Bone tumours – general

Modified WHO classification of benign bone tumours

I. Bone forming tumours
  i. Osteoma
  ii. Osteoid osteoma
  iii. Osteoblastoma
II Cartilage forming tumours
  i. Chondroma
  ii. Osteochondroma
  iii. Chondroblastoma
  iv. Chondromyxoid fibroma
III Giant cell tumours
IV Marrow tumours (none)
V Vascular tumours
  i. Haemangioma
  ii. Lymphangioma
  iii. Glomus tumour
VI Other connective tissue tumours
VII Other tumours (nervous)
  i. Neurilemmoma
  ii. Neurofibroma
VIII Unclassified tumours (none)
IX Tumour like lesions
  i. Solitary bone cyst
  ii. Aneurysmal bone cyst
  iii. Metaphyseal fibrous defect
  iv. Eosinophilic granuloma
  v. Fibrous dysplasia
  vi. Osteofibrous dysplasia
  vii. Myositis ossificans
  viii. Brown tumour of hyperparathyroidism
  ix. Intraosseous epidermoid cyst
  x. Giant-cell (reparative) granuloma

Enneking classification of benign bone tumours

Stage 1
-Latent benign bone tumour
-This does not progress, or heals spontaneously
-E.g. non-ossifying fibroma

Stage 2
-Active benign bone tumours
-This can expand and even deform the bone but is fully confined by the bone
-E.g. giant cell tumour

Stage 3
-An aggressive benign bone tumour
-This invades and destroys the bone and extends into the soft tissue
E.g. aggressive giant cell tumour

Staging system of Musculoskeletal Tumour Society

Stage I
-All low grade tumours

Stage II
-All high-grade tumours

Stage III
-All tumours with metastases

If the neoplasm is intracompartmental it is considered stage A
If it is extracompartmental it is considered stage B

This system is applicable to tumours of bone and soft tissue.
It is not applicable to tumours of bone marrow (e.g. lymphoma, leukaemia) which have their own system.

Low grade tumours have a less than 25% chance of metastasis. High grade tumours have more than a 25% chance of metastasis.

Epidemiology of bone tumours

1. In US bone tumours make up 0.2% of all tumours
2. Soft tissue tumours make up 2%
3. Of primary bone sarcomas:
  i. Osteosarcoma 35%
  ii. Chondrosarcoma 26%
  iii. Ewing's tumour 16%
  iv. Chordoma 8%
  v. MFH and fibrosarcoma 6%
  v. Angiosarcoma 1%
4. Bimodal distribution for bone tumours – second and seventh decade
5. Malignant tumours and patient age
  1-30 Ewing's tumour Osteosarcoma
  30-40 Fibrosarcoma and MFH

		Malignant giant cell tumour
		Reticulum cell sarcoma (primary lymphoma of bone)
		Parosteal osteosarcoma
	40 plus	Mets
		Myeloma
		Chondrosarcoma

6. Gradual increase in incidence of soft tissue tumours
7. Males>females for bone tumours (except GCT and parosteal and periosteal osteosarcomas)
8. Whites>blacks for bone tumours

