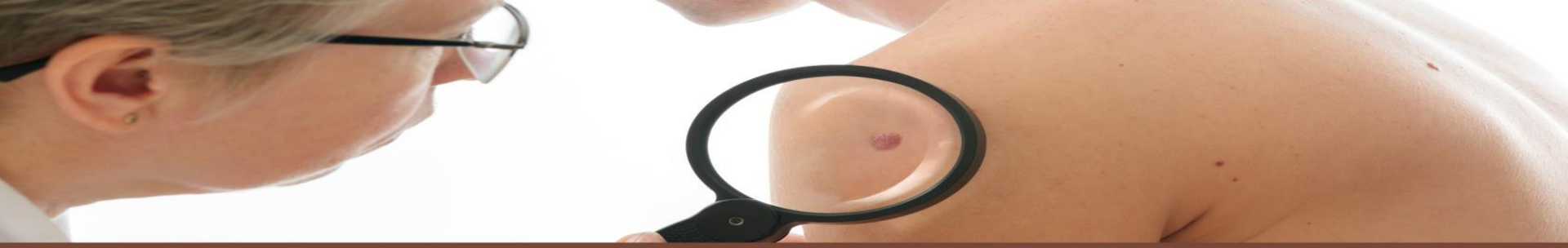

433 Teams

DERMATOLOGY

L9-Purpura and Vasculitis

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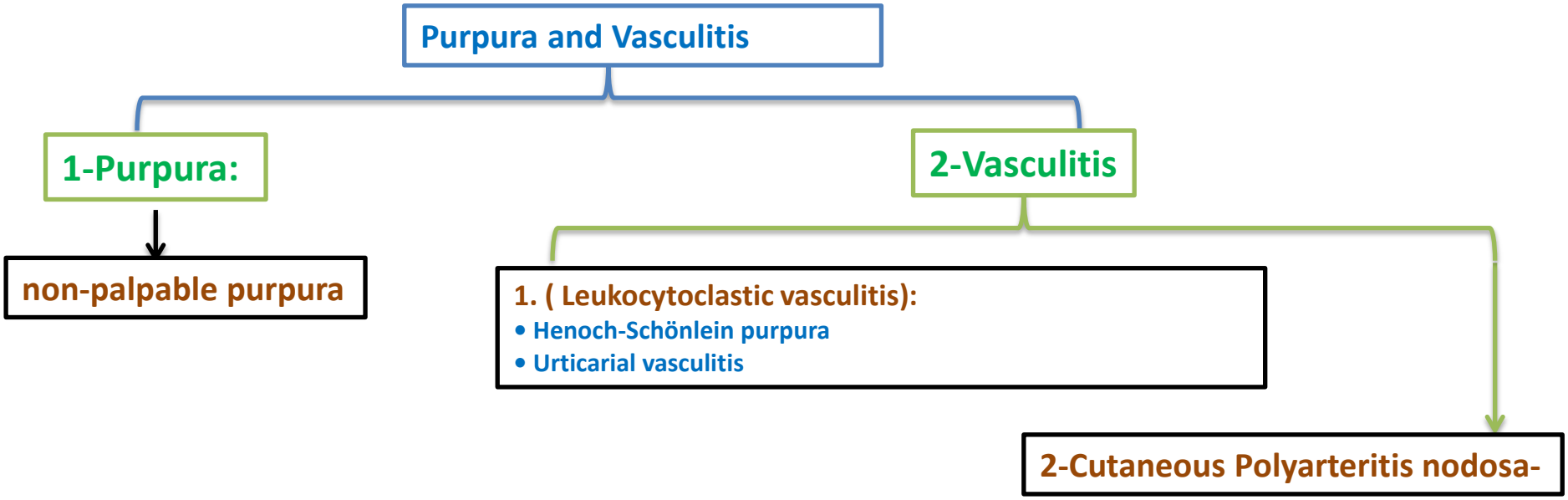
Objectives

- 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

Mind Map



1-Purpura:

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

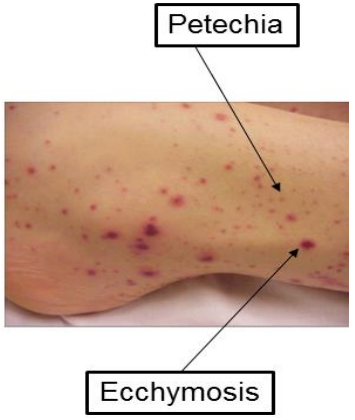
Purpura may be palpable or non-palpable

 # Non-palpable purpura are divided into 2 morphologies based on their size:

1- Petechiae- (< 3mm) superficial, pinhead-sized, hemorrhagic macules
2-Ecchymoses- (> 3 mm) irregularly shaped, bluish-purpulish patches " bruises"

The type of lesion usually indicates the underlying pathogenesis;

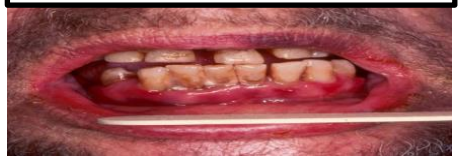
- **Non-palpable purpura is typically→ non-inflammatory**
- **Palpable purpura is usually a sign of → vascular inflammation →"hallmark lesion of leukocytoclastic vasculitis"**



#Causes of non-palpable purpura:

- ❖ Trauma
- ❖ Poor dermal support of blood vessels e.g. " topical or systemi steroid use"

hemorrhagic gingivitis



- ❖ Vascular dysfunction: aging, scurvy, Ehlers-Danlos syndrome

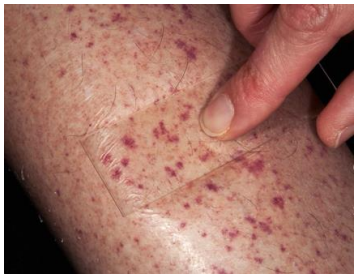
Vitamin C deficiency "Scurvy": (SAILORS)
- perifollicular petechiae
- keratotic plugging of hair follicles
- hemorrhagic gingivitis

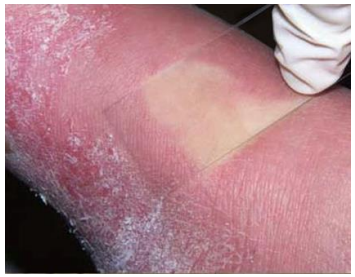
- ❖ Platelet dysfunction or Decreased Count: →
  - Idiopathic thrombocytopenic purpura
  - Thrombotic thrombocytopenic purpura
  - drug-induced thrombocytopenia
  - congenital/acquired platelet function defects

- ❖ Coagulopathies: hemophilia, cryoglobulinemia, anticoagulants, DIC, vitamin K deficincy, hepatic disease

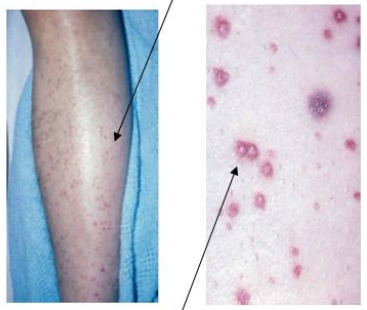
All forms of purpura do NOT blanch with pressure:

- ❖ 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)





Perifollicular petechiae



Keratotic plugging of hair follicles

How do we evaluate a patient with purpura?

