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Paediatric Term
FRH 11th May 2009

PAEDIATRIC MUSCULOSKELETAL TUMOURS

CONTENT LAYOUT

- Classification
- Presentation
- Diagnostic clues
- Benign lesions
- Malignant lesions

CLASSIFICATION

- BONE TUMOURS
  - PRIMARY
    - BENIGN
      - Enchondroma
      - Osteochondroma
      - Chondroblastoma
      - Osteoid osteoma
    - MALIGNANT
      - Osteosarcoma
      - Ewings sarcoma
      - Leukemia
  - METASTATIC
    - Neuroblastoma
    - Rhabdomyosarcoma
- SOFT TISSUE TUMOURS
  - Rhabdomyosarcoma
- TUMOUR LIKE LESIONS
  - UBC
  - ABC
  - Eosinophilic granuloma
  - NOF (FCD)
  - Fibrous dysplasia
  - Osteomyelitis

PRESENTATION

- Pain
  - characteristic night pain
  - deep, dull, toothache type
  - initially intermittent then constant
  - progressive
  - NSAIDS
- Mass
- Fracture
- Incidental

DIAGNOSTIC CLUES

- Age
- Number of lesions
- Location of lesion
- Lesion reaction on bone
- Bone reaction on lesion
- Matrix characteristics

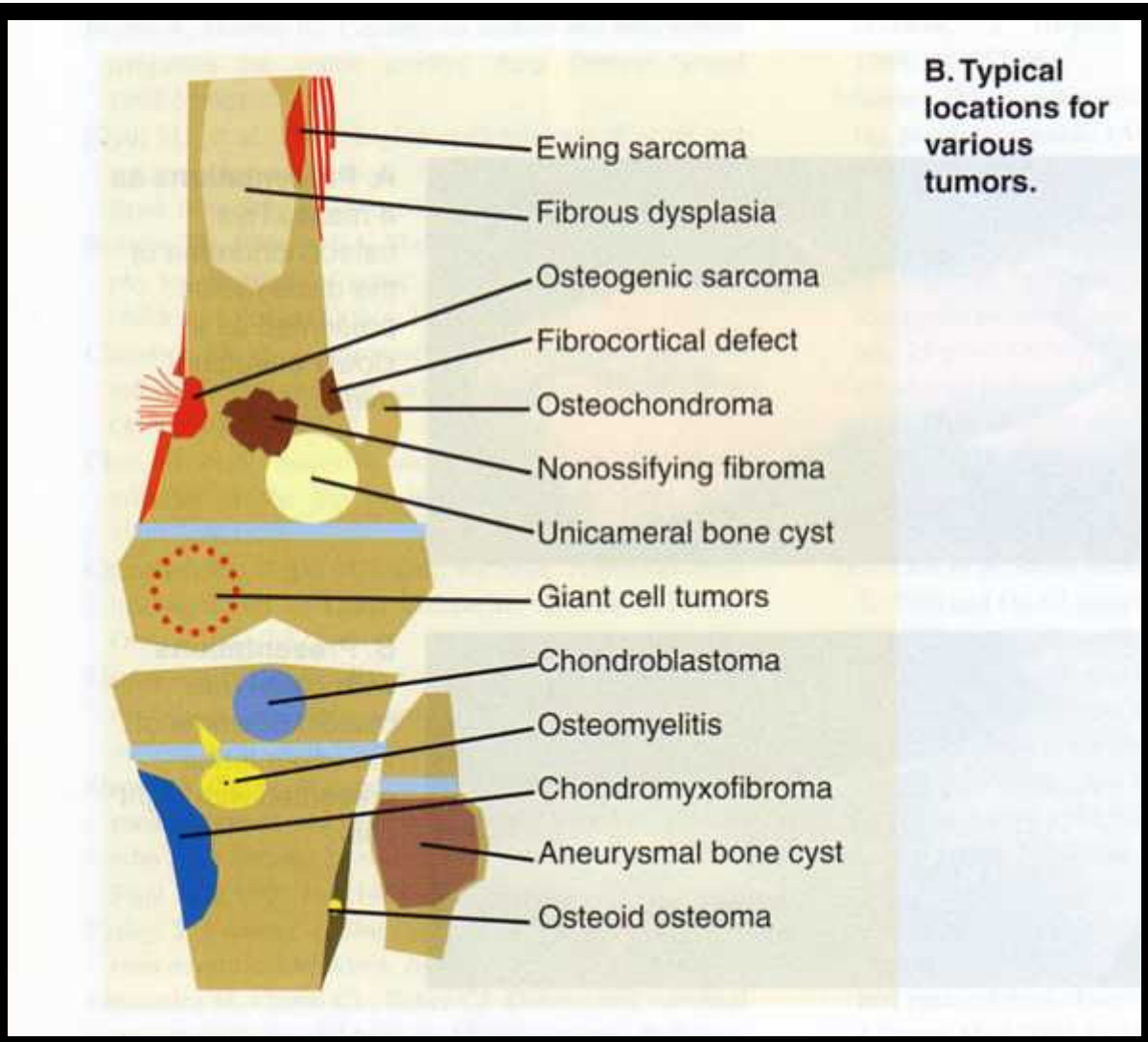
AGE

AGE	BENIGN	MALIGNANT
Below 5 yrs	Osteomyelitis Osteofibrous dysplasia	Leukemia Metastatic neuroblastoma Metastatic rhabdomyosarcoma
10-25 yrs	Eosinophilic granuloma Enchondroma Fibrous dysplasia Osteomyelitis	Osteosarcoma Ewings Leukemia

