Multicystic Dysplastic Kidneys : Antenatal Diagnosis

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Introduction

High resolution ultrasonography allows demonstration of fetal kidneys and urinary bladder from early second trimester and enables to detect a number of major congenital anomalies of the urinary system. The sonographic demonstration of renal anomalies may modify obstetric management, facilitate paediatric and surgical care of the new born or may allow for elective termination of pregnancy. The incidence of congenital renal malformations on routine antenatal ultrasound is about 0.2% [1]. Multicystic dysplastic kidney disease (MCDKD) is usually an incidental finding during routine antenatal sonographic examination. The incidence of MCDKD is about 1 in 5,000-10,000 births [2]. Two cases of fetal bilateral multicystic kidney disease diagnosed antenatally are described.

Case Report-1

A 19 year old primigravida with history of consanguineous marriage reported for routine antenatal ultrasound during second trimester. Gestational age by LMP was 24 weeks. Biparietal diameter and femoral length corresponded to gestational age of 21-22 weeks. The fetal kidneys were enlarged, each measuring 40 x 24 mm and contained multiple sonolucent cystic structures of variable size ranging from 3-9 mm in diameter (Fig. 1). Urinary bladder was not demonstrable and bowel loops were dilated (Fig. 2). Amniotic fluid was markedly reduced consistent with severe oligohydramnios. Four weeks later she aborted spontaneously. No external abnormality was seen in the aborted fetus.

Case Report - 2

A 25 year old primigravida with unremarkable prenatal history reported for routine antenatal ultrasonography. She had amenorrhoea of 27 weeks duration. Calculated gestational age was 23 weeks. Ultrasonography revealed bilaterally enlarged fetal kidneys each measuring 45 x 26 mm with multiple cysts. Urinary bladder was not demonstrable and the bowel loops were dilated (as in case 1). Pregnancy ended in spontaneous abortion at 26 weeks. The fetus was found to

have facial asymmetry with cleft lip and palate which was missed during antenatal ultrasonography. Post abortion ultrasound revealed enlarged kidneys with multiple non communicating cysts (Fig. 3).

Discussion

The antenatal diagnosis of fetal anomalies has improved largely due to the availability of high resolution ultrasound equipment. Ultrasound is able to demonstrate the fetal bladder shortly after the 11th week of gestation when the kidneys start production of urine and it is possible to document the changes of nephrogenesis which continue until 38 weeks of gestation. The proportion of renal abnormalities diagnosed by antenatal sonography has increased from 15% in 1984 to 80% or more in the recent studies [3-5].

Renal dysplasia is a form of abnormal renal parenchymal development resulting from anomalous differentiation of metanephric tissue during embryogenesis. Most dysplastic kidneys are associated with some form of obstruction of the urinary tract during nephrogenesis. The type and severity of renal dysplasia depends on the pattern of malformation and severity of urinary tract obstruction. Based on these criteria renal dysplasia has been broadly subdivided into four groups. 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 ureterocele with ureteral duplication, Group III : cystic renal dysplasia associated with lower urinary tract obstruction e.g. posterior urethral valves and Group IV : hereditary familial renal dysplasia without any obstructive uropathy [6].

MCDKD is usually an incidental finding during routine antenatal ultrasonographic examination. The entire kidney is usually affected in classical MCDKD 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

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Fig. 1 : Ultrasound showing enlarged right fetal kidney with multiple cysts

Fig. 2 : Ultrasound showing dilated bowel loops

Fig. 3 : Postabortion ultrasound showing enlarged kidneys with multiple cysts.

said to be more common in females with a predilection for occurrence on the right side [7]. MCDKD is characterized by dilation of the collecting tubules. It may be unilateral (76%) or bilateral (24%) [8]. The recognition of bilateral MCDKD by ultrasound has grave prognostic implication as it is a fatal anomaly. Some of the ultrasonographic criteria for the diagnosis of MCDKD include identification of grape like clusters of non communicating cysts in enlarged kidneys (Fig. 3), absence of urine accumulation in the bladder and severe oligohydramnios [9]. There may be associated non renal anomalies like dilation of bowel loops (Fig. 2). Serial antenatal ultrasound examination of MCDKD may reveal dramatic changes in the appearance of affected kidneys. The affected kidneys may enlarge or may actually shrink in size (due to involution of some of the cysts) or rarely may disappear [10]. Both our cases had bilateral severe involvement of kidneys with extreme oligohydramnios. There was also evidence of absence of urinary bladder and dilation of bowel loops (Fig. 2). Both the cases showed growth retardation of 3-4 weeks. The parents were explained about the grave prognosis. Both cases eventually aborted spontaneously within 3-4 weeks of diagnosis.

Cystic kidneys discovered incidentally during routine antenatal ultrasonography frequently pose significant diagnostic dilemma. There are diverse etiologies with equally variable implications for prognosis in the affected fetus. Correct and timely antenatal diagnosis of MCDKD with associated fetal anomalies is important so that

proper counselling and appropriate obstetric and paediatric management can be extended.

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