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Case Report
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Odontogenic Carcinoma with Dentinoid Involving the Maxilla: Case Report and Literature Review

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Abstract

Objective: To report a rare case of odontogenic carcinoma with dentinoid involving the maxilla and highlight the challenges in diagnosing and managing this uncommon entity.

Methods: A 19-year-old male presented with nasal congestion, intermittent cheek pain, and left cheek numbness. Imaging revealed a 5.6 cm mineralized mass in the left maxillary sinus with cortical destruction. Multiple biopsies were performed, with the final pathology confirming odontogenic carcinoma with dentinoid.

Results: The patient underwent left infrastructure maxillectomy with microvascular fibula free flap reconstruction. Final pathology showed odontogenic carcinoma with dentinoid without perineural or lymphovascular invasion and negative margins. No adjuvant therapy was recommended, and the patient has shown no evidence of recurrence to date.

Conclusion: Odontogenic carcinoma with dentinoid is a rare, locally destructive malignancy that can be challenging to diagnose due to its rarity and histological similarity to benign odontogenic tumors. This case highlights the importance of maintaining a high index of suspicion for malignancy in odontogenic tumors with aggressive features and the need for thorough diagnostic workup, including multiple biopsies if necessary.

Keywords: Maxillofacial surgery; General dental; Mouth; Oral pathology; Pediatric ORL; Endoscopic sinus surgery; Rhinology; Oncology

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Introduction

Odontogenic carcinomas are uncommon low-grade malignancies arising from remnants of odontogenic epithelium, most commonly found in the mandible but can also rarely present in the maxilla. Odontogenic carcinomas are locally destructive that typically present with asymptomatic swelling and seldomly with pain or discomfort. They have been reported to demonstrate both perineural invasion and adjacent tissue invasion with a high potential for recurrence, making early diagnosis and prompt treatment crucial for a favorable outcome [1-9]. Previously, Odontogenic carcinoma with dentinoid was included in the 2017 World Health Organization (WHO) classification of odontogenic and maxillofacial bone tumors; however, as of 2022, this is no longer the case with many previously described Odontogenic tumors with dentinoid in the literature no longer being formally recognized due to its rarity and does not currently fit into any WHO-defined entity [10,11]. Given the rarity of this disease process with only 11 total cases reported, odontogenic carcinoma with dentinoid remains a poorly described entity.

Case Presentation

A 19-year–old man presented initially with complaints of nasal congestion, nasal drainage, post-nasal drip, intermittent left cheek pain, and ultimately left cheek numbness.

Computed tomography imaging showed a 5.6 cm mass centered on the left maxillary sinus, involving the ipsilateral superior alveolar ridge with extension into the nasal cavity and buccal space. The mass was largely mineralized with multifocal marked cortical destruction along the margins. Magnetic resonance imaging demonstrated a 5.7 × 5.1 × 5.5 cm T2 hyperintense and T1 isointense complex expansile mass involving the left nasal cavity, masticator space, and abutting the inferior orbital wall (Figure 1).

Nasal endoscopy showed a large expansile mass in the maxillary sinus medializing the inferior

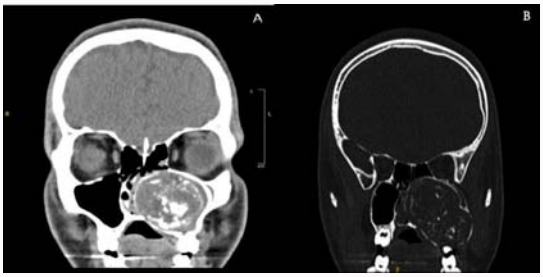

Figure 1: Imaging examinations of odontogenic carcinoma with dentinoid. (A) Computed tomography scan (coronal section), showing involvement of the left orbit and left septum by the tumor. (B) Computed tomography scan (coronal section), showing the heterogeneously enhancing mass centered within the left maxillary sinus.

