



Multiple unilateral subcapsular cortical hemorrhagic cystic disease of the kidney: CT and MRI findings and clinical characteristic

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Abstract

Purpose The aim of this study was to clarify the radiologic and clinical characteristics of multiple unilateral subcapsular cortical hemorrhagic cystic disease of the kidney.

Method Fourteen patients with unique and characteristic multiple hemorrhagic subcapsular cortical cysts of the kidney, not categorized in any existing renal cystic diseases, were retrospectively reviewed. The clinical information including age, sex, symptom, family history of renal or renal cystic disease, and laboratory data were collected. CT and MRI findings including distribution, number and size of cysts, and CT attenuation and signal intensity on T1- and T2-weighted MRI of cysts were analyzed.

Results All patients except one were young and none had a family history of renal or renal cystic disease. Common clinical symptoms were flank or abdominal pain and hematuria. In all cases, only the left kidney was involved at initial presentation. Cysts were small (median cyst size, 4–15 mm), numerous, and distributed mainly along the subcapsular cortex of the kidney. Cysts were hyper-attenuated on unenhanced CT, extremely hypointense on T2-weighted MRI, and mildly hyperintense on T1-weighted MRI. All patients except one had normal renal function. Imaging follow-up revealed stable or mildly progressive disease in seven patients. Two patients developed several hemorrhagic subcapsular cortical cysts in the right kidney at follow-up. Three of five patients with a renal pathology specimen showed concurrent IgA nephropathy.

Conclusion We have identified a unique renal cystic disease with multiple unilateral subcapsular cortical hemorrhagic cystic disease of the kidney that has a characteristic manifestation both radiologically and clinically.

Key Points

- Multiple unilateral subcapsular cortical hemorrhagic cystic disease of the kidney is a unique non-familial renal cystic disease with a characteristic manifestation both radiologically and clinically.
- Most cases of multiple unilateral subcapsular cortical hemorrhagic cystic disease of the kidney are stable or slowly progressive, and do not require invasive intervention.

Keywords Kidney cortex · Cysts · Multidetector computed tomography · Magnetic resonance imaging

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Abbreviations

ADPKD	Autosomal dominant polycystic kidney disease
CT	Computed tomography
MRI	Magnetic resonance imaging
MUCH cystic disease	Multiple unilateral subcapsular cortical hemorrhagic cystic disease

Introduction

Cystic diseases of the kidney comprise a wide range of disease entities, with broad categories being autosomal dominant polycystic kidney disease (ADPKD), autosomal recessive polycystic kidney disease, multicystic dysplastic kidney, autosomal dominant tubulointerstitial kidney disease, glomerulocystic disease, acquired renal cystic disease, and hereditary kidney cancer syndromes such as von Hippel-Lindau syndrome, tuberous sclerosis complex, and hereditary leiomyomatosis and renal cell cancer [1–10]. The renal cystic kidney diseases most commonly diagnosed in adults are ADPKD and acquired renal cystic disease. While renal cystic disease commonly involves both kidneys (with the exception of multicystic dysplastic kidney), unilateral multiple cystic lesions have been reported and described as unilateral polycystic kidney disease, localized cystic disease of the kidney, or unilateral renal cystic disease [11, 12]. Previous reports suggest unilateral renal cystic disease most often demonstrates imaging features similar to ADPKD except that it involves a single kidney or a segmental area of a kidney [13, 14]. Hemorrhagic changes within the cysts in ADPKD are not uncommon, but it is rare to involve almost the totality of cysts. We recently encountered 14 patients who presented with multiple unilateral subcapsular cortical hemorrhagic (MUCH) cysts of the kidney. These patients do not share imaging findings with known existing renal cystic diseases. The purpose of this study is to report the radiological and clinical characteristics of MUCH cystic disease of the kidney.

Material and methods

This study had institutional review board approval. Informed consent was obtained from patients who had genetic testing. Informed consent was waived in other patients. Fourteen patients having unique imaging features on CT or MRI were retrospectively collected through a polycystic kidney disease image conference database at our institutions between June 2007 and January 2018. The unique image features included multiple subcapsular cortical cysts predominantly involving a single kidney and

the majority of cysts having imaging evidence of hemorrhagic contents (hyper-attenuated on unenhanced CT or extremely hypointense on T2-weighted MR images). Eleven patients were from Mayo Clinic (USA) and three patients were referred to the image conference from three different universities (Kanazawa University (Japan), Paris Descartes University (France), and Laval University (Canada)).

