

Surgical resection of a virilizing adrenal mass with extensive tumor thrombus

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Adrenocortical carcinoma with tumor thrombus and concomitant testosterone production is a rare entity. We describe a case of a 53-year-old woman with a testosterone producing left-sided adrenocortical carcinoma with tumor extending to the right atrium and tumor embolus to the right pulmonary artery. To our knowledge, there exist

no such reported cases in the medical literature. We describe our use of techniques derived from transplant surgery for the removal of this mass. Critical components for successful resection included early renal artery ligation, hepatic mobilization off the inferior vena cava, and minimization of cardiopulmonary bypass time thus eliminating the need for deep hypothermic circulatory arrest.

Key Words: adrenocortical carcinoma, tumor thrombus, cardiopulmonary bypass, paraneoplastic syndrome

Introduction

Adrenocortical carcinoma (ACC) is a rare malignancy with an incidence of approximately 1 per million population.¹ ACC often presents at advanced stages and has a 5 year disease-specific survival of 10%-80% depending on prognostic factors.² At the time of diagnosis, up to 75% of tumors will be hypersecreting^{3,4} and many will present with vascular invasion and tumor thrombus.²

Complete primary surgical extirpation is the mainstay of treatment for ACC with tumor thrombus and has been shown to improve survival.⁵ However, surgery for large adrenal masses with tumor thrombus extending to inferior vena cava (IVC) and right atrium (RA) may be associated with significant morbidity and mortality. At our institution, we incorporate organ transplantation techniques allowing for complete abdominal organ mobilization and minimization of cardiopulmonary bypass (CBP) time.⁶⁻⁸ We present the successful surgical management without the need

for deep hypothermic circulatory arrest (DHCA) of a large, virilizing, left-sided ACC with tumor extending to the RA and tumor embolus to the right pulmonary artery (RPA).

Case report

Patient

A 53-year-old woman presented with a several month history of hirsutism, deepening voice and increasing abdominal distension with left flank pain. Abdominal ultrasound showed a large left flank mass and blood tests revealed a testosterone level of > 900 ng/dL. Further investigations with computed tomography demonstrated an 18 cm left-sided adrenal mass with tumor thrombus extending into the retro-hepatic IVC, RA, and RPA, Figures 1a, 1b. Given the significant risk of sudden cardiac death and lack of distant metastasis, the decision was made to perform primary surgical extirpation of all gross tumor.

Surgical technique

Through a chevron incision, the pancreas, spleen, and colon were mobilized *en bloc* and rotated medially as previously described.⁸ The adrenal tumor and left kidney were then dissected free allowing posterior access to the left renal artery for early ligation.

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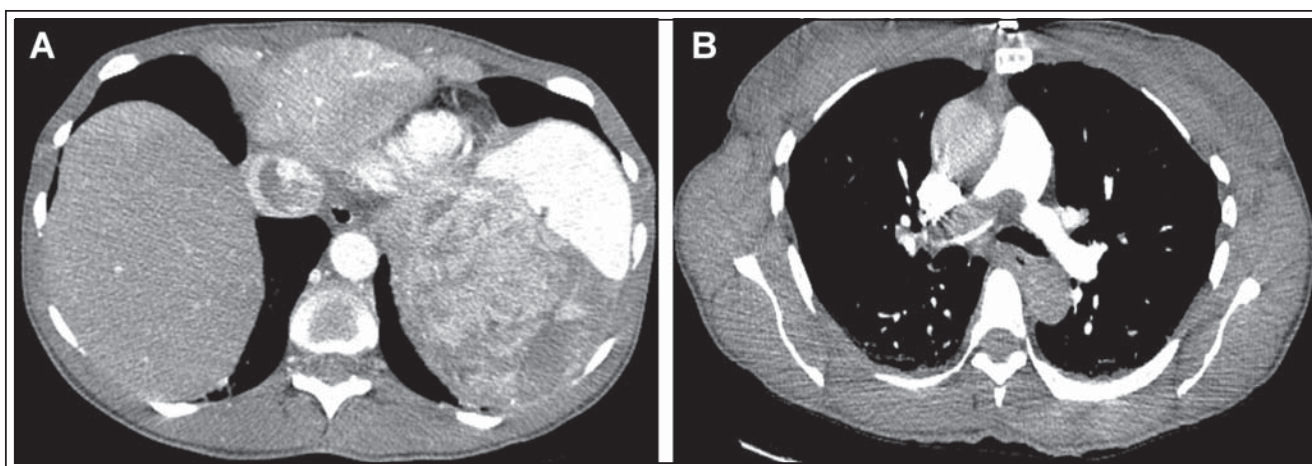


Figure 1. (a) CT scan showing a large left-sided adrenal mass with tumor thrombus in the inferior vena cava and (b) tumor embolus in the right pulmonary artery.

