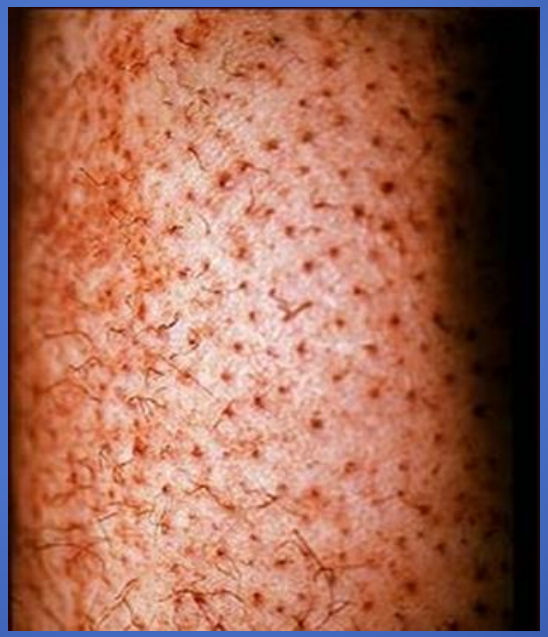
A woman with Systemic Lupus and a new rash



Farhan Tahir MD, FACR
Rheumatic Disease Associates
Willow Grove, PA

Objectives

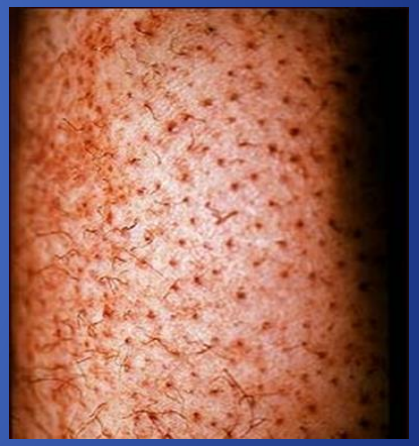
- Case review
- Purpura : causes and types
- Many faces of Purpuric rash
- Purpura from a Rheumatologist's eyes
- Discussion
- Conclusion

Pertinent points in HPI

- Rash-Petechial purpura and ecchymosis (no prior history)
- Chronicity: over 3 months, progressive
- Preceding events: no infectious, traumatic or chemical exposure, no herbal supplement exposure, no new drug
- Systemic features: dyspnea and fatigue, no fever, diarrhea, hemoptysis
- Potential culprit medications: ASA, Prednisone ,Cellcept, Gabapentin
- Nutrition: Poor (peanut butter sandwich, no multivitamins)
- Co morbid conditions: SLE, chronic steroid use, osteoporosis

Pertinent history & exam

Med/Surg hx: Mitral valve prolapse, Hip replacement and partial hysterectomy (no history of excessive bleeding)



Family: no bleeding diathesis

Social: lives alone, no drugs or alcohol abuse

Exam: Poor dental hygiene, tender bruising over thighs, petechial lesions, 1+peripheral edema, normal pulses in feet

Purpura

- **Purpura** (from Latin: *purpura*, meaning "purple")
  - Bleeding under skin or into mucosal membranes
  - Pinpoint area, < 2mm: petechiae
  - Larger confluent lesions: ecchymosis (bruises)
- **Causes of Purpura**
  - Disruption of vascular integrity
  - Primary or secondary hemostatic abnormality

Factors influencing hemostasis

Destruction of vascular wall

- Trauma, inflammation or infection

Impaired platelet plug and fibrin clot

- Injury-> vasoconstriction & retraction->platelet recognition vessel endothelial adhesion ->platelet granules->platelet plug
- Tissue factor-VII complex->activation of coagulation cascade->Fibrin cross links->clot

Impaired collagen synthesis

- Disordered collagen and connective tissue synthesis and structure, congenital versus acquired

When Purpura is vasculitis

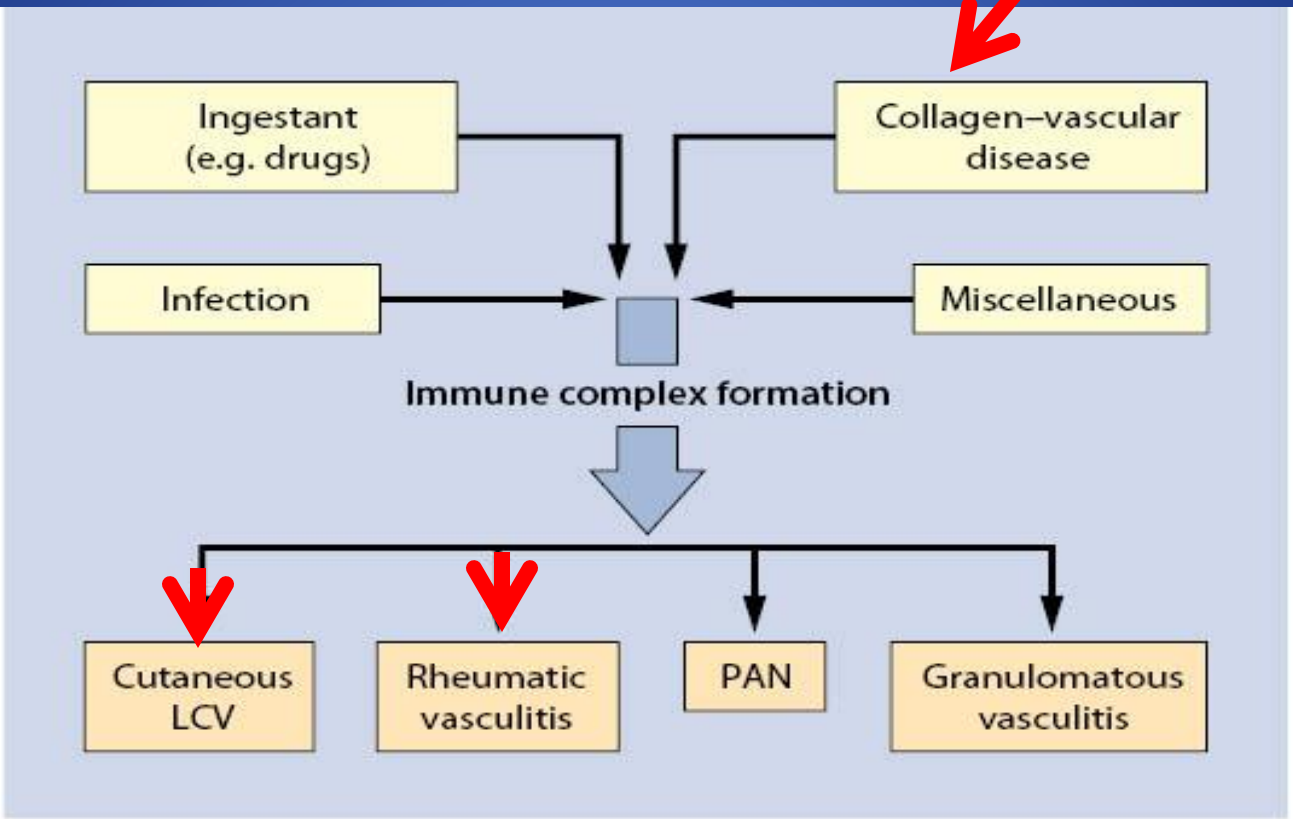
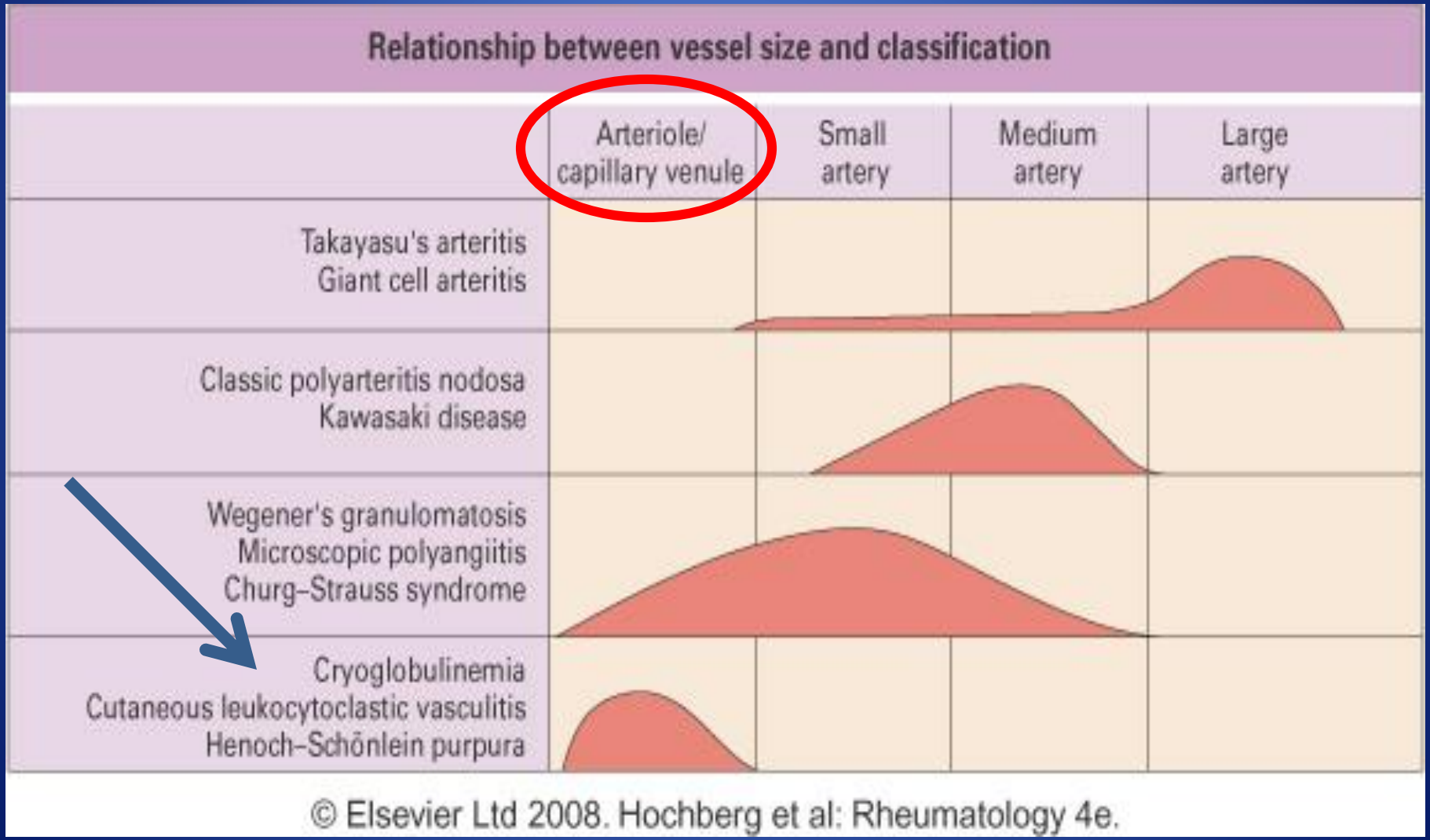
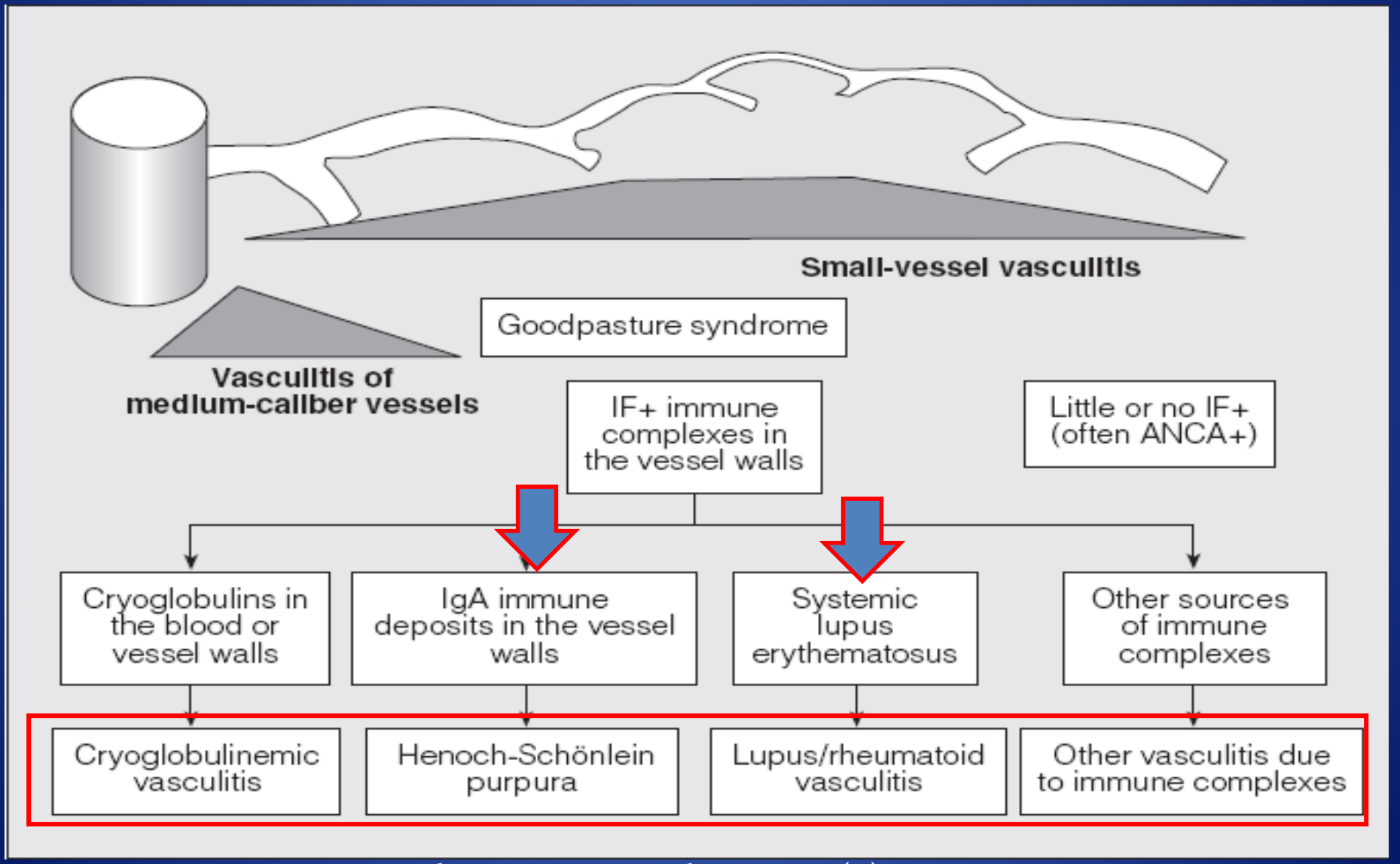


