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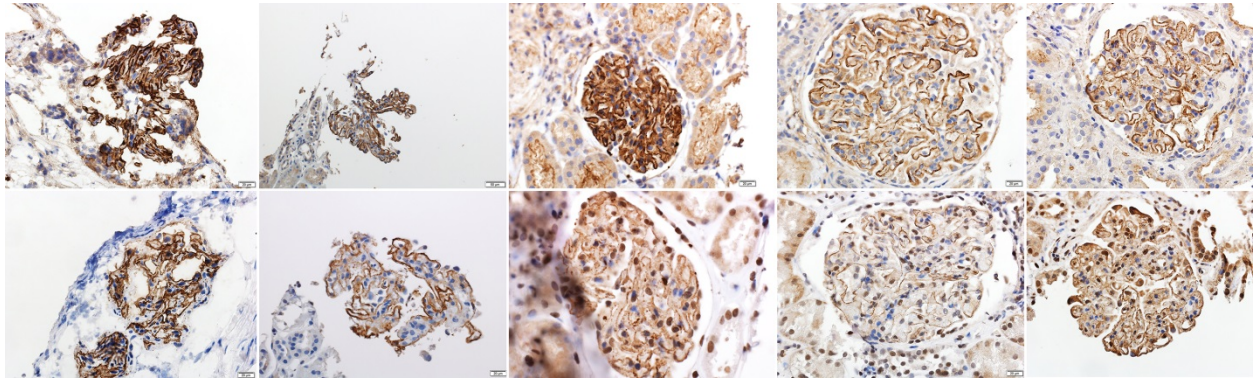
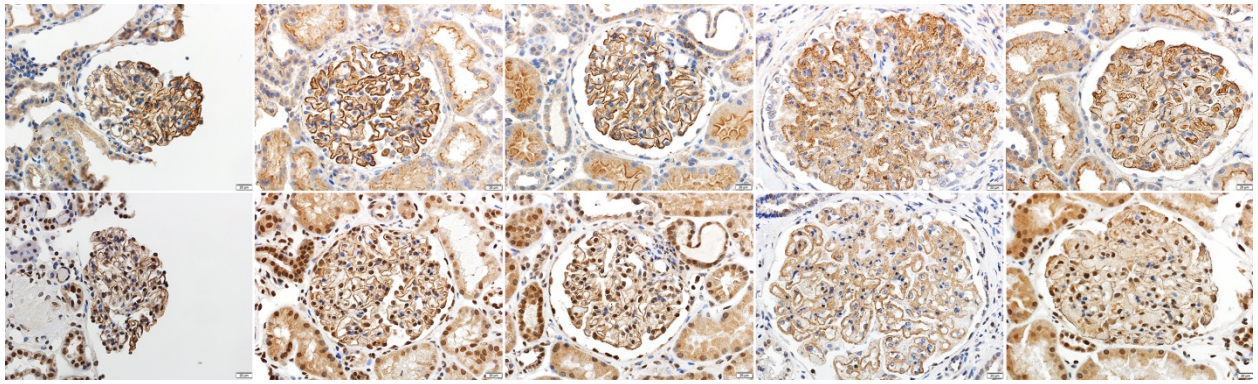
Supplement table 1. Laser microdissection: number of glomeruli dissected and cut area/case.

Case number	Number of glomeruli dissected	Cut area square microns
Case 1	8	127231
Case 2	25	642922
Case 3	8	143755
Case 4	30	572387
Case 5	28	570610
Case 6	10	234746
Case 7	28	573762
Case 8	17	255959
Case 9	21	335912
Case 10	23	350584
Case 11	35	589132
Case 12	20	614729
Case 13	21	531275
Case 14	9	132207
Case 15	NA	NA
Case 16	NA	NA
Case 17	25	538594
Case 18	6	169703
Case 19	12	142972
Case 20	NA	NA
Case 21	28	534491
Case 22	13	195501
Case 23	NA	NA
Case 24	19	542453
Case 25	18	544770
Case 26	NA	NA

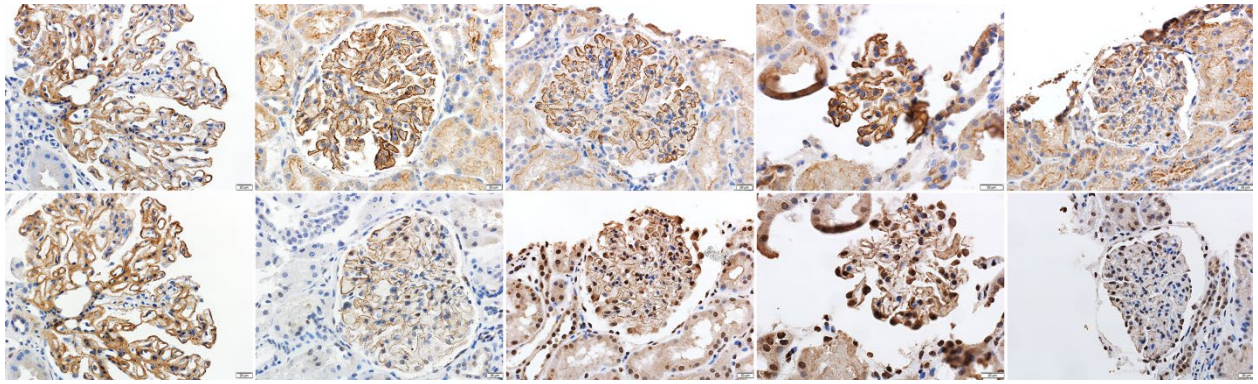
Supplement figure 1A: EXT1/EXT2-associated MN: Granular staining for EXT1 and EXT2 along the GBM in 5 cases of EXT1/EXT2 associated MN. The first column is case 1, column 2 is case 2, column 3 is case 5, column 4 is case 6 and column 5 is case 8 (top row EXT1, bottom row EXT2).



