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Bone and Soft Tissue Sarcomas (with comments on retroperitoneum and adrenal gland)

1. GROSS DESCRIPTION

Specimen

— fine needle aspirate/needle core biopsy/open biopsy (incisional/curettings)/enucleation/wide local excision/compartmentectomy/segmental resection/en-bloc resection/amputation (limb (below/above knee, etc))/complex resection (forequarter/hindquarter/hemipelvectomy).
— right or left.
— size (cm) and weight (g).
— bone tumours often present as severe continuous pain unrelieved by anti-inflammatory agents, swelling or sometimes as a pathological fracture following low-impact trauma. Investigation is by plain X-ray and CT scan looking particularly for signs of periosteal reaction. A tissue diagnosis is obtained by needle core biopsy under radiological control. Osteosarcoma and Ewing's sarcoma are treated by a combination of chemotherapy and surgery, chondrosarcoma by surgery. Most primary bone tumours arise de novo but a minority occur in association with recognizable precursors, e.g. Paget's disease or a history of radiation. Metastatic bone disease can cause hypercalcaemia and is detected by isotope bone scan. Benign soft tissue tumours far outnumber malignant cases. Soft tissue sarcomas occur mainly in the extremities (often thigh) but also the retroperitoneum and trunk wall. They are usually deep seated and progressively increase in size, causing a lump or swelling and sometimes pain with a loss of function in the limb or adjacent organs. Plain X-ray may show focal calcification (e.g. synovial sarcoma) but MRI is the investigation of choice in defining the nature of the mass, its extent and involvement of adjacent structures. CT scan is used for lesions of the trunk. After full clinical and radiological assessment most centres use needle core biopsy to obtain a tissue diagnosis and open biopsy only when needle core biopsy or FNA have proven inconclusive. Surgery is the mainstay of treatment with mainly chemotherapy for Ewing's sarcoma and rhabdomyosarcoma.

Tumour

Site
— osseous: paracortical (paraosteal/periosteal); cortical; medullary (epiphysis/metaphysis/diaphysis); soft tissue extension.
— soft tissues: dermis/subcutaneous tissue/deep fascia/peripheral nerve/muscle/osseous extension/retroperitoneal.
— satellite nodules: size (cm) and distance (cm) from the main tumour.
— location: may be indicative, e.g. pelvis—Ewing's sarcoma or chondrosarcoma; chest wall—Askin (thoracopulmonary neuroectodermal) tumour, alveolar rhabdomyosarcoma.

Size
— length × width × depth (cm) or maximum dimension (cm).

Appearance
— solid/cystic/necrotic/lobulated/fatty/myxoid/cartilaginous/osseous.

Edge
— circumscribed/irregular.

Vessels
— relationship of tumour to vessels.

2. HISTOLOGICAL TYPE

Prior to histological evaluation of any bone or soft tissue sarcoma the pathologist must be aware of the patient's age, anatomical site of the lesion, subsite (e.g. epiphysis, metaphysis or diaphysis of bone) and, crucially, the radiological appearances. For example, a rapidly growing chest wall lesion in a young male may be nodular fasciitis rather than a sarcoma, peripheral chondroid lesions are benign whereas proximal are more likely to be malignant, and an epiphyseal lesion is likely to be a giant cell tumour (adult) or chondroblastoma (child) rather than an osteosarcoma (young/metaphysis). Age also closely correlates with type of soft tissue sarcoma: embryonal rhabdomyosarcoma (infants), synovial sarcoma (young adult), liposarcoma (middle age) and malignant fibrous histiocytoma (elderly). Close clinicopathological correlation is fundamental to the diagnosis.

Osteo-, chondro-, Ewing's/PNET, lipo-, synovial-, fibro-, rhabdo-, leiomyo-, angio-, malignant peripheral nerve sheath tumours, malignant fibrous histiocytoma and variants are amongst the main categories of sarcoma and each comprises variable numbers of subtypes (Tables 36.1 and 36.2).

Morphology is the mainstay of diagnosis but a panel of immunohistochemical antibodies to intermediate filaments and other markers (e.g. alkaline phosphatase on fresh tissue touch imprints for osteosarcoma), electron microscopy and cytogenetic analysis should be used as appropriate. This allows subclassification as to histogenetic type for a