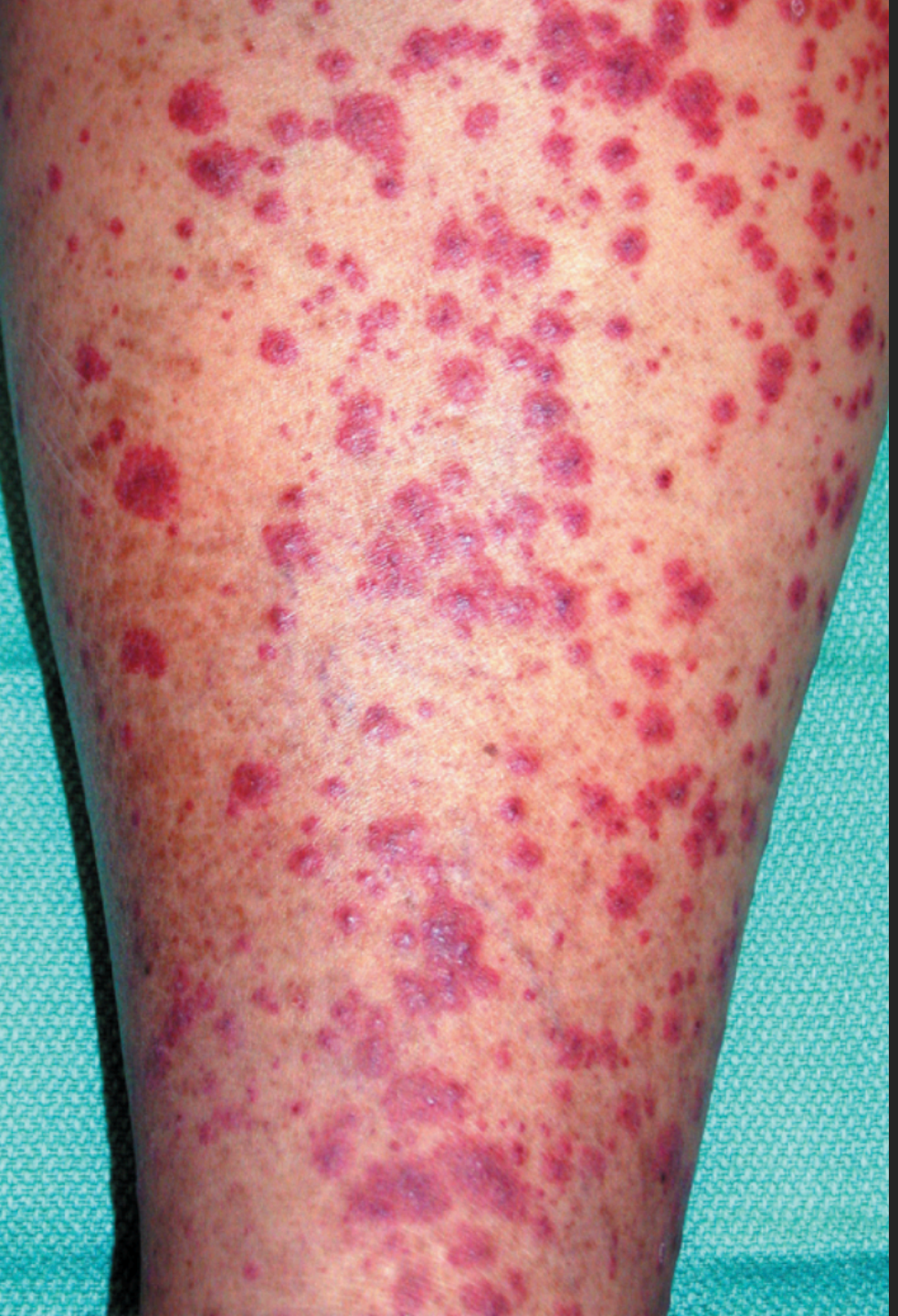

Cutaneous manifestation of Purpura & Vasculitis

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What is purpura?

- Visible hemorrhage into the skin or mucous membrane.
- Mainly divided into 3 categories:

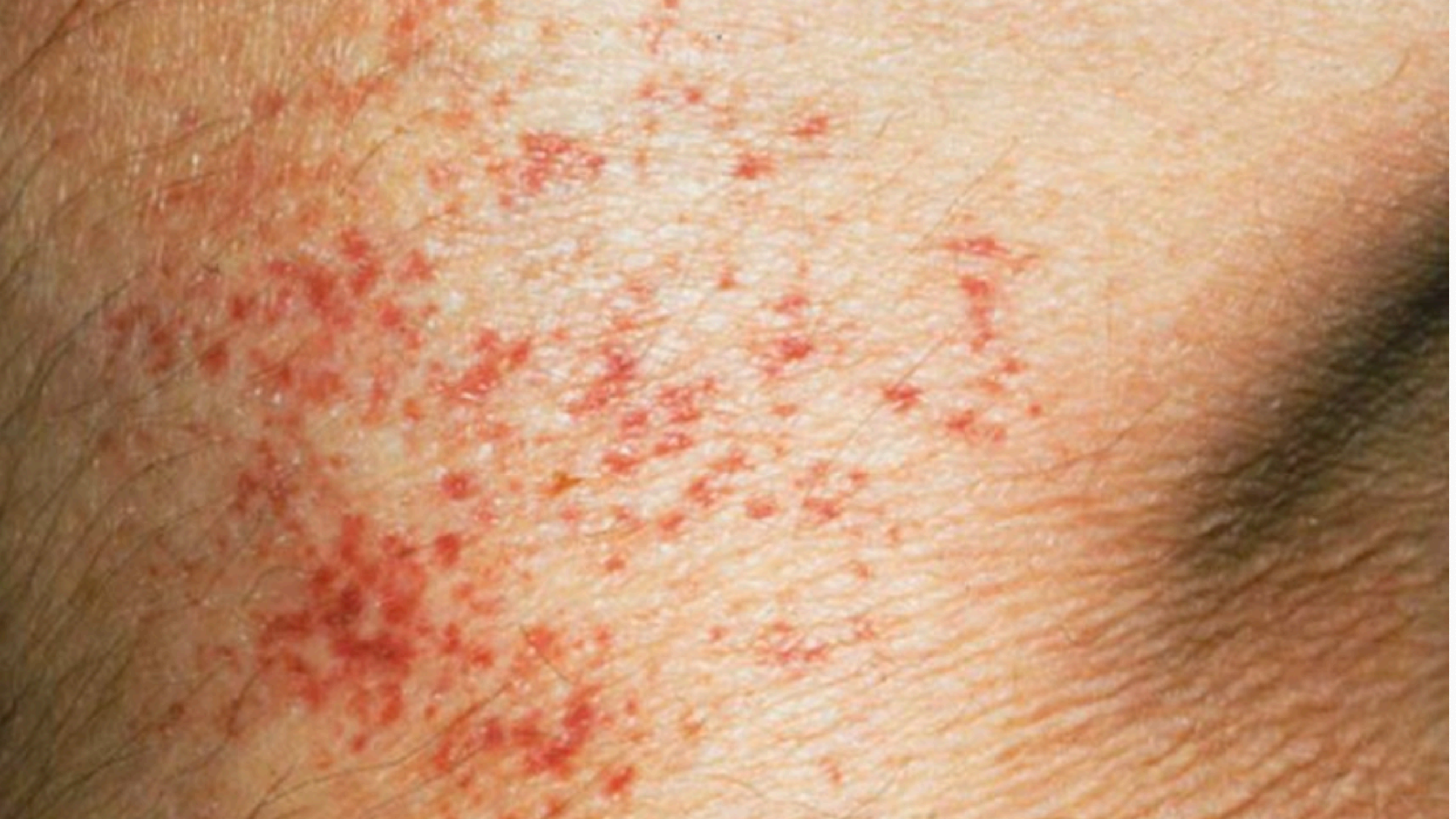
1) Palpable Purpura (Inflammatory)
2) Non-palpable Purpura (non-inflammatory)

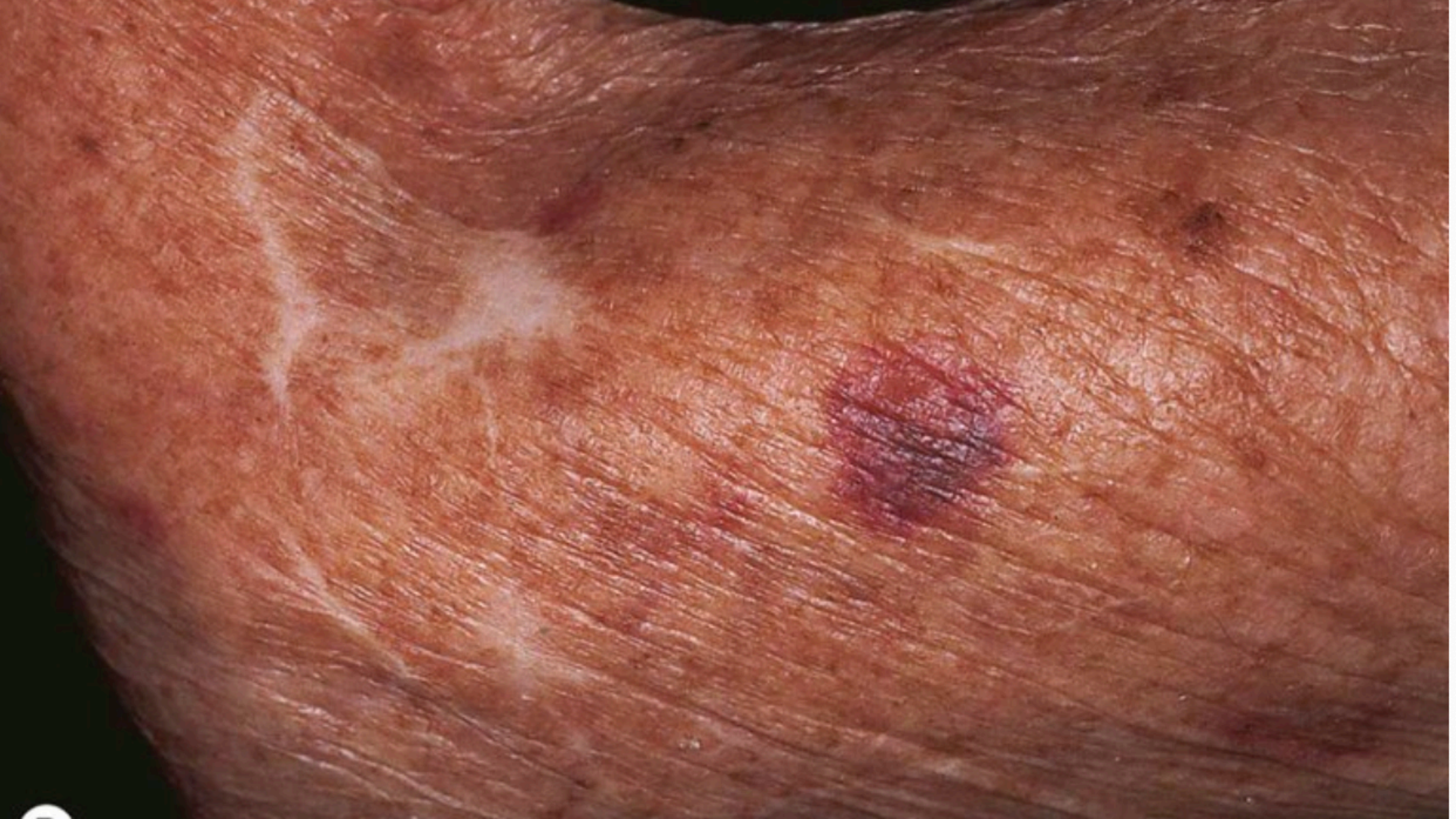
- Macular Petechiae (<4 mm)
- Macular Purpura (5-9 mm)
- Macular Ecchymoses (> 1 cm)

