Case Report

Unicystic desmoplastic osteo-ameloblastoma of posterior mandible: Report of rarest of the rare case and review of literature

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Abstract

Desmoplastic ameloblastoma (DA) is an epithelial odontogenic tumor and a rare variant of ameloblastoma. It was first described by Eversole in 1984. In the World Health Organization (WHO) classification of odontogenic tumors (2005), DA 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. We report here an extremely rare "Hybrid DA" in a 50-year-old female patient with painless hard swelling involving right posterior mandible with detailed clinical history, an unusual radiographic and histopathological presentation. Histopathology revealed odontogenic epithelium in the form of follicles, ameloblastoma with cystic degeneration, and squamous metaplasia at places and elsewhere there were odosntogenic islands compressed by dense fibrocellular stroma suggestive of desmoplasia along with osseous tissue formation. Also review of the literature and possible explanation of etiopathogenesis of cystic change and osseous tissue formation in DA are discussed.

Keywords: Desmoplastic ameloblastoma, hybrid, osteoplasia, rare, unicystic

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INTRODUCTION

The World Health Organization (WHO) has defined the ameloblastoma as a true neoplasm of enamel organ-type tissue which does not undergo differentiation to the point of enamel formation. It was first recognized by Cuzack, in 1827. Ivy and Churchill has coined the term "Ameloblastoma" in 1934. Robinson has described it as "usually unicentric, non-functional, intermittent in growth, anatomically benign, and clinically persistent." It is the second most common odontogenic neoplasm and only odontoma outnumbers it in reported frequency of occurrence.[1] Six histopathologic patterns of ameloblastoma are described including follicular, acanthomatous, granular cell, basal cell, plexiform, and desmoplastic.[1,2]

Among the ameloblastomas, the desmoplastic variation is rare. The desmoplastic ameloblastoma (DA) is characterized by specific clinical, imaging, and histological features. It is more complicated to be treated because of its tendency to penetrate the surrounding bone. This tumor

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usually occurs in third to fifth decades of life and has equal sex predilection. DA has a marked prevalence to occur in the anterior region of the maxilla in direct contrast to conventional ameloblastomas, and radiography seldom suggests its diagnosis because of its mixed radiolucent and radiopaque appearance. It is a rare variant initially described by Eversole in 1984. It represents 4–5% of all ameloblastomas.[3] Takata *et al*. have reported 1.1% hybrid lesion of all ameloblastoma.[2]

DA has distinct histologic features comprised of stroma with intense desmoplasia, few epithelial islands, and cords of various sizes. The mixed radiolucent/radiopaque appearance is due to osseous metaplasia within the dense fibrous septa that characterize the lesion and is not a mineralized product of tumor.[3,4] When DA coexists with other variants of ameloblastomas, it is referred to as "Hybrid DA." Not many cases of DA have been reported in Indian literature and hence this endeavor to report a rare case of hybrid DA with review of literature emphasizing the clinical, radiographic, and histopathologic aspects that are relevant for diagnosis and treatment of this tumor.[3]

CASE REPORT

A 48-year-old female patient reported to the College of Dental Sciences and Research Centre, Ahmedabad with the chief complain of having painless swelling in right lower back side of the face since last 1 month. The patient was relatively asymptomatic before 1 month. Then she had noticed swelling in right mandibular back region. Swelling increased gradually. There were no associated complaints. The patient had past medical history of hyperthyroidism since last 3 years. Patient was on thyronorm medicine for the same (25 mcg 1 OD in morning). The patient had no relevant past surgical history.

On extra-oral examination, 6 × 3 cm sized oval swelling was noted on lower right back side of the face. Swelling was extending anterio-posteriorly from right corner of the mouth to the region 0.5 cm anterior to the lobule of the ear. Superio-inferiorly swelling was extending from the line joining the corner of the mouth to the lobule upto the lower border of the mandible. There was no change in overlying skin with absence of extra-oral draining sinus [Figure 1a and b]. On palpation single firm swelling was non-tender. There was no parasthesia of the lower lip of the same side.

