42 Musculoskeletal tumours

BONE TUMOURS

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(b) Figure 42.4 (a, b) Sclerotic osteosarcoma of the distal femur in a child (arrows).

Figure 42.5 (a) Chondrosarcoma of the proximal humerus with multiple calci /f_i cations. involvement. (c) Excised chondrosarcoma of the proximal humerus. (a) (c) Figure 42.6 (a) Chondrosarcoma of the foot. (b) Computed tomography scan reconstruction showing multiple calci /f_i cations. magnetic resonance imaging scan shows high signal in the chondrosarcoma. (b) Magnetic resonance imaging scan showing extensive (b) (d) (c) T2-weighted (d) Excised chondrosarcoma of the foot.

Figure 42.7 Pathological fracture through a primary chondrosarcoma of the proximal humerus. TABLE 42.1 Conditions associated with an increased risk of malignant disease in bone and cartilage. High risk Moderate risk Low risk Hereditary multiple Chronic Maffucci syndrome exostoses osteomyelitis (enchondromatosis and angiomas of soft tissue) Ollier’s disease Polyostotic Paget’s Osteonecrosis (enchondromatosis) disease Radiation osteitis Fibrous dysplasia, Familial osteogenesis retinoblastoma imperfecta, syndrome osteoblastoma and chondroblastoma

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Biopsy

Biopsy

A biopsy is performed only when local staging investigations have been completed. Because removal of the biopsy track is an important principle in the treatment of sarcomas, and specialist pathology is required, biopsies should be performed either in, or after consultation with, the specialist centre where the definitive surgical procedure will be performed. have a higher diagnostic accuracy because areas of radiological concern can be targeted. If image-guided biopsy is performed, close discussion between radiologist and surgeon is required to ensure an appropriate biopsy route is used (Figures 42.26 and 42.27). Summary box 42.10 Biopsy /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Biopsies for bone tumours are usually taken using a Jamshidi or other hollow needle (Figure 42.28), while Trucut needles are preferred for soft-tissue tumours. Although most biopsies are performed with a needle, some times an open biopsy is required, which should be performed according to the following principles. /uni25CF A tourniquet can be used; but exsanguination by compres sion should be avoided as this may disseminate the tumour. En bloc is French for ‘in a block’. Khosrow Jamshidi , contemporary , Iranian haematologist. /uni25CF Use longitudinal incisions that are part of an extensile approach. /uni25CF Do not cross anatomical compartments or contaminate critical anatomical structures (e.g. nerves or blood vessels). /uni25CF Use a biopsy track that can be excised at the time of definitive surgery . /uni25CF Ensure specimens are sent for microbiology as well as histopathology . /uni25CF Some specimens should be sent fresh to the laboratory for genetic studies.

Figure 42.26 Poorly placed biopsies can make subsequent surgical excision of the track dif /f_i cult or not possible. Figure 42.27 En bloc excised tumour and biopsy track. Only biopsy once local staging is completed Biopsy should be performed at, or after discussion with, the specialist centre Image-guided biopsy is more reliable The biopsy track should be excised at de /f_i nitive surgery Jamshidi needles for bone; Trucut needles for soft tissues Figure 42.28 Bone biopsy instruments.

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Chondrogenic tumours

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These tumours produce chondroid matrix and include a wide range of benign and malignant tumours. Osteochondroma (Figures 42.14 and 42.15) is a benign cartilage-capped bony projection, thought to originate - - from the physis. The bony projection always grows away from the joint towards the diaphyseal region of the bone. It has no structures attached to it. Osteochondromas can be pedunculated (with a stalk) or sessile (without a stalk). The stalk or base is always continuous with the intramedullary cavity of the bone, and the continuity of the cortex of the bone into an osteochondroma is a characteristic radiological feature. They are usually solitary , but some patients have multiple osteochondromas (hereditary multiple exostoses, autosomal dominant inheritance) (Figure 42.16). Osteochondromas can cause local irritation and complications include mechanical symptoms, nerve impingement, vascular pseudoaneurysm, fracture and infarction. Increasing size or pain, particularly after skeletal maturity , is concerning and may indicate malignant transformation. The incidence of malignant transformation is less than 1% in solitary osteochondromas and 1–3% in patients with multiple osteochondromas.