Figure 4–1 Schematic representation of multiple antigenic exposures leading, through the mechanisms of immune complex formation and deposition, to the varying manifestations of vasculitis. LCV = leukocytoclastic vasculitis; PAN = polyarteritis nodosa.

Small vessel vasculitis as purpura



Small vessel vasculitis



Arch Bronconeumol. 2008;44(8):428-36

Is this "Petechial rash" a form of vasculitis

- Palpable Purpura is usually inflammatory or vasculitis
  - Hypersensitivity vasculitis
  - Henoch-Scolein purpura
  - SLE, RA and small vessel vasculitis (ANCA)
  - Infectious
  - Polyarteritis Nodosa
  - Pseudovasculitis

Or it is non inflammatory petchiae?

- Non palpable purpura : Non vasculitic

Corticosteroid use
Idiopathic thrombocytopenic purpura
Thrombotic thrombocytopenic purpura
Disseminated intravascular coagulation
Vitamin K deficiency
Scurvy

Ecchymosis : second concern

- Platelet disorders (quantitative) :
  Thrombocytopenia
- Platelet disorders (functional):
  Glycoprotein disorders
- Hemophilia
- Factor inhibitors
- Leukemia
- Disseminated intravascular coagulation
- Vitamin K Deficiency
- Scurvy

Lab data

Hematology

WBC: 3.0, HGB: 10.5, platelets: 235 MCV: 82

Comprehensive metabolic panel

CR: 0.8 Ca:9.7, AST: 35, ALT:40, Albumin of 2.9

Cagulation profile

PT11.6/INR 1.1, PTT 21,
Factor Vll, lX X and Von WIllibrand Factor assays-normal,
Bleeding time- 6.3min(2.3-9.5min),

Rheumatologic profile

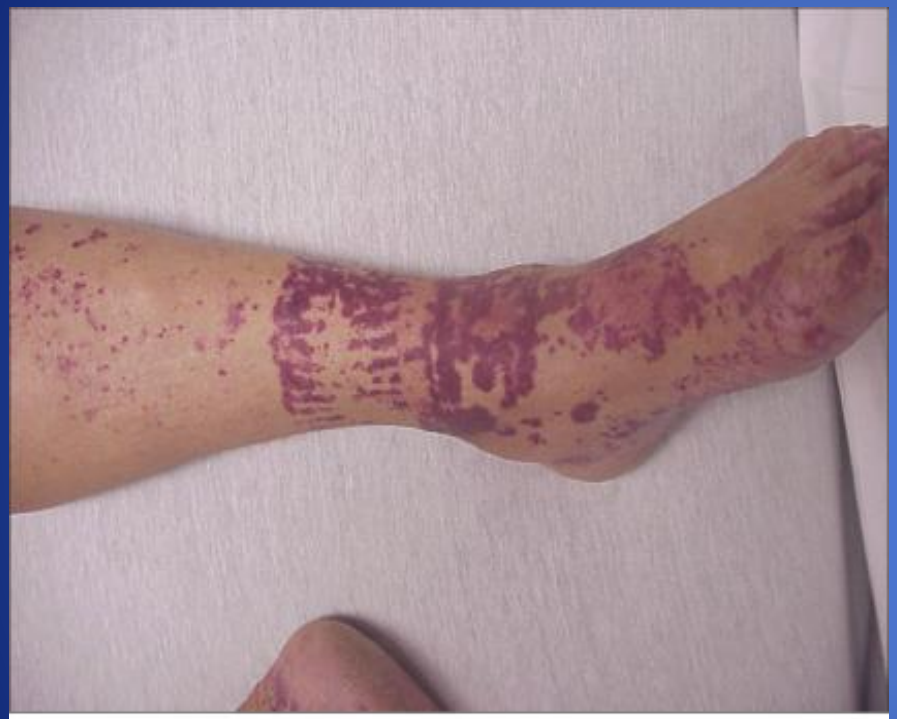
ESR 25, DSDNA:NEG, , C3: 167 (nl), C4:18(nl)
ANCA: NEG , PR3:NEG, Myeloperoxidase Ab: neg

Leukemic, malignancy

Normal wbc profile, SPEP: NEG, UPEP: NEG.

Questions to ask ourselves

What is the cause of this patient's purpura
What is the cause of this patient's ecchymosis
Is it inflammatory or vasculitis
Is it non vasculitic?



