Case Report

Clear cell odontogenic carcinoma of maxilla: A rare case report

ABSTARCT

Clear cell odontogenic carcinoma is a rare, infrequent, aggressive in nature, locally reoccurring odontogenic tumor with a tendency of distant metastasis, occurring during to 4th to 6th decades with a mostly female predilection. Histologically, it is characterized by sheets and islands of vacuolated/clear cells. Till date, only 74 cases have been reported in the literature. We present a case of 45-year-old woman with a tumor mass extending from the maxillary right first premolar till the third molar region measuring 4 cm × 4 cm. The diagnosis was given based on the histopathological findings. Being locally aggressive, the reported data and understanding of this infrequent tumor needs to be strengthened by reporting new cases, and it also demands to be distinguished from other primary and metastatic clear cell tumors of the head-and-neck region.

Keywords: Ameloblastoma, clear cell odontogenic carcinoma, Weber-Ferguson approach

INTRODUCTION

In 1985, clear cell odontogenic carcinoma (CCOC) was first described by Hansen *et al*., and Waldron *et al*. Considering its locally aggressive nature, it was termed clear cell odontogenic tumor.[1] It was defined by the WHO as a benign neoplasm with a capacity for locally invasive growth, and was considered more aggressive than ameloblastoma. In the year 1992 and in 2003, Reichart and Philipsen proposed a revision of the classification, clearly considering the clear cell odontogenic tumor as a carcinoma.[2] Although initially being thought to be benign, because of their aggressive behavior, tendency for local recurrence, regional lymph node metastasis and distant metastasis, the WHO classification of odontogenic tumors was revised in 2005, and CCOC was denoted as a malignant tumor of odontogenic origin.[3] The authors hereby report this rare tumor of the head-and-neck region.

CASE REPORT

A 45-year-old woman reported to the Department of Oral and Maxillofacial Surgery, Subbaiah Institute of Medical and Dental Sciences, Shivamogga, India, with a 5 months history of painless swelling in the right maxillary region. On inspection, the swelling was noted in the right maxillary region extending from the first premolar till the third molar measuring 4 cm × 4 cm with buccal cortical plate expansion. The swelling was smooth surfaced, mucosal colored with no signs of ulceration [Figure 1]. On palpation, the swelling was nontender, nonfluctuant, and firm inconsistency. Grade II mobility of all the five teeth were noted. Bilateral cervical lymph nodes were not palpable. Contrast computed tomography scan revealed a well-delineated unilocular

Anubhav Jannu, Gautam Rao, Mithun Kulambi[1], Arun Kumar Talkal[2], M. S. Suma[3], K. K. Deepa[4], Rachana Srinivas[5]

Departments of Oral and Maxillofacial Surgery, [1]Oral and Maxillofacial Pathology, [2]Prosthodontics, [3]Oral Medicine and Radiology, [4]Oral Pathology, and [5]Pediatric Dentistry, Subbaiah Institute of Medical and Dental Sciences, Shimoga, Karnataka, India

Address for correspondence: Dr. Anubhav Jannu, Department of Oral and Maxillofacial Surgery, Subbaiah Institute of Medical and Dental Sciences, Shimoga - 577 004, Karnataka, India.
E-mail: anubhavjannu@gmail.com

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radioopaque mass extending from maxillary right premolar till the posterior aspect of maxillary tuberosity region involving the maxillary sinus and the nasal septum [Figure 2]. A provisional diagnosis of intraosseous odontogenic tumor/ameloblastoma/pleomorphic adenoma was made. The patient's personal, family, and medical histories were noncontributory. An incisional biopsy under local anesthesia was performed after all the hematological tests were within the normal limits, and no other systemic abnormalities were observed.

Microscopically, sections stained with H and E revealed sheets and islands of large cells separated by a delicate fibrous connective tissue stroma. The cells at the periphery of the nests occasionally demonstrated nuclear palisading away from the basement membrane. The differential diagnosis that was considered included clear cell ameloblastoma, CCOC, intraosseous clear cell salivary gland tumor, mucoepidermoid carcinoma. Tumor cells were periodic acid–Schiff positive and diastase sensitive, thus confirming the glycogen content of the cytoplasm. The cells did not show positivity for alcian blue, ruling out mucoepidermoid carcinoma [Figure 3]. Based on the radiologic, histopathologic, and histochemical findings, a diagnosis of CCOC was made.

