Case Report

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Desmoplastic Ameloblastoma: A Rare Case Report

Anish Tiwari[1*] Rina Tiwari[2] Nirav Patel[3] Ravi Kalola[4] Adarsh Desai[5]

[1]Professor and Head, Department of Oral and Maxillofacial Surgery, Goenka Research Institute of Dental Science, Gandhinagar, Gujarat, India.
[2]Private Consultant, Department of Oral and Maxillofacial Pathology, Ahmedabad, Gujarat, India.
[3]Reader, Department of Oral and Maxillofacial Surgery, Goenka Research Institute of Dental Science, Gandhinagar, Gujarat, India.
[4]Reader, Department of Oral and Maxillofacial Surgery, Goenka Research Institute of Dental Science, Gandhinagar, Gujarat, India.
[5]Reader, Department of Oral and Maxillofacial Surgery, Goenka Research Institute of Dental Science, Gandhinagar, Gujarat, India.

ABSTRACT

Background: Among the ameloblastomas, the desmoplastic variation is rare. The desmoplastic ameloblastoma (DA) is characterized by specific clinical, imaging, and histological features. DA showed a nearly equal male to female ratio (55/59) with a prevalence within the forth and fifth decades. Sixty-two lesions occurred in the mandible and fifty-one lesions in the maxilla. Clinically, a painless swelling with buccal extension was the most common presentation being found in 48 cases. Histologically, scattered epithelial nests and extensively desmoplasia were prominent features of DA. In conclusion, these retrospective results confirm the statement that DA is a variation among ameloblastomas. DA present clinicoradiographic and histologic distinct features, when compared with "conventional ameloblastomas".

Keywords: Desmoplastic ameloblastoma, Mandible, Maxilla, Histology, Radiology, Treatment.

INTRODUCTION

Ameloblastoma is the second most common odontogenic tumour. It arises from odontogenic epithelium and is known for its distinct aggressive clinical behaviour and characteristic histologic picture. Based on clinical and radiographic characteristics, histopathology, behavioural and prognostic aspects, the subtypes of ameloblastoma are classic solid/multicystic, unicystic, peripheral and desmoplastic types[1]. The first detailed report in English literature on desmoplastic ameloblastoma (DA) was given by Eversole in 1984, who called it an "ameloblastoma with pronounced desmoplasia[2]."

DA is a rare variant of ameloblastoma accounting for approximately 4-13% of ameloblastomas[3]. It is characterized by an unusual histomorphology, including extensive stromal collagenization or desmoplasia. A characteristic feature is an almost equal distribution in location between the maxilla and mandible. Radiologically, the desmoplastic variant exhibits atypical and varied radiographic features such as: Localized irregular multilocular radiolucency with indistinct borders, or a mixed radiopaque-radiolucent appearance with ill-defined margins similar to fibro-osseous lesion, or a massive expansile osteolytic lesion with honeycomb, mottled or multilocular appearance[4]. Tooth displacement is a common feature. DA may be defined as a benign but locally invasive variant consisting of proliferating, irregular, often bizarrely shaped islands and cords of odontogenic epithelium of varying sizes embedded in a desmoplastic connective tissue stroma. Formation of metaplastic bone trabeculae (osteoplasia) rimmed by active osteoblasts has been described in a few cases[5]. A possible transitional form of DA, showing microscopic features of desmoplastic variant together with areas of classical follicular/plexiform ameloblastoma has been described as a "hybrid lesion[6]."

CASE REPORT

A 45-year-old female presented to the Department of Oral and Maxillofacial surgery, Goenka Institute of Dental Sciences and Research, Gandhinagar,

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***Correspondence Dr. Anish Tiwari.**
Department of Oral and Maxillofacial Surgery, Goenka Research Institute of Dental Science, Gandhinagar, Gujarat, India.
Email: dr.anish.ti@gmail.com

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Gujarat, India with an asymptomatic swelling of spontaneous origin in her left maxilla. The lesion originated one year ago. There had been gradual increase in the size of swelling to its present size. There was absolutely no pain, bleeding or pain. She also gave no history of trauma or any non-relevant medical history. The physical examination revealed facial asymmetry due to swelling on the left side of face which was oval in shape and had a smooth surface. The skin over the swelling appeared normal. It was not tender on palpation.

