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Prenatal Diagnosis of Multicystic Dysplastic Kidney and its Histopathological Correlation

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Abstract

Multi Cystic Dysplastic Kidney (MCDK) develops in utero and the prenatal diagnosis is often made by an ultrasound scan. The vast majority are sporadic and non-familial. Rarely autosomal dominant forms are seen. It is the commonest cause of abnormally enlarged kidney, diagnosed on antenatal ultrasound examination. It is typically a unilateral disorder; bilateral condition is incompatible with extra uterine life. Association of this disease with abnormalities of various organs is common. We report a rare case of unilateral MCDK associated with contralateral atrophic kidney with hydroureter, and hence the pregnancy was terminated.

Keywords: Multicystic dysplastic kidney, Prenatal diagnosis, Unilateral MCDK.

Introduction

Multicystic dysplastic kidney (MCDK), a variant of renal dysplasia, is one of the most frequently identified congenital anomalies of the urinary tract[1]. MCDK is characterized by the presence of multiple, non-communicating cysts of varying size separated by dysplastic parenchyma and the absence of a normal pelvocaliceal system. The condition is associated with ureteral or ureteropelvic atresia, and the affected kidney is nonfunctional. It is typically a unilateral disorder.

Renal anomalies detected in antenatal period in sonography may modify obstetric management, facilitate paediatric and surgical care of the new born or may allow for elective termination of pregnancy. On routine antenatal ultrasound, the incidence of congenital renal malformations is about 0.2%. The incidence of MCDKD is about 1 in 5,000-10,000 births[2]. Up to 39% of patients present with associated anomalies in the contralateral kidney. The left kidney is involved in 55% of cases, and the right kidney is involved in 45% cases[3]. Multicystic dysplastic kidney is the most common cause of an abdominal mass in the newborn period and is the most common cystic malformation of the kidney in infancy.

We hereby, describe a case of multicystic dysplastic kidney disease with contralateral atrophic kidney with hydoureter diagnosed antenatally and the correlation between histopathology and sonography is discussed.

Case Report

A 22 year old primigravida with unremarkable prenatal history reported for routine antenatal ultrasound during second trimester. Gestational age by her dates was 19

weeks. Biparietal diameter and femoral length was corresponding to the gestational age of 18-19 weeks. The left fetal kidney was enlarged, measuring 42 x 28 mm and contained multiple sonolucent cystic structures of variable size ranging from 3-7 mm in diameter ***[Figure – 1a]***. No corticomedullary differentiation was noted. The right kidney was small, echogenic without any corticomedullary differentiation. Renal pelvis was dilated with a diameter of 6 mm and right ureter was also dilated. Surprisingly, amniotic fluid was normal in this case which indicates late onset of obstruction in the atrophic kidney or the mucticystic kidney was partly functional. So, the USG findings were suggestive of Left Multi Cystic Dysplastic Kidney (MCDK) along with Right Atrophic Kidney with UVJ obstruction.

Fetal Karyotype was done which was normal. Guarded prognosis was explained to the parents. Parents decided for termination of pregnancy. Consent for autopsy was obtained, and performed post-expulsion of male abortus. Autopsy findings were in concordance with ultrasonographic findings. The gross appearance of bilateral kidneys showing left polycystic kidney and right atrophic kidney ***[Figure – 1a]***. Gross appearance of hydroureter on the right side and atretic ureter on the left side was also noted during autopsy ***[Figure – 1c]***. On histopathology, both kidneys had dysplastic changes ***[Figures – 2a & 2b]***. No other external abnormality was noted.

Discussion

Renal dysplasia, defined as abnormal metanephric differentiation, has variable presentations that cover a spectrum of conditions, including hypoplasia, multicystic dysplasia and aplasia. Most dysplastic kidneys are associated with some form of obstruction of the urinary tract during nephrogenesis. Based on these criteria, renal dysplasia has been broadly subdivided into four groups[4]:

- Group I: classical multicystic dysplasia resulting from ureteral atresia at pelviureteric junction
- Group II: focal and segmental cystic renal dysplasia usually caused by obstruction related to ectopic uretrocele with ureteral duplication
- Group III: cystic renal dysplasia associated with lower urinary tract obstruction e.g. posterior urethral valves
- Group IV: familial renal dysplasia without any obstructive uropathy.

In classical MCDKD the entire kidney is involved and is said to be more common in males. In contrast to classical MCDKD segmental dysplastic kidney disease typically affects upper pole of a duplex kidney and is usually noted in females. Associated contralateral renal tract abnormalities are common (seen in ~50% of cases) and include: vesicoureteric reflux (VUR): most common and seen in up to 20%, pelviureteric junction (PUJ) obstruction, ureteral ectopia, vesicoureteric junction (VUJ) obstruction, ureterocoele[5].

Ultrasonographically detected fetal renal abnormalities may be unilateral or bilateral and different abnormalities may be seen in the same fetus. According to the type and severity of abnormality, the number of nephrons of such fetuses is reduced at various degrees. These abnormalities are the leading cause of renal failure at childhood. Genetic disorders are responsible for some of these abnormalities[6]. In order to better understand the etiology and pathogenesis of renal dysplasia, many efforts have been made to further classify them as syndromic and non-syndromic and to find the gene or genes involved in the development of the disease[7].

