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Original Research Article

Duplex kidney anomalies and associated pathology: a single centre retrospective review

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ABSTRACT

Background: Duplex kidneys are common developmental renal anomaly with an incidence of 1% in healthy adult population. Adult individuals may present as non-functional moiety, calculus disease or an incidental finding. Duplex kidney is defined as a renal unit comprised of two pelvicalyceal systems. Based on the degree of fusion, it can present as bifid renal pelvis, partial ureteric duplication (Y-shaped ureter), incomplete ureteric duplication with ureters joining near or in bladder wall (V-shaped ureter) and complete ureteric duplication with separate ureteric orifices. The purpose of this study is to retrospectively review the various duplex kidney anomalies and associated pathology.
Methods: This is a retrospective study, performed at Department of Urology, Gauhati Medical College Hospital, a tertiary centre, from September, 2018 to August, 2019. Preoperative imaging, plain intravenous urography or computed tomography intravenous urography was done.
Results: 29 patients were diagnosed with variants of duplex kidney anomaly. It includes right side, left side and bilateral anomalies. Among all the patients 4 had incidental findings. 16 patients had associated renal or ureteral calculus. 5 patients had associated ureteropelvic junction obstruction while 1 patient had ureterovesical junction stricture. We found single patient in each group of duplex kidney anomaly associated with non-functional moiety with renal cell carcinoma, ureterocele with urothelial malignancy and horseshoe kidney with ureteropelvic junction obstruction respectively.
Conclusions: Duplex kidney anomaly in most individuals is of no clinical significance. High index of suspicion along with good quality imaging can accurately detect specific anomaly and associated condition.

Keywords: Duplex kidney anomaly, Bifid pelvis, Complete ureteral duplication

INTRODUCTION

Duplex kidneys are a common abnormality of renal tract development, carrying an incidence of approximately 1% in healthy adult population and 2-4% of patients investigated for urinary tract symptoms.[1-4] It also accounts for 7.2% of patients with congenital urinary tract anomalies.[5] Although it is one of the most common anomalies of the kidney, this condition is under reported because most of such patients remain asymptomatic throughout their life.[6] Symptomatic children develop serious complications in their childhood or in the early adolescent period with recurrent breakthrough urinary tract infections, urinary incontinence or progressive deterioration of renal function, needing some form of medical or surgical intervention.[7] Those who present in adulthood may present as non-functional moiety, calculus disease or an incidental finding. Duplex kidney is defined as a renal unit comprised of two pelvicalyceal systems.[8,9] Based on the degree of fusion, it can present as bifid renal

pelvis, partial ureteric duplication (Y-shaped ureter), incomplete ureteric duplication with ureters joining near or in bladder wall (V-shaped ureter) and complete ureteric duplication with separate ureteric orifices.[10,11] The majority of duplex abnormalities are asymptomatic and carry no clinical significance.[3] Duplex kidneys can however be associated with complications. Recognising these patients early is important as severe complications are symptomatic and can be treated effectively by early intervention.[12]

The purpose of this study is to retrospectively review the various duplication anomalies of the kidneys that we have encountered in our institution and also the different modes of presentation, associated pathology and modalities of treatment.

METHODS

This is a retrospective study and was performed at our institution, Department of Urology, Gauhati Medical College Hospital, a tertiary care centre. Ethical committee clearance taken. Study period was from September, 2018 to August, 2019. Those patients presented in that time period to outpatient department or underwent surgical intervention for some conditions or pathology associated with different variants of duplex kidneys were included in this study. Total 29 patients were included in the study. Preoperative imaging, plain intravenous urography or computed tomography intravenous urography (CTIVU) was done. All the parameters were entered in Microsoft Excel sheet (version 2007). Percentages were calculated. As our study didn't require any complex statistical calculation or comparison between groups, no special statistical software required in our study. Asymptomatic patients with incidental detection of variants of duplex kidney anomaly did not require any surgical intervention. While other symptomatic patients with definite pathology and conditions associated with variants of duplex kidney anomaly underwent definite surgical intervention depending upon their individual condition.

