Small Round Blue Cell Tumors
Eosinophilic Granuloma
Ewing’s Sarcoma
Lymphoma
Multiple Myeloma

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Unknown Case 1
Answer

- Eosinophilic Granuloma
Reticulin Stain
Answer

- Ewing’s Sarcoma
Small Round Blue Cell Tumors

- Small Round Blue Cells—resemble hematopoietic cells
- No Matrix Production
Small Round Blue Cell Tumors

- **Benign:**
  - Eosinophilic Granuloma (Langerhans Cell Histiocytosis; Histiocytosis X)
  - Osteomyelitis

- **Malignant:**
  - Ewing Sarcoma/PNET
  - Lymphoma
  - Metastatic Neuroblastoma
  - Multiple Myeloma (Plasmacytoma)
  - Metastatic Small Cell Carcinoma
  - Rhabdomyosarcoma (rare in bone)
Eosinophilic Granuloma

- **Definition:** Solitary or multiple lesions confined to bone
- **Hand-Schuller-Christian Disease** (1-5 years): chronic disseminated histiocytosis
- **Letterer-Siwe disease** (<1 year): acute or subacute disseminated histiocytosis
- **Solitary EG** is twice as common as multifocal EG
Eosinophilic Granuloma

- **Sites:**
  - Flat Bones (most common—70%)
    - Skull
    - Pelvis
  - Femur
  - Humerus
  - Hands and Feet are rare in solitary disease
Eosinophilic Granuloma

- Age: 5-15 years
- 95% are white
- Pain, tenderness and fever
Eosinophilic Granuloma

- **Radiology:**
  - Early: Permeative with periosteal reaction (lamellated)
  - May be sharply delineated (geographic)
  - May have rind of sclerosis
  - 5-10% of patients have an associated soft tissue mass
  - Sequestrum (button-like); Hole in a Hole
Eosinophilic Granuloma

- Skull: beveled edge; button sequestrum
- Flat bone: hole in a hole
- Spine: vertebra plana
- Long bone:
  - diaphysis: (58%)
  - Metadiaphysis (18%)
  - Metaphysis (28%)
  - Epiphysis (2%)
Eosinophilic Granuloma

- **Treatment:**
  - Curettage and bone graft
  - Observation of spine lesion—usually spontaneously regress
  - Intraleisonal prednisone
  - Low Dose XRT (300-1000 rads)
Hand-Schuller-Christian Disease

- **Triad:**
- Destructive skeletal lesions
- Exophthalmos
- Diabetes Insipidus
- 10% of patients with unifocal EG develop multifocal and extraskeletal disease
- Usually <5 years old
- Hepatosplenomegaly, adenopathy, anemia, fever, neurological complaints
- Fatal in 15%
- Any bone but 90% have skull involvement
Letterer-Siwe Disease

- Develops in 1\textsuperscript{st} year of life
- Disseminated disease and small bone lesions
- Fatal in 95\% who develop before 1 year of life
EG CD-10
Ewing Sarcoma

- Malignant
- 5% of all biopsied tumors
- Sites:
  - Long Bones—Most common
    - Femur
    - Humerus
  - Flat Bones
    - Pelvis
    - Ribs
Ewing Sarcoma

- Age: 10-25 years (5 months to 83 years)
- Diaphysis and Metadiaphysis most common
- Soft tissue mass—90%
- Chromosomal Translocation T11;22
Ewing Sarcoma

**Distribution:**

- Metadiaphyseal: 44%
- Mid-diaphysis: 33%
- Metaphysis: 15%
- Metaepiphysis: 6%
- Epiphysis: 2%
Ewing Sarcoma

- **Soft tissue mass**—90%
- **Permeative or moth eaten bone destruction**
- **Periosteal Reaction**—50%
  - Due to irritation, edema, tumor permeation
  - Onion Skin (colic pattern of irritation)
  - Hair on End (rapid continuous lifting of periosteum)
- **Reactive Bone**—10%
- **No cartilage or bone production by tumor**
- **Pathologic fracture** in 10-15%
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Ewing Sarcoma
Ewing Sarcoma
Ewing Sarcoma
Ewing HBA71 Stain
Ewing PAS
Ewing Reticulin
Ewing Electron Microscope
Lymphoma

- **Primary Lymphoma of Bone—rare** (must rule out metastatic)
- Non Hodgkins (most common); Hodgkins also occurs
- Rare—3% of primary tumors
- Any age but rare <10 years old (average age: 42 years; range: 2-88 years)
- **Sites:**
  - Long Bones—most common
  - Flat bones
  - Spine and small bones
Lymphoma

- Permeative or moth eaten bone destruction (55%)
  - Geographic (11%); Blow out (1%); Blastic (2%); Normal XR (5%)
- Metadiaphysis (75%)
- Periosteal reaction—may look benign
  - Interrupted or solid single layer (66%)
  - Onion Skin 10%
  - Sunburst 2%
- Soft tissue mass—by CT (80%); by MRI (100%)
Lymphoma

- Pathologic Fracture (22%)
- Sequestra (16%)
- Cross Joint (5%)
- Diff Dx:
  - Metastatic Lymphoma
  - Ewings
  - Neuroblastoma
  - Rhabdomyosarcoma
  - Osteomyelitis
  - Eosinophilic Granuloma
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Lymphoma
Lymphoma Reticulin
Myeloma

- Plasma Cell Dyscrasia
- Age: > 50
- Clinical: pain, osteopenia, monarthropathy, pathologic fracture, bleeding diathesis, infection, renal insufficiency
- Labs: monoclonal spike; Bence-Jones proteinuria, anemia, elevated sed rate, hypercalcemia
- 10% with coexisting amyloidosis
- Palsmacytoma: Solitary bone involvement
Myeloma

- Bone scan positive for 80% of lesions
- Skeletal Survey
- XR: Solitary lesion: Bubbly lytic lesion with any margin (may have soft tissue mass)
- Multiple: punched out lytic lesions; endosteal scalloping
- Generalized osteopenia
- Osteosclerotic Myeloma <3%--- associated with POEMS syndrome
Myeloma

- **POEMS Syndrome**
  - Polyneuropathy (100%)
  - Organomegaly (24%)
  - Endocrinopathy (39%)
  - Monoclonal gammopathy (52% IgA; 36% IgG;)
  - Skin Changes (58%)
Myeloma
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Osteosclerotic Myeloma
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