NUMBER OF LESIONS

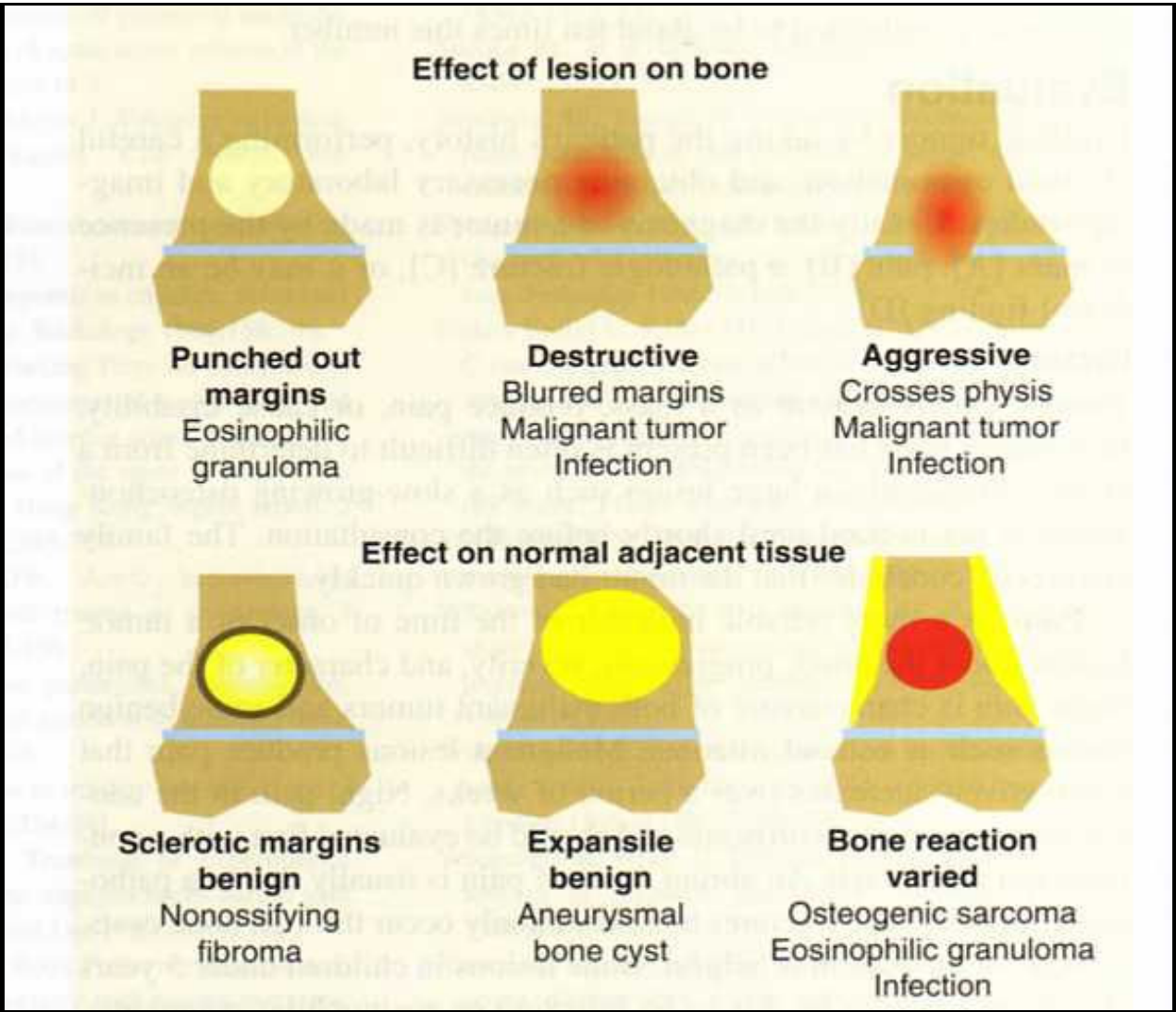
- Below 5 yrs
  - Multiple destructive lesions – neuroblastoma or wilms tumour
- Histiocytosis X
- Poly ostotic fibrous dysplasia

