Early experience with transurethral endoscopic de-roofing of single system orthotopic ureteroceles in adults

Abstract :

BACKGROUND:

Ureteroceles are cystic dilatations of the distal ureter that occur due to congenital ureteric wall weakness. Only 20% are termed single-system orthotopic ureteroceles and occur in normal ureteric locations typically known as “adult ureteroceles”. It is a rare condition with incidence ranging from 1:500 to 1:4000 affecting more females than males in a ratio of 6:1. There is no consensus in literature regarding the best endoscopic technique in treating ureteroceles in adults. Here we describe our experience with endoscopic de-roofing of adult orthotopic ureteroceles using the transurethral cutting loop.

METHODOLOGY

This is a 6 year retrospective analysis of 11 patients aged18years and above with 13 orthotopic adult ureteroceles who underwent endoscopic de-roofing. Their data were retrieved from case notes for analysis.

RESULTS

This series consisted of nine women and 2 men (F: M 4.5:1) with a mean age of 36.5 +/-14.8 years (range 21 - 57). The dominant presenting complaint was flank pain which was observed in all but two patients (n = 9, 81%). Others symptoms included hematuria (n = 4,

36%) and those of urinary tract infection (n=8, 73%). A total of 11 patients and 13 orthotopic ureteroceles were operated upon as elective cases 10 (91%), while 1 (9%) case, with bilateral obstruction from stones, was operated upon as an emergency. The patients were all discharged home after an over-night stay in hospital following the procedure except the case with bilateral renal obstruction.

CONCLUSION

The endoscopic approach to treating adult single-system orthotopic ureteroceles using transurethral resection cutting loop at low current is an effective, safe and easily adaptable procedure.

KEYWORDS

Ureter, urine, stone, anomaly, incision

INTRODUCTION

Ureterocele is the pseudo-cystic dilatation of the distal end of the ureter due to congenital ureteric wall weakness. (1)(2)(3)(4)(5)(6) It is often diagnosed during antenatal ultrasonography in children as heterotopic ureteroceles,(3)(4)(5) located in ectopic ureters or ectopic duplex renal systems. (3) In 20% of cases it occurs in the normal ureteric location (orthotopic ureteroceles) and this is typically seen in adults hence the name “adult ureteroceles”.(1)(2)(7) It is a rare condition with incidence ranging from 1:500 in children to 1:4000 in adults and it has a predilection for females in a ratio of 6:1.(2)(5)(6)(7)(8)

The etiology of ureterocele to date is not very clear. (2)(4) While ureteroceles in children are commonly regarded as congenital, in adults it may be considered acquired as a result of inflammation in or around the vesical wall or from the passage of stone. (1)(2) One of the most popular theories behind its congenital origin posits that it develops from the incomplete resorption of the chwelle’s membrane. (2) Ureteroceles can be referred to as either a single system or duplex system ureteroceles. Ericsson in 1954 classified ureteroceles into simple and ectopic, while Stephens in 1958 classified them into, stenotic, sphincteric, sphincterostenotic and cecouretrocele.(2)(3) Brueziere on the other hand classified ureteroceles as follows; type A (intravesical ureterocele on single ureter), type B (ectopic ureterocele on single ureter), type C (intravesical ureterocele on pyeloureteral duplicity) and type D (ectopic ureterocele on pyeloureteral duplicity).(3)(9)

It commonly occurs unilaterally but in 10 - 15% of cases, it is bilateral. (2)(7) In adults it is not unusual to find ureteroceles incidentally since a good number are asymptomatic. (2)(5) In a few circumstances the patients may present with symptoms such as recurrent flank pain, hematuria and features of urinary tract infection. (2)(4)(7)(5) Radiological imaging is the hallmark of diagnostic evaluation of ureteroceles. (2)(3)(5)(10)

Currently ,there are no clear guidelines in the management of this condition and various procedures have been described for its treatment.(1)(4)(5)(6)(7) Over the last 3 decades the surgical approach to the treatment of ureteroceles has evolved from open surgery to minimally invasive endoscopic procedures.(3)(4)(11)(12) Several endoscopic techniques have been described in the literature for the treatment of ureteroceles albeit mostly in children. None of these methods can be said to be the most ideal technique since there are no documented randomized controlled studies in the literature.(1)(4)(5)(6) The general objectives of treatment are to prevent infection and preserve renal function.(1)(2)(3)(6)(7) These endoscopic techniques involve puncture or incision in diverse ways using any of the following: cold knife, Collins knife, bugbee electrode and laser.(1)(7)(13)(14) Puncture of ureteroceles risks potential bladder perforation and grave intra-peritoneal complications.(1)(15) Herein, we describe our experience with the management of adult orthotopic ureteroceles using transurethral endoscopic de-roofing with the cutting loop.

