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## Renal pelvis squamous cell carcinoma with inferior vena cava infiltration: Case report and review of the literature

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## ABSTRACT

**INTRODUCTION:** Renal squamous cell carcinoma (RSCC) is a rare tumor that is usually diagnosed late as a locally advanced malignancy with adjacent structure involvement. Radical surgical resection with negative margins is the mainstay of treatment, as it is correlated with improved survival, while other modalities of treatment have been shown to have limited efficacy.

**PRESENTATION OF CASE:** We report a case of a 56 year old gentleman with right RSCC with tumor encasing the inferior vena cava (IVC), treated successfully with surgical resection.

**DISCUSSION:** The surgical management of vascular involvement of similar tumors has not been discussed in-depth in the literature. Surgical resection of the IVC without reconstruction can be done successfully in the circumstance of good collateral circulation; otherwise IVC resection with reconstruction will be necessary.

**CONCLUSION:** Radical resection with clear margins of RSCC tumors with vascular involvement is feasible in selected circumstances.

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## 1. Introduction

Cancers of the upper urinary tract (including renal parenchyma, renal pelvis and ureters) are rare, occurring in only 8% of all renal malignancies.<sup>1</sup> Furthermore, primary RSCC is less commonly seen, with a prevalence of 1.35% of the malignancies of the upper urinary tract.<sup>2</sup> Other histological types more commonly found are urothelial carcinoma. RSCC usually progresses insidiously, resulting in a late presentation; such cancers present at a later clinical stage and appear to be a more aggressive disease when compared to other histological types.<sup>2</sup> Prognosis is usually poor with a mean survival period of 7 months,<sup>1</sup> more so with neoplastic extension of the tumor into the inferior vena cava.<sup>3</sup> Surgical resection is the management of choice, as alternative modalities of treatment have not been proven to be effective.<sup>1,4</sup> We report a case of a 56 year old gentleman with right RSCC with tumor encasing the IVC, treated successfully with surgical resection.

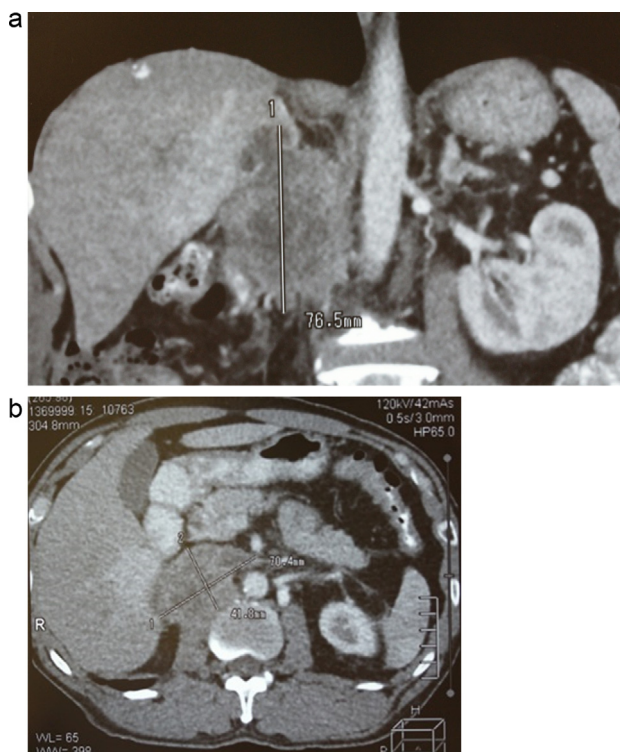
## 2. Presentation of the case

A 56-year-old man with a past medical history of hypertension, first presented to a district hospital with the complaint of hematuria. Investigations done included an intravenous pyelogram, demonstrating a horseshoe kidney with right kidney staghorn calculi. The patient underwent an exploratory laparotomy with the primary intention of removing the staghorn calculi. Intraoperatively, a large retroperitoneal mass was found, encasing the IVC down to the bifurcation of the iliac veins. In view of these unexpected findings, a debulking surgery was performed via a right nephrectomy. Histology of the resected specimen showed squamous cell carcinoma of the renal parenchyma. Subsequently, 6 cycles of carboplatin adjuvant chemotherapy was instituted to reduce the size of the remaining tumor. The patient then presented to our hospital for further treatment.

