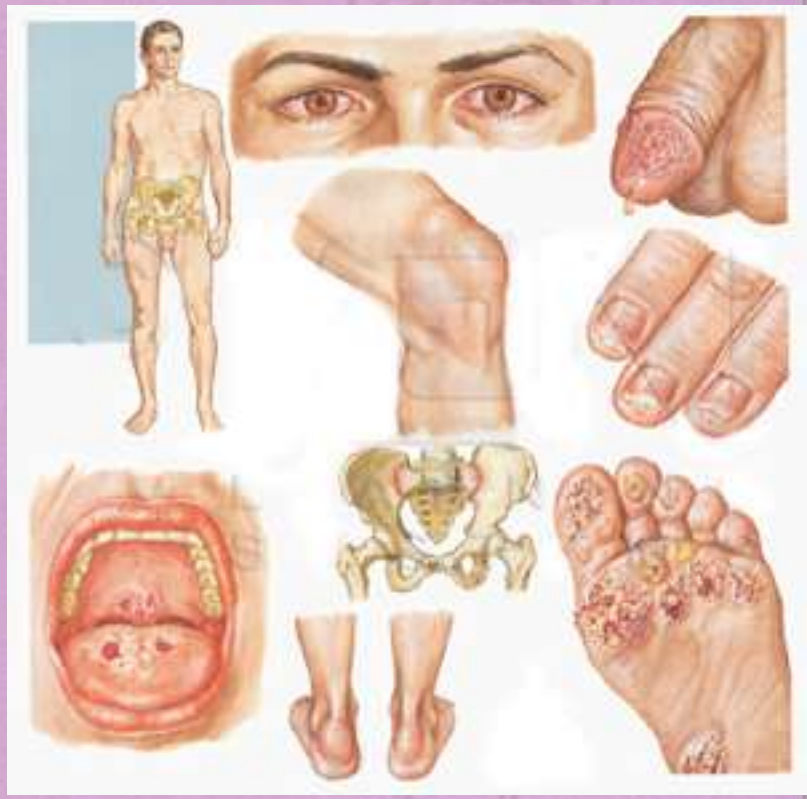
Reiter's Syndrome:
Clinical Features

- Arthritis lower extremities
- Enthesitis
- Spondylitis
- Tenosynovitis
- Dactylitis
- Urethritis
- Uveitis
- Oral ulcers
- Keratoderma
- Balanitis
- HLA B-27 Positive (80%)



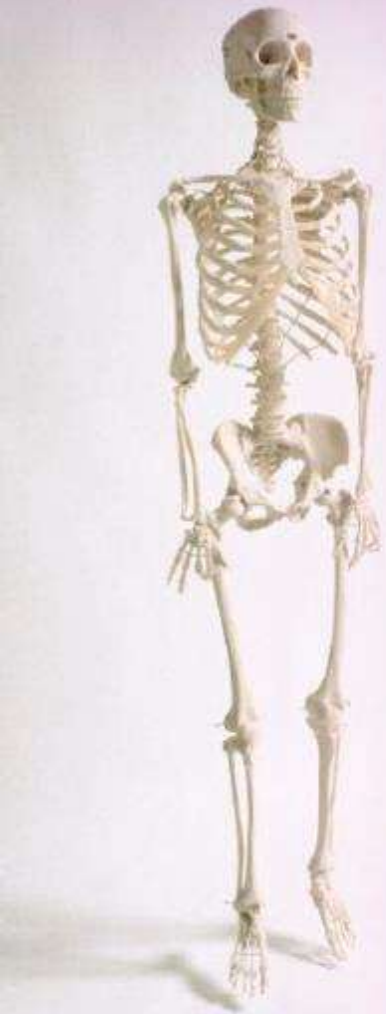
Reactive Arthritis:
Laboratory Findings

- Clinical !!! (History is vital)
- Laboratory (confirmatory)
  - ESR and CRP - Elevated during acute phase
  - Negative RF, ANA
  - Synovial fluid:
    - High WBC count, (often w/ elevated PMNs)
    - Gram stain and culture (to exclude septic arthritis)
  - Throat, stool, or urogenital tract cultures if indicated to isolate causative organism.

Reactive Arthritis:
Treatment

- Infectious Disease:
  - Eliminate “triggering infection”
- Extra-articular Disease
  - Typically self-limited
  - Topical steroids: keratoderma blennorrhagicum
  - Uveitis: Ophthalmologic Referral
- Articular Disease
  - NSAIDS (foundation of Tx): Indomethacin 150 mg/d
  - Recurrent “chronic” conditions: Intra-articular corticosteroids (SI joints require US guidance)

- Refractory Disease
  - Remission common in 2 – 6 months
  - If recurrence persistence and/flare can used DMARD
    - Sulfasalazine 2 – 3 gm/day** drug of choice
    - MTX, azathioprine, cyclophosphamide (variable success)
    - TNF alpha blocking agents: (variable success)



Rheumatoid Arthritis

Rheumatoid Arthritis

RA	DJD/OA
Inflammatory Joint Dx	Non-Inflammatory Joint Dx
Sym/Bilat MCP & PIP	HIPS, Knees, Spine, & DIP
Soft Tissue Swell & Ulnar Dev	Bony Swelling & Nodes
Rheumatoid Nodules & RF	No Nodules
Erosions, Osteopenia	Sclerosis & Osteophytes
Synovitis, Ankylosis, & Pannus	Reactive Changes: Cysts
Systemic	Local

