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Epidemiology of Cutaneous Lupus Erythematosus Among Adults Over Four Decades (1976–2018): A Lupus Midwest Network (LUMEN) Study

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POTENTIAL COMPETING INTERESTS

The authors report no competing interests.

SUPPLEMENTAL ONLINE MATERIAL

Supplemental material can be found online at http://www.mayoclinicproceedings.org. Supplemental material attached to journal articles has not been edited, and the authors take responsibility for the accuracy of all data.

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Abstract

Objective: To characterize the epidemiological trends and mortality of cutaneous lupus erythematosus (CLE) between 1976 and 2018 in Olmsted County, Minnesota.

Patients and Methods: In this retrospective population-based cohort study, all incident and prevalent CLE cases among adult residents in Olmsted County, Minnesota, between January 1, 1976, and December 31, 2018, were identified and categorized by subtype through medical record review using the resources of the Rochester Epidemiology Project.

Results: The overall incidence rate of CLE between 1976 and 2018 was 3.9 (95% CI, 3.4 to 4.5) per 100,000. The incidence of CLE was relatively stable, with no major trend across sexes or age groups. The age- and sex-adjusted prevalence of CLE was 108.9 per 100,000 on January 1, 2015. Mortality in CLE patients was similar to that of the general population, with a standardized mortality ratio of 1.23 (95% CI, 0.88 to 1.66) with no observed trends in mortality over time.

Conclusion: In the past 4 decades, the incidence of CLE remained stable. Patients with CLE have mortality comparable to that of the general population.

Cutaneous lupus erythematosus (CLE) is a chronic heterogeneous autoimmune disease unified through shared histopathologic features. The clinical manifestations are local or generalized skin lesions with varying characteristics that can have complications such as atrophy, scarring alopecia, dyspigmentation with telangiectasias, and photosensitivity.[1] Patients with CLE report impaired quality of life with poor emotional well-being.[2,3]

Cutaneous lupus erythematosus is classified into 3 major subgroups: acute cutaneous lupus erythematosus, subacute cutaneous lupus erythematosus (SCLE), and chronic cutaneous lupus erythematosus (CCLE). Acute cutaneous lupus erythematosus is seen almost exclusively in the setting of systemic lupus erythematosus (SLE), while SCLE and CCLE can exist alone or concomitantly with SLE. Chronic cutaneous lupus erythematosus is further classified into discoid lupus erythematosus (DLE), tumid lupus (TL), chilblain lupus (CHLE), and lupus panniculitis (LP).[1,4,5] The CLE subtypes differ in their morbidity, association with SLE, and clinical characteristics.[6–8]

Cutaneous lupus erythematosus incidence estimates have ranged from 2.7 to 4.4 per 100,000,[7,9–12] while those reporting CCLE ranged from 2.6 to 3.9 per 100,000.[13,14] Although these studies provided a wealth of data regarding the CLE epidemiology, prior literature has several important gaps. First, there are limited contemporary data regarding secular trends in the incidence of the disease that can result from changes in established risk factors such as smoking.[9,13,14] Second, it is unclear whether CLE is associated with increased mortality.[9,15] Third, there is a lack of incidence estimates on CLE subtypes such as TL and CHLE.[9,10,13] Finally, we recently reported a rise in SLE incidence in Olmsted County, Minnesota, likely due to increasing racial and ethnic diversity in its population.

It is possible that the CLE incidence might be affected by changes in the population demographics.[16]

We aimed to investigate the incidence, prevalence, and mortality of CLE and its trends between 1976 and 2018 in Olmsted County, Minnesota.

PATIENTS AND METHODS

Study Design

The Lupus Midwest Network (LUMEN) is a population-based CLE and SLE registry that utilizes the Rochester Epidemiology Project (REP) resources, a medical records linkage system of the population of Olmsted County, Minnesota. The REP is a well-suited tool for investigating the epidemiology of CLE because comprehensive medical records for all residents of Olmsted County, Minnesota, seeking medical care are available.[17] The characteristics, strengths, and generalizability of the REP have been described elsewhere.[18,19]

Case Identification and Capture

We identified all patients in Olmsted County who had an SLE- or CLE-related diagnosis code and/or positive test result for the following laboratory measures between January 1, 1976, and December 31, 2018: antinuclear antibodies, anti–double-stranded DNA, anti-Sm, complement (C3, C4), anticardiolipin (IgG/IgM/IgA), and anti–β_2-glycoprotein 1 (IgG/IgM/IgA). Medical records, including clinic notes, pathology reports, and medical photographs, were reviewed. Data abstractors were extensively trained until each abstractor achieved 95% agreement with the senior author for all elements. Audits of 10% random samples of the abstracted cases were performed, and retraining was done as needed to maintain the 95% agreement.

