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Desmoplastic Fibroma: A Rare Pathological Midshaft Femoral Fracture Treated With Resection, Acute Shortening, and Re-lengthening

A Case Report

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Investigation performed at Royal Infirmary Edinburgh, Edinburgh, Scotland

Abstract

Case: We report a rare case of desmoplastic fibroma (DF) of the midshaft femur presenting as a pathological fracture. This rare benign bone tumor was treated with an acute en bloc excision and femoral shortening over an intramedullary nail. Once union of the acute shortening had been achieved, further surgery was undertaken to lengthen the femur with the use of Intramedullary Skeletal Kinetic Distractors. At 3 years after fracture, our patient had achieved equal leg lengths, had normal knee function, and was disease free.

Conclusions: DF resulting in pathological fracture of the midshaft femur is extremely rare and has not been reported to occur in the femoral diaphysis. This location is important as preservation of the joint above and below is preferable and en bloc excision is recommended. Restoration of bone stock after en bloc excision is difficult and recurrence needs to be monitored.

Desmoplastic fibroma (DF) is an exceedingly rare and locally aggressive benign bone tumor. The reported incidence of this tumor is 2.5 cases for every population of 100 million[1]. As initially described by Jaffe[2], the lesion resembles an aggressive fibromatosis and is histologically similar to the soft tissue desmoid tumor. The World Health Organization described the appearance of DF as "tissue from bone being composed of slender, spindle to stellate cells with minimal cytological atypia and abundant collagenous matrix"[3]. Radiographically, DF typically displays a well-demarcated lytic pattern or extensive bone destruction with a "moth-eaten" appearance, and it characteristically lacks matrix calcification or sclerotic margins while often showing internal pseudotrabeculation[4].

Because the recurrence rate ranges from 15.4% to 72%[1,5], resection of the affected bone is the preferred treatment, and ideally as en bloc excision. Recent studies have suggested extraosseous tumor growth to be a poor prognostic sign[1]. Wide resection is the treatment of choice, especially where limb salvage surgery can provide restoration of function[1]. To our knowledge, a midshaft femoral DF lesion presenting as a pathological fracture has not previously been reported. The aim of this study is to document and demonstrate this rare site and pathological fracture presentation. We also describe the mid-term outcome of en bloc resection, acute shortening, and re-lengthening over an intramedullary nail (IMN).

The patient was informed that data concerning the case would be submitted for publication, and he provided consent.

Case Report

A previously healthy 24-year-old male presented to our emergency department following a low-energy fall on ice from a standing height. He was unable to bear weight through his right leg and deformity in the thigh was observed. He was treated at the scene by emergency services who placed him into traction before transfer to the hospital.

On admission to the hospital, the patient gave a history of knee and thigh pain, which had been present for approximately 1 year. The pain was usually dull, transient in nature, and exacerbated by weight-bearing activities. He reported night pain and at 1 stage sought advice from physiotherapists who thought muscle strain was the likely etiology. He could not run for longer than 5 minutes because of thigh pain and on occasion had a limited walking distance because of thigh and knee pain. He did not take analgesia for the pain. He had no family history of musculoskeletal tumors. On examination in the emergency department, he was noted to have deformity of the thigh adequately managed in a splint, he had no abnormalities to neurology or vascularity, and the injury was closed. Radiographs demonstrated a pathological midshaft femoral fracture (Fig. 1).

Disclosure: The **Disclosure of Potential Conflicts of Interest** forms are provided with the online version of the article (http://links.lww.com/JBJSCC/A788).

Fig. 1

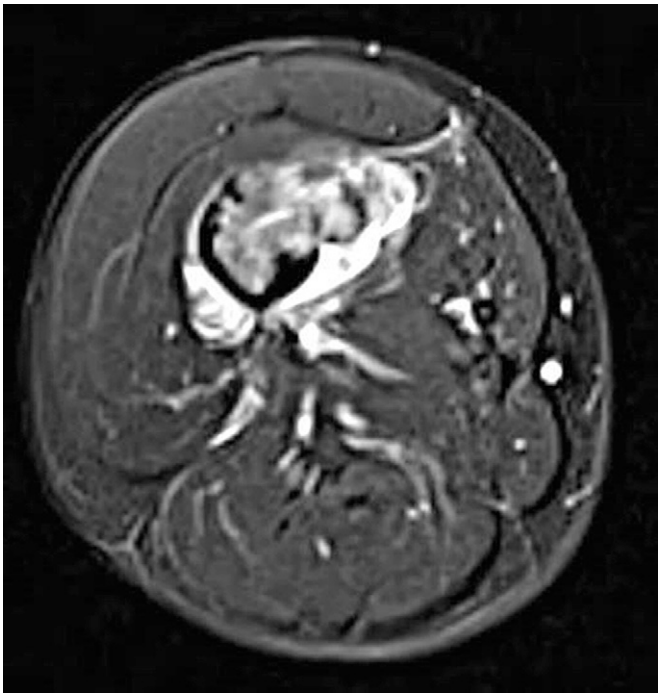

Fig. 2

Fig. 1 Plain film radiograph at presentation. **Fig. 2** Magnetic resonance imaging axial slice though pathological fracture.

