Desmoplastic Fibroma of Bone: a Case Series and Review of Literature

Yashwant Singh Tanwar^{1,2} · Yatinder Kharbanda² · Ruchi Rastogi³ · Ruchi Singh⁴

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Abstract

Desmoplastic fibroma is an uncommon locally aggressive benign tumor of the bone. Patients usually present with a long-standing history of pain and swelling. Radiologically and histologically, it can mimic a variety of tumors. This article presents a report on three cases of desmoplastic fibroma in the proximal humerus, distal femur, and neck femur region. The varied radiographic features and the management of the three cases are described in detail. All the three patients were treated by extended curettage. Follow-up ranged from 2 to 5 years. There were no local or systemic recurrence until the last follow-up. Tumor resection with adequate margins is the preferred treatment, although extended curettage can be an acceptable alternative treatment modality when resection is not possible or acceptable.

Keywords Desmoplastic fibroma · Extended curettage · Fibular graft

Introduction

Desmoplastic fibroma is a rare primary bone tumor which morphologically resembles its soft tissue counterpart the desmoid-type fibromatosis commonly seen in the abdominal wall. It was first described by Jaffe in 1958 and there are around 270 reported cases in literature accounting for less than 0.1% of primary bone tumors [1]. It has been reported in a diverse age group ranging from 20 months to 75 years; however, most of the lesions have been reported in the 3rd decade [2–4]. Long bones are commonly involved especially the distal femur and proximal tibia; however, the lesion also shows a predilection for the mandible and pelvis [5].

It is important to diagnose the lesion correctly, as radiologically and histologically, the lesion may be mistaken for an indolent, benign fibrous lesion or more aggressive spindle-cell sarcomas and the treatment can vary drastically [6]. As it is a

relatively rare tumor, the literature on desmoplastic fibroma of the bone is confined to few scattered case reports in the orthopedic, pathology, and radiology literature. We present our experience of three cases all of whom had varied presentations and were subsequently diagnosed as desmoplastic fibroma on biopsy.

Material and Method

An informed consent was taken from all the patients to publish before publishing the report.

Patient 1

A 14-year-old male boy presented in the Outpatient Department with complaint of gradual onset slowly progressive pain in the left arm since last year. Roentgenograms of the upper limb showed presence of a well-marginated osteolytic lesion in humeral diaphysis causing osseous expansion, cortical thinning, and endosteal scalloping (Fig. 1a). No periosteal reaction was seen and the physis was spared. Internal pseudo-trabeculations which are quite characteristic of desmoplastic fibroma were also seen. MRI in coronal T1- and fat-suppressed T2-weighted images of the upper humerus revealed that the mass was isointense to hypointense compared to adjacent muscles on T1W sequence and heterogeneously hyperintense on fat-suppressed T2W sequence with few interspersed T2 hypointense areas

✉ Yashwant Singh Tanwar
tanwar_yashwant@yahoo.co.in

¹ HNo299, Pocket B, DDA Flats, Sarita Vihar, New Delhi, India
² Department of Orthopedics, Apollo Hospital, New Delhi 110076, India
³ Department of Radiology, Apollo Hospital, Sarita Vihar, Delhi 110076, India
⁴ Department of Radiation Oncology, Lok Nayak Hospital, Delhi, India

Fig. 1 **a** Well-margined osteolytic lesion in humeral diaphysis causing osseous expansion, cortical thinning, and internal pseudo-trabeculations. **b, c** Showing a large exophytic growth arising from the posterior aspect of the distal femur

(Fig. 2a). FNAC was suggestive of fibrous tumor possibly desmoplastic fibroma.

After all preliminary investigations, patient was planned for surgery. Keeping in the view the large size of the tumor, it was

Fig. 2 **a** MRI image of the proximal humerus lesion. **b** MRI image of the desmoplastic fibroma in the neck femur. **c** Showing the coronal and sagittal MRI images of the distal femur demoplastic fibroma. **d** Post-

contrast axial T1-weighted image showing moderate heterogeneous post-contrast enhancement

