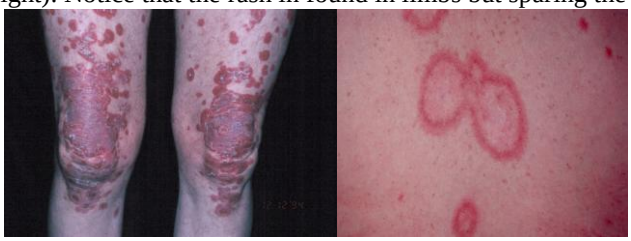
Unit IX – Problem 4 – Clinical: SLE

- **There are three major subtypes of cutaneous lupus erythematosus (CLE):**

Subtype	Description
Acute CLE	• Mainly in SLE. • Sharply demarcated, macular erythematous lesions (indurations التيبُّس and scaling might occur). • Localized (face: butterfly) or generalized (in any area exposed to sunlight). • Typically painful and itchy. • Heals without scars or atrophy.
Subacute CLE	• The most photosensitive one of all types. • Patients have anti-SSA/Ro antibodies. • Rash is scaly (مُتَقَشِّر), erythematous, papulosquamous (image: left) or annular (image: right). Notice that the rash in found in limbs but sparing the face.  • Mostly induced by medications (example: hydrochlorothiazide). • Heals without scars or atrophy.
Chronic CLE	• The most common subtype is DLE (Discoid Lupus Erythematosus). • Discoid term refers to the disc-shaped appearance. • Sharply demarcated, raised plages lesion with adherent scale with erythematous ring around the lesion. • Frequently localized to the scalp, face and neck. • Typically painless and not itchy. • Heals with scars and atrophy thus leading to permanent alopecia and disfigurement.

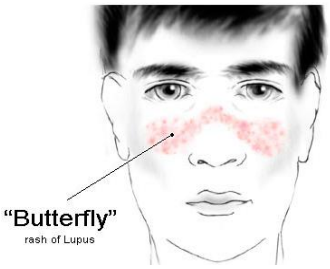
- **Epidemiology:**
  - **Female to male ration is (9-15):1** → 90% of these females are in their childbearing years (15-40 years).
  - Notice that people of all genders, ages and ethnic groups (المجموعات العرقية) are susceptible to the disease but the prevalence is variable between different sex and ethnic groups:
    - ✓ More in females than males.
    - ✓ More in blacks than white people.
- **Clinical manifestations of SLE:**
  - **General**: fatigue/ malaise, fever, anorexia and weight loss.
  - **Musculoskeletal**: arthralgia/ myalgia, non-erosive polyarthritis and myopathy/myositis.
  - **Cutaneous**:

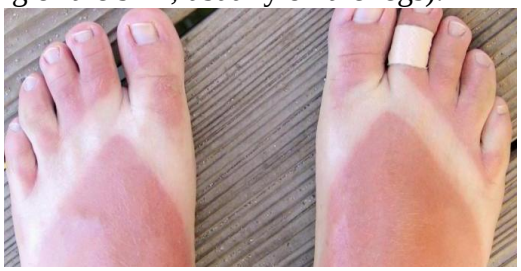
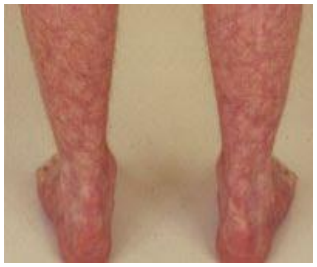
Common (20%-50%)	• Photosensitivity • Butterfly rash • Purpura/ petechiae • Chronic discoid lesions • Non-scaring alopecia  • Vasculitis of digits
Less common (5%-20%)	• Hyperpigmentation • Urticaria • Mucosal ulcers

	• Limb ulcers • Subcutaneous nodules
Occasional (< 5%)	• Periorbital edema • Pruritis • Paniculitis • Psoriaform lesion • Bullae • Jaundice • Severe scaring alopecia 

- **Hematologic**: anemia (normocytic normochromic/ autoimmune hemolytic anemia), leucopenia, lymphopenia and thrombocytopenia.
- **Renal**: lupus nephritis.
- **GIT**: abdominal pain.
- **Vasculitis**.

