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CASE REPORT

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A rare case of dentinogenic ghost cell tumor with concomitant odontoma

Tatsuo Okui[1] | **Reon Morioka**[1] | **Teruaki Iwahashi**[2] | **Yuhei Matsuda**[1] | **Shinji Ishizuka**[1] | **Satoe Okuma**[1] | **Hiroto Tatsumi**[1] | **Takahiro Kanno**[1]

[1]Department of Oral and Maxillofacial Surgery, Shimane University Faculty of Medicine, Izumo, Japan

[2]Department of Surgical Pathology, Shimane University Faculty of Medicine, Izumo, Japan

Correspondence
Tatsuo Okui, Department of Oral and Maxillofacial Surgery, Shimane University Faculty of Medicine, 89-1 Enya-cho, Izumo, Shimane 693-8501, Japan.
Email: tokui@med.shimane-u.ac.jp

Key Clinical Message

A case of dentinogenic ghost cell tumor occurring simultaneously with a clinically diagnosed odontoma. The occurrence of epithelial and mesenchymal tumors at the same site is very rare, but should be kept in mind during pathological diagnosis.

Abstract

Dentinogenic ghost cell tumor (DGCT) is a rare and benign odontogenic tumor composed of ghost cells, calcified tissue, and dentin. We present an extremely rare case of a 32-year-old female who was clinically diagnosed with an odontoma presenting with a painless swelling in her maxilla. Radiographic examination showed a well-defined radiolucent lesion with tooth-like calcified areas. The tumor was resected under general anesthesia. No recurrence was noted at the 12-month follow-up. Histopathological examination of the surgically resected tumor yielded a diagnosis of DGCT with odontoma.

KEYWORDS

dentinogenic ghost cell tumor, odontogenic tumor, odontoma

1 | INTRODUCTION

Dentinogenic ghost cell tumor (DGCT), a rare odontogenic tumor accounting for less than 0.5% of all odontogenic tumors, arises from the epithelial remnants of dental lamina or enamel.[1] Odontoma, a benign tumor-like lesion, consists of dental tissues such as enamel, dentin, cementum, and pulp.[2] Although both tumors are benign, they may cause complications depending on their location and size. DGCT is usually asymptomatic and presents as a painless swelling in the oral and maxillofacial region. Radiographically, DGCT appears as a well-defined radiolucent lesion with calcified areas.[3] Histopathologically, it is characterized by the presence of ghost cells, dentin, and calcified tissue.[4] Although DGCT is considered a benign tumor, it can be locally aggressive and has the potential to recur.[5] Here we present an extremely rare case of combined DGCT and odontoma, in which lesions with each pathology occurred at the same site simultaneously in a middle-aged female patient.

2 | CASE REPORT

A 32-year-old female presented to Shimane University Hospital with a painless swelling in her left maxilla that had

been present for approximately 3 months. Clinical examination revealed a firm, non-tender swelling in the left maxilla region (Figure 1A). Radiographic examination showed a well-defined radiolucent lesion with tooth-like calcified areas (Figure 1B,C). Under the clinical diagnosis of odontoma, the tumor was resected from the left side of the maxilla with an intraoral approach under general anesthesia (Figure 2A). The tumor presented morphological findings consistent with an odontoma (Figure 2B). The patient had an uneventful postoperative course, and her 12-month follow-up radiographs showed no evidence of recurrence (Figure 2C,D).

Histologically, immature dentin without dentin tubules and enamel formation were seen around the dentin tissue, seeming to form a complex odontoma (Figure 3A). However, findings in the soft tissue of the tumor were different from those of a usual odontoma. Ghost cells were

