Management of synchronous vascular and ductal anomalies in living donor liver transplantation for hepatic epithelioid

LETTER TO THE EDITORS

Management of Synchronous Vascular and Ductal Anomalies in Living Donor Liver Transplantation for Hepatic Epithelioid Hemangioendothelioma

Received July 25, 2013; accepted October 22, 2013.

TO THE EDITORS:

The presence of intrahepatic vascular and ductal variants in the liver of a suitable donor should not be considered a contraindication to living liver donation. When a suitable living donor is available, a good understanding of such anomalies and meticulous surgical technique make living donor liver transplantation one of the best therapeutic options, especially when recipients are disadvantaged by their weight range in terms of long waiting times for cadaveric donation.

We present a case of a complex donor anatomy during a right lobe living donor transplant performed to treat a patient with hepatic epithelioid hemangioendothelioma. This is a rare, low-grade vascular malignancy whose therapeutic algorithm is far from standardized.[1,2] As demonstrated by different authors,[3-6] liver transplantation is a valuable treatment for hepatic epithelioid hemangioendothelioma, even in cases of extrahepatic metastasis. In our case, the 26-year-old donor and the 26-year-old recipient were twins (brother and sister, respectively). The recipient presented with obstructive jaundice and abdominal pain. The computed tomography scan showed multiple, large, central, and peripheral liver lesions suspected to be hemangioendotheliomas. She was stented via endoscopic retrograde cholangiopancreatography, and the histological examination confirmed the radiological diagnosis. An attempt at removing the stent 6 months after its positioning failed because of multiple intrahepatic strictures. She deteriorated with progressive jaundice and weight loss (44 kg) and had a body mass index of 16 kg/m^2 before transplantation. Her Model for End-Stage Liver Disease score, though not representative of her deteriorating clinical condition, had progressed from 7 at listing to 14 (UK End-Stage Liver Disease score = 50) just before transplantation. Her twin brother, a fit and healthy male weighing 68 kg (body mass index = 23 kg/m^2), came forward as a possible donor. He was blood group–identical (O Rh+) and had excellent liver function. However, his liver had several anatomical anomalies. The portal vein had a type 4 variation: a segment VIII portal vein branch (6.2 mm in diameter) originating from the distal main left portal vein (Fig. 1). A large vein (7 mm in diameter), draining segment VII, joined the inferior vena cava just inferiorly to the main right hepatic vein; another vein (4 mm in diameter) was identified as an accessory low inferior right hepatic vein. An intraoperative cholangiogram showed a right posterior bile duct draining into the main left bile duct (Fig. 2). The artery to segment VIII arose from the right system, so there was no concern about arterial ischemia for this segment after graft removal. On the back table, the graft was prepared as follows:

- The segment VIII portal branch was detached from the left portal branch origin and reconstructed with the right portal vein into a single vein orifice, which was elongated with a fresh cadaveric, ABO-identical iliac vein interpositional graft.
- Venoplasty between the right hepatic vein and the vein draining segment VII was first performed, and this was followed by quilt plasty using an ABO-identical, cadaveric iliac vein sutured around the common venous orifice.
- The stumps of the branches of the middle hepatic vein draining segments V and VIII (V5 and V8), were reconstructed with a fresh cadaveric, ABO-identical iliac artery.

Implantation was performed through the creation of an extended midline orifice in the recipient's inferior vena cava, which accommodated the venoplasty patch of the graft. The reconstructed right portal vein and the right hepatic artery were regularly anastomosed with 6/0 and 7/0 Prolene sutures, respectively. The 2 ducts were drained with a Roux-en-Y loop because the extent of the tumor in the recipient precluded the

[6]The study protocol received a priori approval by the hospital review committee.

