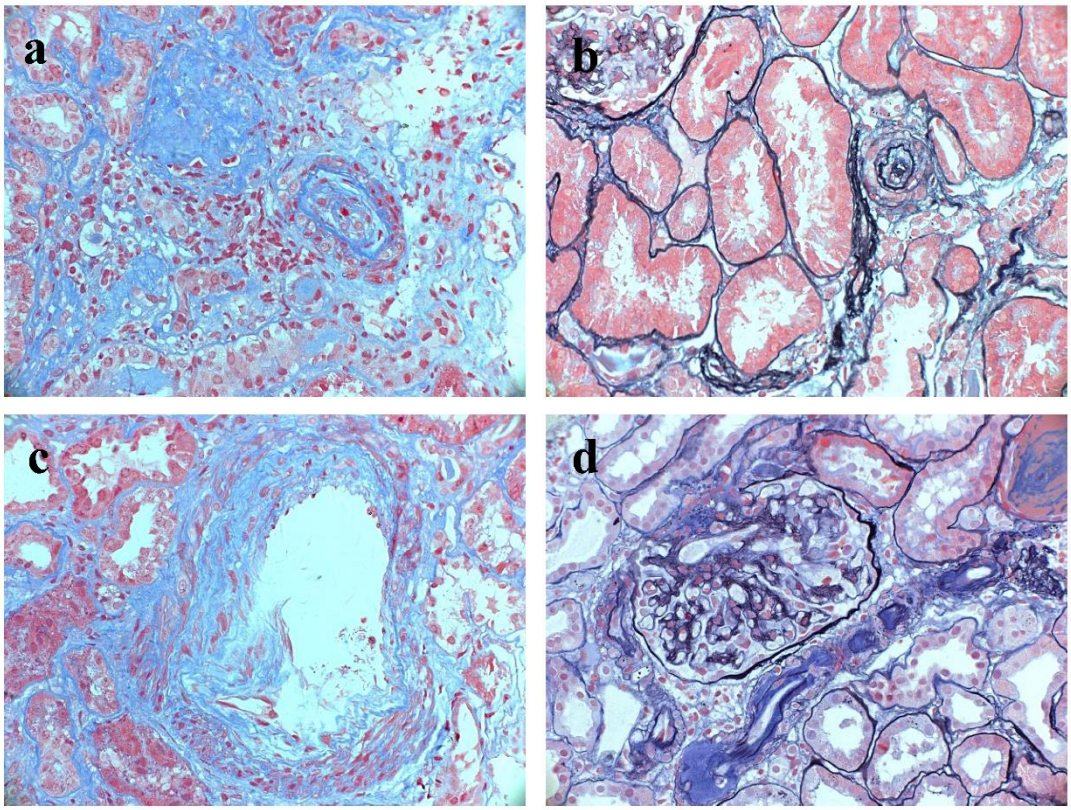

Figure S1. Typical examples of MA lesions and arteriolosclerosis in IgA nephropathy. (A) Subintimal edema in an arteriole (Masson trichrome stain). (B) Arterioles with onion-skin lesions (PASM-Masson stain). (C) Arterial intimal fibrosis arteriole (Masson trichrome stain). (D) Arteriolar hyalinosis (PASM-Masson stain); A-D: original magnification, ×400)

Supplementary Table 1. Clinicopathological features of groups with non-vascular lesions, simple arterio-/arteriolosclerosis and microangiopathic lesions.

