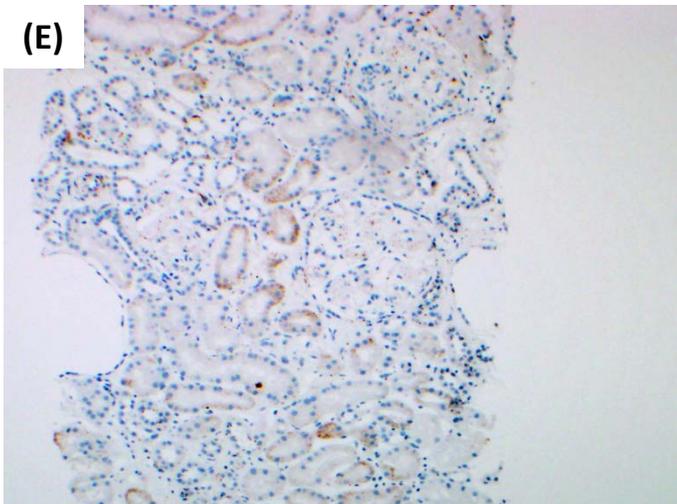
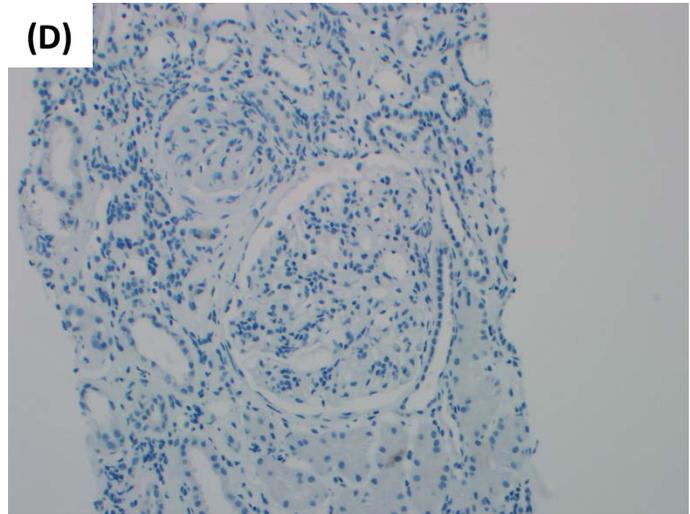
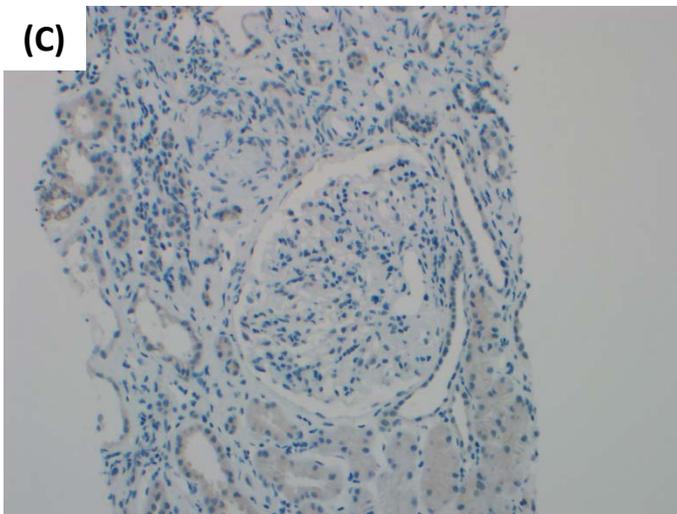
