

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr



Case Report

The importance of CT Urography in early diagnosis of anatomical variations in urogenital tract: case presentation[☆]

Bardha Dervishi, MD^a, Fjolla Hyseni, MD, PhDc^b, Juna Musa, MD^c, Kristi Saliaj, MD^d, Valon Vokshi, MD^e, Loran Rakovica, MD^f, Fareeha Nasiri, MD^g, Arlind Decka, MD^h, Eram Ahsan, MDⁱ, Ineida Boshnjaku, MD^j, Essa Mohamed, MD^k, Rilind Sylaj, MD^{l,m}, Dijon Musliu, MD^{l,m}, Krenare Shabani, MD^{l,m}, Arif Musli, MDⁿ, Guri Hyseni, MD^o, Zana Gafurri, MD, PHD^{p,q,*}

^a Faculty of Medicine, University of Prishtina, Kosovo, Albania

^bNYU Langone Health, New York, USA

^c Department of General Surgery, Mayo Clinic, Rochester, MN, USA

^d Mother Teresa Hospital, Tirana, Albania

^e Department of Anesthesiology, University Clinical Center of Kosovo, Kosovo, Albania

^fInternal Medicine, St. Barnabas Hospital, Department of Medicine, New York, USA

^gHarlem Hospital Center Manhattan, New York, USA

^h Department of General Surgery, Westchester Medical Center, Valhalla, NY, USA

ⁱDivision of Nephrology and Hypertension, Mayo Clinic, Rochester, MN, USA

^jHospital Mother Teresa, Tirana, Albania

^k Department of Cardiovascular Desiease, Mayo Clinic, Rochester, MN, USA

¹Clinic of Radiology, University Clinical Center of Kosovo, Kosovo, Albania

^m Faculty of Medicine, University of Prishtina, Kosovo, Albania

ⁿDepartment Radiology, Clinical Center of Tetovo, North Macedonia

° Department of Pediatric Surgery, University Clinical Center of Kosovo, Kosovo, Albania

^p Clinic of Medical Biochemistry, University Clinical Center of Kosovo, Kosovo, Albania

^qFaculty of Medicine University of Prishtina, Kosovo, Albania

ARTICLE INFO

Article history: Received 9 May 2022 Revised 13 July 2022 Accepted 17 July 2022

АВЅТ Я А С Т

Anatomic variations in the urogenital tract have generally been diagnosed through intravenous urography as a modality of choice. In recent years, computerized tomography (CT) urogram has replaced the traditional intravenous imaging of the genitourinary tract. Hematuria, tumoral mass, obstructive uropathy, and congenital collecting system abnormalities are indications for CT imaging. In this report, we present a young woman with intermittent right flank pain and recent urinary tract infection. Her history was also positive for spon-

^{*} Competing Interests: Authors declare no conflicts of interest.

^{*} Correspoding author.

E-mail address: zana.gafurri@uni-pr.edu (Z. Gafurri). https://doi.org/10.1016/j.radcr.2022.07.074

^{1930-0433/© 2022} The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Keywords: CT urography Renal anomaly Ectopic kidney Urogenital

