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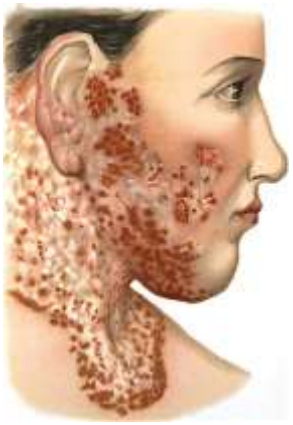
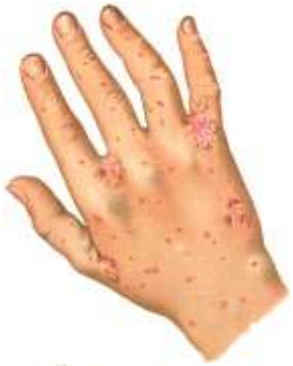
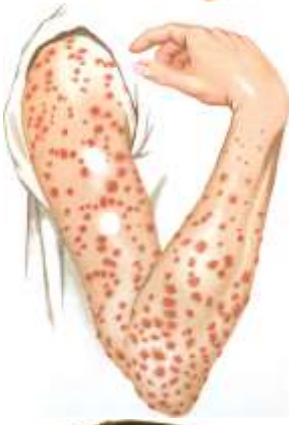
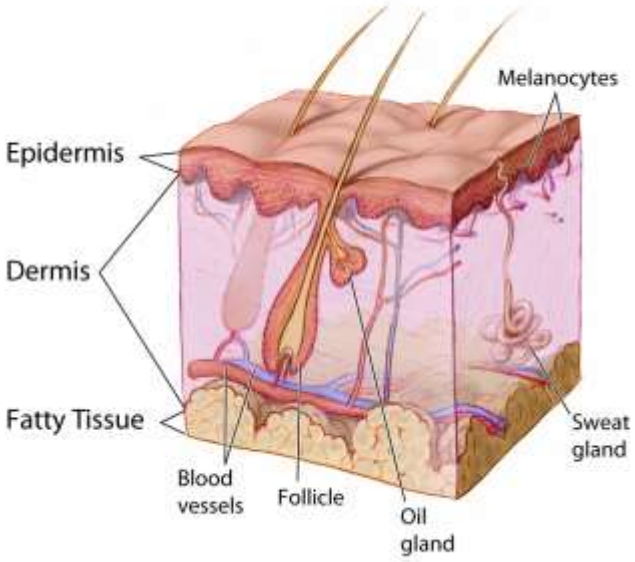






432 Teams

Dermatology



Purpura and Vasculitis

Color Code: Original, Team's note, Important, Doctor's note, Not important, Old teamwork

Done by: ***Jawaher Enani***

Reviewer: ***Lama Al Tawil***

Team Leader: ***Basil AlSuwaine &Lama Al Tawil***

Objectives- MALES OBJECTIVES -

1. differentiate between different types of purpura.
2. To know the difference between inflammatory and non inflammatory purpura.
3. To have an approach to diagnose purpuric lesions.
4. To be familiar with serious and non serious conditions and when to refer to a specialist.

When we asked the doctor about the difference in our slides in comparison to the male's , she said : I only want what I explained and everything you need is in the lecture.

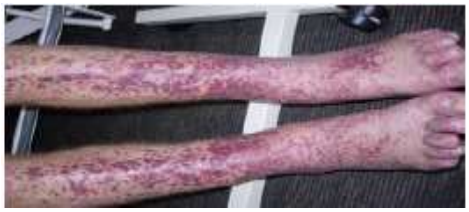
However, If you want to take a look over the extra topics check the male's teamwork.

PURPURA

DEFINITION :

Visible hemorrhage into the skin or mucous membrane subdivided as a follow:

-Petechiae less than or equal 4 mm
-Purpura (>4mm - < 1cm)
which can be either Palpable or non-palpable(macular)
-Ecchymoses > or equal to 1 cm



CAUSES :-

1-Platelet Disorders :-
Thrombocytopenia
Platelet Dysfunction
renal failure
Hepatic failure
"thrombotic thrombocytopenic pupura" "autoimmune disease"

The doctor said that this slide is not our main interest in this lecture, it's just to refresh you memory

2-Coagulation Factor Deficiency :-
Congenital
Factor VIII Deficiency
Factor IX Deficiency
Von Willebrands disease
Acquired
Disseminated Intravascular Coagulopathy
Liver disease
Uremia
Vitamin K deficincy

3-Vascular Factors :-
Congenital
Hereditary Hemorrhagic Telangectasia
Ehlers-Danlos Syndrome (Type IV) "defect in the collagen fiber –remember that collagen is presented in skin and blood vessels and other organs like joints- so patients usually die at an early age because of aortic dissection . They also have saggy skin
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.

