

Supplementary Material

Supplementary Table S1. Comparison of clinical characteristics between the ANCA-positive LN group and ANCA-positive non-LN group

	ANCA -positive LN N=25	ANCA -positive non-LN N=21	<i>P</i>
Age (years)	45.73 ± 14.17	41.52 ± 13.97	0.314
Male, n (%)	1 (3.8)	1 (4.0)	0.699*
Time-gap from diagnosis of SLE to ANCA positive (months)	15.00 (5.50, 96.00)	12.00 (1.00, 30.00)	0.177
WBC (*10 ⁹ /L)	3.97 (2.76, 5.55)	4.25 (2.75, 5.99)	0.850
HGB (g/L)	91.54 ± 22.70	100.95 ± 14.57	0.118
Serum albumin (g/L)	36.21 ± 13.03	39.21 ± 9.74	0.098
CRP (mg/L)	3.00 (3.00, 19.15)	9.90 (9.90, 18.18)	0.810
ESR (mm/h)	60.25 ± 36.88	53.35 ± 32.80	0.601
CREA (umol/L)	107.95 (49.75, 298.25)	43.00 (37.51, 53.00)	0.001
eGFR (mL/min/1.73m ²)	54.48 (14.81, 114.15)	123.42 (107.44, 137.16)	0.001
24h urine protein (g)	1.36 (0.66, 3.39)	0.10 (0.05, 0.25)	0.000
IgG, g/L	18.42 ± 9.79	23.14 ± 10.19	0.118
IgM, g/L	0.85 (0.44, 1.47)	1.39 (1.10, 1.91)	0.007
IgA, g/L	2.99 ± 1.36	3.35 ± 1.59	0.416
C3, g/L	0.54 ± 0.27	0.75 ± 0.26	0.011
C4, g/L	0.14 ± 0.09	0.13 ± 0.07	0.946
Anti-dsDNA, n (%)	12 (50.0)	9 (60.0)	0.542
Anti-dsDNA titer (IU/mL)	6.63 (2.29, 100.00)	1.79 (0.98, 16.41)	0.185
Anti-SmD1, n (%)	11 (47.8)	9 (60.0)	0.469
Anti-ANA, n (%)	15 (57.7)	14 (66.7)	0.529
Anti-RNP, n (%)	4 (28.6)	10 (66.7)	0.040
Anti-MPO, n (%)	11 (44.0)	5 (23.8)	0.152
Anti-PR3, n (%)	14 (56.0)	16 (76.2)	0.152
Anti-MPO titer (RU/mL)	73.63 (63.84, 105.82)	39.90 (24.71, 122.30)	0.117
Anti-PR3 titer (RU/mL)	55.20 (31.86, 72.55)	71.94 (35.90, 131.42)	0.188

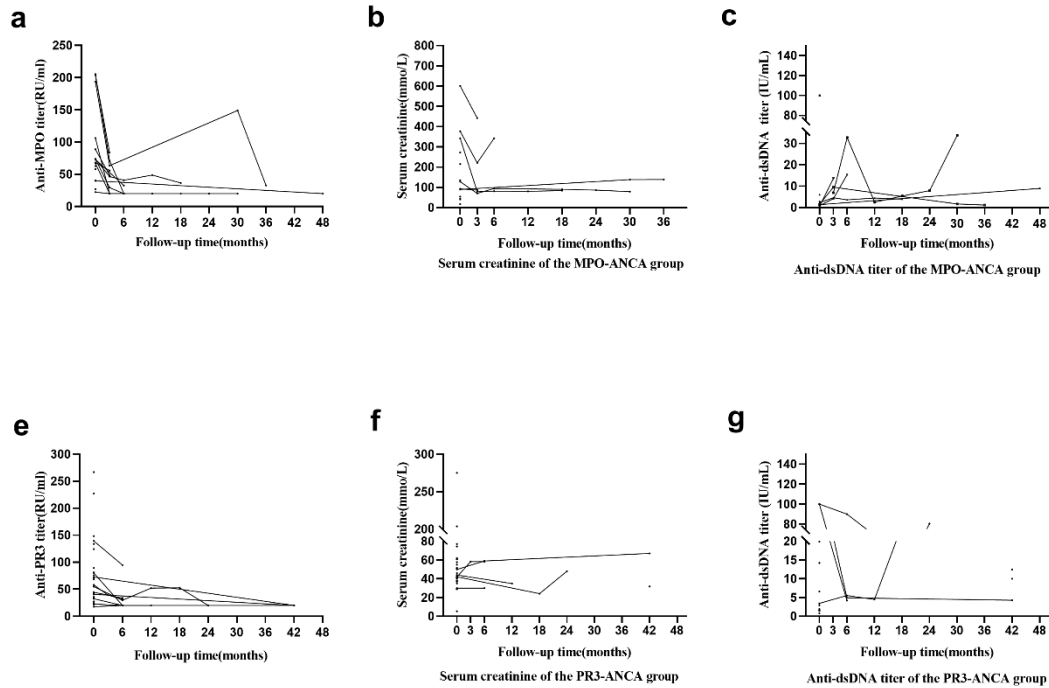
*Fisher test; WBC: white blood cell; HGB: hemoglobin; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; CREA: serum creatinine; eGFR: estimated glomerular filtration rate; IgG: serum immunoglobulin G, Normal values: 7-16g/L; IgA: serum immunoglobulin A, Normal values:0.7-3.8g/L; C3: complement 3, Normal values: 0.8-1.85g/L; C4: complement 4; Normal values: 0.1-0.4 g/L; ds-DNA: double stranded DNA antibodies; Normal values:0-7IU/ml; SmD1: Smith antigen D1; ANA: antinuclear antibody; MPO: myeloperoxidase; PR3: proteinase 3; Anti-MPO titer: Normal values:< 20 RU/ml; Anti-PR3 titer: Normal values:< 20 RU/ml;

