2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

Osteosarcoma

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Treatment

- Primary tumour excision-Wide local excision with mega prosthesis reconstruction.
- Open right multiple pulmonary metastatectomy with right segmentectomy
- On chemotherapy

OSTEOSARCOMA

- Primary malignant tumour of bone

- derived from undifferentiated connective tissue and forms neoplastic osteoid.

• ***CLASSIFICATION OF OSTEOSARCOMA***
• **A. INTRAOSSEOUS OS** **- CONVENTIONAL** **- TELANGIECTETIC** **- SMALL CELL** **- LOW GRADE** **- CORTICAL**
• **B. SURFACE OS** **- PERIOSTEAL** **- HIGH GRADE PERIOSTEAL** **- PAROSTEAL/ JUXTACORTICAL**
• **EXTRAOSSEOUS OS**
• **MULTICENTRIC OS**
• **SECONDARY OS**

CENTRAL OSTEOSARCOMA

- Central osteosarcoma is the second most common primary malignant bone tumor (multiple myeloma is the most common).

- Osteosarcoma is approximately twice as common as chondrosarcoma and three times more frequent than Ewing's sarcoma

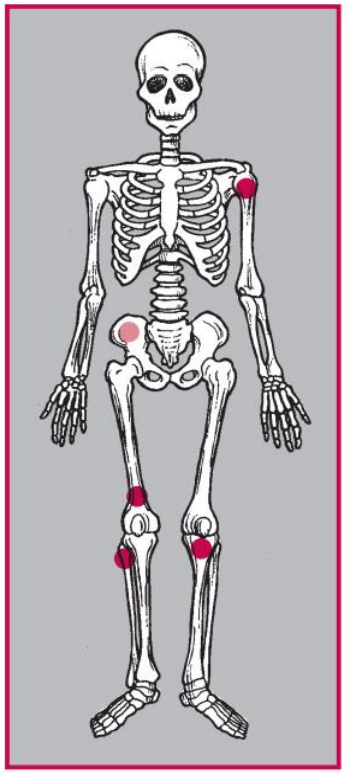
- **Age Distribution;**
- most commonly in the age group from 10 to 25 years (75% of cases); few cases occur before age 5 or after age 30

- **Signs and Symptoms**
- Painful swelling at the site of the lesion is the presenting complaint in approximately 85% of patients

- The pain initially is insidious and transitory, it eventually becomes severe and persistent.

Location

- The long bones of the extremities are the target sites for osteosarcoma.
- The knee and shoulder are the most commonly affected joints.
- 58% of all osteosarcomas occur around the knee.
- The metaphysis abutting the physis is the classic location in 75% of cases; diaphyseal, a well as epiphyseal, lesions have been reported (25%)
- Other bones that may be involved are the calvaria, sacrum, pelvis, mandible, maxilla, scapula, clavicle, ribs, hand, calcaneus, and spine
- Primary osteosarcoma of the spine is rare. Osteosarcoma also occurs in vertebrae affected by Paget's disease.



1-91 SKELETAL DISTRIBUTION

Metastases

- Metastases to the lungs are common
- Multiple lesions are the usual presentation, and these actually represent sarcomatous bone growths within the lungs (cannonball metastases)
- Spontaneous pneumothorax is common because subpleural nodules that have undergone excavation lead to rupture into the pleural space.
- Skeletal metastases (skip lesions) are also found but do not occur with the same frequency as in Ewing's sarcoma and are poor prognostic signs.
- the most frequent metastatic sites are the lungs (95%), bones (50%), and kidneys (12%).

Figure 11-99 **CANNONBALL METASTASES. Lung.** Note the multiple radiopacities scattered throughout both lungs, representing cannonball metastases from osteosarcoma of the distal femur. These radiopacities actually represent sarcomatous growth of bone within the lung parenchyma.

- **Pathologic Features**

- Three basic patterns exist:
  - 50% of cases are sclerotic,
  - 25% are purely lytic,
  - 25% are mixed lesions. •

Figure 11-97 **OSTEOSARCOMA: TIBIA. A. AP Knee. B. Lateral Knee.** Observe the diffuse ivory osteosarcoma in the proximal tibia. *COMMENT:* Approximately 50% of the osteosarcomas present as a sclerotic lesion, rendering an ivory or sclerotic appearance.

