Primary Bone Lesions

Classification into 3 types

- Malignant bone tumours (sarcomas
- Benign bone tumours
- Lesions that simulate bone tumours (Tumour like conditions)Which may be reactive and miscellaneous

Common lesions that are not of mesenchymal origin include

- Metastatic bone disease
- Myeloma
- Lymphoma

	Classification of musculoskeletal tumours based on origin This includes tumour- like conditions
1. Bone origin	**Benign**;Osteoid Osteoma, Osteblastoma, **Malignant**;Osteosarcoma
2. Cartilage origin	**Benign**;Osteochondrama,Chondrobastoma,Chondromyxoid,Fibroma, Enchondroma , Periosteal Chondroma, **Malignant**;Chondrosarcoma
3. Fibrous origin	Nonossifying Fibroma, Fibrous Dysplasia, Osteofibrous Dysplasia, Desmoplastic Fibroma **Malignant**; Malignant Fibrous Histiocytoma
4. Miscellaneous	Unicameral bone cyst, Aneuysmal bone cyst,Giant cell tumour, Langerhans histiocytosis Ewing's Sarcoma
5.Musculoskeletal manifestations of leukemia	
6.Bone lymphomas	
7. Metastatic tumours	Neuroblastoma, Retinoblastoma, Hepatoblastoma, Lung, Renal Prostate Breast Thyroid

Bone Tumour Mimics

- 1) Soft tissue haematoma
- 2) Myositis Ossificans
- 3) Stress fractures
- 4) Tendon avulsion injuries
- 5) Infection – osteomyelitis especially chronic type
- 6) Gout

Lesions(Tumour like conditions)that are often confused with bone tumours

Simple bone cysts

1) Solitary bone cysts
2) Aneursymal bone cysts – benign expansile lesions composed of blood filled cystic spaces which are destructive but histologically benign. Patients typically present with a painful swelling in the metaphyseal region and neurological symptoms if the spine is involved

3) Fibrous Dysplasia which may involve one bone (Monostotic) or several (Polyostotic)

osteosarcoma

- Classical Intramedullary osteosarcoma
- Variants include
- Periosteal
- Parosteal

May be regarded as surface tumours

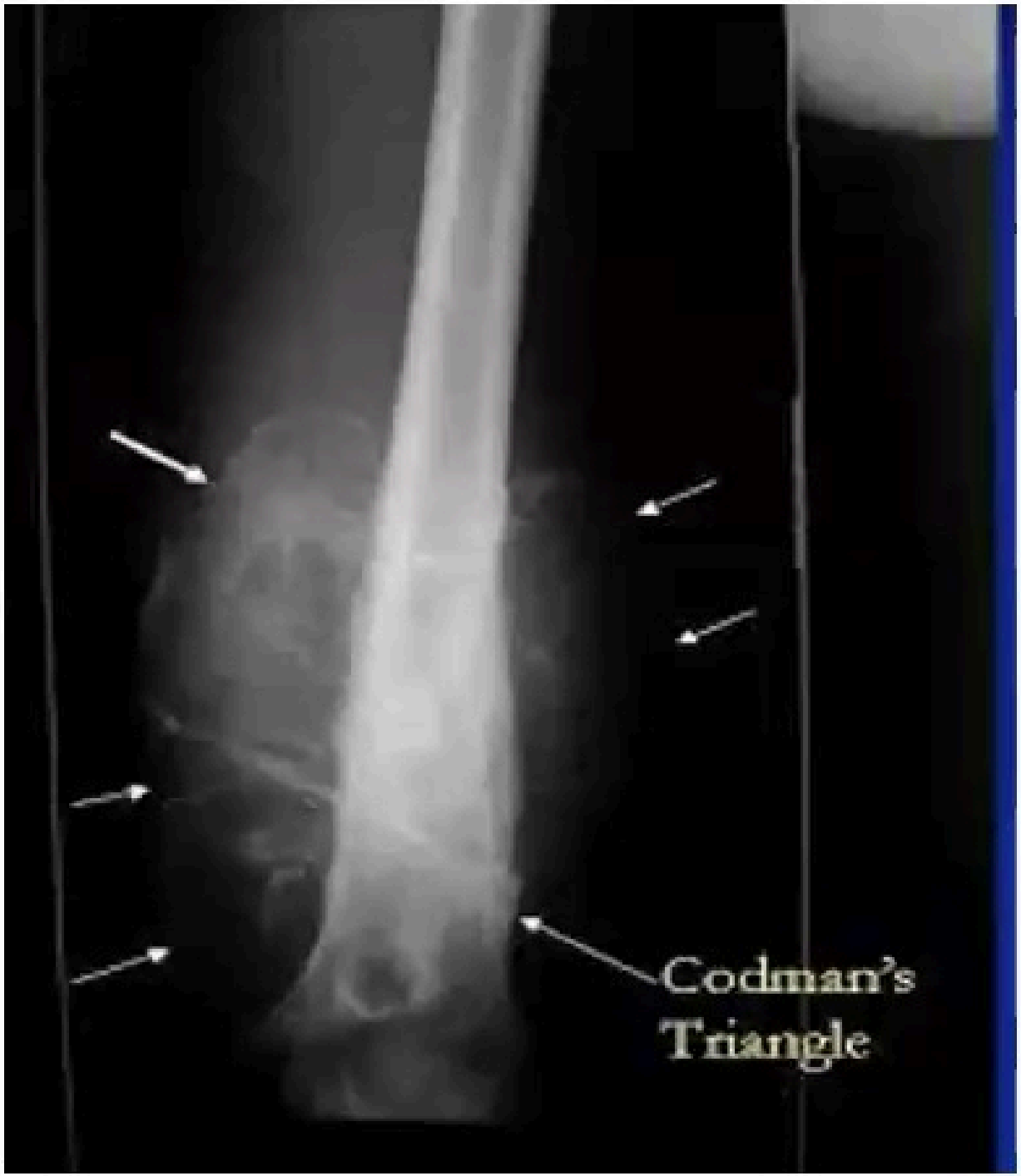
Osteosarcoma may be described as primary(denovo) or secondary (arising from abnormal bone)

