Small Round Blue Cell Tumors

Eosinophilic Granuloma
Ewing's Sarcoma
Lymphoma
Multiple Myeloma

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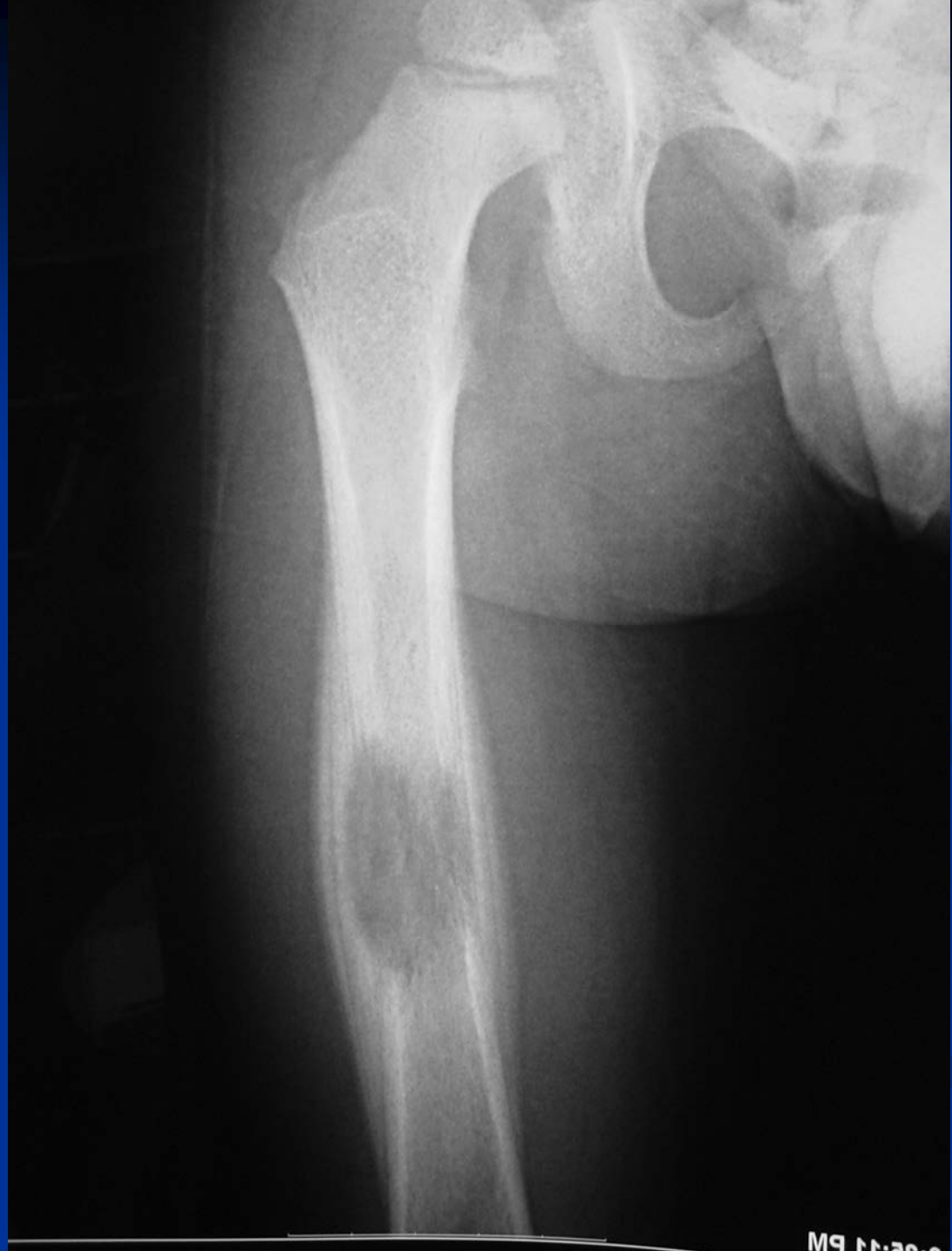
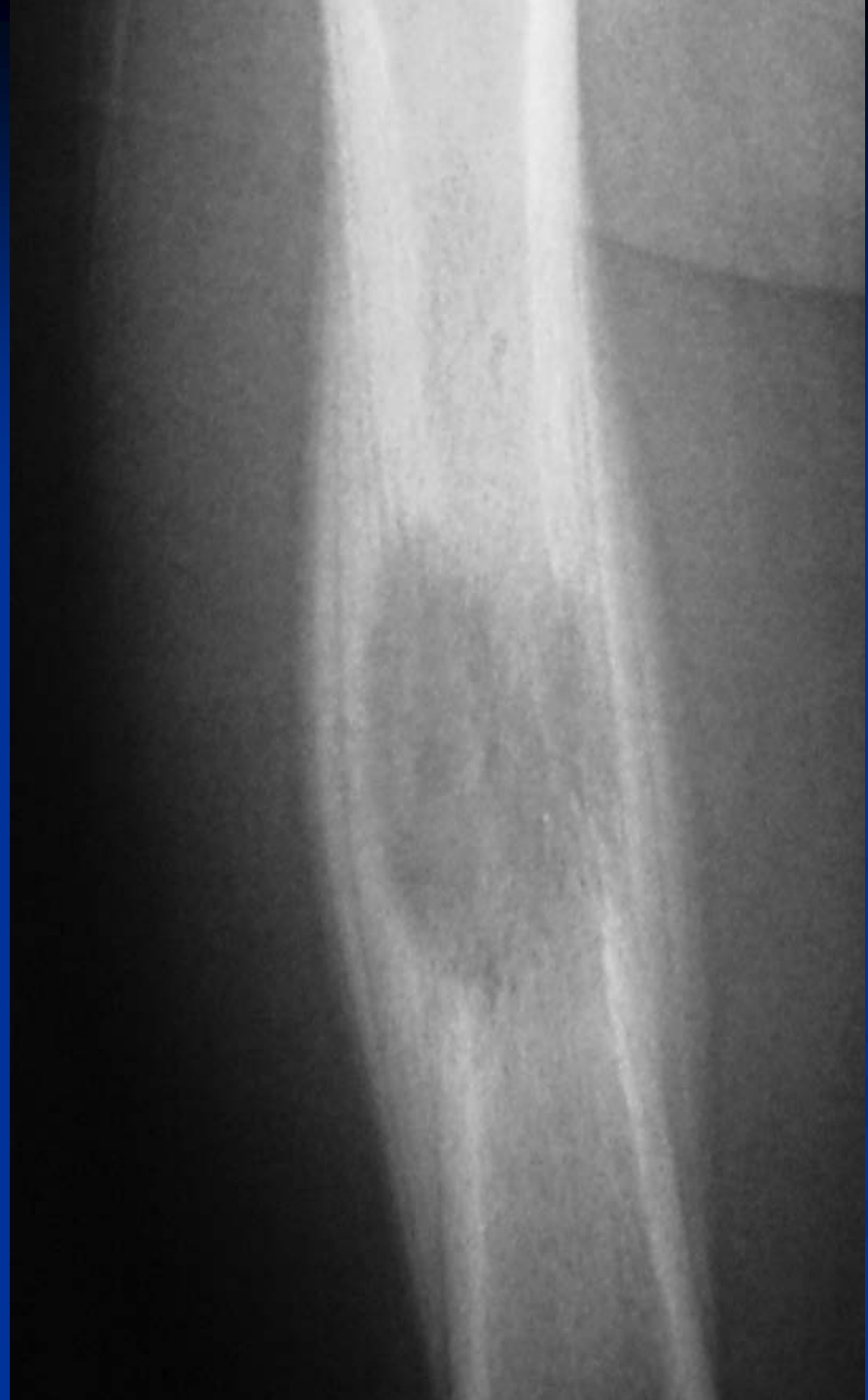
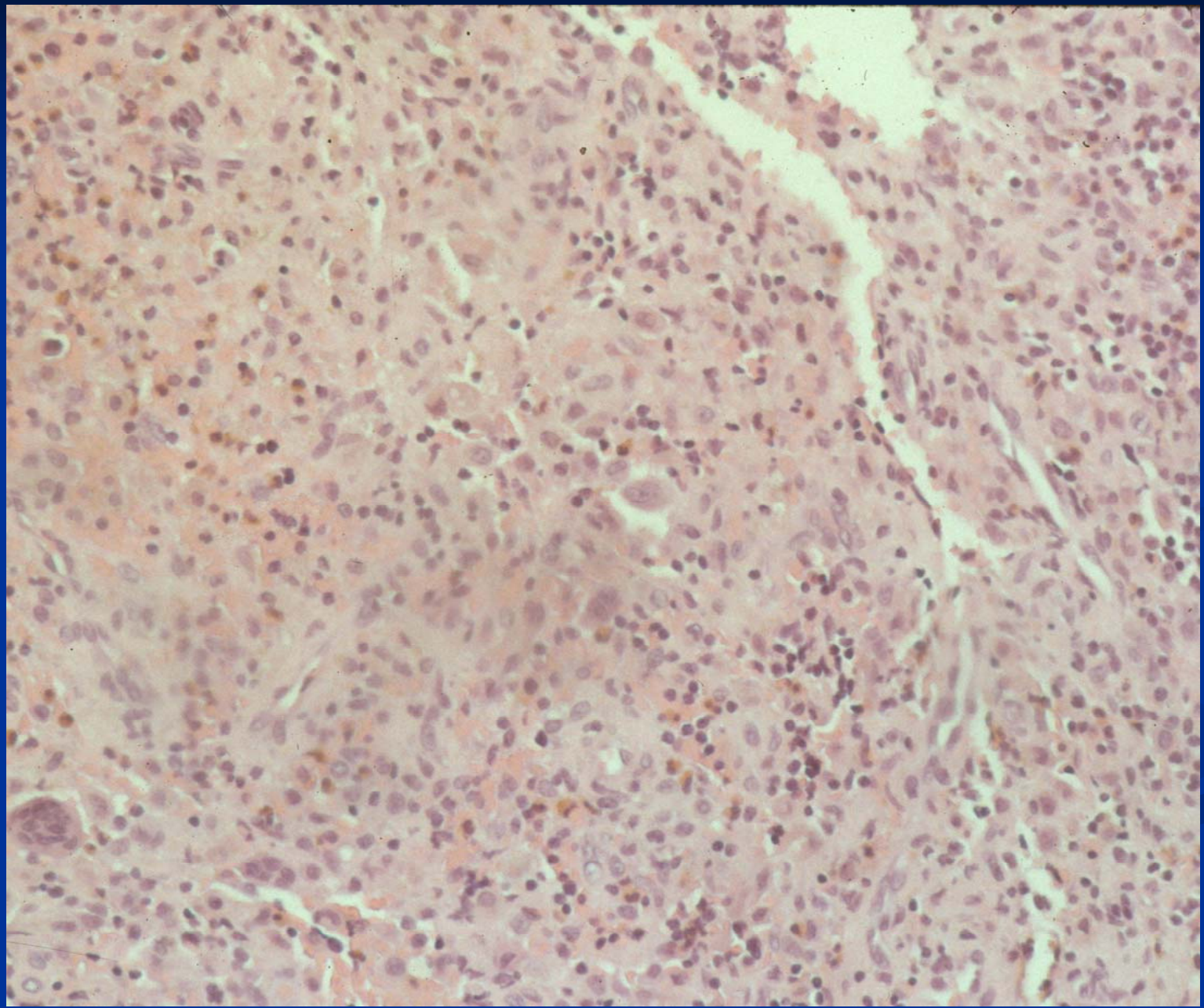
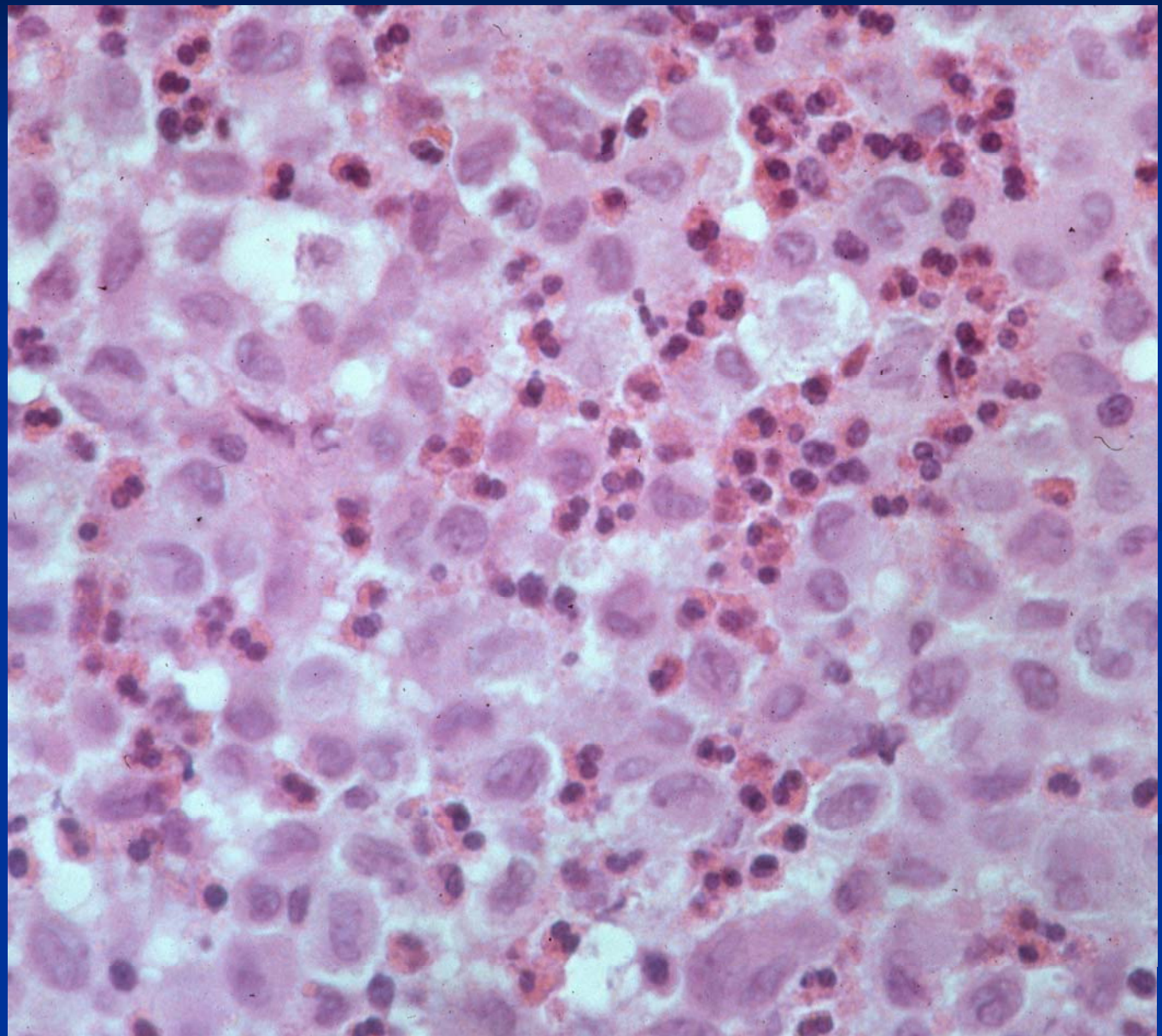
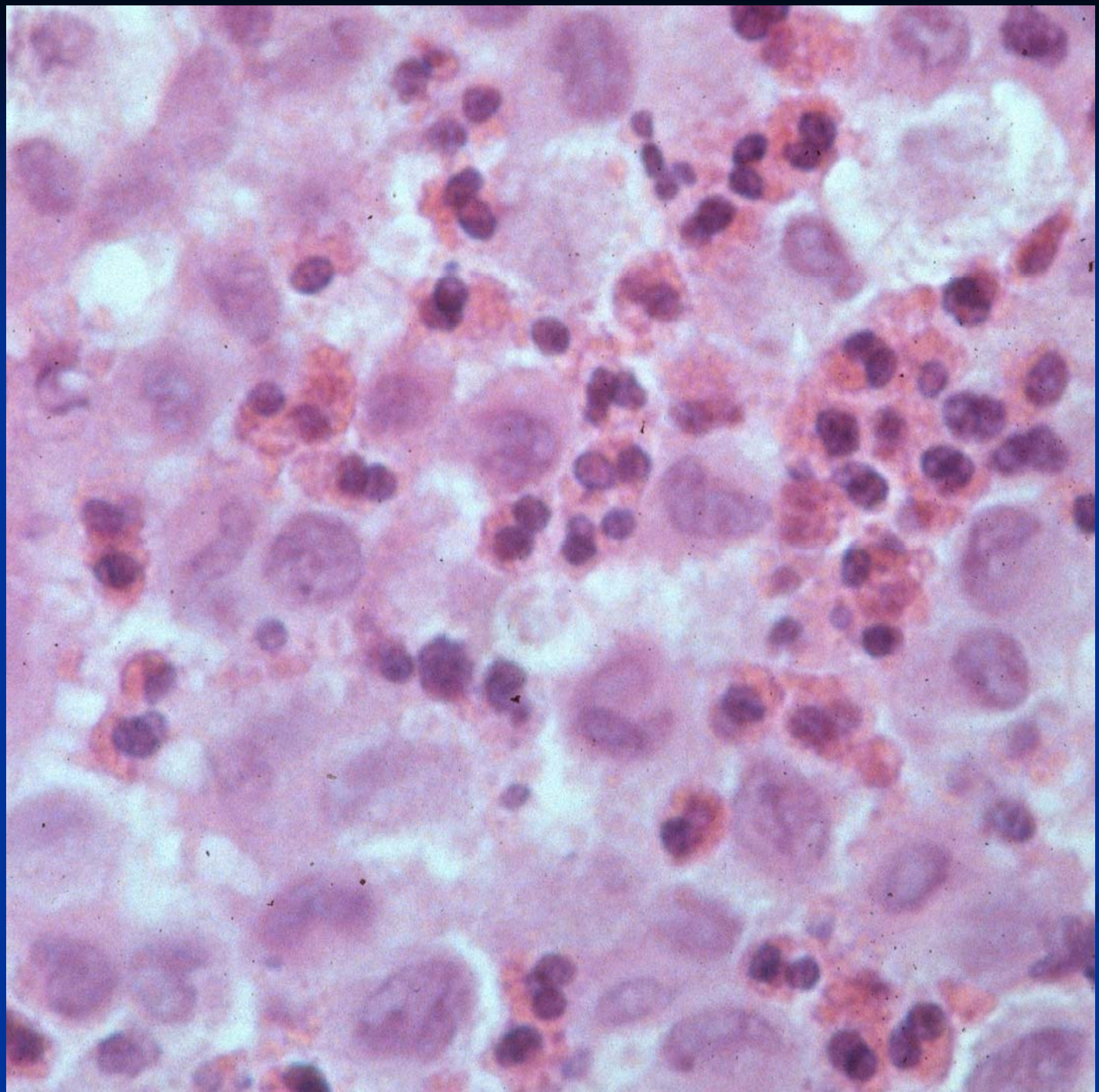
Unknown Case 1

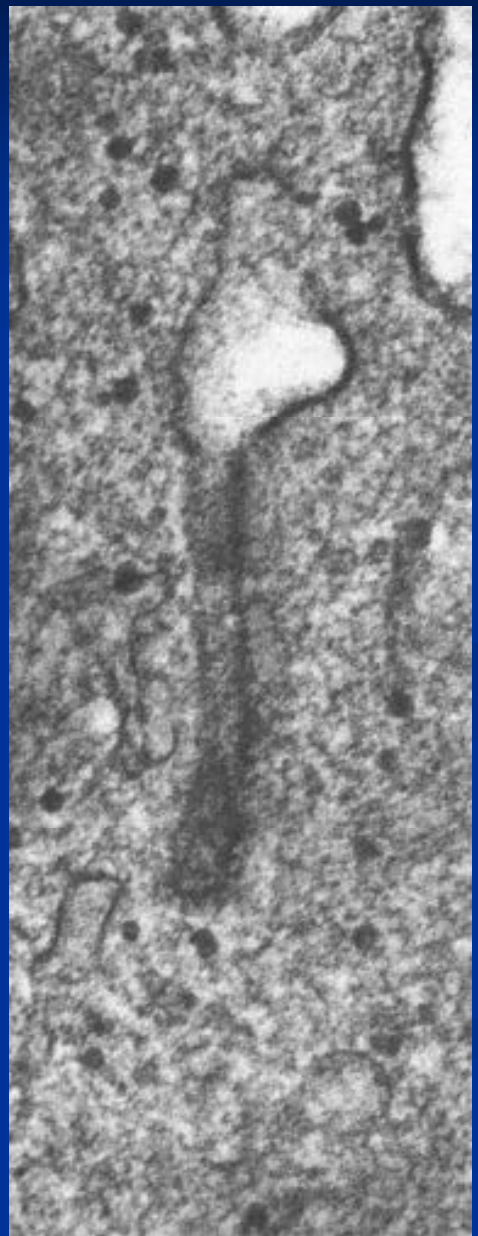












Answer

- Eosinophilic Granuloma

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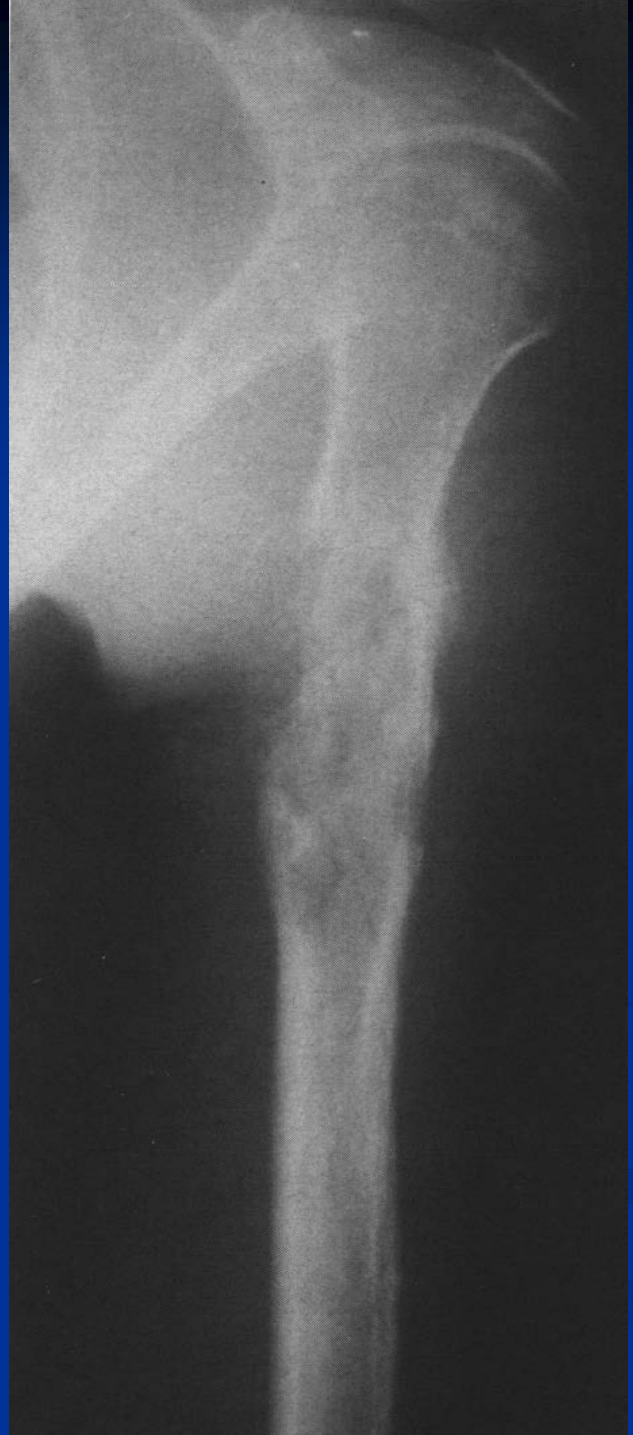
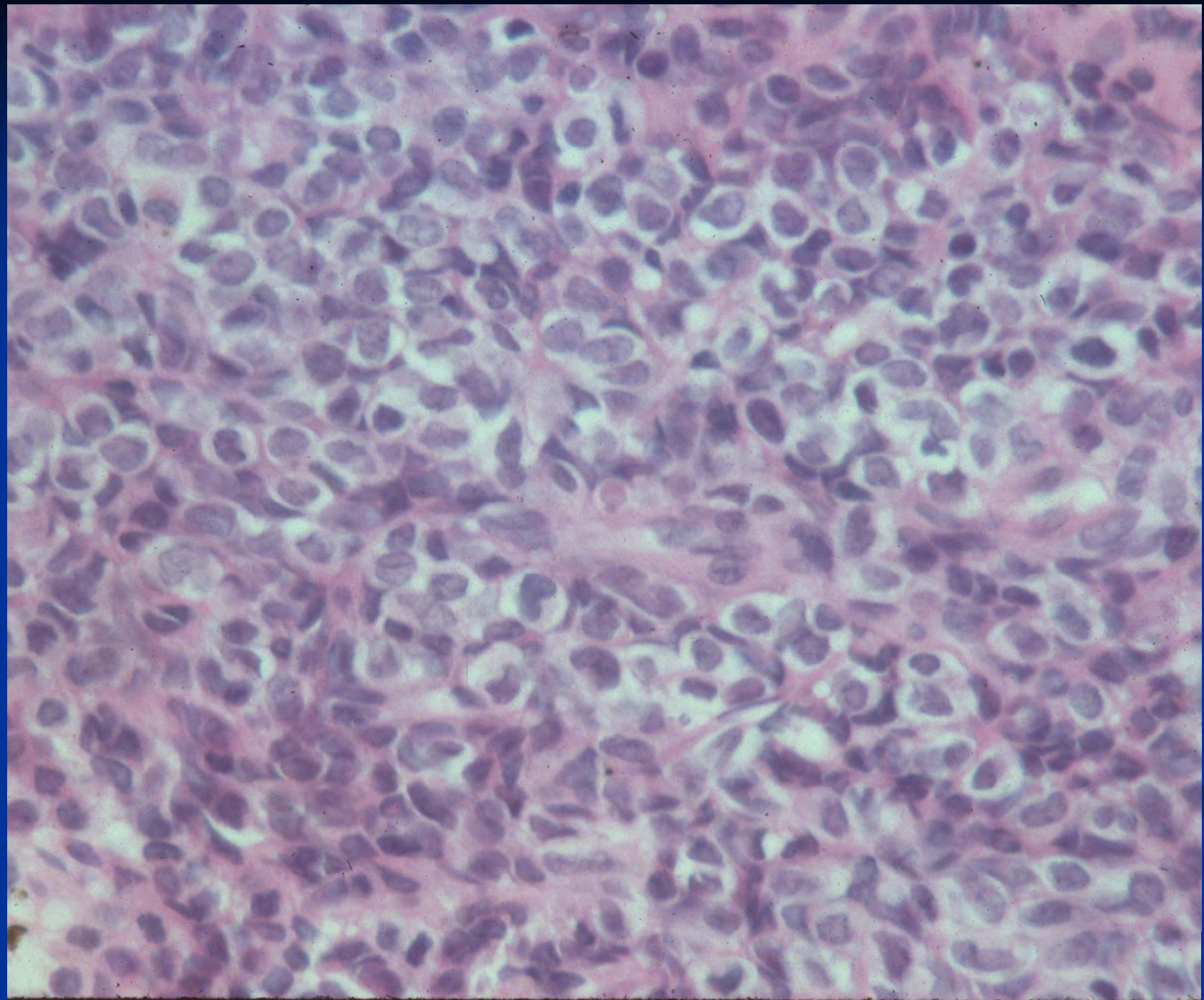
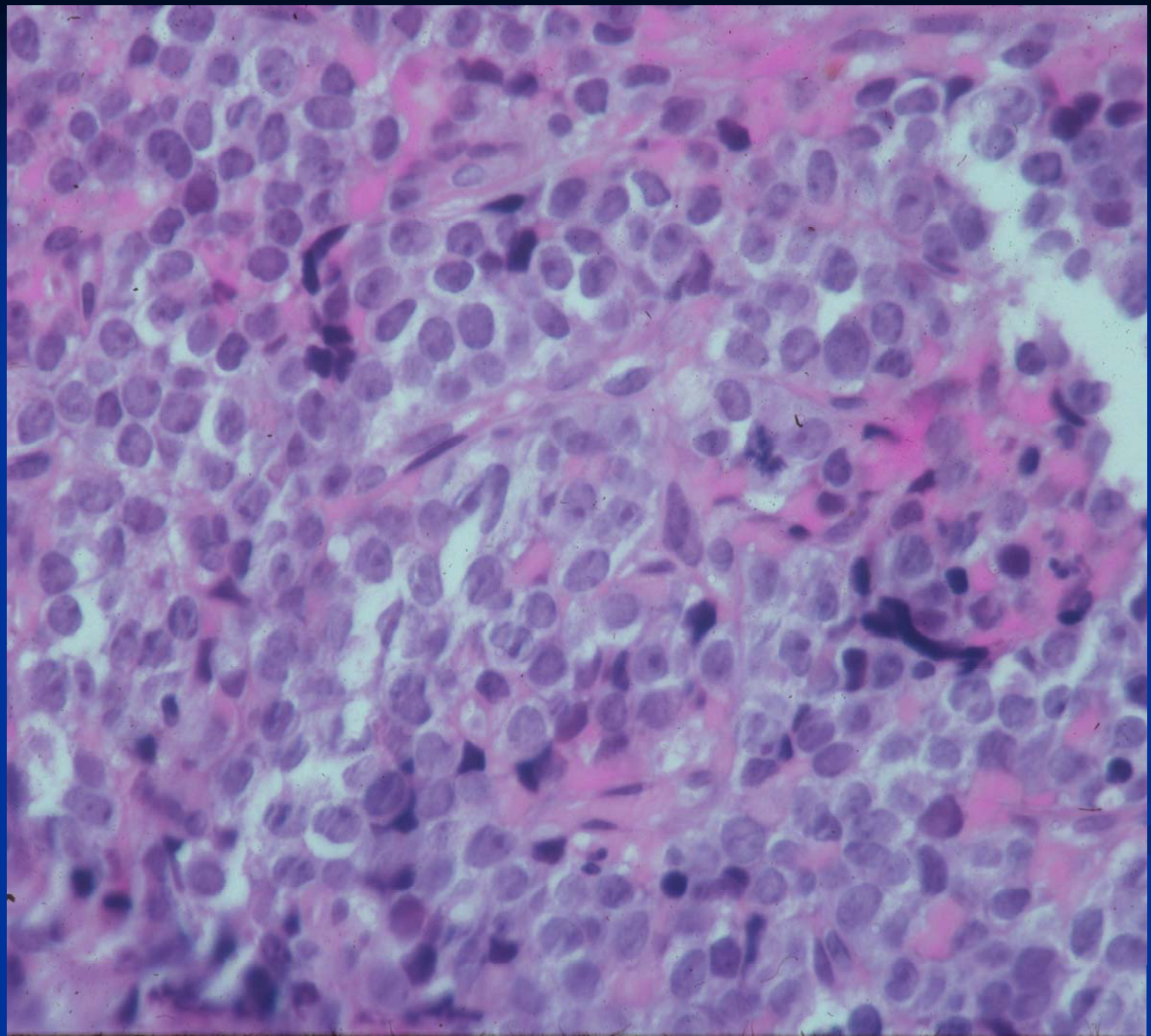
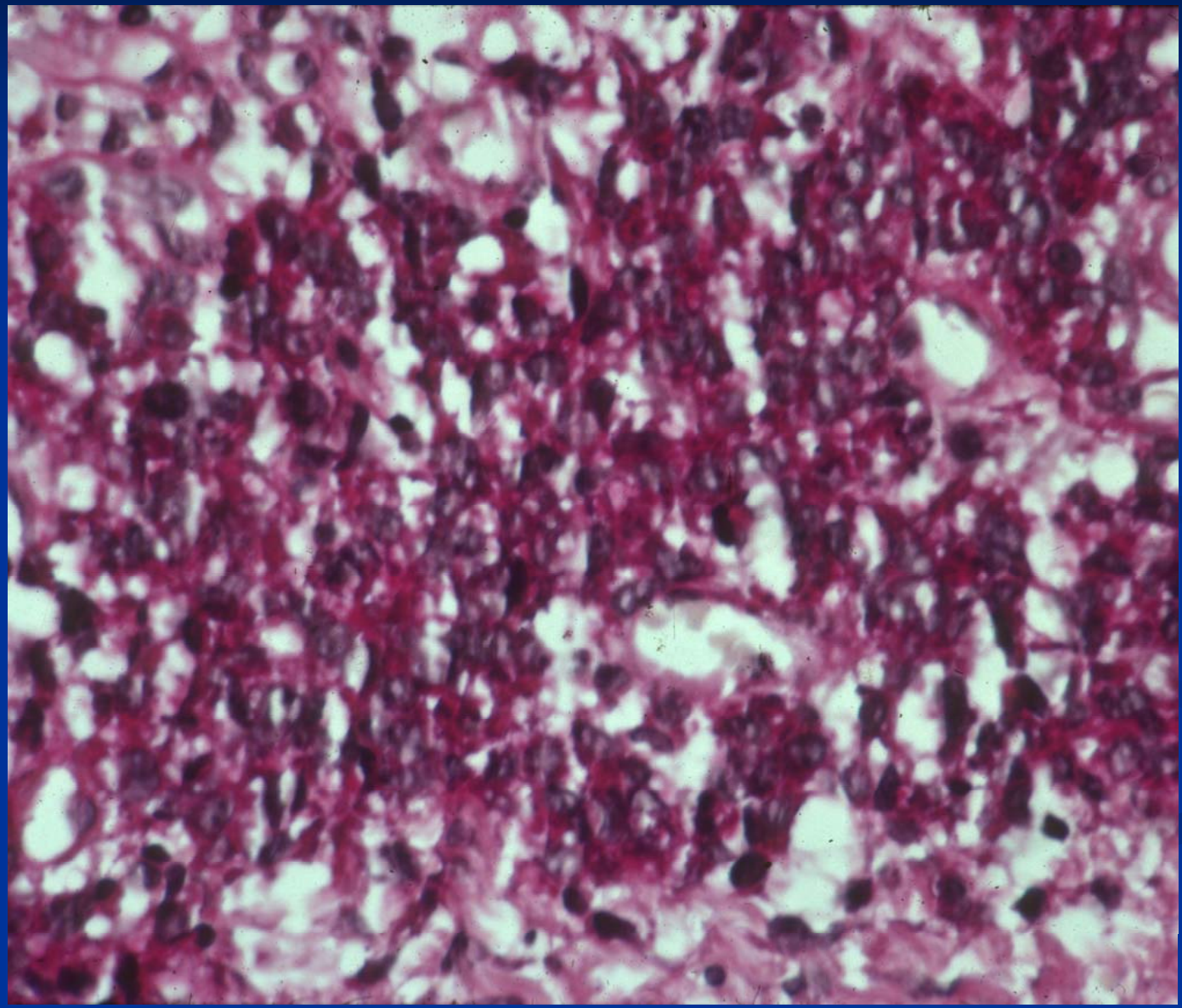






