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REVIEW ARTICLE

Ureterocele: Review of Presentations, Types and Coexisting Diseases

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Abstract

Introduction: Ureterocele is swelling in one of the ureters that carry urine from the kidney to the bladder, which can block urine flow. An ureterocele usually occurs in the lower part of the ureter, where the ureter enters the bladder. Ureteroceles are equally common in both left- and right-side ureters. Some persons with ureteroceles are asymptomatic. Often, the diagnosis is made later in life due to kidney stones.

Materials and methods: A systematic review was accomplished using Medline to obtain articles related to ureteroceles in English, Spanish, Italian, and French. Interests were focused on presentations, types and coexisting diseases for ureterocele.

Results: Ureterocele can present with urinary tract infection, urinary retention, urinary incontinence, abdominal or loin pain, abdominal or vulvar mass, abdominal distention, hematuria, or vaginal bleeding. We can type the ureteroceles based on its location (ectopic or intravesicle), its association with triplicate system, duplication system, or single system, prenatal or postnatal, pediatric or adult, female or male, and thick or non-thick. The co-existing diseases include ureteral calculus, tumor, and congenital urinary tract abnormalities. Large ureteroceles are usually diagnosed earlier than smaller ones. An ureterocele may be discovered during a pregnancy ultrasound. Children with this condition often have severe urinary tract infections.

Conclusions: The review suggests that there are various presentations, types and coexisting diseases for ureteroceles. Findings implicate the importance of considering these variables when making treatment decision in patients with ureteroceles.

Keywords

Ureterocele, Presentation, Types, Coexisting diseases

Introduction

An ureterocele is a congenital abnormality that ureter balloons at its opening into the bladder, forming a sac-like pouch. Ureteroceles occur in about 1 in 500 to 1 in 4,000 people, at least four fifths of who are female. Patients are frequently Caucasian. An ureterocele usually occurs in the lower part of the ureter, where the ureter enters the bladder. Ureteroceles are equally common in both left- and right-side ureters. It is most often associated with a double collector system. Simple ureteroceles, where the condition involves only a single ureter, represents only 20% of cases. [1-29]. Two key steps in the urinary tract ontogenesis may be related to ureterocele development: formation and migration of the ureteric bud and its incorporation in the bladder [30]. Some persons with ureteroceles are asymptomatic. Often, the diagnosis is made later in life due to kidney stones. Since the advent of the ultrasound, most ureteroceles is diagnosed prenatally. The pediatric and adult conditions are often found only through diagnostic imaging performed for reasons other than suspicious ureteroceles. The signs and symptoms of ureteroceles in the latter two forms can easily be confused with other medical conditions. Symptoms can include: frequent urinary tract infection (UTI), urosepsis, obstructive voiding symptoms, urinary retention, failure to thrive, hematuria, and cyclic abdominal pain. A minority of ureteroceles is discovered incidentally during ureteral reimplantation for vesicoureteral reflux [1-29].

Many other complications arise from ureteroceles. Because of the distal ureteral obstruction, the ipsilateral renal moiety often is hydronephrotic or dysplastic. The degree of hydronephrosis may fluctuate depending on the amount of urine produced by the renal moiety. Ureteroceles is also associated with poor kidney function due to frequent blockage of the ureter. Blood

