



CLINICAL FEATURES OF SYSTEMIC LUPUS ERYTHEMATOSUS

ORIGINS OF LUPUS

- ❖ Lupus is latin for wolf
- ❖ From the term for a french style of mask, (loup)



EPIDEMIOLOGY

Prevalence: 20-240 per 100/000 persons

Iran 40 per 100/000 person

Incidence: 1-10 per 100/000 person-years

Female to male ratio:

- * in adults: 10-15/1

- * in older onset: 3/1

- * in children: 8/1

age: 15-64y

SLE is more severe in men and children.

DEFINITION

SLE is a multisystem autoimmune disease

- ❖ Constitutional symptoms %90-%95
- ❖ Mucocutaneous involvement %80-%90
- ❖ Musculoskeletal involvement %80-%90
- ❖ Serositis %50-%70
- ❖ Glomerulonephritis %40-%60
- ❖ Neuropsychiatric involvement %40-%60
- ❖ Autoimmune cytopenia

MUCOCUTANEOUS

1. Acute: malar rash
2. Subacute
3. Chronic: Discoid lupus

Lupus panniculitis

LE tumidus / chilblain LE

1-2-3 are specific skin lesions.

❑ Non Specific: Oral lesions

Livedo reticularis

Raynaud's phenomenon

Vasculitis

Urticaria / Lupus hair / Alopecia

ACUTE CUTANEOUS LE

❖ ACLE

- Localized
- Generalized

❖ **100%** ACLE have SLE

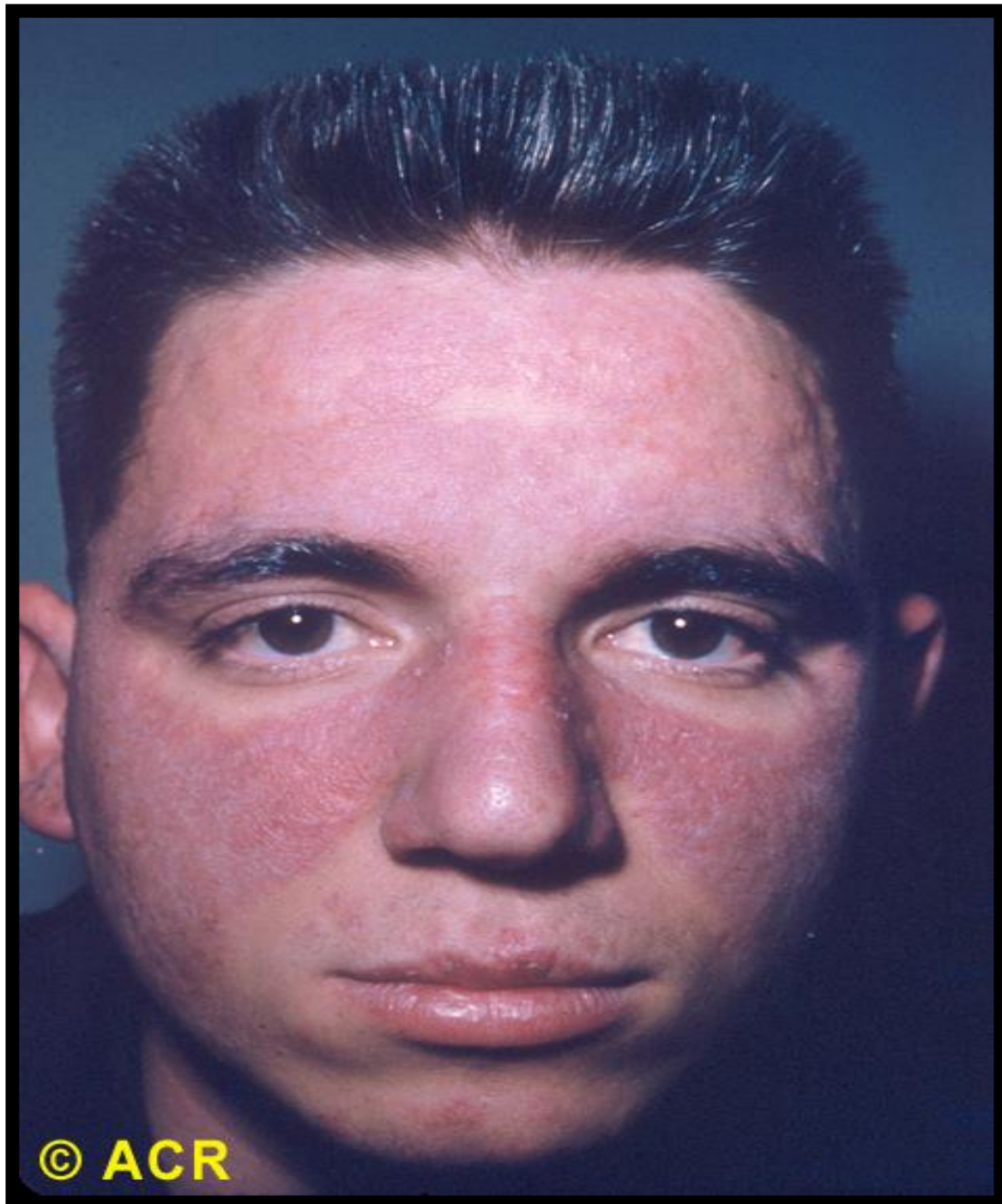
❖ ACLE lesions lasting days to weeks.

