

# 17 y/o female with incidental finding on knee radiograph

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RADIOLOGY



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A large, stylized oak leaf graphic in a dark blue color, positioned on the left side of the slide. The leaf has a prominent central vein and several smaller veins branching off it. The leaf's edge is serrated.

# Fibrous Dysplasia



- Well defined expansile lesion with smooth sclerotic margins and a hazy matrix in the distal femur
- No periostitis

# Fibrous Dysplasia

- Non-inherited bone disease with abnormal differentiation of osteoblasts
  - Normal marrow and cancellous bone replaced by immature bone and fibrous stroma
- Usually an incidental finding
- May be complicated by pathologic fracture
- Very rarely can undergo malignant degeneration
- Will not have periostitis
- Not painful (unless with an associated fracture)
  - Occult fractures occur in long bones (weight bearing)
  - Fractures in non-weightbearing bones should not occur

# Histopathology

- Irregular spindles of woven bone scattered throughout a fibrocellular matrix
- Degree of radiographic haziness of the lesion correlates directly with the histopathology
  - More radiolucent: composed of mostly fibrous elements
  - More radiopaque: composed of more woven bone

# Monostotic Fibrous Dysplasia

- 80% of all cases.
- 10-70 years old
- Most commonly involves ribs, femur, tibia, mandible, skull, humerus
- Generally uncomplicated and do not cause significant deformity
- General rule:
  - Won't convert to polyostotic form
  - Lesions do not increase in size over time
  - Becomes inactive at puberty



# Polyostotic Fibrous Dysplasia

- May involve many or few bones.
- Often unilateral
- Most commonly involves skull and facial bones, pelvis, spine, and shoulder.
- Involves large segments of bone and is frequently associated with fractures and severe deformities.
  - Bowing deformity with varus angulation of proximal femur (shepherd's crook)
  - Cranial nerve dysfunction with visual and hearing impairment if skull is involved
- Does not proliferate or spread
- Usually becomes inactive at puberty
  - Existing deformities may progress

# Complications

- Pathologic fractures
- Malignant degeneration (<1% of all cases)
  - Presents as pain and swelling
  - Cortical destruction
  - Soft tissue mass
  - Osteosarcoma, fibrosarcoma, malignant fibrous histiocytoma

# Associations

- McCune-Albright syndrome
  - Endocrinopathy mostly in girls
  - Triad: precocious puberty, polyostotic fibrous dysplasia, and café au lait spots
- Mazabraud syndrome
  - Fibrous dysplasia and soft-tissue myxomas (benign)
  - Soft-tissue myxomatous tumors develop in muscle near the most extensively involved bones years after the initial diagnosis
  - Higher incidence of transformation to osteosarcoma
- Adamantinoma
  - Appears very similar to fibrous dysplasia, but almost exclusively occurs in the tibia and jaw
  - Malignant tumor
  - Very rare

# References

Imaging Findings of Fibrous Dysplasia with Histopathologic and Intraoperative Correlation

Kimberly A. Fitzpatrick, Mihra S. Taljanovic, Donald P. Speer, Anna R. Graham, Jon A. Jacobson, George R. Barnes, and Tim B. Hunter

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