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A complete duplicated collecting system with giant ureterocele in adult: Case report

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ABSTRACT

INTRODUCTION: Ureteroceles is a developmental anomaly with cystic dilation of the distal aspect of the ureter and are often associated with some urological anomaly such as a duplicated system or stenotic ureteric orifice.
PRESENTATION OF CASE: This study reports an ectopic ureterocele in duplication of collecting system associated with double ureters and ureteral ectopia in a woman aged 24 years with minor flank pain. Cystoscopy deroofing of the ureterocele performed and followed by secondary surgery laparoscopic heminephrectomy.
DISCUSSION: Ureteroceles have various clinical manifestations and complications. Treatment for ureterocele depends on age, type of the ureterocele, obstruction to the draining system, and complications. No single method is sufficient for all cases, and management must be individualized. Endoscopic treatment has gradually broadened as a safe, minimally invasive, and effective procedure, but there is no consensus on its effectiveness for treating ectopic ureterocele. However, it is reported that 50–80% of cases after initial endoscopic treatment require secondary surgery.
CONCLUSIONS: Ureterocele is reported rarely in adults, especially with duplication of the collection system in the nonorthotopic (extravesical) position in women. Cystoscopy deroofing of the ureterocele can be performed to decompress the hydroureteronephrosis, and laparoscopic heminephrectomy can be performed due to dysfunctional uppers moiety. Long-term follow-up is required to monitor renal function, symptoms, and occurrence of vesicoureteric reflux.

1. Introduction

Ureteroceles are cystic dilation of the distal aspect of the ureter. It is a developmental anomaly and can be located intravesical (inside the bladder) or in extravesical (the bladder neck and urethra). Some urological anomalies are often associated with Ureteroceles. The anomaly such as a duplicated system or stenotic ureteric orifice. Ureteroceles incidence is 1: 4000 people, occurring four times more often in women with some predominance on the left side, and 10% of cases are bilateral [1].

Ureterocele presentations can vary greatly, ranging from urinary incontinence, urinary tract infections, tenesmus, incomplete bladder emptying, supra-pubic pain, bladder tension, and ureterocele prolapse. Ureteroceles, especially in adults, are generally asymptomatic and rarely diagnosed [2]. The low incidence of ureteroceles in adults and unclear physical findings cause the diagnosis to be often delayed. There is almost no literature on adequate diagnostic algorithms and appropriate available treatment nowadays [3].

In this study, we reported a case of an ectopic ureterocele in duplication of collecting system associated with double ureters and ureteral ectopia in a middle-aged woman. Cystoscopy deroofing and later laparoscopic heminephrectomy were performed due to the dysfunctional upper moiety. This study is reported in line with the SCARE checklist [4].

2. Case presentation

After a medical check-up, a twenty-five-year-old female patient was referred from the Obstetrics and Gynecology Department with suspicious right renal cyst and uterine cyst through ultrasonography findings. There was a history of minor recurrent right flank pain for the last five months and no urinary flow symptoms. Physical examinations were within normal limits, with a soft, non-painful, and depressible abdomen, non-painful and non-distended right flank region, and no mass nor other abnormality in external genitalia. The laboratory investigations of complete blood count,

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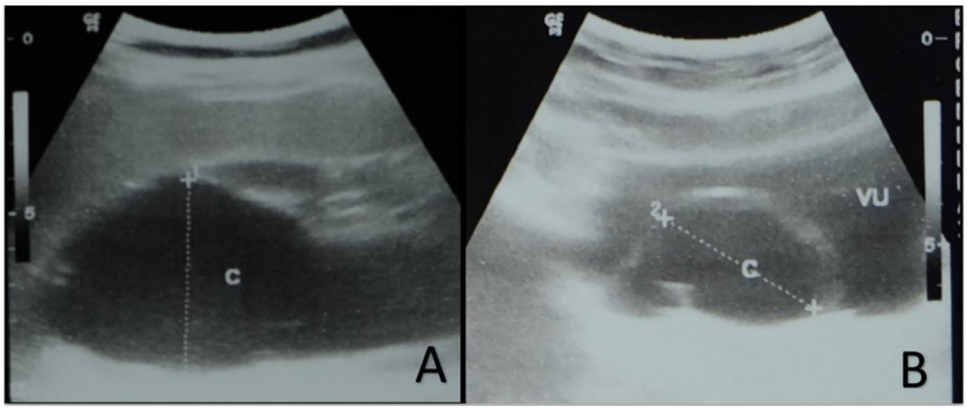

Fig. 1. Ultrasonography showing: A. right proximal hydroureter, and B. Giant ureterocele.