❖ ACLE lesions heal without scarring. postinflammatory hyperpigmentation may be observed.

MALAR RASH(LOCALIZED ACLE)

- ❖ The most classic form of rashes
(**30-60%**, IRAN :**61%**)
- ❖ Fixed erythema; flat or raised
- ❖ Abrupt in onset
- ❖ Initiated and/or exacerbated by exposure to sun light
- ❖ Photosensitivity
- ❖ F > M





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GENERALIZED ACLE

- Widespread macular or maculopapular erythem
- Distribute on any **photosensitive** area
- Palmar surface, dorsa of the hands, extensor surfaces of the fingers are commonly involved.
- Typically is located between the **IP joints**.



SUBACUTE CUTANEOUS LUPUS **ERYTHEMATOSUS**

❖ 7-27%

- Papulosquamous lesions
- Psoriasis like
- Annular-polycyclic lesions
- Peripheral scale & central clearing



- Sun exposed areas
- Back, neck, shoulder and extensor surfaces of the arms
- Usually spares the face.
- Superficial, non-scarring
- HLA-DR3
- Anti SSA/RO

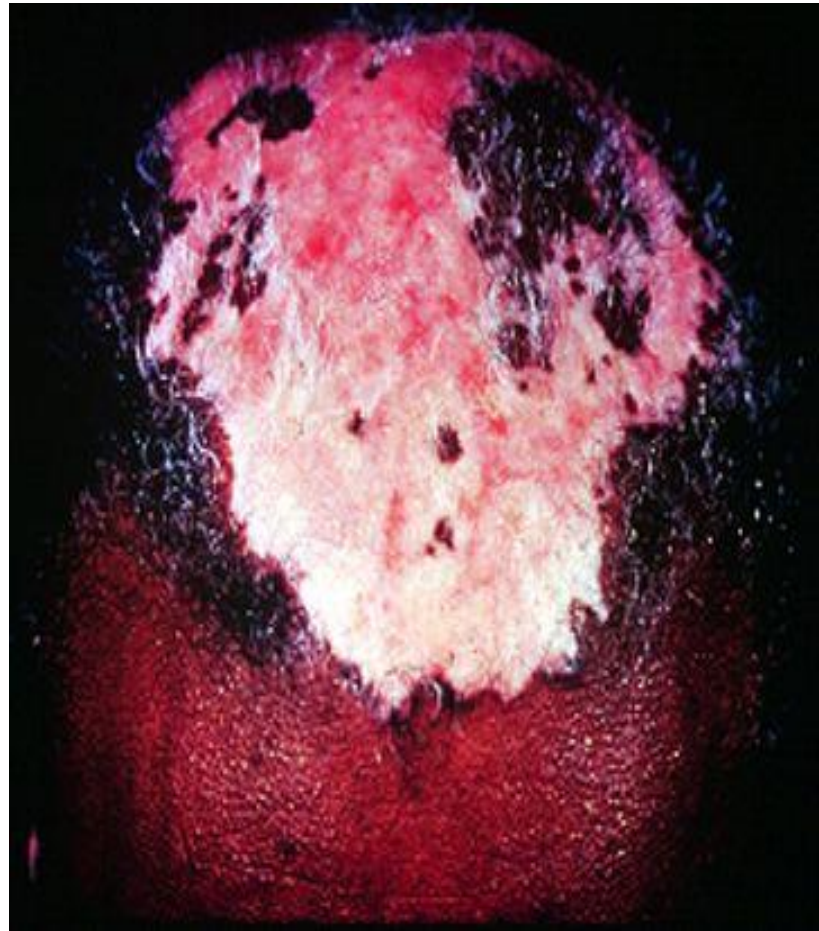


DISCOID LUPUSE

- ❖ **25%** of SLE (Iran: **15%**)
- ❖ risk of developing SLE **5-10%**
- ❖ Disk-shaped ,raised erythematosis plaques with adherent scale
- ❖ Localized and generalized
- ❖ Face, neck, scalp, ear, arm
- ❖ Diff Dx: lichen planus, eczema, actinic keratosis, psoriasis

DISCOID LE OF THE SCALP

- ❖ Localized discoid LE of the scalp with scarring, alopecia, and hypopigmentation





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GENERALIZED DLE

- ❖ Cutaneous lupus involving the face, ears, scalp, arms, and torso in a woman with leukopenia and a positive ANA but no systemic symptoms.