The patient underwent right hemimaxillectomy through the Weber-Ferguson approach under general anesthesia [Figure 4]. The tumor was resected with the help of Straight handpiece and straight bur. It was later detatched from the normal tissues through chisel and Mallet [Figure 5]. On examination, a tumor measuring 4 cm × 4 cm was found in the right maxilla. The tumor was firm in consistency with all five teeth present. It was seen to involve the underlying bone along with the involvement of the right nasal septum and maxillary sinus regions. Postoperative histopathologic examination showed a tumor with morphology similar to that in the incisional biopsy. The diagnosis of CCOC was confirmed. All the resected margins were free of tumor. The postoperative period was uneventful, and the patient did not show any recurrence of tumor in 1 year of follow-up.

DISCUSSION

CCOC is a rare, aggressive, locally reoccurring odontogenic tumor with a tendency of distant metastasis, occurring during to 4th to 6th decades with a mostly female predilection. It was formerly known as clear cell odontogenic tumor as described by Hansen *et al*. and Waldron *et al*. in 1985.[1] The WHO described it as a benign neoplasm with a capacity for locally invasive growth, and was considered more aggressive than ameloblastoma.[3,4] Clinically, it presents as asymptomatic, slow-growing swelling of long duration with a mean size of 4 cm diameter. Most of the patients present with gingival swelling and loosening of teeth, other manifestations include delayed healing of wound after extraction of teeth, pain and bleeding.[5,6] Radiologically,

