Inferior Vena Cava Resection and Hemihepatectomy for Leiomyosarcoma, Utilizing Cardiopulmonary Bypass, in Situ Hepatic Perfusion, and Distal Hypothermic Circulatory Arrest

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IN THIS CASE REPORT, the authors describe a novel hybrid perfusion approach for a large inferior vena cava (IVC) leiomyosarcoma, extending into the right atrium and right lobe of the liver, and further complicated by thrombus extending into the renal and femoral veins.

The objective of the authors' approach was to maintain cerebral perfusion via upper body cardiopulmonary bypass while providing a bloodless surgical field below the diaphragm to allow resection of the tumor from the liver and IVC and right atriotomy. The authors also outline the in vivo use of organ preservation solution during vascular exclusion to maintain viability of the hepatic remnant.

The authors present a review of the literature and an associated description of the anesthetic, surgical and perfusion considerations, and techniques used in this case.

CASE REPORT

A 55-year-old previously well woman presented to the authors' institution with a 2-day history of right upper quadrant pain, nausea, and vomiting. Clinical examination noted tenderness in the same region, and associated hepatomegaly. Laboratory testing demonstrated acute renal impairment, anemia, and deranged hepatic synthetic function (Table 1).

An upper abdominal ultrasound revealed IVC occlusion and reduced flow in the main portal and intrahepatic veins consistent with incipient Budd-Chiari syndrome. Computed tomography (CT) confirmed these findings and demonstrated extensive invasion of the right adrenal gland and right lobe of the liver by a heterogenous mass apparently arising from the perirenal IVC (Fig 1). The tumor extended to the renal veins inferiorly and 2.8 cm into the right atrium superiorly. The distal IVC, iliac, and common femoral veins were occluded with clot. Moderate ascites, collapse of the right lower lobe of the lung, and associated pleural effusion were identified. On transthoracic echocardiography (TTE), a large echogenic mass was seen in the right atrium, measuring 4.6 cm × 2.9 cm. The tricuspid valve was not obstructed. Despite abnormalities in her laboratory coagulation profile, the presence of IVC thrombus necessitated commencing therapeutic, low–molecular-weight heparin.

Aggressive palliative debulking was proposed to avoid imminent IVC occlusion and Budd-Chiari syndrome. The decision to proceed with this significant operation was supported by the patient's young age and physical fitness. The authors planned resection of the tumor from the right atrium, surgical IVC resection, right hemihepatectomy, and IVC reconstruction together with on-table assessment of the resectability of other lesions.

Two major issues were apparent: Firstly, circulatory arrest below the diaphragm would be necessary as there would be no other means of preserving lower body venous return, due to occlusion of the femoral vessels. Circulatory arrest also would have the advantage of limiting visceral organ perfusion and, hence, portal blood flow, improving surgical conditions for hepatic resection. Secondly, the length of time required for complex surgery and on-table assessment required careful consideration of organ protection. Deep hypothermia would contribute to organ protection by limiting warm ischemia, and cerebral perfusion would further reduce the risk of brain injury. Hepatic protection also would be particularly important as the planned liver resection was extensive, and residual liver dysfunction was a significant possibility. To address these issues, lower body deep hypothermic circulatory arrest (DHCA) by means of an aortic cross-clamp at the level of the diaphragm was planned with continued perfusion of the upper body by the bypass circuit. In situ hepatic perfusion via the residual left portal vein to the liver remnant using Custodiol® HTK (Dr Franz-Kohler Chemie GmbH, Bensheim, Germany) also was to be utilized.

After induction of anesthesia and intubation, vascular access was further secured with two 8-French rapid infusion catheters and bilateral brachial arterial catheters, which were continuously monitored throughout the case. A right internal jugular central venous catheter was inserted, as was a temperature-sensing, indwelling urinary catheter to measure core temperature. A nasopharyngeal temperature probe was placed separately to measure cerebral temperature. Bispectral Index™ (Covidien PLC, Dublin, Ireland) was used to monitor anesthetic depth. Transesophageal echocardiography confirmed the findings of the preoperative study (Fig 2). Point-of-care thromboelastography (TEG) was consistent with a highly thrombogenic state.

A laparotomy extended by sternotomy revealed a tense, engorged liver with substantial venous obstruction additionally, two small deposits of tumor were discovered in the left lobe. Frozen section and histopathologic analysis confirmed the diagnosis of leiomyosarcoma.

Initial hepatic mobilization was complicated by approximately 1 L of blood loss. A decision was made to institute cardiopulmonary bypass earlier than planned to allow for salvage of shed blood. After administration of 500 units/kg of heparin and 15 mg/kg of tranexamic acid, a 34F malleable single-stage basket (Medtronic, Minneapolis, MN) was placed

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Table 1. Results of Admission, Postoperative Day 1, and Discharge Laboratory Studies With Relevant Reference Ranges

