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شعبة الإمارات لطب الأسرة

Rheumatology

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Lecture Modules

- Systemic Lupus Erythematosus
- Progressive Systemic Sclerosis/scleroderma
- Dermatomyositis/Polymyositis
- Proximal myopathies
- RA vs OA

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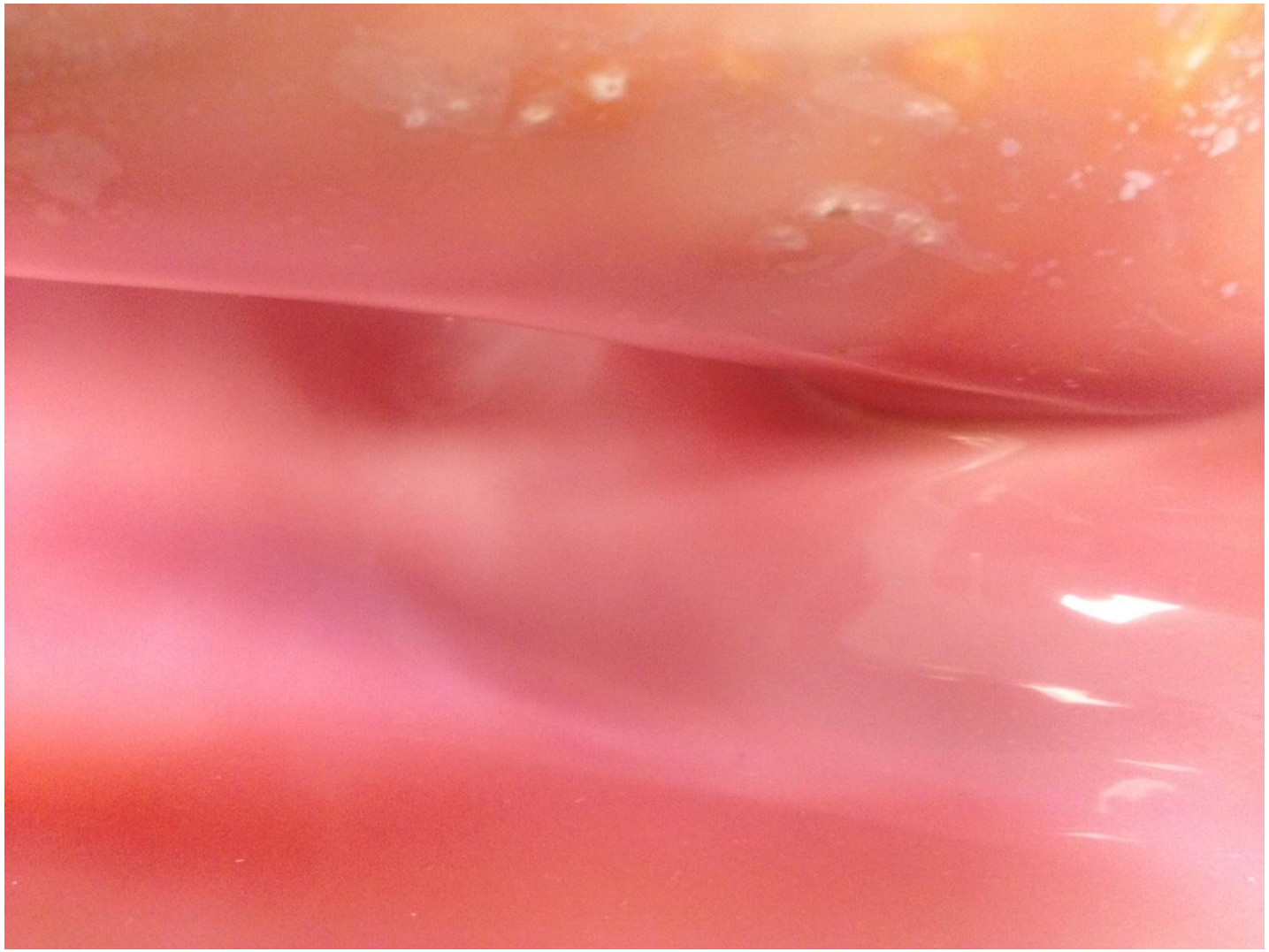
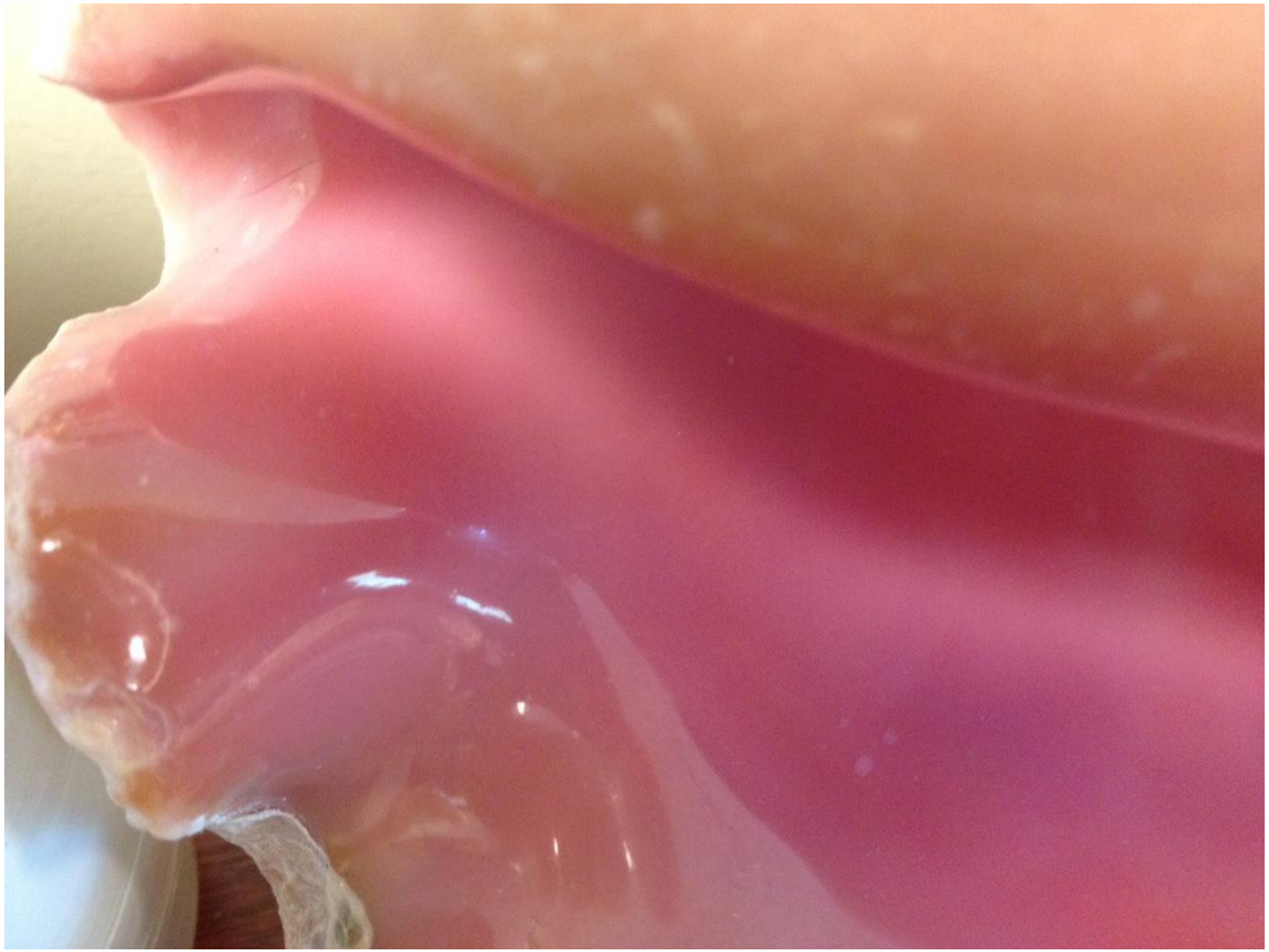
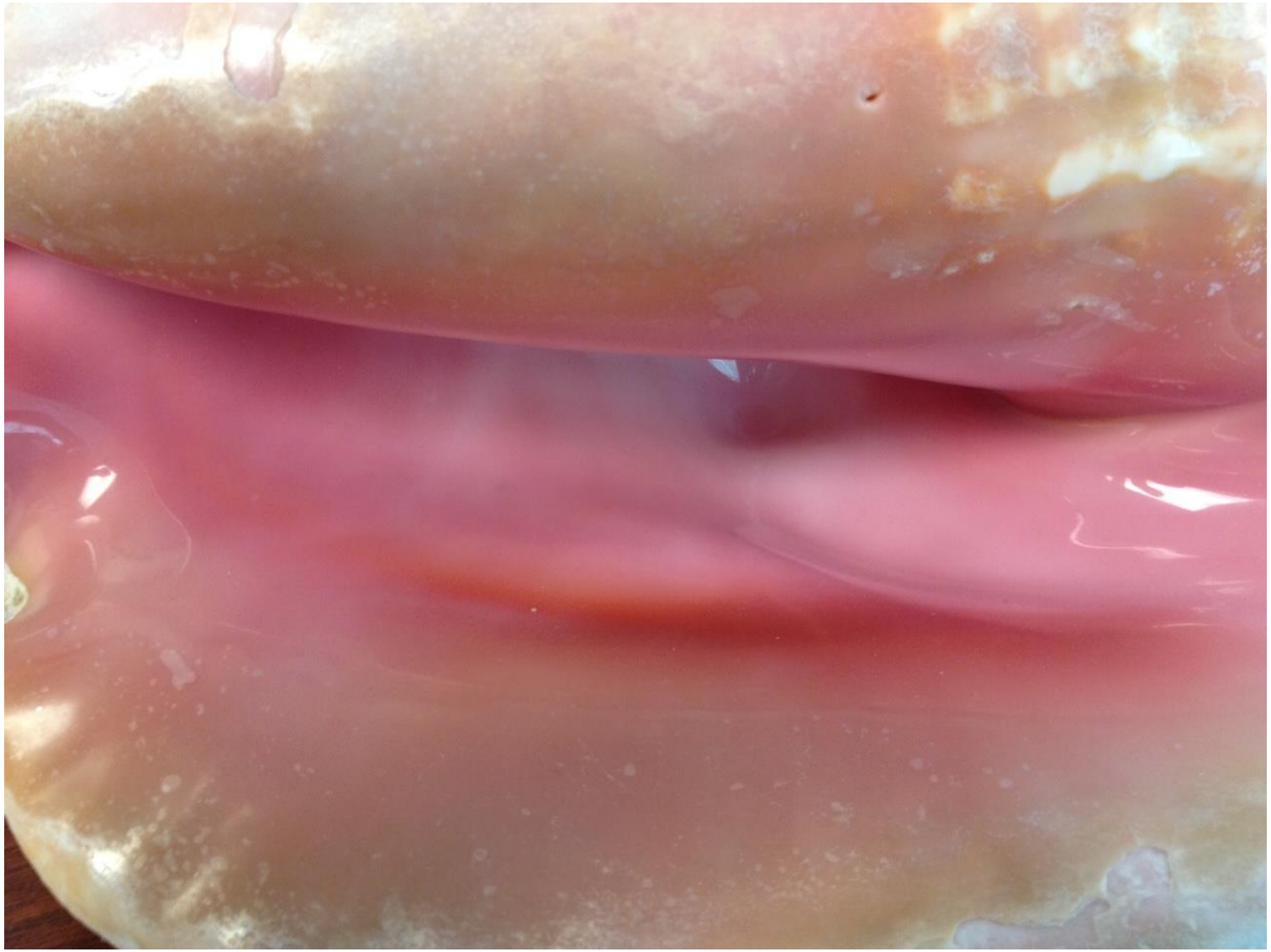
General Comments re: CTD (Connective Tissue Diseases)

- Pattern recognition is invaluable in assessing for potential CTDs
- Women are affected much more frequently than men for almost all CTDs
- Most CTD patients should be managed with the help of a rheumatologist when prescribing immunomodulators or biologic agents







Patient Case

- 37 yo female presents with progressive shortness of breath.
- Has given birth to 5 healthy children previously without difficulty.
- Other complaints include:
  - Knee and elbow pain
  - Patchy hair loss

Patient Case

- Labs of note:
  - ANA (+) at 1:320
  - Platelet count of 96
  - WBC 3.5
  - Lymphocyte count of 1200
- Exam notable for bilateral pleural effusions

37 yo female with dyspnea, arthralgias, (+) ANA, mild pancytopenia, and pleural effusion. What is the most likely diagnosis?

A. Rheumatoid arthritis
B. Systemic lupus erythematosus
C. Wegener's granulomatosis
D. Sarcoidosis

Systemic Lupus Erythematosus (SLE)

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- SLE is a systemic inflammatory autoimmune disease with protean manifestations.
- Female: male incidence of 9:1
- Typically affects women of child bearing years
- US prevalence of 0.1% (1 per 1000)
- More prevalent and severe among blacks and Hispanics

SLE

- Estimated survival rates of 96% at 5 years, 93% at 10 years, and 78% at 15-20 years.
  - Previously, 50% at 5 years in the 1950's
- Major causes of mortality is premature coronary artery disease with a three fold increase over general population.
- Increased risk of osteoporosis
  - From disease and drugs (steroids)
  - 3 months of prednisone 5mg/day or more → DEXA

SLE

- The diagnosis is clinical
- (+) ANA – not always helpful
- Must have 4 of 11 diagnostic criteria
- Lab tests cannot substitute for pattern recognition in the history and physical by the physician!

SLE Diagnostic Criteria

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- Mucocutaneous signs – 4 of them
- Brain – seizures, psychosis
- Lab – (+) ANA
- Lab – other immune criteria
- Arthritis – nonerosive (vs Rheumatoid Arthritis)
- Serositis
- Hematologic disturbances
- Renal disease

Lab

- (+) ANA
  - 90% of patients with SLE have a (+) ANA
- Negative at < 1:40 serum dilutions
- Indeterminate at 1:40 – 1:160
- Clearly positive at > 1:320

Lab

- False (+) RPR
- Prolonged PTT (Lupus anticoagulant)
- (+) Anti-Smith antibodies
- (+) Anti-double stranded DNA antibodies
- (+) Anticardiolipin antibodies
- (+) Lupus erythematosus cell prep

Renal involvement

- Proteinuria > 0.5 grams/day
- > 3+ protein on dipstick
- Cellular casts indicative of renal disease
  - RBC casts suggest glomerulonephritis
  - Tubular casts suggest inflammation of the tubules

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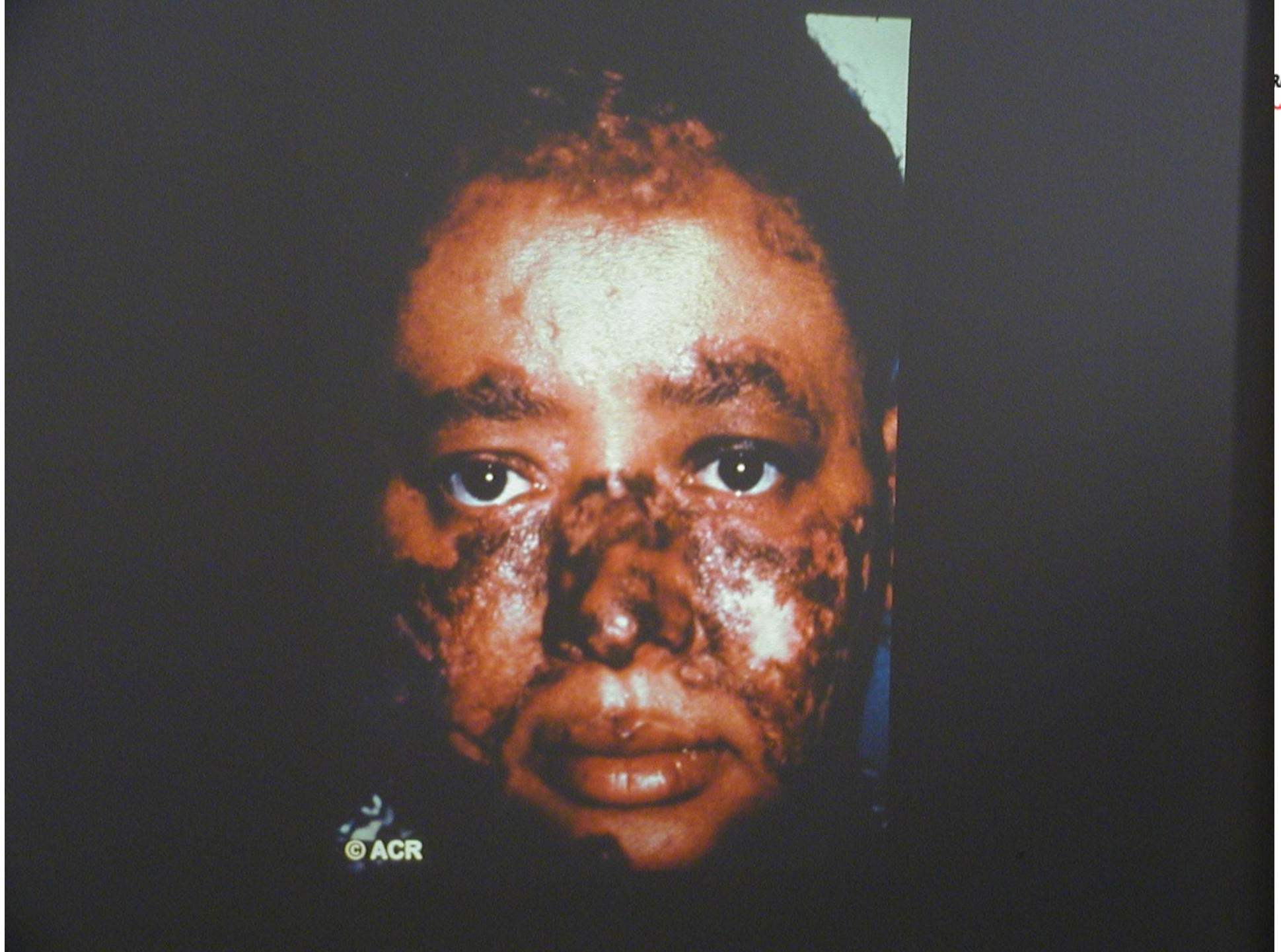
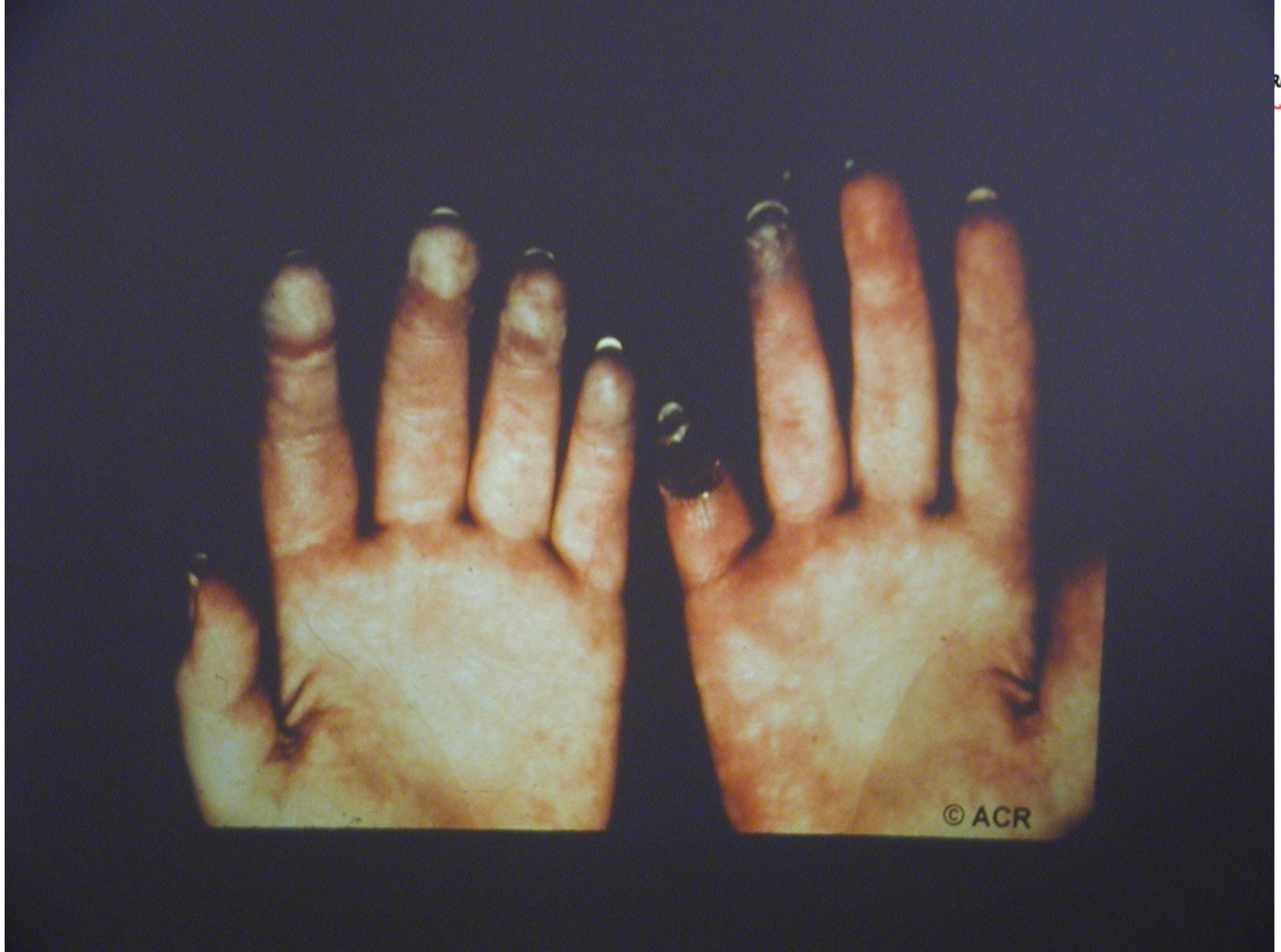
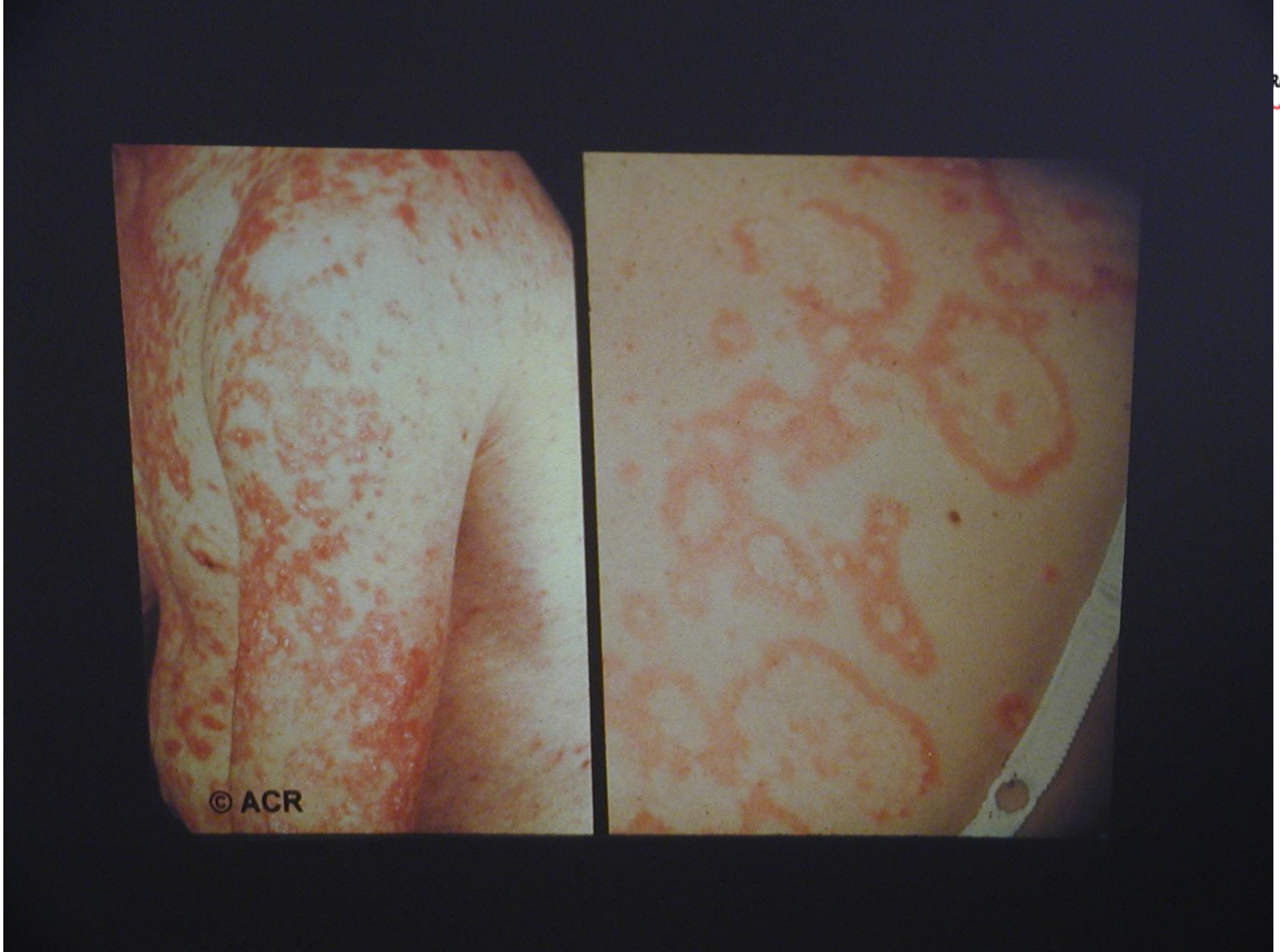
Clinical Manifestations of SLE

