



Unilateral Renal Cystic Disease: A Case Report of A Rare Disease and Review of Literature

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Abstract

Unilateral renal cystic disease (URCD) is a rare entity with few reported cases. This condition is often misdiagnosed for other cystic renal diseases like autosomal dominant polycystic kidney disease, cystic dysplastic kidney disease and cystic nephroma. Nevertheless, this is a benign entity with a potential for good prognosis. Imaging features, supported by background clinical and biochemical findings, diagnosis of URCD is possible. A case of URCD is reported here, having diagnosed on imaging features and followed up conservatively.

Introduction

Unilateral renal cystic disease (URCD) is a rare cystic renal disorder. It is characterized by affected kidneys having multiple cysts, normal excretory system and a near normal contralateral kidney. It is not related to autosomal dominant polycystic kidney disease (ADPKD) [1,2]. There is no genetic predisposition, renal function deterioration or extra renal manifestations [1,2]. Knowledge of imaging features, with supportive clinical and biochemical evidence, help radiologist to arrive at the correct diagnosis and appropriate follow up. We report a case of URCD diagnosed by computed tomography (CT) and magnetic resonance imaging (MRI) characteristics.

Case Presentation

Seventeen year old female patient was presented with intermittent right sided abdominal pain for 6 months duration. No family history of renal disease was identified. Physical examination was unremarkable. Her blood pressure was 110/70mmHg.

Biochemical investigations, including serum creatinine, full blood count, and urinalysis were normal. Her serum creatinine was 0.6mg/dl.

On abdominal ultrasonography (USS) the right kidney was enlarged with a bipolar length of 12.4cm. There were multiple cysts of varying sizes involving cortex and medulla of the right kidney and some of them having mural calcification. Ultrasonically the left kidney was normal. There were no cysts in the spleen, pancreas or liver.

Technetium-99m-dimercaptosuccinic acid (DMSA study)

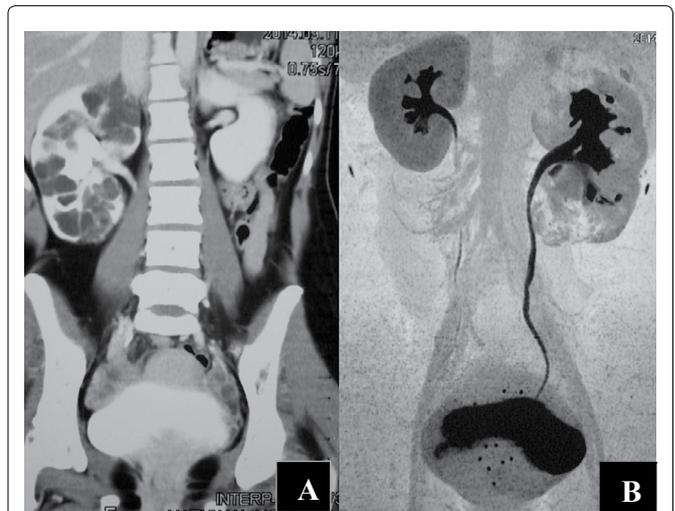


Figure 1: CECT abdomen (A) coronal image demonstrating multiple simple cysts in Right kidney and normal left kidney and (B) CT urography image (posterior) showing normal excretory system of right kidney.

