

# Clear Cell Odontogenic Carcinoma Of The Maxilla: A Case Report

K Adebiyi, V Ugboko

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## Abstract

Clear cell odontogenic carcinoma (CCOC) is a rare odontogenic tumor that tends to occur in the mandible of older adults with a predilection for women. We describe a case which occurred in the maxilla and metastasized to the cervical regional lymph nodes of a 45 year old female Nigerian.

This article highlights the clinico-pathologic features of CCOC that were confirmed by histopathological examination of the case. Other secondary or primary sites of the lesion were excluded by clinical and radiological examinations. Special staining for mucin ruled out a salivary gland tumor. The aggressive potential of this neoplasm is well documented and resection with negative margins is the treatment of choice.

In view of the notable number of cases with metastasis, elective neck dissection and adjuvant radiotherapy should be considered as part of the treatment modality. The present report is an addition to the sparse literature.

## INTRODUCTION

Clear cell odontogenic carcinoma, initially designated as clear cell odontogenic tumor (CCOT) was first described as a benign separate entity by Hansen et al [1] in 1985. They described it as a benign but locally aggressive epithelial neoplasm of odontogenic origin. The World Health Organization classification of odontogenic tumors [2] also defined CCOT as a benign but locally invasive neoplasm of odontogenic epithelium characterized by sheets and islands of uniform, vacuolated and clear cells. Subsequent reports in the literature [3, 4] indicated that the tumor has the potential to metastasize and current opinion is that it should be designated as clear cell odontogenic carcinoma (CCOC).

This additional report to the English literature documents the clinicopathologic features of a rare case which occurred in the maxilla.

## CASE REPORT

A 45 – year old female Nigerian presented at the Maxillofacial Unit of the Obafemi Awolowo University Teaching Hospital, Ile-Ife, Nigeria with a six months history of left maxillary swelling that had gradually increased in size. There was no history of trauma but associated left nasal discharge. This was the first episode of such a swelling and

the family history as negative. The systemic review was also non-contributory.

Extra oral examination revealed a firm to bony hard left maxillary swelling measuring about 8 X 10 cm (Fig 1).

### Figure 1

Figure 1: Shows left maxillary swelling in a 45-year-old female.



There was paraesthesia over the region supplied by the left infraorbital nerve. The upper left cervical lymph nodes were

palpably enlarged, bony hard and fixed to the underlying structures. Intra orally, there was a pinkish fleshy mass with buccal and palatal expansion of the maxilla extending from the upper left first premolar to the third molar on the same side (Fig 2). Paraesthesia of the overlying mucosa with no area of ulceration was also observed. The related teeth were mobile. Based on the foregoing an impression of mucoepidermoid carcinoma of the maxillary antrum was made.

**Figure 2**

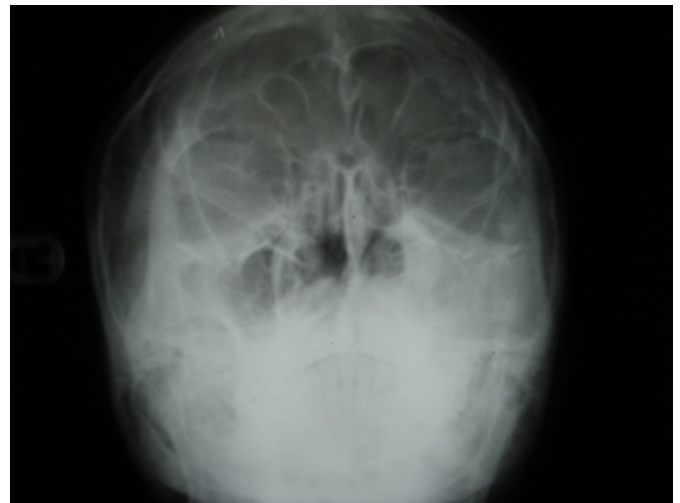
Figure 2: Intra oral view of the same patient showing pinkish fleshy mass with buccal and palatal expansion of the maxilla



Plain radiographs showed multiple irregular osteolytic lesions of the left maxilla and zygoma with opacity of the maxillary antrum on the affected side (Fig 3).

**Figure 3**

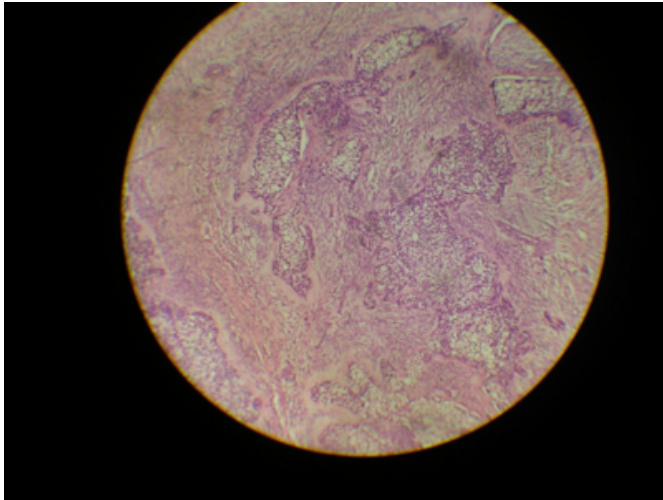
Figure 3: Occipitomeatal view of the skull shows multiple irregular osteolytic lesions of the left maxilla and zygoma with opacity of the maxillary antrum on the affected side



Abdominal ultrasound of the kidneys, liver and spleen were essentially normal. Hematological and biochemical parameters were within normal limits. The patient subsequently had an incisional biopsy under local anaesthesia that was reported as clear cell odontogenic carcinoma of the maxilla. Histopathological examination of the specimen revealed irregularly shaped tissue mass with a stratified squamous epithelium. Solid sheets, islands and nests of neoplastic clear epithelial cells separated by fibrous connective tissue septae densely infiltrate the stroma. No evidence of ameloblastic differentiation was observed. Majority of the cells were isomorphic with well-defined outlines. Prominent atypia and mitotic figures were absent (Figs 4a & 4b). The tumor cells were negative for alcian blue but PAS stain for glycogen was positive.

