

## Dentinogenic Ghost Cell Tumor: An Unusual Aggressive Oral Lesion

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### Abstract

The purpose of this report was to document a case with a 1-week and 2-month follow up and contribute to the small body of existing literature to better understand this rare odontogenic tumor. There have been few aggressive and large dentinogenic ghost cell tumor case reports found in this literature.

**Keywords:** Dentinogenic ghost cell tumor, Ghost cell, Dentinoid material.

### Introduction

The head and neck regions are well known to be afflicted with a great variety of odontogenic and non-odontogenic cysts and tumors. Dental professionals are very familiar with the appearance of most of the common lesions as well as some of the uncommon, yet “classic” lesions. Some uncommon lesions may present with an appearance that can confound a clinician as to its diagnosis and treatment. Many different terms have been used to describe the dentinogenic ghost cell tumors (DGCT). This tumor represents a subset that makes up 11.5% of calcifying cystic ghost cell odontogenic tumors and has throughout the years gone by a variety of names such as “dentinoameloblastoma” by Shear and “epithelial odontogenic ghost cell tumor” by Ellis and Shmookler.<sup>1</sup> In 2005, the World Health Organization coined the term “dentinogenic ghost cell tumor,” initially used by Praetorius in 1981, due to the differentiating formation of dentinoids noted in its histology. This tumor is classified as a locally invasive neoplasm with ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma. There can be aberrant keratinization in the form of ghost cells found in association with varying amounts of dysplastic dentin.<sup>2</sup>

### Case Presentation

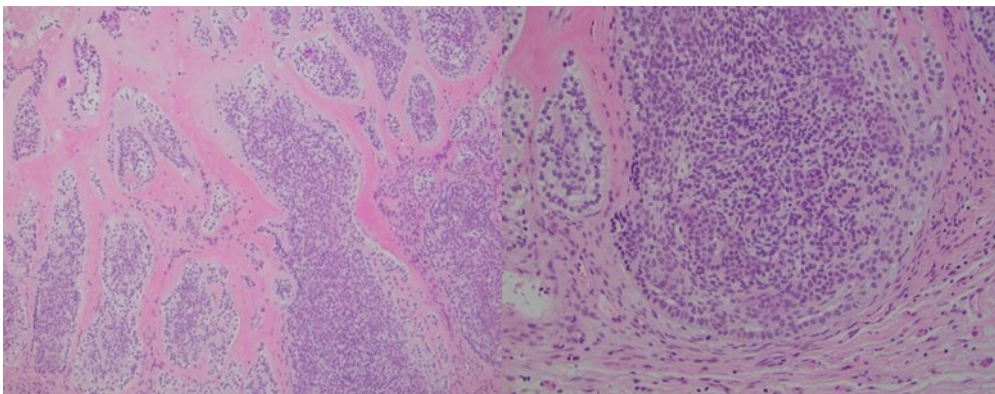
A 60-year-old African American male presented to the oral surgery suite at the Rocky Mountain Regional Veterans Administration Medical Center, USA. He was referred by his primary dentist who noted the lesion during a routine dental visit. At presentation his chief complaint was “my gums are tender up here,” pointing to his upper right quadrant around the buccal aspect of tooth #3. His history of present illness consisted of tenderness, in addition to intermittent hot and cold sensitivity for three months. Medical, dental, and family history was not relevant, and all vital signs were within normal limits. The patient reported no history of trauma.

The intraoral clinical examination revealed a 12mm x 8mm x 4mm enlargement of buccal tissue extending from the free gingival margin of #3 to the depth of the vestibule. The tissue was pink, firm, and highly keratinized in appearance with rolled gingival margin borders. The swelling was firm and tender to palpation. There was no drainage or apparent communication when periodontal probing was performed. The periapical radiographs revealed a similarly sized radiolucency at the periapical of #3 with non-corticated, moderately well-defined borders extending towards the mesial of tooth #2 and distal of tooth #4. Severe resorption of the mesio- buccal and disto-buccal roots of #3 was noted. The cone-beam computed tomography revealed severe resorption of mesio-buccal and disto-buccal roots of tooth #3 with the lesion expanding through the buccal plate.

An incisional biopsy of the firm mass was performed on the upper right buccal mucosa above #3: A 15-blade was used to make 10mm x 5mm incision to periosteum of excision of both normal and atypical tissue. The final specimen measured 10mm x 6mm x 5mm. The differential diagnoses consisted of pleomorphic adenoma, fibroma, or mucoepidermoid carcinoma.

