Synovial Diseases in Children

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Disclosures

• Pediatrics Lead Author for Amirsys-Elsevier
  – Royalties/Fees

• Lecture heavily weighted for MRI
Overview

• Anatomy
• Imaging: Techniques and value
• Specific diseases
  – Etiology
  – Distribution of joints
  – Acute & chronic manifestations
  – Useful imaging clues
Anatomy: Types of Joints

- Synarthroses (immovable)
- Amphiarthrosis (slightly movable)
- Diarthrosis (freely movable)
Anatomy: Diarthrodial Joints

- Cartilage
- Capsule
  - Ligaments
  - Fibrous tissue
- Synovium
  - Lines capsule, not cartilage
  - Invests intra-articular ligaments & tendons
Anatomy: Synovium

- Two layers
  - Intima
    - Synovial lining cells (synovioocytes, SLCs)
    - Discontinuous layer 1-3 cells thick
    - Incomplete basement membrane
  - Subintima
    - Loose connective tissue
    - Merges with fibrous capsule
Anatomy: Synovium

• Purposes
  – Fluid formation
  – Nutrition of cartilage
  – Movement
Synovium also found...

- Bursae
- Tendon sheaths
- Synovial cysts
- *NOT* synovial sarcoma
Joint pathophysiology

- Variety of processes may incite synovitis
- Single vs. recurrent event
  → destructive cycle
- Long term effects
  - Bone
  - Articular cartilage
  - Growing epiphysis
Imaging: Why?

- Establish diagnosis
- Localize/define extent of involvement
  - Within single joint
  - Tendon sheath/bursa
  - Additional unsuspected joints
- Pick up complications early
- Optimize management
General imaging: Radiographs

- **Pros**
  - Baseline
  - Exclude other etiologies
  - Cheap, quick

- **Cons**
  - No early cartilage damage
  - No synovial detail
General imaging: Radiographs

• Secondary findings
  – Synovial hypertrophy
    • Effusion/synovial complex
    • Osteoporosis
    • Epiphyseal changes
  – Osteocartilaginous damage
    • Joint space loss
    • Erosions
    • Ankylosis
General imaging: Ultrasound

• Pros
  – No radiation/sedation
  – Readily available
  – Easy contralateral comparison
  – Great for effusions, synovitis
  – May demonstrate osteocartilaginous changes

• Cons
  – Operator dependent
  – Limited window
General imaging: MRI

• Pros
  – Most sensitive, specific method for all tissues
  – Entire joint
  – Early changes (even asymptomatic)
  – Quantitative assessments

• Cons
  – Sedation (age dependent)
  – IV contrast
  – Comparison not readily available without planning
  – Time intensive
  – Scheduling
  – Cost
Specific Diseases
Differential:
Intra-articular “loose” bodies

- Tiny, numerous, no $\text{Ca}^{2+}$
  - Rice bodies/necrotic synovium
    - JIA
    - Granulomatous diseases
    - Other?
  - Chondromatosis

- Larger, single, +/- $\text{Ca}^{2+}$
  - Osteochondral fragment
    - OCD
    - Acute trauma
  - Retained foreign body
Differential: Intra-articular “loose” bodies

- Tiny, numerous, no $\text{Ca}^{2+}$
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    - Granulomatous diseases
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    - OCD
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  - Retained foreign body

Is it really loose vs. attached?
Juvenile Idiopathic Arthritis

- Most common chronic pediatric arthropathy

- 16-150/100,000 children worldwide

- Most common 1-3YO, less 8-10YO
Juvenile Idiopathic Arthritis

- Arthritis of unknown etiology
  - Onset prior to 16YO
  - Lasts at least 6 weeks
- 9-83% persistent disease into adulthood
- Mortality < 0.3%
JIA: ILAR Classification

- Systemic arthritis (10-20%)
- Oligoarthritis (50-60%)
- Polyarthritis (Rh factor negative) (>30-35%)
- Polyarthritis (Rh factor positive)
- Psoriatic arthritis (<15%)
- Enthesis-related arthritis (<7%)
- Undifferentiated arthritis
Differential:
Distorted epiphyses

