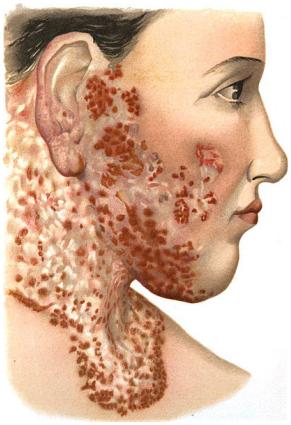
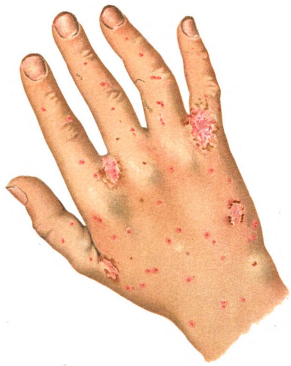
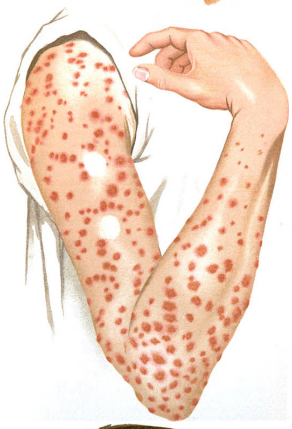
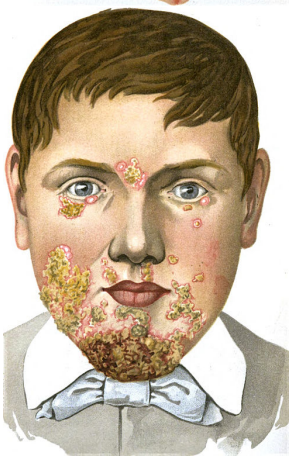
جامعة الملك سعود
King Saud University



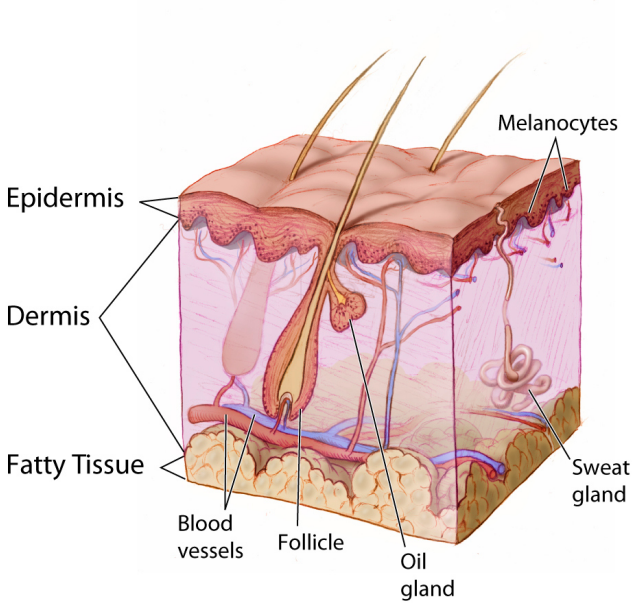






432 Teams

Dermatology



Purpura and Vasculitis

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

Done by: ***Turki AlOtaibi***

Reviewer: ***Fahad AlTurki***

Team Leader: ***Basil AlSuwaine***

1

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.

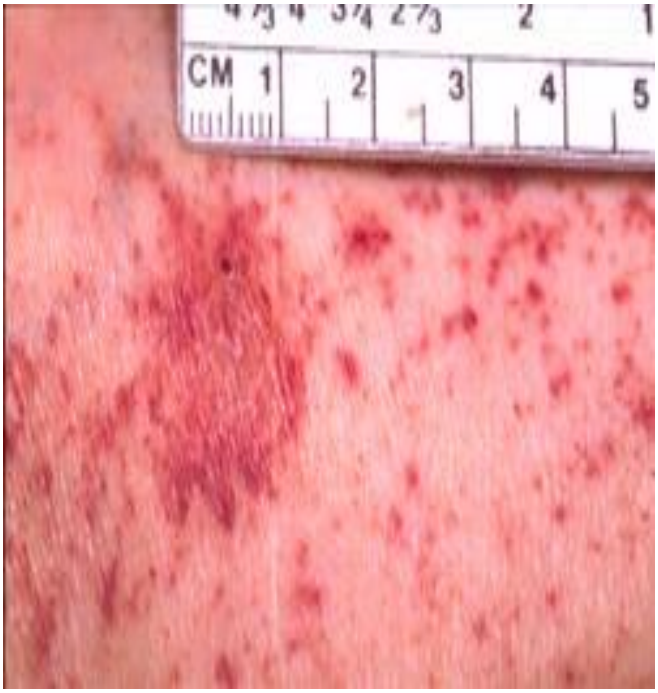
Purpura

Reddish-purplish skin lesions from extravasation of RBCs into the skin.

