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Multicystic dysplastic kidney complicated by pyelonephritis

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 21
Final Diagnosis: Multicystic Dysplastic Kidney Disease complicated by pyelonephritis
Symptoms: Left flank pain (CVAT) • dysuria • fever
Medication: Levofloxacin
Clinical Procedure: Dimercaptosuccinic acid scan • voiding cystourethrogram
Specialty: Nephrology

Objective: Rare disease





Background: Multicystic dysplastic kidney (MCDK) is a renal dysplasia characterized by the presence of multiple cysts that are non-communicating, separated by dysplastic parenchyma that consumes the renal cortex resulting in a non-functional kidney. MCDK has an incidence of 1: 4300 of live births and is usually unilateral, most commonly occurring in the left kidney. Simple MCDK is defined as unilateral dysplasia with a normal contralateral kidney but with compensatory hypertrophy of the contralateral kidney, and no associated genitourinary anomalies.

Case Report: A 21 year old Hispanic American female, presented with intermittent, sharp, severe left flank pain, fever and dysuria for two days but had gradually worsened within the last 24 hours prior to presentation. Previous history of multicystic dysplastic kidney, diagnosed four years ago. No pertinent physical examination findings except left costovertebral angle tenderness (CVAT). Urinalysis findings were positive for infection and urine culture grew pan sensitive Escherichia coli. A CT scan of abdominal and pelvis without contrast revealed a normal right kidney and left kidney had multiple non-communicating dilated cystic spaces, but no hydronephrosis, left ureteropelvic junction obstruction and finding were consistent with multicystic dysplastic kidney and also noted perinephric stranding.

Conclusions: VUR is the most common renal abnormality in patients with MCDK, occurring in about 25% of contralateral kidney. Infections involving the MCDK are rare. In fact, cases of infections such as pyelonephritis or an infected renal cyst of MCDK are almost non-existent in the current literature. This patient presented with findings consistent with MCDK complicated by pyelonephritis.

Key words: multicystic dysplastic kidney • pyelonephritis • voiding cystourethrogram

Full-text PDF: <http://www.amjcaserep.com/download/index/idArt/889557>

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Background

Multicystic dysplastic kidney (MCDK) is a renal dysplasia characterized by the presence of multiple cysts that are non-communicating, varying in size, separated by dysplastic parenchyma that consume the renal cortex resulting in a non-functional kidney with the absence of a normal pelvocaliceal system [1]. It is due to the atresia of the ureteral bud system during the metanephric stage of uterine development with failure of normal nephrogenesis [2]. The incidence of MCDK is approximately 1:4300 of live births, with males being affected more often than females. MCDK can be familial disease but most often occurs sporadically. Most cases of MCDK are detected during fetal ultrasonography and are reported as early as 15 weeks gestation. Studies have shown that MCDK occurs 53% of the time in the left kidney and 47% of the time in the right kidney.

MCDK can be unilateral, bilateral or segmental and can be further sub classified as either simple or complex. Simple MCDK is defined as unilateral dysplasia with a normal contralateral kidney but with compensatory hypertrophy of the contralateral kidney, and no associated genitourinary anomalies. Complex MCDK is defined as bilateral dysplasia of the contralateral kidney or genitourinary anomalies. The contralateral urinary tract may be associated with a variety of other defects, including rotational or positional anomalies, hypoplasia, areas of dysplasia, vesicoureteral reflux (VUR), ureterocele, ureteropelvic junction obstruction (UPJ obstruction), or genital abnormalities [3]. VUR is the most common renal abnormality in patients with MCDK, occurring in about 25% of contralateral kidney. Bilateral MCDK leads to absent fetal and neonatal renal function associated with pulmonary hypoplasia and therefore is incompatible with extra uterine life [4]. The disease is found to occur bilaterally in 19% to 34% of cases.

Environmental influences, such as maternal antiepileptic drugs and chromosomal defects on the occurrence of MCDK have been identified. Most cases of MCDK seem to result from sporadic malformations, although familial association has been recognized. One research study identified a mutation in exon 2 of PAX2 gene that causes renal coloboma syndrome and is associated with MCDK [5]. It was suggested that PAX2 may also play a role in early ureteric obstruction. The protooncogene B-cell lymphoma-2 (BCL-2) is associated with the formation of renal cysts [6]. Exposure to viral infections in utero has been associated with MCDK, such as cytomegalovirus, enterovirus, and adenovirus have been implicated in the development of renal dysplasia. We hereby present a case of a young adult female with unilateral multicystic dysplastic kidney disease complicated by pyelonephritis versus an infected renal cyst.

