

A rare case of left kidney and ureter duplication with ectopic orifice in vagina. Diagnosis, complications, treatment

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KEY WORDS

kidney ▶ urinary tract developmental defects ▶
enuresis ureterica ▶ ectopic ureter orifice
▶ heminephroureterectomy

ABSTRACT

The objective of the study was to present a rare case of a double pyelo-calyceal system and duplicated ureter leaving the left kidney diagnosed in a female patient. The ureter was descending from the upper segment of the left kidney with an ectopic orifice into the vagina. The condition resulted in enuresis ureterica. We present diagnostic and therapeutic processes that often pose difficulties for many specialists, including urologists, gynecologists, surgeons, radiologists and pediatricians.

INTRODUCTION

Ectopic ureter orifice is a rare congenital defect of the urinary tract resulting from abnormal embryonic development. Upper urinary tract duplication is a consequence of two ureteric buds or a bifurcated single bud formed soon after it bulged into Wolff's duct [1, 2]. The displaced ureter orifice can be located in the urinary bladder or outside it. Outside the bladder ureter orifice is observed in 1 per 20,000 births, five times more frequent in females than males and in 75% cases it is associated with a duplicated ureter [3]. According to Weigert-Meyer law, outside the bladder ectopic ureter orifice observed in females refers to the upper ureter descending from the upper pole of the kidney (duplicated in many cases). Typically, it produces involuntary, continuous leakage of urine from a ureter that is displaced beyond the external sphincter. The condition is called enuresis ureterica and is associated with difficult outflow of urine from the kidney segment containing the ectopic ureter. In males the ectopic ureter can open into the epididymis causing inflammations [4] and frequent urinary tract infections. The case report presents a patient with ectopic vaginal orifice of a double ureter, diagnosed and treated for incontinence and pseudo-vesicovaginal fistula.

CASE REPORT

The patient, a 33-year old female was admitted to the Clinic of Urology and Urological Oncology, at the Medical University of Lublin on May 11th, 2008 for septic fever with pyonephrosis of the left kidney's upper segment. The patient was in a severe condition. The patient's history revealed that she had suffered incontinence

and lumbar pains since her kindergarten years, but was never treated. She was pregnant 3 times. The first pregnancy progressed normally with spontaneous delivery in due term but, the second pregnancy (2003) was terminated by spontaneous miscarriage. She carried her third pregnancy (2004) until spontaneous full-term delivery of a healthy baby. However, during her third pregnancy she suffered from frequent infections of the vagina and urinary bladder and incompetence of the uterine cervix, which required a surgical procedure. Towards the end of her pregnancy she developed acute pyelonephritis.

Urographic examination performed in the Urological Outpatient Department in May 1998 detected a widened pyelo-calyceal system in the right kidney and incomplete turning of the left kidney. Ultrasound of her abdomen detected a bilaterally widened pyelo-calyceal system. Micturating cystography found no abnormalities. The patient was released with the diagnosis of right ureteropelvic junction stenosis with a recommendation for further diagnosis and treatment.

Follow-up ultrasound examination of the abdomen, performed in September 2001, showed two kidneys of normal size. The right kidney presented normal without features of retention; the left kidney's pyelo-calyceal system was slightly distended, suggestive of



Fig. 1. Intravenous pyelography. No leakage of contrasted urine in the upper segment of left kidney.

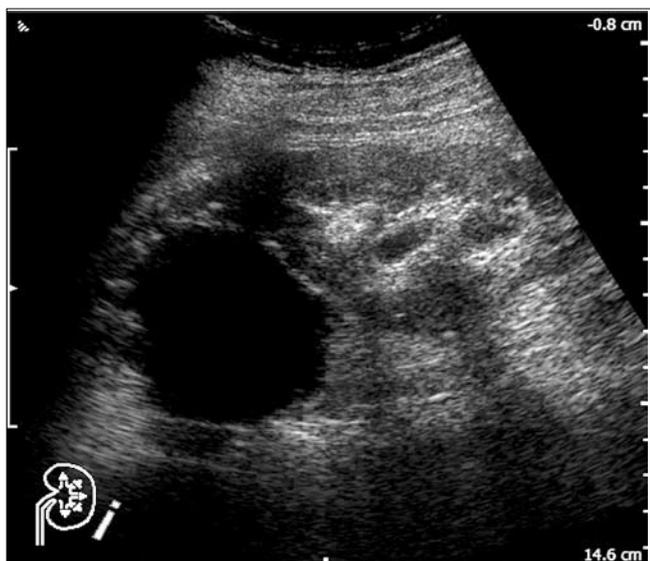


Fig. 2. USG of left kidney. Hydronephrotically changed upper segment of the kidney that has been mistaken for a cyst. The change pulls the lower segment affected by inflammation away from the spine.



