Bone Tumours benign and malignant: A synopsis for medical stud

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Table of Contents

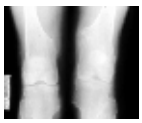
Bone Tumours

Department of Orthopaedic Surgery - Stellenbosch University

Bone tumour index page

Menu:

To
begin
click
Button
below



Benign Tumour Video

Malignant tumour video

Printable Version (All above pages, HTML)

PDF version (Printable booklet)

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Benign Tumours

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Page contents

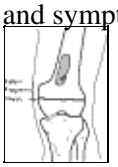
Differences between benign and malignant tumours

Benign tumors are unable to metastasize and generally grow slowly.

Benign	Malignant
Well defined margin	Poorly defined margin
Slow growth	Rapid growth
No metastases	Distant metastases

Simple bone cyst

This is a common bone tumor in children and may lead to pathological fractures. If it is painful, the cause is usually mechanical stress (pre fracture stage) and symptomatic



lesions should be treated. Cysts in the region of the hip are particularly prone to fracture.

Characteristics Simple bone cyst

- Children
- Mildly Expansile
- Filled with serous fluid
- Migrate to diaphysis
- Path. fractures common

Treatment

Symptomatic or cysts in high stress areas such as the femur neck need treatment The cyst needs to be decompressed. Methods such as curettage and bone graft, injection with cortisone and simple drilling with K wires are all effective. Fractures through a cyst are treated conservatively (plaster cast). In high stress areas such as the hip, internal fixation is needed. The cyst usually resolves after the fracture unites.

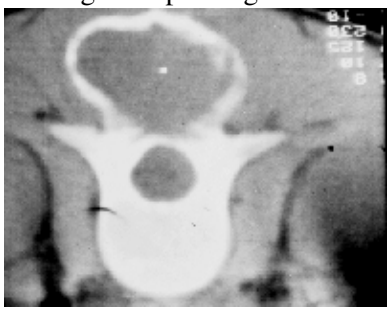
If a cyst recurs a biopsy is needed, as the diagnosis may be a more aggressive entity such as the aneurismal bone cyst.

Aneurysmal Bone Cyst

Is a cystic expansile bone tumor seen in the first and second decades. It may appear in any bone.

Aneurysmal bone cyst of the clavicle. Expansile bone cyst, loculated

The cyst is filled with blood. 30% are associated or contain elements of another 
primary lesion such as a GCT, chondroblastoma, fibrous dysplasia, chondro myxoid fibroma, EG, simple cyst, osteoblastoma, non ossifying fibroma The ABC can become large, and can also be the cause of a pathological fracture. Some ABC 's can be fast growing and locally aggressive. Biopsy and histological diagnosis is mandatory if ABC is suspected. Treatment is by curettage and packing with bone chips.



Commonly seen in the vertebrae, the ABC usually involves the posterior elements.

Enchondroma

A benign cartilaginous lesion appearing in adult life, seen often in short tubular bones e.g. the hand. The lesions are usually single but may be multiple.

Characteristics

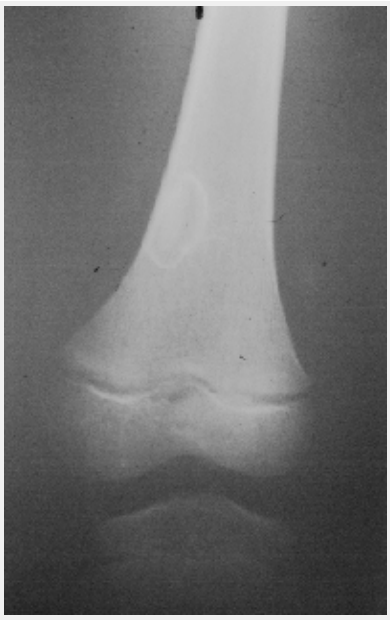
- Age: 2nd - 5th decade
- Cartilage
- Short cylindrical bones,often in hand
- Problem: ? Low Grade Chondrosarcoma

X-rays show scalloped erosions on endosteal surface. There may be flecks of calcification. In *multiple enchondromatosis* (Ollier's disease) there may be associated deformities, such as genu varus or valgus. In the systemic form (Ollier's), there is a high (10 -20%) incidence of malignant transformation.