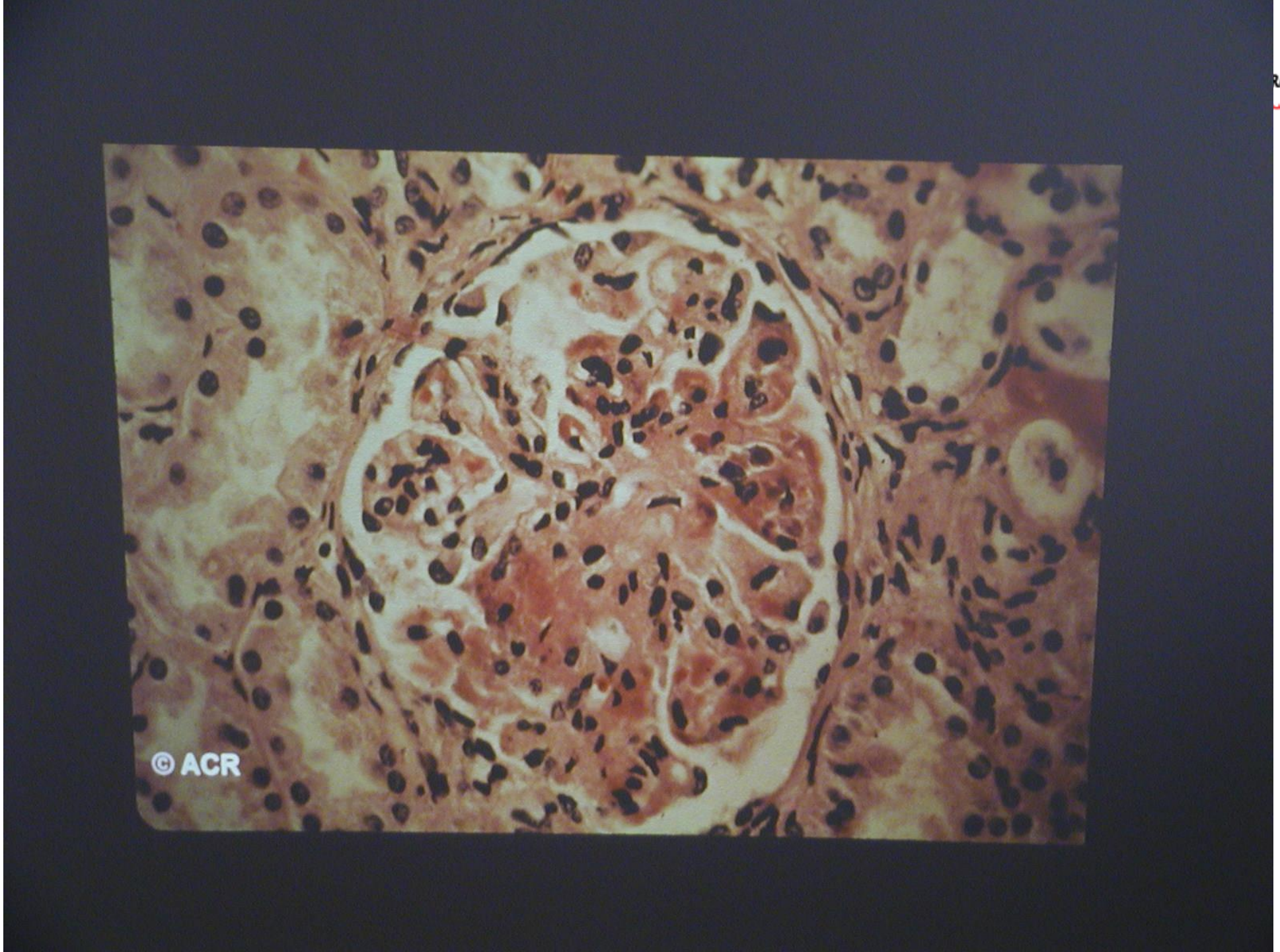
American College of Rheumatology

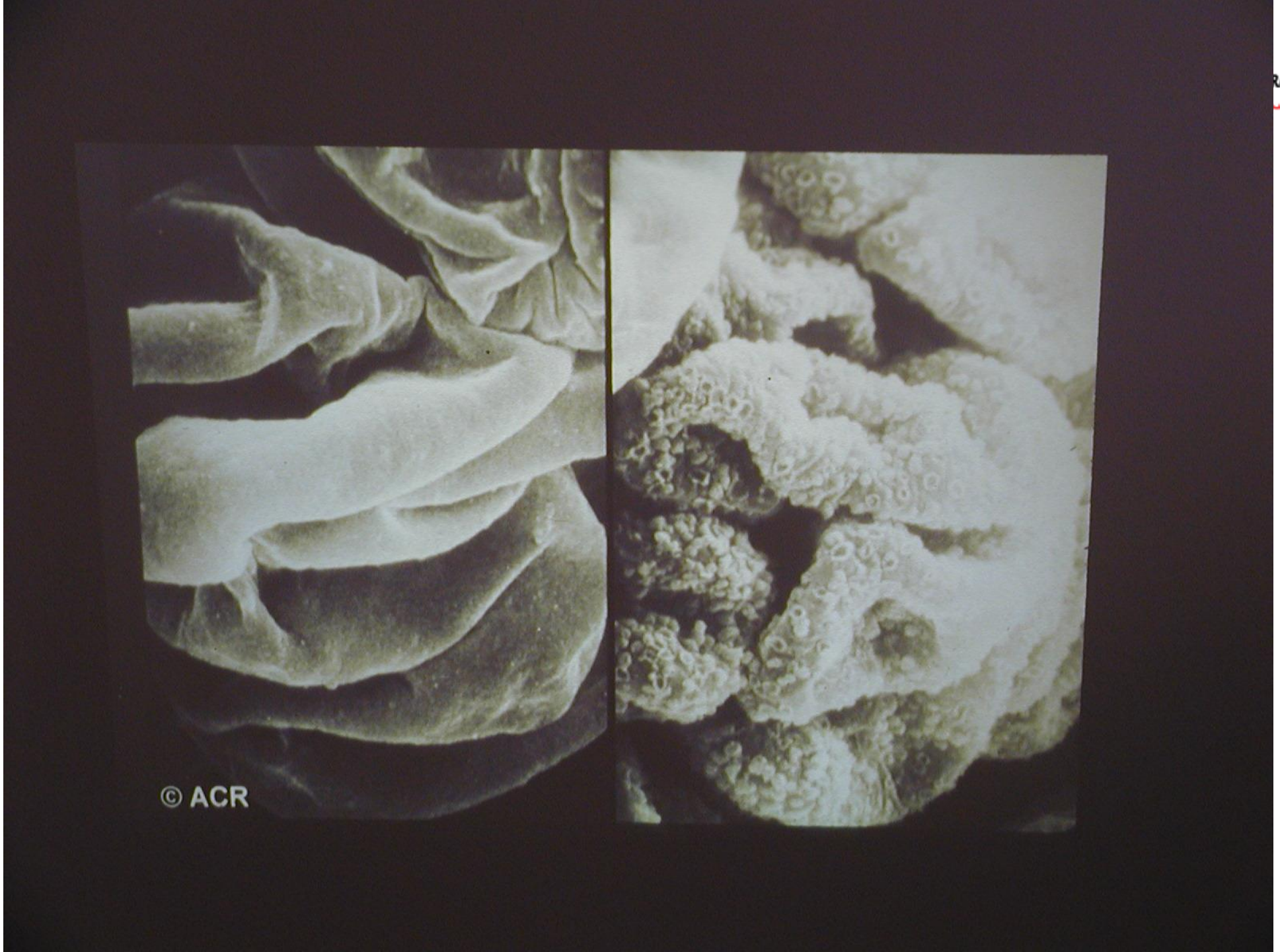
Slide Set





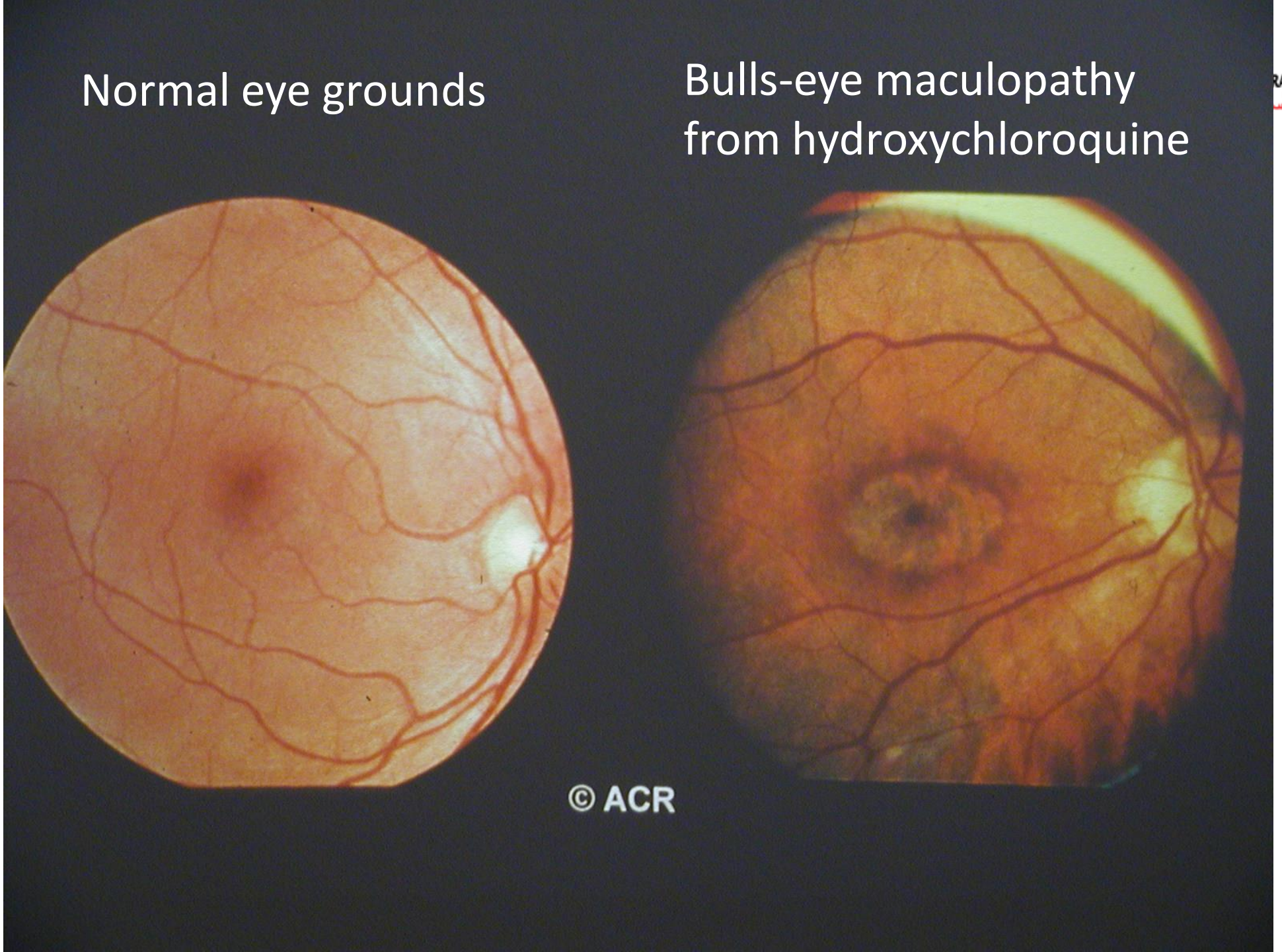






Normal eye grounds

Bulls-eye maculopathy from hydroxychloroquine



SLE Treatment

- NSAIDs
  - Can be used early in SLE treatment
  - Effective for arthralgias and serositis
  - Avoid with lupus nephritis and renal insufficiency
  - Avoid Cox-2 inhibitors in patients with an increased risk of CVD

SLE treatment

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- Steroids
  - High dose steroids can be used to manage severe disease episodes
  - May serve as bridge therapy until slow-acting drugs become effective
  - Low dose (5mg daily) can be used in the treatment of mild SLE
  - Topical steroids for localized skin manifestations
  - Intra-articular steroids for joint disease
    - Rule out septic arthritis first

SLE treatment

- Hydroxychloroquine
  - All SLE pts should be on hydroxychloroquine unless not tolerated
  - Requires 6-12 weeks to show benefit.
  - Can be combined with NSAIDs for mild disease
    - Cutaneous manifestations and arthralgias

The Canadian Hydroxychloroquine Study Group: A randomized study of the effect of withdrawing hydroxychloroquine sulfate in systemic lupus erythematosus. N Engl J Med. 1991;324(3):150-154

SLE treatment - Other immunomodulatory drugs

- Methotrexate – some benefit
- Mycophenolate mofetil (Cellcept)
- Cyclophosphamide
  - Historically used for lupus nephritis
- Biologics – belimumab and rituxamb (off label)

Fanouriakis A, Kostopoulou M, Alunno A, et al. 2019 update of the EULAR recommendations for the management of systemic lupus erythematosus Ann Rheum Dis. 2019;78(6):736. Epub 2019 Mar 29.

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Which of the following is not a cause of an elevated ESR?

A. Acute gout flare
B. SLE/Lupus
C. Polymyalgia rheumatic
D. Fibromyalgia

Etiologies of elevated ESR > 100mm/hour

- Acute gout
- Polymyalgia rheumatica/temporal arteritis
- Rheumatoid arthritis
- SLE/active CTDs
- Infections:
  - Osteomyelitis, Subacute bacterial endocarditits, deep tissue abscess
- Cancer:
  - Carcinoma, leukemia, lymphoma, multiple myeloma

Zacharski LR, Kyle RA. Significance of extreme elevation of erythrocyte sedimentation rate. JAMA. 1967;202(4):264-266.

A 47 yo female presents with progressive dyspnea and edema. Exam notable with tight skin over hands bilateral (picture below) and face/cheeks. Labs show ANA (+) with nucleolar pattern. PFTs show restrictive lung disease. Which of the following is the most likely cause?

