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Case Report

Radiology Section

Prenatal Ultrasonographic Diagnosis of Posterior Urethral Valve: A Case Report

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ABSTRACT

A case report on prenatal ultrasonographic diagnosis of posterior urethral valve.

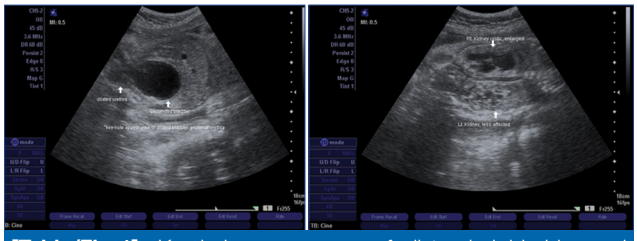
Several anomalies of lower urinary tract leading to obstructive uropathy have been demonstrated in literatures either isolated of urethra or in combination with other syndromes like Prune-belly syndrome or anorectal malformation.

In this present case report, we would like to elucidate the prenatal ultrasonographic features of posterior urethral valve (PUV) and formulate a brief discussion on the treatment option including prognosis of such a case. This case was detected incidentally by routine ultrasonographic examination of foeto-placental profile to monitor growth of the foetus. Mother was counselled for prenatal treatment but she opted for immediate postnatal treatment. Foetal surgery of such type of cases has not been reported much. It is associated with high risk of preterm delivery. Here we have diagnosed it in prenatal period and managed this case after delivery at 34 weeks of gestation.

Keywords: Bilateral hydrouretero-nephrosis, Bladder outlet obstruction, Congenital urethral obstruction, Prenatal kidney damage

CASE REPORT

A 30-year-old G2L1 was booked at our antenatal service at 11 weeks of gestation. She had a normal child of a consanguineous marriage. Her anomaly scan at 20 weeks of gestation was normal. At 28 weeks of gestation she was diagnosed to have Gestational Diabetes Mellitus and admitted in the hospital for the initiation of Insulin. Scan was repeated after admission which showed a distended bladder of foetus with dilated proximal urethra [Table/Fig-1] and bilateral hydroureter & hydronephrosis more prominent on right kidney [Table/Fig-2]. It was found to be at 28 weeks of gestation and normal amniotic fluid index for the gestation. Repeated on the same day and on the next day revealed the same findings. No other system anomalies were identified. PUV was suspected when urinary bladder of the baby was seen persistently distended on repeated ultrasonography at 3rd trimester; however, it was not noted during single anomaly scan at 20th week of pregnancy.



[Table/Fig-1]: Key-hole appearance of distended bladder and dilated proximal urethra due to obstruction at urethra.
[Table/Fig-2]: kidneys show cystic dysplastic changes, though it was more marked on the right side.
NB: right kidney was found non- functional on renogram after PUV fulguration procedure in the neonate

Patient was informed about the problem and a follow-up was planned after discussion with another hospital having facility of neonatal surgery. She was referred to that hospital at 34weeks for delivery as there was increasing bilateral hydronephrosis, (more on the right side) and decreasing amniotic fluid index.

But the patient did not go there for her personal reason.

She was admitted in our hospital at 37weeks when she came for routine checkup and elective LSCS was done at earliest possible time.

After delivery, male baby was found to have distension of lower abdomen with bladder and poor stream of urine. Newborn was referred to a center with paediatric surgery unit, after one week of delivery as earlier shifting could not be arranged by the patient. At that hospital, renal biochemical profile of the newborn was within normal limit. Neonatal ultrasound showed bilateral hydronephrosis and thickened bladder. The Micturating Cysto-Urethrogram (MCU) showed thickened and trabeculated bladder with dilated and elongated posterior urethra. There was grade-II reflux into right ureters and kidney

but no reflux was noted on the left side. A cystourethroscopy was performed and a posterior urethral valve was noted which was fulgurated at the same time. Newborn had uneventful post-op and catheter drainage for 4 days. After the fulguration procedure,Tc99m-DTPA renogram (F+0) was performed for evaluation of renal function and drainage of collecting system which showed nonfunctioning right kidney and hydroureteronephrotic left kidney with good cortical function. Since then, the patient has been under regular follow-up in paediatric unit of that hospital to follow-up the renal function.

DISCUSSION

There is a wide range of anomalies which can cause lower urinary tract obstruction. Numerous anomalies which have been described in literatures are posterior urethral valve, urethral stricture, urethral atresia, urethral membrane, urethral polyp, mullerian duct remnants like enlarged prostatic utricle and mullerian duct cyst, anterior urethral valve and diverticula, megalourethra, urethral duplication and urethroperineal fistula and in combination with other disorders like prune-belly syndrome and anorectal malformation. Among all these anomalies of the urethra, the posterior urethral valve is the commonest one [1,2]. Many of these conditions are difficult to differentiate by antenatal ultrasonography as sonographic features of lower urinary tract obstruction may be pretty same in many cases.