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pressure may be high if there is kidney damage. In other cases, a small upper portion of the kidney is congenitally non-functional. Though often benign, this problem can necessitate the removal of non-functioning parts. Redundant collection systems in double system are usually smaller in diameter than single, and predispose the patient to impassable kidney stones. The effective "bladder within a bladder" compounds this problem by increasing the collision of uric acid particles, the process by which uric acid stones are formed. A urinalysis may reveal blood in the urine or signs of urinary tract infection. The following tests or procedures may be performed: pyelogram, abdominal ultrasound, CT (computerized tomography) or MRI (magnetic resonance imaging) [31] scan of the abdomen, VCUG (voiding cysto-urethrogram), radionuclide renal scan, and cystoscopy. To guide surgical management, intraoperative retrograde ureterocelogram to further define the ureterocele anatomy could be valuable in some complex cases [32]. Antibiotics are usually given to prevent further infections until surgery can be done. The goal of treatment is to get rid of the blockage. Stents may provide short-term relief of symptoms. Surgical repair of the ureterocele usually cures the condition [1-29,33]. Primary endoscopic ureterocele treatment seems to be an appropriate option for children with a clinically significant ureterocele. The rate of secondary procedures was higher for ectopic ureteroceles (EUCs) but acceptable compared to the upper tract approach [34]. Other study suggested that primary endoscopic deroofing with double-J stenting for obstructing ureterocele is the best initial approach for adequate decompression and reducing the rate of secondary surgery [35]. In some complex cases heminephrectomy, ureteroureterostomy, or ureterocystoplasty may be required and many of these can be completed robotically [36-41].

Materials and Methods

A systematic review was accomplished using MEDLINE and article bibliographies to obtain articles related to endoscopic management of ureteroceles in English, Spanish, Italian, and French. Exposures of interest were presentations, types and coexisting diseases for ureterocele.

Results

Presentations

Urinary tract infection; urosepsis: Urinary tract infection is usually the most common postnatal presentation of ureterocle, patients presented with cloudy urine, urinary pain, and fever [42,43]. In a study with 40 children cases, first clinical manifestation was UTI in 87.5% [44]. In another study with a total of 37 patients who were diagnosed and treated for ectopic ureterocele, UTI was the most frequent mode of presentation (59%) [45]. A case of ectopic ureterocele in a 50-year-old male is reported. He was admitted to the hospital with complaints of miction pain, cloudy urine and fever attack. He had had pain at voiding from time to time during the past 10 years. The prostate was swollen, edematous and tender. Cystoscopy revealed a large bulge in the left trigone to bladder neck, and two ureteral orifices in the right, but none in the left. Complete duplication of the right ureter, lateral deviation of left ureter, and bladder deviation to the right were detected on the drip infusion pyelogram. An ectopic ureteral opening was present in the posterior urethra [46]. In the setting of untreated UTIs and hydronephrosis, affected older children and adults may reveal signs and symptoms of pyonephrosis and/or frank urosepsis, hyperammonemia. This sometimes could be life threatening [47-49]. One study report on a boy with ureteroceles that obstructed the bladder outlet and ureters, who presented with sepsis and hyperammonemia despite normal liver function. The hyperammonemia was most likely caused by excessive absorption of ammonia produced by Proteus mirabilis in the obstructed urinary tract [45]. In other report 3 infants with subphrenic abscess, pyonephrosis, and obstructive ureterocoele respectively had grossly increased concentrations of plasma ammonia. This was considered to be a result of infections with urea splitting organisms. All died in spite of intensive care support, including specific measures to reduce plasma ammonia [48].

Urinary retention; obstructive voiding symptoms; incontinence: Hydroureteronephrosis and bladder outlet obstruction are very common presentations of ureteroceles. This may lead to significant decline in kidney function [50]. During the physical examination, particular attention should be paid to the abdomen and the genitalia [51]. In a study on children who have single-system ureteroceles, there were 35 ureteroceles in the 32 patients; prenatally detected hydronephrosis or cystic renal dysplasia was the most common presentation (24 patients) [52]. Obstruction of the bladder outlet is commonly caused by prolapse of the ureterocele into the urethra. A prolapsing ureterocele in a female patient may cause physical obstruction of the bladder neck. Anatomic obstruction of the bladder neck by the cystic ureterocele may incite obstructive voiding symptoms or may precipitate acute urinary retention in both pediatric and adult populations [5,6,53].

