

Supplementary Table S1. Clinical association with acro-osteolysis.

Data	Overall n 120 (%)	No acro-osteolysis n (%)	Acro-osteolysis n (%)	OR (95%CI)	p-value
Female	75 (62.5)	28 (65.1)	47 (61.0)	0.84 (0.36-1.95)	0.66
Diffuse SSc	64 of 114 (56.1)	24 (60.0)	40 (54.0)	0.78 (0.33-1.83)	0.54
Age at onset (mean ±SD)	54.8 ± 10.6	52.3 ± 10.5	56.2 ± 10.4	NA	0.05
Age at study date (mean ±SD)	56.8 ± 10.5	54.2 ± 10.3	56.2 ± 10.4	NA	0.04 ^a
Duration (mean ±SD)	2.0 ± 1.3	1.9 ± 1.2	2.1 ± 1.3	NA	0.50
Anti-topoisomerase I antibody positive	90 of 107 (84.1)	29 (74.4)	61 (89.7)	3.00 (0.92-10.22)	0.04 ^a
Anti-centromere antibodies positive	6 of 120 (5.0)	2 (4.7)	4 (5.2)	1.12 (0.15-12.9)	0.90
Clinical characteristics at onset					
Raynaud's phenomenon	97 of 118 (82.2)	34 (79.1)	63 (84.0)	1.39 (0.47-4.00)	0.50
Ischaemic ulcer	22 of 118 (18.6)	4 (9.3)	18 (24.0)	3.08 (0.91-13.35)	0.049 ^a
Digital gangrene	2 of 118 (1.7)	0 (0.0)	2 (2.7)	NA	NA
Telangiectasia	20 of 117 (17.1)	7 (16.3)	13 (17.6)	1.10 (0.36-3.56)	0.86
Salt and pepper	83 of 118 (70.3)	30 (69.8)	53 (70.7)	1.04 (0.42-2.54)	0.92
Oedematous	75 of 118 (63.6)	36 (83.7)	39 (52.0)	0.21 (0.07-0.57)	<0.001 ^a
Tendon rub	35 of 117 (29.9)	13 (31.0)	22 (29.3)	0.93 (0.38-2.32)	0.85
Hand deformities	26 of 118 (22.0)	7 (16.3)	19 (25.3)	1.74 (0.62-5.40)	0.25
Synovitis	25 of 118 (21.2)	5 (11.6)	20 (26.7)	2.76 (0.89-10.18)	0.05
Muscle weakness	14 of 117 (12.0)	2 (4.8)	12 (16.0)	3.81 (0.78-36.48)	0.07
Dysphagia	46 of 118 (39.0)	10 (23.3)	36 (48.0)	3.05 (1.23-7.90)	0.01 ^a
Heart burn	43 of 117 (36.8)	19 (45.2)	24 (32.0)	0.57 (0.24-1.33)	0.15
Stomach symptoms	27 of 118 (22.9)	13 (30.2)	14 (18.7)	0.53 (0.20-1.40)	0.15
Constipation	15 of 118 (12.7)	6 (14.0)	9 (12.0)	0.84 (0.24-3.11)	0.76
Weight loss	63 of 119 (52.9)	25 (58.1)	38 (50.0)	0.72 (0.31-1.63)	0.39
MRSS (mean ±SD)	12.9 ± 8.8	12.6 ± 7.4	13.1 ± 9.6	NA	0.76
ESR (mean ±SD)	45.3 ± 23.6	48.3 ± 25.0	43.6 ± 23.2	NA	0.57
CRP (median (IQR))	4.1 (1.6-27)	3.6 (2.4-12.7)	10.4 (1.2-27.4)	NA	0.57
Pulmonary fibrosis	22 of 103 (21.4)	9 (25.7)	13 (19.1)	0.68 (0.23-2.07)	0.44
Alveolitis	41 of 102 (40.2)	15 (42.7)	26 (38.8)	0.85 (0.34-2.12)	0.69
Myositis	19 of 102 (18.6)	6 (17.1)	13 (19.4)	1.16 (0.36-4.14)	0.78
Pulmonary arterial hypertension	6 of 102 (5.9)	1 (2.9)	5 (7.5)	2.74 (0.29-133.42)	0.35
Renal crisis	2 of 102 (2.0)	1 (2.9)	1 (1.5)	0.52 (0.01-41.61)	0.64
Clinical characteristics at study date					
Raynaud's phenomenon	48 of 118 (40.7)	16 (37.2)	32 (42.7)	1.26 (0.55-2.93)	0.56
Ischaemic ulcer	11 of 118 (9.3)	0 (0.0)	11 (14.7)	NA	NA
Digital gangrene	2 of 118 (1.7)	0 (0.0)	2 (2.7)	NA	NA
Telangiectasia	25 of 118 (21.2)	7 (16.3)	18 (24.0)	1.62 (0.57-5.06)	0.32
Salt and pepper	78 of 118 (65.6)	27 (62.8)	51 (67.1)	1.21 (0.51-2.83)	0.63
Oedematous	27 of 118 (22.9)	16 (37.2)	11 (14.7)	0.29 (0.11-0.77)	0.01 ^a
Tendon rub	17 of 117 (14.5)	5 (11.6)	12 (16.2)	1.47 (0.44-5.74)	0.50
Hand deformities	43 of 118 (36.4)	9 (20.9)	34 (45.3)	3.13 (1.24-8.42)	0.01 ^a
Synovitis	6 of 118 (5.1)	2 (4.7)	4 (5.3)	1.15 (0.16-13.27)	0.87
Muscle weakness	7 of 118 (5.9)	3 (7.0)	4 (5.3)	0.75 (0.12-5.40)	0.71
Dysphagia	26 of 118 (22.0)	6 (14.0)	20 (26.7)	2.24 (0.77-7.44)	0.11
Heart burn	29 of 118 (24.6)	9 (20.9)	20 (26.7)	1.37 (0.52-3.83)	0.49
Stomach symptoms	13 of 118 (11.0)	6 (14.0)	7 (9.3)	0.63 (0.17-2.48)	0.44
Constipation	11 of 118 (9.3)	2 (4.7)	9 (12.0)	2.80 (0.54-27.64)	0.19
Weight loss	25 of 119 (21.0)	10 (23.3)	15 (19.7)	0.81 (0.30-2.27)	0.65
MRSS (mean ±SD)	9.6 ± 8.6	9.5 ± 8.8	9.7 ± 8.5	NA	0.92
ESR (mean ±SD)	40.7 ± 24.6	40.3 ± 21.8	41 ± 26.0	NA	0.93
CRP (median (IQR))	4 (1.7-21.0)	29.5 (11.3-55.5)	3.7 (1.6-5.8)	NA	0.16
Pulmonary fibrosis	28 of 115 (24.4)	12 (29.3)	16 (21.6)	0.67 (0.27-1.77)	0.36
Alveolitis	56 of 115 (48.7)	21 (51.2)	35 (47.3)	0.85 (0.37-1.97)	0.69
Myositis	21 of 115 (18.3)	10 (24.4)	11 (14.9)	0.54 (0.19-1.60)	0.21
Pulmonary arterial hypertension	6 of 115 (5.2)	2 (4.9)	4 (5.4)	1.11 (0.15-12.83)	0.90
Renal crisis	2 of 115 (1.7)	1 (2.4)	1 (1.4)	0.55 (0.01-44.08)	0.67

