HAWAII ACADEMY OF FAMILY PHYSICIANS

FEBRUARY 16–18 GRAND NANILOA HOTEL

SKIN SIGNS OF SYSTEMIC DISEASE

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No Disclosures

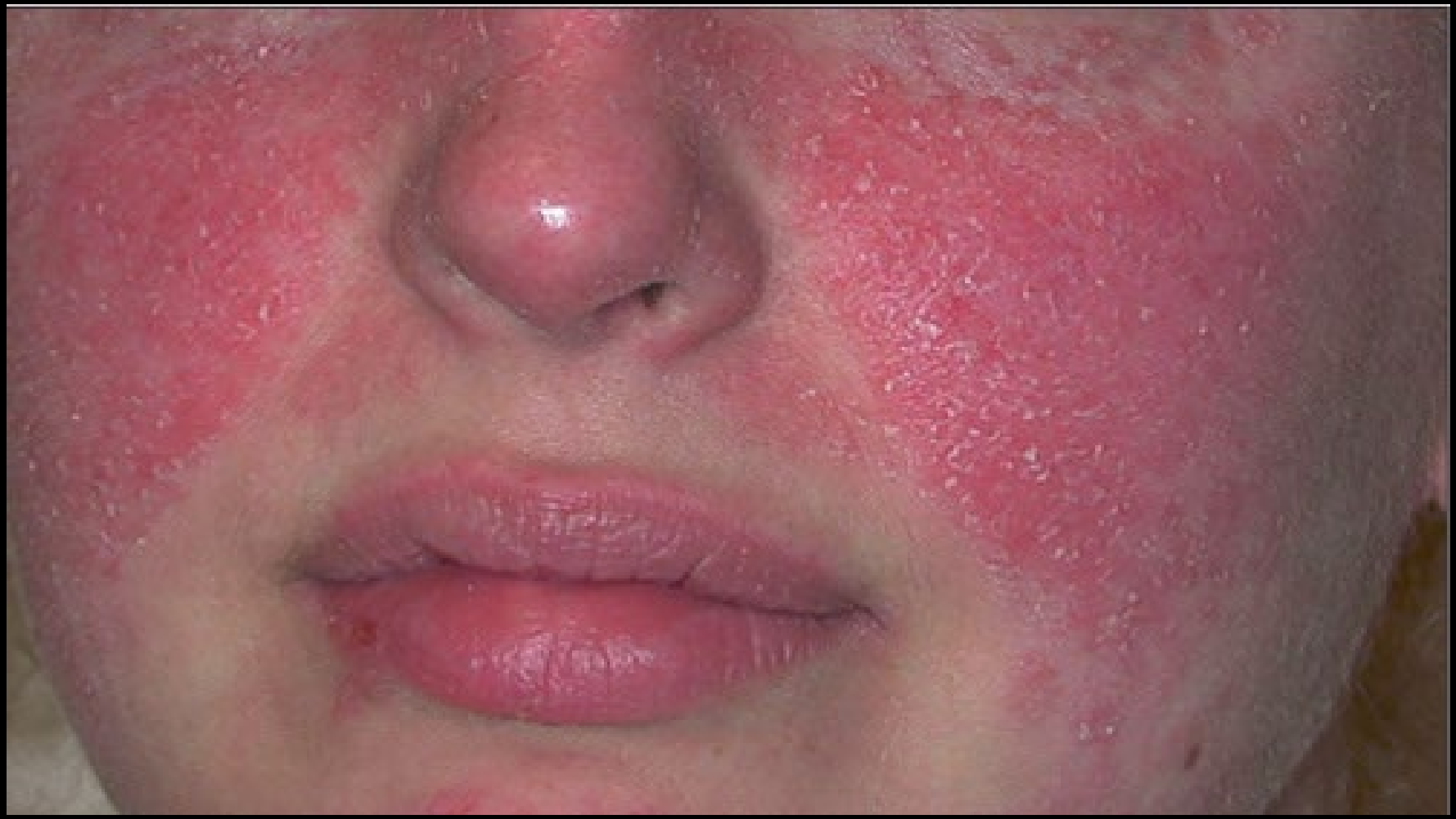
Objectives

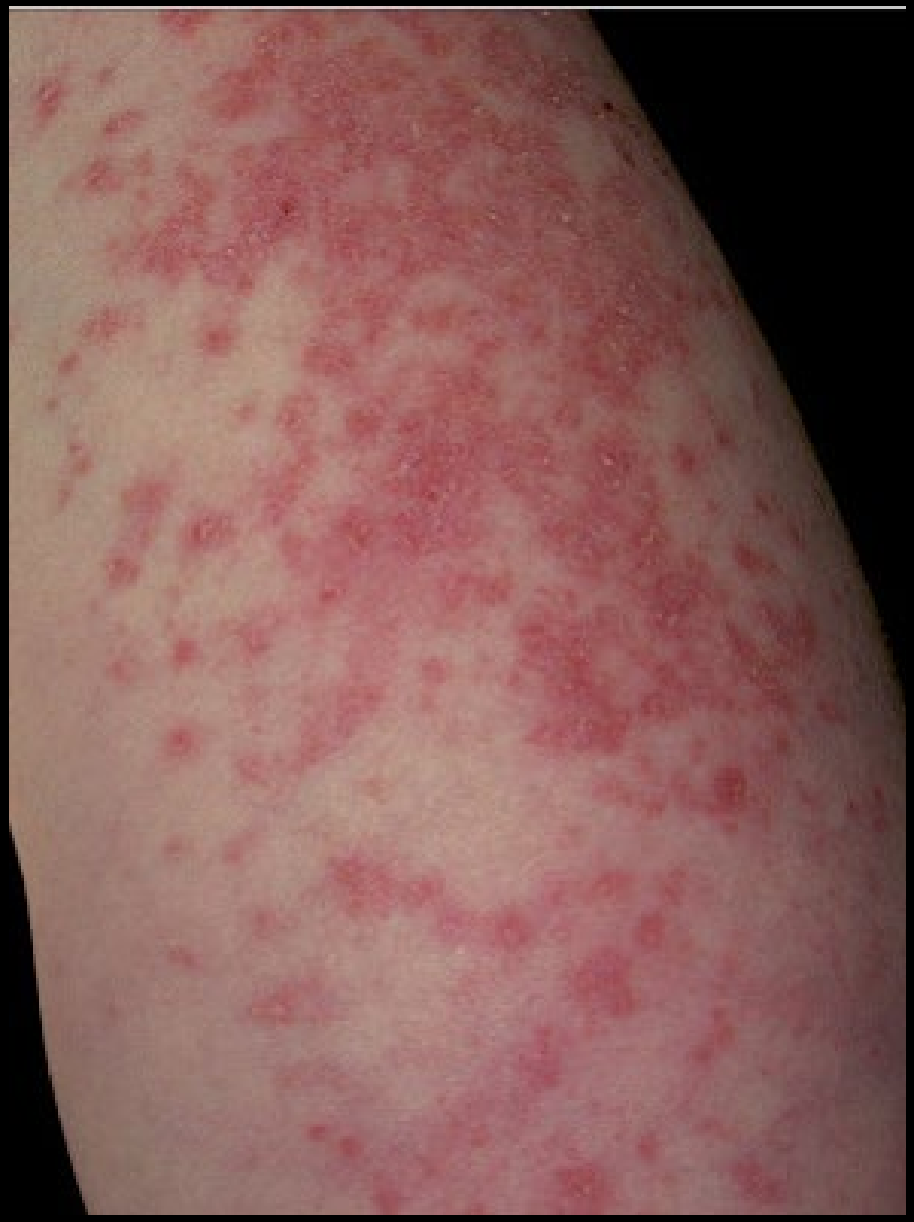
- Recognize major skin manifestations that can indicate underlying systemic diseases
- Develop appropriate differential diagnoses based on skin examination findings
- Discuss skin findings you may see in your clinic that may indicate inflammatory and autoimmune conditions, internal malignancy, and endocrine disorders

SIGNS OF AUTOIMMUNE DISEASE…

CASE 1:

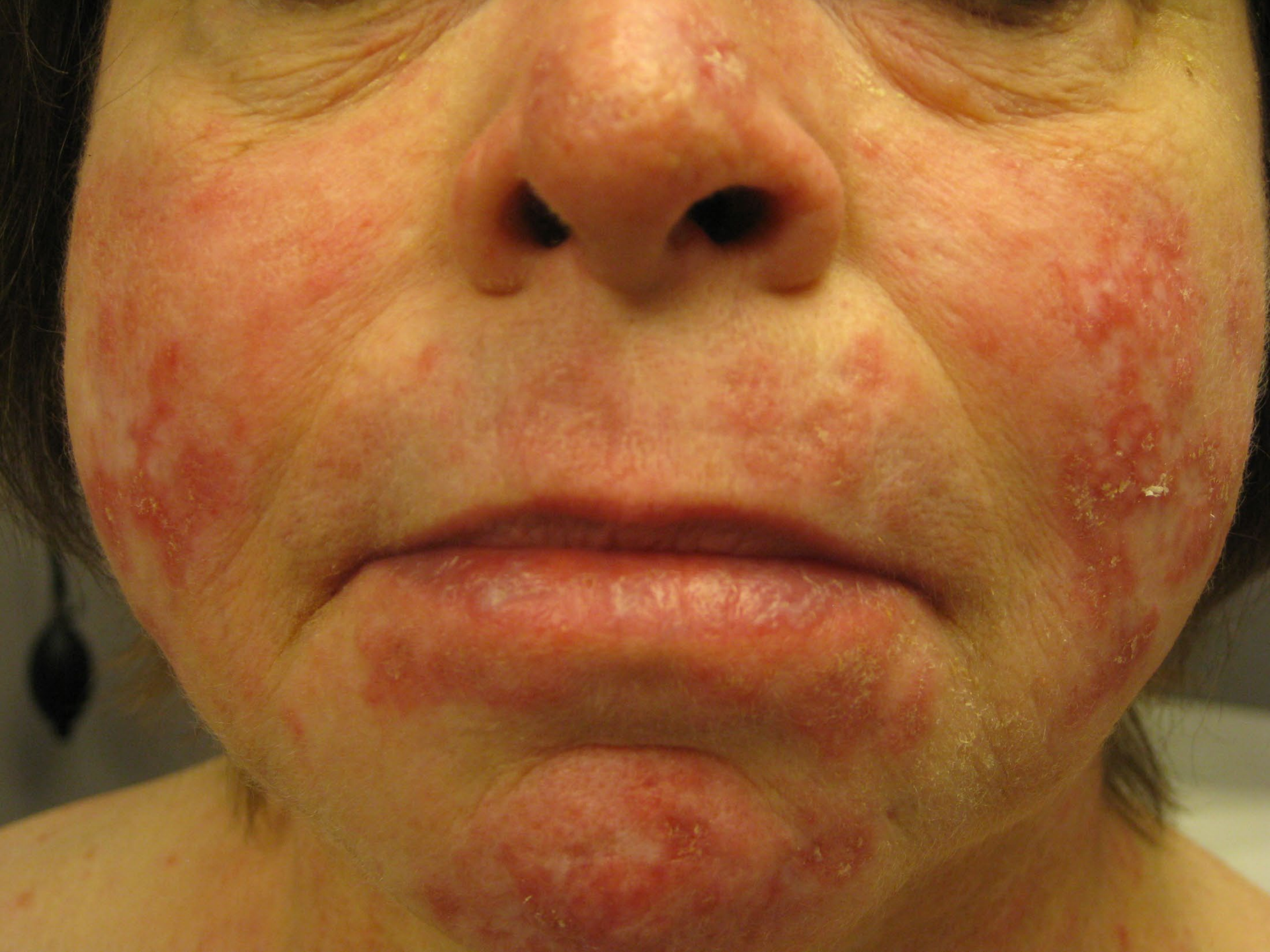
- 32 F presents with several week history of worsening:
  - Malaise, arthralgias, intermittent fevers, oral ulcers and pleuritic chest pain *… AND FACE FEELS RED AND SWOLLEN …*

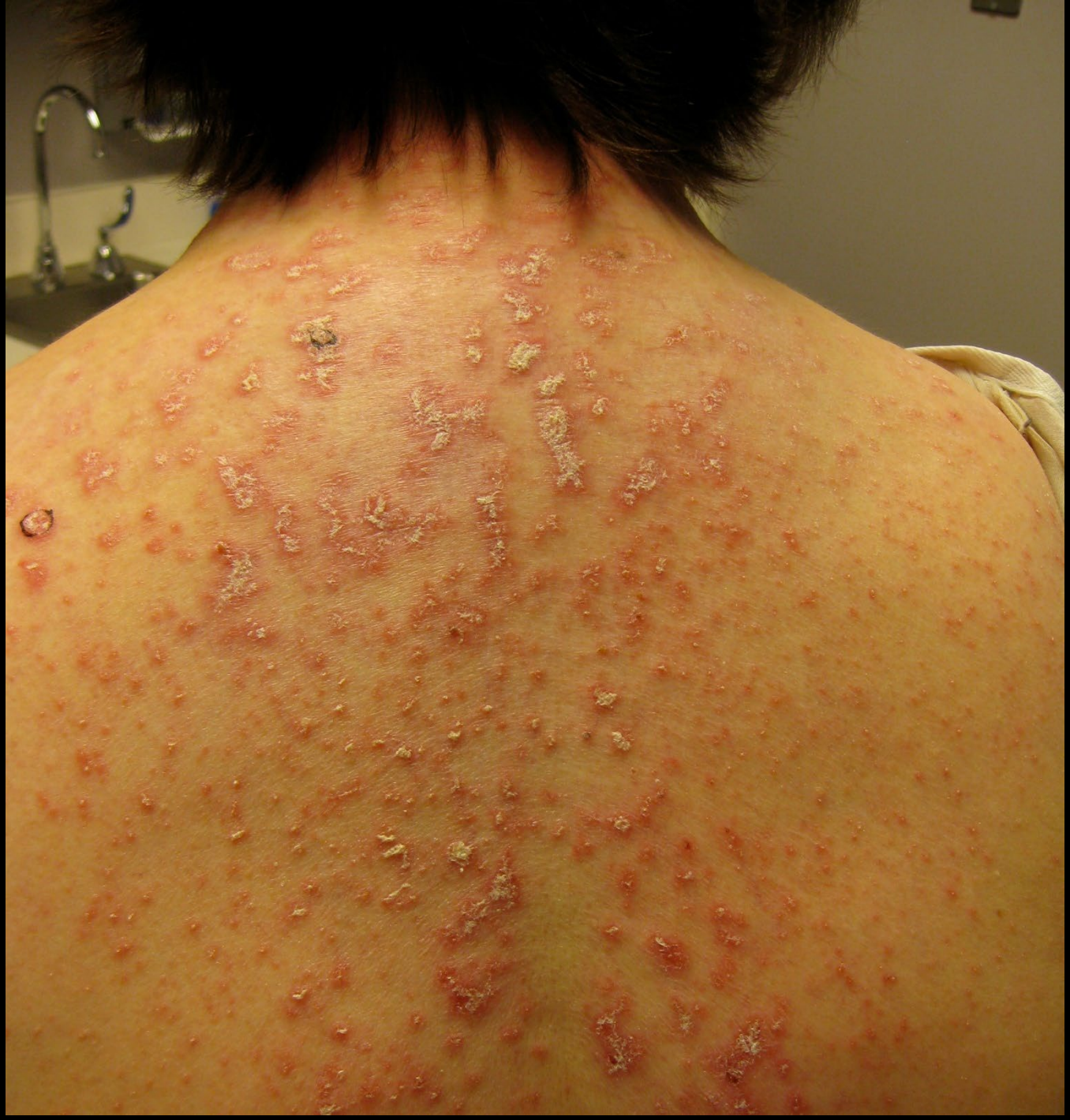




CASE 2:

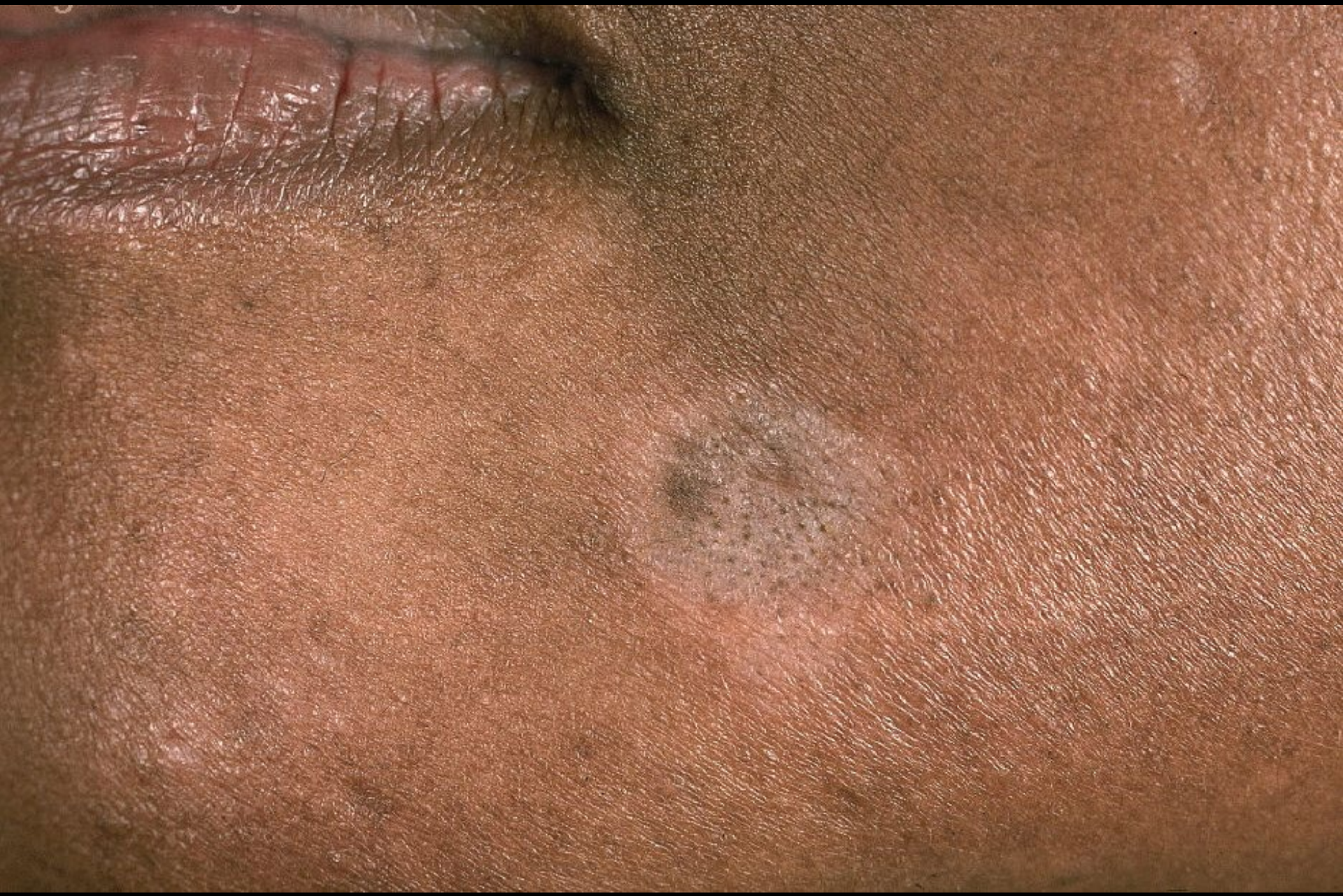
- **55 F presents with several year history of:**
  - ***– SCALY RASH, WORSENS WITH SUN***

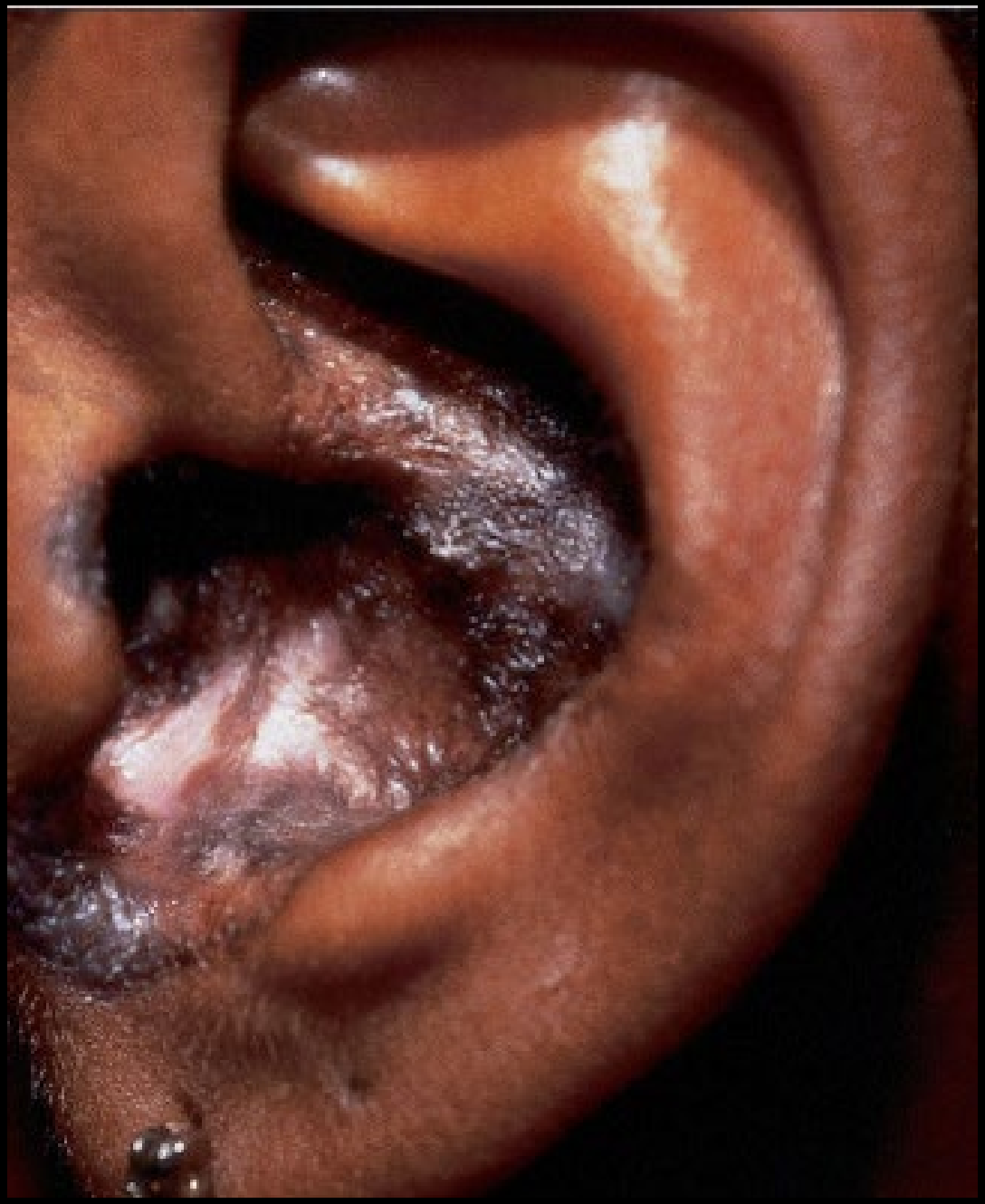




CASE 3:

- **48 F presents with several month history of hair loss and:**
  - ***SCALY SKIN LESIONS ON FACE AND EARS THAT ARE SCARRING …***



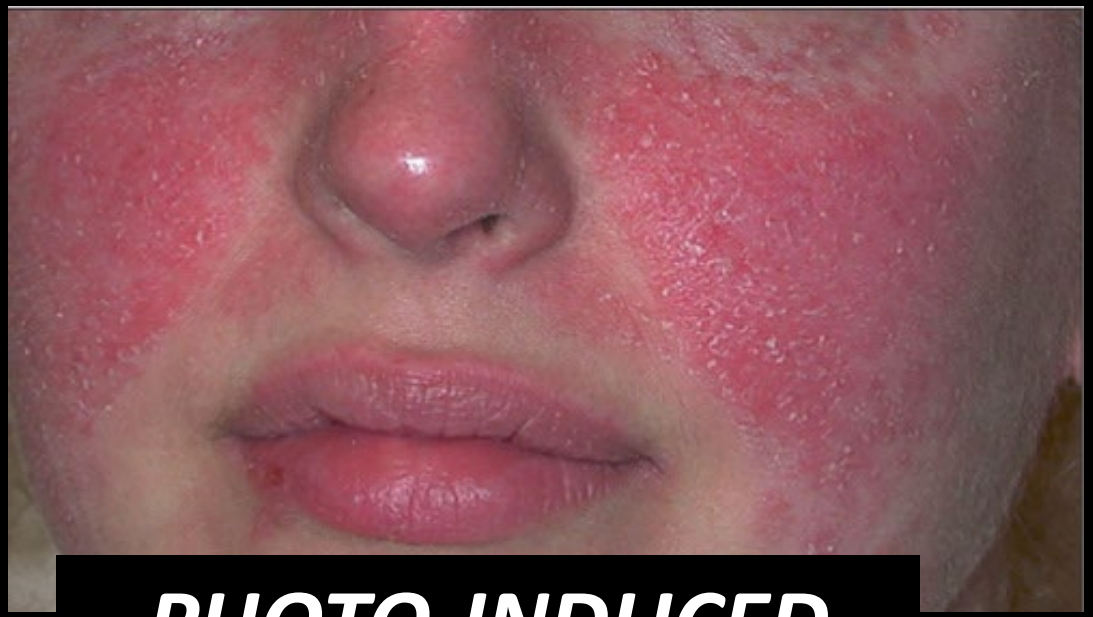


LUPUS ERYTHEMATOSUS

- ***Cutaneous findings specific to lupus may be divided into <u>3 distinct categories</u>:***

1. **ACUTE CUTANEOUS LUPUS ERYTHEMATOSUS**
2. **SUBACUTE CUTANEOUS LUPUS ERYTHEMATOSUS**
3. **CHRONIC CUTANEOUS LUPUS (DISCOID LUPUS ERYTHEMATOSUS)**

