Case Report

An Expansile Well–defined Radiolucency in Anterior Mandible: Case Presentation with Treatment Approach and Literature Update

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Abstract

A 15–year–old Indian male patient presented with a history of pain and swelling in left mandible. Imaging studies revealed a well–defined unilocular radiolucency in the body of the mandible. Patient also gave the past history of the surgery of the jaws, which was histopathologically diagnosed as dentigerous cyst. Following this patient underwent incisional biopsy and later excisional biopsy. The histopathologic diagnosis for incisional biopsy was unicystic ameloblastoma but final diagnosis was dentinogenic ghost cell tumor for the excised tissue. To the best of our knowledge, this appears to be the first case of dentigerous cyst transforming into dentinogenic ghost cell tumor. The clinical presentation of the case, differential diagnosis and treatment modalities are being discussed.

Keywords: dentigerous cyst, dentinogenic ghost cell tumour, mandible, unicystic amelobalstoma

Case presentation

A 15–year old Indian male patient presented with a history of pain and swelling in lower left region of jaw since 6 months at Maulana Azad Institute of Dental Sciences, New Delhi, India. Swelling was initially small in size which had gradually increased. Pain was insidious and intermittent in nature. The patient was in apparently good health with his medical history being non–contributory. Past dental history revealed patient had undergone surgery in the same region of the jaw four years back which was histopathologically diagnosed as dentigerous cyst (DC), previous radiographs were not available. Marsupialization had been carried out with respect to the impacted canine previously so that it can erupt, following which the patient did not return for follow up, and then he reported back after 4 years with this swelling

On examination, left facial swelling was noted in the region of mandible. Left submandibular lymph node was enlarged, palpable and tender. Intraoral examination revealed swelling of 3.5x 1.5 cm in size extending from distal aspect of mandibular right central incisor to mandibular left second premolar. Lateral incisor and canine were missing on the left side of the mandible. There was displacement of mandibular right canine, right lateral incisor, right central incisor and left central incisor of the mandible. Palpation revealed firm to fluctuant swelling with well–defined borders. Marked buccal expansion was also noted.

Panoramic view revealed a well–defined unilocular radiolucency (RL) with sclerotic border in the mandible extending from medial aspect of root of right first premolar to root apex of left first premolar. Displacement of right canine, lateral incisor and central incisor with impacted left lateral incisor and canine were also observed (Fig.1). CECT revealed a large well circumscribed cystic lesion involving the body of the mandible in midline and left side with marked cortical expansion and thinning, along with unerupted impacted teeth in the inferior aspect. No perforation was seen.

Consequently, incisional biopsy was performed and histopathological section revealed a cystic lumen lined by characteristic 4–8 layer of cells with basal cells

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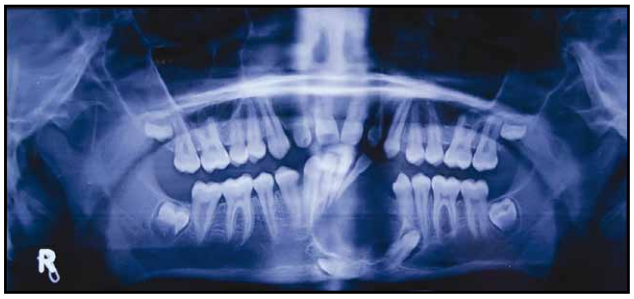
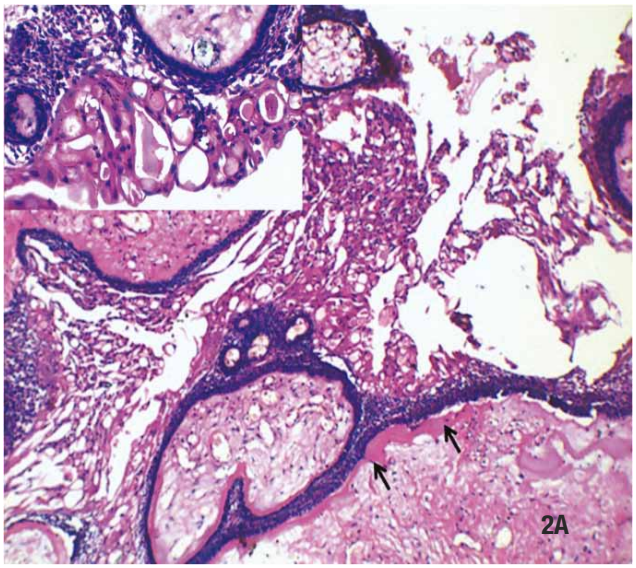


