Endoprosthetic treatment of primary bone sarcomas with pathological fractures

Dr Thomas L Hilton MBChB(UCT), DA(SA), DipPEC(SA), FCS(Orth), MMed(UCT)
Dr Keith Hosking MBChB(UCT), FCOrth(SA)
Vincent Pallotti Life Orthopaedic Hospital, and Princess Alice Orthopaedic Unit, Groote Schuur Hospital, University of Cape Town

Correspondence:
Dr TL Hilton
11 Clive Street
Vredehoek
8000 Cape Town, South Africa
Email: thilton@hotmail.com
Cell: 0027 (82) 796 7608

Abstract

Background: Primary bone sarcomas that are associated with a pathological fracture are rare and as a group have a worse prognosis than their non-fractured counterparts.

Questions/purposes: Traditionally limb ablation was advised; however, recent evidence suggests that limb salvage is a safe and acceptable form of treatment for both surgeon and patient.

Patients and methods: We present a retrospective review of a series of six patients referred to our unit with pathological fractures. These were treated by initial traction and neo-adjuvant chemotherapy where indicated with subsequent resection and endoprosthetic replacement.

Results: The age range of our series is from 20 to 81 years, with four males and two females. All had distal femur involvement with a 60% incidence of osteosarcoma and 40% chondrosarcoma. Three patients required total femur resection due to extensive tumour involvement. Our results show 100% of patients had clear margins at post-operative histology. Due to the aggressive nature of these types of tumours they carry a worse long-term prognosis and as such we had three deaths in our series. One patient died of a myocardial infarction post-operatively, and two patients developed lung metastases and died 2 years later.

Conclusion: Our conclusion is that with careful planning, a safe margin can be achieved. Endoprosthetic replacement allows for rapid reconstruction and mobilisation in this group of patients facilitating further oncological management.

Key words: pathological fracture, sarcoma, limb salvage, endoprosthetic replacement

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Introduction

Primary bone sarcomas that are associated with a pathological fracture (Figures 1–4) are rare and as a group have a worse prognosis than their non-fractured counterparts. It was previously thought that this problem should be treated with amputation to prevent local recurrence, reduce incidence of metastases and improve survival. More recent studies have shown that careful selection of patients, initial immobilisation, and neo-adjuvant chemotherapy where indicated, leads to fracture healing and facilitates adequate resection of margins subsequently. This treatment results in comparable survival rates to amputation with the advantages of a cosmetically and functionally superior limb.

Materials and methods

We performed a retrospective review of a series of six patients referred to our unit with pathological fractures from 2009 to 2014. The patients were admitted and treated with bed rest and skin traction until fracture union. Skeletal traction was avoided to minimise septic complications.
During their admission, an MRI scan of the entire involved femur was done to assess the site and size of the lesion, its proximity to neurovascular structures, and to exclude skip and satellite lesions. A CT chest and abdomen including a nuclear medicine bone scan was performed to stage the patient and assess for the presence of metastases. A needle biopsy for histological confirmation of diagnosis and grading of the tumour was performed.

Osteosarcoma patients underwent neo-adjuvant chemotherapy prior to surgery to facilitate fracture union and tumour shrinkage. No patient received radiotherapy prior to surgery. Patients were consented and counselled about the possibility of an amputation if the macroscopic appearance of the tumour was unfavourable at the time of surgery.

Surgical treatment included limb salvage surgery with wide excision of the tumour and endoprosthetic replacement (Figures 5–7). Rehabilitation was per the unit’s protocol.

Results

Of the six patients, the age range was from 20 to 81 years, with four males and two females. The follow-up period was at a median of 12 months from time of presentation (range min 5, max 61). All patients had distal femur involvement with a 60% incidence of osteosarcoma and a 40% incidence of chondrosarcoma. Four of the six patients were diagnosed with a primary bone sarcoma and pathological fracture at their index presentation. Two of the patients were undergoing neoadjuvant chemotherapy for their primary bone sarcoma at the time of fracture. No patients sustained an intra-operative fracture. Three patients required total femur resection due to extensive involvement of their disease.

MRI was essential in surgical planning to ascertain the degree of soft tissue involvement, intramedullary extent, as well as proximity to the neurovascular bundle.

All of the sarcomas were high-grade and extra-compartmental in nature. A marginal or wide excision margin was achieved in all cases. Due to the aggressive nature of these high-grade sarcomas, they carry a worse long-term prognosis and we had three deaths in our series. One patient died of a myocardial infarction post-operatively, and two patients developed lung metastases, undiagnosed at initial presentation and died 2 years after completion of initial treatment. One of the two patients who died from the disease developed local recurrence 2 years after initial presentation and underwent limb ablation.
Figures 5, 6 and 7. Intra-operative image of excised tumour and endoprosthetic insertion

Figures 8, 9 and 10. X-ray and intra-operative images of a primary sarcoma with pathological fracture treated with intramedullary nailing. The result was extensive spread of tumour throughout the femur, necessitating endoprosthetic replacement.
Discussion

The principal goal in the management of patients who have a primary bone sarcoma is prolonging their survival. Most studies comparing amputation and limb salvage report no adverse effects of the long-term survival of patients with primary bone sarcomas treated with limb-sparing surgery. The current trend for the treatment of pathological fractures as a consequence of primary bone sarcomas is still towards limb salvage; however, this has not always been the case. Amputation was the most common procedure of choice. It was recommended because it was thought that the local fracture haematoma disseminated tumour cells into the adjacent tissue and joints. Also, it was thought that damage to the microvascular circulation facilitated metastases. Factors that changed the trend towards limb salvage included the efficacy of neo-adjuvant chemotherapy, fracture union during pre-operative chemotherapy, and the newer functional reconstructive modalities of treatment and imaging.