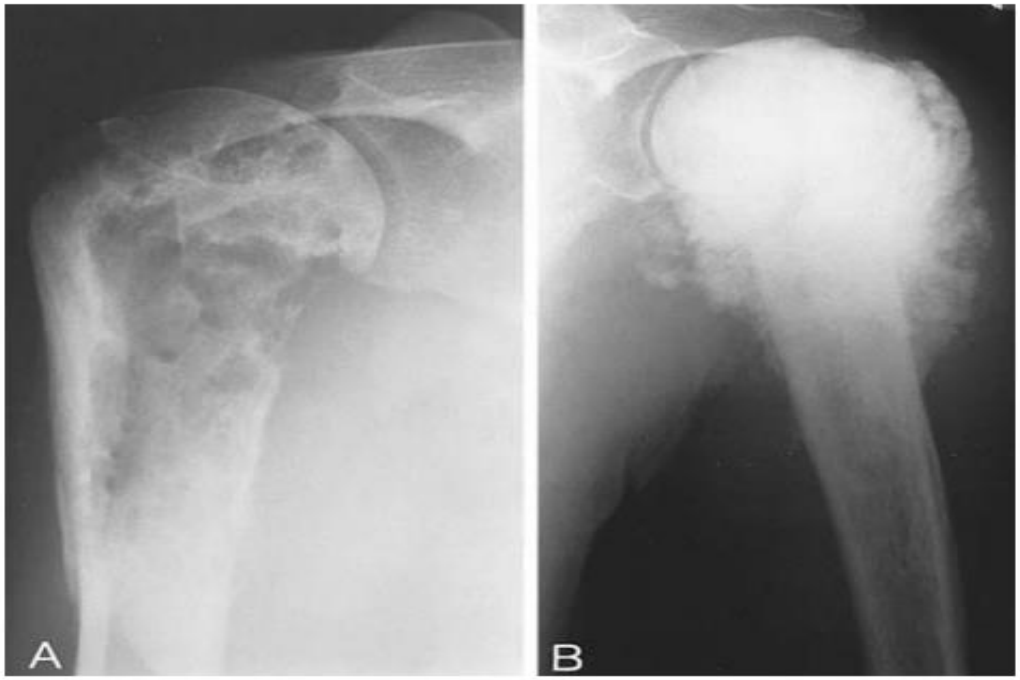


Figure 11-101 **THE RADIOGRAPHIC APPEARANCE OF LYTIC VERSUS SCLEROTIC OSTEOSARCOMA. A. Lytic Presentation: Proximal Humerus.** Note the focal lesion in the metaphysis of the proximal humerus, demonstrating mottled, permeative destruction. This appearance is characteristic of a lytic presentation in osteosarcoma. **B. Sclerotic Presentation: Proximal Humerus.** Note the dense radiopaque appearance to the humeral head and its metaphysis. A large soft tissue mass and spiculated periosteal response are associated with this sclerotic or ivory type of osteosarcoma. *COMMENT:* Approximately 50% of osteosarcomas present as sclerotic lesions; the remainder are lytic in presentation. Of the lytic cases, 25% have a mixed pattern if moderate osteoid matrix is present.

Radiologic Features

- Osteosarcomas usually arise in the metaphysis (75%) of long bones,
- most frequently at the distal end of the femur, the proximal end of the tibia, and the proximal end of the humerus.
- Rarely affect the diaphysis or epiphysis (25%) and may involve any bone
- The epiphyseal plate functions as a barrier to tumor migration in some cases.

- The classic presentation is that of a focal lesion in the metaphysis, creating either a mottled, permeative lesion with a poorly defined zone of transition or a dense ivory or sclerotic region filling the medullary space.
- Cortical disruption is common.
- Periosteal new bone formation occurs and is highly irregular.
- This periosteal reaction often takes place within an extracortical, dense soft tissue mass that displays transverse spicules or radiating striations
- This characteristic pattern of periosteal response has been referred to as sunburst, or sunray, and is the appearance for which osteosarcoma is best known.

Figure 11-102 **OSTEOSARCOMA. Proximal Humerus.** Note the ill-defined, destructive lesion assuming the mixed pattern of presentation in the metaphysis and proximal diaphysis of the humerus. Malignant spiculated periosteal response is observed on both sides of the humeral cortex, with extension of the osteosarcoma into the soft tissue structures.