Figure 1. OPG showing well defined unilocular radiolucency with sclerotic border in the mandible extending from medial aspect of root of right first premolar to root apex of left first premolar

having tall columnar appearance showing reversal of polarity and subnuclear vacuolization. Juxta–epithelial hyalinization was seen at many areas. The epithelial lining showed intraluminal proliferation as well as few islands of ameloblastic follicles in the capsule. Evidence of aberrant keratinization in the form of ghost cells was evident at certain areas in the lumen. Few ameloblastic follicles also showed squamous metaplasia. Juxta–epithelial hyalinization with probable dentinoid formation was evident in focal areas. The capsule was moderately fibrocellular in nature. So it was diagnosed with a histopathological differential diagnosis of (1) UA type 1.2.3 (luminal+ intraluminal + mural) and (2) DGCT.

Considering the young age of patient and the nature of the lesion, conservative approach was followed. The operation was performed under general anesthesia intra–orally through degloving incision. Complete extirpation of the tumor mass along with removal of impacted lower left lateral incisor and canine teeth was done. In addition, the involved mandibular teeth– right central incisor, right lateral incisor, right canine, left central incisor and deciduous canine were also removed. Considering the aggressive behavior of both tumors (UA/DGCT), peripheral ostectomy followed by the application of Carnoy's solution (absolute alcohol 6 ml, chloroform 3 ml, glacial acetic acid 1 ml, ferric chloride 1 mg) was done. Around 5 mm of the surrounding bone was shaved off and Carnoy's solution was applied twice initially 5 minutes followed by 3 minutes. The reconstruction plate was fixed across the defect strengthening the inferior border of mandible as due to the lesion less than 1 cm of normal bone was remaining. The bony cavity was thoroughly irrigated with betadine solution before closing with 3–0 silk interrupted sutures.

The biopsy specimen was sent for histopathological examination. Microscopic tissue examination revealed cystic lining composed of tall columnar basal cells with hyperchromatic polarized nuclei and superficial stellate reticulum like cells. Numerous ghost cells having indistinct cytoplasmic borders were noted in groups extending into the lumen. (Fig. 2A and 2B) A zone of atubular dentinoid was present subepithelially at many areas. (Fig. 3A) The dentinoid nature was confirmed by Van–Gieson stain. (Fig. 3B) Follicles of odontogenic epithelium lined by tall columnar preameloblast like cells and containing stellate



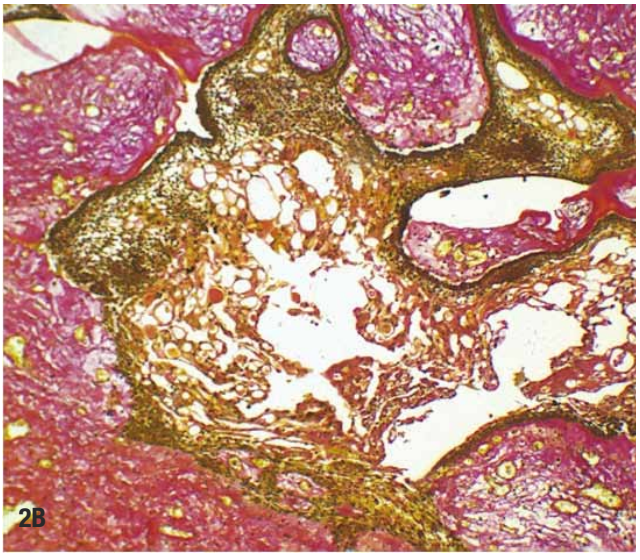


Figure 2A. Microphotograph showing a cystic lumen lined by dark staining tall columnar cells and superficially by loosely arranged cells. Juxta–epithelial hyalinization is seen at many areas shown by black arrows. (H&E X 100)– Inset shows area of aberrant keratinization within the cystic lining. (H&E X 400);

Figure 2B. Van Gieson stained microphotograph showing a characteristic cystic lining composed of tall columnar basal cells and superficial stellate reticulum like cells. Abundant ghost cells having indistinct cytoplasmic borders are prominent in groups extending into the lumen. (Van Gieson X 100)

