Desmoplastic Ameloblastoma: Case Report and review

Dr. Varsha B. Aher[1], Dr. Swapnali Y. Chaudhary[2], Dr. Rajendra S. Birangane[3]

ABSTRACT

Desmoplastic Ameloblastoma is a rare histological variant of Ameloblastoma. Approximately 115 cases of desmoplastic Ameloblastoma have been reported so far in literature. The World Health Organization Classification of Head and Neck Tumors (WHO-2005) included desmoplastic Ameloblastoma as a variant of Ameloblastoma with specific clinical, image, and histological features in the year 2005.

We have presented a case of desmoplastic ameloblastoma in the premolar region of mandible with its clinical and radiographical viewpoints. This case report focuses on a DA which occurred in the mandible of a 26 year-old female. The main signs and symptoms included painless swelling with buccal expansion. Panoramic and periapical radiographs and computed tomography demonstrated an image similar to a fibro-osseous lesion. The lower border of the mandible was intact, after an incisional biopsy was performed; the DA was given as diagnosis. A marginal resection, without involving the lower border of the mandible, was performed in the affected region .There were no post-operative complications. The patient is undergoing routine follow up.

Key words: Desmoplastic Ameloblastoma, fibro-osseous lesion

[1]Senior Lecturer, [2]Reader, [3]Professor & Head

Department of Oral Medicine and Radiology, Pandit Deendayal Upadhyay Dental College, Sholapur, Maharashtra, India

Corresponding author mail: drvarshaaher2013@gmail.com

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INTRODUCTION

Ameloblastoma are the commonest odontogenic neoplasm affecting the jaws[1]. Despite their locally destructive nature, they are considered benign[2]. The most common histological types of Ameloblastoma are the follicular and plexiform, followed by the acanthomatous and granular cell types, comparatively less common variants include desmoplastic, basal cell, and clear cell.

When the desmoplastic type coexists with other types, it is called a 'hybrid' Ameloblastoma[3]

Desmoplastic ameloblastoma (DA) was first reported by Eversole et al. [4], in 1984 and was included in the World Health Organization's Classification of Head and Neck Tumors (WHO-2005). DA is a tumor with specific clinical, image, and histological features. In additional, DA is a rare form of tumor and has been described in 115 cases in English literature [5]. Radiographically, 50% of DA shows an image suggesting a benign fibro-osseous lesion [5,6]. Eventually, the clinical-radiographic features of DA may prove as a diagnostic delima to the surgeons and radiologists .Thus, to provide additional clinical-radiographic-histological data about DA, this article aims to describe a case of DA which occurred in the mandible of a 26 year-old female.

CASE REPORT

A 26-year-old lady presented with a painless hard swelling in the anterior part of the lower jaw on the right side which gradually increased in size in the past one year. Clinical examination revealed a mild facial asymmetry with a non-tender, non-compressible, hard swelling measuring 2 × 2 cm on the right premolar region of mandible, Overlying skin was normal in color, texture and consistency and was not adherent to the underlying swelling (Figure **1).**

