AN UNCOMMON PRESENTATION OF AN UNCOMMON NEPHROPATHY: THE KARYOMEGALIC INTERSTITIAL NEPHRITIS.

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INTRODUCTION

Karyomegalic interstitial nephritis (KIN) is a rare nephropathy (hitherto, less than 30 cases have been described in literature) firstly described in 1974, of unknown etiology, histologically characterized by interstitial nephritis and hyperchromatic, abnormally enlarged nuclei of tubular epithelial cells, and presenting in young patients with progressive renal failure, proteinuria and/or hematuria and a history of recurrent respiratory infections. An unspecific elevation of liver enzymes has been also observed. Herein, we report a case of KIN with atypical clinical presentation diagnosed in a young patient with progressive kidney failure without proteinuria or hematuria or history of recurrent respiratory infections. In this patient, the unusual clinical presentation did not help in including KIN in the initial differential diagnosis. Thus, this case emphasizes once more the importance of performing a renal biopsy in all cases of nephropathies of unclear etiology, even in patients with established chronic kidney disease (CKD).

CASE PRESENTATION

A 33-years old Caucasian man was referred to our Unit because of impaired renal function. The history was negative for familial kidney diseases, recurrent respiratory infections and occupational exposure to dangerous substances. At the time of admission, physical examination was unremarkable. GFR was 36.8 ml/min and no urinary abnormalities were detected. Liver enzymes were slightly increased (Table 1). Serology for hepatotropic and nephrotropic viruses was negative and serum complement and the titers of auto-antibodies were normal. Renal ultrasound evidenced a longitudinal diameter of 90-95 mm, a slight reduction of the renal cortex thickness without evidence of urinary stones or urostasis, and a normal Doppler velocimetric pattern on both renal arteries. A ^{99m}Tc-MAG3 renal scan evidenced a slightly reduced bilateral kidney perfusion (effective renal plasma flow 260 ml/min) and a regular radiotracer excretion. Thus, a renal biopsy was performed.

RESULTS

Immunofluorescence microscopy detected only a weak positivity for IgM in some scarred glomeruli. Light microscopy obsolescence of 17 out of 26 glomeruli was observed, the remaining showing only a mild expansion of mesangial matrix, with normal glomerular cellularity and normal glomerular capillary basement membranes. Tubulo-interstitium showed areas with various degrees of fibrosis, mononuclear infiltrates and presence of large hyperchromatic nuclei with irregular outlines in epithelial cells lining proximal and distal tubules (Figure 1A). Arterial vessels showed mild intimal fibrosis. Electron microscopy evidenced irregular distribution of chromatin within the enlarged nuclei of the tubular epithelium (Figure 1B), but no viral particles or electron-dense deposits. Thus, in consideration of these histological findings, a diagnosis of KIN was made, and therapy with anti-angiotensin agents and low dose anti-aldosteronic agent was started. After 12 months of follow-up, no progression of renal insufficiency was observed and urinalysis was still normal (Table 1).

Figure 1: (A) Hematoxilyn/eosin staining, x400. (B) Uranyl-lead staining, x6,000.

