Case Report

Calcifying Epithelial Odontogenic Tumor (Clear-Cell Variant) A case report and literature review

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ABSTRACT

Calcifying epithelial odontogenic tumor (CEOT), or the Pindborg tumor, is very rare neoplasm, which accounts up to 1% of all odontogenic tumors. CEOT is locally aggressive benign odontogenic neoplasm arising from epithelial tissue. CEOT is rarely reported in Libya.

Here in, we present an additional case of 55 year old female patient with a clear cell Variant (CCEOT) of CEOT located in the left posterior maxilla with maxillary sinus extension, with an emphasis on clinical, radiographic and histopathology features and review of literatures.

Keywords: calcifying odontogenic tumor, clear cell type, maxilla.

INTRODUCTION

Odontogenic tumors (OT) are lesions that derive from the tooth-producing tissues or their remnants that remain entrapped either within the jaw bones or into the adjacent soft tissues.

These lesions represent hamartomas with varying degrees of differentiation, while the rest are benign or malignant neoplasms with variable aggressiveness and potential to develop metastasis. Odontogenic tumors are rare, some even extremely rare, but can pose a significant diagnostic and therapeutic challenge. Calcifying epithelial odontogenic tumor (CEOT) is one of a rare benign, locally invasive, most frequently presenting as a painless slow growing swelling. This tumor is commonly associated with impacted teeth. It usually involves the posterior mandibular. Therefore CEOT exclusively must be considered in differential diagnosis of tumors involving the jaw bones. Pindborg a Dutch pathologist in 1955 described very rare odontogenic tumor. He referred to this tumor as a calcifying epithelial odontogenic tumor (CEOT) [1-2]. The term "Pindborg's tumor" was first used by Shafer and colleagues in 1963 [3]. This type of tumor is considered benign yet its behaviour consider rather aggressive in nature, with a recurrence rate of 10 to 15% [4]. Malignant transformation and metastasis are very rare [5].This tumor occurring mainly intraosseous, but extra-osseous cases rare reported [6] .The mandible is more commonly affected than the maxilla in ratio of 3:1[6]. About 65% of all reported cases have occurred in the molar-premolar area of mandible, which is associated with un-erupted or embedded teeth [7]. CEOTs more frequently affects adults in an age range of 20-60 years, with no sex predilection [8]. The etiology of this tumor is unclear, but it is generally accepted to be derived from stratum intermedium or dental lamina remnants[9]. Radiographic features of CEOTs may present variable radiographic appearances, depending on stages of development, either uni or multilocular radiolucent area with impacted tooth, mixed radiolucent and radiopaque pattern or totally radiopaque [10, 11], due the maturation of the tumor.

Histopathology is the main stay for definitive diagnosis. The classical histopathological characteristics of CEOT comprise of plates of polyhedral epithelial cells with highly eosinoplilic cytoplasm, nuclear polymorphism, intercellular bridges in fibrous conjunctive tissue, associated with calcifications as well as deposition of an amyloid-like substance; however, occasionally, focal areas of clear cells can be observed in the clear-cell variant of CEOT (CCEOT) [12].

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The method of treatment varies and depends mostly on the size and anatomic location of the lesion, range from simple enucleation and curretage to radical excisions. Maxillary CEOT should be treated more aggressively, as they tend to grow more rapidly and usually poorly localized ,the best treatment is hemi- maxillectomy [12].

CASE REPORT

A 55-year-old female referred to university private dental clinic for evaluation of facial swelling. She complained of progressive, painless swelling of left side of the face , which she had had for 1 year. Patient had taken medications but there was no reduction in the size of the swelling . The patient's medical history was unremarkable. A clinical examination revealed an extra orally diffuse swelling of the left cheek, which is hard non-tender swelling involving the left posterior maxillary region. Regional submandibular lymphnodes were palpable, mobile, firm and tender. Intraoral examination revealed a diffuse swelling extending from the maxillary left first premolar to the tuberosity region, obliterated the buccal vestibule. The overlying mucosa was intact. On palpation, at the level of maxillary alveolar process the swelling was hard and painless.