Introduction

Congenital abnormalities of the kidney and urinary tract occur in 3-6 per 1000 live births [1]. They include a wide range of anomalies, from asymptomatic ectopic kidneys to lifethreatening renal agenesis [2]. When presenting with symptoms, they are associated with hypertension, proteinuria, and renal impairment. Many of them are detected in the antenatal or immediate postnatal period. However, a significant proportion is found in the adult population with varying severity, with subsequent renal complications like stone formation, infection, and renal failure. They are the leading cause of end-stage renal disease and kidney transplantation or dialysis in children age lower than 5 [1]. Congenital anomalies of the kidney and urinary tract (CAKUT) involves hypoplastic kidney, dysplastic kidney, horseshoe kidney, number (agenesis or supernumerary) and shape abnormalities, cystic renal disease, and ureteropelvic junction obstruction (UPJO) and vesicoureteral reflux (VUR), complex renal calyces and megacalycosis, calyceal diverticula, duplication anomalies, ureteroceles, and more. The most common anomaly of the urinary tract is a simple renal ectopia with one organ lying outside the renal fossa, most commonly in the pelvis [3]. Although anatomic and histologic evaluations are the gold standard for CAKUT diagnosis, histology is often not available [4]. Consequently, renal and urinary tract developmental anatomic variants and their complication can be evaluated with imaging studies alone, which can differentiate the anatomical variants of the normal renal urinary tract from the pathologic changes visible on renal calyces, infundibulum and renal pelvis morphology [5]. CT urography is the ideal imaging technique for scanning the retroperitoneal organs, such as the kidney and has become the most useful diagnostic of urinary tract (UT) congenital abnormalities, including complex anomalies [5,6]. Renal ectopia, crossed ectopia, horseshoe kidney, and renal agenesis are all detected easily by computerized tomography urogram (CTU). CTU can also be beneficial in the detection of UPJO and assessment of the potential of living renal donors. It can determine which ureteropelvic obstructions are due to extrinsic such as an aberrant renal artery or crossing vein, playing an essential role in evaluating potential living renal donors [7]. Urinary tract duplication also can be diagnosed by CTU, which should be considered a diagnostic test in isolated superior pole renal collecting system dilation [8,9]. CTU can reveal the exact site of the typically ectopic upper ureter insertion along with evaluation for the presence of an ureterocele, which appears as a round or oval filling defect within the urinary bladder at or near the site of ureter insertion. Ectopic ureteral insertions and ureteroceles are rarely detected in the setting of the non-duplicated upper urinary tract [7]. CTU may also play an important role in surgi-

taneous abortion. She was referred to the Radiology Clinic for a CT urography. Our aim, in this case report, is to highlight the role of CT urography in the early diagnosis of anatomical variations of the urogenital system and appropriate prevention of clinical progression. © 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license

(http://creativecommons.org/licenses/by-nc-nd/4.0/)

cal decision-making by providing necessary anatomic knowledge about the renal vasculature and the urinary tract [10]. The only limitation of CTU is radiation exposure ordering it only when indicated in pediatric, young adult, and pregnant patients [5].

Case presentation

A 28-year-old female patient presented to the urologist with the complaint of intermittent non defined pain in the right lumbar region. The pain was radiated to the lower part of abdomen. There were about 4 or 5 similar episodes during the month, and each episode lasted several days. There was no associated nausea or vomiting. Recent medical history showed a urinary tract infection (UTI)6 months ago. At that time patient complained of pelvic pain, dysuria, strong persistent urge to urinate and cola colored urine. She was treated and the symptoms and UTI were resolved after the antibiotic course. She had a history of a spontaneous abortion followed by a complicated pregnancy. A healthy baby was delivered, and an arcuate uterus was discovered intraoperatively by visual examination. She suffered from post-partum depression, but now she claims she is in a stable mental health and is not taking any medication. The patient's family history was unremarkable. Patient reported no smoking, no alcohol consumption. Being asked about any changes in urinary frequency or color, she reported that was urinating more frequently. A recurrent UTI was primarily suspected.

Initial vital signs, including blood pressure, pulse rate, respiratory rate, and body temperature were within normal range. Physical examination revealed right lumbar tenderness without guarding or rigidity, but she was otherwise fit and well. Examination findings were essentially normal, and the patient was sent for a complete blood count, urinalysis and electrolyte, urea, and creatinine.

Laboratory tests were performed at Clinic of Medical Biochemistry, showed hematuria and sterile pyuria in urine analysis. Liver function test (aspartate aminotransferase and alanine aminotransferase), serum creatinine, blood urea nitrogen, and electrolytes level were within normal limits. For further evaluation the patient was referred to the Radiology Clinic for a CT urography of the abdomen and the pelvic area.

