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7th EMIRATES FAMILY MEDICINE SOCIETY CONGRESS 2024

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Connective Tissue Disease Update

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Learning Objectives

1. Identify the **major symptoms and risk factors for connective tissue disorders** systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), scleroderma, and dermatomyositis.
2. **Differentiate** between systemic lupus erythematosus, rheumatoid arthritis, scleroderma, dermatomyositis, and other conditions with similar symptoms.
3. Counsel patients on **treatment regimens** to manage symptoms to include first-line medications like **hydroxychloroquine for SLE and methotrexate for rheumatoid arthritis**.

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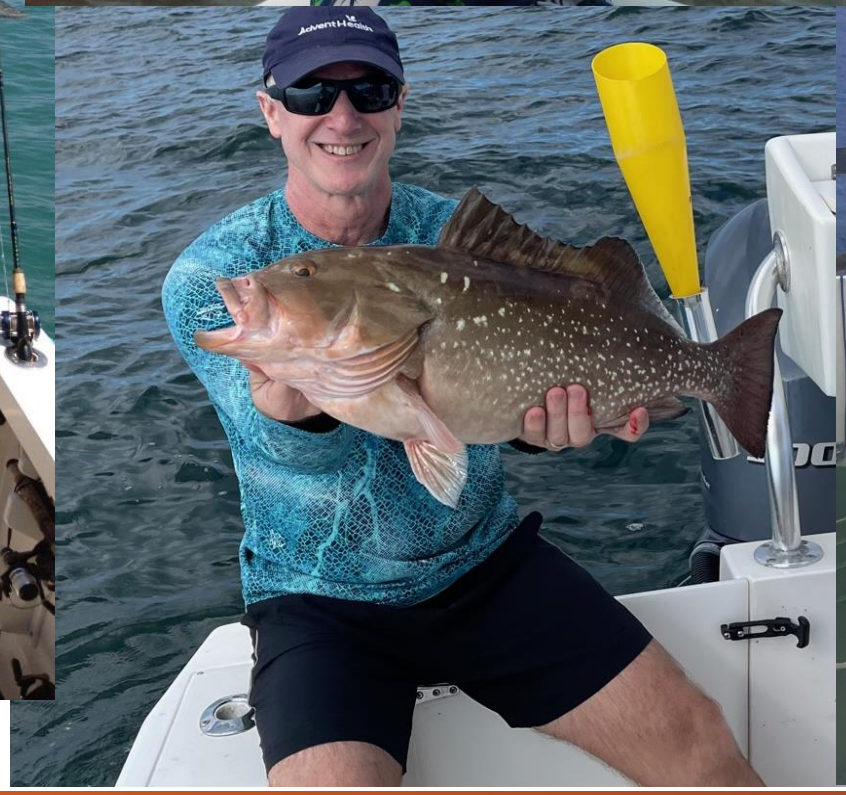
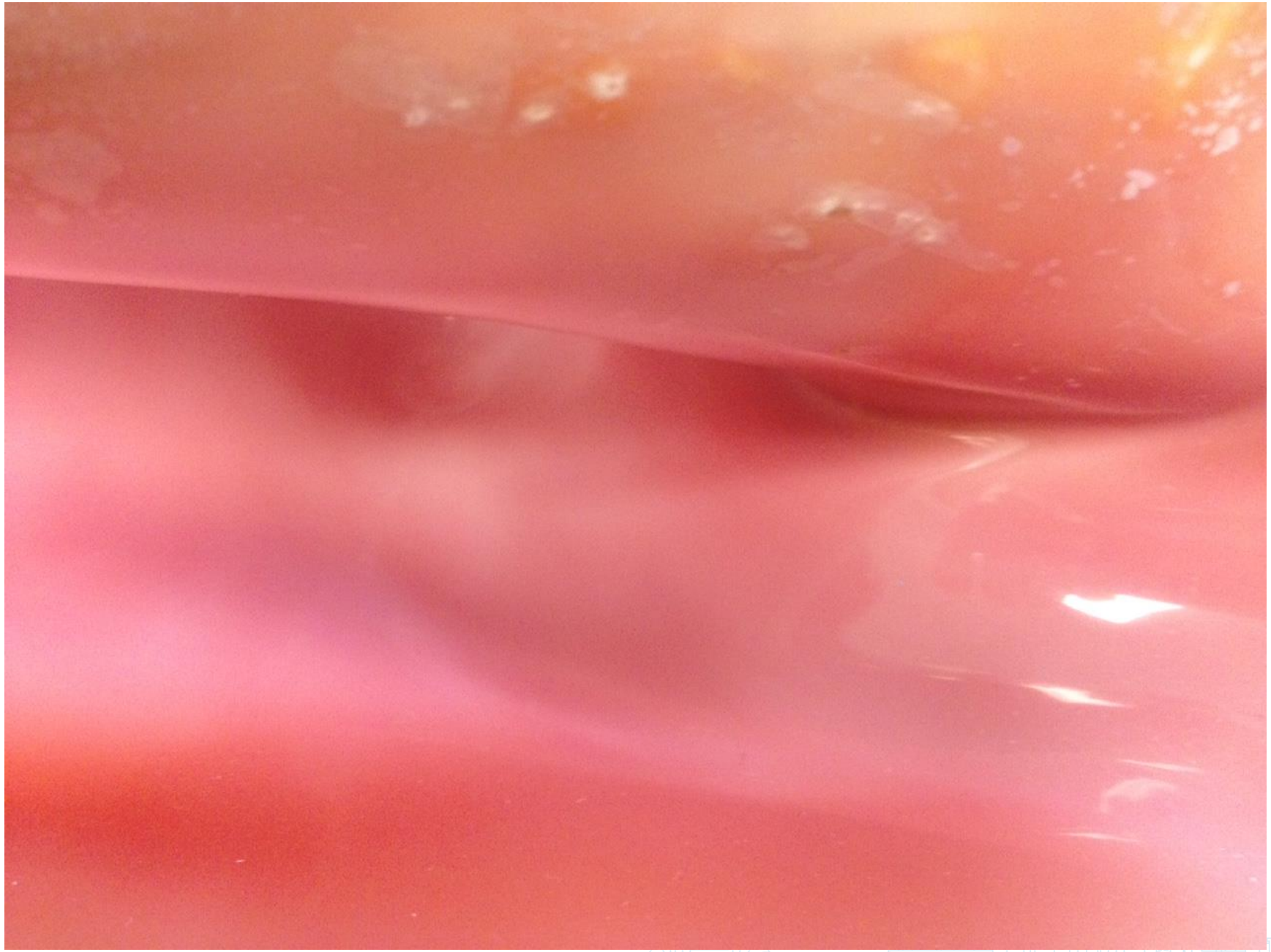
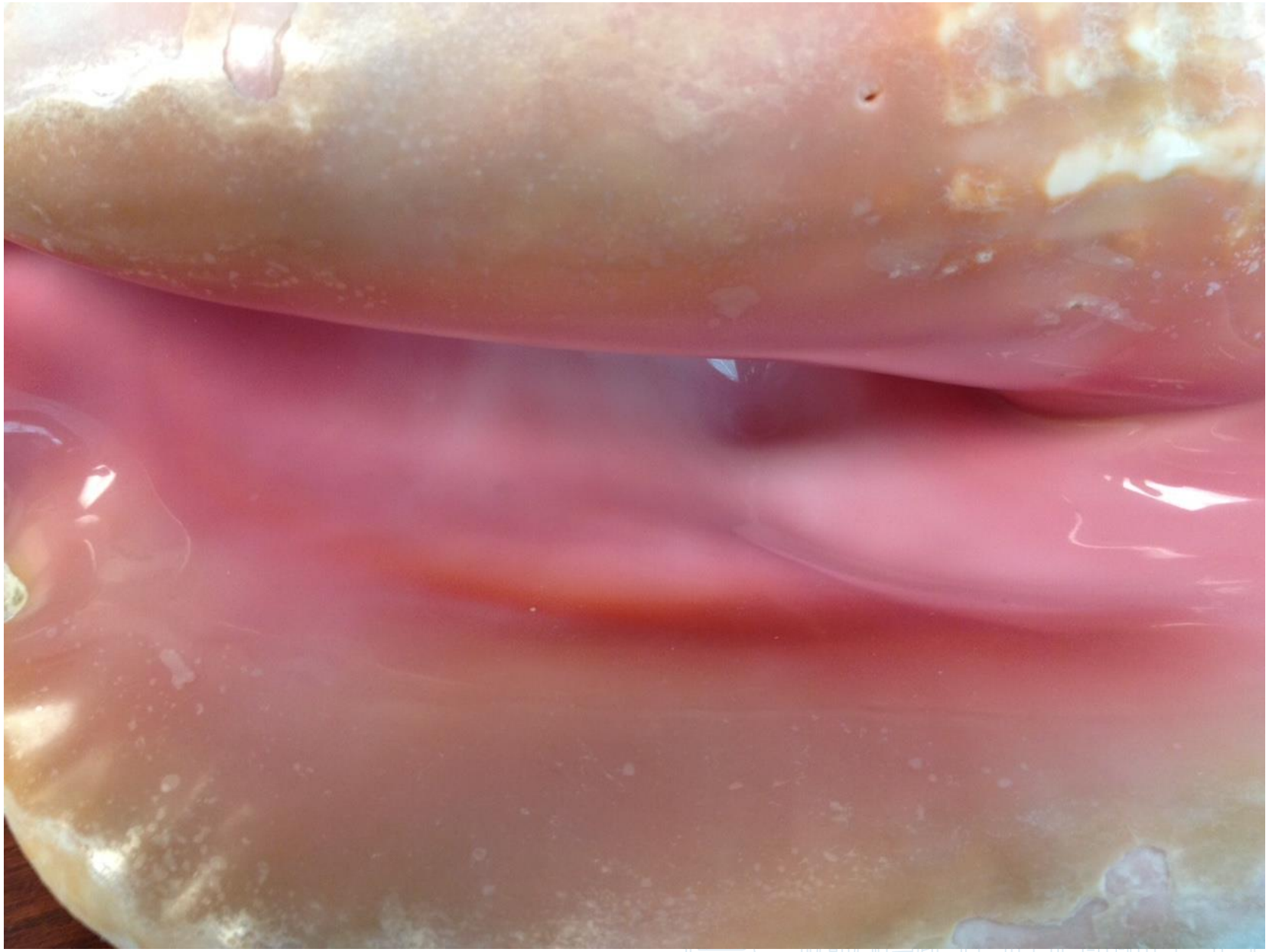


General Comments re: CTD (Connective Tissue Diseases)

- 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 7 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

AES Question

Question #1:
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

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D. Sarcoidosis

Systemic Lupus Erythematosus (SLE)

- 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

Systemic Lupus Erythematosus

- 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 3-5 fold increase over general population.
- Increased risk of osteoporosis
  - From disease and drugs (steroids)
  - 3 months of prednisone 5mg/day or more → DEXA

Systemic Lupus Erythematosus

- The diagnosis is clinical
- (+) ANA – not always helpful
- Must have 4 of 11 diagnostic criteria, ACR 1997 criteria
- 10 points - 2019 European League Against Rheumatism/ American College of Rheumatology (EULAR/ACR) criteria
- Lab tests cannot substitute for pattern recognition in the history and physical by the physician!

Criteria for Classifying Systemic Lupus Erythematosus

System	ACR criteria (1997)*	SLICC criteria (2012)†	EULAR/ACR (2019)‡

TABLE 1

Criteria for Classifying Systemic Lupus Erythematosus

System	ACR criteria (1997)*	SLICC criteria (2012)†	EULAR/ACR (2019)‡
Cardiovascular/pulmonary	Pleuritis (pleuritic pain or rub or pleural effusion) or pericarditis (documented by electrocardiography, rub, or pericardial effusion)	Serositis (pleurisy for more than one day, pleural effusion, or pleural rub; pericardial pain for more than one day, pericardial effusion, pericardial rub, or pericarditis)	Pleural or pericardial effusion (5); acute pericarditis (6)
Constitutional	—	—	Fever > 100.9°F (38.3°C) (2)
Hematologic	Hemolytic anemia, leukopenia (< 4,000 cells per mm^3), lymphopenia (< 1,500 cells per mm^3), or thrombocytopenia (< 100,000 cells per mm^3)	Hemolytic anemia; leukopenia (< 4,000 cells per mm^3) more than once or lymphopenia (< 1,000 cells per mm^3) more than once; thrombocytopenia (< 100,000 cells per mm^3)	Leukopenia (3); thrombocytopenia (4); autoimmune hemolysis (4)
Immunologic	Positive test result for antinuclear antibodies; elevated anti-dsDNA, anti-Smith, or antiphospholipid antibodies; discoid rash; photosensitivity; oral or nasal ulcers	Positive test result for antinuclear antibodies; elevated anti-dsDNA, anti-Smith, or antiphospholipid antibodies; low complement (C3, C4, or CH 50) or direct Coombs test (in the absence of hemolytic anemia); chronic cutaneous lupus, nonscarring alopecia, or oral or nasal ulcers	Anticardiolipin immunoglobulin G or anti-$beta_2$-glycoprotein 1 antibodies or lupus anticoagulant (2); low C3 or C4 (3); low C3 and low C4 (4); anti-dsDNA or anti-Smith antibodies (6)
Integumentary/mucosal	Malar rash	Acute cutaneous lupus or subacute cutaneous lupus	Nonscarring alopecia (2); oral ulcers (2); subacute cutaneous or discoid lupus (4); acute cutaneous lupus (6)
Musculoskeletal	Nonerosive arthritis involving two or more joints	Synovitis involving two or more joints or tenderness at two or more joints and at least 30 minutes of stiffness in the morning	Joint involvement (6)
Neuropsychiatric	Seizure or psychosis	Seizure, psychosis, mononeuritis complex, myelitis, or peripheral or cranial neuropathy	Delirium (2); psychosis (3); seizure (5)
Renal	Persistent proteinuria (> 0.5 g in 24 hours or > 3+ on urine dipstick testing) or cellular casts	Urinary creatinine (or 24-hour urinary protein) > 500 mg or red blood cell casts	Proteinuria > 0.5 g in 24 hours (4); renal biopsy class II or V lupus nephritis (8); renal biopsy class III or IV lupus nephritis (10)

ACR = American College of Rheumatology; anti-dsDNA = anti–double-stranded DNA; EULAR = European League Against Rheumatism; SLICC = Systemic Lupus International Collaborating Clinics.

*—ACR criteria have a sensitivity of 82.8% and a specificity of 93.4%; at least four of 11 criteria required for classification.

†—SLICC criteria have a sensitivity of 96.7% and a specificity of 83.7%; at least four of 13 criteria, including at least one clinical criterion and one immunologic criterion, are required for classification, or the patient must have had lupus nephritis confirmed by biopsy with a positive antinuclear or anti-dsDNA antibodies test result.

‡—EULAR/ACR criteria have a sensitivity of 96.1% and a specificity of 93.4%; a weighted score of 10 or more criteria is required for classification. The numbers in parentheses are the weighted score and should be added together to reach a total score.

Information from references 3, 7, and 8.

Lam NV, MD, Brown JA, Sharma R. SLE: Dx and Rx. *Am Fam Physician*. 2023;107(4):383-395

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2019 EULAR/ACR Criteria

Required ANA (+) at 1:80

10 pts = Dx

Domain	Item	Original	Modification	Revised	Simplified
Constitutional	Fever	13		13	2
Hematological	Leukopenia	12	+7	19	3
	Thrombocytopenia	26		26	4
	Autoimmune hemolysis	28		28	4
Neuropsychiatric	Delirium	12		12	2
	Psychosis	20		20	3
	Seizure	34		34	5
Mucocutaneous	Alopecia	13		13	2
	Oral ulcers	14		14	2
	SCLE/DLE	29		29	4
	ACLE	38		38	6
Serosal	Effusion	34		34	5
	Acute pericarditis	38		38	6
Musculoskeletal	Joint involvement	34	+4	38	6
Renal	Proteinuria	27		27	4
	Class II/V	55		55	8
	Class III/IV	74		74	10
APL antibodies	Anti-phospholipid	13		13	2
Complements	C3 or C4 low	19		19	3
	C3 and C4 low	27		27	4
SLE-specific antibodies	Anti-Sm	40		40	6
	Anti-dsDNA	38		38	6

Aringer M, Costenbader K, Daikh D, et al. 2019 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Systemic Lupus Erythematosus. Arthritis Rheumatol. 2019 Sep;71(9):1400-1412. doi: 10.1002/art.40930. Epub 2019 Aug 6. PMID: 31385462; PMCID: PMC6827566.