A. Scleroderma
B. Dermatomyositis
C. Rheumatoid arthritis
D. Raynaud's phenomenon

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Scleroderma
(Progressive Systemic Sclerosis)

Scleroderma

- A chronic condition characterized by fibrosis of the skin and internal organs
- Raynaud's phenomenon is present in most patients at some stage of the disease
- Prevalence between 20-250 patients per 10^6
- Women have a 5 fold increased risk
- Survival of 78% at 5 years, 55% at 10 years, 37% at 15 years, and 27% at 20 years
- 60% of patients die from pulmonary disease

Scleroderma

- Major criterion is symmetric sclerosis
  - Skin thickening
- Minor criteria
  - Sclerodactyly – thickening/tightening of the fingers
  - Digital pitting or loss of finger tip pad substance
  - Bilateral basilar pulmonary fibrosis

CREST syndrome

- Calcinosis
- Raynaud’s phenomenon
- Esophageal dysmotility
- Sclerodactyly
- Telangiectasia

Lab testing

- Most patients have a (+) ANA
- Nucleolar pattern is present in 30%
- Anti-topoisomerase-1 Ab (Scl-70) are associated with diffuse scleroderma
  - Present in 40% of patients
- Anti-centromere Ab are present in 75% of patients with limited scleroderma and CREST

Routine monitoring every 6 months

- Complete blood count
- Creatinine level
- ESR
- Urinalysis
- ECG
- Echocardiogram (for right heart failure/pulmonary HTN)
- PFTs: +/- DLCO to check for fibrosis

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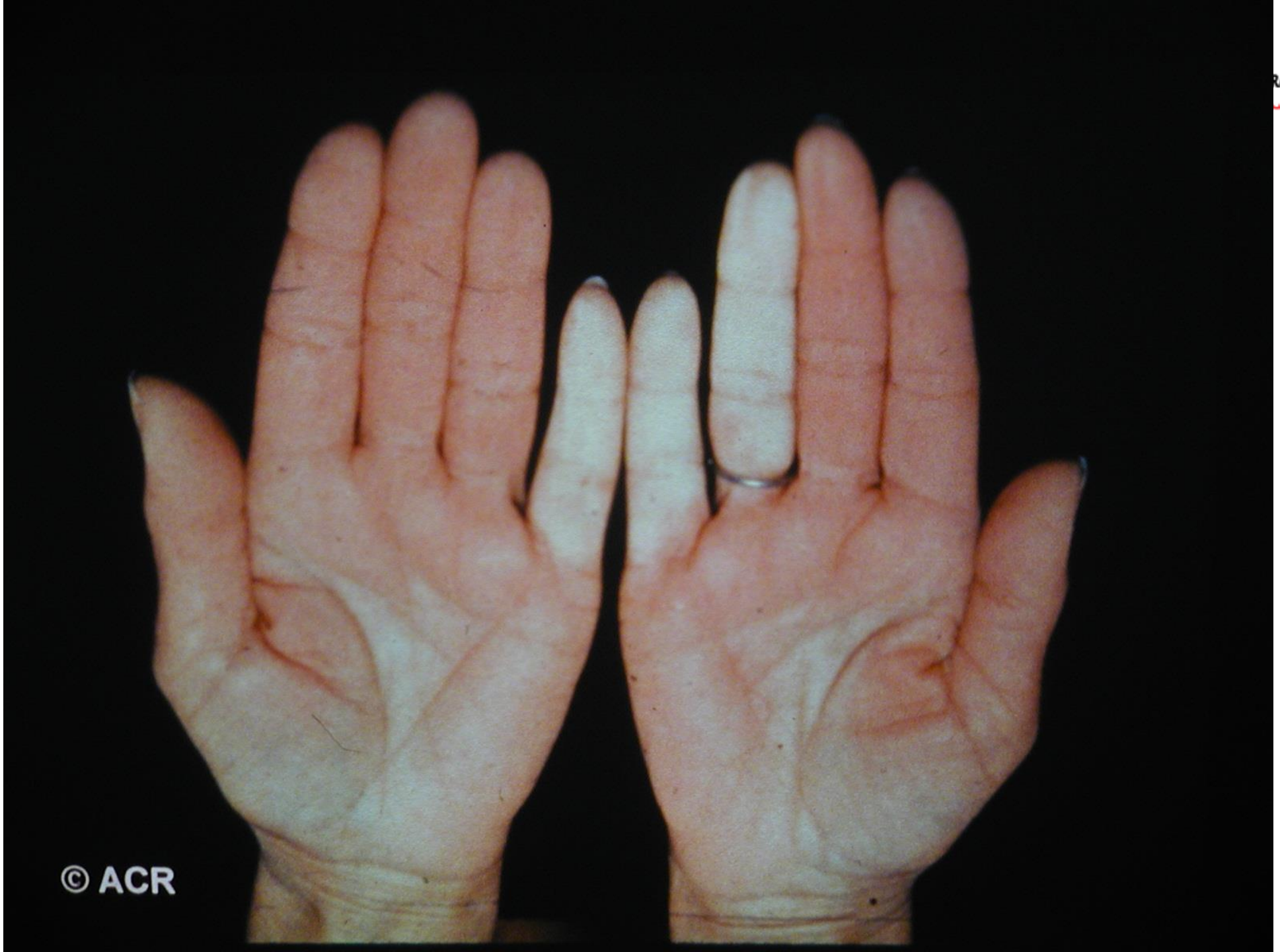
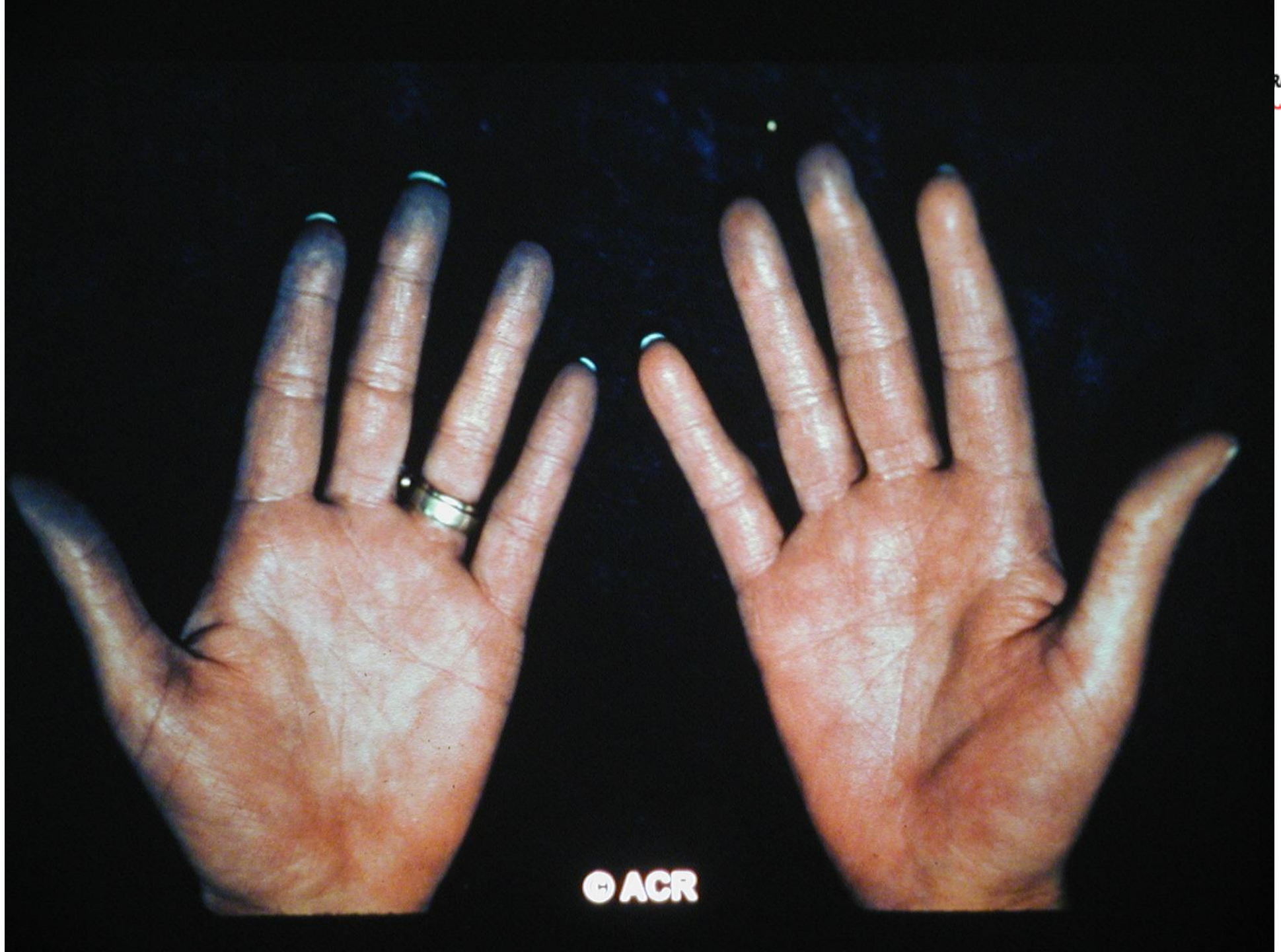
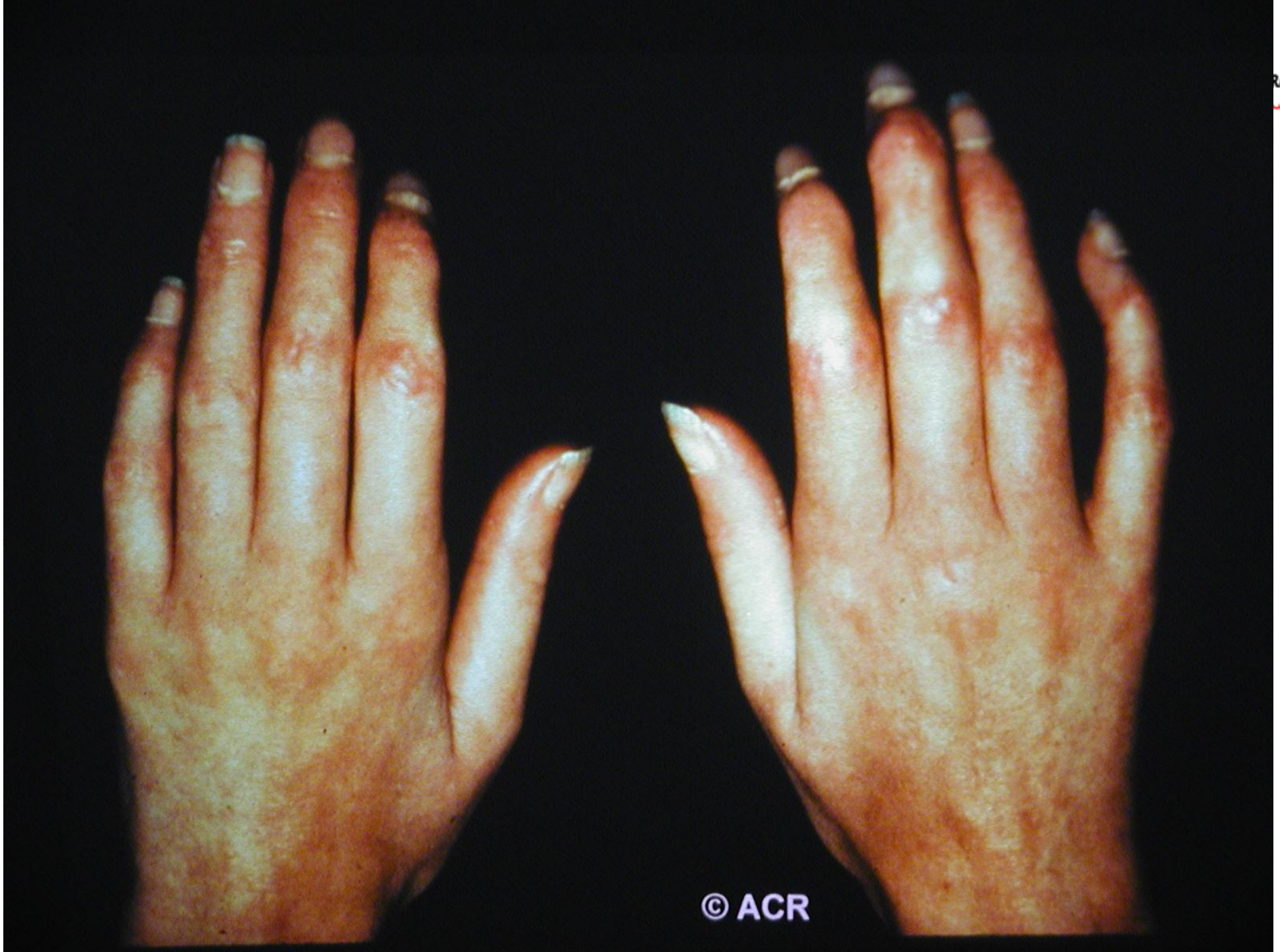
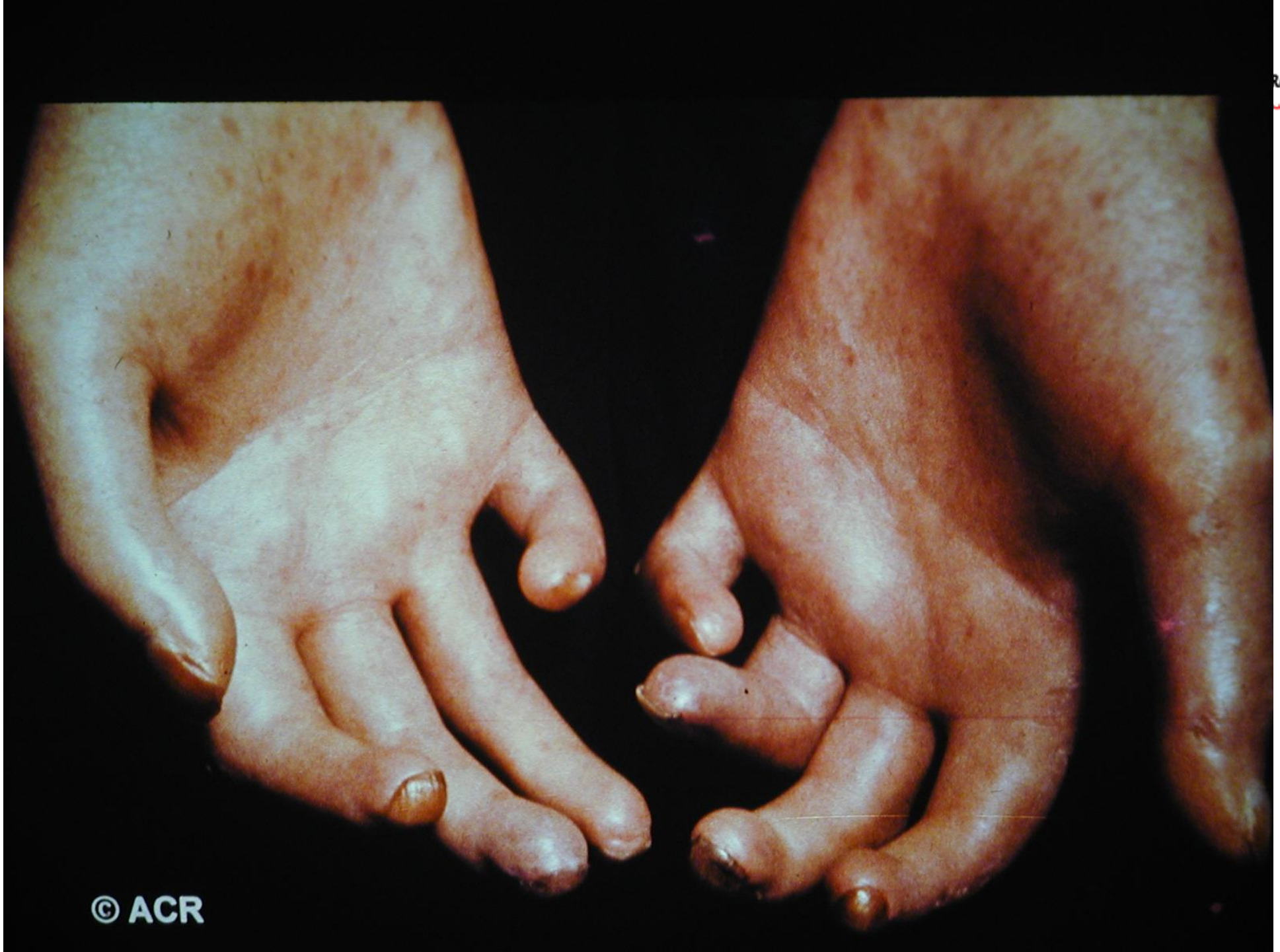
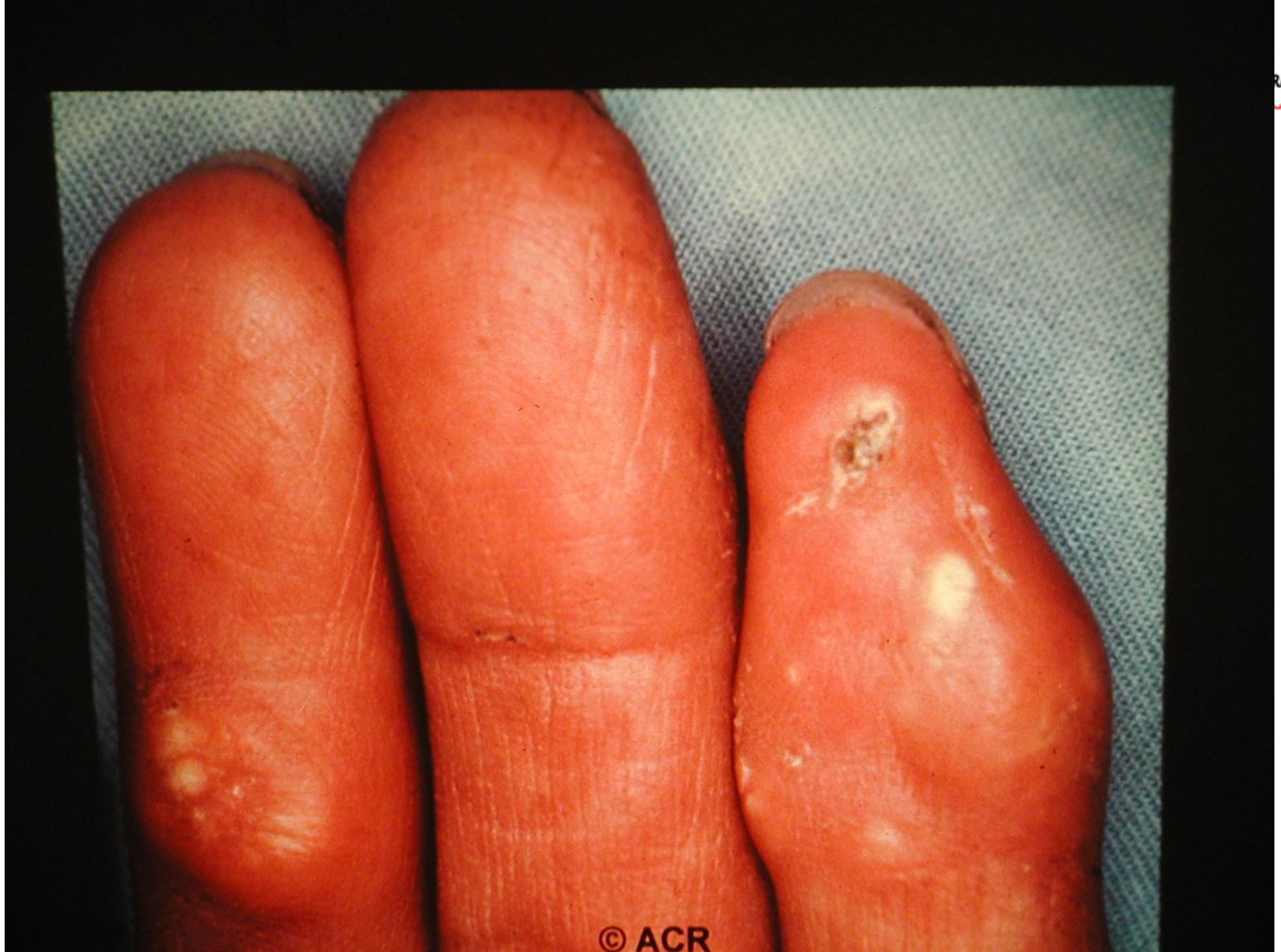
Clinical manifestations of Scleroderma



