

## Figure S1. Immunostaining for CTGF in skin samples from PBS or BLM treated WT or *Fli1*<sup>+/-</sup> mice.

**A.** The representative pictures of immunostaining for CTGF in skin samples from WT and *Fli1*+/- mice treated with PBS or BLM. **B.** The relative number of CTGF-positive fibroblasts and FSP1/CTGF double positive fibroblasts in the dermis. The number per high-power field is adjusted to that in PBS-treated WT mice set at 1 (n = 5). **C.** The representative pictures of immunofluorescence for FSP1 (green) and CTGF (red) in skin samples from WT and *Fli1*+/- mice treated with PBS or BLM. Double positive cells were indicated by arrows. Values are the means  $\pm$  SEM. \*P < 0.05. Bars, 10 µm.



# Figure S2. Immunostaining for integrin $\beta$ 3 in skin samples from PBS or BLM treated WT or *Fli1*<sup>+/-</sup> mice

**A.** The representative pictures of immunostaining for integrin  $\beta$ 3 in skin samples from WT and *Fli1*+/- mice treated with PBS or BLM. **B.** The relative number of FSP1/integrin  $\beta$ 3 double positive fibroblasts in the dermis. The number per high-power field is adjusted to that in PBS-treated WT mice set at 1 (n = 5). **C.** The representative pictures of immunofluorescence for FSP1 (green) and integrin  $\beta$ 3 (red) in skin samples from WT and *Fli1*+/- mice treated with PBS or BLM. Double positive cells were indicated by arrows. Values are the means  $\pm$  SEM. \*P < 0.05. Bars 10µm



## Figure S3. Immunostaining for integrin $\beta$ 5 in skin samples from PBS or BLM treated WT or *Fli1*<sup>+/-</sup> mice.

**A.** The representative pictures of immunostaining for integrin  $\beta$ 5 in skin samples from WT and *Fli1*+/- mice treated with PBS or BLM. **B.** The relative number of FSP1/integrin  $\beta$ 5 double positive fibroblasts in the dermis. The number per high-power field is adjusted to that in PBS-treated WT mice set at 1 (n = 5). **C.** The representative pictures of immunofluorescence for FSP1 (green) and integrin  $\beta$ 5 (red) in skin samples from WT and *Fli1*+/- mice treated with PBS or BLM. Double positive cells were indicated by arrows. Values are the means  $\pm$  SEM. \*P < 0.05. Bars, 10 µm



#### Figure S4. The expression profiles of cytokines, chemokines, and M2 macrophage markers in the lesional skin of PBS-treated mice.

mRNA levels of the *ll1b, ll4, ll6, ll10, ll12a, ll17a, lfng, Tnfa, Mcp1, Arg1, Fizz1,* and *Ym1* genes were measured in the skin of WT and *Fli1+/-* mice with PBS treatment. Values are the means  $\pm$  SEM (n = 4-8). ND; not determined. AU, arbitrary unit.



## Figure S5. The evaluation of inflammatory cell infiltration in mice treated with PBS or BLM.

The representative pictures of F4/80, toluidine blue, CD4, and CD8 staining are shown in the skin of WT and  $Fli1^{+/-}$  mice at day 7 and 28 after PBS or BLM injection (n = 5).

![](_page_5_Figure_0.jpeg)

#### Figure S6. mRNA levels of the *Col1a2* gene and the levels of soluble type I collagen in the skin of WT and *Fli1+/-* mice.

**A.** mRNA expression of the *Col1a2* gene in the skin tissue of WT and *Fli1+/-* mice at day 28 after PBS injection were assessed (n = 10). **B.** The levels of soluble type I collagen were elevated in *Fli1+/-* mice. Pepsin-soluble collagen was stained with Coomassie blue (a left panel). Arrows indicate collagen  $\alpha$ 1(I) and  $\alpha$ 2(I) subunits.  $\beta$ -components represent cross-linked  $\alpha$ -chain dimers. Collagen levels were quantitated using public domain software ImageJ (n = 3; a right panel). Values are the means  $\pm$  SEM. \**P* < 0.05, \*\*\**P* < 0.001. AU, arbitrary unit.

![](_page_6_Figure_0.jpeg)

## Figure S7. mRNA expression of the *ICAM1*, *GlyCAM1*, *SELP*, and *SELE* genes in the skin tissue of healthy controls and SSc patients.

Skin sections from diffuse cutaneous systemic sclerosis (dcSSc) patients with disease duration of  $\leq 1$  year, dcSSc patients with disease duration of >1 year, and healthy controls were assessed (n = 4-6). Values are the means  $\pm$  SEM. \*P < 0.05. AU, arbitrary unit.

