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CASE REPORT

Surgical resection of adrenocortical carcinoma with invasion into the inferior vena cava: a case report and literature review

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Key Clinical Message

Adrenocortical carcinoma (ACC) is a malignant endocrine tumor. Moreover, ACC with invasion into the inferior vena cava is rare. Early diagnosis and treatment are crucial for such cases. Radical surgical resection is the key therapeutic option in ACC.

Keywords

Adrenocortical carcinoma, inferior vena cava, invasion, surgical resection.

Introduction

Adrenocortical carcinoma (ACC) is a rare and highly aggressive malignancy with an incidence rate of 0.7–2.0 per million per year [1, 2]. ACC is mostly presented in advanced stages with poor prognosis as there are limited treatment options. According to the clinical guideline for the surgical management of ACC, extension of ACC to the adrenal, renal vein, or inferior vena cava (IVC) occurs in approximately 25% of the patients with ACC [3]. Owing to the low incidence of ACC, tumor invading into IVC is still regarded as an uncommon case. The mainstay of treatment for these cases is surgery. However, there are no randomized controlled trials on the treatment of ACC with invasion into IVC. Current knowledge is based on retrospective studies and expert opinion. Hence, we report a case of ACC with invasion into IVC in a young patient who was successfully treated by radical surgical resection.

Case Report

A 23-year-old male with new-onset abdominal pain was transferred to our hospital. No specific abnormal physical signs were disclosed in physical examinations. However, the patient provided us with a report of abdominal ultrasonography from a community hospital. The report showed the presence of a giant retroperitoneal mass.

Laboratory workup revealed the following results: hemoglobin 126 g/L (120~160 g/L), aspartate transaminase 25 U/L (15~40 U/L), alanine aminotransferase 18 U/L (9~50 U/L), total bilirubin 20.5 μmol/L (3.4~22.2 μmol/L), serum

creatinine 71 μmol/L (44~133 μmol/L), serum natrium 143 mmol/L (137~147 mmol/L), serum potassium 3.7 mmol/L (3.5–5.3 mmol/L). Comprehensive endocrine workup was unremarkable: free urinary cortisol 334 nmol/day (153.0~789.4 nmol/day), basal serum cortisol 417.89 nmol/L (118.60~618.00 nmol/L), and ACTH 9 pg/mL (0~46 pg/mL), DHEA-S 813.21 ng/mL (690.00~2960.00 ng/mL), 17-OH-progesterone 1.51 ng/mL (0.61~3.34 ng/mL), androstenedione 3.64 ng/mL (1.86~5.44 ng/mL), free testosterone 14.50 pg/mL (6.20~28.10 ng/mL), aldosterone/renin ratio 13 ng/dL per ng/mL/h (<30 ng/dL per ng/mL/h), plasma-free metanephrine 0.19 nmol/L (≤0.50 nmol/L), plasma-free normetanephrine 0.50 nmol/L (≤0.90 nmol/L).

Computed tomography (CT) scan and magnetic resonance imaging (MRI) confirmed the ultrasonography findings. Both CT and MRI revealed a bulky, heterogeneous, and lobulated shape tumor (151 mm × 96 mm × 157 mm) on the right side of retroperitoneum, originating from right adrenal. They also demonstrated the oppression and direct invasion of neighbor organs including the liver, the right kidney, and IVC (Fig. 1A and B). A whole-body bone scan was negative for bone metastases. Chest CT scan showed no sign of metastasis.

With the cooperation of a multidisciplinary team, the patient received radical surgery treatment. The surgical procedure started by the laparotomy from the right upper quadrant. Then, a large adrenal mass was revealed. The mass adhered to the right lobe of liver and it invaded into IVC. The tumor along with the right adrenal gland and a part of the liver was removed, while the right kidney was reserved. With the assistance of cardiopulmonary bypass, segmentectomy of the IVC was performed, and the IVC was reconstructed with vascular prosthesis (Fig. 1C and D). No intra- or postoperative complications were encountered, and no significant bleeding occurred during resection and reconstruction of the IVC. The estimated blood loss was 850 mL. Therefore, the patient received four units of blood transfusion during operation.

