DERMATOLOGY

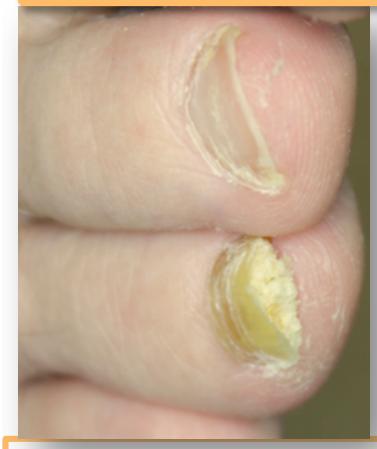
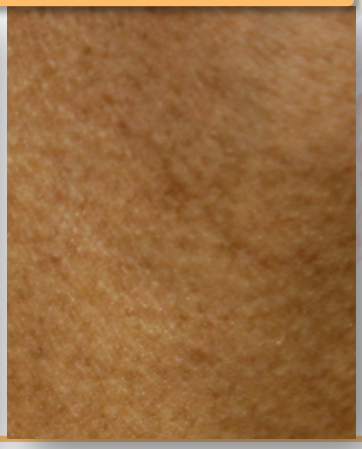
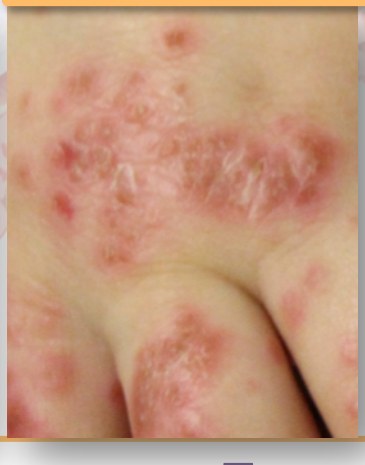
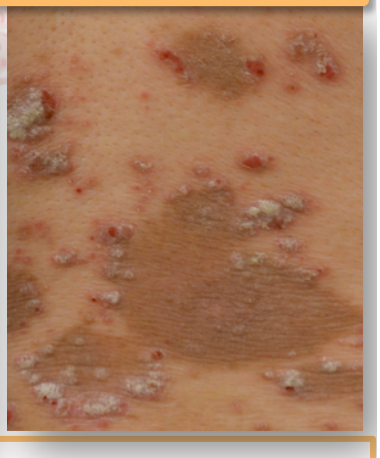
Cutaneous Manifestations of Connective Tissue diseases

DONE BY

Musab Almoshiqeh

TEAM LEADER

Meshaal AlOtaibi

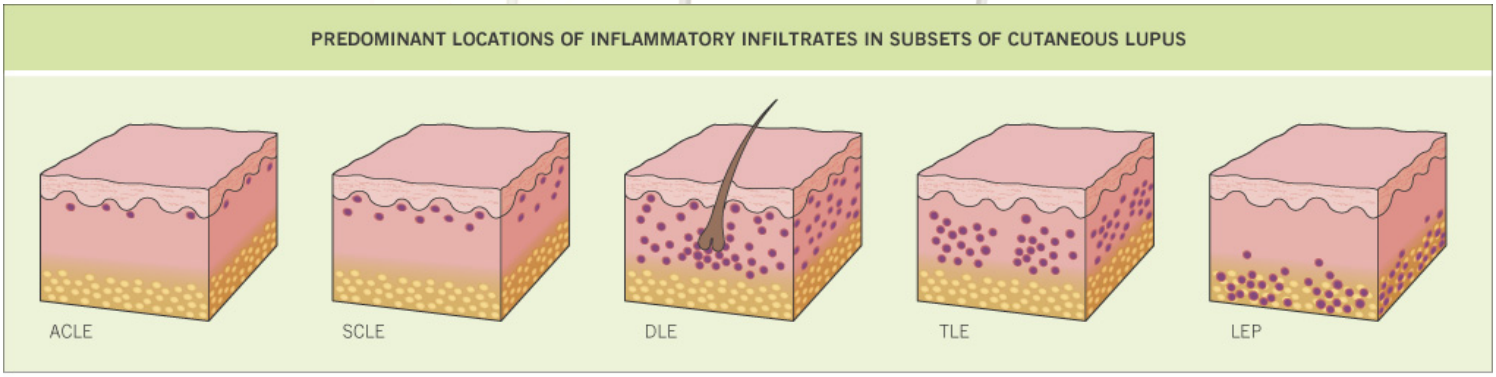





FROM SLIDES DOCTOR'S NOTES BOOK NOTES TEAM' NOTES IMPORTANT

Lupus Erythematosus :

- **Introduction:**
- LE is as an autoimmune diseases associated with antibodies directed against components of cell nuclei .
- Lupus may affect any tissue, skin, kidneys, CNS, lungs and others.
- Cutaneous lupus subtypes: " Each one has its own appearance not related to each other, but may it overlap"
- **Types:**

✓ Discoid lupus erythematosus
✓ Subacute lupus erythematosus
✓ Neonatal lupus erythematosus
✓ Lupus tumidus
✓ Lupus profundus
✓ Chilblain lupus erythematosus
✓ Drug-induced lupus erythematosus
✓ Systemic Lupus Erythematosus



ACLE : Redness , scale , malar rash, the pathology is superficial , close to epidermis .
SCLE : Redness , scale , superficial not cause scarring .
Discoid : more deep " the inflammation seen in the dermis , the pathology in the dermis and surrounding the hair follicle " important " it make alopecia and scarring .
Tumid : " meaning swelling " Dermal proses without any epidermal change " no scale".
Profundus : affect the subcutaneous layer " appear as nodules ' then it become atrophy .

❖ Discoid Lupus Erythematosus : Discoid meaning "coin shape"

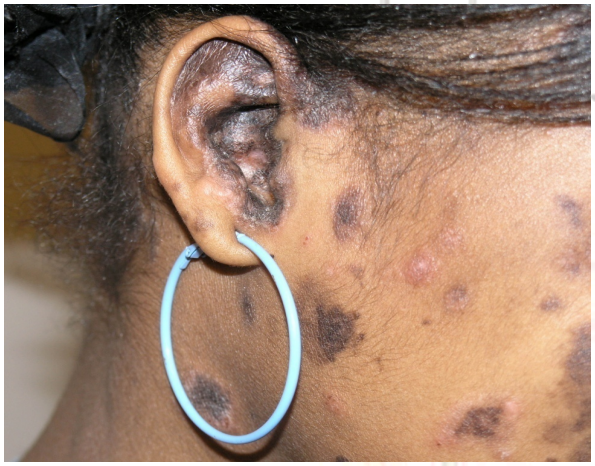
- **Definition :** It is the commonest form of cutaneous lupus (purely coetaneous 90% of cases) usually presents as red scaly patches or plaques (on light exposed areas (photosensitivity)) , that leave dyspigmentation and scarring (Mostly Hypopigmented or depigmented scars) .