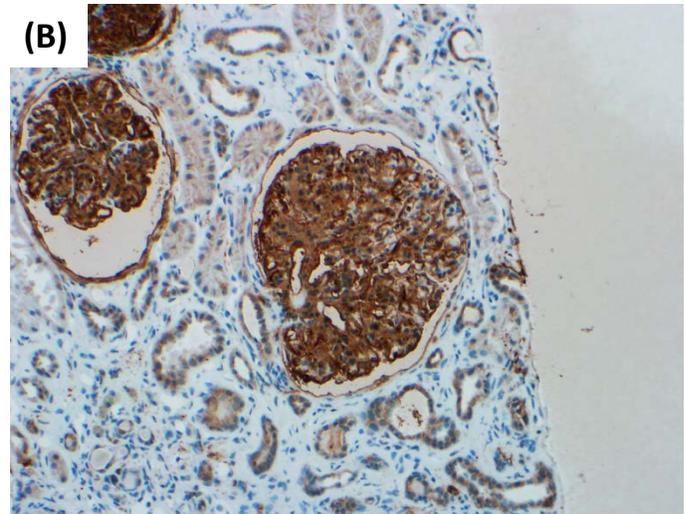
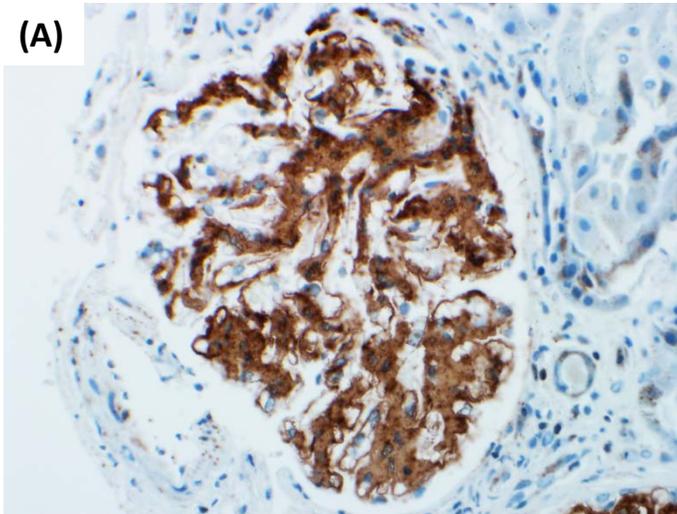


