J. Int Oral Health 2011

Hybrid lesion of desmoplastic and conventional Ameloblastoma – A Case Report

BG Harsha Vardhan* Ekta Ingle† K Dornadula‡ K Saraswathi Gopal§

*M.D.S, Associate Professor, †Post Graduate Student, ‡M.D.S, Professor, §M.D.S, Professor and Head, Department of Oral Medicine and Radiology, Meenakshi Ammal Dental College, Chennai.
Contact: ektaingle@ymail.com

Abstract:

Desmoplastic ameloblastoma is an unusual variant of ameloblastoma with specific radiographic and histologic features and anatomic distribution differing from those in conventional types. In this article, we present a patient with desmoplastic and conventional variant (hybrid lesion) localized in the posterior part of mandible. The radiographic features presented with a well defined, irregular, multilocular lesion extending from the canine on the left side till the ascending ramus on the right side. A mandibular segmental resection was performed followed by transport distraction osteogenesis. The biologic profile of this tumor is not fully understood because of its complexity and limited numbers of cases in the medical literature, coupled with inadequate long term follow up.

Key words: Ameloblastoma, Desmoplastic, Hybrid lesion, Mandible .

Introduction:

Odontogenic tumors represent a spectrum of lesions ranging from benign and malignant to dental hamartomas, all arising from odontogenic residues, i.e. odontogenic epithelia or ectomesenchyme with variable amounts of dental hard tissues in the same sequence as in normal tooth development[1]. Ameloblastomas are the most common, clinically significant odontogenic neoplasms affecting the jaws[2]. Follicular and plexiform patterns are the most common histological

P- ISSN
0976 – 7428

E- ISSN
0976 – 1799

Journal of International Oral Health

Oral Medicine and Radiology

Case Report

Received: Dec, 2010
Accepted: Feb, 2011

Bibliographic listing:
EBSCO Publishing
Database, Index Copernicus, Genamics Journalseek Database, Proquest, Open J Gate.

variants, less common cellular variants include basal cell, clear cell, unicystic, desmoplastic, keratoameloblastoma and papilliferous keratoameloblastoma[3].

A desmoplastic variant with features of other histological types is termed as a "Hybrid" lesion. The hybrid lesion of desmoplastic ameloblastoma and conventional ameloblastoma is an unusual variant that was first described by Waldron and El – Mofty in 1987[4]. This lesion shows typical areas of follicular or plexiform patterns together with microscopic features of desmoplastic ameloblastoma which are characterized by pronounced desmoplastic stroma. To our knowledge, less than 10 cases of hybrid lesion have been published in medical literature[4-6]. The clinicopathological and biological characteristics of this lesion have not been clearly established. In our article, we add a case of hybrid tumor to share our experience of this interesting lesion.

Case Report:

A 29 year old woman reported to our department with a complaint of swelling with pain in the right side of face since one week (Fig.1). History dates back to 5 years, when she noticed a small swelling which progressively increased to reach the present size. An incisional biopsy and extraction of two teeth in the same region was carried out 3 years ago, following which the swelling subsided, but reappeared in six months. The histopathology gave an impression of an odontogenic cyst.

Clinical examination revealed a single, well defined swelling in the right lower half of the face roughly measuring about 7 x 6 cm extending medially from the corner of mouth on left side, crossing the midline, to laterally, the angle of mandible. A line drawn superiorly between the angle of mouth to the tragus of the ear and inferiorly about 3cm below the lower border of mandible. Skin over the swelling appeared smooth with obliteration of right nasolabial fold. The swelling was hard in consistency with no signs of paraesthesia.