At our hospital, physical examination did not reveal any significant findings, with no palpable masses or lower limb swelling. Blood investigations, were not significantly deranged. Computer tomography (CT) scans of the chest, abdomen and pelvis (Fig. 1a and b) showed a retroperitoneal mass measuring 7.6 cm by 7.0 cm by 4.0 cm in the right renal fossa, contiguous with the anterior aspect of the paravertebral muscle, with invasion into the inferior vena cava, right adrenal gland and right psoas muscle. No distant metastasis to the liver, lungs or bone was detected.

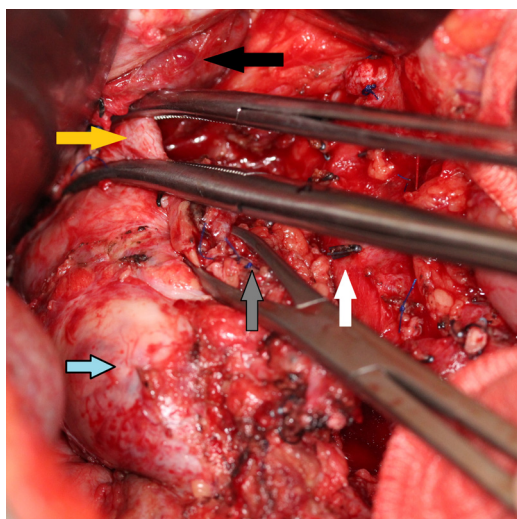
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**Fig. 1.** (a) Computer tomographic scan (axial view) showing retroperitoneal tumor, invading into the IVC. (b) Computer tomographic scan (coronal view) showing the retroperitoneal tumor, invading into the IVC.

The patient was electively admitted for exploratory laparotomy and resection of the right retroperitoneal tumor and IVC. Intraoperatively, a large tumor was found involving the IVC, extending caudally to just above the confluence of the iliac veins and superiorly to just below the insertion of the hepatic veins, lateral to the aorta, and medial to the psoas muscle (Fig. 2). Upon further dissection, proximal and distal control of the IVC was obtained. The left renal vein was then isolated and clamped distal to the gonadal and lumbar veins, but as close to the IVC as possible; the left



**Fig. 2.** Intraoperative view of the retroperitoneal tumor and surrounding structures from top clockwise: liver (black arrow), abdominal aorta (white arrow), tied renal artery (gray arrow), IVC tumor (blue arrow), proximal segment of the IVC double clamped (yellow arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

kidney was observed to continue to produce urine, hence the decision was made to ligate the left renal vein close to the IVC. Since the IVC clamping was well-tolerated by the patient, the IVC was resected en bloc with the tumor without reconstruction required.

Histology of the resected tumor showed moderately differentiated invasive squamous cell carcinoma, and margins were negative for malignancy.

Post-operatively, the patient wore compression stockings and was given subcutaneous clexane. No lower limb swelling was noted. His recovery was uneventful and he returned home on the 5th day.

Post-operative follow up at the 6th month with a positron emission tomography–computed tomography (PET–CT) scan showed no recurrence of the tumor.

### 3. Discussion

RSCC is frequently associated with risk factors such as chronic inflammation, nephrolithiasis, a non-functioning kidney, horseshoe kidney, analgesic abuse and smoking. SCC of the upper urinary tract is believed to arise via a process of squamous metaplasia of the urothelium.<sup>5</sup> There is relatively equal gender distribution in RSCC. Common presentations of RSCC have been frequently described as one or a combination of the following: hematuria, hydronephrosis, nephrolithiasis, loin or flank pain, paraneoplastic syndromes.<sup>5</sup> In our case, the patient had two of these risk factors that contributed to squamous metaplasia: staghorn calculi and a horseshoe kidney, and one of the described presentations of hematuria.