LUPUS PANNICULITIS

- ❖ Subcutaneous nodular lesions
- ❖ Firm, 1-3 cm
- ❖ Head, arms, chest buttocks, thighs

Tcell lymphoma
mimicking panniculitis



LUPUS PROFUNDUS

Sclerotic plaque with
center depression
(arrow) consistent
with lupus profundus



CHILBLAIN LUPUS

- ❖ Chilblain lupus characterized by reddish-blue nodules (in this case, on the fingers) occurring in **cold weather**. The lesions improved after the administration of nifedipin
- ❖ **Rarely** develop to **SLE**



LE TUMIDUS

- ❖ Rare
- ❖ Pink-violaceous papules ,
nonscarring plaques,
nodules



PHOTOSENSITIVITY

- ❖ The most common > 90%.iran 56%
- ❖ Abnormal cutaneous reaction to ultraviolet radiation (uvr)
- ❖ Occurring 1-2 weeks after exposure
- ❖ Worsening systemic symptoms (fatigue and joint pain)

LUPUS VASCULITIS

- ❖ Palpable purpuric lesions on the shins in a patient with lupus and necrotizing vasculitis of the skin, kidney, and brain.



VASCULITIS



VASCULITIS



LIVEDO RETICULARIS

- ❖ Patient with lupus and antiphospholipid antibodies with livedo reticularis (manifested by a reddish-cyanotic, reticular pattern of the skin) which has resulted in ulcer formation (arrows).





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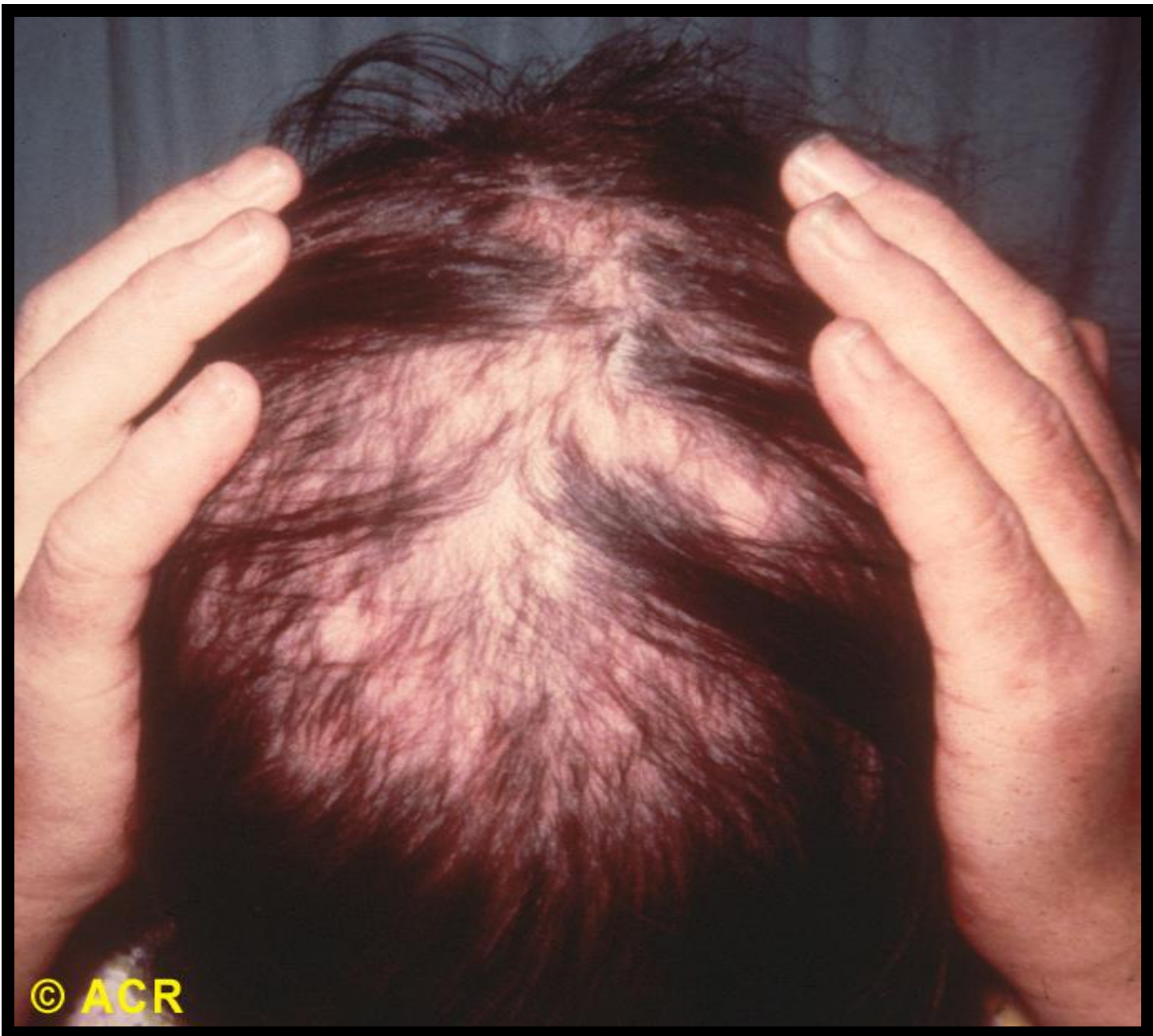