- Synovitis
- Prior infection
- Syndromic
- Gorham disease
- AVN
JIA
JIA: Imaging

- Radiographs
  - Periarticular soft tissue swelling, effusion
  - Periarticular osteoporosis
  - Accelerated maturation of secondary ossification centers
  - Periosteal reaction
  - Joint space loss, erosions
  - Subluxation, dislocation (ligamentous laxity)
  - Ankylosis (wrist, c-spine), degeneration
JIA: Imaging

• Radiographs—Specific joints
  – Knee: Widened intercondylar notch, flat patella
  – Phalanges: Boutonniere, Swan Neck, flexion
  – Hip: Protrusio acetabuli, osteonecrosis
  – Spine: Atlanto-axial instability, fusion of posterior elements, hypoplastic vertebral bodies
JIA: Imaging

- **MRI**
  - Fat pad edema
  - Prominent lymph nodes
  - Effusion, synovial thickening
  - Cartilage wear, bony edema, erosions
  - Enthesitis
  - Tenosynovitis, tendon rupture
  - Synovial cyst (popliteal or “Baker’s” cyst)
- **Specific joints:**
  - Knee: Hypoplastic menisci, cruciate ligament atrophy
Grading System of Knee: JAMRIS

- Synovial hypertrophy
- Bone marrow changes
- Cartilage lesions
- Bone erosions
### Juvenile Arthritis MRI Scoring (JAMRIS) system for the knee

<table>
<thead>
<tr>
<th>Synovial hypertrophy score (maximal synovial thickness)</th>
<th>Location</th>
<th>0-2mm</th>
<th>≥2-4mm</th>
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<td>Cruciate ligaments</td>
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<td>Medial posterior condyle</td>
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<tr>
<th>Bone marrow change score (involvement of bone volume)</th>
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<th>Cartilage lesion score (involvement of cartilage surface area)</th>
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OMERACT 11

• Special interest group on MRI in JIA
• Further develop MRI as outcomes marker
• Established goals include
  – Age-based MRI atlas of normal joint development
  – Standard protocols
  – Clinically-validated imaging grading scales
    • Knee, ankle (large)
    • Wrist, hand (small)
    • TMJ
OMERACT 12

• JIA SIG plus Health-e-Child Radiology group
  – Knee and wrist

• Agreements on protocols and scoring
  – Terminology/Definitions
  – Locations
  – Scales
JIA MRI Protocol

- T1 no FS
- T2 with FS
- Pre/post T1 with FS (subtraction)
  - Immediate, dynamic vs. 5/10 minutes
- Others?
  - DWI/ADC
  - 3D imaging
    - GRE or PD (cartilage)
JIA Quantitative Imaging

- MRI
  - Synovial volumes
  - Dynamic enhancement
  - BOLD
  - Cartilage mapping (T2, T1rho, dGEMRIC)

- US
  - Hyperemia
  - Cartilage/bone erosions
JIA TMJ

- Mild enhancement, small amount of fluid may be normal
- Surrounding soft tissue enhancement abnormal
  - Other than normal posterior venous plexus
- Active disease
  - Signal to noise ratios pre/post
  - Dynamic enhancement
  - Condylar edema
Whole Body Screening

- MRI
- PET
Ultrasound Features

- Non-compressible hypoechoic synovium
- Compressible hypo/anechoic effusion
- Hypo/anechoic tissue in tendon sheath
- Hypoechoic, thickened bony insertions of
  - Tendons
  - Ligaments
  - Capsule
- Cartilage thickness
- Bone erosions
- Doppler
JIA Therapies

- NSAIDs
- Steroids (systemic or local injections)
- DMARD
  - Methotrexate, sulfasalazine, leflunomide
- Biologics (monoclonal antibodies, soluble receptors)
  - TNF-α inhibitors
  - IL-1, IL-6
- Stem Cell Transplant
Differential:
Intra-/peri-articular calcifications

