Successful Treatment Using Coil Embolization of a Symptomatic Intrahepatic Portosystemic Venous Shunt Developing through a Patent Ductus Venosus in a Noncirrhotic Adult

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Abstract

The patient was a 60-year-old man with encephalopathy without liver cirrhosis. CT angiography revealed a patent ductus venosus between the anterior segmental branch of the portal vein and the middle hepatic vein. Coils were framed in the patent ductus venosus and then used to fill in the frame. After treatment, transarterial portography showed that the shunt flow of the ductus venosus had decreased significantly. After one day, the patient’s disturbance of consciousness disappeared. Our case involved the adult-onset of a patent ductus venosus, which is extremely rare. This case is the first in which coil embolization was successfully achieved in a noncirrhotic elderly patient with a patent ductus venosus.

Key words: coil embolization, hepatic encephalopathy, intrahepatic portosystemic shunt, patent ductus venosus

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Introduction

Portal-systemic venous shunts are formed by portal hypertension accompanied by hepatic fibrosis in cirrhotic cases that subsequently leads to the development of hepatic encephalopathy accompanied by hyperammonemia (1). On the other hand, shunts are also formed in noncirrhotic patients (1). The cause of shunt formation in the liver is sometimes unclear (2). Consequently, cases of hepatic encephalopathy can be misdiagnosed as dementia or other psychoneurological diseases (3). Detection of noncirrhotic cases may increase in association with progress in and spread of abdominal imaging diagnostic techniques such as abdominal ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI). One adult case of a single shunt has been reported in which the shunt was formed by a patent ductus venosus developing between the left branch of the portal vein and the left hepatic vein (4). The ductus venosus, which connects the umbilical vein and the inferior vena cava during the fetal stage, closes immediately after birth (5). Therefore, adult cases of patent ductus venosus are extremely rare, with only eight cases having been reported globally (6-10). Moreover, there are no reports describing the use of coil embolization as a treatment for a patent ductus venosus as a shunt vessel.

We herein report an extremely rare adult case of a symptomatic intrahepatic portosystemic venous shunt developing through a patent ductus venosus. Moreover, we describe the first noncirrhotic elderly patient with a patent ductus venosus to undergo successful treatment using coil embolization.

Case Report

The patient was a 60-year-old man with a mild consciousness disturbance classified as 1 on the Japan coma scale. He had a past history of a skull fracture due to a head injury that occurred in an accident without abdominal injury in...
March 2002. The patient became a heavy drinker, consuming 40 g/day every day, starting in March 2002. He was diagnosed with alcoholic liver dysfunction and diabetes mellitus; however, he did not take any medications. He did not have hypertension, hypercholesterolemia, heart disease or mental retardation and did not have a family history of liver disease, heart disease or diabetes mellitus. In a local hospital in February 2008, he was evaluated for a flapping tremor. Head MRI showed the basal nucleus to have high intensity on T1-weighed images, although no brain atrophy was found. Therefore, the patient was suspected of having hepatic encephalopathy. For the treatment of the encephalopathy, nutritional support by a dietitian was provided to ensure a low-protein diet, and lactulose was administered. However, the disturbance of consciousness did not improve. The patient was admitted to our hospital for further examination of the encephalopathy.

On admission in April 2008, the patient’s body temperature was 36.2°C and his body height and weight were 162.4 cm and 65.8 kg, respectively. His body mass index was 24.9 kg/m². His abdomen was soft and flat and his bowel sounds were normoactive. The liver was not palpable and was without tenderness. The blood test results obtained on admission were as follows: white blood cells, 3,600/μL; hemoglobin, 8.5 g/dL; MCV, 70 fl; MCH, 21.9 pg; platelet count, 11.8×10⁹/μL; prothrombin time (PT), 73.0%; C-reactive protein, 0.12 mg/dL; albumin (Alb), 3.1 g/dL; total bilirubin (T-Bil), 1.3 mg/dL; aspartate aminotransferase (AST), 27 IU/L; alanine aminotransferase (ALT), 15 IU/L; lactate dehydrogenase, 288 IU/L; alkaline phosphatase (ALP), 573 IU/L; γ-glutamyltransferase, 37 IU/L; choline esterase, 196 IU/L; ammonia, 100 μg/dL; hyaluronic acid (HA), 49.4 ng/mL; total bile acid, 136.7 μmol/L; HbA1c, 8.7%; HBs Ag, 0.00 (-); and HCV Ab, (-).

