Desmoplastic Fibroma of the Distal Femur in a Young Man: A Rare Case Report

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Abstract

Introduction: Lytic lesions in the distal femur in a mature skeleton though a common presentation for various tumors, desmoplastic fibroma (DFB) of bone is a rare occurrence. Review of the literature shows its incidence of 0.06% to 0.3%. In the majority of reported cases, the diagnosis has been obtained on histopathological examination. Treatment varies from aggressive curettage to amputations. We describe a novel surgical technique for dealing such lesions.

Case Report: A 24-year-old male presented with swelling around with DFB the left distal femur presented with pain for 4 months which progressed in severity and led to inability to bear weight. On clinical examination, he had a tender discrete swelling over the medial aspect of the left distal femur. Radiographic examination showed an eccentric lytic lesion in the metaphyseoepiphyseal region of the left distal femur. An extended curettage using phenol (10%) as adjuvant therapy was performed. The cavity was packed with bone cement and the distal femur was fixed by spanning the lytic lesion with a distal femoral locking plate. At months follow-up, he reported complete resolution of symptoms, and on examination, he had pain free and full range of motion, without any signs of recurrence.

Conclusion: An effort should be made to obtain a pre-operative histopathological diagnosis of the tumor type. In cases of equivocal findings, a diagnosis of DFB of the bone should be considered. Extended curettage, phenol ablation, and bone cement with plate augmentation offer an effective treatment modality in the treatment of DFB.

Keywords: Desmoplastic fibroma, benign bone tumors, surgery, knee, femur.

Introduction

Of all the primary bone tumors, desmoplastic fibroma (DFB) is a relatively rare entity [1, 2]. The reported incidence in literature ranges from 0.06% to 0.3%. DFB is a benign but locally aggressive tumor with border line activity with no site, sex, or age predilection. Like its soft-tissue counterpart, DFB is notorious for its recurrence. One study [2] refers the recurrence rate being as high as 42%. Cortical destruction and soft-tissue spread portend a more aggressive presentation [3]. Treatment varies from aggressive curettage to wide excision during index surgery to amputations or radical excision and distal femoral replacement with prosthesis [4]. Literature review shows that a pre-operative diagnosis of DFB is as a rule never made and therefore appropriateness of surgical excision during the index procedure can always be debated [5]. We report a case of DFB of the distal femur treated by a novel technique using extended curettage, adjuvant phenol application, and bone cement with plate fixation.

Case Report

Clinical presentation

A 24-year-old young male presented with a history of the left knee and lower thigh pain and swelling for the past 4 months. The pain started acutely, without any history of trauma, and progressed over the past 4 months. For the past month, he was unable to weight bear on the left lower limb, and there were no constitutional symptoms. There was a moderately large and tender swelling present over the lower medial left thigh with painful knee movements.

Imaging

On plain radiographs, a large destructive osteolytic lesion involving the distal femur with breakage of the medial cortex was seen (Fig. 1). Magnetic resonance imaging revealed a large heterogeneous altered signal in the lower diaphysis and metaphysis of the distal femur, with an asymmetrical expansion of the involved bone.

Laboratories and tissue diagnosis

All laboratory investigations were within normal limits. The core biopsy was non-conclusive. Curetted material from the lesion obtained intraoperatively revealed a mesenchymal neoplasm of the bone, of mild-to-moderate cellularity. It was composed of

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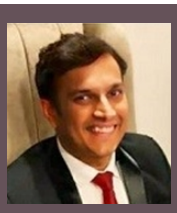
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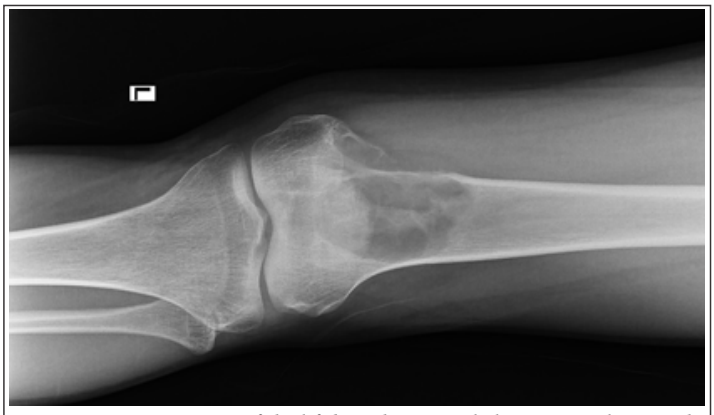