LOCATION

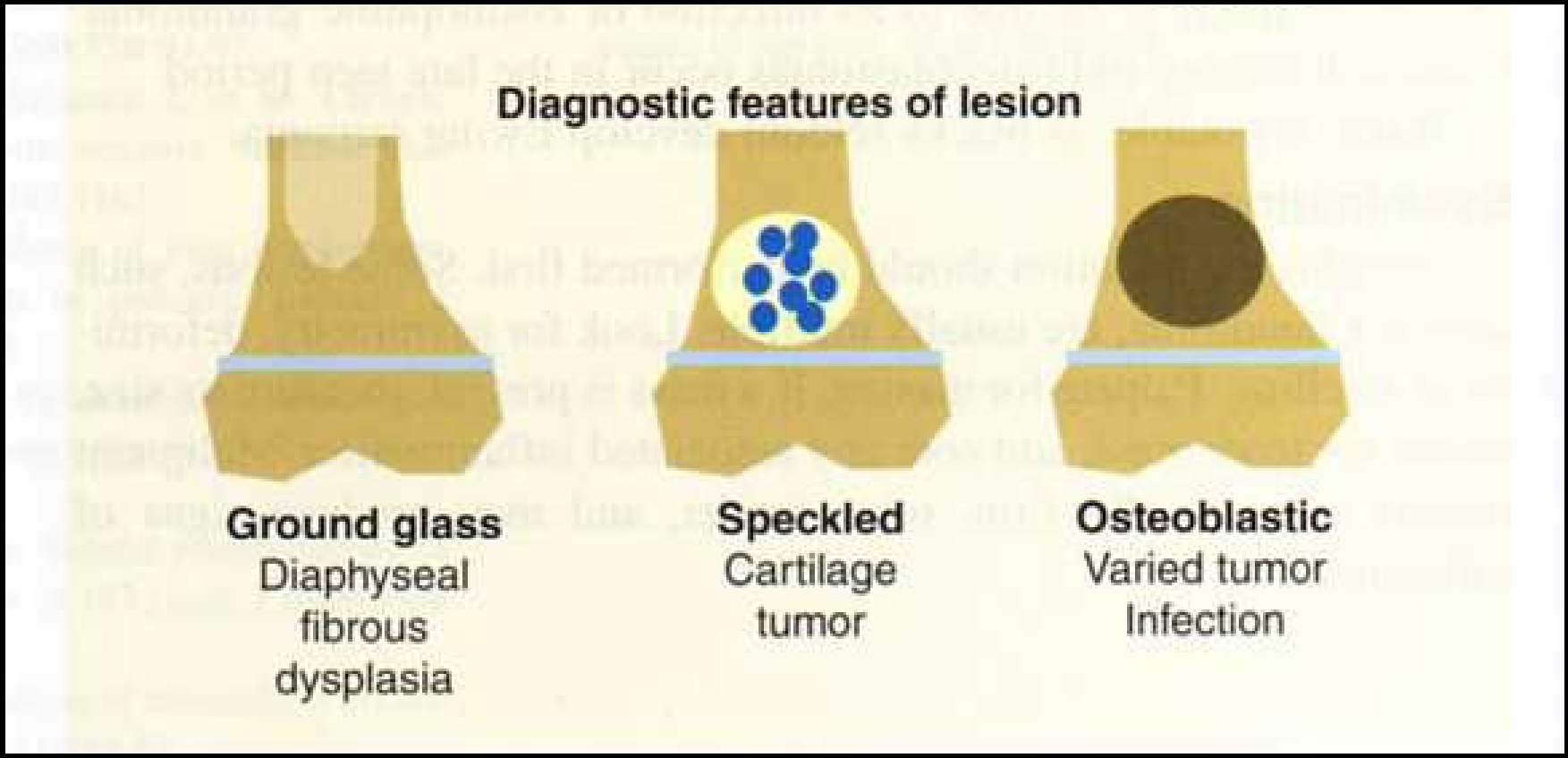


B. Typical locations for various tumors.

BONE AND TUMOUR REACTION

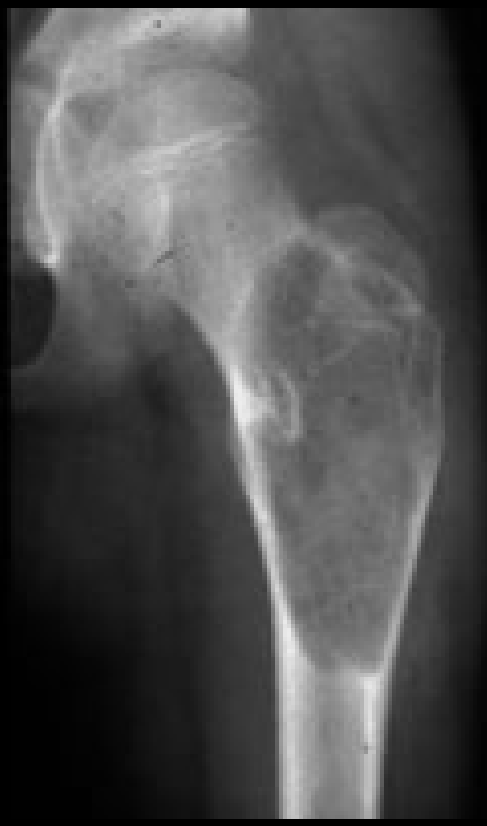


MATRIX


Diagnostic features of lesion
Ground glass
Diaphyseal
fibrous
dysplasia
Speckled
Cartilage
tumor
Osteoblastic
Varied tumor
Infection

BENIGN

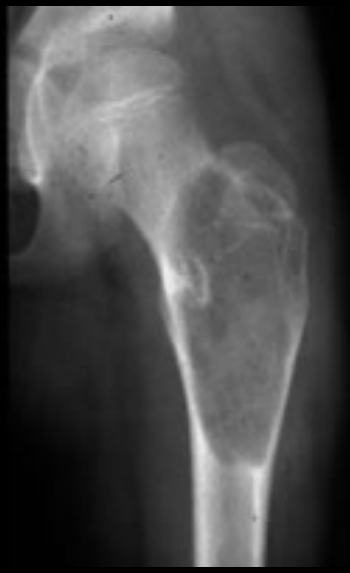
Unicameral Bone Cyst



- A benign cyst filled with straw coloured fluid
- Tends to heal spontaneously
- Children and Adolescents
- Typically in the metaphysis of a long bone
  - proximal humerus and proximal femur.
- **Usually asymptomatic**
  - Incidental x-ray finding
  - Pathological fracture
- **X-ray**
  - Well demarcated radiolucent area
  - Metaphysis
  - Often extends to physis
  - Cortex may be thinned
  - Bone may be expanded, SYMMETRICALLY
- **Active** – when cyst abuts the physeal plate, enlarging in sequential x-rays
- **Latent** – when normal bone intervening

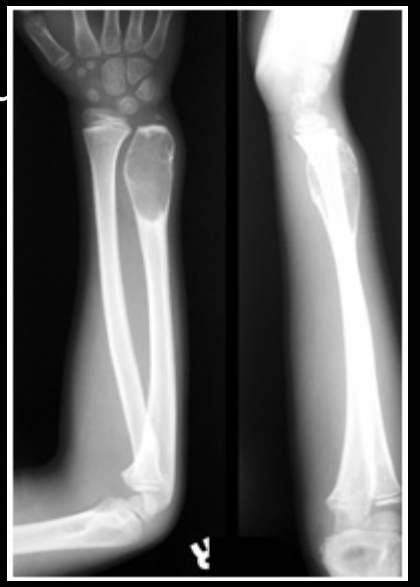
Unicameral Bone Cyst 2 Treatment

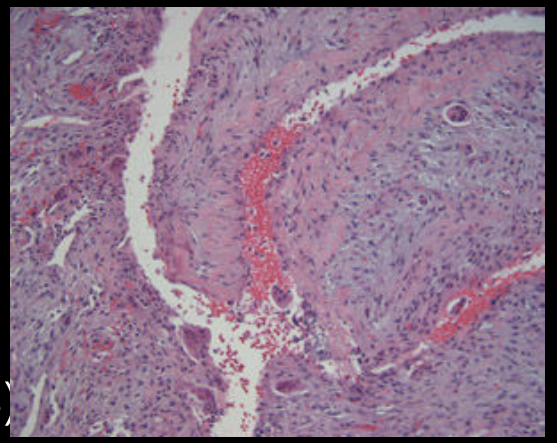
- Asymptomatic Latent lesions can be monitored
- Aspiration to confirm diagnosis
- **Active cysts**
  - Aspiration and injection of Methylprednisolone
  - May need to be repeated
  - Recalcitrant proximal humerus – curettage and grafting
  - Proximal femur – curettage, graft and internal fixation to avoid fractu
- **Pathological fracture**
  - Proximal humerus
    - Monitor – often stimulus for healing
    - Curettage, bone graft +- fixation
  - Proximal femur
    - Curettage, graft and internal fixation



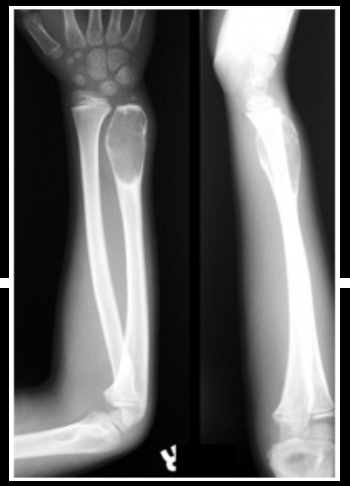
Aneurysmal Bone Cyst

- Benign but often very AGGRESSIVE with a high rate of recu
- Cyst may expand the bone and significantly weaken it
- Blood filled spaces without an endothelial lining
- Isolated or associated with other lesions
  - GCT
  - Chondroblastoma
  - Chondromyxoid fibroma
  - Fibrous dysplasia
- Swelling and pain
  - may have a soft tissue mass
  - May have been present for months or years
- Most common in second decade of life (10–20 years)