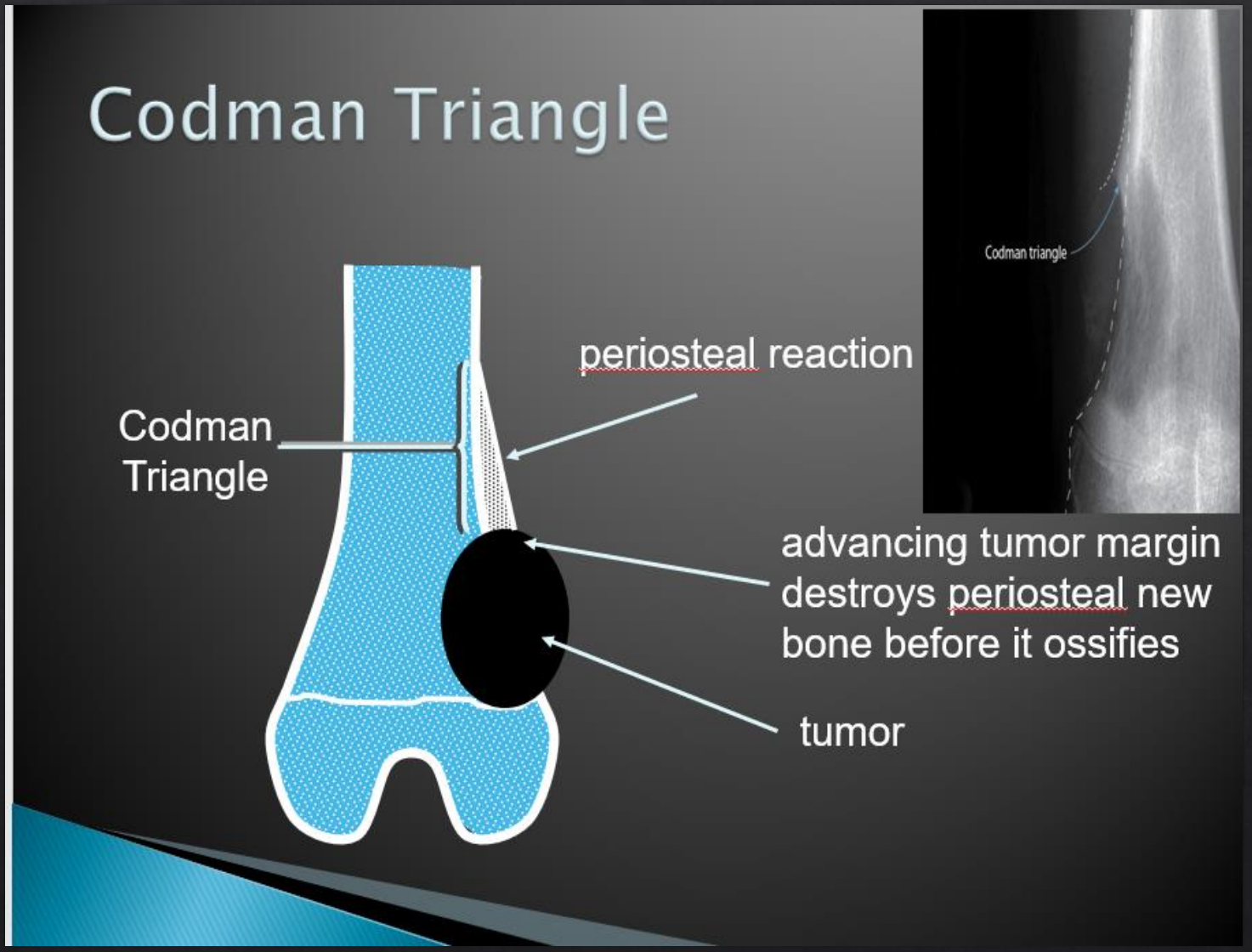
Figure 11-103 **SCLEROTIC OSTEOSARCOMA: MALIGNANT PERIOSTEAL RESPONSE IN THE DISTAL FEMUR A. AP Knee. B. Lateral Knee.** Observe the increase in bone density in the metaphysis and distal diaphysis of the femur. There is a sunburst or malignant spiculated periosteal response present surrounding the entire distal metaphysis of the femur. This is a classic radiographic presentation for osteosarcoma.

- Occasionally, the periosteum is found to be elevated by tumor tissue on the upper and lower margins of the lesion, creating Codman's reactive triangles.

- This is a reactive response to the lifting of the periosteum and is not pathognomonic for osteosarcoma .

- Also seen in traumatic periostitis, osteomyelitis, eosinophilic granuloma, and thyroid acropachy.