Intraorally, the swelling measured about 5 x 2 cm from the mesial aspect of 41 to the mesial aspect of 47 with marked obliteration of the buccal vestibule. Expansions of both buccal and lingual cortical plates were evident with lingual displacement of 44 (Fig.2). Indentations of the opposing teeth were well appreciated on the surface of the swelling with areas of erythema. 44 and 47 showed negative response to vitality test. Following clinical examination, the patient was subjected to plain film radiography. IOPA of 43, 44, 45 and 46 region revealed complete absence of normal trabeculae with presence of multilocular radiolucency with irregular septae. Resorption of apical portion of 43 and 44 were evident (Fig.3). Mandibular occlusal revealed a single multilocular radiolucency extending from mesial aspect of 33 to distal aspect of 47 with marked buccal and mild lingual expansion, traversed by thin, irregular and discontinuous septae and lingually displaced 44. The normal trabecular pattern was present in the anterior part of the lower border of mandible and completely absent as the lesion progressed posteriorly (Fig.4). Panoramic radiograph revealed a well defined, irregular, multilocular radiolucency extending from 33 region to the mesial aspect of 48 traversed by coarse, curved septae. The buccal cortical margin appeared thin with discontinuity on the lingual side. The degree of radiolucency was variable within the lesion, appeared more cystic in the anterior aspect and less radiolucent as the lesion extended posteriorly. Loculations of varying sizes with septae of different thickness were seen within it. Scalloping was well appreciated in the anterior part when compared to the posterior part of the lesion. Mental foramen was not evident on right side, however, the path of mental nerve was visible till the mesial aspect of 48 (Fig.5).

USG of the lesion exhibited an expansile lytic heterogenous area of 4 x 3 cm with cystic and echogenic solid component with multiple internal septations. Computed Tomography showed a well circumscribed, multilocular

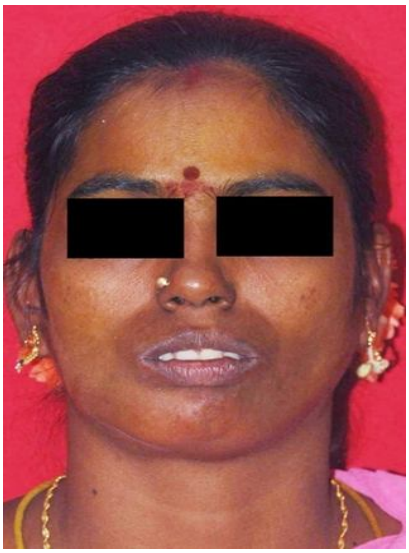

Fig. 1: Clinical photograph showing mild asymmetry on the right side

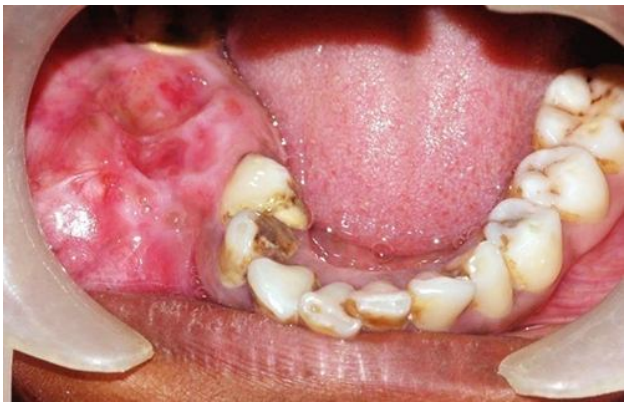

Fig. 2: Intraoral lesion with indentations of upper molar teeth and obliteration of the vestibule.

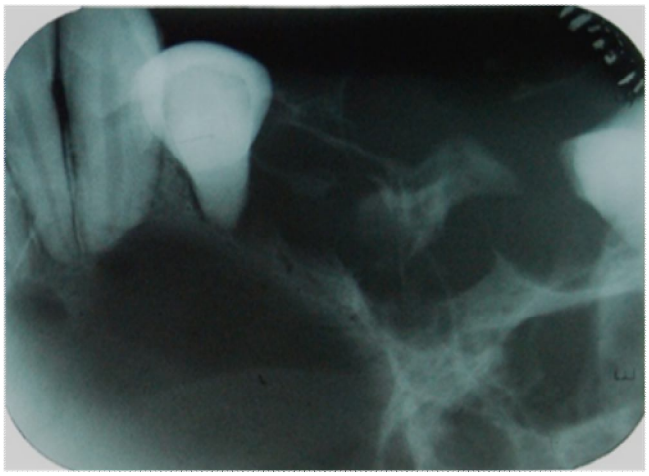

Fig. 3: IOPA in relation to 43, 44, 45 & 46 with multilocular radiolucency.

