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Case Report

A Case Report of Distinctive Entity - Dentinogenic Ghost Cell Tumor

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Abstract

The Dentinogenic ghost cell tumor (DGCT), a benign neoplasm, causes bone destruction and cortical expansion, despite being locally invasive and associated with a risk of local recurrence. This article portrays a rare case of 22-year-old male patient who reported with a chief complaint of recurrent swelling on right side of the face since 1 month. Multi-slice multiplanar plain CT was carried out which revealed abnormal lytic expansile lesion involving the alveolar process of the maxilla on right side. The final diagnosis of the lesion was made Dentinogenic ghost cell tumor with histopathological investigation which revealed it to be rarest entity.

Keywords: Calcifying Epithelial; Odontogenic Cyst; Dentinogenic Ghost Cell Tumor; Maxilla; Recurrence

Abbreviations

CEOC: Calcifying Epithelial Odontogenic Cysts; WHO: World Health Organization; COC: Calcifying Odontogenic Cysts; CCOT: Calcifying Cystic Odontogenic Tumor; DGCT: Dentinogenic Ghost Cell Tumor

Introduction

Dentinogenic ghost cell tumor is a rare neoplasm thought about as solid variants of calcifying epithelial odontogenic cysts (CEOC) constituting soley 11.5% of all COCs [1]. Calcifying odontogenic cyst was first delineated by Gorlin and his colleagues in 1962. In 1971, WHO included enclosed it under classification of histological typing of odontogenic tumor, jaw cyst and allied lesions [2]. In 1981, Praetorious., *et al.* classified COC into cystic and solid neoplastic type and so the term DGCT was planned for the neoplastic type [3]. The term dentino-ameloblastoma, was used by Shear in 1983 due to resemblance of the features to the ameloblastoma and dentinoid production [4]. It was renamed COC as calcifying cystic odontogenic tumor (CCOT) and was included by WHO in 2005 and maintained the term DGCT for the neoplastic type [5].

They are locally invasive neoplasms which usually occurs in elderly persons with a slight male predilection. Their main characteristic features are ameloblastoma like odontogenic epithelial proliferation and exhibits aberrant keratinization in the form of ghost cells and dysplastic dentin [1]. Hong., *et al*. classified it in two forms intraosseous (central) and extra osseous (peripheral), among which intraosseous is more aggressive variety which requires careful monitoring and local resection to prevent recurrence.

This article depicts an interesting case of DGCT, treatment adopted in this case and review of this case in the indexed literature.

Case Presentation

A 22-year-old male patient was reported to the department of Oral Medicine and Radiology with the chief complain of swelling on

right side of the face since one month. As reported by him, the swelling was slowly progressive in nature and did not give any history of pain associated with it.

It was reported that he had similar swelling in the same region of the jaw in November 2016, for which he had undergone incisional biopsy. The histopathological reports revealed presence of spindle cells suggestive of Adenomatoid tumor. In December 2016, excision of right maxillary tumor was performed under General anesthesia. Enucleation was executed by taking an incision from region 12 to 18 and a mucoperiosteal flap was raised, following this extraction of 16,17,18 was performed. Again, the specimen was sent for histo-pathological evaluation, which revealed presence of ghost cells suggestive of Dentinogenic ghost cell tumor. Patient was under follow up and did not reveal any signs of recurrence for 3 years. No relevant Family history was noted and all the vital and peripheral signs were within normal limits.

Extra-Oral examination on inspection exhibited a solitary, diffuse swelling about 3x4 cm in size, extending Anterio-posteriorly from the lateral margin of nose to preauricular region, Supero-inferiorly extending from the infra orbital margin to middle 1/3rd of face on right side. On inspection the swelling was non- tender and soft in nature (Figure 1).

Figure 1: Extra-Oral Photograph showed a solitary, diffuse swelling.

Intra-Oral examination on inspection revealed a solitary swelling about 3x4cm arising from the buccal and lingual alveolar bone extending from the distal surface of 14 to the right maxillary tuberosity. On palpation it was mildly tender, bony hard in nature. Clinically missing 15,16,17,18 was noted (Figure 2).

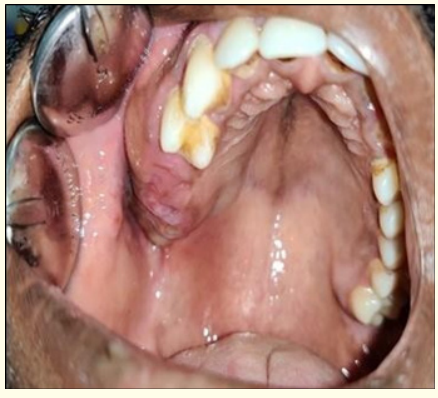

Figure 2: Intra-oral photograph revealed a solitary swelling about 3x4cm arising from the buccal and lingual alveolar bone extending from the distal surface of 14 to the right maxillary tuberosity.

