Bilateral Giant Orthotopic Ureterocele Appearing as Kissing Cobra in a Nigerian Child

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ABSTRACT

The cobra head sign is classically seen when a unilateral intravesical ureterocele is present. This type of ureterocele is also termed orthotopic. Orthotopic ureteroceles are usually unilateral and commoner in females, but bilateral in 10% of cases.The cobra head sign is characterised by bulbous dilatation of the distal end of the ureter with a surrounding radiolucent halo seen within the contrast-filled urinary bladder on intravenous urograms. Although, the cobra head sign is classic for an intravesical ureterocele, only about 50% of cases show this sign. This is a rare case of an eleven-year old boy with giant bilateral ureterocele touching each other on intravenous urogram, hence appearing as 'kissing cobra'.

Key words: Kissing cobra; orthotopic; ureterocele

Introduction

Ureterocele is a cystic dilatation in the terminal section of the ureter that is located inside the bladder, the urethra, or both.[1] It may be unilateral or bilateral and may be associated with a single or duplex system in some cases.[2,3] The incidence is reported to be between 1 in 5,000 and 1 in 12,000 of pediatric hospital admissions;[1] however, a number of autopsy studies have suggested a much higher incidence of up to 1 in 500.[2]

It occurs 4–6 times more frequently in girls and is observed almost exclusively in the Caucasian population.[4,5] Ureteroceles have no predilection for side and 10% of cases show bilateralism.[4,6]

The cobra head sign is characterized by bulbous dilatation of the distal end of the ureter with a surrounding radiolucent halo, seen within the contrast-filled urinary bladder on intravenous urograms.[7] The cobra head sign is classically seen with an intravesical ureterocele.[2,8] This type of ureterocele is also termed orthotopic, since it arises from a ureter with a normal insertion into the trigone.[7]

Herein, we present a rare case of an 11-year-old boy with giant bilateral ureterocele touching each other on intravenous urogram, hence, appearing as 'kissing cobra'.

Case Report

J.A. is an 11-year-old boy referred from a private hospital on account of abdominopelvic ultrasound (US) diagnosis. He presented at a private hospital about two months ago, on account of recurrent abdominal pain of one-month duration and fever of 2-days duration. There was no history of urinary symptom. He had a US done which revealed a right ureterocele and it was on this basis that he was referred. Physical examination was essentially normal. No significant finding was found on system examination.

Repeat abdominopelvic US shows bilateral hydrocalycoses worse on the right and two round anechoic cystic structures closely apposing each other were also demonstrated within the urinary bladder [Figure 1]. The intravenous urogram (IVU) showed bilateral hydrocalycoses and hydroureters with distal dilatation and clubbing touching each other in the urinary bladder [Figure 2].

A diagnosis of bilateral giant ureterocele was made and was managed surgically to relieve the obstruction with the aid of endoscopy.

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Quick Response Code:	Website: www.wajradiology.org
	DOI: 10.4103/1115-1474.146151

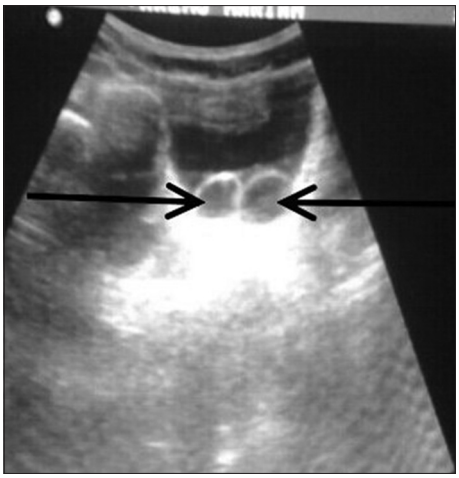

Figure 1: Pelvic sonogram (transverse scan) showing two round well defined echo-free structures closely apposing each other within the urinary bladder consistent with bilateral ureterocele (black arrows)

