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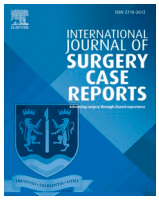

Case report

A case of prune belly syndrome: Experience from a regional hospital in Togo

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ABSTRACT

Introduction and importance: Prune Belly syndrome is a rare congenital condition first reported in 1939. It is a malformative disease associated with high mortality. We report a neonatal case in a regional hospital in Togo.
Presentation of case: A 4-month-old male infant was brought to the hospital by his uncle because of a "deformity of the abdomen and absence of testicles". The examination revealed hypoplasia of the abdominal wall associated with bilateral cryptorchidism. Abdominal and urinary tract ultrasound revealed a left pyelocaliceal dilatation and a right megaureter. All these data allowed the diagnosis of Prune Belly syndrome.
Conclusion: Prune Belly syndrome is a rare and complex disease with a high mortality rate.

1. Introduction

First described by Frolish in 1839, Prune Belly syndrome or Eagle-Barrett syndrome is a rare and complex condition [1–3]. It is predominantly male [1,2,4]. The prune-like appearance of the belly is the most characteristic manifestation of the disease [1]. The diagnosis is based on a radio-clinical triad described in 1895 by Parker [1]. The disease is a malformative entity involving the abdominal wall, the testicles and the urinary tract [1–4]. It is a neonatal disorder [1]. Prune Belly syndrome is a condition with an undefined etiopathogenic. The management of this syndrome has improved due to a better understanding of its pathophysiology [4]. However, this management remains complicated in countries with limited resources. We report in our work a case of Prune Belly syndrome observed in a 4-month-old male infant in a regional hospital in Togo. This manuscript was written according to the rules of the SCARE [5].

2. Presentation of case

This is a 4-month-old male infant brought to the surgery by his uncle for "abnormal appearance of the abdomen and absence of testicles". This was the first medical consultation for this symptom. The history reports a vaginal delivery after a poorly monitored monofetal pregnancy. Two (02) episodes of dysuria were reported by the parents, as well as traditional management. Moreover, we did not note any notion of consanguineous marriage between the parents. No family history was found. Inspection of the unclothed infant showed abdominal distension with a bilobed appearance of the abdomen and subcutaneous visualisation of the intestinal ansae (Fig. 1). Palpation reveals laxity and severe atrophy of the anterolateral abdominal wall allowing subcutaneous palpation of the intraabdominal organs. Examination of the bursae revealed that they were empty. Biologically, we noted normal renal function with uraemia at 0.2 g/l and creatinemia at 11 mg/l and a normal urine cytobacteriological examination. Abdominal and urinary tract ultrasound revealed normal-sized testes in the abdominal position, pyelocaliceal dilatation with left hydronephrosis and a megaureter with right ureterohydronephrosis. *Trans*-thoracic ultrasound did not report any cardiac abnormality. IVUS, CT scan, cystography and karyotype were not available in the centre. The diagnosis of Prune Belly syndrome was therefore retained in the light of these radio-clinical arguments. Care was provided by general surgeons. Due to the inadequacy of our technical facilities, our patient was referred to a reference centre with a pediatric surgery department for appropriate management. The patient has been put under close surveillance (quarterly). He is waiting for a testicular reduction at the age of 2 years.

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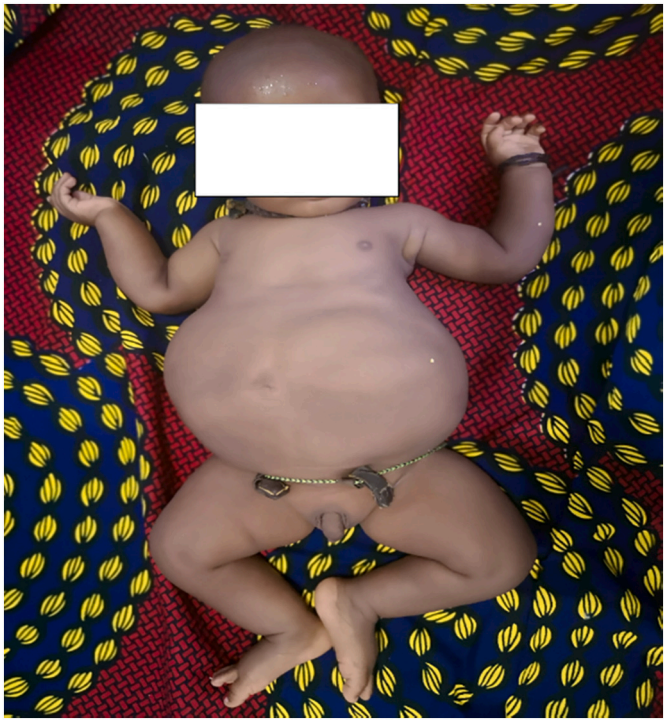

Fig. 1. Abdominal distension with bilobed appearance and subcutaneous visualisation of the digestive tract.