- Features and distribution :
- The lesion may be localized or widespread.
- Usually affects the cheeks, nose and ears, but sometimes involves the upper back, neck, and backs of hands.

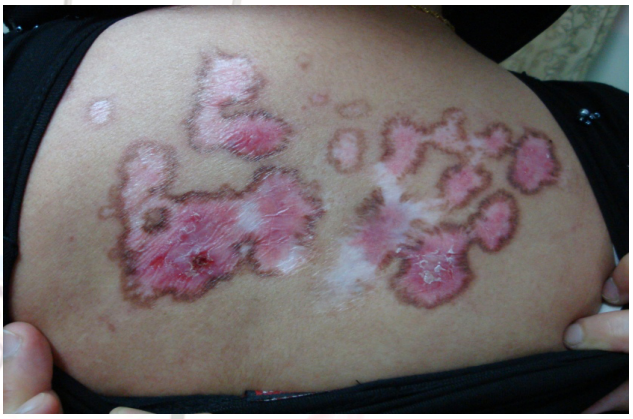
- Involvement of hair follicles will lead to scarring alopecia.

- **Diagnosis :**
- Mostly diagnosed by morphology.
- Need skin Biopsy to confirm the diagnosis.
- Serologic abnormalities are uncommon.
- Discoid lupus one of the criteria of SLE , but can be present alone and 10% of DLE patients develop SLE. Because of that "Need to check ANA regularly and follow the patient every 3-6 months "why ??" to avoid systemic involvement and early control of SLE and prevent its complications.

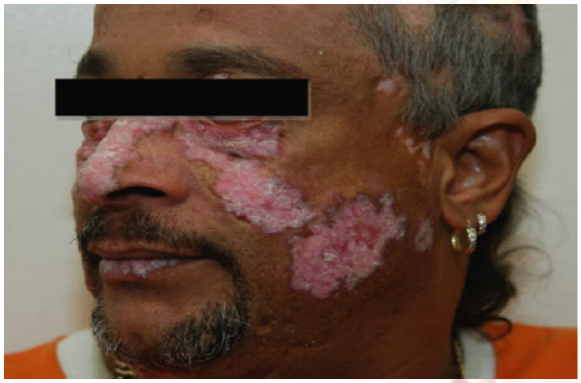
- **Treatment :**
- Moderate to strong topical steroid and intralesional steroid , avoid the strong steroid if the lesions in the face, around the eye and the axilla to avoid atrophy.
- Resistant cases: immunosuppressive (azathioprine, cyclosporine a, mycopheolate).



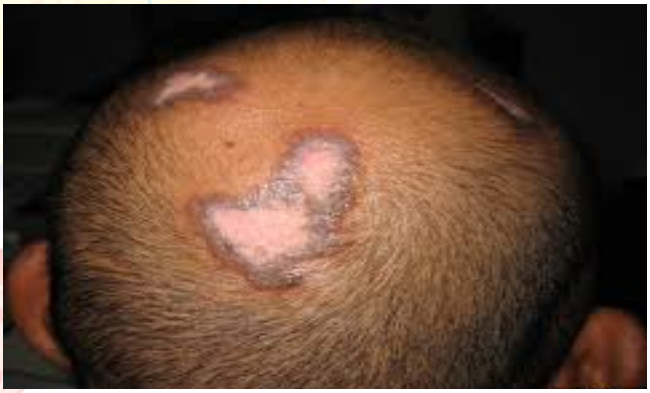
Multiple patch, scared hyperpigmemnted lesion "depend on patient color"
Darky skin " hypopigmentation "



Multiple Red scaly plaques hyperpigmented margin, hypopigmented center with some redness.
As the lesion progresses, the scale may thicken and become adherent to underlying epidermis (carpet-tack sign), and run into hair follicle



If the lesion affect the face "which most likely " need to treat it very aggressively with systematic medication from the beginning to prevent scarring formation.



If the lesion involve the scalp it will lead to Scarring Alopecia.

❖ Subacute lupus erythematosus :

- **Definition** : Non-itchy dry rash (macule and plaques) appears on the upper back and chest, often following sun exposure.
- Photosensitivity is prominent in about half of patients.
- Subacute LE does not cause scars.
- Systemic involvement is not usually severe." Usually limited"

- **Morphology** :

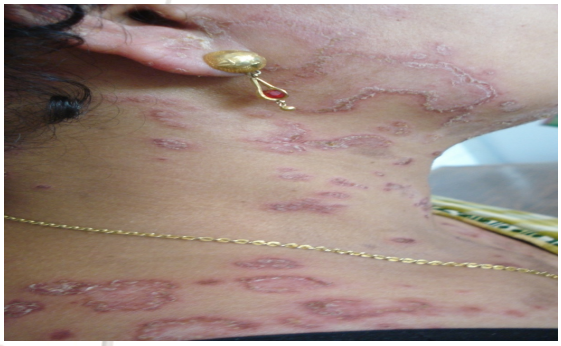
Annular or polycyclic (ring-shaped) or as papulosquamous (scaly patches and plaques)

- **Diagnosis** :
- A strong association exists with anti-Ro (SS-A).
- Anti Ro/La antibodies, is nearly always present in patients with subacute LE.

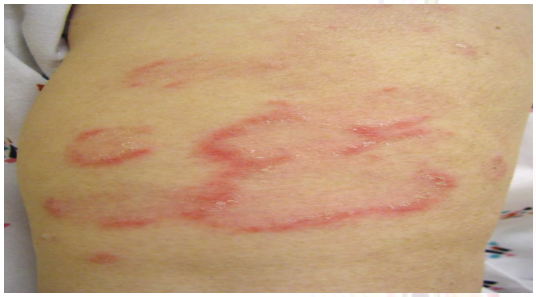
- Biopsy and immuoflurescence studies are helpful.

- **Treatemnt** :

Topical creams and systemic antimalarial.
Systemic steroid may be needed.



