Comparison of Disease Characteristics, Course and Outcome between Adults with Juvenile and Adult- onset Systemic Lupus Erythematosus

A Thesis

Submitted for Partial Fulfillment of

M.D Degree In Internal Medicine

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(2018)

بسم الله الرحمن الرحيم

Acknowledgement

In the name of Allah, the Most Gracious and the Most Merciful

Alhamdulillah, all praises to Allah for the strengths and His blessing in completing this thesis.

Special appreciation goes to my supervisor, **Prof. Dr. Abd El-Azeim Mohammad ELhefny,** *for his supervision and constant support. His invaluable help of constructive comments and suggestions throughout the experiment and thesis works have contributed to the success of this work.*

I am also deeply indebted to **Prof. Dr. Reem Abd El-Monem** *for her great support throughout the whole work.*

I would like to express my appreciation to **Prof. Dr. Maryam Abd El-Rhaman** *-for the tremendous effort she has done in meticulous of revision of this work.*

My profound thanks for **Dr. Nashwa Aly Morshedy** *for her useful advises and continuous help in achievement of this thesis.*

Many thanks for **Dr. Amro Abdelzaher** *for his meticulous assistance and guidance through out the study especially in the statistical and puplication work.*

Last but not least, my deepest gratitude goes to my parents and also to my wife for their endless love, prayers and encouragement.

Mohammed Al Azab

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List of Abbreviations

Abb.	Meaning
ACLE	Acute cutaneous lupus erythematosus
ACR	American College of Rheumatology
AIHA	Autoimmune hemolytic anemia
ANA	Antinuclear antibody
Anti-dsDNA	Antinuclear double stranded DN
APL	Antiphospholipid antibodies
APO	Adverse pregnancy outcomes
APS	Antiphospholipid syndrome
ARDS	Acute respiratory distress syndrome
ASLE	Adult-onset SLE
AZA	Azathioprine
BAFF	B-cell activating factor
BILAG	British Isles Lupus Assessment Group
CHB	Congenital heart block
CHD	Coronary heart disease
CNS	Central nervous system
CSs	Corticosteroids
CVST	Central venous thrombosis
DHEA	Dehydroepiandrosterone
DM	Diabetes mellitus
ECG	Electrocardiography
ESRD	End stage renal disease
EULAR	European League Against Rheumatism
GI	Gastrointestinal
HAQ	Heath Assessment Questionnaire

List of Abbreviations

Abb.	Meaning
IA	Intra-articular
IFN-alpha	Interferon-alpha
ILD	Interstitial lung disease
ITP	Idiopathic thrombocytopenic purpura
IUGR	Intra-uterine growth restriction
IVIG	Intravenous immunoglobulin
JSLE	Juvenile onset systemic lupus erythematosus
LAC	Lupus anticoagulant
LE	Lupus erythematosus
LN	Lupus nephritis
LVEF	Left ventricular ejection fraction
MTX	Methotrexate
MMF	Mycophenolate Mofetil
NPSLE	Neuropsychiatric lupus
NSAIDs	Nonsteroidal anti-inflammatory drugs
PAH	Pulmonary arterial hypertension
PDCs	Plasmacytoid dendritic cells
PML	Progressive multifocal leukoencephalopathy
RA	Rheumatoid arthritis
RNP	Ribonucleoprotein
SAH	Subarachnoid hemorrhage
SF36	Short Form 36
SLAM	Systemic Lupus Activity Measure
SLE	Systemic lupus erythematous
SLEDAI	Systemic Lupus Erythematosus Disease Activity Index
SLICC	Systemic Lupus International Collaboration Clinics
TPMT	Thiopurine S-methyltransferase
TTP	Thrombocytopenic purpura

Abstract

Background:

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with a broad spectrum of clinical manifestations that predominantly affects young women during childbearing age. The disease onset of SLE can also occur in pediatric and older populations.

Patients and Method:

One hundred and fifty consecutive patients with SLE fulfilling the SLICC criteria for the classification of SLE (Petri et al., 2012) were included in the study. The patients were classified according to the age at disease onset.

Results:

This study was conducted on One hundred and fifty patients with SLE fulfilling the SLICC criteria for the classification of SLE (Petri et al., 2012). SLE patients were 141 females (94.00%) and 9 males (6.00%).

Conclusion:

- The age at onset of SLE influences its clinical and serological manifestations.
- JSLE patients have a more aggressive presentation and course than ASLE patients with a higher frequency of lupus nephritis, more cutaneous symptoms and hematological manifestations.
- JSLE patients have more severe disease activity with very high SLEDAI and SLICC damage scores.
- ASLE patients have more musculoskeletal manifestations

Keywords: Systemic Lupus International Collaboration, Thiopurine S-methyltransferase, Thrombocytopenic purpura

INTRODUCTION

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with a broad spectrum of clinical manifestations that predominantly affects young women during childbearing age ***(Cervera et al., 2009).*** The disease onset of SLE can also occur in pediatric and older populations ***(Mak et al., 2007)***.

Although the highest prevalence of SLE is among women of childbearing age, juvenile-onset SLE (JSLE) represents 10–20% of all SLE cases ***(Kamphuis and Silverman E, 2010)***. SLE is diagnosed early in life (before age 16) in up to 20% of cases ***(Hersh et al., 2011)***.

Several factors, including age and ethnicity, can affect the nature and severity of SLE, and response to treatment also varies by race, ethnicity, and age ***(Borchers et al., 2010)***. Childhood-onset lupus patients display some differences in their disease profile and are associated with higher disease severity, and a more-rapid damage accrual compared with adult-onset SLE (ASLE) ***(Kamphuis et al., 2010)***.

It has been established JSLE tends to have a more aggressive presentation and course, with high rates of organ involvement and increased need for long-term immunosuppressive medication ***(Beatriz et al., 2014)***. JSLE patients have more severe disease activity, and renal involvement was more frequent ***(Choi et al., 2015)***.

While in adults a predominance of clinical features such as serositis and sicca symptoms was observed, in adolescents

the disease was significantly more associated with the development of LN, which is considered an indicator of a worse prognosis. A notable percentage of adolescent-onset patients also developed cardiovascular events or cancer, which are uncommon diseases in young patients ***(Beatriz et al., 2014).***

There are notable differences among the manifestations of the disease between children and adults. A study comparing 56 children with JSLE and 194 patients with ASLE found that renal involvement, encephalopathy and hemolytic anemia, were significantly more common in JSLE as compared to ASLE ***(Hersh et al., 2009).*** There is a higher prevalence of anti-dsDNA, antiSm, and anti-nucleosome antibodies in JSLE patients ***(Choi et al., 2015).***

Despite a dramatic improvement in mortality in both adult and adolescent-onset disease in the last few decades, owing largely to significant advances in the therapy of SLE, patients diagnosed with SLE at an early age remain at high risk for early mortality in their young adult years ***(Hersh et al., 2011)***.

The poorer outcome observed in younger children may be explained by a stronger genetic predisposition, a more severe disease expression (e.g. more frequent neuropsychiatric disorders), a higher infectious susceptibility and a more aggressive therapy, particularly within the first 6 months of disease course ***(Elodie et al., 2009)***.