Figure 1: Pre-operative X-ray of the left knee showing single lytic eccentric lesion in the diaphysiometaphyseal region of the distal femur showing scalloping margins and lamellation, with destruction of the medial cortex of the femur.

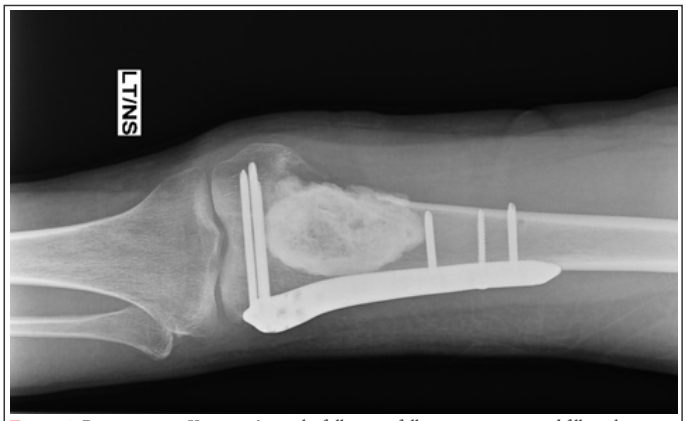

Figure 4: Post-operative X-ray, at 6 months follow-up, following curettage and filling the cavity with bone cement, the distal femur has been prophylactically stabilized with a distal femoral locking plate on the lateral aspect.

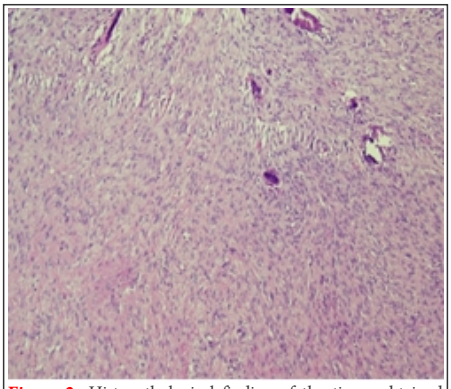

Figure 2: Histopathological finding of the tissue obtained intraoperatively showed a mesenchymal neoplasm of the bone of mild-to-moderate cellularity. It composed of short spindle cells, which were distributed haphazardly, alternating with thick sclerotic collage bundles. The background stroma was of dense fibrous tissue.

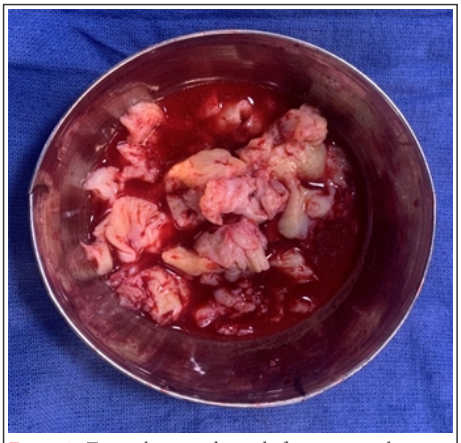

Figure 3: Tissue that was obtained after curetting the cavity shows white and firm fibrous tissue.

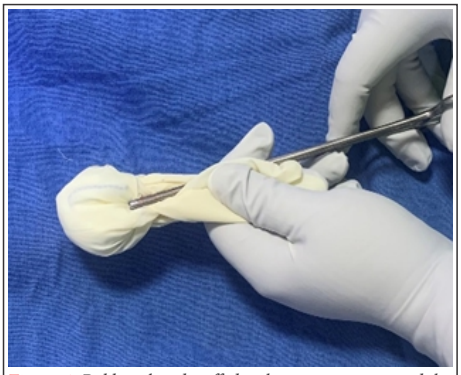

Figure 5: Rubber gloved stuffed with cotton gauze is used the pressurize the cement within the cavity.

short spindle cells, which were distributed haphazardly, alternating with thick sclerotic collagen bundles. The background stroma was of dense fibrous tissue (Fig. 2), with no evidence of malignancy. The histology features were suggestive of a DFB.

