ISSN: 2572-3235

Sangey T et al. Int J Radiol Imaging Technol 2022, 8:091
DOI: 10.23937/2572-3235.1510091
Volume 8 | Issue 1
Open Access

International Journal of Radiology and Imaging Technology

CASE REPORT

Multi Imaging Modality in Diagnosis of Unusual Giant Hydronephrosis of the Non Functioning Lower Moiety in A Partial Duplex Kidney with Distal Ureteric Stricture

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Abstract

Duplex collecting system of the kidney is a common congenital anomaly of the urinary tract. It is usually asymptomatic clinically with normal renal function. In rare instances, it may complicate with giant hydronephrosis. The following is a case of a middle aged woman with history of recurrent urinary tract infections (UTI) and flank pain diagnosed with bilateral partial duplex kidney complicated with unusual non-functioning lower moiety left giant hydronephrosis due to distal left ureteric stenosis which probably enhanced the reverse flow of urine together with YO-YO reflux. The left upper moiety was preserved and functioning. There was also right bifid ureter seen (Y Shaped). Advanced cross sectional imaging modalities has simplified the diagnosis of this condition.

Introduction

Duplex collecting system of the kidney is a common congenital anomaly of the urinary tract, with an incidence of 0.8 - 1% [1-3]. They are bilateral in 20% of cases and are almost twice as common in females as males. They are usually diagnosed and treated in children, but in some instances may go unnoticed up to adulthood [3,4].

Duplex kidney is result of partial or total duplication of the ureteric bud. It is characterized by incomplete fusion of upper and lower kidney moieties accompanied by complete or incomplete duplications of pyeloureter [5].

Most duplex kidneys are asymptomatic and diagnosed incidentally in adult hood. We describe a case of middle aged woman with history of repeated urinary tract infections (UTI) and left flank pain. The diagnosis of bilateral partial duplex kidney with unusual non-functioning left lower moiety giant hydronephrosis due distal ureteric stenosis which probably enhanced the reverse flow of urine together with YO-YO reflux.

Case Report

A 46-year-old female, with no history of chronic illness presented in our out-patient department with complaints of epigastric pain and flank pain, more pronounced on the left side than the right. She had no other complaints. There was no reported urine incontinence or change in urine stream. She had a long-standing history of recurrent Urinary Tract Infection.

On physical examination, her vital signs were within normal range - temperature 36.7 C, blood pressure 120/70 mmHg, pulse rate was regular at 102 beats per minute, and Oxygen saturation was 99% at room air. Abdominal exam was unremarkable. All laboratory findings were normal and within range. Urine analysis, in particular, showed no leucocyturia, pyuria, or bacteriuria.

On radiological examination, ultra-sonogram of the abdomen revealed a large left side intra-abdominal cystic lesion occupying the left hemi-abdomen pushing the bowel and other solid organs to the right side.

Computed Tomography Urography (CTU) with delayed images showed bilateral partialrenal duplex kidney. The right kidney appeared as partial duplex

Citation: Sangey T, Khan ZD, Hassanali LS (2022) Multi Imaging Modality in Diagnosis of Unusual Giant Hydronephrosis of the Non Functioning Lower Moiety in A Partial Duplex Kidney with Distal Ureteric Stricture. Int J Radiol Imaging Technol 8:091. doi.org/10.23937/2572-3235.1510091

Accepted: June 16, 2022: **Published:** June 18, 2022

kidney (Y-Shaped) with moderate Pelvic Ureteral Junction (PUJ) dilatation and narrowing of the proximal ureter. The left kidney showed a marked cystic dilatation (15.3 × 6.24 cm) of non-functioning lower moietypelvic-calyceal region with no remaining visible parenchyma. The functional upper renal moiety was medially compressed having normal enhancing parenchyma and contrast excreted with dilated proximal ureter (9 mm) on delayed scan. No evidence of calculus on non-contrasted images.

Magnetic Resonance Urography (MRU) showed upper moiety ureter insertion inferiorly to the non-functioning lower moiety giant hydronephrotic Pelvic-calyceal system. The ureter proximally appears dilated and tortuous; distally there is beak mouth narrowing of the ureter noted (stricture). The vesico ureteric junction was patent. No evidence of ureterocele seen (Figure 1, Figure 2, Figure 3 and Figure 4).

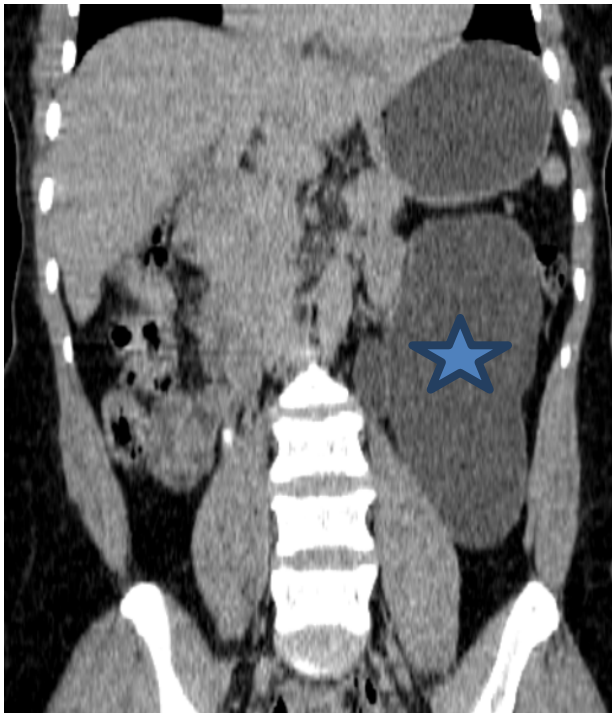

Figure 1: Coronal reformatted CE-CT showing a left sided Giant hydronephrosis occupying the left hemi abdomen denoted by star.

