

Supplementary material has been published as submitted. It has not been copyedited, typeset or checked for scientific content by *Acta Dermato-Venereologica*

**Table SI.** Criteria for the histologic grade of sclerosis

Mild	Presence of no fibrosis in the papillary dermis and light fibrosis in the superficial reticular dermis or in the median reticular dermis or in the deep reticular dermis;
Moderate	All cases that did not belong to mild or severe
Severe	Presence of severe fibrosis in the deep reticular dermis and in the median reticular dermis irrespective of the degree in the superficial reticular dermis and in the papillary dermis or Severe fibrosis in the deep reticular dermis plus moderate fibrosis in the median and in the superficial reticular dermis as well as in the papillary dermis

**Table SII.** Definition for the patterns of sclerosis

Top heavy	Thickened collagen bundles exclusively in the papillary dermis to the superficial reticular dermis.
Bottom heavy	Thickened collagen bundles in the deep reticular dermis and subcutis, and sparing from the papillary dermis to the median reticular dermis.
Full thickness	Thickened collagen bundles throughout the full dermis.

**Table III. ACR-EULAR criteria for SSc patients (n=15)**

			1	2	3	4	5	6	7	8	9	10	11	12	13	14	15
			F/78	F/53	M/58	M/30	F/53	F/38	F/75	F/70	F/61	M/52	F/47	F/34	F/42	F/46	F/66
Items	Sub-items	score															
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints (sufficient criterion)		9	0	0	<b>9</b>	0	<b>9</b>	0	0	<b>9</b>	<b>9</b>	0	0	0	0	0	0
Skin thickening of the fingers (only count the highest score)	Puffy fingers	2	2	2	2	0	<b>2</b>	0	<b>2</b>	<b>2</b>	0	2	0	0	<b>2</b>	0	<b>2</b>
	Sclerodactyly of the fingers (distal to MCP but proximal to the PIPs)	4	<b>4</b>	<b>4</b>	<b>4</b>	<b>4</b>	0	<b>4</b>	0	0	<b>4</b>	<b>4</b>	0	<b>4</b>	0	0	0
Finger tip lesions (only count the highest score)	Digital Tip Ulcers	2	0	<b>2</b>	0	0	0	0	0	<b>2</b>	0	0	0	0	0	0	0
	Finger Tip Pitting Scars	3	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Telangiectasia		2	<b>2</b>	0	0	<b>2</b>	<b>2</b>	<b>2</b>	<b>2</b>	0	<b>2</b>	<b>2</b>	0	<b>2</b>	<b>2</b>	<b>2</b>	<b>2</b>
Abnormal nailfold capillaries		2	<b>2</b>	0	0	<b>2</b>	<b>2</b>	<b>2</b>	<b>2</b>	0	<b>2</b>	<b>2</b>	0	<b>2</b>	<b>2</b>	<b>2</b>	<b>2</b>
Pulmonary arterial hypertension and/or Interstitial lung Disease* (*Maximum score is 2)	PAH ILD	2	0	<b>2</b>	<b>2</b>	0	<b>2</b>	0	<b>2</b>	0	<b>2</b>	<b>2</b>	0	<b>2</b>	<b>2</b>	<b>2</b>	0
Raynaud's phenomenon		3	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	<b>3</b>	0	<b>3</b>	<b>3</b>	<b>3</b>
Scleroderma related antibodies** (any of anti-centromere, anti-topoisomerase I (anti-Scl 70), anti-RNA polymerase III) (**Maximum score is 3)	anti-centromere anti-topoisomerase I anti-RNA polymerase III	3	<b>3</b>	<b>3</b>	0	0	0	<b>3</b>	0	<b>3</b>	<b>3</b>	0	<b>3</b>	<b>3</b>	0	0	0
<b>Sum</b>			<b>14</b>	<b>14</b>	<b>18</b>	<b>11</b>	<b>11</b>	<b>14</b>	<b>11</b>	<b>19</b>	<b>25</b>	<b>13</b>	<b>6</b>	<b>13</b>	<b>11</b>	<b>9</b>	<b>9</b>

Only numbers in bold were included in the score.

