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Dentinogenic ghost cell tumour: A case report

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Abstract

A 23-year-old Malay female patient presented with a history of pain and swelling over right maxilla. Imaging showed a well-defined unilocular radiolucency with areas of radiopacity in the right maxilla. The lesion was initially thought to be a unicystic ameloblastoma. However, histopathology of the excised lesion proved otherwise with a final diagnosis given as dentinogenic ghost cell tumour. The clinical presentation of the case, subtypes of DGCT, similarities with ameloblastoma, and treatment modalities are discussed in this paper.

Keywords: *calcifying odontogenic cyst, dentinogenic ghost cell tumour, Gorlin cyst*

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Introduction

Dentinogenic ghost cell tumour (DGCT) presents as a rare invasive neoplasm characterized by islets of ameloblastoma-like epithelial cells in mature connective tissue. Aberrant keratinization can be found in the form of ghost cells in association with varying amounts of dysplastic dentin (Agrawal, *et al.,* 2017). This tumour makes up for only 2%-14% of all calcifying odontogenic cysts and less than 0.5% of all odontogenic tumours which owes to its rarity (Kumar, et al., 2010). It usually occurs in elderly persons with a male predilection (Pinheiro, *et al.*, 2019). The purpose of this article is to report a case of dentinogenic ghost cell tumour in a 23-year-old female, which is at a comparatively younger age.

Case Presentation

A 23-year-old Malay female was referred to Department of Oral and Maxillofacial Surgery, Segamat Hospital due to swelling over the anterior upper jaw which she noticed in the past one month. The lesion started as a small swelling at the upper sulcus of anterior teeth then gradually increasing in size. She also complains of intermittent throbbing pain over that region upon biting. Patient claims to have no known medical illness with no relevant family history. However, when vital signs were taken prior to biopsy, it was noted that patient had persistent tachycardia. Patient was then referred to Emergency Department and was later diagnosed as hyperthyroidism secondary to Graves' disease. Patient was then started on carbimazole and propranolol by medical team for her condition which has improved her symptoms.

Extraoral examination revealed right nasolabial fold obliteration secondary to swelling of right upper lip and philtrum region. Intraoral examination revealed swelling over right labial sulcus extending

from 11 to 14 region with overlying mucosa appearing bluish, soft in consistency and non-tender on palpation (Figure 1a). Bony expansion was also noted on palatal region extending from 11 to 14 (Figure 1b). Teeth involved were firm, non-displaced, but non vital with electric pulp test. Upon aspiration from right labial sulcus, noted brown coloured fluid within lesion (Figure 1c). An incisional biopsy was performed and based on histopathological examination correlating with clinical and radiographical findings, differential diagnosis of ameloblastoma and calcifying odontogenic cyst were considered.

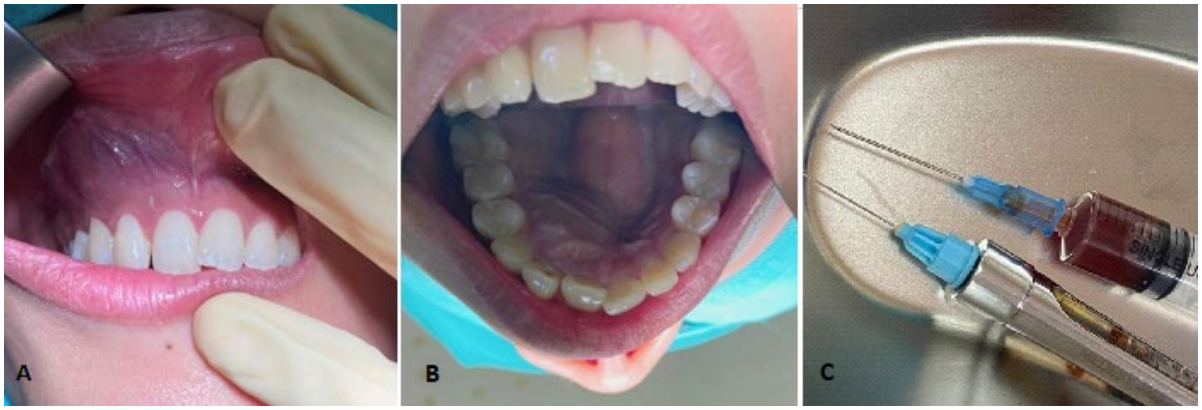

Figure 1. (A) Swelling over right labial sulcus extending from 11 to 14 region with overlying mucosa appearing bluish. (B) Bony expansion palate region extending from 11 to 14. (C) Brown coloured aspiration fluid.

