# Purpura and Vasculitis

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# Objectives

- Differentiate between different types of purpura
- Identify the morphology of different types of purpura
- Recognize palpable purpura as a hallmark lesion of leukocytoclastic vasculitis
- Outline an initial diagnostic approach to diagnose purpura

# Purpura

## What is the definition of Purpura ?

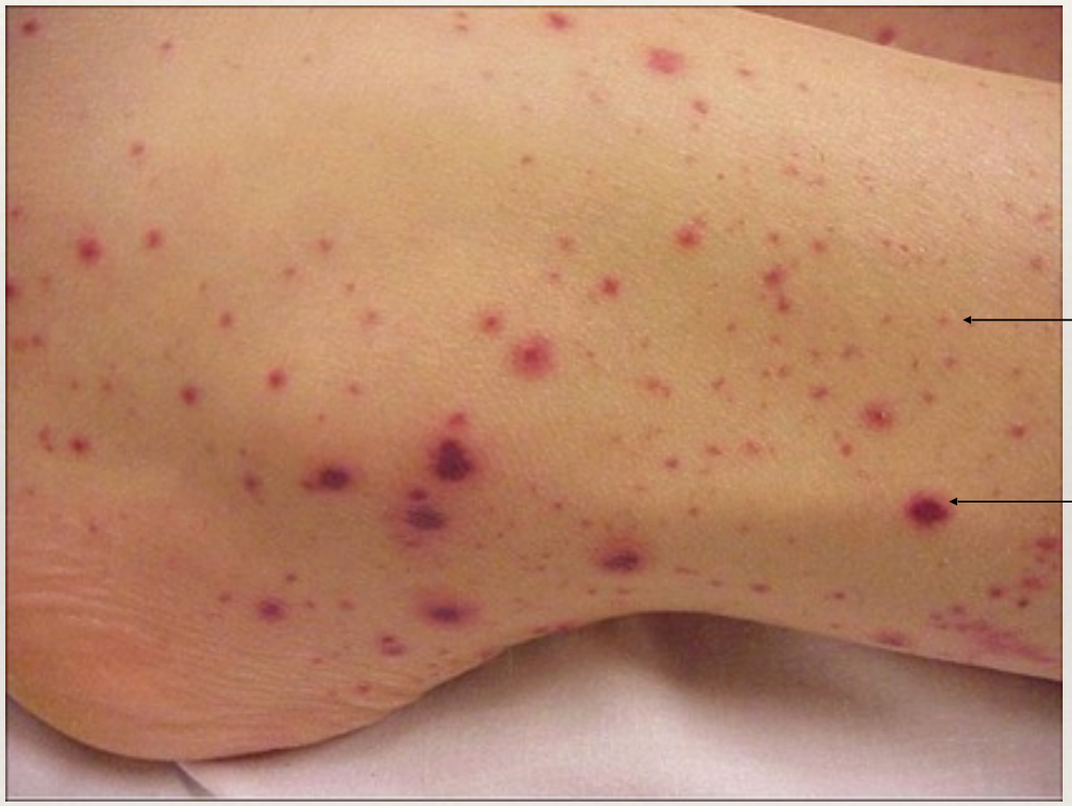
# Purpura

- Purpura is multifocal extravasation of blood into the skin or mucous membranes

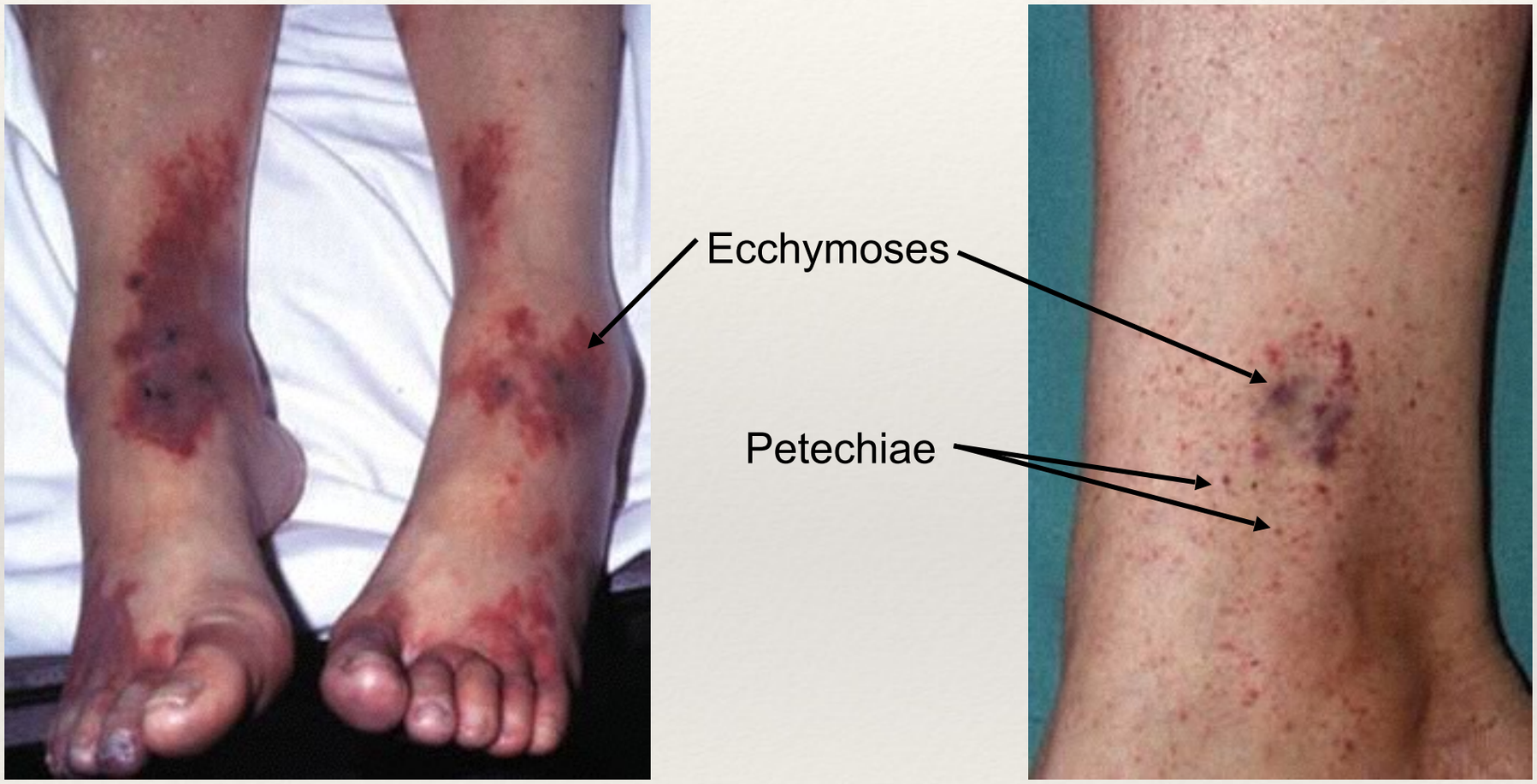
# Purpura

- Purpura may be palpable or non-palpable
- Non-palpable purpura are divided into 2 morphologies based on their size:
  - Petechiae- (< 3mm) superficial, pinhead-sized, hemorrhagic macules
  - Ecchymoses- ( > 3 mm) irregularly shaped, bluish-purpulish patches “ bruises”

# Purpura



# Purpura



# Purpura

- The type of lesion usually indicates the underlying pathogenesis;
  - non-palpable purpura is typically non-inflammatory
  - palpable purpura is usually a sign of vascular inflammation "hallmark lesion of **leukocytoclastic vasculitis**"

