

(CASE REPORT)



Nephrectomy of Multicystic Dysplastic left Kidney in an 18-year-old female: A case report and literature review at Sunyani Teaching Hospital (Ghana)

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Abstract

We herein present our patient with left MCDK, the associated symptoms were the left flank pain and left flank mass. Though conservative management is proven to have some favourable outcomes, a nephrectomy was done for our patient due to the increasing intensity of pain and abdominal mass which was affecting the patient's quality of life. Left sub-costal extraperitoneal nephrectomy was performed successfully at the CJ Oppong theatre of Sunyani Teaching Hospital in Ghana which gave a new life full of joy to our patient. We, therefore, recommend nephrectomy for a patient with increasing symptoms that reduce quality of life and early childhood screening for MCDK.

Keywords: Multicystic; Dysplastic kidney; Abdominal mass; Nephrectomy

1. Introduction

Multicystic dysplastic kidney (MCDK) is a congenital kidney abnormality with an overall incidence rate of 1/4300 live births (Pourpashang et al., 2023). The condition occurs more commonly in males than females (2.4:1) and the left kidney is more affected than the right kidney (Mehtap et al., 2021., Cambio et al. 2008). It is a congenital developmental anomaly consisting of multiple variably sized cysts, with little or no normal renal tissue discernible. MCDK is usually unilateral and involves the entire kidney, with an incidence of 1:2400–4300 live births. Histopathologically, the appearance of the kidney characteristically varies with number and size of cysts and the amount of solid tissue (Fong et al., 2021).

It is part of the spectrum of Congenital Abnormalities of the Kidney and Urinary Tract and an increasing number of children are being diagnosed on antenatal ultrasound (Paul W. & Lyn S. C. 2007). MCDK is a unique entity in the broad spectrum of cystic diseases of the kidney, with classical features including nonfunctioning renal parenchyma accompanied by multiple noncommunicating cysts. In the era of routine antenatal ultrasound scanning, MCDK is being diagnosed more frequently (Mushtaq, et al., 2023). Although the use of antenatal and postnatal ultrasonography (US) has become wide spread in the past 20 years, MCDK may still be asymptomatic and may not be detected until adulthood (Mallik & Watson, 2008). In the past, some patients may not have been detected until adulthood, usually following investigations and management for other illness, or even as incidental findings at post mortem, unless there was severe bilateral dysplasia leading to Potter's sequence or renal failure in childhood (Paul W. & Lyn S. C. 2007). Multicystic dysplastic kidney (MCDK) is the most common renal cystic disease in infancy and the second most common neonatal abdominal mass. It represents an extreme form of renal dysplasia. The key features in MCDK include variability in size, loss of kidney shape, absence of function and obstruction of ureter. Current management is still controversial. The prognosis is largely depended upon the problems of associated anomalies and the condition of the contralateral kidney (Chang et al., 1993)

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It is the most common prenatal diagnosed cystic lesion of the kidney that occurs unilaterally or bilaterally and is associated with other structural ureteral abnormalities, such as ureteropelvic junction obstruction, vesicoureteral reflux and ureterovesical obstruction (Schreuder et al., 2009). It is characterized by the enlargement of the kidney, multiple, noncommunicating cysts of varying size, no functioning kidney parenchyma, and atresia or hypoplasia of the ureter (Winyard & Chitty, 2008). For those cases not diagnosed prenatally, presenting symptoms usually include a palpable abdominal mass, pain, urinary tract infections, haematuria, and hypertension (Imaran M. et al., 2023).

The failure of mesenchymal metanephros to differentiate into ureteral bud epithelial cells may be the cause of MCDK. The presence of numerous thin-walled cysts without connections within a kidney with irregular outline is the classic MCDK ultrasound appearance. This appearance could be seen for the first time between 15 and 20 weeks of gestation (Al Naimi et al., 2013).

Most MCDK are unilateral, more frequently affecting the left kidney, and have a favourable prognosis. Contrarily, bilateral MCDK is a rare condition with a poor prognosis. Infants with bilateral kidney disease frequently die during the neonatal period. External anomalies, anhydramnios, pulmonary hypoplasia and chromosomal anomalies often are associated with bilateral MCDK (Brown et al., 2019). MCDK is mostly an isolated condition; however, the contralateral urinary tract could be affected by other abnormalities such as vesicoureteral reflux (VUR), ureteropelvic junction obstruction (UPJO) and ureterovesical obstruction (UVJO) (Mansoor et al., 2011).