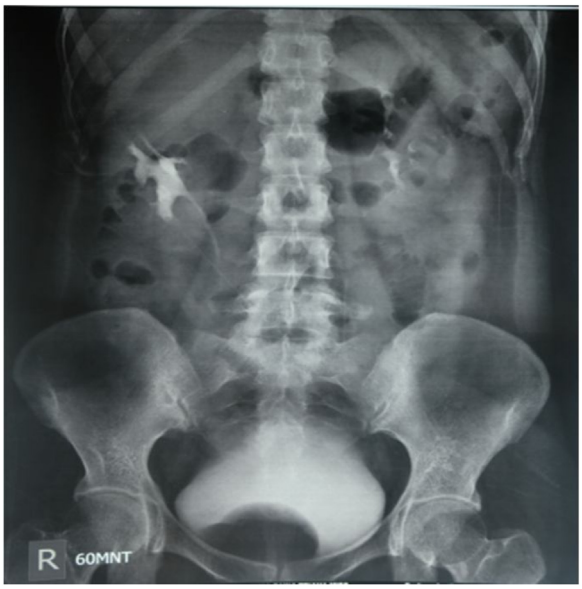

Fig. 2. IVP (60 min) showed a 'dropping lily' sign and a large filling defect in the bladder neck suggestive of giant ureterocele.

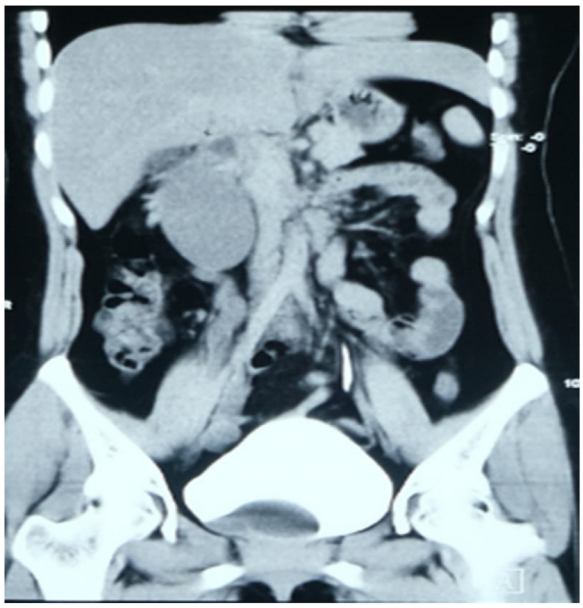

Fig. 3. Contrast CT urography revealed right upper pole hydronephrosis grade III-IV, right proximal-distal hydroureter, and giant ureterocele in the bladder neck.

creatinine, coagulation profile, urinalysis, including renal function tests, were within normal limits. The patient has no history of allergies and has no history of taking any routine medications. There was no hematuria nor urinary tract infection history and no family history of the same complaint. The patient did have a medical report at the age of 2 years old when she had a mass that burst out from her urethra. She was examined in a hospital, performed urethral catheterization, and removed the catheter a week later. No further investigation was done, and since that period, she had no symptoms until now.

Abdominal ultrasonography showed right upper pole hydronephrosis grade III-IV and right proximal-distal hydroureter (Fig. 1). Intravenous pyelography showed a 'drooping lily' sign in the right urinary system (Fig. 2). Computerized tomography (CT) urography with contrast confirmed right upper pole hydronephrosis grade III-IV, right proximal-distal hydroureter, and a giant ureterocele in the neck of the bladder. The left pelvicalyceal system and ureter were visualized normally (Fig. 3).

The patient underwent a cystoscopy performed by the main author (Postgraduate Specialist, Urology Master of Surgery) in a general hospital. The Cystoscopic examination found the ureterocele had filled the bladder neck area and identified upper pole ureteral insertion in posterior urethra, normal lower pole ureteral insertion, and no decompensated bladder [Fig. 4]. Unfortunately, no Isotope renal scintigraphy scan was performed due to under maintenance. In this case, the differential diagnosis was a duplication of the right collecting system associated with double ureters, hydroureteronephrosis in the upper pole, and ureteral ectopia with giant extravesical ureterocele.

