Localized cystic disease of the kidney: a rare entity

Zafar Neyaz[1*], Sunil Kumar[1], Hira Lal[1], Rakesh Kapoor[2]

1. Department of Radiodiagnosis, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India

2. Department of Urology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India

* **Correspondence:** Dr. Zafar Neyaz, MD, Assistant Professor, Department of Radiodiagnosis, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Rae Bareilly Road, Lucknow, Uttar Pradesh, 226014, India
(*zafarneyaz@hotmail.com*)

Radiology Case. 2012 Jul; 6(7):29-35 :: DOI: 10.3941/jrcr.v6i7.1026

ABSTRACT

Localized cystic disease of the kidney is a benign nonsurgical entity and presents with multiple cystic lesions in just one portion of the kidney or involves the entire one kidney. We report a case of localized cystic disease of the kidney in a 16 year-old-male. This patient underwent an ultrasound examination and incidentally found to have multiple cysts in the right kidney whereas the left kidney was normal. Diagnosis was confirmed by typical MRI findings, absence of any family history, normal results of urine analysis and renal function tests.

CASE REPORT

CASE REPORT

A 16-year-old male underwent an ultrasound examination for acute abdominal pain in the periumbilical region. It revealed multiple small cysts in the right kidney with relative sparing of the upper pole (Fig.1A). Right Kidney was mildly enlarged in size. The left kidney was normal in appearance (Fig. 1B). Remaining abdominal organs showed normal appearance. The abdominal pain subsided with conservative management; however, patient was referred to urology department for the evaluation of renal cysts. Past history did not reveal hematuria or any other urological symptoms. Physical examination was normal with arm blood pressure of 110/80 mm Hg. Urine analysis and renal function parameters were within normal limits. There was no history of renal disease in the family.

An abdominal MRI examination was performed on 3-Tesla GE scanner to characterize the renal lesions. Fat suppressed T2 weighted, Diffusion weighted sequence, pre-contrast T1 weighted gradient sequences were carried out in axial plane. Post contrast T1 weighted gradient sequence was acquired in axial and coronal planes. MR imaging revealed multiple simple cysts mainly involving middle and inferior polar region of the right kidney (Fig. 2-5). The upper polar region was relatively spared with normally enhancing renal tissue. The cysts were small in size with thin walls without any solid component or thick septa inside. However, at some places, attenuated renal parenchyma was present wedged between cystic lesions. No diffusion restriction was present inside any cystic lesion (Fig. 6). The left kidney was entirely normal. The right renal vein was normal and there was no retroperitoneal lymphadenopathy. No cystic lesions were seen in liver, pancreas or spleen.

Based on laboratory parameters, sonographic and MRI imaging findings, a diagnosis of localized cystic disease of the kidney was made. Patient has been informed about the benign non-progressive nature of the condition; however, he has been advised to undergo annual ultrasound, urine analysis and renal function test.

DISCUSSION

Localized cystic disease of the kidney is a non hereditary, non-progressive, and benign renal condition [1-3]. This disease is characterized by multiple cysts replacing a variable portion or an entire kidney, while the other kidney remains normal. The cysts are separated by normal or atrophic renal tissue [1]. This is a rare entity with approximately 63 cases reported in the literature [1-23]. Initially localized cystic disease was considered to be a form of autosomal dominant

polycystic kidney disease (ADPKD), but Cho et al in 1979 first suggested this entity to be distinct from ADPKD [4]. Levine et al proposed the term "unilateral renal cystic disease" for this condition; however, the term "localized cystic disease of the kidney" was preferred by Slywotzky et al [1, 5].

Exact etiology and pathogenesis of this condition is unknown [1]. Most of the affected patients are males in the fifth decade; however, some cases have been reported in the younger age group [1, 4-6]. Common clinical presentation of this condition includes hematuria, hypertension, abdominal mass, and flank pain [1-3]. In a few cases proteinuria, mild azotemia, renal stone formation and urinary tract infection have also been described [2-8]. Pathological examination reveals multiple smooth walled cysts containing clear yellow fluid with regions of attenuated normal or atrophic renal tissue separating these cysts [1, 4, 14]. Microscopic examination shows thin walled cysts of various sizes lined by flattened cuboidal epithelium.

