Lupus and Friends
Perspectives on common syndromes
and
Primary care responses

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Lupus and Friends

Systemic Lupus Erythematosus
What it is and what it’s not

Sjogren's Syndrome
Perspectives on dryness

Raynaud's Phenomenon
When to worry
Disclosures

- No speakers bureaus
- Clinical trials contracts:
  Gilead, Roche - Genentech, UCB, Amgen, Merck, BMS, Sun Pharma, Janssen, Abbvie, Pfizer, AstraZeneca/MedImmune, Mallinckrodt, HGS/GSK, Lilly, Sanofi, Celgene, Takeda
Systemic Lupus Erythematosus (lupus)

A multisystem disease that is caused by tissue damage resulting from antibody and complement - fixing immune complex deposition.
Epidemiology

- Prevalence in the U.S. is estimated about 1/20,000
- SLE clearly more prevalent in women with a ratio of about 9:1 as compared to men
- Three times more common in African-Americans than in whites in the U.S.
Classification Criteria

- Serositis
- Oral Ulcers
- Arthritis
- Photosensitivity
- Blood
- Renal manifestations
- ANA
- Immunologic tests
- Neurologic manifestations
- Malar rash
- Discoid rash

4 of these 11 criteria
Dermatologic features of SLE

- Malar rash
- Discoid
- Patchy alopecia (scarring)
- Psoriasiform
- Palpable purpura

Subacute cutaneous lupus
Dermatologic features of SLE

- Not just a flush
- Persistent, raised, scarring, ulcerating
- Non-lupus rashes occur in patients with lupus
Oral manifestations

Oral ulcers
- painless
- shallow
- hard and soft palate in location
Musculoskeletal features

- Arthralgias and/or arthritis are the most common presenting manifestations
- Swelling tends to be relatively mild
- Non-erosive arthritis that is reducible - Jaccoud’s arthropathy
Musculoskeletal features

• Myositis, myalgias and weakness can occur

• Histologically different from idiopathic polymyositis-displaying microtubular inclusions and mononuclear infiltrate with occasional CPK elevation and fiber necrosis
Neurological manifestations

- Seizures- grand mal, petit mal, focal, temporal lobe
- Movement disorder - choreiform
- Headaches – not responsive to narcotics
- Cranial neuropathies - visual defects, blindness, papilledema, nystagmus, ptosis or facial palsy
- Retinal vasculitis
- Transverse myelitis
Psychiatric features

- Overt psychosis
- Organic brain syndrome
  delirium
- emotional inadequacy
- cognitive impairment
Renal Manifestations

- Proteinuria of > 0.5g per 24 hrs.
- Presence of RBC casts
- Renal biopsy - WHO classification
  - class II  Mesangial
  - class IIIa Focal segmental glomerulonephritis
  - class IIIb Focal proliferative glomerulonephritis
  - class IV  Diffuse proliferative glomerulonephritis
  - class V   Membranous glomerulonephritis
  - class VI  Advanced sclerosing glomerulonephritis
Renal Manifestations

Biopsies

Class II

Class III

Class IV

Class V

Class VI
Renal Manifestations

- Active urinary sediment – proteinuria and/or rbc casts

- Bilateral ankle edema

Call rheumatologist/nephrologist today
Pulmonary features

- Lupus pneumonitis
- Lupus pleuritis
- Pulmonary hemorrhage - due to pulmonary vasculitis
- Pulmonary hypertension - r/o APLS
- Pulmonary embolism - linked to APLS
Pulmonary features

- Pleuritis
- Interstitial pneumonitis
- Shrunken lung - fibrosis
- Pulmonary hemorrhage

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Cardiac features

- Pericarditis - most common cardiac manifestation 20 - 30% may see leukocytosis, ANA and low complement in fluid
- Myocarditis - suspect in patients with arrhythmias, conduction defects or unexplained cardiomegaly
- Endocarditis - Libman- Sacks- nonbacterial vegetations
- Coronary Artery Disease - coronary vasculitis, coronary occlusion- linked to APLS and CAD from steroid use.
Gastrointestinal features

- Mesenteric vasculitis
- Lupus peritonitis
- Pancreatitis - occurs in 8% of patients
- Lupoid hepatitis - now called autoimmune hepatitis
Types of lupus

• Non-visceral
  skin, joints, serosa, mucosa, blood

• Visceral
  kidney, brain, lung, liver, pancreas, artery
# Laboratory features

- **Immunological tests**
  - ANA positive (screen positive / zero titer is a negative test)
  - Anti- Double-stranded DNA antibodies
  - Anti-smith antibodies (Anti-Sm)
  - Anti-Histone antibodies - drug induced lupus
  - Anti Ro/La  SSA/SSB

- **Pancytopenia**
  - Hemolytic anemia - Coombs +
  - Leukopenia- in particular lymphopenia
  - Thrombocytopenia
Treatment

• NSAIDs - pericarditis and musculoskeletal
• Steroids- for acute flare of systemic disease

  *The “P” in prednisone is for “poison”* – Michelle Petri

• Hydroxychloroquine- prevention and steroid sparing
• Azathioprine - Rx for nephritis or steroid sparing
• Cyclophosphamide - for DPGN
• Methotrexate- cutaneous and articular manifestations
• Mycophenolate mofetil – renal and pulmonary features
• Belimumab IV – most non-renal features
• IV IG- when all else fails
• Plasmapheresis - when all else really fails- last ditch effort
Cautions

• Never diagnose lupus by antibodies alone (ANA, dsDNA, SS-A, SS-B, RNP, etc.)
• Never diagnose lupus without the concordance of a rheumatologist
• Always get a UA with a microscopic if you suspect SLE
• Avoid starting corticosteroids – call the rheumatologist
• Avoid the default attribution trap
• Avoid the hypermobility trap
Sjogrens Syndrome

- Autoimmune Exocrinopathy
- Occasionally primary
- Usually occurs with other rheumatic disorders
  - rheumatoid arthritis
  - systemic lupus erythematosus
  - scleroderma
  - autoimmune hepatitis
  - primary biliary cirrhosis
I’m still waiting...
Sjogren's Syndrome

Sicca complex

• Dry eyes
• Dry mouth
• Genital dryness
• Skin dryness
• Usually Sjogren's is low on the list for persons complaining of dryness
More common reasons for dryness

- Advanced age
- Female gender
- Hormonal changes (primarily due to decreased androgens)
- Systemic diseases (e.g., diabetes mellitus, Parkinson disease)
- Contact lens wear
- Systemic medications (antihistamines, anticholinergics, estrogens, isotretinoin, selective serotonin receptor antagonists, amiodarone, nicotinic acid)
- Ocular medications (especially those containing preservatives)
- Nutritional deficiencies (e.g., vitamin A deficiency)
- Decreased corneal sensation
- Ophthalmic surgery (especially corneal refractive surgery)
- Low humidity environments
Sjogren's Syndrome

- SS-A and SS-B
- Minor salivary gland biopsy
Raynaud’s Syndrome
Raynaud’s Syndrome

Raynaud’s (blanching)  Acrocyanosis
Raynaud’s Syndrome
Nail Fold Capillaroscopy

A
B
C
D
Nail fold capillary damage
Buergers Syndrome (thromboangiitis obliterans)

- cigarettes
- cigar smokers
- marijuana users (cannabis arteritis)
- chewing tobacco
- snuff
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