ACUTE CUTANEOUS LUPUS ERYTHEMATOSUS



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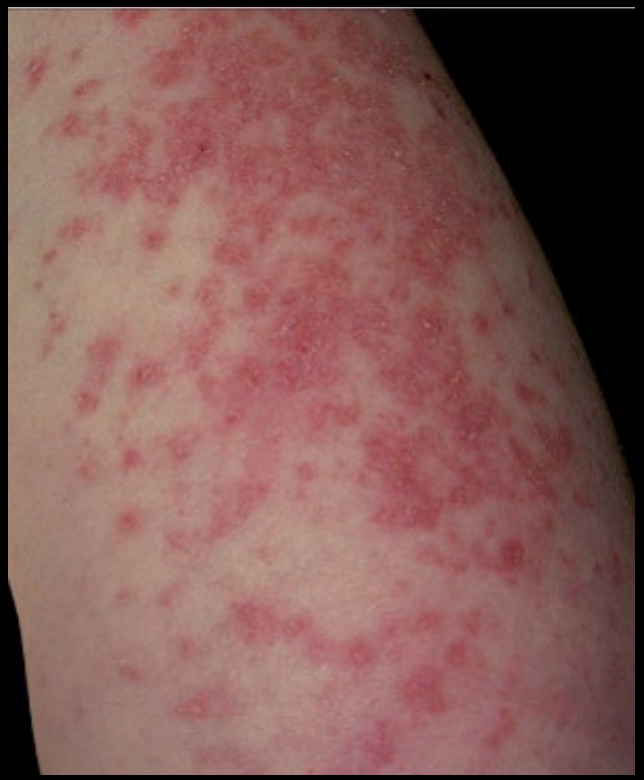
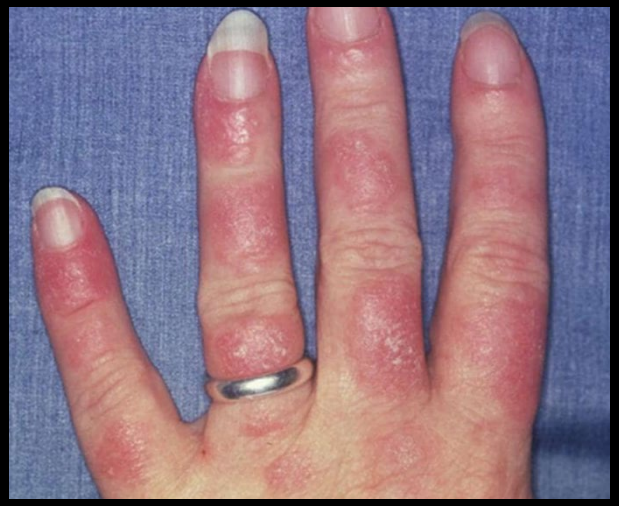


RED, SCALY, EDEMATOUS

ACUTE CUTANEOUS LUPUS ERYTHEMATOSUS

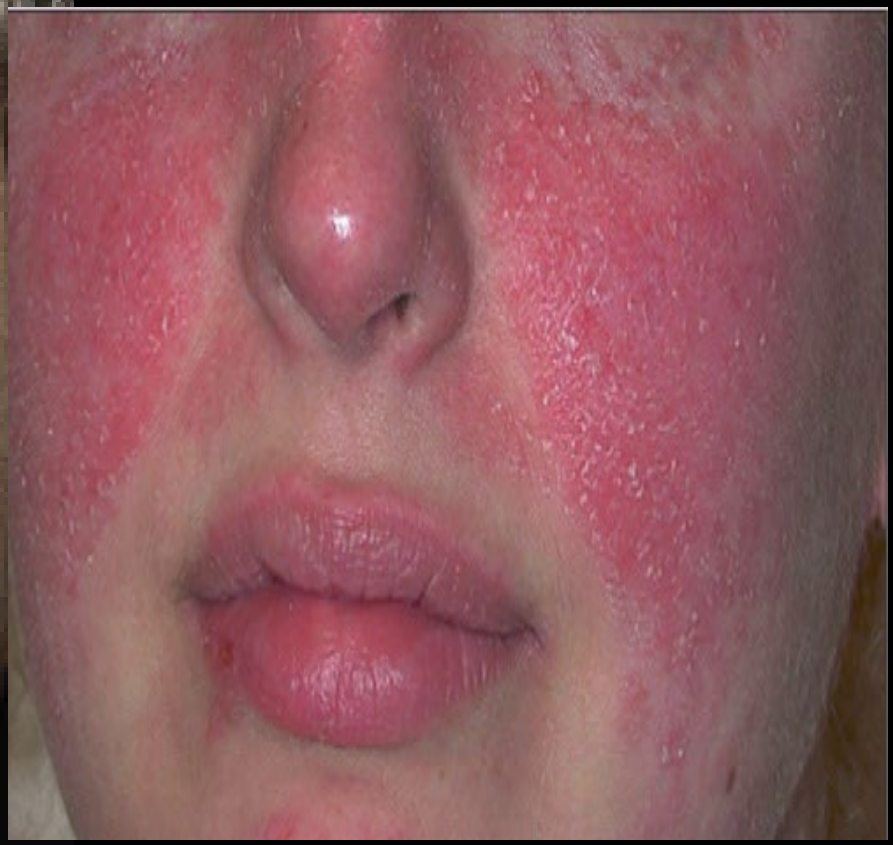
- **RASH OF ACUTE CUTANEOUS LUPUS MAY:**
  - ***Last hours to weeks***
  - ***Extend beyond the face and be "photo-distributed"***
  - ***Affect the hands, sparing the knuckles***



SYSTEMIC LUPUS: DIAGNOSTIC CRITERIA

- **4/11 CRITERIA MUST BE MET:**
  1. **Malar rash**
  2. **Discoid rash**
  3. **Photosensitivity**
  4. **Oral ulcers**
  5. **Arthritis (2+ joints)**
  6. **Serositis (pleuritis or pericarditis)**
  7. **Renal disorder (proteinuria)**
  8. **Neurologic disorder (seizures, psychosis, etc)**
  9. **Hematologic disorder (anemia, leukopenia, etc)**
  10. **Immunologic disorder (Anti-dsDNA, anti-Sm, etc)**
  11. **Antinuclear antibodies (ANA+)**

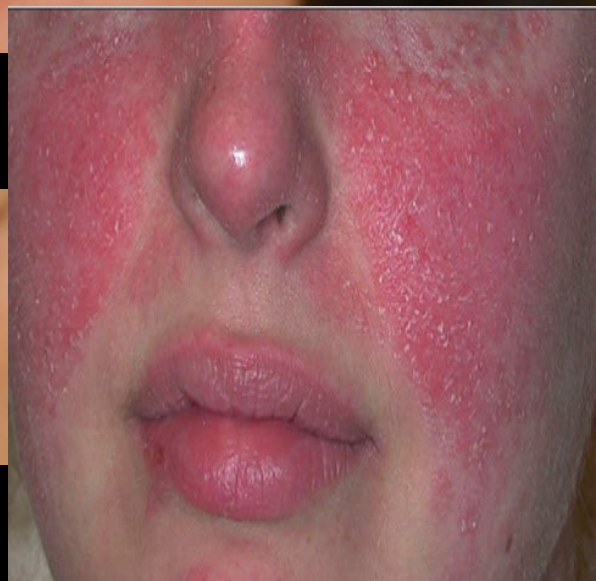
OTHER BUTTERFLIES…



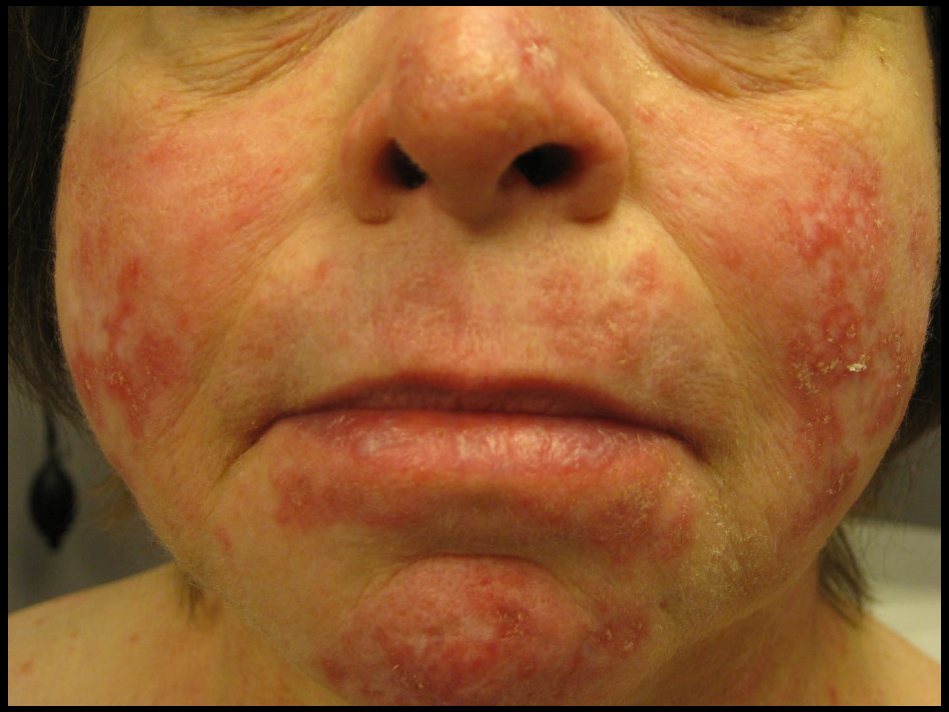
SEBORRHEIC DERMATITIS

OTHER BUTTERFLIES…

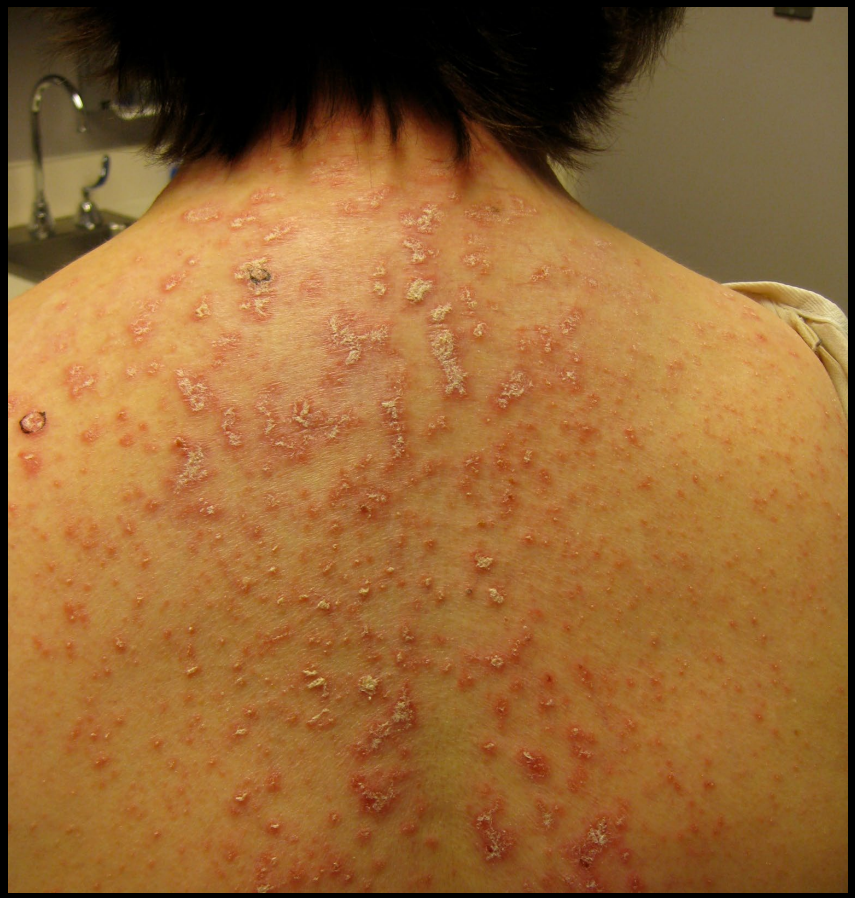
ROSACEA

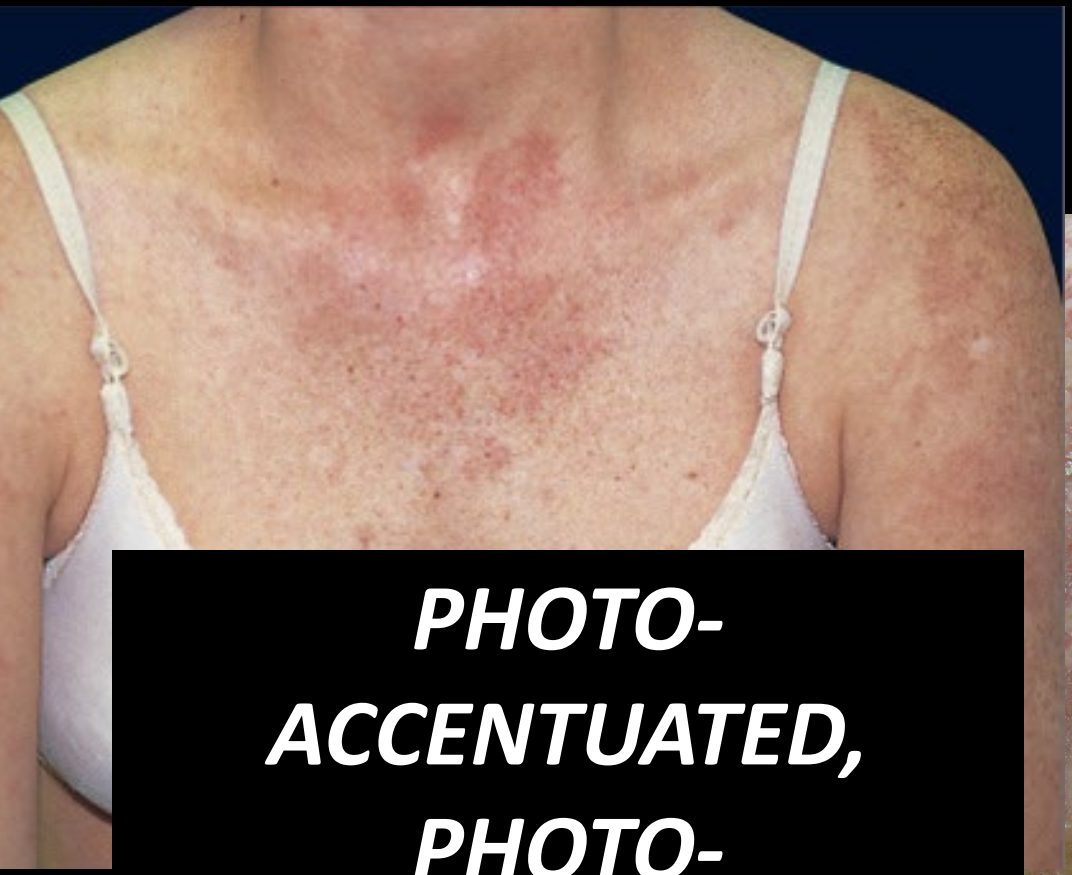


SUBACUTE CUTANEOUS LUPUS ERYTHEMATOSUS (SCLE)



'PSORIASIFORM'





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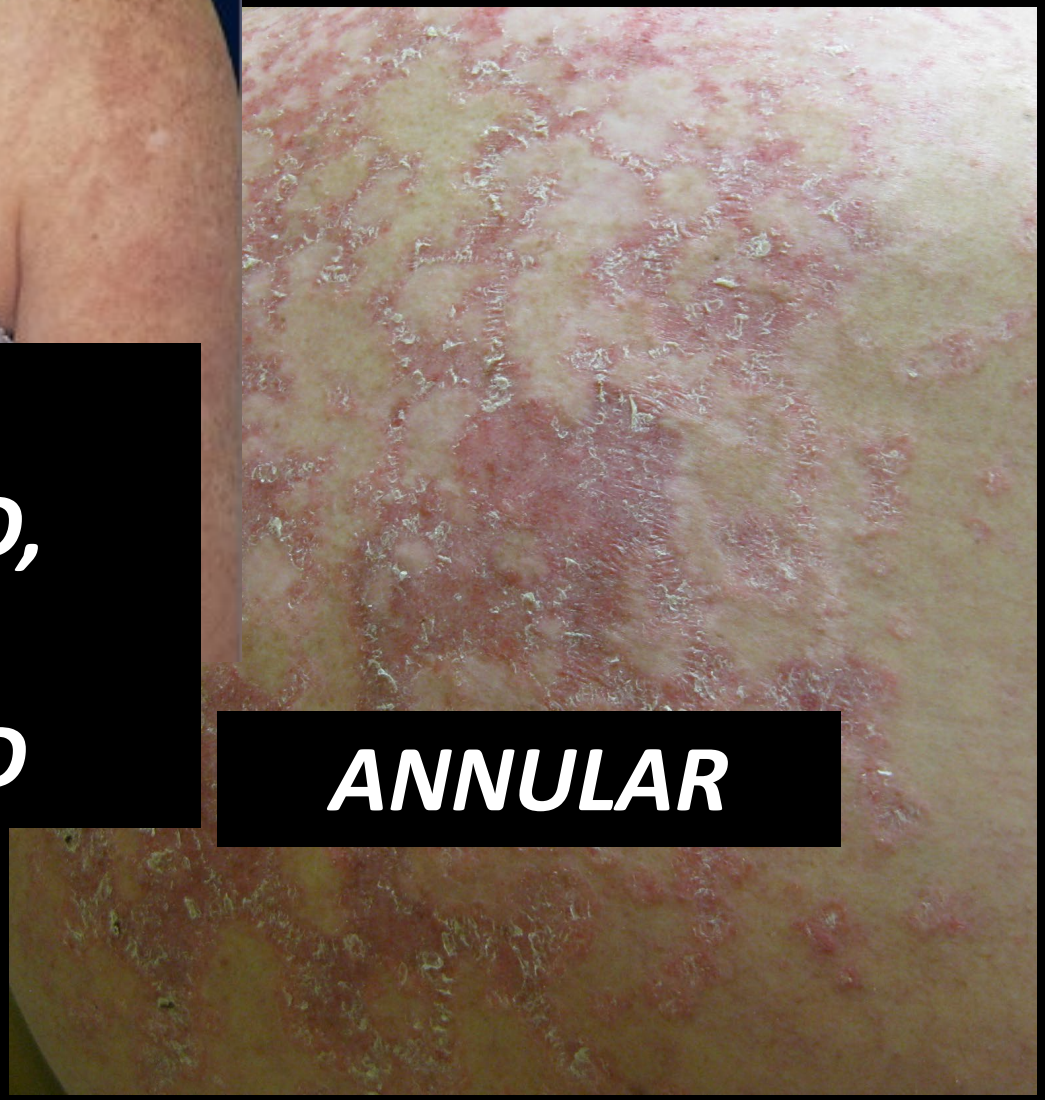
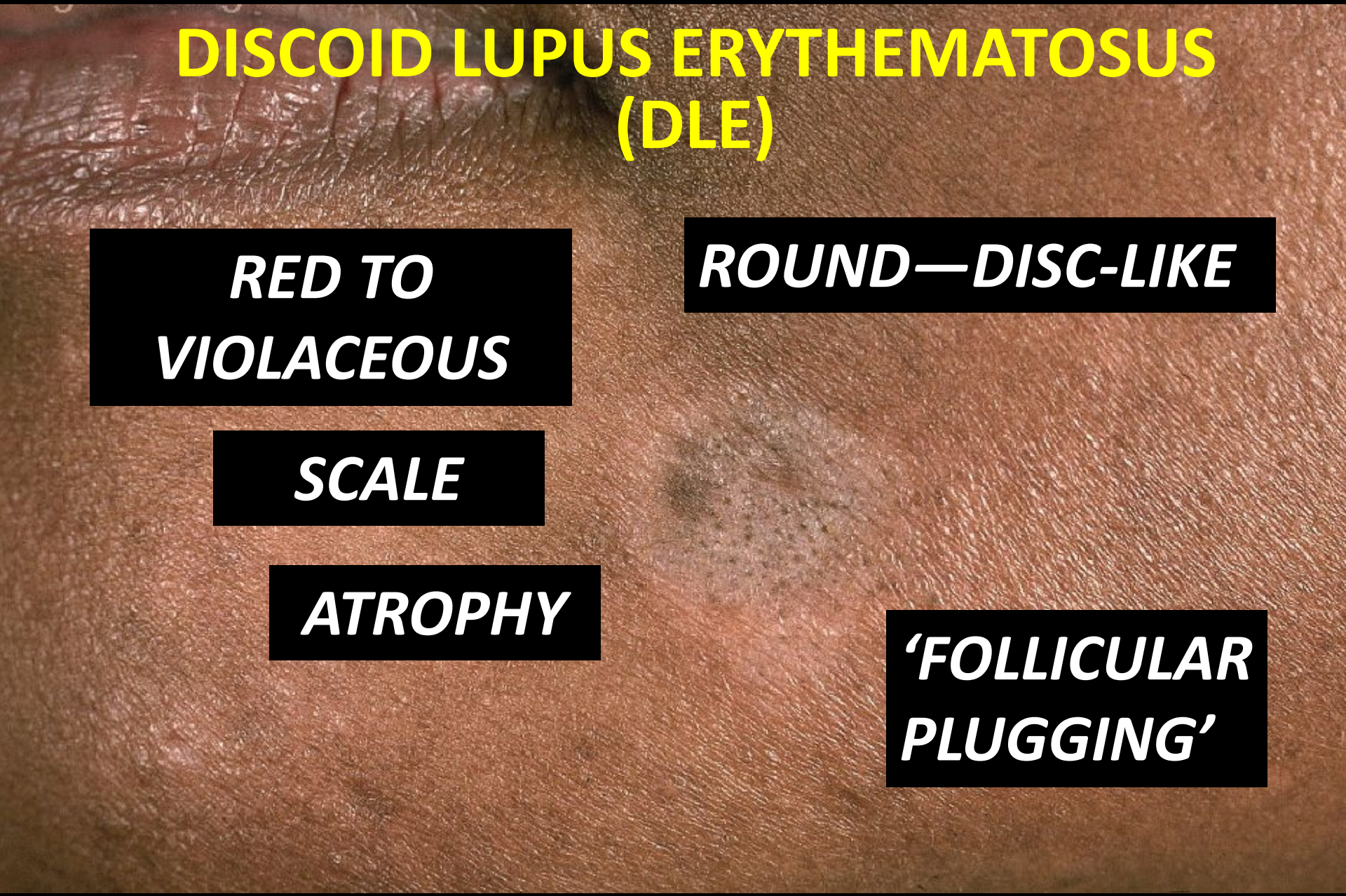


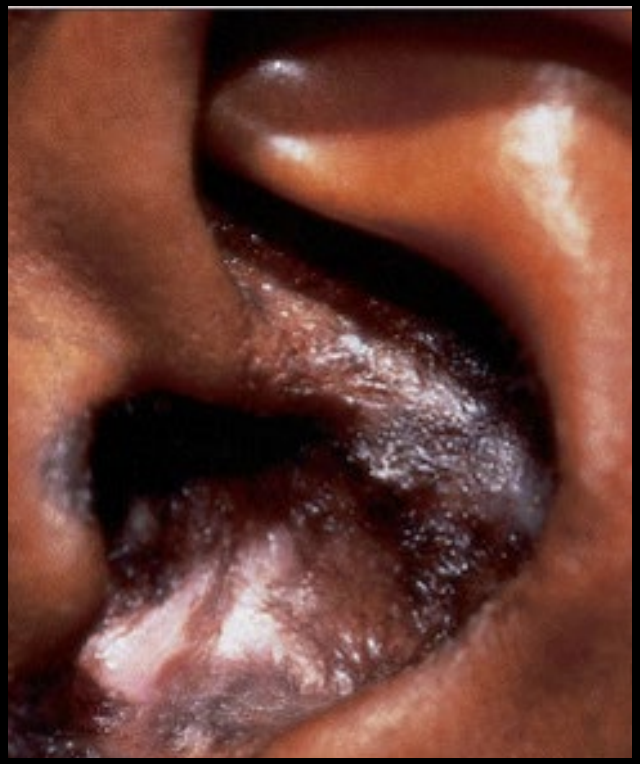
ANNULAR

SUBACUTE CUTANEOUS LUPUS ERYTHEMATOSUS (SCLE)

- ***~50% of patients meet criteria for SLE***
  - ***severe manifestations (arthritis, CNS, renal) uncommon***
- ***Majority patients have anti-Ro/SSA antibodies***
- ***DRUG-INDUCED:***
  - ***HCTZ, Calcium Channel Blockers, TERBINAFINE, ACEi***

DISCOID LUPUS ERYTHEMATOSUS (DLE)