Recognition of kidney abnormalities (MCDK with or without contralateral Obstructive Kidney Dysplasia [OKD]) are extremely important especially for cases requiring intrauterine interventions. Fetuses with obstructive uropathies are candidates for such

interventions. Furthermore, antenatal detection permits pregnancy termination, in terms of renal diseases with a worse prognosis. 90% of fetal kidneys may be visualized at 17-20 weeks and 95% up to 22 weeks[8].

The majority of dysplastic kidneys are associated with urinary tract obstruction commencing in early embryonic life. Embryologically, MCDK may result from abnormal renal morphogenesis. MCDK is a form of renal dysplasia, where cystic elements are found in the kidney along with immature, undifferentiated primitive tissue[9]. Bilateral MCDK malformations are frequently associated with Potter anomaly. This fatal anomaly is associated with renal agenesis with oligohydramnios, bilateral pulmonary hypoplasia and may result in still birth or neonatal death[10]. In the present case, unilateral MCDK was noted, most likely to be classical type with atretic ureter. The contralateral dysplastic kidney and hydroureter could be due to obstruction at vesico-ureteric junction (VUJ). Posterior urethral valve was ruled out in our case as it results in oligohydramnios with a distended urinary bladder and bilateral hydronephrosis.

Involvement of other organs is common in patients with MCDK. Esophageal atresia, tracheoesophageal atresia, ventricular septal defect and patent ductus arteriosus are the most common extra-renal abnormalities.

Conclusion

Correct and timely antenatal diagnosis of bilateral kidney abnormalities in the form of dysplastic changes with or without associated fetal anomalies is important so that proper counselling and appropriate obstetric management can be offered. Performing autopsies on such fetuses can result in confirmation of ultrasonographic findings and finding out other associated anomalies or related syndromes.

Compliance with Ethical Standards

Approval of ethical committee was taken for this study and informed consent was obtained.

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PHOTOGRAPH SHEET

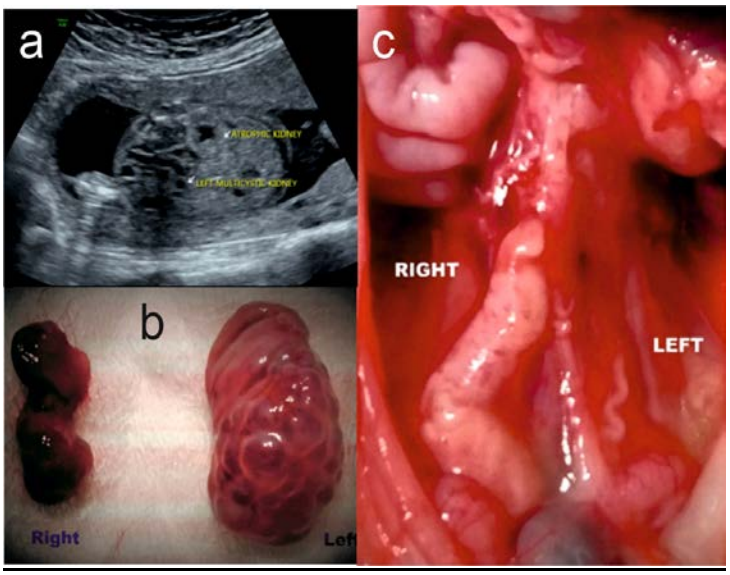

Figure 1 : (a) USG image showing left multicystic kidney and right atrophic kidney in the fetus (b) Gross appearance showing left multicystic kidney and right atrophic kidney.

(c) Autopsy revealing hydroureter on the right side and atretic ureter on the left side

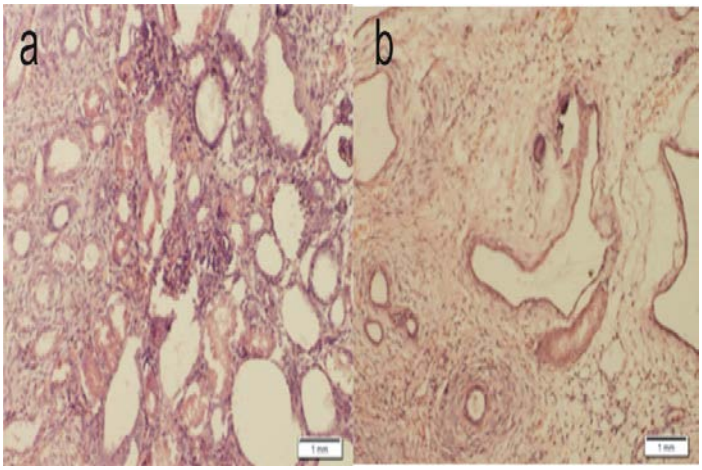

Figure 2. (a). H&E-stained kidney section with 10× lens, original magnification: MCDK

showing multiple cysts and primitive tubules

(b). Dysplastic changes with dilated tubules lined by flattened epithelium