Figure 42.13 Parosteal osteosarcoma of the distal femur in an unusu ally young patient. There is no continuity between the tumour and the intramedullary cavity of the femur (arrow). Figure 42.14 Pedunculated osteochondromas (arrow) of the

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Enchondroma (Figure 42.17) is a benign cartilaginous neoplasm within the intramedullary cavity of bone. Approximately 50% are in the hands and feet: enchondromas are the most common bone tumours in the hand. Although they can present with pain, swelling or pathological fracture, many are entirely asymptomatic and are detected incidentally . Patchy calcification, expansion and scalloping can be visible on radiographs, but some are only diagnosed on magnetic resonance imaging (MRI) scan. Ollier’s disease is a developmental condition characterised by multiple enchondromas. In Ma ff ucci syndrome, multiple enchondromas are associated with multiple angiomas. Malig tion to chondrosarcoma can occur in approx nant transforma imately 20% of patients with Ollier’s disease and is almost inevitable in patients with Ma ff ucci syndrome. Chondroblastoma (Figure 42.18) is a benign cartilage- producing tumour that occurs in the epiphyses of bones in en. It is most common around the knee. Pain is often childr tion and possibly joint e ff usion. severe, with associated inflamma On plain radiographs, there is an often barely visible lytic lesion in the centre of the epiphysis. Previously , the diagnosis was often missed, but this has become less frequent with MRI scanning, which usually identifies the lesion with an intense inflammatory response. Chondrosarcoma (Figures 42.5, 42.6 and 42.7) is a malig nant tumour with cartilage di ff erentiation. The biological Summary box 42.5 Tumours producing cartilage /uni25CF /uni25CF /uni25CF /uni25CF behaviour ranges from very low-grade lesions to highly aggressive dedi ff erentiated tumours. Patients usually present with pain and/or swelling and symptoms may be longstand - ing. Many chondrosarcomas arise in pre-existing lesions such as

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EVALUATION AND INVESTIGATION OF THE PATIENT WITH A SUSPECTED BONE OR SOFT-TISSUE TUMOUR

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The diagnosis and treatment of patients with primary bone and/or soft-tissue tumours requires a high index of suspicion, appropriate and prompt investigation, and early referral to a specialist multidisciplinary team for diagnosis, biopsy and appropriate treatment. When a musculoskeletal tumour is suspected, clinicians should: /uni25CF stop; /uni25CF think; /uni25CF investigate. bone or soft-tissue tumour can be divided into three phases. The first two phases can be performed at the referring hospi tal, but the third phase may be best done in a specialist centre (Table 42.5). /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF

TABLE 42.5 The three phases of assessment of lesions. Phase 1 Phase 2 Phase 3 (within 24 hours) (within /f_i rst week) (at specialist centre) History and Bone scan CT scan lesion examination Ultrasound scan MRI scan lesion abdomen Blood tests Biopsy Radiograph CT scan chest whole bone Chest radiograph CT, computed tomography; MRI, magnetic resonance imaging.

