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Congenital intrahepatic arterioportal fistula presenting as severe undernutrition and chronic watery diarrhea in a 2-year-old girl

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Abstract Intrahepatic arterioportal fistula (IAPF) is a rare cause of portal hypertension in young children. We report the case of a 2-year-old girl with severe undernutrition, chronic watery diarrhea, and gastrointestinal bleeding because of a congenital intrahepatic arterioportal fistula. Radiographic embolization and surgical ligation of the left hepatic artery were attempted, with no resolution of the symptoms. So, a left lobectomy was performed, with excellent results and prompt disappearance of the diarrhea. Hepatectomy should be considered as a definitive and reliable therapy for congenital IAPF.

Intrahepatic arterioportal fistula (IAPF) is a rare cause of portal hypertension in young children. Most often it is acquired and develops either after trauma or iatrogenically after transhepatic interventions [1]. Congenital lesions are very rare with few cases described in the literature [2].

Arterioportal fistulas in infancy usually have initial symptoms including gastrointestinal (GI) bleeding episodes, ascites, and splenomegaly. Also, children can present with failure to thrive, diarrhea, and malabsorption [1,3].

The options for the treatment of this disease are percutaneous transarterial embolization [4-6], surgical ligation of the implicated hepatic artery [7,8], partial hepatectomy [9,10], portocaval shunt [3], and liver transplantation [11].

We report the case of a 2-year-old girl with a congenital IAPF and severe undernutrition successfully treated with left hepatectomy.

1. Case report

A 2-year-old girl was admitted to our pediatric surgical department in September 2008 with a history of severe melena. This admission was the seventh time in the last 6 months that she presented GI bleeding. In addition, she had had abdominal enlargement and chronic watery

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diarrhea since birth, with 10 to 15 episodes of liquid defecation a day. She had had 2 previous hospital admissions because of respiratory and urinary tract infections. Medical history showed an unremarkable pregnancy and delivery. Birth weight was 3 kg. There was no history of trauma or surgery.

On admission, physical examination revealed a pale undernourished child (body weight, 6.3 kg; lower than 2.5th percentile) with abdominal distension without any visceral enlargement. She underwent upper GI endoscopy that showed thick esophageal and gastric varices with signs of recent bleeding, beyond that of severe hypertensive gastropathy. A Doppler ultrasonography suggested an IAPF in the left lobe with hilar portal flux inversion.

As the clinical conditions were poor, an endovascular procedure was performed, and the left hepatic artery branch relating to the fistula was embolized (Figs. 1 and 2). Postembolization ultrasonography revealed persistent filling of the fistulous area with arterial blood. The watery diarrhea persisted, and she had another episode of GI bleeding.

Considering the extreme undernutrition of the child, the surgical team decided on a surgical ligature of the left hepatic artery, to control the portal hypertension and reduce the diarrhea. An eventual hepatectomy would be left for a second phase of intervention under better nutritional conditions. The procedure was accomplished without complications, but collateral arteries from the right lobe were responsible for persistence of the arterioportal fistula and continuation of the diarrhea.

We opted then for a short cycle of enteral nutrition followed by left hepatic lobectomy. The surgery was performed without hemorrhagic complications, and hemostasis of the cut surface was achieved by using an ultrasound dissector (CUSA, Valley Lab, Boulder, CO) complemented by coagulation of small vessels with bipolar diathermy and ligation of larger structures. The patient did not need any blood transfusions during or after the surgery.

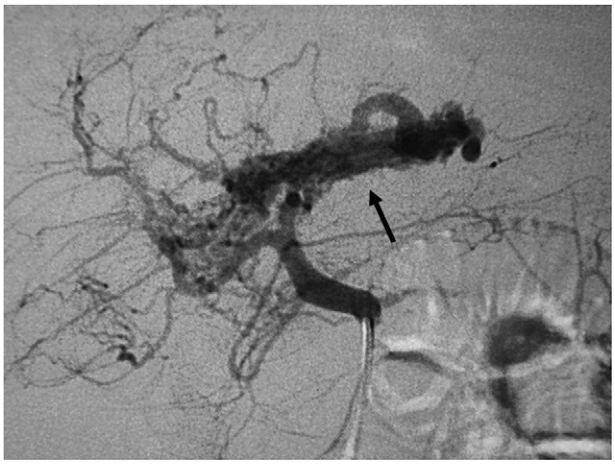

Fig. 1 Preintervention selective hepatic artery angiography showing a large arterioportal fistula between left hepatic artery branches and left portal vein branches (arrow).

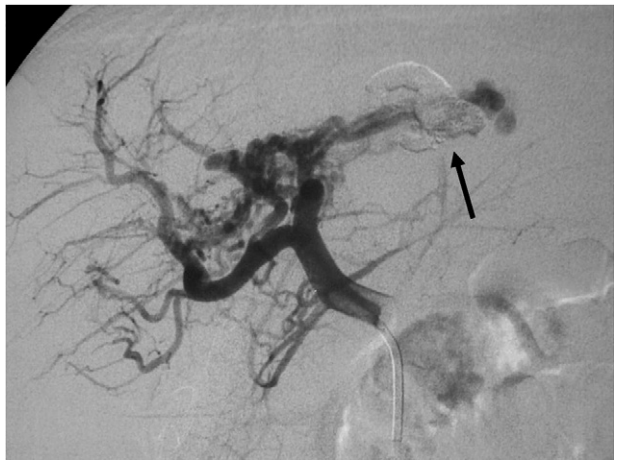

Fig. 2 Radiologic intervention on APF. Embolization (arrow) resulted in partial obliteration of the fistula.

In gross appearance, the resected liver, except segment IV, displayed normal parenchyma. The cut surface of segment IV showed a large portal vein branch with multiple holes, which were arterioportal fistulae. The histologic findings showed arteries filled with emboli and dilated venous branches (Fig. 3).

She had an uneventful postoperative period and was discharged on the seventh postoperative day. The watery diarrhea disappeared soon after the surgery. An upper GI endoscopy 10 days after the hepatectomy revealed a significant reduction in the esophageal and gastric varices.

By 30 days after the surgery, she had gained 2 kg, was having 2 to 3 solid evacuations per day, and had no more episodes of GI bleeding.

2. Discussion

Congenital IAPF can be defined as an intrahepatic communication between the hepatic artery and the portal venous system, without any communication with the systemic venous circulation, with no secondary cause or primary hepatic or biliary disease, and presenting before 18 years of age [2]. To date, a total of 33 cases have been reported in the literature, with the first report nearly 40 years ago [12].

Diffuse or multiple IAPFs are virtually always congenital in origin [9,13], whereas a solitary fistula is typically acquired [14]. Less than 10% of all arterioportal fistulas that involve the hepatic artery are congenital [15]. Secondary causes are more common: major blunt [16] or penetrating abdominal trauma [17], surgical procedures such as needle liver biopsy [18], Kasai portoenterostomy [19] and segmental liver transplantation [20], hepatic artery aneurysms [21], cirrhosis [22], hepatocellular carcinoma [23], biliary atresia [24], and hereditary hemorrhagic telangectasia [25].