



## **Desmoplastic Ameloblastoma in Maxilla: A Report of Rare Case with Review of Literature and Classic Histopathology**

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### **Authors' contributions**

*This work was carried out in collaboration between all authors. Author VGD designed the study, performed the statistical analysis, wrote the protocol, and wrote the first draft of the manuscript. Authors SS and PS managed the analyses of the study. Author DR managed the literature searches. All authors read and approved the final manuscript.*

### **Article Information**

DOI: 10.9734/ARRB/2017/38186

Editor(s):

- (1) Manikant Tripathi, Department of Microbiology, Dr. Ram Manohar Lohia Avadh University, Faizabad, India.  
(2) George Perry, Dean and Professor of Biology, University of Texas at San Antonio, USA.

Reviewers:

- (1) Sucheta Bansal, Himachal Institute of Dental Sciences, India.  
(2) Manas Bajpai, NIMS Dental College, India.  
(3) César Luiz Da Silva Guimarães, Federal University of Rondônia, Brazil.  
Complete Peer review History: <http://www.sciedomain.org/review-history/22475>

### **Case Study**

**Received 16<sup>th</sup> November 2017**

**Accepted 21<sup>st</sup> December 2017**

**Published 27<sup>th</sup> December 2017**

### **ABSTRACT**

Ameloblastoma is a very rare odontogenic tumor of the oral cavity, with different histological variants. One of the types of ameloblastoma is desmoplastic ameloblastoma (DA) which has 4-5% of incidence. Here we review and reported the desmoplastic ameloblastoma is the least of occurrence of all the variants of ameloblastoma. The uniqueness of this lesion can be further enlightened with respect to its site of occurrence, the radiographic feature and the histological

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appearance different from the classical type of ameloblastoma. This case report focuses on a DA that occurred in the maxilla of 25-year-old women and explains about clinical, CT scan, histopathological finding, and treatment plan. The patient is undergoing routine follow-up and is presently free of disease.

From this case, the clinicians should remember to consider the desmoplastic ameloblastoma as differential diagnosis, if a patient complaint of swelling in and around the premolar region of the maxilla.

**Keywords:** *Odontogenic tumor: ameloblastoma; desmoplastic variant; maxilla; benign fibrooseous lesion.*

## 1. INTRODUCTION

Ameloblastoma is the most common odontogenic tumor of epithelial tissue origin. It is the second most common odontogenic neoplasm [1]. Ameloblastoma are rare benign, slow-growing but locally aggressive, polymorphic neoplasm of proliferating odontogenic epithelial origin. Ameloblastoma is a benign tumor whose importance lies in its potential to grow into enormous size with resulting bone deformity [2]. The incidence, clinical, radiological features, histopathological feature, and behavior of ameloblastoma have been extensively reviewed in numerous literatures. However, desmoplastic ameloblastoma do not show a typical clinical or radiographic or histopathologic feature of ameloblastoma [3].

Ameloblastoma is the most common benign odontogenic tumor with various geographical prevalence, but it is most common in China and Africa, while it is the second most common in the United States and Canada (odontoma being most common) [4]. According to some studies, desmoplastic ameloblastoma was most commonly found in Japanese and Chinese [3].

Ameloblastoma (AM) of the jaw is a neoplasm that accounts for about 1% of all tumors of the jaws and 18% of the various odontogenic neoplasms [5]. Desmoplastic ameloblastoma is rare, accounting for approximately 4% to 13% of ameloblastoma [5,6]. According to a review of literature carried out by Gandhi et.al, 2017 found that 163 cases [6].

Leon Barnes has categorized ameloblastoma into four types on the basis of the behavioral pattern, clinical and radiographic characteristics, histopathology and prognostic aspects into [7] –

1. The classic solid/ multicystic ameloblastoma
2. The unicystic ameloblastoma

3. The peripheral ameloblastoma
4. The desmoplastic ameloblastoma.

However recent studies declared that the desmoplastic variant of ameloblastoma is a separate entity in itself, based on its clinical, site of occurrence, unusual radiologic appearance and histopathological features [5,7]. The purpose of this article is to present a case of desmoplastic ameloblastoma in clinical, radiologic and histopathological aspect and to provide a brief review of the literature.

## 2. CASE ANALYSIS

A 24-year-old female patient complained of a swelling in the posterior region of the right maxilla, which had been present for about six months. The swelling was solitary, dome-shaped, slowly enlarging, indurated mass covered by intact mucosa. There was no bleeding, pain, or any sensory changes. Involved teeth were vital and no mobility. The swelling extended from the right lateral incisor to right second premolar and right side of the hard palate and buccal cortex was intact, and normal in color and appearance. No lymphadenopathy or sinus or fistulae were observed. There was no previous medical history or trauma. The patient had no related past dental/medical history. On oral clinical examination, first premolar and canine on the left side of the maxilla were absent. Based on these findings a clinical diagnosis of fibro-osseous lesion was given (Fig. 1).