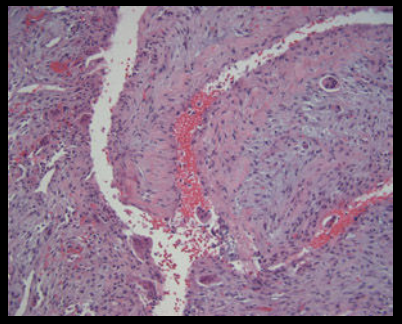




Aneurysmal Bone Cyst 2

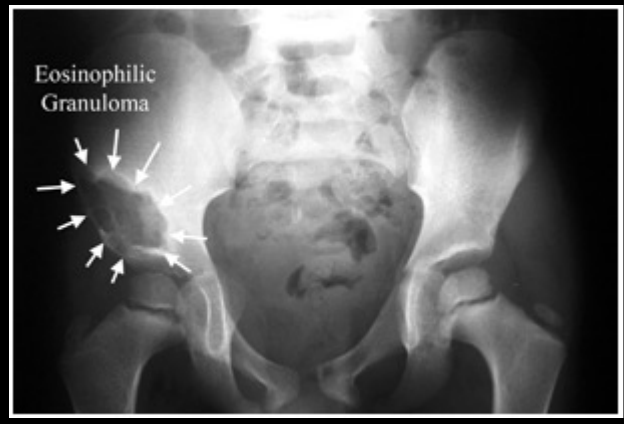


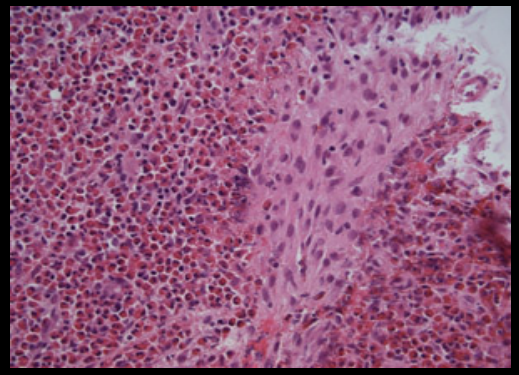
- Long bones usually in metaphysis
- Femur, tibia, vertebrae (however, may be seen in any bone)
- **X-ray**
  - Well defined radiolucent cyst , trabeculated and eccentric
  - Metaphysis/ PERIOSTEAL NEW BONE FORMATION
- **MRI** : fluid-fluid levels indicate blood within the lesion. There are often multiple cavities and there may be an associated SOFT TISSUE MASS
- **Treatment**
  - Curettage and bone graft
  - Curettage and cement
  - Occasionally , curettage, grafting and internal fixation



Eosinophilic Granuloma

- Langerhans Cell Granulomatosis or Histiocytosis X
  - Unusual group of disorders
  - Cells of R-E system form granulomatous lesions resembling bone tumours
  - Marrow containing bone is resorbed
  - Lytic lesions in flat bones or metaphyses of long bones
- 5-15 years
- Usually no pain
  - Incidental x-ray finding
  - Pathological fracture
- Skull, femur, pelvis, ribs, and spine—but any bone
- **X-ray**
  - Well demarcated oval radiolucency
  - Reactive sclerosis
  - May have multiple lesions
  - Skull – punched out appearance
  - Vertebral collapse – vertebra plana

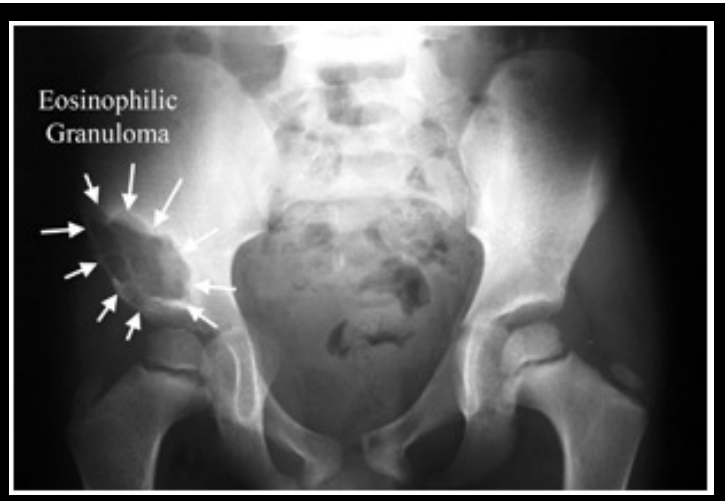




Eosinophilic Granuloma 2

Hand-Schuller-Christian disease

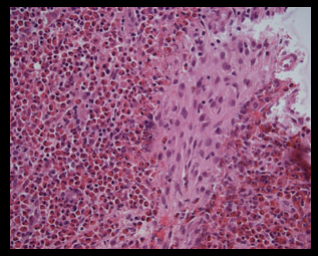
- Disseminated form
- Lesions in skull, vertebra, liver and spleen
- Anaemia and recurrent infection
- Individual lesions – curettage and radiotherapy
- Complete remission unlikely



Letterer-Siwe disease.

- Extremely rare
- Severe
- Seen in infants and usually progresses rapidly to fatal outcome

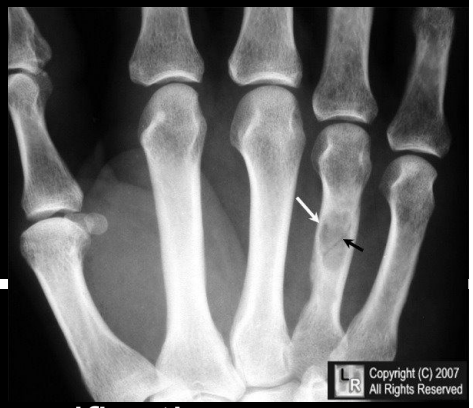
Symptoms: pain, swelling, occasionally loose teeth or excessive thirst (diabetes insepidus)



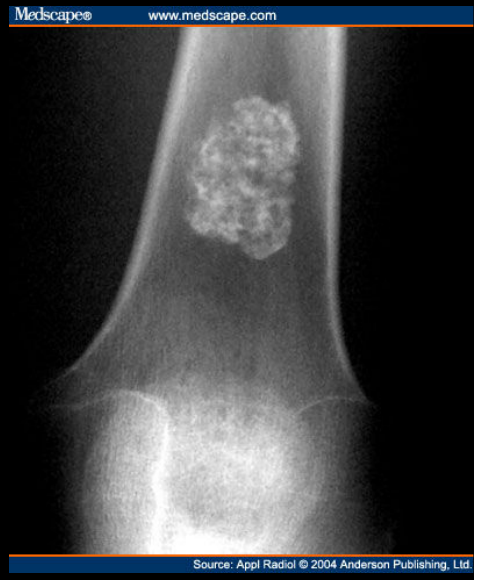
Treatment:

- Injection of solitary lesions with methylprednisolone is often curative.
- Curettage and bone grafting if risk of fracture.
- Low dose irradiation can be effective
- If there are multiple lesions or the vertebrae are involved and causing neurologic problems, systemic chemotherapy may be necessary.

