

Dialogue: Cutaneous lupus erythematosus: a lone Wolf?

Andrew G Franks Jr,¹ Antonio Guilabert²

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¹Skin Lupus & Autoimmune Connective Tissue Disease Section, NYU School of Medicine, New York, New York, USA

²H de Granollers, Granollers, Spain

Correspondence to

Professor Andrew G Franks Jr; andrew.franks@nyumc.org

The expansion of genetic, genomic, environmental and metabolic knowledge that contributes to the immunological dysregulation in lupus is extremely challenging to those attempting to establish a 'gold standard' classification of systemic lupus erythematosus (SLE).¹ An approach that incorporates this information into its classification may help in attaining more uniform comparison of patient data across all populations. The Systemic Lupus International Collaborating Clinics (SLICC) represented an 8-year effort of review, consensus and statistical analyses which sought dermatological expert opinion and strived to improve on the revised American College of Rheumatology (ACR) classification.² These criteria included a more detailed classification of skin manifestations and attempted to address the issue of excessive weighting of skin criteria in the ACR SLE scheme. SLICC included additional components of cutaneous lupus that were absent in the ACR criteria such as alopecia and eliminated the somewhat redundant photosensitivity, but one can have discoid rash, malar rash, alopecia and a positive antinuclear antibody (ANA) and still be labelled as having SLE. Collaboration will continue to be necessary to further modify classification of SLE with increased and diverse sample sizes. A study group of the European Society of Cutaneous Lupus Erythematosus (EUSCLE) has defined a core set of variables for the evaluation of the characteristic features of cutaneous lupus, resulting in the development of the EUSCLE Core Set Questionnaire.³ This database provides standardised assessment and monitoring of cutaneous lupus erythematosus (CLE) and has begun to explore prospective therapeutic trends and efficacy in the different subtypes of CLE.⁴

Merola *et al*⁵ have published a provocative advance in the arena of classification models. Using a 'pre-Delphi exercise' and in the future moving to a 'Delphi consensus technique', their work may supplement ACR, SLICC and EUSCLE databases and assist in defining better therapeutic options for our patients through more refined clinical trials based on

better classification of CLE. While the Delphi technique is appealing, overall the track record of the Delphi method over the years has been mixed. One conclusion of Merola *et al* was the suggestion to completely separate CLE from SLE based on the question 'defining CLE from SLE is important'. No doubt separating these two poles of the 'nomenclature' spectrum emphasises important prognostic and therapeutic differences as pointed out in their paper. By doing so, the role of all dermatologists is then modified from managing CLE to mandating the knowledge of appropriate screening for active systemic involvement and its severity. All dermatologists who care for patients with CLE must then perform the necessary review of systems, clinical exams and laboratory studies required to screen for other organ system disease.⁶ Besides the ACR and SLICC criteria, knowledge of the Cutaneous Lupus Disease Area and Severity Index and knowledge of Systemic Lupus Erythematosus Disease Activity Index (2000)⁷ which measures a range of disease activity as recognised by the clinician and the modified version of the British Isles Lupus Assessment Group's Activity Index (2004) used to classify the severity of systemic involvement will need to be used by all dermatologists caring for people with CLE.⁸ If more than mucocutaneous lupus erythematosus (LE) is identified, dermatologists must then as now refer the patient to a rheumatologist or other appropriate specialist. If CLE classification into a standalone disease as suggested by Merola *et al* occurs, it will be interesting to follow the effects it has on the current validated instruments for classification, activity and damage.

Currently the ACR, SLICC and EUSCLE are attempting to refine the classification of lupus in order to clearly define the CLE and SLE poles of the LE immunopathogenic spectrum which of course is especially important in clinical trials and the development of better therapeutic strategies. In addition, the very recent Lupus Foundation of America-Rapid Evaluation of Activity in Lupus is a pilot application with simplified SLE outcome measures

and is also being developed for use in both clinical trials and clinical care.⁹ Improvement in clinical outcomes by integrating all this new knowledge while extremely challenging is also exhilarating and relies on the continued dedication, passion and perseverance of all those who do research and/or care for people with this disease.

Competing interests None.

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