Surgical treatment

The lesion was exposed through a lateral incision. There was a large tumor involving the distal femur, with breach of the medial cortex. It was fibrous in consistency which was filling the whole cavity of the femur (Fig. 3). An extended curettage was done and the cavity was treated with phenol (10%) as adjuvant therapy and was filled with bone cement. The distal femur was fixed by spanning the lytic lesion with a distal femoral locking plate (Fig. 4) to protect the distal femur. At 6 months follow-up, he reported complete resolution of symptoms and had regained 90° of knee flexion and a full extension of the knee. The plain radiographs did not show any recurrence of the tumor.

Discussion

Jaffe (1958) was the first to describe DFB, as a distinct clinical entity and a type of osseous fibrous tumor. It has a close resemblance to a desmoids tumor or fibromatoses of the soft tissues [1, 2]. It is an extremely rare benign tumor, which is locally aggressive with high recurrence rates. Its reported incidence is only around 0.06% of all the primary and 0.3% of all the benign bone tumors [1, 2]. DFB commonly affects young individuals in the first three decades of life [3] and the flat bones of the face, and the involvement of long bones of the extremities is uncommon. Among all the long bones, the involvement of the femur is the most common [1, 2, 3, 4]. Awareness about this entity is mandatory as only a few case reports have been published on it so far [5]. DFB can mimic other osteolytic lesions, such as fibrous dysplasia, giant cell tumors (GCTs), brown tumor, aneurysmal bone cyst and malignant bone tumors (fibrosarcoma and osteosarcoma) [6, 7, 8, 9, 10, 11]. Surgical removal of the tumor is the recommended treatment [12]. There has been a debate about the choice of preferred surgical treatment between extended curettage and wide excision of the tumor [13]. The reported recurrence rates are higher after intracapsular or marginal curettage as compared to wide excision with bone grafting. Furthermore, the wide excision in a young patient may be associated with significant dysfunction and disability [13, 14, 15] with possible early failure of endoprosthesis when implanted. Another extremely rare complication is malignant transformation into osteosarcoma [16, 17].

We describe an innovative technique of using phenol as adjuvant along with polymethyl methacrylate (PMMA) to fill the cavity supplemented with plate osteosynthesis in the treatment of DFB of distal femur which has not been reported to the best of our knowledge, although the similar use of phenol, PMMA, and support plate has been used in other tumors such as locally aggressive and recurrent GCT. The bone PMMA cement is packed in the bony cavity after pressurizing it with a surgical glove (filled with a cotton gauze), similar to the technique of pressurizing the bone cement for the acetabular cup as performed in total hip arthroplasty (Fig. 5). This allows bone

cement to disperse uniformly with in the contours of the walls of the cavity, leaving no residual dead space. Further, we believe that the heat generated during the exothermic reaction of the PMMA, effectively acts to destroy the tumor cells, and the tight packing of the cavity does not leave any residual space for the tumor cells to grow.

The early outcomes were satisfactory, however, a long-term follow-up is warranted to check for the recurrence of DFB.

Conclusion

An effort to obtain a pre-operative histopathological diagnosis is gold standard. However, rare entities like DFB of bone should be kept in mind. These are locally aggressive tumors with high recurrence rates. Hence, extended curettage, phenol ablation, and bone cement with plate augmentation offer an effective treatment modality in the treatment of DFB.

Clinical Message

Pre-operative diagnosis of DFB in lytic lesion of the distal femur should always be entertained.

Extended curettage, phenol as adjuvant with adequate bone cementing, and plate osteosynthesis offer an alternative treatment modality for the management of DFB of the distal femur.

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