

Review Article

A rare case of multilocal calcifying epithelial odontogenic tumor

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Abstract: Calcifying epithelial odontogenic tumor (CEOT) is a rare odontogenic tumor which was first described by Pindborg in 1956. CEOT accounts approximately 1% of all odontogenic tumors. 65% of reported cases are in the mandible. A 18-year-old female patient was referred to our clinic with a chief complaint of missing teeth. Intraoral examination revealed a large number of missing teeth and buccal and lingual cortical plate expansion was seen from premolar to molar regions of mandible and maxilla. Impacted teeth including #45-48-37-13-17-18-27-28 were extracted and lesions were enucleated. Histopathological examination confirmed the diagnosis of calcifying epithelial odontogenic tumor. The postoperative course was uneventful and the patient was asymptomatic in the 57 months period of follow-up.

Keywords: Pindborg tumor, multilocular, enucleation, embedded teeth

Introduction

Calcifying epithelial odontogenic tumor (CEOT) is a rare tumor first described by Pindborg in 1956 that accounts for approximately 1% of all odontogenic tumors [1]. There are two variants of Pindborg tumor, intraosseous (central), which constitutes the great majority of cases (94%), and a rare (6%) extraosseous (peripheral) type.

Although the exact origin is unknown, it is believed to be caused by oral epithelium, enamel epithelium, stratum intermedium and dental lamina remnants [1]. Histologically, the tumor exhibits polyhedral eosinophilic epithelial cells and deposition of an amyloid-like substance [2].

It usually occurs between 30 and 50 years of age, irrespective of gender. Sixty-five percent of reported cases are in the mandible, and frequently in the posterior region [3].

Clinically, CEOT is characterized as a slow growing, painless swelling [2]. Radiographically, it is usually observed in association with unerupted teeth, and as a mixed radiopaque radiolucent

lesion. Although small lesions are unilocular in appearance, larger lesions are multilocular [4].

We report a unique case of Pindborg tumor affecting multiple sites in the maxilla and mandible, together with a review of the relevant literature.

Case report

An 18-year-old female patient was referred to the oral and maxillofacial surgery clinic with a principal symptom of missing teeth. Hepatitis B was present in her medical history. The family history was non-contributory, and there was no history of trauma or any developmental abnormalities. The patient was a non-smoker and had no other tobacco habits. Intraoral examination revealed poor oral hygiene, hypertrophic gingiva, and a large number of missing teeth. Buccal and lingual cortical plate expansion was observed from premolar to molar regions in the mandible and maxilla. Extraoral examination revealed no involvement of the lymph nodes or facial asymmetry. Radiographic investigation including panoramic radiography revealed impacted #13, 17, 18, 23, 27, 28, 35, 37, 38, 45 and 48 teeth (**Figure 1A**). Dentigerous cyst,

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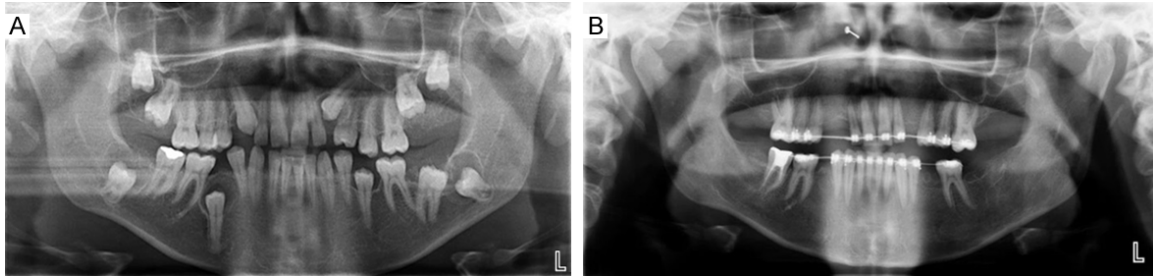


Figure 1. A. Panoramic radiograph showing similar pathologic lesions involvement of both jaws and the association with the teeth. B. Postoperative panoramic radiograph. The postoperative course was uneventful, and the patient was asymptomatic in the 57-month follow-up period.