DYSPIGMENTATION

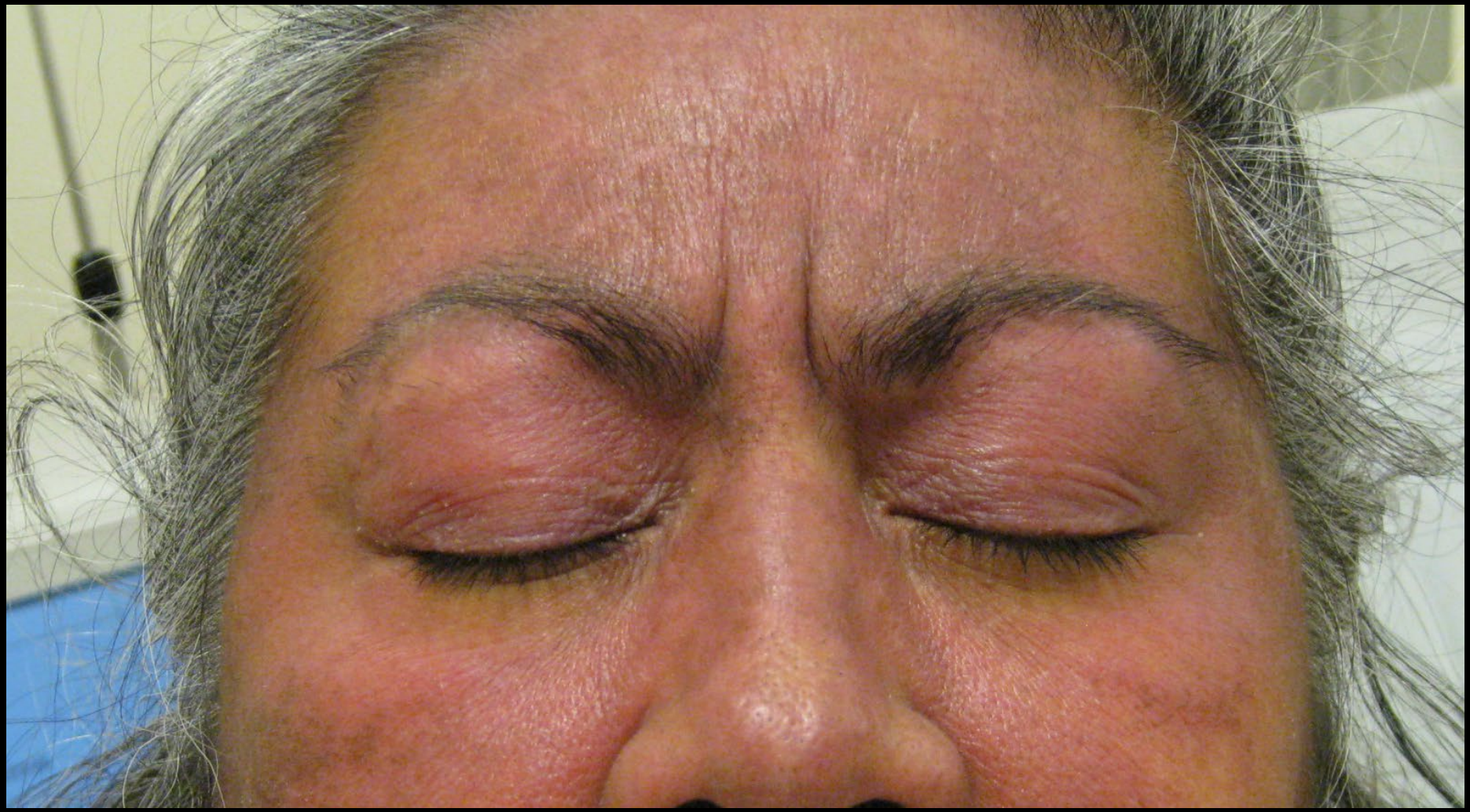
SCARRING

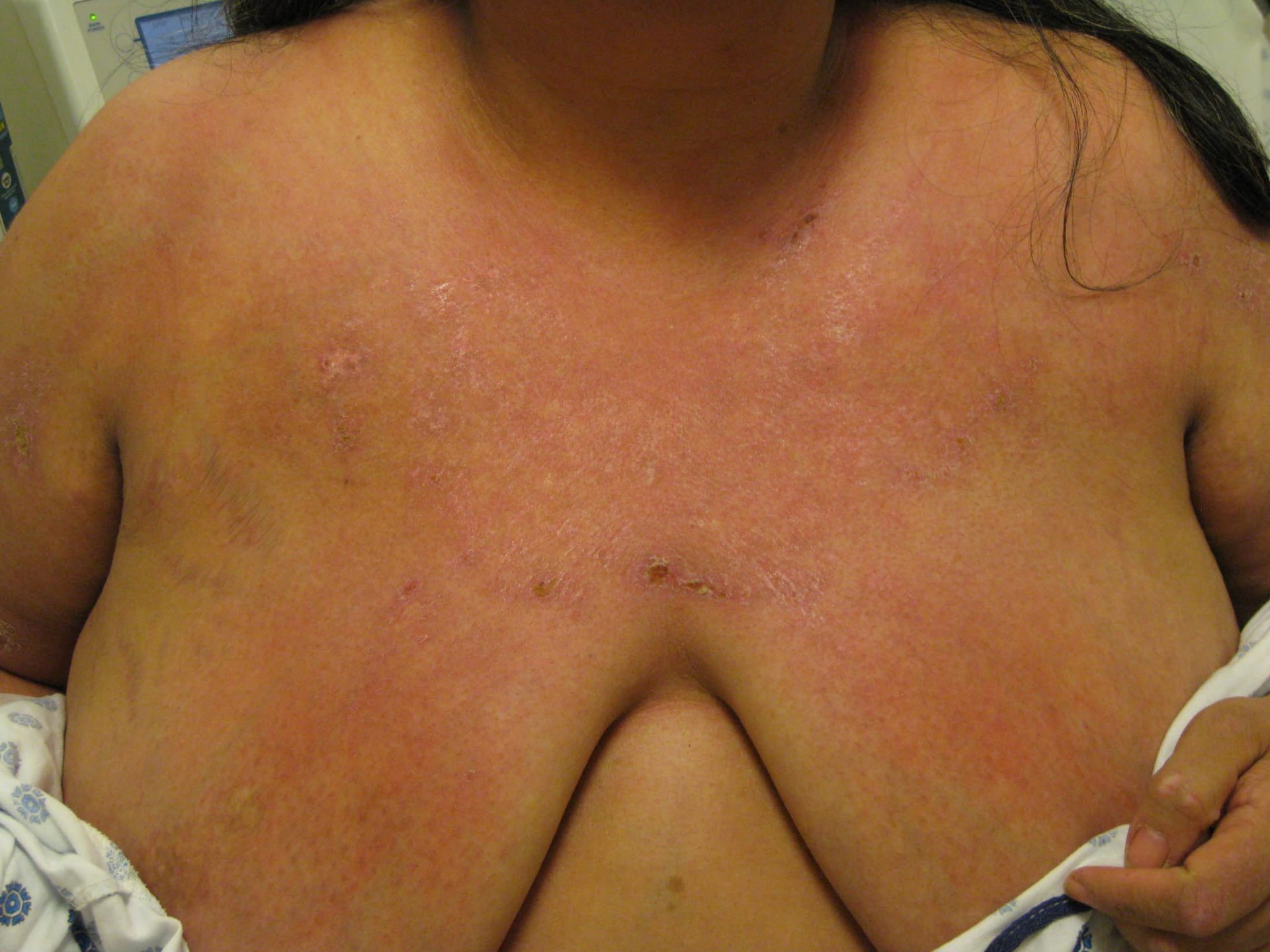
DISCOID LUPUS ERYTHEMATOSUS (DLE)

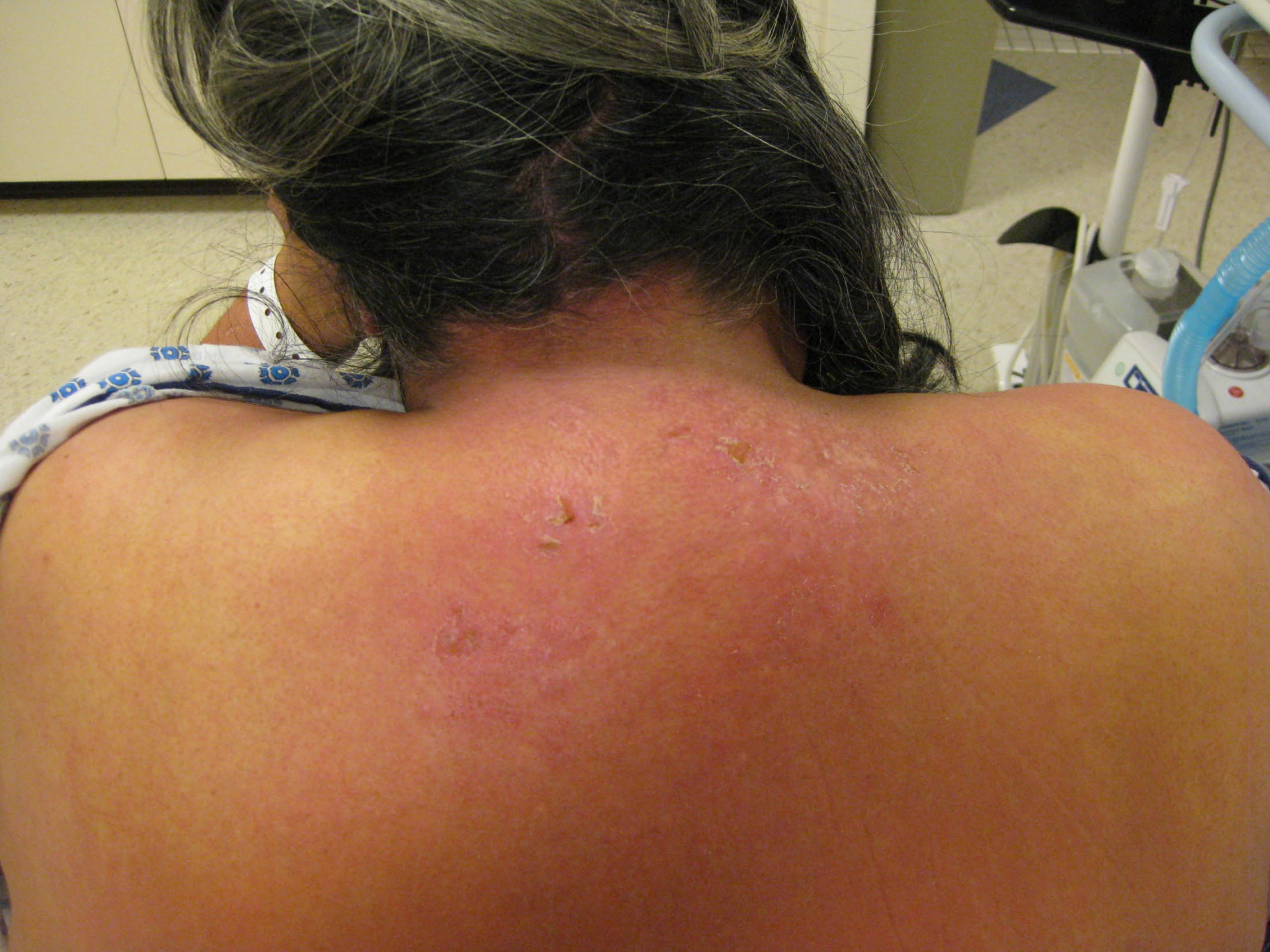
- **Only 5-10% of patients with DLE will develop SLE**

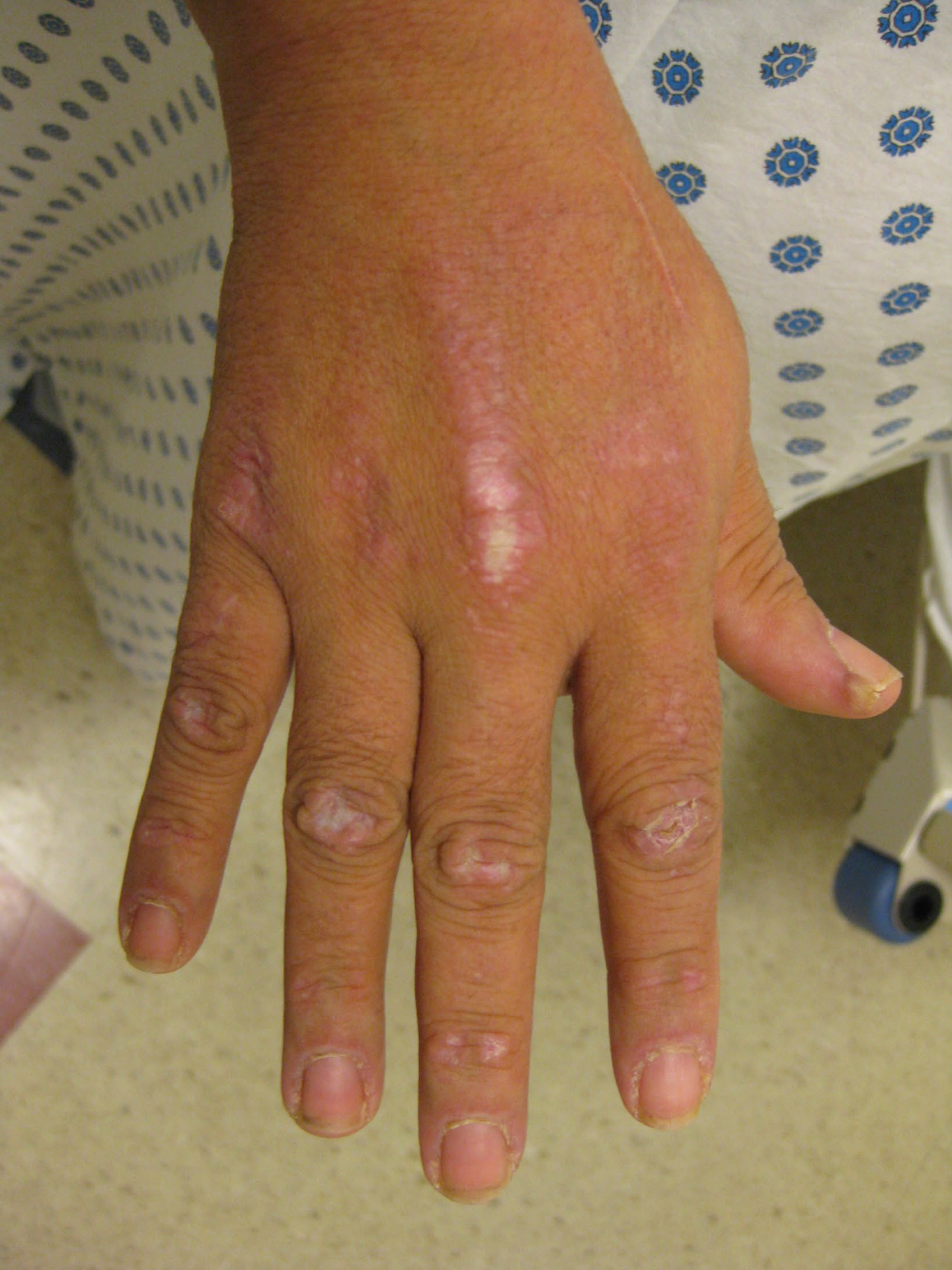
CASE 4:

- **47 y/o F presents with a 2-3 month history of:**
  - ***Proximal muscle weakness** (e.g. difficulty raising arms above head, difficulty standing from a chair)*
  - ***Fatigue***
  - ***New skin changes …***









DERMATOMYOSITIS

- ***Bimodal age of onset:***
  - ***10-15 y/o ("juvenile") and 45-57 y/o ("adult")***

- ***~60% present with muscle weakness and skin findings simultaneously***

DERMATOMYOSITIS: Systemic features

- ***MYOSITIS W/ PROXIMAL MUSCLE WEAKNESS***

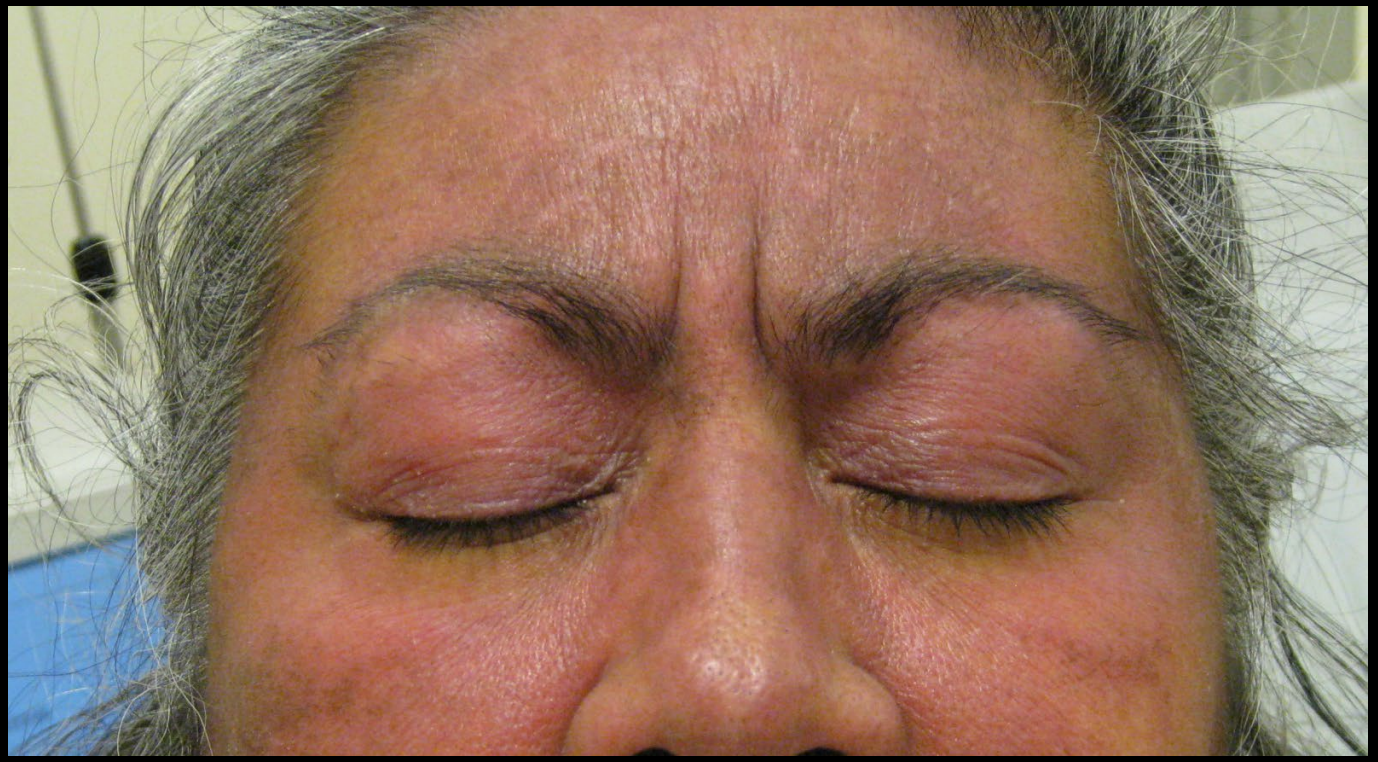
- ***Dysphagia -15-50%***
- ***Pulmonary Disease -15-30%***
  - **Respiratory failure due to muscle weakness, Diffuse interstitial lung disease, Dysphonia**
- ***Cardiac***
  - **Cardiomyopathy, Conduction defects**

DERMATOMYOSITIS: Systemic features

- ***Internal malignancy:***
  - ***~25% of adult patients have an associated occult malignancy***
    - ***Most common: OVARIAN and COLON***

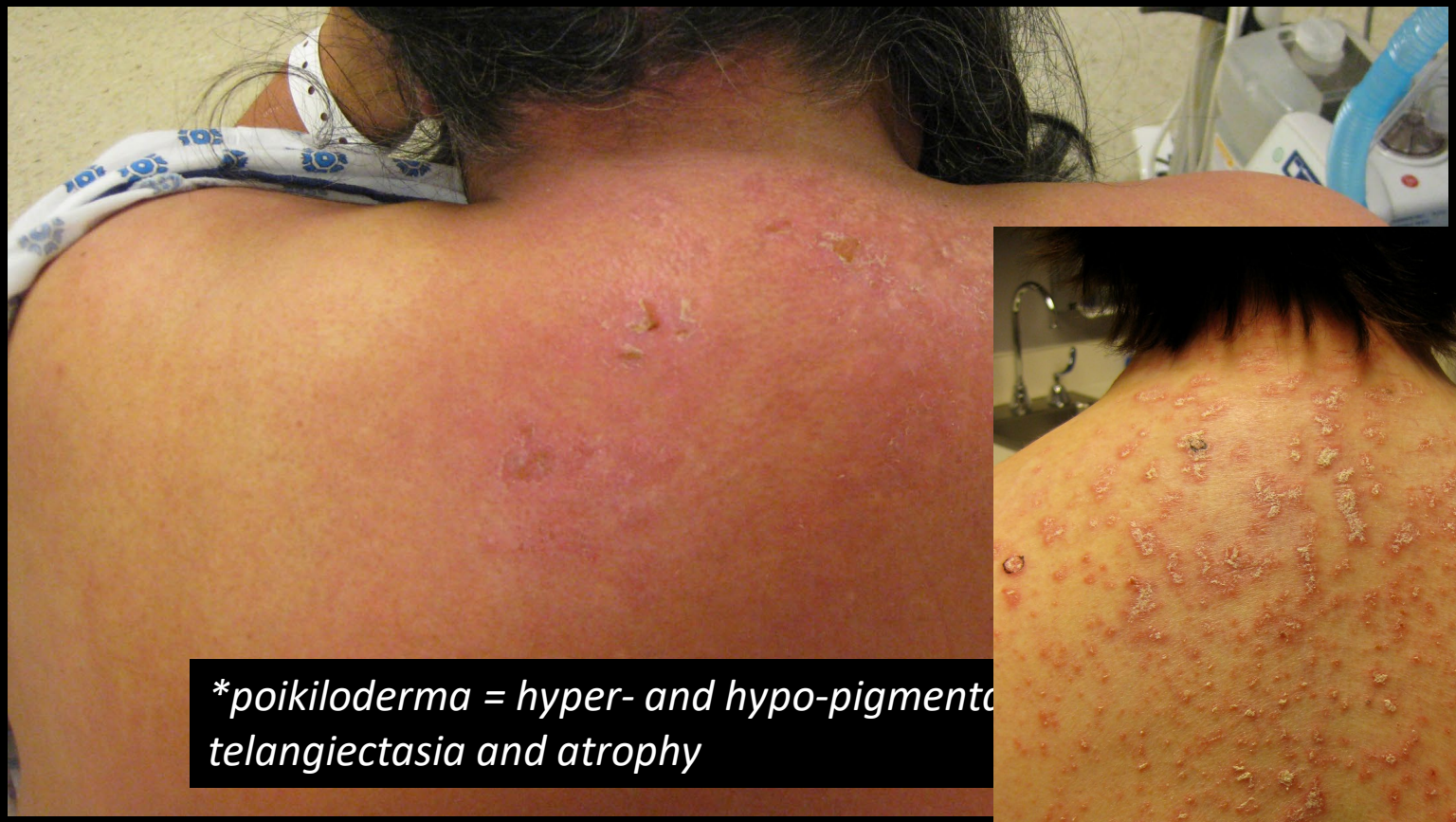
DERMATOMYOSITIS

- *Characteristic cutaneous findings:*
  - **Heliotrope sign:** *violaceous, edematous patches around the eyes*

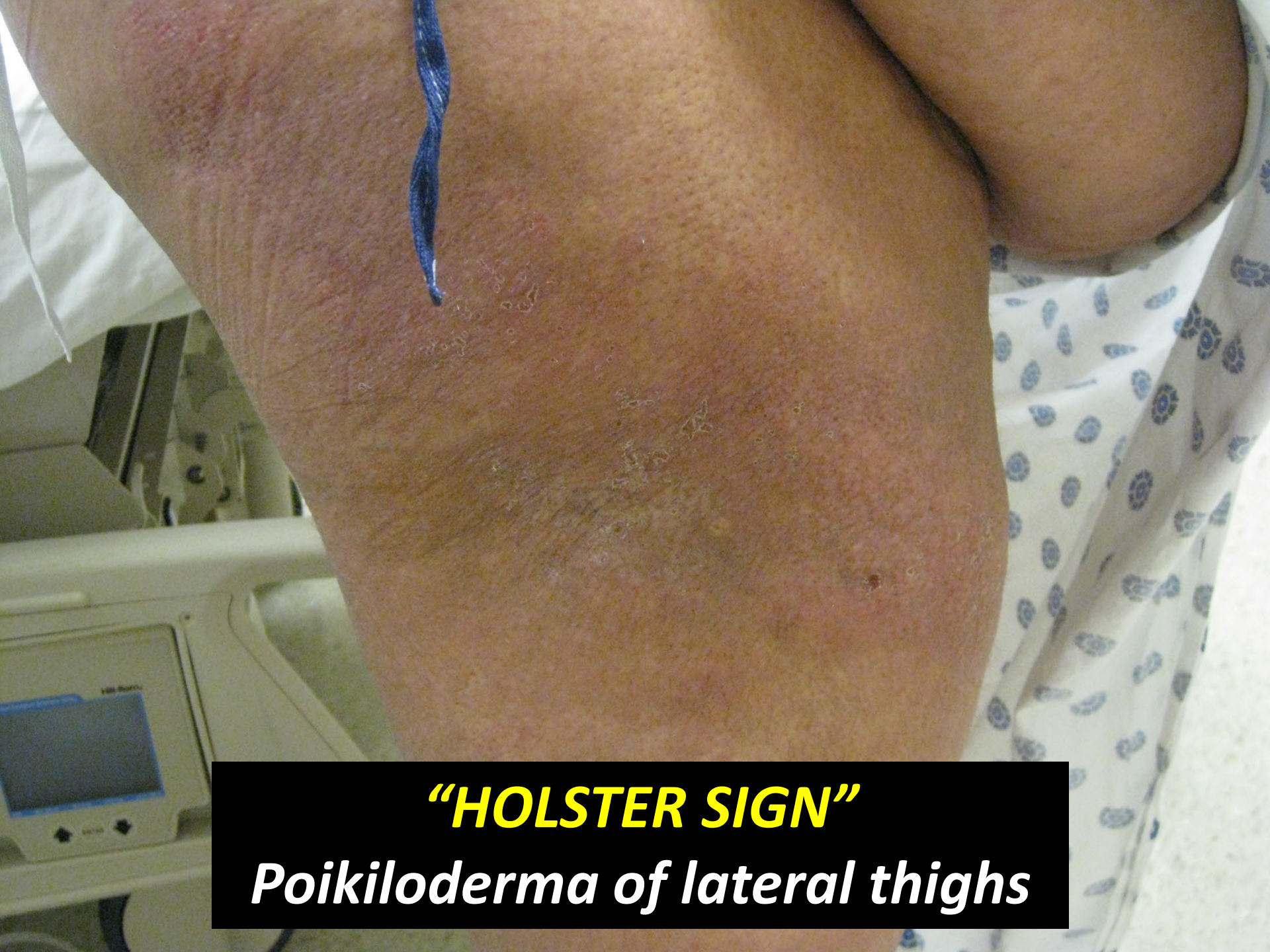


DERMATOMYOSITIS

- **Shawl sign:** *violaceous poikilodermatous* patches/thin plaques on upper chest and upper back, mild scale*