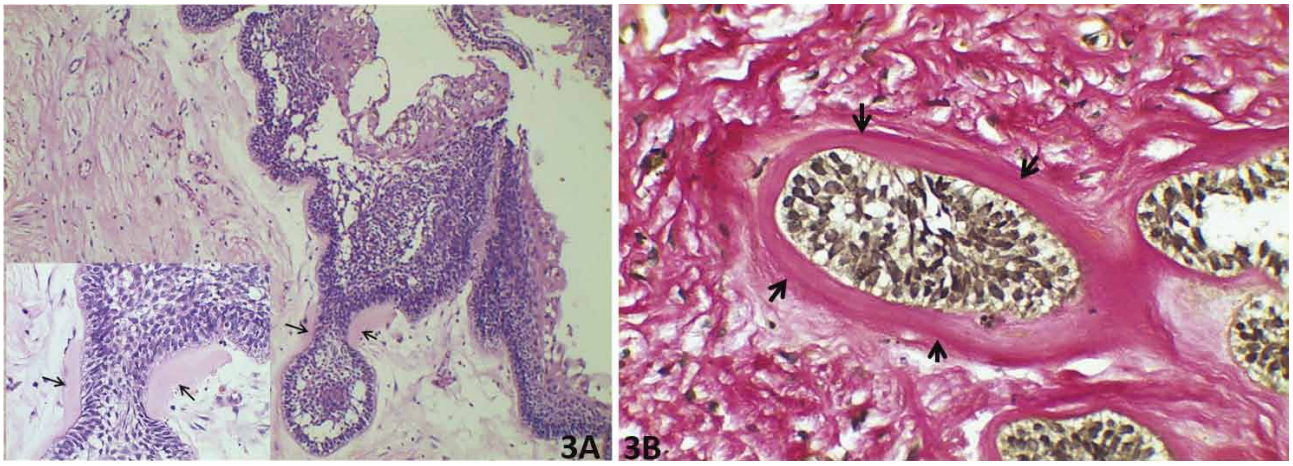


Figure 3A. Microphotograph showing zone of atubular dentinoid subepithelially (H&E X 100), Inset (H&E X 400).

Figure 3B. Van Gieson stained microphotograph showing zone of dentinoid around odontogenic follicles. (Van Gieson X 400)

reticulum like cells were observed at many places in the stroma. A zone of hyalinization was present around some follicles. The supporting stromal tissue was densely collagenous in most part with few fibroblasts and blood vessels. A few quiescent appearing rests of odontogenic epithelium were seen at some areas. These features confirmed the final diagnosis as DGCT.

The post–operative healing was uneventful. As the recurrence and local invasion is characteristic of this lesion, but not distant metastasis. Patient was followed up clinically and radiographically for two years and is still under observation with no recurrence. (Fig. 4) Informed consent was obtained from the patient for the publication of this case.

Discussion

Praetorius et al., (1981) concluded that COC actually comprised of two entities: a cyst and a solid neoplasm (calcifying ghost cell odontogenic tumor, CGCOT). [1–3] The retitling of CGCOT to DGCT was done in 2005 WHO (Praetorius and Ledesma–Montes, 2005).[1]

The PubMed (Medline) electronic database was searched and it disclosed 31 cases of intraosseous DGCT from 1972 to 2008 (Juneja and George, 2009), 15 cases from 2009 to 2013 (Konstantakis et al., 2013) and 5 cases in a recent review of 37 ghost cell odontogenic tumors, a 44 year analysis in Iranian population (Etemad–Moghadam et al., 2014).[5–7] This indicates the rare occurrence of this entity.

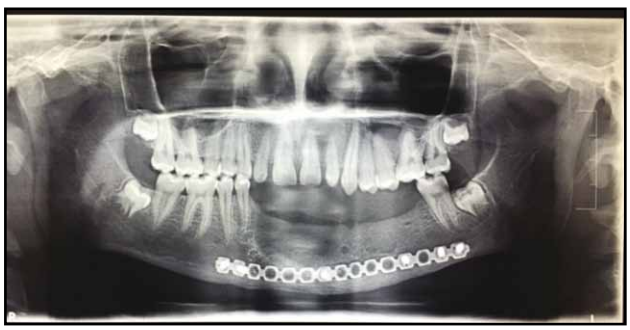


Figure 4. Two year follow up panoramic view showing good bone formation along with reconstruction plate.