Fig. 4. Close to the bladder, the part of the ureter descending from upper segment of the left kidney. Ectopic orifice into the vagina.

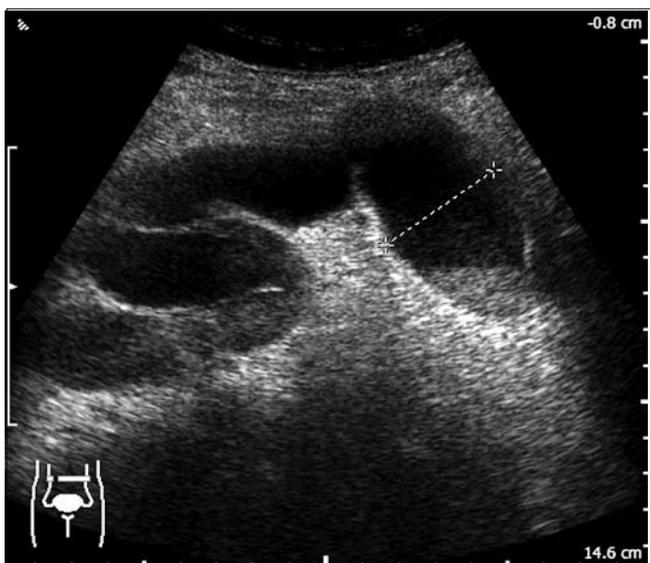


Fig. 3. Middle and lower segment of substantially widened (up to 36 mm) duplicated ureter filled with thick contents. Harmonic option.



Fig. 5. Descending pyelography. Contrasted upper segment of the left kidney. Visible nephrostomy.

obstructed urine outflow, possibly vesicoureteral reflux. Uroflowmetry did not find any abnormalities.

In 2006 in the Gynecological Ward, the tension-free vaginal tape (TVT) surgery was performed. In February 2008 the patient was admitted to the Clinic of Surgical Gynecology. She underwent surgery to remove the vesico-vaginal fistula. She was discharged from the hospital in good general condition. Incontinence subsided. After 5 days she developed a high fever and tenderness in the lumbar area. Pyuria and incontinence recurred soon. In May 2008 she returned to the Clinic of Surgical Gynecology and underwent the reoperation of the vesico-vaginal fistula. On the 6th day after the reoperation she developed high fever with tremor and vomiting. Physical examination found positive Goldflam's sign on the left side. Basing on the clinical examination, she was diagnosed with pyonephrosis of the left kidney with urosepsis. Emergency admission was necessary and the patient was hospitalized at the Clinic of Urology and Urological Oncology. Intravenous pyelography showed bilateral concomitant excretion of urine of the same concentration by both kidneys. The left kidney was incompletely turned and

a widened pyelo-calyceal system in the left kidney with an altered lower anterior calyx. The renal pelvis was slightly widened on the right side when compared to the left one and a bent stenotic subpyelar section of the right ureter without evidence of retention in the pyelo-calyceal system (Fig. 1). Ultrasound of the abdomen showed a hydronephrotically widened double pyelo-calyceal system in the left kidney with a widened total length of the ureter of 22 mm in the upper part and up to 38 mm near the bladder (Figs. 2, 3 and 4). Emergency nephrostomy of the left kidney's upper segment drained thick pus. Selective antibiotic treatment was introduced producing improvement in the patient's general condition while returning temperature to normal. A descending pyelography visualized a widened renal pelvis and a widened total length of the ureter – ureterohydronephrosis (Fig. 5). The ureter of the lower renal segment was normal and opened into the bladder, however, the ureter descending from the upper segment opened ectopically into the vaginal vestibule. Nephrostomy was performed and the patient was released home with selective recommendations to wait for an elective urological operation.

