Case Report

Clinical, radiological and histological features of an unique case of calcifying epithelial odontogenic tumor

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Abstract

Calcifying epithelial odontogenic tumor (CEOT), also known as Pindborg tumor, is a rare benign but locally aggressive odontogenic neoplasm, accounts for <1% of all odontogenic tumors. CEOT is usually seen in the posterior area of the mandible in-between 30 and 50 years of age without definite sex predilection. A painless, slow-growing swelling with bone expansion is the most common clinical feature of CEOT. Radiographically, it presents as a mixed radiographic lesion may or may not be associated with any impacted tooth. Confirmation of the diagnosis is made by histopathological examination. The tumor has a recurrence rate of 10%–20% and so periodic follow-up is necessary. A unique case of CEOT involving the right mandibular molar–premolar in a 25-year-old female patient with clinical behavior, radiological, histopathological features and surgical managements is discussed herewith.

Keywords: Amyloid, calcifying epithelial odontogenic tumor, Congo red, odontogenic tumor

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INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT) or Pindborg tumor is a rare, slowly growing, nonencapsulated, locally invasive epithelial odontogenic neoplasm characterized by the presence of irregular sheets and islands of polyhedral often pleomorphic odontogenic epithelial cells along with amorphous eosinophilic substances and calcified areas.[1,2] The origin of CEOT is controversial, although it is believed to be derived from the oral epithelium, reduced enamel epithelium, stratum intermedium or remnants of dental lamina.[3] Pindborg tumor is prevalent among 30–50 years of age without any definite sex predilection and recognized as either intraosseous (96%) or extra osseous (4%), being mandible is commonly affected than maxilla with a ratio of 2:1.[3,4] This neoplasm may present as painless, expansile bony hard swelling causing expansion and distortion of the cortical plates along with subsequent soft tissue infiltration. It may cause tipping, migration, rotation and/or mobility of the adjacent tooth and may also be associated with impacted/unerupted teeth. Root resorption is found only in 4% of cases.[5]

Radiologically, CEOT is characterized by unilocular or multilocular radiolucency that often exhibits mixed radiographic feature (65%) due to the presence of scattered flecks of calcifications, often produce a typical "snow driven" appearance.[3,4]

The classical histopathological features of CEOT comprise sheets, cords and/or islands of polyhedral, neoplastic

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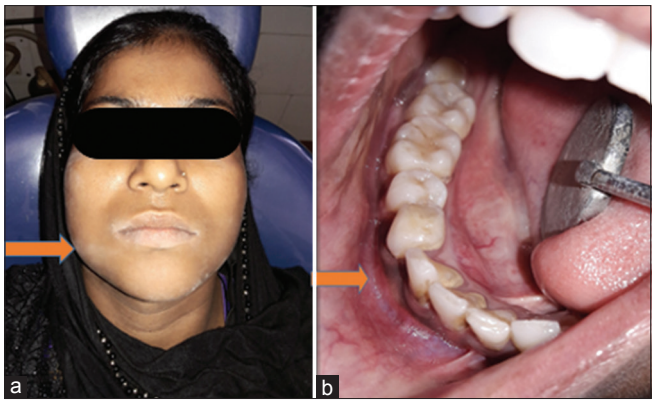

Figure 1: (a) Extra oral photograph showing diffuse swelling involving the right lower third of the face. (b) Intraoral photograph showing a swelling involving 41–47 region

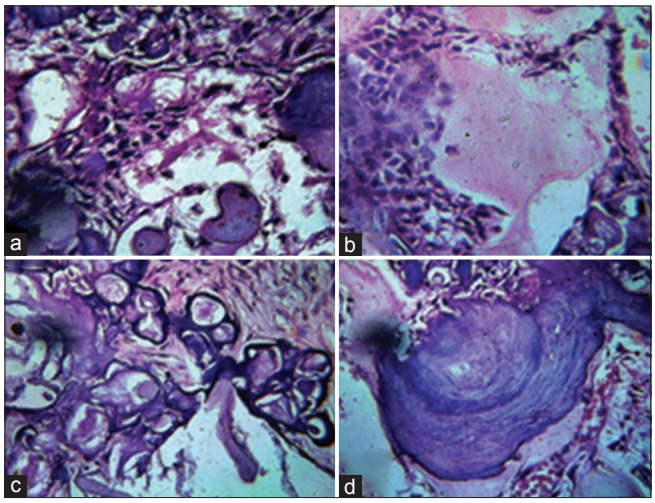

Figure 3: High power photomicrographs (H&E, ×40) showing the presence of sheets of polyhedral neoplastic odontogenic epithelial cells with distinct intercellular bridges (a and b), amyloid-like material and areas of concentric ring-like calcifications within the connective tissue stroma (c). Areas of hard tissue formation (d)