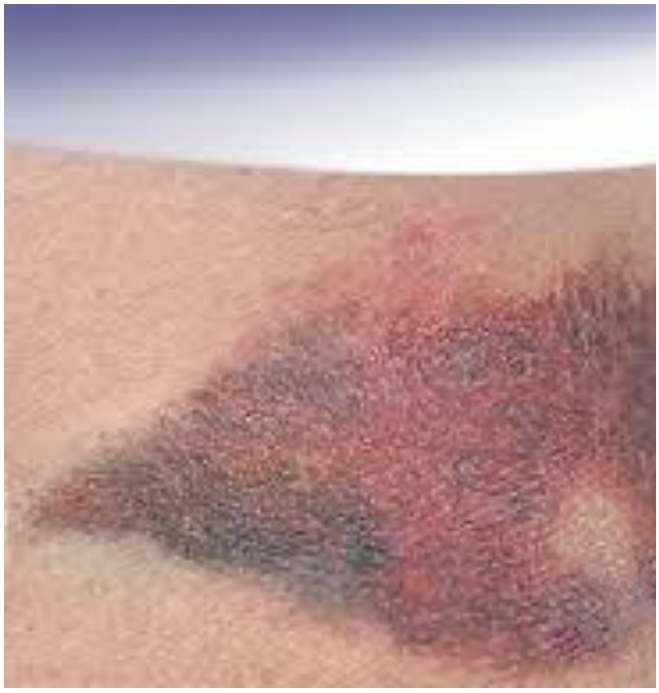
- **Non palpable purpura:** classified according to size to:
  Petechiae less than 2 mm, ecchymosis or bruises more than 2 mm

- **Palpable purpura** is the inflammatory type (presence of inflammatory cells) of different sizes, the main sign of vasculitis.

Petechiae



Ecchymosis or bruises



CAUSES OF PUPURA:

- **Vascular damage** from trauma, aging (decrease support around the vessels collagen & fat), nutritional (Scurvy "vit C deficiency") or vasculitis.
- **Decreased platelets numbers or function.**
- **Coagulopathy** like DIC, drug induced bleeding like heparin and warfarin, or other clotting factors deficiency.

Vasculitis

- **Large vessels**: Giant cell arteritis (temporal arteritis), Takayasu's arteritis. (aorta)

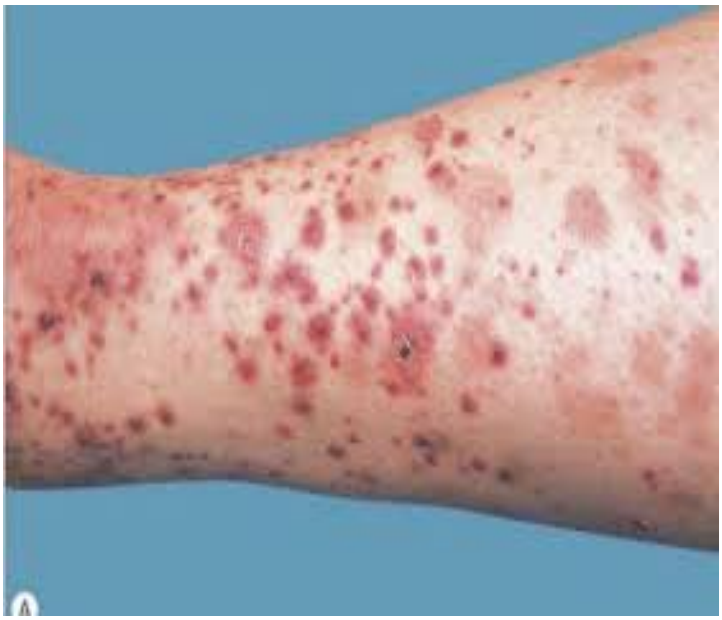
- **Medium sized vessels**: polyarteritis nodosa, Kawasaki disease. (fever > 5 days, cervical lymph nodes are enlarged, rash and aneurysm around the heart.)