A panoramic showed a mixed radio-opaque radiolucent lesion, not well-demarcated from the surrounding bone, with radiopaque specks scattered throughout, involving the left maxillary tuberosity, alveolar process, and the maxillary sinus with erosion of lateral wall and floor of the sinus. No evidence of any impacted tooth (Figure 1) .

Using a computerized tomography, The coronal view of facial CT reveals a large osteolytic mass with amorphous and stippled calcifications involving the left maxilla (Figure 2).

An incision biopsy was done under local anaesthesia. Undertaken and the specimen was sent to the Oral Pathology laboratory, department of Oral Medicine, Pathology and Diagnosis .

Histopathological examination revealed, islands and sheets of polyhedral epithelial cells with prominent intercellular bridges and amyloid-like material. Cellular and nuclear pleomorphism and calcification in the form of Liesegang rings were seen figure 3. These bodies were present between the epithelial cells as well as in the stroma. Some areas showed a proportion of clear cells with vacuolated cytoplasm. This histological picture is in consistent with CCEOT.

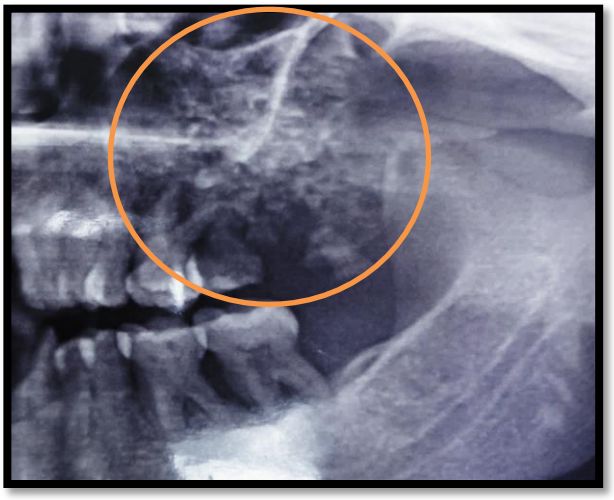

Figure 1: A cropped image of an orthopantomograph showing a large mixed radio-opaque radiolucent lesion on left maxilla .

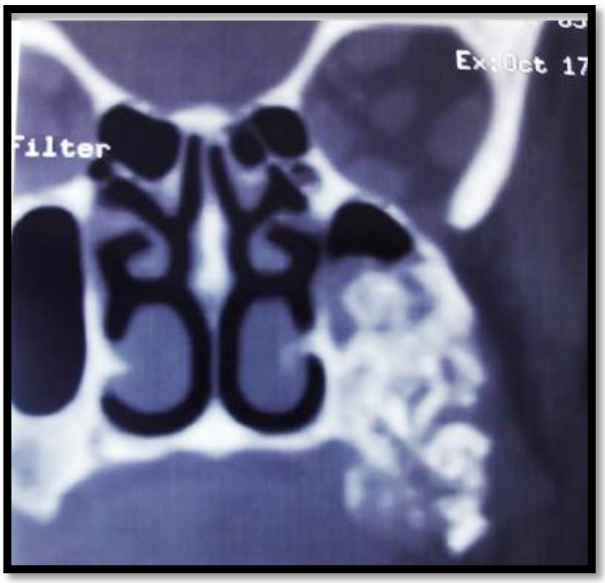

Figure 2: CT coronal section showing a large expansile lesion involving the left maxilla and extending up to maxillary sinus .

