Rheumatology Board Review

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Overview of Today’s Review
(At least that which can be covered in two hours)

- Inflammatory Arthropathies and Autoimmune Diseases
  - Rheumatoid Arthritis
  - Systemic Lupus Erythematosus
  - Seronegative Spondyloarthritis
- Non-Inflammatory Arthritis
- Crystalline-Induced Arthritis
  - Gout
  - CPPD
Case I

- A 35 year old woman reports 6 weeks of morning stiffness in bilateral wrists, meta-carpal phalangeal joints, and feet that lasts for 2 hours each morning and is worse with inactivity. In evaluating the arthritis, her primary care doctor notes that she has swelling in her hands and wrists, a negative Rheumatoid Factor, and X-rays of her hands that demonstrate peri-articular osteopenia but no joint space narrowing or erosions.

Case I

Which of the following statements regarding this patient’s symptoms is correct?

A. Her age of onset is too young for RA
B. Her morning and gelling symptoms are most consistent with an inflammatory arthritis
C. The radiologic findings show no joint deformities, making an RA dx. unlikely
D. She can’t have RA if her rheumatoid factor is negative
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Rheumatoid Arthritis

- RA is 2\textsuperscript{nd} most common form of chronic arthritis (behind osteoarthritis)
- Has a prevalence of 1\% in US adults
- Gender incidence 3:1 women:men
  - onset 4\textsuperscript{th}-5\textsuperscript{th} decade
- Marginal joint erosions distinguish RA from most other forms of arthritis

Rheumatoid Arthritis

Clinical Manifestations

- Musculoskeletal
  - Bilateral, symmetric, polyarthritis (> 5 joints) often affecting small joints of hands and feet (Wrists, MCPs, PIPs, MTPs, not DIPs)
  - Morning stiffness and gelling are common
  - 90\% insidious onset over weeks to months
- Systemic signs/symptoms
  - Fatigue common
  - Significant weight loss (10\%)
  - Low grade fevers (<38.3)
Rheumatoid Arthritis
Extra-articular Manifestations

- Rheumatoid Nodules
  - More prevalent with RF+ patients
  - Most commonly develop on extensor surfaces (arms, fingers)

- Eye
  - Keratoconjunctivitis sicca - dry eyes
  - Scleritis - painful, injected
  - Scleral ulcers

- Pulmonary
  - Effusions
  - Interstitial lung disease
  - Nodules

Rheumatoid Arthritis
Laboratory testing

- Common labs
  - Anemia of chronic inflammation
  - Thrombocytosis
  - Elevated ESR, CRP

- Rheumatoid factor
  - Negative in up to 20% of RA patients (up to 50% negative at time of diagnosis)
  - Positivity increases with disease duration

- Anti-cyclic citrullinated peptide (CCP) has 70-80% sensitivity and 90+% specificity
Imaging

Rheumatoid Arthritis
• Marginal erosions (mid-late)
• Peri-articular osteoporosis (early-onward)

Osteo-Arthritis
• Sclerosis
• Osteophytes
• Joint space narrowing

Case I, question #2

The patient receives prednisone, methotrexate and is started on infliximab since she does not want to give herself shots. Within 2 weeks her morning stiffness lasts less than 10 minutes, and the swelling in her fingers is markedly reduced. She feels great. 6 weeks following initiation of therapy she develops a low grade temperature accompanied by a mildly productive cough. After a short course with a macrolide, the cough continued so her primary provider orders a chest x-ray which revealed a bi-basilar faint, diffuse infiltrate.
**Case I, Question #2**

Which of the following is the next best step?

A. Admit to the hospital to initiate parenteral antibiotics in an immunosuppressed patient.

B. Admit to the hospital to rule out an atypical presentation of pulmonary tuberculosis.

C. Stop the methotrexate because of presumptive diagnosis of an hypersensitivity syndrome.

D. Increase the methotrexate, because of presumptive diagnosis of rheumatoid lung.

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![Bar chart](chart.png)
Commonly used medications
Monitoring and toxicity

- **Glucocorticoids (e.g. prednisone)**
  - Dosed daily, maintenance 5 - 15 mg/day
  - Observe rapid (3-7 day) response to inflammatory symptoms
  - Toxicity
    - Hyperglycemia
    - Cushingoid changes - truncal obesity, “moon facies”
    - Adrenal insufficiency - must consider during acute stress
    - Osteoporosis – initiate bisphosphonates, Ca, Vitamin D Rx

- **Methotrexate (cornerstone of RA DMARD therapy)**
  - Early initiation of DMARD therapy now standard of care
  - Clinical Response sub-acute – 3-6 weeks
  - Dosed one day per week, 7.5mg-20mg total
  - Toxicity
    - Hepatotoxicity - transaminitis
    - Myelosuppression - especially lymphocytes
    - Hypersensitivity pneumonitis and interstitial lung disease
    - Mucosal irritation – Use concurrent folate supplements
  - Monitor CBC, LFTs q 4 weeks until achieve stable dose then q 4-8 weeks as long as take medication
Commonly used medications
Monitoring and toxicity

- **Hydroxychloroquine**
  - Dosed daily
  - Clinical effect 2-3 months (mild to mod)
  - Toxicity – retinal damage (generally after long use)
  - Monitor – baseline retinal exam then q 6-12 months

Commonly used medications
Monitoring and toxicity

- **Anti-Tumor Necrosis Factor Agents**
  - Clinically – **New paradigm initiate early on in RA**
  - **Infliximab**
    - Dosing - IV infusion on day 0, 2 and 6 weeks, then q 8 weeks
    - Toxicity
      - Infusion reaction within 1-2 hours of treatment
      - Reports of serum sickness
Commonly used medications
Monitoring and toxicity