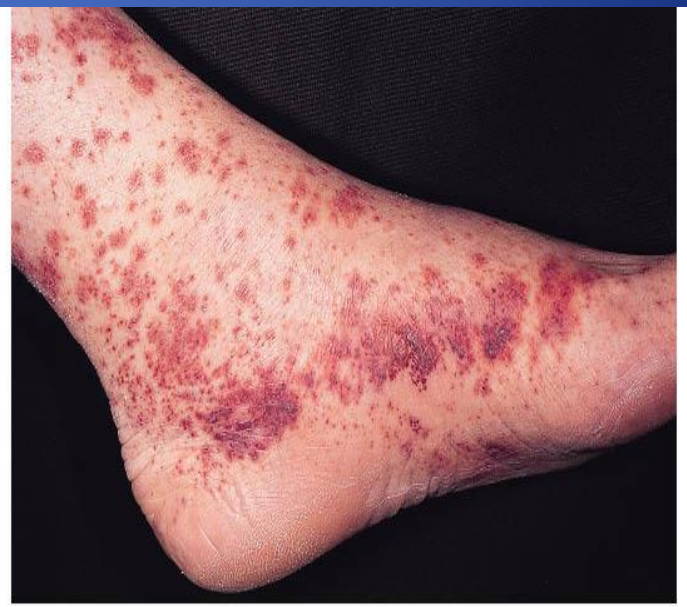


Figure 4–2 Typical palpable purpuric lesions seen in a patient with hypersensitivity vasculitis/small-vessel vasculitis.

Hypersensitivity vasculitis ?

[22.13.01] Traditional Diagnostic Approach for Hypersensitivity Vasculitis (ACR Criteria)

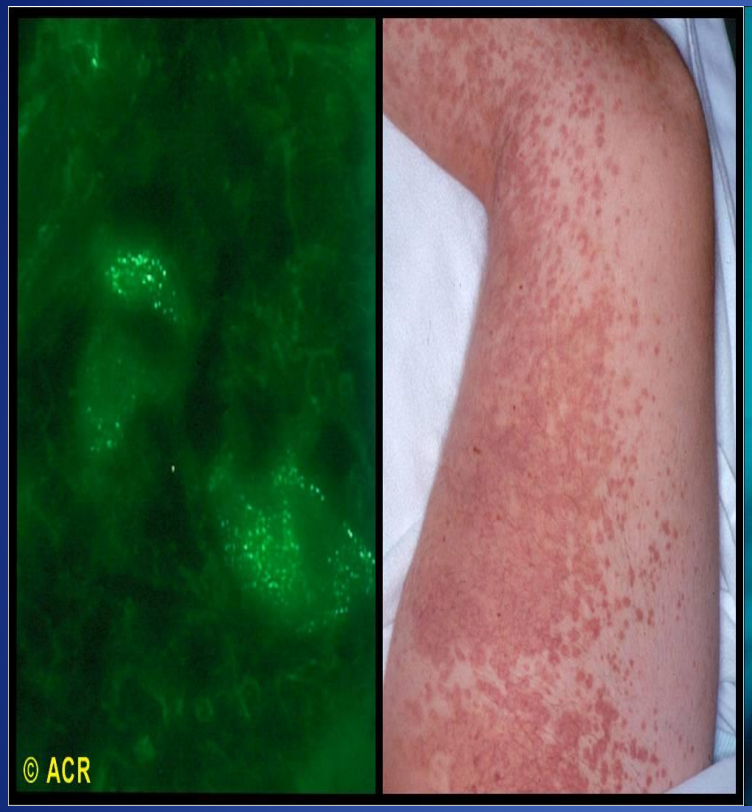
Purpose: To diagnose Hypersensitivity vasculitis by the presence of certain findings.

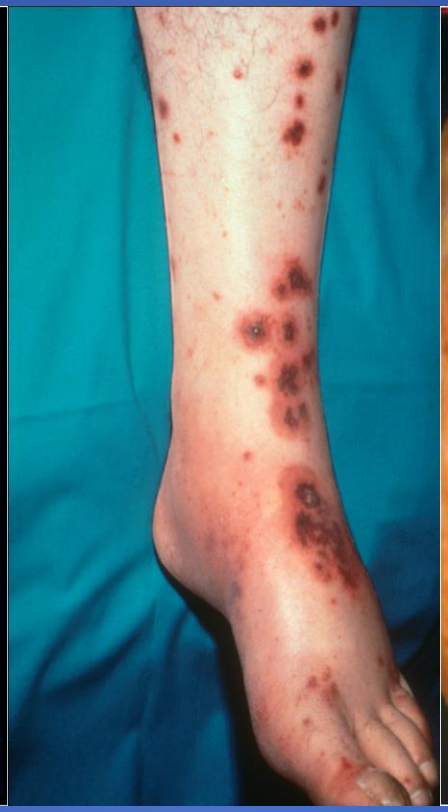
Age of patient in years at the time of onset	54 Years
Does the patient show palpable purpura, which do not blanch on pressure and which are unrelated to thrombocytopenia? (Y or N)	(•) Yes ( ) No
Does the patient show a maculopapular rash? (Y or N)	( ) Yes (•) No
Was a medication taken at the onset of symptoms? (Y or N)	( ) Yes (•) No
Does a biopsy show granulocytes in a perivascular or extravascular location, involving arterioles and/or venules? (Y or N)	( ) Yes (•) No

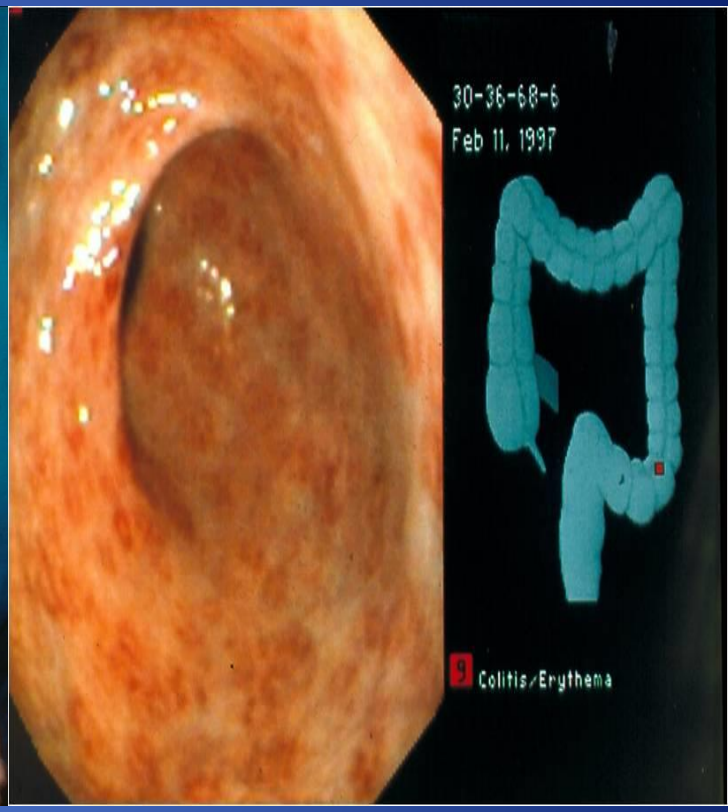
Please Click calculate if any changes are done.

Calculate Reset

Evaluation	Result
Data complete?	Yes
Number of factors present	2
Is hypersensitivity vasculitis likely present?	No







Henoch-Schönlein purpura ?

[22.14.01] Traditional Diagnostic Approach for Henoch-Schonlein Purpura (ACR Criteria)

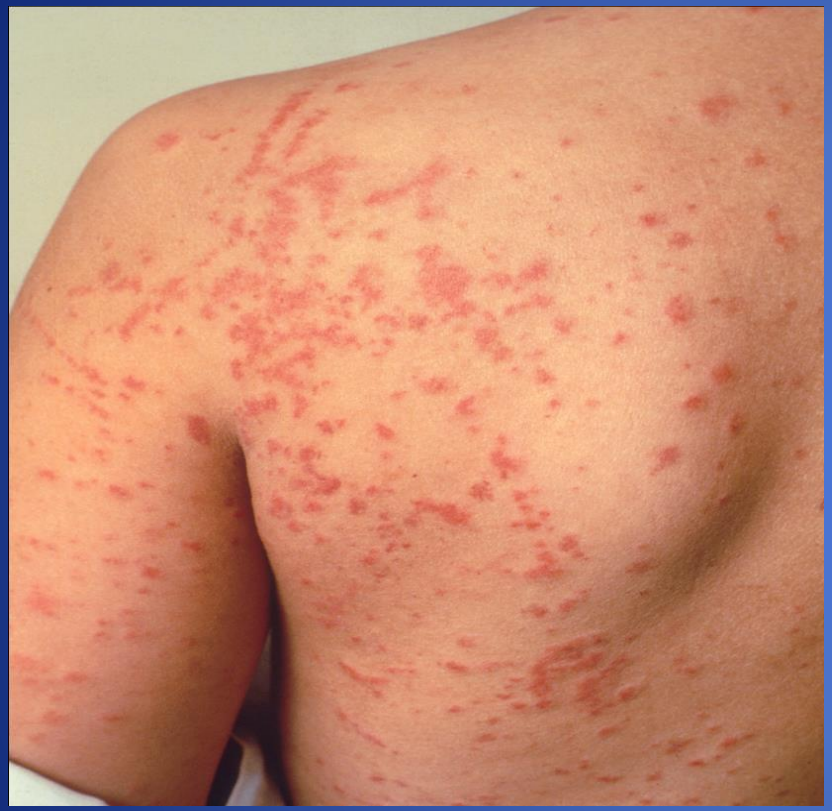
Purpose: To diagnose Henoch Schonlein Purpura by the presence of certain findings.

