

Desmoplastic ameloblastoma - a report of two clinical cases

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Abstract

Desmoplastic ameloblastoma is a relatively rare histological variant of ameloblastoma with specific clinical, radiological, and histological features. Although radiographic examination of ameloblastomas usually reveals unilocular or multilocular radiolucency, desmoplastic ameloblastoma may appear as a mixed radiopaque-radiolucent lesion resembling benign fibro-osseous lesions. Histologically, desmoplastic ameloblastoma is characterized by small nests and strands of "compressed" odontogenic epithelium supported by pronounced collagenized stroma. This report describes two cases of desmoplastic ameloblastoma in the anterior maxilla of a female patient and the anterior mandible of a male patient, mimicking clinically as an odontogenic cyst.

Keywords: ameloblastoma, desmoplastic, multilocular, anterior maxilla, anterior mandible.

Introduction

Ameloblastomas are tumors arising from the odontogenic epithelium and most commonly encountered odontogenic tumors. Despite their locally destructive nature, they are considered benign¹. Histologically, ameloblastoma has been classified as follicular, plexiform, acanthomatous, granular cell, desmoplastic, and basal cell¹⁻³.

In 1984, Eversole *et al.* discovered a new and unusual histologic variant known as desmoplastic ameloblastoma⁴, but only in 1992, the World Health Organization (WHO) recognized this entity as a variant of ameloblastoma³.

Till date, 145 cases of desmoplastic ameloblastoma have been reported in Japanese, Chinese, Malaysian, Western, and African populations, with very few cases described in Indians⁵. This report describes two cases of desmoplastic ameloblastoma in the anterior maxilla of a female patient and the anterior mandible of a male patient, mimicking clinically as an odontogenic cyst.

Case 1

A 47-year-old female patient came to the Department of Oral Medicine with complaints of swelling in the right anterior region of the upper jaw with 6 months of duration. She revealed a history of insidious onset as a small nodule, gradually reaching the present extent without any paresthesia, discharge but was associated with mild, intermittent type of pain. No other associated symptoms were reported by the patient.

Intraoral examination revealed a well defined, non-tender, hard swelling in the

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right anterior maxilla with intact mucosa causing bicortical expansion (Figure 1). The maxillary right canine was vital with grade I mobility. The provisional diagnosis of a fibro-osseous lesion of right maxilla was made.

Intraoral periapical radiograph revealed a diffuse, ill-defined, radiolucent lesion interspersed with radiopaque septae, producing a multilocular appearance with widening of the periodontal ligament space and loss of lamina dura of the right canine (Figure 2). Computed tomography (CT) (Figure 3) revealed that the lesion was extending on buccal and palatal aspects from the right lateral incisor to the right second molar, and the walls of the maxillary antrum were intact and not involved.

The overall clinical and radiographic features were suggestive of an odontogenic tumor, probably an ameloblastoma, with differential diagnosis of monostotic fibrous dysplasia, or adenomatoid odontogenic tumor.

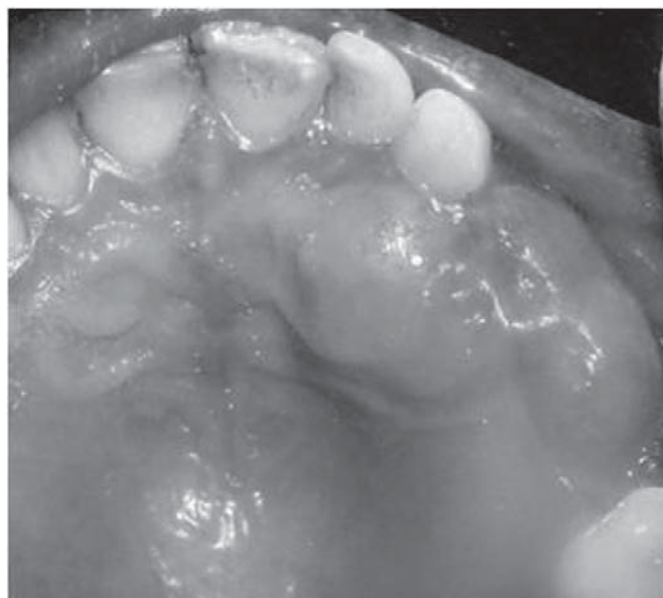


Fig. 1. Photograph showing bicortical expansion on maxilla with intact mucosa.



