

Management of Ureterocele: Review Article

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Abstract

A ureterocele is a cystic dilatation of the distal ureter with variance in anatomy and presenting symptoms. Ureterocele can be associated with single or duplex collecting systems. The clinical presentation can be so diverse as to be ranging from totally asymptomatic to overt urinary tract infection. It is multimodal management when it comes to managing a case of ureterocele for optimal outcomes. Literature is still struggling to find best practices guidelines in managing ureterocele. Basic measures are taken to preserve renal function. Primarily these cases are managed by adopting transurethral incision of the ureterocele which is an easy, minimally invasive and practicable option for dealing patients who have ureterocele. In this review we tried to explain the importance of diagnosing this rare ailment in time and taking the measures to deal with it in terms of preservation of renal function. New advances and surgical options have also been discussed here in managing such cases.

Keywords: Cystoscopy • Ureterocele • Complications • Surgery

Introduction

A ureterocele is basically a cystic dilatation of the distal ureter, a challenging clinical situation because of variance in anatomy and presenting symptoms [1,2]. Ureterocele can be associated with single or duplex collecting systems. Additionally, it may be either intravesical (orthotopic) or open at extravesical site (ectopic ureterocele) [2]. Naturally. Vesicoureteric Reflux (VUR) may be the result and can be present in the upper moiety or both of the moieties.

The clinical presentation can be so diverse as to be ranging from totally asymptomatic to overt urinary tract infection [2]. Ureterocele display varying signs and symptoms such as urinary tract infection, obstructive lower urinary tract symptoms, incontinence, and on very rare occasions a ureterocele might hang out of the urethra. Protruding ureterocele are found very rarely [3-5].

It is multimodal management when it comes to managing a case of ureterocele for optimal outcomes. Literature is still struggling to find

best practices guidelines in managing ureterocele. Broadly speaking, management aims frequently sought for include persevering the renal function up to the maximum possible extent, preventing vesicoureteric Reflux (VUR) and treating VUR in a timely manner in case it arises or if it is already present, keeping the kidney drainage intact (treating ureteric narrowing congenital or acquired), in order to preserve renal parenchymal tissue, preventing bladder outlet obstruction, maintaining subject's continence mechanisms. Besides these, minimizing chances of any potential infection is the hallmark of overall management [1,2]. Primarily these cases are managed by adopting transurethral incision of the ureterocele which is an easy, minimally invasive and practicable option for dealing patients who have ureterocele [2].

Materials and Method

We did search on PubMed, Medline database publications using: ureterocele, etiology, Infant, diagnosis, complications, surgery treatment options. The publications included were case reports, reviews, books and research studies regarding the subject matter over last 50 years.

Discussions and Literature Review

As mentioned earlier ureterocele is a cystic dilatation of the terminal section of the ureter (intramural part) that leads to obstruction of urine flow, consequently there is gradual dilation of the ureter and renal pelvis resulting in decline in renal function [3].

Ericson, in 1954 made the first attempt at classifying this disease [4]. He mentioned that it can occur as simple or ectopic (determined by the site of opening of the ureteric orifice). The opening of the ureter in a simple non-duplex system is located in the trigone area of the urinary bladder but in case of an ectopic ureterocele this orifice can be located anywhere between the bladder neck and the posterior urethra [1].

Different Classifications Used for Severity and Nature of Ureterocele

Stephens had classified ureterocele into four types (categorized on the basis of size and location of the ureteric opening): Stenotic (orifice is narrow and lies within the bladder), sphincteric (broad open within the internal sphincter), sphinctero-stenotic (when the orifice is narrow and resides within the internal sphincter) and caeco-ureterocele (blind-ending ureterocele that extends down the urethra) [5].

Churchill et al. utilized the concept of extent of the renal tissue at risk of damage secondary to the stasis (obstruction) or high-grade reflux [6]. Reflux Grade 1 when only the ureterocele segment is affected, Grade 2 when both segments of one kidney are affected and Grade 3 when both kidneys are affected.

