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Demographic and clinical characteristics of cutaneous lupus erythematosus at a paediatric dermatology referral centre

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Abstract

Background—Paediatric cutaneous lupus erythematosus (LE) is uncommon and inadequately described in the literature. Similar to adults, children with cutaneous LE develop LE-specific and/or LE-nonspecific skin findings. Similarities and differences in demographics and clinical course between paediatric and adult cutaneous LE have not been sufficiently described.

Objectives—The purpose of this study is to detail the demographic and clinical features of paediatric cutaneous LE and then compare these findings to those reported in the adult literature.

Methods—A retrospective chart review was performed of 53 children seen in a paediatric dermatology clinic with cutaneous manifestations of LE.

Results—Patients presented with all five major subtypes of cutaneous LE, with some notable differences from adult cutaneous LE and previously published reports of paediatric cutaneous LE. Progression from discoid LE to systemic lupus erythematosus (SLE) did not occur in our cohort. Patients with subacute cutaneous LE were more likely than adults to have lesions below the waist as well as concomitant SLE. Sex distribution for cutaneous LE in our study was equal prior to puberty and female-predominant in post-pubertal patients.

Conclusions—Children with cutaneous LE have variable clinical presentations and progression to SLE that may be different from adult disease. Specifically, children with acute and subacute cutaneous LE may be more likely than adults to have systemic disease; therefore, patients with these subtypes should be monitored closely for evidence of SLE. Study limitations included small patient numbers that may limit ability to generalize this data and relatively short follow-up intervals.

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Introduction

Lupus erythematosus (LE) is an autoimmune disease that presents as a vast spectrum of clinical manifestations involving many organ systems, including the skin. Dermatologic findings of LE are termed cutaneous LE and may be classified as either LE-specific or LE-nonspecific.[1] LE-specific cutaneous lesions characteristically show interface dermatitis on histology and include the subtypes chronic cutaneous lupus erythematosus (CCLE), subacute cutaneous lupus erythematosus (SCLE), and acute cutaneous lupus erythematosus (ACLE), each with its own subcategories.[2,3] LE-nonspecific cutaneous findings, such as vasculitis or alopecia, occur frequently in LE patients but may also be seen outside of LE and do not show histologic features characteristic for LE.[2–4]

There are known differences in demographics and disease patterns between childhood-onset and adult-onset systemic lupus erythematosus (SLE).[5] Demographic and clinical characteristics for cutaneous LE in adults are also described in the literature; however, similar information specific to paediatric cutaneous LE is lacking. The pathophysiology and clinical features of paediatric disease may be fundamentally different from that in adults, and therefore, understanding of paediatric disease patterns is necessary. The purpose of this study is to describe in detail the demographic and clinical features of paediatric cutaneous LE and compare these findings to those reported in the adult literature.

Materials and Methods

The study was approved by the Children's Hospital of Wisconsin Institutional Review Board. A retrospective chart review was conducted of patients aged 0–18 years seen at a paediatric dermatology referral centre from January 2000 to June 2012. The Children's Hospital of Wisconsin paediatric dermatology clinic serves paediatric patients primarily in the Milwaukee, Wisconsin metropolitan area, but also draws referrals from throughout the state of Wisconsin and northern Illinois. Patients were identified by the International Classification of Diseases, 9th revision codes 710.0 (SLE), 695.4 (lupus erythematosus), or 373.34 (discoid lupus erythematosus). Only those children with LE-related cutaneous disease (including LE-specific, LE-nonspecific, or neonatal LE) seen in dermatology clinic were included. Cutaneous LE subtype was determined based on clinical, laboratory, and pathology findings upon review of the record by two of the authors (BZD and YEC). The modified Gilliam criteria were used to classify the subtypes (Table 1).[1,2,6] Puberty status was assigned by age cut-off of 12 years based on United States national averages.[7] This study was conducted in Milwaukee, Wisconsin, United States of America.

Results

Overall epidaemiology and clinical findings: cutaneous lupus erythematosus

Complete demographic, clinical, and laboratory findings are summarized in Table 2. Fifty-three children were included in the study. Of these, 15 patients had neonatal lupus and will be discussed separately. The remaining 38 patients had paediatric cutaneous LE, either LE-specific skin disease, LE-nonspecific skin disease, or both. Seventeen had CCLE, 6 had SCLE, and 13 had ACLE; 3 patients were diagnosed with 2 LE-specific subtypes and 2

patients with 3 or more subtypes. Fifteen children had LE-nonspecific skin disease. Mean follow-up time was 47.8 months (median 38, range 2–138).