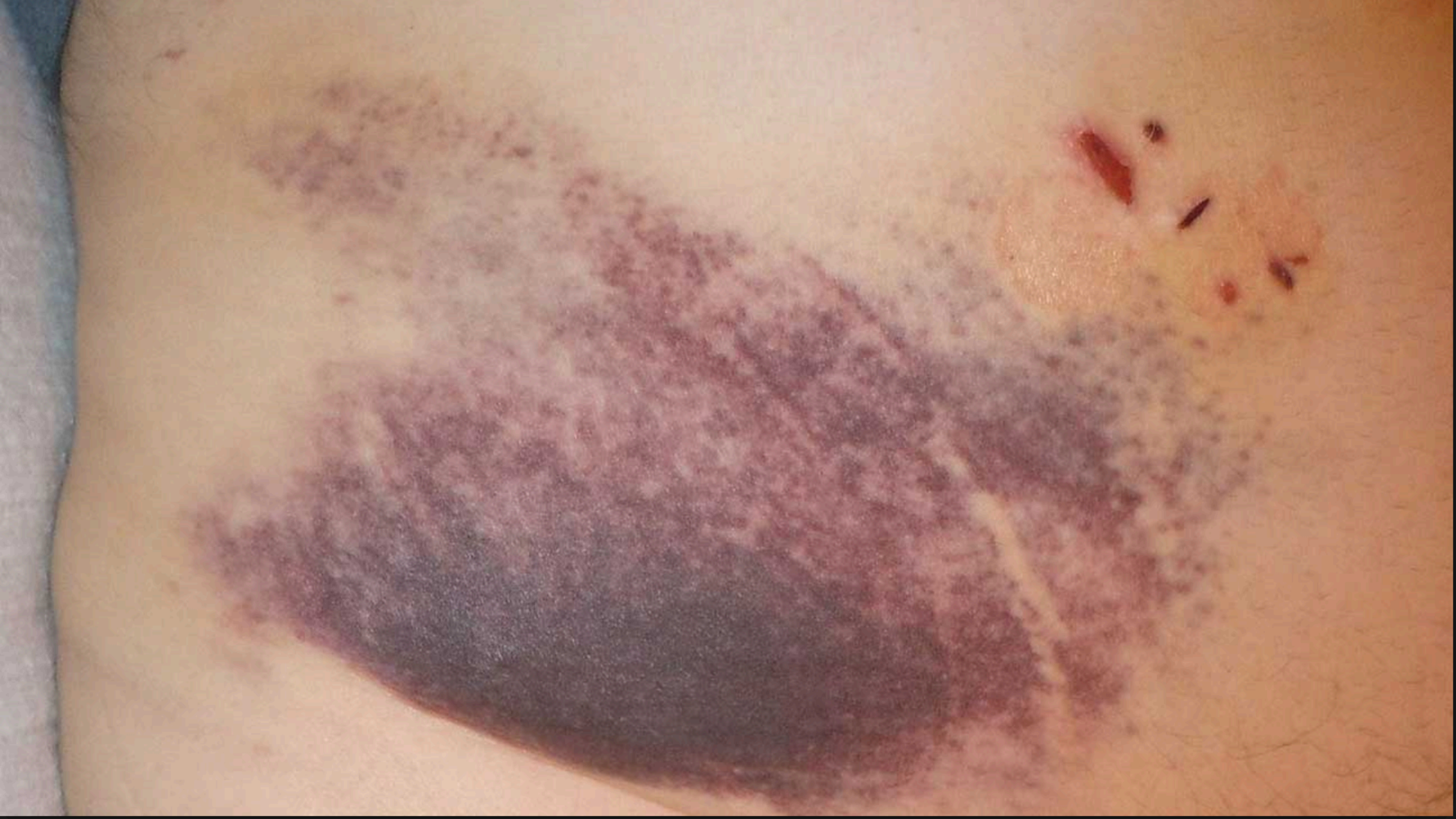
3) Retiform Purpura

Purpura

- **Causes of non-palpable purpura:**
  - Trauma, pigmented purpura, actinic (solar) purpura.
  - Poor dermal support of blood vessels e.g. " topical or systemic steroid use"
  - Vascular dysfunction: aging, scurvy, Ehlers-Danlos syndrome.
  - Platelet dysfunction or Decreased Count: ITP, TTP, drug-induced thrombocytopenia, congenital/acquired platelet function defects
  - Coagulopathies: hemophilia, cryoglobulinemia, anticoagulants, DIC, vitamin K deficiency, hepatic disease.







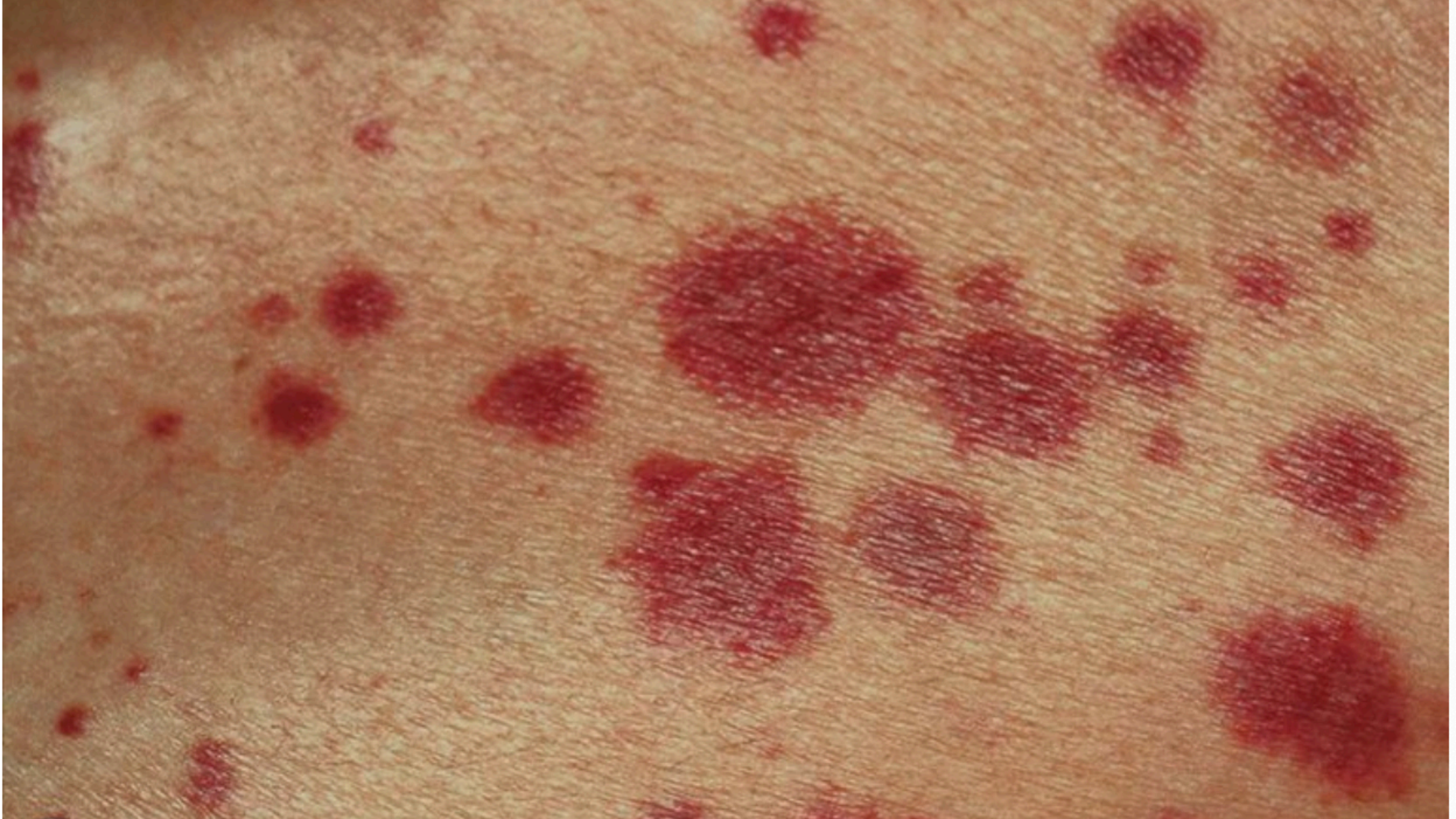
Causes of palpable Purpura

- **<u>Leukocytoclastic Vasculitis:</u>**

- Small Vessels
- Medium Vessels
- ANCA-associated
- Others

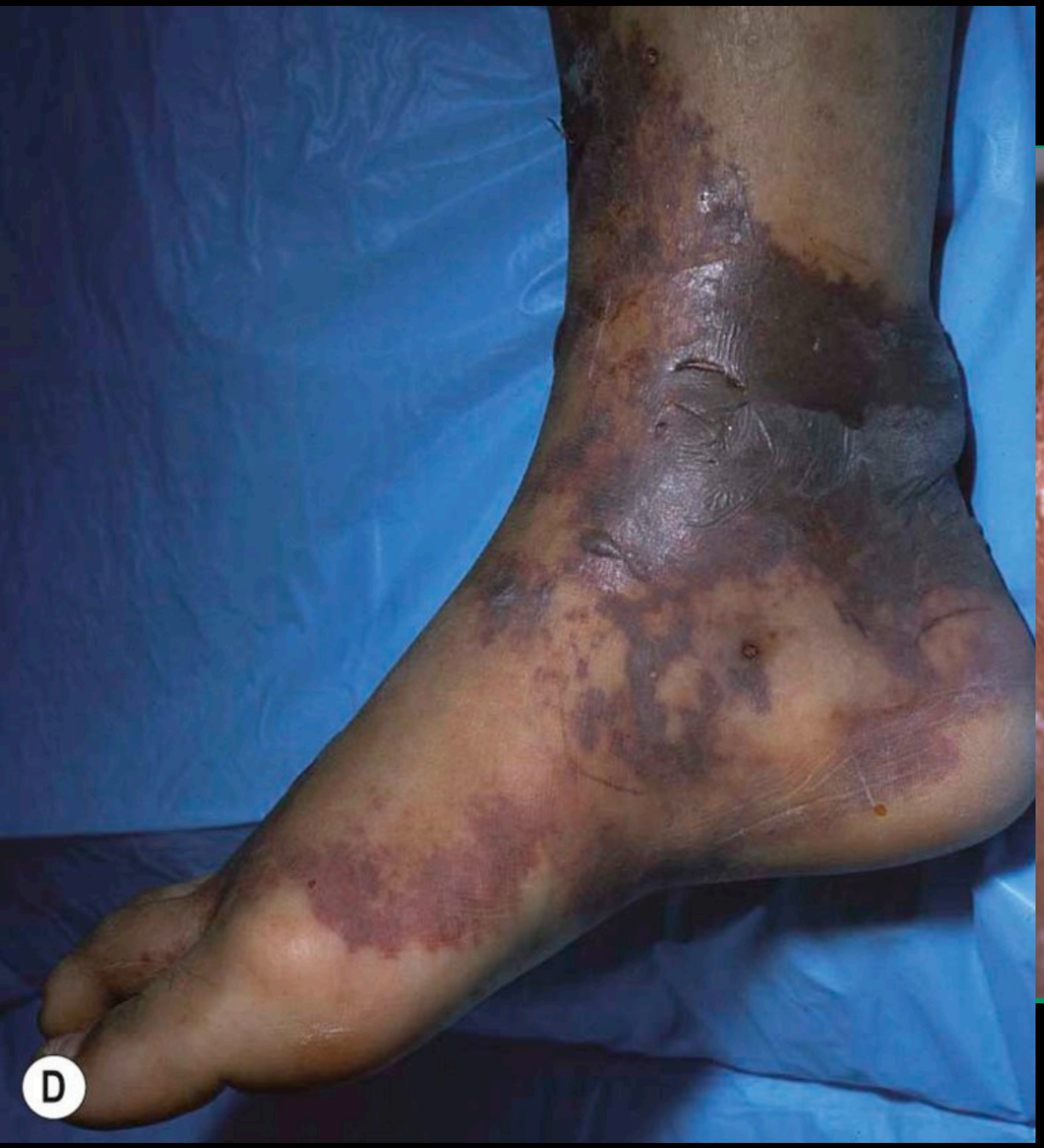
- **<u>Not Leukocytoclastic Vasculitis:</u>**