Transesophageal ultrasound (TEE) was employed throughout the surgery to ensure stability of the thrombus in the RA and RPA.

A sternotomy incision was made and canulas were placed in the ascending aorta and superior vena cava in preparation for CPB once the IVC dissection was to begin. Liver mobilization was performed with division of the supporting ligaments and was rolled to the left using techniques previously described for liver

transplantation.⁹ Mobilization was further performed by dividing the small hepatic veins to the caudate and right lobe of the liver so that all portions of the IVC were completely exposed, Figure 2a.

The patient was placed on CPB without circulatory arrest for removal of the tumor, tumor thrombus and tumor embolus. A Pringle maneuver was performed, and vascular clamps were placed on the right renal and adrenal veins and on the IVC below the tumor

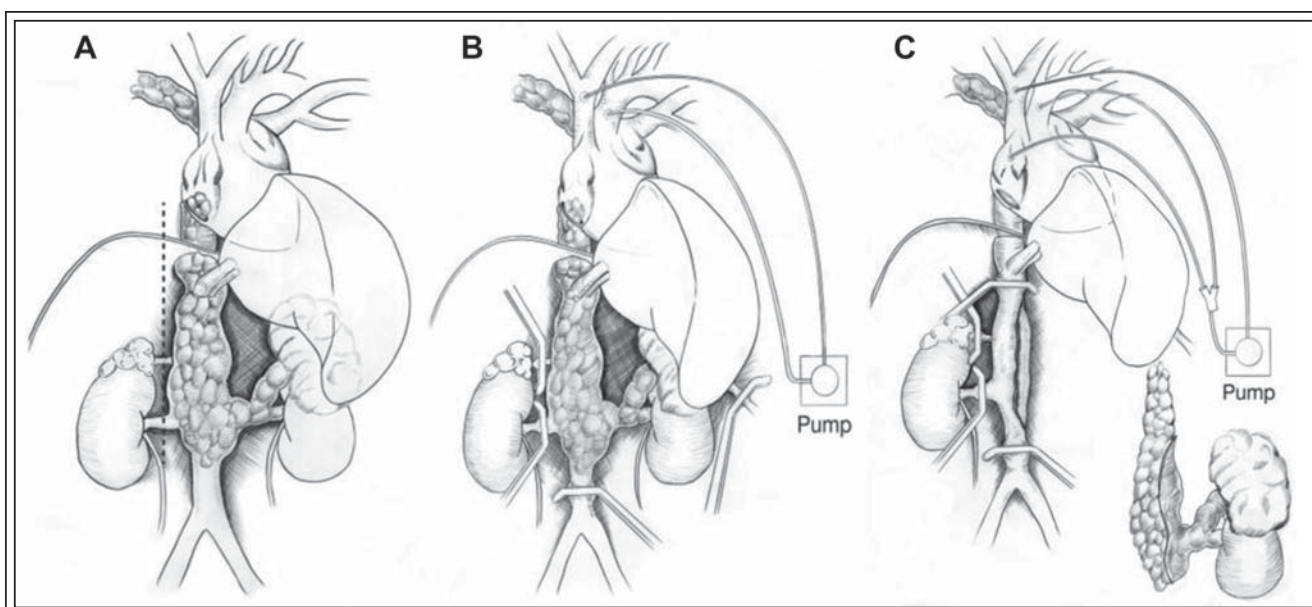


Figure 2. (a) Medial reflection and mobilization of the liver off the IVC is performed after early left renal artery ligation. (b) Vascular clamps are placed on the right renal and adrenal veins and on the IVC below the tumor thrombus. A Pringle maneuver is performed and CPB is initiated. (c) Adrenal tumor and IVC tumor thrombus are removed *en bloc* through an incision in the IVC. A vascular clamp is placed below the major hepatic veins and the Pringle maneuver is released returning blood flow to the liver.