A contrast enhanced computed tomography scan (CECT) showed a unilocular expansile intraosseous lesion measuring 2.5 x 2.8 x 2.7cm at the right paramedian anterior maxillary region with no significant enhancement. There are specks of hyper density within the lesion, suggestive of calcifications (Figure 2). There is associated root resorption of adjacent right upper incisors up to the first premolar. The lesion has caused abutment of anterior wall of maxillary sinus and right nasal cavity without invasion.

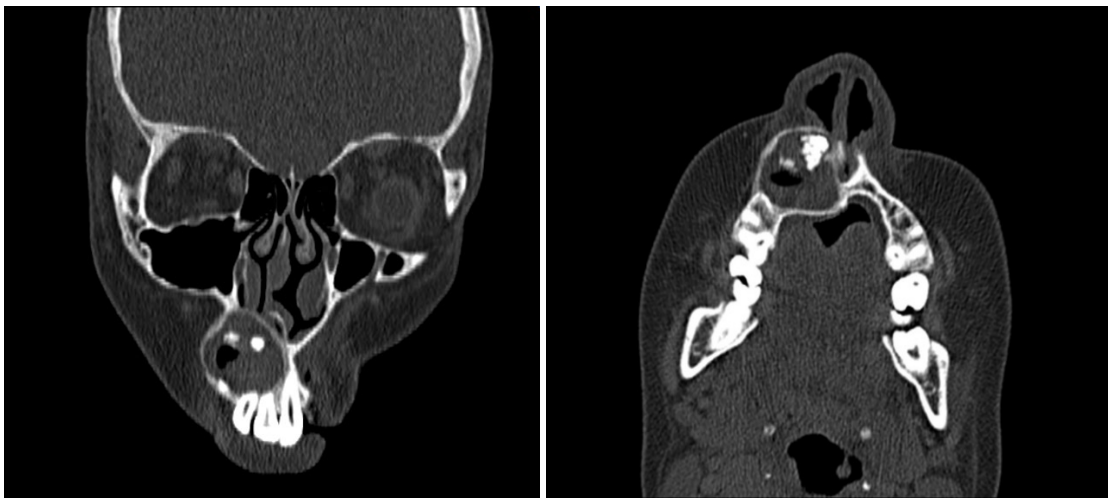

Figure 2. Contrast enhanced computed tomography showing unilocular expansile lesion with specks of calcification within (coronal and axial view).

Enucleation of lesion (Figure 3) with ostectomy of approximately 1-2mm at the periphery margins of the lesion was done under general anaesthesia and the tissue was submitted for histopathological examination.

