Derma Team 436

Purpura and Vasculitis

Objectives: not given

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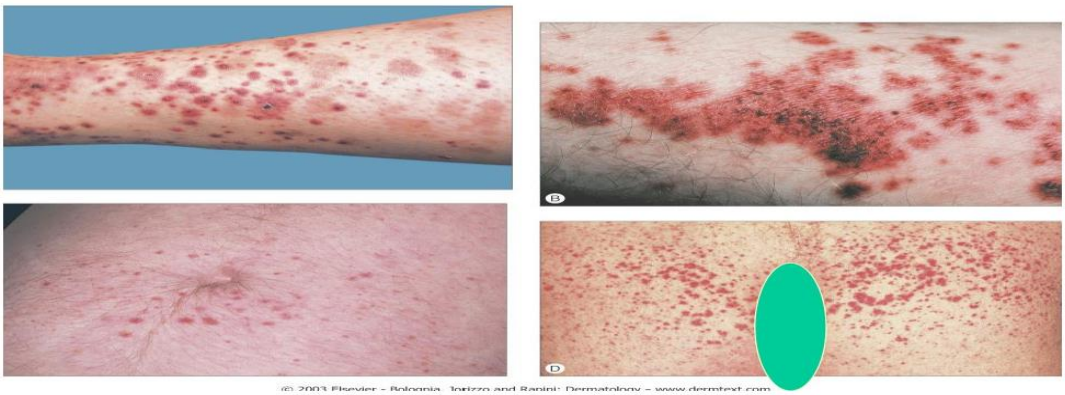
Before you start.. CHECK THE EDITING FILE

Sources: doctor's slides and notes + FITZPATRICK color atlas
[Color index: Important | 435 notes | gold | doctor notes | Extr]

Purpura

Definition: Visible hemorrhage into the skin or mucous membrane
It's a condition of red or purple discolored spots on the skin that do not blanch (doesn't disappear) on applying pressure using glass if blanchable → Erythema (Dilated vessels), if not blanchable → purpura
They subdivided as a follow:
-Petechiae less than or equal 4 mm
-Purpura (>4mm - < 1cm)
which can be either Palpable or non-palpable(macular) Most important DDx of palpable purpura is vasculitis. You have to role out vasculitis if it is palpable purpura
-Ecchymoses > or equal to 1 cm



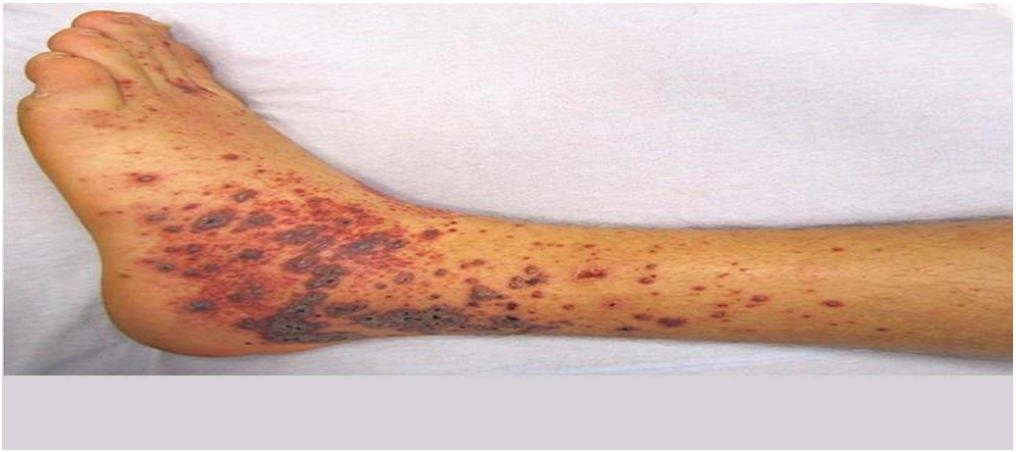
Causes:

PLATELET DISORDERS	*THROMBOCYTOPENIA, PLATELET DYSFUNCTION*
Coagulation Factor Deficiency	*Congenital:* *FACTOR VIII DEFICIENCy - fACTOR IX DEFICIENCY* *VON WILLEBRANDS DISEASE* *Acquired:* DISSEMINATED INTRAVASCULAR COAGULOPATHY LIVER DISEASE -UREMIA -VITAMIN K DEFICINCY
Vascular factors	CONGENITAL HEREDITARY HEMORRHAGIC TELANGECTASIA EHLERS-DANLOS SYNDROME (TYPE IV) ACQUIRED: INFLAMMATION(VASCULITIS) TRAUMA - **VITAMIN** C DEFICIENCY (SCURVY)

Vasculitis

Definition: A clinicopathologic process characterized by inflammatory destruction of blood vessels that results in occlusion or destruction of the vessel and ischemia of the tissues supplied by that vessel.