- ❖ **History (Family hx , Drug hx & Medical hx)**
- ❖ **Physical examination (Size, Type ,Distribution & Mucous membranes)**
- ❖ **CBC & Differential**
- ❖ **Bleeding time**
- ❖ **PT & PTT**

2-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 OR 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:

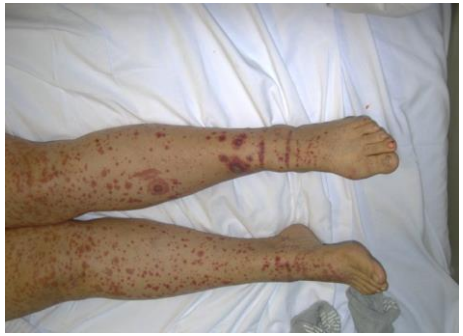
- Giant-cell arteritis
- Takayasu arteritis

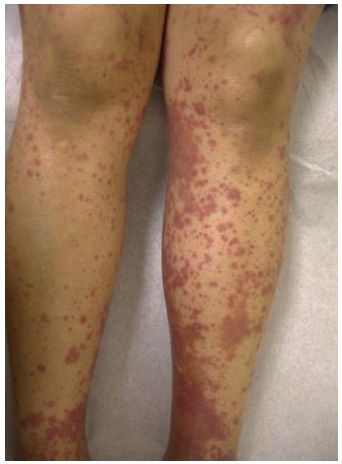
1. Cutaneous small vessels (Leukocytoclastic vasculitis):

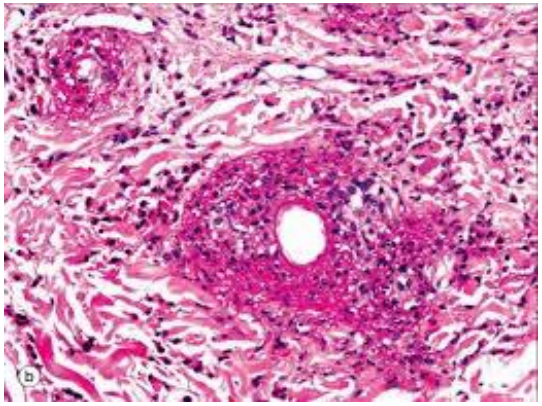
- Could occur as a primary process or could be secondary to an underlyingcause
- The majority of cases follow an acute infection or exposure to a newmedication
- Palpable purpura is the hallmark of this disease
- pinpoint to- several mm in diameter
- They predominate on the ankles and lower legs, affecting mainly dependent areas
- They resolve within 3-4 weeks with residual post-inflammatory Hyperpigmentation

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







1. Cutaneous small vessels (Leukocytoclastic vasculitis):

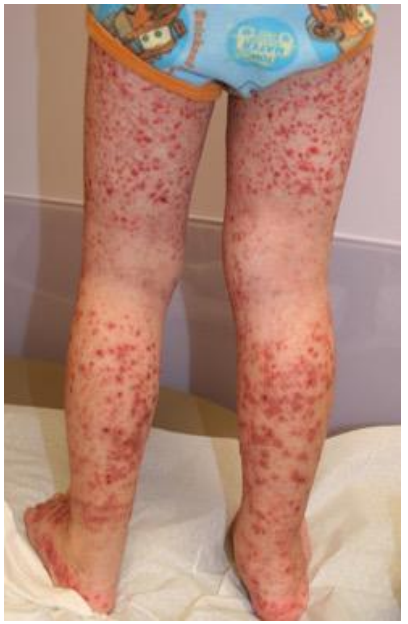
A- Henoch-Schönlein Purpura

- 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 (characteristic)
- 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



Course of the disease & possible complications:

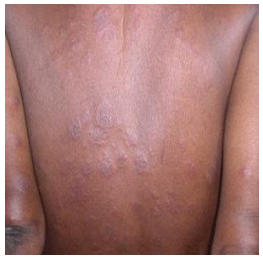
- The duration of the illness is 6-16 weeks
- In most patients the disease usually resolves without sequelae
- 5-10 % of patients will have persistent or recurrent disease
- Arthalgias may progress to arthritis producing periarticular swelling
- around the knees and ankles
- GI bleeding, acute surgical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

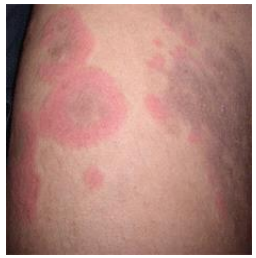
Treatment :