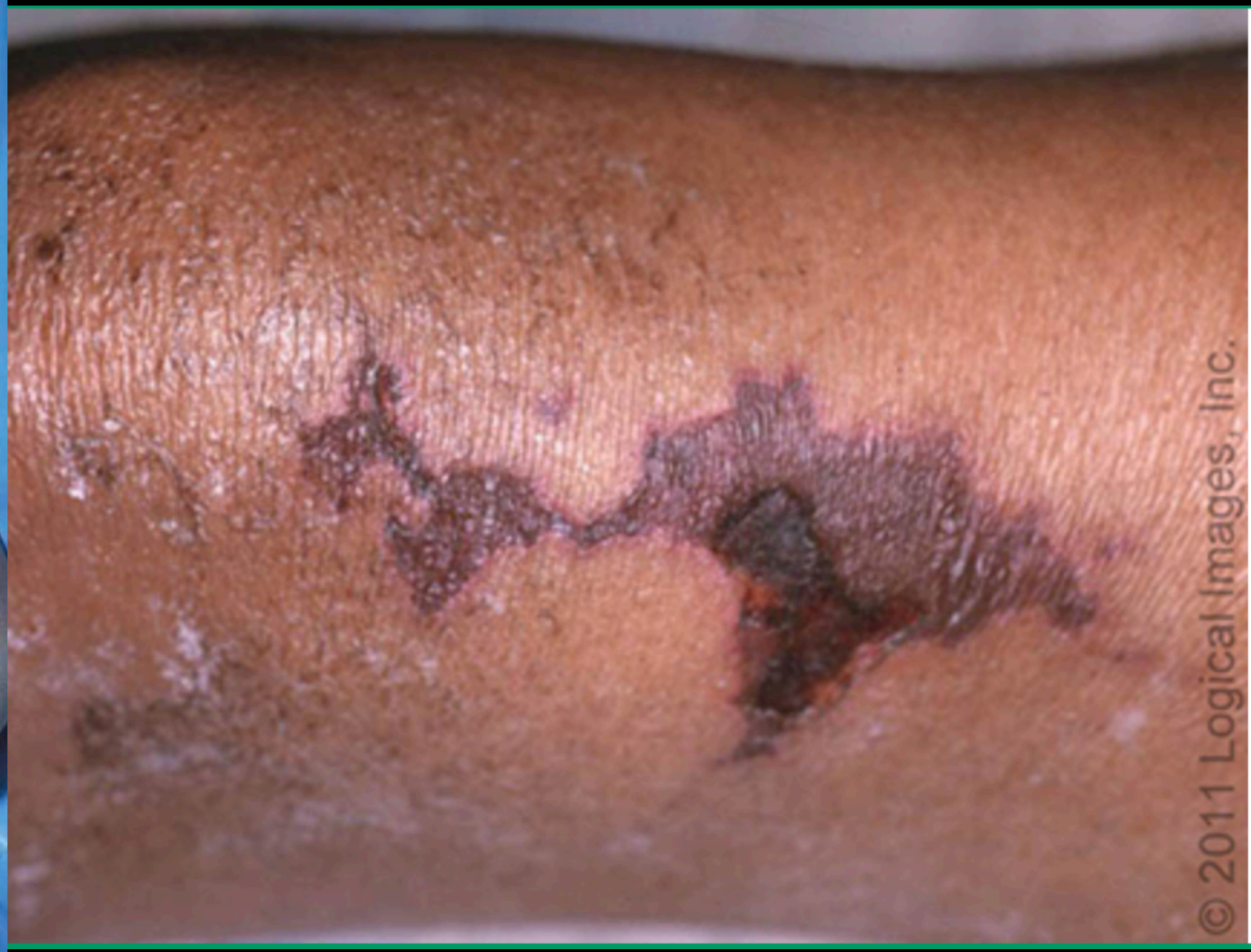
- Erythema Multiforme
- Pityriasis Lichenoides et varioliformis acuta (PLEVA)
- Pigmented purpura



Causes of Retiform Purpura

- Heparin/Warfarin Necrosis
- Cryoglobulinemia
- Invasive Fungi
- Ecthyma Gangrenosum
- Protein C- and S- related
- Livedoid Vasculopathy
- Malignant atrophic papulosis (Degos' disease)
- Sneddon Syndrome.
- Cholesterol Emboli
- Calciphylaxis
- Polyarteritis Nodosa
- Microscopic Polyangiitis
- Wegner's granulomatosis
- Chrugg-Strauss Syndrome



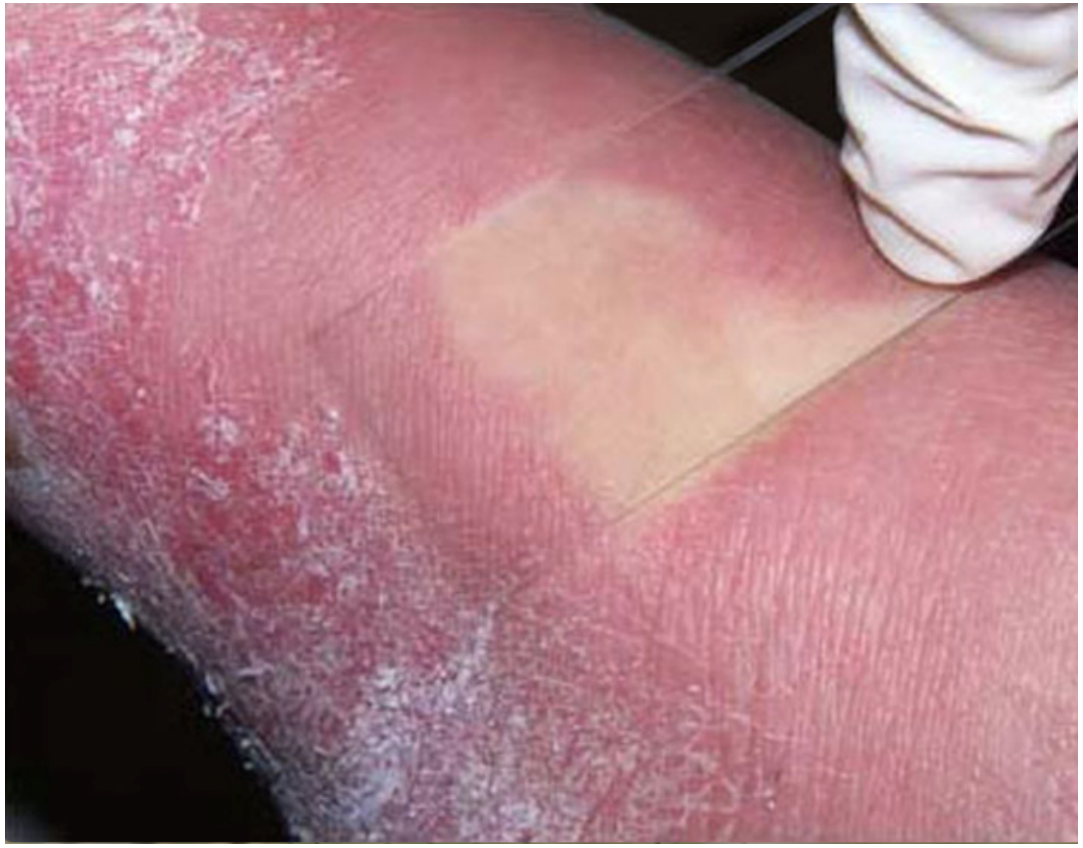


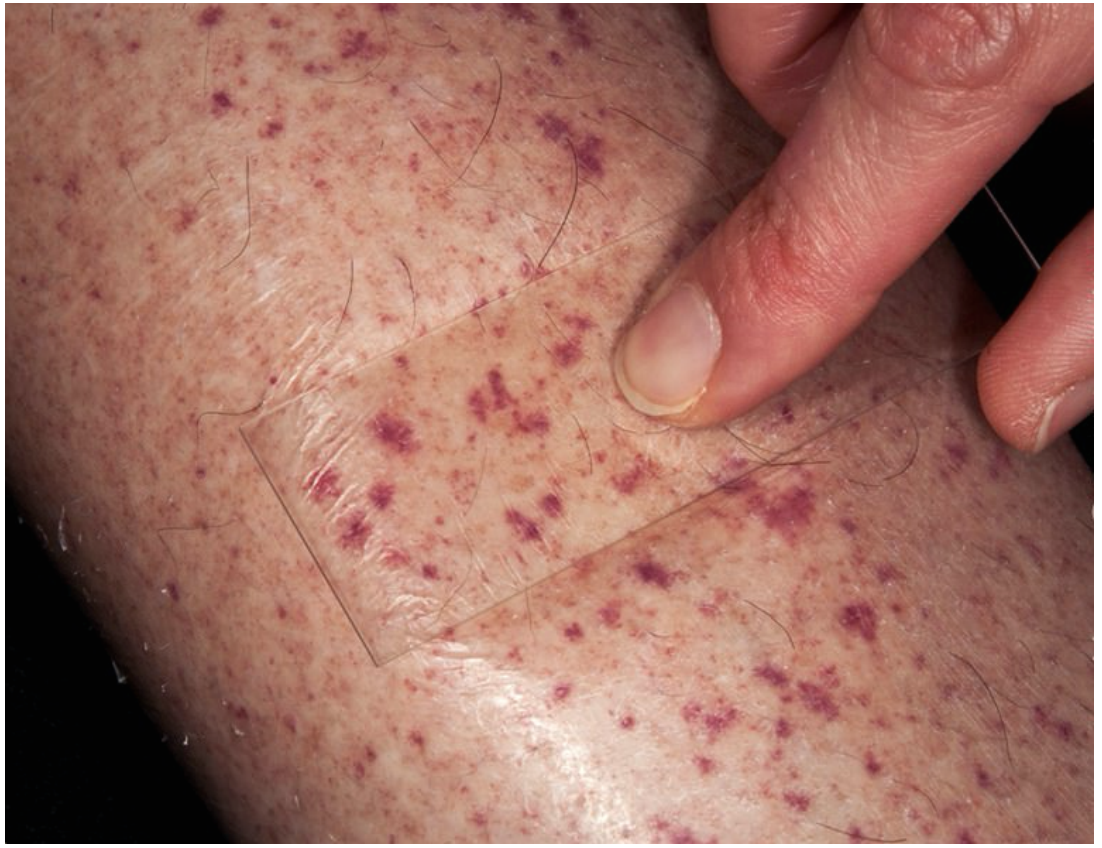
Clinical exam

- purpura does **NOT** blanch with pressure

- **Diascopy: use of a glass slide to apply pressure to the lesion to differentiate erythema secondary to vasodilation (blanchable with pressure), from extravasation of blood (non-blanchable)**

Diascopy test





Evaluation

- History

Family hx, Drug Hx, Medical hx

- Physical examination

Size, Type, Distribution, Mucous membranes.

- Labs

CBC & Differential, Bleeding time, PT & PTT

Vasculitis

Vasculitis

- Vasculitis represents a specific pattern of inflammation of the blood vessel wall.
- It can occur in any organ system of the body.
- Cutaneous vasculitis could be limited to the skin, have secondary systemic involvement or a manifestation of systemic vasculitis.
- Can affect small, medium or large vessels (arterial and venous)
- Cutaneous involvement occurs almost exclusively with vasculitis of small and medium-sized vessels.

Vasculitis classification

- Vasculitis is classified by the vessel size affected (small, medium, mixed or large)
- Clinical morphology correlates with the size of the affected blood vessels
  - Cutaneous small vessels- palpable purpura, urticarial lesions "urticarial vasculitis"
  - Small-medium vessels- subcutaneous nodules, purpura, livedo reticularis, ulceration and necrosis of mainly medium vessel
  - Large vessels- claudication, ulceration and necrosis

Vasculitis

Cutaneous small vessels (Leukocytoclastic vasculitis)

Henoch-Schönlein purpura

Urticarial vasculitis

Other

- idiopathic
- infection- streptococcal, bacterial endocarditis, parvovirus B19, HIV, hepatitis, TB
- drugs- NSAID,sulfonamides, penicillins, barbiturates, propylthiouracil
- malignancy- leukemias, lymphoma, multiple myeloma, renal, lung, prostate,breast

Mixed (small and medium) vessels

ANCA associated vasculitides

- Churg-Stauss syndrome
- Microscopic polyangiitis
- Granulomatosis with polyangiitis (Wegener)

Essential Cryoglobulinemic vasculitis

Medium vessels

Polyarteritis nodosa- Cutaneous & systemic

Large vessels

Giant-cell arteritis

Takayasu arteritis

Cutaneous small vessel vasculitis
‘’Leukocytoclastic vasculitis’’

Cutaneous small vessel vasculitis "Leukocytoclastic vasculitis"

- Primarily involves the dermal postcapillary venules.
- Could occur as a **primary** process or could be **secondary** to an underlying cause.
- **Palpable purpura** is the hallmark of this disease.
- pinpoint to- several mm in diameter.
- They predominate on the ankles and lower legs, affecting mainly dependent areas.
- Usually asymptomatic and lesions resolve with residual post-inflammatory hyperpigmentation.
- Constitutional symptoms (Fever, weight loss and myalgias) may accompany flares of CSVV including systemic symptoms. (rule out systemic vasculitis)
- 90% of patients will have spontaneous resolution of lesions within weeks, months.

Cutaneous small vessel vasculitis "Leukocytoclastic vasculitis"

Work-up:

History and physical examination .

