Biomedical Communication

Clear Cell Odontogenic Carcinoma: A Rare Case Report

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ABSTRACT

Clear Cell Odontogenic Carcinoma (CCOC) is a slow-growing, locally invasive odontogenic tumor affecting the jaws. It usually has confusing clinical characteristics, radiographical and histological features, making its recognition more challenging. In (2005), the WHO has reclassified CCOC as a malignant odontogenic tumor due to its aggressive behavior. This case report is about a woman who aged 42 years old, experienced swelling of the lower jaw and complained of paresthesia of the lips for two years. Radiographic findings showed an extensive, large, multilocular radiolucency lesion associated with scalloping, non-sclerotic border, and crossing the midline of the mandible. Histopathologically, the high-power view illustrated lobules of clear epithelial cells with clear cytoplasm. A review of English literature in PubMed Medline revealed few similar cases of CCOC affecting the mandible. The definitive diagnosis was consistent with CCOC; therefore, the patient was admitted to the hospital and surgical resection of the mandibular tumor was performed under general anesthesia. Long-term follow-up visits showed no signs of recurrence or post-surgical complications. We explain the signs and symptoms of CCOC such as symptomatic or asymptomatic jaw swelling, teeth loosening, displacement and mobility of teeth, and thinning of the mandible. Also, differential diagnosis and the nature of CCOC under the microscope were discussed and explained. The presentation of clear cell tumors is challenging, and it needs a meticulous investigation to determine the type of the tumor for proper diagnosis and management. CCOC should be included in the differential diagnosis list of jaw swelling that associated with slow-growing mass and paresthesia in the affected jaw.

KEY WORDS: CASE REPORT, CLEAR CELL ODONTOGENIC CARCINOMA, MANDIBLE, ORAL TUMOR, PARESTHESIA.

INTRODUCTION

Clear cell odontogenic carcinoma (CCOC) was first reported in (1985) still the existing reported cases of CCOC are very few due to which the behavior and pathology pattern of this tumor is still considered mysterious and not fully understood. It is a rare, aggressive, odontogenic malignant tumor (Hansen, Eversole and Green 1985). Approximately 74% of CCOC reported cases to affect the mandible with high incidence in females (Guastaldi and Faquin 2019). Previously, CCOC was classified as a benign tumor, and it was also known as clear cell ameloblastoma. CCOC owns aggressive features such as destructive growth patterns, regional lymphadenopathy, distant metastasis, and a high recurrence rate. Therefore, in (2005), the WHO reclassified it as a malignant odontogenic tumor (Guastaldi and Faquin 2019). A thorough review of literature in PubMed Medline revealed few similar cases of CCOC in the mandible. The reported case of CCOC in a 42-year-old, female patient with a swelling in the mandibular region. The work has been reported in line with the SCARE criteria (Agha et al. 2020).

Case Presentation: Forty-two years old women presented with paresthesia of the lower lip and large swelling in the mandible for more than a year and a half. The patient reported no previous history of trauma and no abnormal discharges such as pus or blood coming out of the mass. However, paresthesia and slight mobility were noted. She was diabetic and on medications for two years and she was allergic to pineapple which is associated with urticaria. Moreover, family medical history and genetic conditions were not remarkable. Moreover, the patient had a clean social record without smoking or abuse alcohol drinking. She had updated routine blood tests which were within normal limits. Extraoral examination revealed a significant enlargement of the chin with intact, and normal colored skin.

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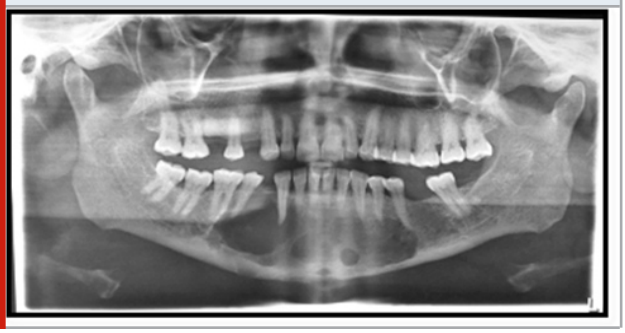
The examination illustrated no regional lymphadenopathy was noted. Remarkably, the extraoral examination confirmed the loss of sensation in the lower lip.