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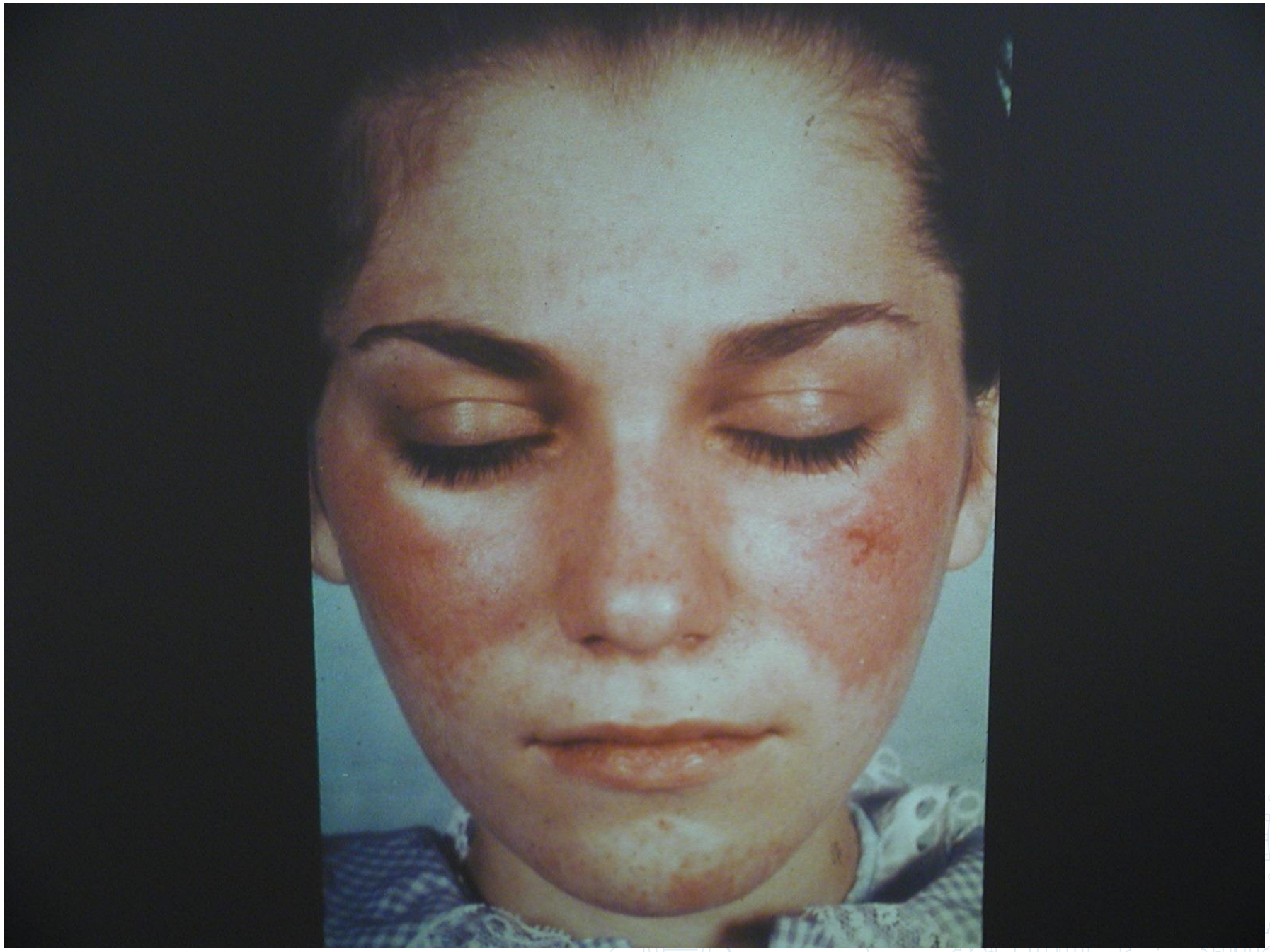
Clinical Manifestations of SLE

American College of Rheumatology

Slide Set

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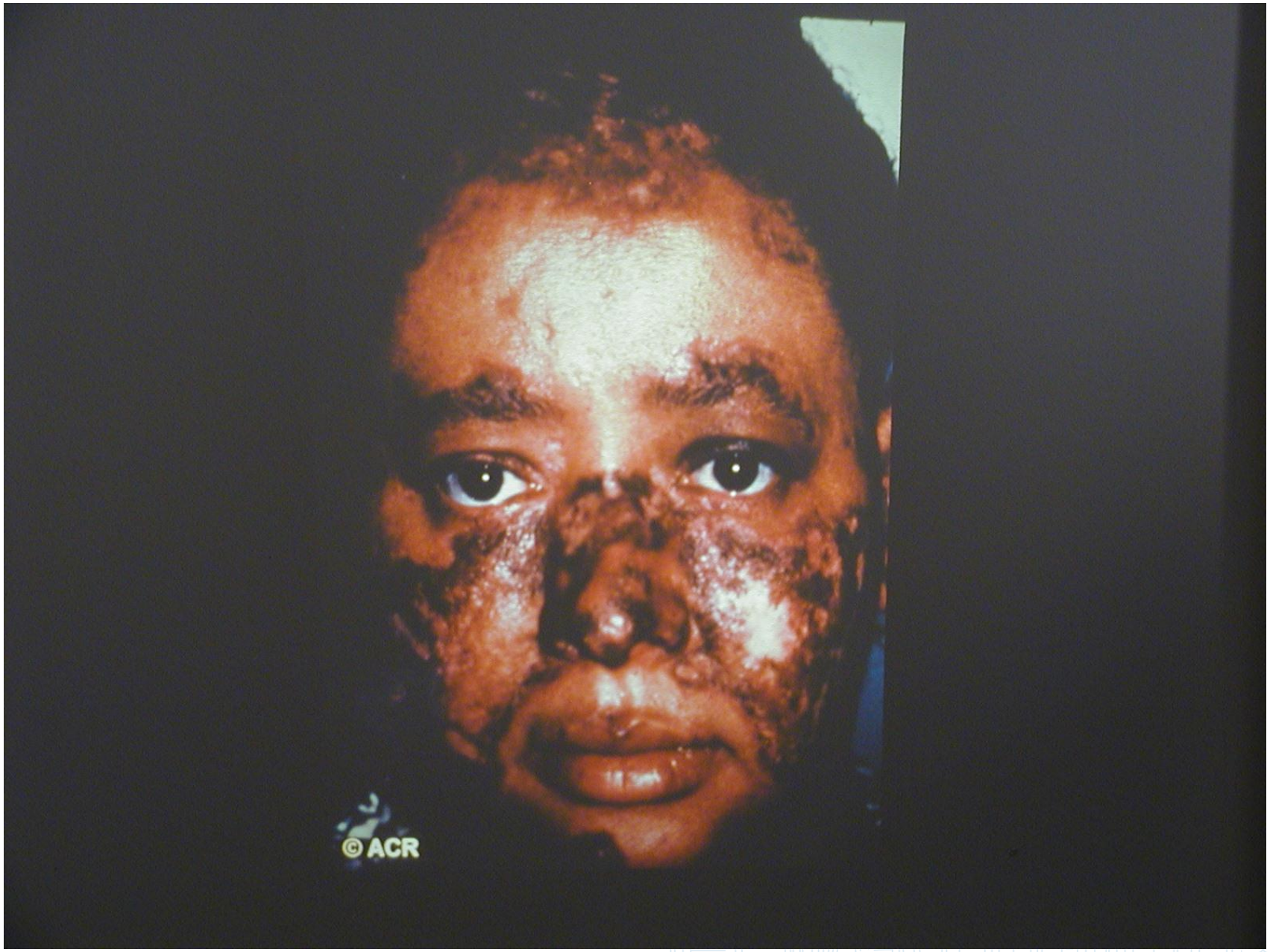
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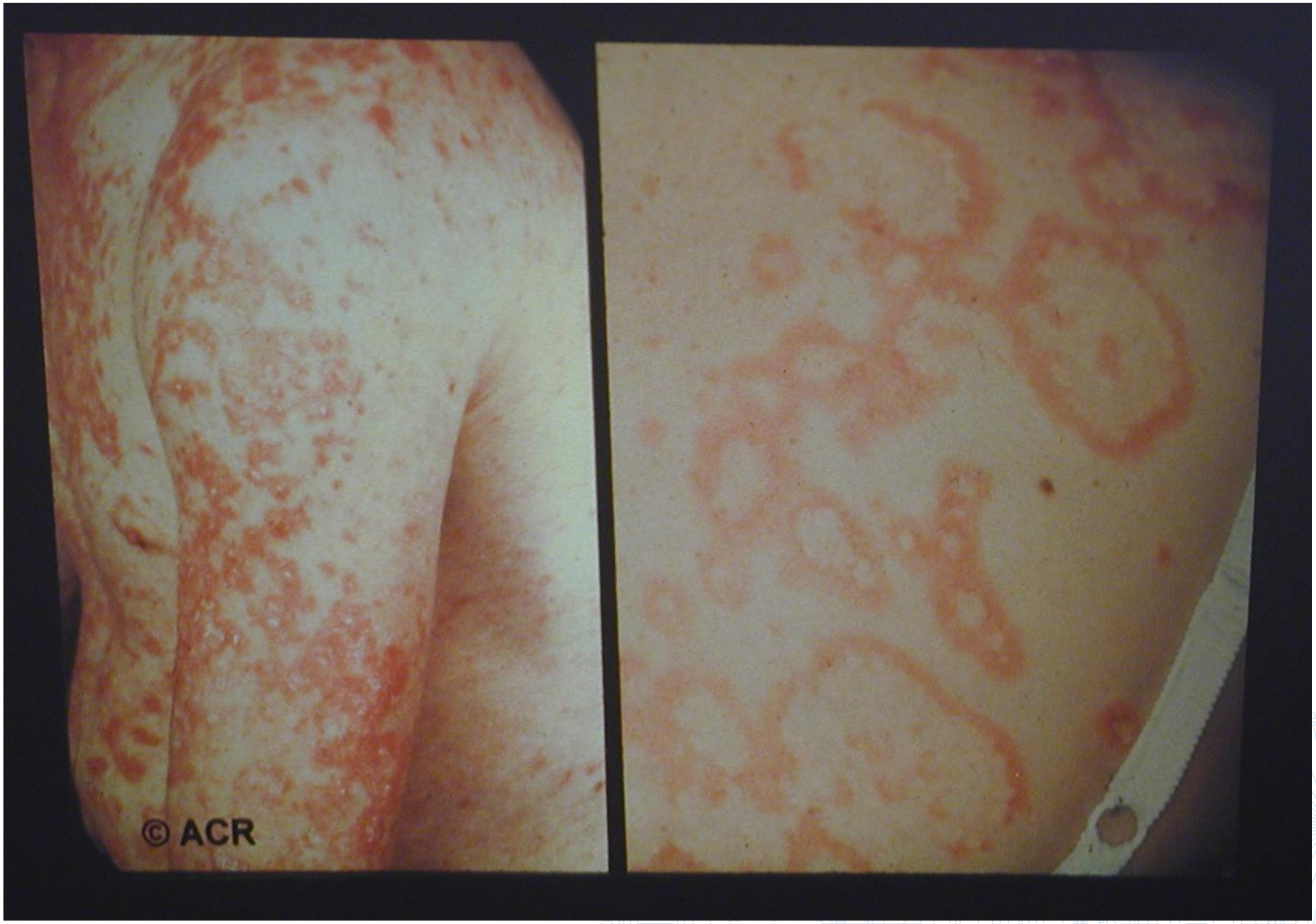
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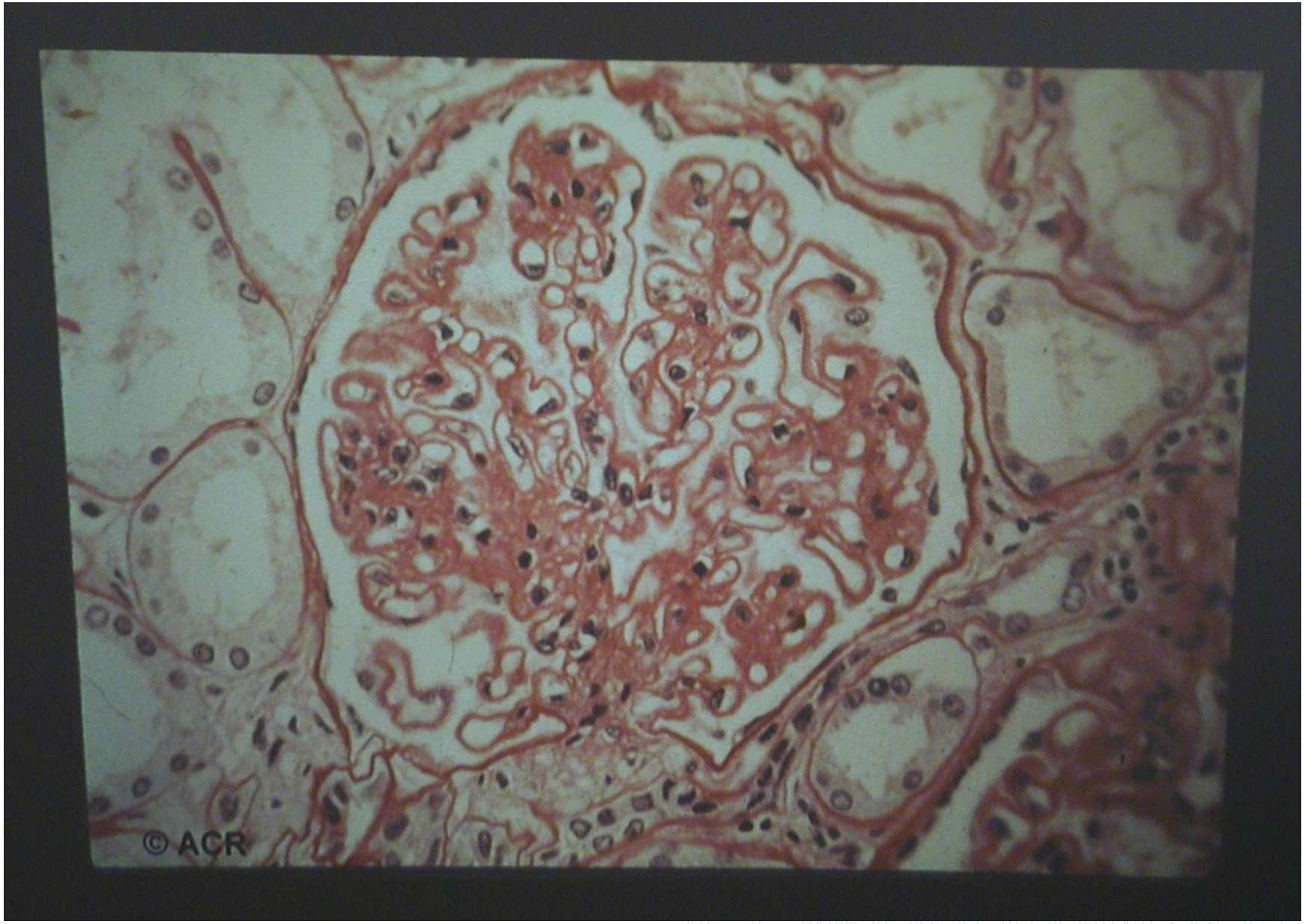
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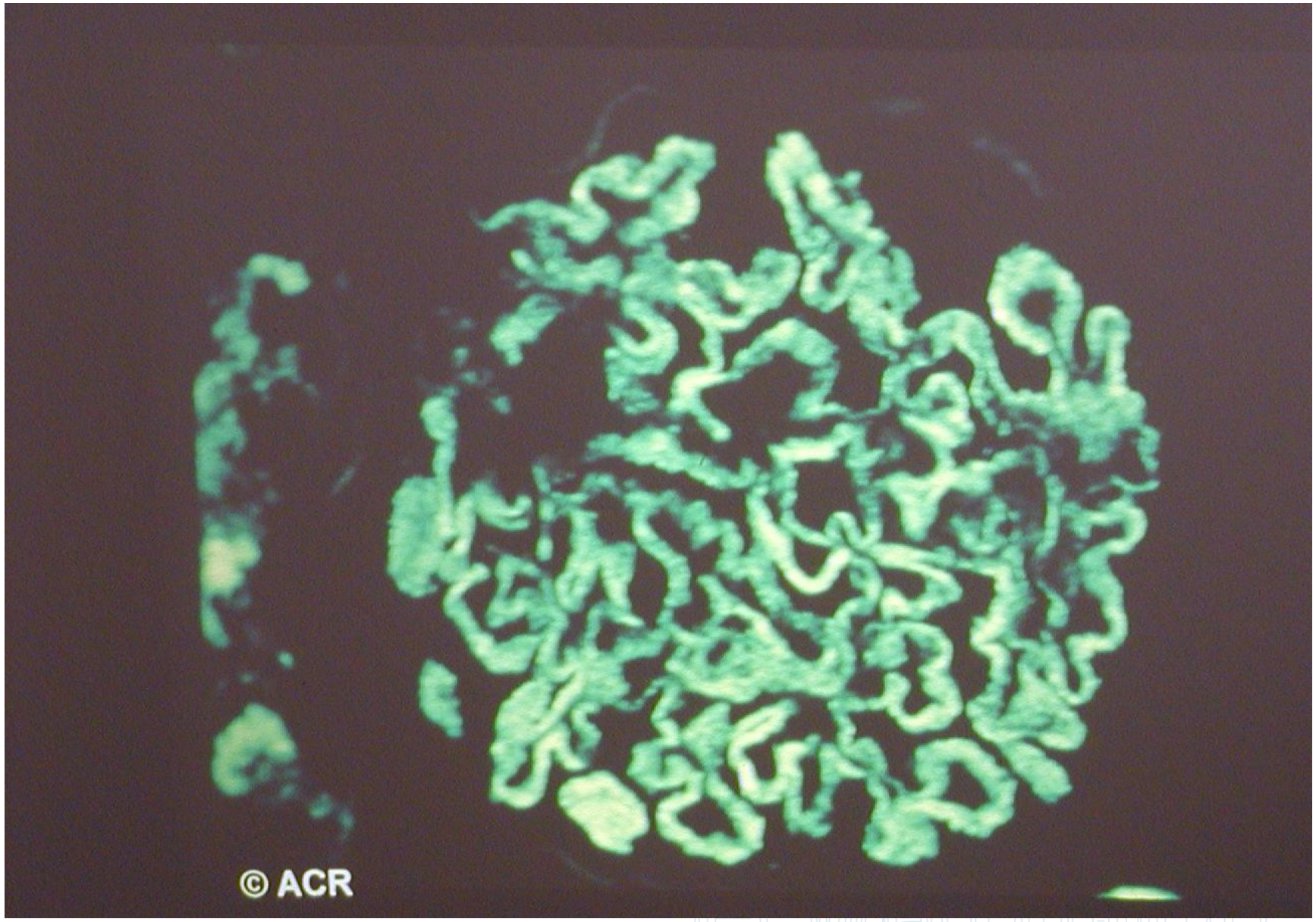
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Diagnostic Tests for CTDs