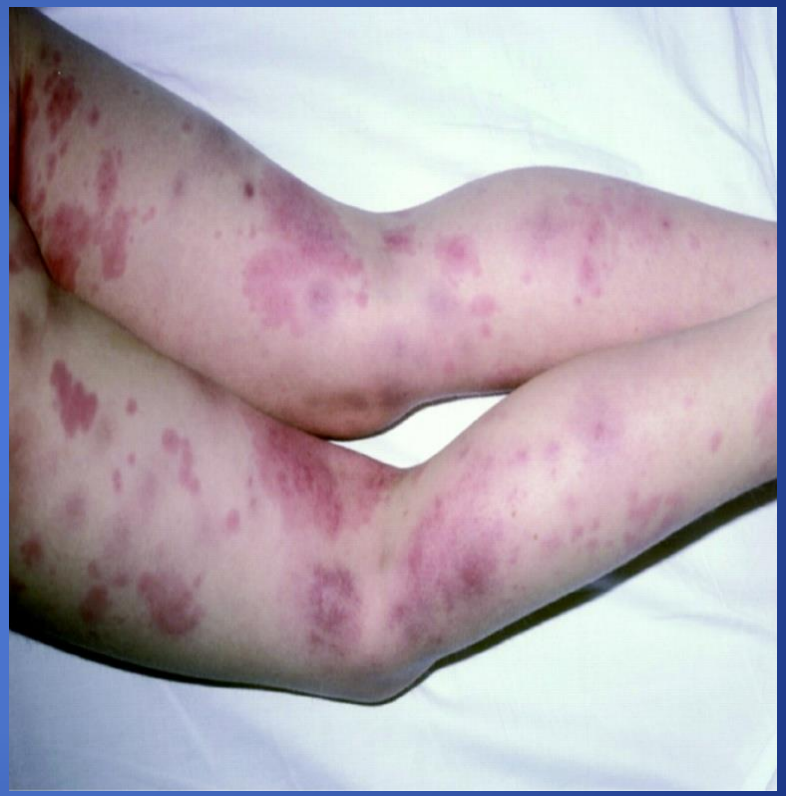
Age in years of patient at onset of first symptom	54 Years
Does the patient have slightly raised and palpable purpura? (Y or N)	(•) Yes ( ) No
Are the occurrence of purpura unrelated to thrombocytopenia? (Y or N)	(•) Yes ( ) No
Does the patient have diffuse abdominal pain, sometimes worse after meals? (Y or N)	( ) Yes (•) No
Does the patient have evidence of bowel ischemia, usually with bloody diarrhea? (Y or N)	( ) Yes (•) No
Are granulocytes seen on biopsy to be located in the walls of arterioles and/or venules? (Y or N)	( ) Yes (•) No

Please Click calculate if any changes are done.

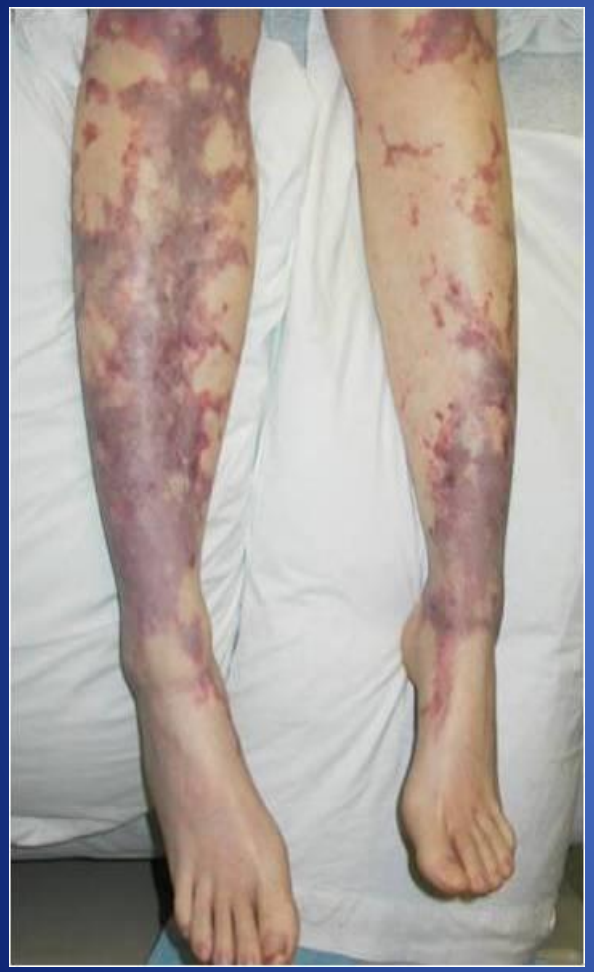
Calculate Reset

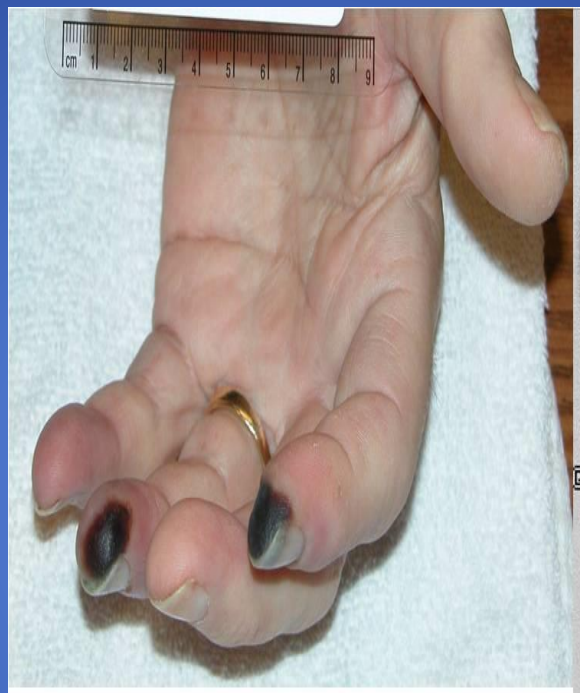
Evaluation	Result
Data complete?	Yes
Number of factors present	1
Is Henoch Schonlein Purpura likely present?	No

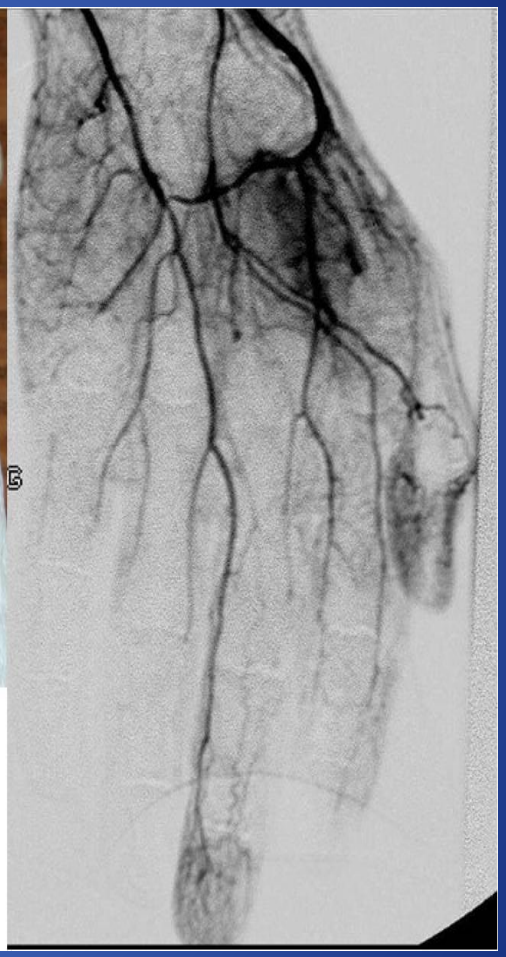




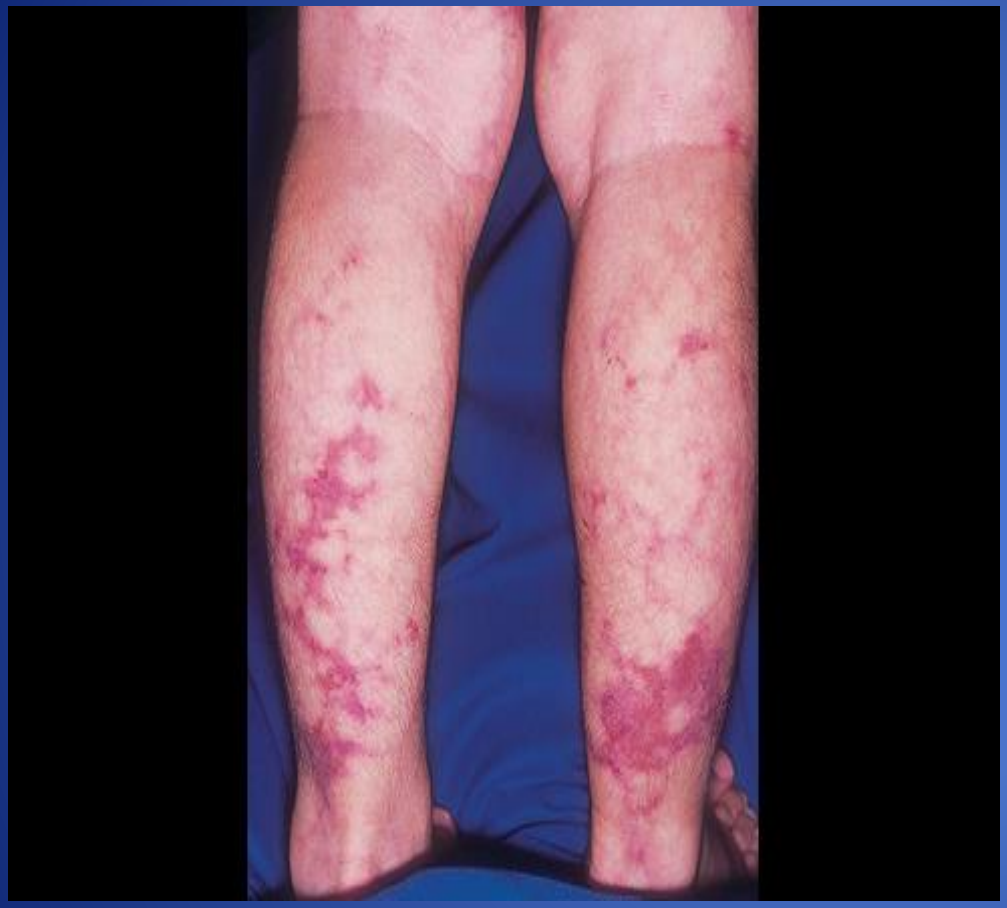
Urticarial vasculitis and purpura?

