

Supplementary Table 2. Itemized analysis of SSc patients who did not have ANCA but reclassified as having the overlap syndrome of SSc with AAV according to the 2022 ACR/EULAR criteria for MPA, GPA, or EGPA (n = 3) (patient 3 = GPA, patients 4 and 5 = EGPA)

Patients number	Scores based on the 2022 ACR/EULAR criteria for MPA	1 (-3)	2 (+6)	3 (+3)	4 (+3)	5 (-1)	6 (-4)
3	3	0	0	1	0	0	0
4	-1	0	0	1	0	0	1
5	-4	0	0	0	0	0	1

1 = Nasal involvement (discharge, ulcers, crusting, congestion, septal defect/perforation); 2 = MPO-ANCA (or P-ANCA) positivity; 3 = Fibrosis or interstitial lung disease on chest imaging; 4 = Pauci-immune glomerulonephritis on biopsy; 5 = PR3-ANCA (or C-ANCA) positivity; 6 = Serum eosinophil count $\geq 1,000/\mu\text{L}$.

Patients number	Scores based on the 2022 ACR/EULAR criteria for GPA	1 (+3)	2 (+2)	3 (+1)	4 (+5)	5 (+2)	6 (+2)	7 (+1)	8 (+1)	9 (-1)	10 (-4)
3	5	0	0	0	0	1	1	1	0	0	0
4	-2	0	0	0	0	1	0	0	0	0	1
5	-3	0	0	0	0	0	0	1	0	0	1

1 = Nasal involvement (discharge, ulcers, crusting, congestion, septal defect/perforation); 2 = Cartilaginous involvement; 3 = Conductive or sensorineural hearing loss; 4 = PR3-ANCA (or C-ANCA) positivity; 5 = Pulmonary nodules, mass or cavitation; 6 = Granuloma, granulomatous inflammation, or giant cells on biopsy; 7 = Nasal/paranasal sinusitis or mastoiditis on imaging; 8 = Pauci-immune glomerulonephritis on biopsy; 9 = MPO-ANCA (or P-ANCA) positivity; 10 = Serum eosinophil count $\geq 1,000/\mu\text{L}$.

Patients number	Scores based on the 2022 ACR/EULAR criteria for EGPA	1 (+3)	2 (+3)	3 (+1)	4 (+5)	5 (+2)	6 (-3)	7 (-1)
3	2	1	0	0	0	0	0	1
4	7	1	0	0	1	0	0	1
5	7	0	0	1	1	1	0	1

1 = obstructive airway disease; 2 = nasal polyps; 3 = mononeuritis multiplex; 4 = Serum eosinophil count $\geq 1,000/\mu\text{L}$; 5 = Extravascular eosinophilic-predominant inflammation on biopsy; 6 = PR3-ANCA (or C-ANCA) positivity; 7 = hematuria.

SSc, systemic sclerosis; ILD, interstitial lung disease; ACR, American College of Rheumatology; EULAR, European Alliance of Associations for Rheumatology; MPA, microscopic polyangiitis; GPA, granulomatosis with polyangiitis; EGPA, Eosinophilic granulomatosis with polyangiitis; MPO, myeloperoxidase; P, perinuclear; PR3, proteinase 3; ANCA, antineutrophil cytoplasmic antibody; C, cytoplasmic.