Medical records were reviewed and clinical data including age, sex, symptoms, presence or absence of hypertension, and family history of renal or renal cystic disease were recorded. Laboratory findings including serum creatinine levels were recorded. Six patients underwent genetic evaluation. Genetic analysis involved Sanger sequencing of the coding regions of specific PKD genes including *PKD1*, *PKD2*, and in some cases *HNF1B* and *UMOD*, and/or analysis using a next-generation sequencing panel of 65 known or candidate PKD genes in two cases. In one patient, a DNA sample was obtained from the involved kidney.

CT was available in all patients (unenhanced CT only in four patients, contrast-enhanced CT only in one patient, unenhanced and contrast-enhanced CT in nine patients) and MRI in ten patients (unenhanced MR only in two patients, unenhanced and enhanced MR in eight patients). MR images included fast spin-echo T2-weighted image with fat suppression ($n = 10$), two-dimensional or three-dimensional T1-weighted gradient-echo in-phase and opposed-phase images ($n = 10$), and three-dimensional spoiled gradient-echo T1-weighted image with fat suppression before and after gadolinium contrast enhancement ($n = 8$). In one patient, multi-echo gradient-echo images were available for R2* quantification. Images were reviewed for laterality, distribution (subcapsular cortical, cortical, or medullary), number (< 10 , 10 to 20, or > 20) and size of the cyst, and CT and MR signal characteristics of the cysts (CT attenuation on unenhanced CT, T1 signal, T2 signal, signal change in in-phase and opposed-phase images, R2*). For CT attenuation, the region of interest was placed on three to five representative cysts and the mean CT value was calculated. For signal change in in-phase and opposed-phase images, the region of interest was placed on three cysts and the mean signal drop between opposed-phase and in-phase images was calculated as follows: $(\text{Signal}_{\text{opposed-phase}} - \text{Signal}_{\text{in-phase}}) / \text{Signal}_{\text{opposed-phase}}$. The volume of the kidneys was measured using Aquarius 3D Workstation (TeraRecon). The volume of the kidneys was also measured in 28 age- and sex-matched controls (who underwent CT for potential renal donor evaluation). For very young patients (< 25 years old) lacking the age-matched control, the youngest sex-matched controls from the potential renal donor cohort were selected.

Pathological findings were reviewed in three patients. Follow-up was available in seven patients.

Results

Clinical findings at presentation are summarized in Table 1. The mean age of the patients was 32 years old (range, 13–66 years). Nine patients were female and five were male. The most common clinical symptom was flank or abdominal pain ($n = 6$) followed by hematuria ($n = 2$). Six patients were asymptomatic. None of them had hypertension or a family history of renal or renal cystic diseases. Renal function was normal in 12 of 13 patients, and one patient (case 1) had mild renal insufficiency but also had coexisting immunoglobulin A (IgA) nephropathy and interstitial nephritis. Three patients (cases 1, 2, and 10) were diagnosed with IgA nephropathy. In one patient (case 1), IgA nephropathy was diagnosed concurrently with the renal cystic disease, but persistent proteinuria and progressive renal insufficiency preceded it by over 10 years. In two patients (cases 2 and 10), IgA nephropathy was diagnosed 18 and 5 years prior to the diagnosis of the renal cystic disease. Two patients (cases 5 and 9) had splenomegaly due to portal vein thrombosis as a neonate or myelofibrosis, respectively.

DNA samples were available in six patients (cases 1, 2, 4, 7, 8, and 12) but the genetic analysis did not identify any likely pathogenic variants.

Radiological findings are summarized in Table 2. In all patients, the hemorrhagic cysts were present only in the left kidney at initial presentation, the majority in the subcapsular cortex. The cysts were small and relatively uniform in size with the median cyst diameter ranging from 4 to 15 mm with the largest cyst diameter ranging from 6 to 34 mm (Figs. 1, 2, 3, and 4). Ten patients had more than 20 cysts, three had 10–20 cysts, and one patient had 7 cysts. In two patients (cases 1 and 4), nearly the entire cortical surface of the left kidney was