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

1. Cutaneous small vessels (Leukocytoclastic vasculitis):

B-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 is an eruption of erythematous wheals that clinically resemble urticaria but histologically show changes of leukocytoclastic vasculitis.**
- **Urticarial vasculitis may be divided into normocomplementemic and hypocomplementemic variants.**
- **The hypocomplementemic form more often is associated with systemic symptoms and has been linked to connective-tissue disease (ie, systemic lupus erythematosus [SLE]).**

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

Normal complement levels:

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

Low complement levels:

- **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)

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)

Medium vessels:
Polyarteritis nodosa-→ Cutaneous

Cutaneous polyarteritis nodosa

- Necrotizing vasculitis affecting small and- mediumsized 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

The Manifestations of 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

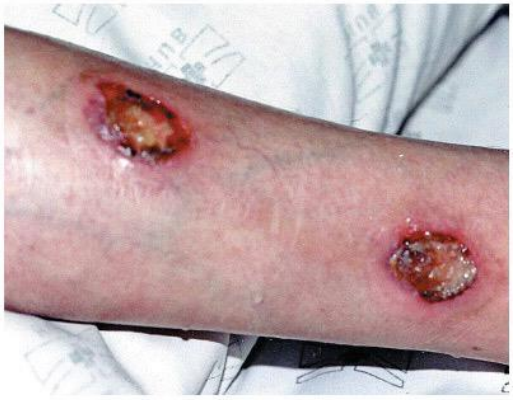
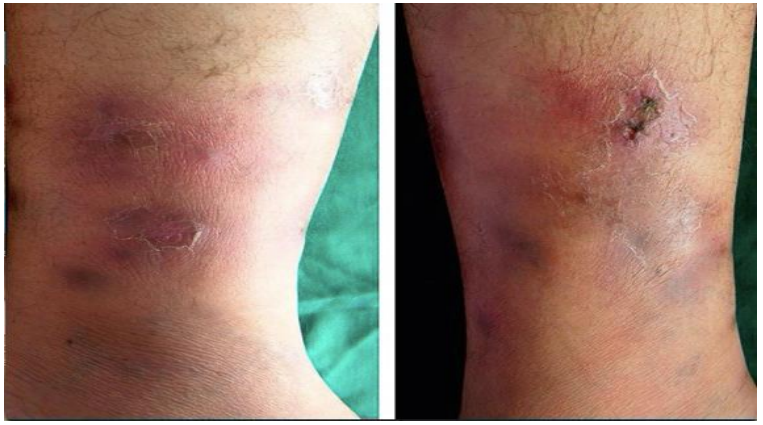
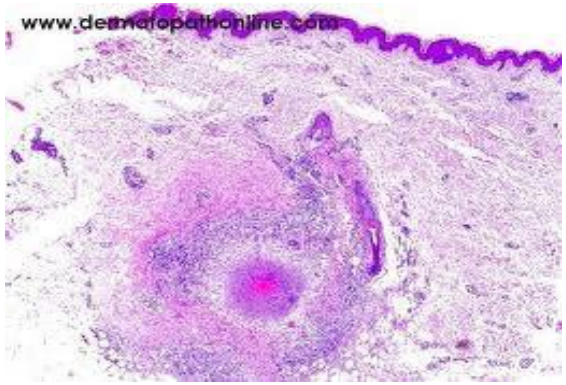


FIGURE 2: Ulcers in lower limbs



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 ofsmall vessel leukocytoclastic vasculitis)