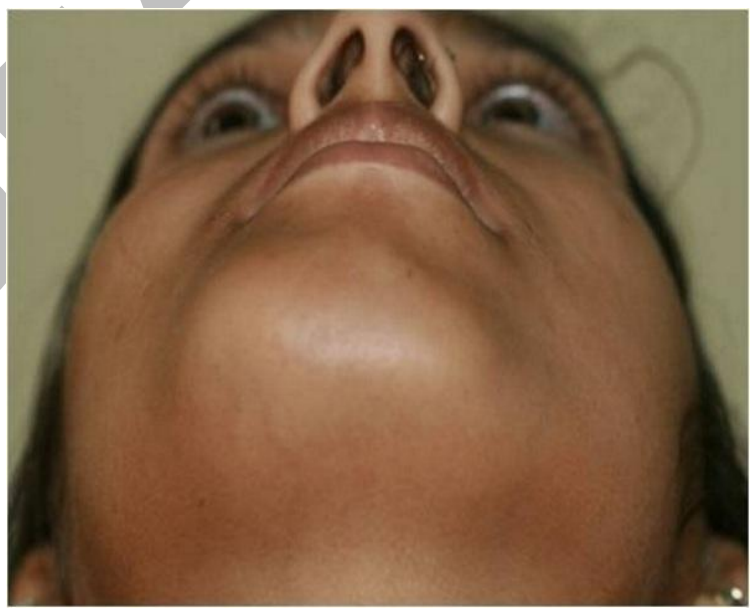

Figure 1: Extra-oral picture - mild facial asymmetry on the right anterior region of mandible with the overlying skin normal in color and texture.

The Intraoral examination (Figure 2) showed that there was a mild painless swelling in the vestibule from right mandibular canine to the distal aspect of second premolar on the same side. The swelling on palpation was firm with uniform hard consistency and there was expansion with the buccal cortical plate of mandible considering these findings, provisional diagnosis of benign tumor or odontogenic cyst was given.

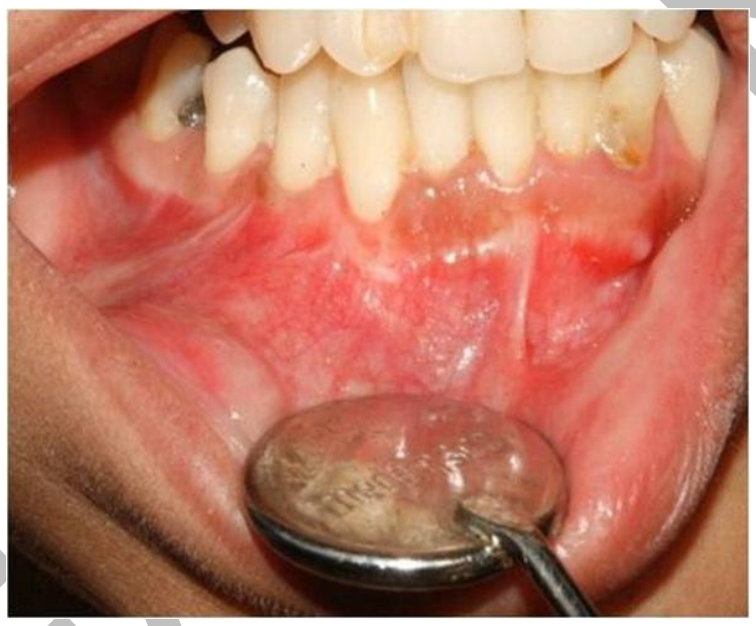

Figure 2: Intraoral picture - mild swelling in the vestibule from right mandibular canine to the distal aspect of second premolar on the same side.

Radiographically, the lesion in an intraoral peri-apical radiograph(Figure 3), showed as a unilocular, mixed lesion, well circumscribed by a radio-opaque border, about 3 × 2 cm, extending from the root of canine to the root of the first premolar posterior. The interior of the region showed a multiple round radiolucencies flocked together in some areas, while few radio opaque patches were also seen suggestive of a multi locular nature.

