Rheumatology Core Content

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❖ Introduction

- Hippocrates: *On the Locations in the Human Body*, c. 350 B.C.E.: Rheuma = earlier catarrhos, humors (flowing from the brain)
- Guilliame Baillou (Ballonius): *The Book on Rheumatism and Back Pain* (1642): associated arthritis and rheumatism
- Thomas Sydenham:
  - fibromyalgia (*Hysteric Diseases*, 1681)
  - oligoarticular gout (1683)
  - febrile polyarticular arthritis of the young (rheumatic fever) (1685)
  - podagra: foot; chiagra: wrist, gonagra: knee.
- More modern spectrum of rheumatology:
  - DJD
  - Fibromyalgia
  - Inflammatory arthritis of various types (RA, gout, etc.)
  - System autoimmune diseases including those against joint synovium, cartilage, tendons, or connective tissue including vasculitis

❖ General Approach to Acute Arthritis

- oligoarticular
  - Stick a needle in it (unless classic recurrent gout, ? overlying cellulitis/infected bursitis only)
- Is it really bursitis?
  - Elbow, knee, shoulder, trochanteric, deltoid, subacromial
  - Septic Bursitis: warmth most distinguishing feature
- Gout/pseudogout
  - Well-fed men who drink plenty of port
  - How useful is serum uric acid? Useless.
  - How do you tell septic joint from gout? Only 25% of GC joints gram stain or culture +!
  - Joint fluid WBC >50-100K suggests infection
  - May have gout crystals in joints without clinical gout
  - Gout and Pseudogout x-rays? Bony erosions, bony destruction (may look like osteo; hard to tell apart)
  - Criteria
    - more than 1 attack
    - max inflammation within a day
    - mono articular

- redness
- podagra (1st MTP joint)
- culture negative
- unilateral 1st MTP joint
- unilateral subtarsal joint
- tophus
- hyperuricemia
- …

- Gout Rx:
  - colchicine IV?
  - NSAIDs?
  - Indocin?
  - Narcotics?
  - Steroids?
  - Inject joint?
  - Allopurinol?

- GC
  - vag or penile DC
  - Classic triad:
    - 50% dermatitis-arthritis syndrome with hemorrhagic pustules
    - Fleeting tenosynovitis of flexor hand tendons (immune complex inflammation)
    - Migratory hot red joints: Wrist, KNEE, ankle (immune complex, later septic)
- Non-GC septic joint

- ♦ KNEE, hip, shoulder, ankle, elbow, hand wrist (19% mortality if also RA)
- ♦ IVDA: sternoclavicular, SI (FABER test: Forced ABduction, External Rotation of hip causes pain; Patrick test: Lotus position, press down on knee; causes pain), manubriosternal joints
- ▪ "Inflammatory DJD"
- ▪ "Toxic synovitis" in kids
- ▪ IBD inflammatory arthritis

➢ polyarticular: see below

❖ **RA/JRA**

➢ Autoimmune against IgG ("RF"): deposit in synovium (don't retest; retesting ESR OK)

➢ Familial tendency, more in women

➢ ARA RA criteria: 4 of 7:

- ▪ morning stiffness*
- ▪ arthritis 3 or more joints*
- ▪ PIP or wrist swelling*
- ▪ Rheumatoid nodules
- ▪ +RF
- ▪ erosions or periarticular osteopenia on films
- ▪ (* more than 6 weeks, else likely viral)

➢ May involve arytenoids: hoarseness, dyspnea, respiratory arrest

- **May develop atlantoaxial (-dens) subluxation:**
  - **nl: <3.5 mm (<4 mm if <12 yrs), flexion view (how do you get flexion view?)**
  - **may get cord compression slowly or after minor trauma**
  - **normal signs of cord compression:**
    - **bowel/bladder**
    - **focal neuro sx or signs**
    - **Lhermitte's Sign: electric shock down back with neck flexion**
    - **If intubating someone with RA, treat like trauma patient: immobilize neck**
- **Pulmonary effusions: low glucose like TB**
- **Pericarditis common but seldom serious**
- **Sjogren's: + dry eyes (Schirmer test)**
- **Felty's: + splenomegaly and thrombocytopenia**
- **Red eye:**
  - **episcleritis? Triangular, up against corneal limbus: benign, painless, self-limiting**
  - **scleritis? Diffuse, painful, changes in vision, may rupture: consult, steroid drops**

