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Case Report

Pelvic Nephroureterectomy for Renal Cell Carcinoma in an Ectopic Kidney

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We present a case of an ectopic renal tumor in a 61-year-old morbidly obese man with a pelvic kidney found after presenting with hematuria and irritative voiding symptoms. The mass, along with the ectopic kidney and ureter, was radically resected through an open operation that involved removing both them and the renal vessels from the underlying iliac vessels. Pathological analysis demonstrated an 8.3 cm papillary renal cell carcinoma (RCC) with oncocytic features, Fuhrman nuclear grade 3, with angiolymphatic invasion and negative margins. The patient has been recurrence-free for over four years since tumor resection.

1. Introduction

Renal ectopia is a rare condition involving a failure of the mature kidney to reach its normal location within the renal fossa. This congenital anomaly comprises about 0.001% of all autopsies. Pelvic, iliac, abdominal, thoracic, contralateral, and crossed ectopic kidneys can occur. Apart from the development of hydronephrosis and urolithiasis, the ectopic kidney is no more susceptible to disease than the normally positioned kidney [1]. Nevertheless, when considering surgical resection, particularly with the intent to remove a tumor, an accurate understanding of the surrounding anatomy is crucial in order to avoid causing both unnecessary damage to blood vessels and leaving remnants of tumor within the patient.

2. Case Report

The patient presented as a 61 year old morbidly obese male with a history of chronic kidney disease describing three months of gross, painless hematuria and irritative lower urinary tract symptoms. The remainder of the review of systems was unremarkable. He initially went to an outside hospital after his symptoms were refractory to antibiotic management where he was found on noncontrast CT to have a large left renal mass arising from an ectopic pelvic kidney. Mercaptacetyltsiglycine-3 (MAG3) renography was performed at our institution following referral, showing an essentially nonfunctional left kidney and delayed perfusion, uptake, and excretion in the contralateral kidney. Two weeks later, the patient was further worked up with a complete history and physical, labs, and abdominal MRI with and without contrast. Physical exam and labs were unremarkable except for a creatinine of 2.2. MRI showed an 8.6 cm centrally necrotic, peripherally enhancing solid mass in the superior-interpolar region of the ectopic left kidney worrisome for renal malignancy. The superior margin of the mass rested anterior to the left common iliac vessels (Figure 1). Two anomalous renal arteries supplied the kidney, with the proximal artery branching off of the distal aorta and the distal artery branching of the left common iliac artery (Figure 2). The left renal vein drained into the left internal iliac vein (Figure 3).
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Figure 1: T2-weighted MRI image (coronal view) showing orientation of mass (star) to major vessels. Normal kidney (diamond). Aorta (short arrow). IVC (long arrow). Note that aortic bifurcation occurs at level of superior pole of ectopic left kidney.

Figure 2: Arterial Phase T1-weighted MRI image (transverse view) showing anomalous arterial supply to ectopic left kidney and mass (star). Proximal left renal artery (short arrow) branching off distal aorta. Distal left renal artery (long arrow) branching off left common iliac artery.

Figure 3: Venous Phase T1-weighted MRI image (transverse view) showing anomalous venous drainage from ectopic left kidney and mass (star). Left renal vein (short arrow) draining into left internal iliac vein (long arrow).

The patient was thoroughly discussed at our institution’s weekly genitourinary (GU) preoperative conference with all urology physicians present and, after weighing the risks and benefits, the decision was made for definitive surgical management. The following week, the patient underwent an open transperitoneal radical left pelvic nephroureterectomy. Cystourethroscopy at the onset of the procedure was done to ensure patency of the contralateral ureteral orifice. A midline incision was made from the level of the pubic symphysis to the xiphoid. The space of Retzius was developed bilaterally until there was visualization of the external iliac vessels. After entering the retroperitoneum and transecting and ligating the proximal and distal urachus, the bladder was dissected down to the level of the vas deferens to aid in visualization. The sigmoid colon was medially displaced and the left colon mobilized medially in order to better access the retroperitoneum. A renal artery was draped over the anterior portion of the kidney, originating from the left common iliac artery. The renal vein took a medial course to reach the left common iliac vein. The kidney was carefully dissected superiorly and inferiorly until the ureter was visualized and tagged. The tumor and kidney were fibrotically adhered to the left common iliac artery, and dissection was done in the midst of confirming palpable lower extremity pulses before and after this stage in the operation. The left renal artery and vein were transected with good hemostasis. The ureter was then clipped distally to the level of the intramural ureter, transected and removed from the pelvis along with the remainder of the specimen (Figure 4) prior to irrigation and layered closure of the peritoneum and abdominal wall. There were no intraoperative complications and the patient tolerated the procedure well. After a low grade fever self-resolved on postoperative day three, the patient progressed as expected and was discharged from the hospital on postoperative day seven shortly following removal of his urethral catheter. Pathology revealed an 8.3 cm papillary renal cell carcinoma (RCC) with oncocytic features, Fuhrman nuclear grade 3, with angiolymphatic invasion and negative ureteral, renal vein, and renal artery margins (Figure 5). The patient was followed up with yearly imaging and, although he was eventually placed on hemodialysis due to end stage renal disease, he has been recurrence-free for over four years since tumor resection.

3. Discussion

There is a dearth in the literature discussing renal malignancy in the ectopic pelvic kidney [2, 3]. The advent of improved imaging studies has allowed for a higher incidence in the identification of ectopic kidneys, including those with malignancy. Despite this fact, these patients can still be misdiagnosed, particularly with atypical presentations as in
Figure 4: Coronal cut of pelvic/ectopic kidney showing a fairly well circumscribed 8.3 × 6.9 × 6.8 cm reddish-brown markedly hemorrhagic mass (star), consistent with papillary renal cell carcinoma. Other structures include normal renal parenchyma (diamond), ureter (narrow arrow), and left renal artery (thick arrow).

Figure 5: Papillary renal cell carcinoma with oncocytic features (hematoxylin and eosin stain, high magnification). Note distinct papillae with eosinophilic cytoplasm (bottom arrow), with fibrovascular core (top arrow).

This report describes a case of successful surgical management of a pelvic kidney with cancer. Imaging was carefully analyzed to determine the orientation of the tumor, kidney, renal vessels, and collecting system, and their association with the underlying major vascular structures enabling the surgeons to avoid significant blood loss during dissection of the affected tissue. Negative margins were acquired and close clinical and radiographic follow-up confirmed appropriate surgical technique while helping ensure long-term patient success.

References