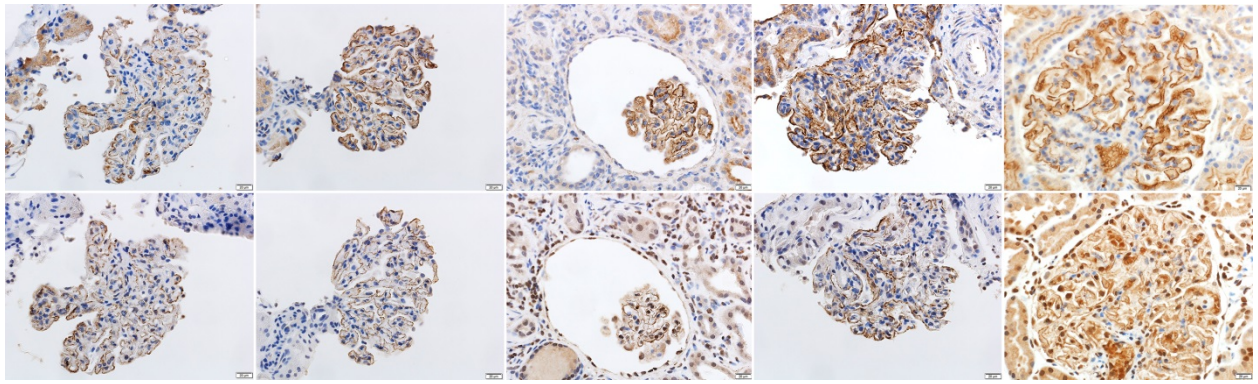
Supplement figure 1B: EXT1/EXT2-associated MN: Bright granular staining for EXT1 and EXT2 along the GBM in 5 cases of EXT1/EXT2 associated MN. The first column is case 9, column 2 is case 10, column 3 is case 11, column 4 is case 12 and column 5 is case 13 (top row EXT1, bottom row EXT2).



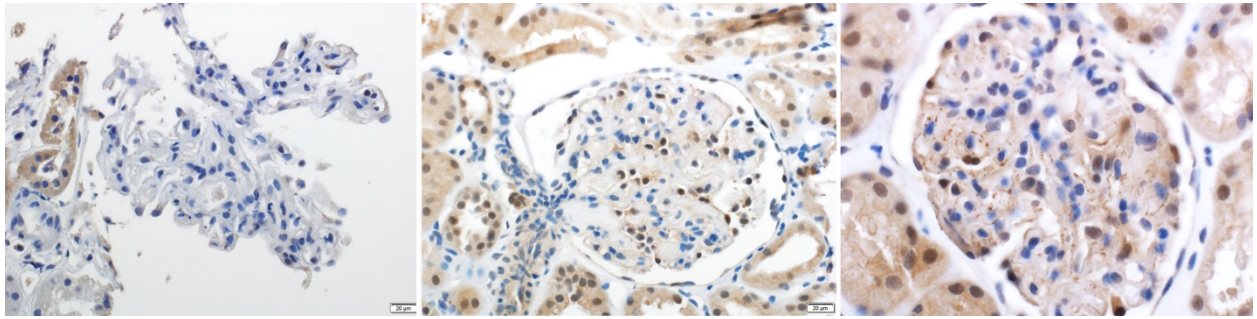
Supplement figure 1C: EXT1/EXT2-associated MN: Bright granular staining for EXT1 and EXT2 along the GBM in 5 cases of EXT1/EXT2 associated MN. The first column is case 15, column 2 is case 16, column 3 is case 17, column 4 is case 18 and column 5 is case 19 (top row EXT1, bottom row EXT2).



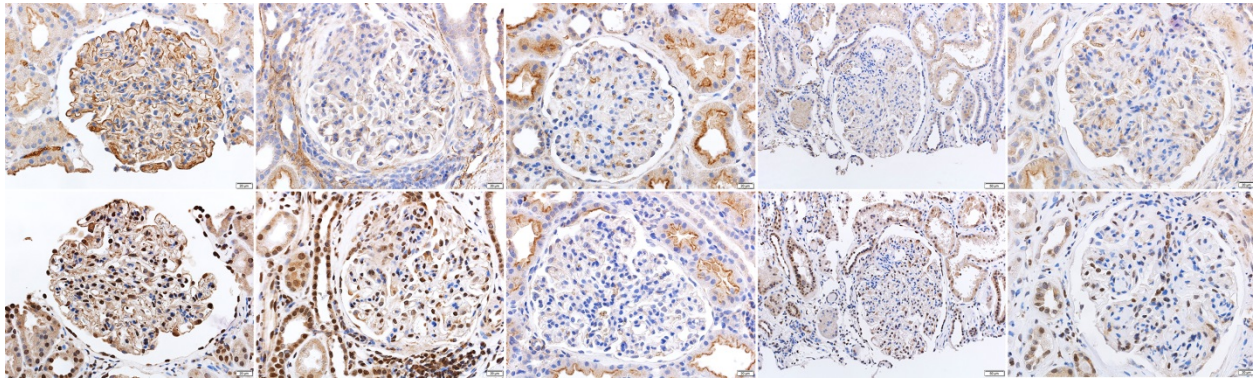
Supplement figure 1D: EXT1/EXT2-associated MN: Bright granular staining for EXT1 and EXT2 along the GBM in 5 cases of EXT1/EXT2 associated MN. The first column is case 20, column 2 is case 21, column 3 is case 22, column 4 is case 23 and column 5 is case 26 (top row EXT1, bottom row EXT2).



Supplement figure 1E: EXTL2 staining in 3 cases of EXT1/EXT2-associated MN: 2 cases of EXT1/EXT2-associated MN are negative for EXTL2and one shows minimal (1+) granular staining for EXTL2 (case 5, that showed very bright EXT1/EXT2 staining).



Supplement figure 1F: EXT1/EXT2 staining in control cases compared to EXT1/EXT2-associated MN: Bright granular staining for EXT1/EXT2 along the GBM in a case of EXT1/EXT2-associated MN (column 1- case 24), and negative in control cases (column 2- IgA nephropathy, column 3- minimal change disease, column 4- FSGS and column 5-diabetic glomerulosclerosis) (top row EXT1, bottom row EXT2).

