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CASE REPORT

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Prune Belly Syndrome in Neonatology at Dschang District Hospital : About Two Cases

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Abstract

Prune Belly syndrom is a typical triad of abdominal muscle deficiency, severe urinary tract abnormality and bilateral cryptorchidism. It is very rare and account for about 1 case in 40,000 lives birth. Diagnosis can be made antenataly, but more ofen postnataly. The surgical procedure, including simultaneous abdominoplasty, bilateral orchiopexy, and urinary tract reconstruction, is performed according to individual needs. The severity of the renal dysplasia is the main prognostic factor. We report two cases of a rare syndrom diagnosed in Dschang district hospital within 8 month of interval. They were all boys newborns with the typical triad and had urinary tract infection. Both are still waiting for this surgical managment. The prognosis was not severe since, they did not have any renal failure. Prune belly syndrome is described as a rare triad, and is not kwnoun by all health personnal. It was important for us to present these cases.

Summary

Prune Belly syndrome is a typical triad of abdominal wall defect, severe urinary tract abnormalities and bilateral cryptorchidism in boys. It is very rare, with one case per 40,000 births. Its diagnosis can be made antenatal, but very often postnatal. The treatment is surgical. The triple surgery consisting of tummy tuck, bilateral orchidopexy and urinary tract repair is performed according to individual needs. Renal hypoplasia is the major prognostic factor. We report two cases of Plum Belly Syndrome diagnosed at Dschang District Hospital within 8 months. They were all boys with the classic form of this syndrome. They all had a UTI with good kidney function and are still awaiting corrective surgery. Their prognosis is therefore not committed. Prune Belly syndrome is described as rare and too little known to health workers. We found it important to present it.

Keywords: Plum Belly; Urinary Tract Infection; Skin Fistula; Dschang

Introduction

Prune Belly syndrome or Eagle-Barett syndrome, described by Frolich in 1839, is very rare, characterized by the triad associating: aplasia or hypoplasia of the abdominal wall, malformations of the urinary tract and bilateral cryptorchidism [1]. This pathology mainly affects boys in 98% of cases and has an incidence of 1 in 40,000 births [1,2]. Its diagnosis can be made prenatal by obstetric ultrasound, clinically postnatal and by ultrasound [3]. In 75% of cases, this syndrome is associated with other malformations that can be life-threatening. The progression without treatment is to short-term respiratory failure when there is pulmonary hypoplasia. In the medium and long term, the disease leads to renal failure and infertility [3,4]. Medical treatment depends on the complications found and may require triple surgery combining orchidopexy, abdominoplasty and urinary tract repair [4]. We report two cases of Prune Belly syndrome received at the neonatal department of the Dschang district hospital in Cameroon, in eight months apart.

Clinical case N °1

This is a newborn baby, 2 days old, male, who was referred from a nearby health center for treatment of laparoschisis.
The parents were not consanguineous and had no known chronic pathology. He was the first child of a 19-year-old mother. The pregnancy followed was without major risk and an obstetric ultrasound performed at 34 weeks was normal. The mother had had an episode of malaria in the third trimester of pregnancy which was successfully treated. The delivery was vaginally at the end of 38 weeks with a birth weight of 2850 grams and an Apgar score of 9/10 at the fifth minute. On arrival at the neonatal ward, the baby was awake, alert, with good muscle tone. He did not have facial dysmorphia. His weight was 2800 grams, his height 49cm and his head circumference 36cm. Under ambient air, his oxygen saturation (SPO2) was 92%, his heart rate (HR) was 123 beats per minute, his respiratory rate (RF) was 48 cycles per minute, and his temperature (T °) was 36. , 7 ° C. His neurological exam was normal. Cardiopulmonary auscultation was without abnormality. We also noted: a flaccid, asymmetrical abdomen with an arch by complete muscular atrophy allowing the viscera to be palpated on the right flank, expansive when crying. Periumbilical wrinkles were noted (Figure). The testicles were absent in the purses. The rest of the exam was normal. The diagnosis of Prune Belly syndrome had been suggested and an abdominal ultrasound requested. The result of this abdominal ultrasound was: defect of the abdominal wall on the right flank with herniation by complete muscular atrophy and only the presence of the skin layer, bladder, ureteral and pyelocaliceal distension suggesting a valve of the posterior urethra or a syndrome of the pyelocalic junction. The diagnosis of Prune Belly syndrome has been confirmed. The additional assessment included: A CBC which was normal for the age with a hemoglobin level of 16 g / l. An ECBU showed a urinary tract infection, the causative agent of which was identified as Proteus species, sensitive to Gentamicin and intermediate to Ceftriaxone. Urea and serum creatinine were normal at 13 mg / dl and 0.61 mg / dl, respectively. The chest x-ray and transthoracic cardiac ultrasound were also normal.