- Synovial chondromatosis
- Synovial venous malformation
- Neuropathic joint
- Synovial sarcoma
- Injection granulomas
- Heterotopic ossification
- Osteosarcoma
- Dermatomyositis
- Tumoral calcinosis

Smaller, Intraarticular
Larger, Periarticular
Synovial Chondromatosis

- Benign subsynovial neoplasia
  - Hyaline cartilage nodules
  - May mineralize (60%) and/or become loose

- Usually monoarticular process
  - Knee (50%) > elbow, hip, shoulder
  - Bilateral knee involvement < 10%

- M:F 2:1

- Most common 3rd-5th decades

- Rare malignant transformation
Synovial Chondromatosis: Imaging

- Radiographs:
  - Joint fullness
    - With/without calcified nodules
    - Nodules typically uniform in size
  - May have erosions (esp. tight capsule joints)
  - Late secondary DJD
Synovial Chondromatosis: Imaging

- **MRI:**
  - Appearance depends on nodule content
    - Cartilaginous
      - Intermediate T1/high T2
      - Low signal intensity septa enhances
    - Mixed cartilage/calcification
      - Small foci of low T1/T2 signal within nodules
    - Ossified with fatty marrow:
      - Follows fat signal (high T1, low w/ fat suppression)
Neuropathic joint
Dermatomyositis
Tumoral calcinosis
Tumoral calcinosis
Osteosarcoma
Differential:
Intra-articular hemosiderin

- Hemophilia (or other systemic bleeding disorder)
- Pigmented villonodular synovitis
- Synovial venous malformation
PVNS

• Classification
  – Intra-articular diffuse
    • Knee 80%, hip, ankle, shoulder, elbow
  – Intra-articular localized
  – Extra-articular (GCTTS)

• M=F

• Most commonly 3rd-4th decade

• Benign neoplastic process
PVNS

- Radiographs:
  - Soft tissue mass
  - Late demineralization and joint space loss
  - Erosions
    - Prominent in joints with tight capsule (hip & ankle)
    - Prefer bare areas (rather than subchondral)
  - Periosteal reaction, calcifications <10%
PVNS

• MRI
  – Low signal
    • Secondary to hemosiderin deposition
    • GRE (T2*) accentuates susceptibility
  – May be components of variable T1/T2 signal due to:
    • Mix of hemosiderin/fat/fibrovascular elements
    • Recent hemorrhagic effusion (subacute blood products)
    • Inflamed synovium
PVNS
PVNS (GCTTS)
Differential:
Focal articular/tendon sheath mass

- Clot
- Foreign body granuloma
- PVNS
- Venolymphatic malformation
- Pannus
- Synovial lipoma
- Synovial sarcoma
- Fibroma of tendon sheath
Synovial sarcoma
Differential:
Fat-containing mass

- Synovial lipoma/lipoma arborescens
- Lipoblastoma
- Fibrous hamartoma
- Involuted hemangioma
- Venous malformation
Lipoma Arborescens/Synovial Lipoma

- **Monoarticular**
  - Usually knee, suprapatellar
  - Rarely bilateral

- **Subsynovial tissue replaced by fat cells, inciting synovial proliferation**

- **Metaplasia (not neoplasia)**
Lipoma Arborescens/Synovial Lipoma

- Usually chronic
  - Gradual swelling
  - Painless
  - Recurrent effusions
  - Rarely restricted movement
  - Most common 5<sup>th</sup>-7<sup>th</sup> decade
  - Can be secondary to JIA, trauma
Lipoma Arborescens: Imaging

- **Radiographs**
  - Radiolucent foci

- **Ultrasound**
  - Echogenic mass
  - May be frond-like and mobile

- **MRI**
  - Follows fat signal on all sequences
  - Nonfatty portions and synovium may enhance
  - May be accompanied by effusion
  - Chemical shift artifact at fat-fluid interface
Lipoma Arborescens
Case 5
Septic Arthritis