MATERIAL AND METHODS

This is a 6-year retrospective study of 11 patients with 13 orthotopic single-system adult ureteroceles who consented to and underwent transurethral endoscopic de-roofing in our practice in Abuja between January 2013 and June 2019. Data from their case notes were retrieved and analyzed with Excel version 2013. Unilateral ureterocele was present in 9 cases (82%) while two (18%) had bilateral ureteroceles. There was a preponderance of left-sided pathology (78%) amongst the unilateral ureteroceles. All the patients had prophylactic antibiotics followed by subarachnoid block anesthesia before being placed in lithotomy position for the procedure. Routine cystoscopy was carried out to further confirm the diagnosis and exclude any other lesions. The endoscopic de-roofing procedure was performed using either a Karl Storz or Vega German resectoscope with either monopolar or bipolar cutting settings at the lowest possible energy. Figure 1. The cutting loop when used properly has the safety of not making cuts too deep to risk injury to the bladder.Figure 2. A stone was harbored in the left ureterocele in one of the patients who presented to our

emergency ward with severe pain and anuria. Figures 3 and 4. She was found to have bilateral stones obstructing the right renal pelvis and in the left ureterocele. She had emergency endoscopic de-roofing and the stone dropped into the bladder following which litholapaxy was performed to crush it. Figure 5. Double J stents were passed bilaterally in this patient with bilateral obstruction following the de-roofing procedure. The right pelvic stone was later on operated. We routinely left in place a size 6Fr 26cm double J stents in all ureterocele units after de-roofing for 6 weeks.Figure 6 The patients were followed up clinically and radiologically as well as with urinalysis for complications such as vesicoureteric junction reflux and urinary tract infection. The procedure is as depicted in the figures 1 - 5. All patient's data were entered into an Excel proforma and analyzed using descriptive statistics.

RESULTS

This series consisted of eleven patients, nine females and 2 males (F: M 4.5:1) with a mean age of 36.5 +/-14.8 years (range 21 - 57). Figure 7. The dominant presenting complaint was flank pain, observed in all but two patients (n = 9, 81%), followed by hematuria (n = 4, 36%) and features of urinary tract infection (n=8, 73%) Figure 8. The diagnosis was made based on clinical presentation and imaging studies (abdominal ultrasonography and intravenous urography). Figure 9 and 10. The classic appearance of the terminal ureters as cobra head was reported on intravenous urography in all our patients. Figure 8. Abdominal sonography, however, missed the ureterceles in 2 of the patients (18%) whom intravenous urography had correctly diagnosed. In the majority of cases n = 9 (81%), the pathology was unilateral while it was bilateral in only n = 2 (18%). All 11 patients presenting with 13 orthotopic ureterocele units were type A according to the Brueziere classification. Ten of the surgeries (91%) were elective cases while one was treated as an emergency (9%) with bilateral obstruction from stones in the left ureterocele and contralateral impacted right renal pelvic stone. The laterality of the pathology was as follows; left side (n=7, 64%), right side (n=2, 18%) and bilateral ureteroceles (n=2, 18%). Transurethral endoscopic de-roofing was performed on all

patients who had given consent for the procedure. The mean operating time was 39 minutes taking into account the treatment of the solitary stone in the ureterocele in the patient with acute kidney injury. No surgical incident was noted during the de-roofing procedures and the average hospital stay was a day except in the patient with bilateral renal obstruction who spent 4 days in the hospital. Transient hematuria which was self-limiting was recorded in 4 of our patients following the surgical procedure. No significant complications were recorded during the period of follow-up (range 3 to 9 months).

