Scleroderma
Systemic Lupus Erythematosis
Dermatomyositis

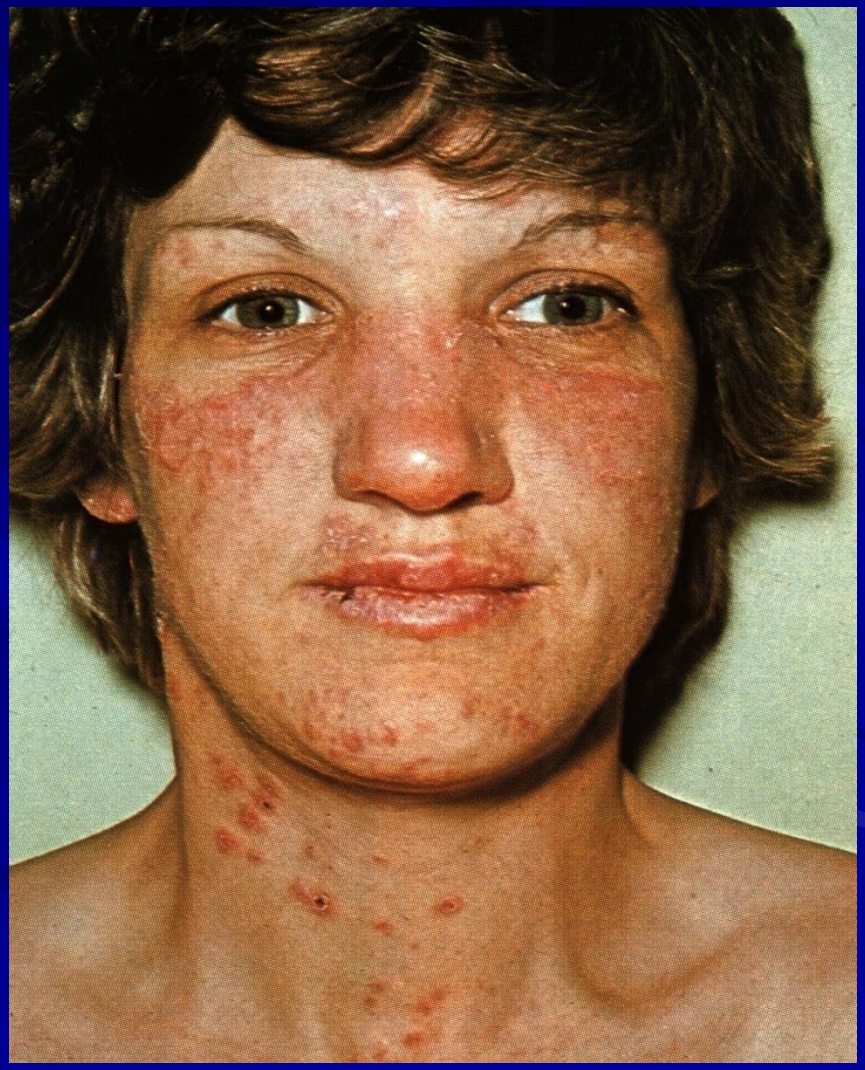
A.C.O.I. Board Review 2018

Howard L. Feinberg, D.O., F.A.C.O.I., F.A.C.R.

(No Disclosures)

Systemic Lupus Erythematosis

- Incidence 15-50/100,000
- Peak age 17-40
- Female:Male
  - 5:1 in peak ages
  - 2:1 for all age groups



Variants

- Subacute Cutaneous Lupus
  - non-fixed rash
  - non-scarring
  - associated with SS-A antibody
- Discoid
  - Scarring rash with central atrophy
- Lupus Pernio
  - variant of sarcoidosis
  - violacious plaques over the face, ears,
  - 86% - hepatic granulomas
  - 20%-hepatomegaly
- Systemic (SLE)

DRUG INDUCED LUPUS

- procainamide
- isoniazide
- hydralazine
- methyldopa
- chlorpromazine
- dilantin
- quinidine
- penicillamine
- possible association - griseofulvin antibiotics, gold salts

