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Glomerulocystic kidney identified in older patients by magnetic resonance imaging

Relation to renal function and renal corticomedullary differentiation

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Abstract

Glomerulocystic kidney (GCK) is often associated with genetic disorders and identified in children or adolescents. However, there are some case reports describing sporadic adult GCK identified by magnetic resonance imaging (MRI). The purpose of this study was to evaluate relationship of GCK identified by MRI in older patients to renal function and renal corticomedullary differentiation (CMD) assessed by MRI.

GCK was identified in 16 older patients (mean age, 79.2 years) by T2-weighted imaging. The cysts of GCK were numerous, homogeneously small, and located in the renal cortex on T2-weighted images. Ten of the 16 patients with GCK had renal impairment (estimated glomerular filtration rate <60 ml/min/1.73 m^2). Six patients who had GCK, chronic liver disease, and renal impairment showed moderate or good CMD.

GCK identified by MRI may be related to renal impairment in some older patients, including those with preserved CMD as a result of chronic liver diseases.

Abbreviations: CMD = corticomedullary differentiation, eGFR = estimated glomerular filtration rate, GCK = glomerulocystic kidney, HCC = hepatocellular carcinoma, IPMN = intraductal papillary mucinous neoplasm, LC = liver cirrhosis, MRI = magnetic resonance imaging.

Keywords: chronic liver disease, corticomedullary differentiation, glomerulocystic kidney, magnetic resonance imaging, renal impairment

1. Introduction

Glomerulocystic kidney (GCK) disease is characterized histologically by uniform cystic dilatation of Bowman capsule.[1,2] The cysts are multiple, uniform in size, and located predominantly in the renal cortex; these features differ from those in other renal cystic diseases.[2,3] GCK disease is often associated with genetic disorders and identified in children or adolescents.[2–6] The disease has also been reported in adults who have some metabolic disorders or neuromuscular degeneration and progressive renal impairment.[7,8] Lennerz et al classified GCK into 5 categories and defined the GCK "diseases" strictly as including those with a genetic basis.[5] However, they also reported sporadic adult GCK, the cysts of which are observed in only 1 kidney, is associated with aging and vascular diseases.

T2-weighted magnetic resonance imaging (MRI) is a sensitive method for detecting cystic lesions and fluid collection.[3,9] The use of T2-weighted MRI to identify GCK disease, including adult cases, has been reported.[5,10,11] In these studies, T1-weighted MRI detects renal impairment as the loss of corticomedullary differentiation (CMD).[10,12] Therefore, MRI may be an ideal imaging tool for identifying GCK. We identified 13 older patients with GCK during routine abdominal MRI. The purpose of this study was to evaluate the relationship of GCK identified by MRI in older patients to their renal function and CMD.

2. Patients and methods

2.1. Patients

From November 2016 to February 2019, 3507 abdominal MRI examinations were performed to evaluate hepatic tumors, chronic liver diseases, pancreatic tumors, pancreatitis, and biliary diseases in our institution. We identified 30 MRI examinations (0.86%) of 19 patients with possible GCK by searching our radiological reporting database (ProRad, FINDEX, Tokyo, Japan) using the term "GCK". We excluded 3 patients: 1 with a history of nephrectomy, a young patient with tuberous sclerosis, and the other who was under hemodialysis. For the remaining 16 patients, we excluded a family history of polycystic

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There was no ethic approval because all diagnostic procedures and treatment were performed in the clinical routine. Written consent to participate was given by the patient. The authors have no conflicts of interest to disclose.

