Derma Team 436

Cutaneous manifestations of SLE and other CTD (dermatomyositis, scleroderma)

Objectives:

- Differentiate between the various types of Lupus
- Recognize how Lupus affects the various systems of the body
- Identify all of the current treatment options available for Lupus
- To learn how to diagnose and investigate dermatomyositis.
- How to manage dermatomyositis.
- To learn the presentation of morphea and systemic sclerosis and ways to manage them.
- 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.

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Members: Reem Alsergani
Revised by: Shrooq Alsomali
Template designed by Group B

Before you start.. CHECK THE EDITING FILE

Sources: doctor's slides and notes

[Color index: Important | doctor notes | Extra | slides + slzides]

A) LUPUS ERYTHEMATOSUS

- It's a designation of a spectrum of diseases that are linked by distinct clinical findings and distinct patterns of polycolonal B cell immunity. (antigen-antibody)
- It ranges from life threatening manifestations of SLE to the limited and exclusive skin involvement in CCLE. Like discoid lupus erythematosus. It can range from mild nonselective like purpura (differentials of purpura: infection (hep B or C), Drugs, autoimmune (that's why with any purpura we have to check ANA to rule out autoimmune))
- More than 85% of patients with LE have skin lesions, which can be classified into LE-specific & non-specific*.

Revised ACR's Criteria for Classification of SLE

Any 4 of the following criteria are required to make the diagnosis:

- Malar rash
- Discoid rash
- Photosensitivity
- Oral ulcer
- Arthritis
- Serositis
- Renal dis.
- Neurological dis.
- Hematological inv.
- Immunological dis.
- ANA

Classification of Cutaneous Disease in Lupus Erythematosus

Lupus Specific eruptions	Lupus- nonspecific eruptions
1 . Acute cutaneous LE (ACLE) Mallor rash> nonscarring Localized, generalized (photodistributed)	• Nonscarring alopecia • Telangiectasia • Livedo Reticularis (autoimmune) • Palpable Purpura • Periungual erythema (in dermatomyositis and systemic sclerosis) • Urticarial vasculitis urticaria : evanescent wheals that disappear within less than 24 hrs, if it persists more than 24 hrs > this is called urticarial vasculitis and we have to look for the cause. • Raynaud's syndrome mostly in Systemic sclerosis. Vasospasm so it turns white then blue due to cyanosis then red due to hyperemia and dilation) • Photosensitivity we have to check ANA and anti Rho to rule out autoimmune
2. Subacute cutaneous LE (SCLE) usually nonscarring but could be mixed A. Annular B. Papulosqamous C. Syndromes commonly exhibiting similar morphology 1. Neonatal LE (NLE) 2. Complement deficiency syndromes 3. Drug induced (drug induces SCLE is different drug induced lupes)	
3. Chronic Cutaneous LE A. Discoid LE (DLE) 1. Localized, head and neck face and scalp: Differentials:1. Lichen planus 2.DLE so BIOPSY is important. Also, don't forget to check the ears for brown scaly plaques and follicular plugging 2. Disseminated (higher chance to have SLE) 3. Verrucous (hypertrophic) DLE B. Tumid Lupus (non scarring no epidermal changes) C. Lupus panniculitis (it will scar because its an inflammation of subcutaneous fat so requires a DEEP biopsy and aggressive treatment. Its also related to morphea (discussed later)) D. Chilblain LE (cold areas, fixed lesion)	
4. Other variance Bullous SLE acute Rowell's syndrome (SLE clinically looks like erythema multiform) looks like Steven- johnson syndrome. Which involves the SKIN + MUCOS MEMBRANE. Causes of Steven Johnsen syndrome: 1.mycoplasma pneumonia 2.Drugs 3. SLE (but in lupus it will only affect the skin sparing the lips)	

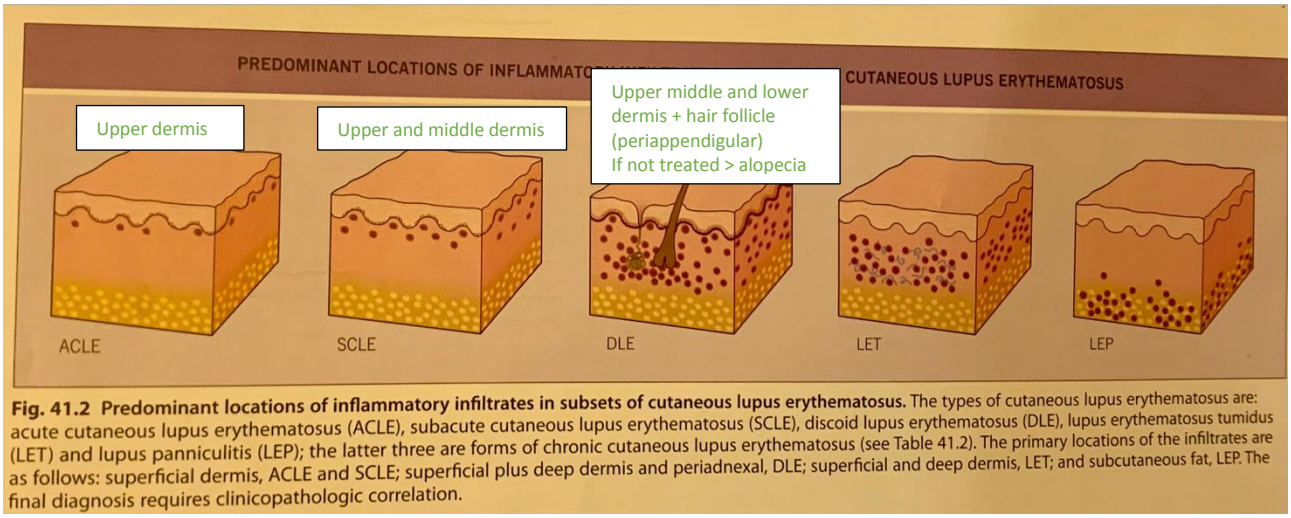


Fig. 41.2 Predominant locations of inflammatory infiltrates in subsets of cutaneous lupus erythematosus. The types of cutaneous lupus erythematosus are: acute cutaneous lupus erythematosus (ACLE), subacute cutaneous lupus erythematosus (SCLE), discoid lupus erythematosus (DLE), lupus erythematosus tumidus (LET) and lupus panniculitis (LEP); the latter three are forms of chronic cutaneous lupus erythematosus (see Table 41.2). The primary locations of the infiltrates are as follows: superficial dermis, ACLE and SCLE; superficial plus deep dermis and periadnexal, DLE; superficial and deep dermis, LET; and subcutaneous fat, LEP. The final diagnosis requires clinicopathologic correlation.