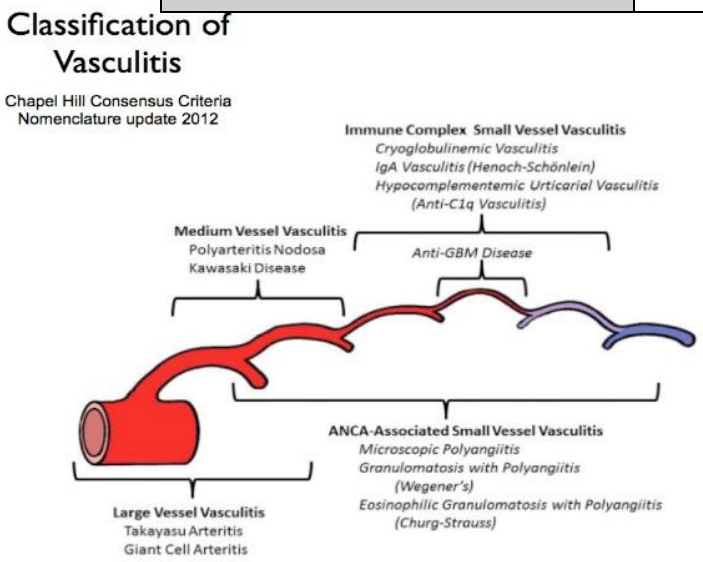
Vasculitis could present with either one of these → Purpura, nodules, ulceration, livedo reticularis.



Characteristic: Purpura, palpable, lower extremities

Classification:

Large-vessel vasculitis	*Aorta and the great vessels (subclavian, carotid)* *Claudication, blindness, stroke*
***Medium-vessel* vasculitis**	*Arteries with muscular wall* *Mononeuritis multiplex (wrist/foot drop), mesenteric ischemia, cutaneous ulcers*
Small-vessel vasculitis	*Capillaries, arterioles, venules* *Palpable purpura, glomerulonephritis, pulmonary hemorrhage*



CHAPEL HILL CONSENSUS CLASSIFICATION
Large-vessel vasculitis
• Giant cell arteritis • Takayasu's arteritis
Medium-vessel vasculitis
• Classic polyarteritis nodosa • Kawasaki disease
Small-vessel vasculitis
• Wegener's granulomatosis • Churg–Strauss syndrome • Microscopic polyangiitis (polyarteritis) • Henoch–Schönlein purpura • Essential cryoglobulinemia • Cutaneous leukocytoclastic vasculitis

Cutaneous small vessel vasculitis:

-most common type of vasculitis and it primarily affect post-capillary venules

-Palpable purpura is the hallmark. pinpoint to several centimeters

Can be presented as erosion and ulceration

-Early on lesion may not be palpable, Papulonodular, vascular, bullous, pustular or ulcerated forms may develop. may be localized to the skin or may manifest in other organs.

-The internal organs affected most commonly include the joints, GIT, and the kidneys.

-Renal involvement present as glomerulonephritis

- The prognosis is good in the absence of internal involvement

-Predominate on the ankles and lower legs i.e. dependent areas

Pathogenesis:

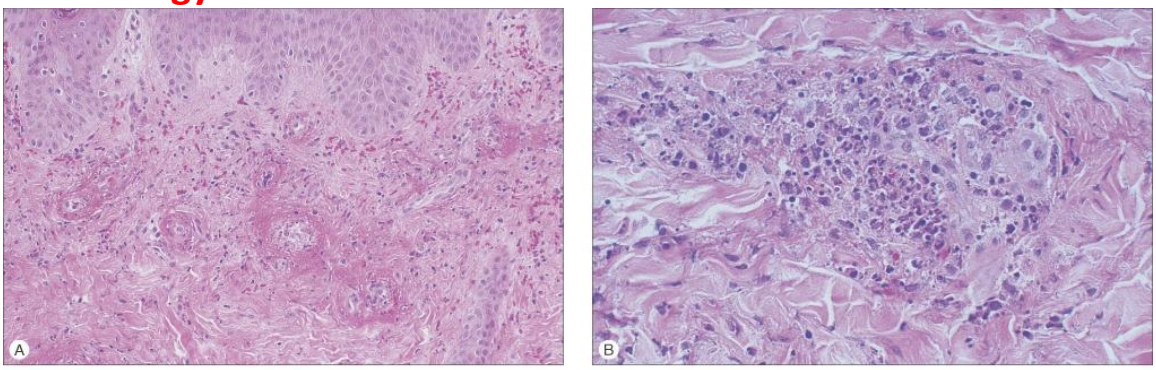
-Many forms of small-vessel vasculitis are felt to be caused by circulating immune complexes

-These lodge in vessel walls and activate compliment.