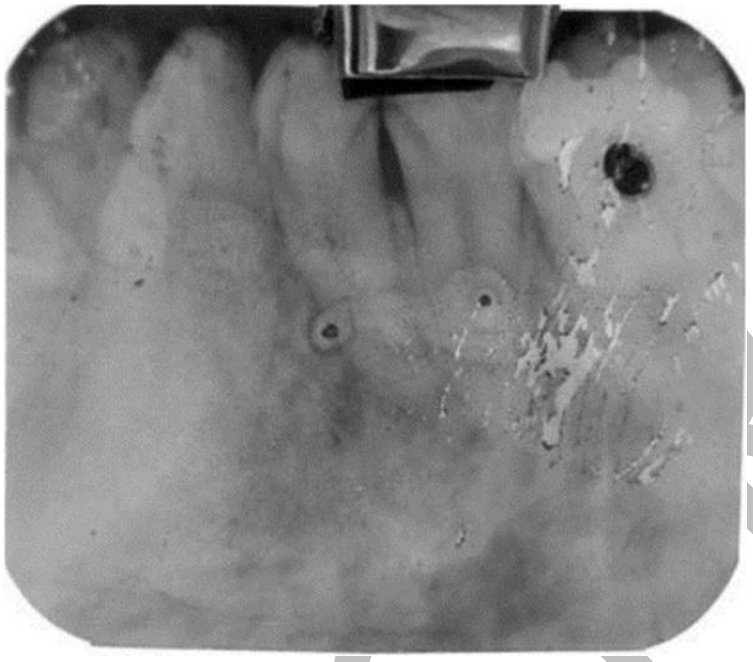

Figure 3: Intraoral periapical radiograph - well circumscribed mixed lesion with both radiolucent and radio-opaque areas within, extending from the root of canine to the root of the first premolar posteriorly, suggestive of a multilocular nature.

The Occlusal radiograph(Figure 4) of the mandible showed expansion and distortion of the buccal cortical plate in the area of 43 to 45, the lesion shows diffuse large radioluscency with ill defined borders along with few small radio-opaque patches interspersed within the main lesion.

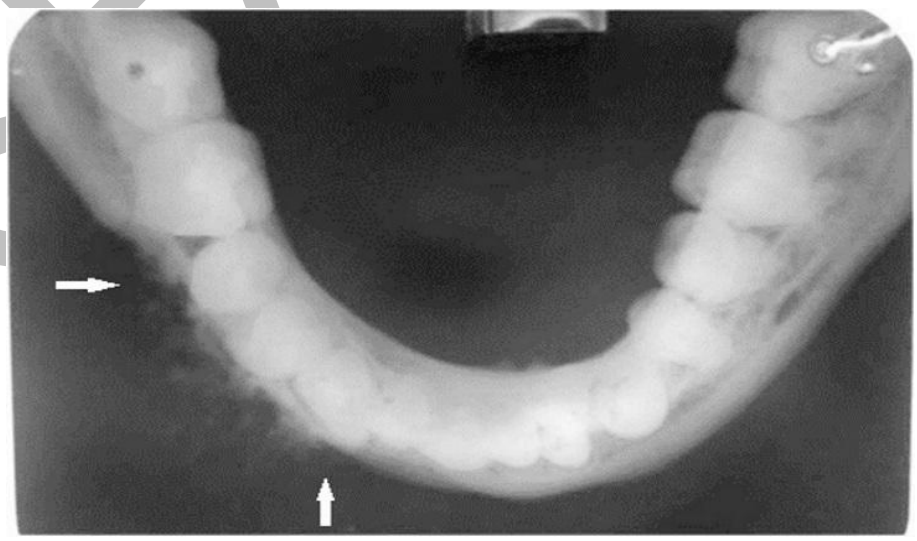

Figure 4: Occlusal radiograph showed expansion and distortion of the buccal cortical plate in the area of 43 to 45 (white arrows), the lesion shows diffuse radiolucency with borders along with few small radio-opaque patches interspersed within the main lesion.

Panoramic radiograph (Figure 5) demonstrated large diffuse radioluscenct lesion in the right side of mandible extending from the periapical region of the lower right central incisor to the medial aspect of the medial root of the first molar of same side with few small radiolucencies interspersed with few radiopaque areas. The periphery of the lesion is thin well defined, not surrounded by any sclerotic border. The internal aspect of the lesion was multilocular with presence of many septae within the lesion, forming multiple loculi, giving honeycomb like pattern. The inferior border of the mandible is intact. The cortices of mandibular canal also are intact.

