Fibrous bony lesions

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SpR Histopathology
Fibrous bony lesions

- Fibrous cortical defect and non-ossifying fibroma
- Fibrous dysplasia
- Fibrosarcoma of bones
- Adamantinoma of long bones
Outline:

- Epidemiology
- Pathogenesis
- Presentation
- X ray features
- Histological features
- Treatment
Fibrous cortical defect and Non ossifying fibroma

- Developmental defects
- Non neoplastic
- Fibrous lesions
- 30-50% children
- Metaphyseal
- Site: long bone, usually LL.
Fibrous cortical defect and Non ossifying fibroma

- Similar histologically
- Differences:
  - Size
  - Natural history.

- FCDs
  - Small (<3 cm), eccentrically located
  - Metaphyseal *cortical* defects
  - Asymptomatic
  - Most of these spontaneously disappear
Eccentric Cortical Oval/round

No soft tissue mass

Sclerotic margin

Fibrous cortical defect
Fibrous cortical defect and Non ossifying fibroma

- Non ossifying fibroma:
  
  i. (≥3 cm) are larger

  ii. Eccentric, intramedullary lesions that abut the cortex; they have a typical, superficial, scalloping pattern in the adjacent cortex

  iii. Might be symptomatic
Intra-medullary
No soft tissue mass

Non ossifying Fibroma
Fibrous cortical defect and Non ossifying fibroma

• Presentation:

  i. Incidental finding

  ii. Pain

  iii. Pathological fracture; larger lesions
Fibrous cortical defect and Non ossifying fibroma

- Histology: both have same histological appearance
  - Spindle-shaped fibroblasts, oriented in a cartwheel or storiform whorled pattern,
  - Scattered giant cells (osteoclast-like multinucleated cells),
  - Foam (Lipid laden macrophages) cells, and
  - Small amounts of collagen.
- No cellular atypia.
Fibrous cortical defect and Non ossifying fibroma

- Treatment:
- Conservative: regress spontaneously
- Large lesion: might fracture, requiring fixation + curettage.
Fibrous Dysplasia

- Tumour-like
- Benign
- Localised developmental arrest
- All bone components present
- Lack of differentiation
Fibrous Dysplasia

- Woven bone with fibrous tissue
- Lack of lamellar bone
- Gene mutation

- Osteoblasts produce fibrous tissue in the bone marrow instead of normal bone

- Results in: fragile bones and bony deformity
Fibrous Dysplasia

- Site:
  - Long bones, jaw, maxilla, ribs
  - Single bone: monostotic 70%
  - Multiple bones: polystotic. Associated with endocrine disorders 30%
Fibrous Dysplasia

- Presentation:
  - Asymptomatic in single bone involvement
  - Pain
  - Deformity e.g. in facial bones
  - Fractures

- X-ray:
Fibrous Dysplasia

Ground-glass lesion

Expansile

Thin cortex
Subendosteal scalloping

Fibrous Dysplasia
Fibrous Dysplasia

- Histology:
  - Curved trabecular bone, fishhook shape
  - Fibrous tissue in between, variable cellularity
  - No lamellar bone
Treatment

- Conservative
- Curettage
- Excision
- ORIF for fractures
- More?
- Rare risk of sarcoma e.g. osteosarcoma
Fibrosarcoma

- Malignant
- Mesenchymal: malignant fibroblasts
- Metaphyseal, 50% femur and tibia
- Primary: de novo lesion
- Secondary: pre-existing lesions
  - Chronic osteomyelitis
  - Paget’s
  - Bone infarction
  - Fibrous dysplasia
Fibrosarcoma

- Presentation:
  - Pain
  - Swelling
  - Fracture

- X-ray:
  Radiolucent, poorly defined, destructive
  +/- soft tissue mass
Fibrosarcoma

- **Histology:**
  - Malignant fibroblasts
  - Variable cellularity + atypia from well to poorly differentiated
  - Mitotic rate increased in high grade tumours
Moderate cellular atypia
Treatment

- Surgical excision
- Reconstruction
- Recurrence 60% following surgery

- Adjunctive chemo- and radiotherapy
- Recurrence 25%
Adamantinoma

- Rare tumour
- Malignant, slow growing
- Cell of origin: epithelial plus fibrous background
- Biphasic?
- Site: long bones, common in Tibia 80%
  Diaphysis and metaphysis
Adamantinoma

- Peculiar relation to:
  - Osteofibrous dysplasia (ossifying fibroma)

- Similar to fibrous dysplasia, but the presence of lamellar bone and osteoblasts
Woven and lamellar bone
Adamantinoma

- Presentation:
  - Pain
  - Swelling
  - Deformity: bowed tibia
  - Fracture
Adamantinoma

- Radiology: non specific
  - Radiolucent
  - Expansile
  - Sclerotic rim
  - +/- Local spread
Adamantinoma

Sclerotic Rim

Expansile Radiolucent
Adamantinoma

- Histology: different patterns

- Epithelial cells in basaloid, squamous, spindle or tubular patterns.

- Palisading at the periphery

- Pseudovascular pattern

- Background fibrous tissue: bland spindle fibroblasts
Fibroblasts

Spindle epithelial cells
Fibroblasts

Pseudovascular pattern
Treatment

- Excision: risk of recurrence
- Wide local excision
- Amputation?
- Other? Radio-resistant and chemo-resistant