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Prune Belly Syndrome: A Rare and Special Case

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Abstract

Prune Belly syndrome (PBS) is an extremely rare anatomo-radiological syndrome that combines aplasia of the muscles of the anterior abdominal wall, dilatations of the urinary tract and testicular malformations, thus forming the classic triad of the syndrome. Up to 75% of patients with PBS have pulmonary, skeletal, cardiac, and gastrointestinal malformations. We report the case of a full-term male infant with no particular pathological history admitted for etiological work-up of bilateral ureterohydronephrosis diagnosed antenatally with a distended, prune-shaped abdomen with bilateral testicular ectopia on clinical examination. The outcome can be very variable, ranging from stillbirth due to major renal and respiratory dysplasia to a practically normal child, all of which explains the great diversity of opinions on the attitude to adopt when faced with this syndrome.

Keywords: ***Prune belly syndrome; Prune belly; Polymalformative syndrome; Variable prognosis***

1. Introduction

Prune Belly syndrome (PBS) or Eagle-Barrett syndrome is a congenital disorder that typically associates aplasia or severe hypoplasia of the muscles of the anterior abdominal wall, urinary malformations and bilateral cryptorchidism [1,2]. However, other malformations may be associated, such as pulmonary, skeletal, cardiac and gastrointestinal malformations. There are also so-called incomplete or partial forms more frequent in females, where the hypoplasia of the abdominal wall is partial or unilateral associated or not with renal, urinary and osteoarticular malformations, which are generally on the same side. These partial forms are known as Pseudo-Prune Belly Syndrome (PPBS) [3,4].

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The clinical forms can be very variable, ranging from stillbirth due to major renal and respiratory dysplasia to the practically normal child. 95% of carriers of this syndrome are male, however it is generally more serious in girls due to a higher incidence of urethral atresia. This explains the diversity of opinion on the attitude to adopt when faced with this syndrome.

We report a case of the complete form of Prune Belly syndrome discovered in a male newborn at birth.

2. Clinical Case

Newborn male, from a non-consanguineous marriage, from a well-monitored pregnancy carried to term, delivery by cesarean section for scarred uterus, Apgar 8/10 increased to 10/10 at the 5th minute. Mother aged 30, G2P2, 2 living children, without particular pathological history, negative infectious history.

The clinical examination found a pink, reactive, hypotonic newborn with a weak sucking reflex, a weight of 3 kg, a height of 50 cm and a head circumference of 35 cm. The abdominal examination found a distended abdomen, a thinned abdominal wall with palpation of the intestinal loops subcutaneously, and a lumbar contact. The cardiovascular, pleuropulmonary and osteoarticular examination was without particularities. The paraclinical examinations included: a thoraco-abdominal radiograph which showed a bilateral interstitial syndrome, with a cardio-thoracic index of 0.62 as well as a deviated gastric air pocket (FIG. 3). An abdominal ultrasound showed a significant bilateral ureterohydronephrosis, diverticular bladder with globe (posterior urethral valve) with left ureterocele. A trans-thoracic ultrasound and a transfontanellar ultrasound were performed without particularities.

The biological assessments are unremarkable, in particular renal function which is normal.

FIG. 1. **Image of our patient showing the thinned appearance of the abdominal wall.**

