Prune Belly Syndrome. A case report

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Background
Prune belly syndrome is a rare anomaly that comprises a specific constellation of features. It consists of three main findings: gross pelvicalyceal and uretric dilatation with renal dysplasia, anterior abdominal wall underdevelopment, bilateral undescended testes in males (1). Is presented the case of a guatemalan 1-month-old male patient, with abdominal distension, presenting distortion of the anatomy, with palpable masses in both iliac fossae, in addition to hypospadias, no testicles are observed in the scrotal area. The presence of Megabladder, bilateral hydroureter, and bilateral hydronephrosis is documented. He had a creatinine level of 1.09 and urea nitrogen of 11.2

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Prune belly syndrome is a rare anomaly that comprises a specific constellation of features. It consists of three main findings: gross pelvicalyceal and uretric dilatation with renal dysplasia, anterior abdominal wall underdevelopment, bilateral undescended testes in males. However, in recent years the prognosis of patients with this diagnosis has been improving since there are a variety of diagnostic methods that allow the clinician to provide a better approach depending of each patient.

Case report

It is presented the case of a 1-month-old male patient, originally from Tecun Uman, San Marcos, product of a transperitoneal cesarean section, who in his 34th week in prenatal ultrasound is diagnosed with hydronephrosis and mega bladder; at 35th week the patient´s mother develops high blood pressure, is referred from a private hospital to the ¨Hospital Regional de Occidente¨, in Quetzaltenango city, when the pregnancy is resolved without complications.

Patient on physical examination; Weight: 1.94kg height/length: 40cm head circumference: 30cm HR: 143 SaO2%: 93% RR: 43x T: 37C GMT: 74 mg/dl. The patient had abdominal distension, presenting distortion of the anatomy, with palpable masses in both iliac fossae, in addition to hypospadias, no testicles are observed in the scrotal area. The presence of Megabladder, bilateral hydroureter, and bilateral hydronephrosis is documented. He has a creatinine level of 1.09 mg/dl and urea nitrogen of 11.2 mg/dl.

Discussion

Prune belly syndrome, also known as Eagle Barrett syndrome or triad syndrome, is a rare anomaly that comprises a specific constellation of features. It consists of three main findings: gross pelvicalyceal and uretric dilatation with renal dysplasia, anterior abdominal wall underdevelopment, bilateral undescended testes in males (1). It is estimated that there are currently a little more than 300 cases reported in the medical literature, its incidence is one per 40,000 live births, being more common in men than in women (2).

Conclusion

Prune Berry syndrome continues to be a difficult diagnosis to manage, since it requires a comprehensive approach secondary to the multiple anomalies that the pathology represents; however, in recent years the prognosis of patients with this diagnosis has been improving since there are a variety of diagnostic methods that allow the clinician to provide a better approach depending of each patient (2). The diagnostic approach of all patients with a presumption of this diagnosis should include a simple abdominal x-ray as an initial method that allows evaluation of the abdominal wall; abdominal ultrasound to establish the appearance of renal anomalies and testicular ultrasonographic evaluation to demonstrate the presence or absence of testicles within the scrotal bag (3).

In recent years, abdominal tomography and magnetic resonance imaging have been taken into