Informed written consent was obtained from the patient for publication of this case report and accompanying images. Intraoral examination showed a large, painless intra-bony swelling with a firm consistency and extending from the right lower first molar to the left lower first molar crossing the midline; area of teeth No. 20 to 30 (according to the universal numbering system). Notably, the patient had multiple missing teeth in the mandibular swelling area as follows tooth No.20, and No. 28. Vitality testing of the involved non-carious mandibular teeth was performed which reflected no abnormal responses. Radiographic analysis including orthopantomogram radiograph depicted an extensive, large, multilocular radiolucency associated with scalloping, non-sclerotic border lesion extending from the right lower first molar to the left lower first molar crossing the midline from tooth No. 19 to 30 without root resorption of the aforementioned teeth. Additionally, the radiographic film showed thinning of the inferior alveolar border in correspondence with the anterior mandible. Also, shifting of tooth No. 29 distally (Figure 1a and 1b).

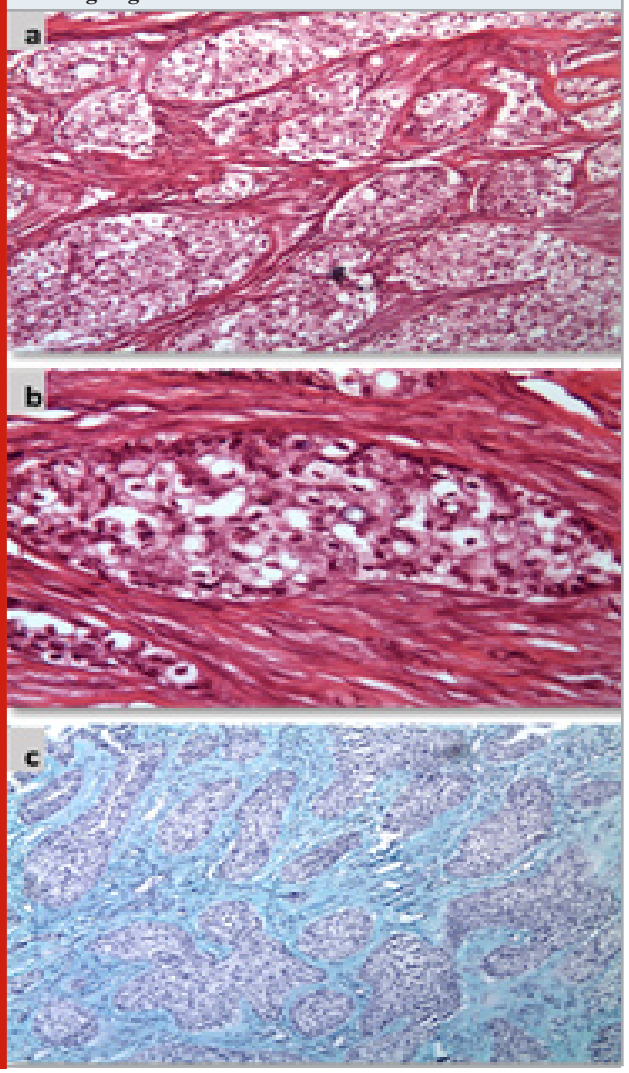
Figure 1: Panoramic radiograph shows an extensive, large, multilocular radiolucency associated with scalloping, non-sclerotic border lesion extending from the right lower first molar to the left lower first molar crossing the midline from tooth No. 19 to 30.



