

CASE REPORT

Vena Cava-Atrium Anastomosis Liver Transplantation on Patient with Budd-Chiari Syndrome Caused by a Tumor Thrombus in the Inferior Vena Cava: A Case Report

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Abstract: Owing to the difficulty in removing hepatocellular carcinomas associated with extrahepatic metastases and the fact that they may further metastasize along the vessels, liver transplantation is not currently recommended for patients with these conditions. We report a case of Budd-Chiari syndrome (BCS) caused by hepatic carcinoma and tumor thrombus in the inferior vena cava (IVC). Backed by the strong desire of the patient to undergo liver transplantation, we decided to perform vena cava-atrium anastomosis liver transplantation (VCAALT) on the patient after a comprehensive evaluation of the patient's condition. The patient recovered quickly and was discharged after 1 month of consolidation chemotherapy. BCS caused by an IVC tumor thrombus is a rare clinical case, and selecting an appropriate treatment is difficult. In this case, we successfully solved the clinical problem of BCS in a patient with hepatocellular carcinoma combined with an IVC cancer embolus using VCAALT.

Keywords: Hepatic carcinoma, Inferior vena cava tumor thrombus, Budd-Chiari syndrome, Vena cava-atrium anastomosis liver transplantation

1. Introduction

Budd-Chiari syndrome (BCS) is a rare disease characterized by an obstruction of the hepatic vein outflow tract. While BCS can be caused by various etiologies, thrombosis remains the main cause^[1,2]. Regardless of the cause, BCS has serious clinical sequelae as the pressure in the sinusoids increases^[3]. Current treatments for BCS include anticoagulation, angioplasty, stent implantation, angioplasty of the inferior vena cava (IVC), a transjugular intrahepatic portosystemic shunt (TIPS), and liver transplantation^[4]. In most cases, BCS can be effectively managed, and only 10–20% of patients need liver transplantation^[5]. Different treatments for patients with BCS caused by hepatocellular carcinoma combined with tumor thrombus in the IVC have been reported, but there is still no optimal therapy due to a scarcity of data attributed to BCS rarity (3–4%)^[6–8]. Liver transplantation seems to be the only radical cure, although hepatectomy combined with IVC thrombectomy is associated with a fatality rate of 15%^[9]. Herein, we report a case of utilizing vena

cava-atrium anastomosis liver transplantation (VCAALT) on patients with BCS caused by a tumor thrombus in the IVC. This therapeutic approach successfully resolved BCS and highlights its efficacy of liver transplantation on hepatocellular carcinoma associated with IVC tumor thrombus.

2. Case presentation

A 57-year-old man was admitted to the hospital on June 8, 2018, due to abdominal distention, lower extremity edema, and hematochezia that had persisted more than 2 months. He had a 30-year history of hepatitis B viral infection. After performing relevant examination and evaluation, our diagnoses regarding his condition were hepatitis B virus-related decompensated liver cirrhosis, hepatocellular carcinoma coupled with a tumor thrombus in the IVC, and BCS. The consent of the patient and his family was obtained for this case report.

Several tests were performed. The peripheral white blood cell count was $2.82 \times 10^9/L$, red blood cell count $2.56 \times 10^{12}/L$, hemoglobin 71 g/L, and platelet count $78 \times 10^9/L$. The level of serum alanine aminotransferase was 65 U/L, total bilirubin 91 $\mu\text{mol/L}$, albumin 33.7 g/L, and potassium concentration 3.27 mmol/l. HBsAg, HBeAb, and HBcAb were confirmed to be positive, and the quantity of hepatitis B virus DNA detected was <500 IU/mL. The alpha-fetoprotein was $>20,000$ ng/ml. The prothrombin time (PT) was 14.4 s, the international normalized ratio 1.32, and the PT activity percentage 63%. Enhanced computed tomography (CT) scans of the abdomen showed two large, mixed-density masses ($22 \text{ cm} \times 18 \text{ cm} \times 13 \text{ cm}$, $19 \text{ cm} \times 11 \text{ cm} \times 5 \text{ cm}$) in the right lobe of the liver and a possible tumor thrombus in the IVC (Figure 1A).

