Research Paper Medical Sciences

Muticystic Kidney - Dilemma

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ABSTRACT

Background : Development of kidneytakes various stages from pronephros to metanephros and metenephros in intermediate mesoderm. Various stages indicates increasing complexity as need arises through evolution. Ureteric bud develops and migrates in metanephric blastima where secretary portion joins to ureteric bud to form nephron and conducting duct. Developmentally joining between them may go wrong giving rise to cyst formation. Still precise cause it not known. Once developed it has to migrate from pelvic region to lumbar region with changing blood supply. Here we are to discuss cystic kidney diagnosed on ultrasonograpy with varied postnatal outcome. One on cadaver and two delivered normally with different late outcome. Different category of terminology are utilised such as multicystic kidney, polycystic kidney and congenital hydronephrosis then unilateral and bilateral. It is attempt to conscious wait and watch and increasing difficulty to decide and council parents.

KEYWORDS Metanephric blastema, MCDK, Wnt Family, pax-2.

Introduction:
Kidney is a vital organ for not only excretion but maintain osmolality, Ph. of blood and blood pressure as well water and electrolyte balance. It also act as site for secretion of erythropoietin.

For its role as simple excretory function to more complex function of water and electrolyte regulation kidney develops through three stages in intermediate mesoderm at various positions. Second it ascends from iliac fossa to final lumbar region.

Definitive kidney develops from epithelial outgrowth i.e. ureteric bud and metanephric blasema both from same mesoderm. They induce each other so as to form secretary portion (nephron) and collecting system (minor calyx till ureter).

At 22nd day transcription factor Pax-2 is expressed caudal to pronephros, which induces Li-1 in the intermediate mesoderm. It leads to formation of mesonephric duct. By the end of 4th week of gestation the mesonephric duct attaches cloaca. WT-1gene near cloaca on posterior aspect of mesonephric duct develops uretric bud[1].

Outgrowth of the ureteric bud from the mesonephric duct is a response to the secretion of glial cell line derived neurotrophic factor (GDNF). The formation of GDNF in the metanephric mesenchyme is regulated by WT-1. Sprouty represses action of GDNFon anterior mesonehric region so as to form uretric bud anteriorly[1].

FGF-2 and BMP7 and Wnt- 11 plays role in branching pattern of tubular system. Uretric bud develops into 14 to 15 branches[1].

Glomerular end levels of Pax-2 expression decrease as WT-1 becomes strongly . At other end at future distal convoluted tubule Wnt-4 and E- cadherin remains prominent and at proximal convoluted tubule K-cadherin is a prominent cellular marker[1].

Case Report:
Patient A; 24 years old, Female pateint with H/o of primigravida with 4 years of infertility for antenatal ultrasonography with gestational age 29 weeks. Ultrasonography showed single live intrauterine pregnancy of 29 weeks 3 days with slightly less liquor with e/o unilateral multicystic dysplastic kidney on left side with small crystic lesions randomly distributed less than 5 mm. Right kidney normal.

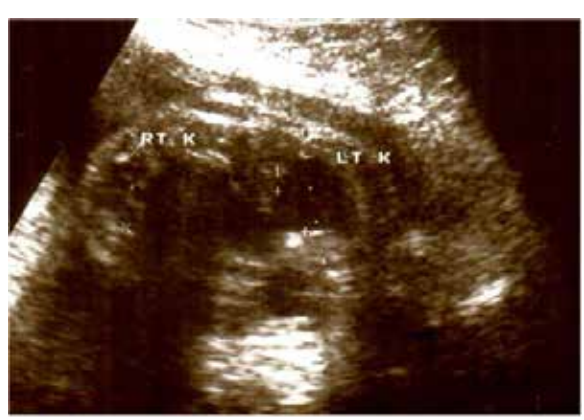

Figure 1: Left multicystic kidney with right normal

Patient B; Same time second patient with ultrasonography of 28 years old patient with ultrasonography at 28 weeks showed Right hypoplastic kidney withLeft kidney lobulated enlarged with ? normal echotexture.

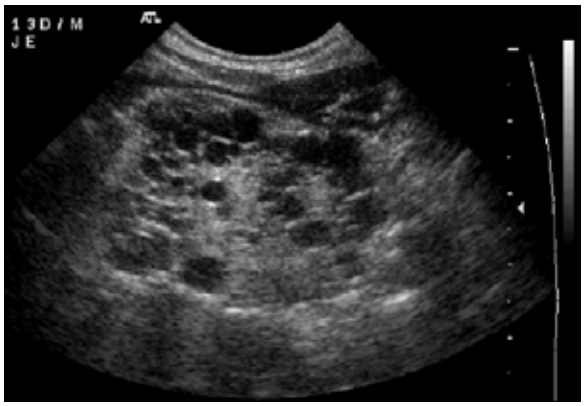

Figure 2: Left polycystic kidney with Right hypoplastic

Patient C; Third patient 26 years old with 25 weeks of gestation of G3A2L0. Ultrasonography showed 25 weeks 4 days with right kidney agenesis and Left kidney enlarged showing cystic mass more than 6 mm. Patient went into preterm delivery and fetus survived for few minutes. In department of anatomy during dissection Left kidney was grossly enlarged with 5 x 4 cm. Multicystic kidney with ranging from 3mm to 15mm. Also had hypoplastic lung and cardiomegaly was seen.

Figure 3: polycystic kidney dissected Pateint C

While follow up of patient A and B dilemma was how to council and follow up during pregnancy and post natal.