We included patients with SCLE and CCLE with or without a preceding or concomitant (within 3 months of incidence date) diagnosis of SLE. Patients with isolated acute cutaneous lupus erythematosus were not included because it is almost exclusively seen in the setting of SLE. We used the definitions and keywords of Drenkard et al[13] to guide the screening process. An incident case had to meet the definition of CLE based on the modified Gilliam classification scheme classifying CLE into SCLE or the following CCLE sub- types:DLE, LP, TL, or CHLE.[1,4,5,20] Medical records for patients with an uncertain diagnosis or subtype were reviewed, confirmed, categorized by subtype by a CLE expert dermatologist (M.D.), or excluded. The incidence date was the date of CLE diagnosis in the medical records and an incident case patient had to reside in Olmsted County for a year before the diagnosis of CLE.

We considered the first diagnosed subtype as incident for patients who had more than one CLE subtype. Incidence estimates for CLE, SCLE, CCLE, and CCLE subtype DLE were calculated for the period January 1, 1976, through December 31, 2018. Incidence estimates for the TL CCLE subtype were calculated for the period January 1, 2000, through December 31, 2018, because of its recent characterization as a clinical subtype.[21] Those with CCLE subtypes CHLE and LP had insufficient numbers for separate subtype estimates.

A prevalent case was defined as an individual who met our case definition for CLE and was a resident of Olmsted County on January 1, 2015. An SLE diagnosis was reported if a patient met the European League Against Rheumatism/American College of Rheumatology criteria[22,23] before or within 3 months after the CLE diagnosis date.

Statistical Analyses

Descriptive statistics (means, standard deviations, percentages) summarize continuous and categorical data. Age- and sex-specific incidence rates were calculated per 100,000 population for CLE overall and for each subtype by using the number of adult (age ≥ 18 years) incident cases as the numerator and adult (age ≥ 18 years) population counts from the REP census as the denominator.[17] Overall incidence rates were age- and/or sex-adjusted to the US total population in 2000 (unadjusted rates reported in Supplemental Tables 1 and 2, available online at http://www.mayoclinicproceedings.org). Trends in incidence rates were examined using Poisson regression methods with smoothing splines for age and calendar year. The CLE point prevalence per 100,000 was determined using the number of prevalent cases on January 1, 2015, as the numerator and the Olmsted County population based on the REP census on January 1, 2015, as the denominator. To compute 95% CIs for incidence and prevalence rates, it was assumed that the number of cases followed a Poisson distribution. Mortality rates following CLE diagnosis were estimated using Kaplan-Meier methods and were compared with the expected survival rates in the Minnesota population. Patients in whom SLE developed before or on the date of CLE diagnosis were excluded from the survival analysis. Patients in whom SLE developed after CLE were censored at the time of SLE diagnosis. The standardized mortality ratio (SMR) was estimated as the ratio of the observed and expected number of deaths. Trends in SMR over time were examined using Poisson regression models. The 95% CIs for the SMR were calculated assuming that the expected rates were fixed and the observed rates followed a Poisson distribution.

Statistical analyses were performed using SAS statistical software, version 9.4M6 (SAS Institute) and R version 4.0.3 (R Foundation for Statistical Computing). The study was approved by the institutional review boards of Mayo Clinic and Olmsted Medical Center (19-000255 and 003-OMC-19, approval date February 5, 2019).