On intra-oral examination, swelling on buccal side was extending from distal half of right firsts premolar upto the right mandibular third molar. Mild swelling was present on lingual side in premolar–molar teeth region. There was no abnormality in relation to overlying mucosa. Also there was no obliteration of the vestibule [Figure 1c]. On palpation swelling was firm and non-tender. But the center area of the buccal and lingual swelling were soft in consistency suggestive of possible buccal and lingual perforation. On Lingual plate expansion was present in premolar–molar region. There was no pus or fluid discharge on palpation. Grade I mobility was noted in mandibular right first molar only. Rest of the mandibular right posterior teeth were not mobile. Right mandibular posterior teeth were neither discolored nor displaced. All the teeth were vital on pulp vitality testing.

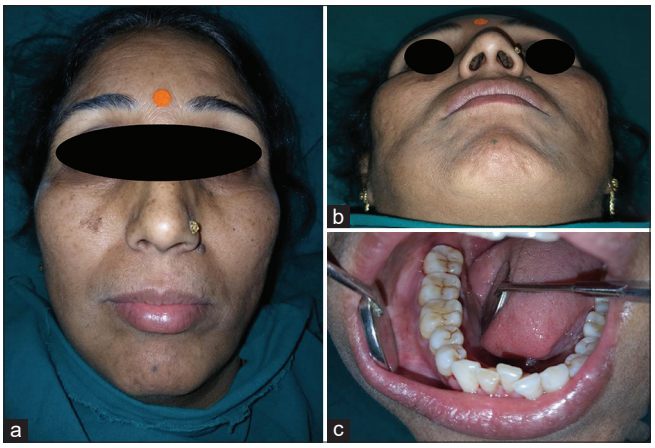

Figure 1: Preoperative clinical views. (a, b) Extra-oral views (c) Intra-oral view

On radiographic examination, orthopantomagram (OPG) revealed a single well-defined unilocular radiolucency in mandibular right symphysis-angle region. Radiolucency mesio-distally appeared extending from distal aspect of mandibular right first premolar to approximately 2 cm distal to the root of mandibular right third molar. Inferio-superiorly it appeared extending from inferior border of mandible to the alveolar crest of all mandibular posteriors in undulating fashion. The internal aspects of pathology appeared to be totally radiolucent without any evidence of calcification. Characteristic radiolucency within radiolucency appearance suggestive of cortical plate perforation. The periphery of radiolucency appeared to be thin, continuous, well-defined, and corticated causing gross thinning of inferior border of the mandible. Roots of mandibular right molars showed smooth knife edge resorption. The shadow of greater cornu of hyoid bone, inferior alveolar nerve, and mental foramen was evident within shadow of radiolucency [Figure 2a].

The computed tomography (CT) scan performed after OPG to exactly delineate the margins of the lesion which can aid while planning surgery [Figure 2b]. The axial section