- ANA positive for up to 1 year
- Anti-histone antibody positive in 95%
- No change in complement
- CNS and renal disease are rare
- usually mild disease

ACR SLE Criteria

(must have at least 4 of 11)

- 1) Malar Rash
- 2) Photosensitive Rash
- 3) Discoid Rash
- 4) Oral Ulcers
- 5) Serositis
- 6) Arthritis
- 7) CNS
- 8) Renal
- 9) ANA
- 10) Hematologic
- 11) Other Lab (anti-double stranded DNA, ENA, VDRL)

2012 SLICC SLE Clinical Criteria

- Acute cutaneous lupus
- Chronic cutaneous lupus
- Oral ulcers
- Non scarring alopecia
- Synovitis (2 or more joints)
- Serositis
- Renal (>500mg protein or RBC casts)
- Neurologic (Seizure, psychosis, myelitis, mononeuritis multiplex, acute confusion, cranial neuropathy)
- Hemolytic anemia
- Leukopenia
- Thrombocytopenia

2012 SLE Laboratory Criteria

- ANA
- Anti-DNA
- Anti-SM (Smith)
- Antiphospholipid
- low complement
- Direct coombs without hemolysis

2012 SLE Diagnostic Criteria

Option One

- 4 criteria
  - At least one clinical criteria
  - At least one immunologic criteria

