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Ante situm liver resection with inferior vena cava replacement under hypothermic cardiopolmunary bypass for hepatoblastoma: Report of a case and review of the literature

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ABSTRACT

INTRODUCTION: Hepatoblastoma with tumour thrombi extending into inferior-vena-cava and right atrium are often unresectable with an extremely poor prognosis. The surgical approach is technically challenging and might require major liver resection with vascular reconstruction and extracorporeal circulation. However, which is the best surgical technique is yet unclear.

PRESENTATION OF CASE: A 11-months-old boy was referred for a right hepatic lobe mass(90 × 78 mm) suspicious of hepatoblastoma with tumoral thrombi extending into the inferior-vena-cava and the right atrium, bilateral lung lesions and serum alpha-fetoprotein level of 50.795 IU/mL. After 8 months of chemotherapy (SIOPEL 2004-high-risk-Protocol), the lung lesions were no longer clearly visible and the hepatoblastoma size decreased to 61 × 64 mm. Thus, *ante situm* liver resection was planned: after hepatic parenchymal transection, hypothermic cardiopulmonary bypass was started and *en bloc* resection of the extended-right hepatic lobe, the retro/suprahepatic cava and the tumoral thrombi was performed with concomitant cold perfusion of the remnant liver. The inferior-vena-cava was replaced with an aortic graft from a blood-group compatible cadaveric donor. The post-operative course was uneventful and after 8 months of follow-up the child has normal liver function and an alpha-fetoprotein level and is free of disease recurrence with patent vascular graft.

CONCLUSIONS: We report for the first time a case of *ante situ* liver resection and inferior-vena-cava replacement associated with hypothermic cardiopulmonary bypass in a child with hepatoblastoma. Herein, we extensively review the literature for hepatoblastoma with thumoral thrombi and we describe the technical aspects of *ante situm* approach, which is a realistic option in otherwise unresectable hepatoblastoma.

Abbreviations: HLB, hepatoblastoma; IVC, inferior vena cava; FAP, familial adenomatous polyposis; LT, liver transplantation; PV, portal vein; TVE, total hepatic vascular exclusion; UVC, upper vena cava; PRE-TEXT, pre treatment extent of disease; POST-TEXT, post treatment extent of disease; SIOPEL, Société Internationale d'Oncologie Pédiatrique-Epithelial Liver Tumor Study Group.

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1. Introduction

Hepatoblastoma (HBL) is the most common paediatric liver tumour and it occurs usually in the first 3 years of life. The incidence of HBL has increased in the recent years up to 1.5 cases per million, and it is frequently associated with low birth weight or genetic anomalies. The management of HBL has impressively improved due to combined neoadjuvant chemotherapy and liver resection or liver transplantation (LT), increasing the overall 5-years survival rate up to 75% [1]. The risk stratification with the pre-treatment extent of disease (PRE-TEXT) system, the worldwide multicentre trials experience and the multidisciplinary management, improved HBL prognosis and surgical resectability [2]. However, HBL with tumour thrombi extended into the inferior vena cava (IVC), with or without the involvement of the right atrium, may result challenging to define the best surgical technique.

Different surgical procedures, mainly reported in adults, have been proposed for liver tumour with IVC infiltration, including total hepatic vascular exclusion (TVE) [3]. These techniques are effective to control haemorrhage and air embolism during liver resection, but may cause severe hepatic ischemia/reperfusion injury, hemodynamic instability and potential renal injury. Recently, liver resection under hypothermic liver perfusion with cytoprotective solution (including *in situ, ex situ* or *ante situm* techniques) has been proposed for preventing ischemic liver injury [4]. Yet, no experience of *ante situm* liver perfusion associated with hypothermic cardiopulmonary bypass has been reported in children.

Herein we report a successful case of *ante situ* liver resection and IVC replacement under hypothermic cardiopulmonary bypass (CPB), performed in a 21 months-old male with HBL and tumour thrombi into the IVC and right atrium.

The current case has been reported in line with the SCARE criteria [5].

2. Case report

The patient was a 11-months old child referred for right upper quadrant abdominal mass. He was born on term (birth weight of 2.470 kg) and had a familial history of adenomatous polyposis (FAP). At the time of admission, the patient was asymptomatic with normal vital signs, but physical examination revealed hepatomegaly, abdominal bloating and umbilical hernia. The ultrasonography (US) showed a large hepatic mass (10 cm in diameter) in the right liver. Serum alpha-fetoprotein (AFP) level was 50.795 IU/mL. Liver function, coagulation, serum B-human chorionic gonadotropin, blood cell count, thyroid function were within normal limits, except for the evidence of thrombocytosis (805.000/UL). Computed tomography (CT) showed a mass of the right hepatic lobe, 90 × 78 mm in size extending in segment IV, with dyshomogeneity and calcifications. The tumour displaced posteriorly the right kidney, dislocated the aorta and the IVC to the left side, stretching the celiac trunk and the superior mesenteric artery (Fig. 1). Tumoral thrombi was present, extending from the right hepatic vein into the IVC up to the right atrium. Bilateral lung lesions, suspicious for HBL metastases, were found as well. Heart involvement was confirmed by echocardiography, which detected a 2.6 cm echoic mass through the tricuspid valve.