PAS Stain



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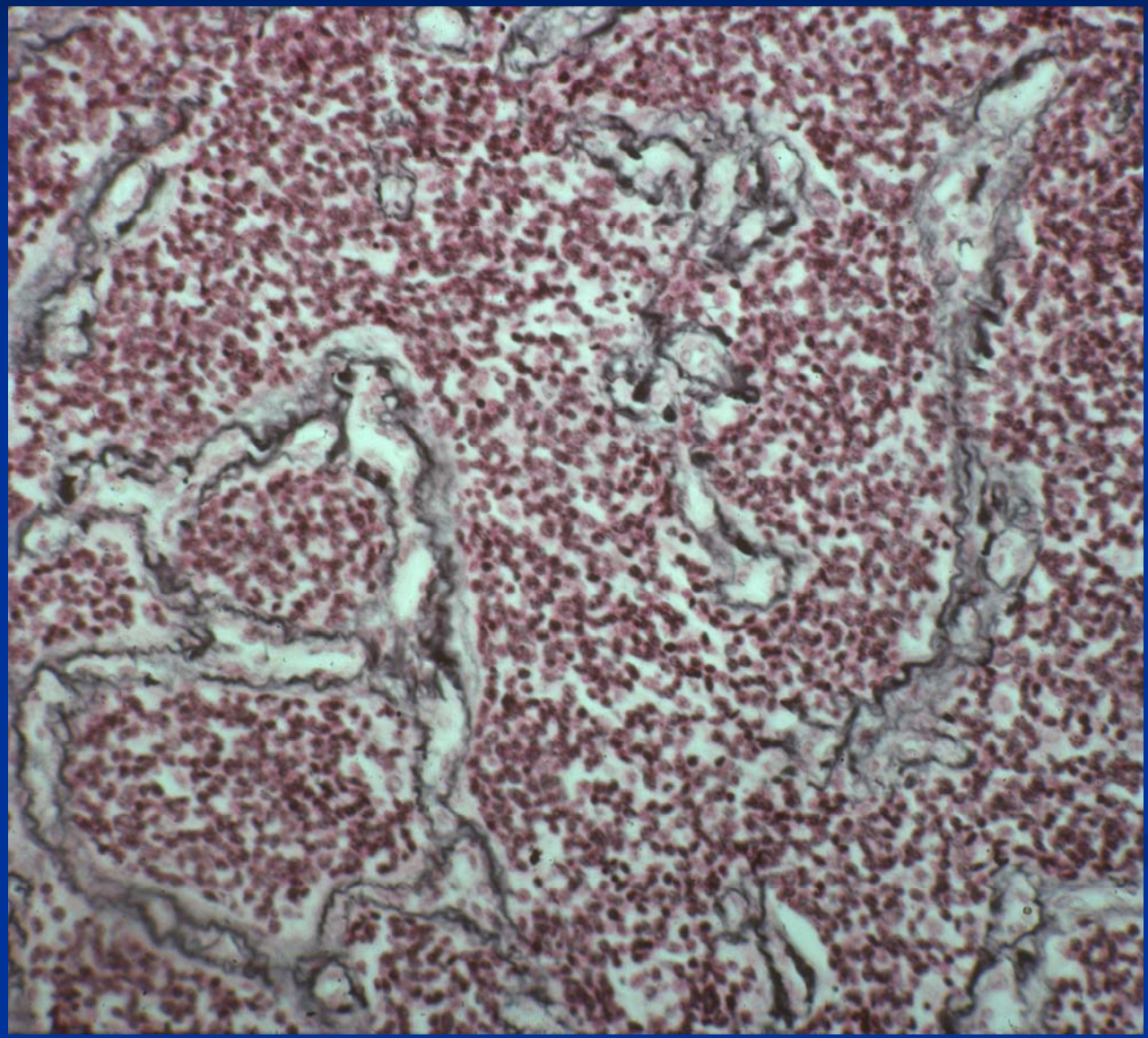

Answer

- 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 graftr
  - 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 1st year of life
- Disseminated disease and small bone lesions
- Fatal in 95% who develop before 1 year of life

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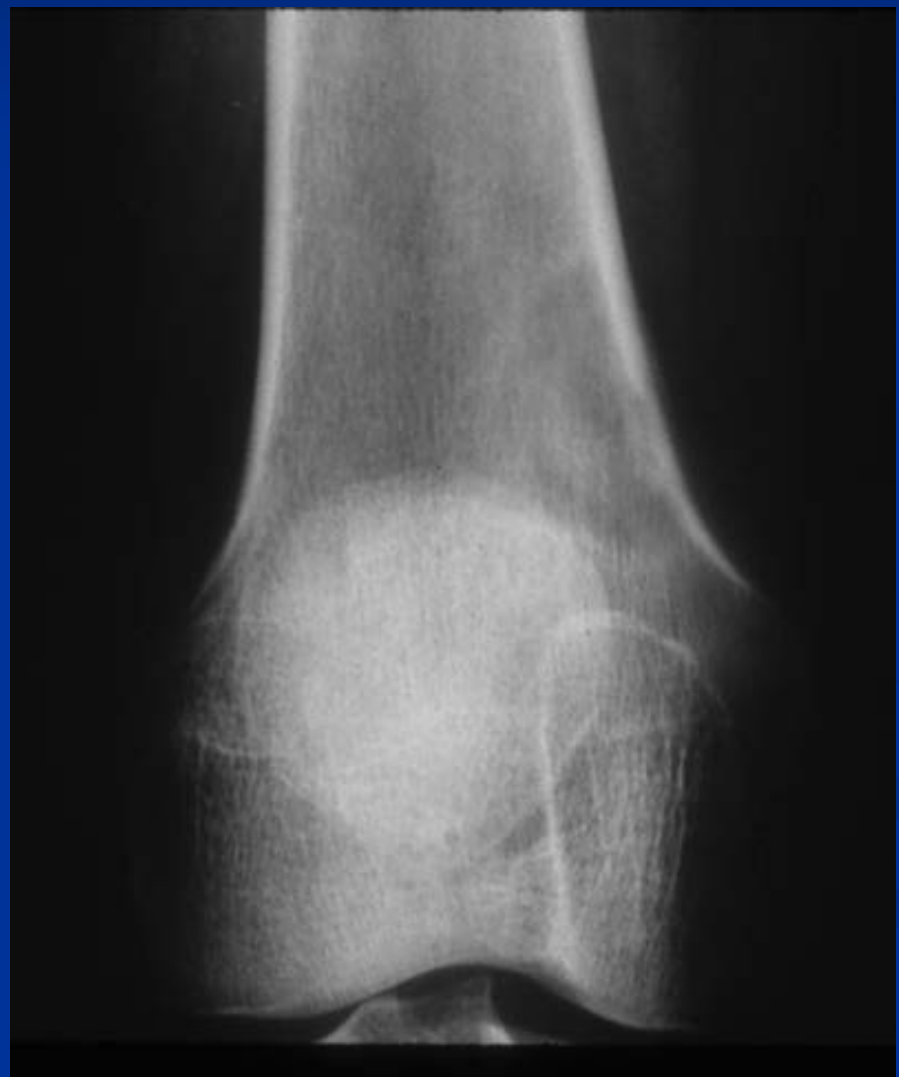

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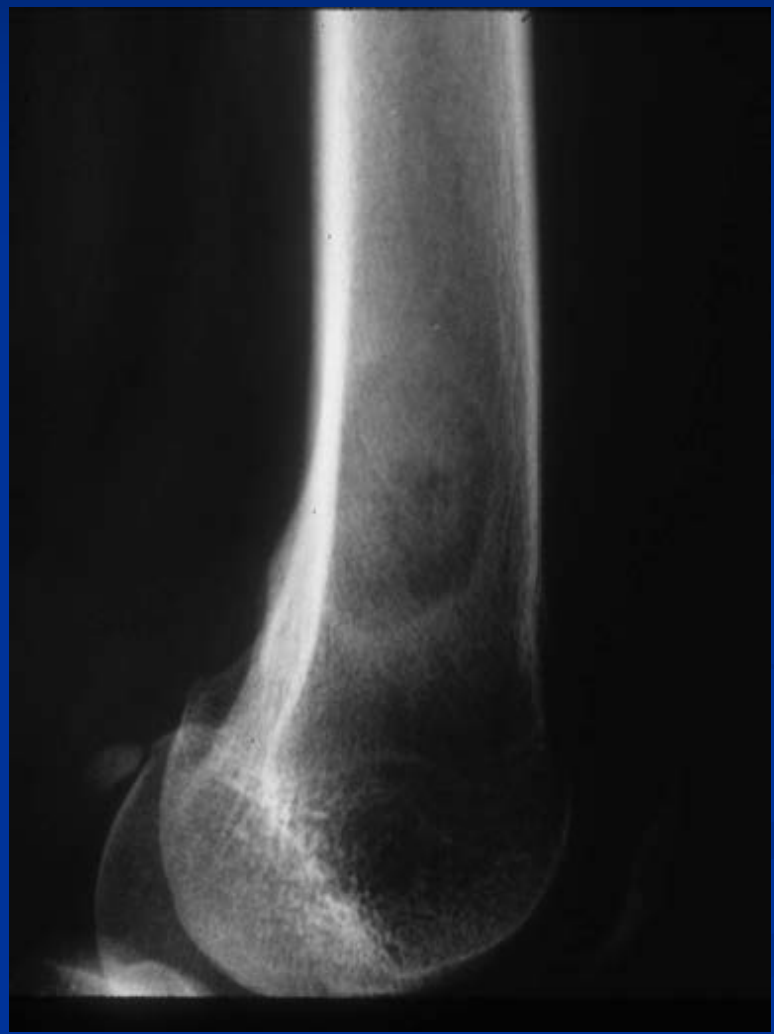

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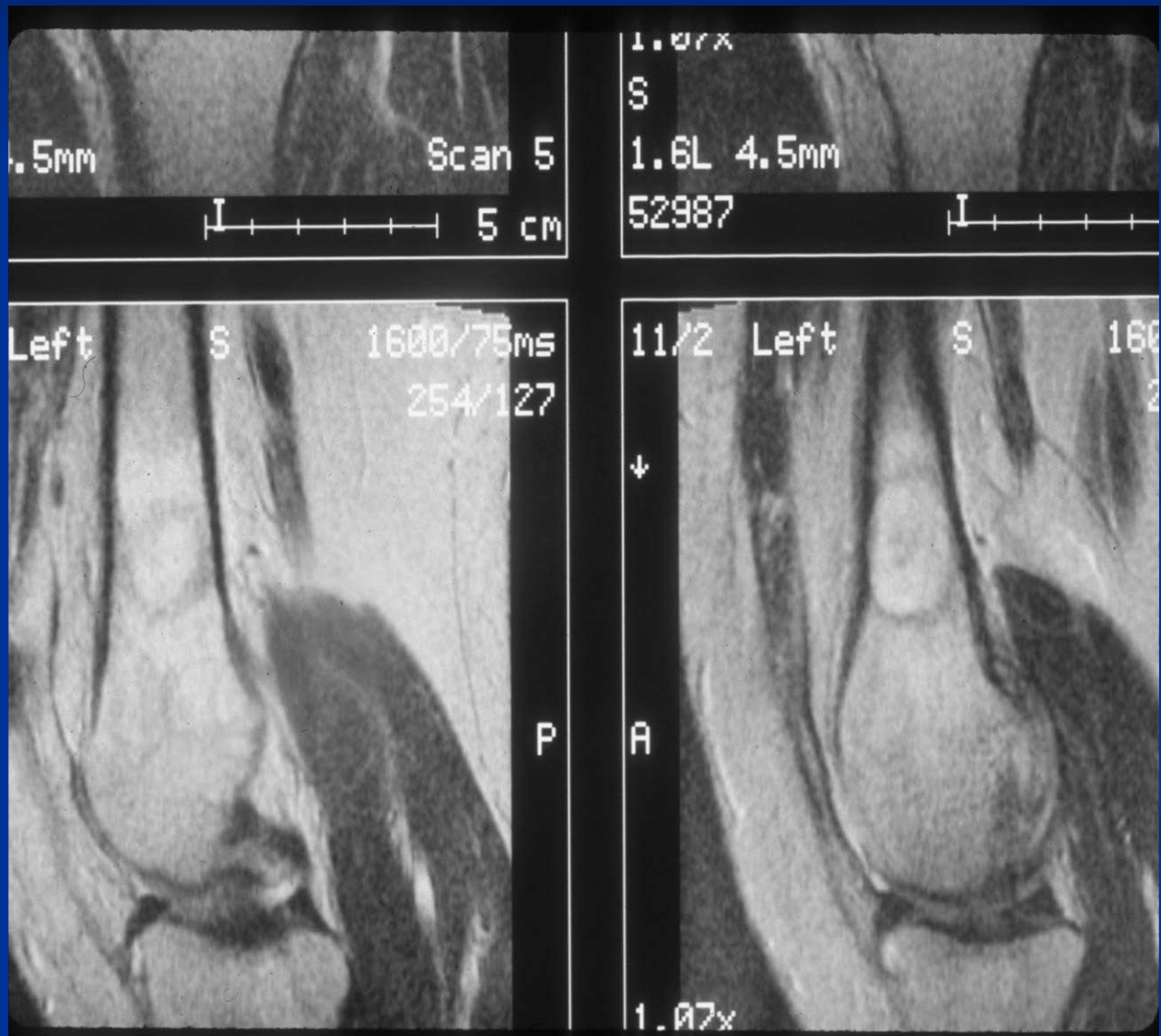
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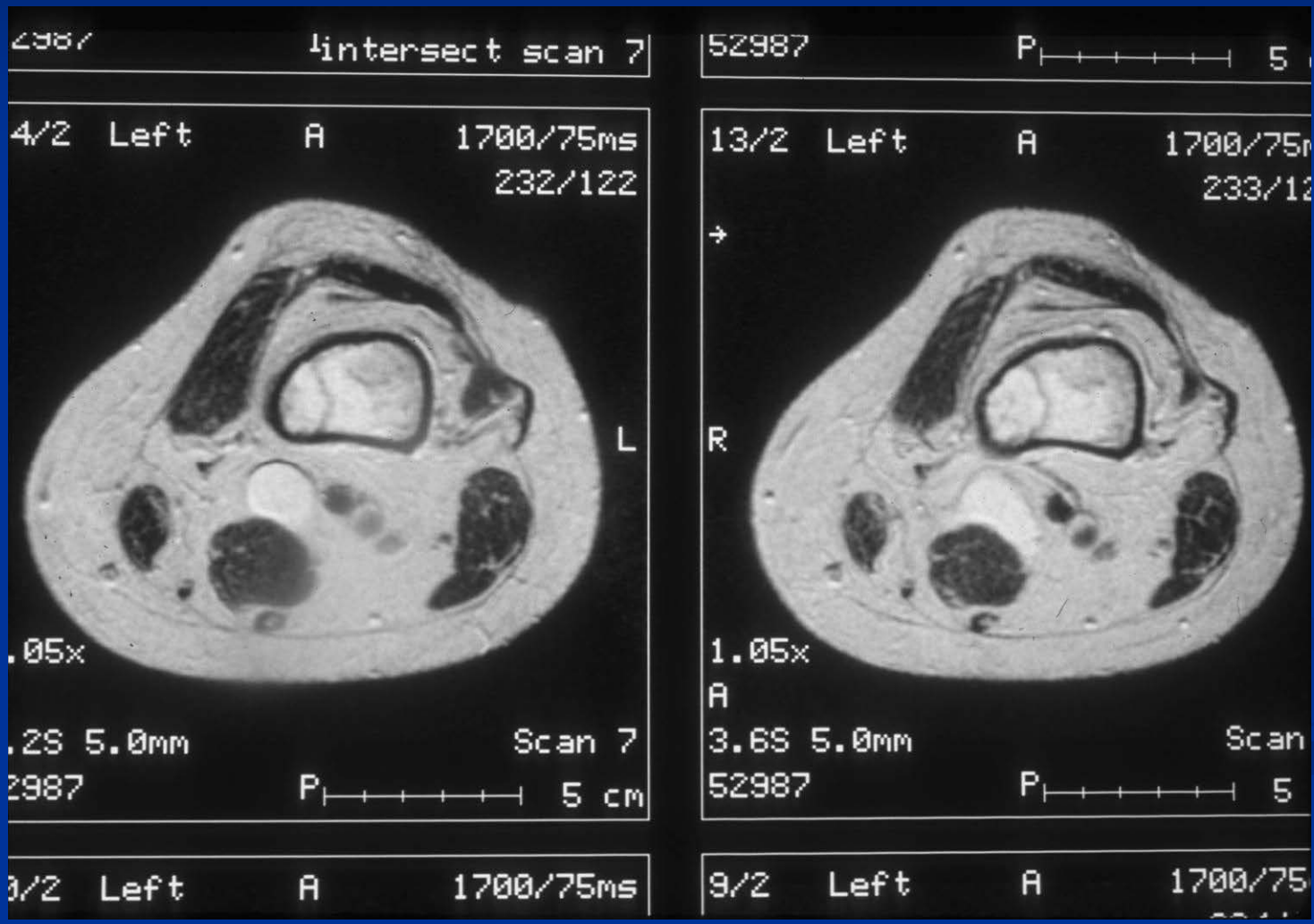
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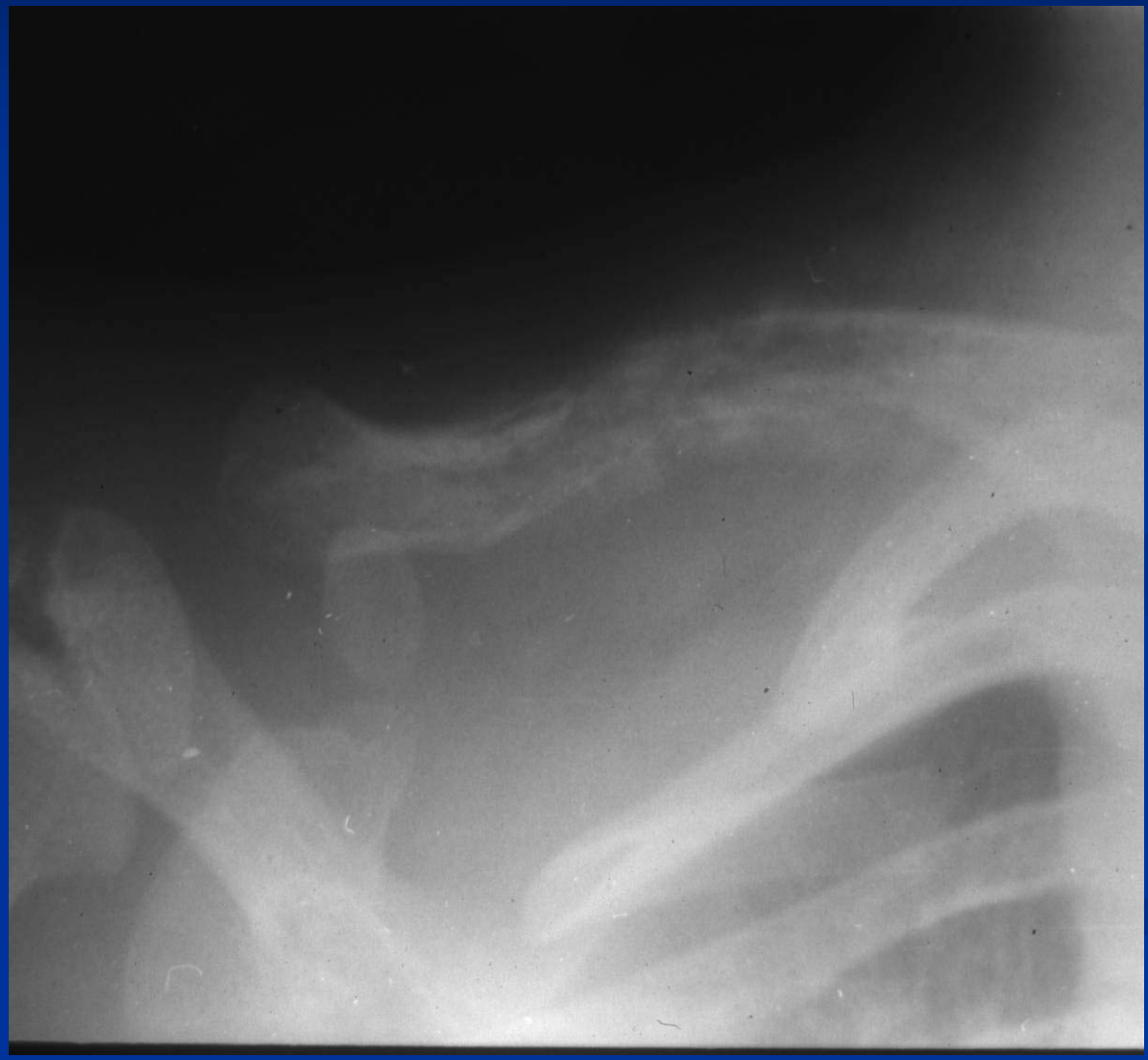

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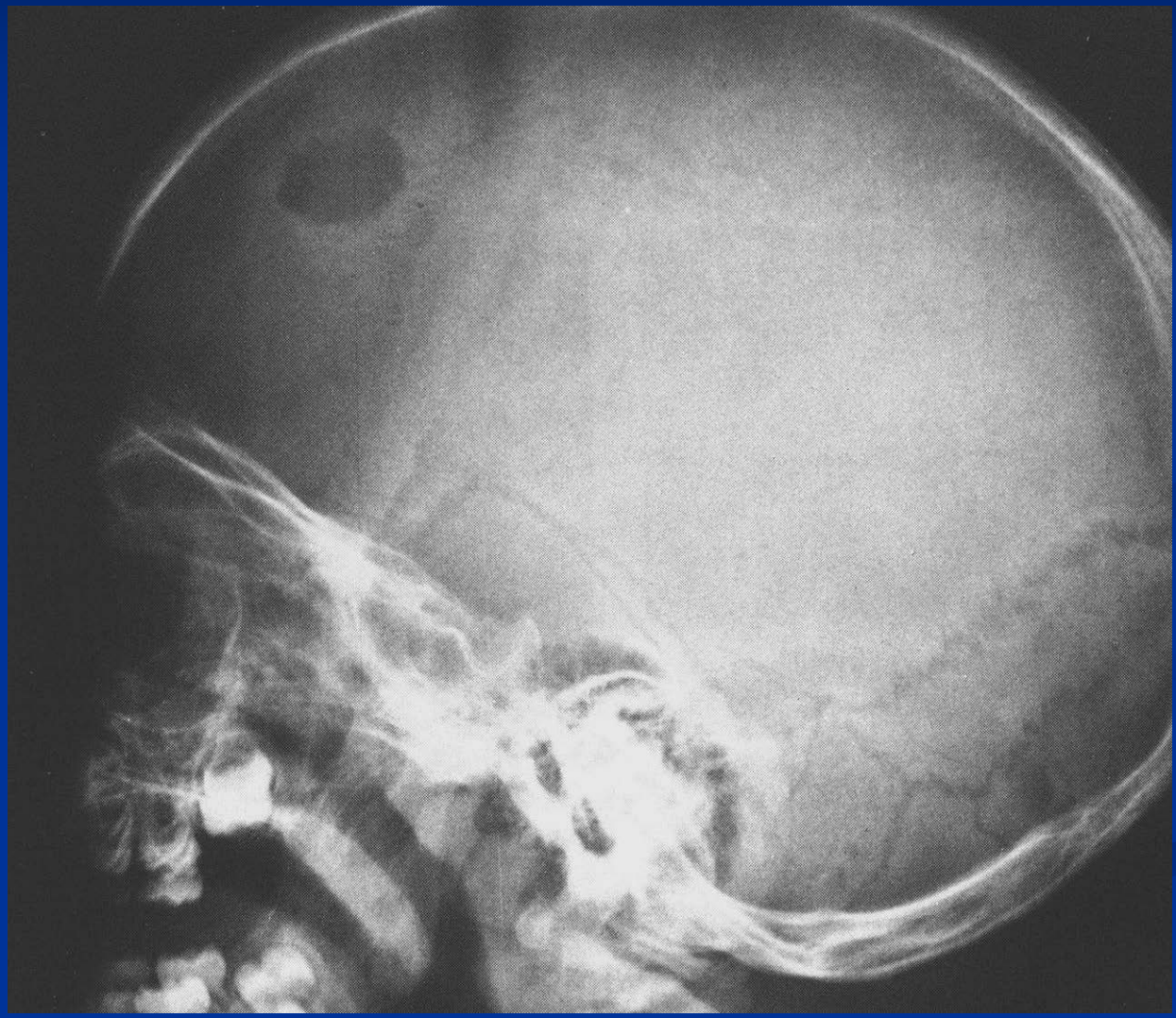

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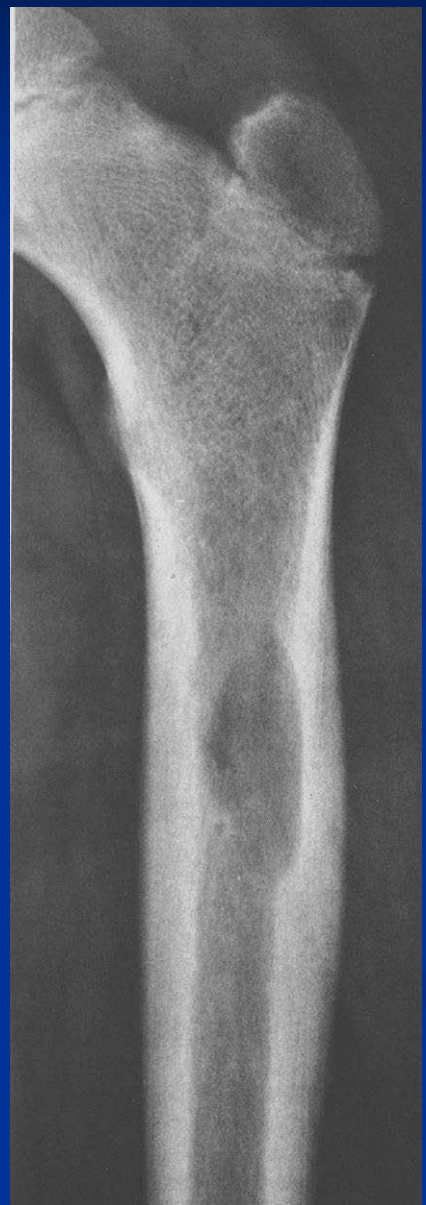

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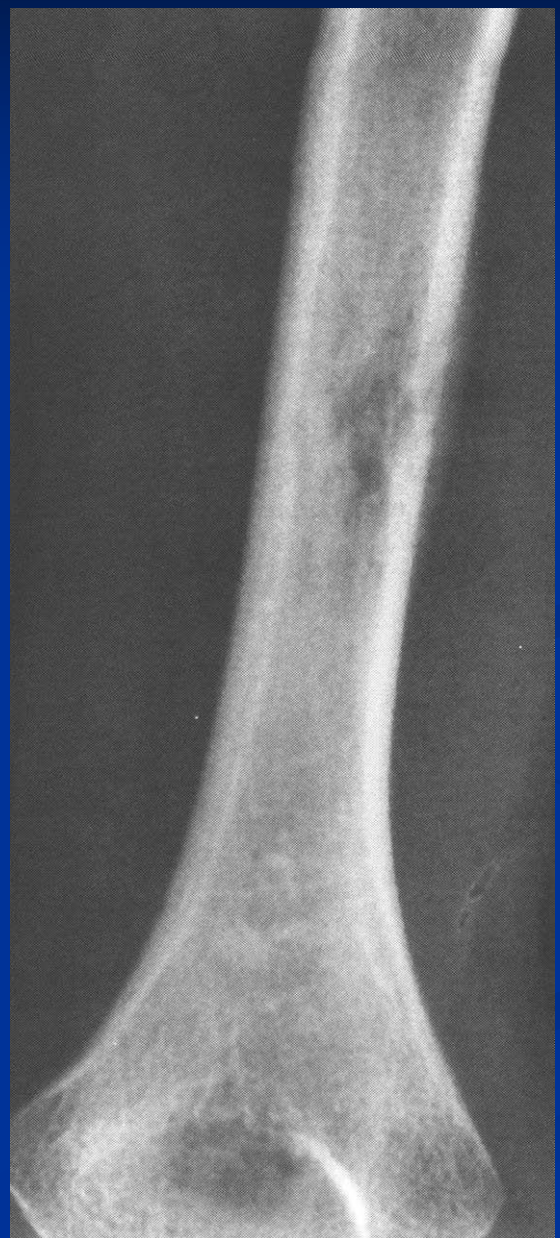

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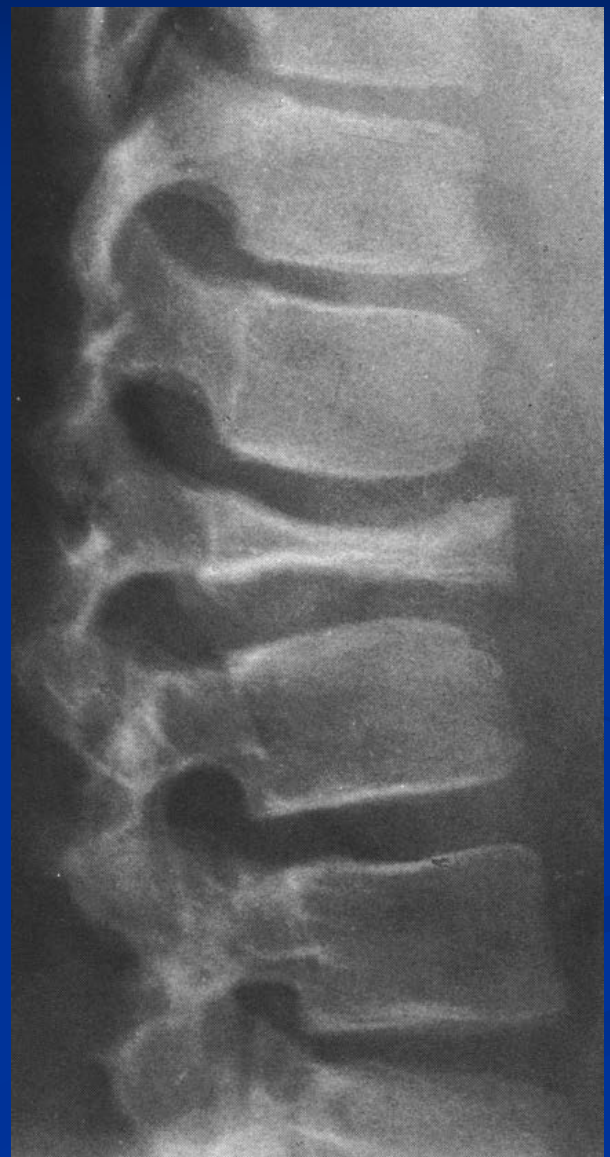

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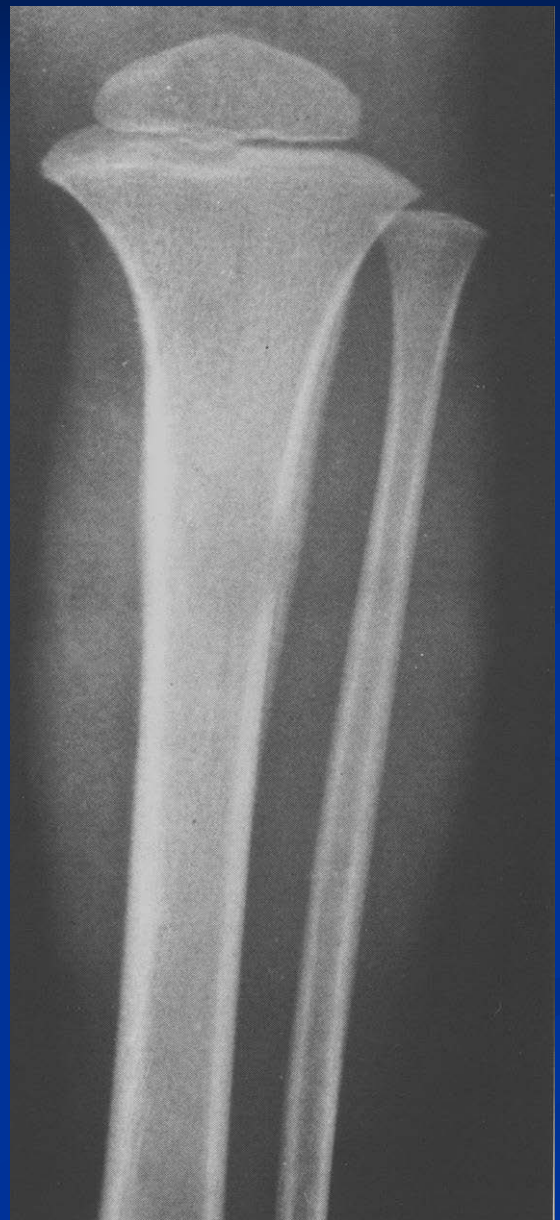

