CASE REPORT

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Dentinogenic Ghost-Cell Tumor: An Uncommon Entity

Dentinojenik Hayalet Hücre Tümör: Yaygın Olmayan Entite

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Abstract

Objectives: Dentinogenic Ghost Cell Tumor (DGCT) constitutes less than 3% of all odontogenic tumors, making it a rare entity characterized by local invasion. The histopathological features of DGCT are the presence of ghost cell layers and the formation of dentinoid material with an ameloblastoma-like odontogenic epithelial proliferation. The peripheral variant of DGCT behaves less aggressively than the intraosseous variant and usually occurs on the anterior region of both jaws.

Case Report: A 60-year-old male patient was referred to the oral and maxillofacial surgery clinic with a complaint of a painless swelling extending from the left upper molar region to the canine region. In the radiographic examination, destruction was observed in the maxillary bone where the lesion was located. Incisional biopsy revealed the diagnosis of DGCT. The patient was scheduled for surgery, but the lesion could not be excised because he refused the treatment.

Conclusion: DGCT may present as a malignant-like lesion in the maxillary posterior region. Therefore, the histopathological differential diagnosis must be made.

Keywords: *Maxilla, Neoplasm, Odontogenic Tumor*

Özet

Amaç: Dentinojenik hayalet hücreli tümör (DHHT), tüm odontojenik tümörlerin %3'ünden azını oluşturur ve bu da onu lokal invazyonla karakterize nadir bir antite haline getirmektedir. DHHT'nin histopatolojik özellikleri, hayalet hücre tabakalarının varlığı ve dentinoid materyalin oluşumu ile ameloblastoma benzeri odontojenik epitelyal proliferasyonudur. DHHT'nin periferik varyantı, intraosseöz varyantına göre daha az agresif davranır ve genellikle her iki çenenin ön bölgesinde görülmektedir.

Olgu Sunumu: 60 yaşındaki erkek hasta, sol üst çene molar bölgesinden kanin bölgesine kadar uzanan ağrısız şişlik şikayeti ile ağız, diş ve cerrahisi kliniğine yönlendirildi. Radyografik incelemede, lezyonun bulunduğu üst çene kemiğinde rezorpsiyon gözlendi. İnsizyonel biyopsi sonucunda DHHT tanısı konuldu. Hasta ameliyat için planlandı ancak tedaviyi reddettiği için lezyon çıkarılamadı.

Sonuç: DHHT, maksiller posterior bölgede malign benzeri bir lezyon olarak ortaya çıkabilir. Bu nedenle mutlaka histopatolojik ayırıcı tanı yapılmalıdır.

Anahtar Kelimeler: *Maksilla, Neoplazm, Odontojenik Tümör*

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Introduction

Odontogenic tumors are lesions that are classified from benign to malignant lesions in the maxillofacial region, and their clinical features and behaviors can vary.[1] DGCT was first reported in the literature in 1946 by Thoma and Goldman.[2] Praetorius et al.[3] have classified DGCT as a solid form of keratocystic odontogenic tumor (KOT) (Type II). The naming dilemma of the lesion was resolved by the World Health Organization (WHO) in 2005. WHO classified this lesion in three groups. These groups are; calcified cystic odontogenic tumor, DGCT, and ghost cell odontogenic carcinoma. WHO has defined DGCT as "a locally invasive neoplasm in a mature collagenous stroma, including epithelial cell islands resembling ameloblastoma".[4] Nevertheless, in the WHO classification of in 2017, DGCT was classified under tumors of benign mixed epithelial mesenchymal origin.[5] In the WHO classification in 2022, the DGCT classification has not been changed and is in the subclass of benign mixed odontogenic tumors.[6]

DGCT is a benign neoplasm with biphasic morphology and shows local infiltration. It consists mostly of ameloblastomatous cell proliferation and to a lesser extent basaloid cells. The characteristic finding of DGCT is abnormal keratinization with a variable amount of ghost cells. The hard tissue formation resembles osteodentin or dentinoid deposits.[5]

There are two variants of DGCT; intraoseous and peripheral. The intraosseous variant is mostly seen in the premolar region. Peripheral variant is rare but mostly seen in the anterior mandibular region.[7] The dimension of intraosseous DGCTs can range from 1 to 10 cm or larger. Clinical features of intraosseous DGCTs can include obliteration of the maxillary sinus, noticeable swelling, enlargement of the jaw, facial asymmetry, infiltration of soft tissues, associated pain, tooth mobility or displacement, and root resorption.[8,9] The majority of peripheral DGCTs are seen in edentulous patients and originate from the gingiva and oral mucosa. It usually appears as an exophytic pedunculated or sessile lesion.[10] Since the recurrence is very rare, a conservative approach is sufficient in most cases.[2] In this case report,a case of DGCT occurring on the maxillary posterior region is presented with a detailed literature analysis.

