

Intraosseous Dentinogenic Ghost Cell Tumor – A rare entity : a case report

Shubham P. Palve¹, Neelam N.Andrade¹, Trupti M.Gandhewar¹, Varsha Gupta¹, Shelly Sharma¹, Reshma Gaikwad¹.

¹Department of Oral and maxillofacial surgery, Nair Hospital Dental College, Mumbai

Corresponding Author

Shubham P. Palve

E-mail ID: drshubhampalve@gmail.com

Abstract

Dentinogenic ghost cell tumor (DGCT) is an infrequent, neoplastic form of the Calcifying Odontogenic Cyst (COC). It has local invasive behavior and accounts for 2-14% of all COCs. The intraosseous DGCT is less likely to occur and less literature is available for the recurrent lesion. Hereby, we present the case of a 22-year-old male patient having recurrent DGCT in the right posterior maxilla. This patient was managed by hemimaxillectomy followed by immediate obturator and close monitoring for detection of recurrence.

Keywords: Calcifying Odontogenic Cyst (COC), Dentinogenic, Ghost cell tumor, Central (Intraosseous), Hemimaxillectomy

Introduction

Dentinogenic ghost cell tumor (DGCT) is an infrequent odontogenic neoplasm and solid type of the calcifying odontogenic cyst (COC)⁽¹⁾. It is characterized by ameloblastoma like epithelial islands, ghost cells and dentinoid material⁽²⁾. Literature reveals two types of DGCT, central (intraosseous) and peripheral (extraosseous). Central lesion is locally aggressive with bony enlargement and has high recurrence rate compared to the peripheral lesion^(3,4). Various treatments have been described for DGCT like marsupialization, enucleation and aggressive resection^(5,6).

Case presentation

A 22-year old male patient reported to our department with chief complaint of soft-tissue overgrowth in the upper right posterior region of the jaw since 3 months. Initially, growth was smaller in size which gradually increased over a period of time. It was associated with occasional mild pain and epistaxis. Patient had history of surgery for DGCT in the same region 4 years ago. No significant findings noticed on extraoral examination. Intraoral examination revealed solitary, globular soft tissue swelling on the right maxillary posterior region of jaw extending from mesial side of the first premolar to tuberosity region along with buccopalatal expansion (Figure 1-A).

The outgrowth was firm in consistency and non-tender on palpation. Maxillary right second premolar to third molar teeth were missing due to previous surgery. Indentations of lower teeth were seen over the swelling.

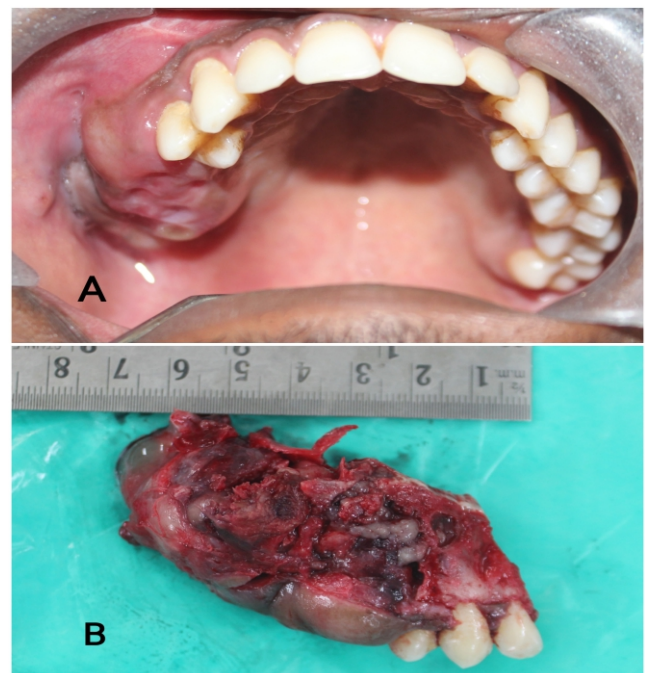


Figure 1-A Preoperative clinical image : showing globular swelling of the right posterior maxillary region with bucco-palatal expansion

Figure 1-B Hemimaxillectomy : Excisional mass showing the resected lesion

Orthopantomogram showed well-defined unilocular radiolucency from first premolar to right maxillary tuberosity region along with root resorption of the first premolar. Computed Tomography scan was carried out to check the extent of lesion. It showed well defined large, rounded,

expansile lesion (4.6x3.3x4cm) involving right posterior maxillary alveolar process, adjacent hard palate and also extended into lateral wall of right maxillary antrum. Mucosal thickening was evident in right maxillary sinus (Figure 2).

