CASE REPORT

URINARY TRACT INFECTIONS

The remnant of a congenital, blind- ended megaureter in a 23-year-old woman causing chronic pain and urinary infections

Tomislav Pejcic[1], Biljana Markovic[2], Zoran Dzamic[1], Milan Radovanovic[1], Jovan Hadzi-Djokic[3]

[1]Clinical Center of Serbia, Urological Clinic, Belgrade, Serbia
[2]Clinical Center of Serbia, Institute of Radiology, Belgrade, Serbia
[3]Serbian Academy of Sciences and Arts, Belgrade, Serbia

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Correspondence
Tomislav Pejcic
129/9, Bulevar Zorana Djindjica
11070 Belgrade, Serbia
phone: +38 111 212 1616
tomislav_pejcic@vektor.net

Multicystic dysplastic kidney (MCDK) is a congenital anomaly as the result of abnormal interaction between the ureteric bud and metanephric mesenchyme. Unilateral MCDK can be associated with other anomalies of the genitourinary tract. Relatively rare associated anomaly is the presence of ipsilateral refluxing blind megaureter.
The patient reported herein is a 23–years–old woman with involuted MCDK and ipsilateral blind megaureter causing chronic urinary infection and chronic abdominal pain. Preoperative and intraoperative examination failed to detect the communication between megaureter and the urinary bladder.

Key Words: multicystic dysplastic kidney ‹› refluxing blind megaureter

INTRODUCTION

Multicystic dysplastic kidney (MCDK) is a congenital anomaly that is the result of abnormal interaction between the ureteric bud and metanephric mesenchyme, early ureteral obstruction, or ureteral atresia. MCDK is usually presented as the formation consisted of multiple cysts of varying sizes, without normal renal parenchyma. The significant number of MCDKs involute and disappear during the fetal period and the early childhood.
Unilateral MCDK can be associated with other anomalies of the genitourinary tract. A relatively rare associated anomaly is the presence of ipsilateral refluxing blind megaureter, which is the result of abnormal development of the vesicoureteral junction; it is usually presented as elongated tubular pseudo-intestinal cystic structure [1]. Refluxing blind megaureter can be associated with chronic urinary infection and chronic abdominal pain. However, blind megaureter rarely requires extirpation in the adult age.

CASE REPORT

A 23–year–old woman from a small village was sent to the urologist from the gynecologist, due to solitary right kidney, cystic mass on the left side of the urinary bladder and the presence of chronic pain and urinary infection. Gynecological exam was normal.
Girl's mother stated that fetal ultrasonography (USG) in 9th gestational month revealed a cystic left kidney. At the age of two, abdominal USG revealed 22 mm large cystic formation in the position of the left kidney. Intravenous urography (IVU) and renal scintigraphy revealed normal right kidney and the absence of the left kidney.
During the next years, the girl was admitted several times to regional hospital, due to recurrent urinary infections and febrile episodes. At the age of 14, cystography revealed vesicoureteral reflux (VUR) grade II on the right side and VUR grade I in the blind megaureter. After that, there were no more urinary infections and VUR spontaneously disappeared on the both sides. The young lady refrained from medical controls until she was married.

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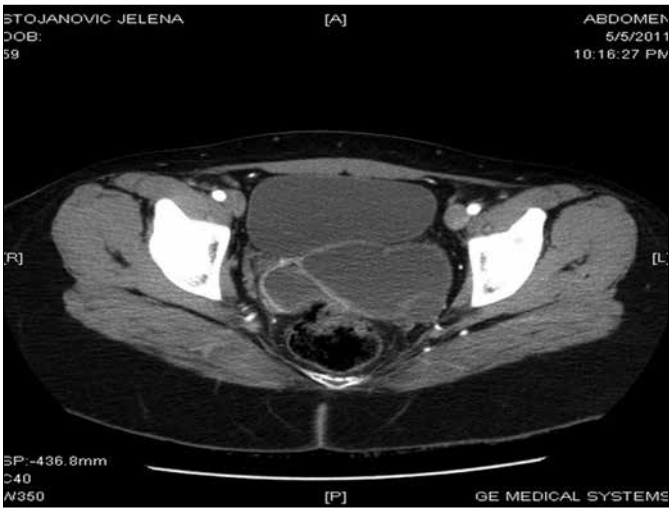

Figure 1. *CT showing pseudocystic formation on the left and beneath the urinary bladder.*

At the age of 23, she visited gynecologist, due to chronic pelvic pain and inability to stay pregnant. The gynecological examination revealed bizarre cystic mass in the pelvis, caudal and left to the urinary bladder. A computerized tomography (CT) scan revealed the presence of the 15-cm-long pseudo-cystic mass in the pelvis. The left kidney was absent while the right kidney was hypertrophic (Figure 1).