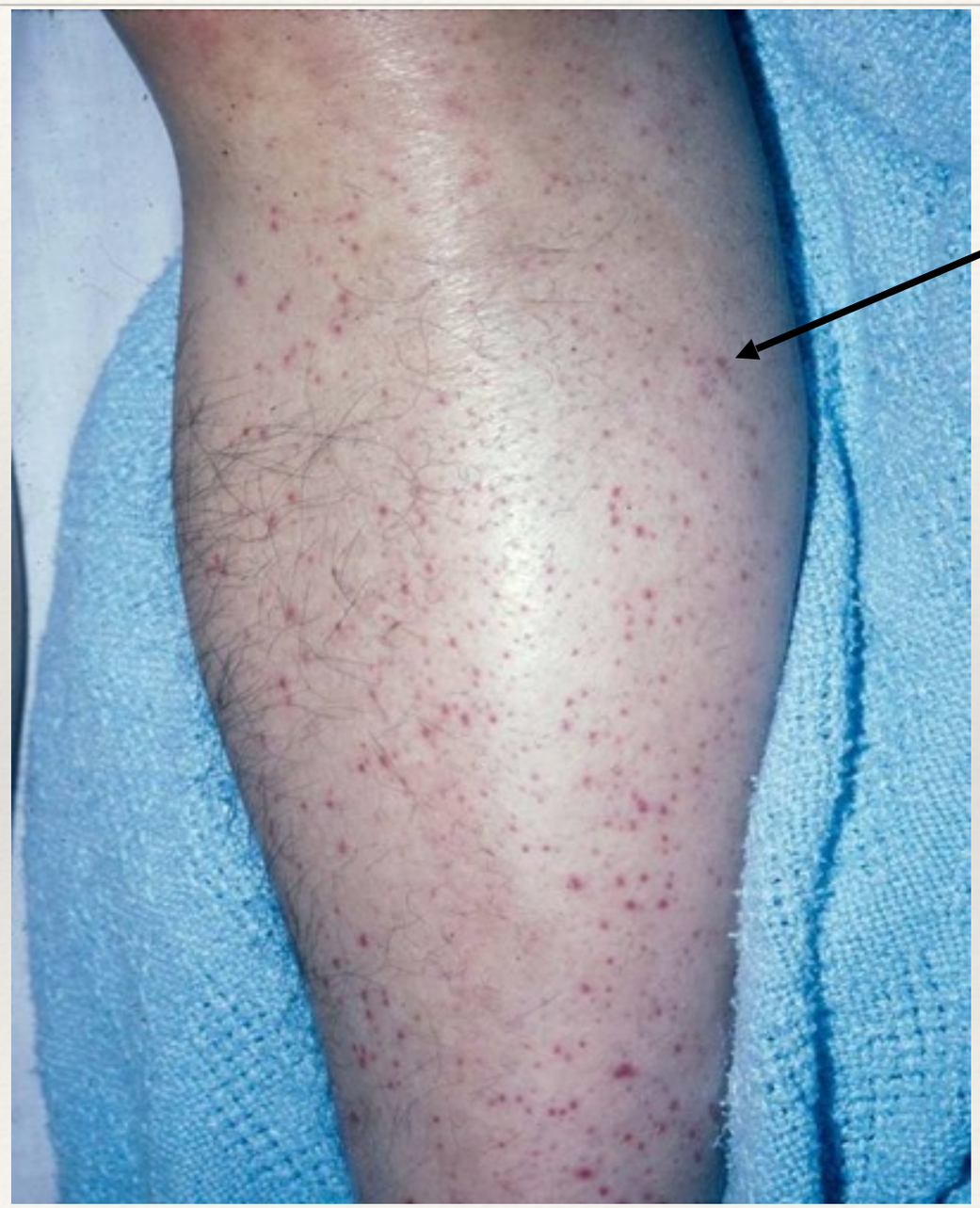
# Purpura

- **Causes of non-palpable purpura:**
  - Trauma
  - 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 deficincy, hepatic disease

# Purpura

- Vitamin C deficiency "**Scurvy**"
  - perifollicular petechiae
  - keratotic plugging of hair follicles
  - hemorrhagic gingivitis

# Purpura



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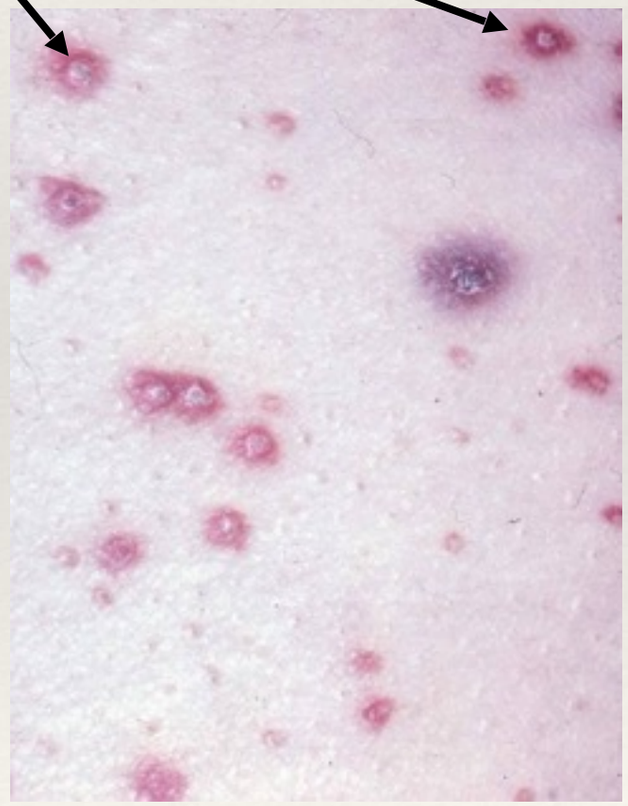
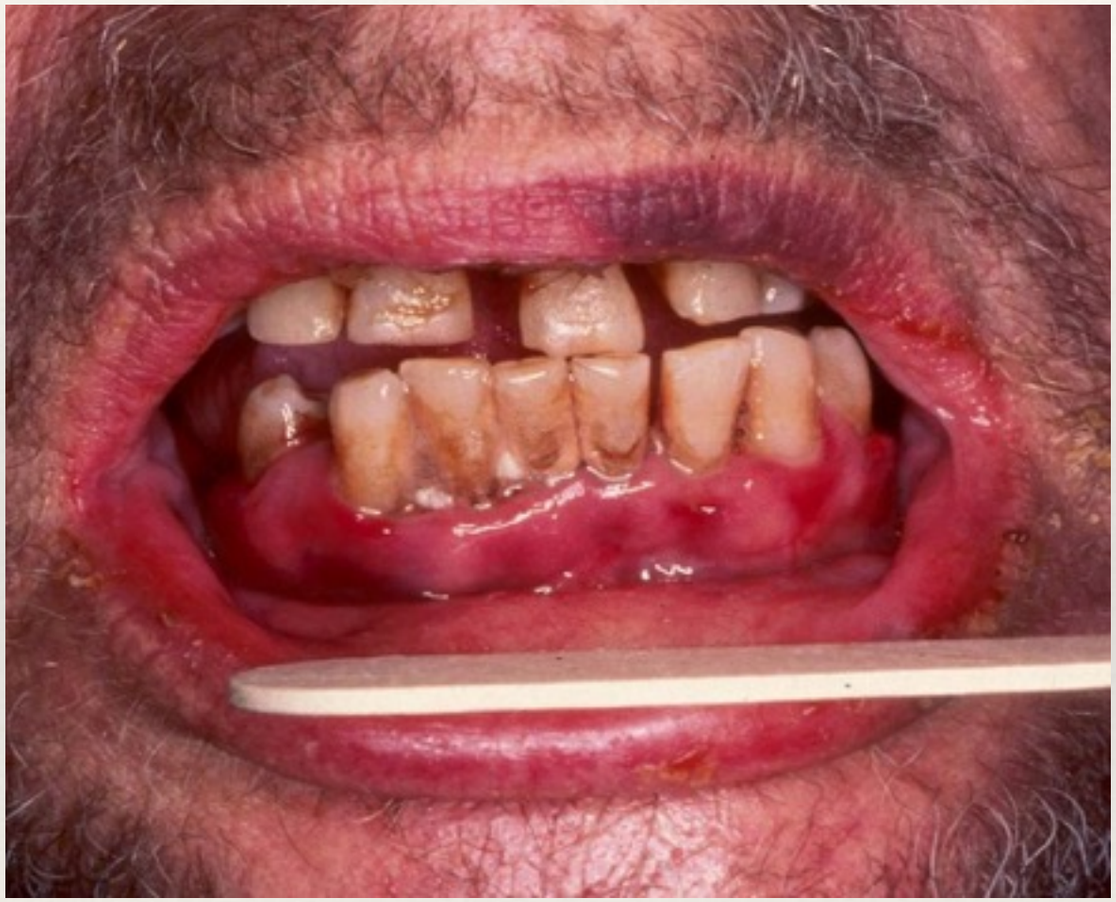


