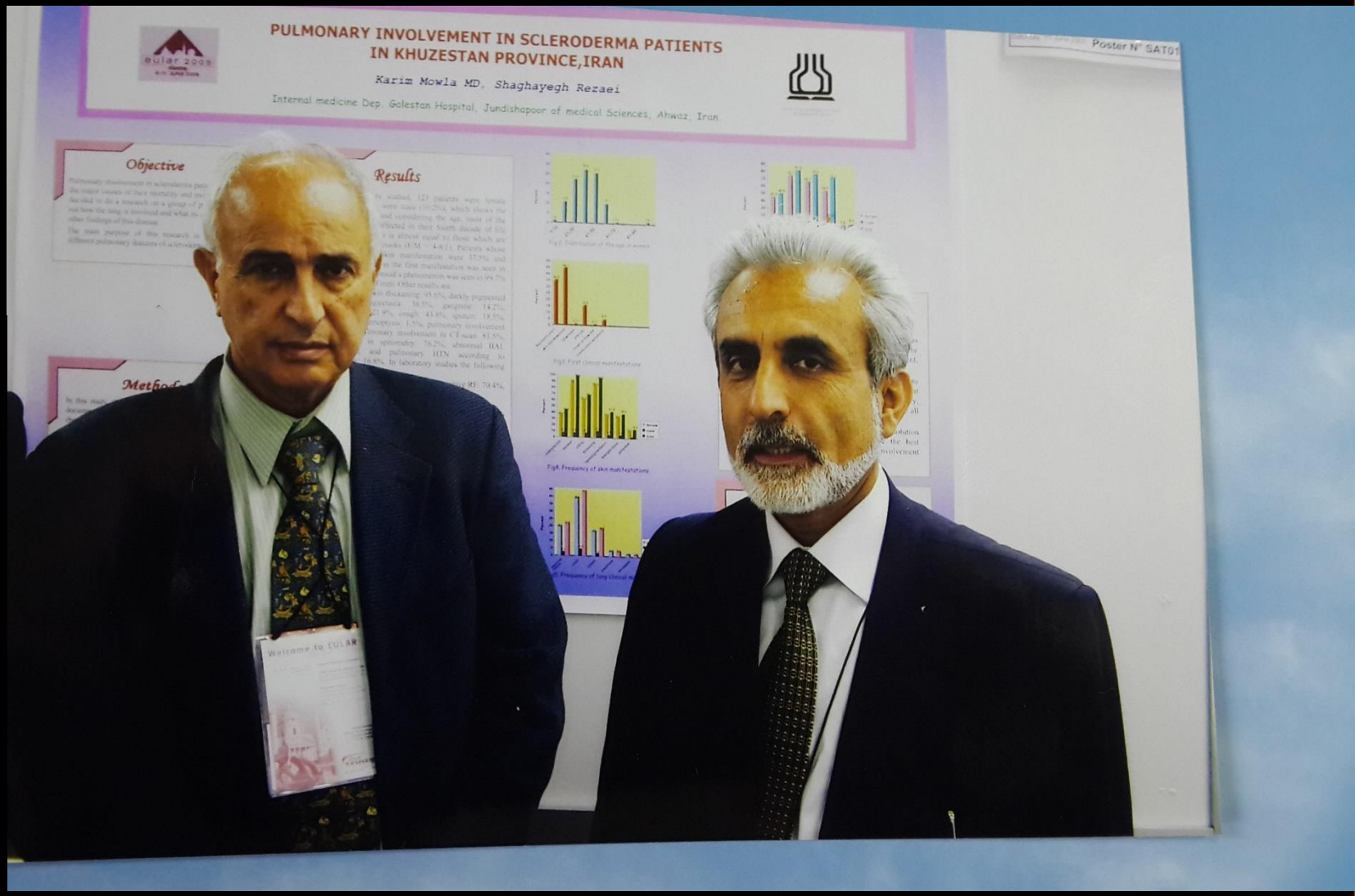

بسم الله الرحمن الرحیم

درود به روان استاد عزیز




May is
LUPUS
Awareness
Month

World Lupus Day™

May 10

The Fifth Annual Seminar of Rheumatology
Memorial of
Professor Mohammad Shafizadeh

How to Predict Flare of Lupus

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Importance of SLE Flare

The rate and the severity of flares during a year-long course of treatment are important predictors of disease outcome.