Case Report

A 60-year-old patient presented to the Oral and Maxillofacial Surgery Department with complaints of swelling in the left maxillary region and difficulty chewing. The anamnesis revealed that the swelling progressed slowly, and there was no complaint of pain in the related area. Clinical examination revealed a firm, well-defined swelling and hardened expansive mass on the alveolar crest extending from the left upper canine region to the molar region (Fig. 1).

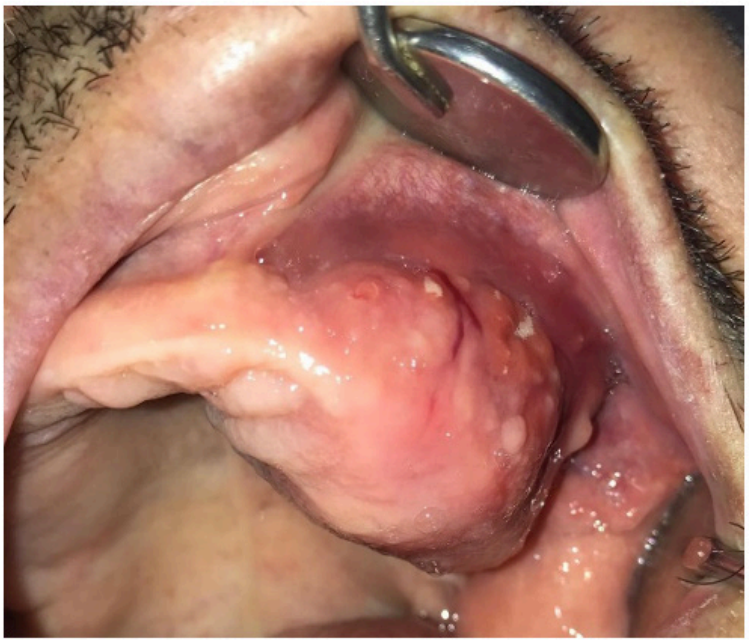

Figure 1. Intraoral soft tissue swelling presenting as a suspected malignancy on the left upper region of the maxilla.

The mucosa was intact and had a slightly erythematous appearance. Panoramic radiography showed a lytic radiolucent bone lesion with a soft tissue counterpart, which degenerated the cortical plate and extended to the oral cavity. Cortical expansion with a diameter of 31.62x32.51 mm was detected in the left maxillary posterior region in the Cone beam computerized tomography (CBCT) examination (Fig. 2 and 3).

