Desmoplastic Ameloblastoma: A Report of Two Cases
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Abstract
Desmoplastic ameloblastoma has been characterised by World Health Organisation as a variant of ameloblastoma with specific clinical, radiographic and histological features. Presenting clinically as an asymptomatic swelling with a mixed radiographic appearance, these lesions are most often misdiagnosed as fibro-osseous lesions. The recurrence rate of these lesions is found to be equal to or higher than the solid multicystic ameloblastoma unlike other variants like the unicystic ameloblastoma or peripheral ameloblastoma which have a much lower recurrence rate. The infiltrative nature of this variant demands a radical surgical approach as recurrence rate has been found to be higher in cases treated with enucleation and curettage. This case report and review aims to highlight the pathologic and diagnostic aspects of Desmoplastic ameloblastoma aiding in its timely diagnosis and appropriate treatment.

Keywords: Ameloblastoma; Desmoplastia; Multicystic; Maxilla; Mandible.

Introduction
Ameloblastoma is a true neoplasm of enamel organ origin which does not undergo differentiation upto the point of enamel formation. Histologically the neoplasm recapitulates the morphology of pre-enamel matrix stage of enamel organ. Robinson describes it as a unicentric, non-functional, intermittent in growth, anatomically benign and clinically persistent lesion. Although most ameloblastomas are microscopically benign and lack cytologic atypia, they are generally considered as locally aggressive and destructive with a high rate of recurrence. World Health Organization (WHO) classified ameloblastoma into four clinico-pathological entities viz. Solid multicystic ameloblastoma (SMA), unicystic, peripheral and Desmoplastic ameloblastoma. Desmoplastic ameloblastoma, first described by Eversole in 1984, is a rare yet unique entity with specific clinical, imaging and histological features. This case report presents the clinico-pathologic features of two cases reported to our institution.

Case Report 1
A 37 year old male patient reported with a complaint of a hard swelling in the lower right back jaw. The swelling appeared about a year back and had shown increase in size for 2-3 months after its first appearance. On examination, a non-tender, bony hard swelling was noted which caused obliteration of the buccal as well as lingual vestibule causing facial asymmetry. On radiographic examination, lateral oblique view revealed an ill-defined mixed radiolucency with displaced roots of adjacent teeth. An occlusal view showed buccal and lingual cortical plate expansion (Figure 1).

Case Report 2
A 35 year old female patient reported with a painless swelling in the upper anterior jaw left side which has been gradually increasing in size for one and a half year. Intra-oral examination revealed swelling on the buccal aspect of the first premolar region. Panoramic radiography revealed a mixed radiolucency in that region. (Figure 2)

Histopathological examination of the incisional biopsy in the first case revealed ameloblastic epithelial islands in a densely collagenized stroma. The islands showed peripheral tall columnar cells with hyperchromatic nuclei resembling preameloblasts and the islands showed stellate reticulum like cells in the centre. The islands appeared compressed due to the hyalinised stroma, a feature characteristic of the desmoplastic ameloblastoma (Figure 3). The H and E stained section of the second case revealed ameloblastic folicies in a densely collagenized stroma. The islands showed hyperchromatic peripheral cells and central stellate reticulum like cells. Some islands showed squamatoid change of the central stellate reticulum like cells. This appearance was consistent with that of a
ameloblastoma with an acanthomatous change (Figure 4). Thus, the histological pictures of both cases are suggestive of Desmoplastic ameloblastoma and the second case was reported as a hybrid variant of Desmoplastic ameloblastoma owing to the acanthomatous change seen in the central cells.

![Image](image1.png)

Figure 1: Intraoral swelling with buccal and lingual expansion and radiograph showing a mixed radiolucency.

![Image](image2.png)

Figure 2: The intra oral swelling in relation to upper left premolar region.

**Discussion**

Desmoplastic ameloblastomas account for 4-13% of all solid multicystic ameloblastomas. The first detailed report was given by Eversole et al in 1984 who called it an ‘ameloblastoma with pronounced desmoplasia.’ The growing knowledge regarding the clinico-radiography as well as pathology of Desmoplastic ameloblastoma has led to its categorization as a distinct variant of ameloblastoma in the WHO classification of odontogenic tumors. DAs are similar to SMAs in gender and age distribution occurring in the average age of 42 years with high incidence in 3-5th decade and male predilection.