1) Acute Cutaneous Lupus Erythematosus

- Acute malar " butterfly rash" or more generalized photodistributed eruption.
- Nearly ALL patients presenting with ACLE will have systemic lupus erythematosus (SLE), often in an acute flare.
- Patients with ACLE will nearly always have a +ve ANA
- ACLE is transient (superficial dermis), improves with improvement of the SLE
- Non scarring

Are there any common skin eruptions that may be confused with acute coetaneous lupus erythmeatosus?

ACLE > no need biopsy because I know for sure the patient has SLE
But for Discoid> I need a biopsy

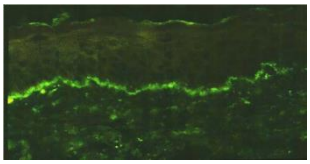
- Seborrheic dermatitis erythema and scales (nasolabial folds, eyebrows and chest)
- Vascular type of rosacea chronic and triggered by sun exposure and heat like ovens
- Polymorphous light eruption
- Photo reaction to systemic medications (doxycycline and psoralen) and topical products
- Certain types of porphyria(related to heme synthesis)

What is the initial workup of ACLE?

1. History & Physical examination (they usually come due to a photoreaction)
2. Skin Biopsy (based on the location, if it was in the face > no biopsy)
3. **Serology:**
   a) ANA :+ve in 95% ,VERY SENSITIVE BUT NOT SPECIFIC
   b) Anti-dsDNA (anti- native DNA): Specific but not very sensitive, indicates high risk for renal disease. (SLE nephritis)
   c) Anti-smith : most specific +ve in 30%
   d) Anti-histone Ab (drug induced lupus)
   e) Rheumatoid factor : +ve in 30%
4. CBC, ESR (leukopenia in SLE)
5. Urine analysis (protein and casts because the kidney is silent)
6. C3,C4: : low levels indicate active disease, often with renal involvement.

7. Lupus Band Test :

- It's preferred to be done on nonlesional nonexposed skin (for SLE) , but can be performed on lesional skin.(for DLE)
- Positive DIF supports the diagnosis of Cutaneous LE, negative DIF does not exclude the diagnosis.

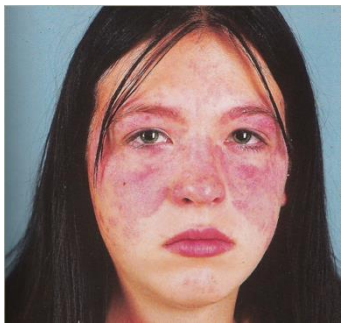


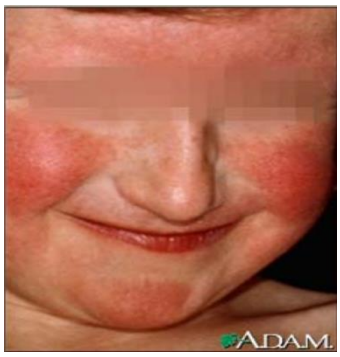
- Clinical, histopathology, serology are more important.
- Positive antibody of normal appearing skin correlates with SLE.
- Granular deposits of immunoglobulins and complement are detected in a band-like pattern at the dermal-epidermal junction.

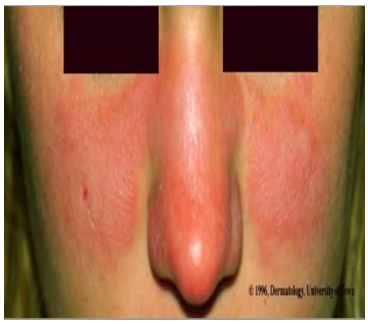
How is ACLE managed ?

• The aim of treatment for cutaneous LE is to alleviate symptoms and to prevent scarring.

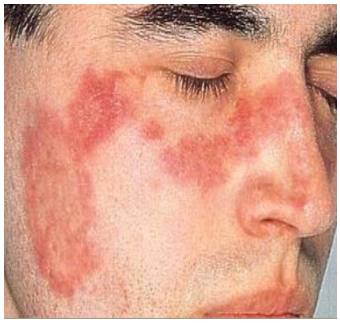
1. Sun protective measures number one method is sun avoidance
2. Potent topical steroids
3. Antimalarial drugs. Hydroxychloroquine to treat skin and joint. S.E> retinal toxicity and flashes
4. Oral steroids.
5. (if not responding) Methotrexate, azathioprin, mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG(immunoglobulin but not first line), and Rituximab.

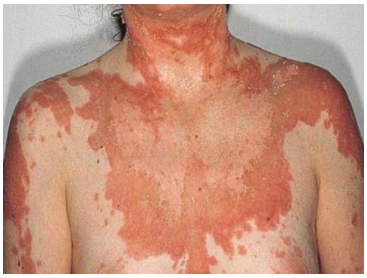


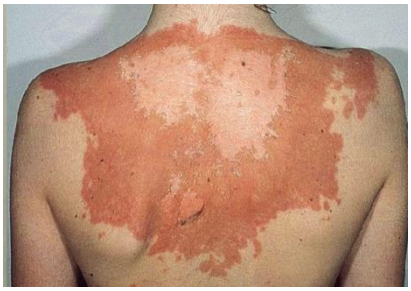




Sparing of nasolabial folds
unlike seborrheic dermatitis







2) Subacute Cutaneous Lupus Erythematosus

- More persistent than those of ACLE (weeks- months)
- Scaly, superficial, inflammatory macules, patches, papules and plaques that are photodistributed, particularly on the upper chest & back, lateral neck, and dorsal arms & forearms.
- **Morphologic subtypes:**
  - Annular/polycyclic lesions " ring-shaped"
  - Papulosquamous lesions:
    - psoriasiform
    - pityriasiform

Do patients with SCLE have SLE?

