

# Cutaneous Manifestations of *Vasculitis and the Differential Diagnosis*

S.Zahra Ghodsi  
Prof of Dermatology  
Rheumatology Research Center  
Shariati Hospital  
Tehran University of Medical Sciences

TABLE I.—*Chapel Hill Consensus Conference nomenclature of vasculitides 2012.*

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Nomenclature of vasculitides

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**Large vessel vasculitis (LVV)**

- Takayasu arteritis (TAK)
- Giant cell arteritis (GCA)

**Medium vessel vasculitis (MVV)**

- Polyarteritis nodosa (PAN)
- Kawasaki disease (KD)

**Small vessel vasculitis (SVV)**

- Antineutrophil cytoplasm antibody (ANCA)-associated vasculitis
  - Microscopic polyangiitis (MPA)
  - Granulomatosis with polyangiitis (Wegener's) (GPA)
  - Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) (EGPA)
- Immune complex SVV
  - Anti-glomerular basement membrane (anti-GBM) disease
  - Cryoglobulinemic vasculitis (CV)
  - IgA vasculitis (Henoch-Schönlein) (IgAV)
  - Hypocomplementemic urticarial vasculitis (HUV) (anti-C1q vasculitis)

**Variable vessel vasculitis (VVV)**

- Behçet's disease (BD)
- Cogan's Syndrome (CS)

**Single-organ vasculitis (SOV)**

- Cutaneous leukocytoclastic angiitis
- Cutaneous arteritis
- Primary central nervous system vasculitis
- Isolated aortitis
- Others

**Vasculitis associated with systemic disease**

- Lupus vasculitis
- Rheumatoid vasculitis
- Sarcoid vasculitis
- Others

**Vasculitis associated with probable etiology**

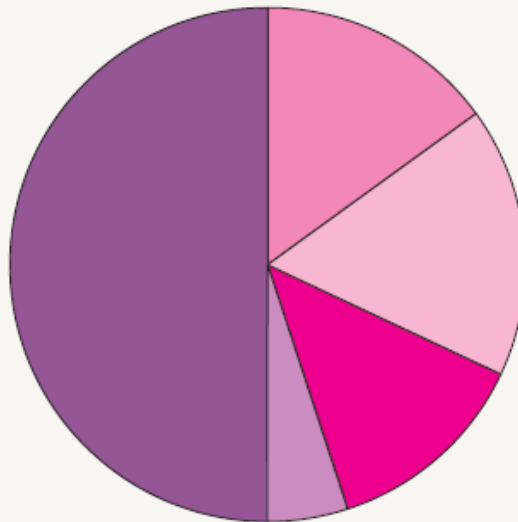
- Hepatitis C virus-associated cryoglobulinemic vasculitis
- Hepatitis B virus-associated vasculitis
- Syphilis-associated aortitis
- Drug-associated immune complex vasculitis
- Drug-associated ANCA-associated vasculitis
- Cancer-associated vasculitis
- Others

- ▶ The majority of cutaneous vasculitides involve small vessels (SVV), particularly venules.
- ▶ leukocytoclastic vasculitis (LCV)