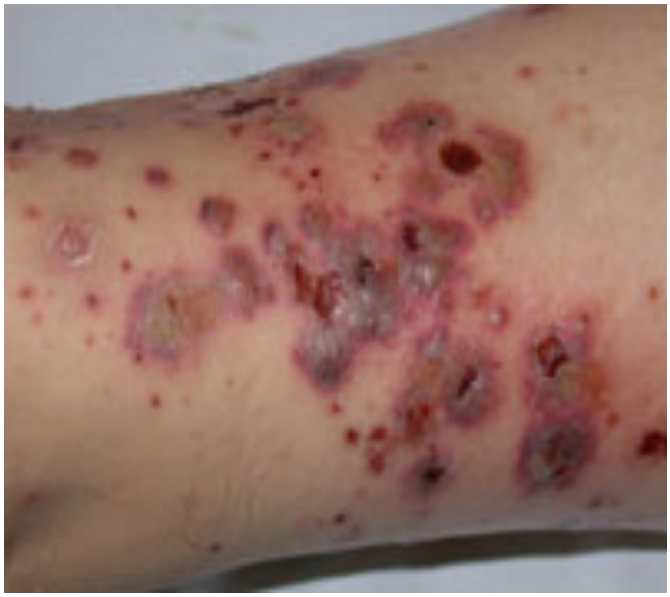
- **Small vessels**: Wegener's granulomatosis, Churg-Strauss syndrome, Microscopic polyangiits, Essential cryoglobulinemia vasculitis, Cutaneous small-vessel vasculitis (CSVV), Henoch-Schönlein purpura, Urticarial vasculitis. Cutaneous polyartiritis nodosa *Pigmented Purpuric Dermatoses.*

- Cutaneous small-vessel vasculitis (CSVV):

Also known as Leukocytoclastic vasculitis

- Characterized by neutrophilic infiltration into the peripheral small dermal blood vessels.
- Purpura, urticaria, erythema-multiforme-like erythema, papules, nodules, pustules, blistering, erosion and ulceration occur, mainly in the lower extremities.





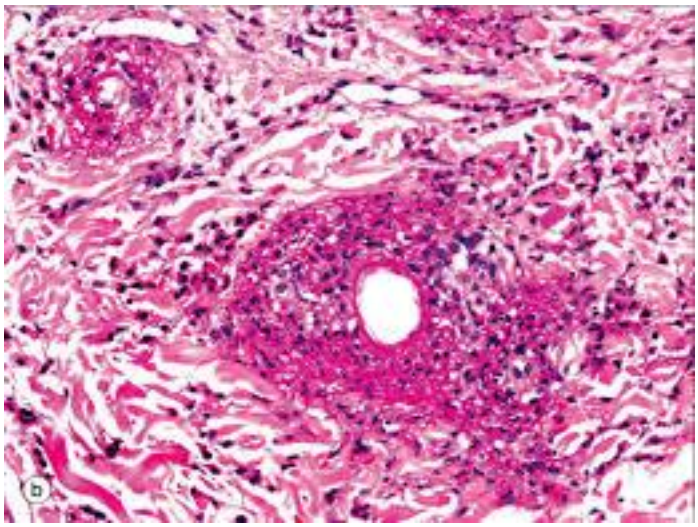
- An immune complex of an antigen (e.g., bacterium, virus, drug) and the antibody against that antigen deposit on the arteriolovenular walls. These activate the immune system and cause vasculitis (type III allergic reaction). Penicillin, sulfa drugs, thiazides, NSAIDs and other drugs, chemical substances, hemolytic streptococcus bacteria, or viruses may be the foreign antigen. Collagen diseases and antibodies against malignant tumors can also be causes.
- 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 Viral Mycobacterial	• Streptococcal, meningococcal, urinary tract infections • Hepatitis B and C, HIV • 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

- Pathology:

In the upper and middle dermal layer, fragments of nuclear debris and leakage of erythrocytes are found in the peripheral arteriola. Neutrophilic infiltration occurs in the arteriolovenous small blood vessels and capillaries. Thickening of the blood vessel walls and fibrinoid necrosis (important in diagnosis).



- Investigations:

- CBC, urea, creatinine.