Rheumatoid Arthritis
Clinical Features

Functional Consequences

Hand

- Swan Neck and Boutonniere Deformity
- Ulnar Deviation

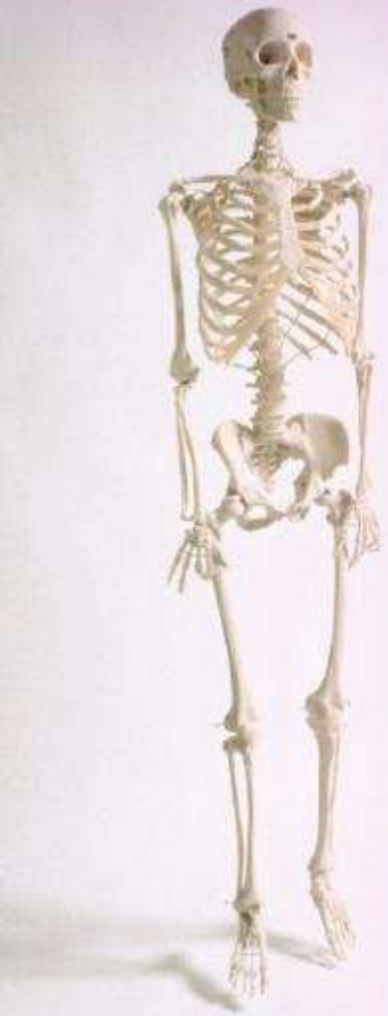
Wrist

- Extensor Tendon Rupture

Rheumatoid Arthritis
Clinical Features

Extra-Articular

- Constitutional
  - Fever, weight loss
- Ocular
  - Sjogren Syndrome
- Vasculitis
  - Finger Infarcts, Leg Ulcers, Stroke
- Respiratory
  - Pleural Effusions, Nodules, ILD
- Cardiac
  - Arrhythmia, Pericardial Effusions, Coronary Vasculitis
- Nodules
- Neurological
  - CTS, Cord Impingement
- Hematological
  - Felty's Syndrome Anemia
    - Seropositive RA/Splenomegaly/Leukopenia (Neutropenia)/Recurrent Infections/Leg Ulcers
- Osteopenia and Osteoporosis
  - 2X increased incidence



A 52-year-old woman presents with a 2 months of pain and stiffness in her hands and fingers, along with malaise and a 5-lb weight loss. On examination, she has swelling and tenderness of her MCP and PIP joints. Lab work reveals the presence of anti-CCP antibodies. What is the most effect treatment to prevent bone erosion?

a. Disease-modifying antirheumatic drugs (DMARDs)
b. High-dose corticosteroids
c. Nonsteroidal anti-inflammatory drugs (NSAIDs)

Rheumatoid Arthritis: Treatment

DMARDS

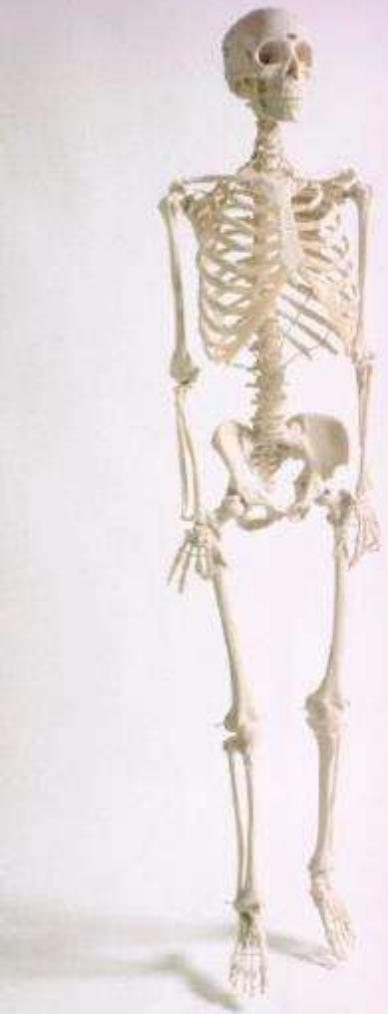
- Methotrexate
- Antimalarials:(hydroxychloroquine) Plaquenil
- Sulfasalazine
- Gold
- Penicillamine
- Minocycline

Biological

- Anti-TNF antibody
  - Etanercept, Infiximab, Adalimumab, certolizumab pegol, Golimumab
- IL-1 blockers
- Monoclonal antibodies against B cells
  - Rituximab
- T cell costimulation blocker
  - Abatacept
  - IL-6
    - Tocilizumab

Methotrexate therapy is to be initiated in a 47-year-old woman for treatment of rheumatoid arthritis. The patient should be advised to do which of the following?

a. Have liver function tests performed every 4 - 8 weeks
b. Have thyroid function tests performed every 4 months
c. Have annual bone marrow examination performed
d. Eat a high-purine diet
e. Avoid intake of magnesium-containing antacids



A 29-year-old women has the acute onset of severe, anterior chest pain that is eased by sitting forward and increased during swallowing and changes in body position. For the past 3 weeks she has had fatigue and generalized arthralgia. Examination shows the presence of a pericardial friction rub. Which of the following is the most likely diagnosis?

a. Pericarditis due to coxsackievirus
b. Pericarditis due to *Staphylococcus* infection
c. Systemic Lupus erythematosus
d. Constrictive pericarditis
e. Superior vena cava syndrome