Annular and polycyclic , erythematous , scaly affect the face and upper trunk



May appeare as psoraiziform or as Annular and polycyclic appearance

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❖ Neonatal lupus erythematosus : " rare but important to know"

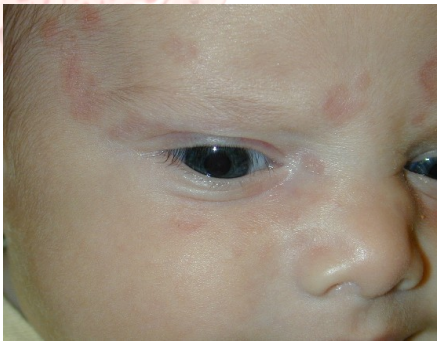
- **Definition** : annular erythematous plaques of newborn babies (usually) born to mothers with subacute LE, develop annular rash, that resolve spontaneously (Usually for babies
- of Anti-Rho +ve mothers)

- **Location** :

Appear predominately on the scalp, neck, or face (typically periorbital in distribution)

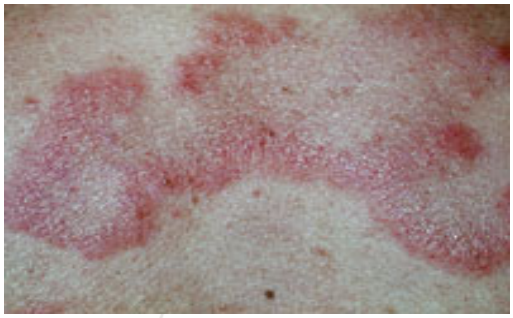
*** The neonates could be at risk of complete heart block.

(You must refer patients with neonatal lupus to cardiologist or ask for ECG)



Annular and semiannual, Erythematous lesion , usually on the face .

❖ Lupus Tumidus : "Tumidus means swelling"



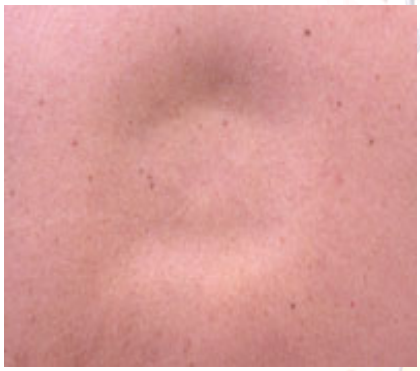
- **Definition**:
  Red, swollen, urticarial-like bumps and patches or swelling, The rash is Characteristically photosensitive, so it affects sun-exposed sites.

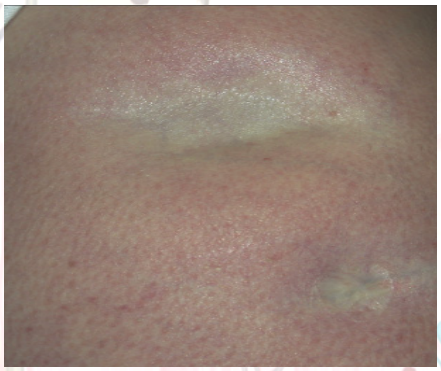
- **Treatment**:
  Topical corticosteroids are not effective as treatment for LET, but many will respond to chloroquine.

Swelling without scale " look like Urticarial"

❖ Lupus Profundus : "Profundus means deep"

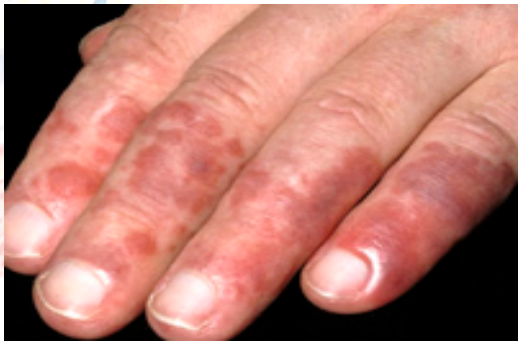
- **Definition** :
- lupus affecting the fat underlying skin lupus panniculitis (other name of lupus profundus), Inflammation of the fat results in firm deep nodules for some months . It may develop at any age, including children. The face is the most common area to be affected.
- The end result is deep scars on fat layer or lipodystrophy.





Appeare as nodule " because of the inflammation " then appeare as depression as " Lipodestrophy"

❖ Chilblain Lupus Erythematosus : "usually on cold area"

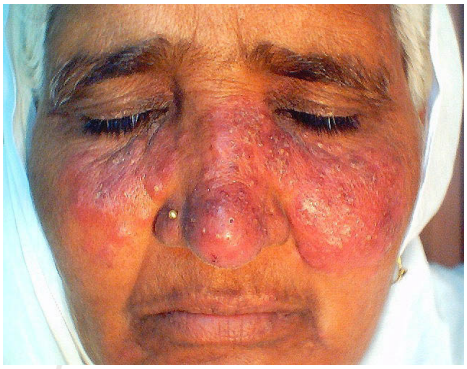


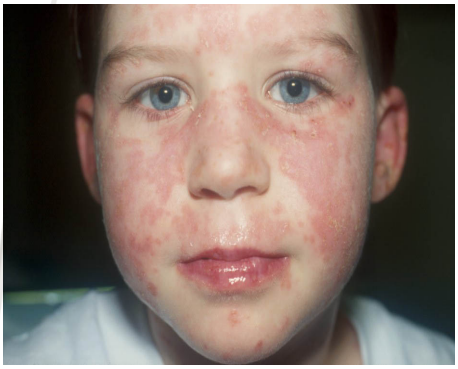
- **Definition**: Itchy and/or tender red or purple bumps that usually come on from cold exposure but can sometimes be precipitated by sun exposure or smoking.
- They are considered to be a form of skin vasculitis (blood vessel inflammation).
- Usually they have no circulating antibodies.
- The main treatment is to avoid precipitating factors.

❖ Drug-Induced Lupus Erythematosus : "usually no skin manifestation"

- Drug induced lupus does not usually affect the skin. The most frequent drugs are: Hydralazine, Carbamazepine , Lithium , Phenytoin , Sulphonamides , Minocycline.

❖ Systemic Lupus Erythematosus :





Force the check and cross the nose bridge with spare of nasolabial fold , get worse with sun exposure .