- Antinuclear antibodies (ANA)
  - SLE, scleroderma, Sjogren's, poly-/dermatomyositis, mixed CTD → ANA patterns can be suggestive
- Anti-SSA (Ro), Anti-SSB (La): SLE and Sjogren's syndrome
- Anti-Jo-1: Polymyositis
- ANCA (antineutrophilic cytoplasmic antibodies)
  - Wegener's (c-ANCA) and vasculitides (p-ANCA)
  - Confirm Wegener's with antiproteinase 3 Ab
  - Confirm vasculitis with antimyeloperoxidase Ab
- Anti-ribonucleoprotein (Anti-RNP): Mixed CTD
- Antitopoisomerase Ab (anti-Scl-70): diffuse scleroderma
- Anti-centromere antibodies
  - Limited scleroderma, CREST syndrome

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

NSAIDs and risk of GI bleed

2010 meta-analysis of relative risk of bleeding or perforation with NSAID use. Overall risk was increased fourfold

Celecoxib RR = 1.4

Ibuprofen RR = 2.2

Diclofenac RR = 3.6

Meloxicam RR = 4.2

Naproxen RR = 4.6

Indomethacin RR = 5.1

Ketoprofen RR = 5.1

Piroxicam RR = 8.0

Ketorolac RR = 14.5

Masso-Gonzalez EL, Partignani P, Tacconelli S, et al. Variability among nonsteroidal anti-inflammatory drugs in risk of upper gastrointestinal bleeding. Arthritis Rheum. 2010; 62(6);1592-1601

SLE treatment

- 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

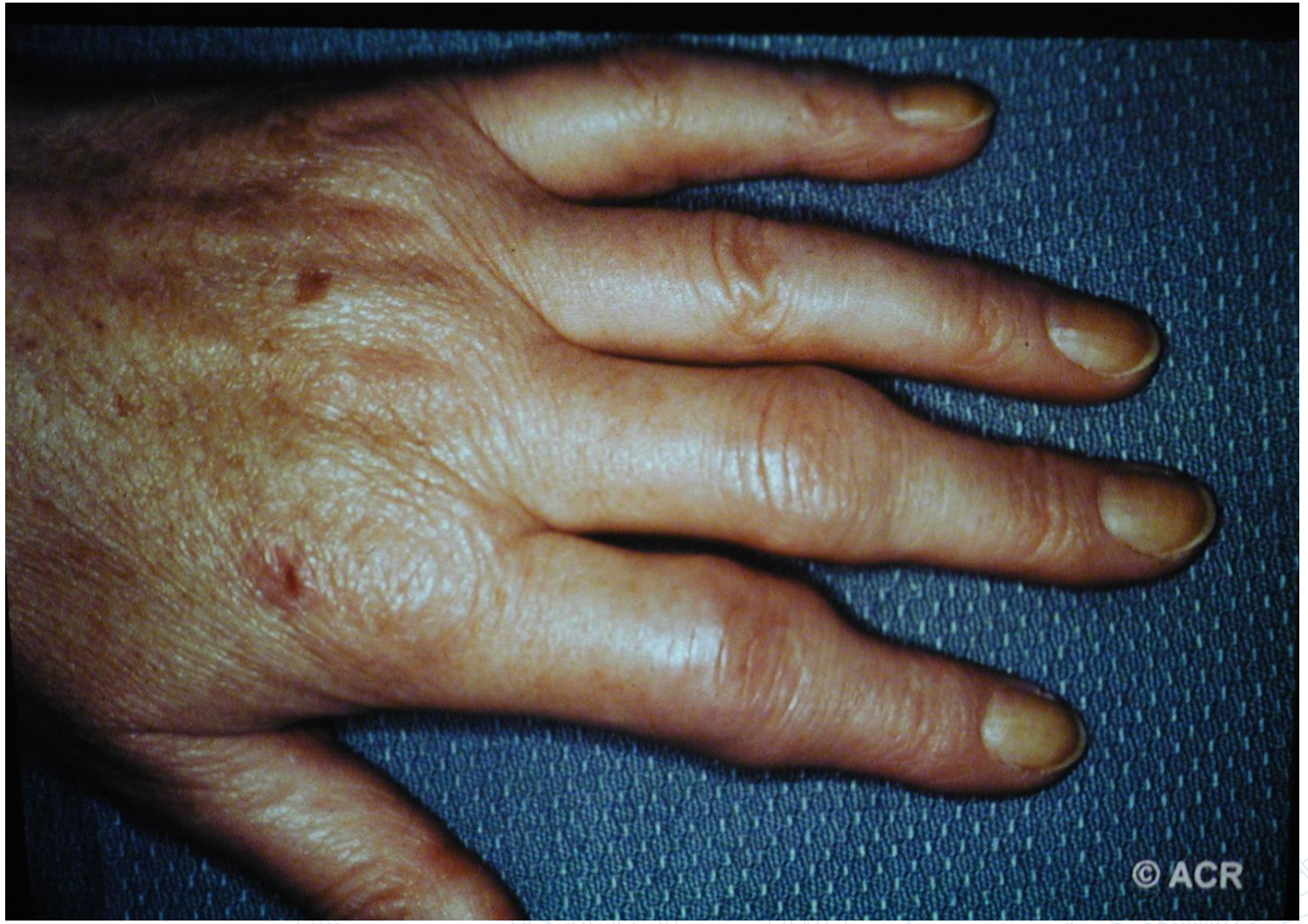
SLE treatment - Other immunomodulatory drugs

- Methotrexate – some benefit
- Mycophenolate mofetil (Cellcept)
- Cyclophosphamide
  - Historically used for lupus nephritis
- Biologics – Mab’s

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.

Rheumatoid Arthritis

Symmetric, inflammatory, polyarthritis of unknown etiology



Rheumatoid Arthritis Epidemiology

- RA is the most common autoimmune inflammatory arthritis
  - Gout is inflammatory, but not autoimmune
  - OA is arthritis but not actively inflammatory
- 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

Initial presentation

- 77% female
- Mean age 48
- Seven swollen joints and eight tender joints
- CRP elevated in 39%
- 44% had (+) RF (Rheumatoid factor)
- 39% had (+) ACPA (Anti-cyclic citrullinated protein antibody)

- Trials of patients with long-standing RA demonstrated more elevated (+) rates of RF and ACPA ~ 90%

Combe B, Benessiano J, Berenbaum F, et al. The ESPOIR cohort: a ten-year follow-up of early arthritis in France: methodology and baseline characteristics of the 813 included patients. *Joint Bone Spine*. 2007;74(5):440–445.
Keystone EC, Haraoui B, Guérette B, Mozaffarian N, Liu S, Kavanaugh A. Clinical, functional, and radiographic implications of time to treatment response in patients with early rheumatoid arthritis: a posthoc analysis of the PREMIER study. *J Rheumatol*. 2014;41(2):235–243.

Tests specific for Rheumatoid Arthritis

- 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

RA: 1987 vs 2010 criteria from the American College of Rheumatology (ACR)

- 1987 criteria
- Criteria designed to **distinguish RA from other CTDs**
- RA vs SLE
- RA vs Sjogren's syndrome
- RA vs Scleroderma

- 2010 criteria
- **Focus on early identification and early initiation of DMARD to optimize outcomes**

Diagnostic criteria : Need ≥6 points to diagnose Rheumatoid Arthritis

- Joint involvement (disease of small joints)
  - 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.

Diagnostic criteria : Need ≥6 points to diagnose Rheumatoid Arthritis

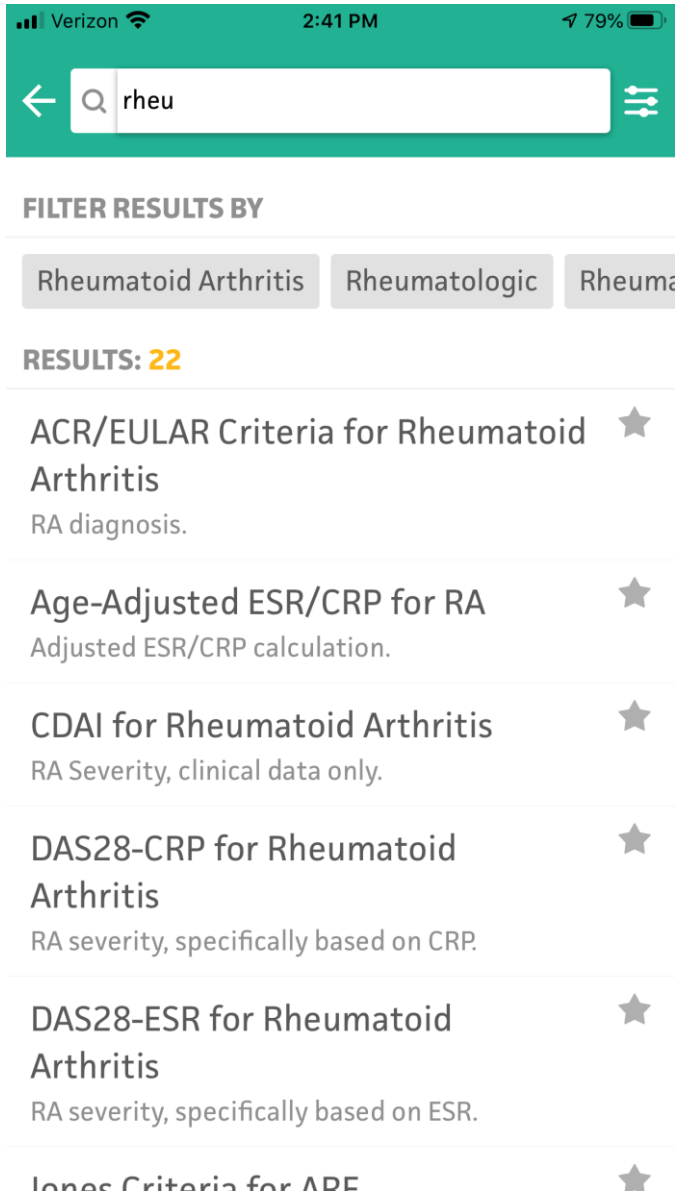
- Acute Phase Reactants
  - Normal CRP and normal ESR 0
  - Abnormal CRP or abnormal ESR 1
- Duration of symptoms
  - < 6 weeks 0
  - ≥ 6 weeks 1
- Total all points for final score
  - ≥6 points = definite RA diagnosis

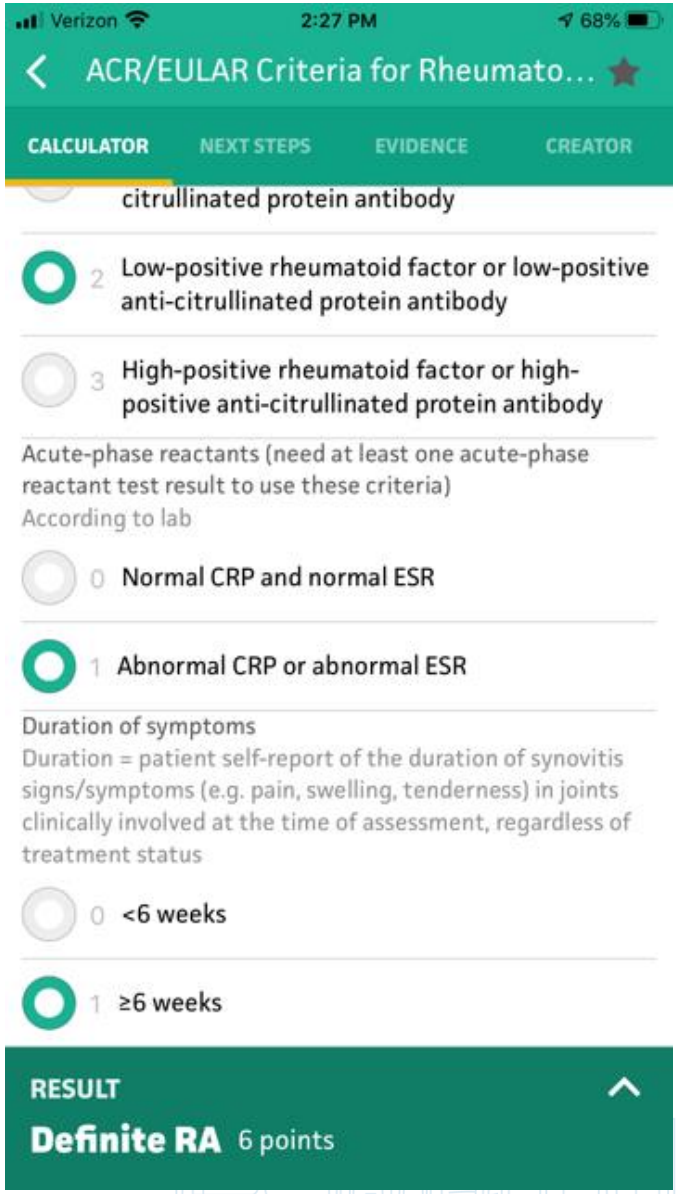
CRP = C-reactive protein; ESR = erythrocyte sedimentation rate

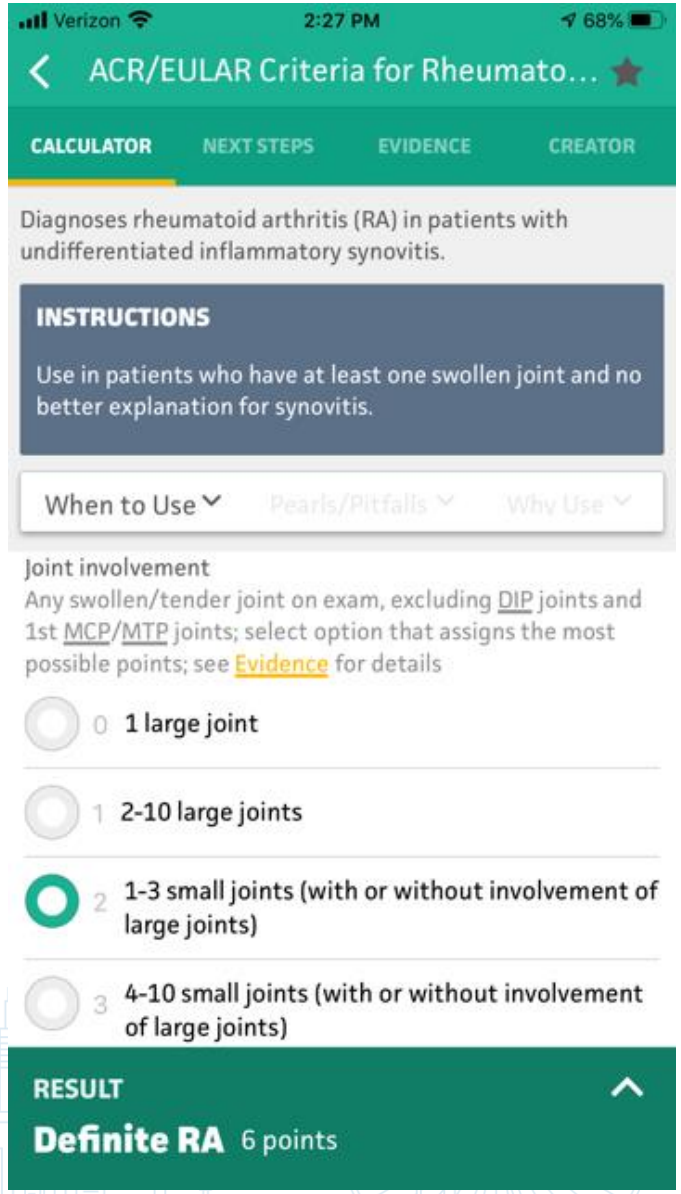
Aletaha D, Neogi T, Silman AJ, et al. 2010 Rheumatoid arthritis classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative. Arthritis Rheum 2010; 62:2569.