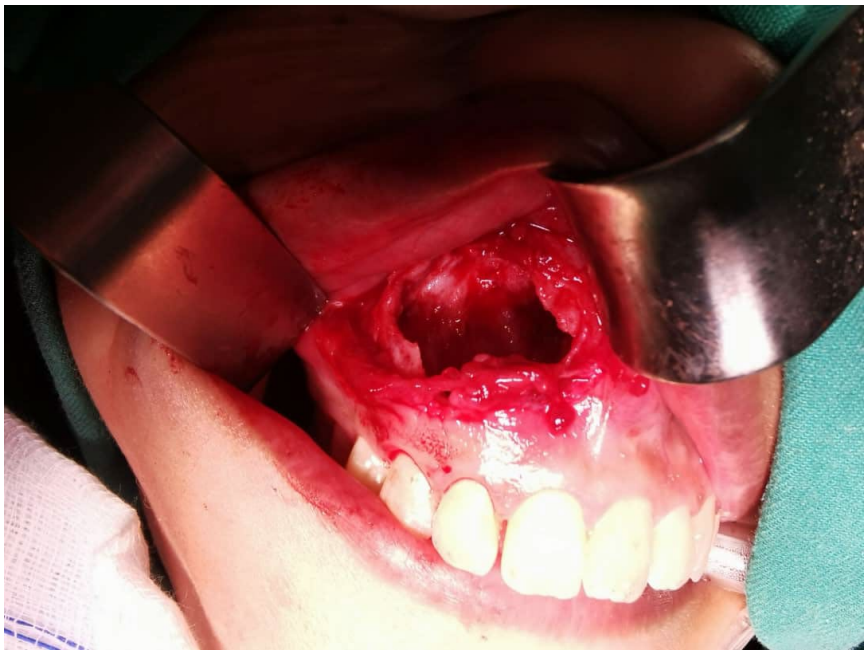

Figure 3. Enucleation of lesion with ostectomy under general anaesthesia.

Histopathological examination showed an odontogenic tumour lined by epithelium of varying thickness composed of loosely cohesive epithelium with palisading of basal cell layer with reverse polarity and areas having accumulation of ghost cell admixed with scattered calcifications (Figure 4a,4b). An ameloblastomatous island consisting of ghost cells and calcifications undergoing cystic degeneration and several small ameloblastomatous islands are seen in the fibrous tissue (Figure 4d).

Present within the fibrous tissue are dentinoid material (Figure 4c) adjacent to the lining and at the deeper aspects, the dentinoid material is seen associated with basaloid cells. Presence of dentinoid materials are also noted between sheets of cholesterol clefts with associated multinucleated giant cells.

Upon one year of follow-up examination, patient presented with well recontoured bone of right maxilla regaining normal morphology (Figure 5A, 5B) and did not show any signs of recurrence. Teeth involved with lesion, tooth 11 to 14, did not show any signs or symptoms clinically. Radiographic examination showed new bone formation filling the previous enucleated cavity while teeth involved with lesion previously did not show further resorption of root tips (Figure 5C). Electric pulp testing was done on involved teeth and noted tooth 13 and 14 regained vitality while tooth 11 and 12 remains non vital. All non-vital teeth were subjected for root canal treatment and apicoectomy with root end filling.