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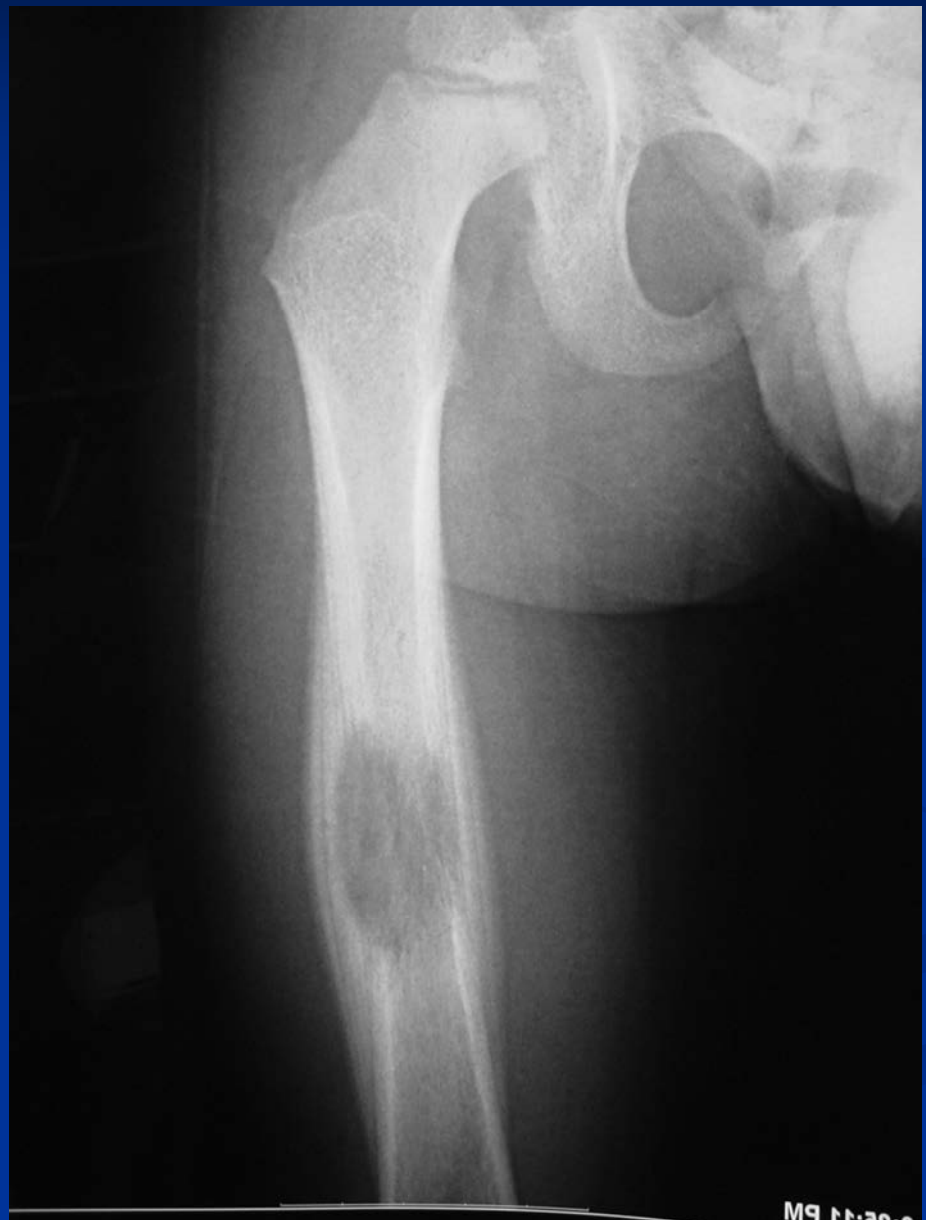

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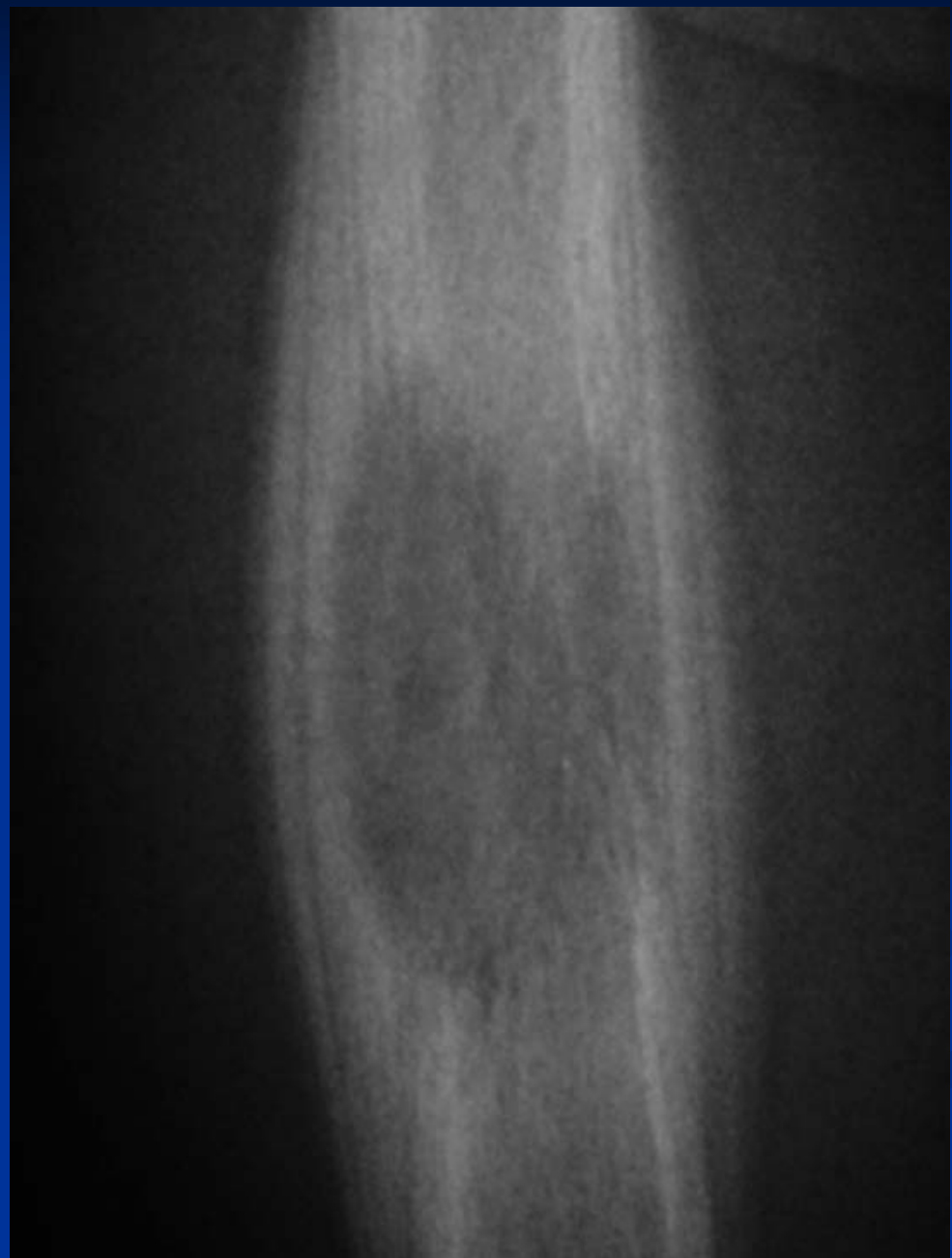

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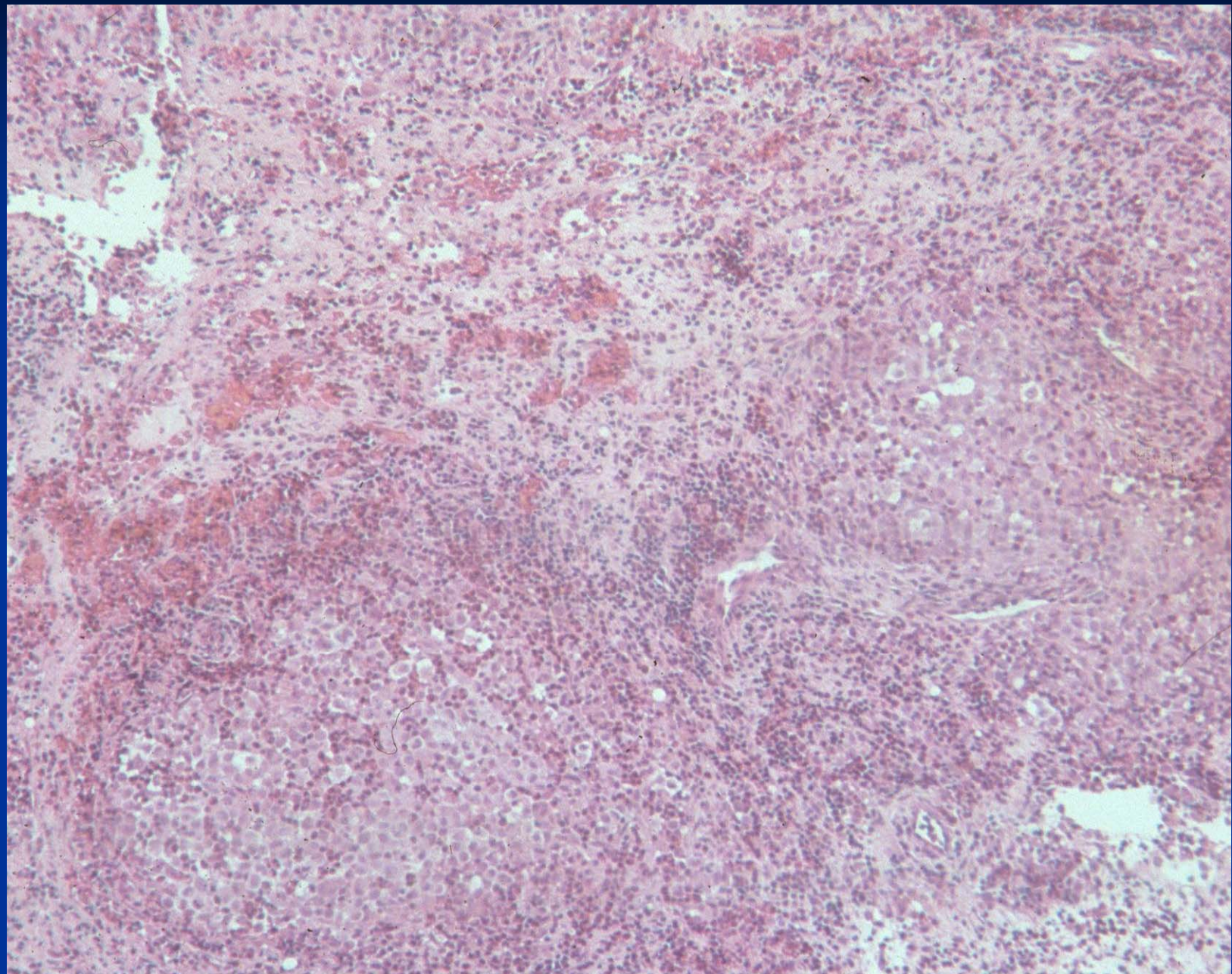

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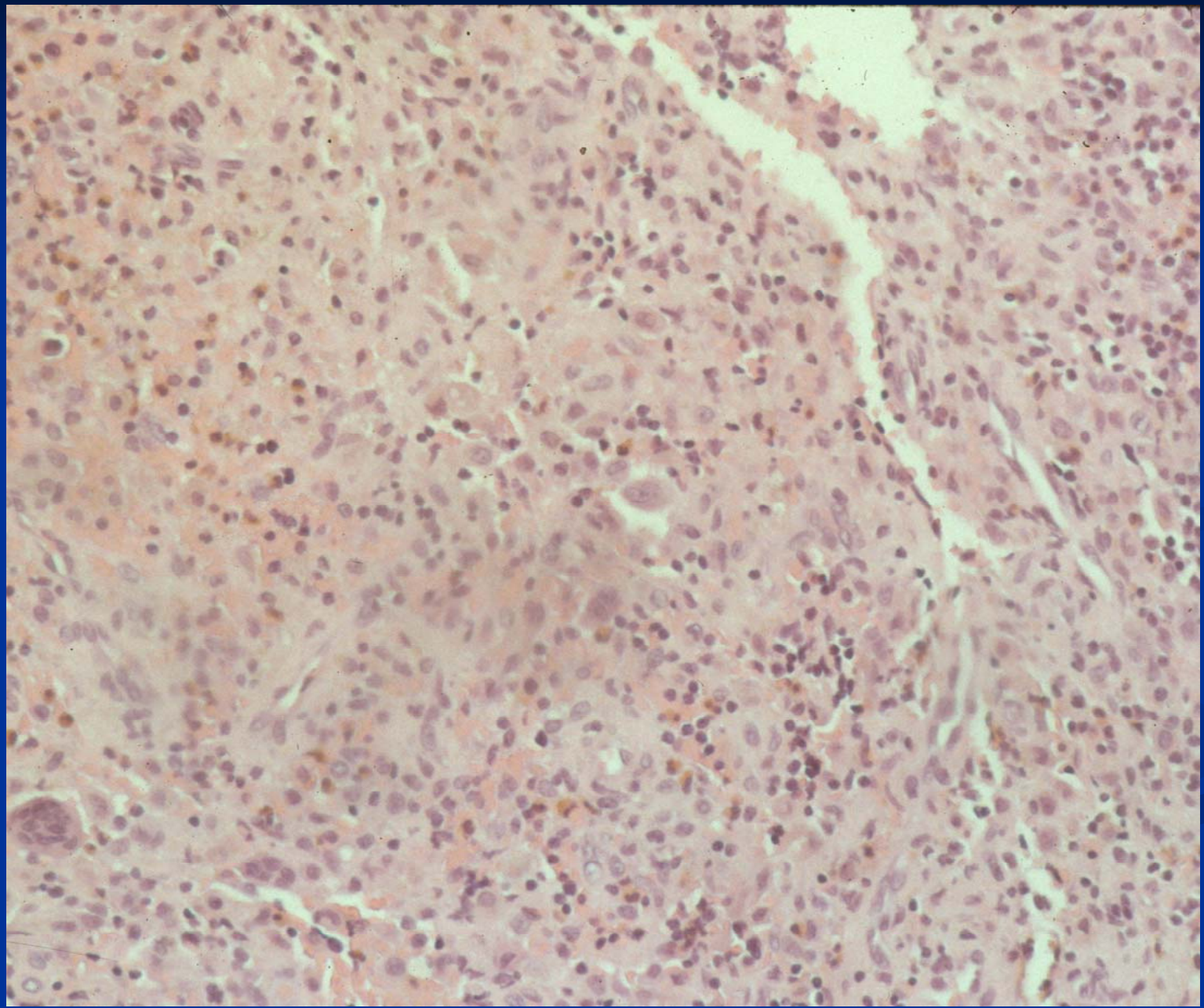

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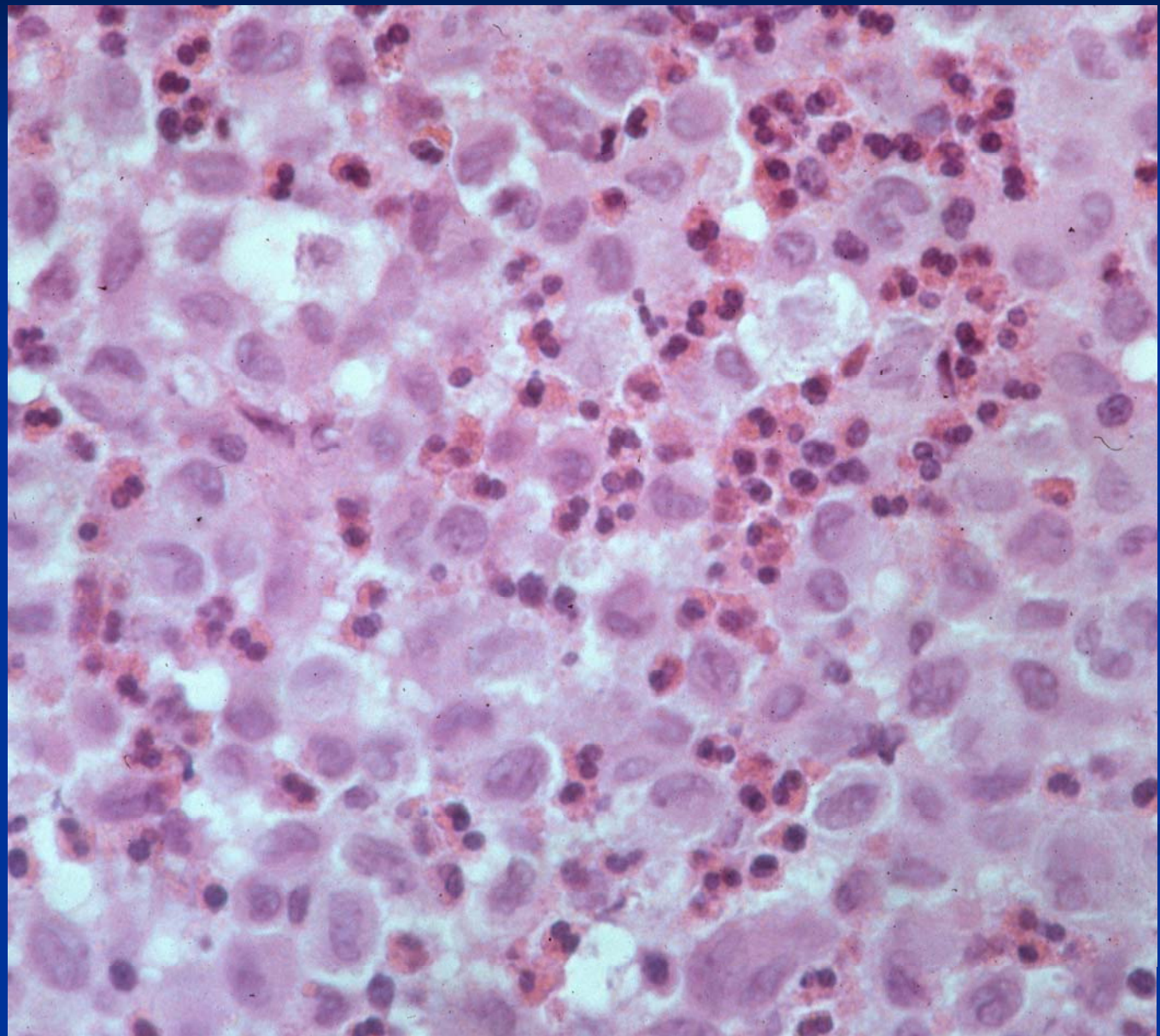

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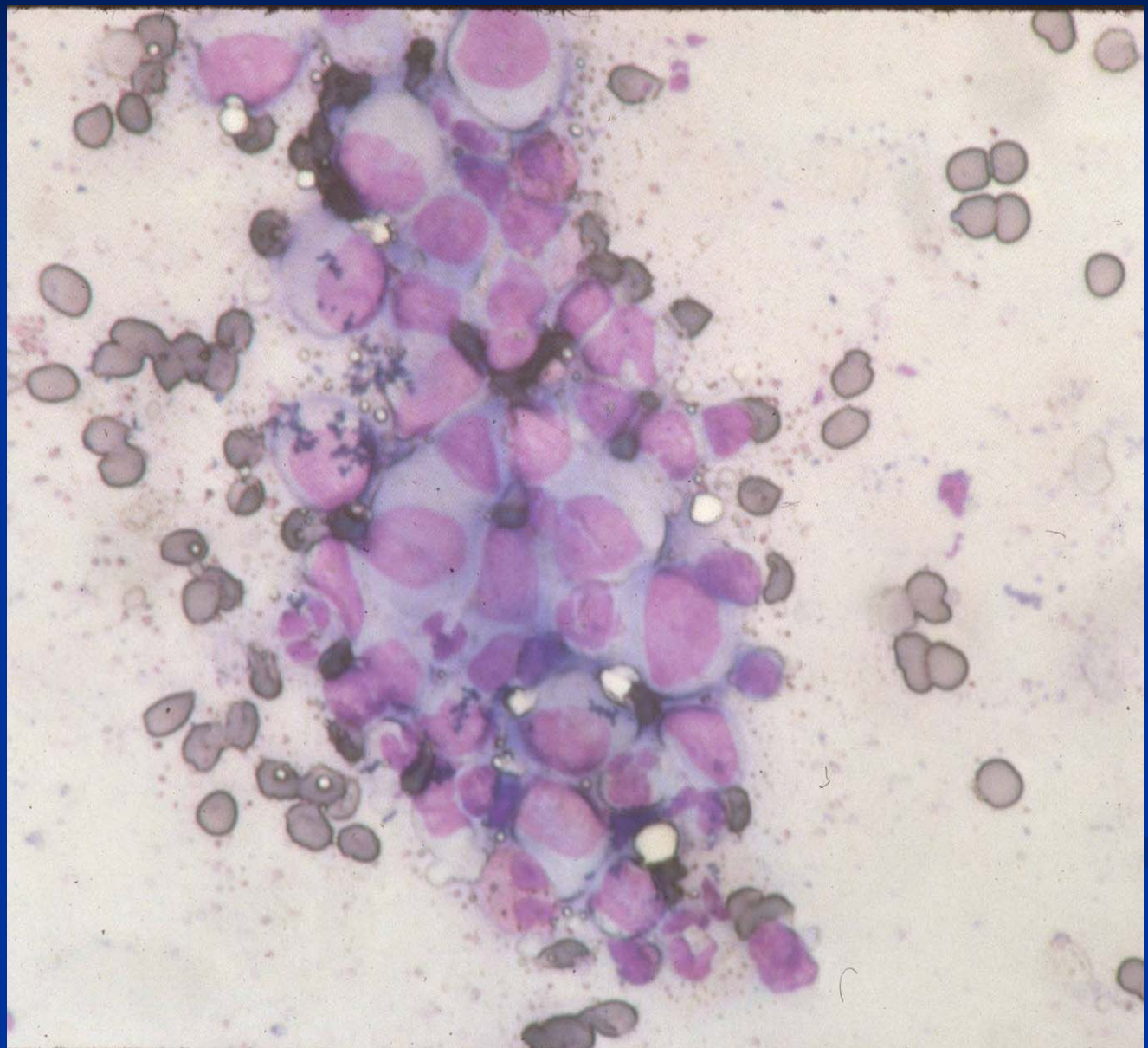

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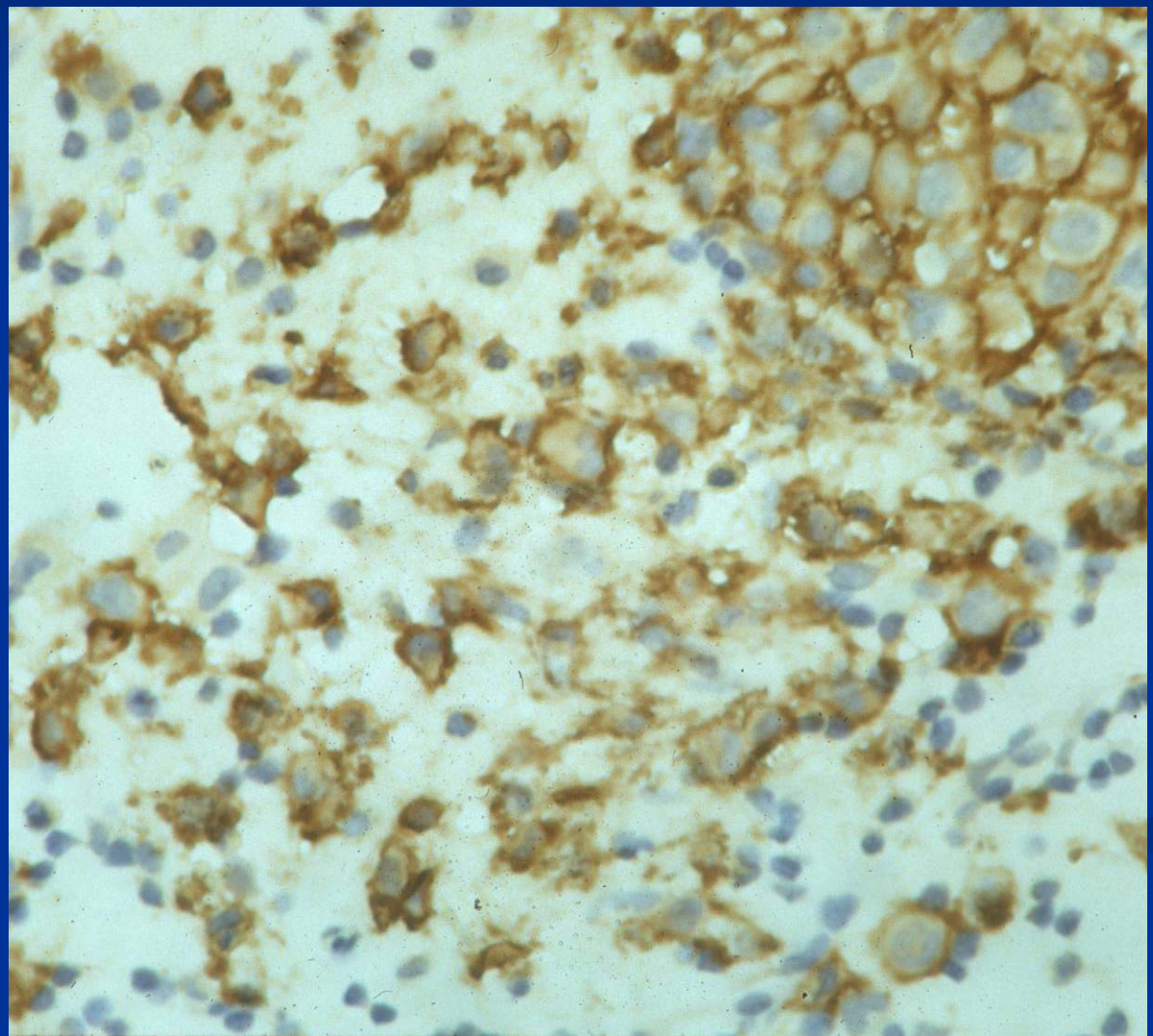

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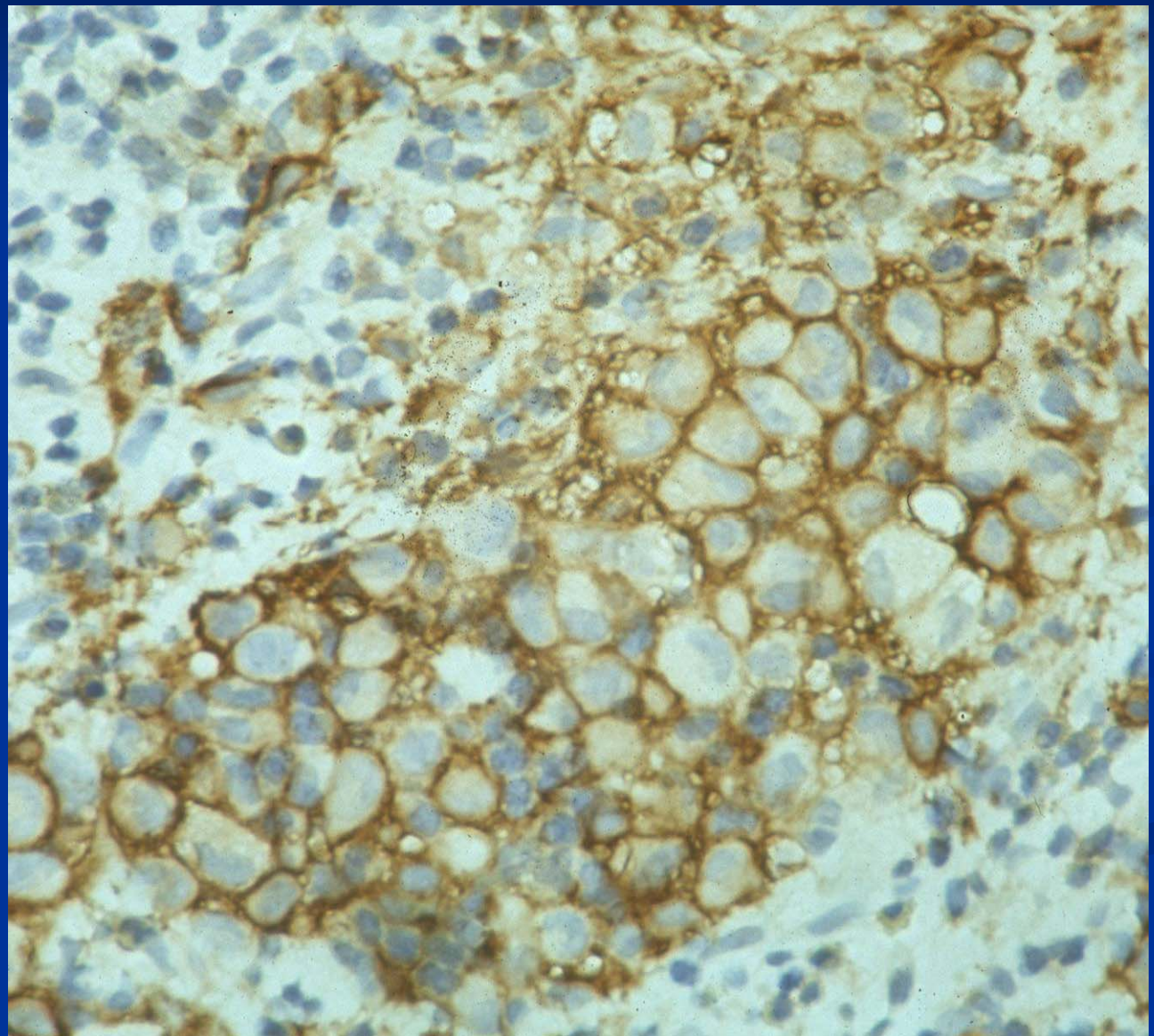

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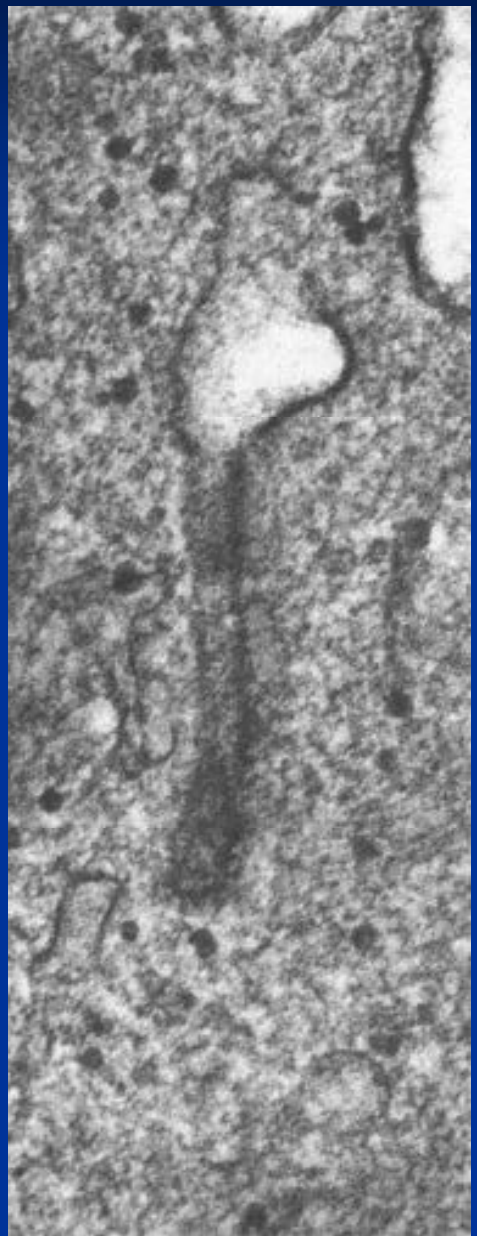
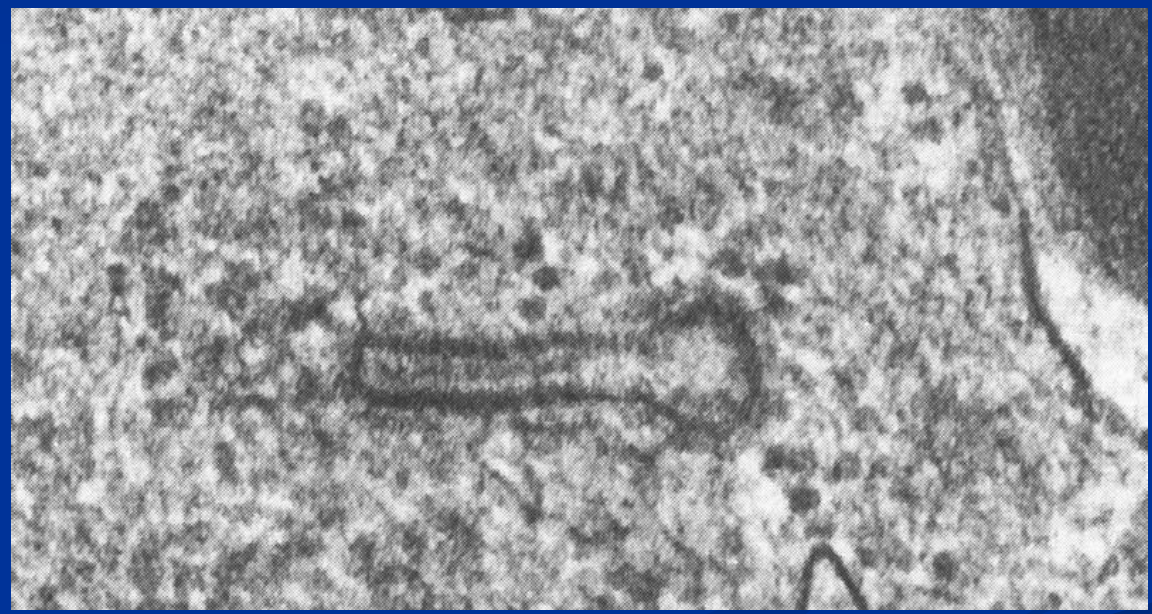