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Haematopoietic tumours

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History and examination

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It is important to take a thorough history , including a pain history . Non-mechanical and/or night pain, particularly in the young adolescent, is a concerning symptom and a primary bone tumour should be suspected. Relief with non-steroidal anti-inflammatory drugs may suggest an osteoid osteoma. Patients with a history of malignancy who present with back pain should be considered to have metastatic bone dis ease until proven otherwise. Plain radiographs of the spine and routine blood tests are the minimum that is required. An MRI of the spine is a more sensitive test for the detection of a malig nant tumour and may demonstrate soft-tissue extension into the spinal canal. Multiple my eloma (Figure 42.3) is the most common primary malignancy of bone in adults and should be considered in all patients over 65 years of age with back pain. Back pain associated with an ESR >100 /uni00A0 mm/h indicates m ultiple myeloma until proven otherwise. Monoclonal gam mopathy or elevated urinary and serum Bence Jones proteins are diagnostic. All patients with suspected cancer in the spine should be examined for signs of spinal cord compression, a potential surgical emergency . Great care should be taken when managing a patient with an apparently ‘solitary’ bone metastasis. This could be a pri mary bone tumour, and further investigation including biopsy is required. Soft-tissue tumours are common and the vast majority are benign. Ho wever, a soft-tissue mass meeting any of the fol lowing criteria may be malignant and the pa tient should be referred to a specialist centre: /uni25CF painful; /uni25CF increasing in size; /uni25CF more than 5 /uni00A0 cm in diameter. In addition, tumours that have recurred after previous excision and tumours located deep to the fascia are more likely to be malignant. It is important to note that tumours that appear mostly superficial but involve the deep fascia are classified as deep tumour s. Henry Bence Jones , 1813–1873, physician, St George’s Hospital, London, UK. The investigation of a patient with a suspected primary bone - or soft-tissue tumour should include the following. /uni25CF Local investigations: /uni25CF ultrasound scan (for soft-tissue tumours); /uni25CF plain radiographs of the whole a ff ected bone or soft- tissue lesion (Figure 42.1); /uni25CF MRI of the whole a ff ected bone or soft-tissue mass; /uni25CF computed tomography (CT) scan may be helpful in ad - dition to an MRI scan. /uni25CF Distant: /uni25CF blood tests, including full blood count, ESR, urea and electrolytes, bone profile and protein electrophoresis; /uni25CF plain radiographs or CT scan of the chest (more sen - sitive); /uni25CF whole-body isotope bone scan (for suspected primary or metastatic bone tumours); /uni25CF ultrasound or CT scan of the abdomen (if renal metas - tasis is a possibility). Plain radiographs are usually the most useful imaging investigations in determining the diagnosis of a primary bone tumour, but further appropriate scans are usually required for confirmation and staging . Imaging should always include the whole of the a ff ected bone to look for satellite lesions and skip metastases. Satellite lesions occur within, whereas skip lesions - occur beyond, the reactive zone of the tumour, which is the layer of compressed tissues, inflammatory cells and tumour infiltration that surrounds the tumour. - Both primary bone and soft-tissue sarcomas metastasise to the lungs, and a CT scan of the chest is an essential part of staging. Patients who present with a lytic bone lesion could have a primary renal carcinoma and an ultrasound or CT scan of the abdomen is advised. Surgery to a

renal metastasis can lead to - significant b lood loss. Summary box 42.9 Staging - /uni25CF /uni25CF /uni25CF /uni25CF -

Plain radiography is most informative for bone tumours Always image the whole bone in the case of skip lesions CT of the lung detects lung metastases Lytic lesions require imaging of the abdomen to check for a primary renal carcinoma

