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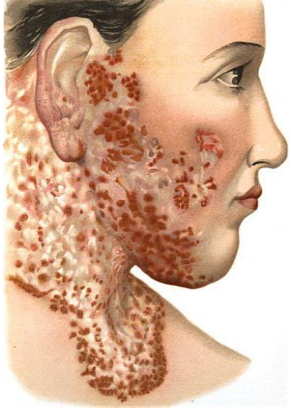
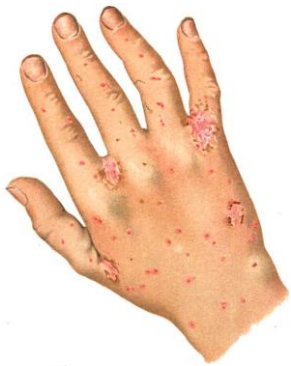
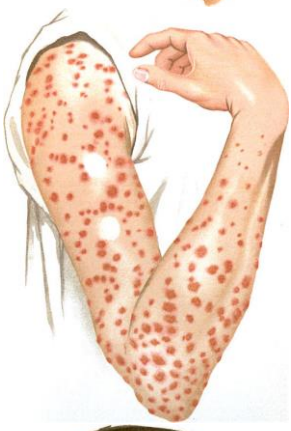
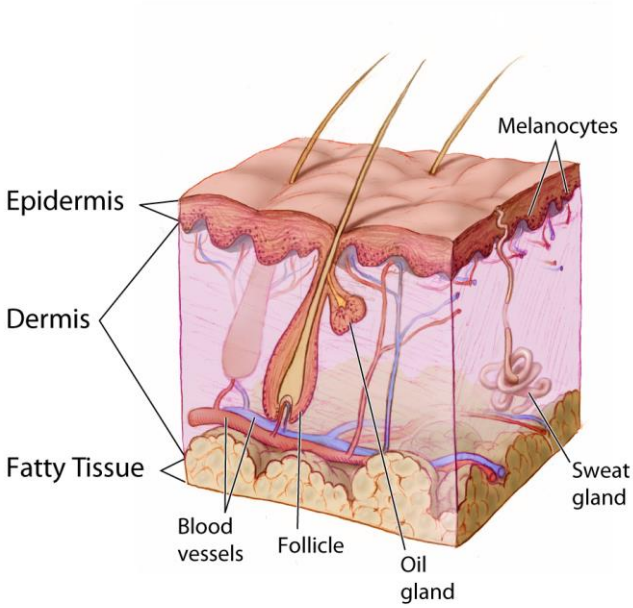






432 Teams

Dermatology



Connective Tissue Diseases

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

Done by: ***Arwa Almashaan***

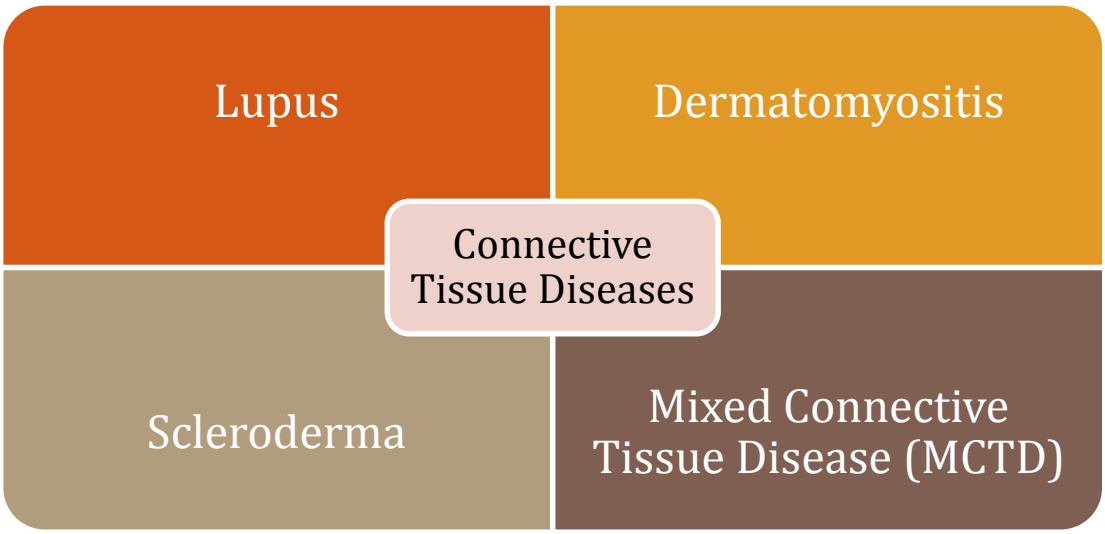
Revised by: Lama AlTawil

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

Objectives

At the conclusion of these lectures, the student will be able to:

1. differentiate between the various types of Lupus
2. recognize how Lupus affects the various systems of the body
3. identify all of the current treatment options available for Lupus
4. recognize the psychosocial effects that Lupus has on the patient and their family
5. To learn how to diagnose, investigate and manage dermatomyositis.
6. To learn the presentation of morphea and systemic sclerosis and ways to manage them.
7. To recognize other diseases like Rheumatoid nodules and mixed CTD.
8. This lecture is not meant to be inclusive of all the information about these diseases but to highlight important aspects in their diagnosis and management.



Lupus Erythematosus:

- LE is an autoimmune diseases associated with antibodies directed against components of cell nuclei.
- Is a multisystem disorder that may affect any tissue, kidneys, CNS, lungs and others. It might affects only skin without systemic involvement
- It is an immune complex disease that target collagen or ground substance. Type 3 hypersensitivity
  Some manifestations are not specific for lupus as livedo reticularis, and telangiectasia, and some are specific like butterfly rash and discoid lupus. Lupus Erythematosus can presented only as cutaneous manifestation and we call it **Cutaneous Lupus Erythematosus,** we follow them for years without presence of systemic involvement. If we talk about **Systemic lupus**, we need 4 out of 11 criteria to diagnose it.

Classification of Lupus Erythematosus (LE):