# Purpura

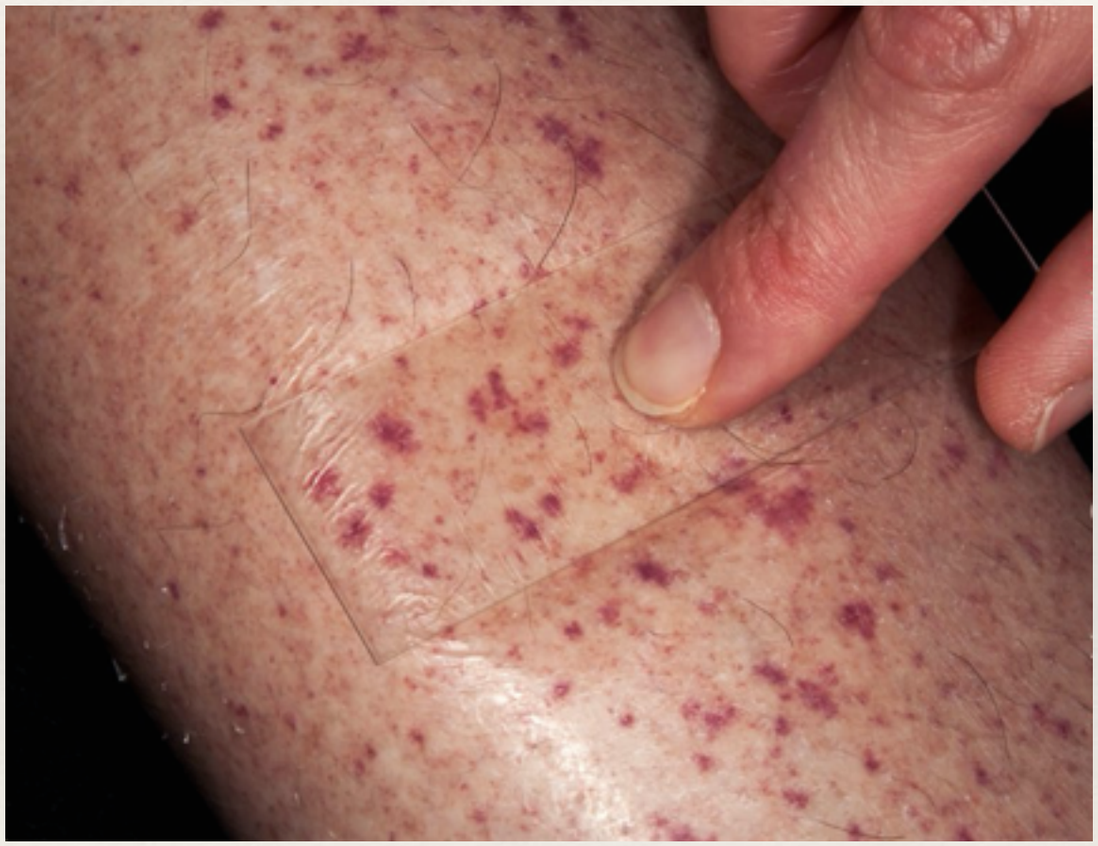


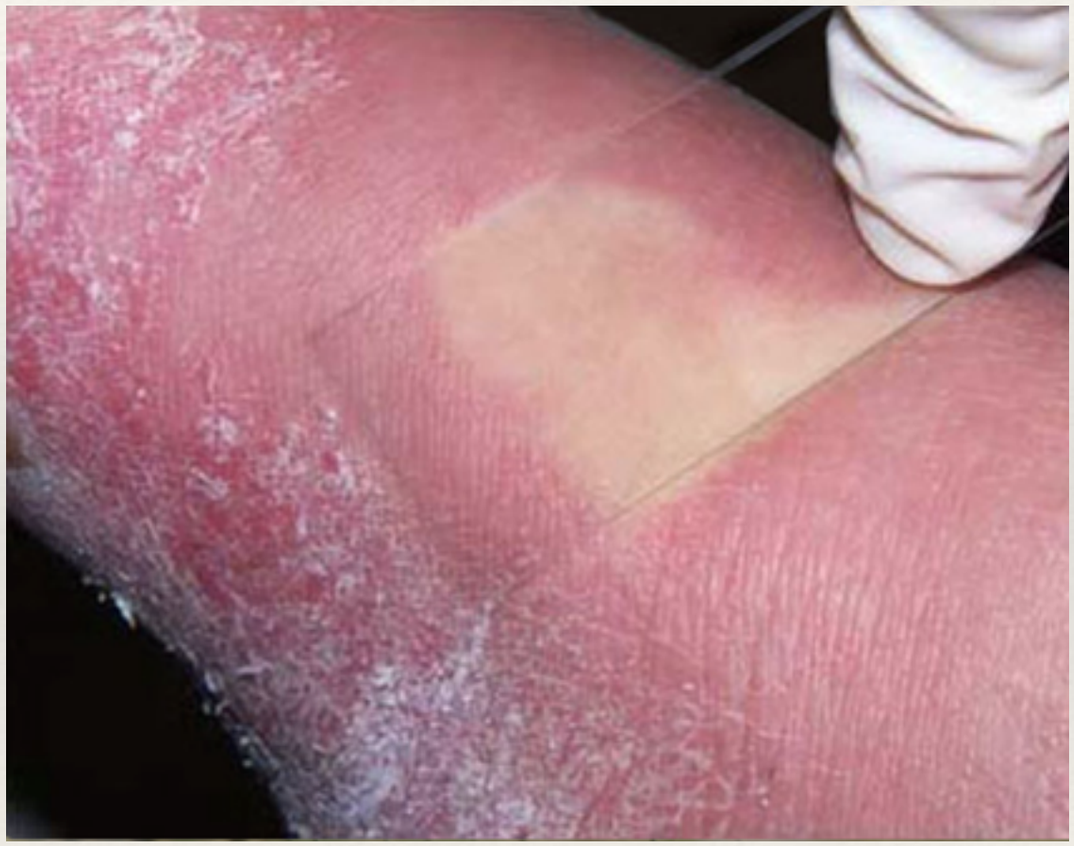
- hemorrhagic gingivitis

# Purpura

- All forms of purpura do **NOT** blanch with pressure
- **Diascopy**- use of a glass slide to apply pressure to the lesion to differentiate erythema **secondary to vasodilation** ( planchable with pressure), from **extravasation of blood** ( non-blanchable)