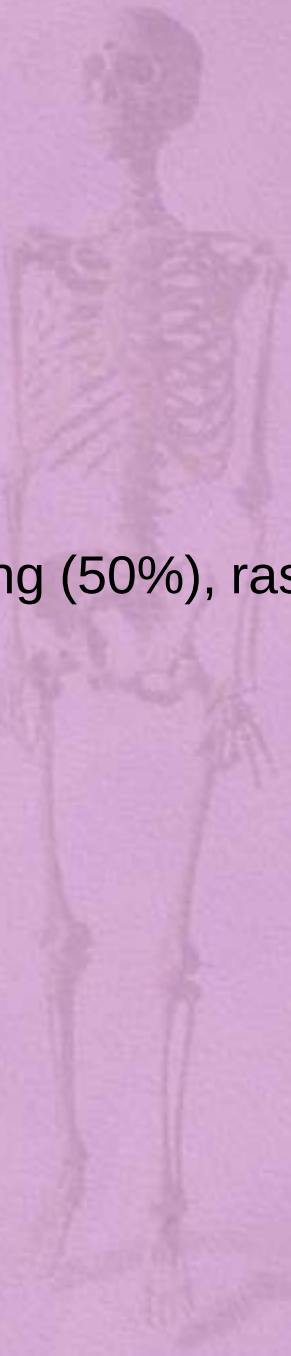
Systemic Lupus Erythematosus

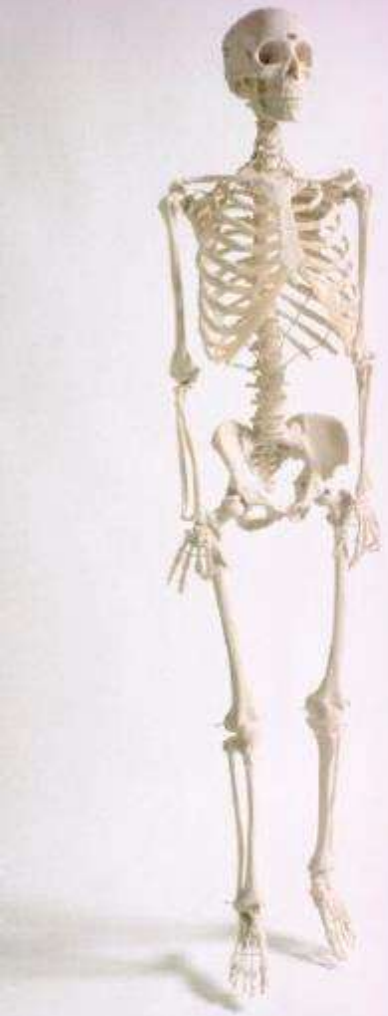
Systemic Lupus Erythematosus:
General Characteristics

- Failure of self tolerance.
  - Involves T cells, IgG autoantibodies, immune complex formation, and Types II and III hypersensitivity reactions.
- Prevalence ratio of women to men: 9:1
- Most common age of onset is from 20-40 y/o.
- Most common in African-American females.

Systemic Lupus Erythematosus:
Clinical Features

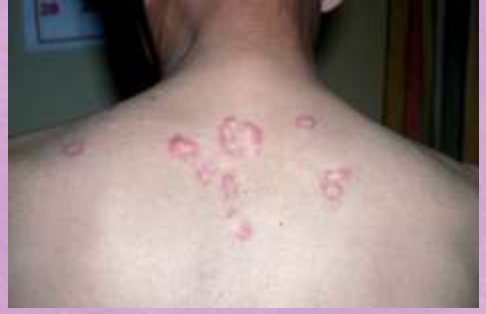
- Initial symptoms
  - Systemic: fever, weight loss and fatigue
  - Most common initial symptoms: joint pain/swelling (50%), rash (20%)
  - Fatigue (10%)
  - Arthralgia: present in 53-95% of patients.
  - Rash: present in 55-90% of patients.
  - Mean length of time to diagnosis is: 5 yrs.

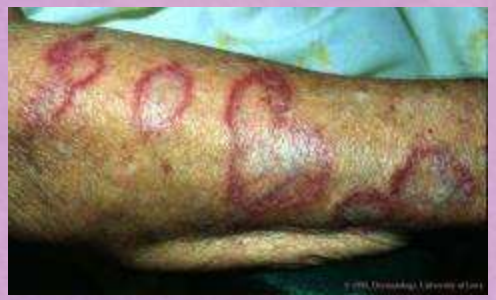




A patient with systemic lupus erythematosus has a rash in malar area and on the extremities. Which of the following is the appropriate counseling to be given to the patient?

a. Apply coal tar and salicylate gel topically on weekly basis
b. Avoid direct sunlight
c. Avoid sunscreen preparations
d. Take weekly cornstarch bath
e. Avoid hydrocortisone topical medications





