

# **Ocular Manifestations in Egyptian SLE patients**

Thesis

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Presented by

**Mostafa Adel Mohammed Mohammed**  
*M.B.B.CH*

Under supervision of

**Prof.Dahlia Abdel Mohsen Hussein**

*Professor of Internal Medicine and Rheumatology  
Ain-Shams University*

**Prof.Samah Abdel Rahman Elbakry**

*Professor of Internal Medicine and Rheumatology  
Ain-Shams University*

**Dr. Nashwa Aly Morshedy**

*Lecturer of Internal Medicine and Rheumatology  
Ain-Shams University*

Faculty of Medicine  
Ain-Shams University

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# List of Contents

Title	Page No.
List of Tables.....	i
List of Figures.....	iv
List of Abbreviations.....	vi
Abstract .....	ix
Introduction.....	1
Aim of the Study.....	xi
Review of Literature	
▪ Systemic Lupus Erythematosus.....	1
▪ Ocular Manifestations in Systemic Lupus Erythematosus .....	36
Subjects and Methods.....	51
Results.....	54
Discussion.....	80
Summary .....	87
Conclusion .....	89
Recommendation .....	90
References.....	91
Arabic Summary	

# List of Tables

Table No.	Title	Page No.
<b>Table (1):</b>	SLEDAI score.....	14
<b>Table (2):</b>	Recent ACR Classification Criteria of systemic lupus.....	17
<b>Table (3):</b>	2015 ACR/SLICC revised criteria for diagnosis of SLE .....	19
<b>Table (4):</b>	Autoantibodies of systemic lupus erythematosus.....	24
<b>Table (5):</b>	Comparison between SLE patients and controls regarding their demographic data.....	54
<b>Table (6):</b>	Clinical manifestations among SLE patients .....	55
<b>Table (7):</b>	Disease Duration (D.D.) and Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) among SLE Patients. ....	56
<b>Table (8):</b>	Different laboratory results among SLE Patients.....	57
<b>Table (9):</b>	ANA and DNA titre among Patients.....	58
<b>Table (10):</b>	Urine analysis among Patients.....	58
<b>Table (11):</b>	Comparison between SLE patients and normal controls regarding Ocular symptoms.....	60
<b>Table (12):</b>	Comparison between SLE patients and normal controls regarding Ocular Examination.....	62
<b>Table (13):</b>	Retinal abnormal findings among SLE patients. ....	64
<b>Table (14):</b>	Comparison between SLE patients with Ocular Affection and SLE patients with no Ocular Affection regarding demographic data.....	65

## List of Tables cont...

Table No.	Title	Page No.
<b>Table (15):</b>	Comparison between SLE patients with Ocular Affection and SLE patients with no Ocular Affection regarding disease duration and SLEDAI score.....	66
<b>Table (16):</b>	Comparison between SLE patients with Ocular Affection and SLE patients with no Ocular Affection regarding SLE symptoms.....	67
<b>Table (17):</b>	Comparison between SLE patients with Ocular Affection and SLE patients with no Ocular Affection regarding laboratory results.....	68
<b>Table (18):</b>	Comparison between SLE patients with Ocular Affection and SLE patients with no Ocular Affection regarding urine analysis parameters.....	71
<b>Table (19):</b>	Comparison between SLE patients with dry eye and those without dry eye regarding demographic data.....	72
<b>Table (20):</b>	Comparison between SLE patients with dry eye and those without dry eye regarding disease duration and SLEDAI score.....	73
<b>Table (21):</b>	Comparison between SLE patients with retinal affection and those without retinal affection regarding demographic data. ....	73
<b>Table (22):</b>	Comparison between SLE patients with retinal affection and those without retinal affection regarding disease duration and SLEDAI score.....	74
<b>Table (23):</b>	Comparison between SLE patients with retinal affection and those without retinal affection regarding anticardiolipin antibodies and lupus anticoagulant. ....	75

## List of Tables cont...

Table No.	Title	Page No.
<b>Table (24):</b>	Comparison between SLE patients with reduced visual acuity and those without reduced visual acuity regarding demographic data. ....	77
<b>Table (25):</b>	Comparison between SLE patients with reduced visual acuity and those without reduced visual acuity regarding disease duration and SLEDAI score.....	77
<b>Table (26):</b>	Ocular symptoms in SLE patient groups classified according to SLEDAI score.....	78
<b>Table (27):</b>	Ocular examination in SLE patient groups classified according to SLEDAI score.....	79

