INTRODUCTION

Ameloblastomas are common and benign, but locally invasive polymorphic neoplasms that consist of a proliferating odontogenic epithelium.\(^1,2\) The common histological patterns of ameloblastomas have been classified as follicular, plexiform, acanthomatous, granular cell, basal cell, desmoplastic, and keratomatous.\(^2\) Desmoplastic Ameloblastoma is a rare variant of ameloblastoma. The first detailed report on the desmoplastic variant of ameloblastoma (DA) in English literature was given by Eversole, in 1984, who called it an ameloblastoma with pronounced desmoplasia.\(^2\) However, Takigawa et al. and Uji et al. were the early ones to draw attention to this unusual variant, characterized by extensive stromal dysplasia with small compressed nests and strands of odontogenic epithelium.\(^3,4\)

Desmoplastic Ameloblastoma has predilection for occurrence in the anterior regions of the maxilla and mandible. The desmoplastic variant appears as a mixed radiolucent-radiopaque lesion, which resembles a benign fibro-osseous lesion.\(^6\) This unusual variant is characterized histologically by extensive stromal collagenization or desmoplasia with small nests and strands of odontogenic epithelium. This article presents an unusual case of DA seen anterior to the premolar region crossing the midline of the mandible.

CASE REPORT

A 24-year-old male patient had reported to the Department of Oral Medicine and Radiology with a complaint of swelling on the lower left side of the face. On extraoral examination, a single ovoid swelling was noticed in the left parasymphysial region, measuring approximately 3 × 2 cm in size, anterioposteriorly and superoinferiorly. The borders of the swelling were well defined, the skin over the swelling was normal, and the swelling was extending anteriorly up to the symphysis region. The inspection findings were confirmed on palpation, the swelling was hard in consistency, nontender in nature, with localized rise of temperature (Figures 1 and 2).

On intraoral examination, the swelling extended buccally from 41 till the mesial aspect of 36, and lingually the swelling extended with well-defined borders, oval in shape, with a
smooth surface over the swelling. Displacement of 31, 32, and 41 was noticed in relation to the lower anterior teeth [Figure 3].

The inspection findings of the swelling were confirmed on palpation. The swelling was hard in consistency, noncompressible, nonfluctuant, and nontender in nature. The swelling was not fixed to the mucosa and seemed to rise from within the bone. The teeth in the affected area were not mobile or nontender on percussion. An electric pulp test revealed the teeth were vital.

The aspiration of the lesion was unproductive. Based on the patient's chief complaint and clinical examination, a provisional diagnosis of ameloblastoma was made. Further investigative procedures such as radiographs and incisional biopsy were performed to establish a definitive diagnosis of the tumor.

**Radiographic features**

The cross-sectional occlusal radiograph of the mandible revealed a mixed radiolucent-radiopaque lesion with expansion of the buccal and lingual cortical plates [Figure 4]. The panoramic radiograph showed the displacement of 31, 32, and 41 [Figure 5].

The axial section of computed tomography (CT) demonstrated permeative destruction of the buccal bony cortex with a mesh of radio-opaque structures with ill-defined borders, suggestive of osteolysis and sclerotic changes, and the lingual bony cortex demonstrated thinning and partial destruction [Figure 6].

The coronal CT demonstrated the sclerotic expansile destructive lesion with displacement of the teeth [Figure 7].

The incisional biopsy revealed small irregular islands and cords of odontogenic epithelium in a dense collagenous stroma. The odontogenic epithelial islands showed peripherally arranged cuboidal cells with hyperchromatic nuclei. The surrounding dense stroma showed bundles of collagen fibers with irregular areas of osseous trabeculae. These features were suggestive of desmoplastic ameloblastoma [Figures 8 and 9].
The tumor was treated by block resection of the mandible [Figure 10] followed by distraction-osteogenesis with an extraoral distractor [Figure 11]. The patient was comfortable and no recurrence was noticed after one year four months, with regular check-ups.

**DISCUSSION**

Desmoplastic Ameloblastoma has been characterized by WHO as a variant of ameloblastoma with specific clinical radiographic imaging and histological features. They
are more likely to occur in the premolar region of the jaws, with equal predilection to the maxilla and mandible. The present case showed predilection for the anterior mandible. The maxillary tumors are more insidious than the mandibular tumors, owing to the proximity of vital structures like the maxillary sinus. The maxilla forms a weak barrier for the spread of tumors; consequently maxillary ameloblastomas may be able to spread easier and more quickly than mandibular neoplasms.[8] The incidence of DA is 0.9-12.1% among all ameloblastomas.

Wang et al. reported three radiological features of DAs. The unilocular form containing varying amounts of radiopaque islands of material. The multilocular destruction containing irregular or line-like radiopaque areas, and the mixed destruction showing plexiform radiopaque material and unilocular change.[9] This case showed well-defined, mixed, radiolucent and radiopaque structures on the conventional radiograph. Takata et al. attributed the mixed radiological appearance and ill-defined margins of DA to its infiltrative nature.

The CT scan delineates the internal structure of the lesion more accurately and is particularly helpful in determining margins and extension into adjacent structures. Root resorption is a common finding in DA, but was not seen in our case, although displacement of teeth was noticed in the mandibular anterior teeth.

The histological appearance of DA is a characteristic. The stromal connective tissue shows extensive dense collagenization, with occasional areas of bone. The odontogenic islands are irregular and appear to be compressed by the stromal tissue, giving rise to a stretched out, tail-like appearance. This variant exhibits odontogenic islands made up of peripheral flattened, cuboidal or squamous cells with rare foci of palisaded columnar and central hypercellular spindle or polygonal cells. This case has presented with the typical histopathological features of DA. Spicules of mature lamellar bone trabeculae have been reported to be in intimate contact with the tumor, and invasion has been demonstrated.

Various immunohistochemical studies have reported DA tumor cells, showing variable expression of S-100 protein and desmin.[10]

Desmoplastic Ameloblastoma may exhibit a more aggressive behavior than other types of ameloblastomas. The factors that suggest the aggressiveness are:
1) A potential to grow to a large size.
2) The diffuse radiographic appearance and the histological finding of bone invasion.
3) DA may have a propensity to recur with a frequency equal to that of ameloblastomas treated with curettage. Curettage leaves islands of tumor within the bone, which later manifest as a recurrence. Therefore, block excision is the most widely used treatment to avoid recurrence.

**Conclusion**

Thus to conclude, a desmoplastic ameloblastoma showing a radiographic appearance resembling fibro-osseous lesions, many times needs a perfect diagnosis based not only on the clinical and radiological appearance, but also on the histopathological findings. The lesion in the present case deviates from the usual desmoplastic variant of ameloblastomas in terms of site and radiological appearance. Therefore, it is essential on the oral diagnostician’s part, to establish a definitive diagnosis, to prevent any error in formulating treatment modalities.

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