Case Report

Renal cell carcinoma in a lumbar ectopic kidney

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Abbreviations & Acronyms

CT = computed tomography RCC = renal cell carcinoma

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Introduction: Lumbar ectopic kidney is a rare developmental renal anomaly. We report a case of renal cell carcinoma in lumbar ectopic kidney with an abnormality of the main renal artery piercing the renal parenchyma.

Case presentation: A 70-year-old female was referred to our division with an incidentally diagnosed ectopic kidney. Computed tomography angiography detected the right kidney at the lumbar (L3-L4) level with an early-enhanced 44-mm-diameter mass. The kidney had a laterally facing renal hilum and renal arteries piercing the renal parenchyma from the medial side. An open radical nephrectomy was performed using a peritoneal approach, and no perioperative complications were observed. The pathological diagnosis was clear cell carcinoma (pT1).

Conclusion: This is the first case report of renal cell carcinoma in a lumbar ectopic kidney. It highlights the importance of preoperational imaging for aberrant artery and careful surgical management.

Key words: ectopic kidney, lumbar kidney, renal artery, renal cell carcinoma, renal ectopia.

Keynote message

RCC in ectopic kidney involves abnormalities in the renal vessels. This case reports a lumbar ectopic kidney with the main renal artery piercing the renal parenchyma. The case depicts rare vessel abnormalities in ectopic kidneys and emphasizes the importance of preoperative imaging.

Introduction

Ectopic kidney is a relatively common congenital disease, with an incidence of approximately 1 in 1000 autopsies. Letopic kidneys below the diaphragm are classified into abdominal, lumbar, or pelvic based on their position. Among these, lumbar kidney is comparatively rare, with a reported incidence of 12% in all ectopic kidneys.

RCC occurring in ectopic kidneys has been previously reported in the literature. While RCC in pelvic ectopic kidneys has been described in approximately 10 cases, 3-12 RCC in lumbar ectopic kidney has not been previously reported.

We report a case of RCC in a lumbar ectopic kidney, including a rare abnormality of the main renal artery piercing the renal parenchyma. This case required careful interpretation of preoperational imaging and surgical management.

Case presentation

A 70-year-old female was referred to our division with a right ectopic kidney incidentally diagnosed via magnetic resonance imaging for chronic lumbago. The patient had no remarkable family or medical history, including no abnormal development or pregnancy and delivery complications. A fist-sized mass was palpable at the right side of the naval, and an ultrasound examination detected a heteroechoic mass.



Fig. 1 Arterial phase CT images at diagnosis. (a) Transverse section: an early-enhanced 44-mm-diameter mass (yellow arrow) was detected. (b) Sagittal section: the tumor (yellow arrow) was located in the lower pole of the right kidney at the lumbar (L3-L4) level. (c) Coronal section: the renal hilum (yellow arrow) faces the lateral direction.

CT detected the right kidney located at the lumbar (L3-L4) level and an early-enhanced 44-mm-diameter mass at its lower pole (Fig. 1a,b). The renal hilum of the kidney faced the lateral direction (Fig. 1c). CT angiography indicated that the largest artery originated from the abdominal aorta and the two smaller arteries originated from the left and right common iliac arteries (Fig. 2a). In a horizontal CT section, the largest artery appeared to pierce the renal parenchyma from a medial to lateral direction (Fig. 2b). Figure 2c shows a preoperative sketch that presumed the perirenal anatomy based on CT images. The patient was diagnosed with RCC of clinical stage cT1bN0M0.

An open radical nephrectomy with a peritoneal approach was performed. An abdominal incision was made and the omentum was moved aside, promptly identifying the main renal vein on the surface of the renal parenchyma (Fig. 3a). The two small renal arteries originating from the iliac arteries were resected (Fig. 3b,c), and the largest renal artery that pierced the renal parenchyma was ligated (Fig. 3d). The renal vein was resected, followed by resecting the main artery. The tumor did not adhere to adjacent organs. The operating time was 1 h and 57 min and blood loss was 20 mL.

The resected surgical specimen included the renal vein, main renal artery, tumor, and ureter and was consistent with CT images (Fig. 4a). The main renal artery (Fig. 4b) and the small artery from the left iliac artery (Fig. 4c) macroscopically

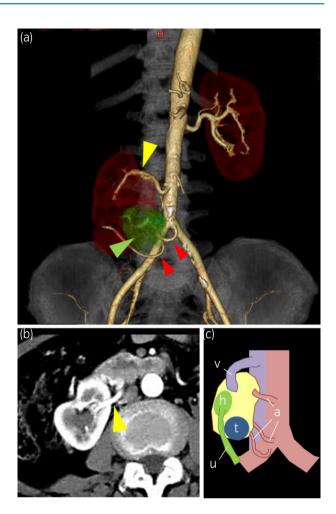


Fig. 2 CT angiography image and additional information on renal arteries. (a) An image of CT angiography: the aberrant main renal artery (yellow arrow), originating from the aorta, appeared to overlap with the renal parenchyma. The two smaller arteries (red arrows) originated from the left and right common iliac arteries. The renal tumor (green arrow) was located at the level of the aortic bifurcation. (b) A transverse CT section: the main renal artery pierced the renal parenchyma (yellow arrow) and bifurcated in the renal medulla. (c) A preoperative sketch that assumed the perirenal anatomy, which was consistent with the actual anatomy. The renal vein, renal arteries, renal hilum, tumor, and ureter are represented by v, a, h, t, and u, respectively.

pierced the renal parenchyma. The tumor was lipid abundant and surrounded by a clear boundary of normal parenchyma (Fig. 4d). The pathological diagnosis was clear cell carcinoma (pT1a, G2, INFa, v0, ly0) (Fig. 4e). No postoperational complications were observed, and the patient experienced no recurrence for 2 years post-surgery.