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LUPUS HAIR

- ❖ Short irregularly sized hair at the frontal hair line
- ❖ Associated with active SLE





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MUCOSAL ULCERS

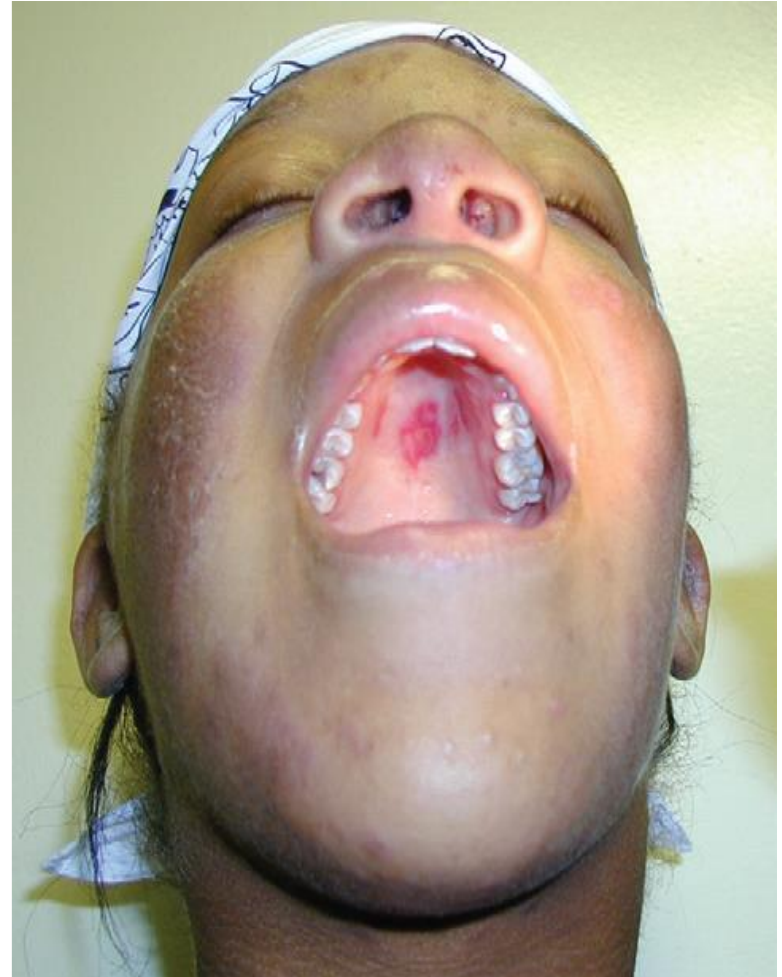
Acut: red macules, Palatal erythema or petechiae, erosions, painless.iran **38/5%**

Oral discoid lesion: painful, Red lesions with White radiating hyperkeratotic striae



ORAL ULCER IN LUPUS

- Chronic lesion



MUSCULOSKELETAL **INVOLVEMENT**

❖ Arthritis

❖ Avascular necrosis:iran 4/6%

❖ Myositis

ARTHRITIS

- ❖ Very common, 80-90%.iran 51/9%
- ❖ Mild joint pain.
- ❖ Symmetric inflammatory arthritis
- ❖ Hand deformity can occur but is reducible
- ❖ Jaccoud`s like arthropathy (hand and foot)



MUSCLE INVOLVEMENT

- ❖ Myalgia & muscle tenderness (%40-80)
- ❖ Myositis (%5-10)iran 2/9%
- ❖ Drug induced myopathy;glucocorticoid,Hcq
- ❖ Metabolic disorders, thyroid disease

RENAL INVOLVEMENT

- ❖ 90% in biopsy
- ❖ 50% clinically significant nephritis
- ❖ ↑ Morbidity and mortality
- ❖ Typically develops with the First 36 mo of the disease.