Enchondroma

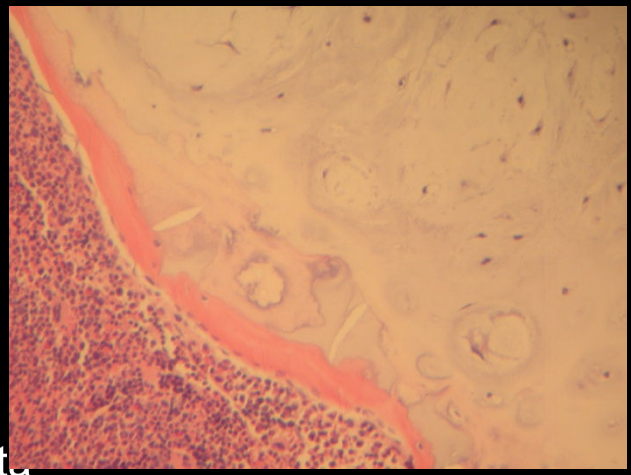


- Benign tumour within the medullary canal
- Islands of Cartilage persist in bones formed by enchondral ossification
- 2nd decade, but may be found in any age group
- Tubular bones of the hands and feet, femur and humerus
- **Symptoms:** often asymptomatic
  - Pathological fracture
  - Malignant transformation <2%
    - Pain
    - Enlargement
    - Cortical erosion
- **X-Ray** :
  - Well defined radiolucent lesion
  - Centrally placed junction of Metaphysis and Diaphysis
  - Punctate calcification within the lesion, expansion of the bone



Enchondroma 2

- MRI : well circumscribed lobular lesion within the medullary canal (bright on T2, low signal on T1)



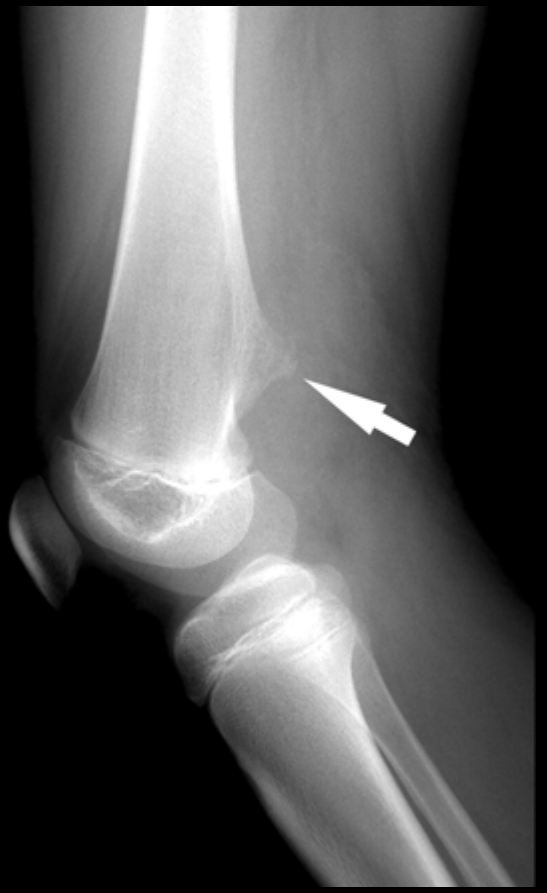
- **Ollier's disease**
  - multiple enchondromas
  - tend toward unilaterality
  - Dysplasia of involved bones
  - Sporadic inheritance
- **Mafucci's syndrome**
  - Ollier's + multiple soft tissue haemangiomata

There is a higher risk of malignant transformation when the enchondromas are associated with a syndrome.

- **Treatment**:
  - Asymptomatic – monitor with annual x-rays and exam.
  - Painful, enlarging or impending pathological fracture – curettage + bone graft +- ORIF
  - Pathological fracture - curettage + bone graft +- ORIF

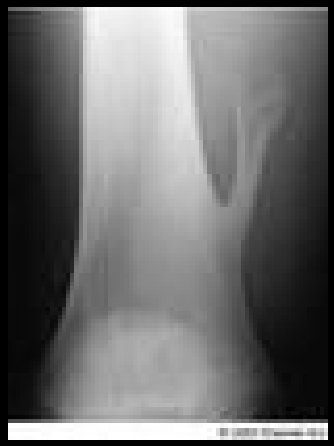
Osteochondroma

- A benign bony protuberance with a cartilage cap
  - Begins as overgrowth of cartilage at edge of physis
  - Enchondral ossification
  - Stops growing at skeletal maturity
- 2nd decade of life
- Ends of long bones and iliac crest
-
- **Symptoms:**
  - Mass
  - Pain
    - Overlying bursa
    - Impingement of soft tissues
  - Rarely paraesthesia
- **X-Ray**
  - Well defined exostosis emerging from metaphysis
  - Base co-extensive with parent bone
  - Sessile or pedunculated
- **MRI**
  - Cartilage cap on the bony stalk is well visualized

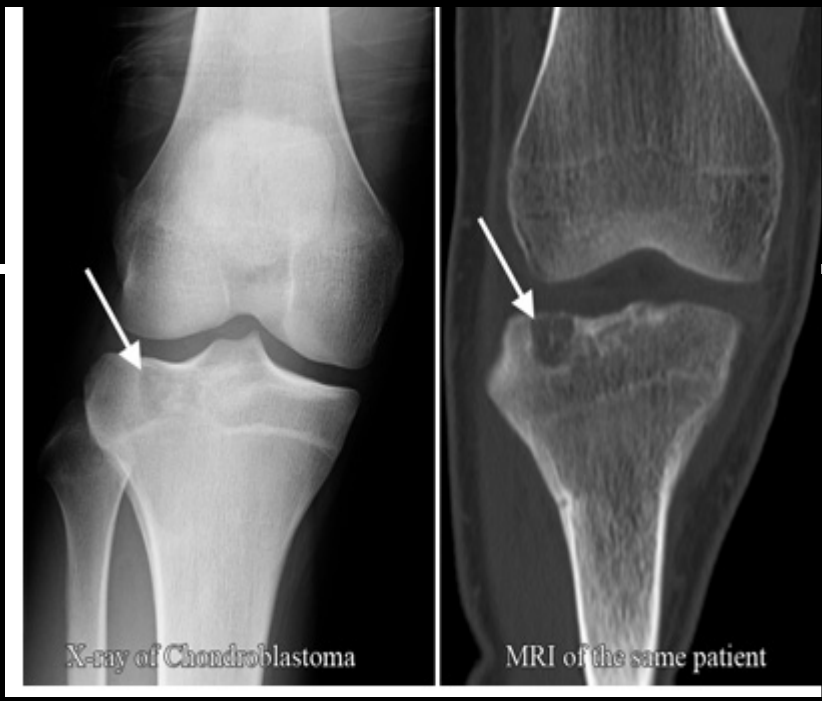


Osteochondroma 2

- **Multiple hereditary exostoses**
  - Multiple osteochondromas
  - Most common skeletal dysplasia
  - Autosomal dominant transmission
  - 1-2 % risk of transformation to chondrosarcoma
- **Treatment**:
  - Asymptomatic - observation
  - Surgical excision
    - Pain
    - Soft tissue impingement
    - Paraesthesias
  - Suggestion of malignant transformation
    - Pain
    - Growth after skeletal maturity
  - Osteotomy for deformity correction



