Tibial Lesions in Children

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Tibial Lesions in Children

- Fibrous Dysplasia
- Osteofibrous Dysplasia
- Adamantinoma

Differential Diagnosis
- Infection
- Eosinophilic Granuloma
- Nonossifying Fibroma
- Unicameral Bone Cyst
- Aneurysmal Bone Cyst
- Giant Cell Tumor
- Hemangioma
- Osteosarcoma
- Fibrosarcoma
- Ewing’s Sarcoma
- Multiple Enchondromatosis (Ollier’s)
- Brown Tumor (Hyperparathyroidism)
- Von Recklinghausen’s (neurofibromatosis)
Fibrous Dysplasia (FD) – Definition

- Developmental hamartoma
  - Benign tumor-like growth composed of differentiated cells in abnormal number or distribution

- Areas of the skeleton fail to mature normally and remain indefinitely as immature

- Poorly mineralized trabeculae
FD – Demographics

- Age: 5-15 years old
- Gender: Female > Male
- Sites: Solitary or Multiple
FD – Pathogenesis

- Developmental failure of primitive bone to mature and realign along stress lines
- Immature trabeculae enmeshed in primitive dysplastic fibrous tissue that are constantly turning over but never mature
- Immature matrix does not mineralize normally
- Lack of stress alignment + insufficient mineralization
  - Significant loss of mechanical strength
FD – Forms

Monostotic Form (Solitary)
- 70% of cases, Ages 10-30 years old
- Usually asymptomatic and commonly found incidentally
- Ribs > Femur > Tibia > Maxilla > Mandible > Skull > Humerus
- Bone deformity less severe
- **During skeletal immaturity** → lesions grow quicker than normal bone (causing expansion and potential complications)
- **After skeletal maturity** → gradual remodeling and slow maturation

Polyostotic Form (Multiple)
- 25% of cases, Unilateral distribution
- Femur > Craniofacial > Tibia > Humerus > Ribs > Fibula
- Gradual bowing deformities due to the loss of mechanical strength
- Leg Length Discrepancy
- **May continue enlarging after skeletal maturity with risk of pathologic fracture**
- Associated with McCune-Albright Syndrome
- Malignant transformation rare (<1%)
  - Fibrosarcoma
  - Osteosarcoma
  - Malignant fibrous histocytoma
FD – Forms

**Craniofacial Form**
- 20% in Monostotic Form, 50% in Polyostotic Form
- Frontal, Sphenoid, Maxillary, Ethmoidal bones
- **Frontal bossing, facial asymmetry**
- Inferolateral orbital displacement
- **Blindness**

**Cherubism**
- Autosomal-Dominant with variable penetrance
- Children, Males > Females
- Regression after adolescence
- Jaw is broad and protruding
- **Maxilla and Mandible are symmetric**

**General Associations**
- 2% Endocrinopathy (Hyperthyroidism, Hyperparathyroidism, Precocious Puberty)
- Polyostotic form has rare association with intramuscular myxomas (adjacent to affected bone) = Mazabraud Syndrome
FD – Natural History

McCune-Albright's syndrome (2-5%)
- Polyostotic fibrous dysplasia
- Females > Males
- Skin pigmentation
  - cafe au lait spots with irregular border
- Precocious puberty and Short stature
- Endocrine Abnormalities
  - Cushing’s, Hyperthyroidism, Acromegaly
- Skeletal lesions tend to be larger, more persistent and with greater complications than when only the skeleton is involved
FD – Clinical Presentation

Monostotic/Polyostotic Lesions

- **Asymptomatic, Incidental Finding**
- Large lesions can cause deformity (especially in maxilla and skull)
- **Classic deformity is “Shepherds' Crook” deformity of the proximal femur**
- **Pain with pathologic fracture**
- Extremity deformity may be present
- **Limb length discrepancy**
- **Hip motion defect**
  - Varus femur leads to abduction defect
FD - Radiology

