Higher specificity of the new EULAR/ACR 2019 criteria for diagnosing systemic lupus erythematosus in patients with biopsy-proven cutaneous lupus

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Abstract

Objective

The risk of developing systemic lupus erythematosus (SLE) in patients with cutaneous lupus erythematosus (CLE) varies, ranging between 5 to 23%, depending on the disease subtype. Interestingly, most of these patients do not manifest clinically significant internal organ features of SLE. The aim of our study was to evaluate the percentage of CLE patients who fulfilled SLE criteria introduced by the American College of Rheumatology (ACR 1997) and Systemic Lupus Erythematosus International Collaborating Clinics (SLICC 2012), as well as the new criteria developed by the European League Against Rheumatism and ACR (EULAR/ACR 2019).

Methods

Patients were evaluated at baseline and during follow-up, and the severity of systemic symptoms was assessed. We retrospectively analysed the medical histories of 184 patients with CLE (75 with discoid lupus erythematosus and 109 with subacute cutaneous lupus erythematosus). The mean duration of follow-up after CLE diagnosis was 58 months (24–120 months).

Results

Of the analysed patients, 23.4%, 17.4% and 14.7% met the ACR 1997, SLICC 2012 and EULAR/ACR 2019 classification criteria for SLE at baseline, respectively. There was no significant difference in this proportion after follow-up. All of the CLE patients fulfilling SLE criteria demonstrated no-to-mild internal organ involvement and laboratory abnormalities such as cytopenia or complement levels were mild or only slightly decreased.

Conclusion

The EULAR/ACR 2019 criteria are characterised by higher specificity for SLE diagnosis when compared to previously introduced criteria sets. We conclude that patients with CLE, even those meeting the criteria for SLE, have low risk of serious complications of SLE.

Key words

lupus, skin, EULAR, ACR, diagnostic criteria

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Introduction

Cutaneous lupus erythematosus (CLE) is a chronic autoimmune disease that can be a separate entity or the skin manifestation of systemic lupus erythematosus (SLE). It has been shown that a subset of patients with CLE only may go on to develop SLE later (1). Depending on the subtype of cutaneous lupus, the cumulative risk of SLE is between 5% and 23% within 5–25 years (1-3). The three classification criteria sets for SLE introduced by 1) the American Rheumatism Association (final amendment in 1997; ACR 1997) (4), 2) in 2012 by the Systemic Lupus Erythematosus International Collaborating Clinics (SLICC 2012) (5), and 3) those developed in 2019 by the ACR in collaboration with the European League Against Rheumatism (EULAR/ACR 2019) (6) include clinical, laboratory and immunological features. As shown in previous studies, most patients with CLE who met the ACR 1997 and SLICC 2012 criteria for SLE did so mainly based on the mucocutaneous criteria such as photosensitivity, discoid rash and oral ulcers accompanied by the presence of antinuclear antibodies (ANAs) (1, 7). However, it has been recently suggested that CLE patients meeting the SLE criteria should not be considered as evidence of transition to SLE since most of these patients do not manifest clinically significant internal organ features of SLE such as nephritis, cerebritis or prominent cytopenias even during follow-up (8-10).

The new EULAR/ACR 2019 classification criteria set was shown to have greater sensitivity and specificity for earlier diagnosis of lupus. However, the greatest innovation of EULAR/ACR 2019 classification criteria set is that each feature has a differential weighted value. Thus, for example mucocutaneous manifestation is no longer equally weighted compared to renal involvement (6). Therefore, we considered it important to determine whether the new EULAR/ACR 2019 diagnostic criteria provide better prognostic information regarding outcome in patients with cutaneous lupus in regards to the risk of systemic involvement.

We performed a retrospective study to determine the percentage of CLE patients who fulfilled EULAR/ACR 2019 criteria for SLE diagnosis both at the time of diagnosis of cutaneous lupus and during a follow-up. Moreover, we assessed the number of patients meeting the other two classification sets (*i.e.* ACR 1997 and SLICC 2012). We categorised which features from the ACR 1997, SLICC 2012 and EULAR/ACR 2019 criteria were seen in the included patients. Furthermore, we attempted to identify the type and severity of systemic symptoms during follow-up among individuals with CLE.

