Dermatology Team 441

MED441
KING SAUD UNIVERSITY

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REVIEWED BY

Cutaneous manifestations of systemic diseases + Purpura & Vasculitis

Objectives:

- No objectives have been found for this lecture

Color index:

- Main text
- Important
- Dr's explanation
- Golden notes
- Extra

Dermatology Team 439

DERMATOLOGY TEAM 438

This lecture is originally done by both 438 & 439 teams.
So great thanks to them

Cutaneous Manifestations Of Systemic Diseases

- Skin is the gate of the body, you have to diagnose it early to prevent complications.
- Detection of Systemic disease or internal maligna through skin presentation.
- Systemic diseases associated with skin diseases.
- **the lecture contents will be classified into:**

- Connective Tissue Disease:
- Lupus
- Dermatomyositis
- Scleroderma

- GIT:
- Chronic Liver Disease (CLD)
- Acrodermatitis entropathica
- Peutz Jeghers Syndrome
- Pyoderma Gangrenosum

- Neurocutaneous diseases:
- Neurofibromatosis
- Tuberous Sclerosis

- Nutritional deficiency disorders:
- Scurvy
- Pellagra

- Cutaneous manifestations of internal malignancy.
- Acanthosis Nigricans
- Dermatomyositis

- Metabolic:
- Hyperlipidemia

- Endocrinological Diseases:
- Diabetes Mellitus
- Hyperthyrodism
- Hypothyeodism
- Cushing's Syndrome
- Addison's disease

- Behcet's Syndrome

- Causes of Generalized pruritus without skin lesions

- Nails:
- Clubbing
- Koilonychia
- Splinter haemorrhages

When to do HIV testing for skin Disease ?

Dr note: To describe any skin lesions you should mention:
1- If it is single / few / multiple / numerous.
2- Well / ill defined (if you can hold your pin and drow around it).
3- Configuration (e.g. Annular).
4- Color: erythematous / brown / blue
5- Hypo / Hyper / de (pigmented).
6- Secondary lesion if present.
7- Primary lesion.
8- Site of predilection.

Cutaneous Manifestations Of Systemic Diseases

Connective Tissue Diseases

Lupus

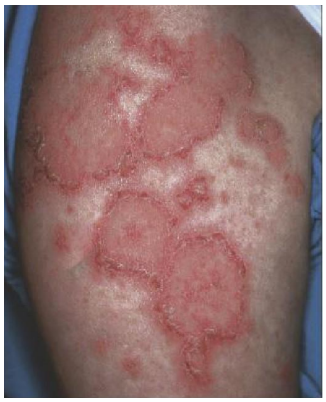
- **SLE:**
  - Facial photosensitivity.
  - Butterfly erythema (malar rash).
  - Multisystem disease (Renal, CNS, Cardiac, Blood, etc ...).
  - Positive ANA and Anti-dsDNA tests.
  - Oral and nasopharyngeal ulcers.
  - Scarring Alopecia.

- **Discoid Lupus (DLE):**
  - Round scarring lesions on light exposed areas.
  - No Systemic involvement.

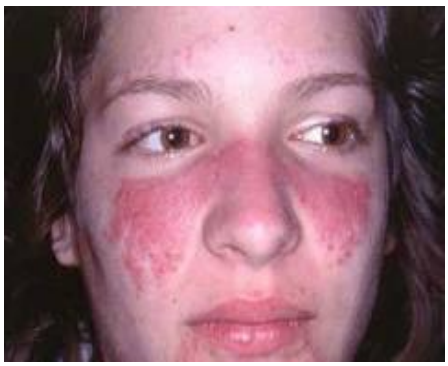
- **Subacute Cutaneous Lupus (SCLE):**
  - Papulosquamous or annular presentation.
  - Photosensitivity.
  - Does not cause scarring.
  - Usually **ANA negative but anti Ro positive.**

- **Neonatal Lupus:**
  - Appears in the first month in a photo-distribution.
  - Patterns (Papulosquamous and annular).
  - **Congenital heart block** (complete & permanent) usually needs pacemaker.
  - **Anti Ro positive.**

- **Drug-induced Lupus:**
  - **Procainamide and Hydralazine.**
  - Anti Histone positive.



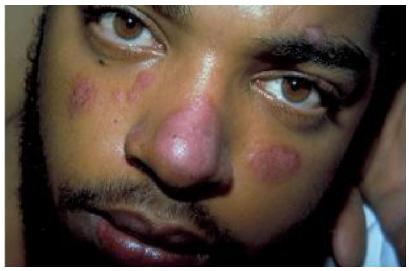
Multiple well defined Polycyclic or annular erythematous plaques with scaly border and some central clearing **(SCLE)**.



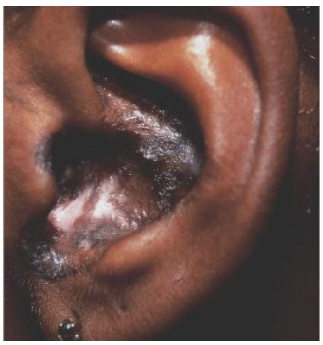
Well defined Erythematous patches and plaques over the cheeks (malar eminence) and over the nose sparing nasolabial fold.

Cutaneous Manifestations Of Systemic Diseases

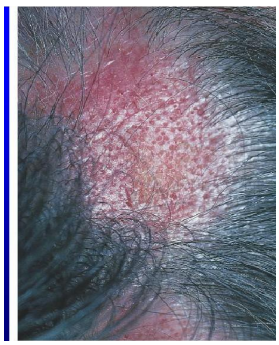
Lupus (cont'):



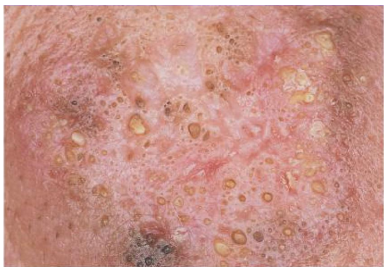
Few well defined erythematous to violesious discoid (Round not annular) plaques with loss of hair over the eyebrows **(DLE).**



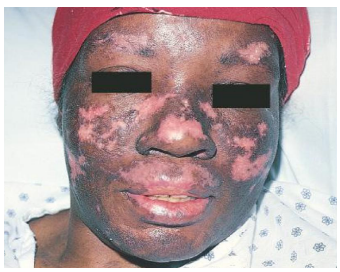
Discoid lesion with hyper & hypopigmentation.



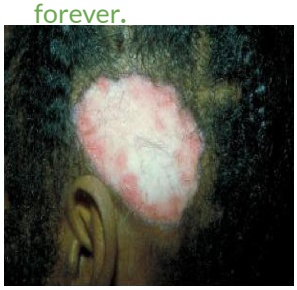
Follicular bulging (keratotic plugging), indicate early **(DLE),** so if you don't the patients earlier they well get scar and they will lose their hair maybe forever.