- Within medullary canal
- "Ground Glass" pattern
- Thin reactive cortical shell
- Expanding lesion, bounded by cortical shell
- Proximal **Femur**
  "Shepherds' Crook"
- Anterior bowing of **Tibia**
  "Saber Shin"
FD - Radiology

- The callus formed around a pathologic fracture of FD acquires the dysplastic character of the lesion.
FD – Radiology

Bone Scan

- Increased uptake in lesion
- Usually well defined
FD – Radiology

CT Scan

- Homogeneous character
- Contrast shows vascularity in and around tumor
FD – Radiology

MRI
- T1 = Low Intensity Signal
- T2 = High Intensity Signal
FD – Biopsy Principles

- Incision in-line with resection incision
- Longitudinal in extremities
- Intramuscular (to bury hematoma)
- No skin or muscle flaps
- Avoid neurovascular structures and joints
- Meticulous hemostasis
- Tight closure
- Approach through soft tissue mass or weakened area of bone
- Drain if necessary → in-line and distal to incision
FD – Gross Pathology

- Yellowish-white tissue with gritty feel
- Vascular around the shell
- Can be peeled away from bony shell
FD - Pathology

- Fibrous tissue with irregularly shaped trabeculae of woven bone
  - Woven bone = immature bone not undergone remodeling into Lamellar bone

- Bone appears to arise directly from fibrous tissue and not from osteoblasts = unusual shapes
  “Chinese Characters”
FD - Histology

- Dysplastic trabeculae enmeshed in primitive mesenchymal cells
- **Lack** of osteoblastic rimming surrounding the dysplastic trabeculae
FD - Treatment

- Clinical Observation if incidental finding and asymptomatic

- SURGERY:
  - Pathologic Fracture or Impending Fracture
  - Disabling Lesions
  - Usually after skeletal maturity
  - No internal fixation into lesion because poor mechanical fixation strength
  - Curettage has high risk of recurrence/fracture
  - Cortical bone grafts are preferred

- NO Chemotherapy
- NO Radiation
FD - Treatment

- Cortical bone grafting

Pre-op: Pathologic Fracture  
Post-op  
Post-op: 1yr
FD - Complications

- Mechanical insufficiency of bone leads to microfractures and deformity

- Rare (<1%) transformation to sarcoma
  - Osteosarcoma
  - Fibrosarcoma
  - MFH of Bone
Osteofibrous Dysplasia (OD)

- Also known as
  - Ossifying Fibroma
  - Kempson-Campanacci Lesion
OD – Definition

- Congenital hamartoma occurring exclusively in Tibia or Mandible
- Benign active or aggressive
- Resembling fibrous dysplasia
OD – Demographics

- Age: 5-20 years old
- Gender: Male > Female (1.5:1)
OD – Natural History

- < 5 years old, rarely progress
- After skeletal maturity, expansion is rare

- Usually recur if treated with surgery (Curettage or Excision) before skeletal maturity

- Some heal spontaneously
- Some may progress to Adamantinoma (controversial)
OD – Clinical

- Often asymptomatic, incidental finding
- Pathologic fracture = pain
- ANTERIOR BOWING of TIBIA
OD – Radiology

- Multiloculated radiolucent
- Anterior Cortex, Tibia +/- ossification
OD – Radiology

**Bone Scan (Similar to FD)**
- Increased uptake in lesion
- Usually well defined

**CT Scan (Similar to FD)**
- Homogeneous character

**MRI (Similar to FD)**
- T1 = Low Intensity Signal
- T2 = High Intensity Signal
OD – Biopsy Principles

- Incision in-line with resection incision
- Longitudinal in extremities
- Intramuscular (to bury hematoma)
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- Avoid neurovascular structures and joints
- Meticulous hemostasis
- Tight closure
- Approach through soft tissue mass or weakened area of bone
- Drain if necessary → in-line and distal to incision
OD – Pathology

- **Gross**: Fibrous tissue with some mixed bone spicules
  
- **Micro**: Trabeculae of woven bone in fibrous stroma, and poorly mineralized immature trabeculae
  