covered by the cysts. CT attenuation of cysts was hyperattenuated compared with the adjacent normal-appearing renal parenchyma on unenhanced CT (13/13) with the mean CT attenuation ranging from 72 to 111 HU across the patients. The cysts showed extremely low T2 signal intensity (10/10) and mildly high T1 signal intensity (8/8) on three-dimensional spoiled gradient-echo with fat suppression. Signal drop of over 15% was evident between opposed-phase and in-phase images in all cases (8/8) when measurable. In two patients, cysts were too small to measure the signal change. On multi-echo fast GRE images, the signal of cysts decreased with increasing echo time and the R2* value of the cyst was 88.5 s^{-1} ($n = 1$), consistent with increased iron concentration [15]. The left and right kidney volumes of patients and controls are shown in Fig. 5. The affected left kidney was larger by 50 cm^3 in four patients and smaller by 50 cm^3 in two patients compared with the right kidney. The threshold of 50 cm^3 was determined from the standard deviation of the left to right kidney volume difference in the controls (16 cm^3). No calcifications, septations, or solid components were seen in any of the hemorrhagic cysts.

In one patient (case 4), papillary renal cell carcinoma type I coexisted in the left kidney. No extrarenal manifestation of hereditary kidney cancer syndrome was present in this patient. Seven patients had additional simple renal cysts. In one patient (case 12), multiple tiny hepatic cysts were found.

Two patients had surgery of the affected left kidney (cases 1 and 4) and one patient had biopsy of the cyst (case 3). One patient (case 4) underwent partial nephrectomy for coexisting papillary renal cell carcinoma with needle biopsy of cysts at the time of surgery. The other patient (case 1) underwent radical nephrectomy for intractable flank pain and infection. Grossly, the subcapsular cortical cysts were brown to black

Table 1 Clinical information of 14 patients with unilateral multiple hemorrhagic subcapsular cortical cysts of the kidney

Case	Age (years)	Sex	Clinical symptom	Creatinine (mg/dL)	IgA nephropathy
1	35	Female	Flank pain	1.4	Yes
2	37	Male	None	1.0	Yes
3	32	Male	None	0.8	No
4	21	Female	None	0.8	No
5	28	Male	None	1.1	NA
6	41	Female	Flank pain, hematuria	NA	NA
7	29	Female	Flank pain	0.9	NA
8	26	Female	Proteinuria	0.7	NA
9	66	Female	None	0.7	NA
10	13	Female	None	0.6	Yes
11	34	Male	Flank pain	1.0	NA
12	41	Female	Gross hematuria	0.8	NA
13	25	Female	Abdominal pain	0.9	NA
14	22	Male	Flank pain	1.0	NA

NA not available or not assessed

Table 2 Radiological findings of 14 patients with unilateral multiple hemorrhagic subcapsular cortical cysts of the kidney

Case	Modalities	Affected side	Number of cysts	Largest cyst diameter (mm)	Median cyst diameter (mm)	CT attenuation	T2 signal on MR images	T1 signal on MR images	Signal drop on long TE MR images
1	CT and MRI	Left ^a	>20	34	15	High	Low	High	Yes
2	CT and MRI	Left	>20	23	10	High	Low	High	Yes
3	CT and MRI	Left	>20	20	12	High	Low	High	Yes
4	CT and MRI	Left ^a	>20	15	6	High	Low	High	Yes
5	CT	Left	>20	15	5	High	NA	NA	NA
6	CT	Left	>20	13	10	High	NA	NA	NA
7	CT and MRI	Left	>20	11	7	High	Low	High	Yes
8	CT and MRI	Left	>20	11	6	High	Low	High	Yes
9	CT and MRI	Left	>20	7	5	High	Low	NA	NM
10	CT and MRI	Left	>20	6	4	High	Low	High	NM
11	CT and MRI	Left	19	13	10	High	Low	High	Yes
12	CT	Left	13	7	6	High	NA	NA	NA
13	CT and MRI	Left	12	18	8	NA	Low	NA	Yes
14	CT	Left	7	11	7	High	NA	NA	NA

NA not available, NM not measured due to small size, TE echo time

^a These patients developed right renal lesions during follow-up

in color (2/2) and contained brown colloidal material (case 1), suggesting hemorrhagic changes (Fig. 3f). Cyst walls were covered by flat or columnar epithelial cells (3/3) (Figs. 1c and 2d). Bowman's space dilatation was absent (3/3). In one patient (case 1), findings of IgA nephropathy and interstitial nephritis were also present in the kidney, but such findings were absent in other patients (cases 3 and 4).

