Liver Transplantation for Portal Cavernoma Cholangiopathy

Subash Gupta*, Sunil Taneja†

*Department of Surgical Gastroenterology & Liver Transplantation and †Department of Gastroenterology and Hepatology, Centre for Liver & Biliary Surgery, Indraprastha Apollo Hospital, New Delhi 110076, India

Portal cavernoma cholangiopathy (PCC) is a difficult clinical problem, where the portal cavernoma is both the cause of biliary obstruction and the obstacle to its safe surgical treatment. The available endoscopic and surgical treatment is successful in majority and further intervention is seldom required since the native liver is normal. PCC is not an accepted indication for liver transplantation as only a small proportion of patients will fail both endoscopic and surgical treatment and progressive liver failure is rarely seen. Secondary biliary cirrhosis as a result of long standing biliary obstruction is an accepted indication however establishing a portal inflow in these patients is often difficult and challenging. The deceased donor liver transplantation would always be preferable over living donor liver transplantation as PCC is usually a non-emergency transplant and the graft can have portal blood inflow through a conduit to even a small segment of patent portal venous system or even to a cavernoma vessel. (J CLIN EXP HEPATOL 2014;4:S85–S87)

Portal cavernoma cholangiopathy (PCC) is a difficult clinical problem, where the portal cavernoma is both the cause of biliary obstruction and the obstacle to its safe surgical treatment. The incidence of PCC ranges between 5% and 18%.^{1–5} The available endoscopic and surgical treatment is successful in majority.^{6–8} However a subgroup of patients including those with recurrent gastrointestinal bleeding and recurrent cholangitis may be difficult to manage if both endoscopic and surgical intervention fails or is not feasible. In such cases the available treatment options are limited. Liver transplantation is a difficult option for PCC but has the potential of treating symptomatic PCC and varices in one procedure. If there is blood flow to the intestine, it stands to reason that there will be portal outflow from the intestine through collaterals. With modern imaging it may be possible to locate one of these collaterals and plan a liver transplant using this as an inflow vessel. However if no tributary or collateral of the portal venous system is identified, then liver transplant will not be possible as caval inflow will not solve the problem of portal hypertension. For example the transplanted liver

can be perfused through inflow from the bile duct varices that are often present.

LIVER TRANSPLANTATION FOR PCC

The literature is scarce on transplantation as a therapeutic modality for PCC. The standalone indication of liver transplantation for PCC is secondary biliary cirrhosis occurring as a result of long standing biliary obstruction; however this is rarely seen in the present aggressive endoscopic stenting era.

Till now only few case reports are available in literature where liver transplantation has been done for PCC after failure of endoscopic and surgical measures. The first reported case of liver transplantation for PCC was in 2004 in a 40-year-male patient with progressively worsening liver functions with grade IIb PCC.^{9,10} Percutaneous bile drainage did not relieve the jaundice and was complicated by a biliportal fistula with hemobilia. Organ transplantation was judged 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.

Another reported case of liver transplantation for PCC was of a young patient with a 17-years long history of non-cirrhotic portal hypertension and cavernous transformation of the portal vein (predominantly intrahepatic portal cavernoma).¹¹ Patient developed PCC not alleviated by repeated endoscopic stent insertion, clinically mimicking primary sclerosing cholangitis and cholangiocarcinoma. The patient underwent deceased donor liver transplantation (DDLT) with a suspicion of cholangiocarcinoma on imaging studies to take care of both the primary disease and the suspected malignancy.

Keywords: portal biliopathy, pseudosclerosing cholangitis, extrahepatic portal venous obstruction, portal hypertension

Received: 8.2.2013; **Accepted:** 2.1.2014; **Available online:** 17.1.2014

Address for correspondence: Subash Gupta, Room No 1246, Centre for Liver & Biliary Surgery, Indraprastha Apollo Hospital, New Delhi 110076, India. Fax: +91 11 41611010

E-mails: drsubashgupta@hotmail.com; guptasubash@hotmail.com

Abbreviations: PCC: portal cavernoma cholangiopathy; DDLT: deceased donor liver transplantation; LDLT: live donor liver transplantation

<http://dx.doi.org/10.1016/j.jceh.2014.01.001>

Table 1 Profile of Patients of Portal Cavernoma Biliopathy Undergoing Successful Liver Transplantation.

