The ectopic ureter opening into the vulva, which is a rare cause of lifelong urinary incontinence: Treatment with ureteroureterostomy

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ABSTRACT

Persistent urinary incontinence (UI) in adults may indicate a congenital anomaly. Before initiating medical treatment in these patients, a detailed physical examination is necessary for establishing an accurate diagnosis. In this study, we report on a patient who presented with the complaint of congenital UI associated with ectopic ureter and was detected with a right complete duplex collecting system and a ureter opening into the vulva and the surgical treatment applied to that patient.

Introduction

Urinary incontinence (UI) in adult women results from acquired causes rather than congenital factors. However, female patients suffering from persistent UI since birth rarely present with a congenital ectopic ureter opening into the vestibulum. 1 Ectopic ureter is defined as a ureter opening into an area outside the posterolateral extremity of bladder trigone and more than 80% of the cases are associated with the complete duplex collecting system. An ectopic ureter is diagnosed by cystoscopy after taking a detailed clinical history of the patient and performing physical and radiological examinations. 2

In this study, we report on a patient who presented with the complaint of congenital UI associated with ectopic ureter and underwent surgical treatment.

Case

Patient history and physical examination findings

The 33-year-old female patient presented to our clinic with a history of persistent UI (day and night) and occasional abdominal pain since childhood. The patient was using disposable diapers (7–8 diapers/day) due to persistent UI. She had received medical treatment with two different anticholinergic agents in the other centers she had applied for UI, but her symptoms did not regress.

The urogenital examination revealed another small vestibulum under the urethra at 6 o'clock [Fig. 1]. When the Valsalva maneuver was performed, the patient developed a urine leak from this vestibulum. The stress test was normal in the functional ureter.

Imaging findings

Non-contrast computed tomography (CT) showed focal parenchymal thinning areas in the upper portion of the right kidney. On 99mTc dimercaptosuccinic acid (DMSA) static renal scintigraphy, the contribution of the left kidney to total renal function was 59.24% and the contribution of the right kidney was 40.76%. When the one-third of the upper and the two-thirds of the lower components of the duplex collecting system of the right kidney were evaluated, the contribution of the upper and lower components to the function of the right kidney was 33.35% and 66.65%, respectively.

Surgical treatment

The patient was diagnosed as having an ectopic ureter opening into the vulva and thus cystoscopy was performed in the lithotomy position under general anesthesia. Bilateral orifices were normal and no abnormality was observed in other areas of the bladder. Subsequently, a
Ureteric catheter was placed both in the ureter opening into the bladder and in the ureter opening into the vulva and retrograde pyelography (RGP) was performed initially through the catheter in the vulva and then through the catheter in the bladder. On RGP, the ureter opening into the bladder was observed to drain the lower pole of the kidney and the ectopic ureter was draining the upper pole of the kidney [Fig. 2A, Fig. 2B and C]. The ureters were completely separate from each other up to the kidney (complete duplex system).

Afterwards, a “right Gibson incision” was made by placing the patient in the appropriate position. The ureters on the right were exposed and then dissected. At a location 1–2 cm below the iliac crest, the ectopic ureter was excised and an end-to-side anastomosis was performed in the normal ureter, and then a double-J ureteral stent (6Fr 28 cm) was placed in both ureters.

**Postoperative follow-up**

On postoperative day 14, the UI resolved completely and the ureteral stents were removed. The patient had no symptoms in the subsequent follow-up visits and is still under routine outpatient follow-up. Postoperative 6 months, DMSA showed similar renal functions with the preoperative DMSA.

**Discussion**

Ectopic ureter implantation is an extremely rare urinary tract anomaly with an estimated incidence of 0.05–0.025%, and almost 80% of the cases are women. Ectopic ureteral opening is typically associated with single collecting system in men, while in women it often drains the dysplastic upper pole of a duplex kidney.1,2 Since the ectopic ureter is located below the external sphincter in women, they often present with the complaint of UI. However, the affected women have a normal voiding pattern despite persistent UI. In women, the most common localizations of ectopic ureter include bladder neck and upper urethra (33%), vulva vestibule that contains the opening to the urethra and the vaginal opening (33%), vagina (25%), and less commonly cervix or uterus. Although ectopic ureter is a congenital anomaly, sometimes it may not be diagnosed in early ages due to insufficient analysis of etiology and inadequate examinations, and these patients may be treated incorrectly due to misdiagnosis, as in our case.3

Primary goal in surgical treatment of ectopic ureter is to preserve renal function, to prevent recurrent episodes of infection, and to restore continence. The current approach used in the management of ectopic ureter is primarily aimed at making decisions and applying treatment according to the functional state of the kidney part drained by the ectopic ureter. If the kidney part drained by the ectopic ureter is nonfunctional, heminephroureterectomy can be considered; however, if this part has a sufficient level of functioning, more nephron-sparing techniques such as ureteroneocystostomy and ureteroureterostomy can be performed. Wang et al. performed heminephroureterectomy in the upper pole of a nonfunctional kidney and obtained successful results.

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**Fig. 1.** Normal ureteral meatus (right arrow) and the ectopic ureter opening into the area below (left arrow).

**Fig. 2.** Perioperative retrograde pyelography (RGP) images. A. The opaque material delivered through the ectopic ureter is seen to drain the upper pole of the right kidney. B. The opaque material delivered through the ureter in the bladder is seen to drain the lower pole of the right kidney. C. Ureteral catheters inserted for RGP.
outcomes. In another study, Demir et al. performed ureteroneocystostomy for a ureter that drained the upper pole of a functional kidney. On the other hand, ureteroneocystostomy is another technique used in patients with ectopic ureter, through rarely. Wong et al. performed ureteroureterostomy by achieving an end-to-side anastomosis with the ureter originating from the upper pole of the functional kidney and the ureter originating from the lower pole. In our patient, DMSA showed that functional capacity of the patient’s kidney was sufficient for the preservation of the upper pole of the right kidney. Accordingly, ureteroureterostomy, which is a more minimally invasive treatment option than ureteroneocystostomy, was preferred since it does not require bladder incision, and the procedure provided successful outcomes.

**Conclusion**

Although ectopic ureter is a well-known cause of persistent UI, these patients can still be missed. Accordingly, appropriate diagnostic methods should be utilized in the investigation of UI and in the differential diagnosis, it should be noted that the patient may have an ectopic ureter regardless of age.

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**Declaration of competing interest**

There are no conflicts of interest in connection with this paper.

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