# List of Figures

Fig. No.	Title	Page No.
<b>Figure (1):</b>	Classification of SLE patients according to SLEDAI score.....	59
<b>Figure (2):</b>	Comparison between SLE patients and normal controls regarding Ocular Affection.....	61
<b>Figure (3):</b>	Comparison between SLE patients and normal controls regarding dry eye .....	61
<b>Figure (4):</b>	Comparison between SLE patients and normal controls regarding retinal affection. ....	63
<b>Figure (5):</b>	Comparison between the 2 groups regarding disease duration and SLEDAI score.....	66
<b>Figure (6):</b>	Comparison between the 2 groups regarding serum creatinine.....	69
<b>Figure (7):</b>	Comparison between the 2 groups regarding C3 .....	69
<b>Figure (8):</b>	Comparison between the 2 groups regarding Anticardiolipin IgM.....	70
<b>Figure (9):</b>	Comparison between the 2 groups regarding Lupus Anticoagulant.....	70
<b>Figure (10):</b>	Comparison between the 2 groups regarding urinary casts.....	72
<b>Figure (11):</b>	Comparison between SLE patients with retinal affection and those without retinal affection regarding SLEDAI score.....	74
<b>Figure (12):</b>	Comparison between SLE patients with retinal affection and those without retinal affection regarding anticardiolipin antibodies.....	76

## List of Figures cont...

Fig. No.	Title	Page No.
<b>Figure (13):</b>	Comparison between SLE patients with retinal affection and those without retinal affection regarding Lupus anticoagulant .....	76

# List of Abbreviations

Abb.	Full term
ACL .....	Anticardiolipin
ACR.....	American College of Rheumatology
ANA.....	Anti-Nuclear Antibody
anti-dsDNA.....	anti-double stranded Deoxyribonucleic acid
APC .....	antigen presenting cell
APLs.....	Anti-Phospholipid antibodies
aPTT .....	activated partial thromboplastin time
AZA .....	Azathioprine
BAL .....	Bronchoalveolar lavage
BILAG.....	British Isles Lupus Activity Group
BlyS.....	B-lymphocyte stimulator
BUN .....	Blood Urea Nitrogen
C3 .....	Complement 3
C4 .....	Complement 4
CBC .....	Complete blood count
CD .....	Cluster for Differentiation
CH50 .....	50% hemolytic complement
CNS .....	Central Nervous System
CPK.....	Creatine phospho-kinase
CRP .....	C-reactive Protein
CSA .....	Cyclosporin A
CT.....	Computed tomography
CTLA4.....	Cytotoxic T-lymphocyte-associated antigen 4
CVA.....	Cerebrovascular Accident
CVD.....	Cardiovascular disease
CYC .....	Cyclophosphamide
DLE .....	Discoid lupus erythematosus
DMARDs.....	Disease-modifying antirheumatic drugs
DRVVT.....	Dilute Russell viper venom time
ECLAM .....	European Consensus Lupus Activity Measurement
ELISA .....	Enzyme-linked immunosorbent assay
ESR .....	Erythrocyte sedimentation rate



## List of Abbreviations cont...

Abb.	Full term
FDA.....	Food and Drug Administration
FFA .....	fluorescein angiography
G6PD.....	Glucose-6-phosphate dehydrogenase
HCQ .....	Hydroxychloroquine
HGB .....	Hemoglobin
hLL2.....	humanized LL2
ICG.....	Indocyanine green
IgG .....	Immunoglobulin G
IgM.....	Immunoglobulin M
IL.....	Interleukin
ILD.....	Interstitial lung disease
INF- $\gamma$ .....	Interferon gamma
IOP .....	Intra-ocular pressure
IVIg .....	Intravenous Immunoglobulin
LAC .....	Lupus anticoagulant
LAI .....	Lupus Activity Index
LDH .....	lactate dehydrogenase
MAC.....	Membrane attack complex
MMF.....	Mycophenolate mofetil
MPA .....	mycophenolic acid
MRI.....	Magnetic Resonance Imaging
NMDA.....	N-methyl-D-aspartate
NMOSDs.....	Neuromyelitis optica spectrum disorders
NSAIDs .....	Non-steroidal anti-inflammatory drugs
OCT.....	optical coherent tomography
ONTT .....	Optic Neuritis Treatment Trial
P/C.....	Protein/creatinine
PH.....	Pulmonary hypertension
PIP .....	Proximal interphalangeal
PLT .....	Platelets
RBCs .....	Red Blood Cells
RNA.....	Ribonucleic acid
RNP.....	Ribonucleoprotein

## List of Abbreviations cont...

Abb.	Full term
RPR .....	Rapid plasma reagin
SCLE .....	Subacute cutaneous Lupus erythematosus
SLAM .....	Systemic Lupus Activity Measure
SLE .....	Systemic lupus erythematosus
SLEDAI.....	Systemic Lupus Erythematosus Disease Activity Index
SLICC .....	Systemic Lupus International Collaborating Clinics
SPSS .....	Statistical Package for Social Science
SS .....	Sjogren Syndrome
TNF .....	Tumor Necrosis Factor
WBC .....	White Blood Cell
Wt.....	Weight

## **Abstract**

Cotton-wool spots were the most common retinal abnormal finding followed by other rare forms of retinopathy as vasculitis, attenuated blood vessels, papilledema and pale optic disc.