RESULTS

A total of 29 patients were diagnosed with variants of duplex kidney anomaly. Among them 9 (31%) individuals were female and 20 (69%) individuals were male (Figure 1).

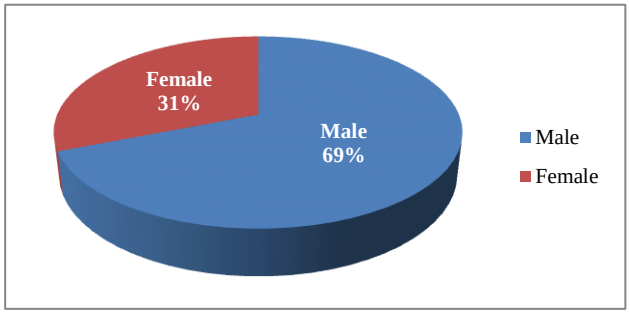

Figure 1: Sex distribution of patients.

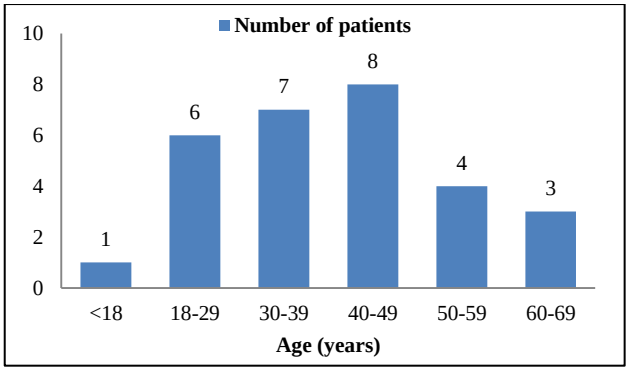

Figure 2: Distribution of patients as per age group.

Patients were divided as per the age group. Lowest age is 14 years and the eldest individual is 65 years old (Figure 2).

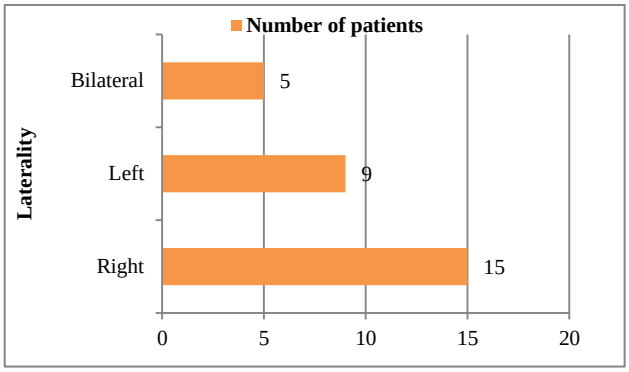

Figure 3: Laterality of variants of duplex renal collecting system anomalies.

Patients were also studied depending on their laterality of variants of duplex renal collecting system anomalies. 9 patients (31.04%) had left sided anomaly, 15 patients (51.72%) had right sided anomaly. 5 patients (17.24%) were found to harbour bilateral anomaly (Figure 3).

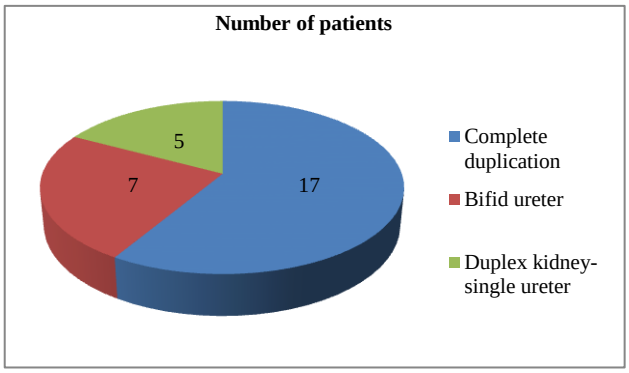

Figure 4: Different variants of duplicated renal collecting system anomalies among the patients.

