Emergency hepatectomy for hepatic arteriovenous malformation combined with pulmonary hypertension in an infant

Naruhiko Murase, Hiroo Uchida, Akihide Tanano, Chiyoe Shirotani, Akinari Hinoki, Takahisa Tainaka, Kazuki Yokota, Kazuo Oshima, Ryo Shirotu

Department of Pediatric Surgery, Nagoya University Graduate School of Medicine, 65 Tsurumai, Showa, Nagoya 466-8550, Japan

Abstract

Patients with hepatic arteriovenous malformations rarely present with pulmonary hypertension. We report the case of a 3-month-old boy who developed severe pulmonary hypertension due to a hepatic arteriovenous malformation. The use of pulmonary vasodilators to treat the patient's pulmonary hypertension worsened his high-output heart failure. This is the first case in which emergency hepatectomy rescued a patient with hepatic arteriovenous malformations who developed pulmonary hypertension.

1. Case report

A 1786 g male was delivered by Caesarean section at 34 weeks of gestation at another hospital. An echocardiogram showed an atrial septal defect, and the child's respiratory condition continued to worsen. Although the patient underwent radical surgery for the atrial septal defect at 3 months of age, his respiratory condition did not improve after the operation, and a postoperative echocardiogram detected pulmonary hypertension. After postoperative treatment with nitric oxide and prostaglandin I2, the patient's respiratory condition worsened considerably. A contrast-enhanced computed tomography (CT) scan of the patient's whole body was performed and showed an arteriovenous malformation in the left lobe of the liver (between multiple feeding vessels from the left hepatic artery and a markedly dilated left hepatic vein) (Fig. 1). The left and right hepatic arteries were normally bifurcated from the proper hepatic artery. There were no findings of hypoperfusion on the right lobe in spite of low resistance in the left hepatic artery. The left and right hepatic arteries were normally bifurcated from the proper hepatic artery. There were no findings of hypoperfusion on the right lobe in spite of low resistance in the left hepatic artery. The patient was referred to our hospital on the 11th day after the cardiac surgery.

When he arrived at our hospital, he was intubated and exhibited a partial pressure of arterial oxygen of 60–70 mm Hg under mechanical ventilation (inspiratory O2 fraction: 100%, positive end expiratory pressure: 10 cm H2O, peak inspiratory pressure: 40 cm H2O). A chest X-ray showed bilateral pulmonary congestion. An echocardiogram demonstrated that the atrial septal defect had closed, but all of his pulmonary veins were markedly dilated, and his estimated systolic pulmonary artery pressure was about 50 mm Hg. A blood test detected a very high brain natriuretic peptide level (8334 pg/ml). On the other hands, the patient did not present with liver dysfunction because his total bilirubin...
concentration was 1.6 mg/dl, serum aspartate transaminase and alanine transaminase levels were 22 and 9 IU/l, and International Normalized Ratio was 1.4. The patient was diagnosed with pulmonary hypertension and high-output heart failure. We had a conference with pediatric cardiologists and interventional radiologists regarding whether transcatheter embolization could be performed to treat the patient’s HAVM; however, they suggested that transcatheter arterial embolization would not effectively improve his high output cardiac failure or flow-PAH due to the presence of multiple collateral arteries. In order to reduce the left-to-right shunts as soon as possible, we performed an emergency left hepatic lobectomy. The patient’s body weight was 3700 g at the time of the operation.

During laparotomy, an arteriovenous malformation was seen in the left lobe of liver, and markedly dilated vessels were observed on the surface of the gallbladder (Fig. 2). The feeding vessels included multiple vessels from the left hepatic artery and collateral arteries from the regions surrounding the hepatoduodenal ligament and left triangular ligament. After dealing with the feeding vessels, a formal left hepatectomy was performed. The operation took 238 min and involved 665 ml of intraoperative blood loss. The patient’s postoperative course was uneventful. After the operation, the patient’s respiratory condition improved markedly, and he was successfully extubated 6 days after the procedure. A postoperative chest X-ray revealed that the patient’s pulmonary congestion had improved, and an echocardiogram did not detect pulmonary hypertension. A blood test performed on postoperative day 2 showed that the patient’s brain natriuretic peptide level had normalized (553 pg/ml). Although the postoperative liver function tests were elevated temporarily (serum aspartate transaminase and alanine transaminase levels on postoperative day 1 were 165 and 52 IU/l, respectively), they were normalized 5 days after the procedure. He was discharged in a healthy condition 13 days after the hepatectomy. A follow-up examination performed at 16 months after the hepatectomy demonstrated that the patient was asymptomatic, and an abdominal CT scan did not show any abnormalities.