# Purpura





# Purpura

## How do we evaluate a patient with purpura?

# Purpura

- History & Physical examination

| History | Examination |
| --- | --- |
| • family Hx | • size |
| • drug Hx | • type |
| • medical Hx | • distribution |
| | • mucous membranes |

- CBC & Differential
- Bleeding time
- PT & PTT

# Vasculitis

- 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

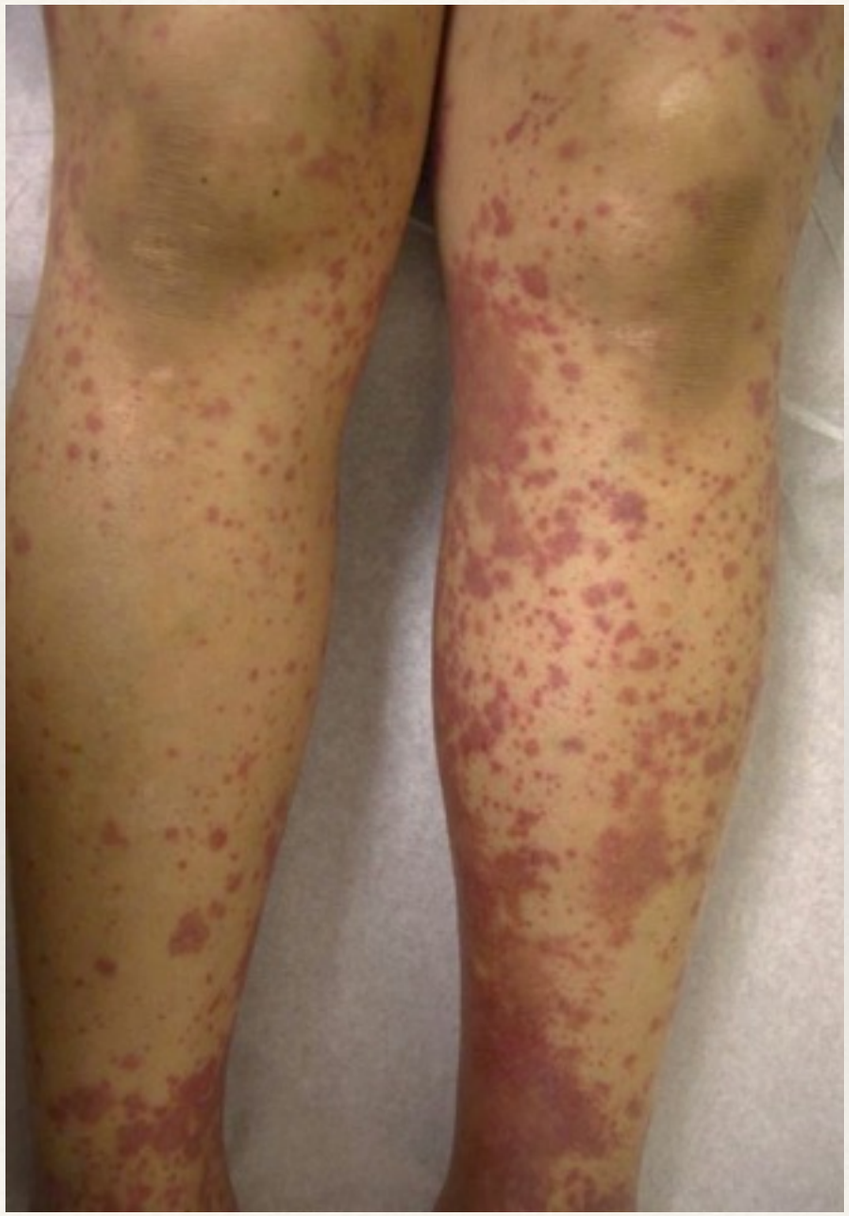
# Vasculitis

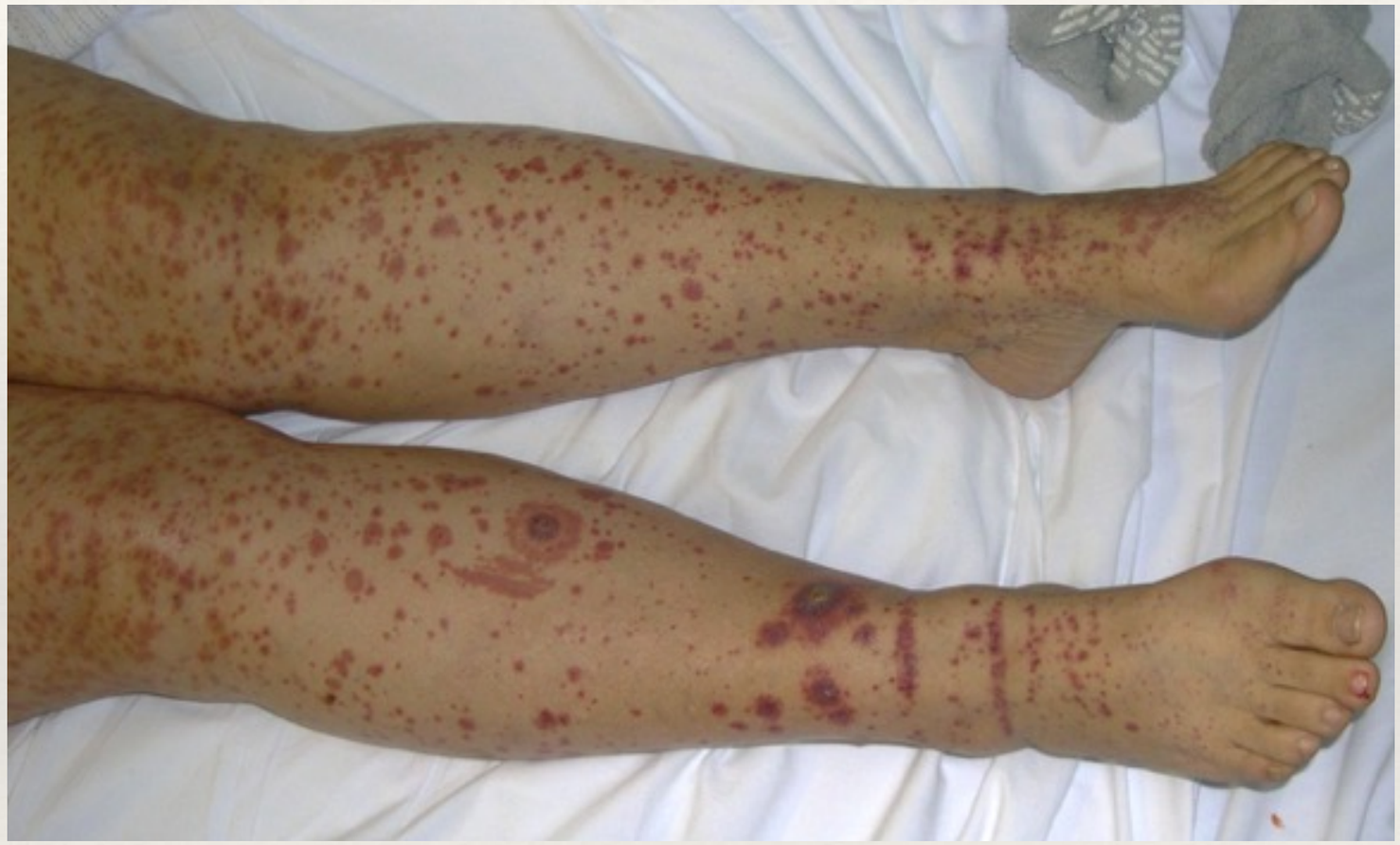
1. **Cutaneous small vessels ( Leukocytoclastic vasculitis)**
   - Henoch-Schönlein purpura
   - Urticarial vasculitis
   - Other
     - idiopathic
     - infection- streptococcal, bacterial endocarditis, parvovirus B19, HIV, hepatitis, TB
     - drugs- NSAID,sulfonamides, penicillins, barbiturates, propylthiouracil
     - malignancy- leukemias, lymphoma, multiple myeloma, renal, lung, prostate,breast
2. **Mixed ( small and medium) vessels**
   - ANCA associated vasculitides
     - Churg-Stauss syndrome
     - Microscopic polyangiitis
     - Granulomatosis with polyangiitis (Wegener)
   - Essential Cryoglobulinemic vasculitis
3. **Medium vessels**
   - Polyarteritis nodosa- Cutaneous & systemic
4. **Large vessels**
   - Giant-cell arteritis
   - Takayasu arteritis