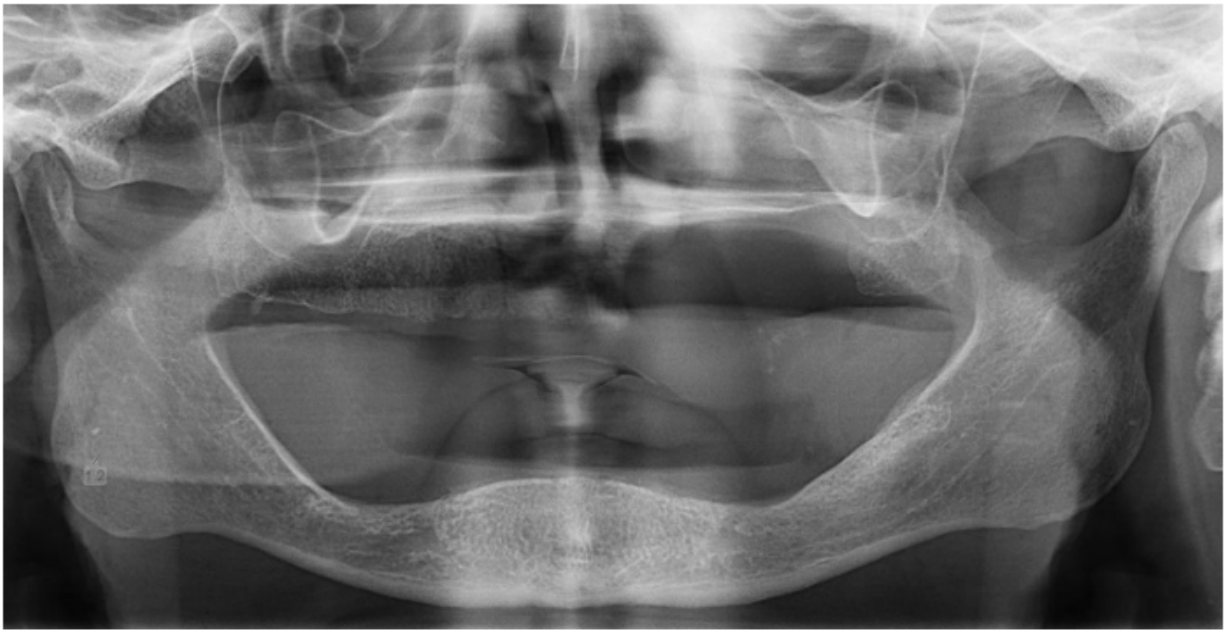

Figure 2. Panoramic radiography shows a large, well-circumscribed radiolucency lesion involving the left maxillary region with a well-defined border.

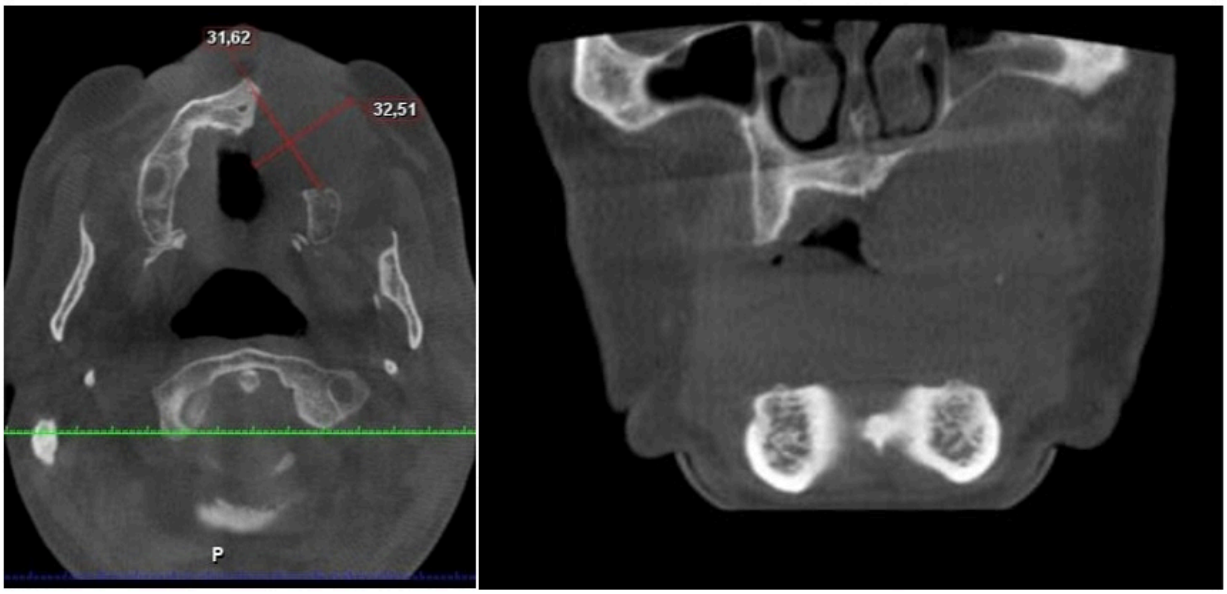

Figure 3. CBCT image showing well-defined, lytic expansile radiolucency in the left maxillary posterior region.

It was decided to perform an incisional biopsy for a definitive diagnosis. After obtaining consent from the patient, an incisional biopsy was taken under local anesthesia, and the specimen was sent for histopathological examination (Fig. 4).