The histological evaluation with the Weiss score confirmed the diagnosis of ACC. According to the Weiss score, the morphological features of the tumor in our case met the following criteria: nuclear atypia, necrosis, invasion of venous structure, invasion of the capsule of tumor. The Ki67 index was 15%. R0 resection (microscopically negative margins) was confirmed histopathologically. At a 6-month follow-up period, there is no sign of locoregional recurrence or distant metastases.

Discussion

Adrenocortical carcinoma is a rare and highly aggressive malignancy with an incidence rate of 0.7–2.0 per million per year [1, 2]. Furthermore, ACC with invasion into the IVC is uncommon. Surgery is the main treatment for ACC as it may provide the best chance of cure for patients with ACC [4]. However, ACC is often associated with poor prognosis. The 5-year survival rate of ACC is between 16% and 47%, but it dramatically drops to 5–10% in metastatic cases [5].

Early diagnosis and treatment are vital for patients with ACC. It has been widely accepted that CT or MRI plays a crucial role in the detection and diagnosis of ACC, especially for nonfunctional tumors. Enhanced CT is a useful imaging technique to assess the relationship between the mass and the surrounding organs. MRI is superior to CT in the diagnosis of venous invasion because MRI has a better resolution of soft tissues [5]. Besides, PET/CT is also useful to characterize adrenal lesions suspected of malignancy in difficult cases and to determine the presence of metastatic disease [2]. The adrenal biopsy in the diagnostic workup of patients with adrenal masses is not recommended as it may increase the risk of tumor dissemination [6].

The final diagnosis of ACC should be confirmed pathologically. It is now well established that the Weiss score is the cornerstone of pathological diagnosis. Weiss score includes nine criteria of proliferation, nuclear abnormality, and tumor extension. Tumors with a Weiss score ≥3 are considered malignant [5]. Our case satisfied four out of the nine criteria: nuclear atypia, necrosis, invasion of venous structure, invasion of the capsule of tumor. Furthermore, Ki67, the proliferation marker, has raised attention for its use in the differential diagnosis of adrenal tumors. The general agreement is that ACCs have a Ki67 labeling index of ≥10%. In addition, Ki67 is the most useful prognostic marker in both localized and advanced ACC [7]. Recently, in a large cohort of patients with localized ACC identified from the German ACC registry, Ki67 provided the best prognostic value for recurrence-free survival [8]. In our case, Ki67 index was 15%, which may indicate a poor prognosis and the great possibility of recurrence.

Surgery is the key therapeutic option in ACC. In the clinical guideline for ACC, it is recommended that adjacent organs should be resected en bloc if they are suspected to be invaded [3]. Owing to the low incidence of ACC with invasion into the IVC, current knowledge is based on retrospective studies and expert opinion. Only case reports or series described such rare pathological condition. Osman et al. [9] reported six cases with adrenal tumors and venous thrombosis. All of them underwent open surgery. Unfortunately, surgical outcome of these cases remained poor. Chiche et al. described their experience in a series of 15 patients, suggesting that surgical treatment could be effective for management of ACC with extension into the IVC [10]. Laan et al. [11] performed a retrospective review of 28 patients of ACC with

