

Supplementary Table 1. presents the clinical characteristics of AAV patients and three normal controls, along with the follow-up information of certain patients.

	C01	C02	C03	P01	P02	P03	P04	P05	P06	P07	P08	P09	P10	P11	P12
Age(years)	10	12	12	8.3	8.9	11.6	11.3	5	13.3	14.2	7.4	17.6	19	65	72
General(female/male)	M	M	F	F	M	F	F	F	F	M	M	F	F	F	M
BVAS/PAVAS															
ANCA levels				MPO+++	MPO++	MPO++	MPO+++	MPO+	MPO+	PR3++	PR3++	PR3++	MPO+++	PR3+++	PR3+++
General				1	0	0	0	0	0	0	3	3	1	1	0
Cutaneous				2	0	0	0	0	0	0	0	0	0	0	0
Mucous membranes & Eyes				1	0	0	0	0	0	0	1	0	0	0	0
Ear & Nose & Throat				0	0	0	4	0	0	0	0	0	6	0	2
pulmonary				6	6	6	0	6	0	0	0	0	6	4	4
Cardio-vascular				0	0	0	0	0	0	0	0	0	4	0	0
Abdominal				0	0	0	0	0	0	0	0	6	0	0	0
Renal				0	12	0	6	0	12	10	6	0	0	0	0
Nervous system				0	0	0	0	0	0	0	0	0	0	0	0
Others				0	0	0	0	0	0	0	0	0	0	0	0
Total				10	18	6	6	10	12	10	10	9	17	5	6
C-reactive protein(mg/L)				<8	<8	<8	<8	<8	<8	<8	<8	<8	10.06	18.95	21.91
Creatinine (μmol/L)				37	266	47	42	29	955.8	81	38	58	54	37	1179
eGFR(ml/min/1.73m2)				176.08	25.04	155.17	178.27	179.39	6.61	103.84	166.33	128.26	238.71	144.55	185.22
24-Hours proteinuria(g)				0.134	0.468	0.004	0.011	0.024	/	7.746	0.023	0.03	/	/	/
Urine protein				-	2+	-	-	-	3+	2+	-	-	-	-	/
URBC				0	924	0	28	0	92.6	260	52	14	/	/	/
GGT(U/L)				14	6	6	/	13	/	28	6	/	/	/	/
Immunosuppressant induction treatment															
Glucocorticoids				+	+	+	/	+	/	+	+	/	/	/	/
Rituximab					+	+	/		/			/	/	/	/
Cyclophosphamide				+	+		/	+	/	+		/	/	/	/
Plasmapheresis				+	+	+	/		/	+	+	/	/	/	/
MMF							/		/		+	/	/	/	/
Last follow-up information of patients after treatment															
C-reactive protein(mg/L)				<8	<8	<8	/	<8	/	<8	<8	/	/	/	/
Creatinine (μmol/L)				37	168	12.96	/	39	/	20	21	/	/	/	/
eGFR(ml/min/1.73m2)				193.17	40.52	600.25	/	142.12	/	149.27	142.82	/	/	/	/
24-Hours proteinuria(g)				0.073	0.826	-	/	0.048	/	0.105	0.346	/	/	/	/
Urine protein				-	+	-	/	-	/	-	±	/	/	/	/
URBC				6	33	0	/	3	/	13	0	/	/	/	/
GGT				25	15	10	/	14	/	34	11	/	/	/	/

Notes: C, Control; P, Patient; /, not applicable; ANCA, anti-neutrophil cytoplasmic autoantibody; MPO, myeloperoxidase; PR3, proteinase 3; -, Negative; URBC, Urinary Red Blood Cell Count; eGFR, Estimated glomerular filtration rate; GGT, Gamma-glutamyl transpeptidase; MMF, Mycophenolate mofetil.