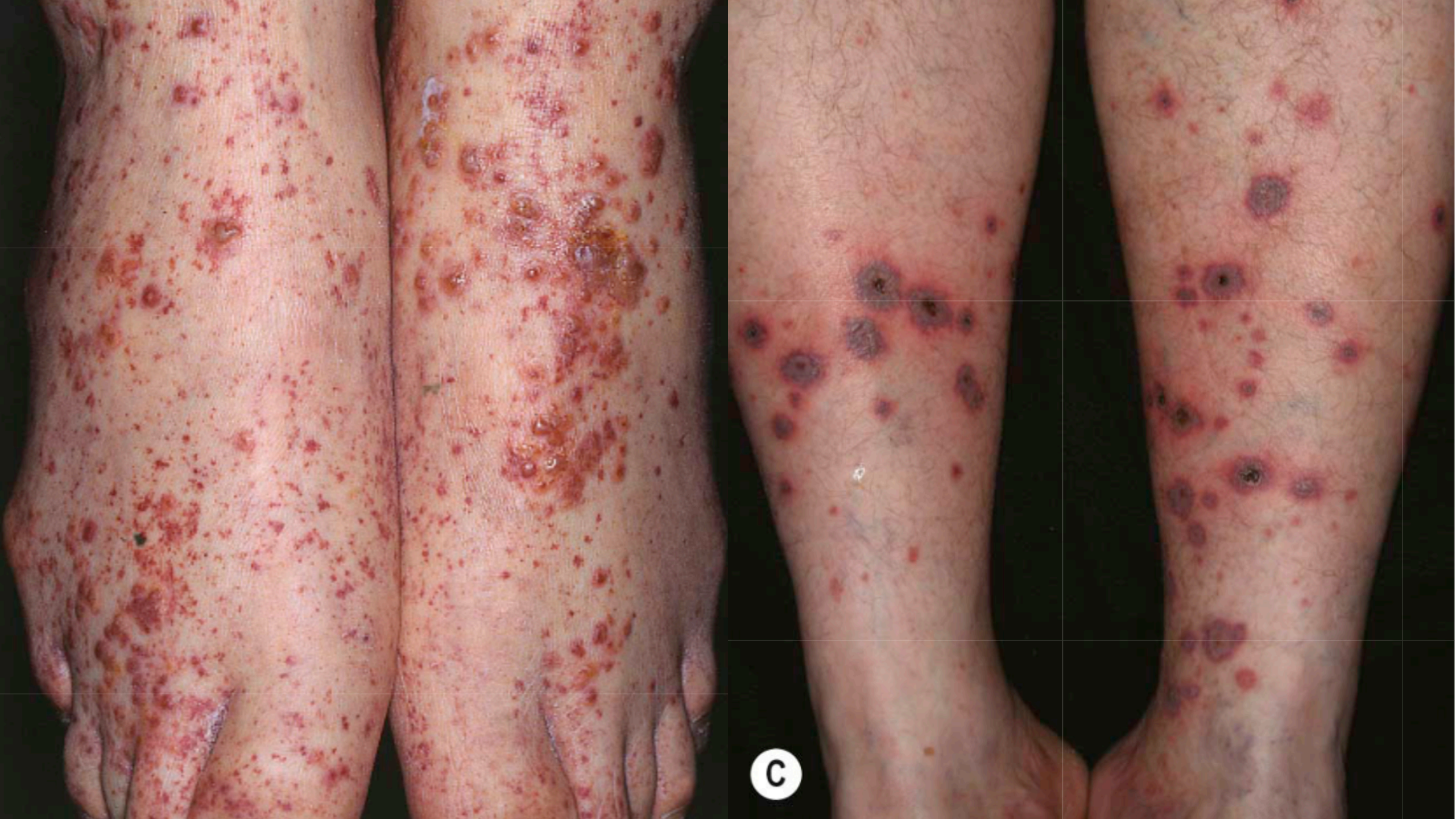
Skin biopsy (+/- DIF)

Labs: CBC, Renal profile, ANA, complement, ANCA.

Treatment

Supportive mainly

If severe, systemic steroids taper.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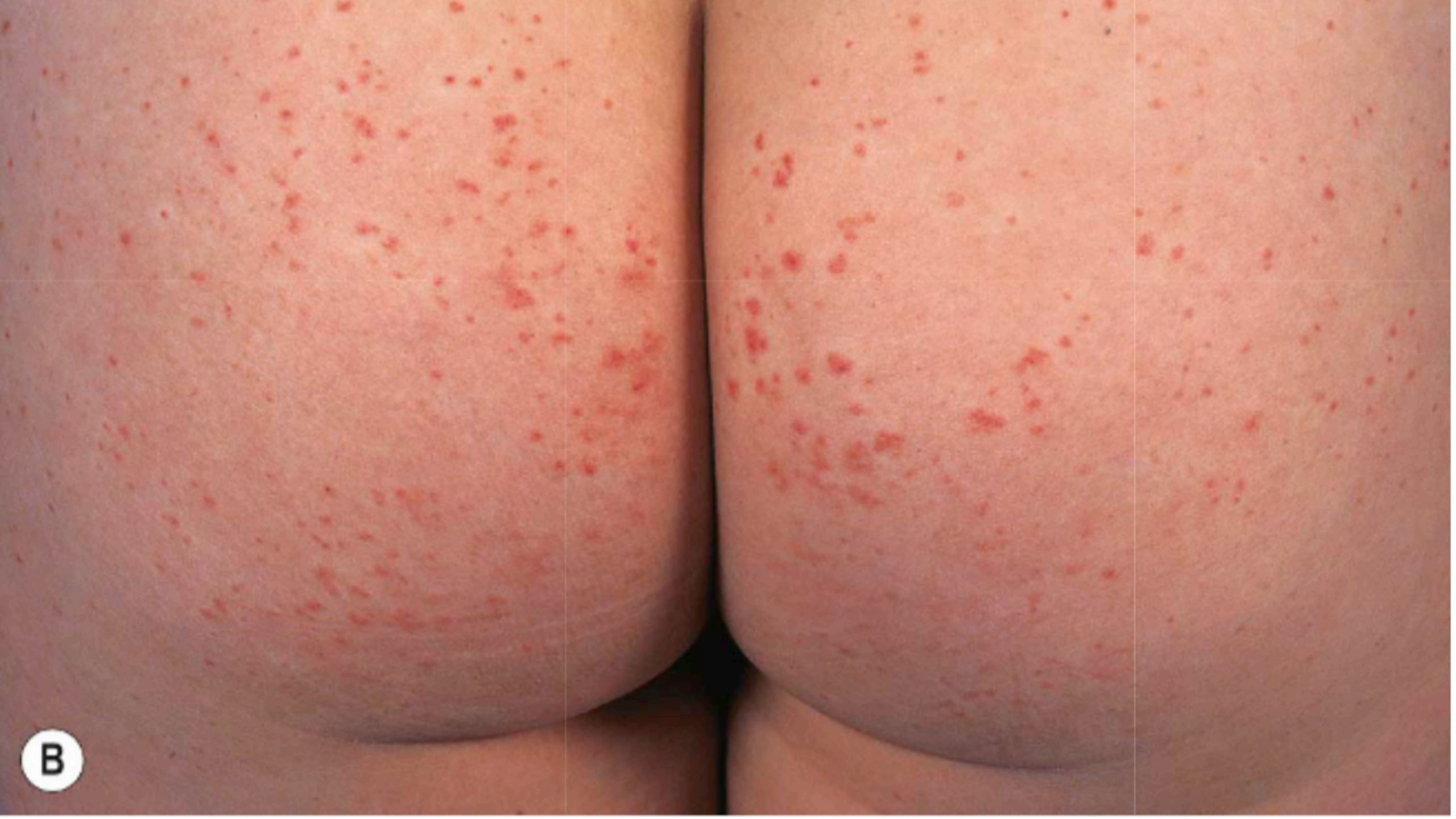
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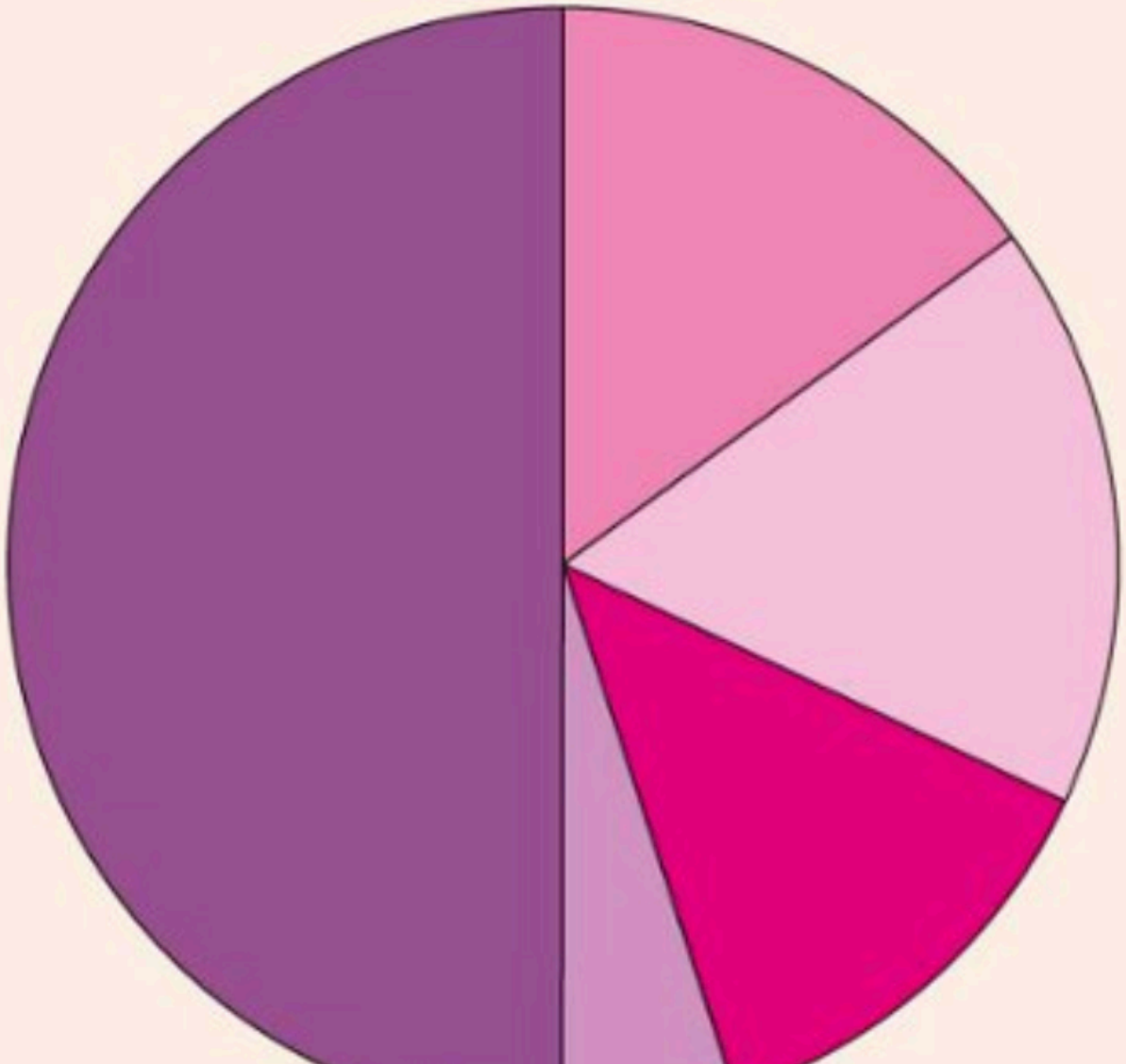
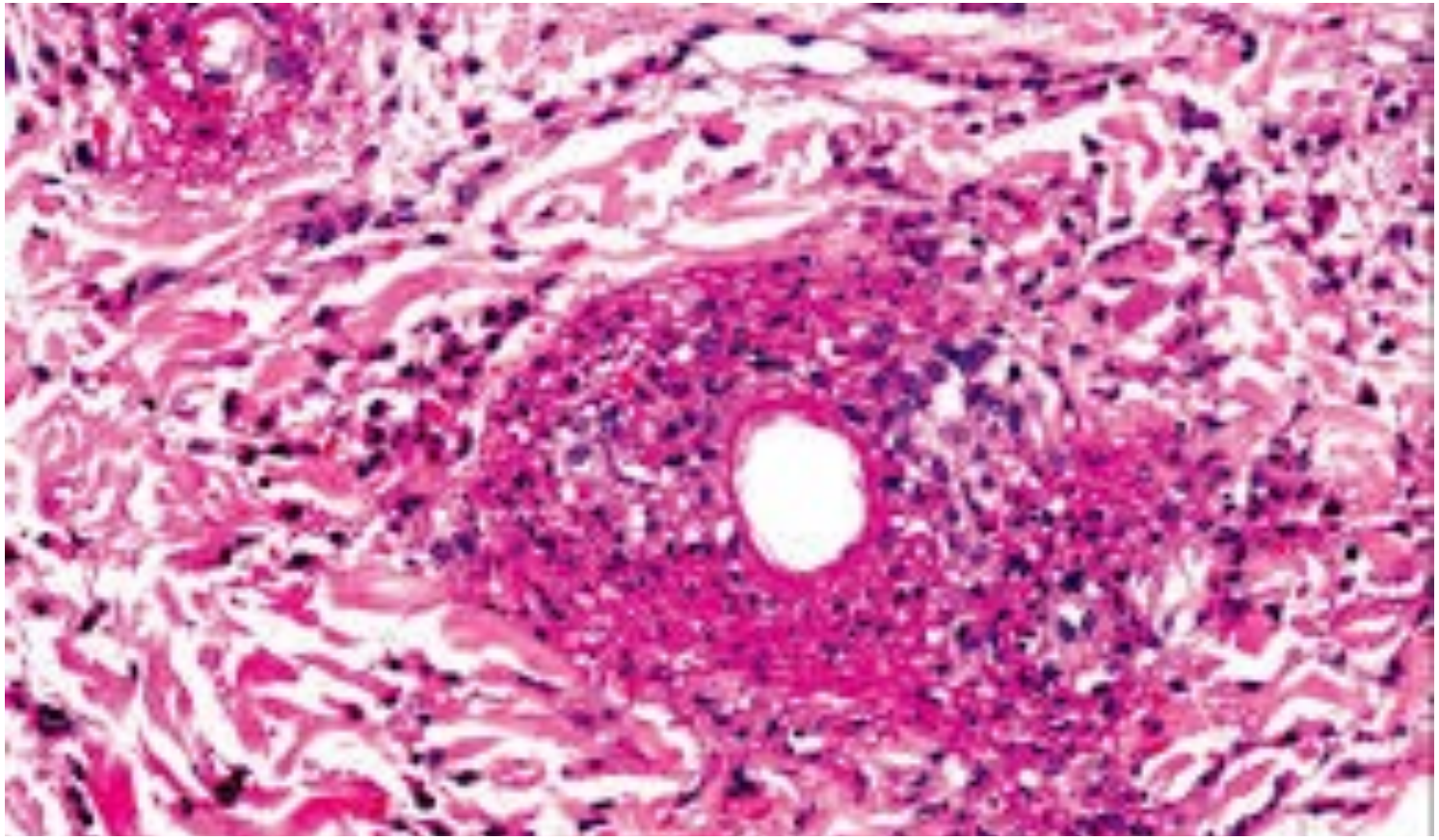
ETIOLOGIES OF CUTANEOUS SMALL VESSEL VASCULITIS