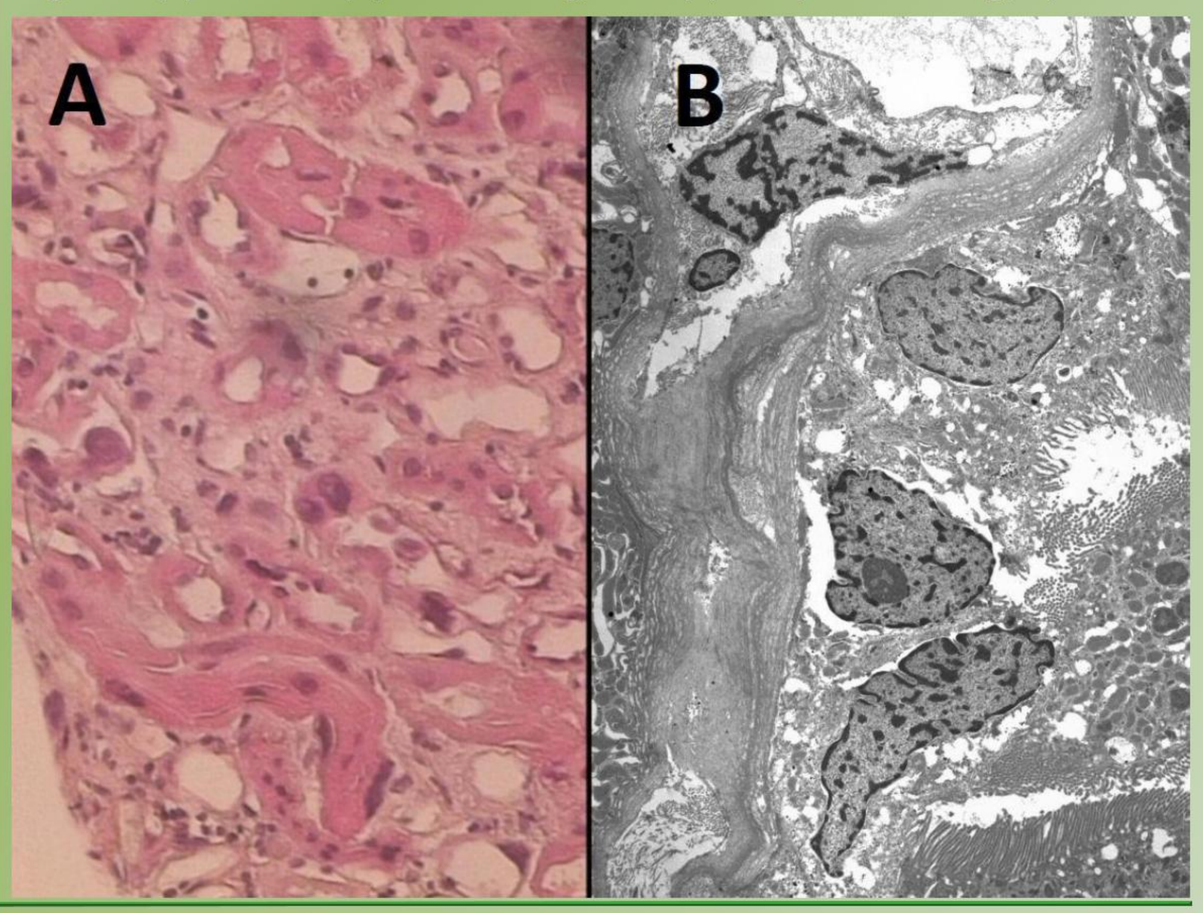

Table 1: parameters collected at the time of kidney biopsy and 6 and 12 months later.

Parameter	Baseline (kidney biopsy)	6 months follow-up	12 months follow-up
Creatinine (mg/dL)	2.2	2.1	2.3
eGFR MDRD (ml/min)	36.8	38.6	34.8
Urea (mg/dL)	63	59	88
Hemoglobin (g/dL)	13.3	13.2	12.8
Aspartate-aminotransferase (U/l)	36	44	44
Alanine-aminotransferase (U/l)	61	82	101
Calcium (mg/dL)	9.9	10.5	9.7
Phosphorus (mg/dL)	3.1	3.1	4.5
Systolic blood pressure (mmHg)	100	110	100
Diastolic blood pressure (mmHg)	80	70	70
Body mass index (kg/m^2)	23.9	23.9	23.5
Proteinuria (g/24h)	< 0.15	< 0.15	< 0.15
Hematuria	Absent	Absent	Absent
Low urinary tract infections	Absent	Absent	Absent

CONCLUSIONS

This case suggests that KIN is probably an underdiagnosed condition, because it can occur also in absence of the traditional clinical and laboratory findings, making the diagnosis absolutely incidental. Therefore, young adults with pauci-symptomatic CKD of unknown origin should be considered for routine renal biopsy to obtain a correct diagnosis that provides information in order to avoid any empirical use of harmful drugs and to encourage living-donor transplant, since no recurrence of the disease has been hitherto described.

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