Follow-up imaging was available in seven patients. In four patients (cases 4, 7, 11, and 14), renal cysts were stable at follow-up imaging up to 7 years, while cysts increased in size and number during the 5-year follow-up in one patient (case 10). In one patient (case 1), renal failure developed after nephrectomy requiring dialysis. In two patients (cases 1 and 4), several subcapsular cortical hemorrhagic cysts developed in the right kidney without symptoms during the follow-up after left nephrectomy or partial nephrectomy.

Discussion

The 14 cases we described here are different from previous known renal cystic disease and have unique clinical and radiological characteristics. Clinically, all patients except one were young and, interestingly, affected only the left kidney at initial presentation. More than half of the patients presented with flank pain or hematuria. None of the patients had renal impairment except in one patient who also had IgA nephropathy and interstitial nephritis.

All cysts exhibited imaging findings suggestive of chronic hemorrhage including increased attenuation on unenhanced

CT, extremely low signal on T2-weighted MR images, mildly high signal on T1-weighted MR images, and signal drop in long echo time MR images. Grossly, cyst fluid was brown to black in color.

The volume of left and right kidneys was discrepant in six of 14 patients. In four patients, the affected left kidney was larger than the right, and this can be attributed to the volume of numerous cysts. In two patients, the affected left kidney was smaller than the right. Both of them had a small number of cysts, and renal parenchymal loss or possibly fibrosis was likely the cause of the volume discrepancy. Repeated hemorrhage into the cysts or infection may be the cause of the renal parenchymal loss.

Given the young onset of the disease, a genetic or developmental abnormality may play a role in development of the cysts. All patients presented with cysts in the left kidney, but two patients later developed cysts in the right kidney during follow-up after left nephrectomy, suggesting the presence of mechanical or other factors that promote progression of cysts more rapidly on the left side. The left renal vein is prone to obstruction due to compression by the superior mesenteric artery, but none of these patients had venous obstruction by imaging. Most unilateral congenital anomalies of the kidney and urinary tract are reported to affect the left kidney more frequently than the right, although the exact mechanism of lateralization remains unclear [16].

Various cystic diseases of the kidney such as ADPKD, ARPKD, autosomal dominant tubulointerstitial kidney disease, and acquired renal cystic disease most commonly involve both kidneys and cysts are commonly distributed throughout the renal parenchyma. The imaging findings of

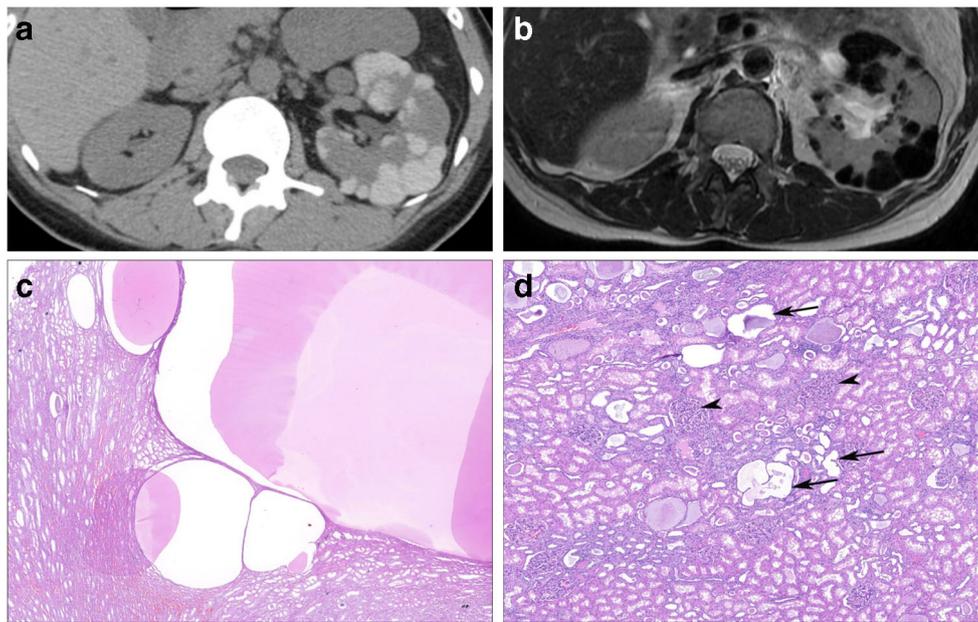


Fig. 1 A 35-year-old female with multiple hemorrhagic subcapsular cortical cysts in the left kidney (case 1). **a** Unenhanced axial CT shows numerous hyper-attenuated cysts along the cortical surface of the left kidney. **b** Cysts are extremely hypointense on axial T2-weighted image (fast spin-echo). **c** Light microscopy shows large

cystic structures distinct from glomeruli at low magnification (upper panel, hematoxylin and eosin, original magnification $\times 2$). **d** There are also variably sized small cortical cystic structures (arrows). These cysts are distinct from glomeruli (arrowheads) (lower panel, hematoxylin and eosin, original magnification $\times 5$)

MUCH cystic disease of the kidney are different from known cystic diseases of kidney.

