CASE REPORT

Desmoplastic Fibroma-A Rare Case Report

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ABSTRACT

Desmoplastic fibroma (DF) is a benign intra-osseous neoplasm, that is, recognized as the intra-osseous counterpart of soft tissue fibromatosis in both gnathic and extra-gnathic sites. It has a propensity for locally aggressive behavior and local recurrence. An occurrence of intra-osseous lesion other than that of odontogenic origin is rare in the jaws. In this case report, we define the clinico-pathological and radiographic features of DF of the mandible in a 35-year-old female, who presented to the Outpatient Department with a 3-year 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.

Key words: Desmoplastic fibroma, fibromatosis, gnathic

INTRODUCTION

Desmoplastic fibroma (DF), a benign locally aggressive lesion of the bone is recognized as an intra-osseous counterpart of soft tissue fibromatosis and is usually seen affecting the long bones, pelvis and only occasionally presents itself as a jaw lesion. Mandible is most commonly affected when compared to the maxilla and the cranium in the head and neck region. A systematic literature search of the PubMed database of national library of medicine using "DF" and "Mandible" as keywords revealed a total of 57 published cases occurring in the mandible alone from the year 1969–2014. 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. 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 35-year-old female patient visited the Department of Oral Medicine and Radiology, with the chief complaint of slowly growing painless swelling in the left lower back tooth region since 3 years [Figure 1]. The swelling was hard and nontender on palpation. The patient's past medical history was noncontributory. Clinical extra-oral examination revealed expansion of the left inferior border of the mandible and intra-oral examination revealed a solitary bony hard swelling measuring about 4.0 cm × 5.0 cm in size with obliteration of the left buccal vestibule in relation to 37 and 38 [Figure 2].

A left lateral oblique view of radiograph showed multilocular radiolucencies with fine trabeculations leading to a soap bubble appearance extending from the left angle of the mandible to the mesial root of the mandibular left first molar. No displacement of teeth or resorption of the root was seen [Figure 3].

A computerized tomography (CT) scan demonstrated buccal and lingual cortical plate expansion and a soap bubble appearance [Figure 4].

Surgical excision of the lesion was done under general anesthesia and sent for histopathological examination.

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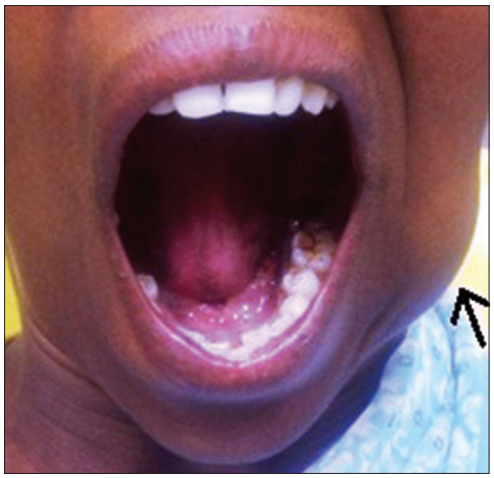

Figure 1: Clinical image showing swelling of left side of lower jaw

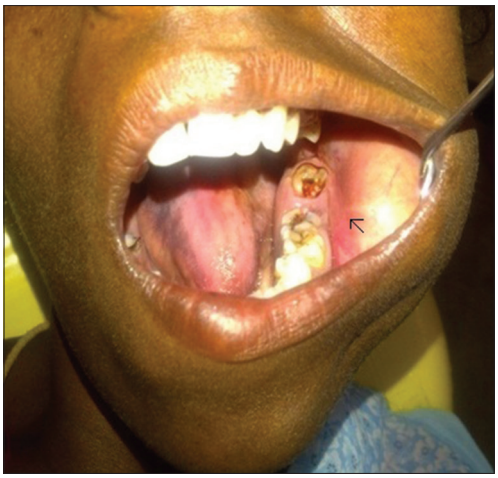

Figure 2: Intra-oral photograph showing obliteration of the left buccal vestibule in relation to 37, 38

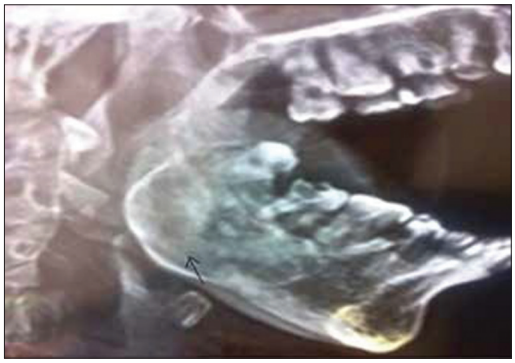

Figure 3: Left lateral oblique view of radiograph showing multilocular radiolucencies