Ewing Sarcoma

- 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

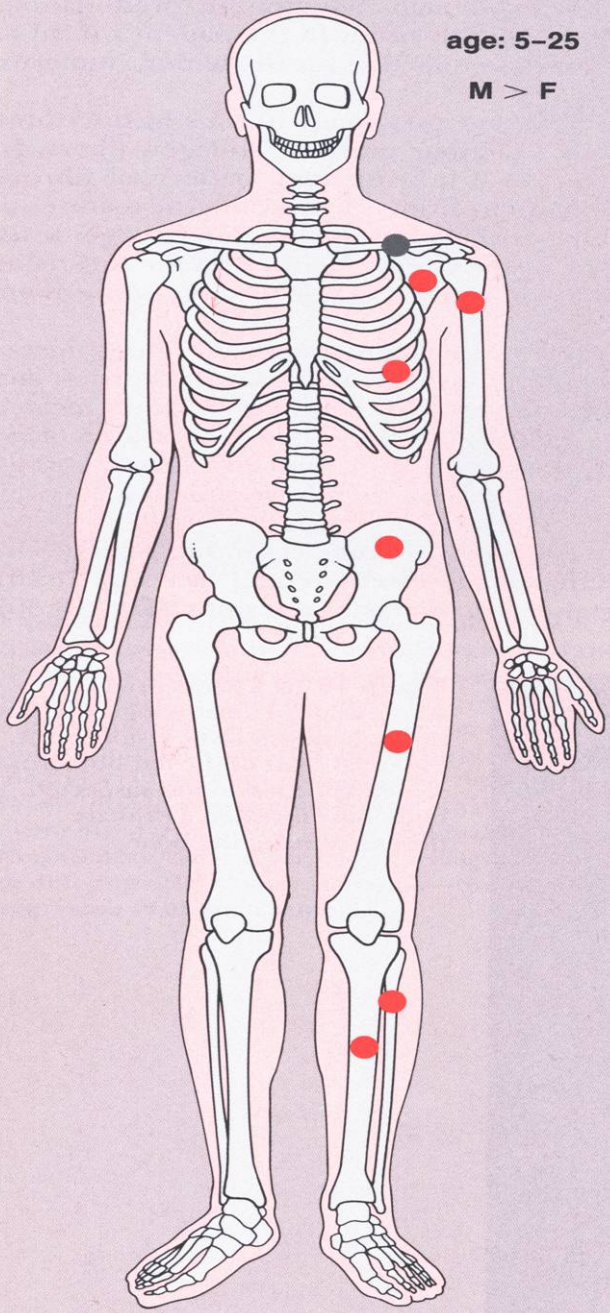
Ewing Sarcoma

- **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%

Ewing Sarcoma



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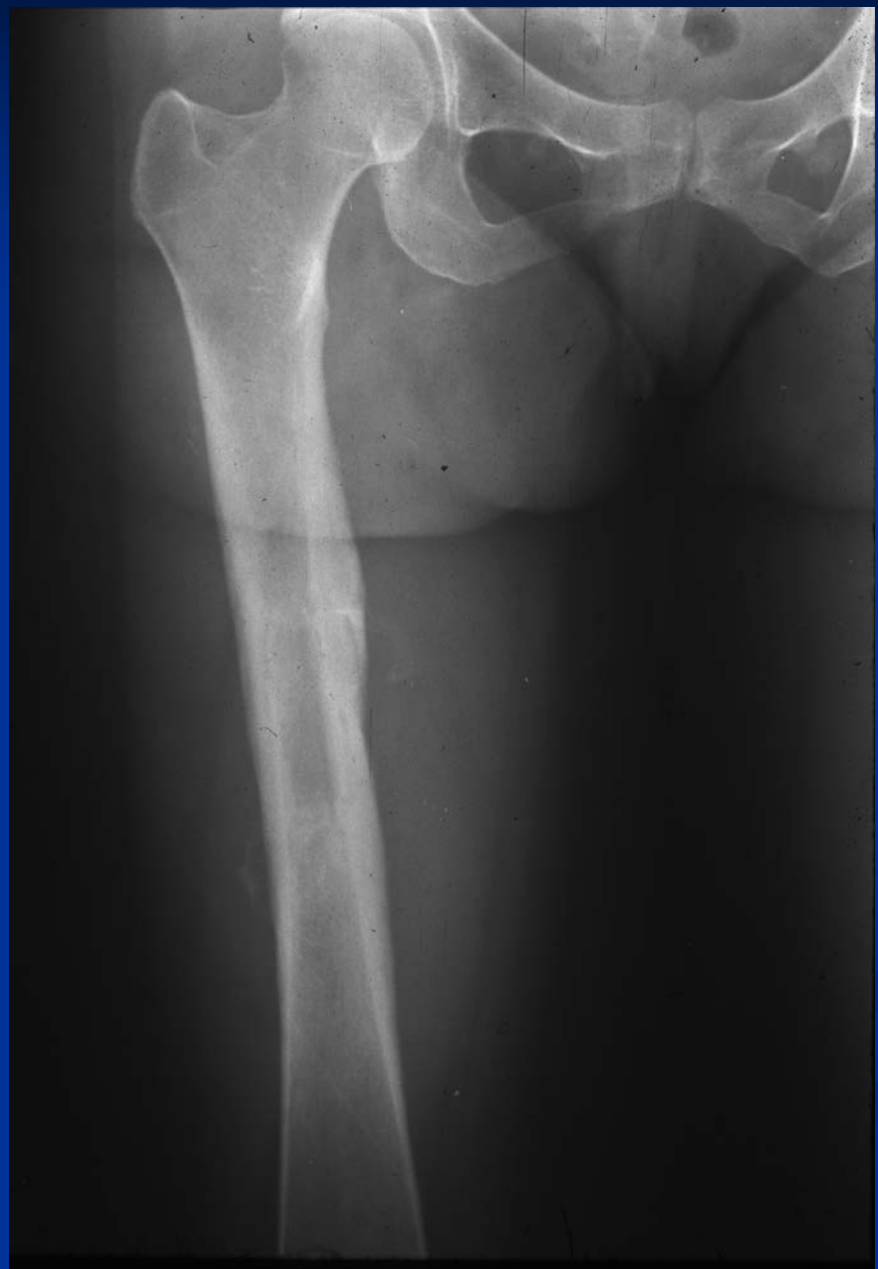

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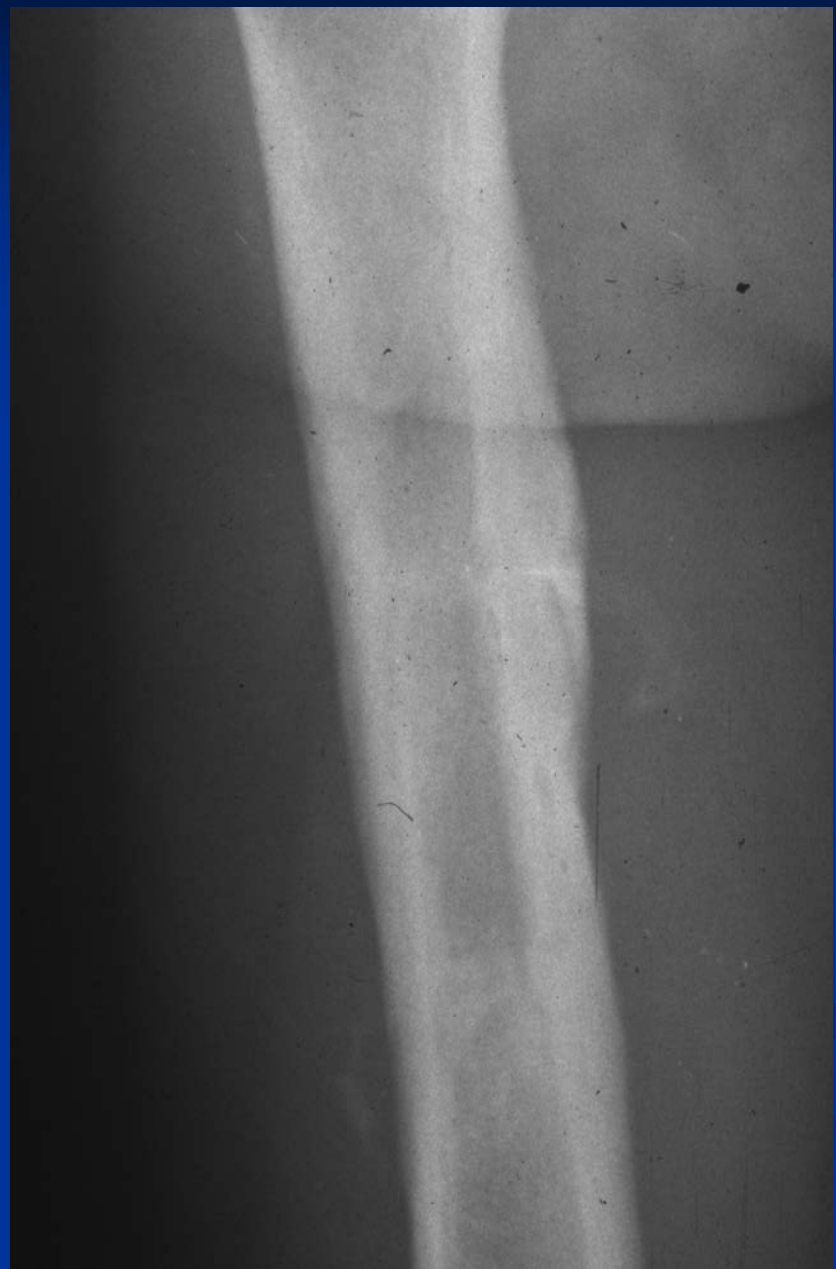

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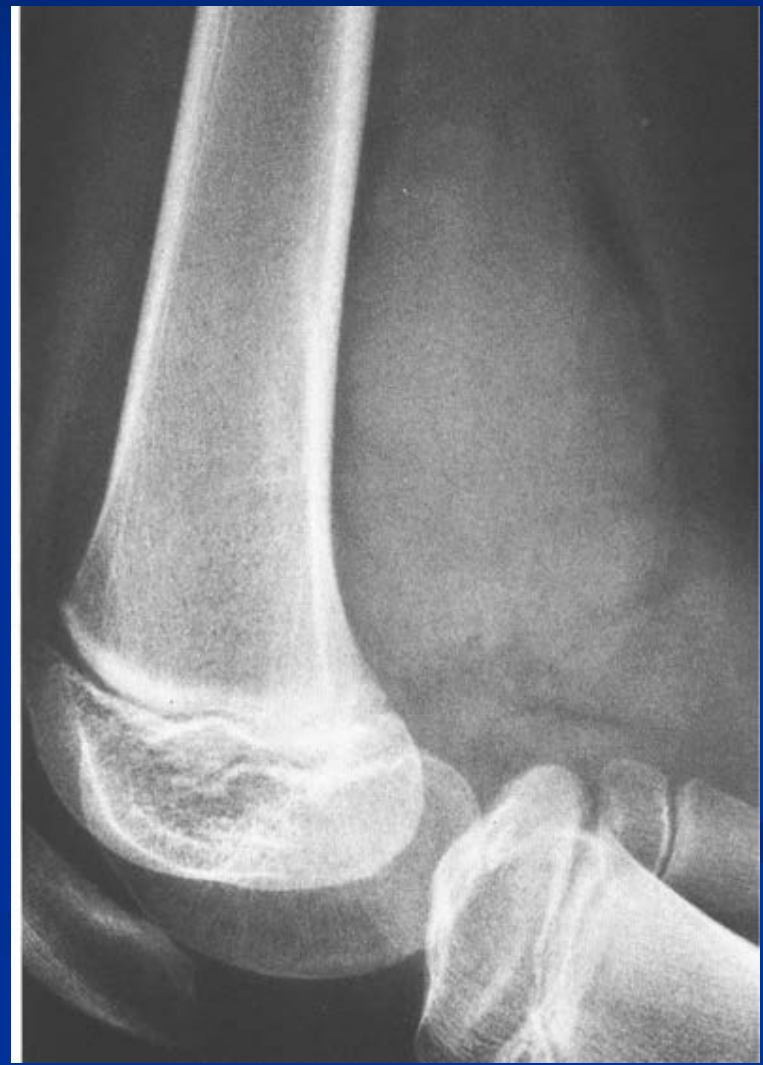

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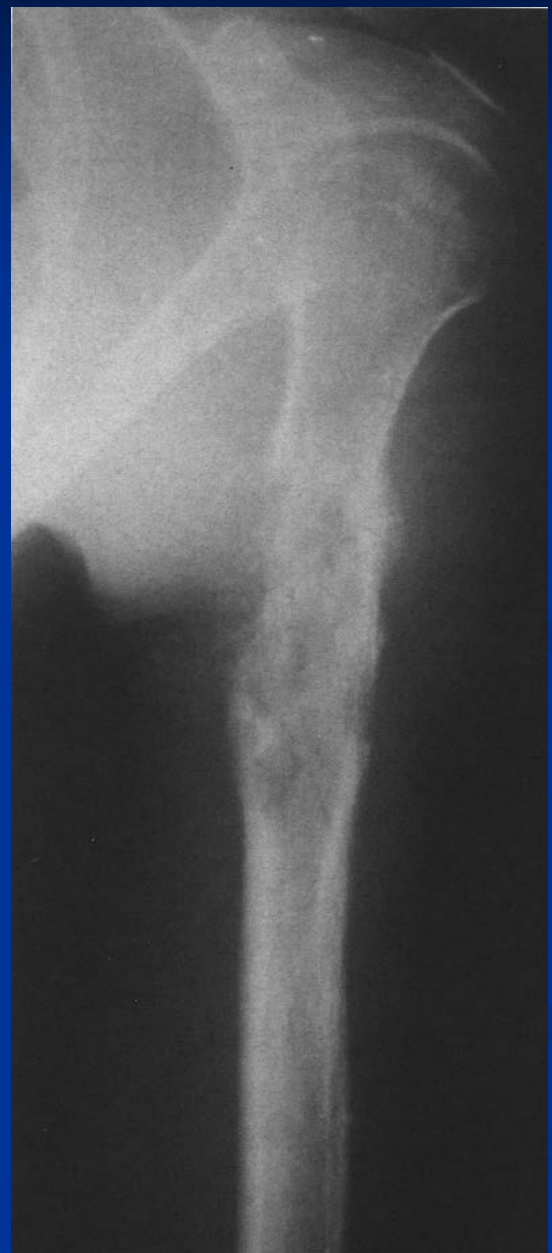

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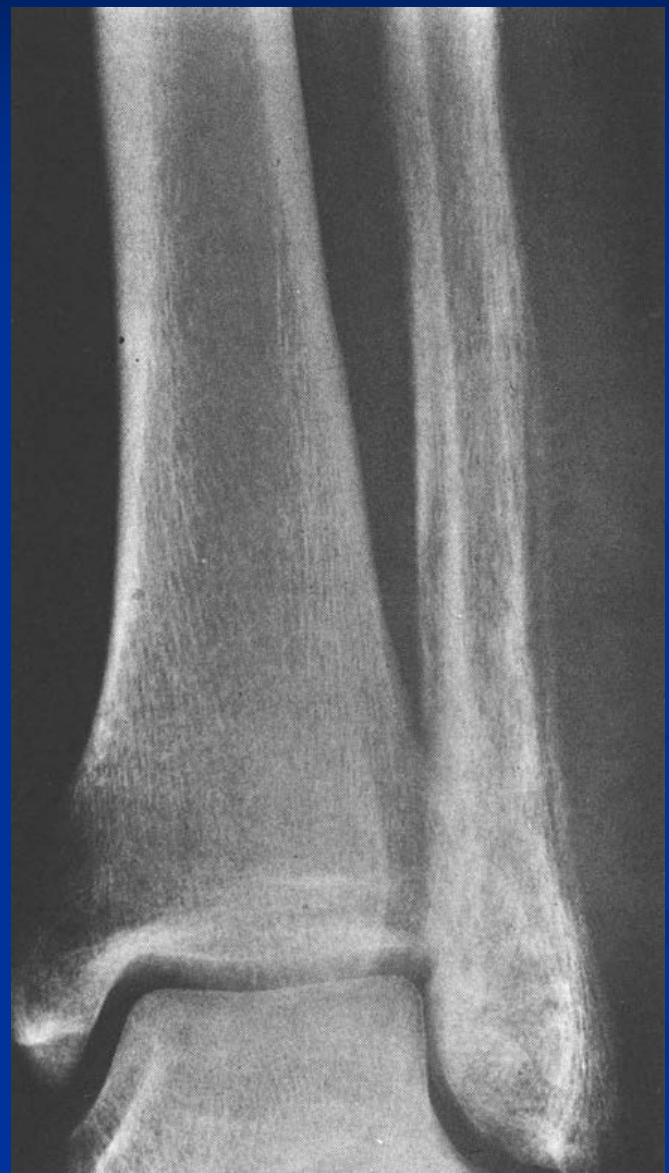

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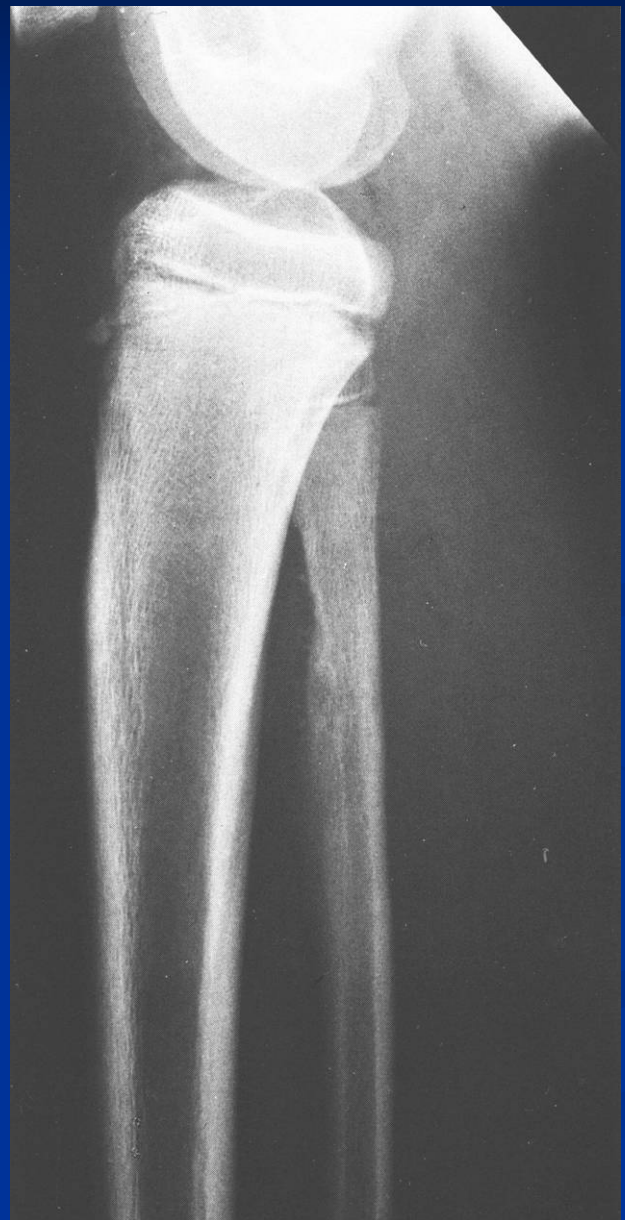

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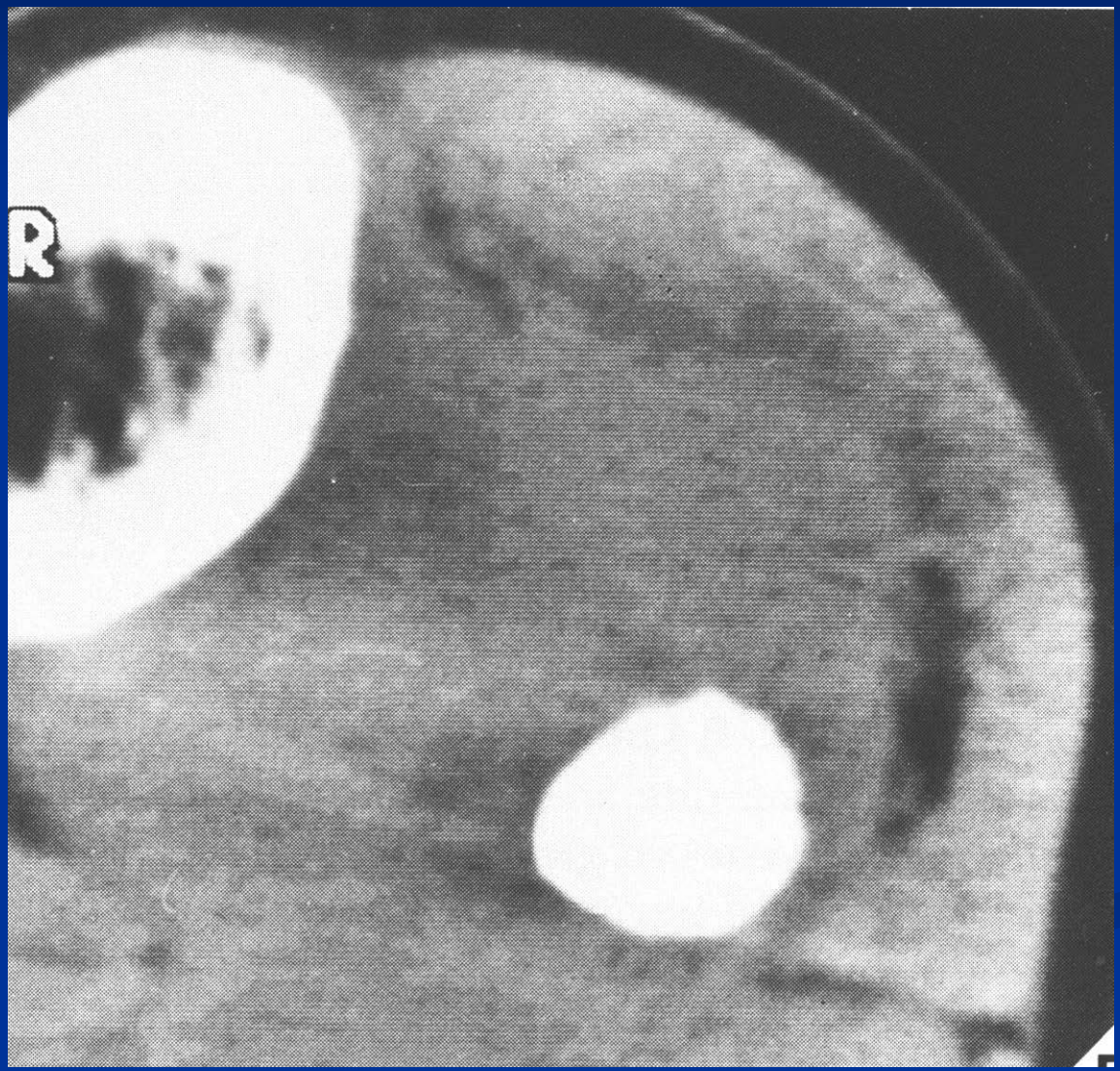

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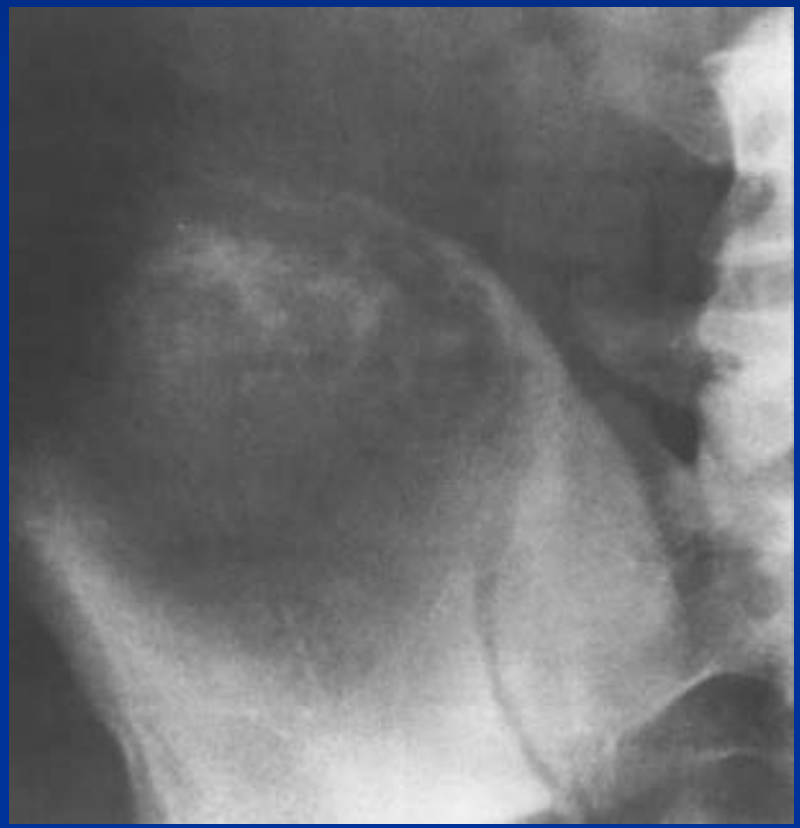

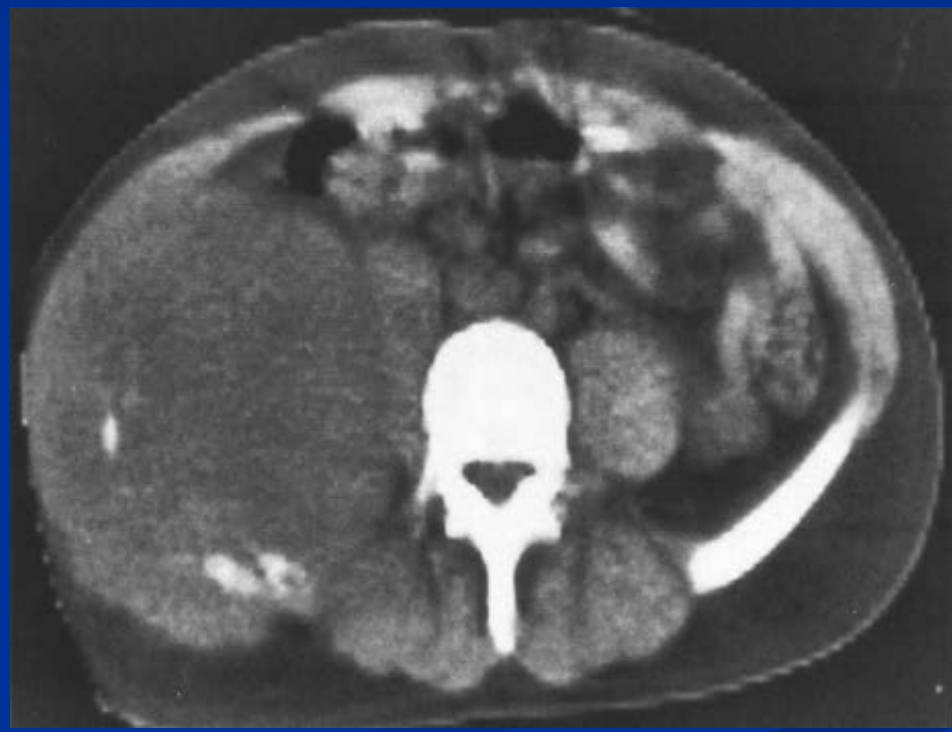

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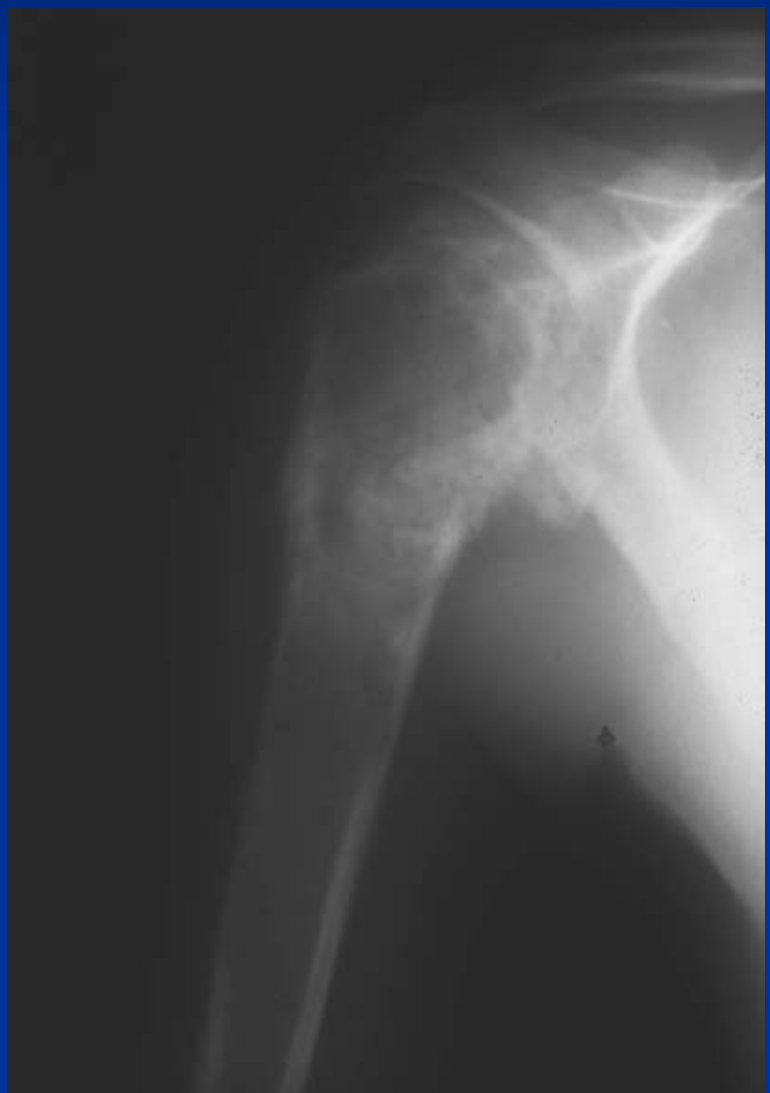