- About 1/2 patients with SCLE will have 4 or more criteria for the classification of SLE
- Skin disease, photosensitivity, musculoskeletal complaints
- 60-100% have anti Ro antibody.
- 10-15% develop significant internal disease including nephritis.
- SCLE could occur in patients with Sjögren syndrome, deficiency of the second component of complement (C2).
- Drug induced SCLE: hydrochlorothiazide, terbinafine, calcium channel blockers, NSAI.

How do you make the diagnosis of SCLE?

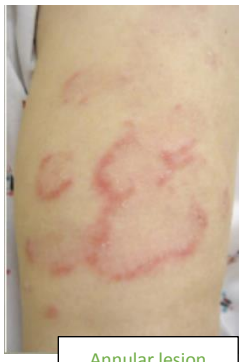
- SCLE is a clinical diagnosis based on presence of:
  - Typical photodistributed eruption
  - Skin biopsy
- Direct Immunofluorescence
- A strong association exists with anti-Ro/SS- A autoantibodies, and a lesser extent will have anti-La/SS-B

What is the initial workup of SCLE?

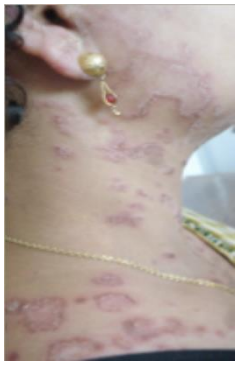
1. History & Physical examination
2. Laboratory testing
3. Medication History

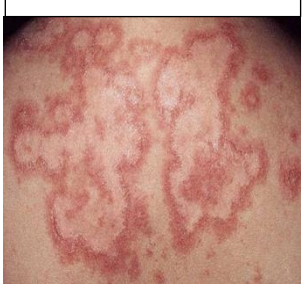
How is SCLE managed ?

- Broad-spectrum sunscreens
- Sun-protective measures
- Topical steroids
- Antimalarial drugs

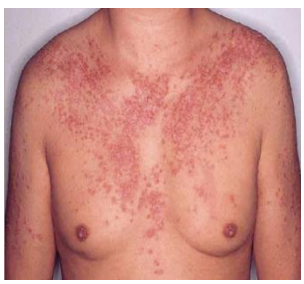


Annular lesion

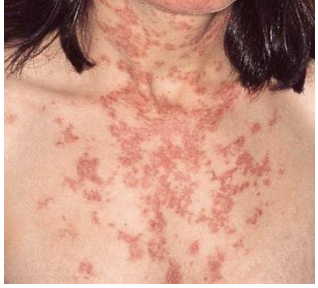




Annular lesion



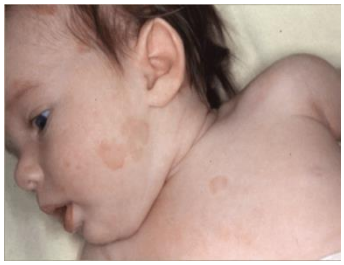
Psoriasiform?

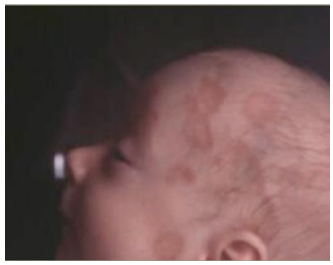


Psoriasiform?

Neonatal Lupus Erythematosus (NLE)

- The skin lesions occur on the face and head, morphologically resemble SCLE lesions(annular), they are transient, resolving within a few months*.
- In NLE , infants develop skin disease (50%), congenital heart block (50%), or both (10%).
- It's usually permanent and may require a pacemaker.
- 10% of infants with NLE and heart disease die from cardiac complications.
- Thrombocytopenia/ liver disease.
- anti-Ro (100%) and other antibodies as well as the mother.
  For mom and baby> we check 1.ANA and anti-rho with anti-la and anti-U1RNP (for CT disease) ,2.ECG, 3. CBC for thrombocytopenia 4. LFT for hepatobiliary disease also 5.biopsy





3) Chronic Cutaneous Lupus Erythematosus

- Several types of cutaneous LE that are very persistent are termed Chronic cutaneous lupus erythematosus.
- The most common of these chronic forms is Discoid lupus erythematosus " DLE"
- Serologic abnormalities are uncommon (only 5% are positive)

A) Discoid lupus erythematosus:

Describe the skin changes that occur with discoid lupus erythematosus?

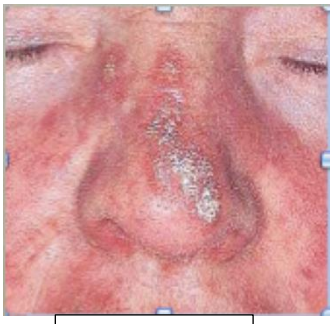
- Localized or generalized
- Chronic, fixed, indurated, erythematous papules and plaques often distributed over the head& neck.
- Scarring (One of the indications of Potent steroid!)
- Pigmentary changes (hypo/hyperpigmentation)
- Epidermal changes: scales, keratotic plugging of hair follicles, crusting
- External ears

Do patients with DLE develop systemic lupus erythematosus?

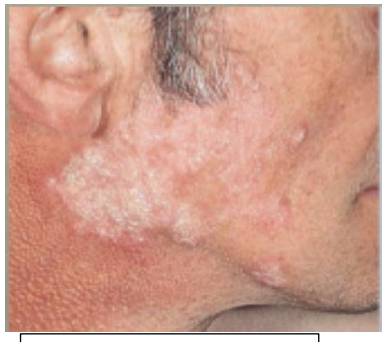
- Risk of developing SLE is 5%-15% (slightly higher risk if DLE is generalized(generalized> 15%)).
- 25% of SLE patients will develop lesions of DLE at some time during the course of their disease.

How is Discoid Lupus treated?

