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Letters to the Editor

Fibronectin glomerulopathy in a 88 year-old male with acute kidney injury on chronic kidney disease: A case report and a review of the literature

Glomerulopatía fibronectina en un anciano de 88 años de edad con lesión renal aguda en enfermedad renal crónica: reporte de un caso y revisión sistemática de la literatura

Dear Editor,

Fibronectin glomerulopathy (FGP) was first recognized through direct staining in 1992 by Mazzucco et al.[1] It was regarded as a rare autosomal dominant disease that tends to happen in various age groups and affects both sexes. The clinical features of this disease were proteinuria, typically nephrotic range, microscopic hematuria, hypertension and a slow decline in renal function.[1–11] However, some patients progress to end-stage renal disease (ESRD) and dialysis has been reported.[5,12–14] Herein we reported a case of FGP that was diagnosed based on the classic ultrastructural features described above. This case was unique in that FGP presents in a male who was 88 years of age and has no known family history of FGP.

An 88-year-old gentleman was admitted with anuria requiring dialysis for uremic encephalopathy and volume overload. He had an extensive past medical history including chronic renal failure (baseline Cr 200–240 μmol/L) one month ago, hypertension, obstructive sleep apnea, mild cognitive impairment, query previous congestive heart failure, diastolic dysfunction, COPD, macular degeneration, benign prostatic hypertrophy and an anemia that was being investigated as an outpatient. Baseline level of proteinuria was unknown. There was no family history of kidney disease.

Relevant initial blood work included a serum creatinine of 1148 μmol/L, urea 82.2 mmol/L and potassium 5.7 mmol/L. During his stay in hospital, a 24-h urine collection was done and only 100 mL of urine were collected with 100 mg of protein and a protein excretion of 1 g/L. The other results of blood examination are as follows: a normal ESR, C3 and C4 values, negative ANA, c-ANCA, p-ANCA, anti-GBM antibody, hepatitis B and C serology, renal ultrasound showed normal size of the kidney. The extranuclear antigen screen was negative except RNP. Immunofixation revealed a serum IgG M-protein with kappa light chain specificity. Bone marrow studies were unremarkable.

A renal biopsy was performed and light microscopy showed a lobular appearance with cellular mesangial nodules that were expanded by matrix. There was also prominent arteriolar fibrointimal hyperplasia and arteriolar hyalinosis. Congo red staining was negative for amyloid. Immunostaining was 2+ for C3 in a granular pattern in the glomerular capillary walls, however staining was negative for IgA, IgG, IgM, kappa, lambda, albumin, fibrin and C1. Electron microscopy showed prominent expansion of the mesangial and subendothelial regions with numerous randomly arranged fibrils seen under high power lens, measuring 10.7–16.8 nm in diameter. Polyclonal rabbit antibody (DAKO, A0245) was used in staining of fibronectin and there was positivity in the mesangium, mesangial nodules and capillary walls (Fig. 1).

The patient received regular hemodialysis and other supportive treatment including anti-hypertension, improving anemia, and preventing renal osteopathy. Unfortunately, this patient died of heart attack half a year later.

To our knowledge, most cases of FGP have presented and been diagnosed in patients under 65-year-old. As this patient was diagnosed with FGP at the age of 88, we reviewed the cases reported in literatures and attempted to explain how

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Table 1 – Reported cases of fibronectin glomerulopathy.

Reporter	No.	Age at Bx	Femal:male	Hypertension	Proteinuria (g/24 h)	Micro-hematuria	Cr (μmol/l)	Time of last outcome	Last renal outcome
Mazzucco et al. (1992)[1]	2	14, 33	2:0	NO	2 patients: nephrotic	N/A	53,141	1 patient: 4 yr	1 patient: Cr was stable
Strom et al. (1995)[2]	23	14–59	7:16	10 patients: YES 8 patient: NO	21 patients: positive (9 patients: nephritic)	12 patients: positive 7 patients: negative	45.8–137	1–15 yr	9 patients: Cr was stable 4 patients: dialysis 4 patients: transplant 4 patients: dead 1 patient: normal Cr, renal cell carcinoma
Assmann et al. (1995)[12]	2	19, 46	0:2	1 patient: YES 1 patient: NO	2.8,6	N/A	76, 100	N/A	N/A
Gemperle et al. (1996)[13]	9	20–46	3:6	8 patients: YES 1 patient: NO	7 patients: positive (1 patient: nephrotic)	N/A	N/A	6–21 yr	2 patients: hemodalysis 2 patients: CAPD 3 patients: dead
Fujigaki et al. (1997)[3]	1	47	1:0	NO	1.4	NO	53	9 yr	0.82
Sato et al. (1998)[15]	1	23	0:1	YES	1–2	NO	80	N/A	N/A
Niimi et al. (2002)[4]	1	3	0:1	YES	>8	YES	80	7 yr	Cr was stable
Castelletti et al. (2008)[5]	8	12–59	1:7	3 patients: YES 4 patient: NO	6 patients: positive (4 patient: nephrotic)	2 patients: positive 1 patients: negative	<194.5	2–23 yr	4 patients: Cr was stable 2 patients: dialysis 1 patients: transplant 1 patients: dead 1 patient: normal Cr, renal cell carcinoma
Yong et al. (2009)[6]	1	41	0:1	YES	Nephrotic	N/A	150	10 mo	1.2
Brcic et al. (2010)[7]	1	34	0:1	YES	6	YES	N/A	21 mo	Cr was stable
Nadamuni et al. (2012)[8]	1	50	1:0	NO	Nephrotic range	YES	70.7	1	0.8
Otsuka et al. (2012)[9]	1	49	0:1	NO	<0.5 g/d	N/A	92	9 mo	1.54
Ertoy et al. (2013)[10]	1	24	0:1	NO	1	NO	72.5	16 mo	
Ishimoto et al. (2013)[14]	1	78	1:0	YES	Nephrotic range	N/A	101	2 mo	Dialysis
Yoshino et al. (2013)[11]	1	67	M	YES	6	N/A	88.4	7 yr	3.54
Chen et al. (2015)[16]	10	19–46	4:6	4 patients: YES 6 patient: NO	10 patients: positive (2 patient: nephrotic)	4 patients: positive 6 patients: negative	47–312	N/A	N/A

HTN: hypertension.
Bx: biopsy.
Note: If a particular piece of information was not available, the space is filled with "N/A".