Skin Manifestations

Malar rash

Discoid rash

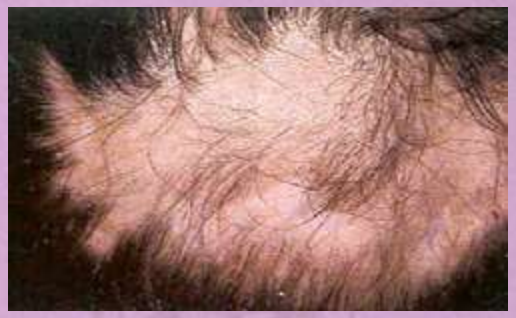
Mucosal ulcerations

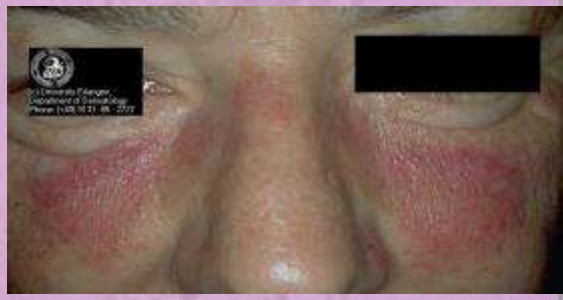
Alopecia

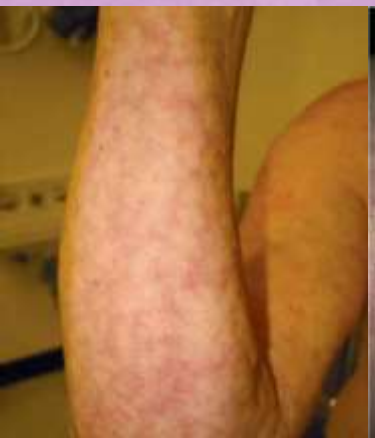
Urticaria

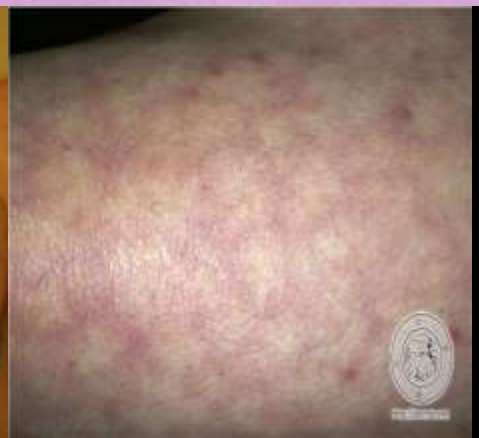
Livedo reticularis

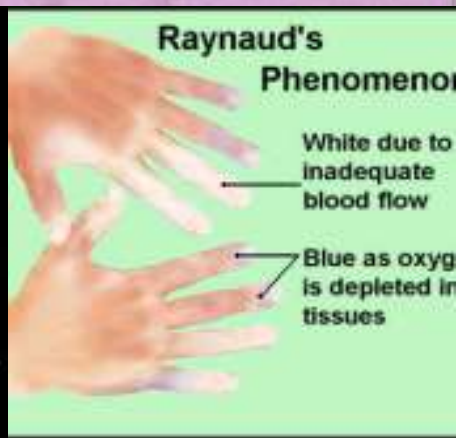
Raynaud's









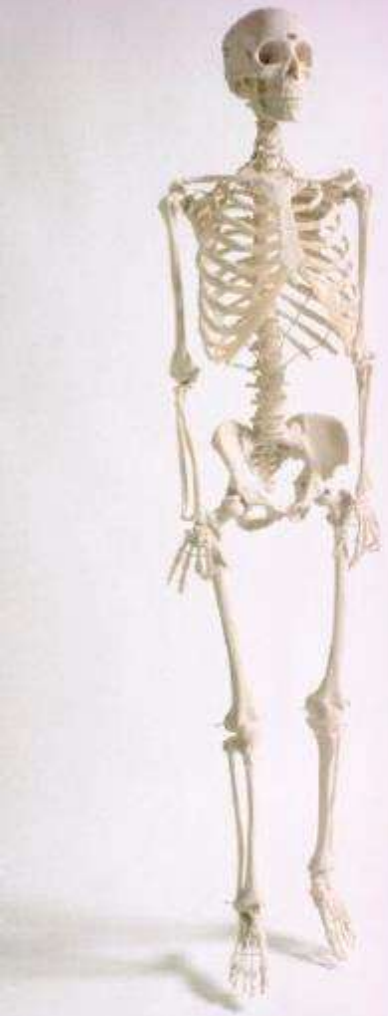


Systemic Lupus Erythematosus:
Laboratory Findings

- ANA: nonspecific, + is not diagnostic, but - virtually r/o dx
  - titre >1:1280 associated w/ renal dx
  - rimmed pattern more specific for SLE
  - sensitive/ but NOT specific
    - 5-15 % of healthy individuals have elevated titres
  - also elevated in:
    - RA, infectious mono, chronic hepatitis, polymyositis.
- Anti ds-DNA (+ in 40-60%)
  - track response to therapy
  - highly specific, esp. nephritis
- Anti Sm (+ in 20-30%)
- Complement: CH50, C3, C4
  - decreases with active disease

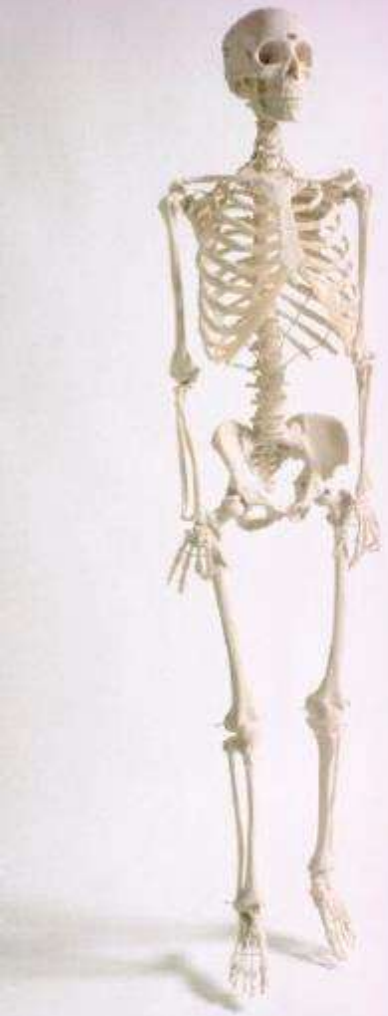
Systemic Lupus Erythematosus:
Treatment

- Plaquenil (hydroxychloroquine): mild disease
- Methotrexate: rash and arthritis
- Corticosteroids: organ threatening disease (heart, lung, kidney, blood, liver)
- Cytoxan (cyclophosphamide): lupus glomerulonephritis
- Cyclosporin:proteinuric nephritis, refractory lupus