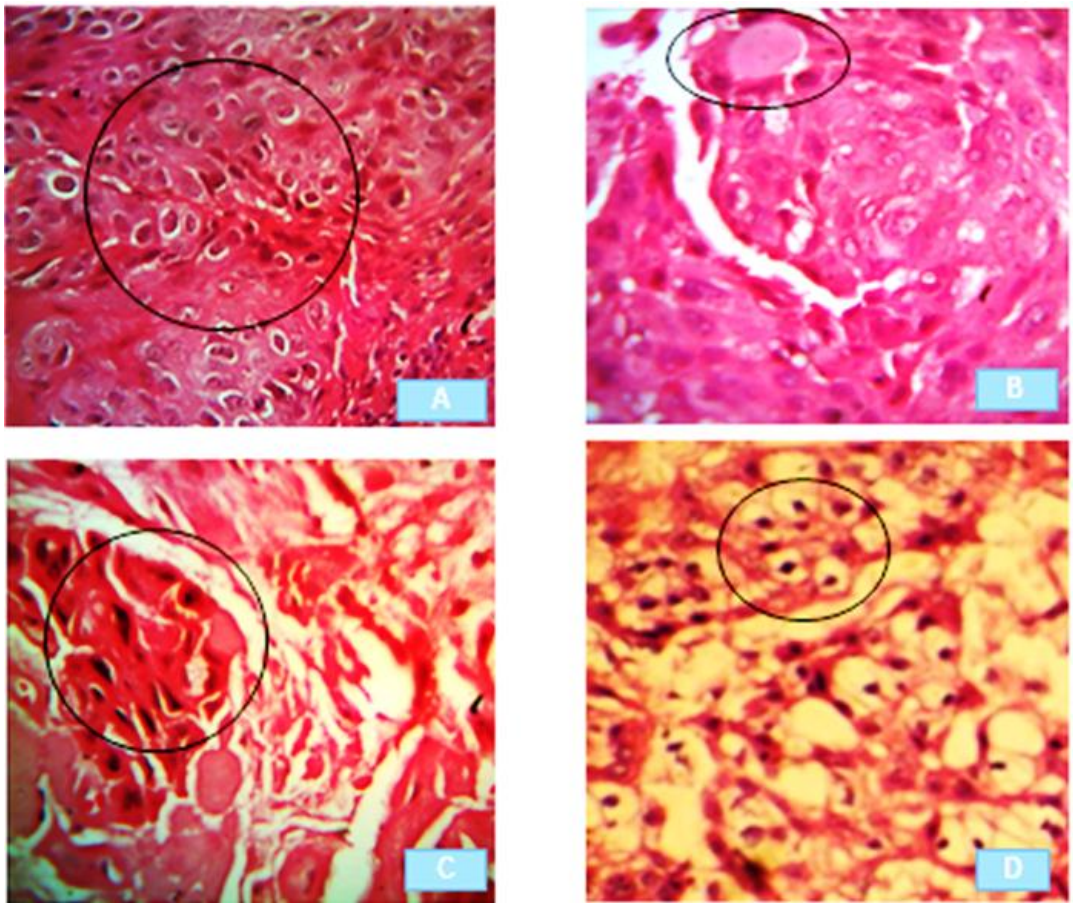

Figure 3: Histopathological photomicrographs of H&S stain ,showing(A)an island of epithelial tumor with evident cellular pleomophism and hyperchromatic nuclei (X100) ,(B) eosinophilic amyloid-like material, (X200) (C) sheets of polyhedral epithelial cells , (X400) (D) clear cell (X400)

DISCUSSION

Since the publication of Pindborg in 1955, there have been numerous articles on CEOT cases have been published. CEOT is a rare benign epithelial odontogenic lesion that comprises from 0.2% to 1.1 of all odontogenic tumors. It's a slow growing, painless neoplasm that occurs as intraosseous (96%) and extraosseous (4%) variants [13]. Some 60% of intraosseous CEOT are associated with an unerupted tooth (or odontoma). Peripheral (extraosseous) CEOT cases have been reported, manifest as nonspecific, sessile gingival masses, commonly on the anterior gingiva. In some cases, this lesion is associated with underlying bone cupped-out erosion [13] .