Methods

Study subjects and data collection

To accomplish the study goals, we used an ongoing database of patients with cutaneous lupus erythematosus who were diagnosed and treated in the Department of Dermatology at the Medical College of Jagiellonian University in Cracow, Poland between January 1, 2008 and February 26, 2018. We selected this time interval to ensure data completeness for all 3 sets of classification criteria and at least 2 years of follow-up. All the patients included in this study had a final diagnosis of cutaneous lupus erythematosus (DLE or SCLE) which was confirmed by histopathological examination. All patients with arthralgia, independent of the time of onset, were examined by a rheumatologist to ensure a clear diagnosis. The following information was extracted from each patient's medical record: demographic characteristics; clinical features such as photosensitivity, alopecia, oral ulcers, and symptoms involving musculoskeletal, nervous and cardiopulmonary systems; laboratory findings including complete blood count (CBC), urinalysis, serum complement levels (C3, C4), antiphospholipid antibodies (APLAs), antinuclear antibody (ANA), anti-dsDNA antibody (determined by CLIFT; [Crithidia luciliae immunofluorescence test]) and antibodies for extractable nuclear antigens (ENA); treatments and follow-up data. Baseline and follow-up data were used to determine which individuals met the diagnostic criteria for SLE proposed by EULAR/ACR 2019, (6) ACR 1997 (4) and those presented by SLICC in 2012 (5).

Table I. Baseline and follow-up characteristics of patients with DLE and SCLE.

	Baseline		*p*	Follow-up		*p*
	DLE (n=75)	SCLE (n=109)		DLE (n=75)	SCLE (n=109)	
Clinical findings						
Alopecia, n (%)	19 (25.3)	21 (19.3)	NS	17 (22.7)	22 (20.2)	NS
Oral ulcers, n (%)	1 (1.3)	3 (2.8)	NS	1 (1.3)	4 (3.7)	NS
Serositis, n (%)	0	0	NS	0	0	NS
Arthritis, n (%)	1 (1.3)	2 (1.8)	NS	0	0	NS
Arthralgia, n (%)	38 (50.7)	32 (29.4)	0.005	19 (25.3)	14 (12.8)	0.03
Photosensitivity	32 (42.7)	45 (41.3)	NS	30 (40)	45 (41.3)	NS
Immunological findings						
Positive for ANA, n (%)	56 (74.7)	77 (70.6)	NS	54 (72)	77 (70.6)	NS
ANA titre, (min-max)	320 (160-10240)	1280 (160-20480)	0.03	320 (160-5120)	640 (160-10240)	NS
Positive for anti-dsDNA, n (%)	4 (5.3)	3 (2.8)	NS	8 (10.7)	5 (4.6)	NS
Anti-dsDNA titre, (min-max)	40 (10-80)	30 (10-160)	NS	30 (10-60)	30 (10-80)	NS
Positive for anti-Sm, n (%)	0	1 (0.9)	NS	1 (1.3)	1 (0.9)	NS
Positive for APLA or LA, n (%)	5 (6.7)	5 (4.6)	NS	5 (6.7)	6 (5.5)	NS
Low C3, (<0.85 g/L), n (%)	11 (14.7)	31 (28.4)	0.03	13 (17.3)	35 (32.1)	0.02
C3 levels, g/L (min-max)	1.1 (0.02-1.5)	1.1 (0.5-1.7)	NS	1.2 (0.08-1.6)	1.1 (0.6-1.8)	NS
Low C4, (<0.15 g/L), n (%)	4 (5.3)	9 (8.3)	NS	4 (5.3)	11 (10.1)	NS
C4 levels, g/L (min-max)	0.2 (0.07-0.54)	0.19 (0.02-1.03)	NS	0.2 (0.1-0.6)	0.2 (0.05-1.1)	NS
General laboratory findings						
Leukopenia (<4000/uL), n (%)	8 (10.7)	17 (15.6)	NS	10 (13.3)	18 (16.5)	NS
WBC, cells/uL (min-max)	5.3 (3.1-8.6)	4.8 (2.7-10.4)	NS	5.8 (3.4-9.2)	5.4 (2.9-9.9)	NS
Lymphopenia (<1000/uL), n (%)	27 (36)	57 (52.3)	0.04	21 (28)	43 (39.4)	NS
Lymphocytes; cells/uL (min-max)	1.8 (0.8-3.4)	1.6 (0.8-5.2)	NS	1.7 (1.0-3.7)	1.9 (0.9-4.9)	NS
Thrombocytopenia (<100000/uL), n (%)	2 (2.7)	2 (1.8)	NS	3 (4)	3 (2.8)	NS
PLT, cells/uL (min-max)	204.5 (94-340)	193.5 (76-364)	NS	210 (91-350)	202 (78-390)	NS
Proteinuria (>0.5 g/24h), n (%)	0	1 (0.9)	NS	1 (1.3)	2 (1.8)	NS
Autoimmune haemolysis, n (%)	0	0	NS	0	0	NS