Pathological fractures occur due to the high-grade and lytic nature of these sarcomas. Fractures can occur spontaneously or after minimal trauma, because of the poor structural properties of the bone affected. The pathological bone often exhibits high cellularity, poor differentiation and significant loss of bone matrix. Stress or mechanical weakness caused by diagnostic biopsy and necrosis of tumour after chemotherapy may contribute to development of fractures.

The incidence of a pathological fracture either at diagnosis or during the peri-operative period is between 5% and 10%. It is important to both make a prompt diagnosis of a pathological fracture and not to miss the diagnosis in the first place, as internal fixation of these fractures may result in tumour spread and impairment of definitive treatment.

As mentioned earlier, amputation was the procedure of choice for these injuries to ensure optimal outcome in terms of recurrence and survival. This is supported by studies that show that amputation produces a better

Figures 11 and 12. Post-operative radiographs demonstrating the endoprosthetic replacement of the pathological lesion

Figures 13 and 14. Clinical photographs demonstrating excellent functional results of megaprosthetic replacement
outcome in the eradication of the local tumour than limb salvage.\textsuperscript{6,7,8,10} However, recent studies have shown no difference in the outcome after limb salvage, including tumour recurrence, the development of metastases and overall survival in patients who presented with a fracture and those who developed a fracture with treatment.\textsuperscript{2,8,29}

Neo-adjuvant chemotherapy improves overall survival in osteosarcoma assisting with tumour shrinkage and union.\textsuperscript{1,12} Without chemotherapy survival rates are as low as 20%,\textsuperscript{13,22} but with chemotherapy increase to 60% overall.\textsuperscript{22} Patients are treated with bed rest and skin traction while undergoing chemotherapy until fracture union. This aids in the manipulation of the limb during the definitive surgery.\textsuperscript{12,22,30} Radiation post limb salvage is not recommended as it has not been shown to be successful in preventing local recurrence or metastases, and it may compromise success of definitive surgery and any potential further surgery.\textsuperscript{2,22}

MRI is extremely useful in the planning of the definitive surgery. A thorough MRI assessment before and after chemotherapy is useful to determine the extent of the initial haematoma and the presence of skip or satellite lesions. This helps to plan the extent of the tumour excision and type of endoprosthetic required. Limb salvage is usually offered unless the tumour cannot be separated from important neurovascular structures, has grossly invaded a joint or where resection of too much muscle would result in a functionless limb.\textsuperscript{3} The reconstructive option is usually with a megaprosthetic or allograft prosthetic composite, which gives the patient a good cosmetic and functional result (Figures 13 and 14). In keeping with Pemister’s law that bone sarcomas have a predilection to occur around joints due to the relative increase in blood supply, the majority of these primary bone sarcomas that sustain pathological fracture occur around the knee and usually involve the joint, necessitating its inclusion in the prosthesis. Sarcomas tend to spread along muscle compartments and are limited by fascial planes. The pathological fracture is usually a low energy process with less soft tissue contamination than conventional fractures, and surgery within certain tissue planes is possible.

The incidence of local recurrence has been reported to be approximately 19%, which is consistent with our study, which showed a recurrence rate of 17%.\textsuperscript{10} The number of deaths unrelated to the tumour or its treatment was slightly higher in our series (17%) compared to reported literature of approximately 8%; however, this may be due to our small numbers.\textsuperscript{2,22,30} Deaths related to the tumour in our series were 33%, compared to 47% reported in the literature.\textsuperscript{2,22,30} The prognosis for these patients is guarded from the outset due to the often aggressive and high-grade nature of the sarcomas that are associated with pathological fracture. In the past the prognosis of patients presenting with this diagnosis was uncertain; however, recent studies suggest that with modern treatment it is similar to those without fracture.\textsuperscript{10,21,22}

Conclusion

Our conclusion is that one can achieve safe local eradication of the tumour and perform limb salvage surgery for primary bone sarcomas with pathological fractures, without affecting patient morbidity or mortality while sparing the patient an amputation. The prognosis for these patients is poor from the outset, but the advantages of limb salvage for the patient are those of improved cosmetic and function gains.

Compliance with Ethics Guidelines

T Hilton and K Hosking declare that they have no commercial associations that might pose a conflict of interest in connection with the submitted article. Work for the article was performed at Groote Schuur Hospital and Vincent Pallotti Life Orthopaedic Hospital.

References


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