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# Clear Cell Odontogenic Carcinoma – A Report Of A Rarity In A Relatively Rare Site

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***Abstract:***
*Clear cell odontogenic carcinomas are uncommon epithelial neoplasms that have metastatic potential. They are rare tumors characterized histologically by epithelial cells with vacuolated cytoplasm and is a potentially aggressive tumor with a tendency for recurrence. It preferentially occurs during the fourth to sixth decades of life and has female predilection with mandible as the most common site. Here we report a case of CCOC of maxilla in a 20 year old female patient. A wide surgical resection under GA was performed and regular follow up is being done. Prognosis was good with no recurrence.*

***Key words:*** *Clear cell, odontogenic, carcinoma, Palate, maxilla*

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## I. Introduction:

Clear cell odontogenic carcinoma is a peculiar odontogenic tumour of the jaw with marked histological features and its unique behaviour. Earlier it was known as clear cell odontogenic tumour (CCOT) or Clear cell ameloblastoma (CCA).[1] Clear cell odontogenic tumour was first classified under benign odontogenic tumour in the year 1985 by Hansen et al.[2] Then in 1992 WHO classified CCOT as benign neoplasm with capacity of undergoing local invasion. Later in 2003, by Reichart and Philipsen CCOC was identified as carcinoma, after which, WHO again in the year 2005, classified CCOC as a low-grade malignant tumour, characterized by locally destructive and aggressive behaviour with local recurrence, regional lymph node metastasis, and sometimes distant metastasis.[3] Even though the first case was documented in 80's, CCOC is a rare tumour. Literature search suggest that less than 120 cases reported till date. It occurs in $5^{th}$ -$7^{th}$ decade of life. 73.8% occurs in the mandible, more common in females (1.5:1 to 2:1) in the mean age of 60 years.[4] Here we present a case report of a 20-year-old female with CCOC in maxilla treated with segmental resection and rehabilitation with obturator. The case is discussed from the diagnostic point of view with additional pathological details pertaining to the case.

## II. Case summary:

A 20 years old female patient reported with a chief complaint of swelling in the palate of 1 year duration, which was insidious in onset, initially of peanut sized and overtly increased to the present size in last 2 months. The swelling was associated with pain, which was dull, throbbing, non-radiating, and continuous in nature. No postural or diurnal variations. No associated toothache, trismus and paraesthesia. Swelling was associated with itching, difficulty in breathing and thin watery nasal discharge since 2 months. No H/O local trauma, fever or chills. H/O similar type of swelling in the same site 8 years back for which the patient got it surgically operated. H/O treatment for Pyelonephritis and complete resection of left side kidney, 2 years back. No contributory family history. On general physical examination, patient was moderately healthy. Examination of head and neck showed no abnormalities of TMJ and lymph nodes. On extra-oral examination no abnormalities were detected. Intraoral examination revealed a single, well-defined swelling on the left side of the palate, extending anterio-posteriorly from 22 to the maxillary tuberosity, medio-laterally extending from the palatal gingiva of 26, 27 to the midline of the hard palate. The colour of the mucosa over the swelling was normal with nodular surface and was fixed to the underlying tissues. There was no associated discharge or secondary changes. Associated teeth were 25, 26 & 27 with slight palatal displacement of 26 and buccal displacement of 25. On palpation, swelling was tender, uniformly firm to hard in consistency, non-fluctuant, non- compressible, did not blanch on pressure, fixed to the underlying tissues with well-defined borders. Surface over the swelling was nodular with no discharge (Fig.1). Associated teeth 25, 26 and 27 were tender on percussion and non-vital. So, provisional diagnosis of Malignant minor salivary gland tumour IRT left side hard palate was considered. Differential diagnosis of Adenoid cystic carcinoma,

Malignant Pleomorphic adenoma, Mucoepidermoid carcinoma of left side palate and metastatic carcinoma of renal origin were included. IOPAR in the region of 25-27 revealed break in the continuity of lamina dura, widening of periodontal ligament space irt 25, 26 and 27 with destruction of floor of the maxillary sinus. (Fig.2). Maxillary left lateral occlusal view revealed a diffuse multilocular radiolucency, extending antero-posteriorly from 26 to the maxillary tuberosity and medio-laterally from palatal gingiva to mid-palatine raphe, with ill-defined borders (Fig.3). OPG revealed an ill-defined multi-locular radiolucency with larger sized locules, extending antero-posteriorly from left lateral border of the nose to the line joining maxillary tuberosity and superior-inferiorly from left infra-orbital margin to the periapical aspect of 25, 26, 27 with destruction of floor of the maxillary sinus and encroachment of the radiolucent lesion into the left nasal cavity (Fig.4). CT scan of the maxilla revealed an ill-defined radiolucent lesion (5x6x5cm) involving left posterior palate, maxillary sinus and encroachment into the left nasal cavity with destruction of cortical boundary of floor of the maxillary sinus. (Fig.5a&5b). Radiographic DDs of Clear cell odontogenic carcinoma, Ameloblastic carcinoma, Malignant minor salivary gland tumour in relation to left side hard palate, Metastatic carcinoma of renal origin was considered. All laboratory findings were found to be within normal limits. Incisional biopsy (Fig.6) was suggestive of Clear cell odontogenic carcinoma. So, after all investigation's final diagnosis of Clear cell odontogenic carcinoma in relation to 25-27 region was established. A wide surgical resection under GA(Fig.7) was performed and was given with palatal obturator (Fig.8). Regular follow up is being done. Prognosis was good with no recurrence.