epithelial cells with well-defined cell borders and distinct intercellular bridges which may demonstrate pleomorphism and occasional mitosis. Other characteristic findings are the presence of eosinophilic amyloid-like substances and concentric calcified areas as "Liesegang" ring-like pattern. Amyloid like substances stained positively with congo red and produce typical apple green birefringence under polarized light in confocal microscope. Histological variation includes CEOT with Langerhans cell, CEOT displaying cementum and bone-like materials and CEOT with clear cell variant.[3,4,6]

Treatments of CEOT depends on tumor size, location, general condition of the patient and operator's skill, and it ranges from enucleation or curettage to radical surgical resection followed by reconstruction. According various literatures recurrence rate is 10-20% following conservative management.[7-9]

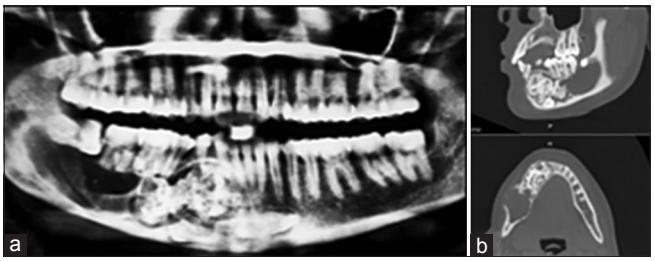

Figure 2: (a) Orthopantomogram showing the presence of a multilocular, well-defined radiolucent lesion involving the right side of the whole of the body of the mandible extending from 34 to 48 region along with scattered flecks of calcifications. 43, 44 and 48 are impacted (b) computed tomography scan of the mandible demonstrating anteroposterior extension of the tumor and destruction of buccal cortical plate

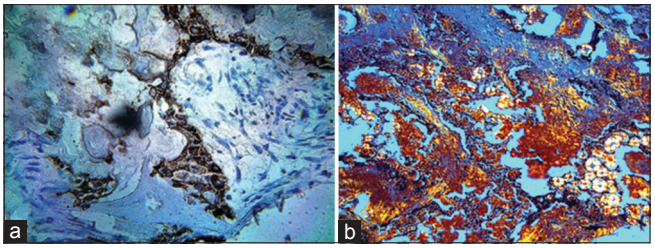

Figure 4: High power photomicrograph (H&E, ×40) showing cytokeratin (CK19)-positive odontogenic epithelial cells (a), low power photomicrograph (H&E, ×10) showing apple-green birefringence of amyloid-like materials under polarized light after Congo red staining (b)

Here, we present an interesting case of CEOT involving the right side of the mandible associated with impacted canine and premolar in a 25-year-old female patient with unique histologic features.

CASE REPORT

A 25 years old female patient from semi-urban area reported with chief complaint of pain and swelling involving the right side of lower jaw for the last 1 year. The patient was aware of the lesion for the last 2 years. However, as the lesion was small and asymptomatic, she did not seek any treatment. The lesion gradually increased in size with localized intermittent pain for the last 1 year. Her past medical and family history was noncontributory. On extra oral examination, there was the presence of a diffuse swelling involving the lower right third of face extending from parasymphyseal to the angle of the mandible leading to mild facial asymmetry without any regional lymphadenopathy [Figure 1a].

Intraoral examination revealed the presence of a diffuse swelling extending from 41 to 47 region covered by normal oral mucosa. On palpation, the lesion was bony hard, mildly tendered and associated with expansion of buccal and lingual cortical plates. 43 and 44 were absent along

with over retained 84 [Figure 1b]. Routine hematological and biochemical investigations were within normal limit.