# Cutaneous small vessel vasculitis
# "Leukocytoclastic vasculitis"

- Could occur as a **primary** process or could be **secondary** to an underlying cause
- The majority of cases follow an acute infection or exposure to a new medication
- **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
- They resolve within 3-4 weeks with residual post-inflammatory hyperpigmentation

# Cutaneous small vessel vasculitis
# “Leukocytoclastic vasculitis”

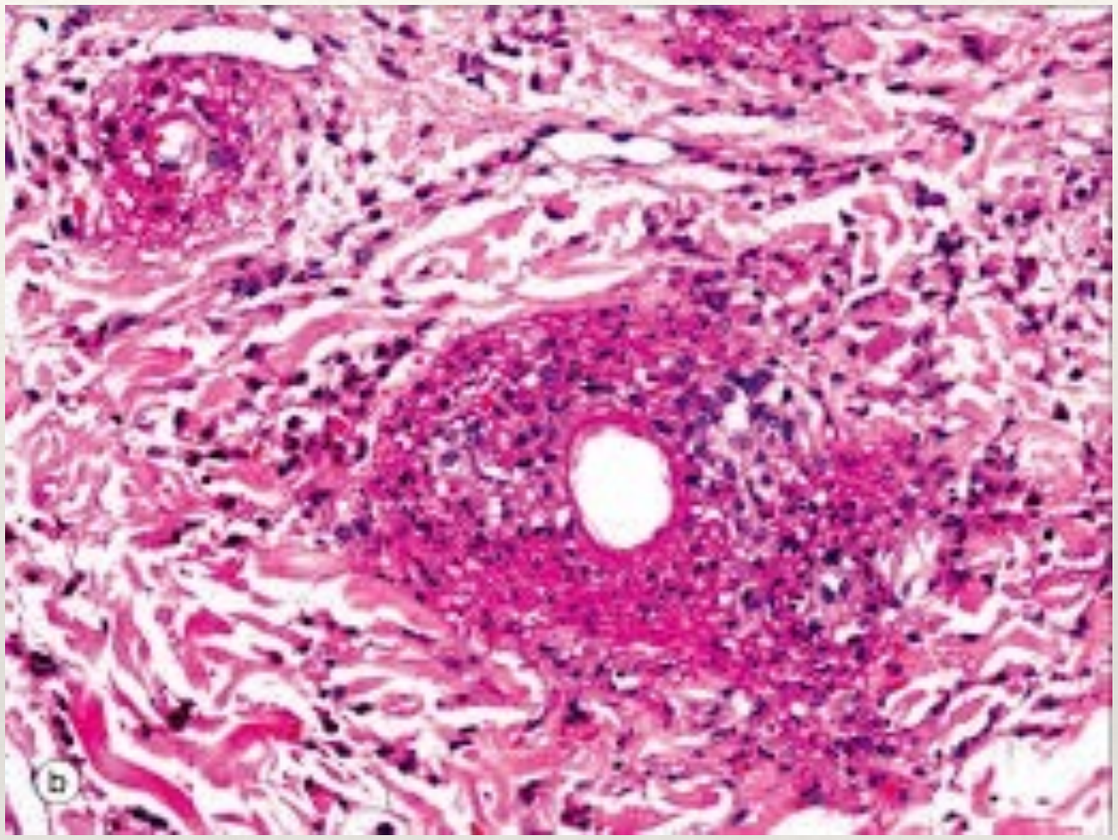




# Cutaneous small vessel vasculitis
# "Leukocytoclastic vasculitis"

## ❖ Histopathology:

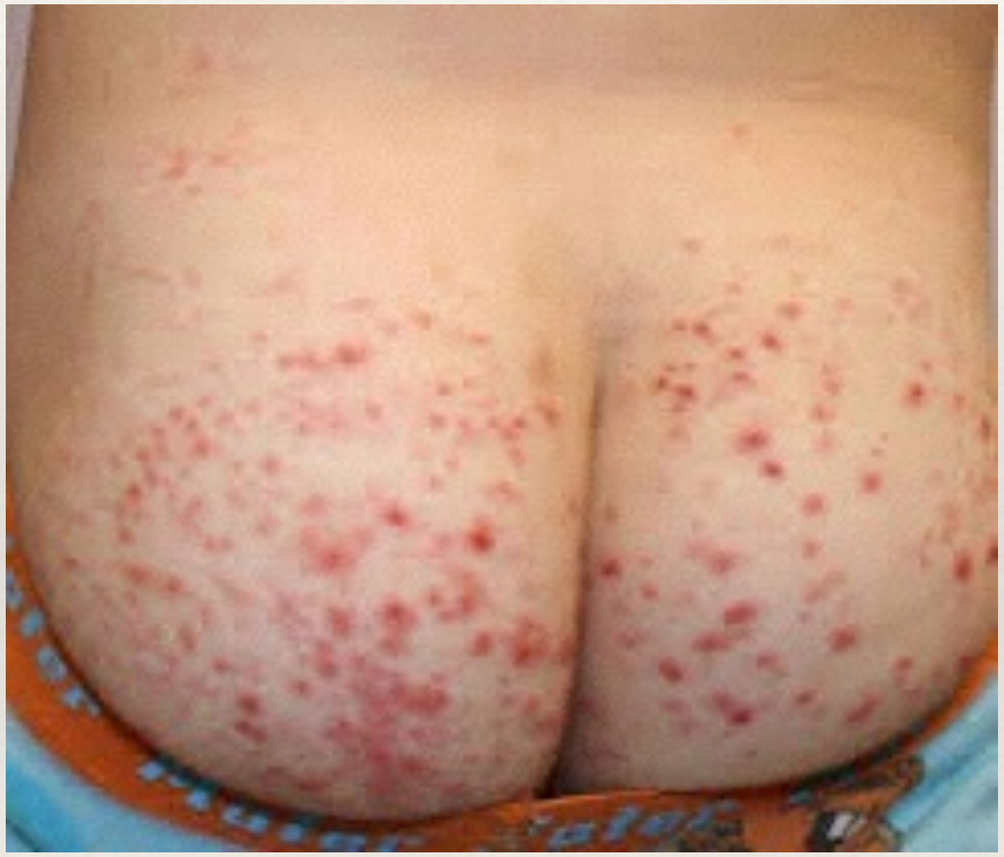
- Inflammation in the form of perivascular infiltrate comprised of intact and fragmented neutrophils ( nuclear dust), hence, "leukocytoclastic vasculitis"
- Blood vessel wall thickening
- Errythrocyte extravasation
- Fibrin deposits within the blood vessel wall
- Endothelial necrosis ( more serious illness)
- immunoglobulin & complement deposits