Among the patients 17 patients (58.62%) were found to have complete duplicated ureter with each ureter draining separately into urinary bladder. No patient with complete

duplicated ureter was found to have ureter draining into ectopic location such as vagina in female or urethra in male and female. 7 patients (24.14%) had bifid ureter. They had duplex kidney with two pelvicalyceal system and bifid ureter, bifid ureter joining to become single ureter and draining into the urinary bladder as single ureter. 5 patients (17.24%) had duplex kidney i.e. two pelvicalyceal system from two renal moiety joining at pelviureteral junction to become single ureter and draining into the urinary bladder as single ureter. This is also known as bifid pelvis (Figure 4).

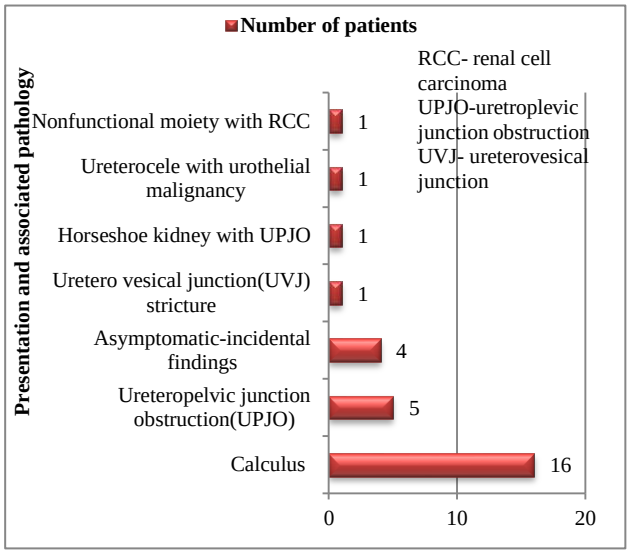

Figure 5: Spectrum of variants of duplex renal collecting system anomaly and the specific pathology and conditions associated with them.

We have analysed in detail the specific variants of renal collecting system anomaly and pathology associated with it. Four patients (n=4) presented with incidental findings as they were investigated outside and referred to us for incidental duplex kidney anomaly finding in urography images. Five patients (n=5) had duplex kidney anomaly associated with ureteroplevic junction obstruction (UPJO). Sixteen patients (n=16) in our study group had duplex kidney anomalies associated with urolithiasis. One patient (n=1) had non-functional moiety with renal cell carcinoma. Duplex kidney anomalies with ureterocele and urothelial malignancy, horseshoe kidney with UPJO, ureterovesical junction stricture groups had one patient (n=1) in each of them (Figure 5).

Some unique cases are highlighted here. One patient had bilateral complete ureteral duplication with bilateral non-functional upper moiety. He underwent bilateral heminephrectomy of non-functional upper moiety. Histopathology revealed a component of clear cell variant of renal cell carcinoma (RCC) in left hemi-nephrectomy specimen (Figure 6).

One patient presented with hematuria and right flank pain. CTIVU revealed right side complete ureteral duplication with ureterocele of upper moiety and a mass lesion within the ureterocele. This resulted in non-functional upper moiety. He underwent transurethral resection of bladder tumour. Histopatholgy of the mass revealed urothelial malignancy. Then laparoscopic right side radical nephroureterctomy with bladder cuff excision was done with curative intent (Figure 7).

One patient had horseshoe kidney with left side duplex kidney with bifid ureter. The left lower moiety had UPJO. She underwent pyeloplasty of left lower moiety (Figure 8).

