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Renal cell carcinoma with tumour thrombus extending in duplicated inferior vena cava: A rarity with management conundrum

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Abstract

DIVC may play a role as a confounding factor in imaging diagnostic tests and also may represent a hazard for inadvertent injury and bleeding during surgery.

Careful interpretation Radiological investigations help to define such anomalies and avoid significant morbidity during surgical exploration.

We present a case of renal cell carcinoma involving Right kidney with tumor thrombus extending into both venae cava. Triphasic Contrast-Enhanced Computerized Tomography (CECT) scan of abdomen with three-dimensional reconstruction showed duplicated left sided IVC, Heterogeneously enhancing intraluminal soft tissue density filling defect was seen along the entire length of right renal vein, also crossing the midline to the left sided duplicated IVC -s/o tumour thrombus. Patient underwent right radical nephrectomy with tumour thrombectomy. Cavotomy was done at the junction of right renal vein and right IVC.

Entire thrombus was delivered intact. Histopathological examination showed papillary renal cell carcinoma (type II) stage pT3bN1Mx. IVC thrombus showed clusters of malignant cells of same morphology as tumor entangled in thrombus. The patient is doing well at 3 months of follow-up.

Keywords: DIVC; RCC; Tumor thrombus; Thrombectomy.

Abbreviations: DIVC: Duplicated Inferior Vena Cava; RCC: Renal Cell Carcinoma; CECT: Contrast Enhanced Computerized Tomography.

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Introduction

Congenital anomalies of inferior vena cava are extremely rare, reflecting the complexity of the embryological development of these structures. Duplication of IVC (DIVC) occurs in 0.7% of population. DIVC may play a role as a confounding factor in imaging diagnostic tests and also may represent a hazard for inadvertent injury and bleeding during surgery.

We describe a patient with right RCC with DIVC and presence of tumour thrombus involving right renal vein and both IVCs, treated by right radical nephrectomy with vena caval thrombectomy.

Case presentation

A 74 year old diabetic man was admitted with complaints of right flank pain since two months and history of passage of blood clots in urine. Physical examination, haematological and biochemical parameters were unremarkable.

Ultrasonography of abdomen showed a bulky kidney with irregular solid mass of size 4 x 3 cm in right kidney. Triphasic Contrast-Enhanced Computerized Tomography (CECT) scan of abdomen with three-dimensional reconstruction revealed heterogeneously enhancing soft tissue density lesion predominantly in the right lower pole extending into the renal pelvis, extending into the calyces and the proximal ureter. Duplicated left sided IVC was seen as a continuation of left common iliac vein, crossing anterior to the aorta just above the level of left renal vein to join the right sided IVC. Heterogeneously enhancing intraluminal soft tissue density filling defect was seen along the entire length of right renal vein, extending into the supra and infrarenal portion of right IVC (for a length of (14.0 cm) and also crossing the midline to the left sided duplicated IVC (for a length of (4.0 cm) - s/o tumour thrombus. Left renal vein was free from tumour thrombus (Figure 1 and Figure 2).

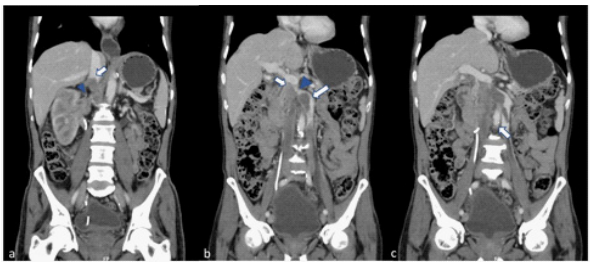

Figure 1: Conronal sections of Right renal cell carcinoma with thrombus in both IVC (duplication of IVC).
Image a: Showing right renal vein with tumor thrombus (blue arrowhead) with extension of the thrombus into the right IVC (white arrow mark).
Image b: showing communication between right and left IVC (blue arrowhead) with thrombus in right IVC (short white arrow mark) and left IVC (long white arrow mark).
Image c: Throwbus in both the vena cava surrounding Aorta (white arrow mark)