FIG. 2. **Image of our patient showing the appearance of the large kidney.**

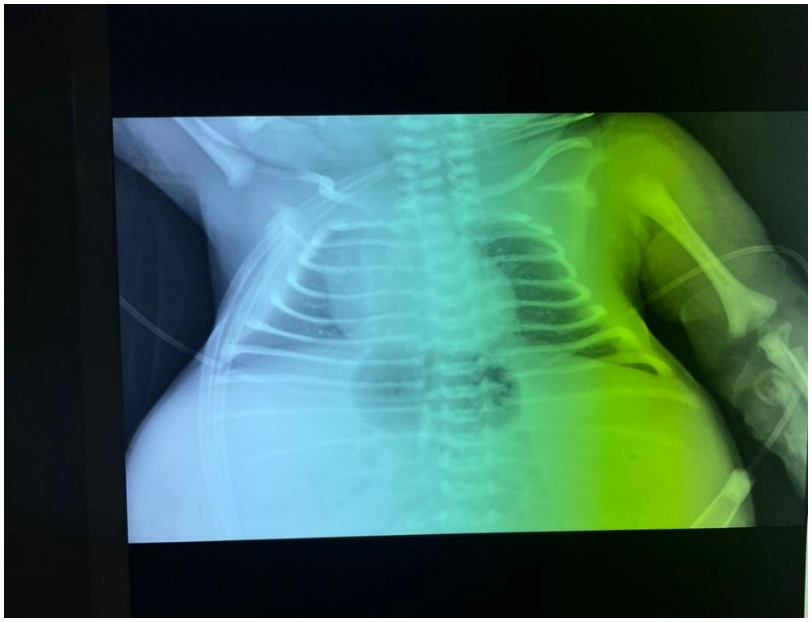

FIG. 3. **Thoraco-abdominal radiograph showing bilateral interstitial syndrome with cardiomegaly and a deviated gastric air pouch.**

3. Discussion

Eagle-Barret syndrome is a complex and rare malformation. According to various studies, the incidence of Prune Belly syndrome is estimated at one case in 40,000 births [1]. This condition is marked by a clear male predominance and by the rarity of complete forms in girls [1,2]. More than 95% of patients are male [1]. This clear male predominance has raised the possibility of genetic involvement with sex-linked autosomal recessive transmission.

The term Prune Belly syndrome was first described in 1839 by Fröhlich. Later in 1895, Parker made the first description associating anomalies of the urinary tract. And it was not until 1950 that Osler named it "Prune Belly" in view of the appearance of the abdominal wall [1,2]. According to various studies, the incidence of Prune Belly syndrome is estimated at one case in 40,000 births [2]. This condition is marked by a clear male predominance, more than 95% of patients are male, and by the rarity of complete forms in female children who generally do not present with urinary malformations [1,5,6]. The exact etiology of PBS is unknown. 3 theories predominate: firstly, the one that proposes a prenatal urinary obstruction, secondly, the one that is based on embryology and that proposes the failure of primary mesodermal differentiation between the 6th and 10th week of gestational age which leads to defective muscularization of the abdominal wall and urinary tract. Thirdly, that of the bladder sacs, which proposes dysgenesis of the velum sac and the allantois [2]. Clinically, the main components of this syndrome are urinary malformations, namely a megabladder, dilated ureters and ureters but which can also be stenotic or atretic with a higher incidence in girls, polycystic kidney disease, hydronephrosis, and sometimes a diverticulum near the vesicoureteral and urethral junction [1,2,6]. The state of renal function is an important prognostic determinant [1,2]. Our case having a complete form of the syndrome, presents vesico-renal malformations such as bilateral ureterohydronephrosis, posterior urethral valve with left ureterocele and normal renal function. However, up to 75% of patients with PBS associate other malformations, namely pulmonary, cardiac, skeletal, gastrointestinal and genital malformations. These malformations have been reported by Routh et al with an incidence of 25% for cardiovascular, 24% for gastrointestinal, 23% for musculoskeletal, 58% for respiratory and 15% for genital [2]. The respiratory malformations found are pulmonary hypoplasia and cystic adenomatoid malformation which can lead to different degrees of respiratory failure, the main reason for neonatal mortality. Gastrointestinal malformations such as mesenteric malrotation, atresia, stenosis, volvulus, imperforate anus, splenic torsion, Hirschsprung's disease and gastroschisis. Osteoarticular malformations such as clubfoot, hip dysplasia, vertebral malformations and scoliosis. Cardiovascular malformations such as patent ductus arteriosus and tetralogy of Fallot [1]. Genital malformations such as cryptorchidism occur in almost all male patients, which is the case in our patient, however, anomalies of the corpus cavernosum or prostatic hypoplasia have also been reported. In women, genital malformations include vaginal atresia, bicornuate uterus and urogenital sinus. There have never been cases of infertility in either women or men [7,8]. In rare cases, hypoplasia of the abdominal wall muscles is unilateral, as are other associated malformations, renal, testicular and bone, generally found on the same side as the abdominal muscular hypoplasia, thus describing the incomplete form generally more frequent in women, also called Pseudo Prune Belly Syndrome (PPBS) [3,4].