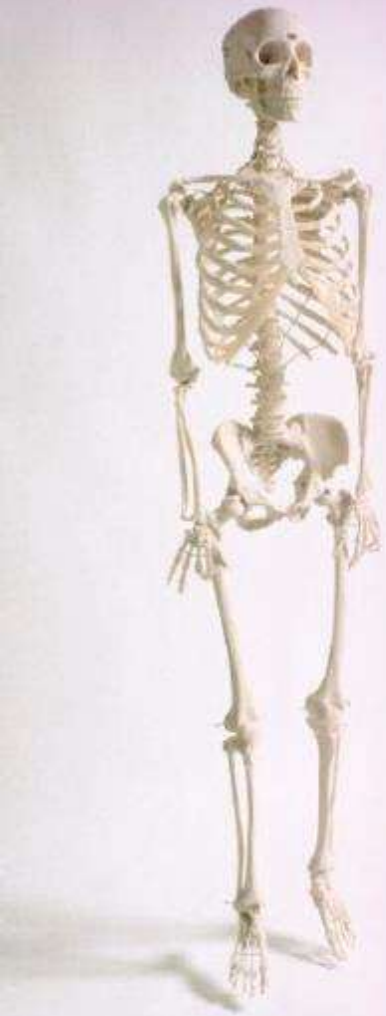
A 26-year-old woman is being placed on hydroxychloroquine (Plaquenil) for systemic lupus erythematous. Which of the following needs to be completed at east annually as part of regular monitoring for medical side effects?

a. Kidney function tests
b. Liver function tests
c. Retinal exams
d. Upper GI endoscopy



A 40-year-old woman has a 2-year history of Raynaud's phenomenon. She now has heartburn and dysphagia to both solids and liquids. Vital signs are normal. Examination shows thickening of the skin of the fingers with loss of creases. Which of the following is the most likely diagnosis?

a. Sjogren's syndrome
b. Achalasia
c. Squamous cell carcinoma of the esophagus
d. Carcinoma of the lung with paraneoplastic syndrome
e. Scleroderma



Scleroderma

Scleroderma:
General Characteristics

- Most prominent feature: thickening or fibrosis of the skin
- F>M = 3:1
- 35 – 64 yr
- Rare in children and in men under 30 yr
- More common in black women during childbearing age otherwise no predominance among blacks

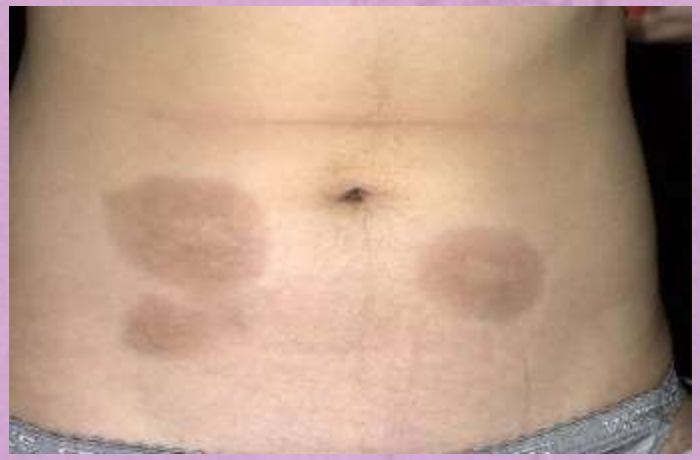
Scleroderma:
Classifications

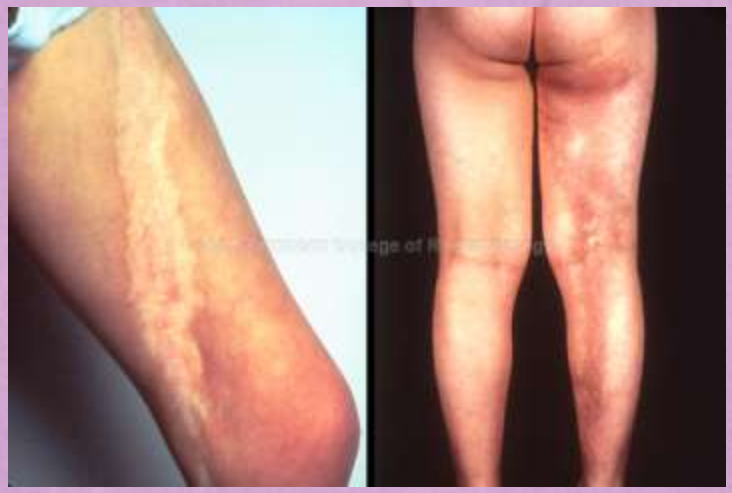
1. Localized Scleroderma

Cutaneous changes consisting of dermal fibrosis without internal organ involvement

a. Morphea: single or multiple plaques commonly on the trunk

b. Linear scleroderma: bands of skin thickening commonly on the legs or arms (typically follow a linear path)