Discussion

A ureterocele refers to a cystic dilatation of the distal ureter protruding into the bladder lumen.[1,2] This malformation is most commonly diagnosed in the pediatric population for two reasons: Firstly, due to prenatal ultrasound screening for congenital anomalies and secondly, due to the high incidence of post-natal urinary tract infections, resulting in ultrasonography of the renal tract as a first-line investigation.[2]

The incidence is reported to be between 1 in 5000 to 1 in 12000 of pediatric hospital admissions;[2] however, a number of autopsy studies have suggested a much higher incidence of up to 1 in 500.[1] They are more common in Caucasian individuals and females, who are affected 4-7 times more often than males.[4] This present case occurs in an 11-year old Nigerian boy. Approximately 10% of ureteroceles occur bilaterally,[4,8] as is the case in this patient.

The American Academy of Pediatrics, classifies ureteroceles contained entirely within the bladder as intravesical or orthotopic, while those with some portion of the ureterocele permanently located at the bladder neck (or rarely urethra) as ectopic.[2] Bilateral orthotopic ureteroceles as found in this case is an uncommon finding.[8]

Single-system ureteroceles are distinguishable clinically from the more common duplex-system ureterocele by their frequent occurrence in male children (in direct contrast to female predominance in duplex-system ureteroceles) and their association with multicystic dysplastic kidney.[2]

This case is also a single system ureterocele seen in a male child with no evidence of dysplastic kidney. Although single system ureteroceles usually manifest in adulthood, they have been reported with increasing frequency in recent years amongst

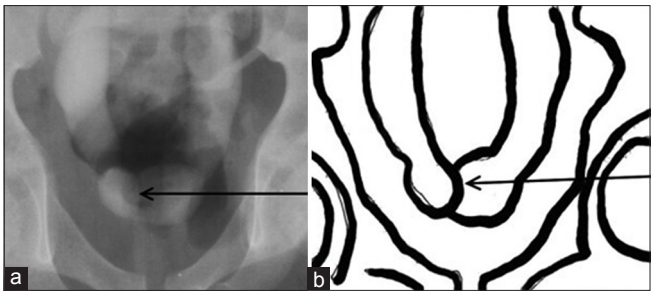

Figure 2: (a) Intravenous urogram (post micturation) showing bilateral dilated distal ureters appearing as "kissing cobra".(b) Line diagram demonstrating the "kissing cobra" appearance

urologists and radiologists in pediatric practice.[2,4] This case is seen in an 11-year-old male child.

Up to 80% of pediatric ureteroceles occur with ureteric duplication anomalies and are termed duplex-system ureteroceles, with 60-80% of these having an ectopic drainage orifice.[2] Ureteroceles vary in size from small, clinically asymptomatic lesions to large lesions, which can obstruct the ipsilateral and contralateral ureters and prolapse into the urethra, causing bladder outlet obstruction.[3] This patient had giant bilateral ureteroceles causing bilateral ureteral obstruction and bilateral hydrocalycoses.

There is no characteristic clinical picture caused by this condition, but fever, pyuria and recurrent abdominal pain are often present in children.[8] The main clinical presentation in this patient was recurrent abdominal pain.

This condition is traditionally diagnosed after urologic investigations triggered by repetitive episodes of a severe urinary tract infection (UTI), although, occasionally, abdominal pain, urinary obstruction, palpable abdominal masses, and lithiasis have been reported as the main initially appearing features.[3,8]

Urinary tract infection is still the most common clinical presentation of ureterocele, in 50% of the patients, prompting physician to make thorough evaluation of the urinary system.[9] However, no evidence of UTI was found in this patient.

Renal and/or bladder US should be the initial study for the evaluation of a neonate with fetal hydronephrosis, with or without a suspected ureterocele.[2]

Every postnatal ultrasonographic evaluation of antenatal hydronephrosis should include examination of the bladder. The sonographic finding of a well-defined cystic intravesical mass within the posterior bladder wall suggests ureterocele.[8] The characteristic finding in ureterocele is a cystic ballooning or spherical dilatation of the lower end of the ureter with associated radiolucent halo (cobra head) as seen on intravenous urography.[10] These features are demonstrated in this present case.