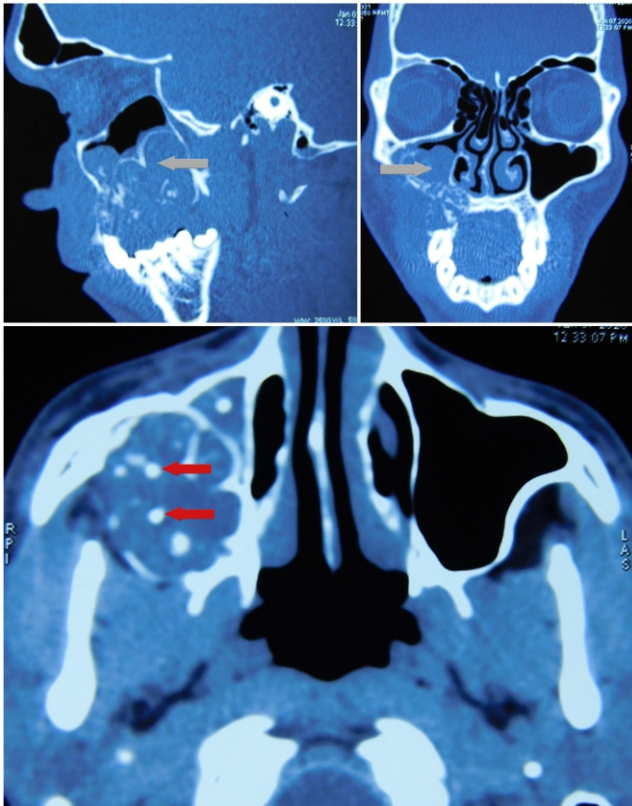


Figure 2-Preoperative Computed Tomography scan : Sagittal,Coronal and Axial planes showing well defined large, rounded radiolucency(Grey arrow) along with the radio-opaque material(red arrow) in right maxillary antrum.Note the involvement of the lesion into right maxillary alveolar process and lateral wall of the maxillary sinus

The overall appearance at that stage was suggestive of a Recurrent DGCT of the right maxilla. The procedure to be performed was explained to the patient preoperatively followed by written consent. Under general anesthesia, vestibular incision was given from the left lateral incisor to right maxillary tuberosity region. A full-thickness mucoperiosteal flap was raised to expose the underlying tumor mass. Hemimaxillectomy (Figure 1-B) was carried out and post-operatively obturator was placed over the defect and stabilized with the help of interdental wires to the left maxillary teeth. The patient was kept on regular follow-up. Histopathologically excisional tissue showed solid lesion comprising of odontogenic epithelium with prominent ghost cells which was suggestive of DGCT (Figure 3).

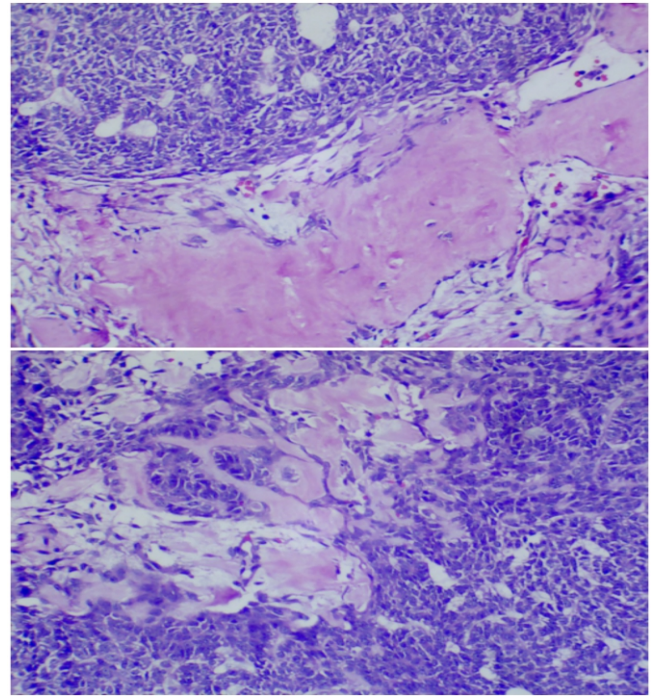


Figure 3-Histopathology of the Resected tissue : Haematoxylin and eosin staining of odontogenic epithelium with prominent ghost cell formation

Result

After 1 year, a CT scan was done which revealed no evidence of tumor recurrence. (Figure 4).

