Congenital anterior urethral diverticulum presenting as a scrotal mass in a two-year-old child

Ali Gürağaç;[1] Hasan Cem Irkilata, MD;[2] Nahit Yunusov, MD;[2] Zafer Demirer, MD[3]

[1]Tatvan Military Hospital, Department of Urology, Bitlis, Turkey; [2]Gülhane Military Medical Academy, School of Medicine, Department of Urology, Ankara, Turkey; [3]Eskisehir Military Hospital, Department of Urology, Eskisehir, Turkey

Cite as: *Can Urol Assoc J* 2016;10(11-12):E395-7. http://dx.doi.org/10.5489/cuaj.3724
Published online November 10, 2016.

Abstract

Here, we describe a case of congenital anterior urethral diverticulum (CAUD) in a two-year-old boy, who presented with right inguinoscrotal swelling that mimicked a spermatic cord cyst or hydrocele. Accurate diagnosis was made intraoperatively by retrograde urethrography. Open diverticulectomy and primary urethroplasty were performed for its management. The clinical presentation, diagnosis, and the management of this rare condition is discussed.

Introduction

Anterior urethral diverticulum may be classified as congenital or acquired. Acquired diverticulum is more common than the congenital variety and urethral trauma, catheterization or instrumentation, infection, and obstruction can play a role in its pathophysiology.[1] Although congenital anterior urethral diverticulum (CAUD) is less common, it can occur as an isolated entity or in association with an anterior urethral valve, which can cause severe obstruction of the lower urinary tract and may also damage the upper urinary tract with serious repercussions.[1,2] However, most cases are diagnosed on prenatal ultrasonography (US) in early life if patients present with bilateral hydronephrosis and/or azotemia. The clinical presentation of CAUD is highly variable, depending on the child's age and the degree of obstruction.[3-6] Variable treatment choices, i.e., endoscopic or open interventions with variable outcomes, have been previously reported in the literature.[7] This current study reports a case of anterior urethral diverticulum large enough to involve the right inguinoscrotal region and which was erroneously diagnosed as a spermatic cord cyst or hydrocele preoperatively. An accurate diagnosis was made intraoperatively by retrograde urethrography and then managed with an open diverticulectomy and a primary urethroplasty.

Case report

An otherwise asymptomatic two-year-old boy was referred for evaluation of right scrotal swelling. His previous medical history was negative for urinary problems and an indwelling bladder catheter had not been used before. On physical examination, bladder was not palpable, bilateral testes were in the scrotum, and a palpable, soft, and fluctuant mass between right scrotum and radix penis, which was also extending toward right groin (diameter approximately 3 cm), was evident. The mass was not compressible completely by manual pressure. There was no swelling on the ventral aspect of the penis and scrotum during voiding. Urine analysis, routine blood counts, and blood urea and creatinine levels were within normal limits. US revealed normal kidneys and urinary bladder, and an inhomogeneous, hypoechogenic, hypovascular cystic mass at right upper scrotum extending into the right groin and radix penis was noted (Fig. 1A). However, in light of the findings, this entity was misdiagnosed as a spermatic cord cyst or hydrocele and a right spermatic cord cystectomy or hydrocelectomy was planned for its management. After a right hemiscrotal incision, the scrotal cystic mass was dissected carefully. It had limited mobility and involved the upper one-half of the right hemiscrotum, reached up to the midline radix penis, and extended toward right groin. The connection between the cystic mass and the penoscrotal junction of the urethra was noted during surgical exploration. On exerting pressure, the swelling disappeared and urine coming out per the external urethral meatus was seen, which was highly suggestive of CAUD. A retrograde urethrogram was performed intraoperatively to confirm the diagnosis and a large anterior urethral diverticulum arising from the penoscrotal junction of the urethra was revealed (Fig.1B). A midline incision was made over the diverticulum and dissection extended until the normal corpus spongiosum was visible. The incision also extended distally toward the penoscrotal junction of the anterior urethra to divide the attenuated tissue in this region. After excision of the diverticulum, a primary urethroplasty was performed over the transurethral catheter in