Dermatologist



### ETIOLOGIES OF CUTANEOUS SMALL VESSEL VASCULITIS



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

Rheumatologist



① *What kind of skin lesion leads you to diagnosis vasculitis?*

SVV

- ▶ Palpable Purpura (most common)
- ▶ Petechiae, Macular purpura



Typically develop in crops

Dependent areas

± pruritus, pain, or burning sensation

Round, red-purple papules and plaques ± inflammation

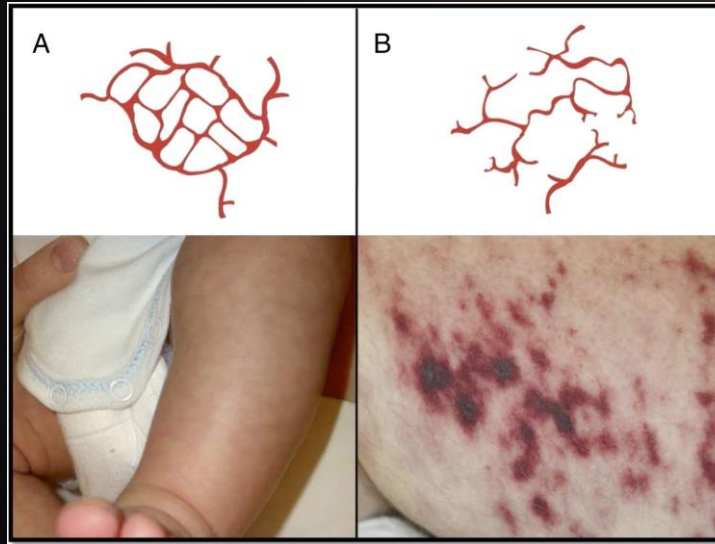


- ▶ Urticarial papules
- ▶ Vesicles
- ▶ Pustules
- ▶ Targetoid papules and plaques



# Livedo racemosa

**MVV**



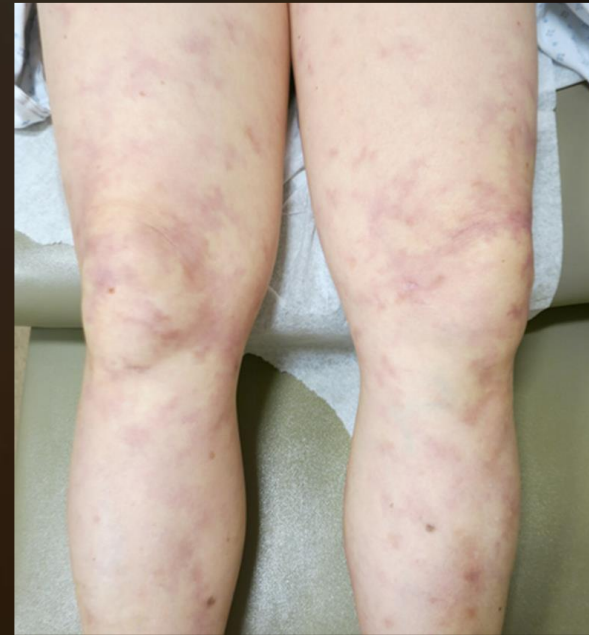
## Retiform purpura



► Ulcers



► Subcutaneous nodules



► Digital necrosis



| Caliber of the predominantly affected vessel | Classification                                        | Subclassification or etiologies                                                                                                                                                                                                                                                                                                                                                                                                  | Morphology of cutaneous lesions                                                                                                         |
|----------------------------------------------|-------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------|
| <b>Small</b>                                 | Cutaneous small vessel vasculitis ( <b>CSVV</b> )     | <ul style="list-style-type: none"> <li>-Henoch-Schonlein purpura</li> <li>-Acute hemorrhagic edema of infancy</li> <li>-Urticarial vasculitis</li> <li>-Cryoglobulinemia Types II and III</li> <li>-Erythema elevatum diutinum</li> <li>-Secondary causes of CSVV: <ul style="list-style-type: none"> <li>AI-CTD</li> <li>Drug exposure</li> <li>Infections</li> <li>Malignancies, most often hematologic</li> </ul> </li> </ul> | <b>Palpable purpura</b><br>Petechieae<br>Macular purpura<br>Urticarial papules<br>Vesicles<br>Pustules<br>Targetoid papules and plaques |
| <b>Small and medium-sized</b><br>("mixed")   | <b>ANCA-associated</b><br><br><b>Secondary causes</b> | Microscopic polyangiitis<br>Wegener's granulomatosis<br>Churg-Strauss syndrome<br><br>Infections<br>Inflammatory disorders (e.g. AI-CTD)                                                                                                                                                                                                                                                                                         | Petechieae<br>Palpable purpura<br>Livedo racemosa<br>Retiform purpura<br>Ulcers<br>Subcutaneous nodules<br>Digital necrosis             |
| <b>Medium-sized</b>                          | Polyarteritis nodosa ( <b>PAN</b> )                   | Classic (systemic) PAN<br>Cutaneous PAN                                                                                                                                                                                                                                                                                                                                                                                          | Livedo racemosa<br>Retiform purpura<br>Ulcers<br>Subcutaneous nodules<br>Digital necrosis                                               |
| Large                                        | Temporal arteritis<br><br>Takayasu's                  |                                                                                                                                                                                                                                                                                                                                                                                                                                  | Cutaneous manifestations are rare                                                                                                       |