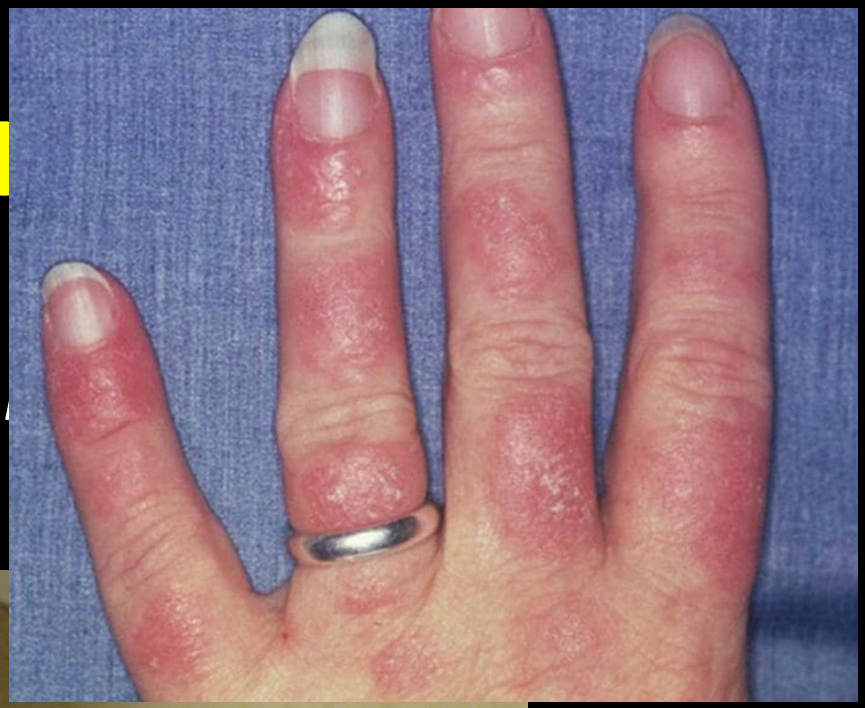


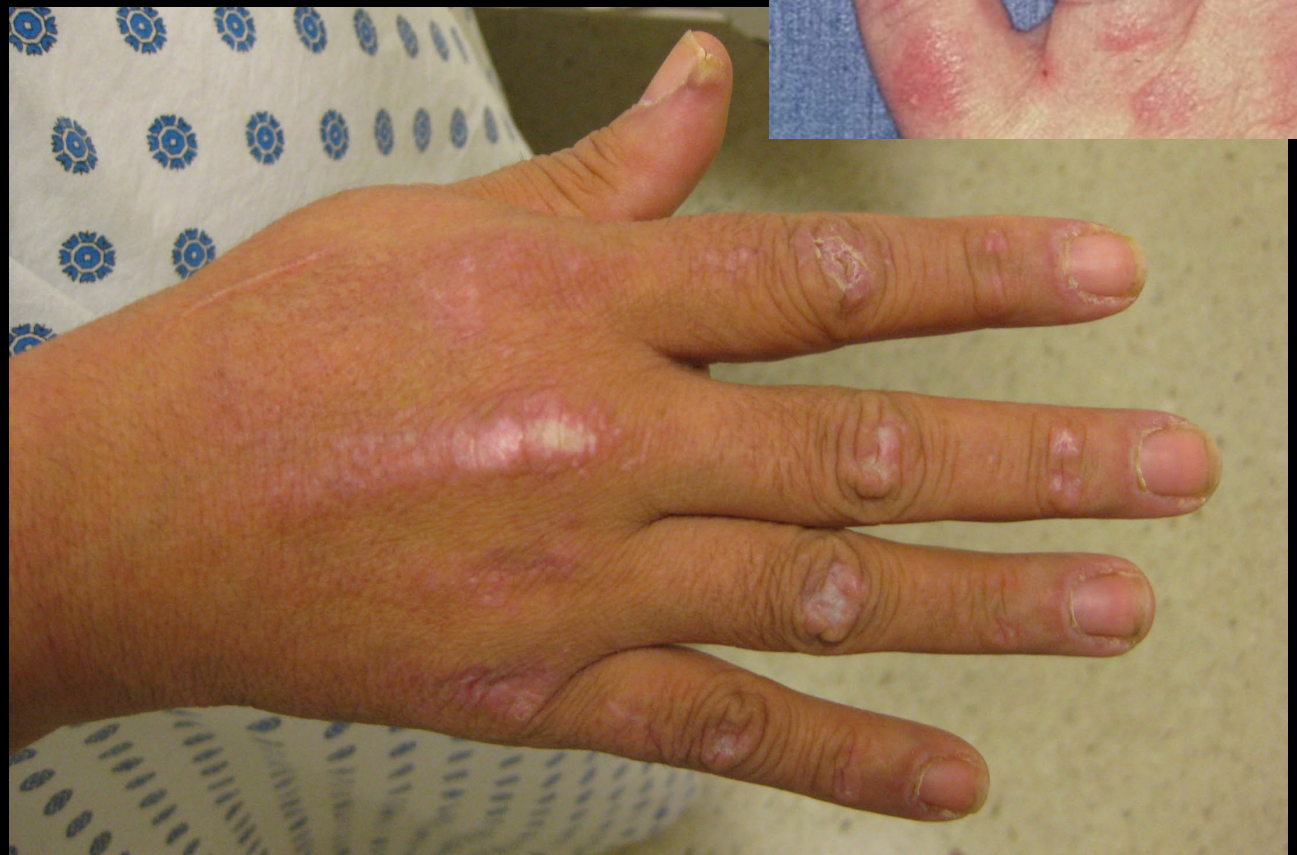
**poikiloderma = hyper- and hypo-pigmento*
telangiectasia and atrophy


"HOLSTER SIGN"
Poikiloderma of lateral thighs

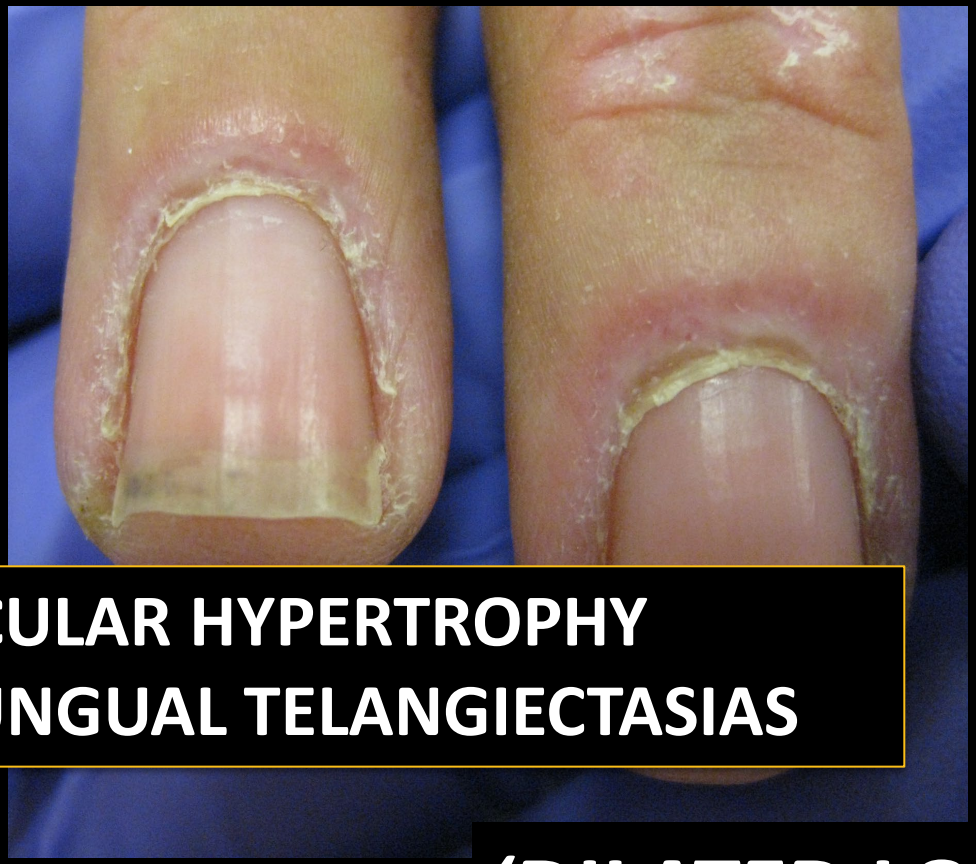
DERMATOM

- **Gottron's papules:** *violaceous*
  over knuckles





DERMATOMYOSITIS:



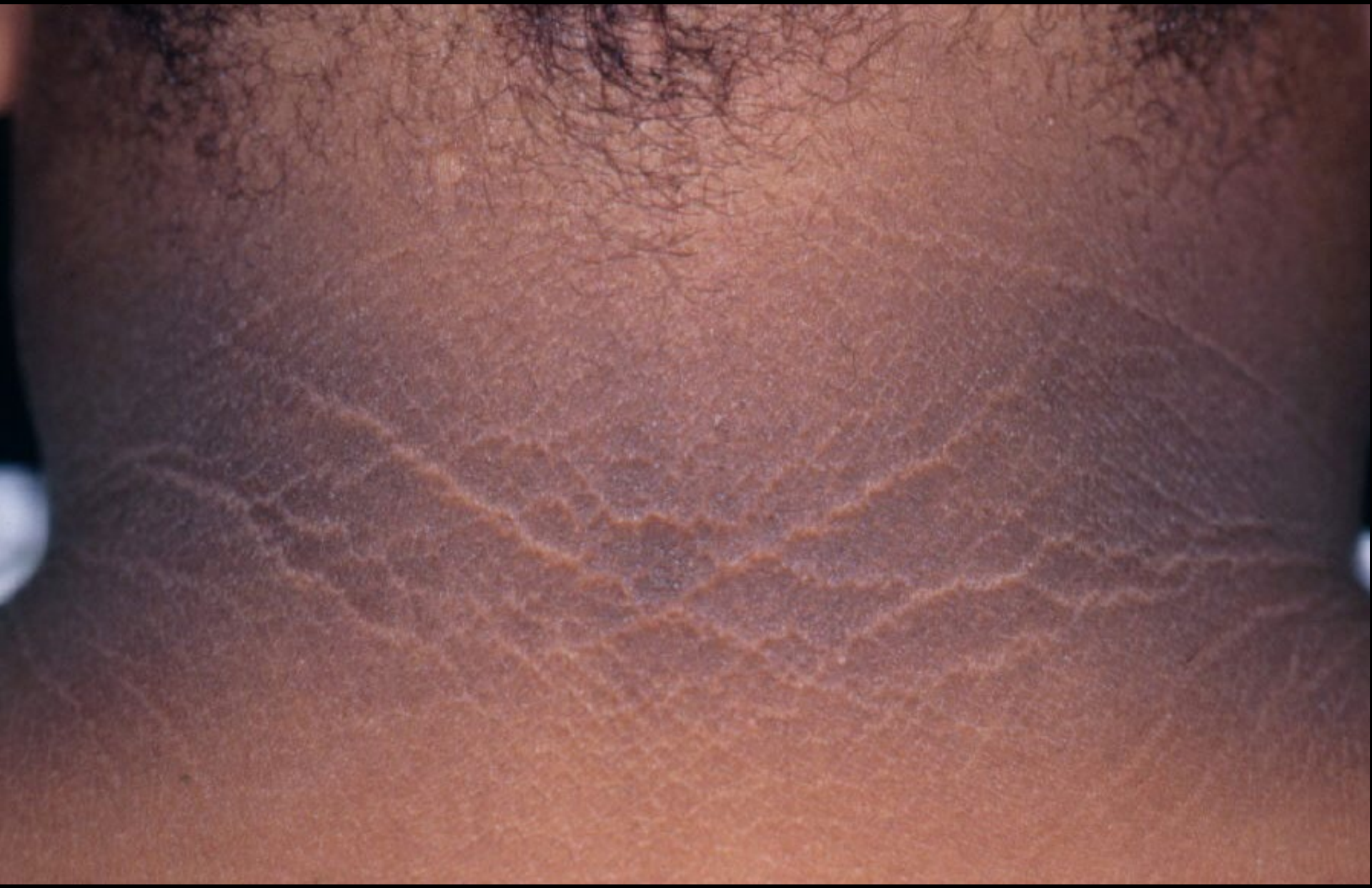
CUTICULAR HYPERTROPHY
PERIUNGUAL TELANGIECTASIAS

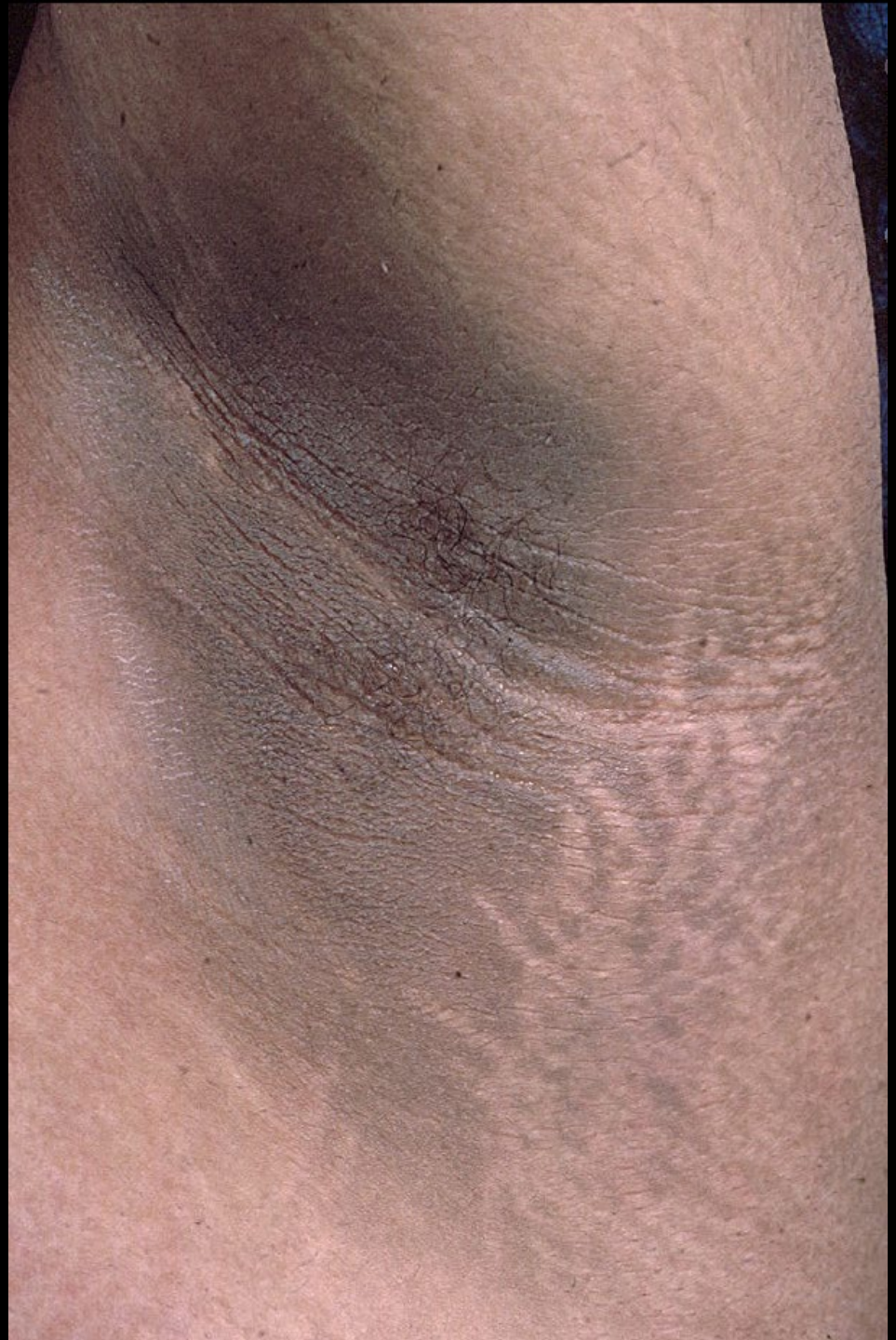
‘DILATED LOOPS’
‘DROP-OUT’

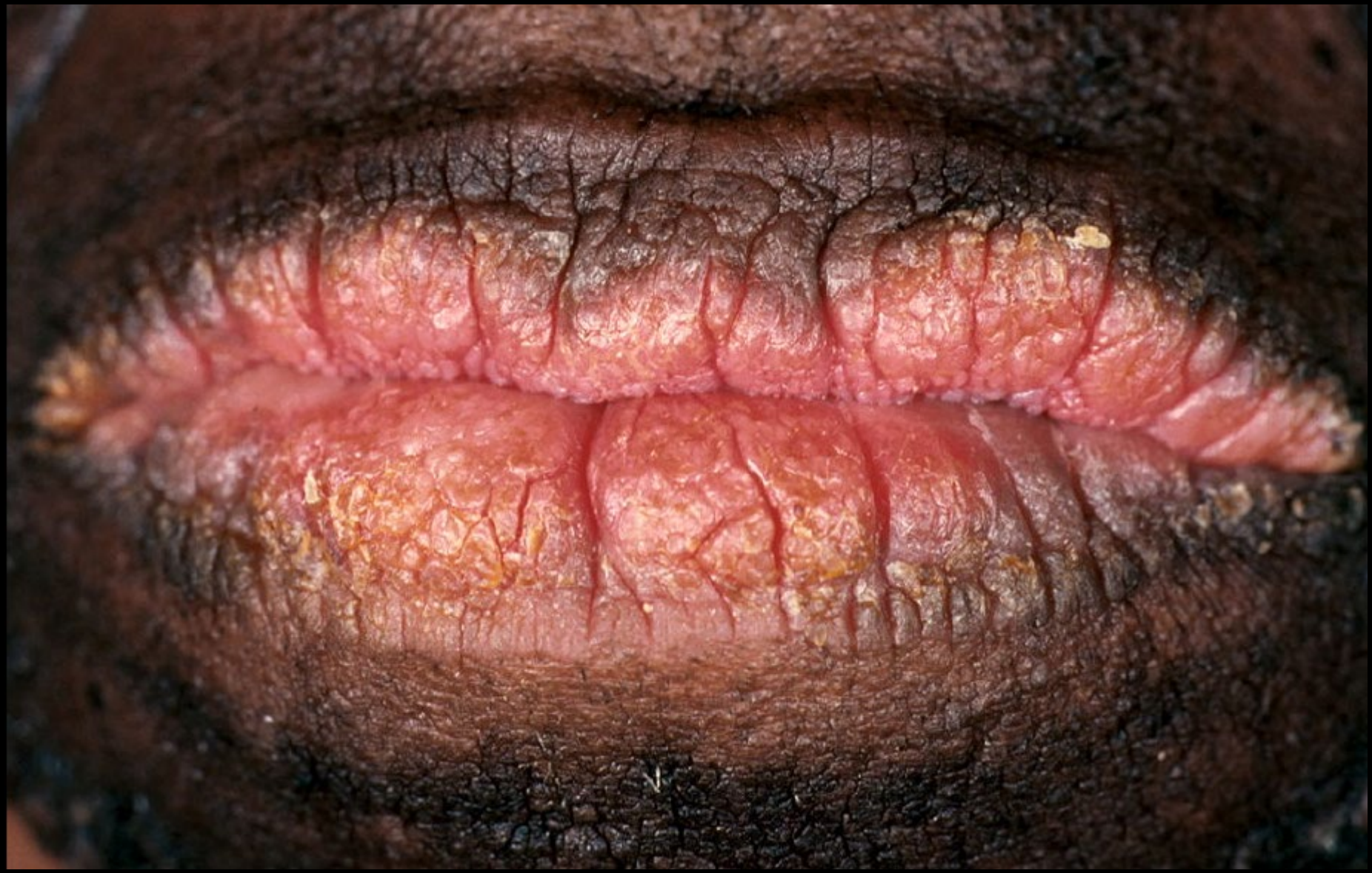
SIGNS OF INTERNAL MALIGNANCY…

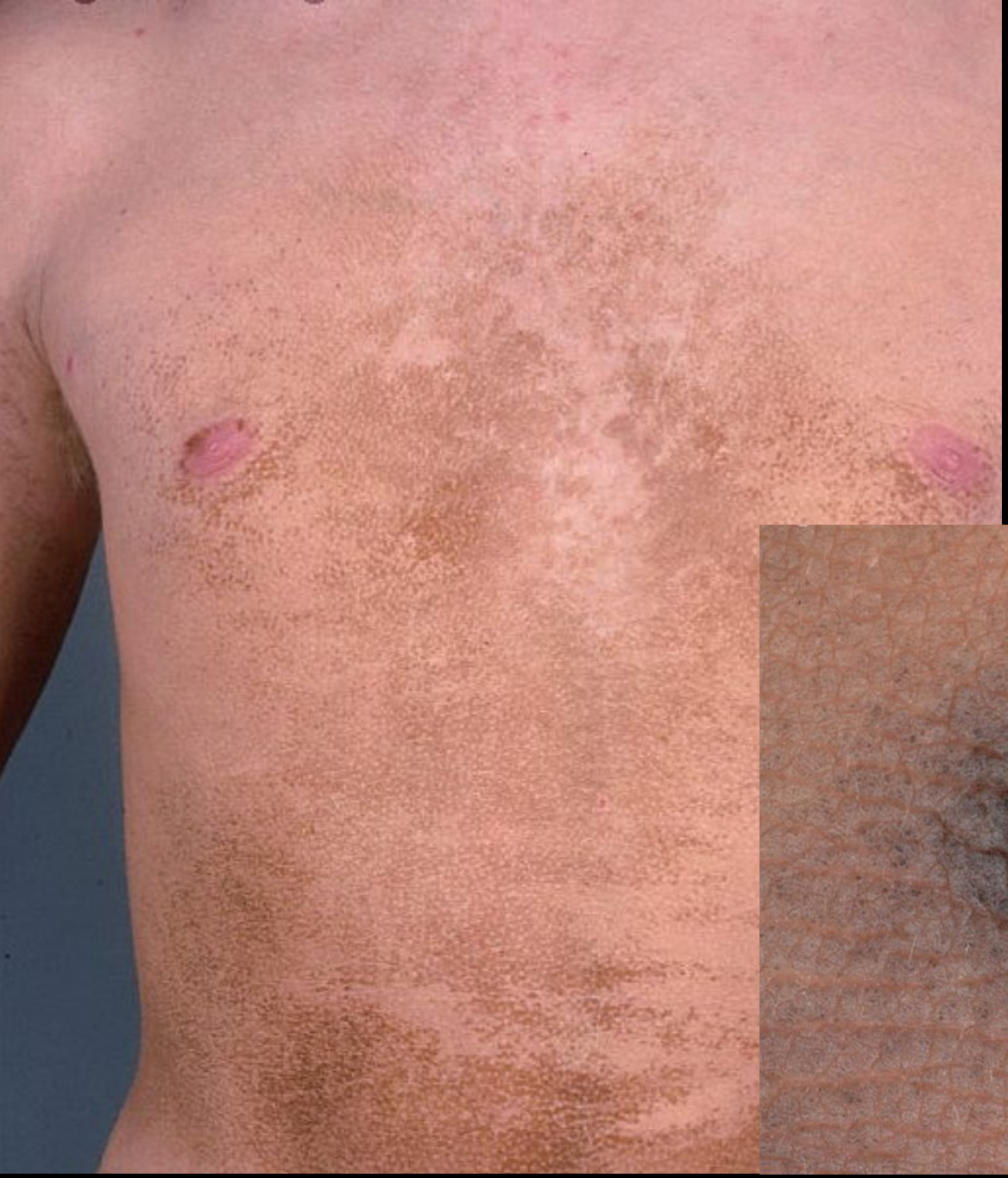
CASE 5:

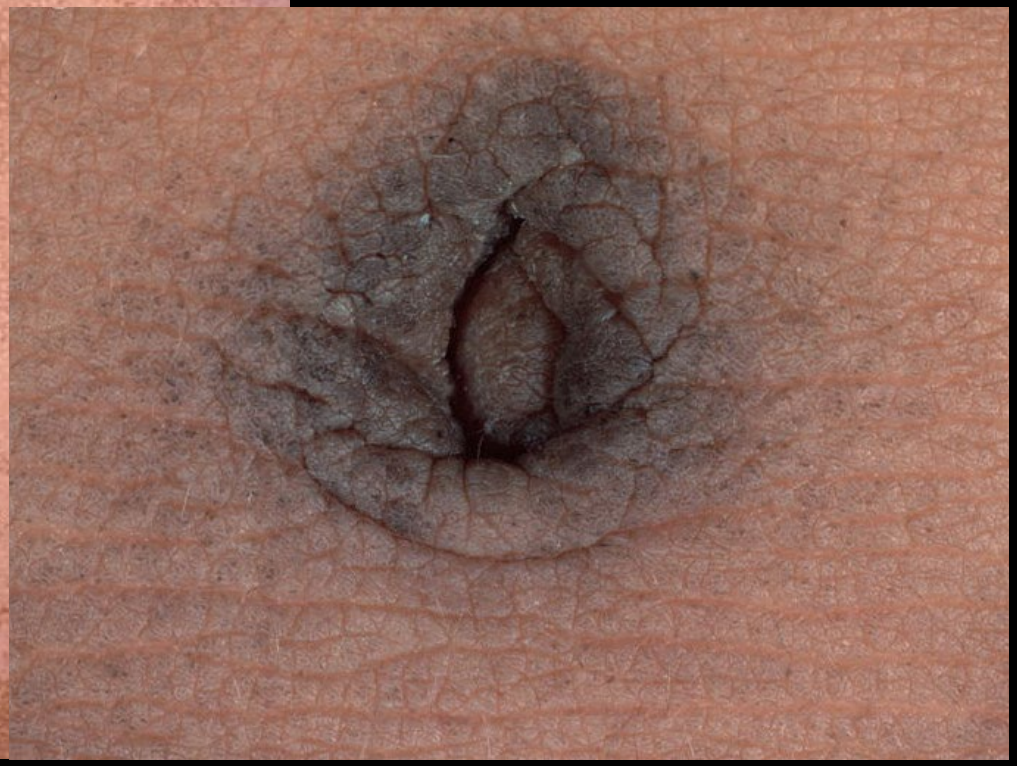
- **45 M presents with brown discoloration on his skin:**
  - ***Thickened velvety plaques are present on his neck and axilla…***







