

**Table S1** Base line clinical characterizations of the patients with MPGN

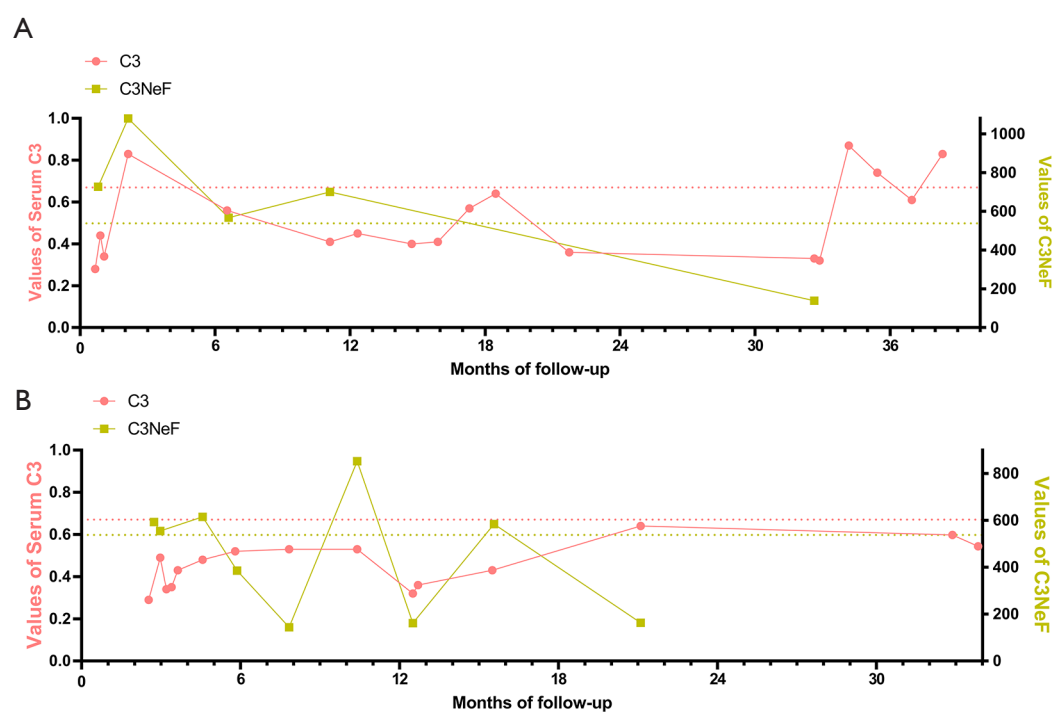
Patient No.	Age (y) / Sex	Prior infection	Edema	Hypertension	Hematuria (/ HPF)	24-hour urinary protein (g)	P/C	Lipstick of proteinuria	Serum albumin (g/L)	Other renal/extrarenal manifestations	Positive family history	LM/EM
1	9/F	(-)	(+)	(-)	(-)	8.53	NA	3+	↓ <sup>a</sup>			Glomeruli hyalinized, mesangial and endocapillary proliferation, crescents, tubular atrophy/NA
2	4/M	Dermal	(+)	(-)	71.3	2.57	8.92	3+	27.2			Mesangial and endocapillary proliferation, individual crescents, 10% glomeruli hyalinized, tubular atrophy/Mild mesangial electron-dense deposits
3	5/M	Dermal	(-)	(-)	135	1.53	2	2+	30.8			Mesangial and endocapillary proliferation, mild interstitial fibrosis, segmental sclerosis, individual crescents, double contours of GBM/ Mesangial, subendothelial and rare subepithelial electron-dense deposits
4	11/M	(-)	(+)	(-)	112	2.86	0.62	2+	38		Consanguineous marriage; A cousin had nephrotic syndrome	Mesangial and endocapillary proliferation/Mild mesangial electron-dense deposits
5	6/M	(-)	(-)	(-)	12.32	0.26	0.81	2+	43.8	Cataract; intellectual disability; hearing impairment		Mesangial and endocapillary proliferation, interstitial fibrosis/Renal afferent arteriolar dense deposits
6	4/M	(-)	(-)	(+)	67	0.54	3.95	3+	40.5	Hearing impairment	The elder sister: diagnosed kidney disease at 4 years old and deceased at 6; Mother: SLE	Mesangial and endocapillary proliferation, individual crescents/ Subepithelial and rare subendothelial electron-dense deposits
7	11/F	Respiratory	(-)	(-)	195	0.8	1.06	3+	18.4			Mesangial proliferation/NA
8	10/M	Dermal	(-)	(-)	35	2.54	1.42	3+	26.4		Parents: recurrent urticaria; Cousins: history of NS or hematuria	Mesangial and endocapillary proliferation, mild interstitial fibrosis, double contours of GBM /No observed electron-dense deposits
9	12/M	Respiratory	(+)	(+)	25	7.098	NA	3+	↓ <sup>b</sup>			Mesangial and endocapillary proliferation, tubular atrophy /No observed electron-dense deposits
10	10/F	(-)	(+)	(-)	50	2.6	NA	2+	↓ <sup>c</sup>			Mesangial and endocapillary proliferation, interstitial fibrosis /Mesangial and subepithelial electron-dense deposits
11	4/M	Dermal	(-)	(-)	6	5.9	NA	3+	Normal <sup>d</sup>	Renal malrotation (Right)	Aunt and uncle: history of edema and proteinuria; Uncle: diabetes insipidus	Mesangial and endocapillary proliferation, double contours of GBM, glomeruli hyalinized, interstitial fibrosis/NA
12	11/F	(-)	(+)	(-)	70	2.78	NA	2+	↓ <sup>e</sup>			Mesangial and endocapillary proliferation, double contours of GBM, tubular atrophy/NA
13	12/F	(-)	(-)	(+)	7	2.53	2.18	3+	35.5			Segmental sclerosis, mesangial and endocapillary proliferation, crescents, tubular atrophy, interstitial fibrosis, glomeruli hyalinized/No observed electron-dense deposits
14	12/F	Respiratory	(+)	(+)	8	4.17	4.34	3+	23.9			Mesangial and endocapillary proliferation/Mild mesangial and subendothelial electron-dense deposits
15	9/F	Respiratory	(+)	(-)	(-)	14.9	NA	3+	↓ <sup>f</sup>			Mesangial and endocapillary proliferation, tubular atrophy/NA
16	12/M	(-)	(+)	(-)	20	4.88	NA	3+	31			Crescents, mesangial and endocapillary proliferation, segmental sclerosis, tubular atrophy, interstitial fibrosis/Severe subendothelial and subepithelial electron-dense deposits
17	7/M	(-)	(-)	(-)	∞	1.5	NA	2+	NA			Mesangial and endocapillary proliferation, crescents, tubular atrophy, / Mild mesangial electron-dense deposits

