoregional control. Assess of recurrence is the presence or absence of surgical margins. This information was not reported in most cases. Nevertheless with curettage, its margins were completely negative as compared with resection. A higher recurrence rate is there with curettage. Lack of tumour free margins likely contributed to the large number of patients who experienced multiple recurrences. Unfortunately, insufficient numbers of patients received adjuvant radiation therapy or neck dissections with treatment where recurrence developed –

average 7 yrs follow up. On the basis of data presented, surgical control of CCOC with an enbloc resection of bone and soft tissue involvement decreases the risk of recurrence. It is imperative that the surgical margins are free of tumour. If positive margins are noted on permanent section – the treatment will be re-resection to attain tumour free margins. A regional lymph node dissection can be performed for staging and treatment of regional diseased.

Adjuvant radiation therapy may contribute to local control in patients with extensive soft tissue invasion which tumour free margins are not possible or in patients with positive nodes and/or extra capsular spread.

#### **Conclusion:**

At the end we can conclude that the clear cell odontogenic carcinoma is not a very common tumour. Very few cases are reported in the posterior region of the maxilla measuring 5cm x 3cm. The actual origin of the tumour is very difficult to understand whether it is from the odontogenic epithelium or from the salivary glands or it is from the secondary metastasis from the kidney. To rule out distant metastasis bone scanning (scintigraphy) is a best modality to evaluate any tumour in the kidney of the

clear cell variant. To study the tumour in detail the best radiographic imaging modality in the maxillofacial region is 3D reconstructive image in the CT scan and further detail evaluation and surgical treatment, the best radiographic modality is cone beam CT. It is said to conclude that a follow up of 3 years shows satisfying results.

#### **References:**

1. Hansen LS, Eversole LR, Green TL, Powell NB. Clear cell odontogenic tumour-a new histologic variant with aggressive potential. *Head Neck Surg.* 1985;8:115–23
2. R.A Cawson, J D Langdon, JW Eveson. *Surgical pathology of the Mouth and jaws.* Page7-8.
3. Charles S et al. Clear cell Odontogenic carcinoma. A comprehensive analysis of treatment strategies. *Head & Neck.* June 2005;100:536-541
4. K. Adebisi & V. Ugboko : Clear Cell Odontogenic Carcinoma Of The Maxilla: A Case Report . *The Internet Journal of Dental Science.* 2005 Volume 2 Number 2
5. Eugenio Maiorano, Mario Altini, Giuseppe Viale, Adriano Piattelli, and Gianfranco Favia, Clear Cell Odontogenic Carcinoma. Report of Two Cases and Review of the Literature. *Am J Clin Pathol* 2001;116:107-114