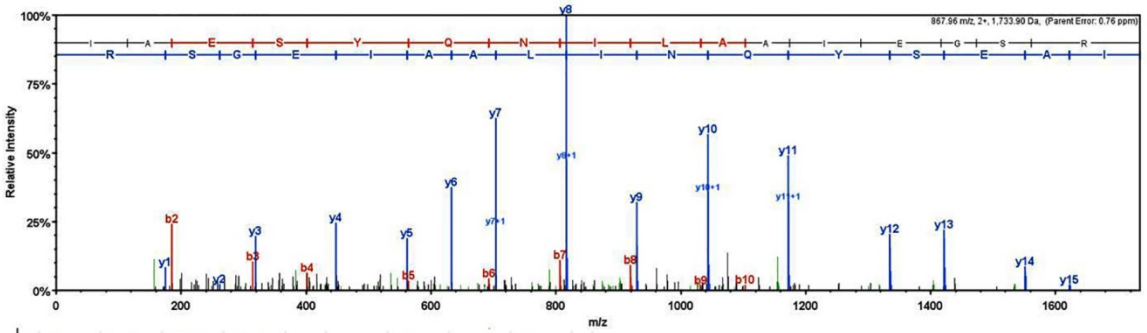


Supplement figure 2. Representative mass spectrometry findings of control cases: Low spectral counts of PLA2R and no detectable spectral counts of EXT1 or EXT2 are present in the control cases. The last 2 columns are from 2 cases of PLA2R-negative MN that were also negative for EXT1 and EXT2. Proteins inherent to the GBM such as laminin, nestin, nidogen are also shown (MCD-minimal change disease; PLA2R negative case-PLA2R-negative primary membranous nephropathy).

Probability Legend: over 95% 80% to 94% 50% to 79% 20% to 49% 0% to 19% Bio View: 2323 Proteins in 2107 Clusters With 2321 Filtered Out	Accession Number	Molecular Weight	Protein Grouping Ambiguity	Diabetes Case 1	Diabetes Case 2	IgA Case 1	IgA Case 2	Primary FSGS Case 1	Primary FSGS Case 2	MCD Case 1	MCD Case 2	PLA2R negative Case 1	PLA2R negative Case 2
Exostosin-2 OS=Homo sapiens GN=EXT2 PE=1 SV=1	sp|Q93063|EXT2_HUMAN	82 kDa		(0)	(0)	(0)	(0)	(0)	(0)	(0)	(0)	(0)	(0)
Exostosin-1 OS=Homo sapiens GN=EXT1 PE=1 SV=2	sp|Q16394|EXT1_HUMAN	86 kDa	★	(0)	(0)	(0)	(0)	(0)	(0)	(0)	(0)	(0)	(0)
Secretory phospholipase A2 receptor OS=Homo sapiens GN=PLA2R1 PE=1 S...	sp|Q13018|PLA2R_HUMAN	169 kDa	★	5	3	2	4	3	3	(0)	(0)	3	4
Laminin subunit beta-2 OS=Homo sapiens GN=LAMB2 PE=1 SV=2	sp|P55268|LAMB2_HUMAN	196 kDa	★	161	166	230	176	154	239	224	188	205	128
Nestin OS=Homo sapiens GN=NES PE=1 SV=2	sp|P48681|NEST_HUMAN	177 kDa	★	81	74	93	97	100	133	163	177	90	102
Nidogen-1 OS=Homo sapiens GN=NID1 PE=1 SV=3	sp|P14543|NID1_HUMAN	136 kDa	★	78	90	113	86	96	101	98	87	146	90
Vinculin OS=Homo sapiens GN=VCL PE=1 SV=4	sp|P18206|VINC_HUMAN	124 kDa	★	75	77	91	89	106	103	119	129	147	78
Heat shock protein HSP 90-alpha OS=Homo sapiens GN=HSP90AA1 PE=1 ...	sp|P07900|HS90A_HUMAN	85 kDa	★	35	31	29	30	32	47	36	26	19	32
Heat shock protein HSP 90-beta OS=Homo sapiens GN=HSP90AB1 PE=1 S...	sp|P08238|HS90B_HUMAN	83 kDa	★	31	31	28	29	37	33	38	35	21	30
Glyceraldehyde-3-phosphate dehydrogenase OS=Homo sapiens GN=GAP...	sp|P04406|G3P_HUMAN	36 kDa	★	35	41	44	37	35	47	67	65	35	40

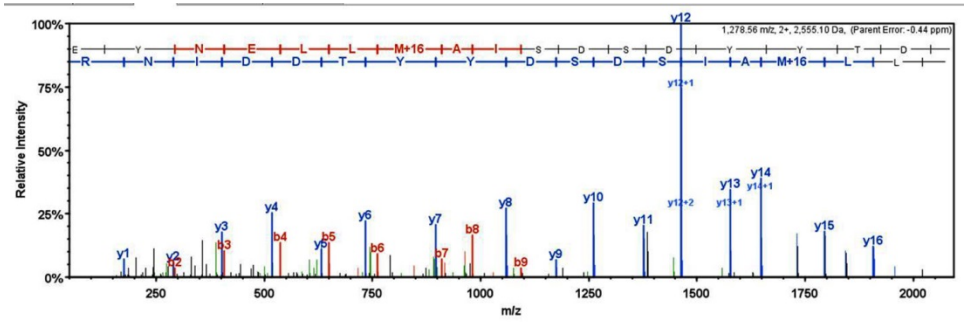
Supplement Figure 3.

A. The MS/MS spectra figure from Scaffold viewer of ion 867.96 $[M+2H]^{2+}$ highlighting the detected b-ions (red) and y-ions (blue) matching the theoretical fragment masses listed in the table for the Exostosin-1 peptide IAESYQNILAAIEGSR.



B	B Ions	B+2H	B-NH3	B-H2O	AA	Y Ions	Y+2H	Y-NH3	Y-H2O	Y
1	114.1				I	1,734.9	868.0	1,717.9	1,716.9	16
2	185.1				A	1,621.8	811.4	1,604.8	1,603.8	15
3	314.2			296.2	E	1,550.8	775.9	1,533.8	1,532.8	14
4	401.2			383.2	S	1,421.7	711.4	1,404.7	1,403.7	13
5	564.3			546.3	Y	1,334.7	667.9	1,317.7	1,316.7	12
6	692.3	346.7	675.3	674.3	Q	1,171.6	586.3	1,154.6	1,153.6	11
7	806.4	403.7	789.3	788.4	N	1,043.6	522.3	1,026.6	1,025.6	10
8	919.5	460.2	902.4	901.4	I	929.5	465.3	912.5	911.5	9
9	1,032.5	516.8	1,015.5	1,014.5	L	816.5	408.7	799.4	798.4	8
10	1,103.6	552.3	1,086.5	1,085.6	A	703.4	352.2	686.3	685.4	7
11	1,174.6	587.8	1,157.6	1,156.6	A	632.3	316.7	615.3	614.3	6
12	1,287.7	644.4	1,270.7	1,269.7	I	561.3		544.3	543.3	5
13	1,416.7	708.9	1,399.7	1,398.7	E	448.2		431.2	430.2	4
14	1,473.8	737.4	1,456.7	1,455.7	G	319.2		302.1	301.2	3
15	1,560.8	780.9	1,543.8	1,542.8	S	262.2		245.1	244.1	2
16	1,734.9	868.0	1,717.9	1,716.9	R	175.1		158.1		1