A CT urography following the protocol was performed and revealed an ectopic and dysmorphic right kidney, located in the right upper pelvic cavity (Fig. 1A). Double collecting system is evident with upper collecting system emerging from the upper pole. Located where the 2 collecting systems merge is a non-obstructive, but large calculus in the right renal pelvis, just proximal to the upper ureter. The calculus is obscured in the contrast excretory phase (Fig. 1D). There is a single



Figure 1 – (A) CT urography coronal plane present: Ectopic and dysmorphic right kidney is seen, located in the right upper pelvic cavity. Double collecting system is evident with upper collecting system emerging from the upper pole. (B) Ct urography -MPR images axial plane present: Incidentally noted is the arcuate uterus, as an anatomical variant. (C) Ct urography -excretory phase axial plane present: Incidentally noted is the arcuate uterus, as an anatomical variant. (D) CT urography excretory phase sagittal plane present: located where the two collecting systems merge is a non-obstructive but large calculus in the right renal pelvis, just proximal to the upper ureter. The calculus is obscured in the contrast excretory phase. (E) CT urography excretory phase coronal plane present Double collecting system single ureter. (F) CT urography excretory phase coronal plane present: There is single ureter, despite the double collecting system. Barely enough total ureter length to join urinary bladder makes this an ectopic kidney case, rather than a mobile kidney where the ureter is usually longer with folding to adjust the kidney mobility between right upper quadrant and pelvic cavity.

ureter, despite the double collecting system. Barely enough total ureter length to join urinary bladder makes this ectopic kidney case, rather than a mobile kidney where the ureter is usually longer with folding to adjust the kidney mobility between right upper quadrant and pelvic cavity. (Fig. 1E and F) Incidentally noted is the arcuate uterus, as an anatomical variant (Fig. 1B and C) Surgical removal of the stone was advised but refused by the patient. Given the fact of the presence of a large stone and a history of UTI patient is now on a regular follow-up with assessments of kidney function and ultrasound scans.

Discussion

Duplex kidney or duplex collecting systems of the kidney is the most common congenital anomaly of the renal system and one of the most frequent developmental abnormalities in the pediatric population, with an estimated incidence of 1% [11–13]. Duplex kidneys are characterized by the presence of 2 pyelocaliceal systems in a single renal unit [12,15]. It presents bilaterally in approximately 20% of cases and occurs with a higher propensity in females. They are twice as likely to be affected, compared to males [13–15].

The condition stems due to anomalies during the normal embryogenesis of the renal system in the fourth week of gestation [12–15]. Ureteral development is characterized by the formation of a single ureteric bud from the mesonephric duct, subsequently branching and differentiating into the different components of the renal pyelocaliceal system [13]. Complete kidney duplex develops when 2 ureteric buds arise from the mesonephric duct, to give rise to 2 independent pyelocaliceal systems with their respective ureters that fuse with the renal mesenchyme independently, creating 2 renal section (the upper and lower moiety) that are drained separately [12,15-17]. Partial renal duplex is characterized by a varying degree of incomplete duplication of the collecting system, due to the premature division of the ureteric bud, ahead of its fusion with the mesenchyme [12,16,17]. Phenotypically, it may present as bifid renal pelvis, partial ureteral duplication (Y-shaped ureter) or incomplete ureteral duplication with ureters joining near or in bladder wall (V-shaped ureter), in any case the systems ultimately drain in a single ureter [15].

This variant is usually asymptomatic, but may occasionally be associated with serious complications and morbidity, depending on the involved moiety. Complications associated with the upper moiety include multicystic dysplasia and an ectopic insertion of the ureter in a complete duplex, with or without the coexistence of an ureterocele, that may lead to urinary incontinence [12,15,17]. Complications involving the lower moiety include VUR, that predisposes to recurrent UTIs, stone formation, renal scarring and functional impairment [12,15,17]. Pyeloureteral obstruction may also develop [15]. Symptoms of duplex kidney include flank pain, urinary incontinence, hematuria, recurrent UTIs and they present overwhelmingly in pediatric patients [13,14]. Adults with duplex kidney are generally asymptomatic and they are diagnosed incidentally during imaging exams, the variant is usually of no clinical importance in these patients [12].