	Microangiopathic lesions n=181	**Simple arterio-/ arteriolosclerosis n=299**	**No vascular lesion n=414**	**p**	**p***	**p****	**p*****
Clinical information							
Sex, male (n, %)	108(59.7%)	156(52.2%)	187(45.2%)	**0.004**	-	-	-
Age (year)	37(30-47)	39(30-48)	30(25-37)	**<0.001**	**<0.001**	**<0.001**	0.994
Systolic BP (mm Hg)	129.03±18.31	122.73±14.64	120.99±14.60	**<0.001**	0.205	**<0.001**	**0.002**
Diastolic BP (mm Hg)	82.32±12.94	78.8±10.2	77.52±11.09	**<0.001**	0.294	**<0.001**	**0.008**
Proteinuria (g/24 h)	2.34(1.40-4.00)	1.48(0.82-3.21)	1.21(0.64-2.38)	**<0.001**	**0.004**	**<0.001**	**<0.001**
Scr (μmol/L)	127(93.5-175.05)	91(74-122)	83(67.75-103)	**<0.001**	**<0.001**	**<0.001**	**<0.001**
eGFR (ml/min/1.73 m^2)	59.69±26.67	78.28±27.88	91.48±26.55	**<0.001**	**<0.001**	**<0.001**	**<0.001**
Hemoglobin (g/L)	132.03±19.49	133.26±18.32	134.5±19.41	0.328	0.395	0.147	0.493
Platelet count (10^9/L)	239.28±63.51	235.49±61.56	250.09±62.71	**0.006**	**0.002**	0.053	0.519
Albumin (g/L)	36.94±5.45	37.05±6.68	38.09±6.22	**0.035**	**0.028**	**0.039**	0.858
Pathological features							
Oxford classification (n, %)							
M1	154(85.1%)	202(67.6%)	250(60.4%)	**<0.001**	0.13	**<0.001**	**<0.001**
E1	80(44.2%)	122(40.8%)	180(43.5%)	0.702	NS	NS	NS
S1	148(81.8%)	211(70.6%)	273(65.9%)	**<0.001**	0.542	**<0.001**	**0.027**
T1/T2	65(35.9%)/59(32.6%)	106(35.5%)/19(6.4%)	79(19.1%)/12(2.9%)	**<0.001**	**<0.001**	**<0.001**	**<0.001**
C1/C2	71(39.2%)/25(13.8%)	135(45.2%)/30(10%)	168(40.6%)/33(8%)	0.13	NS	NS	NS

p, p-values among the three groups; p*, p-values between groups with no vascular lesions and simple arterio-/arteriolosclerosis; p**, p-values between groups with no vascular lesions and microangiopathic lesions; p***, p-values between groups with simple arterio-/arteriolosclerosis and microangiopathic lesions. Unless otherwise indicated, the values represent n(%), the mean±SD, or the median (25^{th}-75^{th} centiles). Bolded values are statistically significant. Abbreviations: BP, blood pressure; Scr, serum creatinine; eGFR, estimated glomerular filtration rate; M, mesangial hypercellularity; E, endocapillary hypercellularity; S, segmental glomerulosclerosis; T, interstitial fibrosis and tubular atrophy; C, crescents.

Supplementary Table 2. Clinicopathological features of 94 patients

Clinicopathological information	IgAN-MA
Clinical information	
Male(n,%)	59(62.8%)
Age(years)	35(28.75-45.25)
Hypertension(n,%)	77(81.9%)
Scr (μmol/L)	164.0(116.6-203.6)
eGFR(ml/min/1.73m2)	43.9(31.8-60.5)
Proteinuria (g/24 h)	2.64(1.34-5.30)
Oxford classification	
M1	81(86.2%)
E1	46(48.9%)
S1	80(85.1%)
T1/2	33(39.3%)/43(45.7%)
C1/2	45(53.6%)/8(8.5%)

Abbreviations: Scr, serum creatinine; eGFR, estimated glomerular filtration rate; M, mesangial hypercellularity; E, endocapillary hypercellularity; S, segmental glomerulosclerosis; T, interstitial fibrosis and tubular atrophy; C, crescents