- Anti-Tumor Necrosis Factor Agents
  - Etanercept
    - Dosing – SQ injection, 25 mg biweekly; 50 mg 1x/week
    - Toxicity – injection site reaction
  - Adalimumab
    - Dosing – SQ injection 40mg every other week
    - Toxicity – injection site reaction

- Class Toxicity
  - Reactivation of latent tuberculosis
    - 50% of patients within 12 weeks of initiating treatment
    - 50% of reactivation manifests with extra-pulmonary TB
    - Screen with PPD, and chest xray
    - treat latent infection with INH prior to RX
  - Increased rate of soft tissue infections
  - Contraindicated in Class III-IV CHF
  - Question of increased risk of malignancy
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Case II

36 year old man complains of 3 weeks of a painful left knee and right ankle, dysuria, and new onset “severe athlete’s foot”. He had no previous illnesses. He endorses having had food poisoning for which he was treated successfully 6 weeks ago. On exam he has active synovitis at his left knee and right ankle, with heel spur pain and plantar fasciitis on his right foot. Pustular scaling plaques with onycholysis were seen on his bilateral feet. Analysis of synovial fluid from his left knee revealed 14,000 WBC (91% PMNs) with negative cultures and gram’s stain. Urinalysis and urethral swab studies are negative for STDs.
Pustular scaling plaques

Case II

Which of the following treatment approaches is least appropriate for this patient?

A. Prescribe diclofenac 75 mg TID until symptoms improve
B. Intra-articular injection of triamcinolone into the knee
C. Sulfasalazine therapy for persistent symptoms
D. Empiric ciprofloxacin therapy
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The Family of Seronegative Spondyloarthropathies

- Ankylosing Spondylitis
- Psoriatic Arthritis
- Reactive Arthritis
- Enteropathic Arthritis (associated with Inflammatory Bowel Disease)

Seronegative Spondyloarthropathies

- General Characteristics
  - ANA, RF negative (seronegative)
  - Inflammatory arthritis of axial skeleton (SI joints and spine)
    - Sacro-iliitis
    - Syndesmophytes; bamboo spine
  - Oligoarticular, asymmetric peripheral arthritis
  - Enthesitis
    - Inflammatory pain at point of tendon or joint capsule insertion on the bone
      - E.g. Achilles tendon or plantar fascia insertion at heel pain
A Word About HLA-B27

- Ankylosing Spondylitis  Reactive Arthritis  Psoriatic Arthritis  IBD Arthritis
  90%  75%  50%  40%

- Prevalence of B27 is high in Caucasians
- 95% of patients with B27 never develop spondyloarthritis
- B27 is not useful diagnostic test in most situations
- Presence of HLA-B27 associated with more axial disease, uveitis

Reactive Arthritis

- Arthritis is a reaction to an infectious exposure that occurred 1-4 weeks before
  - Venereal Exposure
    - Chlamydia trachomatis
    - Male Patient may report an antecedent urethritis
    - Female patient often asymptomatic
  - Invasive enteropathic process
    - Salmonella, Shigella, Campylobacter, Yersinia
    - Patients report a prior episode of bloody or severe diarrhea
- Up to 50% may not identify an exposure
Classic Triad

- Described by the Prussian physician Hans Reiter in 1916
  - Arthritis
  - Conjunctivitis
  - Non-gonococcal Urethritis
- Triad is present MINORITY (< 10%) of Reactive arthritis cases

Reactive Arthritis
Clinical

- Disproportionately men, 3rd-5th decade
- Disproportionately affects lower extremities
  - Plantar fasciitis with heel spur pain on exam
  - Achilles tendonitis
  - Asymmetric knee or ankle arthritis
- Muco-cutaneous lesions
  - Painless oral ulcers
  - Keratoderma blenorrhagica (difficult to differentiate from pustular psoriasis)
  - Circinate balanitis
- Axial Arthritis
  - Asymmetric Sacro-iliitis
  - Asymmetric, bulky vertebral body osteophytes
Reactive Arthritis: Treatment

- NSAIDs of symptomatic benefit
- Sulfasalazine of some modest benefit for chronic peripheral arthritis
- Methotrexate likely of some benefit
- Anti-TNF medications for spinal and peripheral arthritis
- If chronic, institute aggressive physical therapy to prevent morbidity

Case III

32 yo man with a 6 year history of persistent low back and buttock pain that often awakens him from sleep, now presents with acute onset of recurrent severe blurred vision and photophobia, in his bloodshot left eye.
Case III

Which of the following is the most likely diagnosis?
A. Scleritis
B. Bacterial conjunctivitis
C. Anterior uveitis
D. Acute angle closure glaucoma

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Ankylosing Spondylitis: Quick Facts

- **Demographic**
  - Men:Women 3:1
  - Most common onset during 3rd-5th decades

- **Clinical**
  - Axial Skeleton
    - Squared off vertebral bodies
    - Shiny corners of vertebral bodies
    - Syndesmophyes - symmetric fine, bony growths that bridge vertebral bodies
    - **Bilateral sacro-iliac joint**
  - Peripheral arthritis
    - Oligoarticular, affecting hip, ankles, knees
Ankylosing Spondylitis: Quick Facts

- **Clinical**
  - Extra-articular manifestations
    - Anterior uveitis - photophobia, often unilateral
    - Pulmonary fibrosis - classically apical
    - Aortitis, aortic regurgitation
Psoriatic Arthritis: Quick Facts

Demographic
- Male:Female - 1:1
- 10% prevalence in psoriatic patients

Cutaneous
- 15% of patients who develop arthritis, joint manifestations may precede the skin changes
- More commonly skin changes precede arthritis
- Examine hairline, umbilicus, gluteal folds for signs of skin disease
- Nailbed changes – pitting, onycholysis
- Severe Psoriasis +/- arthritis consider HIV

