Bilateral vesicoureteral reflux secondary to anterior urethral valves: a new review


ABSTRACT

The presence of valves in the anterior urethra is a rare pathology, with only a few cases reported in the national and international literature. Patients present with recurrent urinary infections or urinary obstruction data that can be serious. The presence of valves in the anterior urethra is an uncommon congenital malformation but it can cause significant damage to the rest of the urinary tract above the obstructive process. Anterior valves are more frequently located at the bulbar level and are associated with urethral diverticuli. Clinical manifestations are secondary to the obstructive process and can have mild to severe urodynamic repercussions. Treatment is always surgical, and can be endoscopic or open surgery. The aim of medical attention in these patients is to have enough elements to make diagnosis and to perform opportune and adequate surgery for the obstructive uropathy and its consequences. The objective of the present article is to present the case of a patient with anterior urethral valves and urodynamic repercussions resulting in secondary bilateral megaureter and to report on management and present patient progression.

RESUMEN

La presencia de valvas en la uretra anterior es una patología poco común, con escasos casos publicados en la literatura nacional e internacional. Los pacientes cursan con infecciones urinarias de repetición o datos de obstrucción urinaria que pueden ser graves. La presencia de valvas en la uretra anterior es una malformación congénita poco frecuente, pero puede causar un daño trascendente al resto de la vía urinaria por arriba del proceso obstructivo. Los lugares que con mayor frecuencia se localizan las valvas anteriores son a nivel bulbar y se asocian a divertículos de la uretra. Las manifestaciones clínicas son secundarias al proceso obstructivo que provoquen ya sea leve a severo con sus repercusiones urodinámicas. El tratamiento de las mismas siempre será quirúrgico ya sea por vía endoscópica o cirugía abierta. El contar con los elementos suficientes para realizar diagnóstico, y un tratamiento quirúrgico oportuno y adecuado de la uropatía obstructiva y de las secuelas de la misma, es el objetivo en la atención de estos pacientes. El objetivo del estudio es presentar un caso de paciente con valvas uretrales anteriores, con repercusión urodinámica, condicionando megaureter.
Key words: Valves, anterior urethra, Mexico.

Palabras clave: Valvas, uretra anterior, México.

INTRODUCTION

The first descriptions of diverticuli in the anterior urethra were made in 1906 and 1917 by Watts and Engleard. It was not until 1969 that the first large series of 17 cases of anterior urethral valve (AUV) was reported by Williams and Retik. This series was followed by 13 cases in 1978, 18 in 1982 and 14 in 1985. In 1960 an attempt was made to isolate AUV from an obstructive diverticulum. In 1981, Lewis found only 30 AUV cases reported in the medical literature published in English. 1

Anterior urethral valve or valves (AUV) is a rare congenital disorder. Its incidence is 1 in 5000-8000 births.2 AUV are less frequent than posterior urethral valves and often are not discovered due to their rareness. 1,2 Etiology is uncertain but they may be the result of a failed attempt at urethral duplication.4 Kusuda and Das suggested that valves may represent an early event in urethral duplication development.5 Another hypothesis is that these valves occur due to cystic dilatation of the periurethral glands that are joined to the urethra as a result of the formation of folds.6 Although they are called valves, these obstructive structures often present in the form of a diverticulum or accompanied by a urethral diverticulum.

In AUV, the urethra has a secular or bulbar dilatation. They are generally produced when there is a defect in the corpus spongiosum, leaving a thin urethral wall. This segment of the urethra distends during micturition, simulating a mass that is sometimes visible along the ventral wall of the penis. It is estimated that the valves in a third of AUV patients are associated with a diverticulum that presents as a lump at the root of the penis after micturition and when compressed results in a dripping of urine from the meatus.7,8 Some authors state that the most common localization is in the distal urethra. 8 Others refer to the anatomical presence of anterior valves in the urethral tract in percentages, documenting more in the bulb urethra (40%), at the junction of the penile urethra and bulb urethra (30%), and in the penile urethra (30%). It is not easy to radiologically demonstrate AUV, especially if they are not associated with urethral diverticuli.7,9 Approximately 70% of AUV are peak or half moon-shaped and 30% are iris-shaped.9

In addition to diverticuli, differential diagnosis includes syringoceles, Cowper’s duct abnormalities and megalourethra.10 Anatomically, differential diagnosis can be made or associated with urethral diverticulum, which is a saccular process that communicates predominantly with the urethral opening (Image 1). The lesion is on the ventral side and varies in size (3 - 5 cm in diameter). In the neck it is not clearly marked except in its distal portion where it can be a very developed labium or spur that becomes the lesion obstruction. The diverticulum is confined by a urothelium-covered fibrous wall. Valves present as mucosal folds in the diaphragm, or more often as a dome, that is generally found along the length of the ventral surface of the urethra (Image 2). This fold enlarges during micturition and is flattened against the urethral roof, becoming a potentially serious obstacle. The effect on the proximal urethra is variable and appears to be separate from bladder and upper urinary tract repercussions.1

AUV have been reported less frequently than posterior urethral valves, at a ratio of 1:10. This urethral anomaly can lead to varying urinary tract symptoms depending on the patient, age, and degree of obstruction. Clinical symptoms are varied, from irritative bladder symptoms, hematuria, urinary infection, reduction of urinary strength and caliber, dripping, urinary retention, incontinence, enuresis, to terminal kidney failure. Urethral obstruction can lead to detrusor hyperactivity and partial imbalance of the sphincter, predisposing the patient to incontinence or nocturnal enuresis. Symptoms are principally dependent on age at time of presentation and degree of obstruction.4

Micturition cystourethrography is the study of choice for this pathology, revealing anatomical characteristics of the bladder and urethra. Today videourodynamic studies can be done to show the connection between the anatomy and the functioning of the urinary tract and the pelvic floor. Clinical evaluation should be accompanied by renovesical echogram, cystourethrogram, excretory
urography, and urethrocystoscopy. The treatment of choice is endoscopic surgery with valve ablation using transurethral electrofulguration, cold knife, or laser. The objective of the procedure is to achieve valve ablation and free urinary flow.