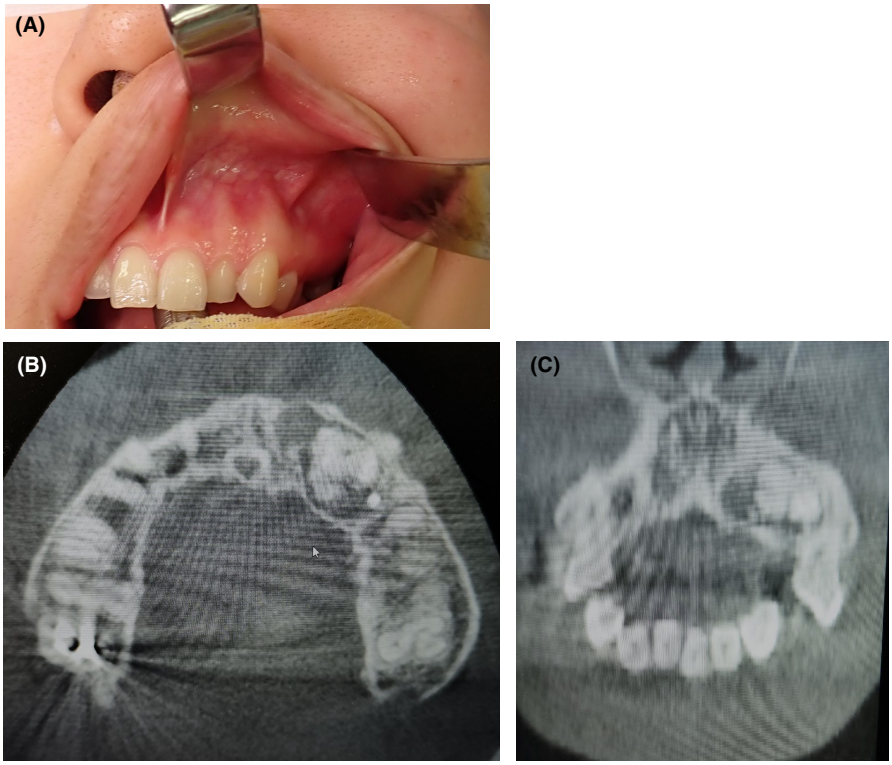

FIGURE 1 Intraoral photograph and X-ray images: (A) Intraoral photograph: A painless gingival mass in the left maxilla region. (B) Conventional CT images (horizontal plane). (C) Conventional CT images (coronal plane).

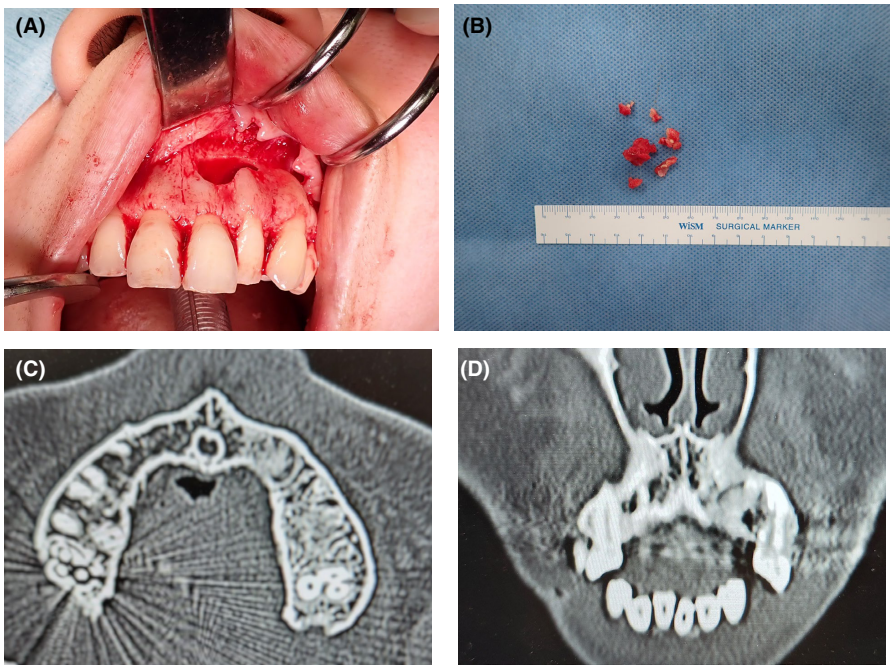

FIGURE 2 (C) Intraoperative photograph and postoperative X-ray image. (A) Intraoperative photograph. (B) Gross appearance of the surgically excised odontogenic tumor specimen. (C) 3-month postoperative conventional CT images (horizontal plane). (D) 3-month postoperative conventional CT images (coronal plane).

also present in the odontogenic epithelium lining the cyst wall (Figure 3B).

Immunohistochemical analysis showed strong cytoplasmic and membranous beta-catenin expression in basaloid cells surrounding ghost cells (Figure 3C). CD138, CK19, and CK14 were diffusely positive in basaloid cells surrounding the ghost cells (Figure 3D–F). CK7 was focally positive, indicating that there were no duct structures (Figure 3G).

