

Desmoplastic Ameloblastoma-An unusual presentation

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Abstract

Desmoplastic ameloblastoma (DA) is an epithelial odontogenic tumor, a rare variant of ameloblastoma. It was first described by Eversole in 1984. In the World Health Organization Classification of odontogenic tumors (2005), desmoplastic ameloblastoma has been considered as a distinct entity from conventional/multicystic ameloblastoma. DA differs strikingly in its clinical, radiological and histopathological presentation when compared to other variants of ameloblastoma. Herewith, we report an unusual case of DA in a 30 year old female patient involving anterior mandible with detailed clinical history, an unusual radiographic picture & histopathological presentation showing osseous tissue formation with areas of cystic change. Also review of literature and possible explanation of etiopathogenesis of osseous tissue formation & cystic change in DA will be discussed.

Keyword: Ameloblastoma, Desmoplastic ameloblastoma, Cyst.

Introduction

Desmoplastic ameloblastoma (DA) is a relatively unusual odontogenic epithelial tumor. It was first described by Eversole et al in 1984. DA presents with clinical and histopathological features that vary significantly from the conventional/multicystic ameloblastoma.⁽¹⁾ It shows marked predilection for the anterior jaws, and radiographic presentation varies from radiolucent to a mixed radiolucent/radiopaque appearance with ill-defined margins.^(2,3) Age ranges from 17 to 83 years with mean age of 41.9 years.⁽⁴⁾ Unlike conventional ameloblastoma which shows predilection for the mandibular posterior region, it has predilection for anterior or premolar region of both jaws.^(1,2)

Case Report

A 30 year old female patient reported to the Department of Oral Medicine and Radiology with a chief complaint of a long standing swelling in the lower right premolar-molar region since 15 years. The swelling was asymptomatic and non-tender in nature earlier but then gradually increased in size for the last 3 years and is now tender on palpation. Patient also gave history of past incomplete dental treatment for the same. Patient gave past medical history of right ovarian carcinoma for which she underwent chemotherapy in 2013.

Extraoral examination revealed facial asymmetry (Fig. 1A) due to swelling on right side of face extending from the right commissure to the mid body region of mandible anteroposteriorly and to approximately 2cm below the inferior body of mandible superoinferiorly of size approximately 3 x 2.5 cm. On palpation the swelling was bony hard and tender. The submandibular and cervical lymph nodes were non palpable. The overlying skin appeared normal.

Intraoral examination revealed a bony hard, tender lesion extending from the right lateral incisor to the right second premolar region. There was expansion of buccal and lingual cortical plates with marked expansion seen on buccal side obliterating the right buccal vestibule. Labial displacement of the right lateral incisor and lingual displacement of the right first premolar was also evident (Fig. 1B). The overlying mucosa showed prominent blood vessels (Fig. 1C). On palpation the swelling was non-pulsatile and did not show blanching on the application of pressure. Teeth from 42 - 46 were tender on percussion. The involved teeth were vital on electric pulp testing and exhibited no mobility.

On the basis of above clinical features provisional diagnosis of benign odontogenic tumor was given as the lesion was slow growing in nature, showed bucco-lingual expansion and had caused displacement of teeth.

The patient was then subjected to radiographic examination. Radiographic features showed contrasting appearance of the lesion. The true mandibular occlusal radiograph showed an ill-defined mixed radiolucent-radiopaque area along with a well-defined unilocular radiolucent lesion involving 47. Expansion of bucco-lingual cortical plates were also evident (Fig. 1D).

The Orthopantomogram (OPG) showed a lesion extending from 33 to mesial of 47. The posterior part of the lesion showed a well-defined unilocular radiolucency with corticated margin causing root resorption of 46 while the anterior part showed mixed radiopaque-radiolucent appearance with ill-defined borders crossing the midline and extending upto the 33. Lesion had caused displacement of 41, 42 and 44 along with the inferior displacement of the right mandibular nerve canal (Fig. 1E).

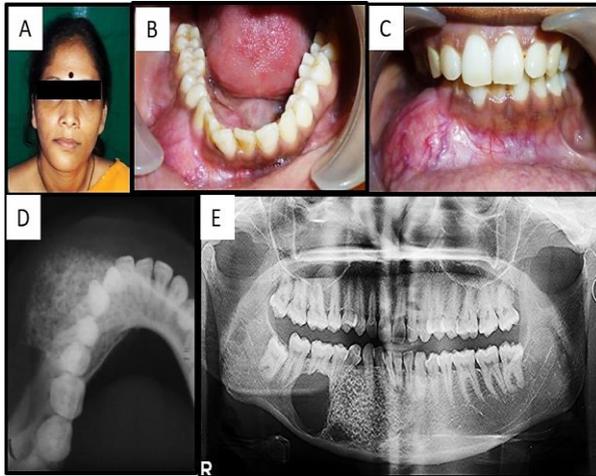


Fig. 1: A: Diffuse swelling in the right parasympyseal region of mandible. B & C: Bony hard swelling with bicortical expansion causing obliteration of buccal vestibule; prominent blood vessels on the surface. D & E: Occlusal radiograph and OPG shows an ill-defined mixed radiolucent-radiopaque lesion in the anterior half and a well-defined unilocular lesion in the posterior half

The patient also got her previous radiographs taken in year 2011 and 2015 (Fig. 2A, 2B). On comparing the previous radiographs with the present radiograph (Fig. 2C), it was observed that there was a progressive increase in the extent of the lesion antero-posteriorly with subsequent development of a radiolucent lesion posteriorly.

