Antenatally Diagnosed Posterior Urethral Valves : A Dilemma

Surg Cdr R Panicker*, Lt Col DS Grewal†

MJAFI 2010; 66 : 167-169

Key Words : Antenatal; Urinary obstruction; Vesicocentesis

Introduction

Posterior urethral valves are the commonest cause of lower urinary tract obstruction (LUTO) diagnosed in the male fetus in the antenatal and immediate postnatal period. The estimated incidence of this condition was thought to be 1 in 10,000 deliveries. However more recent studies have suggested that the incidence could be higher at about 1 in 2500 – 5000 [1] and this appears to be due to the widespread use of high resolution antenatal ultrasonography.

Case Report

A 27 year old lady in a non consanguineous marriage, presented in her fifth month of pregnancy to the out patient department (OPD) for a routine checkup. She was a second gravida with a previous history of a first trimester missed abortion two years ago. By her dates the lady was in her 18th week of pregnancy. She was essentially asymptomatic as her early pregnancy symptoms had subsided. She did not give any history in the first trimester of bleeding per vaginum, febrile illness or drug intake. Her first trimester ultrasound report was normal and the calculated gestational age corresponded to the period of amenorrhoea. All her routine antenatal investigations were normal.

On examination, her general physical examination was within normal limits. Obstetrical examination revealed that the uterine fundal height corresponded to 16-18 weeks gestation. A second trimester ultrasound was requisitioned and the findings showed a single live fetus of gestational age 18 weeks $\pm$ 1 week. There was significant oligohydramnios and the amniotic fluid index was calculated as 5 cm (normal range at this gestation is 9-20 cm). The fetal urinary bladder was grossly overdistended and revealed the "Keyhole" sign (Fig.1). The bladder neck to fundal height was 74 mm (normal range is upto 35 mm). Both the fetal ureters were dilated and tortuous. Both kidneys showed increased cortical echogenicity, fullness of the central pelvis and hydronephrosis. One fetal foot revealed features of talipes

equino varus deformity.

Since the couple were extremely anxious to continue with the pregnancy, the lady was subjected to a fetal vesicocentesis. The sodium content and osmolality of the fetal urine were evaluated. The sodium level in the fetal urine was 145 meq/l and the osmolality was 228 mosm/l.

The couple were called for counselling and were explained the poor prognosis in view of the fetal kidney involvement. They were also explained the pros and cons of vesicoamniotic shunting. The couple, after debating all aspects of the problem and consulting with paediatric surgeons and paediatric nephrologists, decided to go in for a medical termination of pregnancy (MTP). A second trimester MTP was hence carried out on eugenic grounds using vaginal misoprostol.

The fetus on examination showed a grossly distended urinary bladder (Fig. 2). The male external genitalia were normally developed. A fine venous cannula could be passed through the meatus but could not be passed into the urinary bladder due to an obstruction at the level of the urethrovessical junction.

Discussion

The in utero development of the renal system is initiated in the 5th week with the formation of the metanephros and the mesonephric ducts. The metanephros differentiates into the fetal kidneys and this process is completed by the 10th week, when urine starts flowing from the kidneys. Simultaneously the lower urinary tract also develops between the 5th – 10th weeks. The terminal end of the hind gut differentiates posteriorly to form the cloaca and anteriorly into the urogenital sinus. The urinary bladder starts developing from the pelvic part of this urogenital sinus, around the 8th week. The terminal part of mesonephric ducts enters the urinary bladder and traverses caudally and medially to open into the phallic part of the urogenital sinus and it is here that there could be an abnormal fold of tissue

*Classified Specialist (Obstetrics & Gynaecology), †Graded Specialist (Radiodiagnosis), INHS Jeevanti, Vasco da Gama, Goa-403802.

Fig. 1 : Oblique axial scan through fetal abdomen and pelvis shows grossly distended bladder with thickened bladder wall, "keyhole" sign, dilated tortuous ureters, oligohydramnios. B: urinary bladder, U: ureters and S: spine, White arrow: Keyhole sign.

which manifests as the posterior urethral valve. Alternately the posterior urethral valve could also be due to the persistence of the urogenital membrane in between the phallic part of the urogenital sinus and the bulbomembranous urethra.

LUTO seen in the second trimester on ultrasonography, in a male fetus is almost always due to posterior urethral valves [2]. The other causes include prune belly syndrome and urethral atresia which are extremely rare [3]. The classical evidence on ultrasonography of LUTO is the presence of severe oligohydramnios, a grossly distended urinary bladder with increase in bladder wall thickness. The so called "keyhole" sign can be demonstrated if the proximal urethra too is distended.

The outcome of pregnancy and long term prognosis in cases of posterior urethral valves diagnosed antenatally at less than 24 weeks is extremely poor. This is primarily due to the prolonged oligohydramnios and the damage to the fetal kidneys due to the involvement of the upper tracts. The degree of involvement of the fetal kidneys can be ascertained ultrasonographically as well as biochemically [3]. The osmolality, urinary sodium levels and the beta 2 microglobulin levels in the fetal urine will give an indication of the fetal renal function. Poor renal function is indicated if the urinary sodium levels are greater than 100 meq/l, osmolality is greater than 200 mosm/l and the beta 2 microglobulin levels are greater than 2 mg/l [4]. However it may be noted that the specificity of these tests increases if these tests are done serially.