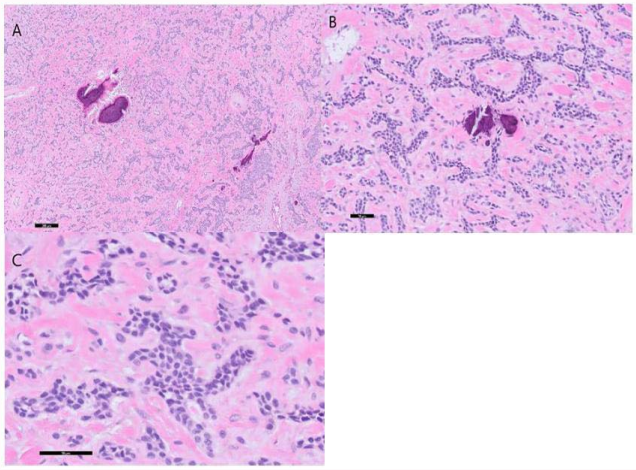

Figure 2: Histopathological examinations of odontogenic carcinoma with dentinoid. (A) 5X image showing cords of odontogenic epithelium, some with a pseudoglandular appearance with foci of small to large calcifications (B) 20X image showing cords of odontogenic epithelium, some with a pseudoglandular appearance with foci of eosinophilic mineralized material consistent with dentinoid. (C) 40X image showing round to oval cells with scant to moderate cytoplasm lacking significant atypia and pleomorphism. The cytoplasm ranges from amphophilic to clear at places with minimal mitosis.

meatus that appeared "bony in consistency." The patient subsequently underwent left-sided endoscopic sinus surgery including a left medial maxillectomy for access, revealing the tumor to have a fibrous shell. Final pathology demonstrated an epithelioid neoplasm with microcalcifications and a fibrous component on the left medial margin displaying bland spindle cell proliferation suggestive of odontogenic fibroma. Given the aggressive imaging features and presentation there was a high degree of suspicion for malignancy, and the patient was taken back to the operating room for repeat biopsy via a Caldwell-Luc approach. Final pathology from this biopsy showed thin cords, nests and strands of odontogenic epithelium embedded within a dense collagenous stroma with zones of hard tissue formation ranging from dentin-like material to bone formation, consistent with odontogenic carcinoma with dentinoid.

The patient subsequently underwent a left infrastructure maxillectomy with microvascular fibula free flap reconstruction, concurrent dental implant surgery, and direct placement of teeth within the fibula or "jaw in a day" procedure" allowing a comprehensive approach to reconstructing the jaw for definitive treatment. Final pathology from this resection revealed odontogenic carcinoma with dentinoid without perineural or lymphovascular invasion with negative margins (Figure 2). The patient was presented at a multidisciplinary tumor board, during which it was determined that no post- operative adjuvant therapy was recommended. Upon follow-up, there has been no discernible evidence of recurrence thus far, as ascertained through physical examination and imaging modalities.

Discussion

Odontogenic carcinoma with dentinoid is a rare subtype of odontogenic malignancy that is quite difficult to diagnose given its rarity and difficulty in distinguishing from benign odontogenic tumors on pathology, often leading to delays in treatment [12-14]. Our case report reflects the difficulty in diagnosing this disease process with multiple deep biopsies required to obtain an accurate diagnosis. Our experience underlines the importance of maintaining a high index of suspicion for malignancy in cases of odontogenic tumors with aggressive features based on signs or symptoms as well as imaging. The difficulty in accurately diagnosing a malignant odontogenic tumor histologically was highlighted in a recent systematic review [15]. Marin et al. [15] reported that out of 507 patients with malignant odontogenic tumors, 24.7% had an initially benign diagnosis before an eventual malignant diagnosis after repeat biopsy. They found 60 different unique diagnoses recorded before reaching the final diagnosis [15]. An additional study reported out of 22 reported malignant odontogenic tumors, only 9 were considered malignant on diagnosis [16]. Due to the infiltrative and destructive nature of many benign odontogenic tumors, it is difficult to discern them from malignancy based off clinical features. Additionally, many histological features such as nuclear and cellular pleomorphism, crowding and budding, and vascular or perineural infiltration can be shared between both tumors [15,17,18]. In our case, initial pathology on deep biopsy was most consistent with odontogenic fibroma. Out of the 11 prior reports of odontogenic tumor with dentinoid found in the literature, there were 7 males and 5 females with a mean age of 41.8 [1-7,9,19]. 6 of these tumors occurred in the mandible, four in the maxilla, and one case reported found in the nasal cavity [1-7,9,19]. The most common presenting sign among the cases was asymptomatic swelling, reported in 9 cases [1,3,5-7,9].

