

Supplementary Fig. 1: RP105 negatively correlates with ASMA in SSc and healthy control skin biopsies. **A.** Skin biopsies from SSc (n=7) and healthy controls (n=4) were immunolabeled using antibodies to RP105 (green) and ASMA (red). Representative immunofluorescence images. Dotted line showed the epidermis and dermis intersection. Scale bars: 100 μm , 20 μm , and 5 μm . **B.** Quantitative data represent the mean fluorescent intensity of both RP105 and ASMA measured in same cell from three different fields containing at least 3-4 cells/hpf from the indicated number of subjects. Pearson's rank correlation.

Supplementary Fig. 2: RP105 KO mice showed increased accumulation of leukocytes and myofibroblasts in the skin. RP105 KO mice and wild-type mice received daily s.c. injections of bleomycin or PBS for two weeks (5 days/week). Mice were sacrificed on day 22, and skin was harvested for analysis. **A.** Immunolabelling. Quantitation showing numbers of F4/80-positive cells /hpf (left panel) and CD3-positive cells /hpf (right panel) from four different areas from each skin section from four mice. One-way ANOVA followed by Sidak's multiple comparison test. **B.** Immunohistochemistry using antibodies to ASMA (left panel). Quantitation of immunohistochemistry images represented as the number of positive cells /hpf from four different areas from each skin section of three mice (right panel). One-way ANOVA followed by Sidak's multiple comparison test. **C.** Immunolabelling using antibodies to Fn-EDA and tenascin-C. Representative images. Bar=100 μm .

Supplementary Fig. 3: RP105 KO mice showed increased proinflammatory gene expressions.

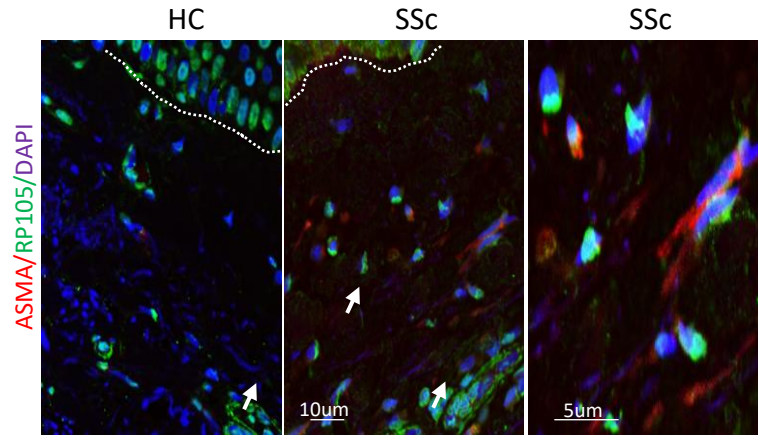
RP105 KO and wild-type mice received daily s.c. injections of bleomycin or PBS for two weeks (5 days/week). Mice were sacrificed on day 22, and RNA isolated from skin was subjected to qPCR analysis. Results, normalized with GAPDH, represent the mean \pm SD of triplicate determinations from four mice. One-way ANOVA followed by Sidak's multiple comparison test.

Supplementary Fig. 4: Spontaneous skin fibrosis in RP105 KO mice.

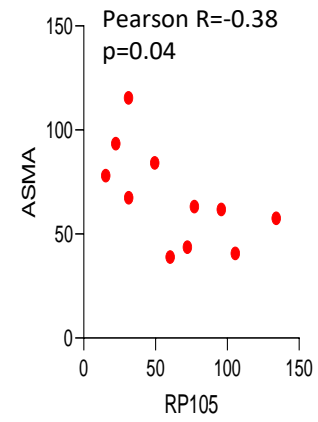
At 6 months of age, female RP105 KO and wild-type mice were sacrificed, and skin tissues were harvested for analysis. **A.** Left panel, H&E stain. Representative images. Bar = 100 μ m. Right panel, dermal thickness (means \pm SD of eight determinations/hpf from the indicated number of mice). The black arrow shows dermal thickness. Mann-Whitney *U* test. **B.** Immunolabelling with antibodies to F4/80. **C.** Skin collagen content. Dots represent the means \pm s.d. from duplicate determination from the indicated number of mice. One-way ANOVA followed by Sidak's multiple comparison test. Bar=100 μ m.

