Large left moiety renal tumor with renal vein thrombus in a horseshoe kidney

Abhishek Bhat* and Bruce Kava

Abstract

Background: Management of large renal tumors in a horseshoe kidney has received little attention due to the highly infrequent occurrence. Surgical management for tumors in horseshoe kidney is challenging due to complex aberrant vascular anatomy, relative fixity of the renal moieties and other associated conditions such as ectopia and malrotation. An added component of a renal vein thrombus in this scenario often adds to the surgical conundrum. Inadequate peri-operative management may cause devastating complications and increased morbidity and mortality. Careful pre-operative planning and meticulous intra-operative dissection limits blood loss and thereby complications.

Case presentation: We present the peri-operative management of a large renal tumor with a renal vein thrombus in a horseshoe kidney moiety as a case report with literature review focusing on the intra-operative surgical approach that may be adopted for these tumors. A 12 cm x 10 cm left renal moiety mass with renal vein thrombus with no distant metastasis was diagnosed. After confirmation of thrombus extent with CT as well as MRI Abdomen, the patient underwent left moiety radical nephrectomy with renal vein thrombectomy. Careful intra-operative dissection and division of the vascular supply of the left renal moiety was performed. Vessel stapler was used for division of isthmus that resulted in minimal blood loss, complete oncologic clearance and minimal post operative morbidity.

Conclusions: Large renal tumor with renal vein thrombus in horseshoe kidneys is an uncommon condition. Pre operative cross-sectional imaging and intra-operative use of vessel sealants like Ligasure, harmonic scalpel and vessel staplers contribute to minimizing blood loss and thereby result in excellent patient outcomes. We report a case of a large left renal moiety tumor with renal vein thrombus successfully managed with open radical left moiety nephrectomy and isthmectomy with tumor thrombectomy.

Keywords: Kidney neoplasms, Renal cancer, Horseshoe kidney, Renal vein thrombus, Surgical stapler

1 Background

Horseshoe kidney is one of the most common anomalies of the renal variants with an occurrence of 1–4 per 1000 individuals [1]. Although the incidence of renal tumors in horseshoe kidneys is the same as normal kidneys, the management of these tumors especially with renal vein thrombi is more challenging due to variations in anatomy and vasculature, most often supernumerary arteries. The association of horseshoe kidneys with variant vascular anatomy, limited mobility and the location of the tumor with respect to the moiety contributes to the complexity of surgical management. Another challenge in horseshoe kidneys is the often rich communicating blood supply through the isthmus that may be present as a thick parenchymal bridge and can cause significant blood loss when divided despite having a good control of the arterial supply. The optimum usage of pre operative cross-sectional imaging allows visualization of the aberrant vessels and thereby planning the surgical approach. Meticulous dissection of the vessels, the affected renal moiety and the tumor allows significant reduction in intra-operative blood loss thereby reducing surgical morbidity. The division of the renal isthmus may be done with vascular

*Correspondence: bhatabhishek@gmail.com
1 Department of Urology, Jackson Health System, Miami, FL, USA
Full list of author information is available at the end of the article

© The Author(s) 2022. Open Access This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article’s Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article’s Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit http://creativecommons.org/licenses/by/4.0/.
stapler that produces a clean, quick and efficient division with minimal blood loss and minimal chances of urinary leak. There is no large reported series on this condition to the best of our knowledge and our attempt by presenting this case report, is to familiarize practicing urologists with a rational management plan with an in-depth surgical approach to avoid misadventures during a surgery that can frequently be complicated by bleeding as well as the need for blood transfusion.

1.1 Case presentation
A 57-year-old healthy gentleman was referred to us by his primary care physician with complaints of significant inadvertent weight loss of approximately 10–15 kg in 2–3 months, right sided flank pain and occasional bouts of diarrhoea. No gross haematuria or loss of appetite was reported. There were no other comorbidities. There was no history of smoking or occupational hazards with no prior surgical procedures that had been performed. Family history was unremarkable as well. Physical examination revealed a large, non-tender, solid but movable, ill-defined mass measuring about 15 cm × 10 cm in the left side of the abdomen.

Laboratory investigations revealed normal complete blood count (CBC), kidney and liver function tests. Imaging performed by the primary care practitioner in the form of computerized tomography (CT) scan of the abdomen revealed a horseshoe kidney with a relatively thin isthmus and a large, exophytic mass with a deep endophytic component measuring 12 cm in the left renal moiety (Fig. 1a, b). Note was made of a tumor thrombus within the left renal vein (Level I thrombus) (Fig. 2). The right moiety was found to be normal. The inferior vena cava (IVC) appeared normal. We performed a metastatic workup with a CT scan of the chest, Abdomen and Pelvis along with a magnetic resonance imaging (MRI) abdomen for better anatomical delineation of the thrombus (Fig. 3). Metastatic workup confirmed organ-confined disease and the tumor thrombus was noted to be limited to the left renal vein only. We made the diagnosis of horseshoe kidney with a large left moiety renal tumor measuring 12 cm × 10 cm TNM stage cT3aN0 with Level I tumor thrombus.