Magnetic resonance imaging demonstrated the fracture through a lytic lesion within the femoral diaphysis with extensive soft tissue hemorrhage, particularly within the deep and mid-substance of vastus intermedius. There was marked endosteal thinning of the cortex and a large defect in the anterior cortex with significant soft tissue involvement (Fig. 2), and also intramedullary abnormal signal extended over a distance of 9 cm. The differential diagnoses included nonossifying fibroma, metastasis, myeloma, and eosinophilic granuloma.

The fracture was stabilized with inline skin traction, and an urgent open biopsy through an anteromedial approach in theatre was organized. Computed tomography–guided biopsy was considered; however, the risk of not achieving a representative sample due to fracture hematoma leads to the decision to perform open biopsy. The anteromedial approach was chosen so that only 1 muscle (vastus medialis) would be contaminated to access the site. The representative specimens demonstrated a lesion consisting of bland spindle cells within a collagenous stroma (Fig. 3). There was little cellular or nuclear pleomorphism and mitotic activity was not readily identified. There was no well-defined histiocytic component, and no evidence of metaplastic bone or cartilage was seen within the tumor. At the edge of one area of lesional tissue, there was some bone formation, but this seemed to be reactive in nature and probably related to the pathological fracture. The major histological differential diagnosis in this case included DF or fibrohistiocytic lesion in which there was no histiocytic component. At the multidisciplinary sarcoma tumor meeting with a 2 pathologist consensus, the diagnosis of DF was made.

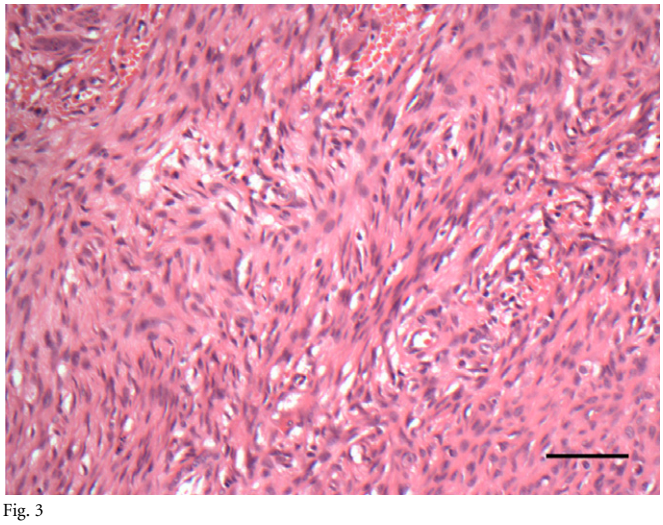

Fig. 3

The tumor comprises bland spindle to stellate cells within a collagenous matrix. The cells are arranged in whorls and fascicles. Hematoxylin and Eosin stained section. Bar = 100 mM.

Fig. 4

Fig. 5

Fig. 6

Fig. 4 Acute femoral shortening after en bloc excision with intramedullary nail. **Fig. 5** An 80 mm lengthening over Intramedullary Skeletal Kinetic Distractor. **Fig. 6** New callus is seen after lengthening.

Shortly after the benign nature of this pathological fracture was confirmed, en bloc excision in theatre was planned. Under a general anesthetic, an anteromedial midline incision incorporating the biopsy tract was performed. The femur was osteotomized above and below the tumor. Careful dissection was performed, and the tumor and pathological fracture were excised. The femur had been shortened a total of 10 cm and was stabilized with the IMN technique (Fig. 4).

The macroscopic specimen was a 10 cm length of bone with attached skeletal muscle. Both cut surfaces of the bone were unremarkable. The longitudinal section showed the displaced fracture with pale grey tumorous material within the medullary cavity. There was extensive hemorrhage relating to the fracture. The lesional material appeared excised by 2.3 cm and 1.2 cm on the bony margins.

Microscopic sections showed that the lesional tissue was composed of irregular fascicles and haphazardly arranged, minimally pleomorphic, spindle cells lying within a variable collagenous stroma. There was an occasional focus of metaplastic bone formation, but this was minimal. Indeed, it was unclear whether or not this represented residual preexisting bone or bone forming within the matrix. However, this was only a minimal part of the overall lesion which, again, was best characterized as DF.