The classification used by the American Academy of Pediatrics is based on the location of ureterocele-intravesical (when located inside urinary bladder) or ectopic (some portion of the ureterocele lies at the bladder neck or the urethra) is the most widely used currently [7].

Discussions and Literature Review

Incidence

Ureterocele are known to happen roughly in 1 in 4000 children, being most common in Caucasians [1,3]. They are more frequently seen in females (almost 4 times as compared to males). This ratio holds true from pediatric age to adult age ranges [1-4].

Left side is considered to be affected more frequently as compared to the right kidney. In up to ten percent of patients affected it may be seen bilaterally [4]. Incidence of an ectopic type of ureterocele has been seen almost four times as compared to the intravesical type of ureterocele [5-8].

It is estimated that in initial infancy age 3/4th of these ureterocele are found in association with the upper pole moiety in a duplex system [6-9]. Importantly, the upper pole in duplex system is prone to be dysplastic or having a poor functioning. It is very rare to find ectopic ureterocele in a single-system and are detected by and large in male gender [7-9].

Genetics involvement

Studies have revealed that there might be probability of genetic predisposition for ureterocele [10]. However, the etiology still remains unknown. Multiple theories have been presented in this respect; however, they have not explained fully the etiology [11-13].

Presenting complaints of ureterocele

The majority of studies have stated that most of the patients are diagnosed before they reach the age of 3 years [14-16]. In the last few years the number of antenatally diagnosed children has increased. In the early years of life children manifest urinary tract infection. In young children, the most common clinical presentation is a UTI. Others may present with ambiguous symptoms of failure to thrive or nonspecific pain in pelvic or abdominal region. At other times a renal unit may be obstructed with a sequel of a palpable abdominal mass. At very rare occasions ectopic ureterocele may result in urethral obstruction if prolapsed. In case of female gender, prolapsed ureterocele may mimic as a vaginal mass [15,16]. There may be a very unusual phenomenon of urinary incontinence if a huge intraurethral ectopic ureterocele is present that may lead to lax external urinary sphincter. Hematuria is a very uncommon presentation. While ureterocele are simple in older age children and adults, these are either normal or mildly dilated (have a single collecting system). They are incidentally uncovered, however urinary tract infection, stasis and consequent stone formation might be observed in these older age children or adults [1,16].

Anatomy and histology of ureterocele

Ureterocele may present in form of minute cystic dilatation (of distal segment of the intramural ureter) or as a huge balloon shaped that obliterates the bladder space. Microscopically, its wall comprises of diminished amount of smooth muscle fibers and fibrous tissue. The luminal surface is lined with ureteric mucosa [1,4,8,13,17].

Investigations

Initially, ultrasound can be performed, a non-invasive imaging tool to reach the diagnosis [14-18]. Voiding cystourethrogram (VCUG) facilitates in detecting the ureterocele, it also helps in detecting the grade of vesicoureteral reflux (VUR) if present. VCUG is a useful investigation that facilitates not only to diagnose VUR but it also detects any new reflux that may come up post endoscopic treatment. Reflux can be detected in ipsilateral lower pole in more than 50 percent of the affected patients, however, the contralateral system can be affected in almost 1/4th of these patients. Intravenous pyelogram (IVP) and Radionuclide studies have also shown to be useful tools to identify the non-functioning poles thus making it easy to plan the surgical management [13-16]. Dimercaptosuccinic acid (DMSA) or mercaptoacetyl triglycine (MAG3) renal scans are used to assess the distribution of function in the duplex kidney [17-22].

Management

Factors influencing management: Management strategy may vary from patient to patient [18-21] and is influenced by various factors such as age, presenting complaints, presence of any reflux, functional capacity of each renal segment in case it is associated with a duplex system and complications like urinary tract infection [20,21].

Goals of management: The primary target while formulating the management strategy of ureterocele incorporates the utmost

safeguarding of renal function. It thus necessitates arresting progressing severity of VUR in early stages. Secondly maintaining unimpeded flow of urine from the functioning parenchyma. Thirdly, halting any development of bladder wall defects due to the bladder outlet obstruction and treating it in time if detected. It is pertinent here that care should also be taken for maintaining continence mechanisms. Lastly and most importantly to get rid of any probable source that may lead to infection to lessen chances of overall surgical morbidity [19-22].

