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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.

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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
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.

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 telangiectasia [25].
Most symptoms and signs are caused by the development of portal hypertension [8,9,26]. As a result, splenomegaly, hypersplenism, esophageal varices, and ascites commonly develop. Failure to thrive and chronic diarrhea are observed in nearly 50% of the cases of congenital IAPF [2]. Intestinal dysfunction is related to venous congestion and stasis secondary to reversal of flow in the portal and superior mesenteric veins [12,27]. Furthermore, protein-losing enteropathy, steatorrhea, or evidence of fat malabsorption may occur and contribute to the malnutrition [2]. Interestingly, in our case, the development of chronic watery diarrhea occurred much earlier than the variceal bleeding, and the girl had no signs of portal hypertension on initial physical examination. This fact in combination with the rarity of congenital IAPF was probably responsible for the delay in diagnosing the cause of the diarrhea, which then led to this extreme undernutrition.

Definitive therapy is aimed at the obliteration of the shunt and the restoration of normal portal and hepatic arterial hemodynamics [1]. The options for treatment are surgical, percutaneous transarterial embolization (TAE), and a combination of TAE and surgery. Surgical approaches have included ligation of the implicated hepatic arteries or branches [9,26,27], fistula excision and direct vascular repair [28], and hepatectomy [9,10]. Liver transplantation has been proposed as a long-term option for cases of recurrent portal hypertension after failed embolization [11]. Recently, end-to-side portocaval shunt has been described as an alternative to hepatic artery embolization or ligation as a means to reduce portal pressure [3].

The interventional radiologic procedure has a high success rate in cases of unilateral lesions with a single feeding artery [2]. Vauthey et al [15] reviewed 88 cases from the literature, with IAPF from various etiologies and at various ages and found that TAE was successful in 42% of all cases. Complex congenital IAPFs are prone to collateralization or recurrence after radiologic intervention [1]. Hence, they may be difficult to treat without a combination of surgery and embolization. Partial hepatectomy is associated with high rates of mortality and morbidity [29,30]. In the current case, our team resisted the idea of performing such an extensive procedure because of the extreme malnutrition of the child. However, the intensity of the diarrhea and the abundance of the esophageal varices (without any response to embolization or surgical ligation of left hepatic artery) led us to attempt definitive aggressive treatment of the IAPF. Surprisingly, the child tolerated the surgery well, with no complications and an early hospital discharge.

A key point that probably contributed to this favorable outcome was the previous surgical ligation of the left hepatic artery. Although it could not stop the symptoms of portal hypertension, the reduction of blood flow to the cut surface of liver during left lobe resection led to few hemodynamic disturbances, with no need for red blood cell transfusion. Although many authors recommend multiple TAEs as the best choice to treat complex IAPF [2,31,32], we believe that such procedures have a considerable risk of complications (as thrombosis of femoral artery and portal vein) [33], with a high percentage of failure or recurrence of symptoms. By the other hand, partial liver resections, performed by groups with great experience in hepatic surgery, may be a procedure with low rates of morbidity, even in cases of children with extreme undernutrition and other comorbidities [34].
In conclusion, our experience in the present case suggests that hepatectomy, preceded by a surgical procedure to diminish arterial flow to the lesion, should be considered as a definitive and reliable therapy for congenital IAPF.

References