

Systemic lupus erythematosus in a male patient

H Sibarani^{1,2*} and Z Zubir^{1,2}

¹Division of Allergy and Immunology, Department of Internal Medicine, Faculty of Medicine, Universitas Sumatera Utara, Medan, Indonesia

²Haji Adam Malik General Hospital, Medan, Indonesia

*Corresponding author: henrysibarani81@gmail.com

Abstract. Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder with a broad spectrum of clinical presentations. Female to male ratio is approximately 9:1. A 20 years old male was admitted to HAM Hospital 3 months ago with chief complaint pain in both knees joint. After anamneses, physical examination and laboratory test the patient was diagnosed with systemic lupus erythematosus. The patient tested positive for ANA and anti-ds-DNA antibody test. The patient was with giving non-biologic DMARDS @myfortic 360mg, methylprednisolone, chloroquine and other symptomatic drugs.

1. Introduction

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder with a broad spectrum of clinical presentations.¹ Lupus can be a mild disease, a severe and life-threatening illness, or anything in between.²⁻⁵ Although the specific cause of SLE is unknown, there have been identifying multiple genetic predispositions and gene-environment interactions. There are genetic-susceptibility factors, environmental triggers, B-cell and T-cell interactions, antigen-antibody (Ab) responses, and immune clearance processes interact to generate and perpetuate autoimmunity in systemic lupus erythematosus (SLE). HLA = human leukocyte antigen; UV = ultraviolet light.⁶

Prevalence: 2–140/100,000 worldwide but as high as 207/100,000. Incidence: 1–10/100,000 worldwide. The population at highest risk: women in their reproductive years. Female to male ratio is approximately 9:1 post-puberty and premenopausal. Environmental and exposure-related causes of SLE are less clear. Possible early-life risk factors include the following: low birth weight (<2,500 g), preterm birth (≥ 1 month early), childhood exposure to agricultural pesticides.⁷⁻¹⁰

The pathogenesis of lupus remains unclear although the concept of apoptosis goes some way to explaining how the immune system may recognize predominantly intracellular antigens. Autoantigens are released by necrotic as well as apoptotic cells. Defects in the clearance of apoptotic cells have been described in SLE which may lead to aberrant uptake by macrophages which then present the previously intracellular antigens to T and B cells thus driving the autoimmune process.¹¹⁻¹⁵

Table 1. Classification criteria for SLE.

Classification criteria	Definition
Malar rash	flat or raised, fixed erythema, tending to spare the nasolabial folds, over the malar eminences
Discoid rash	Adherent keratotic scaling and follicular plugging is raised by erythematous; atrophic scarring may occur in



Photosensitivity	older lesions Skin rash is a result of unusual reaction to sunlight, by physician observation or patient history
Oral ulcer	nasopharyngeal or oral ulceration is not usually pain, observed by a physician
Arthritis	Non-erosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling or effusion
Serositis	Pleuritis: Convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion
a) <i>Pleuritis</i>	Pericarditis: documented by ECG or rub or evidence of pericardial effusion
b) <i>Pericarditis</i>	Proteinuria: more than 0.5 grams per day or greater than +++ if quantification not performed
Renal disorder	Casts: may be red cell, hemoglobin, granular, tubular or mixed
a) <i>Persistent proteinuria</i>	See ACR definitions of 19 separate syndromes
b) <i>Cellular casts</i>	With reticulocytosis
Neurologic disorder	Less than 4000/mm ³ total on 2 or more occasions
Hematologic disorder	Less than 1500/mm ³ total on 2 or more occasions
a) <i>Hemolytic anemia</i>	Less than 100,000/mm ³ in the absence of offending drugs
b) <i>Leukopenia</i>	Antibody to native DNA in abnormal titer
c) <i>Lymphopenia</i>	Presence of antibody to Sm nuclear antigen
d) <i>Thrombocytopenia</i>	1. Abnormal serum level of IgG or IgM antinuclear antibodies
Immunologic disorder	2. A positive test result for lupus anticoagulant using a standard method
a) Anti-DNA	3. A false positive serologic test for syphilis, known to be positive for at least 6 months and confirmed by <i>Treponema pallidum</i> immobilization or fluorescent treponema antibody absorption test
b) Anti-Sm	Abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay are at any point in time and there are no drugs known to associate with 'drug-induced lupus' syndrome
c) Positive finding of antiphospholipid antibodies	
Antinuclear antibody	

Table 2. Treatment of SLE.