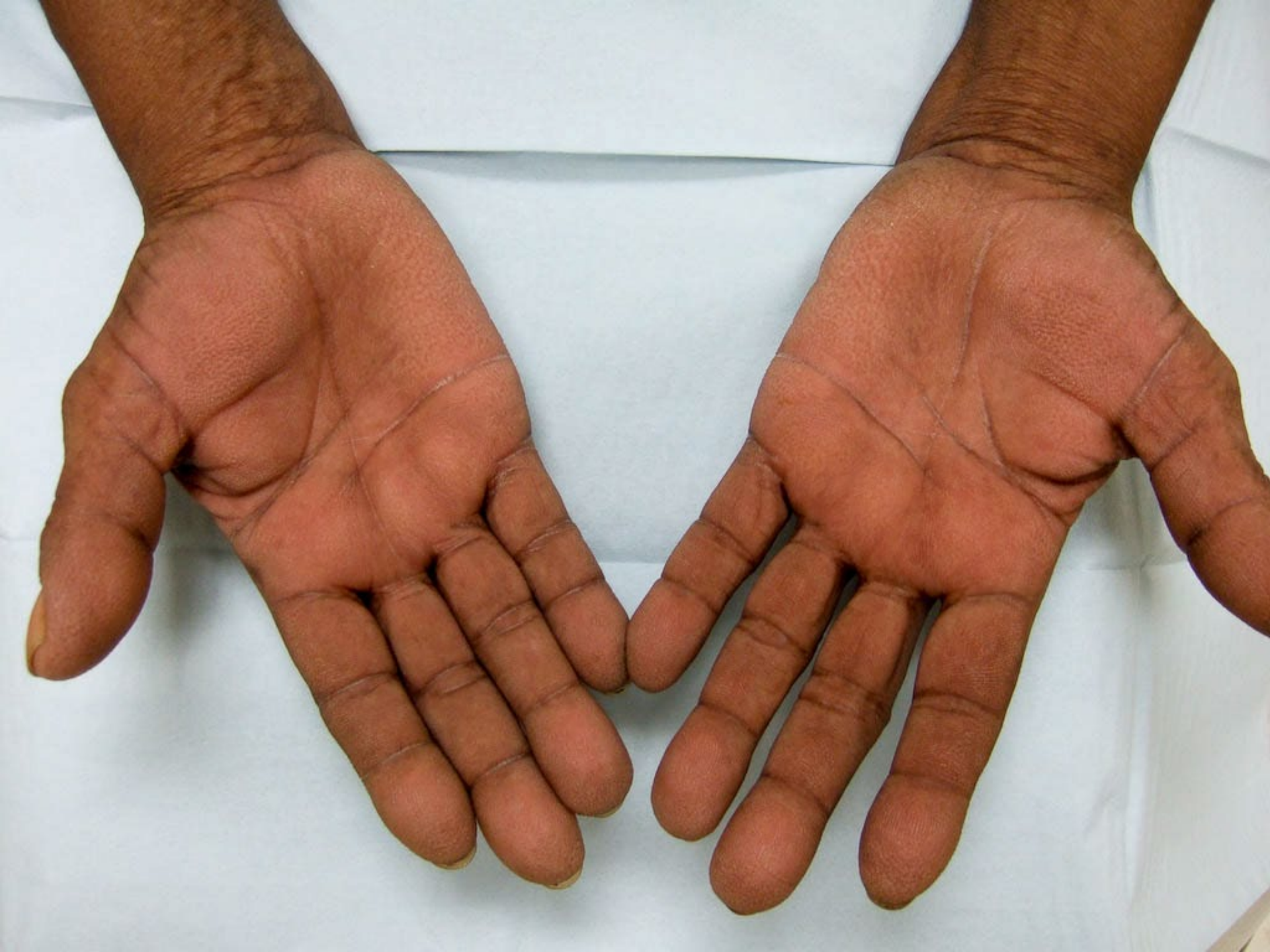


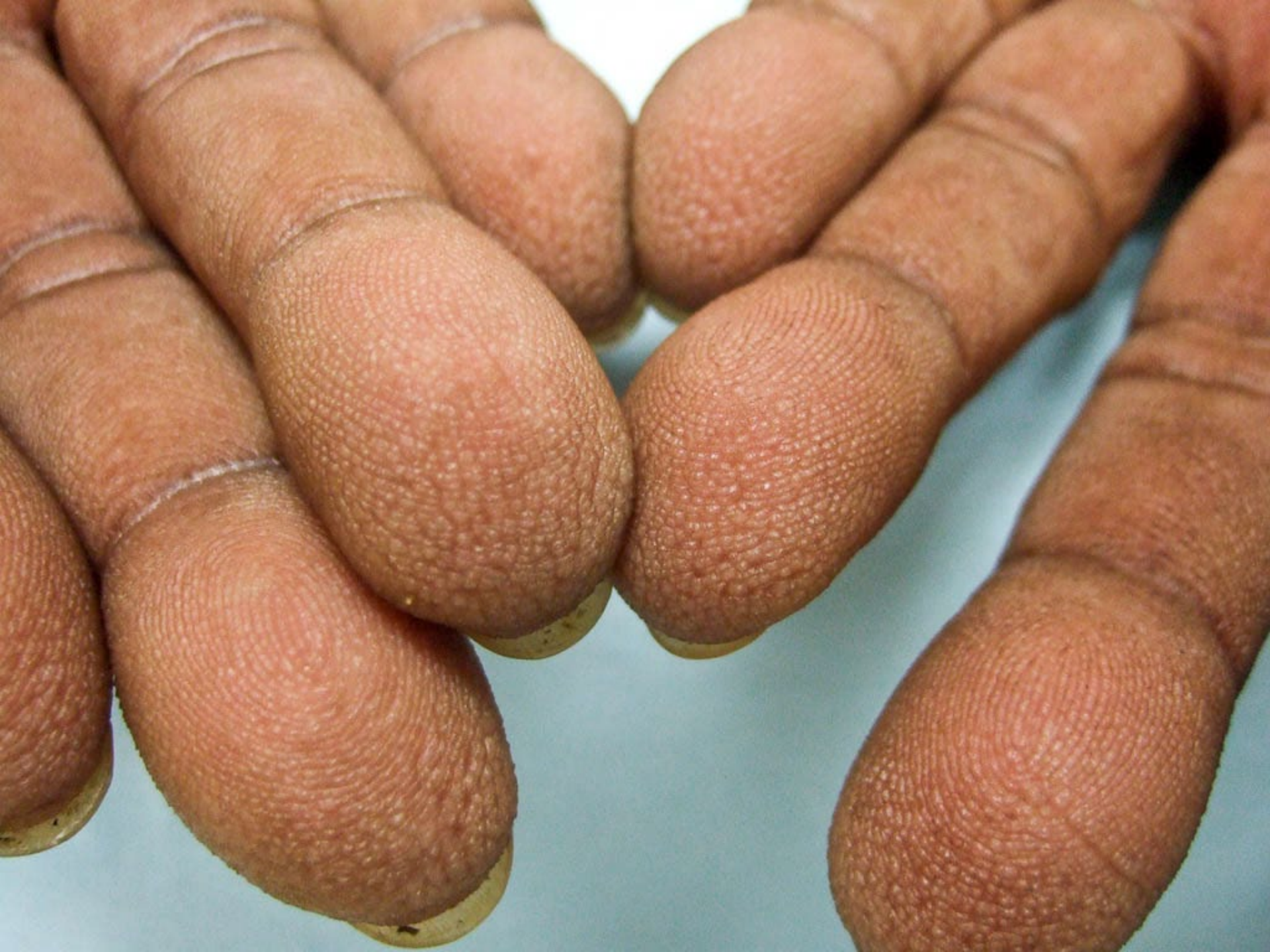


ACANTHOSIS NIGRICANS & CANCER

- **MALIGNANT ACANTHOSIS NIGRICANS IS RARE**
  - **ASSOCIATED MALIGNANCIES:**
    - **ADENOCARCINOMA: GASTRIC, LUNG, BREAST**

- ***WHEN TO WORRY:***
  - ***RAPID ONSET***
  - ***EXTENSIVE, DIFFUSE***
  - ***ATYPICAL LOCATIONS (E.G. MUCOSAL)***





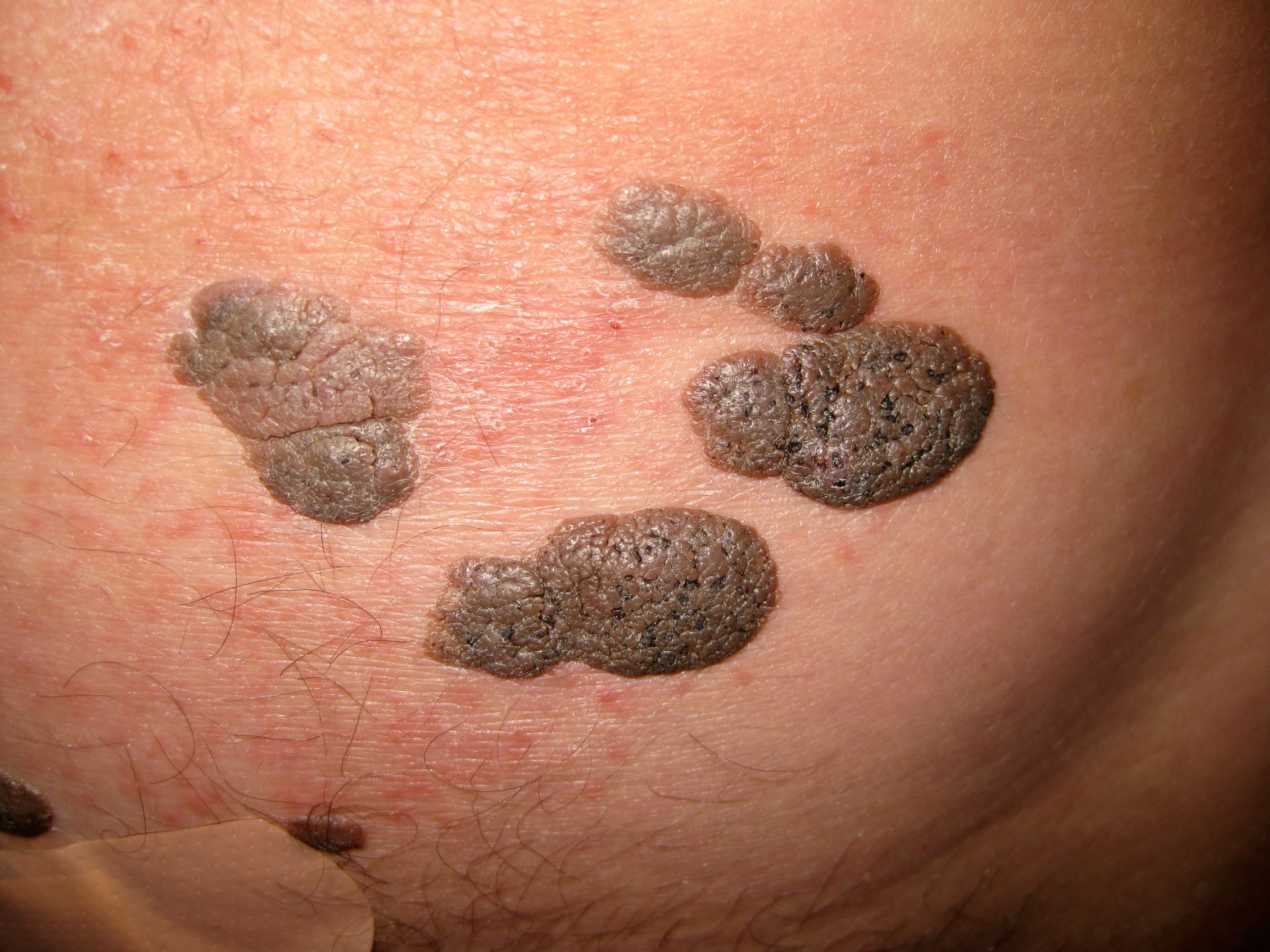

TRIPE PALMS

TRIPE PALMS

- **'RUGOSE' APPEARANCE TO SKIN**
  - ***Said to resemble bovine foregut***
- **ACCENTUATED OVER PRESSURE POINTS**
  - **ASSOCIATED MALIGNANCIES:**
    - **PULMONARY CARCINOMA**
    - **GASTRIC ADENOCARCINOMA**

CASE 6:

- **82 M presents with these “barnacles” that appeared over the last month**
  - ***Exam shows stuck on brown papules***



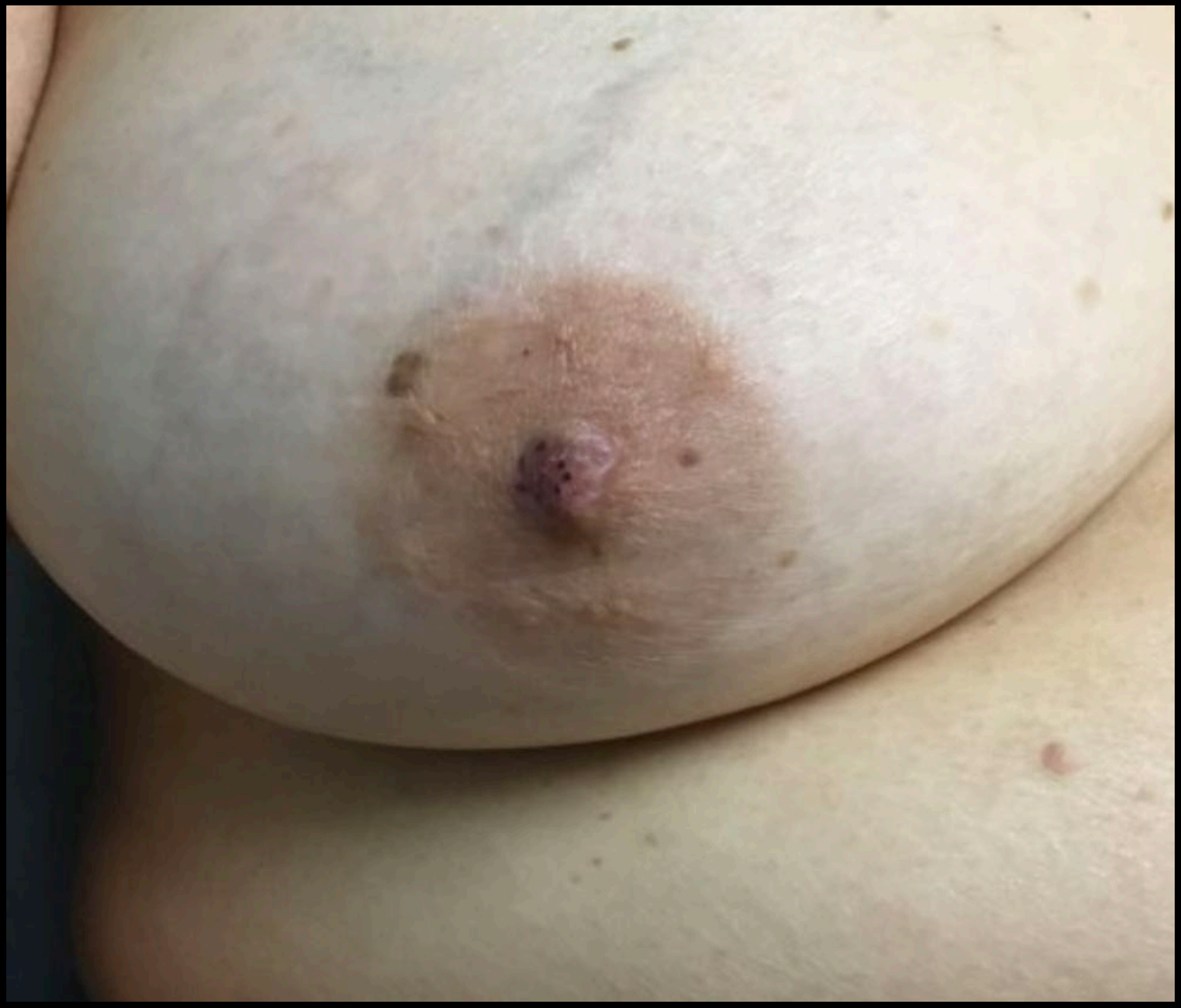
SIGN OF LESER-TRELAT

- **SUDDEN APPEARANCE OF MULTIPLE SEBORRHEIC KERATOSES**
  - *AS SIGN OF INTERNAL MALIGNANCY*

- **ASSOCIATED MALIGNANCIES:**
  - **LYMPHOPROLIFERATIVE DISORDERS, OTHERS**

CASE 7

- 50 W with scaling around areola
  - Remote history of atopic dermatitis
  - Not itchy, no discharge
  - Mammogram recently was normal



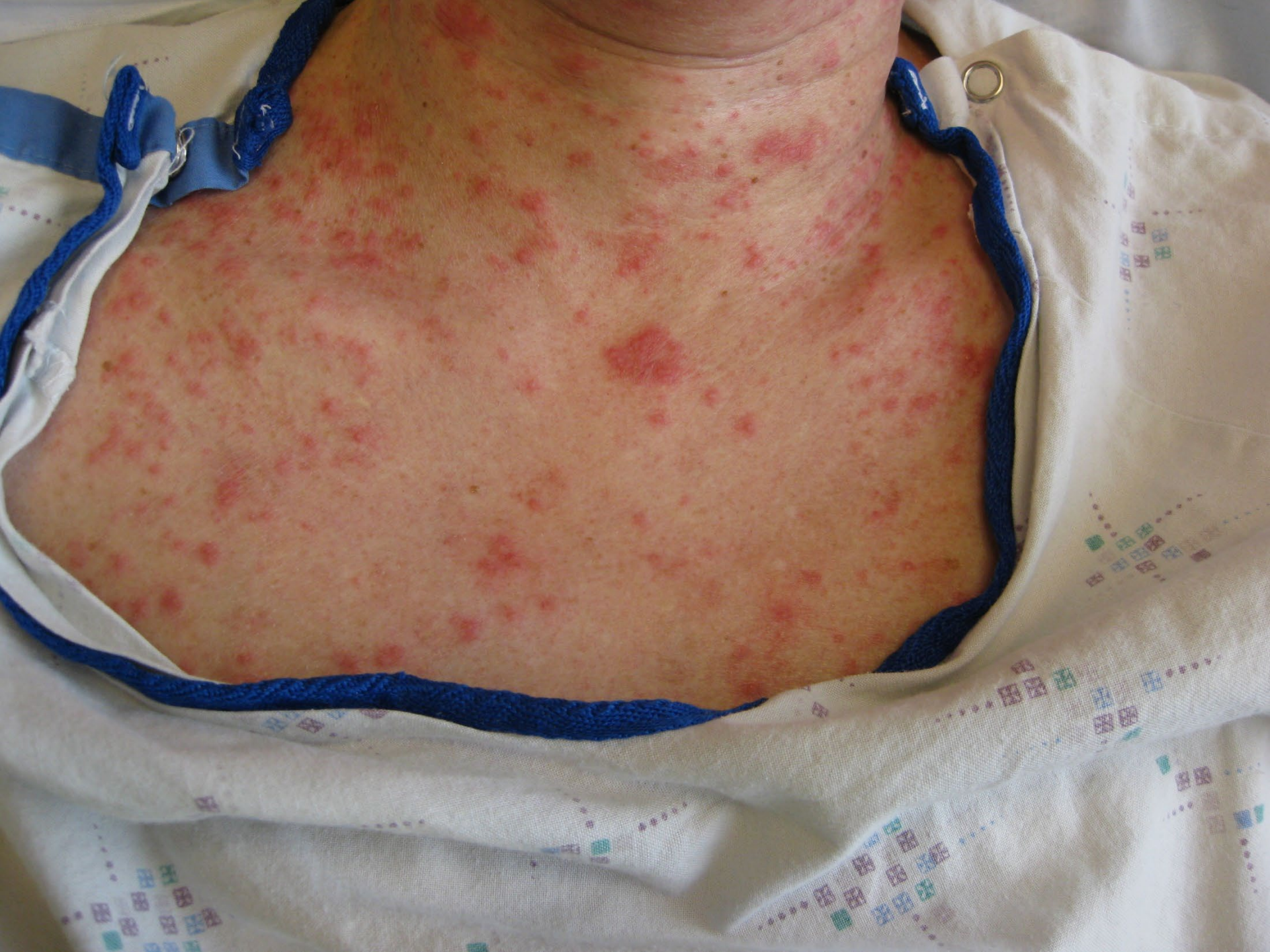
Paget's Disease

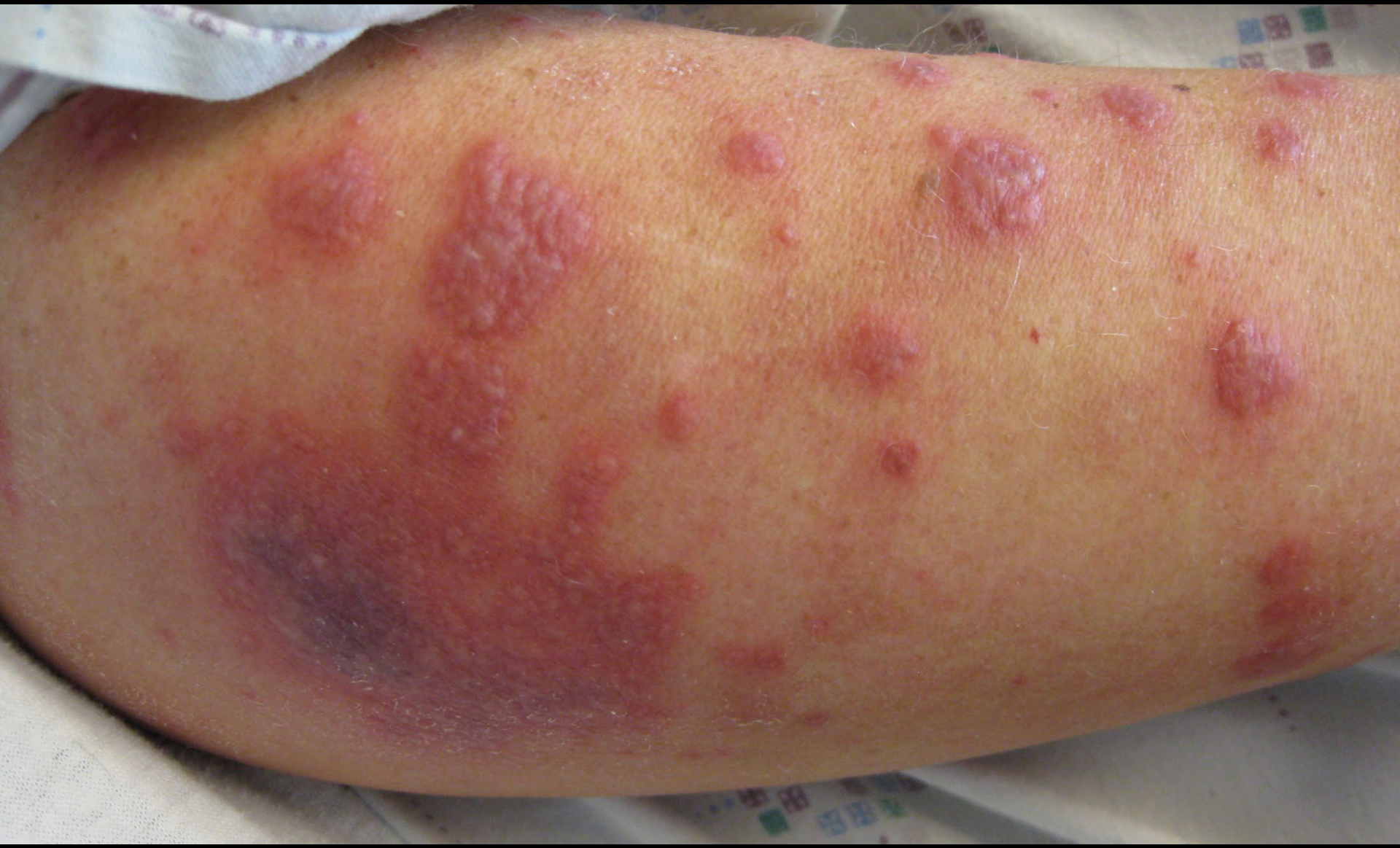
- Eczema like changes around the nipple
- Nipple may be inverted
- May have straw colored discharge
- Underlying ductal carcinoma
  - Clues:
    - Area not itchy
    - Not bilateral
    - Not responsive to topical steroids