The anatomic location is rather unlike the conventional ameloblastoma which occurs most commonly in the posterior mandible. DA’s are seen most commonly in the anterior or premolar region with equal predilection for upper and lower jaw. Contrary to this report, some authors
suggest that DA’s have slightly higher rate of occurrence in the mandible. Sun et al. also suggested that the desmoplastic variants of ameloblastoma are usually smaller in size compared to the conventional ameloblastoma.

Desmoplastic ameloblastoma usually presents as a uni or multilocular mixed radiolucency with ill-defined borders. Sivapathasundaram et al. have stated that the mixed radiographic appearance could be due to the infiltrative process into the marrow spaces with simultaneous osteoblastic activity around remnant of original bone. Others have hypothesized that the mixed appearance on radiograph may be attributed to metaplastic foci. Takata et al attributed the ill-defined margins and mixed radiolucency to the infiltrative nature of the lesion.

Figure 3: The photomicrograph of case 1 showing the desmoplastic variant of ameloblastoma.

Figure 4: The photomicrograph of case 2 showing the desmoplasia in the connective tissue with ameloblastomatous islands.

Philipsen et al. stated that the radiographic appearance of DA is suggestive of an infiltrative potential with high tendency for recurrence. Thus, the pronounced
desmoplasia can be viewed as a defensive response of the host to an ‘aggressive’ tumor. The pattern of infiltration and lack of demarcation of fibrous connective tissue may be of prognostic significance in predicting tumor behaviour. Sun et al have reported that this radiographic appearance with ill-defined borders mimic fibrous osseous lesions or malignant tumors. The most prominent feature of the Desmoplastic ameloblastoma is the dense hyalinised moderately cellular fibrous connective tissue with areas of bone formation occasionally. The odontogenic islands are irregular and appear to be compressed or squeezed by the stromal tissue, giving a stretched out tail like appearance. Neither the palisaded peripheral tall columnar cells nor the inner stellate reticulum like cells are observed in DA. The centres of islands in DA appear hypercellular with spindle, squamatoid, occasionally keratinised epithelial cells. Such lesions are called ‘hybrid’ variants of ameloblastoma. The second case reported belongs to this variety.

Philipsen et al., have suggested that tumor islands induce proliferation of mesenchymal cells resulting in desmoplasia and metaplastic bone formation. This argument has not been accepted as the same inductive potential was not observed in SMA. Zhi Jun Sun et al., stated that the pronounced desmoplasia is a defensive response to the aggressive tumor. Some authors argue that desmoplasia can be a maturation change in a SMA, as similar dense collagenisation is seen during maturation of long standing tumors. The occasional osteoplasia has been suggested to be metaplastic change rather than a part of a reparative process.

Immunohistochemical studies state that desmoplasia originates from de novo synthesis of extracellular matrix proteins. Marked positivity with TGF beta has also been seen suggesting its role in matrix formation. High expression of caspase, Fas, p63 and decreased expression of CK19 has been shown by tumor cells. Desmoplastic stroma of DA’s has reported to show a strong positivity for collagen VI ruling out scar tissue. Immunonegativity for tenasin and strong immune-positive reaction for fibronectin and type I collagen. Kishino et al., demonstrated oxtalan fibres in the stromal tissues of one case, suggesting that the tumor had derived from the epithelial rest of malaseez in the periodontal membrane of a neighbouring tooth.

The infiltrative nature of the lesion demands a radical surgical approach, though surgical resection, enucleation and curettage have been performed, studies show higher recurrence rate with enucleation and curettage. The prognosis of this lesion is a question of doubt as authors suggest Desmoplastic ameloblastoma may exhibit aggressive behaviour as compared to other types of ameloblastoma. Some consider its recurrence rate to be as much as other variants of ameloblastoma. Kezler et al., have predicted higher recurrence rate and stated the possible causes as follows:

1. DAs are apt to be mismatched with fibro-osseous lesions. The accurate diagnosis of a DA is hard to achieve before the operation;
2. DA frequently present with ill-defined border making it difficult to investigate the exact interface of the lesion with normal bone;
3. The more common location in the maxilla may produce an early invasion of adjacent structure.

**Conclusion**

The Desmoplastic ameloblastoma is often regarded as a mere histological variant of ameloblastoma with ignorance about its biologic behaviour. Its unique behaviour, diverse from the conventional ameloblastoma or its other variants in regards to anatomic location, radiographic appearance, atypical histology and recurrence warrants a special attention to its diagnosis. Most often considered as a fibro-osseous lesion, its clinical behaviour is underestimated and inadequate treatment follows. Consideration of the Desmoplastic ameloblastoma as a distinct clinical as well as pathologic entity is the first step towards prevention of such mismanagement of the lesion.

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