Linear morphea: Clinical characteristics, disease course, and treatment of the Morphea in Adults and Children cohort

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Background: Prospective, longitudinal studies examining the features of linear morphea are limited.

Objective: To utilize the Morphea in Adults and Children cohort to determine clinical characteristics, impact on life quality, and disease course of linear morphea in a prospective, longitudinal manner.

Methods: Characteristics of linear morphea versus other subtypes were compared in a cross-sectional manner. Next, linear morphea participants were examined in depth over a 3-year period.

Results: Linear morphea was the most common morphea subtype (50.1%, 291/581) in the cohort. Deep involvement was more common in linear (64.3%, 187/291) than other morphea subtypes. Linear morphea participants with deep involvement were more likely to have a limitation in range of motion (28.6%, 55/192) than those without (11.1%, 11/99, $P < .001$). Adult-onset disease occurred in 32.6% (95/291) of those with linear morphea. Frequency of deep involvement was similar between pediatric (66.8%, 131/196) and adult-onset linear morphea (58.9%, 56/95, $P = .19$). Quality of life and disease activity scores improved over time, while damage stabilized with treatment.

Limitations: Results of the study are associative, and the University of Texas Southwestern Medical Center is a tertiary referral center.

Conclusion: A substantial number of linear morphea patients have adult-onset disease. In all age groups, linear morphea with deep involvement was associated with functional limitations. (J Am Acad Dermatol 2019;80:1664-70.)

Key words: en coup de sabre; linear morphea; localized scleroderma; MAC cohort; Morphea in Adults and Children cohort; Parry-Romberg syndrome; quality of life.

Morphea, or localized scleroderma, is an inflammatory disorder of the skin that can affect soft tissue with devastating functional and cosmetic impairment. Morphea is divided into 4 subtypes: linear, generalized, plaque (circumscribed), and mixed.[1] Linear morphea is the predominant subtype in children and is most frequently associated with musculoskeletal, cosmetic, and neurologic irregularities.[2-4]

To date, most studies of linear morphea are limited by focus on cases with pediatric onset, with few participants having adult-onset disease.[1,5-10]

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Also, the largest studies examining linear morphea have relied on retrospective data or were compiled from numerous sites in which multiple examiners reported results. This can cause variation in how patients are characterized and assessed. With few longitudinal studies available to date, our understanding of the clinical characteristics of patients with linear morphea, as well as the clinical course of patients on standard of care therapies, is limited. As a result, little is known about how patients with linear morphea compare with other morphea subtypes, the clinical characteristics of adults with linear morphea, or long-term patient outcomes. This knowledge gap limits the clinical evaluation and care of these patients and inhibits planning for clinical trials.

CAPSULE SUMMARY

- Linear morphea, which is most common in children, has not been systematically compared with the other morphea subtypes in children or adults.
- Deep involvement is most common in the linear subtype and signals risk for functional limitations warranting more aggressive treatment. Appropriate treatment decreases activity and stabilizes damage.

To address this knowledge gap, we utilized the Morphea in Adults and Children (MAC) cohort to conduct a 2-part study to determine how linear morphea differs from other morphea subtypes and examine the demographic and clinical features of linear morphea in greater detail.

Study design

First, we performed a cross-sectional analysis of all participants in the MAC cohort who met inclusion criteria to compare linear morphea with other morphea subtypes. Next, we prospectively examined patients with linear morphea for 3 years to determine changes in clinical scores and quality of life.

Participants

Details of the MAC cohort have been described previously.[11] MAC cohort participants with a diagnosis of morphea during 2007-2017 as confirmed by 1 author (Dr Jacobe) based on clinical examination (and pathologic examination when the clinical diagnosis was in question) were included in the study (N = 581). This study was approved by the institutional review board of the University of Texas Southwestern Medical Center. Written informed consent was provided by all participants or guardians.

Variables

At each study visit, a dermatologist (Dr Jacobe) examined patients and assigned a subtype as delineated by Zulian and Laxer[1] and physical examination scores. The Localized Scleroderma Cutaneous Assessment Tool (LoSCAT) component scores for activity (modified Localized Scleroderma Skin Severity Index [mLoSSI]) and damage (Localized Scleroderma Skin Damage Index [LoSDI]), along with those of the Physician Global Assessment of Disease Activity (PGA-A) and Damage (PGA-D) were used.[12-14] Remission was defined as mLoSSI score change from ≥1 to 0 during the study period, while recurrence was defined as mLoSSI >0 after previous remission. Demographic and clinical data, photographs, treatment information, Dermatology Life Quality Index (DLQI), and Children's Dermatology Life Quality Index (CDLQI) scores were collected. Dermal involvement was defined by the absence of involvement of the subcutis or beyond. Deep lesions are defined as extension into or beyond the subcutaneous fat as confirmed by pathology, imaging, or clinical examination.