Prolapse of the ureterocele and its presentation as a vulval mass is an extremely rare condition. During a female genital examination, a prolapsing cystic mass may be seen emerging from the external meatus in young girls or older women. This is a sign of a prolapsing ureterocele. However, the differential diagnosis of a prolapsing mass in children also should include urethral prolapse, sarcoma botryoides, and urethral caruncle. A 36-year-old female teacher, presented with a bleeding mass in the introitus, difficult micturition, and dysuria of four months duration was studied. On examination the mass was firm, round in shape about 2.5 cm in diameter, dark red in color with a catgut stitch and areas of necrosis. The patient was investigated and a diagnosis of a prolapsing ureterocele was made [5]. Other case report presented a newborn girl whose ureterocele ex-

tending through the urethra [6]. Ureterocele prolapse is a rare presentation of single system ureteroceles and is usually found early in childhood. One study reported a rare case of recurrent prolapse of a single system ureterocele that did not present until the patient was 17 years of age [53]. Bladder neck obstruction can even be caused by a large simple nonprolapsing intravesical ureterocele; it has been reported in an adult male [54].

Once an ureterocele is identified sonographically, a VCUG to detect vesicoureteral reflux (VUR) and a 99 m-technetium dimercapto-succinic acid renal scan to evaluate the function of the different portions of the kidney are mandatory. Vesicoureteral reflux in the lower pole is observed in 50% of cases and in the contralateral kidney in 25% [55]. In a study with 12 patients who had ectopic and 2 orthotopic ureteroceles. Vesicoureteral reflux was present in 8 of 12 (67%) ectopic systems before incision and 9 of 10 (90%) after. None had resolved reflux during follow-up [56]. In another study of 51 patients with a duplex system and associated ureterocele 19 (37%) required a secondary open procedure. The ureterocele was intravesical and ectopic in 22 (43%) and 29 (57%) cases, respectively. Reflux was associated with the ureterocele in 27 patients (53%) [57].

Children with EUC are at high risk for a high capacity bladder with incomplete emptying. In a study, 34 patients with a mean age of 10 months were treated for large or medium ectopic ureteroceles at our institution and 32 participated in postoperative followup. Of the 32 patients 19 had infrequent voiding and 3 had incontinence. Cystometric bladder capacity was increased to greater than 150% of the normal value for age in 15 of 27 patients (55%). Uroflowmetry revealed greater than 50 mL residual urine in 15 patients (56%). Postoperatively no radiological signs of bladder neck obstruction were found. Increased bladder capacity and residual urine did not correlate with ureterocele size or location, or surgical procedure. There was no progression of bladder dysfunction with age [58]. In another study of 39 cases with EUCs, one patient presented with urinary incontinence [59].

Abdominal pain or loin pain: The ectopy of the ureter orifices and ureterocele are always followed by either mechanical or dynamic obstruction of the ureter and obstructive pyelonephritis. These anomalies are periodically accompanied by abdominal pains which make their appearance during an attack of acute pyelonephritis. Cyclical expansion and decompression of the renal pelvis manifests as intermittent abdominal pain in older children and adults. Ureterocele is a cause of abdominal pain or loin pain [60-63]. These abdominal pains can be erroneously taken for symptoms of appendicitis or intestinal obstruction and the patients are subjected to appendectomy or laparotomy by mistake. In one study, the erroneous appendectomy or laparotomy was occurred in 47 of 201 patients with ectopy of ureter orifices and ectopic ureterocele. The differentiation of genesis of abdominal pains may be more exact with the help of chromocystoscopy, excretory urography and isotopic renography [63]. Symptomatic ureteroceles with hydronephrosis also manifest as signs of abdominal tenderness to palpation.

Abdominal mass, abdominal distention, and vulvar mass: Ureterocele can be huge in size [52,64,65] and present as abdominal mass [52], abdominal distention [59,65]. An abdominal mass due to a large hydronephrotic kidney may be appreciated in the upper abdominal quadrant in thin adults and young children. Flank tenderness often accompanies the abdominal findings. In infants an abdominal mass due to hydronephrosis may be noted by transillumination in a dark room. In a study of ectopic ureteroceles duplicated system of the kidney was involved in 35 cases (42 kidneys) and single system in 4 cases (4 kidneys). Abdominal distention was the second most common mode of presentation [59]. Very rarely, a prolapsed ureterocele could present like a vulvar tumor [66].

