Malignant bone tumors

Incidence
Myeloma 45%
Osteosarcoma 24%
Chondrosarcoma 12%
Lymphoma 8%
Ewing's Sarcoma 7%

Commonest primary bone sarcoma is osteosarcoma

X-ray Questions to ask

1. Solitary or Multiple
2. What type of bone involved?
3. Which part of the bone: Epiphyseal, Metaphyseal or Diaphyseal
4. Margins: Well defined/ill defined
   Geographic Benign
   Malignant: Moth eaten
   Permeative
5. Periosteal reaction
6. Any calcification in the lesion

Geographic Permeative Moth Eaten
**Zone of transition**

It is the most reliable indicator to differentiate benign from malignant lesion. The zone of transition is the border between the lesion and the normal bone. Benign lesions are well defined [narrow zone of transition = fine line or sclerosed]. In malignant lesions there is ill defined [Wide zone of transition]. Zone of transition is applied to lytic lesion only.

**Beware:** Eosinophilic granuloma

- Infection
- Osteoid Osteoma
- Stress fractures

These benign lesion may have radiological appearance of malignancy.

**CT, MRI & BONE scan Assessment is essential**

**CT**

- Bone involvement better seen [eg Osteoid osteoma]
- Difficult areas: Pelvis and spine and scapula
- Pulmonary metastasis

**MRI**

- It is a gold standard
- Defines soft tissue involvement well seen
- Skip lesion
- Marrow spread
- Activity of the tumor [Gadolinium]
- Neurovascular relation
- T1 good for anatomy and T2 for pathology

Gadolinium: Only rim enhancement means benign cyst
Enneking Staging of bone sarcomas

Stage I: Low grade sarcomas

Stage II: High grade sarcomas

Stage III: High grade + Metastasis

(A: Intracompartment
B: Extracompartment)

SURGICAL MARGINS

RADICAL RESECTION
WIDE EN BLOC
MARGINAL
INTRACAPSULAR
Osteosarcoma

1970’s 20% 5 yrs survival; Present 80% 5 yr survival.

Increased frequency: with Li Fraumeni Syndrome
TP 53 deficiency
Retinoblastoma

Age: I Peak is in the II decade [75%]; II peak is after 60 yrs

Sex : Male: Female = 1.5: 1

Common sites : Around knee 50%
Proximal shoulder

Biochemistry
Elevated alkaline phosphotase 50% patients
Recently LDH: Prognostic

I Classic osteosarcoma Chondroblastic
Osteoblastic
Osteoblastic

II The surface [Juxtacortical]
Parosteal,
Periosteal,
High-grade surface tumors.

III Secondary osteosarcomas:
Paget disease
Post-irradiation sarcomas,
Fibrous dysplasia and bone infarcts.
Presence of secondaries in the lungs [15% Survival Vs 80% survival at one year]

Paget’s osteosarcoma has worse prognosis.
Treatment
1. Discuss with tumor centre and biopsy
2. Principle of management:
   Surgical: Amputation Vs Limb preservation.
   Mortality rate is similar and presently limb preservation

Limb preservation: I biopsy
   Chemicaltherapy
   Chemotherapy: <90% necrosis +ve cells
   En bloc excision of the tumor
   Part is replaced by artificial joint or allograft
   Continue Chemotherapy
   Observation for recurrence

Patient with secondaries in the lungs: May need excision of secondaries

MACI Methotrexate, Adriamycin, Cisplatin, and Ifosfamide [MACI].
   are commonly used. Also doxorubicin is used

Prognosis
Recent report based on 560 cases of Osteosarcoma
Patients with inadequate surgical margin, should undergo immediate amputation especially with poor chemotherapy response
Currently: long term survival is 60 to 80% in non-metastatic tumor
   Those with metastases: 10-20% at 5 yr

Parosteal Osteosarcoma

4% all Osteosarcoma
Common in females over 20 years
Best prognosis [94% survival]
75% distal posterior femur.
Rest: Proximal humerus and tibia.
**X ray**
Lobulated sclerotic mass
No involvement of medullary canal usually
Pasted on appearance

**Treatment**
Wide excision
No chemotherapy is required

**Chondrosarcoma**

Most common sarcoma of bone in patients over 40 years
Il common bone sarcoma [after Osteosarcoma ]
It represents about 25% of all bone sarcoma
Typically occurs in 40 and 60 years.

