Transplantation

Issue: Volume 77(2), 27 January 2004, pp 326-327

Publication Type: [LETTERS TO THE EDITOR]
ISSN: 0041-1337
Accession: 00007890-200401270-00035

[LETTERS TO THE EDITOR]

Portal biliopathy treated by liver transplantation

Filipponi, Franco; Urbani, Lucio; Catalano, Gabriele; Iaria, Giuseppe; Biancofiore, Gianni; Cioni, Roberto; Campatelli, Alessandro; Mosca, Franco; Campani, Daniela; Romagnoli, Paolo

Author Information
Liver Transplant Unit, University of Pisa, Pisa, Italy

Postsurgical and Transplant Intensive Care Unit, University of Pisa, Pisa, Italy

Diagnostic and Interventional Department, University of Pisa, Pisa, Italy

General and Transplantation Surgery, University of Pisa, Pisa, Italy

Division of Pathology, University of Pisa, Pisa, Italy

Department of Anatomy, Histology and Forensic Medicine, University of Florence

Address correspondence to: Franco Filipponi, M.D., Liver Transplant Unit, Ospedale Cisanello, Via Paradisa 2, 56124 Pisa, Italy. E-mail: f.filipponi@med.unipi.it.

Received 6 February 2003.

Revised 31 March 2003.

Accepted 5 June 2003.

Portal biliopathy is an uncommon cause of obstructive jaundice (1). The proposed therapies include endoscopic dilatation and stenting of the common bile duct or a portocaval shunt with splenectomy and sometimes hepatico-jejunostomy (2).

We recently treated a white man with obstructive jaundice who had growth-hindering diarrhea in childhood. He experienced abdominal pain at the age of 33 years, and ultrasonography and computed tomography examinations revealed mesentericoportal thrombosis and splenomegaly. One year later, endoscopy showed esophageal varices; a liver biopsy showed normal histology. Obstructive jaundice first occurred when he was 40 years old; the patient was admitted to another hospital, and endoscopic retrograde cholangio-pancreatography showed concentric stenosis of the common bile

duct, with no evidence of stones (Fig. 1). The jaundice recovered spontaneously, but biochemical signs of cholestasis persisted. At 46 years of age, the patient's blood bilirubin levels increased to 29 mg/dL. Nuclear magnetic resonance with contrast failed to show the carrefour and extrahepatic bile ducts; stenting was excluded because the stenosis affected more than one duct. Grade IIIb portal biliopathy was diagnosed according to Chandra et al. (1). Percutaneous bile drainage did not relieve the jaundice and was complicated by a bilioportal fistula with hemobilia. Computed angiotomography revealed a splenoportal cavernoma with a spleno-renal shunt and abundant intrahepatic angioma-like tissue (Fig. 2). Liver protein synthesis markedly decreased (serum albumin 2.7 g/L and prothrombin activity 35% with an international normalized ratio of 2.34), partially because of the impaired absorption of lipophilic vitamins, ascites appeared, and muscle mass decreased. For these last reasons, and the persistence of severe jaundice and hemobilia, the patient was considered at risk of imminent death. We judged organ transplantation as the best therapy because of his poor general condition, the abundant angioma-like intrahepatic tissue, and the presence of both extrahepatic and bilateral intrahepatic bile duct stenosis.

During surgery, a large splenoportal cavernoma was found encircling and compressing the common bile duct. After ligation of the splenic artery to avoid posttransplantation blood steal, portal pressure decreased abruptly, indicating the existence of an intrasplenic arteriovenous shunt and the need for splenectomy. Because of the high risk of thrombosis, a cavoportal hemitransposition was performed (3,4). Briefly, the cavernoma was connected to the recipient vena cava through an iliac vein graft; the recipient suprarenal vena cava was anastomosed to the donor infrahepatic vena cava (end-to-end) and portal vein (end-to-side). Graft portal perfusion was ensured by calibrating the donor infrahepatic vena cava.

The removed liver weighed 2,340 g and was bile-green; histology showed signs of recurrent thrombosis of the portal branches, cholestasis, and fibrosis, as well as abundant loose connective tissue extremely rich in blood vessels surrounding the branches of the portal and hepatic arteries and bile ducts. To the best of our knowledge, this is the first report of such a finding in a patient with portal biliopathy. The spleen weighed 1,770 g; the red pulp was replaced by newly formed vascular channels lined by factor VIII-related antigen-positive endothelial cells.

Despite appropriate prophylaxis with low molecular weight heparin immediately after surgery (4), the grafted vena cava and vena porta, the iliac vein graft, and the right hepatic artery developed thromboses with severe ascites. We therefore decided to perform a retransplantation on the eighth postoperative day using an end-to-end cavoportal anastomosis; the drainage of the obstructed cavernoma was not restored. On the following day, severe congestive gastropathy caused hemorrhaging, which was addressed by means of a thromboendovenectomy of the cavernoma and its draining into the donor infrahepatic vena cava through a new (end-to-end) iliac vein graft, thus achieving a complete cavoportal transposition. The patient was discharged after 2 months with anticoagulant and antiaggregant therapy (4); he is still well 12 months later.

To the best of our knowledge, this is the first report of portal biliopathy treated by liver transplantation and shows that this therapy may be indicated in the treatment of otherwise intractable disease.

Franco Filipponi

Lucio Urbani

Gabriele Catalano

Giuseppe Iaria

Gianni Biancofiore

Roberto Cioni

Alessandro Campatelli

Franco Mosca

Daniela Campani

Paolo Romagnoli

REFERENCES

1. Chandra R, Kapoor D, Tharakan A, et al. Portal biliopathy. J Gastroenterol Hepatol 2001; 16: 1086. SFX Buy Now Bibliographic Links

2. Chaudhary A, Dhar P, Sarin SK, et al. Bile duct obstruction due to portal biliopathy in extrahepatic portal hypertension: surgical management. Br J Surg 1998; 85: 326. SFX Bibliographic Links

3. Tzakis AG, Kirkegaard P, Pinna AD, et al. Liver transplantation with cavoportal hemitransposition in the presence of diffuse portal vein thrombosis. Transplantation 1998; 65: 619. Ovid Full Text SFX Bibliographic Links

4. Urbani L, Cioni R, Catalano G, et al. Cavoportal hemitransposition: patient selection criteria and outcome. Transplant Proc 2002; 34: 3331. SFX Bibliographic Links

---