2. Discussion

Patients with HAVM rarely develop pulmonary hypertension in early infancy [2–6]. Pulmonary hypertension is categorized into 5 groups: pulmonary arterial hypertension (Group 1; PAH), pulmonary hypertension due to left heart disease (Group 2), pulmonary hypertension due to chronic lung disease and/or hypoxia (Group 3), chronic thromboembolic pulmonary hypertension (Group 4), and pulmonary hypertension due to unclear multifactorial mechanisms (Group 5) [7]. Most cases of pediatric pulmonary hypertension involve PAH (Group 1). As the use of a specific threshold for
increased pulmonary vascular resistance was abandoned as a diagnostic criterion for PAH, Group 1 also includes patients with pulmonary hypertension caused by increased pulmonary blood flow due to large left-to-right shunts (flow-PAH), who can exhibit normal pulmonary capillary wedge pressure levels and normal or even low pulmonary vascular resistance [7,8]. In a study of a large registry of patients with pediatric pulmonary hypertension conducted in the Netherlands, it was found that 2845 of 3265 (87%) pediatric patients with pulmonary hypertension had PAH. These patients were divided into two categories according to their prognosis; i.e., progressive PAH (n = 154; 5%) and transient PAH (n = 2691; 82%) [9]. The patients with transient PAH included numerous patients with flow-PAH (n = 1112). However, almost all of the patients with flow-PAH (n = 1110) had congenital heart defect-associated PAH, and there were few cases in which peripheral left-to-right shunts were an important component of the patient’s PAH [9]. In particular, flow-PAH resulting from HAVM in early infancy seems to be very rare, and only 7 case reports (including ours) about the condition were found in the literature [2–6]. Most cases (6/7) were associated with high-output heart failure, and the prognosis of HAVM-induced flow-PAH is not very good as 43% of the patients (3/7) died (Table 1). Our case is the first in which emergency hepatectomy rescued a patient with HAVM who developed flow-PAH.

Flow-PAH and PAH resulting from increased pulmonary vascular resistance are poles apart in terms of pulmonary blood flow, and it is necessary to treat flow-PAH with care because the use of pulmonary vasodilators will worsen high-output heart failure associated with flow-PAH [4]. In our case, the HAVM-induced flow-PAH could not be detected before the start of pulmonary vasodilator treatment. However, it is possible that the emergency procedure could have been avoided if pulmonary vasodilators had not been used. Indeed, a neonatal case was reported in which medical therapy in the form of diuretic agents and captopril improved HAVM-induced flow-PAH without the need for invasive procedures [6].

Although no standardized recommendations for the management of HAVM have been developed due to the rarity of the condition, transcatheter embolization followed by surgical resection is often employed [1]. In our case, we consulted interventional radiologists and pediatric cardiologists, but they concluded that transcatheter arterial embolization would not be effective against our patient’s HAVM because embolizing the dominant feeding vessels would lead to rapid enlargement of the minor feeding vessels [10]. Transcatheter arterial embolization or ligation of the proximal feeding arteries is only temporarily effective, and the rapid recruitment of collateral arteries makes further treatment difficult or impossible [10,11]. Emergency hepatectomy is associated with significant risks, especially in small, sick infants. If our patient presented with preoperative liver dysfunction, the postoperative course might be troublesome. However, emergency hepatectomy is sometimes the only definitive treatment for cases of HAVM involving urgent complications. Among the literature, we found a case in which hepatectomy was performed for a hepatic arterioportal malformation and intestinal hypoperfusion in a neonate (2700 g) [12].

In conclusion, HAVM-induced flow-PAH is rare in early infancy. However, care should be taken when treating flow-PAH because the standard treatment for PAH resulting from increased pulmonary vascular resistance; i.e., the administration of a pulmonary vasodilator, can worsen high-output heart failure. Emergency hepatectomy is an effective treatment for HAVM-induced flow-PAH, even in small, sick infants.

Conflicts of interest

The authors declare that they have no conflicts of interest.

Informed consent

Informed consent was obtained from participants in this article.

References


Table 1

Reported cases of hepatic arteriovenous malformations combined with persistent pulmonary hypertension in early infancy.

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Onset</th>
<th>Location</th>
<th>Other clinical presentations</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bamford</td>
<td>1980</td>
<td>At birth</td>
<td>Left lobe</td>
<td>HOHF</td>
<td>Digoxin, furosemide</td>
<td>Died on day 3</td>
</tr>
<tr>
<td>Paley</td>
<td>1997</td>
<td>1 month</td>
<td>NA</td>
<td>HOHF, skin hemangioma</td>
<td>Hepatic artery ligation</td>
<td>Died after 22 months</td>
</tr>
<tr>
<td>Alexander</td>
<td>2006</td>
<td>At birth</td>
<td>Left lobe</td>
<td>Hepatomegaly, skin hemangioma</td>
<td>Embolization, liver transplantation</td>
<td>Alive</td>
</tr>
<tr>
<td>Thatrimontrichai</td>
<td>2012</td>
<td>At birth</td>
<td>Left lobe</td>
<td>HOHF, skin hemangioma</td>
<td>Embolization</td>
<td>Alive</td>
</tr>
<tr>
<td>Agha</td>
<td>2015</td>
<td>At birth</td>
<td>Left lobe</td>
<td>HOHF</td>
<td>Digoxin, furosemide, captopril</td>
<td>Alive</td>
</tr>
<tr>
<td>Our case</td>
<td>2015</td>
<td>3 months</td>
<td>Left lobe</td>
<td>HOHF</td>
<td>Hepatectomy</td>
<td>Alive</td>
</tr>
</tbody>
</table>

NA: not available, HOHF: high-output heart failure.