B. The MS/MS spectra figure from Scaffold viewer of ion 1278.56 $[M+2H]^{2+}$ highlighting the detected b-ions (red) and y-ions (blue) matching the theoretical fragment masses listed in the table for the Exostosin-2 peptide EYNELLMAISDSDYYTDDINR.



B	B Ions	B+2H	B-NH3	B-H2O	AA	Y Ions	Y+2H	Y-NH3	Y-H2O	Y
1	130.0			112.0	E	2,556.1	1,278.6	2,539.1	2,538.1	21
2	293.1			275.1	Y	2,427.1	1,214.0	2,410.0	2,409.1	20
3	407.2		390.1	389.1	N	2,264.0	1,132.5	2,247.0	2,246.0	19
4	536.2		519.2	518.2	E	2,150.0	1,075.5	2,132.9	2,131.9	18
5	649.3		632.3	631.3	L	2,020.9	1,011.0	2,003.9	2,002.9	17
6	762.4	381.7	745.3	744.4	L	1,907.8	954.4	1,890.8	1,889.8	16
7	909.4	455.2	892.4	891.4	M+16	1,794.7	897.9	1,777.7	1,776.7	15
8	980.4	490.7	963.4	962.4	A	1,647.7	824.4	1,630.7	1,629.7	14
9	1,093.5	547.3	1,076.5	1,075.5	I	1,576.7	788.8	1,559.6	1,558.7	13
10	1,180.6	590.8	1,163.5	1,162.5	S	1,463.6	732.3	1,446.6	1,445.6	12
11	1,295.6	648.3	1,278.6	1,277.6	D	1,376.6	688.8	1,359.5	1,358.5	11
12	1,382.6	691.8	1,365.6	1,364.6	S	1,261.5	631.3	1,244.5	1,243.5	10
13	1,497.6	749.3	1,480.6	1,479.6	D	1,174.5	587.8	1,157.5	1,156.5	9
14	1,660.7	830.9	1,643.7	1,642.7	Y	1,059.5	530.2	1,042.4	1,041.5	8
15	1,823.8	912.4	1,806.7	1,805.8	Y	896.4	448.7	879.4	878.4	7
16	1,924.8	962.9	1,907.8	1,906.8	T	733.3	367.2	716.3	715.3	6
17	2,039.8	1,020.4	2,022.8	2,021.8	D	632.3		615.3	614.3	5
18	2,154.9	1,077.9	2,137.8	2,136.9	D	517.3		500.2	499.3	4
19	2,268.0	1,134.5	2,250.9	2,249.9	I	402.2		385.2	384.2	3
20	2,382.0	1,191.5	2,365.0	2,364.0	N	289.2		272.1	271.2	2
21	2,556.1	1,278.6	2,539.1	2,538.1	R	175.1		158.1		1

Supplement Methods

LASER MICRODISSECTION AND MASS SPECTROMETRY

Formalin fixed paraffin embedded (FFPE) renal biopsy materials were sent to the Mayo Clinic renal biopsy laboratory for diagnosis of membranous nephropathy. For each case, 10μM thick paraffin sections were obtained and mounted on PEN membrane laser microdissection slides. The sections were deparaffinized using xylene and alcohol. Using a Zeiss Palm Microbean microscope and Robopalm software, multiple glomeruli were microdissected to reach approximately 250-500,000μM^2 per case, and catapulted into 35μl of digest buffer (100mM Tris, pH 8.5/0.002% Zwittergent Z3-16) in the cap of a 0.5 ml tube. The tube was removed from the collection plate and spun at 14000g x 2minutes. The samples were frozen until all samples were collected. Upon thawing, samples were heated to 98°C, then proteins were reduced and alkylated by sequential addition of TCEP (Tris(2-carboxyethyl) phosphine hydrochloride) and iodoacetamide to 10mM for 30 minutes each. Trypsin (0.05 μg) was added to each tube and proteins were digested overnight at 37°C for 16-18 hours. After digestion, the samples were acidified with trichloroacetic acid, dried down and resolubilized with A solvent for mass spectrometry.

The trypsin digested peptides were identified by nano-flow liquid chromatography electrospray tandem mass spectrometry (nanoLC-ESI-MS/MS) using a Thermo Scientific Q-Exactive Mass Spectrometer (Thermo Fisher Scientific, Bremen, Germany) coupled to a Thermo Ultimate 3000 RSLCnano HPLC system. The peptide mixture was loaded onto a 250nl OPTI-PAK trap (Optimize Technologies, Oregon City, OR) custom packed with Michrom Magic C 8, 5μm solid phase (Michrom Bioresources, Auburn, CA). Chromatography was performed using 0.2 % formic acid in both the A solvent (98%water/2%acetonitrile) and B solvent (80% acetonitrile/10% isopropanol/10% water), and a 5%B to 40%B gradient over 90 minutes at 400 nl/min through a PicoFrit (New Objective, Woburn, MA) 100μm x 35cm column handpacked with Agilent Poroshell 120 EC C18 packing. The Q-Exactive mass spectrometer experiment was a data dependent set up with the MS1 survey scan from 340-1500 m/z at resolution 70,000 (at 200m/z), followed by HCD MS/MS scans on the top 15 ions having a charge state of +2, +3, or +4, at resolution 17,500. The ions selected for MS/MS were placed on an exclusion list for 30 seconds. The MS1 AGC target was set to 1e6 and the MS2 target is set to 1e5 with max ion inject times of 50ms for both.