F, female; LM, light microscopy; EM, electron microscopy; GBM, glomerular basement membrane; HPF, High power field; SLE, systemic lupus erythematosus; M, male; NS, nephrotic syndrome; y, years; NA, not available; P/C, urinary protein/creatinine. Serum album of patient 1, 9, 10, 11, 12 and 15 were evaluated by serum protein electrophoresis. 33% (a), 59.9% (b), 55.1% (c), 66.7% (d), 52.8% (e) and 58.4% (f).

**Table S2** Clinical characteristics and laboratory parameters of patients with MPGN at last follow-up

Patient No.	Complications	Hematuria(/HPF)	eGFR(ml/min·1.73m <sup>2</sup> )	24-hour urinary protein (g)	Lipstick of proteinuria	C3 (g/L)	C3 NeF	Time from disease onset to renal biopsy (months)
1	Hypertension dental ulcers	(-)	5.9	4.8	2+	NA	NA	19.6
2	dental ulcers	72.18	148.7	NA	1+	NA	NA	0.6
3	Hypertension	63.39	109.5	0.11	(-)	0.83	(-)	1.3
4	Hypertension	21.55	110.3	NA	micro	1.1	NA	1.8
5	(-)	(-)	190.9	NA	1+	NA	NA	1.7
6	(-)	(-)	66.1	NA	2+	NA	NA	1.2
7	(-)	(-)	148.0	NA	+	1.65	NA	0.3
8	(-)	90	115.1	1.08	3+	0.1	NA	13.1
9	(-)	(-)	134.9	0.49	(-)	NA	NA	4.4
10	(-)	(-)	104.0	NA	(-)	NA	NA	2.0
11	(-)	(-)	33.9	NA	2+	NA	NA	84.4
12	(-)	(-)	110.0	NA	(-)	NA	NA	1.6
13	(-)	(-)	5.6	NA	NA	NA	NA	7.3
14	High intraocular pressure Binocular cataract Urinary tract infection	(-)	135.2	0.11	(-)	0.64	(-)	2.6
15	Urinary tract infection	(-)	70.8	(-)	(-)	NA	NA	15.4
16	Urinary tract infection	(-)	NA	NA	3+	NA	NA	7.1
17	blurred vision	(-)	75.4	NA	(-)	NA	NA	63.7

C3NeF, C3 Nephritis Factor; HPF, High power field; NA, not available; eGFR was calculated by the Schwartz formula using a local k-factor of 49 in CKD 1-2, 36.5 in CKD 3-5.



**Figure S1** Evolution of serum C3 and C3NeF in the whole course of disease. (A) patient 3; (B) patient 14.