On account of clinical examination, a provisional diagnosis of Benign Odontogenic tumor most likely to be an Adenomatoid tumor, Dentinogenic ghost cell tumor, Ameloblastoma were specified.

Various radiographic investigations were carried out which consists of an Orthopantomograph and Computed tomography scan. Orthopantomograph revealed an ill- defined radiolucency, homogenous in nature extending antero-posteriorly from the periapical region of 14 to right maxillary tuberosity. Supero- inferiorly involving the alveolar crest bone from 14 to18 region. Evidence of External root resorption with 13,14 and thinning of alveolar crest bone were noted (Figure 3).

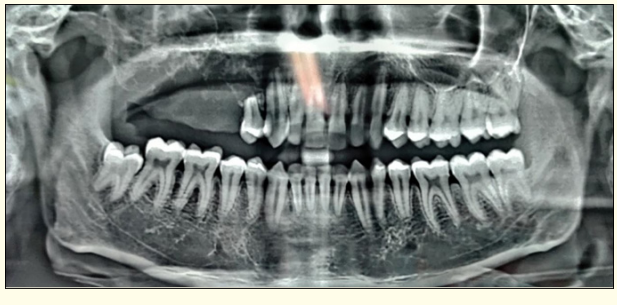

Figure 3: Pre-op Orthopantomograph shows an ill-defined homogenous radiolucency, extending antero-posteriorly from the periapical region of 14 to maxillary tuberosity. Supero-inferiorly involving the alveolar crest bone from 14 to18 region. External root resorption with 13,14 and thinning of alveolar crest bone.

Multislice multiplanar plain CT of maxilla revealed abnormal lytic expansile lesion involving the alveolar process of the right-side maxilla. It showed multiple thin septa within, involves hard palate and also extends into the maxillary antrum involving its lateral wall (Figure 4). Radiographic differential diagnosis included adenomatoid tumor, Ameloblastoma.

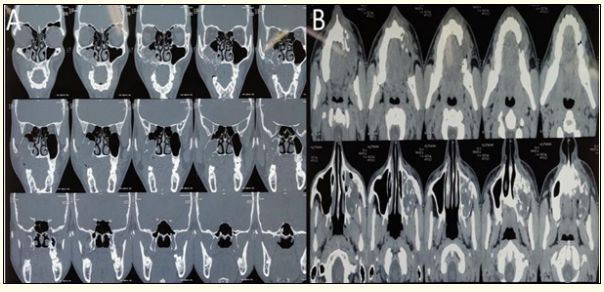

Figure 4: CT of maxilla revealed abnormal lytic expansile lesion involving the alveolar process of the maxilla on right side. It showed multiple thin septa within, involves hard palate and also extends into the maxillary antrum involving its lateral wall.

Followed by radiographic examination all necessary hematological investigation were performed and was immediately posted for surgery, hemimaxillectomy under general anesthesia was performed and was sent for histopathological evaluation for definitive diagnosis of lesion. Patient was under observation for 7 days and advised follow up after every 6 months.

Histopathological reports revealed areas of keratinization, islands of ghost cells and had foci with increased mitotic activity (Figure 5). Overall features were suggestive of Dentinogenic ghost cell tumor. Orthopantomograph was performed on follow up which showed well defined radiopacity in the form of wire from 11-28 suggestive of interdental arch bar wiring. Well defined radiolucency extending from distal surface of 11 to right maxillary tuberosity suggestive of surgical defect. It revealed no evidence of recurrence. Patient was provided with an interim obturator (Figure 6) and recalled after 6 months for follow-up.

Discussion

In the literature since 1962 till now, terminologies and classifications of COC have been proposed and practiced. Fejerskov and Krogh in 1972 suggested the term "calcifying ghost cell odontogenic tumor" [5].

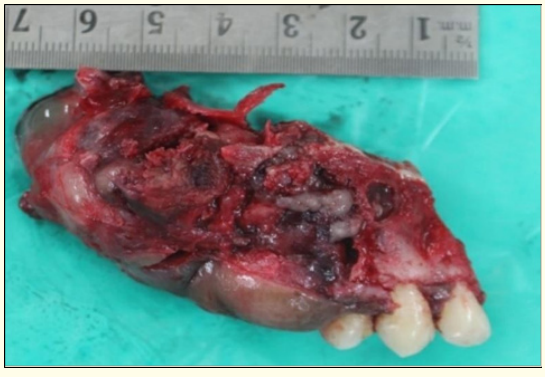

Figure 5: Tissue Specimen after biopsy.

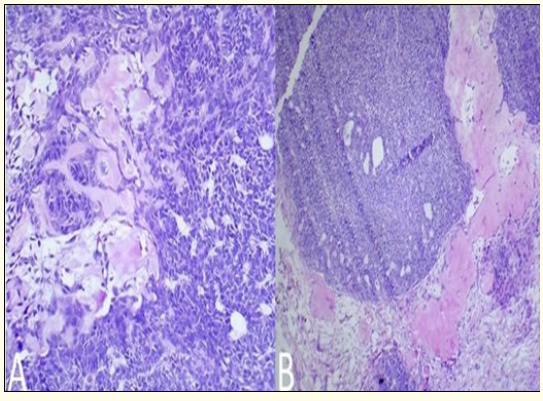

Figure 6: Histopathological slide shows areas of keratinization, islands of ghost cells and foci with increased mitotic activity.