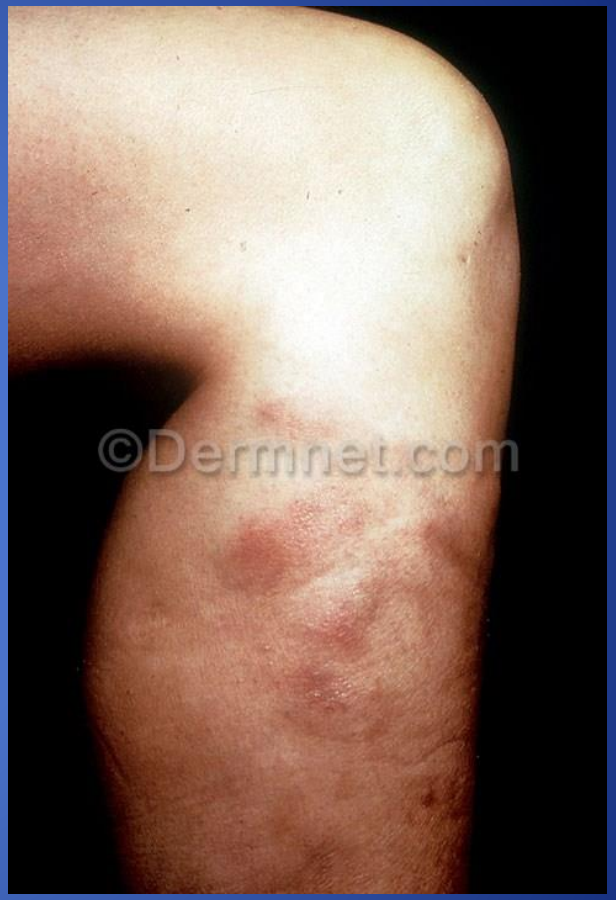






Cryoglobulinemia ?





Polyarteritis Nodosa ?

Polyarteritis Nodosa

Criteria	Comment
weight loss >= 4 kilograms	weight loss since illness began, not due to dieting or other factors
livedo reticularis	mottled reticular pattern over the skin of portions of the extremities or torso
testicular pain or tenderness	pain or tenderness not due to infection, trauma or other causes
myalgias, weakness or leg tenderness	diffuse myalgias (excluding shoulder and hip girdle), or weakness of muscles, or tenderness of leg muscles
mononeuropathy or polyneuropathy	may include multiple mononeuropathies
diastolic blood pressure > 90 mm Hg	
elevated BUN or creatinine	BUN > 40 mg/dL or creatinine > 1.5 mg/dL, not due to dehydration or urinary tract obstruction
hepatitis B antigenemia	presence of hepatitis surface antigen in serum, with immune complex formation
arteriographic abnormalities not resulting from arteriosclerosis, fibromuscular dysplasia or other noninflammatory causes	arteriogram showing aneurysms or occlusion of visceral arteries, not due to noninflammatory causes
Biopsy specimens of small or medium muscular artery show granulocytes or granulocytes and mononuclear leukocytes in the artery wall.	Transmural pleomorphic cellular infiltrate, fibrinoid necrosis, thrombosis, and aneurysm formation.

[22.09.01] Traditional Diagnostic Approach for Polyarteritis Nodosa (ACR Criteria)

Purpose: To diagnose polyarteritis nodosa based on the presence of certain clinical findings.

Gender of patient (enter M or F)	○ Male ◉ Female
Has rheumatoid arthritis been excluded? (Y or N)	◉ Yes ○ No
Has SLE with vasculitis been excluded? (Y or N)	◉ Yes ○ No
Usual body weight in kilograms, before onset of illness	167 ○ kilograms ◉ pounds
Body weight after onset of illness, in kilograms	167 ○ kilograms ◉ pounds
Could dieting or heavy exercise by the patient explain the weight loss? (Y or N)	○ Yes ◉ No
Can the increase in BUN and/or creatinine be explained by dehydration or urinary tract obstruction? (Y or N)	○ Yes ◉ No
Is hepatitis B antigen present in blood? (Y or N)	○ Yes ◉ No
Does a biopsy of a small or medium muscular artery show an infiltrate of neutrophils with or without mononuclear cells? (Y or N)	○ Yes ◉ No
Does an arteriogram show aneurysm or occlusion? (Y or N)	○ Yes ◉ No
Can the arteriogram findings be explained by arteriosclerosis, fibromuscular dysplasia or other noninflammatory cause? (Y or N)	○ Yes ◉ No

Please Click calculate if any changes are done.

Calculate Reset

Evaluation	Result
Data complete?	Yes
Evaluation appropriate?	Yes
Number of criteria present	2
Polyarteritis nodosa likely present?	No

Pseudovasculitis

Conditions that may present with a clinical syndrome mimicing vasculitis:

(1) atrial myxoma

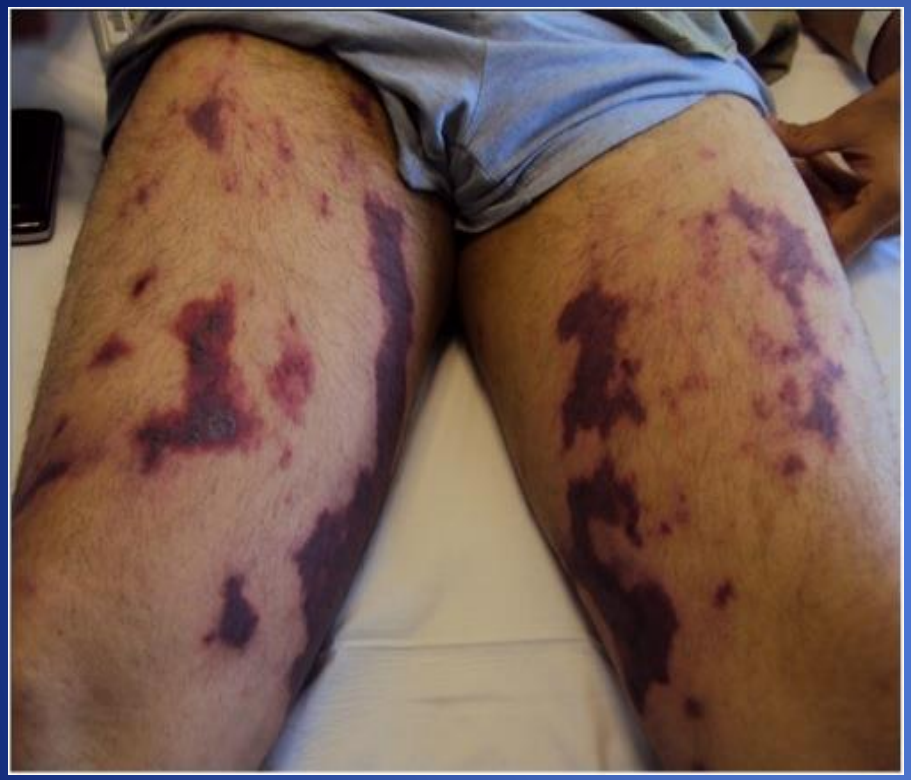
(2) septicemia

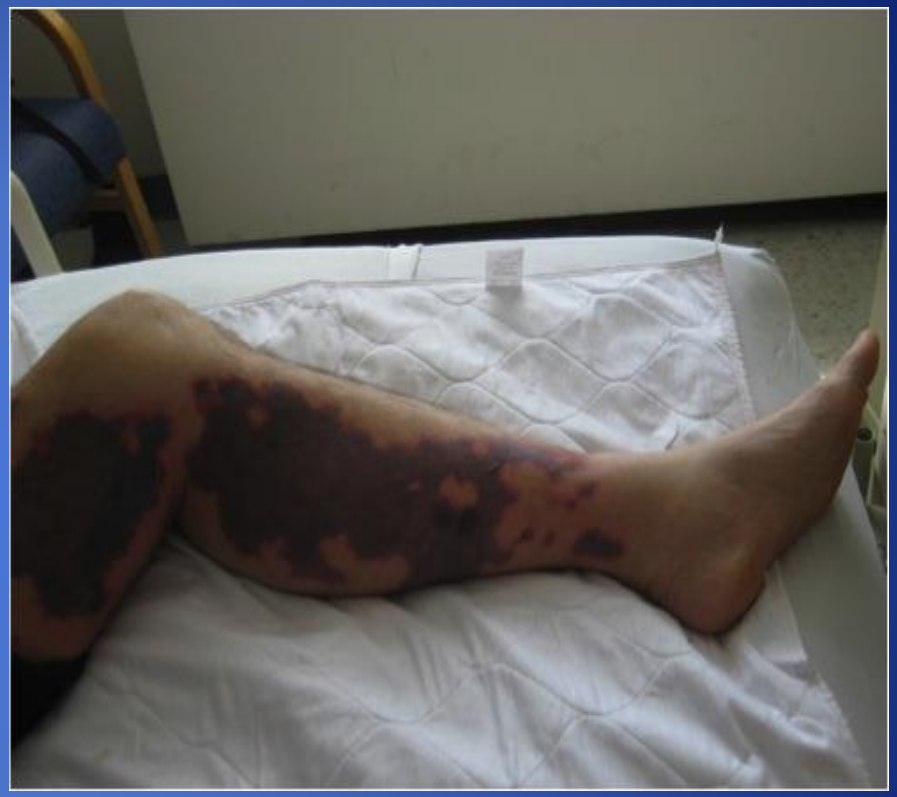
(3) chronic microthromboembolism

(4) infective endocarditis (both septic and microthrombemboli)