RESULTS

Demographic and Clinical Characteristics of the Incident and Prevalent CLE Cohorts

Our initial screening criteria identified 3292 patients, and 254 CLE cases were identified as possible CLE. Further review by an expert dermatologist (M.D.) excluded 50 cases for failing to meet our case definition due to insufficient available data or atypical clinical or pathologic features. Three patients were less than 18 years of age and were excluded from further analyses. Overall, our study identified an incident cohort of 201 adult cases of CLE diagnosed between January 1, 1976, and December 31, 2018, among Olmsted County residents (Supplemental Figure 1, available online at http://www.mayoclinicproceedings.org). The mean ± SD age was 50±17 years; 146 patients (73%) were female. Of the 201 patients, 174 (87%) had a skin biopsy; 6 (3%) had prior or concomitant SLE. Our cohort's racial and ethnic distribution was 85% White (170 patients),

2% Hispanic (4), 7% Asian (14), and 6% Black (12). As noted in the Table, the racial makeup of the incident cohort changed throughout our study period, with an increasing representation of Black and Asian patients in recent decades. The subtype distribution of the entire CLE incident cohort was 50 cases of SCLE (25%) and 151 cases of CCLE (75%). The CCLE distribution was as follows: 125 DLE cases (62%), 3 LP (1%), 5 CHLE (2%), and 18 TL (9%). The proportion of DLE cases decreased throughout our study period, while the SCLE and TL case proportions increased. The demographic and clinical characteristics of the prevalent cohort were similar to the incidence cohort (Table).

Incidence Rates of CLE and CLE Subtypes Between 1976 and 2018

The overall incidence rate of CLE between 1976 and 2018 was 3.9 (95% CI, 3.4 to 4.5) per 100,000 person-years. The sex-specific incidence rates were 5.3 (95% CI, 4.5 to 6.2) in females and 2.3 (95% CI, 1.7 to 3.0) in males, respectively. The overall incidence rate ranged from 3.0 to 4.5 in the periods between 1976 and 2018(Supplemental Table 3, available online at http://www.mayoclinicproceedings.org).

There were no clear trends in the incidence rates, but there was a cyclical pattern with a notable increase in CLE incidence for females from 1985 to 1990 and males from 1982 to 1988, with a subsequent return to average rates (Figure 1). The same incidence trend was observed in all age groups (Figure 2). There was no statistically significant change in age at diagnosis throughout the past 4 decades (P=.32; Figure 3).

The overall SCLE incidence rate between 1976 and 2018 was 1.0 (95% CI, 0.7 to 1.2) per 100,000, with a 3 times higher incidence rate in females compared with males (Supplemental Table 4, available online at http://www.mayoclinicproceedings.org). In the same period, the CCLE overall incidence rate was 2.9 (95% CI, 2.5 to 3.4) with an incidence rate twice as high in females than in males. For DLE, the overall incidence was 2.5 (95% CI, 2.0 to 2.9) and was twice as frequent in women. Our estimate for TL overall incidence rate between 2000 and 2018 was 0.6 (95% CI, 0.3 to 0.9) with similar incidence across sexes. The overall CHLE incidence rate between 2000 and 2018 was calculated at 0.1 (95% CI, 0.0 to 0.3). The data for long-term overall incidence did not reveal trends in the CLE subtypes. Overall, CCLE incidence between 1976 and 2018 remained stable due to increasing TL incidence offsetting the decreasing DLE incidence (Supplemental Table 4).

Age-specific incidence rates revealed different trends in patients with SCLE compared with other CLE subgroups. The incidence of SCLE in patients younger than 50 years was 0.5 per 100,000 and increased steadily, with age reaching 4.2 in patients older than 80 years. For both CCLE and DLE, there was a peak incidence in the fifth decade of life with a declining incidence in elderly patients (Supplemental Figure 2, available online at http://www.mayoclinicproceedings.org).

Prevalence of CLE

There were 170 Olmsted County residents with prevalent CLE on January 1, 2015. The overall estimated point prevalence per 100,000 was 108.9 (95% CI, 92.2 to 125.6). Prevalence was 146.5 (95% CI, 119.7 to 173.3) for females and 67.8 (95% CI, 48.7 to 86.9) for males (Supplemental Table 3).

Mortality of CLE

A total of 192 incident CLE cases were included in the survival analysis. From 1976 to 2018, there were 41 deaths. The mean ± SD length of follow-up was 12.1±9.1 years; 33.45 deaths were expected based on Minnesota life tables. The most common causes of death were cardiovascular (34% of deaths [14 of 41]) and infection (22% [9 of 41]). The SMR for CLE between 1976 and 2018 was calculated at 1.23 (95% CI, 0.88 to 1.66). There was no significant change in SMR across the past 4 decades, with SMRs at 1.81 (95% CI, 0.99 to 3.04; P = 02) in 1976–1988, 1.49 (95% CI, 0.83 to 2.45; P=.12) in 1989–1998, 0.53 (95% CI. 0.19 to 1.15; P=.11) in 1999–2008, and 1.4 (95% CI, 0.51 to 3.04; P=.41) in 2009–2018. As shown in Figure 4, there was no difference between observed and expected survival in patients with CLE.