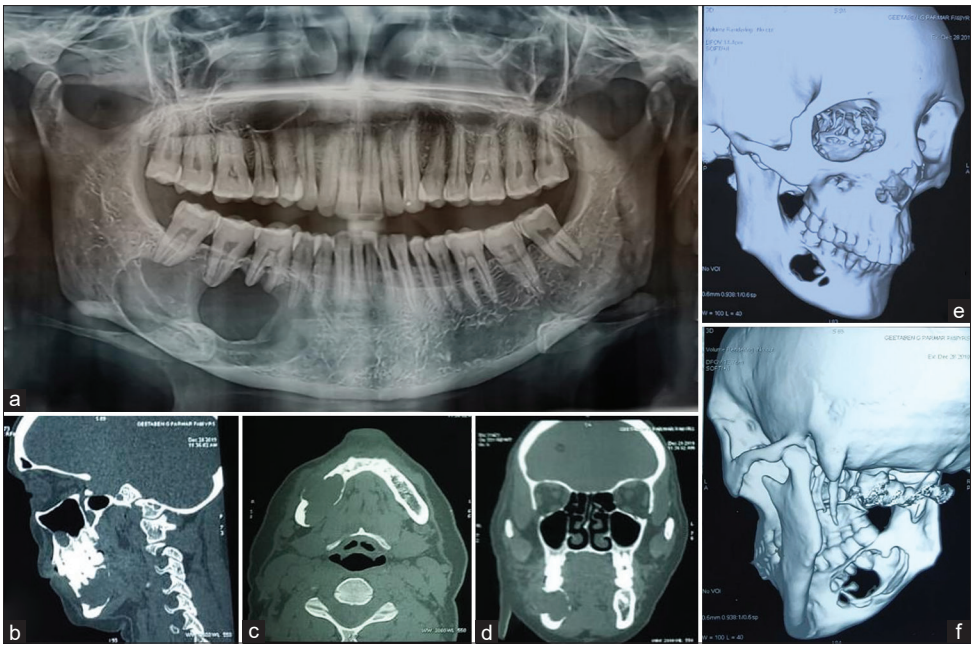

Figure 2: Preoperative radiographs. (a) OPG (b) Sagittal view (c) Axial view (d) Coronal view (e, f) 3D views

Figure 3: Intraoperative photographs. (a) Inferior alveolar and mental neurovascular bundle separated and saved (b) Application of Cornoy's solution (c) Aspirated fluid with specimen

showed bucco-lingual expansion with perforation in both buccal and lingual plates [Figure 2c]. Lesion appeared to be extending from para-symphysis region toward angle with more predominant expansion of lingual cortex. The periphery of lesion appeared to be smooth, coarse, and regular with dense internal radiolucency. In coronal section, gray shadow of soft tissue appeared to be continuous with internal lumen of pathology. [Figure 2d–f]

On the basis of above clinical and radiographic findings a provisional diagnosis of benign odontogenic tumor with cyst formation was given with differential diagnosis of ameloblastoma, keratocystic odontogenic tumor, and ossifying fibroma were given.

All preoperative routine blood investigations were reported normal except elevated T3 and T4 level. The same patient was referred to the physician, who advised her to increase the dose of thyronorm to 100 mcg per day in the same manner. After 15 days T3, T4, and TSH reports were repeated, which were normal. After the physician and anesthetic reference, the enucleation of the lesion was planned followed by application of Cornoy's solution.

Enucleation was performed under general anaesthesia. A 10 cc of dark red colored fluid was aspirted from the lesion. Inferior alveolar and mental neurovascular bundles were separated and saved [Figure 3a]. Root resection of mandibular right first molar was performed. Cavity was packed for 3 mins with the ribbon gauze dipped in Cornoy's solution [Figure 3b]. Before application of the Cornoy's solution, neurovascular bundle was painted with glycerine to prevent chemical injury to neurovascular bundle. Watertight suturing was performed using 3.0 silk sutures. Aspirated fluid and specimen were sent for histopathological examination [Figure 3c].

Histopathology

Histopathological examination of blood stained aspirated fluid was insignificant. H and E stained sections of the specimen showed the following:

- Section 1 (scanner view) showed cystic lumen lined by odontogenic epithelium and capsule showed dense

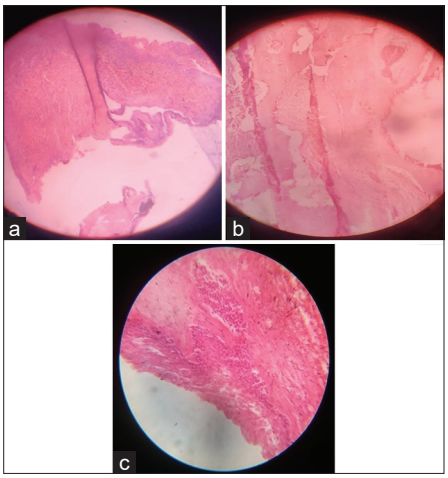

Figure 4: Microscopic features. (a) Odontogenic epithelium lining cystic lumen with dense fibrous connective tissue (scanner view). (b) Degenerated odontogenic epithelium with hyalinization and ossification (low power). (c) Cystic lumen lined by odontogenic epithelium with lining epithelial cells proliferating within connective tissue capsule (high power)

fibrous connective tissue [Figure 4a]. Low power showed cystic lumen lined by odontogenic epithelium with columnar basal cells and degenerated suprabasal cells, dense fibrous connective tissue with some areas with hyalinization and few islands of odontogenic epithelium showing desmoplasia.