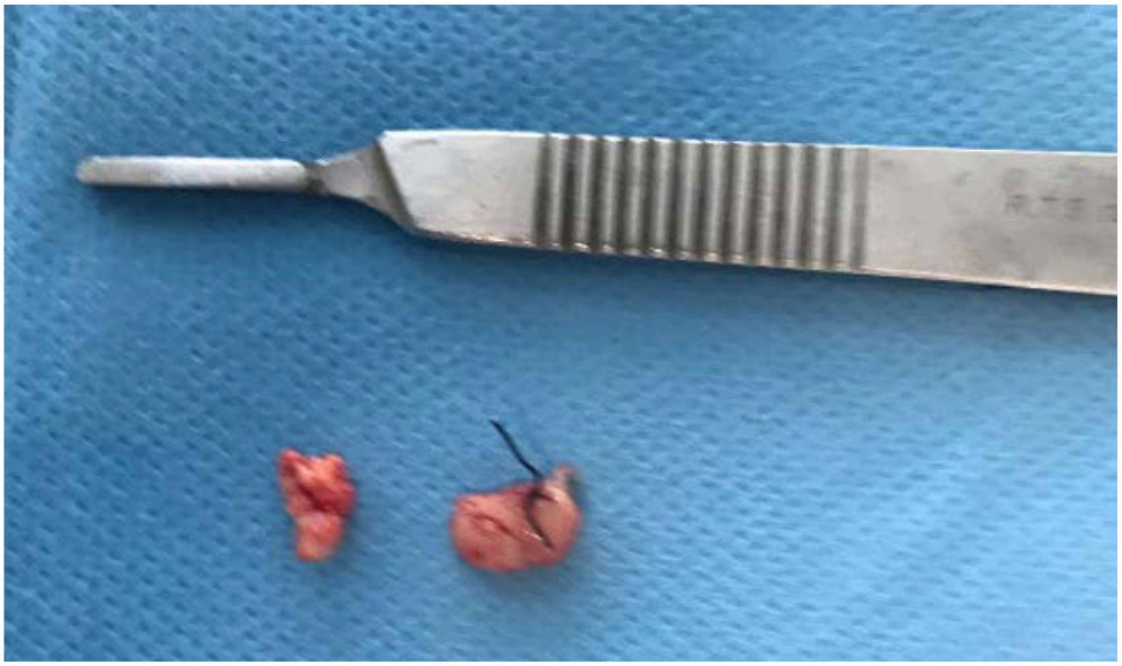

Figure 4. Macroscopic appearance of incisional biopsy specimen.

The lesion had ameloblastoma-like epithelial neoplastic proliferation accompanied by ghost cell foci in the histological examination (Fig. 5). There was no evidence of malignancy. A diagnosis of benign DGCT was made. Surgical excision was planned under general anesthesia. However, the patient refused the treatment and lost the chance to follow up.