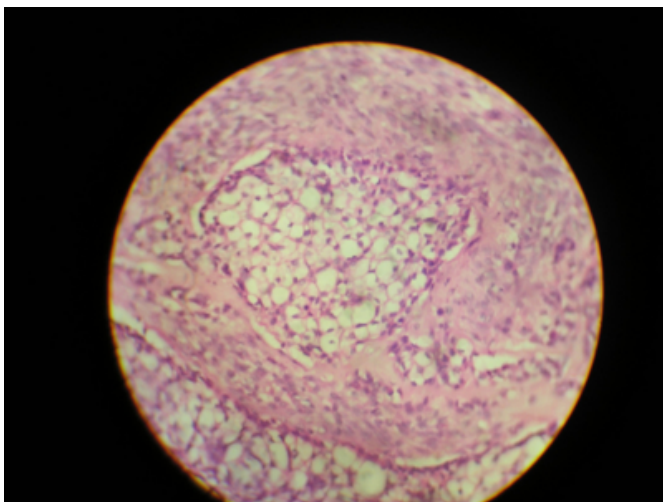
### Figure 4

Figure 4a: Shows sheets and cords of the neoplastic clear epithelial cells (H&E X80)



### Figure 5

Figure 4b: Shows well defined outlines of the cells and the fibrous septa separating the islands (H&E X200)



The patient was scheduled for a hemimaxillectomy and subsequent radiotherapy but declined and requested that she be referred to another centre where it will be convenient for family members to adequately attend to her needs while receiving treatment.

## DISCUSSION

CCOC is a rare odontogenic tumor with female predilection and peak age incidence in the 5<sup>th</sup> to 7<sup>th</sup> decades of life. This is consistent with the demographic characteristics of our patient.

In contrast with the case being reported where the lesion occurred in the posterior region of the maxilla, studies have

shown that the anterior portions of the jaws especially the mandible are most frequently affected [5, 6].

Generally, the histologic feature is that of solid epithelial islands and nests and strands of clear and basaloid cells with scanty eosinophilic cytoplasm that are separated by septae of fibrous connective tissue. They are unencapsulated and show aggressive infiltration of the surrounding tissues [5, 7]. Braunshtein et al [5] reviewed the literature and found 27 cases of CCOC reported worldwide that showed a high rate of recurrence (50%) and metastasis (33%). Kumar et al [4] also documented a case which metastasized to the 5<sup>th</sup> lumbar vertebra and hip 3 years after the initial diagnosis, thus emphasizing the need for long-term follow up. Local recurrence is a common finding and cytologic atypia appears to worsen with recurrent lesions [7].

Three histomorphologic patterns have been described [8] with the commonest a biphasic tumor characterized by oval and linear nests of clear cells intermixed with smaller islands of polygonal cells with eosinophilic cytoplasm. Occasionally these two cell-types co-exist in a tumor nest yielding a “glomeruloid” appearance. The second variant is represented by islands that show only the clear cell phenotype whereas the third and least common variant is comprised of clear cell nests with a tendency for ameloblastoid palisading around the periphery. The case being reported is typical of the second variety. The presence of eosinophilic hyaline fibrillar dentin/bone like structures between tumor cell nests and fibrous stroma also suggests that some of these tumors possess epithelial-mesenchymal inductive capacity, a feature shared by many odontogenic epithelial tumors [9].

It is noteworthy that ultrasound evaluations of the liver, kidneys and spleen did not reveal any metastatic lesions because tumors with 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 the kidneys. Even additional special staining for mucin is recommended to rule out mucoepidermoid carcinoma. Furthermore, CCOC can be distinguished from the clear cell variant of calcifying epithelial odontogenic tumor because it lacks the characteristic calcification and amyloid deposition.

There are no definite universal treatment protocols for CCOC in the literature probably due to the recurrences and/or metastasis that have been recorded following aggressive surgery. Hence, CCOC should be regarded as a

low-grade malignancy with capacity for sinister biologic behavior. Treatment should be by wide surgical resection with at least 1cm of tumor-free margins. In addition, elective neck dissection and adjuvant radiotherapy is suggested.

### CORRESPONDENCE TO

Dr Kehinde E. Adebiyi Department of Oral/Maxillofacial Surgery & Oral Pathology Faculty of Dentistry, Obafemi Awolowo University, Ile-Ife, Nigeria. E-mail: kenad@justice.com

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**Author Information**

**Kehinde Adebisi, FWACS**

Department of Oral/Maxillofacial Surgery & Oral Pathology, Faculty of Dentistry, Obafemi Awolowo University

**Vincent Ugboko, FMCDS, FWACS**

Department of Oral/Maxillofacial Surgery & Oral Pathology, Faculty of Dentistry, Obafemi Awolowo University