Malignant bone tumors

Incidence

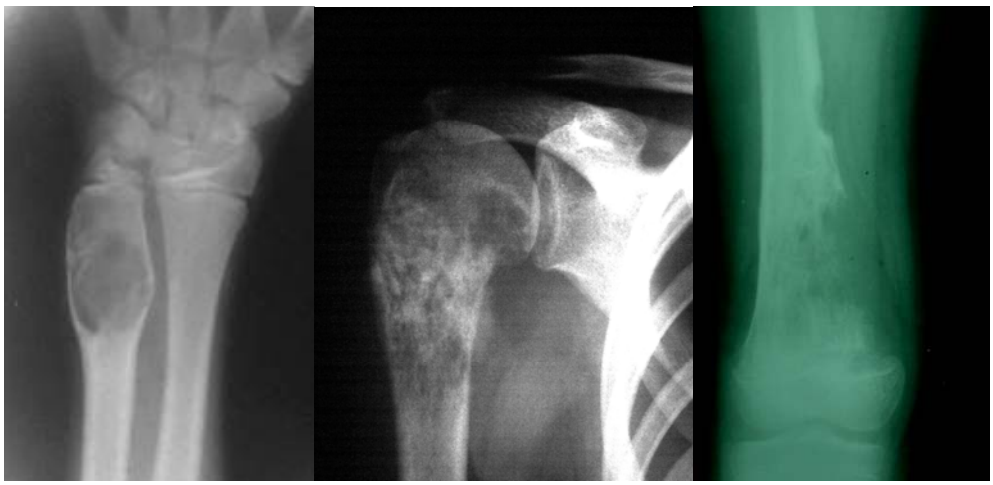
Myeloma	45%
Osteosarcoma	24%
Chondrosarcoma	12%
Lyphoma	8%
Ewing's Sarcoma	7%

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

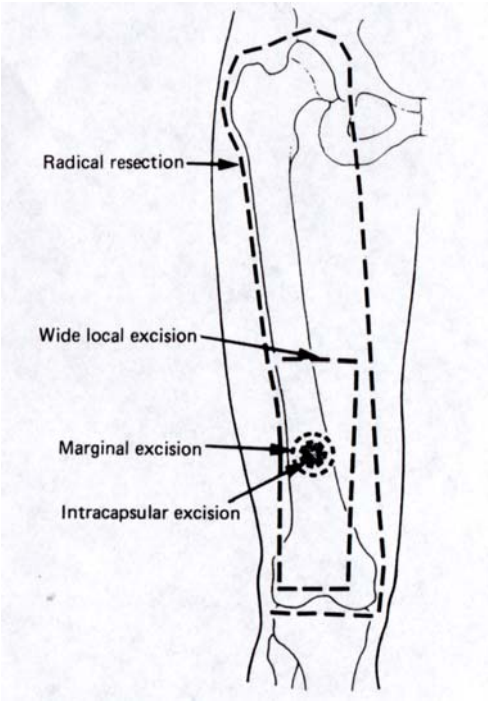
Stage I: Low grade sarcomas

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

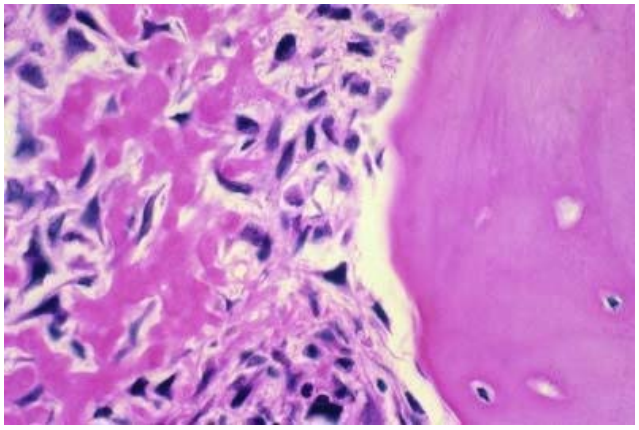
Common sites : Around knee 50%
Proximal shoulder

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.

