ORIGINAL RESEARCH PAPER | Medical Science | Volume : 6 | Issue : 7 | July 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

A case of Prune Belly Syndrome Complicated by Pneumoperitoneum

KEYWORDS: Prune Belly Syndrome, Pneumoperitoneum

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ABSTRACT *Prune belly syndrome, a rare congenital anomaly, affecting about 1 in 40000 live births; of which 95% are male.PBS is constellation of anterior abdominal defect, bilateral hydroureteronephrosis and abnormalities of the genitourinary tract. Here we report a case of a male baby with PBS because of its rarity, acute complication of pneumoperitoneum & fatal course.*

Introduction:

Prune Belly Syndrome consists of a constellation of congenital malformations, with the classical triad of, anterior abdominal defect , bilateral hydroureteronephrosis and abnormalities of the genitourinary tract. While the unusual "prune" like appearance of abdomen is the hallmark of PBS, the underlying renal function is the most important factor in determining their overall survival.

CASE REPORT:

A male baby was born to a 23year old, primigravida mother, with no h/o consanguinity,by LSCS, indication being breech presentation with oligohydroamnios at 35 weeks of gestation. Birth weight was 3.45kg.Antenatal USG revealed oligohydroamnios with bilateral gross hydroureteronephrosis with dilated urinary bladder. There was no h/o radiation exposure, fever with rash or any drug intake in this pregnancy. Neither she was a known case of Diabetes Mellitus nor there was any h/o genetic or congenital anomaly in the family.

Baby had delayed cry, requiring PPV for 1minute. Baby was kept in NICU in view of respiratory distress and gross distension of abdomen.

Examination findings:

Baby was in respiratory distress (RR-68/min),with mild cyanosis in room air(SPO2-88%)with good perfusion. Baby had facial dysmorphism, low set ears and mal-developed right pinna, with grossly distended abdomen, with wrinkled appearance and bilateral lumbar swellings with hypogastric lump. Scrotal skin showing little rugae and no testis in the sac. Baby had bilateral clubfeet [Fig.1].Rest of the general and systemic examination were apparently normal.

Course:

Baby was kept under oxygen support and iv maintenance fluids. Paediatric surgery consultation was sought and as per his advice baby was catheterised.USG abdomen was suggestive of **multiple cysts in the right kidney** with bilateral hydronephrosis, ascitis, thickened urinary bladder with left undescended testes. Blood investigations were normal, Kidney function tests were also normal(S.urea-11,s. creatinine-0.6,Sr.Na-135,Sr.K-4.6).On day2 of life, gradually trophic feeds were introduced, suddenly after few hours, baby developed significant abdominal distension and severe respiratory distress.On examination, baby was having tachycardia, tachypnea and severe subcostal retractions and SPO2-60% with oxygen,abdomen was tense,shiny and distended.ABG suggested severe metabolic acidosis. The baby was intubated and put on mechanical ventilation. ray showed pneumoperitoneum.[Fig.2]Repeat kidney function tests were normal.Pediatric surgery reference was taken.On abdominal paracentesis, foul smelling purulent discharge seen. The general condition of the baby kept on deteriorating with worsening of shock. Fluid resuscitation and multiple isotopes were added. Antibiotics were upgraded. Despite everything, baby expired on day3 of life.

DISCUSSION:

We wish to highlight this case of PBS for its rarity and its stormy course, as it was complicated by pneumoperitoneum.

Exact cause of pneumoperitoneum could not be established. We presume either it is because of sepsis with NEC, or it has been originated in the thorax from a pulmonary air leak with underlying pulmonary hypoplasia or may be due to rupture of G.I tract with underlying G.I abnormality.

In addition to the classical triad, a broad spectrum of mu sculoskeletal(50%),cardiovascular (10%) and genital malformations are noted[1,2].When the urinary tract mal-development is associated with severe obstructive uropathy, this syndrome can lead to oligohydroamnios and pulmonary hypoplasia.The embryological defect that is responsible is still not known but there is a theory suggesting that a defect exists in the mesoderm of the anterior abdominal wall and urinary tract. Between 6-10 weeks of gestation,aberrant development of derivatives of 1st lumbar myotome leads to patchy muscular deficiency of abdominal wall. Alternate theory suggests that urethral obstruction leading to bladder distension causes pressure atrophy of abdominal wall muscles and interfere with the descent of testes. The mechanism responsible for urinary tract dilatation and distension is a flap valve mechanism resulting from hypoplasia of stromal and epithelial elements of prostatic urethra[3].

Neonatal management includes radiological evaluation. BUN, creatinine and serum electrolyte level should be obtained for baseline and carefully followed during the 1st week.A rising level suggest poor prognosis. Severely affected patients require early surgery(supravesical diversion or primary reconstruction)to provide adequate drainage

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and avoid recurrent infections. Renal replacement therapy for those with ESRD includes dialysis and renal transplantation [4]. Orchiopexy should be performed to prevent malignancy. Abdominal musculature reconstruction for both aesthetics and function is necessary.

One third babies are stillborn or die within 1st few months because of pulmonary complications. As many as 30% of survivors develop end stage renal damage due to reflux and eventually require transplantation[5].

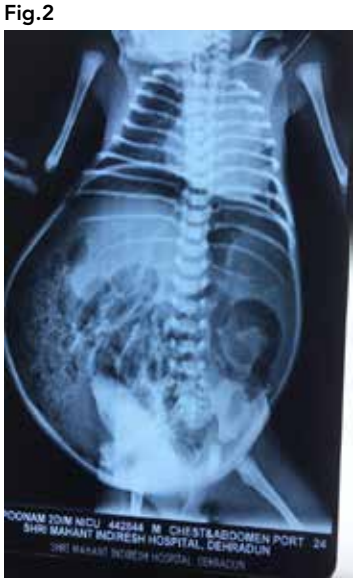
Poor prognostic factors include oligohydroamnios, huge urinary ascites, dystrophic bladder ,pneumoperitoneum and peritoneal calcification.

CONCLUSION

PBS is a rare congenital anomaly which has no known prevention other than the routine use of screening for fetal anomalies. Routine antenatal care with ultrasonography will help in detecting renal anomalies early and optimal treatment will ensure optimum outcome.

Fig.1

Fig.2



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