MDCalc App

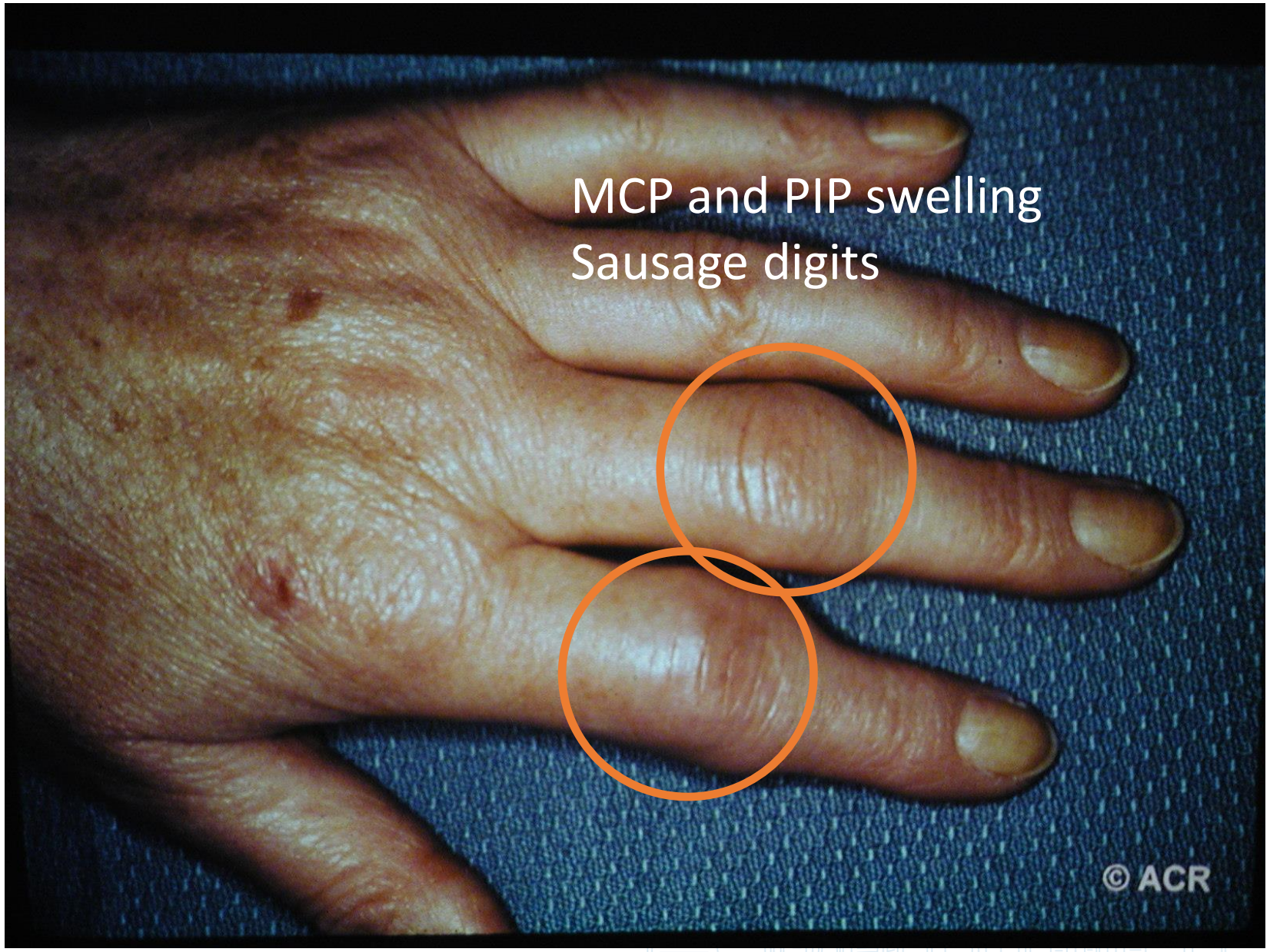
"Work Smarter, Not Harder"

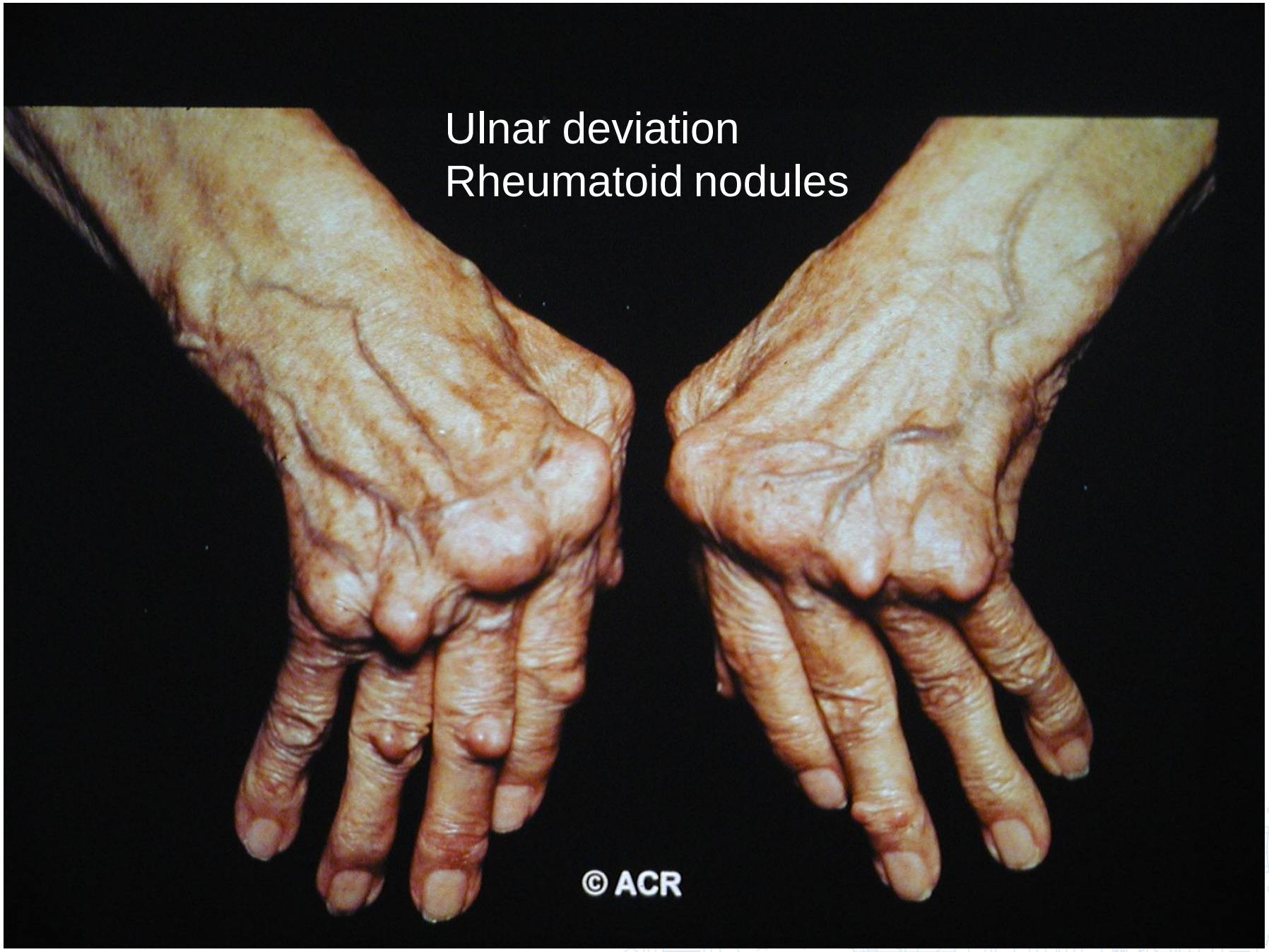


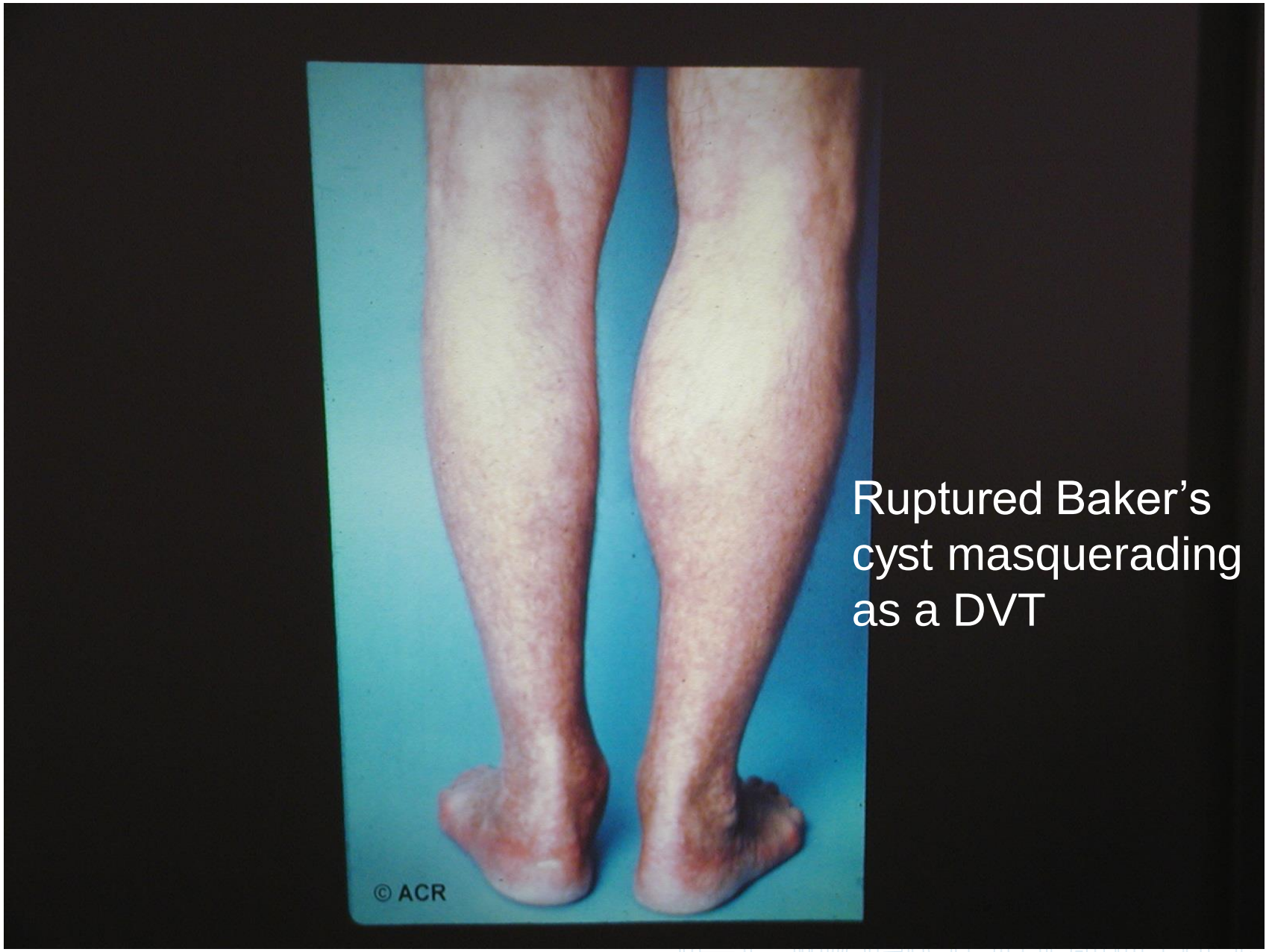




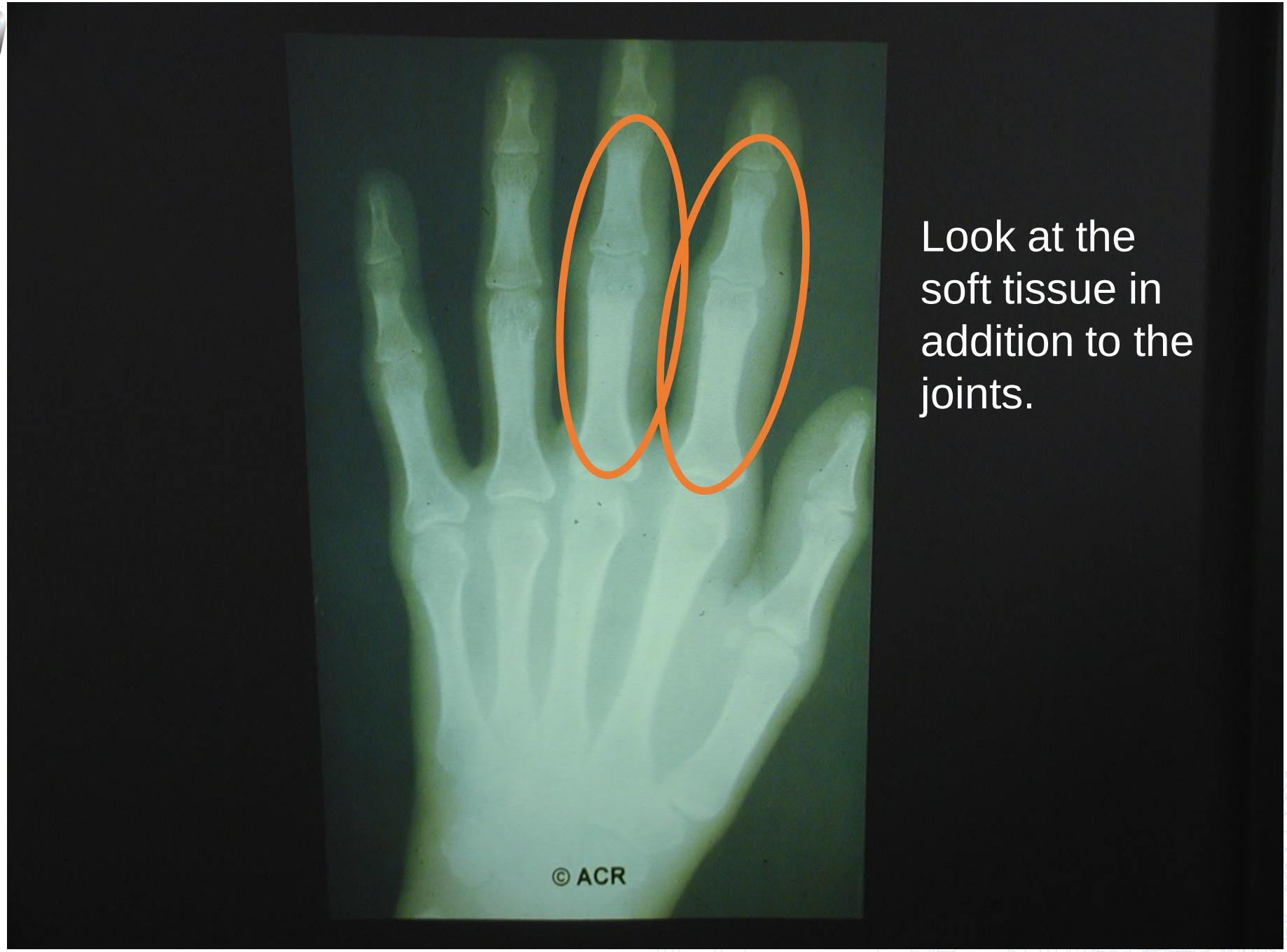
Clinical Findings in Rheumatoid Arthritis