A renal cyst is defined as an enclosed sac or nephron segment lined by epithelial cells dilated to more than 200µm. A cystic kidney is having three or more cysts present. Multiple dysplastic kidney (MCDK) is the second most common disease causes of a flank mass in newborn. MSDK was first described by Schwartz in 7 month old child in 1929 as a 'Bunch of grapes' replacing the kidney [2].

MCDK is typically unilateral but bilateral MCDK's have been reported in stillborn infants with oligohydramnios. In unilateral MCDK the contralateral kidney is normal but at high risk for other abnormalities [3].[4].

MCDK often presents as an asymptomatic abdominal mass in the neonate. However, currently approximately 85% to 100% of MCDK are detected by prenatal sonographic examination. Among 64 fetus diagnosed prenatally with MCDK at the Hospital for Sick children in London, 33% were diagnosed at 18 to 20 weeks of gestation, 44% between 20 to 30 weeks and 33% 30 to 40 weeks of gestation [5][6].

The pathogenesis of this disorder is poorly understood but it is thought to involve failure of the uretric but to integrate and branch appropriately into the metanephrous during the development of the kidney[7].

The incidence of bilateral MCDK is estimated at 1 : 10000 live births. The male to female ratio is 2:1in contralateral MCDK [8] [9].

The sonographic features of MCDK include a cystic paraspinal flank mass. The cysts are usually of various sizes are distributed along the periphery and absence of normal renal sinus echoes and renal parenchyma[10].

The cyst may regress or enlarge and then subsequently regress in size. Once the condition is identified the kidney opposite to the MCDK becomes of paramount importance because of the 40% incidence of contralateral anomalies [11].

Preservation of normal amniotic fluid volume suggests normal renal function in the contralateral kidney. However oligohydramnios and absence of bladder filling suggest a lethal fetal renal disease, which occurs in 30% of fetuses with MCDK [12].

The main differential diagnosis of MCDK includes ureteropelvic junction obstruction, adult or infantile forms of polycystic kidney disease, trisomy 13 and Mecket – Gruber syndrome [13] .

The sonographic detection of renal parenchymal cysts that communicate with the renal pelvic suggest the diagnosis of ureteropelvic junction obstruction [14]

In adult polycystic kidney disease the cysts are randomly distributed in contrast to be at the periphery [15]

The Meckel – Gruber syndrome the cysts are small uniform and scattered throughout the kidney. Infentile polycystic kidney disease are very large and echogenic and discrete cyst not visible. In trisomy 13, the kidney is bright echogenic and has small cyst scattered in the renal parenchyma. He observed complete involution in 33% during the first 2 years, 47% at 5 years and 59 % at 10 years. [16]

In unilateral MCuDK with normal contralateral kidney an excellent prognosis is anticipated and no special care at time of delivery. However if oligohydramnios or a contralateral renal abnormality is present then newborn resuscitation may be complicated by pulmonary hypoplasia. Postnatal studies will be 1) Renal ultrasound examination 2) Diuretic renal scan with cystourethrography 3) serum creatinine and blood urea nitrogen level will begin at 36 to 48 hours 4) the infant should be treated with antibiotics till vesicourethral reflex is excluded. It has been reported that spontaneous total involution occurs in 67% cases [17]

Prenatal involution was observed in 6 cases (5%) and there were an addition 16 cases (17.5%) of involution postnatally. MCDK less than 5mm in length will get smaller and either disappear or remain unchanged [181920]

Those with MCDK size greater than 6mm often do not involute. There was a high incidence of associated genitourinary abnormalities. Uteropelvic junction obstruction and megaureter were the most common abnormalities affecting the contralateral kidney. Hypertension develops and resolves with nephrectomy in all instances [2122]

Discussion:

Patient C showed bilateral involvement with cyst on left side larger than 6mm and Right kidney agenesis. It has been associated with hypoplastic lung as it correlates with findings with Rabelo et al, Snodrass and Narchi. [17, 21-22].

Patient A and B delivered full term normal delivery. Both required minimum resuscitation as observed by Diana W. Biachi and Rabelo. [12,17].

Pateint A's neonate went through ultrasongraphy after birth shows similar finding. Biochemical markers where normal throughout two years. But ultrasound changes with reduce size of cyst seen at 6th month and totally regressed at one year

as consistent with Aslam et al and Gordan et al and Gaugh et al. [16-19]. Baby is having normal milestones.

Pateint B's neonate went through ultrasongraphy after birth shows similar finding. Biochemical markers slowly deranged intwo years. Cysts show no regression on left side. Hypertension prevailed. Milestones are retarded. With poor prognosis [16-21].

Conclusion:

Early diagnosis of cystic or dilated kidney on ultrasonography makes critical decision about management of pregnancy and concealing patient.

As it is difficult to decide about prognosis as still no standard markers not available rather it is still unclear what goes wrong so abnormality in development.

Certain guidelines can be used as present findings correlate with few previous study To rule out urteropelvic junction obstruction [14, 17].

Bilateral involvement either agenesis or bilateral cysts with oligohyramnios has poor prognosis [12,17].

Unilateral cysts less than 5mm and normal biochemical markers has better prognosis and routine follow up can help [18-20].

Cysts larger then 6mm with reduced liquor may indicates more followup and indicator of poor prognosis [21-22].

As advances in genetic marker and better understanding of developmetal genetics will make diagnosis and concealing better. [1].

Isolated MCDKs are usually sporadic with no risk of recurrence in subsequent pregnancies. However any family history with renal agenesis to congenital hydronephrosis risk increases by 50% [12].

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