







Renal cell carcinoma in ectopic-pelvic kidney: A rare case with review of literature

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ABSTRACT

The incidence of renal cell carcinoma in a pelvic kidney is rare, and has only been reported in a small number of cases. We report a 50-year-old male patient presented with vague abdominal pain and without hematuria. CT scan showed large heterogeneous soft tissue mass arising from a left pelvic kidney with areas of necrosis measuring 8.4x8.4x7.5 cm. Histopathology after radical nephro-ureterectomy showed grade II clear-cell renal carcinoma. Renal cell carcinoma of ectopic kidney is a rare disease. Even though the presentation might be atypical and challenging, the treatment strategy is still the same as for tumors of orthotopic kidneys.

Keywords: Ectopic kidney; pelvic ectopia; pelvic kidney; renal cell carcinoma; renal mass.

Introduction

Renal Cell Carcinoma (RCC) is the most common solid tumor affecting adults, accounting 2% of the worldwide and in Europe.^[1] The diagnosis of RCC can be challenging as only almost 10% of the patients present with the classic triad of flank mass, flank pain and hematuria and thus it may present with diverse manifestations including weight loss, general weakness, malaise, hypertension and hypercalcemia. Renal tumors of ectopic kidney are extremely rare. We report a case of a left pelvic kidney with a large renal cell carcinoma diagnosed in a patient presented with abdominal pain but without hematuria.

Case presentation

A 50-year-old male smoker known to have hypertension well controlled with valsartan/hydrochlorothiazide (80/12.5 mg) presented to the outpatient clinic in June, 2016 complaining of vague abdominal pain of two weeks duration. He described a discomfort in the lower abdomen which was not associated with nausea, vomiting or hematuria. General examination showed a male patient with BMI of 23.4 kg/m² (weight: 80 kg, height 1.85 m) and abdominal examination revealed a palpable

mass in the lower abdomen. Routine laboratory workup, including complete blood count, renal, and liver function test results, and electrolytes were within normal range and urinalysis showed RBCs of 1-2 per HPF. Ultrasonography of the abdomen and pelvis showed left pelvic kidney with a large renal mass. Contrast-enhanced CT scan of abdomen and pelvis showed large heterogeneous soft tissue mass arising from the left pelvic kidney with areas of necrosis measuring 8.4x8.4x7.5 cm demonstrating significant corticomedullary phase enhancement with no metastasis. (Figures 1, 2). The vascular supply was provided from a single left renal artery arising from the left common iliac and a single left renal vein draining into the left common iliac vein.

After a multidisciplinary team discussion and the patient's informed consent, the patient underwent radical left pelvic nephro-ureterectomy via transperitoneal approach in July, 2016. The post-operative period was unremarkable. Specimen showed a unifocal tumor limited to the kidney and histopathology revealed clear-cell renal carcinoma grade II with negative margins. TNM staging was pT2 Nx M0 (Figure 3). The patient went abroad and he was instructed to attend his follow-up visits in the country of his residence.

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Table 1. Review of cases from the literature

| Reference, year | Age, Gender | Presentation | Comorbidities | Side | Vascularity | Surgery | Histopathology | Metastasis | Follow up |
|---|-------------|---|----------------------------|-------|---|-----------------------------------|-------------------------|----------------------------------|---|
| Fischer et al. ^[3] , 1999 | 48, Male | Hematuria | None | Right | Single renal artery originating from the common iliac artery. Single renal vein draining into the right external iliac vein | Open radical nephrectomy | RCC, Clear, Fur 1/4 | Tumor thrombus in the renal vein | Not reported |
| Terrone et al. ^[4] , 2004 | 52, Male | Abdominal pain | Not reported | Left | NA | Open radical nephroureterectomy | RCC, Fur 4/4 | No | 9 months after surgery the patient had no recurrence |
| Karaman et al. ^[5] , 2009 | 66, Male | Weight loss, abdominal pain and palpable mass | NA | Left | Renal artery originating from left common artery | Nephrectomy | RCC, Fur 3/4 | No | The patient is under control program without any evidence of local recurrence or metastasis for 24 months. |
| Mahmoudnejad et al. ^[6] , 2009 | 53, Female | Abdominal pain | Not reported | Left | Two renal arteries originating from left internal iliac artery. Single vein identified | Open radical nephrectomy | RCC, Clear, Fur 2/4 | No | Within postoperative first year the patient underwent physical examinations and metastatic work up and no delayed complications occurred. |
| Dash et al. ^[7] , 2010 | 55, Male | Acute urine retention and weight loss | Posterior cortical atrophy | Left | NA | No surgical intervention was done | RCC (post mortem) | No | Patient did not have surgery and died four months after diagnosis |
| Baldie et al. ^[8] , 2012 | 61, Male | Hematuria and irritative symptoms | Morbid Obesity | Left | Two renal arteries originating from left common iliac artery. Vein drains into left internal iliac | Open radical nephroureterectomy | RCC, Papillary, Fur 3/4 | No | Patient is on dialysis and recurrence-free for 4 years since surgery |
| Nowroozi et al. ^[9] , 2015 | 53, Female | Hematuria | None | Left | NA | Open nephrectomy | RCC, Clear, Fur 4/4 | No | No local recurrence was detected for 18 months after surgery |
| Parashari et al. ^[10] , 2015 | 45, Male | cervical spondylitis myopathy (cervical metastasis) | Smoking | Left | Arterial supply from the left common iliac artery | No surgical intervention was done | NA | Vertebra | Not reported |