DLE: discoid lupus erythematosus; SCLE: subacute cutaneous lupus erythematosus; ANA: antinuclear antibodies; APLA: antiphospholipid antibodies; LA: lupus anticoagulant; WBC: white blood count; PLT: platelets.

All study participants gave their informed written consent, and the study was approved by the Jagiellonian University Bioethical Committee (approval no. 1072.6120.76.2020)

Statistical analysis

Statistical analysis was performed with the Statistica v. 7.1 PL package (StatSoft, 2005). Data are expressed as median and min-max unless otherwise stated. Between group comparisons were performed using the Mann-Whitney U-test and Wilcoxon signed-rank test. *p*-values <0.05 were considered statistically significant.

Results

Patient characteristics

Patients (n=184) with histologically and clinically confirmed cutaneous lupus erythematosus were included in this study (75 with DLE and 109 with SCLE), 75.5% of them were females. Mean age at the time of diagnosis was 59 years (range: 20–85 years). The mean duration of follow-up after diagnosis of cutaneous lupus was 58 months (range: 24–120 months). All individuals in our cohort received topical corticosteroids. Due to the lack of clinical improvement, therapy with oral hydroxychloroquine (n=132) or oral corticosteroid, or a combination of both (n=98) was initiated in 72% of the patients. Most patients in this group showed complete or partial response to therapy in, on average, less than 1 year. The duration of treatment with equivalent of ≥20 mg/day of prednisolone was no longer than 6 weeks among individuals who required systemic corticosteroids. Relapses were seen after drug discontinuation in approximately 70% (n=92) of these patients.

Table I shows baseline and follow-up characteristics of included patients based on the subsets of cutaneous lupus erythematosus (*i.e.* DLE *vs.* SCLE).

Photosensitivity was the most common finding in the patients at baseline and follow-up (41.8% and 40.7%, respectively). About 40% of individuals complained of arthralgia, which was significantly more common among DLE patients when compared to those with SCLE ($p<0.05$, Table I). Interestingly, a significant decrease in prevalence of joint pain was observed during follow-up.

Antinuclear antibodies were present in 72.3% of all included patients. Low serum complement C3 levels were found in 22.8% individuals and this was significantly more common among patients with SCLE in comparison to those with DLE (Table I).

Lymphopenia (<1000 cells/μL) was found to be the most frequent complete blood count abnormality (overall incidence of 45.7%). Mean lymphocyte count among these patients was 870 cells/μL. Prevalence of thrombocytopenia was 2.2%.

