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Case Report

Desmoplastic Ameloblastoma – A Case report and review of literature

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INTRODUCTION

Desmoplastic ameloblastoma is a rare variant of ameloblastoma, a benign odontogenic tumor that typically arises from the epithelial lining of the tooth-forming structures. Desmoplastic ameloblastoma is characterized by a unique and diverse growth pattern and histopathological features[1].

The word "desmoplastic" refers to the extensive presence of dense fibrous connective tissue that surrounds the tumor cells, giving them a mature, differentiated, and well-organized appearance. Desmoplastic ameloblastoma is a rare variant of ameloblastoma and accounts for approximately 4% to 13% of all ameloblastoma cases. This variant affects both genders equally and has been reported to occur in people of all ages ranging from 6 to 83 years, but the highest incidence is between 30 to 60 years of age[2].

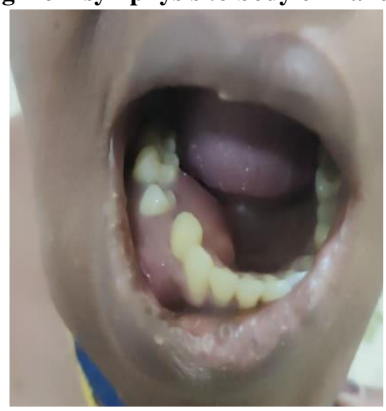
Desmoplastic ameloblastoma has been reported to occur primarily in the posterior mandible, followed by the anterior mandible, maxilla, and posterior maxilla, in descending order of frequency[3].

Due to the rarity of the tumor and its similarity in appearance to other more common fibrous lesions, the diagnosis of desmoplastic ameloblastoma can be challenging. A definitive diagnosis requires careful histological evaluation by an experienced pathologist, aided by appropriate immunohistochemical and molecular techniques[4].

CASE REPORT

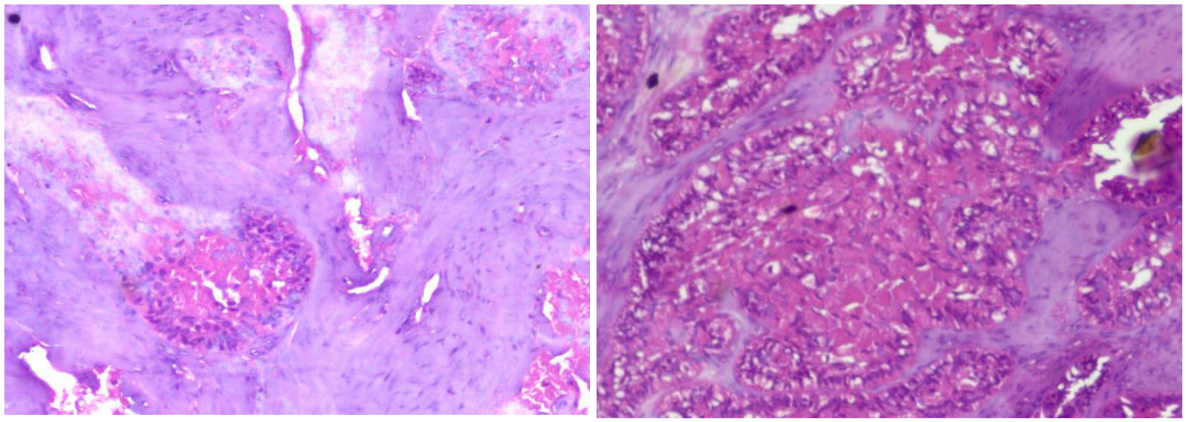
A 54 Year female patient present to the Hospital with the chief complaint of swelling in the right side of lower jaw since 6 months. On intra oral examination, the swelling is ovoid is shape, firm in consistency, surface texture is irregular, erythematous ,and measuring about 3x3cm is size extending from symphysis to the body of mandible (Fig 1).

Fig. 1 The swelling is ovoid is shape, firm in consistency, surface texture is irregular, erythematous ,and measuring about 3x3cm is size extending from symphysis to body of mandible.



The lesion is excised and the samples are sent to the Department of Oral Pathology & Microbiology. On macroscopic examination of the received specimen which was blackish brown in colour, firm in consistency and it was sectioned into 3 equal halves and were labeled as A, B, & C. A, B, & C were measuring about 2.2x1.5cm, 2x1cm, 0.9x0.8cm in size respectively. A, B, C were kept for processing. On microscopic features, the hematoxylin and eosin stained soft tissue section A exhibits connective tissue stroma with few ameloblsatic islands with peripheral palisading follicles and bone with resting and reversal lines, marrow spaces and with presence of osteoblastic rimming(Fig 2).

Fig. 2 The hematoxylin and eosin stained soft tissue section A exhibits connective tissue stroma with few ameloblsatic islands with peripheral palisading follicles.



The connective tissue stroma also exhibits areas of heamorrhage, extravasated red blood cells, micro orgamisms and fibrin. The connective tissue stroma is hyalinised which is getting calcified. Section B exhibits numerous capillaried and follicles. Section C exhibits hyalinised connective tissue stroma pinching into follicles.

Based on overall clinical, radiological and its orientation with histopathological features the final diagnosis is suggestive of "DESMOPLASTIC AMELOBLASTOMA"

DISCUSSION

The tumor can occur in any part of the mandible or maxilla and is usually found in adults between the ages of 30 and 60 years. Clinical features of desmoplastic ameloblastoma depend on the location and size of the tumor and may include pain, swelling, difficulty in opening the mouth, and loosening of the adjacent teeth[5,6]. The histopathology of desmoplastic ameloblastoma is characterized by a dense fibrous stroma and strands or nests of odontogenic epithelium[7]. The tumor cells of desmoplastic ameloblastoma are small, spindle-shaped, and arranged in long, linear strands or cords within the fibrous connective tissue.

The stroma or fibrous tissue is mature and well-developed, forming a background and a net-like mesh that surrounds the tumor cells. Unlike conventional ameloblastoma, the tumor cells in desmoplastic ameloblastoma are not arranged in a typically circular pattern around a central stellate reticulum-like structure. The cells have a basaloid appearance with scanty cytoplasm, hyperchromatic nuclei, and prominent nucleoli. Mitotic activity is typically very low to absent.

In some cases, the tumor cells may fuse together to form interconnected anastomosing strands or cords. These strands/cords of the tumor are further surrounded by a well-vascularized fibrous stroma. The presence of these strands and cords is what provides the "desmoplastic" or "fibrotic" appearance to the ameloblastoma[8].

Additionally, areas of calcification or ossification can be seen in up to 50% of cases. The calcification can occur within the neoplastic epithelial tissue or within the fibrous stroma surrounding it[9].

In summary, the histopathology of desmoplastic ameloblastoma is characterized by the presence of a dense, fibrous connective tissue that surrounds the tumor cells. Tumor cells are small and spindle-shaped, with linear strands or cords found in concert with the fibrous stroma. The presence of calcification or ossification is common. This unique presentation distinguishes desmoplastic ameloblastoma from other variants of ameloblastoma.

TREATMENT

Treatment of desmoplastic ameloblastoma involves surgical excision of the lesion, and in most cases, the removal is curative. Recurrences of this type of tumor are rare compared to other types of ameloblastomas[10].

SUMMARY AND CONCLUSION

In summary, desmoplastic ameloblastoma is a rare variant of the benign odontogenic tumor that has a unique histopathological appearance characterized by a densely fibrous connective tissue and elongated spindle cells. It occurs in adult individuals and is treated by surgical excision[11].

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