KIDNEY

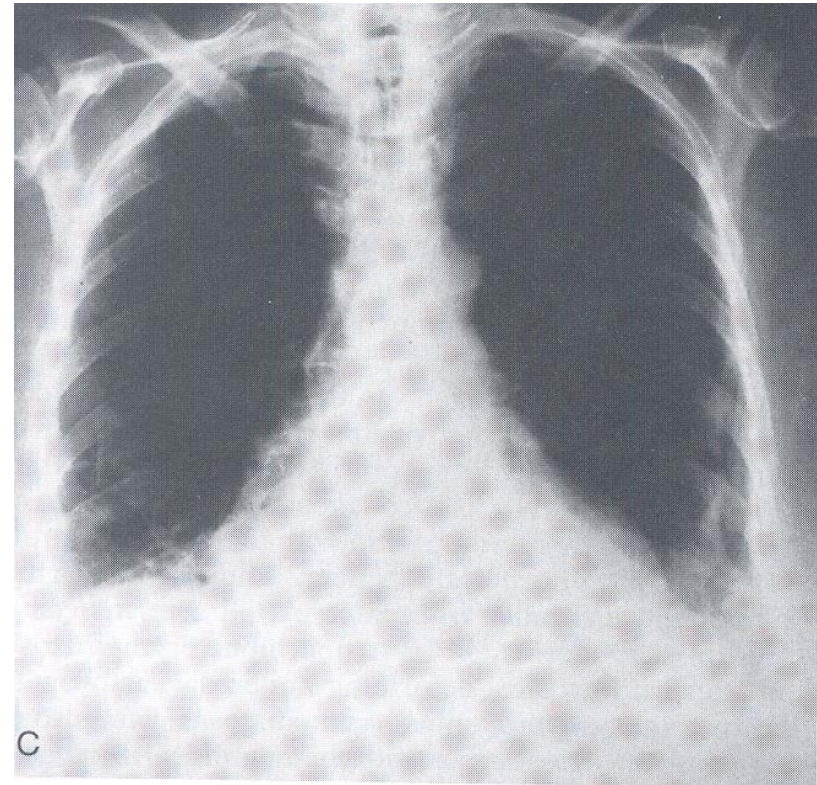
- ❖ Glomerulonephritis (most common form)
- ❖ Tubulointerstitial nephritis
- ❖ Vascular disease:
 - * Immune complex deposition
 - * Thrombotic microangiopathy
 - * Renal vein thrombosis
 - * Vasculitis
- ❖ Drug induced renal disease
- ❖ Infection

CLINICAL TYPES OF **LUPUS NEPHRITIS**

- ❖ Occult
- ❖ Chronic active nephritis
- ❖ RPGN
- ❖ Nephrotic syndrome
- ❖ HTN

PLEURITIS

- ❖ 50%(Iran: 16%)
- ❖ Pleural effusion:
 - ❖ SLE
 - ❖ Infection
 - ❖ Heart failure
 - ❖ Uremia
 - ❖ Malignancy
- ❖ Bilateral, Mild to moderate, Exudative
- ❖ Pleuritic chest pain, Friction rub
- ❖ Pleuritis corresponds to active SLE



ACUTE PNEUMONITIS

- ❖ Rare.iran 2%
- ❖ Fever. Cough. Pulmonary infiltration. Hypoxemia
- ❖ Cxray: bilateral, lower lobe, acinar Infiltration.
Often occur in conjunction
With a pleural effusion.
- ❖ Excluding other causes such
infection is necessary.
(bronchoscopy)
- ❖ High mortality rate > 50%



CHRONIC INTERSTITIAL LUNG DISEASE

- ❖ Rare
- ❖ May develop after acute pneumonitis or insidious fashion.
- ❖ Dyspnea on exertion, pleuritic chestpain, cough
- ❖ Must exclude infection & malignancy
- ❖ HRCT