Undesired treatment consequences can be urethral lesion with complications such as fibrosis and urethral stricture. If equipment for endoscopic ablation is not available, open valve resection is an equally good alternative. Other options are vesicostomy in children with severe vesicoureteral reflux, urethroplasty in cases involving the urethra, and diverticulectomy in results associated with diverticulum.

Savage et al suggested a treatment algorithm based on disease seriousness. They recommend vesicostomy in babies not yet eating solid foods that present with high grade reflux and emptying of the urinary tract. If urethral caliber is sufficient, transurethral fulguration is recommendable. Open urethroplasty is useful in patients with large urethral diverticulum and thin urethra.

**CLINICAL CASE PRESENTATION**

Patient is a four-month-old male infant, product of a first pregnancy. Mother presented with mild preeclampsia and oligohydramnios. Prenatal ultrasound detected left pyelocaliectasis at sixth month of pregnancy. Patient was delivered by cesarean section at 37 weeks of gestation, weighed 2450 g, and was 49 cm long, and cried and breathed at birth. He was hospitalized for 10 days at three months of age due to urosepsis, with fever of 39°C. During hospital stay he presented with leukocytes, elevated serum creatinine and urea, and positive urine culture with 800,000 *Candida albicans*.
colonies. He was released due to clinical improvement.

When admitted prior to surgery, patient did not manifest symptoms although he occasionally cried during micturition and had reduced urinary stream caliber. Physical examination showed patient weighed 5.4 kg, was 52 cm long, and had slight, non-painful bulkiness in left abdominal quadrant. There were no other alterations. Preoperative laboratory results: complete blood count, full blood chemistry, serum electrolytes, and prothrombin time were normal. Urinalysis: pH 6, density 1.005, leukocytes: 15 per field, scant bacteria. Urine culture: 500,000 colony forming units of E. coli.

Radiology: Plain film of the urinary tract with no evidence of pathology.

Ultrasound: Right kidney 7.1 cm x 3.6 cm, 8 cm parenchyma with pyelocaliectasis. Left kidney: 7.7 cm x 3.7 cm, 6 cm parenchyma, both kidneys with ureteropelvicaliectasis (Images 3 and 4). Bladder with 0.5 cm thick mucosa but no other echographic alterations except dilatation of left ureter at ureterovesical junction (Image 5).

Urethrocystogram: showed left grade V high and low pressure vesicourethral reflux, right grade II high pressure, bladder diverticuli image, “pumping” at anterior urethra level with stagnation of contrast medium (Images 6, 7 and 8). Surgical exploration was decided on in order to carry out urethrocystoscopy and bilateral ureteral reimplantation.

Urethrocystoscopy: Permeable, anterior urethra with iris-shaped anterior valves at the penoscrotal junction. Permeable posterior urethra, bladder with grade III trabeculations, meatuses in position, both in position A; horseshoe-shaped right meatus and golf hole-shaped left meatus. Both were permanently open (Image 9).

Surgical findings: Left ureter was 16 cm in length and 3 cm in diameter and right ureter had an extravesical
length of 5 cm and a 2 cm diameter. Anterior valve fulguration and bilateral ureteral restoration with Kalicinski technique for the right side and Hendren technique for the left side were performed. Bilateral ureteral Hendren-type reimplantation was carried out (Image 10). Cystostomy was left in.

Early diagnosis and treatment had satisfactory results. The obstructive process was resolved with anterior valve fulguration with Bugbee electrode and both ureters were repaired with bilateral restoration and reimplantation. Up to the present date the patient has good transurethral micturition caliber, kidney function is conserved, and his general condition is good.

**DISCUSSION**

Success with this pathology is achieved not only through detection and diagnosis of the obstructive process but also through adequate urinary diversion and definitive attention in time to avoid urodynamic repercussions prior to obstruction. Often urethral valve elimination is not enough.

**CONCLUSIONS**

Obstructive congenital pathologies of the anterior valves (valves, diverticuli) are rare and can be difficult to diagnose. The authors reviewed the existing international reports on AUV. Anatomical interpretation of these lesions is far from unequivocal and it is necessary to distinguish between valves and diverticuli. The main difference lies in abnormality and corpus spongiosum adjacency. Clinical presentation is age-dependent and diagnosis depends essentially on cystourethrogram. Generally, valve treatment is simple and is carried out with endoscopic resection-fulguration. It is not always necessary or recommendable to resect diverticuli. After surgery the patient should present with clinical
improvement and improved urinary flow. Long-term follow-up in which micturition parameters are checked is recommended to opportently identify possible urethral obstruction as a consequence of treatment.

BIBLIOGRAPHY