MCP and PIP swelling
Sausage digits
© ACR


Ulnar deviation
Rheumatoid nodules
© ACR

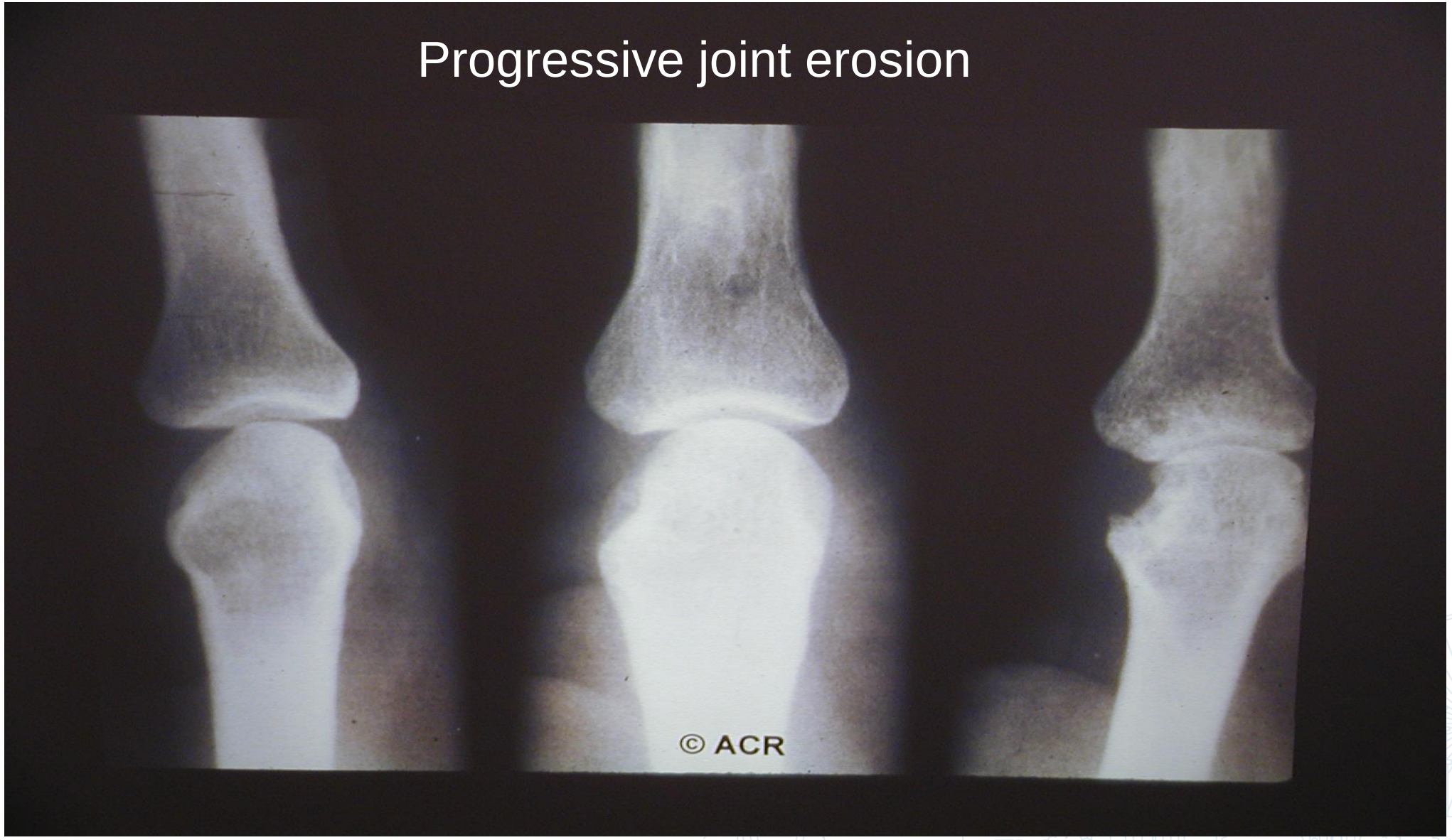


Ruptured Baker's cyst masquerading as a DVT

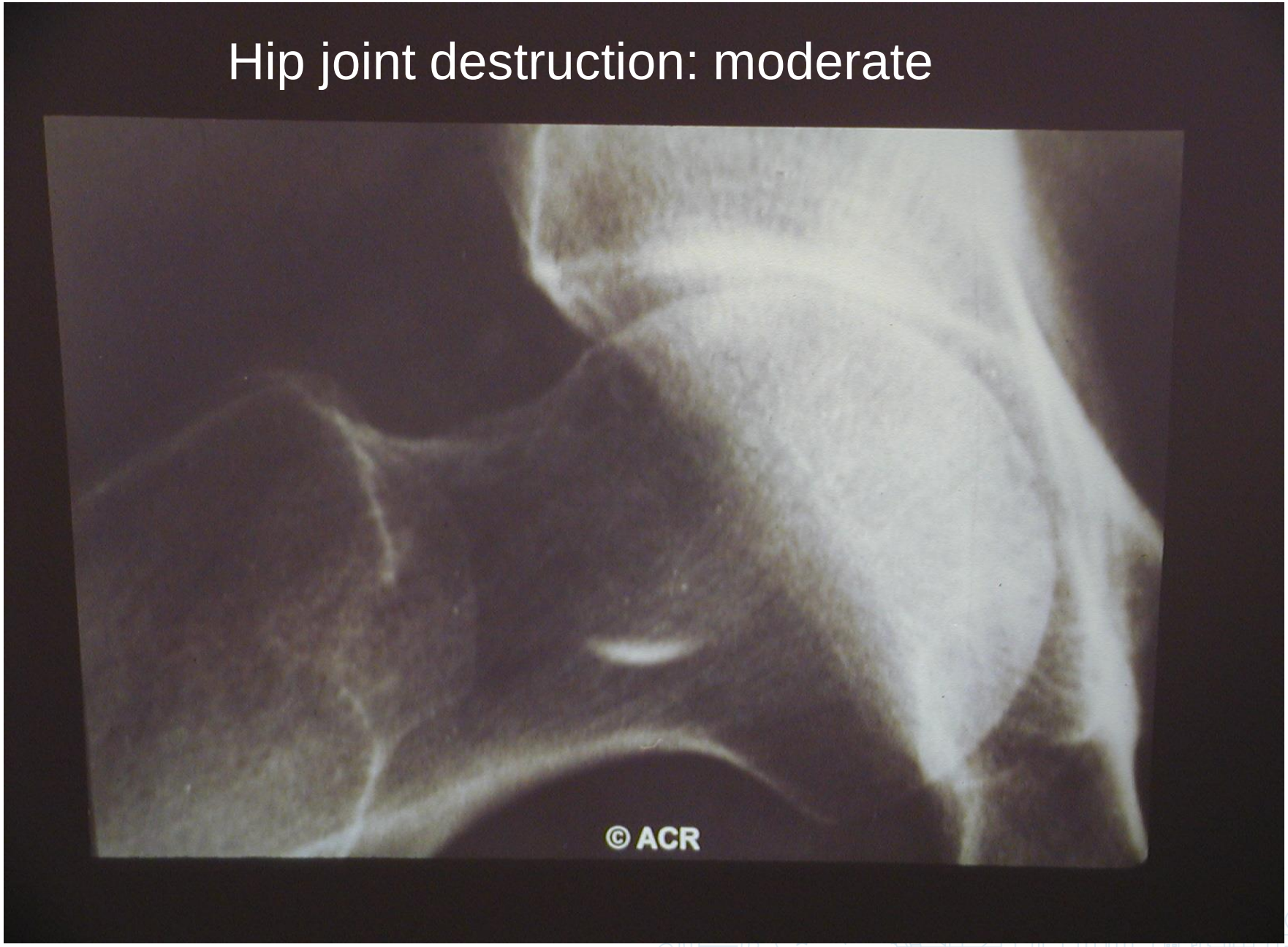


Look at the soft tissue in addition to the joints.

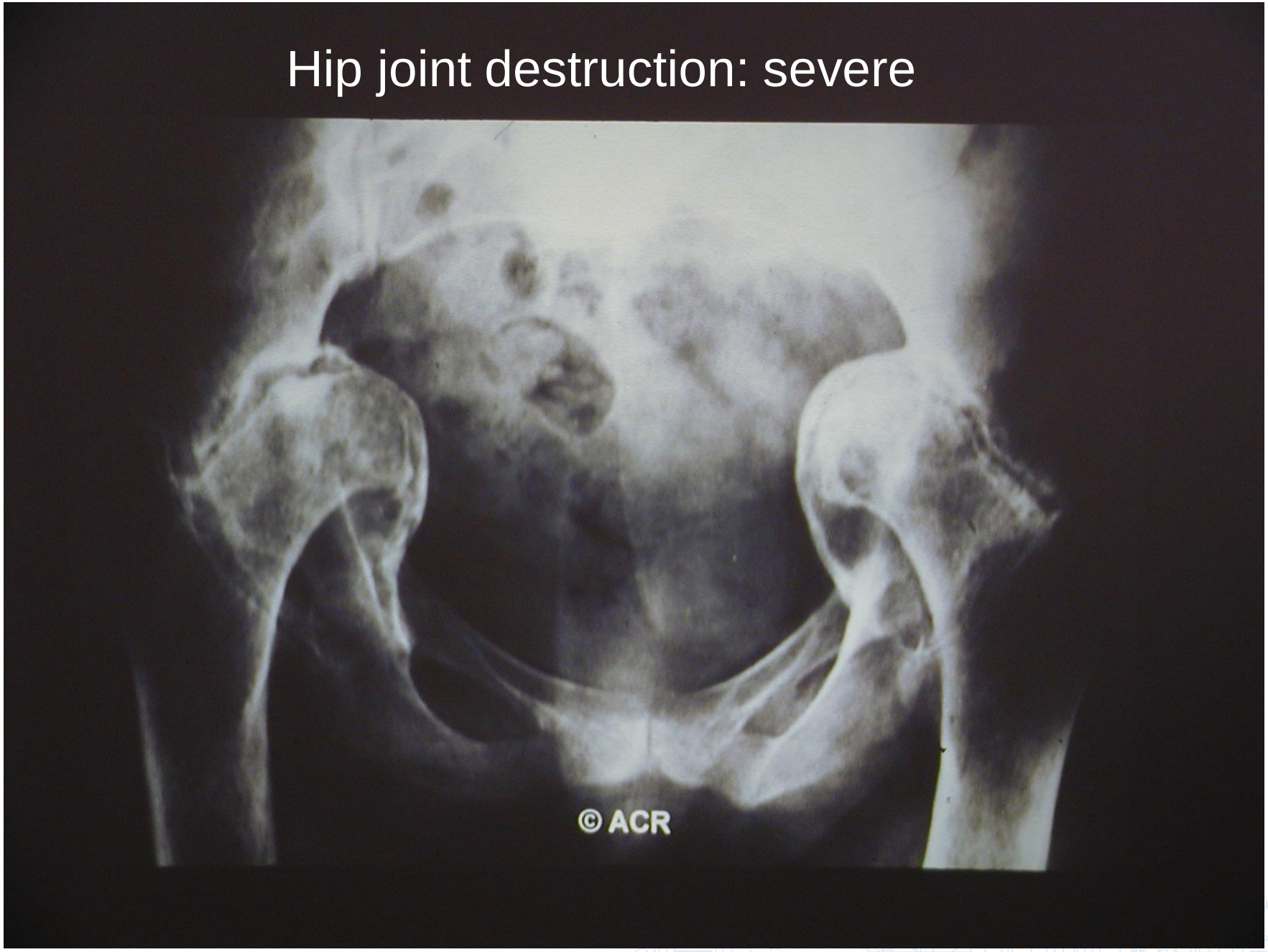
Progressive joint erosion

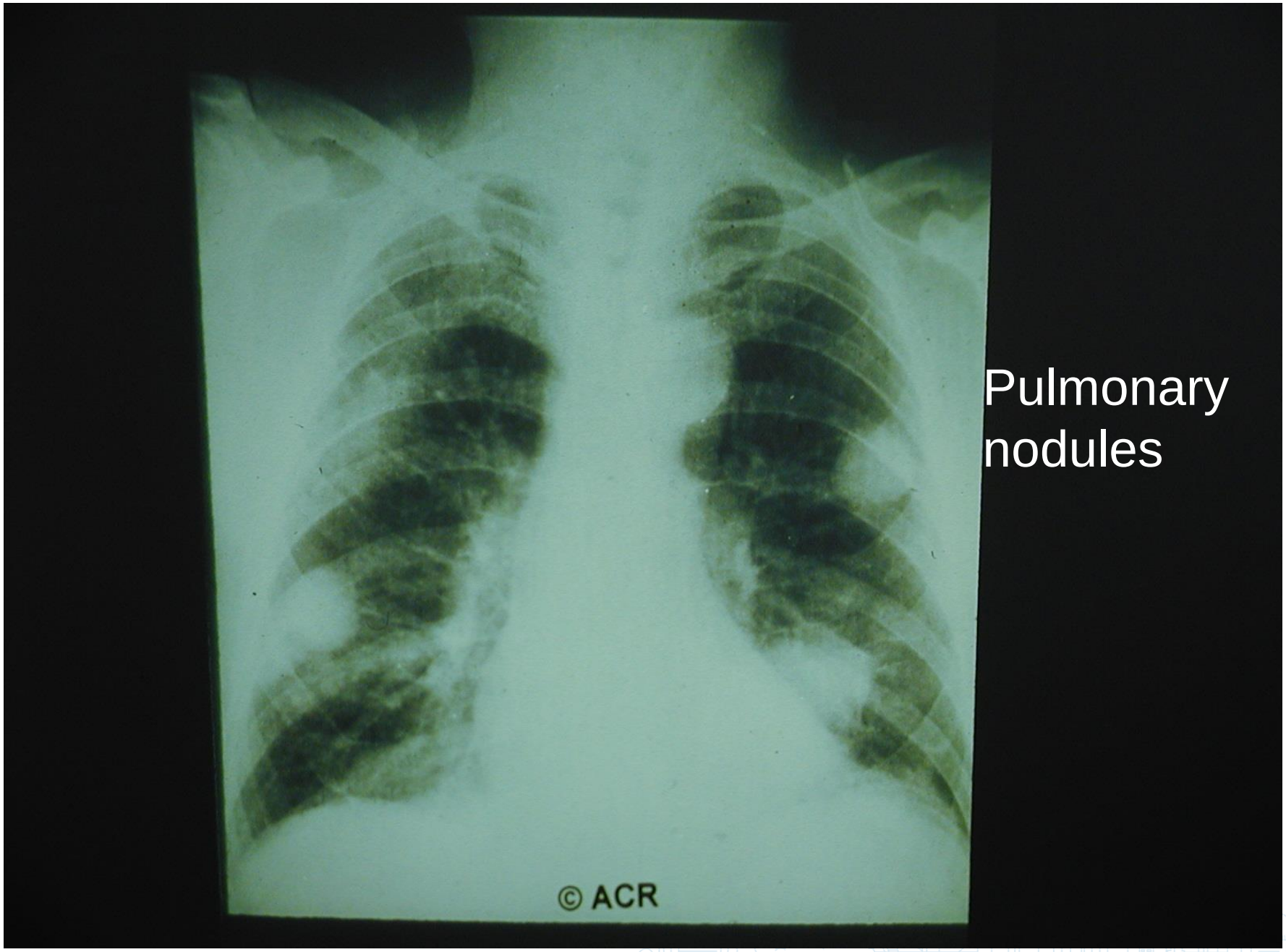


Hip joint destruction: moderate



Hip joint destruction: severe





Pulmonary nodules

Exam hints

- When assessing a joint, look for swelling, tenderness and warmth (STW)
- RH 2F MCP – S2 T2 W1; 3F MCP – S2 T3 W1; 2F PIP – S3 T2 W2
- Scale 0-3
- Attempt to quantify physical exam findings
- Can then compare future exams with baseline exam
- “Make a fist” – marked inflammation of MCPs and PIPs accentuated
- Feel for the ulnar styloid – common place for inflammation/erosion

Extra-articular manifestations
RA is *Systemic Inflammation*

- Assess lungs – pulmonary nodules, interstitial lung disease/fibrosis
- Assess the C1/C2 joint, especially pre-op before the anesthesiologist extends the neck for intubation
- Occasion pericarditis with RA

Treatment for RA

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

AES Question

Question #2

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

Question #3

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

A. Hydroxychloroquine

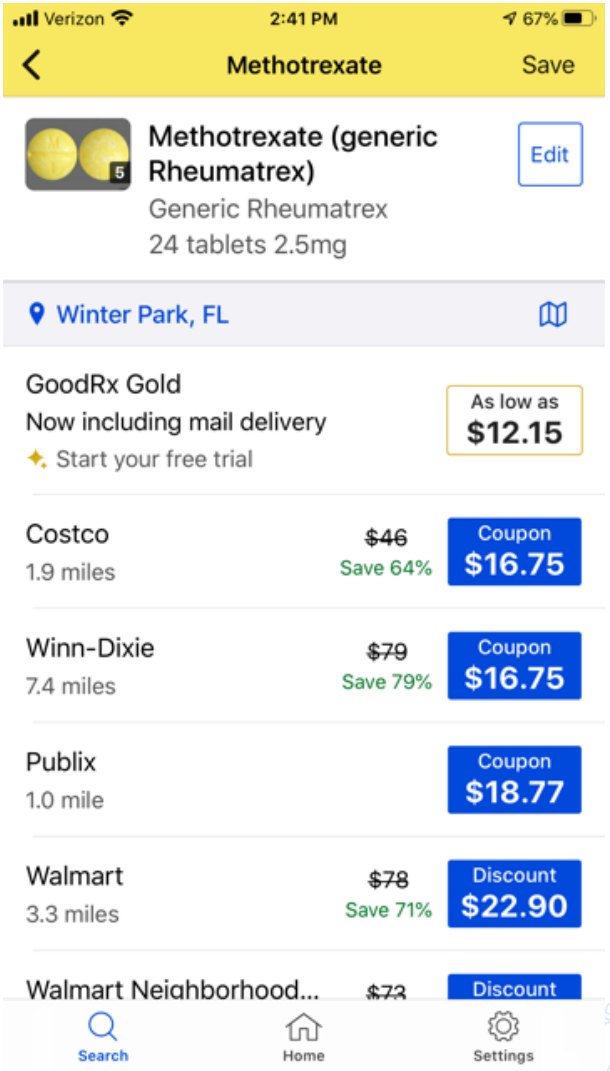
B. Sulfasalazine

C. Methotrexate

D. Any of the biologic agents

Treatment for RA

- Acute treatment
  - NSAIDs and Glucocorticoids
- Chronic – DMARDs
  - Nonbiologic
    - **Methotrexate – best choice**
    - Sulfasalazine, hydroxychloroquine, leflunamide
  - Biologic
    - TNF inhibitors:
    - Etanercept, adalimumab, others



For a newly diagnosed patient or RA patient having a flare

- Consider NSAIDS for initial treatment
- Reassess in 2-4 weeks
- Consider starting or switching to oral steroids for acute flare.
  - Prednisone at 5-20 mg daily
- May need to taper.

- General opinion: taper steroids when using for more than 2-3 weeks.
- In chronic RA patient, may taper by 1 mg prednisone per day over one month at a time.