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kidney diseases, associated neuromuscular degenerations, or use of lithium.[2,3,5,7] Thereafter, 2 investigators with 30 years and 7 years of experience in diagnostic radiology, independently diagnosed the GCK as defined in previous studies.[5,11] The final diagnosis was determined when both radiologists made the diagnosis of GCK without the need for consensus. Consequently, 27 abdominal MRI examinations of 16 patients with GCK were enrolled. The patients included 5 men and 11 women, ranging in age from 67 to 90 years (mean, 79.2 years; SD, 5.6 years). The reasons for the MRI examinations were intraductal papillary mucinous neoplasm (IPMN, n=6), liver cirrhosis (LC) and hepatocellular carcinoma (HCC; n=4), LC (n=2), chronic hepatitis (n=2), hepatic hemangioma (n=1), and pancreatic carcinoma (n=1). The estimated glomerular filtration rate (eGFR) was measured within 35 days of the MRI (median, 0 day), and 11 of the 16 patients underwent MRI and blood sampling on the same day. All patients gave informed consent for the MRI examinations; 1 patient underwent 7, 1 underwent 3, 3 underwent 2 examinations, and the remaining 11 underwent 1 MRI study. This retrospective study was approved by our institutional review board, and informed consent for the review of medical charts was waived.

2.2. MRI

MRI examinations were carried out using a 1.5 tesla (8 patients) or 3.0 tesla (8 patients) imager (Ingenia, Philips Medical Systems, Best, The Netherlands). A torso phased-array coil was used for signal reception. Breath-hold transverse T1-weighted and transverse and coronal T2-weighted imaging with or without fat-suppression was performed. The typical imaging parameters of MRI are summarized in Table 1.

2.3. Analysis

The 2 radiologists assessed 27 abdominal MRI of 16 older patients with GCK independently. First, the characteristics of GCK on MRI, such as the numbers of cysts, their location, and the signal intensity, were described. In the 5 patients who underwent multiple MRI studies, the most recent study was assessed. Second, the number of patients who had both GCK and renal impairment (ie, Egfr $< 60\,ml/min/1.73\,m^2$) was noted. Third, CMD as observed on the T1-weighted images was defined as good, moderate, or poor, based on previous studies as follows: good, CMD was visualized clearly throughout the kidney; moderate, CMD was partially lost in the kidney; poor, CMD was diminished.[12,13] The CMD was compared with renal function in these older patients with GCK. A kappa analysis was used to evaluate the radiologists' assessment of CMD. When there was disagreement for CMD assessment between them, the consensus reading was performed.

3. Results

The cysts of GCK as visualized on the T2-weighted images were numerous, uniformly small, and located in the renal cortex in all 16 older patients (Fig. 1A, B). The cysts showed homogeneously high intensity on the T2-weighted images and were difficult to detect on the T1-weighted images (Fig. 1).

Table 2 summarizes the clinical data and CMD of the 16 patients. Ten (62.5%) of the 16 patients with GCK had renal impairment, as did 6 (75.0%) of the 8 patients with both GCK and chronic liver diseases. The CMD assessment was concordance for 29 of the 32 kidneys of the 16 older patients between the 2 readers (k=0.81), which indicated excellent agreement. The CMD on the T1-weighted images was identical for both kidneys in all but 2 patients. Five older patients with GCK and normal eGFR showed good CMD in both kidneys and 1 showed good and moderate CMD (Table 2). In the 20 kidneys of 10 patients with GCK and reduced eGFR, 12 (60%) kidneys showed good CMD, 5 (25%) showed moderate CMD, and 3 (15%) showed poor CMD. Among these 10 patients, 5 of the 6 patients with good CMD of both kidneys had eGFR $> 45\,ml/min/1.73\,m^2$. Six (75.0%) of the 8 patients with both GCK and chronic liver disease had renal impairment, but 11 (91.7%) of their 12 kidneys showed good or moderate CMD and only 1 showed poor CMD (Table 2, Fig. 1C).

4. Discussion

This study demonstrated that numerous, uniformly small, cortical cysts were identified by T2-weighted MRI in both kidneys of all older patients with GCK, and that their presence was associated with renal impairment in 10 of the 16 patients. The MRI features of GCK in these patients were identical to those of GCK in children and adolescents.[2–6] In this study, 6 of the 8 patients who had GCK, chronic liver diseases, and good or moderate CMD had renal impairment, whereas 5 of the 16 patients with GCK had normal renal function and good CMD.

