CASE REPORT

Desmoplastic ameloblastoma of anterior mandible: A rare case report
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Abstract
Desmoplastic ameloblastoma (DA) is an uncommon variant of ameloblastoma that accounts for 4-13% of ameloblastomas. It differs from classical ameloblastoma in its site of occurrence, radiographic features, and histologic appearance. The World Health Organization (2005) classified this tumor as a variant of ameloblastoma. DA is a benign tumor with typical histopathological and radiographic features. Its anatomic distribution differs from the conventional type of ameloblastoma. This article reports a case of DA in a female patient aged 49 years with a huge swelling in her anterior mandible region for 2 years. Multilocular radiolucency was seen on the anterior mandibular region. Histopathological examination of the biopsy specimen confirmed the diagnosis of DA. Segmental mandibulectomy was performed from teeth 35 to 44. Prognosis was good. The patient is on regular follow-up.

Keywords
Ameloblastoma, desmoplastic ameloblastoma, segmental mandibulectomy

Introduction
Ameloblastoma is a benign, locally aggressive epithelial odontogenic tumor with slow growth. Most common site in the jaw is posterior aspect of the mandible, but it can also arise in the maxilla and anterior aspect of the jaws. Microscopically, this tumor shows cells resembling ameloblasts, with no formation of calcified material. Imaging characteristics range from a unilocular to multilocular radiolucency. Ameloblastomas are commonly seen in the age group of 20-50 years.[1] Most common types of ameloblastoma are follicular type and plexiform type, and least common types are acanthomatous and granular cell types. Other rare variants are desmoplastic ameloblastoma (DA), clear cell ameloblastoma, basal cell ameloblastoma, keratoameloblastoma, and unicystic ameloblastoma.[2,3] Approximately 0.9-12.1% of all ameloblastomas have been reported to be DAs. DA is an uncommon variant of ameloblastoma which has a tendency for the anterior jaw region, occurs more frequently in the anterior region of the jaw, and appears like a fibro-osseous lesion in radiographs with mixed radiopacity and radiolucency.[4] The first case of DA was reported by Eversole et al. in 1984, it was called as an ameloblastoma with pronounced desmoplasia.[4] The World Health Organization has classified DA as a distinct variant of ameloblastoma in the Classification of Odontogenic Tumors in 2003.[6] In this article, we report a case of DA of the anterior mandible in a 49-year-old female patient.

Case Report
A 49-year-old female patient reported to the Department of Oral Medicine and Radiology, Sri Rajiv Gandhi College of Dental Sciences and Hospital, Bengaluru, Karnataka, India, with a chief complaint of swelling in the chin area for 2 years. History of presenting illness revealed that the swelling started 2 years back after a trauma as a result of physical assault. Initially, the swelling was small and gradually increased to its present size. Swelling was associated with mild pain for 6-7 months.

Medical history revealed that she was suffering from hypothyroidism for 2 years and was hypertensive for 1 year and on medication for the same. On extraoral examination, a large well-defined swelling was seen in the chin region, about 10 cm × 7 cm in size, roughly oval, extending superoinferiorly from lower border of lower lip up to submental region, and bilaterally extending up to parasymphyseal region [Figures 1 and 2]. Skin over the swelling appeared tense and color appeared normal. On palpation, tenderness was present and swelling was firm to hard in consistency.

Intraoral examination showed a diffuse swelling in the labial sulcus and alveolar ridge extending from 44 to 36 causing lingual
Displacement of 43 and mobility of 31, 32, 41, and 43 [Figure 3]. Based on the history, slow duration of growth, and clinical features, a provisional diagnosis of infected benign odontogenic tumor was given. Ameloblastoma and traumatic bone cysts were given as differential diagnosis.

Orthopantomogram showed a well-defined multilocular radiolucency in the anterior mandible extending superiorly up to apices of mandibular anterior teeth causing knife-edge type of resorption and displacement of teeth, medially laterally from mesial aspect 46 up to mesial aspect of 36. Large locule was completely radiolucent and smaller locules showed the presence of radiopaque flecks. Inferiorly, it involved lower border of the mandible [Figure 4].

The patient was then referred to the Department of Oral Surgery where incisional biopsy of the lesion was performed. Specimen was sent to the Department of Oral Pathology. Histopathologically, it showed extensive desmoplastic stroma with abundance of collagen and moderate amount of cellular connective tissue, giving the final diagnosis as DA. The patient underwent segmental mandibulectomy followed by graft placement.