The intraoral examination disclosed a large mass, approximately 3.5 × 2.5 cm in size, extending from the left canine to the left Second premolar buccally as well as palatally. The swelling was bony hard in consistency, nonfluctuant, nonreducible, noncompressible and nonpulsatile.

Radiographic examination of the maxilla revealed a diffuse ill-defined mixed radiolucency with some areas of radiopacity, extending from the distal surface of the left canine region to the mesial surface of the left second premolar, with an approximate size of 3.3 × 2.3 cm (Figure 1). Floor of maxillary sinus appeared to be intact. Based on the clinical and radiographic appearance, a provisional diagnosis of odontogenic myxoma was made. Aspiration of the lesion was non-productive and a complete blood picture showed values within the normal limits. An incisional biopsy was performed under local anaesthesia for a definitive diagnosis. Histologically, the features were consistent with those of desmoplastic ameloblastoma (Figure 2).

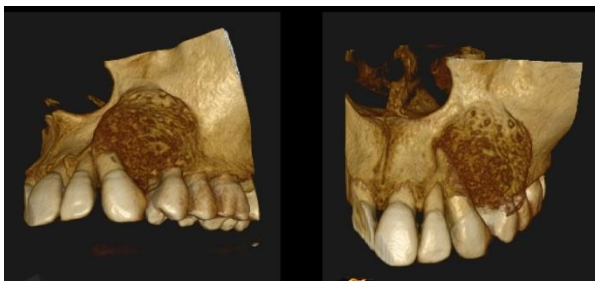

Fig 1: Radiograph shows the extent of the lesion.

A partial maxillectomy for tumour resection was performed (Figure 3) under general anaesthesia and a transitional maxillofacial prosthesis was placed. The resected specimen was 3 × 2 cm in dimension and consisted of three teeth and it was again sent for histological observation.

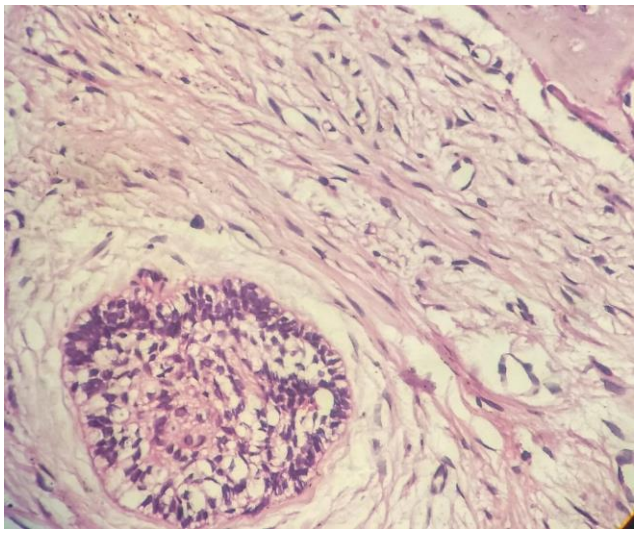

Fig 2: Histological specimen showing desmoplastic features.