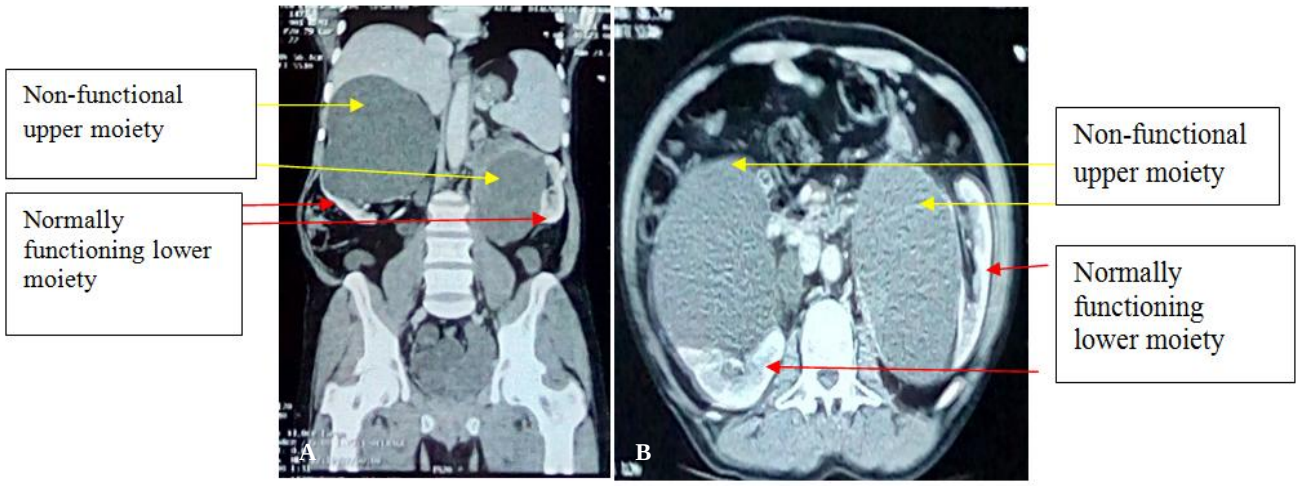

Figure 6 (A and B): Complete ureteral duplication with bilateral non functional upper moiety (yellow arrow) and normally functioning lower moiety (red arrow); he underwent bilateral heminephrectomy of upper non functional upper moiety; histopathology revealed a component of clear cell variant of renal cell carcinoma (RCC) in left heminephrectomy specimen.

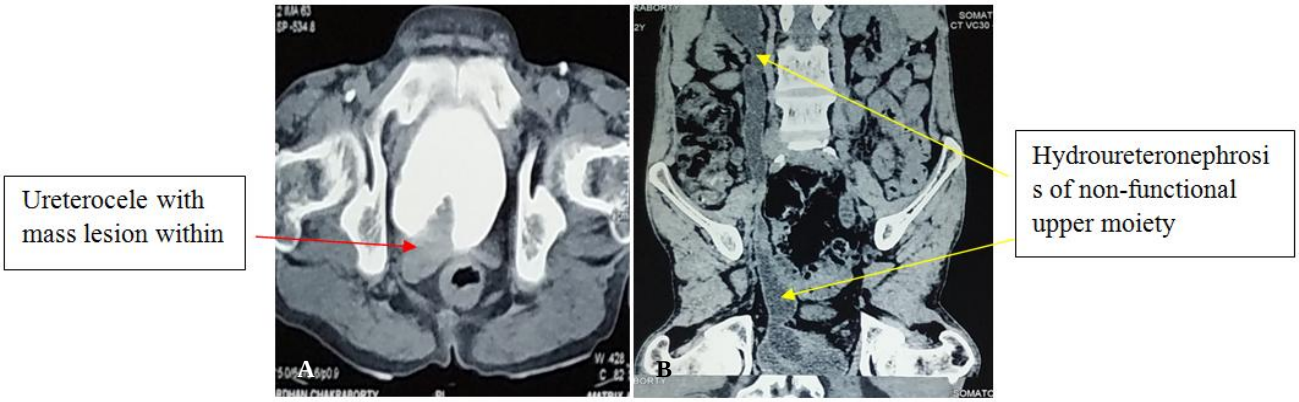

Figure 7 (A and B): Right side complete ureteral duplication with ureterocele of upper moiety and a mass lesion within the ureterocele (red arrow); resulted in hydroureteronephrosis of non-functional upper moiety (yellow arrow).