DISCUSSION

In this population-based study, we investigated the epidemiology of CLE in a welldefined region in the United States. Our results revealed no secular incidence trends except a notable increase in CLE incidence within the 1985–1990 period. The overall incidence rate of CLE between 1976 and 2018 was 3.9 per 100,000. Our data confirmed that the clinical characteristics of the disease have been changing, with a declining proportion of DLE cases and an increasing proportion of SCLE and TL cases in our incidence cohort through recent decades. We provided the first estimates of the incidence of TL and CHLE in the literature, which were 0.6 and 0.1 per 100,000, respectively. Our study did not find increased mortality associated with CLE.

Our findings are difficult to compare with those of previous population-based studies,[9,13] which relied on different methodologies and case definitions or were conducted on study populations with different racial and socioeconomic backgrounds. Our data provide an updated estimate for the overall incidence rate of CLE in this predominantly White US population. The incidence rate in the prior study in Olmsted County between 1965 and 2005 was similar to our updated estimate (4.3 vs 3.9 per 100,000).[10] Although a study from Sweden reported a rate similar to ours (4 per 100,000),[7] another one from Denmark had a lower incidence (2.7 per 100,000)[11]; both relied on *International Classification of Diseases* coding information for case ascertainment, which can lead to overestimation of cases compared with ascertainment by medical record review. Our estimate is also close to the recently reported estimate from South Korea at 4.3 per 100,000 despite differences in racial backgrounds.[12] The specific incidence estimates of DLE and CCLE for Olmsted County are lower than previously reported estimates from the Atlanta metro area, which has a higher proportion of Blacks in the population. Although the CCLE incidence in Olmsted County was lower than in the Atlanta metro,[13] it was higher than reported in the predominantly Black population of French Guiana (2.6 per 100,000), which has more limited health care access.[14] In addition, the estimates from Denmark, Sweden, South Korea, and Olmsted County included CLE patients with SLE.[7,9–12] Conversely, the studies from Atlanta and French Guiana excluded CLE patients with concomitant SLE.[13,14]

Our findings regarding no increased mortality in patients with CLE differ from those of the prior study from Denmark, which reported increased all-cause mortality in patients

with CLE.[15] This discrepancy could be due to several potential reasons. The Danish study had a large sample size and might have detected a difference due to higher statistical power. Also, it only included hospital-based *International Classification of Diseases*, *Tenth Revision* coding for case capture. This factor may lead to the inclusion of more severe cases because milder cases are less likely to be followed up by the hospital-based health care professional; also, code-based studies are at risk for classification bias. These codes have not been validated, thus classifying CLE as SLE. Conversely, our study included both hospital and community-based health care professional in its geographic region, and all cases were confirmed. Our results are similar to those of a prior Rochester, Minnesota, study[9] but expand the findings regarding no increased mortality with a larger sample size and study period (62 vs 192 patients and 12 vs 43 years, respectively).

Our findings on the secular trends of CLE reveal a stable incidence of the disease, with a notable peak observed in the 1985 to 1990 period. Smoking has been associated with CLE; however, although the smoking rates in the United States have declined in recent years, we did not observe a concomitant decline in CLE incidence. Based on the Minnesota Heart Survey study,[24] the smoking rates in Minnesota, where our study population is located, declined significantly from 32.9% in men and 31.8% in women in 1980–1982 to 20.6% in men and 19.5% in women in 2000–2002. However, the CLE rates continued to rise from 1980 to 1990 when smoking rates were already declining. It is possible that the accumulated nature of cigarette smoking in the 1960s and 1970s contributed to persistently high CLE rates observed in later decades. It is unclear why the peak CLE incidence was observed in the late 1980s.

Our group recently reported an increase in the SLE incidence in Olmsted County during the same study period of this study, likely explained by the increased racial and ethnic diversity of Olmsted County in the recent decades.[16] We did not observe the same trends in CLE. The Atlanta study reported a higher CLE incidence in Black than in White patients with SLE. During the study period, the racial diversity in the county was increased not only in the proportion of Black individuals but also in Asians and Hispanics. The comparative CLE incidence in other racial and ethnic groups is unknown. Further studies are needed to better characterize CLE risk in people with different racial and ethnic backgrounds.