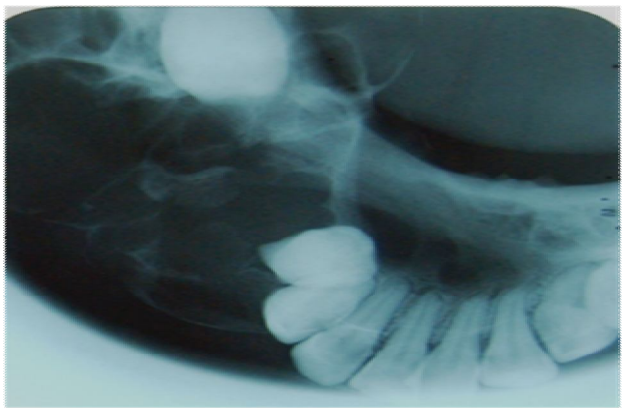

Fig. 4: Mandibular occlusal with buccal plate expansion

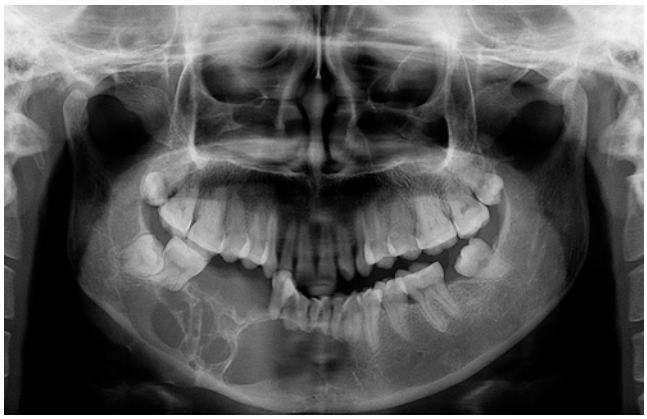

Fig. 5: Orthopantomograph

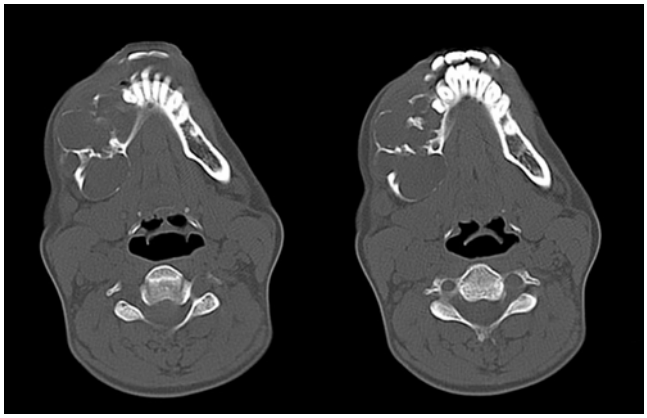

Fig. 6: Axial Images of CT mandible showing expansile multilocular hypoattenuating lesion on the right side with sclerotic changes and displacement of teeth and adjacent structures.

expansile lytic lesion measuring 5 x 4.2 cm with thin septations and a narrow zone of transition involving the body of mandible on right side. CT also confirmed the break in the inner cortex of the mandible (Fig.6,7). Aspirate from the

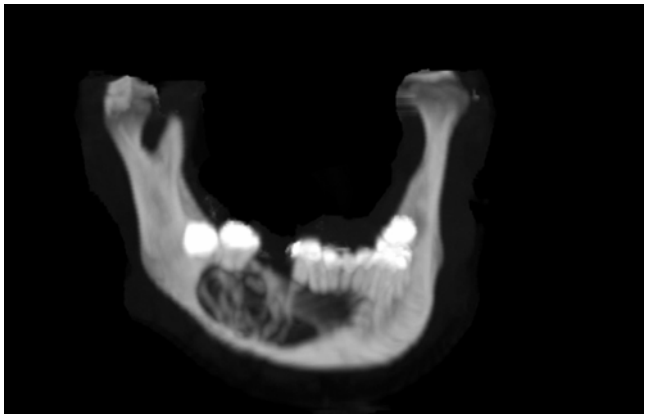

Fig. 7: 3D Image of mandible showing sclerotic changes and displacement of teeth.