Once the antenatal diagnosis of this condition is made, the dilemma for the clinician is : when to intervene?

Fig. 2 : Male Fetus with arrow pointing to grossly distended bladder.

Whether intervention in such patients improves prognosis at all, is a subject matter of ongoing research, though evidence available so far does not show any significant increase in long term prognosis [3]. The options for intervention include vesicoamniotic shunting, fetal endoscopic ablation of valves and of course termination of pregnancy. Most of the available data relates to vesicoamniotic shunting, as fetal endoscopic ablation is a relatively newer interventional modality first reported in 1995 by Quintero et al [5].

It has been seen that vesicoamniotic shunting in cases of posterior urethral valves does correct the existing oligohydramnios, though the perinatal mortality and long term survival after this procedure remains poor [6,7]. This fact is important as the parents need to be counselled that the incidence of renal failure in childhood, even after vesicoamniotic shunting, could be as high as 50% [8]. Moreover the procedure has a 45% complication rate which includes shunt blockage, preterm delivery and urinary ascites.

The couple have been counselled that though the etiology of posterior urethral valves is believed to be multifactorial with teratogenic embryopathy being the most likely single important factor, there is some evidence that this condition occurs more commonly amongst certain cohorts of population [9]. As a result autosomal dominant inheritance has been suggested though this theory has not been substantiated.

Conflicts of Interest

None identified

References

1. Chatterjee SK, Banerjee S, Basak D, Basu A. Posterior urethral valves: The scenario in a developing country. *Paediatr Surg Int* 2001; 17:2-7.
2. Chowdhary SK, Wilcox DT, Ransley PG. Posterior Urethral Valves: Antenatal diagnosis and management. *J Ind Ass Paed*

Surg 2003; 08: 164-8.

3. Agarwal SK, Fisk NM. In utero therapy for lower urinary tract obstruction : Prenatal diagnosis does it alter outcome? Prenatal Diagnosis 2001; 21:1004-11.
4. Nicolini U, Fisk NM, Rodeck CH, Basehan J. Fetal urine biochemistry: an index renal maturation and dysfunction. Br J Obstet Gynaecol 1992; 99:46-50.
5. Quintero RA, Hume R, Smith C, Johnson MP, Cotton DB, Romero R. Percutaneous fetal cystoscopy and endoscopic fulguration of posterior urethral valves. Am J Obstet Gynecol 1995; 172:206-9.
6. Freedman AL, Johnson MP, Smith CA, Gonzalez R, Evans MI. Long term outcome in children after antenatal intervention for obstructive uropathies. Lancet 1999; 354: 374-7.
7. Holmes N, Harrison MR, Baskin LS. Fetal surgery for posterior urethral valves: Long term postnatal outcomes. Pediatrics 2001; 108: E7.
8. McLorie G, Farhat W, Khoury A, Geary D, Ryan G. Outcome analysis of vesicoamniotic shunting in a comprehensive population. J Urol 2001; 166: 1036-40.
9. Rajab A, Freeman NV, Patton M. The frequency of posterior urethral valves in Oman. Br J Urol 1996; 77:900-4.

Quiz

Radiological Quiz

Lt Col U Rajesh*

MJAFI 2010; 66 : 169

A 31 year old male presented with complaints of recurrent pain in the right side of abdomen. Pain was colicky, intermittent and used to radiate to the right testis. Physical examination revealed no abnormal findings. Haematological and biochemical parameters were normal and showed preserved renal functions. Urine analysis was also within normal limits.

Patient was taken up for ultrasonography (USG) of the kidney, ureter and bladder region. The right kidney measured 119 x 54 mm and showed mild dilatation of

the pelvi-calyceal system (Fig. 1). The right kidney showed normal cortical echotexture and preserved cortico-medullary differentiation. Right proximal ureter was also dilated. No obvious calculus could be seen in the right kidney or the dilated right ureter. Left kidney, left ureter and the urinary bladder were within normal limits.

Patient underwent intravenous urography the next day. Fig. 2 shows 30 minute supine intravenous urogram.

What is your diagnosis?

Fig. 1 : Sagittal sonogram of the right kidney. Right kidney measures 119 x 54 mm and shows mild dilatation of the pelvi-calyceal system. The echotexture of the right kidney is normal and the cortico-medullary differentiation is maintained. No calculus is seen in the right kidney.

Fig. 2 : 30 min supine intravenous urogram shows mild dilatation of the right pelvi-calyceal system. The right proximal ureter, till the level of fourth lumbar vertebra, is also dilated. Medial deviation of the right ureter is seen at the level of L4. Distal one-third of right ureter is normal in course, contour and caliber.

Answer to Radiological Quiz : pg. 182

*Graded Specialist (Radiodiagnosis), 150 GH, C/o 56 APO, PIN-900 129.

Received : 19.10.07; Accepted : 30.09.09 E-mail : uddandamrajesh@yahoo.com