



Priapism in a 4-year-old boy with late presentation posterior urethral valves: A case report

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ABSTRACT

A four-year-old boy presented painful episodes of priapism always before emptying the bladder since he was two years old, which led to investigation for a possible arteriovenous fistula of the perineal vascular system or a cavernous artery hyper flow. Following different exams in an attempt to establish the diagnosis, the child, actually, was found to have posterior urethral valves. The diagnosis was totally unforeseeable and the urethral valves endoscopic treatment reduced the priapism events with a positive effect on the disease.

Key Words: Posterior urethral valves, priapism, urethra abnormalities, late diagnostic, treatment outcome, child.

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Introduction

Priapism, considered a medical emergency, is a relatively rare condition. Priapism is defined as a condition in which the penis remains erect for hours either in the absence of stimulation or after the stimulation has ended. Patients presenting with ischemic priapism require immediate intervention in an attempt to mitigate the symptoms. High-flow priapism, also known as arterial priapism, resulting from an exaggerated flow in the cavernous artery, is not always an emergency and at first its treatment requires nothing but clinical observation [1,2].

Posterior urethral valves (PUV) constitute the most common cause of lower urinary tract obstruction in boys, most commonly leading to chronic urinary tract disorder in children.

Children with late posterior urethral valves manifestation, as a rule, have voiding dysfunction, enuresis, urinary frequency, recurrent orchiepididymitis, or a history of recurrent urinary tract infections [3].

We report the case of a four-year old child with posterior urethral valves. The event that prompted parents to seek medical attention was a nosebleed. The investigation carried out by a hematologist did not reveal any precise cause of the bleeding, but it called his attention to the existence of many episodes of priapism before bladder emptying since he was two years old. Apparently, this is the first reported case of a child with posterior urethral valves that presents frequent priapism events as main manifestation.

Case report

A 4-year old boy sought medical care due to a nosebleed. He was seen by a hematologist who found no cause for nosebleed but decided to investigate a maternal complaint that the child had been experiencing daily and frequent episodes of painful priapism before emptying his bladder, since the age of two (Fig. 1).



Fig. 1. Penile erection (priapism) of our 4-year-old patient before urinating.

A Magnetic Resonance Angiography (MRA) was performed but showed no arteriovenous fistulas. The radiologist, however, did mention the existence of a “prostatic utricle cyst”. A voiding cystourethrography showed a narrowing in the region of the posterior urethra, no image of cyst, and no vesicoureteral reflux (Fig. 2).



Fig. 2. Narrowing at the transition from the posterior urethra to the anterior urethra without vesicoureteral reflux (arrow).

An urethrocystoscopy was then performed under anesthesia using a 10.8 Fr cystoscope sheath and a 0° telescope, surprisingly revealing an image of a posterior urethral valve, and the bladder with thickened and trabeculated walls (Fig. 3).



Fig. 3. Posterior urethral valves with valve flaps evolving from the verumontanum to the anterior wall of the urethra.

He also had an enlarged prostatic utricle with no cyst observed. After identifying the valves, a 600 U quartz bare tip end-firing laser fiber was passed through the side channel of the cystoscope and brought in direct contact with the valves that had been fulgurated. The laser power setting ranged from 20 to 25 W while the total energy delivered ranged from 1000 to 2000 J. Fulguration was done at the 5, 7 and 12 o'clock positions.

He had normal levels of creatinine and BUN; normal DTPA and DMSA, no signs of obstruction or renal scars. During the initial one-year of follow-up an ultrasonography of the urinary tract was performed showing both kidneys to have a discreet hydronephrosis, the bladder with thickened walls; however, after voiding, the urinary residue was negligible. With the valves ablation the erection events slowly reduced; currently they occasionally occur when the child wakes up, in the morning.

He now comes back every six months for follow-up.

Discussion

PUV are thought to develop in the early stages of fetal development [4]. If the diagnosis is not made in the first months of life, the children usually have enuresis (60%), urinary frequency (17%), infections of the urinary tract (17%), or even, arterial hypertension [3].

The existence of episodes of priapism led the hematologist who decided to investigate the cause of the episodes of priapism to perform an angioresonance that revealed nothing but the existence of a possible utricle cyst. As episodes of priapism always occurred before emptying the bladder, we decided to perform a voiding cystourethrography. A voiding cystourethrography was performed to investigate the possible existence of an utricle cyst, which could be compressing the posterior urethra. We did not find any utricle cyst but a narrowing of the posterior urethra (Fig. 2), which made us perform a cystourethroscopy, which revealed the existence of posterior urethral valves (Fig. 3), with their typical insertion at the base of the verumontanum, in addition to a thickened and trabeculated bladder.

Venous priapism is a medical emergency and is the result of an obstruction of the corpora cavernosa of the penis, usually caused by altered red blood cells that prevent normal blood flow. In such cases, one should intervene as early as possible to prevent the formation of fibrosis of the corpora cavernosa and a consequent future erectile dysfunction. Arterial priapism is caused by an increase in blood flow to the cavernous artery. It occurs in general through the existence of an arteriovenous fistula caused in general by an accident involving the perineal region [2].

We hypothesize that the urinary retention typical of the existence of posterior urethral valves may have had the same effect on the corpora cavernosa that morning urinary retention causes in normal individuals and may have caused repeated episodes of priapism. This can be considered an unusual case in which the posterior urethral valves diagnosis was made based on frequent priapism events, probably resulting from bladder retention. This is one more unusual manifestation of posterior urethral valves which urologists should be aware of.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Patient confidentiality has been maintained and written consent has been obtained from the patient's parents for the publication of patient information and clinical pictures and can be provided as required.

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