decided to proceed with extended curettage and bone grafting rather than en bloc excision and reconstruction. Humerus was approached antero-laterally via the delto-pectoral interval. Anterior cortex was expanded and thinned out. Drill holes were made and a large rectangular window measuring 7×3 cm was removed. A firm soft tissue mass was found inside the medullary cavity. Care was taken to remove the mass in toto (Fig. 3a). The cavity was thoroughly curetted and lavaged. Hydrogen peroxide and cryo-therapy were applied to the walls of the cavity to prevent recurrence. Autogenous ipsilateral fibular shaft was harvested with a length of approx. 10 cm. It was divided in length into two pieces and multiple drill holes were made in the cortex to help in early integration of the graft. Autogenous ipsilateral iliac crest cortico-cancellous graft was also harvested. The recipient cavity was packed with graft from fibula and iliac crest (Fig. 4). Hemostasis was achieved and a meticulous closure was done, with no drain. Soft dressing was applied and arm was placed in U slab for 4 weeks. Gentle active range of motion exercises were started at the elbow and shoulder joint. Follow-up radiographs were done at 4, 8, 16 weeks and at 6 months and 1 year. Graft gradually incorporated in the host bone (Fig. 5a). There was no recurrence until the last follow-up at 5 years.

Patient 2

A 65-year-old female presented with complain of pain in the right hip region since 6 months. On examination, tenderness was present in the groin region anteriorly on deep pressure. Leg lengths were equal and there was no distal neuro-vascular deficit. X-rays of the hip showed a large radiolucent lesion confined to the neck of femur. MRI showed classical features suggestive of a fibrous tumor. Coronal T1- and T2-weighted images at the level of hip joint demonstrated a well-marginated lesion in the femoral neck causing mild osseous expansion and cortical thinning. The lesion was showing iso-intense signal to adjacent muscles on T1W and isointense to hypointense signal on T2W images (Fig. 2b). There was no extra-osseous soft tissue component or periosteal reaction. FNAC was suggestive of fibrous tumor possibly desmoplastic fibroma.

Patient was planned for a hip conservation surgery in the form of excision of the mass and bone grafting. Hip was exposed via the anterior Smith-Petersons approach. The interval between TFL and gluteus was identified and the neck femur was exposed. The anterior cortex of neck was found to be intact but thinned out and slightly bulged. Drill holes were

Fig. 3 Showing the excised specimen from the proximal humerus (a), the neck femur (b), and the distal femur (c)

Fig. 4 Showing a large residual defect in proximal humerus after excision of tumor and extended curettage which was filled with ipsilateral fibular graft and iliac crest graft

made with a 3.5-mm drill bit and a rectangular cortical window measuring 3 cm on each side was taken out. There was a solid fibrous tumor filling the medullary cavity (Fig. 5a). The mass was excised (Fig. 3b) and the walls were curetted with burr and liquid nitrogen. Ipsilateral non-vascularized fibular shaft graft was harvested and it was split longitudinally into three pieces (Fig. 5d). Cancellous graft was also harvested from the ipsilateral iliac crest. Four guidewires were passed from the trochanter into the femur head across the defect in the neck and a cannulated 4.0-mm drill bit was drilled over them,

through a separate stab incision over the greater trochanter (Fig. 5b). The fibular grafts were passed over the guidewires and the two partially threaded 7.0-mm cancellous screws were passed over the remaining two guidewires. Cancellous bone harvested from the iliac crest was packed into the remaining defect (Fig. 5c). In bed, exercises were started and the patient was kept non-weight bearing for 6 weeks. Partial weight bearing was continued until 12 weeks after which full weight was allowed. The follow-up at 4 years showed incorporation of the graft and no evidence of recurrence (Fig. 6b).

Fig. 5 **a** Showing anterior view of the neck femur, the anterior cortical window has been removed and the fibrous tumor can be seen filling the neck. **b** Anterior view of the neck femur showing the cavity and two guidewires passed from the base of GT into the head femur. **c** Showing

filling of the defect in the neck femur by fibular grafts and cancellous bone chips. **d** Showing autogenous fibular graft split longitudinally into three sections

Fig. 6 Post-op X-ray of the proximal humerus (a) and the neck femur (b) showing incorporation of the graft. c Showing the post-operative image after resection of the distal femur tumor and cementing

Patient 3

A 31-year-old female presented with a long-standing history (2 years) of mild pain and swelling over the posterior aspect of the left femur distally. On inspection, superficial dilated veins could be seen, the overlying skin was normal and not adhered to the underlying swelling. On palpation, the overlying temperature was normal and the swelling was firm to hard in consistency. X-rays showed a large well-delineated osteolytic lesion with bony expansion and cortical breach arising at the posterior aspect of the distal femoral metadiaphysis (Fig. 1b, c). Coronal and sagittal T1- and T2-weighted images of the lower femur demonstrates a large mass in the posterior and medial compartment of the thigh consisting of both intra-osseous and extra-osseous components with osseous expansion, cortical destruction, and bone lysis (Fig. 2c). The intra-osseous component of mass showed well-demarcated margins with absence of any surrounding marrow edema. The mass was heterogeneous with isointense to hyperintense signal compared to adjacent muscles on T2-weighted sequence.