Peripheral Arthritis
- Oligoarticular, monoarticular (66% of patients)
- Polyarticular (pseudo-rheumatoid presentation)
  - Affects the DIPs commonly

Axial Arthritis
- Early sacroiliitis usually unilateral
- Bulky asymmetric vertebral body osteophytes

Dactylitis – Sausage digits
Psoriatic arthritis – Pencil in cup

Psoriatic Arthritis: Quick Facts

- Medications
  - NSAIDs - given to all who tolerate
  - Sulfasalazine - some benefit to peripheral arthritis
  - Methotrexate - improves cutaneous and arthritis
  - Anti-TNF - excellent relief of skin and arthritis
  - Avoid systemic corticosteroids - tapering can trigger pustular psoriasis flare
A 71 year old woman presents to you in urgent care complaining of a 6 month history of progressive shortness of breath with ambulation. Since moving to San Francisco (from Tulsa) one year ago, she reports worsening of “numbness” in her fingers and toes. She denies chest pain, cough or fevers. You order a chest X-ray which was interpreted as normal and a 12 lead ECG which reveals LVH strain pattern, right atrial enlargement with normal R wave progression and no Q waves.

On physical exam she has mildly swollen fingers, peri-ungal erythema and no synovitis. She had multiple erythemaous, blanching, non-palpable lesions scattered across her face and chest.

PMH
Hypertension
Severe GERD
Post-herpetic truncal neuralgia
Case IV

Which of the following is the least appropriate next step in the workup?

A. Left heart catheterization
B. Echocardiogram to assess Pulmonary artery pressures
C. High resolution CT of the chest
D. Pulmonary function tests including measurement of diffusion capacity
Scleroderma - Key Points

- **Serology**
  - ANA >95% for both forms
  - Anti-centromere pattern 20-40% (specific for CREST)
  - Anti-SCL-70 pattern 20-40% (specific for Progressive scleroderma)

- **Pulmonary Disease**
  - Isolated pulmonary hypertension seen more often in patients with CREST (common cause of death)
  - Interstitial lung disease, without pulmonary HTN more common in Diffuse scleroderma

- **Overlap Syndromes**
  - Scleroderma with elements of polymyositis, dermatomyositis

Limited Scleroderma (CREST): Quick Facts

- **Background**
  - Women:men 4:1
  - Most common onset during 3rd-5th decades
- **CREST (Criteria - diagnosis 3 of 5)**
  - Calcinosis - subcutaneous deposits, fingers extensor surfaces
  - Raynaud’s
    - Most common first sign (>90%)
    - May involve fingers and toes,
  - Esophageal
    - GERD often severe, provider does not appreciate association
    - Dysphagia (food sticks in the mid-esophagus)
  - Sclerodactyly
    - Swelling of fingers
    - Skin thickened distal to the MCPs
  - Telangectasia
    - Squared off appearance on face, palms, mucosal surfaces
Progressive Systemic Sclerosis: Quick Facts

- **Demographic**
  - Same as CREST

- **Clinical (Different than CREST)**
  - **Cutaneous**
    - Early disease may see scleredema of fingers, hands
    - Sclerosis extends to dorsum of hands, forearms
    - May involve the face and trunk
  - **Renal Crisis**
    - Hypertensive emergency picture
    - Most common during scleredema phase
    - Associated with prednisone use
    - ACE-I’s are life saving therapy

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**Case IV**

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B. **Echocardiogram to assess Pulmonary artery pressures**
C. **High resolution CT of the chest**
D. **Pulmonary function tests including measurement of diffusion capacity**
Case V

55 yo woman 2 week history of “funny red bumps” on her legs reports 1 week of increasing shortness of breath with mild cough. Today notes blood tinge to her sputum. On exam her BP is 185/90. No arthritis, oral lesions, normal neurologic exam. She had palpable purpura.

Lab studies revealed serum creatinine 2.7 elevated from her baseline 0.9, myelo-peroxidase antibody positive, proteinase-3 antibody negative

Case V

Which of the following is the most likely diagnosis?

A. Cryoglobulinemia
B. Wegener’s granulomatosis
C. Sub-acute bacterial endocarditis
D. Microscopic polyangiitis (MPA)
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Gross framework to organize vasculitis

- Multiple organ involvement
- Large vessels
  - Limb ischemia, stroke
- Medium vessel
  - Organ ischemia, neuritis, skin ulcers
- Small vessel
  - Capillaritis - Diffuse alveolar hemorrhage, palpable purpura, GN
Small vessel vasculitis framework
ANCA Associated Vasculitis

- Microscopic polyangiitis
  - Clinical
    - Skin - palpable purpura, ulcers
    - Lungs - Diffuse alveolar hemorrhage
    - Kidneys - glomerulonephritis
    - Neuro - mononeuritis multiplex, > 60%
  - p-ANCA - anti-myeloperoxidase abs
    - MPA - 75% sensitive
    - Also abnormal with Churg-Strauss

ANCA Associated Vasculitis: Quick Facts

- Wegener’s Granulomatosis
  - Clinical
    - Sinus - chronic sx, necrotizing disease
    - Lungs - nodules, cavities, alveolar hemorrhage
    - Kidneys - pauci-immune glomerulonephritis; normal complements
  - c-ANCA - anti-proteinase-3 abs are highly sensitive

- Treatment of both: Cytotoxic therapies and corticosteroids
ABIM Board Review
2008

Rheumatology Section
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Overview of Today’s Review:
The Field of Rheumatology: That giant black hole
Overview of Today’s Review:

- Vasculitis
- Scleroderma
- Inflammatory Myopathies

Important topics covered in other talks
- Viral, spirochetal, and septic arthritis (covered elsewhere in course)
- Metabolic bone disease and osteoporosis (covered elsewhere in course)

Case #1

A 66 yo Caucasian woman with ischemic heart disease and congestive heart failure notes development of arthritis in her wrists, knees and ankles that is worse in the morning and associated with occasional chest pain of a possible pleuritic quality. She denies skin lesions or oral ulcers. She has no family history of rheumatic conditions. Past medical history also includes hypertension and diabetes. Her medications include: aspirin, carvedilol, hydralazine, benazepril, and metformin. Her physical examination is consistent with mild synovitis in her hands and wrists and x-rays show small pleural effusions but are otherwise negative.
Case #1

She is found to have a negative RF and an ANA that is 1:320 in a diffuse pattern. Her chemistry panel is otherwise unremarkable and a urine dipstick in the office is only positive for 1+ proteinuria. What is the LEAST appropriate next step?

A. Check a urinalysis with micro, CBC, C3, C4
B. Check anti-ds DNA, Smith, and other ANA sub-serologies
C. Substitute another agent for hydralazine
D. Perform a kidney biopsy to rule out early lupus associated proteinuria
Case #2

She is found to have a negative RF and an ANA that is 1:320 in a diffuse pattern. Her chemistry panel is otherwise unremarkable and a urine dipstick in the office is only positive for 1+ proteinuria. What is the LEAST appropriate next step?

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Case #1

A. Check a urinalysis, CBC, C3, C4
   Search for systemic involvement from SLE or other disease (low WBC, low complements, etc.)
B. Check anti-ds DNA, Smith, and other ANA sub-serologies
   Again, looking for evidence of systemic lupus
C. Substitute another agent for hydralazine
   Addresses most likely cause of symptoms
D. Perform a kidney biopsy to rule out early lupus associated proteinuria
   Most likely has drug induced LE which is not often associated with organ involvement. Has other reason for proteinuria
Drug Induced SLE

- Well Characterized with the following drugs:
  - Hydralazine, Procainamide, Quinidine, Isoniazide
  - Minocycline can have somewhat different LE presentation

- Clinically
  - Patients are often > 50 (bias given medications involved?)
  - ANA is a diffuse or homogenous pattern
  - Sub-serologies are usually negative
  - Experience arthritis, serositis, cutaneous disease
  - Infrequent visceral involvement

- Disease improves with cessation of the agent
  - ANA may be persistently positive

The ANA
is not synonymous with “Lupus Test”

- Abnormal ANA does not equal SLE
  - 99.9% Sensitive for SLE
  - A negative ANA rules out most SLE
  - Poor Specificity
    - A Positive ANA does not rule in SLE
    - Other autoimmune diseases (scleroderma, sjogren’s, thyroid disease), medications, neoplasms, etc... associated with positive ANA
Using the ANA in the appropriate context

- Women: men 9:1
- Disproportionately affects African Americans
- Usually post-pubertal onset, affecting 3rd-5th decade
- Patterns of clinical involvement characteristic of SLE
  - Although not intended for use in diagnosis, the ACR “Diagnostic” criteria of SLE can help define the clinical context of patients

SLE: ACR Criteria:
4 of 11 Criteria without better explanation (Not diagnostic)

- Malar Rash
- Discoid Rash
- Photosensitivity
- Oral Ulcers
- Arthritis
- Serositis
- Renal Disorder
SLE Criteria Cont.

- Hematologic Disorder
- Immunologic Disorder
- Positive ANA
- Neurologic Disorder

Malar Rash

- Fixed malar distribution of erythema, flat or raised
Discoid Rash

- Erythematous raised patches with keratotic scaling and follicular plugging; some atrophic scarring in chronic lesions

Photosensitivity

- Skin rash as an unusual reaction to sunlight, by patient history or physical examination
Oral Ulcers

- Oral or nasopharyngeal ulcers, usually painless, observed by a physician

Arthritis

- Non-erosive arthritis involving two or more peripheral joints, characterized by tenderness, swelling, or effusion
Other SLE Criteria

- **Serositis**
  - Pleuritis (convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion)
  - Pericarditis (documented by EKG, rub, or evidence of pleural effusion)

- **Renal Disorder**
  - Persistent proteinuria >0.5g/day (or >3+)
  - Cellular casts of any type

Hematologic Abnormalities

- **Hemolytic anemia** (usually coomb’s positive)
- **Leukopenia** (WBC < 4,000 on at least 2 occasions)
- **Lymphopenia** (<1500 on 2 or more occasions)
- **Thrombocytopenia** (PLT<100,000 on 2 or more occasions)
**Immunologic Disorder**

One of the Following

- Anti-dsDNA
- Anti-Smith
- Positive findings of anti-phospholipid Abs
  - Abnormal level of either IgG or IgM CLIP Abs
  - Positive test for Lupus Anticoagulant (RVVT)
  - False positive RPR/VDRL > 6 months neg. FTA

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**Positive ANA**

- An abnormal titer of ANA in the absence of drugs known to be associated with “drug-induced lupus syndrome”
Neurologic Disorder

- Classically defined only as:
  - Seizures (in the absence of other causes)
  - Psychosis (in the absence of other causes)

SLE: Useful Facts of which the Internist should be Aware

- SLE patients are doubly susceptible to infectious complications 1. from SLE, itself, and 2. from the use of immunosuppressive therapies

- Infectious complications can mimic disease activity – so don’t be fooled by questions like this:
  - A 40 year old woman with a six year history of lupus develops fever, worsening respiratory failure, and infiltrates on chest CT. Your next best step is to:
    • A. Start prednisone therapy immediately
    • B. Check her anti-dsDNA titer
    • C. Arrange for an open lung biopsy
    • D. Start antibiotics, pan culture, and arrange for bronchoscopy
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SLE: Useful Facts of which the Internist should be Aware

- Infectious complications were classically thought of as leading cause of mortality

- Growing evidence that ischemic heart disease now leading cause of mortality
  - Risk of CV disease in SLE patients is 7-50 fold greater (dwarfs diabetes and cholesterol and other “traditional” CV risk factors!!)
  - Cardiac symptoms should be treated seriously in all SLE patients, including young women, and on all board exams!!!!

