Fibrous Dysplasia in Images

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Agenda

• Patient Presentation
• Radiographic Differential Diagnosis
• Fibrous Dysplasia
  – Bone biology
  – Types
  – Clinical Presentation
• Imaging of Fibrous Dysplasia
Our Patient History

• A 37 year-old woman
• CC: Left distal thigh pain
• HPI:
  – Vague complaints of left lower extremity pain for ~2 years
  – Over last 5-6 months, distal thigh pain on her left side causing her to favor this leg and have difficulty with daily living activities
  – Denies any history of trauma, prior surgical or other problems with this leg
Our Patient: Femur Plain Films
37 yo female, pain in L lower extremity

Quiz: Do you see anything abnormal?
Features of this lesion:

- Location: eccentric, distal metaphysis
- Relatively thick, sclerotic border
- No definite internal mineralization
- Non-aggressive features
- No periosteal reaction, no cortical destruction, no apparent endosteal scalloping
Our Patient: Femoral lesion on CT

- Lucent lesion
- Eccentric location
- No periosteal reaction, no endosteal scalloping
- Thick sclerotic border of lesion with narrow zone of transition
- No soft tissue abnormalities seen
Radiologic Differential Diagnosis for a Lucent Lesion of Bone

- Fibrous dysplasia
- Osteoblastoma
- Giant cell tumor
- Metastasis / Myeloma
- Aneurysmal bone cyst
- Chondroblastoma / Chondromyxoid fibroma
- Hyperparathyroidism (brown tumors) / Hemangioma
- Infection
- Non-ossifying fibroma
- Eosionophilic granuloma / Enchondroma
- Solitary bone cyst
Non-Ossifying Fibroma versus Fibrous Dysplasia

Question: Does our patient’s lesion look more like fibrous dysplasia or a non-ossifying fibroma?

Fibrous Dysplasia

www.bonetumor.org

Our patient’s lesion
BIDMC, PACS

Non-ossifying fibroma

www.bonetumor.org
Non-Ossifying Fibroma versus Fibrous Dysplasia

**Answer:** The question cannot be answered without a biopsy. So, our patient underwent a biopsy...
Biopsy – Pathology Report

• Trabecular bone with an intertrabecular proliferation of fibrous tissue

• No significant mitotic figures nor atypical mitoses

• Not an adequate explanation for the two-year history of pain except to suggest that there could have been periodic microfractures

• Most consistent with fibrous dysplasia
Treatment

• Goal: prevention of deformity and fracture

• Patient was taken to the OR:
  - Curettage and allograft bone packing
  - Prophylactic fixation using four hole locking plate of left distal femur
Fibrous Dysplasia
Etiology

- Activating missense mutation in the Gs alpha gene
- Leads to over activity of adenylate cyclase
- Result is increased cell proliferation, inappropriate cell differentiation leading to disorganized fibrotic bone matrix
Bone Biology

- Normal medulla space of the bone is replaced by fibroosseous tissue
- Microscopic features: irregularly shaped trabeculae of immature (woven) bone in a cellular, loosely arranged fibrous stroma

Shah, 2005
Conditions with FD

• Monostotic
• Polyostotic
• McCune-Albright (3% of patients with FD)
  – Polyostotic FD
  – Café-au-lait spots
  – Hyperfunctioning endocrinopathies
• Mazabraud’s syndrome
  – Intramuscular myxomas
  – Usually seen with polyostotic FD
Monostotic Fibrous Dysplasia

- Six times more common than polyostotic FD
- Lesions may be asymptomatic
- Can present at any age, but most common in first or second decade of life
- Symptoms when present: pain, tenderness, swelling, stress or overt fracture
- Generally requires a bone biopsy for diagnosis, in addition to radiographs

Kransdorf, 1990
Polyostotic Fibrous Dysplasia

- Presents earlier than monostotic, mean age of 8
- 2/3 of patients have symptoms before the age of 10: leg pain, limp, pathologic fracture
- 85% develop pathologic fractures
- Can be diagnosed based on clinical symptoms and radiological images (biopsy unnecessary for diagnosis)
- “Monomelic” distribution is common

Bone scan of patient with FD

Commonly affected bones:
- Femur (91%)
- Tibia (81%)
- Pelvis (78%)
- Foot (73%)

Kransdorf, 1990
Complications of Fibrous Dysplasia

- Deformity
  - Leg length discrepancy
  - “Shepherd’s crook”
  - Facial asymmetry
  - Tibial bowing
  - Rib deformity
- Pathologic fractures
- Recurrence following surgery
- Associated tumors (< 1% develop osteosarcoma)

Gould, 2007
Menu of tests for imaging fibrous dysplasia

- Scintigraphy
- Plain film (of areas demonstrating increased uptake of the radioisotope)
- CT is useful for confirming the diagnosis and the extent of the FD in the craniofacial skeleton
- * MR is increasingly being studied and used as an imaging modality of FD
Companion Patient #1: FD on Scintigraphy

• Typically exhibits markedly increased radionuclide accumulation in early perfusion and delayed bone imaging (but non-specific)

• Useful for visualizing polyostotic involvement where only monostotic disease was suspected

• Useful for determining when plain radiographs of the spine are needed for clinical management

Bone scan showing femur and tibia FD
Companion Patient #2: FD on Plain Film

- Lesions medullary in origin
- Sometimes eccentric in location
- May be lucent, sclerotic, mixed, or resembling ground glass
- Often causes “scalloping” of the endosteum
- Lesions typically sharply marginated

Frontal film: right femur

Gould, 2007
Fibrous Dysplasia on CT

• Similar to findings on plain films
• Lesions can be radiolucent or radio opaque
• Confirms extent, specific dimensions and radiodensity of FD with a high degree of precision
• Useful in determining any long-term changes associated with the progression of FD, or reactivation of the lesion growth
• Especially useful in evaluating craniofacial FD
Craniofacial FD

• Clinical Presentation
  – Atypical facial pain or headache
  – Sinus congestion or infection
  – Orbital involvement (proptosis, diplopia, visual changes)
  – Facial/skull enlargement / Asymmetry
  – Hearing loss

• Sphenoid and ethmoid are commonly involved

• Most frequent site for sarcomatous transformation is the craniofacial skeleton

Kransdorf, 1990
Posteroanterior view: obliteration of frontal and sphenoid sinuses, inferior displacement of the left orbit

Lateral view: thickening of the occiput

Kransdorf, 1990
Fibrous dysplasia involving the sphenoid and temporal bones. Patient presented with symptoms of orbital compression and proptosis. No symptoms

Companion Patient #4: Craniofacial FD on CT

Extensive fibrous dysplasia of the entire skull base

Lustig, 2001
Fibrous Dysplasia on MRI

- Shows location and extent of lesions better than radiographs.
- On T1, lesions are largely isointense to skeletal muscle with areas of hypointensity.
- On T2, lesions appear heterogeneously hyperintense with hypointense, isointense or markedly hyperintense areas within.
Companion Patient #5: FD on MRI

Expansile lesion in femoral neck and upper shaft with a thick sclerotic rim and matrix calcifications

T1: lesion iso-intense to muscle

T2 fat suppressed: heterogeneously hyperintense lesion with small hypointense foci distally

Images from Shah, 2005
Summary of Radiographic Appearance of FD

- Location: centrally within the metaphysis or diaphysis (can be eccentric)
- Sharp margins
- “Ground glass” appearance of the matrix
- Absence of cortical destruction
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References