History and examination

It is important to take a thorough history , including a pain history . Non-mechanical and/or night pain, particularly in the young adolescent, is a concerning symptom and a primary bone tumour should be suspected. Relief with non-steroidal anti-inflammatory drugs may suggest an osteoid osteoma. Patients with a history of malignancy who present with back pain should be considered to have metastatic bone dis ease until proven otherwise. Plain radiographs of the spine and routine blood tests are the minimum that is required. An MRI of the spine is a more sensitive test for the detection of a malig nant tumour and may demonstrate soft-tissue extension into the spinal canal. Multiple my eloma (Figure 42.3) is the most common primary malignancy of bone in adults and should be considered in all patients over 65 years of age with back pain. Back pain associated with an ESR >100 /uni00A0 mm/h indicates m ultiple myeloma until proven otherwise. Monoclonal gam mopathy or elevated urinary and serum Bence Jones proteins are diagnostic. All patients with suspected cancer in the spine should be examined for signs of spinal cord compression, a potential surgical emergency . Great care should be taken when managing a patient with an apparently ‘solitary’ bone metastasis. This could be a pri mary bone tumour, and further investigation including biopsy is required. Soft-tissue tumours are common and the vast majority are benign. Ho wever, a soft-tissue mass meeting any of the fol lowing criteria may be malignant and the pa tient should be referred to a specialist centre: /uni25CF painful; /uni25CF increasing in size; /uni25CF more than 5 /uni00A0 cm in diameter. In addition, tumours that have recurred after previous excision and tumours located deep to the fascia are more likely to be malignant. It is important to note that tumours that appear mostly superficial but involve the deep fascia are classified as deep tumour s. Henry Bence Jones , 1813–1873, physician, St George’s Hospital, London, UK. The investigation of a patient with a suspected primary bone - or soft-tissue tumour should include the following. /uni25CF Local investigations: /uni25CF ultrasound scan (for soft-tissue tumours); /uni25CF plain radiographs of the whole a ff ected bone or soft- tissue lesion (Figure 42.1); /uni25CF MRI of the whole a ff ected bone or soft-tissue mass; /uni25CF computed tomography (CT) scan may be helpful in ad - dition to an MRI scan. /uni25CF Distant: /uni25CF blood tests, including full blood count, ESR, urea and electrolytes, bone profile and protein electrophoresis; /uni25CF plain radiographs or CT scan of the chest (more sen - sitive); /uni25CF whole-body isotope bone scan (for suspected primary or metastatic bone tumours); /uni25CF ultrasound or CT scan of the abdomen (if renal metas - tasis is a possibility). Plain radiographs are usually the most useful imaging investigations in determining the diagnosis of a primary bone tumour, but further appropriate scans are usually required for confirmation and staging . Imaging should always include the whole of the a ff ected bone to look for satellite lesions and skip metastases. Satellite lesions occur within, whereas skip lesions - occur beyond, the reactive zone of the tumour, which is the layer of compressed tissues, inflammatory cells and tumour infiltration that surrounds the tumour. - Both primary bone and soft-tissue sarcomas metastasise to the lungs, and a CT scan of the chest is an essential part of staging. Patients who present with a lytic bone lesion could have a

primary renal carcinoma and an ultrasound or CT scan of the abdomen is advised. Surgery to a renal metastasis can lead to - significant b lood loss. Summary box 42.9 Staging - /uni25CF /uni25CF /uni25CF /uni25CF -

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Introduction

INTRODUCTION

Musculoskeletal tumours include primary and secondary benign and malignant tumours of bone and soft tissue. The most common malignant tumours in bone are metastatic carci nomas (Figure 42.1). Advances in oncological treatment mean that the number of patients living with metastatic bone disease is increasing. The most common carcinomas that metastasise to bone originate in the breast, prostate, lung, kidney , thyroid and colon (Figure 42.2). Haematopoietic tumours may also arise in bone: multiple myeloma (Figure 42.3) is a malignant neoplasm arising from - plasma cells in the bone marrow , leading to multiple lesions in the skeleton. When solitary , this type of tumour is called a plasmacytoma.

(a) (b) Figure 42.1 (a) Pathological fracture of the proximal femur through metastatic breast carcinoma. (b) Radiographs of the whole femur show a further, more distal metastatic deposit. Understand the principles of biopsy • Describe the principles of surgical treatment of • musculoskeletal tumours List the aims of surgical treatment for metastatic bone • disease Understand how to manage patients with an impending or • completed pathological fracture Evaluate the risk of pathological fracture • Thyroid Lung Breast Kidney Colon Myeloma Prostate Figure 42.2 Common malignant tumours involving bone (courtesy of Mr Andy Biggs, The Robert Jones and Agnes Hunt Orthopaedic Hospital NHS Foundation Trust).

Malignant primary bone tumours (sarcomas) are very rare, but notably can occur in children and young adults. The most common malignant primary bone tumours are osteosarcoma (Figure 42.4), chondrosarcoma (Figures 42.5, 42.6 and 42.7 and Ewing’s sarcoma (Figure 42.8). Soft-tissue tumours are common. However, only one in a 100 is malignant (Figure 42.9).

(b) Figure 42.3 (a) Multiple myeloma affecting the left humerus with a pathological fracture (arrows). (b) Multiple myeloma with multiple deposits in the skull (arrows).