**Table SIV.** Summary of patient characteristics and clinical findings between the localized morphea, generalized morphea, and systemic sclerosis groups

	LM (n=150)	GM (n=16)	SSc (n=15)	P-value
	N (%)	N (%)	N (%)	Overall
Female sex	124 (82.7)	13 (81.3)	12 (80.0)	0.930
Age at diagnosis, y (median, IQR)	33.5 (20.0–49.8)	27.5 (21.0–61.0)	53.0 (42.0–66.0)	<b>0.004</b>
Duration of symptoms, m (median, IQR)	15.0 (6.0–45.0)	30.0 (8.3–75.0)	35.0 (6.0–86.0)	0.323
Subtype	Circumscribed: 80 (53.3) Linear: 66 (40) Mixed: 4 (2.67)	Generalized: 13 (81.3) Pansclerotic: 3 (18.7)	Limited: 5 (33.3) Diffuse: 5 (33.3) Overlap: 5 (33.3)	-
<b>Anatomical location<sup>a</sup></b>				
Scalp	15 (10.0)	1 (6.3)	0 (0.0)	0.667
Forehead (En coup de sabre)	23 (15.3)	1 (6.3)	0 (0.0)	0.212
Face and neck	27 (18.0)	1 (6.3)	4 (26.7)	0.337
Trunk	61 (40.7)	<b>15 (93.8)</b>	6 (40)	<b>&lt;0.001</b>
Upper extremities	24 (16.0)	<b>11 (68.8)</b>	<b>12 (80.0)</b>	<b>&lt;0.001</b>
Lower extremities	31 (20.7)	<b>14 (87.5)</b>	7 (46.7)	<b>&lt;0.001</b>
Hands or feet	2 (1.3)	<b>6 (37.5)</b>	<b>8 (53.3)</b>	<b>&lt;0.001</b>
Blaschkoid distribution	20 (13.3)	2 (12.5)	1 (6.7)	0.908
<b>Functional limitations or clinical symptoms<sup>a</sup></b>				
Pain or arthralgia of affected joints	13 (8.7)	5 (31.3)	<b>10 (66.7)</b>	<b>&lt;0.001</b>
Pruritus	18 (12.0)	4 (25.0)	<b>6 (40.0)</b>	<b>0.010</b>
Joint contracture or decreased ROM	3 (2.0)	<b>4 (25.0)</b>	<b>5 (33.3)</b>	<b>&lt;0.001</b>
Sclerodactyly	0 (0.0)	1 (6.3)	<b>8 (53.3)</b>	<b>&lt;0.001</b>
Raynaud phenomenon	0 (0.0)	0 (0.0)	<b>14 (93.3)</b>	<b>&lt;0.001</b>
<b>History of autoimmune disease</b>				
	20 (13.3)	4 (25.0)	<b>9 (60.0)</b>	<b>&lt;0.001</b>
<b>Non-cutaneous manifestation<sup>b</sup></b>				
Musculoskeletal	8 (5.3)	4 (25.0)	<b>15 (100.0)</b>	<b>&lt;0.001</b>
CNS	0 (0.0)	0 (0.0)	0 (0.0)	>0.99
GI	0 (0.0)	0 (0.0)	<b>7 (46.7)</b>	<b>&lt;0.001</b>
Pulmonary	0 (0.0)	0 (0.0)	<b>9 (60.0)</b>	<b>&lt;0.001</b>
<b>Laboratory finding</b>				
≥1 abnormal laboratory value	41/128 (32.0)	<b>13/16 (81.3)</b>	<b>15/15 (100)</b>	<b>&lt;0.001</b>