Codman Triangle



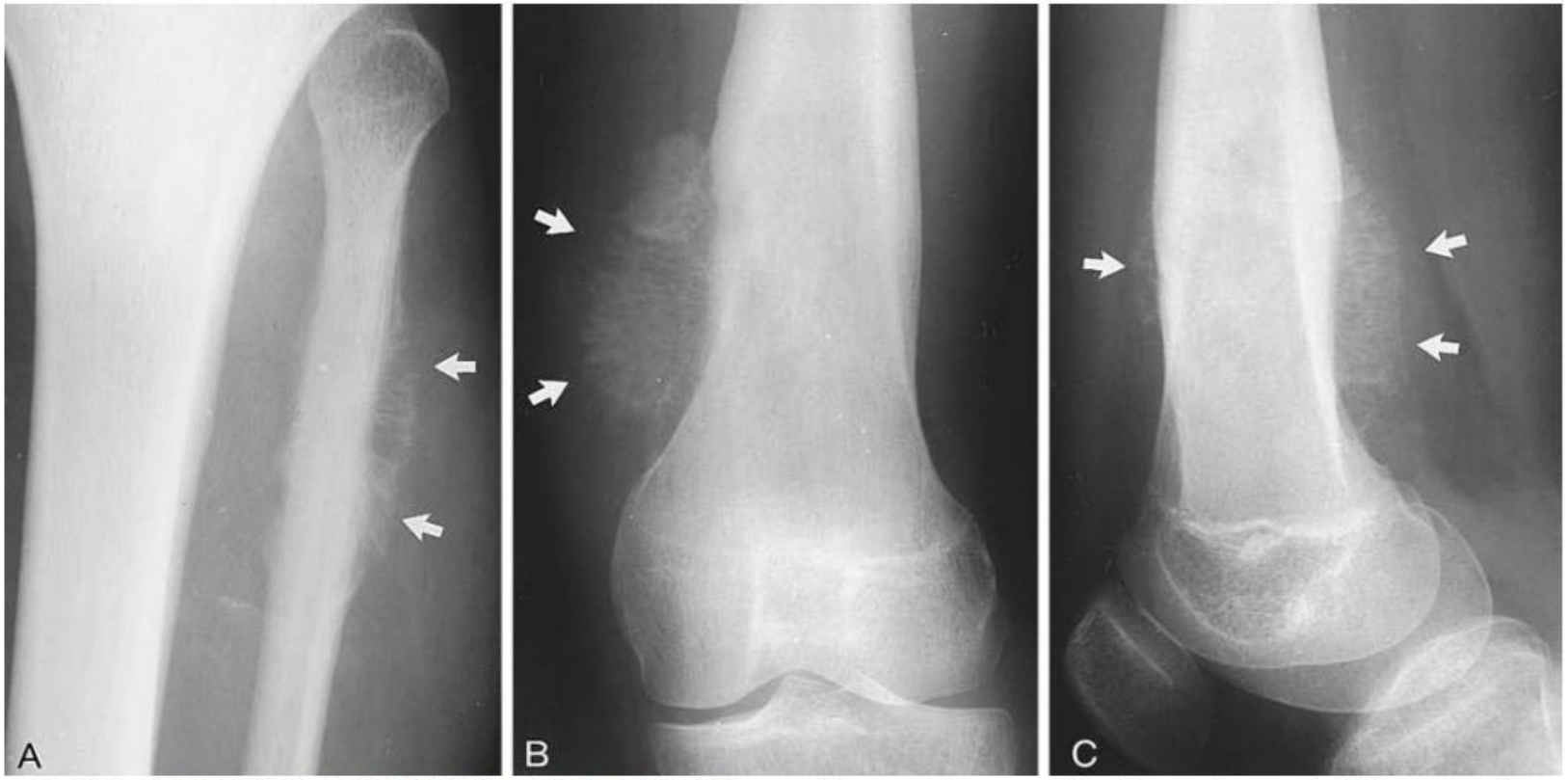


Figure 11-105 **PERIOSTEAL RESPONSE IN OSTEOSARCOMA. A. Fibula.** Observe the spiculated periosteal response on the lateral surface of the proximal fibula (*arrows*). **B. AP Distal Femur. C. Lateral Distal Femur.** Observe the classic sunburst or spiculated periosteal response seen in the distal metadiaphyseal area of the femur (*arrows*). This sunburst appearance, the soft tissue mass, and the destructive lesion of the medullary portion of the femur are the cardinal radiographic features of osteosarcoma.

- As the lesions continue to grow, bone expansion may occur.
- pathologic fracture may follow some form of trivial trauma.
- 50% of these lesions are radiopaque or sclerotic and may develop a roughened lobulated margin, referred to as the cumulus cloud appearance.
- The remaining 50% are split between a purely osteolytic lesion (25%) and a mixed lesion (25%)

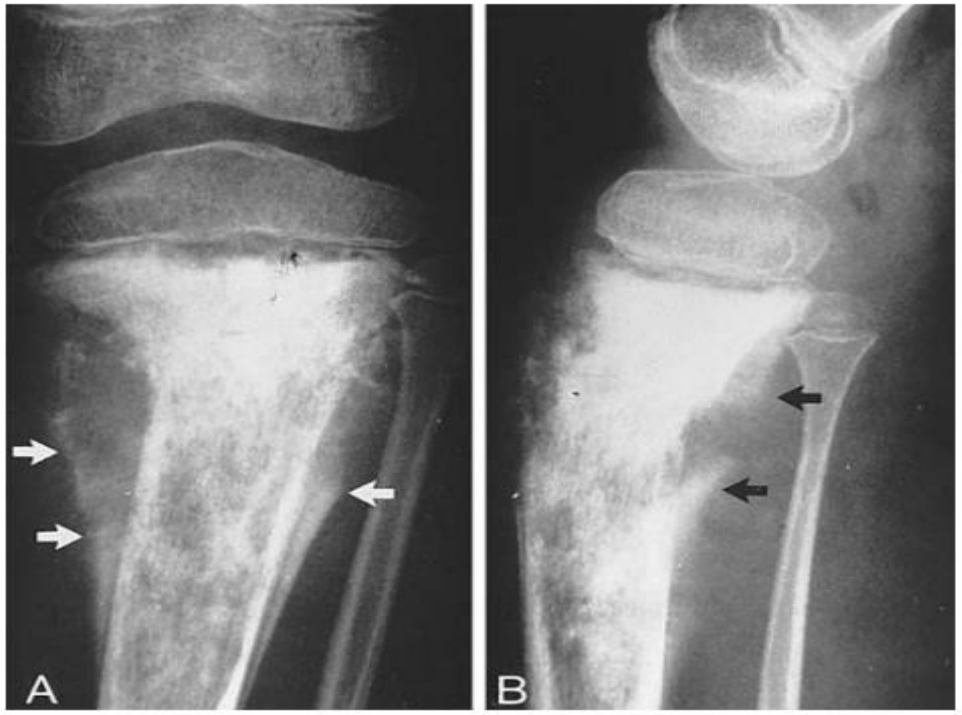


Figure 11-106 **CODMAN'S REACTIVE TRIANGLE AND OSTEOSARCOMA OF THE PROXIMAL TIBIA. A. AP Tibia. B. Lateral Tibia.** Note the mixed pattern of osteosarcoma in the proximal metaphysis of the tibia. There are large Codman's reactive triangles present surrounding the cortex of the proximal tibia (*arrows*). *COMMENT:* Codman's reactive triangle is frequently found in osteosarcoma of bone; however, it is not pathognomonic of osteosarcoma and can be found in infectious lesions, traumatic periostitis, and peculiar callus formations.