Of the 38 paediatric cutaneous LE patients, 44.7% were black, 34.2% were white, 13.2% were Latino, and 7.9% were Asian. Mean age of onset was 11.7 years (median 12.5, range 2–17). Overall, female patients predominated, with a female to male ratio of 2.2:1; however, there was equal sex distribution in children with pre-pubertal disease onset before age 12 (female:male ratio 1:1), and a striking female predominance in post-pubertal disease onset at or after age 12 (female:male ratio 4.5:1). These ratios differed substantially by cutaneous LE subtype. Males developed cutaneous LE earlier than females, with a mean age of 9.8 years for males versus 12.5 years for females.

Laboratory values by cutaneous LE subtype are summarized in Table 3. Laboratory parameters were evaluated at various points during the disease course according to the clinical judgement of the providers. Additionally, not all subjects had laboratory testing performed. Overall, 25 patients (66%) met diagnostic criteria for SLE. Even in the 13 children who did not meet diagnostic criteria for SLE (12 had CCLE, 1 had SCLE), laboratory abnormalities were common; 5 out of 12 had hematologic abnormalities, 1 out of 7 had abnormal urinalysis findings, and 7 out of 12 had an auto-antibody present. Skin biopsy with subsequent histological examination was performed in 21 children, as well as in 1 infant with neonatal LE.

Topical treatments included corticosteroids and calcineurin inhibitors, while systemic agents included corticosteroids, hydroxychloroquine, mycophenolate mofetil, azathioprine, methotrexate, and cyclophosphamide.

Chronic cutaneous lupus erythematosus

Among the 38 patients with cutaneous LE, CCLE was the most common subtype (17 patients, 45%). There were 10 patients with discoid lupus erythematosus (DLE) localized to the head, 4 patients with generalized DLE, 2 patients with lupus panniculitis, and 1 patient each with lupus tumidus and chilblain LE. This group had the earliest age of disease onset, with mean of 9.9 years (median 11, range 2–15). These children had nearly equal female to male sex distribution of 1.1:1, and this group was the most evenly distributed by race. When compared to other cutaneous LE patients, this group was the least likely to have family history of autoimmune disease, with only 23.5% of patients reporting autoimmune disease in either immediate or distant family members. This group also had the lowest rates of laboratory abnormalities in hematologic, urinary, and auto-antibody tests. Four patients with localized DLE and one patient with lupus panniculitis were the only ones in the study to be treated with topical therapies alone.

Out of 17 total patients with CCLE, 5 had concomitant SLE (1 with localized DLE, 2 with generalized DLE, 1 with localized DLE and lupus panniculitis, and 1 with chilblain LE). Four of these patients had either pre-existing SLE or concurrent development of cutaneous and systemic disease. Notably, the only patient in the study observed to progress from skin-limited to systemic disease was a female patient who had chilblain LE diagnosed at age 12

and progressed to SLE at age 16. No patient with DLE progressed to SLE over a mean follow up time of 22.9 months (median 23.5, range 2–56).

Subacute cutaneous lupus erythematosus

SCLE was the least commonly observed subtype, present in only 6 patients (16%). The skin lesions were most often described as being annular (4 subjects) or papulosquamous (2 subjects), but toxic epidermal necrolysis-like lesions were described in one patient who also had annular lesions. Five of the 6 patients (83.3%) had concomitant SLE, and the remaining patient was followed for 50 months without evidence of systemic disease. Distribution of SCLE lesions on the body was more widespread than in other subtypes, as all 6 patients had lesions located on at least one other body area in addition to the head. The mean age of onset was 11.3 years (median 12, range 2–16). This was the only group in which no patient reported immediate family history of autoimmune disease; however, 3 patients (50%) had distant family history. All 6 patients required systemic therapy, and 4 were prescribed topical treatment in addition.