In the whole group of patients, 10 (5.4%) and 11 (5.9%) cases were identi-

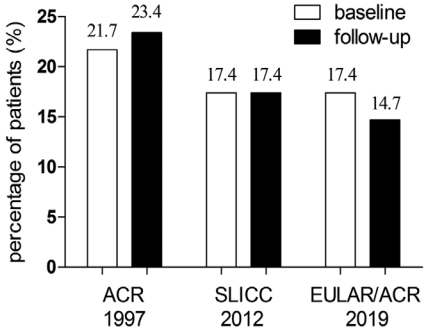

Fig. 1. Percentage of patients with cutaneous lupus erythematosus who met ACR 1997, SLICC 2012 and EULAR/ACR 2019 criteria sets for systemic lupus erythematosus at baseline and after follow-up.

fied who were positive for antiphospholipid antibodies at baseline and during follow-up, respectively. At baseline, the mean titres (min-max) for these antibodies were as follows: aCL (anticardiolipin) IgM 7.5 (1–32.1) MPL/mL, aCL IgG 2.95 (1–16.8) GPL/mL, β2GPI (β2glycoprotein) IgM 2.8 (0.72–180.2) SMU/mL, β2GPI IgG 0.35 (0.1–4.1) SGU/mL. None of patients fulfilled clinical criteria for antiphospholipid syndrome (both at baseline and during follow-up).

Systemic involvement

None of the patients with CLE (including those fulfilling SLE criteria and those that did not) had fever that could not be explained by infection, cardiopulmonary, neurologic, kidney involvement or laboratory and clinical findings characteristic of haemolytic anaemia both at baseline and follow-up. In one patient with SCLE, low-grade proteinuria (0.7 g/24 h) was identified at the time of cutaneous lupus diagnosis. During the follow-up (60 months), neither an increase in proteinuria nor an appearance of urinary cellular casts was noted. In one patient in the DLE and one in the SCLE group, a low-grade proteinuria (<1 g/24 h) was found during follow-up. Uncontrolled hypertension was diagnosed by the consultant nephrologist as the cause of proteinuria.

Fulfilling classification criteria for SLE

The percentages of patients fulfilling the ACR 1997, SLICC 2012 and EULAR/ACR 2019 classification criteria for SLE at the time of cutaneous lupus diagnosis were 23.4%, 17.4% and 14.7%, respectively. There were no significant differences in percentage of patients who met classification criteria between the DLE and SCLE groups (data not shown). Interestingly, there were also no significant differences in percentage of individuals fulfilling classification criteria for SLE at the baseline and after follow-up (Fig. 1). Due to this finding, further analysis was performed only including patients who met classification criteria for SLE at the time of CLE diagnosis. Figure 2 shows the incidence of specific clinical and laboratory findings among groups fulfilling the EULAR/ACR 2019 (Fig. 2A), SLICC 2012 (Fig. 2B) and ACR 1997 (Fig. 2C) criteria.

In the EULAR/ACR 2019 criteria patients must have antinuclear antibody levels of at least 1:80. Among individuals fulfilling the EULAR/ACR 2019 criteria (27/184), low serum complement C3 levels (59.3%), leukopenia (48.1%), presence of antibodies against dsDNA (37%) and low serum C4 levels (33.3%) were found frequently (Fig. 2A). In this group of patients, mean serum complement C3 and C4 levels were 0.76 g/L and 0.14 g/L, respec-

