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BILATERAL MULTICYSTIC DYSPLASTIC KIDNEYS: CASE REPORT
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BILATERAL MULTICYSTIC DYSPLASTIC KIDNEYS: CASE REPORT

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SUMMARY

Multicystic dysplastic kidney (MCDK) is a congenital, non-heritable maldevelopment in which the renal cortex is replaced by numerous cysts of varying sizes. Typically, MCDK is a unilateral disorder in 76% of cases and bilateral in 24%. The latter is incompatible with life. We present the case of a male child with bilateral MCDK who was followed up with ultrasound scan (USS) from 22nd week of intrauterine life till death on the 19th day after delivery. Serial antenatal USS revealed oligohydramnios and both kidneys showed multiple cysts which appeared to be communicating. Postnatal USS however, revealed multiple sub-cortical thin walled cysts of varying sizes which apparently appeared to be communicating. A diagnosis of bilateral MCDK was made and was confirmed at autopsy.

INTRODUCTION

Multicystic dysplastic kidney (MCDK) is a congenital, non-heritable mal-development in which the renal cortex is replaced by numerous cysts of multiple sizes (1). It is unilateral in 76% of cases and bilateral in 24%. Bilateral occurrence is incompatible with life (2). Our case was the rarer bilateral MCDK. Atresia of the pelvis and ureter and an absent arterial supply is also common (3).

High resolution ultrasound examination of the foetus enables early detection of congenital malformations of the urinary tract (4). Such information is of value in determining foetal prognosis, in deciding the method of delivery and in alerting the pediatricians to supportive and corrective measures that may need to be taken in the post-natal period. In bilateral renal dysplasia, renal failure ultimately results.

CASE REPORT

A19-day-old male, with antenatal diagnosis of severe oligohydramnios from ultrasound (USS) done at 22nd and 30th week gestational age (GA). He was delivered at 36 weeks GA via emergency Caesarean section.

Post-natal USS done on the first and fifth days of life revealed bilaterally enlarged kidneys with multiple sub-cortical cysts. Sinuses were demonstrated and the pelves were not dilated in both kidneys (Figure 1). Both ureters were not outlined and urinary bladder was empty. Micturating cystourethrogram (MCUG) performed on the fifth day post-delivery revealed a normal urinary bladder with uniform outline and no vesico-ureteric reflux (VUR). The posterior and anterior urethrae were normal in outline and calibre. On the basis of USS and MCUG a radiological diagnosis of bilateral MCDK was made.

He was anuric with worsening renal functions and generalised oedema. He had peritoneal catheter insertion and peritoneal dialysis was commenced on the 10th day post-delivery but his condition deteriorated with worsening respiratory distress and temperature instability. He developed apneic attacks and subsequently died on the 19th day.

At autopsy, body was that of a male neonate that measured 42cm in length with crown-rump length of 29cm and occipito-frontal diameter of 35.5cm. There was surgical incision in the left peri-umbilical region representing the site of peritoneal dialysis. Also present were generalised oedema, bilateral talipes vara and moderate palor.

Internal examination revealed right and left kidneys that were multicystic with loss of reniform appearance (Figure 2) and weighed 20gm each. Capsule stripped with ease to show numerous cysts of varying sizes irregularly distributed within the cortex and medulla. The cortico-medullary junction was absent. The ureters were atretic while the bladder and urethra were hypoplastic.

Histology sections from the kidneys showed renal tissue with islands of haphazardly arranged tubules and abortive glomeruli (Figure 3) with absence of cortico-medullary differentiation. There were numerous cysts lined by flattened epithelial cells distributed within the renal tissue. The intervening stroma was made up of immature and myxoid tissue.

Assessment of bilateral multicystic dysplastic kidney (BMCDK) with bilateral talipes vara was made

Figure 1
Renal USS showing multiple non-communicating cysts and central hyperechoic sinus