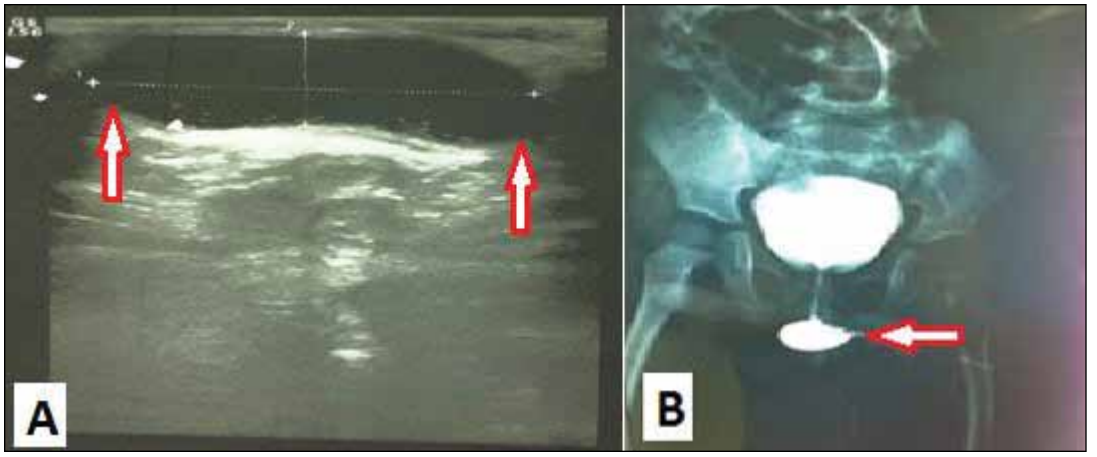

Fig. 1. (A) US demonstrated an inhomogeneous, hypoechogenic, hypovascular cystic mass at right upper scrotum extending right toward groin and radix penis; ***(B)*** intraoperative retrograde urethrogram confirmed the diagnosis; large anterior urethral diverticulum was shown arising from penoscrotal urethral junction of the urethra.

layers (Fig. 2). The patient made an uneventful recovery, and at the tenth postoperative day, the catheter was removed. Histopathological examination results were consistent with the diagnosis. On followup at three months, the patient had no voiding complaints and the micturating cystouretrography was negative for fistula formation, recurrence of the diverticulum, urethral stenosis, vesicoureteral reflux, or other postoperative complications.

Discussion

CAUD is a saccular outpouching arising from the ventral surface of the anterior urethra. This uncommon entity may cause urethral obstruction in childhood and can present at any age.[8] Although some authors distinguish between urethral diverticula and anterior urethral valves, others believe they belong to a spectrum of obstructive anterior urethral conditions.[9] While there is little doubt of the congenital origin of these lesions, no family pattern of inheritance was detected in this case.[9]

The etiology of this rare condition has provoked controversy for years. Various proposed hypotheses include a ruptured syringocele, incomplete hypospadias, cystic dilatation of the periurethral glands, developmental defects of corpus spongiosum, incomplete fusion of a segment of the urethral plate, sequestration of an epithelial nest after closure