1. **Cutaneous**
2. **Systemic**

Classification of Cutaneous Lupus Erythematous

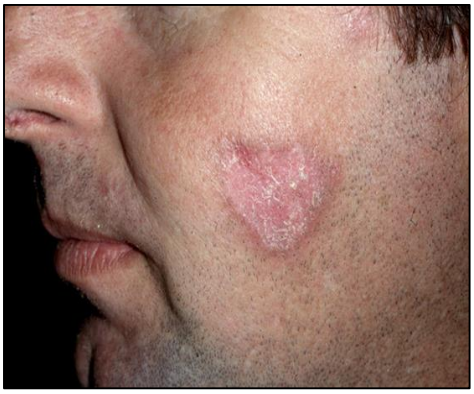
Acute	Subacute (SCLE)	Chronic Commonest

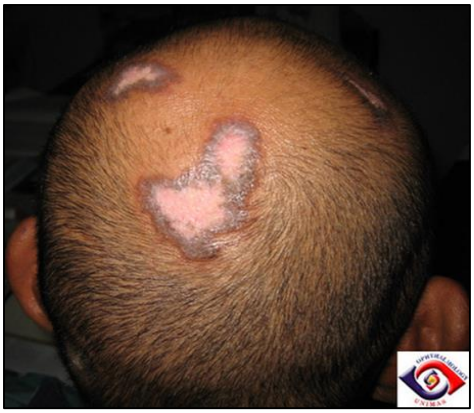
Specific: biopsy will show LE histopathology
• Malar eruption or 'butterfly rash'
• Erythematous papular rash on arms
• Photosensitivity
• Cheilitis and mouth ulcers
• Blisters (bullous LE)
Non-specific: as Hair fall Telogen Effluvium non-scaring "Telogen = last phase of hair fall", Raynaud's phenomena start as white pallor ischemia then blue stenosis then red active hyperemia, Periungual Telangiectasia , Vasculitis (small vessel)
	• Annular
	• Papulosquamous
	• Associated with syndromes :
	  •Neonatal LE
	  •Complement deficiency syndrome (C2 and C4)
	  •Drug induced SCLE (Subacute cutaneous LE)
		• Discoid LE commonest
		• Verrucous (hypertrophic) LE
		• Lupus erythematous – Lichen planus overlap
		• Chilblain LE
		• Tumid LE
		• Lupus panniculitis
		• All chronic manifestation end up with scaring not like in acute and subacute.

First, we will talk about the chronic cutaneous LE

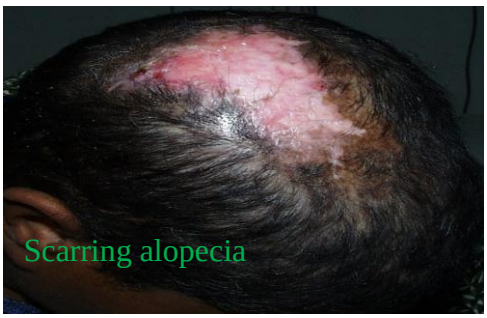
1-Discoid LE (DLE)

- Commonest form of cutaneous LE
- Common in young females
- Started as **red indurated plaques** and evolve with **atrophy**, **scarring** and **pigment** changes (mostly Hypopigmentation or depigmentation)
- **Hyperkeratosis** characterized by **follicular plugging** (scarring alopecia). Irreversible
- Common above **head and neck** nose, ear and ear canal and scalp. On the sun exposed areas.
- **Localized** form associated with **5%** with SLE.
- **Generalized** form associated with **20%** with SLE.
- May involve M.M Mucus membrane
- Aggressive SCC squamous cell carcinoma may arise from long standing DLE. 2% of patients so follow up is important.
- Generalized form associated more with SLE, +ve ssDNA, +ve ANA, leukopenia, high ESR.
- Treated by **superpotent topical steroid**, **antimalarial**, **retinoid** (oral isotretinoin and acitretin). For active lesions only. i.e erythematous verrucous not burnt out
- **Approach**: history for other systemic manifestation of lupus, lab and serology

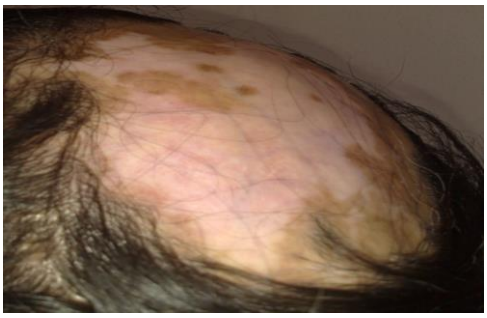


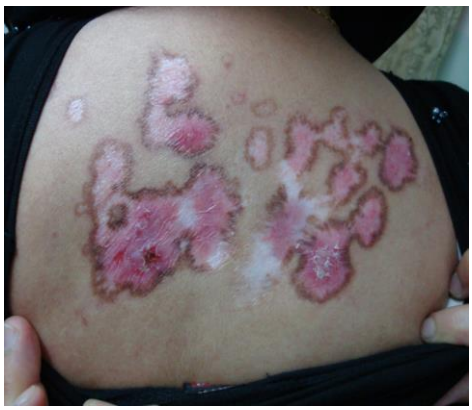


Lesion start to be depigmented in the center and no hair follicles opening end up as scarring alopecia



Scarring alopecia





Hypopigmentation in the central or inactive area and hyperpigmentation at the active border with Scaring and telangiectasia.

Pigmentation of the ear lobe with follicular plugging if you zoom in it looks like black comedones

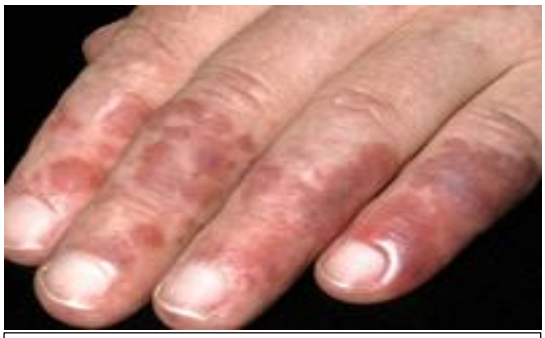
2- Hypertrophic LE (Verrucous):
Commonly seen on the extremities manly over **shins**. Hyperkeratosis, wart like, thick lesions. Not as common as discoid LE.

3- LE-LP (Lichen planus) overlap:
Common on the **extremities**, palm and soles associated with **oral ulcers** and **nail** involvement.

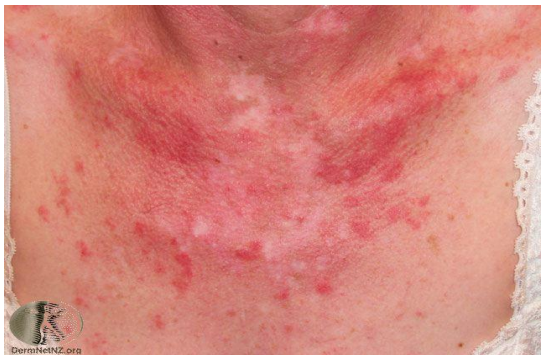
4- Chilblain LE (pernio):
Presents as tender red or purple skin lesion that involve fingertips, rim of the ears, calves and heals, **exacerbated by** cold exposure. DDX frost bite

5- Tumid LE:
Presented with edematous erythematous plaques on the trunk and respond to antimalarial.

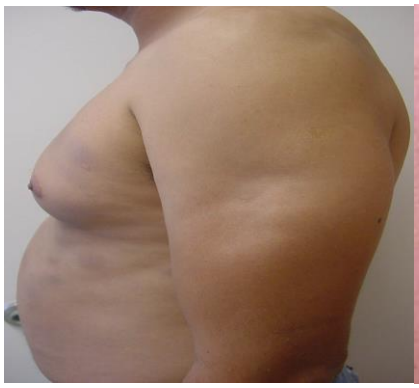
6- LE panniculitis:
Presented as non-tender subcutaneous nodules on **face** or **proximal** extremities that heals with atrophy. Lupus profundus is another name for LE panniculitis **if** on top of it discoid LE.