CLASSIFICATION :- depending on the vessel's diameter

Table 26.2 Chapel Hill consensus classification.

CHAPEL HILL CONSENSUS CLASSIFICATION	
Large-vessel vasculitis	Like aorta and it's main branches
• Giant cell arteritis • Takayasu's arteritis	Not important because they don't present with skin disease so forget about them
Medium-vessel vasculitis	Arteries that supplies the organs like renal arteries and coronary arteries
• Classic polyarteritis nodosa	Might affect small vessels as well as organs. (skin might be affected)
• Kawasaki disease	In children, affect skin and coronary artery so if you suspect Kawasaki, R/O coronary artery disease
Small-vessel vasculitis	Mainly affect the skin, because it affect capillaries and post capillary venules
• Wegener's granulomatosis • Churg–Strauss syndrome • Microscopic polyangiitis (polyarteritis)	Might affect small and/or medium blood vessels
• Henoch–Schönlein purpura	
• Essential cryoglobulinemia	Due to high amount of cryoglobulin deposition
• Cutaneous leukocytoclastic vasculitis	

Behcet's disease is considered small vessel vasculitis

CLASSIFICATION CONTINUE :-

-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

Cutaneous small vessel vasculitis (Leukocytoclastic vasculitis)

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

PATHOLOGENESIS :-

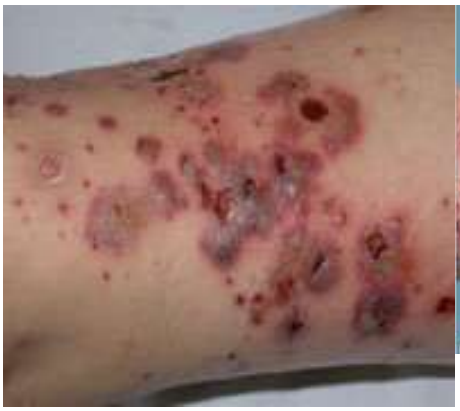
-Many forms of small-vessel vasculitis are felt to be caused by circulating immune complexes
-These lodge in vessel walls and activate compliment
It's due to type III hypersensitivity reaction due to immune complexes which leads to activation and deposition of the complement

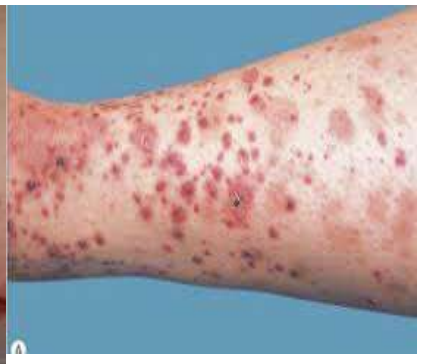
CAUSES :-

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

- streptococcal infection is common but **idiopathic is the most common type**
- Wegener granulomatosis, polyarteritis nodosa, churg-strauss syndrome → they all can present as small and/or medium blood vessel vasculitis.