Scleroderma
(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 (1:10,000)
- 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

Scleroderma:
Two forms of the disease

- Limited
  - Morphea – localized thick, hardened skin patches
  - Linear scleroderma – bands of hard skin
  - CREST syndrome
  - Systemic sclerosis sine scleroderma
- Diffuse

CREST syndrome

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

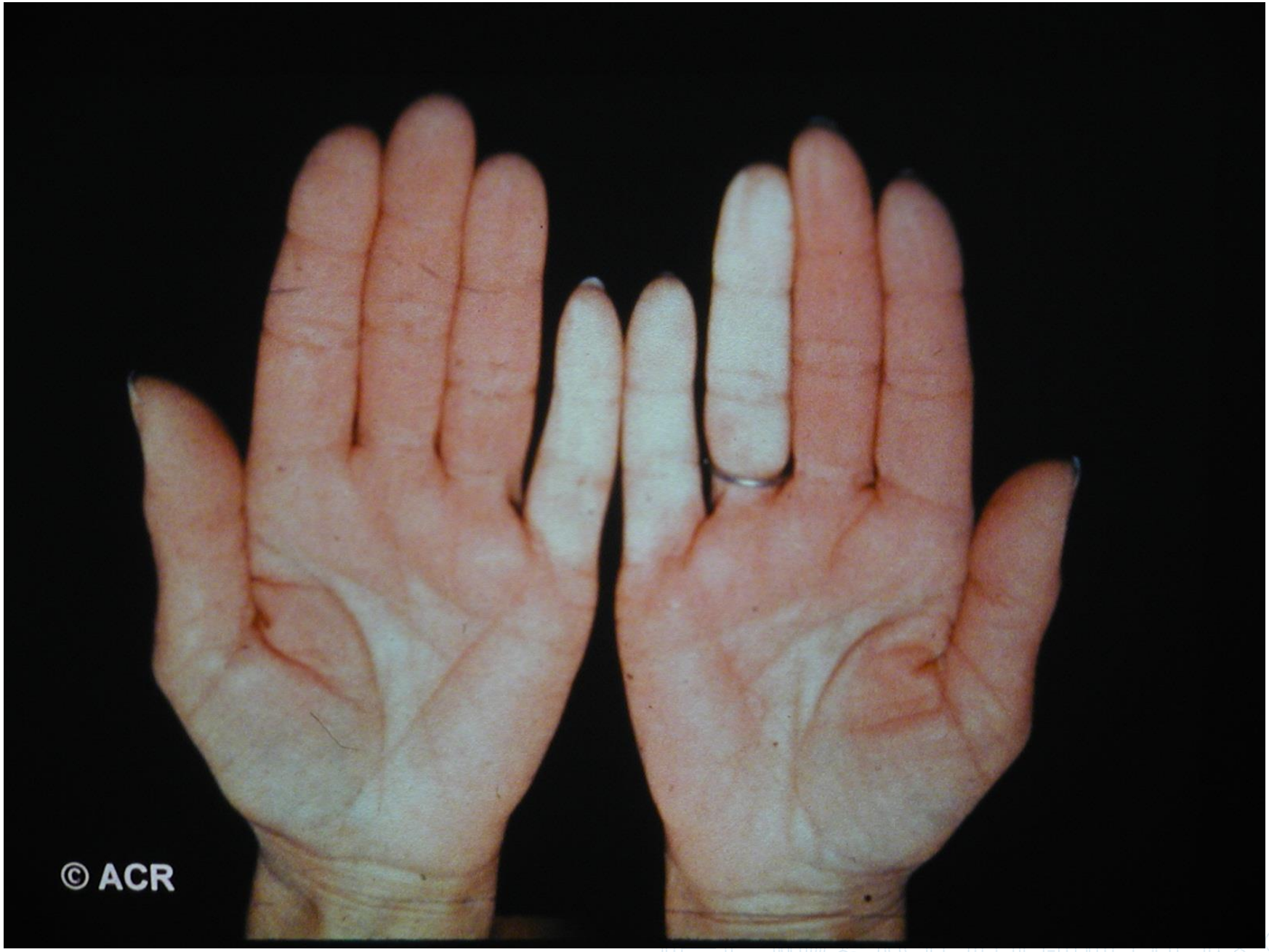
Lab testing

- 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 – look for RV hypertrophy (Pulm HTN)
- Echocardiogram – look at PA pressures (Pulm HTN)
- PFTs: +/- DLCO to check for fibrosis

Clinical manifestations of Scleroderma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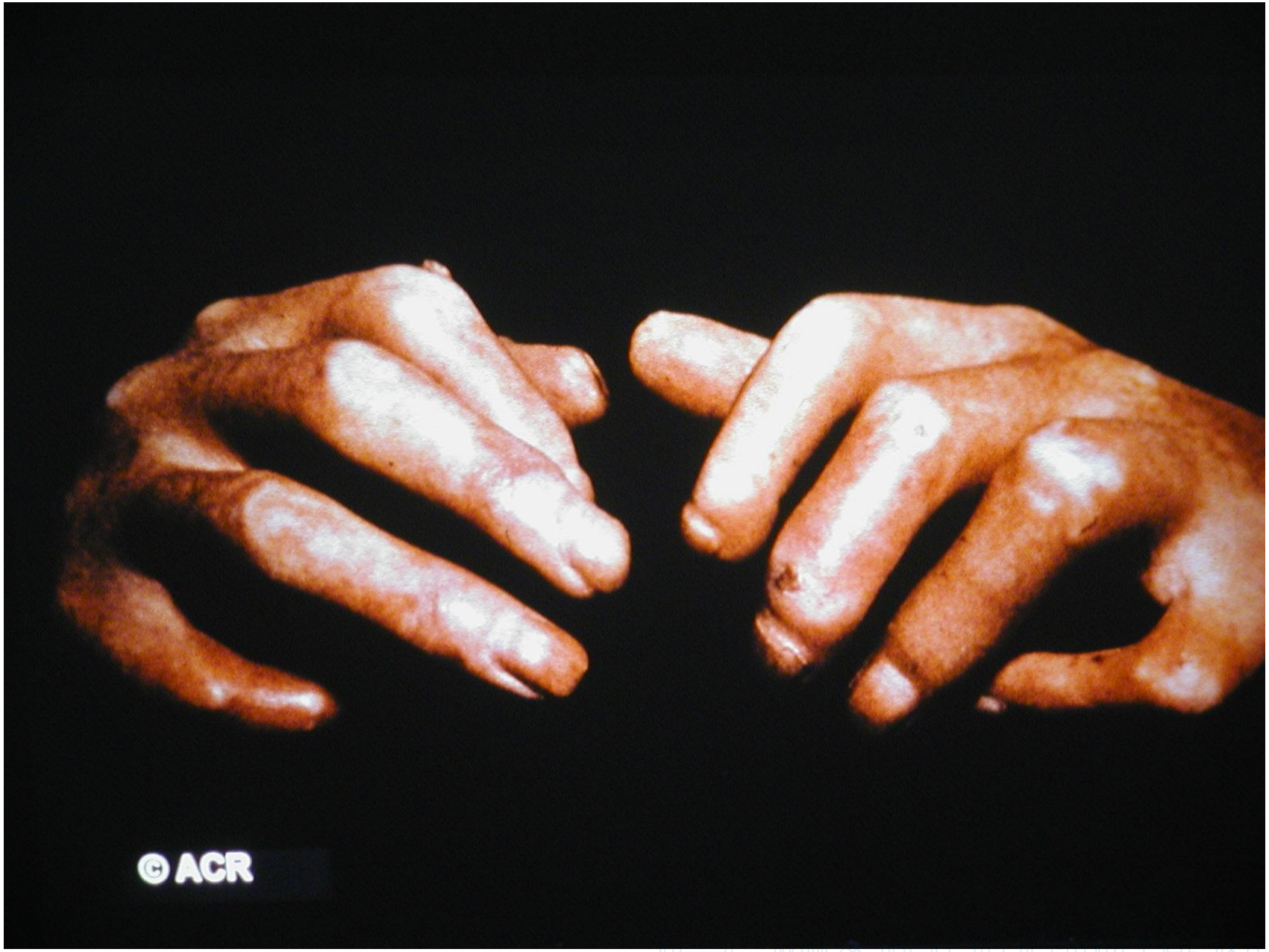
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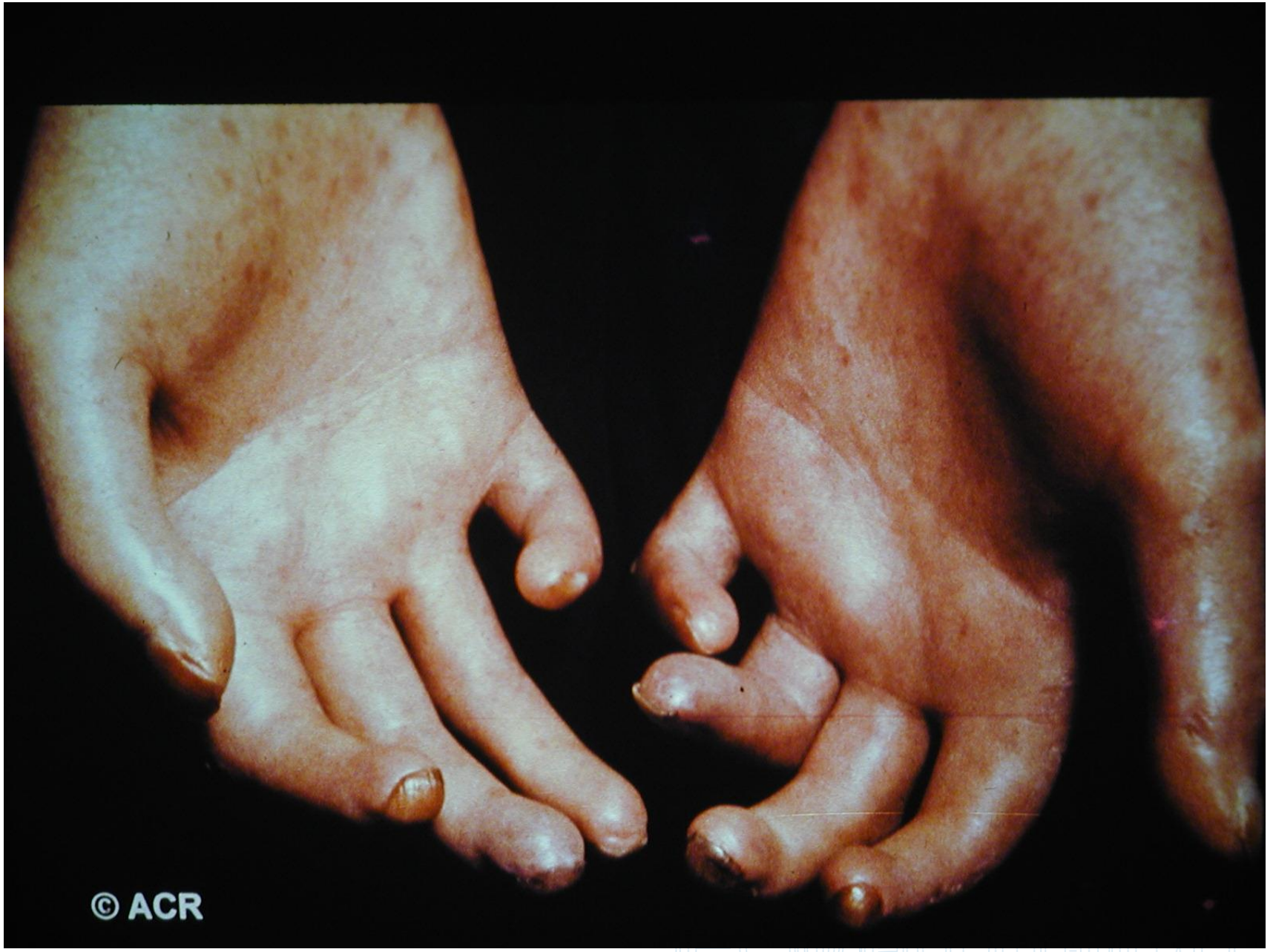
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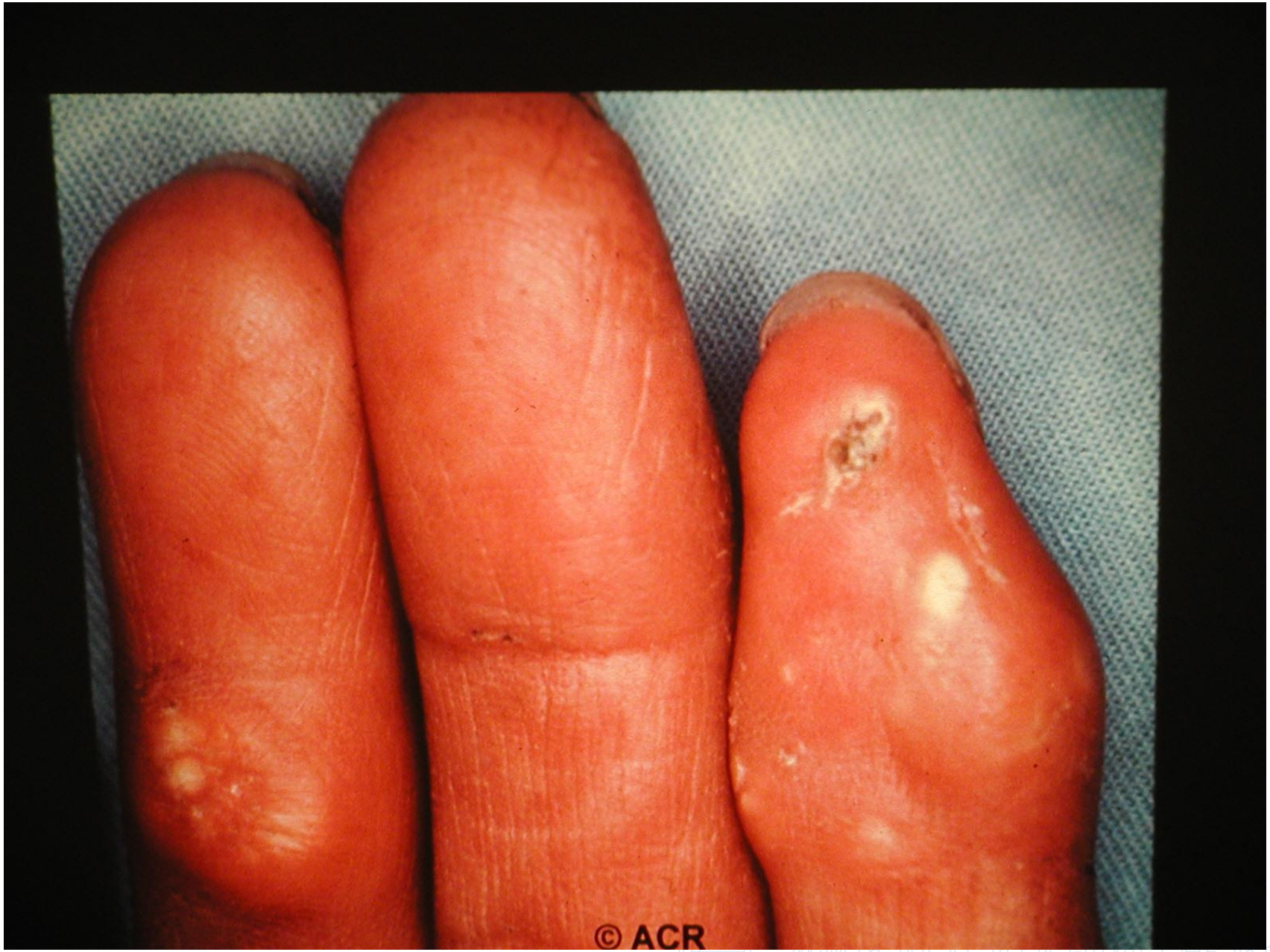
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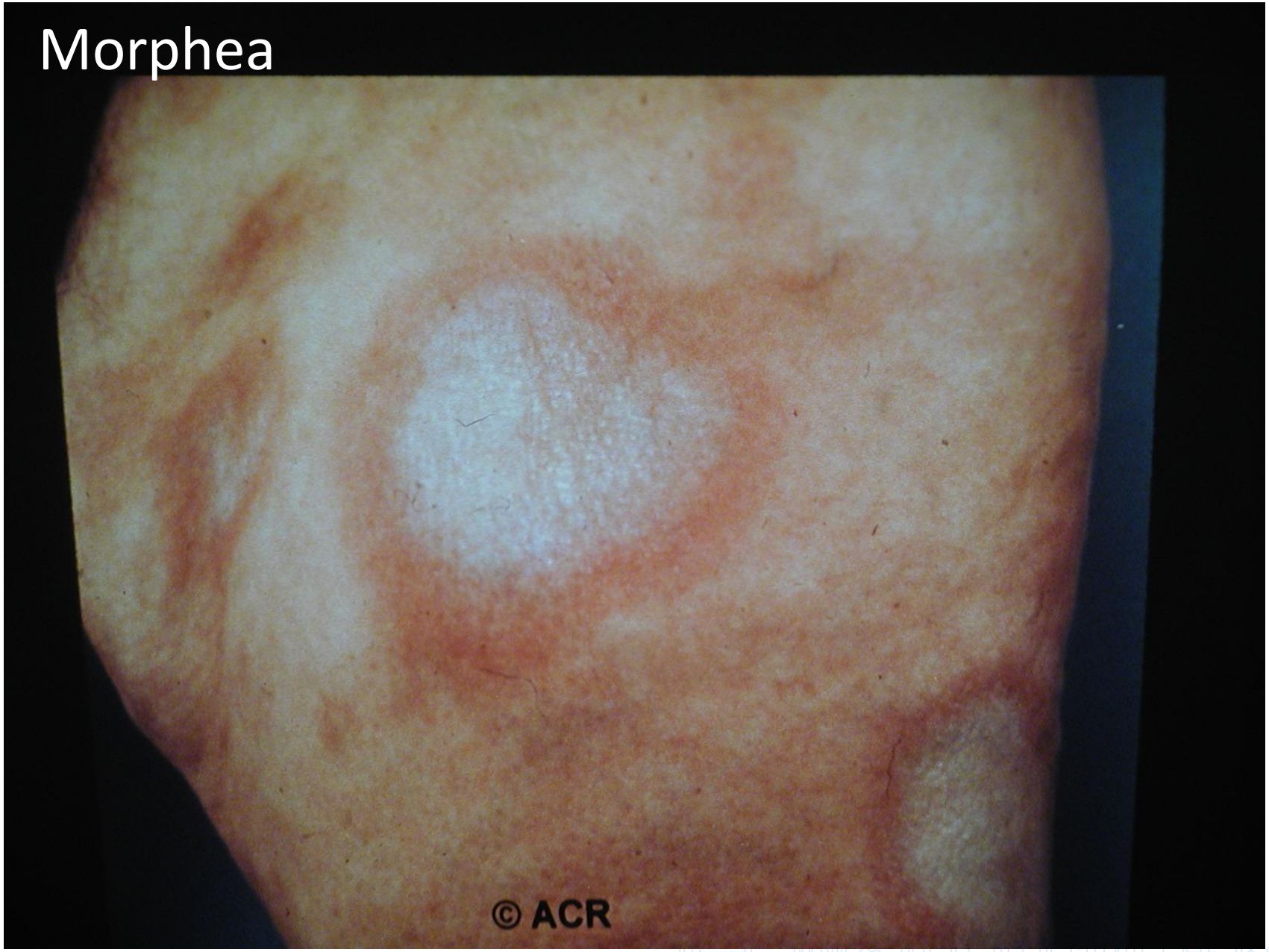
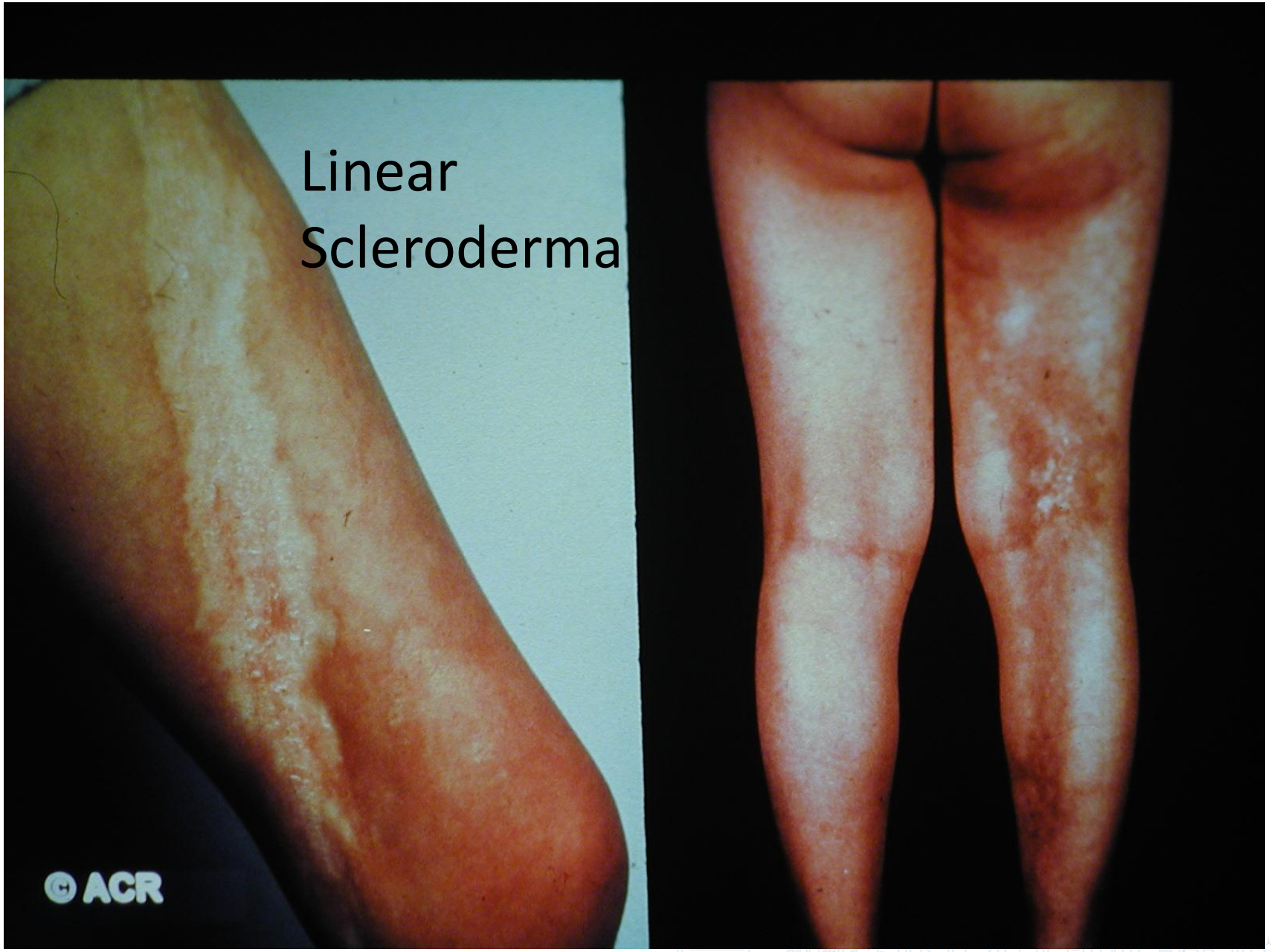
Mauskapf
Mouse Face