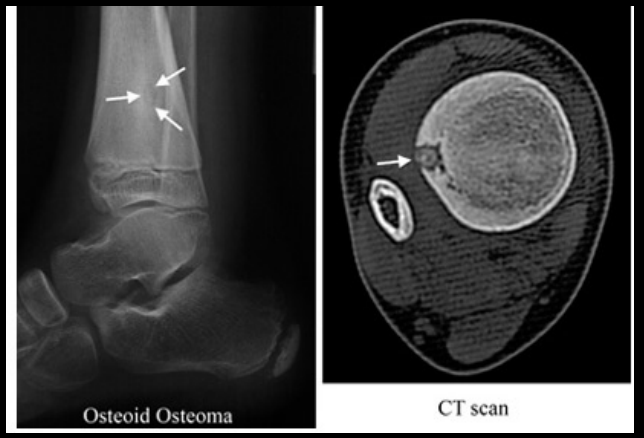
Chondroblastoma

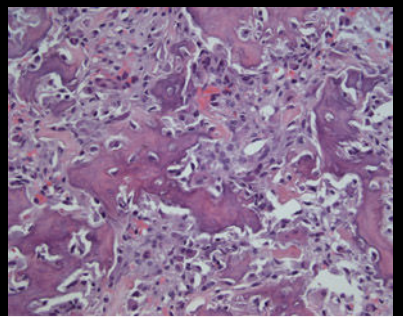


- Benign slow growing lesion
  - Immature cartilage cells
- Presents 15-25 years
- Epiphysis
  - proximal humerus, distal femur and proximal
- **Symptoms**
  - joint pain, often present for months to years
- **X-Ray**
  - well circumscribed
  - Radiolucent
  - Occasionally breaches physis and/or articular surface
- **Treatment**
  - End of growth period is preferable
  - Marginal excision if possible + bone graft
  - Curettage and bone grafting (rate of recurrence 10-20%)
  - Inherent risk of joint damage

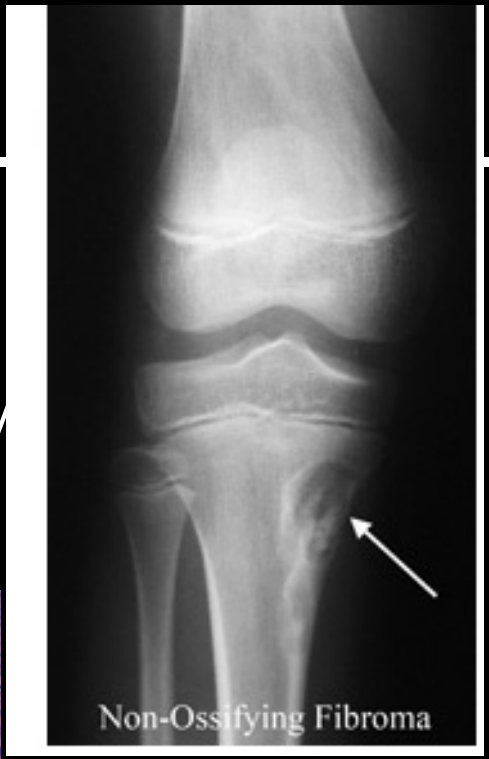
Osteoid Osteoma

- Benign, small nest of osteoid surrounded by osteoblasts
- 2nd decade. 3 males: 1 female
- **Location**
  - Any bone except skull
  - proximal femur most common
- **Symptoms**
  - Persistent pain - often worse at night
  - Limp, muscle wasting
  - Spine – intense pain, muscle spasm, scoliosis
  - May be relieved by anti-inflammatory medications (Ibuprofen, Aspirin).
- **X-Ray** small radiolucent nidus surrounded by dense reactive bone
- **Bone scan**: intense localised activity
- **CT** : best study to visualize the nidus within the dense reactive bone
- **Treatment**
  - CT guided Radiofrequency ablation
  - Surgical excision is usually curative

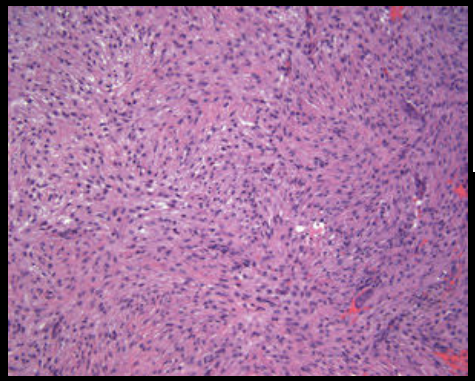




Non-Ossifying Fibroma (fibrous cortical defect)



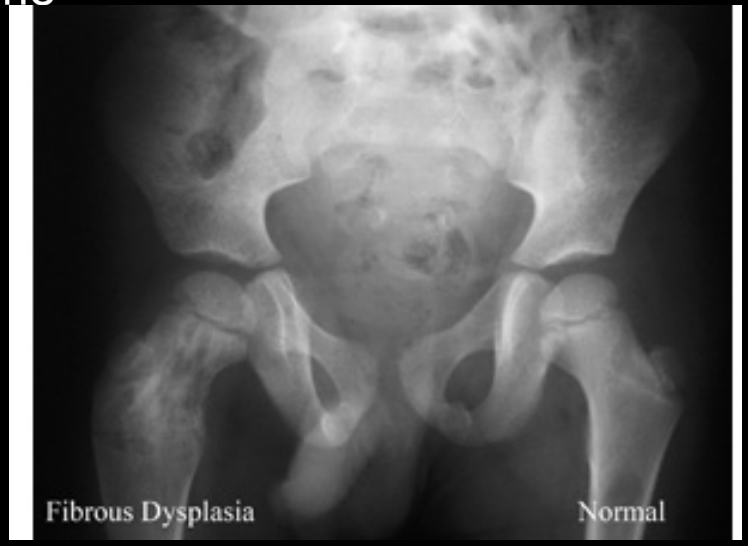
- Commonest benign bone lesion
  - Persistent nest of fibrous tissue within a bone
- Distal tibia and distal femur, usually in the metaphy
- **Symptoms**
  - Asymptomatic
  - Incidental finding
  - Rarely pathological fracture
- **X-ray**
  - Oval radiolucent area
  - Thin margin of dense bone
  - Adjacent to or within cortex



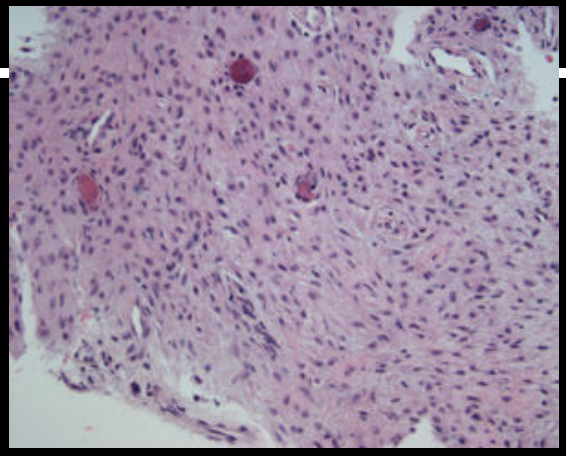
- **Treatment**
  - Small lesions can be observed, resolve near skeletal maturity
  - Large lesions that risk fracture can be treated with curettage and bone grafting

