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Ectopic Ureters in Siblings

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Abstract : We report a case of single system ectopic ureters that developed in siblings. The elder sister had continuous urinary incontinence after toilet training. She had a left dysplastic kidney and a left ectopic ureter which terminated in the vagina. The younger brother presented with febrile urinary tract infection. A small cystic kidney and an ectopic ureter on the left side were detected incidentally. His left ureteral orifice was found in the posterior urethra. The two children underwent nephroureterectomy and the surgical specimens showed renal dysplasia.

Key Words : Abnormalities, Siblings, Ureter

Introduction

Ectopic ureter is an abnormal termination of the ureter at the bladder neck or distally into the mesonephric duct structures. There have been many studies on the evolutional mechanisms, locations of the ectopic orifice and the correlation with the kidney [1,2]. However, the true incidence of ectopic ureter is unknown. There have been no data reported about the familial or genetic backgrounds of ectopic ureter. To the best of our knowledge, this is the first report of ectopic ureters that developed in siblings.

Case

An elder sister and a younger brother had consulted from the pediatric department. The 3-year-old girl presented with continuous urinary incontinence despite successful toilet training. Continuous urinary dribbling and pooling of urine in the vagina were detected

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on physical examination. On ultrasonography (USG), the right kidney and bladder were normal, but the left kidney was not found in the renal fossa. There was no reflux demonstrated on voiding cystourethrography (VCUG). The technetium-99m dimer-captosuccinic acid (^{99m}Tc-DMSA) renal scan showed a normal right kidney and a minute isotope uptake in the left lower abdominal area (Fig. 1). The computed tomography scan revealed a small left ectopic kidney in the para-aortic area at the level of bifurcation of the inferior vena cava, the left ureter entered the vaginal wall and contrast media was seen in the vagina (Fig. 2). The left hemitrigone and ureteral orifice were not detected on cystourethroscopy. Left nephroureterectomy was performed and the girl was free from symptoms immediately after the procedure. Pathological finding as renal dysplasia with chronic inflammation.

The 7-month-old boy with febrile urinary tract infection (UTI) was also transferred from the pediatric department. With a normal renal parenchyma on the right side, the USG showed a cystic lesion with thin parenchyma on the left side, and a dilated left ureter behind a normal bladder. VCUG demonstrated reflux into the left ureter, which originated from the posterior urethra (Fig. 3). The ^{99m}Tc-DMSA scan revealed no radioactivity in the left kidney. Cystourethroscopy showed an ectopic ureteral orifice in the prostatic urethra and the absence of the ureteral orifice and hemitrigone on the left side. The boy underwent left nephroureterectomy. The surgical specimen showed severe renal dysplasia.

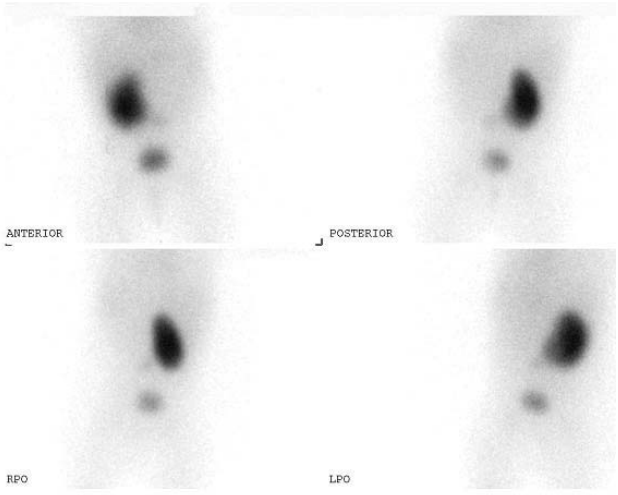

Fig. 1. Technetium-99m dimercaptosuccinic acid renal scan shows a normal right kidney and a minute isotope uptake in the left lower abdominal area. RPO, Right posterior oblique; LPO, Left posterior oblique

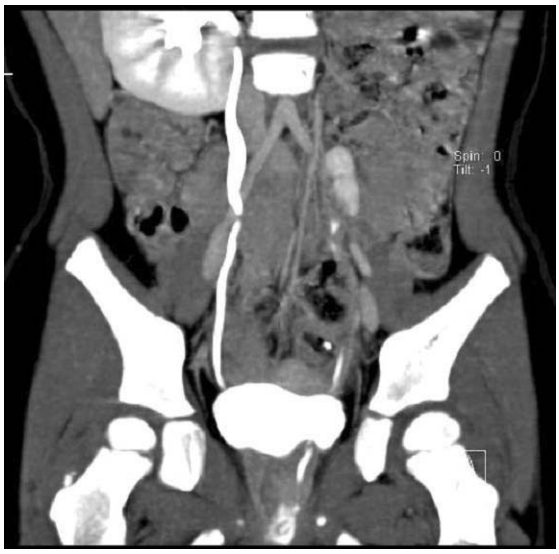

Fig. 2. A computed tomography scan reveals 25 mm-sized left ectopic kidney in the para-aortic area at the level of bifurcation of the inferior vena cava. The left ureter entered the vaginal wall and contrast media was seen in the vagina.

