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Obstruction to Hepatic Venous Drainage after Liver Transplantation: Treatment with Balloon Angioplasty[1]

Stenosis of the suprahepatic inferior vena caval anastomosis is a rare but serious vascular complication after liver transplantation. It may cause significant obstruction to venous drainage from the allograft liver and result in the Budd-Chiari syndrome with massive ascites and pleural effusion causing respiratory compromise. The authors report two such cases in which percutaneous transluminal angioplasty (PTA) of the stenotic anastomosis was performed. This nonsurgical approach resulted in resolution of ascites, pleural effusion, and respiratory distress in both patients. They conclude that PTA is a therapeutic alternative with minimal risk compared with surgical repair or retransplantation and should be considered the initial treatment of choice in selected patients.

ORTHOTOPIC liver transplantation is an accepted therapy for liver failure caused by irreversible hepatic disease (1); however, technical complications remain a significant cause of morbidity and mortality (2-4). Complications related to arterial and biliary tract reconstruction occur most frequently, with incidences of 7.4% and 13.2%, respectively (5,6). Venous complications, especially those involving the inferior vena cava (IVC), are unusual (4,7). Herein, we describe two pediatric patients who developed significant stenosis of the suprahepatic IVC anastomosis approximately 4 and 5 months after orthotopic liver transplantation. The resulting obstruction to hepatic venous drainage resulted in the Budd-Chiari syndrome. Both cases were successfully treated nonoperatively by means of percutaneous transluminal angioplasty (PTA) of the stenotic anastomosis.

CASE REPORTS

Case 1.—A 1-year-old girl with biliary atresia underwent three successive orthotopic liver transplantations at the Children's Hospital of Pittsburgh. Allograft failure in the first two transplants occurred after thrombosis of the hepatic artery. After the third transplantation she did well, was discharged after 5 weeks, and returned to her home in France with normal liver function.

Four months after transplantation, she presented with acute respiratory distress with fever and a large right pleural effusion. Upon admission to the University Hospital St-Luc in Brussels, she was found to have hepatomegaly and significant ascites. Her total bilirubin level was 2.0 mg/dL (34.2 μmol/L).

Initial evaluation consisted of real-time sonography, which demonstrated an area of increased echogenicity surrounding the region of the suprahepatic IVC anastomosis; this area was thought to most likely represent scar tissue. In addition, there was no evidence of a patent lumen between the confluence of the right hepatic veins and the right atrium. The IVC could not be identified. Because of these findings, inferior vena cavography was performed via a common femoral vein approach; this study demonstrated occlusion of the hepatic segment of the IVC. For evaluation of the suprahepatic IVC anastomosis, percutaneous catheterization of the IVC via the right internal jugular vein was performed. This study demonstrated severe stenosis of the suprahepatic IVC anastomosis at the level of hepatic venous drainage (Fig 1a).

Treatment consisted of balloon dilatation of the stenotic anastomosis via the right internal jugular vein. Because of the severity of the stricture, a 5-mm balloon catheter could not initially be advanced across the anastomosis. Dilation was first performed with 5-9-F vessel dilators, which permitted introduction of the balloon catheter. For technical reasons, a pre-PTA pressure gradient was not measured. At initial inflation, an hourglass deformity of the balloon was noted. Continued inflation was performed until the indentation of the lumen of the balloon was eliminated. The balloon was inflated five times. Because of arrhythmias, the balloon could not be inflated for longer than 10 seconds each time. After dilation there was slight improvement in the lumen diameter. Use of a larger balloon catheter was attempted; however, because of resistance at the stricture, the catheter could not be advanced through the anastomosis.

After 6 days repeat angiography was performed to reevaluate the suprahepatic IVC anastomosis and to reattempt PTA with a larger balloon. Before PTA, a pressure gradient of 10 mm Hg across the anastomosis was recorded. A 9-mm-diameter, 3-cm-long balloon was positioned across the anastomosis. As done previously, the balloon was inflated to eliminate the waist on the balloon lumen. The balloon was inflated ten times, again for 10 seconds each time. After PTA, there was no pressure gradient across the anastomosis (Fig 1b). The serum bilirubin level returned to normal, the large pleural effusion resolved, and the ascites decreased.

The stricture recurred two times during the following 5 months. The signs of recurrence were pleural effusion and ascites. At angiography, a stricture at the anastomosis was noted, but it was not as severe as the first stricture. In addition, the clinical manifestations were much less severe. PTA was repeated, with balloon diameters of 9 mm the first time and 15 mm the second. Each time, a significant pre-PTA pressure gradient was recorded, with marked reduction after PTA. The patient is without recurrence of

symptoms 31 months after the last PTA.

