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2015 ACR/SLICC Revised Criteria for Diagnosis of Systemic Lupus Erythematosus

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Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune systemic disorder with unknown etio-pathogenesis. Upon the susceptible genetic, hormonal and abnormal immunologic background, the environmental factors especially ultraviolet rays may play role as trigger to permit disease development [1].

Auto-antibodies especially Antinuclear Antibodies (ANA), anti-double stranded DNA (anti-dsDNA), anti-smith antibody (anti-Sm), anti-phospholipid antibodies (aPLs), antibodies against RBC, WBC, platelets, anti-neuronal antibodies and consumption of complements and production of Immune-complexes can contribute to creation of all clinical/laboratory manifestations of SLE [2, 3].

It occurs predominantly among women of childbearing ages and involves all organs in the body [4]. Malar rash, discoid rash (DLE), photosensitivity, alopecia, oral/nasal ulcers, polyarthralgia/myalgia, polyarthritis, pleurisy/pericarditis and peritoneitis, leukopenia, thrombocytopenia, hemolytic anemia, hematuria, proteinuria, azotemia, psychosis/seizures, peripheral/cranial neuropathies are the classic features of disease. Other organ involvements are including cardiovascular, pulmonary, ophthalmic, gastrointestinal, and so on [5, 6].

The diagnosis of SLE can be made by clinical/laboratory judgment of an expert rheumatologist and there is not any diagnostic criteria for early detecting it yet. The 1997 American College of Rheumatology (ACR) criteria [7] and its complementary criteria; the 2012 Systemic Lupus International Collaborating Clinics (SLICC) criteria [8], both are designed for classification of SLE and they are not diagnostic. The 2012 SLICC criteria are very complex/extended criteria and it can be used when the ACR criteria cannot classify SLE. Application of two separate criteria for classification of one disease is not a normal/natural way. So we need single criteria instead of them for SLE not only for classification but also for early diagnosis of it. About two years ago the corresponding author of this letter created the 2013 ACR revised criteria by Iran for diagnosis of SLE and he delivered it to his colleagues within the largest center of SLE in Iran. Despite the good cooperation of our colleagues in that center, the project of evaluation of that criteria was failed due to many problems including low financial facilities and some defects in the data of the profiles of patients with SLE eg the absence of Anti-Sm or Renal pathology in many cases and so on [9].Right now by this letter corresponding author deliver his newest criteria for diagnosis of SLE entitled “2015 ACR/SLICC revised criteria for diagnosis of SLE” that is presented in table A.

Table A: 2015 ACR/SLICC revised criteria for diagnosis of SLE [a b c]

Acute/subacute cutaneous lupus rash	Up to 2 points
• Malar rash	2.p
• Subacute cutaneous Lupus erythematosus (SCLE) rash • Palpable purpura or urticarial vasculitis	1.p 1.p
• Photosensitivity	1.p
Discoid lupus erythematosus (DLE) rash or hypertrophic Lupus rash	1.p
Non-scarring frank alopecia [d]	1.p
Oral/nasal ulcers	1.p
Joint disease	1.p
Pleurisy and/or pericarditis	1.p
Psychosis and/or seizure and/or acute confusion	1.p
Kidney involvement	Up to 2 points
• proteinuria≥ 3+ or ≥ 500 mg/day or urinary casts	1.p
• Biopsy-proven nephritis compatible with SLE	2.p
Hematologic	Up to 3 points
• WBC count < 4000/mm^3 or lymphocyte count < 1500/mm^3 on ≥ 2 occasions or WBC count < 4000/mm^3 along with lymphocyte count < 1500/mm^3 in one occasion	1.p
• Thrombocytopenia < 100,000/mm^3	1.p
• Hemolytic Anemia	1.p
Serologic tests	Up to 3 points
• Low titer positive ANA	1.p
• High titer FANA with homogenous or rim pattern	2.p
• Positive anti-ds DNA	2.p
• Positive anti-Sm	2.p
• Anti-phospholipid antibodies (aPLs)	1.p
• Low serum complement (C_3 and/or C_4 and/or CH_{50})	1.p

a: for each criteria: No other prominent disease or condition is likely to cause the presence of the criteria according to the patient's clinical and drug history or physical examination.