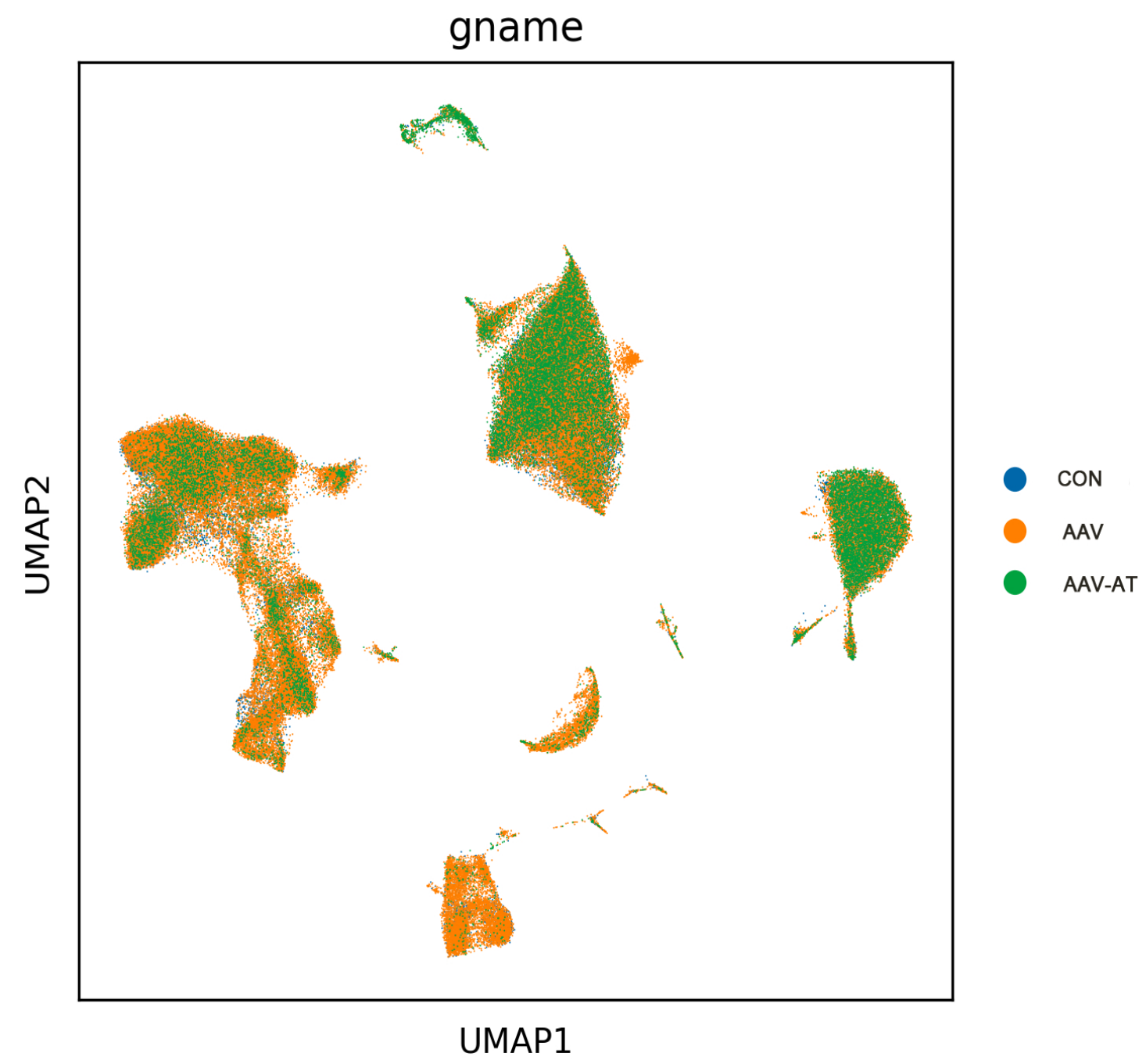
Single-cell blood samples in the after-treatment group were derived from treated patients (P01, P02, P03). P01, P03, and P05 were samples of the MPO clinical phenotype without kidney injury; P02, P04, and P06 were those with the MPO clinical phenotype and kidney injury. Samples P07-P09 were PR3-ANCA-positive cases, while P10-P12 were adult AAV specimens.

Supplementary Table 2. Clinical information of AAV patients treated in our hospital, excluding those whose samples were used for single - cell sequencing.