Plane x-ray may be diagnostic. Look for codman triangle and sunburst appearance

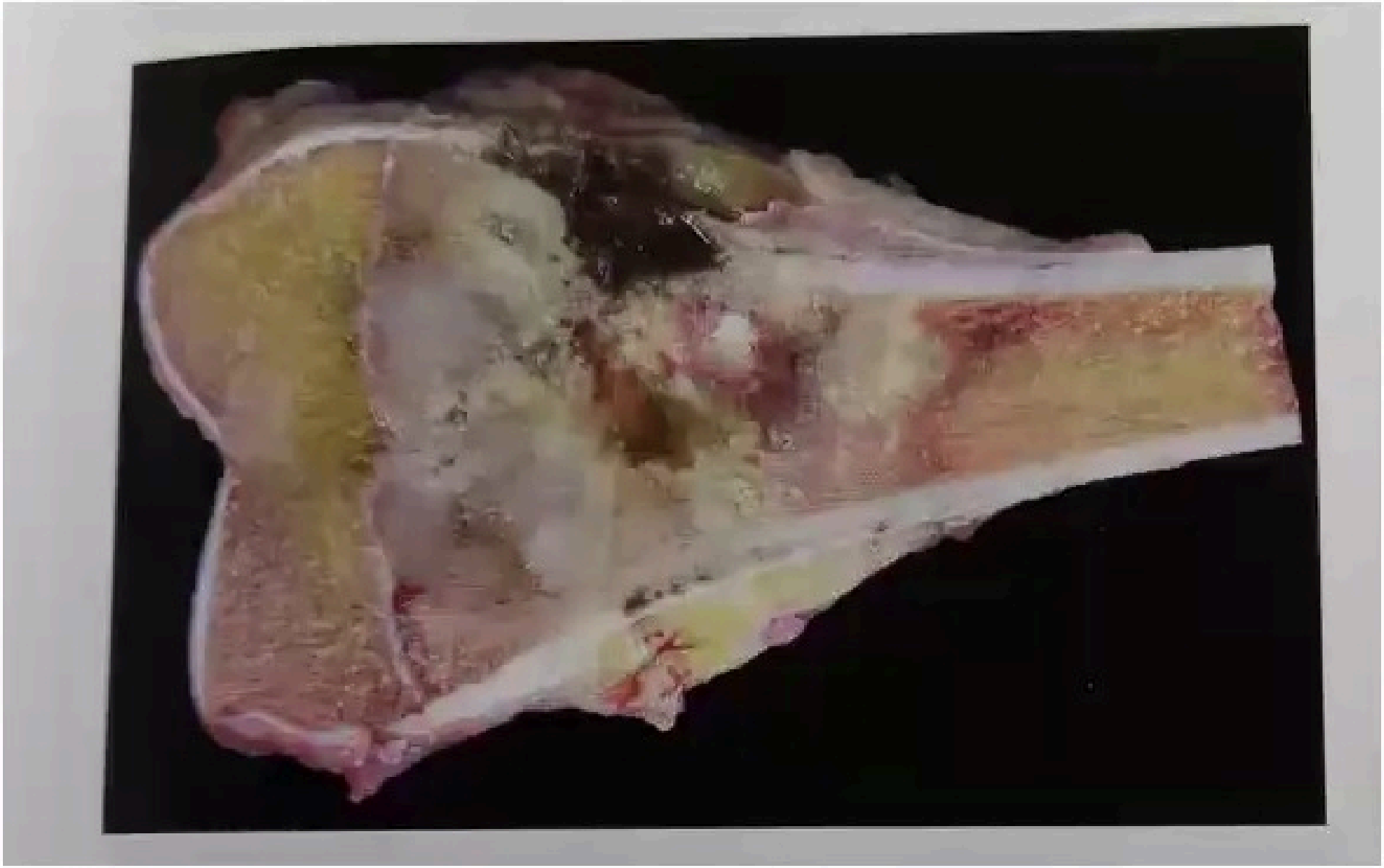
- The tumours may be variably mineralized and there may be a codmans triangle where new bone forms in response to periosteal elevation and a sunburst appearance where the periosteum does not have enough time to lay a new layer of bone and instead sharpeys fibers stretch perpendicular to the periosteum.

- Classically 80% are extracompartmental at presentation i.e extending through the cortex through what is regarded as cortical destruction not to be confused with cortical expansion and thining as seen in benign bone cysts
- A biopsy will show cells that demonstrate severe anaplasia and pleomorhism producing primitive woven bone and osteoid and display a permeative pattern of replacing host bone.

- Conventional osteosarcoma is subclassified according to predominant extracellular matrix evident in the tissue such as osteoblastic, chondroblastic and fibroblastic variants.



Osteogenic sarcoma



Osteosarcoma

- MRI of the whole bone will delineate the medullary and extraosseaus extent
- Nuclear medicine scintigraphy- skip lesions and distant bone tissue metastasis.
- CT chest is mandatory

Ewings Sarcoma

- Believed to arise from mesenchymal stem cells in the bone marrow
- Femur and tibial respectively are the commonest long bones but overall, pelvis accounts for the majority of cases
- Pain is the earliest symptom
- Radiography reveals aggressive permeative poorly defined lesion with lamellar “onion skin” appearance causing fusiform bone enlargement

- Local and distant staging include bone scintigraphy and chest CT.
- MRI – intraosseous and extraosseous extent
- Chemotherapy preoperatively results in tumour shrinkage and may be combined with adjuvant radiotherapy if surgical margins are poor

Evaluation

clinical presentation- history

Musculoskeletal pain- initially may be intermittent but becomes continuous and deep-seated.

Physical exam

Define the swelling in terms of site size, surface consistency etc most will be firm/hard consistency

Investigations

- Plane x-ray AP+Lat. May give a clue whether the lesion is benign, malignant or is a tumour like lesion.
- Can be useful in defining the effect of the lesion on the cortex(malignant tissue destroy the cortex whereas tumour like lesions will cause cortical thinning.)
- Useful in defining zone of transition between normal and abnormal tissue. MRI will enhance the diagnosis in terms of tumour that have spread in to the normal tissue.
- If x ray is normal do selected studies which will be guided by clinical suspicion

Laboratory studies cont

- Younger age group -blood count with diff, peripheral blood smear, ESR
- Older age group- blood count with differential, ESR, Ca P04, Pho4, serum or urine protein electrophoresis, tumour markers
- Peripheral blood film as well as bone marrow studies – may help complete investigations for lymphoreticular disorders (leukemias,lymphomas)

Biopsy

- Biopsy – performed after complete evaluation of the patient so that the pathologist and the surgeon already have a narrow working diagnosis. Points to remember orientation and location of the biopsy, meticulous haemostasis.
- Is the matrix having cartilage. Is there calcification or mineralization of osteoid. Cartilage calcification often appears stippled or show arcs or rings
- Osteoid mineralization is often cloud-like
- types of biopsy may be; frozen biopsy, needle biopsy versus open biopsy in bone tumours. Needle biopsy is almost always inadequate. Open biopsy is recommended in bone tumours.
- Benign bone tumours- should be excised in total.

Staging and grading

Value

- To develop evaluation strategies
- Planning treatment
- Predicting prognosis
- Most popular and useful for musculoskeletal lesions is Enneckking system

Grading

- Grading – a lesion is high grade if it has high potential for distant metastasis or low grade if with lower potential for metastasis
- Grading is difficult and requires a morphologic range and most grading systems are based on 3 grades
- Grade I – well differentiated
- Grade II – moderately differentiated
- Grade III – poorly differentiated

Cont.

Grading is difficult and is based on

- **Nuclear anaplasia** (degree of loss of structural differentiation)
- **Pleomorphism** (variation in size and shape of the cells in question)
- **Nuclear hyperchromasia** (nuclear staining)

Staging

The staging system of musculoskeletal tumour society (Enneking system) and the American joint commission on cancer (AJCC) are the most popular.

Enneking system based on knowing the

- Histologic grade of the lesion(Low grade or high grade) ,
- The anatomic features (intracompartmental or extra compartmental) and
- The absence (M0) or presence of metastasis (M1)

The recent edition of AJCC system has become more popular than Enneking among medical oncologists and many orthopaedic oncologists use it as well

Formulations of a differential diagnosis.

- Age of patient
- No of bone lesions – monostotic or polystotic
- Anatomic location
- Effect of lesion on bone
- Response of bone to the increased blood supply and adjacent elevation of periosteum in low grade malignancy causing periosteal elevation. (codman triangle is made of normal bone)

Treatment principles

- Neoadjuvant chemotherapy if limb salvage procedure is possible to shrink tumour and define better surgical margins
- Multiagent chemotherapy
- Above measures has improved survival rate as well as limb salvage survival.
- Other than resection margin and chemotherapy worse outcome tends to be seen in large tumours, age under 14 years, male gender, high ALP, local recurrence, p-glycoprotein expression and absent Er2 expression.

Treatment

- Management should consider whether lesion is benign or malignant.
- Tumour-like conditions are basically benign lesions. Methods of surgical excision in benign lesions and tumour-like conditions are similar whereas in malignant tumours and those doubtful lesions, an incision biopsy is done after which more radical surgery can be considered.
- This radical surgery can include limb saving procedures or a more radical surgical intervention like amputations.