Radiographic features of CEOTs may present variable radiographic appearances, depending on stages of development. The lesion usually consists of a radiolucent area, which is well defined associated with an impacted tooth .The area is often unilocular when small and larger lesions tend to have honeycomb or soap bubble appearance. This radioluceny, then become mixed radiolucent and radiopaque pattern or totally radiopaque [10, 11]. Advanced imaging technique of CT plays an important role in evaluating the extent of the tumor in relation to facial bones and important structures .

Histogenesis of CEOT , is controversial and it is thought to be derived from the oral epithelium, reduced enamel epithelium, stratum intermedium or dental lamina remnants uncertain. Pindborg was initially of the opinion that the CEOT was of odontogenic origin and developed from, the reduced enamel organ of the unerupted tooth [14]. Furthermore the appearance of reports cases of peripheral and

intraosseous CEOT without associated unerupted tooth, may indicate other sources than reduced enamel epithelium. The occurrence of peripheral lesions suggests that CEOTs arise from the basal cells of the oral epithelium or remnants of the dental lamina [15]. Other investigators suggested that tumor cells originate from stratum intermedium dental lamina as an idea based on the morphologic similarity of the tumor cell to the normal cells of stratum intermedium and a finding of high activity of alkaline phosphatase and adenosine triphosphate at both sites [15].

Histologically, four distinct patterns of CEOT have been described [16]. Although two or more patterns may coexist in the same tumor, a predominance of one type is often seen. Briefly stated, the four patterns are the following:

(1) Sheets of polyhedral cells with intracellular bridges;
(2) A cribriform pattern with numerous spaces containing eosinophilic (amyloid-like) material;
(3) Densely populated tumor cells with interspersed multinucleated giant cells;
(4) Clear cell predominant tumors with pseudoglandular architecture and centrally placed eosinophilic material .

A clear cell variant has been recognized, which was first reported by Abrams and Howell in 1967 [17]. Few cases has been reported in the literature of clear cell variant of CEOT [18]. While it is not surprising to find clear cells in odontogenic lesions, the exact nature of the clear cells in CCEOT has not been elucidated.

The diagnosis of CCEOT is usually based on the presence of typical epithelial clear cells within the tumour. The nuclei show considerable variation in size and shape, mitotic figures rarely seen .Microscopic differential diagnosis between clear cell variant of CEOT , should include other tumors showing clear cells differentiation n the jaws, such as metastatic tumors from liver, kidney, thyroid, colon and prostate, central mucoepidermoid carcinoma and clear cell odontogenic carcinoma [19,20]. The presence of amyloid material and calcified formation in our case were essential features to establish the final diagnosis of clear cell predominant CEOT.

The treatment plan is dependent on the size of the tumor, the location of the tumor, histological findings, general condition of patient and operator skill. It ranged from simple enucleation or curettage to radical and extensive resection such as hemimandibulectomy or hemimaxillectomy. In our case which is a clear cell variant, she may needs hemimaxillectomy, with free bone margins ,because this tumor grow more rapidly and usually not well confined .

The prognosis of the CEOT is generally good with infrequent recurrence, but long-term follow-up is essential because there is a recurrence risk if the tumor was incompletely resected and in particular with the clear cell variant [21]. The recurrence rate may range from 14% to 20% [22]. Malignant behavior is extremely rare [23].

CONCLUSION

Evidence supports that clear cell variant is one of a rare type of CEOTs, it has more aggressive biological behavior and higher chances of recurrence. The classic histopathological feature will always confirm the diagnosis. The presence of clear cell directs the surgeon towards more definitive surgical excision of the lesion. Maxillary involvement with CCEOT should be treated more aggressively because they grow faster and possess close proximity to important structures. This variant is often associated with recurrence, long term follow-up is recommended.

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