DIFFUSE ALVEOLAR HEMORRHAGE

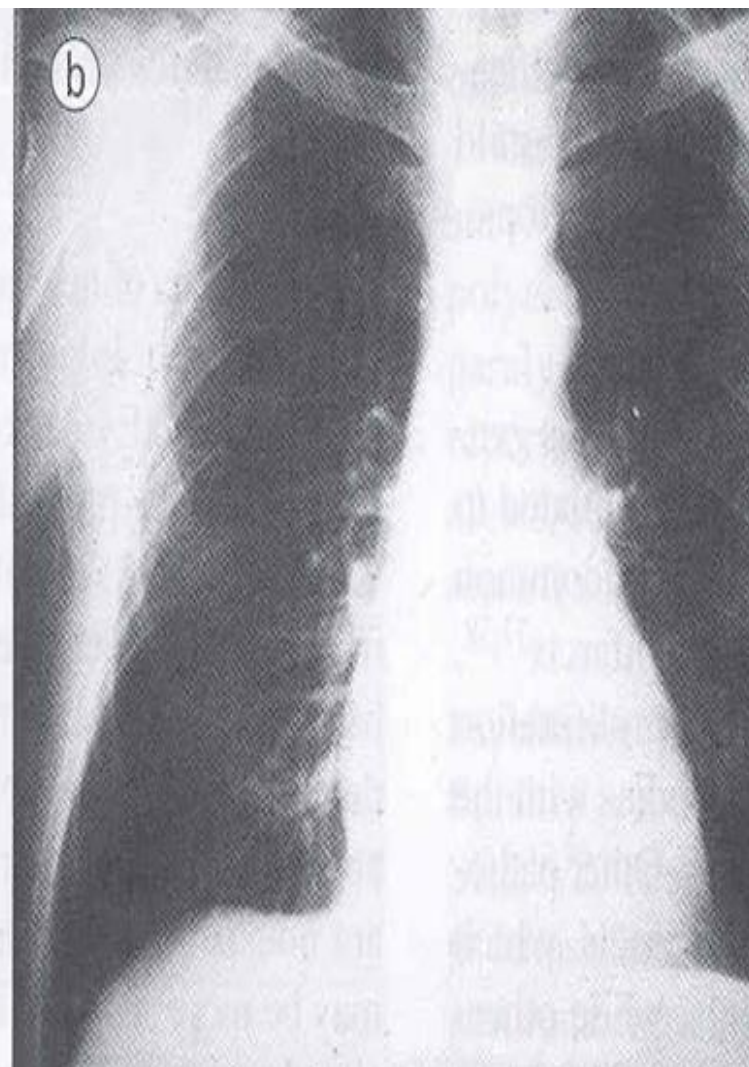
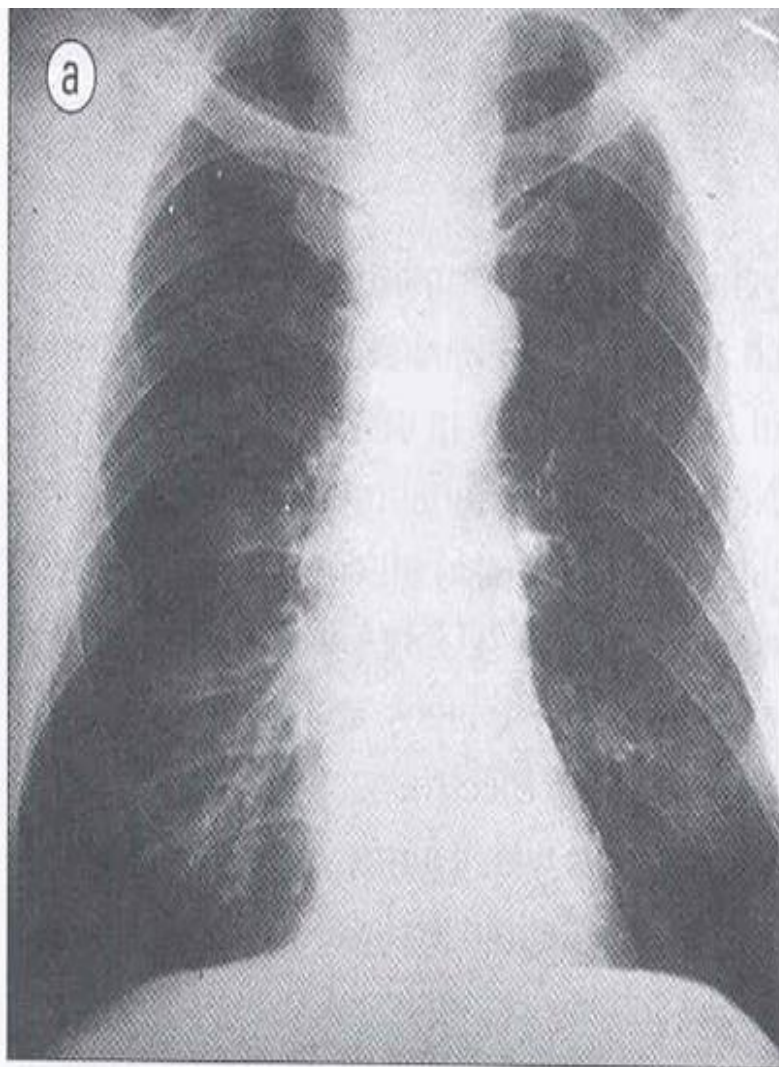
- ❖ Less than 2%
- ❖ Acute or sub acute onset of dyspnea and cough
- ❖ Falling in HB $\pm$ hemoptysis
- ❖ DLCO
- ❖ BAL is diagnostic
- ❖ High mortality rate > 50%