	P01	P02	P03	P04	P05	P06	P07	P08	P09	P10	P11	P12	P13	P14	P15	P16	P17	P18	P19	P20	P21
Age(years)	12	5	13	11.8	11.2	12	2.5	6.9	11.7	6	13	12.9	14.9	9.8	7.8	12.4	12.9	3	4.6	14.7	9.9
General(female/male)	M	M	F	F	F	F	F	M	F	M	F	F	F	M	M	F	F	M	F	F	M
ANCA levels	MPO++	MPO+	MPO+	MPO+	MPO+	MPO+	GMB+	PR3+	MPO+	MPO+++	MPO+	PR3+++	MPO+++	PR3+++	PR3+	MPO+++	MPO+	MPO+++	MPO+++	MPO+	MPO+
BVAS/PAVAS																					
General	0	0	0	3	1	1	1	0	0	0	2	1	2	1	0	0	0	0	0	0	1
Cutaneous	5	1	0	0	1	1	1	0	0	0	1	0	0	0	0	0	1	0	1	0	0
Mucous membranes & Eyes	2	0	0	0	0	0	0	0	0	0	0	0	2	0	0	0	0	0	0	0	0
Ear & Nose & Throat	0	0	0	4	0	0	0	0	0	0	2	4	2	2	0	4	0	0	0	0	0
pulmonary	0	0	0	0	0	0	0	0	0	4	6	6	0	0	0	0	0	6	0	6	0
Cardio-vascular	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Abdominal	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Renal	12	12	10	12	6	6		10	12	6	12	12	6	6	12	12	6	6	12	6	6
Nervous system	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Others	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Total	19	13	10	19	8	8	2	10	12	10	23	27	12	9	12	12	7	12	13	12	7
C-reactive protein(mg/L)	8	11	<8	8	18	43	8	<8	8	<8	11	<8	31	<8	8.72	<8	<8	9.87	<8	/	11.61
Creatinine (μmol/L)	91	438	96	151	34	49	27	25	154	42	132	53	49	36	160	260	62	24	37	46	43
eGFR(ml/min/1.73m2)	80.14	11.32	82.55	49.91	200.2	143.88	151.26	246.99	48.94	127.33	55.99	146.78	150.82	191.78	41.02	28.42	130.96	198.53	139.29	170.17	163.95
24-Hours proteinuria(g)	1.203	0.09	8.66	1.386	0.26	/	/	/	5.08	0.25	2.12	0.177	0.272	0.107	1.107	3.438	0.291	/	0.394	0.516	0.289
Urine protein	1+	3+	3+	2+	1+	1+	-	3+	2+	1+	2+	-	1+	-	2+	3+	-	-	2	-	1+
URBC	630	4307	112	1146	33	10	0	2948	430	44	74	483	39	30	13662	1134	41	139	864	0	543
GGT(U/L)	8	7	21	10	14	12.3	9	6	14.2	9	6	45	10	10	6	9	10	10	7	6	12
Immunosuppressant induction treatment																					
Glucocorticoids	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+
Rituximab	+	+		+				+						+	+	+	+	+	+	+	+
Cyclophosphamide	+	+	+	+		+			+	+	+	+	+			+				+	
Plasmapheresis	+			+	+					+	+	+				+		+			
MMF				+					+												
Last follow-up information of patients after treatment																					
C-reactive protein(mg/L)	<8	/	<8	<8	<8	/	/	<8	/	<8	<8	<8	23	<8	<8	<8	<8	<8	<8	<8	<8
Creatinine (μmol/L)	101	132	47	47	46	99.2	29	33	218	43	110	72	48	35	50	381	63	34	32	44	44
eGFR(ml/min/1.73m2)	103.54	47.88	170.69	165.51	163.83	71.07	144.18	191.53	34.57	133.42	66.3	109.39	153.96	201.42	131.27	19.39	128.88	143	161	177.91	160.25
24-Hours proteinuria(g)	1.38	/	1.1	0.121	0.15	0.58	0.03	/	/	0.131	2.533	/	0.03	0.046	/	6.624	0.172	/	0.45	0.146	0.218
Urine protein	1+	-	-	2+	-	1+	-	-	3+	-	1+	1+	-	-	-	3+	-	-	-	-	-
URBC	0	1	0	73	0	1	7	2070	36	0	12	8	7	0	0	438	0	0	0	0	6
GGT	10	29	79	15	10	15	10	11	17.9	13	13	76	21	16	11	15	11	19	16	9	19

