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Case Report

Duplex collecting system with renal malrotation: an unusual combination

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ABSTRACT

Both duplex collecting system and renal malrotation are congenital developmental anomalies of kidney. They usually present asymptomatic and accidentally diagnosed while patients undergo radiology examinations. We are reporting a case of a 33-year-old-male with a combination of duplex and malrotation kidney. Although these two are common anomalies of kidney, their combination is extremely rare. We found only two reports that had similar combined cases, so our case is the third reported.

Keywords: Bifid renal pelvis, Duplex kidney, Malrotation kidney, Renal fusion anomaly, Reverse rotation kidney

INTRODUCTION

Duplex collecting system is a common anomaly in renal tract development that affects 2-4% of people with urinary tract symptoms and occurs in about 1% of healthy adult populations. 7.2% of individuals with urinary tract abnormalities were included in this.[1] Kidney malrotation is a rare case, more commonly present in men than women. Prevalence of renal malrotation is 1 in 2000 autopsies.[2] The incidence of renal malrotation in a separate study was found in 1 in 939 cases. Of these the lateral also known as reverse rotated kidney is the rarest type.[3] The combination of a duplex collecting system and renal malrotation is a rare case. Our case is the third case from the available literature.[4,5] We aim to describe this case and provide the most recent information from the literature.

CASE REPORT

A 33-year-old male presented to our outpatient urology department with the chief complaint of unspecific right flank pain which was on and off for about a week. He had no history of kidney disease. Vital signs and physical examinations were within normal limits. Laboratory tests revealed no significant changes in complete blood count and urinalysis. Blood urea was 22.3 mg/dl (20-40 mg/dl), creatinine serum was 0.59 mg/dl (0.6-1.2 mg/dl), and uric acid was 5.89 mg/dl (3.4-7.0 mg/dl). Ultrasonography of urinary tract revealed multiple calcifications in right pelvic calyceal system. There were no abnormalities in left kidney.

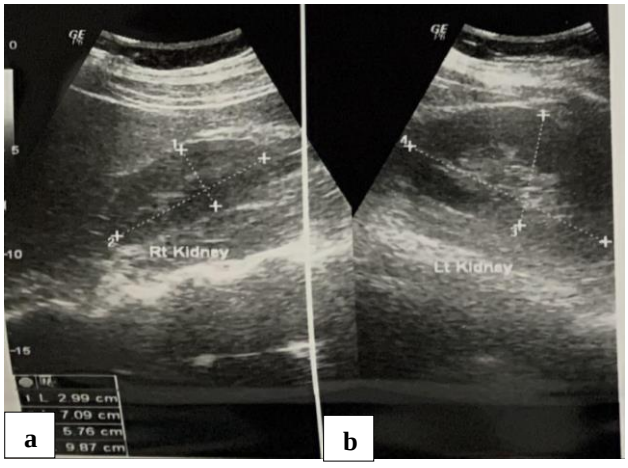

Figure 1: Ultrasonography of the kidneys (a) right kidney, and (b) left kidney.

He was thus sent to have another radiology examination, a non-contrast computed tomography (CT) urogram. It revealed that the right kidney had a double collecting system with proximal ureter fusion (bifid pelvis), accompanied by rotational abnormalities. The hilum is facing laterally and the vessels are anterior to the kidney, consistent with reversed kidney rotation. Left kidney was normal. No therapy was given, and he was followed up every 3-6 months to evaluate the kidney function.

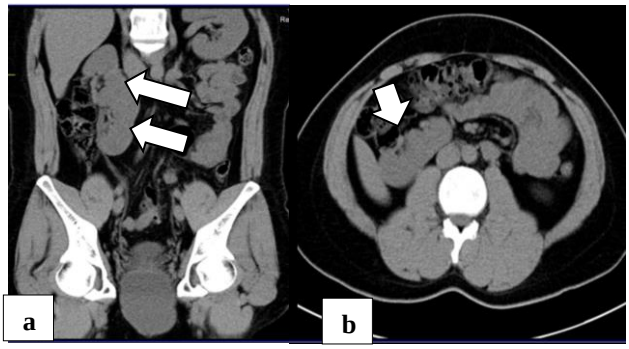

Figure 2: CT urography (a) coronal view: upper pole moiety (thin white arrow), lower pole moiety (thick white arrow); (b) axial view: malrotation of right kidney (white arrow).