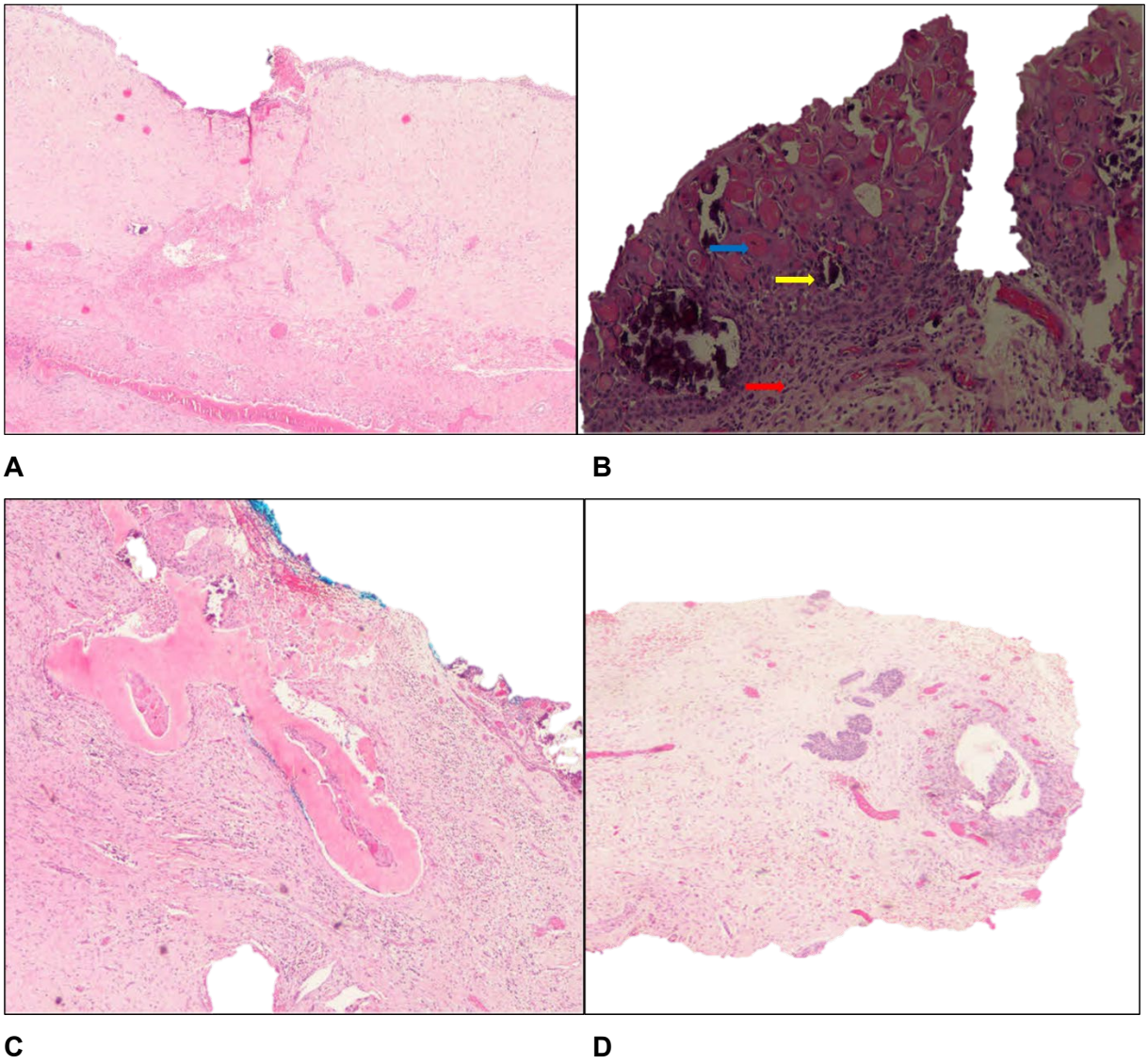

Figure 4. (A) Fibrous wall containing bland odontogenic rests lined by loosely cohesive epithelium with focal areas having palisading of basal cell layer with reverse polarity. Parts of the cells near the luminal areas resemble stellate-reticulum like-cells. Presence of dystrophic calcifications and trabeculae of vital woven bone rimmed by osteoblasts is noted. [Hematoxylin and Eosin stain, ×4], (B) Odontogenic tumour lined by epithelium of varying thickness composed of loosely cohesive epithelium with palisading of basal cell layer (red arrow) with reverse polarity. Areas having accumulation of ghost cells (yellow arrow) admixed with scattered calcifications (blue arrow). [Hematoxylin and Eosin stain, ×10], (C) Dentinoid materials [Hematoxylin and Eosin stain, ×10], and (D) Ameloblastomatous islands [Hematoxylin and Eosin stain, ×10].

