

What urethrogram sees and a surgeon does not

P. Salcedo Arroyo¹, C. Domínguez García², M.R. Delgado Alvira¹, J. Pisón Chacón¹, Y. González Ruiz¹, M. González Herrero¹, P. Bragagnini Rodríguez¹, M.V. Cobos Hernández³

¹Pediatric Surgery Department. Miguel Servet University Hospital. Zaragoza (Spain). ²Pediatric Surgery Department. Torrecárdenas ³University Hospital. Almería (Spain). Radiodiagnosis Department. Pediatric Radiology Unit. Miguel Servet University Hospital. Zaragoza (Spain).

ABSTRACT

Introduction. Urethrorrhagia is an infrequent sign in childhood. It should be distinguished from hematuria, since they have a different etiology.

Clinical case. 11-year-old male patient with significant urethrorrhagia. Urinary sediment analysis: red blood cells++. Pelvic ultrasonography: fusiform anechoic image in the corpus spongiosum of the penile root. Retrograde urethrogram: normal anterior urethra, extraluminal contrast passage in the ventral aspect of the bulbar urethra. Cystoscopy: no pathological findings in the urethra or the bladder. Control retrograde urethrogram: cystic dilatation of Cowper's gland duct; Maizels' type 3 perforated syringocele.

Discussion. Cowper's syringocele is a rare pathology. It can occur at any stage of childhood in the form of urinary infection, obstructive voiding symptoms, or urethrorrhagia. Urethrogram is key for diagnostic purposes, since most Cowper's syringoceles are detected following urethrogram or cystoscopy. Cases with functional repercussions for the urinary system require surgical treatment. Otherwise, a wait-and-see approach is feasible.

KEY WORDS: Urethrorrhagia; Hematuria; Bulbourethral glands; Child.

LO QUE UNA URETROGRAFÍA VE Y UN CIRUJANO NO

RESUMEN

Introducción. La urethrorragia es un signo infrecuente en la infancia que debe distinguirse de la hematuria dada la diferente etiología de las mismas.

Caso clínico. Varón de 11 años con urethrorragia franca. Sedimento urinario: hematíes++. Ecografía pélvica: imagen anecoica fusiforme en cuerpo esponjoso de raíz peneana. Uretrografía retrógrada: uretra anterior normal, paso de contraste extraluminal ventral

en uretra bulbar. Cistoscopia: sin hallazgos patológicos en uretra ni vejiga. Uretrografía retrógrada de control: dilatación quística del conducto de las glándulas de Cowper; siringocele perforado tipo 3 de Maizels.

Comentarios. El siringocele de Cowper es una patología infrecuente que puede debutar en cualquier momento de la infancia como infección urinaria, síntomas miccionales obstructivos o urethrorragia. La uretrografía es fundamental en su diagnóstico ya que la mayoría se objetivan por este medio o cistoscopia. Los casos con repercusión funcional del sistema urinario requieren tratamiento quirúrgico. En caso contrario podrá realizarse actitud expectante.

PALABRAS CLAVE: Urethrorragia; Hematuria; Glándulas bulbouretrales; Niño.

INTRODUCTION

Macroscopic hematuria is defined as an increase in red blood cells in the urine visible to the human eye, whereas urethrorrhagia is defined as urethral bleeding. In the pediatric population, it is important to distinguish hematuria from urethrorrhagia, as well as asymptomatic hematuria from symptomatic hematuria, glomerular hematuria from extraglomerular hematuria, and self-limited progression from persistent progression.

The most frequent causes of glomerular hematuria include post-infectious glomerulonephritis, IgA nephropathy, etc. Urinary tract infection, urethral-perineal inflammation, and trauma are the most common causes of extraglomerular hematuria⁽¹⁻³⁾. Urethrorrhagia used to be interpreted as a self-limited, benign sign⁽⁴⁾, with urinary tract infection, stenosis, and urethral tumors being the most frequent causes⁽⁵⁾.

Syringocele is a cystic dilatation of Cowper's bulbourethral glands that may cause urethrorrhagia. It is typically more frequent in the pediatric population, but cases in adulthood are increasing. The real incidence of syringocele in the general population is unknown. The largest series of cases published so far –Wagemans et al.⁽⁶⁾– includes 122

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Corresponding author: Dra. Paula Salcedo Arroyo.

E-mail address: paulasalcedo93@gmail.com

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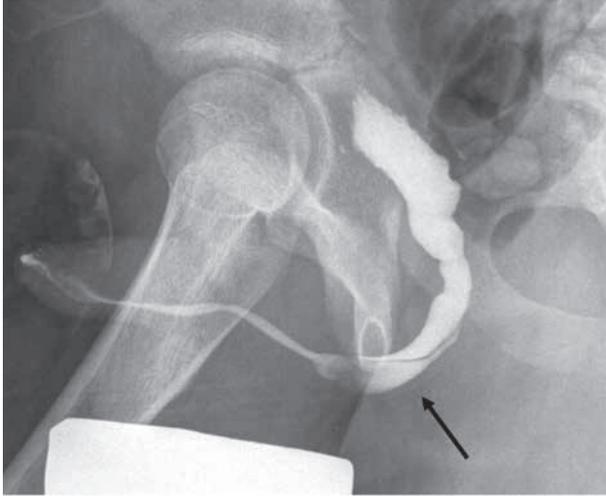


Figure 1. Retrograde urethrogram at diagnosis: saccular urethral structure (black arrow).