HEART

- ❖ Pericarditis: 50%.iran 9/2%
- ❖ Libman sacks endocarditis: 43%
- ❖ Valvular involvement: up to 61% iran 3/8%
- ❖ Myocardial involvement :8-25% iran 2/8%
- ❖ Coronary artery disease :25-40%
- ❖ Conduction disorder :35-70%

PERICARDITIS

- ❖ Most common cardiac manifestation
- ❖ Usually small. Asymptomatic
- ❖ Clinically may be present with sharp, precordial chest pain, friction rub, muffled heart sounds
- ❖ Chest pain improve in the upright position.
- ❖ Is related with active SLE
- ❖ ECO: choice
- ❖ ECG: Generalized ST elevation



MYOCARDITIS

- ❖ %8-25
- ❖ Anti RNP, anti RO/SS-A
- ❖ Clinical: Fever, dyspnea, palpitation, heart murmur, tachycardia
- ❖ CXR: Cardiomegaly

MYOCARDITIS

- ❖ **EEG**: ST-T Changes, sinus tachycardia, conduction disorder
- ❖ **ECO** :Left ventricular systolic or diastolic dysfunction,global hypokinesis
- ❖ **Endomyocardial** biopsy may be helpful in confirming the diagnosis and excluding other causes such as hydroxychloroquine

VALVULAR DISEASE

- Prevalence > 61%
- Valvular thickening, The most common abnormality. mitral and aortic valves
- Valvular stenosis
- Valvular regurgitation
- Libman – sacks endocarditis
- **Presence and progression of valvular disease were not associated with SLE disease activity**

LIBMAN – SACKS **ENDOCARDITIS**

❖ **43%**

❖ Classic cardiac lesion

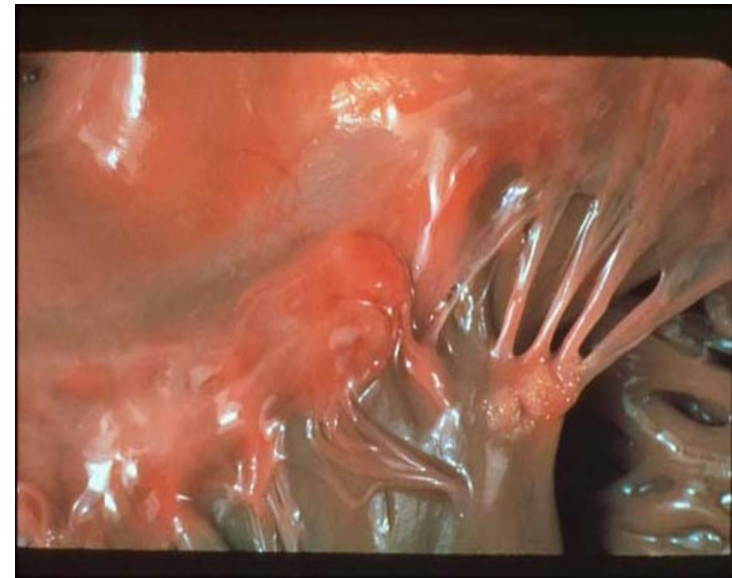
❖ Verrucous vegetation

❖ Most commonly on the ventricular aspects of the mitral valve

❖ All four valves involve. predominance of left sided

CLINICAL MANIFESTATION

- ❖ Frequency asymptomatic
- ❖ Emboli (rare)
- ❖ Bacterial endocarditis
- ❖ Valvular insufficiency
- ❖ First test:
- ❖ TEE is superior to TTE



CORONARY ARTERY DISEASE

- ❖ Atherosclerosis. (25-40% in a autopsy.)
- ❖ Young women: 50-fold higher risk of MI
- ❖ Risk factors: classic risk factors +SLE it self
- ❖ Chestpain, shortness of breath & silent MI & CHF
- ❖ Antiphospholipid antibodies: ↑IHD

- **NEROPSYCHIATRIC**
INVOLVEMENT

HEADACHES

- ❖ More than 50% of SLE patients
- ❖ Migrainous and tension-type headaches
- ❖ Evaluation of headache should be similar to that in a non SLE patient
- ❖ Evaluation of fever, Meningismus
- ❖ Altered mental state and focal neurologic signs.

