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CASE REPORT

Prune Belly Syndrome

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Abstract

Prune belly syndrome is a rare congenital disorder, characterized by a triad of abnormalities, such as deficiency or absence of abdominal wall musculature, bilateral cryptorchidism, and malformation of the urinary tract. This is a case report of a male newborn vaginally delivered at full term, with a birth weight of 3,335 g and an Apgar score of 9 at the 1st minute and 10 at the 5th minute. Physical examination revealed bilateral cryptorchidism, reduced abdominal wall tone, and wrinkled appearance of the abdominal skin, which presented a “prune” appearance. The patient underwent an ultrasound examination of the urinary tract, which showed kidneys in the normal position with bilateral hydronephrosis. The newborn progressed with urinary retention and increased nitrogenous waste products. The bladder was catheterized for 5 days, with spontaneous urination after the catheter was removed. Before discharge, the newborn underwent another urinary ultrasound, which showed mild bilateral hydronephrosis. In this patient, only the alterations inherent to the triad itself were observed. Therefore, the goal of the treatment became the prevention of urinary tract infection with prophylactic antibiotic therapy, because there was no longer any obstruction of the urinary tract. The newborn was referred for pediatric surgery in order to evaluate the appropriate time to perform cryptorchidism correction and abdominoplasty. The objective of this case report was to raise awareness of the syndrome, to facilitate an early diagnosis, preferably in utero, and ensure that the treatment, which chiefly focused on urinary system abnormalities, is not delayed.

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INTRODUCTION

Prune belly syndrome (PBS), also called triad syndrome or Eagle-Barrett syndrome, is characterized by a triad of abnormalities, namely, deficiency or absence of the abdominal wall muscles, bilateral cryptorchidism, and malformation of the urinary tract¹.

PBS is a type of fetal uropathy that lacks a clear etiology, but several theories have been proposed to clarify it, among which the obstructive theory and the mesodermal arrest theory have been the most frequently cited¹⁻³. According to the obstructive theory, the syndrome arises from a urethral and/or ureteral obstruction, which would cause gross dilation of the fetal urinary tract, consequently distending the abdominal wall, compromising its appropriate formation, and preventing testicular descent. According to the mesodermal arrest theory, the syndrome is caused by a disorder in mesodermal development during the third week of pregnancy, as this germ layer is responsible for the formation of the urinary tract, kidneys, and abdominal muscles¹⁻⁴.

The estimated incidence of PBS is 1:40,000 live births, and 95% of the affected children are male¹, with only 3%-5% of the cases occurring in females - the latter being called pseudo-prune belly syndrome, due to the absence of cryptorchidism⁴.

Individuals with this syndrome generally present loose and wrinkled abdominal wall skin; approximately 30% of cases show absence of the anterior abdominal wall muscles. The skin folds in the periumbilical region resemble the surface of prunes, hence the name of the syndrome, "prune belly."

Bilateral cryptorchidism is an essential component of the syndrome, with the intra-abdominal testes¹ located along the dilated ureteral segments at the height of the iliac vessels.

In the urinary tract, clinical changes are manifested in varying degrees, although the kidneys can be normal; renal dysplasia and hydronephrosis are common¹.

As for the prognosis, up to 30% of patients surviving for long periods develop chronic renal failure and occasionally require kidney transplants¹.

CASE REPORT

The patient was a male newborn (NB) vaginally delivered at term, with a birth weight of 3,335 g, length of 52 cm, 1-minute Apgar score of 9, and 5-minute Apgar score of 10.

Prenatal imaging exams reported the following observations:

- Obstetric Ultrasound (US) at 28 weeks: presence of bilateral hydronephrosis, more evident in the right kidney; overdistended bladder with wall thickening; and "keyhole" configuration, suggesting the presence of a posterior urethral valve.

- Obstetric US at 33 weeks: both kidneys featured pyelocalyceal dilatation, and the bladder dimensions and volume slightly increased, suggesting the presence of a posterior urethral valve.

In spite of these ultrasound changes, antenatal care was uneventful.

According to the delivery room (DR) description, the NB was born in good vital conditions and presented urination in the DR.