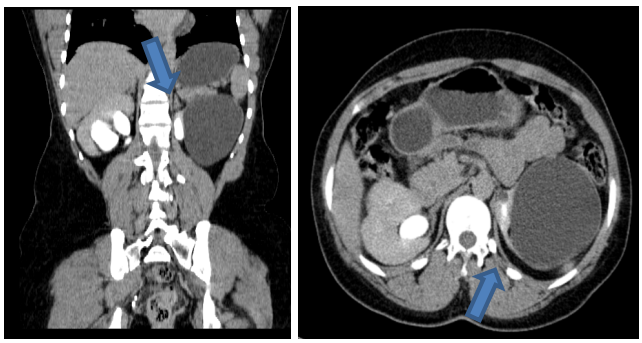

Figure 2: Coronal and axial delayed phase showing medially compressed and contrast excretion of the left upper moiety (blue arrow) and excretion of both right kidney moiety.

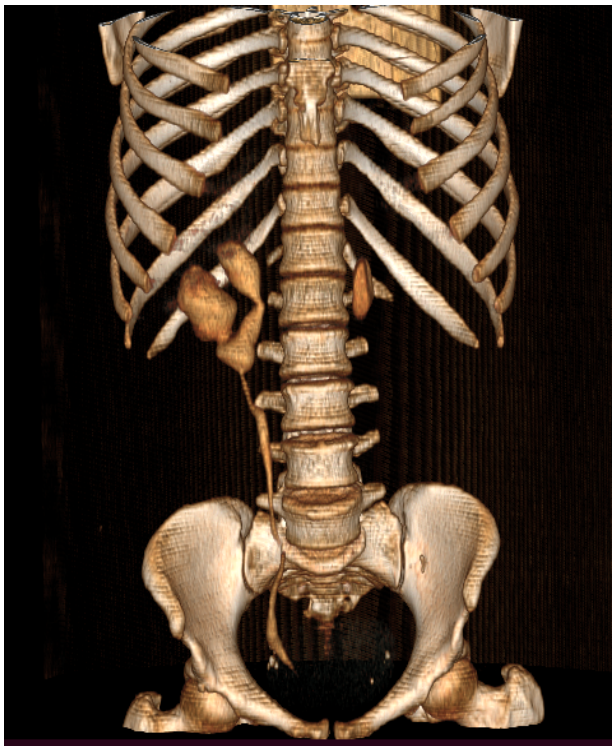

Figure 3: Reformatted image showing Right bifidureter with mild dilated PUJ and proximal ureteric narrowing;

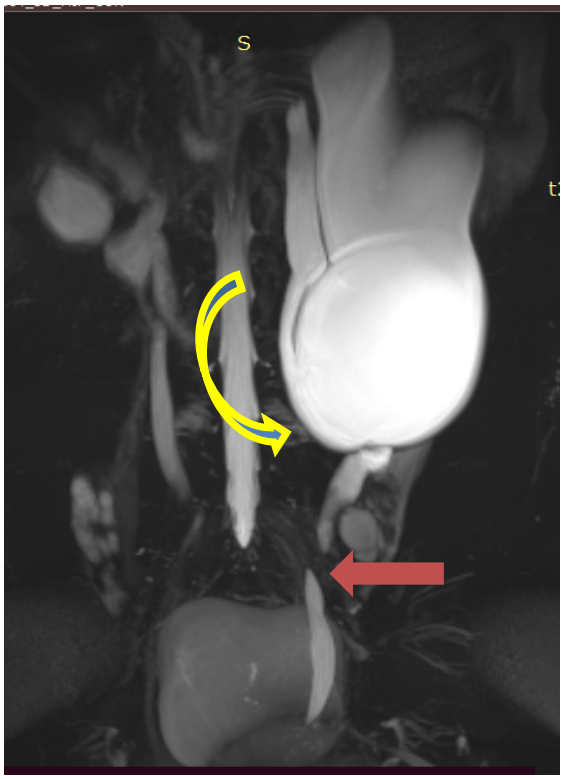

Figure 4: MRU showing a left Giant hydronephrosis with distal lower moiety ureteric stenosis (red arrow). Upper moiety ureter insertion inferiorly to the lower moiety PUJ (curved yellow arrow).

