Case report

Distal Femoral Replacement as a Salvage Treatment After Desmoplastic Fibroma Resection

Norberto J. Torres-Lugo, MD a, *, David Beatón-Comulada, MD a, Roberto Colón-Miranda, MD a, José P. Bibiloni-Lugo, BS b, Norman Ramírez, MD c, Juan Bibiloni-Rodríguez, MD a

a Department of Orthopaedic Surgery, University of Puerto Rico, Medical Sciences Campus, San Juan, Puerto Rico
b School of Medicine, Universidad Central del Caribe, Bayamón, Puerto Rico
c Department of Orthopaedic Surgery, Mayagüez Medical Center, Mayagüez, Puerto Rico

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A B S T R A C T

Desmoplastic fibroma is an extremely rare primary bone tumor that can mimic the presentation of other bone lesions. We describe the case of a middle-aged male with a mass on the left distal femur initially diagnosed as fibrous dysplasia that underwent a wide margin excision followed by a distal femoral replacement to restore anatomy and functionality. Histologic examination of the complete surgical specimen was consistent with a desmoplastic fibroma. This case is the first report of a successful application of endoprosthetic reconstruction after desmoplastic fibroma resection.

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Introduction

Desmoplastic fibroma (DF) is an extremely rare primary bone tumor characterized by a nonmetastatic aggressive behavior [1]. These locally destructive neoplasms represent around 0.06% of all bone tumors and 0.03% of benign bone tumors [1,2]. DF was first described in 1958 by Jaffe, who highlighted their histological resemblance to a desmoid tumor [3]. Further histologic characterization portrayed a neoplasm consisting of spindle to stellate cells with minimal cytological atypia and an abundant collagenous matrix [4]. The most affected bone by DF is the mandible (22%). Other less-frequent sites include the femur (15%), pelvic bones (13%), radius (12%), and tibia (9%) [1,5]. Patients typically present before their third decade of life with a painful mass requiring wide resection to decrease the risk of recurrence [6]. However, even when wide margin resection is preferred due to better results (ie, 17% recurrence rate), it has been associated with poor functional outcomes in massive tumors [7–9]. In this report, we present the wide resection of an extensive DF on the distal portion of the femur reconstructed with a distal femoral replacement as a salvage treatment alternative to preserve the patient’s functionality and quality of life. This case adds to the literature the first successful use of endoprosthetic reconstruction after wide resection of a DF on the distal femur.

Case history

A healthy 41-year-old Hispanic man was diagnosed with fibrous dysplasia (FD) of the left distal femur 14 years ago (Fig. 1). He was managed with a retrograde intramedullary nail as a prophylactic fixation on the weakened bone (Fig. 2). Thirty-six months later, the patient complained of ongoing pain in his left knee, but he remained able to perform his daily activities and ambulate without difficulty. Orthogonal views of the femur were remarkable for significant local progression (Fig. 3). The patient was scheduled for an open bone biopsy to rule out any malignant transformation. The pathologic report was consistent with FD, and the patient...
continued under observation without indications for further management.

Five years later, the patient reported a new onset of ongoing knee pain that intensified over a few months. He denied any recent trauma or injury. No constitutional symptoms such as fever, malaise, weight loss, or night sweats were reported. Physical examination was only remarkable for mild tenderness to palpation over his left knee. Radiographs of the knee and femur showed hardware in place, with no signs of pathologic fracture or dislocation. A second biopsy was performed to evaluate for any malignant behavior. The pathologic assessment remained consistent with FD (Fig. 4), and the patient continued with observation requiring sporadic over-the-counter pain management.

The patient continued without further complications for the next 4 years. However, one morning, he felt a sudden sharp pain on his left knee when he was walking at home, followed by difficulty ambulating. At a local emergency room, anteroposterior and lateral radiographs of the left knee showed the well-known osteolytic lesion, hardware failure with a broken screw, and decreased intraarticular space (Fig. 5). The patient was discharged home with instructions to avoid weight-bearing on the left lower extremity and requested follow-up with an orthopaedic surgeon. At the
musculoskeletal oncology clinic, the patient was scheduled for removal of hardware after providing consent. The intervention proceeded without complications. The patient was oriented about the substantial femoral bone weakening and the concomitant joint degeneration that were causing joint instability and affecting his range of motion (−5° to 75°). The case was discussed in a multidisciplinary team led by the musculoskeletal oncology service and joint reconstruction service. Based on the patient’s age, absence of comorbidities, and functional recovery potential, wide resection of the tumor with a distal femoral replacement was recommended. The patient was fully informed about the current scenario and the recommended treatment alternative. The risks and benefits of the procedure were explained to the patient, to which he agreed and consented.

The patient was taken to the operating room for wide excision of the left mid to distal femur with reconstruction using an oncologic rotating hinge prosthesis (DePuy Synthes, Warsaw, IN). The tumor was approached through an anteromedial longitudinal incision followed by a parapatellar arthroscopy with soft-tissue dissection. The femur was osteotomized based on disease-free margins, and the mass removed (Fig. 6). Then, the femoral canal was reamed to 17 mm to place a 17.5-mm stem. The proximal tibia was cut 14 mm from the surface, and a large tibial metaphyseal sleeve without stem was placed after canal preparation. After an adequate reduction trial, the permanent components, including 125-mm augments and a 14-mm insert, were implanted press fit (Fig. 7). In addition, polymethyl methacrylate and antibiotics were applied to the tibial baseplate. The joint was irrigated, a drain was placed, and a layered closure was performed (Fig. 8). The patient was discharged home on the third postoperative day without complications. The mass was sent to a major referral center for pathologic evaluation. The report described a paucicellular benign tumor with a collagenous matrix but no significant atypia or mitotic activity consistent with a DF (Fig. 9a and b).