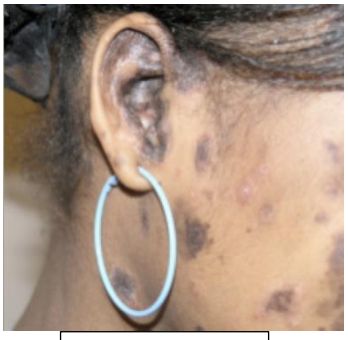
- Sunscreens
- Sun-protective measures
- Potent topical steroids/ intralesional steroids
- Antimalarial drugs(for skin and joint)



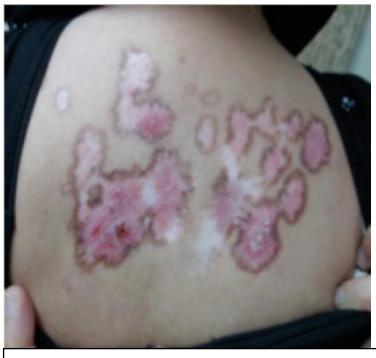
Keratotic plaques



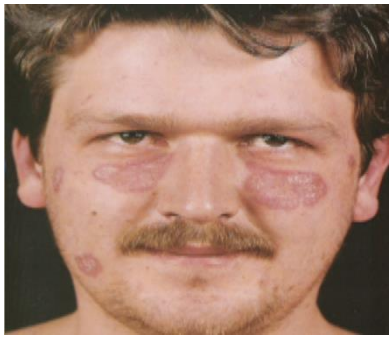
Hypopigmentation and scarring

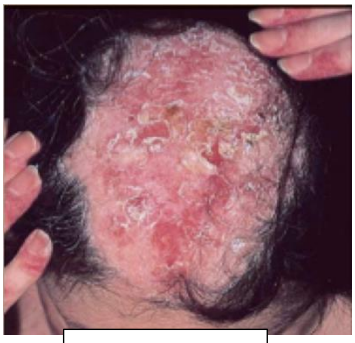


Brown scaly DLE

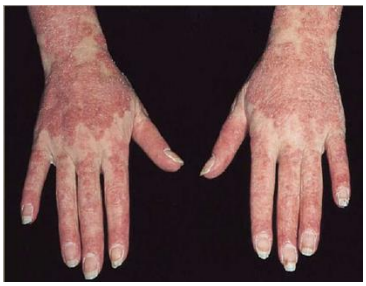


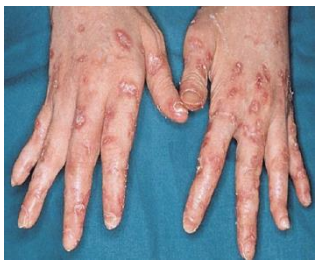
Annular lesions that are pigmented on the periphery with scarring





Scarring alopecia





B) Drug-Induced Lupus Erythematosus

Drug-induced lupus differs from SLE by the following features:

- Sex ratios are nearly equal.
- Arthralgia, myalgia, pleuritis, fever, lacking nephritis, CNS disease.
- Procainamide, hydralazine.
- Antihistone antibodies.
- No antibodies to native DNA or hypocomplementemia are present.
- When the drug is discontinued, the patient has resolution of clinical manifestations and reverting of abnormal laboratory values to normal.

What is minocycline-induced lupus? (minocycline Is used for acne)(you have to follow up)

- Polyarthralgia, fatigue, fever, elevated liver enzymes, pneumonitis, anemia, lupus specific skin eruption have not been reported
- All patients have positive ANA

  If you have cough, arthritis , or fever > stop the drug

Drugs associated with lupus erythematosus:

Chlorpromazine	Isonizide
Hydralazine	procinamide
Methyldopa	Quinidine
Anti- TNF	Minocyclin
Penicillamin	IFN-alpha, IFN-beta

2) DERMATOMYOSITIS

- Autoimmune disease of uncertain etiology.
- Involving skin and proximal muscle weakness. (shoulder and pelvis)
- Juvenile, adult onset.
- Amyopathic dermatomyositis- in some instances, muscle involvement may not be detectable.(both clinically and radiologically)
  However if clinically there wasn't muscle involvement, but radiologically there was > this is hypo myopathic dermatomyositis.
- Making early diagnosis is important because it is a serious but treatable multi system disease

Criteria for diagnosing dermatomyositis:

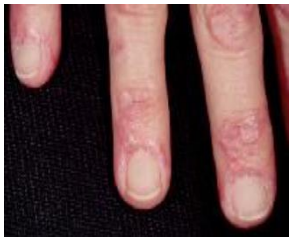
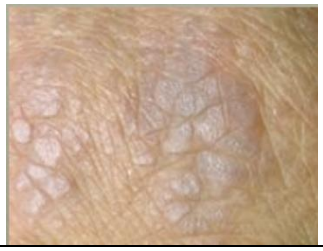
- Progressive proximal symmetrical weakness (difficulty combing hair, sitting up, dysphagia)
- Elevated muscle enzyme levels
- Abnormal findings on electromyograms
- Abnormal findings from muscle biopsy.
- Compatible cutaneous disease.

Diagnosis :