Discussion

DA is a benign tumor with typical clinical, radiographic, and microscopic features. It occurs more frequently in 40-50 years of age groups and affects both genders equally. DA occurs most commonly in the anterior region of the jaw and usually presents as a painless slow-growing swelling. As reported in previous literatures, the reported case of DA was also found in a middle-aged female who reported with a slow-growing swelling. DA differs from the other types such as solid/multicystic ameloblastomas, which are commonly seen in the mandibular molar/ramus regions, pain tends to be rare, but variable sizes of tumor causing root resorption of adjacent teeth are common.

Radiographically, solid/multicystic types of ameloblastomas appear as a multilocular radiolucency, giving a soap bubble or a honeycomb appearance, with well-defined borders. Reichart et al. reported that 51% of the DAs were unilocular with well-
defined margins. However, usually, it appears as an ill-defined, mixed, and radiolucent-radiopaque lesion resembling a benign fibro-osseous lesion, especially when evaluating panoramic and periapical radiographs. The present case also showed a multilocular radiolucency with flecks of radiopacities resembling a fibro-osseous lesion.

Microscopically, classical solid/multicystic type of ameloblastomas contains two main histopathologic patterns: Follicular and plexiform. It shows proliferating, irregularly shaped islands of the narrow cords of odontogenic epithelium embedded in a connective tissue stroma. However, DA has appears as irregularly shaped odontogenic epithelial islands surrounded by a thin zone of loosely arranged connective tissue embedded in desmoplastic stroma.[12]

Significance of desmoplasia
The presence of extensive desmoplasia in epithelial tumors is of unknown significance. However, extensive stromal desmoplasia in these tumors might inhibit the growth of tumor cells.[12]

Solid/multicystic ameloblastomas and DA show a high recurrence rate. Hence, the management is controversial, especially in conservative treatments such as enucleation and/or curettage.[12] Hence, the treatment of choice is surgical resection, due to the ill-defined border of tumor.[13]

Conclusion
The DA is considered as an uncommon tumor, but literature suggests that it can be considered as a distinct variant of ameloblastoma. It is important to remember that new case reports regarding DA are important in an attempt to better define observations regarding its nature and biological behavior.

References
1. Sun ZJ, Wu YR, Cheng N, Zwahlen RA, Zhao YF. Desmoplastic ameloblastoma - A review. Oral Oncol 2009;45:752-9.
2. Thompson IO, van Rensburg LJ, Phillips VM. Desmoplastic ameloblastoma: Correlative histopathology, radiology and CT-MR imaging. J Oral Pathol Med 1996;25:405-10.
3. Barnes L, Eveson JW, Reichart P, Sidransky D, editors. Pathology and Genetics of Head and Neck Tumours. Lyon: IARC Press; 2005.
4. Nair PP, Bhat GR, Neelakantan S, Chatterjee R. Desmoplastic ameloblastoma of mandible. BMJ Case Rep 2013;2013. pii: Bcr2013200082.
5. Eversole LR, Leider AS, Hansen LS. Ameloblastomas with pronounced desmoplasia. J Oral Maxillofac Surg 1984;42:735-40.
6. Barnes L, Eveson JW, Reichart PA, Sidransky D. World Health Organization Classification of Tumors. Pathology and Genetics of Head and Neck Tumors. Lyon: IARC Press; 2005. p. 284.
7. Kawai T, Kishino M, Hiranuma H, Sasai T, Ishida T. 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 Pathol Oral Radiol Endod 1999;87:258-63.
8. Durmus E, Kalayci A, Ozturk A, Gunhan O. Desmoplastic ameloblastoma in the mandible. J Craniofac Surg 2003;14:873-5.
9. Smallin SE, Faquin W, Susarla SM, Kaban LB. Peripheral desmoplastic ameloblastoma: Report of a case and literature review. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;105:37-40.
10. Torres-Lagares D, Infante-Cossio P, Hernández-Guisado JM, Gutiérrez-Pérez JL. Mandibular ameloblastoma. A review of the literature and presentation of six cases. Med Oral Patol Oral Cir Bucal 2005;10:231-8.
11. Reichart PA, Philipsen HP, Sonner S. Ameloblastoma: Biological profile of 3672 cases. Eur J Cancer B Oral Oncol 1995;31B:86-99.
12. Nakamura N, Higuchi Y, Mitsuysatu T, Sandra F, Ohashi M. Comparison of long-term results between different approaches to ameloblastoma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2002;93:13-20.
13. Keszler A, Paparella ML, Dominguez FV. Desmoplastic and non-desmoplastic ameloblastoma: A comparative clinicopathological analysis. Oral Dis 1996;2:228-31.