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Renal Cell Carcinoma of Pelvic Kidney with Atypical Nodal Metastasis: A Diagnostic Challenge

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ABSTRACT

Ectopic pelvic kidney is a known congenital anomaly; however, the presence of renal cell carcinoma (RCC) in an ectopic kidney is rare with the evidence available in the form of a few case reports only. In this case report, we present a case of metastatic RCC in the pelvic kidney which became a diagnostic challenge because of atypical contrast-enhanced computed tomographic (CT) characteristics and unusual pattern of lymph node involvement including cervical lymph node in the absence of visceral metastasis. Because of its unusual location and uncertain vascular anatomy, ectopic kidney poses a surgical challenge. Owing to the rarity of this condition, optimal surgical approach, metastatic potential, routes of metastasis, and effectiveness of systemic agents in pelvic RCC compared to RCC in a normally located kidney, are largely unknown.

Key Words: *Renal cell carcinoma, Pelvic kidney, Lymph node metastasis.*

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INTRODUCTION

Renal ectopia is a known congenital anomaly where the kidney fails to ascend to its usual adult position. The most common site of ectopia is the pelvis, seen in 1 of 2,100 to 3,000 autopsies with the left side favoured slightly over the right.¹ However, renal cell carcinoma (RCC) in the pelvic kidney is a very rare phenomenon with the evidence available in the form of a few case reports only.²⁻⁹ RCC is notorious for being an incidental diagnosis and for atypical presentation following uncharacterised pathways.¹⁰ We herein present a case of RCC in a pelvic kidney with an atypical pattern of nodal metastasis.

CASE REPORT

A 48-year male with no prior co-morbidities presented to the outpatient department of our hospital with lower abdominal swelling, generalised abdominal pain, and weight loss for 1 month. On general physical examination, he had bilaterally palpable, firm, matted cervical and supraclavicular lymph nodes (Levels II to V); however, no axillary or inguinal lymph nodes were found. On abdominal examination, he had a palpable infra-umbilical, non-tender, firm mass of approximately 15×15 cm with a smooth surface and ill-defined margins.

He was a clerk by profession with no history of addictions and negative family history of malignancy or tuberculosis. He had normal complete blood count (CBC) and renal functions. On presentation, he had a plain computed tomography (CT) scan with him, in which, he was reported to have an absent left kidney with bulky psoas muscle and bilateral para-aortic nodal masses (6×10 cm). Hence, his contrast CT scan abdomen and pelvis was performed which showed the left pelvic kidney superolateral to the urinary bladder with a large 101×90 mm cystic lesion arising from the upper pole containing solid enhancing component along its left lateral wall and retroperitoneal lymph nodes. The right kidney was normal (Figure 1).

Tuberculosis, lymphoma, RCC, Upper tract transitional cell carcinoma, germ cell tumor, and colorectal cancers were the major differential diagnoses. His sputum Acid Fast Bacilli (AFB) cultures and Gene Xpert were negative. He underwent a CT-guided biopsy of paraaortic lymph node masses, which showed a neoplasm composed of cords and tubules of polygonal cells exhibiting minimal nuclear atypia and moderate pale cytoplasm with occasional mitoses. The cells demonstrated focal cytokeratin (CK) CAM 5.2 and CKAE1/AE3 expression suggesting epithelial differentiation. However, immunostains such as Epithelial membrane antigen (EMA), CK 7, and CK 20 were negative. Metastasis from prostate and kidney were excluded by the negativity of immunostains such as Prostate-specific antigen (PSA) and Renal cell carcinoma (RCC). Neuroendocrine markers, synaptophysin and chromogranin A, germ cell marker, SALL4 and vascular marker, Cluster of differentiation 34 (CD34), were also performed and were negative. Hence, a diagnosis of a neoplastic lesion with epithelial differentiation which cannot be further characterised despite extensive stain, was given.