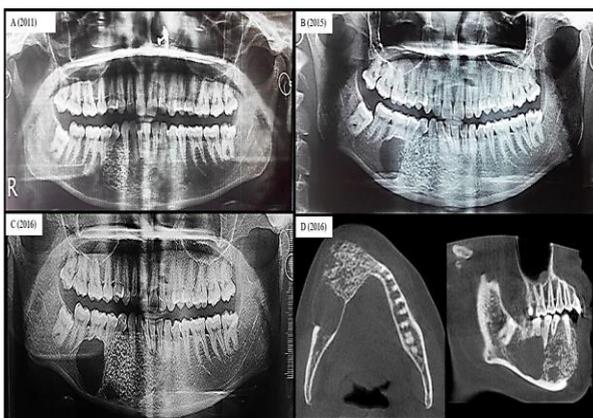


Fig. 2: A, B & C: OPG's of subsequent years shows progressive increase in the size of lesion with the development of radiolucent lesion. D: CBCT shows buccal cortical plate perforation and thinning of lingual cortical plate

Cone beam computed tomography (CBCT) examination was done to know the exact extent of the lesion. In addition to above mentioned radiographic findings CBCT examination revealed areas of

perforation in the buccal cortical plate and thinning of the lingual cortical plate (Fig. 2D).

On the basis of above clinical and radiographic findings a provisional diagnosis of benign odontogenic tumor with cyst formation was given with a differential diagnosis of ossifying fibroma, desmoplastic ameloblastoma, fibro-osseous lesions (fibrous and cement-osseous dysplasia) and hybrid tumor was given.

An incisional biopsy was performed to establish the definitive diagnosis. Before biopsy routine blood investigations were done which came out to be within normal limits. As lesion showed radiographic variation over the time, biopsy was taken from anterior (mixed radio-opaque area) as well as posterior (completely radiolucent area) of lesion. An aspiration was done from the posterior part. 1.5 ml of straw colored, thin, transparent cystic fluid was received which on wet mount showed RBCs and few pus cells. This confirmed the cystic nature of the lesion.

Anterior of the lesional area histopathologically showed odontogenic epithelial islands and cords scattered in a pronounced desmoplastic stroma with no clear ameloblastic polarization. However, in some areas, the odontogenic epithelial cells showed peripheral cuboidal and/or columnar cell alignment, surrounding stellate reticulum like cells. Connective tissue stroma consisted of dense collagen fibers and seemed to compress the odontogenic epithelial islands. Areas of new bone production with osteoblastic rimming were also seen within the lesional area (Fig. 3A, 3B & 3C).

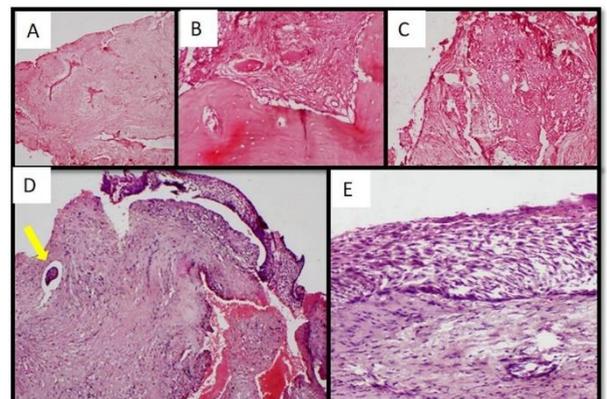


Fig. 3: A: Compressed odontogenic islands in dense hyalinized connective tissue stroma (H & E X40). B & C: Mature lamellar bone and odontogenic follicle (H & E X100). D: Cystic lumen, lining odontogenic epithelium, connective tissue capsule & odontogenic follicle within connective tissue wall (arrow) (H & E X40) E: Odontogenic lining epithelium consisting of hyperchromatic, tall columnar basal cells & subbasal stellate reticulum like cells (H & E X100)

However, biopsy from posterior region of lesional area showed a cystic lumen lined by odontogenic epithelial lining and connective tissue capsule. Lining

epithelium showed hyperchromatic, tall columnar basal cell with reverse polarity and suprabasal stellate reticulum like cells. Connective tissue capsule consisted of mature fibrous connective tissue interspersed with fibroblast. At one or two areas odontogenic epithelial islands were also seen within the connective tissue (Fig. 3D & 3E).