The term COC is not entirely appropriate, because there could be the possibility of cystic degeneration taking place in the center of proliferating epithelial islands rather than epithelial changes developing in a pre-existing cyst wall. The presence of the ghost cells which might exhibit calcification and its proliferative potentiality may give rise to lesions of considerable size were some concerns raised by the authors.

In 1991, Buchner., *et al.* clinically classified COCs into central and peripheral lesions and further sub- classified each of them into cystic or neoplastic variants [6].

WHO classification given in 2005 has solved the conundrum related to the term "cystic" which was considered similar for "non-neoplastic" and could be misleading as there may be lesions which

demonstrates cystic architecture and still have extensive proliferative capacity. According to the WHO, the spectrum of odontogenic ghost cell tumors comprises CCOT, DGCT and ghost cell odontogenic carcinoma [7].

The latest edition of the World Health Organization Classification of Head and Neck Tumors described DGCT is a benign neoplasm, locally invasive, classified as a mixed epithelial and mesenchymal odontogenic tumor with about 60 cases described in the literature [7,8].

It can be central/intraosseous or, sporadically, peripheral/extraosseous, the former having a more aggressive behavior which influences its evolution. In our case the intraosseous variety of DGCT was found that is most commonly noted in the posterior region of the jaw [9].

The peak incidence is not yet consensual, but occurs mostly from second to fourth decade like the patient in our case.

The clinical features of DGCT variants include visible swelling, which can be painful or painless with obvious facial asymmetry due to expansion of the jaw, with occasional occurrence of obliteration of the maxillary sinus or infiltration of the soft tissues which may occasionally accompanied by pus discharge, tooth displacement or mobility. According to the findings stated by konstantakis., *et al.* in 2013, DGCT may appear radiographically as radiolucent, radiopaque or mixed lesion. Lesions can be unilocular or multilocular with either well-defined or ill-demarcated margins [10]. This case portrayed an unilocular radiolucent lesion with ill-defined borders. Thinning of maxillary sinus walls was noted in our case and external root resorption of the involved teeth was noted.

Histo-pathologically DGCT is characterized by sheets and rounded islands of odontogenic epithelial cells seen in a mature connective tissue stroma [10]. A characteristic feature of DGCT is ghost cells, they are ellipsoidal keratinized epithelial cells with basic cellular outlines but loss of nuclei, with formation of foreign body granuloma and has potential to calcify. All these features can be identified using special stains such as van Gieson, Goldner, or Ayoub-Shklar histochemical stains.

The transformation of epithelial cells, metaplastic transformation of odontogenic epithelium, and squamous metaplasia with secondary calcification due to ischemia, degeneration of epithelial cells or as a result of apoptotic process are some of the theories of origin of ghost cells postulated in literature. The ghost cells are identified in other neoplasms such as odontomas, ameloblastomas and ameloblastic fibro- odontomas hence alone are not pathognonic of DGCT.

The one of the characteristic histopathological feature of DGCT is formation of dentinoid or osteoid material. This material is usually found in relation to ghost cells. According to Bafna., *et al*. dentinoid or osteoid material is mesodermal in origin because it is not found in luminal proliferation unless there is breach in the continuity of basement membrane due to outgrowth of connective tissue between the epithelial ghost cells [11]. Gorlin and his colleagues described it an inflammatory response of the body tissue to the masses of ghost cells. On further investigation by Abrams and Howell further stated that these masses of "ghost cells" induce granulation tissue formation which lays down juxtra-epithelial osteoid that may further calcify. On the other hand, Singhaniya., *et al.* were of the opinion that dentinoid stands for a metaplastic change in the connective tissue without the participation of granulation tissue [3].

It was stated by Soluk Tekkesin., *et al*. [12] that intraosseous DGCTs are more aggressive than their extraosseous counterparts. For intraosseous variant, if the tumor is showing ill-defined borders radiographically complete removal of the tumor is highly recommended in such cases. Depending upon its size or anatomic extent, tumor may require block excision or segmental mandibular resection or partial maxillectomy Our case was treated by partial maxillectomy. Sun., *et al.* found that DGCT's have a high rate of recurrence [13]. Our case has shown recurrence once. Currently, no significant signs of recurrence were noted.

Conclusion

Dentinogenic ghost cell tumor (DGCT) accounts about 1.9% to 2.1% of all odontogenic tumors and can be considered as a very rare neoplasm. In conclusion, we presented a rare recurrent case of DGCT in posterior maxilla highlighting its clinical, radiological and histopathological features. For accurate diagnosis circumspective investigation of the specimen by an adept oral pathologist should be requested in order to choose an appropriate treatment approach.

Acknowledgements

None.

Conflict of Interest

None.

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