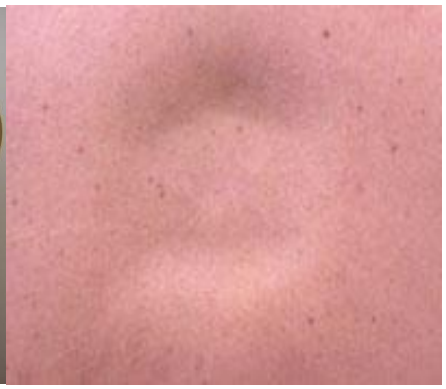


Chilblain LE
Erythematous violaceous **macules** over the **fingers**.



Tumid LE
Erythematous, edematous **plaques**, urticaria-like or eczematous wheel , on the **chest** exacerbated by **heat** Hx"



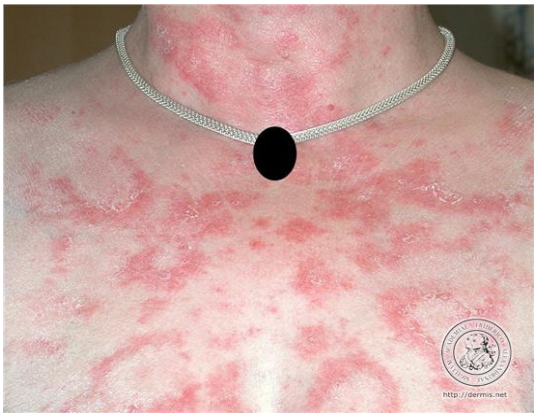


LE panniculitis

- Burnt out lesion end result is deep scars on fat layer or lipoatrophy
- Common in **face** which tend to be disfiguring.
- **Treatment** by intralesional steroid or antimalarial (has anti-inflammatory effect on lupus and maybe used if monotherapy is not affected). They are used in the active phase
- If atrophy happened, it is difficult to inject fillers because of the atrophy fibrosis in subcutaneous area.
- 30% association with SLE. So, do Hx, lab, …etc. and to start aggressive treatment

Then, we will talk about the subacute cutaneous LE.

Subacute cutaneous lupus erythematosus:

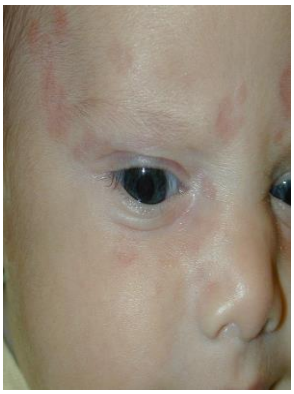


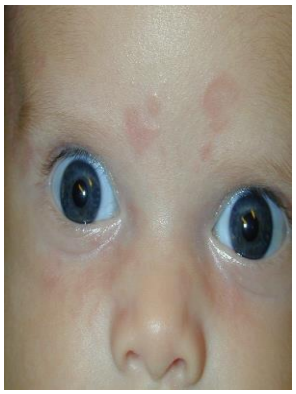
- 10 to 15% of LE.
- Common on young females.
- A non-itchy rash appears on the upper back and chest (on sun-exposed regions).
- Associated with photosensitivity and arthralgia.
- Systemic involvement is **not** usually severe. But 1/3 will get SLE
- Can be **annular** or **psoriasiform** "papulosquamous = papule + scale" and heals without scarring.
- Team 431:" Papulosquamous, differentiate it from psoriasis by location. Psoriasis usually appears over joints "knees, elbows, sacral area" but SCLE is usually on the chest and upper back"
- **Labs**: 80% ANA and 70% **anti-RO**, anti-LA. (the underlined are mainly for sub-acute)
- **Treated** by **antimalarial** and sun protection.
- Must follow up once a year to make sure they don't convert to SLE

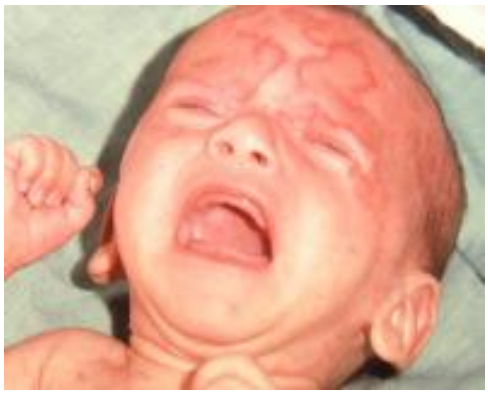
Complement deficiency sub-acute cutaneous lupus →deficiency in C2 and C4

Neonatal lupus erythematosus

- Newborn babies born to mothers with subacute LE even if she has no clinical symptoms but she have +ve anti-RO, anti-LA
- May develop annular rash, known as neonatal LE that resolve spontaneously without scaring. Because the antibodies will clear after they are born
- The neonates could be at risk of complete heart block may end up with pacemaker, thrombocytopenia, and increased liver enzymes.
- Therefore, you should take Hx, and PEx of **first** the mother to see if she has the disease clinically, and for the baby order CBC, liver enzymes, serology, and refer to cardiology "ECG and echocardiogram".







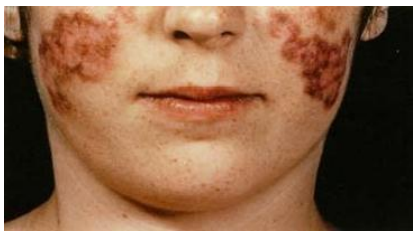
Neonatal LE
Same as subacute LE. It could be annular or papulosquamous. No need for skin biopsy

Systemic Lupus Erythematosus:

- Only a few patients with cutaneous LE also have SLE. The **most common presentation is with a malar eruption or butterfly**. Other skin changes in SLE are **photosensitivity**, **mouth ulcers**, and **diffuse hair loss**.
- May also affect joints, kidneys, lungs, heart, liver, brain, blood vessels and blood cells
- ACR criteria for the diagnosis: 4 out of 11 need for the diagnosis.

1. **Malar rash.**
2. **DLE.**
3. **Photosensitivity**.
4. **Oral ulcers**.
5. Arthritis **small joint** of the hand.
6. Serositis (pleuritis , pericarditis, peritonitis)
7. Renal : proteinuria (protein more than 0.5 g/day)
8. CNS: Seizure or psychosis
9. Blood: anemia of chronic disease, hemolytic anemia, lymphopenia, leukopenia, thrombocytopenia
10. +VE ANA
11. +VE anti-ds DNA and anti-smith and anti-phospholipid antibodies. If one of them +ve account as criteria

- First Four are cutaneous criteria "non-painful".
- If you have SLE patient with anemia you have to do coombs test **"Autoimmune hemolytic anemia"**
- Repeat this for the patient with cutaneous LE every 1 year. You must screen for systemic involvement
- Anticoagulant lupus seen in anti- phospholipids syndrome so look for their antibodies
- Almost all patients with acute cutaneous will get SLE



Drug induced Lupus:

- Associated with **ANA** and **anti-histone** antibodies.
- Does **not** usually affect the skin.
- Hydralazine, procainamide, sulfonamide, anticonvulsants, Minocycline = a tetracycline for acne but it a has side effect that it induce lupus Erythematosus.