Suppl Fig. 1

A

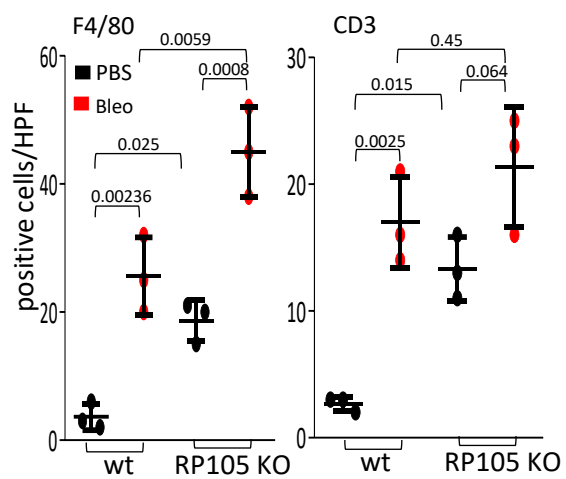


B

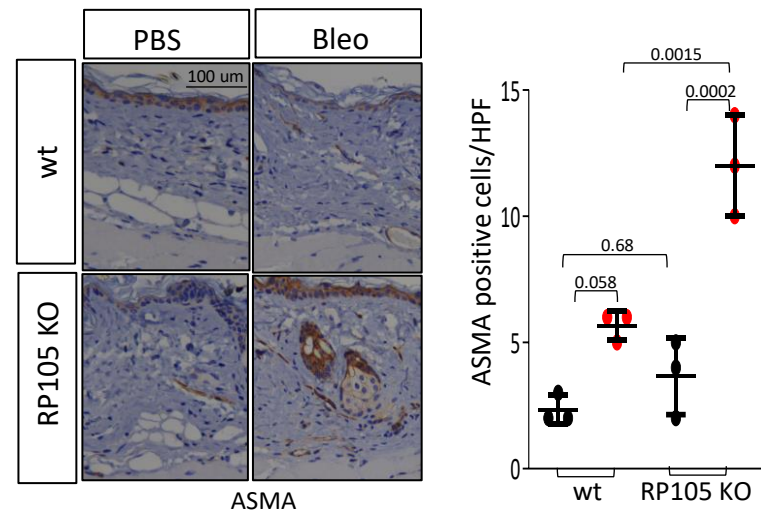


Suppl Fig. 2

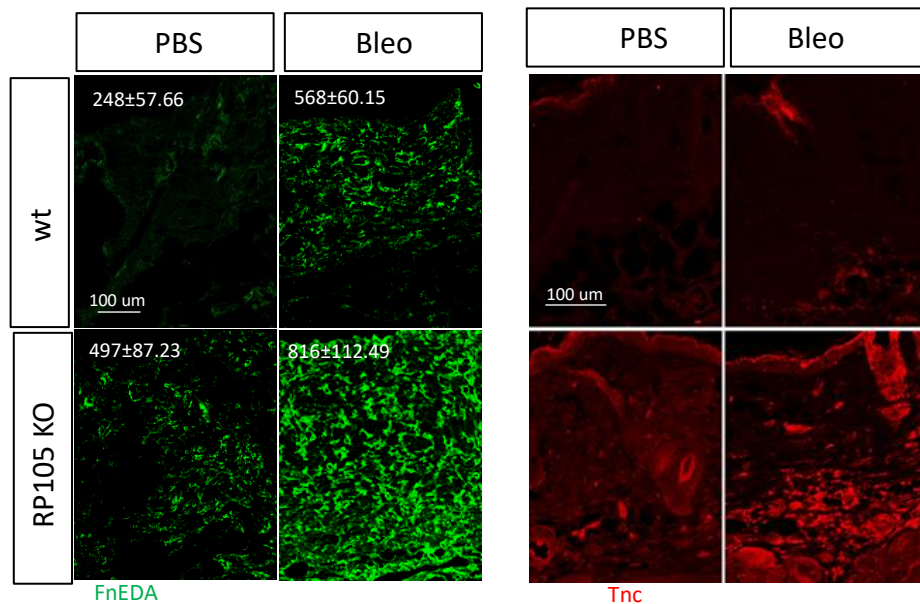
A



B