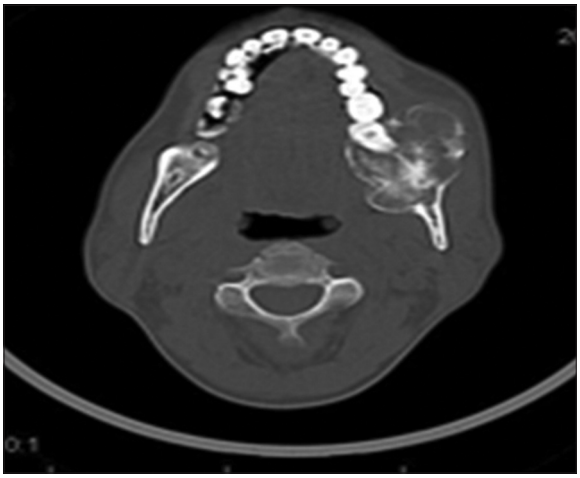

Figure 4: Computed tomography scan showing buccal and lingual cortical plate expansion

The hematoxylin and eosin stained tissue section showed hypo and hyper-cellular areas with proliferation of plump fibroblasts arranged in interlacing fascicles and dense collagen. Fibroblasts were not atypical and mitosis figures were absent [Figures 5-7]. Focal areas of the section also revealed dense collagenous stroma with foci of hyalinization [Figure 8].

A final diagnosis of DF was arrived at after histopathological examination.

DISCUSSION

DF is a rare, locally aggressive myofibroblastic benign tumor of connective tissue origin. As an intra-osseous lesion, it commonly occurs in the metaphysis of long bones.[1] DF was first described by Jaffe in 1958[2] and named as DF. In 1965, the first report about a DF of the jaw was presented by Griffith und Irby.[3]

The Histologic criteria for DF as defined by the World Health Organization is "a rare benign bone tumor composed of spindle-shaped cells with minimal cytological atypia and abundant collagen production."[4] Desmoid tumor also called as "aggressive fibromatosis," was described before DF. Desmoid tumors can occur both intra-abdominally and extra-abdominally. About 69% of desmoid tumors are abdominal; the extra-abdominal variety occurring in the bone is the DF.[5] Although DF can affect any age group, most patients are affected in the first three decades of life.[5] In our case, the patient was a 35-year-old female. The average age of patients at the time of the final diagnosis is 15.1 years.[3] Metaphysis of long bones especially tibia, scapula and femur are the most frequent sites of involvement. Mandible is the fourth most common site of involvement and sex predilection remains unclear.[6,7] In the mandible, the lesions tend to occur posteriorly at the ramus-angle region. This is similar to our case report, where the lesional tissue is in relation to the second and third molars. DF comprises 0.06% of all osseous tumors and 3% of all benign bone tumors.[8]

The symptoms are nonspecific including diffuse, moderate pain in the region of the tumor, both at rest and on movement or when bone bears any weight.[9] In the maxillofacial region, DFs usually are painless, slow-growing firm masses.[10]

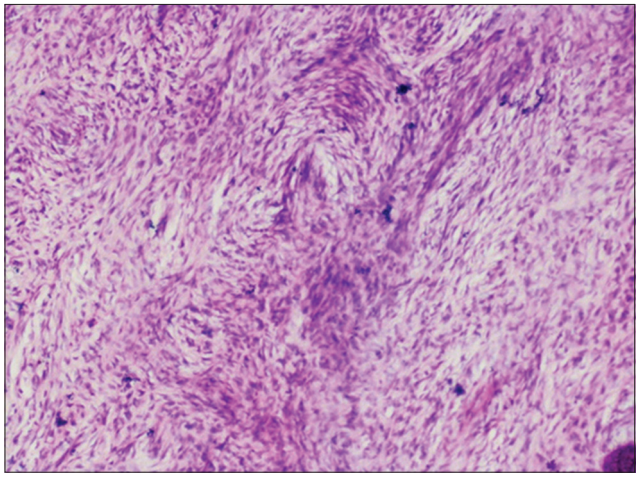

Figure 5: Photomicrograph showing hypo-cellular and hyper-cellular areas with spindle cells arranged in interlacing fascicles (H&E stain, ×10)

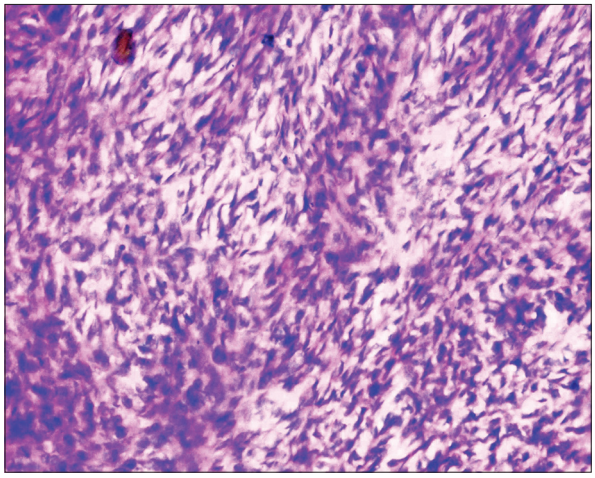

Figure 6: Photomicrograph showing hyper-cellular area with proliferating plump fibroblasts, (H&E stain, ×20)