Lab Findings for SLE:

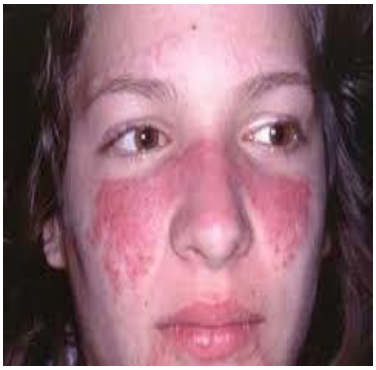
- CBC: anemia , Leucopenia , thrombocytopenia
- High ESR
- Coombs test: hemolytic anemia
- Urinalysis and 24 hr urine collection : for proteinuria
- U &E.
- Serology: ANA (+ve in 95% of SLE patients, **screening** test), anti-ds DNA (specific), anti-ss DNA in generalize DLE, anti-smith (specific), anti-RO, anti-LA, lupus anticoagulant and anti cardiolipin (in anti-phospholipid), anti-n RNP(mixed connective tissue disease). For screening ANA and we do with it as baseline anti-ds DNA
- Low C2 and C4. Subacute CLE associated with Complement deficiency syndrome.

- **Skin biopsy** may be diagnostic especially in DLE (you will see lymphocyte infiltration (chronic), atrophic epidermis, follicular hyperkeratosis, inflammation and deposition of immune complexes in the basement membrane and thickening). **Direct IF** may show positive antibody deposition along the basement membrane due to chronic inflammation **(lupus band test)**. The older the lesion the higher chance of finding antibody deposition.

Table 2. Clinical Features of SLE

System	Presentation
Constitutional	Fatigue, fever (in absence of infection), weight loss
Musculoskeletal	Arthritis, arthralgia, myositis
Skin	Butterfly rash, photosensitivitiy, mucous membrane lesion, alopecia, purpura, urticaria, vasculitis
Vascular	Raynaud's disease
Renal	Hematuria, proteinuria, renal casts, nephrotic syndrome
Gastrointestinal	Nausea, vomiting, abdominal pain
Pulmonary	Pleurisy, pulmonary hypertension
Cardiac	Pericarditis, endocarditis, myocarditis
Reticuloendothelial	Lymphadenopathy, splenomegaly, hepatomegaly
Hematologic	Anemia, thrombocytopenia, leukopenia
Neuropsychiatric	Psychosis, seizures, organic brain syndrome, transverse myelitis, cranial neuropathies, peripheral neuropathies

SLE: systemic lupus erythematosus.
Source: Reference 13.



Malar rash

Treatment of cutaneous lupus

- The aim of treatment for cutaneous LE is to alleviate symptoms and to prevent scarring. The treatment in general depends on the stage.

Local therapy:

- Sun protection. By the use of physical and chemical sunscreen against UVA, UVB light. Also, shade the car glass, wear sun glasses, and if working outdoor change to indoor
- Topical or intralesional steroid
- Topical calcineurin inhibitor e.g. tacrolimus

Systemic antimalarial therapy:

- **Hydroxychloroquine** 200 mg OD or BD in adults up to 6.5mg/kg.
- **Chloroquine** (125-250 OD in adults up to 3.5-4 mg/kg.
- **Quinacrine** 100 mg OD.

Combination: we wait for 3 months if no response we combine two antimalarial drugs .

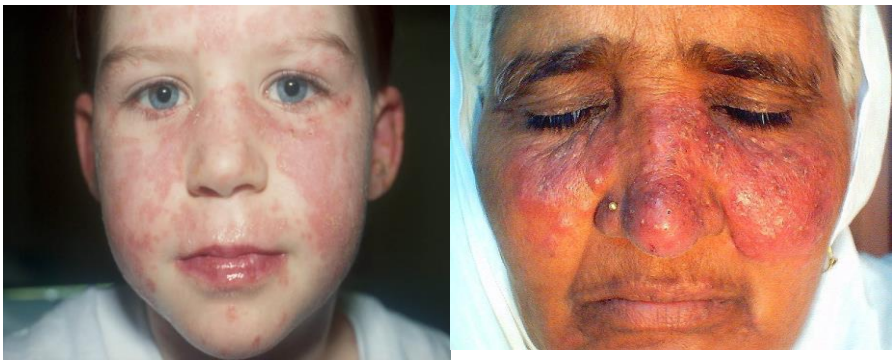
Side effects of Hydroxychloroquine: cause pigment deposition

- Eye toxicity. In lens its **reversible** but in **retina** irreversible. Baseline ophthalmology screening.
- Skin eruption. Mainly pigmentation. With Chloroquine yellow discoloration of the skin. Other ADR: Drug induced psoriasis.
- Leukopenia and thrombocytopenia
- Hemolytic anemia in G6PD def.
- Liver toxicity

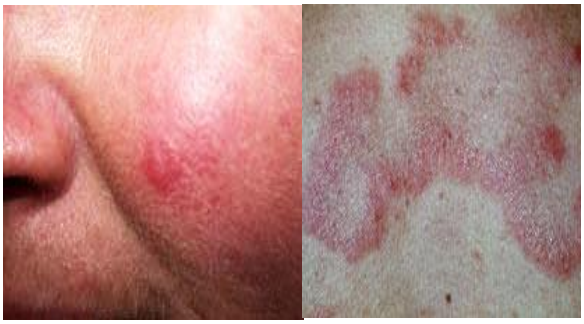
Therefore, you should do CBC, liver function test, G6PD, and refer to ophthalmologist.

Other Systemic therapy

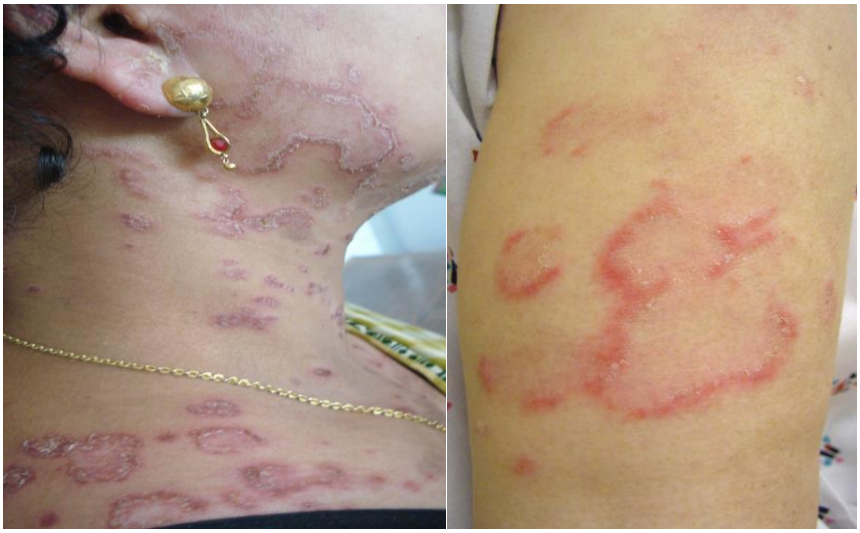
- Retinoid. Used also for DLE
- Thalidomide. Used in past for hyperemesis in pregnant women. SE: Phocomelia "short limbs of the fetus", neuropathy "take patient consent before using it". Not preferable.
- Dapsone for bullous lupus
- Systemic steroids in SLE you should go aggressive in systemic involvement , otherwise antimalarial is good
- Steroid sparing agents: Azathioprine, Methotrexate, Mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG, and Rituximab "Biological treatment". As add ons



