Urinary incontinence in a young girl due to ectopic ureter: the importance of history in a diagnostic challenge

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Abstract

In girls who are otherwise well and whose history is that of continuous wetting day and night, despite successful toilet training, for a lifelong history, an extravesical infrasphincteric ectopic ureteral orifice should be strongly suspected and imaging should be vigorously pursued. Here, delayed diagnosis of vaginal ectopic ureter in a young girl with a lifelong history of urinary incontinence is presented. The importance of history and imaging procedures are also discussed.

Wetting is a common problem during childhood which includes majority of patients who have no underlying abnormality of the urinary tract. A minority of children are incontinent secondary to structural abnormalities. It is important to recognise these cases which will not improve spontaneously.

Ectopic duplex ureter is a rare cause of urinary incontinence in girls. A constant dribble of urine, every day and every night in a girl who has been successfully toilet-trained is the characteristic story of a young girl who has a ureter that drains ectopically outside the bladder and below the sphincter. This ectopic ureter usually carries urine from upper pole of a duplex kidney.

In this report we present 20-year-old girl with a lifelong history of urinary incontinence due to left duplicated ectopic ureter opening to the vagina. Also, imaging findings of intravenous urography (IVU), ultrasonography (USG) and computerised tomography (CT) are described.

Case report

A 20-year-old girl presented with a lifelong history of urinary incontinence. There were no other associated urinary symptoms and bowel control was normal. She received medical therapy including imipramine, anticholinergics, desmopressin and used alarm devices for diagnosis of enuresis nocturna but, could not be cured. There was no history of trauma or pelvic operation.

Physical examination didn’t reveal any evidence of neurological deficit of the lower limbs and perianal region, the anal tone was normal. There was no expressible or distented urinary bladder. Her chief complaint was wetting at night, however, it was detected in detailed anamnesis that she had wetting not only at night but also during the day, the amount of wetting declined through years but not ceased completely. IVU demonstrated left kidney with a missing of upper pole calices (Figure 1).

USG examination showed a cystic mass at the upper pole of the left kidney (Figure 2). Especially in the light of history and IVU findings ectopia of the ureteral orifice draining the upper moiety of a duplex kidney was strongly suspected and CT imaging was performed to confirm this and to determine the side of abnormality. CT scan
revealed a dysplastic duplex kidney located at left upper pole unit and drained by a
dilated ureter extending to the vagina (Figure 3-a and 3-b).

**Figure 1. IVU demonstrating missing upper pole calices at the left kidney (arrows)**

![Figure 1](image1)

**Figure 2. USG demonstrating left kidney with normal lower pole (solid arrow) and dilated dysplastic upper pole (curved arrow).**

![Figure 2](image2)
Figure 3-a. CT scan shows dilated dysplastic upper pole segment (left duplicated kidney)

Figure 3-b. CT scan shows normal left lower pole kidney (solid arrow) and dilated ureter of the left dysplastic upper pole moiety (curved arrow)

Exploration of the left kidney demonstrated a dysplastic upper pole segment with a draining dilated ureter. Partial nephrectomy (upper pole heminephrectomy) was performed and the dilated ureter was excised as far as possible with an extensive care not to compromising the blood supply to the ureter of normal lower pole segment. Postoperative recovery was uneventful and the patient was cured of her wetting.

Discussion

Wetting is a common symptom in children and may occur at night, during the day or at both times. Those who wet during the day or both day and night are the ones that need to be investigated. The majority of cases have functional causes. Organic causes are much less common but important because it will not improve spontaneously and may be curable with surgical intervention. Anatomic abnormalities causing incontinence of urine include spinal dysraphism, sacral agenesis and epispadias. Such conditions are usually evident on careful physical examination of the back, perineum, and lower extremities.

In girls who are otherwise well and whose history is that of continuous wetting day and night, despite successful toilet training, for a lifelong history, an extravesical infrasphincteric ectopic ureteral orifice should be strongly suspected and imaging should be vigorously pursued.

An ectopic ureter as a cause of wetting is well documented [1]. Ureteral ectopia with incontinence is uniquely female, because the most caudal location for an ectopic ureteral orifice in a male is always above the urethral sphincter [2]. Its diagnosis may be delayed due to inadequate medical history but often may be suspected from detailed medical history and the clinical presentation with a characteristic pattern of wetting, as exemplified in the present case.
As a result, a detailed history, including the pattern of wetting, and a thorough physical examination supplemented with appropriate investigations usually lead to a diagnosis [3]. In the majority of cases the ectopic ureters are derived from the upper moiety of duplex kidneys. Ectopia of the ureteral orifice is often associated with dysplasia of the kidney or of that portion of the kidney drained by the ectopic ureter. As a rule, the more ectopic the orifice the worse will be the dysplasia [4].

The majority of ectopic ureters associated with renal duplication can be diagnosed by clinical history, renal and bladder USG or IVU. USG often shows evidence of a duplicated collecting system with hydronephrosis of the upper pole of the collecting system. A dilated ureter is often visualized posterior to the bladder.

Findings on IVU vary based on amount of renal function present, and range from functioning upper pole moieties associated with hydroureteronephrosis to poorly functioning upper moieties with downward and lateral displacement of the lower pole collecting system (The “drooping lily” sign). However, when the upper pole ectopic ureter is not dilated and the kidney is small, dysplastic, poorly functioning, findings on USG and IVU are often inconclusive. Therefore, additional diagnostic procedures including, renal scintigraphy [5], CT [6, 7] and MR [8, 9] are recommended. In our patient IVU suggested poorly functioning segment at the upper pole of left kidney, USG revealed a cystic mass in the upper pole. To maximize our diagnostic sensitivity we performed CT and detected a left duplex kidney located at left upper pole unit and drained by a dilated ureter extending to the vagina.

In conclusion, girls with continuous wetting should be considered to have an ectopic ureteral orifice until proved otherwise. IVU with CT is indicated to confirm the suspicion and to show the side or sides of involvement. In most cases IVU will be diagnostic. However, when the history is highly suggestive and the urographic findings seem normal, enhanced CT may show the abnormality, which is almost certainly present.

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