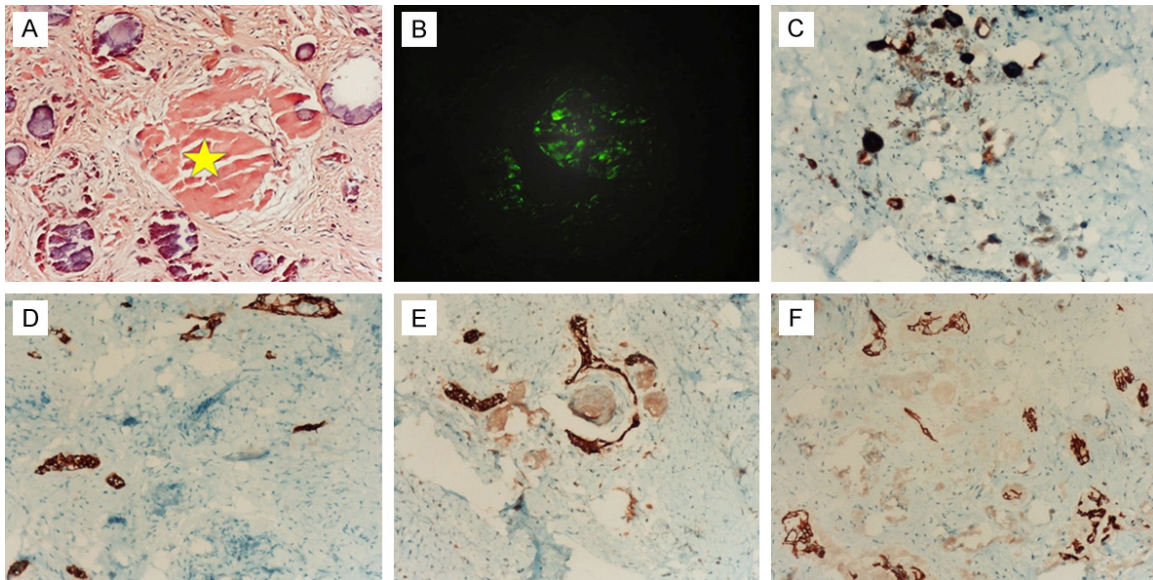


Figure 2. A. Amorphous eosinophilic material in stroma positive with Congo Red stain, $\times 200$. B. Kongo Red positive area shows apple green birefringence under polarized light, $\times 200$. C. Immunohistochemical positive staining with amyloid, $\times 200$. D. Epithelial cells positive with CK19 with immunohistochemical staining, $\times 200$. E. Epithelial cells positive with pancytokeratin CK19 with immunohistochemical staining, $\times 200$. F. Epithelial cells positive with CK5.6 with immunohistochemical staining, $\times 200$.

odontogenickeratocysts, unicystic CEOT and ameloblastoma were considered at differential diagnosis. We decided to treat the lesions with extraction of the involved impacted teeth and enucleation. Under local anesthesia, the left mandibular third molar tooth was extracted first. Pindborg tumor was diagnosed following histopathological examination. Impacted teeth #45, 48, 37, 13, 17, 18, 27 and 28 were then extracted and the lesions enucleated. All lesions removed after surgeries were diagnosed as CEOT at histopathological examination (**Figure 2A-F**). Radiolucent areas of bone formation were observed in the postoperative period at radiographic examination. The postoperative course was uneventful, and the

patient was asymptomatic in the 57-month follow-up period (**Figure 1B**).

Discussion

CEOTs are rare benign, but locally aggressive tumors. They are frequently seen in the posterior region of the mandible and constitute 1% of all odontogenic tumors [5]. The mandible is affected more than the maxilla, at a ratio of 3:1 [6]. A local tumor recurrence rate of 10-15% has been reported, and malignant transformation is rare [5]. Tumor location in our case was consistent with the literature. More unusually, the tumor involved both jaws and was present as a plurality. Additionally, all lesions were

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Table 1. Distribution of age, gender, location, symptoms, number of lesions, symptoms, associated lesions and complication of patients with Pindborg tumor

Authors/year	Gender	Age	Number of lesions	Location	Symptoms	Associated lesion	Complication
Parish P/2007	M	51	5	1-Distal to the right maxillary second premolar. 2-Distal to the left maxillary central incisor, 3-Left maxillary second premolar 4-Distal to the maxillary left first molar, 5-Distal to the left mandibular second premolar	No clinical signs	No	NA
Tarsitano A/2012	M	55	2	1-Associated with the roots of teeth 45, 44, 43, 42, 41, 31, 32, and 33.2-Right maxilla in the 11, 12, 13 area	No clinical signs	Squamous odontogenic tumor	42 months, no complication
Present case	F	18	11	Associated with teeth 13, 17, 18, 23, 27, 28, 35, 37, 38, 45, and 48	No clinical signs	No	57 months, no complication

associated with impacted teeth. This is the only such case in the literature. No recurrence was observed during 57-month follow-up.