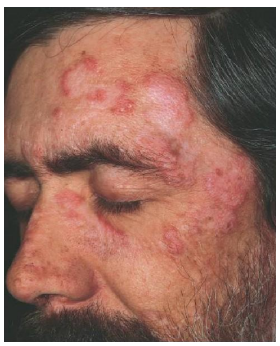
Extensive Follicular bulging.



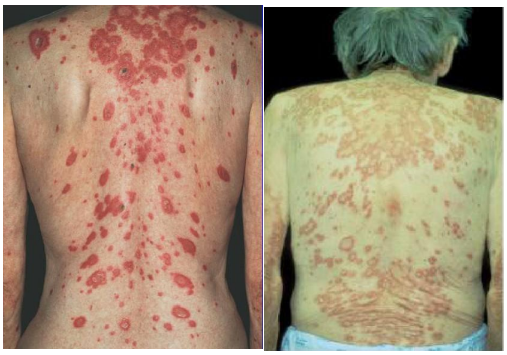
Dyspigmentation (hypo & hyper) with scaring, loss of vermilion border of the lip, also scarring alopecia on the eyebrow **(Aggressive DLE).**



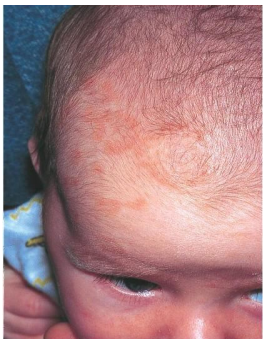
Big scarring alopecia with depigmentation and erythema .



Multiple polycyclic and annular scaly plaques **(SCLE).**



Psoriasiform annular scaly red lesions **(SCLE).**

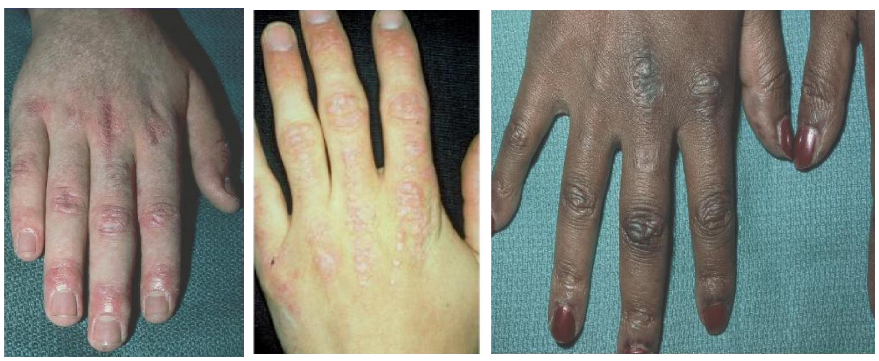


Annular lesions **(Neonatal lupus)**
Risk of heart block, so we have to check by ECG

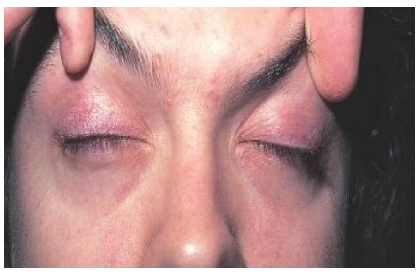
Connective Tissue Diseases (cont')

Dermato -myositis (Skin Rash + Muscle Weakness)

- **Heliotrope:** Violaceous color over the upper eyelids. .
- **Gottron's papules:** Flat- topped violaceous papules over knuckles of hands.
- Calcifications especially in kids.
- **Bilateral proximal muscle weakness** (with high CPK, positive EMG and muscle biopsy).
- In adults (especially over 50 years) associated with internal malignancy e.g. GI, Prostate, Ovary & breast. (A female above 50 newly diagnosed with dermatomyositis, you must rule out ovarian cancer first, then other types of cancer).



Violicious Gottron's papules



Heliotrope rashes

Cutaneous Manifestations Of Systemic Diseases

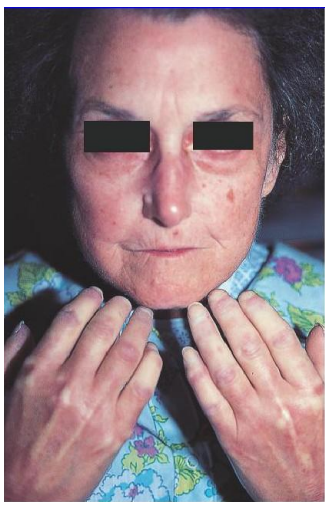
Connective Tissue Diseases (cont')

Scleroderma (systemic sclerosis)

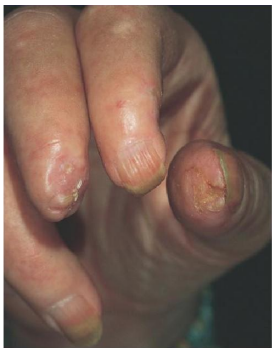
- **Scleroderma (Systemic sclerosis):**
  - Tight and thickened skin. Due to excessive collagen deposition
  - Sclerodactyly: fibrotic thickening and tightening of the skin of the fingers and hands
  - Face: Loss of forehead lines, beaked nose, small mouth, radial furrowing around the mouth.
  - Telangiectasia and calcification.
  - In diffuse type there is more systemic involvement (Lung, GI, Kidneys) and positive anti scl-70.

Other types of scleroderma include:

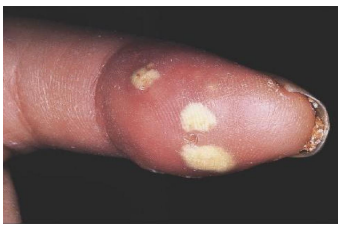
- **CREST:**
  - A milder type of scleroderma, **C** = **C**alcification, **R** = **R**aynaud's phenomenon, **E** = **E**sophageal dysfunction, **S** = **S**clerodactyly, **T** = **T**elangiectasia.
  - **Positive anti-centromere** with less systemic involvement.
- **Morphea:**
  - A localized scleroderma without systemic involvement, there is a firm, white patch of skin surrounded by violaceous ring.
- **En coup de sabre** (ضربة بالسيف) **:**
  - Linear scleroderma on the scalp and face which may give scarring alopecia and it may affect muscles or even bones.



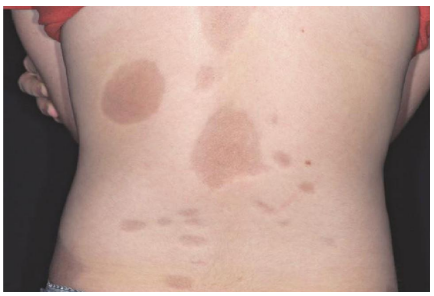
Minimal forehead wrinkle, very thick tight skin, peaked nose, small mouth, Sclerodactyly, Raynaud Phenomenon.



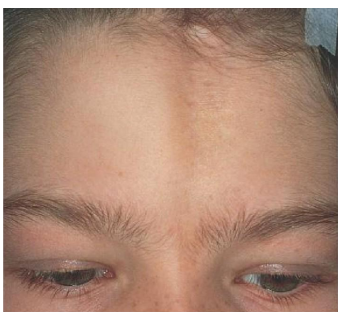
Sclerodactyly with infarction and loss of the tips of fingers.



