

Cutaneous Lupus Erythematosus: Disease Area and Severity Index reliability in clinical evaluation and assessment of cutaneous lupus erythematosus lesions in pediatric cases

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Abstract

Background: Children having systemic lupus erythematosus clinically reflect a management and diagnostic challenge. Cutaneous lupus erythematosus reflects the dermatological signs of SLE categorized into LE-specific and LE-nonspecific features. The Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) and Physician Global Assessment (PGA) are clinical tools categorizing the dermatological disease severity in cases having cutaneous lupus disease

Aim of study: Assessment of CLASI scoring reliability and usefulness in pediatric cutaneous lupus.

Methodology: A clinical research trial conducted on 100 research study subjects with pediatric cutaneous lupus erythematosus. Cases had an age range of 6–18 years. All research study subjects were assessed using both the CLASI and skin Physician's Global Assessment clinical tools.

Results: CLASI activity had excellent inter-rater and intra-rater reliability scores among dermatologists and rheumatologists (ICC = 0.965, 0.993, 0.924, 0.975 consecutively), CLASI damage had good interrater and intra rater scores among dermatologists (ICC = 0.810, 0.893 consecutively) on the other hand among rheumatologists it had poor interrater score (ICC = 0.397) and good intra rater score (ICC = 0.786). PGA activity had good inter rater reliability (ICC = 0.830) and excellent intra rater variability (0.912) among dermatologists and among rheumatologists had fair inter rater and intra rater reliability (ICC = 0.754, 0.784 consecutively) PGA damage had moderate inter rater reliability (ICC = 0.593, 0.684) and fair intra rater reliability (ICC = 0.783, 0.756) among dermatologists and rheumatologists consecutively.

Introduction

Systemic Lupus erythematosus is a systematic chronic autoimmune illness characterized by a broad range of clinical signs. Even though SLE in childhood is more severe at pathological and clinical levels of presentation with a great percentage of children having considerable renal or central nervous system affection at the time of clinical diagnosis, interestingly the raised severity may could arise from delayed clinical diagnosis and poor levels of compliance to management protocols [1,2].

Children having systemic lupus erythematosus clinically reflect a management and diagnostic challenge. Early onset permits the observation of the natural history of SLE free from the confounding issues usually observed in older cases. Continuing physical and emotional development and growth impacts the medications choice particularly in presence of cutaneous manifestations affecting the probability of management protocol success. Clinical satisfactory outcome for SLE in a child is not 5-year or 10-year survival rate,

on the other hand 50-year or 60-year survival is considered the real clinical challenge [3,4].

Cutaneous lupus erythematosus reflects the dermatological signs of SLE categorized into LE-specific and LE-nonspecific features. LE-specific features are characterized by a typical vacuolar change within basal cell layer in conjunction to lichenoid tissue. The features and clinical categories of cutaneous lupus erythematosus in correlation to systemic disease have been considerably and extensively investigated in adult age groups however research data in pediatric population affected is scarce. Around 10% to 20% of cases having systemic lupus Disease have their first symptom under 18 years of age. Above 70% of children suffering systemic lupus develop skin lesions, the term 'lupus erythematosus' reflects a wide spectrum of correlated system disorders [5,6].

Around 80% of lupus cases eventually develop characteristic dermatological lesions at some time point during their illness.

Clinicians and pharmacological manufacturers require track clinical effectiveness of treatment precisely that requires reliable assessment tools. The Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) and Physician Global Assessment (PGA) are clinical tools categorizing the dermatological disease severity in cases having cutaneous lupus disease [7,8].

Cutaneous lupus in pediatric patients is a chief issue causing morbidity, quality of life impairment and psychosocial insults. Proper assessment permits proper and improved management protocols evaluation therefore aiding in enhancement level of care in those categories of cases among pediatric population [9,10].

Aim of study

Assessment of CLASI scoring reliability and usefulness in pediatric cutaneous lupus.

