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A diagnostic and management conundrum in a rare case of a small renal mass UTUC with IVC and bilateral renal vein tumour thrombus

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ABSTRACT

Upper Tract Urothelial Carcinomas (UTUC) are generally uncommon, accounting for approximately 5% of all urinary tract tumours. This report describes a unique Case of a 52-year-old-male with no known risk factors or symptoms of UTUC, who presented with bilateral sub-massive pulmonary embolus (PE). Subsequent computed tomography (CT) demonstrated a small (<2cm) right cortical based mass a discordant venous tumour thrombus (VTT) extending in the IVC, up to the level of the hepatic vein and bilateral renal veins. The patient had surgical excision in the form of right radical nephroureterectomy, IVC resection with bovine pericardial graft reconstruction and left renal autotransplant.

1. Introduction

Upper Tract Urothelial Carcinomas (UTUC) with concomitant renal vein (RV) and/or inferior vena cava (IVC) tumour thrombus involvement is an uncommon finding. Huber et al. reviewed their series of 102 cases of UTUC managed over 20 years and found an incidence of 5%, however fewer than 40 cases have been reported to date.[1]

2. Case report

A 52-year-old man presented to the Emergency Department (ED) with chest pain and desaturation. Computed Tomography Pulmonary Angiogram (CTPA) showed sub-massive bilateral pulmonary emboli (PE). Lower limb doppler ultrasonography was negative for deep vein thrombosis. Despite compliance with his anticoagulation the patient represented with worsening PE. He had an inferior vena cava (IVC) venogram which demonstrated an extensive filling defect in the right renal vein (RV), as well as the IVC up to the level of the hepatic vein. The patient had a subsequent suprarenal IVC filter inserted.

Urology was consulted following CT findings of a right upper pole solid cortical lesion with contrast enhancement, measuring 15 × 18mm with bilateral RV and IVC thrombosis (Fig. 1). Given the discordance between the small renal mass and the extensive tumour thrombus into the IVC, a renal mass biopsy was taken to further characterise the lesion. The biopsy demonstrated inconclusive findings of a poorly differentiated carcinoma favouring urothelial differentiation. Multidisciplinary team (MDT) consensus involving Urology, Oncology, Vascular, Anaesthetics, Intensive Care Unit (ICU), Respiratory and Haematology was that the patient was extremely high risk for surgery given the significant cardiorespiratory distress due to the extensive PE, and therapeutic anticoagulation. The decision was made to commence neoadjuvant chemotherapy and reassess feasibility of surgery.

Following the completion of 5 cycles of chemotherapy (cisplatin and gemcitabine), repeat CT staging confirmed locally advanced disease. The patient subsequently underwent an open right radical nephroureterectomy (Fig. 2), IVC excision and reconstruction with tubularised bovine pericardial graft (Fig. 3), and left renal autotransplant. Intraoperative findings showed that the entire infrahepatic IVC was indurated and woody, concerning for wall involvement. The left RV was also noted to be non-mobile and fixed, concerning for tumour thrombus involvement and was confirmed with an intraoperative ultrasound. Given the above findings, the option between anastomosis of the left RV to the caval graft and autotransplant was discussed. Due to the predisposition for altered flow and resultant significant thrombosis risk associated with any type of vascular graft up to 22%, decision was made to autotransplant the left kidney.[2]

Final histopathology revealed pT4, high grade urothelial carcinoma with involvement of the right kidney, perinephric fat, adrenal gland, IVC vessel wall, bilateral RV and IVC thrombus. Bladder was clear of urothelial malignancy on cystoscopy.

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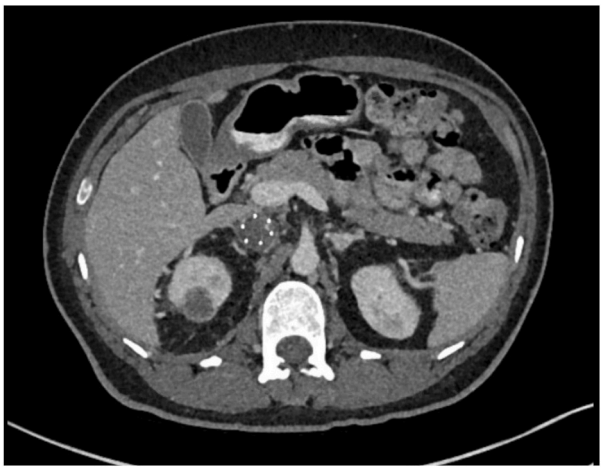

Fig. 1. Right upper pole solid hypovascular lesion measuring 15 × 18mm with IVC tumour thrombus.

