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Prune Belly Syndrome (Eagle Barrett Syndrome): A Rare Cause of Abdominal Wall Defect with Renal Anomalies in Children

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ABSTRACT

Prune belly syndrome is an interesting clinical case which is infrequently described in medical literature.

Case presentation: A 1 year old boy presented to our institution with recurrent urinary tract infections and Prune belly syndrome.

Conclusion: We report the rarely described disease entity of Prune belly syndrome.

Keywords: Prune belly syndrome, Urinary tract infections

INTRODUCTION

Prune belly syndrome (PBS) (Eagle Barrett syndrome) is a rare disorder characterized by the triads of deficient abdominal wall muscles, urinary anomalies (hypoplastic kidneys or hydronephrosis and hydroureter) and undescended testes [1,2]. It is caused by urethral obstruction early in development resulting in massive bladder distension and urinary ascites, leading to degeneration of the abdominal wall musculature and failure of testicular descent. The impaired elimination of urine from the bladder leads to oligohydramnios, pulmonary hypoplasia and Potter's facies. The syndrome has a broad spectrum of affected anatomy with different levels of severity. The exact etiology of PBS is unknown, although several embryologic theories attempt to explain the anomaly [3]. It results from congenital absence of anterior abdominal wall muscles. The term "prune-belly" reflects the characteristic wrinkled appearance of the abdominal wall in the newborn due to the complete or partially complete absence of abdominal wall muscles. In adolescent and adult life, the abdomen often assumes a smooth "pot belly" configuration and 95% of patients with this syndrome are males. There are rare case reports of this disorder in females. This syndrome has derived its name from the wrinkled prune appearance of the abdominal wall. Prune Belly syndrome is a rare anomaly seen in one in 35,000-50,000 live births. It occurs in all races. Prune Belly syndrome almost exclusively occurs in males (M:F, 20:1). The diagnosis can be made in utero by ultrasonography at 21 weeks of gestation or in the Neonate with characteristic clinical findings [4].

This syndrome is thought to be due to mesenchymal genetic defect, which remains unknown. It has also occurred in association with other chromosomal abnormalities, including trisomy 13, 18 and 21. The severity of renal pathology is the primary factor that affects the clinical manifestations of PBS. Dysplasia is the main renal abnormality of PBS, and it is manifested by incomplete nephron differentiation and dilatation of the tubules. About one-half of patients have severe dysplasia and develop end-stage renal disease (ESRD) and require renal replacement therapy. In addition, insufficient fetal urine production leading to oligohydramnios results in pulmonary hypoplasia and neonatal mortality among patients with severe PBS. Less commonly, gastrointestinal malformations (e.g. malrotation of the midgut or anorectal malformations), skeletal abnormalities (e.g. clubfoot) and rarely cardiac anomalies are observed [5-7].

CASE REPORT

A 1 year old male was brought to the Department of Pediatrics and Child Health of TikurAnbessa Specialized Hospital due to fever of 2 days duration. The patient was the only child of unrelated parents, born at term by spontaneous vaginal delivery. There was no history of maternal drug or alcohol abuse, x-ray exposure or infection during pregnancy.

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The child suffered from recurring urinary tract infections starting from the age of 3 months, with five episodes till the age of 1 year.

Physical examination upon admission revealed a stature (81 cm) and body weight (9.3 kg) within the normal ranges for age. Other physical findings were fever (37.9°C), lax abdominal wall muscles, bimanually palpable kidneys and bilaterally undescended testes **(Figure 1)**.

