



Prune belly syndrome associated with bilateral multicystic dysplastic kidneys and urethral obstruction: A case report

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Abstract Prune belly syndrome is a rare congenital disorder defined by a characteristic clinical triad: Abdominal muscle deficiency, severe urinary tract abnormalities, and bilateral cryptorchidism. We describe a preterm neonate of Prune Belly syndrome who had abdominal muscle deficiency, multicystic dysplastic kidney, urethral hypoplasia and pulmonary hypoplasia. We presented this rare case with the data gathered from the literature.

Key Words *Newborn; prune belly syndrome; pulmonary hypoplasia; multicystic dysplastic kidney; urethral obstruction.*

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INTRODUCTION

Prune belly syndrome (PBS) is a rare congenital disease occurring about 1 per 50.000 live births. It is characterized by

anomalies such as deficiency of abdominal wall musculature, bilateral cryptorchidism and urinary tract abnormalities. 97% of PBS cases are males, and 3% of are females [1-3].

PBS is also associated with pulmonary, cardiovascular, gastrointestinal and musculoskeletal abnormalities [1-3]. Oligohydramnios and pulmonary hypoplasia

are common complication in PBS. The prognosis is depending on severity of pulmonary hypoplasia and renal dysplasia [1,4]. We present a case of prune belly syndrome with bilateral multicystic dysplastic kidney and urethral.

CASE REPORT

The male neonate was delivered by vaginal delivery after 32 weeks of gestation to a 35-year-old gravida 6, para 5 mother. In family history, mother had gestational diabetes that regulated with only diet arrangement and another feature out of it was not. In prenatal history of the infant; oligohydramnios, bilateral clubbed feet and bilateral infantile polycystic kidney had found in obstetric ultrasonography at gestational age of 20 weeks and termination of pregnancy had recommended to family. The patient's 1st and 5th minute APGAR scores were 1 and 3, respectively. He required intubation and ventilation support due to respiratory distress that developed after birth and was admitted to the neonatal intensive care unit. He weighed 2240 g (90th centile), his length was 40 cm (25-50th centile) and head circumference was 28 cm (25-50th centile). Physical examination on admission showed multiple dysmorphic features, including flattened nasal bridge, micrognathia, cryptorchidism, bilateral clubfoot.

Abdominal examination revealed distended abdomen with thin wrinkled skin and visible peristalsis (Fig. 1) with mass in left lower quadrant size of approximately 4x4 cm.

Bilateral pulmonary hypoplasia and pneumothorax were found in radiographs and a chest tube was placed. The patient had absolute anuria and trial of insertion of urethral catheter was unsuccessful. This condition was considered associated with urethral hypoplasia. Ultrasonography of the abdomen showed bilateral multicystic dysplastic kidneys and mesenteric cyst of 4 cm size in left lower quadrant. Based on the present findings, PBS was diagnosed.

Although mechanical ventilation, hypoxia was continued and at eighth hour of postnatal period cardiopulmonary arrest occurred. Cardiopulmonary resuscitation was performed but the patient dead.



Fig. 1. Flaccid abdomen due to abdominal muscle deficiency.

DISCUSSION

PBS is a rare congenital disease characterized by a triad; deficiency of abdominal wall musculature, bilateral cryptorchidism and urinary tract abnormalities. The disease occurs in approximately 1 in 50.000 live births and 97% of patients are male. There is wide variability in disease severity, and patients may also experience concomitant cardiopulmonary, gastrointestinal and musculoskeletal anomalies [1-3]. PBS rarely can occur in females with anomalies of uterus and vagina [5].

The etiology and pathogenesis of PBS is not yet fully understood. It has been suggested that the underlying defect in PBS is abnormal mesoderm development affecting the embryogenesis of the abdominal wall musculature, the mesonephric and paramesonephric ducts, and the urinary organs [1,4].

All of the male PBS cases have cryptorchidism and follow and treatment are necessary [3]. Malrotation and anal atresia are frequently seen as gastrointestinal abnormalities in PBS [1,3]. Our case had cryptorchidism too, and mesenteric cyst had found by ultrasonography.

One of the remarkable features of PBS is urinary tract abnormalities. The most common urinary abnormalities are

megaureter and megabladder [1-4]. Others are renal dysplasia and hydronephrosis. In our case, bilateral multicystic dysplastic kidney and urethral hypoplasia were determined. It has been suggested that early urethral obstruction by a valve-like mechanism is the primary abnormality in PBS, which can then lead to the other clinical manifestations [6,7]. We thought that the lesion presented as mesenteric cyst on ultrasound, may belong to the bladder.

Rarely, complex cardiac anomalies may occur with PBS (10%). The most common cardiac anomalies are atrial septal defect, ventricular septal defect and pulmonary stenosis [3]. In our case, 2/6 systolic murmur was heard on mesocardiac focus. But echocardiography could not be performed because of death at postnatal eighth hours.

Potter face which includes hypertelorism, micrognathia, flattened nasal bridge, epicanthal folds occurs due to uterine press causing oligohydramnios. Also musculoskeletal abnormalities are considered associated with oligohydramnios [1]. Our case had micrognathia, flattened nasal bridge, bilateral clubfoot.

The prognosis in PBS is due to severity of pulmonary hypoplasia and renal dysplasia [1,4]. The present case died due to severe urinary abnormalities and associated

pulmonary hypoplasia at early postnatal period.

This case report to suggest that untreatable urinary abnormalities are part of the broad spectrum of clinical features of prune belly syndrome. Mortality may be decreased with early diagnosis and treatment. These patients need multidisciplinary management of a

neonatologist, nephrologists, and pediatric urologist for an optimal outcome.

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