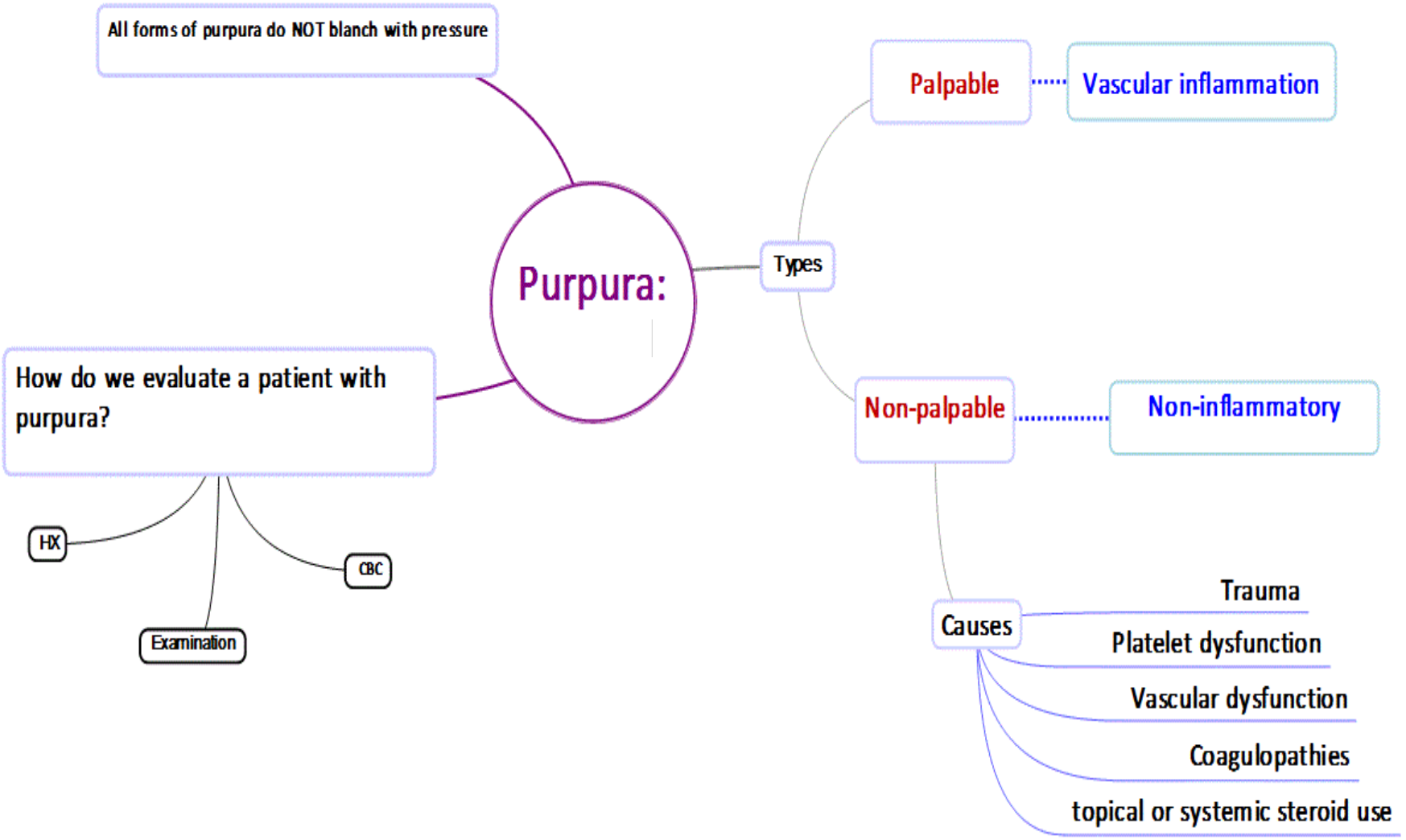
- **Cutaneous PAN- has been associated with HBV & HCV infection, Crohn's disease, streptococcal infections, TB, and medications (minocycline)**