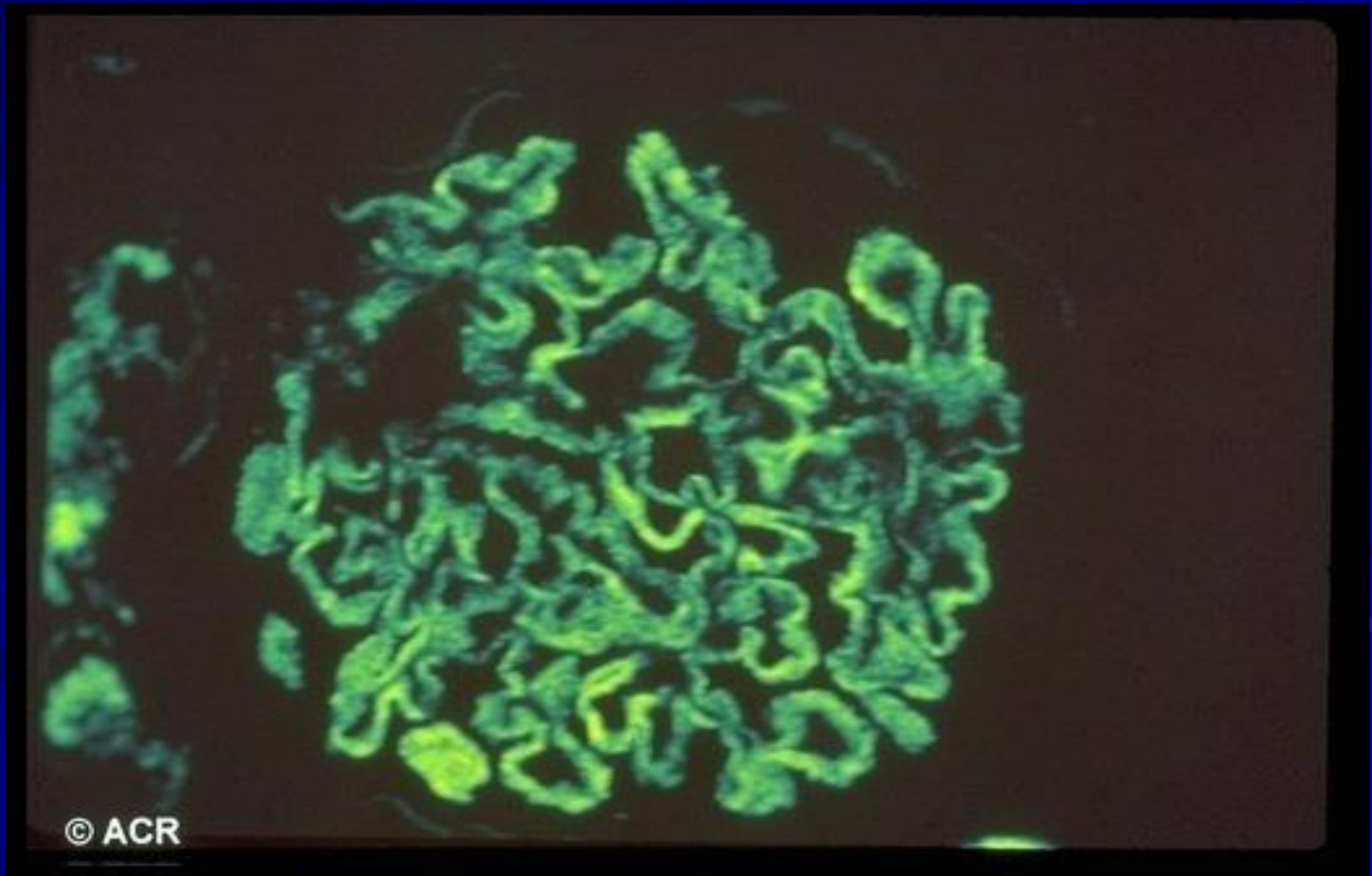
Option Two

- Biopsy proven nephritis
- Positive ANA or anti double stranded DNA

Systemic

- CNS
  - seizures
  - psychosis
- Cardiac
  - Libman-Sacks Endocarditis
  - arrhythmia
- Renal
  - Normal
  - Mesangial lupus nephritis
  - Focal proliferative lupus nephritis
  - Diffuse proliferative glomerulonephritis
  - Membranous glomerulonephritis
  - Sclerosing

