

Adrenal cortical carcinoma with pulmonary emboli: A unique presentation of a rare tumor with extensive tumor thrombus and inferior vena cava extension

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ABSTRACT

Introduction: Adrenal cortical carcinoma (ACC) is rare, and presenting symptoms vary widely depending on functional or non-functional endocrine status. These tumors are most commonly treated with surgical resection and postoperative mitotane administration. **Case Report:** This is an unusual presentation of a 23-year-old female with no significant past medical history, admitted to the hospital with syncope and dyspnea. Computed tomography angiography (CTA) demonstrated extensive bilateral pulmonary embolisms, with an associated 16-cm assumed right lobe hepatic mass with suprahepatic vena cava tumor thrombus extension beyond the level of the hepatic veins. The patient underwent a complete resection of the right adrenal mass, with inferior vena cava resection, thrombectomy, and placement of caval interposition graft without the use of bypass. Pathology was consistent with adrenal cortical carcinoma. **Conclusion:** This case of an adrenal cortical carcinoma, with a rare presentation

of bilateral pulmonary embolisms, was treated with a surgical RO resection. This included a right adrenalectomy with IVC resection and interposition graft. Tumors with IVC involvement and tumor thrombus can be treated with surgical resection and IVC grafting, without the use of bypass.

Keywords: Adrenalectomy, Adrenal cortical carcinoma, Inferior vena cava

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INTRODUCTION

Adrenal cortical carcinoma (ACC) is an extremely rare condition, with an incidence of 1–2 per million populations [1–5]. Patients may present with symptoms of steroid surplus such as Cushing's syndrome, virilization, hirsutism, or oligomenorrhea. In atypical cases, presentations may include hypertension or electrolyte disturbances associated with hyperaldosteronism. However, nonfunctioning tumors may present with abdominal or back pain and vague gastrointestinal

symptoms. These are extremely aggressive tumors requiring a complete R0 resection and adjuvant treatment with mitotane. Large-vessel invasion has been demonstrated as a poor prognostic indicator, with a <30% 3-year survival rate [2, 3]. Resection of ACC with caval extension and tumor thrombus has been described previously and has required a cavotomy for thrombus removal or venous patch, conducted with the use of cardiac bypass or veno-veno bypass [4, 5]. This report describes a right ACC with rare presentation of initial bilateral pulmonary embolisms and with suprahepatic vena cava tumor thrombus extension above the level of the hepatic veins. The patient required a right adrenalectomy, inferior vena cava (IVC) resection, thrombectomy, and IVC replacement with composite graft, conducted without the use of bypass.

CASE REPORT

A 23-year-old woman with no significant past medical history was admitted to the hospital with dyspnea and syncope. The admitting laboratory values included: Na 141 mEq/L, Cr 0.79 mg/dL, glucose 129 mg/dL, troponin 0.412 ng/mL, D dimer 10.4 ug/mL FEU, lactate 2 mmol/L. A computed tomography scan of the head was normal. However, a computed tomography angiogram of the chest demonstrated extensive bilateral pulmonary emboli in the right and left main pulmonary arteries, with extension into lobar, segmental, and subsegmental branches. Doppler ultrasound of the lower extremities was negative for venous thrombosis. An echocardiogram showed significant right atrial and ventricular dilation with markedly reduced systolic function but preserved ejection fraction and a well-circumscribed mass in the IVC consistent with thrombus. She was treated with tissue plasminogen activator and continuous heparin infusion.

Ultrasound of the abdomen revealed a 16-cm mass thought to arise from the right hepatic lobe with a 5-cm IVC thrombus. On subsequent magnetic resonance imaging, a 16.6-cm mass was noted in the right upper quadrant adjacent to the posterior right hepatic lobe with extension into the intrahepatic IVC and prominent tumor thrombus extending just distal to the right atrium (Figure 1A–1C). Urine metanephrine and normetanephrines were negative, but the free cortisol level was elevated. Her dehydroepiandrosterone level was significantly elevated, while her morning cortisol level was within normal limits. Tumor activity was negative for adrenocorticotropic hormone, alpha-fetoprotein, and carbohydrate-associated antigen 19-9. A transjugular biopsy of the tumor thrombus was attempted; however, a large multilobular filling defect in the IVC was noted extending beyond the level of the hepatic veins, resulting in an 80% stenosis (Figure 1D). Percutaneous biopsy was consistent with ACC, with positive immunohistochemistry staining for inhibin and MART-1 but negative staining for

adrenocorticotropic hormone.