Duplex collecting systems: The management strategy of duplex collecting systems is guided by various factors such as the extent of renal impairment, recurrent infections and related morbidities. For example in case of simple duplex systems where there is absence of dilatation of either moiety, with no associated obstruction or when there is no reflux, no intervention is required. However hemi nephrectomy can be opted in case of complex duplex systems where there is evidence of diminishing renal function secondary to renal impairment or when the function of a moiety is below 10%. Heminephrectomy can also be considered in cases where conservative treatment with antibiotic prophylaxis has failed [22].

Non-surgical management: Patients antenatally diagnosed with ureterocele but without any symptoms can be put on conservative approach [23-25]. However due to insufficient data regarding conservative approach and their resultant longer follow-up outcomes, this approach cannot be deemed as a judicious option. In case of adults, it may be detected incidentally and as such does not require treatment unless it is complicated by calculus. In adults after ureterocele puncture for stones the chances of postoperative reflux are less likely [12,19,25,27].

Optimal timing of intervention: The optimal time regarding the timing of surgical procedure has remained a dilemma, however some authors are of the view to adopt early endoscopic incision of ectopic ureterocele in neonatal age-group. It not only relieves the obstructed drainage system of both the involved segments but also stops progression of further renal function impairment and recurrent infections [22-27]. This early drainage permits more time even if delayed reconstruction of the affected system is required later on. It is imperative to follow these children up for development of UTI, vesicoureteral reflux, renal function and longterm sequel in form of hypertension [23-28]. Some authors think on the contrary and stress more on wait and see policy in children having no preexisting VUR [25-29]. Despite varying opinions, endoscopic ureterocele incision can be opted in early stages as it is easy to perform, less invasive and can be done as a day case procedure [28-30].

Transurethral endoscopic incision: Some studies suggested that simple transurethral incision can relieve the overall patient condition and the patients may only require a long-term regular follow up [22-24]. Blyth et al. suggested that endoscopic incision of ureterocele proved to be an ultimate success in most of the children treated for ureterocele in their cohort [21-25].

Electrosurgery and laser technology: Transurethral endoscopic decompression of ureterocele can be undertaken by various techniques depending on each center's expertise and availability of equipment. Electrosurgery has been utilized widely in the recent past with promising results, however, new advances are taking place with the introduction of laser technology called as holmium: yttrium-aluminum-garnet (Ho:YAG) laser [27-29].

Advantages of laser

Accurate vaporization: It is an accurate technology as it vaporizes tissue site without much need for manipulation as is seen in cauterization and incision techniques utilized while applying electrocautery and cold knife incision, respectively. Furthermore, the incision and puncture tissue do not reseat as noted in other techniques [27-28]. Single or multiple punctures, or incisions can be utilized while treating ureterocele. Recently utilization of multiple punctures with holmium laser has been illustrated as an efficacious and effortless procedure, with shortened hospital stay in newborns, infants, and children alike [28-29].

Precise thermal effect: While utilizing laser fibers, there is less danger of spread of thermal effect beyond the site of incision. While in electrosurgery there is danger of thermal injury to surrounding tissue. This gives the surgeon room for precise multiple small punctures in the ureterocele [27-29]. Multiple small punctures with a laser fiber result in ample vaporization of obstructing tissue and thus relieve the obstruction adequately.

Studies regarding comparisons of ureterocele treatment outcomes by utilization of either Ho:YAG laser or electrosurgery are still lacking and as such definitive superiority of one procedure over the other mandates further research studies.

Half-loop technique in bilateral ureteroceles: Ureteroceles are rare in adult age and few cases have been encountered having bilateral ureteroceles. It is a challenging task to deal with such difficult cases. Half-loop technique has been proposed recently to treat bilateral ureteroceles [29].