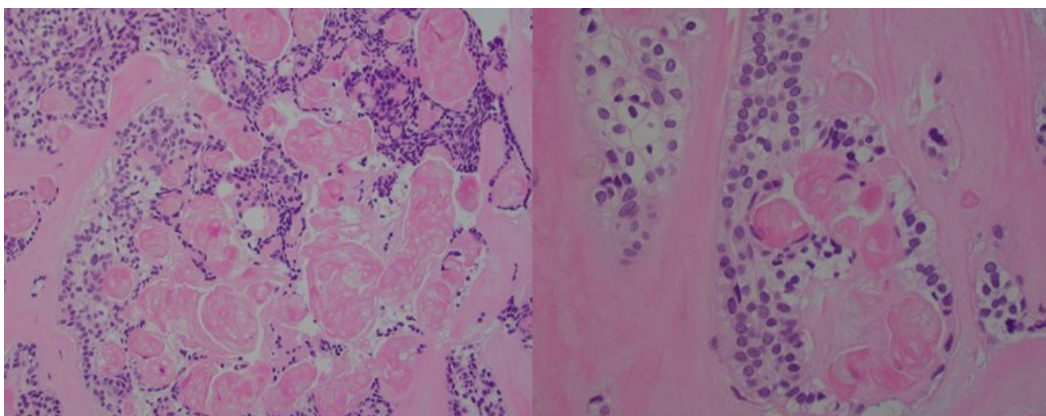
Histopathological examination determined that the lesion was a dentinogenic ghost cell tumor (DGCT). The sections showed a bisected portion of oral squamous mucosa with an underlying circumscribed epithelial neoplasm that exhibited a pushing but infiltrative border. The lesion was composed of variably sized nests, islands, and cords of odontogenic epithelium, many of which that had an ameloblastic-like peripheral palisading. Prominent ghost cell keratinization was observed in the tumor nests and frequent clear cell change. The neoplastic tissue was supported by a densely hyalinized stroma. Immunostains with adequate controls showed the tumor was positive for beta-catenin (strong nuclear staining), CK 5/6, and CK 19. Ki-67 was slightly elevated in the peripheral cells of the islands. Negative stains included BRAF, S100 and SOX-

This odontogenic tumor had potentially locally aggressive characteristics, and complete removal was recommended.

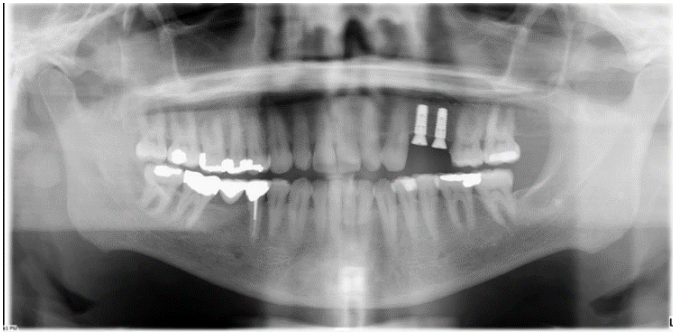


**Figure 1:** Variably sized nests, islands and cords of odontogenic epithelium in a background of densely hyalinized stroma (H &E, 100X)

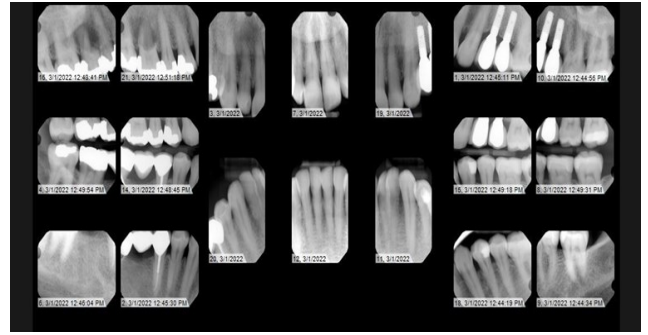
**Figure 2:** Ameloblast-like peripheral palisading seen around one of the nests (H & E, 200X).



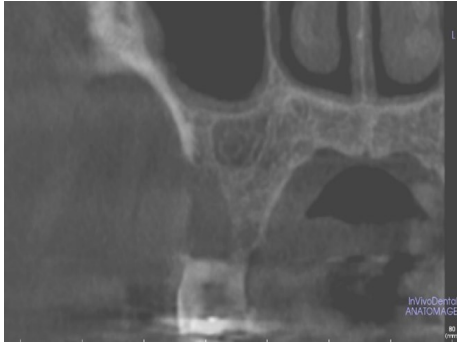
**Figures 3 and 4:** Prominent ghost cell keratinization in the tumor nests (H & E, 200X).