The patient underwent cystoscopy deroofing of the ureterocele. The operating surgeon was the main author (Postgraduate Specialist, Urology Master of Surgery) in a general hospital. The patient's recovery progressed well after doing the procedure. Three months later, she complained about an increasing dull right flank pain but had no incontinence nor sign of urinary tract infection. The laboratory investigations were within normal limits. CT urography with contrast was performed and showed a non-functional upper pole of the right kidney. Next, the patient underwent laparoscopic heminephrectomy for the non-functional upper moiety of the right kidney [Fig. 5]. The postoperative period remained uneventful. She followed up in outpatient surgery clinics. At follow-up, one month and six months after the surgery, the patient had recovered well and had no urological complaints.

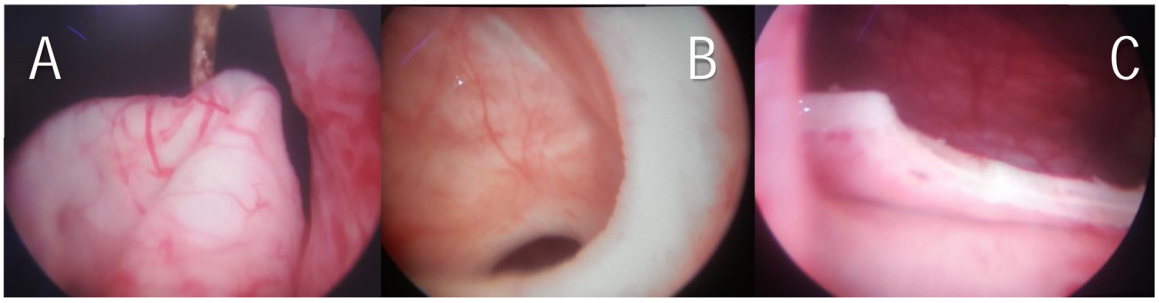

Fig. 4. Ureterocele endoscopic deroofing: A. Ureterocele filled internal urethral orifice, B. The right ureteral orifice in posterior urethra, and C. After deroofing.

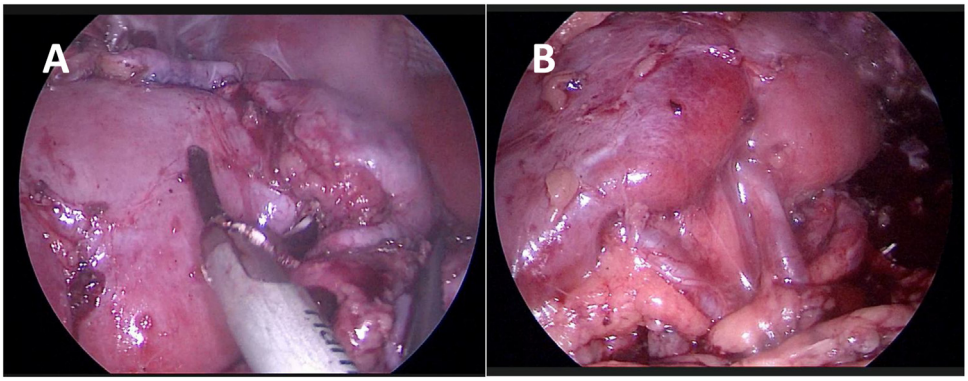

Fig. 5. Laparoscopy Heminephrectomy: During (A) and after (B) upper pole right kidney removal.

3. Discussion

A ureterocele involves a cystic dilation of the distal aspect of the ureter and often presents with other anomalies such as stenotic ureteric orifice or duplex upper tract and other clinical symptoms. The incidence is four times more common in women with some predominance on the left side [1]. The etiology of the occurrence of ureteroceles is still unclear. It is generally accepted that ureteroceles are a congenital disease in pediatric populations, and in adults, some authors believe it is an acquired disease [4].

Ureteroceles are classified into intravesical (entirely in the bladder) or ectopic (located in the bladder neck or the urethra), and ectopic is more common than intravesical ureterocele [5]. Pediatric ureteroceles often present with duplication of the collection system in the nonorthotopic (extravesical) position, while adult ureteroceles usually involve a single unilateral system in the orthotopic (intravesical) position [6]. In this case, we found a giant extravesical ureterocele on the right side with other anomalies, that is, duplex renal associated double ureters in an adult woman.

