ABSTRACT

The calcifying epithelial odontogenic tumor (CEOT) is a benign neoplasm of odontogenic origin as described by Dr. J J Pindborg in 1958. It is a rare entity and represents 1% of odontogenic tumors and it was coined as Pindborg tumor by Shafer et al. CEOT is a locally aggressive tumor arising from odontogenic epithelium that invades adjacent bone and soft tissues. It is commonly seen in 4th-6th decades of life and can be seen equally in both the genders. In this article we report and discuss a rare case of a cystic variant of CEOT arising in the left anterior maxilla in a 27 years old male individual associated with an impacted tooth.

Keywords: Calcifying epithelial odontogenic tumor; CEOT; Pindborg tumor; Odontogenic tumor.

I. INTRODUCTION

Pindborg introduced a rare entity in 1958, a tumor arising from odontogenic epithelium that represents 1% of the odontogenic tumors. Later it was coined as Pindborg tumor by Shafer et al [1,2]. Generally it presents as a slow growing tumor associated with an impacted tooth. It is found commonly in the fourth to sixth decades of life and does not show any gender predilection [3]. On a radiograph it shows mixed radiolucency. Histopathologically it is characterised by the presence of islands of epithelial cells, amyloid like material and leisgang rings of calcification. The differential diagnosis for CEOT include adenomatoid odontogenic tumor (AOT), calcifying odontogenic cyst (COC), ameloblastic fibro odontoma (AFO), and odontoma [2]. It is usually treated with surgical excision of the lesion, with a good prognosis. However, 14% of recurrence rate is reported in the literature [4]. In this report we present a rare case of cystic variant of CEOT occurring in the left anterior maxillary region.

II. CASE REPORT

A 27-year-old male reported to the department of oral and maxillofacial surgery with a painless swelling in the left anterior maxillary region measuring 6 × 8 cm extending from Left alar base to the left infra orbital region and left zygomatic region (fig.1). The swelling was smaller in size which increased over the period of 4 years. Rapid growth was noted in the past four months after the alleged history of trauma (hit on the face while playing). The swelling was mildly tender on extraoral palpation. Intraoral examination revealed a sinus tract opening in the left canine-premolar region with retained deciduous canine; also there was an occasional purulent discharge from the opening (fig. 2). On radiographic examination, a well defined radiolucency associated with impacted permanent canine was noted in the 21 to 27 tooth region (fig. 3); however the pre-surgical CT scan images showed osteolytic lesion with foci of calcification (fig. 4). The provisional clinical diagnosis was made as adenomatoid odontogenic tumor, calcifying epithelial odontogenic tumor, ameloblastoma, and dentigerous cyst. The incisional biopsy showed epithelium of odontogenic origin characterised by hyperplastic non keratinised stratified squamous epithelium of variable thickness. Few areas of dystrophic calcifications were evident within the epithelium.

The lesion was treated conservatively with surgical excision followed by peripheral ostectomy. The excised specimen sent for the histopathological examination was greyish brown in colour with firm consistency measuring 4 X 5 X 2.8 cm (fig. 5). Microscopic examination showed similar epithelial lining as described in the incisional biopsy report and numerous amyloid like deposits and concentric basophilic calcifications (Leisgang ring calcifications) in odontogenic islands in the dense hyalinised connective tissues. On congo red staining it
showed focal birefringence of amyloid deposits confirming as a cystic variant of calcifying epithelial odontogenic tumor (fig. 6). There was no recurrence noted after the follow up of six months.

Fig. 1: Photograph showing swelling in the left anterior maxillary region extending from Left alar base to the left infra orbital region and left zygomatic region.

Fig. 2: Intraoral swelling extending from 22-27, sinus tract opening in the left canine-premolar region with retained deciduous canine.