Renal ectopia is defined as an abnormal positioning of the kidney, outside the renal fossa, due to abnormalities in its ascent during early renal embryogenesis [18,19]. It develops as a result of an aberration during the normal migration of the metanephros and the ureteric bud from the pelvis to the lumbar region, between the sixth to eighth week of gestation [20]. Depending on its location, ectopic kidneys are classified into thoracic, abdominal, iliac and pelvic, with pelvic kidney being the most prevalent variation [21]. The incidence of pelvic kidneys is reported to be approximately 1:2200-3000 patients [22]. The ectopic kidney may present ipsilaterally to its ureteral insertion, simple ectopia or contralaterally to its ureteral insertion, cross ectopia, that is often associated with partial fusion with the orthotopic kidney, as well [19]. Pelvic kidneys may present with malrotation, hydronephrosis due to obstruction or VUR, extrarenal calices, nephrolithiasis and UPJO [18,20,-23]. Obstruction has been hypothesized to derive from the high insertion of the ureters into the pyelocaliceal system, impeding the normal drainage and facilitating urinary stasis [23]. Patients are largely asymptomatic and ectopic kidneys are incidental findings, nevertheless they show a slightly increased susceptibility to UTIs, nephrolithiasis and traumatic injuries [22]. There have been reports of an association of ectopic kidneys with concurrent genital developmental anomalies or variations, as was the case with our patient [21,23].

To the best of our knowledge, our patient represents one of the few cases reported in literature of a concomitant presence of two congenital renal developmental anomalies, a pelvic and partial duplex kidney, associated with a concurrent uterine malformation, an arcuate uterus.

Conclusion

In our patient, an ectopic and dysmorphic kidney associated with uterus variation was noticed. The structural visualization of kidney parenchyma and urogenital tract through CT urography was of high quality. This can lead to earlier detection of the anomalies and proper treatment suggestions with no delay. Given the good imaging outcomes, CT urography can be the gold standard in diagnosing anatomic variations of the urogenital tract.

Patient consent statement

Informed Consent Statement: Written informed consent has been obtained from the patient to publish this paper.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2022.07.074.

REFERENCES

- [1] Yosypiv IV. Congenital anomalies of the kidney and urinary tract: a genetic disorder? Int J Nephrol 2012;2012:20.
- [2] Ramanathan S, Kumar D, Khanna M, Al Heidous M, Sheikh A, Virmani V, Palaniappan Y. Multi-modality imaging review of congenital abnormalities of kidney and upper urinary tract. World J Radiol 2016;8(2):132.
- [3] Thorner P, Bernstein J, Landing BH. Kidney and lower urinary tract. In: Diseases of the fetus and newborn. London: Chapman and Hall Medical; 1995. p. 609–91.
- [4] Ramanathan S, Kumar D, Khanna M, Al Heidous M, Sheikh A, Virmani V, et al. Multi-modality imaging review of congenital abnormalities of kidney and upper urinary tract. World J Radiol 2016;8(2):132.
- [5] Quaia E, Martingano P, Cavallaro M, Premm M, Angileri R. Normal radiological anatomy and anatomical variants of the kidney. Radiol Imaging Kidney 2014;Second Edition:17–74.
- [6] Abou-El-Ghar M, Refaie H, Sharaf D, El-Diasty T. Diagnosing urinary tract abnormalities: intravenous urography or CT urography. Rep Med Imaging 2014;7:55–63. doi:10.2147/RMI.S35592.
- [7] Mitsumori A, Kotaro Y, Akaki S, Togami JJ, Hashimoto H, Komon H, et al. Evaluation of crossing vessels in patients with ureteropelvic junction obstruction by means of helical CT. RadioGraphics 2000;20:1383–93.
- [8] Caoili EM, Cohan RH, Korobkin M, Platt JF, Francis I, Faerber JE, et al. Urinary tract abnormalities: initial experience with multi- detector row CT urography. Radiology 2002;222:353–60.
- [9] Chow LC, Sommer FG. Multidetector CT urography with abdominal compression and three- dimensional reconstruction. AJR Am J Roentgenol 2001;177:849–55.
- [10] Raman SS, Pojchamarnwiputh S, Muangsomboon K, Schulman PG, Gritsch HA, Lu DS, et al. Surgically relevant normal and variant renal parenchymal and vascular anatomy in preoperative 16-MDCT evaluation of potential laparoscopic renal donors. AJR Am J Roentgenol 2007;188:105–14.
- [11] Privett J T, Jeans WD, Roylance J. "The incidence and importance of renal duplication.". Clin Radiol 1976;27(4):521–30. doi:10.1016/s0009-9260(76)80121-3.