Table 3. Causes of cutaneous vasculitis[5,6]

Infections	
Bacterial	• Streptococcal, meningococcal, urinary tract infections
Viral	• Hepatitis B and C, HIV
Mycobacterial	• Tuberculosis
Connective tissue disorders	• SLE and related conditions • Rheumatoid arthritis • Systemic sclerosis, Sjogren syndrome • Dermatomyositis • Medium vessel vasculitides (Wegener granulomatosis, polyarteritis nodosa, Churg-Strauss syndrome)
Malignancy	• Haematologic — myeloproliferative — lymphoma — monoclonal gammopathy — multiple myeloma
Drugs	Including antibiotics, antihypertensives
Idiopathic	Henoch-Schonlein purpura

Histology:



Agiocentric segmental inflammation, endothelial cell swelling, fibrinoid necrosis of blood vessel walls and a cellular infiltrate composed of neutrophil with RBC extravasation.

Work up:

-Detailed history and physical examination
-History should focus on possible infectious disorders, prior associated diseases, drugs ingested, and a thorough review of systems
-CBC, strep throat culture or ASO titer, Hep B & C serologies and ANA are a reasonable initial screen, renal profile
-URINALYSIS FOR RBC, PROTIEN & CAST
- Skin biopsy most important tool

ble 26.4 Laboratory evaluation in known or suspected vasculitis.

LABORATORY EVALUATION IN KNOWN OR SUSPECTED VASCULITIS	
ystem	Technique
leme	Complete blood count with differential and platelet count, erythrocyte sedimentation rate (ESR), C-reactive protein
enal	Urinalysis, BUN, creatinine
iver	Abnormal liver function tests, hepatitis B and C antibody, cryoglobulins
mmunologic	Serum complement, rheumatoid factor, antinuclear antibody, anti-dsDNA, extractable nuclear antigen, antineutrophil cytoplasmic autoantibodies (ANCA)
nfectious	Blood and cultures
lead and neck	Sinus radiographs and CT
ulmonary	Chest radiograph or CT
ardiovascular	Electrocardiogram, creatine phosphokinase, echocardiogran
leurologic	Nerve conduction studies
Musculoskeletal	Electromyography

DIAGNOSTIC WORKUP

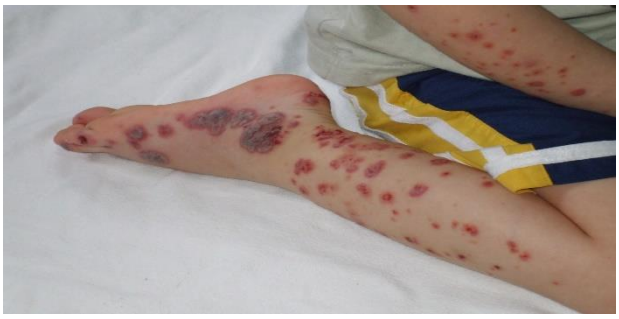
Laboratory investigations

- Full blood count with differential white cell count
- Markers of inflammation: ESR,CRP
- Electrolytes and hepatic transaminases, glucose
- Urinalysis for protein and blood
- Blood cultures (if pyrexial)
- Serology—ANA ,dsDNA , ANCA,C3 and C4,ASLO titre, viral titres (e.g. hepatitis B and hepatitis C, possibly HIV, CMV, parvovirus B19 and others if recent infection).
- Others-rheumatoid factor, electrophoresis, immune complexes.

Treatment: treatment of cause.

-Symptomatic treatment (if skin is only involved): rest, NSAIDS, Antihistamine
-severe visceral involvement may require high doses of corticosteroids with or without an immunosuppressive agent
-Immunosuppressive agents for rapidly progressive course and severe systemic involvement Systemic steroids if there is internal organ involvement.

Henoch-Schönlein purpura HSP:



Primarily occurs in male children
-peak age 4-8 years
-Adults may be affected
-A viral infection or streptococcal pharyngitis is the usual triggering event

-In about 40 % of the cases the cutaneous manifestations are preceded by mild fever, headache, joint symptoms, and abdominal pain for up to 2 weeks
-Characterized by intermittent purpura, arthralgia, abdominal pain, and renal disease
-Typically, purpura appears on the extensor surfaces of the extremities
-Become hemorrhagic within a day and fades in 5 days
-New crops appear over a few weeks

May be associated with:
pulmonary hemorrhage
Abdominal pain and GI bleeding
-GI radiographs may show "cobblestone" appearance
-Renal manifestations may occur in 25% or more but only 5% end up with ESRD
The long-term prognosis in children with gross hematuria is very good; however, progressive glomerular disease and renal failure may develop in a small percentage you have to do urine analysis even if skin manifestations subsides
-IgA, C3 and fibrin depositions have been demonstrated in biopsies of both involved and uninvolved skin by immunofluorescence techniques

Treatment: supportive. (bed rest, pain relieve, etc)

Test your knowledge:

Q: what is the best treatment for Cutaneous small vessel vasculitis without systemic involvement?
a- methotrexate b-bed rest c-steroid

b

Q: what is pathognomic sign for small vessel vasculitis?
a- vesicles b- bulla c- palpable purpura d-scar

c