Learning objectives

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List the symptoms and signs associated with a • musculoskeletal tumour Understand why a patient with a suspected • musculoskeletal tumour should be referred to a specialist centre for staging, biopsy and multidisciplinary management Understand why staging should be completed before • biopsy Explain why a diagnosis is required before treatment • Learning objectives

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Metastatic bone disease

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Most tumours that metastasise to bone are carcinomas. Some times, despite further investigations, the primary tumour is never found: these patients are described as having ‘carcinoma of unknown primary’. However, with advanced diagnostics and laborator y investigations the origin of most bone meta stases can be identified. Carcinomas usually spread to bone by the haematogenous route: the spine is the third most common site for metastases, after the lung and liver. Although most patients with metastatic cancer will have bone metastases in the spine before they die, tic. only 10% are symptoma Tumour cells metastasise to the spine via Batson’s venous plexus. These retroperitoneal veins have no valves and allow retrograde embolic spread to the spine and proximal long Figure 42.10). bones (Bone metastases can be lytic, sclerotic or mixed. Lytic metastases are usually highly vascular or locally aggressive such Angelo Ma ff ucci , 1847–1903, Professor of Pathological Anatomy , Pisa, Italy , described enchondromatosis in association with soft tissue haemangiomas in 1881. Louis Xavier Edouard Léopold Ollier , 1830–1900, Professor of Surgery , Lyons, France, described enchondromatosis in 1899. Oscar V Batson , 1894–1979, American otolaryngologist. - - that there is no healing response from the bone. Metastases from prostate cancer may appear sclerotic. Metastases are rare in children, but bone metastases can occur from neuroblastoma, rhabdomyosarcoma and clear cell carcinoma of the kidney .

Figure 42.8 Ewing’s sarcoma of the proximal /f_i bula. The tumour is metadiaphyseal in location with a periosteal reaction and subtle onion-skinning. (a) (b) Figure 42.9 (a) Large fungating soft-tissue sarcoma of the buttock. /uni00A0 (b) Magnetic resonance imaging scan from the same patient showing a large fungating sarcoma of the buttock.

Summary box 42.1 Most common tumours metastasising to bone (93%) /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Summary box 42.2 Most common sites of bone metastases /uni25CF /uni25CF /uni25CF

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Metastatic bone disease

Patients with confirmed metastatic bone disease may require resuscitation for electrolyte imbalance, anaemia, cardiorespi - ratory problems or hypercalcaemia before surgical treatment can be considered. Hypercalcaemia can be treated e ff ectively with fluid resuscitation and bisphosphonate infusion. Surgical treatment in patients with metastatic bone disease is usually palliative; although complete resection of solitary metastases in selected patients may confer some survival bene - fit, the evidence for this is not strong. Surgery of the spine may be requir ed for

stabilisation and/ or decompression when tumour extension puts the spinal cord at risk. Surgery in the peripheral skeleton is mainly for treat - ment of pain and (impending) pathological fracture. Renal metastases tend to be very v ascular and massive blood loss can be encountered during surgery . Therefore, pre - operative embolisation should be consider ed just before sur - gery to prevent blood loss (Figure 42.30). Treatment of myeloma is mainly haematological. Non - surgical treatments including radiotherapy can lead to healing of bone lesions in some cases. Surgical treatment is often required for complications such as fracture and spinal cord compression. The following factors should be considered when contem plating surgical treatment for patients with metastatic bone disease: /uni25CF likely survival (Figure 42.31) – consider the primary diag nosis and performance status; /uni25CF quality of life; /uni25CF fitness for anaesthesia and surgery; /uni25CF fracture risk; /uni25CF single or multiple bone lesions; /uni25CF response to adjuvant treatment such as radiotherapy and hormonal treatment; /uni25CF radiotherapy can be administered pre- or postoperatively . The risk of pathological fracture can be assessed using the Mirels score (Table 42.7). However, this scoring system is prone to inter- and intraobserver variation. Hilton Mirels , contemporary , South African orthopaedic surgeon who now practises in the USA. - - Bone healing following a fracture through a metastasis is unpredictable and there can be local recurrence of the tumour after treatment. The approach is therefore di ff erent from the treatment of other fractures. The aim of surgery should be to improve pain and maintain mobility . Approaches that require prolonged protected weight-bearing to allow healing are not appropriate in this group of patients with reduced life expec - tancy . Therefore, as a general rule, prosthetic replacement of bones is preferred, particularly for epiphyseal and metaphyseal lesions. Metastases in the diaphysis may be most appropriately osthetic treated with an intramedullary nail. In the shoulder, pr replacements have a poor function and internal fixation may give better physical functioning. However, for hip lesions, the best treatment is often replacement surgery . Patients with solitary breast and renal metastases can have prolonged disease-free survival, so excision and replacement rather than fixation should be considered.