SEIZURE

❖ 7-20% iran 13%

❖ The most common; complex partial seizure

❖ Grand, Petitmal, focal, temporal lobe

❖ Etiology: Acute inflammatory episode
old scar

❖ Risk factors: Antiphospholipid syndrome, uremia, malignant hypertension, infection, stroke, vasculopathy, high dose chloroquine, trauma

STROKE

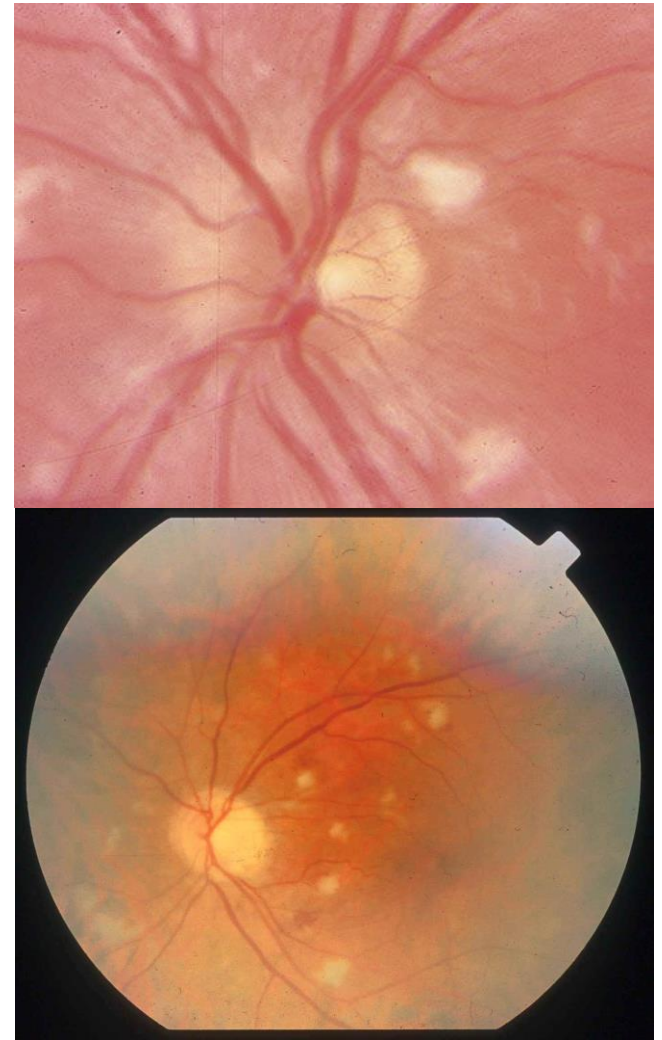
- ❖ 14/5 %
- ❖ The first 5 year
- ❖ Recurrent
- ❖ Risk factors: Hypertension, atherosclerosis, Infection, emboli, vasculitis, antiphospholipid syndrome, valvular disease

PSYCHOSIS

- ❖ 5%.iran 4/9%
- ❖ The first year
- ❖ Hallucination, delusion, fluctuate delirium, aggressive behavior, change in mood or behavior or affect
- ❖ Etiology: SLE(90%), Steroid(10%)
- ❖ Anti-ribosomal, Anti-neural Ab
- ❖ Metabolic disorder, infection, schizophrenia

OPHTHALMOLOGIC

- ❖ The most common: kerato conjunctivitis sicca (kcs)
- ❖ Retinal abnormalities:
 - ★ Retinal hemorrhages
 - ★ Vasculitis-appearing lesions cotton wool spots
- ❖ Episcleritis & scleritis
- ❖ Uveitis is rare.
- ❖ Side effects of drugs (hydroxychloroquine and prednisolon)



GASTROINTESTINAL & HEPATIC

- ❖ Gastritis and peptic ulcer due to drugs
- ❖ Pancreatitis, peritonitis ,mesenteric vasculitis due to SLE
- ❖ Sign&symptoms:
 - Anorexia
 - Nausea& vomiting
 - Diarrhea
 - Abdominal pain
 - Ascites
- ❖ Liver involvement (lupoid hepatitis)

HEMATOLOGIC

- ❖ Anemia: * Chronic disease 37%
 - * Autoimmune hemolytic anemia 14%
 - * Iron deficiency 36%
 - * Microangiopathic hemolytic anemia(TTP-HUS)
- ❖ leukopenia: * 50%
 - * Lymphopenia: in active SLE 75%
 - * Side effect of glucocorticoids or other immunosuppressive agents.

HEMATOLOGIC

- ❖ Neutropenia: * Immune-mediated destruction
 - * Bone marrow suppression
- ❖ Thrombocytopenia:
 - * ITP
 - * TTP
 - * Splenomegaly
 - * Antiphospholipid antibody

LYMPHADENOPATHY AND SPLENOMEGALY

❖ LYMPHADENOPATHY:

- * 50%

- * Associated with active SLE

❖ SPLENOMEGALY:

- * 10-46%

- * Associated with active SLE

- * Usually with hepatomegaly

- * Isolated splenic infarction