A PRE-TEXT III staging (P0, V3, M1) with lung and atrium-cava metastasis at the outset was defined. The child underwent neoadiuvant chemotherapy (SIOPEL 2004 high risk protocol; cycles A1-3 and cycle B) for 8 months: 3 cycles with cisplatin (70 mg/m^2, 9 doses administered) and doxorubicin (30 mg/m^2, 6 doses); 4 cycles with carboplatin (6 mg/Kg, 4 doses) and doxorubicin (0.83 mg/Kg, 10 doses); and 2 cycles with carboplatin (25 mg/Kg, 2 doses), vincristine (0.05 mg/Kg, 5 doses) and 5-fluorouracil (33 mg/Kg, 6 doses). During the treatment, the child presented transient severe thrombocytopenia and one episode of sepsis successfully treated with antibiotics. After neoadiuvant therapy AFP decreased to 879 IU/mL. CT scan showed size reduction of the HBL (61 × 64 mm), still involving the IVC as the right and middle hepatic vein. A left accessory hepatic artery from the left gastric artery and a replaced right hepatic artery arising from the superior mesenteric artery were documented; lung lesions were no longer clearly visible. Cavography documented retrohepatic IVC infiltration by HBL (Fig. 2). After multidisciplinary team meeting (involving surgeons, oncologist, anaesthesiologist and radiologist), the small patient was proposed for an extended right liver resection, with IVC and intracardiac thrombus removal, which was performed by a senior liver transplantation and hepato-biliary-pancreatic surgeon.

2.1. Surgical procedure

The patient was placed in supine position and the abdomen was explored through a bilateral sub-costal incision with xyphoid extension. There was no evidence of ascites or peritoneal metastasis and intraoperative US documented that the tumour did not involve the left lateral segment of the liver. The Arantius' ligament was dissected and the left hepatic vein was looped. After cholecystectomy, the common bile duct, the right hepatic artery and the anterior and posterior branches of the right portal vein (PV) were ligated and divided. The left PV and the left hepatic arteries were identified and looped. The Rex recess was then exposed and vessels for segment IV were divided. Parenchymal transection, along the line of the falciform ligament, was performed via an anterior approach, using the hanging manoeuvre with "no touch approach" of the tumour. Biliary and vascular structures were divided between clips or tie. Pringle manoeuvre was not used. A vessel loop around the IVC above the renal veins was then placed.

The xiphoid incision was extended up to the jugulum with a median sternotomy and the pericardial sac was opened. After systemic heparinization, the ascending aorta, the upper vena cava (UVC) and the infra-renal IVC were cannulated and clamped, and the extracorporeal circulation with CPB was started. Body temperature was reduced to 28 °C, in order to protect the organs. The diaphragm was incised vertically down toward the suprahepatic IVC and the diaphragmatic veins were divided. After clamping the left hepatic arteries and the PV, the left PV was cannulated though the right PV stump. The left hepatic vein was divided and *ante situ* hypothermic liver perfusion with Celsior solution (4 °C) was started. The liver was further cooled with ice on his surface. After division of the right triangular ligament, an *en-bloc* resection of the extended-right hepatic lobe (segments I + IV-VIII), of the retro- and supra-hepatic IVC and of the neoplastic thrombus (extending from the right hepatic vein to the right atrium) was performed (Fig. 3).

The IVC was reconstructed with a fresh aortic graft from cadaveric donor with identical blood group. The aortic conduit was end-to-end anastomosed with the right atrium (through the diaphragmatic ostium) and inferiorly with the supra-renal IVC by 5/0 prolene continuous running sutures. The neo-IVC was opened immediately below the diaphragmatic ostium and end-to-side triangular anastomosis was performed between the left hepatic vein and the neo-IVC by 6/0 prolene. After 40 min of hypothermic liver perfusion, the portal flush was interrupted. UVC, IVC, aorta, left hepatic arteries and PV were de-clamped, and the left lateral segment was reperfused. The patient was gradually rewarmed and, once hemodynamic stability and good haemostasis were confirmed, the CPB was weaned off, after a total time of 71 min. Roux-and-Y end-to-side hepaticojejunostomy with 6/0 PDS was performed for biliary reconstruction. Before thoraco-abdominal closure, Doppler-US established a good flow through the neo-IVC, left hepatic vein, left hepatic arteries, and PV. The total operation time was 8 h and