Multiple enchondromata or Ollier's disease

Treatment is by curettage and bone grafting. Because many of these may be low grade malignant chondrosarcoma (which is difficult to distinguish histologically from benign enchondroma) additional techniques, such as cryosurgery are also added to make sure the residual cells are killed.

Non Ossifying Fibroma



Fibrous cortical defect
X-ray features
Margin well defined, sometimes scalloped, and often sclerosed

This is also known as a fibrous cortical defect or a metaphyseal fibrous defect. Occurs in the metaphysis in the first two decades of life. Histologically it consists of fibrous tissue.It is often  primary lesion such as a GCT, chondroblastoma, fibrous dysplasia, asymptomatic and found incidentally on X-rays Large lesions may cause a pathological fracture.

Treatment is only required in a symptomatic lesion. The defect is curetted and packed with bone.  If found incidentally on x rays and the lesion is small, *leave it alone*. Larger lesions are rare, but if > 50% of diameter of the bone consider surgery, as pathological fractures can take place.

Fibrous dysplasia


Histologically also fibrous tissue. Is a more severe and often systemic form non ossifying fibroma. It begins in childhood and affects one (monostotic) or many bones. It may cause deformities such as coxa vara and facial deformities.

X-ray features

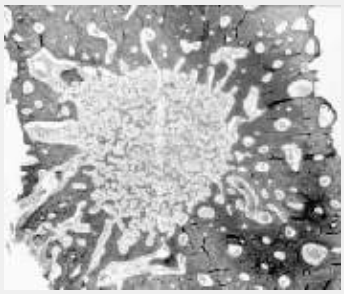
Radio lucent or opaque lesions may be lobular or scalloped. The cortex is eroded and expanded.

Osteoid Osteoma

Osteoid osteoma presents as a sclerotic cortical lesion. It is painful and the pain is relieved by asprin

There is an oval lytic centre known as the nidus. It is this center that produces benign osteoid.

Osteoid Osteoma of femoral neck. Notice the sclerosis of the neck and the central nidus



Histology - the nidus produces osteoid it may contain giant cells

Differential diagnosis of a sclerotic cortical lesion in a child

- Osteoid osteoma
- Stress fracture
- Chronic osteomyelitis
- Malignant tumor e.g. osteosarcoma

Management
To locate the *nidus* computer tomogrammes are helpful. A technetium scan will show a hot spot. Excision of the nidus will cure the pain. An *en block* excision is done.

Osteochondroma

The osteochondroma is common and presents as a bony outgrowth near an epiphysis.

Multiple osteochondromas

Lesions be single or multiple. It consists of a bony outgrowth with a cartilage cap. The lesion may have a narrow neck (pedunculated) or may have a broad base (sessile) Growth stops with skeletal growth. If the lesion enlarges the cartilage becomes thickened in adult life consider malignant change in your diagnosis. Malignant change to osteosarcoma or chondrosarcoma may occur in up to 10% of multiple osteochondromas.

Causes of pain in an Osteochondroma

- Mechanical eg ileotibial band impingement
- Fracture
- Malignant change

Management Not all lesions require excision. Excise symptomatic lesions and do histology. In multiple osteochondroatosis yearly technetium scans are done and hot lesions are excised. All symptomatic lesions must be excised.

Giant Cell Tumours

The Giant Cell Tumor grows in the epiphysis of adults and undermines the mechanical integrity of the joint.