IgG DEPOSITS IN SLE GLOMERULOUS

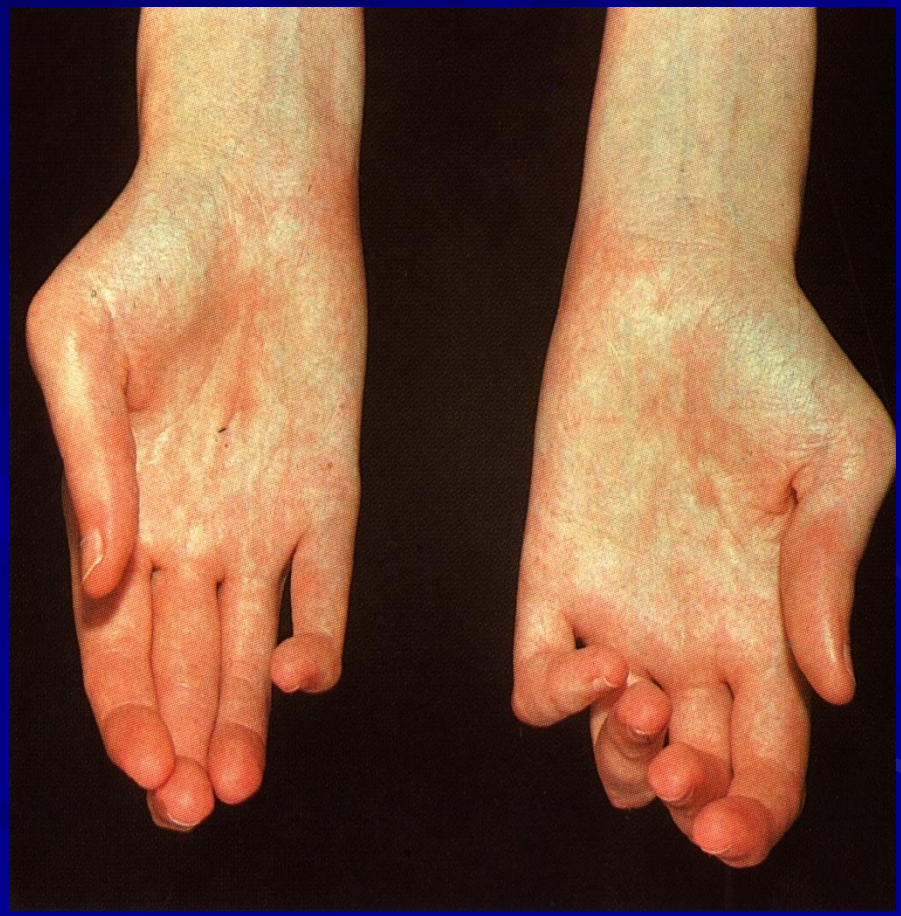


Treatment

- Symptomatic
- Hydroxychloroquine (Plaquenil)
- Azothiaprim (Imuran)
- Methotrexate
- Steroids
- Cyclophosphamide (Cytoxan)
- Mycophenolate mofetil (CellCept)
- Belimumab (Benlysta)
- Bone Marrow Transplant

Scleroderma

- Incidence approximately 0.4-1/100,000
- Peak age 30-55
- Female:Male 7.5:1



Variants

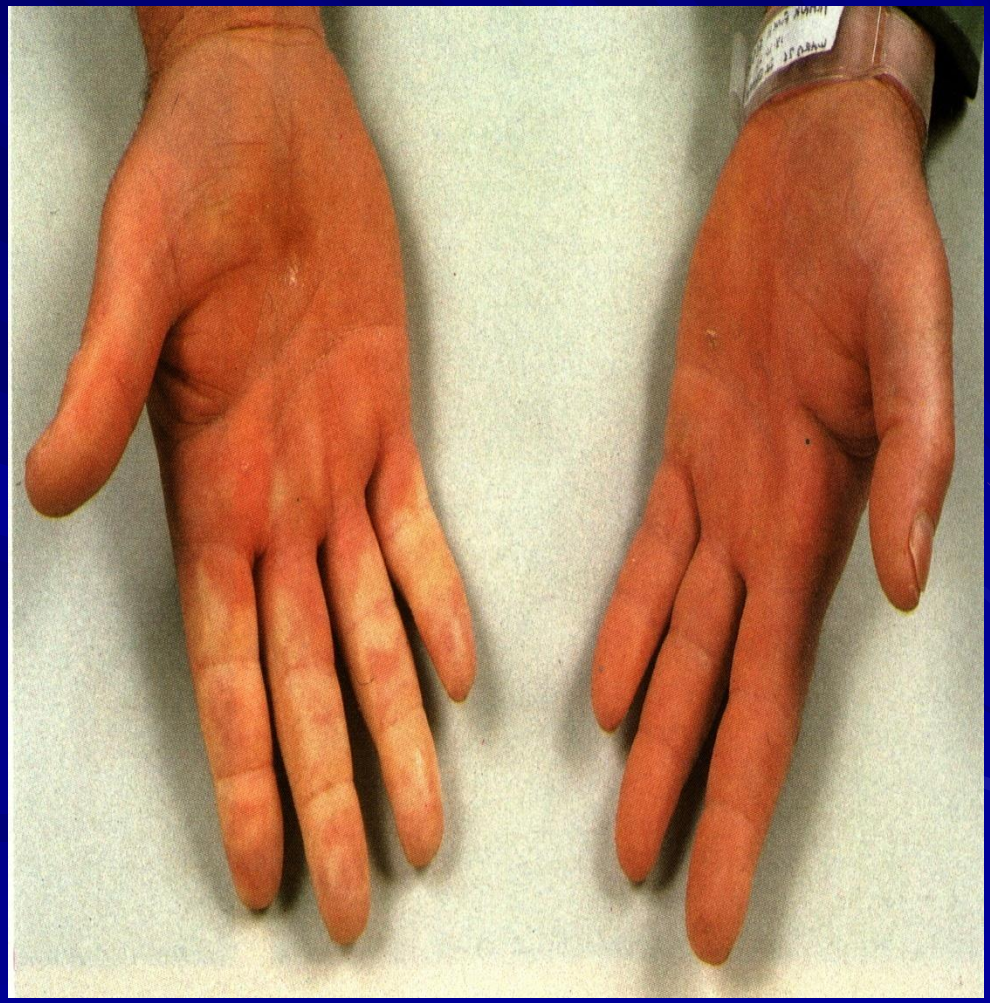
- Localized
  - anticentromere antibody
  - Linear
  - morphea
- CREST
- Toxic
- Progressive Systemic Sclerosis
  - ANA
  - SCL-70 antibodies

