Desmoplastic ameloblastoma with mucous cell differentiation: A Rare case report

ABSTRACT

Ameloblastomas are tumors arising from the odontogenic epithelium. Ninety cases of desmoplastic ameloblastoma have been reported so far in the literature, out of which only five cases with mucous cell differentiation have been reported. We are presenting a case of 24-year-old female having a chief complaint of a painless swelling on the left side of the face for 7 months. After radiological, histopathological findings lesion was diagnosed with unicystic ameloblastoma which was treated by segmental resection. The purpose of this article is to present a case of desmoplastic ameloblastoma that has occurred in an unusual site and has unique histopathological features.

Keywords: Desmoplastic ameloblastoma, mucous secreting cells, segmental resection

INTRODUCTION

Ameloblastoma can be defined as a tumor that is usually unicentric, nonfunctional, anatomically benign, intermittent in growth, and clinically persistent.[1]

Eversole et al., in 1984 was the first to describe three cases of the desmoplastic variant of ameloblastoma. Later, Waldron and El Molly reported an additional of 14 cases and since then 73 cases have been reported in literature taking the total no of reported cases to 90.[2‑4] This unusual variant was characterized histologically by extensive stromal collagenization. Immunohistochemical studies suggest that the desmoplasia originate from de novo synthesis of extracellular matrix proteins.[5]

CASE REPORT

A 24-year-old female patient was referred to the department of oral and maxillofacial surgery at our institute in august 2010 with the chief complaint of a painless swelling on the left side of the face for 7 months. The swelling had gradually grown to the present size [Figures 1 and 2]. She also noticed the swelling intraorally in relation with the left mandibular molars. There was no history of trauma or any remarkable medical history.

Physical examination revealed a 4 cm×2 cm bony hard swelling which was nontender, nonfluctuant, noncompressible, and nonpulsatile. Swelling extended beyond the inferior border of the mandible and up to the left parasympyseal region anteriorly. The patient showed no signs of paraesthesia. A single swelling extending from the lower left premolars to the first molar with the expansion of buccal (maximal) and lingual (minimal) cortical plates noted [Figure 3]. Overlying mucosa appeared normal. Teeth 34 and 36 were 1° mobile; while 35 was 2° mobile. No occlusal discrepancy was noted.

On radiographic evaluation, a well-defined multilocular radiolucency with bony septa extending from 32 to 36 was seen (soap bubble appearance). Resorption of the apices of...
34, 35, and 36 was noted. Pathologic migration of 34, 35 was seen [Figure 4]. There was thinning and expansion of buccal and lingual cortical plates [Figure 5]. Inferior border of the mandible was thinned out in relation to the lesion. Fine septae-producing locules were noticed. The inferior border of the mandible was intact with no expansion.

From these clinical and radiographic findings, a possible diagnosis of a dentigerous cyst, ameloblastoma, or odontogenic keratocyst was made. To obtain a correct diagnosis an incisional biopsy was taken and the lesion was diagnosed as unicystic ameloblastoma. Specimen showed marked stromal desmoplasia characterized by moderately cellular fibrous connective tissue with abundant collagen formation and the presence of small ovoid or follicle-shaped islands of odontogenic epithelium.

Informed consent explaining the surgical procedure and outcome of the treatment was obtained from the patient. Segmental resection of the left side of the mandible extending from the distal aspect of 41 to the distal aspect of 38 using a combined transoral and extraoral approach was carried out [Figure 6]. Excised specimen was given for histopathological reporting [Figure 7]. A few large odontogenic islands were seen in different sections of staining. The islands were very irregular in shape with a pointed, stellate appearance. Slides showed compressed odontogenic islands in dense collagenous stroma suggestive of desmoplastic ameloblastoma [Figure 8]. Typical “animal like” pattern could be appreciated. Some slides also showed few dilated follicles in the collagenous stroma. Thus, both compressed and dilated follicles were seen [Figure 9]. On higher magnification signate ring-shaped cells were seen undergoing mucous cell differentiation along with squamous metaplasia within the follicle [Figure 10]. Mucous retained pools were also seen in different sections [Figure 11]. Immediate reconstruction using a titanium reconstruction plate was carried out followed by primary closure of the mucosa and skin [Figure 12].

**DISCUSSION**

The desmoplastic ameloblastoma is a rare and infrequent tumor, characterized histologically by marked stromal

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**Figure 1: Extraoral appearance**

**Figure 2: Worms view showing the swelling**

**Figure 3: Expansion of cortical plates**

**Figure 4: OPG showing the extent of lesion**
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desmoplasia.[2] The common age of presentation is from the third to the fifth decades. Among patients, men outnumber women.[6] The majority of desmoplastic ameloblastomas occur in the mandible, commonly in the anterior part.[6] On histopathology, desmoplastic ameloblastoma reveals small areas and thin cords of odontogenic epithelium distributed between dense, fibrous connective tissue. Regions of mature lamellar bone may be seen and invasion may be

![Figure 5: Coronal section at the level of the first molar](image)

![Figure 6: Surgical exposure](image)

![Figure 7: Excised lesion](image)

![Figure 8: Photomicrograph showing compressed odontogenic islands (A) in dense collagenous stroma (B) suggestive of desmoplastic ameloblastoma](image)

![Figure 9: Photomicrograph showing few dilated follicles (A) in collagenous stroma (B)](image)

![Figure 10: Higher magnification inset from Figure 9 indicating signate ring mucous cell differentiation (A) along with squamous metaplasia within the follicle (B)](image)
Desmoplastic ameloblastoma is therefore considered more aggressive than other common variants of ameloblastoma. In this reported case, we found with the compressed islands also a few dilated islands. Thus, slides stained at a higher magnification revealed signate ring mucous cell differentiation (A) along with squamous metaplasia within the follicle. This type of mucous cell development strikes out from other variants of desmoplastic ameloblastoma, with this being only the 5th reported case of its kind. The biologic behavior of the desmoplastic ameloblastoma with mucous cell differentiation, including recurrence rate, still cannot be fully appreciated due to the relatively few reported cases with sufficiently long follow-up periods. The possible pathogenic mechanism of this case would appear to be a reflection of the pluripotential character of the odontogenic epithelium. This case indicates that multi-potential odontogenic epithelial tissue can develop diverse differentiation. The prognosis, in this case, will probably be as expected for conventional desmoplastic ameloblastoma.

Treatment must be guided by consideration of the behavior and potential of the tumor. Less than complete excision is equivalent to planned recurrence. Curettage and enucleation of desmoplastic ameloblastoma either separate or in combination, have resulted in recurrence. Since desmoplastic ameloblastomas tend to infiltrate between bone trabeculae, curettage often leaves islands of tumor within the bone, which eventually leads to recurrences. Therefore, block excision is the most widely accepted form of treatment.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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Figure 11: Photomicrograph showing a odontogenic follicle with squamous metaplasia (A) and mucous pool and mucous production (B)

Figure 12: Postoperative radiograph