Imaging findings depend on the extent of involvement [1]. Cysts may involve only a portion of the kidney or the entire kidney. Scattered focal calcifications may be noted in the cyst wall [1, 13]. The contralateral kidney remains entirely normal. In some cases few small cysts have been reported in uninvolved kidney but they were thought to represent simple cysts, a common finding in adults [1]. Intravenous urography (IVU) shows enlarged kidney with multiple masses distorting the pelvicaliceal system. The affected kidney shows normal excretion of the contrast medium, even in patients in whom a large portion of the kidney is replaced by the cysts [1]. Ultrasound shows multiple cysts of variable size, which may appear complex due to the presence of intervening attenuated parenchyma simulating septae. CT or MR imaging best reveals the imaging characteristics of localized cystic disease [1-3, 5].

The major differential diagnosis of localized cystic disease in an adult patient are autosomal dominant polycystic kidney disease, multicystic dysplastic kidney, multiple simple cysts, multilocular cystic nephroma, and cystic renal cell carcinoma.

Although localized cystic disease morphologically resembles ADPKD, it is a unilateral condition without any genetic background [3]. In addition, ADPKD is associated with extra renal manifestations which include cysts in other organs (liver, pancreas, and spleen), valvular disorder, cerebral aneurysm and colonic diverticula. Deterioration of the renal function, hemorrhage and renal calculi are common in ADPKD, but usually not seen in localized cystic disease [10]. However, distinction may be difficult when ADPKD has unilateral presentation, especially in children [11]. In children with unilateral diffuse renal cysts with functioning kidney, imaging should be performed in the parents to rule out ADPKD. In such cases, long term follow up is also useful as there may be progression of unilateral into asymmetric bilateral disease [2].

When localized cystic disease involves a small segment of kidney, it may be confused with focal cystic masses such as multilocular cystic nephroma and cystic neoplasm. However, localized cystic disease does not show encapsulation and often shows other cysts clearly separate from the main cluster of cysts [1]. Cystic renal masses are usually discrete, encapsulated and do not contain enhancing renal parenchyma inside. Evaluation of contiguous axial CT images and multiplanar reconstruction are particularly useful in these situations.

Multicystic dysplastic kidney can manifest in an adult as a unilateral multicystic condition. The affected kidney is usually severely dysplastic and non-functioning due to ureteral atresia or agenesis, whereas symmetrical contrast excretion is seen in localized cystic kidney. Due to morphological resemblance of multiple simple cysts with localized cystic disease on imaging as well as pathology, differentiating the two conditions may be challenging especially when multiple simple cysts are involving one kidney predominantly and some authors have proposed that localized cystic kidney disease may represent a variant of multiple simple cysts [13,17, 18]. However, strict unilateral involvement and appearance of cysts as early as in 1st to 2nd decade is very unusual with multiple simple cysts. In addition, simple cysts are usually not as numerous, scattered without cluster formation and more cortically oriented [3, 4, 8]. Localized cystic disease does not require any management and patient can be followed up on serial imaging [1, 12]. Usually the disease remains stable; however, in some case increase in cyst size has been observed with age [1].

To conclude, the purpose of this case report is to familiarize radiologists with this rare entity. Localized cystic disease of kidney must be carefully differentiated from other cystic renal conditions like ADPKD, multicystic dysplastic kidney and cystic neoplasms, as this condition can be managed conservatively.

TEACHING POINT

Localized cystic disease of kidney is a rare benign entity. Familiarity with imaging findings of this condition may be helpful in differentiating it from other cystic renal conditions which require different treatment approaches like ADPKD, multiloculated cystic nephroma and cystic neoplasms. These patients can be safely followed up and unnecessary surgeries can be avoided.

