

An Unusual Case of Desmoplastic Variant of Ameloblastoma in the Anterior Maxilla – A Case Report

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Case Report

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Abstract: Desmoplastic ameloblastoma (DA) is a rare subtype of ameloblastoma, exhibiting eloquent variation in its clinical, radiological and histologic appearance. In 2017 WHO classification of a head and neck tumour has further modified this entity and titled it as a histologic variant of conventional ameloblastoma. A tumour mimics benign fibro-osseous lesion for being often prevailing in the anterior region of jaws as a mixed radiopaque-radiolucent lesion. We present a case of desmoplastic ameloblastoma in a 55-year-old male with a painless swelling in the anterior region of right maxilla. Ct scan shows a mixed lesion with multilocular appearance. An incisional biopsy verified it to be a case of desmoplastic ameloblastoma. Partial maxillectomy was performed on the right side. The patient is on routine follow-up with no recurrence. The present case deserves emphasis because of its unfamiliar appearance, potentially aggressive nature and deceptive radiologic appearance maximizing the chances of misdiagnosis. So, the clinician should be alert enough to include desmoplastic ameloblastoma in the differential diagnosis of any lesion/growth with mixed radiolucent-radiopaque appearance having ill-defined borders and occurring in anterior maxilla or mandible.

Keywords: Ameloblastoma, Desmoplastic, Maxilla, Recurrence.

INTRODUCTION

In 1984 Eversole *et al.* primarily described desmoplastic ameloblastoma and termed it as “ameloblastoma with pronounced desmoplasia”[1]. Since then numerous histological sub-classification has been published which commonly encompass: follicular, acanthomatous, plexiform & desmoplastic types [2].

Desmoplastic ameloblastoma (DA) is an unusual histologic variant of ameloblastoma characterized by a distinctive histomorphology, in conjunction with substantial stromal collagenisation or desmoplasia. Although, the latest 4th edition of the World Health Organization classification of head & neck tumours labelled it as the histological variant of conventional ameloblastoma and not as a discrete clinic pathological entity[3]. In the past, around about 238 cases of this unique entity have been recorded in the literature [4] which nearly accounts for 4%-13% of ameloblastomas with likely to be foreseen in the anterior region of the jaw as a mixed radiopaque-radiolucent lesion[5]. Recurrence rate of this rare tumour is relatively high in contrast with the conventional ameloblastomas [6,7] in spite of innumerable incidence of desmoplastic ameloblastoma have been documented in the scientific literature, the definite biologic outline is still ambiguous.

This report depicts a rare case of Desmoplastic ameloblastoma highlighting the peculiarity of a tumour along with emphasizing to figure out the myriad of tumours in the maxilla and its management.

CASE PRESENTATION

A 55-year-old male reported to the Department of Oral & Maxillofacial Surgery with the complaint of swelling on the right front side of the face since 6 months. The swelling had started 6 months ago and since then there had been a gradual increase in size to that observed at presentation. He underwent extraction of upper right canine and first premolar by a general dentist with no remission of symptoms, hence referred to us for further management. His medical history was non-contributory.

Extraoral examination showed a diffuse swelling measuring approximately 3 × 4 cm in the

upper right anterior side of the face extending from ala of nose superiorly to upper lip inferiorly and from philtrum of a lip to commissure of lip mediolaterally incorporating the body of maxilla on the right side. The skin overlying the swelling was normal with the marked deviation of an upper lip. On palpation, it was bony hard and fixed to underlying structures. No signs of lymphadenopathy or fistulae were present.

The intra-oral examination revealed a large mass approximately 3 × 4 cm in size, extending from upper right lateral incisor to Upper right 2nd premolar. Expansion of labial and palatal cortex was evident. A healing socket was noted with the overlying mucosa inflamed and erythematous (fig.1). On intraoral palpation, the swelling was found to be firm, bony hard in consistency and non-tender.

Further investigations included Facial computed tomographic (CT) with axial section image that revealed a diffuse ill-defined multilocular mixed radiopaque radiolucent lesion 3 x 4 cm extending from maxillary right lateral incisor to right second premolar (fig. 2).

Perforation of both buccal and palatal cortical plates along with marked bony expansion & thinning

were eminent along with an expansion of the lesion into the right maxillary sinus with the destruction of the anteromedial wall and mucosal thickening. A provisional diagnosis of a fibro-osseous lesion or benign/malignant tumour of the maxilla was considered with a biopsy warranted to confirm the diagnosis.