Case Report

A previously healthy 21 year old Hispanic American female, whom recently delivered a healthy baby girl one month prior via normal spontaneous vaginal delivery, presented with intermittent, sharp, severe left flank pain with no radiation for two days but had gradually worsened within the last 24 hours prior to presentation. Other complaints included a high grade fever of 40.1°C, chills, nausea, vomiting of three episodes, and dysuria, all during the last two days prior to admission. She denied any complaints of headaches, blurry vision, chest pain, shortness of breath, diarrhea or hematuria. The patient had no past medical problems other than a history of multiple left renal cysts that were noted on a previous hospitalization, four years prior to this admission. Genetics tests were done at that time were performed to rule out causes for renal cystic diseases. Mutations in the PKD1, PKD2, and PKHD1 genes that cause polycystic kidney disease, MUC1 gene that cause medullary cystic kidney type 1 and UMOD gene that cause medullary cystic kidney disease type 2 were all genetically tested and negative.

Patient stated that she was not taking any medications. Family history was not significant for intracranial hemorrhages, subarachnoid hemorrhages, berry aneurysms, polycystic kidney disease or renal failure. She denied using drugs, alcohol or tobacco. The imaging studies that were performed four years prior included a renal ultrasound that showed a left kidney measuring 13.7 centimeters longitudinally and right kidney measuring 11.6 centimeters longitudinally. There was marked dilation of the central collecting system and calyces in the left kidney with marked thinning of the renal cortex.

Vitals signs revealed a temperature of 39.3°C, blood pressure of 107/53 mmHg, tachycardia (HR: 92 to 105) beats per minute, respiratory rate of 18 breaths per minute. No pertinent physical examination findings except left costovertebral angle tenderness (CVAT). The white blood cell count on admission was $11.7 \times 10^3/\mu\text{L}$, hemoglobin of 14.2 g/dL, platelet count of $345 \times 10^3/\mu\text{L}$, sodium level of 138 mmol/L, potassium level of 4.0 mmol/L, BUN of 10 mg/dL, creatinine level of 0.8 mg/dL and glucose level of 113 mg/dL. Liver function tests were normal. Urinalysis findings included a 3 + bacteria, moderate blood, RBC 3–5, WBC 10–15, WBC casts, positive leukocyte esterase and nitrite. Due to the urinalysis result, ceftriaxone 2 g IV daily was initiated empirically. The urine culture grew pan sensitive *Escherichia coli*. On hospital day 2 the WBC elevated to $14.6 \times 10^3/\mu\text{L}$, BUN of 18 mg/dL, and creatinine of 1.6 mg/dL.

A dimercaptosuccinic acid (DSMA) scan was then ordered to assess the renal function, essentially revealing that the left kidney consisted mostly of non-functional renal parenchyma. The right kidney was normal appearing. A voiding cystourethrogram

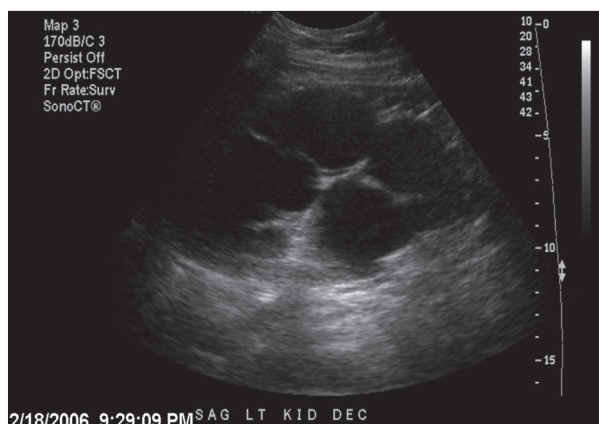


Figure 1. Renal ultrasound – left kidney consists of multiple non-communicating thin walled cysts that are randomly distributed.