Tx malignant tumors

- **Surgical procedures**

Limb salvage

Local control

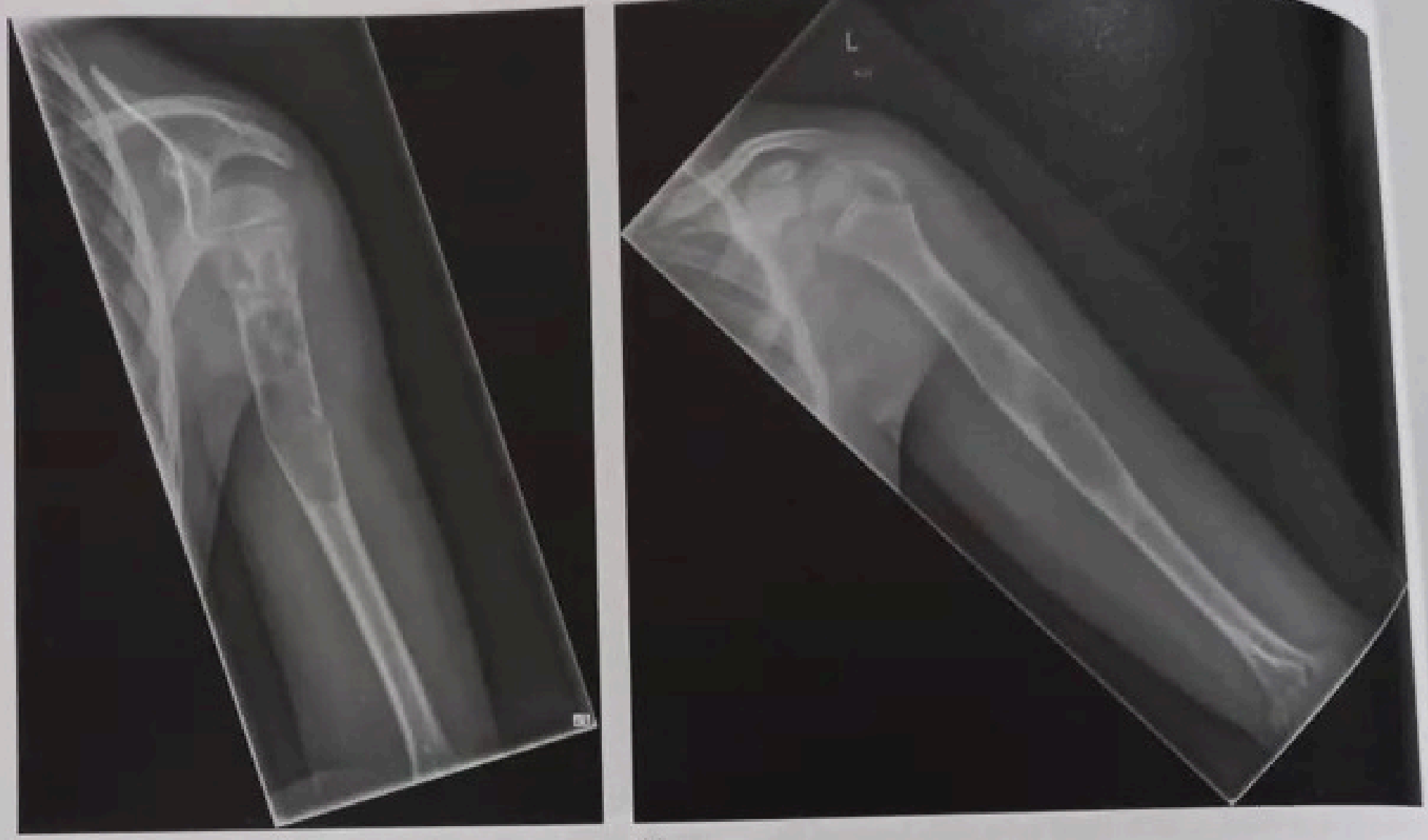
Saved limb functional

Surgical margins

- radical, wide margins, intralesional margin
- Radical – tumour and all surrounding muscles, ligaments and connective tissues are removed either by amputation or disarticulation.
- Above procedures are combined with Adjuvant chemotherapy.
- **neoadjuvant chemotherapy is for patients where limb salvage is possible, and that limb must be functional**

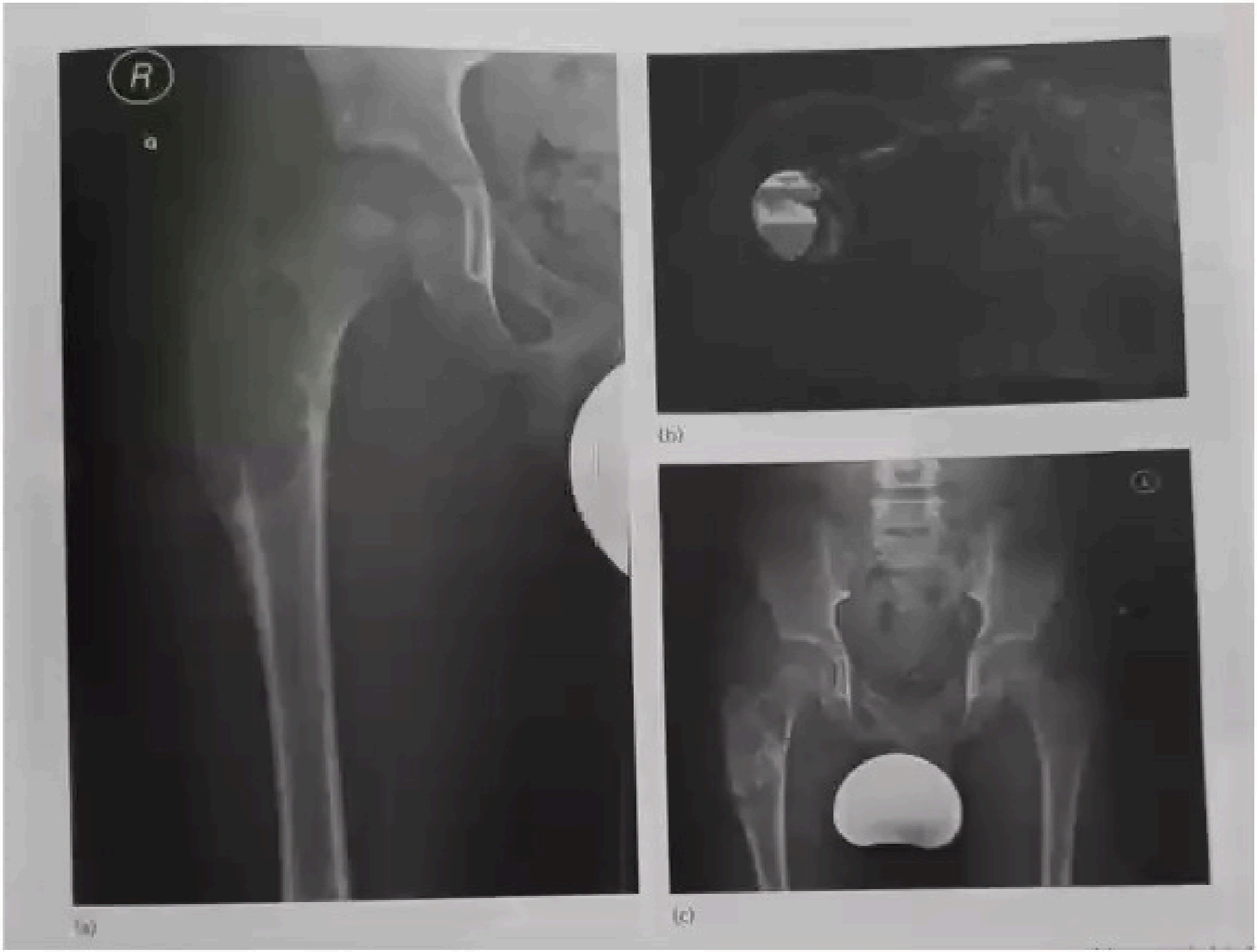
a) Multiagent chemotherapy after surgery for further local control and distant metasates

- Radiation therapy (External beam) Local control Ewings tumour, lymphoma, myeloma, metastatic bone disease
- Please note that radiotherapy is used for pain control in secondaries to the spine e.g prostate ca mets to the lumbosacral spine