Acute cutaneous lupus erythematosus

Thirteen patients (34%) were seen in the paediatric dermatology clinic for ACLE lesions. Of these, all 13 had previously diagnosed SLE. Two had concomitant CCLE, and 3 had concomitant SCLE. There were 7 patients who also had LE-nonspecific skin disease. Generalized ACLE was the most frequent type (8 patients), followed by localized ACLE (5 patients), and toxic epidermal necrolysis-like ACLE (1 patient). Interestingly, the 1 patient with both generalized and toxic epidermal necrolysis-like ACLE was the same patient who also presented with toxic epidermal necrolysis-like SCLE. Generalized lesions were located on the head (75%), trunk (50%), upper extremities (50%), and/or lower extremities (38%). This group had the highest age at onset, with a mean age of 13.6 years. Female to male ratio was 12:1, strikingly higher than in other LE-specific subtypes. Systemic therapy was necessary in all 13 patients, while topical treatment was used adjunctively in 9 patients.

Lupus erythematosus nonspecific skin disease

Out of the 38 patients presenting with cutaneous LE, 15 (40%) were diagnosed with at least one of the LE-nonspecific skin diseases. All 15 of these patients had SLE, and 8 of them also had LE-specific skin disease. The majority had only 1 LE-nonspecific subtype; however, 3 patients had multiple subtypes. The most common category of LE-nonspecific skin disease was cutaneous vascular disease (9 patients); 3 had small vessel vasculitis, 3 had Degos' like vasculopathy, and the subtypes periungual telangiectasia, livedo reticularis, and Raynaud's phenomenon were each diagnosed in 1 patient. Additional LE-nonspecific subtypes identified were non-scarring alopecia (4), urticaria (3), bullous SLE (2), and erythema multiforme (1).

Neonatal lupus erythematosus

There were 15 patients with neonatal LE, two of whom were sisters. All 15 patients (100%) had lesions on the head, and 6 patients (40%) presented with lesions localized to the head only. Seven patients (47%) had mothers with known autoimmune disease, while 6 patients

(40%) had asymptomatic mothers later found to have serum auto-antibodies. There were 2 patients whose mothers did not undergo autoimmune disease testing. Anti-Ro/SS-A and anti-La/SS-B antibodies were present in 93% and 50% of patients, respectively. Three patients had cytopoenia, 6 patients had hepatitis, and no patients had heart block on electrocardiography.

Discussion

Our study summarizes the demographic and clinical characteristics of paediatric cutaneous lupus erythematosus seen at a paediatric dermatology referral centre over a 12.5 year period. Differences in disease course between childhood-onset and adult-onset SLE have been reported, but similar data specific to cutaneous LE is lacking.[5] This study furthers understanding about cutaneous LE in children and elucidates differences between adult and paediatric patients. The limitations of our study were the small study population, relatively short follow-up time, retrospective nature, and restriction of subjects to those seen in the paediatric dermatology clinic, thus excluding cutaneous LE patients managed by other departments. Even so, this is the largest cohort of paediatric cutaneous LE patients reported to date, and prospective studies are difficult given the rarity of these disorders in children.

In contrast to adult cutaneous LE, where a striking female predominance is seen, we found an equal sex ratio in prepubertal patients diagnosed before age 12. After puberty, cutaneous LE was more likely to occur in females, with a ratio of 4.5:1 in those diagnosed at or after age 12. These numbers are comparable to those previously reported for paediatric SLE, where the sex ratio is equal until puberty when a striking female predominance is then seen.[8] The peak incidence of cutaneous LE at 11–13 years is similar to that seen in childhood SLE and was comparable across cutaneous LE subtypes.[9] Thus, our findings of sex and age distribution for cutaneous LE are different from that of adult cutaneous LE but similar to childhood-onset SLE.

CCLE was the most common subtype in our study, with the majority of these children having DLE. The age of onset at 9.9 years and sex ratio of 1.1:1 seen in our population are similar to other studies.[10,11] A major difference observed in our study compared to the paediatric DLE literature is rate of progression to SLE. Four patients (29%) had SLE at the time of development of DLE, but of the 10 patients with skin-only DLE, none progressed to SLE after a mean follow-up period of 22.9 months. Our rate of previously-diagnosed SLE is similar to the 23.5% of paediatric DLE patients who also met criteria for SLE observed by Sampaio.[12] Other studies of paediatric DLE patients, however, have reported various progression rates of 5.9%, 24%, and 26%.[10–12] In general, these studies suggest that progression rate is higher in paediatric patients than in adult DLE patients, who progress to SLE at a rate of 5–10%.[13] The low progression rate in our study may be attributed to a shorter follow-up time with a mean of only 22.9 months, compared to means of 36 and 49.8 months in the two studies with the highest progression rates.[10,12]

SCLE was uncommonly observed in our study population with just 6 cases over the 12.5 year period. Even though SCLE is uncommon below the waist in adults, many of the children in our study had lesions on their lower extremities.[3,4] This may suggest different

patterns of sun exposure in the paediatric population. Another key point of difference was that the majority of SCLE patients (83%) in our study had concomitant SLE, in contrast to adult literature reports of 50%.[1,14] Since children with SCLE may be more likely than adults to develop SLE, providers must be vigilant in monitoring for evidence of systemic disease and its subsequent complications.

