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CASE REPORT

A Rare Case of Gorlin Cyst in the Maxillary Anterior Edentulous Region

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ABSTRACT

The Gorlin cyst (calcifying epithelial odontogenic cyst) is a rare lesion. Its clinical and radiological features are not pathognomonic, and is characterized by histological diversity. Radiographically, it presents as a unilocular or, occasionally, multilocular radiolucency that may contain calcified radiopacities or tooth-like densities. Microscopic appearance of odontogenic epithelium admixed with focal areas of dentinoid formation or calcification along with sheets of ghost cells gives the definitive diagnosis of Gorlin cyst. A rare case of Gorlin cyst, which present as swelling in the maxillary anterior edentulous region along with emphasis on review of literature, is presented in this paper.

Keywords: Cysts of jaws, Gorlin cyst, Calcifying epithelial odontogenic cyst.

INTRODUCTION

Calcifying epithelial odontogenic cyst (CEOC) is a unique jaw lesion, first recognized as a distinct entity by Gorlin et al, and hence the eponym Gorlin cyst.[1] It is an uncommon lesion that demonstrates considerable histopathologic diversity and variable clinical behavior. Due to its diverse histological picture, several terms have been used by different authors to describe this lesion, such as dentinogenic ghost cell tumor, calcifying ghost cell odontogenic tumor, keratinizing ameloblastoma, cystic calcifying odontogenic tumor (CCOT), peripheral odontogenic tumor with ghost cell keratinization, dentinoameloblastoma, ameloblastic dentinoma, epithelial odontogenic ghost cell tumor and odontogenic ghost cell tumor.[2] There have been disagreements on the terminology as well as whether to classify CEOCs as a cyst or a neoplasm. Although the WHO classification (2005) includes it as a benign odontogenic neoplasm and now recently it is renamed as CCOT.[3]

CASE REPORT

A 55-year-old female reported with a chief complaint of swelling in the upper front teeth region since one year. History revealed that one year back she was hurt by cow horn accidentally in her upper front teeth region, due to which she lost her upper left lateral incisor. After 4 days of injury, she developed a small swelling in the same region which slowly progressed to current size. She had mild discomfort and pain in the area on and off. Patient had undergone extraction of 21 about 20 days before reporting to this department as it was extremely mobile.

A solitary, oval-shaped well-defined swelling was present in the edentulous region with respect to 21 and 22 measuring 4.5 cm in its greatest diameter. It was extending from mesial surface of 11 up to mesial surface of 23 and was obliterating the labial vestibule (Fig. 1). Overlying mucosa was having one healing socket. On palpation, it was not tender, firm in consistency with smooth surface. Egg shell crackling was present with thinning of the labial cortex. Vitality test was done for all the maxillary teeth with electric pulp tester which did not elicit any abnormal response. A provisional diagnosis of residual cyst in the region of missing 21 and 22 was made with differential diagnosis of traumatic bone cyst, odontogenic keratocyst, Gorlin cyst, adenomatoid odontogenic tumor, Pindborg's tumor, traumatic neuroma and ameloblastoma were considered.