Antibiotic treatment for urinary tract infection has been proposed by Ceftriaxone at 50 mg / kg of body weight for ten days and gentamycin at 5 mg / kg of body weight for three days. A triple surgery had been considered. The parents had left against medical advice for a hospital in the capital by interrupting the treatment of the urinary tract infection on the third day, under pressure from the family. Tracking this baby from then on was difficult. He remains awaiting surgery, but his physical growth was normal at the 14-week follow-up visit with a weight of 5.2 kg (-1 SD).

Clinical case N °2

Newborn male, referred to the neonatal ward by the maternity team at Dschang District Hospital, where he was born. He was admitted on the 2nd day of life for abdominal malformation and absence of urine since birth.

The mother was 32 years old, with no known pathology, of pregnancy formula G3P2002, and the 32-year-old father, with no known pathology. The parents were not consanguineous. It was the 3 rd child in a family of 3, all in apparent good health. Pregnancy was unrecognized until the age of 22 weeks, as the mother continued to have periods. The obstetric ultrasound performed at 32 WA

was normal. The toxoplasmosis, rubella and cytomegalovirus serologies were not performed. No pathology was noted during pregnancy. He was born at the end of 39SA +5 days, vaginally in the maternity ward of the Dschang District Hospital with a birth weight of 3200 grams and an Apgar score of 10/10 by the fifth minute. On physical examination he was alert, with good muscle tone and no signs of facial dysmorphism. Its parameters were: T ° 37.2 ° C FC 132 beats per minute FR 41 cycles per minute SPO2 97% under ambient air.

Cardiopulmonary auscultation was normal. In the abdomen, there was a wrinkled appearance with multiple folds of the wall (Figure), without any real localized defect, on an otherwise very soft stomach. A fluid discharge under the umbilicus, resembling urine, suggested a vesico-cutaneous fistula. The urine had not been emitted naturally. The testicles were absent in the purses. The kidneys and bladder were not palpable; no organomegaly was noted. No other anomalies were observed. We have mentioned the diagnosis of Plum Belly Syndrome associated with a very probable vesico-cutaneous fistula. Abdominal ultrasound showed two kidneys of respected size and parenchyma, bilateral pyelic dilation, severely dilated ureter (bilateral mega ureter), thick-walled empty bladder a thin anterior abdominal wall with anterior abdominal muscle hypoplasia and cryptorchidism bilateral. The ECBU had shown a urinary tract infection with *Escherichia coli* . Renal function was preserved with urea at 22 mg / dl and creatinine at 0.67 mg / dl. The chest radiograph was normal and transthoracic echocardiography showed an atrial septal defect (ASD) type ostium secundum. Unable to obtain antibiogram results, the urinary tract infection had been treated, empirically and probabilistically, by a bi-antibiotic therapy combining ceftriaxone at 50 mg / kg per day for 10 days and Gentamicin at 5 mg / kg / 24 h for 3 days. Subsequently, the baby was transferred for reconstructive surgery to a referral hospital in the capital, while his growth was normal. The chrurgy had been programmed and was still pending. The follow-up of the evolution of this baby was difficult and was done, most often, by phone with the father, given the difficulties of movement.