There is still significant controversy and differences of opinion in regard to the possibility of malignant transformation, and for this reason the management of MCDK is mostly conservative and dictated primarily by the coexisting abnormalities. Nephrectomy is only indicated in symptomatic cases or as a consequence of parental choice (Carmack et al., 2006).

Confusion between MCDK and fetal hydronephrosis may occur, particularly in cases with a single predominant large cyst. Antenatal diagnosis may be changed in some patients owing to imaging results and affects prenatal counseling of parents and postnatal therapeutic planning (Fong et al., 2021).

There are three major issues in patients with unilateral MCDK and the fate of the contralateral functioning kidney (Erich, Lipsky & Braga 2019). Firstly, MCDK may undergo complete or partial involution within time (Kiyak et al., 2009). Secondly, although it is presumably thought to have increased risk for the incidence of Wilms tumor in the past, it is not widely accepted today [Calaway et al., 2014]. And thirdly, the risk of hypertension in MCDK has been found to be high but no higher than in the general pediatric population (Erich, Lipsky & Braga 2019).

A study found that girls with bilateral MCDK were twice as likely as boys to have non-renal associated anomalies and four times more likely to have chromosomal anomalies as boys, this could imply that male fetuses with severe malformations are less likely to survive. Moreover, MCDK is typically a sporadic anomaly rather than a familial one, hence screening of relatives is not relevant (Winding et al., 2014).

The management of MCDK has changed greatly over time. Previously, nephrectomy was performed to avoid infection, pain, hypertension, and malignancy (Chang et al., 2018); however, the risks of these complications were ill-defined (A. Chang et al., 2018). Recently, management is primarily conservative due to favourable outcome of patients (Okada et al., 2003).

We report on a 17yr old who presented with 2year history of left flank pain, erroneously interpreted as a severe hydronephrosis. Details of the history, radiological evaluation, morphological characteristic, and clinical aspect of this case are presented.

2. Case Presentation

A 17-year-old was seen in our facility with 2 years history of left flank pain. The pain was of gradual onset that worsened during menstruation, causing significant limitations in her usual daily activities. Pain was described to be dull, cramping and sometimes felt as fullness, lasting for days or weeks and rated 6 to 8 out of 10 on a visual analogue scale. Usually, exacerbation of the pain is unpredictable and relieved by analgesics. About six months later, patient started to feel a mass at the left flank which further worsened the intensity of the pain, sometimes causing her to skip school. Patient's

presentation did not associate with edema, anuria or hypertension. She has three other siblings with no similar complaints.

Prior to presentation at our facility, patient had been managed for pyelonephritis in one of the peripheral facilities.

On clinical examination, there was a round palpable mass at the left flank area measuring about 6x4cm, tender, soft in consistency, well defined edges, smooth surface, not adherent to overlying skin with positive left renal angle tenderness. An impression of multicystic left kidney was made.

Ultrasound of the abdomen and pelvis: first scan showed enlarged left kidney measuring 17x9.8cm with multiple communicating pelvicalyceal system and hydronephrosis of left kidney. Scan was repeated at a different facility and showed left kidney containing multicystic fluid measuring 4.9cmx5.5cm, and appears to be dilated, suggestive of hydronephrosis. From both scans, the right kidney was normal with good corticomedullary differentiation, fig 1A below.