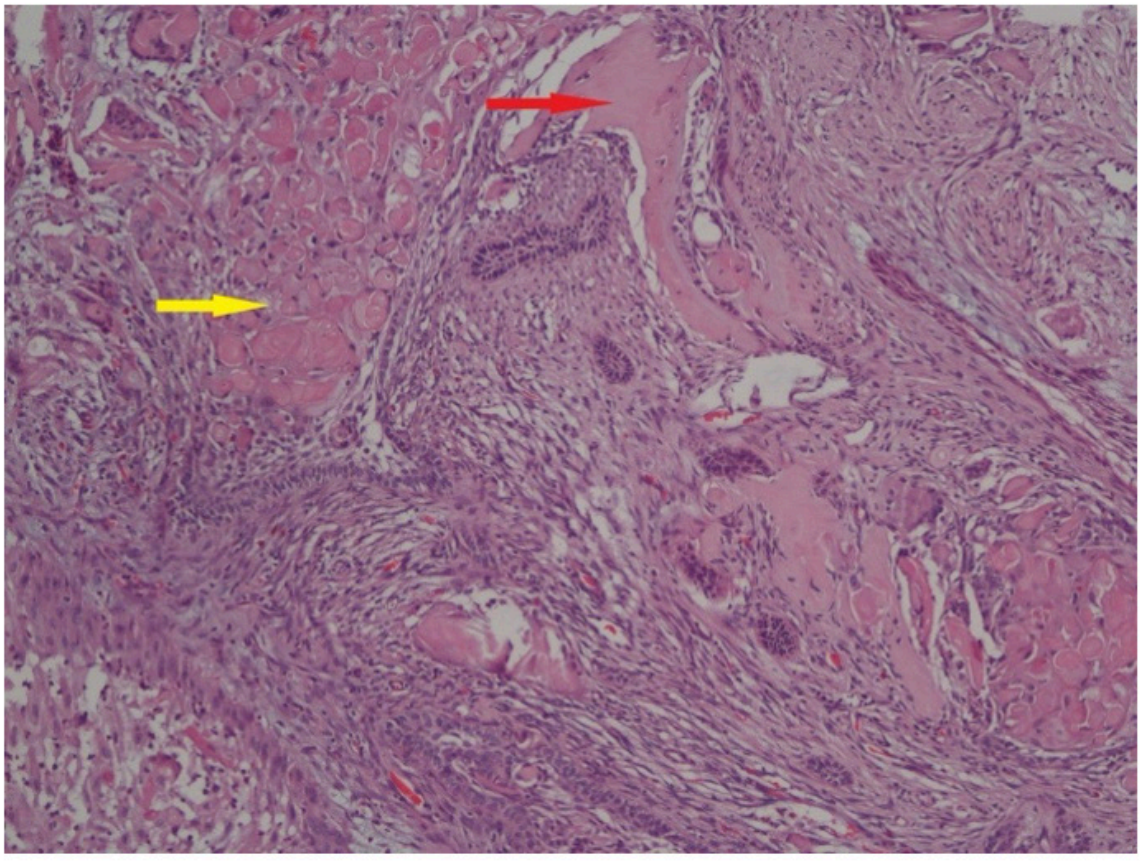

Figure 5. Histopathological image of DGCT lesion showing odontogenic epithelium with ghost cells (yellow arrow) and dentinoid material (red arrow) (Hematoxylin-Eosin stain ×100).

Discussion

DGCT histopathologically demonstrates layers of odontogenic cells of odontogenic origin and round islets in a mature collagenous stroma. The epithelium of the tumor islands is ameloblastoma-like. These tumor cells do not have mitotic figures. Small cysts may form on epithelial islands.[11] Central DGCT is more aggressive than periperal DGCT. It has a growth pattern with localized invasion, and a high recurrence rate has been reported even after resection. Peripheral DGCT is less common. It occurs on the gingival or alveolar mucosa. It shows restricted growth potential.[12] A total of 130 cases of DGCT have been seen worldwide in recent literature reviews.[13] It accounts for less than 3% of all odontogenic tumors.[14] Although it is more common in the anterior region of the jaws, it affects both jaws. Clinically, the lesion is asymptomatic but causes noticeable swelling with facial asymmetry, which is dependent on the size of the lesion.[15] In the current case, DGCT was located on the posterior region and maxilla, in contrast to the anterior region, which is the majority of the literature. The painless growth and swelling in the patient is consistent with the literature. The male-female ratio of DGCT is 3:2, and it occurs more frequently in males.[13] Although it is seen with a wide incidence in the age range of 20-70 years, it is most commonly seen in the age range of 40-60.[16] In this case, as consistent with the literature, the patient was 60 years old. DGCT appears radiologically as a well-circumscribed, unilocular, multilocular radiolucent, or mixed lesion.[17] In this case, an unilocular well-circumscribed radiolucent lesion was seen.

The treatment approach for DGCT varies depending on the variant. Conservative surgery is typically employed for peripheral DGCT, while more aggressive surgical resection with adequate safety margins is recommended for central DGCT due to its higher recurrence rate.[2,13,18] Regular postoperative follow-up is essential to monitor for recurrence, which can be as high as 71% in central DGCT.[18] An operation was planned for our patient, but the surgery could not be performed because the patient refused treatment due to his psychological disorder.

Conclusion

DGCT may occur as a protruding malignant-like soft tissue lesion on the maxillary posterior region of the oral cavity. Clinical information about DGCT is very scarce in the literature. In cases where a definitive diagnosis cannot be made, histopathological differential diagnosis by a pathologist specializing in maxillofacial lesions is important to determine the presence of ghost cells and dysplastic dentin. Therefore, more studies on this entity are needed in order to diagnose and treat patients with DGCT in the most effective way.

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Conflicts of İnterest

The Authors declare that there is no conflict of interest.

Authors' Contribution

Concept/Idea: K.N.S, D.O Design: K.N.S, D.O Supervision/Consultation: K.N.S Analysis and/or Interpretation: K.N.S Literature Search: T.G Manuscript Writing: G.T., K.N.S Critical Review: K.N.S, D.O, G.T., G.M.K., K. Y.Ç, A.M.F.

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