Congenital giant megaureter: Neonatal presentation of a congenital giant megaureter is a very rare unilateral urinary anomaly. It can be secondary to ureterocele [67].

Hematuria: Gross hematuria most often had a benign cause in children and adolescents. In a study with 342 patients, 2 boys and 1 girl had ureterocele. It was more common in boys for almost all etiologic categories and ages [68]. Patient with ureterocele can also present with isolated macrohematuria [69].

It is important to exclude other causes of hematuria before making a diagnosis. In a study a young male patient that had been misdiagnosed as having an ureterocele causing ureterohydronephrosis and hypertension had undergone nephrectomy. Pathological analysis of the surgical specimen revealed the underlying cause of the patient's condition. Pheochromocytoma is a rare disease entity whose most common form of presentation is hematuria or that which results from catecholamine produced by the tumor. Lesions located close to the ureteral meatus may cause obstruction. If this condition is not suspected and the findings of diagnostic imaging are inadequately interpreted, its diagnosis and treatment may be delayed thereby increasing the surgical risk due to its secretary nature [70].

Vaginal bleeding: Rarely ureterocele can present as vaginal bleeding [5,61], as it did with the 36-year-old female teacher previously mentioned [5].

Forniceal rupture with increased urine flow: Very rarely, ureterocle presented as forniceal rupture with increased urine flow. The ureterocele ruptur followed hypophysectomy for Cushing's disease and high urine outflow [71].

Genetic ureterocele?: Definitive causes of ureteroceles have not been found. While the abnormal growth occurs within the uterus, it has not been substantiated that genetics are to blame. One study reports two pairs of twins one of each set had ureteroceles, and one other a related urinary tract malformation (ureterocele, polycystic kidney disease). These cases raise the question of whether the siblings of children with ureteroceles should be screened for urogenital abnormalities [72].

Types

Types based on location

Bilateral or unilateral: Ureteroceles are equally common in both left- and right-side ureters. In a study with 39 cases, the left EUCs were found in 15 patients, the right in 17 patients, and the bilateral in 7 patients [59]. Another study on children who have single-system ureteroceles: There were 35 ureteroceles in the 32 patients, 29 were unilateral (14 right-sided, 15 left-sided) and 3 were bilateral [52].

Ectopic ureterocele or intravesical ureterocele: An EUC may be defined as a cystic dilatation of the distal submucosal or intravesical portion of a ureter that is associated with the ectopic moiety of a completely duplicated system or, in the absence of duplication, associated with a ureter draining into an ectopic position. More than 50 per cent were less than 3 years old at the time of presentation. The female preponderance was 3 to 1. Urinary infection was the most frequent mode of presentation [73]. In a study, all 9 of the patients with a single system ureterocele had an intravesical ureterocele. No patient had associated reflux nor did any require a secondary open procedure. In 3 cases new onset ipsilateral reflux into the ureterocele spontaneously resolved. In another group of 51 patients with a duplex system and associated ureterocele 19 (37%) required a secondary open procedure. The ureterocele was intravesical and ectopic in 22 (43%) and 29 (57%) cases, respectively. Reflux was associated with the ureterocele in 27 patients (53%) [57]. In the majority of ureterocele cases it is accompanied by a double-collector system and affects 6:1 female child patients. There were 40 cases in an observational, longitudinal, transverse, descriptive, retrospective, and open study: 50% male, average age 20 months; ectopic type was found more frequently with 62.3% and orthotopic or simple, in 37.5% [44].

One study described four patients with unusual anatomic presentation of EUCs and their surgical treatment. Over a 3-year period four cases of unusual EUCs were encountered. A 6-month-old girl had a complex cloacal anomaly with an ECU within the cloaca. A 10-year-old boy had two large diverticuli within a EUC combined with a blind-ending ipsilateral ureter. A 3-year-old girl had a EUC combined with a periureteral diverticulum and a completely duplicated ipsilateral kidney. A 4-year-old girl was found to have a vaginal EUC. Despite thorough radiological investigation in all patients a correct assessment of the anatomic defect was achieved only by surgical exploration or endoscopic evaluation. If preoperative radiological evaluation is equivocal, a high index of suspicion and intraoperative recognition of an unusual anatomic presentation of the EUC are essential for appropriate management and a successful outcome [74].