- **BONE** is rimmed with Osteoblasts and is distinct from stroma. This differentiates OD from Fibrous Dysplasia (FD)
OD – Histology

- Osteoblastic rimming
- Poorly mineralized immature trabeculae
OD - Treatment

- Asymptomatic – Observation
- Delayed excision + bone grafting for painful deformity
  Preferably AFTER skeletal maturity, because has high rate of recurrence if done before skeletally maturity
- NO Chemotherapy
- NO Radiation
Adamantinoma

- Rare, low-grade malignant tumor of bone
- Cell of origin uncertain
- May arise from Osteofibrous Dysplasia (controversial)
- 1% of malignant bone tumors
Adamantinoma – Demographics

- Age: 10-40 years
- Gender: Male > Female
- Tibia, Fibula, Mandible
Adamantinoma – Pathology

- Epithelial and Mesenchymal cells

- The presence of mesenchymal cells, makes this tumor look like a malignant variant of fibrous dysplasia (FD) or Osteofibrous Dysplasia (OD)

- But, the presence of epithelial cells, makes this a distinctly different tumor than FD/OD
Adamantinoma – Natural History

- Low-Grade
- Slow-Growing
- 15% Metastasize
  - Sites: Bones, Lymph Nodes, Lungs
  - Death usually @ 30-50 years old if metastasize
Adamantinoma – Clinical

- Localized insidious pain
- Gradual swelling, deformity in lower leg
- Pathologic fracture = pain
Adamantinoma – Workup

- Local and Systemic Staging
  - Bone Scan (increased uptake of Tc-99m)
  - CT Chest, Abd, Pelvis
  - MRI: Best for Local staging
    - T1 = low intensity signal
    - T2 = high intensity signal
  - Angiogram *** Highly Vascular Lesion
  - Blood work
Adamantinoma – Radiology

- “Bubbly" radiolucent lesion
- Anterior Tibial cortex
Adamantinoma – Radiology

- Angiogram *** Highly Vascular
Adamantinoma – Radiology

- MRI
Adamantinoma – Biopsy Principles

- Incision in-line with resection incision
- Longitudinal in extremities
- Intramuscular (to bury hematoma)
- No skin or muscle flaps
- Avoid neurovascular structures and joints
- Meticulous hemostasis
- Tight closure
- Approach through soft tissue mass or weakened area of bone
- Drain if necessary in-line and distal to incision
Adamantinoma – Pathology

- **Gross**: gray-tan tissue

- **Micro**:  
  - Epithelial cells in fibrous stroma  
  - Immature mesenchymal cells
Adamantinoma – Histology

- Spindle pattern has nests or columns of *epithelioid* spindle cells in an alveolar pattern
- **Mesenchymal**: loose arrangement of immature spindle cells with little collagen
Adamantinoma – Histology

- Hematoxylin and Eosin (H & E) stain
  - H = Blue-Purple (nucleic acids) = Basophilic
  - E = Red-Pink (intracellular or extracellular proteins) = Eosinophilic

- Keratin Stain (for epithelial cells)
Adamantinoma – Treatment

- **Wide Surgical Excision**
  - Reconstruction with allograft, autograft, or vascularized autograft (fibula)
  - Amputate if negative margins can not achieved
  - Consider **Embolization**

- **No Curettage = High rate of Recurrence**

- **No Chemotherapy**
- **No Radiation**
Adamantinoma – Treatment
Summary

- **Fibrous Dysplasia**
  - Benign, Slow-Growing, Monostotic vs. Polyostotic
  - Asymptomatic, usually incidental finding
  - Classic deformity = “Sheppard’s Crook”
  - Radiology = “Ground Glass” Cortically Based Diaphyseal lesion, NO periosteal expansion
  - Pathology = Disorganized Fibrous Tissue, NO osteoblastic rimming, NO epithelial cells
  - Treatment = Observation, Surgery if Fracture/Deformity
Summary