Considering the above histopathological features a diagnosis of DA with cystic change was given. Patient did not report for further treatment.

Discussion

Desmoplastic ameloblastoma (DA) is an uncommon odontogenic tumor, considered as a variant of ameloblastoma. It was documented initially in the literature by Eversole et al in 1984 and named it as "ameloblastoma with pronounced desmoplasia".^(3,5,6) In 2005, the World Health Organization classified ameloblastoma into four different types as solid/multicystic, extra-osseous, desmoplastic and unicystic.⁽⁷⁾ Due to varied clinical, radiographic and histopathological features, DA is considered as a distinct entity.

DA occurs in the age range from 17 to 83 years with a mean age of 41.9 years.^(3,4,5) It has strong propensity to occur in anterior region of both the jaws⁽⁶⁾ with no gender predilection.^(4,6) On radiographs, it frequently present as a mixed radiolucent – radiopaque lesion with ill-defined borders.⁽⁶⁾

Li B et al. in their study of 23 cases of DA, described three different radiological presentations: type I (14 cases) with radiolucent and radiopaque appearance (osteofibrosis type); type II (6 cases) had a completely radiolucent appearance (radiolucent type); and type III (3 cases) showed a radiolucent and radiopaque appearance combined with a large radiolucent change (compound type). They concluded that osteofibrosis type (type I) was the most common pattern, and the compound type to be the least common (type III).⁽⁵⁾ According to this, our case had radiographic features similar to type III in which the unilocular radiolucent lesion developed subsequently adjacent to the mixed radiopaque-radiolucent lesion.

This unusual radiographic appearance of DA could be because of the presence of the residual bone or due to new bone formation. Li B et al. believed that radiolucent-radiopaque appearance is attributed to the density of the compressed odontogenic epithelium, which is supported by desmoplastic stroma, and the residual bone invaded by the tumor.⁽⁵⁾ However, Okada et al. & Philipsen et al. were of the opinion that there is formation of new bone by the tumor itself within the densely collagenized stroma.^(8,9)

Histologically, DA is characterized by proliferating, irregularly shaped islands and cords of odontogenic epithelium of varying sizes embedded in a desmoplastic connective tissue stroma. The epithelial islands may show a pointed, stellate or "kite-like" (animal like)

appearance. The cells at the periphery of the islands are cuboidal and occasionally show hyperchromatic nuclei.^(10,11) Also, small cystic changes within tumor mass of a DA have been reported in the literature. Kishino et al. in their study of 10 cases of DA, found only 2 cases with mixed solid and cystic areas on histopathologic examination.⁽¹²⁾ On the other hand, Iida et al.⁽¹⁾ & Kawai et al.⁽¹³⁾ in their case reports described larger cystic lesions in the DAs. To date, it remains debatable whether the mural or unicystic ameloblastoma arises from epithelial cells of a preexisting cyst or a cyst develops within an ameloblastoma lesion.⁽¹³⁾ Iida et al. also suggested that the cyst was formed by cystic degeneration of the tumor epithelial nests.⁽¹⁾

In the present case, biopsy from the posterior region showed a large cystic cavity lined by odontogenic epithelial cells. This may be either due to fusion of small cystic degenerations (microcyst) in the solid mass of DA or it could be that unicystic ameloblastoma has developed in the vicinity of lesion which later on get fused with primary lesion (DA).

It is also likely that there was a simultaneous occurrence of the two lesions (UAB & DA) but the UAB was not obviously noticeable radiographically at the time of initial reporting.

Radiographically DAs may be misdiagnosed as cysts of jaws when they show a complete radiolucent picture.⁽⁵⁾ When radiographically present as a mixed radiolucent-radiopaque picture, it can mimic fibrous dysplasia, chronic osteomyelitis, and ossifying fibroma.⁽¹⁾

DA has a recurrence rate of 21% following simple enucleation, which could be attributed to the fact that the tumor is without capsule and hence infiltrate beyond the radiographic margins. Therefore, resection of the tumor is the treatment of choice. This brings the recurrence rate to as low as 3.1% which is comparable to that of solid ameloblastoma (4%).^(4,6)

Conclusion

DA is one of the variant of ameloblastoma, which clinically and radiographically can mimic an odontogenic cyst or a fibro-osseous lesion. The present case of DA radiographically showed a mixed radiolucent radiopaque picture in the anterior half of the lesion, and posterior half of the lesion showed a well-defined unilocular radiolucency. Histopathological examination revealed a combined appearance of desmoplastic ameloblastoma and a cystic epithelial lesion. This emphasize that diagnosis of DA should be based not only on clinical and radiographic findings but also on histopathological examination which is mandatory for the final diagnosis.

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