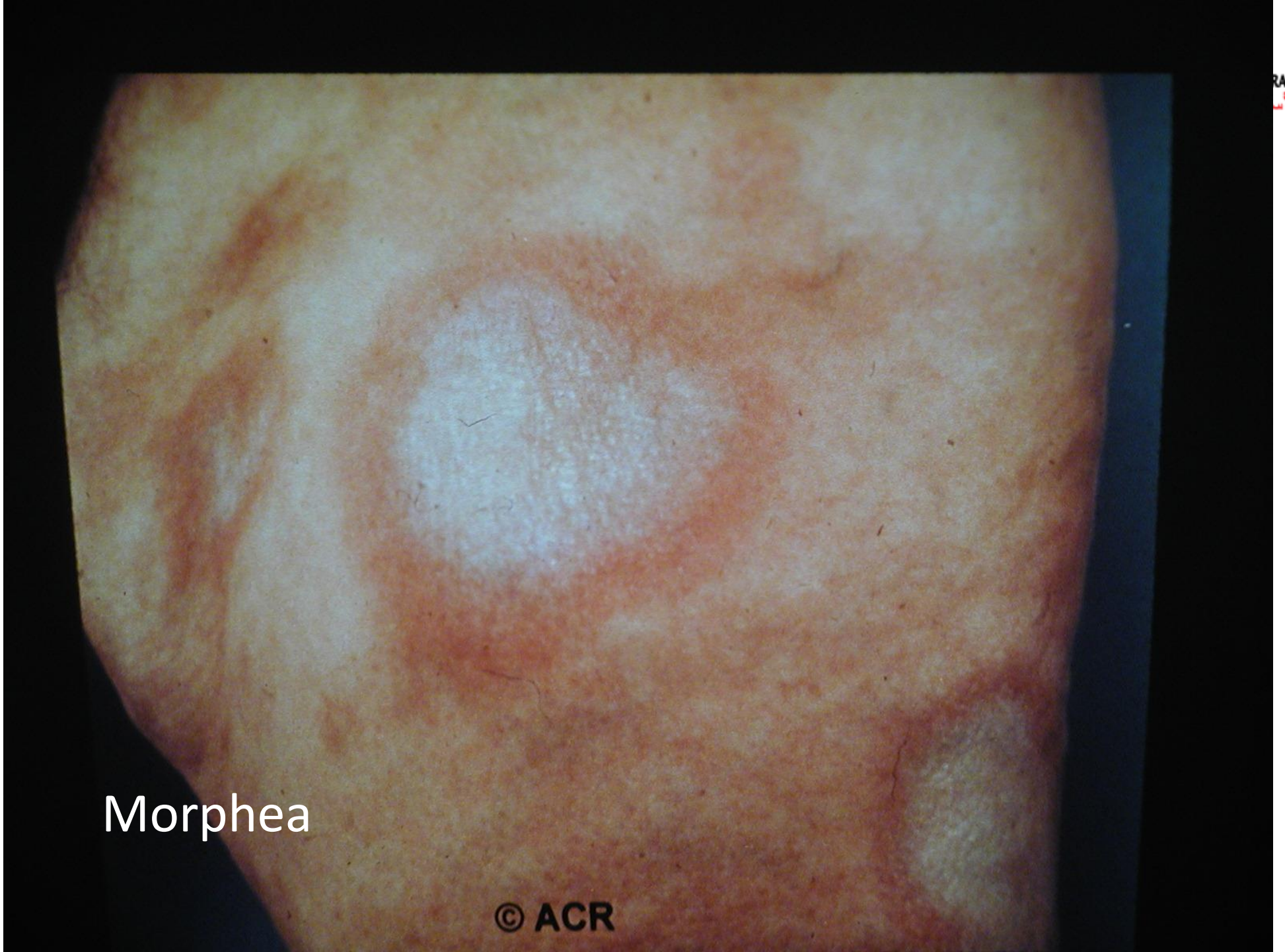




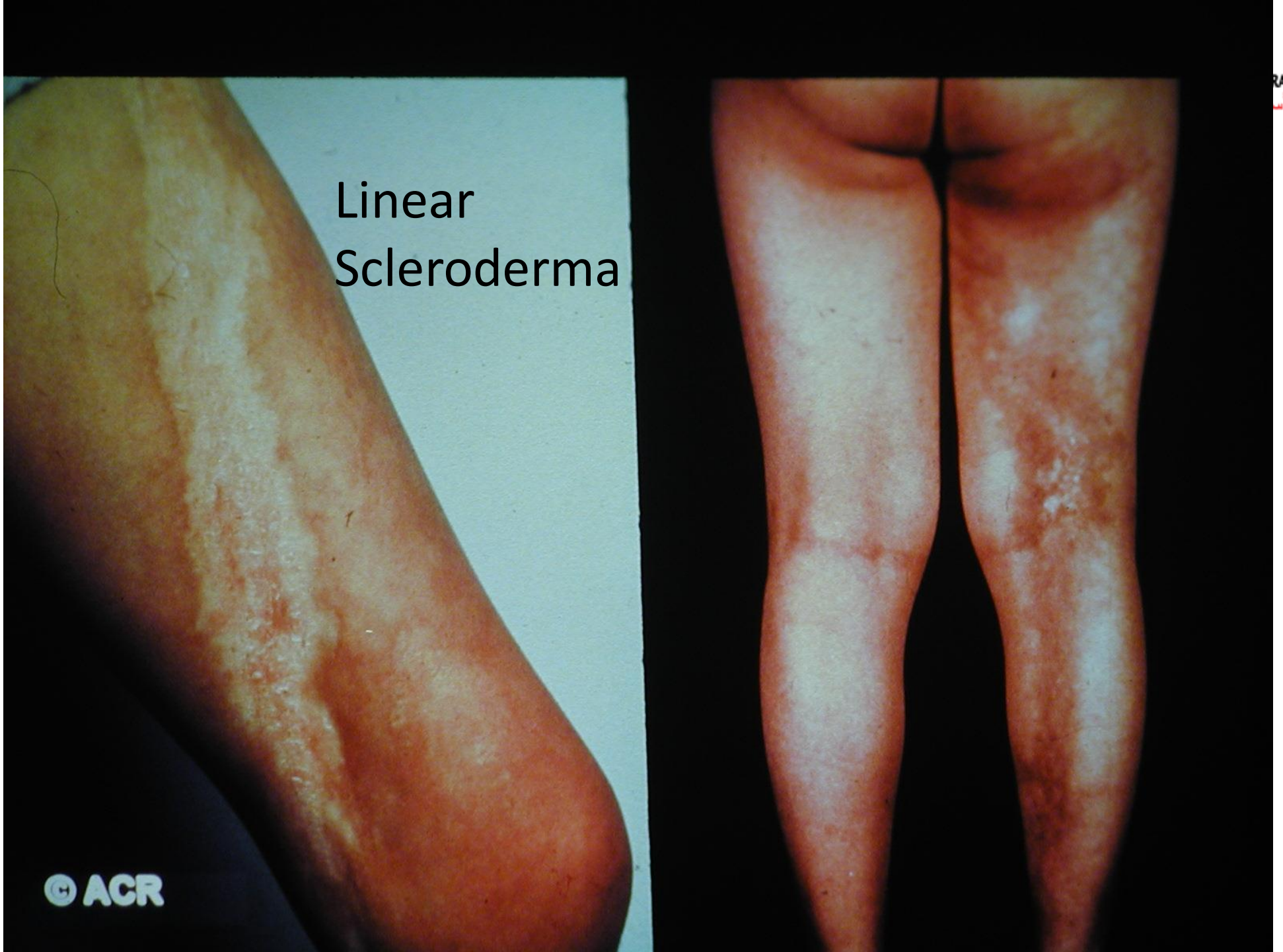


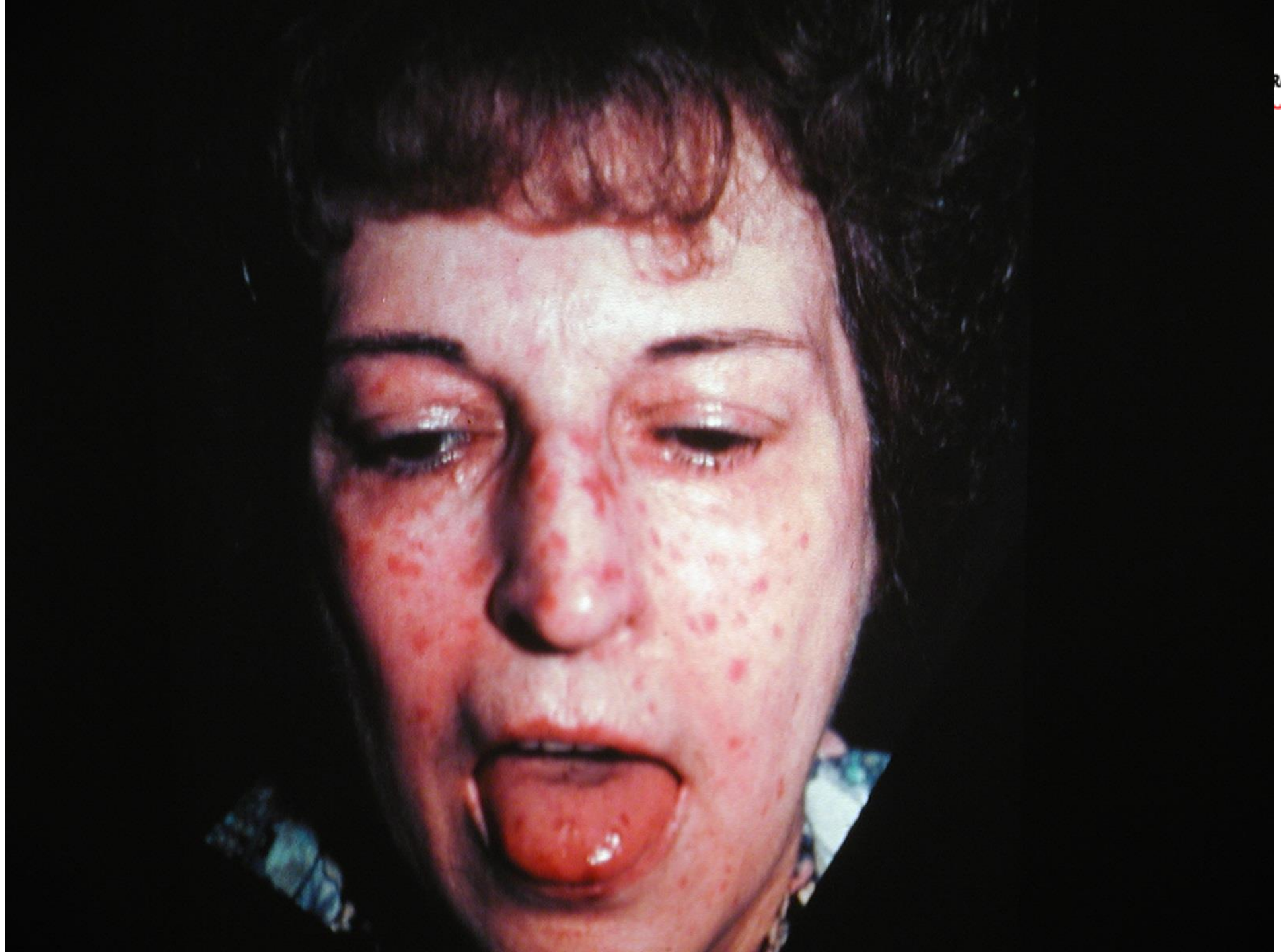


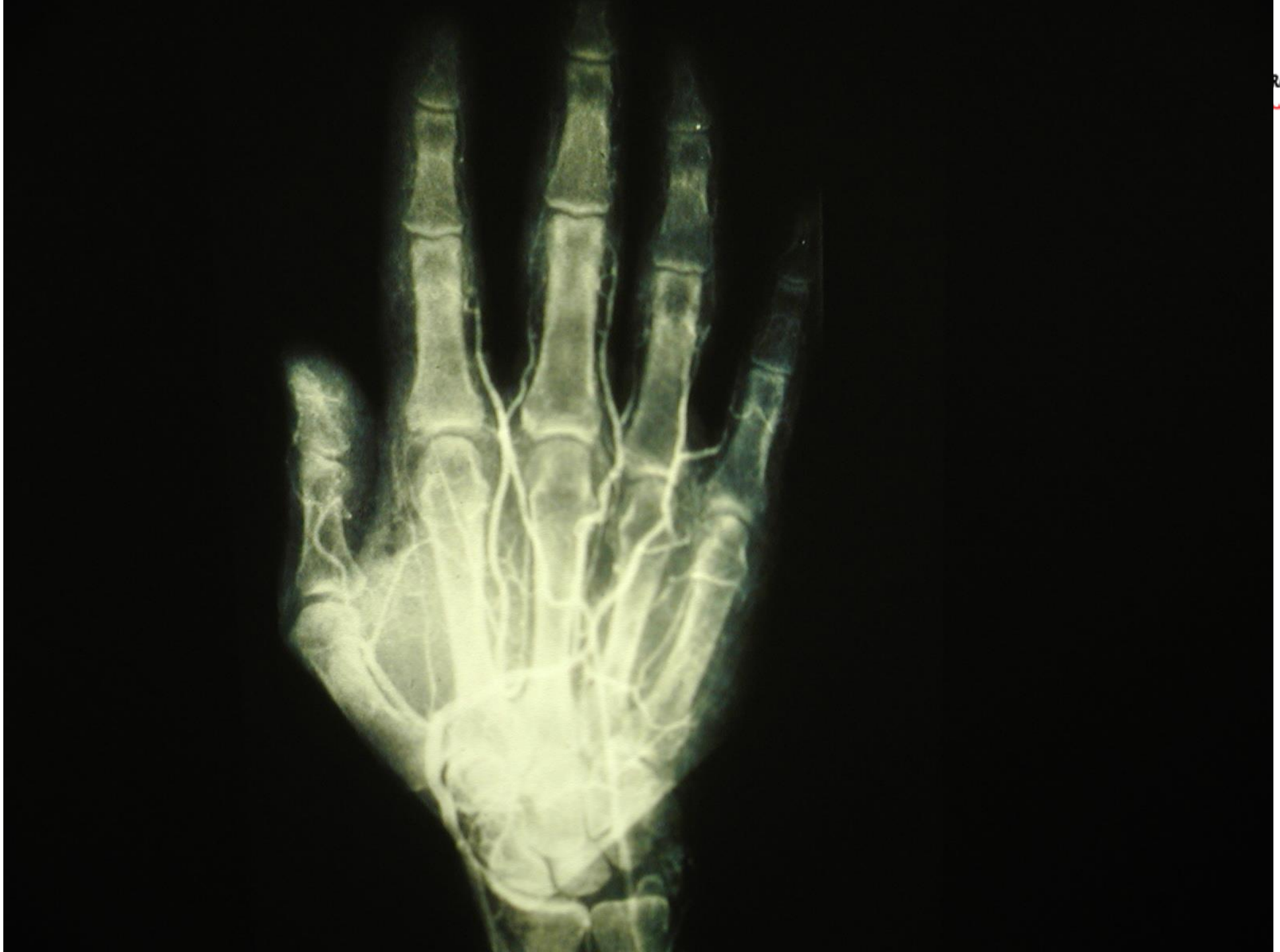

Mauskapf
Mouse Face

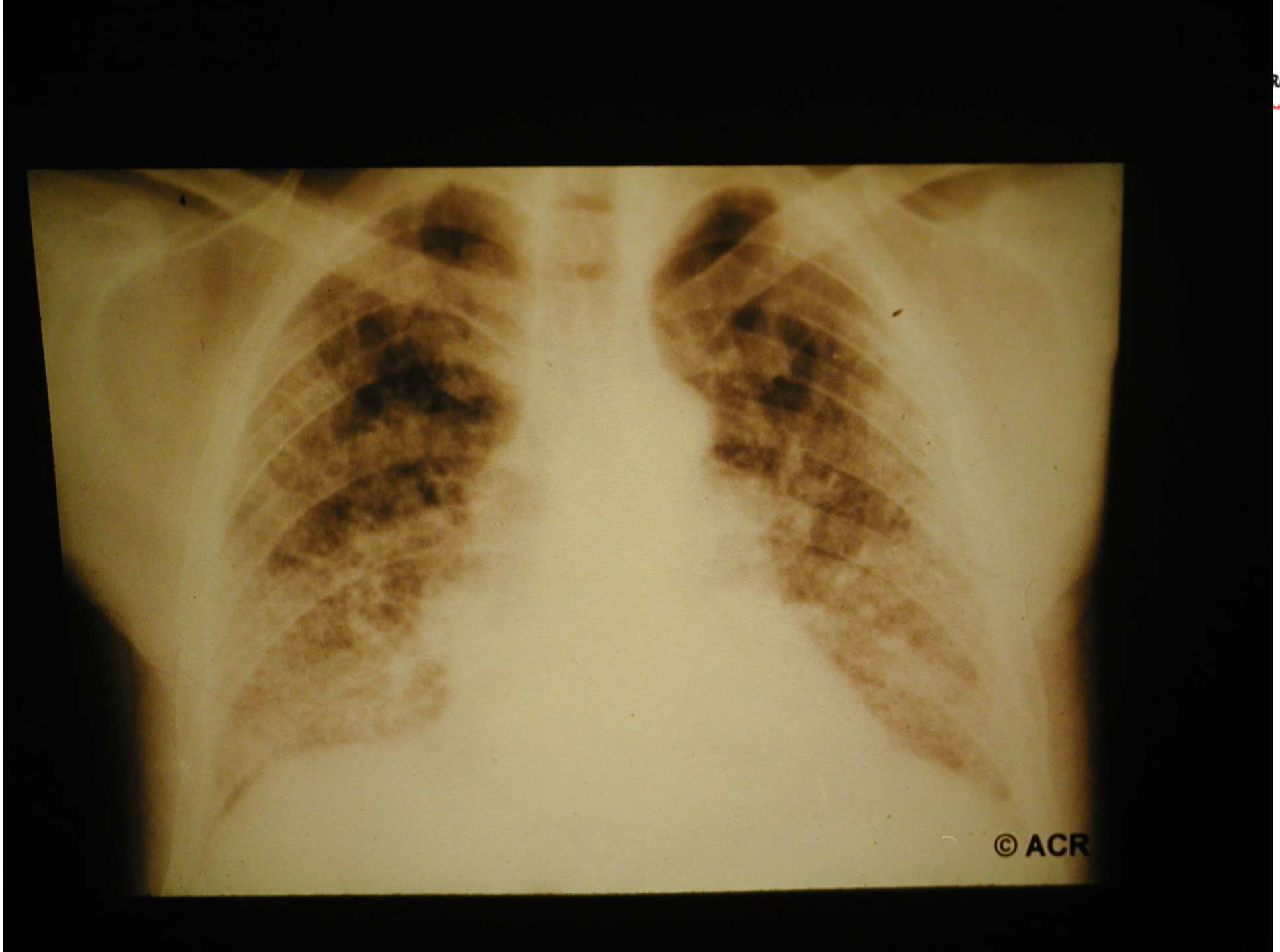


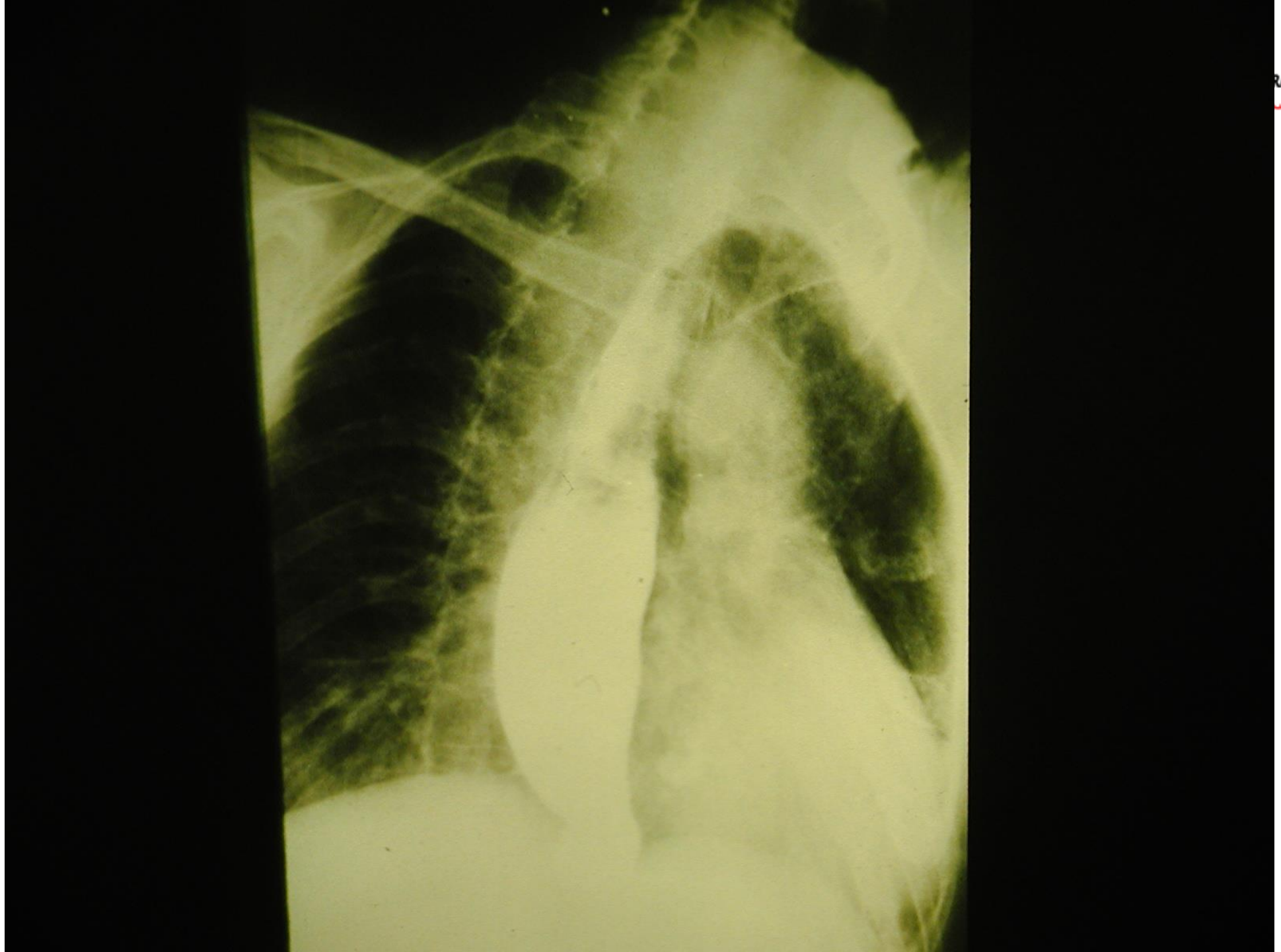
Morphea

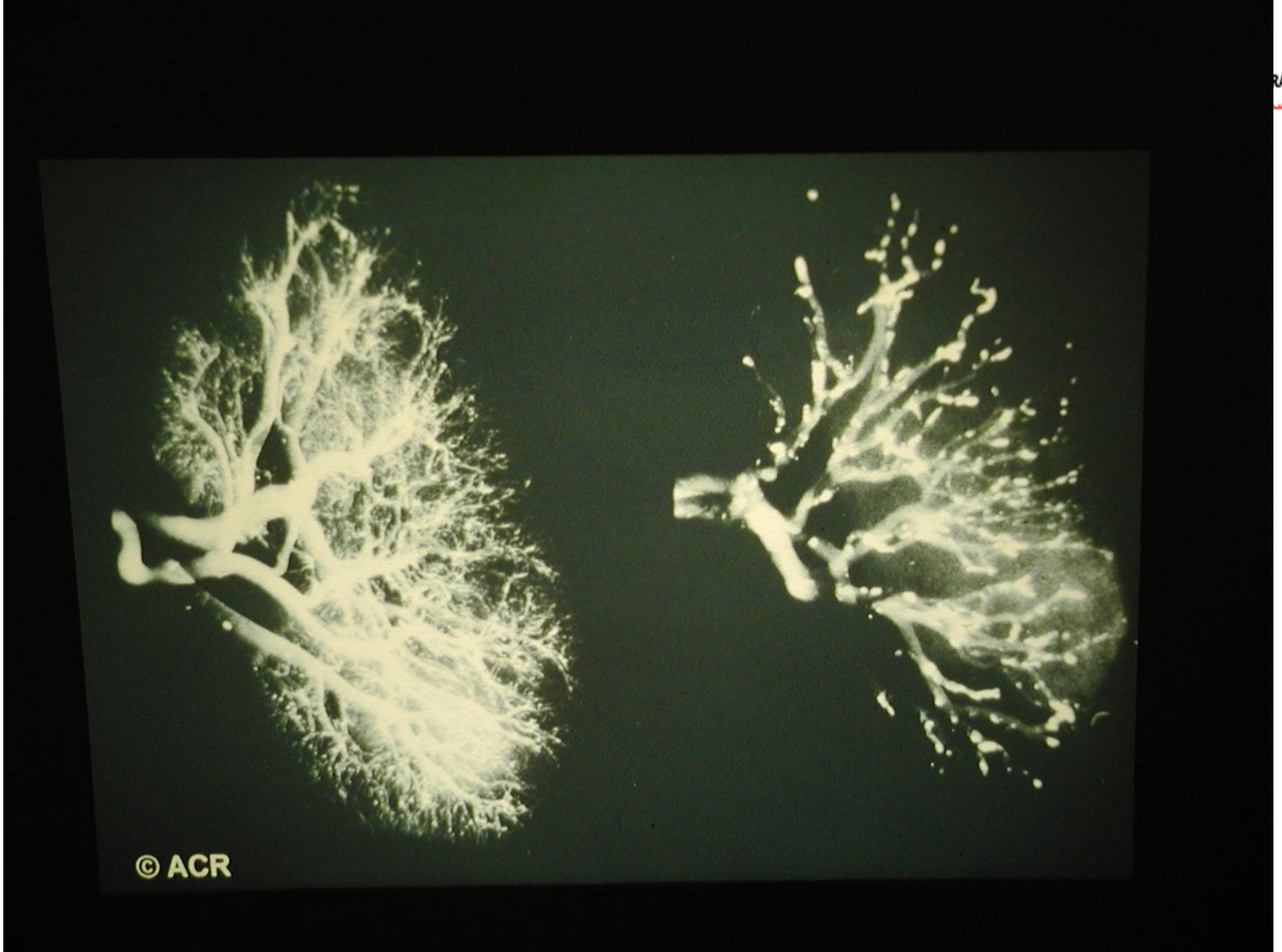












Scleroderma Treatment

- Arthralgias can be treated with:
  - NSAIDs, hydroxychloroquine, MTX, azathioprine, or mycophenolate
- Inflammatory episodes: steroids
- Any patient with scleroderma and HTN should be on an ACEI to preserve renal function
- Esophageal disease/reflux: PPI
- Pulmonary Disease: Cyclophosphamide

Scleroderma Treatment

- Raynaud's phenomenon:
  - Calcium channel blockers: extended-release
    - Nifedipine or amlodipine
  - IV iloprost (prostacyclin analogue – vasodilator)
  - Sildenafil (phosphodiesterase inhibitor leading to vasodilation)

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Rheumatology #2

Patient Case

- 47 yo male with three-year history of progressive weakness.
- He has trouble brushing his hair or reaching over his head.
- MRI brain and spine normal 3 years ago
- 6’ 5” male sitting in NAD
- Can’t get out of a chair without leaning far forward; cannot raise arms past 90 degrees
- He is on no meds at present

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47 yo male with shoulder girdle weakness and normal brain MRI. Blood pressures are 130/70. No recent viral symptoms. Most likely diagnosis?

A. Poliomyelitis
B. Polymyositis
C. COVID myopathy
D. Cushing's syndrome

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Polymyositis

Patient Case

- Patient with similar complaints with:
  - Erythematous plaques on the dorsal MCPs and PIPs of both hands
  - Erythematous patches and plaques on the upper eyelid

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Dermatomyositis

Gottron’s papules