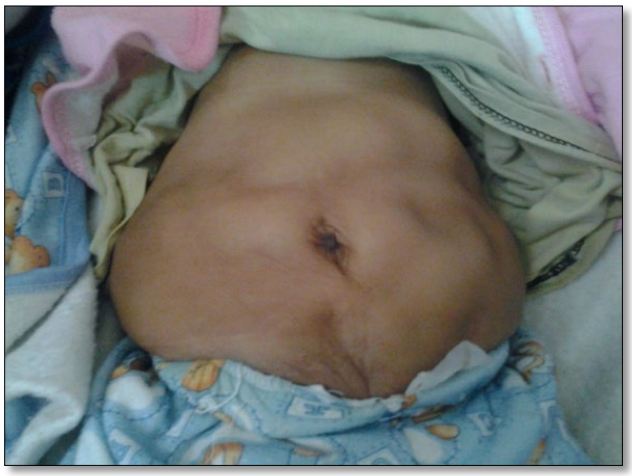

Figure 1. Lax abdominal wall muscles and peristalsis seen upon inspection of the patient's abdomen.

The psychomotor milestones of development were not impaired.

Investigations showed: A WBC count of 26,900 with a neutrophil percentage of 76.7%; urinalysis revealing many WBC/HPF. Urine culture revealed *Enterobacter fecalis*. Bilateral hydronephrosis was diagnosed on an abdominal ultrasound **(Figure 2)**. Renal function tests were within normal limits. IVP (intravenous pylography) showed mega ureter.

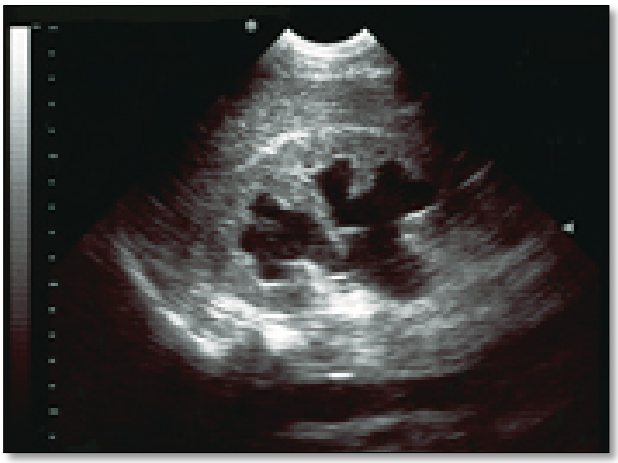

Figure 2. U/S showing severe hydronephrosis in this patient.

He was diagnosed with febrile urinary tract infection and Prune belly syndrome and treated with a course of parenteral antibiotics.

DISCUSSION

We presented the infrequently diagnosed case of Prune belly syndrome. The diagnosis was made due to deficient abdominal wall muscles, undescended testes and bilateral hydronephrosis with mega ureter and recurrent urinary tract infections.

Surgical management may be needed to provide adequate urinary drainage and avoid urinary tract infections in some patients. Renal replacement therapy for those patients with ESRD includes dialysis and renal transplantation.

Other interventions include additional genitourinary procedures to improve bladder control and capacity, orchiopexy and abdominal wall reconstruction.

Renal hypoplasia, hydroureter, hydronephrosis, ureterovesical and ureteropelvic junction obstruction,

posterior urethral valve and vesicoureteric reflux, have all been associated with this syndrome [8]. PBS has rarely been reported in association with Down syndrome (DS) [9].

Current theories on the pathogenesis of PBS suggest some yet unknown mesodermal injury and or in utero urinary tract obstruction [10]. A genetic cause may also be possible. However, this does not exclude modification of the severity of PBS by the associated chromosomal anomaly [11]. It has been recognized recently that many genes involved in renal nephrogenesis either reappear or are expressed to a markedly greater degree in renal disease [12]. The prognosis of PBS is poor with stillbirths and early infant deaths being common [13].

CONCLUSION

The diagnosis of PBS is difficult for it is associated with many congenital anomalies. Clinical criteria like lax abdominal wall muscles and multiple urinary congenital anomalies with recurrent UTI and abdominal U/S with renal contrast studies like IVP are important diagnostic methods for this syndrome. Our patient fulfills all the diagnostic criteria; hence, it is a real case of PBS.

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