Case Report

# Clear cell odontogenic carcinoma: A rare case

**Garima Jain, Padmaraj Hegde[1], Pushparaja Shetty**

Departments of Oral Pathology and Microbiology and [1]Oral and Maxillofacial Surgery, A. B. Shetty Memorial Institute of Dental Sciences, Deralakatte, Mangalore, Karnataka, India

## ABSTRACT

Clear cell odontogenic carcinoma is a rare neoplasm with very few cases reported in the literature. We report a case of a 50-year-old female patient with the malignancy at a less common location. Diagnosis was given based on the histopathologic findings. The demographic data and understanding for this tumor needs to be strengthened by reporting all new cases, which are diagnosed, in literature.

**Key words:** Clear cell, malignancy, odontogenic

## INTRODUCTION

Clear cell odontogenic carcinoma (CCOC) is a very uncommon neoplasm, and few cases have been reported in English-language literature. According to a recent paper by Kalsi *et al*., only 74 such cases have been described so far.[1] In addition, odontogenic tumors with a significant number of clear cells are rare.[2] First described by Hansen *et al*. in 1985, CCOC was initially considered as benign, but locally invasive.[3] Later, World Health Organization in 2005 reclassified it as a malignant tumor of odontogenic origin, owing to its aggressive nature, infiltrative behavior, local recurrence tendency, lymph node and distant metastasis, especially pulmonary.[3] The authors hereby report this rare case.

## CASE REPORT

A 50-year-old female patient reported to hospital, with the chief complaint of swelling and a nonhealing ulcer in the left upper jaw since 2 weeks. Onset of the lesion was insidious, although the patient relates the increase in size of the lesion after extraction of teeth from left maxillary posterior region, 1-week prior to the complaint. According to the patient, all the teeth in that area were grossly decayed and nonmobile. Patient had a habit of pan chewing since 1-year. There was no relevant family or medical history, and physical examination was noncontributory.

On extraoral examination, there was a diffuse swelling of 4 cm × 3 cm approximately on the left side of the face, making the extraoral appearance asymmetrical. This swelling was mainly present over maxilla and zygomatic arch, extending below up to the corner of the mouth and medially 1 cm away from the ala of nose. Posterior extension was up to the lateral canthus of the eye. None of the lymph nodes were palpable.

On intraoral examination, a swelling was present on the maxillary ridge which was approximately, 5 cm × 5 cm and extending in the region of 25 to 28. The intraoral lesion was pink red in color and ulcero-proliferative [Figure 1]. Lesion appeared to be intrabony, was tender on percussion and indurated.

Orthopantomograph showed an osteolytic area extending from the distal aspect of 25 to 28 region [Figure 2]. This area was covered with a slightly radiopaque soft tissue shadow. Paranasal sinus view revealed haziness of the left maxillary sinus.

There were no known primary tumors in any other part of the body. Considering a clinical differential diagnosis of squamous cell carcinoma of the maxilla, an incisional biopsy was performed. Histopathological examination showed epithelium and minimum connective tissue. Epithelium consisted of basal columnar and squamous cells; and suprabasal polygonal cells [Figure 3]. Suprabasal cells showed a biphasic pattern containing both clear cells and more hyperchromatic appearing polygonal cells [Figure 4].

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**Address for correspondence:** Dr. Garima Jain, Department of Oral Pathology and Microbiology, A. B. Shetty Memorial Institute of Dental Sciences, Deralakatte, Mangalore - 575 018, Karnataka, India. E-mail: id- drgarimajain7@gmail.com

Epithelial islands were separated by zones of mature, fibrous and partially hyalinized connective tissue stroma [Figure 3]. Some epithelial cells showed highly dysplastic features like nuclear hyperchromatism, increased nuclear-cytoplasmic ratio and abnormal mitotic figures.

A diagnosis of Clear cell odontogenic carcinoma (CCOC ) was made.