Correct diagnosis and treatment should include a careful assessment of clinical, radiological and histopathological findings. Clinically, the lesion may occur as an expansile lesion of the jaw, therefore, the growing tumor may cause displacement of teeth or pain-like symptoms. These findings can be common to many lesions affecting the jaws, particularly odontogenic tumors and cysts [7]. However, our patient referred to our clinic with no clinical signs or symptoms.

Although numerous CEOT cases have been reported in the literature, cases of multilocular CEOT are rare. To the best of our knowledge, there have been only two reports of cases of multilocular CEOT in the English-language literature [7, 8]. This is the first report in the English-language literature of numerous multilocular CEOTs in both the mandible and maxilla associated with embedded teeth. The patients in these two previous reports were aged 51 and 55. Both were male. In the first case, lesions were located distal to the left maxillary central incisor and left maxillary second premolar, and distal to the maxillary left first molar, and one lesion was distal to the left mandibular second premolar. In the second case, the first lesion appeared to be associated with the roots of teeth 4.5, 4.4, 4.3, 4.2, 4.1, 3.1, 3.2, and 3.3, while the second lesion was located in the right maxilla in the 11, 12, 13 area. In the literature, diagnosis is often made using panoramic radiographs. However, some cases have been identified with CBCT. Both lesions were enucleated surgically (Table 1). We report this case in the light of this literature review. Our patient was an

18-year-old woman. Two lesions in our patient were located at the left mandibular second and third molars, one at the right mandibular second premolar, one at the right mandibular third molar, two at the right maxillary second and third molars, and two at the left maxillary second and third molars. All the lesions were also associated with embedded teeth.

In a study of 113 cases, Frankin and Pindborg [9] reported that CEOTs were observed in individuals aged 8-92 years. Cicconetti et al. [10] reported that CEOTs are observed in the age range 20-60 and that distribution among man and women is equal. In contrast to the literature, our patient was an 18-year-old female.

Radiographically, calcifying odontogenic tumor most commonly appears as a diffuse or a well-circumscribed unilocular radiolucent area. In some cases, the lesion becomes multilocular with a honeycomb pattern [4]. In others, calcified structures of different sizes are seen in the lesions [11]. Radiographically, the tumor may be mistaken for ameloblastoma or dentigerous cyst [12]. Diagnosis of this rare tumor may be problematic at clinical and radiographic examination. Histopathologic evaluation plays an important role in correct diagnosis and treatment planning. Histologically, the tumor contains polyhedral epithelial cells covered intimately in large sheets and distinct intercellular connective tissue [4]. Mitotic activities are rarely seen. Amyloid-like substance and calcified rings are other typical findings [13]. In our case, all the lesions were associated with impacted teeth and were diagnosed by histological examination.

The treatment of calcifying odontogenic tumor usually begins with surgical excision. Small

intrabony mandibular and maxillary lesions can be treated by simple enucleation or curettage with careful excision of a thin layer of bone adjacent to the tumor [14]. Aggressive surgical approaches such as segmental resection, hemimandibulectomy or hemimaxillectomy are required for large tumors. In these cases, bone discontinuity necessitates reconstruction procedures such as grafting or distraction osteogenesis [2]. Although CEOT has a lower recurrence rate than that of ameloblastoma and malignant transformation and metastasis is rare, long-term follow-up is recommended in the literature [15]. In the present study, small intrabony lesions were removed surgically with a bone safety margin, and associated teeth were extracted. No recurrence was reported during 57-month follow-up. The patient was also referred to the department of orthodontics and prosthodontics for treatment of malocclusion and edentulism.

This case is a very interesting and rare manifestation of CEOT due to multicentric presentation, the involvement of both jaws and the association with the teeth.

Disclosure of conflict of interest

None.

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