The cobra head sign is an indicator of an uncomplicated ureterocele.[7] However, a cobra-shaped distal end of a ureter may be seen with incomplete distal ureteral obstruction caused by a tumor or calculus.[7] This appearance is termed pseudoureterocele. The appearance of the radiolucent wall surrounding the dilated distal ureteral segment is an important differentiating point between a ureterocele and a pseudoureterocele.[7]

The lucency or halo surrounding a pseudoureterocele is thicker than that of a uterocele and is poorly defined; in cases of tumors, it may be irregular and may show a filling defect within the ureterocele.[11,12] It is also important to note that although the cobra head sign is classic for an intravesical ureterocele, only about 50% of cases show this sign.[7,13]

When ureterocele is symptomatic, treatment can generally be accomplished by cystoscopic transurethral incision at the base of the ureterocele.[4,8] Unroofing of the ureterocele, also accomplished via a transurethral approach, is sometimes necessary to relieve obstruction.[8] A more radical approach may be necessary in cases of ectopic ureterocele associated with non-functioning renal moiety; partial nephroureterectomy may be necessary.[8] Endoscopic treatment is the standard modality for bilateral ureteroceles.[4]

In conclusion, we have presented a rare case of an 11-year-old male with bilateral giant orthotopic ureterocele with characteristic imaging appearance of a kissing cobra sign.

References

1. Castellanos-Hernández H, García-González VM, Figueroa-Zarza M, García-Sánchez D, Gutiérrez-Ochoa J. Giant ureterocele: A case report. Rev Mex Urol 2013;73:80-3.
2. Stunell H, Barrett S, Campbell N, Colhoun E, Torreggiani WC. Prolapsed bilateral ureteroceles leading to intermittent outflow obstruction. JBR–BTR 2010;93:312-3.
3. Sepulveda W, Campana C, Carstens E, Rodriguez J. Prenatal sonographic diagnosis of bilateral ureteroceles: The pseudoseptated fetal bladder. J Ultrasound Med 2003;22:841-4.
4. Shamsa A, Asadpour AA, Abolbashari M, Hariri MK. Bilateral simple orthotopic ureteroceles with bilateral stones in an adult: A case report and review of literature. Urol J 2010;7:209-11.
5. Abdulkadir AY, Adesiyun OA, Popoola AA, Adekeye AO. Ureterocele: Self resolved following spontaneous extrusion of calculus. Trop J Health Sci 2008;16:39-40.
6. Shokeir AA, Nijman RJ. Ureterocele: An ongoing challenge in infancy and childhood. BJU Int 2002;90:777-83.
7. Chavhan GB. The cobra head sign. Radiology 2002;225:781-2.
8. Tahboub A, Herskovits M, Loberant N. Bilateral orthotopic ureterocele. Imaging Sci Today 2010;321.
9. Günşar C, Mir E, Şencan A, Ertan P, Özcan CÜ. Pediatric ureteroceles: Diagnosis, management and treatment options. Iran J Pediatr 2010;20:413-9.
10. Thiel DD, Petrou SP, Broderick GA. Orthotopic ureterocele masquerading as a bladder tumour in a woman with pelvic pain. Int Braz J Urol 2005;31:549-51.
11. Gordon NS. The cobra raises its head: Transitional cell tumor progressing as acquired ureterocele. Br J Urol 1987;60:271-2.
12. Mitty HA, Schapira HE. Ureterocele and pseudoureterocele: Cobra versus cancer. J Urol 1977;117:557-61.
13. Hung-Wei C, Shiu-Dong C. Cobra head sign. Incont Pelvic Floor Dysfunct 2010;4:25.

How to cite this article: Adesiyun OM, Oyinloye OI, Akande HJ, Atobatele MO, Adeniyi WA, Abdur-Rahman LO. Bilateral giant orthotopic ureterocele appearing as kissing cobra in a Nigerian child. West Afr J Radiol 2015;22:42-4.

Source of Support: Nil, **Conflict of Interest:** None declared.