Calcinosis cutis



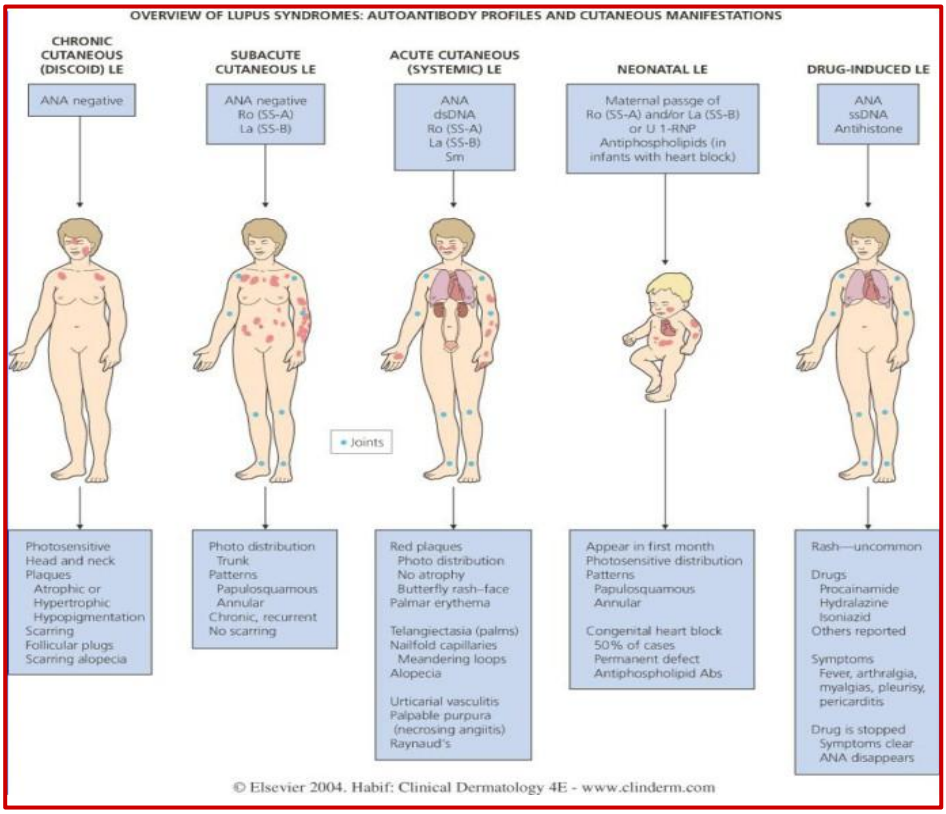
Well defined hyperpigmented atrophic plaque **(morphea).**



Deep morphea **(En coup de sabre).**

Antibody Testing in connective tissue diseases **(Important)**	
Antibody	**Clinical Significance**
ANA	Screening for SLE and other CTD
Anti-Centromere	Marker for CREST
Anti-Histone	Marker for Drug-induced Lupus
Anti-Smith	Specific for SLE
Anti-RNP	For Mixed CTD
Anti-Ro	Neonatal lupus, SCLE
Scl-70 Antibody	For Scleroderma
Anti dsDNA	For SLE

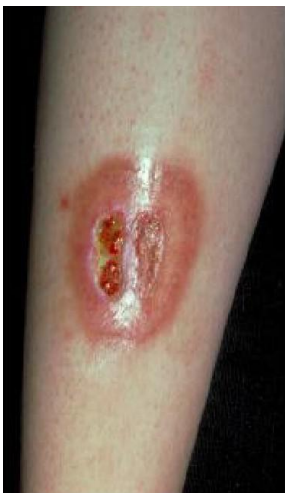
Cutaneous Manifestations Of Systemic Diseases

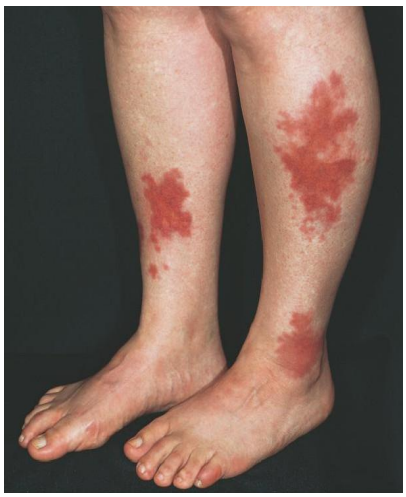


Endocrine system

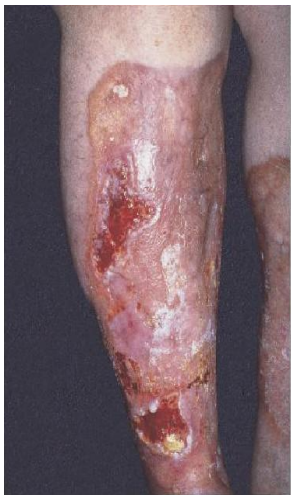
Diabetes mellitus

- **Necrobiosis lipoidica diabeticorum (NLD):**
  - Asymptomatic, usually seen on the shins. may progress to atrophy or ulcers)
  - May predate frank development of diabetes by several years (or with the onset DM).
  - **Shiny atrophic red or yellowish plaques with telangiectasia over their surface + ulceration.**
  - Severity of NLD is not directly related to severity of diabetes.
  - Increased risk of fungal and bacterial infection
  - Histopathology: shows tiered granulomatous Reaction.





Painless erythematous **orange** color plaques with **atrophy +** Telangiectasia and it will end up with ulceration.



Advanced NLD.

Cutaneous Manifestations Of Systemic Diseases

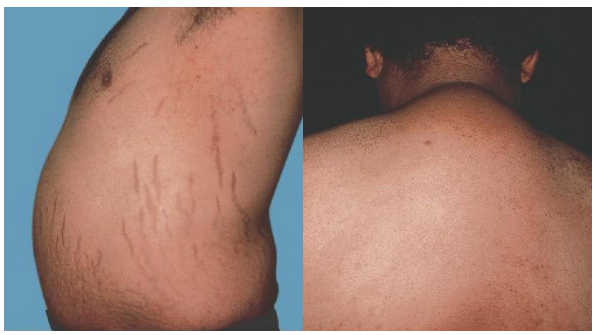
Endocrine system

Thyroid disorders

- **Hyperthyroidism:**
  - Smooth, warm, moist (due to increase sweating) skin and pruritus.
  - Pretibial myxedema (asymptomatic red plaques over shins).
  - Thin & fine hair.
  - Onycholysis (Brittle nails).
  - Clubbing.
  - Hyperpigmentation or vitiligo
- **Hypothyroidism:**
  - Dry and cold skin and pruritus.
  - Edematous skin (myxedema).
  - Hair loss of lateral third of eyebrows.
  - Brittle hair or nails.
  - A yellowish hue to the skin due to carotenaemia.
  - Delayed wound healing.