Methodology

A clinical research trial conducted on 100 research study subjects with pediatric cutaneous lupus erythematosus. Cases had an age range of 6–18 years. All research study subjects were assessed using both the CLASI and skin Physician’s Global Assessment clinical tools.

Skin Physician’s Global Assessment

Estimates overall skin disease severity and has been used in multiple validation studies for skin diseases. It requires physicians to score the cases of skin related health on a 0–10 scale (0 = worst skin condition imaginable, 10 = perfect health) for his or her overall skin score, skin activity and skin damage. It also asks the physician to rate the patient’s overall skin condition as mild, moderate or severe. Assessment and evaluation of inter- and intra-rater reliability Inter-rater reliability was assessed among the dermatologists and the rheumatologists separately and together.

Statistical analysis

Data were collected, revised and entered to the Statistical Package for Social Science (SPSS) version 23. Quantitative data were presented as mean, standard deviations and ranges while qualitative data were presented as numbers and percentages. In each physician group (dermatologists and rheumatologists) we use interclass correlation coefficient (ICC) to assess the intra-rater reliability for each of the four studied scores while within each physician group we estimate inter-rater ICC. The confidence interval was set to 95% and the margin of error accepted was set to 5%. So, the p-value was considered significant at $P < 0.05$ in all analysis.

Table (1): Demographic data and characteristics of the studied patients

	No. = 100
Age at presentation (years) Mean \pm SD Range	13.32 \pm 3.42 6 – 18
Age at diagnosis (years) Mean \pm SD Range	11.4 \pm 3.5 5 – 16
Gender Males Females	15 (15.0%) 85 (85.0%)
Predominant CLE subtype Acute CLE Discoid lupus erythematosus Tumid lupus erythematosus Subacute CLE	35 (35%) 35 (35.0%) 20 (20.0%) 10 (10.0%)
Medications Hydroxychloroquine Mycophenolate mofetil Prednisone Dapsone Methotrexate	75 (75.0%) 32 (32.0%) 35 (35.0%) 20 (20.0%) 10 (10.0%)

Table 1 reveals and displays that among the 100 research study subjects recruited for the study age at presentation Mean \pm SD, age at presentation, diagnosis(13.32 \pm 3.42 yrs , 11.4 \pm 3.5 yrs ,consecutively) males represented 15 percent of the study cohort ,and females represent 85 percent , as regards Predominant CLE subtype Acute CLE represented 35 % of cases , Discoid lupus erythematosus represented 35 % of cases, Tumid lupus erythematosus represented 20 % , Subacute CLE represented 10 % of cases

Table (2): Inter-rater and intra-rater reliability of the studied scores

	Inter-rater ICC (95% CI)	Intra-rater ICC (95% CI)
Dermatologists (no. = 3)		
CLASI activity	0.965 (0.895 – 0.992)	0.993 (0.989 – 1.000)
CLASI damage	0.810 (0.721 – 0.923)	0.893 (0.764 – 0.973)
PGA activity	0.830 (0.746 – 0.945)	0.912 (0.776 – 0.982)
PGA damage	0.593 (0.424 – 0.723)	0.783 (0.654 – 0.883)
Rheumatologists (no. = 3)		
CLASI activity	0.924 (0.846 – 0.984)	0.975 (0.931 – 0.994)
CLASI damage	0.397 (0.164 – 0.685)	0.786 (0.646 – 0.915)
PGA activity	0.754 (0.612 – 0.895)	0.784 (0.631 – 0.919)
PGA damage	0.684 (0.512 – 0.864)	0.756 (0.421 – 0.882)

95% CI: 95% Confidence interval; CLASI: Cutaneous lupus erythematosus disease area and severity index; ICC: Intraclass correlation coefficients; PGA: Physician's Global Assessment

CLASI activity had excellent inter-rater and intrarater reliability scores among dermatologists and rheumatologists (ICC = 0.965, 0.993, 0.924, 0.975 consecutively), CLASI damage had good inter-rater and intrarater scores among dermatologists (ICC = 0.810, 0.893 consecutively) on the other hand among rheumatologists it had poor inter-rater score (ICC = 0.397) and good intra rater score (ICC = 0.786).