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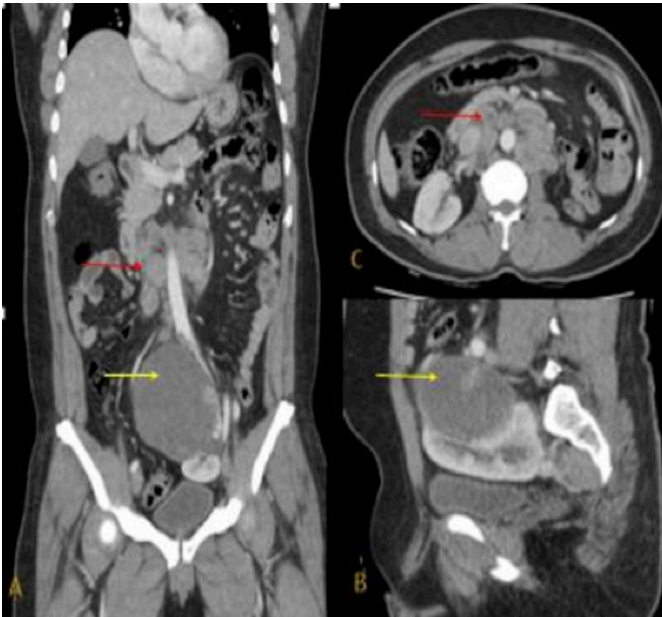


Figure 1: (A & B) Left ectopic kidney lying in pelvis supero-lateral to urinary bladder with a large cystic lesion (101×90 mm in size) arising from upper pole containing a solid enhancing component along its left lateral wall (yellow arrows). Left renal artery and vein both arising from left internal iliac vessels. (C) Grossly enlarged heterogenous enhancing lymph nodal deposits in the para-aortic, aortocaval and preaortic region (red arrows).

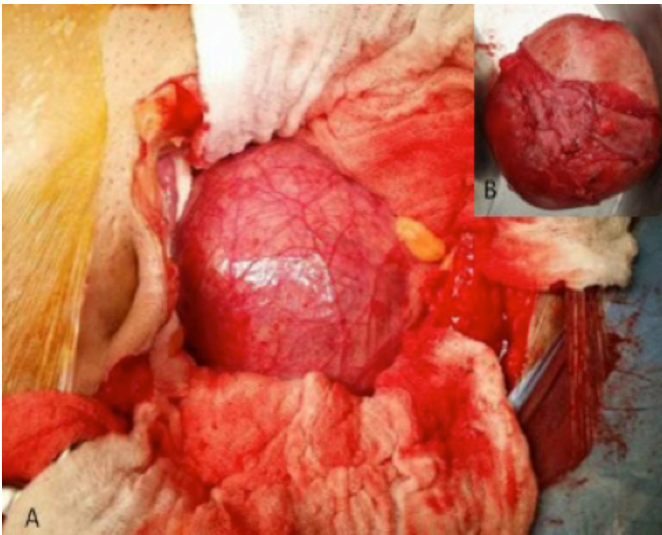


Figure 2: (A) Mass in situ peroperatively, (B) Mass after removal.

To relieve continuous abdominal pain, we decided to perform a cystectomy of the renal cyst along with an excisional biopsy of deep cervical lymph nodes. We used a midline infra-umbilical, intraperitoneal approach. There was a big mass occupying whole of lower abdomen. To help dissection and create space, around 100 ml of coffee ground fluid was aspirated from mass and was sent for cytology. Mass was dissected from surrounding structures and then finally was dissected off the upper pole of the kidney after applying clamps to the upper pole (Figure 2).