Post-contrast sagittal and axial T1-weighted images revealed moderate heterogenous post-contrast enhancement in both intra-osseous and extra-osseous components of mass with presence of few thin linear non-enhancing fibrous septa (Fig. 2d). A low-grade parosteal osteosarcoma was suspected. FNAC of the lesion showed fibrous tumor suggestive of desmoplastic fibroma. Excision of the lesion was planned, along with extended curettage and cementing of cavity. The tumor was exposed using the extensile medial subvastus approach. Surrounding soft tissue was dissected of the mass and the tumor was taken out piece meal (Fig. 3c). The intra-medullary component was taken out and the cavity was curetted with a power burr (Fig. 5e). The cavity was filled with antibiotic impregnated bone cement and the wound was closed in layers over a drain (Fig. 6c). Post-operatively, the limb was protected in an above knee slab for 6 weeks. Recovery was uneventful and partial weight bearing was started at 6 weeks and full weight bearing at 10 weeks.

Discussion

Desmoplastic fibroma is an extremely rare, slowly progressing tumor with well-differentiated cells that produce collagen. Although benign in nature, it is locally aggressive and the peak incidence is between 15 and 25 years. Patients have pain, often for several years, which suggests slow growth. On rare occasions, a desmoplastic fibroma may be an incidental finding. Any bone may be affected by desmoplastic fibroma; however, most lesions occur in the long bones. Although considered to be the counter part of desmoid-type fibromatosis [1], recent study revealed that there were no mutations in exon 3 of CTNNB1, encoding for B-catenin, thus genetically distinguishing desmoplastic fibromas of bone from desmoid-type fibromatosis [7].

In 2013, the WHO described the microscopic appearance of desmoplastic fibroma as being composed of slender, spindle to stellate cells with minimal cytological atypia and abundant collagenous matrix [8] (Fig. 7). The most important histological differential diagnosis is low-grade fibrosarcoma. However, it can be distinguished on the basis of high cell

Fig. 7 Showing the microscopic appearance of desmoplastic fibroma

density, high grade of polymorphism, and a high rate of mitosis.

Radiographically, a wide variety of entities can masquerade as a desmoplastic fibroma including fibrous dysplasia, giant cell tumor, unicameral bone cyst, chondromyxoid fibroma, nonossifying fibroma, periosteal desmoids, eosinophilic granuloma, low-grade intra-osseous osteosarcoma, and adamantinoma, as well as distant metastasis. The most common locations of desmoplastic fibroma are metaphyses and diaphyses in the long bone as seen in two of our cases, though it may also rarely arise in the diaphysis, as seen in our first case. It usually presents as a well-margined osteolytic lesion on radiographs with a narrow transition zone. It may have irregular trabeculated or bubbly appearance with cortical thinning. An extra-osseous soft tissue mass may be present in cases with cortical breach. Periosteal new bone formation and matrix mineralization are rare. Extra-osseous soft tissue component was present in our third case associated with cortical destruction, while it was absent in our remaining two cases.

Although there is a considerable overlap in the MR imaging features with other bone tumors, an interesting MR feature of desmoplastic fibroma is the presence of low to intermediate signal intensity foci on T2-weighted images, which radiographically does not correspond to calcifications. This feature may help narrow the differential diagnosis [9, 10]. Other authors have described varied T2-weighted signal intensities among different cases, including high-SI areas within small low-SI background and low-SI areas with small high-SI background [11, 12]. Low T2 SI areas correspond to abundant collagen fibers while high T2 SI areas correspond to fibroblasts and loose collagen fibers [11, 12]. We found T2 hyperintense signal in all three of our cases. CT has advantage over MRI to show the bony destruction to a better extent while MRI provides better depiction of medullary and soft tissue extent of tumor.

Recurrence rates following simple curettage have been reported to very high [4]. Treatment by surgical excision with wide margins is preferred and has shown to have the least recurrence rates. However, in large tumors, wide resection and reconstruction seems unreasonable, as it poses a considerable reconstructive challenge. Also, the options available for reconstruction are either extremely costly (e.g., allograft reconstruction and mega prostheses) or impair function permanently (e.g., arthrodesis and rotation plasty). These are at many times not acceptable to the patient.