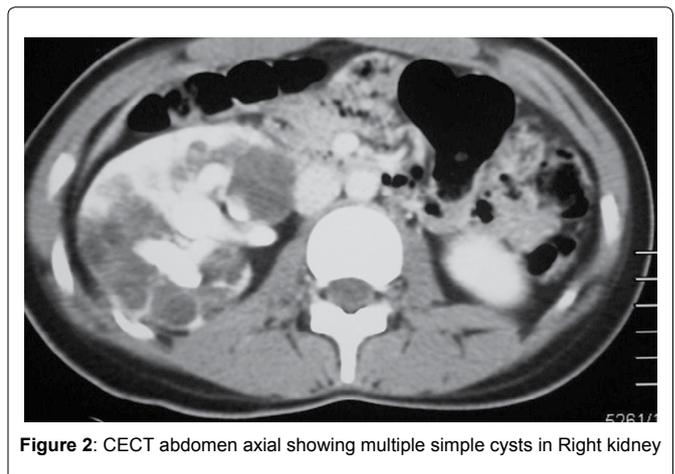


Figure 2: CECT abdomen axial showing multiple simple cysts in Right kidney

showed reduced tracer uptake in the right kidney. Differential renal

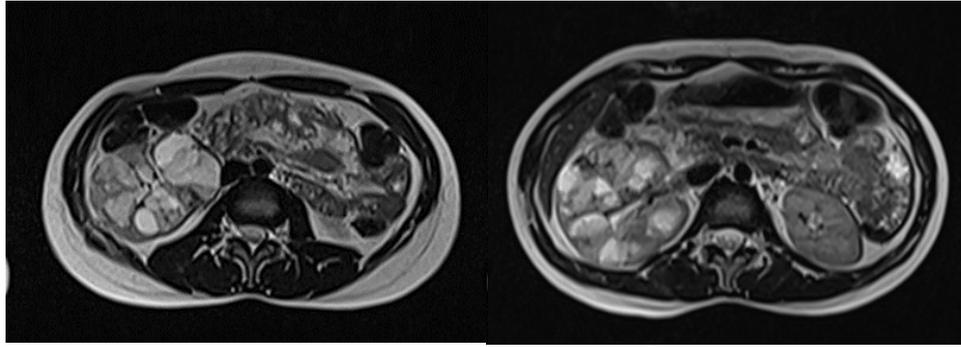


Figure 3: Axial T2 weighted MRI images showing multiple cysts in the right kidney.

Table 1: Cases of URCD reported in the literature with emphasis on radiological imaging features.

Reference	Age/Sex	FH _x	BP	SC	Imaging findings	Opposite kidney /other organs	Follow up
Hazarika et al. 2014 [8]	49/ Male	Negative	NL	NL	CT Diffusely involved with cysts and residual normal renal parenchyma. No enhancing soft tissue component or bands. Normal collecting system.	Not involved	Not mentioned
Miyamoto et al. 2010	52/ Male	Negative	NL	NL	CT Multiple large cysts scattered diffusely.	Not involved	Follow up
Curry N S et al. 1993	57/ Male	Negative	NA	NL	CT A near complete replacement of affected kidney by cysts of varying sizes and calyceal distortion.	2 small, simple cysts in upper pole	CT follow up for 30 years
Bae E H et al. 2013 [9]	51/ Male	Negative	NA	NL	CT Revealed enlarged kidney with variable-sized round, well marginated multiple cysts without capsule formation or solid content. MRI Six months, later showed numerous cysts in the right kidney with no changes according to previous CT	Not involved	Follow up by MRI in 6 months
Park B S et al. 2007 [1]	31/ Female	Negative	NL	NL	CT Enlarged kidney filled with variable sized multiple cysts with enhancing normal renal tissues between the cysts. MRI Enlarged kidney with multiple cysts of varying size without capsule formation. There were no solid portions within the cysts and normally enhancing renal parenchyma seen between the cysts.	Not involved	Laparoscopic Radical nephrectomy
Hwang D Y et al. 1999 [2]	31/ Male	Negative	NL	NL	CT Enlarged kidney, variable size well marginated multiple cysts with no capsule formation. Cysts separate by normal parenchymal bands.	2 simple, cortical cysts. Other organs normal	Follow up with creatinine for 18 months
	44/ Male	Negative	High	NL	Enlarged kidney with variable size multiple cysts separate by enhancing septae	Not involved	Follow up 15 months with Creatinine
Farinha A et al. 2011 [8]	26/ Female	Negative	High	NL	CT Diffusely enlarged kidney containing multiple cysts of varying sizes.	Not involved	Follow up radiologically for 10 years
Zafar Neyaz et al. 2012 [4]	16/ Male	Negative	NL	NL	USS Multiple small, simple cysts with relative sparing of the upper pole parenchyma. MRI Multiple simple cysts mainly involving middle and inferior polar region of the right kidney without any solid component or thick septae.	Not involved	Follow up annually with USS & creatinine
Solack et al. 2013 [6]	16/ Male	Negative	High	NL	USS Multiple variable sized cysts in the upper pole of the kidney CT The upper pole of the right kidney was filled with multiple cysts of different sizes with enhancing normal renal tissue between the cysts. MRI Upper pole cysts without capsule formation. There were no solid areas within and renal parenchyma between the cysts was normally enhancing DMSA Decreased renal uptake at the upper half of the right kidney	Not involved	Right upper pole hemi nephrectomy to control BP 2 year follow up with USS and creatinine

NOTE – FH_x: Family History, BP: Blood Pressure, SC: Serum Creatinine, CT: Computed Tomography, MRI: Magnetic Resonance Imaging, USS: Ultrasonography, DMSA: Technetium-99m-Dimercaptosuccinic Acid, NL: Normal and NA: Not Available

functions were 65% in left kidney and 34% in right kidney.