Diagnosis of RSCC is commonly made after surgical resection, by histological analysis of the resected specimen. Detection of the tumor on via imaging modalities is difficult because the common radiological features are those of renal calculi or hydronephrosis, both arising due to an obstructive process.<sup>6</sup> Several case reports used the non-invasive method of urine cytological to achieve the histological diagnosis, with confirmation after surgical resection.<sup>7,8</sup>

Mainstay of treatment of RSCC is still surgical resection, either by nephrectomy or nephroureterectomy, with clear margins. Benefit from neoadjuvant and adjuvant radiotherapy and chemotherapy is small in the management of RSCC.<sup>1</sup> Median survival of the patients where known is 7–10 months.<sup>1,2</sup>

The spread to the surrounding vasculature is not uncommon as the patients tend to present late, with the spread including that of the hilar vessels and the IVC. In the last 20 years, only 11<sup>9–17</sup> cases of vascular involvement of the RSCC tumor have been reported, of which 8<sup>9,10,14–17</sup> cases involved both renal vein and inferior vena cava, and 3<sup>11–13</sup> cases only the renal vein. However, the management of the vascular involvement of the tumor was not discussed in-depth in these cases.

Compared to renal cell carcinomas, where caval involvement is almost always limited to tumor thrombus within the lumen without significant vessel wall invasion, complete tumor resection can be accomplished without major inferior vena cava resection.<sup>17–19</sup> This is done by removing the tumor within the vessel with limited exposure and thrombectomy techniques. However, in our case of RSCC, the tumor invaded the entire IVC and thus oncologic resection requires complete caval excision. Invasion into the IVC wall was also reported in the other cases of RSCC.<sup>15,16</sup>

IVC resection without reconstruction can be successfully performed with the following caveat:

Good collateral circulation must be present in the form of the lumbar veins, adrenal veins, and gonadal veins. These pathways may be well developed in cases of tumor compression or rarely occlusion of the IVC. Good collateral circulation ensures that ligation will be well tolerated without clinical evidence of venous hypertension. It is confirmed by first clamping the left renal vein

and observing for good urine output, then clamping the IVC and observing that the patient's blood pressure remains stable. Test clamping is crucial because a sudden dip in the systolic blood pressure to approximately 80 mmHg would occur due to a sudden significant decrease in venous return in the case of poor collateral circulation.<sup>20,21</sup> Complete IVC occlusion without edema of the lower extremities post-operatively could be a predictor of the need for IVC reconstruction.<sup>22</sup> In our patient, he presented with no signs of lower limb edema.

If there should be no or poor urine output after clamping the renal vein, then the IVC should be reconstructed and the renal vein replanted. In the absence of potential intraoperative contamination, the ringed, reinforced polytetrafluoroethylene (PTFE) graft is our choice for IVC replacement.<sup>20,21</sup> Should there be a potential contamination, autogenous material is preferred. Options for an autologous graft for circumferential replacement would include the superficial femoral vein. Alternatively, a cryopreserved vein graft can be used.

IVC resection in our case was successfully performed due to sufficient collateral circulations (gonadal veins, lumbar veins, adrenal veins) which would avoid the occurrence of renal failure, and allow the effective drainage of the remaining kidney. Due to fulfillment of this caveat, an IVC reconstruction was not necessary. Preservation of the function of the kidney in this patient is of great importance as he has only 1 kidney remaining.

In view of the late clinical stage of RSCC at first presentation, with poor median survival times of 10 months or less, as well as the lack of effective adjuvant radiotherapy or chemotherapy, radical resection with clear margins may be the only method to prolong survival.<sup>3</sup>

#### 4. Conclusion

RSCC with involvement of surrounding vasculature is not uncommon. Radical excision with clear margins en bloc with the tumor is feasible in selected cases.

#### Conflict of interest

The authors declare no conflict of interest.

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#### Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contributions

Lin Zhimin involved in study design, data collection, data analysis and interpretation, writing the paper. Dr. Chng Jack Kian involved in study design, data collection, data analysis and interpretation, writing the paper, revising the article critically for important intellectual content, final approval of the version to be submitted. Dr. Chong Tze Tec and Dr. Soo Khee Chee helped for revising the article critically for important intellectual content, final approval of the version to be submitted.

#### Key learning points

- The primary curative treatment of RSCC is surgical resection by nephrectomy or nephroureterectomy with clear margins.
- Occasionally, these margins may include surrounding vasculature such as the IVC, which should also be resected.
- However, the decision to resect the IVC without its reconstruction will depend on the presence of good collateral venous circulation already present in the patient.
- This can be predicted if the patient has complete IVC occlusion with no edema of the lower extremities.
- Intra-operatively, test clamping the renal vein and the IVC while watching for changes in the systemic blood pressure and urine output will confirm this.

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