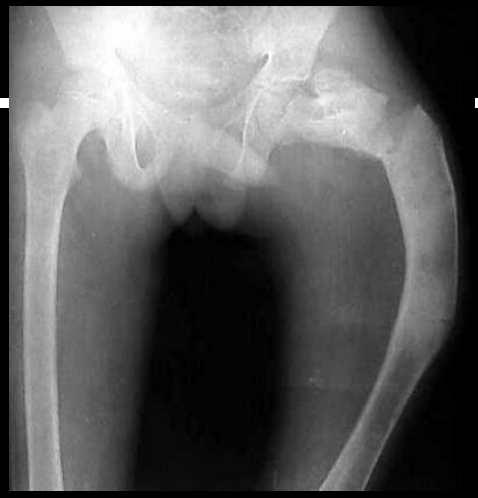
Fibrous Dysplasia

- Developmental disorder of bone
  - Replacement of trabecular bone with disorganized fibrous tissue containing flecks of osteoid and woven bone
  - Genetic mutation - increased cAMP
- 2nd and 3rd decades
- skull, jaw, ribs, proxymal femur, tibia
- 80% **monostotic**
- **Polyostotic**
  - Present earlier with progressive deformity, pathological fractures
  - **McCune-Albright syndrome**
    - Café-au-lait
    - Precocious puberty



Fibrous Dysplasia 2



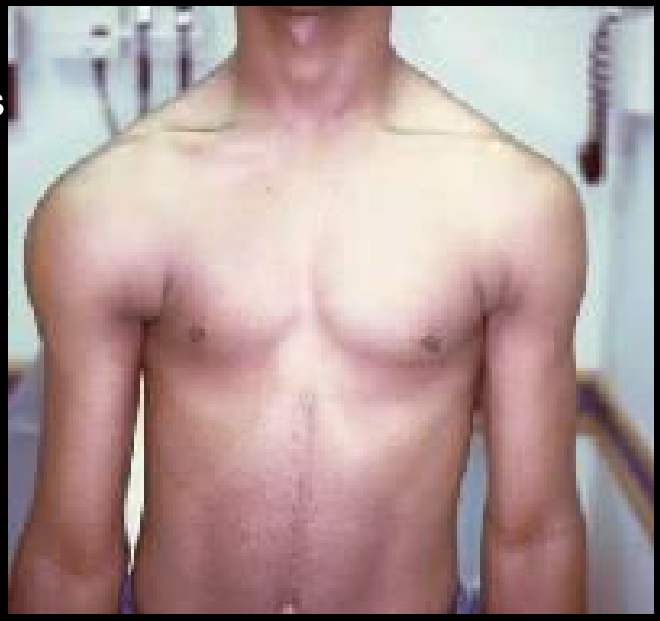


- **Symptoms**
  - Pain
  - Progressive deformity
  - Pathological fracture
- **X-ray**
  - Radiolucent cystic areas in metaphysis and diaphysis
  - Hazy ground glass appearance
  - Deformity of wt bearing bones eg. shepherd's crook deformity of the proximal femur
- **Treatment**
  - Depends on extent of defect and presence of deformity
  - Curettage, bone grafting, +- cement +- stabilization with ORIF
  - Deformity correction
  - Diphosphonates may help with pain associated with multiple lesions

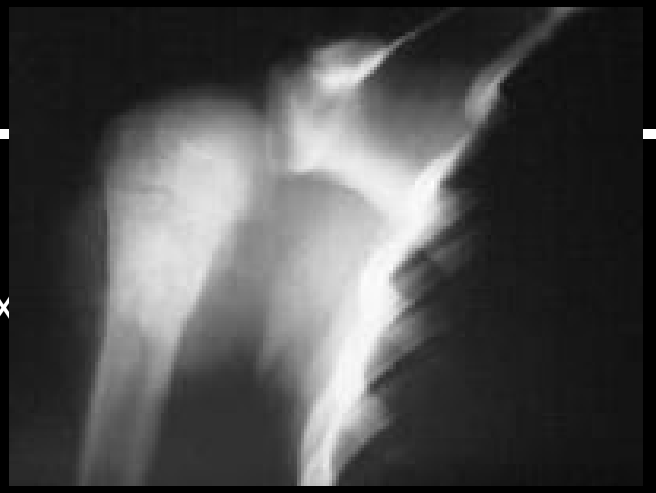
Malignant

Osteosarcoma

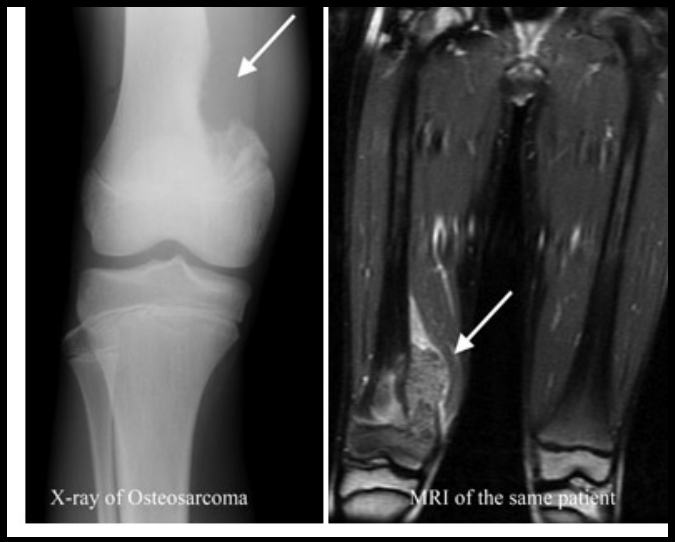
- High grade intramedullary osteosarcoma
- The most common primary malignant bone tumour
  - Primitive mesenchymal bone-forming cells
  - Osteoid or bone synthesized
- Most common age: 10-20 years
- Usually metaphyseal
  - distal femur , proximal tibia and proxymal humerus
- **Clinical Features**
  - Pain
    - Constant
    - Worse at night
    - Gradually increases in severity
  - Mass
  - Pathological fracture rare
  - Weight loss in advanced cases
  - Overlying tissues may be swollen and inflamed
  - Local tenderness
  - Raised ESR and Alk Phos



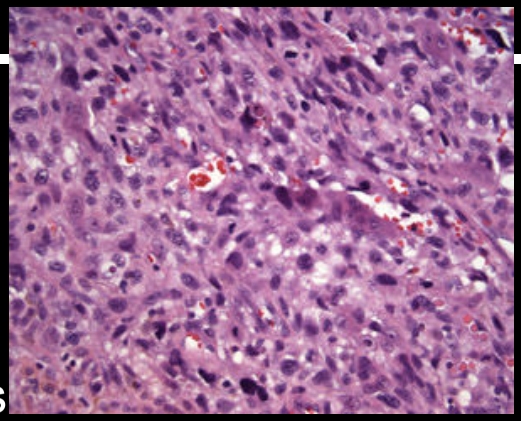
Osteosarcoma 2



- **X-Ray**
  - Poorly circumscribed and may be lytic, blastic or mix
  - Endosteal margin poorly defined
  - Trabecular and cortical bone destruction
  - Cortical breaching (sunburst effect)
  - Periosteal reactive new bone (Codman's triangle)
- **MRI**
  - Extent of mass locally
  - Identify skip lesions
- **Bone scan**
  - Bone mets
  - Skip lesions
- **CT**
  - Staging
    - 10 – 20 % have pulmonary mets at presentation