At the first follow-up visit, the patient was doing well, ambulating with a cane without distance limitations. Physical examination reveals a surgical incision that was healing properly and an adequate range of motion (0° to 100°). After 24 months, the patient remains without signs of recurrence and can perform all the activities of daily living.

**Discussion**

We have described a case of DF of the distal portion of the femur that was initially diagnosed as FD. The initial assessment was based on conventional radiographs showing an expansile lytic lesion and several biopsies performed through the years. It was not until the histopathologic evaluation of the wide resection specimen that the diagnosis of DF was established. Due to the aggressive local behavior of the skeletal lesion, including substantial disruption of the femoral anatomy, we planned a wide resection followed by a distal femoral replacement as a limb-salvage procedure to restore the patient’s functionality and quality of life.

DF is an extremely rare benign bone tumor that may affect patients of any age or sex [10]. Their reported incidence is about 2.5 patients per 100 million people per year [6]. Its clinical...
manifestations usually include pain and swelling in the affected region and symptoms produced by mass compression on the surrounding tissue [2]. Other patients may debut with a pathologic fracture. Radiologically, these tumors are centrally located, expansile osteolytic lesions with honeycombed appearance secondary to an irregular trabecular formation through the lytic regions [2,11,12]. Their diagnosis is challenging on radiographical imaging alone as many tumors such as giant cell tumors, aneurysmal bone cysts, FD, non-ossifying fibroma, chondromyxoid fibroma, and eosinophilic granuloma have similar radiological appearances [4,11]. Yet, the presence of a lytic bone lesion in a young patient involving a typical site like mandible or long bones should alert clinicians of a possible DF [10,11]. Advanced imaging, such as computed tomography and magnetic resonance imaging, are helpful techniques for further characterization and delimitation of the tumor, providing a more precise picture of the bony architecture to guide surgical management [4,11,13].

Bone biopsy remains the gold standard for DF diagnosis [2,4]. The histopathologic assessment is characterized by slender fibroblastic to reticular cells without pleomorphism or mitotic activity embedded in a copious collagenous matrix [14]. However, their histologic appearance could be deceiving as it overlaps with other benign bone lesions such as FD. In that context, the lack of osseous metaplasia serves to aid in differentiation between those 2 entities as there are no specific immunohistochemical markers for DF [14,15]. Furthermore, there have been rare reports of DF and FD coexisting within the same lesion [12].

Due to their local aggressive behavior, DF is usually surgically managed. Treatment alternatives include intralesional curettage, which is considered in areas of significant functional deficit (ie, knee), or resection (intralesional, marginal, or wide), which is associated with better outcomes [4,6,12]. In massive or recurrent lesions, amputation could still be considered [4,7,9]. Bohm et al. evaluated 191 cases of DF reporting a recurrence rate of 55% with curettage and 17% with wide resection [7]. In this perspective, wide resection has remained the primary treatment approach for DF [4,8–10]. However, despite better curative rates with wide tumor resection, the functional prognosis could be compromised when dealing with massive lesions [8,9]. In those patients, conservative surgery (ie, curettage) persists as a treatment option if surgical

Figure 5. Radiographs showing hardware failure and broken screw on the left femur.
resection would translate into significant morbidity or loss of functionality [4,9,10,16]. Ultimately, radiation therapy could reduce the pain and limit tumor progression in inoperable tumors [6].

In the current case, wide resection was considered based on the anatomic location and the aggressive tumor behavior, including articular surface collapse and deterioration of knee function. Centered on the poor functional prognosis, given the substantial proportions of the lesion, an endoprosthetic reconstruction was planned to restore anatomy and functionality. We opted for an oncologic rotating hinge prosthesis with a metaphyseal stemless sleeve for the tibial component based on the surgeon’s preference, the excellent clinical improvement, and the leg alignment restoration that can be achieved with this technique [17]. Previous literature had described the potential of limb-salvage surgery as a replacement for amputation in patients with primary bone tumors of the distal femur [18,19]. However, to the best of our knowledge, the successful use of a distal femoral replacement as a limb-salvage technique after wide resection of a DF on the distal femur has not been previously reported in the literature. Clinical and functional outcomes were satisfactory. Endoprosthetic reconstruction represents a valuable surgical alternative in musculoskeletal oncology to restore function and preserve the quality of life after DF resection.

**Summary**

This case demonstrates the natural history of a lesion on the left distal femur initially diagnosed as FD. We performed a wide-margin excision and a distal femoral replacement due to the aggressive local behavior of the tumor. Histologic examination of the complete surgical specimen revealed a DF diagnosis. Endoprosthetic reconstruction represents an effective treatment alternative in the setting of a DF in the distal femur when the integrity of the bone is compromised after wide tumor resection. Appropriate surgical planning using a distal femoral replacement prosthesis restored anatomy and functionality in our patient.

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**Conflict of interest**

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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**Informed patient consent**

The authors confirm that written informed consent has been obtained from the involved patient and, he has approved this information to be published in this case report.
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Figure 8. Postoperative radiographs of the left thigh immediately after surgery.

Figure 9. Pathology slides showing the desmoplastic fibroma in hematoxylin and eosin stain (a) in medium-power microscopy and (b) high-power microscopy.
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