The first case of combined heart-liver transplantation in a patient with alveolar echinococcosis

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June 22, 2020

Abstract

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Combined heart-liver transplantation

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Funding statement: The work was not supported by any sources

Abstract: We report a rare case of liver alveolar echinococcosis with an invasion of the hepaticocaval confluence, inferior vena cava, pericardium, right atrium, atrial septum, and superior vena cava, and its successful treatment by combined heart-liver transplantation.

Keywords: alveolar echinococcosis, vascular invasion, combined heart-liver transplantation.

Introduction: The first combined heart-liver transplantation (CHLT) was performed in a 6-year-old girl with familial hypercholesterolemia and secondary heart failure in 1984 (1). From 1988 to February 2020, 329 cases of CHLT have been recorded in the United Network for Organ Sharing (2). Indications for CHLT include cardiac cirrhosis, concurrent liver-heart diseases, and amyloidosis, but not alveolar echinococcosis (AE) of the heart (3). AE is a helminthiasis caused by the *Echinococcus multilocularis* larvae, which are retained by the liver while passing into the portal bloodstream from the bowel and form tumor-like neoplasms. Treating

patients with liver AE is difficult as it shows signs of a slowly growing malignant tumor with infiltrative growth, possible invasion of adjacent organs, and ability to metastasize (4). Endemic hotbeds are Western Siberia, Far East, Yakutia, Central Asia, Central Europe, Alaska, and Northern Canada.

CASE REPORT: The patient was a 36-year old woman who experienced severe, colicky pain in the right hypochondrium for about a year. For several years, she had contact with wild animals, mainly foxes, and was involved in processing of fur. She was diagnosed with AE of the liver.

Further examination revealed that in addition to the liver, her heart was affected. Computed tomography showed a parasitic lesion with complete occlusion of the hepaticocaval confluence and inferior vena cava (IVC), narrowing of the right atrium, sinus of the superior vena cava (SVC), and coronary sinus (Fig. 1 A), and a solitary 3-cm AE in the left lung. No brain AE metastases were detected. Echocardiography revealed a new growth in the roof and septum of the right atrium with a clear uneven contour reducing the atrial cavity by approximately 40%-50% and obstructing the mouth of the SVC (Fig. 1 B). As the AE involved the liver, heart chambers, and IVC, isolated liver transplantation was impossible; therefore, CHLT was performed.

The patient was in the waiting list for 76 days. At the time of transplantation, she had ascites, severe shortness of breath, and distended veins of the neck.

The procedure first involved a J-shaped laparotomy. The liver showed typical features of Budd-Chiari syndrome. Parasitic lesion was localized in liver segments 1, 2, 3, 4, 7, and 8 and grew in hepaticocaval confluence, retro- and suprahepatic IVC, diaphragm, and pericardium. The liver was completely mobilized, and anteroposterior phrenotomy was achieved. The diaphragm and pericardium were partially resected in the circumference of the IVC, and its supraphrenic segment was encircled from the abdomen (Fig. 1 C).

The next step involved a median sternotomy. The heart was not enlarged. Cardiopulmonary bypass was initiated with peripheral IVC cannulation through the femoral vein, SVC and aorta were cannulated directly. The completely mobilized heart was lowered through the diaphragm into the abdominal cavity and removed *en bloc* with the liver and retrohepatic segment of the IVC (Fig. 2 A, B).

Orthotopic heart transplantation was performed using the standard bicaval technique. IVC cardiac anastomosis formed with the interposition fresh venous allograft (infrarenal IVC from the same donor) between heart and liver which was placed into the abdominal cavity. The heart was reperfused without IVC inflow. Heart ischemia time was 3 hours 50 minutes. Liver was transplanted using the conventional technique. Superior caval anastomosis was formed using interposition venous allograft (Fig. 2 C). Liver cold and warm ischemia time were 4 hours 7 minutes and 41 minutes respectively.

We used the triple immunosuppression protocol based on tacrolimus in combination with mycophenolic acid and steroids. The postoperative period was complicated by bile leakage, which caused anastomotic arterial bleeding and was successfully treated by Roux-en-Y hepaticojejunostomy with hepatic artery reconstruction. The total hospital stay was 4 months. Before discharge, multiple small AE metastases were detected in both lungs and adjuvant chemotherapy with albendazole was initiated. On 1-year follow-up, the patient was in a good condition and lung metastases remained stable.

DISCUSSION: Radical surgery associated with antiparasitic chemotherapy is the only curative option for liver AE. But due to asymptomatic process, many patients have advanced disease with vascular invasion when standard liver resection is unlikely to be feasible. However, in most of these cases the radical treatment may be provided using total vascular exclusion with *in vivo* or *ex vivo*hypothermic liver surgery (5). Liver transplantation is regarded as a possible treatment option for nonresectable AE and can be performed even if the suprahepatic IVC is involved but not the heart.

In conclusion, CHLT is a rare procedure that treats complex and often fatal conditions, and, to date, does not include AE in the indications.

Acknowledgements: None

Conflict of Interest Statement: There are no conflicts of interest.

Author contribution statement: All authors are familiar and agree with the content of the article

Informed patient consent obtained

Figure legends:

Figure 1. (A) Preoperative computed tomography, (B) Preoperative echocardiography, (C) Parasitic lesion in the liver

Figure 2. (A) Liver of the recipient, (B) Heart removed with liver *en bloc* , (C) Transplanted heart and liver.

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