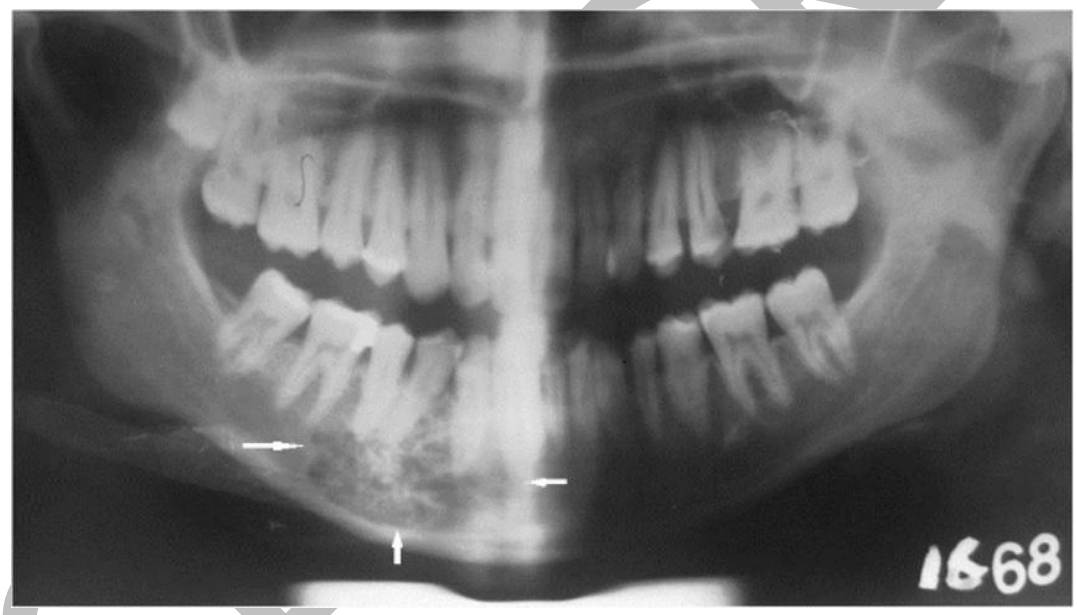

Figure 5: Panoramic radiograph- radiolucent lesion in the right side of mandible extending from the periapical region of lower right central incisor to the right first molar (white arrows) with multilocular appearance and presence of few septae within, giving honeycomb like pattern.

Computed tomography (Figure 6, 7, 8)

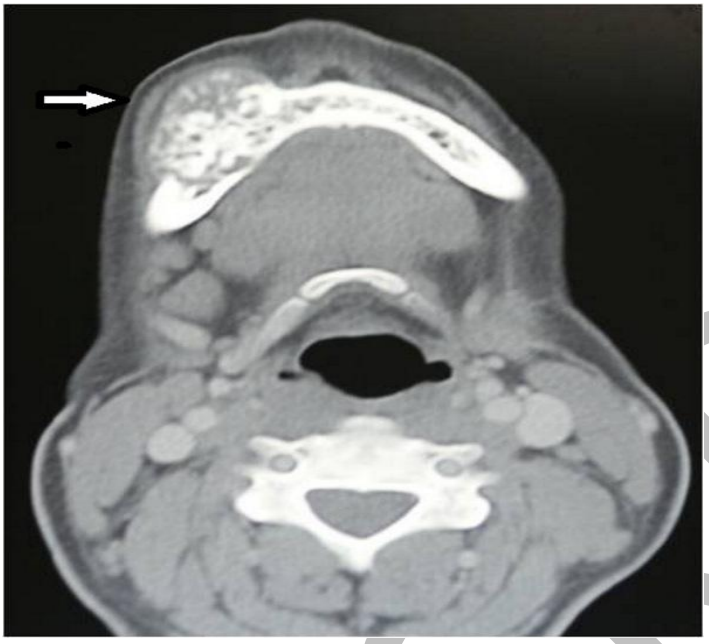

Figure 6: Axial post contrast image of the mandible shows expansile osteolytic lesion (white arrow) along the right half of mandible anteriorly. It shows mixed osteolytic and sclerotic areas within. Mild displacement of the overlying depressor muscles is seen.