In one study types and variants of ureterocele are specified basing on 115 clinical cases and literature data. Three types of this anomaly exist: intravesical, cervicosphincteral and extravesical. Intravesical ureterocele can be orthopedic and ectopic. The latter has three variants: of accessory ureter, of partially double ureter, of non-double ureter. Ureterocele is frequently associated with low ectopy of ostium ureteric, double ureter. Ureterocele occurs neither in the ureter itself nor in high ectopy of the ostium ureteric. Extravesical ureterocele has many variants which differ in males and females. All ureterocele types and variants occur as unilateral or bilateral. The new diagnostic method is proposed: endovesical ureterocelegraphy [75]. The control of infection became easy in all patients except for one with a sphincteric ureterocele [76].

Ureterocele associated with triplicate system, duplicate system or single system: The EUCs are frequently associated with double urinary systems. The EUCs with duplicated system of the kidney were involved in 35 cases (42 kidneys) and single system in 4 cases (4 kidneys) [59]. Single-system ureterocele is an important, although uncommon, cause of hydronephrosis and renal dysplasia in infants and children. Single-system ureterocele is distinguished clinically from the more common duplex-system ureterocele by its frequent occurrence in boys and its association with multicystic dysplastic kidney. Because these ureteroceles are frequently small and have a propensity to evert at VCUG, they can be mistaken for paraureteral diverticula [52]. Duplex systems are more likely to cause urethral obstruction in males although they occasionally can occur with just a single system. In one study, four types of ureteroceles are described: A) ureterocele with single ureter (10%); B) ureterocele with total duplication and intra-vesical development (10%); C) ureterocele with total duplication and extra-vesical development (62%); D) ureterocele with ectopic ureter (3%). Most ureteroceles are now detected by antenatal ultrasonography, allowing early management. The complicated forms may require either meatotomy for decompression or diversion by percutaneous nephrostomy. Strangulation of the ureterocele constitutes an emergency [77]. There were also case reports of ureterocele in a triplicate system [78-80] and a ureteral triplication with a contralateral duplication and ureterocele [81].

Prenatal and postnatal ureterocles: In one study 41 ureteroceles were in duplex kidney. In 33 cases (70%) the prenatal ultrasound had observed hydronephrosis or

duplex kidney or the presence of the ureterocele. These babies were free of urinary infection before treatment [82]. Historically, most patients with a renal duplication anomaly associated with upper pole hydronephrosis underwent upper pole nephrectomy and partial ureterectomy. Prenatal sonography has resulted in increased recognition of these anomalies and, therefore, earlier urological referral and evaluation. Although there is still controversy regarding the impact of prenatal detection of hydronephrosis on the preservation of renal function in patients with a single system, one study revealed a beneficial effect in patients with duplication anomalies [83]. The other study postulated that prenatal detection of ureteroceles has a positive impact on the natural history and clinical outcome of ureteroceles in duplex system. Mean followup in the 2 groups was 3.9 years. Preoperatively the reflux rate was 51% in the prenatal and 66% in the postnatal groups. Preoperatively UTIs were less common in the prenatal group (12% versus 84%). Mean age at initial intervention in prenatally and postnatally diagnosed patients was 6 and 31 months, respectively. Postoperatively the UTI rate was double in postnatally diagnosed patients. Overall prenatal diagnosis is associated with a decreased rate of secondary procedures independent of the type of ureterocele [84].