② *In patients with vasculitis which kind of skin lesions can be related to underlying disease?*

# SLE

- ▶ Vasculitis 11%
- ▶ cutaneous lesions the main clinical presentation, 89%
- ▶ Most SVV
- ▶ but a subset of MVV (nerve and visceral)
- ▶ Ulceration
- ▶ higher lupus activity scores.



# SJS

- ▶ Patients with Sjögren syndrome who develop cutaneous vasculitis frequently have circulating cryoglobulins
- ▶ xerosis



# Dermatomyositis

- ▶ a **rare** manifestation of DM
- ▶ most often in juvenile-onset
- ▶ ↑ **malignancy and poor outcomes**
- ▶ in 9.2% of patients with **MVV**, such as sc nodules, periungual infarcts, and digital ulceration



# Dermatomyositis



# Rheumatoid Arthritis

- ▶ Rheumatoid vasculitis is a severe and uncommon presentation of longstanding ( >10 years), poorly controlled RA with significant morbidity and mortality.
- ▶ MVV closely resemble PAN
- ▶ ANCA-associated vasculitis



# Systemic Sclerosis

- ▶ Vasculitis has **rarely** been associated with SS
- ▶ Most frequently described as being p-ANCA/ MPO positive.
- ▶ rapidly progressive glomerulonephritis, alveolar hemorrhage, limb ischemia, and vasculitis of the skin

③ *What are the differential diagnosis?*

# *Pigmented Purpuric Dermatoses*

- ▶ Clustered petechial hemorrhage
- ▶ Often a background of yellow–brown discoloration due to hemosiderin deposition
- ▶ Lower limbs



14%

calcium channel antagonists,  
beta-blockers,  
angiotensin-converting enzyme inhibitors,  
furosemide and other diuretics,  
antihistamines,  
antidepressants,  
chlordiazepoxide,  
carbamazepine, paracetamol and NSAIDs,  
glipizide,  
bezafibrate,  
medroxyprogesterone acetate,  
raloxifene,  
pseudoephedrine,  
vitamin B derivatives,  
interferon- $\alpha$  (in hepatitis C infection),  
antibiotics including ampicillin and cotrimoxazole,

# Cryoglobulinemia

- ▶ Cryoglobulins cause disease via two mechanisms:

- 1- **Occlusion** (type I)

monoclonal, due to an underlying plasma cell dyscrasia or lymphoproliferative disorder



# *Cryoglobulinemia*

- ▶ 2- **Immune complex-mediated** vasculitis (types II, III)  
mixed cryoglobulinemia , often due to hepatitis C

Palpable purpura

# Cholesterol Embolus

men  $\geq 50$  usually with DM, HPL, tobacco use, and peripheral artery disease.

