Surgical management of complicated bladder exstrophy with genital and bladder duplication


ABSTRACT

Introduction: Complicated bladder exstrophy can be divided into pseudoexstrophy, covered bladder exstrophy, bladder fissure, and double bladder. Double bladder is extremely rare and consists of two bladder chambers separated by a complete fibromuscular wall, with or without double urethra. It is frequently accompanied by congenital abnormalities. Each bladder receives an ipsilateral ureter and drains into an independent urethra. The objective of this article was to present a case of complicated bladder exstrophy with genitourinary duplication, hydronephrosis, and vesicoureteral reflux, along with its management.

Clinical case: Patient is a twenty-year-old woman with past medical history of bladder exstrophy with spontaneous closure. Physical examination revealed ambiguous genititals and double clitoris, double vagina, and double uterus. Patient presented with right flank pain and palpable mass. Computed tomography scan showed right renal hydronephrotic sac, two bladders with independent urethras, and uterus didelphys. Cystography

RESUMEN

Introducción: La extrofia vesical complicada se puede dividir en seudoextrofia, extrofia vesical cubierta, fisura vesical y duplicación vesical. La duplicación vesical es extremadamente rara, consistiendo en cámaras vesicales separadas con pared fibromuscular completa, acompañada o no de duplicación de la uretra, frecuentemente se acompaña de anomalías congénitas. Cada vejiga recibe un uréter ipsilateral y drena a una uretra independiente. El objetivo de este trabajo fue presentar un caso de extrofia vesical complicada, con duplicación genitourinaria hidronefrosis, refluo vesicoureteral y su manejo.

Caso clínico: Femenino de 20 años de edad con el antecedente de extrofia vesical con cierre espontáneo. A la exploración física, se encontró la presencia de genitales ambiguos duplicación de clitoris, vagina y uretra. Presentaba dolor en flanco derecho y masa palpable. La tomografía axial computada mostró bolsa hidronefrótica del riñón derecho, dos vejigas con uretras independientes y útero didelfo. En la cistografía, se evidenció duplicación vesical completa y presencia de refluo vesicoureteral derecho. Se
revealed complete double bladder and the presence of right vesicoureteral reflux. Cystoscopy correlated non-communicating bladders and independent urethras. Complete double genitourinary system was diagnosed along with vesicoureteral reflux and functional exclusion of right kidney. Right nephroureterocystectomy was carried out and patient progression was satisfactory.

**Conclusions:** Fewer than fifty cases of double bladder have been reported and duplication can be complete or partial and sagittal or coronal. There is genitourinary duplication in the majority of cases. Other associated abnormalities are double penis, double vagina, and double uterus. Surgical correction is the treatment of choice in the majority of patients, providing good results and favorable prognosis.

**Keywords:** Bladder exstrophy, double bladder, treatment, Mexico.

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**INTRODUCTION**

Bladder exstrophy is a serious closure defect in the fetal abdominal wall that affects 1 in 30,000 neonates. There are different forms of complicated bladder exstrophy, such as cloacal exstrophy, characterized by two hemibladders and double genitals; in bladder pseudoexstrophy, or ectopic bladder, skin covers a prolapsed bladder; covered bladder exstrophy is characterized by a thin layer of skin covering the bladder; bladder fissure in which there is an infraumbilical defect and communication with an intact bladder that communicates with a normal urethra; and double bladder.1,2 Double bladder is extremely rare. It consists of separate bladder chambers with complete fibromuscular wall. Double urethra may or may not be present. This anomaly is more frequent in men than in women, with a 2:1 ratio, and there are often accompanying congenital abnormalities.1-3 A total of 45 cases have been reported in the medical literature, with genital abnormalities associated with 90% of cases and gastrointestinal abnormalities in 40% of cases.3 Each kidney drains urine through an independent ureter to the ipsilateral bladder, each of which has an independent urethra.2,3 The most accepted embryologic theory is that of urogenital septum persistence with a widening of the cloaca, dividing the endodermic elements and forming two urachuses, two bladders, and two urogenital sinuses. Duplication is generally diagnosed in infancy, correllacionó con cistoscopia vejigas no comunicantes y uretras independientes. Se diagnosticó doble sistema genitourinario completo, con reflejo vesicoureteral y exclusión renal derecha. Se realizó nefroureterocistectomía derecha. La paciente evoluciona satisfactoriamente.