Authors	Age (years)	Bilirubin (mg/dL)	Type of surgery	Portal vein reconstruction or portal inflow	Indication for liver transplantation	Outcome	Follow-up
Francoet al ⁹	40	29	DDLT	Iliac vein graft to achieve a complete cavoportal transposition	Progressive liver failure and intra- and extra-hepatic bile duct stenosis	Successful	12 months
Hajdu et al ¹¹	43	11.8	DDLT	–	Recurrent cholangitis and suspicion of cholangiocarcinoma	Successful; No tumor identified	12 months
Zhang et al ¹³	9	1.01	LDLT	Venous anastomosis of donor portal vein to a large recipient collateral vein	PCC, hypersplenism	Expired	7 days
	7	2.73	LDLT	Short bridging vein graft to portal vein confluence of splenic and superior mesenteric vein	PCC, Hypersplenism	Successful	26 months
	10	12.71	LDLT	Jump bypass graft was anastomosed end-to-side to superior mesenteric vein	PCC, hypersplenism, hepatic encephalopathy	Successful	26 months
Gupta et al ¹²	26	1.50	LDLT	Infracolic collateral	Life threatening sepsis, recurrent cholangitis, recurrent gastrointestinal bleed	Successful	24 months

DDLT, Deceased Donor Liver Transplantation; LDLT, Living Donor Liver Transplantation; PCC, portal cavernoma cholangiopathy.

We reported the first case of live donor liver transplantation (LDLT) for PCC in a 26-year-old Indian man from United States who presented with growth retardation during childhood, recurrent episodes of cholangitis for 11 years, upper gastrointestinal hemorrhage, and generalized weakness for 4 years. At the age of 15-years he developed features of hypersplenism for which he underwent a splenic artery embolization. One year later, he developed obstructive jaundice. Subsequently he had recurrent episodes of obstructive jaundice with cholangitis and upper gastrointestinal haemorrhage which required multiple endoscopic retrograde cholangiopancreatography and stenting and endoscopic variceal ligation. He also had 2 episodes of life-threatening sepsis owing to cholangitis requiring immediate stent exchange and admission to intensive care units. Considering his young age, intractable symptoms, failure of endoscopic therapy, nonfeasibility of other treatment options, poor quality-of-life, and nonavailability of deceased-donor organ, the option of an LDLT was given to him. He underwent a successful right lobe graft without the middle hepatic vein, donor being his 48-year-old mother.¹²

LDLT as a first line therapy was also reported from China in three pediatric patients with extensive intrahepatic cavernous transformations, varices, hypersplenism and liver decompensation in one patient because of concerns regarding the expected low effectiveness of a surgical portosystemic shunt in pediatric population.¹³ The donor was the child's father in two cases and the patient's mother in the third case. All donor grafts were taken from the left

lobe (segments 2, 3, 4, and middle hepatic vein). Graft hepatic vein was used to extend the portal vein for anastomosis in all three cases. In the first patient usual venous anastomosis of the donor portal vein to a large recipient collateral vein of portal vein was made whereas in the second case anastomosis was done to the confluence of the splenic and superior mesenteric veins. Among the three PCC patients, the tortuous, closely packed veins and venules formed a sponge like lesion that extended into the intrahepatic portal lobar branches. Hilar fibrosis and inflammation made hilar dissections around the hepatoduodenal ligament very difficult and laborious. Some technical difficulties were encountered, including the dissection of a portal vein that featured no obvious lumen, hemorrhage and the separation of the bile duct from the portal vein. However the transplantation procedure was technically feasible, and there was no procedure-related mortality in any patient. The profile of patients of PCC undergoing successful liver transplantation is given in Table 1.