The CBCT showed a mixed hyperdense-hypodense image with buccal expansion and the displacement of teeth. There was no perforation present. A panoramic radiograph revealed a poorly defined, mixed, radiolucent-radiopaque in the area of the lesion. Maxillary sinus was not involved and there was no evidence of root resorption. The above features were suggesting that benign fibro-osseous lesion (Fig. 2).



**Fig. 1. Preoperative picture of the patient showing swelling present on the maxillary premolar region**

All laboratory investigations were carried out and were found to be normal. The incisional biopsy of the lesion was performed under local anesthesia and a surgical specimen was sent for the histopathological examination.

The Haematoxylin & Eosin stained tissue section revealed numerous proliferating odontogenic follicles of varying sizes and shapes, immersed in a thick, dense collagenous stroma, along with irregular trabecular bone formation in a few areas. The follicle shows flattened peripheral cells, and polygonal and spindle-shaped central cells compactly arranged. Few follicles are compressed, also exhibit cystic degeneration and squamous metaplasia surrounding connective tissue show extensive desmoplasia. All the above features suggest the diagnosis of Desmoplastic Ameloblastoma of the anterior mandible.

The patient underwent surgical resection and currently routine follow-up done with no signs of recurrence after 12 months.

### 3. DISCUSSION

In the year 1984, Eversole et al. who first reported a case of Desmoplastic ameloblastoma and they termed it as 'Ameloblastoma with pronounced Desmoplasia' [8]. It has been classified by World Health Organization Classification of Head and Neck Tumors (WHO-2005) as a variant of ameloblastoma with typical clinical, imaging and histological features [9]. But now, the newer classification of World Health Organization (WHO 2017) has been dropped and

considered as one of the histological variants of conventional ameloblastoma [10].

Desmoplastic Ameloblastoma (DA) is a rare variant of ameloblastoma, with characteristic clinical, radiological and histopathological features. The biologic behavior of this tumor is not completely understood because of the less number of reported cases [9].

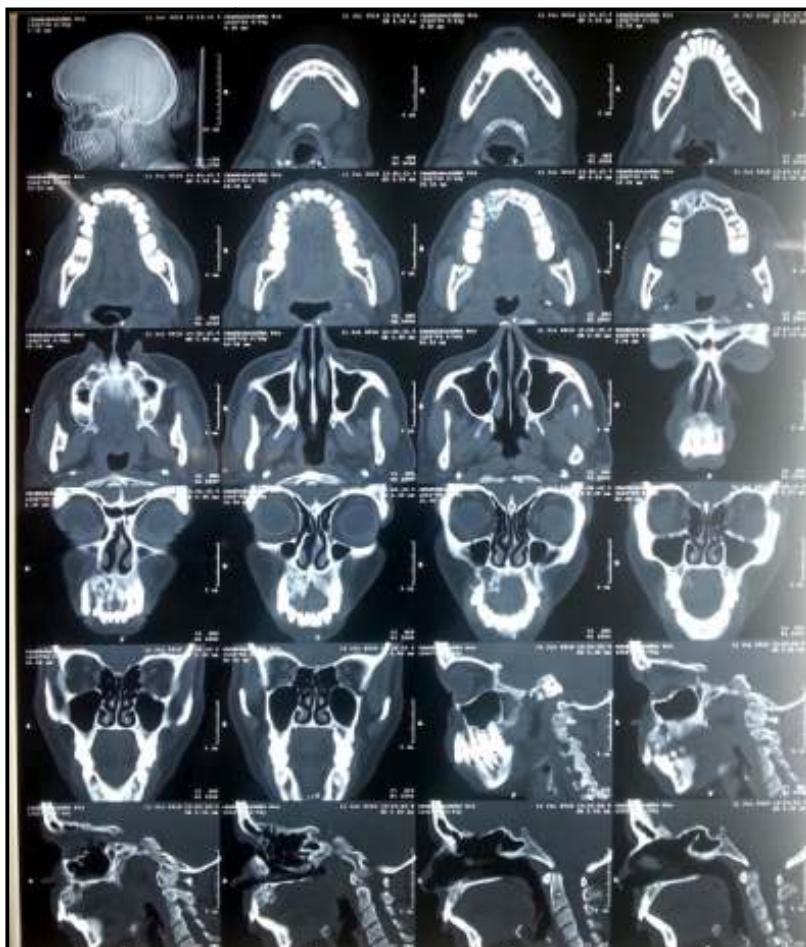
Although desmoplastic ameloblastoma are similar to conventional solid ameloblastoma regarding the age (3<sup>rd</sup> to 5<sup>th</sup> decade of life) and no gender distribution, studies have demonstrated that these tumors present a strong predilection for the anterior region of the jaws, equally in the maxilla and mandible [8,9]. But few authors suggested that desmoplastic ameloblastoma has a slight predilection for male and majority of the cases occurred in the anterior region of the mandible [3,7]. Contrast to this, our present case shows the age of the patient was 24 years-old female in the premolar region of the maxilla.