- Infection (15–20%)
- Autoimmune connective tissue disease (15–20%)
- Drug (10–15%)
- Neoplasm (5%)
- Idiopathic (45–55%)



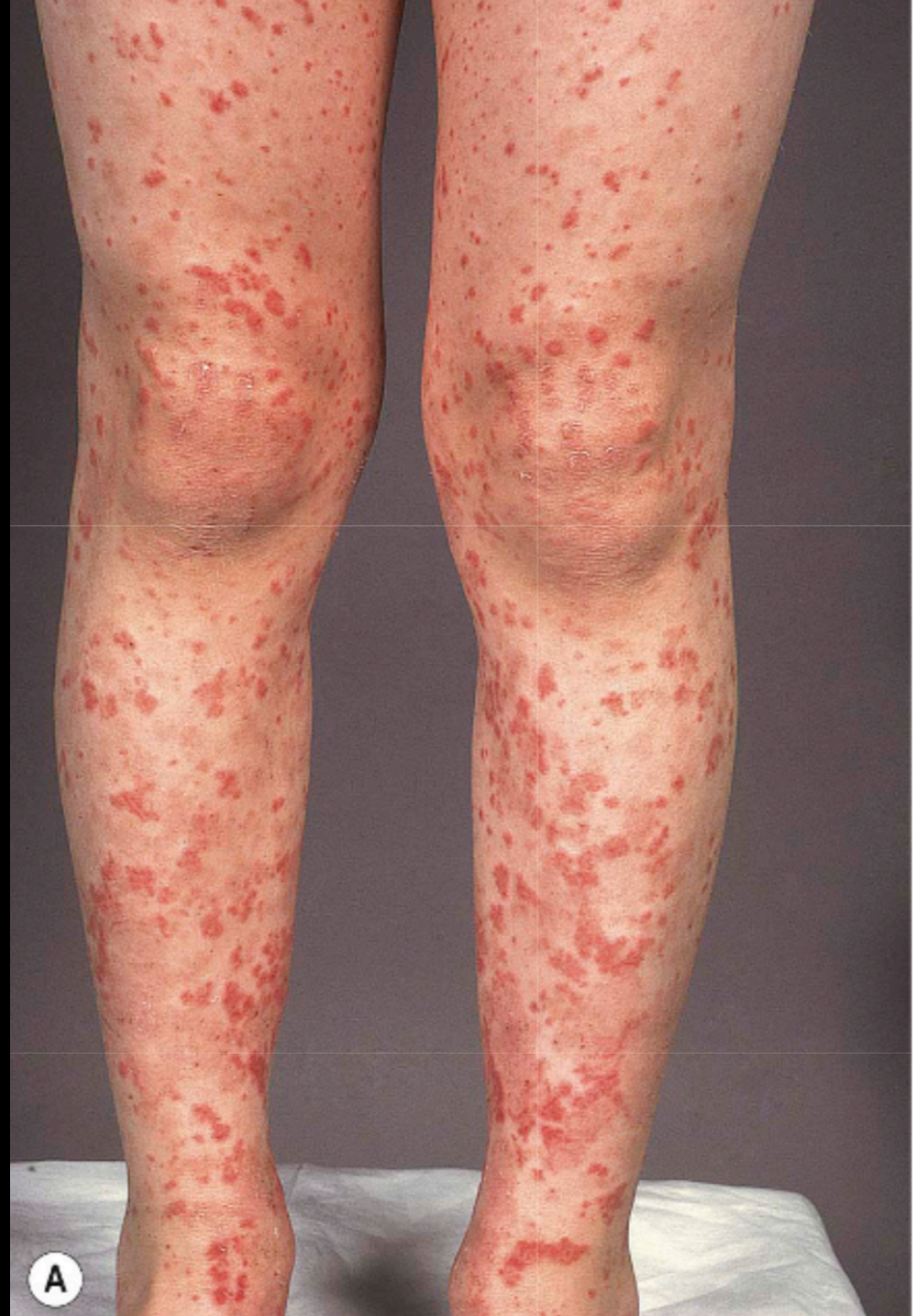
Cutaneous small vessel vasculitis “Leukocytoclastic vasculitis”

Histopathology:

- Inflammation in the form of neutrophils (nuclear dust)
- Vessel wall thickening
- Erythrocyte extravasation
- Fibrin deposit within the vessel wall.

Henoch-Schönlein Purpura

- HSP is a specific form of CSVV.
- Most commonly occurs in children <10 yrs , can be seen in adults.
- Associated with a preceding respiratory tract infection.
- Intermittent palpable purpura on extensor extremities and buttocks.
- **IgA-dominant** immune deposits in walls of small blood vessels.
- Arthralgias and arthritis (up to 75%)
- GI involvement (50-75%): Abdominal pain and/or melena.
- Renal involvement (40-50%): Renal vasculitis often mild but can be chronic.
- May be associated with an underlying malignancy in adults.
- Treatment: supportive.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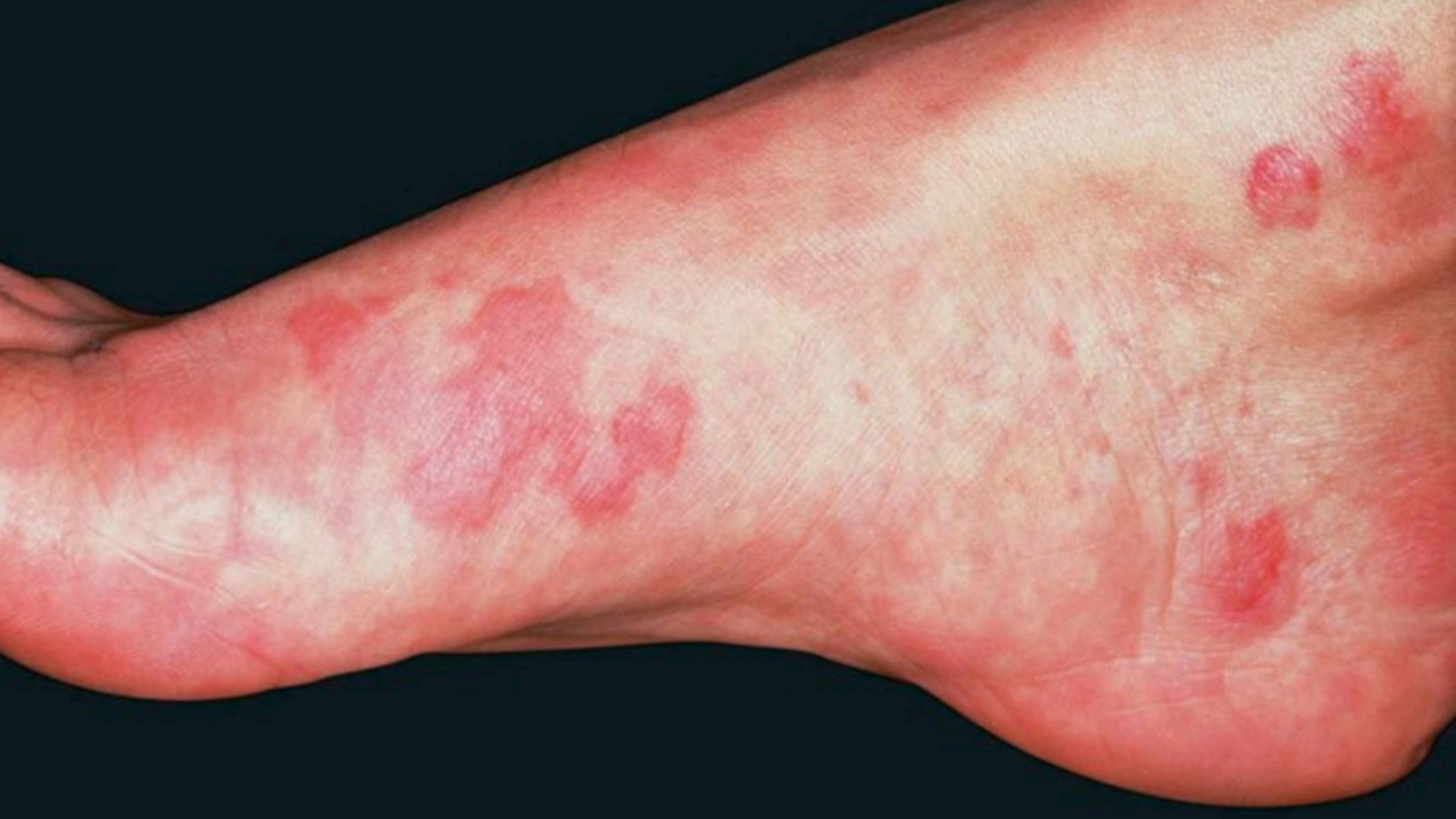
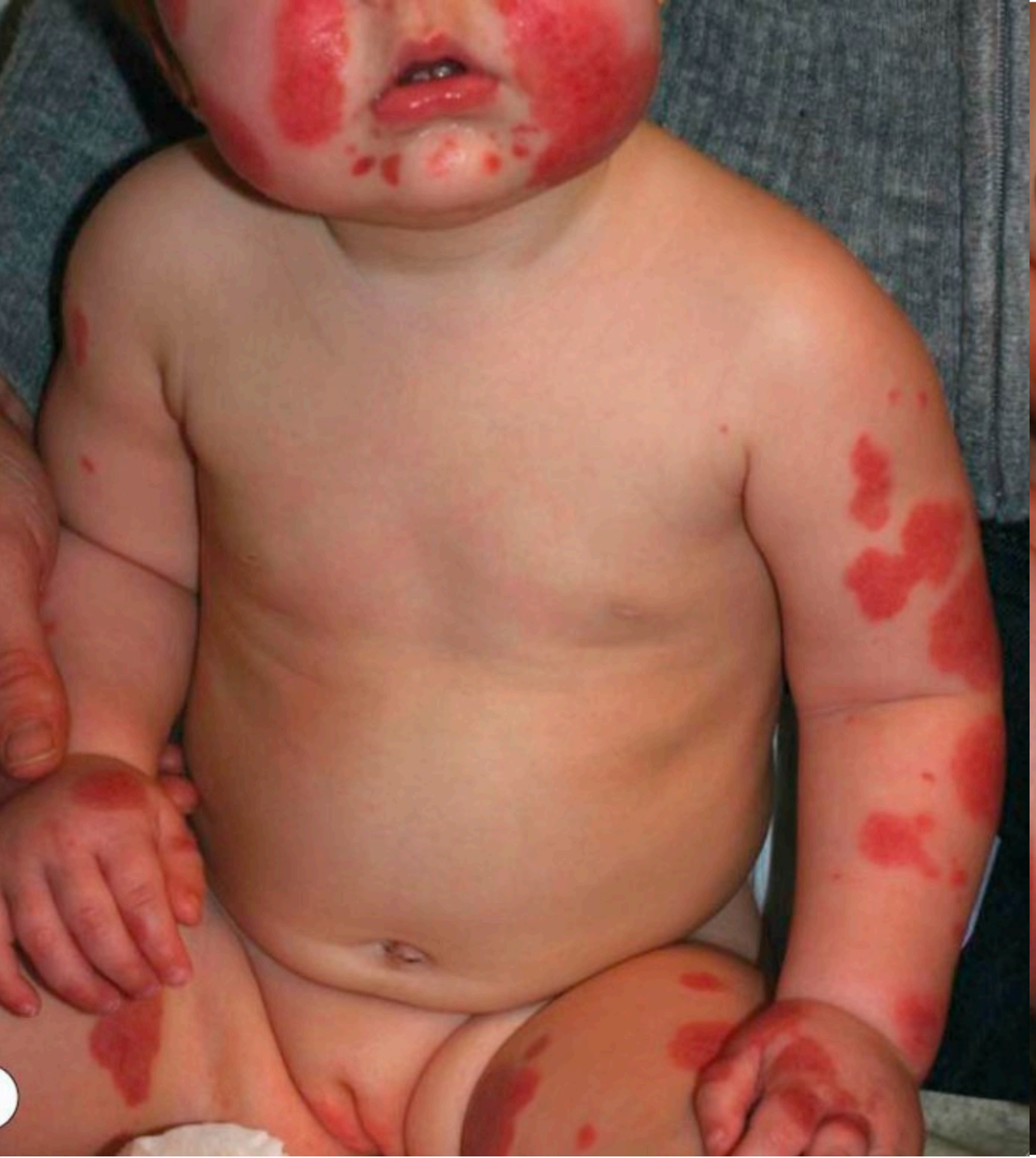
Urticarial Vasculitis