DATABASE SEARCHING

Tandem mass spectra was extracted by msconvert version 3.0.9134. Charge state deconvolution and deisotoping was not performed. All MS/MS samples were analyzed using Mascot (Matrix Science, London, UK; version 2.4.0) and X! Tandem (The GPM, thegpm.org; version X!Tandem Sledgehammer (2013.09.01.1)). Mascot and X! Tandem were set up to search a Swissprot human database with reverse decoy (40570 entries) assuming the digestion enzyme strict trypsin and with a fragment ion mass tolerance of 0.020 Da and a parent ion tolerance of 10.0 PPM. Glu->pyro-Glu of the n-terminus, ammonia-loss of the n-terminus, gln->pyro-Glu of the n-terminus, oxidation of methionine is specified in X! Tandem as variable modifications and carbamidomethyl of cysteine was specified as a fixed modification. Oxidation of methionine and carbamidomethyl of cysteine were specified in Mascot as variable modifications and fixed modifications respectively.

CRITERIA FOR PROTEIN IDENTIFICATION Scaffold (version Scaffold_4.8.3, Proteome Software Inc., Portland, OR) was used to validate MS/MS based peptide and protein identifications. Peptide identifications were accepted if they could be established at greater than 95.0% probability by the Scaffold Local FDR algorithm. Protein identifications were accepted if they could be established at greater than 95.0% probability and contained at least 2 identified peptides. Protein probabilities were assigned by the Protein Prophet algorithm.[7]The protein decoy false discovery rate (FDR) was <1.5.In general, over 1500-2500protein were identified in each sample. Proteins that contain similar peptides and cannot be differentiated based on MS/MS analysis alone were grouped to satisfy the principles of parsimony. Proteins sharing significant peptide evidence were grouped into clusters. Protein comparisons were made with ratios of Scaffold normalized total spectral counts. The 'Spectra' value indicates the total number of mass spectrum collected on the mass spectrometer and matched to the protein using the proteomics software. A higher number of mass spectra is indicative of greater abundance and will typically yield greater amino acid sequence coverage. A higher mass spectra value also indicates a higher confidence in the protein identification.

IMMUNOHISTOCHEMICAL STAINING FOR EXOSTOSIN 1 (EXT1),EXOSTOSIN 1 (EXT2)AND EXOSTOSIN-LIKE 2 (EXTL2):

Tissue sectioning and immunohistochemical (IHC) staining was performed at the Pathology Research Core (Mayo Clinic, Rochester, MN) using the Leica Bond RX stainer (Leica). FFPE tissues were sectioned at 5 microns and IHC staining was performed on-line. Slides for the EXT1 stain were retrieved for 20 minutes using Epitope Retrieval 2 (EDTA; Leica) and incubated in Protein Block (Dako) for 5 minutes. The EXT1 primary antibody (rabbit polyclonal, Thermo Scientific #PA5-60699) and EXT2 antibody (rabbit polyclonal; Abcam) was diluted to 1:100 in Background Reducing Diluent (Dako) and incubated for 15 minutes. Slides for the EXTL2 stain were retrieved for 20 minutes using Epitope Retrieval 1 (Citrate; Leica) and incubated in Protein Block (Dako) for 5 minutes. The EXTL2 primary antibody (rabbit polyclonal, Origene #TA590761) was diluted to 1:1400 in Background Reducing Diluent (Dako) and incubated for 15 minutes. The detection system used was Polymer Refine Detection System (Leica). This system includes the hydrogen peroxidase block, post primary and polymer reagent, DAB, and Hematoxylin. Immunostaining visualization was achieved by incubating slides 10 minutes in DAB and DAB buffer (1:19 mixture) from the Bond Polymer Refine Detection System. To this point, slides were rinsed between steps with 1X Bond Wash Buffer (Leica). Slides were counterstained for five minutes using Schmidt hematoxylin and molecular biology grade water (1:1 mixture), followed by several rinses in 1X Bond wash buffer and distilled water, this is not the hematoxylin provided with the Refine kit. Once the immunochemistry process was completed, slides were removed from the stainer and rinsed in tap water for five minutes. Slides were dehydrated in increasing concentrations of ethyl alcohol and cleared in 3 changes of xylene prior to permanent cover slipping in xylene-based medium.

IMMUNOFLUORESCENCE STAINING FOR EXT1 AND EXT2

Immunofluorescence staining was performed on FFPE sections retrieved for 30 min using target retrieval solution high pH (Dako). The EXT1 primary antibody (rabbit polyclonal, Thermo Scientific #PA5-60699) and EXT2 primary antibody (rabbit polyclonal; Abcam) were diluted to 1:100 in blocking solution (2% fetal calf serum and 2% normal goat serum) and incubated overnight at 4°C with retrieved biopsy sections . Next, the slides were incubated with goat Alexa488-conjugated anti-rabbit Fab IgG antibodies as secondary antibody (Life technologies). Finally, slides were mounted in mounted medium (Thermo Scientific) and covered with LDS2460EP cover glasses. Stained sections were evaluated by using inverse microscope Olympus IX83.

WESTERN BLOTTING

The protein samples, recombinant human EXT1, recombinant human EXT1/EXT2 heterodimer (both R&D Systems) and recombinant human PLA2R1 (Origene Technologies), were diluted with non-reducing Laemmli sample buffer (Bio-Rad) and boiled for 5 min. Samples were loaded into Criterion 4-15% TGX gels (Bio-Rad) and electrophoresed in Tris-glycine-SDS running buffer. Proteins were transferred to poly (vinylidene difluoride) membranes according to standard protocols, then membranes were blocked with Pierce Protein-Free Blocking buffer (Thermo Scientific). Membranes were incubated overnight at 4°C with sera from patients, controls (dilution 1:50) and rabbit polyclonal antibodies against EXT1 and EXT2 (Thermo Scientific and Abcam, respectively). Subsequently, blots were washed and incubated for 2 h at room temperature with goat anti-human IgG, HRP conjugate (Millipore) or with goat anti-rabbit HRP conjugated secondary antibody. Immunoreactive proteins were visualized with SuperSignal West Pico Chemiluminescent substrate (Pierce) followed by luminescence detection with Ozyme Syngene LED imager.