Figure 1 Urological Ultrasound scan of right and left kidneys

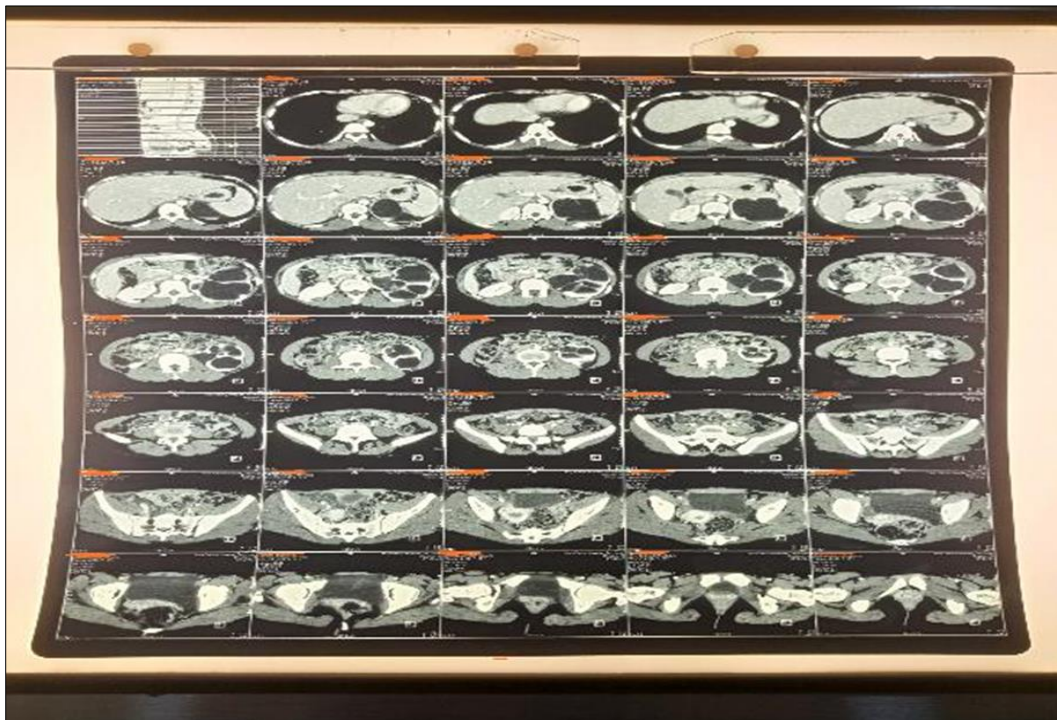


Figure 2 Contrast CT of the abdomen and pelvis showing left multicystic kidney

To further support diagnosis, CT Urography was done and found left kidney with ballooning of the renal pelvis (antero-posterior diameter of 4.4cm), marked dilatation of the calyceal system, severe renal cortical thinning (thickness of 3.5mm) and a collapsed (non-dilated) left ureter in the pre-contrast and corticomedullary phase of the imaging; finding features suggestive of a left pelvic-ureteric junction obstruction, normal right kidney. Figure 2 below.

Other auxiliary investigations including Serum electrolyte levels, liver function test, full blood count, blood urea and creatinine, urine routine examination were all normal.

Patient and relatives were adequately counselled for nephrectomy which they consented but surgery was postponed for 1yr to enable patient graduate from Senior High School.

After successful graduation (patient now 18yrs), patient was reviewed at the Out Patients' Department and booked for nephrectomy. She was admitted and adequately hydrated for at least 24hrs prior to the surgery, with sufficient urine output. Nephrectomy (left) was finally done on 10th October, 2023 at C. J Oppong Theatre – Sunyani Teaching Hospital (Ghana).

Under general anesthesia, subcostal incision was made. Anterior abdominal muscles were bluntly dissected into the extraperitoneal space where the left renal vessels and ureter were ligated. Intra-operative, a large soft mass measuring about 13x7cm containing multiple cysts of varying sizes that are randomly arranged and separated by thin membrane, the largest measuring about 4x3cm. About 500mls of cystic fluid was suctioned to reduce volume for easy access, with no extraperitoneal fluid, refer to figure 3 below.



Figure 3 Intra-operative appearance of the left kidney



Figure 4 Post nephrectomy of left kidney bed, with a drain

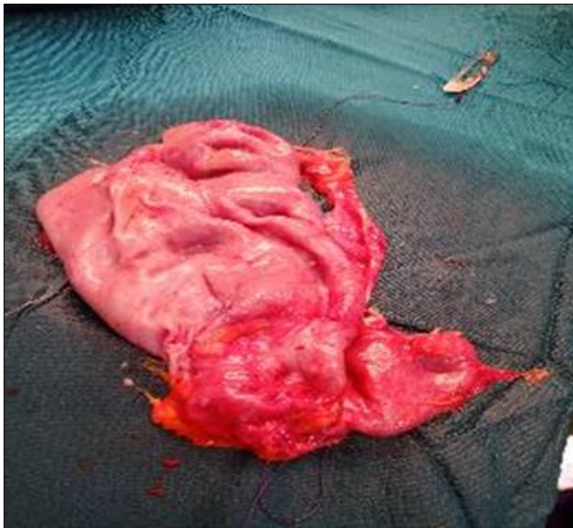


Figure 5A Nephrectomy specimen, solely capsule



Figure 5B Dissected nephrectomy specimen

Nephrectomy specimen contained multiple pockets of cysts of varying sizes with virtually no parenchymal tissues.