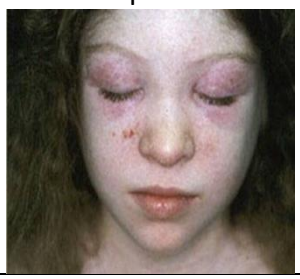
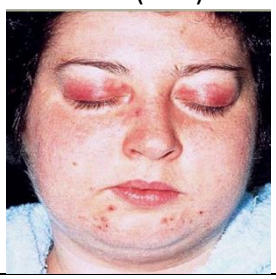
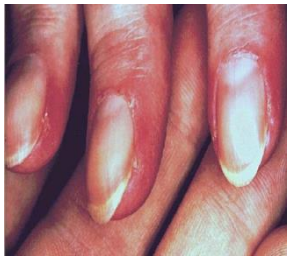
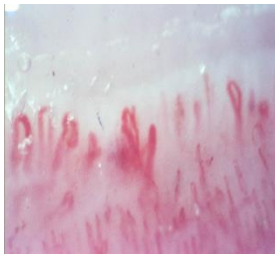
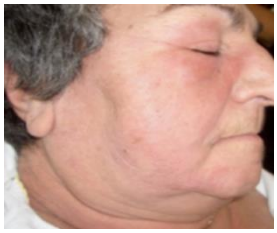
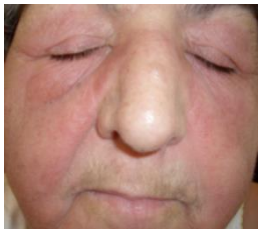
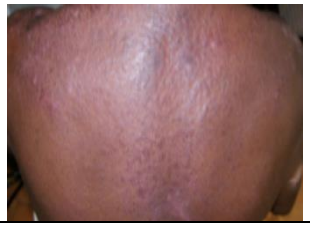
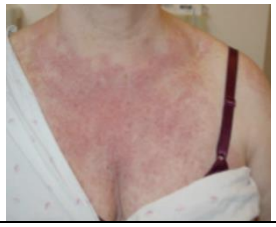
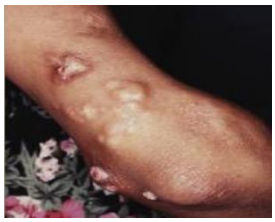
- Proximal muscle weakness with two of the three laboratory criteria

Are there skin changes that are diagnostic of dermatomyositis?

Two cutaneous findings have been describes as "pathognomonic" of dermatomyositis:

1. Gottron's papules	2. Gottron's sign
Erythematous to purplish flat papules on the extensor surfaces of the interphalangeal joints (knuckles)  	Consist of symmetric violaceous erythema, sometimes with edema, over the dorsal knuckles of the hands, elbows, knees, and medial ankles. 

Are there other skin findings that are characteristic of dermatomyositis?

Heliotrope rash	Symmetrical periorbital edema with a violaceous (lilac) dusky erythema  
Periungual talangiectasia with cuticle atrophy (mechanic hands)	 
Photodistrbuted violaceous erythema	Face, sun-exposed areas of the neck, upper chest, shoulders, dorsal arms, forearms, and hands. (blotches of erythema)  
Shawl sign	Highly associated with interstitial lung disease  
Calcinosis	Of skin or the muscle. Manifests as firm, yellow or flesh-colored (chawky) nodules, often over bony prominences. (juvenile DM) in 25%-70% of patients 

Are there any diseases associated with dermatomyositis?

Malignancies (ovarian is difficult to diagnose> need CT of pelvis)

- It can be associated with other connective tissue diseases such as lupus, rheumatoid arthritis, scleroderma and Sjogren's syndrome.
- 25% of adults with DM have associated malignancy.
- Female patients should be carefully screened for ovarian cancer.

1.CT diseases
2. malignancies
3.calcinosis cutis
4.fulminant lung disease

- Juvenile type associated with calcinosis cutis.
- Fulminant lung disease is reported in amyopathic DM and underlying malignancy, therefore both malignancy screening and systemic involvement are recommended irrespective of muscle involvement.(even if amyopathic dermatomyositis, it still could have malignancy)
- Oropharyngeal and upper esophageal muscle involvement may lead to dysphagia, nasal regurgitation or aspiration.
- Respiratory failure due to weakness of the diaphragm and chest wall muscles.
  Respiratory failure can be due to respiratory muscle weakness or interstitial lung disease)

What is a serious complication in DM?

- ILD in 10% of cases
- often occurs with antisynthetase antibodies (t-RNA) antibodies to aminoacyl-transfer ribonucleic acid synthetase enzymes
- 30% of DM have antisynthetase syndrome (ILD raynaud's phenomenon, arthritis, mechanics hands)

Anti Jo >> ILD

How do you diagnose dermatomyositis?

1. History & Physical examination
2. Serum levels of muscle enzymes- creatine phosphokinase (CPK) level is most reliable indicator of disease activity. Other muscle enzymes: aldolase, AST, ALT, LDH.
3. **Serology:**
   - ANA in < 60%
   - Anti-Jo-1 (anti-histidyl-t-RNA synthase) in 30%
   - Anti-Mi-2 (highly specific), but it lacks sensitivity because its present in only 25% of patients, indicates good prognosis.
4. Magnetic resonance imaging (MRI), sensitive, not specific.
5. U/S of muscle.
6. Electromyogram, sensitive. Not specific. Normal in 10% of patients
7. Muscle biopsy- Inflammatory cell infiltrations & necrosis of muscle cells. Best to choose biopsy side based on MRI findings.
8. Skin biopsy- suggestive but not diagnostic (unlike DLE where we have to do skin biopsy), shows interface dermatitis.
9. PFTs, Co diffusion
10. CXR, high resolution chest CT for ILD
11. ECG
12. GIT symptoms, barium swallow
13. CMP, CBC, diff, ESR
14. Malignancy screen for adults

What are the indicators of poor prognosis?

- Malignancy
- Cardiac involvement
- Older age
- Progressive disease
- initiation of therapy after 24 months of muscle weakness
- Longer duration of symptoms before diagnosis
- Pulmonary disease
- Dysphagia

- Extensive cutaneous involvement of the trunk

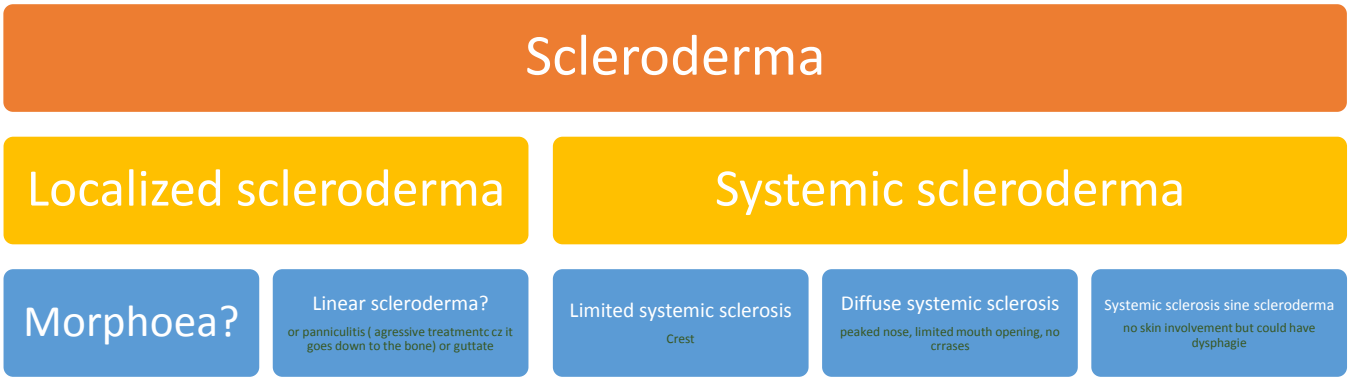
How is dermatomyositis treated?