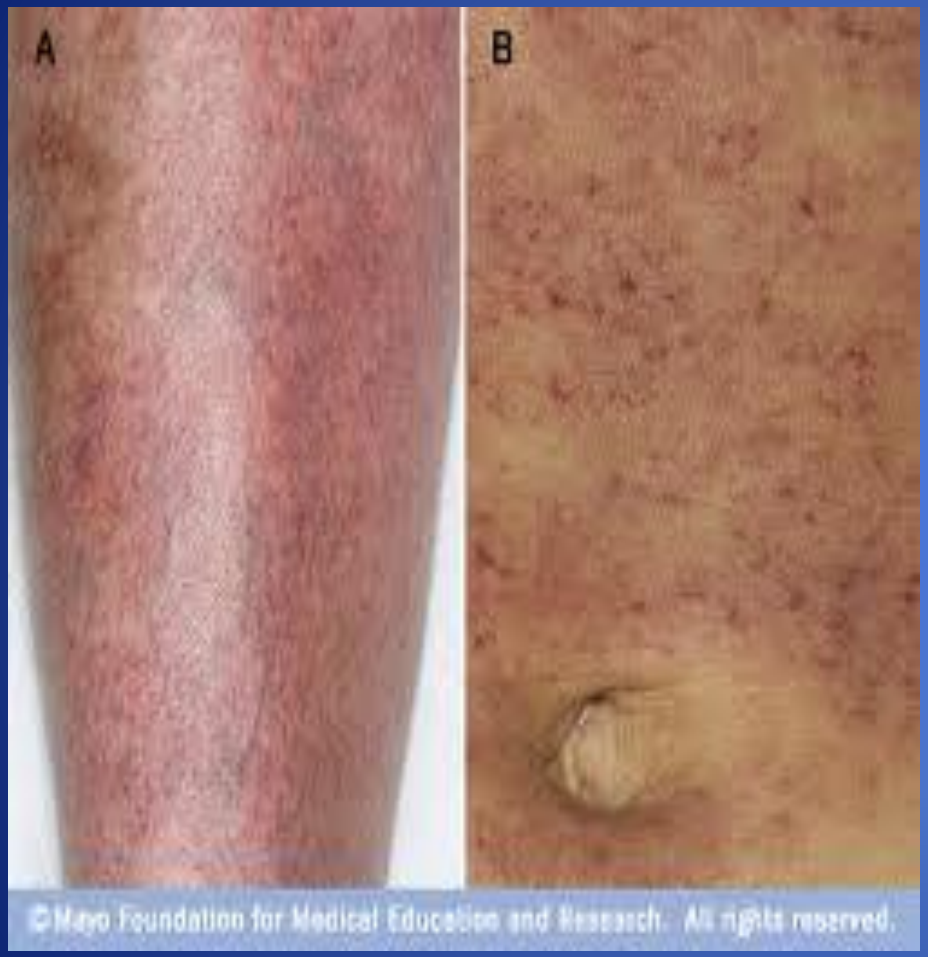
(5) rejection after organ transplantation

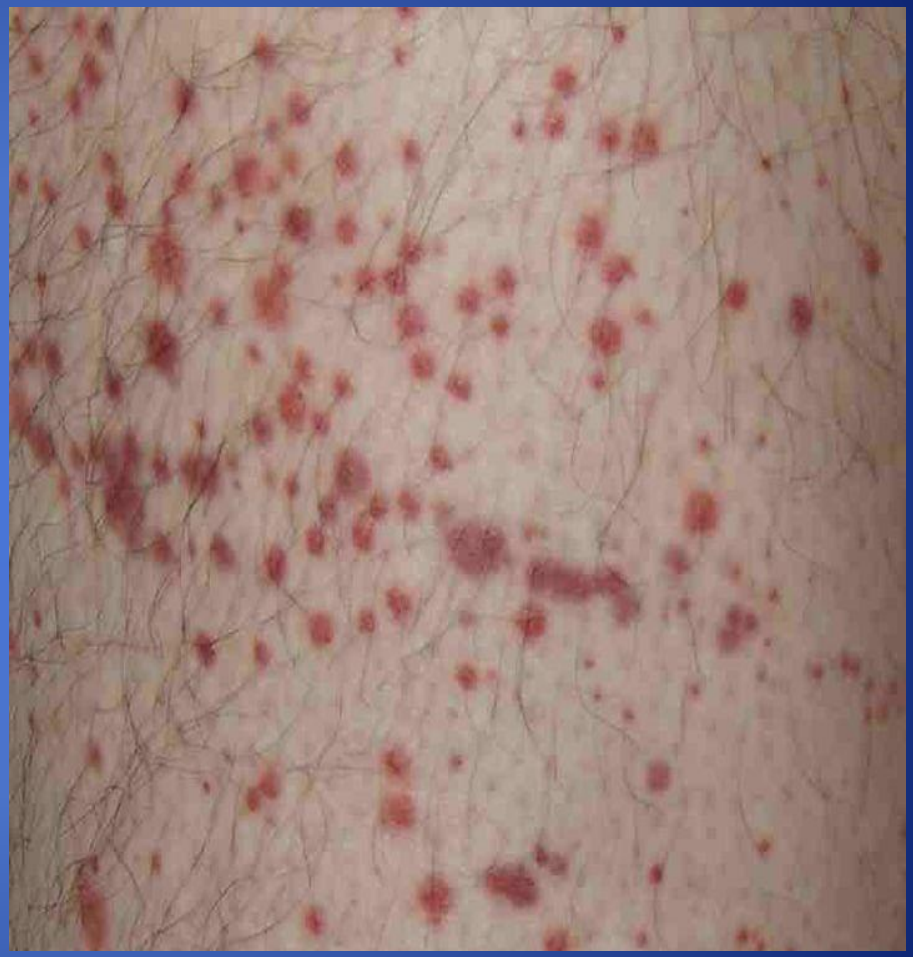
(6) drug or chemical related (ergotism, cocaine, etc)





Pseudovasculitis ?
Cocaine induced…

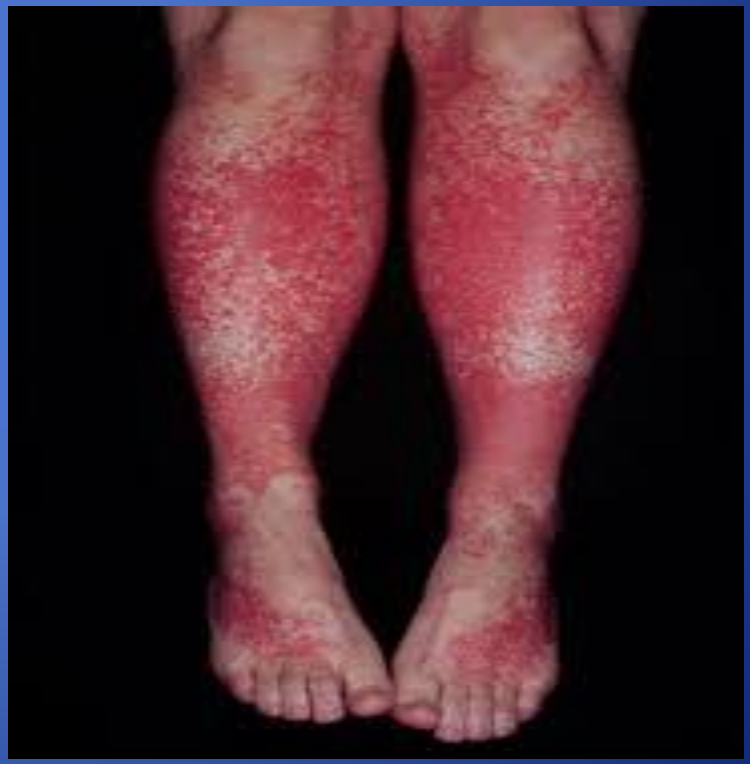




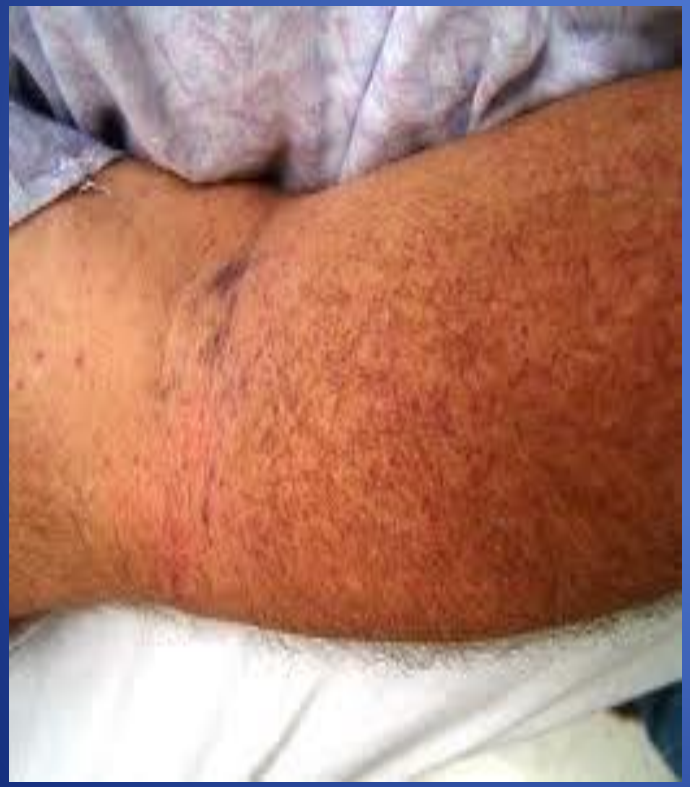
Idiopathic Thrombocytopenic Pupura ?

