EWING'S SARCOMA

- It is the primary malignant tumor of bone arises from reticuloendothelial cells of medullary cavity/ endothelial cells in bone marrow

- **Incidence:**
- - 4th most common primary malignant tumor of bone (6-8%)
- - 2nd most common before the age of 30
- - Most common between 10-20 years (less than 10 years)
- - (6-8%) of all primary malignant tumor

- **Delay of onset of symptoms to diagnosis:** 34 weeks
- (Patient: 15 weeks + Physician- 19 weeks)
- **Course:** Period of remission (decreased size of the tumor) and exacerbation (increase size of the tumor)
- **Metastasis:** 1st in bone (skull , vertebrae , ribs) → then in lungs (via lymphatic+ Haematogenous)

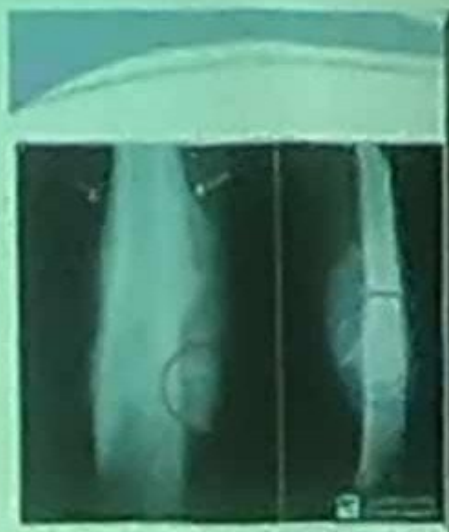
- **Clinical features:**
- - Pain (E.S swelling then pain but in O.S Pain then swelling)
- o Throbbing
- o Worse at night
- - Swelling :
- o warm, tender (suggestive of osteomyelitis)
- o Dilated veins over the skin , firm in consistency, fixed with underlying bone
- o Generalized illness
- o Pyrexia (constitutional symptoms)
- o Metastatic features of lung may present

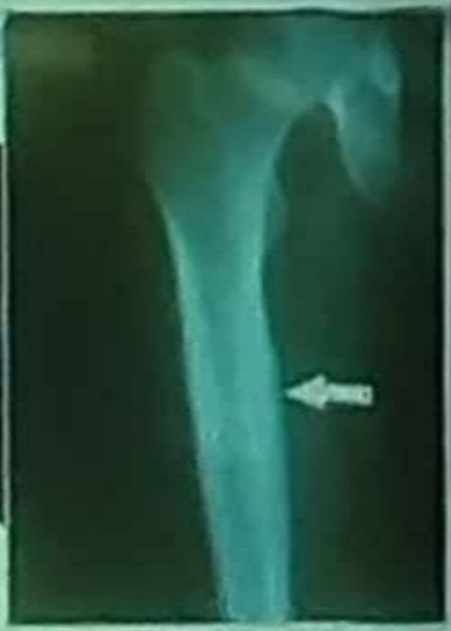
- **Predisposing factor:**
- **Age:** 10-20 years (Apley) , less than 10 years – most common
- **Sex:** Male: Female = 1.5:1
- **Race:** Rare in black and Chinese children
- **Site:**
- - Diaphysis of skeletally immature long bone
- - Often extend in metaphysis
- - Usually in a tubular bone
- - Tibia, fibula ,clavicle
- - Flat bones- shoulder and pelvic girdle ,skull
- - Rarely ,spine and small bones of the hand and feet

- **Differential diagnosis:**
- - Sub-acute osteomyelitis: blood culture
- - Osteosarcoma: biopsy
- - Reticular cell sarcoma: Biopsy (Non- hodgkins's lymphoma)
- - Malignant neuroblastoma
- - Malignant round cell tumor

- **Biopsy:** To confirm the diagnosis (Should be done at last because it distort the tumor appearance)
- **CT + MRI:** Reveals the large extra- osseous component.
- **Bone scan:** Show multiple areas of activity in the skeleton. (Detects skip lesion and secondary silent deposit)

- **X ray of the affected part:**
- - Area of bone destruction, predominant in mid-diaphysis
- - **Onion peel affect:** new bone formation extend along the shaft and it appears as fusiform layers of bone around the lesion





- - Often extend into surrounding soft tissues ,
- with radiating streaks of ossification (sunray appearance) and reactive periosteal bone at proximal and distal margins (Codman's triangle)
- - Sunray appearance and Codman's triangle also present in Osteosarcoma

- **Investigation:**
- CBC- leukocytosis 20000-40000/ mm3
- ESR: ↑↑
- Hb%: ↓↓
- CRP: ↑↑↑

- **Prognosis of Ewing's sarcoma:**
- Age: Older age, poor prognosis (with a cut off 12-15 years)
- Sex: Male poor prognosis than female
- Site: Proximal part poor prognosis than distal
- Size: Larger – poor prognosis
- Metastasis: Distant metastasis – poor prognosis

- For inaccessible site:
- - Neo- adjuvant radiotherapy+
- - Surgery (wide local excision)
- - Adjuvant chemotherapy for 1 year.

- Postoperative (Adjuvant) Chemotherapy: May be added if the resected specimen is found not to have a sufficiently wide margin of normal tissue.

- **Treatment of Ewing's sarcoma:**
- Best result is achieved by combination of radiotherapy, chemotherapy and surgery
- For accessible site:
- - Neo-adjuvant chemotherapy +
- - Surgery (wide local excision/ amputation/ disarticulation) +
- - Adjuvant chemotherapy for 1 year.