Of all adult and paediatric patients with SLE, the most common LE-specific cutaneous finding is the malar erythema of ACLE, reportedly occurring in approximately 80% of patients.[15] Reports differ as to the prevalence in paediatrics specifically, with some authors reporting higher rates of children with malar rash than adults and others describing lower percentages of 50–74%.[5,9,16] Our study suggests an even lower prevalence, with only 38% of children with SLE having a malar rash; however, not all children with SLE and typical malar erythema would have been referred to dermatology at our institution (and thus did not meet study inclusion criteria). For all ages, a reported 72% of ACLE patients meet criteria for SLE.[14] All 13 ACLE patients in our study met criteria for SLE, suggesting that isolated ACLE without systemic disease is rare in childhood. Again, this highlights the need for a high level of clinical suspicion for SLE when malar erythema is present.

In general, this study corroborates neonatal LE findings from previous studies, with a predominance of head and neck lesions and approximately half of mothers with known autoimmune disease.[15,17] Our female:male ratio of 1.5:1 is somewhat lower than the 3.5:1 reported by Lee.[18] Since maternal auto-antibodies typically clear within 6–9 months of age, infants with neonatal LE are not expected to progress to SLE or cutaneous LE in childhood, although predisposition to development of other autoimmune conditions later in life has been reported.[19,20] No children in our study developed another autoimmune disease, although mean follow-up time was only 8.6 months (median 3, range 0–56). There was one patient in our study who had anti-Smith and anti-RNP antibodies instead of anti-Ro and anti-La antibodies, which has been reported in very few cases.[18,21]

Findings of LE-nonspecific skin lesions are generally similar between our study and previous studies regarding higher incidence of cutaneous vascular disease in children than adults. In all patients with SLE, vasculitis has been reported in 10–20%, and in children this is as high as 42%.[1,9] Similarly, 36% of paediatric SLE patients in our study developed cutaneous vascular disease. Non-scarring alopecia, on the other hand, did not occur as frequently in our study as that reported in the adult literature. This was only seen in 16% of our paediatric SLE patients, compared to 40–70% reported for adult SLE patients.[1,22] This may suggest that non-scarring alopecia results from a more chronic disease course, as seen in adults with longstanding SLE, or is underreported by children and parents. The higher prevalence of vascular complications in children again suggests a more severe disease course than adults.

In this report, we detail the demographics and clinical features of 53 children with cutaneous LE in order to further characterize this uncommon disorder. Important differences between childhood and adult cutaneous LE were noted, providing useful information for practitioners caring for children with these rare diseases. Childhood-onset cutaneous LE may have a stronger association with SLE than adult-onset cutaneous LE. Thus, a vigilant approach to

monitoring paediatric cutaneous LE patients for development of systemic illness is recommended. Further studies documenting longer-term morbidity and mortality outcomes comparing childhood-onset and adult-onset cutaneous LE patients are warranted.