Leukemia ?

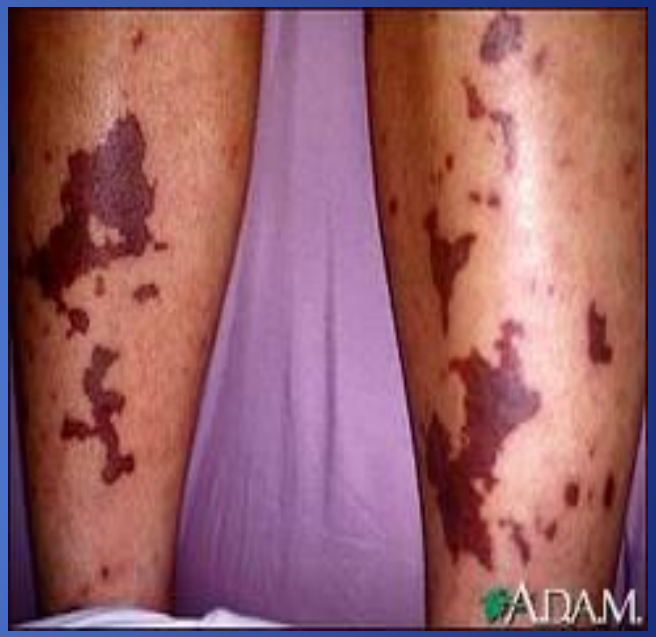
TTP ?

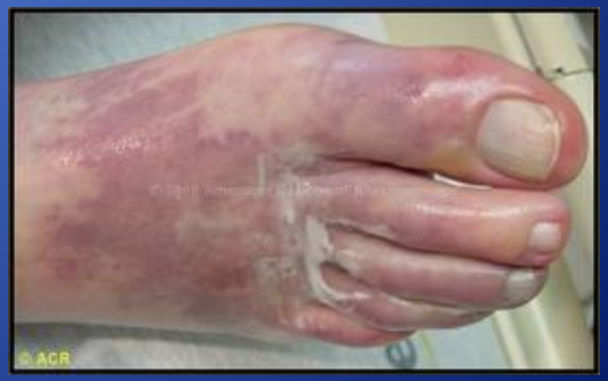


Drug induced Thrombocytopenia ?

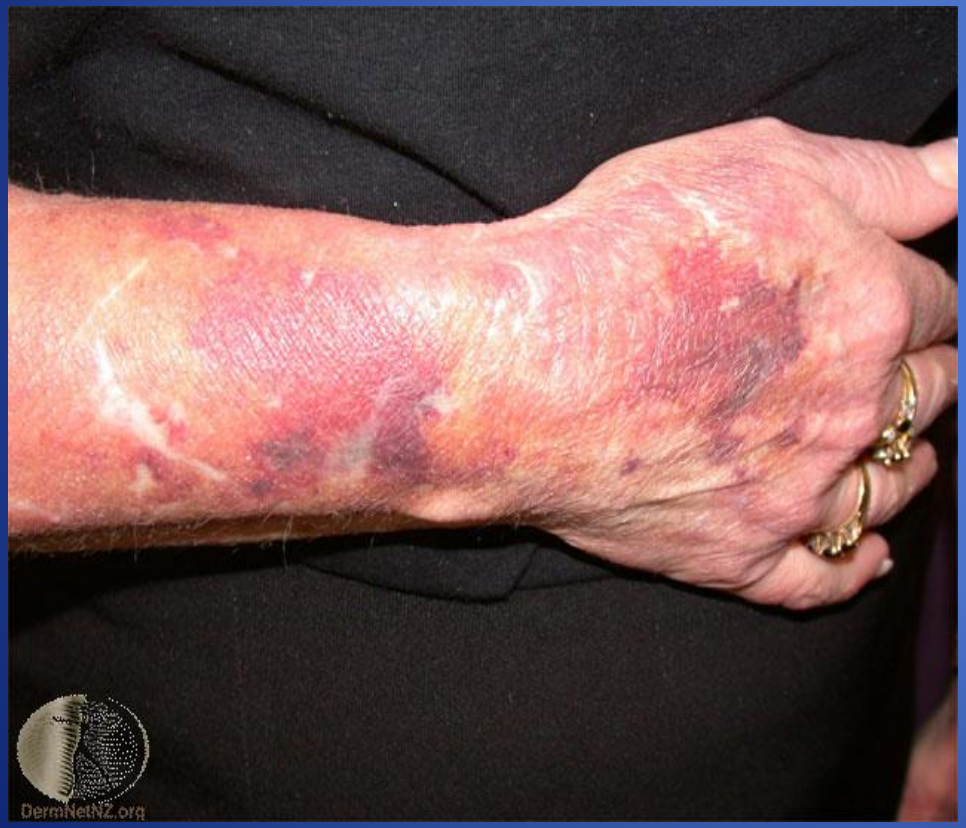


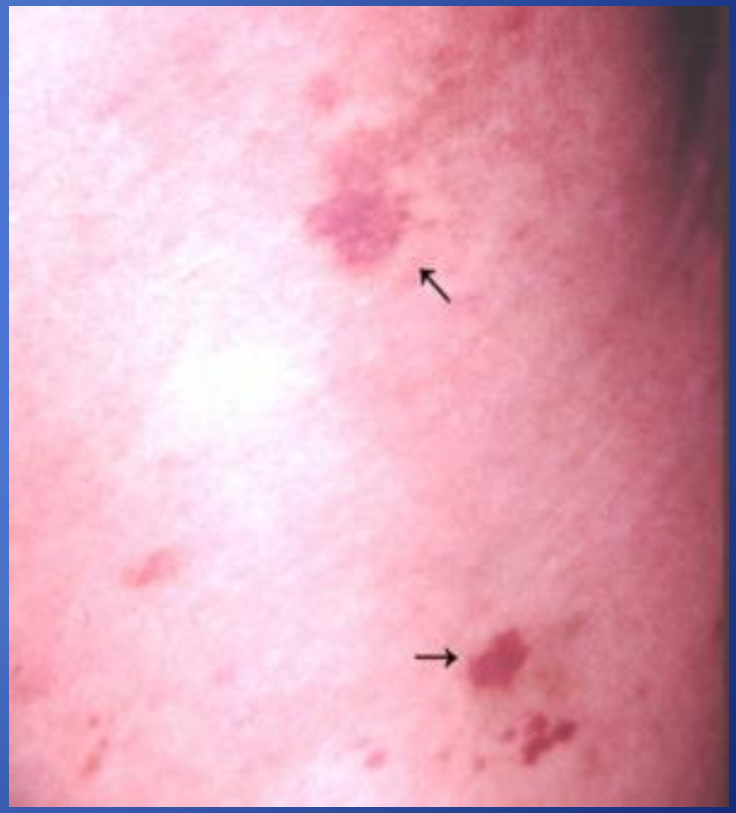
DIC ?