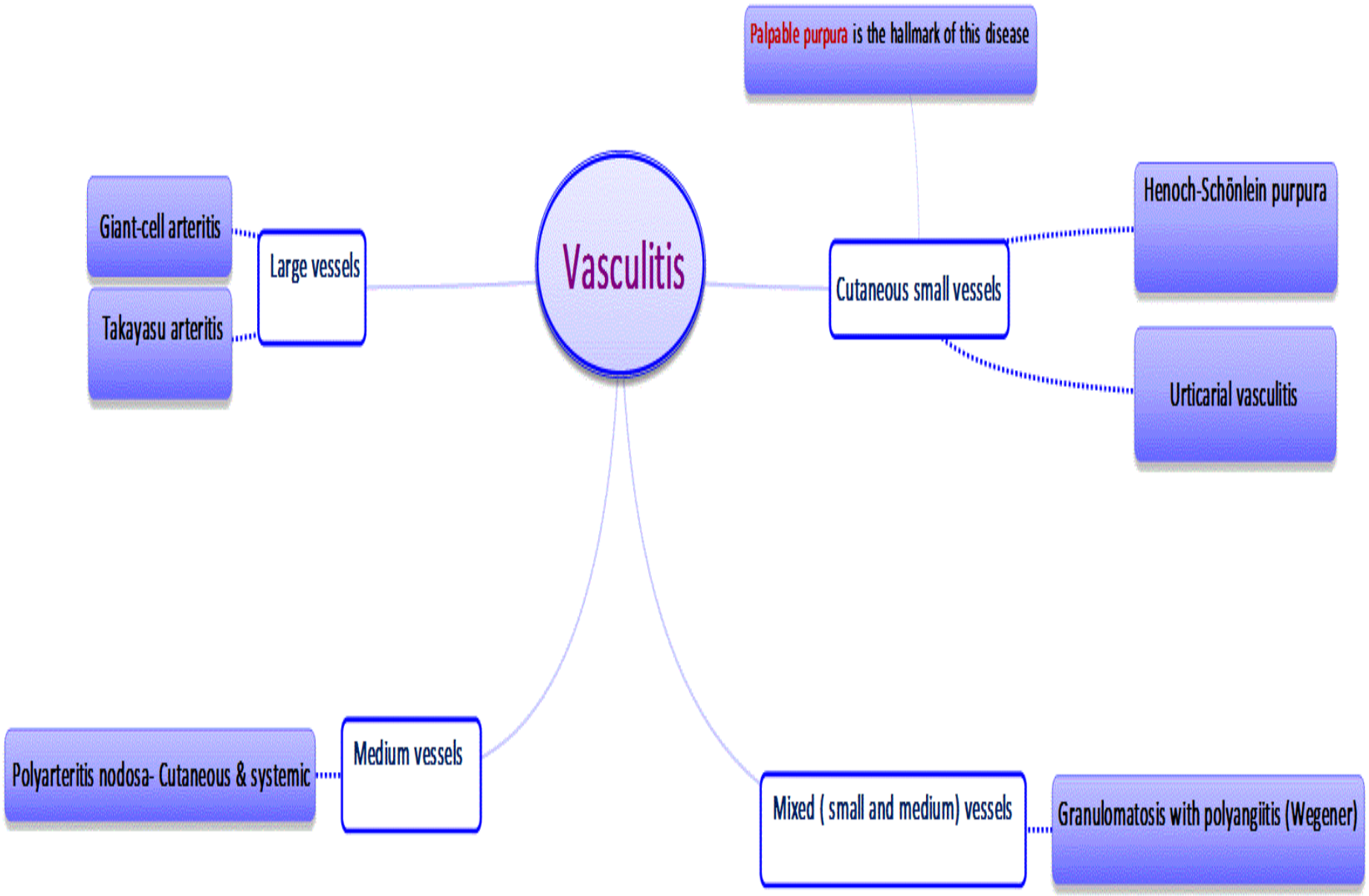
- **Typically the only laboratory abnormality is ESR**

- **Treatment→ Most patients respond well to: aspirin, NSAIDs, prednisone, sulfapyridine, or methotrexate**

PAN= Polyarteritis nodosa

SUMMARY


All forms of purpura do NOT blanch with pressure
Purpura:
Types
Palpable
Vascular inflammation
Non-palpable
Non-inflammatory
Causes
Trauma
Platelet dysfunction
Vascular dysfunction
Coagulopathies
topical or systemic steroid use
How do we evaluate a patient with purpura?
HX
CBC
Examination


Vasculitis
Large vessels
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