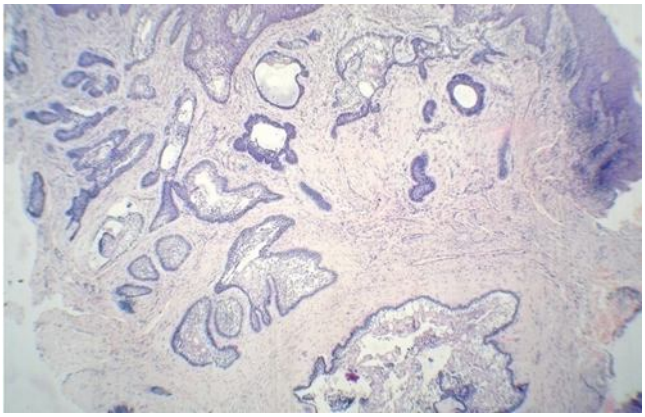

Fig. 8: Photomicrograph showing odontogenic epithelial islands of varying shapes and sizes and some areas of cystic degeneration. (hematoxylin and eosin stain , x 10)

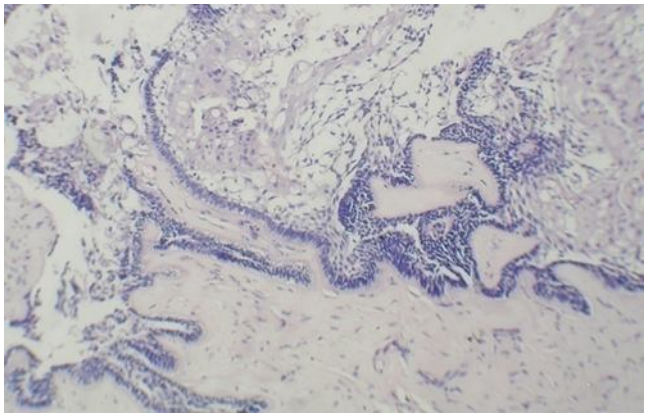

Fig. 9: Photomicrograph showing tall columnar cells and loosely arranged stellate reticulum like cells. (hematoxylin and eosin stain , x 40)

anterior part of lesion yielded a viscous, yellowish white material tinged with blood.

Based on the history, clinical and investigative procedures, a provisional diagnosis of ameloblastoma involving the right mandible was made. With an informed consent, incisional biopsy was performed and the soft tissue section showed odontogenic epithelium supported by a mature connective tissue stroma. The epithelium was seen in the form of islands which were of varying size and shape, lined by a peripheral layer of tall columnar cells with hyperchromatic nuclei exhibiting reversal of polarity. These cells were enclosing a central mass of loosely arranged polyhedral cells resembling the stellate reticulum. Micro and macrocyst formation were seen in these central areas indicating degeneration of the stellate reticulum like cells. In some areas, these central cells showed squamous metaplasia with keratin formation. In one area, the odontogenic epithelium were arranged in the form of cords and strands. The section also showed hyalinization of connective tissue stroma with compressed odontogenic epithelium. The supporting connective tissue stroma were predominantly fibrous with thick collagen fibres arranged irregularly, lined by spindle shaped fibroblasts. The stroma also showed few areas of odontogenic epithelial cells (Fig.8,9). The biopsy was diagnostic of a Hybrid tumor - desmoplastic and conventional ameloblastoma.

Under general anaesthesia, a mandibular segmental resection / hemimandiblectomy was performed followed by transport distraction osteognesis. The patient s postoperative period was uneventful. Postoperative radiographs and clinical follow up examination disclosed no reccurence or residual tumor.