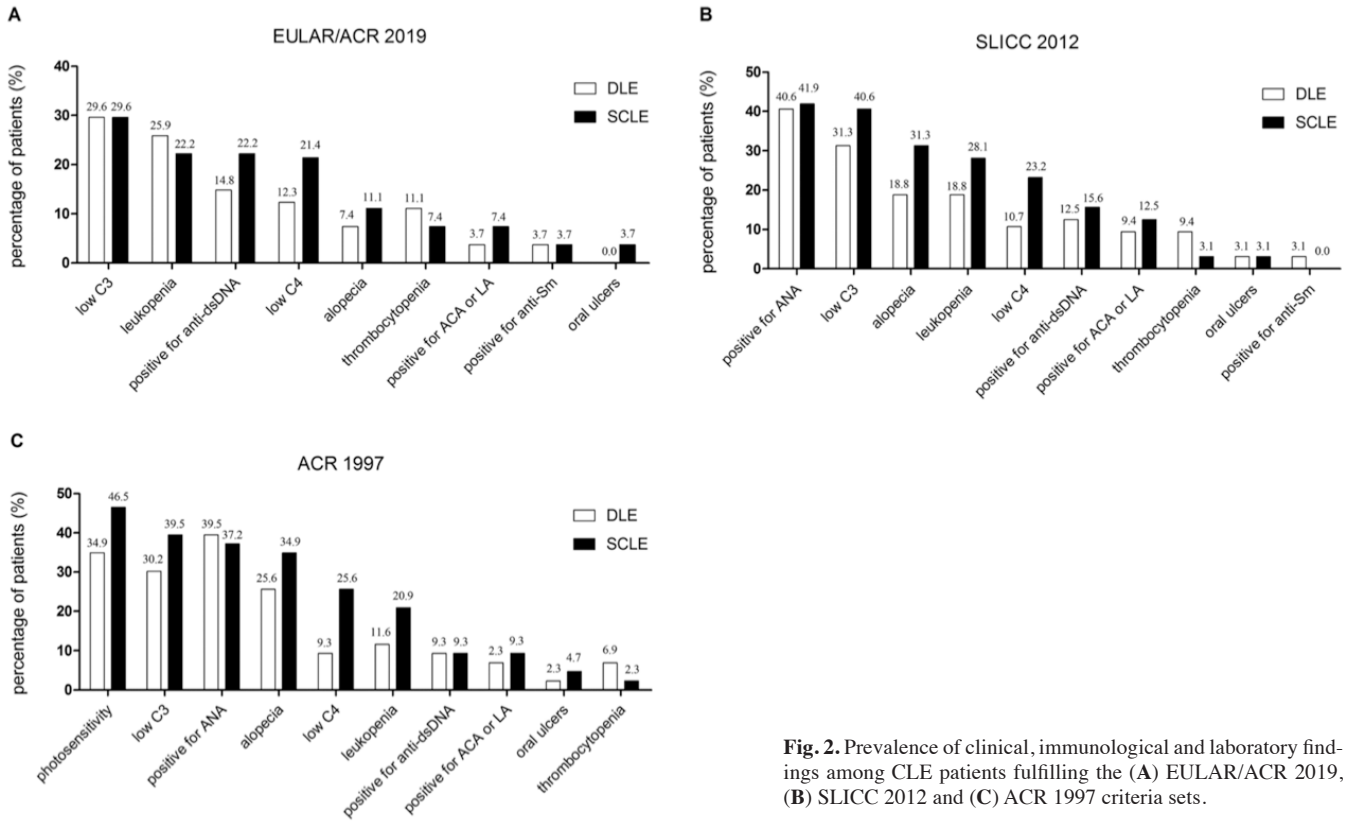

Fig. 2. Prevalence of clinical, immunological and laboratory findings among CLE patients fulfilling the (**A**) EULAR/ACR 2019, (**B**) SLICC 2012 and (**C**) ACR 1997 criteria sets.

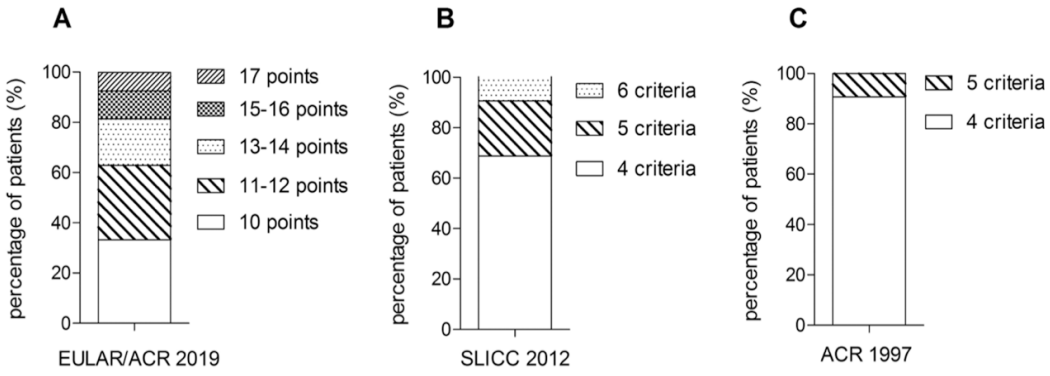

Fig. 3. Number of SLE criteria which occurred in patients who met (**A**) EULAR/ACR 2019, (**B**) SLICC 2012 and (**C**) ACR 1997 criteria sets.