## DISCUSSION

Clear cell odontogenic carcinoma is an epithelial neoplasm of odontogenic origin which is defined as a benign, but locally invasive neoplasm characterized by sheets and islands of uniform, vacuolated, and clear cells.[4] It usually occurs in the fifth to the seventh decade, has female predilection and mostly located in the anterior region of the mandible.[1,2,5] Other common clinical features are jaw enlargement and mild pain in the affected area, which were present in the present case.

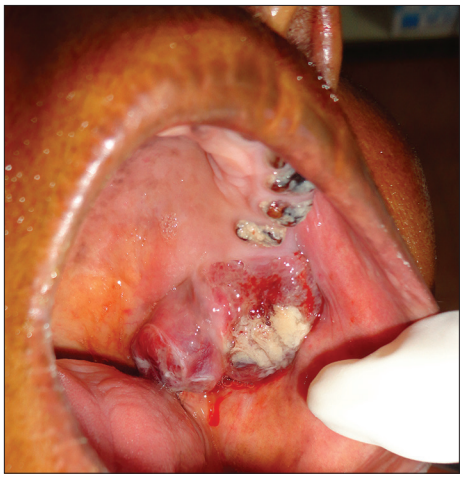

**Figure 1:** Intraoral finding: Swelling on maxillary posterior ridge, ulceroproliferative

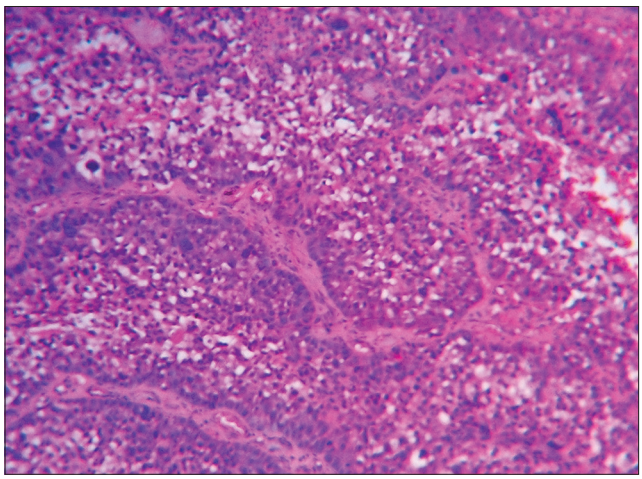

**Figure 3:** Photomicrograph shows abundant odontogenic epithelial islands separated by zones of mature, fibrous and partially hyalinized connective tissue stroma (H and E, ×100)

The present case was unusual as clear cell containing odontogenic tumors are rare, and very few cases are reported to involve posterior maxilla. Most of the authors have reported this disease entity in mandible with few exceptions like Elbeshir *et al*. who reported one case in posterior maxilla, and Kalsi *et al*. and Iezzi *et al*. who reported CCOC in anterior maxilla.[1,3-9] Other odontogenic tumors containing clear cells are clear cell variant of calcifying odontogenic tumor and clear cell ameloblastoma.[4] Clear cell component can be expected in odontogenic tumors, as they arise from dental lamina.[3]

Clear cells are seen in any lesion due to intracellular accumulation of nonstaining compounds, that is, glycogen, lipid, mucin; a scarcity of cell organelles or artefact induced during tissue fixation or processing.[3] Other clear cell containing lesions may be salivary gland tumors, metastatic renal cell carcinoma, melanocytic tumors, and odontogenic epithelial linings in periodontal cysts.[3]

"Clear cell odontogenic carcinoma exhibits three histologic patterns: Biphasic, monophasic, and ameloblastomatous."[3] The present case had a biphasic pattern like most such tumors, with nests of clear cells among small islands of hyperchromic polygonal cells with cytoplasmic eosinophilia. These findings were similar to the findings

