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Case Study

Prune Belly Syndrome with Hydrocephalus and Craniostenosis

Hayat Aichaoui*

Oran, Algeria

***Corresponding Author**: Hayat Aichaoui, Oran, Algeria.

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Abstract

Introduction: The Prune Belly syndrome (PBS) is a complex and rare malformation with a clear male predominance, characterized by the association of a major bladder dilation, a distention with impaired muscles of the anterior abdominal wall that is flaccid and wrinkled and cryptorchidism. PBS is not a genetic disease.

Objectives: To make known this pathology through our observation.

Observation: This is a newborn male, born at home, eutrophic term resulting from a pregnancy not followed. Admitted to the neonatology unit at 2 hours of life. He had an abdominal protrusion whose musculature is loose; anal polyp; the left foot bot talus; cryptorchidism; the anterior fontanel closed. The exploration found craniostenosis, hydrocephalus with cerebral parenchymal hyper-echogenicity, nephromegaly with bilateral hydronephrosis, and mega-bladder. A multidisciplinary management (neuro-surgical, nephrological, urological, orthopedic and pediatric) was programmed for this child but the parents did not show up for the appointments, having required a convocation after which they inform us that their child is dead!

Conclusion: The diagnosis of Belly Prune Syndrome is easy from the 15th week of pregnancy; urologic surgery improves the prognosis and delays the occurrence of renal failure main cause of mortality. In 2019 there are many pregnancy not followed at our country.

Keywords: *Prune Belly Syndrome; Newborn*

Introduction

We had this case in 2009, but it did not have the opportunity to be published. It happened when I was doing my civil service in a mother-child establishment in the city of Oran, in Algeria.

The prune Belly syndrome (PBS) is a complex and rare malformation with a clear male predominance, characterized by the association of a major bladder dilation, a distention with impaired muscles of the anterior abdominal wall that is flaccid and wrinkled and cryptorchidism. PBS is not a genetic disease.

Objectives of the Study

To make known this pathology through our observation.

Our observation

This is a newborn male, born at home eutrophic term from a non-follow-up, admitted to the neonatology unit at 2 hours of life.

Clinical examination at admission

- Weight 3 kg300, PC 32 cm
- Strong cries
- 136 bpm, no heart murmur
- 42 c/min, no respiratory distress
- Anal polyp, good passage of the rectal probe
- Protrusion of the abdomen, absence of abdominal musculature
- small penis, purses are empty
- Left talus clubfoot
- Closed anterior fontanel
- The archaic reflexes are present and normal.

Para-clinical examinations

- HT: 45%
- Blood urea 0.12 g/l, serum creatinine 6.3 mg/l
- Negative CRP
- Radio chest probe in place and ASP are normal
- Trans-fontanellar echography: mega communicating big cistern with the ventricular system, moderately dilated V3 and V4, associated with a cerebral parenchymal hypergenesis.
- Abdominal echo: Enlarged kidneys with significant dilatation of pelvic-calytic cavities bilaterally almost completely laminating the renal parenchyma Thin - walled mega - bladder discharges laterally and upwards the digestive loops. Spleen and liver of normal appearance, and absence of intraperitoneal fluid effusion.

At 24 hours of life

- Generalized mottling, lack of meconium and oligo-anuria
- Hypertonic convulsions of the limbs and perioral cyanosis Supported
- Sedation of convulsions by Gardenal® then related by depakine® after 48 hours
- Antibiotics
- Breastfeeding initiated after hemodynamic and neurological stabilization

- Monitoring of urinary catheter diuresis, transit monitoring: rectal probe stimulation and evacuation lavement.

- An appointment was fixed for the follow - up at Uro-nephro-pediatric service.

Evolution

- Parents do not accept the illness of their child, despite our interview with them and the psychological care within our service.

- New born put out as soon as the feeding became satisfactory, and resumption of a normal diuresis, with a control in the 3 following days at the level of our consultation.

For a follow-up in parallel with the specialized care in the service of pediatric uro-nephrology and psychological care.

The parents did not show up for the appointment, but 2 months later they come back to inform us of the death of their child

The usual treatment

Is surgical:

- Urine diversion by nephrostomy, cutaneous ureterostomy, cystostomy, in severely affected patients

- Reconstitution of the urinary system by reimplantation and modeling of the ureter according to an anti-reflux procedure

- Treatment of cryptorchidism in several times

- Plication of the abdominal wall.

Usual prognosis of the disease:

- Most often lethal the first weeks of life

- Chronic constipation, severe abdominal meteorism

- Pulmonary hypoplasia

- Chronic urinary tract infections, renal lithiasis, kidney failure

- Infertility

- And other complications related to the associated diseases.

Prevention

Antenatal diagnosis is possible to 15 SA, with an antenatal surgical correction.

Conclusion

The diagnosis of Belly Prune Syndrome is easy from the 15th week of pregnancy, urologic surgery improves the prognosis, and delays the occurrence of renal failure main cause of mortality [1-4].

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