The tumor was arising from the right adrenal gland with multiple enlarged venous collaterals enveloping the surface and adherent to the inferior aspect of the liver with no apparent tumor infiltration of the liver or kidney. There was clear tumor infiltration of the IVC and thrombus extending from the suprarenal cava to the level of the hepatic veins (Figure 2A).

The liver was mobilized off of the right diaphragm and IVC. The right kidney was spared by dissecting the tumor from the adrenal, with preservation of the right renal vein. The adrenal mass was completely mobilized from the retroperitoneum. Total vascular exclusion of the liver was performed by circumferential dissection of the infrahepatic suprarenal cava, and the suprahepatic vena cava just below the hepatic veins. Vascular clamps were positioned at the supra and infrahepatic caval positions, ensuring the suprahepatic clamp was placed superior to the thrombus. The right adrenal gland and IVC was removed en bloc (Figure 2B). A polyester composite graft (Hemashield Gold 22 mm- Maquet Getinge Group) was used to reconstruct the IVC in an end-to-end fashion (Figure 2C–D). Once the suprahepatic caval anastomosis was completed, the clamp was moved below the hepatic veins to allow for early reperfusion of the liver and to aid with overall hemodynamics.

Pathology was consistent with an 18-cm, 928-g right ACC with IVC and liver capsule invasion. The margins were negative, with no evidence of perineural invasion. The patient did well postoperatively with no immediate complications and was discharged on a steroid taper. She commenced treatment with mitotane as an adjuvant therapy and therapeutic lovenox for treatment of her pulmonary emboli.

DISCUSSION

Adrenal cortical carcinoma is an extremely rare disease, whose incidence has been reported at 0.78 per million, though other sources may report a higher incidence. It has been noted to be more prevalent in women, occurring either in childhood or in the fourth or fifth decade. They may be glucocorticoid, androgen, estrogen, or aldosterone producing. However, in rare cases these masses may not produce hormones, appear clinically inactive, and are found incidentally. Symptoms of inactive tumors may include abdominal or back pain, fever, weight loss, or may have no apparent clinical symptoms associated with their presence.

Diagnosis of ACCs involves both hormonal work up and imaging studies. If glucocorticoid production is suspected, the following should be performed: dexamethasone suppression test, 24 hr urinary cortisol, serum cortisol, and serum ACTH. In the case of androgen or estrogen production a serum DHEA-S, 17-OH progesterone, androstenedione, testosterone, and a 17β estradiol should be obtained. In cases of aldosterone

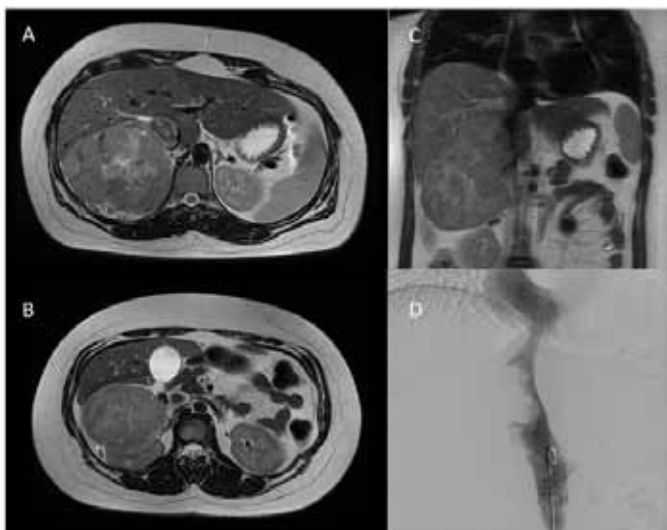


Figure 1: Magnetic resonance imaging demonstrating (A) Right adrenal mass involving the IVC with tumor thrombus, (B) Origin arising from the right adrenal, (C) A coronal view showing IVC extension to the level of the hepatic veins; and (D) A venogram demonstrating IVC thrombus with resulting stenosis.

not amenable to surgical intervention, a biopsy may be required for diagnostic purposes to dictate appropriate medical therapy.