SLE



Lupus Tumidus



Subacute Lupus Erythematosus

Dermatomyositis

- An uncommon inflammatory disease affects adults between 40-60 (females mainly) and children 5-15.
- Skin changes.
- Muscle weakness

Classification:

- Classic DM :
  -Adult type
  -Juvenile type
- Amyopathic DM. Dermatomyositis-sine myositis, also known as ADM, is diagnosed in patients with typical cutaneous disease in whom there is no evidence of muscle weakness and who repeatedly have normal serum muscle enzyme levels
- Paraneoplastic DM
- Overlap with CTD
- Drug induced DM : D-penicillamine, hydroxyurea, NSAID "diclofenac", STATIN and Phenytoin

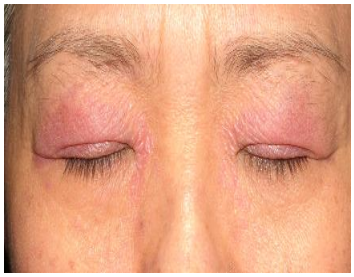
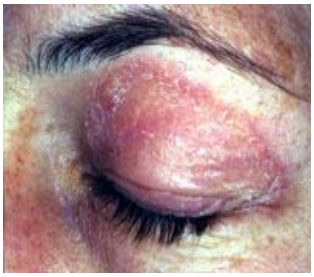
Clinical features

Skin:

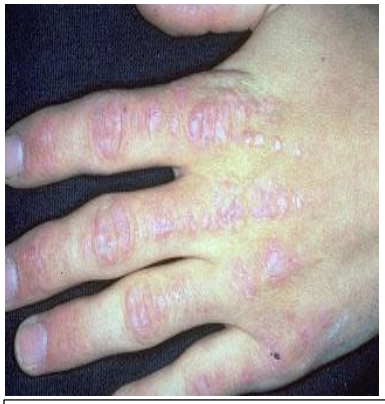
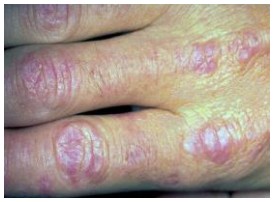
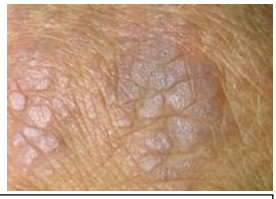
- **Heliotrope rash:** scaly edematous pinkish plaques over both eyelids.
- **Gottron's sign:** scaly erythematous plaques over elbows and knees like psoriasis.
- **Gottron's papules**: pink to purple flat topped papules over knuckles
- Erythema of **upper chest** (**V**) pattern or over **shoulders** (**shawl sign**)
- Periungual telangiectasia.
- **Mechanics Hand**: hyperkeratosis, fissuring, scaling with hyperpigmentation over finger tips, sides of fingers and palms. You will have +ve Anti- Jo-1 and lung disease
- **Holster sign:** scaling and erythema over hips and thigh.
- **Poikiloderma:** atrophy, Mottled hyper and hypopigmentaion, and telangiectasia

Other cutaneous signs and symptoms include: not specific

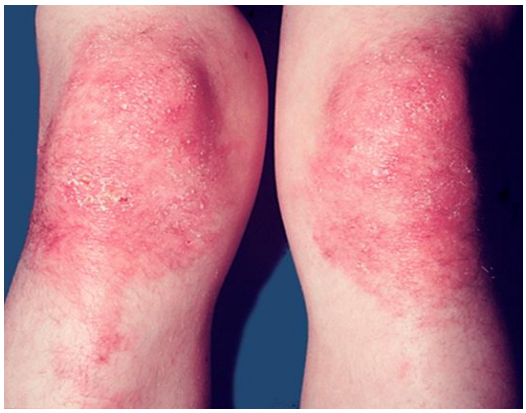
- Ulceration
- Photosensitivity
- Raynaud's phenomenon
- Calcinosis cutis **especially in children**

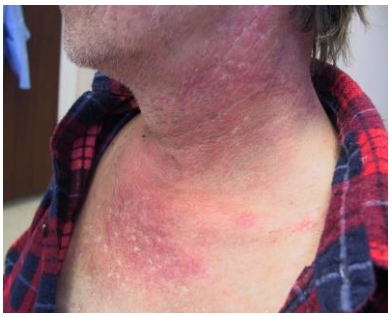
Heliotrope rash
violaceous erythema over the **eyelid** with edema. Indicates an inflammation of the underlying muscle, which is orbicularis oculi.

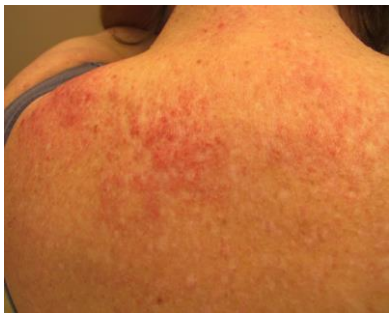
Gottron's papules
Flat toped violaceous nodule over the **knuckles** of the hands**. Look like lichen planes description but over the knuckles**



Gottron's Sign
Erythematous scaly plaque over the knees. Unlike the silver thick ones seen in psoriasis



V-shape erythema of neck



Shawl sign

Muscle:

- Weakness, progressive **proximal** muscle weakness involves the hips, thighs, shoulders, upper arms and neck.
- The weakness is symmetrical and more in the extensor muscles. Difficulties in rising hands above head, and getting up from a chair. Esophageal muscles especially the **upper 1/3 striated** muscles affected with dermatomyositis and the patient presents with dysphagia.

Lungs:

- Interstitial lung disease. If patient came with Mechanics hand, he have high chance to develop interstitial lung disease.

Periungual telangiectasia and cuticle hypertrophy
DLE has a similar picture but its alternating with pallor

Associated disease

It can be associated with:

- Other CTD: lupus, rheumatoid arthritis, scleroderma and Sjogren's syndrome.
- Cancer: take Hx of weight loss, and systemic review. Screen them for age-related CA.
  - Especially in older patients. Age related CA
  - **common** : ovarian CA (the most common), if male GI "Colon"
  - Cancer of the cervix, lungs, pancreas, breasts, lymphoma and gastrointestinal tract.
  - Malignancy could precede, coincide or follow the diagnosis of DM.

Investigations and Diagnosis:

- In general, you will start with Hx, PEx, then ask for CBC, LFT, and U&E!!