An abdominal dynamic CT scan revealed an intrahepatic portosystemic shunt between the anterior segmental branch of the portal vein and the middle hepatic vein during the portal phase (data not shown). On CT arterial portography, the anterior segmental branch of the portal vein was first found to be enhanced and significantly dilated with a diameter of 16 mm (Fig. 1A). Then, the ductus venosus was found to be enhanced and exhibited strong tortuosity (B). The ductus venosus merged with the middle hepatic vein (C), then the inferior vena cava was enhanced (D).

To confirm the presence of a shunt vessel, transarterial portography was performed via the right femoral artery using a 4-French RC catheter (Medikit Co., Ltd., Tokyo, Japan). The ductus venosus was consistent with a shunt vessel (Fig. 2A). To measure the pre-embolization hepatic vein wedge pressure, transvenous portography via a right femoral vein approach was also performed using a 4-French shepherd’s hook catheter (Medikit Co., Ltd., Tokyo, Japan). The hepatic vein wedge pressure was 11 mmHg before and after

![Figure 1. CT arterial portography of the intrahepatic portosystemic venous shunt developing through a patent ductus venosus. On CT arterial portography, the anterior segmental branch of the portal vein was first found to be enhanced and significantly dilated with a diameter of 16 mm (A). Then, the ductus venosus was found to be enhanced and exhibited strong tortuosity (B). The ductus venosus merged with the middle hepatic vein (C), then the inferior vena cava was enhanced (D).]
occlusion by a balloon catheter (Terumo Clinical Supply, Tokyo, Japan). The pressure was within the normal range. The middle hepatic vein as a drainage vein was occluded by a balloon catheter, then five IDC detachable coils (Boston Scientific, Ireland) ranging from 20 to 30 mm in diameter were framed in the ductus venosus. Ten Tornade coils (0.035 inches) (Cook Medical, Bloomington, IN, USA) ranging from 8 to 10 mm in diameter and 35 microcoils (Johnson & Johnson, MA, USA) were used as embolic materials to fill in the frame. Transarterial portography showed that the shunt flow of the ductus venosus had decreased significantly (B). After one day, the patient’s disturbance of consciousness disappeared. The serum levels of ammonia and total bile acid improved (36 μg/dL and 35.9 μmol/L, respectively). After five months, the patient’s liver function also improved as follows: PT, 82.4%; Alb, 4.4 g/dL; and T-Bil, 1.0 mg/dL. He provided written consent to undergo treatment with coil embolization. The treatment complied with the standards of the Declaration of Helsinki and current ethical guidelines.

**Discussion**

The ductus venosus connects the umbilical vein and the inferior vena cava during the fetal stage. It closes rapidly after birth as a consequence of a decrease in pressure in the portal sinus, which results in retraction and narrowing of its origin (5). The ductus venosus is soon obliterated and transformed into the ligamentum venosum that connects with the ligamentum teres. In the adult liver, it runs in the fissure between the anatomic left and right lobes and courses posteriorly to reach the inferior vena cava (8). A patent ductus venosus is a possible cause of an intrahepatic portosystemic venous shunt (11-14), and many pediatric cases have been reported since Barjon et al. described the first case in 1972 (4). When the ductus venosus remains patent, there is a risk of progression to liver dysfunction, including cirrhosis and hepatic insufficiency. There have been many reports of pediatric patients showing hyperammonemia, liver dysfunction, retardation of mental development and portal-systemic encephalopathy due to retention of the portal-hepatic vein shunt (15, 16). In our case, however, the patient grew up without encephalopathy or mental retardation. At 60 years of age, he suffered from hepatic encephalopathy but not cirrhosis or hepatic insufficiency. He did not undergo a liver biopsy; therefore, a detailed examination of liver fibrosis was not performed. The serum level of HA has been reported to be useful as an alternative indicator of liver fibrosis (17, 18). In our case, the serum HA level was slightly elevated within the normal range; therefore, it is speculated that the liver fibrosis would be mild, not cirrhotic, in our case. Adult-onset cases of intrahepatic portosystemic venous shunts developing through a patent ductus venosus are extremely rare, with only nine reported cases (including the present case) (6-10). In this case, the age of onset was 60 years, which is older than all patients in the previous reports (6-10). Adult-onset cases of patent ductus venosus have been shown to exhibit a male predominance: all cases occurred in men. The patients’ ages (in years) were distributed as follows: one: 20-30, two: 30-39, five: 40-49, zero:
50-59 and one: 60-69. There is a peak in the age distribution at 40-49 years of age.