Statistical analyses

Demographic and clinical variables were reported as frequencies or medians and interquartile ranges (IQRs). Chi-squared and Fisher's exact tests were used to compare dichotomous and categorical variables. Kruskal-Wallis and Mann-Whitney U tests were used to compare clinical characteristics between morphea subtypes. Wilcoxon matched-pairs signed rank tests were performed to compare quality of life and physician reported scores at baseline to those at annual visits for a 3-year follow-up period. A 2-sided P value <.05 was set for statistical significance. All statistical analyses were performed with Graphpad Prism v7 (La Jolla, California).

RESULTS

Table I shows the demographic and clinical characteristics of all participants. Linear morphea was the most common subtype (50.1%, 291/581) in the MAC cohort. The proportion of male patients was highest in linear morphea than other subtypes (24.7%, 72/291, P = .001; Table I). Pediatric-onset disease (<18 years) was observed in 40.1% (233/581) of all participants.

Comparison of linear versus other morphea subtypes

Linear morphea participants had lower baseline disease activity than the generalized morphea group

Abbreviations used:

CDLQI: Children's Dermatology Life Quality Index
DLQI: Dermatology Life Quality Index
IQR: interquartile range
MAC: Morphea in Adults and Children cohort
LoSCAT: Localized Scleroderma Cutaneous Assessment Tool
mLoSSI: modified Localized Scleroderma Skin Severity Index
LoSDI: Localized Scleroderma Damage Index
PGA-A: Physician Global Assessment of Disease Activity
PGA-D: Physician Global Assessment of Disease Damage

as measured by the mLoSSI and PGA-A (Table I). Median LoSDI scores were lower for participants with linear morphea (10, IQR 5-16) than the generalized subtype (19.5, IQR 10-29.3). In contrast, PGA-D scores were highest in participants with linear morphea ($P < .001$).

Deep lesions were more frequent in linear morphea (64.3%, 187/291) than the other subtypes ($P < .001$). Genital involvement and overlying lichen sclerosus were less frequent in the linear subtype (0.3%, 1/291) than the generalized subtype (8.8%, 16/182). Limitations in range of motion and joint contractures were found in similar frequencies in participants with linear and generalized subtypes. Neurologic manifestations, including headaches and seizures, were most commonly reported in the linear morphea group.

Supplemental Table I (available at http://www.jaad.org) details the treatments used in linear morphea participants with adult-onset and pediatric onset disease recorded at the initial study visit. Differences in utilization of systemic corticosteroids were identified among morphea subtypes: oral

Table I. Demographic and clinical characteristics of morphea participants by clinical subtype

