Glomerulocystic kidney disease presenting as acute renal failure in an adult patient

Sir,

The diagnosis of glomerulocystic kidney disease (GCKD) in adults is usually an incidental finding during the evaluation of other working diagnoses. Hereby we report the case of a 55-year-old woman admitted for an elective second prosthetic mitral valve replacement for rheumatic mitral valvulopathy who presented acute renal failure (ARF). Four months before, when a diagnosis of granuloma annulare was made, her plasma creatinine was 1.8 mg/dl and urinanalysis showed haematuria without proteinuria. Her father, aged 60 years, had died of chronic renal failure of unknown cause. Twenty days before hospital admission she had pneumonia and was treated with penicillin and diclofenac for 10 days. She was regularly taking atenolol and hydrochlorothiazide. On admission she was normotensive. Urinary volume was ~1000 ml/day. Laboratory tests showed plasma creatinine = 6 mg/dl, blood urea nitrogen = 90 mg/dl, plasma sodium = 140 mEq/l, plasma potassium = 5.2 mEq/l, ionic calcium = 4.4 mg/dl, phosphate = 10 mg/dl, haemoglobin = 9.6 g/l, leukocytes = 6600/mm³ and platelets = 120000/mm³. Urinalysis showed pH = 6, gravity = 1014, leukocytes = 144000/ml, red blood cells = 145 000/ml. Proteinuria was 1.6 g/24 h. Urine culture was negative. Ultrasonography showed normal-sized kidneys with mild hyperechogenicity and a cortical cyst of 2.4 cm in the left kidney. The patient’s renal function worsened and she required haemodialysis for 2 weeks. Renal biopsy showed marked dilatation of Bowman’s space in 40% of glomeruli with the correspondent glomerular tufts collapsed (see Figure 1). No periglomerular fibrosis was found. Dilated tubuli with flattened epithelial cells or with regenerative changes were seen in some areas. The arteries had no alterations. The final diagnosis was GCKD plus regenerating acute tubular necrosis. In the 40th hospital day she was discharged, having undergone uneventful heart surgery on the 20th hospital day, with plasma creatinine = 2.5 mg/dl. Thirty days after discharge, she maintained the same creatinine and microscopic haematuria.

GCKD is more often described in infants and young children than in adults. Nowadays GCKD is categorized into three groups according to its clinical presentation: (i) sporadic or hereditary non-syndromal forms found in children or adults; (ii) hereditary syndromal forms; and (iii) glomerular cysts found in dysplastic kidneys [1]. The sporadic form of GCKD associated with other renal diseases has been designated acquired [2,3]. Although hypercalcaemic ARF had been reported in association with annular granuloma, our patient did not have hypercalcaemia that points to the presence of annular granuloma be incidental [4]. Her father having had renal failure suggests a heritable non-syndromal form of GCKD, where urinanalysis can be normal or show non-nephrotic proteinuria and haematuria as in the case of our patient. Ultrasonography can show characteristic findings, such as a hypoechoic cortical rim or less characteristic patterns, such as exclusive cortical cysts, loss of corticomedullary differentiation, hypoplastic or diffusely echogenic kidneys [5]. The patient’s ultrasound showed mild cortical hyperechogenicity and only a cortical cyst. Although there was no consensus on the histological criteria to define GCKD, Berstein’s definition is the most strict: presence of dilatation of Bowman’s space of two to three times in the plane of section in at least 5% of the glomeruli [1]. Our patient certainly fits this definition: 40% of her glomeruli showed marked dilatation of Bowman’s space. The finding of superimposed acute tubular necrosis can explain the worsening of renal function and subsequent partial recovery in our patient. ARF was attributed to the prolonged use of diclofenac. The reported case points to the importance of performing renal biopsy in ARF of uncertain cause and, to the best of our knowledge, this is the first report of GCKD associated with acute tubular necrosis.

Conflict of interest statement. None declared.

Hospita das Clinicas
Nephrology Division
University of São Paulo
São Paulo, SP
Brazil
Email: kader@usp.br

doi:10.1093/ndt/gfh995