Ureterocele in pediatric or adult patient: In a study with a total of 37 patients who were diagnosed and treated for EUC, 54% were less than 3 years old at operation [45]. In one study on children who have single-system ureteroceles. There were 35 ureteroceles in the 32 patients. Mean age at presentation was 0.7 years (0-9.2 years) [52]. Currently most pediatric ureteroceles are found incidentally during routine screening antenatal ultrasound. Adult ureteroceles also are found incidentally during imaging studies for urologic complaints of usually unrelated symptomatology. Ureteroceles are interesting radiologic curiosities that often do not have clinical sequelae in the adult population. Ectopic ureteroceles, while not uncommon in children, have been reported only rarely in adults. One study presented five adults with EUCs with emphasis on the varied clinical and radiographic manifestations. These findings were compared with those in 32 children with EUC. It was found that the clinical presentation differed in adults and children, though the radiological findings were similar. The diagnosis was in some cases delayed for many years. The anomaly could not be detected by imaging means in two of five adults and eight of 32 children, and was found only at surgery [85].

Ureterocele in female or male patient: For EUC, the female preponderance was 3 to 1 [75]. In a study a total of 37 patients was diagnosed and treated for ectopic ureterocele, female-to-male ratio 3.6:1 [45]. In another study on children who have single-system ureteroceles. There were 35 ureteroceles in the 32 patients. Twenty-five patients were boys (78%) and 7 girls [52]. Relapsing ureteroceles also can occur in boys, but they are much less common. Ureterocele and stone were the main findings in girls and boys with acute urinary retention, respectively, and urinary retention in boys was twice as prevalent as that in girls [86].

Thick or non-thick ureterocele: Ureterocele wall thickness was assessed subjectively via radiographic and cystoscopic methods and categorized as thin, intermediate and thick. Patients with a thick walled ureterocele required repeat puncture more frequently than those with a nonthick ureterocele. The mode of presentation does not predict the need for a repeat open procedure. Thick walled ureteroceles require repeat endoscopic puncture more frequently than thin and intermediate walled ureteroceles [57]. In a study with 5 boys and 9 girls, mean patient age at presentation was 17.5 months. Ureterocele was defined as thick if ultrasound measurement was 4 mm. or greater. Thick ureterocele was present in 4 (28%) patients [56].

Cecoureterocele: The cecoureterocele differs from the usual ectopic ureterocele because it extends outside the bladder and may obstruct the urethra. The differentiation requires careful examination of the urethra by a voiding cystourethrogram and cystoscopy. One study described the management of a girl with bilateral cecoureteroceles who presented as a newborn with urinary retention and sepsis [87]. The more accurate diagnosis of cecoureterocele was not suggested by preoperative radiographic studies. The cecocele component was disclosed only at endoscopy or during open resection of the ureterocele [88,89]. Two studies reported some cases of cecoureterocele to emphasize the salient features of this rare clinical entity. The differentiation of it from the usual ectopic ureterocele requires careful examination of the urethra by VCUG and cystourethroscopy [90,91].

A rare case of single system cecoureterocele associated with ipsilateral dysplastic kidney in a 4-year-old girl was reported. The preoperative radiological and endoscopic investigations identified ureterocele but not its cecal extension. The latter was diagnosed only during the transvesical surgery [92]. The ureterocele within the bladder base and the entire urethra were examined macroscopically and microscopically in serial sections. The ureterocele and its cecal extension were amuscular. The bladder neck and the involuntary and voluntary sphincters were attenuated or devoid of muscle in the quadrant beneath the ureterocele and its cecal extension. These findings were correlated with the clinical features of 2 living patients with comparable ureteroceles who had urinary incontinence following surgical excision of the ureteroceles [93]. In another report, a three-year-old female had a nephroureterectomy for a hydronephrotic left kidney with a total duplicated collecting system. Postoperatively urinary retention developed which was found to be secondary to a previously nonobstructive cecoureterocele [94].

Co-existing diseases

Ureterocele coexists with ureteral calcus: Ureterocele complicated with ureteral calcus is not uncommon

[95-97]. One retrospective study was based on 20 cases of ureterocele in adults complicated by stones. This series consisted of 8 men and 12 women with a mean age of 48.3 years. The clinical features were dominated by low back pain. The ureterocele affected a single ureter in 16 cases (80%) and a duplex ureter at the expense of the upper renal segment in 4 cases. Adult ureterocele complicated by stones is a well-tolerated, rare entity that can often be easily diagnosed [96]. To children with orthotopic ureterocele complicated with ureteral calcus the majority of children have definable metabolic abnormalities [97].