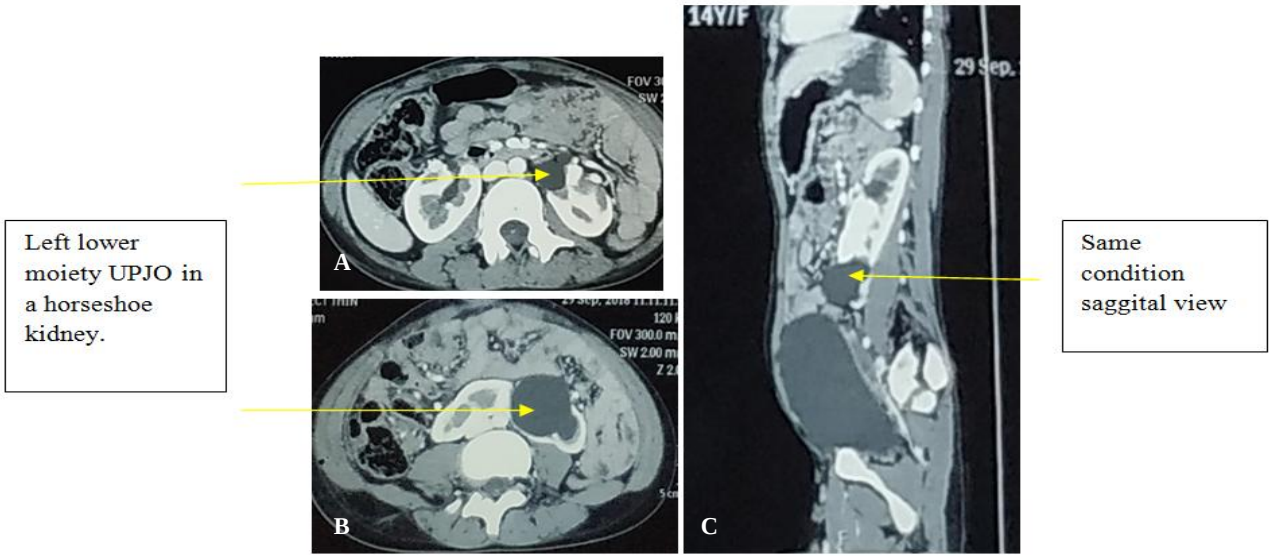

Figure 8 (A-C): Left lower moiety UPJO in a horseshoe kidney with left side duplex kidney and bifid ureter causing hydronephrosis of left lower moiety (yellow arrow).

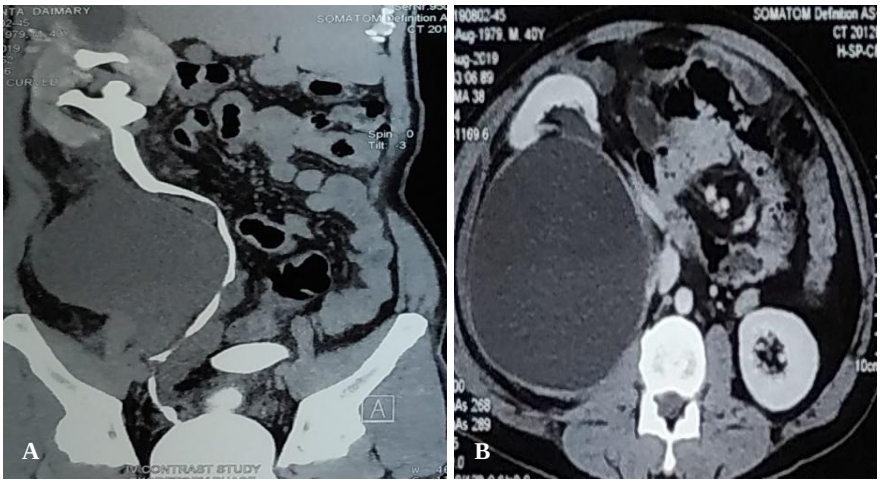

Figure 9 (A and B): Right side complete ureteral duplication with non-functional lower moiety due to ureteropelvic junction obstruction.