		Postoperative		Reference
	Admission	Day 1	Discharge	Range
Hemoglobin (g/L)	109	95	109	115-165
INR	1.4	2.1	1.0	<1.3
aPTT (seconds)	25	42	24	22-38
Fibrinogen (g/L)	2.7	1.3	3.3	2.0-4.0
Urea (mmol/L)	10.4	4.4	2.4	3.5-7.2
Creatinine (μmol/L)	141	105	33	44-80
eGFR (mL/min)	36	51	91	>90
Bilirubin (μmol/L)	11	73	4	<18
ALT (units/L)	532	174	34	<33
AST (units/L)	526	348	44	<32
γ-GT (units/L)	342	27	221	<40
ALP (units/L)	376	42	159	35-105
Albumin (g/L)	38	35	30	35-52

in the right atrium, and a 24F single-stage, right-angle drainage cannula from the same manufacturer was placed in the superior vena cava (SVC). Special care was taken to avoid disturbing the tumor. An aortic root vent was inserted, as well as a left ventricular vent via the left superior pulmonary vein. A 22F EPOA® return cannula (Medtronic, Minneapolis, MN) was placed in the distal ascending aorta, and cardiopulmonary bypass was commenced. Tranexamic acid infusion was commenced at 5 mg/kg/h. Anticoagulation was monitored regularly using activated clotting time (ACT). When adequate mobilization of the liver had been achieved, cooling was commenced. At 26°C, the ascending aorta was cross-clamped, and a single dose of 1,500 mL of Custodiol® histidine-tryptophan-ketoglutarate (HTK) cardioplegia was administered, with immediate diastolic arrest. The patient then was cooled further to a nasopharyngeal temperature of 18°C. One gram of methylprednisolone was administered with the aim of enhancing spinal cord protection. The inominate, carotid, and subclavian arteries were not manipulated. A second cross-clamp was applied to the descending thoracic aorta immediately above the diaphragm. The abdominal organs were packed in ice. Upper body perfusion via the ascending aortic cannula (Fig 3) was maintained at 11 mL/kg/min, yielding a mean arterial pressure of 50 mmHg to 60 mmHg, as measured by the brachial arterial catheters. Hemofiltration was commenced after distal cross-clamping.

The right hemihepatectomy then was completed. To allow for greater surgical access, the left hepatic vein was detached from the IVC. Custodiol® HTK was administered into the left portal vein under continuous hydrostatic pressure (less than 120 mmHg) at approximately 20 mL/min and 4°C in keeping with the authors' institutional protocol for organ retrieval. This preserved the remaining left liver that now was excluded from the portal and systemic circulations. Given the extent of the planned procedure and the operative risk, a decision was made to forego removal of the left liver lobe deposits. The IVC was transected above the renal veins and tumor was extracted from the perirenal IVC without damaging the vein. The clot was retrieved from below the tumor using a Fogarty catheter. A right atriotomy was performed, and the tumor dissected from the atrial wall and displaced into the IVC. The IVC was transected again just below the orifice of the left hepatic vein and the tumor removed from the suprarenal IVC (Fig 4). The IVC then was reconnected to the left hepatic vein and the right atrium closed. Retroperitoneal dissection was completed behind the IVC bed, and the right adrenal gland resected. A 24-mm Dacron graft then was anastomosed to the free ends of the IVC.

Separation from bypass was achieved without difficulty following slow, stepwise rewarming from a nasopharyngeal temperature of 18°C to 36.5°C over 96 minutes, maintaining a maximum gradient of 7°C between the nasopharyngeal and cardiopulmonary bypass (CPB) perfusate temperatures. Total cardiopulmonary bypass time was 4 hours and 30 minutes with a proximal cross-clamp time of 2 hours and 56 minutes. The distal aorta was clamped for 120 minutes. Post-bypass echocardiography showed complete removal of the tumor from the right atrium. After reversal of heparin, the aorta and SVC were

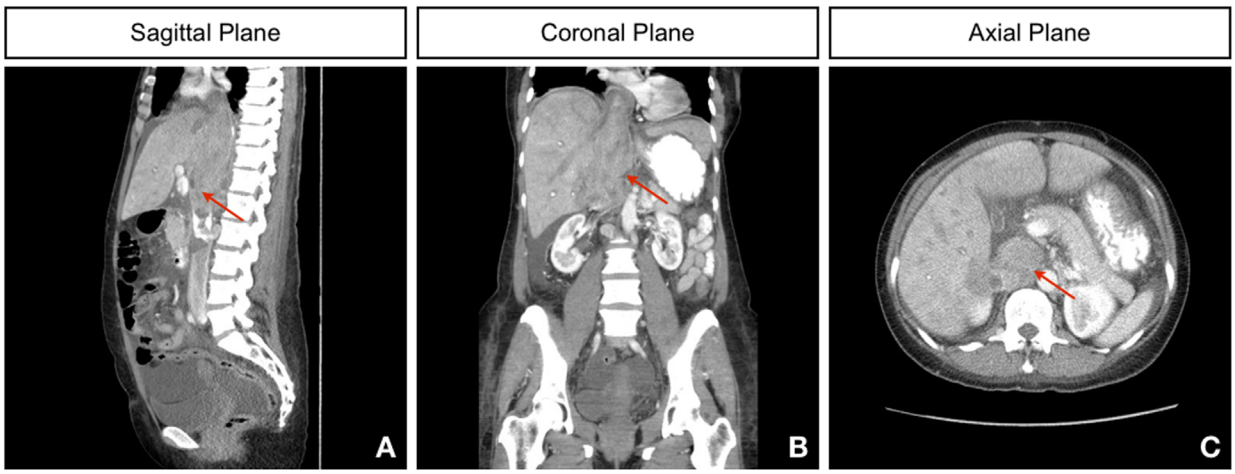

Fig 1. Abdominal computed tomography scan of the patient at presentation, presented in the sagittal (A), coronal (B), and axial (C) planes. The red arrows denote the tumor, which is seen as a heterogenous mass arising from the inferior vena cava and extending into the right lobe of the liver, the right adrenal gland, and the right atrium.