Case #2

A 72 year old woman presents complaining of profoundly increased pain and morning stiffness in her neck, shoulders low back and hips. She notes increased fatigue and a 10 pound unintended weight loss. No focal deficits are appreciated on her neurologic exam. Her PMH is notable for sciatica, hypertension, and COPD. Medications include HCTZ, Advair, and ibuprofen PRN. Xrays of her spine and hips are negative.
Case #2

All of the following are appropriate next steps in evaluating this patient except:

A. Start high dose prednisone 60 mg/day and arrange for urgent temporal artery biopsy
B. Question and examine the patient for more specific signs of Giant cell arteritis
C. Order an Erythrocyte Sedimentation Rate
D. Add 20 mg of prednisone empirically and assess her clinical response in 3-4 days
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Polymyalgia Rheumatica

- Demographic
  - Women:men 2:1
  - Rare before age 50
  - Traditionally most common in whites of northern European lineage

- Clinical
  - Proximal musculo-skeletal pain (shoulder girdle, neck> hips)
  - No true weakness like POLYMYOSITIS (this is not the same family of diseases!!!!!)
  - Morning stiffness, gelling, and feing OLD!!
  - Usually no palpable synovitis, although on ultrasound or MRI can see evidence of large joint bursitis
  - May have malaise, low grade fever
PMR: More Clinical Features

- Elevated ESR and/or CRP
- Association with Giant cell arteritis (But only 10-50% of time)
- Rapid and dramatic response to MODEST doses of prednisone (<20 mg/day)
  - No need to treat PMR with large doses of prednisone unless there is clinical suspicion of GCA
  - However, be wary of patients (and test questions) in whom one expects a diagnosis of PMR but there is no rapid response to modest doses of prednisone

Giant Cell Arteritis
Clinical Manifestations

- Demographics
  - Same as PMR (May be part of spectrum of same disease)
  - 40-50% develop PMR (may precede, follow, or occur concomitantly)
  - Rare before age 50.
  - The most common cause of vasculitis: increases in prevalence with each decade of life (less common in 50 year olds than in 80 year olds)
Giant Cell Arteritis
Clinical Manifestations

- **Headache (70-80% at one time or another)**
  - Commonly dull, aching, often over the temporal area but can be anywhere
  - Scalp tenderness may be present

- **Visual Changes**
  - Present in up to a third of patients
  - Blurred vision, diplopia, amaurosis fugax often presage blindness
  - Monocular blindness can be abrupt without warning
  - If persists >24 hours can be permanent

- **Jaw Claudication**
  - Most specific symptom for GCA
  - Classic presentation is discomfort over masseter muscles with protracted chewing
  - This is not pain at temporal mandibular joint

**Constitutional signs are common in this SYSTEMIC disease**
- Weight loss, Malaise
- Low grade fever in up to half of patients
- Cause of FUO in elderly
Giant Cell Arteritis

Work-up

- Establish pre-test probability of GCA using demographics, history, physical exam

- Laboratory Evaluation
  - ESR
    - >90% patients have an ESR >50; frequently >100
    - C-reactive protein may be more sensitive and be elevated in patients with normal ESR
  - CBC
    - Normocytic anemia, thrombocytosis

Giant Cell Arteritis

- Temporal artery biopsy
  - If elect to pursue biopsy, initiate prednisone 40-60 mg/day
  - Request 3-5 CM segment of artery.
  - Unilateral biopsy is >90% sensitive
  - 2 weeks of empiric prednisone does not significantly affect the sensitivity. Treat with large, long-term doses (40-60mg prednisone daily to start)
Case III
Question #1

- 55 year old male awakens with right knee pain and swelling one morning that worsens over next 48 hours until he has difficulty walking on that knee. He presents to your office complaining of knee pain, swelling, and low grade fever. On a recent Chem. 20 panel, uric acid level was elevated at 9.2. He denies any other joint pains, IVDU, or recent sexual contacts.

- Your next best step in managing this patient should be to:
  - A. Order an Xray of the Knee
  - B. Recheck the patient’s serum uric acid level
  - C. Perform an arthrocentesis
  - D. Treat the patient empirically for presumed gout

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Acute Gout

- Acute, usually self limited monoarticular inflammatory arthropathy
- Inflammatory response directed against monosodium urate crystals in synovium
- Usually but not always associated with hyperuricemia
- After attack, patient returns to normal during an asymptomatic inter-critical period that can last months or years
- Monosodium urate crystals precipitate around a UA concentration of 6.8, below the upper limit of “normal” in most US populations
Distribution of Serum Uric Acid Levels in Japan: 34,000 People

![Graph showing distribution of serum uric acid levels in males and females.](image)

**Acute Gout - Diagnosis**

- **Definitive:** Crystal Identification – the only way!
  - Joint fluid examination under polarized microscopy with red compensator
  - Strongly negatively birefringent needle shaped crystals
- **Suspected:** Characteristic radiographic “gouty” corticated erosions away from joint space
- **Possible:** Classic clinical picture with elevated serum urate – not diagnostic however!!!!
Acute Gout - Key Points

- **Arthrocentesis is required to:**
  - Confirm the diagnosis of gout
  - Exclude infectious arthritis, which can coexist in cases of known gout (gram’s stain and Cx.)
- **X-rays of benefit for suspected diagnoses and to follow radiographic progression**
- **Serum urate level does not confirm or refute a diagnosis of gout but can be used to monitor therapy**
Case III, Question 2

- Plain films demonstrate no acute changes, repeat uric acid level is now 10.7, and aspiration of the synovial fluid reveals numerous PMN's, WBC count of 75,000, intracellular needle-shaped negatively birefringent crystals, and a negative gram's stain. The patient is started on indomethacin and allopurinol and sent home. Which of the following actions in this case was a mistake?