Giant Cell Tumor

Treatment
The GCT consists of giant cells in a spindle cell stroma. The often breaks through the bone and invades the soft tissue. 
primary lesion such as a GCT, chondroblastoma, fibrous dysplasia, On rare occasions it metastasizes to the lungs. The tumor is curetted and packed with bone. This may fail if it is a major joint and block excision may be required with arthrodesis or joint replacement.

Chondroblastoma

The chondroblastoma has a predilection for the epiphysis and is almost always

found here. It does not stretch to the articular surface as the GCT does.

Chondroblastoma

- always in the epiphysis, shows areas of calcification

Peak age incidence 10 to 20 yrs. Almost never undergo malignant transformation. 
primary lesion such as a GCT, chondroblastoma, fibrous dysplasia,

Treatment

Curettage and bone graft.

Chondromyxiod Fibroma

The Chondromyxoid Fibroma is composed of myxoid or primitive cartilage and fibrous tissue. It presents in the second decade or later.

Chondromyxoid fibroma
Eccentrically situated, may stretch to, but not cross, the growth plate

It has a very sclerotic endosteal border. 
primary lesion such as a GCT, chondroblastoma, fibrous dysplasia,

Treatment
Extra capsular marginal excision. Recurrence is rare.

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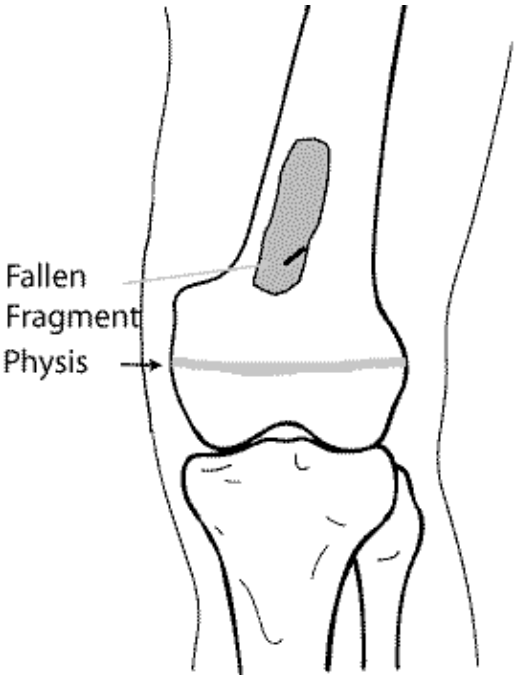
Cystic benign bone tumours

Positions in the bone

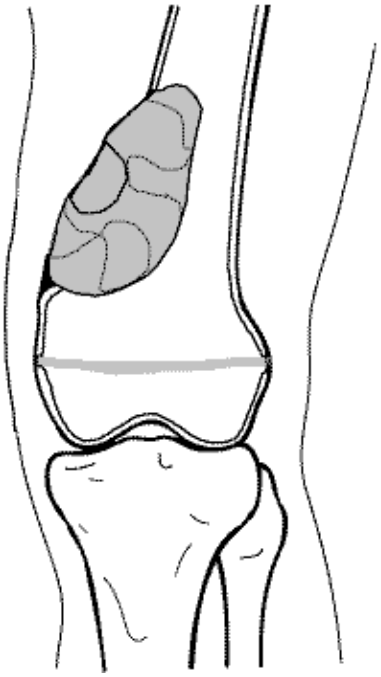


Simple bone Cyst

Eccentric, non expansile. One compartment. May see "fallen fragment " sign. Starts at physis and migrates away with growth. Has a long axis aligned to the bone shaft. Never penetrates cortex to extend into soft tissues.

Aneurysmal Bone Cyst

Usually metaphyseal, may extend into epiphysis Blown out appearance with thinned outer cortex. Multi compartmental.

