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Calcifying epithelial odontogenic tumor in a child: A case report

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ABSTRACT

Calcifying epithelial odontogenic tumor (CEOT) or Pindborg tumor is a rare tumor that accounts for <1% of all odontogenic tumors. It usually affects patients between the 3rd and 4th decades of life, however a wide age range from 8 to 92 years has been reported. This neoplasm may be associated with erupted or unerupted teeth. There are both intraosseous and extraosseous variants of CEOT and the posterior part of mandible is the most common location. We present an interesting case of CEOT involving the left side of the maxilla associated with unerupted canine and premolar in an 11 year old girl.

Keywords: Odontogenic tumor; Pindborg; Pathology.

Introduction

Calcifying epithelial odontogenic tumor (CEOT) or Pindborg tumor is a rare odontogenic tumor that was described by Jens Pindborg the first time [1]. This tumor is a very rare neoplasm that comprises only 1% of all odontogenic tumors [2,3]. Stratum intermedium layer of enamel organ in stages of tooth development or remnants of the primitive dental lamina found in the initial stage of odontogenesis has been suggested as the origin of this tumor [4]. According to most studies, it usually affects patients between the 3rd and 4th decades of life, however a wide age range from 8 to 92 years has been reported [5]. This odontogenic tumor does not show a gender predilection according to most studies [6,7]. Although, 71% of all case reports in children have been observed in females [8]. Although, CEOT is a benign tumor with slowly growing, local aggressive behavior in jaw bone and soft tissue involvement has been observed in some reports [7,9]. Furthermore, malignant transformation with multiple recurrences and patients with metastasis have been reported only in adults and are extremely rare [10,11].

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There are both intraosseous (96%) and extraosseous (4%) variants of CEOT and the posterior part of mandible bone is the most common location [7,12]. This neoplasm may be associated with erupted or unerupted teeth [10]. Tipping, migration, rotation or mobility of the adjacent tooth may occur. In radiographical features, CEOT is characterized by unilocular or multilocular radiolucency. Mixed radiographic feature may be observed due to the presence of scattered flecks of calcifications that create a typical "snow driven" appearance [7]. Histopathologically, sheets of polyhedral epithelial cells with distinct cell borders, prominent intercellular bridges, nuclear pleomorphism, hyperchromatism and few mitoses are the hallmarks of the CEOT. In addition, spherical amorphous calcifications (Liesegang rings) may be present in between tumor cells and connective tissue. There is a homogeneous, eosinophilic, acellular matter intermixed with the tumor cells that is identified as 'amyloid-like' substance and stains with Congo Red and demonstrates apple-green birefringence on polarization [7,11]. To date, more than 362 cases of CEOT have been reported but 15 cases occurred in children that only 3 of them occur in maxilla [8,10,13]. Here, we present a rare case of CEOT involving the left side of the maxilla associated with unerupted canine and first premolar in an 11 year old girl.

Case Report

An 11 year old girl presented with the chief complaint of absence of the left maxillary first premolar and canine was referred to the Oral Surgery Department of Shahrekord Dental School. She was asymptomatic and had no pain, paresthesia or swelling in the jaws. There were no palpable lymph nodes on physical examination. Cone Beam Computed Tomography (CBCT) revealed a tooth-shaped radiopacity in conjunction with an unerupted maxillary first premolar measuring 1.5cm x 1cm (Fig 1). Based on the clinical and radiographic features, odontoma, adenomatoid odontogenic tumor (AOT), calcifying epithelial odontogenic tumor (CEOT), fibro-osseous lesion and osteoma were considered as differential diagnosis. Excisional biopsy was performed under general anesthesia and the lesion with impacted tooth was extracted. Gross examination revealed white color tissue measuring 1.5cm x 1cm x 0.5cm with hard consistency and solid cross-section attached to the unerupted first premolar. In histopathological features, sheets and islands of odontogenic epithelial cells with polygonal shapes, homogenous eosinophilic cytoplasm and large ovoid nuclei were observed. Mitoses and nuclear pleomorphism were rarely seen. The cellular outlines of the epithelial cells were distinct and intercellular bridges were observed. The tumor islands were frequently enclosed by hyaline material and the deposition of amyloid-like substance, resulting in a cribriform appearance. Furthermore, there were multiple round calcified areas forming concentric "Liesegang ring " like pattern in and around the epithelial cells and connective tissue (Fig 2). Considering all the features, the final diagnosis of CEOT was made. The patient was followed up for nine months with no sign of clinical recurrence.

