Adrenocortical carcinoma with inferior vena cava thrombus: Renal preserving surgery

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ABSTRACT

Adrenocortical carcinoma with tumor thrombus extending via an adrenal vein into the inferior vena cava is uncommon. We describe a left side kidney-preserving surgery in such a patient where, after transection of the main renal vein at the point of insertion of the adrenal vein, the left kidney was being drained by lumbar and gonadal veins.

Key words: Adrenocortical carcinoma, inferior vena cava thrombus, renal preserving surgery

INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare malignancy (0.5–2 cases/million/year) with poor prognosis (5-year overall survival: 32–45%) due to delay in diagnosis and the lack of effective adjuvant treatment.^[1,2] Several studies indicate that radical surgical resection can improve survival.^[2] The prognostic value of inferior vena cava (IVC) thrombus extension is debated but does not represent a contraindication to surgery.^[3,4]

Right-sided ACC may grow into the IVC more frequently than left sided tumors. Left side ACC extends into the IVC through the renal vein. In such a situation, a left nephrectomy is usually performed along with resection of the tumor and the thrombus. To the best of our knowledge, this is the first case report of a kidney-preserving adrenalectomy and IVC thrombectomy in a patient with bilateral normally functioning kidneys.

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CASE REPORT

A 55-year-old healthy male was incidentally detected to have a retroperitoneal mass in the left suprarenal location. Laboratory tests including serum creatinine, cortisol, aldosterone, 17-hydroxy progesterone, androstenedione, urinary vanillylmandelic acid, epinephrine, and norepinephrine were normal. Computed tomography and magnetic resonance imaging showed an enhancing left adrenal mass (8 cm) with a tumor thrombus extending into the infrahepatic IVC (thrombus extending up to 5 cm of IVC from renal vein), displacing the left kidney laterally [Figure 1a and b]. A whole-body scintigraphy was negative for bone metastases.

Laparotomy was performed with a Chevron incision and lower sternotomy. We proceeded with an *en bloc* resection of the mass and the para-aortic lymph nodes after mobilization of the spleen and the tail of the pancreas. The infrahepatic IVC was exposed and venous control was achieved by placing four tourniquets; two on the IVC cranial and caudal to the thrombus, one around the left renal vein proximal to the thrombus and one around the right renal vein. After caval clamping, thrombectomy was performed by a longitudinal incision of the cava. The thrombus was also infiltrating the IVC wall, which was

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Figure 1: (a) Illustration showing adrenal cortical carcinoma with tumor thrombus extending into inferior vena cava. (b) Contrast enhanced computed tomography abdomen showing adrenal mass with tumor thrombus extending into inferior vena cava (Star depicting adrenal mass and arrow showing tumor thrombus extending from adrenal vein into renal vein and inferior vena cava)

resected with the specimen. The specimen included the tumor thrombus, infiltrated IVC wall, distal left renal vein, adrenal vein, and the adrenal mass [Figure 2a]. The IVC was reconfigured with a Prolene 5-0 continuous suture. The renal vein proximal to the thrombus was sutured with 6-0 Prolene, preserving the gonadal and lumbar veins [Figure 2b].

Postoperatively on day 1, the serum creatinine started rising due to acute tubular necrosis secondary to intraoperative hypotension. In view of rising creatinine, color Doppler was done which showed normal patency and flow in the IVC and renal vein. The patient needed renal replacement therapy and supportive intensive care unit management for a week followed by gradual return of renal function to normal. At 1 year follow-up, the patient is doing well without any evidence of metastasis on positron emission tomography contrast enhanced computed tomography (PET-CECT) with a serum creatinine of 1.1 mg/dL. CECT abdomen images acquired during the PET-CECT study at 1-year follow-up showed normal contrast uptake and enhancement in both kidneys [Figure 2c]. Histopathology was consistent with poorly differentiated ACC. The tumor thrombus showed the same pathology. Five lymph nodes were present, and all were negative for metastasis.

DISCUSSION

The management of a patient with left ACC extending into the IVC is complex. Considering that prognosis depends strictly on a radical resection, the main aim is to obtain a complete excision of the tumor.^[1,2] Therefore, an aggressive approach is required, however preserving the ipsilateral left kidney whenever possible, is a surgical challenge.



Figure 2: (a) Pathology specimen showing resected adrenal mass along with inferior vena cava thrombus. (b) Illustration showing venous drainage of left kidney after wide excision of adrenal mass with inferior vena cava thrombectomy. (c) Contrast enhanced computed tomography abdomen at 1-year follow-up showing normal contrast uptake and enhancement in both kidneys

The adrenal venous drainage varies by side and both adrenal glands are drained by a single large vein. On the left side, the adrenal vein enters the cranial aspect of the left renal vein and on the right side, the adrenal vein enters the IVC directly on its posterio-lateral aspect. In our patient, the tumor thrombus was extending from the left adrenal vein into the distal left renal vein and then into the IVC as shown in Figure 1a and 1b. Renal preserving surgery on the left side is technically demanding. After resection of the distal left renal vein, venous drainage of the kidney was possible through lumbar and gonadal veins. Thus the left kidney was preserved even though the distal part of the left renal vein was excised [Figure 2b].

A few studies have shown significant survival after radical surgery in patients of ACC with IVC thrombus.^[1,5] Local stage, curative surgery, age below 35 years, and no other organ resection are significantly associated with better survival.^[1] An aggressive approach is not recommended in case of metastasis because even with radical surgery, the 2 year survival rate is 20%.^[1]

CONCLUSION

ACC with tumor thrombus extending into the IVC does not represent a contraindication to surgery and curative resection is feasible. Kidney-sparing surgery, if possible, should be attempted without compromising surgical margins.

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Conflicts of interest There are no conflicts of interest.

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