- A. Allopurinol therapy
- B. Indomethacin therapy
- C. The patient was not admitted and treated empirically with antibiotics pending results of synovial fluid cultures
- D. None of the above
Case III, Question 2

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Acute Gout Therapy

Aimed at reducing the severity and duration of symptoms and reaching the "inter-critical period" sooner

- NSAIDs
  - Effective and rapid relief of symptoms
  - Contraindicated in patients with GI, Renal, or hypersensitivity concerns

- Corticosteroids
  - Intraarticular
  - Systemic

Colchicine: NOT APPROPRIATE THERAPY FOR ACUTE GOUT ANYMORE

- Uric Acid lowering therapy is not appropriate during acute gouty flare
Chronic Gout - Progression

- Recurrent inflammatory arthritic attacks separated by diminishing inter-critical periods of normalcy
  - Monoarticular or polyarticular
    - Same joint
    - Spread to other joints
      - General rule of thumb: most commonly involved joints: distal (podagra) to proximal

- Chronic inflammation/synovitis with no inter-critical period
  - Recurrent attacks blend together and patient's symptoms never return entirely to normal between attacks
  - Eventually, chronic inflammation remains

- Tophaceous gout:
  - Can occur with all of the above
  - Uric acid containing tophi deposit in joints/tendons/soft tissues, can lead to erosions and deformities
  - Chronic synovitis and tophaceous deformities can be difficult to distinguish from other inflammatory arthritis such as RA

Case III, Question 3

- The patient is started on allopurinol at 100 mg/day which is eventually increased to 200 mg/day. However, the patient experiences a second, and subsequently third, painful attack of gout over the next eight months. A repeat serum urate level indicates that the patient's uric acid level is now in the normal range at 6.9, however a foot film reveals the presence of a small tophaceous erosion in the 1st MTP joint. Your next best course of action is to do which of the following:

  A. Discontinue allopurinol for lack of efficacy and switch to daily colchicine
  B. Increase allopurinol to 300 mg./day and target a serum uric acid of less than 6.0
  C. Add colchicine to current allopurinol regimen
  D. Add prednisone to the current regimen
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Chronic Gout - Therapy

- **Goal:** reduce serum uric acid level
  - Lower serum urate associated with fewer attacks
  - Helps remove tophi/stores of uric acid
  - Goals of therapy, especially for tophi removal, are serum urate levels < 6.0. Max. dose of allopurinol is more than 200 mg/day in patients with normal renal function

Chronic Gout - Serum Urate Lowering Therapies

- **Allopurinol**
  - Xanthine Oxidase Inhibitor
  - Blocks metabolism of purines to uric acid
  - Effective for both under-excreters and overproducers of uric acid
  - Careful use in patients with renal failure
  - Associated with hypersensitivity syndrome that is DIFFERENT from rash
    - Fever, Steven’s-Johnson/TEN, hepatitis, marrow suppression, nephritis
Chronic Gout – Serum Urate Lowering Therapies

- Probenecid: Uricosuric blocks tubular re-absorption or uric acid
  - Useful in patients who under-excrete uric acid (90%)
  - If need be, confirm under-excretion with 24 hr. uric acid <800 mg/24 hrs.
  - Do not use if:
    - Tophi
    - Renal insufficiency
    - Clear overproduction syndrome

Gout Therapy - Key Points

- Do not initiate uric acid lowering therapy during acute attack
- DO use prophylaxis with either daily colchicine (0.6/day), NSAIDs, or prednisone when starting urate lowering medications... ? 3 months
- Do not treat asymptomatic hyperuricemia
- Use colchicine properly
  - Not for acute attacks
  - Yes for prophylaxis during urate lowering therapy
  - NOT preferred for chronic, long term management (will not prevent tophaceous complications)
  - But perhaps OK in that special patient intolerant of or refusing allopurinol but wanting to decrease acute flares
Case IV

- A 60 year old white female comes to your office for increasing right shoulder pain and limited range of motion. Her past medical history is notable for hypertension, nephrolithiasis, asthma, and a osteoarthritis of the knee that required a TKA two years prior. She takes only a beta blocker and inhaled corticosteroids. On examination, she has some tenderness of the supraspinatus tendon with minimal impingement with abduction, however - there is crepitus, diminished range of motion, and pain in the right shoulder with both abduction and external rotation. Shoulder and knee films are shown to the right.

Case IV, Question #1

- Which of the following features of this patient’s case, by itself, should prompt a more thorough diagnostic workup for her arthritis?

- A. Osteoarthritis involving the shoulder
- B. Total knee arthroplasty at age 58
- C. Tenderness to palpation of the rotator cuff
- D. Use of beta blockers
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Osteoarthritis – Key Point

- Certain joints do not usually experience degenerative changes
  - Glenohumeral joint shoulder
  - Elbow
  - Ankle
  - Wrist
- Common joints affected by osteoarthritis
  - Acetabular joint of hip
  - Knee
  - DIP/PIP joints of hands/feet
  - AC joint of shoulder
  - Spine

Case IV, Question #1

- B. Her R. knee OA and subsequent TKA is possibly/probably related to shoulder arthritis, but in and of itself, OA knee is not uncommon in people entering their seventh decade.