tively. Median white blood count was found to be of 3500/μL and mean titre of anti-dsDNA antibodies was 1:40.

Presence of ANA (96.9%), low serum complement C3 levels (71.9%), alopecia (50%) and leukopenia (46.9%) were the most common features in patients meeting SLICC 2012 criteria (32/184; Fig. 2B). Among patients who fulfilled the ACR 1997 criteria, the most common features were photosensitivity (81.4%), presence of ANA (76.4%), low serum complement C3 levels (69.8%) and alopecia (60.5%) (43/184; Fig. 2C). One third of patients meeting the ACR 1997 criteria accumulated only 10 points (Fig. 3A). Only four SLE criteria according to the SLICC 2012 and ACR 1997 were identified in 70% and 90% of patients fulfilling these classification criteria, respectively (Fig. 3A-B). Interestingly, similar percentages were found when looking at follow-up data (not shown).

Discussion

Until now, there have been only a few studies devoted to CLE and its transition to SLE (1, 8-11). Depending on the subtype of CLE, the risk of progression to SLE is between 5-23% (1-3). However, in most of these studies, progression to SLE is defined as meeting the diagnostic criteria for SLE. Recently, it has been shown that most of the CLE patients who fulfilled SLE criteria (ACR 1997 and SLICC 2012) did so primarily by meeting mucocutaneous criteria, without manifestation of clinically significant internal organ features of SLE such as nephritis, cerebritis or prominent cytopenias. In fact, even if they develop SLE, the majority have no to mild additional systemic disease (8-10). Thus, it was suggested that this overdiagnosis of SLE among CLE patients may lead to the negative psychological effects such as anxiety and significant lowering of quality of life of affected individuals.

Our study is the first that we are aware of to determine the percentage of CLE patients who fulfilled the new EULAR/ACR 2019 criteria both at the time of cutaneous lupus diagnosis and at the end of follow-up. We compared the EULAR/ACR 2019, SLICC 2012 and ACR 1997 classification criteria sets, seeking to determine a link between criteria scores at onset and outcome.

Using the EULAR/ACR 2019, SLICC 2012 and ACR 1997 criteria, 14.7%, 17.4% and 23.4% patients met criteria for SLE at the time of cutaneous lupus diagnosis, respectively. This proportion of individuals fulfilling each of these sets did not change significantly at the end of follow-up. Interestingly, none of the analysed patients manifested clinically significant internal organ features of SLE (*i.e.* nephritis, central nervous system involvement, prominent cytopenias) both at baseline and follow-up. Interestingly, the laboratory and immunological abnormalities that were identified were mild in almost all of our patients (*i.e.* only small decrease in C3 complement levels, mild leukopenia and thrombocytopenia and low titres of anti-dsDNA antibodies). Thus, it seems that despite improved specificity of EULAR/ACR 2019 criteria when compared to the previous ones (SLICC 2012, ACR 1997), there is still a significant number of CLE patients that are over-diagnosed with SLE.

Since our patients did not progress to SLE (did not fulfill SLE criteria) during the follow-up, we may only hypothesise that the cutaneous lupus phenotype may be determined at the onset of the disease. Thus, there are CLE patients who: 1) remain CLE only, 2) additionally met the SLE criteria, and 3) met the SLE criteria and develop significant internal organ involvement. However, the latter group was not identified in our study.

Recently Alniemi *et al.* (9) showed that only 1 of 90 patients with SCLE (1.1%) had lupus nephritis. Thus, our results align with the previous observations that patients with cutaneous lupus erythematous, including discoid and subacute subtypes, have been found to have a more mild disease course if SLE develops.