Discussion:

Desmoplastic ameloblastoma (DA) is a benign, locally infiltrative epithelial neoplasm believed to be a variant or subtype of solid / multicystic ameloblastoma (SMA)[7]. Desmoplastic Ameloblastoma was first described by Tussole et al in 1984 and reviewed by Waldron and El-Mofty in 1987 and Reichart et al in 1995. In 1992, the World Health

Organisation (WHO) regarded it as a variant of ameloblastoma. Unlike in our case, Desmoplastic Ameloblastomas are most likely to occur in the anterior or premolar region of the jaws. A characteristic feature is an almost equal distribution in location between the maxilla and mandible. The size of the tumor varies between 1.0 and 8.5cm at its greatest diameter. The age range of patients varies between 17 and 72 years (n=72) at the time of diagnosis, the overall mean age being 42.8 years (men=42.9, women = 40.3years)[7]. The incidence of DA is relatively low, ranging between 0.9% - 12.1% of all the ameloblastomas that have been reported.[4,5,8-10]. Geographic distribution suggests that the tumor has been prevalent among Chinese, Japanese, Malaysians and Afro – Carribbeans[8].

The biological behaviour of DA still remains unresolved, it is generally agreed that this tumor is a variant of SMA. Oxytalan fibers have been identified and isolated in the stromal tissue of one of the cases reported by Kawai et al[14]. This finding is a possible indication of this tumor derived from the epithelial rests of Malassez in the periodontal membrane of the related tooth / teeth. Higuchi et al postulated that ameloblastoma in the tooth bearing area has a greater tendency to have an abundant stroma and to be desmoplastic. Genetic or biologic differences of the bone due to the anatomical sites, such as alveolar bone proper of odontogenic origin versus body of the jaw bone of non odontogenic origin, may be attributed to the histopathological variation of ameloblastomas[5].

The radiographic features of this tumor differ in most cases from those of other variants of ameloblastoma. The radiographic appearance indicates a mixed radiolucent - radiopaque lesion[7]. Panoramic radiograph in our case revealed a variable degree of multilocular radiolucency, being more cystic in the anterior aspect and less radiolucent as the lesion extends posteriorly. This variable presentation can suggest a preoperative diagnosis of a fibro-osseous lesion. The radiographic presentation may also suggest that the tumor may be more aggressive than other variants of ameloblastoma[11]. Takata et al also believed that the mixed pattern expresses the infiltrative nature of the tumor. As the DA infiltrates the bone narrow spaces, remnants of the original non - metaplastic or non-neoplastic bone remain in the tumor tissue. The ill - defined borders may also be attributed to the infiltrative behaviour of the tumor[15]. In a reported case of DA, there were prominent osteoplasia, which explained the mixed radiolucent - radiopaque appearance in the tumors[12].

Histopathological interpretation of coexistence of DA and the conventional variant in the "Hybrid" lesion is a matter of conjuncture. It is not clear whether desmoplastic change occurs secondary in the stroma of a pre-existing solid or multicystic ameloblastoma, whether a part of primary desmoplastic ameloblastoma alters into conventional ameloblastoma or whether the hybrid lesion is a kind of collision tumor[12]. Waldron and EL-Mofty described the histological appearance of DA as small ovoid islands and narrow cords of odontogenic epithelium widely separated by dense, moderately cellular and fibrous connective tissue. Columnar cells with reversed polarity within the epithelial islands are also seen, however, they are not a dominant feature. On the contrary, in our case, this finding was predominant. Spicules of mature lamellar bone trabeculae have been seen in intimate contact with the tumor. This finding may indicate the potential for local invasion and diffuse radiographic presentation[4]. The solid desmoplastic tumor may, on occasions undergo a partial morphological alteration to the morphology of the more conventional form of ameloblastoma[12]. Desmoplasia of the stromal connective tissue in DA can be argued to be a maturation of SMA, as similar dense collagenization are seen during maturation of long standing tumors. This arguement can be supported by the existence of Hybrid tumors

wherein the follicles are present in a desmoplastic background. The location of the tumor can influence the maturity of the lesion and, hence, the tumors in anterior jaws may mature sooner than those in the posterior mandible[13].