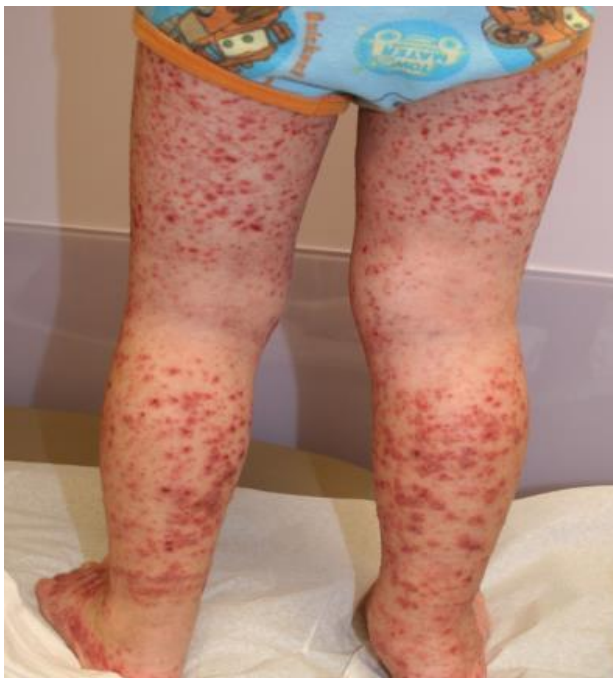
- ESR (usually raised)
- Complements (decreased) (consumed in immune complexes formation).
  o Urinalysis (for protein and hematuria if kidneys involved)
- Occult blood in stool
- ANA (SLE), ANCA (Wegners).
- Cryoglobulins, hepatitis B, C, HIV.
- CRP, ASO titer and throat swab culture for streptococcal infection
- Skin biopsy. (important for diagnosis)

- Management:

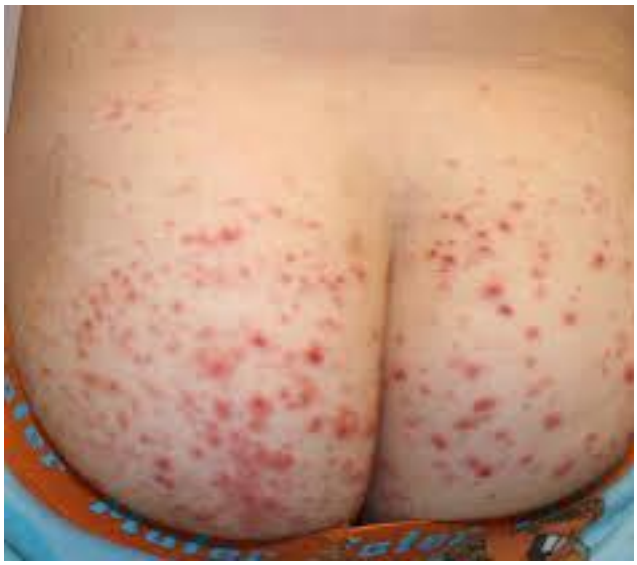
- *-treatment of cause.*
- *-Symptomatic treatment (if skin is only involved):*
- *rest ,NSAIDS ,Antihistamine*
- *-severe visceral involvement may require high doses of corticosteroids with or without an immunosuppressive agent*
- -Immunosuppressive agents for rapidly progressive course and severe systemic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

- Henoch-Schönlein purpura (HSP):

- A specific type of cutaneous small-vessel vasculitis.



- Mostly affects children.
- May be preceded by symptoms of upper respiratory tract infection.
- Presents as multiple palpable purpura occur mostly in the lower legs, buttocks and to lesser extent on forearms.
- Could be accompanied by arthralgia, abdominal pain, nausea, vomiting, melena and kidney involvement with hematuria, proteinuria, or acute nephritis.
- In children, the onset is mostly after upper respiratory infection; association with hemolytic streptococcus has been pointed out.
- Drugs (penicillin, aspirin)
- These antigens combine with antibodies (mainly IgA) in the body, and the immunocomplex deposits on the vascular walls. Immunoreaction is induced to cause vasculitis and purpura.
- Pathology shows leukocytoclastic vasculitis with IgA deposition is observed by direct immunofluorescence.
- The histology of the kidney in HSP patients often shows crescentic glomerulonephritis.