DISCUSSION

Ureterocele is a rare congenital anomaly that refers to the cystic dilatation of the distal ureter commonly found in children.(1)(2)(3)(4)(5) Twenty percent of ureteroceles are termed single-system orthotopic ureteroceles and located in the normal ureteric sites typically in adults hence the name “adult ureteroceles”.(1)(2)(7) The rare nature of this condition is obvious even in our study where only 11 patients were seen over six years. Its etiology remains unclear but several theories have been postulated. (2)(3)(4) Among the postulations is that of abnormal muscular development of the terminal part of the ureter which gives rise to weakness and dilatation of the distal ureter.(2) Another school of thought believes that there is an abnormal stimulus that eventually leads to the dilatation of the distal ureter.(2)(4) The most popular and plausible theory, however, is that of incomplete resorption of the chwalle's membrane, a double-layered portion of the ureteric lumen which normally gets ruptured during the eighth week of gestation. (2)(6) Ureteroceles found in adulthood most probably have an acquired origin, likely due to inflammation in or around the vesical wall or from the passage of ureteric stones.(2)(6)(7) There is yet, no consensus in the literature on the best treatment modality for ureteroceles.(1)(4)(5)(6) This may probably be a result of its rare nature and also the lack of prospective, randomized controlled studies of the various treatment options. (4) The goals of treatment are as follows; the prevention of infection and preservation of renal function.(1)(2)(3)(6)

Over the last few decades, the surgical approach to ureteroceles has gradually evolved from open surgery to minimally invasive endoscopic procedures.(1)(3)(4) The endoscopic approach by transurethral de-roofing of single-system orthotopic ureterocele is easy to learn and teach as well as safe and can be easily adapted to, especially in the era of upsurge in endourology practice and patients quest for minimal access surgeries.(5)(16)(17) The transurethral de-roofing technique preserves the posterior detrusor muscle wall and thereby does not temper with valve mechanism.(18)

The mean age of patients in our study is 36.5 +/-14.8 years (range 21 - 57) and this is similar to that reported by Ahmed et al and other researchers.(2)(6)(16)(17) Ralph et al, however, noted a bimodal age distribution with the first peak being 30 to 39 years age group, consistent with our series, while they noted a second peak between 60 – 69 years. (4) We found a female-to-male sex ratio of 4.5: 1 which is also similar to local and foreign studies. (2)(3)(5)(6)(7) One study, however, in southern Nigeria reported a male preponderance of in the ratio of 3:2.(4)

Surgical intervention becomes indicated in patients with symptomatic ureteroceles such as urinary tract infections, impaired renal function, ureterovesical drainage, and severe vesicouretericreflux.(2) In our series, all the ureteroceles were classified as intravesical on a single ureter (type A according to the Brueziere classification). Amine Oueslati et al in their series documented that the type A ureteroceles constituted the commonest type.(3) The type A ureterocele which is a simple form of the disease may explain the good tolerance as well as the late presentation in adulthood in a disease that is presumed to be congenital. Renal dysplasia and obstruction in the urinary tract which is common with ureteroceles in children are not typically the presentation in adult ureteroceles. (2) Although orthotopic ureteroceles hardly become symptomatic in adults and adolescents, the presence of symptoms such as voiding problems and flank pain may be pointers to its eventual diagnosis especially where the index of suspicion is high. (1)(2)(5)(7) In this study, flank pain and hematuria occurred in 80% of the patients, while lower urinary tract symptoms and urinary tract infection were the

other patterns of presentation reported. Several articles have observed a similar picture in adult ureteroceles.(2)(3)(5)(6) An associated pathology we reported in our series was a stone in the ureterocele in a female patient. This is not an unusual presentation as several studies have shown this to be common in adult ureteroceles. (2)(3)(5)(6)(13)(14) The development of stones in the distal obstructed cobra head which typically has a stenotic ureteric orifice is likely a result of the stasis of urine in the ureterocele.(2)(5) Unlike some authors we did not find in our series, associated pathologies such as genitourinary tuberculosis or schistosomiasis hematobium that may have an etiologic role in the occurrence of adult ureteroceles (2)(19)