and

Heliotrope rash

Findings in both DM and PM

- Elevations in muscle enzymes
  - CPK, muscle aldolase, LDH, AST, ALT
- Symmetric proximal muscle weakness
- Characteristic EMG findings
- (+) anti-Jo-1 antibodies
- Muscle biopsy demonstrating active inflammation
- Additionally in DM
  - Heliotrope rash and Gottron’s papules

DDx for proximal muscle weakness

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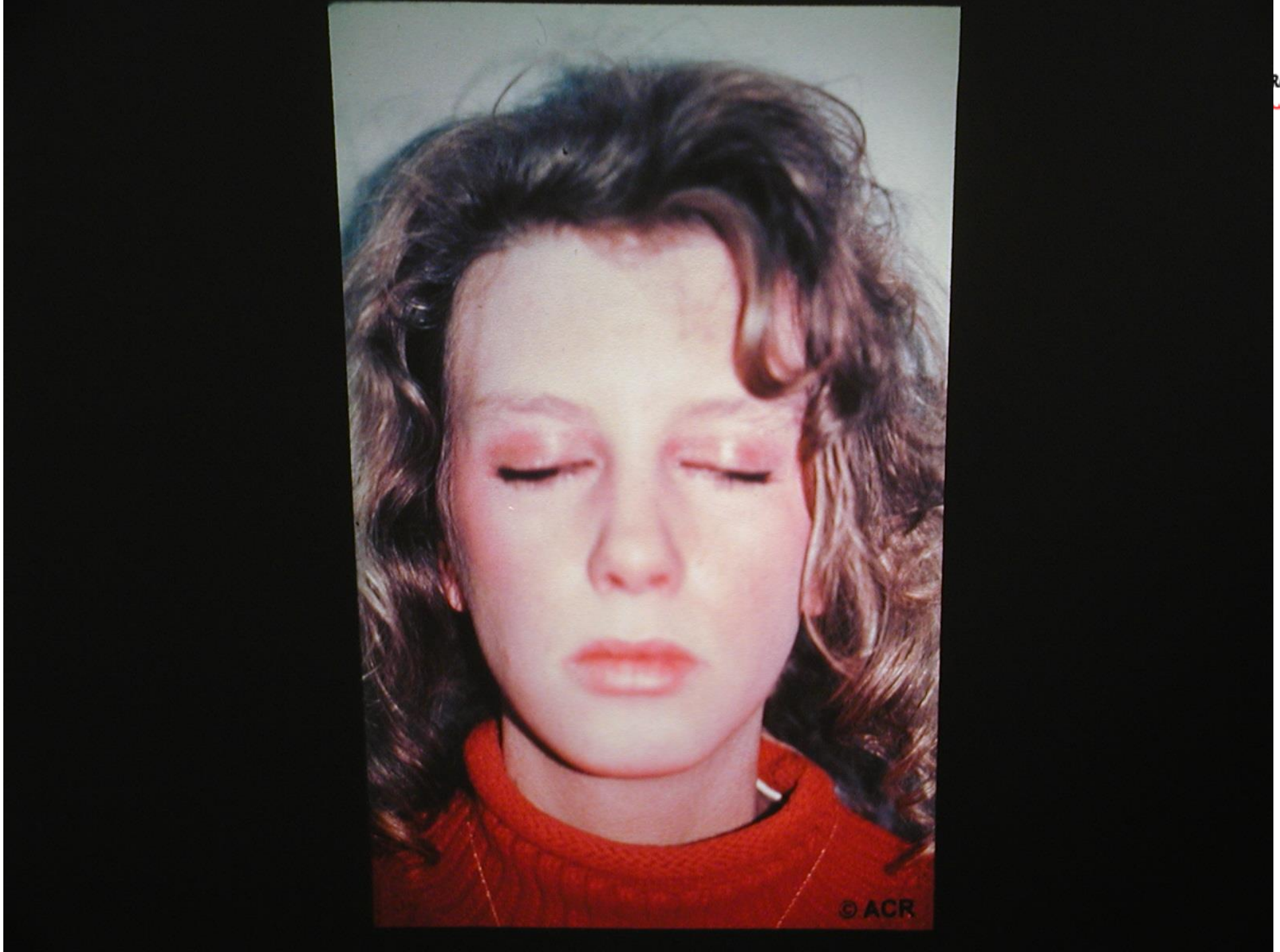
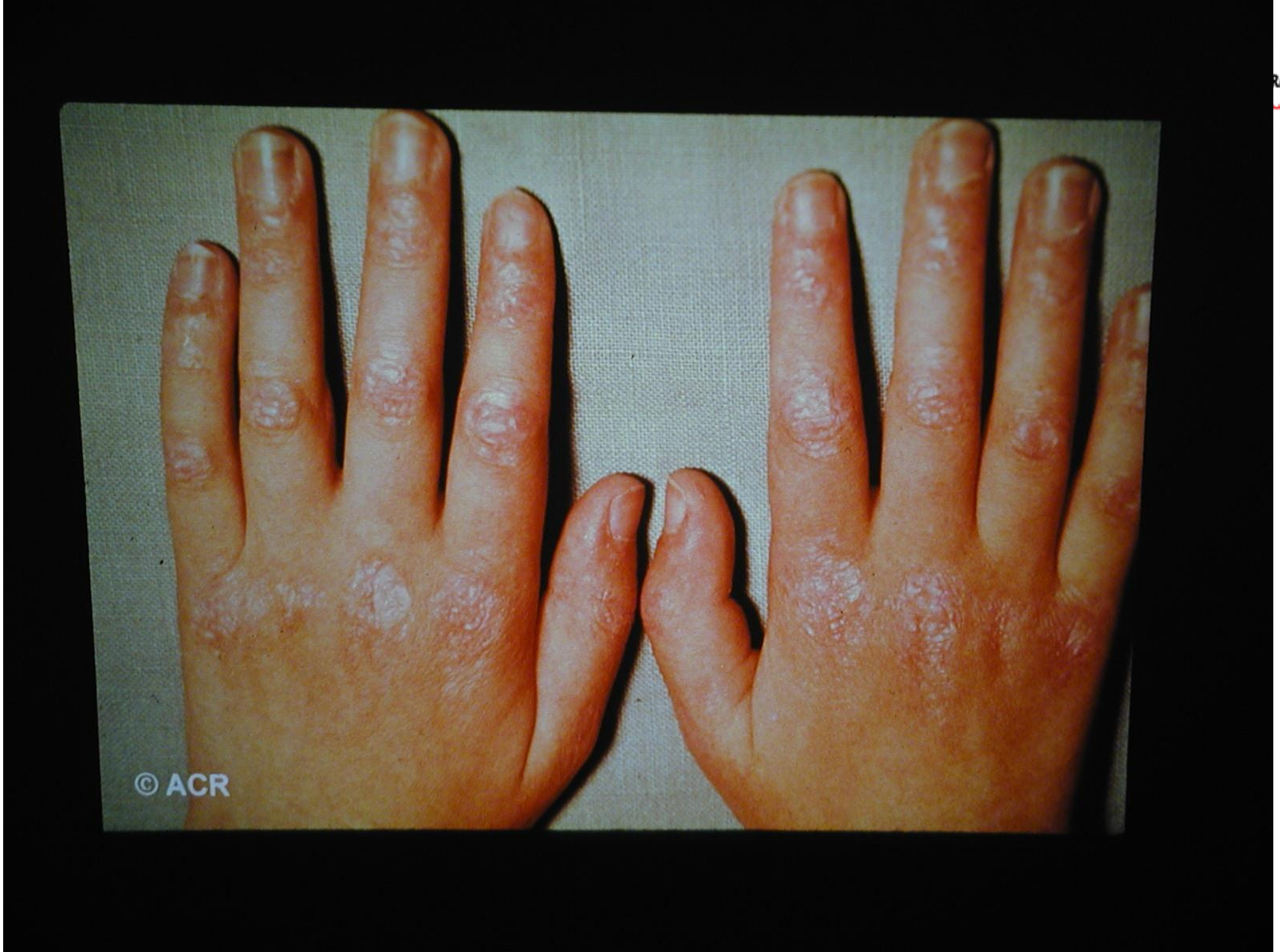
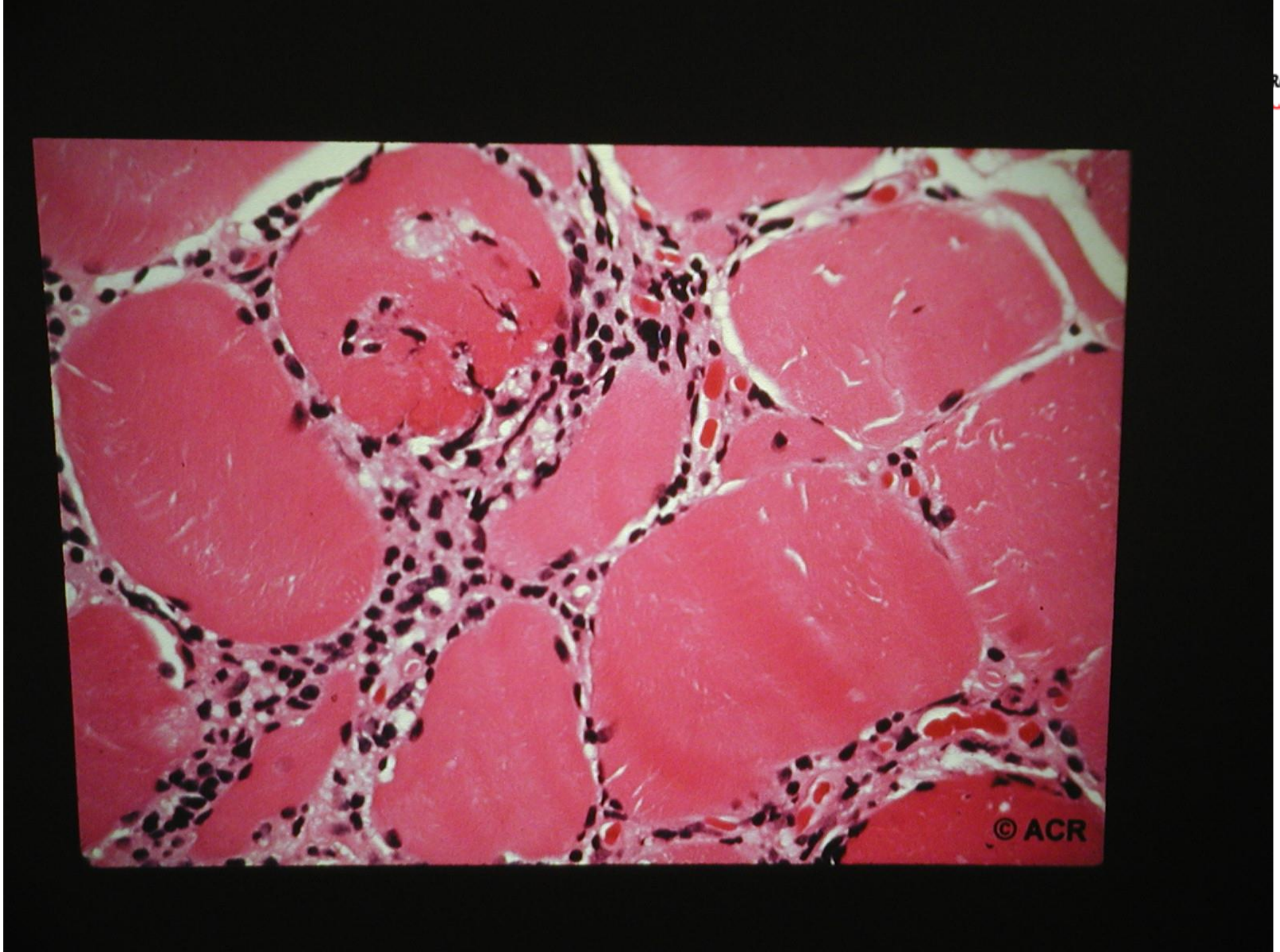
- **DM/PM**
- **Polymyalgia rheumatica**
- **Temporal arteritis**
- Endocrinopathy
  - Thyroid
  - Cushing's disease
  - Parathyroid
- Infections
  - Toxoplasma, trichinosis
  - **Viral**
- Glucocorticoid myopathy
- Statin myopathy
- Neurologic disorders
  - Myasthenia Gravis
  - Eaton-Lambert
  - Amyotrophic lateral sclerosis
- Muscular dystrophies
- Myopathies
- **Electrolyte disturbances**







Dermatomyositis

- Both DM and PM increase the risk of cancer, DM>PM
  - 3 fold increase in RR in DM; 2 fold increase with PM
  - 70% of cancers are peritoneal adenocarcinomas
- Recommend age appropriate cancer screening for patients (Colonoscopy pts > 50 yo, etc…)
- In higher risk patients, consider CT Chest, Abdomen, Pelvis

Sigurgeirsson B, Lindelöf B, Edhag O, Allander E. Risk of cancer in patients with dermatomyositis or polymyositis. A population-based study. N Engl J Med 1992; 326:363.

DM/PM treatment

- Initial high dose steroids to placate the inflammatory myopathy: 60-80mg daily
  - Wean down over ~ 1 year
- Steroid sparing agents:
  - Azathioprine
  - Methotrexate
  - Hydroxychloroquine

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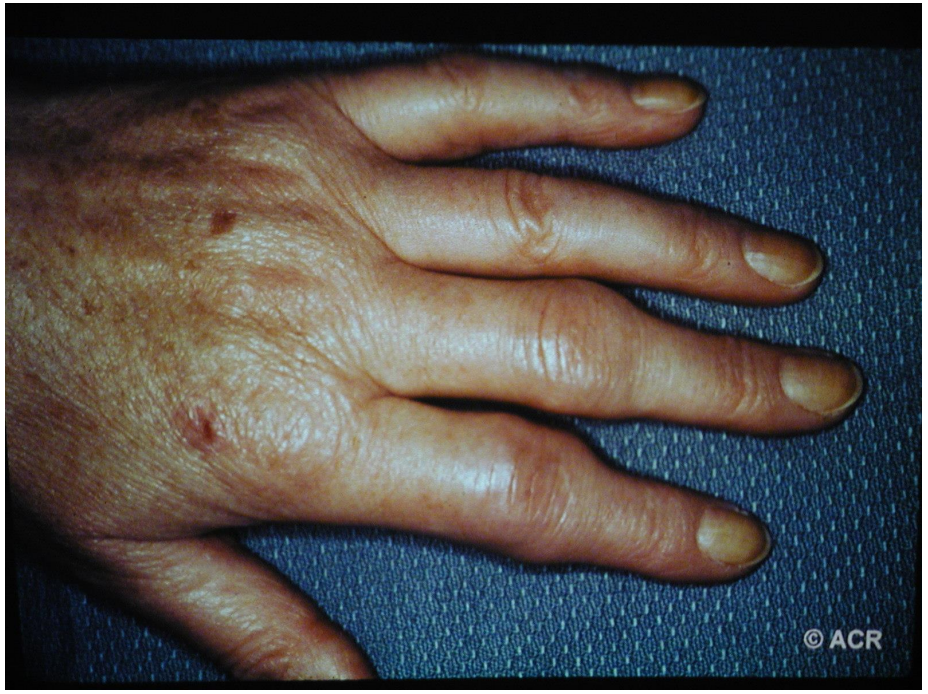
A 37 yo woman presents with painful swollen joints in her hands. No recent URI symptoms. No F/C, (+) general fatigue, weight stable. What is the most likely diagnosis?