Localized Scleroderma

- primarily in extremities
- no internal organ involvement
- usually does not require treatment or intervention
- includes linear scleroderma and morphea

CREST

- Calcinosis
- Raynauds
- Esophageal Motility
- Scleroderma
- Telangiectasia



TOXIC SCLERODERMA

- Toxic Oil - rapeseed oil
- Eosinophilia myalgia syndrome - L-tryptophan (rash, fever, arthralgias)
- Diffuse fasciitis with eosinophilia (Shulman's Syndrome)
  - no systemic features
  - usually follows exercise or trauma
  - mainly affects a single limb

Progressive Systemic Sclerosis

- Pulmonary Fibrosis
- Cardiac Fibrosis
- Raynauds
- Sicca Complex
- Renal
  - accelerated hypertension
  - renal crisis

Criteria (requires 9 points)

- Skin thickening of the fingers of both hands proximal to MCP joint (9 points)
- Skin thickening of the fingers distal to MCP (4 points) or puffy fingers (2 points)
- Telangiectasia (2 points)
- Nailfold capillaries (2 points)
- PAH or ILD (2 points)
- Raynaud's (3 points)
- Autoantibodies (3 points)
  - Anticentromere
  - Anti-topoisomerase
  - Anti-RNA polymerase III

Treatment

- Mycophenolate mofetil (CellCept)
- Penicillamine (Cuprimine, Dpen)
- Steroids
- Methotrexate
- Cyclophosphamide
- PAH
- Other Immuosuppresive Therapy
- Bone Marrow Transplant

IDIOPATHIC INFLAMMATORY MYOPATHIES

Polymyositis

Dermatoyositis

Inclusion Body Myositis

Malignancy Associated Myositis

Juvenile Dermatomyositis

Variants

- Polymyositis
- Dermatomyositis
- Inclusion Body Myositis
- Malignancy Associated Myositis
- Pediatric Poly/Dermatomyositis
- Amyopathic Dermatomyositis
- Collagen Vascular Disease Associated
- Mixed Connective Tissue Disease

POLYMYOSITIS

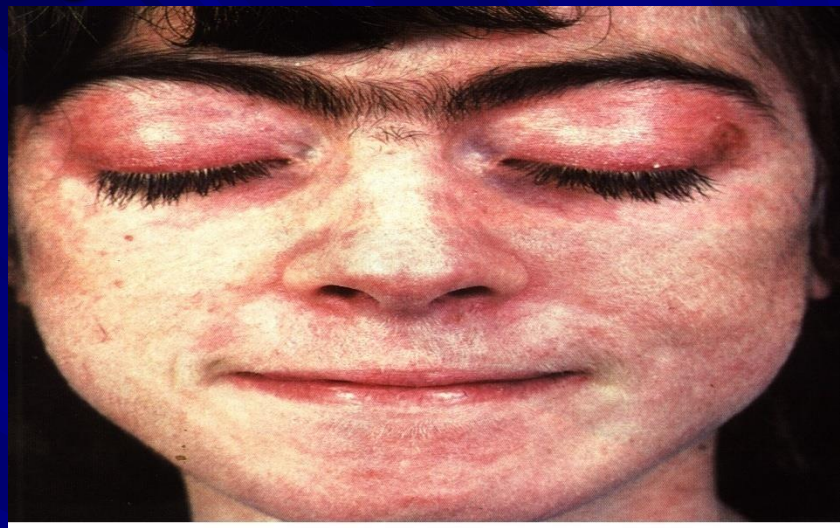
- Female : Male 2:1
- Incidence 0.5-8.4/million
- Peak ages
  - 10-15 - pediatric
  - 45-60 - adult