SSc: systemic sclerosis; MRSS: modified Rodnan skin score; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; SD: standard deviation; IQR: interquartile range; NA: data not available.

^astatistically significant.

Supplementary Table S2. Logistic regression analysis clinical associated with acro-osteolysis.

Data	Crude OR (95%CI)	Adjusted OR (95%CI)	<i>p</i> -value
Anti-topoisomerase I antibody positive	3.00 (0.92-10.22)	13.96 (2.29-85.10) ^a	0.01 ^a
Ischaemic ulcer at onset	3.08 (0.91-13.35)	2.03 (0.29-14.16)	0.48
Oedematous at onset	0.21 (0.07-0.57)	0.05 (0.01-0.32) ^a	0.01 ^a
Synovitis at onset	2.76 (0.89-10.18)	0.80 (0.16-3.93)	0.78
Muscle weakness at onset	3.81 (0.78-36.48)	13.75 (0.90-210.49)	0.06
Dysphagia at onset	3.05 (1.23-7.90)	6.66 (1.77-25.05) ^a	0.01 ^a
Oedematous at study date	0.29 (0.11-0.77)	0.31 (0.07-1.40)	0.13
Hand deformities at study date	3.13 (1.24-8.42)	3.81 (1.03-14.07) ^a	0.045 ^a
Age at onset	NA	1.14 (0.55-1.63)	0.63
Age at study date	NA	0.95 (0.55-1.63)	0.85

^astatistically significant; NA: data not available.