Ureterocele in a duplex system commonly involves the upper renal moiety and results in stenosis and ureter obstruction, which can cause severe ipsilateral hydroureteronephrosis. Meanwhile, hydronephrosis in the lower moiety is usually due to vesicoureteral reflux. This condition will later be related to morbidities, such as recurrent urinary tract infections (UTIs) or chronic pyelonephritis [7].

Ureteroceles have various clinical manifestations and complications. Commonly in pediatric can be found recurrent UTI or urosepsis, incontinence, failure to thrive, urinary calculus, abdominal mass, urinary tract obstruction, and vaginal or urethral prolapse. In the adult population, ureterocele usually stays asymptomatic, and diagnosis is usually made accidentally, sometimes accompanied by intermittent pelvic pain, recurrent UTIs, or calculi [1]. While ureteroceles can cause obstruction, they are less commonly reported in adults, especially with the duplex system [8]. In our case, the 25-years older adult had minor flank pain but no urinary symptoms. Interestingly, there was a duplication in the right kidney, hydroureteronephrosis at the upper pole, and ureteral ectopia with a giant ectopic ureterocele draining the upper pole.

Initial ureterocele diagnostic imaging is generally done with ultrasound to identify cystic dilatation in the bladder wall and can sometimes show a picture of duplication of the system [1]. Intravenous urography (IVU) can be done to assess renal function if there is a decrease in function, which will appear as delayed excretion or even no excretion [4]. If complete ureter duplication is found, renal scintigraphy may be performed to evaluate scar tissue and determine the upper and lower poles' function. This can help in the choice of therapy [9]. In ectopic ureteroceles, 74%–90% are related to the duplex kidney's upper pole, which shows minimal or no function, and the resulting ureteroceles are primarily negative on the radiographic sign [9]. Hydronephrosis deviates the upper pole to the downward side and pushes the functional lower pole laterally and inferiorly, giving a 'drooping lily' sign [5].

The ureterocele management should be individualized based on clinical presentation, type of ureterocele, patient's age, and other clinical variables that may contribute to management's best choice [5]. Endoscopic treatment has the advantages of simplicity and minimally invasive, but there is no consensus on its effectiveness for treating ectopic ureterocele. It has been reported that 50–80% of cases after initial endoscopic treatment need secondary surgery in ectopic ureterocele [9].

A meta-analysis (1965–2005) conducted by Byun and Merguerian found a greater relative risk for reoperation after ureterocele incision in patients with extravesical as compared with intravesical ureterocele. The need for secondary surgery was greater in ureteroceles associated with duplex versus single collecting systems [10]. However, it is generally agreed that endoscopic puncture of an ectopic ureterocele is primarily used to treat uncontrolled sepsis and azotemia with bladder outlet obstruction with or without any ureterocele prolapse [5].

In cases where the upper renal moiety function is poor, the upper pole heminephrectomy is a standard surgical treatment [11]. In our case, the patient underwent endoscopic treatment to decompress the hydroureteronephrosis and preserve renal function. Secondary surgery was performed due to CT urography with contrast indicating a dysfunctional upper moiety. Besides, Renal scintigraphy is useful in planning corrective surgery and should be performed. Renal scintigraphy is a gold standard to assess function in the duplex kidney and to detect and follow up on a malfunctioning upper pole and scar tissue in cases of ureterocele.

After three months of follow-up, the patient recovered well with no symptoms and no further complications. However, the patients required long-term follow-up to monitor renal function, symptoms, and occurrence of vesicoureteric reflux.

4. Conclusion

Ureterocele with duplication of the collection system in the nonorthotopic (extravesical) position in women reported rarely. Cystoscopy treatment can be performed for initial treatment to decompress the hydroureteronephrosis, and later laparoscopic heminephrectomy can be performed due to dysfunctional uppers moiety. Long-term follow-up required to monitor renal function, symptoms, and occurrence of vesicoureteric reflux.

Declaration of Competing Interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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Ethical approval

This type of study does not require any ethical approval by our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution

Prahara Yuri: study concept, drafting and final approval the article.

Eldo Taufila Putra Utama: study concept and revising the article.

Registration of research studies

Not applicable.

Guarantor

Prahara Yuri.

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