The patient medical history, the clinical and radiographic findings were all indicating the odontogenic tumors. The differential diagnosis of the mandibular mass included clear cell variant of ameloblastoma, clear cell odontogenic carcinoma, clear cell variant of a calcifying epithelial odontogenic tumor, mucoepidermoid carcinoma, acinic cell carcinoma, epithelial myoepithelial carcinoma, melanocytic tumors. She was educated about the nature of the tumor, different treatment plan options, possible results, and prognosis. An incisional biopsy was considered as the first line of management to determine the type of the tumor. The biopsy was performed under local anesthesia, an incisional biopsy was obtained and sent for histopathology analysis.

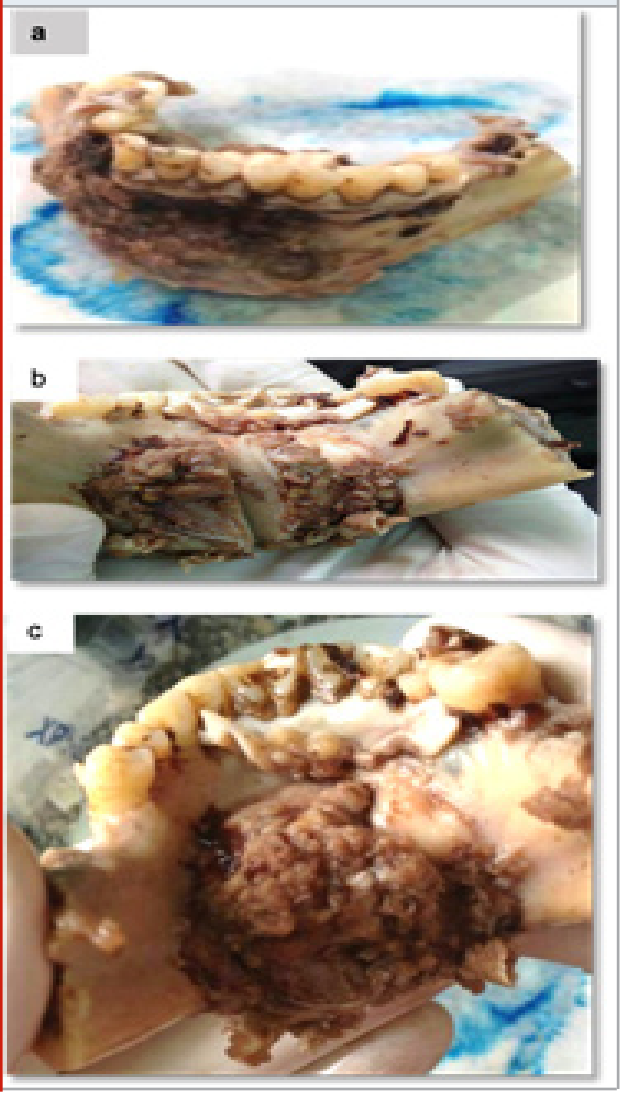
Two different stains were used for histopathologic analysis to provide an accurate diagnosis. Hematoxylin and eosin (H&E) stain; an immunohistochemistry stain, and Alcian blue stain; a non-immunohistochemistry stain. Under the microscope, the lesion appeared to have notable stromal hyalinization with small multiple nests and strands. Also, it showed lobules of epithelial cells with clear to eosinophilic cytoplasm (H&E) (Figure 2a, and 2b). Due to the overlapping and the similarity between odontogenic and salivary clear cell tumors, an Alcian blue stain was used to distinguish between them. The Alcian blue stain was negative for cellular mucin; therefore, salivary clear cell tumors and metastatic tumors were ruled out (Figure 2c).

Figure 2: (a) photomicrograph (H&E stain) shows stromal hyalinization with small multiple nests and strands; (b) a high-power view shows lobules of clear epithelial cells with clear cytoplasm to eosinophilic cytoplasm; (c) Alcian blue staining negative for cellular mucin.



The diagnosis was made based on the histopathologic features under the microscope which were strongly consistent with clear cell odontogenic carcinoma. Consequently, the patient was admitted to the hospital and surgical resection of the mandibular tumor with free margins was performed under general anesthesia (Figure 3a, 3b, and 3c).

