

Desmoplastic Fibroma - Case Report

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Abstract

Desmoplastic fibroma (DF) is considered as an extremely rare locally aggressive bone tumor with the clinical presentation of pain and swelling in the affected site most likely the metaphysic of long bones. Most of the times, a clinical confusion may occur due to the histological and biological similarity with extra abdominal desmoids tumor of soft tissue. In this case report, we define the clinico-pathological and radiographic features of DF of the mandible in a 15-year-old male, who presented to the Outpatient Department with swelling and pain over right wrist. 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) is a benign locally aggressive lesion of the bone is recognized as an intra-osseous counterpart of soft tissue fibromatosis where more infiltrate and locally aggressive was observed [1] and is usually seen affecting the long bones, pelvis and only occasionally presents itself as a jaw lesion [2]. DF represent one of the most uncommon bone diseases as it has an incidence of nearly 0.1% of all primary bone tumours [1, 3]. Metaphysis of long bones and mandible are the most commonly affected when compared to the maxilla and the cranium in the head and neck region [4].

The clinical presentations are non-specific and usually presents as pain over the affected area and rarely palpable mass was observed [5]. The etiology of DF is still unknown by have varied in pathogenesis ranging from endocrine, genetic and traumatic factors to an exuberant reactive proliferation. The disease related to gender specific is not remaining clear but higher rates are recorded among female patients than males. Pathologically the tumor was composed mainly of bland looking spindle cells in abundant collagenous stroma, accompanied with areas of myxofibrosarcomatous and malignant fibrous histiocytomatous components [3].

In most cases, the radiographic examination may provide nice clue where the tumor presenting as expanding, well delineating radiolucency with either

unilocular or multilocular and destroyed cortex. This pattern of bone destruction may sometimes report as malignancy [6]. To overcome that situation, the histological examination is considered as an identical for appropriate diagnosis. This study presents the case of a 36 year old female, who complained of increasing pain for over one and half years.

ABOUT THE CASE

A 15 year old male patient visited the outpatient department with pain and swelling in the right wrist. The pain started one year earlier, at which point the patient sought help from a local surgeon. The pain became worse with movement. He may think that the pain may be due to heavy works and holding heavy materials. During this period, the patient also had intermittent fever with a range of 37 to 38.6°C at daily maximum temperature. He had no history of diabetes, hypertension, hyperlipidemia, asthma, trauma or any major surgeries.

Visceral etiologies should be considered for multiple joint pains and fever which include infectious diseases (viral, bacterial, fungal, or parasitic diseases), crystal-induced arthritis (gout or pseudogout), endocrine disorders (hyperthyroidism, hypothyroidism, or hyperparathyroidism), collagen vascular diseases (rheumatoid arthritis, systemic lupus erythematosus, various vasculitis, or adult Still's disease) or seronegative spondyloarthropathies (psoriatic arthritis, ankylosing spondylitis, enteritis-associated arthritis, or

Behcet disease). Taking detailed clinical history is necessary for focusing to potential pathogens. Review of systems is mandatory for gathering additional important information. Signs and symptoms of malignancy were not present. Initially the patient was treated with physical therapy, chiropractic treatment and pain control medications.

The surgery was performed with the patient under general anaesthesia and the tissue was removed from the right wrist. The tissue was brown in color with hard and soft areas of consistency (Figure-1). The removed mass was sent for further histological examination to the department of pathology.



Fig-1: Intraoperative photograph showing the removed tissue and it has a brown color and hard as well as soft areas of consistency

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

(Figure-2). 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.

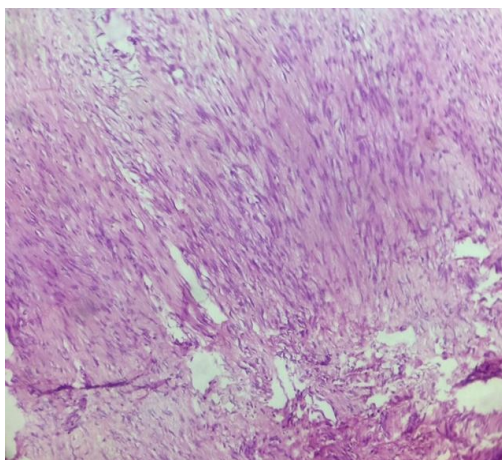


Fig-2: Photomicrograph showing hypo-cellular and hyper-cellular areas with spindle cells arranged in interlacing fascicles (H&E stain, ×10)