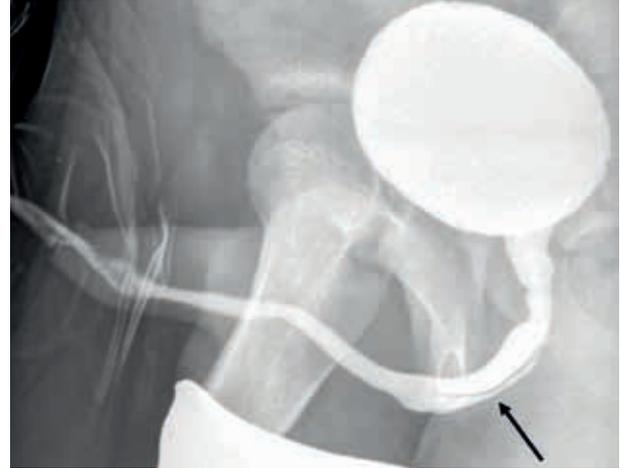


Figure 2. Control retrograde urethrogram at month 6: urethral cystic dilatation compatible with perforated syringocele (black arrow).

patients managed at a third-level hospital over 25 years. Various classifications are available. Maizels et al.⁽⁷⁾ and Bevers et al.⁽⁸⁾ published a description and a classification based on anatomical differences, whereas Campobasso et al.⁽⁹⁾ divided them into non-obstructive (perforated/open) and obstructive (imperforated/simple).

CLINICAL CASE

11-year-old male patient with significant urethrorrhagia during voiding and unrelated spotting associated with dysuria and pollakiuria, without trauma history. At physical exploration, external genitals looked normal, with pain upon perineal palpation. Urethrorrhagia was evident. Rectal examination: the anterior aspect of the rectum was palpated, with inflammation and pain signs. No further abnormalities were observed.

Additional tests:

- Blood test: Normal.
- Urinary sediment analysis: Hemoglobin ++. No further abnormalities were observed.
- Pelvic ultrasonography: Fusiform anechoic image seemingly located in the corpus spongiosum at the level of the penile root.
- Cystourethrogram: Anterior urethra with normal morphology and extraluminal contrast passage in the ventral portion of the bulbar urethra. The voiding study showed greater extraluminal contrast passage towards the saccular structure compressing the urethral lumen. It could be either incomplete urethral duplicity or urethral diverticulum.
- Cystoscopy: Normal-sized urethra, with no lesions or active bleeding areas. Slight mucosal congestion. Normal bladder.

Following cystoscopy, urinary stream was abundant, without difficulty, and without dysuria. The patient had several episodes of urethrorrhagia, especially in the first 24 hours, and subsequently, occasional spotting was observed. Outpatient controls were carried out 6 and 12 months later, with the patient experiencing no symptoms. A new cystourethrogram was performed at month 6. It confirmed the presence of a saccular formation in the ventral aspect of the bulbar urethra, which continued in the form of a duct running into the membranous urethra. It was visible in both retrograde and voiding urethral images. Findings were compatible with cystic dilatation of Cowper's gland duct or type 3 perforated syringocele according to Maizels' classification⁽⁷⁾. Since the patient had no symptoms, a wait-and-see approach was decided upon. Clinical follow-up has been carried out for 16 months so far.

DISCUSSION

Extraglomerular hematuria should be confirmed by means of a urine test strip and a urinary sediment analysis to demonstrate the presence of red blood cells in the urine. Following full anamnesis and physical exploration, and as a previous step to etiological diagnosis, various additional explorations can be conducted, such as urinary sediment analysis and urine culture in case of suspected urinary infection, imaging tests like CT-scan in case of trauma, or renal ultrasonography in case of suspected lithiasis or anatomical abnormalities of the urinary tract. Diagnostic cystoscopy – which can be performed on an outpatient basis in adults but requires general anesthesia in children – should only be used in cases of hematuria with ultrasound visualization of bladder tumor, persistent symptoms of inflammatory cystitis, or history suggestive

of urethral involvement following trauma⁽¹⁰⁾. In up to one third of cases, etiological diagnosis of hematuria is not achieved^(1,2,4).

Clinically speaking, Cowper's gland syringocele can manifest as urinary tract infection or obstructive urinary symptoms, dysuria, post-void urine leakage or residual urine, urethrorrhagia, and perineal pain^(11,12). At early stages, it has also been described as difficulty for bladder catheterization⁽¹³⁾. Symptoms vary according to age (patients under vs. over 4 years old) and type of syringocele (non-obstructive vs. obstructive)⁽⁷⁾.

Diagnosis is usually confirmed by urethrocystoscopy, while cystourethrogram can support suspected diagnosis⁷. Perineal ultrasonography, NMR, and CT-scan are useful in case of imperforated syringocele, whereas flowmetry is useful for the assessment and follow-up of obstructive symptoms^(11,12).

Therapeutic options include loop diathermy opening under direct visualization through urethroscopy⁽¹⁴⁾, laser ablation⁽¹⁵⁾, and open perineal resection^(16,17). Given that spontaneous regression has also been reported, some authors advocate a wait-and-see approach^(7,11,12,18).

In our case, the physical exploration of the patient and the clinical characteristics of urethrorrhagia made us consider urethral pathology from the beginning. The ultrasound visualization of a saccular urethral structure suggested a voiding cystourethrogram should be performed as the next step to confirm diagnosis. Urethrocystoscopy ruled out secondary urethral or bladder involvement, which led us to opt for an approach based on clinical observation.

Since the patient had no history of urinary tract infection or obstructive symptoms, and clinical progression was good – with the patient presenting no symptoms at controls –, follow-up continuation and a wait-and-see approach were considered. After reviewing the literature published, and in agreement with his family, this approach was decided upon.

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