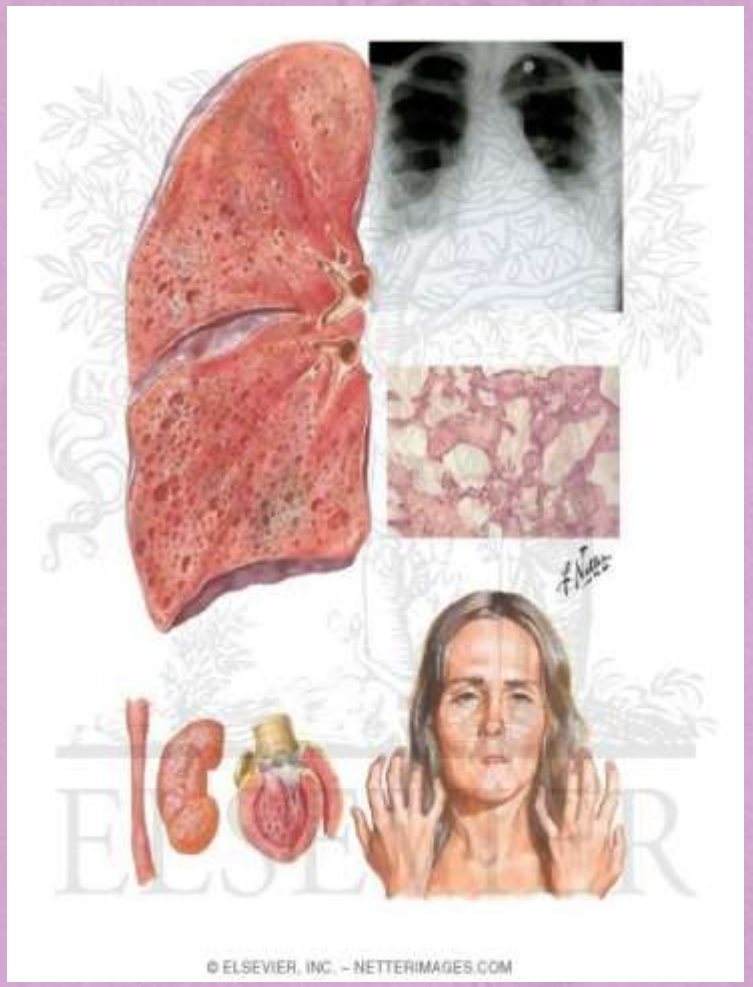


Scleroderma:
Classifications

2. Systemic Sclerosis

a. Diffuse system sclerosis: fibrotic skin proximal to the elbows or knees excluding the faces and neck

- May have onset of Raynaud's within a year of developing disease
- More likely to have pulmonary, renal, or cardiac involvement.
- More likely to have a positive ant-Scl-70 antibody
- Less likely to have an anti-centromere antibody



Scleroderma:
Classifications

2. Systemic Sclerosis

b. Limited Systemic Sclerosis

Fibrotic skin limited to the hands and forearms, feet, neck and face

- Usually Raynaud's for years, may have telangiectasias, skin calcifications, & late incidence of pulmonary hypertension.
- High incidence of anti-centromere antibody

CREST Syndrome

Subgroup of patients with <u>limited systemic sclerosis</u>:

C – **C**alcinosis

R – **R**aynaud's phenomenon

E – **E**sophageal dysmotility

S – **S**clerodactyly

T – **T**elangiectasias

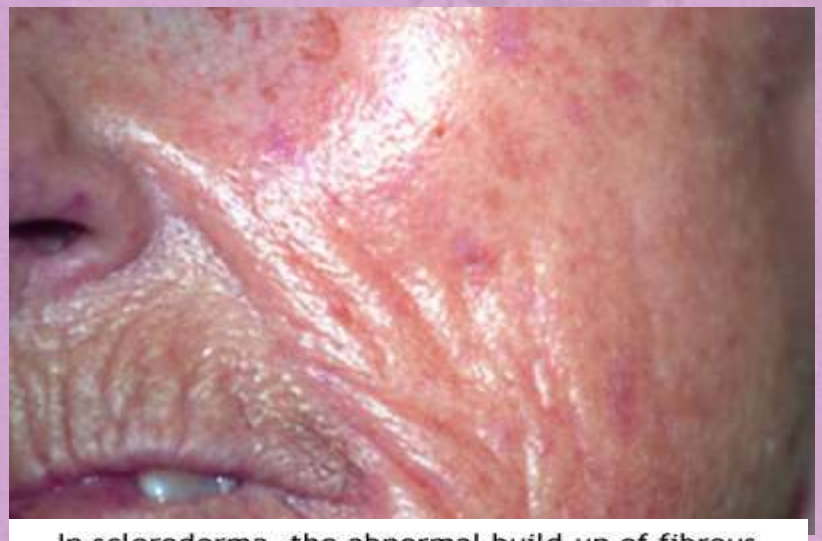
3. Overlap Syndrome: Scleroderma associated with other autoimmune diseases

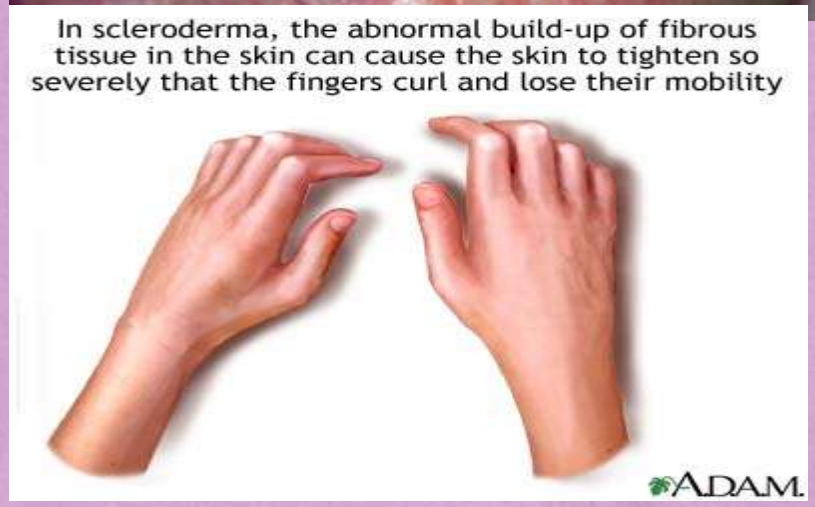
Scleroderma:
Clinical Features

Thickened skin (Hallmark)

- Loss of sweat glands & hair loss in areas of tight skin

- Begins on fingers & hands in all cases of systemic sclerosis
  - If begins elsewhere consider a localized form of scleroderma

