

ONLINE SUPPLEMENTARY MATERIAL.

Supplementary Table 1. Categories of comorbidities.

airway disease	
allergic disease	bronchial asthma, allergic rhinitis
upper airway disease	chronic rhinitis, chronic otitis media, history of sinus surgery, severe caries necessitating extraction
lower airway disease	bronchiectasis, pulmonary infections, emphysema, fibrotic interstitial lung disease (NSIP, UIP)
infectious airway disease	upper airway disease, bronchiectasis, pulmonary infections (history of pulmonary tuberculosis, pulmonary non-tuberculosis mycobacterium infections, fungal infections)
non-infectious airway disease	emphysema, fibrotic interstitial lung disease
heart disease	atrial fibrillation, heart valve disease, chronic heart failure
atherosclerotic disease	hypertension, diabetes, ischemic heart disease, stroke, arteriosclerosis obliterans/aneurysm
others	history/presence of malignancy

Supplementary Table 2. Vasculitis involvements of AAV patients at diagnosis.

	Total	EGPA	GPA	MPA	uAAV	p value
cases	195	24	51	89	31	among AAV
female(%)	58.0%	54.2%	62.8%	55.1%	61.3%	>0.1
age (y.o. mean±SD)	69±12	57±13	68±12	71±11	75±9	<0.0001
MPO-ANCA (%)	75.9%	41.7%	58.8%	91.0%	87.1%	<0.0001
PR3-ANCA (%)	17.4%	8.3%	39.2%	10.1%	9.7%	0.0001
BVAS (mean±SD)	15.4±7.9	17.9±8.5	16.5±9.8	15.9±6.4	10.1±5.7	0.0002
general	85.1%	75.0%	88.2%	85.4%	87.1%	>0.1
muscle joint symptom	54.9%	75.0%	51.0%	52.8%	51.6%	>0.1
skin	25.6%	54.2%	17.7%	27.0%	12.9%	0.0032
ENT	34.3%	41.7%	90.2%	7.9%	12.9%	<0.0001
heart	3.6%	12.5%	3.9%	1.1%	3.2%	>0.1
lung	39.0%	45.8%	62.8%	25.8%	32.3%	0.02
pulmonary hemorrhage	4.1%	0%	5.9%	4.5%	3.2%	>0.1
kidney	59.5%	25.0%	43.1%	85.4%	38.7%	<0.0001
abdominal	2.0%	8.3%	2.0%	0%	0%	0.0605
central nervous system	7.2%	4.2%	15.7%	3.4%	6.5%	0.071
pachymeningitis	2.6%	0%	9.8%	0%	0%	0.0032
sensory neuropathy	42.6%	91.7%	31.4%	39.3%	32.3%	<0.0001
motor neuropathy	14.9%	66.7%	7.8%	10.1%	0%	<0.0001
eGFR (ml/min, mean±SD)	65±33	96±30	71±31	53±31	67±24	<0.0001
use of CY	34.7%	26.1%	33.3%	41.6%	23.3%	>0.1
use of RTX	11.9%	0%	21.6%	12.4%	3.3%	0.0059

Supplementary Table 3. Vasculitis involvements of MPO-ANCA-positive AAV patients at diagnosis.

	EGPA	GPA	MPA	uAAV	p value
cases	10	27	81	25	
female	50.0%	66.7%	55.6%	64.0%	>0.1
age (y.o. mean±SD)	62.0±14.0	70.0±8.9	71.7±10.8	76.0±8.1	0.0054
BVAS score (mean±SD)	18.9±5.5	17.9±8.5	16.3±6.4	10.3±5.9	0.0003
general	100%	88.9%	87.7%	88.0%	>0.1
muscle joint symptom	100%	40.7%	51.9%	52.0%	0.0024
skin	60.0%	22.2%	25.9%	16.0%	0.080
ENT	50.0%	92.6%	7.4%	16.0%	<0.0001
heart	10.0%	0%	1.2%	4.0%	>0.1
lung	50.0%	59.3%	28.4%	32.0%	0.0279
pulmonary hemorrhage	0%	3.7%	4.9%	4.0%	>0.1
kidney	30.0%	44.4%	87.7%	40.0%	<0.0001
abdominal	0%	0%	0%	0%	>0.1
central nervous system	10.0%	14.8%	3.7%	8.0%	>0.1
pachymeningitis	0%	14.8%	0%	0%	0.0031
sensory neuropathy	90.0%	33.3%	38.3%	36.0%	0.0082
motor neuropathy	60.0%	3.7%	8.7%	0%	<0.0001

ENT: ear, nasal and tracheal involvement; eGFR: estimated glomerular flow rate; Statistically significant values are shown in bold ($p < 0.05$).