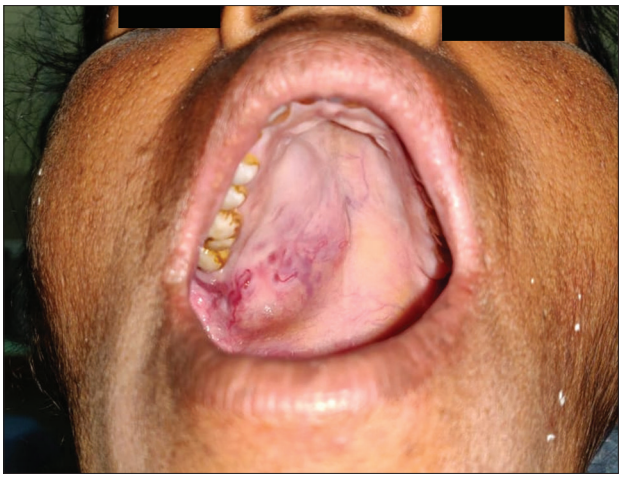

Figure 1: Swelling in the right maxillary region

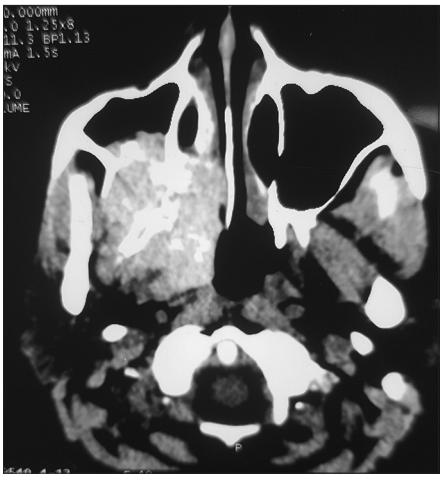

Figure 2: Computed tomographic image of the tumor

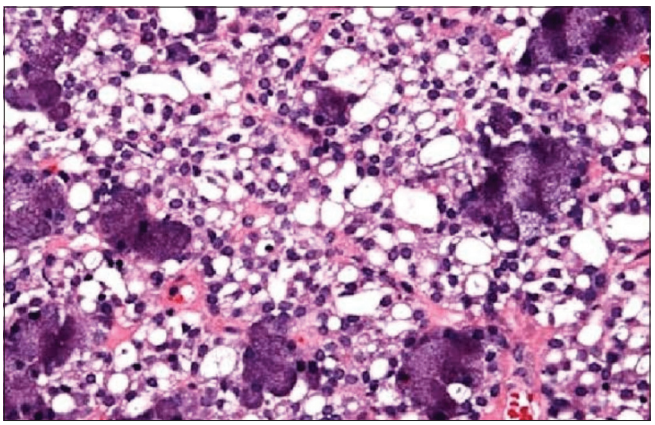

Figure 3: Histopathologic image

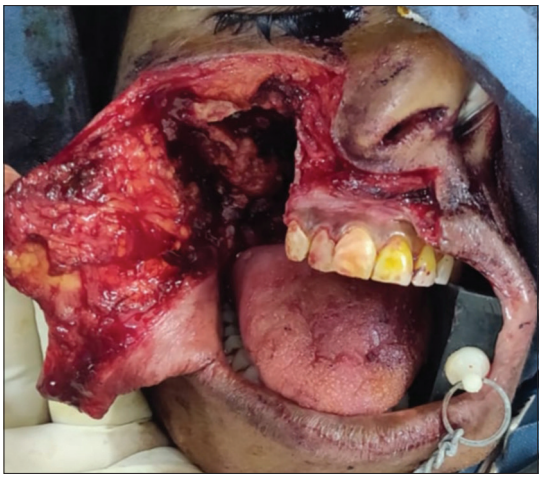

Figure 4: Right hemimaxilletomy through Weber-Furguson approach

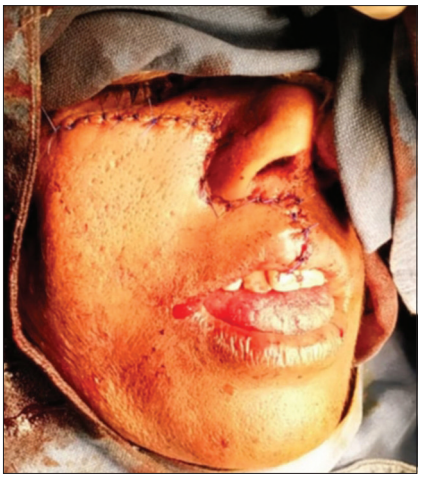

Figure 5: Closure

the tumor is manifested as an ill-defined radiolucency with irregular margins. Aggressive tumor growth often results in root resorption.[6]

Clear cells are seen in any lesion due to intracellular accumulation of nonstaining compounds such as glycogen, lipid, mucin, a scarcity of cell organelles or artifact induced during tissue fixation or processing.[7]

CCOC exhibits three histological patterns: Biphasic, monophasic, and ameloblastomatous.[3,6,7] Most frequently seen is the biphasic pattern with islands of epithelial cells in a fibrous stroma. The cells have eosinophilic cytoplasm due to the presence of cytoplasmic glycogen, well-demarcated cell membranes, and irregular nuclei. The monophasic pattern comprises only of clear cells while the ameloblastomatous pattern resembles the growth pattern of ameloblastoma with nests of cells showing central cystic change and squamous differentiation, and peripheral nuclear palisading with reverse polarity.[8]

A recent review of literature[9] suggests that CCOC has the potential for multiple recurrences (41%), metastasis (31%), thus demanding an aggressive treatment protocol and a long-term follow-up. Surgical resection with wide margins is the treatment of choice for CCOC. For cases showing perivascular and perineural invasion, adjuvant radiotherapy is necessary. Conservative surgery showed a higher recurrence rate as compared to those treated with surgical resection (86.7% vs. 29%).[10-13]

CONCLUSION

CCOC is a rare, infrequent, aggressive in nature, locally reoccurring odontogenic tumor with a tendency of distant metastasis. The clinical, histologic, and immunohistochemical properties of this tumor are not fully understood, as very few cases have been reported in the literature. Currently, treatment depends on the size of the lesion, location, soft-tissue involvement, and nodal or distant metastasis. Treatment is aimed at achieving wide surgical resection with tumor-free margins and local radiation in cases with extensive soft tissue invasion, perineural spread, lymph node metastasis with extranodal involvement, or in those where tumor-free margins are not possible. As these tumors may recur locally or present with late distant metastasis, a long-term follow-up is essential.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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