SLE patients with ocular affection especially retinopathy had significantly higher levels of Anticardiolipin antibodies, Lupus Anticoagulant and also disease duration and SLEDAI score when compared to those patients without ocular affection.

**Key words:** Ocular Manifestations - Food and Drug Administration-Interleukin – Hydroxychloroquine - Mycophenolate mofetil - Optical coherent tomography

## INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic, multisystem, autoimmune disease that can affect any part of the human body. Ocular manifestations in SLE are fairly common and should be taken seriously as they may lead to significant morbidity and visual deterioration. In addition to that, ocular manifestations could represent the disease activity of the lupus (especially in the presence of retinal signs) and serve as an indicator of adequacy of treatment in control of the disease (*Arevalo et al., 2002*).

SLE can affect the periorbital, ocular adnexa, eye, and optic nerve. The most common association is keratoconjunctivitis sicca, while the most visually devastating sequelae occur secondary to optic nerve involvement and retinal vaso-occlusion (*Neal et al., 2012*). Orbital involvement is a rare manifestation of SLE. Vasculitis, myositis, and panniculitis have all been described. Signs and symptoms include proptosis, enophthalmos, orbital pain, blurred vision, chemosis, and restriction of extraocular movements (*Stavrou et al., 2002*). Periorbital edema is an uncommon manifestation of systemic and discoid lupus erythematosus with an overall incidence of 4.8% (*Tuffanelli et al., 2002*). Typical lesions of discoid lupus erythematosus are slightly raised, scaly, and atrophic rarely affecting the eyelids. Episcleritis is generally a benign inflammation of the episclera and typically occurring in

young females with incidence 2.4 % in SLE patients (*Sitaula et al., 2011*). Corneal epitheliopathy, scarring, ulceration and filamentary keratitis can all occur secondary to keratoconjunctivitis sicca. More rare corneal complications include peripheral ulcerative keratitis (*Messmer et al., 1999*), which can be marker of active systemic vasculitis, interstitial keratitis, and keratoendothelitis (*Varga et al., 1993*).

Lupus retinopathy is one of the most common vision-threatening complications of systemic lupus erythematosus with an incidence of up to 29% in patients with active systemic disease. A strong correlation exists between presence of retinopathy and CNS disease (*Stafford-Brady et al., 1988*). The most common pattern of retinopathy is microangiopathy similar to diabetic and hypertensive retinopathy. The earliest findings are small intraretinal hemorrhages and cotton wool spots (*Ushiyama et al., 2000*). Retinal vasculitis, a subset of retinal vasculopathy featuring inflammation of the retinal arterioles or venules, tends to have poorer visual outcomes and present in an acute onset fashion. A large percentage of these patients have concomitant antiphospholipid antibodies including anticardiolipin and lupus anticoagulant (*Montehermoso et al., 1999*).

## **AIM OF THE STUDY**

**I**t is to study Ocular manifestations in Egyptian SLE patients and its correlation with disease activity.

## Chapter One

# SYSTEMIC LUPUS ERYTHEMATOSUS

### Introduction:

Systemic lupus erythematosus (SLE) is a chronic autoimmune connective tissue disorder affecting multiple organ systems often with a relapsing and remitting clinical course. It can affect the skin, joints, kidneys, Eye, brain and other organs. The underlying cause of autoimmune diseases is not fully known (*Palejwala et al., 2012*).

SLE is much more common in women than men. It may occur at any age but appears most often in people between the ages of 10 and 50. African Americans and Asians are affected more often than people from other races (*Ruiz-Irastorza et al., 2010*).

Its pathogenesis is multifactorial mainly genetic and environmental in which it occurs to genetically predisposed individuals resulting in irreversible loss of immunologic self-tolerance (*Ruiz-Irastorza et al., 2010*).

The reported prevalence SLE in the population is 20 to 150 cases per 100,000. In women, prevalence rates vary from 164 (white) to 406 (African American) per 100,000 (*Pons et al., 2010*).

Due to improved detection of mild disease, the incidence nearly tripled in the last 40 years of the 20th century. Estimated