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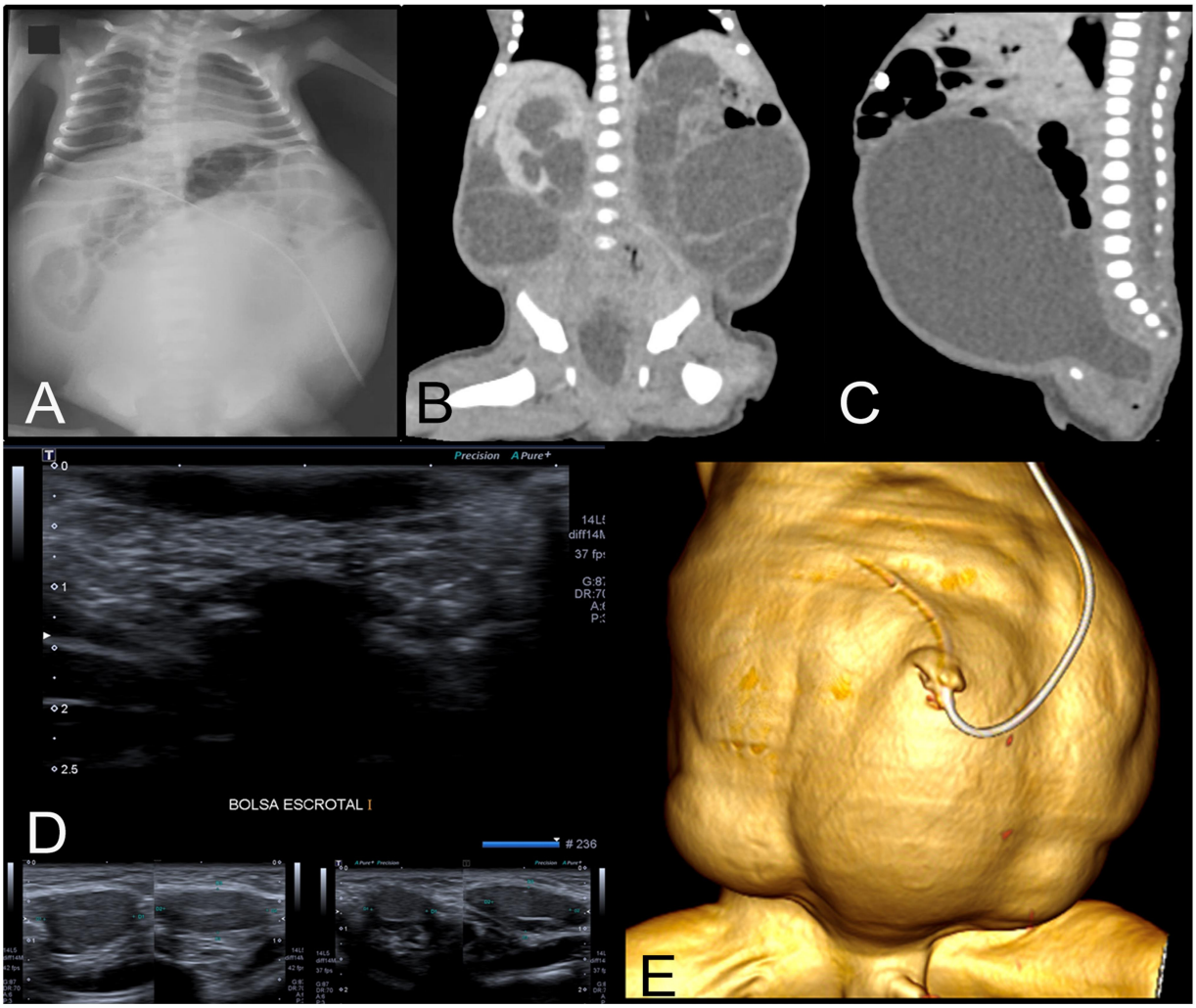

Figure 1. A. Plain Radiograph. Bulging abdomen due to lack of abdominal wall muscles. B. Coronal section tomography. Bilateral hydroureteronephrosis with extremely dilated and tortuous ureters. C. Sagital section tomography. Enlarged urinary bladder. D. Testicular Ultrasound. The testicles are not inside the scrotum. The testicles are located in the middle third of their respective inguinal canal. E. 3D reconstructions. Anterior abdominal wall with ¨Prune-Belly¨ appereance.

account since both sectional studies allow us to evaluate all the anomalies together, the possible complications and also collaborate with the clinician to establish a therapeutic algorithm that gives the patient a better prognosis (4).

Conflicts of interests

There was no conflict of interest during the study, and it was not funded by any organization.

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