Desmoplastic ameloblastoma: A case report

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Abstract

Desmoplastic ameloblastoma (DA) is a rare variant of ameloblastoma, accounting for approximately 4 to 13% of ameloblastomas. It is uncommon, aggressive in nature, and there are high chances of misdiagnosis. Clinical and radiographical features are similar to fibro-osseous lesions of jaw. We reported a case of 35-year-old male patient of DA.

Keywords: Desmoplastic ameloblastoma, histopathology, radiology, treatment

Introduction

Odontogenic tumors (OTs) are assorted lesions derived from epithelial or ectomesenchymal tissues or both. They range from hamartomatous or non-neoplastic tissue proliferation to malignant neoplasms with metastatic potential. In humans, tumors of the odontogenic tissues are comparatively rare, comprising about 1% of all oral and maxillofacial biopsy specimens diagnosed.[1] Ameloblastoma is a neoplasm of odontogenic epithelium, principally of enamel organ-type tissue that has not undergone differentiation to the point of hard tissue formation. It accounts for about 1% of all oral tumors and about 9-11% of odontogenic tumors. It is generally a slow-growing but locally invasive tumor.[2]

Desmoplastic ameloblastoma (DA) is rare, accounting for approximately 4 to 13% of ameloblastomas. It was first described by Eversole et al. in 1984 as a new type of ameloblastoma.[3] World Health Organization (WHO) 2005 histological classification of odontogenic tumor categorize DA as a separate entity and defines it as a variant of ameloblastoma with specific clinical, imaging, and histological features.[4]

We reported a case of DA in 35-year-old male patient based on typical clinical, radiological, and histopathological features.

Case History

A 35-year-old male patient reported with a chief complaint of pain, loose teeth, and swelling in the lower front tooth region since 3 months. On clinical examination, extra orally no abnormality was detected [Figure 1]. Intraoral examination revealed a solitary swelling measuring approximately 3 × 2 cms, extending from a distal surface of 35 to 32 regions and from the marginal gingiva to the alveolar mucosa. Mucosa over the swelling was normal, with no surface changes, no sinus openings or discharge. On palpation, it was hard in consistency and nontender. Grade one mobility with respect to lower left lateral incisor detected. Mouth opening was 40 mm [Figure 2]. On radiographic examination, orthopantomogram (OPG) revealed periapical radiolucency in relation to the lower left lateral incisor [Figure 3]. Computed tomography showed soap bubble appearance with areas of calcifications [Figure 4]. Considering the clinical features and mixed radiolucency and opacities, provisional diagnosis of fibro-osseous lesion was made.

Incisional biopsy was done under local anesthesia and tissue was sent for histopathological examination [Figure 5]. Hematoxylin and eosin-stained sections showed connective tissue with collagen fiber bundles and few areas of hyalinization and thin strands

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and cords of odontogenic epithelial islands containing central cuboidal cells and peripheral flattened epithelial cells [Figure 6].

Correlating with clinical and radiological features, a histopathological diagnosis of DA with respect to anterior mandible was given and en bloc resection was done with respect to mandibular left canine to right premolar region.

Excisional biopsy tissue was sent for histopathological examination. Hematoxylin and eosin-stained tissue sections showed odontogenic islands lined by ameloblast-like cells with central stellate reticulum like cells and cystic degeneration, connective tissue shows collagen fiber bundles and few areas of hyalinization and also evidence of thin strands and cords of odontogenic epithelial islands containing central cuboidal cells and peripheral flattened epithelial cells in some areas. Correlating

Figure 1: No abnormality detected on extraoral clinical examination

Figure 2: Solitary swelling measuring approximately 3 x 2 cms, extending from the distal surface of 35 to 32 regions and from the marginal gingiva to the alveolar mucosa

Figure 3: Periapical radiolucency in relation to lower left lateral incisor

Figure 4: Computed tomography showed soap bubble appearance with areas of calcifications

Figure 5: Incisional biopsy specimen for histopathological examination

Figure 6: Areas of hyalinization and thin strands and cords of odontogenic epithelial islands containing central cuboidal cells and peripheral flattened epithelial cells
with clinical and radiological features, a histopathological diagnosis of DA with respect to anterior mandible was given.