Glucocorticoids/steroids: oral Prednisone 0.5-1 mg/kg/day for severe disease	Hydroxychloroquine: oral 200-400 mg/day
Methylprednisolone: iv. lupus nephritis 1000 mg iv. for 3 days	Mycophenolatemofetil: oral 2-3 g/day
Cyclophosphamide: iv. 7-25 mg/kg/month x6 or oral 1.5-3 mg/kg/day	Methotrexate: oral 10-25 mg once week
Azathioprine: oral 2-3 mg/kg/day	Topical glucocorticoids
	NSAIDs

2. Case Report

A male 20 years old, admitted to HAM Hospital 3 months ago with main complain pain in both knees joint. The patient stated pain, in the beginning, mild but in 1 month gradually increased. He could not walk along distance and easy to feel weak. He also felt redness on face following itch and sensitivity to sunlight on a half month ago. He was consulted to the general practitioner and was given analgetic to reduce the pain. After a month pain not decreased by consuming that tablet.

After anamneses, physical examination and laboratory test the patient was diagnosed with systemic lupus erythematosus. Laboratory test, Hb/WBC/Plt: 9.9 / 12080 / 281,000. Neutrophil / Lymphocyte /

Monocyte / Eosinophil / Basophil: 85/11 / 3.6 / 0.1 / 0.2%. Ureum / Creatinine: 21/ 0.5 mg/dl. Electrolyte Natrium / Kalium / Chloride: 140/2.5/107 mEq/L. The patient tested positive for ANA: 204 and anti-ds-DNA antibody test: 742. Radiology imaging showed the normal result. The patient was hospitalized and given non-biologic DMARDS @myfortic 360mg (Mycophenolatemofetil) 2 times a day, methylprednisolone 12mg three times a day, chloroquine and other symptomatic drugs. After three months of laboratory finding back to normal, treatment continued, and the dose is tapered off. The complaint knee joint pain almost disappears, anitch on the face decreased, and now the patient can start to do this activity.

3. Discussion

Based on criteria ARA patient had six sign and symptoms: malar rash, arthritis, photosensitivity, anemia, immunologic disorder, antinuclear antibody. Patient categorized as mild SLE because there is no sign life-threatening and vital sign is considerable good. Prevalence SLE population at highest risk: women in their reproductive years with a female: male ratio is approximately 9:1 in this case patient was male and had no familial autoimmune disease. The patient was a farmer and often contact with agricultural pesticides, worked under the sun and consumed cigarettes and instant noodle. Immunological test ANA: 204 references strong, anti-ds-DNA: 742 references moderate confirm as Systemic lupus erythematosus.

Although the specific cause of SLE is unknown, multiple factors associated with the development of the disease, including genetic, epigenetic, ethnic, immunoregulatory, hormonal, and environmental factors. Patients with SLE must know to prevent triggers for flare.