CRITERIA:

1. Typical **skin rash**.
2. Muscle **weakness** (proximal upper or lower extremity)
3. +ve **Muscle enzymes** (Creatine Kinase and aldolase)
4. +ve **EMG** (short duration , polyphasic)
5. +ve **muscle biopsy** → best location for the biopsy is **triceps**

- Definite: rash with three muscle findings.

OTHER LABS:

- Serology:
  - ✓ Anti- Jo-1 (histidyl- tRNA synthetase): **lung disease & mechanical hands**.
    Anti- Jo- 1 in general is positive in all dermatomyositis
  - ✓ Mi-2: specific for DM **with skin findings**
- Skin Bx Biopsy
- Magnetic resonance imaging (MRI). Of the muscles

With all of the above, you should screen for malignancy.

Treatment

SKIN:

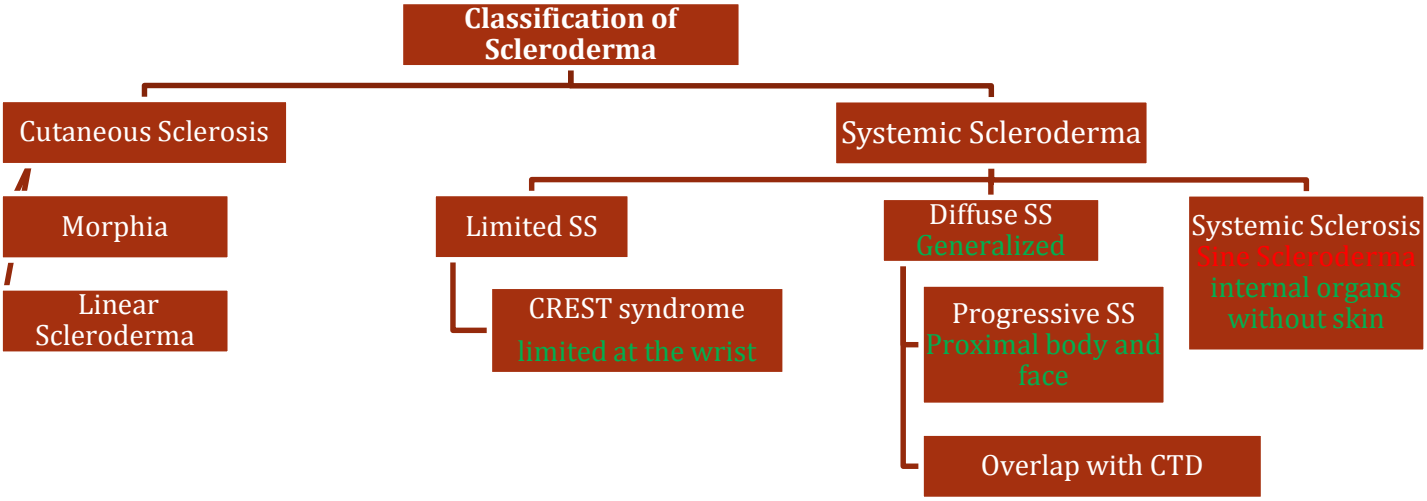
- Sunscreen
- Topical steroid, Topical Tacrolimus
- Hydoxychloroquine antimalaria
- Oral **steroids** are the mainstay treatment. If severe

Systemic therapy:

- Steroid sparing agents are Methotrexate, azathioprine, mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG, and Rituximab.
- **Physiotherapy** to improve strength and flexibility of the muscles.
- **Surgical excision or Co2 laser** could be utilized to remove tender calcium deposits. In Calcinosis cutis. Medical Tx as aluminum hydroxide, Calcium channel blockers.
- Used in systemic involvement of lung and muscle start with steroid and steroid sparing agents

Systemic Sclerosis (Scleroderma)

- A group of diseases that involve the **hardening** and tightening of the skin and connective tissues
- Affects women more often than men.
- Most commonly occurs between the ages of 30 and 50.

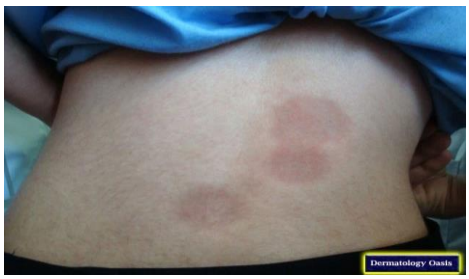


Localized Cutaneous Sclerosis:

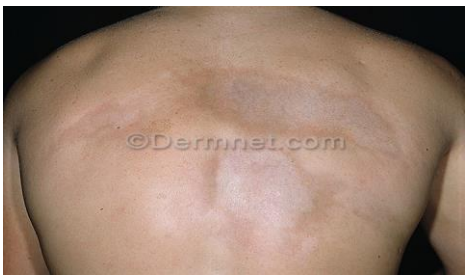
- Pure cutanouse disease (not involving other systems)

Morphia:

- More common in Female
- oval reddish or purplish patches and plaques on the skin that progress to smooth , hard, sclerotic, depressed plaque surrounded by violashous zone
- It subsides **on its own** over time leaving dyspigmentation and atrophy and sclerosis
- Can be localized or generalized **without** visceral involvement

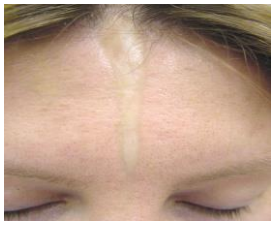
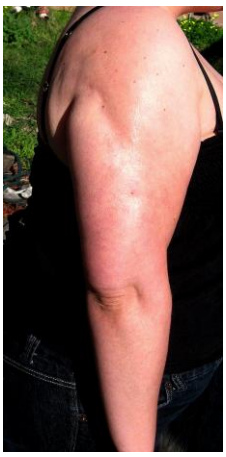


Morphia Violashous or erythematous atrophic plaque this is an active lesion



Morphia Sclerotic shiny skin. This after the active lesion heals. Burnt out notice the brown zone

Linear Scleroderma or Linear Morphia:

- Parasagittaly on the **forehead** (encoup de sabre). Common in forehead and over joints. Can occur over the limbs
- **Parry-Romberg syndrome** manifested as progressive hemifacial atrophy, epilepsy, exopthalmos, and alopecia. + encoup de sabre
- May extend on the extremities associated with muscle atrophy and flexion contracture.
- The woman with the scarf in the picture has a burnt out lesion

Diagnosis

Diagnosed on the base of its morphology and confirmed by skin biopsy, which usually show **thinking of collagen bundles** and **loss of skin appendages** like sweat glands and hair follicles. So homogeneous. The biopsy is rectangular shape here unlike the usual cone type.