- **Systemic Lupus Erythematosus (SLE); lupus**
  - **basics:**
    - **1:1000 women of child-bearing years**
    - **genetic predisposition (HLA), environmental (sun) and hormonal (estrogen)**
    - **triggered by hydralazine, INH, Procan/quinidine (milder) (sulfa, TCN, Macrodantin, Dilantin, PTU, lithium, Thorazine, allopurinol)**
    - **Tan criteria (any 4 of 11): malar or discoid rash, photosensitivity, oral ulcer, arthritis, serositis, renal, neuro (sx/psychosis), hematologic, + immuno tests, + ANA**
    - **Autoimmune:**
      - **ANA, LE Prep, anti-DS DNA (just anti-SS DNA/anti-histone in drug-induced), anti-Smith antigen, v complement levels**
      - **Lupus "anticoagulant":**
        - **may be seen without SLE, especially in HIV,**
        - **procoagulant**
        - **elevated PTT**
        - **anti-phospholipid/cardiolipin antibodies**

- spontaneous Abs
- premature atherosclerosis

(? early MI instead of pericarditis)

- **Clinical:**
  - fever, joint pain, rash, in child-bearing years
  - mild (skin only) to severe
    - ESRD: 50% of SLE
    - cerebritis: sz, CVA, psychosis, migraines, peripheral neuropathy; 10% mortality, but 75% recover; can also be bacterial meningitis
      - transverse myelitis: "an inflammatory process involving both gray and white matter of spinal cord."
    - polyserositis:
      - pericarditis (30%): tamponade rare, responds to steroids; pleurisy; abdominal pain
    - pancreatitis (from SLE or steroids)
    - Anemia, thrombocytopenia
  - PIP and MCP joints (like rheumatoid), tendonitis
  - Butterfly rash, discoid lupus (only 10% of discoid will have SLE); 25% of SLE will have rash
- **Treatment:**
  - general:

- avoid precipitants (sun, estrogen, stress)
- NSAIDs (if kidney OK), avoid estrogen/UV/sleep deprivation
- steroids (Solu-Cortef 100 mg for routine, big doses for cerebritis)
- antimalarials: chloroquine, etc. (may cause blindness)
- immunosuppressants (azathioprine=Imuran cyclophosphamide=Cytoxan)

- ED:
  - cerebritis/sz: work up for other causes, then one gram SoluMedrol, admit
  - nephritis: check urine microscopic, check for elevated PTT and check for renal vein thrombosis (CT?)

Vasculitis

Wegener's Granulomatosis ("Lethal Midline Granulomatosis")

- Sinusitis, nasal ulcers, pulmonary nodules, infiltrates, bronchospasm, hemoptysis

Temporal Arteritis ("Giant-Cell Arteritis")

- why may be hidden timebomb:
  - may cause sudden blindness 3-4 months after onset

- may cause aortic dissection
- when to treat: if suspect enough to get bx
- Treat with prednisone 60/day, still have a week to get a temporal artery biopsy
- when to suspect
  - older women
  - temporal aa tenderness
  - ESR ~60
  - jaw, tongue or upper extremity claudication
  - locally tender scalp in a blood vessel distribution

➢ **Younger women: Takayasu's Arteritis ("pulseless disease")**

- large vessel and coronary ischemia
- both associated with:

➢ **Polymyalgia Rheumatica (PMR)**

- Inflammatory condition of proximal limb girdle muscles
- Normal CPK, elevated ESR
- dramatic response to low-dose steroids (2.5-5 mg/day prednisone)

➢ **Kawasaki's**

- arteritis of medium to small vessels
- likely caused by retrovirus
- gamma globulin combined with high dose aspirin is somewhat

effective in preventing cardiovascular complications.