Contrast enhanced abdominal CT (CECT) scan revealed enlarged right kidney with multiple cysts, of varying sizes without capsular formation. It showed enhanced intervening normal renal parenchyma (Figure 1A) with normal pelvicalyceal system and ureters (Figure 1B). CT scan confirmed that there are no cysts in the contralateral kidney

or other abdominal organs (Figure 2).

MRI scan too confirmed, evidence of enlarged right kidney (BPL 13cm) with multiple renal cysts (Figure 3) with preservation of the normal renal configuration. There were no solid masses or thick septae in cysts.

Based on the clinical, biochemical and imaging findings, and

the absence of family history of renal cystic diseases, the patient was diagnosed as URCD. Follow up with imaging is recommended to evaluate possible complications of renal cysts.

Discussion

Unilateral renal cystic disease is a rare entity, first described in 1964 as unilateral polycystic kidney disease [3]. In 1979 Cho et al., first proposed this condition as a separate entity from ADPKD [4]. Whereas, Levine et al. [5], in 1989 named the condition as unilateral renal cystic disease. There are only 63 reported cases in the literature [4]. Radiological and clinical features of recently reported cases in the literature are summarized in table 1. Clinically, these patients present with hypertension, flank pain, hematuria or a flank mass.

Imaging findings vary depending on the degree of involvement. Cysts can involve only a segment of the kidney or the whole kidney. Solack et al. [6] and Neyaz et al. [4] reported two cases of localized or segmental form of URCD and other reports are of diffused disease as in our case. Ultrasonography displays multiple cysts of various sizes, which may appear complex due to intervening parenchyma, simulating septae of a cystic nephroma. There can also be scattered focal mural calcifications in the cyst wall [4].

Conventional, CT or MR urography demonstrates distortion of the pelvicaliceal system. However, the affected kidney shows normal excretion despite a significant portion of the kidney being replaced by cysts [4]. Hazarika et al. [7] also reported that the excretory system is normal despite pelvicaliceal distortion. CT or MR imaging best reveals the cystic nature of this renal disease [4] and background supportive features to arrive at a correct diagnosis.

URCD need to be distinguished from other cystic renal conditions such as ADPKD, cystic nephroma and cystic dysplastic kidney diseases [8].

Although URCD demonstrate similarity to ADPKD, it is different from ADPKD. As URCD is a non-familial, non-progressive and a unilateral entity with absence of pancreatic, splenic or liver cysts and other organ abnormalities like colonic diverticuli and berry aneurysms in the circle of Willis [7,8]. Similar findings were seen in our case and in most of the cases reported in the literature.

In our patient, family screening using USS was negative for ADPKD and she did not show extra renal manifestations of ADPKD. Most of the cases reported in literature also confirm negative family history normal renal functions. Bae et al. [9] and Hwang et al. [2] as in this case, screening was done using USS whereas, other cases in the literature were reviewed clinically. URCD shows fairly a benign course, compared to ADPKD. Associated renal cell carcinoma has not been reported so far. Due to prognostic and genetic implications, differentiation of these two entities is imperative.

Enhancing and compressed renal tissue in URCD can mimic enhancing septae of a complex cyst in CT. Therefore, in the segmental form of URCD cystic renal cell carcinoma needs to be excluded [9]. Discrete, encapsulated masses in the latter condition would be a distinguishing imaging feature. Non encapsulated nature of cysts and preservation of reniform configuration in URCD is described as a defining radiological feature in Park et al. [1] and Bae et al. [9] as well.

In URCD collecting system is continuous, thus facilitating the differentiation from cystic unilateral dysplastic kidney diseases in which, the affected kidney is non-functional and ureters are atretic [9]

Most patients of URCD are commonly diagnosed in adulthood, although children and even infants and neonates have been reported in the literature.

URCD, due to its benign nature can be managed conservatively, biochemical and radiological surveillance to follow up is sufficient. This was the protocol used in most of the reported cases except, for a few, which offered nephrectomy to control symptoms.

Conclusion

URCD is rare, but is a benign condition with a relatively good prognosis. Imaging together with clinical and biochemical evidence can differentiate it from other similar morphological conditions like ADPKD, cystic nephroma and cystic dysplastic kidney diseases with a relative certainty. Knowledge of this condition helps in arriving at correct diagnosis, thus minimizing unnecessary investigations and patient and family reassurance.

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