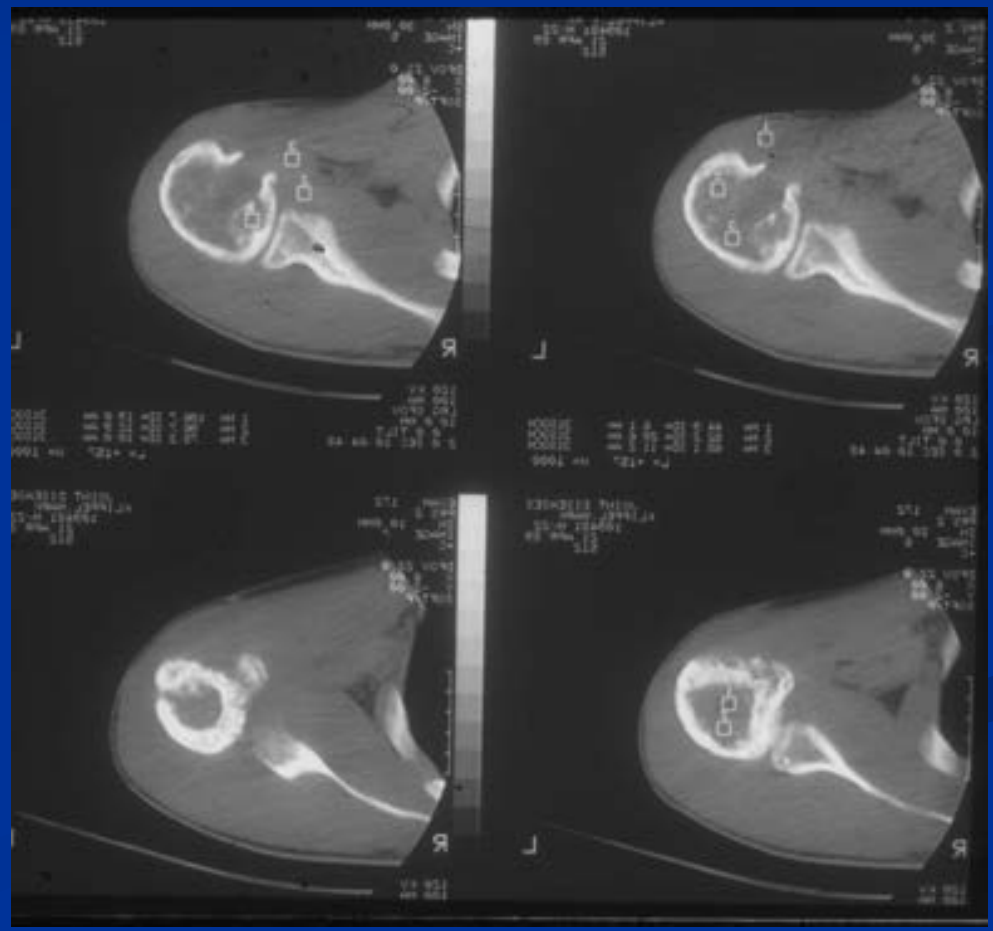

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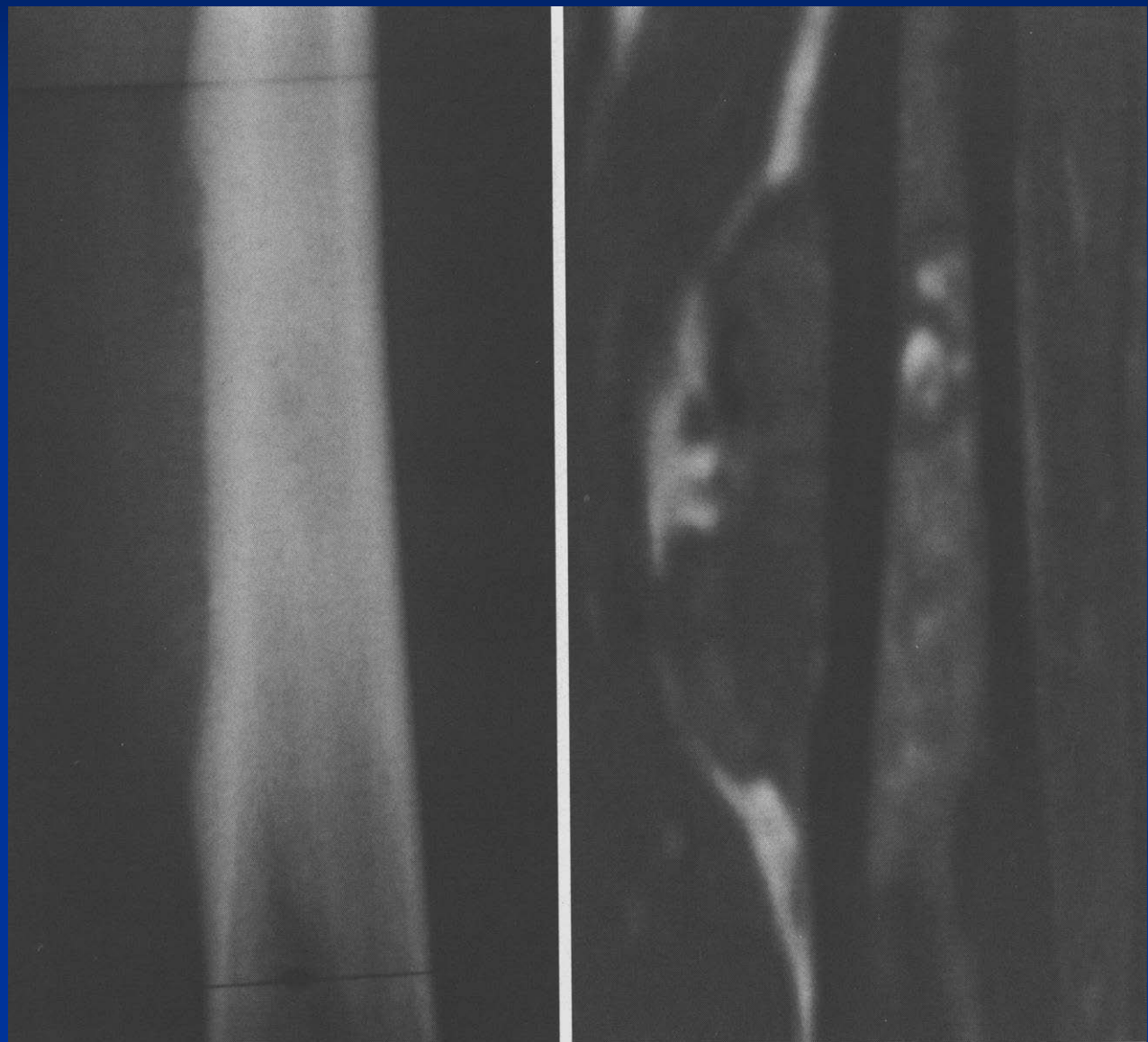

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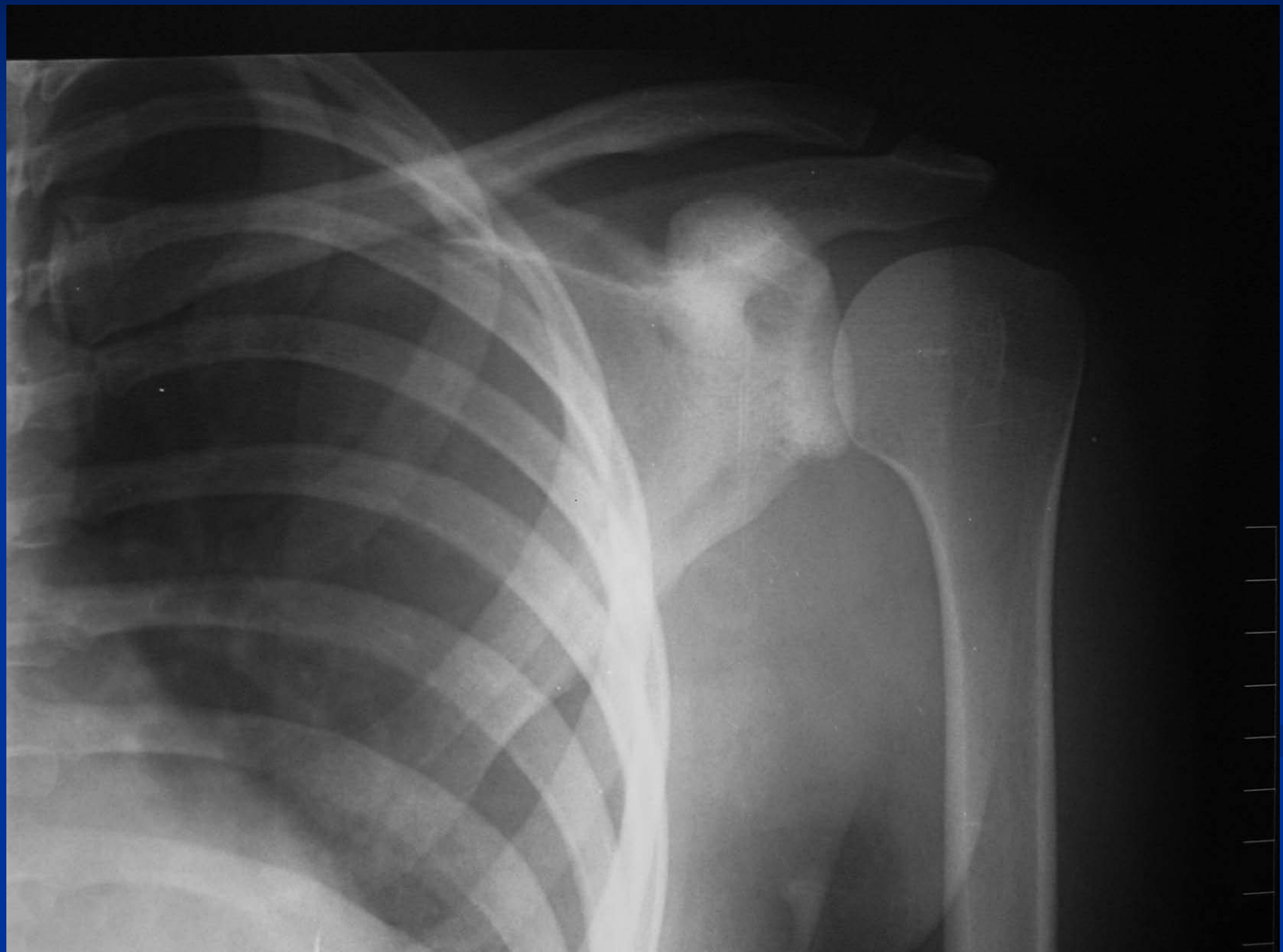

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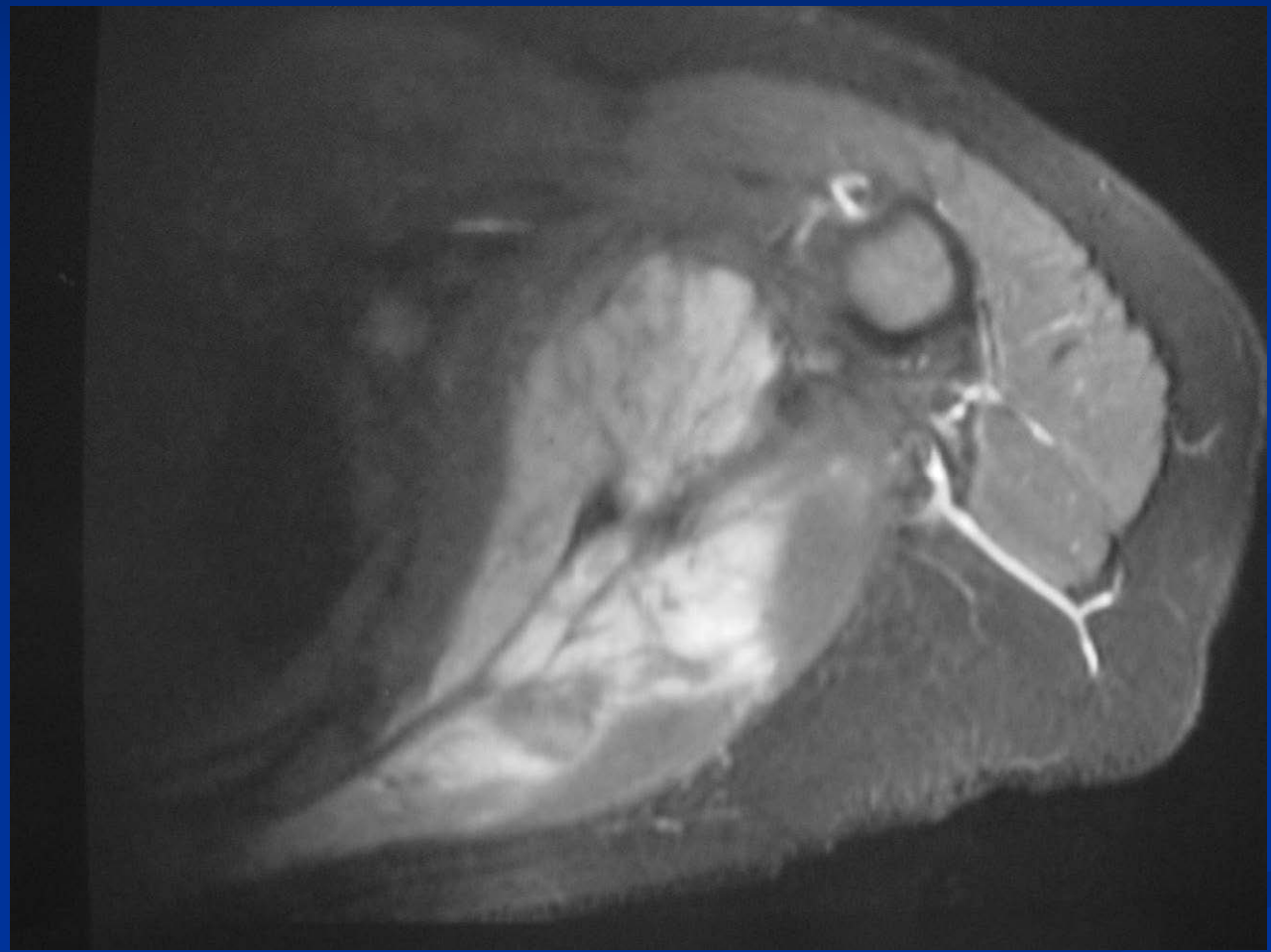

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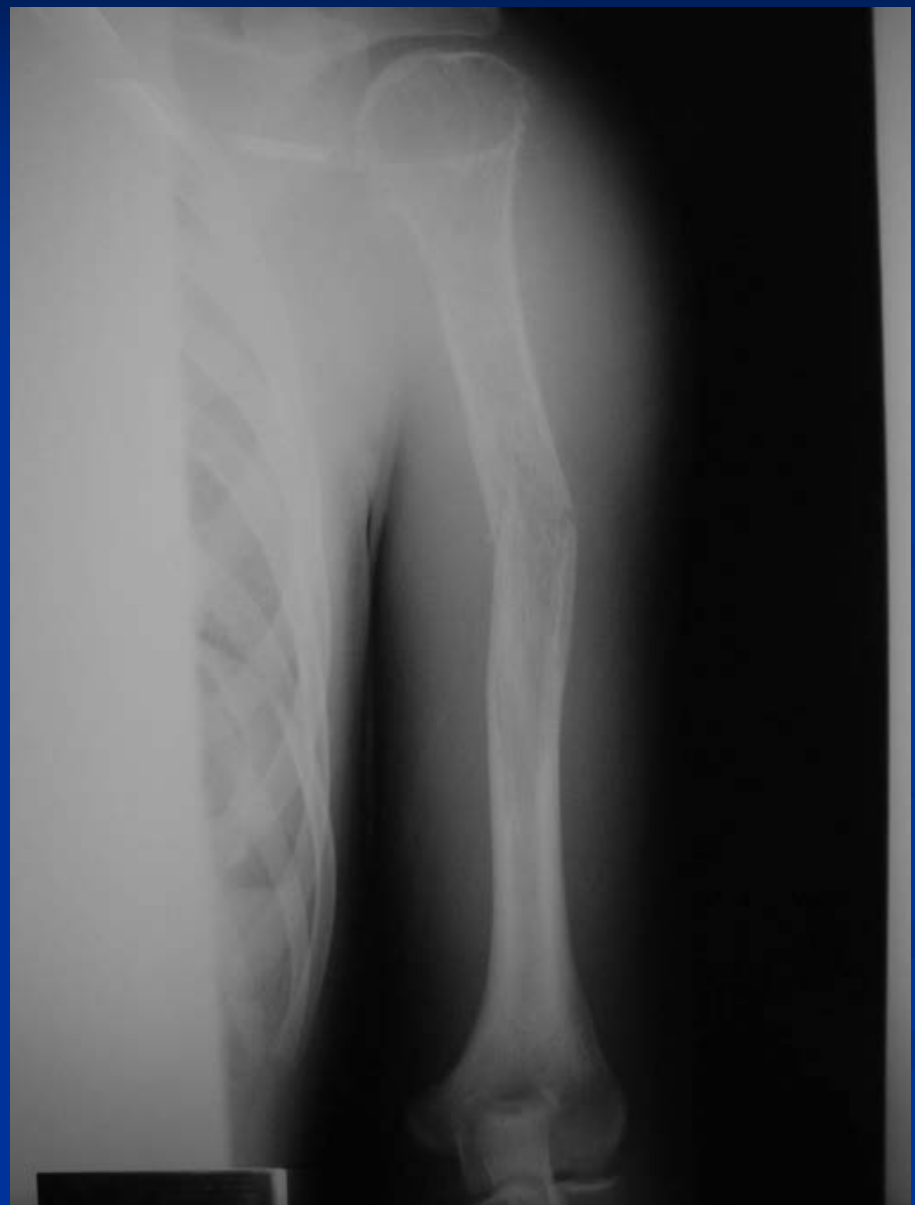

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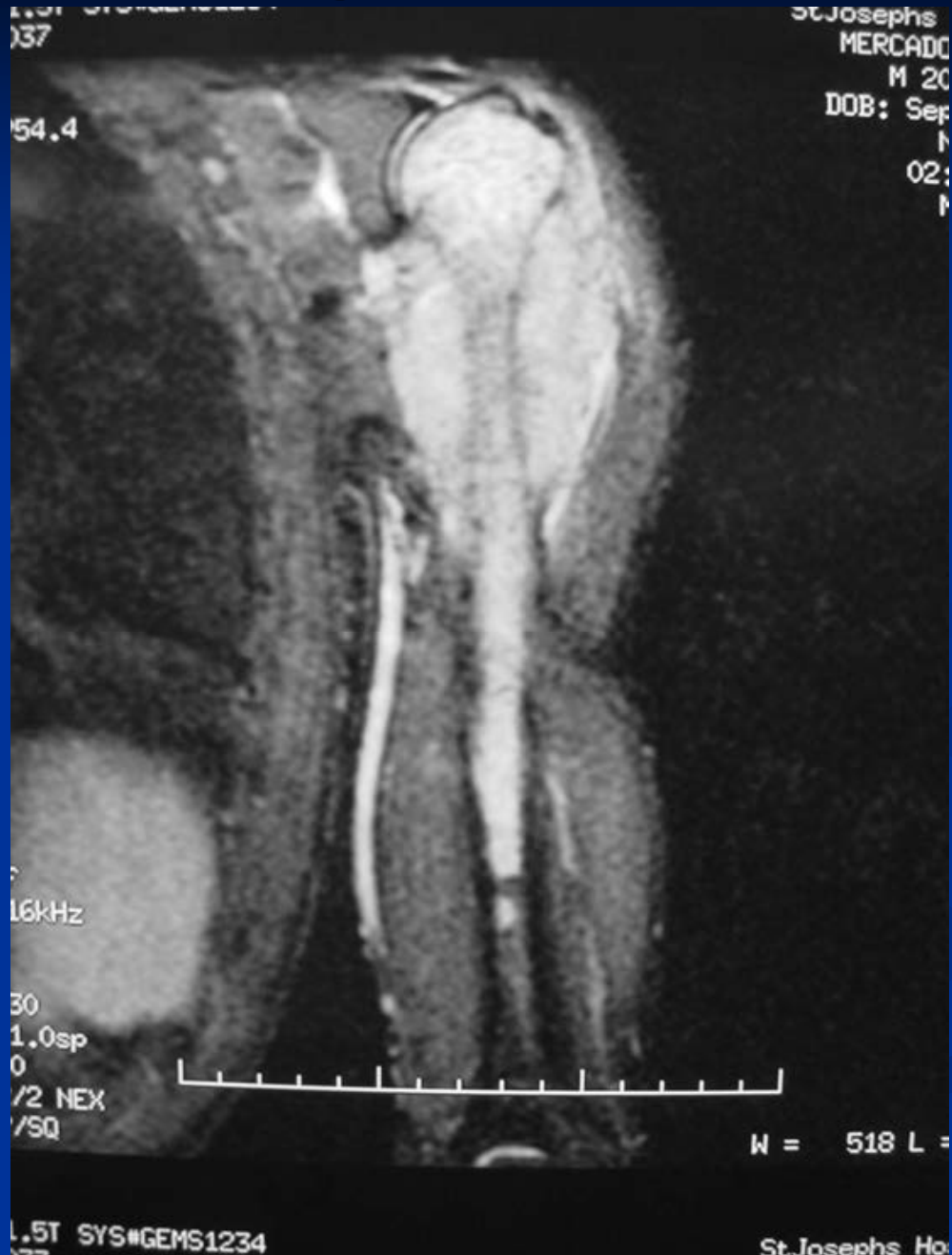

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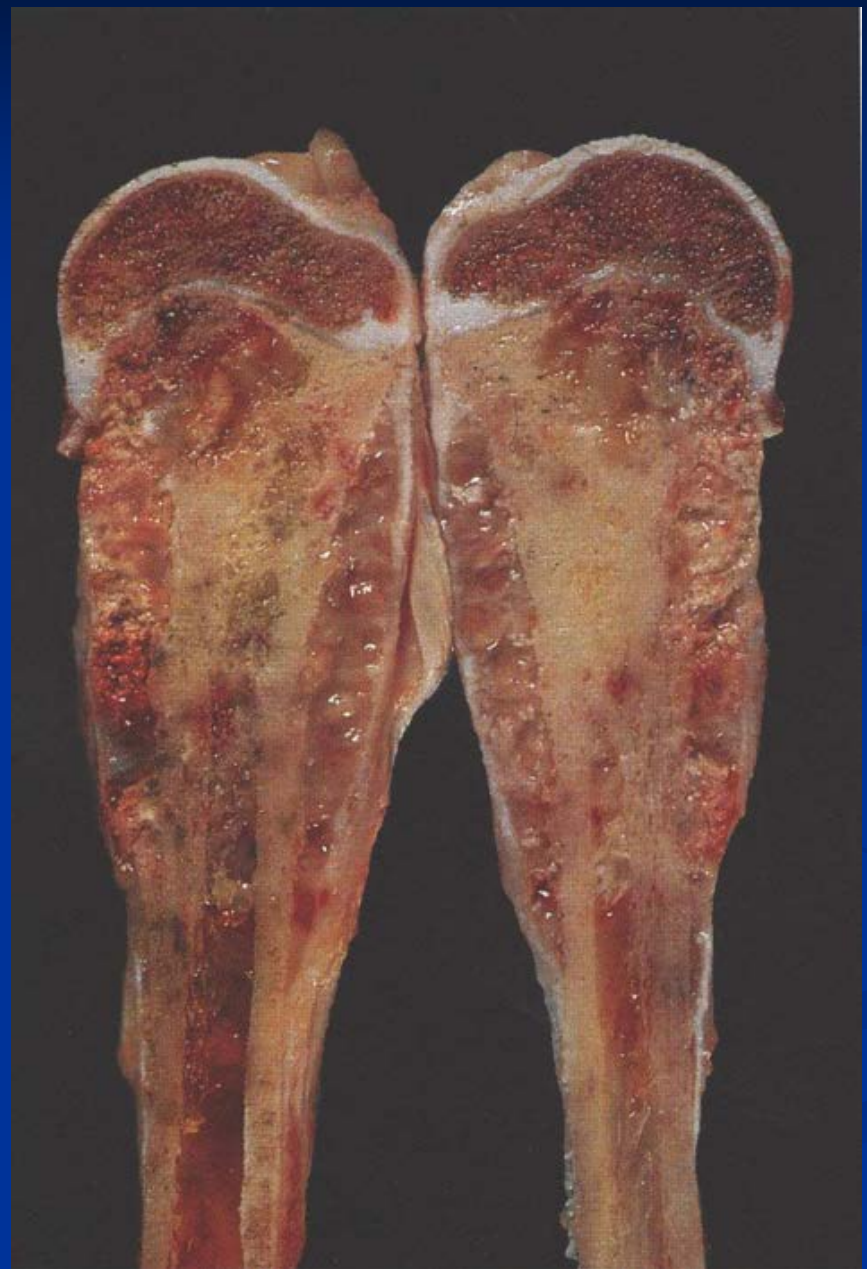

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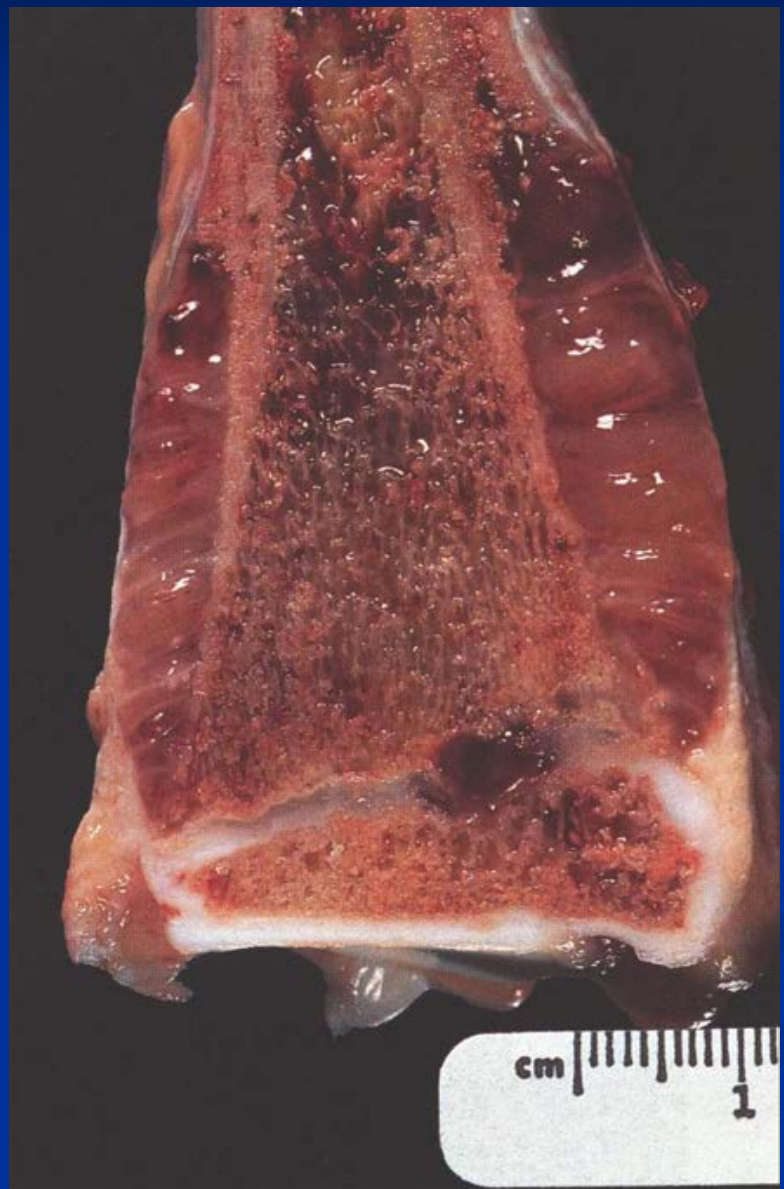
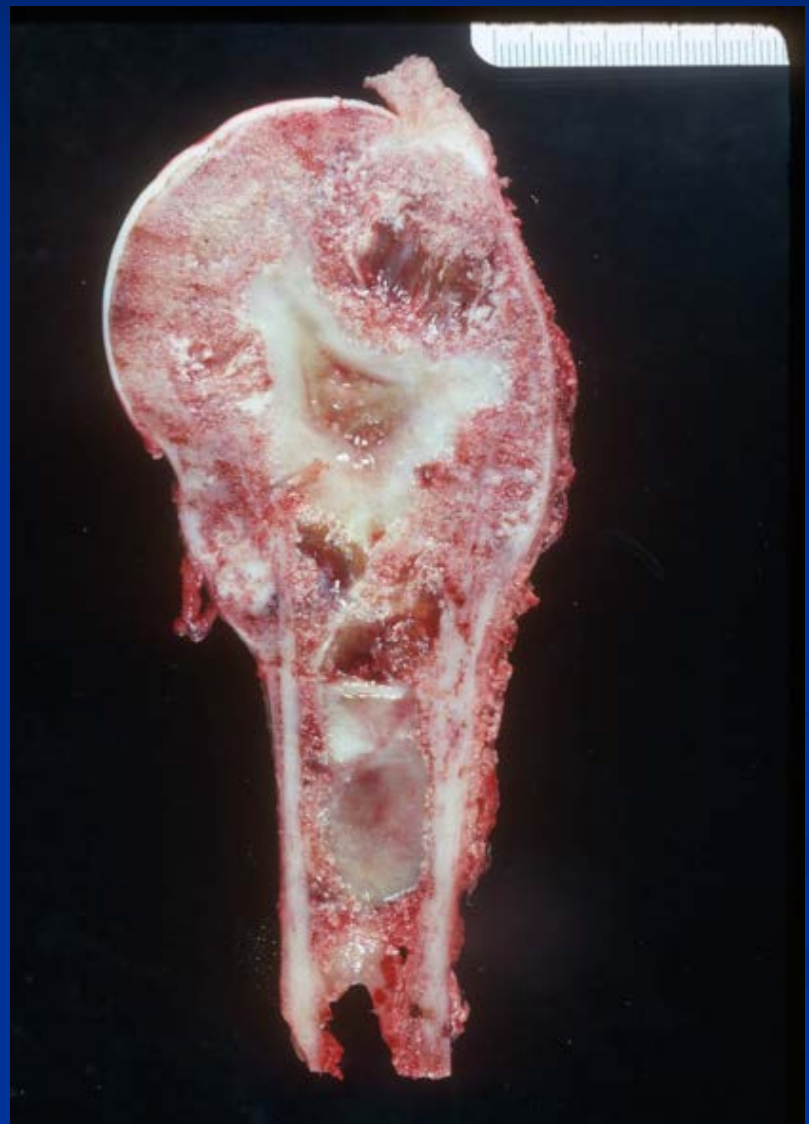


Ewing Sarcoma



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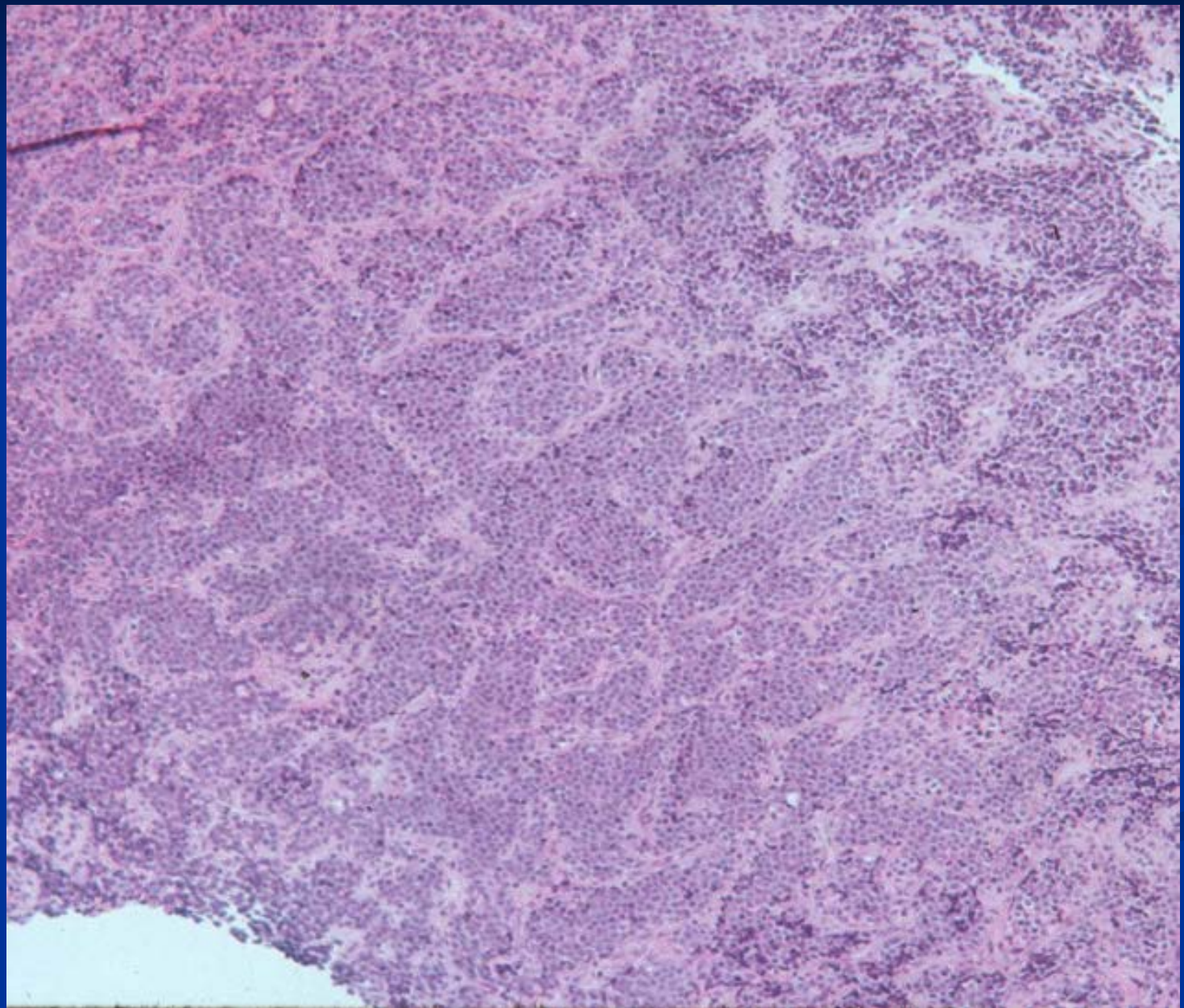

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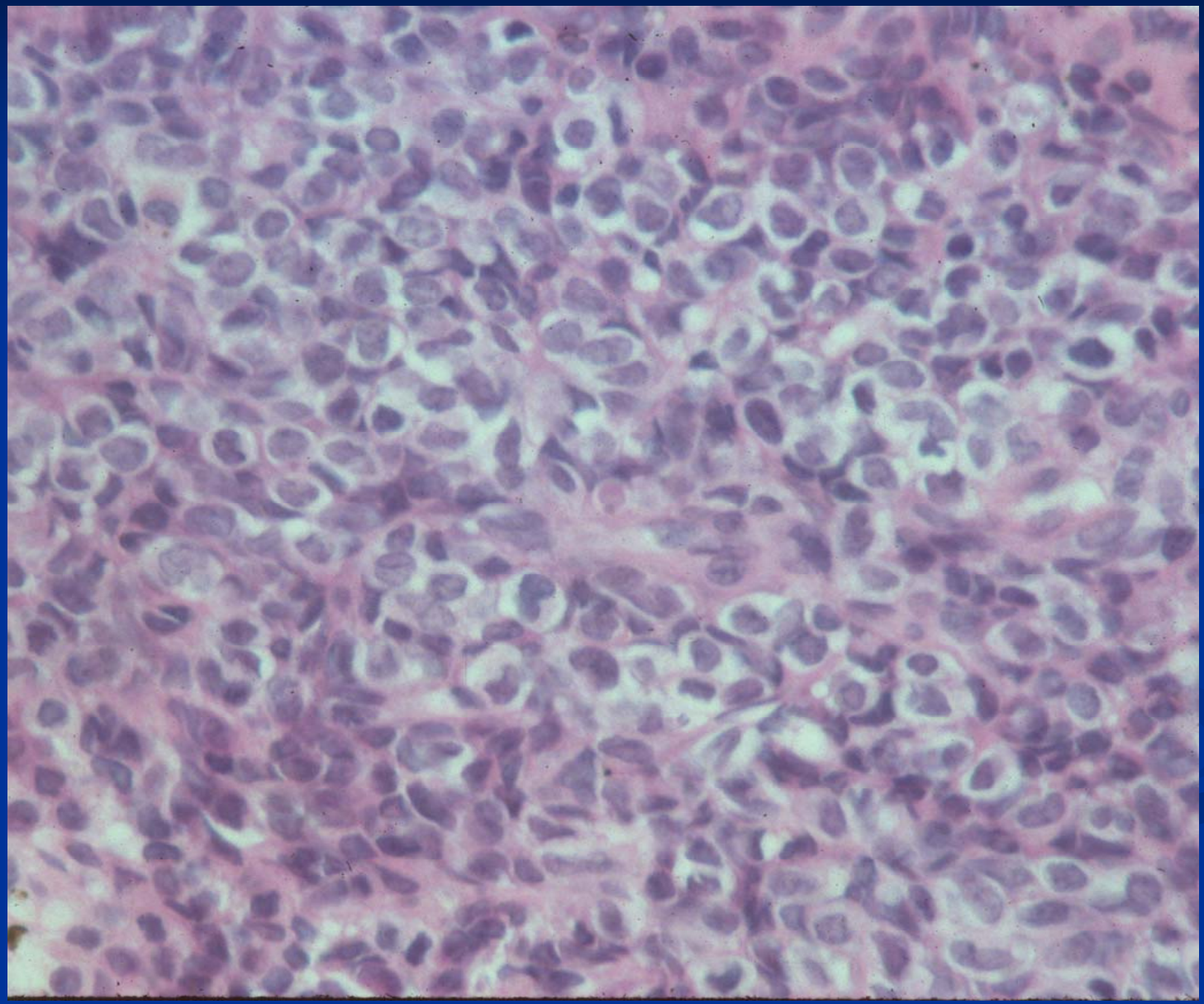