The CLE subtype-specific data underscore the heterogeneous nature of CLE with respect to age and sex associations and reveals changes in the clinical characteristics of the disease throughout the past 4 decades. Increased clinical awareness can explain an increase in the proportion of SCLE and TL cases. Conversely, the relative decrease in DLE incidence may be due to the classification of patients into newly defined subtypes. Historically, the term *discoid LE* was often used generically to refer to all skin lesions that have some form of association with lupus.[25] Clinicians may be using more modern terms in their current clinical descriptions.

This study has several limitations. Our study population was predominantly White, and thus, our findings may not be generalizable to more diverse populations. Our estimates were based on the clinical diagnoses documented in the medical records. However, we had the availability of complete medical records from all health care professionals in the area.

The strengths of this study include virtually complete population case ascertainment for 43 years, detailed clinical characterization, and separate incidence estimates for different CLE subtypes.

CONCLUSION

In this study, we observed evolving clinical characteristics of CLE with a stable incidence in this US population. Patients with CLE have mortality rates comparable to those of the general population.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Abbreviations and Acronyms:

CCLE	chronic cutaneous lupus erythematosus
CHLE	chilblain lupus
CLE	cutaneous lupus erythematosus
DLE	discoid lupus erythematosus
LP	lupus panniculitis
REP	Rochester Epidemiology Project
SCLE	subacute cutaneous lupus erythematosus
SLE	systemic lupus erythematosus
SMR	standardized mortality ratio
TL	tumid lupus

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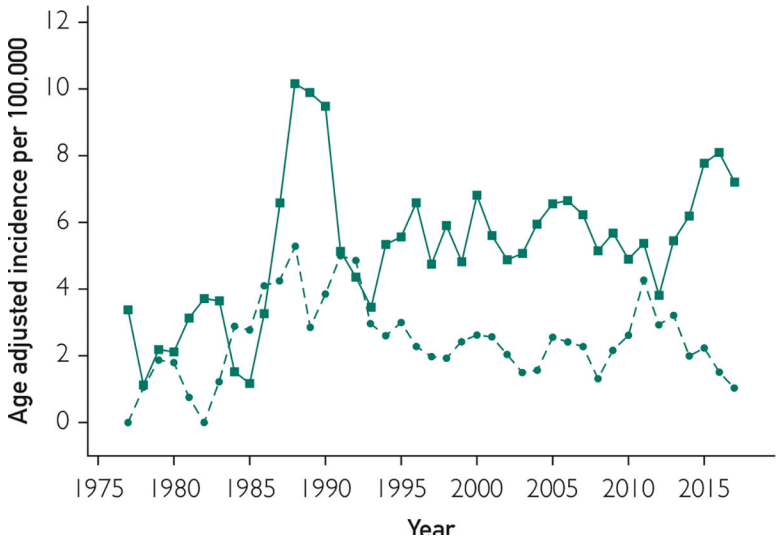

FIGURE 1.

Age-adjusted incidence rates of cutaneous lupus erythematosus in adults (≥18 years) in Olmsted County, Minnesota, stratified by sex. Based on the modified Gilliam classification scheme per 100,000 person-years between January 1, 1976, and December 31, 2018, in females (solid line) and males (dashed line). Incidence rates were age-adjusted to the 2000 US population. Incidence rates were calculated using the number of incident cases as the numerator and population estimates for Olmsted County based on the decennial census counts (1980, 1990, 2000, 2010) as the denominator, with linear interpolation used to estimate population size for intercensal years