3. Discussion

We report on a case of prune belly syndrome discovered in a regional hospital in Togo. This case once again revealed the difficulty of managing patients in facilities with limited technical resources.

Prune Belly syndrome is also known as Eagle-Barrett syndrome [1–3]. It is a rare, congenital and complex disease that was described in 1839 by Frolish [1–3]. It has an incidence of approximately 1 case per 40,000 births [1,4]. The disease affects males preferentially [1–4].

Diagnosis may be antenatal based on obstetric ultrasound [3,4,6–8]. Diagnosis in the 2nd trimester of pregnancy is the most reported [8]. This ultrasound can detect urinary and abdominal parietal abnormalities [6]. These include oligohydramnios, significant bladder dilatation, pyelocaliceal dilatation and abdominal wall weakness with prominence on sagittal sections [6,8].

In the majority of cases, the diagnosis is neonatal as in our case [1]. It is a radio clinical diagnosis. It is based on a triad reported by Parker in 1895 [1,9,10]. This triad associate malformative lesions of the abdominal wall, the testicles and the urinary tract [1,4,10,11]. At the abdominal level, it is aplasia or hypoplasia of the abdominal wall [1,8]. This malformation is the most common reason for discovery [1,3,11]. Cryptorchidism is the testicular lesion involved; it is most often bilateral [1,2,9]. Lesions of the urinary tract concerning the bladder, ureters and pyelocaliceal cavities. This triad was complete in our infant. In some cases, other malformations are associated with this triad. These malformations may be cardiac, pulmonary, osteoarticular, gastrointestinal, and genital [1,9–11]. None of these malformations was detected in our patient. The rarity of this condition is at the origin of its lack of recognition by health personnel, which sometimes leads to a delay in diagnosis [1,2]. Indeed, some authors have reported diagnoses of Prune Belly syndrome in elderly subjects [1,2,9]. The delay in consultation in our case is linked to the initial management by a traditherapist (talismans on the waist and wrist in Fig. 1). In summary, the diagnosis of this disease is based on clinical and abdominal-pelvic ultrasound [6,10]. The search for associated malformations may require, depending on the case, abdominal and pelvic CT scans, trans-thoracic ultrasound, renal workup to assess renal function, hip ultrasound with skeletal X-ray and karyotype for deletion of nuclear factor 1-beta (HNF1beta) [6,10]. In our case, abdominopelvic CT, karyotype, and osteoarticular imaging were not performed.

The pathogenesis of Prune Belly syndrome is not well understood [10]. Hereditary, toxic and hormonal factors have been described [9]. Three theories have been described: prenatal obstruction of urine flow; arrested development of the lateral or intermediate mesoderm during embryonic life leading to hypoplasia of the abdominal wall and urinary malformations; and dysgenesis of the velum sac and allantois [4,8,10].

The management of Prune Belly syndrome is quite complex. It must take into account the three types of malformations. Opinions are divided between surgical and conservative treatment [3,7]. However, the mainstays of treatment are abdominoplasty, a cure for cryptorchidism with lowering of the testicles and orchidopexy, and urinary tract reconstruction [1,10]. Early management of cryptorchidism in the neonatal period or during infancy ensures good testicular function [2,9]. In all cases, therapeutic indications are determined on a case-by-case basis [10]. Major advances have been made in the management of this disease [7]. The key element of this management remains qualified personnel, who guarantee a satisfactory result [7]. Due to the inadequacy of our technical facilities, our patient was referred to a reference centre with a pediatric surgery department for appropriate management.

The mortality rate of this condition in the neonatal period is between 10 and 25 % [3,8]. The prognosis of the disease depends on the severity of the malformations [1,3]. Urological lesions are prognostically important [9,10]. However, renal failure and pulmonary hypoplasia are the main causes of mortality [8,10,11]. Patients with Prune Belly syndrome should therefore benefit from multidisciplinary management due to its complexity [3,4,10].

4. Conclusion

Prune Belly syndrome is a rare, congenital disease affecting mainly males. This rarity is responsible for a delay in diagnosis. The hypoplasia of the abdomen, the main mode of revelation, makes its diagnosis easy. The management of this syndrome is complex and there is no consensus. The condition is associated with a high perinatal mortality rate, requiring early and multidisciplinary management.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Ethical approval

The study protocol met the requirements of the ethics committees of the Dapaong Regional Hospital Center Centre and was approved.

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Author contribution

ETB: Study design and data acquisition.
KSP, KAP: literature review.
ETB, PE and IM: manuscript editing.

All authors: manuscript review.

Registration of research studies

Not applicable.

Guarantor

ESSOBIYOU Tamassi Bertrand.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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