Supplemental Figure 2. DNAJB9 Primary Antibody Specificity. We tested the specificity of DNAJB9 primary antibody using immunohistochemistry (IHC). A total of 6 FGN cases, 9 amyloid cases (3 AL- κ , 3 AL- λ , and 3 ALect2), and 9 healthy controls were chosen from the LC-MS/MS cohort for antibody testing. **(A-B)** DNAJB9 staining from two FGN cases showing smudgy glomerular staining (sparing the nuclei). All FGN cases showed similar staining pattern. **(C-D)** Serial sections from the case shown in (B) were stained with control diluent and polyclonal rabbit IgG as primary stain, instead of anti-DNAJB9 antibody, respectively. This showed that the DNAJB9 staining observed in FGN cases was not due to non-specific secondary antibody staining. **(E-F)** DNAJB9 staining in example cases of AL- λ and healthy control, respectively. As expected, we observed tubular staining, but glomeruli were negative for DNAJB9 in all amyloid and healthy controls.



Supplemental Table 1. Clinical and Pathologic Characteristics of 24 cases of Fibrillary Glomerulonephritis

Case #	DNAJB9 method of testing	Age/ gender	S. Cr. (mg/dl)	Proteinuria (g/day)	Full NS	Hematuria	Concurrent conditions	Glomerular morphology on LM	# of glomeruli sampled for LM/ % global glomerulosclerosis	Degree of TA&IF	Positive immune reactants in glomeruli by IF	Extraglomerular staining for IgG	Fibril distribution on EM	Mean fibril thickness
1	MS + IHC	44/M	12.8	NA (>300 mg/dl on UA)	no	yes	HTN	mes. proliferative GN	12/83%	marked	IgG (3+), C3 (2+), kappa (3+), lambda (3+)	no	mes, GBM	18
2	MS + IHC	60/F	NA	NA (subnephrotic)	no	yes	none	mes. proliferative GN	101/20%	mild	IgG (2+), C3 (2+), kappa (+/-), lambda (1+)	no	mes, GBM	20
3	MS + IHC	71/M	2.6	2.5	no	no	hepatocellular Ca, HTN	mes. proliferative GN	22/23%	mild	IgG (3+), C3 (3+), kappa (2+), lambda (2-3+), IgA (1-2+), C1q (2+)	no	mes, GBM	19
4	MS + IHC	59/F	2.3	NA (2+ on UA)	no	yes	HTN, hemoptysis	crescentic GN	8/25%	none	IgG (3+), C3 (3+), kappa (1-2+), lambda (1-2+), IgM (+/-), C1q (+/-)	no	mes, GBM	18
5	MS + IHC	64/M	1.2	1.7	no	yes	prostate CA	mes. proliferative GN	24/0%	mild	IgG (1-2+), C3 (1+), IgA (1-2+), IgM (1-2+), kappa (+/-), lambda (1-2+)	yes (focal TBM)	mes, GBM	17
6	MS + IHC	53/F	2.8	20	no	no	DM, HTN, COPD	mes. proliferative GN	13/15%	moderate	IgG (3+), C3 (2-3+), kappa (3+), lambda (3+)	no	mes, GBM	19
7	MS + IHC	58/F	1.6	9	no	yes	none	membranoproliferative GN	25/72%	moderate	IgG (2-3+), IgM (1+), IgA (+/-), C3 (1+), kappa (2+), lambda (2-3+)	no	mes, GBM	20
8	MS	55/M	1.9	6.7	no	yes	HCV, HIV, HTN	mes. proliferative GN	12/42%	moderate	IgG (3+), kappa (3+), lambda (3+)(by pronase IF), C3 (2+)	no	mes, GBM	not measured
9	MS	75/M	1.4	6.6	no	yes	HTN	mes. proliferative GN	18/39	mild	IgG (2+), C3 (1+), IgM (1+), kappa (1+), lambda (2+)	no	mes, GBM	16
10	MS	73/M	5.5	8.8	yes	yes	DM	endocapillary proliferative GN	15/ 20%	mild	IgG (3+), C3 (2+), C1q (1+), IgA (+/-); staining for kappa and lambda not done	no	mes, GBM	12
11	MS	55/M	0.7	3.4	no	yes	cutaneous lupus	mes. proliferative GN	12/0%	mild	IgG (3+), C3 (1+), IgM (1+), IgA (1+), C1q (2+); staining for kappa and lambda not done	no	mes, GBM	15
12	MS	53/M	2.4	17	NA	yes	HCV, HTN, morbid obesity	mes. proliferative GN	20/25%		IgG (1+), C3 (1+), kappa (+/-), lambda (1+)	no	mes, GBM	not measured
13	MS	44/F	0.8	5	no	yes	undifferentiated MCTD	mes. proliferative GN	28/11%	none	IgG (2-3+), C3 (3+), IgM (1-2+), IgA (1+), C1q (2+); kappa (2+), lambda (3+)	no	mes, GBM	10
14	MS	37/M	0.9	2.4	no	no	none	mes. proliferative GN	16/0%	none	IgG (3+), C3 (1+), kappa (2+), lambda (3+)	no	mes, GBM	20
15	MS	59/F	4.1	NA (>300 mg/dl on UA)	NA	yes	HTN, CAD	mes. proliferative GN	53/75%	marked	IgG (2-3+), C3 (1+), kappa (1+), lambda (+/-)	no	mes, GBM	17
16	MS	71/F	1.7	1.7	no	yes	HTN, gout	mes. proliferative GN	17/88%	moderate	IgG (2+), C3 (1-2+), IgM (1-2+), kappa (1-2+), lambda (1-2+)	no	mes, GBM	17