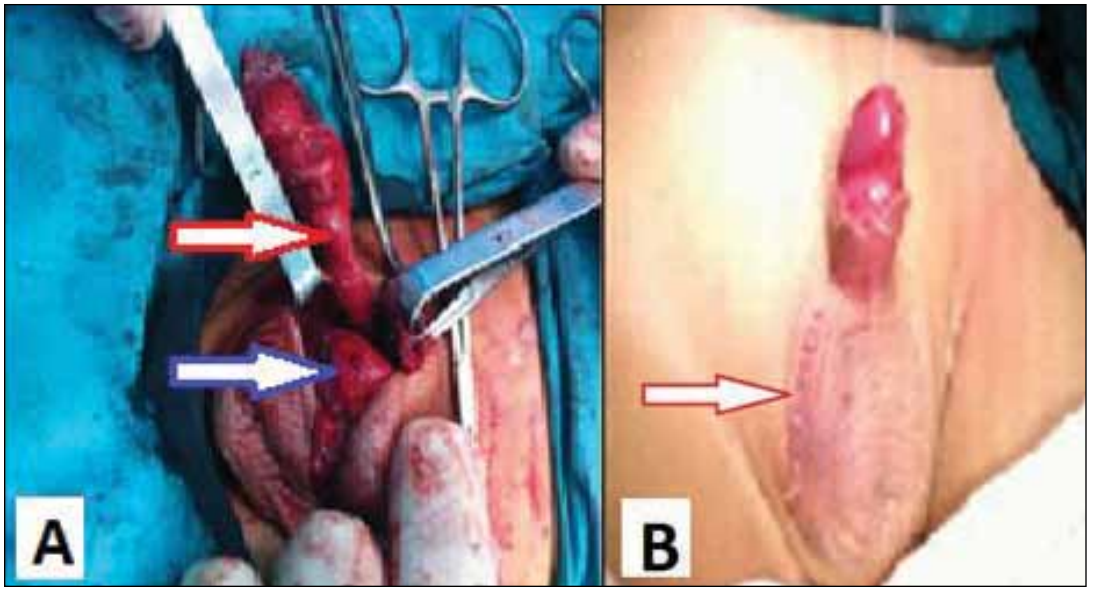

Fig. 2. (A) An excision of the diverticulum was performed (red arrow: spermatic cord; blue arrow: congenital anterior urethral diverticulum); ***(B)*** a primary urethroplasty was performed over the transurethral catheter in layers.

of urethral folds, and the presence of epidermal pockets communicating with the ventral urethral wall.[4-10]

The clinical presentation essentially depends on the child's age and the degree of obstruction. Generally, one-third of cases are diagnosed in the neonatal period and one-half by two years of age in patients who have never been catheterized urethrally.[9] Although infectious symptoms, i.e., fever, diarrhea, vomiting, dehydration, or even septicemia, predominate, especially in newborns and infants, older children usually presents with voiding complaints such as dysuria, weak urinary stream, frequency, incontinence, and sometimes bleeding and urinary retention.[9,11,12] Clinically, swelling on the ventral aspect of the penis or scrotum may appear with post-voiding dribbling. Upon compression of this swelling, urine coming out through the urethra may also be visualized.[9,11]

The diagnosis of CAUD is usually made by micturating cystouretrography or retrograde urethrogram. Micturating cystouretrography has some additional advantages in demonstrating vesicoureteral reflux or other associated anomalies. US can be performed for evaluation of upper tracts. A voiding US can be performed as an alternative diagnostic test to contrast studies.[13,14] Although cystourethroscopy is usually difficult, particularly with young children, this technique is therapeutic as well as diagnostic.[4]

Various treatment modalities have been described for this condition depending on the size of the diverticulum and the degree of the obstruction. A small anterior urethral diverticulum may be treated with an endoscopic resection of the anterior spur. A large diverticulum should be treated with excision of the diverticulum and reconstruction of the urethra performed in the same session. The latter approach may provide a more uniform caliber of urethra, but there is a risk of urethrocutaneous fistula formation.[7,9] If there are back-pressure changes of the upper renal tract with effected renal function, urinary diversion in the form of suprapubic cystostomy or vesicostomy and open treatment of the diverticulum are suggested as safer option.[15]

In conclusion, CAUD can present as a cystic mass between spermatic cord and radix penis, and it can mimic other scrotal pathologies such as spermatic cord cyst or hydrocele, as in this case. The precise diagnosis of uncommon presentation of CAUD in children requires a high degree of suspicion and heightened awareness. Despite the clinical presentation, large CAUDs in children should be treated in order to prevent complications secondary to urinary outflow obstruction. A diverticulectomy with primary urethroplasty should be the chosen treatment modality in such a case, as in this case where the condition was misdiagnosed preoperatively and the accurate diagnosis was made intraoperatively.

Competing interests: The authors report no competing personal or financial interests.

This paper has been peer-reviewed.

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Correspondence: Dr. Zafer Demirer, Eskisehir Military Hospital, Department of Urology, Eskisehir, Turkey; zaferdemirer1903@gmail.com