Fig. 3: A well defined radiolucency associated with impacted permanent canine in 21 to 27 tooth region.
As a group odontogenic tumors are cellular proliferations with a wide range of biologic potentials and behaviours. Pindborg tumor is a rare benign tumor arising from odontogenic epithelium, slowly growing; however, it is locally aggressive and results in localized bone destruction as reported in the case above. According to WHO classification, 1992 it is classified into Group I tumors. Neville and colleagues have stated that this tumor is a rare entity and represents 1% of the odontogenic tumors\[5\]. It is predominant in 4-6th decade of life with no sex predilection. Most commonly it is found in the posterior mandibular region as a painless slow growing mass associated with an impacted tooth however, rare extraosseous lesions have been reported in the literature\[6\]. Few maxillary tumors could be more aggressive in nature and may involve the maxillary sinus and nasal cavity leading to stuffiness and epistaxis. Some authors have proposed that CEOT arises from the dental organ's stratum intermedium or in the dental lamina, however the degree of differentiation of the odontogenic epithelium that
forms the idea of tumor pathogenesis is still debatable[6,7]. It's been proposed that amyloid deposition in Pindborg tumour represents an immune response to these stratum intermediate cells[8]. Others state that it develops from remains of the primitive dental lamina present early in odontogenesis, and that these epithelial rests are the real progenitor cells. The specific cause, however, remains uncertain[9].

CEOT appears as unilocular or multilocular radiolucency with or without radiopacities. It often can be misdiagnosed as a dentigerous cyst as it is associated with an impacted tooth in 50% of cases[2,7]. As it is associated with an impacted tooth, it is completely radiolucent in the early stages, appearing as a dentigerous cyst. In the second phase, small intratumoral calcifications may appear, which is distinctive but not definitive. The later stages are marked by osseous deterioration and calcification of the tumour, giving it a honeycomb appearance.

In this case report, we have reported a rare cystic variant of CEOT occurring in anterior maxilla. Few similar cases have been reported in the literature by Gopalakrishnan et al. and Channappa et al., a cystic lesion associated with the impacted tooth like the present case; however, in the case of Barreras et al., there was no evidence of the impacted tooth. Out of all cases, Gopalakrishnan et al. showed an area of transition from thin dentigerous cyst like lining into thicker CEOT cystic epithelium, which might contribute to the fact that cystic CEOT arises from the neoplastic transformation of the dentigerous cyst but still debatable.

Recently Kamboj et al., 2020 reported a case of similar lesion in maxilla associated with an impacted molar. On orthopantomogram it showed homogenous radiolucency involving apices of left maxillary premolars and molars, extending anteriorly to push the lateral wall of maxillary sinus and superiorly till the orbital roof. On computed tomography, it showed dubious radiolucency in the centre and few flecks of calcified deposits. The lesion showed increase in size, undermined the lateral sinus wall, and protruded into the nasal cavity. Therefore, the provisional diagnosis of dentigerous cyst or CEOT was made[7].

In the presented case similar radiographic findings were noted; however, the tooth impacted was maxillary canine. Provisional diagnosis for our presented case included dentigerous cyst, adenomatoid odontogenic tumor (AOT), calcifying epithelial odontogenic tumor and ameloblastoma. As the incisional biopsy showed features more similar to a cyst, the lesion was treated conservatively with enucleation (fig. 7) followed by peripheral ostectomy. The specimen was greyish brown in colour measuring 4 X 5 X 2.8 cm. On histopathological examination both the cases showed similar epithelium and connective tissue stroma. Special staining with congo-red stain confirmed focal birefringence of amyloid deposits which confirmed it as a cystic variant of CEOT.

As suggested by Franklin and Pindborg, CEOTs are less aggressive with only 14% recurrence rate. Conventional CEOTs are universally treated by conservative surgical approach with adjuvant procedures like peripheral ostectomy[10]; however the treatment modality may differ according to the factors like site, size, and utmost important histomorphological features of the lesion, etc[11]. We treated this lesion conservatively with enucleation followed by peripheral ostectomy according to the cystic characteristics shown by the epithelial lining on the incisional biopsy. No recurrence was noted after 6 months of follow up; however, the patient will be followed up for a long term to rule out the recurrence.

IV. CONCLUSION:

Although these types of lesions can be treated in multiple ways, we conclude that a conservative approach can be a treatment of choice with long term follow up; however, treatment may differ according to the factors like age, site, histopathological features and extent of involvement of the adjacent structures. Also this lesion should be considered in the differential diagnosis for the lesions exhibiting similar clinical and radiographic features occurring in the maxillary region.

REFERENCES:


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