Patient was managed on the ward for 6 days post nephrectomy. Vital signs were all within their normal ranges, post-surgery labs (full blood count, blood urea, creatinine and electrolytes were all unremarkable). Patient's recovery was uneventful and was discharged from hospital on post-operative day 6 on oral antibiotics, analgesics and alternate day wound dressing.

Patient was scheduled for weekly reviews for 6 consecutive weeks, 2 weekly reviews for 8 wks and monthly for 6 months.

Nephrectomy kidney specimen was sent for histopathological analysis, which confirmed dysplastic left kidney.

3. Discussion

Multicystic dysplastic kidney as one of the most common anomalies detected by prenatal ultrasound has an overall incidence of 1/4300 live births (Pourpashang et al., 2023), making it rare in terms of the general population.

The condition occurs more commonly in males than females (2.4:1) and the left kidney is more affected than the right kidney (Mehtap et al., 2021., Cambio et al. 2008). It is a congenital developmental anomaly consisting of multiple variably sized cysts, with little or no normal renal tissue discernible. MCDK is usually unilateral and involves the entire kidney. Histopathologically, the appearance of the kidney characteristically varies with number and size of cysts and the amount of solid tissue (Fong et al., 2021). These facts are consistent with the presentation and histopathology findings of our patient.

Although MCDK is being diagnosed more frequently (Mushtaq, et al., 2023) through antenatal and postnatal ultrasonography, MCDK may still be asymptomatic and may not be detected until adulthood (Mallik & Watson, 2008), such was our case whose condition was neither detected during routine antenatal ultrasound and postnatal scanning until age 17.

MCDK usually occurs unilaterally and is associated with other structural ureteral abnormalities, such as ureteropelvic junction obstruction, vesicoureteral reflux and ureterovesical obstruction (Schreuder et al., 2009). These facts are consistent with the CT findings of our patient. It is characterized by the enlargement of the kidney, multiple, noncommunicating cysts of varying size, no functioning kidney parenchyma, and atresia or hypoplasia of the ureter (Winyard & Chitty, 2008).

The classic type of MCDK has a random configuration of cysts, whereas the hydronephrotic type presents with a discernible and dilated renal pelvis surrounded by numerous peripheral cysts. The peripheral cysts may communicate with each other or with the renal pelvis (Pourpashang et al., 2023). Based on the gross appearance and pathological findings of the MCDK, our case illustrated like a typical hydronephrotic type with classical features including nonfunctioning renal parenchyma accompanied by cysts of varying sizes. It seemed to belong to a hydronephrotic variant of MCDK.

3.1. Ethical issues

Patient and parents have given consent for publication as a case report. ethical issues including plagiarism, data fabrication, double publication have all been completely observed by the authors.

4. Conclusion

MCDK is a rare congenital kidney abnormality common in males than female, usually unilateral affecting left kidneys than the right. Despite advancement in prenatal and postnatal diagnostics, MCDK can be misdiagnosed mostly in low resource countries in sub-Sahara Africa including Ghana. Though our patient had left MCDK, the associated symptoms were the left flank pain and left flank mass. Though conservative management is proven to have some favourable outcome, nephrectomy was done for our patient due to increasing intensity of pain and abdominal mass which was affecting patient's daily activities and decreasing her quality of life.

It is also important to note that other abnormality, left pelvic-ureteric junction obstruction, was seen on supportive CT scan. MCDK was also confirmed by histopathology report, surgery and clinical findings.

We therefore recommend nephrectomy for a patient with increasing symptoms that reduce his or her quality of life. We also recommend early childhood screening for MCDK.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

Ethical issues including plagiarism, data fabrication, double publication have all been completely observed by the authors. Study approved by the institution ethical committee.

Statement of informed consent

The patient and parents have given consent for publication as a case report

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