Figure 42.30 (a) Lytic metastasis of renal cell carcinoma. (b) Angiogram shows increased vascularity. 1.0 Breast Lung 0.8 Myeloma 0.6 Other Prostate 0.4 Renal Cum. survival Thyroid 0.2 0 0 10 5 Time (years) Figure 42.31 Cumulative survival curves of patients who present with bony metastasis. (c) Following embolisation. TABLE 42.7 The Mirels scoring system for risk of pathological fracture. Score 1 2 3 Site Upper limb Lower limb Peritrochanteric Pain Mild Moderate Functional Size <1/3 1/3–2/3

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Osteogenic tumours

Osteogenic tumours

These tumours characteristically produce osteoid or bony matrix, which may be seen on imaging studies or on histo logical examination. Osteoid osteoma (Figures 42.11 and 42.12) is a benign bone-forming lesion that is small but very painful. Usually , pain occurs at night and is typically relieved by non-steroidal anti-inflammatory medica tion. Osteoid osteomas usually occur in children and adolescents. They can arise in any bone, particularly the proximal femur, and cause a dense cortical reaction in the centre of which is a nidus (Figure 42.12 - -).

Figure 42.11 Radiograph showing an osteoid osteoma of the tibial diaphysis with reactive bone formation (arrow). (a) (b) Figure 42.12 (a) Axial computed tomography (CT) scan showing an osteoid osteoma nidus in the distal tibia (arrow). (b) CT-guided radio- frequency thermocoagulation of an osteoid osteoma of the distal tibia. The scan shows the electrode in situ (arrow).

Malignant bone tumours /uni25CF /uni25CF /uni25CF /uni25CF Osteoid osteomas can cause irritation and e ff usions if they occur close to a joint. Osteoblastoma is the larger (>2 /uni00A0 cm), more aggressive coun terpart of osteoid osteoma and more typically occurs in the spine. Osteosarcoma (Figure 42.4) is a malignant bone-forming tumour, most common in the distal femur, followed by the proximal tibia, pro ximal humerus and distal tibia. The radio logical and histological classification of osteosarcomas includes sclerotic (Figure 42.4), chondroblastic, telangiectatic and other more unusual forms. Usually , osteosarcomas are intraosseous, b ut they can also arise from the surface of bones. Parosteal osteosarcoma (Figure 42.13) is a low-grade osteosarcoma that arises from the surface of the bone, typically of the distal femur or proximal tibia. Symptoms are often mild and longstanding. Summary box 42.4 Tumours producing bone /uni25CF /uni25CF /uni25CF

Plasmacytoma – solitary form of multiple myeloma Osteosarcoma – usually secondary to Paget’s disease and radiotherapy in older patients Chondrosarcoma Ewing’s sarcoma Osteoid osteoma – small, painful; produce dense cortical reaction Osteoblastoma – larger and more aggressive than osteoid osteoma Osteosarcoma – malignant; commonest in lower femur and upper tibia

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Malignant bone tumours /uni25CF /uni25CF /uni25CF /uni25CF Osteoid osteomas can cause irritation and e ff usions if they occur close to a joint. Osteoblastoma is the larger (>2 /uni00A0 cm), more aggressive coun terpart of osteoid osteoma and more typically occurs in the spine. Osteosarcoma (Figure 42.4) is a malignant bone-forming tumour, most common in the distal femur, followed by the proximal tibia, pro ximal humerus and distal tibia. The radio logical and histological classification of osteosarcomas includes sclerotic (Figure 42.4), chondroblastic, telangiectatic and other more unusual forms. Usually , osteosarcomas are intraosseous, b ut they can also arise from the surface of bones. Parosteal osteosarcoma (Figure 42.13) is a low-grade osteosarcoma that arises from the surface of the bone, typically of the distal femur or proximal tibia. Symptoms are often mild and longstanding. Summary box 42.4 Tumours producing bone

