

Clinical report

Intraosseous dentinogenic ghost cell tumor: Case report and treatment review

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ARTICLE INFO

Article history:

Received 11 March 2013

Accepted 26 March 2014

Available online 20 November 2014

Keywords:

Dentinogenic ghost cell tumors

Calcifying odontogenic cyst

Aggressive local resections

Palabras clave:

Tumor dentinogénico de células fantasma

Quiste odontogénico calcificante

Resección local amplia

ABSTRACT

Dentinogenic ghost cell tumors (DGCT) are uncommon neoplasms classified as solid variants of calcifying odontogenic cyst and are defined as a locally invasive neoplasm, characterized by ameloblastoma-like islands of aberrant keratinization of odontogenic epithelium in the form of ghost cells in association with dysplastic dentin. We present the case of a 46-year-old woman who was referred to us due to dental mobility and swelling of the jaw. The different imaging and histological studies confirmed the diagnosis of a dentinogenic ghost cell tumor. Treatment was based on aggressive local resection with adequate safety margins, and monitoring the patient for detection of recurrences. The purpose of this paper was to describe a case of DGCT and the treatment adopted in our case, and to provide a review of the treatment of the cases reported in the indexed literature.

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Tumor dentinogénico de células fantasmas tipo central (intraóseo): un caso clínico y revisión del tratamiento

RESUMEN

El tumor dentinogénico de células fantasma (TDCF) es una rara neoplasia clasificada como una variante sólida del quiste odontogénico calcificante, definida como una neoplasia localmente invasiva caracterizada por la presencia de islas ameloblastomatosas con queratinización aberrante en forma de células fantasma, coexistiendo con displasia dentinaria. Presentamos un caso clínico de una paciente de 46 años que nos consultó por presentar movilidad dental con inflamación a nivel mandibular. Los estudios radiológico e histológico revelaron el diagnóstico de tumor dentinogénico de células fantasma. Se realizó resección local amplia con márgenes, revisando periódicamente a la paciente para detectar recidivas. El objetivo de este artículo es presentar un caso clínico de un TDCF, describir el tratamiento realizado en nuestro caso y revisar el tratamiento realizado en los casos publicados en la literatura indexada.

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<http://dx.doi.org/10.1016/j.maxilo.2014.03.007>

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Introduction

Calcifying odontogenic cyst (COC) was first described as a distinct pathology by Gorlin et al. in 1962.¹ However, in 1971, COC was officially defined by the World Health Organization (WHO) as a “nonneoplastic cystic lesion in which the epithelial lining shows a well-defined basal layer of columnar cells, an overlaid layer that is often many cells thick that may resemble stellate reticulum and masses of ghost epithelial cells that may be in the epithelial cyst lining or in the fibrous capsule. Dysplastic dentin may be laid down next to the basal cell layer of the epithelium”.² Over the past years, many classifications have tried to set the different histological and clinical features of this group of tumors. However this new classification supposed a self-contradiction since the lesion was again classified as “benign odontogenic tumor”.² Buchner resolved this classification issue by splitting these lesion categories into two: cysts and neoplasms. Buchner also found that only 2–14% of COCs were solid tumors.

These solid tumors were considered as dentinogenic ghost cell tumors (DGCTs).^{3,4}

The current WHO classification of 2005 recognizes DGCT as the early solid variant of COC (type II).

The objective of this article was to present a DGCT case study along with a review of published cases of central DGCT in order to discuss the appropriate treatment to be provided for this kind of lesion in the maxillary bones.

Case report

A 46-year-old woman was referred to the Department of Oral and Maxillofacial surgery due to painless swelling and dental mobility on the anterior dental sector of the jaw for the previous 3 months. Clinical examination disclosed grade III mobility of teeth [42–35] with rhizolysis of teeth 41, 31 and 32, and painless swelling from the right lateral incisive of the jaw to the left second molar area of the jaw. The patient referred a solid mass in the area described. The oral mucosa also displayed an irregular surface and a protuberant mass on the left mandible.

A mixed radiolucent–radiopaque well-defined unilocular lesion was found on an orthopantomographic examination, affecting roots 42–35 (Fig. 1).

A histological examination (after incisional biopsy) revealed islands of ameloblastoma-like odontogenic epithelial proliferation, eosinophilic ghost cells and dentinoid-like material.

A computed tomography scan (CT) for preoperative evaluation showed a 2.6×1.3 expansive lesion with a combined pattern of a well-circumscribed unilocular radiolucency with radiopaque images that corresponded with calcification. Thinned cortical expansive lesion was observed on the lingual edge, and no septums were found (Fig. 2).

The patient was treated using extensive curettage with an aggressive local resection consisting of marginal mandibulectomy surrounding the lesion (Fig. 3) with safety margins without loss of continuity of the jaw.

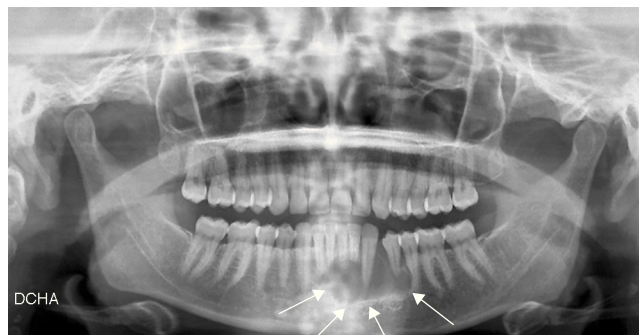


Fig. 1 – Mixed radiolucent–radiopaque unilocular lesion, encompassing the roots from 42 to 35.

