CALCIFYING GHOST CELL ODONTOGENIC CYST: A CASE REPORT & A REVIEW OF LITERATURE

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Abstract:
Background: This is a case of Calcifying Ghost Cell Odontogenic Cyst (CGCOC) which aims to collate the findings of studies done on ghost cells, to pave the path for further investigative research to better understand the pathogenesis of lesions showing ghost cell formation. Materials & Methods: A 13 year old female patient presented with a swelling on the right lower jaw. Histopathological evaluation of biopsied tissue was done using haematoxylin and eosin stain. Results: An abundance of ghost cells both in the cystic lumen and connective tissue stroma was revealed in the tissue specimen. Conclusion: The occurrence of non-neoplastic and neoplastic variants of the lesion with similar cellular and histomorphologic features determine choice of therapeutic modalities and follow up. The unravelling of the true nature of ghost cells can throw light on the dualistic nature of this lesion.
Key-words: Calcifying odontogenic cyst, ghost cells, aberrant keratinisation.

INTRODUCTION

The Calcifying Odontogenic Cyst (COC) was first described by Rywkind as in 1932 and separated as a distinct pathological entity by Gorlin et al in 1962.[1] It represents two percent of all Odontogenic Cysts and Tumours.[1,2] The nature of this lesion is controversial, where the term cystic seems to be synonymous with non-neoplastic. The histopathogenesis of the centrally located COC is attributed to the reduced enamel epithelium or remnants of odontogenic epithelium. The exact origin of ghost cells seen in this lesion is not yet known but several theories have been proposed.

CASE REPORT

A 13 year old female patient reported to the department of Oral Medicine & Radiology with a six month history of a progressively increasing swelling on her right lower jaw. There were neither accompanying symptoms nor any significant medical and family history.

The extra-oral examination denoted a subtle increase in volume over the right cheek, not involving the nasolabial fold. The intraoral examination revealed a solitary well circumscribed swelling, measuring three by two centimetres in the right buccal vestibule and alveolus, extending from right lower canine to right lower second premolar. Obliteration of the buccal vestibule was seen along with bony expansion of the buccal cortical plate. Oral mucosa appeared taut and blanched over the swelling. Right lower first premolar was not clinically evident. The swelling was soft, non-fluctuant and non-tender. Teeth of the right lower quadrant were vital on electric pulp testing and not associated with mobility. Routine haematological tests revealed normal values. There was no fluid yield on aspiration.
Radiographic investigations showed a well-defined, mixed radiolucent-radiopaque lesion associated with the impacted first premolar, extending from its distal margin to the mesial margin of the second premolar root, abutting on the mandibular foramen. Mild displacement of first and second premolars was observed, with no evidence of root resorption. The mandibular true occlusal radiograph confirmed the gross expansion of the buccal cortical plate. The differential diagnosis included: Calcifying Epithelial Odontogenic Tumor, Calcifying Odontogenic Cyst, Adenomatoid Odontogenic Tumor and Ameloblastic Fibro-odontoma. The lesion was enucleated under general anaesthesia by raising a mucoperiosteal flap and submitted for histopathological evaluation. Gross: The enucleated specimen was cystic, oblong in shape, one to one and a half centimetres in diameter. The haematoxylin and eosin stained section showed a cystic lesion with a fibrous connective tissue capsule lined by non-keratinizing odontogenic epithelium with cuboidal –columnar palisaded basal cells with polarized hyperchromatic nuclei. (Figure 1)

Some ghost cells showed dystrophic calcification. (Figure 2)

![Figure 2: (100 X) Haematoxylin and eosin stained section showing basal layer of cuboidal columnar palisaded cells with polarized & hyperchromatic nuclei.](image)

The connective tissue stroma was fibrovascular with irregular foci of atubular dentin. (Figure 3)

![Figure 3: (100X) Haematoxylin and eosin & stained sections showing irregular foci of atubular dentin within connective tissue stroma.](image)

The postoperative course was satisfactory, with no sign of recurrence on six monthly follow-up, till date.

**DISCUSSION**

Shear (1983) preferred the term “Dentinoblastoma”; Ellis & Schmookler (1986) suggested the term “Epithelial Odontogenic Ghost Cell Tumor” as epithelial cells appearing like ghost cells were the most distinctive feature of this neoplasm; Colmenero et al (1990) put forth the term “ Odontogenic Ghost Cell Tumor” for the neoplastic form of COC. \[3,4\]

The COCs were sub-divided into three distinct entities \[5,6\]
- Calcifying Odontogenic Tumor- locally invasive
Dentinogenic Ghost Cell Tumor - with clinical and histopathological similarities with an ameloblastoma.

Ghost Cell Odontogenic Carcinoma - very aggressive with high recurrence rate.

The two variants of COC are the central (intraosseous) and the peripheral (extraosseous) variant. Our case is the former. The radiographic features depend upon the maturity of the lesion when detected.

The COC lacks pathognomonic clinical, radiographic features. Histopathological evaluation remains the gold standard to arrive at a conclusive diagnosis.

The classic features include a fibrous capsule lined with odontogenic epithelium, with elliptical eosinophilic epithelial cells (Ghost cells), the presence of dystrophic calcification, and dentinoid in the stroma.

A number of immunohistochemical studies have been undertaken to analyse the true nature of ghost cells and their formation. Results of some of these studies have been summarized in Table I.

Ghost cells are also considered to be foreign bodies within connective tissue which induce granulation tissue response. This response further initiates juxta-epithelial degeneration of ghost cells which form foci for dystrophic calcification via the Notch1-Jagged1 ‘lateral –induction’ pathway.

Further research on the molecular pathogenesis of ghost cells might shed light on the etiopathogenesis of this rare odontogenic cyst thus paving a way for a targeted treatment protocol.

REFERENCES


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