Slot blot native blotting. Protein slot blotting was done using Bio-Dot SF assembly apparatus (Bio-Rad). EXT1 and EXT1/2 proteins were added to the wells. We used nitrocellulose membrane to slot recombinant EXT1 and EXT1/2 proteins under vacuum. All blocking and washing steps of the membrane were done according to manufacturer's instruction. Subsequently, membranes were removed from the apparatus and small pieces with antigens were prepared for overnight incubation with sera from patients (dilution 1:50) and rabbit polyclonal antibodies against EXT1 and EXT2. Subsequently slots were washed and incubated with goat-anti human or goat anti-rabbit IgG, AP conjugate (Sigma). Immunoreactive proteins were visualized with BCIP/NBT liquid substrate system (Sigma).

Table 1: Clinical and laboratory findings of Validation Cohort

N°	Code	Year of biopsy	Exostosin	Age at kidney biopsy/ Gender/ Ethnicity	Rash/Arthritis/Other	Serum Creatinine (mg/dL)	Proteinuria (g/24 hours)	C3 (0.74-1.43)/ C4 (0.19-0.43g/L)	ANA/ dsDNA/ Other*	Clinical presentation**
1 "Primary" (double negative PLA2R and THSD7A) MN										
1	T2N00129	2012	-	28/M/African	N/N/N	0.7	5.3	2.18/0.62	80/neg/NS	Nephrotic Sd
2	12N00288	2012	-	58/M/African	N/N/N	1	2.9	1.19/0.49	ANA neg/neg/NS	Nephrotic Sd
3	14TN330	2014	-	61/M/Caucasian	N/N/ polyneuropathy, Sjögren	0.8	4.8	NA/NA	ANA neg/neg/NS	Nephrotic Sd
4	15TN00498	2015	-	27/F/African	N/N/N	0.6	6.1	NA	NA	Nephrotic Sd
5	11N00262	2011	-	54/F/Caucasian	N/N/N	0.6	12	1.7/0.41	1280/neg/Anti-centromere	Nephrotic Sd
6	T08E00791	2008	-	40/F/Caucasian	Y/N/N	0.9	17.3	1.12/0.26	80/neg/neg	Nephrotic Sd
7	05H07788	2005	-	58/F/Sri Lanka	N/N/Y myalgias, polyadenopathies	0.7	2.8	1.47/NA	200/neg/neg	Nephrotic Sd
8	09H05897	2008	-	42/M/Caucasian	Y/N/Y Raynaud	0.9	0.75	NA/NA	160/neg/neg	Proteinuria
9	08H0466	2008	-	76/M/Maghrebian	N/N/N	1	6.0	NA/NA	ANA neg/neg/neg	Nephrotic Sd
10	07E 00106	2007	-	34/F/Caucasian	N/N/Raynaud, sicca syndrome	0.7	1.5	0.93/0.18	160/neg/neg	Proteinuria
11	06H06087	2006	-	84/F/Caucasian	N/N/N	0.7	0.7	0.97/0.13	NA	Nephrotic Sd
12	18TN00253	2018	-	26/F/African	N/N/N	0.6	3.4	1.23/0.20	1280/neg/SSA	Proteinuria
13	12N00418	2012	-	41/M/African	N/N/N	0.9	0.2	1.12/0.12	640/neg/NS	Nephrotic Sd
14	**06H11586**	**2006**	+	24/F/Maghrebian	N/N/N	0.6	1.2	0.96/0.17	ANA neg/neg/SSA	Nephrotic Sd

	12N271	**2012**	+		Y/Y/thrombocytopenic purpura	0.5	1.8	0.73/0.11	1280/neg/Sm.RNP.SSA.B	Clinical lupus
15	**18TNN00285**	**2018**	+	70/M/African	N/N/N	2.2	13.3	0.70/0.30	1280/neg/Sm.RNP.SSA	Nephrotic Sd
16	**T07H03197**	**2007**	+	21/F/Caucasian	N/N/N	0.7	NA	1.43/0.35	1000/neg/RNP	Nephrotic Sd
	T08H07707	**2008**	+		Y/N/Y pleuritis	0.7	5.7	0.86/0.37	320/neg/RNP	Clinical lupus
	18TN00273	**2018**	+		N/N/N	0.5	4.8	0.71/0.12	1200/neg/RNP	Asymptomatic lupus
	2.Class V lupus MN									
17	**17TN00433**	**2017**	+	20/F/Caucasian Hispanic	N/Y/Y	0.5	6.3	1.04/0.30	No/8/SSA.Ro	Clinical lupus
18	**18TN00197**	**2018**	+	49/F/African	N/N/thrombocytopenic purpura	0.7	2.6	N/**0.15**	320/ neg/Sm. SSA. RNP	Proteinuria
19	**T08H10866**	**2008**	+	27/F/African	Y/Y/N	0.8	5.3	1.26/low C4	1280/NA/SSA	Clinical lupus
	17TN00426	**2017**	+		Y/N/N	0.7	0.5	0.68/0.10	1280/26/Sm. SSA. RNP	Clinical lupus
20	**T05H08855**	**2005**	+	26/F/Guyane	Y/Y/Y lung emboli	0.8	10	0.52/0.04	1000/340/Sm. SSA. RNP	Clinical lupus
	T06H02989	**2006**	+		N/N/N	0.8	2	0.71/0.08	1000/68/SSA	Asymptomatic lupus
21	**T00H02015**	**2000**	+	46/F/African	Y/Y/Y sicca syndrome	0.5	3	0.74/0.2	1000/neg/RNP	APLS & clinical lupus
22	**T01H01069**	**2001**	+	38/F/African	N/N/Y hemolysis Coombs pos	0.7	7	N/0.16	1000/23/SSA	Renal lupus
23	**T13N00303**	**2013**	+	38/M/African	N/Y/Y lung, lymph nodes	1.0	1.4	N/N	1280/neg/Sm. RNP. SSA	Clinical lupus
	17TN00061	**2017**	+		N/N/Y lymph nodes	0.7	2.8	0.19/0.07	1280/neg/Sm. RNP. SSA	Clinical lupus
24	**T13N00177**	**2013**	+	45/F/African	Y/Y/Y pericarditis	0.7	3.2	0.85/0.15	1280/neg/Sm.RNP	Clinical lupus
25	14TN00361	2014	-	32/F/African&Caucasian	Y/Y/N	0.7	0.7	0.62/0.15	1280/119/SSA	Clinical lupus
26	T02H10849	2002	-	40/F/Maghrebian	Y/Y/Y vasculitis	0.7	1.1	0.49/0.08	4000/27/neg	Clinical lupus
27	T03H00890	2003	-	41/F/Caucasian	Y/Y/N	0.9	<0.1	1.09/0.29	200/neg/NS	Clinical lupus
28	T04H09991	2004	-	Pakistani	N/N/Y diarrhea, lymph node	1.0	6.1	N/ 0.04	1000/neg/Sm. RNP	Clinical lupus
29	T11N00384	2011	-	35/F/Guyana	N/Y/N	0.7	0.4	0.93/0.13	1280/150/SSA	Clinical lupus
30	T05H01461	2005	-	54/M/Maghrebian	Y/N/Y colitis, spinal amyotrophy	0.7	0.4	0.51/0.05	NA/NA	Clinical lupus
31	18TN00204	2018	-	58/F/African	N/Y/Y pericarditis	0.6	2	1.68/0.33	1280/neg/Sm. RNP	Clinical lupus
32	T12N00232	2012	-	45/F/Caucasian	Y/Y/Y neurolupus	0.5	3	0.74/0.22	320/24/NS	Clinical lupus
33	17TNN00327	2017	-	35/M/African	Y/Y/Y	0.6	0.8	0.58/0.12	1280/157/Sm. RNP.SSA	Clinical lupus