Histopathological examination of the surgical specimen revealed a solid tumor composed of sheets and rounded islands of odontogenic epithelium that resembled ameloblastoma-like cells, transformation of the epithelial cells into ghost cells in the fibrous connective tissue similar to foreign body reaction. Ghost cells were seen trapped in the dysplastic dentin and mitosis could not be observed (Fig. 4).

Currently, two years later, there is no evidence of re-sprouting or malignant transformation (Fig. 5).

Discussion

In 2005 the WHO classification defined specific types of cystic lesions and solid tumor mass as well as their development. DGCT was then defined as a locally invasive neoplasm characterized microscopically by ameloblastoma-like odontogenic epithelial proliferation, an aberrant keratinization in the form of ghost cells and dysplastic dentin.^{5,6} Two variants were



Fig. 2 – CT showed a expansive lesion with lingual cortical thinning.



Fig. 3 – Aggressive local resection: marginal mandibulectomy without loss of the continuity of the jaw.

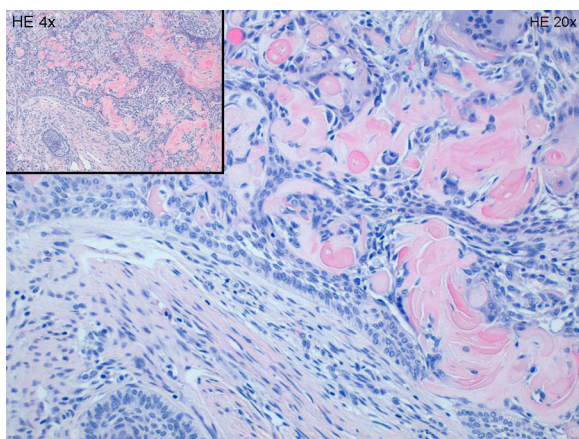


Fig. 4 – Transformation of the epithelial cells into ghost cells in the fibrous connective tissue.



Fig. 5 – Panoramic radiography did not show recurrence two years later.

identified: aggressive central and non-aggressive periphery located tumors.⁷

A search of indexed literature found 41 cases of central DGCT (excluding ours)⁸⁻¹¹ with a wide range of patient age distribution (from 12 to 75 years). Among these 41 cases, gender and location data were specified in only 27 cases: 19 cases (70.4%) were males and 8 (29.6%) females. There were 15 (55.6%) cases of DGCT in the mandible and 12 (44.4%) in the maxilla.

The treatment of each case described was different. The early solid variant of COC (type II) seemed to have a more aggressive development than cystic lesions; this could explain the common recurrent local sprouting after more traditional treatment protocols.

Gary L. Ellis⁶ affirms that several histological features of DGCT are similar to ameloblastoma; therefore, such lesions should be treated as ameloblastoma. Kasahara et al.¹² proposed similar recommendations: among 11 patients reviewed, locally recurrent DGCT occurred in 4 of them. So they recommended treating DGCT as ameloblastoma and suggested that conservative treatment protocols (curettage or/and enucleation) were not suitable options for the treatment of DGCT. In addition, recently, Sun G. et al.⁸ reviewed 7 patients with DGCT of whom 5 had been treated using only curettage and all of them had recurrences. The other 2 patients treated using local resection had not shown any recurrence so far. Thus, the authors concluded “that intraosseous DGCT should be treated with resection with an adequate safety margin, at least 0.5 cm, similar to recommendations for ameloblastoma”.

Our team proposes an appropriate local wide resection for these lesions. In our case the patient was treated using curettage and additional marginal osteotomy but preserving the mandibular basal bone. After two years there is no evidence of re-sprouting. The patient did not want to be rehabilitated with dental implants, settling for a removable dental bridge. We had in mind Kasahara et al.’s treatment recommendations in order to avoid any recurrences due to enucleation: they found that the recurrence rate was 36% (4 of the 11 DGCTs treated¹²); therefore, the prognosis of DGCT is determined by the treatment provided.¹³ Although non-concrete DGCT treatment has been proposed, we consider there is enough evidence to suppose that the appropriate treatment should be an aggressive local resection with or without safety margin, and the patient should remain in long-term follow-up since a recurrent malignant neoplasm from a previously diagnosed DGCT has been demonstrated.¹⁴ In the event of a recurrence, retreatment with wide local resection should be considered.

Conclusion

In our case study the patient was treated using local resection and curettage and two years later, there was no evidence of recurrence.

It should be noted that there is no consensus regarding the best treatment option in cases of DGCT. The 41 cases reported to date were treated with different surgical techniques with inconclusive long-term results. Irrespective of treatment options, patient monitoring should be inexcusably undertaken in order to detect recurrences or re-sprouting.

We hope that additional studies in future will contribute to determining the best treatment options for DGCT in this line, as well as a better explanation of the precise histopathological, biological and clinical development of DGCT and to definitively determine whether aggressive resection is the best treatment for DGCT.

Ethical disclosures

Protection of human and animal subjects. The authors declare that procedures conformed to the ethical standards of the responsible committee on human experimentation and in accordance with the World Medical Association and the Declaration of Helsinki

Confidentiality of data. The authors declare that they have followed the protocols of the workplace on the publication of data from patients and all patients included in the study have received sufficient information and gave written informed consent to participate in the study consented.

Right to privacy and informed consent. The authors have obtained informed patients and/or subjects referred to in article consent. This document is in the possession of the author of correspondence.

Funding

None.

Competing interests

None declared.

Ethical approval

Not required.

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