17	MS	67/F	6.8	NA	NA	NA	none	membranoproliferative GN	30/53%	marked	IgG (3+), C3 (3+), kappa (1+), lambda (3+)	no		19
18	MS	76/F	1.6	NA	NA	NA	NA	mes. proliferative GN	11/45%	mild	IgG (3+), C3 (2+), kappa (3+), lambda (3+)	no	mes, GBM	17
19	MS	63/F	1.4	0	no	yes	COPD	mes. proliferative GN	6/50%	mild	IgG (2+), C3 (1+), kappa (1-2+), lambda (2+)	no	mes, GBM	16
20	MS	66/M	1	3.7	NA	yes	DM, HTN	mes. proliferative GN	19/5%	mild	IgG (2+), C3 (1+), kappa (+/-), lambda (+/-)	no	mes, GBM	18
21	MS	62/M	1.3	6	no	yes	HTN	mes. proliferative GN	25/20%	mild	IgG (2-3+), C3 (2+), kappa (3+), lambda (1-2+), IgA (2-3+)	no	mes, GBM	14
22	MS	50/F	0.9	0.6	no	yes	HTN	mes. proliferative GN	34/26%	mild	IgG (3+), C3 (3+), kappa (1-2+), lambda (1+), C1q (1+)	yes (TBM)	mes, GBM, TBM	13
23	MS	65/M	2.4	5.3	NA	NA	DM	endocapillary proliferative GN	32/34%	marked	IgG (3+), IgM (2+), C3 (2+), kappa (3+), lambda (2+)	no	mes, GBM	14
24	MS	40/F	2.3	2.7	no	yes	NSIP	mes. proliferative GN	31/0%	mild	IgG (3+), C3 (3+), kappa (2+), lambda (1-2+)	no	mes, GBM	13

CA, carcinoma; CAD, coronary artery disease; COPD, chronic obstructive pulmonary disease; DM, diabetes; GBM, glomerular basement membrane; HTN, hypertension; IHC, immunohistochemistry; MCTD, mixed connective tissue disorder; mes, mesangial; NSIP, nonspecific interstitial pneumonia; mes, mesangial; NS, nephrotic syndrome; MS, mass spectrometry; NA, not available; TBM, tubular basement membranes;

Proteins	Median SpC ^a		Description
	FGN	Amyloid	
gi 4501885 ref NP_001092.1	132	93	actin, cytoplasmic 2 [Homo sapiens]
gi 4501889 ref NP_001606.1	93	69	actin, aortic smooth muscle [Homo sapiens]
gi 62414289 ref NP_003371.2	77	91	vimentin [Homo sapiens]
gi 9558755 ref NP_036460.1	54	0	dnaJ homolog subfamily B member 9 [Homo sapiens]
gi 115298678 ref NP_000055.2	54	18	complement C3 precursor [Homo sapiens]
gi 119703755 ref NP_002283.3	49	15	laminin subunit beta-2 precursor [Homo sapiens]
gi 4557325 ref NP_000032.1	47	118	apolipoprotein E precursor [Homo sapiens]
gi 4504349 ref NP_000509.1	45	44	hemoglobin subunit beta [Homo sapiens]
gi 12667788 ref NP_002464.1	41	21	myosin-9 [Homo sapiens]
gi 240255535 ref NP_476507.3	39	10	collagen alpha-3(VI) chain isoform 4 precursor [Homo sapiens]

Supplemental Table 2: This table displays top 10 proteins detected in the glomeruli of fibrillary glomerulonephritis (FGN). (a) Total number of spectra matched to a protein is considered as a semi-quantitative measure of its abundance in a sample. Protein spectral counts were normalized to account for protein loading differences between LC-MS/MS experiments. Median protein spectral counts (SpC) for each the protein detected in FGN and amyloid cohorts was computed. Rows were ordered by the decreasing abundance of the protein in FGN cohort.

ID	Name	#Gene	FDR
R-HSA-977606	Regulation of Complement cascade	5	4.39E-08
R-HSA-166658	Complement cascade	5	2.71E-07
R-HSA-174577	Activation of C3 and C5	3	2.05E-05
R-HSA-166663	Initial triggering of complement	3	4.83E-04
R-HSA-168249	Innate Immune System	6	1.39E-02

Supplemental Table 3: This table displays the pathway enrichment results for proteins that are overabundant in FGN glomeruli. For this, proteins that had a corrected differential expression p-value of ≤ 0.05 and a $\log_2(\text{fold change}) \geq 0.3$ were uploaded to WebGestalt for pathway enrichment analysis (<http://www.webgestalt.org/option.php>). Pathways that had an corrected enrichment FDR ≤ 0.05 were considered as significant for reporting.