Orthopantomogram revealed the presence of multilocular, well-defined radiolucent lesion involving the right side of the whole of the body of the mandible extending from 34 to 48 region. Most striking feature was the presence of multiple radiopaque foci in the symphyseal and parasymphyseal region, radiodensity of which were similar to that of the normal bone. 43, 44 and 48 were impacted along with root resorption of 46 and 47. 84 was retained [Figure 2a]. Computed tomography scan showed the presence of expansile bony lesion along with the presence of radiopaque foci involving the symphyseal and parasymphyseal region of the right side of the lower jaw [Figure 2b]. Destruction of buccal cortical plate and thinning of lingual cortical plate were also noted.

Based on the above clinical and radiological findings, the provisional diagnosis of fibro-osseous lesions and odontogenic neoplasms were made.

Ethical clearance of this study was received. Following this, incisional biopsy was performed from representative site of the lesion after obtaining informed consent from the patient, and the specimen was processed for histopathological examination.

The light microscopic features revealed the presence of scanty areas of polyhedral, neoplastic, odontogenic epithelial cells with prominent cellular outlines and intercellular bridges along with the presence of homogeneous, eosinophilic and amorphous materials at places [Figure 3a and b]. The most characteristic feature is the presence of multiple rounded calcified areas, forming concentric "Liesegang ring-"like pattern in and around the epithelial cells and connective tissue [Figure 3c]. Areas of osteoid tissue formation within the neoplasm were also noted [Figure 3d]. The histological diagnosis was made as "CEOT."

To confirm this diagnosis, immunohistochemical (IHC) evaluation was performed. IHC marker such as cytokeratin 19 confirmed the presence of odontogenic epithelium within the neoplasm [Figure 4a]. Amyloid-like material after Congo red staining produces typical apple-green birefringence viewed under confocal microscope with polarized light [Figure 4b].

After considering the clinical, radiological and histological features, confirmatory diagnosis of CEOT was made, and the patient was referred to the Department of Oral and Maxillofacial Surgery for further treatment and management.

DISCUSSION

CEOT was first described and defined by prof. J. J. Pindborg in the year 1955 and was histologically characterized by the presence of three components – polyhedral often pleomorphic epithelial cells, amorphous amyloid-like depositions and calcifications.[1-4,6]

The patient under discussion was a 25-year-old female, having mildly tendered swelling involving the right side of the mandibular symphyseal and molar region associated with expansion of buccal cortical plates. The side of involvement and age of occurrence of CEOT in our case are in accordance with the cases published in previous literatures.[3,6]

Radiologically, CEOT exhibits as a unilocular or multilocular radiolucent lesion together with scattered flecks of calcification throughout the radiolucency giving rise to "driven snow" appearance. However, 52% of the reported cases have been definitely associated with impacted/unerupted tooth. Our case also revealed similar radiological features along with the presence of impacted right canine, premolar and third molar teeth and retained deciduous first molar. Resorption of roots of 46 and 47 were also noted although it is an uncommon finding according to some authors.[1,3,6]

Histological sections of CEOT are composed of polyhedral, closely packed epithelial cells with prominent intercellular bridges, having nuclei of varying form and size associated with the presence of extracellular, eosinophilic, amyloid-like material and areas of concentric calcifications in the form of "Liesegang ring-"like fashion with the connective tissue stroma. Amyloid-like material showed apple-green birefringence under polarized light after Congo red staining.[3,6,10] Odontogenic epithelial cells stain positively with cytokeratins such as AE1/AE3, CK5/6, Cam52, CK 14, CK19 and P63.[11] All these histolopathological features were also present in our case too. The presence of calcification in CEOT may have prognostic implications. The absence of calcification indicates a poorly differentiated tumor with more chances of recurrence.[12]

Surgical managements of mandibular lesions range from simple enucleation to hemimandibulectomy.[13] Maxillary lesion is usually treated with maxillectomy since they grow more rapidly and occur in vicinity of some important structure.[3] Recurrence occurs more frequently in maxilla (14%). The incidence of malignant transformation of Pindborg tumor is extremely low.[14,15]

CONCLUSION

Because of pleomorphism of epithelial cells, sometimes, CEOT is mistaken as malignancy. Complete surgical excision is essential to prevent recurrence of this neoplasm. Hence, proper clinicopathological and radiological correlation is essential to diagnose this benign neoplasm.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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