- [12] Doery A J, Eileen A, Michael R. "Duplex kidney: not just a drooping lily.". J Med Imaging Radiat Oncol 2015;59(2):149–53. doi:10.1111/1754-9485.12285.
- [13] Davda S, Vohra A. Adult duplex kidneys: an important differential diagnosis in patients with abdominal cysts. JRSM Short Rep 2013;4(2):13–15. doi:10.1177/2042533312472126.
- [14] Clement K, Hendry J, Keeley FX, Aboumarzouk OM. "Heminephrectomy in adults: a systematic review with cumulative analysis.". Urol Int 2018;101(1):1–6. doi:10.1159/000485466.
- [15] Giorlando Francesca, et al. "Duplex collecting system in a pelvic kidney - an unusual combination.". J Radiol Case Rep 2017;11(12):8–15. doi:10.3941/jrcr.v11i12.2991.
- [16] Whitten S M, Wilcox D T. "Duplex systems.". Prenatal Diagn 2001;21(11):952–7. doi:10.1002/pd.206.
- [17] Jain S, Chen F. "Developmental pathology of congenital kidney and urinary tract anomalies.". Clin Kidney J 2018;12(3):382–99. doi:10.1093/ckj/sfy112.
- [18] Editor(s): Donnelly Lane F. CHAPTER SIX Genitourinary. In: Donnelly Lane F, editor. Pediatric Imaging W.B. Saunders; 2009. p. 125–55. PagesISBN 9781416059073. doi:10.1016/B978-1-4160-5907-3.00006-4.
- [19] Editor(s): Engen Rachel, Hingorani Sangeeta. 87 -Developmental abnormalities of the kidneys. Avery's Diseases of the Newborn. Gleason Christine A, Juul Sandra E, editors. (10th Edition) Elsevier; 2018. 1250-1259.e2, ISBN 9780323401395. doi:10.1016/B978-0-323-40139-5.00087-5.
- [20] Bingham G, Leslie SW. Pelvic kidney. StatPearls [Internet] [Updated 2021 Aug 12] In:, Treasure Island (FL): StatPearls Publishing; 2021. Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK563239/.
- [21] Editor(s): Merguerian Paul A, Rowe Courtney K. 88 -Developmental abnormalities of the genitourinary system. Avery's Diseases of the Newborn. Gleason Christine A, Juul Sandra E, editors. (10th Edition) Elsevier; 2018. Pages 1260-1273.e4, ISBN 9780323401395. doi:10.1016/B978-0-323-40139-5.00088-7.
- [22] Cinman Nadya M, Okeke Z, Smith AD. "Pelvic kidney: associated diseases and treatment.". J Endourol 2007;21(8):836–42. doi:10.1089/end.2007.9945.
- [23] Eid Seif, Iwanaga J, Loukas M, Oskouian RJ, Tubbs RSH. "Pelvic kidney: a review of the literature.". Cureus 2018;10(6):2775 9 Jun. doi:10.7759/cureus.2775.