The CT and MRI images all pointed to a diagnosis of renal malignancy in the left moiety of a horse shoe kidney with a Level I renal vein thrombus extending till the ostium of the IVC. The patient was counseled about the management options and the intra/post-operative complications outlined to him.

Considering the patient’s favorable age and Eastern Cooperative Oncology Group (ECOG) status, a plan was made to treat the renal mass definitively with surgery. Open radical left moiety nephrectomy with isthmectomy and tumor thrombectomy was planned. After formal informed consent, we proceeded to surgery after ensuring compatible blood was available for transfusion and sterile urine cultures. Chevron incision was made. Post bowel mobilization and duodenal kocherization, IVC was identified and intense desmoplastic reaction was noted in the area of both the renal veins. The thrombus was easily palpable, and the ostium of left renal vein entering the IVC seemed uninvolved. A vessel loop was placed at this distal level to prevent escape of thrombus. The left colon was mobilized and the mass was noted to occupy almost the entire left side of the abdomen. The left renal moiety was mobilized in all directions with all parasitic vessels.
carefully divided using the vessel sealing device Ligasure™. The ureter was identified, clamped and divided. The left renal artery was dissected and divided between ties, which substantially improved the parasitic vessel bleeding and reduced the size of the left moiety. The left renal vein was noted to be encased by lymph nodes and Ligasure™ was used to free up the remaining attachments to the retroperitoneal musculature. The remnant part of the renal vein was then tied off at the IVC ostium after ensuring that there was no tumor thrombus migration and sent separately for analysis. The isthmus was divided with an Echelon linear stapler™. The specimen was delivered and sent for pathology analysis (Fig. 4). We noted small enlarged lymph nodes around the great vessels and performed a modified left retro-peritoneal lymph node dissection and removed all the visibly enlarged para-aortic, and inter-aortocaval lymph nodes as safely as possible.

The estimated blood loss was 400 ml from surgery and he remained hemodynamically stable throughout the surgery. He was monitored overnight in the ICU, transferred to the floor on postoperative day number 1, and then discharged on post-operative day 5, with continued improvement in bowel function and ambulation. Histopathology revealed a 12.5 cm Renal Cell Carcinoma (RCC) Fuhrman grade 4 invading the perirenal fat with negative surgical margins. The renal vein margin was negative for tumor and all dissected nodes were negative for cancer spread (TNM staging–pT3aN0). He was seen post-operatively at one-month follow up wherein he had already gained about 5 kg post-surgery, had normal laboratory results and had no complaints from surgery. No adjuvant treatment was administered in view of favorable surgical pathology and will continue with CT chest/abdomen and pelvis surveillance protocol as per the NCCN guidelines for RCC. At the time of last follow-up when this case report has been written, he has rejoined work and is asymptomatic with no evidence of recurrence on follow-up CT chest/abdomen and pelvis at 1 year.

2 Discussion
Renal cell carcinoma has a predilection for vascular invasion with reported rates of roughly 10% [2]. Wagner et al. reported that although the tumor level did not affect the prognosis significantly tumor size, fat invasion, involvement of lymph nodes and metastasis carried poorer outcomes [3]. As a consequence, an aggressive and effective tumor thrombus clearance should always be attempted for best results.
Although horseshoe kidney is the most common renal congenital anatomical variant, the coexistence of a large renal tumor along with renal vein thrombus is very rare. There is a higher incidence of renal carcinoids and Wilms tumors in horseshoe kidneys [4], but the incidence of RCC seems to be unaffected. A total of about 200 cases of RCC in a horse shoe kidney have been reported in literature [5], but only a handful of cases exist wherein there is described vein invasion into either the IVC or the renal vein [6]. A Level I thrombus can be safely resected with minimal mobilization of the IVC [7]. An important step to reduce bleeding when dealing with a vascular thrombus is early ligation of the renal artery which immediately decongests the veins and reduces bleeding.

The complexity of management of RCC in a horseshoe kidney also arises from the aberrant vascular anatomy in horseshoe kidneys [8]. Linxweiler et al. described the case of a similar complex endophytic renal tumor with tumor thrombus in the IVC, with multiple renal veins completely occluded by the tumor thrombus. After rightly proceeding with full dose anticoagulation and placement of IVC filter, the authors performed an open surgical extirpation of the involved kidney and isthmus, which involved an operative duration of 7 h, blood loss of approximately 5 L and a relatively uneventful hospital stay of 13 days. In our case, a pre-operative anticoagulation or placement of IVC filter was not indicated as such, as we took the patient up for surgery relatively rapidly. We were however aware of the potential for aberrant vessels and blood loss, which prompted us to use intraoperative autologous blood transfusion, if the need arose. We were also meticulous with the dissection around the kidney obtaining wide exposure and identification of all anomalous vessels before dividing them.

