NDT Plus (2010) 3: 349–350
doi: 10.1093/ndtplus/sfq048
Advance Access publication 13 April 2010

NDT PLUS
Nephrology Dialysis Transplantation

Case Report

Glomerulocystic disease

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Abstract

Glomerulocystic disease is a rare cause of cystic kidney diseases and can occur at any age. It is characterized by cystic dilatation of the Bowman's capsule and normal tubules, and needs to be differentiated from other cystic renal diseases. It commonly presents as renal failure. We present a case of a 52-year-old female, with renal failure who was subsequently found to have glomerulocystic disease on renal biopsy.

Keywords: cysts; glomerulocystic disease; kidney

Introduction

Glomerulocystic kidney disease is an uncommon type of cystic renal disease. It is characterized by cortical microcysts, which are represented by cystic dilatation of Bowman's spaces. It differs from the better known cystic renal diseases.

Case report

Mrs B, a 52-year-old female, presented to the hospital with chief complaints of vomiting, loss of appetite and swelling feet of 3 months duration. There was no history of oliguria, dysuria, haematuria or lithuria. She had no obstructive urinary symptoms. There was no fever or drug abuse prior to the onset of these symptoms. The patient denied any history of rash or joint pains. The family history was not significant. She had one child who was normal. Physical examination was unremarkable but for pallor and hypertension [blood pressure (BP) of 160/90 mmHg]. All peripheral pulses were well felt. Complete urine examination revealed trace protein and no red blood cells. Blood biochemistry showed serum creatinine 9 mg/dL (686.24 mmol/L), blood urea 180 mg/dL (128.52 mmol/L), serum sodium 138 mmol/L, serum potassium 4.4 mmol/L, serum calcium 8.8 mg/dL (2.2 mmol/L), serum phosphorus 4.9 mg/dL (1.58 mmol/L), serum bilirubin 0.8 mg/dL (13.68 μmol/L), serum aspartate amino transferase (AST) 13 U/L, serum alkaline phosphatase 140 U/L, plasma proteins 66 g/L, plasma albumin 40 g/L, haemoglobin 80 g/L, total leucocyte count 8.9×10^9/L and platelet count 350×10^9/L. Ultrasound examination showed a right kidney measuring 9.3 cm and a left kidney measuring 9.8 cm with a grade 2 echotexture. There were no cysts in any other organs. Computed tomography (CT) scan showed an altered echotexture in the renal cortices with sparing of medullary pyramids. The patient received two sessions of haemodialysis with blood transfusion. Subsequently, a renal biopsy was performed which showed features of glomerulocystic disease (Figure 1). The patient subsequently was initiated on maintenance dialysis.

Discussion

Roos first described glomerulocystic disease as an isolated abnormality in 1941 [1]. However, Taxy and Filmer proposed the term of glomerulocystic kidney disease (GCKD) in 1976 and described glomerulocystic kidney in which cystic dilatation of Bowman's space was observed [2]. It

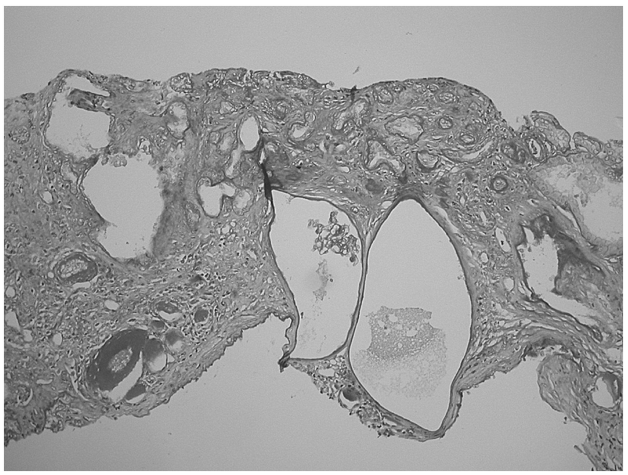

Fig. 1. Renal biopsy showing glomerular cysts (H&E) high power view—black and white. A primitive glomerulus can be seen inside the cyst 67 × 50 mm (300 × 300 DPI).

Table 1. Classification of glomerulocystic diseases

I	Familial GCKD	Autosomal dominant GCKD
II	Familial/sporadic heritable syndromes	Autosomal dominant polycystic kidney disease (ADPKD)
		Cystic renal dysplasia
		ARPKD
		Zellweger's cerebral–renal–hepatic syndrome
		Tuberous sclerosis
		Trisomy 13
		Juvenile nephronoptdisis
		Orofacial digital syndrome type 1
		Brachymeromelia renal syndrome
		Majewski-type short rib polydactyly syndrome
		Jeune's osteodystrophy
		Goldston syndrome
		Lissencephaly
III	Glomerulocystic kidneys as component of otder cystic diseases	Diffuse cystic dysplasia
		Renal–hepatic–pancreatic dysplasia syndrome
		Meckel syndrome, glutaric aciduria type 2
IV	Sporadic	
V	Acquired	Associated witd haemolytic–uraemic syndrome

is rare and is mainly reported in neonates and infants. In 1984, Dosa *et al.* described the sporadic adult form of GCKD [3]. GCKD must be diagnosed by excluding other cystic renal disorders. Characteristically, there is widening of Bowman's space (2–3× normal) with at least >5% cysts containing atrophic glomeruli [4].

It is believed to be caused by glomerulotubular neck obstruction with proliferation of epithelial cells of the Bowman's capsule, but recent studies are contradictory [5,6]. This is associated with remodelling of the basement membrane of the Bowman's capsule. Some cases are thought to be due to urinary tract obstruction [7] *in utero* or due to ischaemia [8].

GCKD can be categorized into five major groups [9] (Table 1). Familial GCKD can be associated either with hypoplastic or normal sized kidneys. In hypoplastic variant, the kidneys are glomerulocystic and small with abnormal pyelocalyceal anatomy. A mutation in the hepatocyte nuclear factor-1beta gene has been identified in hypoplastic GCKD variant [10]. The second group includes sporadic and familial disease in older children and adults. No differences between familial and sporadic cases have been identified apart from the family histories. The sporadic cases represent new mutations of the same disease. The third group includes glomerulocystic kidneys as components of other cystic diseases. The fourth group comprises all the sporadically occurring GCKD. Acquired GCKD (Group 5) has been described following haemolytic–uraemic syndrome [11].

Differential diagnosis

The main differentiating feature to distinguish GCKD and autosomal recessive polycystic kidney disease (ARPKD) is abnormal medullary pyramids in the latter. The imaging findings of small renal cysts with a predominant cortical and subcapsular distribution allow for distinction from other, more common, polycystic kidney diseases. CT scan and magnetic resonance imaging (MRI) may be valuable in distinguishing these two diseases [12].

Histology reveals glomeruli with marked dilatation of the Bowman's capsule with collapsed capillary tuft projecting into the lumen covered by a prominent layer of visceral epithelial cells. Glomeruli appear ischaemically obsolescent and small. The blood vessels show fibrointimal hyperplasia and luminal narrowing. Interstitium shows infiltrations of lymphocytes. The tubules are normal (*vs* PKD). Immunofluorescence examination reveals insignificant deposits.

Thus, GCKD is a rare cause of chronic kidney disease (CKD) and needs to be recognized in the appropriate setting.

Conflict of interest statement. None declared.

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Received for publication: 2.11.09; Accepted in revised form: 16.3.10