Types
- Primary
- Secondary chondrosarcoma: <1% of Osteochondroma
  - 1-2 multiple exostosis
  - 50% with Olliers and Maffucci’s Enchondroma

These secondary sarcomas are almost always histologically low-grade

Site: Pelvis is the commonest site
- Femur, Humerus, scapula, ribs

Painful encondroma in a long bone:
- Suspect malignancy

**X ray**
Botchy calcification
- Pop corn appearance
Cortical involvement destruction
- Soft-tissue extension.
- Periosteal reaction
CT: Investigation of choice
MRI: For soft-tissue extension
  and Relation to neurovascular bundle.

Medullary fill greater than 90% is predictive of chondrosarcoma.

Open biopsy preferred. Biopsy specimens should be taken from the areas of most concern.

A major drawback is sampling error due to tumor heterogeneity.

The low-grade chondrosarcoma pattern consists of cartilage cells that permeate marrow spaces and completely replace the marrow fat. The cartilage cells directly abut and surround the lamellar bone in the chondrosarcoma pattern.

**Treatment**

It requires surgical excision for cure.

Current chemotherapy and radiation have no significant role.

Grading systems are not standardized.

Low grade tumours amenable to limb sparing procedures.

Radical radiotherapy may be used to control inoperable.

**EWINGS SARCOMA**

Common site is Femur [20%]

More common in males, less than 20 years

**Clinical**

  - Pain, tenderness, fever
  - Anemia, leukocytosis, ESR
  - May mimic osteomyelitis
With Modern Chemotherapy: Survival is 70%

Present treatment
Pre-op Chemotherapy
Surgical resection when possible [otherwise radiotherapy]
Limb sparing surgery like osteosarcoma
Monitor for recurrence

Chemotherapy
Doxorubicin + Vincristine + cyclophosphamide + Actinomycin D + Adriamycin. Recently: added Ifosfamide

Lung radiotherapy is indicated for patients with lung metastases
**Myeloma**

Usually occurs in 50-80 years
Common primary bone tumor 45%
Males more commonly affected
Lymphocytes produce the same immunoglobulin. These Ig are called the paraproteins is common. IgG 55%
In some patients only part of the Ig is produced, usually the light chain and these appear as Bence-Jones proteinuria.
IgA myeloma [21%] carries poor prognosis

**Clinical**

Bone pain - usually ribs and vertebral
Pathological fracture
Deposition of amyloid [15%]
Hypercalcaemia
Amyloidosis

**Investigations**

<table>
<thead>
<tr>
<th>General</th>
<th>Hemoglobin and ESR</th>
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<tbody>
<tr>
<td>State of renal function</td>
<td>Urea and electrolytes</td>
</tr>
<tr>
<td>Bone destruction</td>
<td>Blood Calcium is high</td>
</tr>
<tr>
<td>Presence of bone fractures</td>
<td>X ray, Alkaline Phosphatase</td>
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<tr>
<td>Degree of immune</td>
<td>Plasma Ig [Electroporesis]</td>
</tr>
<tr>
<td>Degree of bone marrow</td>
<td>blood counts, Plasmacytes</td>
</tr>
<tr>
<td>Degree of haemostasis</td>
<td>bleeding time, coagulation screen</td>
</tr>
<tr>
<td>Disease activity</td>
<td>Serum beta2 microglobulin</td>
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</tbody>
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**Radiography**

Classic punched out lesions
Osteopenia or collapse vertebra
Skeletal survey: Skull, Chest, Pelvis
Spine: vertebral fracture
Treatment
1. Melphalan
2. Prednisone
   Combination chemotherapy shown to prolong overall survival
3. Radiation is very effective for localized lesions.
4. Surgical intervention: Prophylactic fixation in impending
   Spine with instability neurologic: Decompression

Monitoring
Total count and platelets
ESR
Serial serum protein electrophoresis or test for quantitative Ig
Serum B microglobulin
X ray: Resolution of the skeletal disease.

HYCALCEMIA

Common in Myeloma
Secondaries in the bone

Clinical
Confusion. Lethargy, Coma
Heart stops in systole

Treatment
Rehydrate
Prednisolone 30-40mg daily
Frusemide
Role of Pamidronate IV [Biphosphonates]

The overall prognosis for patients with myeloma patients tend to die within 3 years

Chemotherapy improves the median survival time to 3 years in the 50% to 60% who respond to treatment.

**Solitary plasmacytoma**

Lab investigations are normal
Younger than multiple myeloma
Solitary bone lesion
When Spine: Paraplegia is more common than myeloma
70% of solitary, will develop multiple myeloma and will usually die within 5 years
Radiation or en-bloc excision
Need close long follow up.