

CASE REPORT

Desmoplastic Fibroma of Condyle: A Case Report

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ABSTRACT

Desmoplastic fibroma (DF) is a benign intraosseous neoplasm that is recognized as the intraosseous counterpart of soft tissue fibromatosis in both gnathic and extragnathic sites. In this case report, we define the clinicopathological and radiographic features of DF of the mandible in a 28-year-old male, who presented to the outpatient department with a history of a slowly expanding painless mass in the left mandibular posterior region. Thus, we present a classic case of DF exhibiting characteristic features along with a review of the literature.

Keywords: Desmoplastic fibroma, Fibromatosis, Gnathic.

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INTRODUCTION

Desmoplastic fibroma (DF), a benign locally aggressive lesion of the bone, is recognized as an intraosseous counterpart of soft tissue fibromatosis, is usually seen affecting the long bones and pelvis, and only occasionally presents itself as a jaw lesion.^[1] The mandible is the most commonly affected when compared to the maxilla and the cranium in the head and neck region.^[2] The cause for DF is unknown and is stipulated to have a varied pathogenesis ranging from genetic, endocrine, and traumatic factors to an exuberant reactive proliferation.^[3] When differentiating it from other neoplasms that behave aggressively, a history of expansion or perforation of the cortical plates along with the histopathological confirmation would be a pointer in the right direction.

CASE REPORT

A 28-year-old male patient visited the Department of Oral and Maxillofacial Surgery, with the chief complaint of slowly growing painless swelling in the right lower back tooth region. The swelling was hard and non-tender on palpation. The patient's past medical history was non-contributory. Clinical extraoral examination revealed the expansion of the right inferior border of the mandible and intraoral examination revealed a solitary bony hard swelling measuring about 3.0 cm × 4.0 cm in size with obliteration of the left buccal vestibule in relation to 46 and 47. A right lateral oblique view of radiograph and computed tomography scan [Figure 1] showed multilocular radiolucencies with fine trabeculations leading to a soap bubble appearance. The hematoxylin and eosin stained tissue section showed hypo- and hypercellular areas with the proliferation of plump fibroblasts arranged in interlacing fascicles and dense collagen. Fibroblasts were not atypical, and mitosis figures were absent. Focal areas of the section also revealed dense collagenous stroma with foci of hyalinization. A final diagnosis of DF was arrived at after histopathological examination. Areas of moderate cellularity were resected through a partial hemimandibulectomy, and condylar prosthesis was placed in the defect [Figure 2]. There has been no recurrence to date and mouth opening is improved [Figure 3].

DISCUSSION

In 1958, Jaffe introduced the term "DF of bone" to describe a densely fibrous entity composed of fibroblasts among rich collagen fibers, resembling the familiar abdominal desmoid tumor.^[1] Griffith and Irby reported, in 1965, the first gnathic involvement of a DF involving the mandible.^[2] According to the World Health Organization, its low to variable cellularity, ovoid to elongated nuclei, and lack of pleomorphism and mitoses categorize it as a benign tumor.^[3] DF can involve any bone but is the most often found in the mandible (22%), followed by the femur (15%), pelvic bones (13%), radius (12%), and tibia (9%).^[3,4] The incidence of DF is estimated to be 0.1% of all primary bone tumors.^[3] Approximately 84% of patients with DF of the gnathic bones are under 30 years of age with a mean age of 16 years. Gnathically, the majority of cases involve the mandible (84%), and in either jaw bone, a

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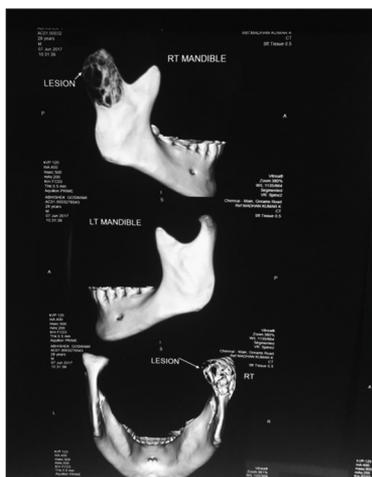


Figure 1: Pre-operative computed tomography image



Figure 2: Intraoperative picture with condylar prosthesis in the defect



Figure 3: Clinical image showing post-operative mouth opening

posterior location is favored. DF is, possibly associated with trauma, endocrine factors, genetic aberrations, or multifactorial etiology have been suggested.^[4,5] Mutations in both β -catenin and adenomatous polyposis coli genes have been implicated in the pathogenesis of desmoid-type fibromatosis.^[6] Fluorescent *in situ*

hybridization analyses have demonstrated trisomy 8 and 20 as nonrandom aberrations in benign fibrous conditions of both bone and soft tissue.^[7] Moreover, DFs of gnathic bones have been diagnosed in patients with the hamartoneoplastic syndrome–tuberous sclerosis, in which they are believed to represent intraoral manifestations of this multisystem genetic disorder.^[8,9] Although many believe the DF to be the bony counterpart of soft tissue desmoid-type fibromatosis, others have suggested that it may be a separate entity, as there have been recent discrepancies with β -catenin staining for DFs.^[10] Cortical perforation and soft tissue involvement require wider margins of resection. Recurrence rates following excision and enucleation have ranged from 20 to 40% and curettage as high as 70%.^[4] It has been noted that tumors with higher cellularity tend to recur more often than those with lower cellularity. Some authors have recommended wider resection in those tumors showing aggressive behavior as demonstrated by cortical perforation, soft tissue involvement, and moderate-to-high cellularity. Following surgical treatment, a follow-up period of <3 years is recommended.^[4]

CONCLUSION

The DF is a rare, benign neoplasm that can occur in gnathic bones and has overlapping histopathologic features with other benign and malignant entities characterized by spindle cell proliferations, trabeculae of bone, and little to no cytological atypia or mitotic activity. Distinguishing DFs from Fibrous dysplasia, low-grade fibrosarcoma, and low-grade osteosarcoma has important prognostic significance and remains a difficult process for the pathologist. As DFs may be locally aggressive and recur if inadequately or conservatively excised, wide resection or *en bloc* resection with long-term follow-up continues to be the treatment of choice.

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