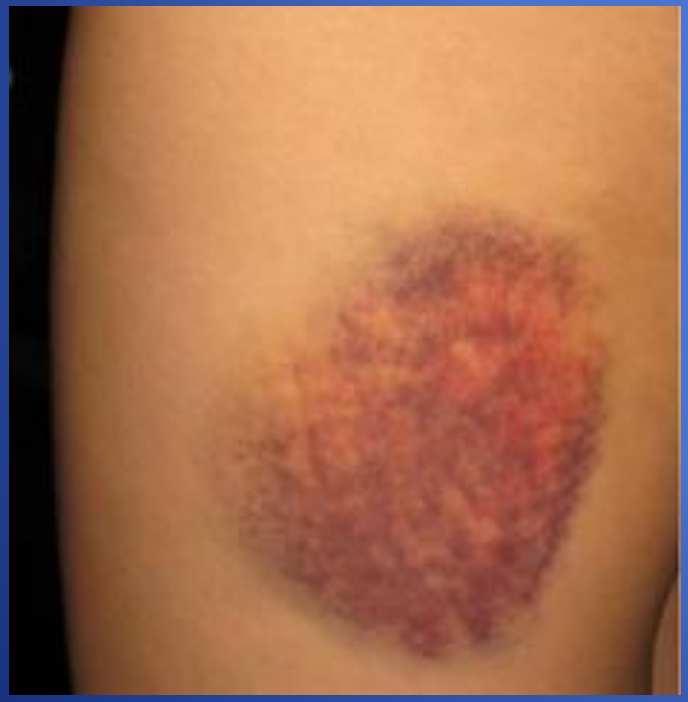


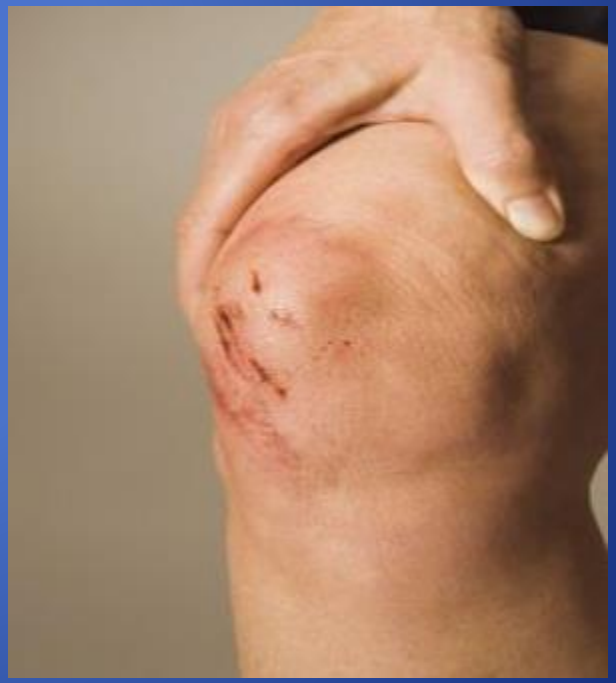
Corticosteroid atrophy and purpura



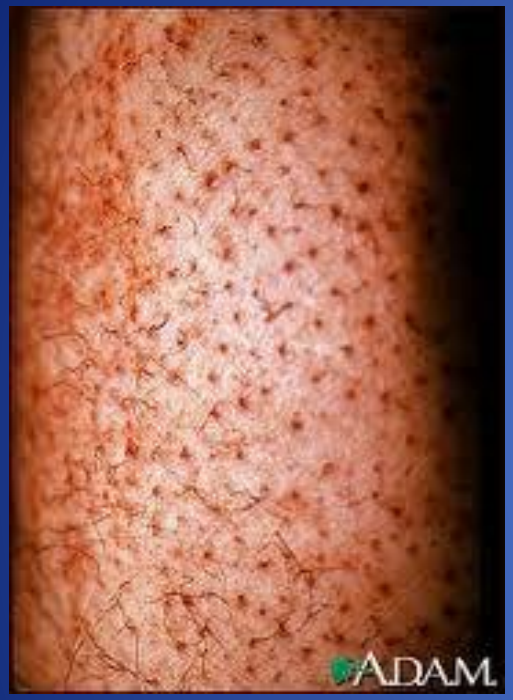


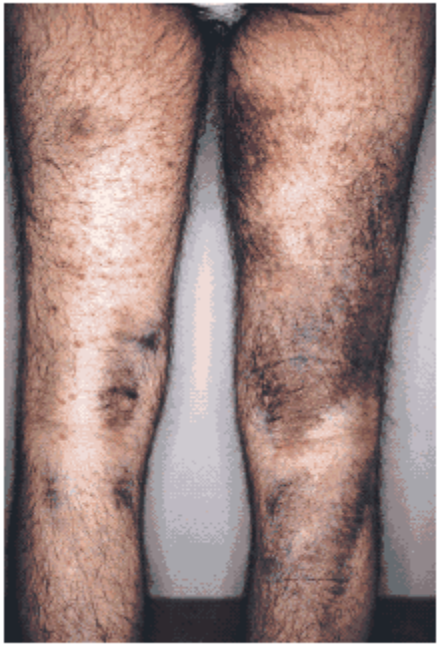
Vitamin K deficiency





Scurvy





Lab interpretation

- Normal platelet and wbc suggesting against thrombocytopenia and bone marrow suppression **: ITP, TTP**
- Normal PT and PPT and bleeding time, goes against coagulation factor deficiencies states **DIC, clotting factor inhibitors and vitamin K deficiency**
- Normal inflammatory markers, absence of elevated dsDNA and hypocomplementemia suggests against a **lupus vasculitis**
- Absence of pulmonary renal failure, negative serologies for **ANCA associated small vessels vasculitis**

Diagnostic workup: Anemia and Dyspnea

- Echocardiogram: normal (excludes CHF, Pulm HTN)
- Doppler negative for DVT

- Colonoscopy- negative (GI bleeding, HSP)
- V/Q scan: low probability for PE

- CT of the lower extremity showed a soft tissue infiltration in the medial thigh (bleeding)

Risk of vitamin C deficiency

- Scurvy: dietary deficiency of vegetables and vitamins
- Deficiency can cause typical perifollicular hemorrhage, collagen defect leads to superficial and deep tissue hemorrhage

Scurvy is the term for clinical deficiency of vitamin C (ascorbic acid). Although encountered less often today than in the past, it still occurs and mild cases may go unrecognized.

Patients at risk for vitamin C deficiency:

(1) infants

(2) adults > 55 years of age, especially males

(3) refugees or displaced persons

(4) malnutrition (including alcoholics)

(5) heavy cigarette smokers

(6) abnormal diets devoid of vegetables and vitamins

(7) mentally ill

(8) dialysis (hemodialysis or peritoneal dialysis)

Could this patient have Scurvy

data	enter		
Are you evaluating a patient for clinical evidence of scurvy? (Y or N)	Y	:-):-):-)	
age of the patient in years	54	years of age	0
Is the patient a refugee or displaced person? (Y or N)	N	:-):-):-)	0
Is the patient malnourished? (Y or N)	Y	:-):-):-)	1
Is the patient a heavy cigarette smoker? (Y or N)	Y	:-):-):-)	1
Is the patient an alcoholic? (Y or N)	N	:-):-):-)	0
Does the patient have a severe mental illness? (Y or N)	N	:-):-):-)	0
Is the patient undergoing hemo- or peritoneal dialysis? (Y or N)	N	:-):-):-)	0
Does the patient eat an abnormal diet devoid of vegetables and vitamins? (Y or N)	Y	:-):-):-)	1
Does the patient show?			
• petechiae? (Y or N)	Y	:-):-):-)	1
• purpura or ecchymosis? (Y or N)	Y	:-):-):-)	1

• subperiosteal hemorrhage? (Y or N)	N	:-):-):-)	0
• bleeding gums? (Y or N)	Y	:-):-):-)	1
• intracranial hemorrhage? (Y or N)	N	:-):-):-)	0
• conjunctival hemorrhages? (Y or N)	N	:-):-):-)	0
• gingival swelling and gingivitis? (Y or N)	Y	:-):-):-)	1
• loose or missing teeth? (Y or N)	Y	:-):-):-)	1
• scurvy buds on the gingiva? (Y or N)	Y	:-):-):-)	1
:-):-):-)	N	:-):-):-)	0
:-):-):-)	N	:-):-):-)	0
• tender or aching limbs? (Y or N)	Y	:-):-):-)	1
• anemia? (Y or N)	Y	:-):-):-)	1
• hyperkeratosis? (Y or N)	Y	:-):-):-)	1
• abnormal wound healing? (Y or N)	Y	:-):-):-)	1
• poor localization of bacterial infections? (Y or N)	N	:-):-):-)	0
• fatigue or weakness? (Y or N)	Y	:-):-):-)	1

Diagnosis : Scurvy

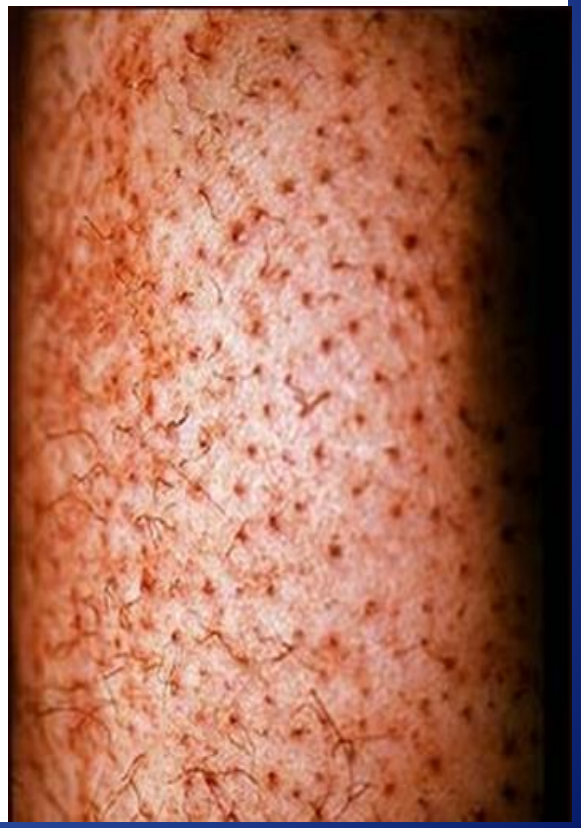
The diagnosis of scurvy involves both of the following:

(1) the presence of clinical features in a person with one or more risk factors for vitamin C deficiency

(2) low blood levels of vitamin C (preferably the concentration within leukocytes)

(3) clinical response to ascorbic acid replacement

calculate	result	
data complete?	Yes	
evaluation appropriate?	Yes	:-):-):-)
number of risk factors for scurvy	3	risk factors
number of clinical findings that could be due to vitamin C deficiency	11	clinical findings
Should blood levels of vitamin C be measured?	Yes	



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Volume 41, Issue 2 , Pages 286-290, October 2011

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Rheumatic Manifestations of Scurvy: A Report of Three Recent Cases in a Major Urban Center and a Review

Marty T. Mertens, MD, Elie Gertner, MD, FRCP(C), FACP

published online 24 December 2010.

- Syptoms including fatigue, purpuric rash, synovitis with effision, anemia and markedly elevated ESR and CRP
- One patient presenetd with severe pulmonary hypertension
- Exam consistnt with hemarthrosis and classic skin findings
- Treatment with Vitamin C 500mg-1000mg daily adequately replensihes
- Body stoes
- 1-3 weeks for resolution of skin findings
- 1-3 moths for hematologic and hemarthrosis

Thank You