As reported in the literature, the desmoplastic ameloblastoma is usually a painless swelling with buccal expansion and tooth displacement with no root resorption [11]. Tooth displacement is a common feature in DAs in almost 92% of the cases and root resorption is seen in 33% of cases [5]. Similarly, our case also showed tooth displacement without root resorption. Origin of desmoplastic ameloblastoma was suggested that it develops from the periodontal membrane of the related tooth and some suggest that it might arise from epithelial rests of Malassez in the periodontal membrane [1].

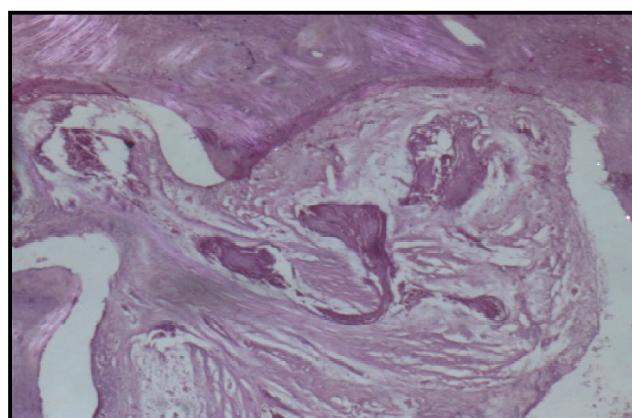
The radiologic picture describes mixed radiolucent and radiopaque areas reflecting the osteolytic and sclerotic areas with ill-defined borders mimicking a benign fibro-osseous lesion [8,9,11]. Shashikanth MC et al. explained that desmoplastic ameloblastoma has 64% of cases of mixed radiolucent/radiopaque appearance and are radiolucent in 36% of cases [5]. Reichart et al. stated that 51% of the tumors were unilocular and rest is multilocular radiolucency, [11] but Effiom and Odukoya stated that multilocular radiolucency is the predominant radiographic presentation of the desmoplastic ameloblastoma [6]. The high incidence of poorly defined borders can be attributed to the pattern of infiltration into marrow spaces and very thin cortical bone of the maxilla represents a weak barrier for the spread of tumors, so that maxillary ameloblastoma may

spread earlier than mandibular neoplasms [1,9]. These features may also be of prognostic significance in predicting tumor behavior [3]. The

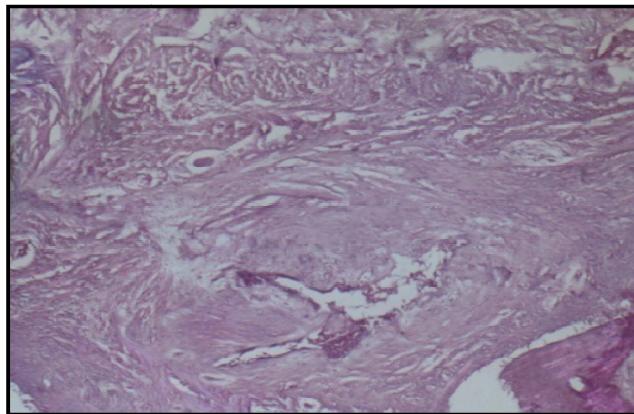
present case shows mixed radiopaque and radiolucent areas with ill-defined borders, cortical expansion, and tooth displacement.



**Fig. 2. Computed tomography revealed a well-defined lytic-expansile lesion in the left maxilla with non-sclerotic margins**



**Fig. 3. Hematoxylin and Eosin stained tissue section shows odontogenic follicles appears in irregular shapes in a desmoplastic stroma (40x magnification)**



**Fig. 4. Haematoxylin and eosin stained section shows epithelial islands in a mature fibrous connective tissue stroma (100x magnification)**

Histological features consisted of proliferating, irregularly shaped islands and cords of the odontogenic epithelium of varying sizes embedded in a desmoplastic connective tissue stroma. The tumor islands may show a pointed, stellate or "kite-like" appearance. The epithelial cells at the periphery of the islands are cuboidal cells with occasionally show hyperchromatic nuclei and microcyst formations in the center were also noted [3]. Columnar cells demonstrating reversed nuclear polarity are rarely conspicuous although some islands may show ameloblast-like cells peripherally [5,11]. Philipsen et al. suggested that the stromal osteoplasia is not a part of a reparative process but may be due to metaplastic changes in the stroma caused by tumor cell stimulation of mesenchymal cells. These metaplastic trabeculae consisted of woven bone, contained large osteocytes and were surrounded by active osteoblasts that were randomly scattered into the stroma. Later it was hypothesized that mixed radiolucent/radioopaque appearance of some lesions may be attributed to these metaplastic foci [3,12].