The final diagnosis was established through incisional biopsy performed under local anaesthesia. The histologic features were corroborating with those of desmoplastic ameloblastoma. Histopathologically the intraosseous tissue section shows the presence of ameloblastic follicles invading the bone.(fig.3 (a)) The follicles are typically arranged in a condensed fashion with focal areas exhibiting tall columnar ameloblasts exhibiting Vickers & Gorlin criteria.(fig.3 (b)) The surrounding connective tissue stroma is collagenous with bundles of collagen oriented parallel to compressed ameloblastic follicles.(fig.3.(c))

Considering extent and aggressiveness of lesion partial maxillectomy of the right maxilla was done under general anaesthesia via intraoral vestibular degloving incision which was followed by interim and final obturator prosthesis (fig 4, 5). The patient is on follow-up routinely and currently, he is free of ailment so far for a period of 5 years.



Fig-1: Clinical picture of lesion in right maxilla



Fig-2: Axial CT scan showing lesion in right maxilla

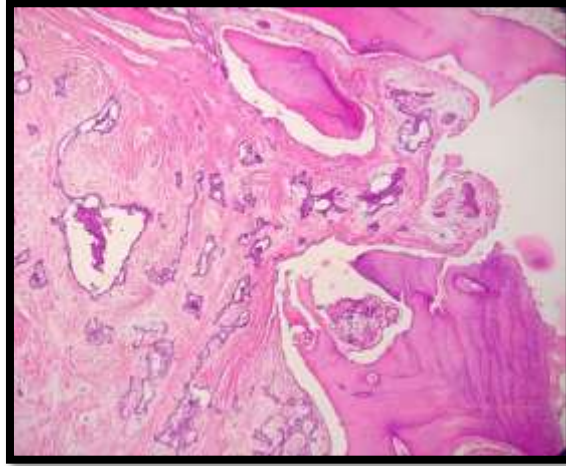


Fig-3a: Histopathological image showing lesion invading into bone

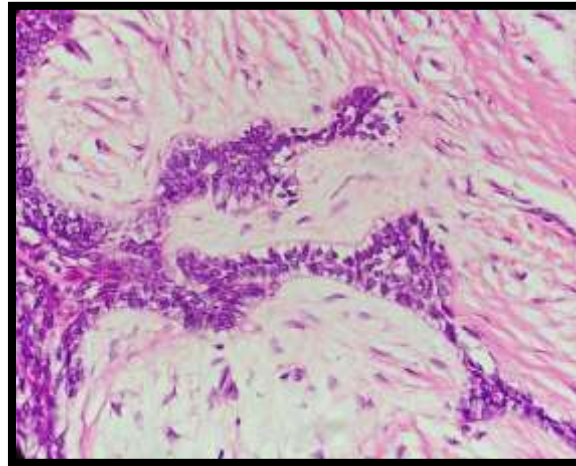


Fig-3b: Histopathological image of Vickers & Gorlin sign

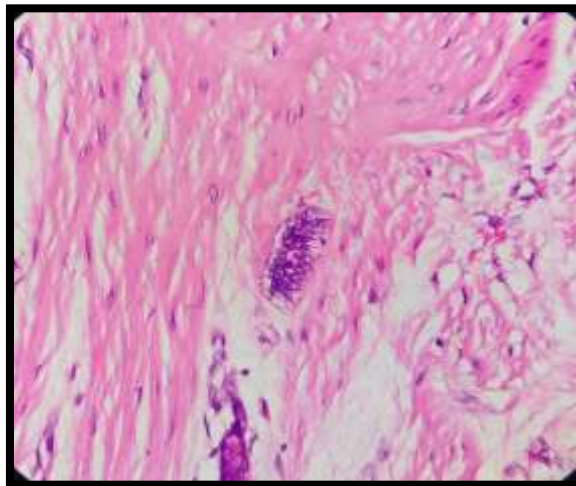


Fig-3c: Histopathological image showing desmoplasia



Fig-4: Post partial maxillectomy defect on the right side



Fig-5: Obturator in place of the defect

DISCUSSION

Desmoplastic ameloblastoma is a rare variant of ameloblastoma outlined by distinctive radiographic and histologic appearance [8]. The prevalence of desmoplastic ameloblastoma is quite low. While desmoplastic ameloblastomas are similar to conventional solid ameloblastomas regarding the age and gender distribution, tumours present a strong proclivity for the anterior or premolar region with equal incidences on both the jaws [9]. Clinically desmoplastic ameloblastoma may develop in all ages yet seen more commonly in the 4th or 5th decades of life with no sex predilection [10, 11]. A painless swelling, with buccal expansion, is a hallmark of desmoplastic ameloblastoma; this is congruent to the clinical finding discussed in our case. Kaffe *et al.* [12] and Li *et al.* [13] reported tooth displacement in 48%-92% of desmoplastic ameloblastomas, whereas root resorption was seen in 8.7%-33% of the cases.