PGA activity had good inter-rater reliability (ICC=0.830) and excellent intra-rater variability (0.912) among dermatologists and among rheumatologists had fair inter rater and intra rater reliability (ICC=0.754, 0.784 consecutively) PGA damage had had moderate inter rater reliability (ICC=0.593, 0.684) and fair intra rater reliability (ICC=0.783, 0.756) among dermatologists and rheumatologists consecutively

Discussion

Cutaneous lupus erythematosus have considerable morbidity and quality of life impairment particularly among pediatric populations affected the earlier the onset the more severe the disease and poorer prognosis is expected among undiagnosed and non-compliant cases to treatment [11,12].

Effective clinical scoring systems are of cornerstone value in proper evaluation and follow up of clinically affected cases with cutaneous lesions. CLASI scoring system was previously evaluated and investigated among adult populations affected by cutaneous lupus issues. However pediatric population affected by cutaneous lupus manifestations were not properly evaluated in prior research studies [13,14].

Mount up pathological progressive damage from cutaneous lupus erythematosus is inversely correlated to onset of age of illness triggering physical and psychological sequela during critical times of

growth and development. On the other hand, it was revealed and displayed by various research groups of investigators that, childhood -onset SLE varies considerably from adult-onset SLE, with higher frequency of end-organ damage issues e.g. renal affection and elevated general rates of mortality [15].

A prior research statistical analysis of 26 research study subjects from pediatric age group having cutaneous lupus erythematosus revealed that seven cases (27%) had neonatal lupus erythematosus (71% females and 29% males). Of the other 19 children with cutaneous lupus erythematosus, 95% were female. The mean and median age of clinically established diagnosis was 11 years. All cases investigated had lupus erythematosus -specific cutaneous lesions and 83% had lupus erythematosus -nonspecific dermal manifestations. On the other hand, it was revealed among the research study findings that atypical initial lesions were observed in 28% of the patients, and 22% of the patients there was rare lupus erythematosus variations. Acute cutaneous lupus erythematosus was observed in 83% of cases, subacute lesions in 44% of cases, and chronic in 22%. Autoimmune correlations were observed in 44% of cases and a positive family history of autoimmune disorders in 61% of cases. The research team of investigators came to the conclusion that there was a considerable female majority affection, greater clinical risk of systemic disease in cases clinically presenting with cutaneous lupus erythematosus although the research study is in harmony with current research findings further research efforts are required to elucidate the clinical spectrum of pediatric cutaneous lupus erythematosus all over the globe [1,3,7].

Another prior research study similar to the current research study conducted in approach and methodology have revealed and displayed that CLASI activity scoring system had excellent inter-rater reliability in comparison to PGA, that revealed good inter-rater reliability. Interestingly it is well known that, the activity scoring is a

more a more valuable measure Than damage scoring since current skin disease activity levels is measured in clinical trials [2,4,9].

Furthermore, in harmony and great similarity to the current research study it was shown that generally CLASI scoring system is more trustworthy than tool than the PGA system especially in evaluation of children diseases activity levels [11,13].

Conclusions and recommendations

CLASI revealed excellent levels of inter- and intra-rater reliability amongst dermatologists and rheumatologists, being superior to the PGA

Scoring system. However future research studies are required to consider larger number of cases to further verify the current research study findings and future research studies are recommended to be multicentric in fashion .Racial and ethnic differences should be taken in consideration in future research efforts to have a valuable research data base that could aid in implementation of useful clinical guidelines in diagnosis and management of cases of pediatric cutaneous lupus lesions.

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