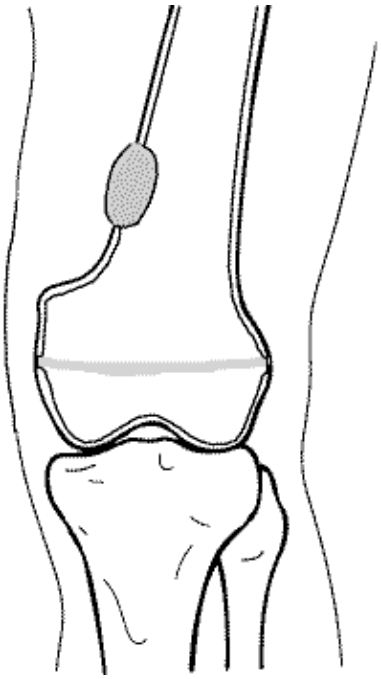


Giant Cell Tumour

Seen in adults. Extends to the subchondral bone of the joint margin. Crosses from metaphysis into epiphyseal region. May be expansile. Outer border may be thinned or absent (may break into soft tissue).

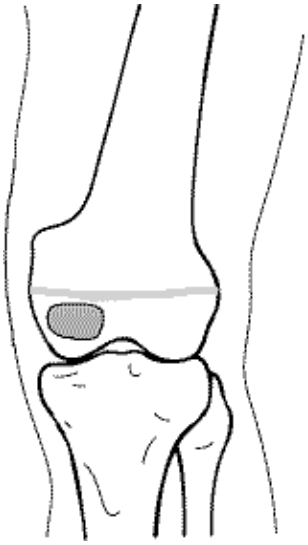
Non Ossifying Fibroma / Fibrous Cortical Defect

Eccentric and attached to cortex. Commonly in metaphysis, but may be seen in shaft. Well defined smooth or scalloped margins.



Chondromyxiod Fibroma

Eccentrically situated lytic lesion with well defined margins in the metaphysis of the lower extremity. The lesion usually has a sclerotic margin of bone and a lobulated contour. Ridges and grooves that appear in the margins secondary to scalloping falsely appear to be trabeculae



Chondroblastoma

Characteristically involves the epiphysis.Well demarcated oval or round radiolucency. Has a thin sclerotic bony margin. Fine calcifications, either punctate or in rings, may be visible.

Malignant Tumours

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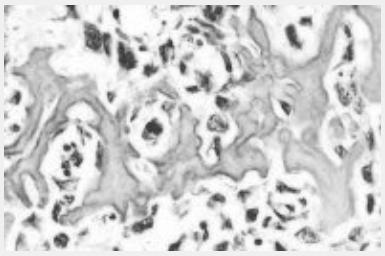
Page Contents

Osteosarcoma

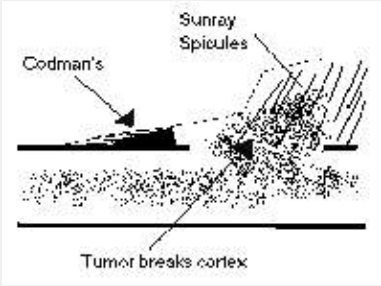
Osteosarcoma is a primary malignancy of bone. The malignant cells produce osteoid.  Histologically the tumor is composed of malignant osteoblasts which produce osteoid Most occur in the metaphysis of long bones especially about he knee. Age - 10 to 20 years. If seen later in life consider a *secondary* malignancy (to Paget's or post irradiation) It metastases to the lungs and to other bones.

Prognosis

Poor in South Africa (20% 5 yr. survival, because of late presentation). International experience is towards a 60% survival.



Histology of an osteosarcoma. Note the malignant osteoid (pink trabeculae)



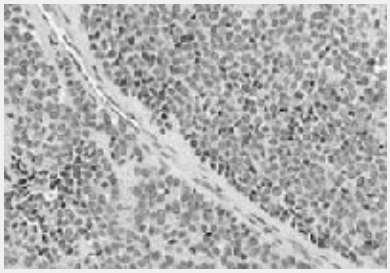
X-ray features
Codman's triangle, *Sunray* spicules

Ewing's Sarcoma

Ewing's is a small cell tumor seen in the 10 to 25 yr. age group. 60% occur in the long bones, but the scapula and pelvis are often affected.