Ureterocele coexists with tumor: One study presented a case of urothelial tumor inside a simple ureterocele with a coexisting contralateral tumor that caused bilateral obstructive uropathy. Follow-up evaluation of a 74-year-old male patient with superficial tumor of the urinary bladder diagnosed 4 years earlier revealed renal failure and bilateral obstructive uropathy. The complementary tests showed a simple left ureterocele containing a lesion suggestive of a tumor and a right ureteral tumor. Later a biopsy confirmed coexisting urothelial disease at 3 levels (intraureterocele, contralateral ureter and in situ carcinoma of the bladder). Multicentric synchronous urothelial tumors warrant a close and long follow-up of the entire urinary tract [98]. Similar intraureterocele tumor was reported in a 71-year-old male [99].

Ureterocele coexists with congenital urinary tract abnormalities

Ureterocele coexists with duplicate system: It is most often associated with a double collector system, where two ureters drain their respective kidney instead of one. Simple ureteroceles, where the condition involves only a single ureter, represents only twenty percent of cases [44,72-74].

Ureterocele co-exists with multicystic disease: In a study with 33 ureteroceles 24 were associated with ipsilateral hydroureteronephrosis and 10 with ipsilateral multicystic dysplastic kidney [52]. One patient had a normal ipsilateral kidney and a contralateral multicystic dysplastic kidney. Single-system ureterocele is an important, although uncommon cause of hydronephrosis and renal dysplasia in infants and children. Single-system ureterocele is distinguished clinically from the more common duplex-system ureterocele by its frequent occurrence in boys and its association with multicystic dysplastic kidney [52]. Multicystic renal dysplasia (MCDK) is a common anomaly well described in the literature, but less well described when involving only a portion of a kidney. Four children had antenatal diagnosis of cystic renal abnormality. In two, with obvious duplex kidneys and associated ureteroceles, the diagnosis of upper moiety MCDK was obvious either antenatally or immediately postnatally. In these children, as in other studies, the most common presentation of segmental MCDK is in the upper pole of a duplex kidney associated with an ureterocele at the end of the atretic ureter [100].

Ureterocele coexists with polycystic kidney disease: Sometimes ureterocele can co-exist with polycystic kidney disease in pediatric patients, as it did with the patients previously mentioned [71].

Ureterocele associated with horseshoe kidney: Horseshoe kidney is a common renal fusion anomaly. Because of the 25% incidence of associated genitourinary anomalies we believe that the diagnosis of horseshoe kidney in pediatric patients should initiate a thorough urologic evaluation including intravenous urography and real-time sonography [101]. Several other cases were reported [102]. There was a report of a horseshoe kidney one side of which contains a multicystic dysplastic element associated with an ureterocele [103].

Ureterocele associated with crossed fused renal ectopia: Ureterocele can be rarely associated with crossed fused renal ectopia. The diagnosis and management become challenging in the presence of renal failure [104-108]. One study described a case of the unique congenital anomaly of cross-fused ectopic multicystic dysplastic kidney with associated ureterocele and demonstrated the usefulness of magnetic resonance imaging in fetal imaging [106]. Some cases were successfully managed by a simple transurethral incision of the ectopic ureterocele [108]. Crossed renal ectopia complicated many anomalies about 50%. Among them anomalies of the urinary tract was most frequent about 30% [108].

Ureterocele coexists with other congenital abnormalities: Symptomatic presentation of orthotopic ureterocele in infancy and its association with posterior urethral valves are rare but were reported [109]. Very rarely ureterocele can also be found to co-exist with hypopidiasis or myelomeningocele [52].

Conclusions

The review suggests that there are various presentations, types and coexisting diseases for ureteroceles. Findings implicate the importance of considering these variables when making treatment decision in patients with ureteroceles.

Conflict of Interests

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

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