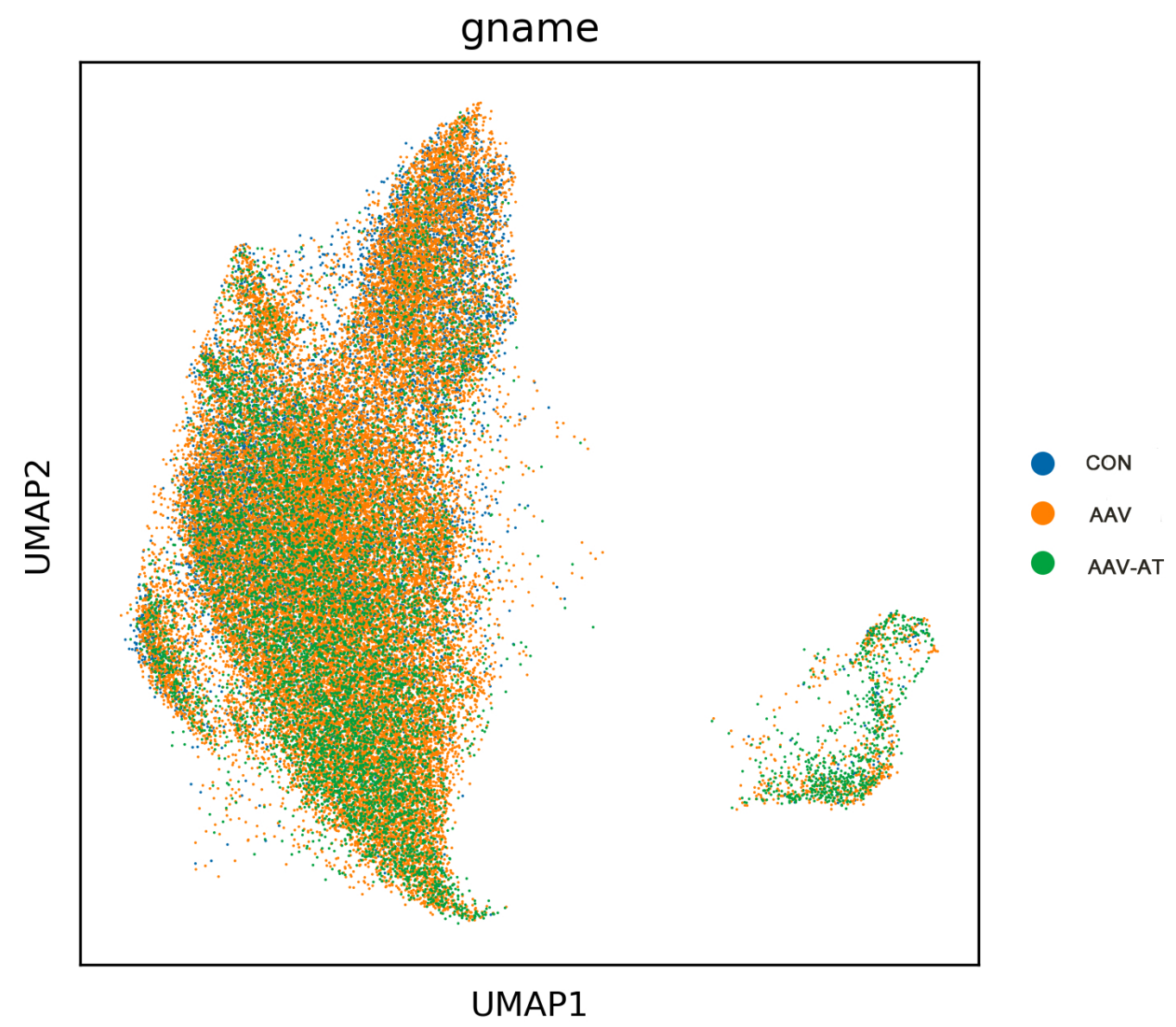
Notes: P, Patient; /, not applicable; ANCA, anti-neutrophil cytoplasmic autoantibody; MPO, myeloperoxidase; PR3, proteinase 3;GBM,Glomerular basement membrane; -, Negative; URBC, Urinary Red Blood Cell Count; eGFR, Estimated glomerular filtration rate; GGT, Gamma-glutamyl transpeptidase; MMF, Mycophenolate mofetil.

Supplementary Figure 1. UMAP plots of immune cells derived from 18 samples.