Based on these immunohistological studies, this case was considered to a case of coexistent odontoma and DGCT.

3 | DISCUSSION

DGCT is defined as a "locally invasive neoplasm characterized by ameloblastoma-like islands of epithelial cells in a mature connective tissue stroma". DGCT can occur in both the maxilla and mandible, and most cases occur in the second to fifth decade of life, with a slight female predilection.[6] The differential diagnosis of DGCT includes calcifying odontogenic cyst, ameloblastic fibroma, adenomatoid odontogenic tumor, and calcifying epithelial odontogenic tumor.[7]

In these diseases, ghost cells can be found in association with varying amounts of dysplastic dentin.[8] However, the true origin of ghost cells is not fully understood though several theories exist.[4] These hypotheses propose that ghost cells could result from aberrant keratinization, the presence of excessive enamel proteins, or coagulative necrosis.[9,10]

In terms of clinical presentation, odontoma and DGCT may have similar features, including a painless, slow-growing mass in the oral and maxillofacial region. However, radiographic evaluation can help to distinguish

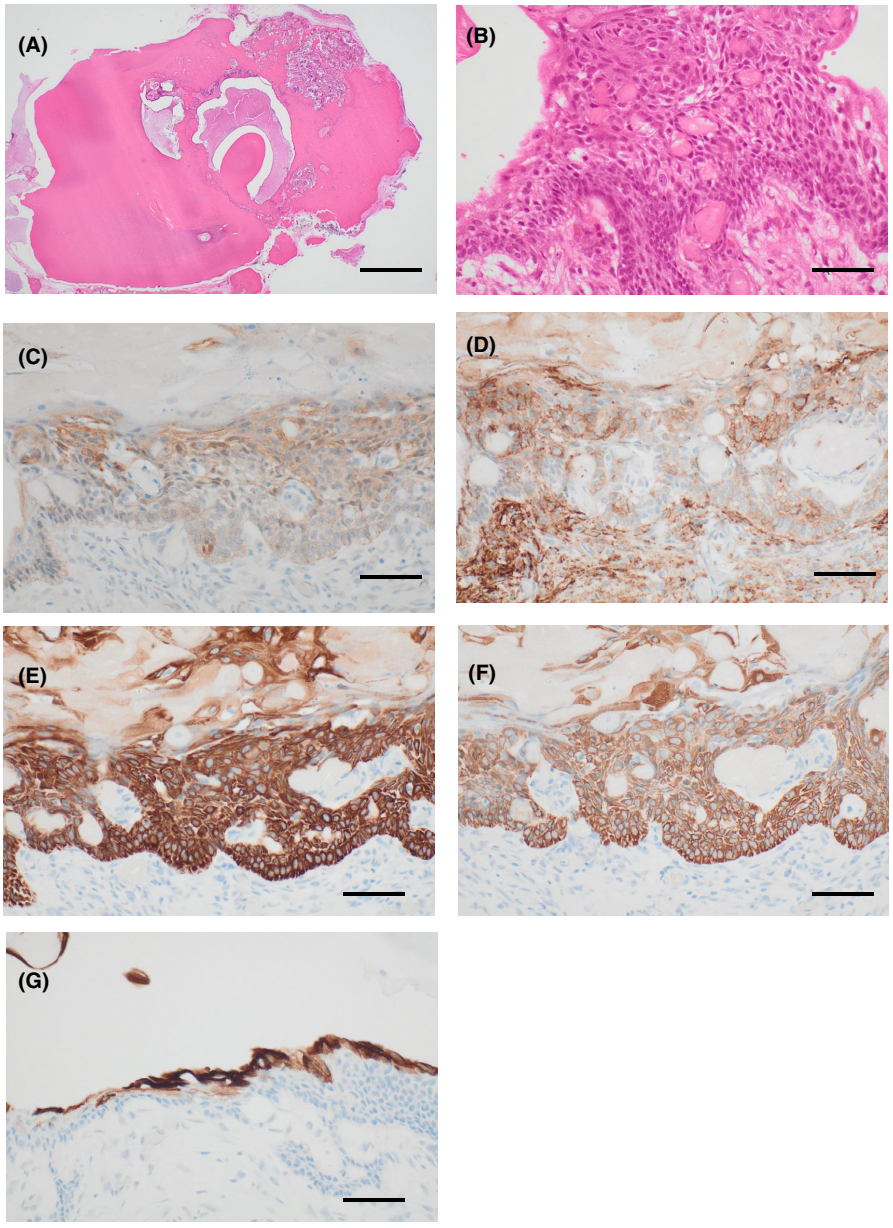

FIGURE 3 Pathological analysis. (A) Photomicrograph of hematoxylin-and-eosin-stained sections showing dentinoid material and tumor (bar 1000 μm). (B) hematoxylin-and-eosin-stained sections showed ghost cells. (bar 40 μm). (C) Immunohistochemical stain (β-catenin) (bar 40 μm). (D) Immunohistochemical stain (CD138) (bar 40 μm). (E) Immunohistochemical stain (CK19) (bar 40 μm). (F) Immunohistochemical stain (CK14) (bar 40 μm). (G) Immunohistochemical stain (CK17) (bar 40 μm).

between the two tumors, as odontoma typically presents as a well-circumscribed radiopaque mass, while DGCT may show a radiolucent component. In our case, odontomas and DGCT coexisted in the same area. This is an extremely rare condition and has only been reported in four other cases besides the present one.[11–14]

Surgical excision is the treatment of choice for DGCT, and the extent of surgical excision required depends on the size and location of the lesion. The recurrence rate of DGCT has been reported to be 10%–20%, highlighting the importance of long-term follow-up.[5] The prognosis of DGCT is generally good, with low potential for malignant transformation.

Odontoma and DGCT also have distinct histopathological features. Odontoma is composed of various dental tissues, including enamel, dentin, cementum, and pulp, forming a disorganized mass.[2] DGCT, on the other hand, is characterized by the presence of ghost cells, which are keratinized cells that have lost their nuclei. DGCT also contains calcified material and a few layers of odontogenic epithelium.[15]