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References

1. Werth VP. Clinical manifestations of cutaneous lupus erythematosus. Autoimmun Rev. 2005; 4:296–302. [PubMed: 15990077]
2. Klein RS, Morganroth PA, Werth VP. Cutaneous lupus and the Cutaneous Lupus Erythematosus Disease Area and Severity Index instrument. Rheum Dis Clin North Am. 2010; 36:33–51. vii. [PubMed: 20202590]
3. Rothfield N, Sontheimer RD, Bernstein M. Lupus erythematosus: systemic and cutaneous manifestations. Clin Dermatol. 2006; 24:348–62. [PubMed: 16966017]
4. Walling HW, Sontheimer RD. Cutaneous lupus erythematosus: issues in diagnosis and treatment. Am J Clin Dermatol. 2009; 10:365–81. [PubMed: 19824738]
5. Livingston B, Bonner A, Pope J. Differences in clinical manifestations between childhood-onset lupus and adult-onset lupus: a meta-analysis. Lupus. 2011; 20:1345–55. [PubMed: 21951943]
6. Gilliam JN, Sontheimer RD. Distinctive cutaneous subsets in the spectrum of lupus erythematosus. J Am Acad Dermatol. 1981; 4:471–5. [PubMed: 7229150]
7. Parent AS, Teilmann G, Juul A, et al. The timing of normal puberty and the age limits of sexual precocity: variations around the world, secular trends, and changes after migration. Endocrine reviews. 2003; 24:668–93. [PubMed: 14570750]
8. Gilliam AE. Skin signs of systemic disease in childhood. Adv Dermatol. 2006; 22:1–30. [PubMed: 17249293]
9. Wananukul S, Watana D, Pongprasit P. Cutaneous manifestations of childhood systemic lupus erythematosus. Pediatr Dermatol. 1998; 15:342–6. [PubMed: 9796581]
10. Moises-Alfaro C, Berron-Perez R, Carrasco-Daza D, et al. Discoid lupus erythematosus in children: clinical, histopathologic, and follow-up features in 27 cases. Pediatr Dermatol. 2003; 20:103–7. [PubMed: 12657003]
11. Van Gysel D, de Waard-van der Spek FB, Oranje AP. Childhood discoid lupus erythematosus: report of five new cases and review of the literature. J Eur Acad Dermatol Venereol. 2002; 16:143–7. [PubMed: 12046818]
12. Sampaio MC, de Oliveira ZN, Machado MC, et al. Discoid lupus erythematosus in children--a retrospective study of 34 patients. Pediatr Dermatol. 2008; 25:163–7. [PubMed: 18429771]
13. Patel P, Werth V. Cutaneous lupus erythematosus: a review. Dermatol Clin. 2002; 20:373–85. v. [PubMed: 12170873]
14. Watanabe T, Tsuchida T. Classification of lupus erythematosus based upon cutaneous manifestations. Dermatological, systemic and laboratory findings in 191 patients. Dermatology. 1995; 190:277–83. [PubMed: 7655105]
15. Kole AK, Ghosh A. Cutaneous manifestations of systemic lupus erythematosus in a tertiary referral center. Indian J Dermatol. 2009; 54:132–6. [PubMed: 20101308]
16. Jones EM, Callen JP. Collagen vascular diseases of childhood. Pediatr Clin North Am. 1991; 38:1019–39. [PubMed: 1870902]
17. Lee LA. The clinical spectrum of neonatal lupus. Arch Dermatol Res. 2009; 301:107–10. [PubMed: 18797891]
18. Lee LA. Neonatal lupus: clinical features and management. Paediatr Drugs. 2004; 6:71–8. [PubMed: 15035648]
19. Bhatt TA, Fatani HA, Mimesh S. Congenital lupus erythematosus. Indian J Dermatol. 2011; 56:734–6. [PubMed: 22345783]

20. Martin V, Lee LA, Askanase AD, et al. Long-term followup of children with neonatal lupus and their unaffected siblings. Arthritis Rheum. 2002; 46:2377–83. [PubMed: 12355485]

21. Provost TT, Watson R, Gammon WR, et al. The neonatal lupus syndrome associated with U1RNP (nRNP) antibodies. N Engl J Med. 1987; 316:1135–8. [PubMed: 3494943]

22. Werth VP, White WL, Sanchez MR, et al. Incidence of alopecia areata in lupus erythematosus. Arch Dermatol. 1992; 128:368–71. [PubMed: 1550369]

What's Known

1. Demographics and clinical course differ between cutaneous lupus erythematosus subtypes in adults.
2. Patterns in demographics and disease course of systemic lupus erythematosus differ between children and adults.
3. Female:male ratio of systemic lupus erythematosus is equal until puberty and then becomes female-predominant.

What's New

1. Demographics and clinical course differ between cutaneous lupus erythematosus subtypes in children as well.
2. Patterns in demographics and disease course of cutaneous lupus erythematosus differ between children and adults.
3. Female:male ratio of cutaneous lupus erythematosus is equal until puberty and then becomes female-predominant.
4. Children are more prone than adults to develop cutaneous lupus erythematosus subtypes that are more strongly associated with systemic disease.