Philipsen *et al.* opined maxillary desmoplastic ameloblastomas to be more aggressive as compared to mandible because of the insidious nature of maxillary tumours to that of mandible which can be attributed to confluence of vital structures and the soft spongy nature of the bone: a gateway for dissemination of tumours

that can camouflage radiographically as normal vital structures [14].

The final diagnosis of desmoplastic ameloblastoma is based on the histopathological evaluation of biopsy specimens. Microscopically extensive stromal desmoplasia is a consistent and distinguishing finding with a propensity to restraint the odontogenic islands at the periphery [15]. The palisading pattern of follicles as noticed in conventional ameloblastoma is absent. A fibrous capsule is also absent corresponding to the radiographically poorly defined tumour margin. [16]. Desmoplasia is a pathological phenomenon involving the stimulation of the connective tissue stroma. We must remain cognizant of the dual nature of the desmoplastic component. Desmoplasia could be an inductive effect of the ameloblastic follicle to prevent its further spread into the stroma. The presence of desmoplasia may also produce a channel for the unhindered infiltration of the ameloblastic follicles to a deeper bone leading to an invasive pattern of ameloblastoma. Histopathologically the diagnosis of desmoplastic ameloblastoma is a challenge as the ameloblastic follicles would be condensed leading to lack of expression of the Vickers and Gorlin Criterion pathognomonic of ameloblastoma.

It is thus imperative to sample adequately to identify a focal area of presence of desmoplastic ameloblastoma exhibiting the tall columnar cells with reversal of polarity and subnuclear vacuolation. Further, the radiographic picture may also be misleading as desmoplasia in a radiograph shows similarities to a fibro-osseous lesion.

The WHO classification of odontogenic tumours has delineated desmoplastic ameloblastoma to have low recurrence rate compared to unicystic ameloblastoma and peripheral ameloblastomas. Sun et al. after analysing 115 cases reported recurrence rate of 21.1 % following enucleation and 3.1% for resection [17].

As a tumour is devoid of a capsule, the cells impregnate amidst the trabeculae of the cancellous bone leaving them unimpaired. Hence, the tumour clearly infiltrates surpassing the radiographic margin which may lead to unusual recurrence rate following curettage. An analysis of 34 mandibular ameloblastomas by Marx and others reviewed 34 mandibular ameloblastoma revealing that a tumour extended 2.3–8.0 mm beyond the radiographic margin [18]. So accordingly, they have advocated resection of a tumour with the safety margin of 1 cm of bone over the radiographic margin. The rationale for reoccurrence can be relatively presumed as desmoplastic ameloblastoma primarily presents with indistinct border forming the unambiguous interface of the lesion with normal bone which can be challenging to assess. Secondly, further recurrent incidence in the maxilla can result in an early invasion of the adjacent structures [19].

In view of the sparsity of such case series and limited knowledge of its biological behaviour and prognosis, appropriate treatment strategies are not completely demarcated so far [7,11]. Hence, such cases warrant meticulous diagnosis and evaluation. It is still an enigma whether the recurrence is due to the nature of a tumour (lack of capsule and precise limit) or due to the unaccomplished surgery. Therefore to prevent recurrence resection is the most usually preferred treatment to intercept reappearance of the lesion [20], while Total maxillectomy or partial maxillectomy is best advocated for the favourable sequel when lesion exists in the maxilla. In our case, we had executed partial maxillectomy of the right side extending from midline to 2nd molar along with partial resection of the palate to avert the relapse following the procedure.

CONCLUSION

The desmoplastic ameloblastoma is characterized by peculiar clinic-radiographic and histologic features. The clinician should be vigilant with regards to such exquisite presentation of this benign tumour and it should be included in differential diagnosis of either fibro-osseous lesions or any mass/growth prevailing in an anterior region of either

jaw. Furthermore, the horizon of differential diagnosis of Desmoplastic ameloblastoma also extends over any mixed radiolucent-radiopaque lesion with inconspicuous radiographic margin presenting in the anterior–premolar region of the maxilla/mandible. There are still ongoing conflicts about the true biologic behaviour of the lesion due to a paucity of adequate cases. The radiological and histological findings of poor encapsulation and ill-defined borders expressive of infiltrative nature warrants in-depth analysis and a long-term follow-up.