Flare in SLE definition

- **Chronic, Incurable**
- **Autoimmune**
- **Inflammatory**
- **Multi-system → Mild to Life threatening condition**
- **Lupus is a disease of Unpredictable Flares and Remission**

Lupus Flare

Unpredictable

Why Lupus Flare is Unpredictable

- **Heterogeneous**
- **Multisystem**
- **Overlap with other CTDs**
- **APS**
- **Comorbidity** **(Metabolic, CAD, Musculoskeletal, Hematologic, Malignancies, Fracture, GI, Lung, Eye, Skin, CNS, Congenital)**
- **Infections**
- **Malignancy**
- **.....**

Why Lupus Flare is Unpredictable

Flare # Activity

Why Lupus Flare is Unpredictable

- **Chronic Active (CA) → The most frequent (50%) → Moderately Severe**

- **Relapsing-Remitting (RR) → Intermediate frequency (30%) → Mild**

- **Long quiescence (LQ) → The least common (20%)**

Lupus Flare Definition

History of Lupus Flare Definition

- **Previous** definitions for Lupus flare
- Several **instruments** have been developed.

Instruments for measurement of acute Lupus activity

- **These have included the:**
  - **SLEDAI 2000**
  - **SLAMR: The Revised Systemic Lupus Activity Measures**
  - **ECLAM: The European Consensus Lupus Activity Measurement**
  - **BILAG: The British Isles Lupus Assessment Group**
  - **CLASI: The Cutaneous Lupus Disease Area and Severity Index**

History of Lupus Flare Definition

- In 2006, the LFA convened an International Consensus Panel to evaluate unmet needs in defining and measuring lupus flares.

SLE Flare definition by Lupus Foundation of America (LFA)

- **Clinical aspect:**
  - Measurable increase in disease activity
  - One or more organ
  - New or worse clinical sign and symptoms
- **Laboratory** measurements
- **Clinically significant** by assessor
- **Change or increase** Treatment

Ruperto N, Hanrahan LM, Alarcon GS, et al. International consensus for a definition of disease flare in lupus. *Lupus*. 2011;20(5):453-462.

Level of physicians agreement on SLE Flare definition

- **The definition had a high (61%) level of agreement across an international group of physicians.**

Ruperto N, Hanrahan LM, Alarcon GS, et al. International consensus for a definition of disease flare in lupus. *Lupus*. 2011;20(5):453-462.

How to Predict Lupus Flare

Warning Signs of a Flare

New manifestations

- **Development of symptoms you haven't had before.**
- **Any new or unexplained symptoms**

Warning Signs of a Flare

- Increased Fatigue
- A new or higher Fever
- Increased Pain
- Hair loss
- The development of Rash
- Abdominal problems
- Headache
- Dizziness, CNS and PNS
- Development of symptoms you haven't had before.

How to Predict Lupus Flare

- **History, History, History**
- **Physical Examination**
- **Paraclinic manifestations**

Etiology of Flare

- **Drugs** Discontinuation or starting some drugs
- **Infection**
- **Stress** (emotional or physical)
- **Sun** Exposure or other **ultraviolet** light
- **Overwork** or not enough rest
- **Pregnancy** or the **postpartum** period
- Unknown , Immunization