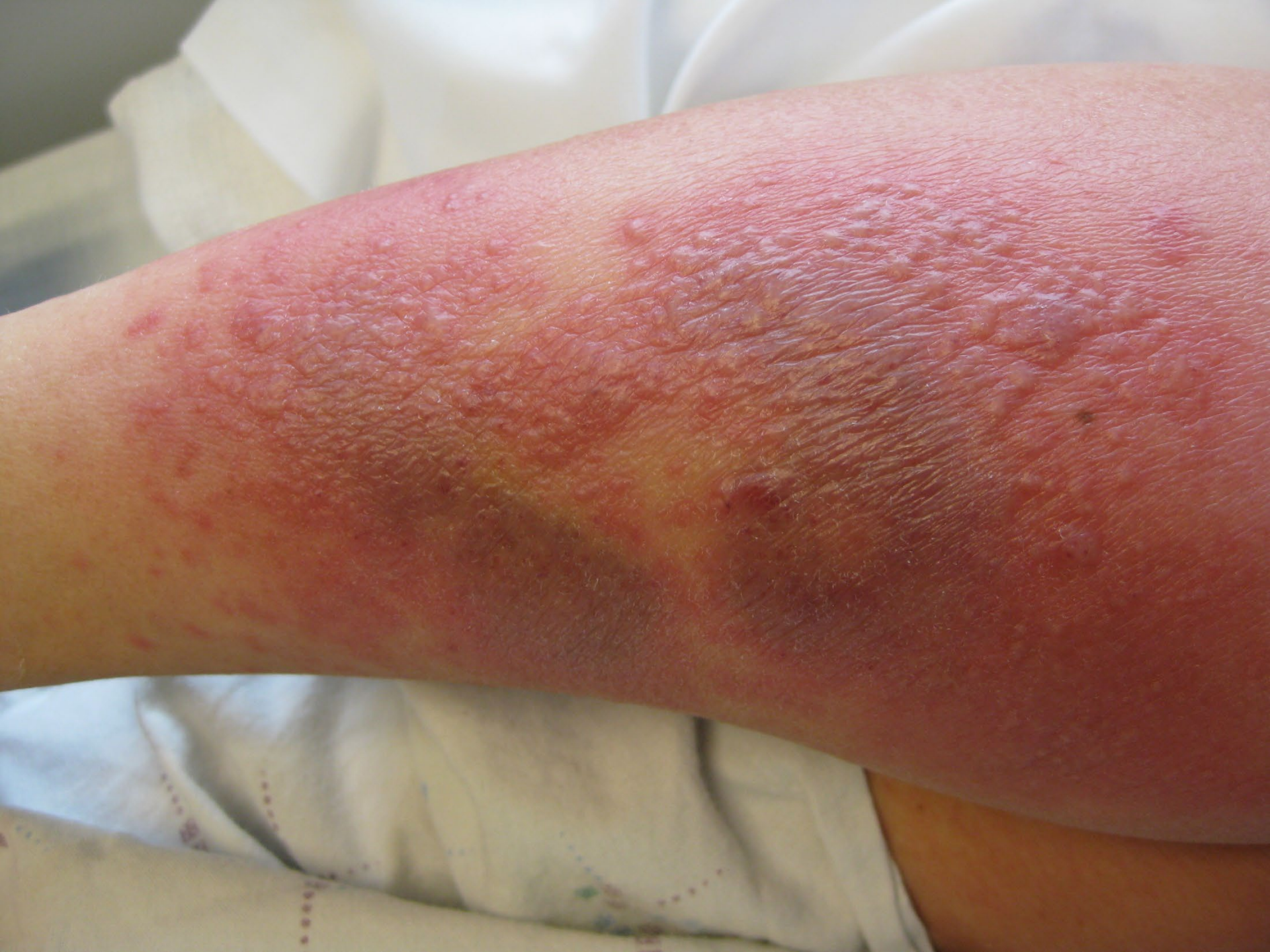
NEUTROPHILIC DERMATOSES …

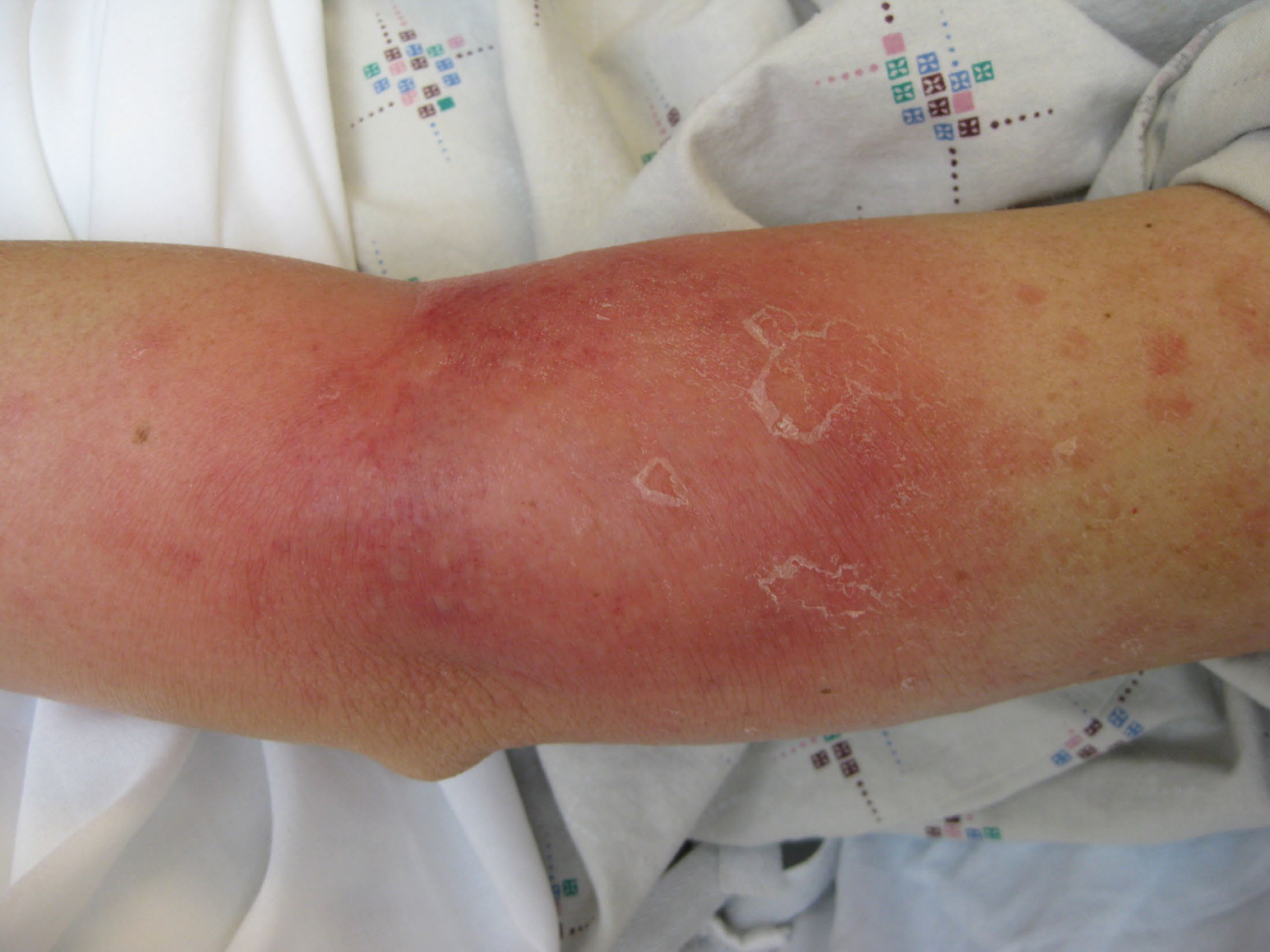
CASE 8:

- **28 F 1 day post partum**
  - ***Fever***
  - ***Edematous pseudovesicular plaques on trunk and extremeties***









SWEET'S SYNDROME

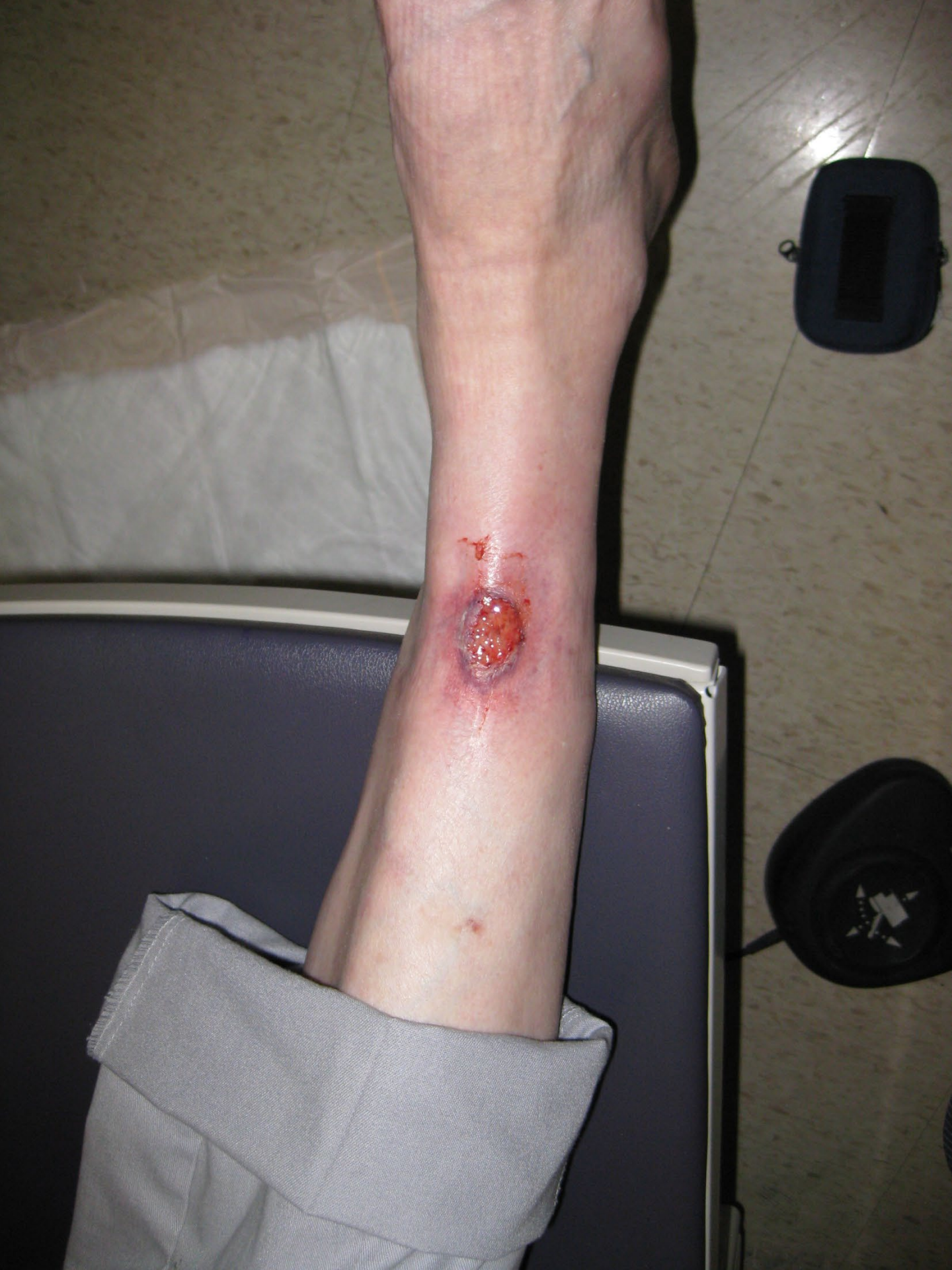
- **TENDER 'PSEUDOVESICULAR' ERYTHEMATOUS PLAQUES + FEVER + NEUTROPHILIA**

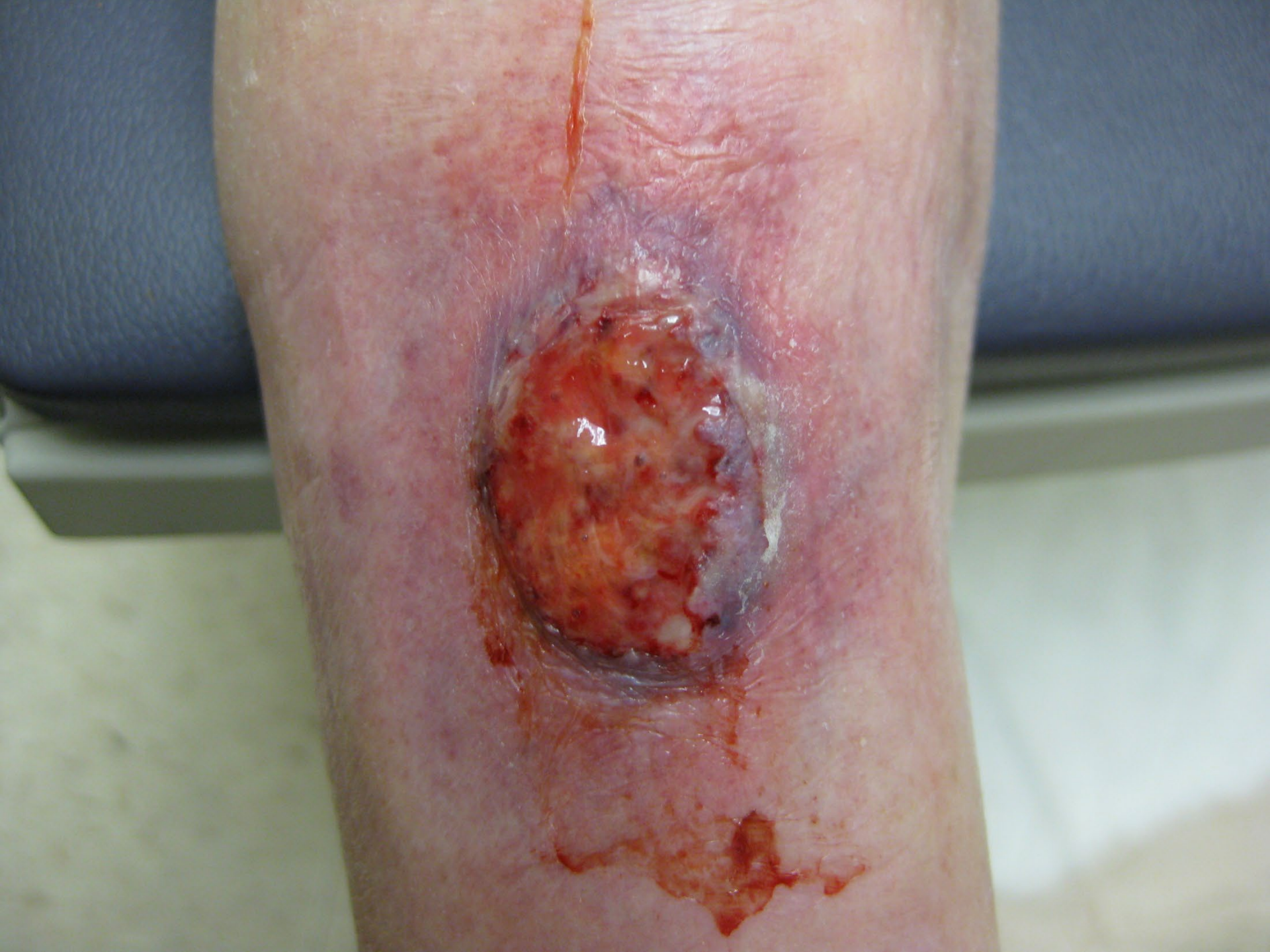
- Dz Associations:
  - *'Classic'-* ***post-infectious, IBD, pregnancy***
  - *Malignancy-assoc:* ***AML, MDS***
  - *Drug-induced:* ***G-CSF (neupogen), All-trans-retinoic-acid***

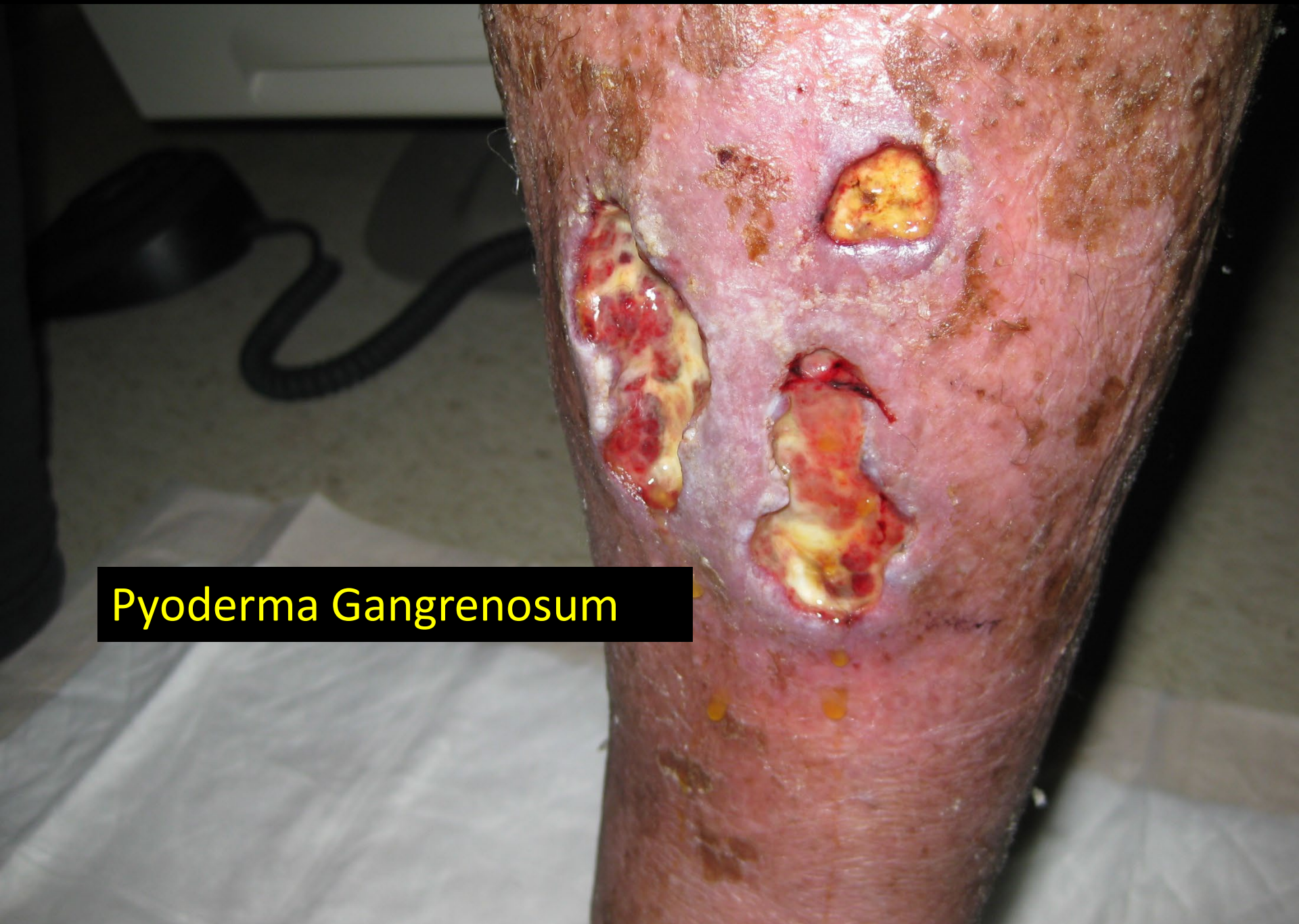
- Rx: Responds quickly to high dose Prednisone

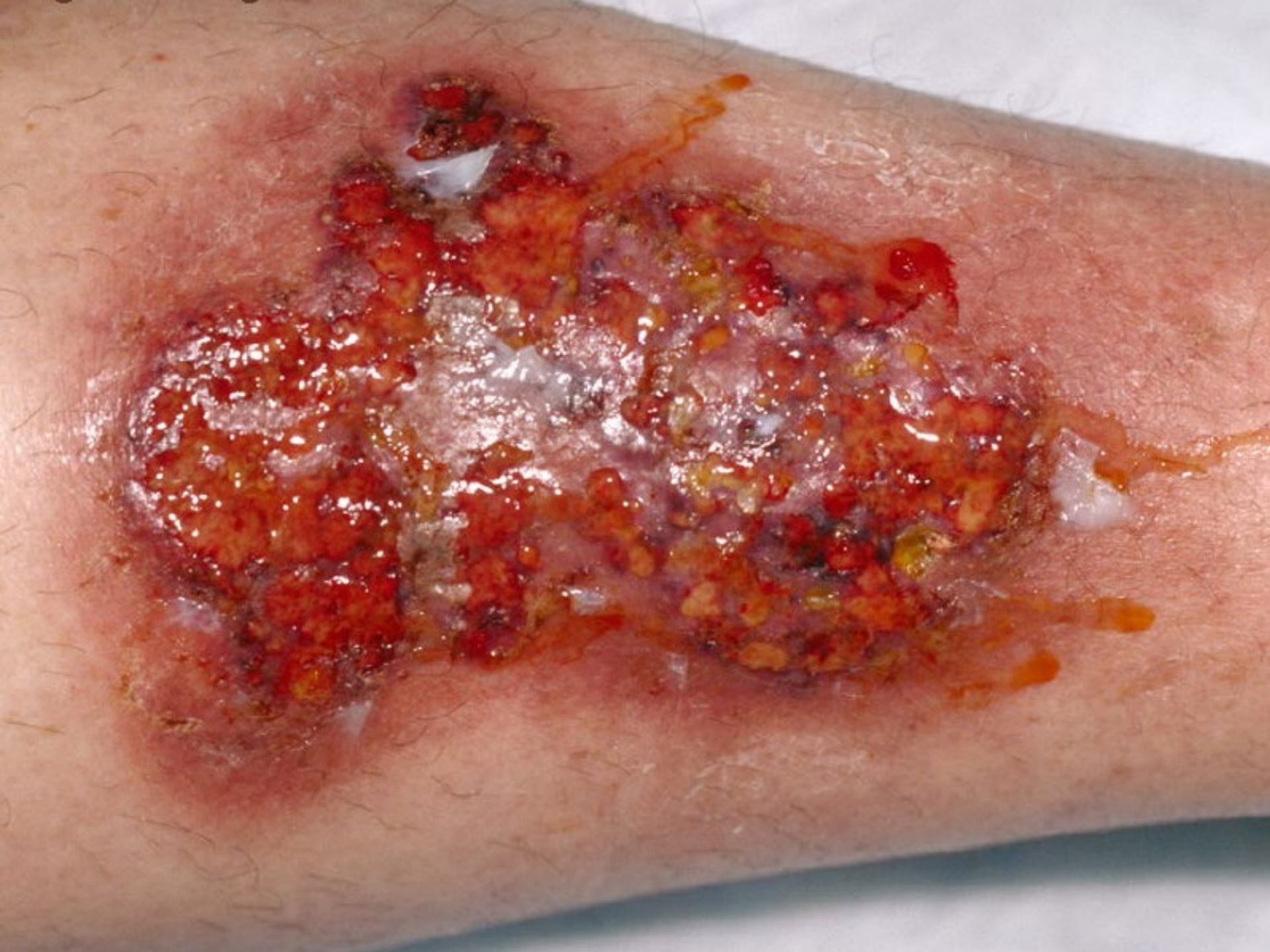
CASE 9:

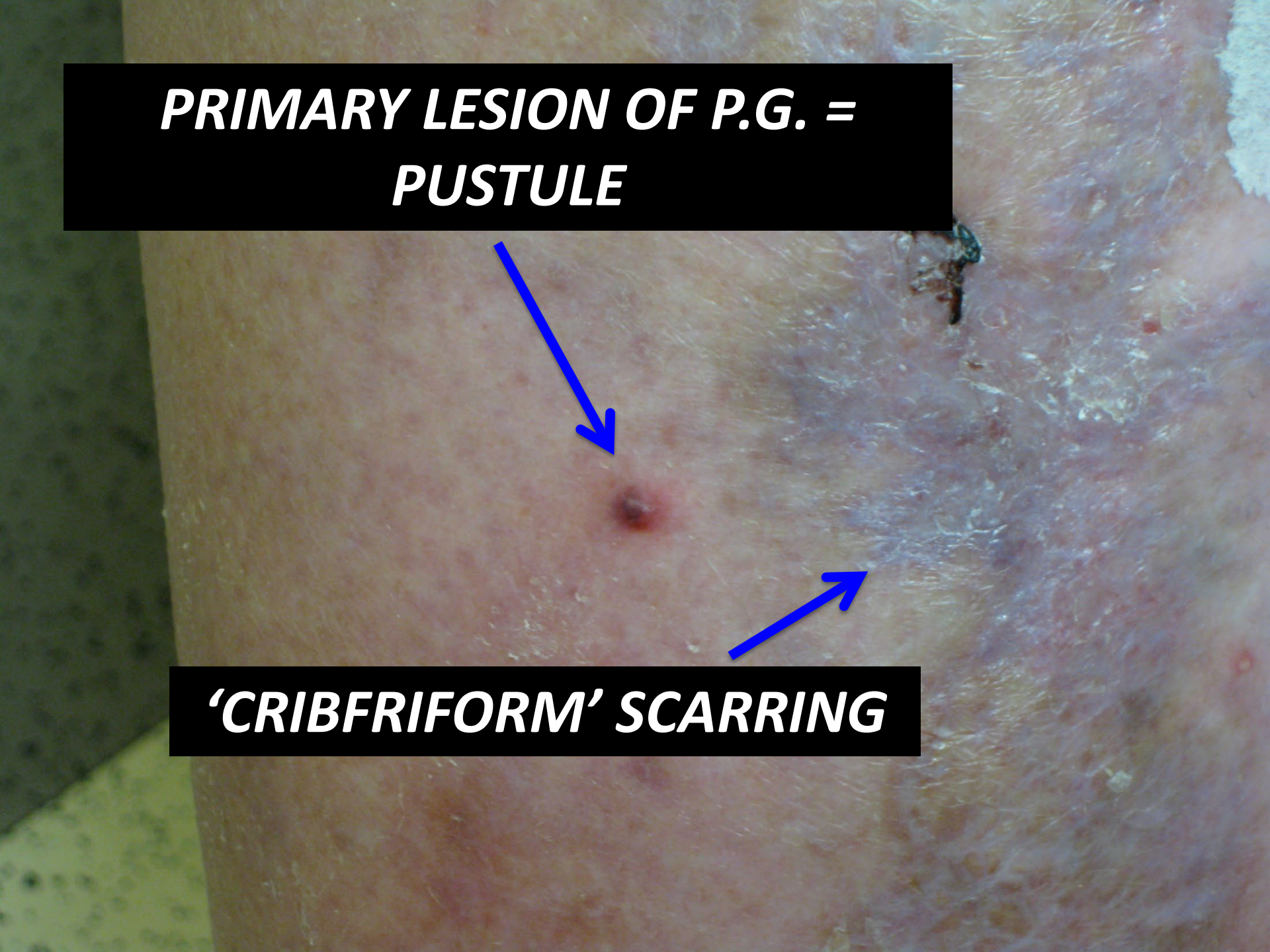
- **60 M with lower leg ulceration**
  - ***Was debrided by wound care but continues to get worse***
  - ***Also notes he has some intermittent diarrhea***
  - ***Painful***






Pyoderma Gangrenosum




PRIMARY LESION OF P.G. = PUSTULE
'CRIBFRIFORM' SCARRING

PYODERMA GANGRENOSUM

- **PAINFUL**, QUICKLY EXPANDING ULCER
- PURULENT, **UNDERMINED VIOLACEOUS BORDER**
- **WORSE WITH DEBRIDEMENT**

- VARIANTS:
  - *Classic: **lower legs most common***
  - *'Atypical PG': coalescing pustules or bullae*
  - *Peristomal PG: around stoma, often for IBD patients*

