

Case Report

Renal Cell Carcinoma in a Right Malrotated Kidney

Tae Joon Lim, Seung Kwon Choi, Hyun Wook You, Myung Joon Kim, Jae Sung Ahn, Tae Gu Kim, Jin Ill Kim, Seung Hyun Jeon

Department of Urology, Kyung Hee University Medical Center, Seoul, Korea

The authors report a case of renal cell carcinoma in a right malrotated (horizontal axis) kidney. The patient was treated by hand-assisted laparoscopic radical nephrectomy. This is the first report of a horizontal axis malrotated kidney with renal cell carcinoma.

Key Words: Carcinoma, Renal cell; Malrotation; Nephrectomy

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Corresponding Author:

Seung Hyun Jeon
Department of Urology, Kyung Hee
University Medical Center, 1,
Hoegi-dong, Dongdaemun-gu, Seoul
130-702, Korea
TEL: +82-2-958-8537
FAX: +82-2-959-6048
E-mail: juro@chol.com

Kidney malrotation is found in 1 of 939 autopsies and is frequently observed in patients with Turner's syndrome. Most often, kidney malrotation is found in conjunction with another renal abnormality, such as ectopia or a horseshoe kidney. In most malrotation cases, the malrotation occurs ventrally, ventromedially, dorsally, or laterally. As far as we know, there are no reports of kidney malrotation with a horizontal axis in conjunction with renal cell carcinoma (RCC). The authors hereby present a case of RCC in a horizontal axis malrotated kidney that was successfully treated by hand-assisted laparoscopic radical nephrectomy.

CASE REPORT

A 51-year-old female had palpitations and radiating back pain and underwent whole-aorta angio 3D computed tomography (CT). The CT revealed a horizontal axis right kidney with a 4.2 cm, lobulating contour multilocular cystic mass at the lower pole. There were multiple irregular, enhancing septations with an enhancing solid portion and central calcification in the mass. The renal artery was located ventrally and the renal vein was located dorsally. The renal pelvis was located caudal to the renal vessels (Fig. 1). She was diagnosed as having cystic RCC in the horizontal axis right kidney and underwent hand-assisted laparoscopic radical nephrectomy in September 2010.

Under general anesthesia, the patient was positioned in the left lateral decubitus position. A 7 cm laparoscopic

hand-assist device was placed in the right lower quadrant with the placement of two additional 10 mm ports directly cephalad along the lateral rectus border, allowing 8 cm of separation between the ports and the hand-assist device. Following dissection into the retroperitoneal space, the kidney was separated from the psoas muscle, adjacent bowel, and liver. There was a 4 cm cystic mass in the horizontal axis right kidney (Fig. 2). The ureter was carefully isolated and divided. The renal artery and renal vein were adhered together, so we used an Endo-GIA universal stapler (Covidien Surgical, Dublin, Ireland) and divided them, thereby isolating the entire right kidney. The specimen was entrapped in a laparoscopic sac and extracted. The procedure was completed successfully with a surgical time of 1 hour and an estimated blood loss of 100 ml. Patient-controlled analgesia with morphine was discontinued on postoperative day one. Postoperatively, the patient did not develop any complications. Pathology revealed the tumor to be a clear cell carcinoma (Fuhrman grade 3) with the tumor limited to the kidney (T1a) and with negative resection margins. In December 2010, follow-on CT revealed no recurrence of the disease.

DISCUSSION

The adult kidney, as it assumes its final position in the 'renal' fossa, orients itself so that the calyces point laterally and the pelvis faces medially. When this alignment is not



FIG. 1. Computed tomography of a horizontal axis right kidney (A) and of a renal cell carcinoma in the lower pole of the horizontal axis right kidney (B) and three-dimensional reconstruction of the kidney malrotation (C).