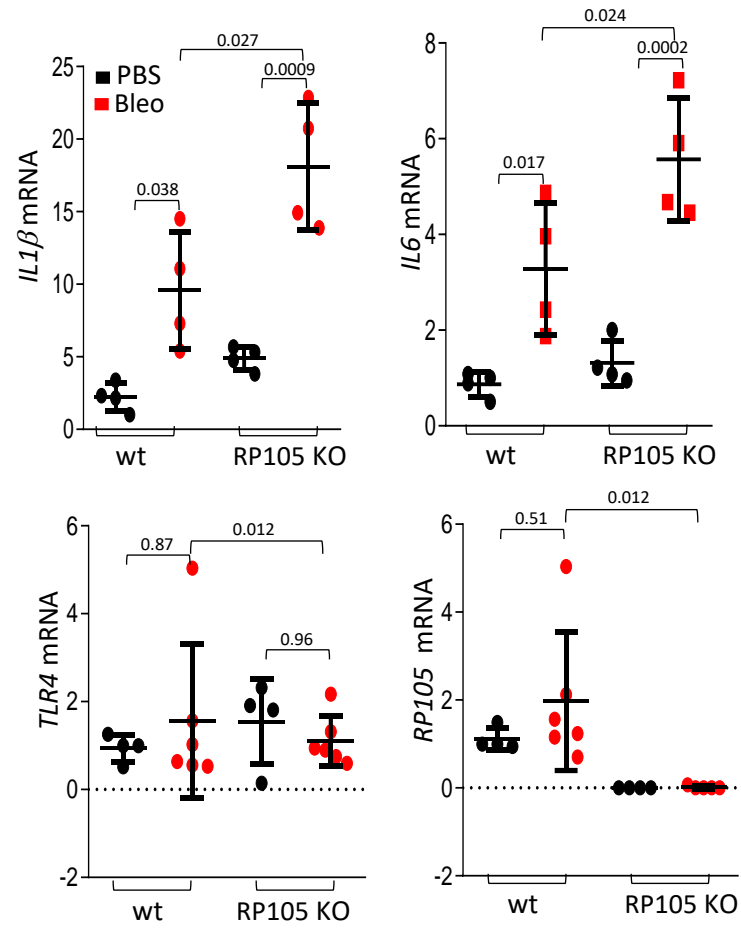


C



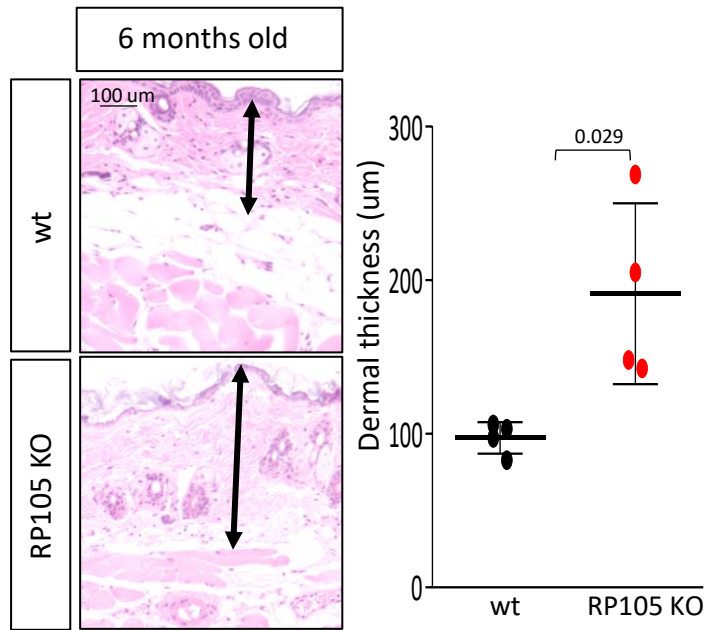
Suppl Fig. 3

Skin mRNA

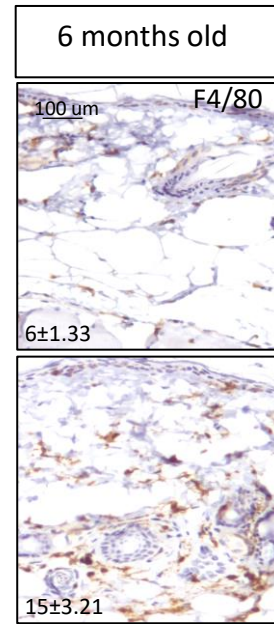


Suppl. Fig. 4

A



B



C