Fig. 2. Intraoral periapical radiograph showing multilocular appearance

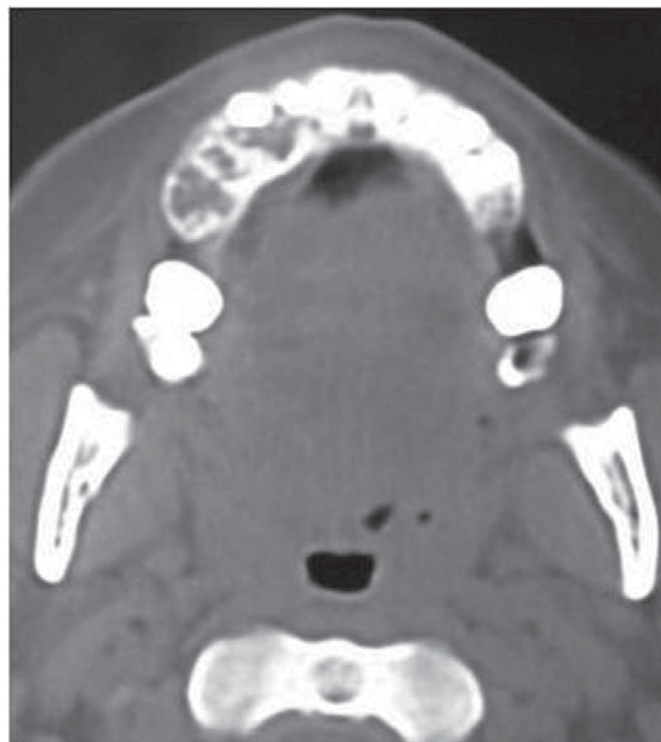


Fig. 3. Axial CT scan showing that the lesion extending on the buccal and palatal aspects from the right lateral incisor to right second molar.

Incisional biopsy was performed and the histopathologic evaluation of the specimen (Figure 4) showed irregular, bizarrely shaped odontogenic epithelial islands and cords in a moderately cellular fibrous connective tissue with abundant thick collagen fibers that appear to compress the odontogenic islands giving them a stellate or an “animal-like” configuration and the diagnosis of desmoplastic ameloblastoma was established.

Excision of the lesion with extraction of right lateral incisor and right canine was done, followed by reconstruction of the defect with buccal pad of fat and primary mucosal

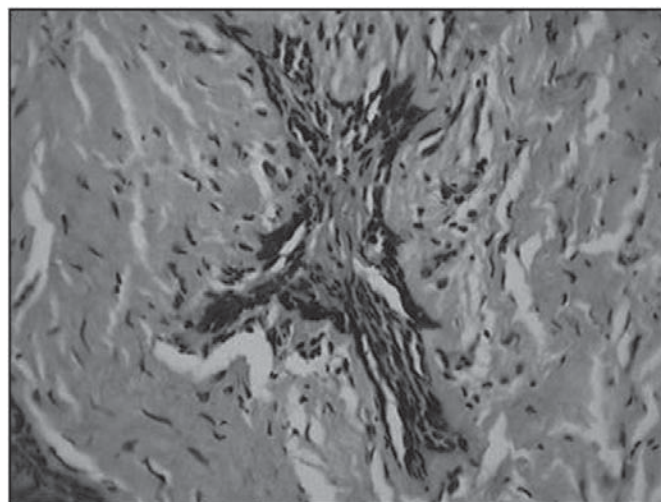


Fig. 4. Histological image showing irregular, bizarrely shaped odontogenic epithelial islands and cords in a moderately cellular fibrous connective tissue with abundant thick collagen fibers that appear to compress the odontogenic islands giving them a stellate or an “animal-like” configuration.

closure under general anesthesia with acrylic splint placement on maxillary arch. Patient was recalled after one week, showing ongoing healing of the site. The patient was reviewed after 1 month, and complete healing of the lesion was noticed. The patient was kept on periodic recall every 6 months during 1 year. However, she could not keep up the recall appointments.

Case 2

A 37-year-old male patient reported with complaints of swelling in the anterior region of the mandible for two months. Patient had history of trauma to this region (biting on hard food), after which he noticed the swelling a week later. Swelling was initially smaller in size and gradually progressed to the present size. There was no associated pain, pus discharge, paresthesia or other symptoms.

Intraoral examination revealed a well defined solitary, non-tender swelling in right mandibular anterior region crossing midline and causing bicortical expansion (Figure 5). On palpation, swelling was soft to firm in consistency with areas of erosion noticed labially with intact mucosa. The mandibular right and left central incisor and mandibular right lateral incisor were vital with grade II mobility.