Figure 11-107 **THE CUMULUS CLOUD APPEARANCE OF OSTEOSARCOMA. Pelvis.** Observe the diffuse, sclerotic osteosarcoma affecting the ilium near the acetabulum. With closer inspection, note the lobulated margins of the medial surface of this osteosarcoma (*arrows*). This lobulated margin of osteosarcoma has been called the cumulus cloud appearance. (Courtesy of Friedrich H. W. Heuck, MD, Stuttgart, West Germany.)

- Soft tissue mass formation is common in osteosarcoma and grow to large dimensions.

- Ossification within this tumor mass is a frequent occurrence.

CT

- Cross-sectional images provide a clearer indication of bone destruction, as well as the extent of any soft tissue mass, than do radiographs.

- CT scanning may depict small amounts of mineralized osseous matrix not seen on radiographs. The modality may be particularly helpful in visualizing flat bones, in which periosteal changes may be more difficult to appreciate

MRI

T_1WI - Low/heterogenous signal intensity

T_2WI - High signal intensity

Contrast - enhancing medullary cavity and solid components.

STIR - High signal intensity and helps in assessing involvement of neurovascular bundles and muscles.

Helps to identify ;

- Preoperative assessment & staging
- Define the intramedullary extent , skip lesions
- Extent of the soft tissue component
- Define the neurovascular relationship
- Define articular cartilage / joint involvement
- Response to therapy

Treatment

- Neo adjuvant chemotherapy, limb salvage surgery with pulmonary metastatectomy , adjuvant chemotherapy

Surface osteosarcoma

- **Includes Periosteal, Parosteal and High grade surface OS.**

Parosteal sarcoma (juxtacortical osteosarcoma, surface osteosarcoma)

- tumor situated on the surface of a bone and is biologically different from its intramedullary counterpart.
- well-differentiated osteosarcoma arises in a juxtacortical location within the periosteum.
- relatively uncommon slow growing lesion representing 3–4% of all osteosarcomas and about 1% of all primary malignant bone tumors.
- symptoms are swelling or mass formation, with a dull, aching pain.
- The typical age of incidence range is from 30 to 50 years

- **Location**

- The majority of cases involve the posterior surface of the distal femoral metaphysis (50% of cases)

- Other common sites are the proximal tibia and humerus (25% of cases).

Radiologic Features

- dense (radiopaque) homogenous juxtacortical mass.
- The classic lesion affects the popliteal surface of the distal femur.
- A peculiar radiolucent cleft separates the majority of the ossified mass from the cortex of the femur
- This cleft (1 to 3 mm in width) stops abruptly at the stalk of the tumor and has been referred to as the cleavage plane or string sign of parosteal sarcoma.
- The periphery of the tumor is usually lobular in outline, and no periosteal (spiculated or laminated) new bone formation can be recognized.

- The treatment will vary, depending on the histologic aggressiveness of the tumor.
- En bloc resection for the less aggressive lesions seems helpful; however, a high rate of recurrence exists (> 50%).
- Amputation should be performed only in large lesions that are determined to be highly aggressive.

