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Case Report

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Severe Neonatal Prune Belly Syndrome Complicated by Terminal Obstructive Renal Failure: A Case Report from CHME Nouakchott, Mauritania

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Abstract

Prune Belly Syndrome is a rare congenital anomaly characterized by the triad of abdominal wall muscle deficiency, severe urinary tract abnormalities, and bilateral cryptorchidism. Prognosis mainly depends on the severity of renal involvement. We report the case of a male full-term neonate born to consanguineous parents who presented with a severe form of Prune Belly Syndrome revealed by marked bilateral ureterohydronephrosis associated with near-complete destruction of the right kidney, progressive renal failure, and post-obstructive diuresis complicated by refractory severe hyponatremia. Despite multidisciplinary management and urinary drainage, the clinical outcome was fatal. This case highlights the severity of neonatal forms of Prune Belly Syndrome and emphasizes the importance of early antenatal diagnosis and specialized multidisciplinary management.

Keywords: Prune Belly Syndrome, Neonate, Obstructive Uropathy, Renal Failure, Post-Obstructive Syndrome, CHME, Mauritania

1. Introduction

Prune Belly Syndrome, also known as Eagle-Barrett syndrome, is a rare congenital malformation affecting almost exclusively males. Its estimated incidence ranges from 1 in 30,000 to 1 in 50,000 live births [1]. The syndrome is defined by a characteristic triad consisting of deficiency of the abdominal wall musculature, severe urinary tract abnormalities, and bilateral cryptorchidism [2].

The pathophysiology of PBS remains incompletely understood. Two main hypotheses have been proposed:

- a primary mesodermal developmental defect;
- early lower urinary tract obstruction leading to chronic bladder distension, secondary abdominal wall muscle atrophy, and impaired testicular descent [3].

The prognosis of PBS mainly depends on the severity of renal and pulmonary involvement. Severe neonatal forms are associated with high mortality, particularly in cases of bilateral renal dysplasia or delayed diagnosis due to lack of antenatal follow-up [4]. We report a severe neonatal case of PBS complicated by terminal obstructive renal failure and fatal post-obstructive syndrome.

2. Case Presentation

A male neonate born to consanguineous parents was delivered at term following an unmonitored singleton pregnancy. Delivery was performed by cesarean section because of a triple-scarred uterus. Apgar scores were 8/10 at both the first and fifth minutes. Maternal infectious history was unremarkable. The mother was 37 years old, gravida 5 para 4 abortus 1, blood group O Rh positive, and from a low socioeconomic background. The neonate was admitted to the neonatal unit for further management.

Physical examination revealed:

- A distended, flaccid abdomen suggestive of severe hypoplasia of the abdominal wall musculature.
- Micropenis.
- Bilateral testicular ectopia.
- Bilateral clubfoot deformity.

- Asymmetric intrauterine growth restriction.

Anthropometric measurements at admission were:
- Weight: 2300 g.
- Length: 52 cm.
- Head circumference: 35 cm.

Overall clinical findings were suggestive of a polymalformative syndrome consistent with Prune Belly Syndrome.

Abdominopelvic ultrasonography demonstrated:
- Severe bilateral ureterohydronephrosis.
- Near-complete destruction of the right kidney.
- Hypofunction of the left kidney.

These findings were consistent with severe obstructive uropathy associated with advanced renal impairment.

Initial laboratory investigations showed:
- Blood urea: 1.67 g/L.
- Serum creatinine: 59 mg/L.
- C-reactive protein: 33.6 mg/L.
- Initially normal complete blood count.

Follow-up laboratory tests revealed progressive deterioration of renal function with:
- Blood urea: 1.8 g/L.
- Serum creatinine: 80 mg/L.

Urine culture revealed more than 1000 leukocytes/mm³ with polymicrobial flora.
The neonate was managed in the neonatal intensive care unit with:
- Supportive care.
- Empiric antibiotic therapy.
- Bladder catheterization.
- Close clinical and biological monitoring.

Due to worsening urinary obstruction associated with progressive renal impairment, a diversion nephrostomy was performed.

Clinical evolution was marked by:
- severe post-drainage polyuria.
- post-obstructive syndrome.
- refractory severe hyponatremia (119 mmol/L).
- profound anemia (hemoglobin 5 g/dL) requiring transfusion of two packed red blood cell units.

Despite intensive neonatal resuscitation measures, the outcome was unfavorable with fatal cardiorespiratory arrest.