The patient had a Child-Pugh score equivalent to Class A. Since the liver tumor was too large to be removed and the fact that vascular metastasis, rather than lymph node metastasis, was present, the patient was first treated with transcatheter arterial chemoembolization after admission. Although the patient and his family members were informed that the condition at the time (i.e., tumor was too large) was unamenable to liver transplantation, they insisted to have liver transplantation as treatment and were willing to take risks of the surgery.

Conventional liver transplantation cannot solve the clinical problems of patients with BCS caused by IVC tumor thrombus. After pre-operative evaluation, we decided to adopt VCAALT on this patient. This procedure requires an anastomosis of the donor's superior vena cava to the recipient's atrium and an end-to-side anastomosis of the donor's IVC (upper pole of the right kidney) after resecting the recipient's retrohepatic IVC. Intraoperatively, we observed that the IVC was filled with tumor thrombus from the plane of hepatic vein to its entrance at the atrium. By opening the diaphragm and pericardial cavity, we blocked the IVC along the right atrium and the first

porta hepatis. A venous bypass was performed using extracorporeal membrane oxygenation. The retrohepatic IVC was cut along the edge of the right atrium. After resecting the diseased liver, the retrohepatic IVC of the donor's liver was sutured continuously with the recipient's right atrium, and the subhepatic IVC of the liver was anastomosed continuously end-to-side to complete liver transplantation (Figures 1B and 2A).

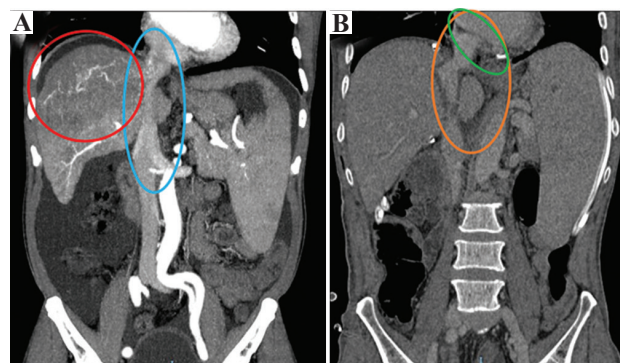


Figure 1. Enhanced computed tomography (CT) scans of the abdomen before and after liver transplantation. (A) Before liver transplantation: the location of hepatocellular carcinoma (red circle) and the inferior vena cava filled with tumor thrombus (blue oval). (B) After liver transplantation: the CT scan shows the inferior vena cava with unobstructed blood flow after transplantation (orange oval) and the anastomosis of inferior vena cava and atrium (green oval).

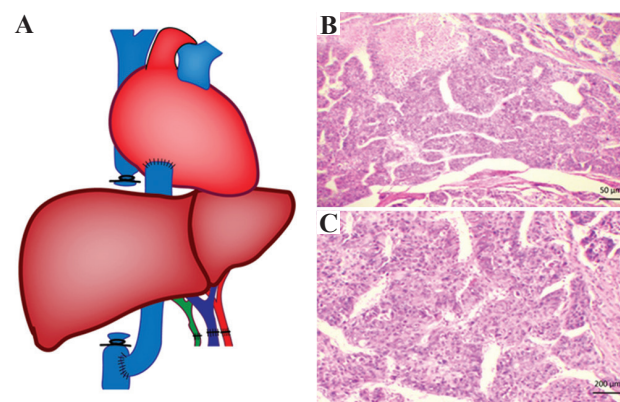


Figure 2. (A) Graphic illustration of vena cava-atrium anastomosis liver transplantation (VCAALT). (B & C) Histopathological examination of hepatocellular carcinoma (grade II) tissues with hematoxylin and eosin staining. Scale bar: 50 μm (B) and 200 μm (C).