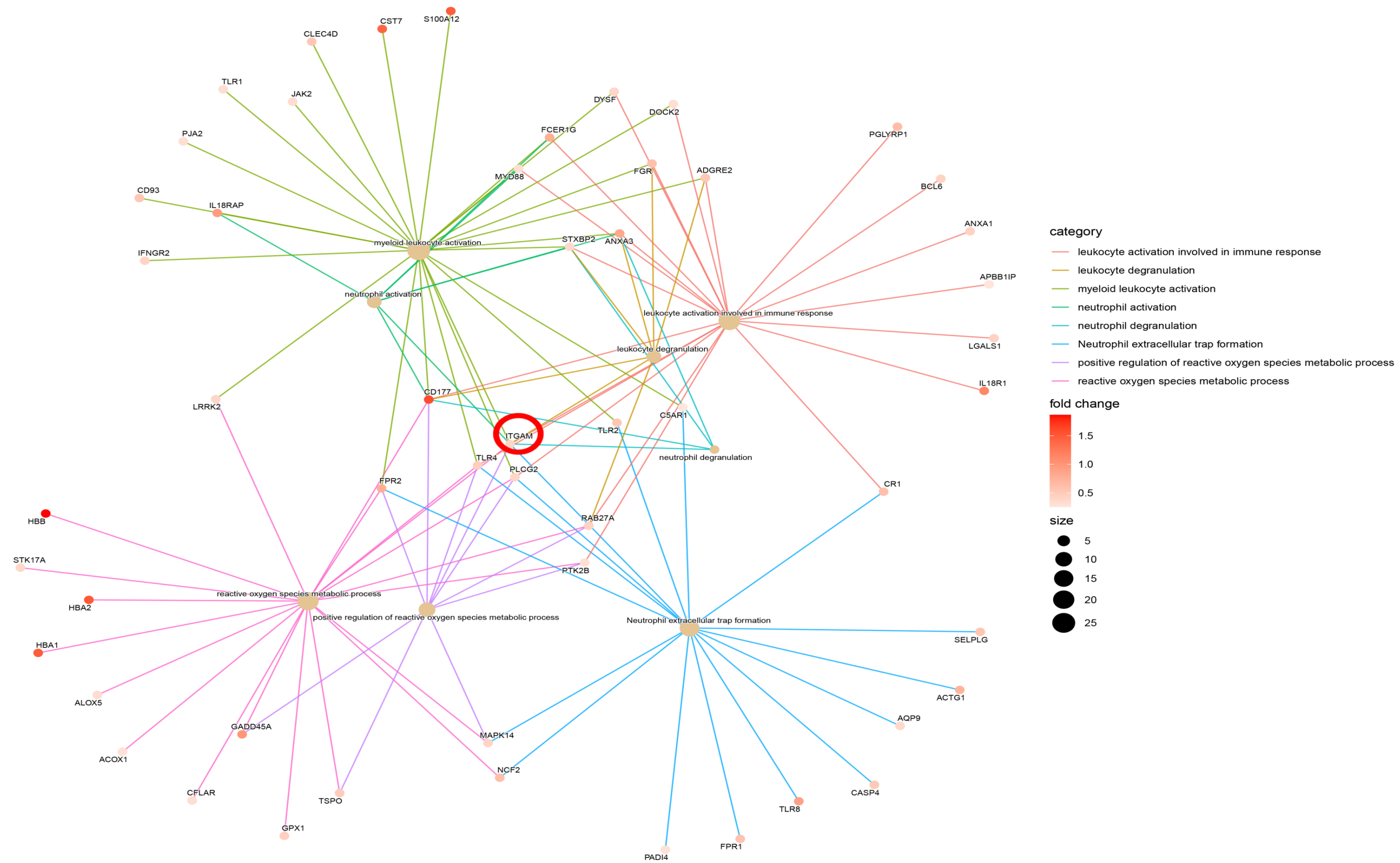
The UMAP displays single-cell data of immune cells obtained from 18 samples(CON group: Blue; AAV group:Yellow; AAV-AT: Green).

Supplementary Figure 2. UMAP plots of neutrophil cells derived from 18 samples.



