



Cystic Renal Disease -- Multicystic Dysplastic Kidney

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ABSTRACT

Multicystic dysplastic kidney is the most common type of renal cystic disease, one of the most common cause of an abdominal mass in infants. A multicystic dysplastic kidney contains non-communicating multiple cystic spaces varying in size and has no functioning renal tissue. there is a possibly results that it from abnormal metanephros differentiation, probably due to disturbed connection of ureteric bud with renal blastema and abnormal division at the stage of metanephros. Contralateral renal and other concomitant abnormalities are important for the prognosis of children with MCDK. We reporting 2 cases.

Keywords : Multicystic dysplastic kidney [MCDK]

Introduction :-

Incidence of unilateral multicystic dysplastic kidney is reported to be 1 in 4300 live births, and the combined incidence of unilateral and bilateral multicystic dysplastic kidney is 1 in 3600 live births. Bilateral multicystic dysplastic kidney occurs in about 20% of prenatally diagnosed cases of multicystic dysplastic kidney. The left kidney is involved in 55% of cases, and the right kidney is involved in 45%. Infants .Multicystic dysplastic kidney is a form of renal dysplasia 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 non functional [1]. Several forms of MCDK have been described . The classic type [simple] and the less common [complex] hydronephrotic type and a third, least common form known as solid cystic dysplasia [2]. We are reporting 2 such cases.

SIMPLE MCDK [case 1]:- A full term male child[Fig:1] , birth weight 2.2 kg , admitted on day of life 1 with respiratory difficulty ,baby had palpable Abdominal mass in left hypochondrium, iliac & lumbar region. Antenatal records were unavailable. Renal function test were abnormal [Urea 64,creatinine2.1] . X-RAY abdomen flat plate was suggestive of mass in left side[Fig:2]. USG abdomen [Fig:3] was performed which showed Grossly enlarged[97 mm] left kidney with variable sized cystic lesions involving whole kidney.right kidney also had multiple tiny cysts .



Fig:1



Fig:2



Fig:3

COMPLEX MCDK[case 2] :-A female child, birth weight 3.kg,Delivered in hospital admitted on day of life 7 with complain of not passed urine for last 4 days. Baby passed urine First time after 3-4 hrs of birth. Mother was first gravida ,antenatal USG at 28 wks &34 wks showed severe oligohydramnios, bilaterally cystic kidneys with dilated ureters. There was no obvious mass on abdominal examination.Kidney function were abnormal [urea 82,creatinine 6.0]. USG abdomen [Fig:4,5] was performed which showed bilateral multi cystic dysplastic kidney[Right>left] with bilateral dilated ureters with vesico ureteric reflux & hydronephrosis.



Fig:4,Left kidney



Fig:5,Right kidney

DISCUSSION:-

We found 2 cases of multicystic dysplastic kidney in our set-up. Both cases presented with different clinical scenario.

In Simple MCDK there is unilateral dysplasia with normal contralateral kidney with compensatory hypertrophy and no associated genitourinary abnormalities [3]. In contrary to this in our case no 1 contralateral kidney also had multiple tiny cysts. Complex MCDK is defined as bilateral dysplasia or abnormalities of contralateral kidney with hydronephrosis [3] as found in our case no 2. Incidence of contralateral VUR is higher in patients those having MCDK [4]. Case no 1 showed only contralateral VUR while case no 2 also had VUR and bilaterally dilated ureters. Right sided ureter was tortuous in whole length due to VUR. MCDK usually occurs as a sporadic event but, rarely autosomal dominant inheritance and sib recurrence has been reported [5]. In Our both cases parental screening was done which showed no abnormality in there kidney anatomy and both cases were first child of there parents. Ultrasonography is preferred first investigation for MCDK and antenatal diagnosis is possible by USG [6]. Case no 2 was diagnosed antenatally while for case no 1 no antenatal record was available.

Contralateral renal and other concomitant abnormalities are important for the prognosis of children with MCDK [7]. MCDK is not treatable. Most MCDKs become smaller and involutes during a period of follow-up. However, the patient is observed periodically for the first few years during which ultrasounds are generally taken to ensure the healthy kidney is functioning properly and that the unhealthy kidney is not causing adverse effects. In some cases like renal hypertension or malignant transformation, the unhealthy kidney is removed entirely [8].

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