Supplementary Table S1. Comparison of the characteristics of 94 patients with SSc and ILD based on ANCA positivity.

Variables	SSc-ILD patients Without ANCA (N=74)	SSc-ILD patients With ANCA (N=20)	p value
At the time of diagnosis			
Chest CT findings			
IPF	36 (48.6)	12 (60.0)	0.368
Pulmonary function test			
FVC (% predicted)	70.0 (21.0)	77.0 (45.0)	0.615
DLCO (% predicted)	60.5 (32.0)	69.0 (38.0)	0.254
During follow-up			
Mortality			
All-cause mortality (N, (%))	11 (14.9%)	2 (10.0%)	0.728

Values are expressed as a median (interquartile range, IQR) or N (%).
SSc: systemic sclerosis; ILD: interstitial lung disease; ANCA: antineutrophil cytoplasmic antibody; CT: computed tomography; IPF: idiopathic pulmonary fibrosis; FVC: forced vital capacity; DLCO: diffusing capacity of the lung for carbon monoxide.

Supplementary Table S2. Cox hazards model analysis of variables at diagnosis for all-cause mortality during follow-up in SSc patients.

Variables	Univariable			Multivariable			Multivariable (Including laboratory results)		
	HR	95% CI	p value	HR	95% CI	p value	HR	95% CI	p value
Age (years)	1.079	1.038, 1.123	<0.001	1.092	1.047, 1.139	<0.001	1.081	1.032, 1.132	0.001
Male gender (N, (%))	0.886	0.204, 3.839	0.871						
Skin thickening of the fingers extending MCP joints	0.683	0.259, 1.804	0.442						
Sclerodactyly with/without puffy fingers	1.621	0.615, 4.275	0.329						
Fingertip lesions	0.641	0.186, 2.206	0.480						
Telangiectasia	7.277	0.943, 56.185	0.057	4.500	0.541, 37.412	0.164	5.769	0.559, 59.505	0.141
Abnormal nailfold capillaries	0.448	0.059, 3.401	0.437						
Pulmonary arterial hypertension	3.619	1.467, 8.928	0.005	5.104	2.000, 13.022	0.001	5.393	1.820, 15.981	0.002
Interstitial lung disease	2.069	0.785, 5.450	0.141						
Raynaud's phenomenon	0.878	0.349, 2.210	0.782						
Anti-centromere	0.885	0.293, 2.669	0.828						
Anti-topoisomerase I	1.000	0.401, 2.494	0.999						
ANCA positivity	1.230	0.439, 3.445	0.693						
MPO-ANCA (or P-ANCA) positivity	1.934	0.694, 5.389	0.207						
PR3-ANCA (or C-ANCA) positivity	0.044	0.000, 113.459	0.435						
White blood cell count (/mm^3)	1.201	1.043, 1.382	0.011				1.188	1.013, 1.394	0.035
Haemoglobin (g/dL)	0.723	0.581, 0.900	0.004				1.172	0.828, 1.660	0.370
Platelet count x (1,000/mm^3)	1.002	0.997, 1.008	0.458						
Blood urea nitrogen (mg/dL)	1.068	1.027, 1.110	0.001				1.066	0.972, 1.168	0.174
Serum creatinine (mg/dL)	1.468	1.064, 2.024	0.019				0.981	0.494, 1.946	0.956
Aspartate aminotransferase (IU/L)	1.003	0.999, 1.006	0.135						
Alanine aminotransferase (IU/L)	1.002	0.996, 1.007	0.552						
Total protein (g/dL)	0.586	0.304, 1.130	0.111						
Serum albumin (g/dL)	0.274	0.147, 0.514	<0.001				0.374	0.123, 1.137	0.083

SSc: systemic sclerosis; HR: hazard ratio; CI: confidence interval; ANCA: antineutrophil cytoplasmic antibody; MPO: myeloperoxidase; P: perinuclear; PR3: proteinase 3; C: cytoplasmic.