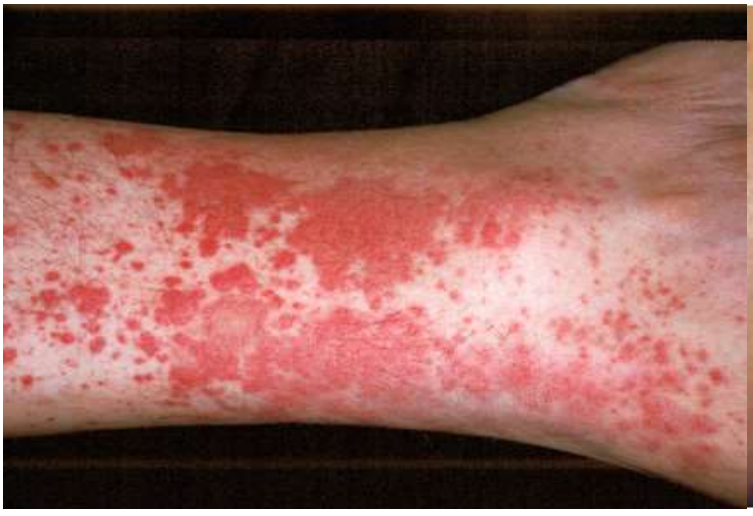


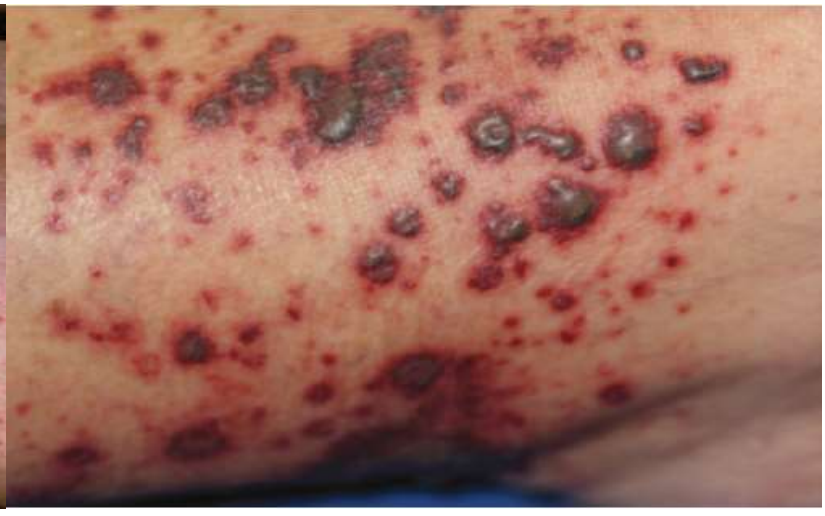
On examination :-

First thing we should do when a patient present :-

- take history and rule out post streptococcal infection and URTI
- We should take history of lupus and connective tissue disease
- History of malignancies and constitutional symptoms (weight loss, fever)
- Drug history

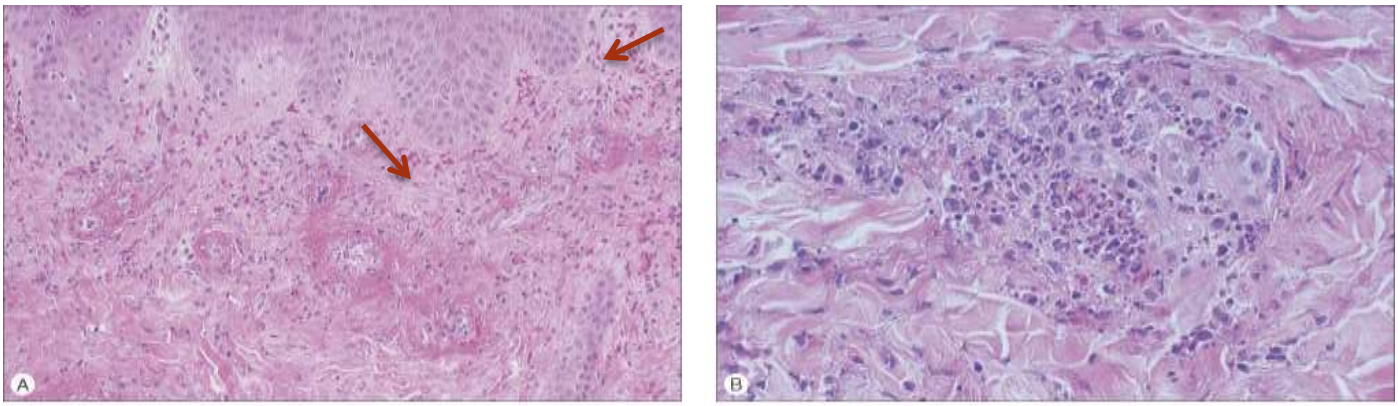
- -Palpable purpura is the hallmark
- -Pinpoint to several centimeters
- -Early on lesion may not be palpable, if the patient present in the first few days it will be raised, palpable, but with time it becomes flat
- -Papulonodular, vascular, bullous, pustular or ulcerated forms may develop
- -Predominate on the ankles , buttocks and lower legs i.e. dependent areas
- -the lesion itself might present with Mild itching , fever, malaise, arthralgia and/or myalgia may occur
- -Typically resolve spontaneously in 3 to 4 weeks
- -Residual postinflammatory hyperpigmentation may be seen
- -Self-limiting
- May recur or become chronic
- -Hemorrhagic vesicles or bullae filled with blood may develop
- *-may be localized to the skin or may manifest in other organs.*
- *Because the post capillary venules are found in skin and in addition to other organs*
- *-The internal organs affected most commonly include the joints, GIT, and the kidneys.*
- *-Renal involvement present as glomerulonephritis*
- *-if only the skin "regardless the cause" in the absence of internal involvement, The prognosis is good because it's self limiting disease but if there is internal involvement there might be a secondary consequences "sequella"*