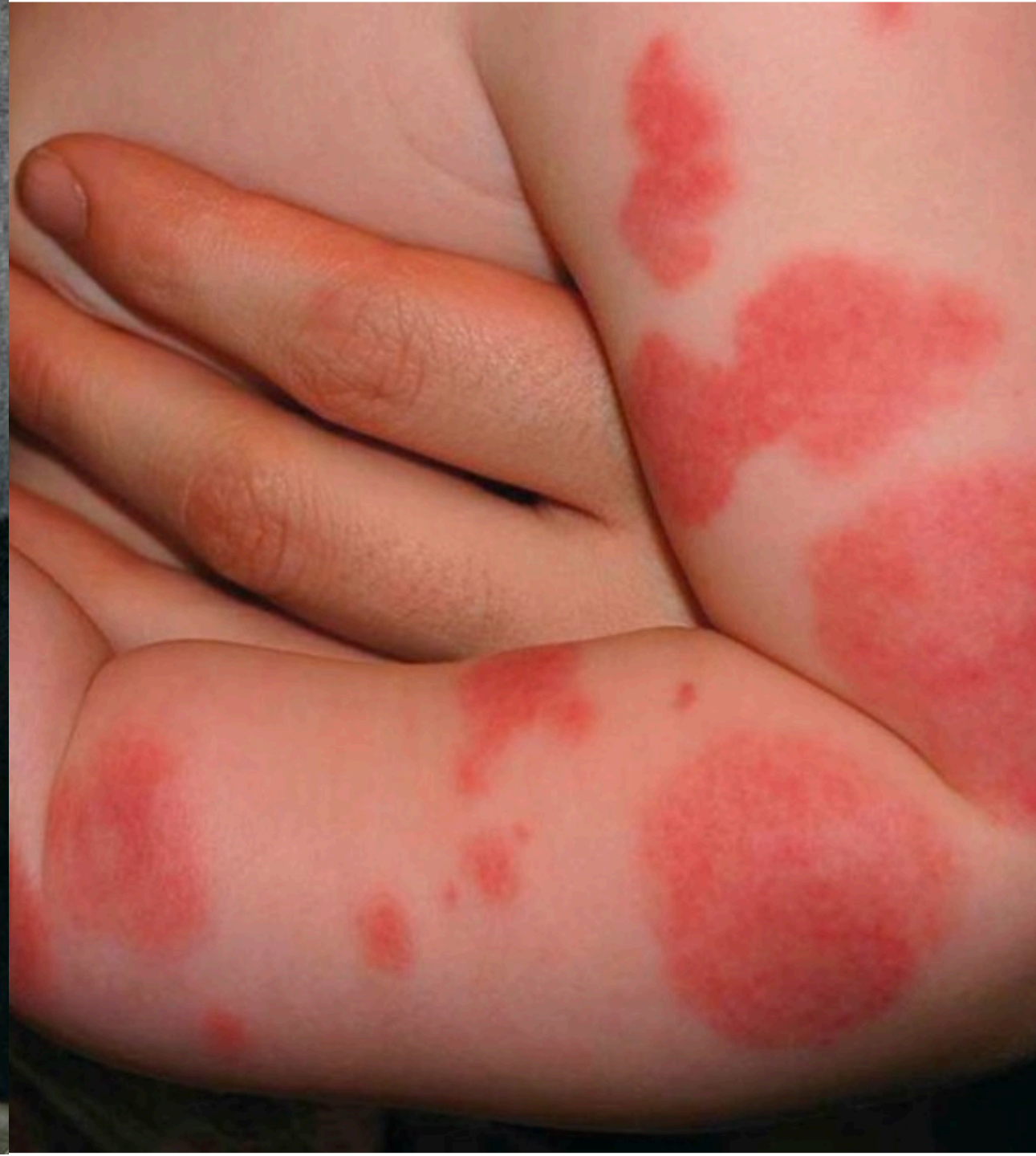
- Recurrent episodes of painful, persistent urticarial lesions that **last > 24 hours** and often resolve with residual hyperpigmentation.
- May occur with or without angioedema.
- May be associated with constitutional symptoms and arthritis.
- Patients with hypocomplementemia are more likely to have systemic involvement (MSK involvement is the most common extracutaneous manifestation).
- Associated disorders include: autoimmune CTD's (Lupus, Sjogren's) and viral infections.
- Labs: elevated ESR, decreased Serum C3, C4 and a positive ANA.



Acute Hemorrhagic edema of infancy

- A very rare form of CSVV.
- The child is well-appearing.
- Seen primarily in children between 4 and 24 months of age.
- Annular, circular or targetoid purpuric plaques on the face and extremities.
- Tender, non-pitting edema of acral sites.
- Extracutaneous involvement is rare.
- Benign clinical course with spontaneous resolution within 1-3 weeks.





Small to medium vessel vasculitis

ANCA-associated Vasculitides

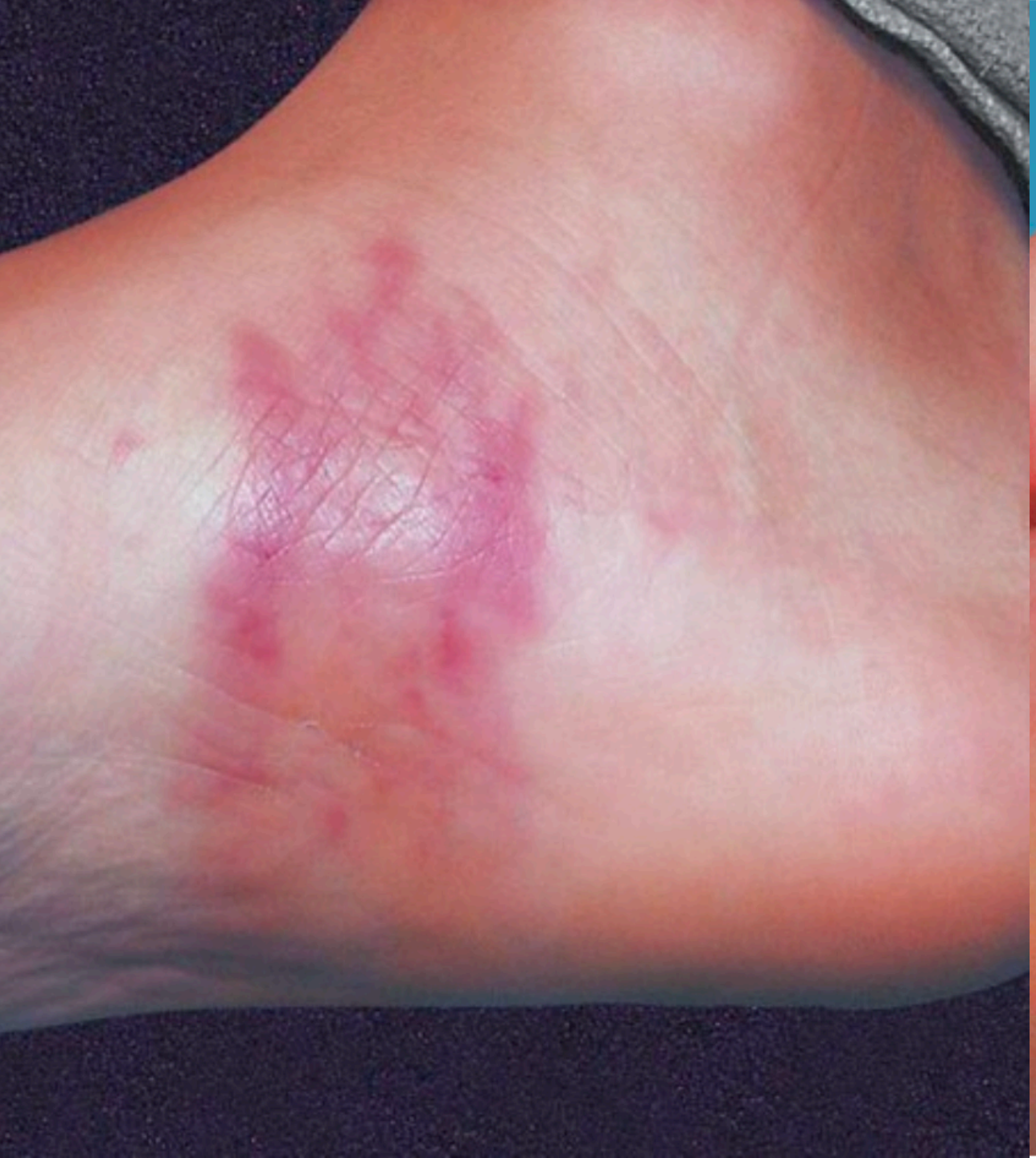
- Characterized by:
- Involvement of small to medium-sized vessels
- The presence of antineutrophilic cytoplasmic antibodies (ANCA)
- An overlapping spectrum of organ involvement

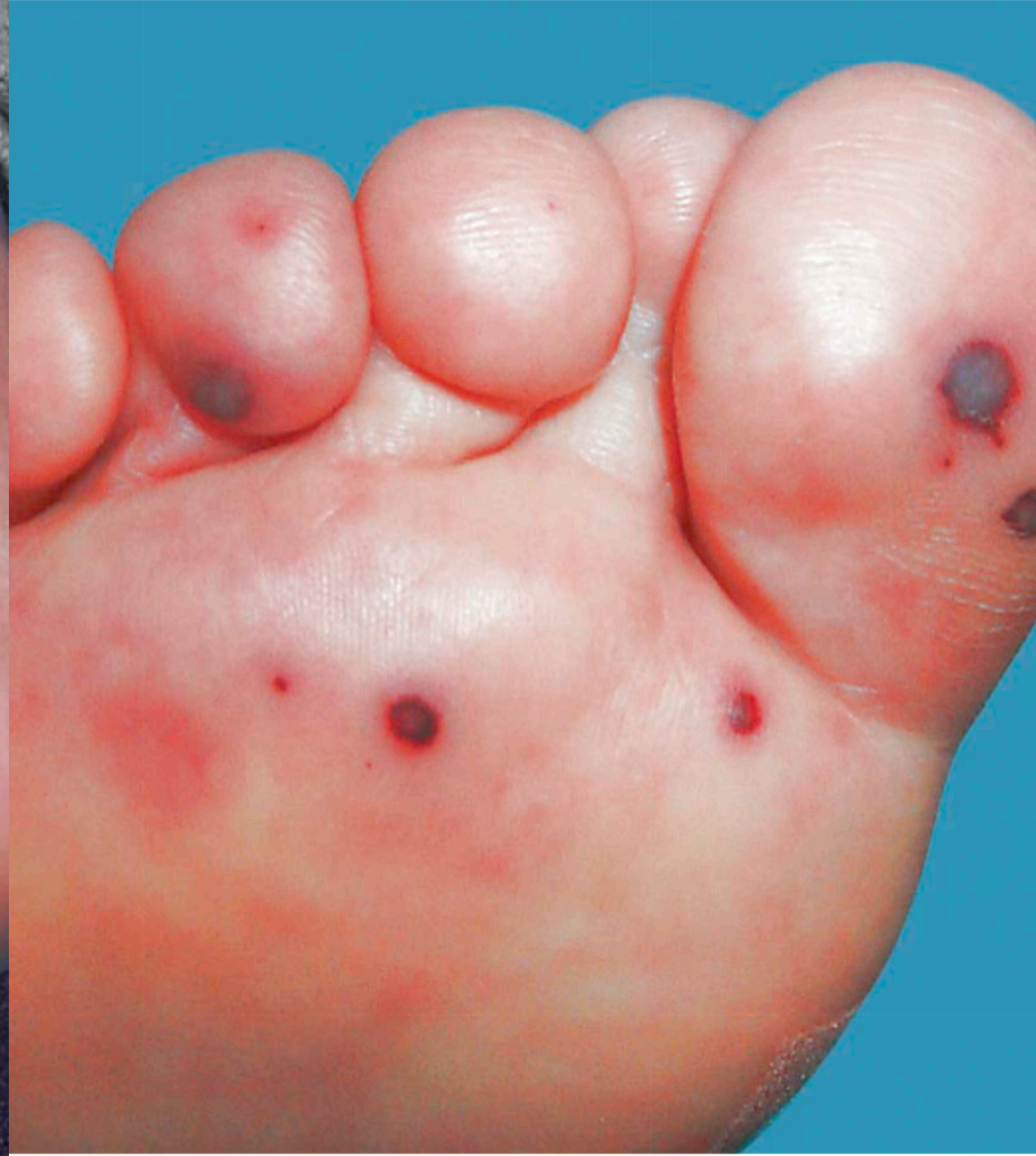
- Each have distinguishing clinical and laboratory features.