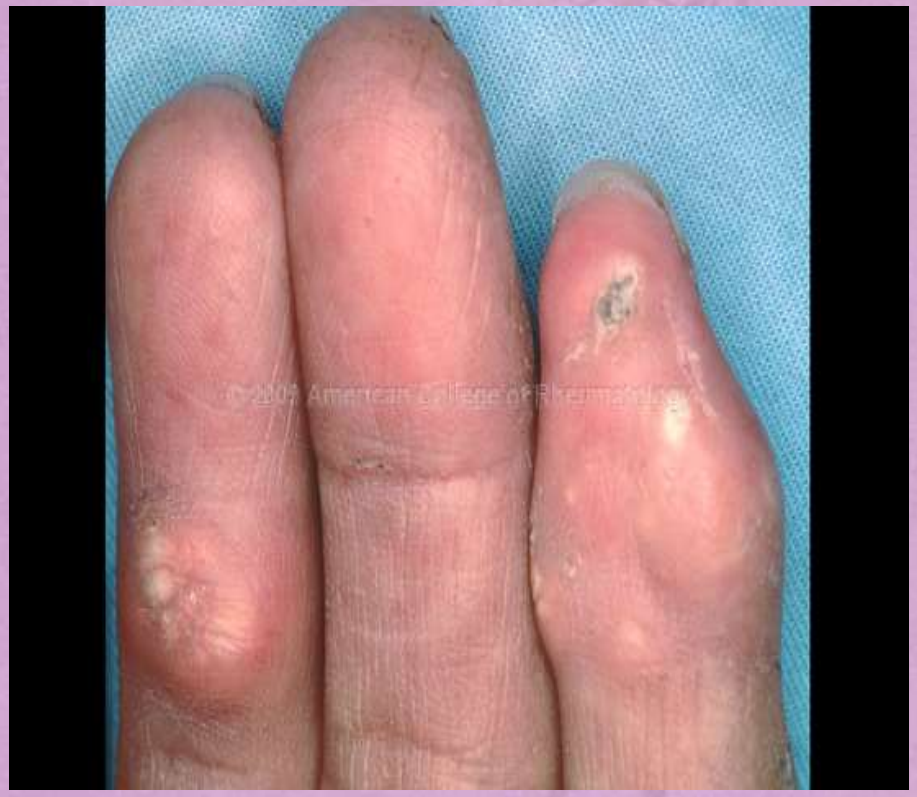




Scleroderma:
Clinical Features

Calcinosis

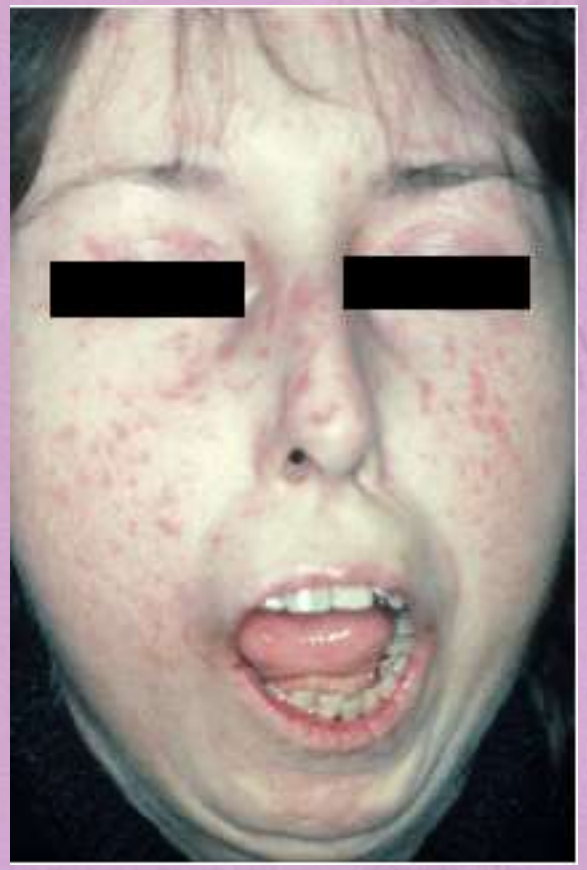
- Cutaneous deposits of calcium phosphate
- Hands, (PIP joints and fingertips), periarticular tissue and bony prominences
- Firm, non-tender, 1mm – several cm.
- Can become inflamed infected or ulcerated



Scleroderma:
Clinical Features

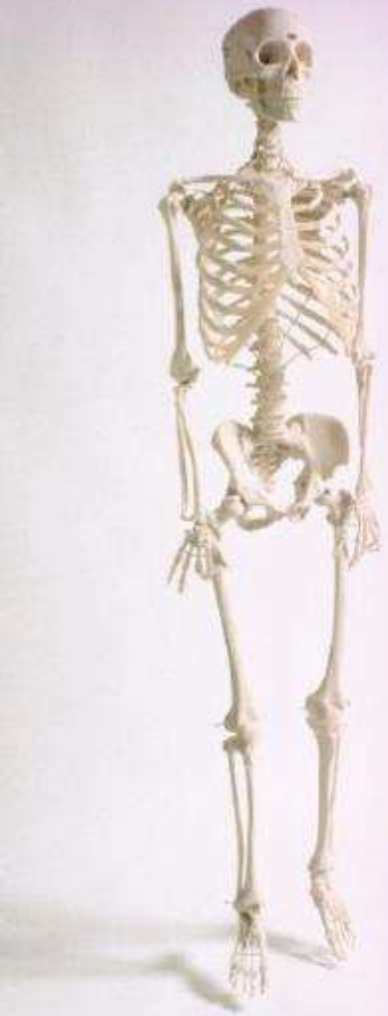
Telangiectases

- Dilated venules, capillaries, and arterioles
- Hands face lips and oral mucosa
- More common in limited systemic sclerosis
- Can occur in GI mucosa (Watermelon stomach) causing blood loss and anemia



