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Surgical Management of Ectopic Kidney with Bilateral Iliac Vein Invasion

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ABSTRACT

Renal cell carcinoma (RCC) is a very rare phenomenon in an ectopic kidney. We come across a 61-year-old gentleman with a history of 2 months of gross, painless haematuria and palpable pelvic mass on examination. CT scan showed 6.5cm X 5.1cm X 5.8cm mass in pelvic kidney with bilateral iliac vein invasion. With the help of intra-operative ultra-sound, tumour thrombus was extracted from both iliac veins with en mass removal of tumour. Patient was well intraoperatively as well as in postoperatively. We also presented an elegant imaging for the case.

Keywords: Nephrectomy, RCC, Thrombus

CASE REPORT

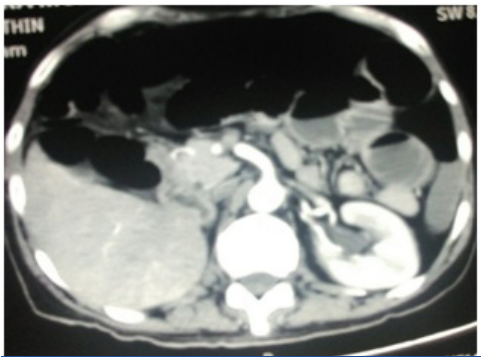
A 61-year-old gentleman with a history of 2 months of gross, painless haematuria not associated with clots or tissue bits passage and irritative lower urinary tract symptoms. There was no other significant history. On examination there was a palpable pelvic mass of size 10cm x 7cm, with well defined margins which was hard in consistency and movable. Other systems was unremarkable.

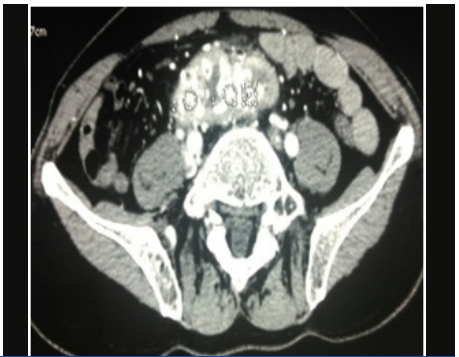
The blood investigations revealed haemoglobin of 11.8 gm%, white blood cell count of 8000/mm^3, serum creatine 1.5mg/dl and normal liver function tests. Chest X-ray was normal. The multislice CT scan was performed using subcentimeter contiguous axial scan of KUB with triple intravenous contrast. The right renal fossa was empty [Table/Fig-1]. The right kidney is malrotated and lying ectopically in pelvic region with renal pelvis facing anteriorly.

A well defined rounded soft tissue density lesion is noted involving lower pole of pelvic kidney [Table/Fig-2]. It measures 6.2cm x 5.1cm in axial plane with cranio-caudal length measuring 5.8cm. The lesion shows significant enhancement on post contrast study with multiple vascular channels supplying the mass and non enhancing necrotic centre [Table/Fig-3].

Single renal artery is noted supplying the ectopic kidney in pelvis arising distal to inferior mesenteric artery [Table/Fig-4] but with a dual venous drainage. One of the renal veins of the ectopic kidney draining into right internal iliac vein and other into the left common iliac vein. The lumen of the renal vein show luminal filling defects on venous phase extending into right internal iliac and left common iliac veins, suggestive of thrombus [Table/Fig-5].

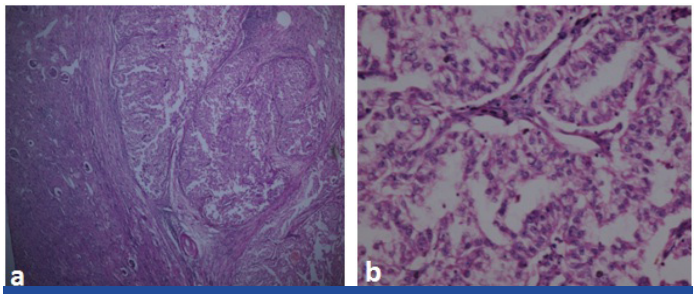
The patient underwent an open transperitoneal radical right pelvic nephro-ureterectomy with thrombus extraction from the left common iliac vein and excision of the right internal iliac vein with the thrombus within. We developed the space of Retzius bilaterally until there was visualization of the external iliac vessels. The sigmoid colon was medially displaced and the left colon mobilized medially. A renal artery was draped over the anterior portion of the kidney, originating from the aorta just distal to the inferior mesenteric artery origin. The renal veins took a medial course to reach the left common iliac vein and right internal iliac vein. We dissected the kidney superiorly and inferiorly until the ureter was visualized and tagged. The tumour and kidney were fibrotically adhered to the tissue overlying the sacrum and dissection in this plane was difficult due to the involvement of the pre sacral venous plexus. Intraoperative ultrasound was done to exactly locate the tumour thrombus within the veins. Tumour thrombus extraction was done from the left common iliac vein by performing venotomy and the right internal iliac vein was excised with the thrombus within after ligating its branches. The renal artery was transacted with good haemostasis. 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. Bilateral iliac group lymphadenectomy was done. There was significant bleeding from the pre sacral venous plexus intraoperatively which was controlled by pressure, packing and suturing. Patient needed six transfusions of blood intraoperatively. On postoperative day three, patient developed right lower limb paresis which improved on physiotherapy. Patient had a low grade fever self-resolved on postoperative day 6 and he was discharged from the hospital





