

Case Report

A rare case of successful excision of a Renal Cell Carcinoma with tumour thrombus in the inferior vena cava on a horseshoe kidney. A case report and description of the surgical technique.

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Introduction

Horseshoe kidney (HSK) is the most common renal fusion anomaly, it occurs in 0.25% of the individuals [1]. Patients are often asymptomatic and the anomaly is often incidentally discovered. However, when symptomatic, it is related to hydro-nephrosis, infection, or calculus formation [2]. HSK is associated with an increased risk of renal malignancy, with a relative risk of 1.76 to 7.93 times higher than that observed in the general population [3]. Renal cell carcinoma (RCC) is the most common neoplasm associated with HSK [2].

HSK is often associated with multiple renal arteries and veins [4], therefore resection of RCC in HSK is associated with challenges. Computed tomography (CT) utilizing routine renal mass protocol and CT angiography is the primary imaging modality of choice for localization and staging of any renal tumour and essential to study the renal vasculature.

We present a case of a 54-year-old male with a HSK, who was found to have RCC in the right moiety of the HSK, with Inferior vena cava (IVC) tumour thrombus along with adrenal metastasis and multiple renal arteries.

Case report

A 54-year-old man was investigated for fever. Abdominal ultrasound revealed, rotated and elongated bilateral kidneys, with medial deviation of the lower poles, which were fused at the midline by an interpolar parenchymal bridge. These findings were consistent with a HSK. A CT scan was subsequently performed, which revealed a large renal cell carcinoma in the right

moiety of the HSK measuring 70x30x20mm, with evidence of tumor thrombus extending into the IVC, through the renal vein up to the level of the hepatic venous openings. Enhancing masses seen in the bilateral adrenal glands measuring 16x18mm in size in left side and 9x7mm in right side. Each side of the HSK was supplied by three renal arteries. The lower polar renal artery to the left moiety was arising from the right common iliac artery. (Figure 1)

Alaparotomy was performed through a reverse L-shaped incision. The caecum, ascending colon and the right colic flexure was mobilised and rotated towards the midline. Kocherization of the duodenum was done. The IVC, and the renal veins were exposed, retrohepatic IVC was mobilised. The renal arteries were isolated, the lower renal artery supplying the right kidney was crossing anterior to the aorta.

The lower end was dissected at the isthmus. The arteries were ligated. A line of demarcation appeared extending across the isthmus. The lower pole was transected. The bleeding vessels from the left sided kidney surface were transfixed. Vascular clamps were applied to the retrohepatic IVC, left renal vein and the infra renal IVC. The IVC was opened near the right renal vein. The tumour thrombus was found to be attached near the right renal vein orifice. The rest of the tumour thrombus was not attached and was mobile within the IVC. Therefore, the tumour thrombus was removed with a cuff of the IVC en bloc with the kidney and the adrenal glands. (Figure 2).

Peri operative blood loss around 800ml, no urinary leakage noted intraoperatively, no immediate and late post operative complication noted.

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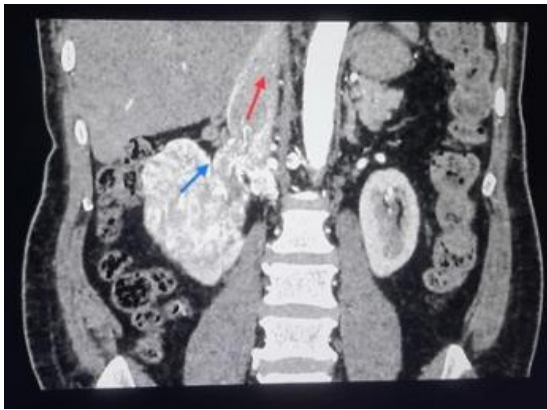


Fig 1- Contrast enhanced abdominal computed tomography scan showing the renal cell carcinoma in the right moiety of the horseshoe kidney (blue arrow), with Inferior vena cava tumour thrombus (red arrow)



Fig 2- Specimen of renal cell carcinoma in right side horseshoe kidney(purple arrow) with isthmus of horseshoe kidney(green arrow), Inferior vena cava tumour thrombus (blue arrow), and right adrenal gland (yellow arrow)

Histology shows clear cell renal cell carcinoma (WHO/ISUP grade 4) involving the upper and mid poles of the right kidney (85×69×90 mm). Rhabdoid and necrotic areas present with renal sinus and perinephric fat invasion, renal vein margin involvement, and adrenal metastasis. Stage pT3a pN0, Leibovich score 9/11 (high risk).

Discussion

Horseshoe kidney (HSK) is the most common type of renal fusion anomaly. HSK occurs in 0.25% of the

individuals [1]. HSK is twice as common in males as in females [2]. The most common mechanism of fusion occurs during abnormal migration of normally formed kidneys, thus having two separate excretory renal units. In more than 95 % of cases, fusion occurs at the lower poles. Depending on the degree of fusion, the isthmus can be composed of renal parenchyma or a fibrous band. This usually occurs between the fifth and ninth weeks of gestation. If large portions of the renal parenchyma fuse, the fusion anomaly loses its horseshoe appearance and appears as a pancake kidney or discoid kidney. Early fusion also causes abnormal rotation of the developing kidney[2].

The arterial supply of the fused kidney is variable in number and the site of the origin of the arteries. The arteries may arise from the common iliac arteries, internal iliac arteries, aorta and middle sacral arteries [2]. There are six different patterns of arterial supply described in Graves' classification [2]. Therefore, meticulous assessment of the preoperative imaging is necessary to lower the complications [3].

There is an increased risk of malignancies in HSK. RCC is the most frequently reported malignancy. However, RCC occurring in a HSK extending into the inferior vena cava (IVC), is not commonly reported. There have been only 200 cases with tumours in the HSK published in the literature so far, in which only 5-10% of cases had IVC tumour thrombus[4].

Imaging is vital in the diagnosis and staging of RCC. On ultrasound scanning (USS), RCC is seen as a heterogenous solid mass. The CT scan is essential in the diagnosis, staging and preoperative planning of RCC [5].

Since the renal tissues were fused in the above mentioned case, the nephrectomy done was technically a "partial" nephrectomy. In addition, the surgery was challenging in the above patient because of the following reasons. The challenges were, the variations in renal vasculature, the isthmus with the renal parenchyma and the presence

of IVC tumour thrombus. The potential surgical complications in HSK include; perioperative blood loss, urine leak and perioperative renal dysfunction.

Therefore, in conclusion, the above case illustrates that a meticulous preoperative evaluation, especially the anatomical evaluation, is essential in the case of HSK. This leads to a successful surgery, fewer complications and a better outcome.

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