Extravesical ureterocele management: As far as an intravesical ureterocele is concerned incision of ureteroceles can avert risk of hydronephrosis and thus conserve the remaining functional capacity of the affected kidney, however, it is not the same for extravesical ureterocele, because incision in such a case has inherent risk of reflux from the incised lesion. For this reason, endoscopic incision is avoided while treating extravesical ureteroceles and other aggressive options including, ureteroureterostomy, partial nephrectomy, and uretero-cystoneostomy are considered to tackle extravesical ureteroceles [30-32].

Other surgical options

Various strategies may include only endoscopic incision which at times may need further reconstruction of the urinary tract [21-30]. Other options include upper pole heminephrectomy with or without need for any lower segment tract reconstruction and watch and see policy [25-28].

Upper pole heminephrectomy: In this option, upper pole heminephrectomy is undertaken without lower tract reconstruction. It is preferred in subjects having ureteroceles with non-functioning upper segment moiety in the presence of mild degree of vesicoureteral reflux [15,18,28].

Lower tract reconstruction in addition to upper pole heminephrectomy: This technique is useful for subjects wherein there is a non-functioning upper pole moiety along with high-degree of vesicoureteral reflux into the same side lower pole ureter or on the contralateral side [21-28]. In neonatal age it may be a difficult approach to reimplant the refluxing ureter [24-27]. For this reason, a two-step procedure may be adopted wherein initially a ureterocele is decompressed along with the heminephrectomy at an early stage then as the child grows up, delayed ureteric reimplantation may easily be undertaken [24-29].

Lower tract reconstruction without upper pole heminephrectomy: Subjects who have functionally intact upper pole segment may do well with lower tract reconstruction without upper pole heminephrectomy. After puncturing the ureteroceles, both the ureters are dissected and reimplanted into the bladder in a submucosal fashion [20-27].

Total nephroureterectomy: It is undertaken in cases where there is severe lower pole ureteric reflux with both upper and lower pole moieties being non-functional [1]. Ureterocele is excised along with total nephroureterectomy [26-33].

New advances in management of ureteroceles

Transvesical laparoscopic surgery: Transvesical laparoscopic surgery has been recently tried wherein transvesical laparoscopic cross-trigonal ureteral reimplantation for correction of vesicoureteral reflux is undertaken. It is associated with diminished postoperative surgical site pain, shortened hospitalization period and acceptable cosmesis [34].

Laparoscopic ureteral ligation of ectopic ureter: In ectopic ureteroceles where the upper moiety is non-functioning, laparoscopic ligation (clipping of ureter) can be undertaken. It is a better and easy approach in the treatment of incontinence secondary to an ectopic ureter [35].

Laparoscopic selective clipping of upper moiety vasculature and ureter: Laparoscopic selective clipping of upper moiety vasculature and ureter has been recently tried to deal with ectopic ureter associated with poor functioning renal moiety. It has an advantage of avoiding partial nephrectomy and potential morbidity that can affect the ipsilateral normal functioning moiety inherent with partial nephrectomy [36].

Endoscopic Urinary Diversion (EUD) for ectopic ureter: It is a practicable, safe and minimal-invasive technique tried recently for the initial management of symptomatic obstructive ectopic ureter. It not only allows an ample ureteral drainage preserving renal function but also buys

more time for a definitive procedure/repair if needed later on [37].

Diagnosing ureteroceles in time and applying a suitable treatment strategy tailored according to each individual case is pivotal for attaining treatment success. Recent advances in treatment strategies should be updated and general surgeons and urologists should be able to do timely referrals to specialized centers for pediatric urology in case of its availability in their city or country. Patient counselling regarding follow up after treatment is also imperative [38,39].

Conclusion

Timely diagnosis and appropriate treatment strategy is the hallmark of success for ureteroceles' management. Primary ureterocele incision by utilizing modern endoscopic techniques has made its management easier and less invasive. Endoscopic incision of ureterocele proves to be the definitive treatment in most of the children. With the advent of modern technologies in the form of laser and laparoscopic techniques, its management has made strides. Follow up after treatment is imperative for management of such cases.

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