

Unilateral renal cystic disease: report of an additional case of a rare disease

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ABSTRACT

Unilateral renal cystic disease is a rare condition characterised by a unilateral enlarged kidney, filled with multiple-sized, well-margined cysts separated by parenchymal bands. Only a few cases have been reported in the literature. It must be distinguished from other renal cystic diseases with which it shares radiological features. The authors describe a case of a 26-year-old woman diagnosed with unilateral renal cystic disease, followed-up for ten years in a Nephrology outpatient clinic.

Key-Words:

Autosomal dominant polycystic kidney disease; multilocular cystic nephroma; renal cystic diseases; unilateral renal cystic disease.

INTRODUCTION

Unilateral renal cystic disease is a rare and benign condition, first described in 1964 as unilateral polycystic renal disease¹. Worldwide, only 54 cases of unilateral renal cystic disease have been described until 2005². The disease is non-familial, non-progressive³ and has been known by a number of different names, including segmental cystic disease, localised cystic disease, multiple unilateral renal cysts, segmental polycystic kidney disease, and unilateral polycystic renal disease⁴. Unilateral renal cystic disease should be distinguished from other cystic entities such as

autosomal dominant polycystic kidney disease (ADPKD), multilocular cystic renal nephroma, cystic dysplasia and multiple simple cysts.

We present a case of a young woman with unilateral renal cystic disease in the right kidney.

CASE REPORT

A 26-year-old Caucasian woman presented to her doctor in 1999 complaining of chronic right flank pain for several months. She denied gastrointestinal, respiratory, cardiovascular or urological symptoms. No family illnesses, including renal diseases, were reported. When examined, she was hypertensive with a blood pressure of 160/100 mmHg, but otherwise the examination was unremarkable. Laboratory findings were unremarkable, namely, renal function (serum creatinine and urea 0.8 mg/dL and 24 mg/dL, respectively, and creatinine clearance 135 mL/min) and urinalysis. A renal ultrasound revealed multiple cysts of varying sizes in the right kidney and a normal left kidney. Computed tomography (CT) scan revealed a diffusely enlarged right kidney measuring 175 mm pole to pole and containing multiple cysts of varying sizes. There were no cysts in the left kidney or in other intra-abdominal organs.

Over the next 10 years of follow-up in our Nephrology outpatient clinic, she maintained a normal renal function (serum creatinine level 0.6 mg/dL). A renal

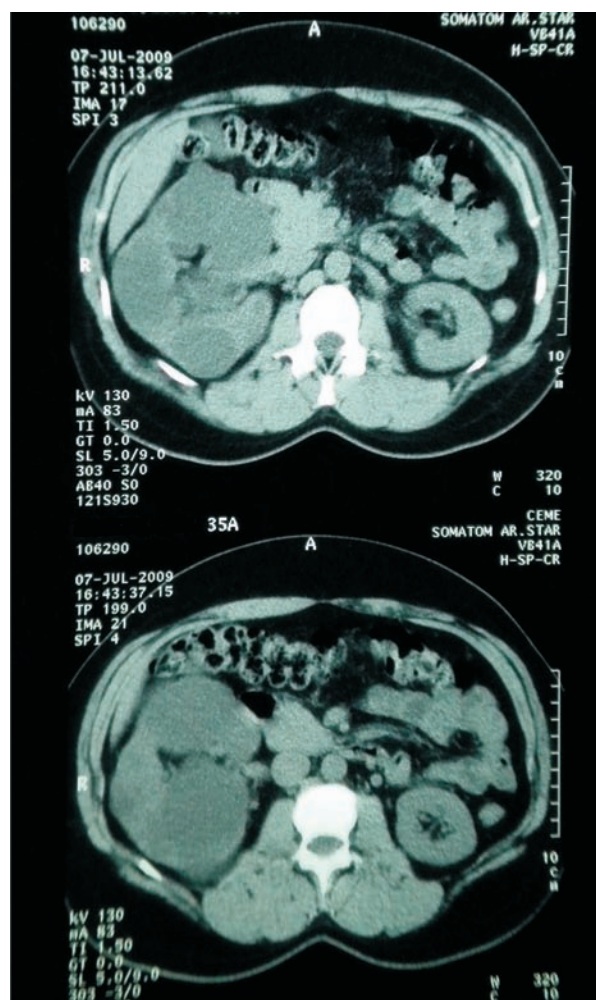


Figure 1

The 2009 abdominal CT shows an enlarged right kidney filled with varying sized round, well-marginated multiple cysts with no capsule formation. The cysts are separated by parenchymal bands (no oral or IV contrast was used)

ultrasound and an abdominal CT scan recently performed showed the same cystic lesions in the right kidney, with no evidence of cysts in other locations (Fig. 1). Her parents and daughter underwent renal ultrasounds that were unremarkable.

DISCUSSION

Unilateral renal cystic disease (URCD) is a rare and poorly understood condition, first described in 1964 as unilateral polycystic renal disease. Until 2005,

only 54 cases of URCD have been described in the literature². Most of these patients present with one or a combination of the following: hypertension, flank pain, haematuria or a flank mass⁵. URCD should be distinguished from other cystic entities such as ADPKD, multilocular cystic renal nephroma, cystic dysplasia and multiple simple cysts.

The diagnosis is made using imaging techniques such as CT scan, ultrasound or magnetic resonance imaging, but additional information is needed to make the differential diagnosis with these other entities.

The main differential diagnosis is with ADPKD. URCD is similar to ADPKD, however, but can be distinguished from ADPKD by five clinical features: unilateral location, negative family history, no progression to chronic renal failure, no cysts in other intra-abdominal organs, and no disorders affecting other body organ systems⁶. Initially, ADPKD may manifest as the involvement of a single kidney, however, as time goes by, cysts will develop in the other kidney as in other organs and other associated conditions may be present as colonic diverticuli or circle of Willis aneurysms (Berry aneurysms). That means that the initial unilateral disease evolves into an asymmetric bilateral disease in a long-term follow-up period. This asynchronous presentation should be excluded by phenotype screening of family members, since ADPKD is an inherited disease that shows Mendelian transmission. Taking only a family history may not be enough.

In our case, family screening was negative for ADPKD. Furthermore, after 10 years of follow-up, no additional cysts have developed.

There are important implications in distinguishing these two entities. Patients with unilateral renal cystic disease should not be labelled as having a hereditary progressive renal cystic disease such as ADPKD, which may have ramifications for individual insurability, prognosis or the need of genetic counselling.

In the spectrum of other cystic conditions that must be differentiated from URCD is multilocular cystic renal nephroma. However, this condition usually forms discrete, encapsulated masses that are well demarcated from the adjacent renal parenchyma, and does not contain patches of non-cystic renal parenchyma interposed among the cysts, as in URCD⁶.

In unilateral dysplastic cystic kidney, the kidney is usually non-functioning, as the collecting system is usually atretic or obstructed. Therefore, in this condition the collecting system is usually not opacified on contrast-enhanced imaging, whereas in URCD the collecting system shows only a displacement^{6,7}.

Multiple cysts may be difficult to distinguish from URCD when confined to one kidney, but they are less numerous than in URCD, and they are predominantly located in renal cortex whereas in URCD they affect both cortex and medulla⁶.

CONCLUSION

URCD is a rare entity that requires recognition as a benign, nonsurgical condition. It demands periodic follow-up, to avoid unnecessary surgery or wrong labels that have different treatment approaches and prognosis.

Conflict of interest statement. None declared.

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