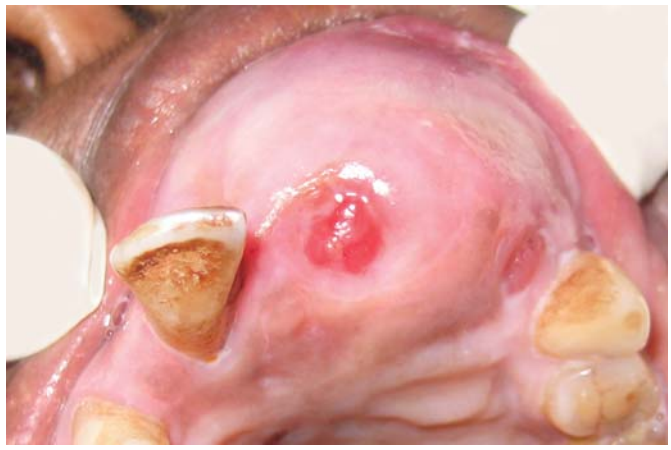

Fig.1: Intraoral swelling

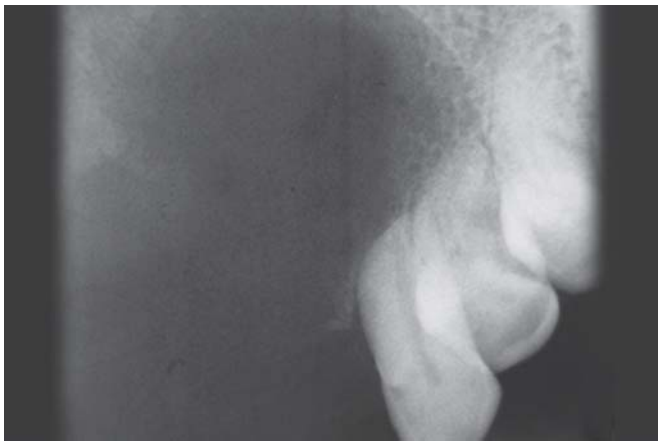

Fig. 2: IOPA

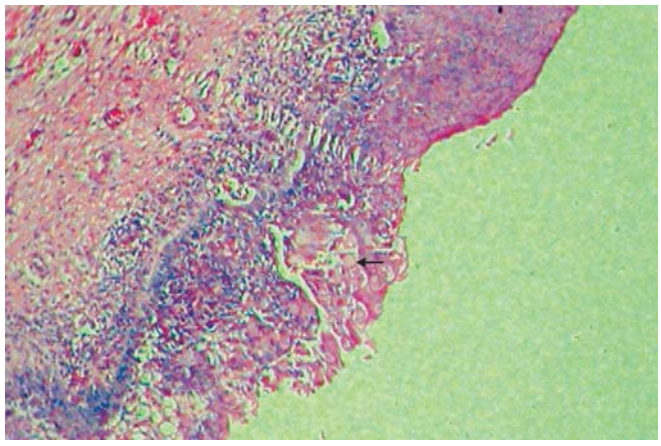

Fig. 4A: Photomicrograph showing cystic epithelial lining, chronic inflammatory cells infiltrate and ghosts cells (black arrow, H&E)

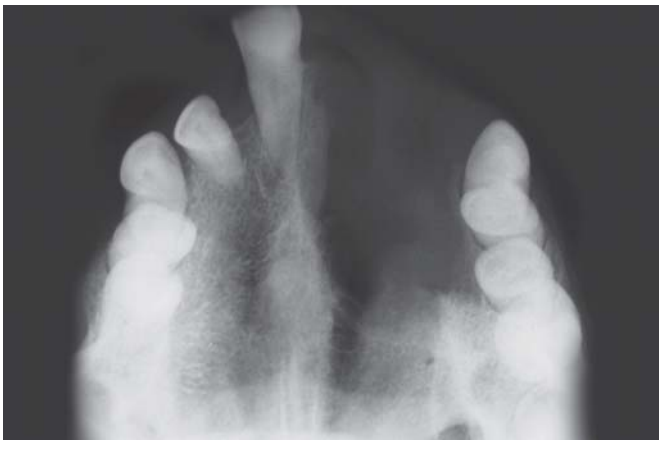

Fig. 3: Maxillary cross-sectional occlusal radiograph

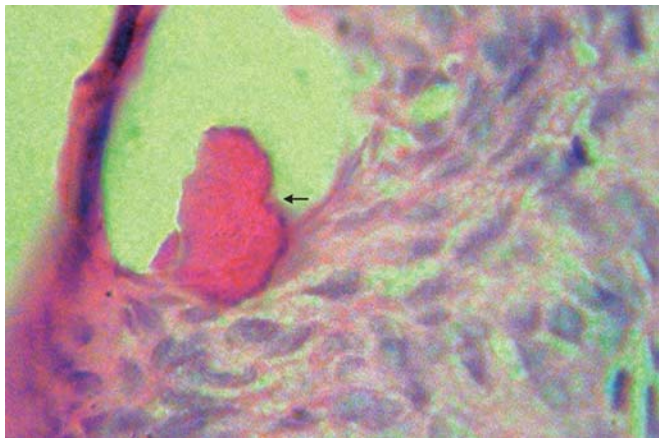

Fig. 4B: Photomicrograph showing calcification (black arrow, H&E)

An intraoral periapical radiograph in the edentulous region with respect to 21 and 22 was taken, which showed a unilocular radiolucency extending up to the distal surface of 24 and was well-defined posteriorly with a corticated margin (Fig. 2).

An occlusal radiograph was taken which showed a unilocular radiolucency extending posteriorly up to the distal surface of 24 and was well defined with a corticated margin except in the anterior region where there was no corticated margin. Radiolucency was extending up to the midline and there was slight expansion of labial cortex in the region of 22 and 23 (Fig. 3). Fine needle aspiration cytology (FNAC) was done which yielded blood tinged fluid. Cytology report was inconclusive.