A recent study indicated the molecular signaling affect for development of these odontogenic tumors. Wnt signaling is also known to play a role in the development and differentiation of odontogenic tissues, including the dental epithelium and mesenchyme. Specifically, the Wnt/beta-catenin pathway has been implicated in the formation of enamel, dentin, and cementum, as well as in the proliferation and differentiation of odontogenic stem cells.

Aberrant Wnt signaling has been observed in odontogenic tumors, including ameloblastomas and odontomas. Some studies suggest that aberrant activation of the Wnt/beta-catenin pathway may play a role in the development of dentinogenic ghost cell tumors.[16] Specifically, it has been observed that the CTNNB1 gene coding beta-catenin, a key component of the Wnt/beta-catenin pathway, shows mutational upregulation in dentinogenic ghost cell tumors.[15–17] These results suggest that abnormalities in the Wnt/beta-catenin pathway may have caused the simultaneous occurrence of odontomas and DGCT as in this case. Further research is needed to fully understand the link between the Wnt/beta-catenin pathway and dentinogenic ghost cell tumors.

These genetic studies could indicate that DGCT and odontomas are diseases with different phenotypes due to identical genetic mutations or activation occurring at different times and locations.

4 | CONCLUSION

DGCT is a rare odontogenic tumor that requires a multidisciplinary approach for diagnosis and management. Surgical excision is the treatment of choice for DGCT, and long-term follow-up is necessary due to the potential for recurrence. The prognosis of DGCT is generally good, with low potential for malignant transformation. Further research is needed to elucidate this entity's etiology.

AUTHOR CONTRIBUTIONS

Tatsuo Okui: Conceptualization; data curation; formal analysis; funding acquisition; investigation; methodology; project administration; resources; software; supervision; visualization; writing – original draft; writing – review and editing. **Reon Morioka:** Data curation; investigation; resources; software; supervision; writing – review and editing. **Teruaki Iwahashi:** Data curation; investigation; resources; software; writing – review and editing. **Yuhei Matsuda:** Data curation; formal analysis; resources; supervision; writing – review and editing. **Shinji Ishizuka:** Data curation; investigation; resources; visualization; writing – review and editing. **Satoe Okuma:** Data curation; investigation; resources; supervision; writing – review and editing. **Hiroto Tatsumi:** Data curation; investigation; resources; software; supervision; writing – review and editing. **Takahiro Kanno:** Conceptualization; data curation; investigation; methodology; resources; supervision; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that they have no conflict of interest.

DATA AVAILABILITY STATEMENT

The datasets analyzed during the current study are available from the corresponding author on reasonable request.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

ORCID

Tatsuo Okui https://orcid.org/0000-0002-7640-3274

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