Location of tumours within bone

Epiphyseal

1. Giant cell tumour
2. Chondroblastoma

Metaphyseal

1. Osteosarcoma
2. Non-ossifying fibroma
3. Chondromyxoid fibroma

Metadiaphyseal

1. Enchondroma
2. Ewing's tumour

Diaphyseal

1. Fibrous dysplasia
2. Eosinophilic granuloma
3. Ewing's tumour

Spine

1. Aneurysmal bone cyst } Posterior
2. Osteoblastoma } elements

Tibia

1. Adamantinoma
2. Osteofibrous dysplasia

Radiology of bone tumours

Questions to ask on reviewing XR

1. Where in the bone is the tumour?
2. What is the tumour doing to the bone?
3. What reaction to the tumour does the bone show?
   -A dense sclerotic margin around the tumour is a characteristic sign of a benign bone tumour
   -Clyde Helm feels the most reliable indicator of benign vs. malignant is the zone of transition
   i. The zone of transition is the border between the lesion and normal bone
   ii. It is narrow if the zone can be so well defined that it can be drawn with a fine tipped pen
   iii. It is wide if it is imperceptible and cannot be drawn
   iv. If a lesion has a narrow zone of transition it is a benign process
   v. If a lesion has a wide zone of transition it is an <u>aggressive</u> (not necessarily malignant) process
   vi. The zone of transition is only valid on plain films, not MRIs
   vii. The zone of transition only applies to lytic lesions, not blastic lesions (which will always have a narrow zone of transition)
4. What periosteal responses are present?
   i. A slow growing benign tumor will cause a thick, wavy, uniform or dense periostitis because it is a low grade chronic irritation that will give the periosteum time to lay down thick new bone and remodel into more normal cortex. Malignant lesions will not cause a benign periostitis; as noted below benign lesions can cause a "malignant periostitis"
   ii. A malignant tumour causes a periosteal reaction that is high grade and more acute and hence the periosteum doesn't have time to consolidate and it may appear lamellated (onion skinned), amorphous or sunburst
   Note that benign lesions can cause an aggressive periosteal reaction. These include:
   a. Infection
   b. Eosinophilic granuloma
   c. Aneurysmal bone cyst
   d. Osteoid osteoma
   e. Trauma

5. Is there any extension into soft tissues?
6. Is there any matrix mineralization?

Note: reactive bone is most mature on the periphery.
Malignant bone is most mature centrally.

Patterns of bone destruction

1. *Permeative destruction*
-Implies rapid growth
-Lesions insinuate themselves between trabeculae of cancellous bone stimulating osteoclastic activity and fine rarefactions

2. *Moth eaten*
-Implies intermed. aggressiveness
-Coarse destructive pattern

3. *Geographic bone destruction*
-Implies low grade destruction
-Provokes resorption of all osseous tissues in their path

Causes of benign cystic lesion
FEGNOMASHIC

F fibrous dysplasia
E enchondroma, eosinophilic granuloma
G giant cell tumour
N nonossifying fibroma
O osteoblastoma
M mets and myeloma
A Aneurysmal bone cysts
S solitary bone cyst
H hyperparathyroidism (brown tumour)
I infection
C Chondroblastoma, chondromyxoid fibroma

Causes of lytic permeative processes

1. Osteosarcoma
2. MFH
3. Ewing's tumour
4. Multiple myeloma
5. Leukaemia
6. Lymphoma
7. Eosinophilic granuloma
8. Infection
9. Aggressive osteoporosis (seen in disuse osteoporosis)

Oncologic margins
Intralesional
The tumour is removed but no attempt is made to obtain normal tissue around it.

Marginal
The tumour is removed completely but the surrounding reactive zone is not completely removed.

Wide marginal
The tumour is removed completely with a surrounding cuff of normal tissue.

Radical marginal
The tumour is removed along with the entire compartment in which the tumour lies. E.g., if the distal femur contains the tumour the whole of the femur is removed..

Other terms
Reactive zone
An area composed of capillary proliferation apparently surrounding a tumour as it grows.

Biopsy remarks

1. The biopsy should traverse only one compartment, and avoid neurovascular structures and joints
2. The biopsy tract should be excised at the definitive operation
3. Haemostasis is mandatory
4. The biopsy should be performed at the interface of normal and tumorous tissue. Areas of fracture should be avoided
5. Adequacy of biopsy should be checked prior to closure
6. The biopsy should be performed at the treating institution as rates of complications are five times higher when done at peripheral hospitals, and there are higher rates of amputation and decreased survival.

Needle biopsy vs. open biopsy
Advantages of needle biopsy

1. Cheaper
2. Avoids need for general anaesthetic
3. Decreased risk of haematoma
4. Can use accurate image guidance (e.g. CT)
5. Less morbidity

Disadvantages

1. Higher risk of sampling error

2. Not enough tissue for special studies such as flow cytometry, gene rearrangement, electron microscopy and immunohisto-chemical staining.

Limb Salvage
For limb salvage to be acceptable, the survival rates must be similar to amputation and function at least equivalent.

Limb salvage is less attractive than amputation with:
1. Pathological fractures, where the adjacent soft tissues are contaminated and there is a higher risk of local recurrence. Patients who have osteosarcoma and local recurrence should go on to immediate amputation.
2. Neurovascular involvement is a relative contraindication; vessels can be reconstructed, and in the upper limb tendon transfers or nerve grafts may retain function.

General points
1. It is rare for primary malignant bone tumours to be discovered or be symptomatic when the lesion is less than 6cm in diameter.

Antigens
S-100 "Every cell is S-100 at some time in its life – Unni"