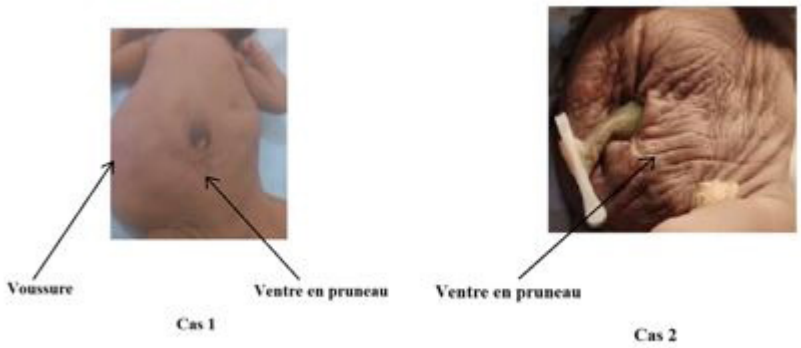

Figure : Images of the prune bellies of the two cases and the arch of the right flank of the first case

Discussion

Prune Belly syndrome is extremely rare, with a prevalence of 3.8 cases per 100,000 live births [1,2]. We had the opportunity to diagnose two cases within 8 months in Dschang, Cameroon. Our two cases were male and this is the most common form in nearly 75% of cases [3]. But, female forms are described in the literature. Thus, seven cases of Plum Belly equivalent have been described by Reinberg et al in Minneapolis, Minnesota, associating hypoplasia of the abdominal wall and abnormalities of the urinary tract. They concluded that the female form was strongly associated with urethral atresia, a factor of poor prognosis [5]. The adult forms also exist and are revealed by various complications according to several authors [6]. Thus, splenic torsion with malrotation has been described in an 18-year-old young man by Trans et al [7], while Wallner et al have described end-stage renal failure in a 35-year-old man [8].

The diagnosis is antenatal. Yamamoto et all described a case of early prenatal diagnosis at 11 weeks of Prune Belly [3], but despite the ultrasounds of our two patients, our diagnoses were postnatal. This diagnosis was not easy, even clinically, for the health personnel who initially saw our patients, who respectively reported laparoschisis and abdominal wall abnormalities.

Urinary tract abnormalities such as megaureteria often form the basis of urinary tract infections [9]. Our two patients had urinary tract infections.

Management is essentially surgical and takes place at varying ages depending on the patients and their need for surgical repair. However, Zugor et all showed in their study on the long-term evolution of 16 babies of their cohort that half of them did well without surgery [10]. Our two patients remain awaiting surgery at the parents' expense and at a significant cost. Their prognosis is not committed and they are growing well, despite the urinary tract infections which risk leading to renal failure in the long term. We encountered a lot of limits with our cases and at various levels ranging from diagnosis to management. These difficulties were linked, on the one hand, to the ignorance of the pathology by health professionals, and on the other hand, to the parents' poverty as well as their ignorance. The lack of health insurance has been a major obstacle to optimal care for our patients. We cannot follow them by not respecting appointments and by a non-optimal collaboration between the different health personnel.

Conclusion

We have presented two cases of neonatal diagnosis of Prune Belly syndrome with the classic triad in male babies at an interval of only 8 months for a syndrome which is however extremely rare. The diagnosis was not easy due to ignorance of the pathology. Appropriate care had many limits linked to ignorance, poverty and insufficient knowledge and technical facilities. We hope, through the publication of these cases, to popularize this pathology and make it better known by health professionals. We would also like to make parents aware of the need for appropriate care within acceptable deadlines. Universal health insurance could help improve the care of these patients.

Conflict of Interest

The authors declare that they have no conflict of interest for the publication of these clinical cases.

Acknowledgments

We thank the parents of these newborns who allowed us to publish these cases.

Authors Contribution

All the authors contributed to the realization of this work. They read and approved the final manuscript.

Ethical Considerations

Parents had been informed of the publication of the cases with photos of their babies and their written informed consent had been obtained.

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