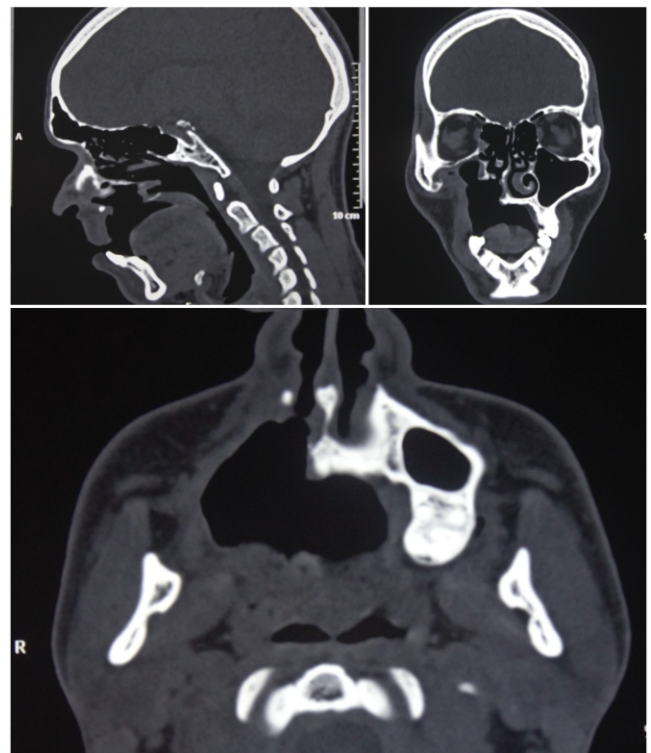


Figure 4 - Follow-up Computed Tomography scan : Sagittal,Coronal and Axial planes showing no recurrence of the lesion on right side of Maxilla

Discussion

COCs were first introduced by **Gorlin et al** in 1962⁽⁷⁾. According to WHO(2005), there are three types of COCs, namely DGCT, ghost cell odontogenic carcinoma (GCOC) and calcifying cystic odontogenic tumor (CCOT)⁽²⁾. COCs represent 1-2% of all odontogenic tumors and among them 2-14% are DGCT⁽³⁾. In 2005, WHO defined DGCT as, “A locally invasive neoplasm characterized by ameloblastoma like islands of epithelial cells in a mature connective tissue stroma, aberrant keratinization may be found in the form of ghost cells along with varying amounts of dysplastic dentin”⁽²⁾. DGCT occurs mostly in posterior mandibular region with slight male predilection and age ranging from 12 to 79 years (mean 39.7+/-19.3 years)⁽⁸⁾. The tumor presented in this case was of a right maxillary intraosseous DGCT in a 22-year-old male individual.

In the literature, various treatments have been described for DGCT such as marsupialization, enucleation, and aggressive resection. **Sun et al (2009)** reported high rate of recurrence during the follow-up period after local curettage⁽⁹⁾. Multiple recurrences have been linked with the malignant transformation of the DGCT into GCOC⁽⁹⁾. Apart from recurrence of DGCT on the primary site, distant metastasis to the graft donor sites have been also reported⁽¹⁰⁾. Preferred mode of treatment of intraosseous DGCT consists of extensive bone resection with safety margin of at least 5 mm as advocated for ameloblastoma⁽⁶⁾. In our case, hemimaxillectomy was performed, reason was the propensity of the tumor to recur after treatment. Recurrence rate was 73 % in lesion treated by conservative surgery and 33 % by radical surgery after 1 year of follow up⁽⁸⁾. The present case has no recurrence after 1.5 years of follow up.

Conclusion

The present case report represents the surgical treatment of recurrent intraosseous DGCT. Although intraosseous DGCTs are rare, the general dental practitioner should not exclude the likelihood of encountering a case of DGCT in routine clinical practice. A thorough clinical and radiographic examination is crucial for this specific entity. Referral of the patient for biopsy, histologic analysis, surgical intervention, and a close long-term follow-up to inspect a recurrence are necessary for the successful treatment of the patient.

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Conflict of interest: Nil

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