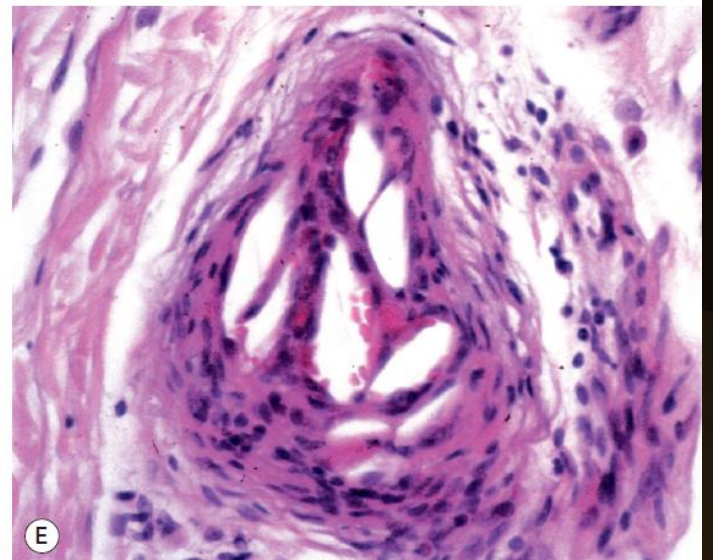
❖ Promotion settings:

1- *arterial or coronary catheterization*, within hours to days of catheterization.

2- *prolonged anticoagulation*, typically occurs after 1 to 2 months of therapy.

“warfarin blue toe syndrome”

3- *acute thrombolytic therapy* given for myocardial infarction or stroke, hours to days of lytic therapy.



# *Antiphospholipid Antibody syndroms*



# *Livedoid Vasculopathy*

Atrophie blanche ,Livedoid vasculitis

- ▶ Favors the distal lower extremities, especially the **ankles**
- ▶ **Female** predominance
- ▶ **Painful**, punched-out ulcers
- ▶ White, round or stellate **scars** with peripheral telangiectasias
- ▶ Reticulated violaceous erythema may extend from ulcer
- ▶ **Unknown** pathogenesis, an **alteration in** local or systemic control of **coagulation** with formation of fibrin thrombi focally within superficial dermal blood vessels.

# *Livedoid Vasculopathy*



# *Malignant Atrophic Papulosis*

## Degos disease

- ▶ systemic and benign (skin-limited)
- ▶ SLE, APLs, Dermatomyositis
- ▶ a rare vaso-occlusive disorder that predominantly affects the skin, GI tract, and CNS.



## *Intravascular Cellular Occlusion*



**Fig. 23.12** Intravascular B-cell lymphoma. The clinical presentation in this

# *erythema ab igne*



- ▶ **Skin biopsy** is **mandatory** in cutaneous vasculitis to confirm the diagnosis and to differentiate it from other non-vasculitic disorders which usually present the same clinical pictures.
- ▶ Biopsy **location**, **depth** and **timing** are three key factors to have a diagnostic picture.