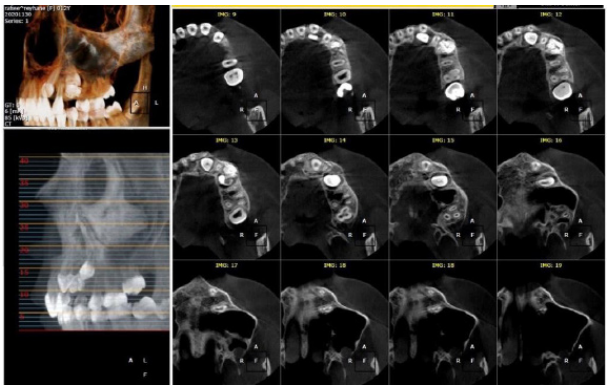
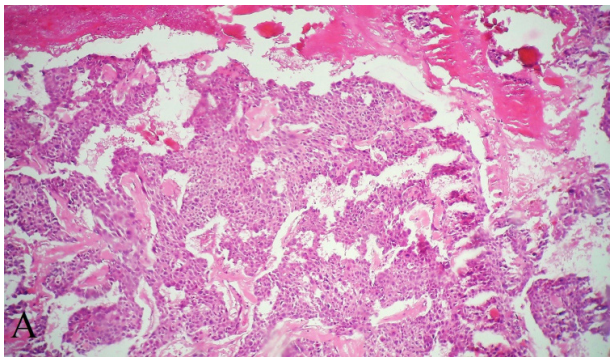
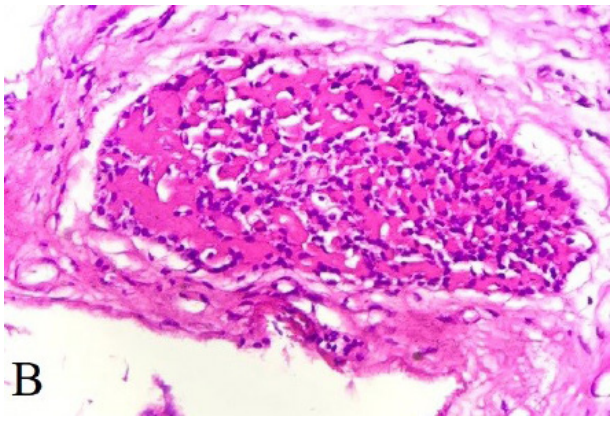


Figure 1. CBCT revealed a tooth-shaped radiopacity in the maxilla.





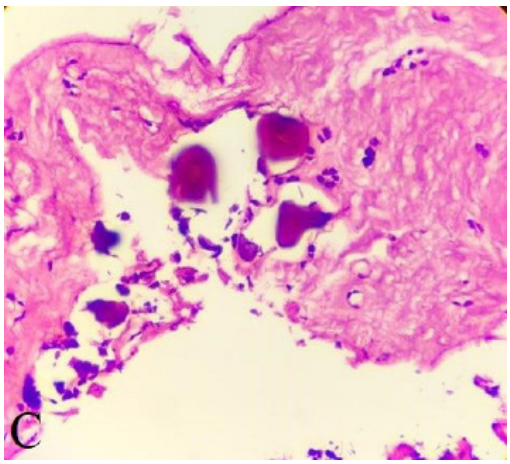


Figure 2. A: sheets and nests of polyhedral neoplastic cells with abundant eosinophilic cytoplasm, B: deposition of amyloid-like substance, C: round calcified areas with "Liesegang ring " like pattern.

Discussion

CEOT is an epithelial odontogenic tumor with incidence between 0.4% and 3% of all odontogenic tumors [3,14]. The prevalence of intraosseous lesions is higher than extraosseous types. This tumor has been reported in a wide age range from 30 to 60-year-old and has equal gender predilection [15]. But, in this report, an 11-year-old child with CEOT was presented. The posterior region of the mandible is the most common site of intraosseous lesions. Almost half of the lesions are associated with impacted teeth or odontomas. According to the most studies, the mandibular molar teeth were the most common teeth associated with this lesion [16]. The lesion was seen on the left part of maxilla and was associated with unerupted first premolar in the present case. Similar to this case, most intraosseous CEOTs in the pediatric group present as asymptomatic, slow-growing masses which may cause cortical expansion [5,8,17]. Radiographic features of CEOT depend on the stage of the lesion and amount of calcified structures [5]. This lesion is also widely reported around the crown of the tooth [15]. The radiographic differential diagnoses include odontoma, ameloblastic fibro-odontoma, CEOT, fibro-osseous lesion and osteoblastoma. Furthermore, in the pediatric patient, the aneurysmal bone cyst, ameloblastoma, odontogenic keratocyst and dentigerous cyst may be referred to as differential diagnoses [8]. Our patient also revealed a tooth-shaped radiopacity related to an unerupted maxillary first premolar [7]. In histopathological featurs, CEOT is composed of sheets and nests of polyhedral neoplastic cells with abundant eosinophilic cytoplasm, prominent intercellular bridges and pleomorphism [15]. Furthermore, accumulation of extracellular eosinophilic material with concentric calcification is characteristic of CEOT that is called Liesgang ring which reacts with specific amyloid staining. Some studies suggest that degradation of lamina densa material causes the formation of this amyloid and it is different from those seen in endocrine-associated amyloid or systemic amyloid [8]. All these histological features were observed in our case. Several histologic variants of CEOT such as clear cell, Langerhans cell, myoepithelial cells, cementum forming, non-calcifying and CEOT associated with other odontogenic lesions have been reported [13,18].

Treatment of CEOT depends on the clinical, radiographic and histopathologic variants and has ranged from conservative enucleation nad marginal resection to extensive resection such as hemi mandibulectomy or hemi maxillectomy for aggressive features [5,7]. Furthermore, the site and size of the lesion are important to choose the better treatment [13]. Because maxillary tumors grow more rapidly and have a higher risk of recurrence (14%), maxillectomy is the recommended treatment [7]. The lesions with calcification and amyloid-like material had more differentiation and a lower risk of recurrence [19]. Malignant transformation of CEOT is extremely rare [7,19]. According to case reporters, malignant transformation or aggressive behavior has not been observed in children [8]. Our case of CEOT did not show local expansion after 9 months, there was no evidence of recurrence of the lesion. Fazeli reported a maxillary CEOT in a 13-year-old girl with locally aggressive expansion to the lateral sinus wall, nasal cavity and orbital floor that was incidentally found on routine dental examination. Only 6 cases of CEOT with impacted/or developing tooth were reported. [8] However, a follow-up period from 5 to 10 years is recommended and more studies in children with CEOT are required [5].

Conflict of Interest

There is no conflict of interest to declare.

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