Correspondence on '2019 European League Against Rheumatism/American College of Rheumatology classification criteria for systemic lupus erythematosus' by Aringer *et al*

The 2019 European League against rheumatism/American College of Rheumatology classification criteria (EULAR/ACR 2019 criteria) for systemic lupus erythematosus (SLE) has introduced a new scoring system to classify SLE.¹ The EULAR/ACR 2019 criteria include positive antinuclear antibody at least once as obligatory entry criterion; followed by additive weighted criteria grouped in seven clinical and three immunological domains and weighted from 2 to 10. Patients fulfilling at least one clinical criterion and accumulating ≥ 10 points are classified. In validation cohort, a classification threshold score of ≥ 10 yielded a sensitivity similar to that of the Systemic Lupus International Collaborating Clinics (SLICC) 2012 criteria (96.1% vs 96.7%) and a specificity similar to that of the ACR 1997 criteria (93.4% vs 93.4%), demonstrating both excellent sensitivity and specificity. However, we have two concerns about its additive criteria and methodology.

First, some gastrointestinal injuries related to SLE, especially lupus enteritis, may be underestimated. Gastrointestinal symptoms are reported to occur in more than 50% of patients with SLE at some point in the course of their disease;² however, these symptoms are usually mild.³ Although lupus enteritis manifestations are non-specific (eg, abdominal pain, nausea, vomiting, anorexia and diarrhoea) and have wide range from mild to life-threatening (perforation and fistulisation), it has relatively specific features ('double-halo' and 'comb sign') on contrast-enhanced CT.⁴ The 'double-halo' (namely 'target sign') is a marker of abnormal bowel wall submucosal thickening, whereas the 'comb sign' correlates with mesenteric vessel prominence.⁴ However, the described abnormalities can also be seen in patients with pancreatitis, mechanical bowel obstruction, peritonitis or inflammatory bowel disease.⁵ Lupus enteritis mainly affects the small intestine; in rare circumstances, the colon and rectum can also be involved.^{6–12} Because of lack of radiology and endoscopy studies on the newly onset SLE, the actual incidence rate of lupus enteritis remains unknown. Recently, we encountered a case of severe lupus enteritis with multiple rectal ulcers and fistulisation formation (figure 1). This is a male patient in his 30s who presented with severe diarrhoea, haematochezia and weight loss for 3 months. He had no dyspnoea, neuropsychiatric, musculoskeletal or mucocutaneous manifestations. Several days before admission, he had cough and low grade fever and this can be explained by mild community-acquired pneumonia and right-side pleural effusion confirmed by his chest CT. After admission, a transthoracic echocardiogram showed a slight pericardial effusion. Pleural or pericardial effusion can be explained by his hypoproteinemia, largely attributable to the protein-losing enteropathy caused by enteritis and rectal ulcers. Although the diagnosis of SLE was subsequently made according to his proteinuria (1.09 g/24 hours), hypocomplementemia (C3: 0.2 g/L, C4: 0.08 g/L) and SLE-specific antibody (anti-dsDNA antibody: >800 IU/mL), in terms of clinical domains in EULAR/ACR 2019 criteria, we felt lupus enteritis 'triumphing over' the seven orthodox clinical domains. Unfortunately, lupus enteritis has not yet been considered in ACR 1997 criteria, SLICC 2012 criteria or EULAR/ACR 2019. It is not even a candidate criteria in patients with early SLE,¹³

Figure 1 Endoscopic examination of the rectum. Multiple deep ulcers with mucosal friability, submucosal haemorrhage and purulent secretion (A–C) and fistulisation (D) were observed. These endoscopic findings are not exactly the same as ulcerative colitis and Crohn's disease.

some of which subsequently being refined and constitute the EULAR/ACR 2019 criteria.

Second, rheumatologists should be informed of exact probability of illness in patients with underlying SLE who are below the threshold (ie, total score <10) so as to provide better decision-making, evaluation and follow-up. It is preferable to use logistic regression and nomogram to predict the probability. In addition, when patients have signs or symptoms suggestive of but not diagnostic of SLE, their physician must decide whether to (1) treat empirically, (2) not treat or (3) perform further diagnostic testing before deciding between options 1 and 2. Under this circumstance, decision-making based on the threshold of 10 generated by the receiver operating characteristics analysis seems risky, especially when clinical and immunologic parameters are ambiguous. Rheumatologists should also be informed of the net benefit¹⁴ from the patients when diagnosis is made and treatment is given at a threshold of 10. This net benefit comparison should be suggested to carry out among ACR 1997 criteria, SLICC 2012 criteria, and EULAR/ACR 2019 criteria.

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