- Oral steroids are the mainstay treatment.
- Steroid sparing agents- Methotrexate, azathioprin, mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG, and Rituximab.
- Topical steroids and antimalarial medications are used to improve the cutaneous rashes.
- Physiotherapy to improve strength and flexibility of the muscles.
- For Calcinosis cutis (juvenile): diltiazem and or surgical excision, low dose warfarin

If we want to put the patient on immunosuppressive medication , we have to do Quantiferon test to rule out Latent TB.

3) Scleroderma

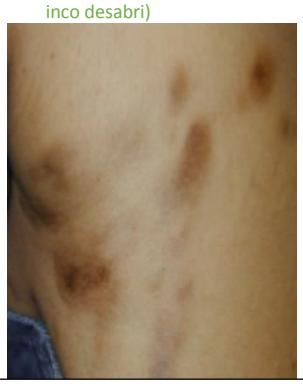
- An autoimmune connective tissue disease of unknown etiology
- Chronic disease that involves the microvasculature and connective tissue and results in fibrosis.
- There is an increase in dermal collagen & decrease in the elastic tissues which leads to typical thickening & immobility (absent creases)
- It may be localized, as in morphea, or more generalized and involving visceral organs, as in progressive systemic sclerosis.



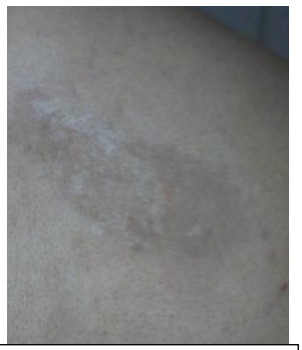
This Graph is very imp!

A) Morphea

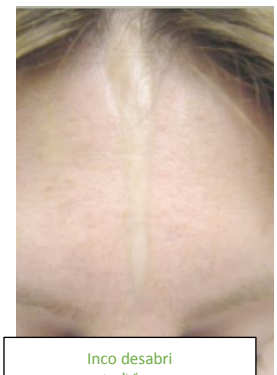
- Sclerotic (hard), indurated plaques that may be solitary, multiple, linear, or generalized or panniculitis
- The surface is usually smooth, with the center of the lesion a whitish or ivory color, whereas the border of active lesions is usually violaceous (in early lesion).
- It usually involves the skin and subcutaneous tissues but involve deeper structures, even bone. (inco desabri)



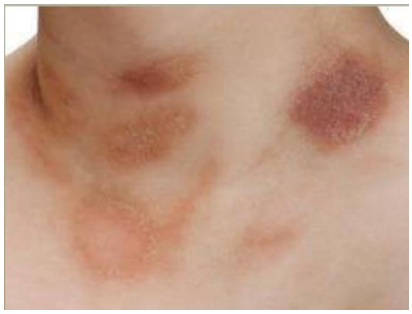
Burned out morphea , pigmentation

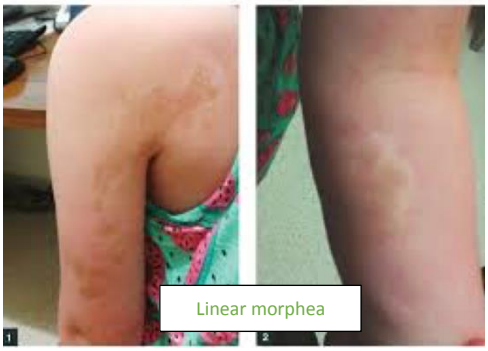


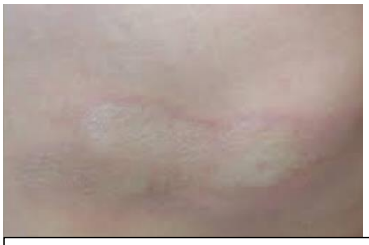
Pigmentation and ivory white
When touch skin, we cant pinch it



Inco desabri
ضربة السيف
Up to bone and muscle







Typical early lesion: violaceous erythema in border and ivory white in center with hardening sclerotic skin

Do patients with morphea develop systemic sclerosis? No

How is morphea treated ? based on location.

- Morphea has no known cure.
- Treatment of morphea focuses on controlling signs and symptoms and slowing spread.
- Topical and intralesional steroids , phototherapy, systemic 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 in movement

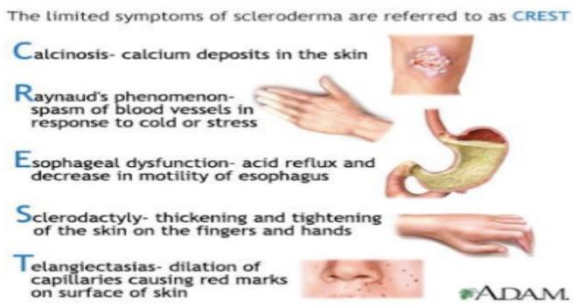
Phototherapy > panniculitis > we use UVA1

Again, with immunosuppressive medication, do quantiferon test to rule out latent TB.

B) limited systemic scleroderma

CREST Syndrome

- It's considered a type of limited systemic scleroderma
  - C = Calcinosis cutis
  - R = Raynaud's phenomenon
  - E = Esophageal dysfunction
  - S = Sclerodactyly (tightening of fingers)
  - T = Talangiectasia
- Most patients with CREST syndrome have circulating antibodies to centromeres, called " anti-centromere antibodies"



C) Progressive Systemic Sclerosis

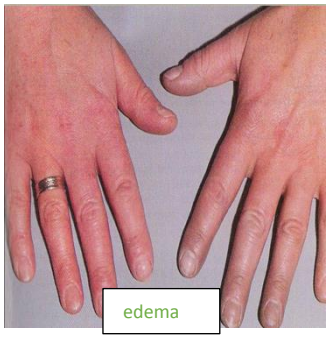
What are the cutaneous findings in progressive/diffuse systemic sclerosis?