A. Rheumatoid arthritis
B. Dermatomyositis
C. Sjogren's syndrome
D. Gout



Rheumatoid Arthritis - Epidemiology

- RA is the most common autoimmune **inflammatory** arthritis
  - Gout is inflammatory, but not autoimmune
  - OA is arthritis but not actively inflammatory
- Inflammation: tumor/swelling, dolor/pain, calor/warmth, rubor/erythema
- Lifetime prevalence of 0.25 - 1%
- Most common in women 30-50 years old, smokers, and patients with a (+) family history of RA
- Most commonly affects the proximal joints of the hands and feet
  - MCP, MTP, and PIP
  - Does not commonly affect DIP or lumbar spine

RA differential diagnosis

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- Osteoarthritis
  - Not inflammatory. Lab tests either negative or false (+)
  - ANA (+) in 10% of population
- Gout
  - Usually monoarticular; Podagra/inflammation at first MTP in 50% gout patients
  - RA specific tests, RA, ACPA, usually negative
  - CRP and ESR can be quite elevated in acute gout
- CTD: SLE/Lupus, Sjogren's syndrome, Dermato-/Polymyositis
  - Diagnostic clues
  - Labs tests: ANA with pattern
- Fibromyalgia
  - No signs of active inflammation
  - Diagnostic tests negative

OA vs RA

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Osteoarthritis

- **Large joints: knees, hips, spine, DIPs**
- DIPs – Heberden's nodes
- Carpometacarpal joint of the thumb is commonly involved.
- Swelling of the joints is hard and bony in OA.
- Stiffness after use – evening
- **Xray – joint space narrowing, osteophytes**

Rheumatoid Arthritis

- **Small joints: MCP, MTP, PIP**
- Swan neck and boutonniere deformities
- Soft, warm, boggy, and tender joints
- Signs of inflammation: tumor, dolor, calor, rubor
- **Stiffness worse after rest – morning stiffness > 30 minutes**
- **Xray – joint space destruction, erosions, soft tissue edema**

Tests specific for Rheumatoid Arthritis

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- Rheumatoid factor (RF)
  - IgM antibody
  - Can be elevated in other conditions than RA
  - 5-10% of general population has a (+) RF
- Anticyclic citrullinated peptide antibodies (ACPA)
- RF and ACPA are both negative in 50% of patients at initial presentation.
- They are both (+) in 80% pts long term.
- Of note, 30% of RA pts have a (+) ANA

Diagnostic criteria : Need ≥6 points to diagnose Rheumatoid Arthritis

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- Joint involvement
  - One large joint 0
  - Two to 10 large joints 1
  - One to three small joints 2
  - Four to 10 small joints 3
  - >10 joints (at least one small) 5
- Serology
  - Negative RF and negative ACPA 0
  - Low (+) RF or low (+) ACPA 2
  - High (+) RF or high (+) ACPA 3

RF = Rheumatoid factor; ACPA = anti(cyclic)citrullinated protein antibodies

Aletaha D, Neogi T, Silman AJ, et al. 2010 Rheumatoid arthritis classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative. Arthritis Rheum 2010; 62:2569.

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Diagnostic criteria : Need ≥6 points to diagnose Rheumatoid Arthritis

- Acute Phase Reactants
  - Normal CRP and normal ESR 0
  - Abnormal CRP or abnormal ESR 1
- Duration of symptoms
  - < 6 weeks 0
  - ≥ 6 weeks 1

- Total all points for final score

CRP = C-reactive protein; ESR = erythrocyte sedimentation rate

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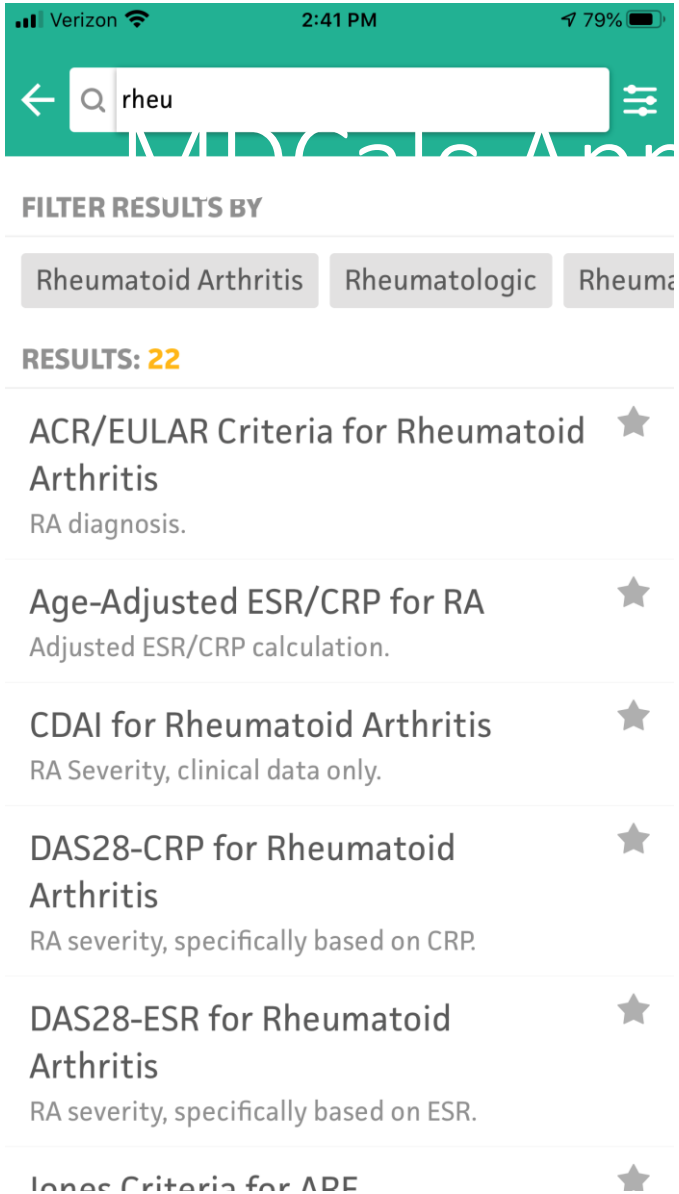
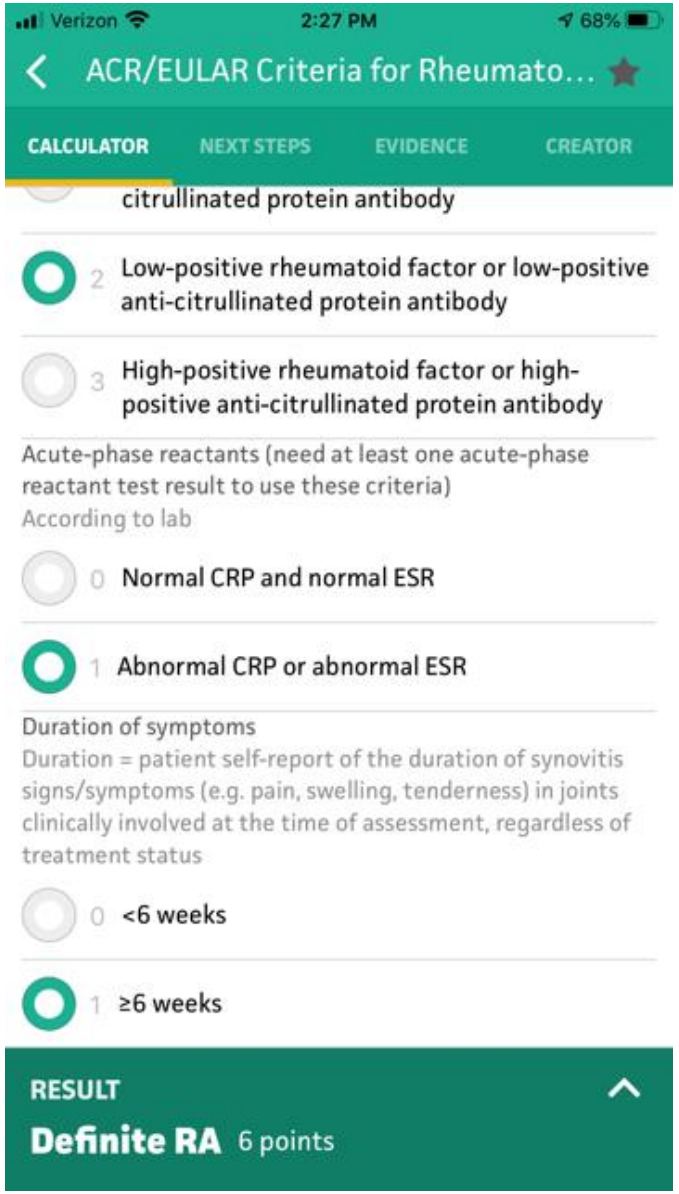
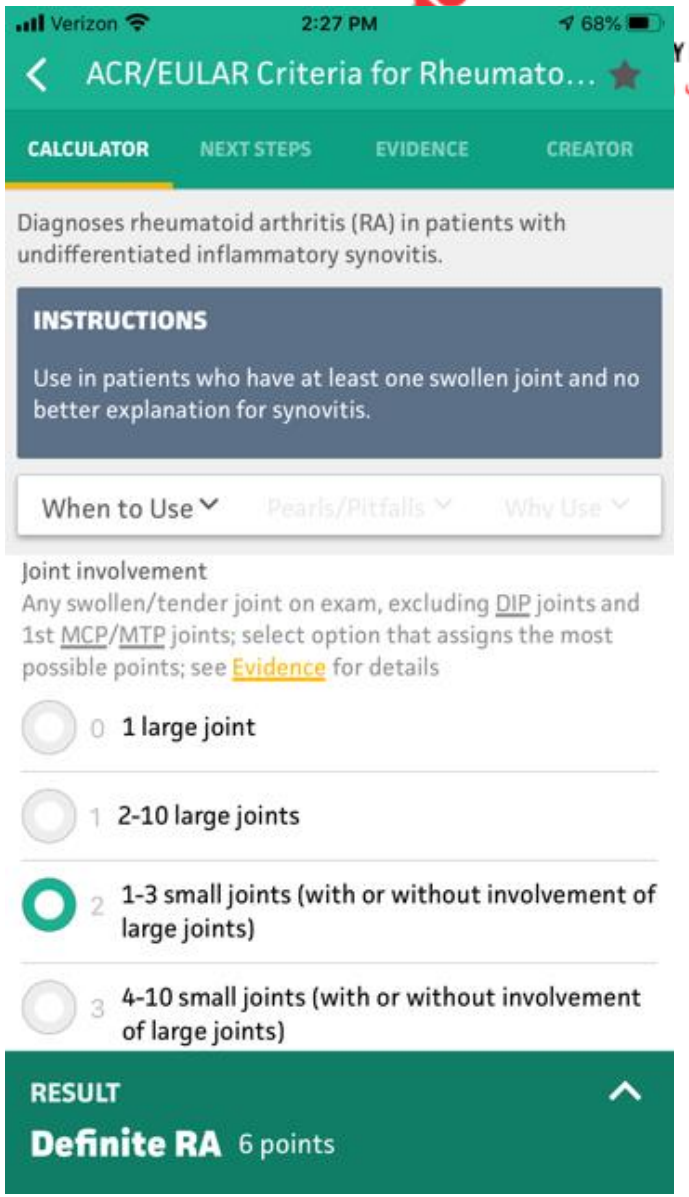
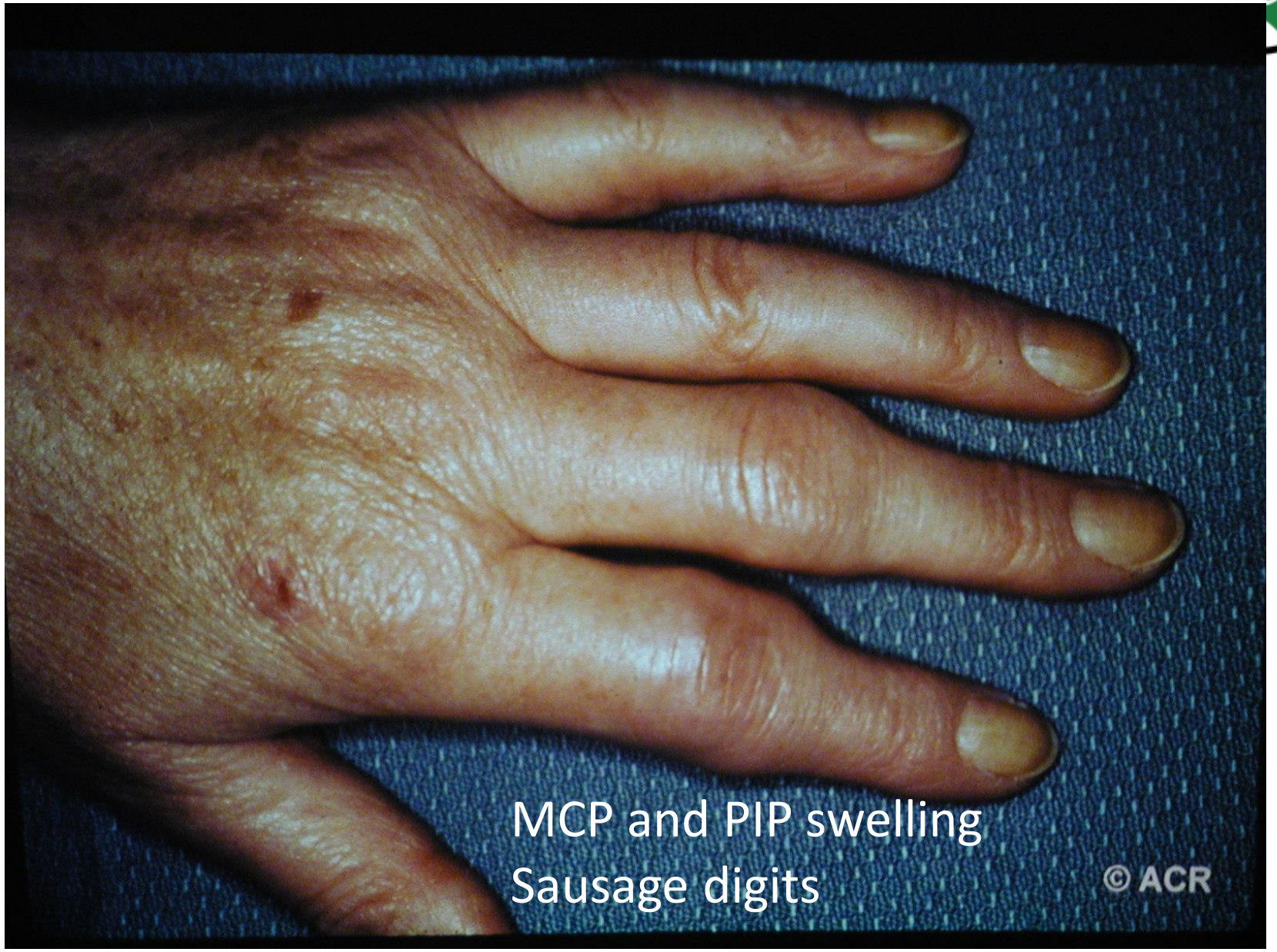
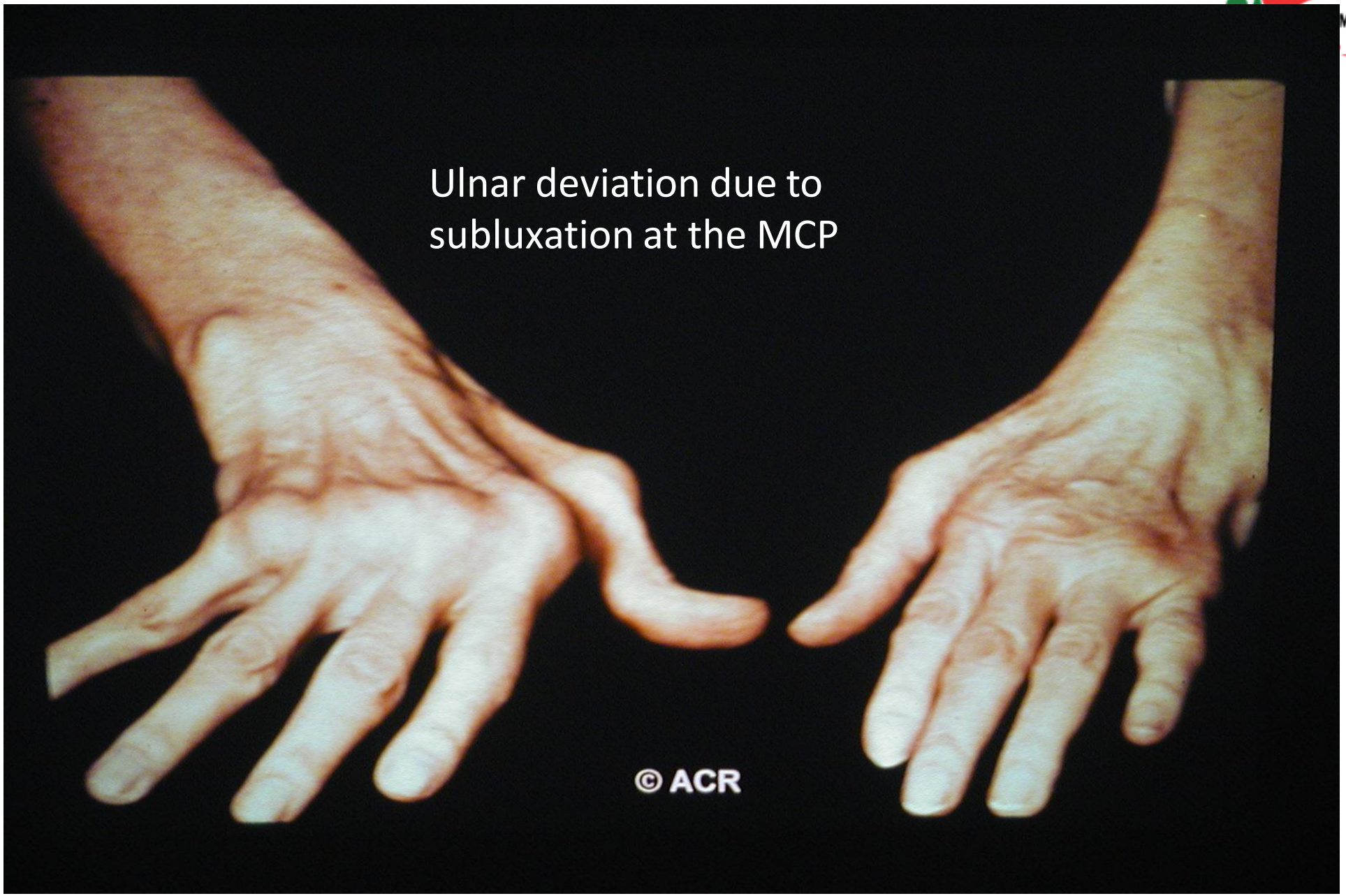
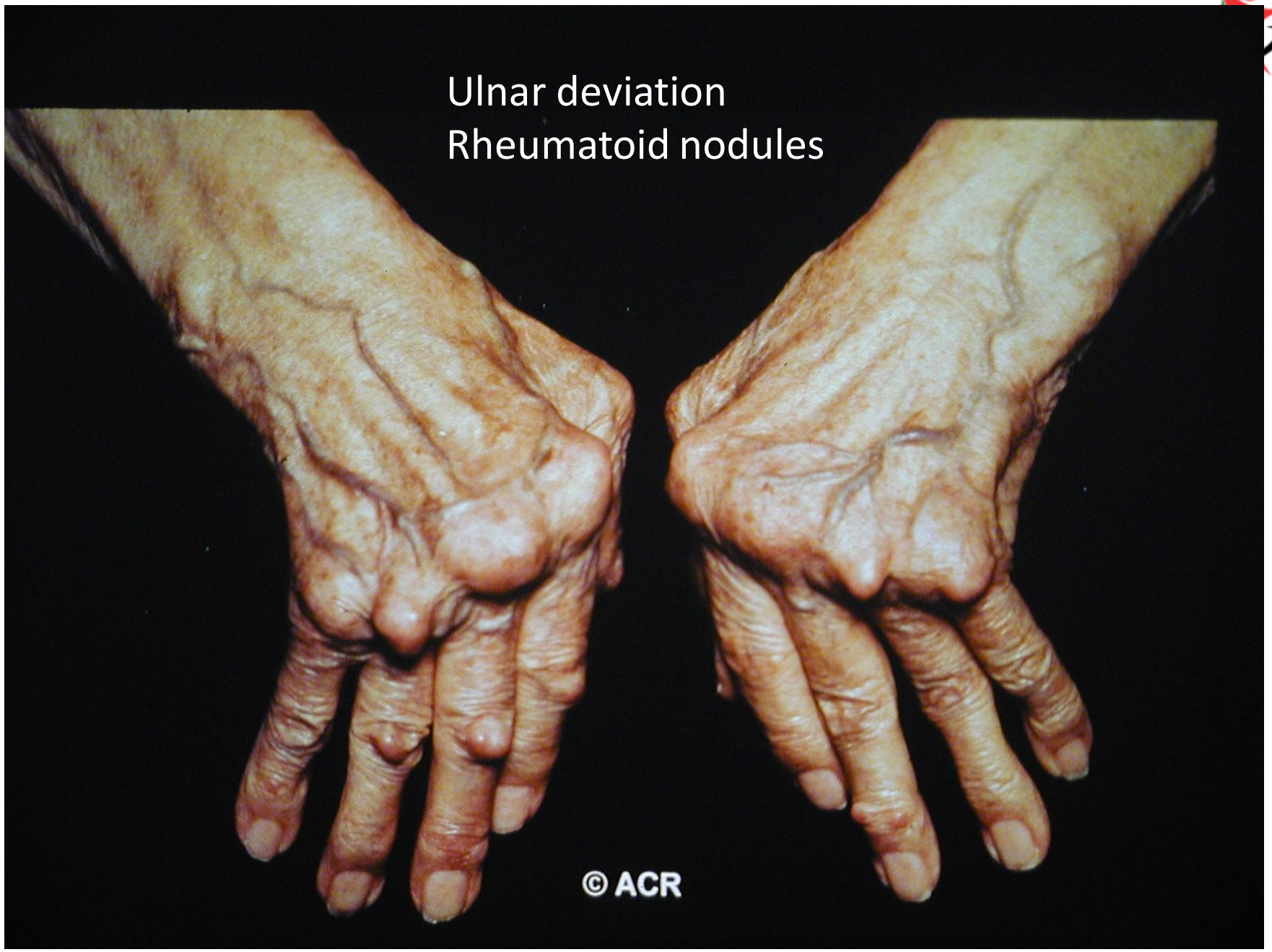