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DOI 10.1002/lt.23781
View this article online at wileyonlinelibrary.com.
LIVER TRANSPLANTATION. DOI 10.1002/lt. *Published on behalf of the American Association for the Study of Liver Diseases*

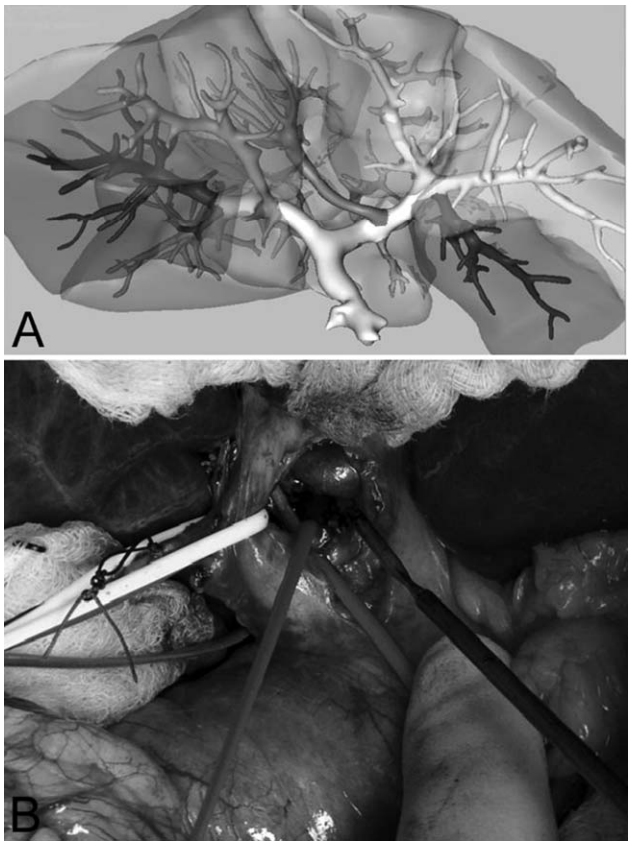

Figure. 1. MeVis reconstruction and intraoperative picture of the donor's liver. (A) MeVis reconstruction showing the donor's liver with its anomalous portal vein branch for segment VIII originating from the distal main left portal vein. (B) Intraoperative picture of the donor's liver with the diathermy indicating the anomalous portal vein branch for segment VIII in the porta hepatis.

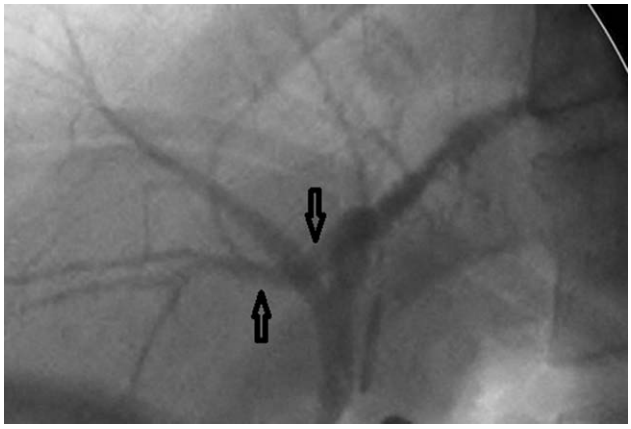

Figure. 2. Intraoperative donor cholangiogram, with the two arrows showing the aberrant bile duct from segment VIII draining into the main left bile duct.

use of the recipient's ducts. Postoperatively, the donor presented with a right pleural effusion of no clinical relevance. He was discharged 6 days after surgery and was free from vascular or biliary complications at 4 months' follow-up. The recipient immediately had excellent liver function, and regular Doppler ultrasound examinations showed patency of the reconstructed vessels, which was confirmed with a computed tomography scan (Figs. 3 and 4) at discharge 11 days after transplantation.