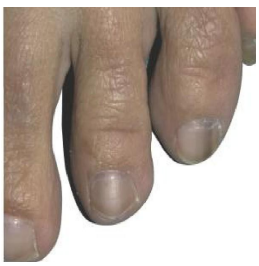
Cushing's Syndrome

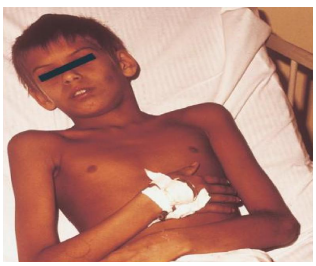
- Rounded face with fullness of cheeks (Moon face).
- Buffalo hump (fat deposit over upper back).
- Central obesity with thin arms & legs "lemon with sticks".
- Atrophy of skin & Striae.
- Purpura.
- Hirsutism & Acne.
- Clitoromegaly and male pattern alopecia (Hamilton pattern).



Addison's disease

- **Hyperpigmentation** at Sun exposed skin, sites of trauma, axillae, palmar creases, old scars, nevi and mucous membranes.
- Diffuse pigmentation on skin and mucous membranes.
- Melanocytes stimulation by ACTH
- Patients usually present with lethargy, postural hypotension, and hyperpigmentation





GIT

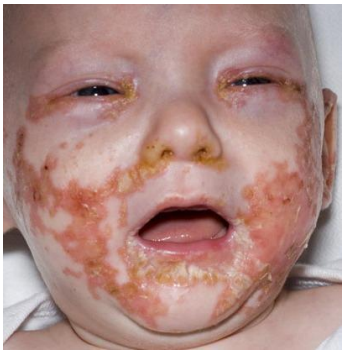
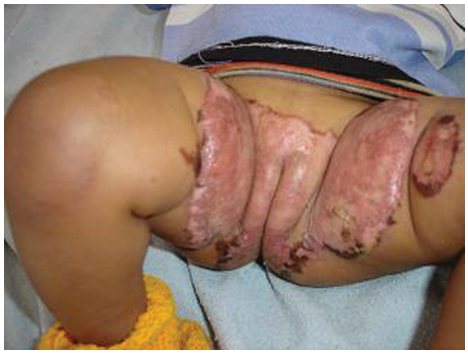
Chronic Liver Disease

- **Jaundice, spider telangiectasia, acne, gynaecomastia, purpura, collateral veins, striae, palmer erythema, dupuytren's contracture, and white nails.**

Acrodermatitis enteropathica (Genetic disease)

- Due to **zinc** deficiency (a rare autosomal recessive disorder that impairs dietary zinc absorption in the jejunum and ileum).
- Seen in infants as inflammatory erythematous erosive scaly rash (patches and plaques similar to atopic dermatitis, but progress to vesicles, crusts, erosions, and pustules) around orifices (mouth, ears, anus) also on hands & feet.
- Alopecia and nail dystrophy.
- **Diarrhea** or abdominal pain.
- Treated with lifelong zinc supplementation (Pt will improve after 2,3 days

Cutaneous Manifestations Of Systemic Diseases

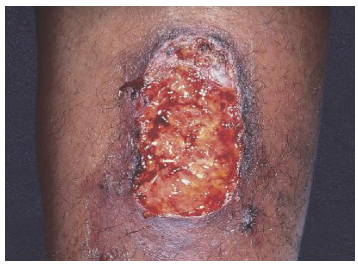



Multiple well defined crusted honey color erosions around the mouth and groins and it will be treated with zinc supplements.

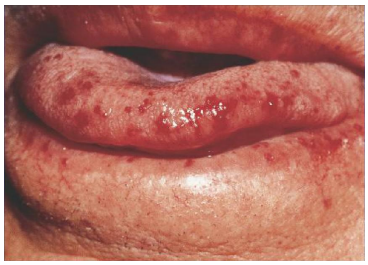
GIT

Peutz Jeghers syndrome	• Small **brown macules** on lips, buccal mucosa and small intestinal polyps. (Asymptomatic except for skin manifestations) • The skin findings first appear in infancy or early childhood. • Rarely polyps can be pre malignant. (Request colonoscopy)
Pyoderma gangrenosum	• Acute painful leg ulceration, surrounded by violaceous border. • Start as small pustules, which subsequently burst and expand to form the larger noninfectious ulcer. • Associated with Inflammatory bowel diseases, rheumatoid arthritis and leukemia.
Hereditary hemorrhagic Telangiectasia	• Telangiectasia (dilated capillaries) over lip, nose, tongue, fingers and toes. • Hx of recurrent epistaxis. • Associated with recurrent upper GI bleed.

Peutz-jeghers syndrome.



Single well defined ulcer with violaceous border.

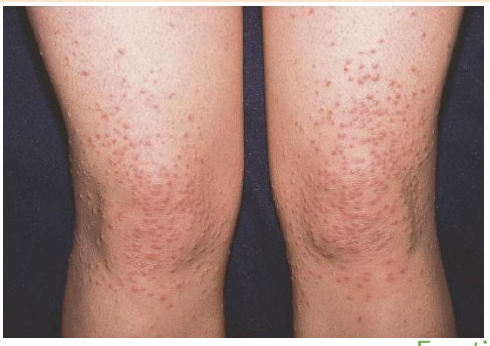
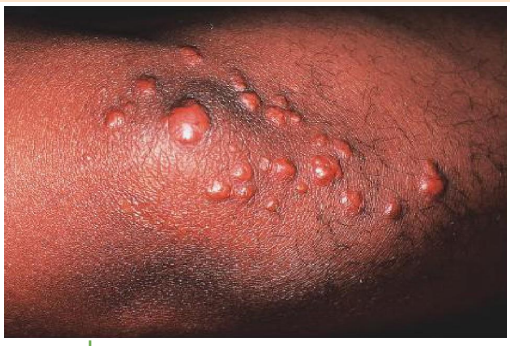


Small telangiectasia with bleeding.

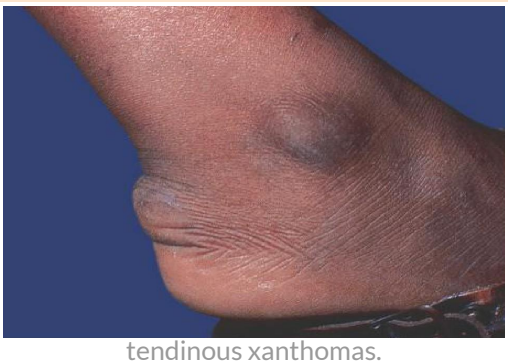
Hyperlipidemia

Hyper -lipidemia	• Present with different types of xanthomas. • Yellow color is characteristic. **Xanthoma may be a pointer to:** • Primary hyperlipemic status due to genetic abnormality. • Secondary hyperlipemic status due to renal, hepatic, endocrine or pancreatic disease. • **Normo-lipemic status.**
Types of Xanthoma	• **Eruptive:** small papules appear in crops over buttocks & extensors. • **Tendinous:** Nodules over tendons e.g. extensor tendons of hands & feet and Achilles tendon • **Palmar crease xanthoma:** on palms. • **Tuberous:** Papules & nodules over knees and elbows. • **Xanthelasma:** Bilateral symmetrical over both eyelids.