At the admission on urology, the young woman was of short stature and morbidly obese. Complete blood count and blood analyses were normal. Cystoscopy revealed clear urine, normal bladder capacity, symmetric trigone, normal right orifice, while the left orifice could not be identified. Transvaginal ultrasonography revealed the presence of 15 cm long gut-like pseudocystic formation on the left side and beneath the urinary bladder.

The patient underwent ureterectomy of the blind megaureter, through the left oblique pelvic incision. The ureter was 24 cm long and 4 cm wide, with the 5-7 mm thick wall. The urine inside the ureter was fetid and cloudy. Pathological diagnosis revealed megaureter and fibroproductive chronic ureteritis (Figure 2).

Postoperative period was normal and the patient discharged in satisfactory general condition. Unfortunately, one year after the surgery, she developed hyperthyreosis, complicated with exophthalmos. At the last visit, her hyperthyreosis was under control.

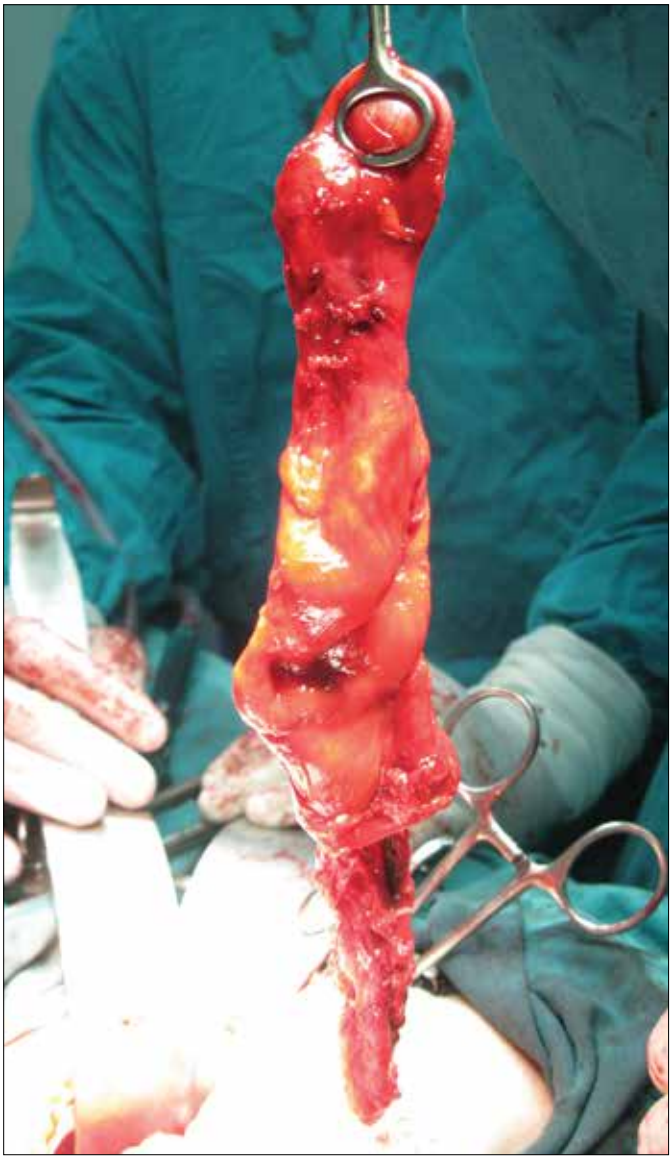

Figure 2. *Removed blind-ended megaureter.*

DISCUSSION

The explanation of the association of MCDK and the primary megaureter (PM) is that both anomalies are the consequence of the abnormal development of the ureter; both anomalies are more often on the left side and boys are more commonly affected. Generally, PM is the result of abnormal development of the vesicoureteral junction; distal ureteral segment can be aperistaltic and fibrotic (obstructed PM), or short, with decreased amount of smooth muscle (refluxing PM). In 10-15% of cases of PM, contralateral kidney is absent or dysplastic [2].

In about 60% of the cases of the unilateral MCDK, the ipsilateral ureter is completely absent. If present, the ipsilateral ureter is always abnormally developed [3]. The most commonly associated anomaly is contralateral VUR, in almost 20%, while the incidence of ipsilateral VUR is relatively low, about 4% [4, 5]. MCDK can be associated with the malformations of the vagi-

na, uterine horn, fallopian tube or ovary in female, or with the absence of the ipsilateral testis, vas deferens, or seminiferous tubules in male [6, 7]. In 93 patients with MCDK, Merrot found six cases with vanishing MCDK and cystic masses close to the urinary bladder. In addition, he found that 15% of patients had malformations of the ipsilateral genitalia [8].