- mostly in kids
- may cause coronary disease
- mnemonic CRASH:
  - C - conjunctivitis
  - R - rash: red lips, palms and soles, desquamation of the fingertips
  - A - adenopathy
  - S - strawberry tongue
  - H - high fever

➢ **Polyarteritis Nodosa:**

- medium-sized arteritis including coronaries

➢ **Scleroderma (systemic sclerosis)**

- prunified skin
- big killer: dehydration leading to hypertensive renal-failure crisis (controlled by ACE inhibitors now)
- Benign free air in belly, ignore
- + ANA

➢ **Polymyositis, Dermatomyositis**

- Proximal muscle involvement; Trouble getting out of chair, up steps, ^ CPK
- Often paraneoplastic
- Treat with big doses of steroids
- May get in respiratory problems but not be able to show costal

retractions, look for nasal flaring instead

- Check bedside PFTs as with Myasthenia (NIF, TV): if <30% predicted, plan to intubate
- May also get fulminant pulmonary fibrosis (as well as pneumonias from immunosuppression)

➢ **Rheumatic Fever: migratory polyarthritis**

- Differential of migratory arthritis: SBE, HSP, Ceclor reaction (kids), sepsis, Mycoplasma, histo, coccidio, Lyme Disease
- "If supported by evidence of preceding group A strep infection, the presence of two major manifestations or of one major and two minor manifestations indicates a high probability of acute rheumatic fever."
  - ♦ Major Manifestations:
    - ◊ Carditis
    - ◊ Polyarthritis
    - ◊ Chorea
    - ◊ Erythema Marginatum
    - ◊ Subcutaneous Nodules
  - ♦ Minor Manifestations:
    - ◊ Clinical Findings: Arthralgia, Fever

- ◊ **Lab Findings: Elevated ESR or CRP**
- ♦ **Supporting Evidence of antecedent Group A Strep infection: positive rapid strep or + culture, elevated or rising strep antibody titre**

➢ ***HLA-B27 "seronegative spondyloarthropathies"*** **(never retest HLA-B27 or ask for a titer)**

- **Reiter's**
  - ♦ **Urethritis, conjunctivitis, arthritis**
  - ♦ **Association with Chlamydia, Salmonella, Shigella**
- **psoriasis**
- **ankylosing spondylitis**
  - ♦ **AS: decreased rib excursion, diminished pulmonary reserve**
  - ♦ **c-spine fracture with AS "bamboo spine": after minor trauma, usually through remains of disk space**

➢ **Fibromyalgia**

- **Inflammatory condition of proximal and spinal muscles; associated with lack of Stage IV sleep; Flexeril helps; normal CPK**
- **nine paired points that are almost invariably tender. Testing for tenderness there, and for control**

points that shouldn't be tender, helps establish the diagnosis.

- **Tender points:**
  - **1. Insertion of nuchal muscles into occiput**
  - **2. Upper trapezius, midpoint**
  - **3. Pectoralis muscle - just lateral to second costochondral junction**
  - **4. 2 cm below lateral epicondyle**
  - **5. Upper gluteal region**
  - **6. 2 cm posterior to Greater Trochanter**
  - **7. Medial knee in area of anserine bursa**
  - **8. Paraspinous, 3 cm lateral to midline at the level of the mid-scapular**
  - **9. Above the Scapula spine near the medial border**
- **Control points:**
  - **1. Middle of forehead**
  - **2. Volar aspect of Mid-forearm**
  - **3. Thumbnail**
  - **4. Muscles of anterior thigh**
  - **Benner RM: Fibrositis. In: Kelly WN, Harris ED, Rudy S, Sledge CB**

(eds): Textbook of Rheumatology, 4th ed. Philadelphia, WB Saunders, 1993.

- **Anterior Spinal Syndrome**
  - thrombosis or embolism or arteritis of the anterior spinal artery "artery of Adamkiewicz" "arteria radicularis magna" "great anastomotic artery," "great radicular artery"
- **Erythema Nodosum**
  - Women on BCPs; also Motrin, Sulfa.
  - May see with sarcoid, SLE, other vasculitis, but usually isolated
  - Treat with high-dose ASA
- ***Extra Credit***
- **Associated S/Sx**
  - psoriasis (psoriatic arthritis)
  - kidney stones (pseudogout)
  - IBD (IBD arthritis)
  - immunosuppression (septic joint)
  - rash
    - erythema migrans (Lyme)
    - erythema marginatum (rheumatic fever)
    - erythema nodosum (SLE)
    - discoid lupus (SLE)
    - keratoderma blennorrhagicum of soles (Reiter's)
    - circinate balanitis (Reiter's)

- **Relapsing polychondritis** (ears, nose and other cartilage: can cause tracheal rings to turn to mush (check flow-volume loops via formal PFTs)
- **Addisonian crisis** from long-term steroid use: if unsure, give Decadron as won't interfere with cortisol assays (also get random cortisol first, should be > 20 mcg/cc)