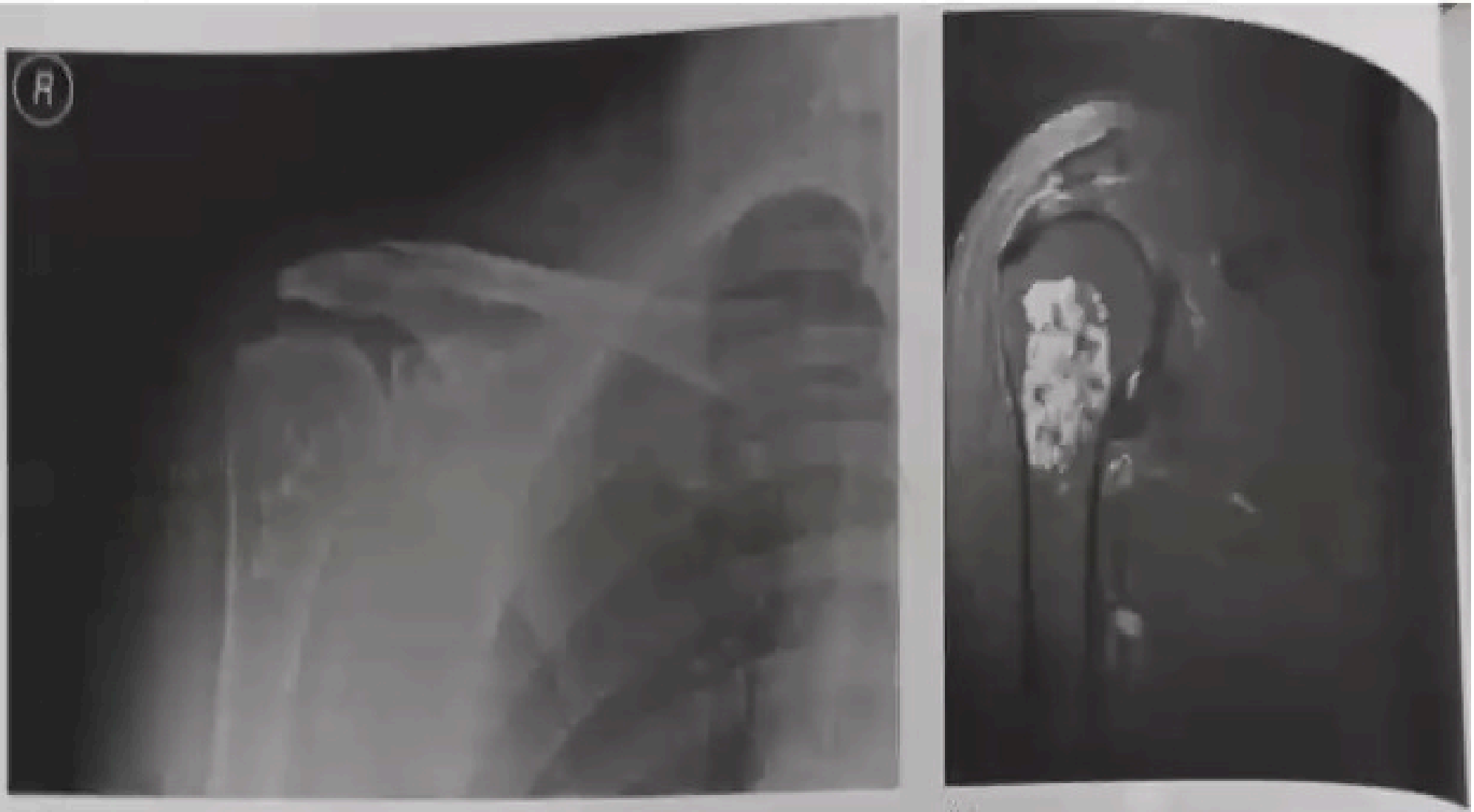


(a) (b)

Figure 9.20 Simple bone cyst (SBC) A 10-year-old boy with a spontaneous fracture through the left humerus. Plain radiographs (a) demonstrate an expansile, lytic lesion in the humeral metadiaphysis with a pathological fracture through the base of the lesion. The imaging features are in keeping with a simple bone cyst. The fracture was managed conservatively with a humeral brace and careful observation. Over time, the lesion consolidated and united (b).

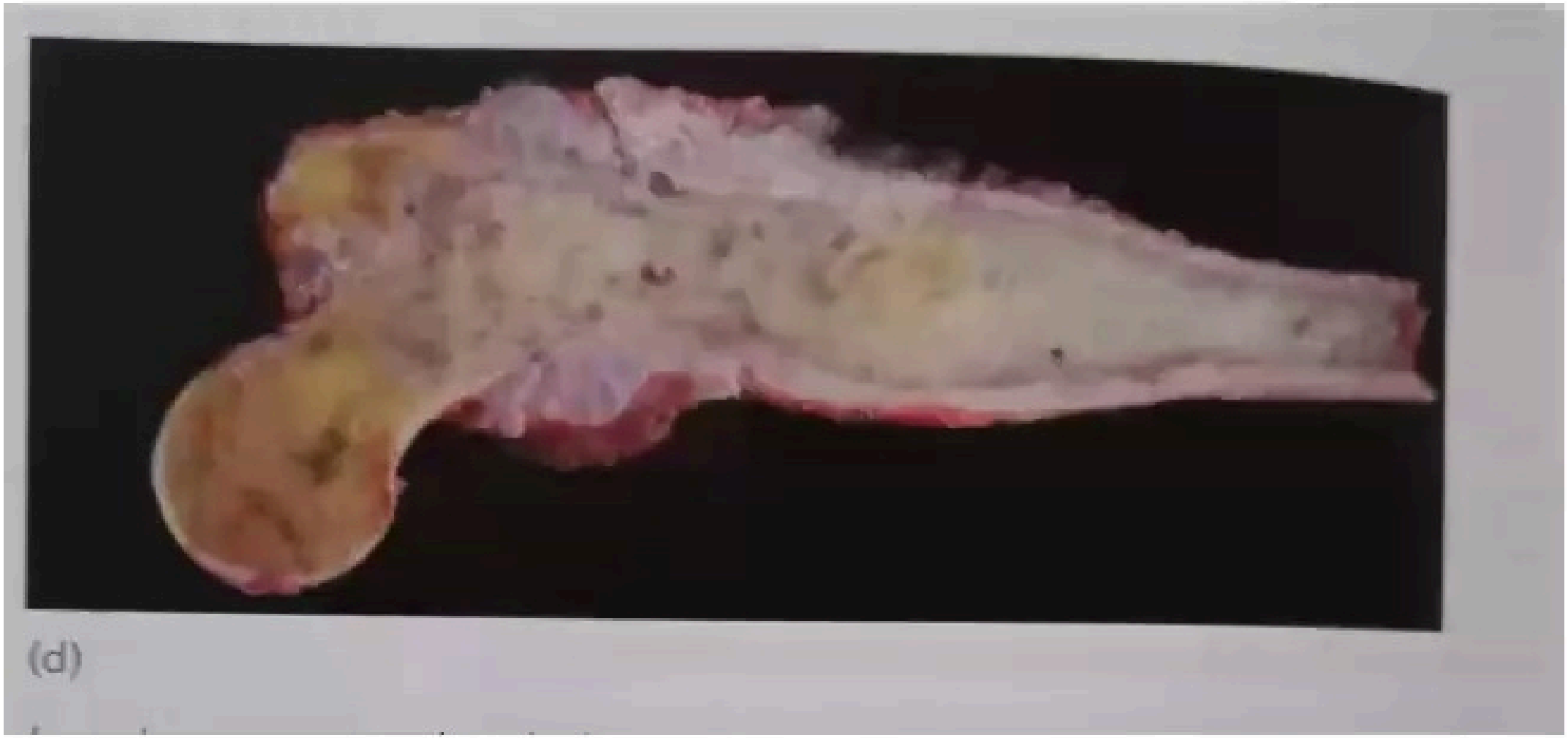


Aneurysmal bone cyst



(a) (b)

Figure 9.12 Enchondroma X-rays taken of a 65-year-old woman who presented with right shoulder pain demonstrate a 4 cm intraosseous chondroid lesion within the right proximal humerus (a). (b) Subsequent MR demonstrates typical features of an enchondroma with a mixed cartilaginous lesion with areas of calcification but no evidence of endosteal scalloping or permeation. The shoulder pain was attributed to a full-thickness rotator cuff tear.