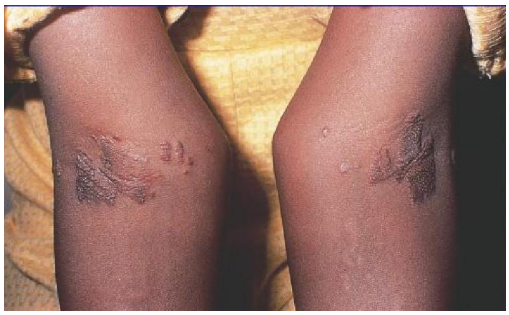
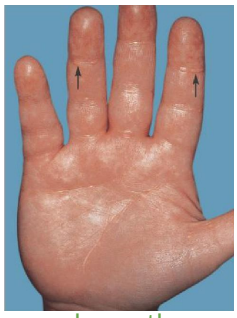
Cutaneous Manifestations Of Systemic Diseases

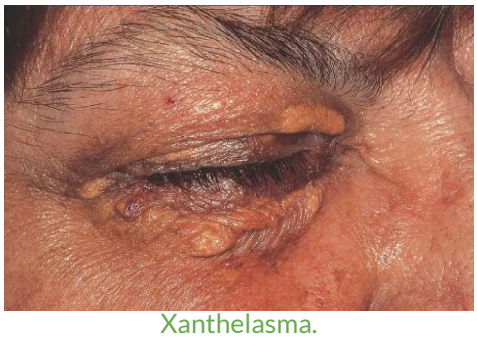
Eruptive papules.



tendinous xanthomas.

Orange color on the creases.

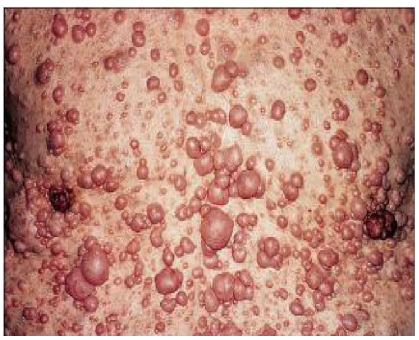


Xanthelasma.

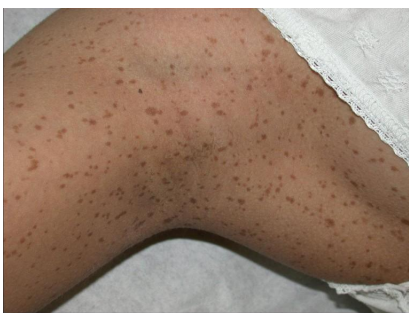
Neurocutaneous Disorders

Neuro -fibromatosis

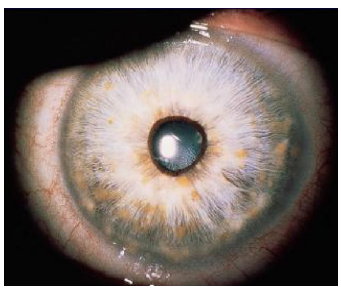
- Autosomal dominant.
- Café-au-lait macules (light brown) can be seen in other disorders but if big size and number suspect neurofibromatosis.
- Neurofibromas (soft pink or skin- colored papules and nodules).
- Axillary freckling (Crowe sign), the presence of the macules on the axillae.
- Optic glioma.
- Lisch nodules (iris hamartoma, seen by slit-lamp examination).
- Associated with Neurological complications e.g. tumors, seizures and mental retardation.



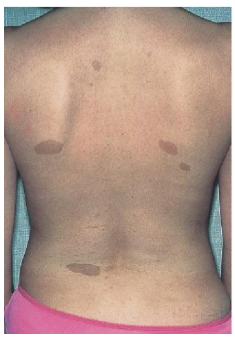
Soft innumerous well defined pink papules and nodules.



Axillary freckling.



Lisch nodules



Café-au-lait macules

Neurocutaneous Disorders (cont')

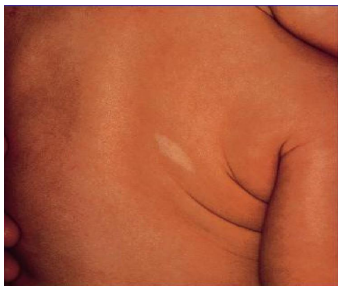
Tuberous Sclerosis (Epiloia)

- **Epi** = Epilepsy / **Loi** = Low intelligence / **A** = adenoma sebaceum.

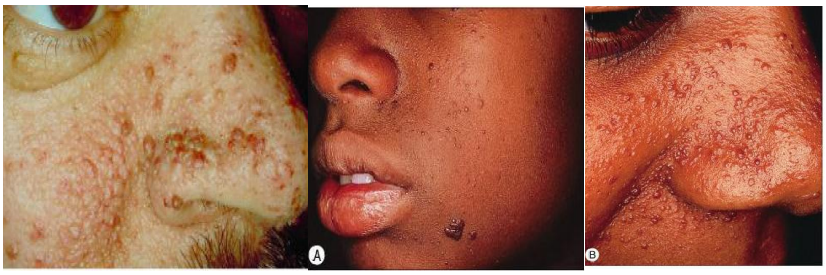
Skin Features:

- Adenoma sebaceum (angiofibroma): red papules around the nose and on chin.
- Ash-leaf hypopigmentation: oval area of hypopigmentation **(This is the earliest sign of TS).**
- **Periungual fibroma:** multiple papules & nodules around the nail **(Pathognomonic)**.
- Shagreen patch: skin colored plaque on the trunk with "orange-peel" Surface.

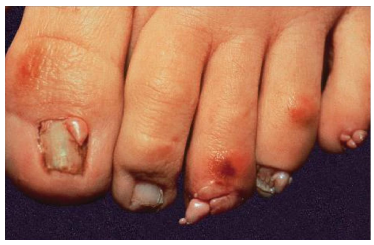
Cutaneous Manifestations Of Systemic Diseases



Ash-leaf hypopigmentation.



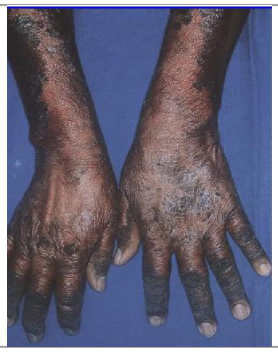
Adenoma sebaceum (angiofibroma).



Periungual angiofibroma.

