Case Report

Desmoplastic fibroma: A rare entity

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Contribution Details: All the five authors have contributed to concept, literature search, data acquisition, data analysis, manuscript editing and review.

[Received-06/08/2014, Accepted-05/09/2014]

ABSTRACT:
Desmoplastic fibroma (DF) is an extremely rare benign intraosseus, locally aggressive bone tumor with an incidence of 0.11% of all primary bone tumors. The most common sites of involvement are the mandible and the metaphysis of long bones. Histologically and biologically, desmoplastic fibroma mimics extra-abdominal desmoid tumor of soft tissue. A 25-year male presented to outpatient department with increasing pain in left hip region since 2 months. Imaging investigations revealed a multiloculated cystic lesion in left femoral region. Curettage material was sent to our department. On histopathology, sections examined showed spindle-shaped cells without cytological atypia and abundant collagen production confirming the diagnosis of desmoplastic fibroma.

Key words: Desmoplastic fibroma, rare bone tumor, benign bone tumor

INTRODUCTION:

In 1958, Jaffe first described desmoplastic fibroma of the bone as a distinct entity when he documented five cases of a previously unclassified osseous fibrous tumor that was histologically similar to abdominal desmoid tumor[1]. World health organisation (WHO) in 2002 defined desmoplastic fibroma as a rare benign tumor characterized by formation of abundant collagen fibers by the tumor cells[2]. The tissue is poorly cellular and nuclei are oval or elongated. The cellularity, pleomorphism, and mitotic activity that are the features of fibrosarcoma are lacking. The clinical presentation of desmoplastic fibroma is non-specific and usually presents as pain over the affected area and occasionally as a palpable mass. Desmoplastic fibroma has recurrence rate of 37% to 72%. Resection of the affected bone is the preferred therapy and ideally an en bloc resection[3]. Cases in which anatomical consideration limit resection, alternative therapy has been curettage and reported to achieve recurrence free survival upto 9 years[3]. Recognition of desmoplastic fibroma is important because on radiology and histology, the lesion may be mistaken for an indolent, benign fibrous lesion or more aggressive...
spindle-cell sarcoma. Because of its relative rarity, desmoplastic fibroma of the bone has been described in only a few small series in the orthopedic, pathology, and radiology literature. Herein we report a case of desmoplastic fibroma in a 25 year male who presented with pain in left hip region of 2 months duration.

CASE REPORT:
A 25 year male presented to orthopaedic out-patient department with pain of increasing severity in the left hip region since 2 months. Patient complained of difficulty in walking. On physical examination the patient had tenderness to palpation over his left hip joint. The patient was in good state of health. Peripheral motor and sensory function were intact. Patient’s haematological investigations were within normal limits. MRI-scan of left hip joint was performed and revealed a sharply defined irregular multiloculated cystic lesion of 50x36 mm in left femoral neck and intertrochanteric region. A thick rim of increased signal intensity was seen around the lesion. Left femoral head demonstrated normal marrow signal intensity pattern and contour. Articular cartilage and periarticular soft tissue appeared normal. There were no signs of malignancy. So most likely diagnosis of an aneurysmal bone cyst was made. Under general anaesthesia patient was treated by thorough curettage. Intra-operatively periosteum was intact. The lesion was greyish white, solid and tough in consistency. Curettage material measuring 4.5x4x0.8 cm was sent for histopathological examination. Microscopically the lesion was composed of interlacing fascicles of mature, moderately cellular fibrous tissue. The cells were spindle shaped & embedded in coarse strands of collagen (figure 1). Nuclei were dense with no nucleoli. Mitosis was not seen. Pleomorphism, atypia and hyperchromatism were absent. Cells and collagen fibres were arranged in parallel fashion and in bundles crossed each other, but the “herring bone” pattern of fibrosarcoma was not evident. Hence a final diagnosis of desmoplastic fibroma was made on these features.