There are two common renal cystic diseases that involve only one kidney. One entity described as various terms including unilateral polycystic kidney disease, localized cystic disease of the kidney, or unilateral renal cystic disease has

imaging features similar to ADPKD, except it involves only one kidney or a segment of one kidney [13, 14, 17–20]. The renal cysts have variable sizes and the distribution is somewhat random and not localized to the renal subcapsular cortex. Hemorrhage into the cysts is not uncommon, but usually involves only a small portion of cysts. It is more common in

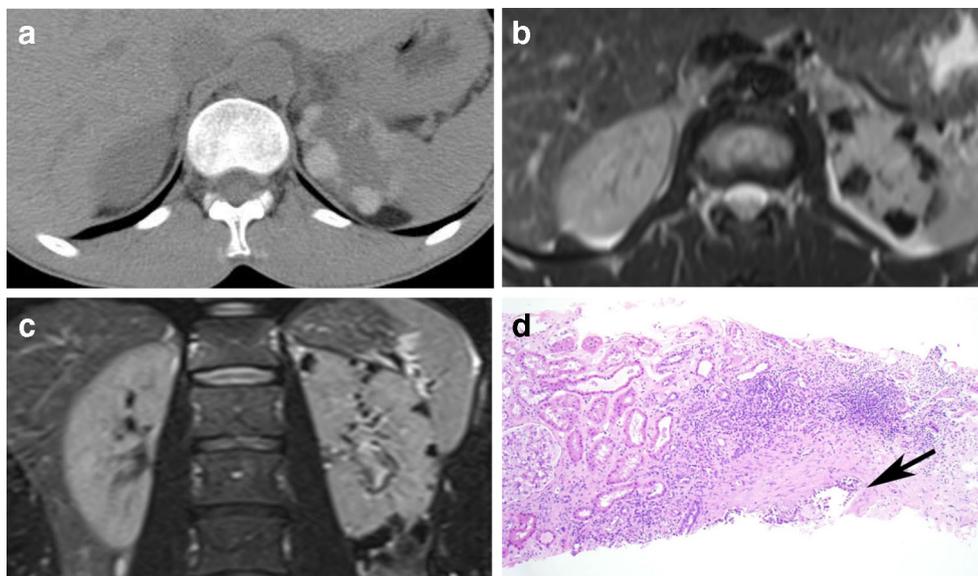


Fig. 2 A 32-year-old male with multiple hemorrhagic subcapsular cortical cysts in the left kidney (case 3). **a** Unenhanced axial CT shows numerous hyper-attenuated cysts along the cortical surface of the left kidney. Cysts are extremely hypointense on axial (**b**) and coronal (**c**)

T2-weighted image (fast spin-echo). **d** Biopsy of cyst lining shows unremarkable renal parenchyma with fibrous cyst wall with surrounding inflammation and bland epithelial cell lining (arrow) (hematoxylin and eosin, original magnification $\times 10$)