Others

Behcet's syndrome	• Oral ulcer (the most common). Painful • Genital ulcers (mainly scrotal). • Iritis and arthropathy (May lead to blindness). • May have CNS involvement.
Scurvy اسقربوط	• Vitamin C deficiency. • Bleeding gums. • Can cause teeth loss (permanent complication). • Easy bruising, they bleed easily through their gum. • Diagnosis: Low ascorbic acid (Vit-C) level in Leukocyte. • Perifollicular hemorrhage and corkscrew hairs (Very brittle hair).
Pellagra	• Nicotinic acid deficiency. **4 "D" s :** • Dermatitis (Photodermatitis). • Diarrhea • Dementia • Death (if not treated)



Causes of Generalized Pruritus Without Skin Lesions (Important)

Causes of Generalized Pruritus Without Skin Lesions	• **Endocrine:** DM, hypo & hyperthyroidism. • **Hematological:** polycythemia rubra vera, iron def anemia. • **Malignancy;** e.g. Lymphoma. • **Hepatic:** primary biliary cirrhosis. • **Renal:** CRF (The commonest manifestation of CRF is pruritus). • **Neurological:** e.g. Tabes dorsalis. • **Others: Psychogenic, Drugs, Idiopathic.**

Table 7.2 Possible laboratory studies in the evaluation of pruritus. These laboratory tests should be performed according to the patient's history, in particular in cases of generalized pruritus of unknown etiology.

POSSIBLE LABORATORY STUDIES IN THE EVALUATION OF PRURITUS
• Erythrocyte sedimentation rate (ESR) • Complete blood cell count (CBC) with differential and platelet count • Blood urea nitrogen, creatinine • Liver transaminases, alkaline phosphatase, bilirubin • Fasting glucose • Thyroid function tests (thyroid stimulating hormone (TSH) and thyroxine levels) • Parathyroid function (calcium and phosphate levels) • Serum iron, ferritin • Chest radiograph • Stool for ova, parasites and occult blood
• Viral hepatitis screen • Serum protein electrophoresis • Serum immunofixation • Antinuclear antibodies (ANA), antimitochondrial antibodies
• Human immunodeficiency virus (HIV) • Allergy panel: total IgE, histamine, serotonin (plasma) • Prick tests of major atopy antigens and additives, patch tests • Urine for sediment, 5-hydroxyindolacetic acid (5-HIAA) and mast cell metabolites • Additional radiographic studies, e.g. abdominal CT scan • Anti-tissue transglutaminase antibody • Anti-smooth muscle antibody

Dr says: Labs are important

Cutaneous Manifestations Of Systemic Diseases

Erythema Nodosum

Erythema Nodosum

- Multiple ill-defined bilateral tender erythematous subcutaneous nodules over shins.
- More common in females.

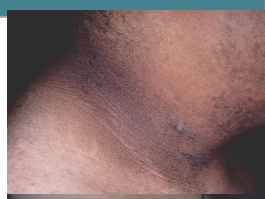
Causes:

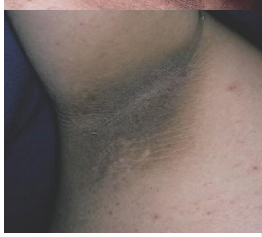
- Infectious: Streptococcus, Tuberculosis, Hepatitis, Chlamydia.
- Sarcoidosis.
- Drugs: Oral contraceptive pills, sulfonamides.
- Lymphoma & leukemia.
- Pregnancy.
- Behcet's disease.
- Idiopathic.

Acanthosis nigricans

Acanthosis nigricans

- Ill defined Brown hyperpigmentation & increased thickening of skin with **velvety texture** at neck, axillae and groin (seen more in body folds).
- Treat the underlying disease (bleaching agents do not help)
- Whenever you see Acanthosis nigrican extensively involving The palms think of malignancy, we call it trip palm





Causes:

- Obesity.
- Endocrinopathy: Diabetes, Thyroid disease, Insulin resistance.
- Internal malignancy: the most common is adenocarcinoma of stomach.
- Drugs: Nicotinic acid.
- Familial.
- Idiopathic.

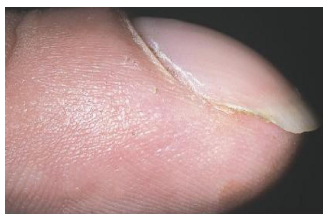
Nails

Clubbing

- Exaggeration of the normal nail curve associated with loss of the normal angle between nail and posterior nail fold.

Causes:

- Thoracic: Lung abscess, Lung CA.
- CVS: Congenital cyanotic heart disease.
- GIT:GI carcinoma, Inflammatory bowel disease.
- Endocrine: Thyroid disease.
- Idiopathic.



Causes of clubbing, From KSUMSC basic clerkship guide

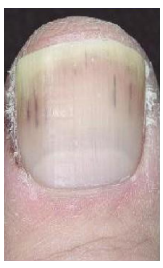
CLUBBING:

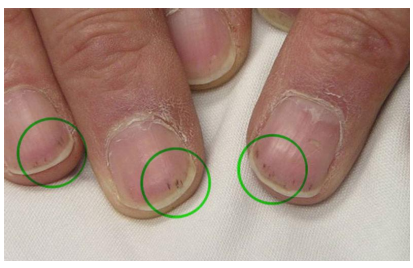
- **C**yanotic congenital heart disease
- **L**ung abscess
- **U**lcerative colitis & crohn's disease
- **B**ronchiectasis, Bronchogenic carcinoma
- **I**nfective endocarditis
- **N**othing (Idiopathic)
- **G**raves

Splinter Haemorrhages

Causes:

- Bacterial endocarditis.
- Septic emboli.
- CTD.
- Trauma.
- Idiopathic.



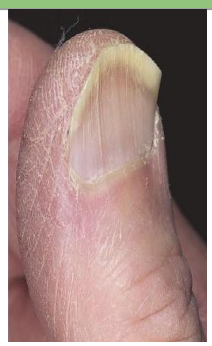


Cutaneous Manifestations Of Systemic Diseases

Nails (cont')

Koilonychia

- Spoon- shaped appearance.
- Causes
  - Iron deficiency anemia.
  - Thyroid disease.
  - Physiological; early childhood.
  - Dermatosis:
    Lichen planus, Alopecia Areata and others .



Some mucocutaneous disorders in which you need to do HIV testing

Oral hairy leukoplakia

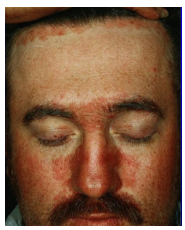
- Corrugated white plaques on the lateral aspect of the tongue.

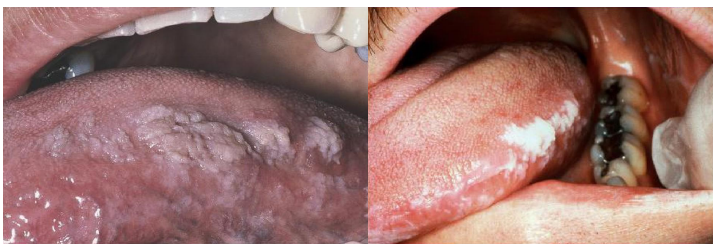
Kaposi Sarcoma

- Caused by HHV -8 (not all are associated with HIV).
- Blue macules, patches or nodules which is in essence a vascular tumor.
- Associated with low CD4 count.
- May resolve or diminish if CD4 count rises
- Types of Kaposi sarcoma: Classic type (in elderly), Immunosuppression associated, HIV associated and African endemic type.
- Metastasis to Lymph nodes, and Viscera.
- Severe seborrheic dermatitis not responding to medications.