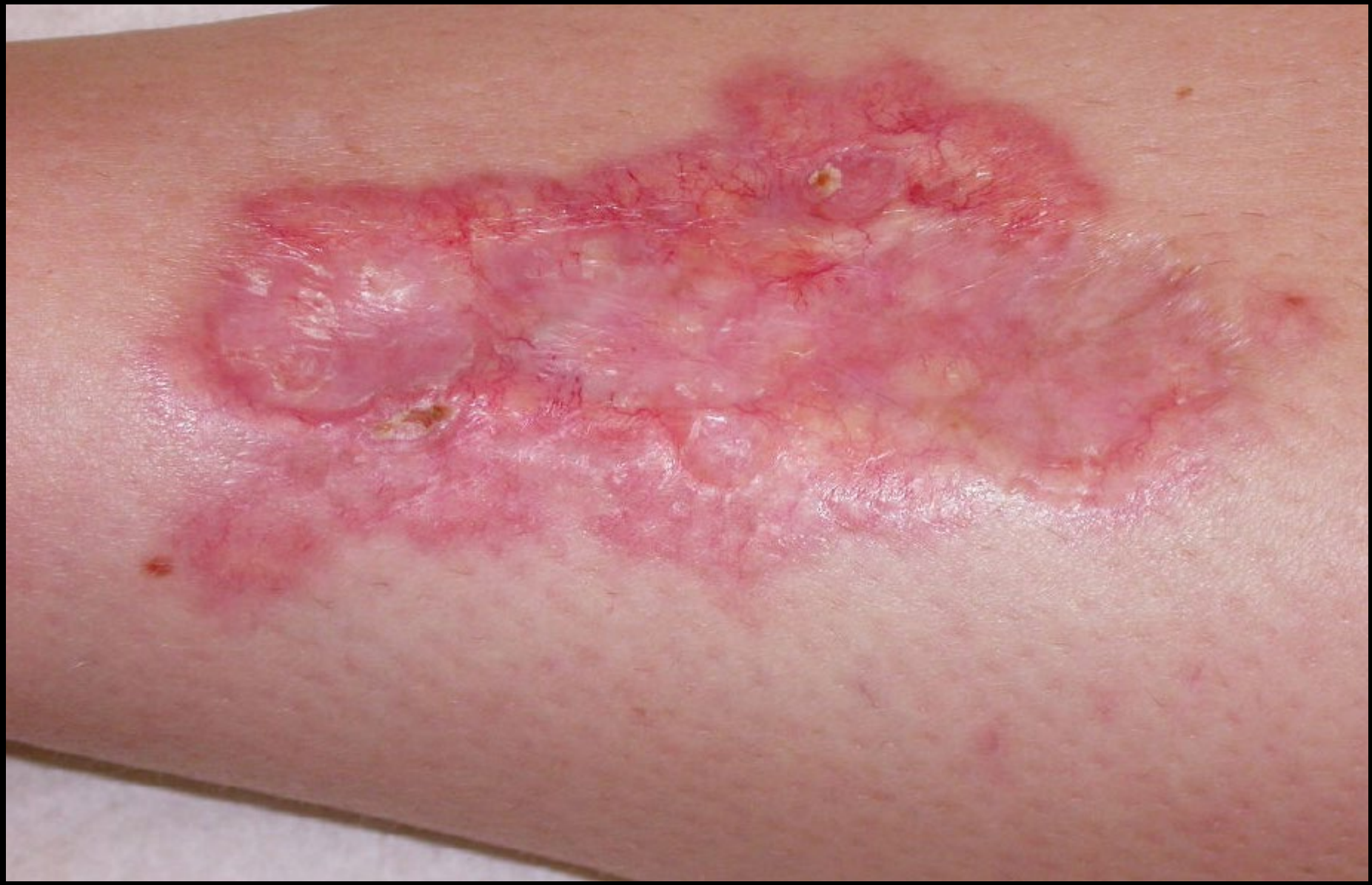
- Disease Associations:
  - **IBD, Arthritis (RA, etc), Hematologic malignancies**
  - Up to 50% idiopathic

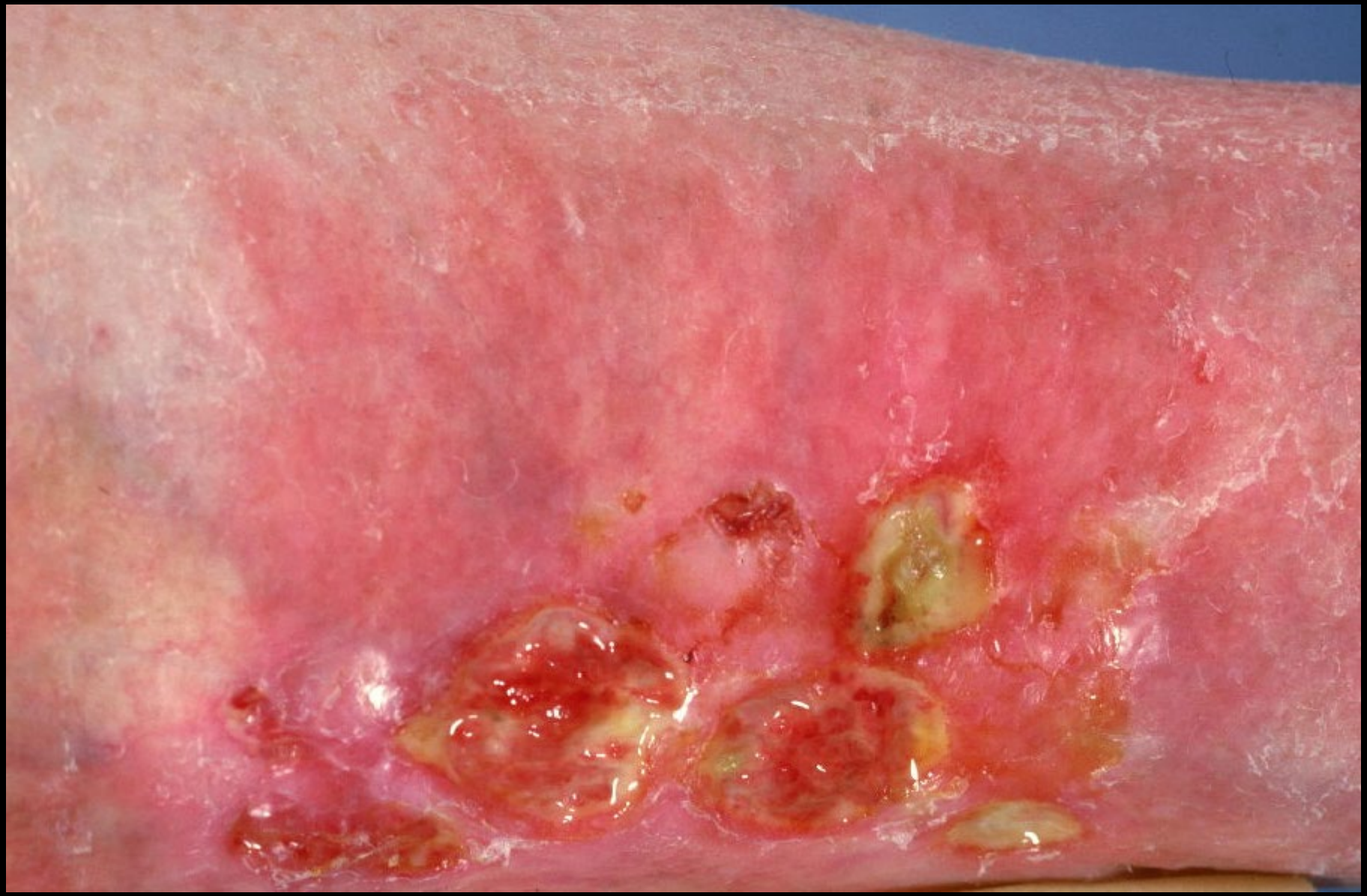
- **DIAGNOSIS OF EXCLUSION**
  - **Rule out vascular causes and infection**

ENDOCRINE DISEASES …

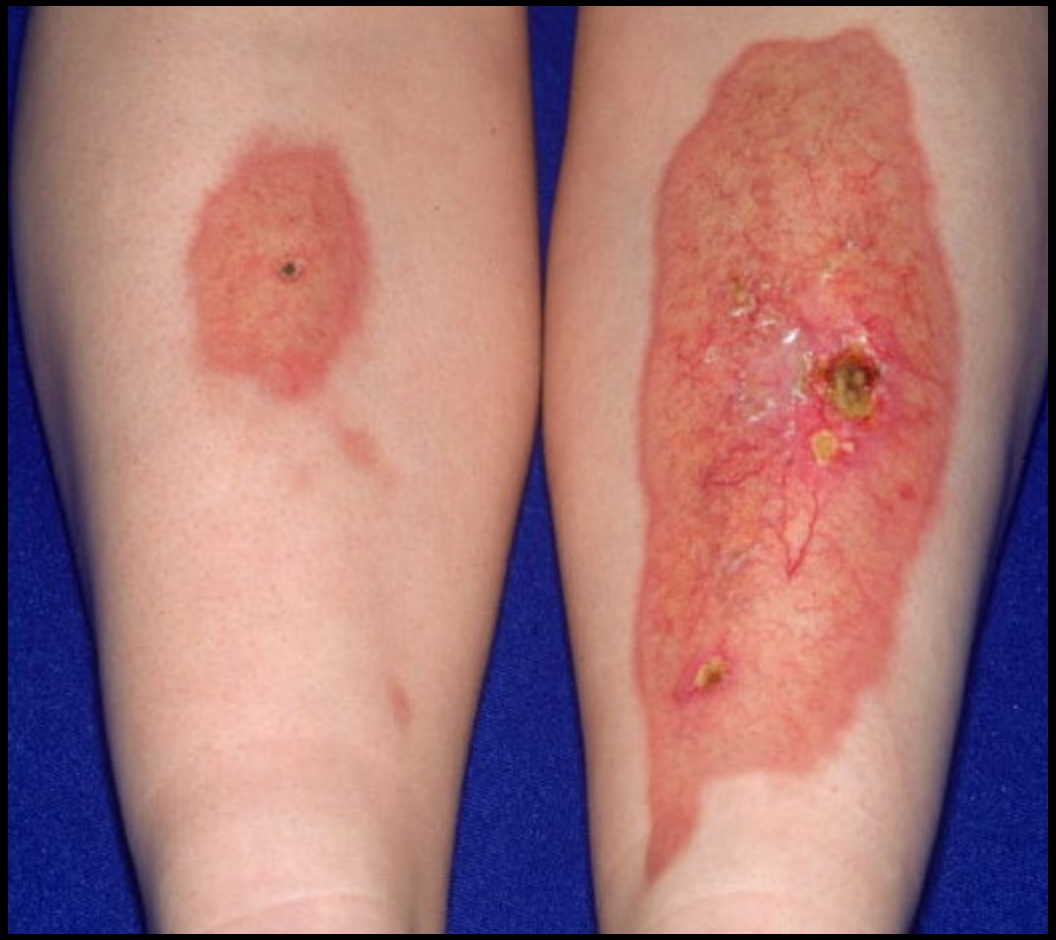
CASE 10:

- **62 F with lower leg skin color changes**
  - ***Not very symptomatic, just doesn't like the color***
  - ***Sometimes it ulcerates***
  - ***HbA1C 8***





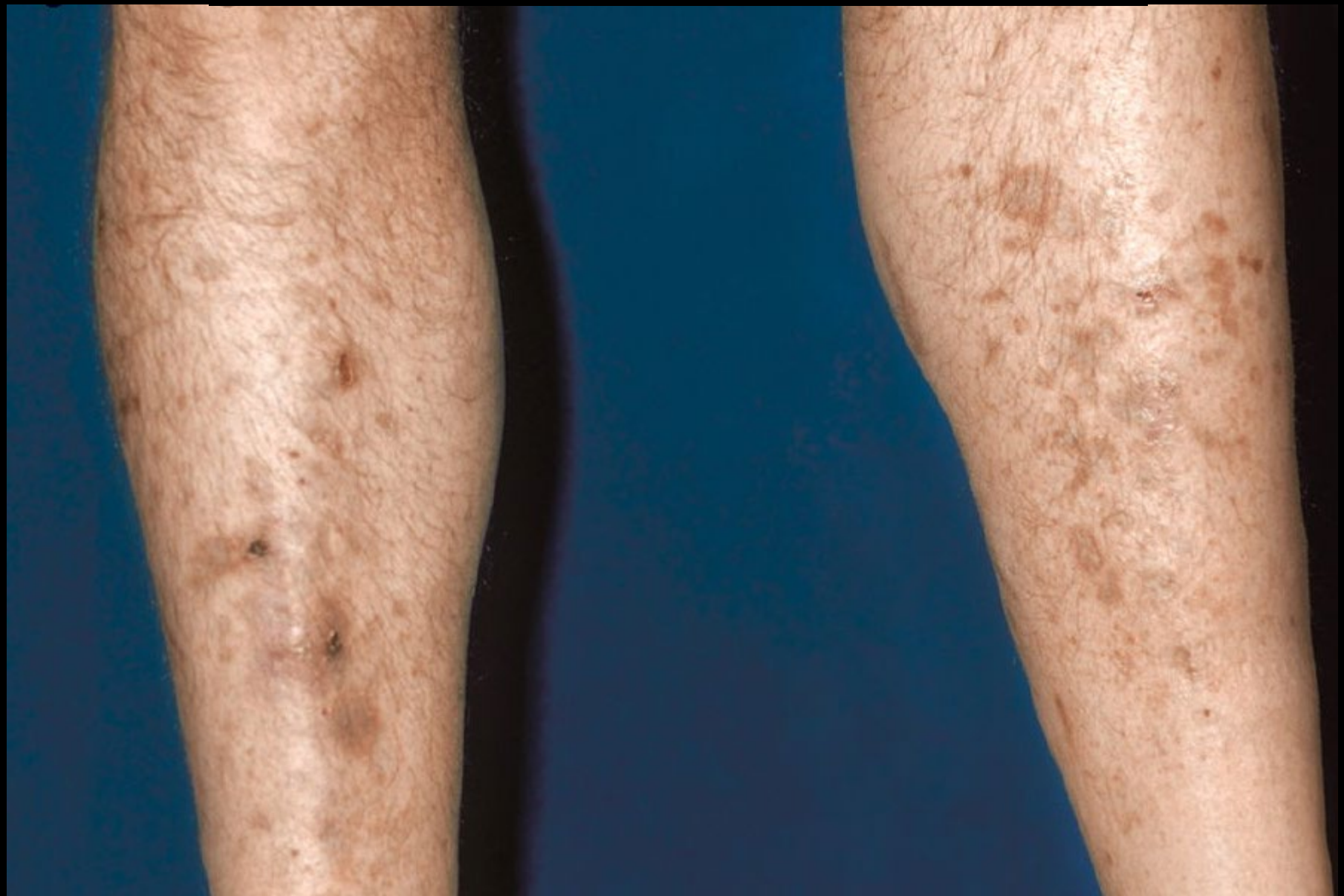
NECROBIOSIS LIPOIDICA (DIABETICORUM)

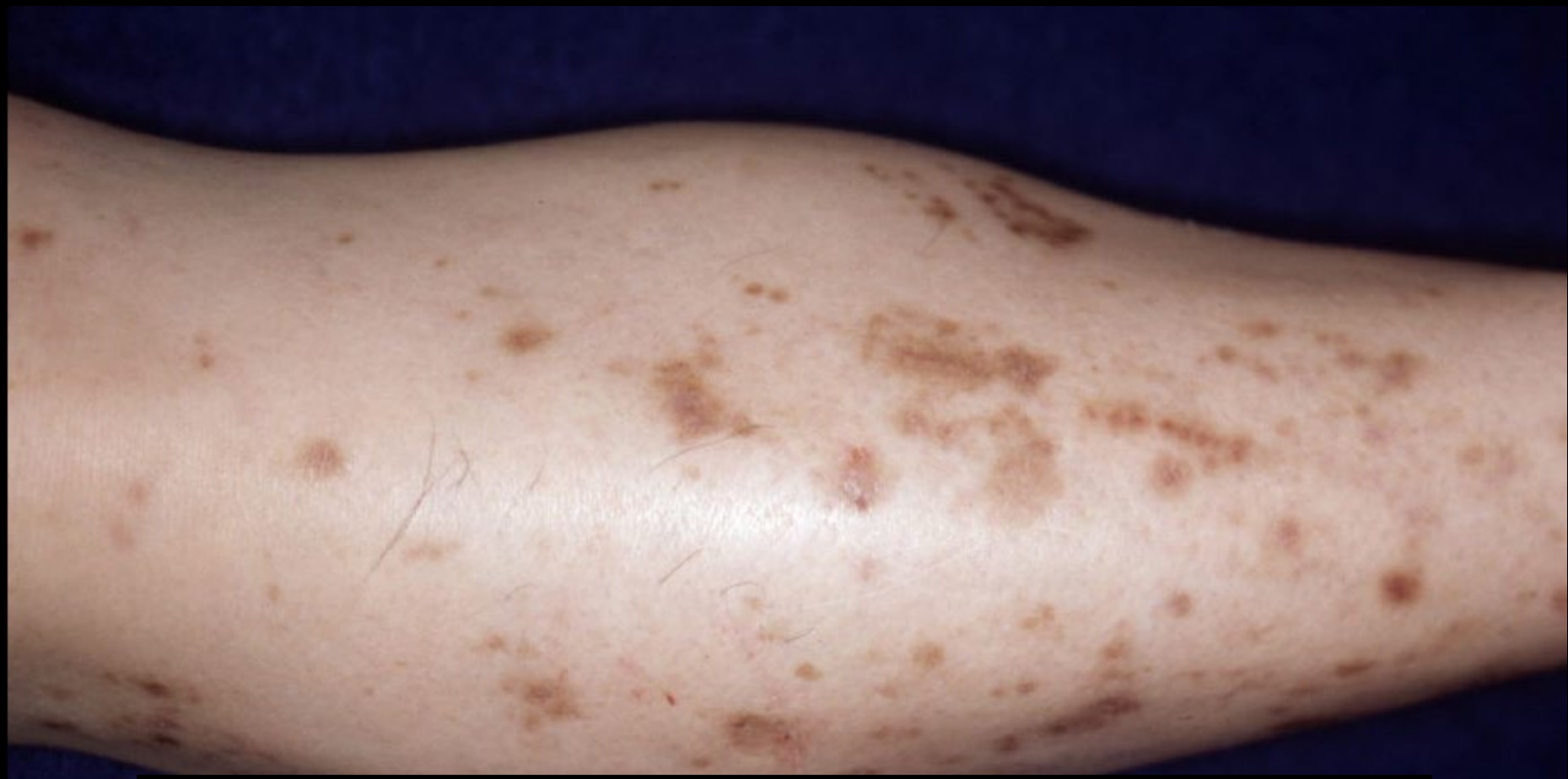


NECROBIOSIS LIPOIDICA DIABETICORUM

- *Type 1 or type 2 DM*
- *Can be seen in prediabetics*
- *Treat with topical steroids*