Chondrosarcoma

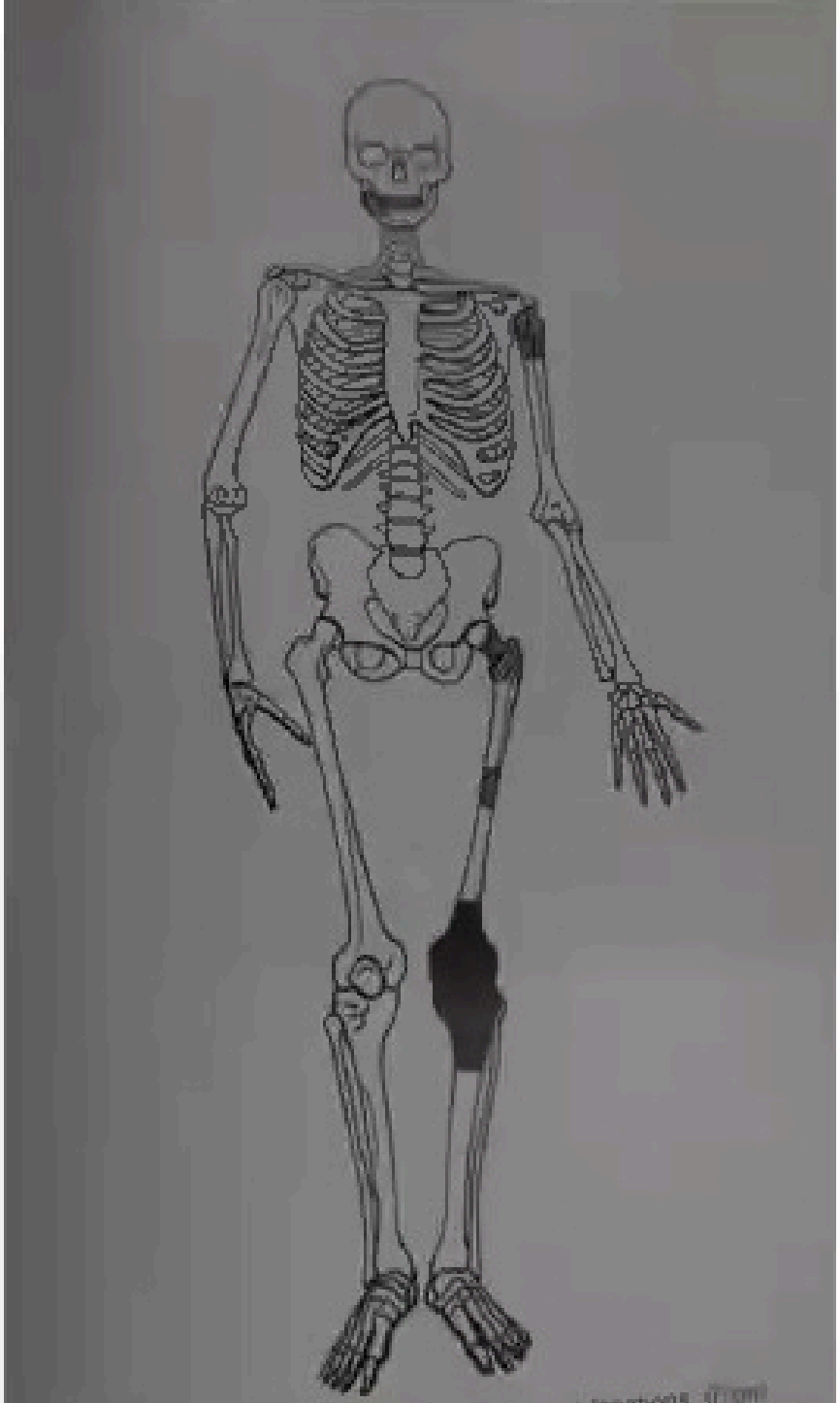
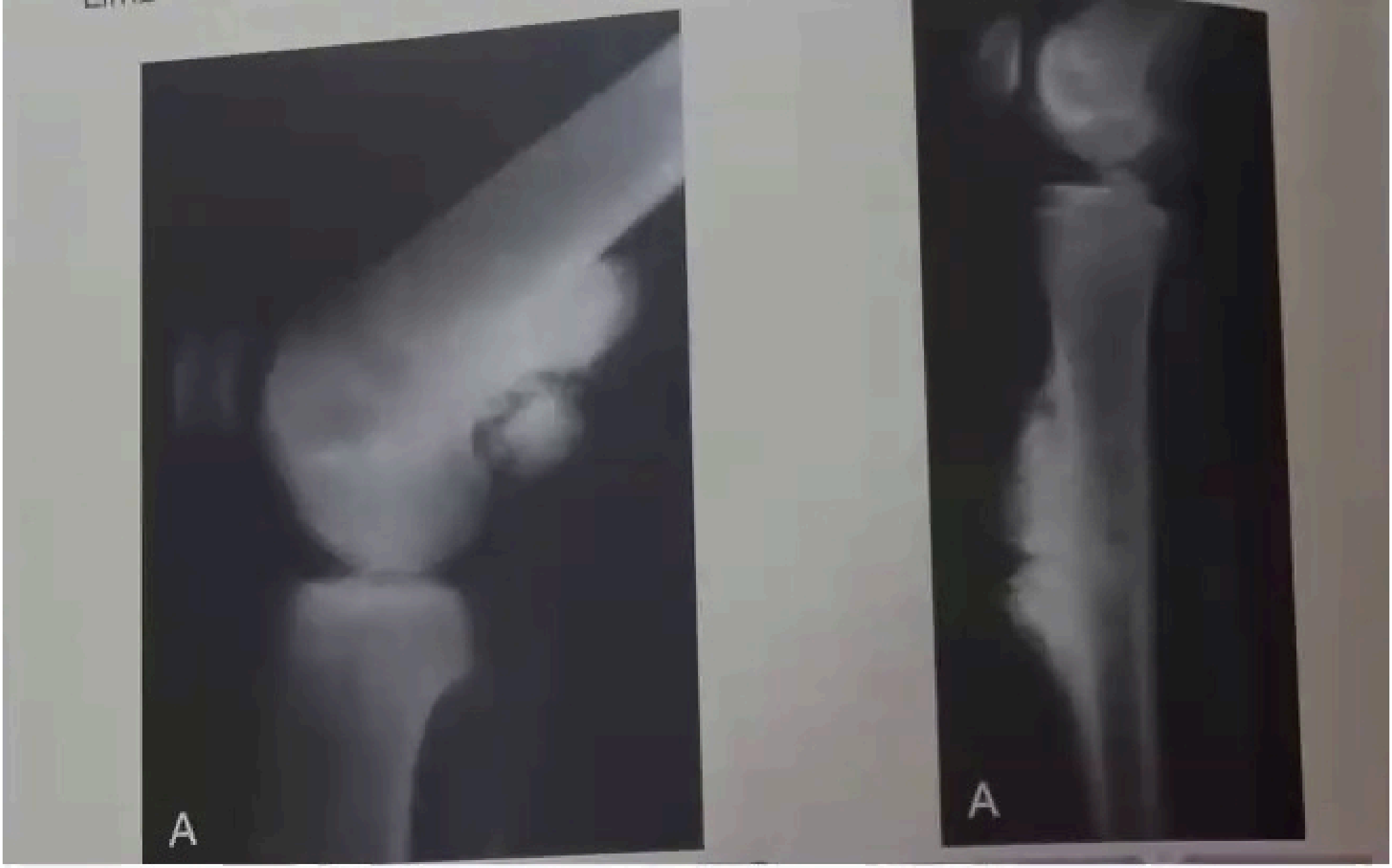
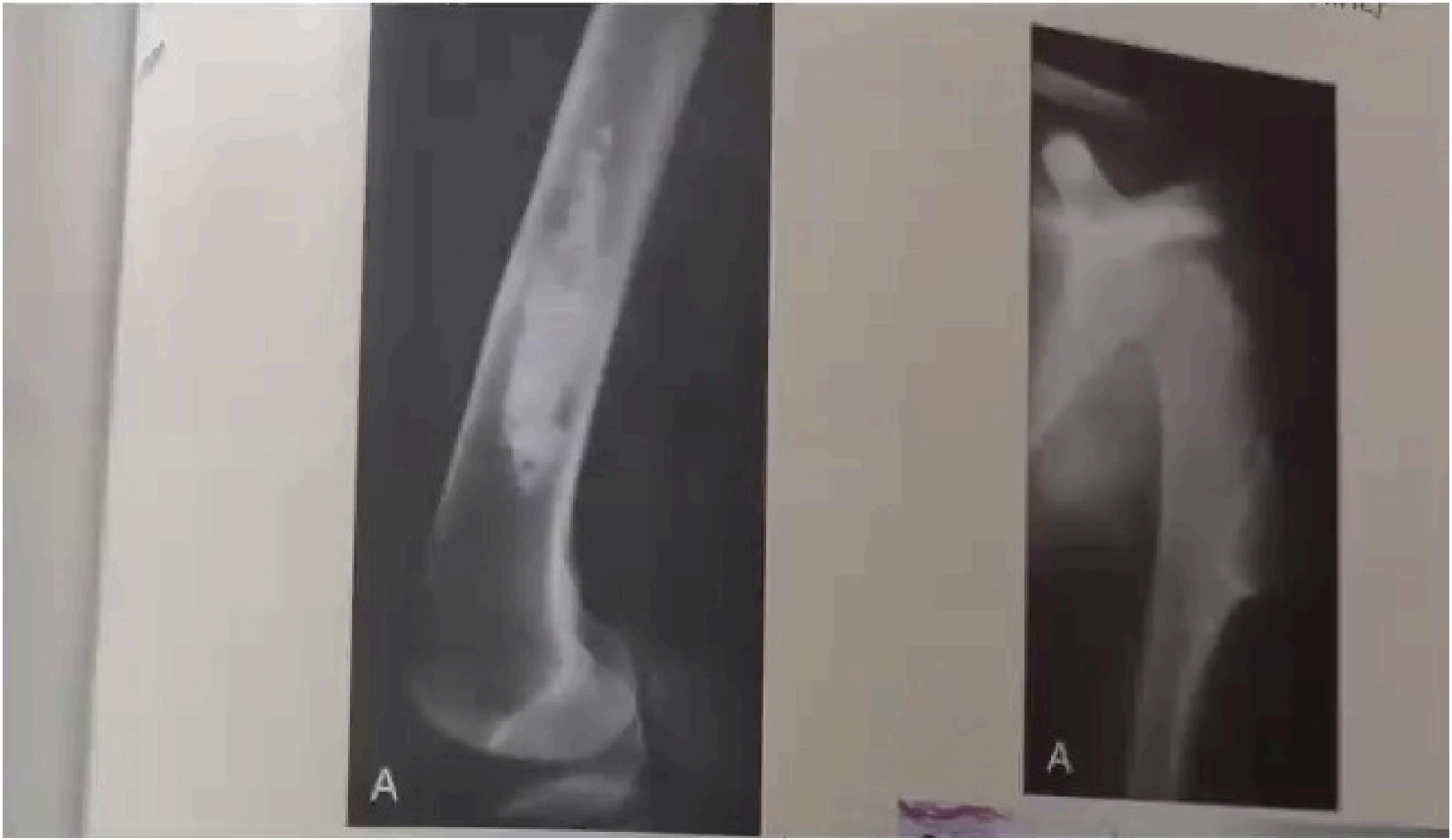