- Section 2 (low power view) showed cystic lumen lined by degenerated odontogenic epithelium and fibrous capsule with some areas of hyalinization as well as ossification [Figure 4b].
- Section 3 (high power view) showed cystic lumen lined by odontogenic epithelium and some lining epithelial cells proliferating within connective tissue capsule [Figure 4c].

On the basis of histopathological features, it was diagnosed as unicystic desmoplastic osteo-ameloblastoma (a hybrid lesion).

Sutures were removed on tenth postoperative day. Extra-oral swelling was absent. There was neither pain nor pus discharge. Also, there was no parasthesia of right lower lip and chin region. Patient was recalled after 1 month, 3 months, and 6 months postoperatively. There were no fresh complaints. There were no signs of recurrence on the clinical and radiographical examination after 6 months of surgery [Figure 5]. Mandibular right first molar which was non-vital on pulp testing after 6 months follow-up, was subjected to endodontic treatment.

DISCUSSION

Eversole *et al.* were the first to describe three cases of DA in 1984. They referred it to as "ameloblastoma with pronounced desmoplasia."[3,4] In 2005, WHO have organized ameloblastoma as solid, extraosseous, desmoplastic, and unicystic types in its classification of odontogenic tumors. Literature review shows DA to be the least frequent odontogenic tumor. About 145 cases of DA have been reported worldwide, but cases reported in India are very few.[3,5]

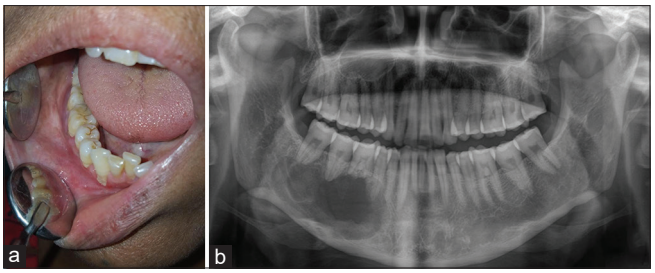

Figure 5: Postoperative follow-up after 6 months. (a) Clinical intra-oral view (b) OPG

DA is similar to conventional ameloblastoma in gender and age distribution, that is, DA is frequent in third to fifth decades of life with equal male-to-female ratio. Frequently, the tumor is asymptomatic. Painless swelling or expansion may be the first presenting sign as reported in many cases, and the same features were also seen in our case.[3,5]

In most of the cases of DA reported so far, there has been a strong predilection for the anterior maxilla. According to Kaffe *et al.* and Manuel *et al.*, >70% of DA cases happen to be in the anterior maxillary region, opposite to conventional ameloblastomas which is usually in the posterior mandibular region. Review done by Desai *et al.* of 89 cases of ameloblastomas has noted DA to occur commonly in anterior region of mandible (57%) than in maxilla (42.7%).[3] So majority of the cases can be seen in anterior maxilla and mandible.[3-6]

In contrast to it, in our case DA was present in posterior mandible. Kawai *et al.* and Utaka *et al.* have reported a case each of DA in the posterior mandibular region. Kishino *et al.* had reported 6 cases in posterior mandible. So considering the site predilection of DA, our case is a rare one. Demographic studies have revealed highest incidence of DA in Japanese race.[3]

Radiographic findings of our case were not like typical of DA. Typical DA appears as mixed radiopaque/radiolucency of indefinite borders. Such findings are seen in 50% of DA imitating fibro-osseous lesions.[2-4,6] The clinical D/D according to radiographic findings includes fibro-osseous lesions, fibrous dysplasias, chronic osteomyelitis, and ossifying fibroma. In our case lesion appeared with definite borders and did not appear like fibro-osseous lesion. But as in all cases of hybrid DA, root resorption was seen.[3]

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). 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.[5]