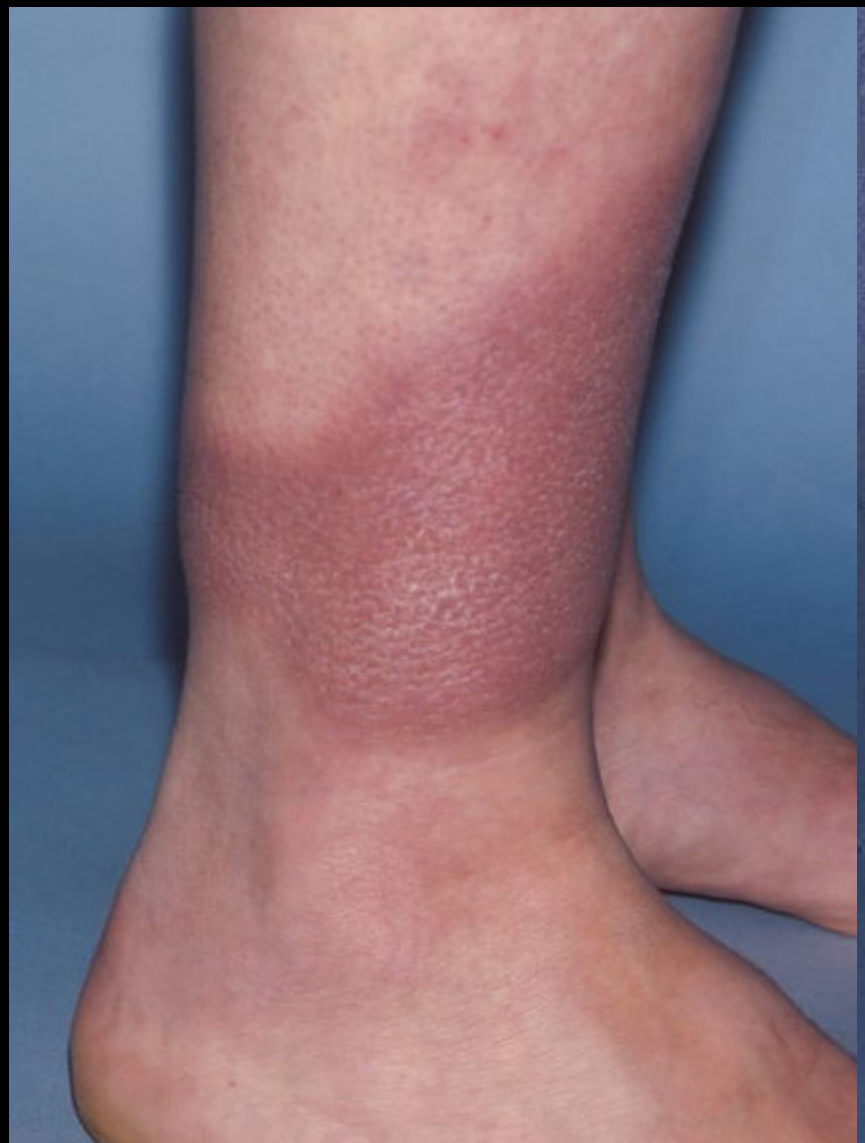
DIABETIC DERMOPATHY

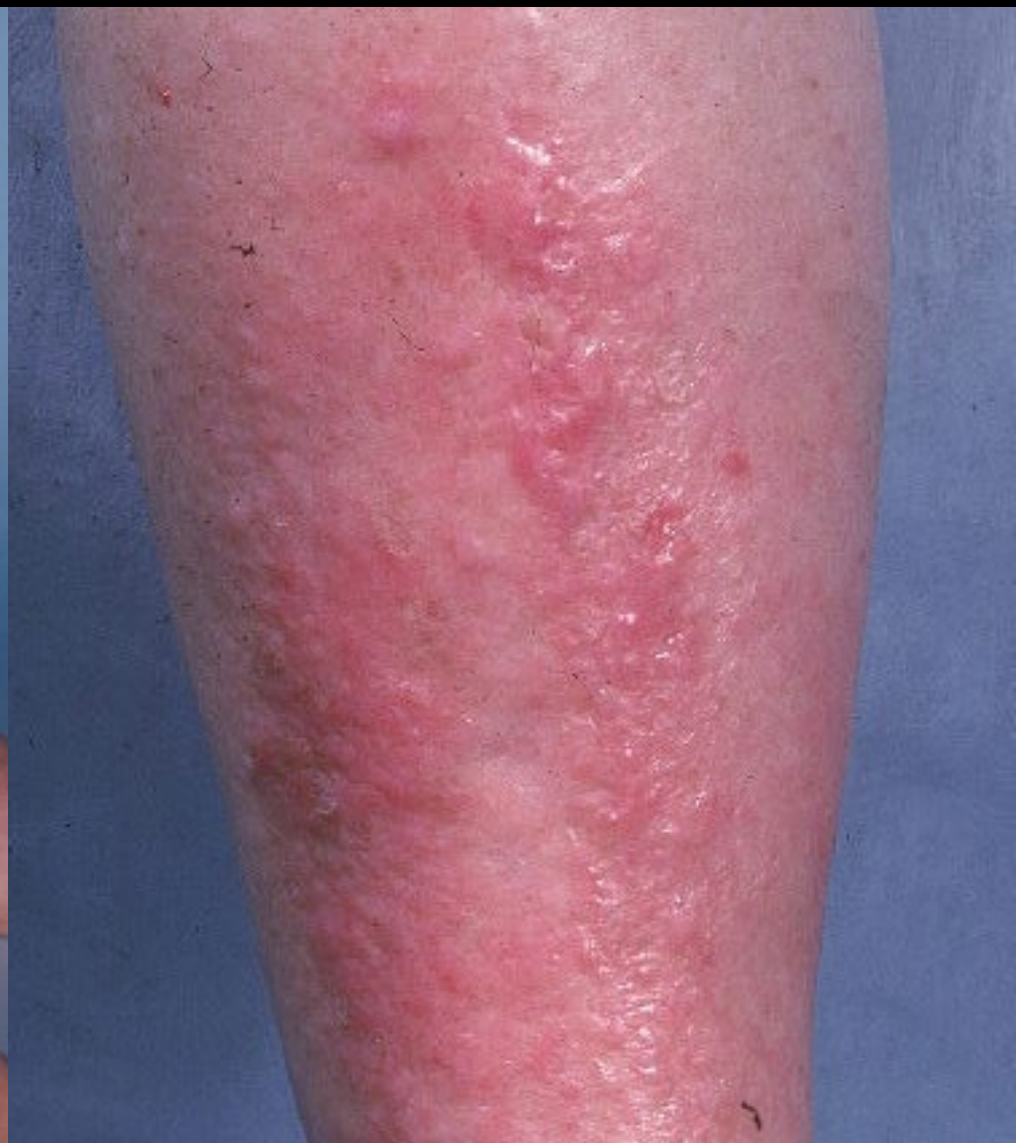




LONGSTANDING DIABETICS
OFTEN WITH OTHER VASCULOPATHY-
ASSOCIATED COMPLICATIONS

PRETIBIAL MYXEDEMA AND GRAVE'S DISEASE

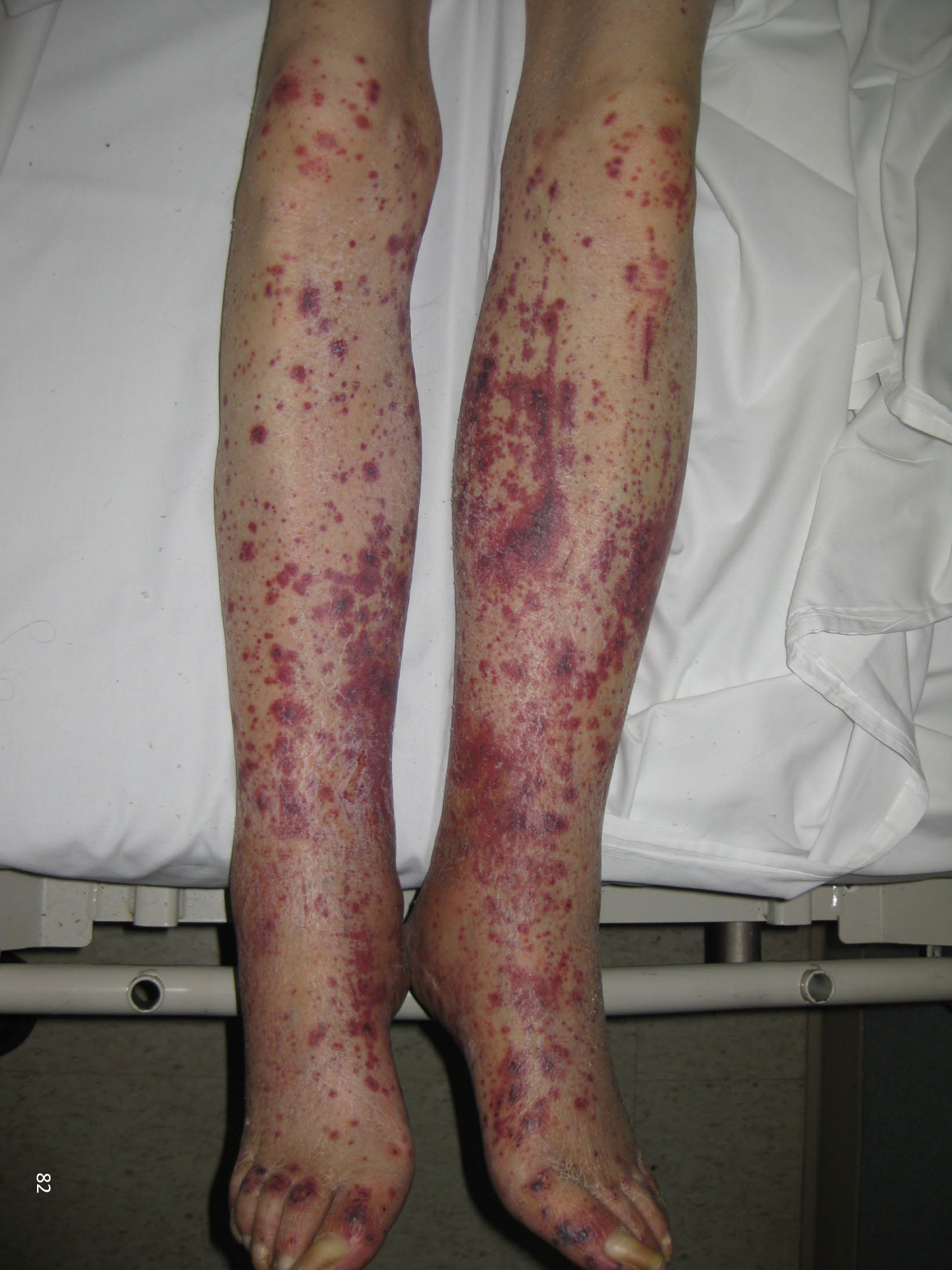


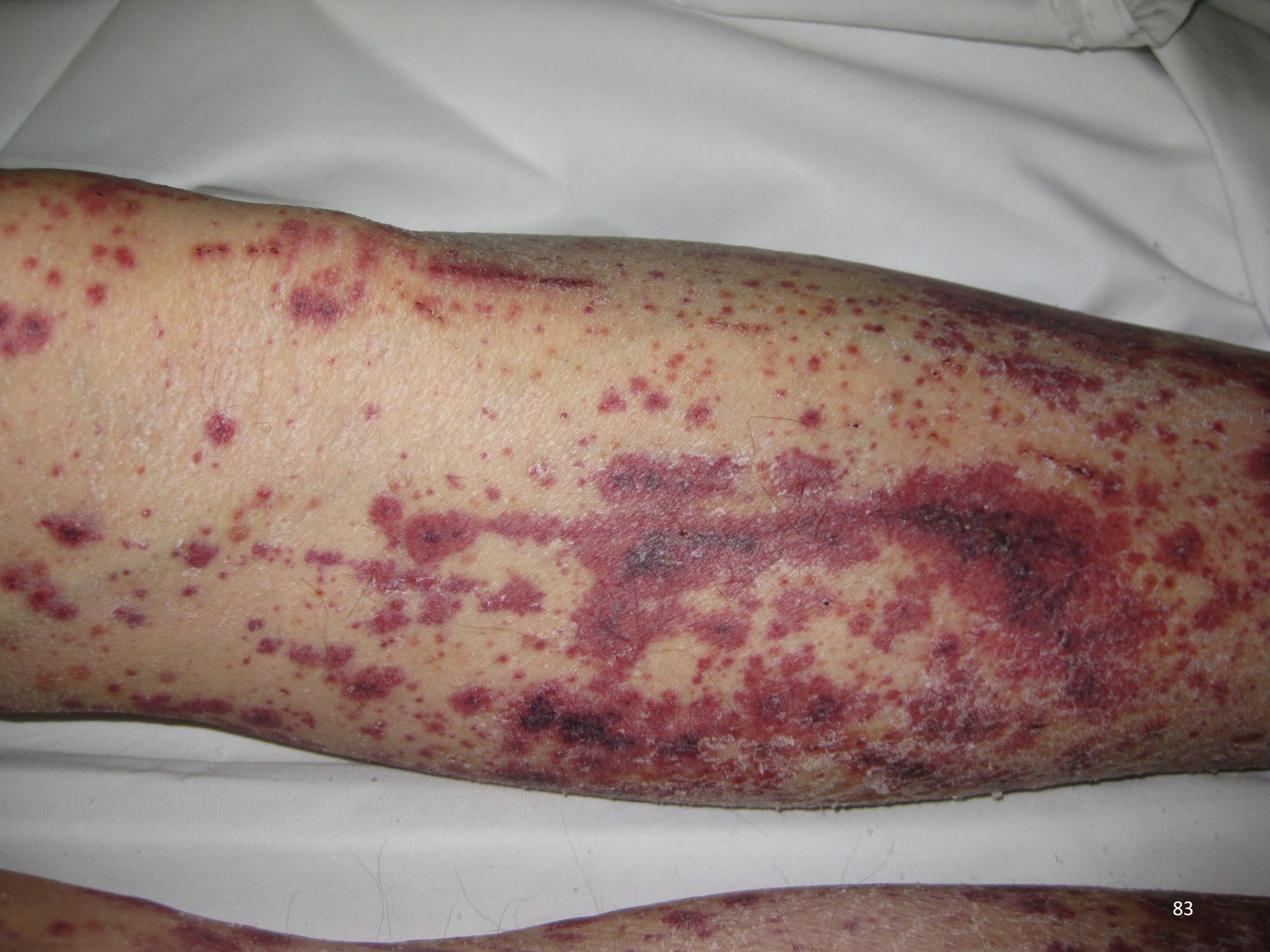


PURPURA …

CASE 11:

- **55 F with positive COVID test**
  - ***Purple papules on legs***





SMALL VESSEL VASCULITIS

- *Aka LEUKOCYTOLASTIC VASCULITIS (LCV)*
  - *palpable purpura favoring dependent areas*
    - » **Pain, swelling, crust, pustules may be present**

SMALL VESSEL VASCULITIS

- **4 Big Categories:**
  1. Idiopathic: 50%
  2. Infectious: 20%
  3. Inflammatory: 20%
  4. Malignancy-associated: 5%

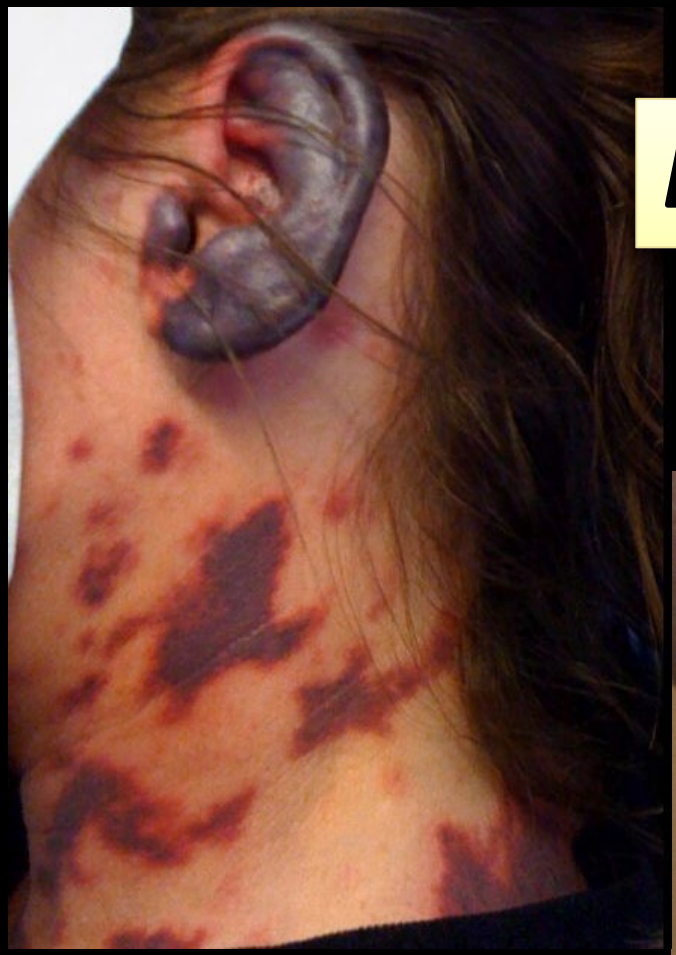
- **Check UA**
  - Glomerulonephritis

V.A.S.C.U.L.I.T.I.S.

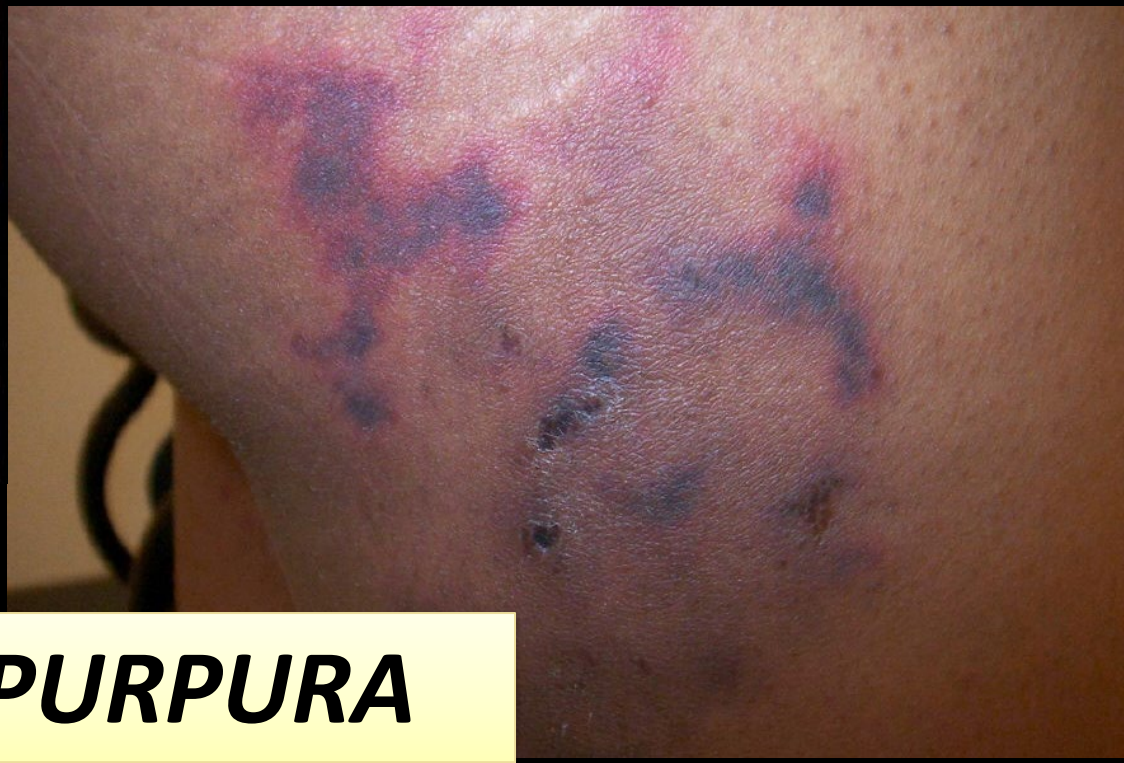
- Various drugs: penicillins, cephalosporins, sulfa, allopurinol
- ANCAs (Wegners, Churgh-Straus, MPA, drug-induced)
- SLE, Sjogren's, RA, IBD
- Cryos – 'MIXED', TYPE II & III (Hep C, Autoimmune, Multiple Myeloma, Waldenstroms)
- Urticarial vasculitis (Hypo vs Normal Complementemic)
- Lymphoma/Leukemia (Hairy Cell Leukemia and PAN)
- Infections (Associative vs. Septic) (Hep B=PAN)
- Tuberculosis ? / TTP?
- Idiopathic, IBD, IgA (HSP)
- Septic vasculitis

CASE 12:

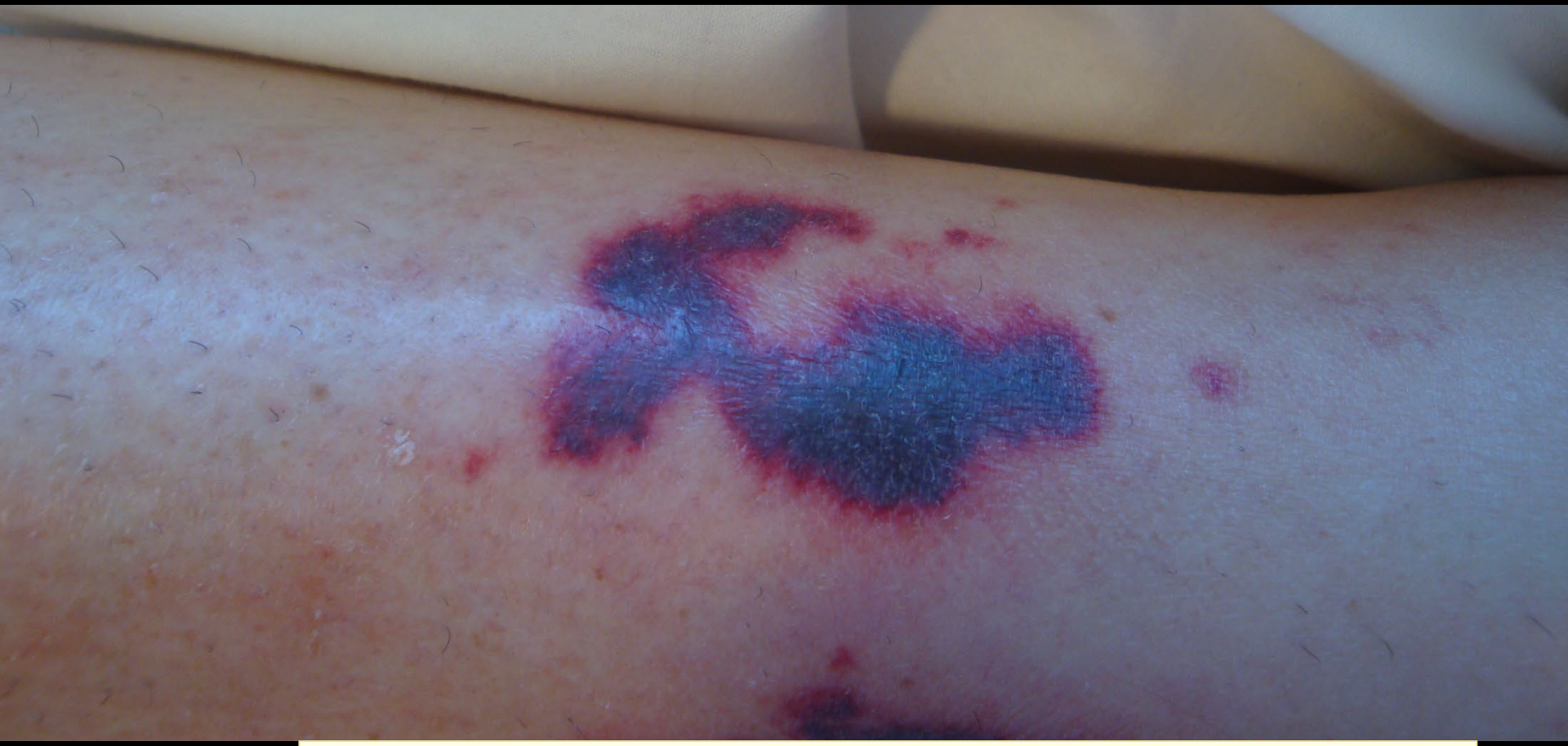
- 45 F with new onset purple plaques presents to the urgent clinic



Labs show + drug screen



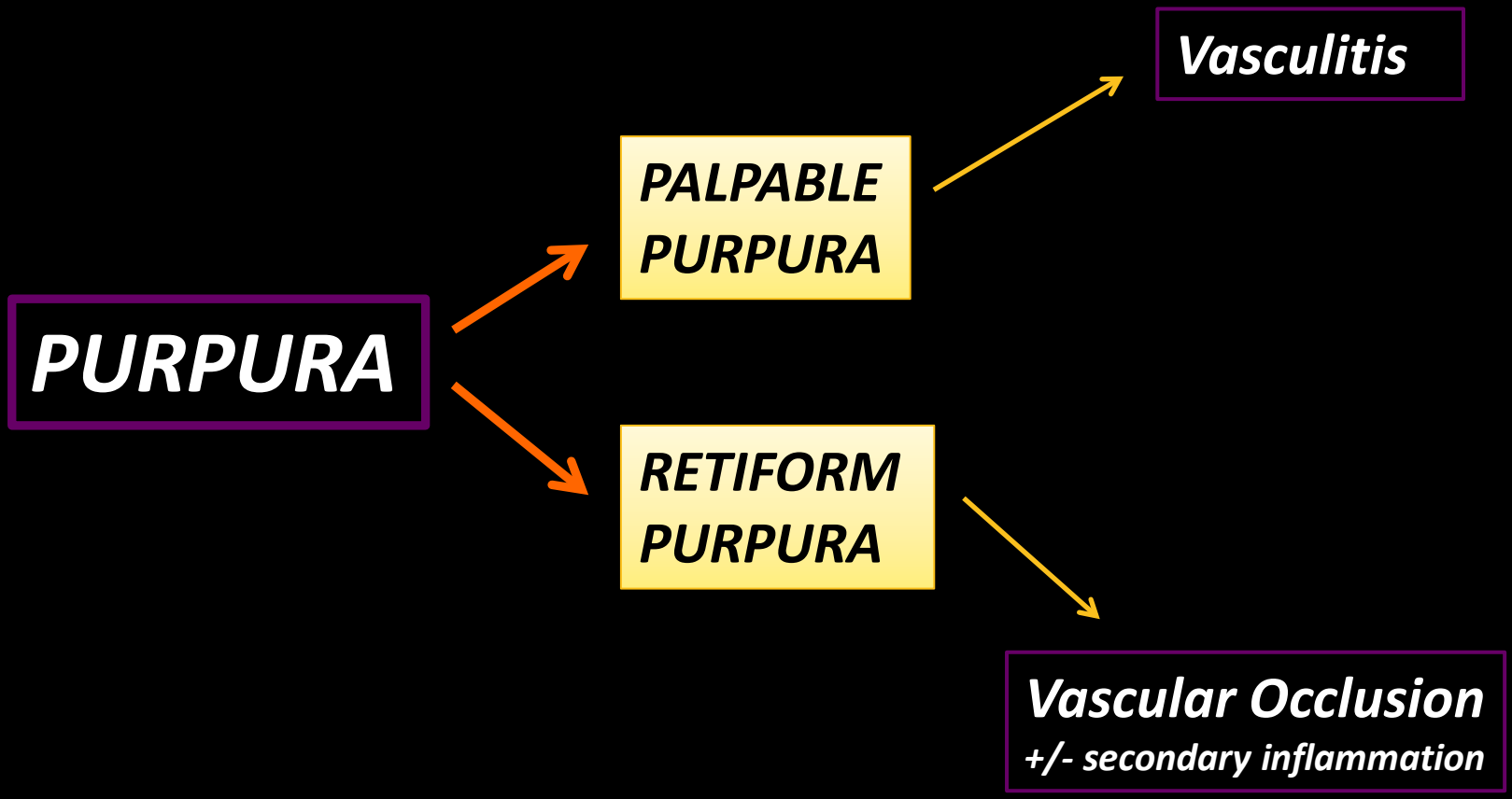
RETIFORM PURPURA

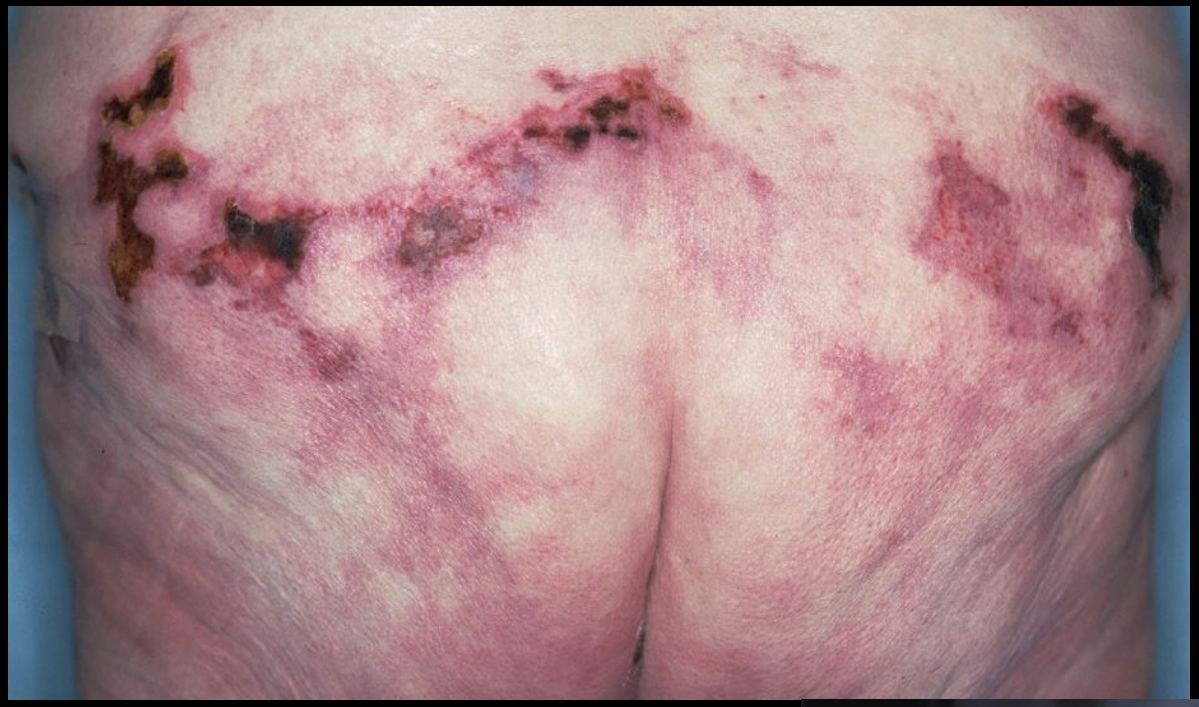


COCAINE LEVAMISOLE TOXICITY

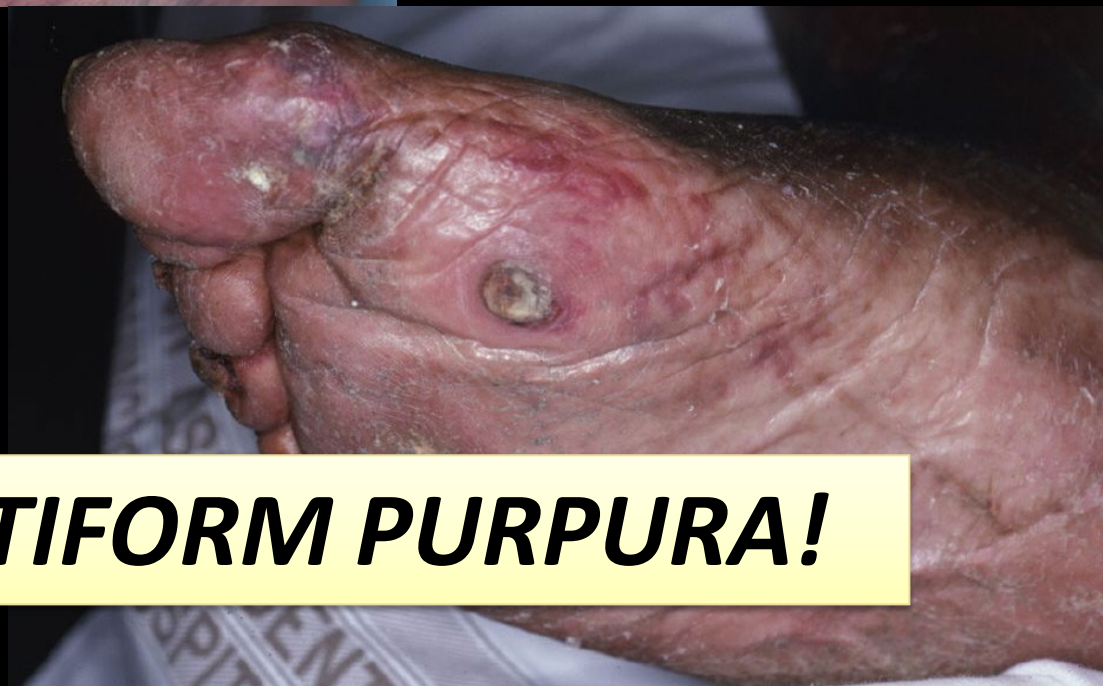
RETIFORM PURPURA

- ***Purpuric macules and patches in net-like, branching or serpentine patterns***
  - **Often with central dusky necrosis**


PURPURA
PALPABLE PURPURA
RETIFORM PURPURA
Vasculitis
Vascular Occlusion
+/- secondary inflammation



CHOLESTEROL EMBOLI



RETIFORM PURPURA!

RETIFORM PURPURA

- *Manifestation of microvascular occlusion of the dermal plexus*

- *Causes (chiefly):*
  - heparin necrosis
  - Coumadin necrosis
  - antiphospholipid antibodies, protein C and/or S deficiency
  - septic emboli
  - cholesterol emboli
  - Type I cryoglobulins --- IgM --- cryogelling occurs
  - calciphylaxis
  - Angioinvasion by organisms (Pseudomonas, Mucor, Aspergillus, Fusarium, Scytalidium)

References

- *Bolognia et al. Dermatology. 4th edition. 2017*
- *Dermnetz*
- *Uptodate*
- *Andrew et al. Diseases of the skin. 12th edition. 2015.*

- ***Big thanks to my numerous patients who gave consent for pictures!***

THANK YOU!