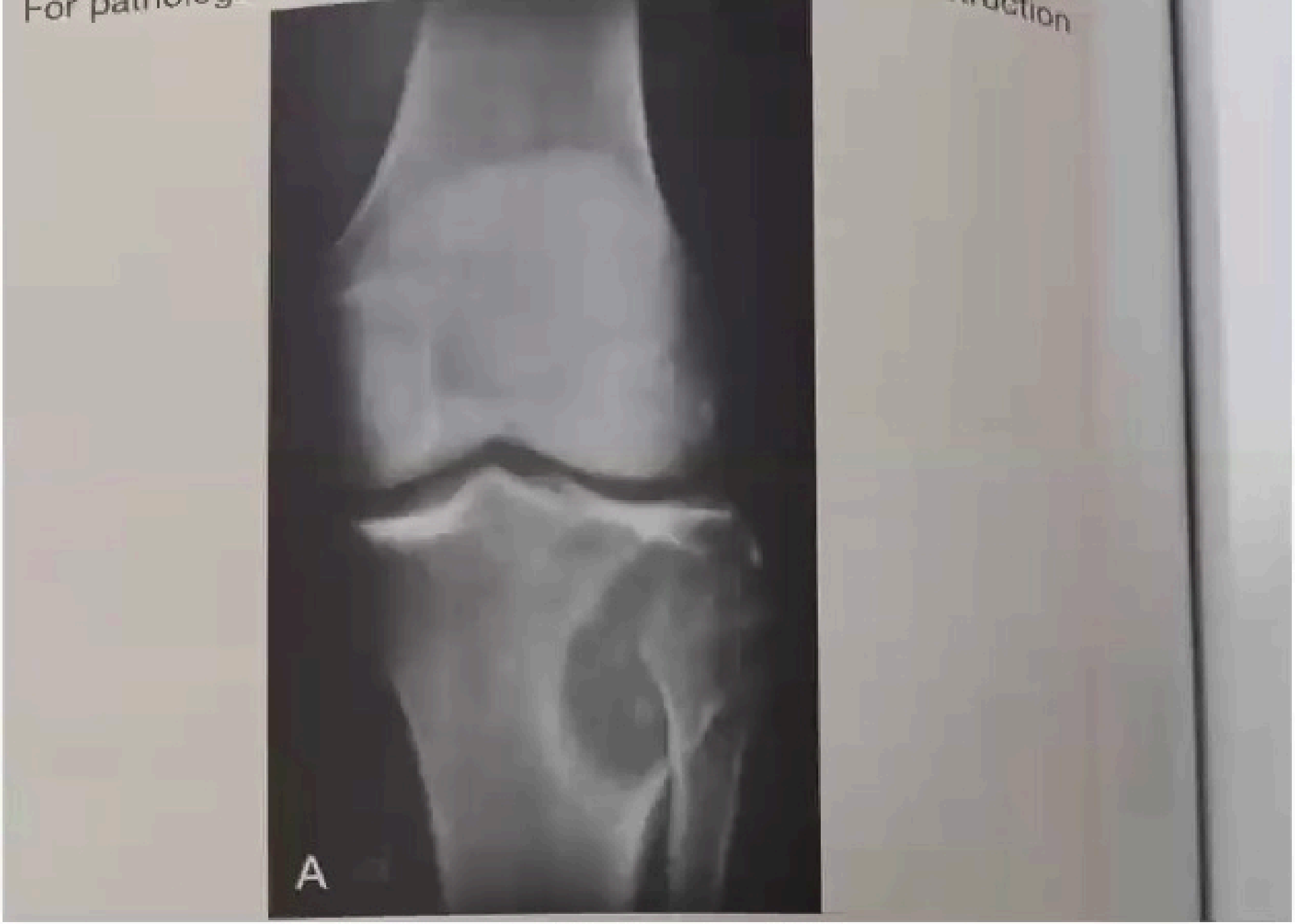
FIGURE 8-8 ■ Conventional osteosarcoma locations. (From McCarthy, E F, and Frassica, F J: Pathology of Bone and Joint Disorders, Philadelphia, WB Saunders, 1998.)