The present lesion differed considerably from the lesions in previous cases with respect to internal appearance; in our case, the lesion appeared primarily as a unilocular radiolucency. In addition, the lesion was situated almost entirely on the surface of the right mandibular cortex; in contrast, all of the previously reported cases affected cancellous portions of the jaws. It can be speculated that the radiographic characteristics of the previous cases might be explained by the growth pattern of this tumor, which characteristically infiltrates into marrow spaces at the periphery of the tumor. For this mode of tumor growth, adjacent bone trabeculae are prone to persist because osteoblastic activity, rather more vigorous than osteoclastic activity, may be induced by these neoplastic cells. This may lead to the numerous bony flecks seen radiographically, which are attributable to radiographic images of unresorbed or newly formed bony trabeculae. Moreover, such a growth pattern of infiltration into marrow spaces and lack of demarcation with fibrous connective tissues would correlate with the indistinct borders in this type of ameloblastoma. In our case, however, no invasive character of the tumor was found.[6]

This unusual radiographic appearance of DA could be because of the presence of the residual bone or due to new bone formation. Li *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. However, Okada *et al*. and Philipsen *et al*. were of the opinion that there is formation of new bone by the tumor itself within the densely collagenized stroma.[5]

Philipsen *et al*. suggested that tumor islands induce proliferation of mesenchymal cells, resulting in desmoplasia and metaplastic bone formation (osteoplasia). Kawai *et al*.[6] have suggested that the thin cortical plate of maxilla forms a weak barrier, to prevent the spread of tumor, than mandible. So, maxillary ameloblastomas may be able to spread earlier and more quickly than mandibular one.[3,4] This is suggestive of a larger size lesion in mandible as diagnosed late.

No desmoplastic variant of ameloblastoma with a large cystic lesion has previously been reported except by Kawai *et al*.[6] The present case is unique in that it was accompanied by such a large cystic lesion. The tumor epithelial cells, however, were not similar to the epithelial cells lining the lumen of the cyst. Nevertheless, it is strongly suggested that the cyst was formed by cystic degeneration of the tumor epithelial nests, because the epithelial cells lining the cyst wall consisted of "degenerated" epithelial cells. To date, it remains debatable whether the mural or unicystic ameloblastoma arises from epithelial cells of a pre-existing cyst or a cyst develops in an ameloblastoma lesion. The features of the present case suggest that if cystic degeneration occurs in the tumor epithelial islands or nests, epithelial cells lining the cyst lumen must be compressed, resulting in the degenerated form.

Hybrid ameloblastoma, first described by Waldron and El-Mofty, is tumor variant where histologically, areas of follicular or plexiform ameloblastoma coexists with areas characteristics of DA.[7] The histologic appearance of DA is characteristic and differs from SMA in the nature of collagenous fibrous stroma and the morphology of constituent cell types in odontogenic islands. Waldron and El-Mofty described the histologic appearance of DA as small ovoid islands and narrow cords of odontogenic epithelium widely separated by dense moderately cellular, fibrous connective tissue. Peripheral columnar cells with reversely polarized nucleus within the epithelial island are not a dominant feature. The odontogenic islands in DA are irregular and appear to be compressed or squeezed by stromalv tissue, giving rise to a stretched out tail like appearance. Microcysts containing either eosinophilic amorphous deposits or appearing empty may occur centrally. Spicules of mature lamellar bony trabeculae have been reported in intimate contact with the tumor and invasion has been demonstrated. Recently, Hirota *et al*. reported a case of DA with few areas of characteristic compressed tumor islands along a predominantly basal cell pattern of odontogenic cells similar to the appearance of basal cell ameloblastoma. In some instances, typical follicular tumor islands as seen in SMA surrounded by less dense collagenous stroma have been observed in DA. This entity is called a "hybrid" lesion.[2-4] No conclusion has been drawn about the biological profile of hybrid type of ameloblastoma due to insufficient no of cases.[7]

Our case showed histologic features very characteristically described in the literature so far for DA but belonged to the hybrid variety. This tumor is somewhat histologically different from other odontogenic tumors like ameloblastic fibroma, odontogenic fibroma, or squamous odontogenic tumor. In our case, diagnosis was not difficult because the stroma showed extensive desmoplasia and ameloblastic polarization. Some of the tumor nests showed clear follicular pattern with cystic degeneration and squamous metaplasia at places. The simultaneous presence of osteoplasia and desmoplasia as seen in connective tissue stroma of this tumor has been reported in only five other cases.[3]

