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CASE REPORT

ABDOMINAL MASS IN A NEONATE: A CASE OF NEGLECTED POSTERIOR URETHRAL VALVE

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ABSTRACT: The finding of abdominal mass in a neonate can be alarming for both parents and physicians. Kidney accounts for about 50% of neonatal abdominal mass presenting as flank mass. We present a stillborn neonate with unusual mass in the hypogastrium which is an over distended bladder due to severe Posterior Urethral Valve detected by postnatal USG. Antenatal USG was not done.

KEYWORDS: Posterior Urethral Valve, Abdominal mass, Neonate.
MESHTERMS: Amniotic fluid, fetal growth retardation, hydronephrosis.

INTRODUCTION: The presence of an abdominal mass in the neonatal period is common occurring in approximately 1 in 1000 live births. The common cause of abdominal mass are 55% are renal, 15% involve the genital tract, 15% involve gastrointestinal tract, 10% are retroperitoneal (non-renal) and 5% involve the liver.[1] Posterior urethral valves are one of most common cause of lower urinary tract obstruction in new born males. In most severe case, obstruction leads to urinary retention, hydronephrosis and renal insufficiency.[2] Severely affected infants are often stillborn.[3] We report a rare presentation of PUV in a stillborn term IUGR as abdominal mass due to severe bladder outlet obstruction.

CASE HISTORY: A 32 year old mother with obstetric score of G3P2L2 from a rural area of Villupuram (INDIA) with 9 months of amenorrhea was brought to our hospital with active labour pains and with the complaint of not perceiving foetal movements since last 6hrs. No antenatal scans were done. Labour pains progressed within 20 minutes of admission and she delivered a term male IUGR baby with birth weight of 1.45kgs.

ON EXAMINATION: Baby was cyanosed, apnoeic with no HR & no spontaneous movements. A mass measuring about 12 X 17cms noted in lower part of abdomen extending from umbilicus to hypogastric region, which was soft in consistency and smooth in surface, however the lower margin of the mass could not be felt which aroused a suspicion of mass arising from pelvis. There was no dysmorphic facies or limb deformities noted. Baby was not revived despite active resuscitation as per NRP protocol.

After due consent from the parents child was taken for USG abdomen to evaluate the cause of the abdominal mass. USG done was suggestive of Posterior Urethral Valve with severe bladder outlet obstruction with thickened bladder wall and bilateral hydroureteronephrosis, the renal parenchyma was dense with poor cortico-medullary differentiation. There was no ascites. The mass is due to the over distended bladder with urine.

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DISCUSSION: Posterior Urethral Valve (PUV) disorder is an obstructive developmental anomaly in the urethra & genito urinary system of male newborns. Incidence is 1 in 8000 babies.[3]

PATHOLOGY: PUV results from the formation of a thick, valve-like membrane from tissue of Wolffian duct origin (Failure of regression of the mesonephric duct) that courses obliquely from the verumontanum to the most distal portion of the prostatic urethra.[4] This is thought to occur in early gestation (5-7 weeks).[5] The valve is actually a diaphragm with a central pinhole, however as it is more rigid along its line of fusion it gradually distends, and becomes distended into a bi lobed sail-like or windsock-like structure.[6]

PHYSICAL PRESENTATION OF PUV: Neonates may present with severe pulmonary distress caused by lung underdevelopment due to oligohydramnios. An appropriate volume of amniotic fluid (Produced by the kidneys) is necessary for complete & proper branching of the branchial tree and alveoli.

PHYSICAL FINDINGS INCLUDE THE FOLLOWING:

- Poor foetal breathing movements, Small chest cavity, Abdominal mass (Ascites), Potter facies, Limb deformities (Skin dimpling) and Indentation of the knees & elbows due to compression within the uterus.

IN OLDER CHILDREN IT INCLUDES:

- Poor growth, Hypertension, Lethargy, A large lower abdominal mass may represent a markedly distended urinary bladder.

On antenatal ultrasound the appearance is that of marked distension and hypertrophy of the bladder, with or without hydronephrosis and hydroureter, and depending on the severity, oligohydramnios and renal dysplasia.

Unfortunately such findings are generally not seen before 26 weeks of gestation, and as such are not frequently identified on routine morphology screening, usually carried out around 18 weeks of gestation.[7]

Following birth the bladder is typically thick walled and trabeculated with an elongated and dilated posterior urethra (keyhole sign). The kidneys in most cases are hydronephrotic, although it is important to note that in up to 10% of cases they appear normal. They may also be hyperechoic with loss of the normal cortico medullary differentiation, a manifestation of renal dysplasia.[7]

VOIDING CYSTOURETHROGRAM (VCUG): It is the best imaging technique for the diagnosis of posterior urethral valves. The diagnosis is best made during the micturition phase in a lateral or oblique views, such that the posterior urethra can be imaged adequately.[8]

FINDINGS INCLUDE:

- Dilatation and elongation of the posterior urethra (Equivalent of the ultrasonographic keyhole sign).
- Linear radiolucent band corresponding to the valve.
- Vesicoureteral reflux (VUR)-seen in 50% of patients.
- Bladder trabeculation/diverticula.

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TREATMENT: Antenatallyvesicoamniotic shunting is possible. Essentially this procedure consists of a supra-pubic catheter performed under USG guidance. The efficacy of this procedure is controversial, as often despite this significant renal & pulmonary morbidity exist.[8] Postnatally, definitive treatment is simple and involves transurethral ablation of the offending valve.[9]

COMPLICATIONS: IN THE NEW BORN: Besides the urological and nephrological manifestations of the PUV, the associated oligohydramnios can lead to pulmonary hypoplasia, facial dysmorphy and deformity of the extremities, in some cases leading to foetal death.[10]

Similar to our case where the child was stillborn baby but without any abnormal facies or limb deformities.

THE OLDER CHILDREN CAN DEVELOP:

- Renal insufficiency, Vesicoureteral reflux, Recurrent UTI and Urinary incontinence.[11]

Long term follow-up of PUV patients by various studies done by parkhouse et al, Kousidis et al, Heikkila et al have concluded a bad long term renal outcome, Chronic renal impairment, Acute renal failure and End stage renal disease in various percentages.[12]

PROGNOSIS: Over the last 30 years, the prognosis of children with PUV has steadily improved. In the past, most children were found to have PUV only after presenting with urosepsis or progressive renal insufficiency. Today, most individuals with PUV are discovered when prenatal ultrasonography reveals hydronephrosis:

- Prompt resolution of bladder obstruction, aggressive treatment of bladder dysfunction, and improved surgical techniques has lowered the neonatal mortality rate to less than 3%.
- Approximately one third of patient's progress to renal insufficiency in their lifetimes. Improved dialysis and transplantation techniques have significantly improved not only the mortality rate for these children but also their quality of life.[13]

CONCLUSION: To conclude posterior urethral valve is a threatening congenital anomaly that needs appropriate surgical treatment and long term follow up. Diagnosing it antenatally and intervening it at the right time can prevent mortality and reduce morbidity due to this dreadful condition.

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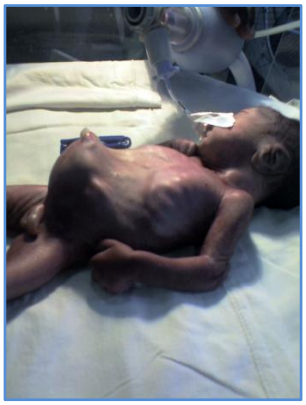

Figure 1

Figure 2

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