thrombus to minimize venous blood in the IVC, Figure 2b. The IVC was opened longitudinally from the left renal vein to the major hepatic veins. The tumor thrombus was gently “milked” down from the IVC and the RA, Figure 2c, and removed *en bloc* with the adrenal tumor, Figure 3. Simultaneously, the cardiothoracic team placed a third canula in the RA to evacuate residual blood. The main pulmonary artery was opened and tumor thrombus was removed from the RPA. The main pulmonary artery was closed, and CPB was discontinued after 12 minutes. Complete thrombus removal was confirmed by TEE. An IVC filter was placed below the right renal vein and the IVC was closed. A vascular clamp was positioned below the hepatic veins and the Pringle maneuver was released to allow early return of blood flow to the liver.

The patient was extubated on the first postoperative day. Renal function was preserved postoperatively with blood urea nitrogen and creatinine peak values of 30 mg/dL and 1.9 mg/dL. On postoperative day 7, her testosterone level decreased to 21 ng/dL and to 8 ng/dL on day 10. Pathologic evaluation of the tumor

reveled ACC positive for inhibin, CAIX and c-kit, and negative for keratin, CK7 and calretinin.

At 6 weeks after surgery, the patient was started on adjuvant chemotherapy with mitotane. Postoperative imaging at 2 months follow up with positron emission tomography scan revealed two metastatic lesions of the lungs. However, re-imaging at 8 months follow up suggests only a slight increase in the size of one lung lesion while the other remained stable in size. At the time of re-imaging the patient continues on mitotane therapy with good performance status.

Discussion

Historically, ACC presents at advanced stages, which may in part explain its overall poor prognosis.^{3,4} Recently, the staging for ACC underwent a proposed revision in an effort to better align stage II, III and IV tumors with outcomes.² In this system, local invasion, nodal status, tumor thrombus into large veins and metastasis are taken into account. Based on the revised system, survival for advanced tumors with extensive venous tumor thrombus involvement (stage III) demonstrated 50% survival at 5 years if all gross tumor could be removed.

Since Skinner et al¹⁰ reported a survival advantage for aggressive surgical resection of IVC thrombus in renal cell carcinoma, steady improvements in the surgical management of these relatively uncommon tumors have been made. We propose three critical technical components for the safe resection of these tumors: (1) early renal artery ligation, (2) hepatic mobilization off the IVC and (3) minimization of CPB time to avoid DHCA.

Early access to the renal artery is crucial during mobilization and resection of hypervascular retroperitoneal tumors. *En bloc* mobilization of abdominal organs avoids the significantly engorged collateral vessels and allows quicker and easier posterior access to the renal artery. We have shown previously that ligation of the renal artery early in surgery leads to decompression of collateral circulation and to decreases blood loss.¹¹

We propose that liver mobilization is advantageous regardless of the level of tumor thrombus, as it provides excellent exposure of the IVC, renal vasculature and major surrounding organs.⁷ Tumors extending into the IVC above the diaphragm can be safely removed via an intra-abdominal approach by “milking” down the tumor without the need for CPB if excellent exposure of the IVC can be gained. However, in cases of RA and RPA involvement, CPB is necessary. CPB time was minimized in this case to 12 minutes because supradiaphragmatic tumor volume was decreased with removal of the RA tumor through an abdominal