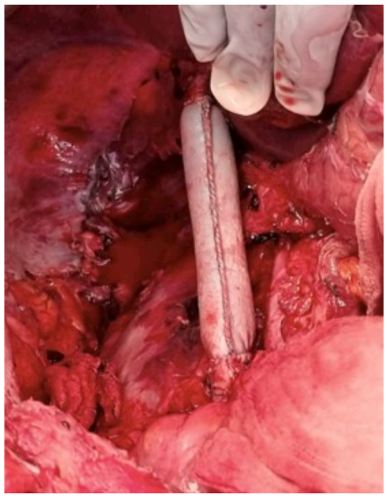

Fig. 2. Right nephroureterectomy.

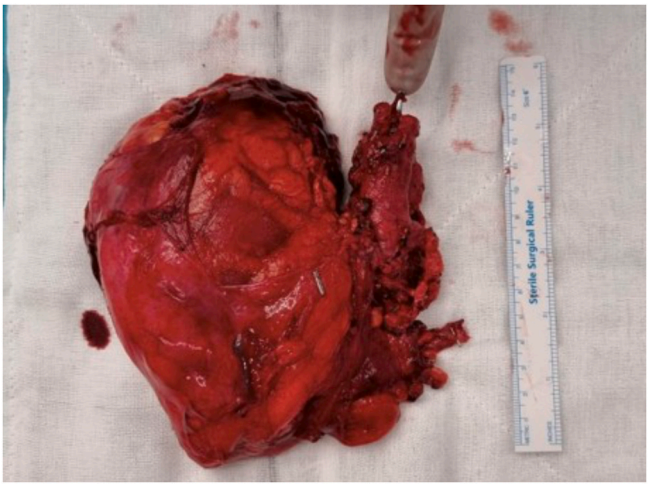

Fig. 3. Tubularised bovine pericardial graft.

Follow up after two months showed that our patient remains well with no evidence of disease (NED) at 2 months, patent IVC graft and good function from his left transplanted kidney. His renal function remains stable with an eGFR of 53 and creatinine of 132. He will be commencing Avelumab based on recent trial findings suggesting survival benefit over observation.

3. Discussion

An unusual feature of this Case was the cortical based renal mass with no evidence of collecting system involvement on MRI or dedicated CT renal studies. A retrospective cross-sectional study evaluated the diagnostic performance of individual CT features and their reliability in differentiating UTUCs of the renal pelvis from centrally located renal cell carcinoma (RCC). The features used for differentiating UTUC from RCC included tumour centre in collecting system, focal filling defect in renal pelvis, renal shape preservation, absence of cystic or necrotic change, homogeneous enhancement of tumour and extension towards ureteropelvic junction.[3] These signs identified UTUC of the renal pelvis with a sensitivity of 68–82% and specificity between 79 and 89%.[3]

One of the biggest challenges in our patient was the extensive tumour thrombus from UTUC. In a retrospective study by Psutka et al. 172 patients with IVC tumour thrombus were evaluated to predict the IVC wall invasion and the need for extensive vascular resection requiring complex vascular reconstruction (complete IVC resection, venous patch graft, tube-interposition graft).[4] The authors concluded that the presence of right-sided tumour, anteroposterior diameter of the IVC at the RV ostium at least 24mm, and radiographic evidence of complete occlusion of the IVC at the RV ostium were associated with a significantly increased risk of need for extensive vascular resection.[4] If a patient had none of these features, the predicted probability of requiring extensive vascular resection was 2% in comparison to 66% of patients with all three risk factors.[4]

The use of neoadjuvant therapy in the preoperative setting is considered experimental and there are no clear guidelines available on its efficacy. There have been several Case series reported on using neoadjuvant systemic therapy agents to debulk and downstage the tumour thrombus level, to permit less invasive thrombectomy approaches, potentially avoiding bypass procedures or entry into the chest. As of yet, there is no established level one evidence to support the use of targeted agents prior to IVC tumour thrombectomy. Even less so, is reported use of neo-adjuvant chemotherapy in the case of UTUC and thus remains an area to be explored in future studies.

Another intriguing aspect of this Case was the use of renal autotransplantation (RAT) which has been used successfully in the management of complex kidney pathology including: centrally located renal tumours, extensive ureteral injuries, renal artery aneurysms, renovascular hypertension, and chronic flank pain. Novic et al. reported on RAT in the largest series to date, 108 cases in total which included 14 cases of complex RCC.[5] They reported a primary graft loss of 14% in the 14 cases of RCC. Of the remaining 12 patients in this group, five were free of cancer at follow-up. There was a 5-year overall survival of 70%. Only four patients died as a result of RCC recurrence or progression.

4. Conclusion

There is paucity of data in existing literature regarding the optimal role and timing of surgical and systemic treatment for UTUC with bilateral RV and IVC tumour thrombus. We highlight that neoadjuvant chemotherapy followed by radical nephroureterectomy with IVC thrombectomy can be a safe and feasible operative method.

Consent

Informed consent was obtained from the patient prior to submission of Case report.

Submission declaration

The Case report is an original article has not been published or submitted elsewhere.

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