Supplementary Table 3. Predicted potentially pathogenic mutations

Gene	Pathway	Nucleotide	AAChange	genotype	MAF	PS
C2	complement	c.1220-1G>A	Splice	Heterozygous	-1	NA
C2	complement	c.2059C>G	p.R687G	Heterozygous	-1	3
C3	complement	c.1625G>A	p.G542D	Heterozygous	0	2
C3	complement	c.3761G>A	p.R1254H	Heterozygous	0	3
C4BPA	complement	c.311A>G	p.Y104C	Heterozygous	-1	3
C5	complement	c.1286C>T	p.T429M	Heterozygous	0	2
C5	complement	c.2789T>C	p.V930A	Heterozygous	-1	4
C6	complement	c.1961T>G	p.F654C	Heterozygous	0	4
C8B	complement	c.757G>T	p.G253C	Heterozygous	-1	2
CFH	complement	c.1817C>A	p.P606H	Heterozygous	-1	3
CFHR2	complement	c.59_63delCAATG	Splice	Heterozygous	-1	NA
CFHR3	complement	c.404C>T	p.S135F	Heterozygous	-1	2
CFHR3	complement	c.634C>T	p.P212S	Heterozygous	0	2
CFI	complement	c.823C>T	p.Q275*	Heterozygous	-1	NA
CR1	complement	c.5438T>G	p.I1813S	Heterozygous	5.7984E-05	2
CR1	complement	c.6472+1_6472+17delGTGCCTTGACTCTCTGG	Splice	Homozygous	-1	NA

CR2	complement	c.472C>T	p.P158S	Heterozygous	-1	3
CR2	complement	c.655G>A	p.G219R	Heterozygous	-1	2
FCN1	complement	c.185C>T	p.P62L	Heterozygous	-1	2
FCN2	complement	c.512G>A	p.G171E	Heterozygous	-1	3
FCN3	complement	c.92-2A>G	Splice	Heterozygous	-1	NA
F5	coagulation	c.1059C>G	p.F353L	Heterozygous	5.7984E-05	4
F5	coagulation	c.1276C>T	p.Q426*	Heterozygous	-1	NA
F5	coagulation	c.6586A>G	p.I2196V	Heterozygous	5.3107E-05	3
VWF	coagulation	c.1307G>A	p.R436H	Heterozygous	0	2
ADAMTS13	coagulation	c.311T>A	p.V104E	Heterozygous	-1	4
F11	coagulation	c.486-2A>G	Splice	Heterozygous	-1	NA
F11	coagulation	c.976C>T	p.R326C	Heterozygous	0	3
F12	coagulation	c.581G>C	p.G194A	Heterozygous	5.3908E-05	2
F2	coagulation	c.1195C>T	p.R399C	Heterozygous	0	3
PROS1	coagulation	c.1021G>A	p.A341T	Heterozygous	5.2994E-05	4
SERPINF2	coagulation	c.103-2A>G	Splice	Heterozygous	5.8025E-05	NA
ACE	RAAS	c.214G>T	p.D72Y	Heterozygous	0	3
ACE	RAAS	c.3629C>T	p.T1210M	Heterozygous	0	2
AGT	RAAS	c.917C>A	p.T306N	Heterozygous	-1	2

Supplementary Table 4. Reported potentially pathogenic mutations

Gene	Nucleotide	AAChange	genotype	MAF	Related disease
CFHR5	c.508G>A	p.V170M	Heterozygous	0.008109	aHUS;IgA nephropathy
CFHR5	c.1357C>T	p.P453S	Heterozygous	0.001166	IgA nephropathy
CFHR5	c.533A>G	p.N178S	Heterozygous	0.005672	IgA nephropathy
CFI	c.848A>G	p.D283G	Heterozygous	0.000348	aHUS
C3	c.1273C>T	p.R425C	Heterozygous	0.002651	aHUS
C7	c.281-1G>T	Splice	Heterozygous	0.001379	Complement component 7 deficiency
F8	c.2535C>A	p.D845E	Heterozygous	0.000933	Hemophilia A
vWF	c.6860G>A	p.R2287Q	Heterozygous	0.005302	Hemophilia

Abbreviations: MAF, minor allele frequency