

# CASE REPORT

## CLEAR CELL ODONTOGENIC CARCINOMA: A RARITY

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### ABSTRACT

**Background:** Clear cell odontogenic carcinoma is an unusual tumor in the buccal cavity, mandible or maxilla. There is slight female preponderance and the peak age of occurrence is in the 6th and 7th decades of life. It shows loco-regional and distant metastasis thereby resulting to great challenges in diagnosis and treatment.

**Case presentation:** We hereby present a case in a 67-year-old woman with a case of right maxillary swelling with extension to gingival labial, central and lateral incisors, and an ipsilateral cervical lymph node enlargement. The lesion was radiolucent on x-ray with associated loss of periodontium around the teeth and erosion of the maxillary bone. The patient had an incisional biopsy and has since been lost to follow-up.

**Conclusion:** One of the differential diagnoses that ought to be considered in a jaw tumor with a prominent clear cell differentiation is a clear cell odontogenic carcinoma. Radical resection is advocated as a mode of treatment more especially when there is an evidence of local or distant metastasis because conservative resection or tumor curettage has been known to be associated with a high recurrence rate. Some of the cases may merit lymph node dissection and adjuvant radiation therapy.

### Background

Clear cell odontogenic carcinoma (CCOC) is not a very common tumor in oral cavity, maxilla or mandible. As at December 2011, less than 70 cases were described in the English literature and complete understanding of the behavior of CCOC was based on limited case reports [1]. We report a case of CCOC in the maxilla of a 67-year-old female with associated lymph node metastasis.

### Case presentation

We present the case of a 67-year-old woman who started complaining of a painful mass on the right side of her jaw at about 6 months prior to presentation to the Dental/ Maxillofacial Surgery

Department of the Federal Medical Centre Makurdi, Benue State. The lesion started as a small nodular out-pouch in the buccal cavity which gradually increased in size. However, about a month before her visit to the clinic, she noticed a sudden growth sprout. A clinical diagnosis of fibrous epulis was entertained at her first visit to the clinic. Some of the teeth (central and lateral incisors on the right maxilla) adjacent to the swelling were loose and one of them became carious at a time and subsequently fell off. Physical examination of the lesion showed an 8 × 7 × 5 cm irregular, firm and slightly tender swelling in the right mandible. The tumor bulged as a pinkish mass when viewed intra-orally. A lymph node was palpable on the lateral aspect of right side of the neck and it measured approximately 2.5 × 2cm. There was also no noticeable loss of sensation over and around the swelling. Radiological examination of the jaw revealed a radiolucent lesion with irregular margins of right alveolar and palatine bone. The maxilla bone demonstrated much tissue loss with severe motility of the segment. Computed tomography (CT) scan of the oral cavity and neck could not be performed owing to financial constraint. A clinical diagnosis of a malignant lesion with a possibility of a cervical lymph node involvement was made.

### Histopathological review

Histologic sections show a malignant epithelial neoplasm composed of mild to moderate pleomorphic cells with clear, foamy and moderate eosinophilic cytoplasm which are separated into irregular nests by broad bands of fibrocollagenous stroma. The columnar cells adjacent to the fibrocollagenous septa appear vaguely palisaded. The cells in the centre of the nests appear large with

abundant clear cytoplasm (**Figures 1 and 2**). Some of the nuclei have coarse chromatin pattern with prominent nucleoli. The overall features are in keeping with a clear cell odontogenic carcinoma. Immunohistochemistry was not done on this case but we subjected the Hematoxylin and Eosin stained slide to extensive review and all the nine persons all of whom are experienced histopathologists independently agreed to the same diagnosis of CCOC.

