Focus On
Earlier diagnosis of bone and soft-tissue tumours

Bone and soft-tissue sarcomas are rare and represent around 1% of all malignant tumours. The incidence of bone tumours is between eight and nine per million population while for soft-tissue sarcomas this figure is about 30 per million population per year.1

A review of a prospective database of over 3000 patients with sarcomas at the Royal Orthopaedic Hospital (Birmingham) has shown that the mean size of sarcoma diagnosed in the past 20 years has been 10 cm. In addition, there is no convincing evidence that over time the size at diagnosis is decreasing for either bone or soft-tissue sarcomas. Delays in diagnosis for both are common at every step of the pathway, usually because of a low level of awareness of the possibility of malignancy.

For soft-tissue sarcomas the main prognostic factors are grade, size, depth, diagnosis and the age of the patient.2 Of these factors, the only one which can be altered, and thus improve prognosis, is the size of the tumour at diagnosis. Numerous studies have confirmed that tumour size is related to survival and correlates strongly with the incidence of detectable metastases at diagnosis.3-5 For every 1 cm increase in the size of a soft-tissue sarcoma at diagnosis there is a 3% to 5% decrease in the chance of cure.5,6 Although the effect of size on prognosis is less marked for bone tumours, large studies have confirmed its significance.7-10

In 1999, guidance was produced highlighting the clinical features suggestive of possible cancer for many tumour types including sarcoma.11 This was updated in 2005 and accompanied by the introduction of two-week-wait referrals which enforced a need for any patient with a suspected malignancy to be seen at a referral centre within two weeks.12,13

The guidance stated that a soft-tissue lump with any of the following characteristics should be considered malignant until proven otherwise.

- Larger than 5 cm
- Increasing in size
- Deep to the deep fascia
- Painful
- Recurrent after previous excision

The guidance also recommended that any patient with bone pain, particularly non-mechanical pain, should be referred for a radiograph and that the presence of any of the following features should lead to further investigation:

- Bone lysis
- New bone formation
- Periosteal elevation
- Soft tissue swelling

The main goal of this article, therefore, is to present simple algorithms (Figs 1 and 2) to help in the proper assessment of patients who present with a soft-tissue or bony lesion. All medical practitioners and orthopaedic surgeons in particular, must continue to have a high index of suspicion of any abnormal bone lesion or soft-tissue mass. To assume that a solitary lytic bony lesion without an obvious primary is a metastasis is dangerous; it should be treated as a primary bone sarcoma until proven otherwise histologically. Again, assuming that a solitary soft-tissue mass is a benign lesion and suitable for intralesional excision is also detrimental and could affect the patient’s prognosis and complicate further surgical treatment if wrongly excised. Any patient with a soft-tissue lump with worrying features (Fig. 2)
should be investigated by proper imaging and referred for biopsy at an appropriate centre.

It cannot be stressed enough that early diagnosis is the key to improving outcome. If there is suspicion of a primary bone or soft-tissue malignancy at any stage of a patient's investigation, medical practitioners should not hesitate to seek advice from an appropriate centre. Seeking a second opinion from a competent authority will never compromise patient outcome. However, operating on a patient with an uncertain diagnosis could have catastrophic consequences and could even compromise that patient's only chance of survival.

**Case reports**

**Patient 1.** A 29-year-old woman with a three-month history of pain and swelling of the left knee accompanied by a progressive cough. Radiographs of the femur showed a lytic lesion suspected to be a primary bone sarcoma (Fig. 3). An MRI of the femur (Fig. 4) and chest radiograph (Fig. 5) were obtained. These
revealed a large mass in the right lung confirmed by a CT of the chest (Fig. 6), which showed a 6 cm mass in the right upper lobe consistent with a metastasis. Isotope bone scan showed a lesion of the L4 vertebra consistent with a bony metastasis. Biopsy of the femur confirmed a highly undifferentiated large cell lung carcinoma. The patient was referred for palliative treatment to a local centre.

Message: Always undertake a systemic investigation in a patient with suspected malignancy.

**Patient 2.** A 12-year-old boy sustained a fracture of his left distal femur as a result of a trivial injury. He had suffered aching in the leg for the previous two weeks. Despite the unimpressive nature of the injury, the radiograph showed no worrying features (Fig. 7) and the patient underwent fixation with a plate. Unfortunately, three months later the fracture demonstrated a delayed union with increasing bone loss. There was also concern about infection although the inflammatory markers were normal (Fig. 8) and the possibility of malignancy was raised. An MRI showed a large soft-tissue mass at the site of the fracture (Fig. 9) and biopsy confirmed a high-grade telangiectatic osteosarcoma. Isotope bone scan showed this to be solitary (Fig. 10), and a CT of the chest showed no evidence of disseminated disease. The patient had a modest response to chemotherapy and required an amputation.
The lack of significant trauma in this young and otherwise healthy adolescent should have raised sufficient suspicion to obtain second imaging (MRI) before surgery. Earlier diagnosis would have allowed proper systemic treatment to commence three to four months earlier and may have allowed limb salvage surgery.