/uni25CF /uni25CF /uni25CF

Plasmacytoma – solitary form of multiple myeloma Osteosarcoma – usually secondary to Paget’s disease and radiotherapy in older patients Chondrosarcoma Ewing’s sarcoma Osteoid osteoma – small, painful; produce dense cortical reaction Osteoblastoma – larger and more aggressive than osteoid osteoma Osteosarcoma – malignant; commonest in lower femur and upper tibia

Others

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Simple (unicameral) bone cyst (Figure 42.20) is a membrane-lined cavity filled with serous fluid within a bone. It usually occurs in the proximal long bones of children. Asso - ciated thinning of the cortex of the bone can lead to fracture. Such fractures usually heal with conservative treatment, but the cyst may only partially resolve . Aneurysmal bone cyst (Figure 42.21) is a benign cystic lesion of bone consisting of blood-filled spaces separated - by fibrous septa. The lesion is more aggressive than a simple bone cyst and often presents with pain and swelling. Plain radiographs commonly show aggressive features with eccentric expansion of the cortex and an open physis. Scans often show multiple fluid levels (Figure 42.21b). Giant cell tumour of bone (Figure 42.22) is a locally aggressive tumour with large osteoclast-like giant cells. It usually occurs between the ages of 20 and 45, after the physes have closed. Giant cell tumour of bone typically extends into the epiphysis of long bones and erodes bone under the articular cartilage, especially around the knee, proximal humerus and distal radius. ‘Benign’ metastases are rare.

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Figure 42.19 (a) Clear cell chondrosarcoma of the medial femoral condyle (arrow). (b) Sagittal T1-weighted magnetic resonance imag

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Eosinophilic granuloma is a rare neoplasm of Lang erhans cells (Figure 42.23). It can be unifocal (eosinophilic granuloma), multifocal (Hand–Schüller–Christian disease) or Paul Langerhans , 1847–1888, Professor of Pathological Anatomy , Freiberg, Germany . Alfred Hand Jr , 1868–1949, American pediatrician, described the eponymous disease in 1893. Artur Schüller , 1874–1957, Austrian neuroradiologist, described the eponymous disease in 1915. Henry A Christian , 1876–1951, American physician, described the eponymous disease in 1919. Erich Letterer , 1895–1982, German pathologist. Sture A Siwe , 1897–1966, Swedish paediatrician. for the skull and the diaphyses of long bones. In the spine it can present with collapse, kno wn as vertebra plana.

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PRINCIPLES OF TREATMENT Primary bone tumours

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SOFT-TISSUE TUMOURS

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Soft-tissue tumours have also historically been classified according to their morphological appearance and presumed cell of origin. The range of biological behaviour is wide and most morphological types have a benign and malignant coun terpart, for example lipoma (Figure 42.25) and liposarcoma. Other more frequent types include undi ff erentiated pleomor phic sarcoma and synovial sarcoma. Patients with suspected or confirmed soft-tissue sarcomas should be assessed and managed in a specialist centre. Summary box 42.8 Warning signs – soft-tissue tumour /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Monique Trojani , contemporary , described the histopathological grading system in 1984. - The Trojani system, based on tumour di ff erentiation, mitotic count and tumour necrosis, is the standard for grading malignant soft tissue tumours. The AJCC / UICC system is used to stage malignant soft tissue tumours. - -

Larger than 5 /uni00A0 cm Increasing in size Painful Deep to the fascia Recurrence after previous excision (b) Figure 42.25 (a) Coronal T1-weighted magnetic resonance imaging scan showing a benign lipoma deep to the quadriceps muscle (arrow). (b) Excised benign lipoma.