DISCUSSION:
Desmoplastic fibroma of bone is a rare benign tumor consisting of thin, wavy fibroblast set in an abundant matrix of collagen fibers. Jaffe in 1958 described the first case of desmoplastic fibroma of bone. WHO in 2002 defined desmoplastic fibroma as a rare, benign bone tumour composed of spindle cells with minimal atypia and abundant collagen production. The sites of predilection are the mandible & long bones, but other sites such as the scapula and os calcis may be involved. It accounts for approximately 0.1% of all primary bone tumours in the body and is most commonly seen in the mandibles of adolescents and young adults.[2]. In a review by BoEHm et al in 1996 of 184 published cases of desmoplastic fibroma the reported mean age of patients with desmoplastic fibroma was 23 years with range from 15 to 75 years[4]. Age of patient in present case is 25 year which nearly correlate with mean age reported by BoEHm et al[4]. Sex predilection remains unclear while some authors report higher rates of desmoplastic fibroma in female patients and others report no detectable sex predilection[3]. Clinical presentation of desmoplastic fibroma is nonspecific and usually include pain over affected area relatively late in the clinical course and occasionally a palpable mass. Because of the non-specific clinical symptoms and absence of palpable mass one has to rely on imaging studies for provisional diagnosis. It may present as an effusion if near a joint. Only 12% patients present with a pathological fracture[5]. No pathological fracture was seen on imaging studies in the present case.

The cellular structure and the morphological arrangement of desmoplastic fibroma are similar to those of aggressive fibromatosis of soft tissues. Plain X ray shows an osteolytic, expansile, medullary lesion with well defined sclerotic margins. The oval tumor is often found in the metaphysis aligned with the long axis of the bone. There is usually thinned cortex and the fine intra-lesional trabeculae give a lobulated appearance that is described as "soap-bubble"
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A CT scan is useful to further demonstrate cortical breakthrough. MRI demonstrates the separation of the intraosseus tumor from the bone. The radiological differential diagnosis includes benign lesions such as non-ossifying fibroma, giant cell tumor, UBC (unicameral bone cyst), ABC (aneurysmal bone cyst) and fibrous dysplasia. Bone tumors in younger adults, that can present as a lytic multiloculated tumor that can expand the cortex include giant cell tumor, chondromyxoid fibroma & aneurysmal bone cyst. A desmoplastic fibroma often behaves in an aggressive manner and macroscopically has a firm consistency with well-defined advancing surfaces that may extend into surrounding soft tissue[7]. The most important histological differential diagnosis is low-grade fibrosarcoma. Fibrosarcoma typically presents with high cell density, high grade pleomorphism, and high rate of mitosis. In rare cases of low grade fibrosarcoma mitotic activity is not present and the tissue is rich with 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. Grossly, desmoplastic fibroma has a grayish to yellowish white color and a rubbery consistency. The edges are irregular, round and blunt. The tumor has occasional cystic foci with clear fluid. Microscopically, the tumor has interlacing bundles of dense collagen and low cellularity. The fusiform cells that are present have no atypia and the nuclei are ovoid or elongated. The differential diagnosis of the tumor includes spindle cell tumors, specially low grade fibrosarcoma. Desmoplastic fibroma does not have the cellularity, mitotic activity or pleomorphism of a fibrosarcoma but the distinction can be difficult and is sometimes made clinically. The edge of the tumor may resemble fibrous dysplasia, but under polarized light lamellar structures are obvious. Differential diagnosis from malignant spindle-cell lesions of bone is important because the treatment of choice for desmoplastic fibroma of bone is simply excision with a thin layer of healthy tissue. In the present case absence of nuclear atypia and mitosis ruled out the possibility of low grade fibrosarcoma. To the best of our knowledge 271 cases of desmoplastic fibroma have been published in the literature till date[3]. Of these 271 cases, 30 were localized in femur diaphysis and metaphysis. This is the only case of desmoplastic fibroma which occurred in neck of the femur which makes it unique. The literature reports the most common treatment for desmoplastic fibroma being a block resection or wide local excision, with enucleation rarely used[8]. The treatment however strongly depends on the region of the body that it affects and on the aggressive nature of the lesion. If possible resection should be performed as it has lowest rate of recurrence[4]. The infrequent use of enucleation is due to the concern that recurrences rates are higher, approximately 20-40%[7,8]. Once the operation is the operation is performed close post-operative observation including clinical and radiographic examination is necessary to detect a recurrent lesion as early as possible. A 3 year follow up period is recommended.

NO CONFLICT OF INTEREST EXISTS; NO FINANCIAL DISCLOSURE

REFERENCES:


Legends:
Figure 1: Interlacing fascicles of spindle cells along with bony trabeculae (H&E 40X).