

## Case Report

# Unilateral renal cystic disease: a case report study

Shameer Hakkim\*, Babu R., Kishan Raj K., Seetha Rami Reddy Mallampati

Department of Medicine, Vinayaka Missions Medical College and Hospitals, Karaikal, Puducherry, India

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**\*Correspondence:**

Dr. Shameer Hakkim,

E-mail: [shameer606@gmail.com](mailto:shameer606@gmail.com)

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### ABSTRACT

Unilateral renal cystic disease (URCD) of kidney is a non-familial, extremely rare condition, characterized by replacement of the renal parenchyma of one kidney by a cluster of multiple cysts of varying size with a normal contralateral kidney. It is morphologically indistinguishable from autosomal dominant polycystic kidney disease (ADPKD); as such, hepatic and pancreatic cysts is not seen and shows no progressive deterioration in renal function; thus, differentiating ADPKD from URCD becomes important. We report a case of URCD documented by clinical and radiological imaging. A 21 year-old female, presented with history of mild lancinating pain in the left flank for 6 years which aggravated in the past 3 days, with no history of lower urinary tract symptoms. No significant family illnesses reported. Examination showed normal vitals and ballotability present and associated tenderness on deep palpation in left lumbar region. Laboratory findings were within normal limits. Ultrasonography of abdomen and pelvis showed left hydronephrosis with multiple cysts. CECT Abdomen revealed an enlarged left kidney (~15×16×10 cm) filled with variable sized round, well-marginated multiple cysts. Renal ultrasound was performed on patient's parents and her siblings and ruled out cystic renal disease. Hence, authors considered the diagnosis of URCD in this patient. In conclusion, treatment and managing guidelines of URCD have not been mentioned in any of the medical literatures. There is little information regarding the progression of URCD. Hence there is a need for further understanding of pathogenesis, progression and management of these patients.

**Keywords:** ADPKD, Case report, Unilateral renal cystic disease

### INTRODUCTION

Unilateral renal cystic disease (URCD) is a distinct, unusual condition characterized by replacement of all or a portion of one kidney by multiple cysts. Kidney diseases are a group of genetically heterogeneous disorders and leading a cause of kidney failure. Focal renal cysts are typically detected in affected subject before 30 years of age. Hundreds to thousands of cysts are usually present in the kidneys of most patients in the fifth decade. Enlarge kidneys can each reach a fourfold rise in length and weight upto 20 times the normal weight. It may be

associated with hypertension or hematuria. While initially thought to be a form of autosomal dominant polycystic kidney disease (ADPKD), unilateral renal cystic disease is nonfamilial, nonprogressive, and not associated with renal failure or cysts in other organs. Patients with ADPKD have bilateral disease, progression to renal failure, associated cysts in the liver and other organs, and relatives with the disorder. Aside from its striking unilaterality, histologically and radiographically, URCD is indistinguishable from ADPKD. Imaging studies usually show marked renal enlargement and either diffuse or partial replacement of the entire kidney with cysts.<sup>1-4</sup> Here, authors are presenting a case of unilateral renal

cystic disease by an adolescent female presenting with lancinating abdominal pain in the left flank.

## CASE REPORT

A 21-year-old female belong to upper lower socioeconomic status of modified kuppuswamy scale, nil premorbid, presented to medical outpatient department with history of mild, lancinating pain in the left flank for 8 years which aggravated in the past 3 days. She denied any history of hematuria, fever, and lower urinary tract symptoms. There were no other cardiovascular, respiratory, gastrointestinal, urological and neurologic symptoms. No significant family illnesses including renal diseases were reported.

On examination, her blood pressure recorded was 100/70 mmHg and ballotability was present in the left side associated with tenderness on deep palpation in left lumbar region while other system examination was unremarkable. Laboratory findings were also within normal limits, including serum creatinine (1.0/\*\* mg/dl) and urinalysis. Ultrasonography of abdomen and pelvis showed left hydronephrosis measuring 17 cm with multiple cysts. CECT Abdomen (Figure 1 and Figure 2) revealed an enlarged left kidney (~15×16×10 cm) gross dilatation of renal calyces causing thinning of the renal parenchyma and filled with variable sized round, well-margined multiple cysts with no capsule formation. No cysts were detected in other intra-abdominal organs with imaging studies. Epithelial membrane antigen (EMA), vimentin and transducin-like enhancer protein 1 (TLE1) were negative and CD99 showed a normal report.



**Figure 1: Enlarged left kidney with well-margined multiple cysts with no capsule formation.**

Renal ultrasound was performed on patient's parents and her siblings and none of them was observed to have cystic renal disease. Hence, authors considered the

diagnosis of Unilateral renal cystic kidney disease in this patient. Patient has been treated conservatively. Patient symptomatically improved during the hospital stay and she has been discharged. She is been under regular follow up.