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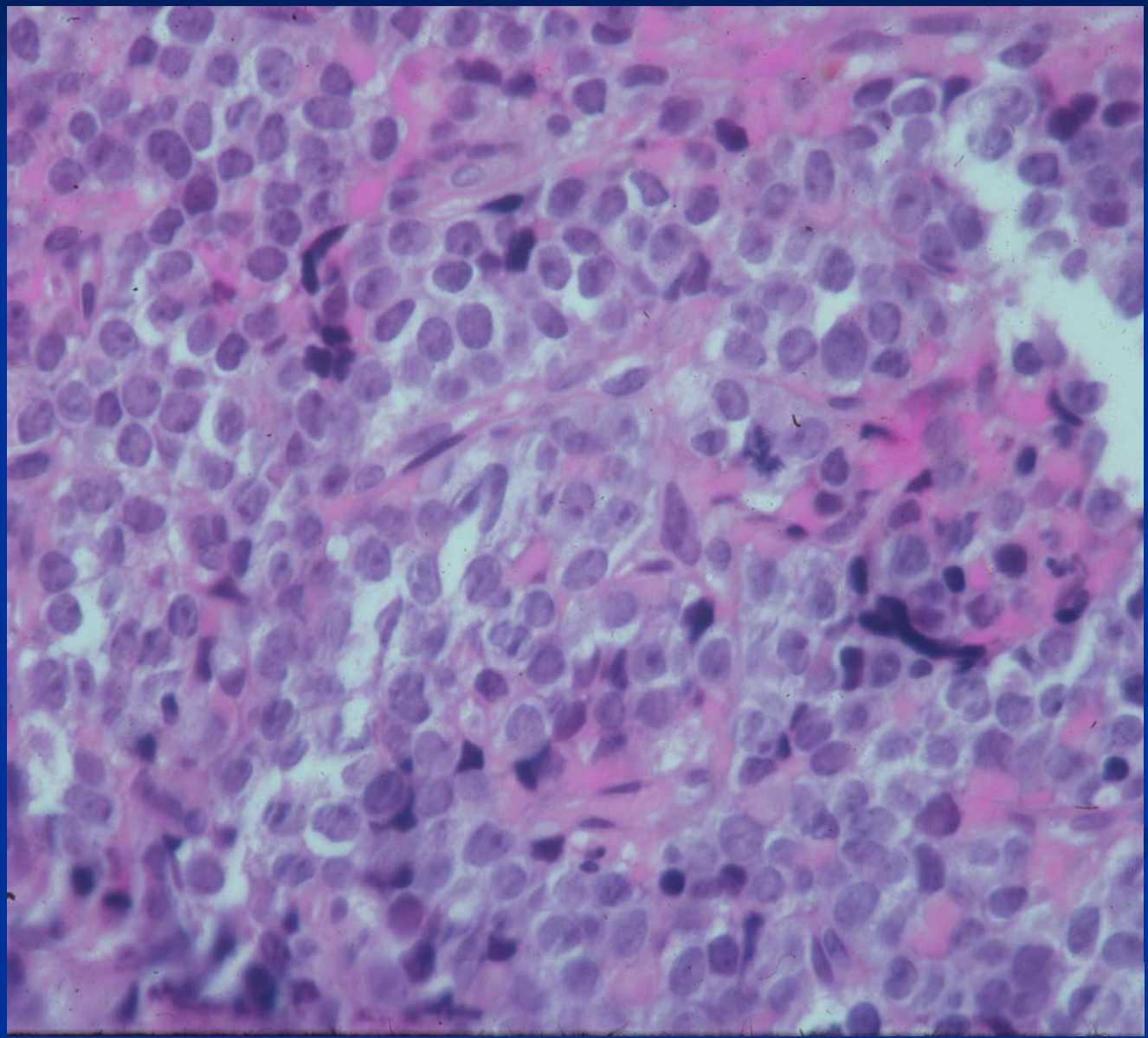
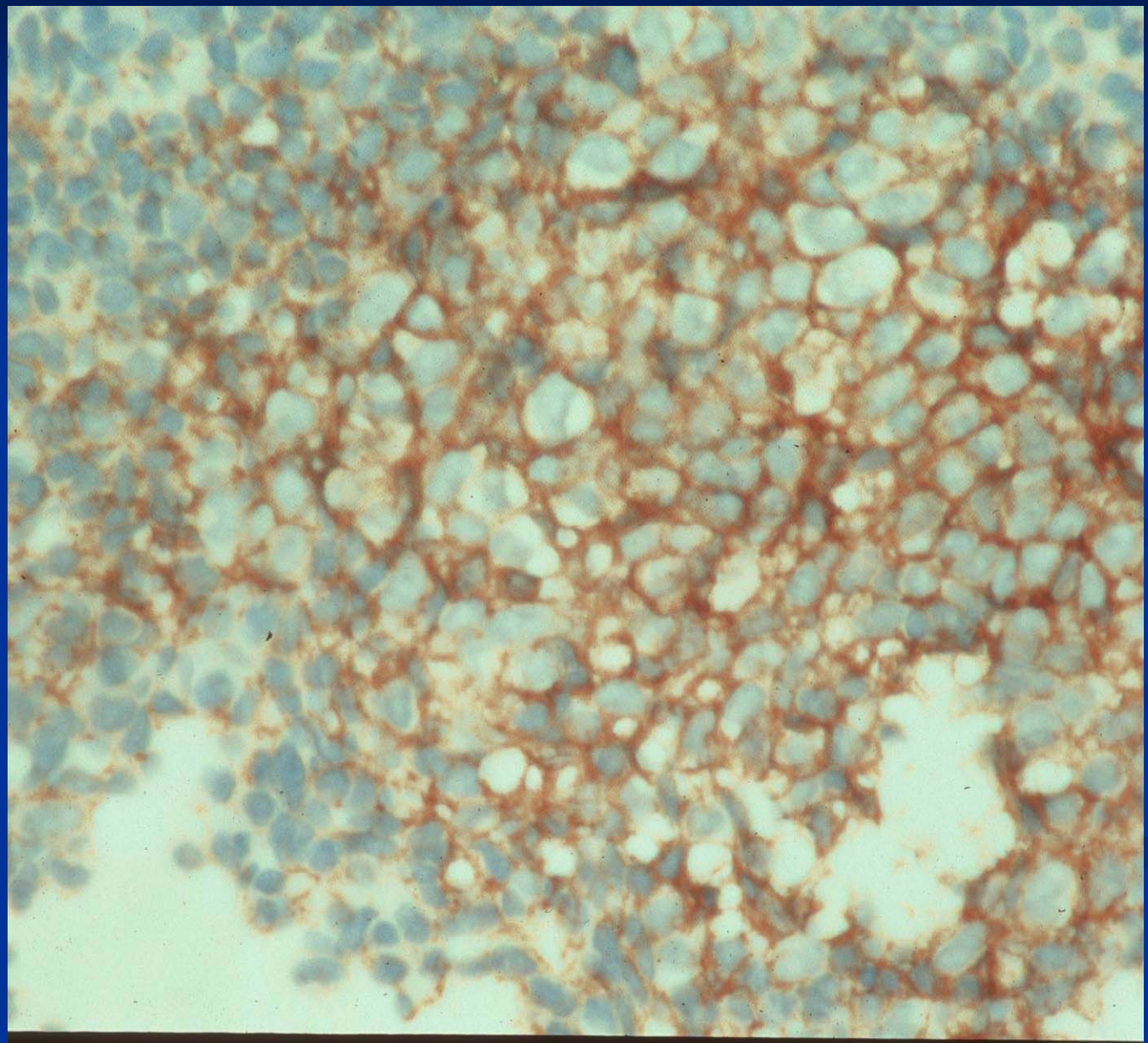


Ewing HBA71 Stain



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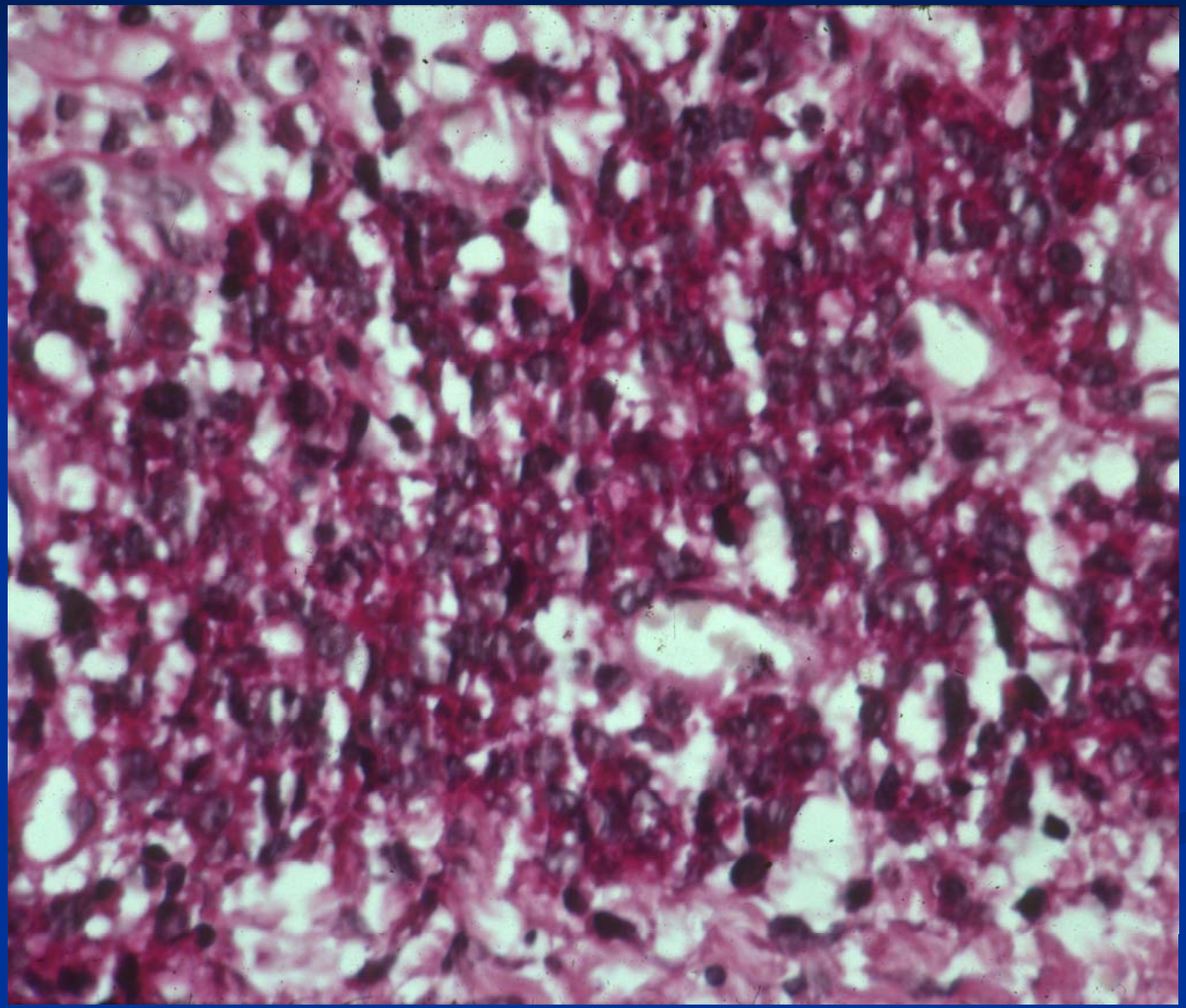
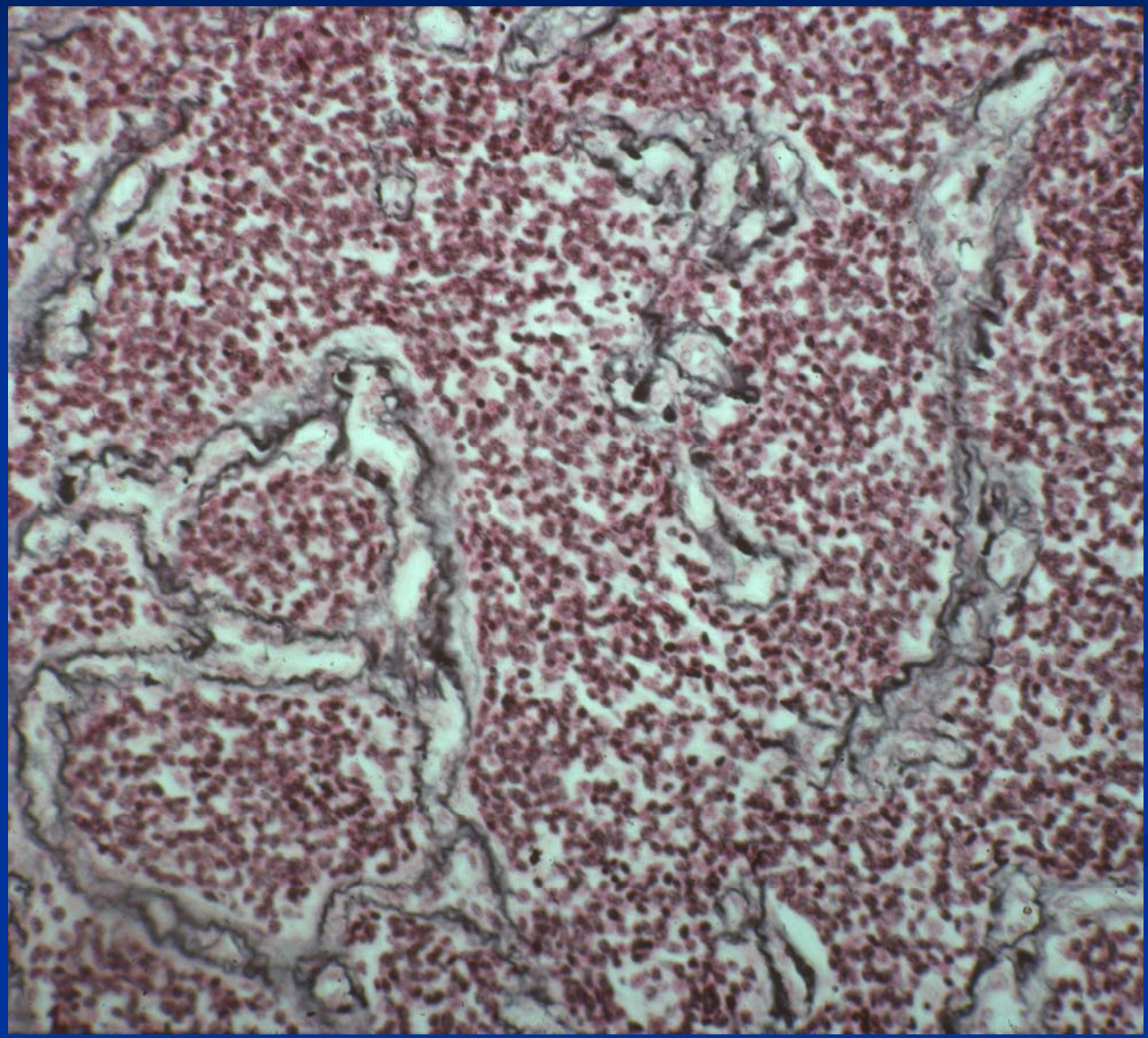
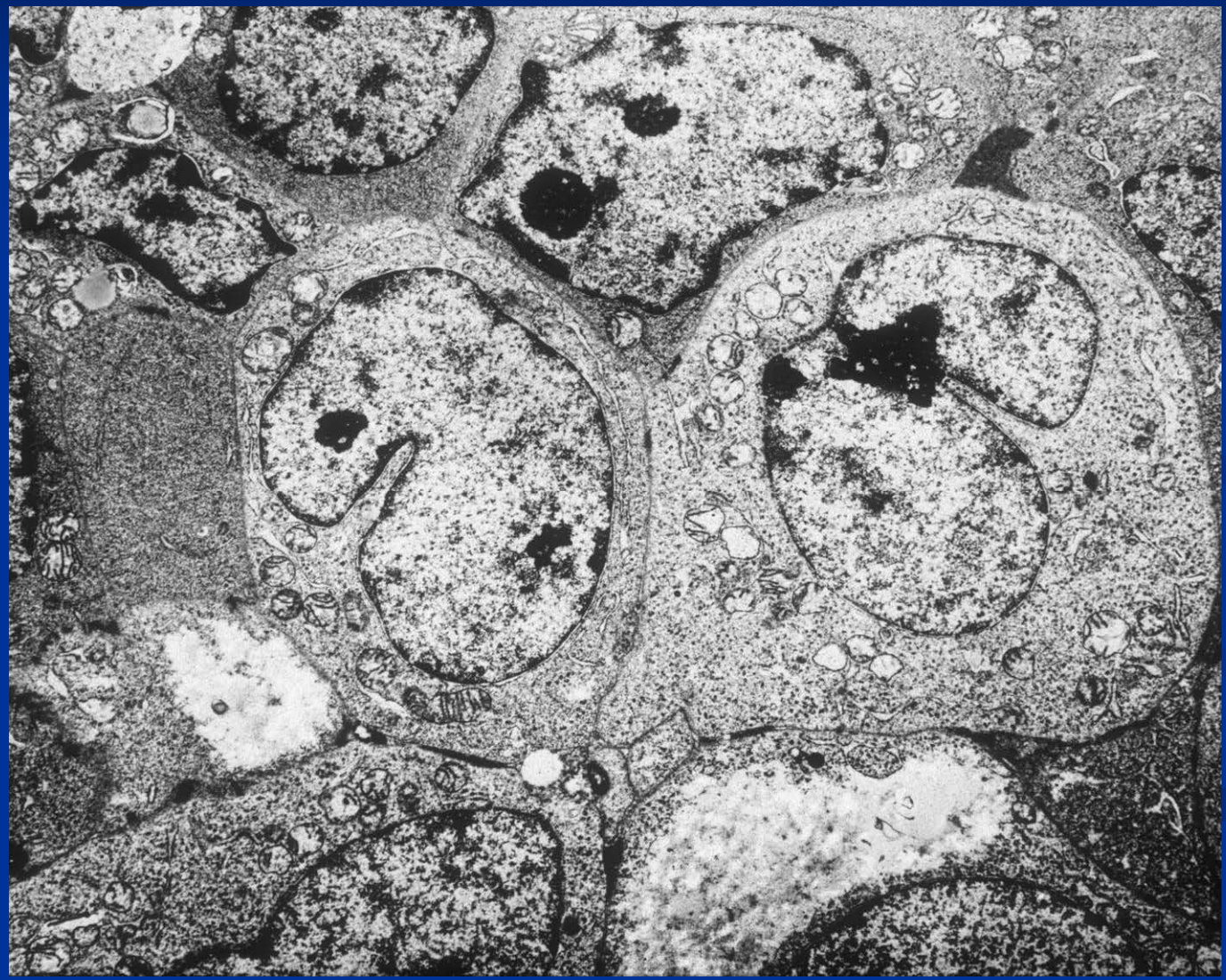


Ewing Reticulin



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

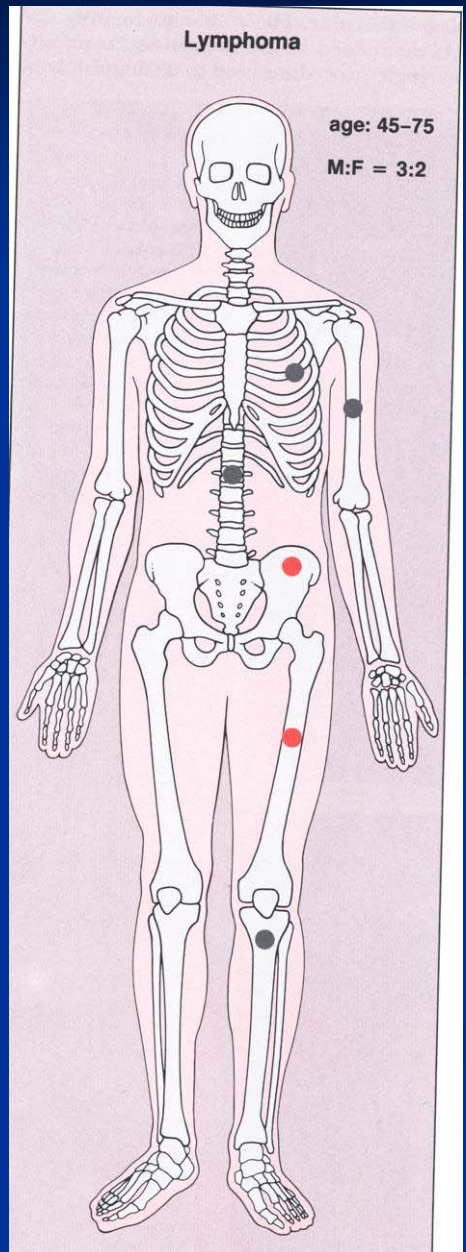
Lymphoma

- 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%)

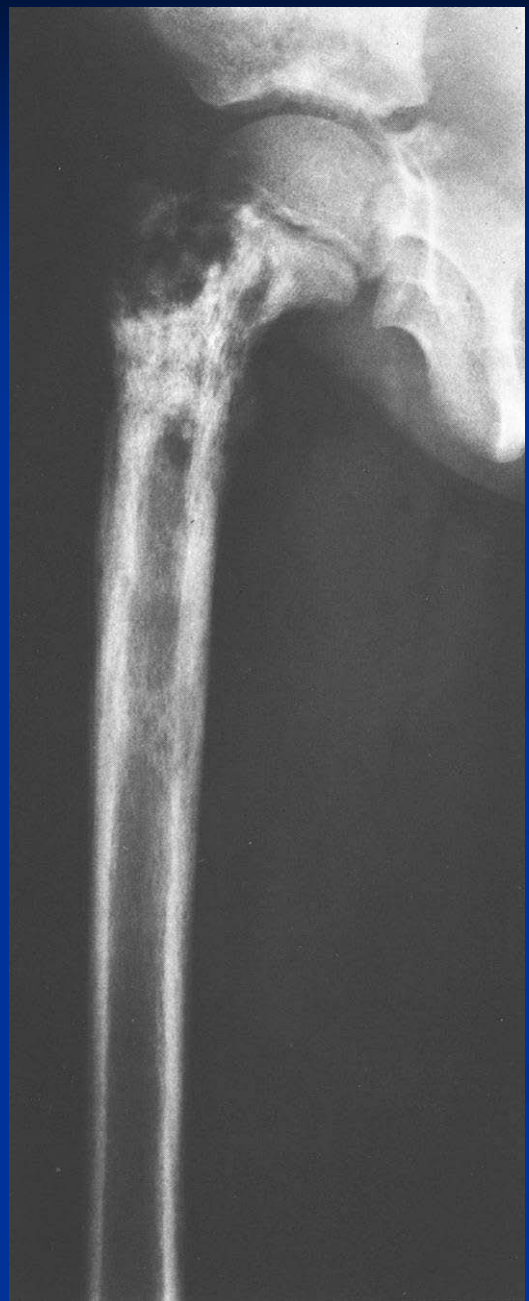
Lymphoma

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

Lymphoma



Lymphoma



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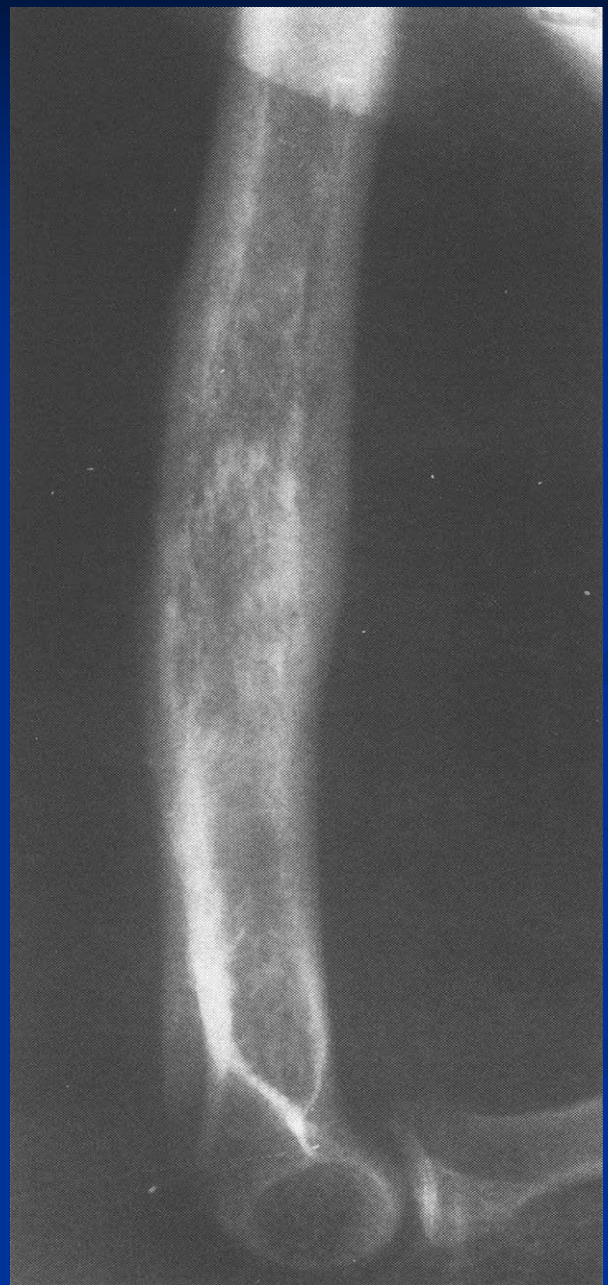

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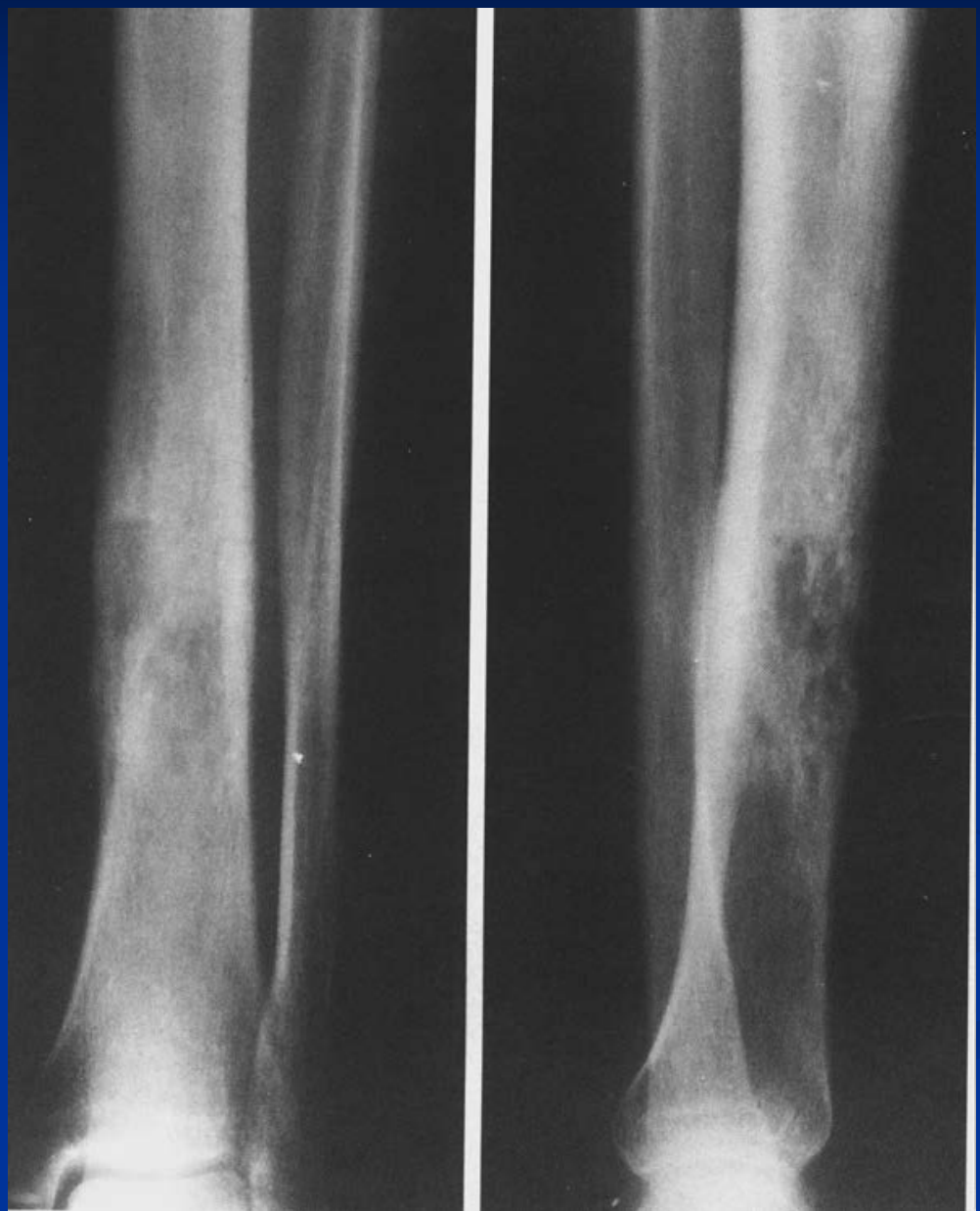
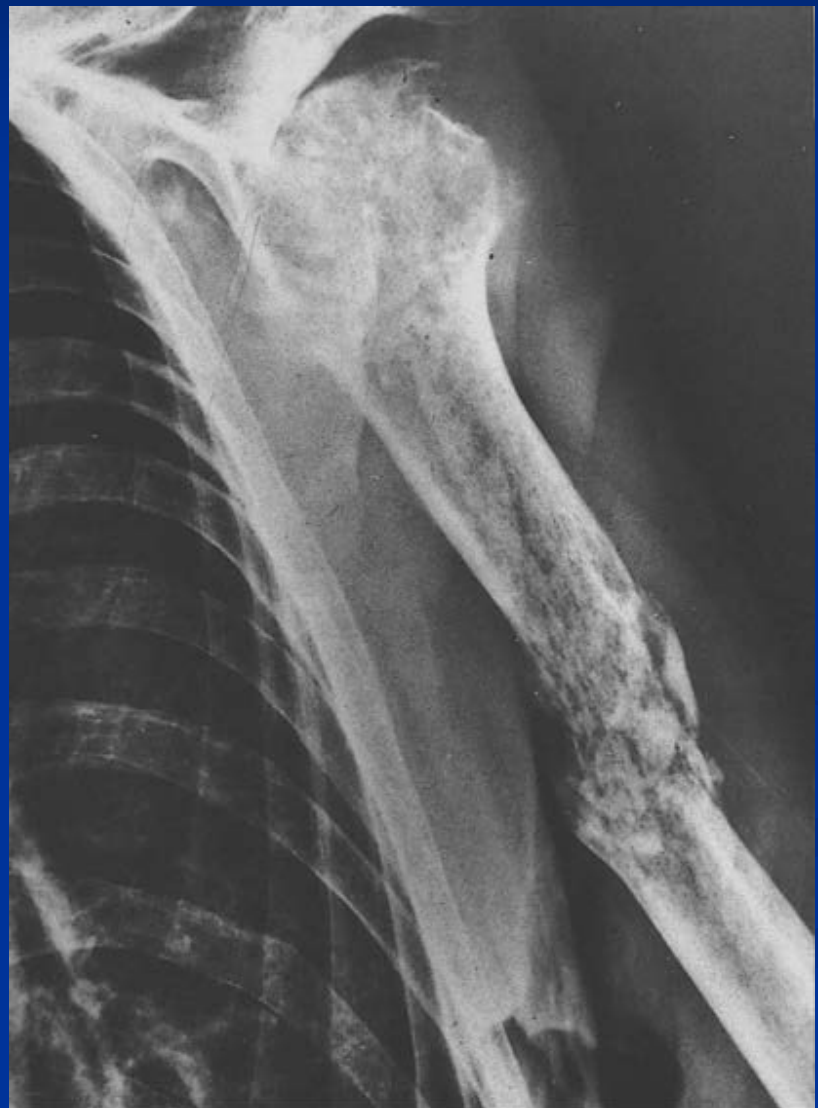