Discussion

We reported a case of RCC occurring in a lumbar ectopic kidney. Although incidences of ectopic kidneys are reported in 1:500 to 1:1200 autopsies, the clinical recognition is estimated to be only 1 in 10 000 autopsies because of vague symptoms. Among several types of ectopic kidneys, lumbar kidney is relatively rare. The incidence of lumbar kidney (12%) in all ectopic kidneys is reported to be lower compared with that

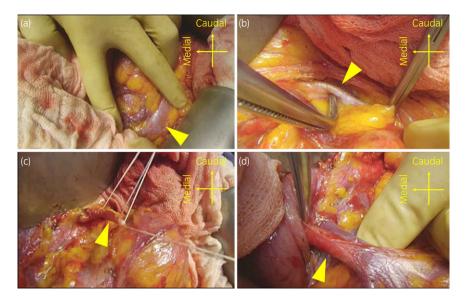


Fig. 3 Gross images of the operation. The direction of the patient's caudal and medial side is shown in each panel. (a) The renal vein (yellow arrow), which existed on the surface of the upper pole of the kidney, was identified after abdominal incision. (b) The small renal artery (yellow arrow) originating from the right iliac artery. (c) The small renal artery (yellow arrow) originating from the left iliac artery. (d) The largest main renal artery (yellow arrow) originating from the artery.

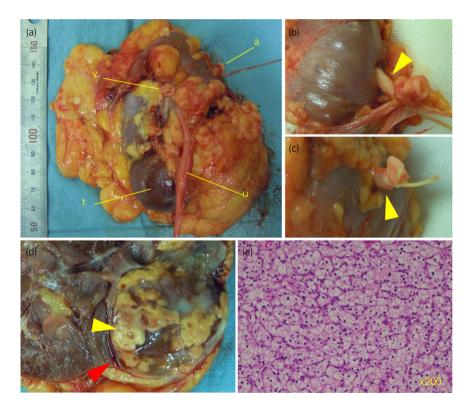


Fig. 4 Gross and microscopic images of the resected specimen. (a) The resected right kidney, the renal vein, main renal artery, tumor, and ureter are represented by v, a, t, and u, respectively. (b) The main renal artery (yellow arrow) macroscopically pierced the renal parenchyma as seen in the CT images. (c) The smaller renal artery, originating from the left iliac artery (yellow arrow), also pierced the renal parenchyma. (d) The tumor was lipid abundant (yellow arrow) and surrounded by a clear boundary (red arrow) of normal parenchyma. (e) Microscopic pathology revealed typical clear cell carcinoma presenting proliferated malignant epithelial cells with abundant clear cytoplasm (Hematoxylin-eosin, ×200).

of pelvic kidney (55%) and cross-fused kidney (27%).² The incidence of RCC in ectopic kidneys is not different from that in normal kidneys.¹ According to the literature review, this case is considered the first to report RCC in a lumbar kidney.

To prepare for nephrectomy, we referred to former cases of RCC in ectopic kidney which presented with a variety of anatomical abnormalities. A case of RCC in pelvic kidney with bilateral iliac vein invasion was reported, in which the kidney and thrombus were successfully removed using intraoperative ultrasound. ¹¹ Another case alarmed that absence of Gerota's fascia in a pelvic kidney may cause adjacent organ involvement. ⁸ In the present case, we identified the right

renal vein on the surface of the upper pole of the kidney soon after skin incision (Fig. 3a). This abnormality may have been caused by the lateral facing of the renal hilum (Fig. 1c) and a scarce amount of Gerota's fascia (Fig. 3a).

In this case, the main renal artery pierced the renal parenchyma from the opposite side of the renal hilum. This phenomenon has not been recorded in former cases of RCC in ectopic kidney.^{3–12} We hypothesized that this rare abnormality of the renal artery is associated with the etiology of lumbar ectopic kidney. Ectopic kidney is caused by the trapping of the kidney in its ascent from the sacral level during fetal development.¹ The kidney rotates and the hilum shifts from

the anterior position to the medial during the ascent.¹³ The present case showed the laterally malrotated kidney that is a rare type of malrotation involving reverse rotation.¹ The laterally malrotated kidney may have been arrested in the ascending process which prompted the main artery to develop through the parenchyma. This type of anomaly may provide new insight into the poorly understood mechanisms that govern the development of the kidney vasculature.¹⁴

Lastly, the possibility of partial and/or laparoscopic nephrectomy in ectopic kidneys can be discussed. A former case has performed a partial nephrectomy for RCC in pelvic kidney. If the present case required nephron sparing, we may have selected a partial nephrectomy with careful identification of abnormal arteries feeding the tumor. Abnormal anatomy of the renal parenchyma and calyces may have an increased risk for urinary fistula. Another case reported a laparoscopic nephrectomy for RCC in pelvic kidney. We did not select a laparoscopic surgery due to the difficulty in preoperatively understanding the vessel abnormalities and the risk for longer ischemic time.

Conclusion

We are the first to report a case of RCC in lumbar ectopic kidney with an aberrant main renal artery that pierced the renal parenchyma. This case highlights the importance of careful evaluation with preoperative imaging in ectopic kidneys.

Acknowledgments

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Conflict of interest

The authors declare no conflict of interest.

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