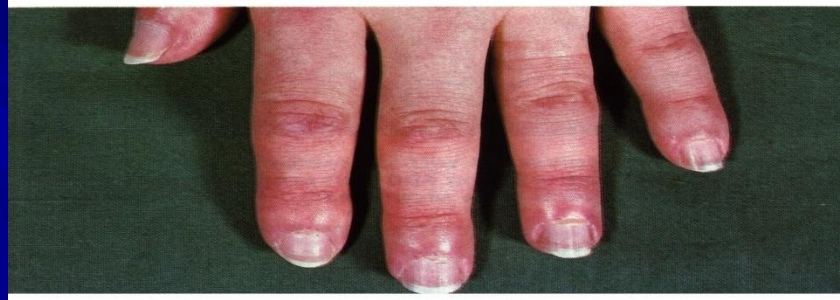
Polymyositis

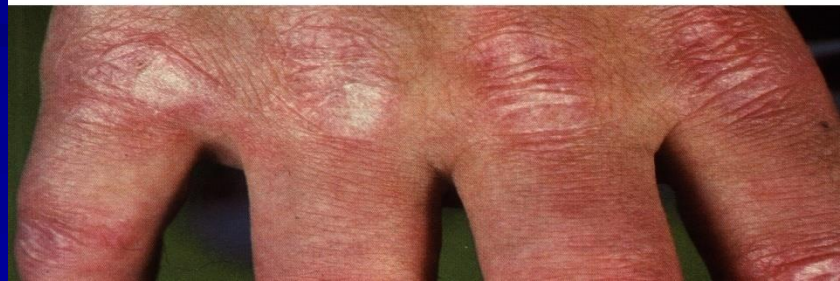
- proximal muscle (hip and shoulder girdle) weakness
- weakness without pain
- no rash
- elevated CPK

Dermatomyositis

- Rash
  - heliotrope rash
  - Shawl sign
  - V - sign
  - mechanics hands
  - Gottrons papules
- All other features of Polymyositis