Supplementary Table S3. Clinical differences stratify by severity of acro-osteolysis.

Data	No	Mild	Moderate	Severe	p-value
Female	28	22	14	11	0.75
Diffuse SSc	24	20	11	9	0.73
Age at onset (mean ±SD)	52.3 ± 10.5	57.0 ± 8.2	54.8 ± 14.7	56.4 ± 9.4	2.22
Age at study date (mean ±SD)	54.2 ± 10.3	58.7 ± 8.1	56.9 ± 14.5	59.0 ± 9.4	0.19
Duration (mean ±SD)	1.9 ± 1.2	1.7 ± 1.2	2.2 ± 1.4	2.6 ± 1.2	0.09
Anti-topoisomerase I antibody positive	29	32	14	15	0.19
Anti-centromere antibodies positive	2	2	1	1	1.00
Clinical characteristics at onset					
Raynaud's phenomenon	34	32	16	15	0.86
Ischaemic ulcer	4	9	3	6	0.11
Digital gangrene	0	0	0	2	0.02 ^a
Telangiectasia	7	5	3	5	0.63
Salt and pepper	30	25	16	12	0.50
Oedematous	36	18	12	9	0.01
Tendon rub	13	12	7	3	0.60
Hand deformities	7	10	5	4	0.69
Synovitis	5	11	5	4	0.21
Muscle weakness	2	8	3	1	0.10
Dysphagia	10	19	10	7	0.049
Heart burn	19	2	7	5	0.52
Stomach symptoms	13	7	5	2	0.41
Constipation	6	5	2	2	1.00
Weight loss	25	20	10	8	0.69
MRSS (mean ±SD)	12.5 ± 7.4	13.6 ± 9.5	15.3 ± 10.8	10.0 ± 8.1	0.29
ESR (mean ±SD)	48.3 ± 25.0	46.1 ± 23.1	44.7 ± 29.5	36.8 ± 9.3	0.84
CRP (median (IQR))	3.6 (2.4-12.7)	24.5 (12.8-143.5)	2.2 (0.4-10.4)	35.8	0.12
Pulmonary fibrosis	9	4	4	5	0.52
Alveolitis	15	11	7	8	0.91
Myositis	6	9	3	1	0.24
Pulmonary arterial hypertension	2	2	1	2	0.62
Renal crisis	1	1	0	0	1.00
Clinical characteristics at study date					
Raynaud's phenomenon	16	19	7	6	0.45
Ischaemic ulcer	0	7	0	4	0.001 ^a
Digital gangrene	0	0	1	1	0.10
Telangiectasia	7	5	6	7	0.11
Salt and pepper	27	28	9	14	0.11
Oedematous	16	10	1	0	0.001 ^a
Tendon rub	5	8	3	1	0.46
Hand deformities	9	13	12	9	0.01 ^a
Synovitis	2	3	0	1	0.75
Muscle weakness	3	2	2	0	0.69
Dysphagia	6	14	4	2	0.05
Heart burn	9	12	5	3	0.58
Stomach symptoms	6	4	3	1	0.91
Constipation	2	4	4	1	0.26
Weight loss	10	9	4	2	0.69
MRSS (median (IQR))	7.5 (2-14)	10 (5-15)	5 (2-13.5)	4 (2-10)	0.30
ESR (mean ±SD)	40.3 ± 21.8	41.8 ± 26.1	32.8 ± 22.6	45.0 ± 29.4	0.82
CRP (median (IQR))	29.5 (11.3-55.5)	8.0 (2.3-136.0)	1.6 (1.0-2.0)	5.1 (3.1-20.9)	0.10
Pulmonary fibrosis	12	7	6	3	0.59
Alveolitis	21	16	20	9	0.97
Myositis	10	5	3	3	0.68
Pulmonary arterial hypertension	2	2	2	0	0.61
Renal crisis	1	1	0	0	1.00

SSc: systemic sclerosis; MRSS: modified Rodnan skin score; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein

^a statistically significant; SD: standard deviation; IQR: interquartile range.