- Only a few patients with cutaneous LE also have SLE.
- The most common presentation is with a malar eruption or butterfly. (Typical presentation)
- Other skin changes in SLE Criteria are photosensitivity, mouth ulcers, and diffuse hair loss.
- For hair loss:
  Usually Patchy, diffuse hair loss is a recognized feature. But, permanent hair loss & scarring is unusual (seen on CDLE) .
- SLE may also affect joints, kidneys, lungs, heart, liver, brain, blood vessels and blood cells.

❖ Investigations :

- Serology:
  1/ ds DNA , Anti Smeth
  2/ SLE is always with positive ANA.
- Skin biopsy may be diagnostic especially in discoid lupus erythematosus with SLE.
  (Acute Lupus usually skin biopsy is not characteristic)
- Direct immunofluorescence tests may show positive antibody deposition along the basement membrane (lupus band test)
- CBC : to see leucopenia , thrombocytopenia, neutropenia .
- Urine analysis, and kidney function test
- And the rest depend on the case and severity of the disease. (For example kidney biopsy for in sever form of Lupus)

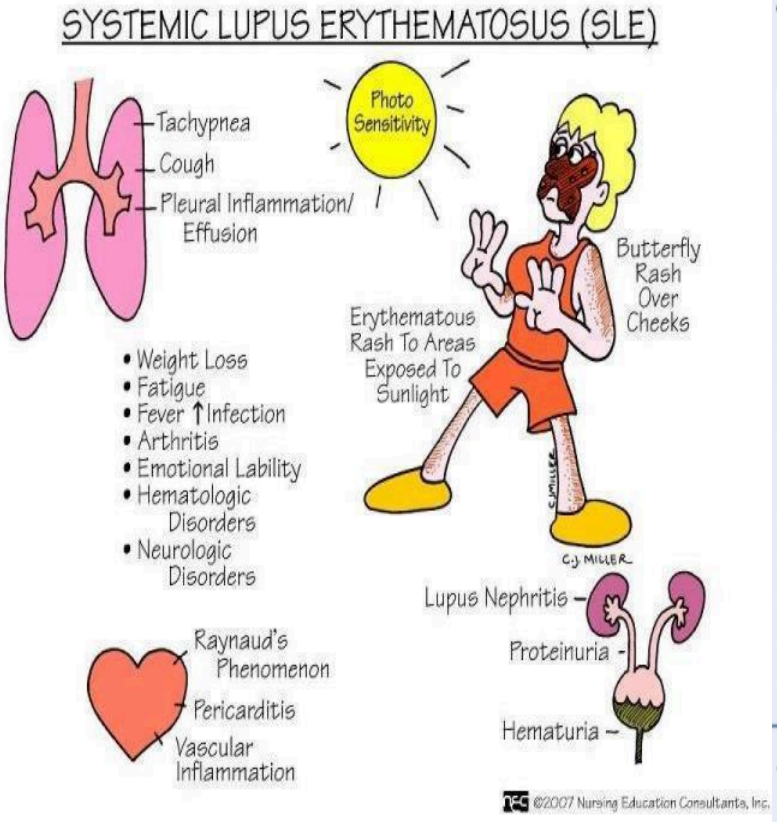


Table 3. ACR Classification Criteria for SLE

Criterion
• Malar rash—also known as *butterfly rash*—is a red and mildly scaly rash that forms over the bridge of the nose and cheeks
• Discoid rash—thick red scaly patches on the skin
• Photosensitivity
• Oral ulcers
• Arthritis
• Serositis—inflammation of the serous tissues of the pleura, pericardium, and peritoneum
• Renal disorder
• Neurologic disorder
• Hematologic disorder
• Immunologic disorder
• Antinuclear antibody

ACR: American College of Rheumatology; SLE: systemic lupus erythematosus.
Source: Reference 15.

❖ **Treatment : (depend on the case and surface area involved)**

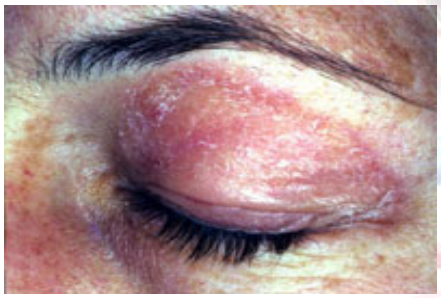
- The aim of treatment for cutaneous LE is to alleviate symptoms and to prevent scarring.
- Smoking cessation will help Raynaud's phenomena and chilblain lupus.
- Sun protection with sun scream . (To avoid photosensitivity)
- Potent topical steroids, Intralesional steroids. (but in the face when don't use super potent steroid because it may cause thinning of the epidermis, folliculitis &telangiectasias).
- Oral antimalarial drugs.
- Oral steroids.
- Immunosuppressive medication : Methotrexate, azathioprin, mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG, and Rituximab.

Dermatomyositis:

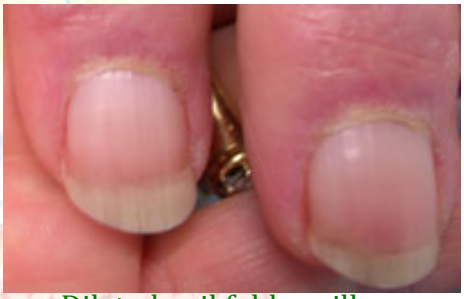
- Definition:
- It is a rare disorder of skin and muscle characterized by a specific rash and proximal weakness.
- An uncommon inflammatory disease affects adults between 40-60 (females mainly) and children 5-15. (The myopathy can come on the onset of skin manifestation or before or after) .
- Some agents may trigger the disease include penicillamine, statin drugs, quinidine, and phenylbutazone.

- Feature and morphology:
- The main symptoms include skin rash and symmetric proximal muscle weakness, which may be accompanied by pain.

- The Rash : A violet-colored or dusky red rash on face and eyelids and on areas around nails, knuckles, elbows, knees, chest and back.
- The rash, which can be patchy with bluish-purple discolorations, is often the first sign of dermatomyositis.



Heliotrope rash: a violaceous -to-dusky erythematous rash with or without edema in a symmetrical distribution involving periorbital skin .



Dilated nail fold capillary
Under magnifying glass
Many kind of disease can present with it

- **Muscle weakness : (**Defect comb hair Defect claim the stairs) :

Progressive symmetric proximal muscle weakness involves the hips, thighs, shoulders, upper arms and neck.
The weakness is symmetrical and more in the extensor muscles.