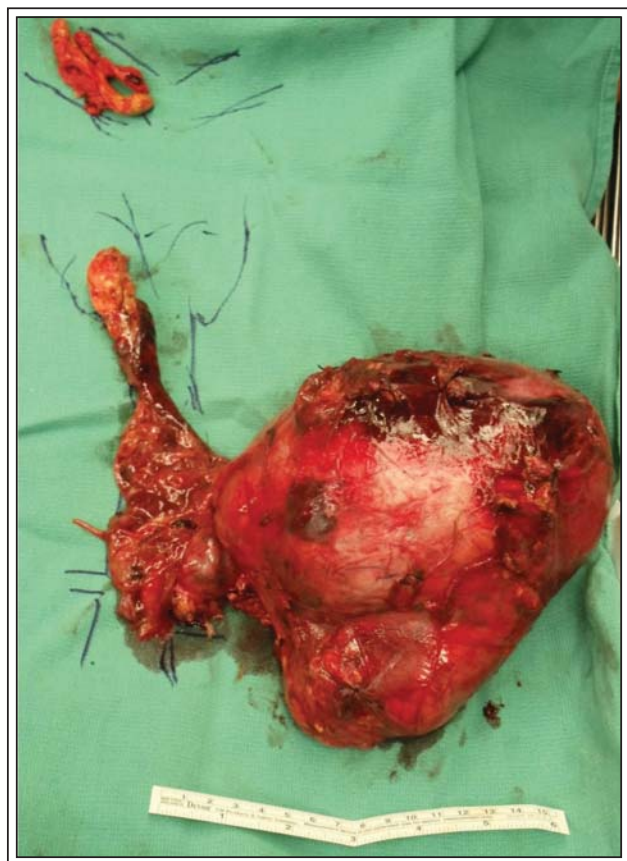


Figure 3. Gross specimen of large adrenal tumor with thrombus.

approach prior to CPB. While some centers propose the use of DHCA, we believe this can be avoided when CPB time is minimal.

To our knowledge, there are no reported cases in the literature of a virilizing ACC with tumor thrombus extending into the RA and tumor embolus to the RPA. Without resection of these tumors, survival is poor with quality of life significantly affected by pain and steroidal excess.

In conclusion, employing the steps described here, ACC with IVC thrombus extending to the RA and RPA can be safely removed. We believe that our approach offers the patient the best opportunity for effective adjuvant therapy and survival. □

References

1. Volante M, Buttigliero C, Greco E, Berruti A, Papotti M. Pathological and molecular features of adrenocortical carcinoma: an update. *J Clin Pathol* 2008;61(7):787-793.
2. Fassnacht M, Johanssen S, Quinkler M et al. Limited prognostic value of the 2004 International Union Against Cancer staging classification for adrenocortical carcinoma: proposal for a Revised TNM Classification. *Cancer* 2009;115(2):243-250.
3. Icard P, Goudet P, Charpenay C et al. Adrenocortical carcinomas: surgical trends and results of a 253-patient series from the French Association of Endocrine Surgeons study group. *World J Surg* 2001;25(7):891-897.
4. Abiven G, Coste J, Groussin L et al. Clinical and biological features in the prognosis of adrenocortical cancer: poor outcome of cortisol-secreting tumors in a series of 202 consecutive patients. *J Clin Endocrinol Metab* 2006;91(7):2650-2655.
5. Hedican SP, Marshall FF. Adrenocortical carcinoma with intracaval extension. *J Urol* 1997;158(6):2056-2061.
6. Delis SG, Bakogiannis A, Ciancio G, Soloway M. Surgical management of large adrenal tumours: the University of Miami experience using liver transplantation techniques. *BJU Int* 2008;102(10):1394-1399.
7. Ekici S, Ciancio G. Surgical management of large adrenal masses with or without thrombus extending into the inferior vena cava. *J Urol* 2004;172(6 Pt 1):2340-2343.
8. Ciancio G, Vaidya A, Shirodkar S, Manoharan M, Hakky T, Soloway M. En bloc mobilization of the pancreas and spleen to facilitate resection of large tumors, primarily renal and adrenal, in the left upper quadrant of the abdomen: techniques derived from multivisceral transplantation. *Eur Urol* 2009;55(5):1106-1111.
9. Ciancio G, Livingstone AS, Soloway M. Surgical management of renal cell carcinoma with tumor thrombus in the renal and inferior vena cava: the University of Miami experience in using liver transplantation techniques. *Eur Urol* 2007;51(4):988-994; discussion 994-995.
10. Skinner DG, Pfister RF, Colvin R. Extension of renal cell carcinoma into the vena cava: the rationale for aggressive surgical management. *J Urol* 1972;107(5):711-716.
11. Ciancio G, Vaidya A, Soloway M. Early ligation of the renal artery using the posterior approach: a basic surgical concept reinforced during resection of large hypervascular renal cell carcinoma with or without inferior vena cava thrombus. *BJU Int* 2003;92(4):488-489.