Table 1

Lupus erythematosus (LE) skin lesions, based on the modified Gilliam and vesiculobullous classification[1,2,6]

- **I.** LE-specific skin disease (characterized by interface dermatitis)
  - **A.** Chronic cutaneous LE (CCLE)
    - **1.** Classic discoid LE (DLE)
      - **i.** Localized DLE
      - **ii.** Generalized DLE
    - **2.** Hypertrophic/verrucous DLE
    - **3.** Lupus panniculitis/lupus profundus
    - **4.** Mucosal DLE
      - **i.** Oral DLE
      - **ii.** Conjunctival DLE
      - **iii.** Nasal DLE
      - **iv.** Genital DLE
    - **5.** LE tumidus/papulomucinous LE
    - **6.** Chilblain LE
    - **7.** Lichenoid DLE (LE/lichen planus overlap)
  - **B.** Subacute cutaneous LE (SCLE)
    - **1.** Annular SCLE
    - **2.** Papulosquamous/psoriasiform
    - **3.** Vesiculobullous annular SCLE
    - **4.** TEN-like SCLE
  - **C.** Acute Cutaneous LE (ACLE)
    - **1.** Localized ACLE (malar rash)
    - **2.** Generalized ACLE (morbiliform)
    - **3.** TEN-like ACLE
- **II.** LE-nonspecific skin disease
  - **A.** Cutaneous vascular disease
    - **1.** Small vessel cutaneous leukocytoclastic vasculitis secondary to LE
      - **i.** Dependent palpable purpura
      - **ii.** Urticarial vasculitis
    - **2.** Vasculopathy

i. Degos' disease like lesions
ii. Secondary atrophie blanche

3. Periungual telangiectasia
4. Livedo reticularis
5. Thrombophlebitis
6. Raynaud's phenomenon
7. Erythromelalgia

B. Nonscarring alopecia

1. Lupus hair
2. Telogen effluvium
3. Alopecia areata

C. Sclerodactyly
D. Rheumatoid nodules
E. Calcinosis cutis
F. LE-nonspecific bullous lesions (bullous SLE)
G. Urticaria
H. Papulonodular mucinosis
I. Cutis laxa/anetoderma
J. Acanthosis nigricans
K. Erythema multiforme
L. Leg ulcers
M. Lichen planus

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Table 2

Demographic characteristics, family history of autoimmune disease, lesion distribution, and treatment modalities distributed by lupus erythematosus (LE) subtype

	CCLE	SCLE	ACLE	LE-Nonspecific	Total Childhood Cutaneous LE	Neonatal LE
N	17	6	13	15	38	15
Sex						
Female (%)	9 (52.9)	5 (83.3)	12 (92.3)	10 (66.7)	26 (68.4)	9 (60.0)
Male (%)	8 (47.1)	1 (16.7)	1 (7.7)	5 (33.3)	12 (31.6)	6 (40.0)
Female:male ratio	1.1:1	5:1	12:1	2:1	2.2:1	1.5:1
Race						
Black (%)	6 (35.3)	3 (50.0)	5 (38.5)	8 (53.3)	17 (44.7)	6 (40.0)
Asian (%)	2 (11.8)	0	1 (7.7)	0	3 (7.9)	0
Caucasian (%)	5 (29.4)	3 (50.0)	6 (46.2)	6 (40.0)	13 (34.2)	8 (53.3)
Latino (%)	4 (23.5)	0	1 (7.7)	1 (6.7)	5 (13.2)	0
Unspecified (%)	0	0	0	0	0	1 (6.7)
Mean age at onset (Median, Range)	9.9 years (11, 2–15)	11.3 years (12, 2–16)	13.6 years (13, 10–18)	13.5 years (13, 9–18)	11.6 years (12.5, 2–17)	3.8 weeks (4, 0–8)
Family history						
Immediate (%)	3 (17.6)	0	4 (30.8)	8 (53.3)	10 (26.3)	13 (86.7)
Distant (%)	1 (5.9)	3 (50.0)	5 (38.5)	3 (20.0)	7 (18.4)	0
None (%)	13 (76.5)	3 (50.0)	4 (30.8)	4 (26.7)	21 (55.3)	2 (13.3)
Distribution						
Head (%)	15 (88.2)	6 (100)	11 (84.6)	8 (53.3)	35 (92.1)	15 (100)
Trunk (%)	3 (17.6)	4 (66.7)	4 (30.8)	7 (46.7)	12 (31.6)	6 (40.0)
Upper extremities (%)	7 (41.2)	5 (83.3)	4 (30.8)	10 (66.7)	20 (52.6)	3 (20.0)
Lower extremities (%)	3 (17.6)	4 (66.7)	3 (23.1)	7 (46.7)	10 (26.3)	4 (26.7)
Genitals (%)	0	0	0	1 (6.7)	1 (2.6)	1 (6.7)
Treatment						
Topical (%)	15 (88.2)	4 (66.7)	9 (69.2)	7 (46.7)	26 (68.4)	11 (73.3)
Systemic (%)	12 (70.6)	6 (100)	13 (100)	15 (100)	33 (86.8)	0