Edema of the fingers is the first sign

1. Swelling of the hands and feet and/or Raynaud's phenomenon (for years)
2. Telangiectasia
3. Proximal nail fold changes (avascular areas)
4. Thickening and sclerotic changes involving the face& extrimities- progressive

- Sclerodactyly- tapering of fingers, with waxy , shiny hardened skin,which is tightly bound down & doesn't permit folding or wrinkling
- Loss of normal facial lines (mask like) patient looks younger than they are
- Thinning of lips, microstomia, radial perioral furrowing , small sharp nose (can't open mouth by 3 fingers)
- Digital ulcers +/- loss of digits


blanching


edema

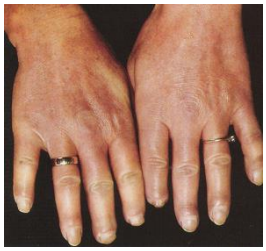
Raynaud's Phenomenon can be primary or secondary

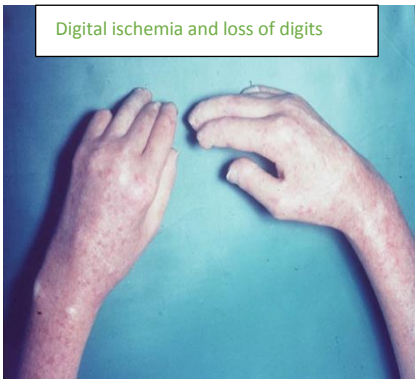
- It is digital ischemia that occurs on exposure to cold and/ or as a result of emotional stress.

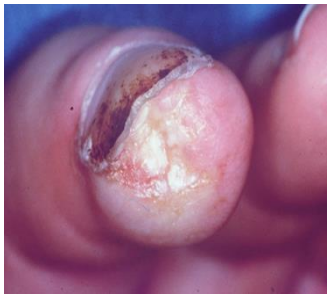
Causes:

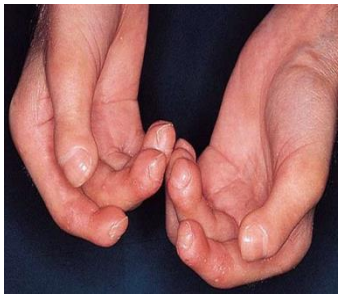
1- Rheumatic disorders(SS 85%,SLE 35%, DM 30%, RA, PAN)
2- Diseases with abnormal blood proteins (cryoprotein, macroglobulins)
3- Drugs (b-adrenergic blockers, nicotine,cyclosporine)
4- Arterial disease (atheriosclerosis obliterans)

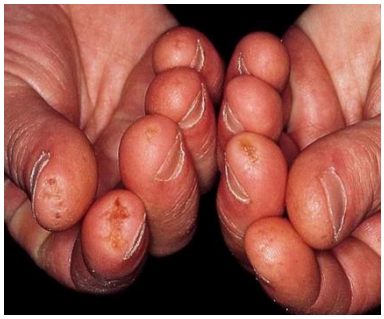
Nonpitting edema of the hands & feet.

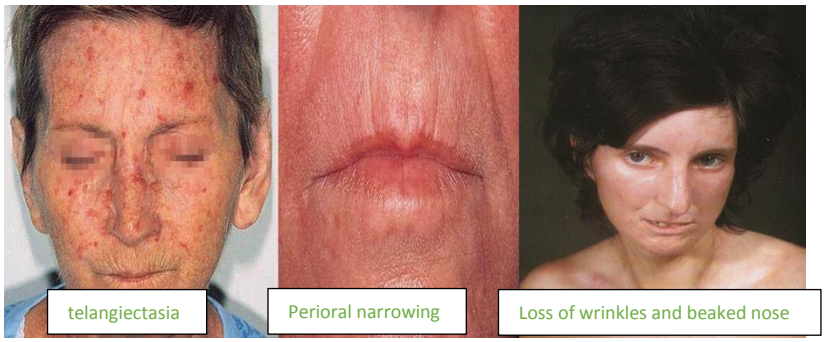



Digital ischemia and loss of digits








telangiectasia

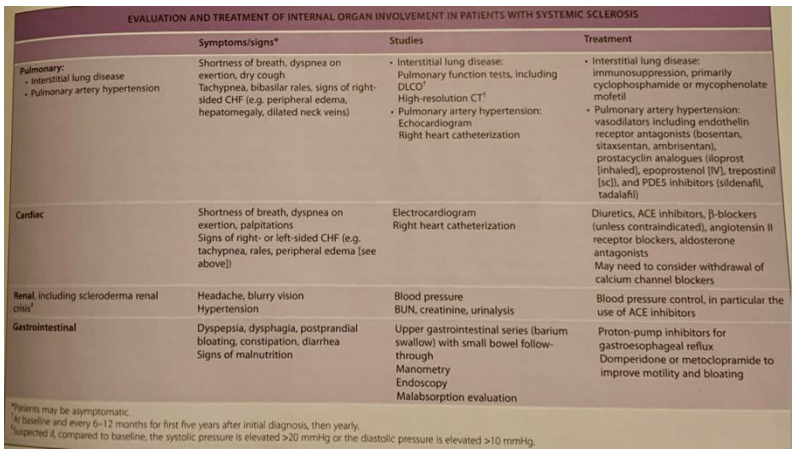
Perioral narrowing

Loss of wrinkles and beaked nose

How do you diagnose scleroderma?