3. Discussion

Prune Belly Syndrome is a rare and severe polymalformative disorder characterized by the association of abdominal wall muscle deficiency, urinary tract abnormalities, and bilateral cryptorchidism [1]. Our case illustrates a complete and particularly severe presentation of the syndrome, including the three major components of the diagnostic triad. In our patient, abdominal distension with a flaccid and hypotonic abdomen was related to marked hypoplasia of the abdominal wall musculature. This abnormality is mainly explained by the obstructive theory, according to which early fetal lower urinary tract obstruction causes chronic urinary tract distension leading to secondary abdominal muscle atrophy [3]. Parental consanguinity, identified in our observation, is also noteworthy. Although most PBS cases are sporadic, recent studies suggest a possible genetic contribution involving abnormalities in embryonic mesodermal development [5,6].

Uro-renal involvement represents the major prognostic factor in PBS [7]. In our case, abdominopelvic ultrasonography demonstrated severe bilateral ureterohydronephrosis associated with near-complete destruction of the right kidney and hypofunction of the left kidney. These findings reflected advanced renal dysplasia with a very poor prognosis. The progressive elevation of blood urea and serum creatinine indicated rapid deterioration of renal function. Several recent studies have reported chronic renal failure as the leading cause of mortality in severe forms of PBS [4,7]. Genital abnormalities observed in our patient, particularly micropenis and bilateral testicular ectopia, are frequently described in this condition. Bilateral cryptorchidism is reported in more than 95% of cases [8].

Associated orthopedic abnormalities, including bilateral clubfoot deformity observed in our patient, are commonly reported in severe forms of PBS. These abnormalities are thought to result from chronic oligohydramnios secondary to fetal urinary tract abnormalities, leading to musculoskeletal deformities caused by intrauterine compression [2]. In our case, the absence of antenatal follow-up probably contributed to delayed diagnosis and poor outcome. Antenatal diagnosis of PBS mainly relies on obstetric ultrasonography, which may reveal megacystis, bilateral hydronephrosis, oligohydramnios, and severe renal abnormalities [1,9]. Early antenatal diagnosis allows better neonatal management planning and may improve prognosis. In selected cases, fetal urinary diversion procedures may be considered to reduce progression toward severe renal dysplasia [10].

Our patient underwent diversion nephrostomy because of worsening obstructive anuria. However, the postoperative course was complicated by post-obstructive syndrome with massive polyuria and refractory severe hyponatremia. Post-obstructive syndrome is a serious complication of severe obstructive uropathy resulting from impaired tubular concentrating ability following drainage of chronic urinary obstruction [3]. The profound hyponatremia observed in our patient probably reflected irreversible tubular injury aggravated by advanced renal failure [3]. The severe anemia that developed secondarily may have resulted from multiple interacting mechanisms, including advanced renal failure, persistent inflammatory state, severe metabolic disturbances, and prolonged critical illness [3,4,7]. Despite intensive neonatal resuscitation and multidisciplinary management, the outcome was fatal. This finding is consistent with the literature reporting high

neonatal mortality in severe PBS associated with bilateral renal involvement, early renal failure, and delayed diagnosis [4].

In low-resource settings, several additional factors worsen the prognosis:

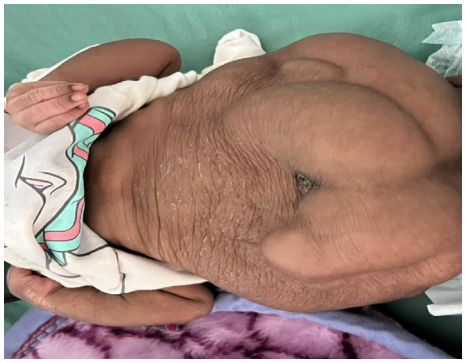
- Limited or absent antenatal screening.
- Restricted access to specialized healthcare facilities.
- Insufficient neonatal intensive care resources.
- Limited availability of pediatric urological surgery and neonatal dialysis techniques [4].

This case therefore highlights the importance of:

- Early antenatal screening for obstructive uropathies.
- Prompt multidisciplinary management.
- Specialized nephrological follow-up.
- Strengthening neonatal intensive care capacities.

4. Conclusion

Prune Belly Syndrome is a rare and severe congenital malformation predominantly characterized by uro-renal abnormalities that largely determine both vital and functional prognosis. Our case illustrates a particularly severe neonatal form complicated by advanced obstructive renal failure and fatal post-obstructive syndrome. This observation emphasizes the importance of early antenatal diagnosis and rapid specialized multidisciplinary management in order to improve prognosis and reduce complications associated with this condition.



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