Morphea




Linear
Scleroderma
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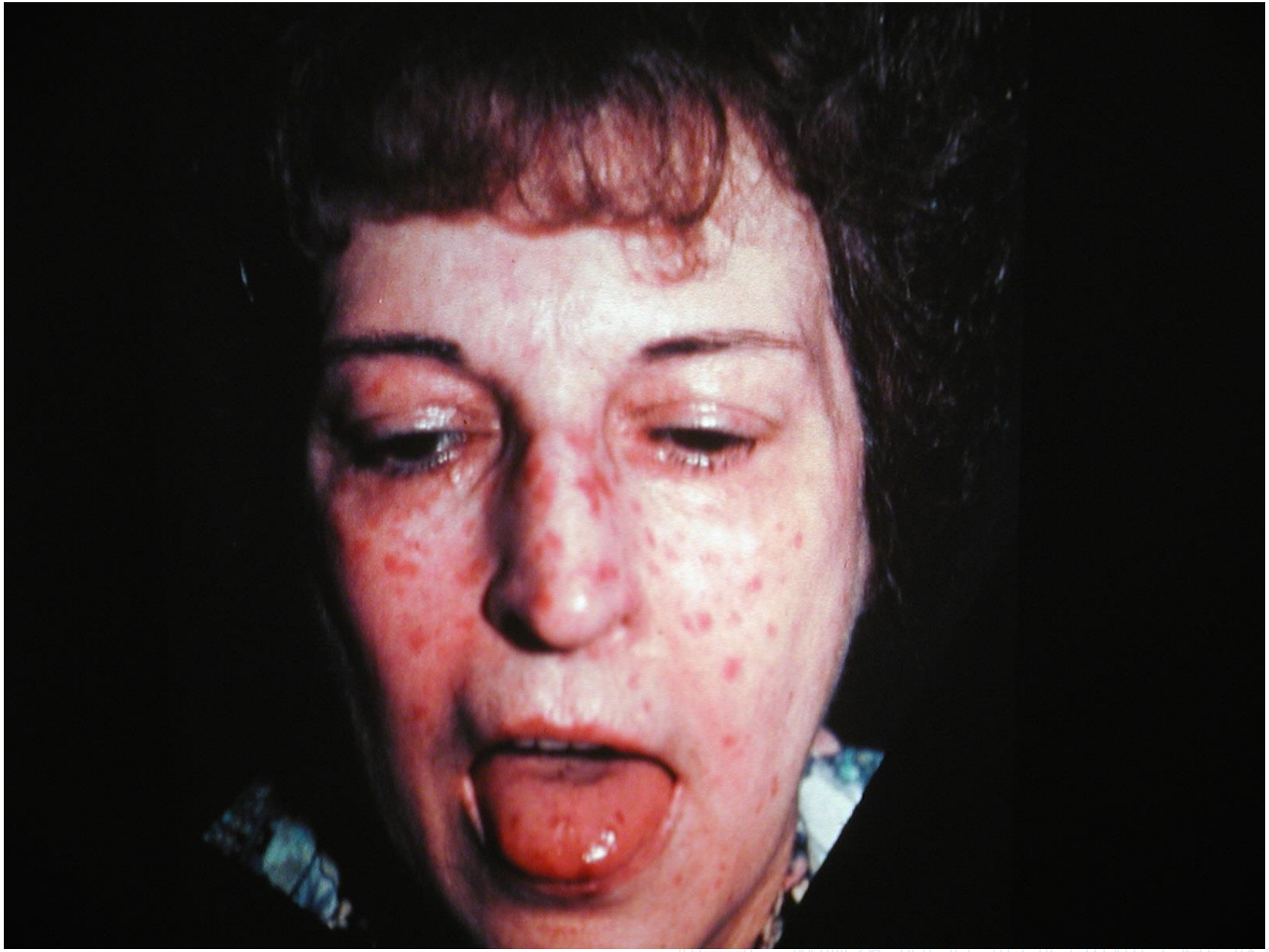

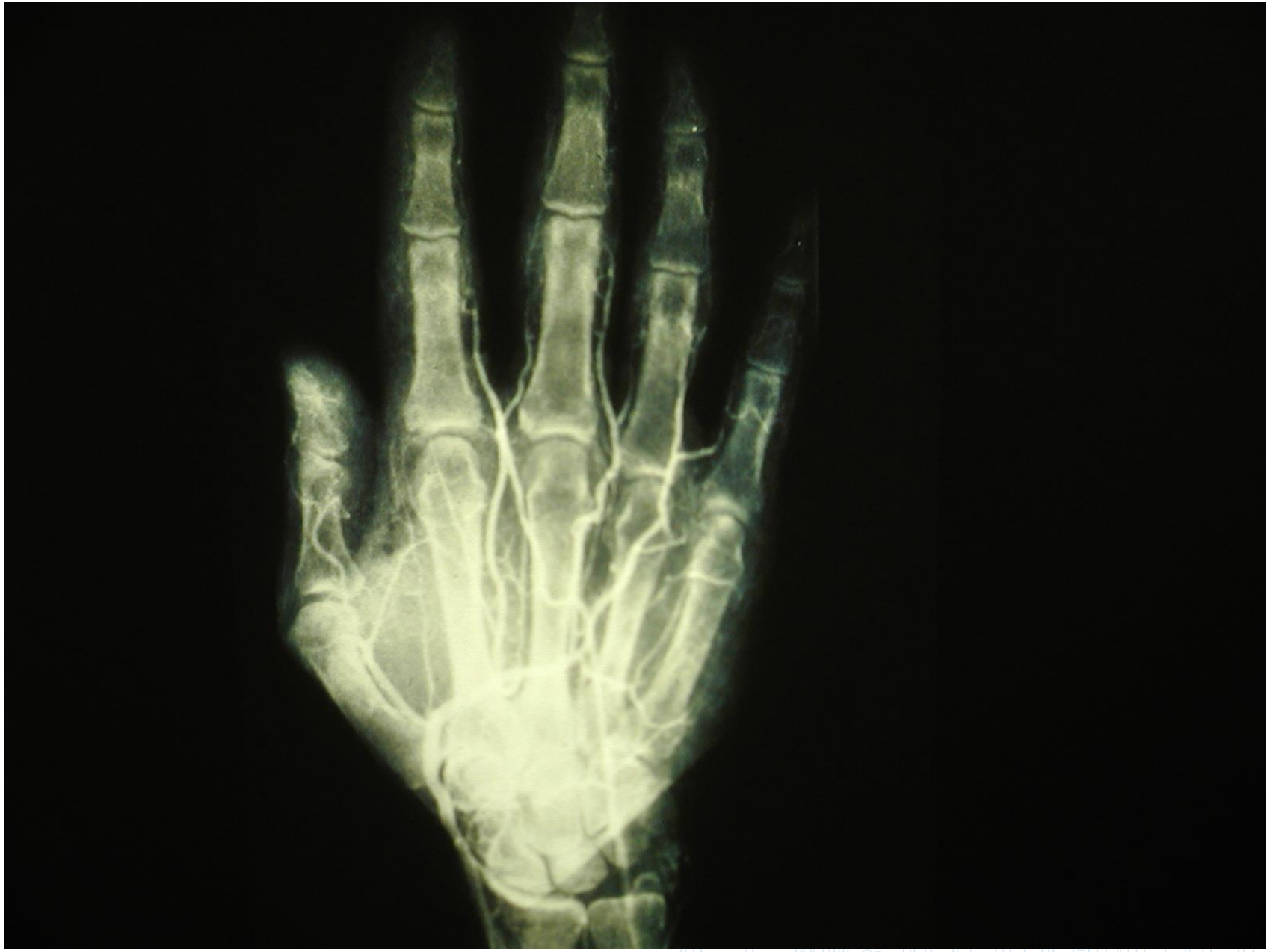

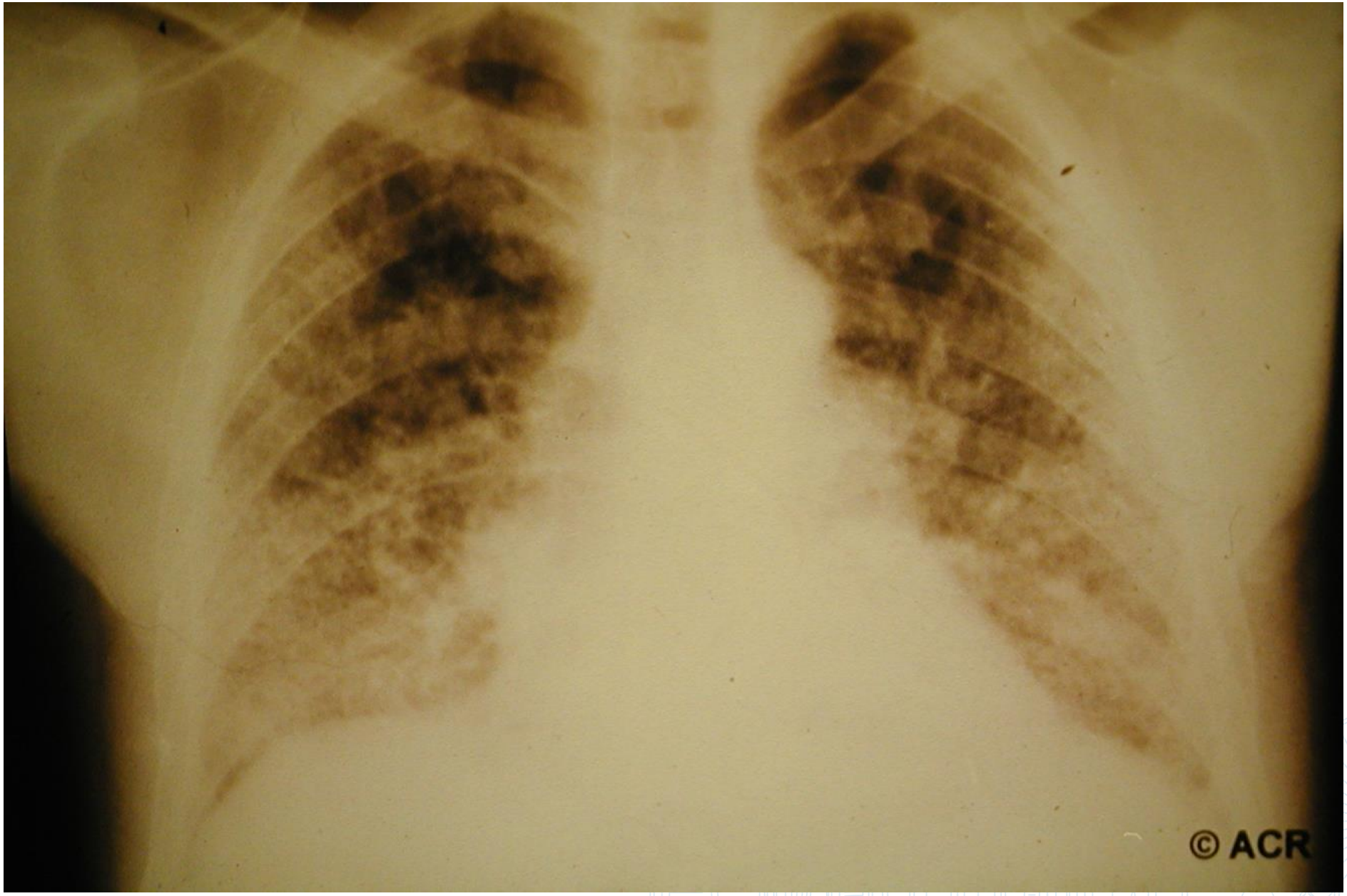
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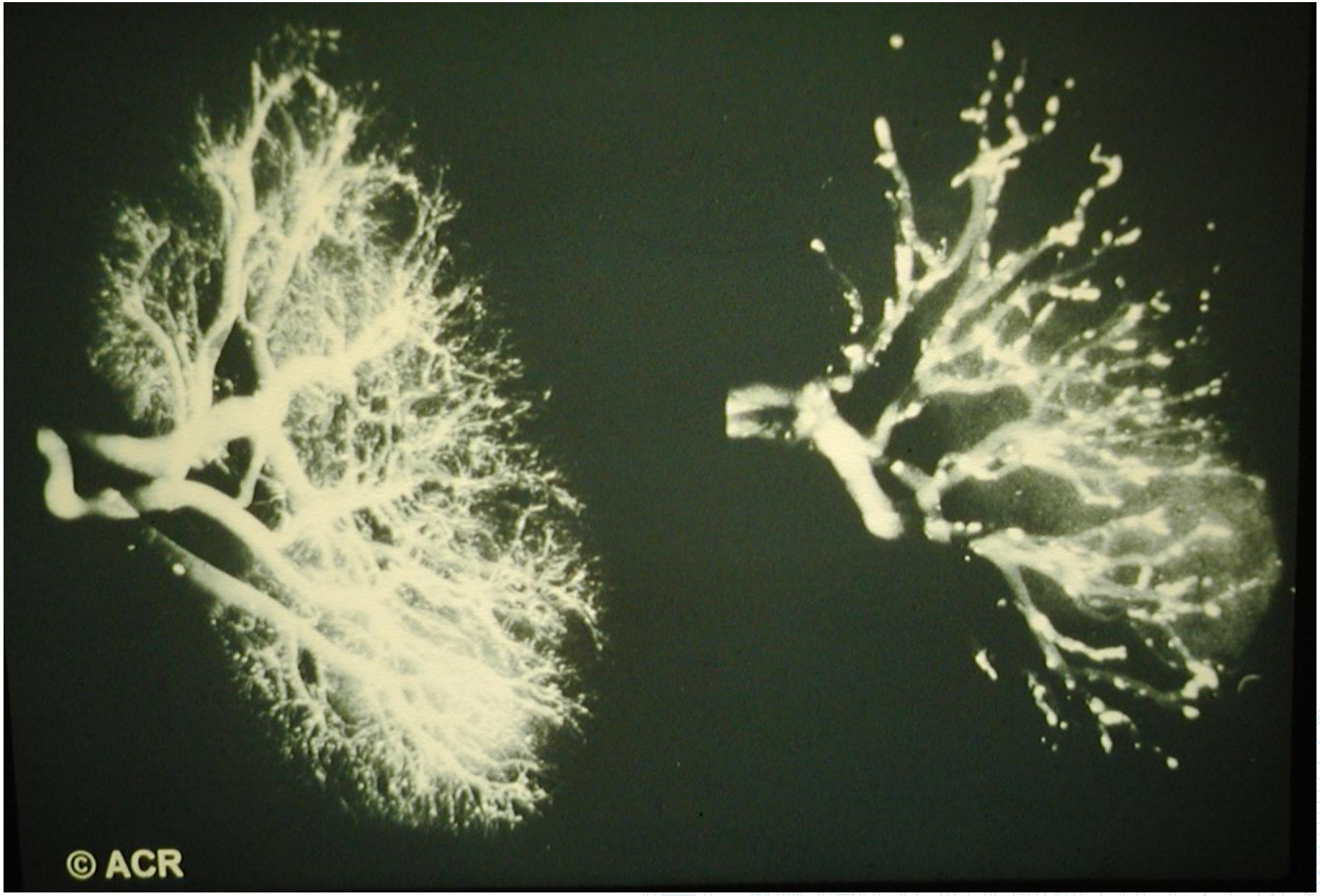
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Scleroderma Treatment

