Case Report

Bilateral Multicystic Dysplastic Kidneys in Association with High Ano-Rectal Anomaly - A Case Report

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Abstract:

Multicystic dysplastic kidney disorder (MCDK) is commonly sporadic but is rarely an inherited congenital disease. 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 bilateral multicystic dysplastic kidney associated with high ano-rectal anomaly which was diagnosed antenatally and pregnancy was terminated.

Key Words: Ano-rectal anomaly, Bilateral, Multicystic dysplastic kidney

Introduction:

Renal dysplasia is a developmental disorder and is categorized as either hypoplastic dysplasia, multicystic dysplasia, agenesis, segmental agenesis or congenital hydronephrosis with or without dysplasia. Multicystic dysplastic kidney (MCDK) is the most common dysplasia variant. [1] It is typically a unilateral disorder.[2] Involvement of other organs is likewise common in patients with MCDK. Esophageal atresia, tracheoesophageal atresia, ventricular septal defect, and patent ductus arteriosus are the most common extrarenal abnormalities.[3] The widespread use of prenatal sonography has led to early discovery of multicystic kidneys.[4] In the present case report, a rare occurrence of bilateral MCDK with associated ano-rectal anomaly is discussed.

Case Report:

A 23 year old female presented at 26 weeks of gestation for a routine antenatal check up. She was G1 P0. Prenatal ultrasonography showed small for date fetus with severe oligohydramnios and multiple congenital anomalies (absent stomach bubble, bilateral multi cystic kidneys, megabladder, dilated bowels). The couple had non consanguineous marriage with no history of congenital anomalies in her or husband's family. The family was distressed and opted for termination of pregnancy. A still born male fetus weighing 600gms was delivered vaginally after induction with Mifepristone and Dinoprost gel. The fetus was sent in formalin to the pathology department.

The fetus in formalin was received in Pathology department after 2 days of delivery. The fetus was showing signs of maceration and was small for gestational age (by anthropometry). The fetus had low frontal hair line, prominent epicanthal folds, bilateral low set and posteriorly rotated ears, flattened nasal tip and micrognathia (Fig. IA). The testes were undescended and the urethral meatus was patent. There was only a dimple present at the site of anus (Imperforate anus; Fig. IB). The rest of the external examination was normal.

As per the parent's wish, the autopsy was immediately carried out and fetus was returned for ritual and final disposal.

The internal examination revealed an enlarged bladder, bilateral multicystic kidneys and dilated rectum (Fig IC).

The kidneys were asymmetric (right kidney was larger than left) with maintenance of reniform shape and presence of multiple cysts on the surface (Fig. IC). Cut section showed presence of multiple cysts with loss of corticomedullary demarcation. Histopathological examination showed features of bilateral multicystic dysplastic kidneys (Fig II).

The left ureter was hypoplastic. The right ureter in the distal one third was dilated (Hydroureter). The bladder showed marked hypertrophy with absence of posterior urethral valves.

There was rectal agenesis with dilatation of

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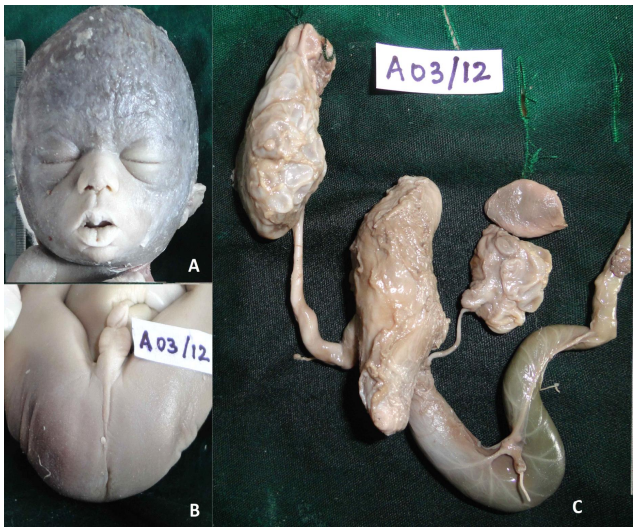

Fig I: (A) Shows dysmorphic facies. (B) Shows imperforate anus. (C) Shows bilateral multicystic kidneys with hypoplastic left ureter, megabladder and dilated rectum.