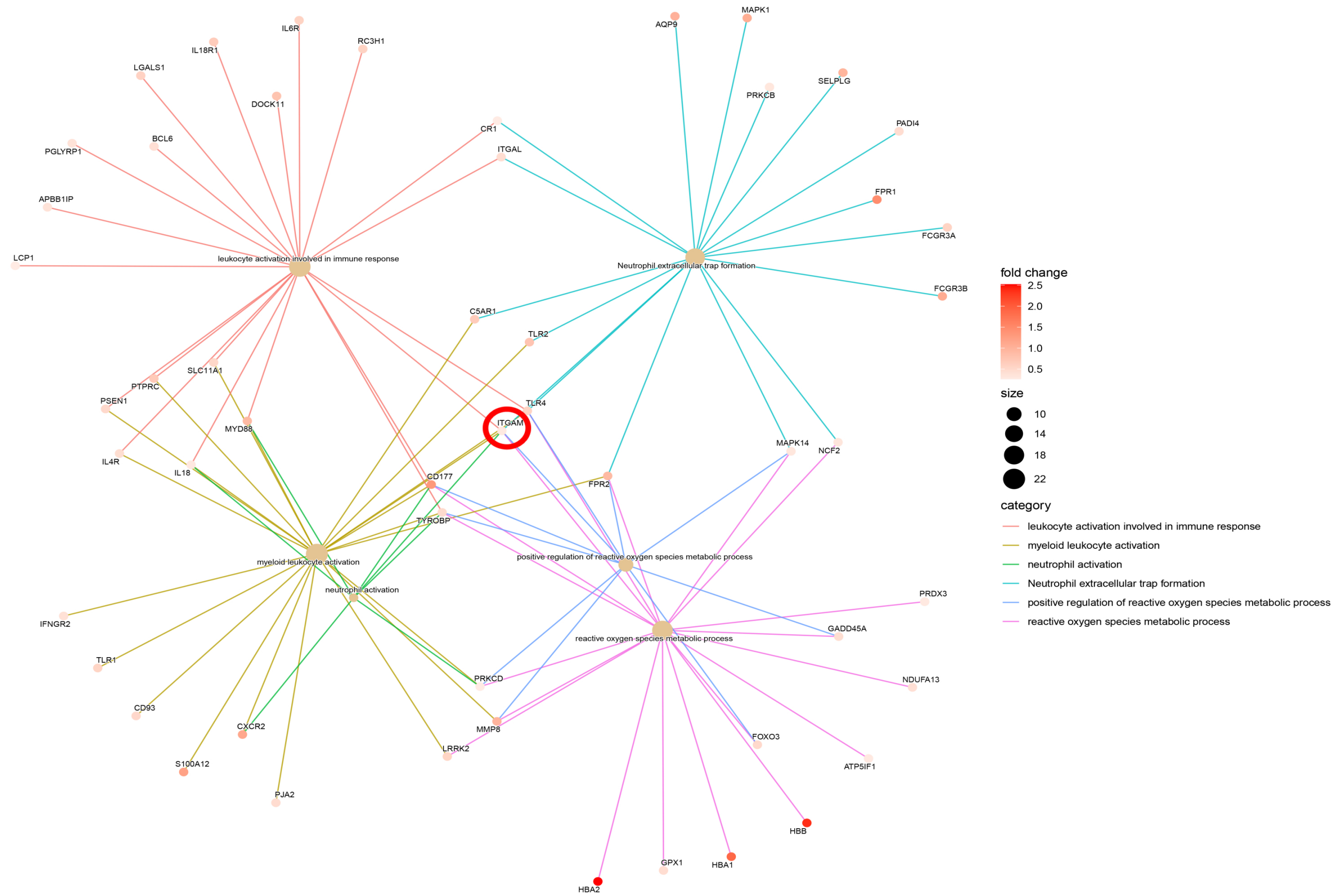
The UMAP displays single-cell data of neutrophil cells obtained from 18 samples(CON group: Blue; AAV group:Yellow; AAV-AT: Green).

Supplementary Figure 3. Association network diagram for enrichment analysis of differentially expressed genes by comparing the CON and AAV groups of clusters (CD10⁺CD11B⁺⁺neutrophils).



The large dots represent pathways, with the dot size indicating the number of genes. The small dots represent genes, and the redder the color, the higher the expression level. The connecting lines indicate that a gene belongs to the corresponding pathway. The area circled in red highlights the gene ITGAM.

Supplementary Figure 4. Association network diagram for enrichment analysis of differentially expressed genes by comparing the CON and AAV groups of clusters (CD10-CD11B⁺⁺neutrophils).



The large dots represent pathways, with the dot size indicating the number of genes. The small dots represent genes, and the redder the color, the higher the expression level. The connecting lines indicate that a gene belongs to the corresponding pathway. The area circled in red highlights the gene ITGAM.

Supplementary Figure 5. Flow cytometry plots and fluorescence data of peripheral blood samples disease model group (N=3) are provided.

