Limb salvage surgery complimented by customised mega prostheses for malignant fibrous histiocytomas of bone

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ABSTRACT

**Purpose.** To assess functional and oncological outcomes of patients with malignant fibrous histiocytomas of bone, after limb salvage surgery complimented by a customised prosthesis.

**Methods.** Between May 1991 and December 2002, 15 men and 5 women (mean age, 42 years) with histologically proven malignant fibrous histiocytoma of bone underwent treatment involving limb salvage surgery complimented by a customised mega prosthesis. Most of the tumours were stage II according to the Enneking system, and located around the knee. Wide resection margins were achieved in 18 patients.

**Results.** Following a mean follow-up of 58 months, 4 patients underwent amputation for local recurrence and 5 died of the disease. Two patients had prosthesis fractures; revision of the prosthesis was carried out in one. The functional result was excellent in 5 and good in 9 patients. The Kaplan-Meier 5-year survival rates of the patients treated without chemotherapy and with chemotherapy were 50% and 76%, respectively.

**Conclusion.** Limb salvage surgery with chemotherapy is a viable treatment option for patients with malignant fibrous histiocytoma of bone. It achieves higher survival rates than resection alone. Such therapy improves quality of life and provides a useful and functional limb.

**Key words:** bone neoplasms; histiocytoma, malignant fibrous; limb salvage; prosthesis implantation; survival rate

INTRODUCTION

Malignant fibrous histiocytoma (MFH) of the bone is a rare tumour of unknown origin, accounting for <2% of all primary malignant bone tumours.² Being aggressive, it is characterised by a high frequency of local recurrence and metastasis to regional lymph nodes and distant sites. A combined approach involving wide surgical resection and neo-adjuvant chemotherapy is recommended.²⁻⁴ Surgical reconstruction techniques include the use of customised prostheses, resection-arthrodasis, allograft, or rotationplasty depending on the tumour’s location and extent of resection.
Customised prostheses give good limb function.

**MATERIALS AND METHODS**

Between May 1991 and December 2002, 15 men and 5 women aged 14 to 60 (mean, 42) years with histologically diagnosed MFH of bone underwent limb salvage and reconstruction with a customised mega prosthesis. The knee was most commonly involved (distal femur [n=9], proximal tibia [n=5]), followed by proximal femur (n=3) and proximal humerus (n=3) [Table]. 14 patients treated after 1996 received chemotherapy. Assessments were made using radiography, computed tomography (CT), magnetic resonance imaging (MRI), technetium bone scanning, and tumour angiography. Tumour staging was carried out according to the Enneking system.\textsuperscript{5,6} All defects were reconstructed with custom-made prostheses.

![Table](image)

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex/age (years)</th>
<th>Involved site</th>
<th>Tumour stage</th>
<th>Prosthesis type*</th>
<th>Resection margin</th>
<th>Chemotherapy</th>
<th>Follow-up (months)</th>
<th>Complications</th>
<th>Functional result</th>
<th>Oncological result†</th>
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<td>Fair</td>
<td>DOD</td>
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<td>TKP</td>
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<td>Wide</td>
<td>Yes</td>
<td>24</td>
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<td>CDF</td>
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</tbody>
</table>

Note: TKP denotes total knee prosthesis, PHP proximal humeral prosthesis, and PFP proximal femoral prosthesis. DOD denotes died of disease, NED no evidence of disease, and CDF continuously disease free.
8–152) months. Survival rates were derived using the Kaplan-Meier survivorship analysis.

RESULTS

Five patients attained excellent, 13 good to fair, and 2 poor functional outcomes. All those with proximal femoral tumours had satisfactory outcomes; the 2 patients with poor outcomes and half of those with fair outcomes had distal femoral tumours. Three of the 5 patients with proximal tibial tumours had excellent outcomes (Fig. 1). Regarding the 3 patients with proximal humeral tumours, one each had excellent, good, and fair outcomes. Overall, functional outcomes in distal femoral tumours were worse than at other sites (Table).

12 patients remained disease free and 3 had no evidence of disease after amputation for local recurrence. Five patients died of the disease with widespread metastases. The 5-year survival rate in the 6 patients who did not receive chemotherapy was 50%, and it was 76% in the remaining 14 patients who received chemotherapy (Fig. 2).

One patient with a distal femoral tumour developed wound infection that settled with antibiotics. One patient with a proximal tibial tumour developed skin necrosis, and was treated by skin cover. Three patients with distal femoral tumours and one with a proximal tibial tumour developed local recurrence; all 4 were treated by amputation. Two patients with total knee prosthesis had fractures of the femoral stem; one of whom underwent revision of the prosthesis (Table).

DISCUSSION

MFH is an unusual, high-grade primary tumour that arises either from fibroblasts or tissue histiocytes. Histologically it consists of rounded, histiocyte-like, spindle-shaped, fibroblast-like cells, arranged in a storiform or whorled pattern. MFH of bone was first described in 1972. It usually arises from soft tissues and accounts for approximately 2% of all primary malignant bone tumours. Up to 25% of these tumours occur secondary to pre-existent benign conditions (Paget’s disease, irradiated bone, bone infarcts, enchondroma, and fibrous dysplasia). Patients with such secondary tumours are often older.