On the first day of life of the NB, physical examination revealed bilateral cryptorchidism, and abdominal examination revealed reduced tone of the abdominal wall and wrinkled skin, creating a "prune" aspect (Figure 1). In addition, the NB progressed with urinary retention. An abdominal US, urinalysis, and laboratory tests were requested. The US showed the kidneys in the normal position, bilateral hydronephrosis, and a full bladder, with regular walls and homogeneous content, with a suspicion of vesical distension. Urinalysis results were normal. Laboratory test results were as follows: hemoglobin: 19.0 g/dL, hematocrit: 58.7%, leukocytes: 23,300/mm³, band cells: 0%, segmented cells: 72%, lymphocytes: 15%, platelets: 163,000/mm³, urea: 41.00 mg/dL, creatinine: 2.02 mg/dL, calcium: 10.5 mg/dL, and magnesium: 4.0 mg/dL.



Figure 1. Absence of abdominal wall tone and wrinkled aspect of the skin.

After receiving the test results, we requested an evaluation from the pediatric surgery staff, who opted for urethral catheterization due to the vesical distension and increased nitrogenous waste products. The NB developed macroscopic hematuria.

We decided to start antibiotic therapy with ampicillin and gentamicin to conduct intravenous hydration and request a cystourethrogram for better evaluation of the urinary tract.

On the next day, new laboratory tests were requested, and the results were as follows: hemoglobin: 16.8 g/dL, hematocrit: 47.5%, leukocytes: 16,450/mm³, band cells: 3%, segmented cells: 62%, lymphocytes: 18%, platelets: 160,000/mm³, urea: 27.00 mg/dL, creatinine: 0.71 mg/dL, potassium: 5.60 mmol/L, sodium: 145.0 mEq/L. Urine culture was negative.

An echocardiogram was performed, owing to the risk of associated cardiovascular changes, and it showed patent foramen ovale and mild pulmonary hypertension.

The urethral catheter was removed after 5 days, with subsequent spontaneous urination. Another urinary tract US was performed, and it showed mild bilateral hydronephrosis and a partially distended bladder, with inconclusive signs of parietal thickening and somewhat irregular internal contours.

We discussed the case with the pediatric surgeon. As the NB was roomed-in, had spontaneous urination for 48 hours, presented a reduction in the degree of hydronephrosis compared with that observed on the US from a week before, and normalization of urea and creatinine levels, we decided to discharge him from the hospital, with subsequent outpatient follow-up.

The NB was discharged with antibiotic prophylaxis (cefalexin) and was referred for pediatric surgery and to nephrology outpatient clinics.

DISCUSSION

PBS is characterized by deficient abdominal muscles, cryptorchidism, and urinary tract abnormalities. Cardiac anomalies are present in 10% of cases¹. Gastrointestinal malformations, such as intestinal malrotation, can also occur. In the case of our patient, only the anomalies inherent to the triad itself were observed.

When the urinary tract is obstructed, temporary drainage procedures, such as vesicostomy, can help preserve kidney function until the child is old enough for surgery. This procedure was originally requested in this case because the NB presented urinary retention on the first day of life, with increased urea and creatinine levels. However, urethral catheterization was performed as an immediate measure, and the NB progressed with spontaneous urination after withdrawal. Therefore, the aim of the treatment became prevention of urinary tract infection with prophylactic antibiotics because the urinary tract was no longer obstructed¹. The reason for

this conduct was that urinary tract infections frequently occur in these patients.

Some children with this syndrome present posterior urethral valves, which was reported in the obstetric US in our case study.

Cryptorchidism is corrected by orchidopexy, with better results when the surgery is performed in the first year of life³.

Abdominal wall reconstruction offers both functional and aesthetic benefits. Abdominoplasty aims at enhancing lung and bowel functions. This procedure should be avoided during growth stages, owing to the development of respiratory disorders by abdominal traction.

In the present case, the NB was referred for pediatric surgery in order to define the appropriate time to perform cryptorchidism correction and abdominoplasty.

The NB was also referred to the outpatient pediatric nephrology clinic for better investigation of the urinary tract changes. Until the completion of this report, the NB had not yet undergone a cystourethrogram due to paperwork issues.

Because PBS is a complex syndrome that may have an adverse outcome if not diagnosed early enough, it is important to emphasize the importance of *in utero* diagnosis, which requires strict surveillance by an obstetrician and monitoring by a radiologist during antenatal care.

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