- C. Tenderness of the rotator cuff may indicate rotator cuff tendinitis, but this entity is not necessarily associated with other medical causes. Treatment usually consists of physical therapy, corticosteroid injections, and/or surgery if damage to the rotator cuff or excessive calcification hinder function.

- D. Use of beta blockers is irrelevant to this case

- A. OA of the shoulder is unusual and usually caused by previous trauma or other medical causes
Case IV, Question #2

Which of her following co-morbid medical conditions may help explain her current symptoms?

- A. Asthma
- B. Hypertension
- C. Nephrolithiasis
- D. Her use of inhaled corticosteroids

Which of her following co-morbid medical conditions may help explain her current symptoms?

- A. Asthma
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- C. Nephrolithiasis
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Case II, Question #2

- Which of her following co-morbid medical conditions may help explain her current symptoms?
- A. Asthma
- B. Hypertension
- C. Nephrolithiasis
- D. Her use of inhaled corticosteroids

CPPD – Calcium Pyrophosphate Dihydrate Deposition Disease
CPPD

- Several distinct clinical forms:
  - Pseudogout (25%): Acute inflammatory mono-arthritis mimics gout
  - Pseudo-RA (5%): Synovitis and degenerative changes of MCP’S (especially 2nd and 3rd)
  - Accelerated OA/DJD of unusual joints
  - Spinal Involvement (fever, neck pain)
  - Asymptomatic chondrocalcinosis

CPPD Associations

- Some Associated Metabolic Disorders
  - Hyperparathyroidism (This patient with renal calculi!)
  - Hemochromatosis
  - Hypothyroidism
  - Acromegaly
  - Ochronosis
  - Wilson’s disease
  - Others
Synthesis of actual ABIM Question
C. 2000

- 50 year old male presents with progressive pain and limited range of motion of right knee.
- Xray shown to left
- Most likely abnormal test finding to be:
  - Elevated glucose
  - Elevated ferritin
  - Abnormal transaminases
  - All of the above!!!!!

Be prepared - know how to test for hyperparathyroidism and hemachromatosis

Case V, Question #1

- A 68 year old woman presents with progressive weakness of her proximal arms and legs to the point where she is having difficulty arising from a chair and combing her hair. On ROS, she notes no joint or muscle pain or stiffness, but admits to a 25 pound weight loss and recent onset of a unilateral temporal headache. Her past medical history is significant for hypercholesterolemia and COPD, and she is treated with only simvastatin and Spiriva. She had previously been on a 2 month prednisone taper for a COPD flare. Which of the following factors in her history is LEAST likely to be associated with her proximal muscle symptoms?

- A. Her history of hyperthyroidism
- B. Her use of “Statins” for elevated cholesterol
- C. Her new unilateral temporal headache
- D. Her use of prednisone
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Myopathies

- A. Both hyperthyroidism and hypothyroidism can be associated with myopathy and proximal muscle weakness.

- B. Statin use is associated with myopathy, muscle pain and/or weakness.

- C. The unilateral temporal headache is not specifically associated with myopathy. It is a harbinger of giant cell arteritis in patients with polyMYLAGIA rheumatica, not polyMYOSITIS.

- D. Her use of prednisone may be relevant. Chronic prednisone use can cause a steroid-induced myopathy that can be associated with weakness and minimal CK elevations. In addition, withdrawal of chronic corticosteroid therapy can induce Addisonian symptoms and myopathy.

Case V, Question #2

- On physical examination, she appears thin, with a erythematous rash over her eyelids and elbows, and has clear muscle wasting over her deltoid and quadriceps muscles. Routine labs reveal a normal CBC and chemistry panel, an elevated CK of 4,300, a normal CXR, and an MRI of her legs followed by a quadriceps biopsy that are both consistent with a proximal inflammatory myositis.
Case V, Question #2

- She is diagnosed with dermatomyositis and started on prednisone 60 mg/day and 20 mg weekly of parenteral methotrexate with marked initial improvement in her weakness; her prednisone dose is slowly tapered to 20 mg/day. However, nine months later, she develops increased cough and dyspnea on exertion that progresses to SOB at rest.
Case V, Question #2

- Exam notable for low grade fevers, crackles in her bilateral lung fields and somewhat improved muscle strength
- Pulse oxymetry low 90’s rest, 80’s with ambulation
- CXR demonstrates bilateral pulmonary infiltrates, CK levels are at 600.
- CT with alveolar GGO and interstitial fibrosis

The most appropriate next step includes which of the following?

A. Initiation of therapy for community acquired pneumonia and PCP
B. Scheduling her for induced sputum and bronchoscopy
C. Discontinuing her methotrexate and considering use of another anti-rheumatic agent
D. All of the above
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D. All of the above
What are the possible causes for this patient’s decompensation??

