Ureteral diverticulum: a review of the current literature

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Introduction: Ureteral diverticulum is a rare urological condition with only 45 cases described in the literature. These previously reported cases vary in their presentation, diagnosis and management and there is no consensus in the literature on the best diagnostic tool available. We describe our experience on diagnosing and managing this condition in two patients and provide a descriptive review of the current literature on ureteral diverticulum.

Materials and methods: A Medline search was performed to identify all reported cases of ureteral diverticulum. Key words used were: ureteral diverticulum; abortive bifid ureter; congenital diverticulum; acquired diverticulum. We also reviewed the records of two patients who presented consecutively to our institution with a ureteral diverticulum. The clinical and radiological characteristics of this entity were then evaluated.

Results: Forty-one manuscripts were identified, encompassing single case reports and case series, the largest of which contained seven patients. Two additional cases were diagnosed in our institution; a true congenital diverticulum and an abortive bifid ureter which is synonymous with a true ureteral diverticulum. Both were uncomplicated cases and were managed conservatively.

Retrograde pyelography was used for definitive diagnosis of this lesion.

Conclusion: Ureteral diverticulum may present as an incidental finding or with a secondary complication. Conservative management is advocated in the literature for non-complicated cases. Retrograde pyelography is our diagnostic tool of choice.

Key Words: retrograde pyelography, review, ureteral diverticulum, abortive bifid ureter, congenital diverticulum, acquired diverticulum

Epidemiology

Ureteral diverticula are rare urological entities with 45 cases described in the literature. The most frequently reported diverticulum is an abortive bifid ureter, ending blindly at one of the proximal ends1-6 and least commonly, acquired diverticulum that arise secondary to the presence of ureteral calculus7 or as a complication of surgery.8,9 Two further cases of ureteral diverticulum were diagnosed in our institution.

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Classification

Ureteral diverticula are sub-classified into three categories: 1) abortive ureteral duplications (a blind-ending bifid ureter: the same embryogenesis as diverticula, arising from disordered ureteric budding, different from diverticulum in its appearance); 2) true congenital diverticulum containing all tissue layers of the normal ureter; 3) acquired diverticulum representing mucosal herniation.10,11

The pathogenesis of an acquired ureteral diverticulum is a distally obstructed ureter. This may be due to a distal calculus causing an increase in pressure that results in a proximal ureteral diverticulum similar to the development of a vesical diverticulum in patients with benign prostatic hypertrophy. These
patients present in a symptomatic fashion with flank pain and fever. In addition, strictures, and ureteral valves may produce a similar picture and present symptomatically. Mucosal herniation may also result from a congenital weakness in the ureter that permits ballooning when the intra-ureteral pressure is increased. Ureteric calculi may also occur in conjunction with true congenital diverticulum, there are four such cases reported in the literature.

The first series of diverticula reported in 1975 concluded that although the potential for the development of these lesions may arise from congenital mal-developments, the natural history also depends on an abnormal hypo-dynamic state that may be acquired in childhood or adult life.

Diverticula can present with painless hematuria or be entirely asymptomatic and present as an incidental finding on imaging. The diverticulum may then become associated with obstructive uropathy when a ureteral calculus sits in the pouch and causes ureteral stenosis proximal to the stone filled pouch and consequent hydronephrosis. Pyelonephritis can then occur, causing sepsis necessitating the insertion of a ureteric stent to relieve the obstruction, as reported by Mori et al. They concluded that a diverticulum, unlike a ureterocele, will not cause obstruction, but a stone in the pouch will. In our institution, one patient had an abortive bifid ureter while the other had a true congenital diverticulum. Visible, painless hematuria was the presenting complaint in both cases and one was associated with a ureteral calculus which passed after cystoscopic insertion of a ureteric stent.

Ureteral diverticula have been described in association with complex pelvic and abdominal abnormalities and have been reported in the pediatric population in association with other congenital anomalies.

**Diagnosis and treatment options**

Radiological imaging is the mainstay of diagnosis among reported cases. Ultrasonography was initially advocated by Wan et al and Herndon then went on to describe the first antenatally detected ureteral diverticulum with the use of antenatal ultrasound. CT is generally used in symptomatic cases where a ureteral stone was subsequently found to be sitting in the pouch causing obstruction. However, the sensitivity of CTU in picking up diverticula associated with ureteric calculi may be debatable. In our experience, CT did not pick up the diverticulum in the patient with a ureteric calculus, Figure 1 but it was diagnosed in the other calculus-free case, Figure 2. The use of non contrast magnetic resonance tomography, CT and repeated target ultrasonography using color mapping and 3D reconstructions has also been reported. We used retrograde pyelography to confirm the diagnosis in both our patients.
In the majority of reported cases, and in our experience, ureteral diverticula do not require treatment. If the diverticulum becomes associated with obstruction, treatment is then initiated. We also elected for conservative management in our patients and at most recent follow up, both patients were well and asymptomatic.

There are early reports of nephrectomy and partial ureterectomy of the affected portion of ureter for management of diverticula associated with obstruction. Later, reconstructive surgery was also shown to have a role in alleviating obstruction secondary to diverticulum.

Diverticulectomy with segmental resection of the ureteral diverticulum with an end-to-end anastomosis is currently advocated in cases where the urine refluxes and stagnates, causing urinary tract infection. This may be done after interval placement of a ureteric stent to allow the sepsis to subside.

More recently, successful laparoscopic resection of acquired ureteral diverticula has been described by Li et al in 2006.

Prognosis

The general prognosis of untreated, uncomplicated ureteral diverticula is excellent however there have been reported cases of some adverse events. The occurrence of TCC in a lower ureteral diverticulum has been described and perforation of a diverticulum can also occur. Fibroepithelial polyps have also become associated with these lesions but it is hypothesized that ureteral diverticula and fibroepithelial polyps are part of a spectrum of the same developmental anomaly and do not have any bearing on prognosis.

Douglas et al reported on patients who developed hydroureteronephrosis and were subsequently treated with surgical excision. At 4 year follow up their renal function remained normal with no deterioration on excretory urography.

Conclusion

Ureteral diverticulum is a rare entity that has been classified in the literature into congenital or acquired lesions. They are generally asymptomatic unless they become associated with ureteral stones causing obstruction. We advocate the use of retrograde pyelography for diagnosis of these lesions as the sensitivity of CTU has not been proven. Conservative management should be employed in asymptomatic cases with no evidence of hydroureteronephrosis.

References

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