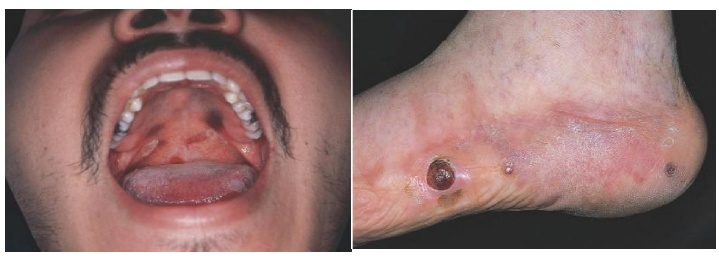
Others

- Multiple mollsucum contagiosum in adult (on face).
- Any STD.
- Seborrheic Dermatitis (extensive & refractory to therapy).
- Severe extensive recalcitrant aphthous ulceration.





Oral hairy leukoplakia



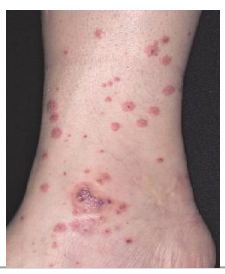
Kaposi sarcoma

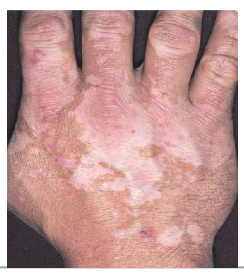
Examples of some skin diseases where you may find systemic associations:

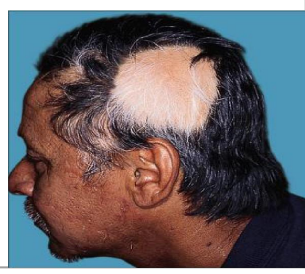
Examples of some skin diseases where you may find systemic associations

- **Lichen planus**; associated with Hepatitis B and C.
- **Vitiligo and Alopecia Areata**: both associated with autoimmune diseases like: Autoimmune Thyroiddis, Diabetes mellitus, Pernicious anemia, Myasthenia gravis, etc.

Lichen planus







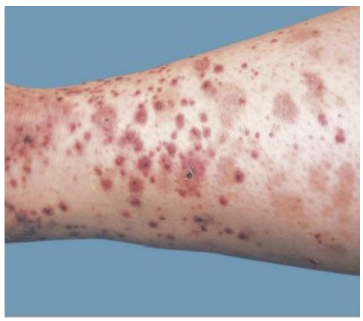
Purpura And Vasculitis

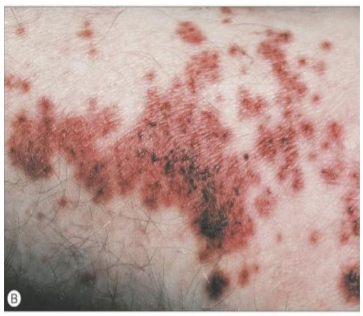
❖ Purpura

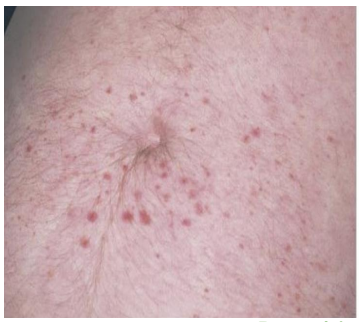
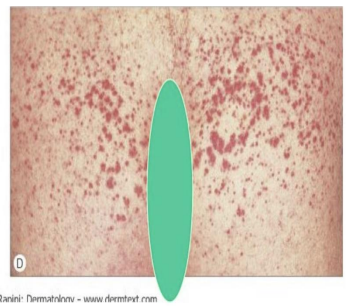
- **Definition**: Visible hemorrhage into the skin or mucous membrane.
- It's a condition of red (Dusky red) or purple discolored spots on the skin that do not blanch (doesn't disappear) on applying pressure .
- if the red spot is blanchable (goes away), then the diagnosis will be Erythema (Dilated vessels).
- You have to use glass, because you will not be able to see it if you apply pressure by your hand.
- The spots are caused by bleeding underneath the skin secondary to platelet disorders, vascular disorders,coagulation disorders or other causes.

They are subdivided as a follow:

- **Petechiae:** less than or equal 3 or 4 **mm**.
- **Purpura:** (>3 or 4 **mm** - <1 **cm**) which can be either Palpable or nonpalpable (macular)
  Palpable purpura is vasculitis until proven otherwise. الدكتور يقول لو ما تطلع من المحاضرة هذي إلا بهذه المعلومة لكفتك. (مجازاً طبعاً لا تصدق)
- **Ecchymoses:** > or equal to 1 **cm**.


Purpura


Ecchymoses: تكون كبيرة



Petechiae: تكون صغيرة

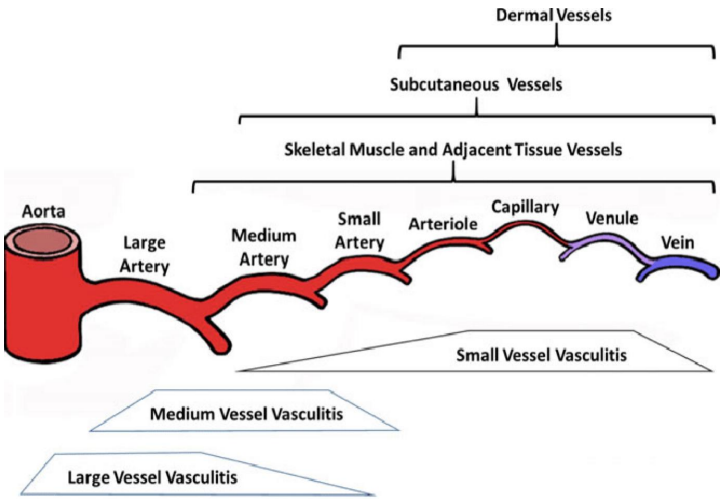
Causes	
Platelet Disorders	- Thrombocytopenia. - Platelet dysfunction.
Coagulation Factor Deficiency	• **Congenital:** - Factor VIII Deficiency. - Factor IX Deficiency. - Von Willebrands disease. • **Acquired:** - Disseminated Intravascular. - Coagulopathy. - Liver disease. - Uremia. - Vitamin K deficiency
Vascular factors	• **Congenital:** - Hereditary Hemorrhagic Telangiectasia. - Ehlers-Danlos Syndrome (Type IV). • **Acquired**: - Inflammation (Vasculitis). - Trauma. - Vitamin c deficiency (scurvy).

Purpura And Vasculitis

❖ 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.
- palpable purpura : If you closed your eyes and put your hands you can feel it

- **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.



