Surgical management of primary bone sarcomas

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Abstract

This article addresses the surgical management of bone sarcoma and includes all the main tumour types, for example chrondrosarcoma, osteosarcoma and Ewing's sarcoma of bone. The article is aimed at the experienced orthopaedic surgeon who may not have specific knowledge of dealing with musculoskeletal tumours. Important principles, such as biopsy, are discussed and appropriate resection methods described. The use of endoprosthetic replacement is a crucial tool in the orthopaedic oncologist's armoury and principles of use are described. Adjuvant therapy such as chemotherapy and radiotherapy is also discussed and some common protocols are also described.

Keywords bone sarcoma; chondrosarcoma of bone; endoprosthetic replacement; Ewing's sarcoma; orthopaedic oncology; osteosarcoma

Introduction

The surgical management of bone sarcomas has changed over the last 30 years from predominantly amputation to an approach that is focused on functional limb salvage as a result of improved oncologic management and advances in biological orthoplastic reconstruction techniques and implant design. Surgery remains pivotal in the treatment of bone sarcoma notably chondrosarcoma and an understanding of the principles involved is essential for any orthopaedic surgeon.

There are a number of surgical reconstructive options which are available, with the use of implants and biological techniques including custom and modular endoprosthesis, allograft, vascularized free fibular grafting (VFFG) and bone transplant. Some bone sarcomas require neo-adjuvant chemotherapy and timing of surgery is of utmost importance.

The management of bone sarcoma may begin with neo-adjuvant therapy in Ewing's sarcoma and osteosarcoma or may rely on surgical treatment alone in chondrosarcoma.

Biopsy

Biopsy is important because it determines the tumour type and grade. Where possible, a biopsy is undertaken under radiological control. This ensures that the most representative region identified by various radiologic methods is biopsied. A percutaneous image-guided biopsy also ensures that there is no delay in starting any neo-adjuvant treatment (which may occur with open biopsy whilst waiting for the wound to heal). When a percutaneous biopsy is inconclusive, an open biopsy is of course mandatory (Tables 1 and 2).

The orientation and location of the biopsy tract are critical. Before biopsy, the surgeon should review the radiographs with the pathologist to plan the biopsy site. As in percutaneous biopsy, one should attempt to factor in the future skin incisions needed for definitive surgery. Whenever possible the surgeon should try to avoid using drains, however if needed these should exit either from the corner of the wound or close to the skin incision that will make resection straight forward to include in future approaches. Transverse incisions should be avoided.

The surgeon must ensure careful attention to haemostasis to prevent haematoma formation and subcutaneous haemorrhage. Biopsy incisions should ideally be made through muscle compartments so that the muscle layer can be closed tightly. Neurovascular structures are avoided. A tourniquet is used to obtain tissue in a bloodless field and are then released so that bleeding points are fully controlled.

All biopsy samples should be submitted for microbiological culture and sensitivity. Antibiotics should not be delivered until the cultures are obtained (Table 3).

Treatment: general principles

The goal of the treatment of malignant bone tumours is to remove the lesion with a clear margin to minimize the risk of local recurrence.

Wherever possible, limb salvage is employed. This is only possible where two essential criteria are met:

- local control of the lesion must be at least equal to that of amputation surgery
- the limb that has been saved must be functional.

Surgical margins are graded according to the system of the Musculoskeletal Tumor Society.

- **Intralesional margin:** The plane of dissection goes directly through the tumour. When the surgery involves malignant tumours, an intralesional margin results in 100% local recurrence. This should only occur when an inadvertent excision has been performed and sometimes in low-grade chondral lesions (see below). (It is also utilized for benign tumours such as giant cell tumours – see below and the article on Management of benign bone tumours elsewhere in this issue; pages 151–160.)
- **Marginal margin:** A marginal line of resection goes through the reactive zone of the tumour; which contains inflammatory cells, fibrous tissue, and areas of tumour cells. When malignant tumours are resected through the reactive zone probably results in a local recurrence rate of 25–50%. A marginal margin may be safe and effective if the response to preoperative chemotherapy has been excellent (95–100% tumour necrosis).
- **Wide margin:** This is the preferred margin. A wide line of surgical resection is accomplished when the entire tumour is removed with a cuff of normal tissue. The local recurrence rate is around 10% when such a surgical margin is achieved.

