

Supplementary Table S1. Demographic characteristics of SSc patients and healthy controls.

| | Healthy (n = 15) | SSc (n = 32) |
|-----------------------------------|------------------|---------------|
| Age, mean ± SD years | 51.07 ± 3.00 | 55.34 ± 11.52 |
| Female, n (%) | 10 (66.7) | 23 (71.9) |
| Disease duration, mean ± SD years | | 1.57 ± 0.95 |
| Disease subtype | | |
| dcSSc, n (%) | | 20 (62.50) |
| lcSSc, n (%) | | 12 (32.50) |
| mRSS | | 16.13 ± 7.73 |
| DU, n (%) | | 9 (28.13) |
| ILD, n (%) | | 10 (38.46) |
| PAH, n (%) | | 8 (27.59) |
| NVC | | |
| Non-SSc pattern, n (%) | | 8 (27.59) |
| SSc pattern, n (%) | | 21 (72.41) |
| early, n (%) | | 0 (0) |
| active, n (%) | | 12 (41.38) |
| late, n (%) | | 9 (31.03) |
| Drugs | | |
| Glucocorticoid, n (%) | | 17 (51.13) |

Immunosuppressive, n (%) 25 (78.13)

dcSSc: Diffused cutaneous SSc; lcSSc: limited cutaneous SSc; mRSS: Modified Rodnan skin score; ILD: Interstitial lung disease; PAH: Pulmonary arterial hypertension; DU: Digital ulcer; NVC: Nailfold videocapillaroscopy;

Supplementary Table S2. Demographic characteristics of SSc patients and healthy controls who underwent skin biopsy.

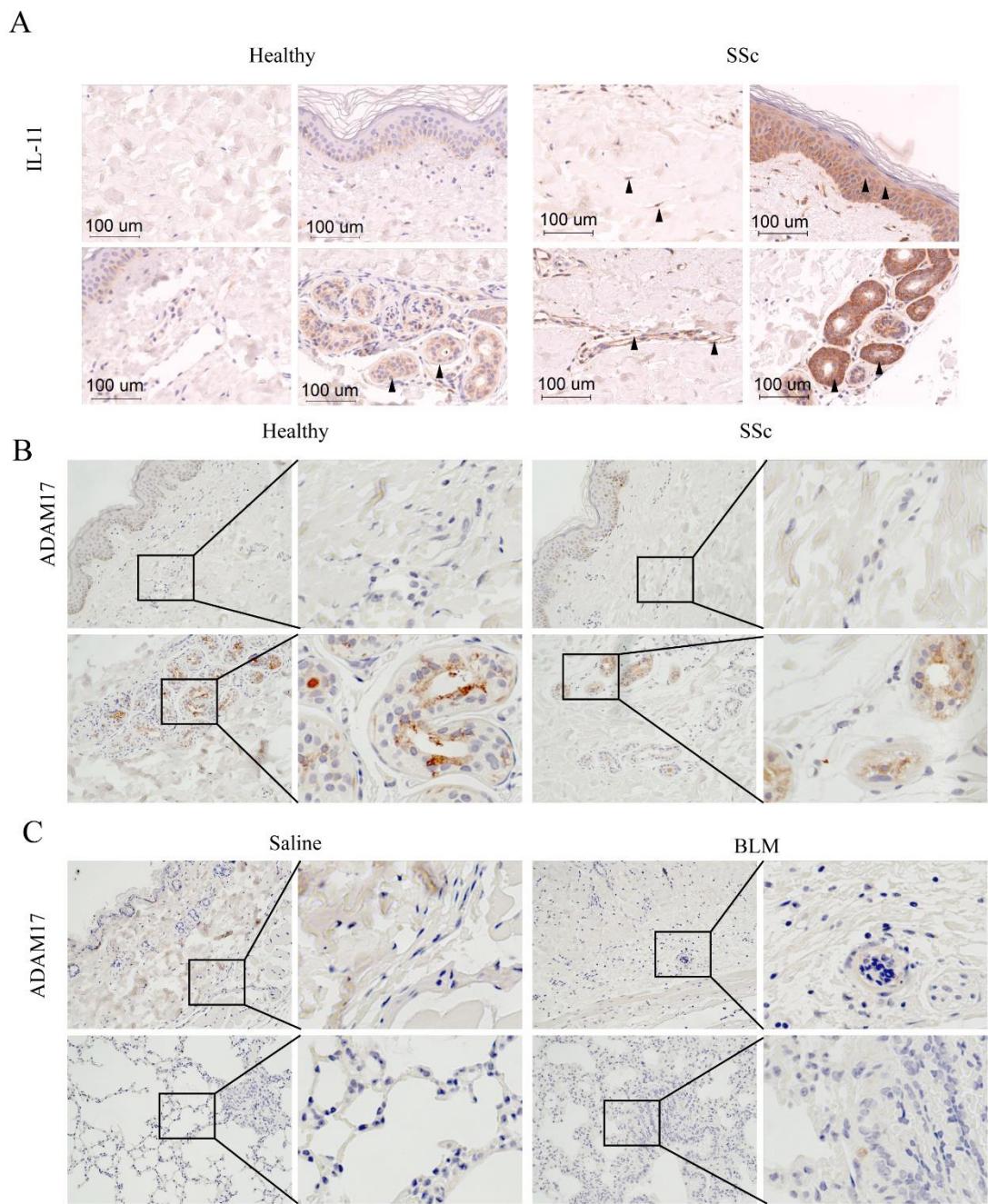
| Skin sample | Sex | Age | Disease subtype | Disease Duration, years | Immunosuppression |
|-------------|-----|-----|-----------------|-------------------------|---------------------------|
| Control 1 | F | 20 | | | |
| Control 2 | F | 33 | | | |
| Control 3 | F | 33 | | | |
| Control 4 | F | 34 | | | |
| Control 5 | F | 59 | | | |
| Control 6 | F | 67 | | | |
| SSC 1 | F | 22 | dcSSC | 48 | None |
| SSC 2 | F | 27 | dcSSC | 13 | Hydroxychloroquine |
| SSC 3 | F | 34 | dcSSC | 13 | Methotrexate |
| SSC 4 | F | 47 | dcSSC | 5 | Hydroxychloroquine; |
| | | | | | Prednisone |
| SSC 5 | F | 48 | dcSSC | 5 | Mycophenolate; Prednisone |

| | | | | | |
|-------|---|----|-------|----|---------------|
| SSC 6 | F | 50 | dcSSC | 24 | None |
| SSC 7 | F | 62 | dcSSC | 36 | Tofacitinib |
| SSC 8 | M | 59 | dcSSC | 24 | Mycophenolate |
| SSC 9 | M | 65 | lcSSC | 9 | Prednisone |

Supplementary Table S3. Levels of plasma IL-11 in SSc and healthy control

| Group | Healthy (n=4/15) | SSc (n=6/32) | P value |
|------------------------|------------------|--------------|---------|
| Female sex, n (%) | 4 (100) | 5 (83.33) | >0.999 |
| Age, mean ± SD years | 51.25±4.35 | 50.83±9.50 | 0.937 |
| IL-11, mean ± SD pg/ml | 2.26±1.74 | 5.90±5.54 | 0.246 |

SSc: Systemic sclerosis;



Supplementary Figure S1. Expression of IL-11 and ADAM17 in skin. (A) Several types of IL-11 positive cells in human skin. (B-C) Immunohistochemical staining of ADAM17 in skin of SSc patients and healthy controls, and in skin/ pulmonary of model and control mice. (magnification = 20X).