Abdominopelvic ultrasound scans confirmed the diagnosis in all but 2 cases in our study. We were not surprised at this, because the ultrasound scan is done in a well-hydrated patient with special focus for this anomaly, it is not unlikely to miss the lesion.(19) Moreover ultrasonography is known to be operator-dependent and the experience of the operator would certainly influence the outcome. The ureterocele is seen as a cystic mass in the bladder or close to the urethra on an ultrasound scan. (2)(19) Intravenous urography on the other hand is a more sensitive diagnostic test and it revealed a cystic dilatation of the distal ureter, radiologically described as cobra head or spring onion deformity with a peripheral halo.(2)(3)(4)(7) Intravenous urography further demonstrates the function and anatomy of each renal unit which becomes useful in planning treatment for the patients.(2) Intravenous urography could not be done in patients with acute kidney injury from bilateral renal obstruction, so we relied on ultrasound and cystoscopy in evaluating her. We did not carry out computerized tomography scans and renal isotope scans due to cost limitations and difficulty in accessing these services by patients in our environment.

Early surgical intervention is required for the cure and prevention of long-term complications in patients with ureteroceles.(2)(7) Minimally invasive endoscopic procedures in the treatment of ureteroceles have in recent years become more commonly practiced than open surgery, not only in adults but also in children.(1)(2) Many different endoscopic techniques

have been described in literature albeit mostly in children.(3)(7) There is no consensus yet in the literature referring to any of these methods as the most ideal for treating this disease.(1)(4)(5)(6) The goals of treatment remain the prevention of infection and preservation of renal function.(1)(2)(3)(6)(7) Endoscopic ureterocele puncture or incision can be performed in many ways using a cold knife, Collins knife, bugbee electrode or laser.(1)(2)(7)(13)(14) Puncture of ureteroceles potentially risks bladder injury and this could give rise to severe intraperitoneal complications.(1) In this study we carried out endoscopic de-roofing of the adult single-system orthotopic ureteroceles using the transurethral resectoscope cutting loop for all our patients. The controversy on whether to stent or not following endoscopic de-roofing remains and the decision is at the discretion of the surgeon.(4)(5)(7) In our series we stented all patients for 6 weeks following the procedure and our choice to do so was to allow healing to take place around the stent which resulted in a near-normal appearance of the orifice. The patients did well and no significant perioperative adverse events were recorded. They were all discharged home within 24 hours except the patient with bilateral renal obstruction. This short hospital stay and early recovery from surgery are the advantages of the endoscopic de-roofing technique over the traditional open methods and this makes the former more attractive. Raphael et al and Chowdry et al used a similar technique of transurethral endoscopic de-roofing of adult ureteroceles and concluded that the technique accords patients a minimally invasive option with minimal morbidity.(4)(5) The follow-up period in our study ranged from 3 to 9 months and all the patients reported satisfactory outcomes. The post-operative ultrasound scan did not show any recurrence or upper tract dilation. We were unable to do a micturating cystourethrogram to exclude post-treatment reflux in our patients and this is one of the drawbacks of retrospective studies. It is pertinent to follow up with these patients for longer durations to monitor their long-term renal function, recurrence of symptoms and post-treatment vesicoureteric reflux

CONCLUSION

Adult ureteroceles even though an uncommon pathology should arouse a high index of suspicion among urologists and other clinicians to clinch the diagnosis. In the absence of consensus on the ideal treatment, we share our experience with the management of this pathology and treatment using transurethral endoscopic de-roofing to help add to knowledge on this subject. The endoscopic approach to treating adult single-system orthotopic ureteroceles using a transurethral resection cutting loop at low current is an effective, safe and easily adaptable procedure.

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Fig 1: The proper cutting loop (not making cuts too deep to risk injury to the bladder)