DECEASED VERSUS LIVING DONOR LIVER TRANSPLANTATION

DDLT is always the preferable surgery as PCC is usually a non-emergency transplant, good graft portal vein length and veins for conduits are available and recipient hepatectomy is easier. However organs from living donors offer many potential advantages over organs from brain-dead donors. The most important advantages of live donation are that it optimizes the timing of transplantation and frees

patients from the waiting list. Preservation time is minimal in LDLT, so there is significantly less ischemic damage to the liver.¹⁴ LDLT is difficult, but may be an option if emergency transplant is needed and patency of some part of portal venous system is there. However the disadvantages to live donor transplantation must be considered carefully. The donor, a perfectly healthy volunteer, faces unequivocal risks of morbidity and even mortality. In LDLT there are no cadaveric venous grafts and it is a difficult surgery as extensive retroperitoneal collaterals are present in these patients with long standing portal hypertension.

CONCLUSION

Liver transplantation can be considered rarely in patients with PCC. However it still remains a difficult option and a last resort when all modalities fail. Secondary biliary cirrhosis is the only accepted indication, however it may not be feasible in all patients due to difficulty in establishing a portal inflow.

CONFLICTS OF INTEREST

All authors have none to declare.

REFERENCES

1. Condat B, Vilgrain V, Asselah T, et al. Portal cavernoma-associated cholangiopathy: a clinical and MR cholangiography coupled with MR portography imaging study. *Hepatology*. 2003;37:1302–1308.
2. Dilawari JB, Chawla YK. Pseudosclerosing cholangitis in extrahepatic portal venous obstruction. *Gut*. 1992;33:272–276.
3. Khuroo MS, Yattoo GN, Zargar SA, et al. Biliary abnormalities associated with extrahepatic portal venous obstruction. *Hepatology*. 1993;17:807–813.
4. Malkan GH, Bhatia SJ, Bashir K, et al. Cholangiopathy associated with portal hypertension: diagnostic evaluation and clinical implications. *Gastrointest Endosc*. 1999;49:344–348.
5. Nagi B, Kochhar R, Bhasin D, Singh K. Cholangiopathy in extrahepatic portal venous obstruction: radiological appearances. *Acta Radiol*. 2000;41:612–615.
6. Dhiman RK, Behera A, Chawla YK, Dilawari JB, Suri S. Portal hypertensive biliopathy. *Gut*. 2007;56:1001–1008.
7. Saraswat VA, Rai P, Kumar T, Mohindra S, Dhiman RK. Endoscopic management of portal cavernoma cholangiopathy: Practice, principles and philosophy. *J Clin Exp Hepatol*. 2013;3(suppl 2).
8. Varma V, Behera A, Kaman L, Chattopadhyay S, Nundy S. Surgical management of portal cavernous cholangiopathy. *J Clin Exp Hepatol*. 2013;3(suppl 2).
9. Filipponi F, Urbani L, Catalano G, et al. Portal biliopathy treated by liver transplantation. *Transplantation*. 2004;77:326–327.
10. Chandra R, Kapoor D, Tharakan A, Chaudhary A, Sarin SK. Portal biliopathy. *J Gastroenterol Hepatol*. 2001;16:1086–1092.
11. Hajdu CH, Murakami T, Diflo T, et al. Intrahepatic portal cavernoma as an indication for liver transplantation. *Liver Transpl*. 2007;13:1312–1316.
12. Gupta S, Singhal A, Goyal N, Vij V, Wadhawan M. Portal biliopathy treated with living-donor liver transplant: index case. *Exp Clin Transpl*. 2011;9:145–149.
13. Zhang M, Guo C, Pu C, et al. Adult to pediatric living donor liver transplantation for portal cavernoma. *Hepatol Res*. 2009;39:888–897.
14. Florman S, Miller CM. Live donor liver transplantation. *Liver Transpl*. 2006;12:499–510.