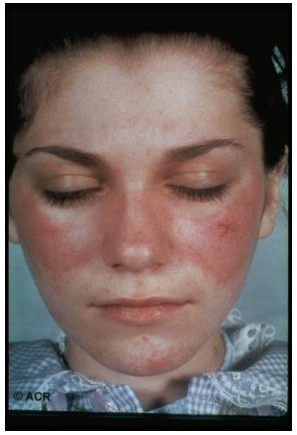
Differential diagnosis

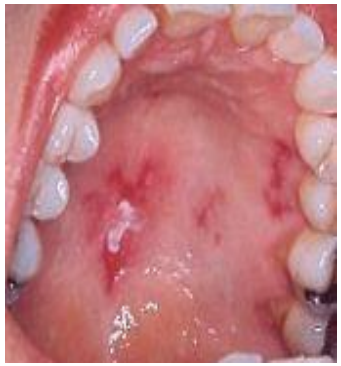
- **SLE**
- **Other CTDs**
- **APS**
- **Comorbidity** **(Metabolic, CAD, Musculoskeletal, Hematologic, Malignancies, Fracture, GI, Lung, Eye, Skin, CNS, Congenital)**
- **Infections**
- **Malignancy**
- **Other internal medicine diseases**

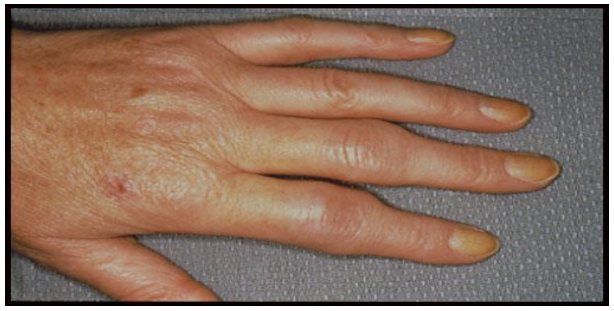
Minor

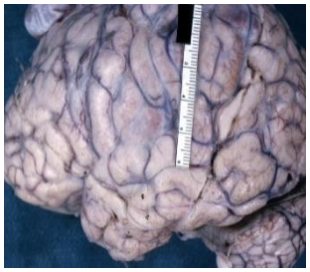
Major

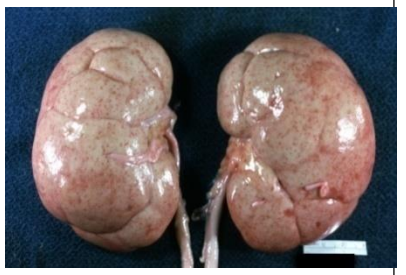
Fatigue, Malaise, Fever, Anorexia,
Skin, Musculoskeletal
Hematologic (mild)

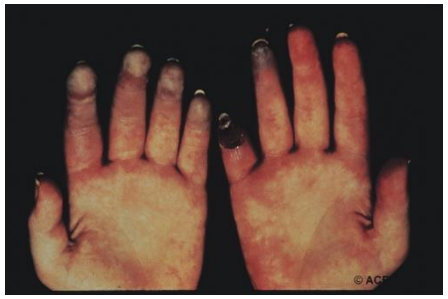


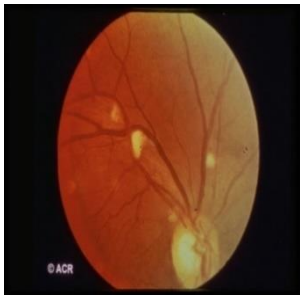


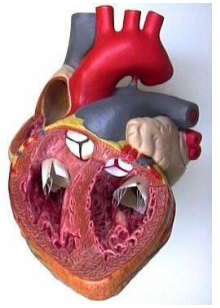




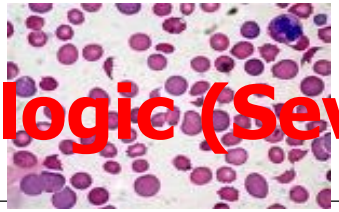








Hematologic (Severe)



Name and Family: Rheumatry-ID: Date:

SLE Follow-Up Form

MUCOCUTANEOUS	PULMONARY	RENAL
(2) ACUTE	• Lupus Pneumonitis	• Edema
• Malar rash	• Alveolar Hemorrhage	• Class I
• Bullous lupus	• ARDS	• Class II
• Toxic epidermal necrolysis	• ILD	• Class III (A/A-C/C)
• Maculopapular lupus rash	• Pulmonary HTN	• Class IV (S-A/G-A/S-C/G-C)
• Photosensitive lupus rash	• PTE	• Class V
SUB ACUTE	• OTHERS	• Class VI
• SCLE- Psoriasiform	CNS	• Activity score (1-24)
• SCLE- Annular polycyclic	• (8) Seizures	• Chronicity score (1-12)
CHRONIC	• (8) Psychosis	IMMUNOLOGIC
• DLE	• Mononeuritis Multiplex	• (2) Anti-dsDNA
• Lupus panniculitis	• Transvers Myelitis	• (2) Low complement (C3, C4, CH50)
• LE tumidus	• (8) Cranial neuropathy	
• Chilblains lupus	• (8) Acute Confusional state	WBC
• Discoid lupus/lichen planus overlap	• Aseptic Meningitis	Hb
LIVEDORETICULARIS	• Autonomic disorders	PLT
URTICARIAL VASCULITIS	• Chorea	ESR
(2) ORAL ULCER	• Guillain-Barre	CRP
NASAL ULCER	• (8) Lupus headache	Cr
(2) ALOPECIA (nonscarring)	• Myastenia Gravis	Anti-DNA
(8) VASCULITIS	• Optic Neuritis	C3
• Gangrene	• Psudotumor Cerebri	C4
• Palpable Purpura	• Vasculitis	CH50
• Periungual Erythema	• (8) CVA	aCL
• Splinter Hemorrhage	• OTHERS	LA
• Lupus Hand	CARDIOVASCULAR	Anti-RO
• OTHERS	• Tamponed	(4) Urinary Cast (RBC- granular)
HEMATOLOGIC	• Myocarditis	(4) Hematuria (>5 RBC)
• Hemolytic Anemia	• Libman Sack	(4) Pyuria (>5 WBC)
• (2) Leukopenia (<4000/mm3)	• Valvular Abnormality	(4) Proteinuria (>500 mg/d)
• Lymphopenia (<1000/mm3)	• Pericardial effusion	24 h- protein:
• (2) Thrombocytopenia	VASCULAR	
• TTP	• DVT	**Vital sign-** BP: PR:
• OTHERS	• Bud-Chiari syndrome	**Height**
MUSCULOSKELETAL	• Portal vein thrombosis	**Weight**
• (4) Arthritis	• Renal vein thrombosis	
• Inflammatory arthalgia	• Cerebral sinus thrombosis	PHYSICIAN GLOBAL ASSESSMENT (0-4)
• Articular erosion	• Jugular vein thrombosis	• Normal (0)
• Jacoud arthropathy	• IVC thrombosis	• Just Serology (1)
• (4) Myositis	• SVC thrombosis	• Mild (2)
• OTHERS	• Arterial thrombosis	• Moderate (3)
SEROSITIS	• OTHERS	• Severe (4)
• (2) Pleuritis	GASTROINTESTINAL	
• (2) Pericarditis	• Peritonitis	CONCLUSION
RETICULOENDOTHELIAL	• Pancreatitis	
• Lymphadenopathy	• Autoimmune Hepatitis	
• Splenomegaly	• PSC	
• Hepatomegaly	• Vasculitis	
CONSTITUTIONAL	• OTHERS	
• Anorexia	EYE	
• (2) Fever (>38)	• Retinal vasculitis	
• Weight loss (10% in 6 months)	• Retinal Hemorrhage	
• Fatigue	• Retinal Cytoid Body	
• OTHERS	• (8) Visual Disturbance	Signature:

PATIENT GLOBAL ASSESSMENT:

PRESENT DRUGS:

Warning Signs of a Flare
Constitutional symptoms

- Increased Fatigue, or extreme Exhaustion
- A new or higher Fever
- Loss of appetite
- Weight loss

Warning Signs of a Flare

Mucocutaneous symptoms

- **Hair loss**
- **The development of Rash**
- **Skin Vasculitis**
- **Oral lesions**

Warning Signs of a Flare

Musculoskeletal manifestations

- Increased Pain
- Joints pain and swelling
- Muscle pain and weakness
- Bone pain

Warning Signs of a Flare

Gastrointestinal manifestations

- **Abdominal discomfort or digestive problems**
- **Upset stomach**
- **Abdominal pain**
- **Liver and Pancreatic sign and symptoms**

Warning Signs of a Flare

CNS and PNS manifestations

- **Head ache**
- **Dizziness of Forgetfulness**
- **Seizure**
- **Focal sign (CNS & PNS)**

Warning Signs of a Flare

Hematologic manifestations

- **Pancytopenia**
- **Thrombocytopenia**
- **Anemia**

Warning Signs of a Flare

Immunologic manifestations

- **Anti ds DNA**
- **C3, C4, CH50, C1q**
- **Other Antibodies**

Warning Signs of a Flare

Acute phase reactant

- **ESR**
- **CRP**
- **Leukopenia**

Physician Global Assessment

- **Normal**
- **Just Serology (SACQ)**
- **Mild Flare**
- **Moderate Flare**
- **Severe Flare**

Mild or Moderate Flare [1]

- Change in SLEDAI > 3 points
- ≥ 1 Increase PGA, but not to more than 2.5

Mild or Moderate Flare 2

- New/worse Minor Organ involvement:
- Skin manifestations:
  - Discoid Lupus, Photosensitivity, Profundus, Cutaneous Vasculitis, Bullus Lupus, Nasopharyngeal Ulcers,
- Serositis: Pleuritis, Pericarditis,
- Arthritis
- Fever (SLE)

Mild or Moderate Flare 3

- **Increase in Prednisolone, but not to > 0.5 mg/kg/day**
- **Added NSAIDs or Hydroxychloroquine**

Severe Flare 1

- Change in SLEDAI > 12 points
- Increase in PGA to more than 2.5

Severe Flare 2

- New/worse Major Organ involvement:
- CNS Lupus manifestations:
- Nephritis
- Vasculitis
- Myositis
- Thrombocytopenia < 60,000
- Hemolytic Anemia: Hb < 7
- Anemia: Hb < 3

Severe Flare 3

- **Hospitalization**
- **Increase in Prednisolone:**
  - **> 0.5 mg/kg/day , and/or Pulse Methyl Pred**
- **New Cytotoxics:**
  - **Cyclophosphamide**
  - **Mycophenolate**
  - **Azathioprine**
  - **Methotrexate**
- **Plasmapheresis and/or IVIg**

Mild, Moderate or Severe Flare

severe and non-severe flares is clinically more important than to distinguish between mild and moderate.

Conclusion 1

- Lupus is a disease of unpredictable Flares and Remission
- In Flare → New or unexplained symptoms and signs → increased disease activity.
- To Predict the SLE Flare → History, Physical Examinations and Paraclinical measurements.

Conclusion 2

- The most important symptoms and signs:
  - Severe Fatigue, Weight loss, Fever
  - Mucocutaneous manifestations
  - Shortness of breath, Chest pain,
  - Abdominal pain,
  - Arthralgia, Myalgia,
  - severe anemia, Cytopenia, and
  - Others … since 7 to 30 days ago.

Conclusion 3

- **Evaluate the patient → Points →**
  - **Normal,**
  - **Just serology, (SACQ)**
  - **Mild to Moderate,**
  - **Severe.**
- **Reevaluate the patient Treatment.**

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