HISTOLOGY :-

Agiocentric segmental inflammation –inflammation of blood vessels- , endothelial cell swelling, fibrinoid necrosis of blood vessel walls and a cellular infiltrate composed of ***neutrophil*** *"main cells in inflammation hence the name (leuko-)" with RBC extravasation the red spots –see the arrow-.*



WORK UP :-

- *-Detailed history to rule out all the causes and physical examination*
- *-History should focus on possible infectious disorders, malignancies and B symptoms prior associated diseases, drugs ingested, and a thorough review of systems*
- *-CBC with differential to rule out malignancies and infections, strep throat culture or ASO titer to rule out streptococcal, Hep B & C serologies and ANA to rule out SLE are a reasonable initial screen, renal profile, ESR*
- ***-URINALYSIS –very important to rule out complications- FOR RBC , PROTIEN & CAST→ if they were elevated then there is renal impairment → you have to involve other teams (medicine, pediatric….) to assess the kidneys .***
- *Two Skin Biopsy for* ***histopathology*** *and* ***DIF for antibodies***

TREATMENT :-

- *-treatment of cause.*
- *-if only skin involvement, it's self-limiting, we only do Symptomatic treatment : rest , NSAIDS , Antihistamine*
- *-severe visceral involvement may require high doses of corticosteroids with or without an immunosuppressive agent*
- -Immunosuppressive agents for ;-
  A) rapidly progressive course –rapidly progressive skin disease-
  B) severe systemic involvement, like renal.

If urinalysis came to be normal we have to follow up the patient because it may present negative at first but up to one month they can still develop impairment "might turn positive at any time, so we should follow the patient"

Henoch-Schönlein purpura (HSP)
SMALL VESSEL VASCULITIS

- *-Primarily occurs in male children*
- *-peak age 4-8 years*
- *-Adults may be affected mainly children*
- *-A viral infection or streptococcal pharyngitis are the usual triggering event or it could be idiopathic*
- *-In about 40 % of the cases the cutaneous manifestations are preceded by mild fever, headache, might have GI symptom which differentiate from the regular Leukocytoclastic vasculitis, joint symptoms, and abdominal pain and for up to 2 weeks*

We have to ask about abdominal symptoms and constitutional symptoms to rule out the causes

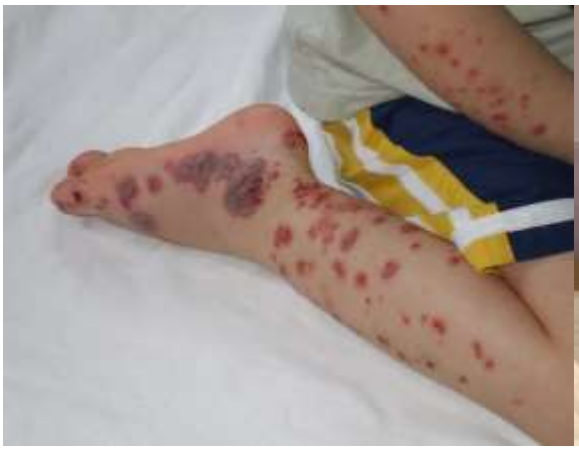
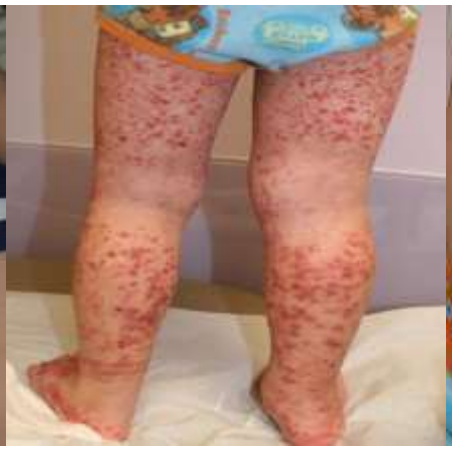
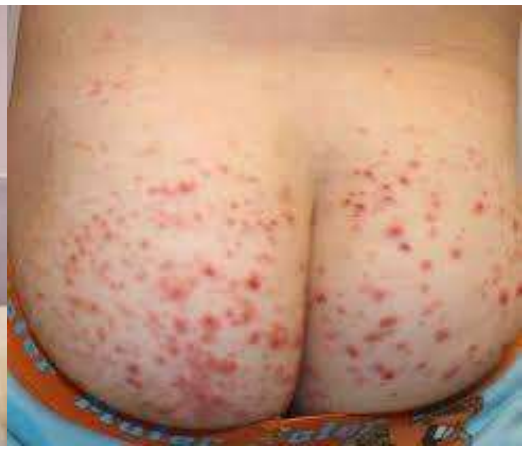
- *-Characterized by **intermittent purpura, arthralgia, abdominal pain, and renal disease***
- *-Typically purpura appears on the extensor surfaces of the extremities*
- *-Become hemorrhagic within a day and fades in 5 days*
- *-New crops appear over a few weeks*
- *-May be associated with:*
- *pulmonary hemorrhage*
- *Abdominal pain and GI bleeding*
- *Arthralgia*
- *→ in patient suspected HSP you have to rule out pulmonary involvement , and abdominal pathology, do x-ray and urinalysis as well but at first we have to call the medicine and pediatric to evaluate the patient*
- *-GI radiographs x-ray may show "cobblestone" appearance*
- *-Renal manifestations may occur in 25% or more but only 5% end up with ESRD so we have to do urinalysis*
- *It is very important to investigate and diagnose it because you don't want your patient to 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*