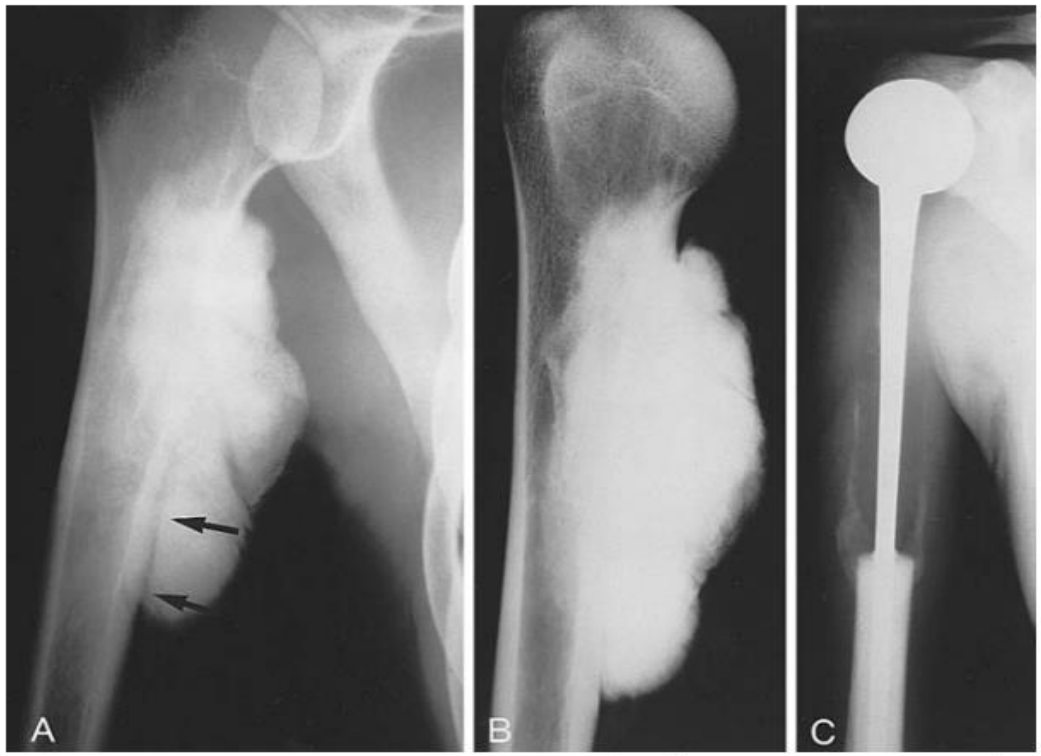


Figure 11-110 **PAROSTEAL SARCOMA. A. AP Humerus.** Observe the large homogeneous radiopaque juxtacortical mass present in the proximal metadiaphyseal area of the humerus. At the caudal end of this mass a clearly defined cleavage plane is visualized (*arrows*). **B. Specimen Radiograph. C. Prosthesis.** Observe the metal prosthesis used in the treatment of this patient's parosteal sarcoma of the proximal humerus. *COMMENT:* The cleavage plane referred to previously actually represents a periosteal fibrous tissue layer separating the tumor from the cortical surface. This has also been referred to as the string sign of parosteal sarcoma and is present in only 30% of cases. (Courtesy of Friedrich H. W. Heuck, MD, Stuttgart, West Germany.)

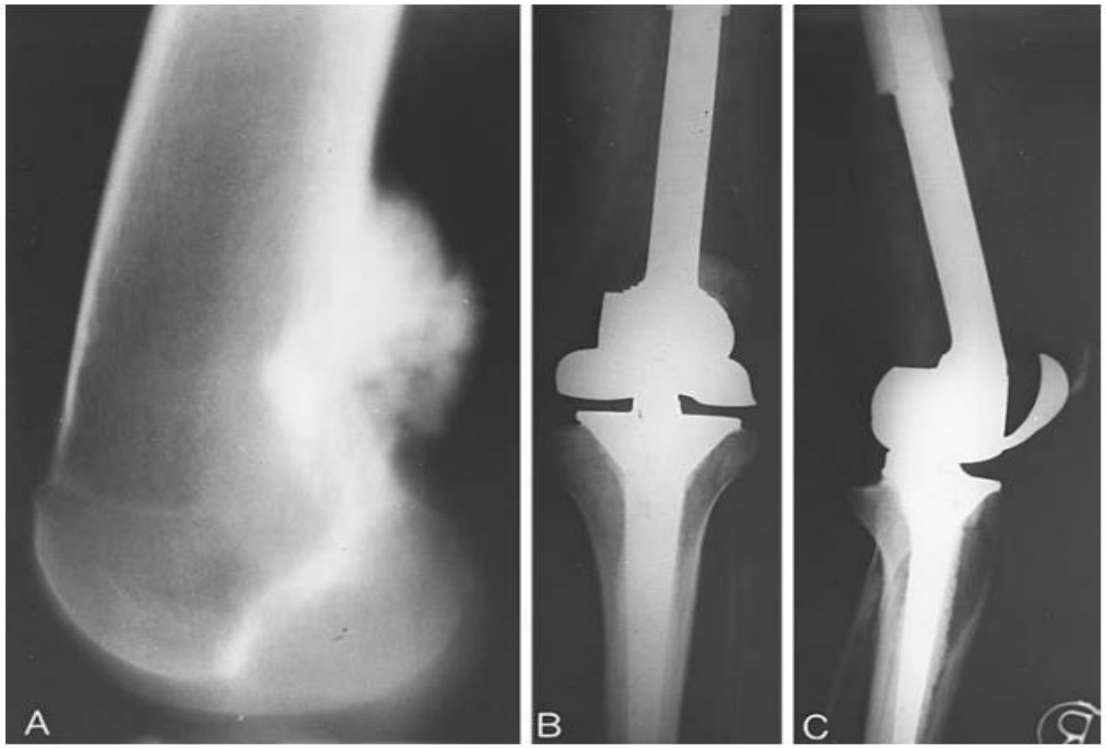


Figure 11-111 **PAROSTEAL SARCOMA: DISTAL FEMUR.** **A. Tomogram, Lateral Knee.** Observe the juxtacortical radiopaque mass on the posterior surface of the distal femur. **B. AP Knee. C. Lateral Knee.** Note that the postoperative radiographs demonstrate a prosthesis that has worked well for this 21-year-old patient. (Courtesy of Cynthia K. Peterson, DC, DACBR, Toronto, Canada.)

Figure 11-112 **PAROSTEAL SARCOMA. Distal Femur.** Observe the nodular ivory radiopaque mass on the posterior surface of the distal femur. This is the classic location for parosteal sarcoma.

PAROSTEAL OS	PERIOSTEAL OS
1. STRING SIGN – Radiolucent line separates tumor from cortex	1.Tends to WRAP AROUND the circumference of the bone.
2. Posterior distal femur	2.Medial distal femur
3. Usually LOW GRADE	3.INTERMEDIATE prognosis(better than conventional OS)
4. Metaphysis	4.Diaphyseal

SECONDARY OSTEOSARCOMA

- Malignant degeneration of benign disorders, such as Paget's disease, polyostotic fibrous dysplasia, hereditary multiple exostosis (osteochondromas), and enchondromatosis (Ollier's disease), to osteosarcoma are documented.