Characteristic	Linear, N = 291	Generalized, N = 182	Plaque, N = 79	Mixed, N = 29	*P* value
Age at enrollment, y, median (IQR)	19 (12-34.5)	58 (46-65)	43 (31-57)	37 (27-48)	<.001
No. with pediatric/adult onset*	196/95	18/164	9/70	10/19	<.001
Sex, n (%)					.001
Male	72 (24.7)	28 (15.4)	6 (7.6)	3 (10.3)	
Female	219 (75.3)	154 (84.6)	73 (92.4)	26 (89.7)	
Race/Hispanic origin, n (%)					.11
White non-Hispanic	202 (69.4)	145 (79.7)	58 (73.4)	19 (65.5)	
Black	5 (1.7)	8 (4.4)	7 (8.9)	4 (13.8)	
White Hispanic	54 (18.6)	23 (12.6)	7 (8.9)	4 (13.8)	
Asian and Pacific Islander	13 (4.5)	4 (2.2)	3 (3.8)	2 (6.9)	
Other	17 (5.8)	2 (1.1)	4 (5.1)	0 (0)	
Clinical feature, n (%)					
Deep involvement	187 (64.3)	59 (32.4)	36 (45.6)	12 (41.4)	<.001
Dermal involvement	71 (24.4)	46 (25.3)	29 (36.7)	8 (27.6)	.15
Lichen sclerosus overlap	24 (8.2)	76 (41.8)	9 (11.4)	2 (6.9)	<.001
Genital involvement	1 (0.3)	16 (8.8)	2 (2.5)	0 (0)	<.001
Hair loss	62 (21.3)	18 (9.9)	3 (3.8)	0 (0)	<.001
Functional abnormalities, n (%)					
Limited range of motion	66 (22.7)	34 (18.7)	2 (2.5)	1 (3.4)	<.001
Joint contracture	3 (1)	3 (1.6)	0 (0)	0 (0)	.62
Limb length discrepancy	13 (4.5)	0 (0)	0 (0)	0 (0)	.004
Neurologic symptoms, n (%)					
Headache	66 (22.7)	20 (11.0)	18 (22.8)	3 (10.3)	.001
Seizure	8 (2.7)	2 (1.1)	1 (1.3)	0 (0)	.48
LoSCAT component score, median (IQR)	n = 226	n = 134	n = 57	n = 25	
mLoSSI	3 (0-7)	17 (6.8-32)	3 (0-5.5)	0 (0-2)	<.001
LoSDI	10 (5-16)	19.5 (10-29.3)	6 (4-8.5)	6 (4-11)	<.001
	n = 225	n = 131	n = 57	n = 25	
PGA-A	10 (0-32.5)	35 (17-70)	10 (0-30)	0 (0-20)	<.001
PGA-D	30 (20-50)	20 (10-40)	10 (5-20)	15 (10-20)	<.001

IQR, Interquartile range; *LoSCAT*, Localized Scleroderma Cutaneous Assessment Tool; *LoSDI*, Localized Scleroderma Skin Damage Index; *mLoSSI*, modified Localized Scleroderma Skin Severity Index; *PGA-A*, Physician Global Assessment of Disease Activity; *PGA-D*, Physician Global Assessment of Disease Damage.

*Pediatric onset was defined as onset of disease at <18 years of age.

Table II. Clinical and demographic characteristics of linear morphea participants by disease onset

Feature	All linear morphea participants, N = 291	Pediatric onset, n = 196	Adult onset, n = 95	*P* value
Age at onset, y, median (IQR)	12.6 (7-23.9)	8.9 (5.4-12.7)	31.1 (24.3-40)	<.001
Multiple lesions, n (%)	154 (52.9)	112 (57.1)	42 (44.2)	.04
Any head or neck involvement, n (%)	109 (37.5)	68 (34.7)	41 (43.2)	.16
En coup de sabre, n (%)	71 (24.4)	44 (22.4)	27 (28.4)	.27
Parry-Romberg or hemifacial atrophy, n (%)	34 (11.7)	21 (10.7)	13 (13.7)	.46
Any trunk or extremity involvement, n (%)	217 (74.6)	152 (77.6)	65 (68.4)	.09
Trunk only, n (%)	28 (9.6)	14 (7.1)	14 (14.7)	.04
Extremities only, n (%)	66 (22.7)	49 (25.0)	17 (17.9)	.17
Above and below neck, n (%)	36 (12.4)	24 (12.2)	12 (12.6)	.93
Laterality, n (%)*	n = 290	n = 196	n = 94	
Left	82 (28.3)	54 (27.6)	28 (29.8)	.69
Right	73 (25.2)	47 (24.0)	26 (27.7)	.50
Bilateral or midline	135 (46.6)	95 (48.5)	40 (42.6)	.34
Deep involvement, n (%)	187 (64.3)	131 (66.8)	56 (58.9)	.19
Neurologic symptom, n (%)				
Headache†	31 (28.4)	21 (30.9)	10 (24.4)	.47
Seizure†	6 (5.5)	6 (8.9)	0 (0)	.08
Vision changes†	15 (13.8)	10 (14.7)	5 (12.2)	.95
Musculoskeletal symptom, n (%)				
Dental issues†	10 (9.2)	5 (7.4)	5 (12.2)	.23
Myalgias	70 (24.1)	45 (23.0)	25 (26.3)	.53
Arthralgias	98 (33.7)	66 (33.7)	32 (33.7)	>.99
Limited range of motion	66 (22.7)	51 (26.0)	15 (15.8)	.05
Joint contracture	3 (1)	3 (1.5)	0 (0)	.55
Limb length discrepancy	13 (4.5)	12 (6.1)	1 (1.1)	.07

IQR, Interquartile range.
*Laterality unavailable for 1 adult-onset participant.
†Headache, seizure, vision changes, and dental issues shown for participants with any head or neck involvement (N = 109).