# Time

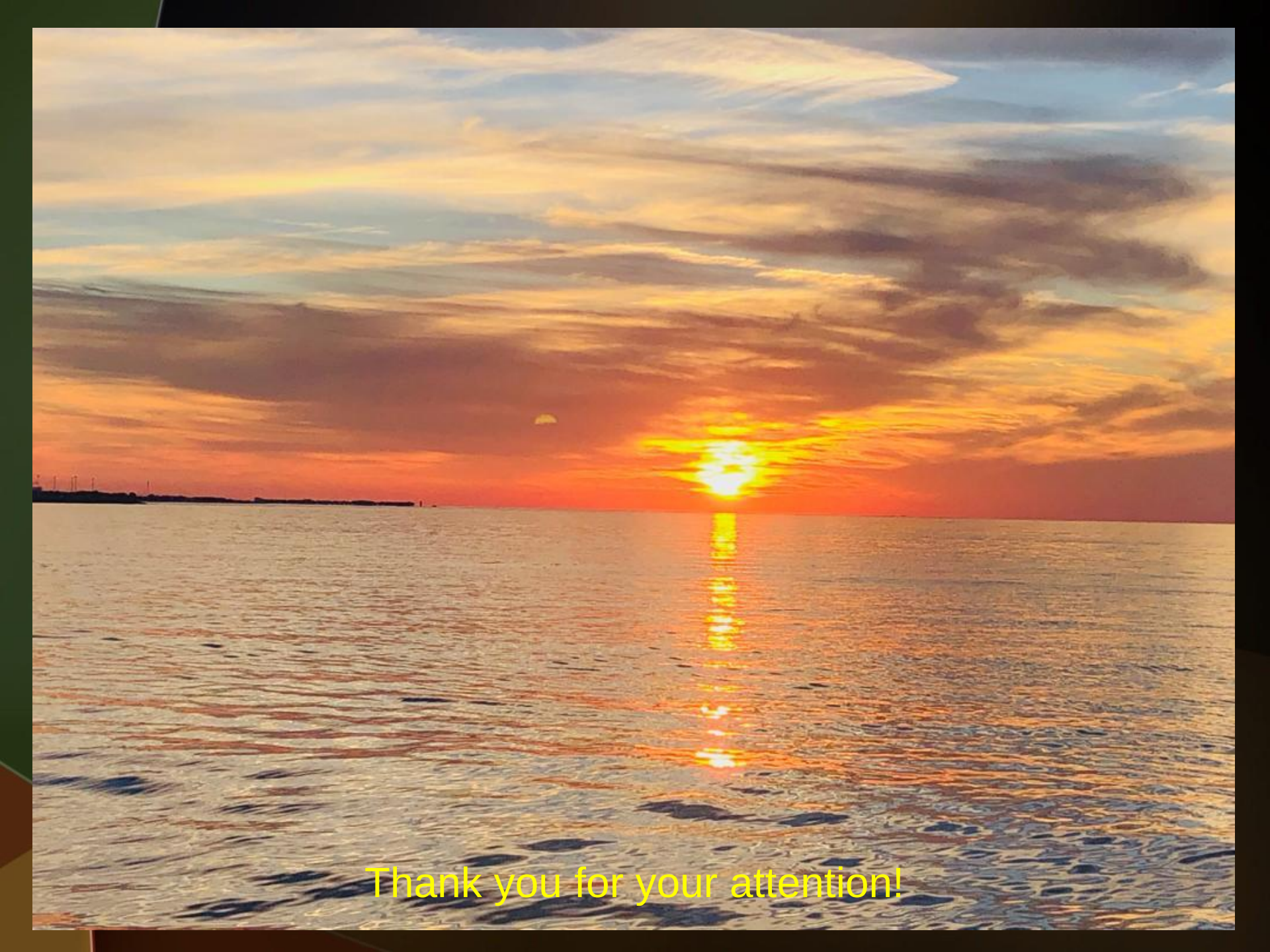
- ▶ Biopsy with DIF should be performed on a fresh lesion, roughly 24-48 hours old.
- ▶ A biopsy, performed too early or too late may be non diagnostic.

## **H&E**

- Granulomatous vasculitis: systemic
- Lymphocytic vasculitis: CTD, viral infection, drug
- Small- and medium-vessel involvement:  
ANCA or CTD
- Urticarial vasculitis with dermal interstitial  
neutrophilic infiltrate: hypocomplementemic  
urticarial vasculitis (consider SLE)
- HSP without eosinophils  
in patients >40 years old: renal involvement

## **DIF**

- IgA predominant: HSP
- Prominent IgM: cryoglobulin or rheumatoid  
vasculitis
- Positive around vessels and BMZ:  
hypocomplementemic urticarial vasculitis  
(consider SLE)
- Negative: pauci-immune vasculitis  
(ie, ANCA-associated)

A photograph of a sunset over a body of water. The sun is a bright, glowing orb on the horizon, casting a long, shimmering reflection down the center of the water. The sky is filled with layers of clouds, some dark and dramatic, others lighter and catching the orange and yellow light of the setting sun. The water in the foreground is textured with small ripples, reflecting the colors of the sky. In the distance, a dark silhouette of a shoreline with some structures is visible on the left.

Thank you for your attention!