The prognosis of hybrid lesions has not been clearly established. It is generally described that most cases of DA have ill-defined margins and may have a propensity to recur, atleast as conventional ameloblastomas. As the biological behaviour, radiographic and histological features of the Hybrid lesion are still not fully understood, the lesion will remain enigma until workers persue a more definite tumor analysis, follow up and tracking many more cases into the medical literature.

References:

1. Shafer, Hine, Levy. Shafer's text book of oral pathology. *6th edition. Elsevier, 2009: 275.*
2. Waldron CA. Odontogenic Tumors. In: Oral and Maxillofacial Pathology. Neville BW, Damm DD, Allen CM, Bouquot JE, editors Philadelphia: W.B. Saunders Company 2002: 611.
3. Rajeev Sivasankar Pillai, Ravikiran Ongole, Auswaf Ahsan, Raghu Anekal Radhakrishnan, Keerthilatha Muralidhar Pai. Recurrent Desomplastic Ameloblastoma of the maxilla: A Case Report. *J Can Dent Assoc 2004; 70(2): 100-4.*
4. Waldron CA, EL-Mofty SK. A histopathologic study of 116 ameloblastomas with special reference to the desmoplastic variant. *Oral Surg. Oral Med. Oral Pathol. 1987; 63: 441-51.*
5. Higuchi Y, Nakamura N, Ohishi M, Tashiro H. Unusual ameloblastoma with extensive stromal desmoplasia. *J. Carniomaxillofac. Surg. 1991; 19(7): 323-7.*
6. Phillipsen HP, Ormiston IW, Reichart PA. The desmo-and osteoplastic ameloblastoma. Histologic variant or clinicopathologic entity? Case reports. *Int. J. Oral Maxillofac. Surg. 1992; 21: 352-7.*
7. Peter A Reichart, Hans P Philipsen. Odontogenic Tumors and Allied lesions. *Quintessence publishing company. 2004: 69-71.*
8. Lam KY, Chan AC, Wu PC, Chau KY, Tideman H, Wei W. Desmoplastic variant of ameloblastoma in Chinese patients. *Br J Oral Maxillofac Surg 1998; 36(2): 129-34.*
9. Ng KH, Siar CH. Desmoplastic variant of ameloblastoma in Malaysians. *Br J Oral Maxillofac Surg 1993; 31(5): 299-303.*
10. Raubenheimer EJ, Van Heerden WF, Noffke CE. Infrequent clinicopathological findings in 108 ameloblastomas. *J Oral Pathol Med 1995; 24(5): 227-32.*
11. Tanimoto K, Takata T, Suei Y, Wada T. A case of desmoplastic variant of mandibular ameloblastoma. *J Oral Maxillofac Surg. 1991; 49: 94-7.*
12. Takashi Takata, Mutsumi Miyauchi, Ikuko Ogawa, Ming Zhao, Yasusei Kudo, Sunao Sato, Toshitsugu Takekoshi, Hiromasa Nikai, Keiji Tanimoto. So-called 'hybrid' lesion of desmoplastic and conventional ameloblastoma: Report of a case and review of the literature. *Pathology International 1999; 49: 1014-8.*
13. Ismail Yazdi, Maryam Seyedmajidi, Ramin Foroughi. Desmoplastic Ameloblastoma (a Hybrid variant): Report of a Case and Review of the Literature. *Arch Iranian Med 2009; 12(3): 304-8.*
14. Kawai T, Kishino M, Hiranuma H, et al. A unique case of desmoplastic ameloblastoma of the mandible: Report of a case and brief review of the English language literature, *Oral Surg. Oral Med. Oral Patho. Oral Radiol. Endo. 1999; 87: 258-63.*
15. Takata T, Miyauchi M, et al. Clinical and histopathological analysis of Desmoplastic ameloblastoma, *Pathol Res Pract 1999; 195: 669-75.*

Source of Support: Nil

Conflict of Interest: No Financial Conflict