Treatment:

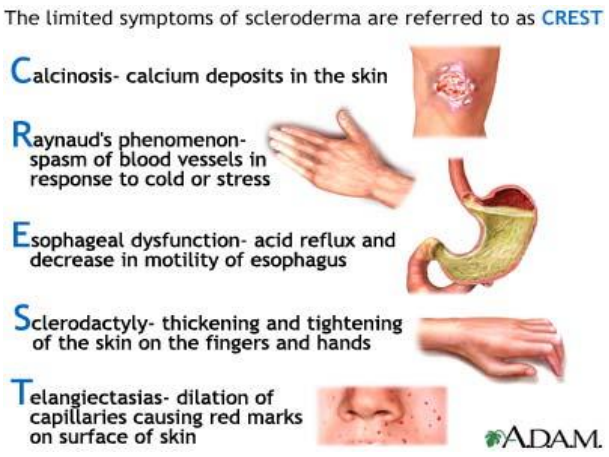
Treatment of morphea focuses on **controlling signs and slowing spread**. Scaring is irreversible

- **Topical steroid.**
- **Topical vitamin D analogue**. Calcipotrine (dovonex) the Keratinocyte has receptors for Vit D, which help in its proliferation and differentiation. usually used with topical steroid in the active stage
- Phototherapy. Mainly UVA for deeper penetration.
- Systemic steroid, azathioprine, methotrexate, and cyclosporine might be used in severe cases. If you have generalized Morphia or linear Morphia over the head and joints use systemic. Because you worry of the contractures + you can give photo therapy
- **Physical therapy** could be of help if the involvement is close to **joints** and cause contracture and difficulty movement.

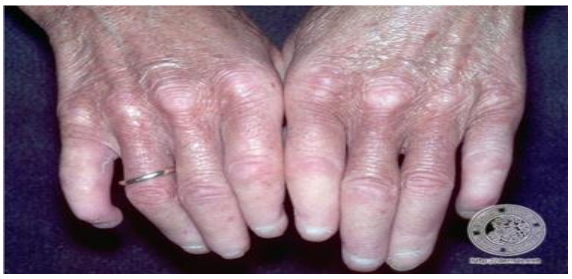
Limited Systemic Sclerosis

CREST syndrome:

- **C** Calcinosis Cutis
- **R** Raynauds Phenomena
- **E** EOSOPHAGEAL DISMOTILITY Lower 2/3 of theb esophagus with SS but upper 1/3 with Dermatomyositis
- **S** SCLERODACTALY skin sclerosing with flexion contracture.
- **T** TELANGECTASIA

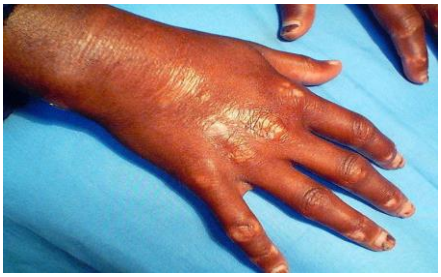


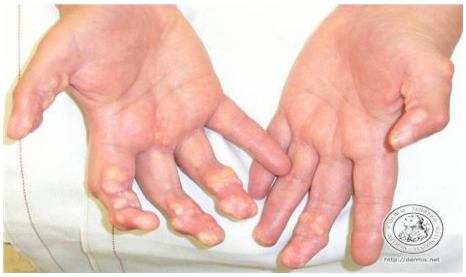
- Good prognosis. No lung and organ involvement.
- Anticentromere antibodies (specific)



Sclerodactaly

Distal sclerotic tight skin with flexion contracture. Unlike progressive which starts proximally





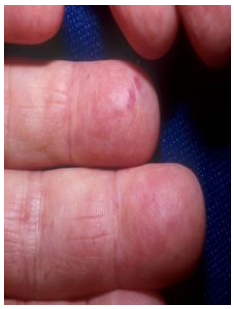
Calcinosis Cutis

Calcium deposition. Treatment: CCB, aluminum hydroxide, and surgical. With hypertension, pericarditis and retinopathy and poly arthritis and renal involvement

Systemic Sclerosis (Diffuse SS):

- An autoimmune multisystem disease that results in fibrosis and vascular abnormalities in association with autoimmune changes.
- Usually starts between 30-40 years. More in women
- Pathophysiology may involve some injury to the endothelial cells and this result 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

Skin findings

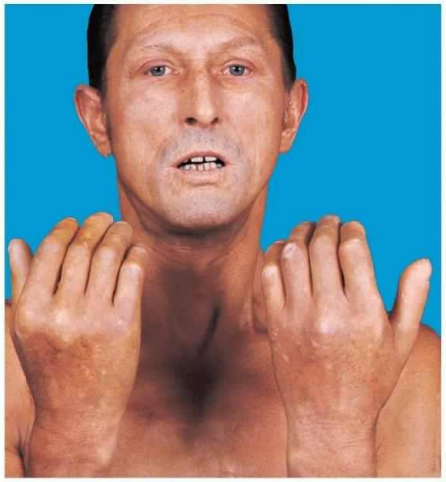
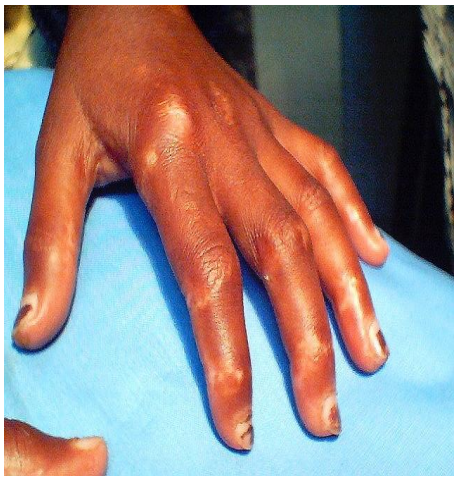


- Raynaud phenomena.
- **Face**: expressionless, constricted mouth, peaked nose. Thin lips with radially furrowed.
- Claw like hands
- **Neck sign**: ridging and tightening of neck on extension.
- Fingertip atrophy, ulceration and gangrene.
- Nail fold capillary hemorrhage
- Telangiectasia appear on the fingers, palms, face, lips, and chest
- Hyper or depigmented spots (salt and pepper skin)

Internal involvement

- Esophageal dysmotility(distal) Esophageal reflux and dysphagia:
- Pulmonary fibrosis
- Pericarditis , HTN
- Retinopathy
- Polyarthritis

- Progressive kidney disease resulting in proteinuria, high blood pressure and eventually renal failure

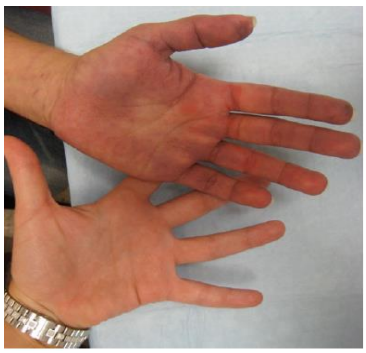



Investigations and Diagnosis:

- Diagnosis made based on clinical features and presentation.
- **ANA** is usually positive. **Anti-topoisomerase I** (Scl 70) is characteristic for it especially in severe cases.
- High **ESR** , thrombocytopenia, hemolytic anemia
- High **CK** with muscle involvement
- **CXR** : diffuse **ground glass and honeycomb lung pattern**
- **CT** :lung fibrosis
- ECG : arrhythmia
- Skin biopsy will show skin atrophy with thickened collagen bundles with absent skin appendages.
- If only morphea no need for screen them for systemic, but if CREST syndrome or systemic , do X-ray and CT to the lung.

