Calcifying Epithelial Odontogenic Tumour: A Case Report

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Abstract Calcifying epithelial odontogenic tumour (CEOT) presenting at an unusual site is rare. The characteristic cytologic findings in association with radiologic features can help the cytopathologist and the surgeon in rendering a firm preoperative diagnosis of Calcifying Epithelial Odontogenic Tumour even at atypical sites such as maxilla. The purpose of this article is to report an additional case of CEOT and discuss the radiologic appearance, histologic features, clinical behavior and surgical management.

Keywords Maxillary tumor · Biopsy · Calcifying epithelial odontogenic tumour · Resection

Introduction

Calcifying epithelial odontogenic tumour (CEOT) is a rare benign odontogenic neoplasm, forming 0.4–3.0% of all intraosseous tumours [1–5]. CEOT was first described by Dr. Jens J. Pindborg, 1955. In honour to this internationally renowned oral and maxillofacial pathologist, this tumor is also known as Pindborg's tumour [4, 6]. It is an uncommon, benign, odontogenic neoplasm that is exclusively derived from epithelial tissue. There is no consensus on the originating cells. Some pathologists suggest that it is derived from the stratum intermedium layer of the enamel organ in the tooth development stage, others favour a different hypothesis in which, this tumour may arise from remnants of the primitive dental lamina found in the initial

stage of odontogenesis. The tumor is similar in behavior to ameloblastoma but has a slightly lesser recurrence rate after surgery.

Report of a Case

We report a case of a 25-year-old male who came to our department with a complaint of pain, swelling and pus discharge from upper left posterior teeth region since 3 months. He noticed a swelling over the posterior maxillary region which gradually increased to the present size. He had undergone an extraction of decayed 26 three months ago assuming the tooth to be the cause of the swelling. Personal history revealed that the patient was a nonsmoker and did not have any other tobacco habits. The family and medical history was non contributory.

On extra oral examination there was mild facial asymmetry on the left side of face because of an oval diffuse swelling measuring 2 × 2 cm in the maxillary sinus region. Overlying skin appeared normal and on palpation the swelling was bony hard and there was mild tenderness present. There was no involvement of the lymphnodes.

Intraoral examination revealed absence of 26 and 27. 21, 22, 23, 24, 25 and 28 were present. There was pus discharge from a sinus in 26 regions where the patient had undergone extraction. The overlying mucosa was normal in colour and consistency. Buccal cortical plate expansion was seen from 24 to 27 regions obliterating the buccal vestibule. The area was mildly tender to palpation. The swelling was oval shaped bony hard and well defined and measured 3 cm × 2 cm in diameter (Fig. 1).

Radiographic investigations included panoramic skia-gram and paranasal sinus computed tomography 2 mm slices in axial and coronal planes (Fig. 2a–d).

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Fig. 1 Pre-operative photograph showing the extent of facial asymmetry in the left midfacial region

Panoramic skiagram showed single, unilocular lesion with a radiopaque tooth like structure in the posterior sinus aspect.

CT showed a large, expansile, lytic, heterogeneously enhancing lesion of approximate size of 42 mm × 16 mm × 28 mm involving left maxilla, inferiorly extending up to alveolar ridge and superiorly extending into the maxillary sinus with an unerupted tooth projecting into the lesion.

A differential diagnosis of residual cyst, dentigerous cyst, odontogenic keratocyst, calcifying odontogenic cyst, benign odontogenic tumour like ameloblastoma, adenomatoid odontogenic tumor, odontogenic myxoma, ossifying fibroma, central giant cell granuloma was considered. Routine preoperative tests like complete blood count and chest skiagram were done which showed normal results.

An aspiration biopsy was negative and an incisional biopsy taken from maxillary buccal aspect of 26, 27 under local anesthesia was reported as calcifying epithelial odontogenic tumor (Fig. 3).

The patient underwent a resection of the left posterior maxilla with a 0.5-cm margin of normal surrounding bone through the intra oral approach. The defect was allowed to heal with secondary intention. A prefabricated heat cure acrylic obturator was fixed in place by clasps on the opposite side molars. Postoperatively patient was kept on the nasogastric tube feeding and started to feed orally on the seventh postoperative day. Patient was followed up for 6 months without any recurrence of the tumour.

Histopathological Report

Gross specimen consisted of left partial maxilla from 24 to 28 region, measuring 40 mm long, 16 mm wide, 25 mm (Fig. 4a, b).

Microscopic section showed a benign tumour with histologic features of CEOT. The closely packed tumour cells exhibit variation in nuclear size and shape. In most areas tumour cells are small with hyperchromatic nuclei. Other areas show large tumour cells with pyknotic nuclei pressed against the cell membrane and faintly eosinophilic cytoplasm. There were stromal and intraepithelial aggregates of amyloid deposition.

Discussion

The CEOT is a benign but rare odontogenic tumor with less than 1% of all odontogenic neoplasms. It is most commonly reported in patients during the 4th, 5th, and 6th decades of life. The mean age range is 33–43 years of age [7–11]. It is usually not found in children and adolescents. They appear to be no sex predilection [8, 9]. Mandible is more commonly affected than the maxilla in ratio of 3:1 [7]. When present in the maxilla, the CEOT is preferentially located in the posterior region. The prevalence in the molar region is three times that in the premolar region [9, 10]. CEOT with extension into the maxillary sinus is uncommon [12].