Pain or discomfort was reported in only 5 of the 11 cases, and tooth resorption was seen in 4 cases [1,2,4]. All tumors appeared as well-defined masses on imaging, with 7 containing calcified material in small patchy areas or larger radiopaque foci and 3 cases described as multilocular [1,6,9]. All cases were treated with surgical excision. 5 patients had local disease recurrence, 4 of which were treated with repeat resection and 1 with adjuvant radiotherapy and chemotherapy [1-7,9,19]. Histological analysis of all cases displays a low-grade but locally destructive malignancy with well-defined margins [1-7,9,19]. In terms of difficulty in diagnosis, 3 of the previously reported cases were diagnosed as several separate entities before arriving at a final diagnosis of odontogenic carcinoma with dentinoid. The first case was initially diagnosed as an "epithelial odontogenic tumor, unclassified" of the left mandible on a biopsy but on definitive resection was diagnosed as clear cell odontogenic tumor with dentinoid [5]. The second case was initially clinically diagnosed as an ameloblastoma also in the left mandible premolar region. Because the tumor showed destruction of the lingual cortical bone and invasion of the inferior alveolar neurovascular bundle and adjacent soft tissue; bilateral central incisors, bilateral lateral incisors, the left canine,

left second molar, as well as the neurovascular bundle were excised extracted and excised along with the tumor. The tumor was later reclassified as an odontogenic tumor with dentinoid. There was no evidence of recurrence or metastasis for 11 months after the surgical treatment [6]. The last case included a mass between the lateral incisor and canine of the left mandible diagnosed as an Adenomatoid Odontogenic Tumor (AOT) after enucleation and subsequent histologic analysis. The patient reappeared with an apical radiolucent lesion in the incisor area with a second biopsy also diagnosed as AOT, recurrent. During follow- up, recurrences were noted twice more and again reported to be AOT. After a fourth recurrence in the maxillary sinus partial maxillectomy was performed, with an eventual appropriate. diagnosis of the odontogenic tumor as an adenoid ameloblastoma with dentinoid, with no further recurrence [3]. Of note, the 2022 WHO histological classification of Tumors no longer specifically recognizes odontogenic carcinoma with dentinoid, as well as many other odontogenic malignancies. The only odontogenic malignancy composed of dentinoid recognized is the dentinogenic ghost cell tumor which is a benign mixed epithelial and mesenchymal odontogenic tumor [20,21]. The lack of formal recognition for these odontogenic malignancy subtypes may impede the ability to efficiently recognize and treat these already difficult- to diagnose tumors. Odontogenic carcinoma with dentinoid is a low-grade locally destructive tumor with a low incidence of perineural invasion and metastasis. Presentation is typically isolated asymptomatic swelling in the mandibular or maxilla. It is crucial to have a high degree of suspicion for malignancy whenever an odontogenic tumor presents with aggressive features clinically or on imaging. Odontogenic malignancies are frequently misdiagnosed initially, and multiple deep biopsies may be necessary to obtain an accurate diagnosis.

Précis of Manuscript

Already known

- Odontogenic carcinomas are rare, low-grade malignancies commonly found in the mandible.
- They are difficult to diagnose due to their rarity and overlap in clinical and histological features with benign tumors.
- Accurate diagnosis often requires multiple biopsies and can be misdiagnosed initially. What This paper adds:
- Provides a detailed case report of odontogenic carcinoma with dentinoid in the maxilla, a location far less commonly affected.
- Demonstrates the necessity of maintaining suspicion for malignancy in odontogenic tumors with aggressive features, despite initial benign histological findings.
- Discusses the implications of the WHO's 2022 reclassification of odontogenic tumors, which may affect the recognition and treatment of these rare tumors.

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