Treatment of SS

- Is symptomatic.
- **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.
- **GI**: proton pump inhibitor, surgery for strictures.
- **Kidney:** ACE inhibitors.
- **In severe cases**: immunosuppressant , D-Penicillamine might be used
- Mainstay is to treat the lung and reflux



Male Team 432

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

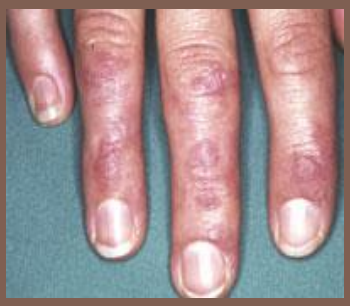
Very Important

Table 2: Useful Antibodies for CTD-ILD Assessment

Autoantibody	Associated CTD
High titer ANA (>1:320 titer)	Many
High titer RF (>60 IU/mL)	RA, Sjögren's disease, SLE
Anti-CCP	RA
Anti-centromere	Systemic sclerosis
Anti-nucleolar-ANA	Systemic sclerosis
Anti-Ro (SS-A)	Many
Anti-La (SS-B)	SLE, Sjögren's disease
Anti-Smith	SLE
Anti-ribonucleoprotein	SLE, MCTD
Anti-dsDNA	SLE
Anti-topoisomerase (Scl-70)	Systemic sclerosis
Anti-tRNA synthetase antibodies	Poly-/dermatomyositis (antisynthetase syndrome)
Anti-PM-Scl	Systemic slerosis/myositis overlap
Anti-Th/To	Systemic sclerosis
Anti-U3 ribonucleoprotein	Systemic sclerosis
ANCA panel	Systemic vasculitis

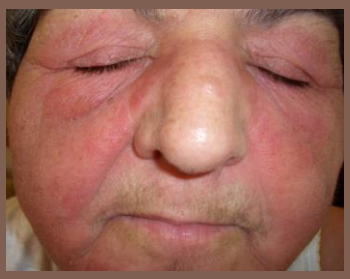
From the males:

Dermatomyositis

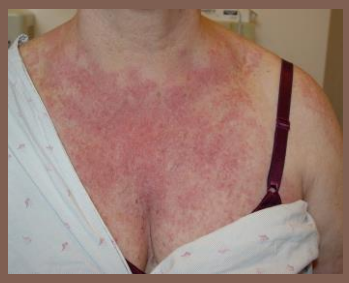


Gottron papules

Raynaud's phenomenon

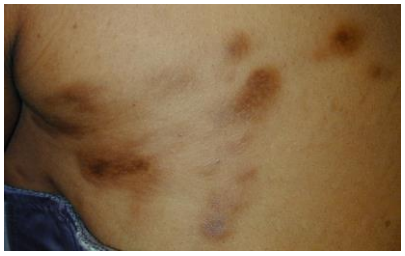


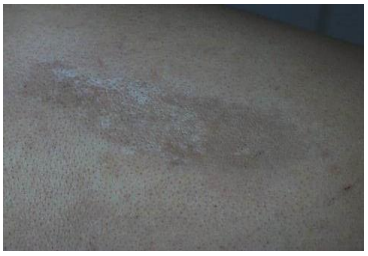
Puffiens of cheek & eyelid

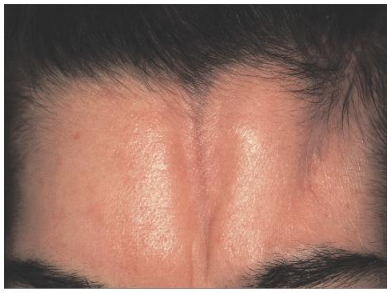


shawl sign rash

Scleroderma linear type:







Summary (From FitzPatrick's book)

Lupus Erythematosus (LE)
ICD-9: 695.4 ◦ ICD-10: L93

- LE is the designation of a spectrum of disease patterns that are linked by distinct clinical findings and distinct patterns of cellular and humoral autoimmunity.
- LE occurs more commonly in women (male to female ratio 1:9).
- LE ranges from life-threatening manifestations of acute systemic LE (SLE) to the limited and exclusive skin involvement in chronic cutaneous LE (CCLE) (Fig. 14-32). More than 85% of patients with LE have skin lesions, which can be classified into LE specific and nonspecific.
- An abbreviated version of Gilliam classification of LE-specific skin lesions is given in Table 14-3.
- Acute cutaneous LE (ACLE) is practically always associated with SLE, subacute cutaneous LE (SCLE) in about 50%, and CCLE most often has only skin disease. However, CCLE lesions can occur in SLE.
- ACLE and SCLE are highly photosensitive.

Dermatomyositis ICD-9: 710.3 ◦ ICD-10: M33.0

- Dermatomyositis (DM) is a systemic disease belonging to the idiopathic inflammatory myopathies, a heterogeneous group of genetically determined autoimmune diseases targeting the skin and/or skeletal muscles.
- DM is characterized by violaceous (heliotrope) inflammatory changes +/– edema of the eyelids and periorbital area; erythema of the face, neck, and upper trunk; and flat-topped violaceous papules over the knuckles.
- It is associated with polymyositis, interstitial pneumonitis, and myocardial involvement.
- There is also a DM without myopathy (amyopathic DM) and polymyositis without skin involvement.
- Juvenile DM runs a different course and is associated with vasculitis and calcinosis.
- Adult-onset DM may be associated with internal malignancy.
- Prognosis is guarded.

Scleroderma ICD-9: 710.1 ◦ ICD-10: M34

- Scleroderma is a not so rare multisystem disorder characterized by inflammatory, vascular, and sclerotic changes of the skin and various internal organs, especially the lungs, heart, and GI tract.
- Limited systemic scleroderma (lSSc) (60%) and diffuse systemic scleroderma (dSSc) are recognized.
- Clinical features always present are skin sclerosis and Raynaud phenomenon.
- Considerable morbidity; high mortality of dSSc.
- *Synonyms:* Progressive systemic sclerosis, systemic sclerosis, systemic scleroderma.

QUESTIONS

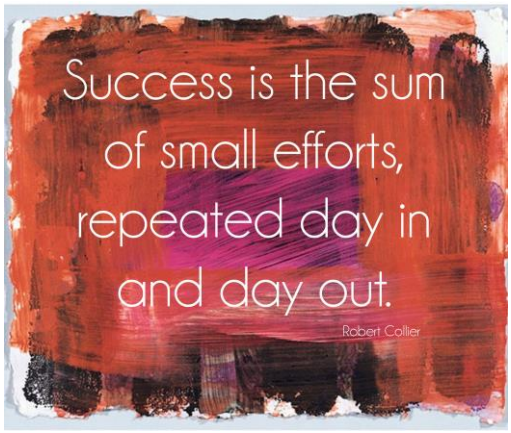
1- A 50-year-old lady with dermatomyositis, you started her on systemic steroids.
To follow up this Patient, which one of the following can be used:
a. ANA
b. CBC and biochemistry
c. Creatine kinase
d. Muscle biopsy

2- 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

3- 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

4- 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.

5- 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.



1- C
2- A
3- A
4- C
5- A