Juneja and George reported that DGCT occurs in age range of 12– 75 years with mean age of 40 years. [5] While Konstantakis et al., mentioned age ranging from 12–51 years with a mean age of 29 years. [6] The current case was of 15–year–old male patient. DGCT occurs more commonly in male.

Intraosseous DGCT more commonly occurs in the anterior mandible and this is in accordance with the published review in the literature. [5–7] The clinical findings of intraosseous DGCT may include expansion of jaws, clinically visible swelling, swelling can be painful to painless and tooth mobility. [5–7] The present case was also in the mandible and showed expansion of jaws with clinically visible facial swelling and displacement of teeth. However, DGCT can present itself as asymptomatic case and incidental radiographic finding during routine examination. [8] The radiographic features in the present case was in consistent with scientific literature. This includes well defined unilocular RL to mixed RL/RO appearance depending on the amount of calcification. [5–7]

Due to paucity of literature regarding the long term follow up of patient following the treatment, the ideal treatment approach has not been conclusively determined. Controversy remains between conservative and radical approach. For example, nomenclature carrying a phrase “cystic” is generally approached relatively less vigorously (enucleation or marsupialization), than nomenclature carrying a phrase “tumor”, which are treated more

aggressively (en bloc resection) and followed–up precautiously for longer period. [9,10] Cogitating the patient age factor, the incisional biopsy report of UA or DGCT and Toida's judgment the lesion was thoroughly curetted and treated with Carnoy's solution. Carnoy's solution (chloroform 3 mL, absolute alcohol6 mL, glacial acetic acid 1 mL, ferric chloride 1gm) was initially used as a sclerosing agent for treatment of cysts and fistulae, and is currently used as a fixative. [11] The use of Carnoy's solution in treatment of odontogenic keratocysts, and central giant cell granuloma has already been studied, and was shown in an animal model to penetrate cancellous bone to a depth of 1.5mm. [12]

The use of Carnoy's solution to decrease chances of recurrence after conservative surgical treatment of UA was initially suggested by Stoelinga and Bronkhorst in 1987, and then by Lee et al., reported success rates with recurrence rates of 10% by using Carnoy's solution as an adjunct to enucleation and curettage, even with a high 93% of the lesions being of the mural type.[13,14] They contended that it is likely that the use of Carnoy's solution does contribute towards a favorable result although a few limitations in the study like a short follow up period do not unequivocally prove this notion. Scientific literature suggests that close long–term clinical and radiographic follow–up evaluation is mandatory for 15 to 20 years in all cases of DGCT. [15]

By noting clinical and radiographic findings i.e. a well–defined lesion of long duration in anterior mandible exhibiting a well–defined unilocular RL with impacted teeth, a broad array of differential diagnosis was made. This included cystic lesions and benign odontogenic tumors like DC, COC, cystic adenomatoid odontogenic tumor (AOT), UA and ameloblastic fibroma (AF). Since the present case was a well–defined radiolucent lesion on radiograph, therefore odontogenic malignancies like ameloblastic fibrosarcoma, ameloblastic fibro–odonto/ dentino sarcoma and non–odontogenic malignancies like osteosarcoma, chondrosarcoma were excluded.

Microscopic features of excisional biopsy of the current case were consistent with the diagnosis of DGCT. The presence of ghost cells within the ameloblastomatous lining is the essential prerequisite for the diagnosis. But however at times it can be confused with ameloblastic fibro–odontoma but absence of enamel matrix helped in DGCT diagnosis. Moreover, ghost cells were also observed in other lesions such as odontoma and ameloblastoma. [4] The presence of large amount of ghost cells and dysplastic dentin observed in the present case differentiated DGCT from ameloblastoma histologically. Considering the previous dental history, this appears to be the first case of DC transforming into DGCT, to the best of our knowledge.

In conclusion, we have presented an interesting case of transformation of DC post marsupialization into a DGCT showing characteristic histopathological features in a very young male individual. Thorough examination of excisional tissue confirmed the incisional biopsy diagnosis. Odontogenic tumors show a lot of diversity in their histomorphological presentation, depending on the stage of initiation of the tumor.

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