ANA level $\geq$ 1:80	22/123 (17.9)	<b>12/16 (75.0)</b>	<b>13/15 (86.7)</b>	<b>&lt;0.001</b>
Peripheral eosinophilia ( $>500/\mu\text{L}$ )	5/124 (4.0)	<b>4/16 (25.0)</b>	<b>4/15 (26.7)</b>	<b>0.001</b>
Elevated ESR or CRP level	17/60 (28.3)	3/14 (21.4)	<b>13/15 (86.7)</b>	<b>&lt;0.001</b>
Rheumatoid factor	6/44 (13.6)	2/9 (22.2)	5/15 (33.3)	0.173
Anti-Scl-70 Ab	0/76 (0.0)	0/14 (0.0)	<b>4/15 (26.7)</b>	<b>&lt;0.001</b>
Anti-centromere Ab	0/46 (0.0)	2/10 (20.0)	3/15 (20.0)	<b>0.005</b>

<sup>a</sup>Patients might be listed in  $>1$  category

<sup>b</sup>Musculoskeletal: Arthralgia or decrease of range of motion. Pulmonary: ILD or pulmonary hypertension, GI: GERD or achalasia

LM: localized morphea; GM: generalized morphea; SSc: systemic sclerosis; IQR: interquartile range; ANA: antinuclear antibody; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; Scl-70: scleroderma-70; Ab: antibody; CNS: central nervous system; GI: gastrointestinal; RO: range of motion.

Bold cells show significant adjusted standardized residuals ( $>2.1$ ) or  $P<0.05$ .

**Table SV.** Comparison of histopathological features between the localized morphea, generalized morphea, and systemic sclerosis groups

	LM (n=150)	GM (n=16)	SSc (n=15)	P-value
	N (%)	N (%)	N (%)	Overall
<b>Pattern of sclerosis</b>				
Top heavy	36 (24.0)	3 (18.8)	1 (6.7)	0.354
Bottom heavy	63 (42.0)	7 (43.8)	9 (60.0)	0.407
Full thickness	51 (34.0)	6 (37.5)	5 (33.3)	0.958
<b>Degree of sclerosis</b>				
Mild	<b>44 (29.3)</b>	3 (18.8)	0 (0.0)	<b>0.024</b>
Moderate	73 (48.7)	5 (31.3)	6 (40.0)	0.362
Severe	33 (22.0)	8 (50.0)	<b>9 (60.0)</b>	<b>0.001</b>
<b>DSQ or fat sclerosis</b>	70 (46.7)	12 (75.0)	<b>15 (100.0)</b>	<b>&lt;0.001</b>
<b>Degree of inflammation</b>				
None to mild	78 (52.0)	5 (31.3)	10 (66.7)	0.134
Moderate	55 (36.7)	7 (43.8)	5 (33.3)	0.816
Severe	17 (11.3)	4 (25.0)	0 (0.0)	0.087
<b>DSQ or fat inflammation</b>	32 (21.3)	<b>7 (43.8)</b>	1 (6.7)	<b>0.046</b>
<b>Specific cell type present</b>				
Plasma cells	81 (54.0)	<b>14 (87.5)</b>	10 (66.7)	<b>0.028</b>
Eosinophils	17 (11.3)	4 (25.0)	4 (26.7)	0.07
<b>Pigmentation</b>				
Basal pigmentation	70 (46.7)	11 (68.8)	10 (66.7)	0.101
Melanin incontinence	31 (20.7)	<b>8 (50.0)</b>	<b>9 (60.0)</b>	<b>&lt;0.001</b>
None	<b>64 (42.7)</b>	3 (18.8)	1 (6.7)	<b>0.006</b>
<b>Presence of epidermal atrophy</b>	59 (39.3)	5 (31.3)	4 (26.7)	0.540
<b>Presence of dermal telangiectasia</b>	47 (31.3)	3 (18.8)	7 (46.7)	0.246

LM: localized morphea; GM: generalized morphea; SSc: systemic sclerosis; DSQ: dermosubcutaneous fat junction.

Bold cells show significant adjusted standardized residuals (>2.1) or P<0.05.