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**Figure 1** Preoperative (a) radiographs and (b) coronal T1-weighted magnetic resonance images showing malignant fibrous histiocytoma of the proximal tibia. (c) Follow-up radiographs after reconstruction with total knee prosthesis.

**Figure 2** The Kaplan-Meier 5-year survival rates of the malignant fibrous histiocytoma patients treated without chemotherapy and with chemotherapy were 50% and 76%, respectively.
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than those with the primary tumours; the secondary type was noted in 3 of our patients, 2 of whom had a history of radiation and one had fibrous dysplasia.

The predilection of locations for MFH of bone is similar to that of osteosarcoma of bone. Commonly involved sites are: distal and proximal femur, proximal tibia, and proximal humerus; approximately 50% of the tumours occur around the knee. In our series, the femur was the commonest site (60%), compared to 32 to 50% reported by others. The mean age of presentation (42 years) and male predominance were similar to findings by others, though female preponderance has also been reported. A special feature of MFH of bone is the bimodal age pattern, which is unlike that for other malignant tumours of bone.

MFH of bone has a tendency to infiltrate adjacent medullary regions gradually, without causing much tissue response or reactive ossification. Hence radiographs must be interpreted with care, as they may underrepresent the true extent of disease involvement. Also, being large destructive lesions, they may lead to pathological fractures (as in 5 of our patients).

The principles of treatment are similar to those for non-metastatic osteosarcoma. Treatment decisions are based on the stage of the disease, as proposed by Enneking. The mainstay of treatment is surgical resection or amputation. Chemotherapy appears to prolong survival and preoperative and adjuvant chemotherapy reduces the risks of microscopic residual and metastasis. Favourable results are reported with limb salvage surgery and chemotherapy for stage-IA, -IB, or -II tumours.

Endoprosthetic reconstruction following local resection of MFH of bone has been reported. In our series, the prostheses were custom-made, using either stainless steel or titanium. Preoperative full-length radiographs were used to estimate the dimensions of the prostheses. Templates were used to manufacture the appropriate endoprostheses. CT and MRI helped estimate the extent of tissue involvement and hence facilitated planning of the level of resection. The accuracy of MRI in determining the extent of tumour spread and its effectiveness in preoperative planning has been reported.

It is important to resect adequate surgical margins to prevent recurrence. Recurrence rates of 64% with inadequate margins, 19% with wide margins, and 6.5% with radical surgery have been described. Also, when wide surgical margins were obtained, there was no difference in the recurrence rates between amputated and limb salvage patients. Two patients in our series with proximal humeral and distal femoral MFH had marginal and contaminated margins respectively. Extensive resections to achieve wide surgical margins in these circumstances would have resulted in large bone defects, posing problems for reconstruction. These 2 patients refused amputation and wanted a functional limb, though they were aware of the increased risk of recurrence and death. One of them later developed recurrence and underwent amputation.

One technical problem with reconstruction for proximal tibial tumours is the restoration of the extensor mechanism. Various methods have been proposed, including: special bands, tubes, and Dacron tapes augmented with bone grafts. We used Dacron tapes to reattach the extensor mechanism to the prosthesis. Two of the 5 patients in our series with proximal tibial tumours had 20° and 10° of extensor lag, which resulted in fair and good functional outcomes, respectively.

Primary MFH of bone is often highly responsive to chemotherapy; some even advocate chemotherapy as the primary treatment and surgery as adjuvant therapy. In our series, patients treated before 1996 did not receive preoperative chemotherapy and had a less favourable 5-year survival rate of 50%; 3 died of the disease after 36, 12, and 8 months of follow-up and 2 developed local recurrence managed by limb ablation. When limb salvage surgery was combined with chemotherapy, the 5-year survival improved to 76%, comparable to other reports of 80% and 66% after 2 and 3 years, respectively. Another series using surgery alone reported a 5-year survival of 28%, which improved to 57% when surgically treated patients also received chemotherapy. Both series noted a worse prognosis in older patients with secondary tumours.

Prosthetic fracture is uncommon and reported in 1.6 to 16% of patients. Poor component design, material properties, and high stresses due to bone deficiency after resection have been implicated as potential causes. Some have reported reduction in implant failures with the use of larger core stems (>12 mm in diameter). Further improvements in the material properties of the prostheses by using forged as opposed to cast material might decrease such complications. Two patients in our series had prosthetic fractures; both were total knee prostheses. One was revised but later developed local recurrence treated by amputation. With the use of larger core diameter stems made of forged material, liability to this complication appears to have decreased.

Limb salvage with custom-made prosthesis was more emotionally acceptable than amputation. In
13 (65%) of our patients, it provided good stability, painless mobility, and an overall satisfactory functional outcome. Seven patients had poor outcomes due either to oncological or mechanical complications. Patients who underwent limb salvage surgery with chemotherapy had higher survival rates than those treated with resection alone. This improved quality of life and provided a useful, functional limb.

REFERENCES