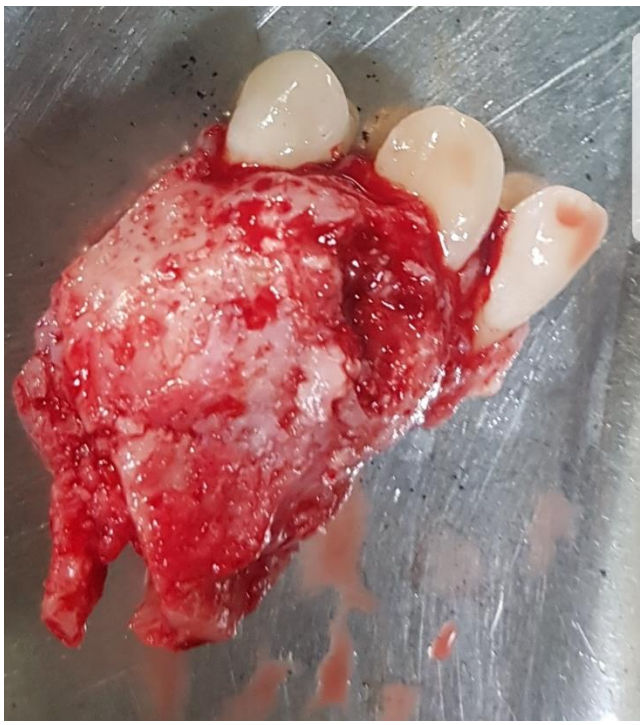

Fig 3: Resected part of maxilla after surgery.

DISCUSSION

Desmoplastic ameloblastoma (DA) continues to remain one of nature's secrets. Till date, less than 150 have been diagnosed with DA. The most common clinical manifestation is painless bony swelling. Desmoplastic ameloblastoma constitutes 0.9% to 12.1% of all ameloblastomas. The mean age at initial presentation is 42.3 years (range 17-70 years) with age and gender predilection similar to

that of other ameloblastomas[7]. The incidence of DA among ameloblastomas ranges from 0.9% to 12.1% in different races. Concerning the race, 69 out of 115 cases DAs with detailed description, of which 25(36.2%) were Japanese, 15(21.7%) were Chinese and 10(14.5%) were Indian. Data from different geographical regions seem to suggest a biogeographical pattern in that the relative frequency of DA is slightly higher in Asian population. However, more systematic studies on DA are needed to verify such suggestions[6, 8, 9]. DA is reported to occur in the anterior or premolar region of either maxilla or mandible. This is in contrast to the conventional ameloblastoma which is found predominantly in the posterior region of mandible.

The radiographic features of DA also differ from the conventional ameloblastoma. A wide range of appearance from radiolucency to radiolucency with flecks of opacity to mixed radiolucent/radio-opaque has been reported[10]. A wide range of appearance from radiolucency to radioluceny with flecks of opacity to mixed radiolucent/radio-opaque has been reported. Conventional ameloblastoma usually presents as a well-defined unilocular/multilocular radiolucency. Again, the borders of DA are usually poorly defined. DA usually infiltrates into marrow spaces and surrounding non-neoplastic bone without a fibrous capsule[11]. Furthermore, a case with associated follicular, plexiform, acanthomatous and basaloid changes has been documented[12]. Cystic degeneration within the tumor has also been described.[14] In our case, few islands with kite-like appearance with spindle cells in the interior of the island and stromal desmoplasia were noted[13].

Extensive stromal desmoplasia is a striking finding with a tendency to squeeze or compress the odontogenic islands at the periphery. The mechanism of desmoplasia is not understood till date. Numerous immunohistochemical studies have shown DA tumor cells as showing variable expression of S-100 protein and desmin. There is also high expression of caspase-3 and Fas. Sometimes decreased expression of cytokeratin 19 and high expression of p63 is also observed[14, 15]. Many a times Intense staining for collagen Type IV adjacent to tumor islands has been observed in DA[3]. This indicates active synthesis of extracellular matrix proteins and that the stroma is not scar tissue.

The biologic behaviour of the lesion is still Confusing. There are lots of Conflicting reports regarding the recurrence because of paucity of adequate samples. The radiographic picture suggests an infiltrative process with the propensity to recur and the stromal reaction may be viewed as a defensive response of the host to the aggressive tumour.

CONCLUSION

Moreover, many vital structures lie close to the lesion and thin cortical bone is a very fragile barrier through which tumour frequently infiltrates into marrow spaces. Hence sharp demarcating border is difficult to identify. The radiological and histological findings of poor encapsulation and ill-defined borders warrants a long-term follow-up. A radical approach to its treatment is recommended as in a case of conventional ameloblastoma.

CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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