Liver Transplantation as Treatment for Arterioportal Fistulae

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The authors present the case of a 14-year-old boy with secondary portal hypertension caused by arterio-portal fistulae. Ascites, edema, severe abdominal pain, and steatorrhea developed in the patient. In an attempt to decrease arterioporal flow the authors carried out vascular embolization that was partially successful. A liver transplant was performed as a last therapeutic resource in view of the fact that it constitutes an accepted therapy for patients with severe liver disease. Orthotopic liver transplantation may be considered as another possible treatment of arterio-portal fistulae.


INDEX WORDS: Arterio-portal fistulae, liver transplantation, vascular embolization.

INTRAHEPATIC arterio-portal fistulae (IAPF) is a rare disease and an unusual cause of portal hypertension (PH) in children. IAPF may be congenital or secondary to various causes such as trauma, family hereditary telangiectasia liver carcinoma, needle liver biopsy, percutaneous cholangiography, or catheterization of the bile duct. Delayed diagnosis is the rule because symptoms initially are subtle and nonspecific. Surgical repair or endovascular occlusion are the current choices for treatment. We report a case of congenital arterio-portal fistulae treated by orthotopic liver transplant as a therapeutic alternative for a complex disease.

CASE REPORT

A 14-year-old boy had been suffering from abdominal pain since he was 6 years old. At the age of 13 he presented with chronic diarrhea with steatorrhea. In the last month he had lost 10 kg and presented a continuous murmur over the hepatic area with normal cardiac functions. Laboratory data included albumin level of 3.0 g/dL; hemoglobin (Hb) value of 9.5 g/dL; hematocrit (Ht) level of 28%; blood platelet count of 160,000/mL; leukocyte count of 4,500 cells per milliliter; prothrombin time of 16 seconds; total bilirubin was 2 mg/dL, aspartate aminotransferase (AST) was 14 mg/dL, alanine aminotransferase (ALT) was 29 mg/dL, and chymotrypsin dosage in feces was within the normal range. Because he did not respond to nutritional therapy, 3 months later he was admitted to the Pediatric Department of the Hospital Italiano of Buenos Aires. On admission the child was chronically and severely ill. He weighed 38 kg and was 155 cm tall, which is the 10th and 25th percentile, respectively according to our standards. His body temperature was normal, but he was pale, and he suffered from intense abdominal pain. On physical examination he was noted to have a marked abdominal distension caused by ascites and splenomegaly, and he presented a continuous murmur over the hepatic area with normal cardiac functions.

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However, selective angiography is the prime standard method to define diagnosis and delineate the intrahepatic vessels, which allows one to pinpoint the location of the fistulae.

The treatment of IAPF is to close the porta-systemic connection to prevent portal hypertension. Porto-cava shunts are not advisable in hepatic arterio-venous fistulae because these procedures may lead to cardiac failure.

Arterio-portal fistulae can be treated by surgery or interventional techniques. The surgical procedures proposed are mainly fistula ligation and vascular reconstruction. Sometimes the liver reacts to increased portal venous flow or pressure by increasing its vascular resistance, so that ligation will not relieve portal hypertension. Intravascular embolization of the arteriovenous fistulae may be highly effective under certain anatomic conditions.

The treatment must be based on the location and extension of the fistulae. With this boy's type of anatomy, the multiplicity of the fistulae prevented a safe and complete embolization.

However, in an attempt to decrease the arterio-portal flow, we treated the high flow fistulae with occlusion techniques, which led to partial clinical improvement. We used two techniques: detachable balloonage to close the larger fistulae and NBCA to occlude some of the remaining microfistulae. The lowering of portal hypertension may have produced an increase in venous mesenteric flow, which decreased his abdominal pain and diarrhea. Although technically feasible, we decided not to perform

DISCUSSION

Fistulae from the hepatic artery to the portal vein are an unusual cause of portal hypertension in pediatric patients. They may occur either intra or extrahepatically. Several disease entities must be ruled out. In our patient the isolated vascular malformation without any other alteration should be considered as secondary to a congenital abnormality.

Clinical symptoms in the disease are initially subtle, and it may take years for bizarre symptoms to develop. It may present itself in a variety of forms. The usual signs and symptoms are abdominal pain and diarrhea and may precede other symptoms such as ascites and bleeding from varicose esophageal veins. The cause of abdominal pain in this case may have been mesenteric congestion, whereas the steatorrhea was interpreted as a manifestation of lymphatic vessel dilation and leak. Our patient did not present with congestive cardiac failure. This may be because of the protective mechanism of the large sinusoidal system of the liver.

Among image methods Doppler ultrasound scan, CT scan, and extremely rapid sequential CT are very useful noninvasive diagnostic methods. However, selective angiography is the prime standard method to define diagnosis and delineate the intrahepatic vessels, which allows one to pinpoint the location of the fistulae.

Fig 1. Microphotography of intestinal villi shows edema at the lamina propria and engorged lymphatic vessels, histological evidence of intestinal lymphangiectasia

Fig 2. Selective hepatic angiogram shows multiple intrahepatic arterio-portal fistulae arising from an enlarged left hepatic artery. The right hepatic artery was small in size and several small fistulae arose from this vessel to the portal vein. It presented a copious hepatofugal flow. P.V., portal vein; H.A., hepatic artery; R.H.A, right hepatic artery; L.H.A, left hepatic artery.
total occlusion of the portal portion of the vascular anomaly because of the high risk of liver failure given the extension of the abnormality and the atrophy of the hepatic right lobe resulting from the hypoplastic right hepatic artery. Failure of the above-mentioned procedures led us to believe that an orthotopic liver transplant was this child’s last chance for survival. Liver transplant presently constitutes an accepted therapy for patients with severe liver disease who do not respond to other forms of treatment. New trends in immunosuppression as well as technical advances have increased the number of pathologies that can be successfully treated. Orthotopic liver transplantation may be considered as another possible treatment for complex arterio-portal fistulae.

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