MRI is an ideal imaging method to identify GCK, because T2-weighted imaging is highly sensitive to cystic lesions and T1-weighted imaging reflects renal function.[3,5,9–11,13] Indeed, its diagnosis was made easily by 2 investigators, because T2-weighted MRI showed numerous, uniformly small cysts located in the renal cortex, which may reflect cystic dilatation of Bowman capsule.[1,3,5] In addition, the assessment of CMD by T1-weighted imaging was concordant between them in almost all kidneys.

In this study, 6 patients with GCK and normal renal function showed good CMD, and 4 of the other 5 patients with GCK and good CMD had eGFR $> 45\,ml/min/1.73\,m^2$. Taking into account some variability of eGFR in the same subjects, their good CMD indicates normal renal function despite cystic dilatation of Bowman capsule. Therefore, the presence of GCK alone may not

Table 1

Magnetic resonance imaging parameters.

Sequences	Tesla	Repetition time (ms)	Echo time (ms)	Flip angle (°)	In-plane resolution (mm^2)	Thickness (mm)	Gap (mm)	reduction factor
T1-weighted imaging	1.5	218.6	4.6	75	1.47 × 1.86	5	1	1.6
(gradient-echo)	3	220	2.3	60	1.47 × 1.76	5	1	2
T2-weighted imaging	1.5	Infinite	80	90	1.47 × 1.28	4	1	1.5
(HASTE)	3	Infinite	90	90	1.25 × 1.87	4	1	3

HASTE = half-Fourier single-shot turbo spin-echo.