- 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)

Polymyositis

Patient Case

- 47 yo male with three year history of progressive weakness. Normal BP.
- 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
- CPK mildly elevated at 350

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

Dermatomyositis

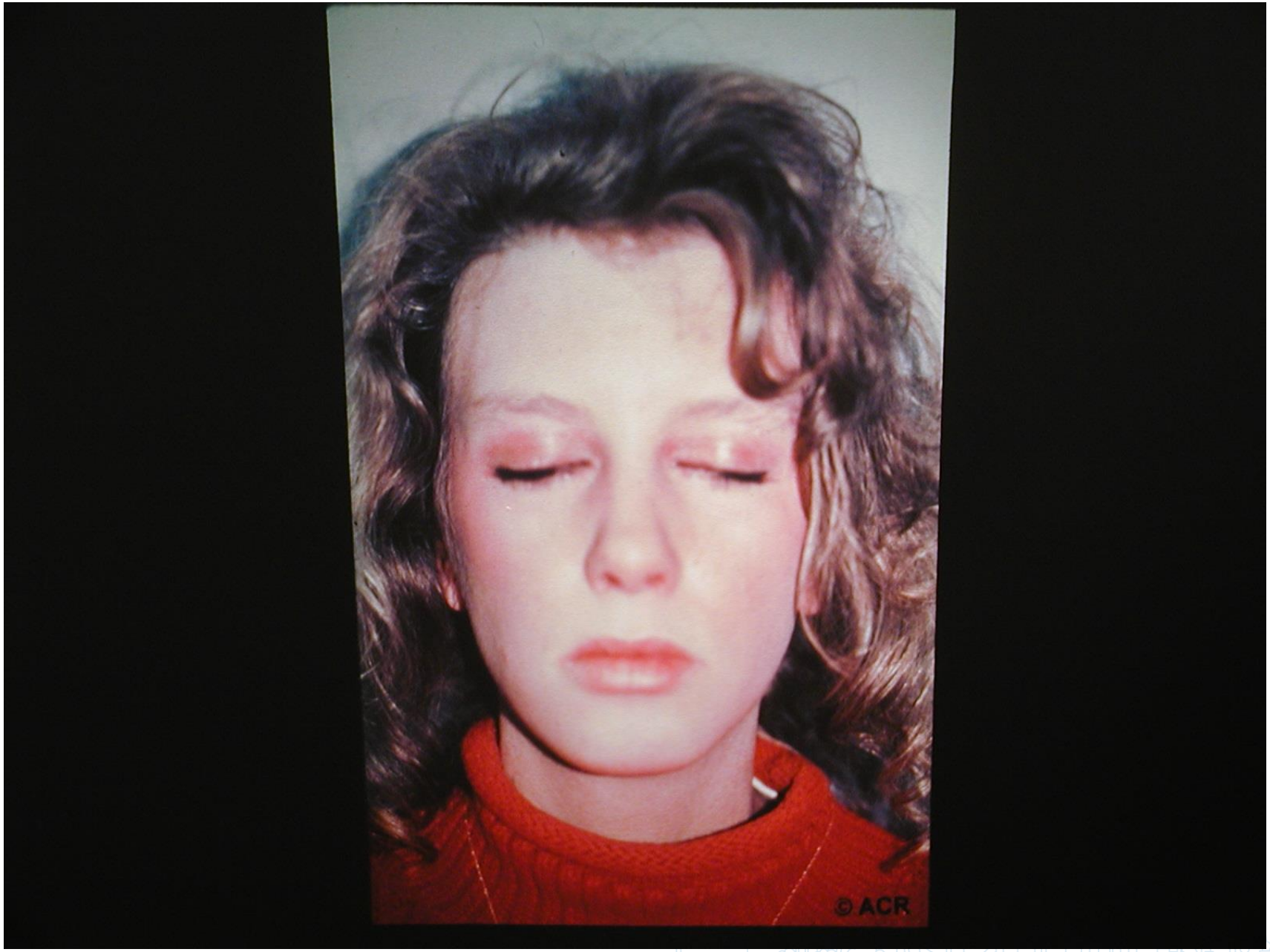
Gottron’s papules

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
- Both rare ~ 1:100,000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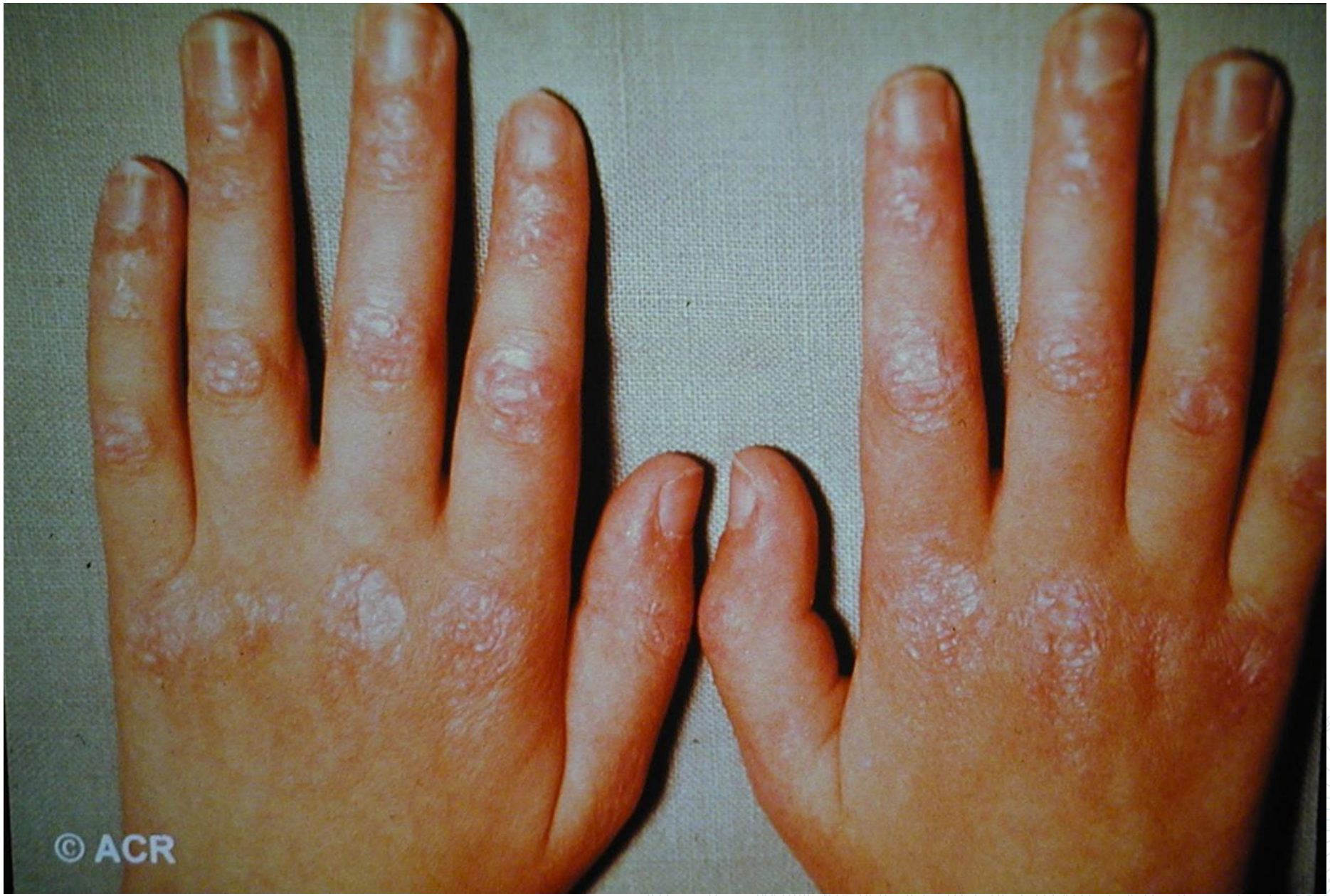
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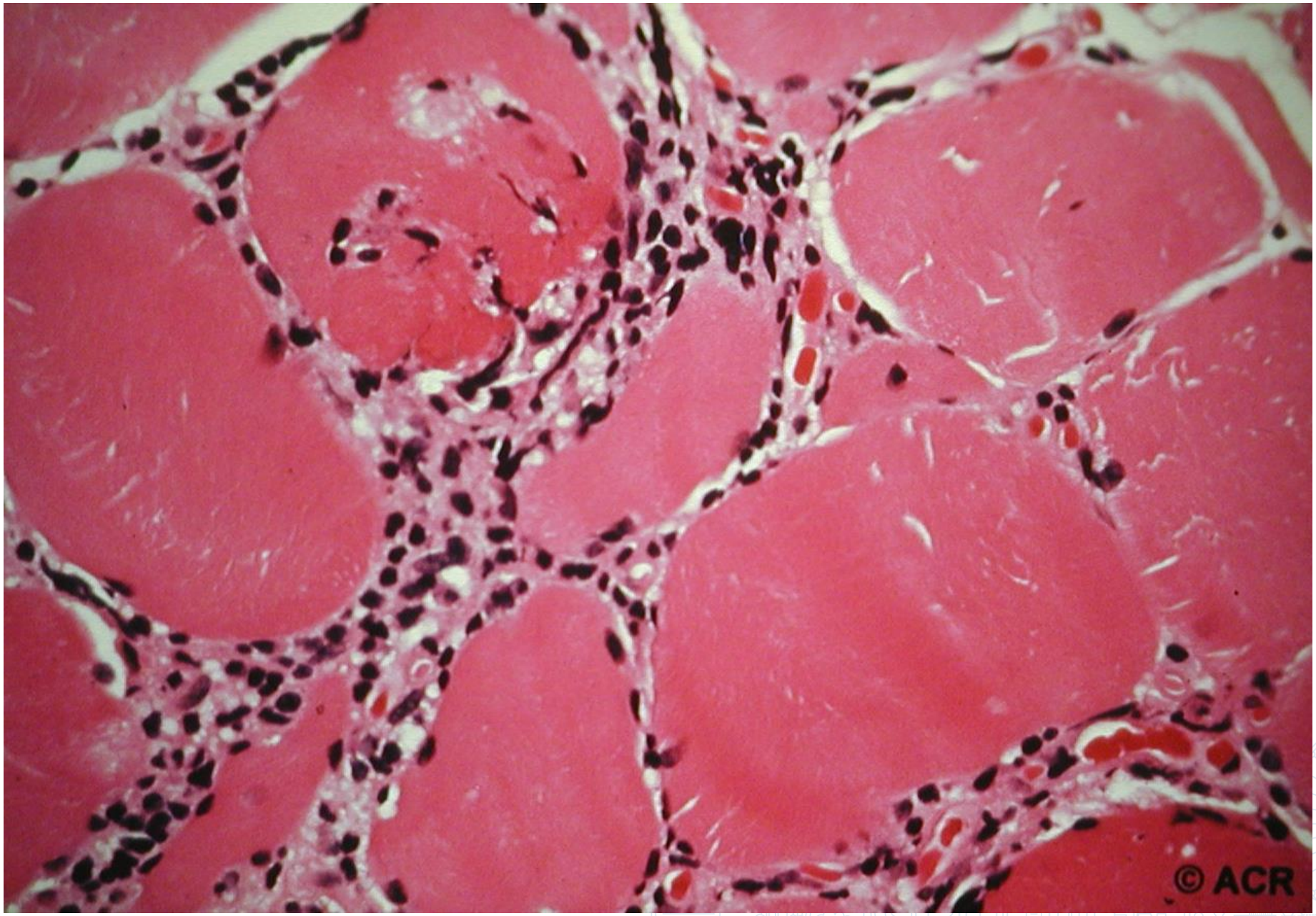
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Key Practice Points

1. The astute history and physical exam is vital when a CTD is suspected. (Pattern recognition). (SORT C)
2. Order studies only as indicated, avoiding the “immune panels” that can give false (+) tests. (SORT C)
3. Use the 2019 EULAR/ACR SLE criteria
4. Consider hydroxychloroquine in all patients with SLE. (SORT A)
5. Select methotrexate (MTX) as the first line DMARD for most RA patients.
6. Work closely with a rheumatologist, when available, in diagnosing and treating our patients. (SORT C)

Thank you for your attention

Contact information:

Eneedham7@gmail.com

Supplemental material

- Using methotrexate to treat rheumatoid arthritis
- Rheumatoid arthritis follow up and remission assessment
- Proximal muscle weakness – DDx
- Treatment comments for PM/DM

How to prescribe methotrexate

- 2.5 mg tablet
- 7.5 – 15 mg once a week (3-6 pills on Monday morning)
- Reassess response every month
- May increase by 2.5 – 5.0 mg per week
- Do not increase more frequently than monthly
- May increase up to 30 mg per week
- Always give with folate
- Mechanism of action
  - Immunosuppressant
  - Inhibits dihydrofolate reductase
  - Inhibits lymphocyte proliferation

MTX toxicities/contraindications to monitor

- Check baseline labs then every 1-2 months.
- hCG – absolute contraindication
- CBC – bone marrow suppression
- Liver enzymes – AST/ALT
  - Rule out Hep B/C
- BUN/Cr
- Remember MTX is a chemotherapeutic agent for many cancers to help decrease cancer cell replication.
- It also slows down the inflammatory process.

Therapy considerations when MTX ineffective

- If inadequate response over 3-6 months with MTX monotherapy:
- Add hydroxychloroquine or sulfasalazine or leflunomide

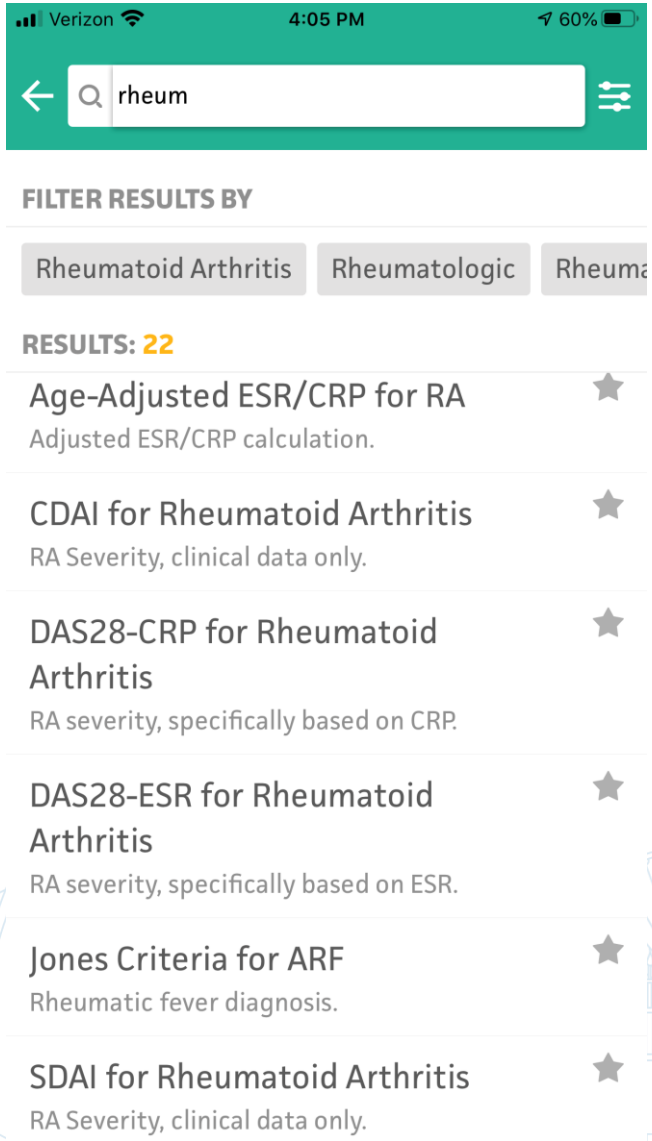
  Or

- Add biologic agent to MTX

- Refer the patient to rheumatology as soon possible with initial Dx
- With a patient on Medicaid, it may take several months for a Rheumatology appt.
- As family physicians, we could start MTX.

Follow up

- Return to clinic q3-6 months in stable patient
- Use a validated severity index
  - MDCalc has several to choose
- Follow exam and laboratory markers
- Consider xrays of hands/feet every 2 years
- If on chronic steroids, check DEXA
- Most common cause of death is CVD.
- Actively assess and treat for prevention of CVD



Remission

- Chronic RA patients are followed q3-6 months when stable
- Labs may include:
  - ESR, CRP, RA, ACPA
  - CBC, CMP
- If inflammatory markers are negative or low, may consider gentle down titration of DMARD and/or steroids.

DDx for proximal muscle weakness

- 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 (C-scope 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