

# Clear cell odontogenic carcinoma of the mandible and temporomandibular joint with cervical lymph nodal metastasis

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## ABSTRACT

Clear cell odontogenic carcinoma (CCOC) is a rare aggressive tumor that has the ability to invade locally as well as cause regional and distant metastasis. The etiology of this neoplasm remains poorly understood and the diagnosis of CCOC is done by exclusion of other clear cell tumors. To date, approximately 75 cases of CCOC have been described in the English literature, all involving a single jaw. The majority of cases have been reported to arise from the mandible, the maxilla is less frequently involved, and no case had involvement of the temporomandibular joint. Lymph node metastasis at initial presentation is reportedly rare (<10%). We describe possibly the first case of CCOC in a 50-year-old woman with involvement of the mandible and the temporomandibular joint along with cervical lymph nodal metastasis. We share our experiences and challenges in the management of this unusual tumor.

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## INTRODUCTION

Clear cell odontogenic carcinoma (CCOC) is an extremely uncommon tumor of odontogenic origin, first described by Hansen in 1985.<sup>[1]</sup> CCOCs were formerly called clear cell ameloblastomas or clear cell odontogenic tumors and were classified under the category of benign tumors in the 1992 World Health Organization classification. However, owing to its high potential for regional spread and distant metastases, it was reclassified as a malignant tumor of odontogenic origin in 2005. To date, of the reported cases, all have presented in a single jaw, with none having TMJ involvement. We report, to the best of our knowledge, the first case of an aggressive CCOC involving the mandible and the temporomandibular

joint (TMJ) with cervical lymph node metastasis at presentation.

## CASE REPORT

A 50-year-old female was referred our institution for evaluation of a swelling in the left mandible of 3 months' duration. She gave a history of extraction of a mobile left lower third molar one month ago elsewhere. The patient's past medical history and family history were unremarkable. On clinical examination, there was an extra oral swelling overlying the left posterior mandible measuring 4 × 5 cm. Intraorally, the swelling extended from the region of the left first molar to the ascending ramus of the mandible, obliterating the left retromolar trigone. The swelling was well demarcated and firm on palpation. The patient was also found to have Grade 1 trismus and numbness of the left tongue. Examination of the neck revealed a significant, firm, submandibular lymph node approximately 1.0 cm in diameter. Orthopantomogram and computed tomography scans revealed an expansile lytic lesion involving the posterior

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body and ramus of the left mandible [Figure 1]. There was perforation of both the buccal and lingual cortical plates and involvement of the adjacent soft tissues. The lesion extended into the left temporomandibular joint. An intraoral trucut biopsy was performed, and a diagnosis of CCOC was established. A chest skiagram and ultrasound of the abdomen, done as a part of the metastatic work-up, were normal.

The patient then underwent a composite resection which included an en bloc excision of the posterior segment of the left mandible, TMJ, infratemporal fossa clearance, and a left modified radical neck dissection [Figure 2]. Reconstruction of the defect was performed using a pectoralis major myocutaneous flap.

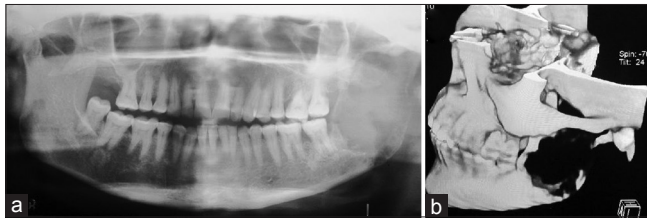
The gross pathology revealed a firm tumor measuring 5 × 3 × 4.5 cm involving the mandible, TMJ, and the adjoining soft tissues. Microscopically, tumor cells were seen arranged in sheets, cords and islands, separated by thin and thick bands of fibrocollagenous stroma with hyalinization. The tumor cells were round to polygonal with well-defined cell outlines and abundant clear cytoplasm. There was mild pleomorphism with centrally

or peripherally placed nuclei and small nucleoli. Mitotic figures were not well made out. Larger islands of tumor showed central necrosis. Focal perineural invasion was also seen. The tumor cells were immunoreactive for keratin, epithelial membrane antigen, smooth muscle actin, and S-100 and negative for vimentin, and HMB-45 [Figures 3 and 4]. The Ki-67 proliferative index was approximately 10%. Two out of the 34 left neck nodes revealed metastatic deposits of CCOC.

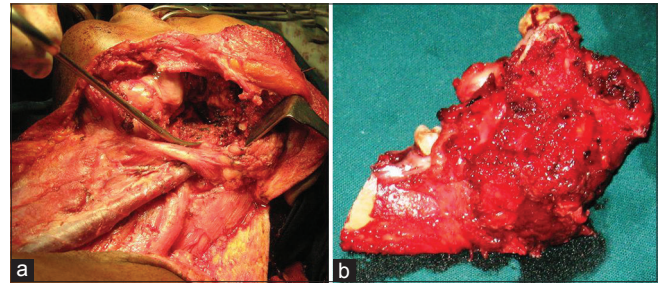
Adjuvant external beam radiotherapy was advised in view of the large erosive tumor with perineural infiltration and the presence of cervical lymph node metastasis. The patient subsequently received 40 Grey of external beam radiotherapy to the tumor bed and neck. She remains asymptomatic and is disease free for more than 2 years.