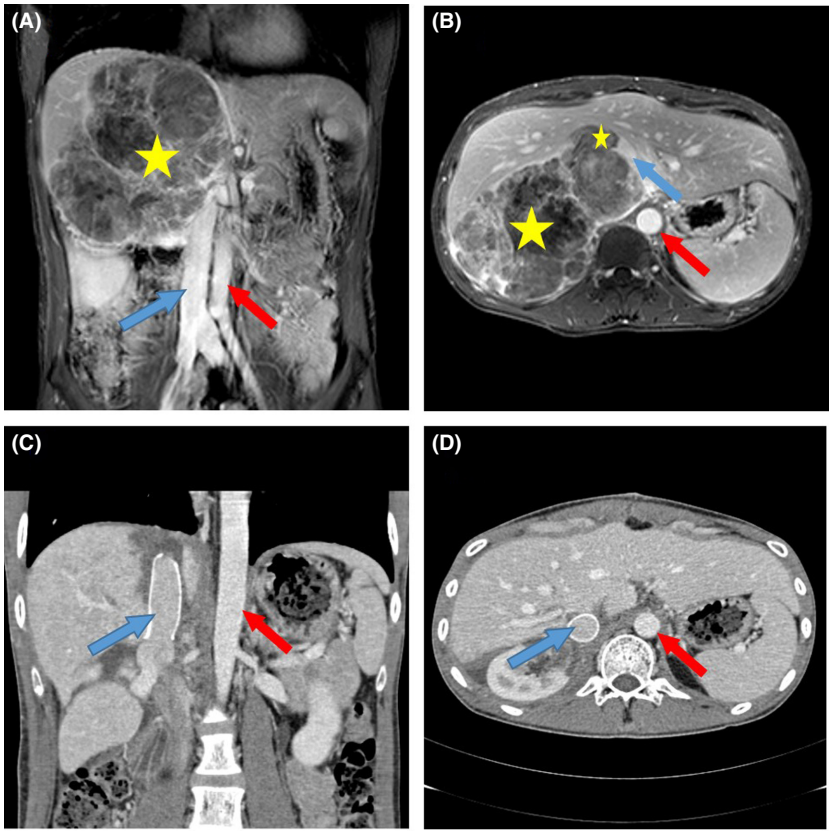

Figure 1. (A and B) Preoperative MRI revealed the bulky and heterogeneous adrenal tumor (151 mm × 96 mm × 157 mm, yellow pentagram) oppressing adjacent organs and the invasion into inferior vena cava (blue arrow), while abdominal aorta was normal (red arrow); (C and D) postoperative CT scan demonstrated the reconstructed inferior vena cava with vascular prosthesis (blue arrow) after radical resection of the tumor, while the abdominal aorta was normal (red arrow).

IVC tumor thrombus during a 30-year period. They drew a conclusion that complete resection of ACC with IVC tumor thrombus seemed justified as the survival was similar to the patients without IVC tumor thrombus. The clinical guideline for ACC recommends that the surgery should be performed by surgeons with expertise in adrenal surgery (open and laparoscopic) and with a volume of more than 15 adrenalectomies per year (benign and malignant) [3]. In our case, the patient underwent radical surgery and reconstruction of the IVC with the use of cardiopulmonary bypass. R0 resection was confirmed pathologically.

In our case, the patient was managed by a multidisciplinary team with extensive experience in adrenal tumors. The multidisciplinary team was consisted of endocrinologists, urologists, hepatobiliary surgeons, cardiovascular surgeons, pathologists, and radiologists. As a matter of fact, the multidisciplinary team provided high-level care for the patient. It improved coordination, communication, and decision-making between team members. Endocrinologists and radiologists played a role in the preoperative assessment. Urologists, hepatobiliary surgeons and cardiovascular surgeons were responsible for the radical resection and reconstruction of the IVC. The diagnosis of ACC with invasion into the IVC was confirmed by pathologists finally. Owing to the close cooperation of a multidisciplinary team, the patient recovered from the surgery without recurrence or metastases of ACC at a six-month follow-up period.

In conclusion, ACC with invasion into the IVC is a rare pathological condition. Early diagnosis and treatment are crucial for ACC. With the coordinated efforts of a multidisciplinary team, excellent short-term outcome could be achieved through radical surgical resection. However, further study is necessary to evaluate the long-term outcome.

Authorship

MJ: clinically managed the patient, reviewed literature, and wrote the article draft. HD: wrote and revised the manuscript. CL: provided the figures and revised the

manuscript. KX and JT: involved in laboratory data collection and prepared the manuscript. YG and SZ: reviewed the manuscript.

Conflict of Interest

The authors have no conflict of interests to disclose.

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