1) **Microscopic polyangiitis (MPA)**
2) **Wegner's granulomatosis**
3) **Churg-Strauss syndrome**

Microscopic polyangiitis

- Vasculitis of capillaries, venules and medium-sized arteries.
- Palpable purpura, erythematous macules and patches, splinter hemorrhages and ulcers.
- Constitutional symptoms, crescentic necrotizing glomerulonephritis and alveolar hemorrhage.
- Presence of **P-ANCA**.
- Absence of granuloma formation.

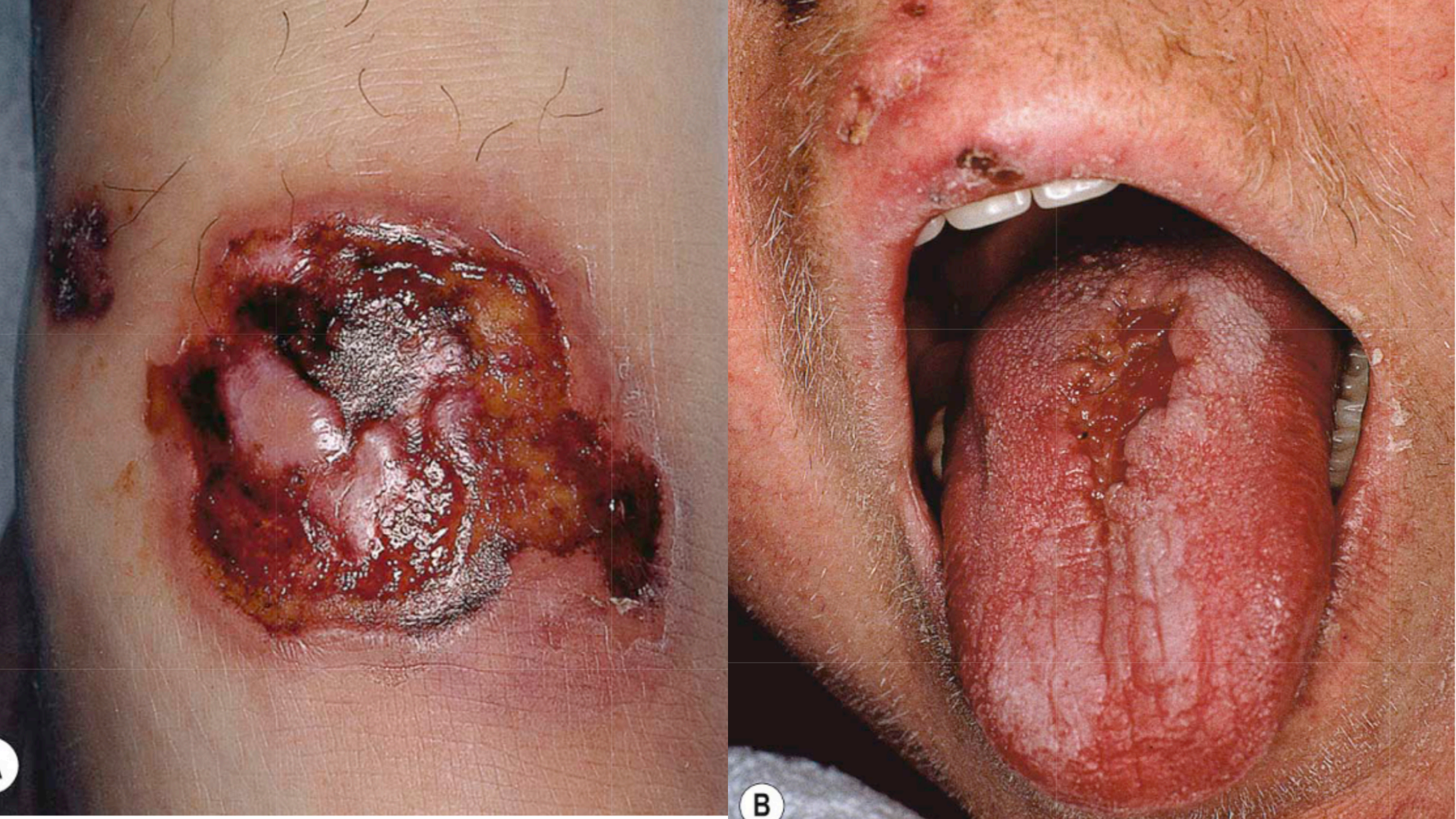




Granulomatosis with polyangiitis (Wegener)

- Necrotizing granulomatous inflammation of the upper and lower respiratory tracts.
- Pauci-immune glomerulonephritis.
- Systemic vasculitis that can involve the skin and oral mucosa.
- Positive **C-ANCA** and Anti-PR3
- The most common mucocutaneous manifestations:

- Palpable purpura
- Oral ulcers
- Strawberry gums
- Papulonecrotic lesions on the extremities.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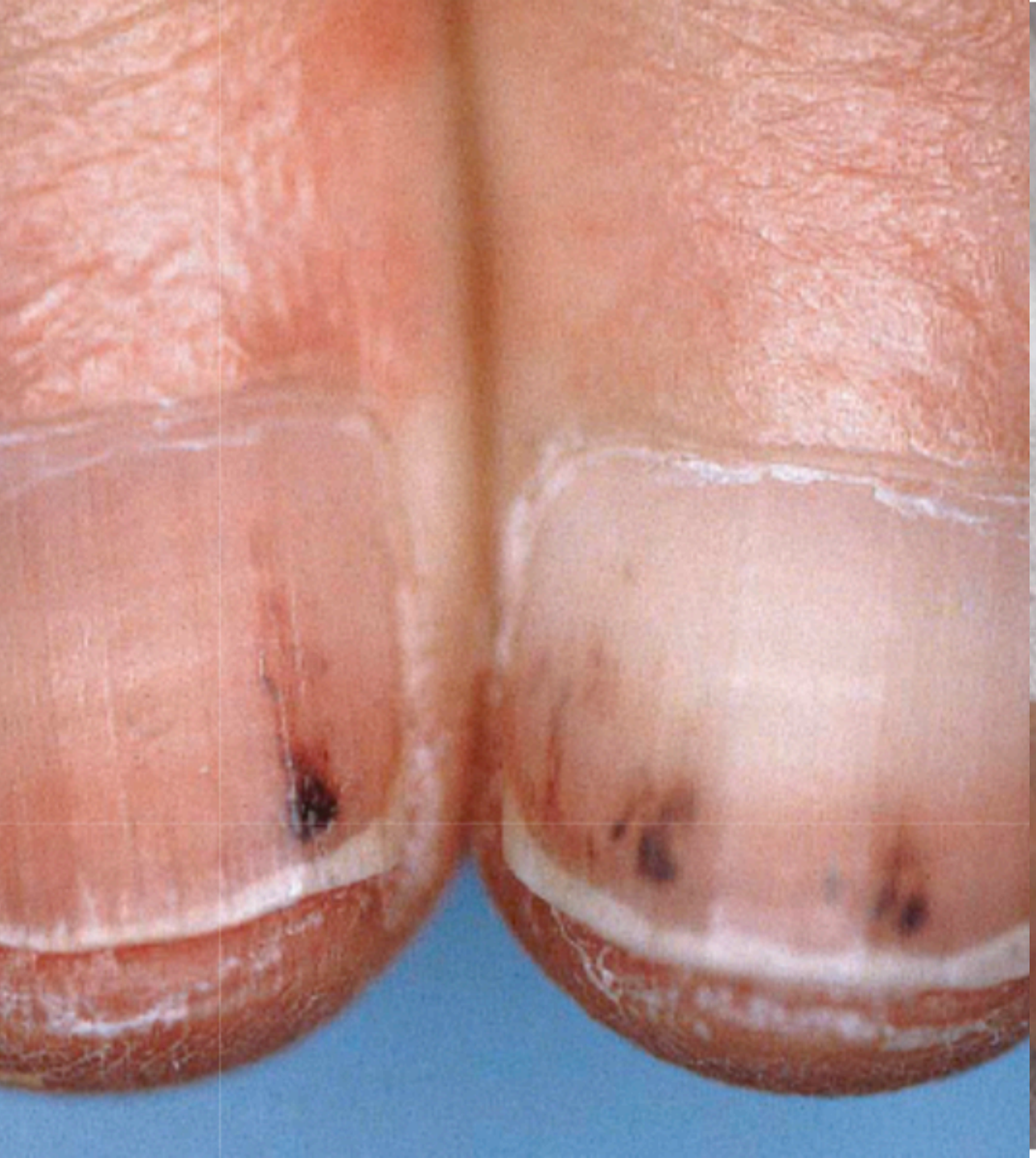
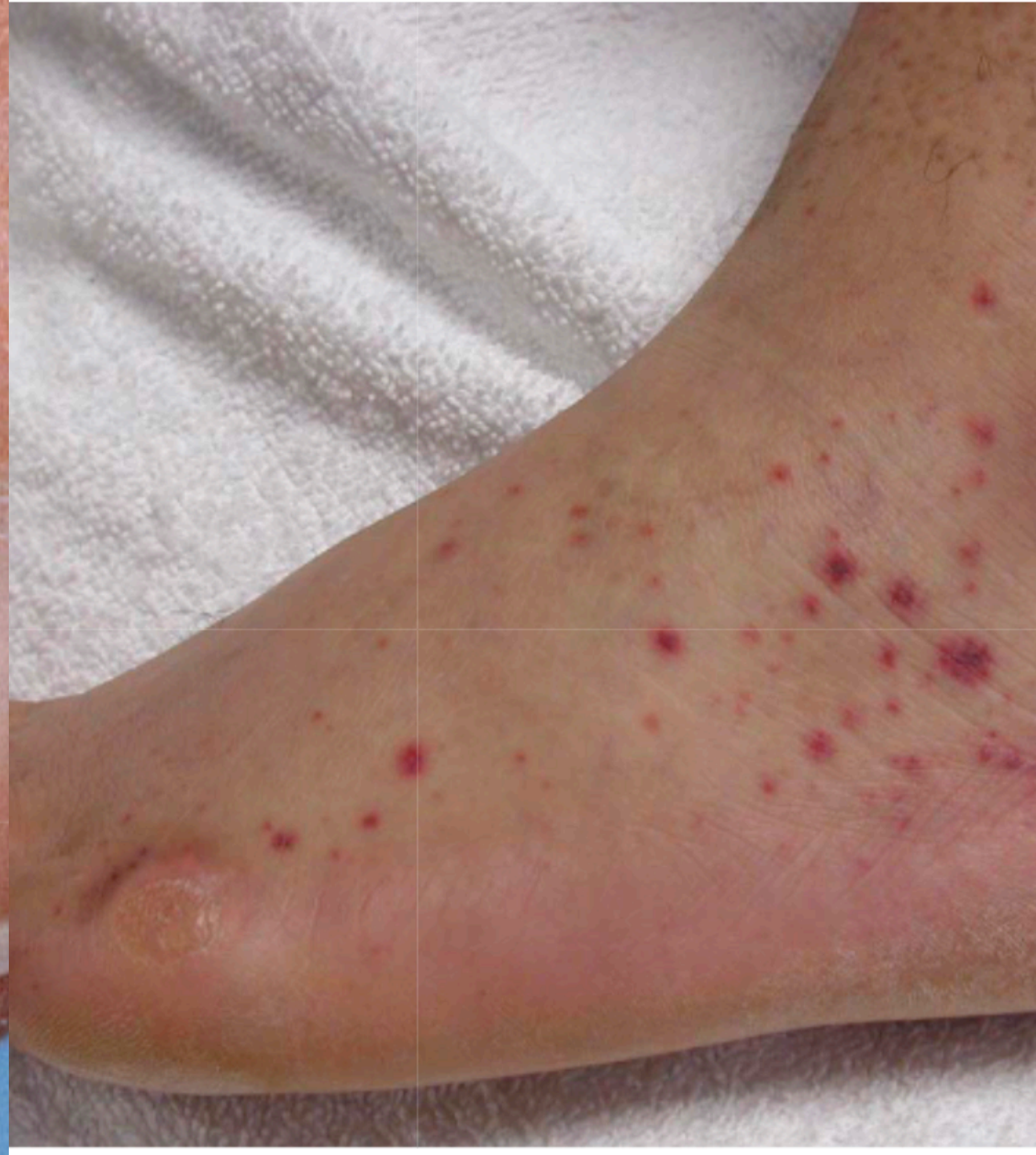