Discussion

Duplex kidney is defined as a renal unit comprised of two pelvicalyceal systems [6]. It is a result of partial or total duplication of the ureteric bud. Depending

on the degree of fusion, it can present as bifid renal pelvis, partial (incomplete) ureteric duplication with ureters joining proximally or near the bladder wall and complete ureteric duplication with separate ureteric orifices [7,8]. In complete duplex, the ureteral drainage pattern is normally governed by the Meyer-Weigert rule, predicting that the ureter of the upper part usually drains infero-medially into an ectopic location, and the lower ureter part usually drains supero-laterally to its normal insertion in the trigon [9,10]. In partial duplex kidney, it is associated with either retrograde peristalsis of urine as a result of pressure gradient between two ureteric segments termed as "Yo-Yo reflux" or pelvic-ureteric junction obstruction [11,12].

Majority of duplex abnormalities are asymptomatic and of no clinical significance [13]. In symptomatic patients, they are commonly associated with recurrent urinary tract infections, flank pain, urinary incontinence and haematuria [13,14].

Giant hydronephrosis is a rare entity and occurs as a complication in duplex kidney [15,16]. It is defined as containing not less than 1 liter of fluid in the collecting system [17] or 1.6% of body weight of fluid in the collecting system or the involvement of five vertebral bodies in length or enlarged kidney occupying the hemi abdomen or crossing the midline [18,19].

The complications relating to duplex kidneys are specific to the moiety. Incomplete duplex kidney, the upper moiety hydronephrosis is usually due to the obstruction at the lower end (ureterocele) while in the lower moiety it is usually due to the vesico-ureteric reflux obeying the Meyer Weigert law [19,20]. However, a few exceptions to the Meyer-Weigert rule has also been published that deal with anatomical variation or ectopic insertion of the ureter of the lower renal pole with vesico-ureteric reflux [21,22] and lower distal ureteric stenosis [23,24].

For partial duplex kidney the uretero-ureteral (Yo-Yo reflux) is mostly responsible for reflux of the reverse urine flow and prevents the urinary tract from ever being completely drained [25]. The pressure of the lower moiety is generally higher than the upper moiety and therefore the urine generally refluxes from the lower moiety to the upper moiety [26]. However this is not a rule as seen in our case where the giant hydronephrosis was in the lower moiety.

Giant hydronephrosis is a slow progressive disease. If undiagnosed and untreated, it can result in long term complications like hypertension, respiratory distress, rupture of the kidney, renal failure and malignant transformation [27-29].

The complications that occur in duplex kidneys are specific to the moiety. The obstruction in the upper pole moiety is associated with anomalies such as ectopic ureter or ureterocele, while Pelvic-Ureteric Junction (PUJ) obstruction and vesicoureteric reflux are associated with the lower pole moiety obstruction [19,20,30].

In regard to imaging modalities, ultrasonography is typically the first imaging modality as it is easily available, cost effective, non-invasive and radiation free in the evaluation of suspected urinary tract duplication but it has a limited role in detection of existing complication in the duplex kidney [31,32]. It cannot precisely distinguish duplex kidneys from a range of other disorders causing renal dilatation like giant hydronephrosis, solitary large renal cyst, polycystic kidney disease [33-35].

Complex duplex renal systems may require further evaluation using cross-sectional imaging, which include CT Urography and MR Urography providing comprehensive morphologic and functional evaluation of complex duplex kidneys [36,37]. Non contrasted CT is useful in detecting presence of calculi causing Pelvic Ureteral Junction (PUJ) or ureteric obstruction [2,38]. Prolonged-delay enhanced CT Urographyis useful for exposing the entire course of the duplicated ureters and existing complications, assessment of the residual parenchyma and also demonstrating the crossing vessels before surgical planning [32,39,40].

High resolution MR Urography is valuable in the evaluation of ectopic ureters as well as non-functioning renal poles even without contrast because they are frequently filled with T2-hyperintense fluid representing excreted urine [37,41,42]. The dynamic renal scintigraphy is the modality of choice to diagnose Yo-Yo reflux [12,43] which was not available at our Health Centre.

Conclusion

Duplex collecting system of the kidney is a common congenital anomaly of the urinary tract. It is usually asymptomatic clinically with normal renal function on laboratory investigation. In rare instances, it may present with giant hydronephrosis. We presented a case bilateral partial duplex kidney complicated with unusual non-functioning lower moiety left giant hydronephrosis due to distal left ureteric stenosis which probably enhanced the reverse flow of urine together with YO-YO reflux. Therefore, it may be a pitfall in clinical practice.

The early detection of the anomalies is helpful in avoiding complications related to the duplex collecting system.

Multiple imaging modalities provide opportunities to diagnose and characterize both anatomical and functional information of the duplicating collecting system with existing complication.

Contributors

TIS was involved with performing the literature search, interpreting the images patient and discussion writing. ZDK was involved in writing of the manuscript

and formulating the final manuscript. Also was involved with proofreading and making amendments to the entire case report and manuscript. LSH was involved in clerking the patient, helping obtain consent from the patient and searching for literature with proofreading of the manuscript.

Funding

This case report was neither funded nor sponsored.

Conflict of Interests

The authors declare that there is no conflict of interests.

Patient Consent

Patient was clearly explained and written informed consent was signed to partake in case report.

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