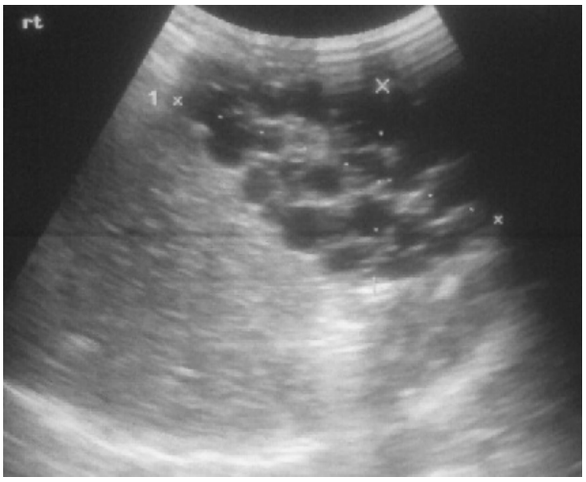


Figure 2
Gross specimen of bilateral multicystic dysplastic kidneys

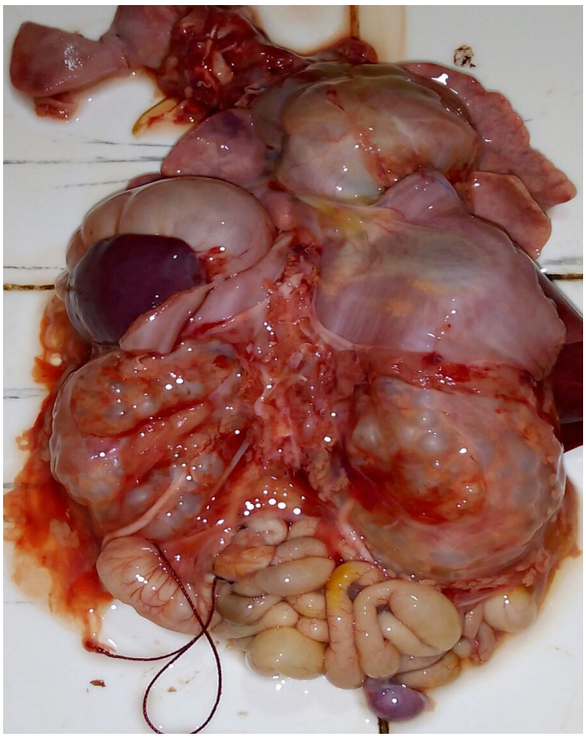
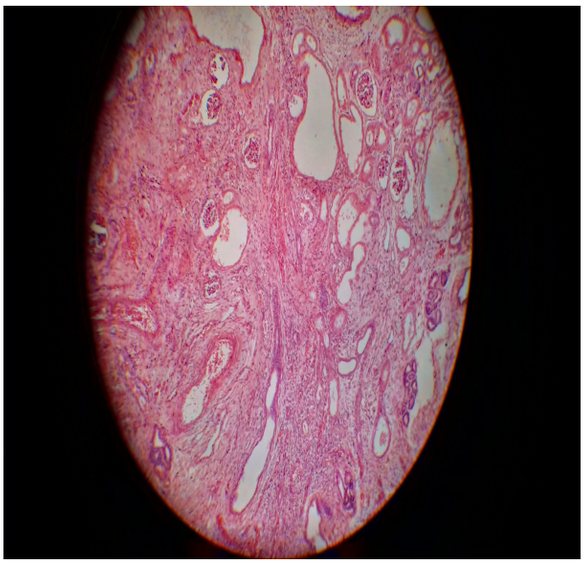


Figure 3
Histology section showing cystic cavities and abortive/ immature glomeruli (x 200)



Both kidneys were riddled with numerous cysts of varying sizes. Atretic ureters originate from medial surface of the kidneys and lie on coils of intestine.

DISCUSSION

Multicystic dysplastic kidney (MCDK) is a congenital, non-heritable mal-development in which the renal cortex is replaced by numerous cysts of varying sizes (1). Several forms of MCDK have been described. The classic (pelvi-infundibular) is the most common type where there are multiple small non-communicating renal cysts, which replace the dilated calyces with atresia of the ureter and renal pelvis. The hydronephrotic type is diagnosed when there is a discernible, dilated renal pelvis surrounded by cysts. The third type is the solid cystic dysplasia which is composed of smaller cysts (microscopic) with a greater amount of non-functional parenchyma (1,5). In this case report, renal pelves were not dilated, ureters were atretic and there were numerous cysts (Figure 1) thus, favoring the classic type.

The etiology of MCDK is not well understood, but it is thought to result from abnormal induction of the nephrons in the mesonephric blastema by the migrating ureteric bud (5). Although the pathogenetic process leading to a MCDK begins at the 8th week *in-utero*, the mean age at the time of the antenatal diagnosis is about 28 weeks, with a range of 21-35 weeks (6). This child had antenatal presentation at the 24th week gestational age.

MCDK is the most documented renal cystic complication in children. It is often unilateral, with a unilateral incidence estimated at 1:2500-4300 live birth, predisposition for the left kidney and

a slightly higher incidence in males with a M:F of 2.4:1.However, the female foetuses are twice likely to have the fatal bilateral disease and associated non-renal abnormalities for example VACTERL, congenital heart disease (2). Studies have shown that the unilateral MCDK have increased incidence of congenital anomaly in the contralateral kidney like VUR which is the most common and seen in 18-20%, pelvi-ureteral junction (PUJ) obstruction, vesico-ureteral junction (VUJ) obstruction and ureterocele (7). This case of bilateral MCDK involved a male child with bilateral talipes vara.

In Nigeria, Aremu *et al* (5) reported two cases of unilateral MCDK. They presented as abdominal masses which are atypical in adults but the most common presentation in children.

Ultrasonography is the first line of study as it is fast, accurate, requires no sedation, radiation or other interventions. As shown in this case, it is useful for prenatal diagnosis and for post-delivery monitoring. Classic MCDK can be diagnosed antenatally as the multiple cysts become evident as early as 15th week GA (8). YBA had antenatal USS which showed these features except that the thin walled cysts appeared apparently communicating which was most likely artefactual, on the basis of which the diagnosis of hydronephrosis was made. In our patient, the antenatal diagnosis of MCDK was missed due to the presence of apparent communication between the cysts, even though other diagnostic criteria such as, bilateral multicystic kidneys, loss of renal architecture, non-visualization of the foetal urinary bladder and oligohydramnios were present (9).

At autopsy, both kidneys were confirmed to be grossly enlarged with multiple cysts of varying sizes. Autopsy is recommended to arrive at a definitive diagnosis and to distinguish between MCDK and other renal cystic diseases. The visualization of primitive ducts with fibromuscular collar (Fig. 3), which is the indispensable feature of renal dysplasia with lobar disorganisation and cysts, confirms the diagnosis (10).

This case shows the importance of the presence of renal sinus which appears as central hyperechogenicity (Fig.1) in the differentiation of MCDK from other entities such as PUJ obstruction which may lead to dilated renal pelvis, calyces and splayed renal sinus in the antenatal period. This is important as precise diagnosis of bilateral MCDK which is incompatible to life, may allow patients the option of an elective abortion or an informed consent.

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