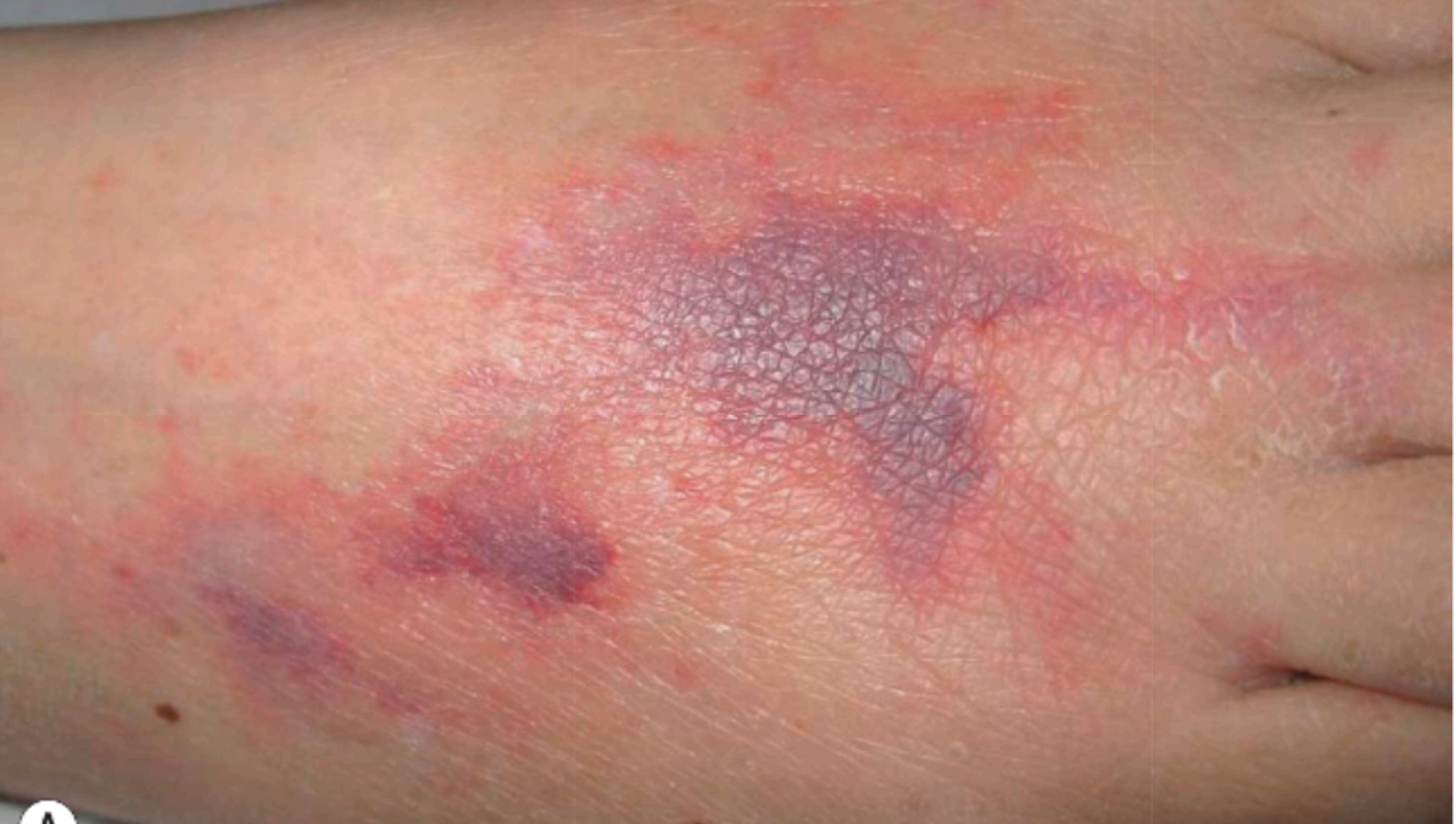
Churg-Strauss syndrome

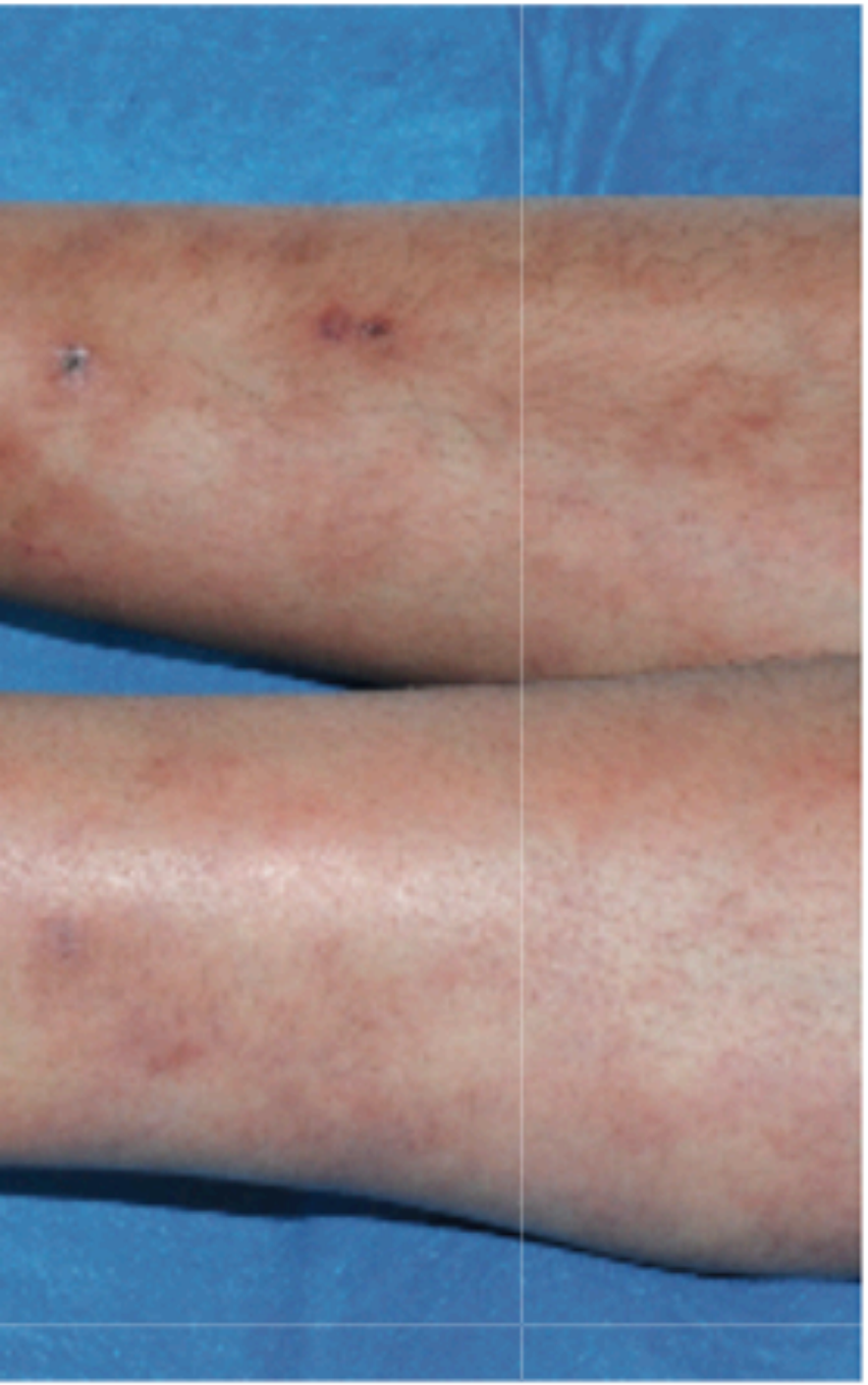
- Asthma and allergic rhinitis typically precede vasculitic phase.
- Elevated blood eosinophil count
- Cutaneous vasculitis in approximately half of patients.
- Histologic features consist of eosinophils, extravascular granulomas and vasculitis.
- Laboratory findings are similar to Wegner's with the additional findings of peripheral eosinophilia and elevated serum IgE levels.

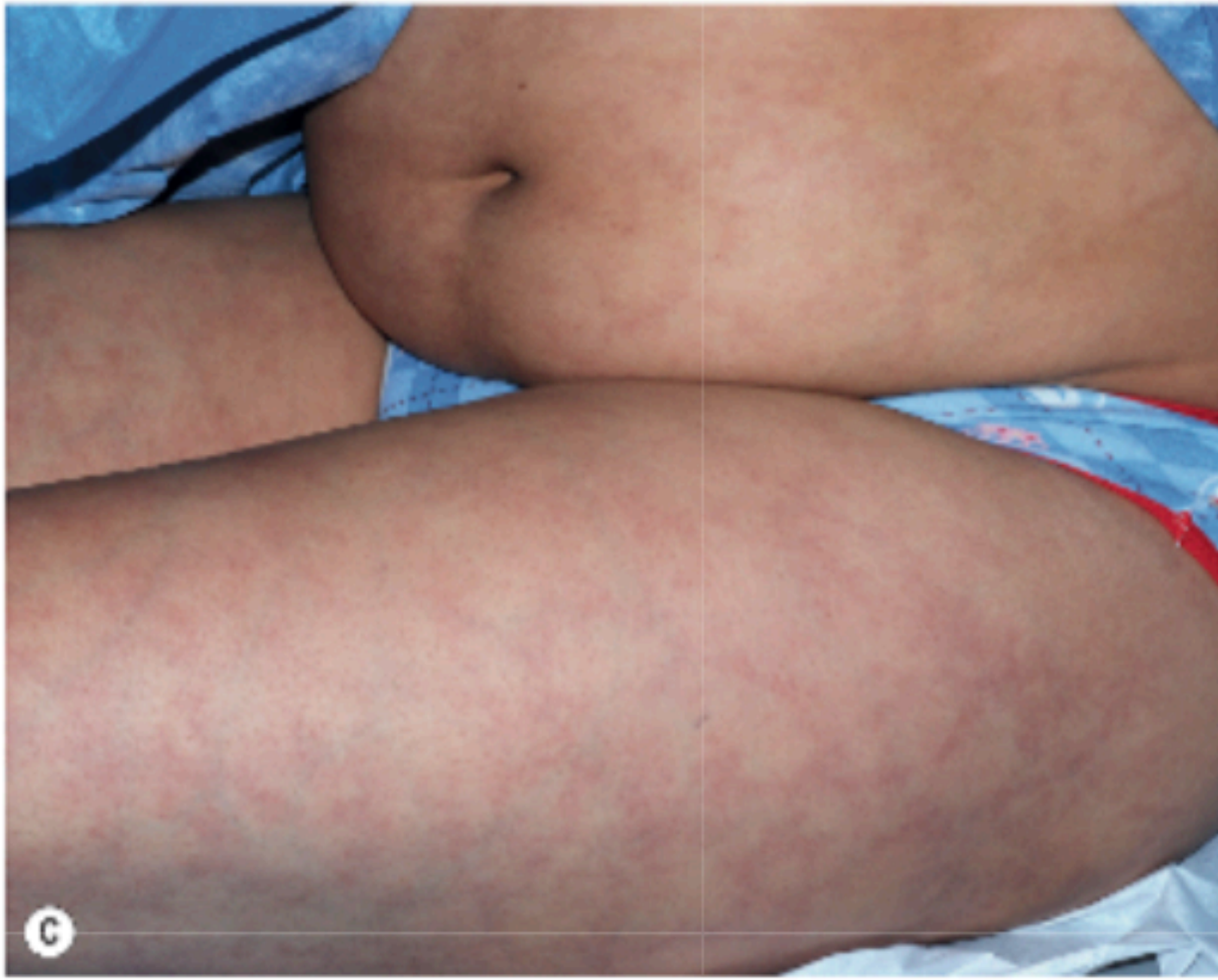
Medium vessel vasculitis

Polyarteritis nodosa

- Segmental multisystem vasculitis of predominantly medium-sized arteries.
- Systemic and cutaneous variants both can present with palpable purpura, livedo racemose, retiform purpura, ulcers, subcutaneous nodules or peripheral gangrene.
- Extracutaneous manifestations of the systemic variants include: Fever, arthralgias, myalgias, paresthesias, abdominal pain, orchitis and renovascular hypertension.
- The cutaneous variant has a chronic, more benign course; it may be accompanied by mild systemic symptoms (fever, myalgias, arthralgias and peripheral neuropathy).



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