- Ionizing radiation may lead to the development of bone sarcoma.

- postradiation sarcoma after treatment for breast cancer, Wilms' tumor, and other primary carcinomas.

- injections of thorotrast, a radioactive contrast medium .

- **The radiographic features**

- permeative, moth-eaten bone destruction with periosteal reaction and soft tissue mass is largely indistinguishable from those osteosarcomas arising de novo

EXTRAOSSEOUS OSTEOSARCOMA

- The most common site is the soft tissues of the thigh, but they can also be found in the pleura, heart valves, dura of the brain, retroperitoneum, buttock, axilla, breast, and renal capsule.
- prognosis is similar to osteosarcoma, with metastases to the lung being common.
- Most occur between 30 and 50 years of age.
- The histologic features are identical to osseous osteosarcoma.
- The radiologic features are those of a large soft tissue mass, which is often not adjacent to a bone.

EWING'S SARCOMA

- Highly malignant primitive primary malignant tumor of bone
- it is composed of tumor cells derived from the connective tissue framework of bone marrow.
- originates from the marrow stem cells with striking predilection for the marrow-rich diaphysis.

- **Incidence**
- 7% of all primary malignant bone tumors
- fourth most common primary malignant bone tumor

Age distribution

- most commonly in the 10- to 25-year age range, with a peak at 15 years.
- 50% of cases occur < 20 years of age.

- **Signs and Symptoms**

  - Localized pain with swelling at the site of the lesion is a consistent presenting complaint.

  - A soft tissue mass is palpable in more than one third of the cases.

  - Systemic symptoms of slight to moderate fever, secondary anemia, leukocytosis, and increased ESR are common in Ewing's sarcoma.

- **Location**

- most frequently in the long tubular bones (50%) and in the flat bones (40%)

- The femur is most commonly involved (23%), with the tibia, fibula, and humerus following (9%).

- The diaphysis is the classic location for these tumors; however, more cases are being seen affecting the metaphysis and meta-diaphyseal region.

- flat bones -the most frequently affected site is the pelvis.

Radiologic Features

- diaphyseal lesion (usually in the lower extremity), permeative in its appearance, with a wide zone of transition.
- A delicate, laminated, onionskin, or onion peel periosteal response is noted in only 25–50% of cases.
- Cortical saucerization is an early and characteristic sign
- irregular defect effaces the outside of the bone occasionally, exhibiting a marginal scalloping effect- occurs when the tumor grows through the haversian system and presents subperiosteally.
- a mixed lytic and sclerotic pattern predominating in the tubular bones.

- **Periosteal Bone Formation-**

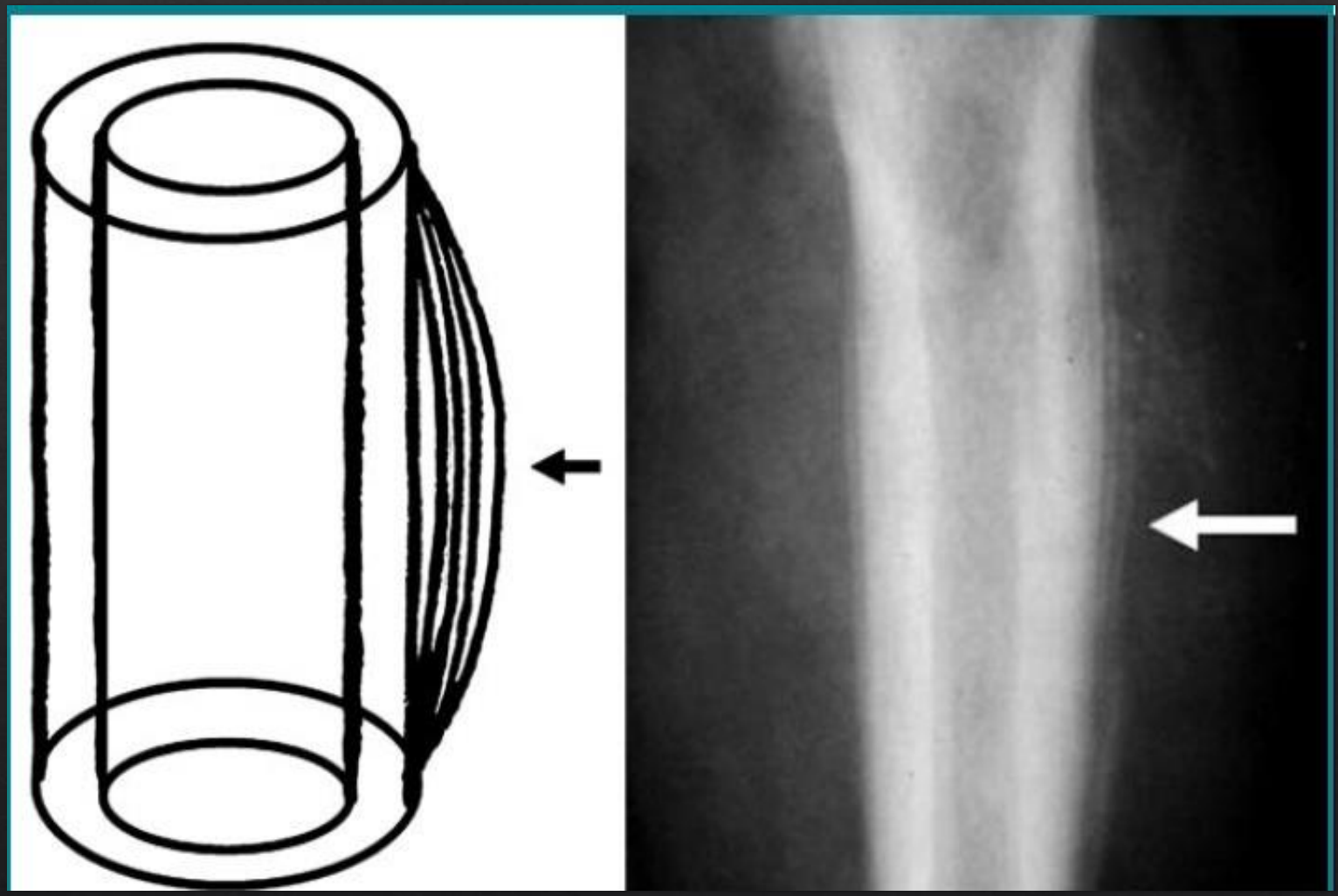
- delicate parallel, laminated, onion skin, or onion peel radiographic presentation is quite striking in its appearance when present.