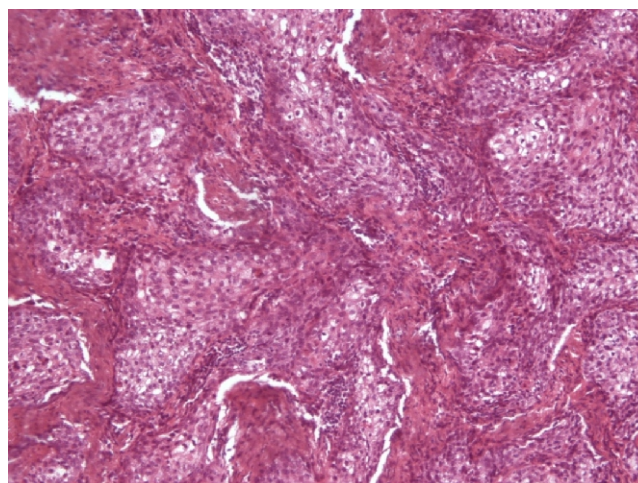
### Discussion

In 1985, Hansen *et al* named a locally aggressive odontogenic neoplasm a clear cell odontogenic tumor [2]. This tumor was classified as benign in the initial report [3]. With emerging and overwhelming evidence supporting tumor aggression, local recurrence, and evidence of pulmonary and lymph node metastases and tumor-related deaths, the tumor was re-defined and re-classified as Clear Cell Odontogenic Carcinoma (CCOC) [4-8]. The usual clinical presentation of CCOC is a painful anterior mandibular swelling in an elderly woman [9]. There may be associated teeth loss and the X-ray findings often show an irregular radiolucent mass, most often in a premolar location [9]. These features are present in this index case. The patient was 67-year-old woman who complained of a painful mass on the right side of her jaw. Physical examination revealed that her central and lateral incisors on the right maxilla, adjacent to the swelling were loose and one of them subsequently fell off. In this case, the tumor is located in the maxilla, whereas the prevalent localization is in the anterior segments of the jaws; the mandible is often affected much more commonly than the maxilla [9, 10].

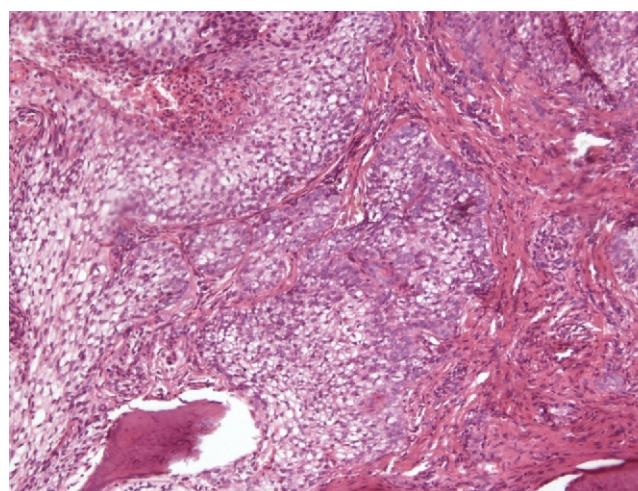
There are three histologically patterns in the literature [11]. The commonest pattern is biphasic in nature, comprising an admixture of cell nests and smaller islands of polygonal cells with eosinophilic cytoplasm characterizing it. The commoner histologic pattern is composed of epithelial islands exclusively made up of clear cells, while the least common variant comprises predominantly clear cell nests with an ameloblastomatous pattern [9]. This index case is composed of admixture of cell nests in intimate relationship with polygonal cells having abundant eosinophilic cytoplasm without ameloblastic differentiation. The case report does not differ from those previously reported in the literature. The histochemical, immunohistochemical, and ultra-structural features of CCOCs, with and without ameloblastic

differentiation were described anecdotally in some literatures [9]. One of the most consistent features of CCOC is glycogen storage [9, 12-16]. However, the use of immunohistochemistry in confirming the diagnosis is controversial. Immunohistochemistry is however not useful in differentiating metastatic clear cell carcinoma (CCC) from CCOC but it is useful in isolating melanoma involving the jaw [17, 18]. The use of histochemical stains is useful in diagnosing amyloidosis.

Based on previously published data and our own experience, the diagnosis of CCOC is usually made by exclusion of other tumors, and we still lack histologic, immunohistochemical, and ultrastructural features to specifically identify this entity [9].



**Figure 2:** Clear cell odontogenic carcinoma is composed of irregular nests of neoplastic epithelial cells separated into nodules by a richly cellular, collagenous stroma. The neoplastic epithelial cells are polyhedral and show distinct cytoplasmic clearing in the center of the neoplastic sheets (H&E, ×4).



**Figure 3:** A higher magnification (X40) of Clear cell odontogenic carcinoma showing sheets of clear cells embedded in a dense fibrous stroma. (H&E)

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