Osteosarcoma 3
Biopsy



- In specialist centre
  - After CT and MRI
  - Open
  - By surgeon likely to be doing definitive surgical excis
  - Site and tract selected to be included in subsequent ablative excision
  - Avoid transverse incisions
  - Approach through muscle to avoid compartment transgression
  - Minimal exposure of tumour
  - Block of tissue removed from boundary zone
    - Normal tissue
    - Pseudocapsule
    - Abnormal tissue
  - If bone removed cover raw area with bone wax or cement
  - Full haemostasis after releasing tourniquet
  - Avoid drains

Osteosarcoma Management – Current principles 1

- Staging of primary lesion and search for other lesions
- MRI of the primary site including the joint above and below
- Bone scan; bone metastases and skip lesions
- CT chest
- Paediatric oncology consultation
- Incisional biopsy
- Intraoperative frozen section
- Line placement for chemotherapy

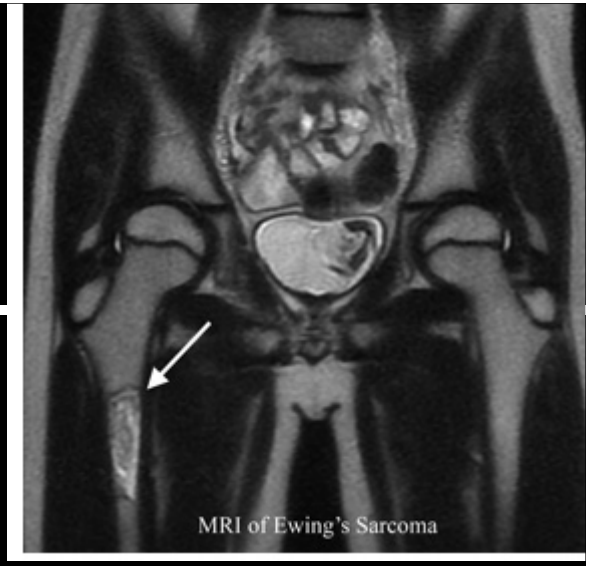
Osteosarcoma Management – Current principles 2

- Preoperative neoadjuvant chemotherapy (usually multiagent chemotherapy)
- Repeat MRI after chemotherapy and prior to definitive surgery,
  - evaluation of the tumour response to chemotherapy
  - change in size
  - change in amount of oedema,
  - involvement of neurovascular structures)
- Surgical planning
- Surgery
  - excision with wide surgical margins
  - Limb salvage
  - reconstruction
- Amputation

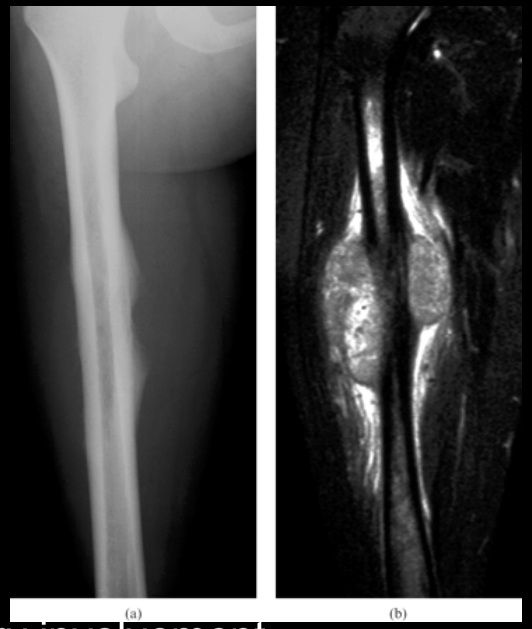
Osteosarcoma Management – Current principles 3

- Histological examination of resection specimen
  - Tumour response to chemotherapy (> 90% tumour necrosis demonstrates good response)
- Verification of wide surgical margins
- Continue chemotherapy (adjuvant chemotherapy)
  - Same protocol if >90% necrosis
  - Change agents if <90% necrosis
  - 6-12 months
- Follow up
  - Every 3 months first year
  - Then 6 to 12 months intervals
- Pulmonary Mets (small and peripheral) – resected with wedge of lung tissue

Ewing's Sarcoma

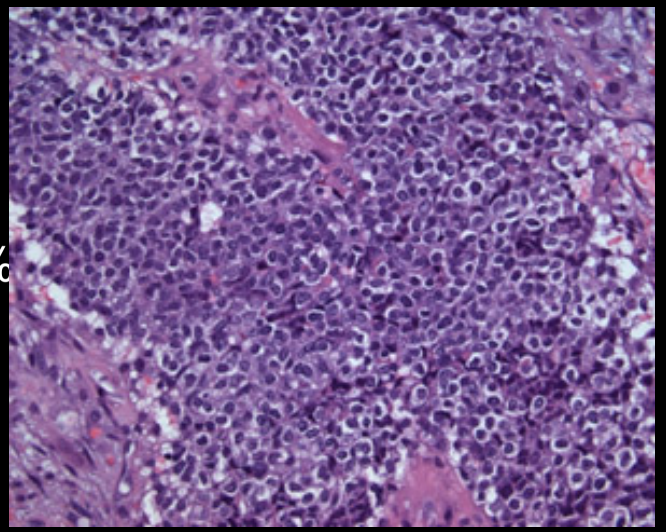


- Malignant small round blue cell tumour
- Consistent chromosomal translocation (t11:22)(q24:12)
- 5-20 years
- Pelvis , distal femur, proximal tibia, femoral diaphysis and prox humerus
- **Clinical features**
  - Pain
  - Mass
  - Generalised illness
  - Fever
  - Weight loss
  - Raised ESR
- **X-ray**
  - Large destructive lesion in metaphysis and diaphysis
  - Periosteal new bone
    - In multiple layers – onion skin appearance
    - Codman's triangle
  - Sunburst appearance
  - Soft tissue mass
- **MRI** : large soft tissue component, long segments of medullary involvement



Ewing's Sarcoma 2

- **Treatment**
  - Aggressive multi-agent chemotherapy
  - Complete resection of the tumour with a margin of normal tissue, which may require amputation or extensive surgical reconstruction
  - Maintenance chemotherapy
  - Radiation therapy may also be necessary



- **Survival**
  - Long term with multi-modality therapy 60-70%
  - Poor prognostic factors
    - Spine and pelvic tumours
    - Tumours >100 cm^3
    - Poor response to chemotherapy (<90%)
    - Elevated Lactate Dehydrogenase levels

Thank you