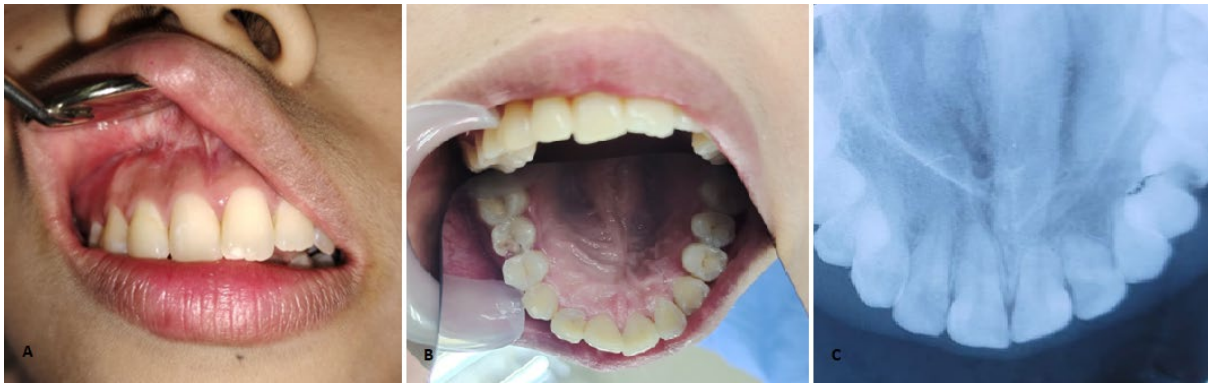

Figure 5. Progress of patient one-year post enucleation. (A) Slight scarring of upper buccal sulcus with no palpable bulge (B) Well recontoured palate, and (C) Upper occlusal radiograph showing new bone formation in the previous enucleated cyst cavity and resorption of root tips teeth 11 to 14 persist.

Discussion

Calcifying odontogenic cysts were first described by Gorlin and colleagues in 1962 as a separate entity of odontogenic origin. Calcifying odontogenic cysts account for 1–2% of all odontogenic tumours, in which 88.5% are cystic and the remaining 11.5% are solid tumours (Agrawal *et al*., 2017; (Singhaniya *et al.*, 2009). As all lesions are not cystic, it is debatable, whether calcifying odontogenic cyst is a cyst or a neoplasm (Singhaniya *et al*., 2009). Some have suggested that there could be a possibility of cystic degeneration taking place at the centre of proliferating epithelial islands rather than epithelial changes developing in a pre-existing cyst wall (Patankar *et al.*, 2019). Based on this dualistic concept, WHO termed all cystic lesions as calcifying cystic odontogenic tumours (CCOT) and the neoplastic entity as dentinogenic ghost cell tumours (DGCT) (Agrawal *et al.*, 2017). 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 in association with varying amounts of dysplastic dentin." (Bafna *et al.*, 2016; Garcia *et al.*, 2015; Patankar *et al.*, 2019). The aetiology of this lesion is still unknown, but it has been suggested that missense mutation in β-catenin during odontogenesis disrupt the proper differentiation process coordinated in wingless integrated (Wnt) pathway, plays a crucial role in the formation of DGCT (Kim *et al.*, 2007).

DGCT may occur as an intraosseous central lesion (68%) and less commonly as an extraosseous peripheral lesion arising in the gingiva or alveolar mucosa (32%). The age may range from 7 to 82 years (mean 45 years) with strong male predilection. A majority of published DGCT cases were reported in the Asian population (65%) (Pinheiro *et al.*, 2019). Both the central and peripheral variant showed a greater predisposition to the mandible than the maxilla (Pinheiro *et al*., 2019). The tendency to occur at the canine to first molar region of the jaw (Agrawal *et al.*, 2017; Patankar *et al.,* 2019; Kumar *et al*., 2010). The behaviour of intraosseous DGCT is more aggressive than extraosseous DGCT (Kelleş *et al.*, 2012).

In this case, the lesion presented with both cystic and tumour characteristics. DGCT may appear radiographically as radiolucent, radiopaque, or mixed lesion amounting to the presence and extent of calcification. The radiopacity seen in the CECT taken was initially thought to be an impacted odontome or supernumerary tooth associated with the lesion. Lesions may appear unilocular or multilocular with either well-defined or ill-defined margins (Patankar *et al.*, 2019). Due to the mixed presentation of this lesion, the initial provisional diagnosis of the lesion was

thought to be a cystic ameloblastoma as it showed clinical features such as resorption of adjacent tooth, association of impacted tooth, buccal and lingual cortical expansion with disruption of the buccal cortex those of which are found in cystic ameloblastoma (Cadavid *et al.*, 2019; Patankar *et al.*, 2019). Ameloblastoma represents approximately 11 to 18% of all odontogenic tumours, being the second most common after odontomas (Cadavid *et al.*, 2019), a much higher prevalence compared to DCGT. The characteristic features of DGCT that distinguish it from ameloblastoma and other odontogenic tumours are presence of numerous ghost cells and masses of dentinoid material (Singhaniya *et al.,* 2009; Martos-Fernández, *et al.*, 2014).