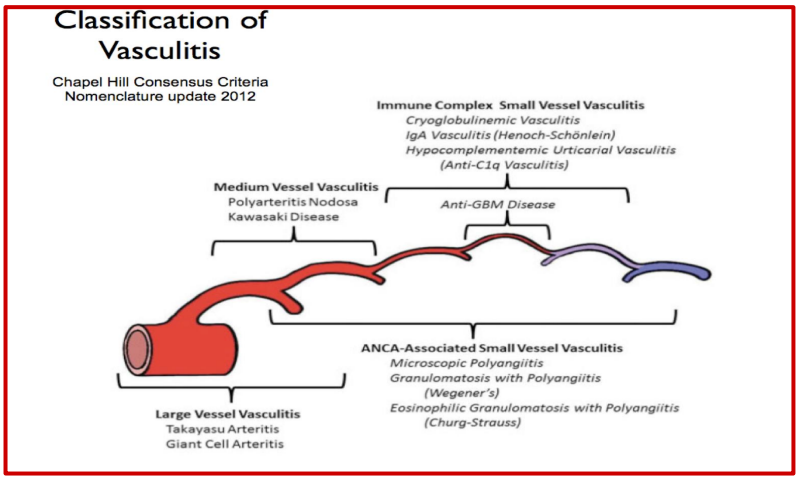


Table 26.2 Chapel Hill consensus classification.

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

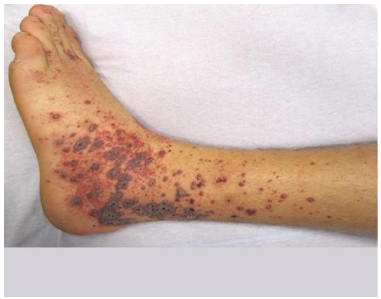
Table 3. Causes of cutaneous vasculitis[5,8]

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

More important than your name
Dr: لا تقولون محد علمنا

Purpura And Vasculitis

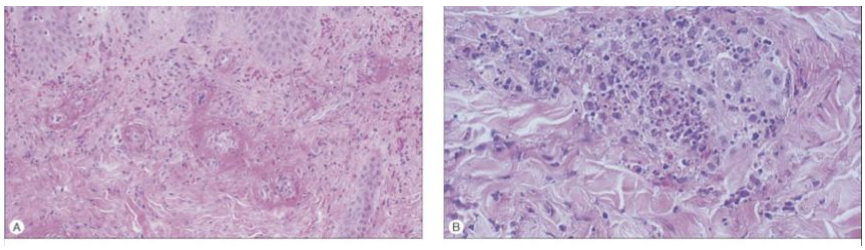
Cutaneous small vessel vasculitis :



Hemorrhagic blisters and Palpable Purpura on lower extremities

- Most common type of vasculitis and it primarily affect post-capillary venules.
- **Pathogenesis:**
  - Many forms of small-vessel vasculitis are felt to be caused by circulating immune complexes.
  - These lodge in vessel walls and activate complement.
- Palpable purpura is the hallmark.
- Pinpoint to several centimeters (Can present as erosion and ulceration).
- Early on lesion may not be palpable, Papulonodular, vascular, bullous, pustular or ulcerated forms may develop.
- Predominate on the ankles and lower legs i.e. dependent areas.
- 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 (may lead to ESRD).
- Renal involvement present as glomerulonephritis.
- The prognosis is good in the absence of internal involvement

- **Histology :**
  - Angiocentric segmental inflammation, endothelial cell swelling, fibrinoid necrosis of blood vessel walls and a cellular infiltrate composed of neutrophil with RBC extravasation.
  - inflammatory Cells inside and outside the blood Vessels.



- **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, PROTEIN & CAST
  - Skin biopsy most important tool to confirm 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
Cardiovascular	Electrocardiogram, creatine phosphokinase, echocardiogra
Neurologic	Nerve conduction studies
Musculoskeletal	Electromyography

DIAGNOSTIC WORKUP

Laboratory investigations

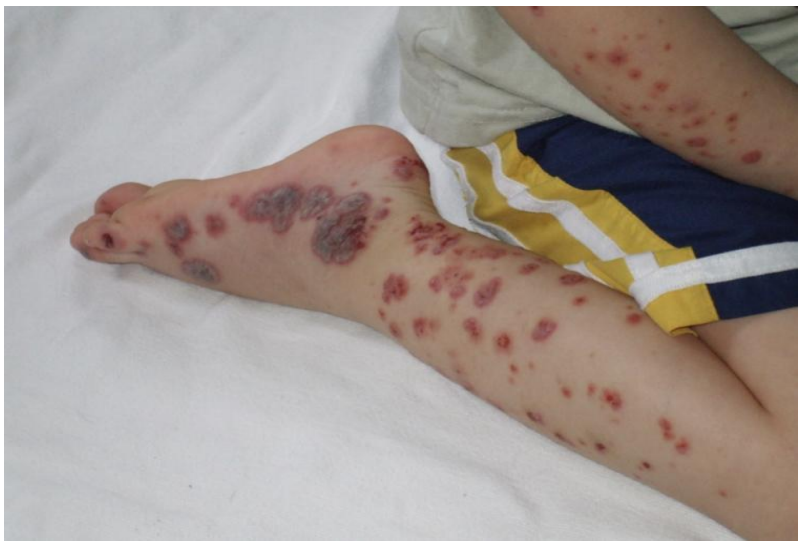
- Full blood count with differential white cell count
- Markers of inflammation: ESR,CRP
- Electrolytes and hepatic transaminases, glucose
- Urinalysis for protein and blood
- Blood cultures (if pyrexial)
- Serology—ANA ,dsDNA , ANCA,C3 and C4,ASLO titre, viral titres (e.g. hepatitis B and hepatitis 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 Systemic corticosteroids with or without an immunosuppressive agent.
  - Immunosuppressive agents for rapidly progressive course and severe systemic involvement e.g Kidney.

Purpura And Vasculitis

❖ Henoch -Schön lein 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 (Good prognosis unless the kidney is affected).
- 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).



Quiz!

1- Which of the following skin rashes is associated with diabetes mellitus?

A) Acanthosis nigricans
B) Palpable purpura
C) Pretibial myxedema
D) Striae

2- Which of the following is not an indication for HIV testing?

A) Any STD
B) Oral hairy leukoplakia
C) Seborrheic Dermatitis
D) lichen planus

3- What is the best treatment for Cutaneous small vessel vasculitis without systemic involvement?

A) Methotrexate
B) Bed rest
C) Steroid
D) Immunosuppressive agents

4- What is pathognomonic sign for small vessel vasculitis?

A) Vesicles
B) Bulla
C) Palpable purpura
D) Scar

Answers:
1 : A , 2 : D , 3 : B , 4 : C

Thanks!!

DERMATOLOGY TEAM 438

Dermatology Team 439

438 Team leader:
Mohsen Almutairi

439 Team leader:
Mohammed Albabtain

438 Done by:
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