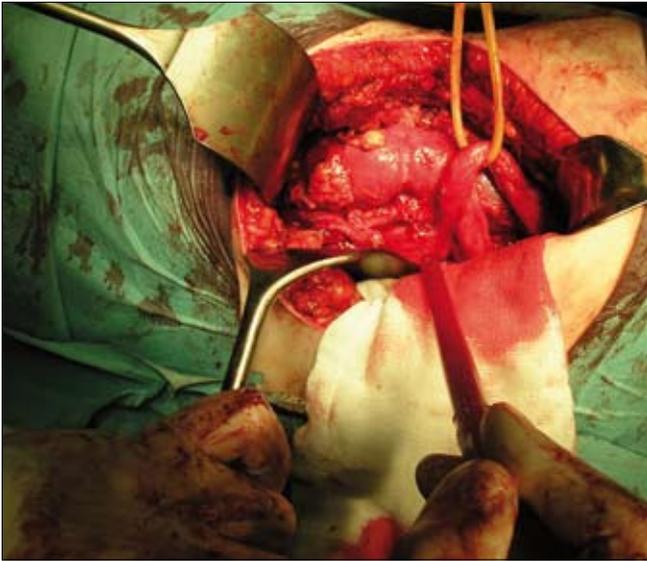


Fig. 6. Left heminephrectomy. Visible widened ureter of the upper segment of the left kidney.

After 3 weeks the patient underwent left side heminephrectomy which was performed in the Department of Urology and Urological Oncology (Fig. 6). The upper segment of the left kidney was removed. A 3cm-wide ureter was submaximally dissected towards the bottom and excised. The contents were evacuated and the ureter stump was left to obliteration. Afterwards, the patient was discharged home in good condition.

In October 2008 the patient was admitted to the Clinic of Urology and Urological Oncology again. She complained of hypogastric pain and episodes of high temperature. Ultrasound and intravenous pyelography (Fig. 7) showed hydroma of the ureteral stump, which developed following left heminephrectomy. It was excised. The postoperative course was uneventful, the wound healed by primary adhesion. The patient was followed up without any complaints.

DISCUSSION

Congenital urinary tract defects in adults cause many diagnostic difficulties. Ectopic ureter orifice, which produces enuresis ureterica is a relatively rare congenital defect, detected and treated mainly in children [5]. Its diagnosis is based on detailed clinical history and imaging examinations. The defect may be signaled by disordered urination and continuously wet diapers and underwear in older children. This is the main reason that causes parents to bring them to the doctor [6]. The absence of continuous leakage of urine does not exclude the defect since the "upper" kidney is likely to be vestigial and produce small amounts of urine. Intravenous pyelography is a basic imaging technique, especially its late pictures that visualize deficient excretory function of the segment with the ectopic ureter. It is difficult to interpret the urographic picture as the "upper" segment minimally changes the direction of the long axis of the kidney, hence detecting the defect based on that examination is not easy, even for an experienced urologists. Other useful imaging techniques are USG and or a nuclear scan with ^{99m}Tc-technetium [7]. If the defect is undetected in early childhood, its diagnosis is even more difficult in adulthood. In adult women, especially after delivery, urinary incontinence should be differentiated from incontinence developed due to functional-anatomical disorders or other diseases such as genitourinary fistulas. Enuresis ureterica may recur after delivery due to mechanical removal of an obstruction from the ureter's orifice into the vagina [8]. Detecting vaginal ectopic ureter orifice by physical examination becomes difficult as



Fig. 7. Intravenous pyelography prior to surgical removal of ectopic ureter stump. Normal excretion of contrasted urine by both kidneys. Visible wide ectopic ureter stump with retained contrast administered during descending pyelography prior to heminephrectomy.

it can hide between folds of mucous membranes, which make it invisible. Urine flowing out of the additional ureter is often uncondensed and the patient does not feel any discomfort, thus they would seek medical consultation much later. In this case report our patient "accepted" disordered urination. She did not seek medical consultation until her complaints increased during pregnancy and after the delivery. Misdiagnosed incontinence and the resulting surgery produced further complications. The patient's urethra was suspended by the TVT technique so iatrogenic vesico-vaginal fistula formation was assumed and ureter orifice into the vagina was sutured as it was mistakenly interpreted as a fistula. That resulted in additional complications which led to heminephrourectomy. During the first operation performed in the Clinic of Urology and Urological Oncology, the doctors tried to spare the young woman additional postoperative scars for aesthetic reasons and left the ureter stump. Unfortunately, reoperation was unavoidable.

The case presented in this report is a very rare congenital defect. Moreover, an array of complications developed as a result of primary misdiagnosis – that is why we decided to report this case.

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