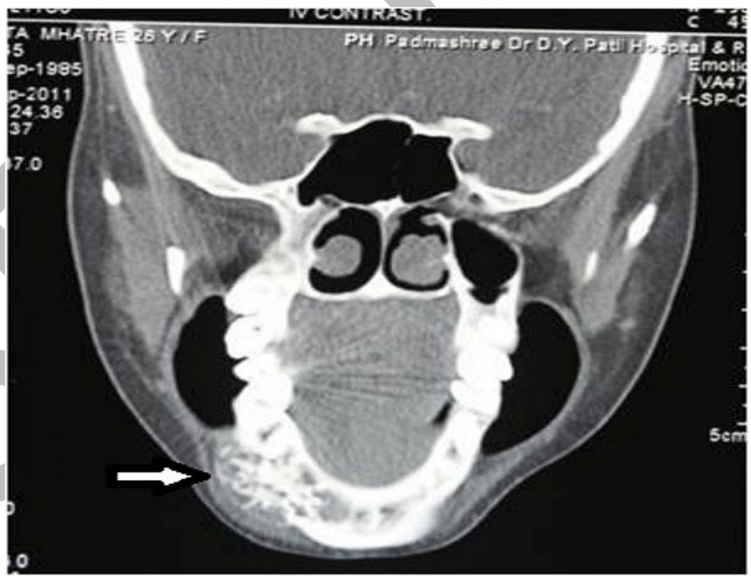

Figure 7: Coronal CT images of the mandible show expansile osteolytic lesion along the right half of mandible anteriorly, expanding along and eroding the buccal cortex (white arrow). Lamina dura of the affected teeth and the buccal cortex were destroyed.

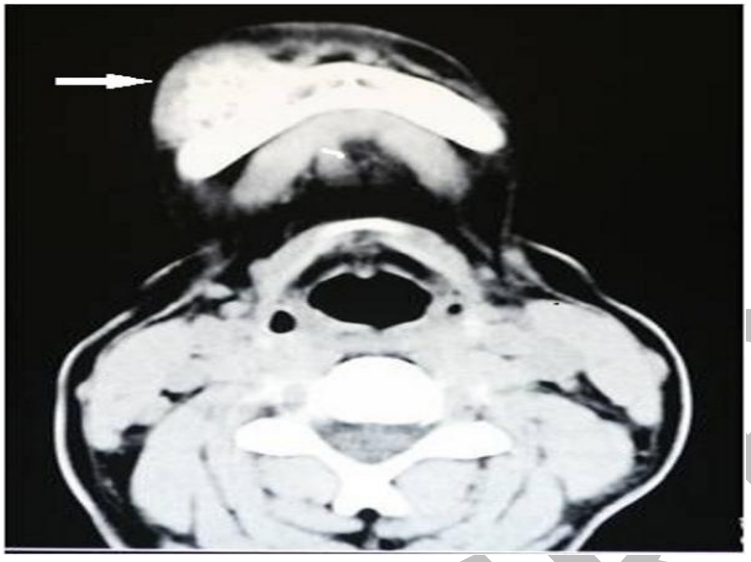

Figure 8: Axial post contrast images of the mandible show expansile osteolytic lesion along the right half of mandible anteriorly eroding the buccal cortex (white arrow) with mild enhancement of the overlying soft tissue.

The CT showed expansile lesion with erosive areas in the lingual plate of mandible and destruction of the buccal plate along with striated calcifications in the interior of the lesion. A computed tomography without contrast clearly revealed a solid, expansible, ill-defined, radio dense mass intermixed with radiolucent areas, arising from the anterior part of the body of the mandible and extending from the right canine to the right premolar region. The lamina dura of the affected teeth, the buccal and lingual cortices were destroyed.

Axial CT of the mandible with bone window settings, shows an expansible, solid, mixed radio dense-radio opaque lesion with poorly defined margins on the right anteriorly, with destruction of the buccal cortical plate and extension into the soft tissues.

Based on the radiological appearances, the diagnostic possibility of either a fibro-osseous mass or an ameloblastoma was suggested. The patient underwent an incisional biopsy, which showed a Desmoplastic Ameloblastoma (DA). A marginal resection in the affected area was then performed with a transitional prosthesis.