- Other signs and symptoms include:

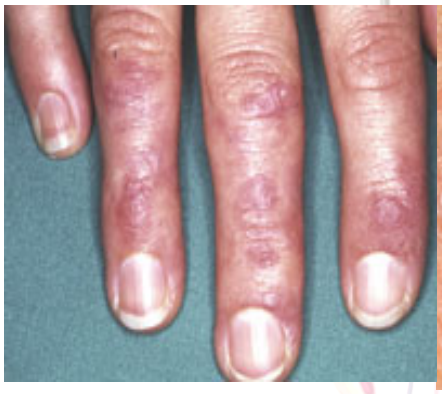
- Photosensitivity :

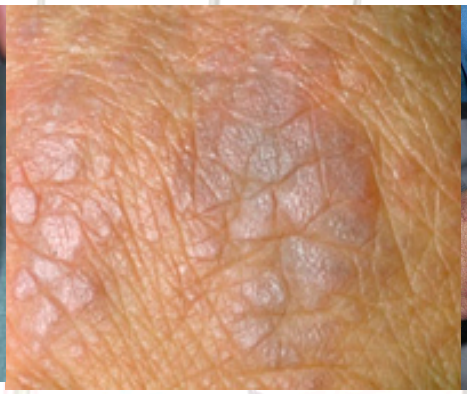
Photosensitivity + telengactesia which could be a sign of malignancy (especially in GIT malignancy + and to some extent ovarian cancer & prostate and breast)

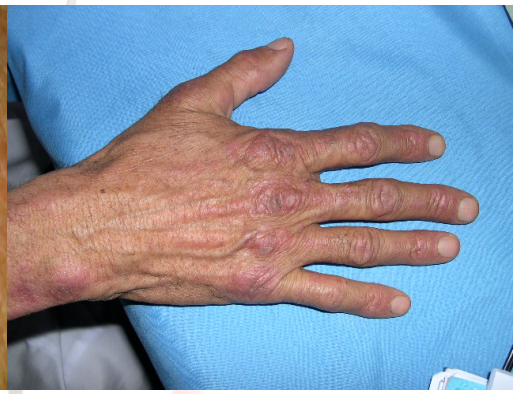
- Raynaud's phenomenon
- Dysphagia, gastrointestinal ulcers
- Muscle pain or tenderness
- Fatigue, fever and weight loss
- Calcinosis cutis especially in children
- Interstitial lung disease.

Raynoid phenomenon :

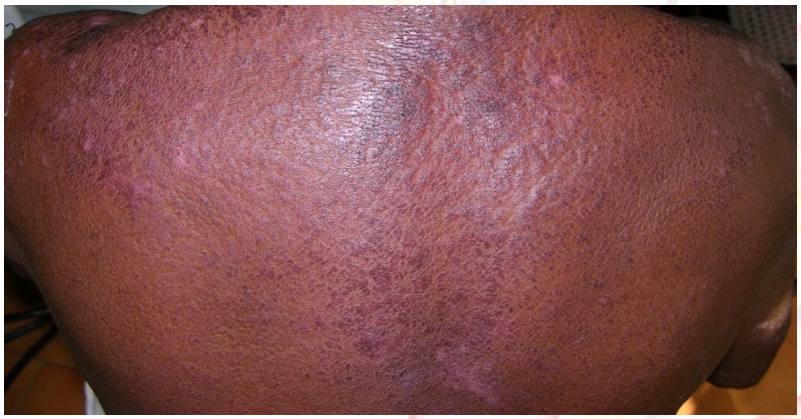
Change of skin in expose to cold (white " vasoconstriction" then blue "cyanosis" then red)
* many kinds of disease can present with Raynoid phenomenon (smoking , SLE , Scleroderma)



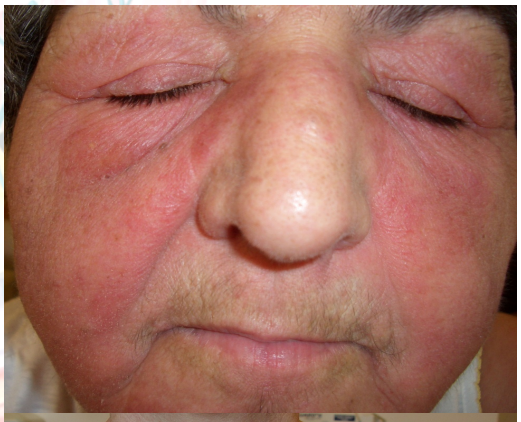


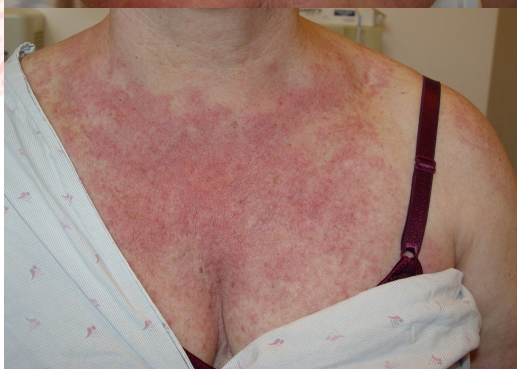


Gottern papule and sign : a pink to reddish purple atrophic or scaling eruption often over the knuckles, knees and elbows.



Shawl sign : shoulders or in a "V" over the posterior neck and back or neck and upper chest, which worsens with UV light. Erythroderma is not a flat, erythematous lesion similar to the shawl sign .





Erythematous, fallacious rash, scaly

- Dermatomysitis : can be associated with:
- Other connective tissue diseases such as lupus, rheumatoid arthritis, scleroderma and Sjogren's syndrome.
- **Cancer,** Especially in <u>older patients</u>, particularly of the

cervix, lungs, pancreas, breasts, ovaries and gastrointestinal tract.

- Cancer could precede, coincide or follow the diagnosis of DM.
- It's important to screen malignancies in patients who are:
  i. older than 50 years .
  ii. had a history of symptoms of malignancy.
  iii. Proximal muscle weakness (triceps , quadriceps) .