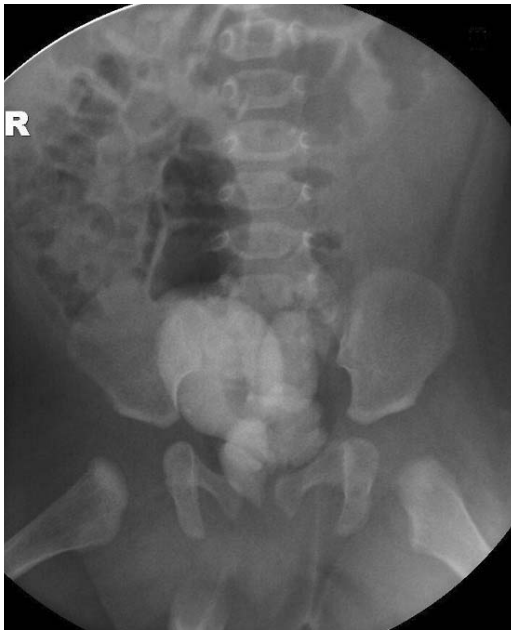

Fig. 3. A voiding cystourethrograph shows reflux into the dilated left ureter originated from the posterior urethra.

Discussion

The incidence of ectopic ureter is unknown because many patients with this condition do not have symptoms.

It has been generally accepted that single system ectopic ureters occur less frequently than ectopic ureters that are associated with a duplicated collecting system. Recently, Chowdhary *et al.* [2] reported that 11 of 127 (8.7%) ectopic ureters arose from a single system. However, the reported incidences of single system ectopic ureter were less than 25% [3,4].

In females, ectopic ureters are commonly associated with a duplicated collecting system, whereas single system ectopic ureters occur more frequently in males[5,6]. In Western countries, most ectopic ureters are associated with a duplicated collecting system, and it has been thought that single system ectopic ureters are rare. In east Asia, However, single system ectopic ureters have been more commonly reported [6].

The clinical manifestations of ectopic ureter are quite different between males and females, due to the locations of ectopic ureteral orifice. The sites of the ectopic ureteral orifices were posterior urethra, prostatic utricle, seminal vesicle, ejaculatory duct and vas deferens in males, but vestibule, urethra, vagina, cervix, uterus and Gartner's duct in females. Males with ectopic ureter do not present with urinary incontinence, but often presented with UTI because their ureteral orifices more commonly open into the prostatic urethra and bladder neck. In females, the urethra, vestibule and vagina are common sites resulting in urinary incontinence or persistent vaginal discharge [6].

Ectopic ureters are often associated with renal dysplasia. An evolutional mechanism of renal damage associated with ectopic ureter might be maldeveloped embryogenesis. Unilateral renal absence or a hypofunctioning kidney, and simultaneous absence of the ipsilateral ureteral orifice and hemitrigone reflect renal agenesis or dysplasia [1]. An abnormally positioned ureteric bud had poor interaction with the metanephric blastema and this mismatch led to renal maldevelopment, resulting in renal dysplasia. The degree of abnormality of the ureteric bud location is correlated with the extent of renal dysplasia [6]. Furthermore, the affected kidney is frequently located in an ectopic position [7].

Treatment of ectopic ureter depends on the functioning of the ipsilateral and

contralateral kidneys. Ureteral reimplantation is indicated for ureters with well-functioning kidneys. Nephroureterectomy is recommended when the ipsilateral kidney is dysplastic or hypofunctioning at less than 10% [8]. In this report, two children underwent nephroureterectomy due to a hypofunctioning kidney.

As in our case, Park *et al.* [9] reported complete dryness after nephrectomy for the patients with single system ectopic ureter with ipsilateral dysplastic kidney. Chowdhary *et al* [2], However, reported that some patients had persistent residual wetting after the surgery. This persistent dampness might be related to a weak urethral sphincter, which was caused by a maldeveloped ipsilateral trigone [10].

In parents and siblings of probands with duplication, the incidence of duplication increases from the predicted 1 in 125 to 1 in 8 [11] or 1 in 9 [12]. The incidence of reflux in the siblings of patients with reflux is greater than the rate in the general population at approximately 32% [13]. However, the prevalence may be as low as 7% in older siblings [14] or as high as 100% in identical twin siblings [15]. However we report a unique presentation of ectopic ureter in sibling. This is the first report in the literature in English of single ectopic ureter developing in siblings. Further studies are necessary to determine the familial or genetic background of ectopic ureters.

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