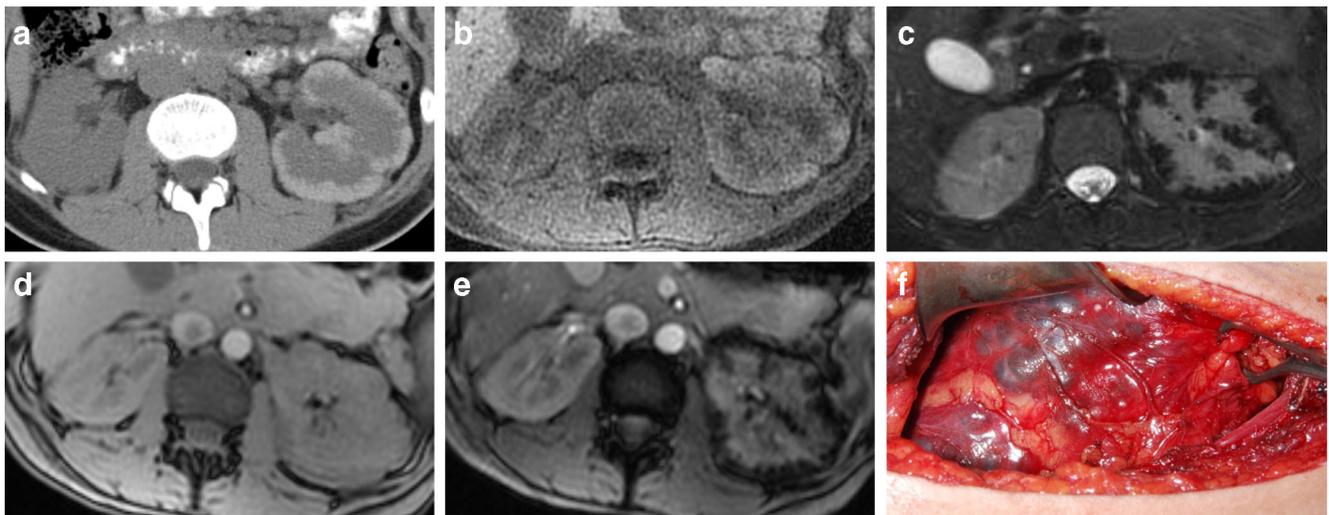


Fig. 3 A 21-year-old female with multiple hemorrhagic subcapsular cortical cysts in the left kidney (case 4). **a** Unenhanced axial CT shows numerous hyper-attenuated cysts along cortical surface of left kidney. Cysts are slightly hyperintense on axial T1-weighted image (**b**, 3-dimensional spoiled gradient-echo), are extremely hypointense on axial T2-weighted image (**c**, fast spin-echo), and shows signal drop between

axial short and long echo time gradient-echo images (**d**, **e**, echo time, 2.4 and 21.5 ms, respectively). **f** During partial nephrectomy for coexisting papillary renal cell carcinoma (not shown), intraoperative photograph shows the left renal surface to be covered by multiple dark subcapsular cysts

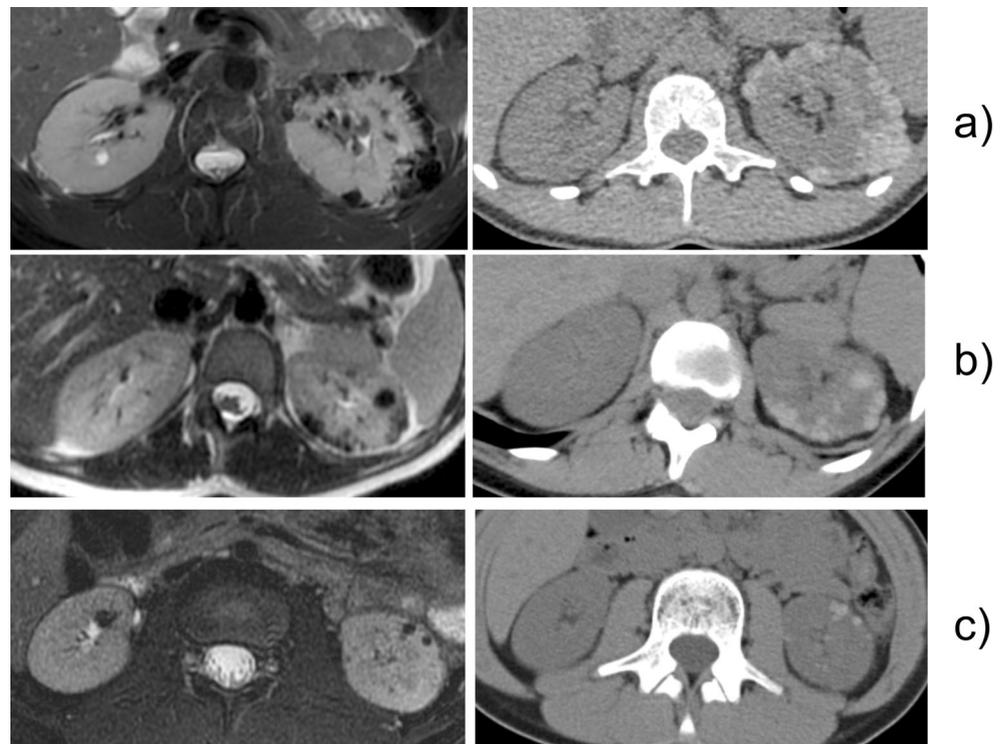
middle-aged males and the patients are evenly divided as to the side of involvement. The mechanism of unilateral cystogenesis has not yet been clarified; some cases have been proven to result from de novo somatic mosaicism of mutations in *PKD1* or *PKD2* [21].