REFERENCES

1. Eversole, L. R., Leider, A. S., & Hansen, L. S. (1984). Ameloblastomas with pronounced desmoplasia. *Journal of oral and maxillofacial surgery*, 42(11), 735-740.
2. Kramer, I. R., Pindborg, J. J., & Shear, M. (1992). The WHO histological typing of odontogenic tumours. *Cancer*, 70(12), 2988-2994.
3. Chung, C. H., Parker, J. S., Karaca, G., Wu, J., Funkhouser, W. K., Moore, D., ... & Shockley, W. W. (2004). Molecular classification of head and neck squamous cell carcinomas using patterns of gene expression. *Cancer cell*, 5(5), 489-500.
4. Anand, R., Sarode, G. S., Sarode, S. C., Reddy, M., Unadkat, H. V., Mushtaq, S., ... & Patil, S. (2017). Clinicopathological characteristics of desmoplastic ameloblastoma: A systematic review. *Journal of investigative and clinical dentistry*.
5. Philipsen HP, Reichart PA, Takata T. (2001). Desmoplastic ameloblastoma (including “hybrid” lesion of ameloblastoma). Biological profile based on 100 cases from the literature and own files. *Oral Oncol*; 37:455- 60.
6. Nair PP, Bhat GR, Neelakantan S, Chatterjee R. (2013). Desmoplastic ameloblastoma of mandible. *BMJ Case Rep.*;17(10):200082
7. Sivapathasundharam, B., Einstein, A., & Syed, R. I. (2007). Desmoplastic ameloblastoma in Indians: report of five cases and review of literature. *Indian Journal of Dental Research*, 18(4), 218.
8. Lam, K. Y., Chan, A. C. L., Wu, P. C., Chau, K. Y., Tideman, H., & Wei, W. (1998). Desmoplastic variant of ameloblastoma in Chinese patients. *British Journal of Oral and Maxillofacial Surgery*, 36(2), 129-134.
9. Rawson, K., Bastian, T. S., Siva, B., & Sharma, M. (2010). A case of Desmoplastic ameloblastoma of the anterior maxilla. *Journal of Indian Academy of Oral Medicine and Radiology*, 22(1), 60-62.
10. Beckley, M. L., Farhood, V., Helfend, L. K., & Alijanian, A. (2002). Desmoplastic ameloblastoma of the mandible: a case report and review of the literature. *Journal of oral and maxillofacial surgery*, 60(2), 194-198.
11. Sun ZJ, Wu YR, Cheng N, Zwahlen RA, Zhao YF. (2009). Desmoplastic ameloblastoma- A review. *Oral Oncol*;45:752- 9.

12. Kaffe I, Buchner A, Taicher S. (1993). Radiologic features of desmoplastic variant of ameloblastoma. *Oral Surg Oral Med Oral Pathol*; 76(4):525–9.
13. Li B, Long X, Wang S, Cheng Y, Chen X. (2011) Clinical and radiologic features of desmoplastic ameloblastoma. *J Oral Maxillofac Surg*; 69: 2173-85.
14. Philipsen, H. P., Reichart, P. A., Nikai, H., Takata, T., & Kudo, Y. (2001). Peripheral ameloblastoma: biological profile based on 160 cases from the literature. *Oral oncology*, 37(1), 17-27.
15. Savithri, V., Janardhanan, M., Suresh, R., & Kumar, R. V. (2013). Desmoplastic ameloblastoma with osteoplasia: Review of literature with a case report. *Journal of oral and maxillofacial pathology: JOMFP*, 17(2), 298.
16. Lamichhane NS, Liu Q, Sun H, Zhang W. A case report on desmoplastic ameloblastoma of anterior mandible. *BMC research notes*. 2016 Dec;9(1):171.
17. Kim, J. D., Jang, H. S., Seo, Y. S., & Kim, J. S. (2013). A repeatedly recurrent desmoplastic ameloblastoma after removal and allobone graft: Radiographic features compared with histological changes. *Imaging science in dentistry*, 43(3), 201-207.
18. Marx, R. E., Smith, B. H., Smith, B. R., & Fridrich, K. L. (1993). Swelling of the retromolar region and cheek associated with limited opening. *Journal of oral and maxillofacial surgery*, 51(3), 304-309
19. Belgaumi, U. I., Sundaresh, K. J., Varma, S., & Mallikarjuna, R. (2013). Desmoplastic ameloblastoma: a rare odontogenic neoplasm with unusual radiographic and histomorphological presentation. *BMJ case reports*, 2013, bcr2013009079.
20. Sheikh, S., Pallagatti, S., Singla, I., & Kalucha, A. (2011). Desmoplastic ameloblastoma: a case report. *Journal of dental research, dental clinics, dental prospects*, 5(1), 27.