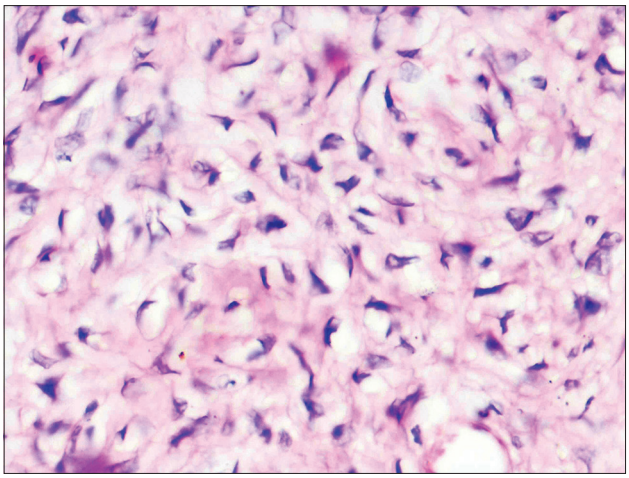

Figure 7: Photomicrograph showing proliferating plump, spindle-shaped fibroblasts in a collagenous stroma (H&E stain, ×40)

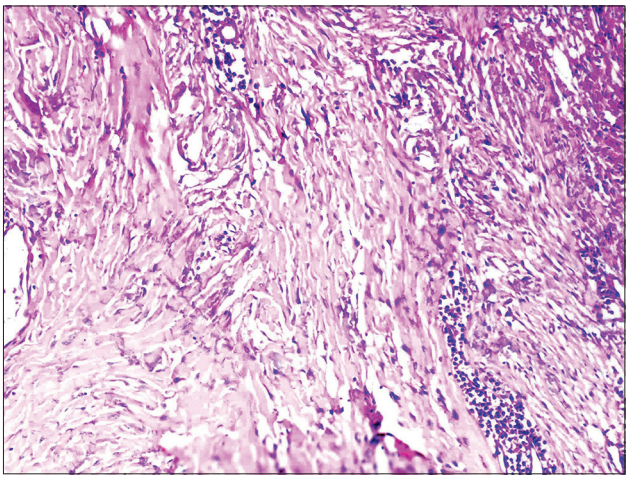

Figure 8: Photomicrograph showing a focus of dense collagenous stroma with focal areas of hyalinization (H&E stain, ×10)

A similar history of a painless, slow-growing lesion was elicited by the patient in our case report.

Radiographic appearance may vary from uni-locular to multi-locular, with or without expansion or perforation of cortical plates according to Frick *et al*. Radiographs showed osteolytic lesions with coarsened ridge-like trabeculae in 63% of cases, osteolytic lesions in 24% of cases and mixed lytic and mildly sclerotic lesions in 13% cases. About 53% of cases showed cortical breaching. CT revealed radiolucent (65%) or mixed radiolucent and mildly sclerotic (35%) matrix patterns. Cortical destruction was seen in 88%. The CT in our case clearly showed cortical expansion without any resorption or displacement of adjacent teeth.

T1-weighted sequences in MRI showed that the signal intensities within the lesions were isointense or hypointense to adjacent normal muscle. Significant T2 shortening was noted on T2-weighted images.[11]

The hypocellular areas of the tumors with abundant collagen are responsible for the areas of T2-shortening while hyper-cellular parts filled with fibroblasts or necrotic areas are responsible for the higher intensity parts within the lesions.[12]

On gross examination, the desmoid tumor appears as firm, rubbery, white, nonencapsulated fibrous growth.[1]

Histologically, the DF contains mature fibrous connective tissue, low to variable cellularity and spindle-shaped fibroblasts/myofibroblasts with uniform long nuclei in an abundant stroma of collagenous matrix lacking cellular pleomorphism, nuclear hyperchromatism or mitoses. The lesions are noncapsulated with poor demarcation within the bone.[1,5,8,10]

The differential diagnosis to be considered histologically would be spindle cell tumors of which low-grade fibrosarcoma is the most important. Tumors such as fibrous histiocytoma, fibrous dysplasia, or a low-grade intra-osseous osteosarcoma; tumor-like lesions such as aneurysmal bone cyst and the

juvenile bone cyst can also present with a similar clinical picture. Though fibrosarcoma exhibits a highly cellular stroma along with high grades of polymorphism and mitosis, the low-grade variant shows a collagen rich tissue with low cell count and no mitotic activity. This is very similar to the histopathological picture of the DF and a definitive diagnosis is possible only with postoperative clinical development.[5]

Hauben *et al*. found that DF has no immunoreactivity of CD117, estrogen and progesterone receptors; and 50% cases showed positivity for muscle-specific markers and the β-catenin pathway does not seem to have the same essential role in the tumorigenesis of DF, as it has in desmoid type fibromatosis.[13] Böhm *et al*. found that the majority of the tumor cells express the mesenchymal marker vimentin and no immunoreactivity for antidesmin and anti-S-100 protein.[14]

DF of maxilla or mandible with extra-osseous extensions is treated with complete excision including a margin of uninvolved soft tissue. These tumors are locally aggressive and can recur with a subtotal resection. The recurrence rate of about 40–47% is seen in lesions treated by curettage or intra-lesional resection[6,9,15] making follow-up a necessity.

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Conflicts of interest

There are no conflicts of interest.

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