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FIGURES

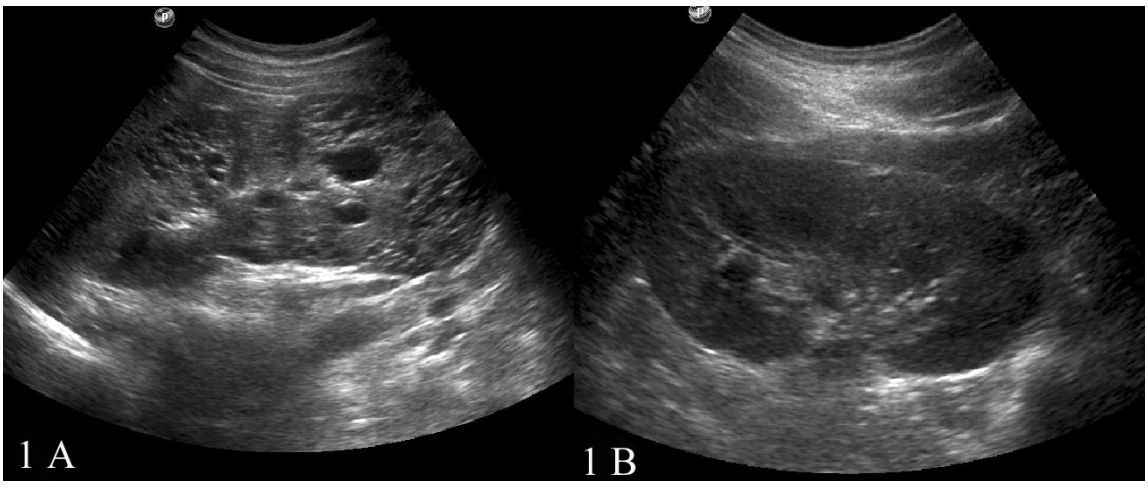

Figure 1: 16-year-old male with localized cystic disease of the kidney presented with acute abdominal pain. (A) Ultrasound of the right kidney shows presence of multiple small simple cysts with relative sparing of the upper pole parenchyma. (B) Left kidney is normal in appearance without any cystic lesion. (Transabdominal ultrasound, HD 11 XE Phillips, C5-2 probe)

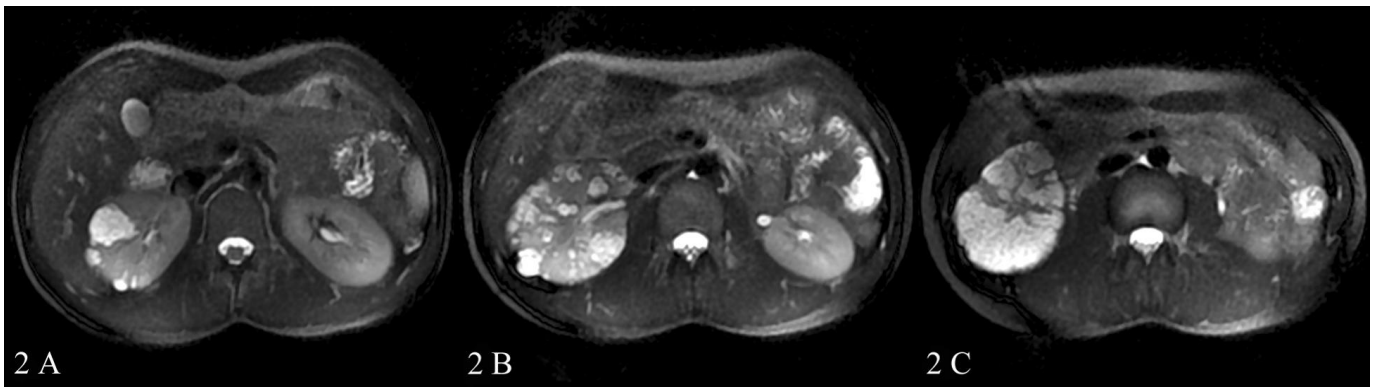

Figure 2: 16-year-old male with localized cystic disease of the kidney presented with acute abdominal pain. (A-C) Axial T2 weighted fat suppressed MR images from cranial to caudal direction. Multiple variable size simple cysts are seen in the right kidney with thin walls. Renal parenchyma of the right lower pole is almost completely replaced by cystic lesions. Left kidney is normal in appearance. (Protocol: 3-Tesla MRI Scanner, GE Healthcare, 5647.06 TR, 77.792 TE, 90 degree Flip angle, 7 mm slice thickness, 8 US TORSOPA coil)