Ewing's

The tumor lifts the periosteum to produce the typical onion skin appearance



Ewing's

Small cells of uniform size. On electron microscopy the cells contain glycogen granules

Ewing's is one of the few tumors that frequently originate in the shaft of long bones. 50% originate in the diaphysis. It is an osteolytic tumor and has a large soft tissue component. It may mimic chronic osteomyelitis and even produce a raised body temperature and ESR as well as white cell count. Histologically the tumor consists of monotonous sheets of small round cells.

Prognosis:
Poor 30% have lung or bony metastases at time of presentation.

Myeloma

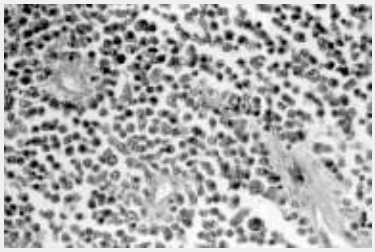
Myeloma is a common primary tumour from the 5th decade onwards. Myeloma presents with lytic bone lesions which commonly lead to pathological fracture.
Site
Common in any bone containing red marrow especially flat bones eg pelvis as well as vertebra. Consider the diagnosis in a vertebral fracture in the elderly. Typically the vertebra is flattened the so called *"wafer"* vertebra. If there is systemic involvement the skull x-ray may show "*punched out*" lytic lesions.
Clinically
Affects bone containing red marrow (skull, ribs, vertebrae, sternum, pelvis)
Weakness, bone pain and pathological fractures
Backache is common and may cause root pain and occasionally paraplegia
Anemia, generalised malaise and cachexia

Effects of Myeloma	**Diagnosis Myeloma**
• Local bone destruction by the tumour	• ESR usually >100 mm/hr • Serum Electrophoresis • Urine Bence Jones Protein

• High plasma protein concentration • Renal effects of abnormal plasma proteins ◊ Renal Failure ◊ Gout	• Serum immunoelectrophoresis • Bone marrow biopsy • Biopsy - only occasionally needed

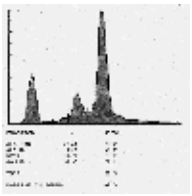
Histology

The tumor is composed of abnormal plasma cells. If the tumor is localised to one bone it is known as a *plasmacytoma* Systemic involvement is known as *multiple myeloma* A bone marrow biopsy must be done from the pelvic rim to determine the extent of spread.



Histology Myeloma

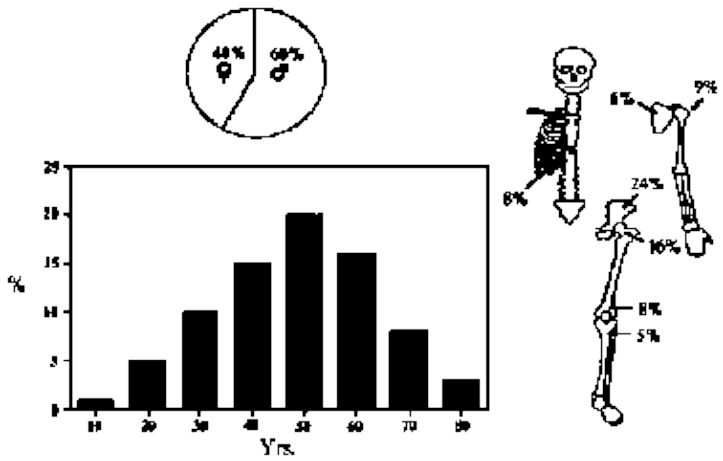
Consists of plasma cells



Monoclonal peaks are seen in multiple myeloma. Peaks such as the one on the right do not correspond to the usual Alpha and Beta peaks

Chondrosarcoma

Chondrosarcoma usually presents after 6th decade.
Characteristically chondrosarcoma is slow growing and seen proximally in the skeleton e.g. prox. humerus and pelvis.