- Infections (immunosuppression)
  - Bacterial, viral, PCP

- Medications
  - Methotrexate: hypersensitivity pneumonitis

- Cardiac (pulmonary edema): myocardial involvement rare

Polymyositis, Dermatomyositis: Effects on the Lung

- Respiratory muscle compromise

- Dysphagia and aspiration pneumonitis

- Interstitial lung disease
Inflammatory Myopathies: Quick Facts

- Polymyositis, Dermatomyositis, Inclusion Body myositis
- Proximal muscle weakness is hallmark of PM and DM, more distal weakness IBM
- Proximal muscle pain is NOT feature
- Severe disease can affect diaphragm and swallowing
  - Interstitial lung disease common
- Elevations of CK and Aldolase are common, higher levels in PM/DM than IBM
- DM>>PM>>IBM association with malignancy (PM/DM: ovarian>breast/lung/GI)
  - Workup for visceral malignancy appropriate for DM
- Treatment with corticosteroids and immunosuppressive therapies mainstay
Question #1

Which of the following is the most life-threatening complication of this disease?:

- A. Respiratory Compromise
- B. Scleritis
- C. Glomerulonephritis
- D. Cardiac Arrhythmia
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- C. Glomerulonephritis
- D. Cardiac Arrhythmia

Relapsing Polychondritis

- Inflammation of the cartilaginous tissues (Type II collagen)
- Elastic cartilage of the ears and nose, the hyaline cartilage of peripheral joints, cartilage in the tracheobronchial tree and collagen in the eye
- Can cause blindness (scleritis), deafness (inner ear ossicles involvement), and systemic symptoms
- Stridor, hoarseness, pneumonia, respiratory compromise are ominous signs
Potpourri #2

67 yo man reports increased falls, fatigue, 15 pound weight loss over 2 months. On exam he has a diffuse, non-palpable, lacy skin lesion across his legs with right foot drop and left wrist weakness. ESR 109.

Which of the following infections is likely contributing to his illness?

A. Hepatitis B
B. Parvovirus B19
C. Coxsackie
D. Epstein-Barr Virus
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Potpourri #2

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Potpourri #2
Livedo reticularis in Polyarteritis nodosa

- PAN is a medium vessel vasculitis
- Clinical findings are secondary to ischemia
  - Cutaneous - livedo reticularis, nodules, ulcers
  - Renal - renal insufficiency, hematuria (no RBC casts)
  - Neuro - mononeuritis multiplex
  - GI - intestinal ischemic
  - Systemic - weight loss, fatigue, fevers
- 10% of PAN associated with Hep B (prevalence falling as HBV vaccine use increases)

Potpourri Question #3

- 45 Year old female with progressive skin tightening of her hands, no history of GERD or Raynaud's phenomenon, and negative ANA.
Potpourri Question #3

The most likely positive test result in this patient would be:

- A. Rheumatoid Factor
- B. Anti Scl-70
- C. Anti-Centomere Antibody
- D. Elevated HgA1C

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  - D. Elevated HgA1C

Soft Tissue complications of Diabetes

- Limited Joint Mobility Syndrome (Cheiropathy – shown)
- Adhesive Capsulitis
- Flexor tenosynovitis, tendon nodules (trigger finger), and dupuytren’s stenosing tenosynovitis
- Carpal Tunnel Syndrome
- Charcot Arthropathy – DJD X 1000, including unusual joints like ankle
- ANA negative patients are unlikely to have positive subserologies like scl-70 or centromere!
Potpourri #4

47 yo woman complaining of dry eyes, dry mouth, who is anti-SS-A+ is diagnosed as sjogren’s syndrome, complains of dry cough, SOB with a chest x-ray demonstrating hilar lymphadenopathy undergoes a gallium scan that shows the accompanying result

Potpourri #4

Which of the following is the most likely diagnosis?
A. Diffuse Infiltrative Lymphocytosis (DILs)
B. Sjogrens
C. Sarcoid
D. Mycobacterium tuberculosis
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Potpourri #4

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C. Sarcoid
D. Mycobacterium tuberculosis
Potpourri #4
Sjogren’s Syndrome

- **Demographic**
  - Female:Male 15:1

- **Clinical**
  - Muco-Cutaneous
    - Dry mouth - aggressive periodontis
    - Dry skin - scaling, pruritus
    - Bilateral parotitis
  - Keratoconjunctivitis - dry, “gritty” eyes
  - May be primary or secondary, accompanying RA/SLE/MCTD

- **Laboratory studies**
  - >90% RF+
  - > 80% ANA+ (speckled pattern)
  - 60% are anti-SS-A (Ro) or anti-SS-B (La)

- **Tissue biopsy**
  - Minor salivary gland - send fungal and TB stains

Potpourri Question #5: Fever, Arthritis, Skin Rash, Renal Failure, and Sensory Neuropathy
Potpourri Question #5

- The least likely positive test result in this patient

- A. Positive rheumatoid factor
- B. Low C4
- C. Positive cold agglutinin test
- D. Positive hepatitis serology

The least likely positive test result in this patient

- A. Positive rheumatoid factor (43%)
- B. Low C4 (15%)
- C. Positive cold agglutinin test (36%)
- D. Positive hepatitis serology (6%)
Potpourri Question #5

- The least likely positive test result in this patient
  
  - A. Positive rheumatoid factor
  - B. Low C4
  - C. Positive cold agglutinin test
  - D. Positive hepatitis serology

Cryoglobulinemia

- Type I
  - Monoclonal IgG or IgM
  - Associated with hematologic malignancy, especially myeloma
  - Acrocyanosis and tissue ischemia

- Type II
  - Monoclonal IgM directed against polyclonal IgG. Immunologically active>ischemia
  - Rheumatoid factor positive
  - Low complement levels
  - Associated with HCV (sometimes Sjogren’s and SLE)

- Type III
  - Polyclonal IgG and IgM
  - RF positive and low complement levels
  - Associated with HCV, Sjogren’s Syndrome, SLE

- Do not confuse Cold Agglutinins and Cryoglobulins!!
Potpourri Question #6

Oral Ulcerations

Which of the following untreated diseases is least likely associated with this oral lesion?

A. Behcet’s Disease
B. Reactive Arthritis (Reiter’s syndrome)
C. Rheumatoid Arthritis
D. Systemic Lupus Erythematosus
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