- caused by the splitting and thickening of the cortex by the tumor cells. The layering is continuous, with reactive ossification in the form of Codman's triangles occurring frequently

- sunray pattern of periosteal new bone formation may also occur in Ewing's sarcoma. These radiating spicules have been referred to as the groomed or trimmed whiskers effect

- these perpendicular bone spicules are thinner and more hair-like than those in osteosarcoma.

Figure 11-128 **EWING'S SARCOMA: DIAPHYSEAL INVOLVEMENT. A. AP Ulna. B. Lateral Ulna.** Observe the lytic destructive lesion in the diaphysis of the ulna. There is moderate expansion of the ulna noted. No characteristic periosteal response is present.



- **Metastases**

- Skeletal metastases occur frequently and early, leading to extensive bone destruction.

- most common primary malignant bone tumor to metastasize to bone

- The spine is a common site for metastasis.

- Multiple lesions in the one bone occur and are described as skip lesions, a phenomenon also seen in osteosarcoma.

- Secondary spread to the lungs is also a common occurrence, with the lung parenchyma and pleura being the favored locations.

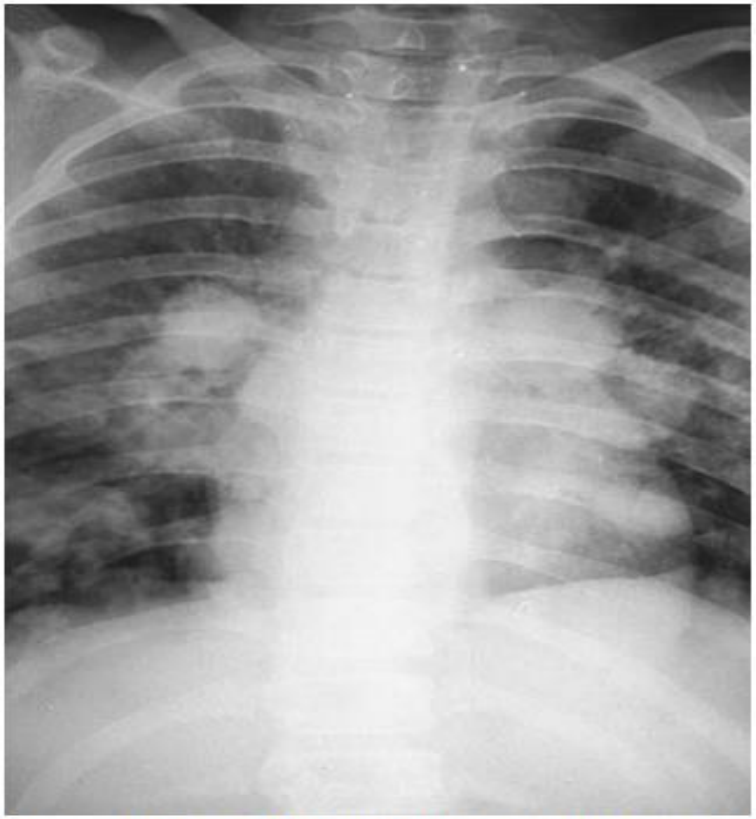


Figure 11-132 **CHEST CANNONBALL METASTASES.** Observe the multiple circular nodular metastatic lesions scattered throughout the central lung fields. These represent cannonball metastases, with sarcomatous growth within the lung parenchyma from Ewing's sarcoma of the femur.

Treatment and Prognosis

- intensive chemotherapy -80% survival rate.

- Amputation has offered the best treatment when the lesion is surgically accessible.