Our patient presents a complete form of the syndrome with abdominal wall involvement with urogenital malformations. The diagnosis is based prenatally on obstetric ultrasound capable of detecting abnormalities of the urinary system associated with the typical appearance of the abdominal wall. Hosbino reported a case of PBS diagnosed at 12 weeks of pregnancy [1,2]. Postnatally it is based on abdominopelvic ultrasound completed by abdominopelvic CT, transthoracic ultrasound to look for cardiac malformation, renal assessment to evaluate renal function, ultrasound of the hips with a skeletal radiograph to look for skeletal malformations as well as a karyotype to look for suppression on nuclear factor 1-beta (HNF1beta) [1,9]. Treatment is mainly surgical: abdominoplasty, orchidopexy, and urinary tract reconstruction [1,2,5,6,10]. For patients with mild abdominal wall dysplasia, postures are acceptable and do not require abdominoplasty. However, for severe cases, surgical treatment is discussed on a case-by-case basis. Pyelostomies, ureterostomies, and cystostomies are also undertaken to temporarily shunt urine in some unstable infants who cannot tolerate surgery. Sometimes kidney transplantation is unavoidable for patients with renal failure. Regardless, whether surgical treatment is undertaken or not, patients with PBS require ongoing multidisciplinary

medical care and close follow-up. The prognosis for patients with PBS varies depending on the severity of pulmonary hypoplasia and urinary tract abnormalities. Pulmonary hypoplasia is the leading cause of mortality in the neonatal period. The severity of urinary tract abnormalities and renal function determine not only mortality but also long-term prognosis [1,2,5,6].

4. Conclusion

Prune Belly syndrome is rare and mainly affects males. Renal failure and pulmonary hypoplasia are the main causes of mortality. In the absence of the classic triad, it is imperative to look for other malformations given the existence of atypical forms, in order to undertake adequate and immediate management.

REFERENCES

1. Xu W, Wu H, Wang DX, et al. A Case of Prune Belly Syndrome. Pediatr Neonatol. 2015;56(3):193-6.
2. Fette A. Associated rare anomalies in plum belly syndrome: a case report. Journal of Pediatric Surgery Case reports. 2015;3(2):65-71.
3. Grover H, Sethi S, Garg J, et al. Pseudo Prune Belly syndrome: diagnosis revealed by imaging? A case report and brief review. Pol J Radiol. 2017;82:252-7.
4. Zugor V, Schott GE, Labanaris AP. The Prune Belly syndrome: urological aspects and long-term outcomes of a rare disease. Pediatr Rep. 2012;4(2):e20.
5. Diao B, Diallo Y, Fall PA, et al. Prune Belly Syndrome?: Epidemiological, Clinical and Therapeutic Aspects. Prog En Urol. 2008;(18):470-4.
6. Seidel NE, Arlen AM, Smith EA, et al. Clinical manifestations and management of Prune Belly syndrome in a large contemporary pediatric population. Urology. 2015;85(1):211-5.
7. Gallo C, Costa W, Favorito L, et al. Prune belly syndrome: is penile structures similar to normal fetuses? In: State University of Rio de Janeiro, Urogenital Research Unit, Rio de Janeiro, Brazil. London, United Kingdom. 2017.
8. Tibor-Denes F, Tavares A, Machado M, et al. Gonadal function and reproductive system anatomy in post puberal plum belly syndrome patients. J Urol. 2017;197(4S):5.
9. Granberg CF, Harrison SM, Dajusta D, et al. Genetic basis of prune belly syndrome: screening for HNF1gene? J Urol. 2012;187(1):272- 8.
10. Dénes FT, Park R, Lopes RI, et al. Abdominoplasty in Prune Belly Syndrome. J Pediatr Urol. 2015;11(5):291-2.