Among case reports reviewed, little has been mentioned regarding aspirated fluid from the lesion. *Agrawal et al* reported thin yellowish colour, blood-tinged aspiration fluid and on cytological examination only red blood corpuscles (RBC) were found (Agrawal *et al.*, 2017) while Kelles *et al.* reported aspiration fluid showed turbid brown fluid and on cytological examination revealed groups of degenerating cells with prominent cytoplasm with foamy histiocytes (Kelleş *et al.*, 2012). There is no conclusive evidence if the aspiration fluid of the cyst provides any diagnostic value.

Central DGCT is considered as locally aggressive neoplasm. Study showed a recurrence rate of 73% after conservative surgical treatment of enucleation or curettage compared with a recurrence rate of 33% after radical treatment of peripheral or segmental resection (Buchner *et al.*, 2016) where recurrence can occur up to 20 years after the initial surgery (Pinheiro *et al.,* 2019). Another author also finds similar finding with recurrence rate of DGCT at 71% and the recurrence tends to occur between 5 to 8 years post initial treatment (Alzaid *et al.*, 2022). Compared to the less aggressive extraosseous counterpart where no recurrence has been reported after conservative treatment (Pinheiro *et al.*, 2019). Although rare, it has been reported that recurrent DCGT have shown malignant potential and is diagnosed as ghost cell odontogenic carcinoma (GCOC) (Martos-Fernández *et al*., 2014; Pinheiro *et al.*, 2019).

Some has proposed that DGCT should be treated as ameloblatomas as there are several histological features like ameloblatomas (Garcia *et al*., 2015). It has been proposed that DGCT should be treated by resection with safety margin of at least 0.5cm, similar to recommendations for ameloblastoma (Buchner *et al.*, 2016; Garcia *et al.*, 2015; Pinheiro *et al.*, 2019). Some authors suggested initial conservative treatment of enucleation and meticulous curettage of the surrounding bony wall around 1 to 3mm for radiographic unilocular well-defined lesions. The initial radical treatment of peripheral or segmental resection is preserved for clinical and radiographic destructive lesions with ill-defined borders (Buchner *et al.,* 2016). Aggressive wide local excision such as en-block resection is proposed to be carried out in intraosseous subtype of DGCT (Patankar *et al.*, 2019) due to its high recurrence rate. However, it is important to note that some cases were treated with surgical enucleation without any recurrence (Pinheiro *et al.*, 2019).

For this patient, considering her young age, gender, and small size and location of the tumour, a more conservative approach was adopted. Radiographically, the lesion involves all right upper anterior teeth, with expansion into palatal region, abutting on right nasal floor and right anterior maxillary sinus. If a wider excision such as en-block excision was done, this patient will end up with large defect which required more complex rehabilitation process. Her appearance, speech and masticatory forces will be greatly impaired and leads to negative outcome on her quality of life. Based on the clinical and radiographical aspects, we decided on a much conservative treatment approach which is surgical enucleation with ostectomy of periphery margin up to 2mm. This treatment approach has been decided in the view of few authors who also practice more conservative approach in treating DGCT. The same approach also taken by other author who reported the enucleation of DGCT was

carried out in a view of small tumour size with no local recurrence (Agrawal *et al.*, 2017).

Due to high risk of recurrence, this patient has been scheduled for a frequent long term follow up with imaging. Imaging is scheduled at least once a year to properly monitor for any recurrence of the lesion as the DGCT has malignant transformation for recurrent cases (Alzaid *et al.*, 2022). One year of follow-up examination on the patient did not show any signs of recurrence. The treatment of choice remains a dilemma for surgeons. However, it is consensus that all patients with DGCT should remain in long-term follow-up due to its high rate of recurrence and possible malignant transformation.

Conclusions

The mixed histopathology characteristic of dentinogenic ghost cell tumour leads to surgical dilemma on the best approach of the treatment. Therefore, a long term follow up is necessary to prevent the recurrence of the disease. The best approach of treatment should be tailored according to individual patient factoring clinical, radiographical, HPE and social impact to prevent unnecessary excision. Quality of life and rehabilitation of the patient should be considered in treatment planning.

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