Gene Forward sequence Reverse sequence mTgfb1 5' -GCAACATGTGGAACTCTACCAGAA- 3' 5' -GACGTCAAAAGACAGCCACTCA- 3' m*Ctgf* 5' -GTGCCAGAACGCACACTG- 3' 5' -CCCCGGTTACACTCCAAA- 3' mIl1b 5' -TTGACGGACCCCAAAAGAT- 3' 5' -GAAGCTGGATGCTCTCATCTG- 3' mIl4 5' -CAACGAAGAACACCACAGAG- 3' 5' -GGACTTGGACTCATTCATGG- 3' m*Il6* 5' -GATGGATGCTACCAAACTGGAT- 3' 5' -CCAGGTAGCTATGGTACTCCAGA- 3' mIl10 5' -TTTGAATTCCCTGGGTGAGAA- 3' 5' -ACAGGGGAGAAATCGATGACA- 3' mIl12a 5' -CACCCTGTTGATGGTCACGAC- 3' 5' -ACTCTGCGCCAGAAACCTC- 3' 5' - CTCCAGAAGGCCCTCAGACTAC- 3' 5' -AGCTTTCCCTCCGCATTGACACAG- 3' mIl17a 5' -TCAAGTGGCATAGATGTGGAAGAA- 3' 5' -TGGCTCTGCAGGATTTTCATG- 3' m*Ifng* 5' -ACCCTCACACTCAGATCATCTTC- 3' 5' -TGGTGGTTTGCTACGACGT- 3' m*Tnfa* mMcp-1 5' -CATCCACGTGTTGGCTCA- 3' 5' -GATCATCTTGCTGGTGAATGAGT- 3' mItgav 5' -GGTGTGGATCGAGCTGTCTT- 3' 5' -CAAGGCCAGCATTTACAGTG- 3' mItgb3 5' -CAGGATATCAGGACCCTTGG- 3' 5' -GTGGGAGGGCAGTCCTCTA- 3' 5' -CACGGACACTTCAAAGGATG- 3' mItgb5 5' -ACCTGCCAAGATGGCATATC- 3' mIcam1 5' -GACGCAGAGGACCTTAACAG- 3' 5' -GACGCCGCTCAGAAGAAC- 3' mGlycam-1 5' -GACGCAGAGGACCTTAACAG- 3' 5' -GACGCCGCTCAGAAGAAC- 3' 5' -TCCAGGAAGCTCTGACGTACTTG- 3' 5' -GCAGCGTTAGTGAAGACTCCGTAT- 3' mSelp mSele 5' -GCCGAGGGACATCATCACAT- 3' 5' -TGAACTGAAGGGATCAAGAAGACT- 3' 5' -CAGAAGAATGGAAGAGTCAG - 3' 5' -CAGATATGCAGGGAGTCACC- 3' mArg1 mFizz1 5' -TCCCAGTGAATACTGATGAGA- 3' 5' -CCACTCTGGATCTCCCAAGA- 3' 5' -CCACTGAAGTCATCCATGTC- 3' mYm1 5' -GGGCATACCTTTATCCTGAG- 3' mGapdh 5' -CGTGTTCCTACCCCCAATGT- 3' 5' -TGTCATCATACTTGGCAGGTTTCT- 3' 5' -GAGGACCTGCCCTCCTTC- 3' hITGAV 5' -GCCGTGGATTTCTTCGTG- 3' hITGB3 5' -CGCTAAATTTGAGGAAGAACG- 3' 5' -GAAGGTAGACGTGGCCTCTTT- 3' 5' -GGAGTTTGCAAAGTTTCAGAGC- 3' 5' -TGTGCGTGGAGATAGGCTTT- 3' hITGB5 h*CTGF* 5' -TTGCGAAGCTGACCTGGAAGAGAA- 3' 5' -AGCTCGGTATGTCTTCATGCTGGT - 3' hICAM1 5' -TAGAGACCCCGTTGCCTAAA- 3' 5' -TCATACACCTTCCGGTTGTTC- 3' hGlyCAM-1 5' -TGAAATTCACTCGGAGACTGC- 3' 5' -TGGCAAGTTTTCCCTCTGA- 3' hSELP 5' -TTAGTTGGACCGGAAGTGGT- 3' 5' -CAGGTGCTGACACTGCACA- 3' **h**SELE 5' -ACCAGCCCAGGTTGAATG- 3' 5' -GGTTGGACAAGGCTGTGC- 3' 5' -CTGGCCCTTGTCACTGGT-3' hVE-cadherin 5' -AAGCCTCTGATTGGCACAGT- 3' hACTA2 5' -CCGACCGAATGCAGAAGGA- 3' 5' -ACAGAGTATTTGCGCTCCGAA- 3' 5' -TGTCCAAGTTGCTCATCAG- 3' hFSP1 5' -GTCCACCTTCCACAAGTAC- 3' hSNA11 5' -ACCCCAATCGGAAGCCTAACT- 3' 5' -GGTCGTAGGGCTGCTGGAA- 3' hFLI1 5'-GGATGGCAAGGAACTGTGTAA-3' 5'-GGTTGTATAGGCCAGCAG-3' 5' -CATACCAGGAAATGAGCTTGACAA- 3' hGAPDH 5' -ACCCACTCCTCCACCTTTGA- 3'

**Table S1.** The sequences of the primers used for qRT-PCR.

#### Table S2. Primers for ChIP.

Gene	Forward sequence	Reverse sequence
hSELE	5' -ATTTCCAAGGGCCATTTACC- 3'	5' -TTCCTTACCCTCCTCCT- 3'
hSELP	5' -TCTCCAGTGGTTGCTGTTGA- 3'	5' -TTGAGGGACAGTGACTGGTG- 3'
hICAM1	5' -CGGTGTAGACCGTGATTCAA- 3'	5' -GCTGCAGTTATTTCCGGACT- 3'
h <i>FSP1</i>	5' -CCCCTAGCTTTTGTGTCACC- 3'	5' -GGTAACGGGTAAGCCCTAGC- 3'
hSNA11	5' -AGAAGCTACCCTTCGGGAGA- 3'	5' -GCATTGACGAGGGAAACG- 3'