1. History & physical examination- characteristic skin changes
2. Serology
    - ANA (often +ve)
    - Anti-centromere antibodies
        - 71% LSSc " CREST"
        - 21% of DSSc " progressive"
    - Anti-Scl-70 " anti-topoisomerase I"
        - 33% of dSSc
        - 18% of CREST

3. Skin biopsy- skin atrophy with preservation of skin appendages.



EVALUATION AND TREATMENT OF INTERNAL ORGAN INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS

	Symptoms/signs*	Studies	Treatment
Pulmonary: • Interstitial lung disease • Pulmonary artery hypertension	Shortness of breath, dyspnea on exertion, dry cough Tachypnea, bibasilar rales, signs of right-sided CHF (e.g. peripheral edema, hepatomegaly, dilated neck veins)	• Interstitial lung disease: Pulmonary function tests, including DLCO† High-resolution CT† • Pulmonary artery hypertension: Echocardiogram Right heart catheterization	• Interstitial lung disease: immunosuppression, primarily cyclophosphamide or mycophenolate mofetil • Pulmonary artery hypertension: vasodilators including endothelin receptor antagonists (bosentan, sitaxsentan, ambrisentan), prostacyclin analogues (iloprost [inhaled], epoprostenol [IV], treprostinil [scl], and PDE5 inhibitors (sildenafil, tadalafil)
Cardiac	Shortness of breath, dyspnea on exertion, palpitations Signs of right- or left-sided CHF (e.g. tachypnea, rales, peripheral edema [see above])	Electrocardiogram Right heart catheterization	Diuretics, ACE inhibitors, β-blockers (unless contraindicated), angiotensin II receptor blockers, aldosterone antagonists May need to consider withdrawal of calcium channel blockers
Renal, including scleroderma renal crisis‡	Headache, blurry vision Hypertension	Blood pressure BUN, creatinine, urinalysis	Blood pressure control, in particular the use of ACE inhibitors
Gastrointestinal	Dyspepsia, dysphagia, postprandial bloating, constipation, diarrhea Signs of malnutrition	Upper gastrointestinal series (barium swallow) with small bowel follow-through Manometry Endoscopy Malabsorption evaluation	Proton-pump inhibitors for gastroesophageal reflux Domperidone or metoclopramide to improve motility and bloating

*Patients may be asymptomatic.
†At baseline and every 6–12 months for first five years after initial diagnosis, then yearly.
‡Suspected if, compared to baseline, the systolic pressure is elevated >20 mmHg or the diastolic pressure is elevated >10 mmHg.

This table is not important

How do you manage a patient with scleroderma?

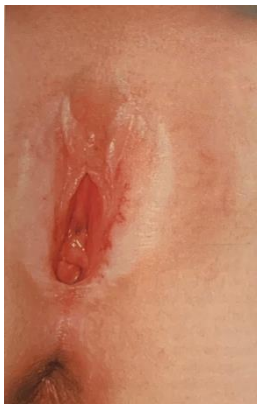
- Treatment is symptomatic.
- Raynaud's phenomena:
    - Stop smoking
    - keep hands warm and decrease trauma
    - calcium channel blockers* (nifedipine)
    - aspirin
    - vasodilating drugs (iloprost)
- Calcinosis cutis: nifedipine, surgical or laser excision.
- Skin sclerosis: physiotherapy, phototherapy.
- GI: proton pump inhibitor, surgery for strictures.
- Kidney: ACE inhibitors. IMP
- In severe cases: immunosuppressant , D-Penicillamine might be used (blocks aldehyde groups involved in intermolecular cross-links in collagen)

***An important thing to remember in SLE is antiphospholipid syndrome in which there will be recurrent abortions and tendency for thrombosis(CVA or stroke) , they will also have elevated PTT

4) Lichen Sclerosus Atrophicus (LSA)

- Sclerotic, ivory white, flat papule and plaques with epidermal atrophy and in extra mucosal sites, follicular plugging . if LSA involves the skin> rule out mucosa

- Most commonly affects female or male genitalia (prepubertal children) , less often extra genital skin may cause scarring of the vaginal introitus or phimosis if not diagnosed and treated early
- Vulval pruritus that's not due to candida or warts
- No systemic manifestations
- Diagnosed by skin biopsy!
- Treatment of choice potent topical steroids (clobetasone dipropionate for longer period to prevent scarring) is not given PRN. If u give 7 year steroids > u will prevent strictures but don't give PRN (intermittent)



Sclerosis around (whitish)
Purpuric lesion
Excoriation

Its an autoimmune condition
Could be a subset of morphea (a lot of similarities between them)
Excoriations > take biopsy
If treated early> you will prevent strictures

Questions

1) You see a patient with chronic scarring alopecia. On examination, you notice fixed, indurated, erythematous papules and plaques on the face and ears. There are also areas of scarring, hyperpigmentation and hypopigmentation. On the scalp, there is scale and keratotic plugging of the hair follicles. What is the most likely diagnosis?
A- Tinea capitis
B- Discoid Lupus Erythematosus.
C- Lichen planus
D- Psoriasis

2) Patient complaining of Raynaud's phenomenon, what is the drug that can cause it?
A- Doxycycline.
B- Beta blockers.
C- Clindamycin.
D- Calcium Channel blockers.

3) Patient came to the ER complaining of SLE like manifestation taking minocycline. What is the first step to manage him?
A- Discontinue the minocycline.
B- Systemic Steroids.

4) Which of the following is a Clinical pathognomic feature of dermatomyositis :
A-Gottron papules
B-vasculitis
C- poikiloderma
D- calcinosis

5) Which one of the following is a very characteristic histopathological feature of scleroderma:
A-increased collagen
B-decreased blood vessel

C-normal eccrine gland size and number
D-normal hair follicle number

6) 19 years old girl presented with arthritis and photosensitive rash on her face, what is the diagnosis:
A- Scleroderma
B- SLE
C- Dermatomyositis
D- Vitiligo

7) Which one of the following is not included in the criteria for diagnosing SLE?
A- Pericarditis
B- Muscle pain
C- Seizure
D- Oral lesion

8) Which one of the following is not considered as part of the diagnostic approach of Dermatomyositis?
A- Magnetic resonance imaging (MRI)
B- Electromyography (EMG)
C- Muscle biopsy
D- Skin biopsy

1	2	3	4	5	6	7	8
B	B	A	A	A	B	B	C