Table 1. Review of cases from the literature (continued)

| Reference, year | Age, Gender | Presentation | Comorbidities | Side | Vascularity | Surgery | Histopathology | Metastasis | Follow up |
|--------------------------------------|-------------|-----------------|---------------|-------|--|---|--|--|--|
| Tanwar et al. ^[11] , 2016 | 61, Male | Gross hematuria | Not reported | Right | Single renal artery. Two veins draining into the right internal iliac and left common iliac | Radical nephroureterectomy with thrombus extraction | RCC, Clear with sarcomatoid differentiation, Fur 3/4 | Tumor thrombus invading both iliac veins | The patient is on surveillance for renal carcinoma and is following the protocol strictly. |
| Present case, 2016 | 50, Male | Abdominal pain | Smoking | Left | Single renal artery origination from the left common iliac artery. Single vein draining into the left common iliac | Radical nephroureterectomy | RCC, Clear, Fur 2/4 | No | The patient went outside the country and is to be followed up in the country of his residence. |

NA: not available



Figure 1. Contrast-enhanced abdominopelvic CT, coronal view showing a large heterogeneous soft tissue mass with dimensions of 8.4x8.4x7.5 cm arising from left pelvic kidney



Figure 2. Contrast-enhanced abdominopelvic CT, Axial view showing a large heterogeneous mass with central necrosis arising from left pelvic kidney

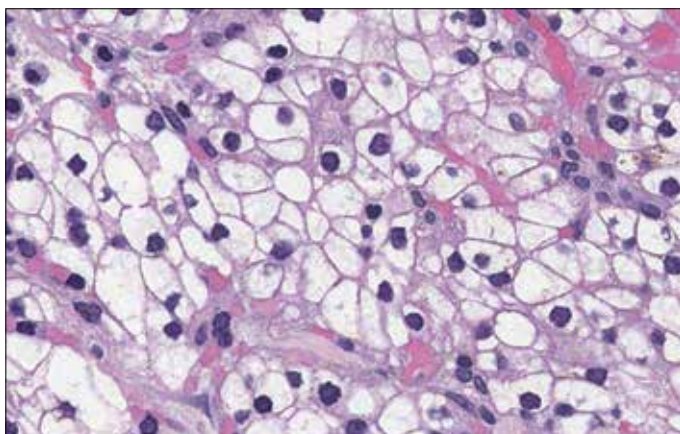


Figure 3. Clear-cell renal cell carcinoma

Discussion

Ectopic pelvic kidney is a rare developmental anomaly. During embryological life metanephros from which the kidney originates is located in the sacral region. During development, both kidneys ascend to their usual location in the retroperitoneal area. Early arrest or failure of ascent results in ectopic kidney.^[2]

An ectopic kidney is mostly discovered incidentally during investigation for other reasons. However, it may be associated with other anomalies involving other systems including skeletal, urinary and cardiovascular systems.^[2]

The average incidence of pelvic kidney ranges between 1/2200 and 1/3000. The relationship between malignancy and ectopic kidneys is still unclear. To our knowledge, three cases of urothelial carcinoma and nine cases of renal cell carcinoma (RCC) in ectopic pelvic kidneys have been reported in the literature and there does not appear to be an increased risk of malignancy in ectopic kidneys. These cases are summarized in Table 1.^[3-11]

The rarity of such cases in daily clinical practice made it a challenge for the urologists. Most cases of ectopic kidneys present with atypical presentations. However, the existence and availability of modern radiological imaging techniques increased the detection rates of such anomalies over the years.

The only reported masses in pelvic kidneys are renal cell carcinomas and upper tract urothelial carcinomas, so the suggested treatment options for tumors in pelvic kidneys are the same as tumors in orthotopic kidneys. The treatment modality and surgical approaches depend on the site, the clinical stage of malignancy, comorbid conditions and the surgeon's experience. Radical nephro-ureterectomy appears to be the standard treatment for localized tumors in ectopic kidneys.

The vascular supply of pelvic kidneys is complex and it is provided from the adjacent blood vessels. So the surgical approach to ectopic kidneys merits caution because of uncertain vascular anatomy.^[6] It is mandatory to perform a detailed preoperative vascular evaluation, and also careful exploration during surgery in order to avoid vascular injury. Follow-up is required to detect any metachronous renal tumors, local recurrence and distant metastases.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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R.K.A., T.Y.L.; Literature Search - R.K.A., H.M.G.; Writing Manuscript - R.K.A., H.M.G., T.Y.L.; Critical Review - T.Y.L., R.K.A., H.M.G.

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