DISCUSSION

Duplex collecting system can be defined as two separate pyelocaliceal systems in one renal unit. This condition is the most common congenital abnormality of the urinary tract.[1] Most patients are asymptomatic throughout their lives, but it can also cause conditions like vesicoureteral reflux (VUR), incontinence, ureterocele or obstructive uropathy as well as renal parenchymal scarring or dysplasia and decreased renal function.[6]

During fetal development, single ureteric bud arises from the mesonephric (Wolffian) duct that migrates to the metanephros (precursor of the kidney). Ureteric bud induces metanephros to form nephrons. In turn, the metanephros induces the ureteric bud to form the renal collecting system including the calyces and renal pelvis. Urinary tract anomalies occur when there is a signaling error between ureteric bud and the metanephros. Researchers have shown that overexpression of the glial cell-derived neurotrophic factor (GDNF)-RET. signaling pathway can induce the formation of multiple ureteral buds which cause duplication of ureter and/or collecting system.[6-8]

Duplex collecting system is classified into incomplete and complete. An incomplete duplex system is one in which the ureters unite before the ureteropelvic junction. The incomplete duplex system is divided into bifid pelvis which is a partially duplicate ureteral pelvis (Y-shaped ureter) and bifid ureter which is an incomplete duplication of ureter with the ureter joining near or at the wall of the bladder (V-shaped ureter). Whereas, complete ureteric duplication with separate ureteric orifices.[9] In our case, the duplex collecting system is classified as an incomplete duplex system with proximal ureter fusion (bifid pelvis).

Abnormal renal rotation, also called renal malrotation, is a variation in the anatomical position of the kidney, especially an anomaly in the orientation of the renal hilum. Renal malrotation usually causes no symptoms.[10]

Normally, the hilum of the kidney faces anteromedial. Malrotation of the kidney occurs in the embryogenesis phase. The kidneys develop from the metaphros which lies in the sacral region and the hilum faces anteriorly. In the sixth week, the kidneys begin to migrate upward and simultaneously rotate 90 degrees toward the midline. In the 9th week, the kidney will be in a lumbar position with the hilum facing medially and slightly ventrally.[10]

Renal malrotation can be divided into several types based on the degree of rotation. The first one is ventral position. There is no rotation at all thus causing the hilum faces ventrally. Ventromedial position or incomplete rotation is when the pelvis faces ventromedially because of an incompletely rotated kidney. Rotation stops during the seventh week of gestation when the kidney and pelvis normally reach this position. Dorsal position or hyper rotation is when renal excursion of 180 degrees occurs to produce this rarest position. The pelvis is dorsal to the parenchyma, and the vessels pass behind the kidney to reach the hilum. The lateral position is when the pelvis faces laterally and the renal parenchyma is medial. There are two types of this position based on its rotation direction. Hyper rotation when kidneys rotate between 180 degrees and 360 degrees and reverse rotation when they are reversed up to 180 degrees. The renal vascular supply can be the clue of the kidney's rotation direction. If the vessels are located anteriorly from the kidney, it's suggested reverse rotation, and if the vessels are located posteriorly, it's suggested excessive ventral rotation. In this patient, the renal hilum faces laterally with vascular places anteriorly, so it is classified into reverse rotation type.[10,11]

The relation between renal malrotation and duplex collecting system is not yet clear and their association is very rare. To the best of our knowledge, this is the third case describing this rare combination of duplex collecting system with reverse malrotation in an adult[4,5] Both of these anomalies can be asymptomatic and only discovered incidentally on imaging. No therapy is needed as long as kidney function is normal and no obstruction of the urine flow.[12]

CONCLUSION

The number of combinations of duplex collecting system and reverse rotation kidney case is still very rare. Urinary tract anomaly, there are no signs and symptoms and there are no complications so the patient is not given therapy. We recommend patients with congenital kidney anomalies

to be followed up every 3-6 months to evaluate the kidney function.

This illustrates a case where fortuitous anomalies have resulted in a normal quality of life in a patient with combined renal developmental anomalies which otherwise would have posed deleterious renal complications.

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