LE, lupus erythematosus; CCLE, chronic cutaneous lupus erythematosus; SCLE, subacute cutaneous lupus erythematosus; ACLE, acute cutaneous lupus erythematosus

Table 3

Laboratory abnormalities in lupus erythematosus (LE) patients, distributed by LE-subtype. Values categorized as abnormal if present at any time during follow-up.

	CCLE	SCLE	ACLE	LE-Nonspecific	Neonatal LE	SLE	Non-SLE
Haematologic							
Leukopoenia (%)	6/16 (37.5)	5/6 (83.3)	11/13 (84.6)	11/15 (73.3)	2/12 (16.7)	17/25 (68.0)	4/12 (33.3)
Anaemia (%)	6/16 (37.5)	5/6 (83.3)	8/13 (61.5)	12/15 (80.0)	2/12 (16.7)	19/25 (76.0)	3/12 (25.0)
Thrombocytopoenia (%)	3/16 (18.8)	1/6 (16.7)	4/13 (30.8)	3/15 (20.0)	1/12 (8.3)	5/25 (20.0)	0/12 (0.0)
Hypocomplementaemia (%)	5/10 (50.0)	5/6 (83.3)	12/13 (92.3)	12/15 (80.0)	0/0	22/25 (88.0)	0/6 (0.0)
Urinary							
Proteinuria (%)	4/12 (33.3)	5/5 (100)	10/13 (76.9)	11/14 (78.6)	0/2 (0.0)	18/24 (75.0)	1/7 (14.3)
Haematuria (%)	3/12 (25.0)	3/5 (60.0)	8/13 (61.5)	10/14 (71.4)	0/2 (0.0)	14/24 (58.3)	0/7 (0.0)
Auto-antibodies							
ANA (%)	8/15 (53.3)	4/5 (80.0)	12/12 (100)	14/14 (100)	9/12 (75.0)	22/22 (100)	3/11 (27.3)
Anti-dsDNA (%)	5/13 (38.5)	5/6 (83.3)	11/13 (84.6)	14/15 (93.3)	0/1 (0.0)	22/25 (88.0)	1/9 (11.1)
Anti-Ro/SSA (%)	3/13 (23.1)	4/5 (80.0)	4/11 (36.4)	5/11 (45.5)	14/15 (93.3)	9/20 (45.0)	1/9 (11.1)
Anti-La/SSB (%)	2/13 (15.4)	2/4 (50.0)	1/10 (10.0)	3/10 (30.0)	7/14 (50.0)	4/19 (21.1)	1/9 (11.1)
Anti-Smith (%)	4/13 (30.8)	2/5 (40.0)	7/10 (70.0)	7/11 (63.6)	3/11 (27.3)	13/20 (65.0)	0/9 (0.0)
Anti-snRNP (%)	5/13 (38.5)	2/5 (40.0)	9/11 (81.8)	9/11 (81.8)	2/11 (18.2)	14/20 (70.0)	1/9 (11.1)
Anti-phospholipid (%)	5/9 (55.6)	6/6 (100)	9/10 (90.0)	9/11 (90.9)	1/2 (50.0)	19/20 (95.0)	2/5 (40.0)

LE, lupus erythematosus; CCLE, chronic cutaneous lupus erythematosus; SCLE, subacute cutaneous lupus erythematosus; ACLE, acute cutaneous lupus erythematosus; dsDNA, double-stranded deoxyribonucleic acid; ANA, anti-nuclear antibody; SSA, single-stranded A; SSB, single-stranded B; snRNP, small nuclear ribonucleoprotein.