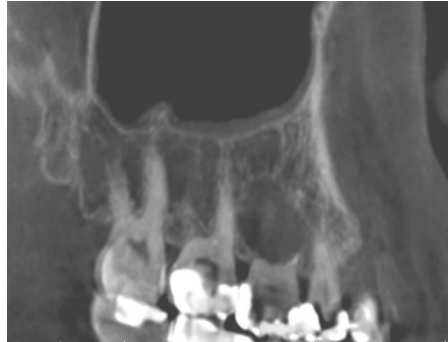
(Pano)



(FMX)



(Coronal)



(Sagittal)



(Axial)



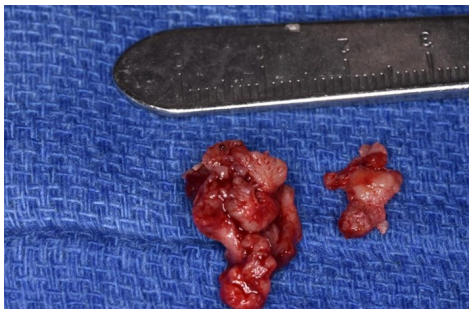
(Biopsy)



(Initial Presentation for Biopsy)



(Pre-Op)



(Lesion)



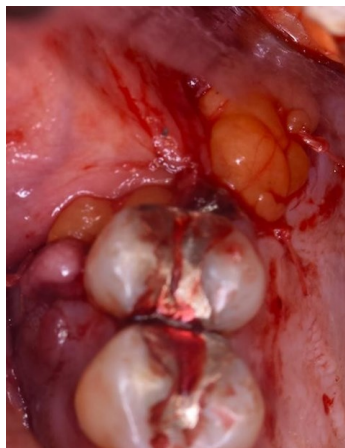
(Ext#3)



(Ext#2/3 and Tumor)



(Post Marginal Resection)



(Post Closure w/Buccal Fat Pad)



(One week f/u; heme from some sutures removed)

*(2 Month Follow-Up)*

## Discussion

A dentinogenic ghost cell tumor (DGCT) is a rare entity with only about 60 cases reported in the literature.<sup>3</sup> DGCT's are benign, mixed epithelial and mesenchymal odontogenic tumors with locally aggressive behavior. They are a subset of Calcifying Odontogenic Tumors with both a cystic type (Type I) and a solid type (Type II).<sup>4</sup> The tumors are more frequently intraosseous in the posterior maxilla and mandible. If extraosseous in position, it is seen typically in the anterior part of the jawbone.<sup>5</sup> DGCTs affect mainly male patients between the fourth and sixth decades, although cases have been presented outside of that range.<sup>5</sup> The symptoms are unspecific, and a vast percentage of patients are asymptomatic. Occasionally, patients experience pain and swelling caused by cortical bone expansion.<sup>5</sup> Radiographically, a unilocular feature is typically found. Commonly, dentinogenic ghost cell tumors cause cortical resorption and extend into the soft tissues.<sup>5</sup>

Microscopically, dentinogenic ghost cell tumors typically show the following histological features: prominent basaloid to stellate reticulum cells lined by peripheral columnar cells, areas of calcification, and keratinization with ghost cells.<sup>5</sup> The two characteristic features of DGCT that distinguish it from an ameloblastoma and other odontogenic tumors are numerous ghost cells and masses of dentinoid material.<sup>3</sup>

Excision is recommended for intraosseous lesions and a more conservative excision is recommended for extraosseous tumors. It is recommended to perform a wide range of resection due to the high recurrence rate, and therefore, long-term follow-up is recommended. In cases where conservative surgery was performed, a recurrence rate of 73% is reported. In cases that are treated with more radical surgery, the recurrence rate, although still noticeable, is reduced to 33%.<sup>3</sup>

In our case following the initial biopsy, the surgical plan was for wide local excision of the tumor with extraction of teeth #2 and #3. Informed consent was obtained, and the patient was taken to the operating room to perform the procedure under general anesthesia. A full thickness mucoperiosteal flap was reflected and teeth #2 and #3 were extracted. #3 was confirmed to have severe root resorption as seen on the pre-operative radiographs. Marginal resection of the tumor was performed, and the site was closed with the patient's buccal fat pad with 3-0 chromic gut sutures in a continuous fashion. The tissue samples were sent to pathology. A one-week, two-week, and two-month follow-up have been performed and the patient is following a normal course of healing. The buccal fat pad has epithelized, and the patient has been slowly regaining full range of motion. Due to the high recurrence rate, the patient will continue to have regular follow-up appointments.

## Conclusion

In conclusion, this case describes the clinical, radiographical, and pathological presentation of dentinogenic ghost cell tumors. Early diagnosis plays a key part in preventing further resorption, bony expansion, and radical surgical approaches. Although odontogenic tumors have a wide range of clinical presentations, in rare instances, a dentinogenic ghost cell tumor should be considered.

## Declarations of Interest

None

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