(linear 24.4%, generalized 29.1%, plaque 12.7%, and mixed 13.8%; $P = .02$) and intravenous pulse (linear 12.4%, generalized 2.7%, plaque 3.8%, and mixed 3.4%; $P < .001$). Methotrexate was the most commonly used steroid-sparing agent (linear 42.1%, generalized 37.4%, plaque 17.8%, and mixed 17.2%; $P < .001$). Ultraviolet A1 phototherapy was used in similar frequency across all subtypes ranging from 6.9% in mixed to 24.7% in generalized morphea. Physical therapy was performed more frequently in patients with linear morphea (12.7%) than patients with other subtypes (generalized 4.4%, plaque 6.3%, mixed 3.4%; $P = .02$).

The median DLQI for linear morphea (3, IQR 1.5-7) was similar to that for the generalized subtype (4, IQR 1-9), correlating with a small effect on quality of life.[15] The median linear CDLQI for linear morphea was 3 (IQR 1-6); this score could not be compared among pediatric patients with other subtypes given their small numbers.

In depth analysis of the linear morphea group

Details of the features of linear morphea participants are found in Table II. Although most linear morphea patients had pediatric onset, adult-onset disease was observed in 32.6% (95/291) of linear morphea participants and occurred at a median age of 31 (IQR 24-40) years.

Within linear morphea, lesions were more commonly found on the trunk and extremities (74.6%, 217/291) than the head and neck (37.5%, 109/291, $P = .04$). Lesions occurred on the right and left side with similar frequency, while bilateral or midline lesions occurred in 46.6% (135/290) of patients. Of those with linear lesions located on the head or neck (N = 109), 65.1% (71/109) were consistent with en coup de sabre lesions, and 31.2% (34/109) had Parry-Romberg syndrome, also known as hemifacial atrophy. Of note, 13.8% (15/109) of those with head and neck involvement had features of both en coup de sabre and Parry-Romberg syndrome. Participants with head and neck lesions had lower median LoSCAT component scores than those with lesions on the trunk and extremities ($P < .001$).

Myalgias and arthralgias were common in linear morphea (24.1% and 33.7%, respectively). Deep involvement was observed in 64.3% (187/291) of

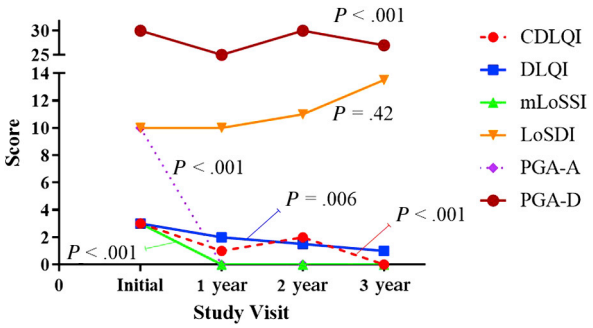

Fig 1. Median quality of life and LoSCAT component scores in linear morphea participants over time. Paired scores were analyzed over a 3-year follow-up period. CDLQI and DLQI scores significantly improved between the baseline and 3-year follow-up study visits ($P < .001$ and $P = .006$, respectively). Activity (measured by mLoSSI and PGA-A) decreased significantly over time ($P < .001$). LoSDI scores showed a trend to increase; however, this did not reach significance ($P = .42$). Median PGA-D scores minimally decreased from 30 (IQR 20-50) at baseline to 27 (IQR 20-40) at 3-year follow-up ($P < .001$). *CDLQI*, Children's Dermatology Life Quality Index; *DLQI*, Dermatology Life Quality Index; *IQR*, interquartile range; *LoSDI*, Localized Scleroderma Damage Index; *mLoSSI*, modified Localized Scleroderma Skin Severity Index; *PGA-A*, Physician Global Assessment of Disease Activity; *PGA-D*, Physician Global Assessment of Disease Damage.

participants with linear morphea, and those with deep involvement (28.6%, 55/192) were more likely than those without (11.1%, 11/99, $P < .001$) to have limitations in range of motion.

Comparison between participants with pediatric and adult-onset linear morphea revealed that multiple lesions were more frequent in pediatric-onset (57.1%, 112/196) than adult-onset (44.2%, 42/95, $P = .04$) disease. There was no significant difference in the frequency of functional abnormalities between the adult-onset and pediatric-onset groups (Table II).