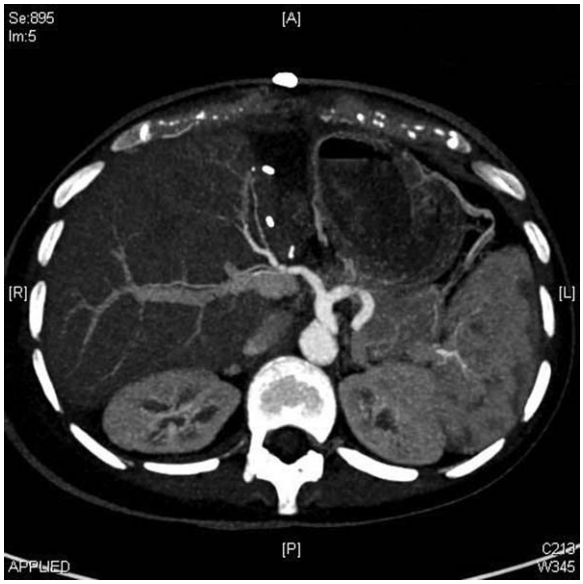

Figure. 3. Postoperative computed tomography scan of the recipient during the portal venous phase. The scan shows patency of the reconstruction of the anomalous segment VIII portal branch and the right portal vein into a single vein orifice, which was then elongated with a fresh cadaveric, ABO-identical iliac vein interpositional graft.

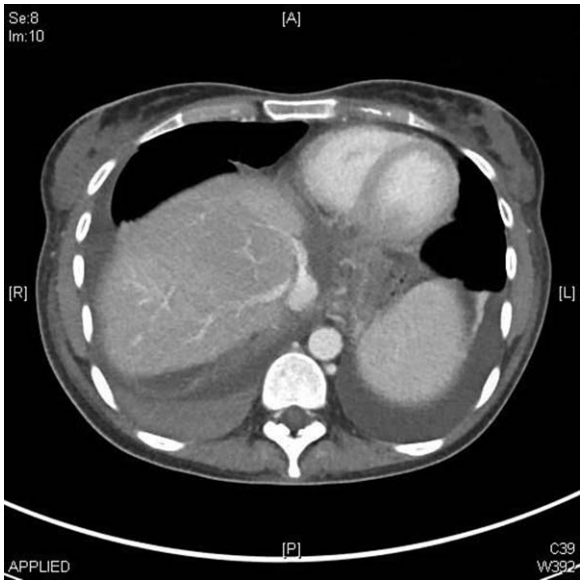

Figure. 4. Postoperative computed tomography scan of the recipient during the portal venous phase. This second image from the same postoperative computed tomography scan shows patency of the anastomosis between the inferior vena cava and the reconstructed venous drainage of the graft. Venoplasty between the right hepatic vein and the vein draining segment VII was performed, and this was followed by quilt plasty of the common orifice performed with an ABO-identical, cadaveric iliac vein.

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REFERENCES

1. Lerut JP, Weber M, Orlando G, Dutkowski P. Vascular and rare liver tumors: a good indication for liver transplantation? J Hepatol 2007;47:466-475.
2. Mehrabi A, Kashfi A, Fonouni H, Schemmer P, Schmied BM, Hallscheidt P, et al. Primary malignant hepatic epithelioid hemangioendothelioma: a comprehensive review of the literature with emphasis on the surgical therapy. Cancer 2006;107:2108-2121.
3. Mosoia L, Mabrut JY, Adham M, Boillot O, Ducerf C, Partensky C, Baulieux J. Hepatic epithelioid hemangioendothelioma: long-term results of surgical management. J Surg Oncol 2008;98:432-437.
4. Grotz TE, Nagorney D, Donohue J, Que F, Kendrick M, Farnell M, et al. Hepatic epithelioid haemangioendothelioma: is transplantation the only treatment option? HPB (Oxford) 2010;12:546-553.
5. Lerut JP, Orlando G, Adam R, Schiavo M, Klempnauer J, Mirza D, et al.; for European Liver Transplant Registry. The place of liver transplantation in the treatment of hepatic epitheloid hemangioendothelioma: report of the European Liver Transplant Registry. Ann Surg 2007;246: 949-957.
6. Lerut JP, Orlando G, Sempoux C, Ciccarelli O, Van Beers BE, Danse E, et al. Hepatic haemangioendothelioma in adults: excellent outcome following liver transplantation. Transpl Int 2004;17:202-207.