**Discussion**

In 1984, Eversole established the report on DA to the English literature thereby describing three cases and called it an 'ameloblastoma with pronounced desmoplasia'. In the WHO’s Histopathological Typing of Odontogenic Tumors 2005, DA is included as a separate clinicopathological entity and classified ameloblastoma into four types as solid/multicystic, extra-osseous, desmoplastic, and unicystic. The term “hybrid lesions” was introduced by Waldron and El-Mofy reporting a condition in which DA was present close to follicular or plexiform ameloblastoma and Wakoh et al. presented a case of a patient demonstrating follicular-type ameloblastoma with desmoplasia, in whom radiological findings suggested the coexistence of a fibro-osseous lesion with a solitary cystic lesion and proposed it to be hybrid follicular/DA.\[5,6\]

The incidence of DA is very low and the literature states that it is reported to range from 0.9% to 12.1%.\[7\] The age of the patient in our case was 35 years. The mean age of DA patients at the initial presentation ranges from 40 to 49, and DA exhibits a similar gender distribution to other ameloblastomas.\[8\]

Approximately half of the desmoplastic lesions are located in the maxilla, and the vast majority of them occur in the anterior mandible or premolar portion of the jaws. Frequently, the tumor is asymptomatic, the tumefaction is painless. and the expansion of the involved bone is the commonly observed clinical manifestations.\[9\] In our case, the location of the lesion was in the mandibular anterior front tooth region also involving left lateral incisor to left second premolar region, clinical signs and symptoms like pain and discharge were absent.

In a review of 115 cases of DAs, the commonly observed radiographical features were mixed radiolucent/radiopaque (56%), multicystic (49%), and with ill-defined borders (64%).\[9\] In this case, multicystic radiolucentcy with radio-opaque areas was observed in the mandibular anterior region.

Based on age, clinical and radiographical features, our provisional diagnosis was fibro-osseous lesion. Such findings are seen in 50% of DA imitating fibro-osseous lesions.\[10\]

In this case, the histopathological features of both incisional and excisional biopsy showed odontogenic island with desmoplastic stroma. These were in consistent with a confirmatory diagnosis of DA that was made by histopathological evaluation. The microscopic features usually include: (1) stromal desmoplasia, in the form of moderately cellular, fibrous connective tissue with abundant collagen, which is the most consistent and distinguishing feature; (2) Islands of different shapes of odontogenic epithelium; (3) peripheral layer of cuboidal cells; and (4) hypercellular central area composed of spindle-shaped or polygonal epithelial cells.\[11,12\]

The case was reported and immunohistochemical (IHC) staining was done by Ramesh et al. in that odontogenic epithelial strands and follicles showed positivity for cytokeratin 19 and negativity for vimentin. Plump fibroblasts as well as pleomorphic cells invading the bony trabeculae were noticed. These cells were negative for cytokeratin 19 and positive for vimentin, thus reflecting their mesenchymal nature.\[12\]

IHC shows variable expressions of S-100 protein, desmin, capsid-3, increased expression of Fas and p63, and decreased expression of cytokeratin. They are strongly positive for fibronectin, type I collagen, and collagen type VI. Oxytalan fibers have been demonstrated in the stroma of DA. By these findings, some authors suggest that DA arises from the epithelial rests of Malassez in the periodontal membrane of the related tooth or originate from de novo synthesis of extracellular matrix proteins.\[13\]

**Conclusions**

The DA is a rare variant of ameloblasoma. The clinical and radiographic features of DA are similar to fibro-osseous lesions, but definitive diagnosis should always be based on the histopathologic findings. Patients present with milder form of the disease and rare diseases hide the symptoms. Possible predictive diagnostic values are lower than the hospital. In such a scenario, these case reports are helpful to family medicine and primary care by identifying the rare cases and unexpected cases, diagnosing, decision-making, and treatment plan. Treatment also varies with fibro-osseous lesion and DA. Histopathological diagnosis of incisional biopsy gives a proper channel for the treatment plan. So, it is needed for the surgeon to find the ultimate diagnosis to plan the treatment and make a regular follow-up of cases, because of its aggressive nature a complete resection and long-term follow-up is required to check any recurrence and even prognosis.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.
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