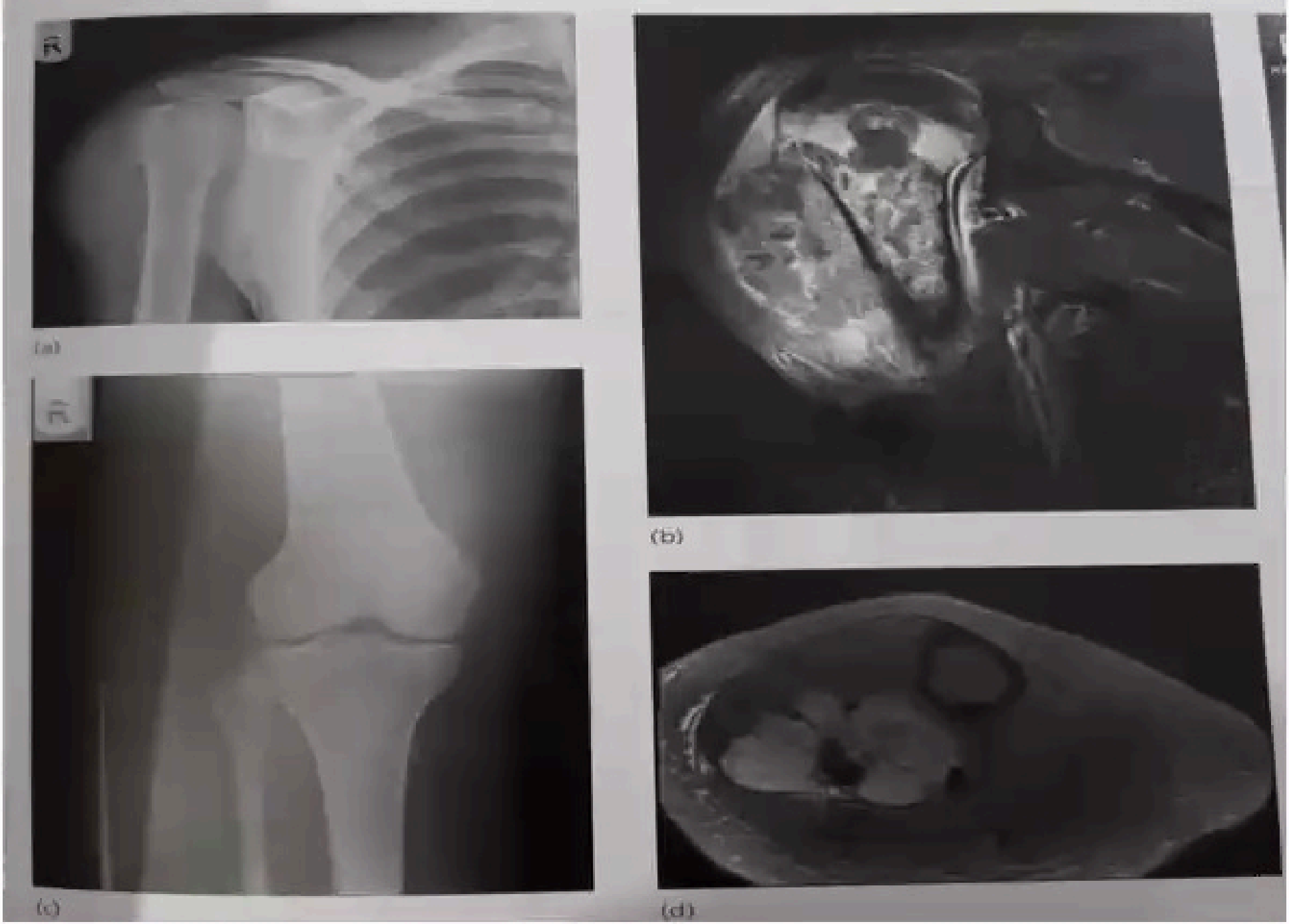
Left:Parosteal osteosarcoma
Right:periosteal osteosarcoms