Longitudinal analyses of linear morphea participants

Quality of life scores and clinical outcomes from 527 linear morphea study visits over a 3-year period are shown in Fig 1. In total, 93 participants completed both baseline and 1-year follow-up quality of life surveys, 59 (63.4%) completed CDLQI and DLQI at 2 years, and 36 (38.7%) participants completed all 3 years.

Median CDLQI scores decreased from 3 (IQR 1-6), indicating an initial small impact on quality of life, to 0 (IQR 0-2) or no effect ($P < .001$). Median DLQI scores also significantly decreased from 3 (IQR 1.5-7) at baseline to 1 (IQR 0-3) at 3 years, suggesting a decreasing effect on life quality ($P = .006$). There was no association between lesion location in cosmetically sensitive sites and life quality in the linear morphea group. However, presence of functional impairment in adult patients showed a trend toward greater quality of life impact (median DLQI [IQR] with vs without, 5 [3-8] vs 3 [1-7]; $P = .05$).

Median mLoSSI scores for linear morphea patients decreased from 3 (IQR 0-7) at baseline to 0 (IQR 0-0) at first annual follow-up ($P < .001$, Fig 1). Of 137 participants with active lesions and mLoSSI >0 at the initial visit, 83 (60.6%) reached remission. Recurrence of activity occurred in 13 linear participants. Median LoSDI scores increased from 10 (IQR 5-15.8) at baseline to 13.5 (IQR 5.8-25) at 3 years; however, this was not statistically significant. PGA-D scores decreased from 30 (IQR 20-50) to 27 (IQR 20-40; $P < .001$). The change in LoSCAT scores were similar in the pediatric and adult-onset groups, suggesting a comparable disease course and response to treatment (Fig 2).

DISCUSSION

In the present study, we compared the characteristics of linear morphea versus other morphea subtypes among participants of the MAC cohort, the largest prospective single-site cohort of adults and children with morphea. We also report results of in-depth cross-sectional and longitudinal examinations of linear morphea participants.

Our results confirmed prior observations by our group and others. We found that linear morphea is predominantly pediatric onset, and this subtype affects a larger percentage of male patients than the other subtypes.[1,6,16] Furthermore, our group and others have reported that current quality of life measures, such as the DLQI and CDLQI, generally reflect that morphea has a mild effect on life quality.[17] However, one third of participants experienced functional limitations. These findings, which likely affect life quality, are not captured by existing dermatology-based quality of life measures. This implies that existing dermatology measures might not capture elements relevant to the experience of patients with linear morphea.

Comparisons of linear morphea with other subtypes yielded novel observations. Although linear morphea has been previously associated with functional limitations, we identified a similar frequency of functional limitations in the generalized morphea group. Moreover, the presence of deep (soft tissue) involvement was strongly associated with functional limitations in both the linear and generalized subtypes. Taken together, these results underscore the

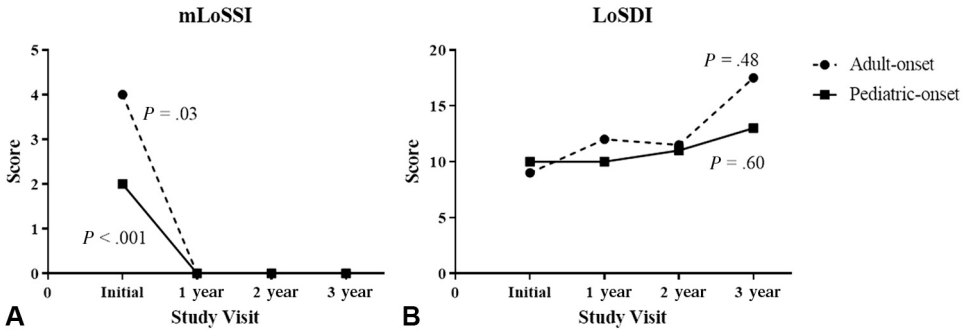

Fig 2. mLoSSI and LoSDI scores in patients with pediatric and adult-onset linear morphea over time. **A**, Median scores for both groups decreased from baseline to first annual study visit and remained stable. **B**, LoSDI scores showed a trend to increase over time. *LoSDI*, Localized Scleroderma Damage Index; *mLoSSI*, modified Localized Scleroderma Skin Severity Index.

necessity to evaluate function not only in linear morphea but also in other subtypes, as well as the need to assess for depth of involvement, which has not been a consideration in most guidelines.[18]