- ***This is VERY IMPORTANT !!***
  -IgA, C3 and fibrin depositions have been demonstrated in biopsies of both involved and uninvolved skin by immunofluorescence techniques
- *That's how we differentiate it from other diseases*
- *Any patient with purpura we have to do two skin biopsies one for histopathology and one for Immunofluorescence*

What to do when a patient present with PURPURA ??

URINALYSIS

SUMMARY –FROM THE BOOK-

- Vessels are involved in most inflammatory processes in the human body.
- **Vasculitis** denotes conditions where vessels are the target of inflammation. The vasculitides can best be classified according to the size of vessels involved.
- Hypersensitivity vasculitis (HV) encompasses a heterogeneous group of vasculitides associated with hypersensitivity to antigens from infectious agents, drugs, or other exogenous or endogenous sources.
    - It is characterized pathologically by involvement of postcapillary venules and inflammation and fibrinoid necrosis (necrotizing vasculitis).
    - Clinically, skin involvement is characteristic, manifested by "palpable purpura."
    - Systemic vascular involvement occurs, chiefly in the kidney, muscles, joints, GI tract, and peripheral nerves.
- Henoch–Schönlein purpura is a type of HV associated with IgA deposits in the skin.
    - Synonyms: Allergic cutaneous vasculitis, necrotizing vasculitis.
    - Henoch–Schönlein Purpura
    - This is a specific subtype of hypersensitivity vasculitis that occurs mainly in children but also affects adults.
    - There is a history of upper respiratory tract infection (75%), by group A streptococci.
    - Histopathologically, there is necrotizing vasculitis and the immunoreactants deposited in skin are IgA.
    - Long-term morbidity may result from progressive renal disease (5%)

MCQ

A 6-year old boy presented with palpable purpuric papules and plaques over the shins and buttoks for 5 days associated with abdominal pain.

If you send skin biopsy for direct immunoflouroscence, which of the following is typical for this disease?

A. C2 and C4 Deposition

B. IgG and IgM deposition

C. IgG deposition

D. IgA and C3 deposition

A 15-year-old boy presented to the emergency department with purpuric papules and plaques over the shins for one day associated with pain of the ankle joints, there symptoms were preceded by upper respiratory tract infection.

Which of the following investigation is helpful to rule out internal organ involvement by vasculitis?

A. Urine analysis for RBC casts

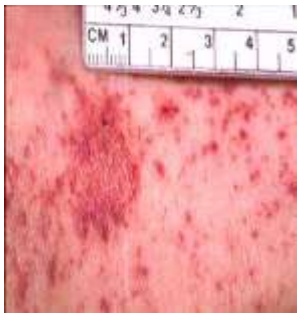
B. Renal function test

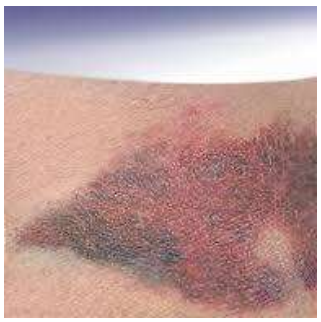
C. Chest X-rays

D. CBC

ANS: *D,A*

Identify The following :





On the left its Petechiae on the right its Ecchymosis or bruises

GOOD LUCK