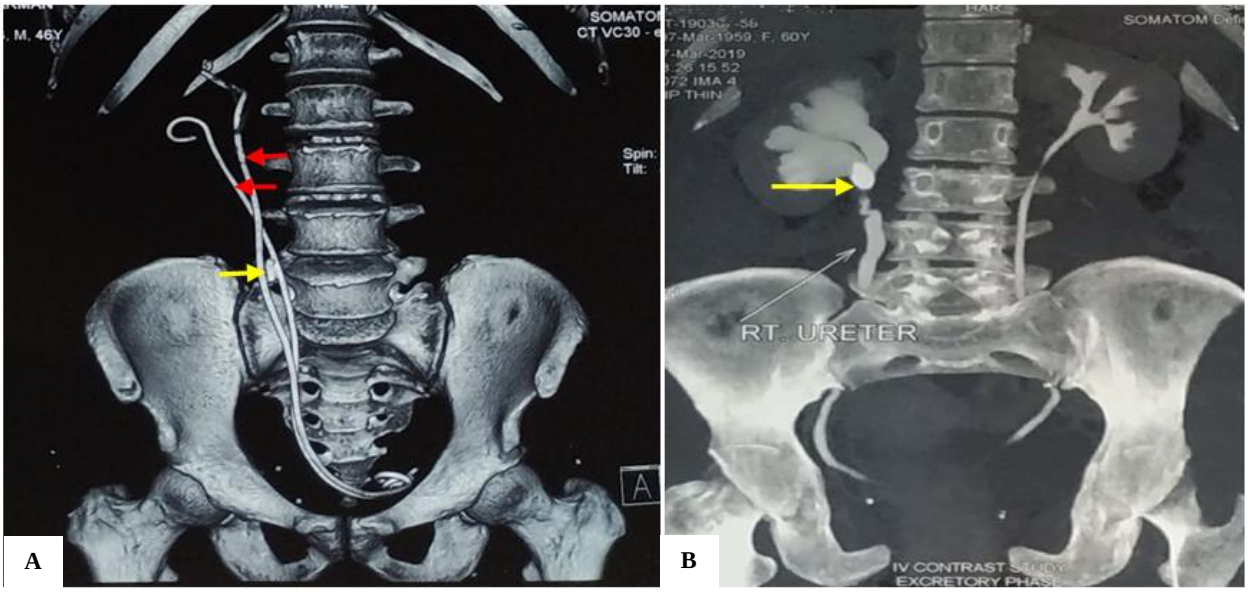

Figure 10: (A) 3D reconstructed computed tomography film showing right side complete ureteral duplication with calculus (yellow arrow) in upper ureter of lower moiety and double J stent (red arrow) in both moiety; (B) CTIVU film showing right side duplex kidney with two pelvicalyceal system joining at pelvis and forming single ureter; he had calculus (yellow arrow) in right pelviureteral junction.

Five patients (13.79%) had different variants of duplex renal system abnormality with non-functional moiety due to ureteroplevic junction obstruction. They underwent heminephrectomy. One such example given below had right side duplex kidney with UPJO of lower moiety causing non-functional moiety (Figure 9).

Sixteen patients (n=16) in our study group had different variants of renal duplex anomaly associated with calculus in upper moiety or lower moiety or pelvis or ureter (Figure 10). Patients with renal calculus underwent percutaneous nephrolithotomy, while patients with ureteral calculus underwent ureteroscopic lithotripsy.