In a child with involuted MCDK, VUR into the blind megaureter may cause abdominal or pelvic pain and chronic urinary infection. Abdominal ultrasonography (US) usually shows multiple cysts without renal parenchyma, or the absence of the kidney. Voiding cystourethrography is necessary to prove, or rule out VUR. Computerized tomography urography (CTU) or magnetic resonance urography (MRU), are sometimes necessary to confirm the diagnosis and to estimate the status of ipsilateral kidney.

The treatment of MDCK is usually conservative, as the majority of MCDKs spontaneously disappear during childhood. However, these conditions sometimes require conservative treatment, due to urinary infection; nephrectomy is indicated very rarely. The treatment of PM is indicated in the cases of renal involvement, chronic urinary infection and calculosis, while in adult age PM is usually asymptomatic and requires only conservative treatment [9, 10].

In the presented case, left MCDK was discovered prenatally and the ipsilateral blind refluxing PM, at the age of two. The girl was followed-up due to recurrent urinary infections, but later, she was asymptomatic and no medical controls. The last cystography failed to detect VUR in the right ureter and the left blind ureter as well.

It is interesting that with time, massive blind megaureter become completely occluded at the level of orifice. We can speculate that VUR in the megaureter existed for a long time, which caused its marked dilatation and tortuosity. It is likely that chronic urinary infection and fibrosis lead to gradual narrowing of the refluxing orifice and, at last, to its closure. Later on, large pseudocystic formation became the cause of chronic abdominal and pelvic pain and the chronic urinary infection.

The patients with involuted MCDK and ipsilateral blind megaureter should have long term follow-up due to possible complications. Complete resection of blind megaureter should be performed in cases of chronic urinary infection and chronic abdominal or pelvic pain.

References

1. La Fianza A, Preda L, Di Maggio EM, Campani R. Blind megaureter with ipsilateral renal agenesis and mullerian anomaly: MR findings in a case. Clin Imaging. 1999; 23: 184-186.
2. Hodges SJ, Werle D, McLorie G, Atala A. Megaureter. Scientific World Journal. 2010; 10: 603-612.
3. Shapiro E, Bauer SB, Chow JS. Anomalies of the Upper Urinary Tract. In Wein AJ, Kavoussi LR, Partin AW, Peters CA and Novick AC, eds. Campbell- Walsh Urology, 10th ed. Philadelphia: Saunders, 2012. Section 17, Chapt 117, pp. 3123- 3160
4. Schreuder MF, Westland R, van Wijk JA. Unilateral multicystic dysplastic kidney: a meta-analysis of observational studies on the incidence, associated urinary tract malformations and the contralateral kidney. Nephrol Dial Transplant. 2009; 24: 1810-1818.
5. Singh JK, Kanojia RP, Narasimhan KL. Multicystic dysplastic kidney in children – a need for conservative and long term approach. Indian J Pediatr. 2009; 76: 809-812.
6. Pavlović S, Kostić M, Milanović D, Popović-Rolović M, Basta-Jovanović G, Jovanović O, Peco-Antić A, Kruscić D. Cystic tumefaction in the minor pelvis--the agenesis-dysplasia syndrome (case report). Srp Arh Celok Lek. 1996; 124 (Suppl 1): 242-243.
7. Pope JC IV: Section 17: Pediatric urology. Chapter 118: Renal Dysgenesis and Cystic Disease of the Kidney. In: Campbell- Walsh Urology, 10th ed. Editor-in-chief: Alan J. Wein. Eds: Louis R. Kavoussi, Alan W. Partin, Craig A. Peters and Andrew C. Novick. Philadelphia: Saunders; 2012: 3162-3196.
8. Merrot T, Lumenta DB, Tercier S, Morisson-Lacombes G, Guys JM, Alessandrini P. Multicystic dysplastic kidney with ipsilateral abnormalities of genitourinary tract: experience in children. Urology. 2006; 3: 603-607.
9. Berrocal T, López-Pereira P, Arjonilla A, Gutiérrez J. Anomalies of he distal ureter, bladder, and urethra in children: embryologic, radiologic, and pathologic features. Radiographics. 2002; 22: 1139-1164.
10. Rathi V. A blind-ending ureter with infection due to vesicoureteric reflux with associated renal agenesis: A rare cause of pain abdomen. Urol Ann. 2011; 3: 100-102.