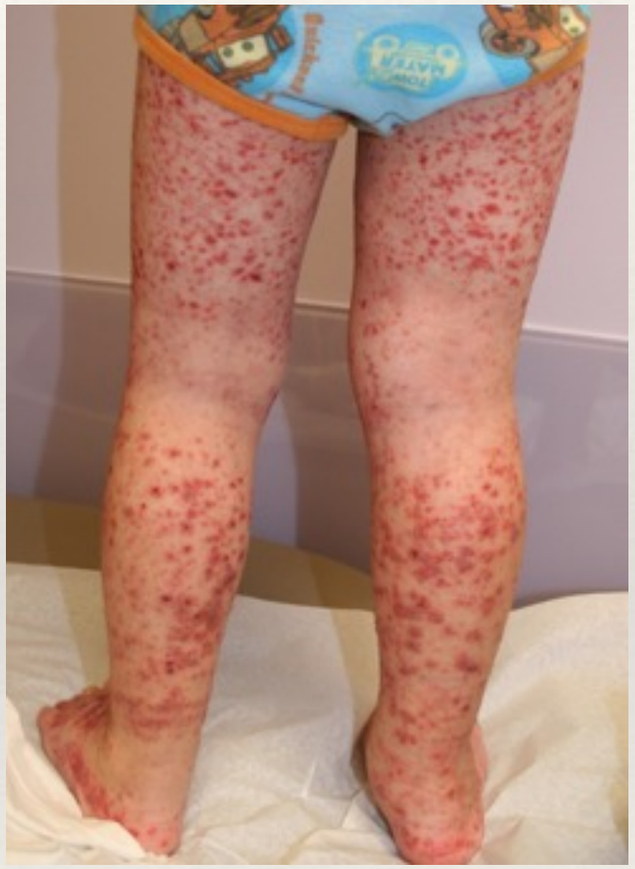


# Henoch-Schönlein Purpura

- Subtype of cutaneous small-vessel vasculitis
- Its a leukocytoclastic vasculitis that mostly affects children, with a predominant IgA-mediated vessel injury
- A viral infection or streptococcal pharyngitis is the usual triggering event, other triggers: bacterial infections, foods, drugs ( aspirin, penicillin), lymphoma
- Characterized by: purpura, arthralgias, abdominal pain and renal disease
- Multiple palpable purpura appears on the extensor aspects of the extremities ( mainly lower legs and to a lesser extent on the forearms) and buttocks
- Histologically; LCV, IgA, C3 and fibrin deposits

# Henoch-Schönlein Purpura





# Henoch-Schönlein Purpura

- **Course of the disease & possible complications:**
  - The duration of the illness is 6-16 weeks
  - In most patients the disease usually resolves without sequelae
  - 5-10 % of patients will have persistent or recurrent disease
  - Arthalgias may progress to arthritis producing periarticular swelling around the knees and ankles
  - GI bleeding, acute surgical abdomen, paralytic ilieus may occur
  - Progressive glomerular disease " crescentic glomerulonephritis", renal failure may occur
  - Pulmonary hemorrhage, can be fatal

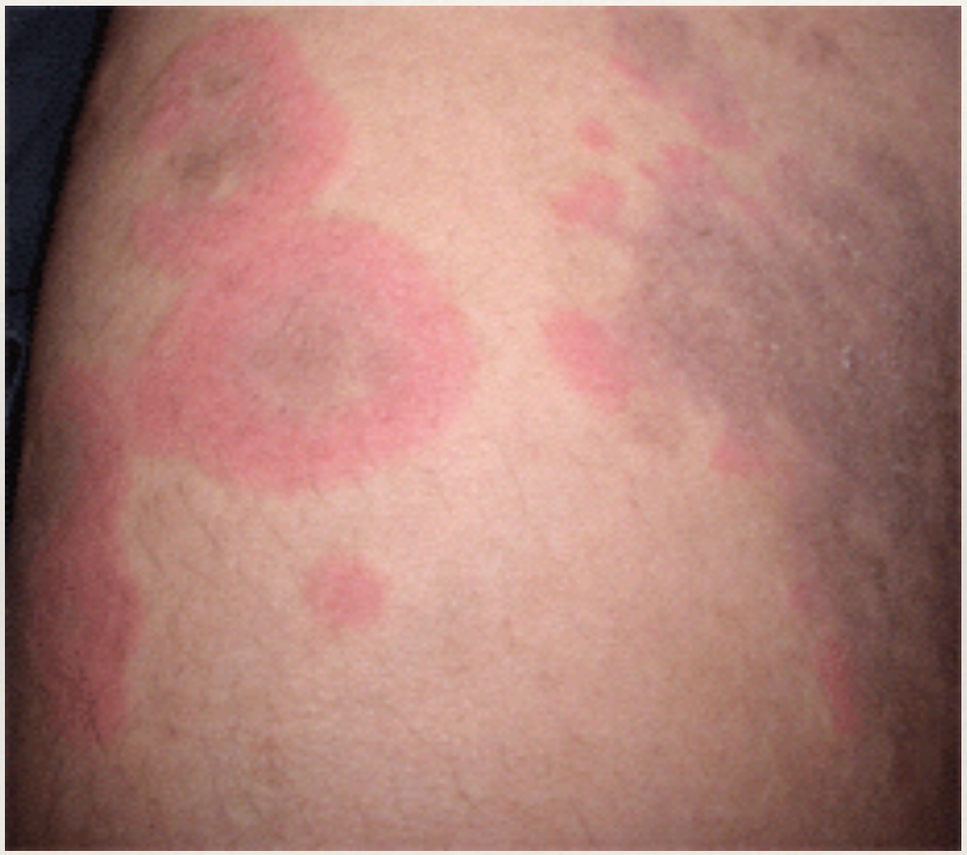
# Henoch-Schönlein Purpura

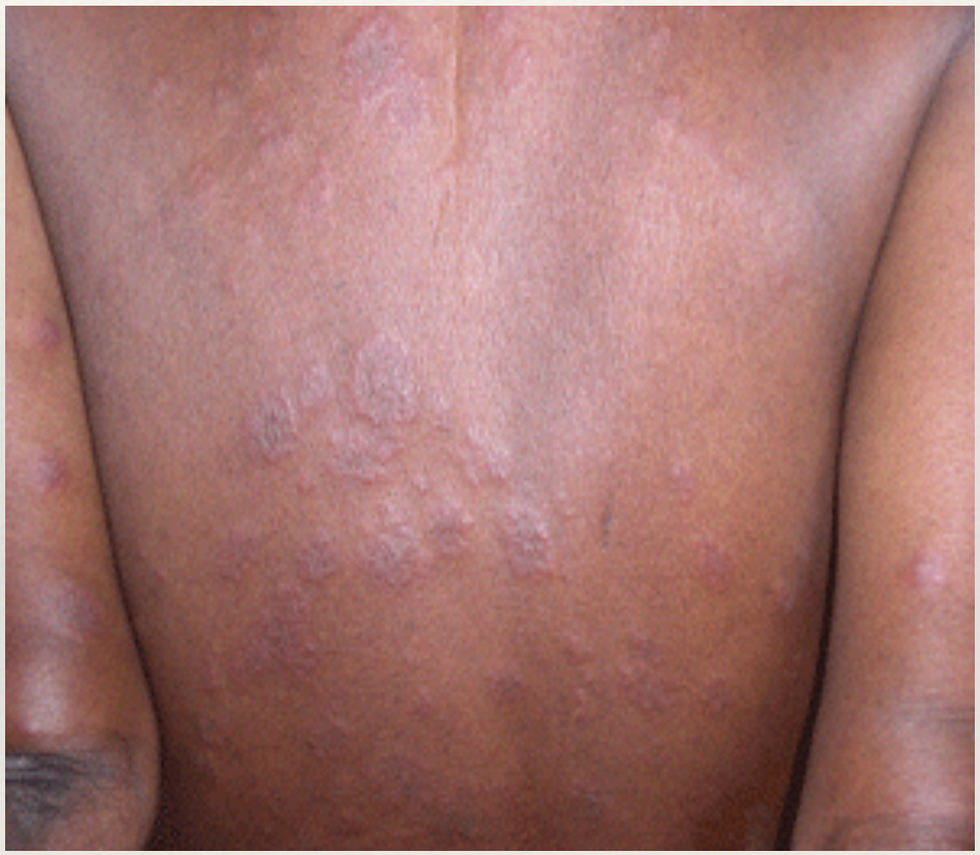
- Treatment
  - Supportive ( bed rest, pain relieve, D/C drugs, treat underlying infection)
  - Abdominal pain- H2 blockers, corticosteroids
  - NSAIDs are best avoided ( renal & GI complications)