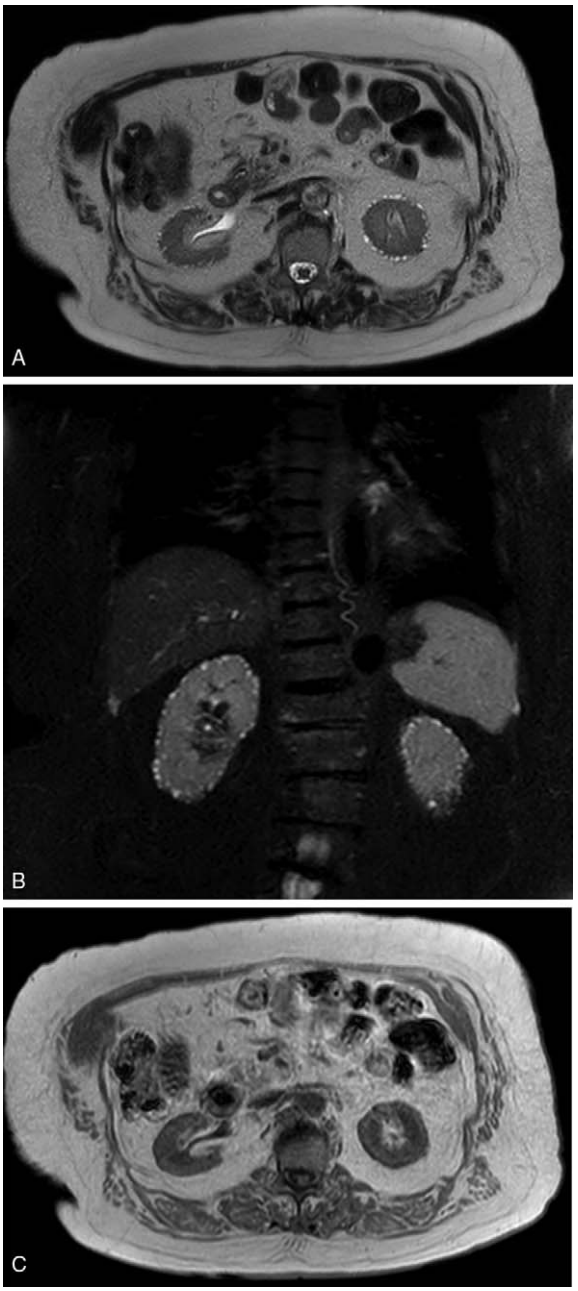

Figure 1. An 84-year-old woman with glomerulocystic kidney and liver cirrhosis. Transverse T2-weighted (A) and coronal fat-suppressed T2-weighted imaging (B) show numerous, uniformly small cysts in the renal cortex. The magnetic resonance imaging features are consistent with glomerulocystic kidney. Transverse T1-weighted imaging does not show any cysts but shows moderate corticomedullary differentiation (CMD) of both kidneys (C). Her estimated glomerular filtration rate (eGFR) is 17.1 ml/min/1.73 m^2. The CMD is likely preserved because of her liver cirrhosis despite the reduced eGFR.

imply renal impairment in older patients without a family history of genetic renal diseases or a past history of renal parenchymal diseases, lithium use, or neuromuscular degenerations.

In contrast, 6 of the 8 patients with GCK and chronic liver diseases had renal impairment, although 11 of their 12 kidneys showed good or moderate CMD. Lee et al[14] have reported that CMD is increased in patients with LC and normal renal function. Yamada et al[13] have shown that in patients with LC, poor CMD reflects renal function, but good and moderate CMD do not.

Table 2

Clinical backgrounds and renal function of 16 older patients with glomerulocystic kidney.

Age sex	Disease	eGFR ml/min/1.73 m	CMD
84 W	LC	17.1	Moderate
79 M	Chronic hepatitis	42.1	Good
77 W	Chronic hepatitis	44.1	Moderate/poor
77 W	IPMN	45.1	Good
87 M	LC HCC	46.2	Good
79 W	LC	46.5	Moderate
82 M	IPMN	51.4	Poor
80 W	IPMN	56.7	Good
90 W	LC HCC	59.1	Good
82 W	IPMN	59.8	Good
80 M	LC HCC	61.2	Good/moderate
79 W	IPMN	65.3	Good
81 M	LC HCC	68	Good
70 W	Liver hemangioma	71.5	Good
67 W	Pancreatic cancer	79	Good
73 W	IPMN	93.4	Good

CMD = corticomedullary differentiation on the T1-weighted images, eGFR = estimated glomerular filtration rate, HCC = hepatocellular carcinoma, IPMN = intraductal papillary mucinous neoplasm, LC = liver cirrhosis, M = man, W = woman. CMD was identical for both kidneys in all but two patients. Ten patients showed reduced eGFR (<60 ml/min/1.73 m^2). In the 6 patients with glomerulocystic kidney, chronic liver diseases, and renal impairment, 11 of the 12 kidneys showed good or moderate CMD.

Therefore, in older patients with chronic liver diseases, the presence of GCK can indicate renal impairment even when good or moderate CMD is observed.[13,15]

This study had some limitations. First, it included only a small sample and was observational because of the rarity and descriptive definition of GCK. However, this study included the largest reported population of individuals with GCK who underwent MRI. Second, the CMD observed on the T1-weighted images was not evaluated quantitatively, because in routine clinical imaging we used a phase-array coil and 2 types of magnetic field strength.[16,17] Although the cysts of GCK were difficult to identify by T1-weighted imaging, we could not entirely exclude that GCK could affect CMD. Lastly, the causes and natural histories of GCK in older patients remain unknown, although aging and renal impairment can be related to GCK.[5,6,10,11] Our study population of patients with GCK included several patients with HCC, LC or IPMN, possibly because these diseases are the main candidates for abdominal MRI and our cohort is representative of the routine studies in the clinical settings. Further studies will be necessary to determine the relationship between GCK and aging, hepatorenal syndrome, and renovascular diseases.[5,8,15]

In conclusion, GCK can be identified in older patients by routine abdominal T2-weighted imaging. GCK is not always associated with renal impairment, but some of older patients with GCK identified by MRI have renal impairment, among those with chronic liver diseases who have preserved CMD on T1-weighted images.

Author contributions

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