Panoramic radiograph (Figure 6) showed multilocular lesion involving left parasymphysis area causing root

resorption of left canine and premolars. The anterior extension of the lesion couldn't be traced out because of superimposition of cervical spine. Mandibular occlusal view showed bicortical expansion and thinning of the buccal cortex (Figure 7). Intraoral periapical radiograph (Figure 8) revealed an ill-defined multilocular radiolucency superiorly causing destruction of alveolar crestal bone with respect to left mandibular lateral incisor and mandibular canine. The inferior of the radiolucency showed scalloping and the interior of the radiolucency showed remnants of trabeculae. There was displacement of the roots of left mandibular lateral incisor and mandibular canine.

The provisional diagnosis of odontogenic cyst was proposed with differential diagnosis of traumatic bone cyst, odontogenic tumor, and central giant cell granuloma.

Aspiration yielded blood tinged straw colored fluid. Incisional biopsy was performed and the histopathologic evaluation of the specimen (Figure 9) showed dense collagen stroma with odontogenic epithelial cells arranged in the form of long, thin strands that gives an animal-like pattern configuration. The epithelial cells were hyperchromatic. This proliferation seemed to be compressed by a dense stroma, and a final diagnosis of desmoplastic ameloblastoma was established.

Peripheral osteotomy of anterior mandible with removal from the mandibular right first molar to the left lateral incisor was done under general anesthesia, and the patient was put on regular periodic recall check up till date (Figure 10). The patient has currently been on follow up for the past 8 months.

Discussion

In the WHO classification, desmoplastic ameloblastoma is considered as a rare variation of ameloblastoma. It accounts for 4% to 5% of all ameloblastoma⁶. A retrospective study was done to correlate the clinical and radiographic features of 115 cases of desmoplastic ameloblastoma reported in



Fig. 5. Photograph showing bicortical expansion on anterior mandible with intact mucosa

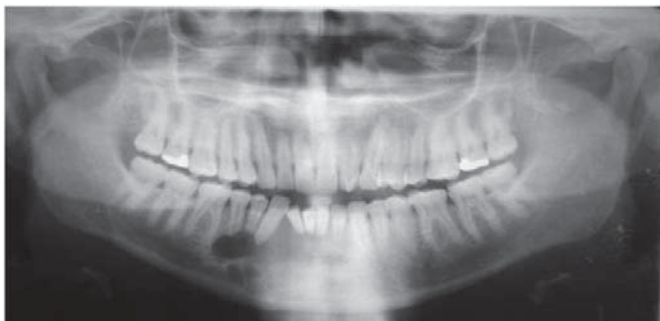


Fig. 6. Orthopantomogram showing multilocular lesion with root resorption of left mandibular premolars and canine.



Fig. 7. Mandibular occlusal view showing thinning of buccal cortex

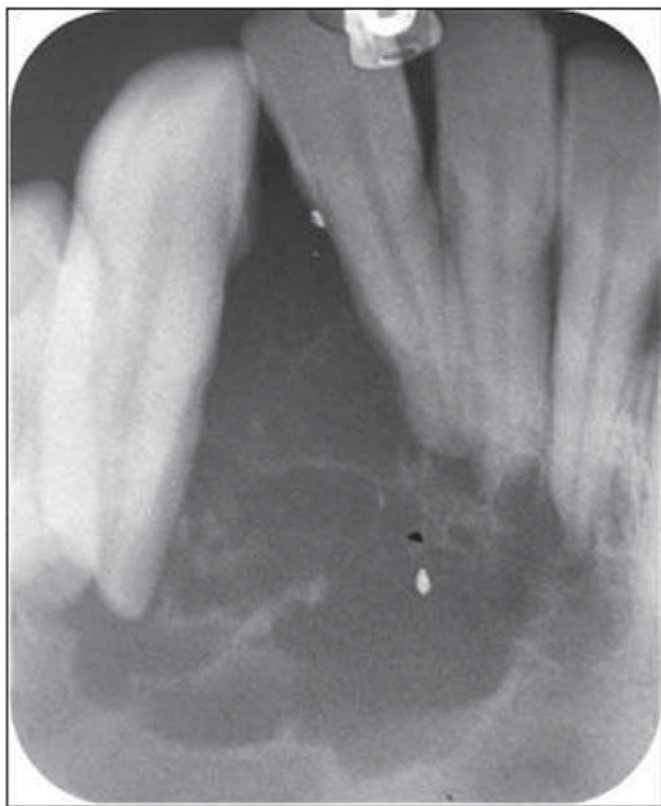


Fig. 8. Intraoral periapical radiograph showing multilocular appearance with divergence of the roots.