A peripheral variant of CEOT also exists which presents clinically as a nodular mass on gingival mucosa in anterior tooth location, but this is extremely rare and only 10 cases are reported (5%) [13].

Pindborg tumour is usually detected in an asymptomatic patient and discovered only through routine radiographic examination by the dentist or it may present as a slow growing, painless, expansile bony hard swelling causing cortical bone to become egg shell thin before perforation and subsequent soft tissue infiltration. It may cause tooth tipping, migration, rotation and/or mobility secondary to tooth resorption [7].

The patient rarely may report pain, nasal stuffiness, headaches, epistaxis or proptosis [12–15] when tumour is in maxilla. Because of the teeth involved within the tumour mass, the lesion closely resembled a dentigerous cyst.

Depending on stages of development, CEOT may present variable radiographic appearances [9]. The lesion usually consists of a radiolucent area, which is well defined. Area is often unilocular when small and larger lesions tend to have honeycomb or soap bubble appearance [7].

Multiple radiopacities of varying size may develop within the radiolucent area and occasionally there are

Fig. 2 **a** Orthopantomogram showing lesion in maxilla, **b** Para nasal spine view, **c** Axial section of computed tomography, **d** Coronal section in computed tomography

extensive areas of calcification that cause the lesion to become radiopaque [15]. The mixed radiolucent and radiopaque pattern occurred most often (65%) followed by the completely radiolucent pattern (32%) and least often the totally radiopaque “wind driven snow” pattern (3%). When tumour is associated with impacted tooth, it may appear as pericoronal radiolucency with or without small radiopacities.

On CT examination, CEOT shows expansion and thinning of buccal and lingual cortical plates. The use of Denta Scan is especially useful to reveal tumour dimensions [16].

When CEOT is analyzed by MRI, it reveals predominantly a hypointense lesion on T1-weighted images and mixed hyperintense lesion on T2-weighted images [7].

According to Pindborg, the characteristic histologic criteria for the diagnosis of CEOT are sheets of large polygonal epithelial cells that have well-defined borders and often show prominent intercellular bridges. There is

Fig. 3 Histological picture of calcifying epithelial odontogenic tumour (H&E stain; 10×)

usually pleomorphism of the epithelial cells. Nuclei are often prominent and show considerable variation in size, shape, and number. Cellular abnormalities are frequent, whereas mitotic figures are rarely seen. Cytoplasm is abundant and eosinophilic. Varying amounts of an extra-cellular amyloid-like material that stains positive with Congo red stain in polarized light is also typical of these tumors. Concentric calcified masses with a Liesegang's rings calcification pattern are also pathognomonic for this tumor.

Ai-Ru et al. [10] have proposed a subclassification of these histologic features into four main patterns. 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:

Pattern 1 Polyhedral epithelial cell arrangements with deeply eosinophilic cytoplasm and prominent nuclei, cell outlines and intercellular bridges are distinct, frequent cell abnormalities, few or no mitotic figures, and calcifications are present in the fibrous stroma.

Pattern 2 Cribriform appearance of the cell arrangements, intercellular bridges might be discrete, few cell abnormalities, and calcification of the eosinophilic material filling the cribriform spaces with a Liesegang calcification pattern.

Pattern 3 Scattered or dense arrangements of the epithelial cells with varying size and multinucleated giant cells are frequent, mucoid material is present in the stroma.

Pattern 4 Nests and cords of epithelial cells containing either an abundant eosinophilic cytoplasm or a clear vacuolated cytoplasm, variable amounts of stroma containing eosinophilic material, and calcifications.

Treatment

Surgical treatment is dependent on the size and location of the neoplasm, the patient's overall medical condition or tolerance to withstand the surgical procedure, and the skill or experience level of the operator.

Small mandibular and maxillary lesions with well-defined borders may be surgically excised by either thorough intrabony curettage or conservative en-bloc resection sacrificing a thin margin of healthy adjacent bone [7].

It has also been suggested that even with an expected low rate of local recurrence compared with solid ameloblastoma, maxillary tumours should be managed more aggressively, with an opportunity to confirm histopathologically tumour free bone margins. For neoplasms greater than 4 cm, en bloc resection is mandatory. Extensive involvement requires partial/hemimaxillectomy requiring postoperative reconstruction and rehabilitation with titanium mesh, grafts and flaps may be required to close the

Fig. 4 **a** Specimen,
b radiograph of specimen

defect. Distraction osteogenesis has shown promising results in such defects [7].

The authors advocate early surgical intervention before such lesions could have widespread involvement. The tumour-free margins are confirmed histopathologically and require radical resection of affected jaw portion and any associated soft tissues with no less than 1 cm in every direction [17].

Prognosis

Most studies of Pindborg tumour report a local recurrence rate between 10 and 20% following conservative treatment. One case of advance disease in patient's maxilla resulted in local extension to the brain [18]. The incidence of malignant transformation of Pindborg tumour is extremely low.

Conclusion

Since the first review in 1966, the number of publications of CEOT cases has increased considerably, from 26 to the present 200 cases [7]. This case report is presented in view of rare involvement of maxilla by CEOT. In our case there was a tooth found embedded in the lesion which on initial appearance gave a radiographic and clinical picture of a dentigerous cyst in a 25-year-old patient. The lesion was discovered due to non healing sinus in 26 regions with a history of extraction. This is to emphasise the need to correlate radiographic, clinical and histopathological findings to arrive at the correct diagnosis and treatment planning.

Conflict of interest None.

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