**Conclusiones:** La duplicación vesical es una entidad con menos de 50 casos reportados, esta puede ser completa o parcial, sagital o coronal. En la mayoría de los casos, existe duplicación genitourinaria. Existen otras anormalidades asociadas como son la duplicación de pene, vagina y útero. La corrección quirúrgica es el tratamiento de elección en la mayoría de los pacientes, con buenos resultados y pronóstico favorable para el paciente.

**Palabras clave:** Extrofia vesical, duplicación vesical, tratamiento, México.
due to recurrent urinary infections. Bladder duplication can occur at the coronal or sagittal plane, the latter being the more common, in which each bladder is separated by peritoneum.

The objective of the present case was to show a case of complicated bladder exstrophy with genitourinary duplication, hydronephrosis, grade IV vesicoureteral reflux, and to describe its management.

**CASE PRESENTATION**

Patient is a 20-year-old woman with past medical history of bladder exstrophy with spontaneous closure. Physical examination revealed duplication of internal and external genitals (clitoris, vagina, and uterus), as well as lack of fusion of pelvic bone (Figure 1). Patient sought medical attention at the urology department for right flank pain and palpable mass. Computed tomography (CT) scan revealed right hydronephrotic sac and the presence of 2 bladders with independent urethras and double uterus (Figures 2 and 3). Magnetic resonance imaging (MRI) identified 2 non-communicating bladders with interface between the two (Figures 4 and 5). Karyotype 46XX was reported by the genetics department. Cystography showed complete bladder duplication and grade IV right vesicoureteral reflux. Cystoscopy with 30° pediatric lens revealed hemibladder in the right urethra with dilated orthotopic ureteral meatus. The left meatus could not be visualized and neither could connection to left hemibladder. The same procedure was carried out in the left urethra, visualizing orthotopic and ejaculating ureter. Complete double genitourinary system was diagnosed with grade IV vesicoureteral reflux and right kidney exclusion. Right nephroureterocystectomy was programmed.

Lumbotomy and extraperitoneal dissection were carried out, revealing dilated right ureter with severe hydronephrosis and no residual kidney parenchyma. Dissection of distal ureter and right hemibladder was continued by means of Gibson incision, identifying both hemibladders and peritoneum and fibromuscular tissue (Figure 6). Patient was released on fourth postoperative day with no complications.

**DISCUSSION**

In 1961, Abrahamson classified double bladder as complete and incomplete. There are two bladders in complete double bladder that are separated by mucosa and muscle layer and each one drains into an independent urethra, as in the present case. In incomplete double bladder there is communication between the two bladders and a single common urethra. 

3 Diagnosis is made by means of bilateral retrograde urethrocytography. Bladder duplication can occur at the sagittal or coronal plane, depending on the axis of...
the septum. The sagittal variant is apparently more common than the coronal, with a 2.5:1 ratio. According to the medical literature, double bladder is more common in men, with a 2:1 ratio. Kossow and Morales reviewed 40 cases, finding gastrointestinal malformations in 42% of cases and genital malformations in up to 90% of cases. Berrocal found genital malformations to be dependent on the axis of the intervesical septum, frequently associated with the coronal plane. Since the first descriptions of double bladder, a total of 45 cases have been reported. The most accepted embryologic theory continues to be that of urogenital persistence and cloacal division proposed by Abrahamson. A variety of bladder duplications are found in the literature and so there is no standard surgical management. Each case must be evaluated individually. In the present case nephroureterocystectomy was carried out because the bladder was nonfunctioning, causing reflux and kidney disorders.

Figure 4. Magnetic resonance image identifying two non-communicating bladders with interface between them.

Figure 5. Another magnetic resonance image showing two non-communicating bladders with interface between them.

Figure 6. Surgical specimen from nephroureterectomy.
exclusion. This is the first documented case in the Mexican urologic literature.

REFERENCES