- **Osteofibrous Dysplasia**
  - Benign active or aggressive
  - Classic deformity = Anterior Tibial Bowing
  - Radiology = “Ground Glass” Cortically Based Diaphyseal lesion, Periosteal Expansion
  - Pathology = Disorganized Fibrous Tissue, Osteoblastic rimming, NO epithelial cells
  - Treatment = Observation, Surgery if Fracture/Deformity
Summary

- Adamantinoma
  - Low-grade malignant tumor
  - Classic deformity = Anterior Tibial Bowing
  - Radiology = “Bubbly” Cortically Based Diaphyseal lesion, Periosteal Expansion
  - Pathology = Disorganized Fibrous Tissue, Osteoblastic rimming, Epithelial cells (Keratin Positive)
  - Treatment = Wide Surgical Resection
Summary – Histology

<table>
<thead>
<tr>
<th>Fibrous Dysplasia</th>
<th>Osteofibrous Dysplasia</th>
<th>Adamantinoma</th>
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<tbody>
<tr>
<td>NO Osteoblastic Rimming</td>
<td>Osteoblastic Rimming</td>
<td>Keratin Stain</td>
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Thank You

If you'd taken better care of yourself, this wouldn't be happening.
IT'S OKAY, IT'S JUST A BENIGN TUNA.
FD – Radiograph – 7 Questions

1. Location
2. Size
3. Tumor effect on Bone
4. Bone response to Tumor
5. Matrix
6. Cortex
7. Soft tissue mass
FD – Radiograph – 7 Questions

Monostotic Lesions

1. Location
   - Ribs > Femur > Tibia > Maxilla > Mandible > Skull > Humerus
   - Usually spare epiphysis, but can occur in any part of the bone

2. Size
   - Variable sizes

3. Tumor effect on Bone
   - “Ground Glass”, Lytic, Pathologic Fracture

4. Bone response to Tumor
   - Well-demarcated sclerotic border, minimal periosteal new bone formation

5. Matrix
   - Depends on quantity of osseous matrix (osseous tissue = ground glass)

6. Cortex
   - Thinned with endosteal expansion

7. Soft tissue mass
   - None
FD – Radiograph – 7 Questions

Polyostotic Lesions

1. Location
   - Femur > Craniofacial > Tibia > Humerus > Ribs > Fibula

2. Size
   - Variable sizes

3. Tumor effect on Bone
   - “Ground Glass”, Lytic, Geographic Border, Pathologic Fracture

4. Bone response to Tumor
   - Endosteal expansion with periosteal new bone

5. Matrix
   - Lytic or Ground Glass, depends on degree of ossification

6. Cortex
   - Endosteal Scallop, Thinned with endosteal expansion

7. Soft tissue mass
   - None
OD – Radiograph – 7 Questions

1. Location
   - Tibia (usually proximal 1/3) anterior cortex, Fibula
   - Eccentric, Diaphyseal > Metaphysis

2. Size
   - Variable

3. Tumor effect on Bone
   - Osteolytic

4. Bone response to Tumor
   - Geographic with periosteal expansion
   - Solitary/Multiple cortical lucency (ground glass surrounded by dense sclerosis)

5. Matrix
   - None, Lytic

6. Cortex
   - Irregular, Thin, Expanded

7. Soft tissue mass
   - None
Adamantinoma – Radiograph – 7 Q’s

1. Location
   - 90% involve Tibial diaphysis, Ulna and Fibula also common
2. Size
   - Variable, may have skip lesions
3. Tumor effect on Bone
   - Osteolytic, Bony Expansion
   - Geographic and well defined
4. Bone response to Tumor
   - Eccentric lucencies with surrounding reactive bony sclerosis
   - Cortex may be eroded with well-organized periosteal expansion
5. Matrix
   - Lytic with mixed ground glass density
6. Cortex
   - Anterior cortically based lesion in the Tibia is characteristic
7. Soft tissue mass
   - May be present, especially in aggressive lesions