Union of the acute shortening was achieved, and at 5 months after en bloc excision, exchange nail was undertaken. The original static nail was removed, and a femoral osteotomy proximal to the union with insertion of an Intramedullary Skeletal Kinetic Distractor (ISKD) construct with the assistance of 2 distal blocking screws was performed. Over a 25-day period, 80 mm of lengthening was achieved over the ISKD nail (Fig. 5).

Six months after insertion of the ISKD, range of movement in the knee was 0 degrees of extension to 120° of flexion. There was 5° of quadriceps lag (Fig. 6). A further 2.5 cm lengthening was required for leg length equality, and the ISKD was exchanged at 8 months (Fig. 7).

The second lengthening of 2.5 cm was achieved and consolidation occurred in a predictable fashion. Two years after the index operation, our patient demonstrated full extension to 130° flexion with Medical Research Council muscle power grade 5. A little less than 3 years after diagnosis, the second ISKD nail was removed (Figs. 8 and 9), and he was able to mobilize without crutches within 6 weeks. Leg lengths were equal. Subsequent clinical and radiographic follow-ups have shown that he has been disease free for the last 7 years.

Discussion

Pathological fracture in a young patient is always a cause for concern. The clinical manifestations of DFs include pain and swelling as the disease progresses. In our experience, diagnosis of DF by radiography alone is not possible. Radiographs

Fig. 7

Fig. 8

Fig. 9

Fig. 7 Second lengthening by 2.5 cm. **Fig. 8** Removal of second Intramedullary Skeletal Kinetic Distractor nail Anteroposterior femur. **Fig. 9** Removal of second Intramedullary Skeletal Kinetic Distractor nail lateral femur.

in this patient demonstrated a classic expansile, osteolytic lesion with a "moth eaten" appearance. Since this lesion has not been reported in the midshaft of the femur previously, histology to establish diagnosis was mandatory.

Crim et al.[4] reported cortical breakthrough in only in 29% of patients with DF. As has been reported above, cortical erosion and soft tissue extension were exhibited in this case. The open biopsy therefore had to be undertaken through the pathological fracture site. The histological diagnosis was based on minimally pleomorphic spindle cells lying within a variable collagenous stroma.

To date, 9 cases of DF arising from the femur have been reported in the English literature. Two were in the proximal femur[6,7] and 7 in the distal femur[8-14]. To our knowledge, this is the first case of midshaft femoral DF lesion. The surgical management of the 10 cases of DF in the femur are summarized in Table I.

TABLE I Published Surgical Management of Desmoplastic Fibroma in the Femur

Author	Year	No. of Cases	Technique	Site
Lichtman and Klein[6]	1985	1	Curette and bone graft	Proximal femur
Gong et al.[7]	2015	1	Curette and bone graft	Proximal femur
Wadhwa et al.[8]	2013	1	Curettage and bone grafting and cementation	Distal femur
Clayer and Oakeshott[9]	1994	1	Distal intralesional curettage with proximal en bloc resection followed by allograph replacement	Distal femur
Rastogi et al.[10]	2008	1	Extended curettage and autologous cancellous bone graft and fibular bone grafting	Distal femur
Chan et al.[11]	1987	1	Nonoperative	Distal femur
Bertoni et al.[12]	1984	1	Excision and endoprosthesis	Distal femur
Gao et al.[13]	2013	1	Radical resection and internal fixation	Distal femur
Yokouchi et al.[14]	2014	1	Extended curettage, heat ablation, and artificial bone grafting	Distal femur

Due to the recurrence rate of up to 72%[5], resection of the affected bone is the preferred treatment for DF. A midshaft femoral lesion presents the ideal location for en bloc resection with preservation of the joints above and below. In our case, 10 cm of bone was resected. The options for bone reconstruction included autograft, vascularized fibular graft, allograft, intercalary endoprosthetic spacer, Masquelet technique, and distraction osteogenesis through bone transportation. Biological reconstruction is our preferred long-term surgical reconstruction as it can restore length and function by means of native bone with the ability to heal fractures and respond to stress. The use of an IMN device in the femur for treatment of a pathological fractures requires careful consideration. There is the potential for local metastasis and a large area of contamination in the setting of positive margins and aggressive tumor. Resection of the tumor should always occur before any preparation for nail insertion to avoid contaminating distant sites with reamers and fixation devices.

Conclusion

Reconstruction with distraction osteogenesis involves an initial decreased function and longer recovery when compared to other reconstructive techniques, but this shortcoming is potentially outweighed by the longevity, function, and durability of this reconstruction method[15,16]. In this case, en bloc resection, acute shortening, and re-lengthening over a nail for the treatment of DF in the femoral diaphysis was a viable option. ■

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