

Periosteal Desmoplastic Fibroma of Radius: A Rare Bone Tumor

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DEAR SIR

Desmoplastic fibroma is an extremely rare tumor of bone with a reported incidence of 0.11 % of all primary bone tumors. The most common site of involvement is mandible (reported incidence 22% of all Desmoplastic fibroma cases) followed by metaphysis of long bones. Involvement of forearm especially involving periosteum is seldom reported. Prompt diagnosis and adequate management is important for limb salvage and restoration of limb function. [1-3]

An 11-year-old boy presented with painful mild swelling of left forearm for a month, with no significant past medical history or any history of trauma. Clinically mild tenderness was elicited at the site of pain. Otherwise rest of physical and systemic examination was unremarkable. The lesion on radiograph appeared to be eccentric, mixed sclerotic lytic area in left distal radius, with marginal sclerosis and solid interrupted periosteal reaction (Fig. 1). Differentials of non-ossifying fibroma, chondromyxoid fibroma, osteosarcoma or sarcomas like Ewing's sarcoma or eosinophilic granuloma were considered. Contrast enhanced CT scan revealed a well-defined, oval, soft tissue attenuated, mass with no post-contrast enhancement, in between left radial and ulnar diaphysis resulting in increased intra-osseous distance (Fig. 2a, 2b). There was an interrupted solid periosteal reaction adjacent to the mass (Fig. 2a, 2b, 2c). Possibility of aggressive lesion was still not ruled out and furthermore MRI was performed which showed a well-defined inter-fascial/ inter-osseous lesion, which was low on T1, high on T2, and STIR with minimal contrast

enhancement on post contrast images and with adjacent bone involvement as was evident by focal cortical interruption, mild endosteal thickening and irregularity and also mild ulnar shaft remodeling (Fig. 3a, 3b). To further characterize the lesion, Tc99 MDP (methylene diphosphonate) bone scan was also performed which showed active bone involvement in left distal radial shaft.



Figure 1: Plain radiograph showing an eccentric area of sclerosis with interrupted solid periosteal reaction with Codman's triangle, in distal radial metaphysis.

Tissue biopsy under local anesthesia was done and examined by two different pathologists. The histopathological evaluation revealed abundant fascicles of fibroblasts with abundant collagen without any mitosis, atypia or necrosis (Fig. 4a). Tumor showed positive staining for B- catenin (Fig. 4b) and focal positive staining for smooth muscle actin (SMA) (Fig. 4c). Final diagnosis of

periosteal desmoplastic fibroma was made after multi-disciplinary team meeting.

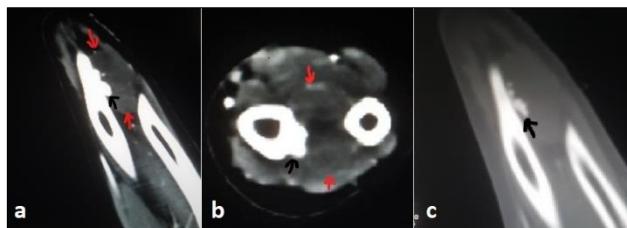


Figure 2: a (coronal) & b (Axial) fairly defined, non-enhancing, soft tissue mass between the radius and ulna (red arrows) along with interrupted solid periosteal reaction (black arrow); c, Interrupted solid periosteal reaction on bone window CT.



Figure 3: (a & b), Fairly defined high T2 lesion on MRI in between radial & ulnar shafts.

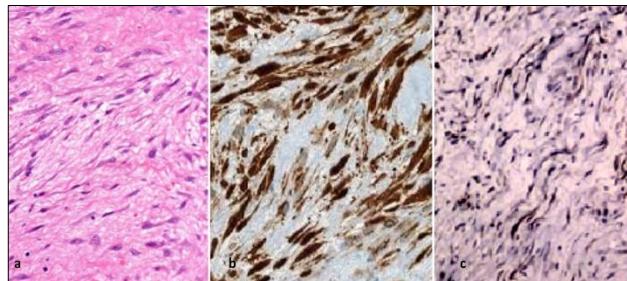


Figure 4: a) Hematoxylin staining showing fascicles of fibroblasts with abundant collagen. b) showing positive staining for B-catenin. c) showing positive SMA staining.

The patient was initially treated with excision of the tumor along with curettage of the adjacent radial shaft. Immediate postoperative radiograph revealed minimal post-surgical sclerosis and he was discharged on 3rd post op day. Histopathology revealed tissues composed of bland looking fibroblasts with abundant collagen showing positive staining for B-catenin and focal positive staining for smooth muscle actin (SMA). Tumor margins could not be assessed as specimen consisted of multiple white tissue pieces collectively measuring 1 x 1 cm. At 6 months follow up, the patient was unremarkable and showed no recurrence. However at 8 months after the first surgery, the patient developed pain and swelling of the forearm. Evaluations revealed recurrence with a size almost double that of the initial tumor. Patient un-

derwent wide en-bloc resection of the tumor with bone curettage. The second specimen on histopathology consisted of oriented soft tissue mass with multiple bony fragments, the margins of the soft tissue mass were clear of tumor. Histological evaluation of the bony fragments after short decalcification showed spindle cell neoplasm with similar histomorphological features as the previous surgical specimen. No allograft was required as the major part of shaft of the bone seemed spared. However patient received radiotherapy. Further 6 month and 1 year follow-ups were unremarkable.

The diagnosis of desmoplastic fibroma is very difficult on radiological imaging alone. The differentials include benign lesions such as fibrous dysplasia, non-ossifying fibroma, chondromyxofibroma and even aneurysmal bone cyst. If cortical destruction along with a soft tissue mass is seen (pressure effects from adjacent soft tissue mass), then differentials of slowly growing lesions like sarcomas (rhabdomyoma/rhabdomyosarcoma, lipoma/liposarcoma, fibromas, nerve sheath tumors, hemangioma and less likely Ewing's sarcoma are considered. [4-6]

It is locally aggressive tumor with propensity to recur locally. Rui et al reported its local recurrence rate as high as 17% with wide en-block resection and up to 55% with curettage. [7] Jamsheidi et al observed that recurrence occurred more frequently in patients with an associated soft tissue component and/or extra osseous extension [5] as also seen in our case. Histopathologically they have resemblance to aggressive fibromatosis (desmoid tumor) and were considered as bony counterpart of extra abdominal desmoid tumors. [5]

Wide resection of the lesion is considered as treatment of choice. Curettage and marginal resection of lesion with limb salvage and joints preservation will cause minimal postoperative morbidity and preservation of limb function but at the cost of increased risk of recurrence. Surgery combined with radiotherapy have promising results for local control of tumor. [6] Hence it is of vital importance that appropriate treatment is selected along with serial follow-ups to detect any recurrence as early as possible, particularly if tumor has a soft tissue component as seen in the index case.

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed

Authors' Contribution: All authors contributed equally in concept, design, literature review, drafting the manuscript, and approval of the final manuscript.

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