

## Supplementary Files

**Supplementary Table 1.** Comparison of serum sPD-1 based on concurrently administered immunosuppressive drugs in AAV patients.

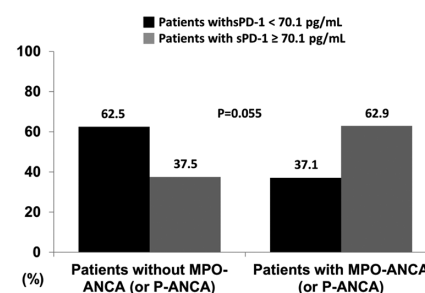
Drugs	Administration of drugs	Serum sPD-1 (pg/mL)	<i>p</i> -value
<i>Prednisolone</i>	Yes (N=43)	204.6 ± 379.5	0.647
	No (N=16)	252.1 ± 262.5	
<i>Cyclophosphamide</i>	Yes (N=9)	416.7 ± 585.3	0.269
	No (N=50)	181.6 ± 283.8	
<i>Rituximab</i>	Yes (N=1)	505.9	0.411
	No (N=58)	212.5 ± 351.1	
<i>Azathioprine</i>	Yes (N=14)	84.7 ± 122.9	0.011
	No (N=45)	258.8 ± 387.0	
<i>Tacrolimus</i>	Yes (N=3)	533.1 ± 881.8	0.581
	No (N=56)	200.6 ± 308.6	
<i>Mycophenolate mofetil</i>	Yes (N=2)	775.5 ± 1,096.7	0.592
	No (N=57)	197.9 ± 306.5	
<i>Methotrexate</i>	Yes (N=2)	336.1 ± 29.1	0.630
	No (N=57)	213.3 ± 355.6	

sPD-1: soluble programmed cell death protein 1; AAV: ANCA-associated vasculitis; ANCA: antineutrophil cytoplasmic antibody.

**Supplementary Table 2.** Comparison of serum sPD-1 based on clinical manifestations in AAV patients .

Clinical manifestations	Presence of absence	Serum sPD-1 (pg/mL)	<i>p</i> -value
<i>General</i>	Presence (N=16)	184.7 ± 208.7	0.665
	Absence (N=43)	229.7 ± 391.3	
<i>Cutaneous</i>	Presence (N=7)	482.1 ± 572.2	0.218
	Absence (N=52)	181.9 ± 300.0	
<i>Mucomembranous/ocular</i>	Presence (N=3)	40.5 ± 70.2	0.374
	Absence (N=56)	227.0 ± 356.8	
<i>Ear nose throat</i>	Presence (N=25)	179.6 ± 233.3	0.480
	Absence (N=34)	245.4 ± 417.1	
<i>Pulmonary</i>	Presence (N=38)	220.7 ± 355.2	0.926
	Absence (N=21)	211.7 ± 349.4	
<i>Cardiovascular</i>	Presence (N=3)	365.8 ± 317.4	0.456
	Absence (N=56)	209.5 ± 352.6	
<i>Abdominal</i>	Presence (N=1)	503.1	0.415
	Absence (N=58)	212.6 ± 351.1	
<i>Renal</i>	Presence (N=32)	279.7 ± 368.4	0.139
	Absence (N=27)	143.7 ± 318.1	
<i>Nervous systemic</i>	Presence (N=12)	102.4 ± 154.9	0.204
	Absence (N=47)	246.9 ± 380.1	

sPD-1: soluble programmed cell death protein 1; AAV: ANCA-associated vasculitis; ANCA: antineutrophil cytoplasmic antibody.

**Supplementary Fig. 1.** Relative risk of MPO-ANCA (or P-ANCA) positivity for serum sPD-1 ≥70.1 pg/mL.

MPO-ANCA (or P-ANCA) positivity showed a tendency of contributing the elevated level of serum sPD-1 in AAV patients, however, it was not statistically significant. MPO: myeloperoxidase; ANCA: antineutrophil cytoplasmic antibody; P: perinuclear; sPD-1: soluble programmed cell death protein 1; AAV: ANCA-associated vasculitis.