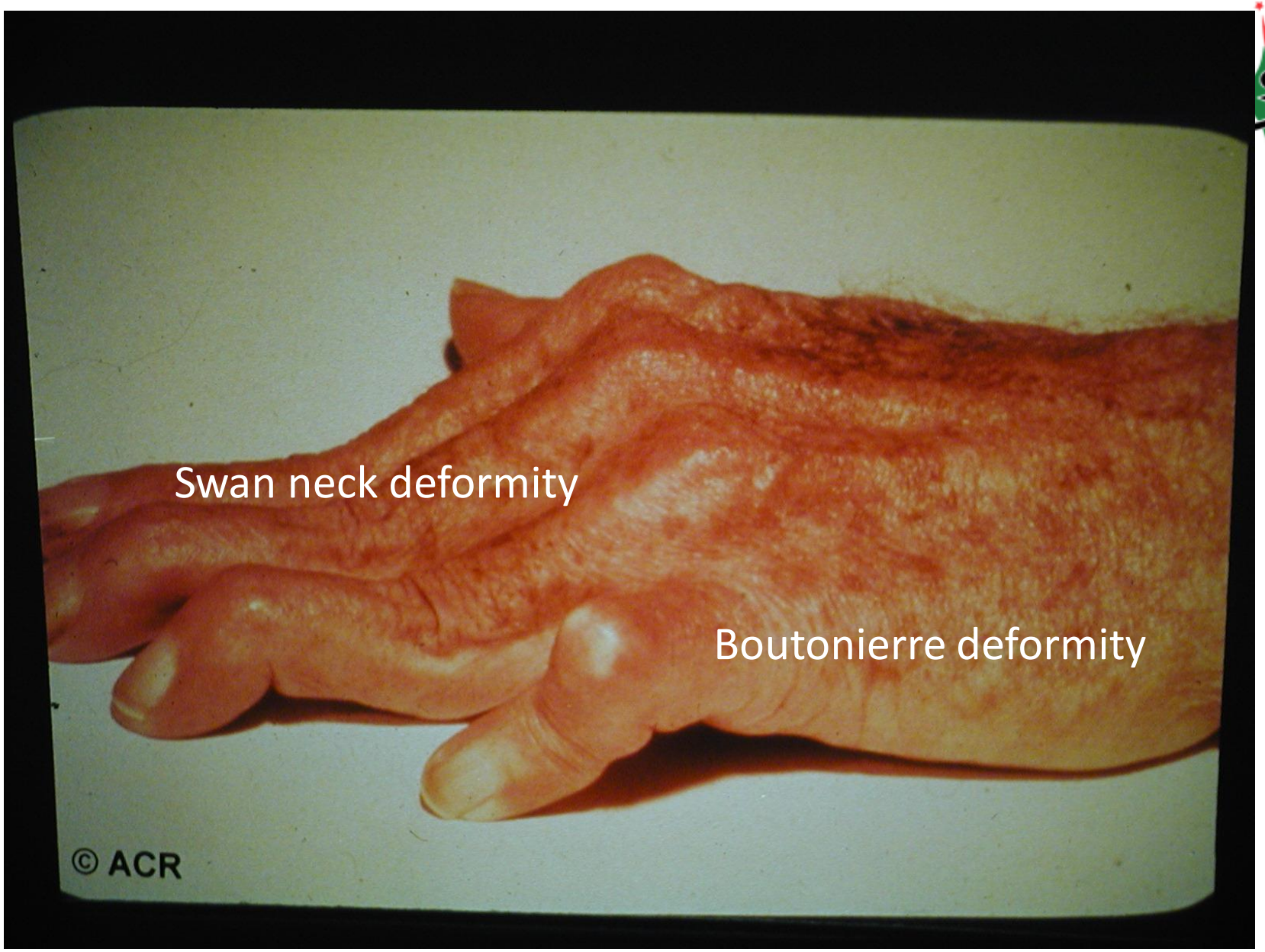




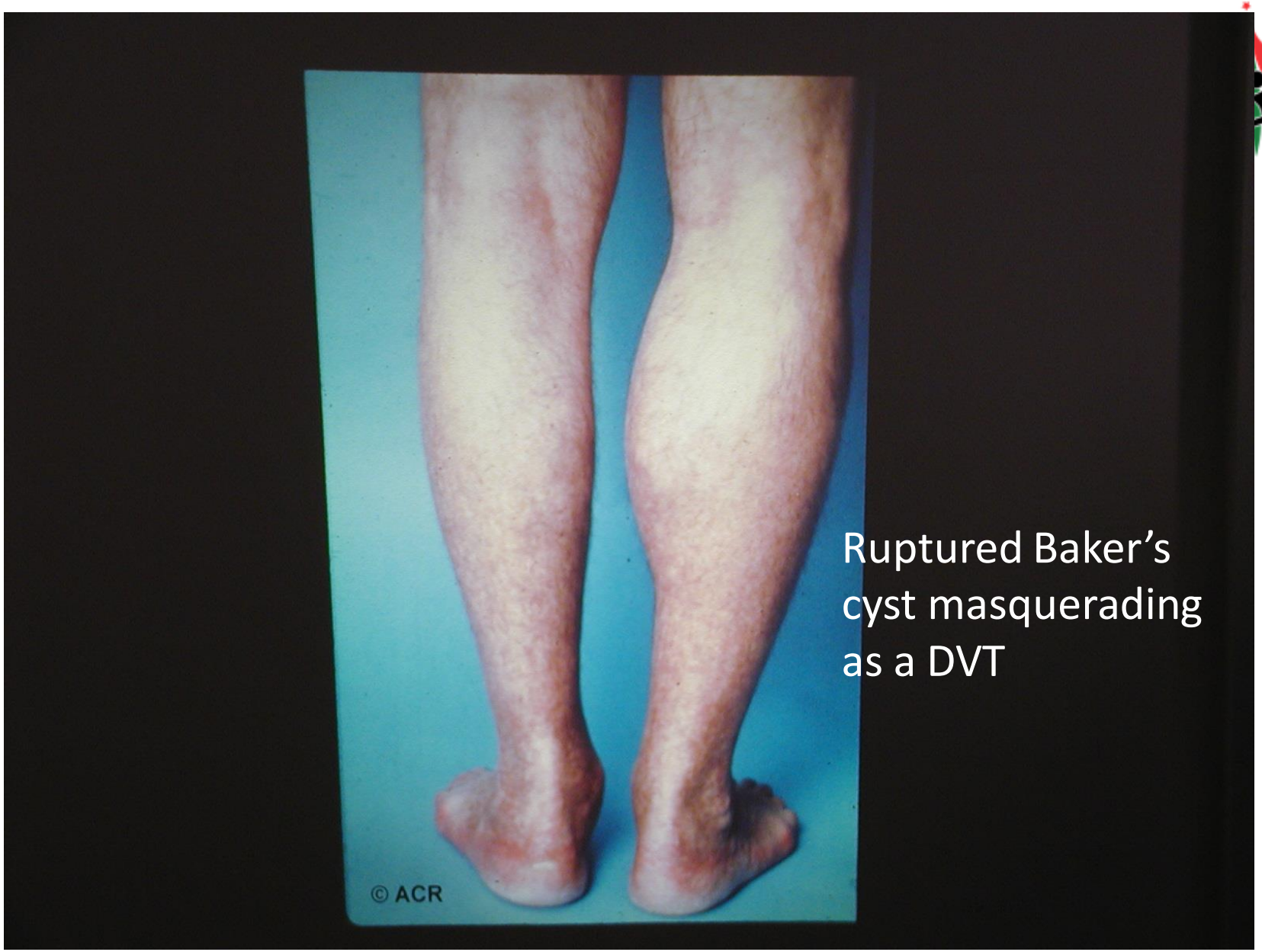

MCP and PIP swelling
Sausage digits
© ACR


Ulnar deviation due to
subluxation at the MCP
© ACR


Ulnar deviation
Rheumatoid nodules
© ACR



EMIRATES FAMILY MEDICINE SOCIETY
شعبة الإمارات لطب الأسرة



Ruptured Baker's cyst masquerading as a DVT

EMIRATES FAMILY MEDICINE SOCIETY
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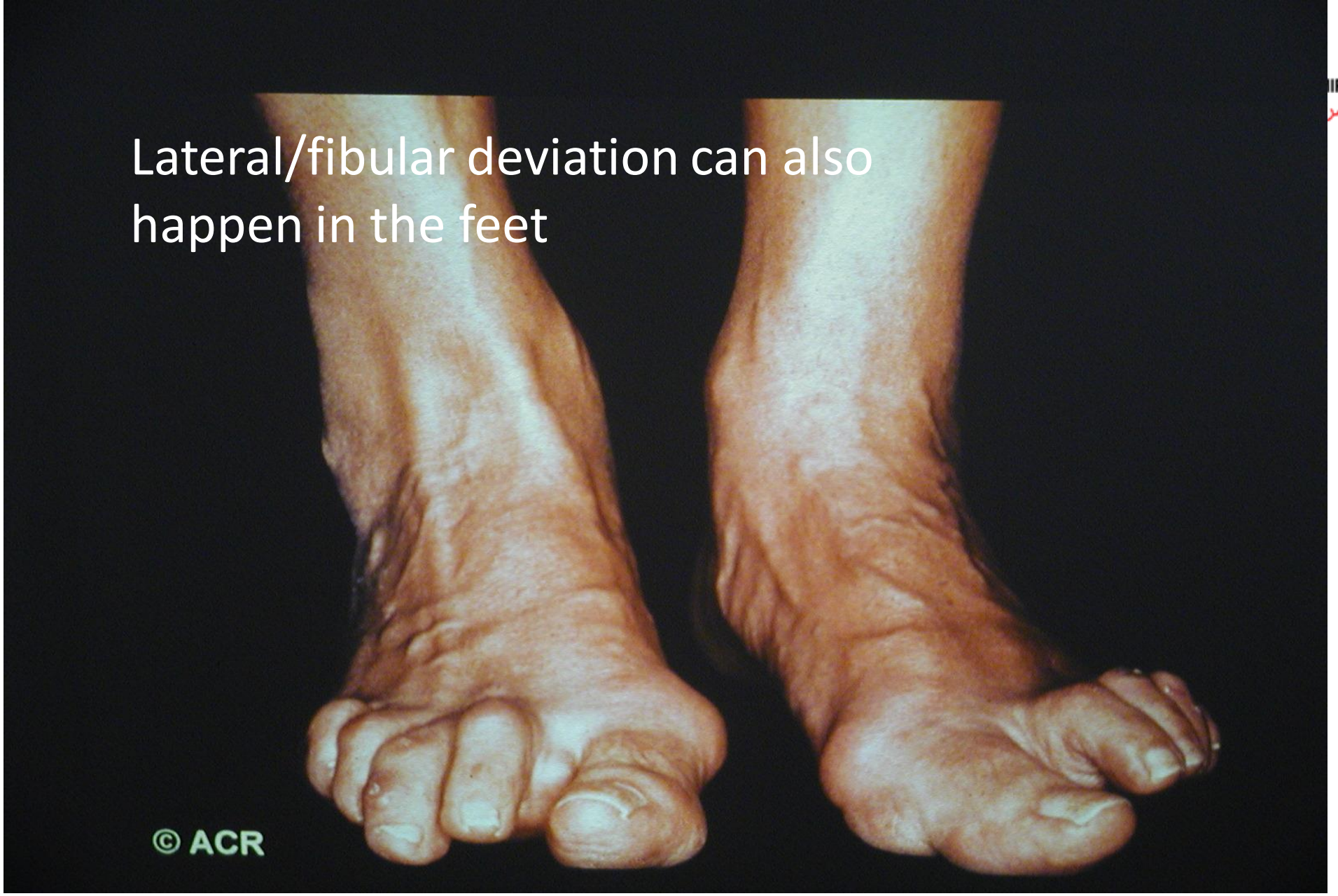
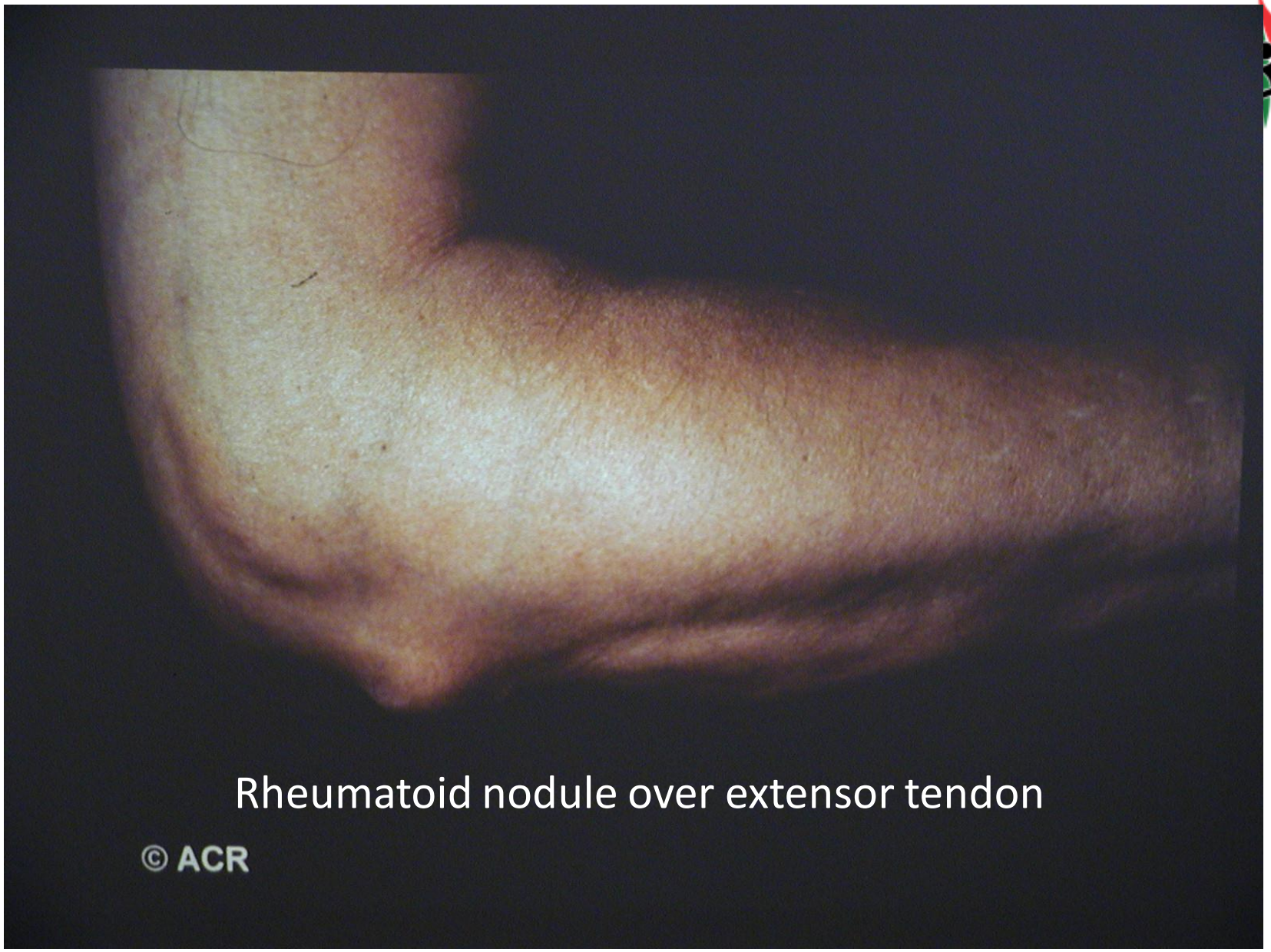
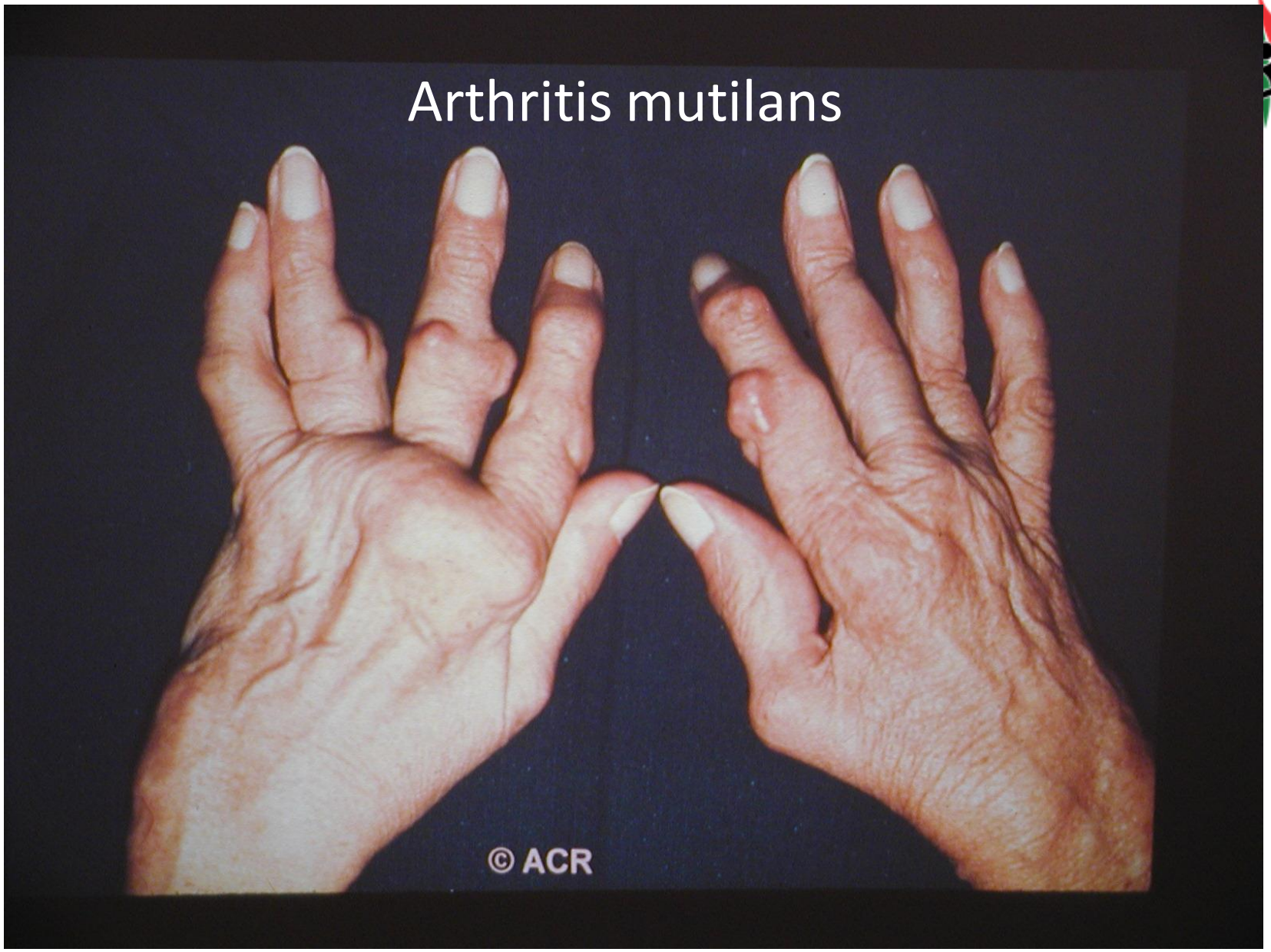
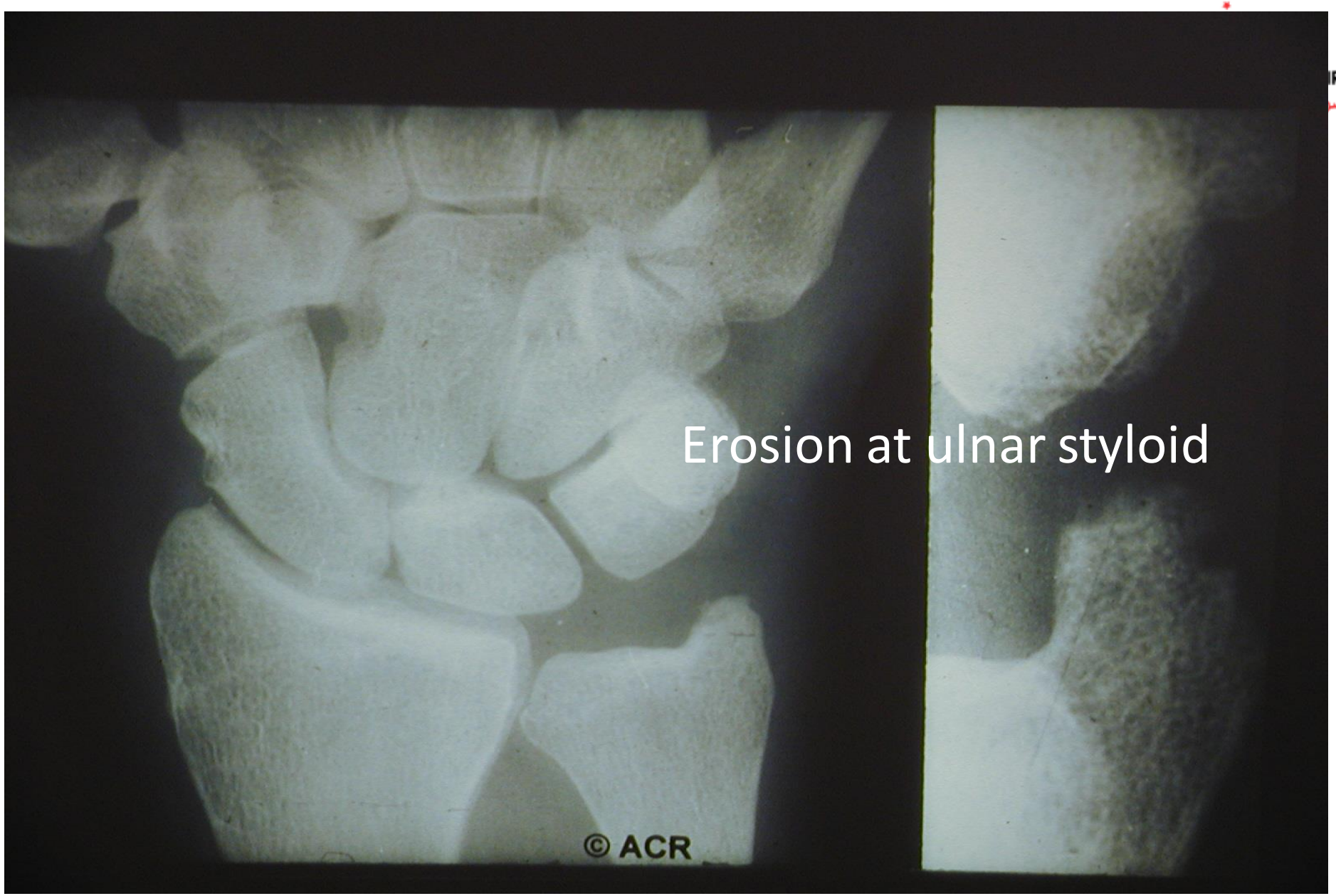