Because the LoSCAT was developed by pediatric rheumatologists, its validation relied heavily on patients with the linear morphea subtype. This raises the question of whether the LoSCAT performs differently in linear morphea versus the other subtypes. We found that those with linear morphea had overall lower LoSCAT component scores (mLoSSI, LoSDI), likely because of the weight the LoSCAT places on quantity of body sites affected. Further, linear morphea patients had higher scores on the PGA-D than patients with other subtypes. This probably occurred because the PGA scores take extra cutaneous manifestations, such as limb length discrepancy, into account, which is not part of the other LoSCAT component scores. Further studies are needed to examine whether the LoSCAT requires separate cutoff values for severity and clinically important difference based on morphea subtype.

Cross-sectional analysis of linear morphea participants yielded new information about the demographic and clinical features of this subtype. First, although linear morphea is thought to predominate in children, over one third of linear morphea patients had disease onset during adulthood, usually in the 20s-30s. Second, many participants with linear morphea had multiple lesions, and 12.4% had lesions above and below the neck (Table II), a distribution that was previously thought to be infrequent.[1] In many of these cases, patients would have met criteria for generalized morphea on the basis of number of lesions and body sites involved. However, the demographic features and morphology of the lesions were linear, suggesting the definition of generalized morphea might need to be revised to exclude those with multiple linear lesions. Lastly, though it is known that lesions which cross joints predispose to limitations in range of motion,[6,19] we found a higher frequency of functional limitations in linear morphea participants with deep lesions. Therefore, we suggest that both lesion location and depth of involvement should be considered when assessing risk for these sequelae.

Longitudinal examination of linear morphea showed most participants achieved disease inactivity with standard of care treatment as measured by the mLoSSI. Further, they remained inactive at the 2-year follow-up (Fig 1). In contrast, LoSDI scores stabilized with successful treatment. This indicates that current treatments abrogate activity or inflammation and stabilize damage in most patients. This should prompt practitioners to initiate appropriate treatment on the basis of the subtype and depth of involvement while the morphea lesions are active and inflammatory.

This study was not population-based; thus, the demographic features, prevalence of different morphea subtypes, and clinical features might not reflect the overall morphea population. Second, although this is the largest sample of linear morphea prospectively examined to date, the sample size still limits analysis among subsets. By design, observational studies provide estimates of association and have the risk of confounders.

The results of our study have several implications for practice. Practitioners should evaluate for functional impairment, particularly if deep involvement is present. Second, there is a high frequency of adult-onset linear morphea, which is associated with similar functional limitations as pediatric onset. Last, current standard of care treatments effectively abrogate activity and stabilize permanent disease damage, underscoring the need to identify patients

with active, inflammatory morphea and promptly initiate appropriate treatment.

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Supplemental Table I. Treatments used in linear morphea participants

Treatment	All linear morphea participants, N = 291	Pediatric onset, n = 196	Adult onset, n = 95	*P* value
Topical steroids, n (%)	173 (59.5)	119 (60.7)	54 (56.8)	.53
Intralesional kenalog, n (%)	42 (14.4)	23 (11.7)	19 (20)	.06
Topical immunomodulators, n (%)	40 (13.7)	20 (10.2)	20 (21.1)	.01
Topical vitamin D analogs, n (%)	81 (27.8)	63 (32.1)	18 (18.9)	.02
Intravenous pulse steroids, n (%)	36 (12.4)	34 (17.3)	2 (2.1)	<.001
Oral steroids, n (%)	71 (24.4)	54 (27.6)	17 (17.9)	.07
Antimalarials, n (%)	37 (12.7)	22 (11.2)	15 (15.8)	.27
Methotrexate, n (%)	123 (42.3)	95 (48.5)	28 (29.5)	.002
Mycophenolate mofetil or mycophenolic acid, n (%)	12 (4.1)	11 (5.6)	1 (1.1)	.11
Ultraviolet A1, n (%)	54 (18.6)	36 (18.4)	18 (18.9)	.91
Narrowband ultraviolet B, n (%)	13 (4.5)	9 (4.6)	4 (4.2)	>.99
Psoralen and ultraviolet A, n (%)	16 (5.5)	12 (6.1)	4 (4.2)	.59
Physical therapy, n (%)	37 (12.7)	32 (16.3)	5 (5.3)	.008
Surgical intervention, n (%)	10 (3.4)	7 (3.6)	3 (3.2)	>.99