Case 2.—A 9-month-old girl with biliary atresia underwent orthotopic liver transplantation at the Children's Hospital of Pittsburgh. Her immediate postoperative course was complicated by thrombosis of the hepatic artery requiring repeat transplantation. Other than several episodes of rejection that responded to treatment, she did well and was discharged 2 months after her second transplantation with essentially normal liver function.

Two months later she developed hepatic dysfunction manifested by elevated liver enzyme levels. This dysfunction was thought to be caused by rejection, and she was treated with steroids. Although liver function improved mildly, her condition deteriorated rapidly, with onset of massive ascites and shortness of breath. The patient was readmitted to Children's Hospital.

Duplex sonography was performed and demonstrated narrowing of the lumen of the IVC at the suprahepatic anastomosis. Pulsed Doppler sonography showed flow in the IVC with markedly increased velocity within the suprahepatic narrowed segment. Because of these findings and the massive ascites, we suspected obstruction to hepatic venous drainage due to stenosis of the suprahepatic IVC anastomosis. The following day, she underwent inferior vena cavography, which documented stenosis at the suprahepatic IVC anastomosis (Fig 2a) corresponding to that identified at sonography.

Several days later the patient underwent PTA via a common femoral vein approach. The IVC stenosis was crossed with a 5-F catheter, and pullback pressure recordings showed a pressure gradient of 7 mm Hg. An 8-mm-diameter, 3-cm-long balloon was placed across the anastomosis and inflated six times for 15–30-second intervals. Upon initial inflation, an hourglass deformity was noted, which eventually disappeared on subsequent inflations. After PTA, there was no pressure gradient across the anastomosis (Fig 2b).

After this procedure, the patient showed progressive clinical improvement, with a decrease in ascites, weight, and abdominal girth. Her affect and appetite also improved. Follow-up angiography was performed after 3 weeks. Because of mild restenosis, PTA was repeated, again with an 8-mm balloon.

Despite successful PTA, her liver enzyme levels continued to deteriorate over the next 3 months. Liver biopsy revealed chronic rejection. Because of her clinical condition a third transplant was necessary. At this operation, a significant amount of fibrous tissue was present at the suprahepatic IVC anastomosis; however, the lumen measured 7–8 mm in diameter. Histologic examination of the anastomosis showed fibrosis of the wall as well as intimal hyperplasia. There was also focal dystrophic calcification and foreign body giant cell reaction to the sutures in the adjacent soft tissues. The patient died because of sepsis and multiple organ failure several weeks after repeat transplantation.

Figure 1. Case 1. (a) Severe stenosis of the suprahepatic IVC anastomosis (arrow) is seen, causing high-grade obstruction of the hepatic veins (arrowheads). The hepatic segment of the IVC is occluded (also documented with inferior vena cavography). (b) After PTA with a 9-mm balloon, there is marked improvement in the lumen of the hepatic vein and the anastomosis (arrow).

DISCUSSION

Obstruction to hepatic venous drainage causing ascites, edema, and hepatomegaly is referred to as the Budd-Chiari syndrome. After orthotopic liver transplantation, ascites may be manifested as a right pleural effusion, as seen in case 1. Although most commonly caused by hepatic venous thrombosis, the Budd-Chiari syndrome may be due to obstruction without thrombosis. In the two cases described here, the Budd-Chiari syndrome was caused by obstruction to hepatic venous drainage due to severe stenosis of the suprahepatic IVC anastomosis after liver transplantation.

Thrombosis of the IVC and stenosis of the IVC anastomoses are rare vascular complications of orthotopic liver transplantation (4,7). In a combined series from two centers of 359 orthotopic liver transplant recipients, thrombosis of the IVC occurred in five patients and anastomotic stenosis in only two (7,8). However, such venous complications are a significant cause of morbidity and may threaten allograft and patient survival.

The role of sonography in the evaluation of suspected IVC and other vascular complications after orthotopic liver transplantation has recently been reported (9–12). Pulsed Doppler sonography is especially important since blood flow can be demonstrated. Findings indicative of anastomotic stenosis are normal flow proximal to the stenosis, high-velocity flow at the stenosis, and turbulence above the stenosis (9,10). Sonography played an important diagnostic role in the initial evaluation of the two patients described herein, leading to angiographic documentation of significant anastomotic stenosis of the IVC in each case.

