

CASE REPORT

MUTICYSTIC DYSPLASTIC DISEASE OF KIDNEY IN FETUS

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Multicystic kidney disease remains 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. Survival is associated with higher risk of Wilm's tumor and Renal cell carcinoma. Diagnosis is facilitated by ultrasonography. Respiratory compromise on account of mass effect may need repeated percutaneous cyst aspiration. Nephrectomy is the definitive treatment option.

Keywords: Multicystic; Dysplasia; Kidney; Fetus

INTRODUCTION

Multicystic disease of the kidney, diagnosed on antenatal ultrasound examination, remains the most common of the numerous causes of abnormally enlarged kidney.¹ It typically presents, on ultrasonography, as numerous cysts of various sizes with a reduced parenchyma. Congenital renal dysplasia may be explained by an abnormal induction of metanephric blastoma by the migrating ureteric bud.¹ It has been suggested that displaced metanephric blastoma interspersed with normal zones of nephrogenesis generates the irregular parenchyma of the multicystic kidney. The subsequent cystic dilatation of dysplastic tissue is believed to compress and permanently damage the normal renal tissue. Following are recognized causes of abnormally sized kidneys on antenatal ultrasound examination:²

- Hydronephrosis
- Multicystic dysplastic kidney
- Congenital nephritic syndrome
- Polycystic disease
- Renal tumour
- Damage from obstructive uropathy

CASE REPORT

A female patient, 35 years old, presented at 24 weeks of gestation for a routine antenatal check up. She was G7 P6 A6. On ultrasound examination, a single alive fetus was seen with fetal heart rate of 153 beats / min. Presentation was breech and placenta was posterior and fundal. According to ultrasound examination, period of gestation was 24 weeks and 4 days. Liquor was scanty and congenital anomalies could be seen. On biophysical profile, fetal movements, fetal tone and breathing were normal but the amniotic fluid index was inadequate.

Colour Doppler showed normal flow in the circle of Willis. Number of vessels in placenta was recognized as three, two umbilical veins and one artery. End diastolic flow was present. The resistance index was 0.45. Detailed fetal abdominal

ultrasonography revealed kidneys with hyper echoic shadows and containing small cystic changes, probably with renal parenchymal disease. No flow could be appreciated on Colour Doppler. A large cyst extending to the pelvis was appreciated in the lower abdomen of the fetus. Umbilical cord blood flow was intact. Oligohydramnios was present. Crown Rump length was 23 cm.

Detailed history did not reveal problems during earlier pregnancies. There was no history of congenital anomalies in her or husband's family. The couple was related. The family was distressed and opted for termination of pregnancy. She was induced with prostaglandin F2 alpha vaginally. As the cervical ripening is very little at 24 weeks, it did not respond very well to prostaglandins. It was quite traumatic for the patient, and decision for hysterotomy was taken. A male fetus with grossly enlarged abdomen was delivered. (Figure)



On gross examination, the fetus had an enlarged abdomen. Numerous distended vessels were seen on anterior abdominal wall. The abdominal wall was very tense and liver and kidneys could not be palpated. Both upper and lower limbs were normal and there were no amniotic bands. The spine was normal. The fetus had a pink colour to it and there were no facial deformities. Head circumference was 43cm. Abdominal girth was 55cm. Placenta weighed

0.5 kg and was examined for abnormalities. It was covered with amnion on fetal side and on the maternal side cotyledons were normal. There were no areas of calcification or infarctions. The fetus did not show any signs of intra uterine growth retardation. The fetus lived for ten minutes after delivery. The abdomen was opened. On gross examination, liver and intestines were normal looking. The kidneys were grossly dilated and cystic. Upon opening fetal kidneys, the kidneys showed multiple cysts of various sizes ranging from 3mm to 7cm. The cyst wall was not very thick and cysts were translucent white and contained clear white fluid. Normal renal anatomy was completely distorted. The cysts completely occupied the medullary region and cortex was pushed towards periphery and formed only a narrow rim surrounding the medulla.

DISCUSSION

Multi cystic disease of the kidney is a common cause of enlarged abdomen in fetus second only to hydronephrosis. Its prevalence is 1 in 2400 births, male fetus being affected in more than two thirds of cases.⁵

It is a congenital maldevelopment, where the renal cortex is replaced by numerous cysts of varying sizes. Dysplastic parenchyma anchors the cysts and the arrangement resembles branches of grapes. Calyceal drainage system is absent. It is typically a unilateral disorder; bilateral condition is incompatible with extra uterine life.

Affected children are still born or die in early post natal period in case of bilateral disease. In unilateral disease, the affected kidney involutes or decreases in size in 60-70% cases.³ Furthermore, there is an increased risk of Wilm's tumor and renal cell carcinoma, in children who survive.⁴ Oligohydramnios occurring before 24 weeks is characteristic.³ It also profoundly affects the fetal lung development; critical transition from the canalicular to alveolar phase.³ Ultrasound can detect the cysts as early as 12- 13 weeks of gestation.⁵

During normal fetal growth, transient dilatation of fetus urinary tract is normal, occurring in 1 out of every 100 fetuses. Measurement of anterior posterior pelvic diameter and its ratio to overall anterior posterior renal diameter have been proposed as criteria for discriminating between normal and abnormal. In fetus younger than 24 weeks a pelvic diameter greater than 10 mm was 76% sensitive in identifying an obstruction.⁶

Antenatal diagnosis is facilitated by ultrasonography that gives a characteristic picture at 24-28 weeks.⁵ Ultra sound shows hypo-echoic cysts of variable sizes and shapes, interfaces between cysts, absence of identifiable renal sinus and minimal surrounding parenchyma.⁵ Ultrasound picture of multiple cystic disease of kidney is an excellent method of diagnosing the condition with a high degree of confidence.⁵ Autosomal recessive Polycystic kidney may not be mistaken for multicystic disease of the kidney, cysts in the former being small and parenchyma is generally hyper-echoic. Other cystic diseases, typically appear with some functional parenchyma.⁵

Bilateral multicystic disease is incompatible with extra uterine life.

Regarding treatment, a newborn child with unilateral disease may need percutaneous cyst aspiration in case of compromised respiratory function, due to mass effect. Follow-up with ultrasound examination, every three months, for the first year of life, is recommended. Nephrectomy may be an option later on.⁷

REFERENCES

1. Sadler TW, editor. Langman's Embryology, 7th edition, Baltimore, Maryland USA: Williams & Wilkins; 1995.
2. Smith NC, Hau C. A six year study of the antenatal detection of fetal abnormality in six Scottish health boards. Br J Obstet Gynaecol 1999; 106:206-12
3. Sukthankar S, Watson AR. Unilateral multicystic dysplastic kidney disease: defining the natural history. Acta Paediatr 2000;89(7): 811-3.
4. Wiener JS. Multicystic Dysplastic Kidney. In: Bellman AB, King, LR Kramer SA. Clinical Pediatric Urology 4th edition, London: Taylor and Francis; 2002. p. 633-45
5. Irsutti M, Puget C, Duga I, Sarramon MF, Guitard J. Mesoblastic Nephroma: prenatal ultrasonographic and MRI features. Pediatr Radiol 2000;30:147-50
6. Peters CA, Carr MC, Lais A, Retik AB, Mandell J. The response of fetal kidney to obstruction. J Urol 1992;148: 503-9
7. Belk RA, Thomas DF, Mueller RF, Godbole P, Markham AF, Weston MJ. A family study and the natural history of prenatally detected unilateral multicystic dysplastic kidney. J Urol 2002;167(2 Pt 1): 666-9.

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