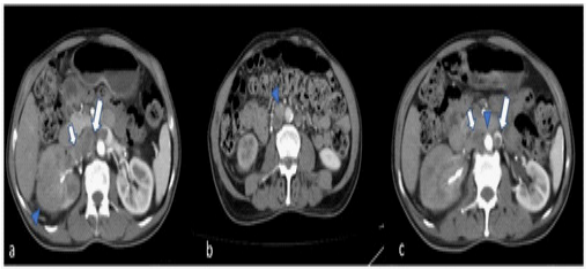

Figure 2: Axial sections of right renal cell carcinoma with thrombus in both IVC (duplication of IVC)
Image a: Non enhancing right kidney with tumor (blue arrowhead) with thrombus extending into the renal vein (short white arrow mark) and crossing through the connection into opposite IVC (long white arrow mark).
Image b: Enhancing lymph node mass (blue arrowhead) seen anterior to the right IVC.
Image c: axial view of three major vessels – Right IVC (short white arrow mark), Aorta (blue arrowhead), Left IVC (long white arrow mark)

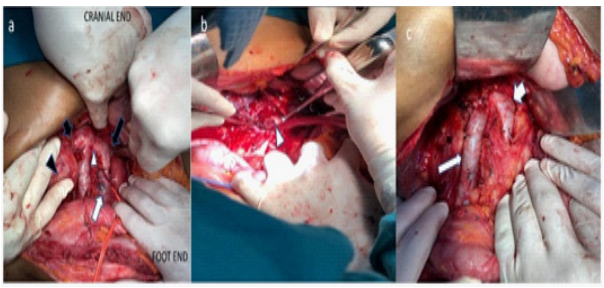

Figure 3: Intraoperative images showing Duplication of IVC with tumor thrombus and lymph node mass
Image a: Shows right kidney with renal cell carcinoma (black arrowhead) with tumor thrombus extending into right renal vein (black short arrow mark). Connection between the right and left IVC can be seen (white arrowhead), Left renal vein can be seen joining the left IVC (black long arrow mark). Lymphnode mass after mobilizing away from the Right IVC can be noted (white arrow mark).
Image b: Shows inferior venacavotomy with extraction of tumor thrombus (white arrowhead).
Image c: Right (long white arrow mark) and left (short white arrow mark) IVC after thrombectomy.

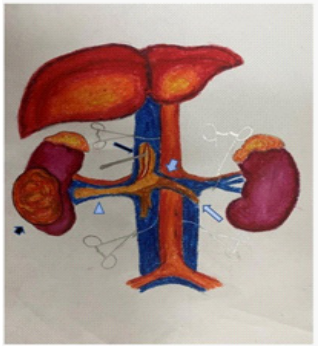

Figure 4: Diagramatic representation: Right renal mass (short black arrow mark) with extension of tumor thrombus into the renal vein (blue arrowhead) and right IVC (long black arrow mark). With bland thrombus extending into the left IVC(long blue arrow mark) via the communication (short blue arrow mark)

Patient was taken up for right radical nephrectomy, with tumour thrombectomy through a transperitoneal Chevron incision with extension in midline upto xiphisternum in supine position. Hilar dissection done and right IVC was bared. Right IVC was seen communicating with left IVC anterior to the aorta with the isthmus at the communication at just superior to the level of left renal vein. Tumour thrombus was seen extending from right renal vein into the suprarenal and infrarenal portion of right IVC and periosteal region of left IVC. Few enlarged retrocaval, precaval and aortocaval lymph nodes were also noted. Right renal artery and ureter were ligated and entire kidney mobilized outside Gerota's fascia, leaving the kidney attached only by the right renal vein. Leftcolon was mobilized and left IVC was bared. Left renal vein was noted to be free from tumour thrombus. Infrarenal Right and left IVCs, left renal vein, and suprarenal IVC just below liver proximal to tumour were clamped in that order. Cavotomy was done at the junction of right renal vein and right IVC. Entire thrombus was delivered intact following which IVC was closed using Prolene 5-0 suture (Figure 3 and Figure 4).

Abdominal closure was done in two layers after placement of a single pelvic drain. Estimated blood loss was 550 ml with one unit of PRBC transfusion done intra operatively. Postoperative period was uneventful with drain removal on third postoperative day. Patient was allowed per orally on second postoperative day and discharged on fourth postoperative day. Histopathological examination showed papillary renal cell carcinoma (type II) stage pT3bN1Mx. Hilar and retroperitoneal lymph nodes were positive for metastasis. IVC thrombus showed clusters of malignant cells of same morphology as tumorentangled in thrombus. The patient is doing well at 3 months of follow-up.