DISCUSSION

Duplex kidney is defined as a renal unit comprised of two pelvicalyceal systems and is the most frequent congenital anomaly of the urinary tract.[8,13,14] There are many variations within this condition.[10] In a complete duplex kidney, there are two separate pelvicalyceal systems that have arisen from two ureteral buds from the mesonephric duct.[6,15] This results in two ureters. These two ureters fuse separately with developing renal blastoma and results in two independent draining sections of kidney. Ureters are termed as upper and lower moiety ureter depending on the draining renal segment. The orifice of the upper moiety is characteristically located medial and caudal than the orifice of the lower moiety that is located cranial and lateral. This relationship is consistent and it is called, Weigert-Meyer rule, which is maintained in more than 90% of the cases.[16] In partial duplication anomaly there is single ureteric budding, but it divides prematurely before meeting the mesenchyme. This leads to spectrum of incomplete duplication within ureter and kidney like bifid ureter or duplex kidney having two pelvicalyceal system, that join at pelviureteral junction leading to bifid pelvis.[1,6,17] In our study we have also found all types of variants of duplex kidney anomalies like complete duplication (n=17), duplex kidney with single ureter (n=5) and two ureter (bifid ureter)joining to become a single ureter (n=7) that drains into urinary bladder.

Despite being one of the most common anomalies of the kidney, this condition is grossly under reported. This is because they are either missed out during routine antenatal screening and/or as they remain largely asymptomatic in the early postnatal period.[6]

However, the risk of renal infection in children is increased by 20 folds with advancement in age. Many of these children develop serious complications in their childhood or in the early adolescent period with recurrent breakthrough urinary tract infections, urinary incontinence or progressive deterioration of renal function, needing some form of medical or surgical intervention.[5]

Some of the most frequent complications are associated with ectopic insertion of the upper moiety ureter of a complete duplication of ureter, often associated with ureterocele.[6] This in female can cause incontinence if insertion of ureter is below the urethral sphincter.[18] In males it can cause a pelvic mass as fluid slowly drains into an accessory sexual structure.[10]

Another common complication is reflux of the lower moiety and is due to the lateral displacement of the

ureteral insertion into the bladder.[3] It can predispose to recurrent urinary tract infections.[1,19]

Some literature has mentioned possible dominant nature of inheritance of duplex kidney variants and predominance in females.[6,20]

Our study has shown wide variation of anomalies on either side as well as both sides. In our study variants of duplex kidney anomalies were associated with wide variation of condition like non-functional moiety, ureteropelvic junction obstruction (n=5), ureterovesical junction stricture (n=1), calculus disease (n=16), ureterocele (n=1), malignant conditions like renal cell carcinoma (n=1) and urothelial malignancy (n=1).

In a study by Kullendorff and Wallin, where dimercaptosuccinic acid scintigraphy was used to characterise renal function, 44% cases with duplex kidneys had severely deteriorated renal function.[21]

Elhadi et al presented a case report of left side duplex kidney with lower moiety renal calculus.[22] Several other studies has also mentioned calculus in urinary tract associated with duplex kidney.[23,24]

Our study has included one patient of complete ureteral duplication with renal cell carcinoma in upper moiety. Mohan et al presented a very uncommon case report of right side duplex kidney with right renal lower polar mass. Patient underwent right radical nephrectomy, post op histopathological examination revealed renal cell carcinoma of conventional type.[25]

In our study one case of urothelial malignancy with ureterocele associated with duplex kidney anomalies found. Association of urothelial carcinoma very rare with duplex kidney. In 2019 a published case report shows ureteral urothelial carcinoma in a North African 52-year-old male patient, in a right duplex system. Radiological explorations concluded a non-functional upper right kidney. A suspect mass was observed in the lumbar part of the ureter of the right upper system. The meatus of the tumorous ureter ended in the right lobe of the prostate. A right hemi-nephro-ureterectomy was performed. A histological examination concluded a pT2G2 urothelial carcinoma.[26]

CONCLUSION

Different variants of duplex kidney anomaly are common congenital anomaly of upper urinary tract. In most individuals it is of no clinical significance. However in some individuals there are other associated congenital and acquired urinary tract conditions. High index of suspicion along with good quality imaging can accurately detect specific anomaly and associated condition. Precise detection of anomaly helps in accurate management of the associated conditions and leads to better quality of life.

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Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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