# Chapter 53
# Case on Adrenal Insufficiency (Addison Syndrome), After Removal of an Adenocarcinoma of the Adrenal Gland

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**Keywords** Adrenal carcinoma • Thrombus caval vein • Addison crisis

## Diagnosis and Indication for Surgery

A 30-year-old female patient was referred to our department because of a tumor of the right adrenal with ingrowth in the inferior vena cava. Since 4 months, she had complaints of tiredness and loss of weight. Recently she had been admitted to the referral hospital because of thoracic pain, edematous legs, and dyspnoea; her complaints were possibly caused by pulmonary emboli. Pulmonary emboli were not found, though, but a tumor of 8 × 5 × 7 cm in the right adrenal with ingrowth into the inferior vena cava (IVC) (Fig. 53.1). There were no distance metastases, but on the CT scan of her abdomen an extensive thrombus was seen in the IVC up to the right atrium and into the right hepatic vein (Figs. 53.2a, b and 53.3a, b). During high fever, a blood culture showed a not-yet specified Streptococcus, but on the endo-ultrasound there were no signs of endocarditis. Moreover, on the PTC a positive tumor and thrombus were found with central necrosis (Fig. 53.4a, b). Hormonal assessment showed normal aldosterone and epinephrines, but high cortisol 0.5 μmol/L, normal 0.03 and 0.28. Diagnosis was established as cortisol-releasing adrenal carcinoma on the right side with thrombus in the ICV. At admission, she was heparinized and an operation plan was made with the cardio surgeon to resect the tumor and thrombus as radical operation.

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M.A. Cuesta, H.J. Bonjer (eds.), *Case Studies of Postoperative Complications after Digestive Surgery*, DOI 10.1007/978-3-319-01613-9_53,

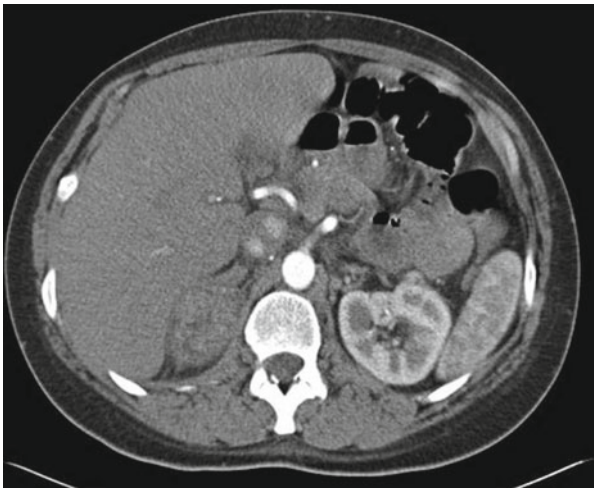

**Fig. 53.1** CT scan showing the tumor in the right adrenal and vena cava

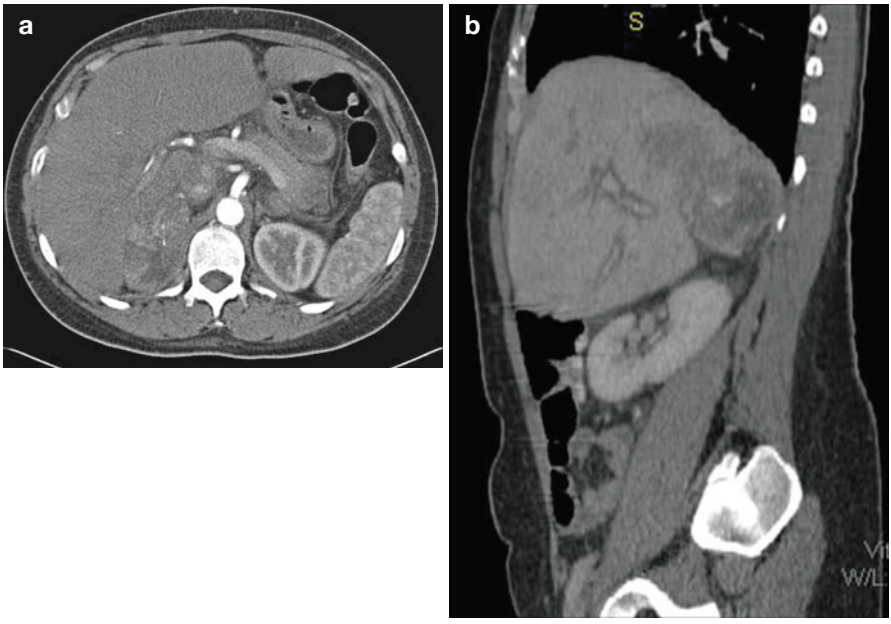


**Fig. 53.2** (**a**, **b**) CT scan showing the tumor in contact with the right hepatic vein

## Operation

At operation, by subcostal right phrenolaparotomy and sternotomy, a resection was performed on the right adrenal and the thrombus in the vena cava after control of the suprahepatic vena cava at the level of the right auricle and proximal of the hepatic veins. Also a Pringle maneuver was done. The tumor and the thrombus were resected en bloc. The thrombus was taken out by a cavotomy. Because of