**Patient 3.** A 44-year-old man had a 2½ year history of progressive left-sided sciatica and pelvic pain, with progressive neuropaxia of the left sciatic territory. The patient was seen by multiple clinicians, including a number of orthopaedic surgeons. He was investigated with a spinal MRI, which was normal, and referred for physiotherapy which did not help. When the patient was finally referred to our unit he presented with pressure sores over his elbows and knees because he could not lie on his back or side; he had been sleeping and washing in a kneeling position. Radiographs showed a destructive lesion of the left ilium with a large soft-tissue mass extending past the midline of the pubis (Fig. 11). MRI further demonstrated a large mass involving the ilium and pushing the entire pelvic cavity across the midline (Fig. 12). Staging studies showed a solitary lesion on the isotope bone scan (Fig. 12) but CT of the chest showed multiple small lesions which were suggestive of metastases. Core-needle biopsy confirmed a Grade II chondrosarcoma of the pelvis. The patient underwent a palliative hindquarter amputation (Fig. 13) in an attempt to obtain local control and reduce the unremitting pain.

Message: Severe pain should be investigated. Imaging of the pelvis in a patient with sciatica and a normal spinal MRI should always be considered. Palliative hindquarter amputation can improve the quality and duration of life, even in patients with metastases.

**Patient 4.** A 34-year-old dental nurse presented with a one-year history of a mass in the posterior aspect of her distal thigh. The lump was not painful but was increasing slowly in size. She was reassured several times by her General Practitioner (GP). When she developed varicose veins on the surface of the mass this led to her request for a second opinion; by this stage the mass had become painful. A radiograph (Fig. 14) showed a soft-tissue mass in the posteromedial thigh and normal bone structures. The mass presented multiple worrying features such as being larger than 5 cm, being deep to the fascia, as well as enlarging and being associated with a recent onset of pain. MRI (Fig. 15) demonstrated a 15 cm mass in the posterior aspect of the left thigh extending into the popliteal fossa, displacing the vessels and sciatic nerve. Trucut biopsy revealed a high-grade soft-tissue sarcoma. The mass was excised with a planned margin.
adjacent to the vessels and sciatic nerve and the patient was treated with adjuvant radiotherapy.

**Message:** Any soft-tissue mass with worrying features should always be investigated by MRI.

**Patient 5.** A 19-year-old student presented with a two-year history of a painless lump in the back of his left calf. He did not seek medical advice because the mass was increasing very slowly in size and was only troubling him when he was crouched. Ultrasound showed a 10 cm solid mass confirmed by MRI (Fig. 16). This mass presented worrying features, as it was larger than 5 cm, deep to the fascia and was slowly enlarging. Imaging also showed an obvious skip lesion proximal to the main mass and adjacent to the vessels. When seen in the outpatient clinic he complained of tw0-week history of backache that sometimes woke him at night. He had been prescribed anti-inflammatory medication by his GP. The patient later complained of paraesthesiae in both legs, although the GP had attributed this to an allergic reaction to the medication. Radio-
graphs of the spine at our unit clearly demonstrated a superior, posterior mediastinal mass over the right upper lobe of the lung (Fig. 17). Urgent MRI confirmed this was because of an extensive thoracic mass which had invaded the spinal canal (Fig. 18). Trucut biopsy of the left calf revealed an extremely rare soft-tissue tumour, a sclerosing rhabdomyosarcoma. The patient underwent an urgent spinal decompression followed by chemotherapy.

Message: Any enlarging soft-tissue mass should be promptly investigated. Back pain that wakes a patient at night is a significant symptom, which requires urgent investigation.

Robert J Grimer FRCS, Consultant Orthopaedic Surgeon
Sophie Mottard FRCS, Consultant Orthopaedic Surgeon
Timothy R Briggs FRCS, Professor of Orthopaedics**

The Royal Orthopaedic Hospital, Birmingham, UK;
**The Royal National Orthopaedic Hospital, Stanmore, UK

Correspondence should be addressed to: Mr RJ Grimer, FRCS, Royal Orthopaedic Hospital, Bristol Road South, Birmingham B31 2AP, Tel: (+44) (0)121 685 4037, Fax: (+44) (0)121 685 4146, E-mail: rob.grimer@roh.nhs.uk
References