X ray

Wider zone of transition

Permeative or Moth eaten

Destruction of the cortex

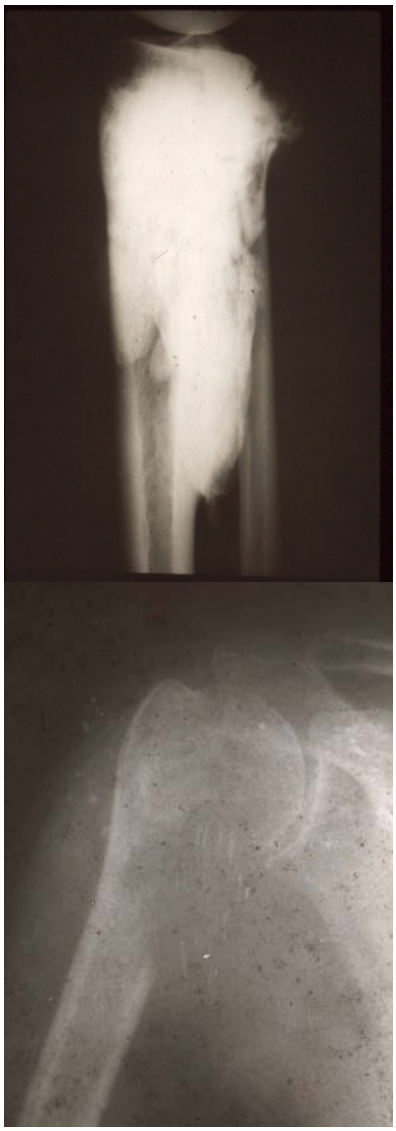
Sclerosis or lysis or mixed

Codmans triangle

Sunburst appearance

Parosteal: Distal femur; Pasted on

Appearance of Parosteal, Periosteal and Telangectatic



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

Chemotherapy

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

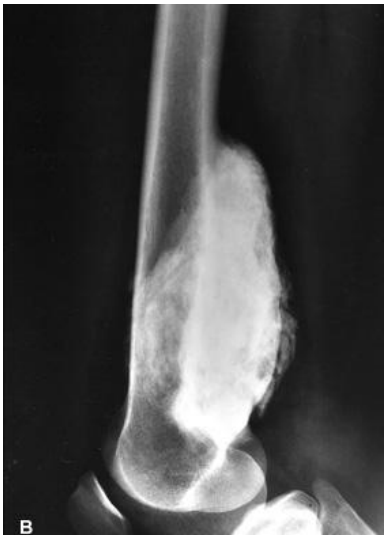
4% all Osteosarcoma

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

II 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

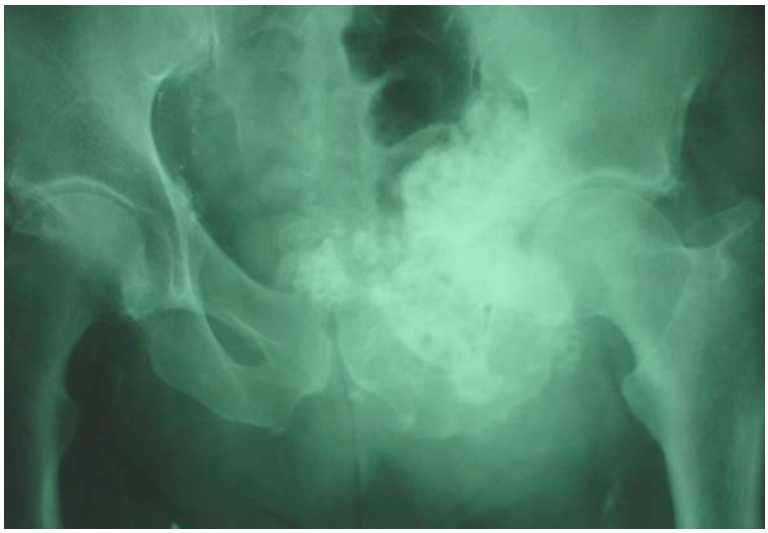
These secondary sarcomas are almost always histologically low-grade

Site: Pelvis is the commonest site

Femur, Humerus, scapula, ribs

Painful encondroma in a long bone:

Suspect maligancy



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

X Ray

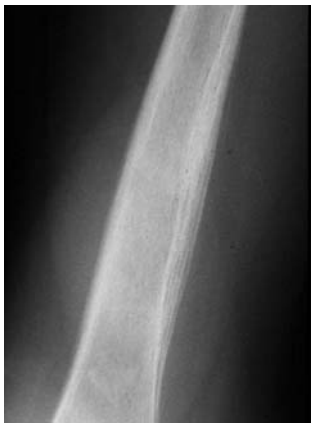
Lytic,

Moth eaten or permeative

Laminated periosteal reaction

(Onion peel) or sub burst

appearance



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 -80years

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

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

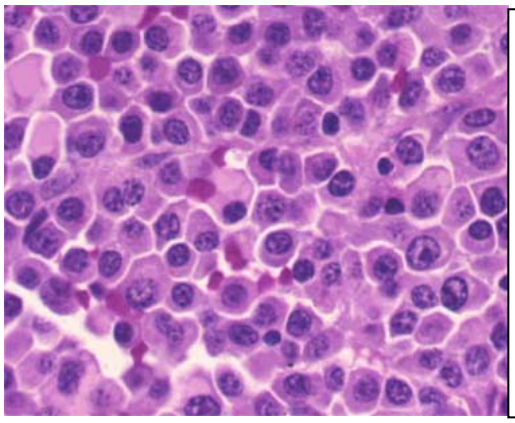
Radiography

Classic punched out lesions

Osteopenia or collapse vertebra

Skeletal survey: Skull, Chest, Pelvis

Spine: vertebral fracture



Sheets of cells

Eccentric nucleus

Cartwheel appearance

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.