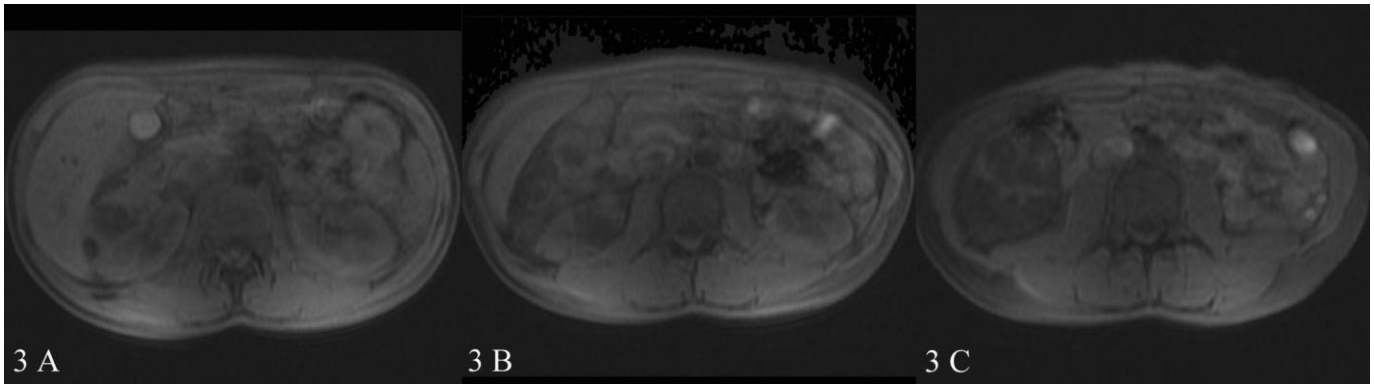

Figure 3: 16-year-old male with localized cystic disease of the kidney presented with acute abdominal pain. (A-C) Corresponding axial pre-contrast T1 weighted gradient MR images from cranial to caudal direction. Cyst contents are hypointense similar to cerebrospinal fluid. (Protocol: 3-Tesla MRI Scanner, GE Healthcare, 3 dimensional, 4.38 TR, 2.108 TE, 12 degree Flip angle, 4 mm slice thickness, 8 US TORSOPA coil)

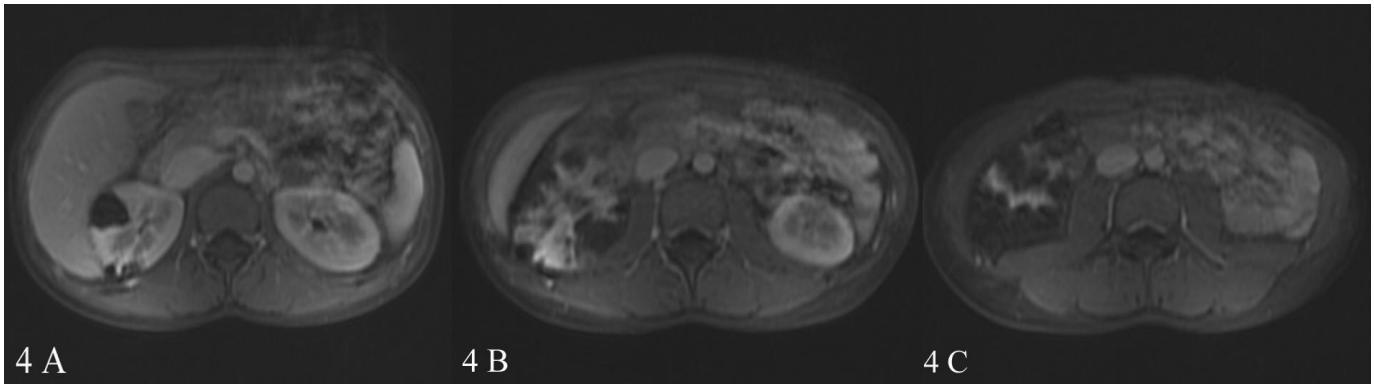

Figure 4: 16-year-old male with localized cystic disease of the kidney presented with acute abdominal pain. (A-C) Corresponding axial post-contrast T1 weighted gradient MR images from cranial to caudal direction. The cystic lesion does not show any wall enhancement, solid component or thick internal septation. Intervening normal enhancing renal parenchyma is noted between the cystic lesions. (Protocol: 3-Tesla MRI Scanner, GE Healthcare, 3 dimensional, 4.38 TR, 2.108 TE, 12 degree Flip angle, 4 mm slice thickness, 8 US TORSOPA coil, 0.2 ml/kg intravenous gadobenate dimeglumine given by hand injection, Multihance, Bracco diagnostics Inc.)