Chondrosarcoma arising from an Osteochondroma.
The patient has multiple enchondromas. The lesion on the left ilium has developed an indistinct border. Compare this to the enchondromas on the proximal femurs.

Click on the image for other views.

Chondrosarcoma often arises secondary to other tumours.

Secondary Chondrosarcoma

- Osteochondromata esp. Multiple
- Enchondroma esp. Ollier's (25%)
- Mafucci (100%)
- Chondroblastoma
- Chondromyxiod fibroma
- Synovial chondromatosis

Treatment

Surgery alone is the only hope of cure with this tumour. Chondrosarcoma is unresponsive to irradiation and chemotherapy. Block excision is recommended.

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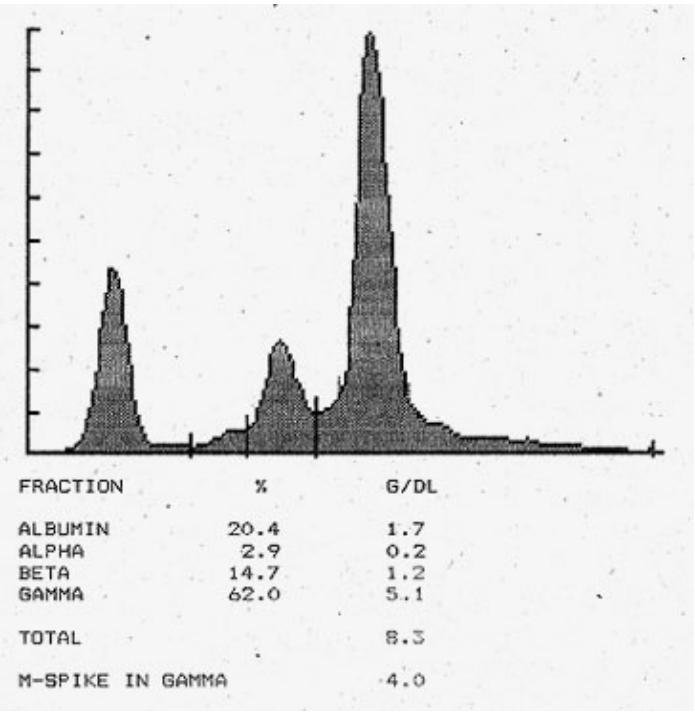
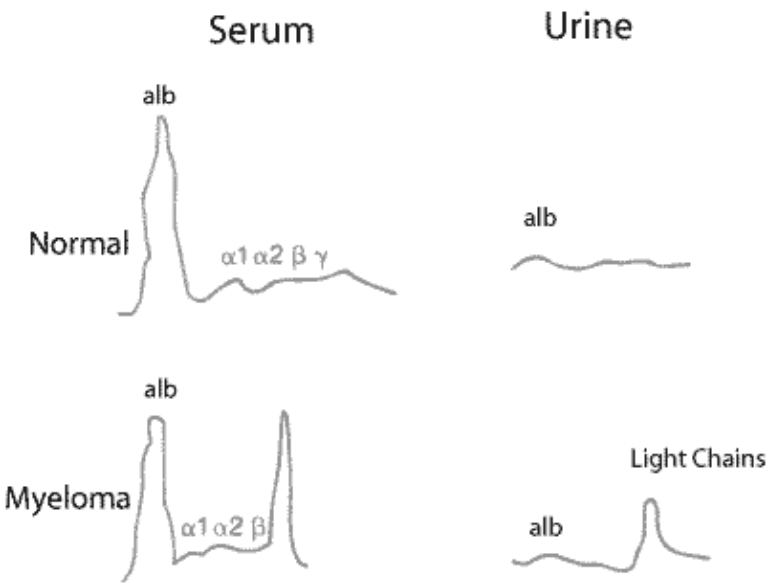
Electrophoresis Home