# Urticarial Vasculitis

- Fixed urticarial lesions that when biopsied will have vasculitis histology
- 3 clinical features distinguish the skin lesion of urticarial vasculitis from urticaria:
    1. Lesions are rather painful, rather than pruritic
    2. Lesions last longer than 24 h and are fixed, rather than pruritic
    3. On resolving there is postinflammatory hyperpigmentation

# Urticarial Vasculitis





# Urticarial Vasculitis

- Determination of complement levels ( CH50, C3, C4, and anti-C1q) is critical in these patients

<u>Normal complement levels</u>

- idiopathic leukocytoclastic vasculitis
- limited to the skin
- self-resolving

<u>Low complement levels</u>

- leukocytoclastic vasculitis + diffuse interstitial neutrophils
- not limited to the skin; clinical features include arthritis, arthralgia, angioedema eye symptoms, asthma, GI symptoms

- Diseases associated with urticarial vasculitis: gammopathies ( IgG & IgM), SLE, Sjögren syndrome, serum sickness, viral infections ( esp. hepatitis C)

# Urticarial Vasculitis

- **Treatment & Management:**
  - History & physical exam
  - Ix- CH50, C3, C4, C1q, ANA, dsDNA, Anti-SSA & Anti-SSB, hepatitis B&C, lupus band test
  - Treatment is based on the systemic effects of the disease, extent of cutaneous involvement and previous response to treatment
  - Cutaneous involvement- NSAIDs & antihitamines, if these fail —> colchicine, hydroxychloroquine, dapsone, if these fail or if the patient has systemic disease —> corticosteroids + steroid sparing agent ( azathioprine, mycophenolate mofetil, rituximab)

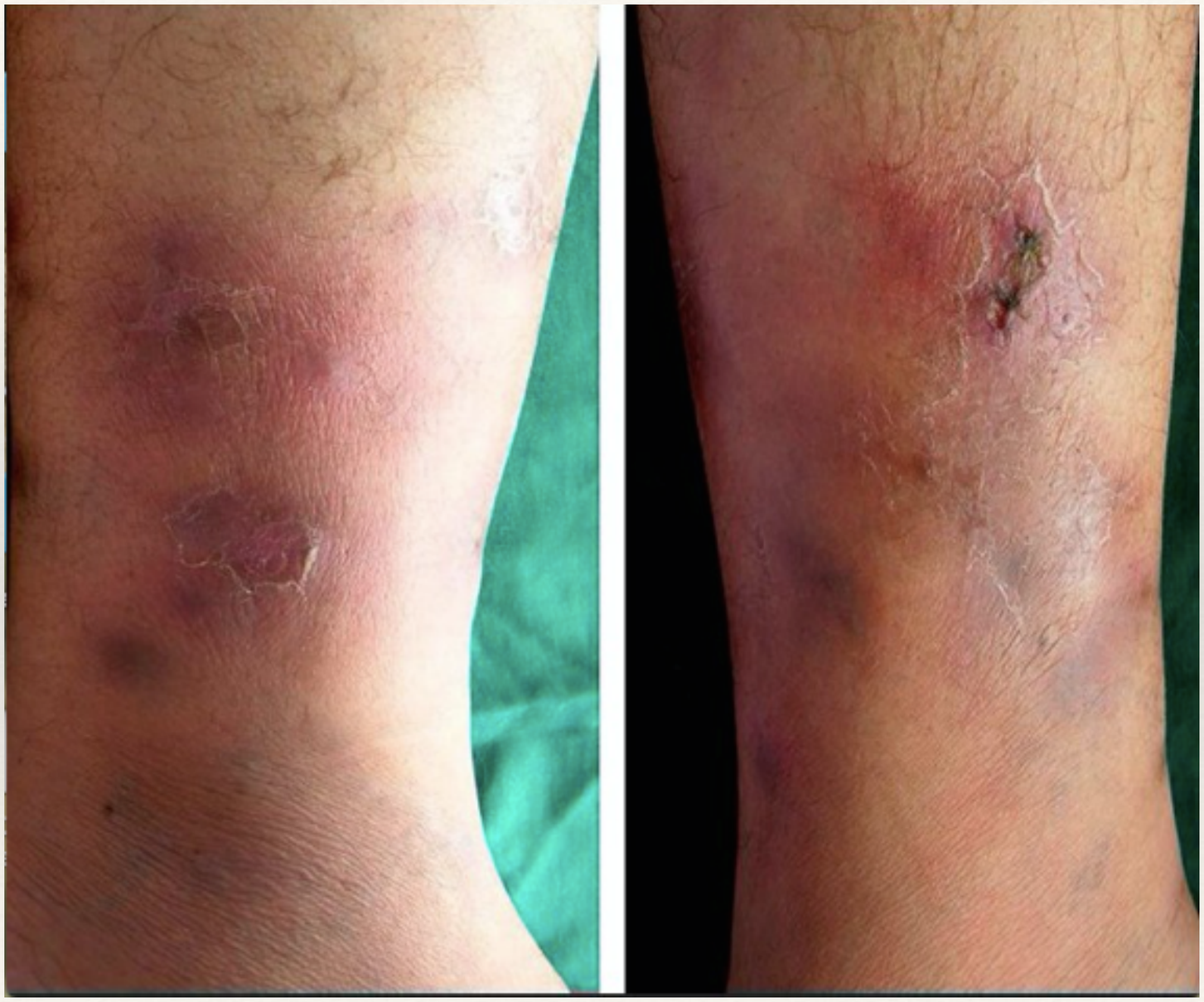
# Cutaneous polyarteritis nodosa

- Necrotizing vasculitis affecting small and- medium sized arteries of the dermis and subcutaneous tissue
- Localized to the skin with limited systemic involvement, usually neuropathy
- Patients should be followed carefully and regularly evaluated to exclude the development of systemic involvement

# Cutaneous polyarteritis nodosa

- Cutaneous findings- almost always subcutaneous nodules associated with livedo reticularis that may ulcerate on the legs and feet
- Peripheral neuropathy- tingling, numbness, sensory disturbances, weakness and absent reflexes