34	12N00258	2012	-	55/F/Caucasian	N/Y/Sjögren	0.7	8.9	1.28/0.24	160/neg/SSA	Clinical lupus
3. Mixed class (V+III/IV) lupus nephritis										
35	**13N00201**	**2013**	+	34/M/African	Y/N/Y stroke	1.1	1.4	0.78/0.04	1280/231/Sm.RNP	Clinical lupus
36	T03H10353	2003	-	48/F/African	N/Y/Y seritis, Sjögren	1.1	1	0.66/0.18	1000/180/NS	Clinical lupus
37	03H00679	2003	-	17/F/African	N/N/ Y seritis, pseudo-lymphoma	1.5	6	0.51/0.03	NA	Clinical lupus
38	07E 00028	2007	-	16/F/ Caucasian	NA	NA	NA	NA	NA	Proteinuria
39	T1N00246	2011	-	42/F/Chinese	N/N/Y Raynaud syndrome	0.6	5.9	1.08/0.21	1280/neg/neg	Clinical lupus
40	T13N00124	2013	-	38/F/African	Y/Y/Y hematological, pancreatitis	0.7	5.0	0.81/0.18	NA/985/RNP.SSA	Nephrotic Sd
41	T13N00089	2013	-	64/F/Maghrebian	NA	ESKD	ESKD	NA	NA	ESKD
42	14TN00098	2014	-	49/M/Asian (Vietnamese)	N/Y/ hematological, pericarditis	1.9	0.8	0.14/0.02	2800/44/ECT	Clinical lupus
43	14TN00109	2014	-	29/ F/Asian	Y/Y/Y seritis, hematological	0.8	1.55	0.42/0.06	640/46/SSA	Proteinuria/hematuria
	14TN00312	sept-14	-			1	0.4	0.96/0.24	NA/NA	Asymptomatic lupus
44	14TN00316	2014	-	26/M/Caucasian	Y/N/Y hematological&SAPL	0.9	1.1	0.17/0.02	1280/50/RNP.SSA	Clinical lupus
45	17N00320	2017	-	55/F/Maghrebian	Y/Y/N	1.3	2	C3 normal/0.18	160/neg/NS	Asymptomatic lupus
46	18TN00279	2018	-	30/F/Asian (Cambodian)	Y/Y/Raynaud	0.8	3.0	0.61/0.07	1280/109/Sm. RNP. SSA	Clinical lupus
47	14TN00235	2014	-	43/F/Caucasian	Y/Y/Y hematological, neurolupus	0.8	3.3	0.6/0.08	320/50/NS	Nephrotic Sd
48	17834	2000	-	34/M/Maghrebian	Y/Y/Y	66	0.2	0.47/0.10	1000/16/RNP.SSA	Asymptomatic lupus

*ANA=antinuclear antibody, dsDNA= anti-double stranded DNA antibody; RNP=ribonucleoprotein, SSA/B=Sjögren syndrome antibody A/B, NA=not available, NS=not specified

**Sd=syndrome

Table 2: Kidney biopsy findings of Validation Cohort

N°	Code	Glomeruli/ sclerosed	Mesangial or endocapillary hypercellularity*	Interstitial Inflammation/ IFTA	Arteries	Immunofluorescence microscopy	Pathological diagnosis	IgG Subclass
1. "Primary" (double negative PLA2R and THSD7A) MN								
1	T2N00129	6/0	Not present	0/0	Normal	G subepithelial	MN stage I	Ig4=3>1
2	12N00288	19/6	Not present	Mild/mild	Intimal fibrosis	G subepithelial, mesangial	MN stage II	IgG1=3>2
3	14TN330	7/0	Not present	0/0	Intimal fibrosis	G, A (weak), C3 subepithelial	MN stage II	IgG1
4	15TN00498	18/1	Not present	0/0	Intimal fibrosis	G subepithelial	MN stage I	IgG4
5	11N00262	26/0	Not present	0/0	Normal	G,C3 subepithelial	MN stage I	
6	T08E00791	19/0	Not present	0/0	Normal	G,C3,C1q subepithelial	MN stage I	
7	05H07788	5/1	Not present	0/0	Normal	G,C3,C1q subepithelial,	MN stage I , segmental	
8	09H05897	12/0	Not present	0/0	Normal	G,M,C3 subepithelial	MN stage II,III	
9	08H0466	15/4	Not present	Mild/mild	Intimal fibrosis	G subepithelial	MN stage II	
10	07E 00106	22/4	Not present	0/mild	Normal	G subepithelial	MN stage III	
11	06H06087	8/3	Not present	Severe (CLL)/mild	Normal	G,C3 subepithelial kappa & lambda	MN stage III	
12	18TN00253	9/0	Not present	0/0	Normal	G,M,C3	MN stage I	IgG4>G1=G2
13	12N00418	28/1	Not present	0/mild	Normal	G subepithelial, C3 arterioles	MN stage II	IgG1>3>2
14	06H11586	31/0	Prolif. mes., small crescents	0/mild	Normal	G, C3 subepithelial	MN stage I-II	
	12N271	22/1	Not present	0/0	Normal	G,C3 subepithelial	MN stage I-II	
15	18TNN00285	10/1	Not present	0/0	Normal	G,C3 subepithelial	MN stage I	IgG4>G1
16	T07H03197	19/0	Not present	0/0	Normal	G, C3 subepithelial; M, mesangial	MN stage I	
	T08H07707	26/0	Prolif.endo., thrombi	Mild/0	Normal	G,A,M,C3,C1q	Class III S-A+ V	
	18TN00273	18/1	Not present	0/0	Normal	G,C3,C1q	Class V	