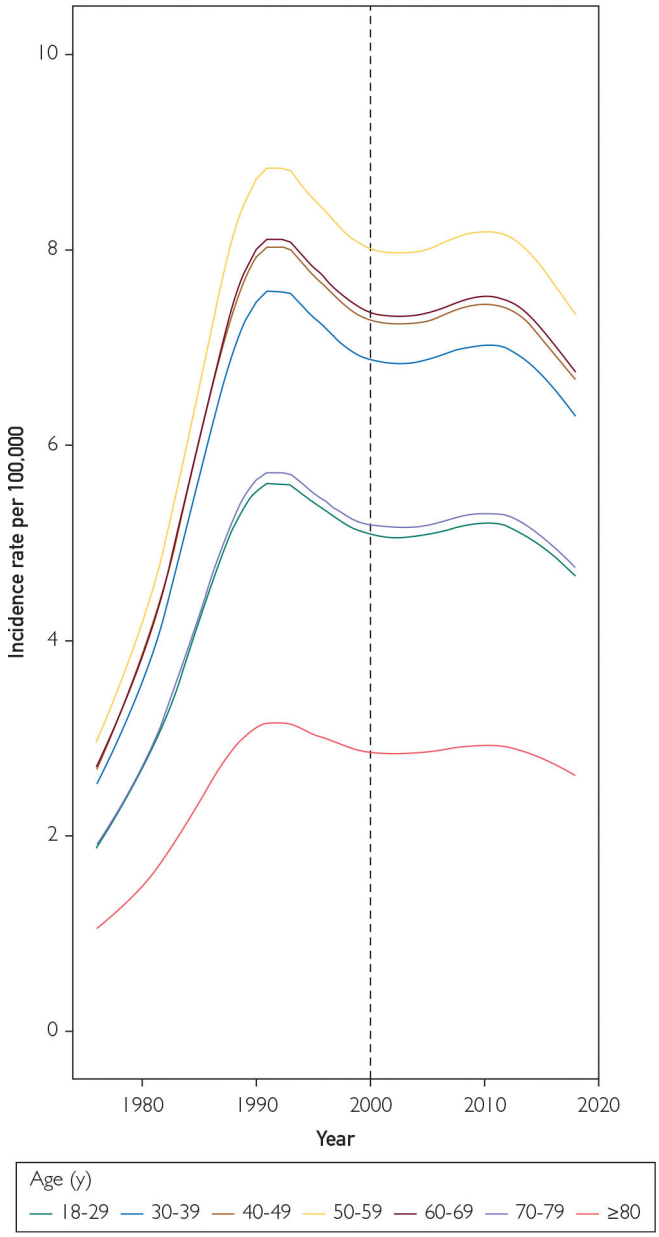

FIGURE 2.

Trends in incidence of cutaneous lupus erythematosus according to age group in adults (≥18 years) in Olmsted County, Minnesota. Based on the modified Gilliam classification scheme per 100,000 person-years between January 1, 1976, and December 31, 2018. Incidence rates were age-adjusted to the 2000 US population. Incidence rates were calculated using the number of incident cases as the numerator and population estimates for Olmsted County based on the decennial census counts (1980, 1990, 2000, 2010) as the denominator, with linear interpolation used to estimate population size for intercensal years.

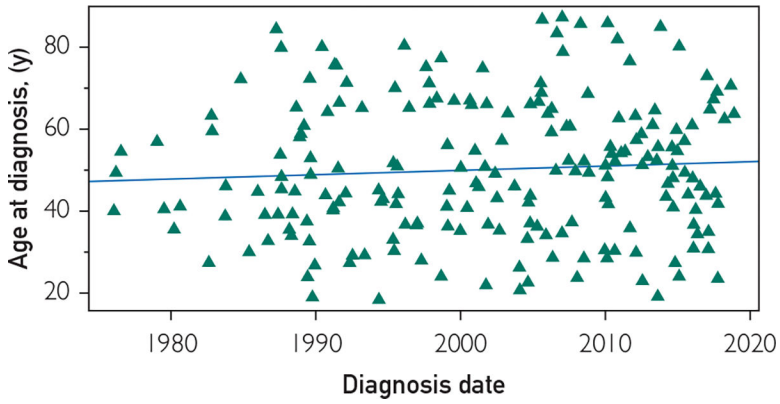

FIGURE 3.

Trends in age at diagnosis of incident cutaneous lupus erythematosus in adults (≥18 years) in Olmsted County, Minnesota, from 1976 to 2018. Cutaneous lupus erythematosus cases were defined according to the modified Gilliam classification scheme. All incident cases were residents of Olmsted County. Age at diagnosis was defined as the date when the diagnosis was documented in the medical record.