Multicystic dysplastic kidney disease is another cause of unilateral renal cystic disease. It is a developmental renal

dysplasia that causes multiple cysts in the kidney. Some patients may present in adulthood with a multiloculated cystic mass with a central region of solid tissue that contains cartilage, undifferentiated mesenchyme, immature glomeruli, and primitive tubules [22–25]. The affected kidney is nonfunctioning.

Glomerulocystic kidney disease is a rare renal cystic disease that is characterized by cystic dilatation of

Fig. 4 Three different patients with multiple hemorrhagic subcapsular cortical cysts in the left kidney (**a**, case 2; **b**, case 8; **c**, case 10). Cysts show extremely low-signal intensity on axial T2-weighted images (left panels) and high attenuation on unenhanced axial CT (right panels)



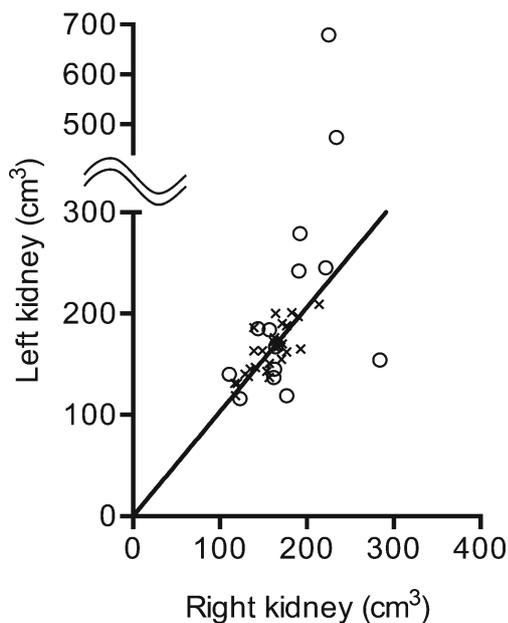


Fig. 5 Scatter plot of kidney volume in 14 patients with hemorrhagic subcapsular cortical cysts (white circle) and 28 age- and sex-matched healthy controls (letter x)

Bowman's capsule. Numerous tiny cysts are located throughout the cortex of kidneys, but not confined to the subcapsular region [26–29]. The glomerular cysts are usually microscopic and only seen on histology. Glomerular cysts have not been described as hemorrhagic. A single case of unilateral glomerulocystic kidney disease in an infant with tuberous sclerosis complex has been described in the literature [30].

The high prevalence of IgA nephropathy (3/14) may suggest association of IgA nephropathy and development of MUCH cystic disease of the kidney. To our knowledge, there have been no reports of the association of IgA nephropathy and renal cystic disease. As renal biopsy did not show findings of IgA nephropathy in two patients, the presence of IgA nephropathy does not appear to be a prerequisite for the development of MUCH cystic disease of the kidney. Two cases had severe splenomegaly; however, the cause of splenomegaly was different.

Our study had several limitations, including the retrospective design and small number of patients included. Many patients did not have pathological confirmation.

The cases presented in this article do not share the CT/MRI findings with known existing renal cystic diseases. We believe this represents a new non-inherited renal cystic disease having a characteristic manifestation both radiologically and clinically. MUCH cystic disease of the kidney demonstrated multiple, tiny, and uniform cysts along the renal subcapsular cortex only in the left kidney at initial presentation, and cysts were hyperattenuated on unenhanced CT and extremely hypointense on T2-weighted MR images.

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Compliance with ethical standards

Guarantor The scientific guarantor of this publication is Naoki Takahashi.

Conflict of interest The authors of this manuscript declare no relationships with any companies, whose products or services may be related to the subject matter of the article.

Statistics and biometry No complex statistical methods were necessary for this paper.

Informed consent Written informed consent was waived by the Institutional Review Board.

Ethical approval Institutional Review Board approval was obtained.

Study subjects or cohorts overlap Any study subjects have not been previously reported.

Methodology

- retrospective
- observational
- multicenter study

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