People with SLE must prevent the ultraviolet light and sun exposure for minimizing worse symptoms from photosensitivity. Diet modification must base on the disease activity. A balanced diet is an important thing, but patients with SLE and hyperlipidemia, for example, doing on a low-fat diet. there are many patients with SLE have low vitamin D because of less sun exposure

Prognosis of the disease course is milder and the survival rate is higher in persons with isolated skin and musculoskeletal involvement than in those with renal disease and CNS disease.^{18,19}

References

- [1] Tan E M, Cohen A S, Fries J F, *et al.* 2013 The 1982 revised criteria for the classification of systemic lupus erythematosus *Arthritis Rheum.* **25** 1271-7
- [2] Fessel W J 1974 Systemic lupus erythematosus in the community: incidence, prevalence, outcome and first symptoms; the high prevalence in black women *Arch. Intern. Med.* **134** 1027-35
- [4] Hochberg M C, Perlmuter S L, Medsger T A, *et al.* 1995 Prevalence of self-reported physician-diagnosed systemic lupus erythematosus in the USA *Lupus* **4** 454-6
- [5] Michet C J, McKenna C H, Elveback L R, *et al.* 1985 Epidemiology of systemic lupus erythematosus and other connective tissues disease in Rochester, Minnesota *Mayo Clin. Proc.* **60** 105-13
- [6] Parks C G, D' Aloisio A A and Sandler D P 2016 Early life factors associated with adult-onset systemic lupus erythematosus in women *Front. Immunol.* **7** 103
- [7] Jakes R W, Bae S C, Louthrenoo W, *et al.* 2012 Systematic review of the epidemiology of systemic lupus erythematosus in the Asia-Pacific region: prevalence, incidence, clinical features, and mortality *Arthritis Care Res.* **64(2)** 159-68
- [8] Ginzler E and Tayar J 2015 *American College of Rheumatology* Available from: http://www.rheumatology.org/practice/clinical/patients/diseases_and_conditions/lupus.pdf#search=sle [Accessed: 25 September 2015]
- [9] Parks C G, Cooper G S, Nylander-French L A, *et al.* 2002 Occupational exposure to crystalline silica and risk of systemic lupus erythematosus: a population-based, case-control study in the southeastern United States *Arthritis Rheum.* **46** 1840-50

- [10] Cooper G S, Parks C G, Treadwell E L, *et al.* 2004 Occupational risk factors for the development of systemic lupus erythematosus *J. Rheumatol.* **31** 1928-3
- [11] Munoz L E, Gaipf U S, Franz S, *et al.* 2015 SLE - a disease of clearance deficiency? *Rheumatology* **44** 1101-7
- [12] Arbuckle M R, Mc Clain M T, Rubertone M V, *et al.* 2003 Development of autoantibodies before the clinical onset of systemic lupus erythematosus *N. Engl. J. Med.* **349** 1526-33
- [13] Mc Clain M T, Arbuckle M R, Heinlen L D, *et al.* 2004 The prevalence, onset, and clinical significance of antiphospholipid antibodies prior to diagnosis of systemic lupus erythematosus *Arthritis Rheum.* **50** 1226-32
- [14] García-Carrasco M, Jiménez-Hernández M, Escárcega R O, *et al.* 2008 Use of rituximab in patients with systemic lupus erythematosus: an update *Autoimmun. Rev.*
- [15] Abelson A K, Delgado-Vega A M, Kozyrev S V, *et al.* 2008 STAT4 associates with SLE through two independent effects that correlate with gene expression and act additively with IRF5 to increase risk *Ann. Rheum. Dis.*
- [16] Zheng Z H, Gao C C, Wu Z Z, *et al.* 2016 High prevalence of hypovitaminosis D of patients with autoimmune rheumatic diseases in China *Am. J. Clin. Exp. Immunol.* **5(3)** 48-54
- [17] Ju C and Tacke F 2016 Hepatic macrophages in homeostasis and liver diseases: From pathogenesis to novel therapeutic strategies *Cell Mol. Immunol.* **13** 316-27
- [18] Cancro M P, D' Cruz D P and Khamashta M A 2009 The role of B lymphocyte stimulator (BLyS) in systemic lupus erythematosus *J. Clin. Invest.* **119(5)** 1066-73
- [19] Lo M S and Tsokos G C 2011 T-cells in systemic lupus erythematosus: progress toward targeted therapy *Rheumatologist*