2. Class V lupus MN								
17	17TN00433	29/0	Not present	0/0	Normal	G,A,M,C3	MN stage I	
18	18TN00197	12/0	Not present	0/0	Normal	G, C3, C1q	MN stage I	IgG1 >IgG2
19	T08H10866	15/0	Not present	0/0	Normal	G,C3,C1q subepithelial, M mesangial	Class V	
	17TN00426	8/0	Not present	0/0	Normal	G,C3	MN stage I	
20	T05H08855	10/0	Prolif. endo., crescents	Mild/mild	Normal	G,A,C3,C1q	Class IV-(G)A+ V	
	T06H02989	6/0	Prolif. decreased	0/0	Normal	G,A,C3,C1q	Class V	
21	T00H02015	10/1	Not present	0/0	Normal	G,A,C3,C1q subepithelial, G,M mesangial	Class V	
22	T01H01069	8/0	Not present	0/0	Normal	G, (A), C3, C1q subepithelial, C1q endomembranous	Class V	
23	T13N00303	20/0	Not present	0/0	Normal	G,C3,C1q	MN stage 1 (Class V?)	IgG1
	17TN00061	34/2	Not present	Severe/0	Normal	G,C3, (A), (C1q)	MN stage 2	IgG1,2,3 &4
24	T13N00177	11/0	Not present	0/0	No artery	G,C3 subepithelial, G M C3C1q mesangial	Class V	
25	14TN00361	15/2	Mild mes. prolif.	0/0	Normal	G,C3, C1q, subepithelial; GAMC3C1q mesangial	Class V	
26	T02H10849	9/2	Not present	0/mild	Normal	G,A, C3,C1q subepithelial, M mesangial	Class V	
27	T03H00890	23/2	Mild mes. prolif.	Mild/mild	Normal	G,A,M,C3,C1q subepithelial, mesangial(small)	Class V	
28	T04H09991	17/2	Prolif. mes.	Focal /0	Mild endarteritis	G,A,C3, C1q subepith, G, A, M, C3 subendothelial	Class V	
29	T11N00384	08/1	Not present	0/0	Intimal fibrosis	G subepithelial, M, C3 parietal	Class V	
30	T05H01461	19/3	Not present	mild/mild	Fibrous endarteritis	G,A,C3,C1q, subepithelial and vascular	Class V	
31	18TN00204	27/2	Not present	0/mild	Intimal fibrosis	G,A,C3 subepithelial	MN stage 1 (Class V?)	
32	T12N00232	18/5	Not present	0/APS-related fibrosis	Thrombosis (APS)	G, C3, C1q subepithelial	Class V + APS	
33	17TNN00327	15/2	Not present	0/mild	Normal	G,C3 subepithelial	Class V	IgG3
34	12N00258	20/2	Not present	0/0	Intimal fibrosis	G subepithelial, mesangial	MN stage 1	IgG4=2>1=3

3. Mixed class (V+III/IV) lupus nephritis								
35	13N00201	6/0	Prolif.mes. (1G)+ 1 crescent	Severe (C/M)/tubulitis	Intimal fibrosis	G,C3 subepithelial, M,C3 mesangial	Class III+V	IgG2>1=3=4
36	T03H10353	23/2	Prolif.endo., necrosis, crescent	Mild/mild	Endarteritis (APLS)	NA	Class III+V	
37	03H00679	26/0	Prolif.endo., mes., crescent	0/mild	One thrombosis	G,A,M,C3,C1qsubepitelial, mesangial	Class III+V	
38	07E 00028	14/5	Not present	0/mild	Normal	G,A,C3,C1q subepithelial	Class IVc +V	
39	T1N00246	24/0	Prolif. endo.	0/mild	Intimal fibrosis	G,M, C3, C1q subepithelial	Class III + V stage II, III	
40	T13N00124	18/3	Scars, synechias	Mild/o	N	G,A,M,C3,C1q subepithelial and endomembranous	Class IV-S (C:68%) + V	
41	T13N00089	18/9	Prolif. endo. &extracapillary	Severe/severe	Intimal fibrosis	G,A,C3 subepithelial	Class IV-G (A, 27%; C, 78%) + V	
42	14TN00098	16/2	Prolif. endo. & thrombi	0/mild	Intimal fibrosis	G=C1q> A=C3, subepithelial and endomembranous	Class IV-S (A) + V	
43	14TN00109	22/1	Prolif. endo. & extracapillary	Mild/0	Intimal fibrosis	G,A, C3, C1q subepithelial and mes.	Class III (A,14%; C, 27%) + V	
	14TN00312	21/5	Scars	0/0	Normal	G,A,C3 subepithelial, C1Q endomembranous segmental	Class IIIC + V	
44	14TN00316	9/0	Prolif. endo.	0/0	Normal	G,C1q,C3 subepithelial &endo, A,M endomembranous	Class IIIA + V	
45	17N00320	11/5	Not present	0/mild	Intimal fibrosis	G,A subepithelial	Class IVC + V	
46	18TN00279	19/0	Prolif. endo.	Mild/mild	Normal	G,A,M,C3,C1q subepithelial & endomembranous	Class IIIA + V	
47	14TN00235	17/4	Prolif. endo.	0/mild	Normal	G,A,M,C3,C1q, subepithelial & endo.	Class IV-G (A/C) + V	
48	INSERM 17834	21/0	Prolif. endo. &extracapillary	Mild/mild	Intimal fibrosis	NA	Class III+V	

*Prolif.=proliferation, mes.=mesangial, endo.=endothelial, 1G=one glomerulus