Many similar locally aggressive tumors like GCT and chondroblastoma are treated by extended curettage. The technique of curettage has improved and extended curettage has been widely described as a unique modality of surgical treatment for bone tumors. It is advised to create a cortical window that is at least as large as the lesion to prevent leaving residual tumor cells “around the corner” adjacent to the near-side cortex. After curettage is complete, a power burr is used to

enlarge the cavity 1 to 2 cm in all directions taking care to avoid perforation of the subchondral bone into the joint. The curettage is further extended with the use of adjuvants, such as liquid nitrogen, phenol, bone cement, electrocautery, or an argon beam coagulator which help to kill any remaining tumor cells [13].

Nishida et al. reported five patients with desmoplastic fibroma treated with aggressive curettage. No recurrences were seen during follow-up periods ranging from 5 years and 6 months to 9 years [14]. Nedopil et al. reported no recurrence in a case of desmoplastic fibroma of the ilium treated by curettage and bone grafting [15]. Evans et al. retrospectively analyzed 13 patients of whom six underwent intra-lesional curettage, three underwent marginal excision, one patient underwent wide excision and endoprosthetic replacement, and one patient underwent below-knee amputation. Of these, two patients had recurrence and both had undergone intra-lesional curettage. Both the recurrences developed within the first year [16]. Rastogi et al. reported three cases of unusual locations and concluded that intra-lesional curettage can be attempted if resection leads to major functional loss [17]. Yokouchi et al. reported the no recurrence at 12-year follow-up after extended curettage and heat ablation for desmoplastic fibroma of the distal femur [18].

We, in the present series, treated three cases by extended curettage. In two patients, the defect was filled with autogenous iliac crest and fibular graft, and in one patient bone cement was used. There was no recurrence until the last follow-up. Recurrence after desmoplastic fibroma has been reported to occur usually within a year. Although uncommon, late recurrences after many years have been reported which suggest the need of keeping these patients under follow-up especially when a conservative surgical procedure has been done. The follow-up time period of the patients in the present series seems to be small in the context of drawing any concrete conclusion regarding recurrence. Even if recurrence was to occur, considering the morbidity of wide excision of such large tumors and given the relative short life span of megaprosthesis, it is valuable to gain even a few years. Bohm et al. in their case report and review of literature observed that the recurrence rate for simple curettage was around 55%; whereas, the recurrence rate decreased to just 25% when an adjuvant was added [4].

As desmoplastic fibroma histologically resembles soft tissue fibromatosis, it has been speculated that radiotherapy may be effective in achieving local control. Effectiveness of radiotherapy in soft tissue fibromatosis is well documented. There have been two case reports describing treatment of desmoplastic fibroma with radiotherapy as the primary modality, one in the ilium and one in the distal femur [19, 20]. Adjuvant RT after a conservative surgical procedure seems an attractive option especially in large tumors with soft tissue components. Yin H et al. reported 12 cases of desmoplastic

fibroma of the spine treated, out of which four were treated by subtotal resection followed by radiotherapy and the rest eight underwent total tumor resection. Post-operative radiotherapy was administered in six cases. Local recurrence was detected in one of the cases that underwent subtotal resection and was not detected in any of the cases involving total spondylectomy. They concluded that radiotherapy may be an acceptable alternative therapy especially when en bloc resection has the potential to result in significant functional impairment [21].

Conclusion

Desmoplastic fibroma is a rare locally aggressive tumor, which is usually diagnosed on biopsy as radiological features can be mimicked by various other tumors. Surgical excision with wide margins is the treatment of choice and is appropriate for all small tumors and for tumors at expendable locations like the fibula, ulna, and ribs. However, extended curettage may be more appropriate for large tumors at meta-diaphyseal locations where en bloc excision would result in a challenging reconstruction. It is difficult to comment on the role of radiotherapy as a primary modality, as the literature is confined to a couple of case reports. However, radiotherapy may be important as an adjuvant after conservative surgery such as extended curettage. Late recurrences have been reported in literature and patients should be kept under regular follow-up to detect the recurrence as early as possible.

Compliance with Ethical Standards

All procedures performed were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Conflict of Interest The authors declare that they have no conflict of interest.

Informed Consent Informed consent was obtained from all individual participants included in the study.

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