[Table/Fig-1]: Absent right kidney in right renal fossa. **[Table/Fig-2]:** Right tumoural ectopic kidney in pelvis with tumor. **[Table/Fig-3]:** Tumour enhancement seen on contrast film.

[Table/Fig-4]: Vascular supply of the ectopic right kidney in pelvis.
[Table/Fig-5]: Tumour thrombosis of the left common iliac and right internal iliac vein.



[Table/Fig-6a]: Low power section showing tumour islands and part of normal kidney.
[Table/Fig-6b]: High power section showing tumour cells few with clear cytoplasm and few with eosinophilic cytoplasm.

on postoperative day 10 shortly following removal of his urethral catheter. Pathology revealed a 7.2cm x 3.3cm renal neoplasm with features of clear cell renal cell carcinoma (RCC) with sarcomatoid differentiation [Table/Fig-6a,b]. The tumour appears to breach the capsule. Fuhrman nuclear grade 3, with angiolymphatic invasion and negative ureteral and renal artery margins. The renal vein and cut margins of the renal vein were involved by the tumour thrombus. Adrenal gland not indentified. The separately send lymph nodal mass had eight lymph nodes all of which were free of tumour.

The patient is on surveillance for renal carcinoma and is following the protocol strictly.

DISCUSSION

The incidence of renal pelvic ectopia has been estimated to occur in 1 of 2100 to 3000 autopsies [1]. It is the failure of the mature kidney to reach its normal location within the renal fossa. Renal ectopia can occur at any of the locations pelvic, iliac, abdominal, thoracic, contra lateral, and crossed ectopic kidneys. Except for the development of hydronephrosis or urinary calculus formation, ectopic kidneys are asymptomatic and there is no increased risk to diseases than the normally positioned kidney [2]. Renal Cell Carcinoma (RCC) is a very rare phenomenon in an ectopic kidney. There are only seven reports of RCC occurrence in pelvic kidney in the literature [3-5]. With better imaging modalities having high sensitivity now-a-days the incidence of the malignancy in ectopic kidney is increasing. After searching the literature it was evident that this is the first case of renal cell carcinoma of the ectopic pelvic kidney with tumour thrombus in the left common iliac and right internal iliac vein. Kidneys and ureters may fail to migrate upwards. Proposed aetiologies are uterine bud maldevelopment, defective metanephric tissue that fails to induce ascent, genetic abnormalities, and maternal disease or teratogenesis [6]. During this period, the pattern of the renal vascular network is dependent on the position of the ectopic kidney and is completely anomalous. Ectopic kidneys may be supplied by one or two main renal arteries arising from the distal aorta, aortic bifurcation, and the common or external iliac arteries [6]. Considering the anomalous vascular supply and abnormal anatomy of the ectopic kidney the surgical approach to it merits caution. It is mandatory to have a detailed preoperative vascular anatomy evaluation and good imaging.

Terrone et al., performed preoperative magnetic resonance angiography for pelvic RCC and has suggested being a better substitute for CT angiography in depicting the renal vessels before nephrectomy [4]. Though they did not found venous thrombus extension as in our case. Mahmoudnejad N et al., did not perform any vascular imaging prior to exploration of pelvic RCC. Fortunately they did not come across any difficult situation intraoperatively [7].

CONCLUSION

This report describes a rare case of ectopic kidney with malignancy with invasion into bilateral iliac veins with a successful surgical management. Imaging plays an important role in such situations to provide the operating surgeon with detailed vascular and surrounding anatomy. So that, a proper surgical plan can be made preoperatively which can minimise the morbidity and mortality.

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