Discussion

The IVC is formed between weeks 6 and 10 of gestation. There are several theories accounting for the duplication. It has been estimated that duplication occurs in 0.2–3.0% of the general population. The infrarenal portion of the IVC is formed from two embryonic veins, the supracardinal veins. The right supracardinal vein persists and develops as IVC while the left supracardinal vein regresses. Persistence of both supracardinal veins results in duplication of IVC [1,2].

Three different variants of duplication of inferior vena cava are known i.e Type I or Major duplication: Two bilaterally symmetrical trunks with a preaortic trunk of the same calibre. Type II or minor duplication with two bilaterally symmetrical trunks, but it is smaller than the preaortic trunk. In Type III or asymmetric duplication there is small left IVC, larger right IVC and even larger preaortic trunk [3]. Our patient was noted to have type I duplication.

Chuang categorised the abnormalities of the postrenal segment of the IVC into four categories: Type A is a persistent right posterior cardinal vein (retrocaval ureter), type B is a persistent right supracardinal vein (normal IVC), type C is a persistent left supracardinal vein (left IVC), and type BC is a persistent right and left supracardinal veins (double IVC) [4].

Locally advanced RCC can have a myriad of presentation from haematuria to fatigue, weight loss to lower extremity oedema, ascites and pulmonary embolism [5]. CT plays an important role in delineating the primary renal pathology and the extent of thrombus invasion into IVC. Because thrombus study requires enhancement phase, a multidetector CT has improved the sensitivity and specificity of CT in detecting RCC by improving accuracy of staging and enhancing surgical planning [6,7].

MRI is however the gold standard for demonstrating level and extent of tumour invasion in IVC with a sensitivity close to 100%. Recent studies have however highlighted equal efficacy of both MRI and multidetector CT for evaluation of tumour thrombus [8]. In our case, CECT revealed double vena cava sign with tumor thrombus in both vena cava extending into suprarenal IVC.

Although IVC duplication is usually asymptomatic, it might have significant clinical implications. As an uncommon anomaly, the duplication of IVC can be misdiagnosed as a pathological lesion such as ureteric dilatation or lymphadenopathy on CT images. The left side of a double IVC might be interpreted erroneously as enlarged retroperitoneal lymph nodes [2,9,10,11]. Which might induce preoperative over staging by radiographic imaging in RCC.

The recognition of IVC anomalies becomes important when radiological, interventional, or surgical procedures involve these structures. Diagnosis of DIVC is important when an IVC filter is to be placed for thromboembolic disease [12]. Reports have showed the importance of recognizing a double IVC when performing renal vein sampling or adrenal venography because blood flow carried by the left vena cava will dilute the left renal vein sample [4].

Early ligation of renal artery is an important step in management of such patients. In a study of 82 patients, Ciancio et al proposed early ligation of renal artery as compared with preoperative embolisation of renal tumours and found it significantly reduced bleeding by collapsing many of the collaterals. They also proposed early control of renal artery by ligating it behind the RV [13].

Recognition of DIVC before or during radical nephrectomy can prevent injury to the anomalous vasculature and resulting exsanguinating hemorrhage. It is also important that control of both IVCs be obtained before cavotomy. In the rare instance that tumor thrombus extends into the anomalous vessels, thrombus extraction from both the IVCs has to be ensured, as was done in our case. If the tumor thrombus is seen invading left IVC, the left IVC can be excised. However, resection of the left IVC despite the absence of an interiliac vein does not reduce the venous drainage from the left femoral vein probably because of abundant pelvic venous channels [14]. Lymphatic drainage tends to follow the vessels. Thus, patients with abnormal venous anatomy may have unusual patterns of lymphatic drainage and lymph node metastases. Therefore, lymph node dissection in a patient with a venous anomaly should be altered accordingly whenever indicated [10].

Conclusion

The recognition of congenital IVC anomalies has major clinical implications. In addition to preventing diagnostic errors, careful interpretation of cross-sectional imaging can help to avoid complications arising from venous anomalies unexpectedly encountered during surgery.

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