(VCUG) revealed a normal appearing bladder with no vesicoureteral reflux. The abdominal ultrasound showed that the right kidney measured 11.5 cm with no cysts. Left kidney measured 14.2 cm (enlarged and irregular) and consisted of multiple non-communicating thin walled cysts that are distributed randomly (Figure 1). No normal renal tissue is apparent on ultrasound. A CT scan of abdominal and pelvis without contrast revealed that the right kidney was normal and not hypertrophic. Left kidney appeared to be grossly abnormal to the point it was described to look as a large hydronephrotic sac with multiple non communicating dilated cystic spaces, but no actual hydronephrosis, left ureteropelvic junction (UPJ) obstruction and finding were consistent with multicystic dysplastic kidney. It was also noted for the presence left perinephric stranding upon further review with radiologist.

The infectious disease consult service was called on second hospital day for further recommendations. In their opinion the patient had left side pyelonephritis however they could not rule out an infected renal cyst. The renal function improved throughout hospital stay and patient was discharged on hospital day six. She was discharged on levofloxacin 500 mg one time per day for a total antibiotic course of 14 days given the possibility of an infected renal cyst. She was advised to follow up with her primary care physician and nephrologist in one to two week after discharge. Her appointment was approximately 1 month after being discharge. At this clinic visit the patient was clinically improved, did not have any complaints and no longer had CVAT. She was advised regarding her condition, follow up, management, and prognosis.

Discussion

Multicystic dysplastic kidney may persist without any change, increase in size, or may undergo spontaneous involution [7].

Most cases of unilateral multicystic dysplastic kidney undergo spontaneous involution [8]. There has been concern for the potential of malignant degeneration in a MCDK. Most case reports have been of Wilms tumor, renal cell carcinoma and urothelial carcinomas that developed in a MCDK [9]. Surgery in dysplastic kidney is required only when the enlarged kidney fails to regress, causes obstructive uropathy, develops malignancy, or the dysplastic kidney become a cause of hypertension or UTI [10]. As the incidence of complications like hypertension, malignancy, or UTI in dysplastic kidney is low, surgical intervention in dysplastic kidney is rarely required. However some studies suggest that the non-functional MCDK may be removed to avoid the risk of hypertension or malignancy, but the patient will only have one functional kidney, and that functional kidney may be at risk due to an increased incidence of contralateral abnormalities [11]. But controversy still exists regarding the role of routine clinical and radiological monitoring in adults due to the risk of hypertension and malignancy [12].

MCDK is usually asymptomatic and can remain undiagnosed until adulthood. Most adults that have MCDK and one normal functioning kidney will lead a normal healthy life. Since they have one functioning kidney there should be regular follow up to check for high blood pressure and kidney insufficiency. Several studies suggest that adults with uncomplicated MCDK are best managed conservatively [13]. Serial ultrasounds also may detect any significant renal scarring to the normal contralateral kidney due to recurrent urinary tract infection [14].

Many concurrent urinary tract abnormalities have been described in patients with MCDK. The most common and potentially significant urologic defect seen is VUR to the contralateral kidney. Other urinary tract abnormalities, such as contralateral ureteropelvic junction (UPJ) obstruction are often seen in patients with MCDK [15]. Although MCDK is rare, the possible complications that may arise highlight the importance of recognizing, appropriately diagnosing, and treating this disorder. Renal ultrasonography is the recommended preliminary diagnostic imaging study and voiding cystourethrography (VCUG) should be performed to look for vesicoureteral reflux (VUR). Patients with unilateral multicystic dysplastic kidney usually have a higher incidence of contralateral vesicoureteral reflux than the general population [16]. Therefore the current recommendations state that the contralateral kidney should also be evaluated for VUR by VCUG [15]. Dimercaptosuccinic acid (DMSA) renal scanning may also be necessary if ultrasonography does not reveal the classic features of multicystic dysplasia of the kidney.

Conclusions

Infections involving the multicystic dysplastic kidney are rare. In fact, cases of infections such as pyelonephritis or an infected

renal cyst of multicystic dysplastic kidney are almost non-existent in the current literature. Pyelonephritis normally occurs in the contralateral normal kidney due to the presence of VUR. However, our patient with MCDK presented with several interesting features, such as a VCUG that demonstrated no VUR, a normal contralateral kidney with no compensatory hypertrophy and a rarely documented infection of the multicystic dysplastic kidney rather than the contralateral kidney. This patient

had the clinical presentation, laboratory findings and imaging that were consistent with MCDK complicated by pyelonephritis.

Disclosures

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