- **Investigations :**
- Muscle biopsy.
- Electromyography. (EMG)
- Magnetic resonance imaging (MRI).
- Blood tests: creatine kinase (CK) (Most Specific) and aldolase. Increased CK and aldolase levels *can indicate muscle damage and CK is useful to monitor the treatment of DM.*

- Serology (Autoantibody) :

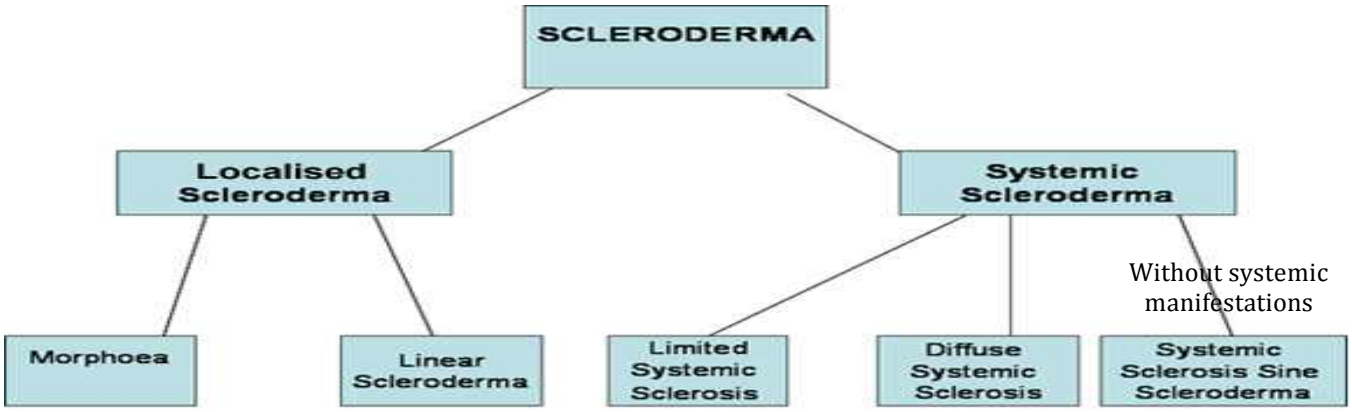
a. Anti-Jo-1 (anti-histidyl-t-RNA synthase) in 30%. It is specific, and has a bad prognosis, because it is associated with pulmonary disease.
b. Anti-Mi-2 is highly specific for DM, but it lacks sensitivity because its
c. Present only 25% of patients with DM, indicates good prognosis.

- Skin biopsy is suggestive but not diagnostic that shows interface dermatitis. (to support the diagnosis , you see inflammatory cells very vascular . Just to tell you it is not other disease)

- **Treatment :**
- Oral steroids are the mainstay treatment.
- Steroid sparing agents are:
  Methotrexate, azathioprin, mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG, and Rituximab.
- Topical steroids and antimalarial medications are used to improve the cutaneous rashes. (if there only skin signs) " can appear as dermatomyositis without myositis " skin manifestation only.
- Physiotherapy to improve strength and flexibility of the muscles.
- Surgical excision or Co2 laser could be utilized to remove tender calcium deposits.
- If there malignancy you need to refer it to oncologist and treat it.
  *** How to treat Raynaud Phenomenon :
  1- Avoid cold exposure .
  2- CCB ,
  3- Control the primary cause

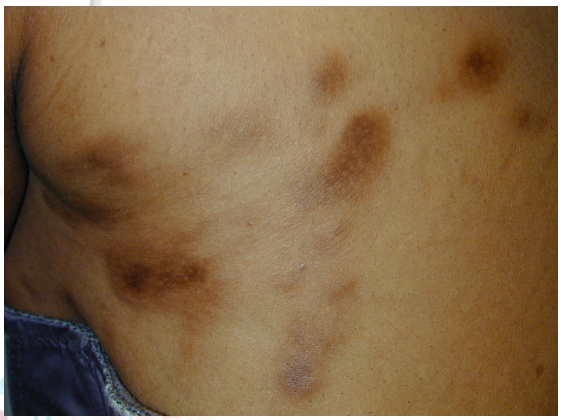
Scleroderma: (hardening and tightening of skin and shinny without wrinkles)

- **Definition:** A group of rare diseases that involve the hardening and tightening of the skin and connective tissues .
- Scleroderma affects women more often than men and most commonly occurs between the ages of 30 and 50.

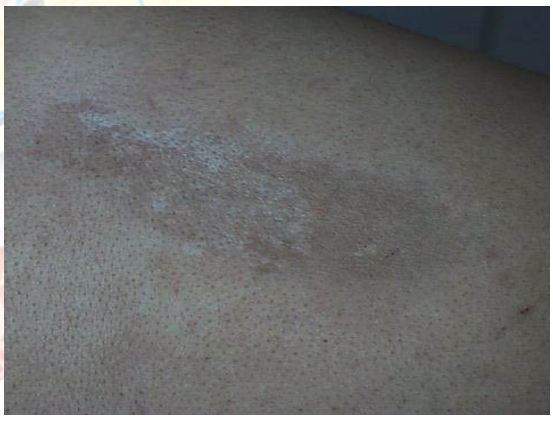


- Localized scleroderma usually affects only the skin on the hands and face. Its course is very slow, and it rarely, if ever, spreads throughout the body (becomes systemic) or causes serious complications.
- There are two main forms of localized scleroderma: Morphea and linear scleroderma

➢ Morphea :" localized patches"



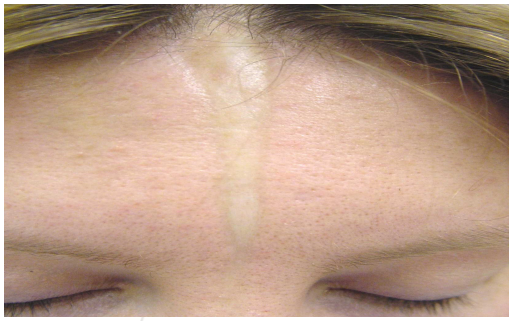
- **Definition:**
  A rare skin condition that causes oval reddish or purplish patches and plaques on the skin.
- Scar like band or plaque on any body site.
- Sometimes in linear distribution on face and extremities.
- It subsides on its own over time leaving dyspigmentation and scars.
- Unlike systemic sclerosis, Morphea lacks features such as Sclerodactyly, Reynaud phenomenon, nail fold capillary changes, telangiectasias, or progressive internal organ involvement.