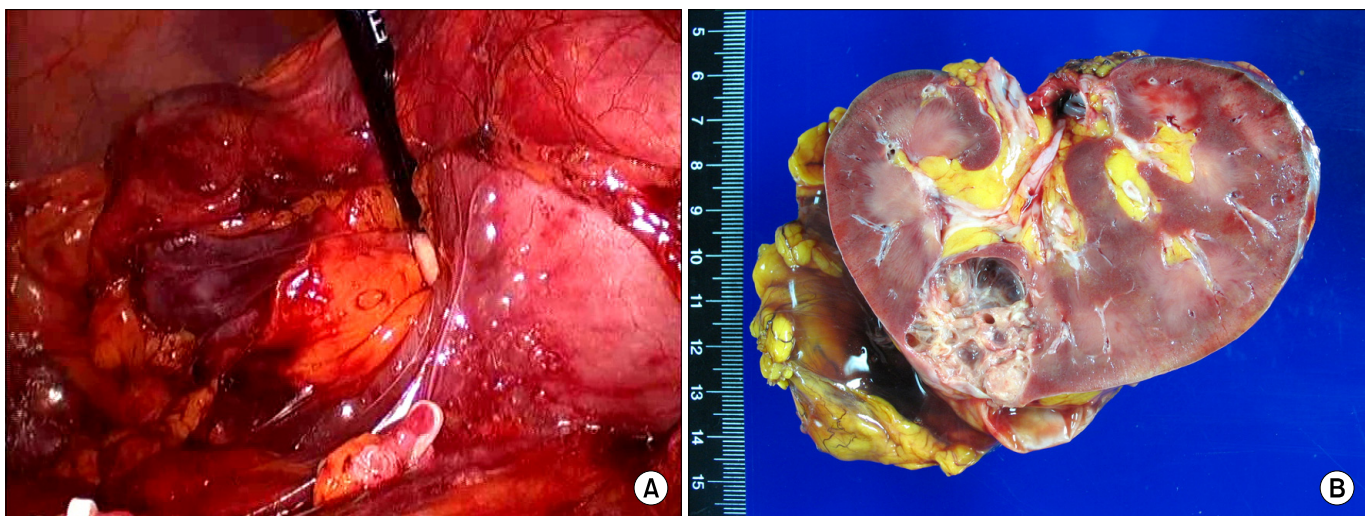


FIG. 2. Operative finding showing 4 cm sized cystic mass in the lower pole of the horizontal axis right kidney (A), gross finding of specimen (B).

exact, the condition is known as malrotation. Most often, the inappropriate orientation is found in conjunction with another renal anomaly, such as ectopia with or without fusion or a horseshoe kidney.

It is thought that medial rotation of the collecting system occurs simultaneously with renal migration. The kidney starts to turn during the 6th week, just as it is leaving the true pelvis, and it completes this process, having rotated 90 degrees toward the midline, by the time ascent is complete at the end of the 9th week of gestation.

It has been postulated that rotation is actually the result of unequal branching of successive orders of the budding ureteral tree, with two branches extending ventrally and one dorsally during each generation or division. Each ureteral branch then induces differentiation of the metanephrogenic tissue surrounding it to encase it as a cap. More parenchyma develops ventrally than dorsally, and the pelvis seems to rotate medially. Weyrauch accepted this theory of renal rotation to be the result of excessive ventral versus dorsal branching of the ureteral tree and concluded that the fault of malrotation lies entirely with the ureter [1]. A

late-appearing ureteral bud may insert into an atypical portion of the renal blastema, leading to a lessened propensity for the developing nephric tissue to shift. Late appearance of the ureteral bud is almost always associated with an aberrant origin from the Wolffian duct; This translates into ureteral ectopia at the level of the lower urinary tract. The renal blood supply does not appear to be the cause or a limiting factor in malrotation but rather follows the course of renal hyporotation, hyper-rotation, or reverse rotation.

There are two divergent opinions in the anatomical literature concerning the definite position of the kidneys. According to the first, the kidney ascends in the retroperitoneal space during precocious ontogenetic development. The renal rudiment occurs in the pelvic region, at the level of the L2 to L3 vertebrae, with the dorsal convex border and the ventral hilum touching the abdominal wall. To place itself in the definite position, the kidney undergoes a process of ascension and rotation. Between the 6th and the 9th week, the kidney ascends to the lumbar region along the dorsal aorta. The exact mechanism is unknown. An in-

ductive substance secreted by the developing kidney may play a role.

The second opinion states that the kidney undergoes a pseudoascension caused by the fast development of the caudal extremity of the fetus [2-4].

In most malrotation cases, malrotation occurs ventrally, ventromedially, dorsally, or laterally. In our case, however, the renal pelvis faced the medial side, but the long axis of the kidney was lying ventral to dorsal. The upper pole and lower pole of the kidney were indistinctive. Therefore, in this case, the process of malrotation likely did not follow the ordinary fashion. It is likely that the kidney rotated cranially or caudally, not medially or laterally.

The ectopic kidney can be located in the pelvis, in the abdomen, or, rarely, in the thorax (0.3%) and can be unilateral or bilateral. The most frequent cases of renal ectopia described in the literature of the field occur in male patients and are located on the right side of the pelvis [5-7]. Generally, an ectopic kidney is smaller, is of irregular shape and variable rotation, and is vascularized by multiple arteries with various levels of origin.

In some cases of ectopic kidney, there is often a bizarre rotation of the kidney as in our case [8-10]. However, the malrotated kidney was located at a similar level to the contralateral kidney and was also located in the retroperitoneal space. The renal vessels were also located at the L2-3 level. Therefore, it was not an ectopic kidney. The process by which this malrotation occurred is not clear, but we can assume that the malrotation occurred during the ascension process or that the axis developed horizontally and proceeded ascension.

We are the first to report RCC in a malrotated kidney. Several cases of renal stones in an ectopic kidney or malrotated kidney have been reported, but no cases of RCC have been reported in an abnormal kidney.

The malrotated, ectopic kidney has clinical significance owing to its atypical location and malrotation. We are the first to report a horizontal axis kidney located in the retroperitoneal space. It is unknown whether the malrotated kidney is vulnerable to RCC development, but we are the first to report RCC in a case of kidney malrotation.

Conflicts of Interest

The authors have nothing to disclose.

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