Soft-tissue tumours

The treatment of soft-tissue tumours should take account of tumour type and the response to other treatments including radiotherapy . Large low-grade or benign lipomatous tumours may be excised in a deliberately marginal or close but complete fashion. Soft-tissue sarcomas should however be excised with a margin of normal tissue around them, which includes the biopsy track, wherever possible (Figure 42.27). Skin involve ment may require resection of the skin and reconstruction with a split-skin graft or skin flap. Following surgical excision of high-grade soft-tissue sarco mas, adjuvant radiotherapy should be considered. Preopera radiotherapy can also have good results, but there is a risk of wound-healing problems following surgery . Chemotherap a limited role in the treatment of soft-tissue sarcomas. British Orthopaedic Oncology Society and British Orthopaedic Association. Metastatic bone disease. A guide to good practice . Oxford: BOOS; London: BOA, 2015. Cool P , Grimer R. Pathological fractures of the extremities. Trauma 2000; 2 : 101–11. Cool P , Grimer R, Rees R. Surveillance in patients with sarcoma of the extremities. Eur J Surg Oncol 2005; 31 (9): 1020–4. Dangoor A, Seddon B, Gerrand C et al . UK guidelines for the management of soft tissue sarcomas. Clin Sarcoma Res 2016; 6 : 20. Enneking WF , Spanier SS, Goodman MA. A system for the surgical staging of musculoskeletal sarcoma. Clin Orthop Relat Res 1980; 153 : 106–20. Gerrand C, Athanasou N, Brennan B et al . on behalf of the British Sarcoma Group. UK guidelines for the management of bone sarcomas. Clin Sarcoma Res 2016; 6 : 7. Mankin HJ, Lange TA, Spanier SS. The hazards of biopsy in patients with malignant primary bone and soft tissue tumors. J Bone Joint Surg 1982; 64-A : 1121–7. Mirels H. Metastatic disease in long bones. Clin Orthop 1989; 249 : 256–64. Wedin R, Bauer HC. Surgical treatment of skeletal metastatic lesions of the proximal femur: endoprosthesis or reconstruction nail? J Bone Joint Surg Br 2005; 87 (12): 1653–7. - World

Health Organization, International Agency for Research on Cancer. WHO classification of tumours. V ol. 3. Soft tissue and bone tumours , 5th edn. Lyon: IARC Press, 2019. - Union for International Cancer Control. TNM classification of malignant tumours , 8th edn. Oxford/Hoboken, NJ: John Wiley & Sons, 2017. tive y has SOFT-TISSUE TUMOURS

Soft-tissue tumours have also historically been classified according to their morphological appearance and presumed cell of origin. The range of biological behaviour is wide and most morphological types have a benign and malignant coun terpart, for example lipoma (Figure 42.25) and liposarcoma. Other more frequent types include undi ff erentiated pleomor phic sarcoma and synovial sarcoma. Patients with suspected or confirmed soft-tissue sarcomas should be assessed and managed in a specialist centre. Summary box 42.8 Warning signs – soft-tissue tumour /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Monique Trojani , contemporary , described the histopathological grading system in 1984. - The Trojani system, based on tumour di ff erentiation, mitotic count and tumour necrosis, is the standard for grading malignant soft tissue tumours. The AJCC / UICC system is used to stage malignant soft tissue tumours. - -

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covered incidentally . Active lesions, such as osteoid osteoma, present with mild symptoms and continue to grow . Aggressive lesions tend to grow rapidly and destroy bone. The Enneking staging system f or malignant tumours combines the local extent of the tumour and the histological grade (Table 42.4). The compartment is the bone in which the tumour arises. A tumour is extracompartmental when it has breached the corte x of the bone. Most primary malignant bone tumours are Enneking stage 2B at diagnosis, meaning they have extended outside the bone of origin but metastases are not detectable. The American Joint Committee on Cancer (AJCC)/Union for International Cancer Control (UICC) stag ing system is also widely used. Summary box 42.7 Warning signs – bone tumour /uni25CF /uni25CF /uni25CF

TABLE 42.4 The Enneking staging system for bone tumours. Low grade Intracompartmental 1A Extracompartmental 1B High grade Intracompartmental 2A Extracompartmental 2B Any grade Metastases 3 Non-mechanical bone pain Especially around the knee in young adolescents Concerning radiographs

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