It is noteworthy that as many as 70% of patients in our study were treated with oral hydroxychloroquine alone or in combination with oral steroids. Thus, that such management could prevent development of systemic manifestation cannot be excluded. Further studies in this area are needed.

When comparing the number of patients with CLE meeting the ACR 1997 and SLICC 2012 criteria in previous studies, our results seem to be similar. On the other hand, Biazar *et al.* (11) showed that almost 40% of individuals with cutaneous lupus fulfilled criteria for SLE. This may be explained by the fact that some patients included in their study had acute cutaneous lupus erythematosus, a subtype with the strongest relationship to SLE with systemic symptoms.

In the new EULAR/ACR 2019 classification criteria set all patients must have antinuclear antibodies levels of at least 1:80. There are 10 criteria, clinical and immunologic, each of which is

weighted toward the highest score attributable to the patient. A total score of at least 10 is necessary for a diagnosis of SLE. Among all of our CLE patients who met EULAR/ACR 2019 criteria, one third received only 10 points. The next one third scored only 11 or 12 points. Similarly, only four ACR 1997 and SLICC 2012 criteria for SLE were met in 90% and 70% of individuals, respectively.

Among all of the analysed features consistent with SLE diagnosis, photosensitivity, presence of ANA and decreased serum complement C3 levels were the most commonly found in our study. Photosensitivity has been reported in 25% to 100% of patients with cutaneous lupus, depending on the subtype of CLE (12). However, this feature is often only evaluated by patient's history and is poorly defined by the classification criteria. Recent studies in which photoprovocation was performed showed that in nearly 40% of cases the results did not correspond to the patient's history of photosensitivity (13). Thus, presumably it would be more useful to replace photosensitivity with the photoprovocation test.

The most frequent features found among patients fulfilling the new EULAR/ACR 2019 criteria were decreased of serum complement C3 levels, leukopenia and presence of anti-dsDNA antibodies. It should be however noted that C3 levels were only slightly decreased (mean serum C3 complement levels of 0.76 g/L), leukopenia was assessed as mild and there were only low titres of anti-dsDNA antibodies.

Some important clinical and pathogenic associations between mucocutaneous items such as alopecia, oral ulcers and cutaneous lupus lesions have been recently suggested (14). Thus, in EULAR/ACR 2019 criteria they were grouped in one domain within only the highest scoring item is being scored for classification. In the ACR 1997 and SLICC 2012 there were classified independently. It could be one of the most important explanation of the relatively smaller number of individuals with CLE who fulfilled EULAR/ACR 2019 criteria in comparison to the previous two sets.

As many as 38% of patients in our study reported having arthralgia. Recently, a very high prevalence of ultrasound joints abnormalities including entheses and tendons was found in lupus erythematosus patients who had even mild joint symptoms (15, 16). Thus, all our patients with arthralgia have been consulted by rheumatologist, and in all of them, inflammatory arthritis had been excluded. Therefore, according to the emerged recent data we strongly recommend referrals to rheumatologists who will be helpful to pinpoint the diagnosis (15, 16).

Our study is limited by its retrospective design and data extraction from medical records. Accordingly, some information may have been missed or underestimated. For example, drug-induced SCLE has recently been reported to be more common than first reported (17). This subtype of cutaneous lupus is generally associated with mild symptoms and no systemic involvement. In the current study, we were unable to confirm drug-induced SCLE in the included patients.

In conclusion, we showed for the first time that a relatively smaller number of CLE patients met the new EULAR/ACR 2019 criteria set for SLE in comparison to previously introduced sets (SLICC 2012 and ACR 1997). The percentage of individuals fulfilling SLE criteria did not change significantly during the follow-up in all classification criteria sets. None of the CLE patients manifested clinically significant internal organ features of SLE during follow-up. Our findings support the hypothesis that patients with CLE, even if they fulfill the SLE criteria, have very low risk of serious complication of SLE.

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