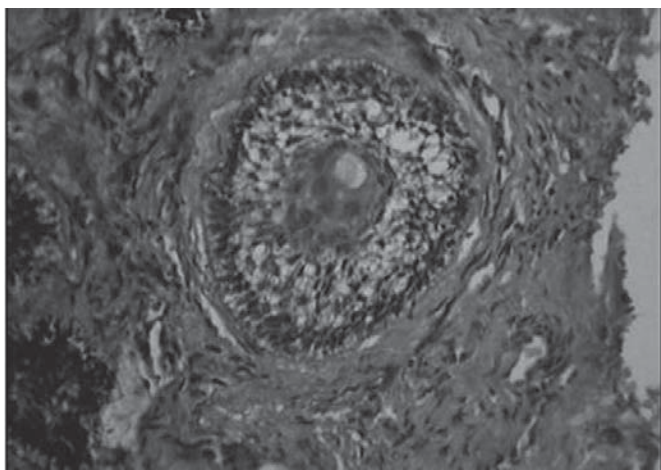


Fig. 9. Histological image showing dense collagen stroma with odontogenic epithelial cells arranged in the form of long, thin strands, giving an animal-like pattern configuration. The epithelial cells are hyperchromatic. These proliferations seem to be compressed by dense stroma.

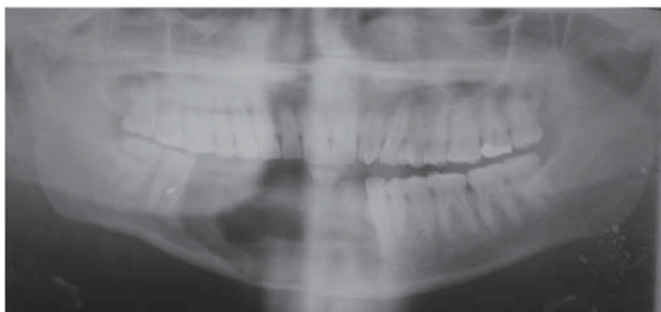


Fig. 10. Postoperative orthopantomogram showing the wide excision of the lesion.

literature from 1984 to 2008, and concluded that it presents distinct clinical, radiographic and histologic features when compared to “conventional ameloblastomas”⁷.

This entity occurs most commonly in the third to fifth decades of life, with an equal male to female ratio^{2,5}. Our cases also showed the same age and sex predilection. More than 70% of the desmoplastic ameloblastoma arose within the anterior or premolar regions of the jaws as seen in our both cases, and roughly half of the tumors occurred in the maxilla^{1,8-9}. Clinically, maxillary lesions are more dangerous than mandibular ones as they can invade the adjacent sinus and orbit and involve vital structures. Additionally, the thin maxillary bone is a weak natural barrier for tumors as compared to the thicker mandible¹⁰.

Radiologically, the desmoplastic variant exhibits atypical and varied radiographic features as: localized irregular multilocular radiolucency with indistinct borders as seen in our second case, or a radiopaque/ radiolucent appearance with ill defined margins similar to fibro-osseous lesion as seen in our first case, or a massive expansile osteolytic lesion with honeycomb, mottled or multilocular appearance^{4,8}. Tooth displacement was a common feature for Desmoplastic ameloblastoma which was seen in our second case. Root resorption was discovered in only 33% of the cases which was seen in our both cases⁸.

CT scan can delineate the internal structure of the lesion more accurately and is particularly helpful in determining its margins and extension into adjacent structures¹⁰. MRI shows heterogeneous low to intermediate signal intensity on T1W images, heterogeneous high signal intensity on T2W images, and strong enhancement on post-gadolinium T1W images¹¹.

Histologically, it is characterized by abundant collagenous proliferation of the stroma, loss of cell rich connective tissue, absence of capsule, and existence of some ameloblastoma like structures with peripherally compressed ameloblasts^{4,12}. A few cases of hybrid lesion of ameloblastoma have been reported in the literature in which histologically, areas of follicular and plexiform patterns are found together with characteristic desmoplastic areas. This type of variant was first described by Waldron².

The tumor cells of desmoplastic ameloblastoma have shown positive immunoreactivities for cytokeratin (CK), CK 8, 13, 19, filaggrin and ameloblastoma antibodies, retaining the odontogenic epithelial characteristics¹³.

Various facts about this lesion may suggest aggressiveness: a potential to grow to a large size; the common location in the maxilla that may produce an early invasion of adjacent structures; the diffuse radiographic appearance. Finally, it is almost impossible to find the exact interface of the lesion with normal bone, making it especially difficult to treat surgically⁹.

Enucleation or curettage alone of the lesion may lead to recurrence¹⁴, as there is indistinct boundary between the tumor and normal tissue. Therefore complete resection and regular follow up is recommended⁴.

In conclusion the desmoplastic ameloblastoma reveals unique radiological and histological features. Further investigation of a larger number of more cases must be carried

out in an attempt to predict the behavior and prognosis of this entity.

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