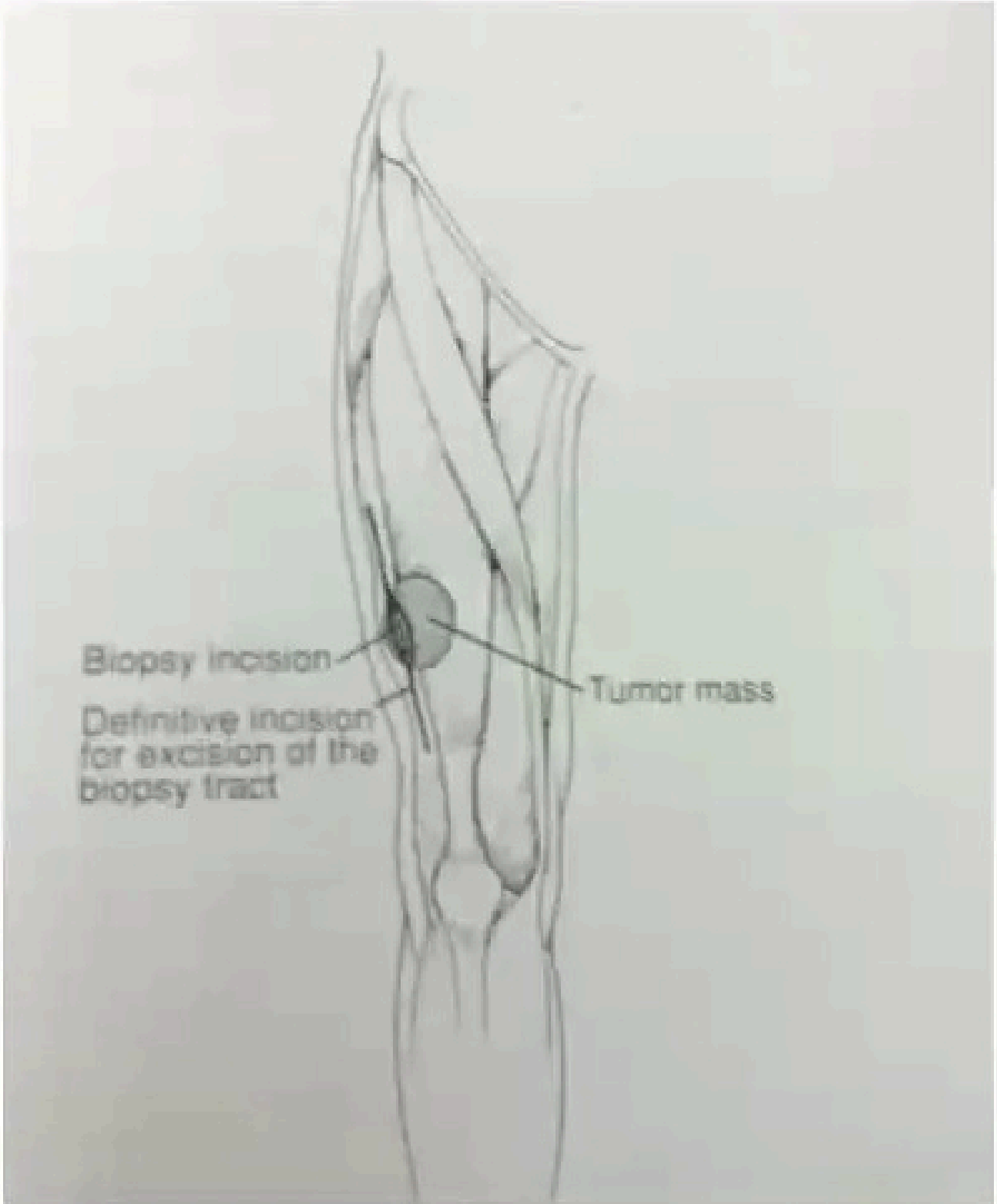
Enchondroma of the distal femur



Giant cell tumor of the proximal tibia



Ewings sarcoma


Biopsy incision
Definitive incision for excision of the biopsy tract
Tumor mass

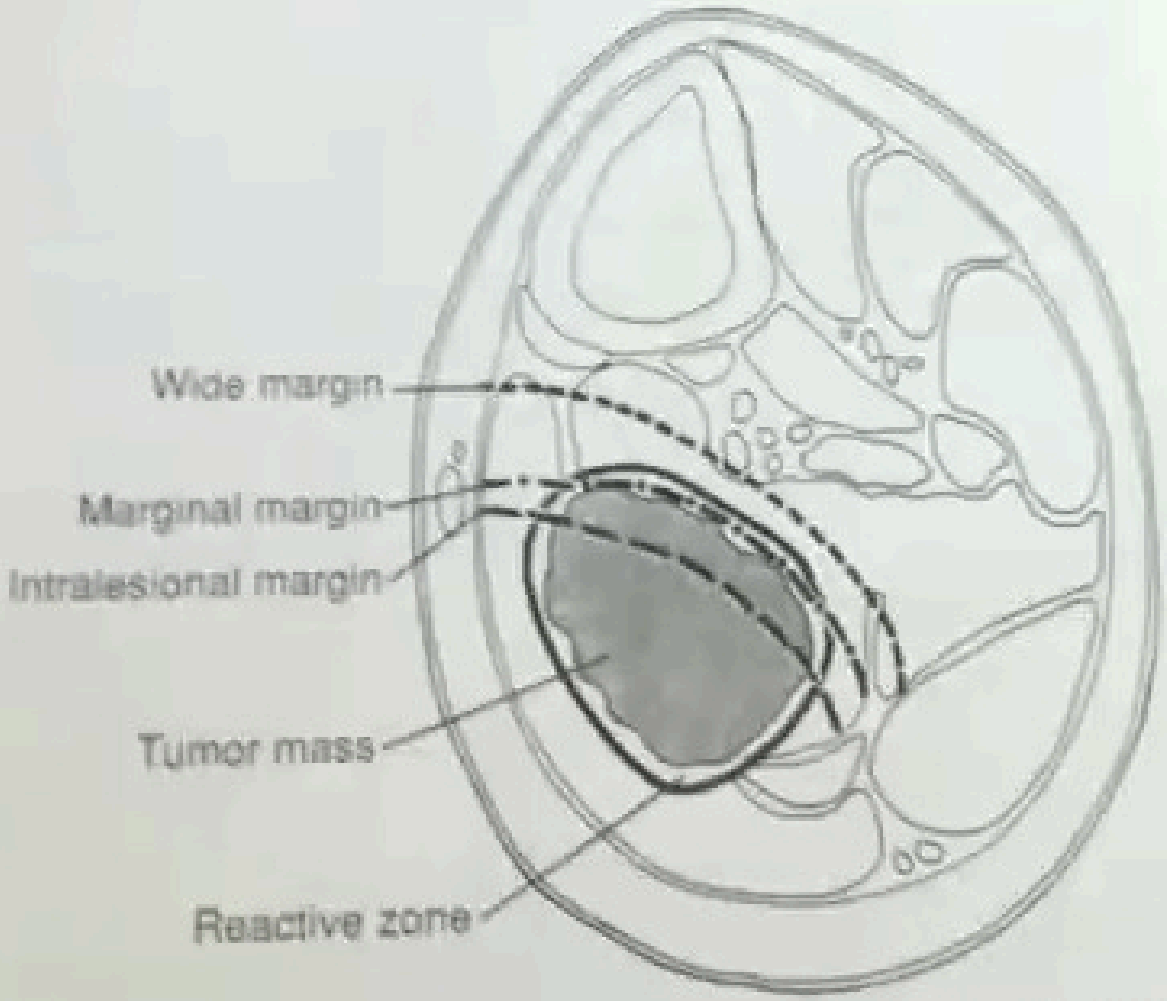
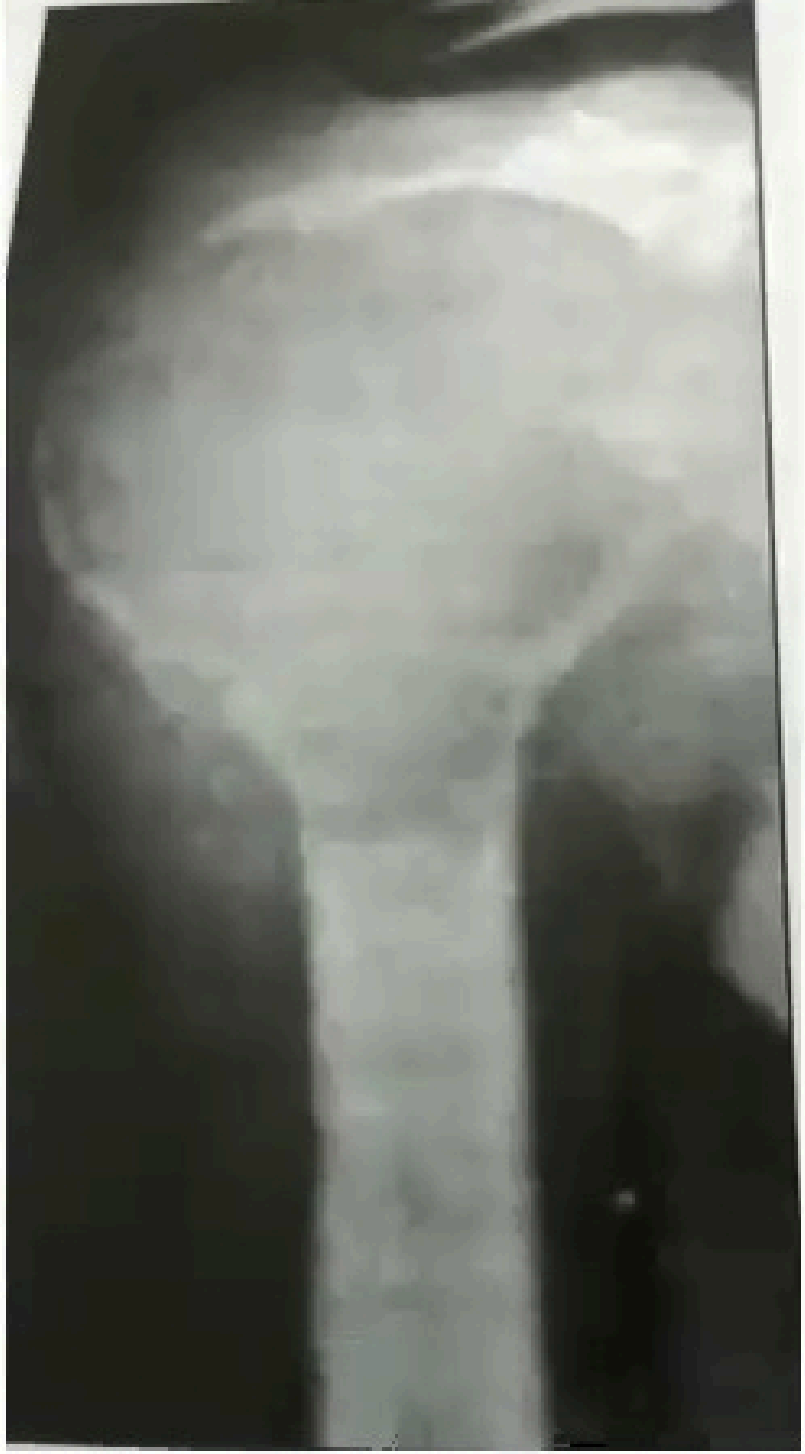


FIGURE 8-2 ■ Types of surgical margin. An intralesional line of resection enters the substance of the tumor. A marginal line of resection travels through the reactive zone of the tumor. A wide surgical margin removes the tumor with a cuff of normal tissue. [From Sim, F.H., Frassica, F.J., and Frassica, D.A.: Soft tissue tumors: diagnosis, evaluation and management. AAOS 2:209, 1994. Copyright 1994 American Academy of Orthopaedic Surgeons. Reprinted with permission.]



Example of a lesion destroying the cortex in the proximal humerus