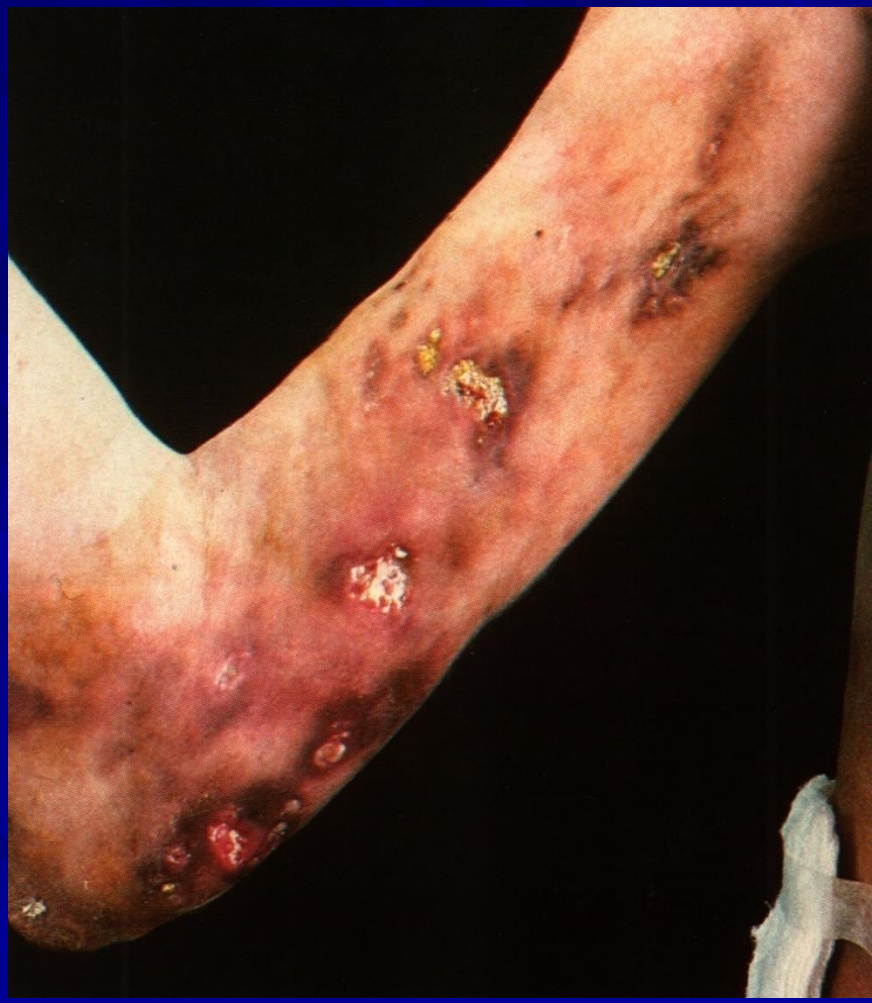






Non Muscle Manifestations

- Calcinosis
- Interstitial Lung Disease
- Cardiac muscle
- Coronary artery disease (rare)
- Dysphagia
- Reflux (50%)
- Diarrhea, constipation, abdominal pain
- Pericarditis



Inclusion Body Myositis

- identical clinical features to Polymyositis/Dermatomyositis
- on electron microscopic evaluation of muscle biopsy specific inclusions are seen
- refractory to treatment
- familial

Malignancy Associated Myositis

- Clinically identical to Poly/Dermatomyositis
- Increasing likelihood with increasing age of patient
- More common in Dermatomyositis
- The most common malignancy for age is the most common seen
- Increased incidence with ovarian cancer

Antisynthetase Syndrome

- 20% of polymyositis/Dermatomyositis patients
- More common in Polymyositis than Dermatomyositis
- MSA on lab
- Raynaud's
- Interstitial Lung Disease
- mechanics hands
- Fever
- non erosive symmetric polyarthritis of the small joints

Signal Recognition Particle Antibody Associated Myopathy

- Associated with severe polymyositis and cardiac involvement
- Test positive for anti SRP antibodies
- Approximately 4% of cases of polymyositis

Amyopathic Dermatomyositis

- Typical skin lesions of Dermatomyositis
- No muscle weakness
- Normal CPK
- May have fatigue
- Due to abnormal ATP in muscles

Diagnosis

- Weakness
- Elevated CPK
- EMG/NCS
- Muscle Biopsy
- +/- Rash
- Lab

Antibody Testing

- Myositis Specific Antibodies (MSA)
  - Seen in 40% of cases
  - Associated with anti-synthetase syndrome
  - Anti RNA synthetase
    - Jo-1 20-30%
    - Anti PL-12
    - Anti-EJ
    - Anti – OJ
    - Anti – PL7
    - Anti – KS
- Anti Signal Recognition Particle Antibody (Anti-SRP)
  - Seen in 4% of patients
  - Associated with poor prognosis and Anti-SRP Myopathy

Antibody Testing

- ANA
  - Seen in 50-80%
  - Pattern
    - Homogeneous
    - Speckled
    - Nucleolar
- Amyopathic DM
  - Anti – CADM 140
- Juvenile
  - Anti – MJ antibody
- Myositis associated antibodies (MAA)
  - Seen in 20-50% of cases
  - Anti – SSA
  - Anti - PM
- Overlap
  - Anti – RNP
  - Anti - Ku

Antibody Testing

- Chromodomain helicase DNA binding proteins 3 and 4
  - Dermatomyositis

Treatment

- varies with disease type
- Prednisone
- Methotrexate
- other immunosuppressive therapy
- Physical therapy

Differential Diagnosis

- Eosinophilic Myositis
- Focal Myositis
- Giant Cell Myositis
- Drug induced myositis/myopathy
- Infectious Myositis
- Metabolic Myopathy

POLYMYALGIA RHEUMATICA

Criteria (4 points)	Without Ultrasound	With Ultrasound
Morning stiffness	2	2
Hip pain/decreased ROM	1	1
Neg. RF or anti CCP	2	2
Absence of other joint pain	1	1
Ultrasound (subdeltoid bursitis, biceps Tenosynovitis, glenohumoral synovitis)		1

Required Criteria – Elevated ESR/CRP, Age >50, Shoulder girdle muscle pain

Contact Information

Howard Feinberg, D.O., F.A.C.O.I., F.A.C.R.
1310 Club Drive
Vallejo, CA 94592

Howard.Feinberg@TU.edu