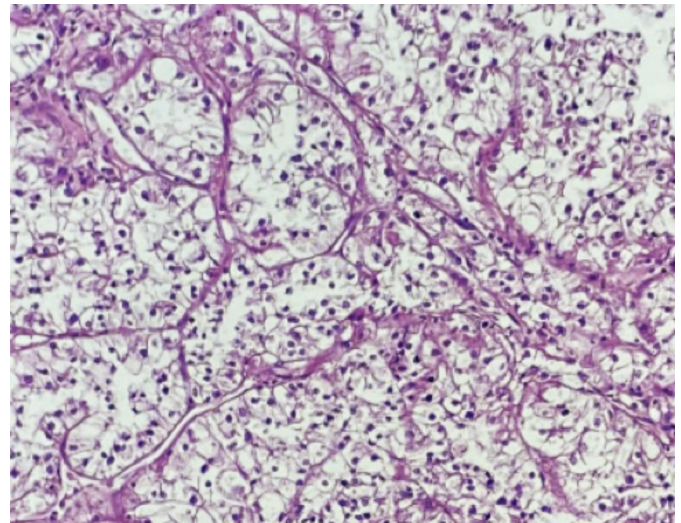


Figure 3: Clusters, nests and aggregates of polygonal cells with moderate clear cytoplasm, mildly atypical nuclei and inconspicuous nucleoli. Thin arborising network of blood vessels is evident (H&E, ×20).

The excised mass was 9 × 8 × 7.5 cm, tan brown, friable tumor. The histology showed clusters and aggregates of polygonal cells with moderate pale to clear cytoplasm, mildly atypical nuclei, and predominantly inconspicuous nucleoli. Thin branching arborizing vasculature was present with upto 40% necrosis. No sarcomatoid or rhabdoid features were noted (Figure 3). Perinephric fat invasion and lymphovascular invasion were absent. Tumor cells showed positive CD10 and PAX8 expression. Hence, based on classic morphology and positive CD10 and PAX8 expression, it was diagnosed as clear cell RCC, WHO/ISUP Grade 2. The renal parenchymal resection margin was 1.4 cm and the perinephric fat margin was 0.2 cm away. Deep cervical lymph node (level II) was sent separately and showed metastasis and the aspirate cytology was also suggestive of RCC. The final pTNM stage was pT2a, N1, M1.

He was referred to oncology for further management of metastatic disease and was advised Pazopanib, though the patient was lost to follow-up and 11 months later, presented with progression in metastasis along with multiorgan failure and expired.

DISCUSSION

There is scarce literature about RCC in pelvic kidneys with the evidence available in the form of a few case reports only.²⁻⁹ This case differs from the reported cases, because, firstly, it had an atypical presentation with enlarged regional and non-regional lymph nodes in the absence of any visceral deposits; secondly, the features of RCC on contrast CT were not typical with a predominantly cystic component which further added to the challenge; thirdly, it was the second reported case of pelvic RCC with metastasis; fourthly, in the reported literature so far, all cases which were managed surgically (eight out of ten) had either nephrectomy or nephroureterectomy and none was managed by partial nephrectomy alone. These features make this case the first of its kind in the reported literature so far.

RCC is known as a great masquerader, owing to its unusual presentations and metastatic potential to almost every organ of the body.¹⁰ It is known to metastasize *via* hematogenous and lymphogenous route. Involvement beyond the regional lymph nodes, *i.e.*, hilar, caval, and aortic is considered metastatic (M1). The basic knowledge of lymphatic drainage of kidney to retroperitoneal lymph nodes has mainly developed in cadaveric studies; however, the nodal metastasis of RCC is largely unpredictable.¹¹ The picture is further complicated by the existence of lymphovenous communication which may be present at the level of inferior vena cava (IVC) or renal vein. Direct drainage into the thoracic duct without passing through nodes has also been described, which explains the presence of visceral metastasis without the involvement of regional lymph nodes.¹¹

Because of its unusual location and uncertain vascular anatomy, ectopic kidney poses a surgical challenge. A few case reports have underscored the importance of preoperative imaging in the form of CT angiography and MR angiography. Owing to the rarity of this condition, optimal surgical approach, metastatic potential, routes of metastasis, and effectiveness of systemic agents in pelvic RCC compared to RCC in a normally located kidney are largely unknown. It seems that the masquerader will continue to amaze us.

PATIENT'S CONSENT:

Written informed consent was taken from the guardian prior to writing this manuscript.

COMPETING INTEREST:

The authors declared no competing interest.

AUTHORS' CONTRIBUTION:

RSR: Initial draft, acquisition of data, critical revision of the manuscript and final approval.

MHA: Operating surgeon, critical revision of the manuscript, and final approval.

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