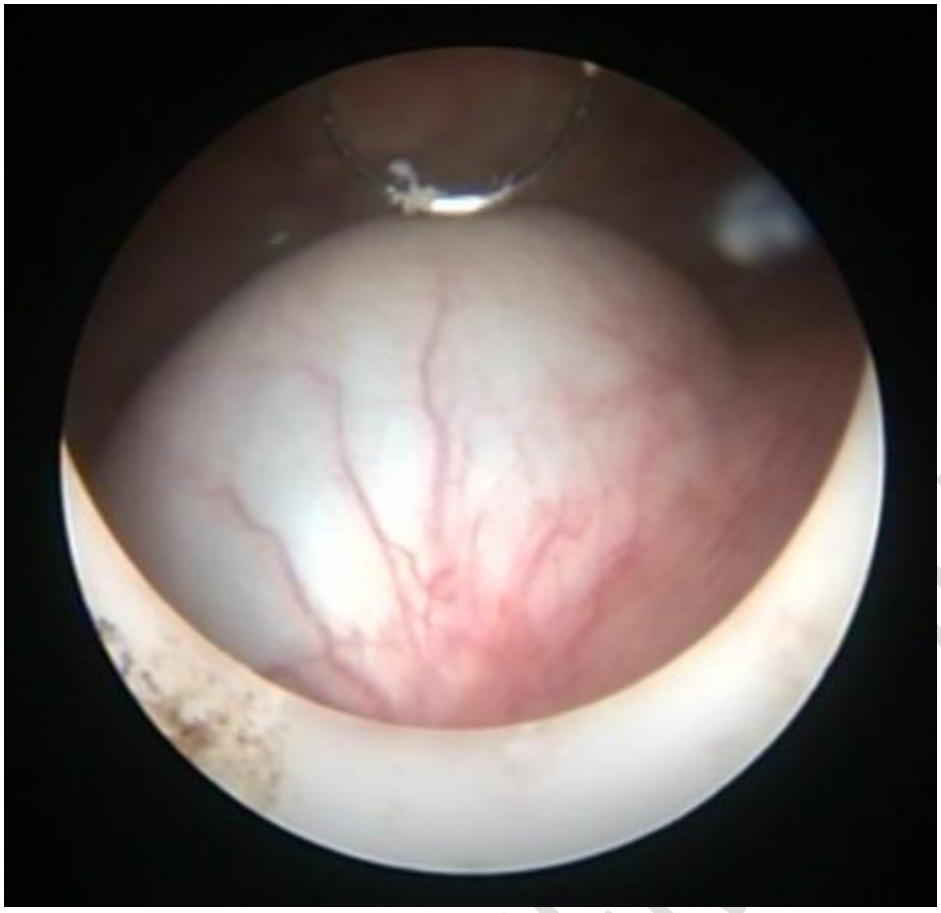

Fig 2 :Stone in the left ureterocele

Fig 3 :Bilateral stones obstructing the right renal pelvis

Fig 4 :Bilateral stones obstructing in the left ureterocele

Fig 5 :Bilateral Double J stents

Fig 6 :Vesicoureteric junction reflux and urinary tract infection

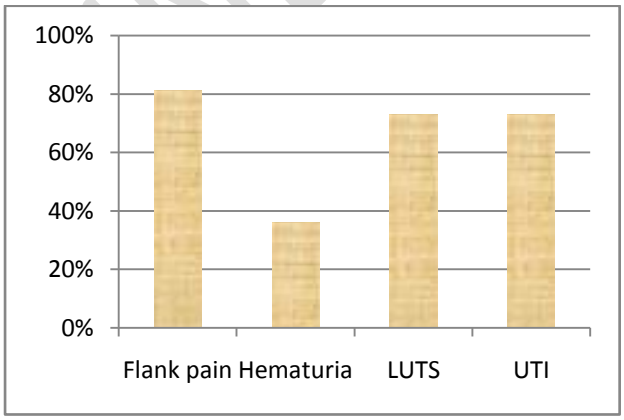

Fig 7 :Complaints of flank pain, LUTS and UTI

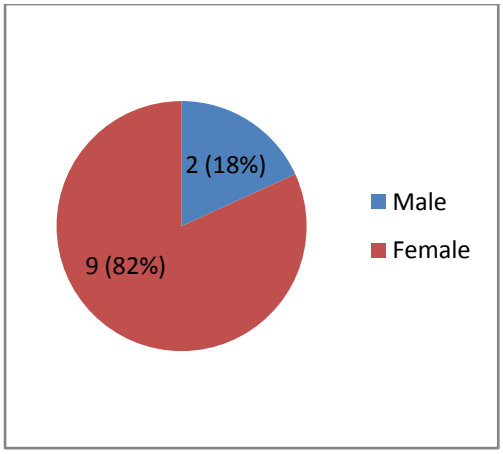

Fig 8: Received Abdominal ultrasonography and intravenous urography

Fig 9: Terminal ureters

Fig 10 :Terminal ureters as cobra head