b: The definitions for malar rash, discoid rash, photosensitivity, oral ulcers, psychosis, seizure and urinary casts are the same as American College of Rheumatologycriteria for SLE and the definitions of nasal ulcers pleurisy/pericarditis and joint disease and acute confusion are the same as Systemic Lupus International Collaborating Clinics criteria for SLE. High titer serologic test means more than 3 times of upper limits of normal.

c: The patients with 4 points out of 16, have definite diagnosis of SLE. With 3 points highly suggestive SLE, with 2 points probable SLE and with one point possible SLE are the diagnosis.

d: Diffuse thinning or hair fragility with visible broken hairs with positive pulling test or apparent alopecia convincing the patient to ask for physician consultation. Not to mention that the related skin should not have any scar.

I think it is a good instrument for early detection of SLE with high sensitivity and specificity but we cannot evaluate it due to many problems mentioned above. However the author would like to ask the ACR and SLICC members and all of the other Rheumatologists in the world to evaluate the 2015 ACR/SLICC revised criteria, 1997 ACR criteria and 2012 SLICC criteria separately in the initial presentation of cases with SLE diagnosed by clinical/laboratory judgment. I will not be surprised if you ask me how I created these criteria. You should know that after many years of visiting the patients with SLE and studying the literatures and evaluating of the classification criteria of SLE in every each one of the patients who have had diagnosis of SLE upon clinical/laboratory judgment, this new criteria can easily be created. Indeed, all of the pitfalls/defects and outstanding items of the both classification criteria of SLE could be detected by corresponding author and after the combination of both criteria (ACR/SLICC), this new criteria could be delivered by giving the compatible points to each item. Here please let me show you some cases of SLE that I have seen in practice. For example in a 31 year-old woman with typical malar rash, frank alopecia and polyarthritis of both hands upon clinical judgment, the diagnosis is SLE, even if all of the biochemistry and serologic tests are normal. This case cannot fulfill anyone of the ACR and SLICC criteria but the 2015 ACR/SLICC revised criteria can be fulfilled by it. In an 18 year-old woman with psychosis and photosensitivity that has high titer of Anti-dsDNA in serologic tests, upon clinical/laboratory judgment the diagnosis is SLE. It cannot fulfill anyone of the ACR and SLICC criteria but the 2015 ACR/SLICC revised criteria can be fulfilled exactly by it. Do you agree with the diagnosis of SLE in a young man with leukopenia of 3200/mm3 and 22% of lymphocyte, thrombocytopenia of 76,000/mm3 along with hemoglobin of 9 gr/dl and reticulocyte count of 12% without any acute blood loss when he has pericarditis and past history of convulsion without any known cause? Upon clinical/laboratory judgment, he is a case of SLE but it cannot fulfill anyone of the ACR and SLICC criteria whereas the 2015 ACR/SLICC revised criteria can confirm it. In a 26 year-old woman with malar rash, discoid lupus rash and urinalysis containing 1+ blood, 1+ protein and 1-2 granular casts with renal pathology compatible to type II Lupus Nephritis and normal serologic tests, upon clinical/laboratory judgment, diagnosis of SLE is the best diagnosis. The 2015 ACR/SLICC revised criteria can establish this diagnosis but anyone of the ACR and SLICC criteria cannot confirm it. If you want, I can present many other cases of SLE that in practice I have seen them while anyone of the ACR and SLICC criteria cannot detect them. Finally, the corresponding author of this letter as the creator of Iran criteria for diagnosis of Ankylosing Spondylitis, Rheumatoid Arthritis and Granulomatosis with polyangiitis (Wegener's)[10, 11, 12] thinks that 2015 ACR/SLICC revised criteria is the best way to approach to the diagnosis of SLE.

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