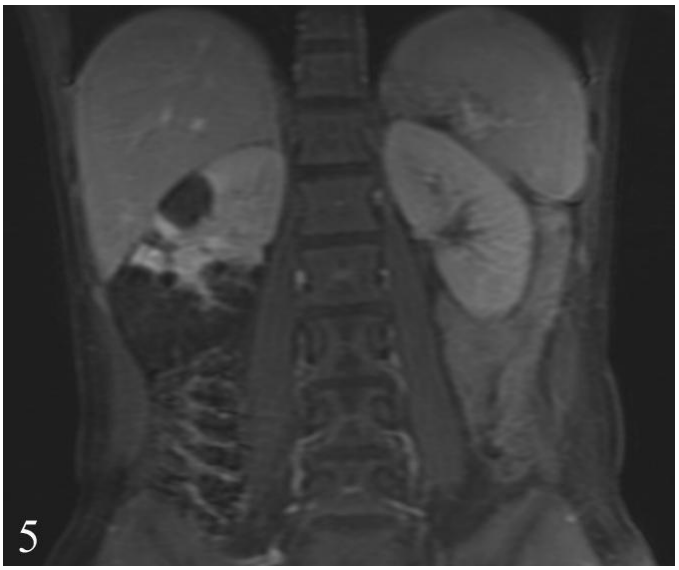

Figure 5 (left): 16-year-old male with localized cystic disease of the kidney presented with acute abdominal pain. Coronal post-contrast T1 weighted gradient MR image. The cysts are more numerous in the right lower polar region with relatively spared upper pole. No cystic lesion is seen in the left kidney. (Protocol: 3-Tesla MRI Scanner, GE Healthcare, 3 dimensional, 4.372 TR, 2.104 TE, 12 degree Flip angle, 4 mm slice thickness, 8 US TORSOPA coil, 0.2 ml/kg intravenous gadobenate dimeglumine given by hand injection, Multihance, Bracco diagnostics Inc.)

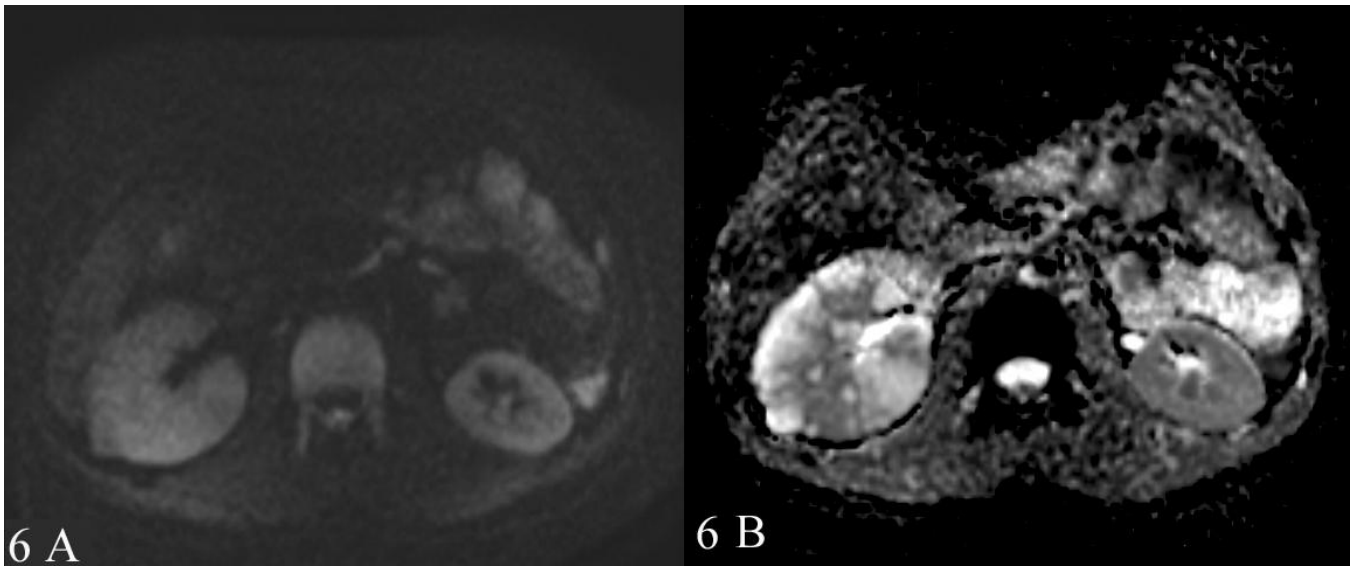

Figure 6: 16-year-old male with localized cystic disease of the kidney presented with acute abdominal pain. (A) Diffusion weighted MR image at the level of right middle pole does not reveal any diffusion restriction in the cystic lesions. (B) Corresponding apparent diffusion coefficient (ADC) image does not reveal any diffusion restriction in the cystic lesions. (Protocol: 3-Tesla MRI Scanner, GE Healthcare, 13333.3 TR, 72.2 TE, 90 degree Flip angle, 7 mm slice thickness, 8 US TORSOPA coil)