### **3.1 Histological Criteria for the Diagnosis of Desmoplastic Ameloblastoma [7] :**

1. Stromal desmoplasia, in the form of moderately cellular, fibrous connective tissue with abundant collagen.
2. Islands of various sizes and shapes in the epithelial component.
3. Peripheral layer of cuboidal cells and
4. Hypercellular central area composed of spindle-shaped or polygonal epithelial cells

The differential diagnosis for desmoplastic ameloblastoma based on histopathologic features, such as ameloblastic fibroma, odontogenic fibroma, and squamous odontogenic tumor must be established [8]. Ameloblastic fibroma shows interlacing scattered thin island of odontogenic epithelium which has columnar cells at the periphery and stellate cells at the center. These odontogenic island scattered in primitive mesenchyme. Odontogenic fibroma composed of mature collagen fiber interspersed with plump fibroblasts and small nests or island of odontogenic epithelium which are inactive. Squamous odontogenic tumor is composed of the island of mature squamous epithelium with flat or cuboidal cells at the periphery. The stroma shows a fibrous i.e mature bundle of collagen fiber without inductive effect. But desmoplastic fibroma shows dense collagen stroma appears hyalinized and hypocellular with thin cord and stand like odontogenic epithelium [13].

Immunohistochemistry shows variable expression of S-100 protein, desmin, capsid-3, increased expression of Fas and p63, decreased expression of cytokeratin. There is strongly positive for fibronectin, Type I collagen and collagen type VI which rules out scar tissue, immune-negativity for tenascin, [8] Oxytalan fibers have been demonstrated in the stroma of DA. By these findings, some author suggests that desmoplastic ameloblastoma arise from the epithelial rests of Malassez in the periodontal membrane of the related tooth, [5] or originate from de novo synthesis of extracellular matrix proteins [3].

Overexpression of transforming growth factor beta (TGF- $\beta$ ) suggested that it is the most potent local factors for modulating extracellular matrix formation. Furthermore, it has been proposed that this phenomenon has its origin from a new protein synthesis that comes from the extracellular matrix which has a fundamental role in supporting, adhering, proliferating, migrating and differing tumor cells, interfering in the behavior and modulation of the tumor cells might be related to the phenomenon of desmoplasia [8,12].

Although resection still is the most common treatment modality followed, some cases are treated by enucleation and curettage. But curettage is considered an inappropriate treatment for desmoplastic ameloblastoma due to incomplete removal of the lesion or may be tumor cells infiltrate the surround bone trabeculae or they fail to produce a capsule which results in recurrence [1,7,9]. Hence it is recommended, desmoplastic ameloblastoma should be always treated by complete surgical resection with proper follow up.

Desmoplastic ameloblastoma exhibits a more aggressive behavior than other types of ameloblastoma. This aggressiveness may be due to [1,9,11]

- 1) Potential to grow to a large size
- 2) Common location in the maxilla leading to an early invasion of adjacent structures
- 3) Diffuse radiographic appearance, and
- 4) Histological finding of bone invasion

From this case, we gained knowledge about DA is considered a rare lesion and it can be considered as a well-defined lesion. If a patient attends with complaints of swelling in and around the premolar region, clinicians should consider the desmoplastic ameloblastoma as one of the diagnosis.

#### **4. CONCLUSION**

The desmoplastic variant of ameloblastoma is comparatively rarer than the other types of ameloblastoma. The clinical and radiographic features of DA resemble that of fibro-osseous lesions but the definitive diagnosis should always be based on the histopathological findings. Furthermore, the lesions in maxilla are close to many vital structures and the thin cortical bone is a weak barrier and the tumor usually infiltrate into marrow spaces. Thus, a clear-cut border may be

difficult to identify. Since the lesion does not have a fibrous capsule, there are chances of recurrence is high. So it is necessary for the surgeon to establish the definitive diagnosis to prevent any error in the treatment plan modalities and a regular follow up of the case. For the proper understanding of such cases, more in-depth knowledge and long-term follow up is required.

#### **CONSENT**

As per international standard or university standard written patient consent has been collected and preserved by the authors.

#### **COMPETING INTERESTS**

Authors have declared that no competing interests exist.

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