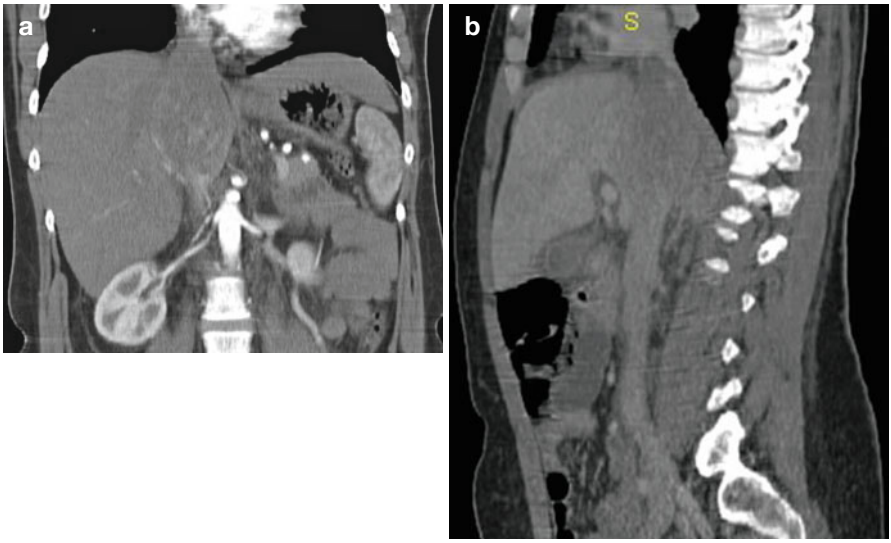


**Fig. 53.3** (**a**, **b**) CT scan showing the tumor with thrombus in the vena cava in direction to right auricle

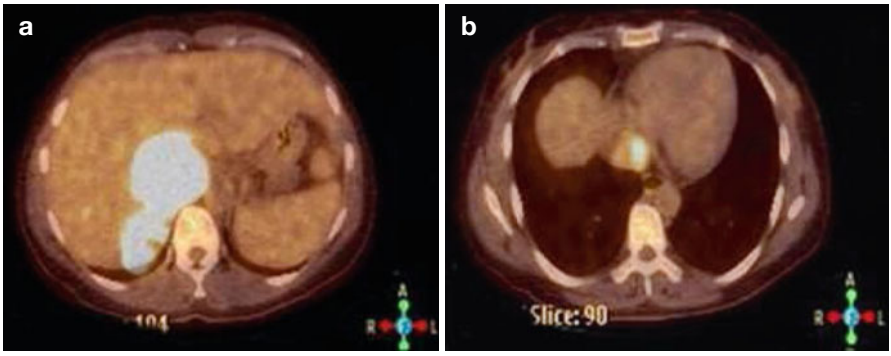


**Fig. 53.4** (**a**, **b**) PET-CT showing avid spot in the tumor and vena cava

ingrowth in the right hepatic vein, a small part was resected and consequently occluded. Pathology showed a radical resected adrenocortical carcinoma with high mitosis index and areas of necrosis and hemorrhage.

## Postoperative Course: Identification and Treatment of Complication

Patient was admitted postoperatively on the IC, where a period of shock occurred and oliguria was observed. Differential diagnosis was made between (a) sepsis (started with iv broad spectrum antibiotics), (b) postoperative bleeding,

(c) thrombosis of the hepatic veins, and (d) presence of Addison crisis (started with hydrocortisone iv). Blood cultures were negative, US showed permeable hepatic veins, and hemoglobin was constant. The Addison crisis was treated by an iv hydrocortisone scheme. Patient went home 11 days after operation with hydrocortisone treatment.

## Pathology

A 7-cm adrenocortical carcinoma (ACC) was found. The tumor was radically resected but showed a high mitotic activity and venous invasion.

## Discussion

Currently, Addison crises are not frequently found in surgical practice. The cause of Addison crisis in this patient is that no corticosteroid scheme was given preoperatively. In spite of a precise diagnosis and preparation of the operation, nobody had taken account of this important point during preparation of patient for surgery. This was especially important in this case because of corticosteroid production of the tumor, the other being adrenal and probably atrophic. Functional variants of ACC have been reported to be more common than the nonfunctional types, and patients mainly present with Cushing syndrome. Recently data suggest that nonfunctional ACCs are more common than the functional types. Adrenocortical carcinoma accounts for approximately 5–10 % of cases of Cushing syndrome and approximately 40 % of patients with both Cushing syndrome and an adrenal mass have an ACC.

Cassinello Ogea et al. described a 70-year-old obese, hypertensive woman taking angiotensin-converting enzyme (ACE) inhibitors and chlorthalidone but with no history of corticosteroid treatment. She underwent a nephrectomy and adrenalectomy under combined general and epidural anesthesia [1]. Severe hypotension with oliguria developed during surgery and persisted during postoperative recovery, with anuria, metabolic acidosis, hyponatremia, and hyperpotassemia. The anuria, metabolic acidosis, hyponatremia, and hyperpotassemia led the authors to consider a diagnosis of an Addison crisis. The patient responded to corticosteroids treatment and low cortisol levels confirmed the diagnosis of adrenal insufficiency.

## Reference

1. Cassinello Ogea C, Giron Nombiela JR, Ruiz Tramazaygues J, et al. Severe perioperative hypotension after nephrectomy with adrenalectomy. Rev Esp Anestesiol Reanim. 2002;49:213–7.