# Cutaneous polyarteritis nodosa



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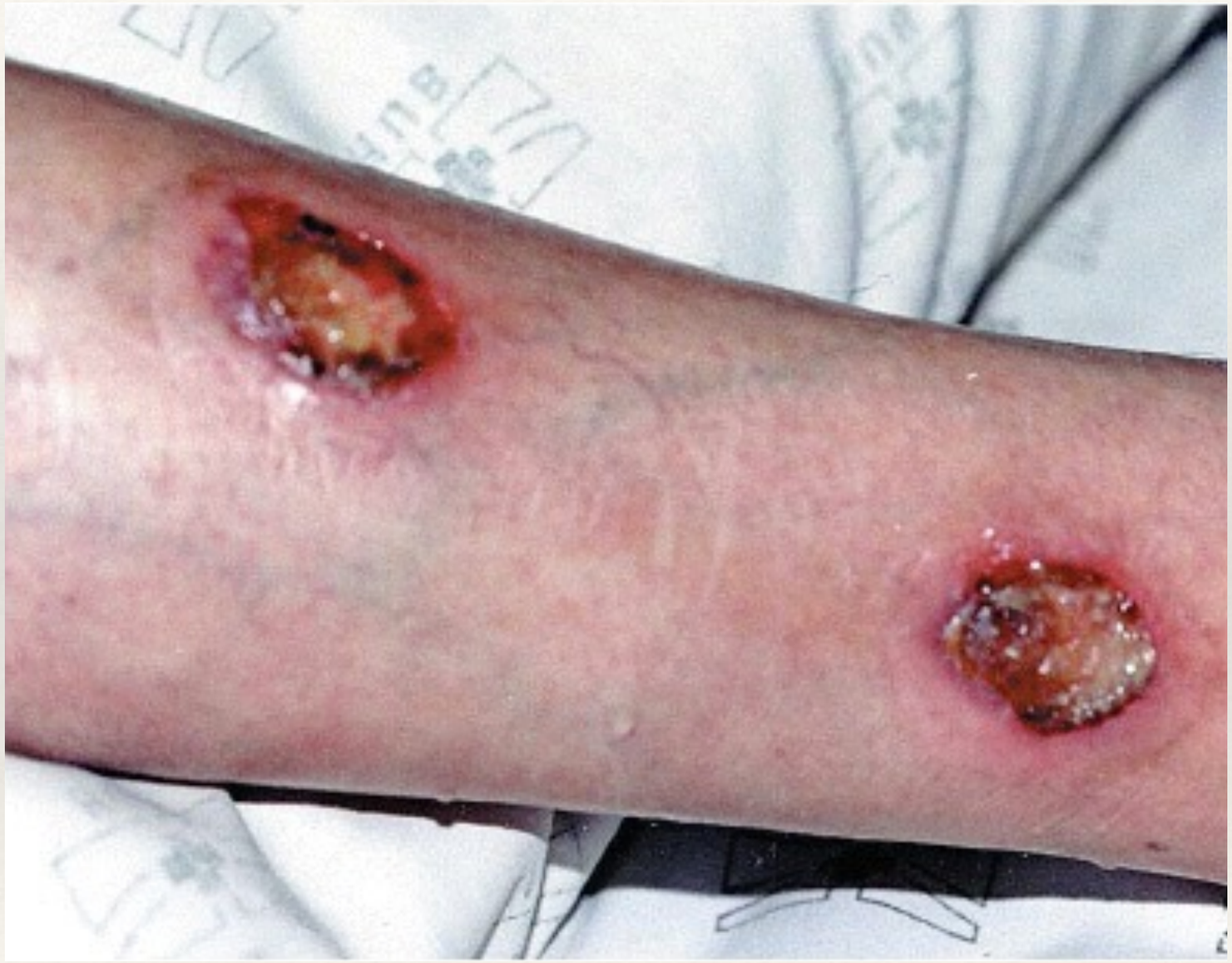
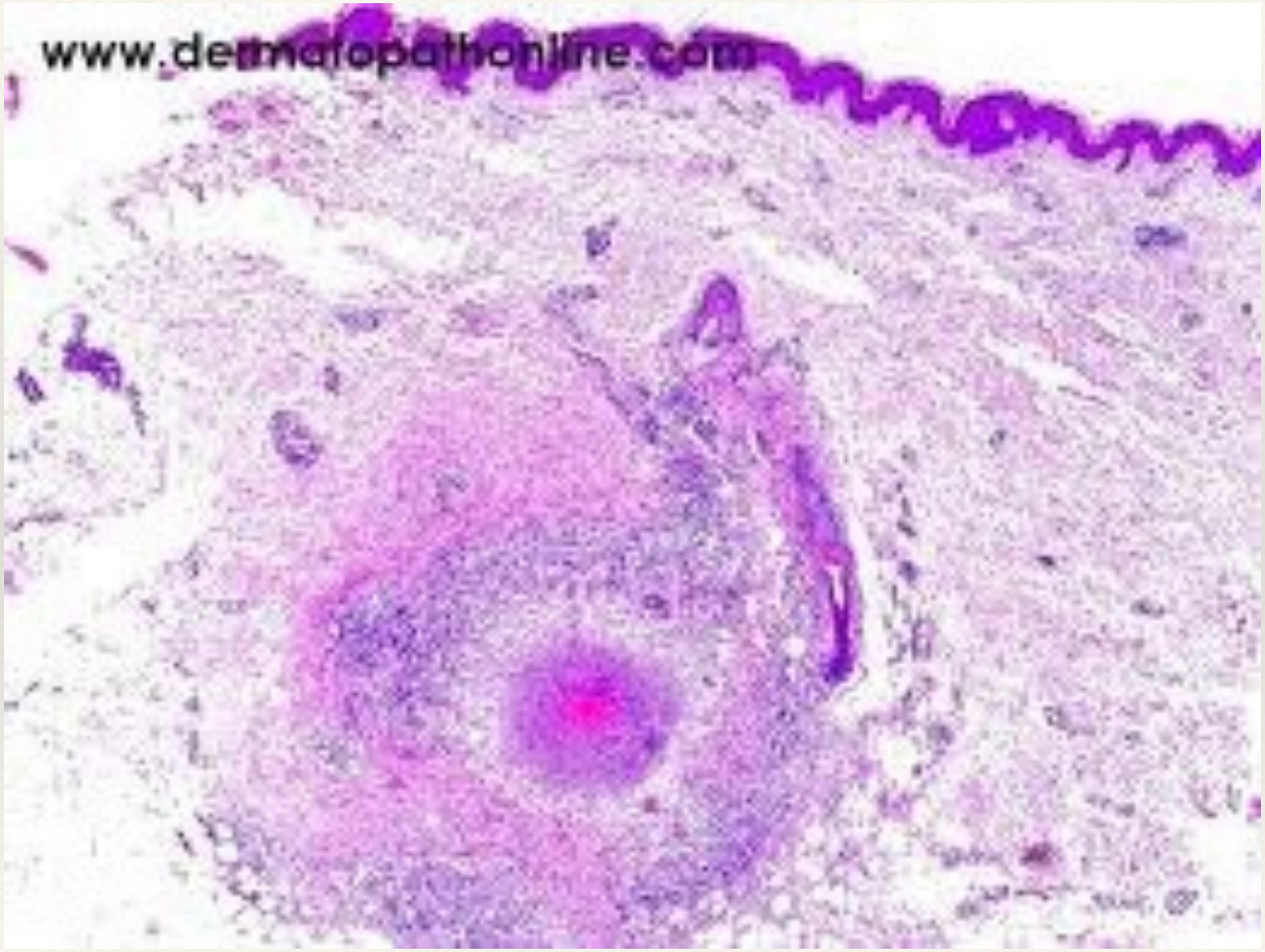


# Cutaneous polyarteritis nodosa

- Histopathology:
  - nodular arteritis + polymorphnuclear infiltrates involving medium sized arteries of the deep reticular dermis and subcutaneous tissue + extensive fibrinoid necrosis ( this is contrast to classical PAN  which rarely shows nodular arteritis and the picture is of small vessel leukocytoclastic vasculitis)

# Cutaneous polyarteritis nodosa



# Cutaneous polyarteritis nodosa

- Cutaneous PAN- has been associated with HBV & HCV infection, Crohn's disease, streptococcal infections, TB, and medications ( minocycline)
- Typically the only laboratory abnormality is ESR
- Most patients respond well to: aspirin, NSAIDs, prednisone, sulfapyridine, or methotrexate