Supplementary Table S2. Immune complex deposition in ANCA positive LN and ANCA negative LN

Direct immunofluorescence microscopy	ANCA positive LN	ANCA negative LN	<i>P</i>
IgG	1.5 (0, 2)	3 (2, 3)	0.013
IgM	2 (1, 2)	2 (1, 2)	0.717
IgA	2 (1, 2)	2 (1, 2)	0.627
C3	3 (2, 3)	2.5 (2, 3)	0.676
C1q	2 (1, 3)	2 (2, 3)	0.648

- Follow-up of ANCA titers, anti-dsDNA titers, CREA in ANCA positive SLE

The change in ANCA titers during treatment in ANCA positive SLE was monitored, and the anti-MPO and anti-PR3 titers in most of the patients decreased to within normal range (20 RU/mL) within six months (Supplementary Figure S1a, S1e). In one patient, high MPO titers recurred one year after self-discontinuation of treatment (Figure S1a). The anti-ds-DNA titers and serum CREA level were also collected during the same period (Supplementary Figure S1b, S1c, S1f, S1g).

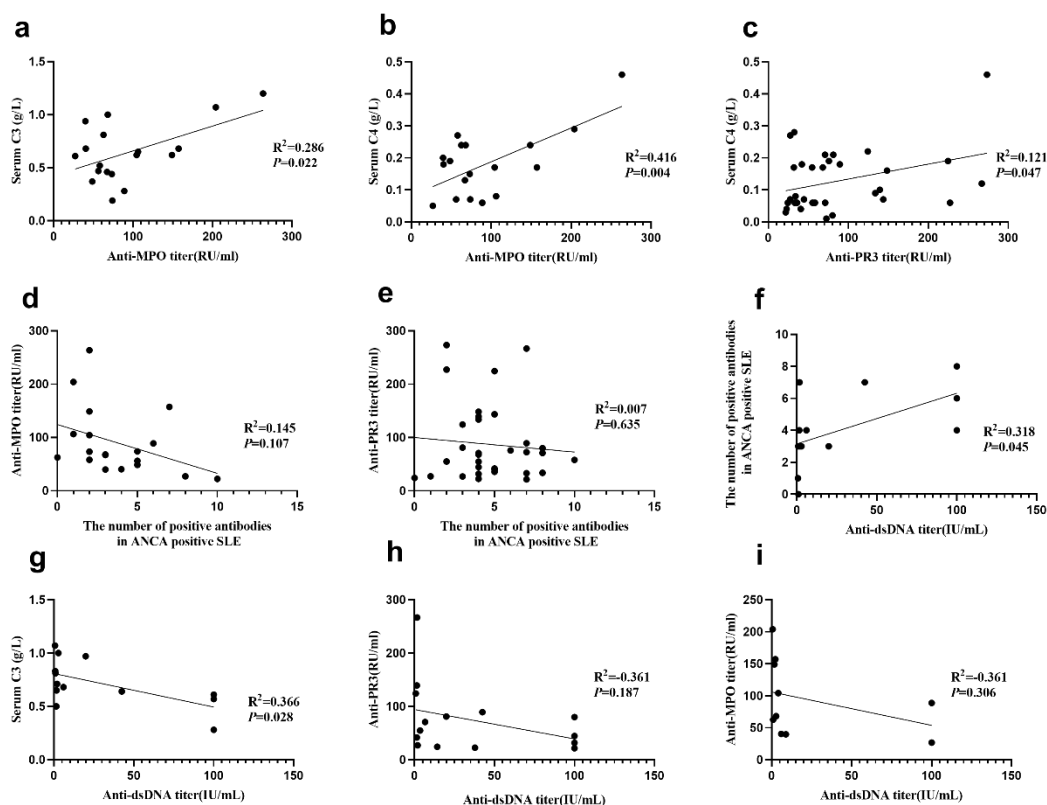


Supplementary Figure S1. Changes in the CREA level and ANCA and anti-dsDNA titers during follow-up (a) changes of anti-MPO titers in SLE with ANCA-positive patients. (b) changes of CREA in MPO-ANCA group. (c) changes of anti-dsDNA titers in MPO-ANCA group. (e) changes of anti-PR3 titers in SLE with ANCA-positive patients. (f) changes of CREA in PR3-ANCA group. (g) changes of anti-dsDNA titers in PR3-ANCA group.

- ANCA serology and correlation of ANCA titers and clinical characteristics

Anti-MPO titers were positively correlated with levels of complement C3 and C4. Anti-PR3 titers positively correlated with levels of complement C4. However, there was no correlation between the ANCA titers and antibody seropositivity in the ANCA-

positive group. Meanwhile, ds-DNA titers were positively correlated with antibody seropositivity in the ANCA-positive group (Supplementary Figure S2).



Supplementary Figure S2. Correlation of ANCA titers and clinical characteristics. (a) the correlation of anti-MPO titers with serum C3. (b) the correlation of anti-MPO titers with serum C4. (c) the correlation of anti-PR3 titers with serum C4. (d) the correlation of positive antibodies numbers in SLE with anti-MPO titers. (e) the correlation of positive antibodies numbers in SLE with anti-PR3 titers. (f) the correlation of positive antibodies numbers in ANCA positive SLE group with anti-dsDNA titers. (g) the correlation of anti-dsDNA titers with serum C3. (h) the correlation of anti-dsDNA titers with anti-MPO titers. (i) the correlation of anti-dsDNA titers with anti-PR3 titers.