Diagnosis of Myeloma: Electrophoresis



FRACTION	%	G/DL
ALBUMIN	20.4	1.7
ALPHA	2.9	0.2
BETA	14.7	1.2
GAMMA	62.0	5.1
TOTAL		8.3
M-SPIKE IN GAMMA		4.0



Electrophoretic patterns in urine and serum. The upper panel represents normal values. Below a "M peak" from the abnormal plasma proteins is added. Light chains may spill over to the urine as "Bence Jones Protein"

Metastatic Tumours

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Metastatic bone tumours

Metastases are the most common bone tumours in older patients. They will present to the orthopaedic surgeon with pain, either because of an actual or threatened pathological fracture, or will present with a lytic or sclerotic bone lesion. The patient may have had a primary diagnosed years beforehand, and a history of previous surgery or investigations must be extracted as such information is not always volunteered.

Tumour	Lytic / Blastic
Breast	Lytic, rarely sclerotic
Prostate	Sclerotic
Lung	Lytic

Thyroid	Lytic, expansile
Renal	Lytic

Q: *When does a patient with a known metastasis require prophylactic fixation?*

A: If the lesion is **painful** it is likely to be at a *pre fracture* stage. If the pain, in a limb with a metastasis, that increases with weight bearing is an indication for fixation. A lesion that is bigger than 50% of the diameter of the bone will also need to be fixed.

Spinal metastases
Click for further discussion

Once a pathological bone has fractured conservative treatment will fail and the bone needs ORIF. After fixation all the bone needs radiotherapy to kill residual cancer cells.

Mirel's Scoring System			
Points	**1**	**2**	**3**
Site	Upper Limb	Lower Limb	Peri-trochanteric
Pain	Mild	Medium	Severe
Lesion	Blastic	Mixed	Lytic
Size			

Management of Tumours

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Basic Investigations

These are the basic investigations needed with most suspected primary tumours.

Investigation	Reason
ESR	High with sepsis and Myeloma, normal or moderate with most tumors
Alk Phosphatase	High - if tumor rapidly replaces bone
FBC	Leucocytosis with sepsis, (sometimes with Ewing's)
Chest X-ray	Pulmonary mets. common in malignant bone tumors.
CT Scan Lung	More accurate than CXR - will detect smaller pulmonary mets.
X-rays of lesion	X-ray, features of bone tumor easily recognisable
Other imaging Modalities	**MRI** excellent for soft tissue component **CT scan** - good alternative especially with bone forming lesions eg osteosarcoma

Staging

In addition the tumor will have to be staged. These investigations can be done at the oncology center to which the patient is referred.

Staging Investigations

- Magnetic resonance of tumour site
- CT scan lungs - picks up smaller mets.
- TC Scan - for other bony mets.
- Biopsy

Once the local imaging is done a biopsy can be done and the histology of the tumour studied A Staging system such as that of Enneking is used to prognosticate and decide on management.

Treatment

Most malignant bone tumors require surgery and chemotherapy. Radiotherapy is reserved for irresectable or marginally excised tumours.
Chemotherapy is usually started about 6 weeks preoperatively once the diagnosis has been confirmed by histology.

Surgery

A wide surgical margin should be achieved.

Low grade **Chondrosarcoma**

After *block excision* and vascularised fibula graft

To achieve this aim, either a *en block excision* with arthrodesis, or custom mage prosthetic joint is required or an amputation is needed. The patient needs

postoperative chemotherapy too, in most cases.

Block excision can also be usedfor the treatment of benign, but aggressive tumours e.g. *giant cell tumour* about the knee.

Postoperative chemotherapy

Chemotherapy is also given postoperatively. Excised lesion is examined histologically to determine if the margins ate tumour free. Another factor looked at is the amount of tumour necrosis caused by the chemotherapy. If there was a good response the same regimen is given postoperatively. If the response was poor the chemotherapy agents are changed, with the hope that alternative drugs will be nore effective.

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