**Figure 2: Enlarged left kidney (~15×16×10 cm) with gross dilatation of renal calyces causing thinning of the renal parenchyma and filled with variable sized round.**

## DISCUSSION

In the late 1970s, several studies reported that URCD might be different from ADPKD.<sup>1-3</sup> Levine et al, proposed the term 'unilateral renal cystic disease (URCD)' as a distinct disease entity in 1989.<sup>4</sup> The clinical importance of URCD is to make a differential diagnosis from ADPKD. As this study shows, URCD has at least three aspects different from ADPKD (i) unilateral localization, (ii) negative family history and (iii) no progression to chronic renal failure.

The clinical importance of URCD is to make a differential diagnosis of such abnormalities including multilocular cystic nephroma, cystic partially differentiated Wilms' tumor, segmental cystic dysplasia, and atypical presentation of polycystic kidney disease such as asymmetric evolution and mosaicism.<sup>5,6</sup> The pathogenesis of this cystic renal disease is unknown. Since there is a morphological similarity of this cystic change to autosomal dominant polycystic kidney disease (ADPKD), it is speculated that pathogenesis is similar.<sup>7</sup> The gross and microscopic features are indistinguishable from ADPKD, and patients may present with hematuria, pain, or a flank mass.<sup>8</sup> However, it can be differentiated from ADPKD by its unilateral localization, negative family history, no progression to chronic renal insufficiency, and no extra renal manifestation. Cystic adenocarcinoma of the kidney can present in a very similar way and it can be differentiated by positive reaction to

epithelial membrane antigen (EMA), vimentin and transducin-like enhancer protein 1 (TLE1), and CD99.<sup>9</sup> Most importantly, it must be differentiated against segmental cystic dysplastic disease. In case of URCD, collecting system should be shown continuously though distorted collecting system. Otherwise it would lean to segmental cystic dysplastic disease.<sup>10,11</sup> The collecting system of this patient was continuous

The pathological findings of URCD are not different from those of ADPKD, showing numerous epithelial cell-lined cysts interposed among patches of non-cystic renal parenchyma. However, grossly, those cysts were localized exclusively in one kidney. In earlier studies, the unilaterality was confirmed by examining nephrectomized specimens or autopsy tissues.<sup>12,13</sup> As more imaging techniques improve, it will become easier to confirm the unilateral localization of lesions of URCD. Since CT became popular, the surgical confirmation of URCD is no longer needed if patients have characteristic CT findings in combination with genetic and clinical manifestations.<sup>14</sup>

Unilateral cystic change of the kidney has been reported in children as an unusual presenting manifestation of ADPKD.<sup>15,16</sup> and has recently been reported to occur in autosomal recessive polycystic kidney disease.<sup>16</sup> The distribution of renal cysts in ADPKD has been found to be asymmetric, particularly in children.<sup>15</sup> In many of these cases, however, investigators relied upon excretory urography to detect the condition. This method of examination is relatively insensitive to subtle changes of cystic disease which are more readily identifiable by ultrasonography or CT. In two reported pediatric cases where there was long-term follow-up, initial unilateral disease evolved to asymmetric bilateral disease on reevaluation 7 and 9 years after the original diagnosis. Family history was also positive for ADPKD in these children.<sup>17,18</sup> Therefore, asymmetric evolution of ADPKD should be excluded either by phenotype screening of family members or by a long period of follow-up.

In our case, Patient has been ruled out for other system involvement in the form of mitral valve prolapse or colonic diverticulum. By other radio-imaging techniques the possibility of cystic lesion has been ruled out. Phenotypic analysis of family members using renal ultrasound was performed in our study. Both parents and siblings of our patient showed no cystic diseases. Therefore, it is concluded that URCD is not a hereditary trait.

Only a few cases of URCD have been reported. Among them, hypertension was found in six, flank or abdominal pain in seven, abdominal mass in seven, and gross hematuria in three cases. A renal stone was found in one case and UTI in one case.<sup>19,20</sup> The clinical manifestations of our two cases such as abdominal mass, flank pain and gross hematuria were consistent with other reported cases.

In the literature, several patients who had both possible URCD and neoplastic disease have been reported. Posso et al, and Hayward WG, reported patients with unilateral polycystic kidney and renal cell carcinoma.<sup>21,22</sup> Cole et al, reported a case of URCD associated with dual adenocarcinomas; Wells et al, reported a case with intracystic papilloma.<sup>23,24</sup> Regrettably, except for the case of Cole et al, no study reported the description of family history; the study by Cole et al, reported a suspicious hereditary renal disease.<sup>23</sup> Although the possibility of misdiagnosis of other cystic renal diseases exists in these cases, the possibility of co-existing renal cell carcinoma in URCD should also be considered.

URCD is a stable disease and patients can be followed up by imaging techniques.<sup>8</sup> In conclusion, unilateral cystic disease of kidney is a rare cystic disease of the kidney diagnosed by imaging techniques and requires nephrectomy only when suspicion of malignancy is strong.

## CONCLUSION

Occurrence of unilateral renal cystic kidney disease is very rare and its treatment and managing guidelines have not been mentioned in any of the medical literatures. Though ADPKD is known to cause multi organ involvement and progressive renal failure, there is little information regarding the progression of unilateral renal cystic kidney disease. Hence there is a need for further understanding of pathogenesis, progression and management of these patients with the help of further case series study and researches on this topic.

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