For further diagnostic workup an open biopsy of the lesion was performed one week later. The histological examination of the tissue demonstrated the formation of abundant collagen fibers and spindle shaped cells with medium to low cell density. The nuclei were without pleomorphism and mitotic activity. In immuno-staining the nuclei of the tumor cells were positively labeled for the adhesion protein β -catenin. In addition there was a partially positive staining recognized for S-100. Morphology and immunophenotype were consistent with either aggressive fibromatosis or desmoplastic fibroma, both of which express β -catenin. Because of its intraosseous

location the working diagnosis of desmoplastic fibroma was made. Accordingly the indication for operative excision *via* curettage and grafting was established.

DISCUSSION

Various literatures suggested that DF is considered as a rare, locally aggressive myofibroblastic benign tumor of connective tissue origin [7-9]. The histopathological description of the DF is “a rare benign bone tumor composed of spindle-shaped cells with minimal cytological atypia and abundant collagen production [10]. Before DF, desmoid tumor was also called as aggressive fibromatosis. In general, desmoid

tumors are abdominal; the extra-abdominal variety occurring in the bone is the DF [1].

Eventhough DF affects all age groups; but predominantly patients are affected in the first three decades of life [1]. In our case, the patient was a 15-year-old male. The average age of patients at the time of the final diagnosis is 15.1 years [9]. Metaphysis of long bones especially tibia, scapula and femur are the most frequent sites of involvement. 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 [11].

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 [12]. In the region, DFs usually are painless, slow-growing firm masses [13] where similar history of a painless, slow-growing lesion was elicited by the patient in our case report.

The characteristics of DF in CT imaging have been well described in this case report [14]. In general, the DF was described as an osteolytic lesion with destruction of the cortical bone, minor pseudotrabeclulation and marginal sclerosis [1]. These findings were also present in our case. The preoperative CT-scans showed increased tumor size indicating bone instability; further there is no extraosseus growth. The study revealed the radiographic characteristics of DF in 83 patients from case reports, reported that only in 29% of patients cortical breakthrough was present [15]. If cortical damage is suspected a CT-scan should be performed for quantifying cortical bone destruction. Further in the case of extraosseus tumor growth is suspected, then MRI is preferred for the evaluation of anatomical landmarks [1].

The differential diagnosis is observed in imaging studies and has some confusion in finalizing DF; the tumors like fibrous dysplasia, giant cell tumor, simple bone cyst, aneurysmatic bone cyst, chondromyxofibroma, non-ossifying fibroma, eosinophilic granuloma, adamantinoma and metastases are mimicking the DF [16-18]. The histological picture of DF is dominated by abundant collagen fibers and low density of cells. The nuclei are long and spindle shaped. Signs of mitosis are usually not present. The histological differential diagnosis includes the spindle cell tumors and tumor like lesions, including juvenile bone cyst and aneurysmatic bone cyst. The most important histological differential diagnosis is low-grade fibrosarcoma.

Fibrosarcoma typically presents with high cell density, high grade of polymorphism, and a high rate of mitosis. In rare cases of low-grade fibrosarcoma mitotic activity is not present and the tissue is collagen rich with a low cell count, such that differentiation between

desmoplastic fibroma and fibrosarcoma is not possible. In these cases the postoperative clinical development helps to establish the definite diagnosis [18-20]. The treatment for DF is highly depends on the affected site and on the aggressive nature of the lesion further analyzed that the resection has the lowest rate of recurrence [21].

In isolated intraosseus lesions without the appropriate evidence of extension in adjacent soft tissues and when resection is associated with a higher risk because of anatomical conditions, a considerate risk-benefit-analysis should be carried out and thorough curettage may be an adequate management as it reduces operation time with a lower risk of infection and facilitates faster recovery. Once the decision has been made in accordance with the patient to perform curettage, close postoperative observation, including clinical and radiographic examinations, is necessary to detect a recurrent lesion as early as possible. The higher risk of recurrence has to be well communicated with the patient preoperatively and has to be included in the decision finding process.

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