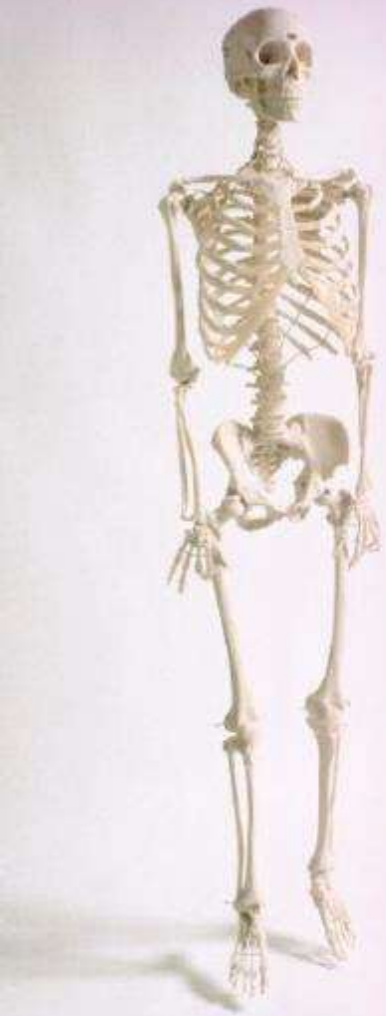
Scleroderma:
Treatment

- No treatment with demonstrated success
  - D-Penicillamine (most studied drug)
  - Following tried in small studies
    - Colchicine, Chlorambucil
    - Corticosteroids:
      - May increase the incidence of renal crisis (should be avoided if possible)
      - Used for comfort in patients with very actively inflamed and tight skin



A 42-year-old woman reports with dry eyes and dry mouth for the past 2 to 3 months. She states she often feels she has “cotton mouth” and her eyes are always itching and burning. Which of the following exam findings is found in one out of three patients with the suspected disorder?

a. Loss of normal skin folds
b. Malar butterfly rash
c. Parotid enlargement
d. Progressive proximal muscle weakness



Sjogren's Syndrome

Sjogren's Syndrome:
General Characteristics

- F>M: 9:1

- Mean age at diagnosis: 50
- Bimodal
  - Childbearing age: mid 30's
  - Post menopausal: mid 50's

- Takes ~ 9 years for Sjogren's patients to be diagnosed even after they develop sicca symptoms.

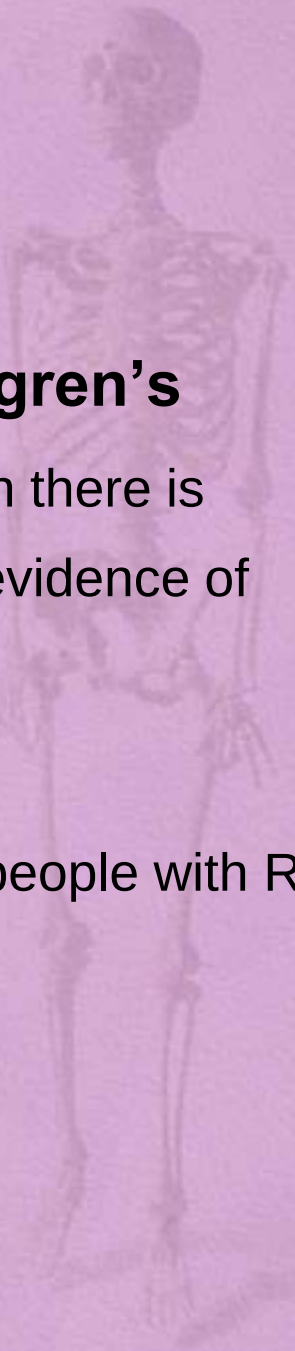
Sjogren's Syndrome:
General Characteristics

Primary Sjogren's

- Absence of another underlying rheumatic disease
- Associated with ANA antibodies to Ro/SSA and La/SSB
- Occurs at rate of 1/1000

Secondary Sjogren's

- Diagnosed when there is accompanying evidence of another CTD
- Occurs in 30% people with RA



Sjogren's Syndrome:
Clinical Features

- Xerophthalmia: 47%
- Xerostomia: 42%
- Arthralgias/Arthritis:28%
- Parotid gland enlargement: 24%
- Raynaud's phenomenon: 21%
- Fever/Fatigue: 10%
- Dyspareunia: 5%

Sjogren's Syndrome:
Clinical Features

Extraglandular Manifestations

Respiratory
- Chronic bronchitis
- Interstitial Pneumonitis

Gastrointestinal
- GERD
- Dysphagia
- Atrophic gastritis

Skin and Mucous Membranes
- Dryness
- Raynaud's phenomenon (20%)

Hematologic
- Neutropenia
- Anemia
- Thrombocytopenia

Neurologic
- Central or peripheral neuropathy
- Depression

Musculoskeletal (60%)
- Myalgia (20 – 30%)
- Arthralgia (60 – 70%)
- Fatigue (60%)

Low grade fever

Lymphatic
- Lymphadenopathy (20%)
- Lymphoma

Renal Involvement
- Interstitial nephritis
- Glomerulonephritis

Sjogren's Syndrome:
Laboratory Findings

- Elevated ESR
- Hypergammaglobulinemia
- Anemia of chronic disease
- Leukopenia
- Thrombocytopenia Rare
- Autoantibodies
  - RF (80 – 95%), ANA (90%), SSA (50 –90%), SSB(50 – 90%)
- Salivary Gland Biopsy
  - demonstrate lymphocytic infiltration

Sjogren's Syndrome: Treatment

Xerophthalmia

- Preservative-free artificial tears (Refresh)
- Lubricant ointments (Refresh PM, Lacrilube)
- Humidifiers
- Punctal occlusion (plugs)

Mucosal Dryness

- Dental Care
  - Fluoridated Toothpaste/Mouthwash
- Cholinergic drugs
  - Salagen (pilocarpine) 5 mg QID
- Treatment of oral candidiasis
  - Nystatin
  - Oral fluconazole for 2 weeks in refractory cases

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IT'S A QUESTION PARTY!
MY FAVORITE!