Classically the diagnosis of PUV consists of distended foetal bladder and a dilated proximal urethra (key-hole appearance) in a male foetus. Depending upon the severity and time of obstruction there will be development of other sequel of obstruction such as oligohydramnios with its effect on pulmonary development and other structural compressive deformities and back pressure effect on ureters and kidneys such as hydroureter and hydronephrosis with enlarged kidneys.

If the obstruction is for long time and severe it may result in dysplastic or fibrotic and small kidneys [3]. Presence of oligohydramnios and dysplastic small kidneys are extremely poor prognostic signs which indicate that the effect of obstruction is severe and has already damaged the kidneys [4]. Oligohydramnios if develops early in the second trimester carries a poor prognosis as this will result in pulmonary hypoplasia which is lethal. Hence the timing of diagnosis and appropriate intervention to mitigate the back pressure effect on kidneys and development of oligohydramnios is very important.

Diagnosis of presence of associated other anomalies as seen in Prune Belly Syndrome is extremely important as the prognosis is different when it is an isolated anomaly only. Presence of chromosomal anomaly will alter the whole prognosis. So every attempt has to be made to verify whether any other anomaly which is incompatible with life is there or not. Risk of renal failure of baby born with PUV and detected immediately after birth is more than those detected later in life [5].Once diagnosis is made it is extremely important to counsel the mother and her partner about the possible intervention and outcome. Interventions include termination of pregnancy, foetal surgical therapy, early delivery and possible neonatal interventions.

Foetal therapy for urethral obstruction has been a controversial subject of intense interest for more than a decade. If the urethral obstruction is associated with other anomalies such as Prune-Belly syndrome, severe oligohydramnios with pulmonary hypoplasia and severe kidney dysplasia diagnosed in early second trimester, anorectal malformation and chromosomal anomalies etc, it is recommended to counsel the parents and terminate the pregnancy. Although much has been learned over the last decade, major questions remain about patient selection criteria, timing of intervention and surgical techniques. As with most urinary tract anomalies, the presence or absence and duration of oligohydraminos are critical factors in the foetal prognosis.

Mahoney and colleague evaluated 40 bladder outlet obstructions to determine the important prognostic prenatal ultrasound features.

More than half of survivors had a normal amniotic fluid volume, whereas only seven percent survived with oligohydramnios. In contrast, 80% of those suffering a subsequent demise, had decreased amniotic fluid, whereas only 12% had a normal volume [6]. Hence follow up with ultrasound and simple measurement of amniotic fluid index is so important to determine the course and time of intervention. In the present case, it was found that the amniotic fluid index was reducing and termination of pregnancy at 34 weeks after administering antenatal steroid was advised though patient could not make it possible.

Presence of renal cortical cyst has a very good specificity and sensitivity of renal dysplastic changes [7]. Increased renal echogenicity is also a prognostic indicator as it indicates renal dysplastic changes. Fresh foetal urine obtained by vesicocentesis and measurement of various parameters like sodium, chloride, calcium, microglobulin and measurement of urine output rate by ultrasound after a vesicocentesis have been used by several investigators for foetal prognostic signs [8].

One of the intrauterine foetal surgical interventions is percutaneous placement of catheter, one end into the foetal bladder and the other end into the amniotic cavity outside the foetal abdomen.

But this is only possible in a centre with all facilities and ongoing experience with proper selection of cases after ruling out other anomalies including chromosomal problem of the foetus and

explaining the procedure related complications. Crombleholme and colleagues reviewed an uncontrolled experience of 40 cases, many of whom underwent shunt procedures. In the group with normal appearing kidneys and normal amniotic fluid, there was 90% survival rates among those treated surgically. However, the nonintervention subgroup had a 70% survival rate with the same favourable criteria. In those with dysplasia, there were no survivors without intervention and this group had a 30% survival rate with shunting [9].

But in most of the situation, when intrauterine treatment is impossible, then early delivery may be of some benefit to shorten the effect of oligohydramnios and the effect of back pressure on the kidneys and ureters. Ablation of posterior urethral valve can be offered as a primary treatment option for all PUV cases detected after birth [10]. Risk of renal failure in patient with PUV depends upon the gestational age at diagnosis and amniotic fluid volume [11]. Hence antenatal steroid and early delivery and post-delivery simple cystoscopic fulguration may be helpful in such a case.

CONCLUSION

Though in the present case, baby with one nonfunctioning kidney, the other kidney was functioning well and the baby is doing well with one functioning kidney. As noted in postnatal MCU that there was no reflux on the left side which perhaps has saved the left kidney and right kidney lost the functionality as more insult of back pressure was on it alone. Long term follow up in childhood is necessary as there may be later manifestation of the reduced function of the kidney. Hence, the patient is on regular follow up.

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