

Rare case adrenocortical carcinoma with tumor thrombus in adrenal vein to inferior vena cava

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Abstract

Adrenocortical carcinoma is a rare neoplasm and difficult to diagnose. This case is an unusual situation meaning that the large adrenocortical carcinoma invaded the liver and the tumor thrombus protruded through the adrenal vein into inferior vena cava. The 38-year-old female presented with Cushing Syndrome (Loss of menstruation, hirsutism, voice change, hypertension, and diabetes mellitus) for 1 year, with no underlying diseases. Operative treatment was performed step by step. The first step was right portal vein embolization. The second step was to explore laparotomy with en-bloc resection of the right adrenal gland tumor, with right hepatectomy with tumor thrombus removal. 1 month after surgery, the patient was discharged without complications, and her condition improved.

Keywords

adrenocortical carcinoma; tumor thrombus; inferior vena cava

Introduction

Adrenocortical Carcinoma (ACC) is a rare neoplasm, with an incidence of 1-2 per million people worldwide [1]. This tumor predominates in bimodal age, mostly in children and men in the fourth and fifth decades of life. ACC is also associated with germline mutation (Li-Fraumeni, Beckwith-Wideman, Multiple endocrine neoplasia type 1, Congenital adrenal hyperplasia, Familial adenomatous polyposis, Lynch Syndrome and carney complex [2]. 50% of adrenocortical carcinoma is non-functioning tumor [3]. Functioning tumors secrete cortisol (30%), androgens (20%), estrogens (10%), aldosterone (2%), or multiple hormones (35%). Patients with a functioning tumor usually present with Cushing's syndrome accompanied by virilizing features. The most important prognosis factor is margin status [4, 5]. ACC with IVC involvement is a controversial risk factor for surgical treatment. This paper describes a successful surgical procedure of ACC with IVC involvement.

Case Presentation

A 38-year-old female, presented with Cushing Syndrome (Loss of menstruation, hirsutism, voice change, hypertension, diabetes mellitus) for 1 year, with no underlying disease. Her first symptom was a loss of menstruation. She then developed hirsutism and voice change. The final symptom presented was

hypertension and diabetes mellitus.

At her first visit, she was consulted in the gynecological clinic due to loss of menstruation and hirsutism. The gynecologist's physical examination didn't find any other abnormalities. She was sent for screening with abdominal ultrasound. The abdominal ultrasound show edanadrenal mass size 12 cm. (**Figure 1**).

She was referred to the general surgery clinic for further evaluation and management. Physical examination showed blood pressure 198/136 mm Hg, pulse rate 123 beat/min., body weight 70 kg. and height 160 cm. Physical examination showed pitting edema +2 but couldn't detect right adrenal mass. Laboratory work included complete blood count, blood chemistry, serum electrolyte, liver function test, cortisol, aldersterone, normetanephrines, total metanephrines, urine

Metanephrines, thyroid function test, HbA_{1c}, sex hormone. Our conclusion was that she had functional adrenocortical carcinoma. She was sent to have a computer tomography scan (CT scan). The CT scan showed a large adrenal mass (size 17 cm.) compressing inferior vena cava (IVC) and right hepatic vein, displacing the right renal vein (**Figure 1,2**). The contrast media could find in IVC the possible thrombosis or compression. We had a multi-disciplinary team to manage this case. We decided to send the patient for investigation by venography to rule out IVC thrombosis or IVC compression from large adrenal mass. The venogram showed that the patient's IVC and tumor thrombus protruded from large right adrenal vein to IVC (**Figure 3**). We planned en-bloc resection in this case but the remnant liver volume of the patient may be insufficient. We discussed with the patient and then sent her to undergo right portal vein embolization. After the right portal vein embolization, we waited 4 weeks to reevaluate her liver volume remnant and condition. The CT scan showed stable size right adrenal mass and increased size left lobe of liver. The operation was performed by en-bloc resection with right hepatectomy, wedge and repaired IVC, preserving the right kidney (**Figure 4-6**). Intraoperative blood loss 4,700 ml. She was admitted for 12 days and discharged without complications.