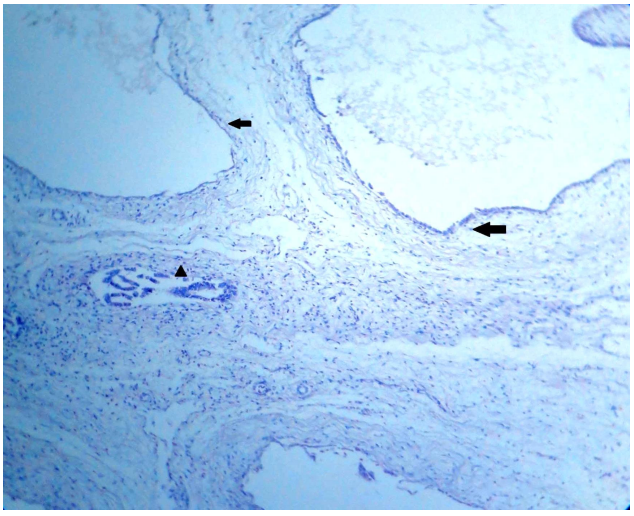

Fig II: Histopathological picture of MCDK showing multiple cysts lined by flattened epithelium with (⬅) few primitive tubules (▲; H & E, 40X).

proximal rectum which ended as a blind pouch and was attached by a thin strand to the posterior wall of bladder (Fig. IC).

The left lung was hypoplastic with a single lobe and a partial fissure. Microscopically, features of hypoplasia could not be determined because of autolysis. The spleen and adrenals showed autolysis with presence of cysts on microscopy. The other organs were normal.

In view of above anomalies, parenteral karyotyping was advised.

Discussion:

Multicystic dysplastic kidney is a congenital renal malformation characterized by the presence of multiple cysts in dysplastic kidneys. It is a form of renal dysplasia, where cystic elements are found in the kidney along with immature, undifferentiated primitive tissue. Embryologically, MCDK may result from abnormal renal morphogenesis.[1] Multicystic disease of the kidney is typically a unilateral disorder.[2] Bilateral MCDR malformations are frequently associated with the fatal Potter anomaly.[1] The sequence, as described by Potter in association with renal agenesis with oligohydramnios, bilateral pulmonary hypoplasia and immature lungs microscopically may result in still birth and neonatal death.[5] Some of the features of Potter sequence found in the present case were oligohydramnios, dysmorphic facies (prominent epicanthal folds, flattened nasal tip and low set and posteriorly rotated ears) and grossly hypoplastic lungs. Bilateral MCDK was found in the present study. Similar findings were reported by various authors.[6-10] The majority of dysplastic kidneys are associated with urinary tract obstruction commencing in early embryonic life.[11] The type and severity of renal dysplasia depends on the pattern of malformation and severity of urinary tract obstruction.[9] In the present case, the fetus had hypertrophied bladder in the absence of posterior uretheral valves, which implies some obstruction in the lower urethra, which were not included during en-bloc removal of organs during autopsy. This obstruction might be the cause for bilateral MCDK. Similar finding of bilateral MCDK with urinary tract obstruction was also found in 2 cases.[9]

Involvement of other organs is likewise common in patients with MCDK. Esophageal atresia, tracheoesophageal atresia, ventricular septal defect, and patent ductus arteriosus are the most common extrarenal abnormalities.[3] In the present case associated rectal agenesis and imperforate anus was found. Similar such association (MCDK with dysplastic empty scrotum and anal atresia) was also found in one case study.[7] In a study of unusual associations with anorectal malformations in 284 children, one case was found to have MCDK.[12]

Correct and timely antenatal diagnosis of bilateral MCDK with associated fetal anomalies is important so that proper counselling and appropriate obstetric management can be extended. Antenatal diagnosis is facilitated by ultrasonography that gives a characteristic picture at 24-28 weeks.[2] In the present case, antenatal diagnosis of bilateral multicystic disease was made at 26 weeks and pregnancy was terminated.

Conclusion:

Routine antenatal scans are done to detect the anomalies in fetus, some of which may be incompatible with life. Performing autopsies on such fetuses can result in confirmation of ultrasonographic findings and finding out other associated anomalies or related syndromes. This can aid the obstetrician in planning for appropriate management.

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Conflict of Interest: None declared.

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