Figure 3: Gross specimen shows CCOC in (a) the labial and (b &c) lingual aspects of the anterior mandible.



Then, the mandible was reconstructed to cope with the issues of functions and esthetic. The diagnosis of CCOC was, also, confirmed by the histopathological analysis of the resected portion of the mandible. Surgical and post-surgical instructions were delivered verbally and in written form to the patient. Also, all patient's concerns and inquiries were taking care of. The patient was monitored at the hospital for five days post-surgically. A two-week, one-month, and two-month, three-month, four-month, six-month, and one-year follow-up visits, she showed no recurrence or complications.

Analysis on the case presentation: CCOC is a slow-growing, and locally invasive odontogenic tumor affecting the jaws. A rare tumor that mostly affects females more than males. It usually has confusing clinical characteristics, radiographical features and poses benign first glance histological features, making its identifications even more difficult. Clinically, most cases of CCOC show pain, however, in this case, the patient had paresthesia of the lower lip with no sign of pain. Moreover, swelling of the jaw, teeth loosening, displacement and mobility, thinning of the mandible are other signs of the tumor. Histopathologically, the name of the clear cells came from the special appearance of the cytoplasm under the microscope which shows a clear cytoplasm with H&E stain. The accumulation of intracellular components such as lipid, mucin, and glycogen resulted in the special appearance of the clear cell (Upadhyay et al. 2019).

The picture of the tumor cells under the microscope varies in each case and it is hard to standardize. It includes different figures of stroma containing clusters and sheets of epithelial cells with color variations of cytoplasm, ranging from colorless to eosinophilic hue. In addition, in some cases, the epithelial cells could be arranged in a palisaded pattern which is similar to ameloblastoma. Alcian blue stain to rule out mucous cells containing tumors such as mucoepidermoid carcinoma and clear cell tumor of salivary glands. Clear cells are, also, seen in odontogenic and non-odontogenic tumors. Clear cells of odontogenic origin such as odontogenic cysts, clear cell ameloblastoma, mucoepidermoid carcinoma, clear cell tumor of salivary glands, calcifying epithelial odontogenic tumor, acinic cell carcinoma, myoepithelial carcinoma, and squamous cell carcinoma. On the other hand, in patients with a history of cancer, distant metastasis is possible. Therefore, undifferentiated clear cell tumors of non-odontogenic origin from kidneys, thyroid, breast, lungs, and colon could be metastasis to the mandible (Swain, Dhariwal and Ray 2013; Liu et al. 2020).

Consequently, a meticulous examination should be done for patients with a possible diagnosis of CCOC to rule out clear cell metastasis from distant organs (Werle et al. 2009). Diagnosis of CCOC depends on the detailed medical history, thorough physical examination, radiographic imaging, and histological analysis of the provided incisional specimen. CCOC ideal treatment is not clear yet due to the rarity of the documented and reported case. Most cases undergo radical surgical resection of the tumor with safe bone tissue margins. However, when palpable lymph nodes or extensive destruction of the bone with tissue invasion are involved, the management would be twisted and shifted to be more aggressive; to reduce the chance of recurrence. It would indicate lymph node removal, radiotherapy, or/and chemotherapy. In our report, the patient did not show signs of lymphadenopathy or metastasis, and the resection of the mandibular tumor was performed with safety margins. Therefore, in this case, the patient did not go through lymph node removal, radiotherapy, or/and chemotherapy. Long-term follow-up of CCOC cases is highly recommended to exclude recurrence and metastasis of the tumor (Upadhyay et al. 2019; Liu et al. 2020).

CONCLUSION

The findings of the present case showed confusing clinical features, radiographical patterns, and poses benign first glance histological forms, making its recognition and diagnosis more difficult. A detailed investigation is essential to reach the definitive diagnosis and to manage the tumor properly. The presentation of clear cell tumors is challenging, and it needs a meticulous investigation to determine the type of the tumor for proper diagnosis and management.

Conflict of Interests: Authors declare no conflicts of interest to disclose.

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