## DISCUSSION

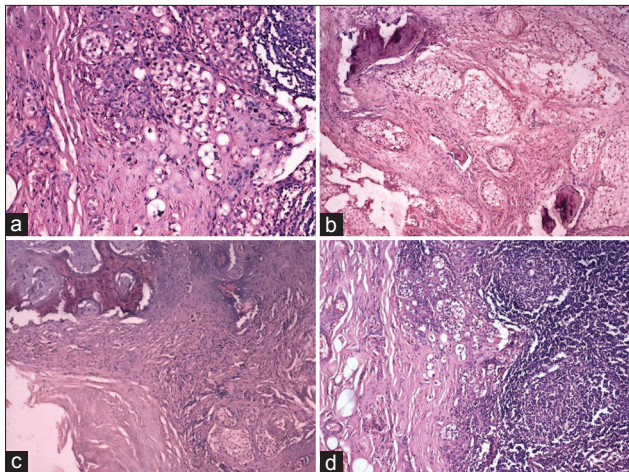
CCOC is presently designated as a malignant tumor of odontogenic origin. Insight into the behavior of this tumor is based on a limited number of case reports. The peak incidence is reported to occur in the fifth to seventh



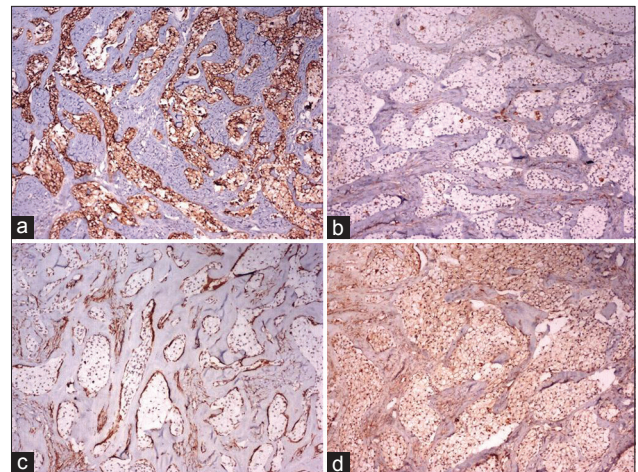
**Figure 1:** (a) OPG showing a radiolucent lesion in the posterior segment of the left hemi-mandible. (b) Reconstructed axial CT scan showing the lytic lesion along the left angle and ascending ramus of the mandible and involvement of the temporomandibular joint



**Figure 2:** (a) Intraoperative clinical photograph following composite resection (b) Postoperative clinical photograph of the operated specimen



**Figure 3:** H and E X10 – (a) The tumor cells were round to polygonal with well-defined cell outlines and abundant clear cytoplasm. There was mild pleomorphism with centrally or peripherally placed nucleus and small nucleolus. (b) Tumor showing infiltration of the mandible. (c) Tumor cell infiltrating the temporomandibular joint. (d) Metastatic tumor deposits in the cervical lymph nodes



**Figure 4:** IHC X100 – (a) Tumor cells showing strong immunopositivity to keratin. (b) Tumor cells showing weak immunopositivity to EMA. (c) Tumor cells showing weak immunopositivity to SMA. (d) Tumor cells showing mild-to-moderate immunopositivity to S-100

decades of life with a female preponderance. (1.62: 1) The mean age at presentation was 54.45 years (range 17–89 years). The most common presenting sign was as a painless swelling of the jaw, often with mobility of the teeth in that region. Palpable lymph nodes on initial presentation were noted in about 10% of the reported cases.<sup>[2-4]</sup>

The etiology of this neoplasm remains poorly understood and the diagnosis of CCOC depends on the exclusion of other clear cell tumors. An accurate histopathologic diagnosis is therefore essential which is often aided by immunohistochemistry. Histopathologically, three subtypes of CCOC have been described.<sup>[5]</sup> The biphasic pattern is the most common, in which clear cell nests are intermixed with polygonal cells having eosinophilic cytoplasm. The monophasic pattern consists of cell nests containing exclusively clear cells. The ameloblastomatous pattern is the least common and has palisaded ameloblastomatous cells at the periphery of clear cell nests. The case that we report was characteristic of the monophasic pattern.

CCOC must be distinguished histopathologically from clear cell carcinoma arising from salivary glands and other benign odontogenic tumors which may also have clear cells, like ameloblastoma and calcifying epithelial odontogenic tumor. Metastatic tumors, such as metastatic renal clear cell carcinoma must also be ruled out. The presence of perineural or vascular invasion is an indicator of malignancy and can be used to identify CCOCs.<sup>[6]</sup>

The treatment of choice as agreed upon by most authors is wide excision of the tumor, including the adjoining soft tissue when involved. The recurrence rate following such treatments appears to be much lower than when treated by enucleation or curettage.<sup>[7]</sup> Elective neck dissection has been performed by several authors and it is interesting to note that none of these patients had recurrences.<sup>[1,7-10]</sup> Adjuvant radiation therapy is generally

considered in cases with extensive soft tissue invasion, perineural spread, lymph node metastasis with extra nodal involvement or in those where tumor-free margins are not possible. Long-term follow-up of patients with CCOC is imperative in view of the aggressive biological potential.

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