Lymphoma



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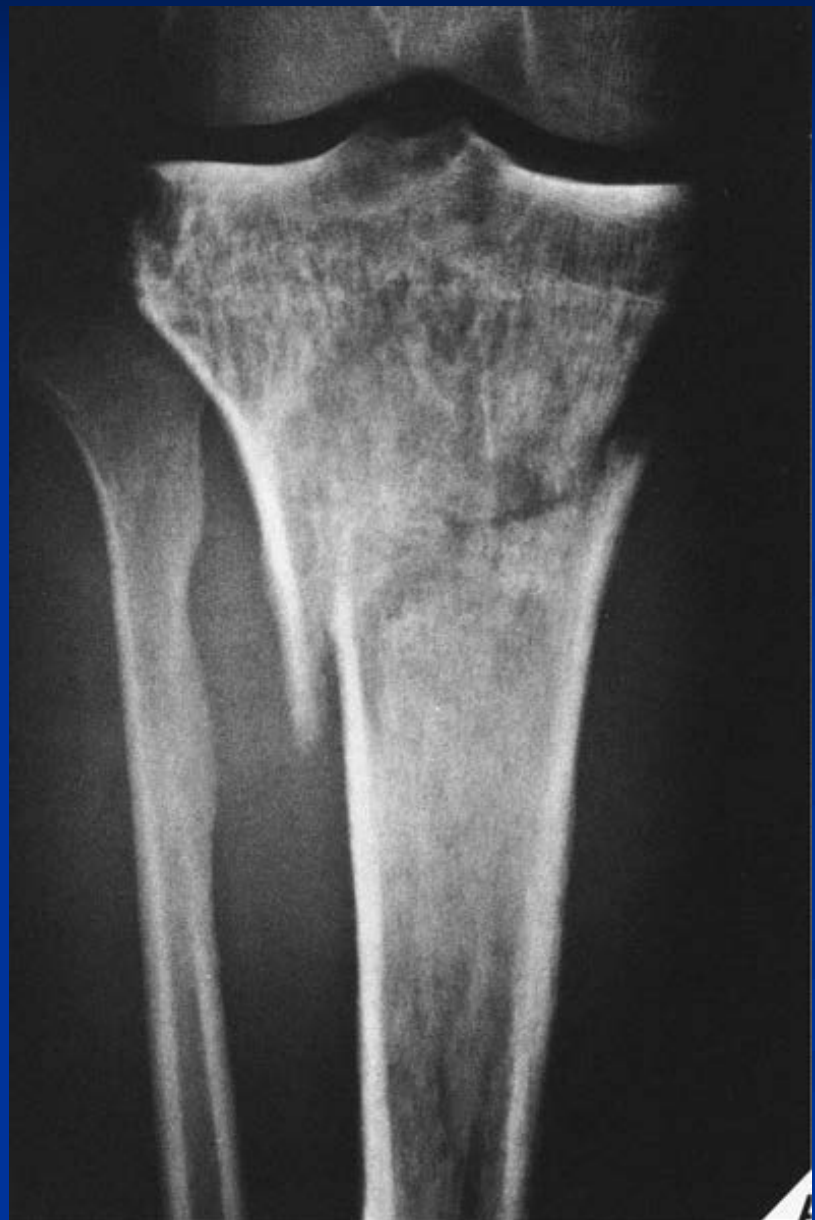

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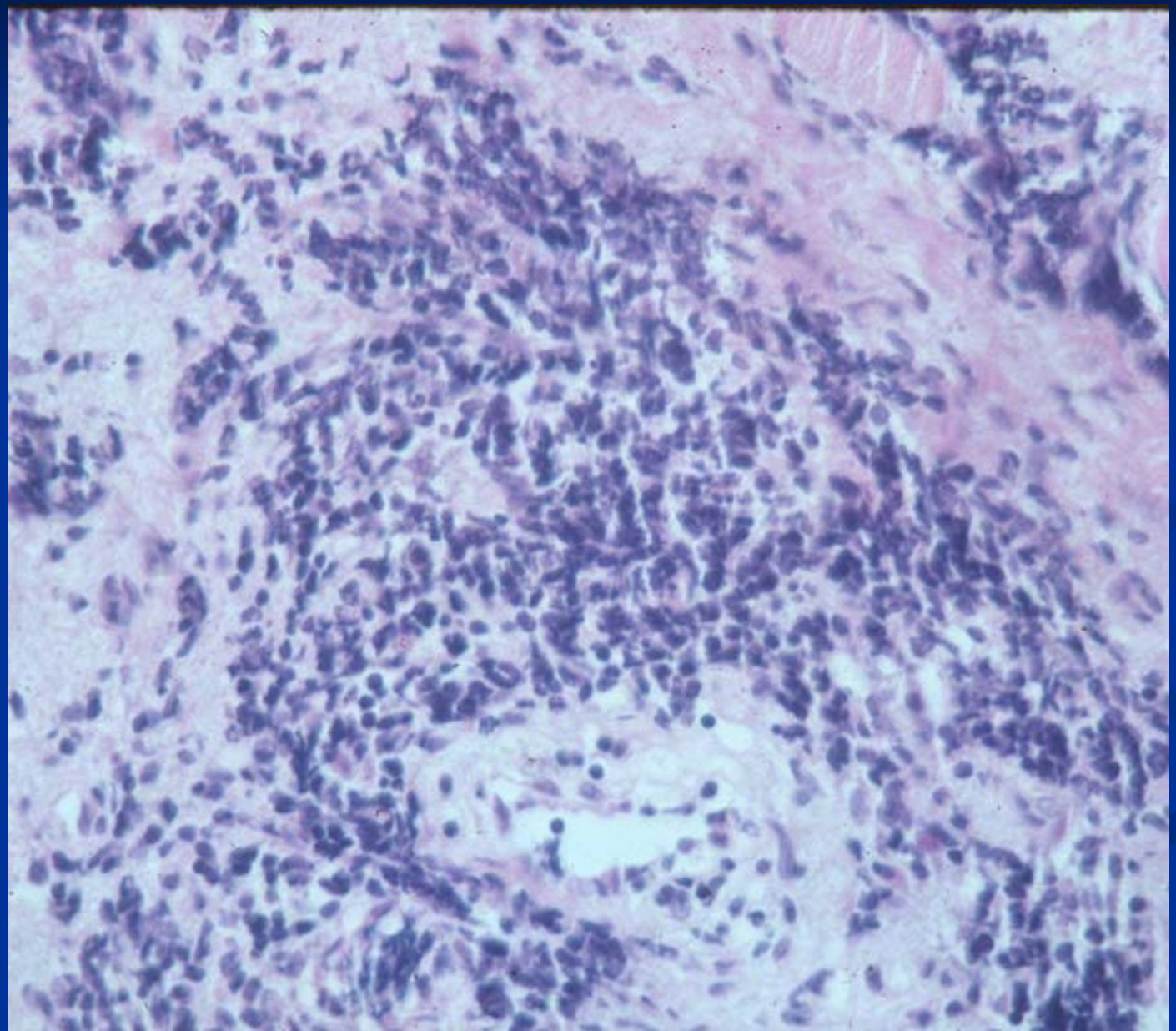

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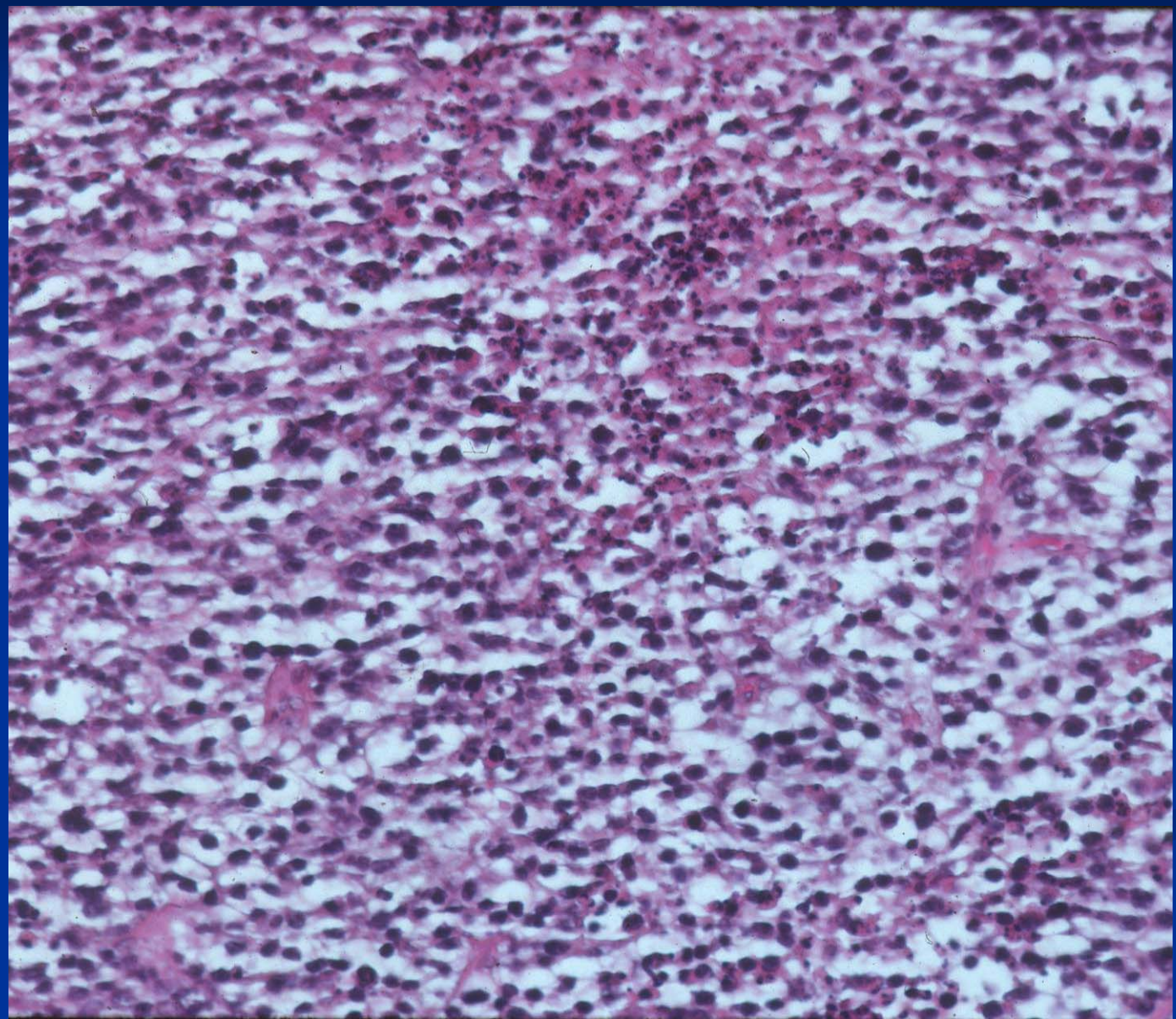

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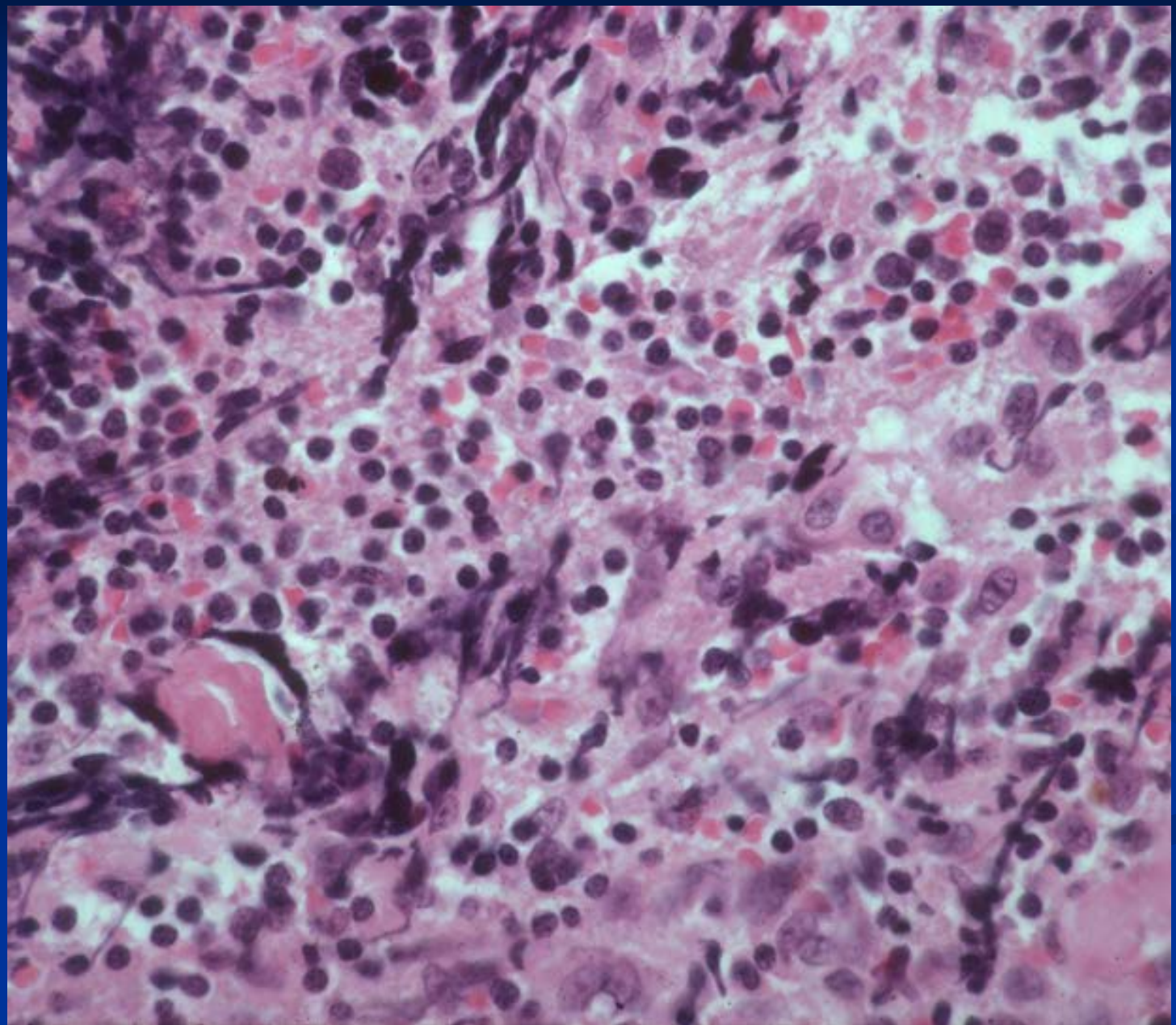

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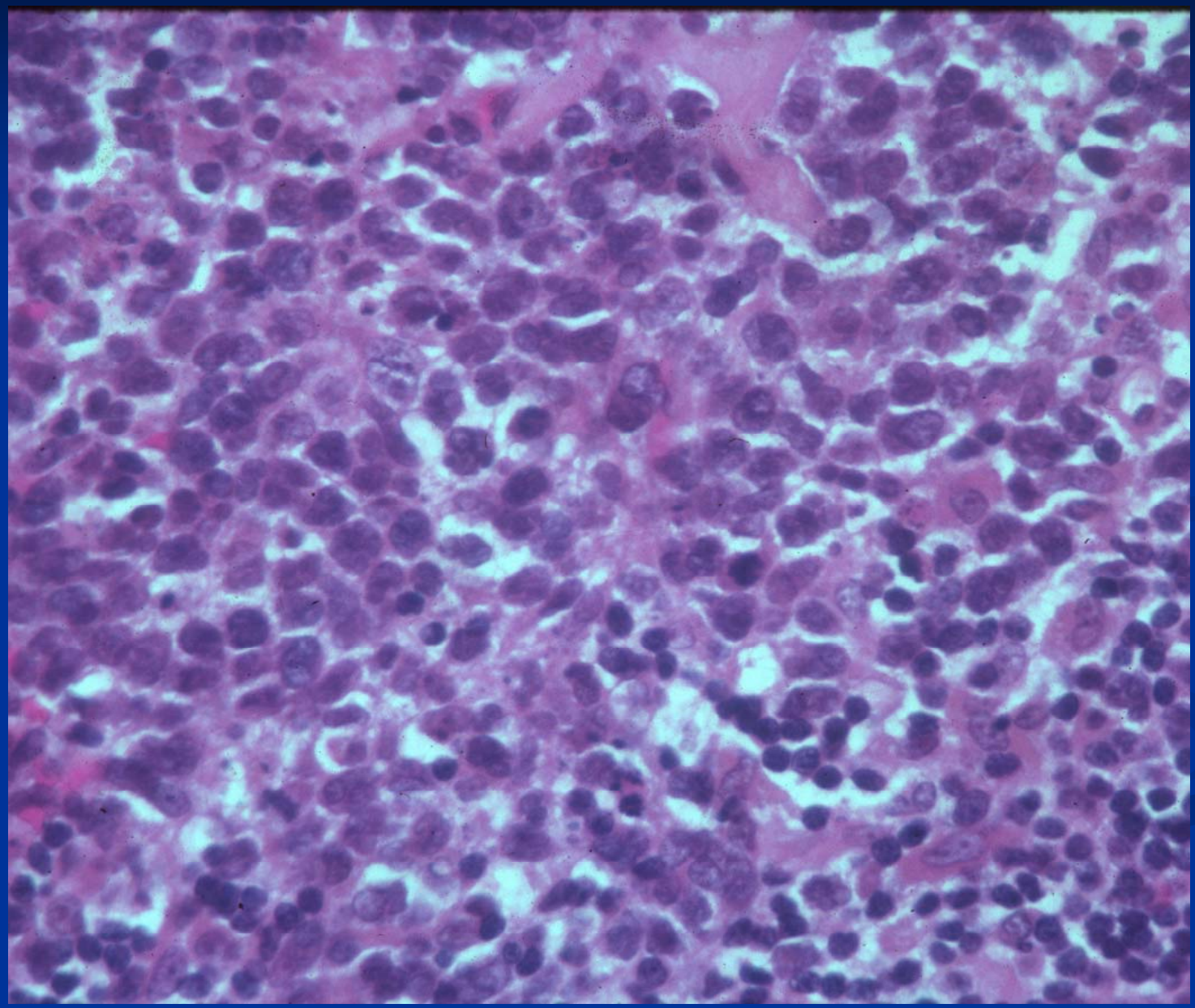

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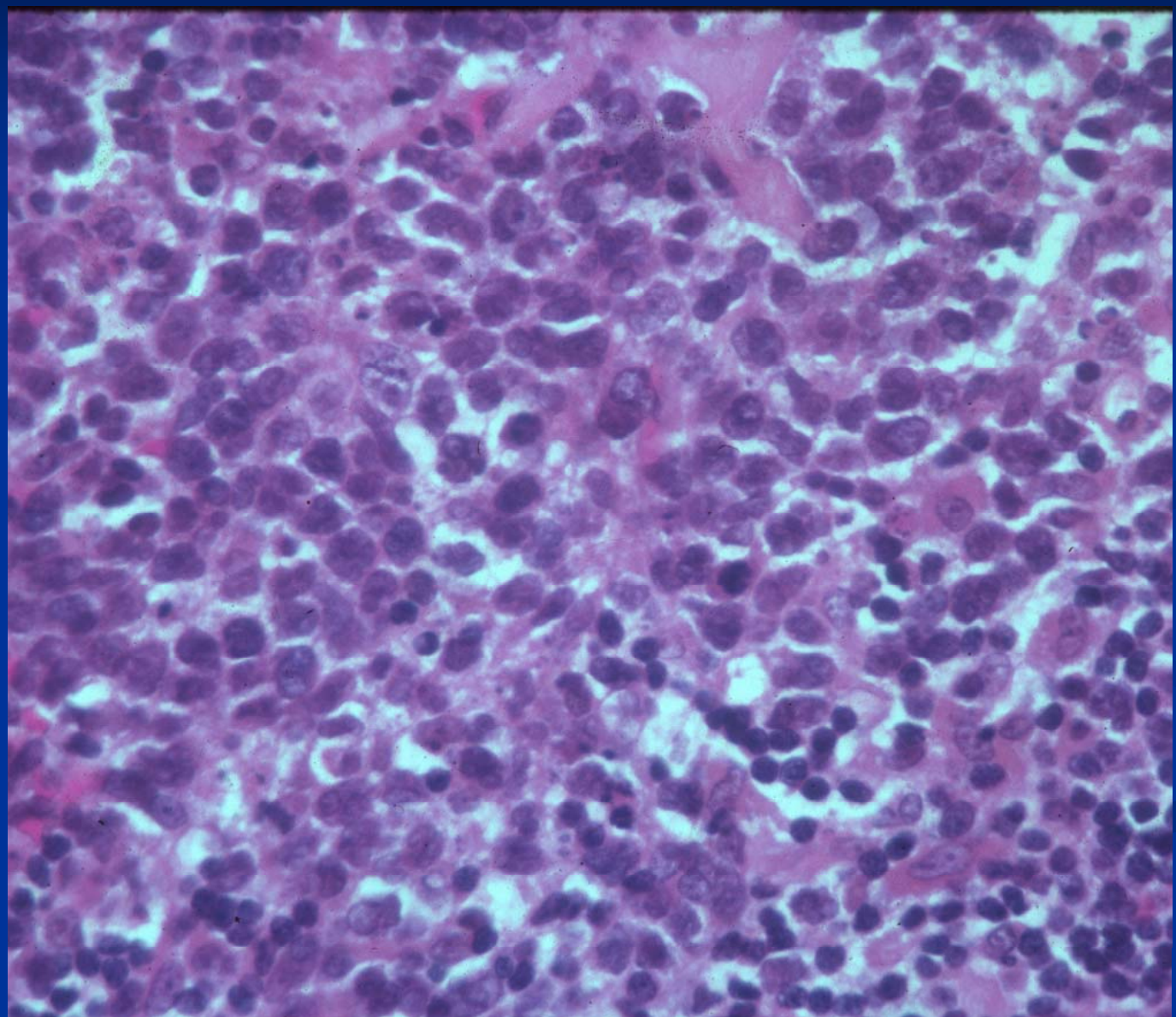

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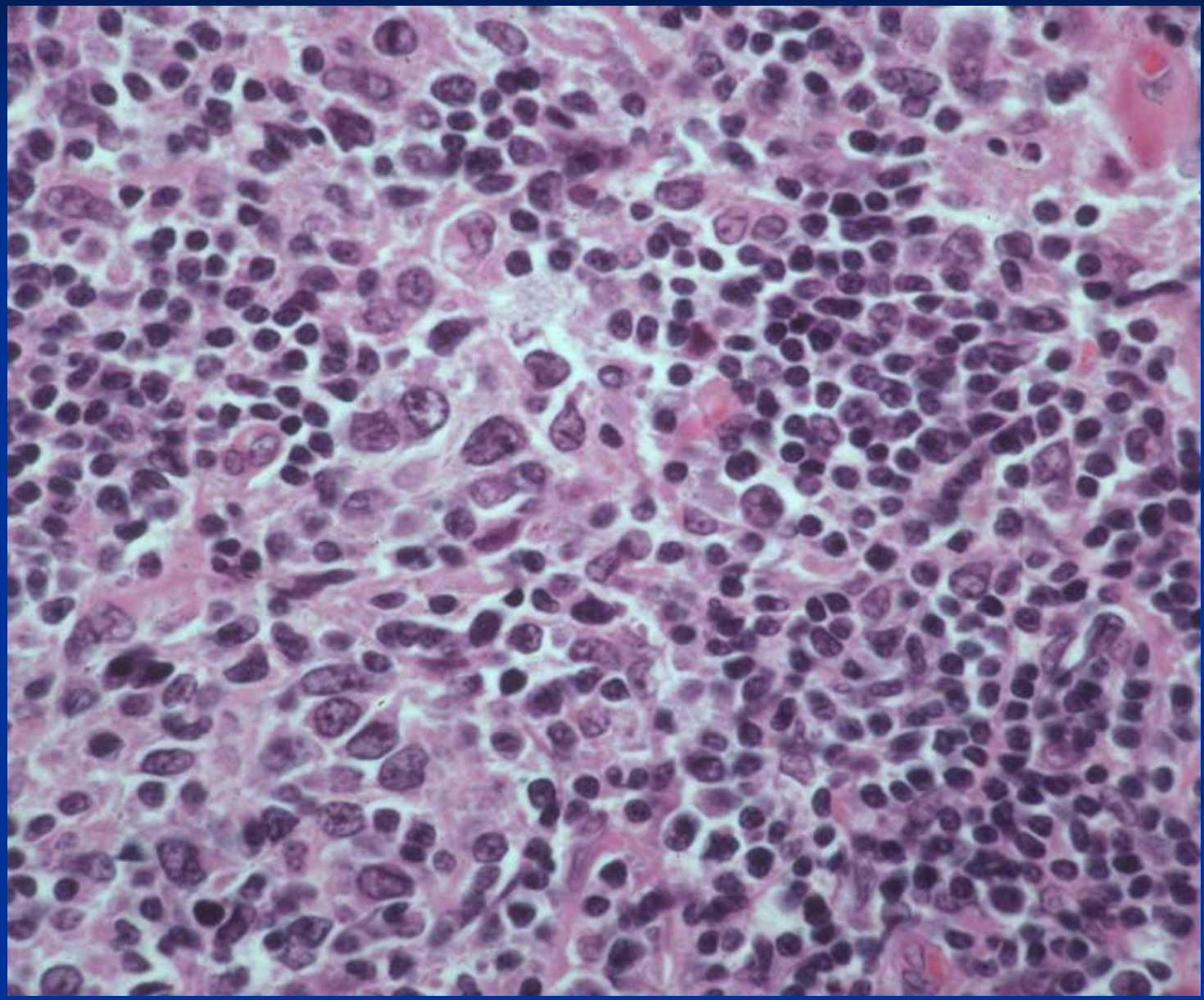

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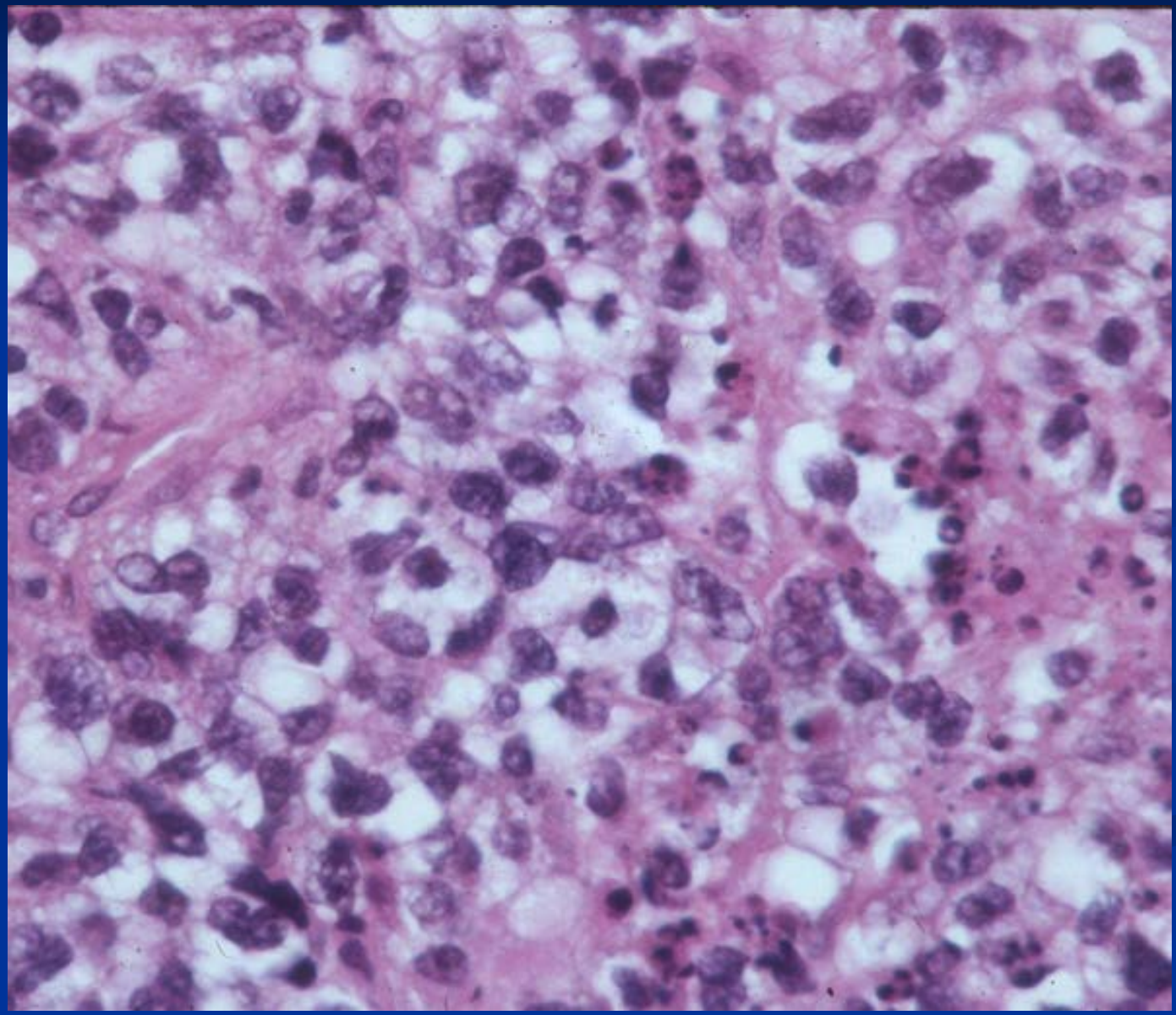
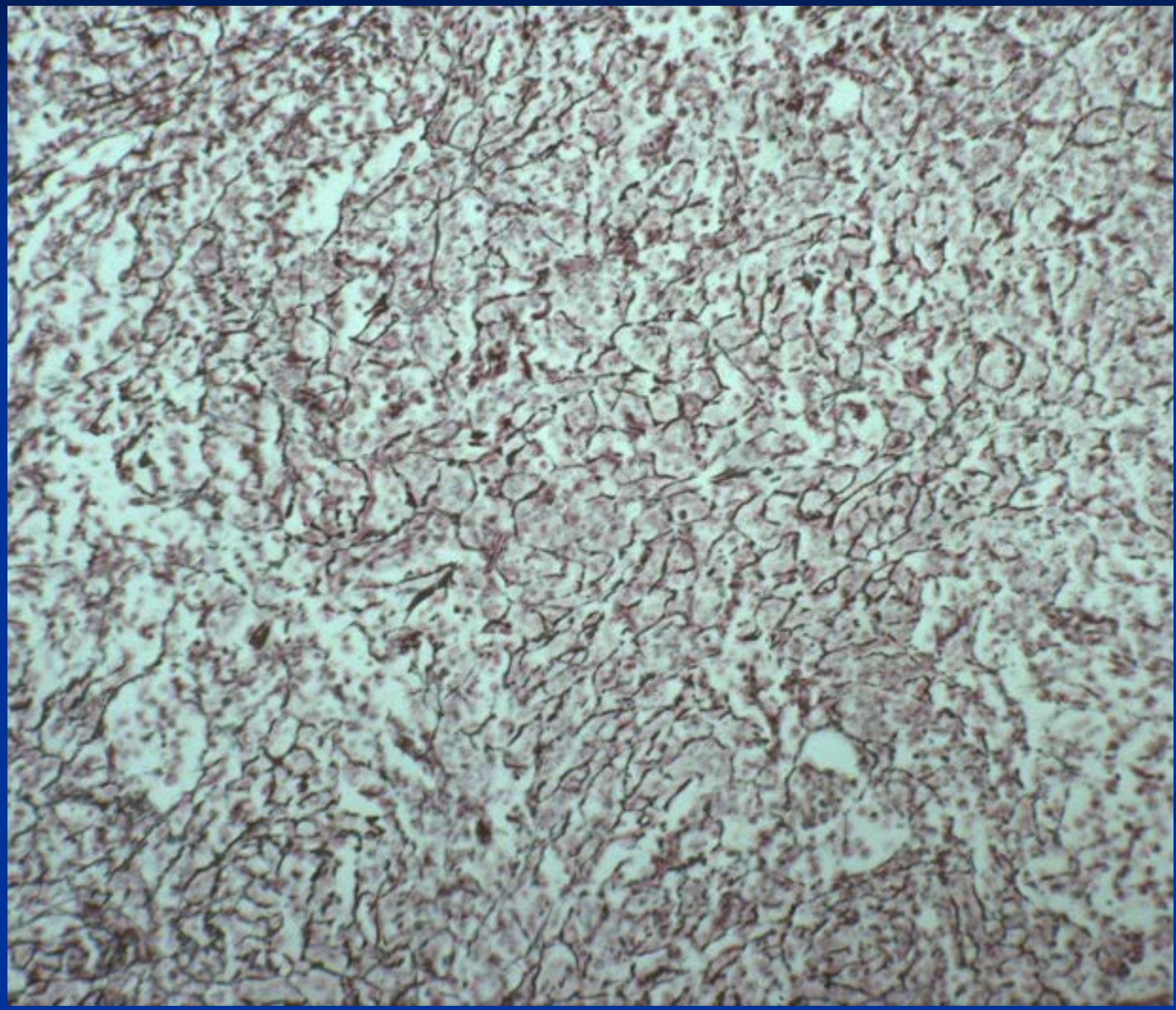