- Often < 10YO
- Knee, hip > elbow, ankle, shoulder
- Sources:
  - Adjacent osteomyelitis
    - Is effusion a septic joint or sterile reactive fluid?
    - Is marrow signal reactive edema vs. osteomyelitis?
  - Hematogenous seeding
  - Direct penetration
Septic Arthritis

- **Joint injury by**
  - Lytic enzymes
  - Increased pressure

- **Imaging**
  - Initially by radiographs, US
  - Correlated with clinical & laboratory parameters (Kocher)
  - Tapped, taken to OR, IV antibiotics
  - MR if no improvement in 48 hours for osteomyelitis

- **Other infections**
  - Granulomatous disease
  - Lyme disease
Septic arthritis
Remote septic arthritis
3 months later...
? Infection

● Toxic Synovitis
  ● Hip (bilateral 25%)
  ● Limp, pain, stiffness
  ● Self-limiting, 3-10 days
  ● 3-10YO

● Imaging
  – Ultrasound cannot distinguish
  – MRI suggests septic joint if periarticular soft tissue or bone marrow signal abnormalities
Toxic synovitis
Hemophilia

• X-linked recessive diseases
  – A: factor VIII deficiency
  – B: factor IX deficiency

• Manifest by spontaneous or traumatic hemorrhage:
  – Intracranial
  – Intra-abdominal, retroperitoneal
  – Musculoskeletal
    • Joints (up to 85% of bleeding episodes)
    • Muscle
    • Bone
    • Fascia
Hemophilic arthropathy

- Synovial bleeding into joint
- Originates at subsynovial vascular plexus
- Chronic deposition of hemosiderin
- Synovial hypertrophy, inflammation, hyperemia
  - Osteoporosis
  - Epiphyseal overgrowth
  - Physeal fusion
Hemophilic arthropathy

- Articular cartilage damaged by:
  - Intra-articular blood products
  - Enzymes breaking down hemosiderin
  - Increased intracapsular pressure

- Subchondral cysts
  - Intraosseous hemorrhage

- Fibrosis
  - Capsular fibrosis, joint contractures
Hemophilic arthropathy: Imaging

• Radiographs
  – General:
    • Increased density effusions
    • Osteoporosis, epiphyseal enlargement, physeal fusion
    • Subchondral cysts
    • Degenerative changes
    • Flexion deformities
Hemophilic arthropathy: Imaging

- Radiographs:
  - Specific joints
    - Knee: Wide intercondylar notch, elongated patella
    - Elbow: Wide trochlear notch and olecranon fossa
    - Ankle: Tibiotalar slanting, flattened talar dome
    - Hip: Protrusio acetabuli, SCFE, coxa valga
Hemophilic arthropathy: Imaging

• MRI:
  – Thickened, low signal synovium on all sequences (most pronounced on T2*)
  – Enhancement post-contrast
Case 7
Synovial Venolymphatic Malformation

- Usually monoarticular
- Knee most common
- Pain, swelling, hemarthrosis
- Focal mass vs. diffuse venous abnormality
  - Malformation may extend through soft tissues to another joint
Synovial Venolymphatic Malformation: Imaging

- **Radiographs:**
  - 50% normal
  - May have phleboliths

- **MRI:**
  - Lobulated mass or serpiginous components
  - High T2 signal
  - Low T1 phleboliths
  - May have fluid-fluid levels
  - May have T2* effect from hemosiderin
  - Variable enhancement
Venolymphatic Malformation
Other causes of synovitis

- Trauma/foreign body
- SCFE
- Infarction
- Tumors
- Leukemia
Foreign body
Chondroblastoma
Summary

- Variety of processes may incite synovitis
- Significant long term effects on intra-articular structures
- Overlapping imaging features
  - Look for specific clues
    - Loose bodies
    - Calcifications
    - Hemorrhage
    - Fat
    - Marrow abnormalities
  - *Always think about septic arthritis!*
Thank you!
References