Lateral/fibular deviation can also happen in the feet
© ACR



Rheumatoid nodule over extensor tendon

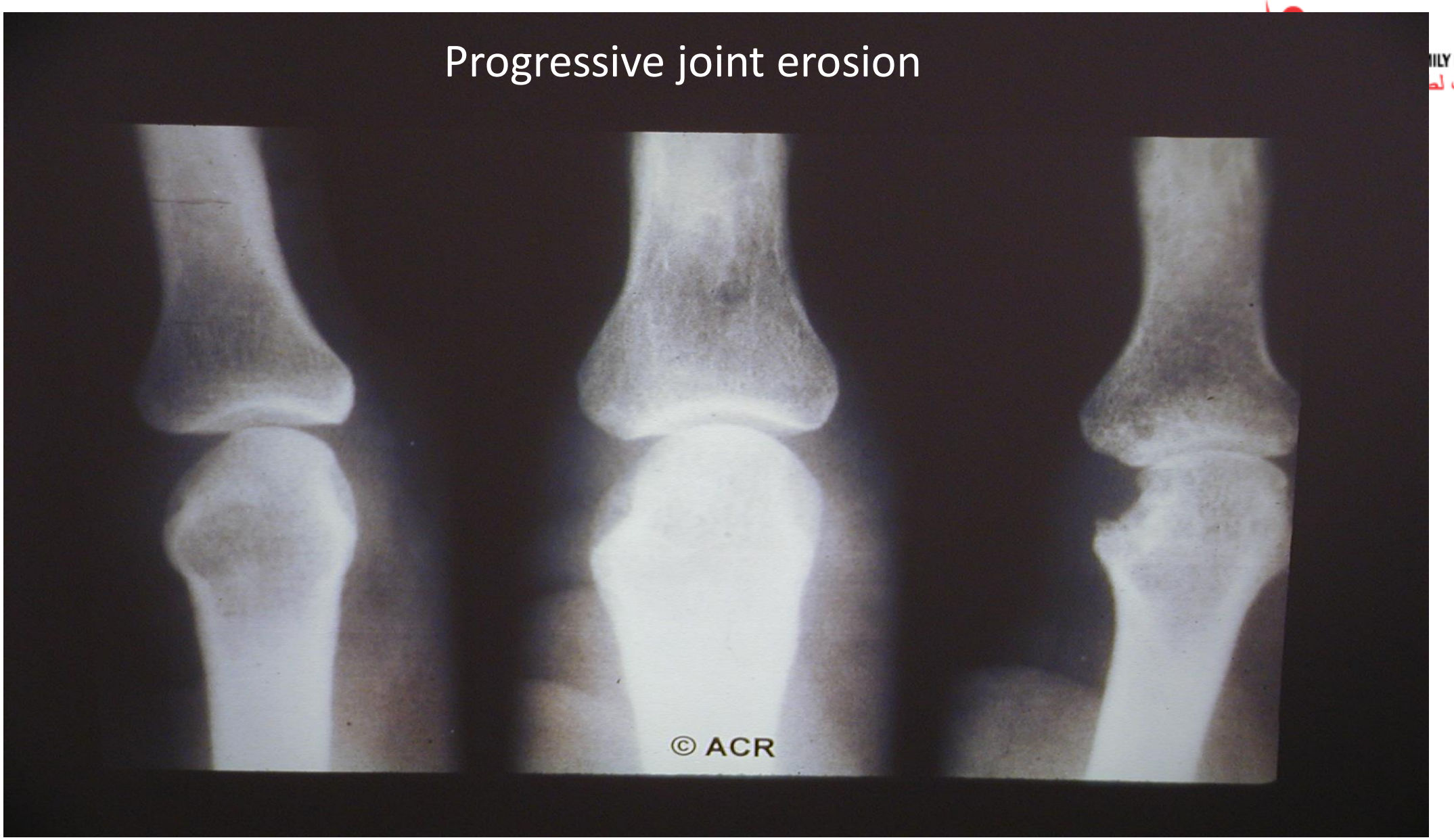
Arthritis mutilans



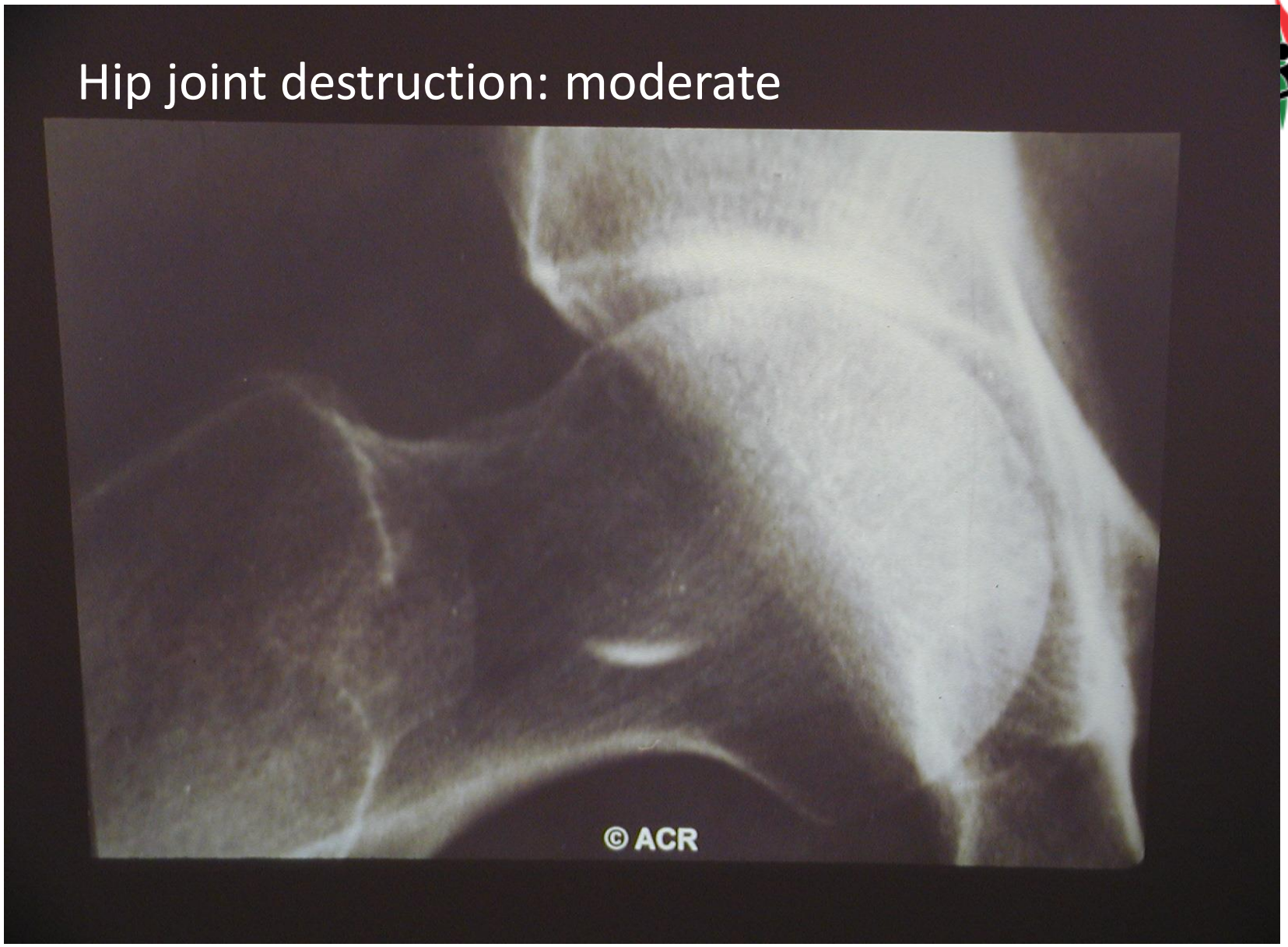


Look at the soft tissue in addition to the joints.

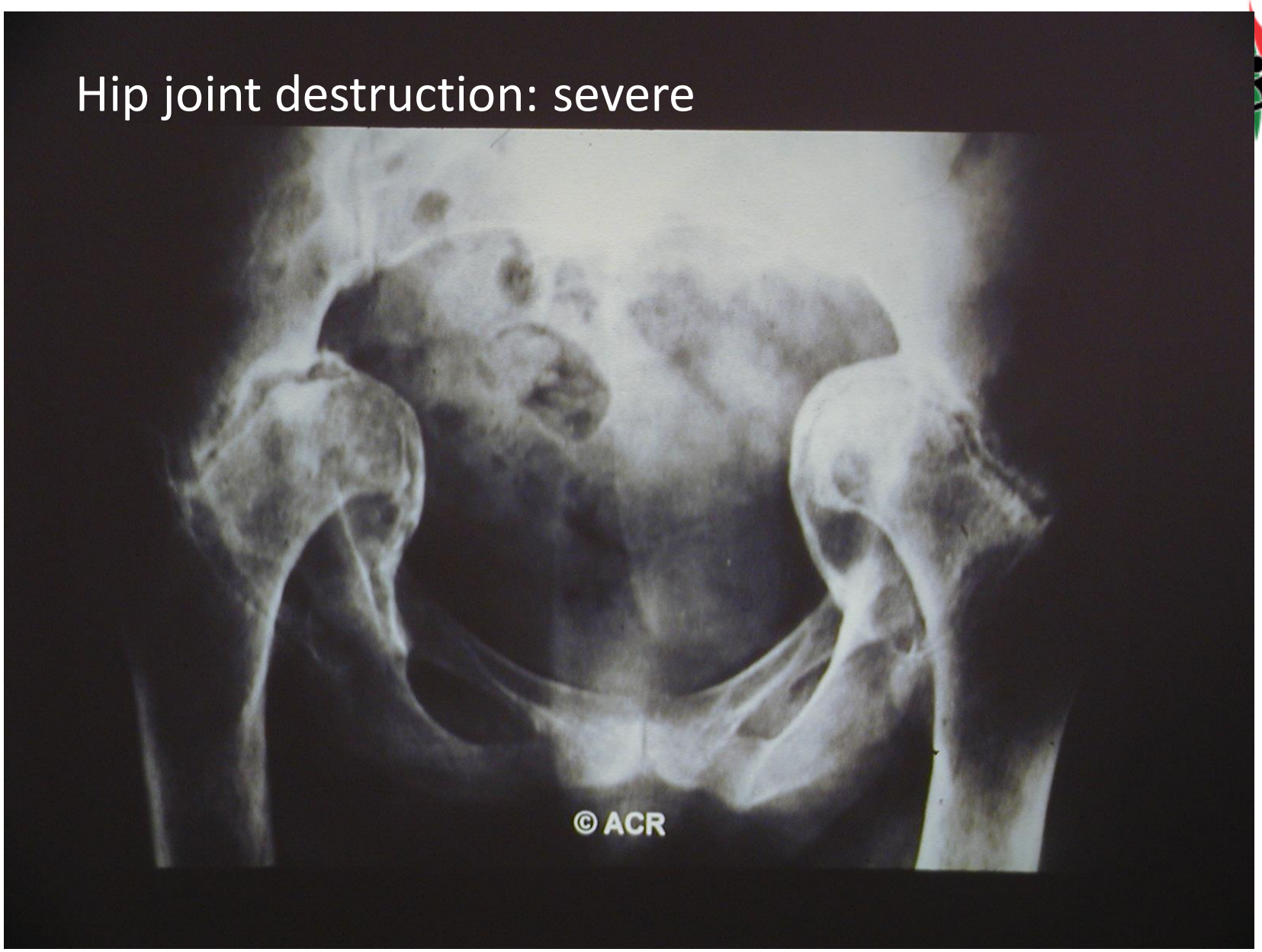
Progressive joint erosion

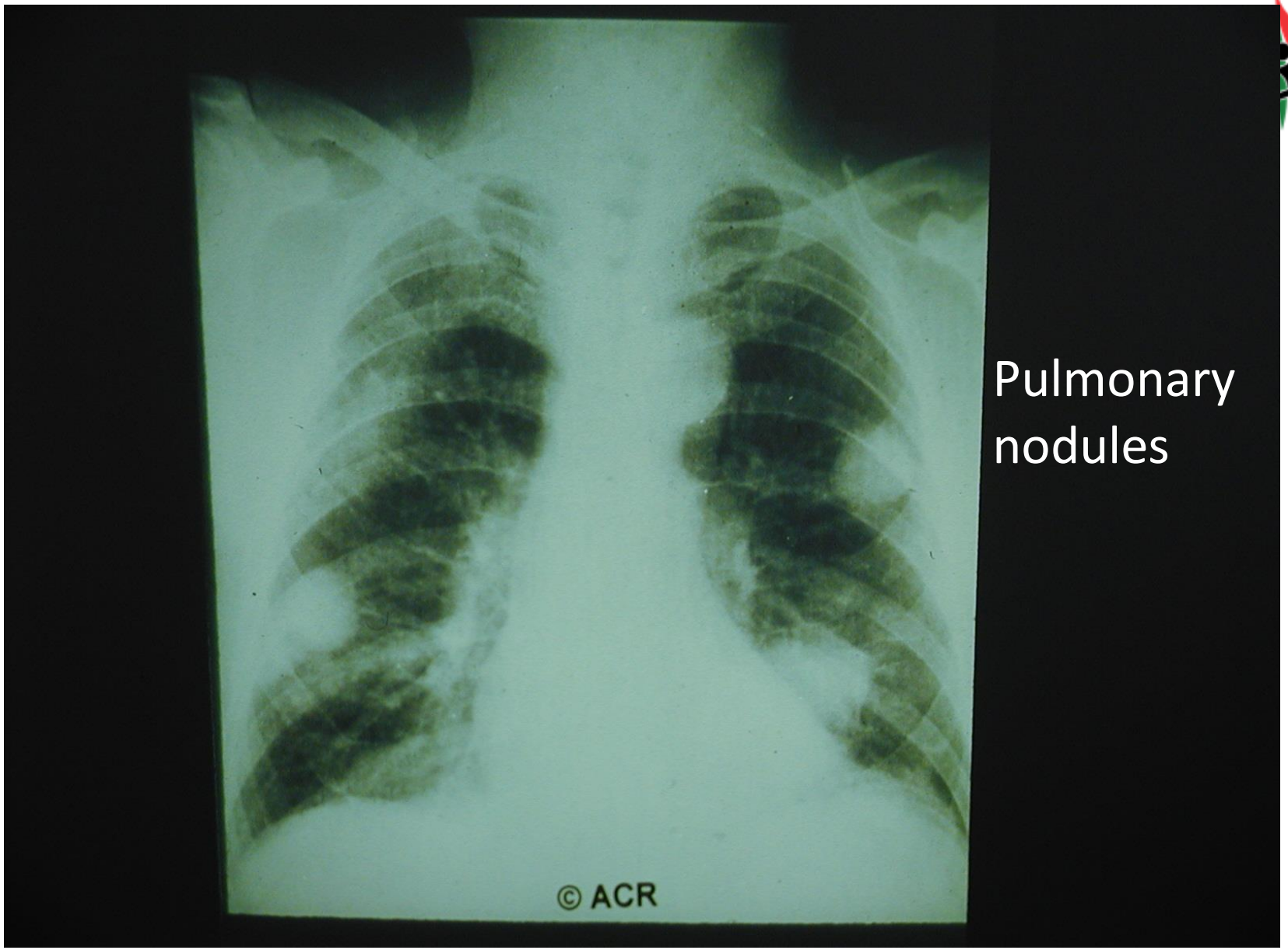


Hip joint destruction: moderate



Hip joint destruction: severe





Pulmonary nodules

Treatment for RA

- Pretreatment screenings:
  - CBC, CMP (Cr, AST, ALT), others as indicated
  - Assessment of comorbidities
  - Immunizations
  - Hep B/C
  - Tuberculosis status
  - CXR
- Vaccinations

Which of the following DMARDs is first choice for a newly diagnosed RA pt?

A. Hydroxychloroquine

B. Sulfasalazine

C. Methotrexate

D. Any of the biologic agents

Practice Recommendations

1. SLE – malar rash, kidneys, joints, (+) ANA/labs
2. Consider hydroxychloroquine in all patients with SLE
3. RA – small joints, (+) RF/ACPA/ESR/CRP
4. Select methotrexate (MTX) as the first line DMARD for most RA patients.


Thank you for your attention
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