Anastomotic stenosis in orthotopic liver transplant recipients may be due to fibrosis, reactive edema, organized thrombus, or neointimal fibrous hyperplasia (8,13). A major contributing factor in the development of stenosis in our two patients may have been excessive fibrous tissue at the suprahepatic anastomosis due to repeat transplantation. A significant amount of fibrous tissue was found at repeat transplantation in case 2 and was noted at sonography in case 1. This fibrous tissue may also account for the need for repeated dilation with larger balloons to achieve long-term success in case 1. Other authors have also reported the need for larger diameter as well as multiple balloons for dilating venous stenoses and recurrent venous stenoses after initial successful PTA (14–18). Balloons up to 50% larger than the size of the vessel have been used to dilate resistant anastomotic venous stenoses associated with dialysis fistulas (19).

The severe ascites in both patients and the large effusion in one were considered life threatening because of marked respiratory distress that was unresponsive to conventional medical management. Surgical correction of the suprahepatic IVC anastomotic stenosis was considered but was thought to be technically extremely difficult and probably impossible to perform. The only other surgical alternative was repeat transplantation; however, liver function in both patients was satisfactory at the time.

On the basis of previous experience with PTA in the successful treatment of vascular stenoses, this approach was considered a therapeutic alternative with minimal risk compared to surgical repair or repeat transplantation. Al-
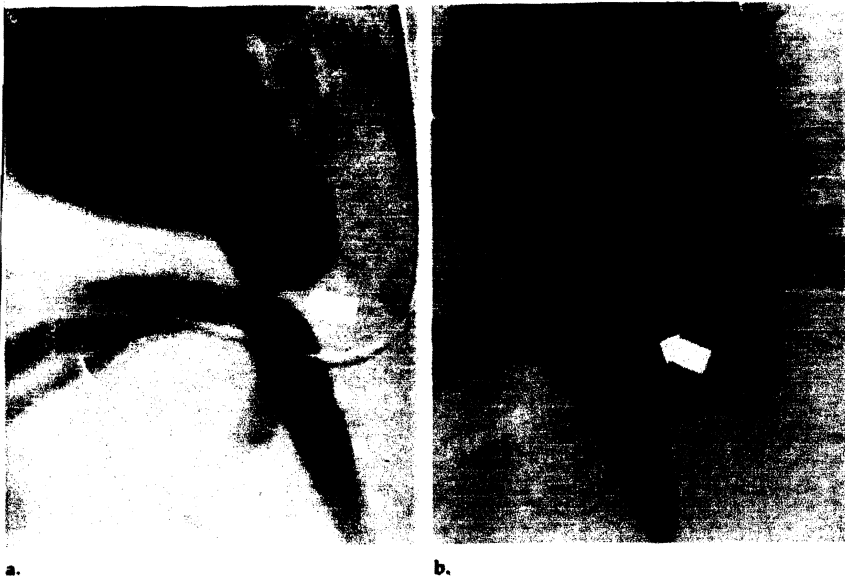
Figure 2. Case 2. (a) Inferior vena cavogram demonstrates stenosis of the suprahepatic IVC anastomosis (long arrow). The hepatic veins (arrowheads) and IVC (short arrows) are patent. (b) After PTA with an 8-mm balloon, the suprahepatic IVC anastomosis (arrow) is widely patent.

though most frequently applied to arterial stenoses, PTA has also been successfully used to treat various types of venous stenoses, including those in saphenous vein bypass grafts (20), the superior vena cava (14), and the portal vein (15). Hepatic vein and IVC stenoses causing the Budd-Chiari syndrome in patients without transplants have also been dilated successfully (16-18,21,22).

Cardella et al (8) reported successful PTA of a suprahepatic IVC anastomotic stenosis in a patient with a liver transplant. Their technique, as well as whether their patient had the Budd-Chiari syndrome, is not known. Recently, Rose et al (23) reported successful PTA of an infrahepatic IVC anastomotic stenosis in a liver transplant patient with massive lower extremity edema. Initial successful dilation in both of these cases was determined by the immediate post-PTA angiogram as well as by the significant reduction in the pre-PTA pressure gradient, as seen in our cases.

Although balloon angioplasty was repeated three times in case 1 and once in case 2, this nonsurgical approach resulted in resolution of ascites, pleural effusion, and respiratory distress in both patients. The patient described by Rose et al (23) also exhibited a dramatic clinical response to PTA. One of our patients (case 1) has remained asymptomatic for 31 months after the last PTA procedure. We believe that in cases of

posttransplantation anastomotic stenoses of the IVC, PTA should be considered the initial treatment of choice in selected patients. ■

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