

**Ewings Sarcoma**  
**Family of Tumors**

**By**  
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**Ewings Sarcoma**  
**Described by. James Stephen Ewings in 1920**  
**Small Round Blue Cell tumor**

**Ewings Sarcoma**  
**Ewings Sarcoma family of Tumors**  
**The term PNET is no longer used**

**Ewings Sarcoma**  
**Bone**  
**Soft Tissue**  
**Thoraco Pulmonary Askins**  
**Breast**  
**Female Genital Tract**  
**Others**

**Age 5 - 20**  
**Below 30**  
**Both sexes**

**Simulates Osteomyelitis**  
**Pain Fever leucocytosis**

**Bones involved**

**Any bone**

**Femur , Tibia ,Humerus , Fibula , Pelvis Ribs ,Vertebrae. Clavicle**

**Site**

**Medullary canal**

**Metaphysis and Diaphysis**

**Penetrates the cortex**

**Invaded the soft tissue**

**Imaging Xrays. CT MRI ,PET CT**

**Permeative destruction of bone**

**Onion shining**

**Periosteal reaction**

**Involvement of soft tissue**

**Microscopic**

**Small round blue cells in sheets Separated by Fibrous tissue , Necrosis.**

**Pseudo rosettes**

**IHC and molecular**

**CD 99 Positive**

**11 / 22 Translocation**

**EWSR 1 fuses with FL1**

**EWSR 1 fuses with ERG**

**NKX 2.2 positive**

**CD 99 Positive and**

**NKX 2.2 Positive**

**Highly specific for Ewings Sarcoma**

**PAS stain is Positive in 50 to 60 % of the cases**

**Mets**

**Lungs**

**Pleura**

**Other bones**

**Skull**

**CNS**

**25 % are multicentric**

**Treatment**

**Radio**

**Chemo**

**Ewings Sarcoma and Adamantinoma**

**Primary Extraskkeletal Ewings Sarcoma**

**Adolescents and young adults**

**Sites Deep soft tissues**

**Lower extremity**

**Paravertebral**

**Any site**

**Round Cell tumor with Ewings like features**

**Ewings like**

**CIC Rearrangement**

**DUX 4 gene**

**CIC - FOXO 4 Fusion**

**Ewings like**

**BCOR - CCNB3 fusion**

**DD**

**Neuroblastoma**

**Alveolar RMS**

**Small cell Osteosarcoma**

**Poorly Differentiated Synovial Sarcoma**

**NHL**

**Desmoplastic Small Round Cell tumor**

**Mets from Pulmonary Small cell Carcinoma**

**Mesenchymal Chondrosarcoma**

**Bad Prognosis**

**Metastasis at the time of presentation**

**Large size**

**Extensive Necrosis**

**Central axis**

**Other sites**

**Ewings Sarcoma**

**of the Female Genital Tract**

**Breast**

**Dr RG. Wiseman Pinto**

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