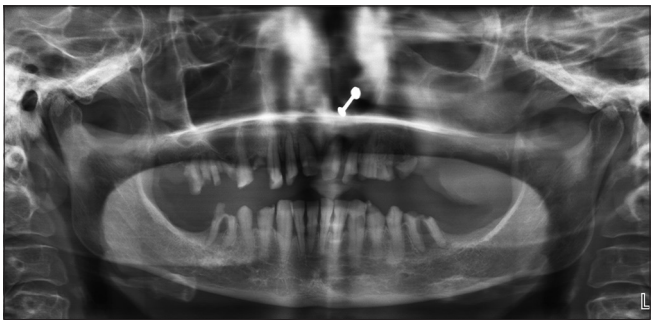

**Figure 2:** Orthopantomograph shows osteolytic area extending from distal aspect of 25 to 28 region (covered by soft tissue shadow)

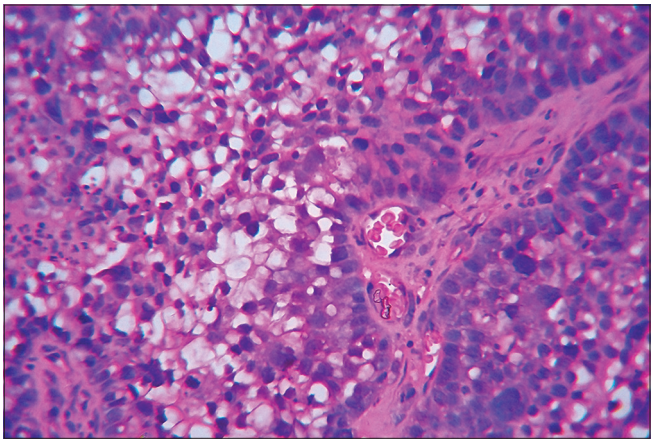

**Figure 4:** Photomicrograph shows basal columnar and squamous cells; and suprabasal polygonal cells in biphasic pattern with clear cells and hyperchromatic polygonal cells (H and E, ×400)

in other cases reported by Kalsi *et al*., Xavier *et al*. and Braunshtein *et al*.[1,3,4] In monophasic pattern, presenting islands consist entirely of clear cells. The ameloblastomatous pattern is characterized by clear cells arranged in nests within networks of follicles.[3,6]

Nuclear pleomorphism and hyperchromatism, and the number of mitoses are quite variable.[4] The present case showed all these features. Furthermore, these lesions are rarely encapsulated and show frequent invasion into the medullary bone, muscle, and neural tissues.[3] The abnormal cells can surround the periphery of the tumor islands or may form ductal structures.[3] Solid cellular aggregates traversed by dense fibrous septa may be usually found and were also seen in the present case.[2]

Mucicarmine staining in the present case showed negative results and thus the salivary gland tumors were ruled out. The histopathology showing biphasic pattern of cells and separation of islands by mature fibrous stroma, were similar to that found by Yamamoto *et al*. and Xavier *et al*., and helped to identify the present case as CCOC.[3,7,8]

Ultrastructural characteristics of epithelial differentiation, glycogen pools, paucity of organelles, and the absence of myoepithelial elements may help in the identification of clear cell component and thus CCOC.[3] Positive immunohistochemical staining for cytokeratins 8, 13 and 19; as well as occasional positivity for epithelial membrane antigen, filaggrin, and antiameloblastoma antigen have also been reported.[5,10] These tumors are usually negative for S-100 protein, smooth muscle actin, glial fibrillary acidic protein, vimentin, and involucrin.[10]

Clear cell odontogenic carcinoma shows a recurrence rate of 34% after resection. Management of CCOC is done by resection with a wide margin and long-term follow-up is required.[9]

## CONCLUSION

As CCOC is a rare lesion, and very few cases have been reported in the literature, the clinical, histologic and immunohistochemical properties of this tumor are not fully understood. The demographic data for this tumor needs to be strengthened by reporting all new cases, which are diagnosed, in literature. Also, it should always be considered as a differential diagnosis in jaw tumors with a clear cell component.

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