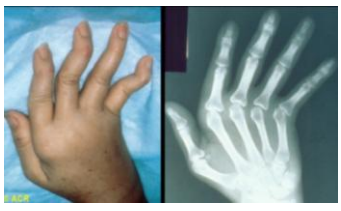
- **Butterfly rash:**
  - It is a fixed erythema (احمرار الجلد) –flat or raised- of the nasal bridge and malar bones (bones of the cheek). Notice that there is sparing of nasolabial folds.
  - This manifestation is found in more than 1/3 of SLE patients.
  - **It is not unique to SLE. It can also be seen in:**
    - ✓ Photosensitivity: which means sensitivity to sunlight. There is another skin condition which aggravated by exposure to cold and is known as livedo reticularis (bluish mottling of the skin, usually on the legs).



    - ✓ Infections.
    - ✓ Allergy (urticaria).
    - ✓ Drug eruption.
    - ✓ Systemic diseases.
    - ✓ Skin vasculitis.

- **Jaccoud's arthropathy (JA):**
  - It is a chronic deformity which is characterized by ulnar deviation and subluxation of the metacarpophalangeal joints. Notice that toes might also be affected.
  - This deformity is voluntarily correctable.
  - It was initially described as a complication of recurrent rheumatic fever.
  - It is associated with:
    - ✓ SLE.
    - ✓ Psoriatic arthritis.
    - ✓ IBD (Inflammatory Bowel Disease).
    - ✓ Malignancy.



- ✓ Pyrophosphate arthropathy.

- **Autoantibodies in SLE and other connective tissue diseases:**

Antinuclear antibody (ANA)	High sensitivity (> 95%) and low specificity for SLE
Anti-dsDNA	High sensitivity and specificity for SLE
Sm	Highly specific for SLE
U1 RNP	Mainly in mixed connective tissue disease
SSA/Ro	SLE
SSB/La	Sjogren's syndrome
Antiphospholipids: • **Anti-cardiolipin** • **Anti-β_2-GPI antibodies** • **Lupus anticoagulants.** • **Anti-prothrombin**	SLE

- **Diagnosis of SLE:**
  - **≥ 4 of the ARA criteria for classification of SLE:**
    - ✓ Butterfly rash.
    - ✓ Discoid rash.
    - ✓ Photosensitivity.
    - ✓ Painless oral ulcers.
    - ✓ Non-erosive arthritis.
    - ✓ Pleurisy/ pericarditis.
    - ✓ Renal features (nephritis).
    - ✓ Neurological features (interactable headache, seizures and psychosis).
    - ✓ Hematological features (autoimmune hemolytic anemia, leucopenia, lymphopenia and thrombocytopenia).
    - ✓ Immunological features (positive anti-DNA, antiphospholipid antibodies or anti-Sm test and false positive syphilis serology).
    - ✓ High titer of antinuclear antibody (ANA).
- **Management of SLE:**
  - **Conservative therapy:**
    - ✓ Rest.
    - ✓ Avoid stress.
    - ✓ Avoid exposure to heat and sunlight.
    - ✓ Diet (↓ fat, ↑ fish oil derivatives and ↑ protein).
    - ✓ Advice the patient to take calcium and vitamin-D.
    - ✓ Bisphosphonates.
    - ✓ Avoid estrogen pills.
  - **Drug therapy:**

Arthralgia/ fever	NSAIDs
Arthralgia/ myalgia and lethargy	Hydroxychloroquine
Malar/discoid rash	Prednisolone, hydroxychloroquine, thalidomide
Arthritis/ serositis/ myositis	Prednisolone
Autoimmune anemia or ITP	Prednisolone, azathioprine
Renal	Prednisolone, azathioprine
CNS	Prednisolone, anticonvulsant

- **Prognosis of SLE:**
  - Poor prognosis (50% mortality within 10 years of diagnosis.
  - 25% of patients may experience remissions, sometimes for a few years, but these are rarely permanent.