Discussion

In general, patients with ACC, an R0 resection was associated with 5 years relative survival (RS) 50%, R1 resection 23% and R2 resection 11% 5 year RS [4]. ACC with IVC involvement was first reported in 1972 by Castleman et al [6]. This condition has a controversial prognostic factor, but the most important prognosis factor is margin status [4, 5]. The surgical treatment can be effective management in ACC with IVC involvement [7]. ACC with IVC involvement prognosis is poor, maybe owing to delays in diagnosis, frequent associated metastatic diseases and lack of effective adjuvant treatment [7].

Most ACC patients with non-functioning tumors are difficult to diagnose [3]. They are commonly presented with an enlarged abdominal mass and abdominal or back pain, but rarely with weight loss or anorexia. Several times the ACC presents symptoms when enlarged mass with nearby organ involvement.

In our case, the enlarged ACC with liver invasion, tumor thrombus in IVC, compressed IVC until contrast couldn't be seen in the IVC lumen. Compression IVC total occlusion made it difficult to differentiate between thrombosis or mass effect. We decided to send to for venogram because thrombosis of IVC maybe contraindicative to surgery. In this case after the venogram showed the

patient's IVC lumen and tumor thrombus protruded into IVC lumen we decided to perform en-bloc resection. However liver invasion was the key factor complication after surgery because future liver remnant may be inadequate. This patient was sent for right portal vein embolization and had to wait for 4 weeks to reevaluate her liver volume with staging tumor by CT scan. The CT scan after portal vein embolization showed adequate future liver remnant. We will further evaluate future liver remnant with indocyanine green test (ICG). In this case ICG is normal. We operated on this case by en-bloc resection with right hepatectomy, wedge and repaired IVC, and preserved her right kidney. The patient was discharged without any complications.

In conclusion, ACC is rare. The prognosis depends on margin status. ACC with IVC involvement is not contraindicative to surgery and needs further investigation for decision making. To accomplish this condition, a multi-disciplinary approach team is required, such as general surgeon, vascular surgeon, cardiothoracic surgeon, transplant surgeon, oncologist.

Figures

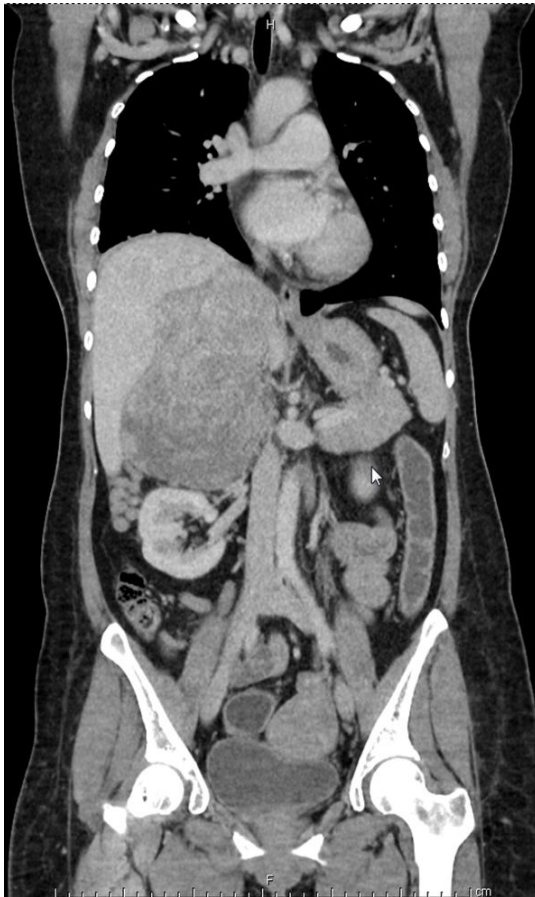


Figure 1: Coronal view CT scan shown large right adrenocortical carcinoma invade right lobe liver and obliterate inferior vena cava and right kidney displace position, several collateral vein lateral side of right kidney and adrenocortical carcinoma