If patent ductus venosus is congenital, one important question arises: Why do patients not develop encephalopathy until 60 years of age? A previous report showed that the shunt ratio is important for determining the age of onset of encephalopathy (15). In our case, we were not able to examine the shunt ratio in the ductus venosus; however, the hepatic vein wedge pressure was 11 mmHg, which was within the normal range. This suggests that the extent of the blood flow in the ductus venosus was not severe. This might explain why the age of onset in our case was older than that reported in other cases. On the other hand, a previous report showed that ammonia production in the intestines was suppressed in a case of congenital absence of the portal vein (19). Homeostatic control of ammonia or other metabolites might become gradually disturbed with age or other influences, such as the excessive intake of alcohol observed in this case starting March 2002.

Conservative therapy for encephalopathy is chosen in most cases to manage the symptoms of patent ductus venosus in adults. The most basic therapy applied for encephalopathy is a low-protein diet (approximately 40 g per day) (1). Disaccharides, lactulose and lactitol (syrup, powder, jelly, etc.) are also administered to clean the intestinal tract in order to acidify the intestinal lumen (1). Furthermore, the oral administration of non-absorbable antibiotics is attempted, if necessary (1). These treatments are similar to those used in cirrhotic patients with hepatic encephalopathy. In our case, lactulose was administered and nutritional support was provided by a dietitian to ensure a low-protein diet. However, these conservative therapies did not ameliorate the encephalopathy. In previous cases, surgical ligation of the ductus venosus was performed (10, 15). However, in some of these cases, the bowel became edematous and engorged postoperatively and the placement of a surgical portocaval shunt was necessary to relieve the overflow of the portal vein (6). In addition, thrombotic thrombocytopenic purpura occurred after surgery, which was speculated to have been induced by injury to the endothelium of the vein by the sudden alteration in the portal blood flow (7). Performing surgical occlusion with a shunt is generally very invasive and does not necessarily achieve good results. Obliterating the shunt using interventional radiology would be less invasive. Therefore, we believed that it was necessary to attempt this in our case. It has been reported that coil embolization using interventional radiology has been performed in some cases of intrahepatic portosystemic venous shunts, but not in cases of patent ductus venosus (20-22). In addition, coil embolization of patent ductus venosus has been performed in many dogs and cats, but not in human subjects (23-25). This patient is the first to undergo coil embolization for obliteration of a patent ductus venosus. After treatment, the patient’s disturbance of consciousness disappeared and the levels of serum ammonia and total bile acids improved significantly. No complications occurred in this case.

Ammonia is an indirect marker of portosystemic venous shunt presence, and the serum bile acid level is also proposed to be a marker of shunt existence (26). Bile acids are synthesized exclusively from cholesterol within hepatocytes, excreted into the biliary tract and stored in the gallbladder. After a meal, bile acids are excreted into the duodenum to help solubilize dietary lipids. They are reabsorbed in the distal ileum and transported back to the liver via the portal vein. In the enterohepatic circulation, almost all bile acids are removed from the portal blood by hepatocytes and recycled back into the biliary system. Increases in the serum bile acid concentrations can be induced by the existence of portosystemic venous shunts (27).

In conclusion, our patient had an extremely rare case of adult-onset intrahepatic portosystemic venous shunt developing through a patent ductus venosus. Moreover, our patient was older than all patients described in previous reports. In addition, our case is the first in which coil embolization was successfully achieved in a noncirrhotic elderly patient with an intrahepatic portosystemic venous shunt developing through a patent ductus venosus.

The authors state that they have no Conflict of Interest (COI).

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