Lymphoma Reticulin



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

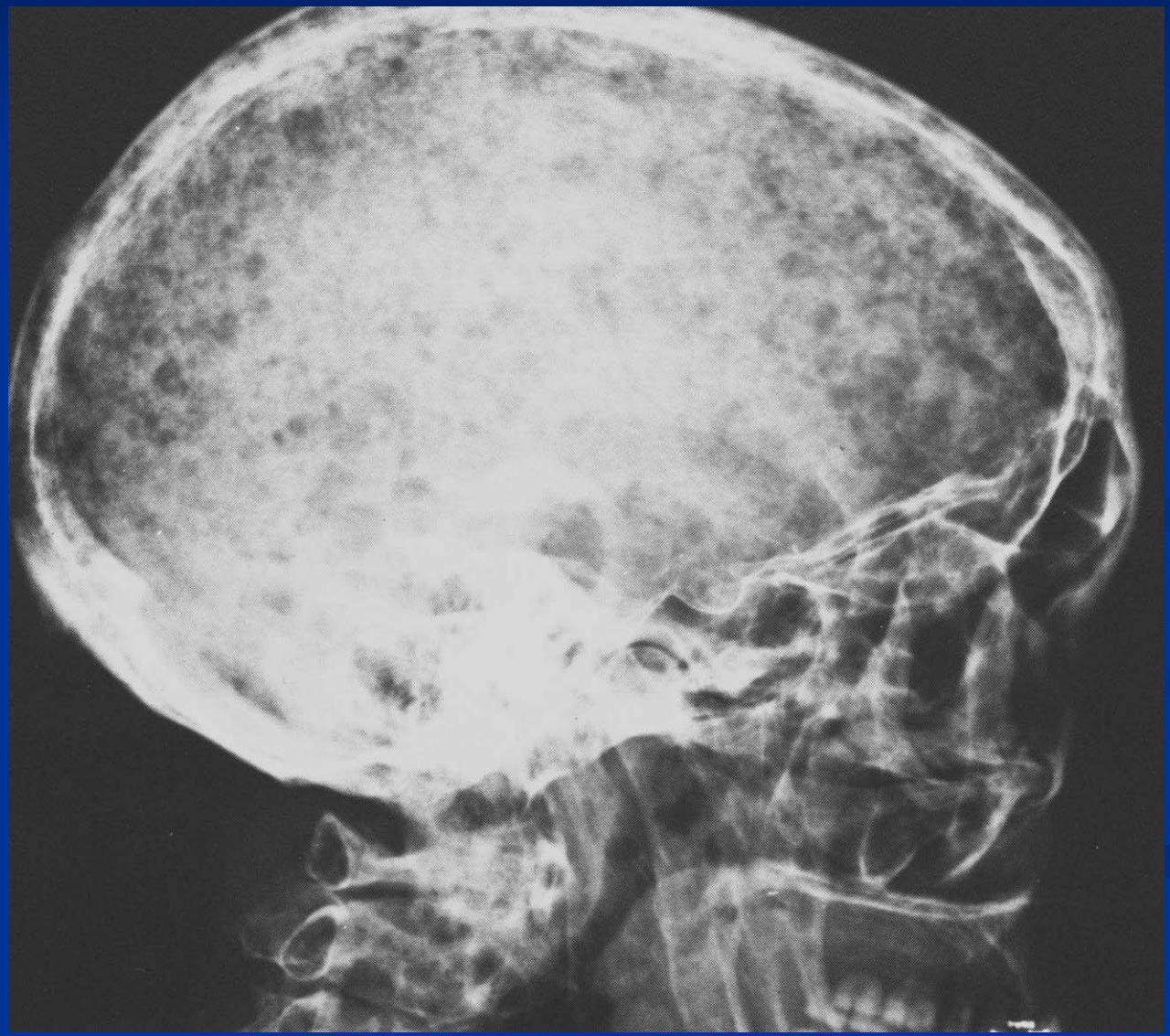
Myeloma

- 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

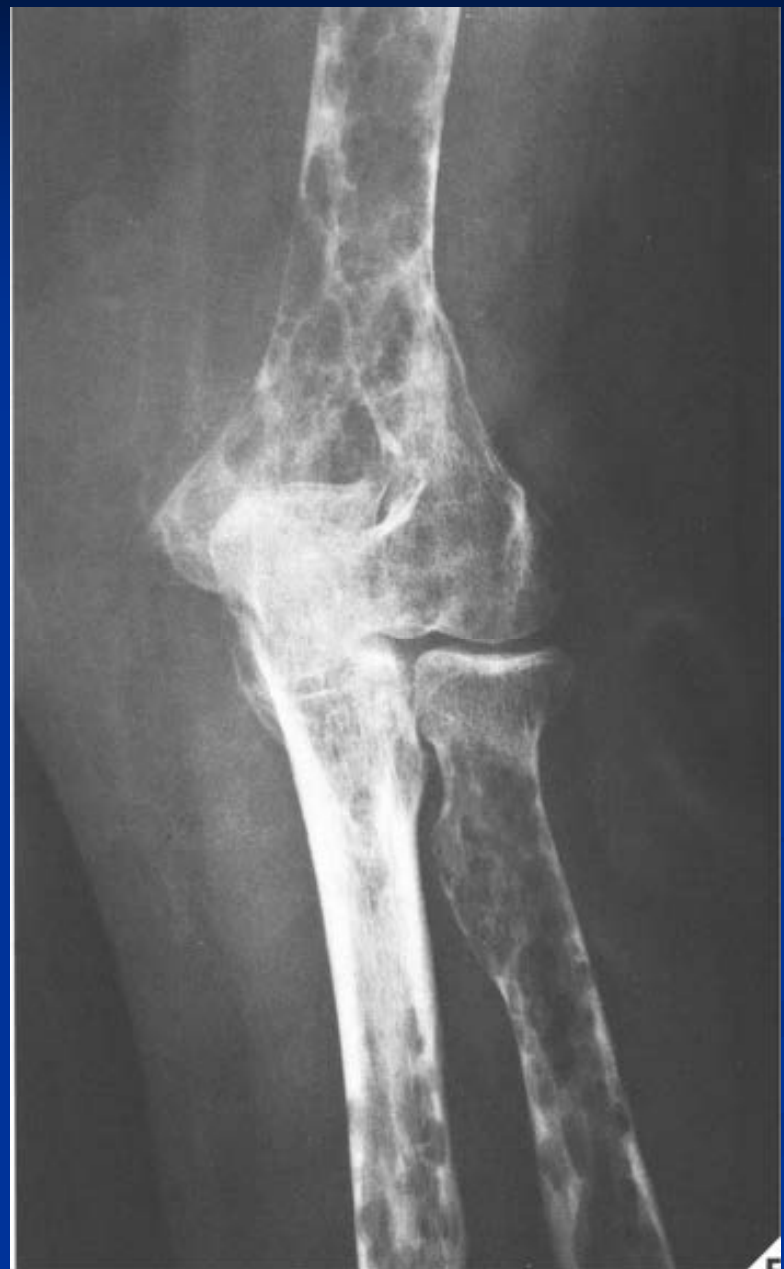
Myeloma

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

Myeloma



Myeloma



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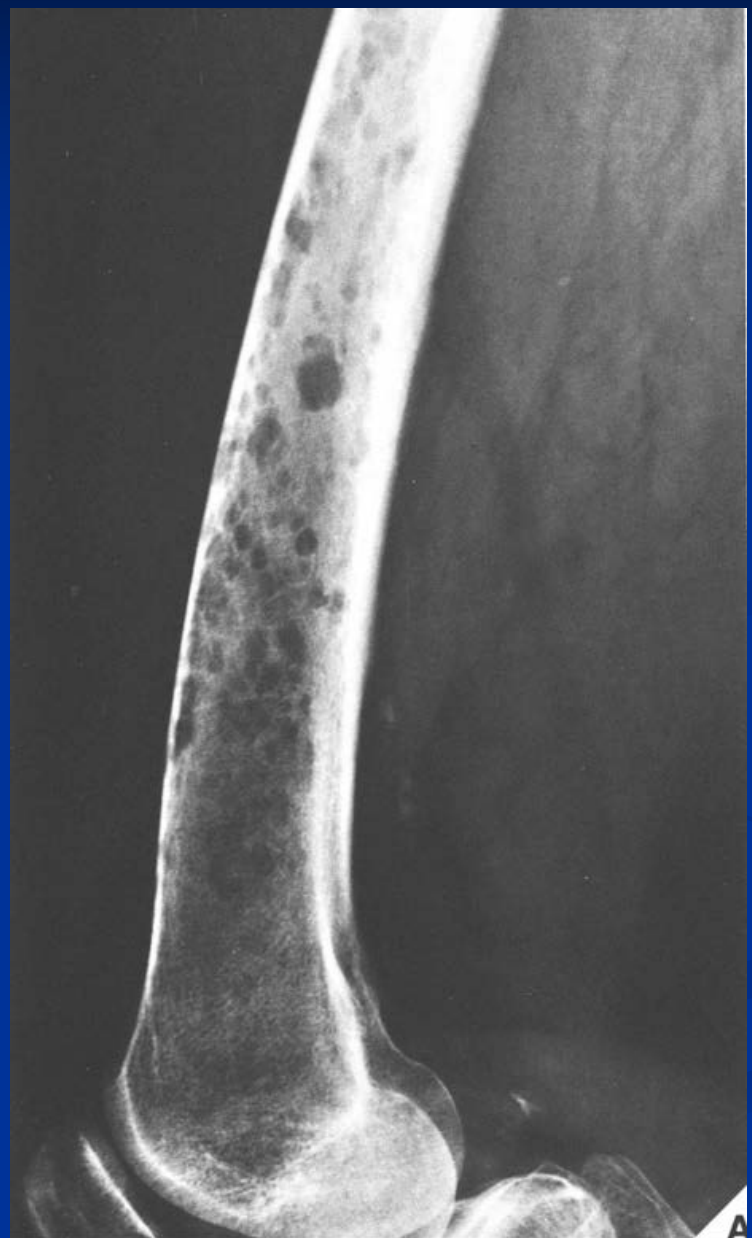

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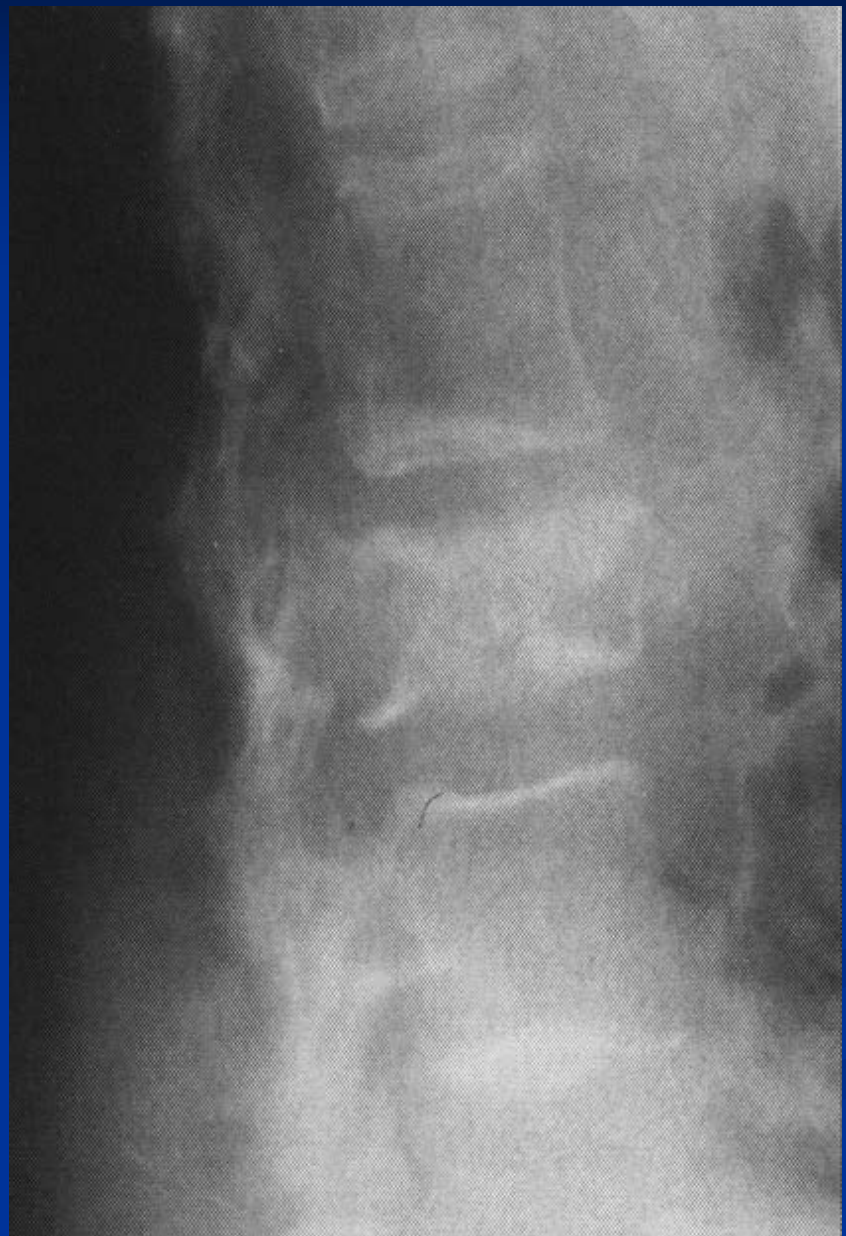

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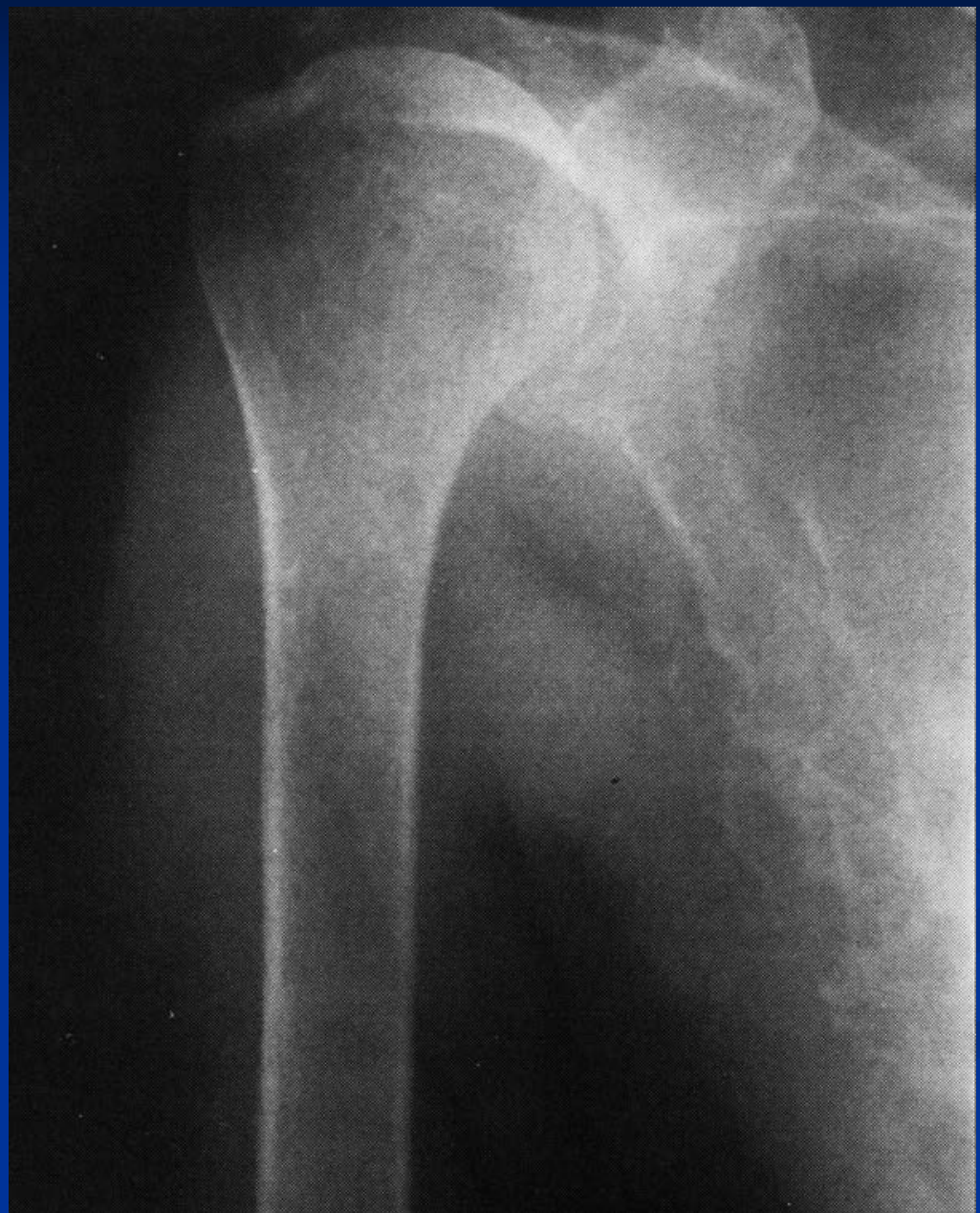
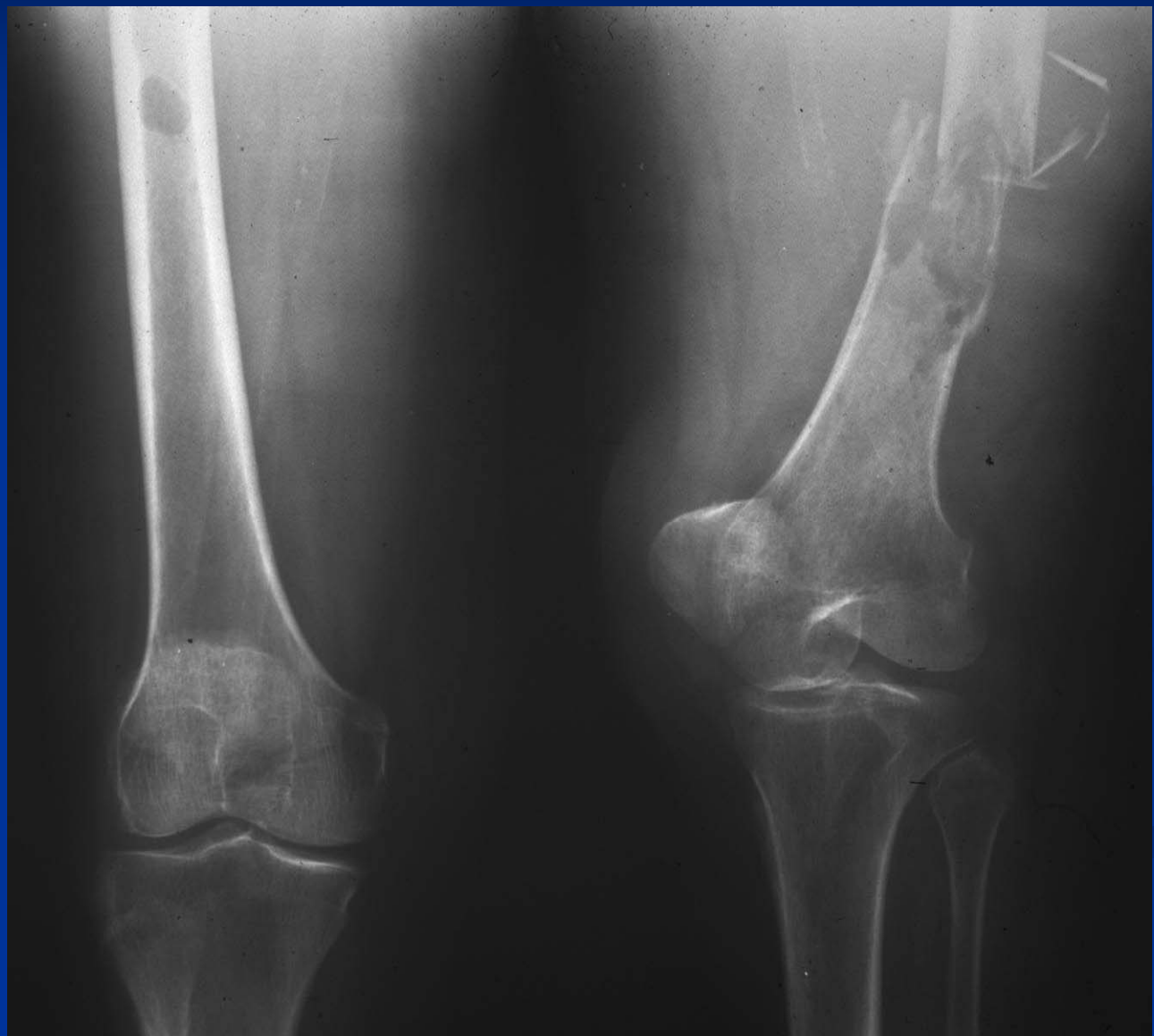
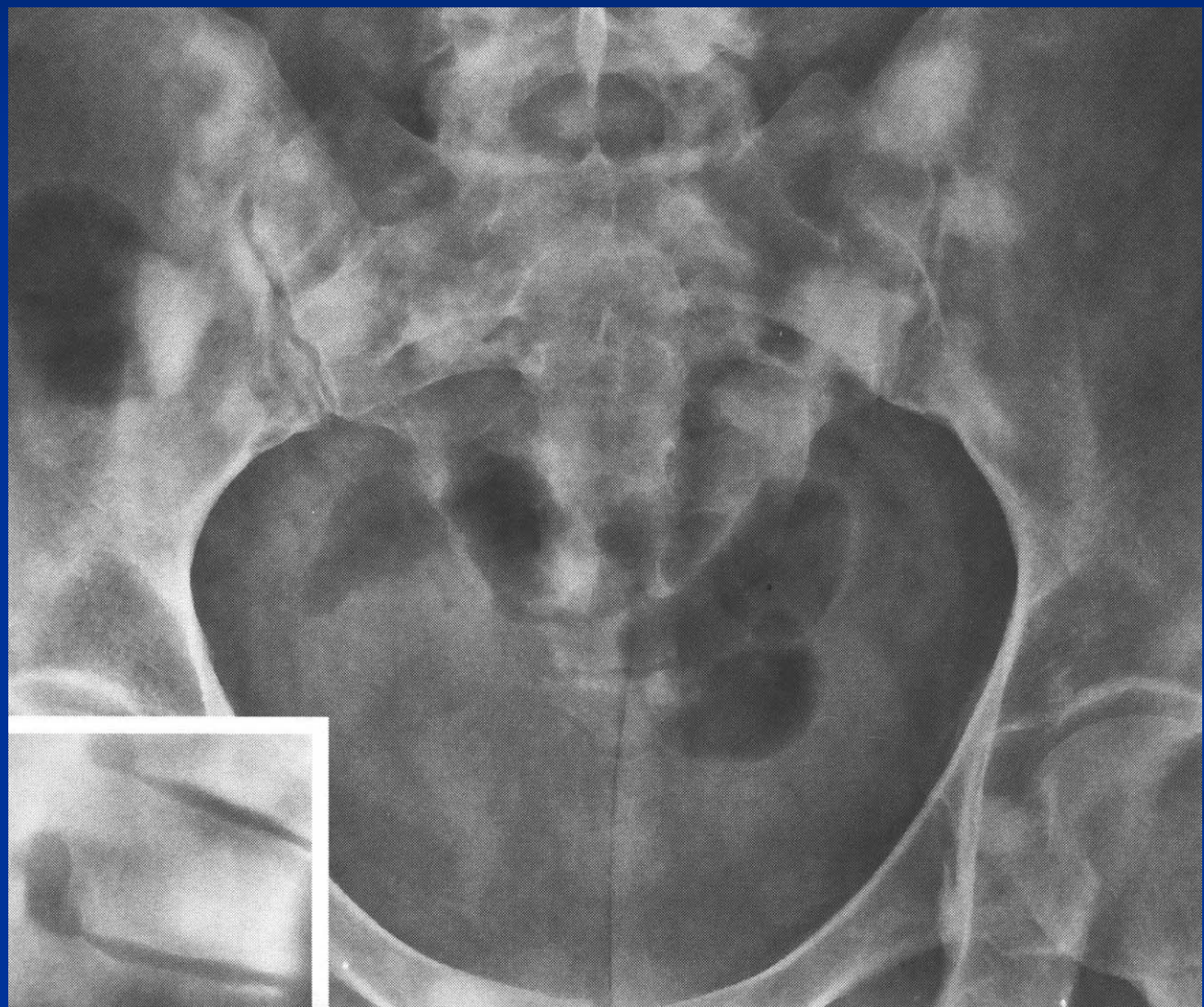


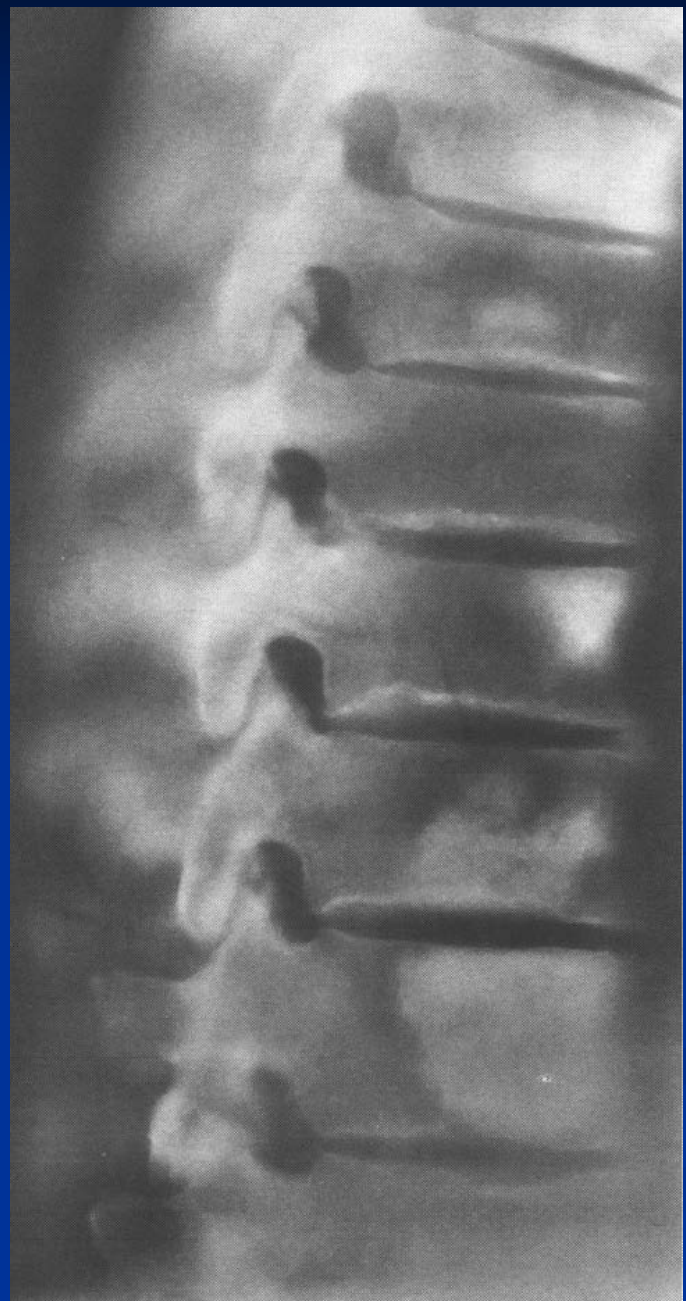
Myeloma



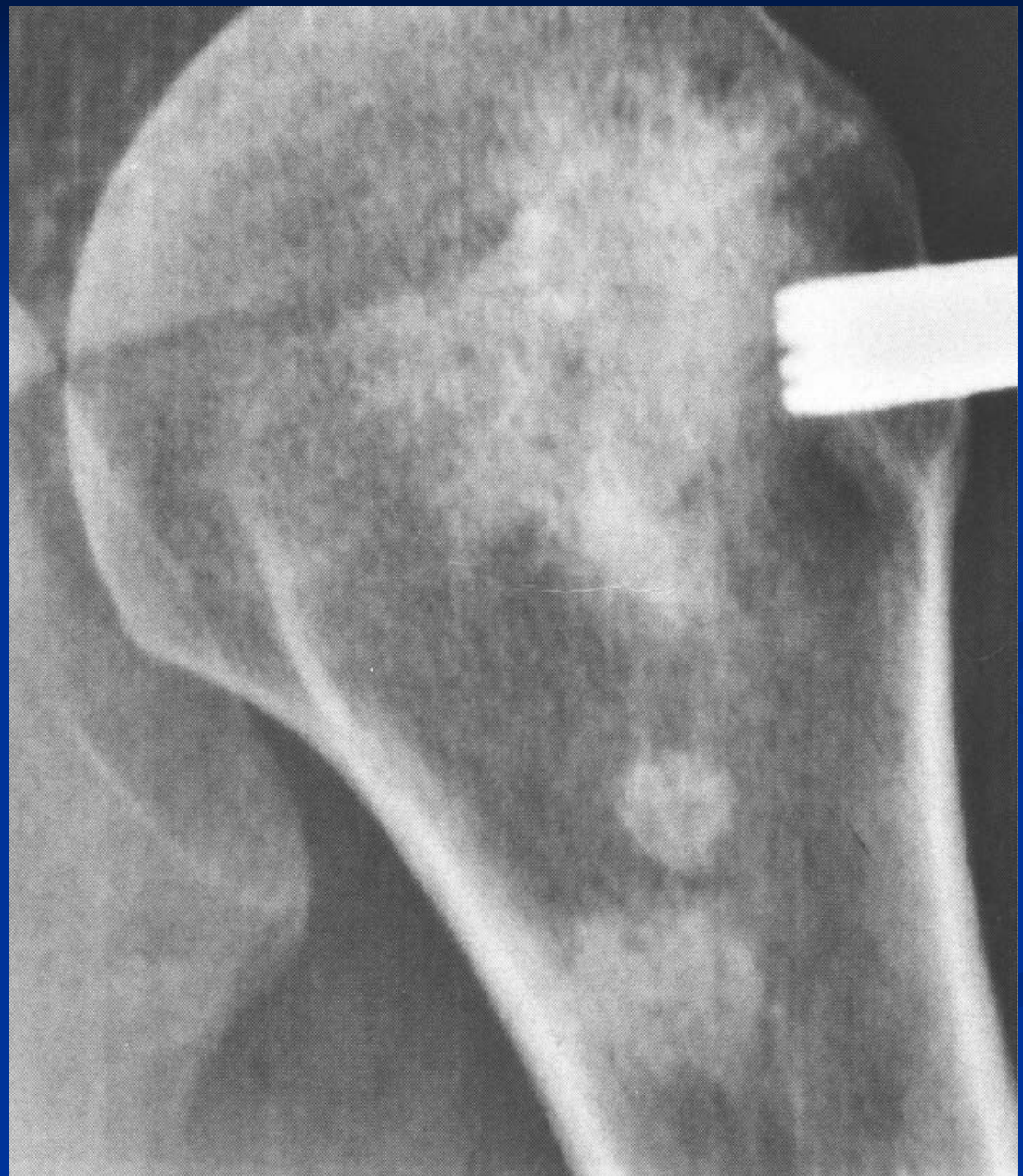
Osteosclerotic Myeloma



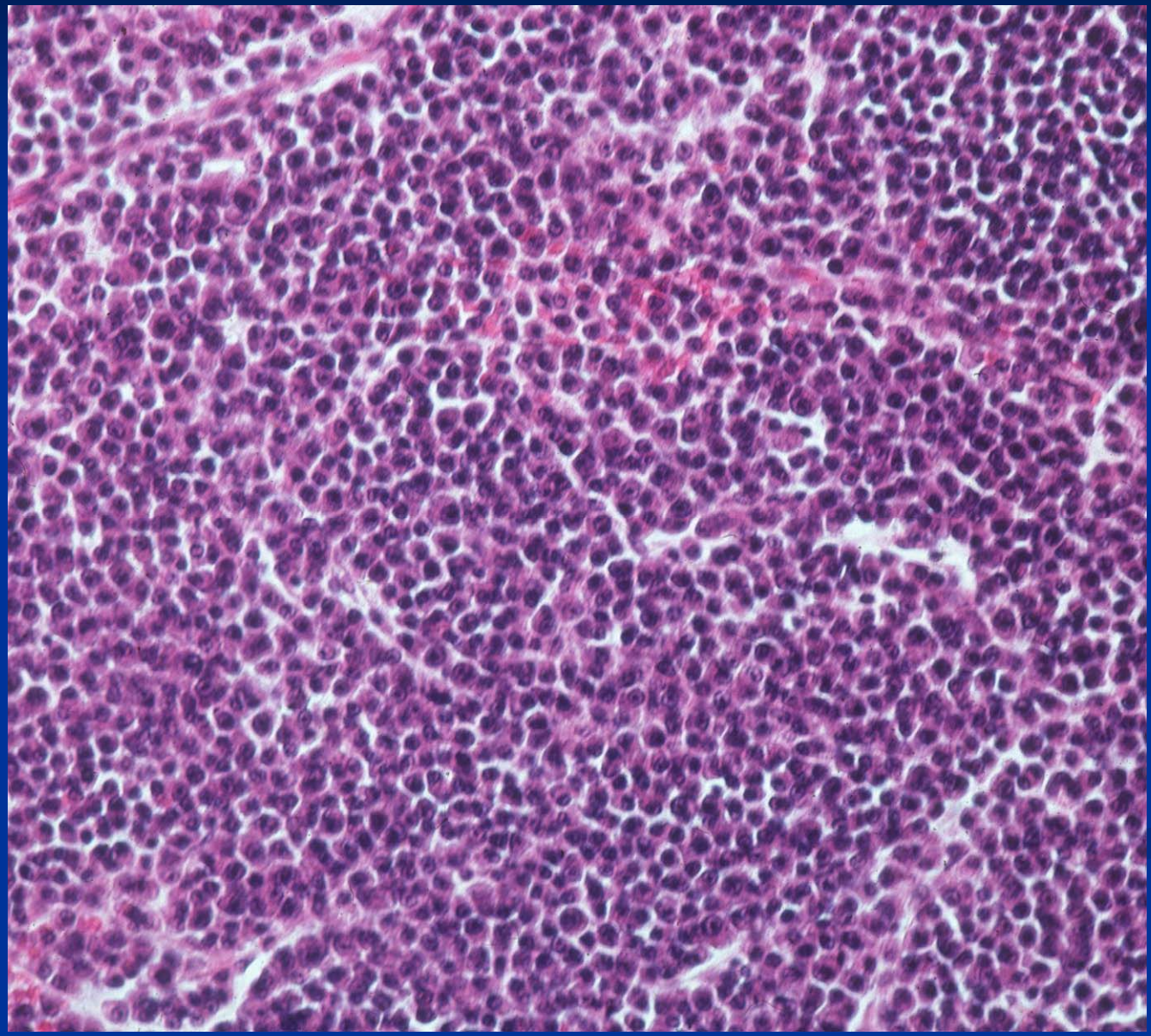
Osteosclerotic Myeloma



Osteosclerotic Myeloma



Myeloma



Myeloma