It has been suggested that DA can behave aggressively and has the potential to grow to a large size. The maxillary prevalence explains its early invasion to adjacent structures as maxilla cannot offer an effective barrier. Diffuse radiographic appearance and histologic findings of bone invasion are the reasons behind not finding the exact interface of the lesion with normal bone making it difficult to be treated surgically. Since DA tends to infiltrate between bone trabeculae, curettage often leaves islands of the tumor within the bone, which eventually leads to recurrences.[3] According to the WHO classification, it is stated that "unicystic, peripheral, and possibly desmoplastic ameloblastoma have lower recurrence rates than other ameloblastomas." The radiological and histological findings of poor capsulation or total lack of capsule requires long-term follow-up and findings indicate that the DA has a potential for recurrence similar to other variants of ameloblastoma. So it is recommended to follow the same treatment protocol as that of "classical" ameloblastoma.[7] Therefore, we treated it with enucleation followed by application of the Cornoy's solution.

Desmoplasia of stromal connective tissue in DA can be argued to be a maturation change in solid multicystic ameloblastomas (SMAs) as similar dense collagenization is seen during maturation of long standing tumors. This argument can be supported by the "hybrid" tumor as seen in our case where follicles were present in the desmoplastic background. But the lesser frequency of DA in posterior mandible compared to SMAs then remains unanswered. It is probable that the location of the tumor can influence the maturity of the lesion and hence tumors in the anterior jaws may mature sooner than those occurring in the posterior mandible. Higher frequency of SMAs in anterior jaws in Blacks, indicating the possible racial influence in the site predilection of ameloblastoma. It is possible that the majority of these ameloblastomas could turn out to be DA if the careful histologic examination is carried out. Second, DA might not actually be a rare entity. Many hybrid lesions may have been misclassified since the presence of typical ameloblastic islands in some cases could have warranted a diagnosis of SMAs.[3,4]

CONCLUSION

Unicystic desmoplastic osteo-ameloblastoma is a very rare and unique hybrid entity. Clinically and radiographically it can mimic an odontogenic cyst or fibro-osseous lesion. It deviates from usual desmoplastic variant of ameloblastoma in terms of locus, radiologic appearance, and cyst formation. Histopathological examination revealed a combined appearance of desmoplstic ameloblastoma and a cystic epithelial lesion. This emphasize that diagnosis of it should be based not only on clinical presentation and radiologic features but also on careful histopathologic examination. A definitive final diagnosis prior to surgery is a must for proper and efficient surgical management especially to prevent recurrence. The biologic behavior of it is still unclear. This lesion will remain an enigma until researchers persue more definitive tumor analysis by immunohistochemistry. Hybrid lesions should be considered collision tumor. Until such time it is recommendable to treat cases of hybrid ameloblastoma like other variants of infiltrative ameloblastoma. Also, long-term follow-up is needed.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. APA. Shafer WG, Hine MK, Levy BM. A textbook of oral pathology. 6th ed. Philadelphia: Saunders; 1983.
2. dos Santos JN, De Souza VF, Azevêdo RA, Sarmento VA, Souza LB. "Hybrid" lesion of desmoplastic and conventional ameloblastoma: Immunohistochemical aspects. Rev Bras Otorrinolaringol 2006;72:709-13.
3. Joshi PS, Golgire S, Majati S. Hybrid desmoplastic ameloblastoma: A case report of rare entity and review of literature. J Dent Allied Sci 2014;3:47-52.
4. Yazdi I, Seyedmajidi M, Foroughi R. Desmoplastic ameloblastoma (a hybrid variant): Report of a case and review of the literature. Arch Iran Med 2009;12:304-8.
5. Khurana D, Thakur A, Kadam S, Tupkari JV. Desmoplastic ameloblastoma-An unusual presentation. J Oral Med Oral Surg Oral Pathol Oral Radiol 2017;3:172-5.
6. Kawai T, Kishino M, Hiranuma H, Sasai T, Ishida T. A unique case of desmoplastic ameloblastoma of the mandible. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1999;87:258-63.
7. Philipsen HP, Reichart PA, Takata T. Desmoplastic ameloblastoma (including "hybrid" lesion of ameloblastoma). Biological profile based on 100 cases from the literature and own files. Oral Oncol 2001;37:455-60.