The post-operative pathological examination confirmed that the tumor was a moderately differentiated hepatocellular carcinoma and revealed a tumor thrombus of IVC (Figure 2B and C). The patient was able to get out of bed 3 days after the operation and returned to normal diet after 1 week, the liver function returned to normal after 2 weeks. However, 2 months after the operation, the levels of hepatocellular carcinoma-related markers were elevated, and abdominal CT scan indicated recurrent mass formation

in the transplanted liver. The above results suggested that hepatocellular carcinoma recurrence occurred in the transplanted liver. Targeted chemotherapy with sorafenib was used. Finally, the patient was discharged after 1 month of consolidation chemotherapy.

3. Discussion

The clinical manifestations of BCS vary greatly from patient to patient, and its treatment follows the principle of a progressive treatment strategy, including anticoagulation, thrombolysis, percutaneous recanalization, transjugular intrahepatic portal shunt (TIPS), surgery, and liver transplantation^[10]. However, anticoagulant therapy is of no use for treating IVC tumor thrombus. TIPS is not always technically feasible because the entire IVC is blocked by the tumor^[11]. Since almost all the cases after IVC thrombectomy can relapse, this technique does not seem to fundamentally resolve the IVC obstruction^[12]. As a last resort for the treatment of BCS, the traditional liver transplantation cannot resolve the BCS caused by IVC tumor thrombus; therefore, the VCAALT is considered an ideal liver transplantation technique.

VCAALT is mainly used in BCS patients with a posterior hepatic vena cava plaque or an extensive thrombus spread to the lower atrial border. The key point of this surgical approach was to block the vena cava from the lower atrium to the upper part of the right kidney and remove a segment of the vena cava. The superior vena cava of the donor's liver was anastomosed with the recipient's atrium, and the donor's liver is anastomosed end-to-end with the recipient's IVC^[13]. Compared with traditional liver transplantation, VCAALT not only avoids the risk of thrombus shedding but also alleviates the complications of IVC tumor thrombus obstruction. It has been reported that the 5-year survival rate after VCAALT can reach 80%, and this technique is also associated with low post-operative BCS recurrence rate and significant clinical efficacy^[14]. Most reports mainly focus on patients with BCS caused by vena cava thrombosis, but reports regarding patients with vena cava tumor thrombus are rare.

At our center, VCAALT has been performed in patients with BCS caused by IVC tumor thrombus. This technique not only resolves the primary liver cancer of the patients but also improves the related complications caused by IVC tumor thrombus, highlighting the uniqueness and superiority of VCAALT in the treatment of liver cancer complicated by BCS. Although the hepatocellular carcinoma status of the patient in this case made him unamenable to liver transplantation, the patient and his family were willing to take the related risks after liver transplantation. The post-operative average survival time for the patients who have had a successful VCAALT in our center is extended to about 1 year. Combined with the

above factors, this operation was performed.

We hereby made some operation recommendations for VCAALT:

- (1) For patients with ortho-sthenic body type, the right atrium and vena cava can usually be exposed from the mediastinum in the middle of the sternum, while for patients with sthenic body type, the mediastinum can be exposed directly from the diaphragm. In children, a diaphragmatic incision is generally performed to expose the right atrium and vena cava.
- (2) The length of the donor's hepatic vena cava that is anastomosed with the recipient's right atrium must be sufficient (uf cm), and a vascular bridge can be installed if necessary. High vascular tension should be prevented from affecting the heart. After performing the vena cava-atrium anastomosis, the diaphragmatic incision and the graft ligaments must be sutured and fixed, making sure that there is no vascular tension during atriotomy.
- (3) When the atrial IVC is blocked, there is no need to perform a bypass operation if the hemodynamics are stable. Thus, a simple diversion from the iliac vein to the jugular vein should be performed.
- (4) A drainage tube is placed after the atrial anastomosis to prevent the hematocoele from affecting blood reflux, and the catheter is removed 48–72 h after the operation without drainage^[15].

4. Conclusion

This case report demonstrates the feasibility and superiority of using VCAALT as a treatment of BCS secondary to tumor thrombus in the IVC, thereby improving the prognosis.

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Conflicts of interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

Author contributions

All authors read and approved the final manuscript.

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