Figure 2: Axial view CT scan shown adrenocortical carcinoma invade right lobe liver segment VI/VII with obliterate right hepatic vein

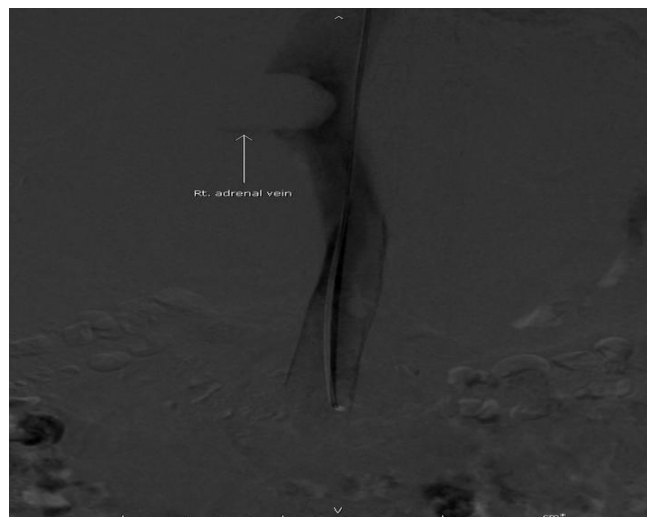


Figure 3: Venogram shown tumor thrombus protrude into inferior vena cava pass through large right adrenal vein, Inferior vena cava still patent lumen



Figure 4: Posterior view of ACC with en bloc resection with right hepatectomy and wedge IVC

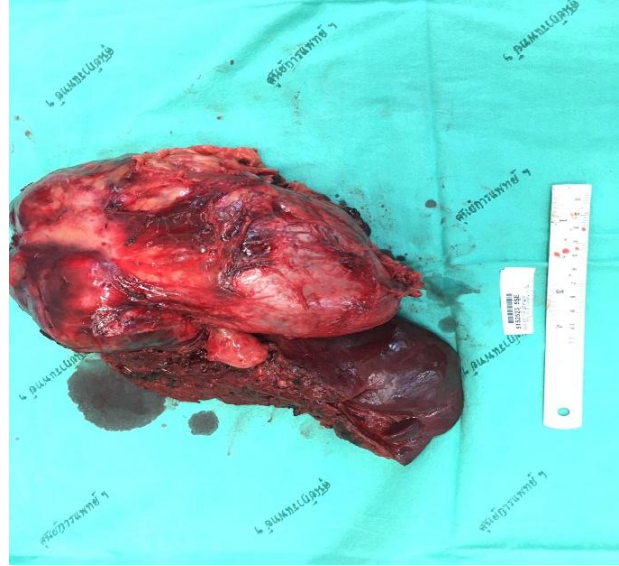


Figure 5: Lateral view of ACC with en bloc resection with right hepatectomy and wedge IVC

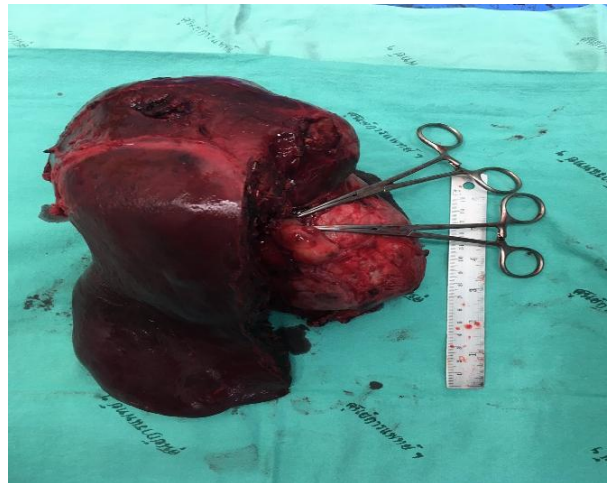


Figure 6: Anterior view of ACC with en bloc resection with right hepatectomy and wedge IVC

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