- Treatment:

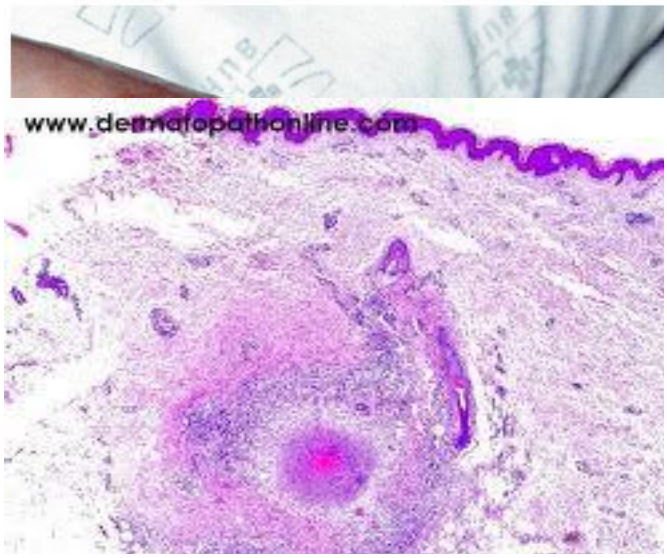
- Bed rest, pain relief and antibiotics if strep throat infection is present.
- Systemic steroids for severe cases with systemic involvement.
- HSP generally has a good prognosis and resolves within several weeks in most cases; however, it may recur.
- Serious complications may occur in other organs, such as nephritis with IgA deposition in the mesangium area, enterorrhagia, intussusception, intestinal perforation, or cerebral hemorrhage.
- Adults may have prolonged kidney involvement.

- Urticaria Vasculitis:

- Presents with urticarial weals that lasts more than 24 hours unlike urticarial, and usually leave hyperpigmentation after resolution.
- Could be associated with fever, arthralgia, abdominal pain and angioedema especially in hypocomplementaemic urticarial vasculitis that is associated with SLE.
- **Urticaria vasculitis has TWO types:**
  1- with low complement (check for SLE). 2- with normal complement.
- Causes include connective tissue diseases, viral and bacterial infections, drugs like ACE inhibitors, penicillin, sulfonamides, fluoxetine and thiazides. Also leukemia could cause it.
- Most cases are idiopathic and improve spontaneously after few months.
- Treatment is to remove the offending agents if present, treat the underlying diseases.
- Symptomatic therapy with antihistamines and NSAIDs.
- Dapsone, colchicine, hydroxycholorquine, steroids, azathioprine, and cyclosporine.

- Cutaneous Polyarteritis Nodosa:

- A rare form of vasculitis which involves small and medium sized arteries of dermis and subcutaneous tissue with unknown pathogenesis.
- Tender subcutaneous nodules and livedo reticularis (reticulated vascular pattern that appears as a lace-like purplish discoloration of the skin) that may ulcerate on legs and feet.
- Systemic involvement is uncommon except for peripheral neuropathy, and progression to classical polyarteritis nodosa is an exception.
- May have neuromuscular involvement in the form of peripheral neuropathy that presents with tingling, numbness, sensory disturbances, weakness, and



absent reflexes.

- In cutaneous PAN, histopathological examination shows features of nodular arteritis with polymorphonuclear infiltrates involving medium sized arteries in deep reticular dermis. There is extensive fibrinoid necrosis. This is in contrast to classical PAN which rarely shows nodular arteritis and the picture is of small vessel leukocytoclastic.
- Cutaneous PAN runs a chronic course lasting months to years, and has a waxing and waning phenomenon.

- Treatment:

- NSAIDs and oral steroids.
- Immunosuppressive drugs can also be used in low doses in more severe kinds of cutaneous PAN and as steroid-sparing drugs.

- Pigmented Purpuric Dermatoses:

- A group of chronic diseases of mostly unknown etiology characterized by extravasation of erythrocytes in the skin with marked hemosiderin deposition. (give orange color)
- Mostly affect males.
- Venous hypertension, exercise, and gravitational dependency are important cofactors that appear to influence disease presentation.
- **Histologically,** a perivascular T-cell lymphocytic infiltrate is centered on the superficial small blood vessels of the skin, which show signs of endothelial cell swelling and narrowing of the lumen.
- Extravasation of red blood cells with marked hemosiderin deposition in macrophages is also found but no vasculitis.
- The lesions are chronic and persist for years. With time, many of the lesions tend to extend and may become darker brown in color.
- No effective treatment available. It is a cosmetic problem mostly.

Orange color because of hemosiderin deposition