Presently, the patient is under follow-up since 1 year without any signs of relapse and with bony restoration in the affected area.

DISCUSSION

DA is a rare odontogenic tumor. According to literature a review carried out by Sun et al. [5] in 2009, 115 cases of desmoplastic ameloblastoma were reported. It is a tumor which shows a characterstic clinical, radiographic, and histological features. The histological picture shows marked stromal desmoplasia.[8] The common age of presentation is from the third to the fifth decades, It is more commonly found in males than females[10] in our case the patient was a 26 year old female., and Demographically it is seen that the highest incidence of this tumor is in patients of the Japanese race.[10]

The most of desmoplastic ameloblastomas tumors occur in the mandible, commonly in the anterior part [10], unlike the solid/multicystic variety which is seen in the posterior part of mandible. Clinically, maxillary lesions are more dangerous than mandibular ones as the thin maxillary bone is a weak natural barrier for tumors as compared to the thick mandibular bone [12].when occurs in maxilla , DA tends to invade the adjacent sinus and orbit and involve vital structures.

The radiographic appereance of a desmoplastic ameloblastoma tumor may be unilocular or multilocular [9] it is usually seen as an ill-defined mass containing osteolytic and sclerotic areas or containing multiplel radiodense flecks within a radiolucent background, suggestive of multifocal nature.. This is because of the infiltration of the tumor cells into the adjacent marrow spaces, along with ongoing osteoblastic activity [12].

CT is usually helpful to delineate the internal structure of the lesion. It is more accurate in determining the contours of the lesion, its internal structure, and its extension into soft tissues [3]. Since Ameloblastoma is an expansive growth within the bony tissue, on CT cystic areas of low attenuation along with isoattenuating solid regions are seen. Contrast-enhanced CT shows an enhancement effect in the solid components, erosion of the adjacent tooth roots can be seen on CT. When large in size, it perforates the bony cortex expanding into adjacent soft tissues, where, CT / CBCT proves to gives a better picture of the nature, extent, location and content of the lesion. Even though the radiation dose is much

higher in CT, its efficiency in identifying the contours of the lesion, its contents and its extension into the soft tissues, makes it preferable for diagnosis since the plain radiographs do not show interfaces between tumour and normal soft tissue; only interfaces between tumour and normal bone can be seen.

In MRI general ameloblastomas demonstrate a mixed solid and cystic pattern, with a thick irregular wall, often with papillary solid structures projecting into the lesion. These components tend to vividly enhance[9] MRI shows heterogeneous low to intermediate signal intensity on T1W images, heterogeneous high signal intensity on T2W images, and strong enhancement on post-gadolinium T1W images [10] MRI can clearly differentiate between solid and cystic components [11]

Both MRI and CT have equal ability to detect the cystic component of the tumor,MRI is essential for establishing the exact extent of an advanced maxillary Ameloblastoma.

On histopathology, desmoplastic Ameloblastoma reveals small areas and thin cords of odontogenic epithelium within the dense, fibrous connective tissue [7] Regions of mature lamellar bone and invasion may be also be seen [7] This histological finding indicates the potential for local invasion and also is the reason for the diffuse appearance on radiographs. Desmoplastic Ameloblastoma is so considered more aggressive than other common variants of Ameloblastoma [7]

As it is difficult to find the exact interface between the lesion and normal bone, it is difficult to cure these tumors surgically [12] Since in desmoplastic ameloblastomas there is infiltration of the bony trabeculae by the ameblastomatous cells,when only curettage is done it, often leaves islands of tumor within the bone, which eventually leads to recurrences. Therefore, block excision is the accepted form of treatment [12]

Thus, desmoplastic ameloblastoma should always be considered in the differential diagnosis of a mixed radiodense-radiolucent lesion with diffuse borders in the anterior premolar region of the jaws. Other possibilities include fibro-osseous lesions like osteitis, cementoblastoma ,cemento-ossifying fibroma, , fibrous dysplasia, calcifying odontogenic cyst, etc. A definitive diagnosis requires

histopathological study, to aid proper surgical management.

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