Radiotherapy for desmoplastic fibroma of bone: A case report

INTRODUCTION

Desmoplastic fibroma of bone is a rare, locally aggressive, and benign skeletal tumour. Though benign in nature, its rate of recurrence after curettage is reported to be very high. Pathological fractures have been reported but metastasis and death are rare. Surgical excision with a wide margin of normal tissue is considered the treatment of choice. Radiotherapy is given in cases where surgery could not be performed. Few cases have been reported to support the effectiveness of radiotherapy in treating the skeletal desmoplastic fibroma; no recurrence was reported in these cases. We present a case of desmoplastic fibroma of the femur of a woman, which was treated primarily with radiotherapy.

ABSTRACT

Desmoplastic fibroma is a rare benign tumour of bone. Diagnosis is not easy and is often made by excluding other tumours. Histopathological diagnosis of this tumour is also sometimes not easy. The treatment modalities for this tumour are non-uniform and often controversial. In the present case surgical options were left aside because the patient did not consent to surgery, so radiotherapy was used, with success at 3-year follow-up. This case is presented here along with a review of relevant literature.

Key words: bone; fibroma, desmoplastic; radiotherapy

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CASE REPORT

A 28-year-old Indian woman presented to the Department of Orthopaedics at the All India Institute of Medical Sciences, New Delhi, India, in August 1996 with a 2-year history of pain around the left knee. The pain had initially been dull, deep-seated, and intermittent, but completely relieved by the occasional oral administration of an analgesic. Pain gradually progressed in intensity, and a noticeable limp had appeared during the month before admission. The patient reported requiring continuous analgesic treatment. Results of general physical examination at presentation were unremarkable. Wasting of the left thigh was noted. There was minimal effusion in the knee joint, and flexion was terminally restricted. Tenderness was localised to the supracondylar region of the femur.

X-rays showed a metaphyseal lesion in the distal end of the femur and a permeative pattern of destruction (Fig. 1). The margins of the lesion were ill-defined and no evidence of matrix mineralisation was visible; the cortex was not breached. The erythrocyte sedimentation rate was 34 mm in the first hour. Total leukocyte count was 0.5x10⁹/L, with neutrophils 66%, lymphocytes 32%, and eosinophils 2%. No abnormal cell was noted in the peripheral smear. Random glucose level was 90 (60–140) mg/dL, urea 34 (10–50) mg/dL, creatinine 0.8 (0.5–1.8) mg/dL, and serum alkaline phosphatase 290 (80–280) U/L.

![Figure 1](a) Anteroposterior and (b) lateral X-rays of the left knee at initial presentation, showing an ill-defined metaphyseal lesion with a permeative pattern of destruction.
Figure 2  Photomicrograph showing spindle cells with dense collagenisation between cells and an entrapped remnant of bone in one corner (H&E, x240).

Figure 3  (a) Anteroposterior and (b) lateral X-rays of the knee at 3-year follow-up, showing sclerosis and regression of the lesion.
The Mantoux test reading was 12 x 12 mm after 48 hours. Incisional biopsy revealed desmoplastic fibroma (Fig. 2). Surgical options (excision of the lower end of the femur with allograft reconstruction, custom-made prosthesis, arthrodesis, or rotation plasty) were deemed unacceptable by the patient. She was therefore referred to the Department of Radiotherapy for radiotherapy. Radiotherapy was given to the left femur by a 6-megavolt beam generated by a linear accelerator (SL 20; Philips, UK) at the anterior and posterior portals. The patient received a dose of 45 grays in 25 fractions delivered over 5 weeks. The entire regimen of radiation was well tolerated by the patient, without any complication or break.

Symptomatically, the patient has been showing improvement. At the last follow-up, 3 years after radiotherapy, she reported occasional pain on exertion and minimal analgesic requirement. She also reported being more concerned about thigh wasting than about pain. The X-rays taken at 36 months showed sclerosis, with no evidence of progression of the lesion (Fig. 3).

DISCUSSION

Desmoplastic fibroma of bone is a rare bone tumour, accounting for only 0.06% of all bone tumours. Fewer than 200 cases worldwide have been reported in the literature. The majority of patients in the reported cases are in their third decade of life, and there is no sex predilection. Although the tumour has been described in various locations of the body, it most commonly arises in the metaphyseal region of long bones. The lesion arises within the central portion of the bone and grows outwards. There is no radiographic evidence of matrix mineralisation. Because the tumour abuts the cortex, it stimulates endosteal resorption and a concurrent periosteal reaction that increases the diameter of the bone. With cortical thinning, a pathological fracture may occur. Radiographic differential diagnosis includes tuberculosis, chondromyxoid fibroma, low-grade intramedullary osteosarcoma, and fibrosarcoma. Histologically, the tumour consists of sheets of fibroblastic cells in a collagenous matrix. The most important differential diagnosis is low-grade fibrosarcoma; however, making the distinction may not always be possible.

Performing an en bloc resection with a wide margin of normal tissue is the treatment of choice, and it may be curative. In our case, limb salvage would have required excision of the lower end of the femur, followed by limb reconstruction. The options available are prohibitively costly (e.g. allograft reconstruction and custom-made prostheses) or seriously impair function (e.g. arthrodesis and rotation plasty). Because the histological features of desmoplastic fibroma are generally similar to those of soft tissue fibromatosis, Enneking has speculated that radiotherapy may be effective in achieving local control. Effectiveness of radiotherapy in soft tissue fibromatosis is well documented. However, there is only one report in the literature in which radiotherapy was used as the definitive treatment of desmoplastic fibroma of the ilium.

In this case, the patient did not consent to the surgical options available, and a trial of radiotherapy was offered as an alternative. After 36 months, the symptomatic benefit was apparent, and X-rays showed regression of the lesion.

Undoubtedly, surgery remains the treatment of choice for this very uncommon tumour. However, in situations in which surgery is not feasible, radiotherapy may be considered as an alternative mode of definitive management. Further follow-up of this case and of more cases treated in a similar manner will help in clarifying the role of radiotherapy in the management of desmoplastic fibroma of bone.

REFERENCES