

CASE REPORT

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Duplex System with Ectopic Ureter Opens into the Posterior Urethra: Case Report

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ABSTRACT

Introduction: Duplicated ureter or Duplex Collecting System is a congenital condition in which the ureteric bud, the embryological origin of the ureter, arises twice, resulting in two ureters draining a single kidney. This congenital anomaly is rare, and even rarer when the duplex system with ectopic ureter is present. This type of congenital anomaly is even more rarely diagnosed and surgically treated in adulthood. **Case report:** This case report presents a case of a 32-year-old male, who had a duplex collecting system with two ureters on the left side. Ectopic ureter, draining the upper pole of the left kidney, opened into the posterior urethra. In our patient, taking into account the clinical perspective, the renal tissue damaging of the upper pole which was not functional, partial nephrectomy and ureterectomy was successfully performed.

Keywords: duplex system, ectopic ureter, nephron-sparing surgery.

1. INTRODUCTION

The kidneys are paired retroperitoneal organs that are normally located between the transverse processes of T12-L3 vertebrae, with the left kidney typically somewhat more superior in position than the right. Duplicated ureter or Duplex Collecting System is a congenital condition in which the ureteric bud, the embryological origin of the ureter, arises twice, resulting in two ureters draining a single kidney. It is the most common renal anomaly, occurring in approximately 1% of the population (1, 2). Ureteral development begins in the human fetus around the 4th week of embryonic development. A ureteric bud, arising from the mesonephric (or Wolffian) duct, gives rise to the ureter, as well as other parts of the collecting system. In the case of a duplicated ureter, the ureteric bud either splits or arises twice. In most cases, the kidney is divided into two parts, an upper and lower lobe, with some overlap due to intermingling of collecting tubules. However, in some cases the division is so complete as to give rise to two separate parts, each with its own renal pelvis and ureter. Interestingly, and explaining Weigert-Meyer rule, the future lower pole ureter separates from Wolffian duct earlier and thus migrates superiorly and laterally as the urogenital sinus grows. According to this Wei-

gert-Meyer law, ureter draining the lower pole should open more cranially, and become refluxive. Presence of various anomalies of the ureter is associated with increased risk of urinary tract infections and many other clinical complications (3).

2. CASE REPORT

A 32-year-old male was hospitalized in the parent institution in November 2016, due to general symptoms of infection accompanied by pain in the left lumbar region, when duplex collecting system of the left kidney was diagnosed. CT urography, performed during the hospitalization indicated the existence of a duplex pyelocaliceal system of the left kidney with a duplicated ureter, dilation of the pyelocaliceal system of the upper pole of the left kidney grade 4, with the kidney parenchyma reduced to 1 mm, as well as the left ureter dilated (12 mm) and curved throughout its length, with ectopic ostium in prostatic urethra (Figure 1 and 2).

During the hospitalization, the patient was treated conservatively with a positive clinical effect. After the inflammatory parameters were stabilized and the pathogenic microorganism was eliminated from urine, the prophylactic antimicrobial therapy ensued, supplemented by further diagnostic procedures.

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Figure 1. Dilatation of the pyelocaliceal system of the upper pole of the left kidney grade 4



Figure 3. Dilatation of the pyelocaliceal system of the upper pole of the left kidney grade 4 by ultrasound

Renal radionuclide imaging, as an integral part of nuclear medicine, provides substantial information on the actual renal function. Static kidney scintigraphy with Tc-99m (DMSA) indicated the absence of accumulation of radiopharmaceuticals in the upper third of the left kidney, except for the slightly preserved function of cortical activity in the marginal area of the upper pole of the left kidney. Dynamic scintigraphy was performed with Tc-99m (DTPA) and it indicated a lower amplitude of the renal curve above the left kidney (lower functional mass), with a relative left-kidney function of 35.3%, while the relative right-kidney function was 64.7%. The patient was surgically treated at the Urology Clinic of the University Clinical Centre of the Republic of Srpska in Banja Luka, in July 2017. Control Ultrasound examination indicated the existence of a duplex pyelocaliceal system of the left kidney with a duplicated ureter, dilation of the pyelocaliceal system of the upper pole of the left kidney grade 4, with the kidney parenchyma reduced to 1 mm, as well as the dilatation of the left ureter (Figure 3). Taking into account the clinical perspective, the degree of damage to the renal parenchyma of the upper pole of the left kidney which was not functional, the indication for nephron-sparing surgery was set, that is, for the partial nephrectomy and ureterectomy. Optimal renal perfusion provided by a hydration regimen of approximately 200 mL/h or more of crystalloids overnight was benefi-