Complete tumor thrombectomy has been shown to be an important prognostic factor [9], and we were able to remove the entire tumor thrombus effectively with a combination of sealant and stapling devices. Management of the isthmus has been described in multiple ways–either with the digital compression and parenchymal suturing, cautery and suturing (monopolar or bipolar), suturing over clamps like the aortic cross clamps, Endo GIA stapler, microwave coagulator, harmonic scalpel, ultrasonic scalpel or with argon beam laser coagulation [10]. We chose the Echelon Flex 45 Stapler with vascular load and it produced a quick, clean and precise division with minimal blood loss. The end result was a very acceptable blood loss and discharge with no post-operative complications.

### 3 Conclusions
Large renal tumors with renal vein tumor thrombus in a horseshoe kidney moiety is a very rare diagnosis. Challenges in horseshoe kidneys include aberrant vascular anatomy, limited mobility and presence of an isthmus that contributes to surgical blood loss. Proper pre-operative imaging with either CT or MRI to define the extent of tumor thrombus is imperative for treatment planning. Use of vessel sealants like harmonic scalpel, Ligasure device and vascular staplers and respecting anatomical aberrations as a normal variation in horseshoe kidney anatomy is essential. Although there are many ways of dealing with the isthmus, one of the best techniques to reduce both blood loss and urinary leak is utilization of a vascular stapler.

### Abbreviations
CBC: Complete blood count; CT: Computerized tomography; IVC: Inferior vena cava; MRI: Magnetic resonance imaging; ECOG: Eastern Cooperative Oncology Group; ICU: Intensive care unit; TAP: Transversus abdominis plane; RCC: Renal cell carcinoma.

### Acknowledgements
None.

### Authors’ information
Both AB and BK are fellowship-trained urologic oncologists working in large academic organizations responsible for resident and fellow teaching.

### Authors’ contributions
Both AB and BK were responsible for concept, primary manuscript preparation and proof reading.

### Funding
None.

### Availability of data and materials
Available if required.

### Declarations

#### Ethics approval and consent to participate
Written and verbal consent was obtained from patient for participation.

#### Consent for publication
Written and verbal consent was obtained from patient for publication.

#### Competing interests
None.

### Author details
1 Department of Urology, Jackson Health System, Miami, FL, USA. 2 Department of Urology, University of Miami Miller School of Medicine, Miami, FL, USA.

### Received: 16 March 2022   Accepted: 13 May 2022
Published online: 31 May 2022

### References
1. Petrovic M, Andrejevic V, Djurasic L, Stamenkovic V, Acimovic M, Pejicic T et al (2012) Tumors of the horseshoe kidney: characteristics and literature review. Acta Chir Iugosl 59(1):53–55
2. Turun S, Banghua L, Zheng S, Wei Q (2012) Is tumor size a reliable predictor of histopathological characteristics of renal cell carcinoma? Urol Ann 4(1):24–28
3. Wagner B, Patard J-J, Méjean A, Bensalah K, Verhoest G, Zigeuner R et al (2009) Prognostic value of renal vein and inferior vena cava involvement in renal cell carcinoma. Eur Urol 55(2):452–459
4. Huang EY, Mascarenhas L, Mahour GH (2004) Wilms’ tumor and Horse-shoe kidneys: a case report and review of the literature. J Pediatr Surg 39(2):207–212
5. Pettus JA, Jameson JJ, Stephenson RA (2004) Renal cell carcinoma in horseshoe kidney with vena caval involvement. J Urol 171(1):339
6. Janitzky A, Meyer F, Halloul Z, Porsch M, Daher M, Kuester D et al (2010) Successful interdisciplinary treatment of renal cell carcinoma with tumour thrombus into inferior vena cava using multimodal protocol and organ-extending R0 resection in rare horseshoe kidney and doubled right organ. BMJ Case Rep 2:1105
7. Hevia V, Ciancio G, Gómez V, Álvarez S, Diez-Nicolás V, Burgos FJ (2016) Surgical technique for the treatment of renal cell carcinoma with inferior vena cava tumor thrombus: tips, tricks and oncological results. Springer-plus 20(5):132. https://doi.org/10.1186/s40064-016-1825-1
8. Linxweiler J, Shayesteh-Kheslat R, Fries P, Schneider G, Jansen M, Ohlmann CH et al (2018) Organ-preserving surgical treatment of a horseshoe kidney occupied by a large renal cell carcinoma with extensive venous invasion: a case report. Urol Int 100(2):245
9. Ljungberg B, Stenling R, Osterdahl B, Farrelly E, Aberg T, Roos G (1995) Vein invasion in renal cell carcinoma: impact on metastatic behavior and survival. J Urol 154(5):1681–1684
10. Khan A, Myatt A, Pailt V, Byani CS (2011) Laparoscopic Heminephrectomy of a Horseshoe Kidney. JSLS J Soc Laparoendosc Surg 15(3):415

Publisher’s Note
Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.