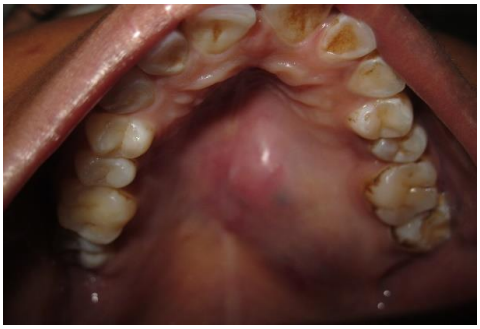

**Fig 1:** Swelling in the left side of the palate

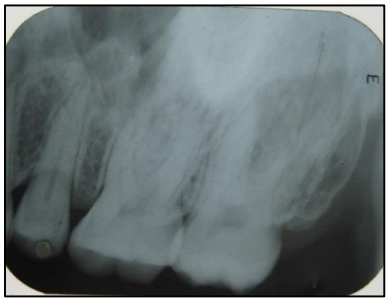

**Fig 2:** IOPAR in the region of 25-27 revealed break in the continuity of lamina dura, widening of periodontal ligament space irt 25, 26 and 27 with destruction of floor of the maxillary sinus.

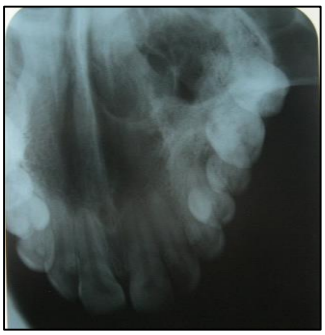

**Fig 3**: Maxillary left lateral occlusal view revealed a diffuse multilocular radiolucency, extending antero-posteriorly from 26 to the maxillary tuberosity and medio-laterally from palatal gingiva to mid-palatine raphe with ill-defined borders.

## III. Discussion:

The term 'Clear cell odontogenic tumour' was coined by Hansen et al in 1985, which is an epithelial odontogenic tumour with predominantly clear cell characteristics. Since all their cases were centrally located in the jaws, they contended that the lesion was of odontogenic origin.[5] As it has tendency to recur and undergo metastasis, the term 'Clear cell odontogenic carcinoma' is more appropriate, and is the one adopted by Reichart and Philipsen in the latest classification of odontogenic tumours approved by the WHO.[6]

Maiorano E, Altini M et al [7] and Yamamoto H, Inui M et al [1] stated the peak incidence of these tumours in the fifth to seventh decades in contradiction to our present case, which occurred in second decade of life. This tumour showed female preponderance (M/F ratio-10:17) as explained by Maiorano E,Altini M et al [7] which is similar to our present case. The most common site of occurrence is in the mandible, as explained by Yamamoto H, Inui M et al [1] and August M, Faquin W et al [8] which is contradiction to our present case, which is reported in the maxilla, similar to Elbeshir EI.Harris M et al [5] and Yazici ZM,Mete O et al.[9]

Many patients complained of lumps, pain, bony expansion or tooth abnormalities.[7, 8] Radiologically, these tumours manifested as radiolucent lesions with irregular margins.[7]Histologically, there were 3 patterns. Most commonly, these tumours show a biphasic pattern characterized by nests of clear cells intermixed with smaller islands of polygonal cells with eosinophilic cytoplasm. The second variant consists of epithelial islands exclusively composed of clear cells, while the least common variant is characterized by clear cell nests with an ameloblastomatous pattern.[7, 10]

The infiltrative growth pattern with a high recurrence rate and local metastasis makes the management of CCOC more challenging for surgeons, demanding long term surveillance. Treatment strategies includes wide surgical resection with tumour free margins or curettage, both with or without lymph node dissection, radiotherapy, and/or chemotherapy. Long term follow up, even when the excision margins have been reported as free of tumour, is necessary as recurrence and metastasis can occur years after the excision of the primary lesion.[5,11-14]

## IV. Conclusion:

CCOC has no specific clinical and radiographic signs representing a diagnostic dilemma, always the concluding diagnosis must be done with adjuvant of pathologic findings, however CCOC is not the only lesion that exhibit clear cells, and it is also seen in Pindborg tumour, ameloblastoma with a component of clear cells and other odontogenic cyst. Hence knowledge about the clinical course, histopathologic pattern, and immune profile of CCOC aids in differentiating it from other clear cell tumours.

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