	Clinical scenario	Ultrasound	CT scan	MRI
Localized cystic disease of kidney	- No family history - Renal function usually normal. - Non surgical condition	- Multiple cysts are seen affect whole of one kidney or a portion of it. - Localized involvement may be confused with complex mass.	- Multiple cysts of variable size seen separated by normal or atrophic renal parenchyma. - Even if the cysts are clustered in one region, they don't form a distinct encapsulated mass. - Affected kidney shows normal contrast excretion.	- Cysts are smooth walled and show water signal intensity.
Multiple simple renal cysts	- Usually seen in elderly individuals. - Normal renal function	- Multiple variable size cortical based simple cystic lesions are seen. - Kidney size usually remains normal.	- Usually both kidneys are involved. - If unilateral, then difficult to differentiate from localized cystic disease of kidney. However, cysts are usually less numerous, scattered and rarely clustered. - Normal excretion of contrast seen on delayed images.	- Multiple simple cortex based cysts are seen with water signal intensity. - Cysts have imperceptible walls. - Intervening renal parenchyma shows normal enhancement.
Multicystic dysplastic kidney	- Developmental condition. - Usually detected at birth or infancy.	- Multiple peripherally located cysts are seen with solid central area.	- Deformed non-reniform multicystic mass. - Non excretion of contrast is seen on delayed images.	- Normal pelvicalyceal system, ureter or renal vessels are not seen or severely atrophied.
Autosomal dominant polycystic kidney disease	- Positive family history - Renal function deranged. - Most patients develop cysts by the age of 30 years.	- Both kidneys are enlarged with multiple cysts of variable sizes. - Some times may be asymmetrical; however, usually the entire kidney is involved and not localized. - Cysts may be seen in liver, spleen or pancreas.	- Usually both kidneys are involved and cysts are seen in cortex as well as in medulla. - Hyperattenuating cysts are more common. - Associated nephrolithiasis may be present.	- Uncomplicated cysts are hypointense on T1w and hyperintense on T2w images. - Hemorrhagic cysts may show variable signal intensity.
Multiloculated cystic nephroma	- Non hereditary benign condition. - Surgery is usually done due to difficulty in differentiating with cystic neoplasm.	- Multilocular cystic mass ranging from few cm to very large in size.	- Discrete encapsulated lesion with thick fibrous capsule and internal septations. - No other cysts in adjacent renal parenchyma. - Pelvicalyceal system may be distorted.	- The capsule and septae are hypointense on T2 w images due to fibrous nature. - Enhancement is seen in the capsule and septae. - Cyst content may be water signal intensity or proteinaceous.
Cystic neoplasms	- Usually 5-7th decade. - Patient may present with gross hematuria or pain abdomen.	- Cystic lesion with thick internal septae or mural nodule.	- Cystic lesion with multiple septae, septal thickening and / or thick calcification. - Mural thickening, nodule formation, and / or calcification. - Invasion of local structures or metastatic spread may be seen.	- Enhancing thick septae and mural nodule may suggest diagnosis of a cystic neoplasm.

Table 1: Differential diagnosis of localized cystic disease of the kidney

Etiology	Unknown, non hereditary
Incidence	Rare entity with approximately 63 cases reported in the literature.
Gender ratio	Male predilection
Age predilection	Mostly in fifth decade; however, sometimes in the younger age group.
Risk factors	Unknown
Treatment	Usually no treatment is required.
Prognosis	It does not lead to renal insufficiency and prognosis is good.
Imaging findings	- Characterized by replacement of variable amount of one renal parenchyma by multiple cysts in a diffusely scattered or a more localized manner. - Intervening tissue between cysts shows normal enhancement. - Sometimes localized involvement of a portion of kidney may simulate a complex mass due to intervening attenuated renal parenchyma; however, it doesn't shows discrete encapsulation and other cysts may be seen clearly separate from the complex mass. - Despite of replacement of a large portion of the affected kidney by cysts, normal excretion is seen after administration of intravenous contrast agents. - Contralateral kidney shows normal appearance.

Table 2: Summary table of localized cystic disease of the kidney

ABBREVIATIONS

ADPKD = Autosomal dominant polycystic kidney disease
CT = Computed tomography
MRI = Magnetic resonance imaging
MR = Magnetic resonance

KEYWORDS

Cysts; Kidney; Localized cystic disease of kidney; MRI; Unilateral renal cystic disease