INTRODUCTION

The calcifying epithelial odontogenic tumor (CEOT) is a rare benign tumor of the jaws first described by Pindborg[1] in 1955 and also referred as Pindborg tumor.[2] The clear cell variant of CEOT was first reported by Abrams and Howell[3] in 1967. Nineteen cases[3-17] of clear cell CEOT (CCEOT) have been described in literature. Topographically two entities have been distinguished: intraosseous (central) and extraosseous (peripheral). Twelve central lesions[3-13] have been reported of which only three cases have been reported in maxilla.[9,11,13]

Typically, CEOT has been described as slow growing, benign and locally aggressive tumor. Hicks et al.,[12] suggested that clear cell variant may show more aggressive behavior with higher recurrence (22%).

Clear cells are present as cellular components of the epithelial lining of lateral periodontal and gingival cysts in adults, or they may be found as clear-cell rests of the dental lamina within the connective tissue wall of these cysts. Rarely clear cells may occur in certain epithelial odontogenic tumors.[18] Philipsen and Reichart[19] reviewed 181 cases of Pindborg tumor, 15 of which are clear cell containing lesions. Some authors have classified CCEOT as separate clinical entity.

In this paper, a rare case of CCEOT of posterior maxilla is reported and clinical, radiographic histological features of the same are discussed.

CASE REPORT

A 36-year-old Indian woman reported to our department with painless, slow growing swelling of right upper jaw and loss of upper teeth due to mobility. Intra oral examination revealed solitary swelling in right posterior maxillary region with missing posteriors. The swelling was about 3 × 2 cm with normal color, intact mucosa and indentations of opposing tooth [Figure 1]. The mass was firm on palpation with slightly more expansion towards the palatal side. OPG showed 3 × 3.5 cm mixed radiolucent-radiopaque lesion in the posterior maxilla with impacted teeth. Radiographically “driven snow” appearance was appreciable [Figure 2]. Computed tomography (CT) revealed radiolucent-radiopaque mass in the right maxilla invading the cortex [Figure 3]. The lesion extended to posterior maxilla and obliterated the right nasal cavity.

An incisional biopsy was taken from the lesion. The specimen was subjected to histopathological examination. Hematoxylin and Eosin stained sections showed fibrous connective tissue stroma with islands of polyhedral epithelial cells containing nuclei of varying form and size. Extracellular eosinophilic amyloid like material was found containing concentric calcifications in the form of Liesegang rings [Figure 4].
Few scattered cells were found with clear vacuolated cytoplasm (clear cells). No cellular atypia or mitotic figures seen. The features were suggestive of CCEOT.

Considering the age of the patient, esthetic concern and benign nature of CEOT, curettage of the lesion along with the associated tooth was performed under general anesthesia (GA).

Patient reported back after 10 months complaining of nasal obstruction and tearing. CT scan confirmed recurrence in the posterior maxilla [Figure 5]. Intranasal biopsy was performed, revealed clear cells in the stroma of CEOT confirming the diagnosis of clear cell CEOT [Figure 6]. Subtotal maxillectomy with Weber Fergusson approach was done. Six weeks postoperatively, an obturator was given. No recurrence was found on 2 years follow-up.

DISCUSSION

Clear cell variant of CEOT is a rare entity and central CCEOT occurring in maxilla is rarest entity. An extensive review of literature on clinical and radiologic features of CCEOT has been done by Anavi et al.[20] that records 19 cases so far comprising of 12 central and seven peripheral lesions. Equal distribution in males and females, age range of 14-68 years; and of the 12 central lesions, nine involving the mandible and three in maxilla (75:25) have been reported.

The typical clinical presentation of CCEOT is painless swelling of the jaw causing cortical expansion. Radiological features of CEOT is unilocular or multilocular lesion with mixed radiolucent and radiopacity of various density. Classically driven snow or wind driven appearance has been described. Mostly associated with impacted tooth.[21] Clinical features and radiological features of the case presented are consistent with that of the other CCEOT reported. The three maxillary lesions[9,11,13] described were radiolucent in OPG, in the presented case lesion was typically of driven snow appearance.

Histological sections of CEOT containing islands of polyhedral epithelial cells, having nuclei of varying form and size with extracellular eosinophilic material–amyloid and concentric
Calcifications within the connective tissue matrix in the form of Liesegang rings are typical. Clear cell variant consists of sheets or cords of clear cells with foamy cytoplasm in the matrix. The clear epithelial cells contain glycogen, demonstrated by PAS reaction. These features were detected in the case presented confirming the diagnosis of clear cell variant. Other variants of CEOT described include pigmented, Langerhan cells containing and non-calcifying variants. Asano et al., in a CCEOT has demonstrated Birbeck granules.

The clear cell variant tends to be more aggressive and shows higher recurrence rates than conventional CEOT. Treatment options range from simple enucleation or curettage to radical and extensive resections. The surgical management modalities in 19 reported cases were complete or partial resection in seven patients (37%), excision in seven (37%), enucleation in four (21%) and curettage in one case (5%). Recurrence in two cases has been reported (17%).

Waldron et al., and Hicks et al., have reported that occurrence of the clear cells may prove to be a sign of increased tumor aggressiveness indicating a more radical approach. Recurrence of tumor following curettage in our case strongly supports the data and enbloc resection is recommended.

**CONCLUSION**

Evidence supports that clear cell variant is a distinct entity, has more aggressive biological behavior and higher chances of recurrence. Hence, it is important that presence of clear cells be included in histopathological diagnosis. The identification of clear cell directs the surgeon towards more definitive surgical excision of the lesion. Maxillary lesions should be treated more aggressively because they grow faster and possess close proximity to important structures.

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