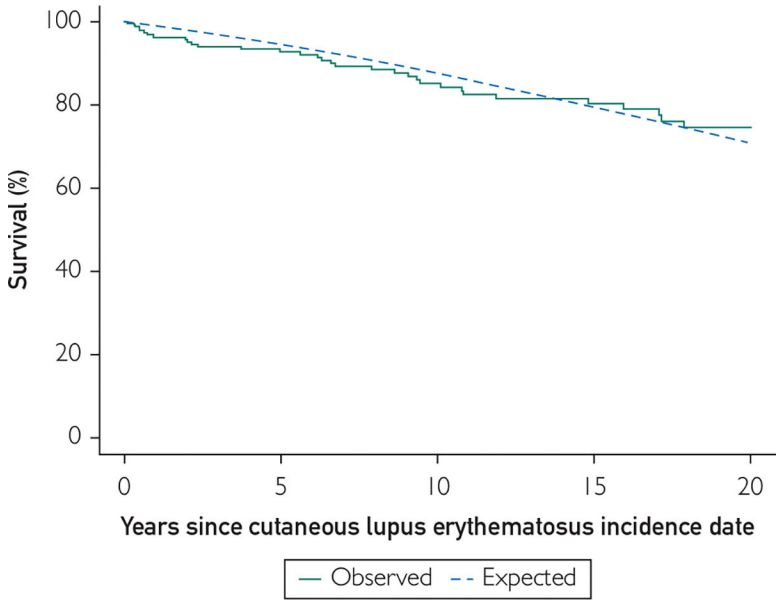

FIGURE 4.

Observed vs expected survival in incident cutaneous lupus erythematosus in adult (≥18 years) patients in Olmsted County, Minnesota, from 1976 to 2018.

TABLE.

Demographic and Clinical Characteristics of the Incident (1976-2018) and Prevalent (2015) Cohorts of Patients With Cutaneous Lupus Erythematosus Among Adults (≥18 Years), Olmsted County, Minnesota[a,b,c]

	Incident cohort, n (%) except for age					Prevalent cohort
Variable	**1976–1988 (n=30)**	**1989–1998 (n=48)**	**1999–2008 (n=58)**	**2009–2018 (n=65)**	**1976–2018 (N=201)**	**01/01/2015 (N=170)**
Sex						
Female	20 (67)	34(71)	44 (76)	48 (74)	146 (73)	120 (71)
Male	10(33)	14 (29)	14 (24)	17(26)	55 (27)	50 (29)
Age (y), mean±SD	48±14	49± 18	51 ± 17	50±16	50±17	45± 16
Race/ethnicity						
Hispanic	1 (3)	0(0)	3 (5)	0(0)	4(2)	2(1)
Asian	1 (3)	2(4)	6 (10)	5 (8)	14(7)	11 (6)
Black	0(0)	2(4)	4(7)	6(9)	12(6)	11 (6)
White	28 (93)	44 (92)	45 (78)	53 (82)	170 (85)	137 (81)
Other/unknown	0(0)	0(0)	0(0)	1 (2)	1 (0)	9 (5)
CLE overall and by subtypes						
CLE overall	30 (100)	48 (100)	58 (100)	65 (100)	201 (100)	170 (100)
SCLE	1 (3)	14 (29)	18 (31)	17(26)	50 (25)	40 (24)
CCLE	29 (97)	34(71)	40 (69)	48 (74)	151 (75)	130 (76)
DLE	29 (97)	33 (69)	28 (48)	35 (54)	125 (62)	109 (64)
LP	0(0)	1 (2)	0(0)	2(3)	3 (1)	1 (1)
CHLE	0(0)	0(0)	2(3)	3 (5)	5 (2)	6 (4)
TL	0(0)	0(0)	10(17)	8 (12)	18 (9)	14(8)

[a]CCLE, chronic cutaneous lupus erythematosus; CHLE, chilblain lupus erythematosus; CLE, cutaneous lupus erythematosus; DLE, discoid lupus erythematosus; LP, lupus panniculitis; SCLE, subacute cutaneous lupus erythematosus; TL, tumid lupus.

[b]Data are presented as No. (percentage) of patients unless indicated otherwise.

[c]Cutaneous lupus erythematosus cases were defined and subtyped according to the modified Gilliam classification scheme. All cases were reviewed through medical record review, and uncertain cases were confirmed by a dermatologist with expertise in CLE. The CLE subtype is defined as the subtype of the incident case in patients with multiple CLE subtypes. All incident cases were residents of Olmsted County, Minnesota. Cases were assigned to 1 of the 5 mutually exclusive race/ethnicity groups based on the self-reported information in the medical records: Hispanic, Asian, White, Black, and Other.