By definition, an ACC with tumor thrombus is classified as a T4 tumor, and stage III cancer. In most cases, these tumors are amenable to surgical resection and come with a high (60–80%) risk of recurrence even with a complete resection. Thrombectomy is performed with the aid of IVC cross clamping, hepatic vascular exclusion, or the use of cardiopulmonary bypass, determined by the margins of thrombus extension. In some cases, replacement of the IVC by use of a vascular graft may be necessary if the thrombus has invaded the vascular wall, or cannot be easily removed, and should be performed if an R0 resection is anticipated. Adjuvant therapy with mitotane is recommended and radiotherapy may be considered. Large vessel invasion independently represents a poor prognostic factor, and confers an inferior disease free survival as well as overall survival. Imaging with CT, and in some cases PET, is recommended every three months for at least two years. Overall prognosis is poor with a 5-year survival rate of <35% in complete resections, and <10% in incomplete resections. Median survival for stage III cases with large vessel invasion are thought to be approximately 9 months, with a significantly decreased disease specific survival with the presence of tumor thrombus.

Surgical resection is the current standard of care in the treatment of ACC, with extent of resection dictated by local invasion of surrounding organs and large vessel invasion. A small number of case reports, case series, and reviews have described resection of ACCs with caval extension and caval tumor thrombus requiring cavotomy or limited IVC resection with emphasis on radical resection and requiring cardiac or veno-veno bypass. While long term prognosis of these progressed ACCs remains poor due to delayed diagnosis, frequent presence of advanced disease, and lack of effective adjuvant treatment, the best management available is complete surgical resection.

This case illustrates the use of radical resection with removal of suprahepatic vena cava tumor thrombus above the level of the hepatic veins and composite graft placement without the use of bypass.

CONCLUSION

An R0 resection and large-vessel venous reconstruction of these extensive adrenocortical carcinomas can be achieved without the aid of cardiac or veno-veno bypass. Imaging is imperative in pre-operative planning, and a multi-disciplinary team approach may be necessary.

Author Contributions

Hoylan T. Fernandez – Substantial contributions to conception and design, acquisition of data, analysis and

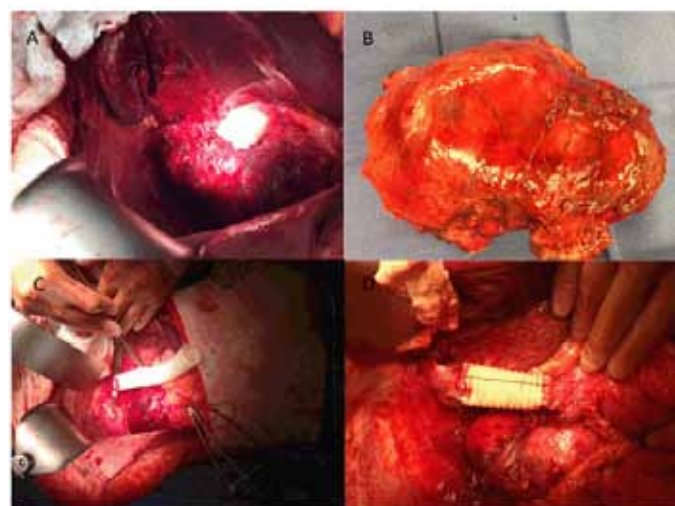


Figure 2: Surgical photos: (A) Right adrenal tumor in situ, with liver rotated medially to expose the IVC, (B) Right adrenal tumor after removal, (C) Composite graft reconstruction of the IVC due to tumor involvement, and (D) Completion of IVC graft reconstruction.

production serum potassium, and an aldosterone/renin ratio are required. A pheochromocytoma should be excluded in the workup. In regards to imaging, CT or MRI offer high specificity, and a PET scan may be helpful in identification of metastatic disease. The presence of tumor thrombus may require Doppler ultrasound, or echocardiography to define the margins of extension. Pursuing a biopsy is discouraged as hormonal work up and imaging are sufficient for diagnosis, and may create the risk of metastasis. However, if the mass is

interpretation of data, drafting the article, revising it critically for important intellectual content, final approval of the version to be published

Peter T. W. Kim – Analysis and interpretation of data, revising it critically for important intellectual content, final approval of the version to be published

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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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