The patient was operated under local anesthesia. Enucleation and curettage were done. Histopathological examination of the enucleated specimen revealed an odontogenic cystic and few foci showing stellate reticulum-like cells along with aggregations of ghost cells and one area with spherical calcification which confirmed the diagnosis of CEOC (Figs 4A and B).

DISCUSSION

CEOC is a rare example of a developmental odontogenic cyst comprising between 2.1 ± 0.37% of all odontogenic tumors.[4,8] Gorlin et al initially regarded it as the oral analog of the cutaneous calcifying epithelioma of Malherbe but later labeled it as the calcifying odontogenic cyst.[1] Prior to this, Rywkind described it as a variant of the cholesteatoma[5] while Maitland regarded it as a type of ameloblastoma.[6] Langlais et al proposed the term calcifying odontogenic lesion (COL), which encompasses both the cystic and tumorous forms as well as combined lesions containing elements of both. The odontogenic origin of the CEOC is widely accepted.[7]

Praetorius et al suggested that it develops in the dental follicle, gingival tissue or bone from remnants of either odontogenic epithelium or reduced enamel epithelium.[7] The pathogenesis is, however, still controversial.[9] According to one study results suggest that CEOC is a neoplasm caused by an activating mutation of beta-catenin.[10]

CEOC is usually a nonaggressive cystic lesion which occur with equal frequency in the maxilla and mandible.[11-13] At least 75% of CEOC occurs in bone. About 75% of cases are found anterior to first molar especially associated with incisors and canines.[14] It is predominantly intraosseous lesion, although 13 to 30% of cases in appeared series have appeared as peripheral (extraosseous) lesions.[15-17] Patient may range in age from infant to elder. The mean age is 33 years and most cases are diagnosed

in the second and third decades of life.[18] Cystic and solid variants of CEOC are described with the cystic variant comprising the majority of cases.[19,20]

Clinically, lesion usually appears as a slowly growing painless swelling unless secondarily infected. In some cases, expanding lesion may cause thinning of cortical plate as seen in present case and if untreated it can further lead to complete destruction of the cortical plate. On fine needle aspiration, the lesion often yields viscous, granular yellow fluid, which was not observed in the present case.[14]

It presents as unilocular radiolucency with irregular calcified bodies of varying size and opacity which is a typical feature, although not pathognomonic. Root resorption or divergence of adjacent teeth is suggested as one of the most important radiographic features, while in the present case there was only slight resorption in the apex of the root of upper left canine. Occasionally, it may appear multilocular.[18] In about one-third of cases, the radiolucent lesion is associated with an unerupted tooth, most often a canine.[14,21,22]

Histopathologically, a well-defined cystic lesion is found with a fibrous capsule and a lining odontogenic epithelium of 4 to 10 cells in thickness. The basal cells are cuboidal or columnar and similar to ameloblasts. The most characteristic histopathologic feature of CEOC is presence of variable numbers of ghost cells within epithelial component.[18] Occasionally, a small amount of true dentinoid material is formed in the adjacent connective tissue and can be differentiated from the ghost epithelium by means of the phloxin-tartrazine stain.[23] The histological features of the CEOC are complicated by its frequent association with other odontogenic tumors, such as the complex or compound odontoma, ameloblastoma, odontoameloblastoma, ameloblastic fibroma, ameloblastic fibro-odontoma and the adenomatoid odontogenic tumor.[9,19,24] Five malignant tumors have been reported.[15,25,26]

Ghost cell keratinization, the characteristic microscopic feature of this cyst is also a defining feature of the cutaneous lesion known as calcifying epithelioma of Malherbe or pilomatrixoma.[27] Actually these cysts share more histologic features with the rare intracranial neoplasm known as craniopharyngioma.[28]

In the jaws, apart from CEOC, ghost cells may also be seen in other odontogenic tumors including odontomas, ameloblastomas, adenomatoid odontogenic tumors, ameloblastic fibro-odontomas and ameloblastic fibromas.[27,28]

Enucleation is the preferred mode of treatment for simple cystic lesions.[29] Recurrence is unusual.[30] A follow-up period of at least 10 years is, therefore, recommended.[29]

CONCLUSION

To find CCOT in the edentulous region is an extremely rare and new finding which is seen in our case. So, whenever a patient comes with an intraoral swelling anterior to first molar region whether dentulous or edentulous, as an oral physician we should always keep CEOC as a differential diagnosis in our mind. As it does not have any pathognomonic, clinical and radiological features, so the definitive diagnosis of this lesion remains dependent on histological evaluation.

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