- Confirmed by skin biopsy (diagnosis), which usually shows thickening of collagen bundles and loss of skin appendages like sweat glands and hair follicles
- No known cure.
- Treatment of morphea focuses on controlling signs
- and symptoms and slowing spread.
- Topical (local) and intralesional steroids ,
- phototherapy, systemic(Generalized) steroids,
- azathioprine, methotrexate, and cyclosporine
- might be used in severe cases.
- Physical therapy could be of help if the involvement
- is close to joints and cause contracture and difficulty
- movement.

Ill-defined, sclerotic patch, sometime be a white , and after resolution it is become depression and atrophy , end up by pigmentation .

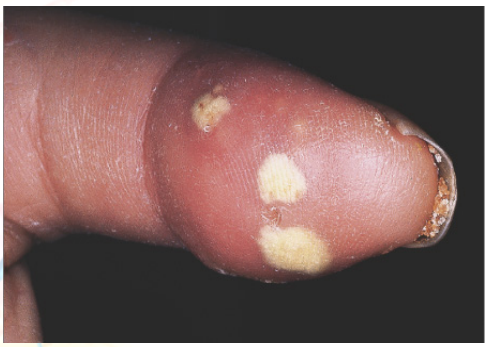
➢ Linear Scleroderma : "Line like appearance" , Consider a type of Morphea :



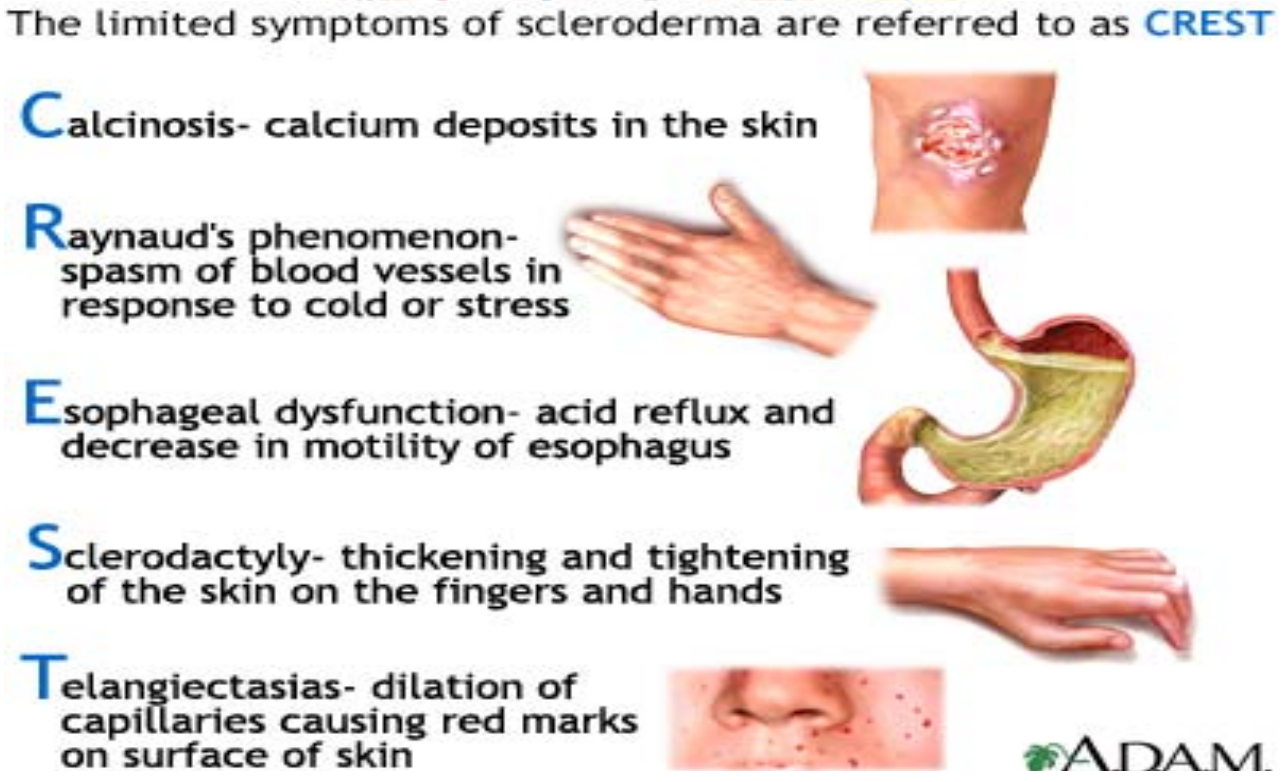
Depressed atrophic linear plaque cause alopecia and may cause growth irregularities of the skull in children

- **Definition:** Linear lesions extend to length of arms or leg , Begin first decade of life
- type of linear scleroderma that presents on the frontal or frontoparietal scalp due to inflammatory disease (Morphea).
- May also occur parasagitally down the forehead, known as en coup de sabre.
- Histology: Morphea, there's loss of all skin appendages which is the major feature in histology.
- **Remember:** Morphea + internal organ involvement = systemic sclerosis , it can affect the bone and muscle If it is so severe .
- **Treatment :** Potent corticosteroids (topical or intralesional)à if active.

➢ Limited Systemic sclerosis (CREST Syndrome) :

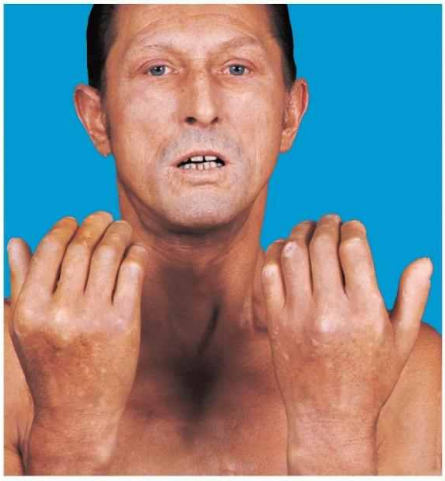


- Is a limited form of systemic sclerosis in which there is **C**alcinosis, **R**aynaud's phenomenon, **E**sophageal involvement, **S**clerodactyly and **T**elangiectases.
- *Anticentromere antibodies are characteristic for this syndrome.*

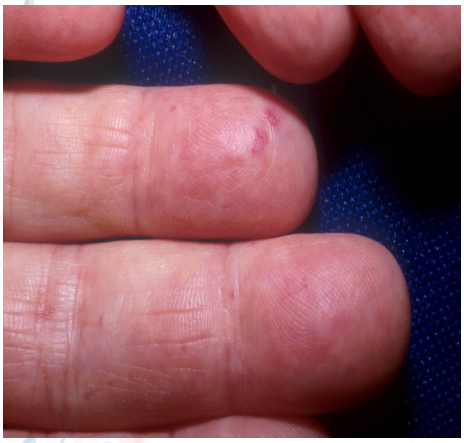


Systemic Sclerosis : (more than CREST)