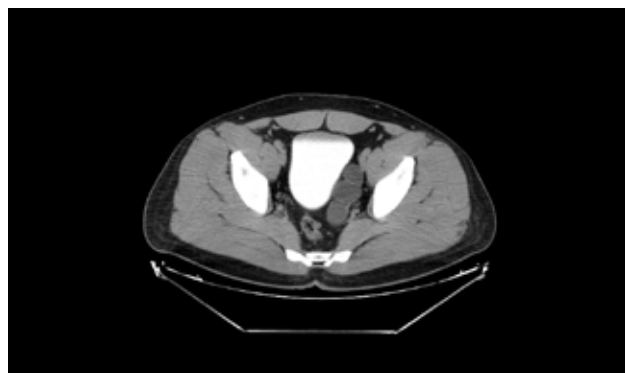


Figure 2. Ectopic ostium of the duplicated left ureter in prostatic urethra

cial. Before positioning the patient for lumbar incision, an urethrocytoscopy was performed, detecting no ectopic ureter orifice in prostatic urethra. Afterwards, JJ stent was placed through the orthotropic left orifice, in order to accurately preserve the ureter for the distal two-thirds of the left ureter. After the patient was adequately positioned, a standard left side lumbar approach was performed, which provided excellent and rapid exposure to the kidney and the hilum. Optimal renal exposure is the key to a successful outcome. We identified the renal pedicle and defined the vasculature and ureter. Then, we isolated the renal artery and placed a vascular loop. After that, we excised the hydronephrotic upper pole with sharp dissection and put suture on the edge to ligate any bleeding vessel with 3-0 absorbable sutures with in situ techniques. Then, the ureter was released with blunt and sharp dissection to the distal third of it, as far as possible from the lumbar incision, the ureter was cut and preparations of the resected upper pole of the left kidney were removed continuously from the proximal 2/3 of the ureter. After that, the aspiration of the contents of the un-resected distal third of dilated left ureter was performed and ligature was placed on it. The resected surface of the upper pole, approximately max 20x20 mm, was additionally cauterized with argon, covered with the Gerota's fascia, the drain was placed, and the lumbar incision was closed by layers. The patient had a neat post-surgery course. PH findings indicated that there was a renal parenchymal dysfunction, nephritis interstitial chronica and ureteritic chronica. A control ultrasound performed after six months showed a regular post-surgery finding for the remaining distal two thirds of the left kidney, without the accumulation of fluid in the retroperitoneal space.

3. DISCUSSION

The Duplex collecting systems diagnosis is rare in adulthood, as most such anomalies are detected and surgically corrected in childhood. Generally, duplex collecting system can be variable. At one end of the spectrum, there is merely a duplication of the renal pelvis, draining via a single ureter. At the other extreme, two separate collecting systems drain independently into the bladder or ectopically. In men, ectopic ureter commonly opens into the posterior urethra, and these men remain continent. However, ectopic ureters opening into the genital tract can manifest symptoms as epididymitis, vesiculitis,



Figure 4. Resected duplicated collecting system of the left kidney

prostatitis, bloody, and painful ejaculation. Ectopic ureteral opening is more often associated with single collecting system in men, and in women it is more frequently associated with double collecting system (4). The present study reports the case of a 32-year-old male, who had duplex collecting system with two ureters on the left side. Ectopic ureter, draining the upper pole of the left kidney, opened into the posterior urethra. Most duplicated systems are asymptomatic and diagnosed incidentally. However, where symptoms do occur (infection, reflux or obstruction), the patient is likely to have completely duplicated ureters. The patient from our case report was treated in hospital due to general symptoms of infection accompanied by pain in the left loin, when he was diagnosed with duplex collecting systems et ureter duplex. As the abnormality is an anatomic alteration, all modalities able to image the renal tract may be able to visualize the typical features. Ultrasound provides excellent anatomic information but does not necessarily differentiate a bifid renal pelvis from a bifid ureter or two complete ureters. In our case, ultrasound provided exact diagnosis of duplex collecting systems and ureter duplex, but did not describe exactly where the ostium of the ureter of upper pole was. CT scanning can help to determine if an obstruction exists and can aid in assessing the renal parenchyma. CT scanning can also help to determine if the insertion of the duplex ureter is intravesical or extravesical (5). Using 16-multidetector computed tomography (16-MDCT) scanning on 126 consecutive potential kidney donors, Raman and colleagues described and quantified renal anatomic variants and looked at the frequency of various findings associated with the renal parenchyma and with the collecting system (6). In our case report, CT scan confirmed the duplex collecting system and ureter duplex diagnosis, showing that the ectopic ostium of the ureter for the upper pole of the left kidney is in the prostatic urethra. The presence of a duplex kidney and ureteral duplication, suggested by findings from excretory urographic, ultrasonographic or CT images, can

be confirmed with scintigraphy. The following radionuclides are used for dynamic imaging: Tc-99m-diethylene triamine pentaacetic acid (DTPA) (Pentatate; 492 Da) and Tc-99m-MAG3 (Mertiatide; 350 Da). Tc-99m-dimercaptosuccinic acid (DMSA) (Succimer; 281 Da) is used for static imaging. The use of dimethyl succinic acid (DMSA) scanning to assess parenchymal function in a duplex kidney is of great value in the management of duplex kidney (7). In our case report, static scintigraphy of the kidney with Tc99m (DMSA) was performed in the patient, indicating the absence of accumulation of radiopharmaceuticals in the upper third of the left kidney, except for the slightly preserved function of cortical activity in the marginal area of the upper pole of the left kidney. Dynamic scintigraphy was performed with Tc99m (DTPA) and it indicated a lower amplitude of the renal curve above the left kidney (lower functional mass), with a relative left-kidney function of 35.3%, while the relative right-kidney function was 64.7%.

Current approach in the management of pyelon et ureter duplex in this case is to remove anomalous upper pole of kidney together with ureter draining into prostatic urethra using nephron-sparing surgery (8, 9). In our patient, taking into account the clinical perspective, the renal tissue damaging of the upper pole which was not functional, partial nephrectomy and ureterectomy was successfully performed

4. CONCLUSION

This case report presents an excellent surgical treatment of the congenital anomaly duplex system with ectopic ureter opening into the posterior urethra in adulthood. Having in mind the clinical perspective and degree of damage to the parenchyma of the upper pole of the left kidney, partial nephrectomy with ureterectomy was an optional treatment method. However, if there had been regular ultrasound examinations during childhood, this innate anomaly would have been surgically corrected in childhood, thus preserving the renal parenchyma and the renal function of the upper pole of the left ureter.

- **Conflict of interest:** none declared.

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