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Bone lesions by age

Age (years)	< 5	< 30	> 30
Malignant	LCH (Letter-Siwe) LCH (Hand-Schüller-Christian) Metastatic Rhabdomyosarcoma Metastatic Neuroblastoma	Ewing's sarcoma Osteosarcoma	Chondrosarcoma Metastases Lymphoma Myeloma Chordoma Adamantinoma
Benign	Osteomyelitis Osteofibrous Dysplasia	Osteoid osteoma Osteoblastoma Chondroblastoma Aneurysmal bone cyst LCH Osteofibrous dysplasia Non-ossifying fibroma	Giant cell tumour Paget's disease

LCH, Langerhans cell histiocytosis.

Table 1

- **Radical margin:** A radical margin is achieved when the entire tumour and its compartment (all surrounding muscles, ligaments, and connective tissues) are removed (Tables 4 and 5).

Adjuvant therapy

- Chemotherapy
- Radiation therapy

Principles of musculoskeletal biopsy

Principle	Rationale
Longitudinal Incision in line with future resection	Longitudinal incision is extensile
Biopsy through a single compartment	Biopsy tract can be excised with final resection remaining extensile
Avoid critical structures, i.e. neurovascular bundles	Contamination of critical structures precludes limb salvage
Biopsy the soft tissue component when present	Bone is weakened when its cortex is disrupted
	Bone requires decalcification for evaluation and this process may affect pathology
Maintain strict haemostasis	Avoid increased contamination outside of the biopsy tract by iatrogenic tumour spread
Use a drain in line with the incision when needed	

Table 2

Tumour–bone interaction (from Lodwick)

Lesion	Type I	Type II	Type III
Radiographic appearance	Geographic A-sclerotic B-distinct C-indistinct	Moth eaten	Destructive
Examples	A-Non-ossifying fibroma B-Unicameral bone cyst C-Giant cell tumour	Osteomyelitis Metastases	Ewing's sarcoma

Table 3

The role of chemotherapy and radiotherapy are discussed in detail in the article on The non-surgical management of musculoskeletal malignancy elsewhere in this issue (pages 195–203; see also Table 6).

Osteosarcoma

- Spindle cell neoplasms that produce osteoid are arbitrarily classified as osteosarcoma.
- Many types of osteosarcoma (see below).
- The most common subtypes are 'classic' osteosarcoma, parosteal osteosarcoma, periosteal osteosarcoma, telangiectatic osteosarcoma, osteosarcoma occurring with Paget's disease, and osteosarcoma after irradiation.
- Historically, osteosarcoma was treated by amputation; long-term studies demonstrated a survival rate of only 10–20%, with metastatic lung disease being the commonest cause of death.

Classification of primary tumours of bone and bone matrix[a]

Histologic type	Benign	Malignant
Hematopoietic		Myeloma Lymphoma
Chondrogenic	Osteochondroma Chondroma Chondroblastoma Chondromyxoid fibroma	Primary chondrosarcoma Secondary chondrosarcoma Dedifferentiated chondrosarcoma Mesenchymal chondrosarcoma Clear cell chondrosarcoma
Osteogenic	Osteoid osteoma Osteoblastoma	Osteosarcoma Parosteal osteosarcoma Periosteal osteosarcoma
Unknown origin	Giant cell tumour (fibrous) histiocytoma	Ewing's tumour Malignant giant cell tumour Adamantinoma

[a] Classification is based on that advocated by Lichtenstein L: Classification of primary tumours of bone, *Cancer* 4:335–341, 1951.

Table 4