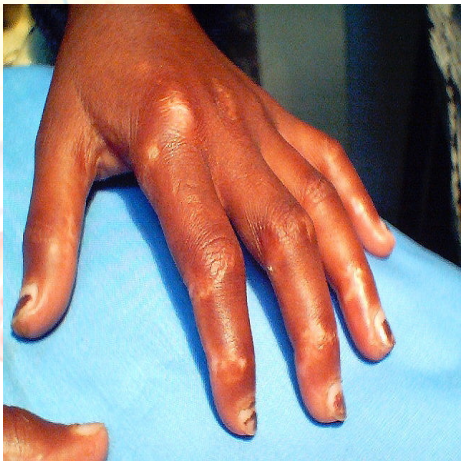
- ✓ **Definition :** An autoimmune multisystem disease that results in fibrosis and vascular abnormalities in association with autoimmune changes.
- ✓ Usually starts between 30-40 years in women who are more affected and later in men.
- ✓ Pathophysiology : may involve some injury to the endothelial cells and this results in excessive activation of the dermal connective tissue cells, the fibroblasts.
- ✓ Usually presents with Raynaud's phenomena,
- ✓ Thickening of the skin of the fingers, then atrophy and sclerosis.
- ✓ The fingers become spindle-shaped (sclerodactyly) from resorption of the fingertips.
- ✓ Fragile nails become smaller with ragged cuticles
- ✓ The tight shiny skin may affect most parts of the body, including the face, resulting in loss of expression and difficulty opening the mouth properly.
- ✓ Telangiectasia appear on the fingers, palms, face, lips, and chest.
- ✓ Ulcers may follow minor injuries over the joints, or on the tips of fingers and toes. Ulceration can lead to dry gangrene and eventual loss of the tips of the fingers
- ✓ Joint contractures. Patients will be bed ridden with time.
- ✓ Esophageal reflux and dysphagia.
- ✓ Lung and heart involvement may manifest as shortness of breath, high blood pressure, chest pain, pleurisy, pneumothorax, pericarditis arrhythmias, general heart enlargement and heart failure.
- ✓ Progressive kidney disease resulting in proteinuria, high blood pressure and eventually renal failure.



Small opening mouth, Finger No facial expertion , Small eye opening , Skin tight , shiny



Red spot (telangiectasia)



Thick shinny skin

- ✓ **Diagnosis:** made based on clinical features and presentation.
- ✓ Skin biopsy will show skin atrophy with preservation of skin appendages.
- ✓ ANA is usually positive.
- ✓ Anti topoisomerase I (Scl 70) is characteristic for it especially in severe cases.
- ✓ Need to take biopsy from kidney, heart and lung .

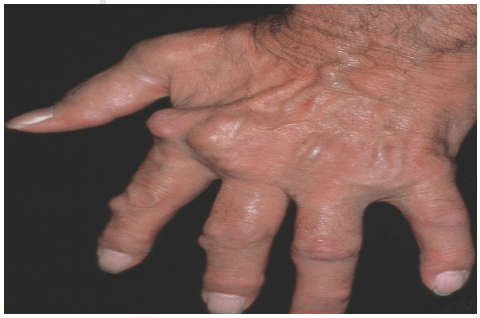
Treatment:

- ✓ Need Symptomatic treatment.
- ✓ Raynaud's phenomena:

Stop smoking, keep hands warm and decrease trauma. calcium channel blockers, aspirin and vasodilating drugs including nifedipine and iloprost infusions.

- ✓ Calcinosis cutis: nifedipine, surgical or laser excision.
- ✓ Skin sclerosis: physiotherapy, phototherapy.UVA
- ✓ GI: proton pump inhibitor, surgery for strictures.
- ✓ Kidney: ACE inhibitors.
- ✓ In severe cases: immunosuppressant , D-Penicillamine might be used.

Rheumatoid Nodules : (Usually a joint disease but Subcutaneous nodules the only skin manifestation)



- 20-30% of RA patients
- Subcutaneous nodules
- Found anywhere on the body
- Histologically shows dense foci of fibrinoid necrosis surrounded by histiocytes in palisaded arrangement.

Mixed Connective Tissue Disease:

- Mixed features of scleroderma, SLE, and dermatomyositis
- Raynaud phenomenon, sausage-shaped fingers, and swelling of the dorsa of the hands that never becomes sclerodactyly are the most typical features
- Alopecia, facial erythema, periungual telangiectasia, and pigmentary disturbances.
- Painful dermal nodules may appear on the hands or elbows.
- Cutaneous ulceration due to subcutaneous dystrophic calcification
- Anti-RNP (The auto antibody is U1-RNP) serologic marker

MCQ : (From Previous Exams of 430) :

A 50-year-old lady with dermatomyositis, you started her on systemic steroids. To follow up this Patient, which one of the following can be used:

a. ANA
b. CBC and biochemistry
c. Creatine kinase
d. Muscle biopsy

A 20 years old male patients presented with dark red atrophic plaque his skin biopsy showed thickening of the dermal collagens and loss of the skin appendages. What is the most likely diagnosis?

a. Morphea
b. Systemic sclerosis
c. Subacute lupus
d. Acute Lupus

A 40 year old woman presented with an eight months history of discoloration on the periorbital region and papules and plaques on the forearms and knuckles with thickened cuticles.
Which one of the following is the correct diagnosis :

a. Dermatomyositis
b. Discoid lupus erythematosus
c. Subacute lupus erythematosus
d. Systemic Sclerosis

Lady with scaly plaque with raised edges and hypopigmented depressed center
What is the most proper investigation to reach the diagnosis?

a-Anti smith.
b-Anti Ro.
c-Skin biopsy.
d-No need to investigate.

18-Patient came complaining of Muscle weakness with red edematous bilateral plaque over her eye lids.
Which one of the following signs she has?

a. Heliotrope.
b. Gottron papules.
c. Shawl sign.

References :

- **Doctor Al-Ghamidy Lecture**
- **Team 429**
- **Fitzpatrick's Color Atlas and Synopsis of Clinical Dermatology**
- **Old MCQ**

GOOD LUCK