Hepatic encephalopathy secondary to porto-systemic shunt satisfactorily treated with interventionist radiology


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RESUMEN

La encefalopatía hepática es un estado reversible de alteración en la función cognitiva, que puede ocurrir en pacientes con enfermedad hepática aguda o crónica o shunts porto-sistémicos, en el que puede aparecer cualquiera de los signos neurológicos o psiquiátricos conocidos. Las sustancias nitrogenadas procedentes de la digestión intestinal alcanzan el cerebro sin la depuración que supone su paso por el hígado, debido a las derivaciones porto-sistémicas, y dan lugar a los signos característicos de la encefalopatía hepática. A continuación presentamos dos casos clínicos de pacientes con shunt porto-sistémicos, diagnosticados de encefalopatía hepática crónica recurrente refractaria al tratamiento médico convencional, tratados satisfactoriamente con embolización de dicho shunt mediante técnicas de radiología intervencionista.


INTRODUCTION

Patients with chronic hepatopathy and portal hypertension frequently present with spontaneous porto-systemic shunts. Although helping to reduce portal pressure, the presence of these shunts causes an increase in toxic substances in the circulation and favors the development of hepatic encephalopathy. This type of patient needs to be very carefully managed since, as a direct consequence of the elimination of the shunt, there will be an inevitable increase in portal pressure (1). The most frequent types of shunts are spleno-renal and the gastro-renal shunts (2).

We present two cases of recurrent chronic hepatic encephalopathy secondary to the presence of a porto-systemic shunt. The condition was satisfactorily treated using interventionist radiology techniques.

CASE 1

A 74-year woman with hepatitis C virus (HCV) cirrhosis and associated hemochromatosis, stage B8 on the Child-Pugh scale, was admitted to hospital on multiple occasions (17 admissions in 20 months) with the diagnosis of severe recurrent hepatic encephalopathy. Conventional abdominal echography showed a heterogeneous
hepatic parenchyma without space-occupying lesions consistent with chronic liver disease, a patent portal vein 13 mm in diameter, and an oversized homogeneous spleen 13 cm in diameter. A doppler assessment showed inverted flow in the portal and splenic veins, with no clear porto-systemic shunt. Hence we performed an angio-CT, which demonstrated the presence of a marked collateral circulation associated with a porto-renal shunt (Fig. 1). The patient had persistently elevated levels of ammonia. Using interventionist radiology techniques we proceeded to partially embolize the shunt with micro-coils. A notable slowing of blood flow through the shunt was achieved (Fig. 1). Abdominal echography post-embolization showed a portal hepato-petal flow with persistent hepato-fugal flow in the splenic vein, together with a decrease in peri-splenic collateral circulation.

Following embolization the patient experienced immediate clinical improvement, and ammonia levels returned to normal (Fig. 2). Within 18 months after embolization the patient needed hospitalization on three occasions for mild hydropic decompensation, hepato-renal syndrome, and grade-I hepatic encephalopathy. A post-embolization oral endoscopy was performed, in which no esophageal-gastric varices were observed.

CASE 2

A 49-year-old male with alcoholic cirrhosis together with HCV infection, stage B7 on the Child-Pugh scale, was admitted to hospital because of altered consciousness. Ammonia levels during hospitalization were persistently elevated. Abdominal echography indicated a peri-pancreatic collateral vessel that was very dilated and tortuous, heterogeneous hepatic parenchyma without space-occupying lesions, and homogeneous splenomegaly 15 cm in diameter. A subsequent angio-CT showed the presence of abundant collateral circulation associated with a porto-systemic spleno-renal shunt (Fig. 3). Using interventionist radiology techniques we performed a partial embolization of the shunt with micro-coils (Fig. 3). An abdominal echography post-embolization indicated that the spleno-renal shunt had disappeared. The patient had a rapid improvement of clinical (disappearance of somnolence) and laboratory parameters (normalization of ammonia levels) following embolization (Fig. 2). He developed neither gastro-esophageal varices nor ascites. He has not been admitted to hospital over the past 10 months of follow-up.

DISCUSSION

The metabolic derangements that accompany end-stage cirrhosis can favor the development of hepatic encephalopathy. However, a patient with porto-systemic shunt can develop hepatic encephalopathy despite having a well-preserved hepatic function (1,2). Up to 46% of cirrhotic patients with marked collateral circulation will develop hepatic encephalopathy. Traditionally, hepatic encephalopathy has been treated medically, and only in exceptional cases invasive procedures have been employed. Essentially, medical treatment consists of eliminating precipitating factors, reducing the load of nitrogenated products from the intestine into the circulation by administering non-absorbable disaccharides and/or...
The first surgical correction of a congenital porto-systemic shunt was performed in 1982. Two years later the first occlusion of a shunt was performed using interventionist radiology techniques. Kimura et al. (4) studied 460 porto-systemic shunts in patients with portal hypertension. Of these, 14% (48 patients) had left-sided gastro-renal shunts, 5% (23 patients) had posterior gastro-renal shunts, 6% (28 patients) had short gastro-renal shunts, 7% (32 patients) had spleno-renal shunts, 2% had inferior mesenteric shunts, 1.3% had superior mesenteric shunts, and 0.4% had pancreatic-duodenal shunts. Takashi et al. (5) reported 15.7% spleno-renal shunts and 5.6% gastro-renal shunts in patients with portal hypertension and marked collateral circulation.

There have been cases reported of hepatic encephalopathy with highly elevated levels of ammonia in non-cirrhotic patients without portal hypertension due to presence of porto-systemic shunts, either congenital or acquired. The exact etiology is not known, and frequency appears to be increasing due, perhaps, to improved imaging techniques (6,7). Possibly, some of these cases had been erroneously diagnosed as psychiatric disorders such as dementia and depression (8). Hepatic encephalopathy secondary to the presence of a porto-systemic shunt needs to be considered in the differential diagnosis when neurological signs reliably indicate hepatic encephalopathy, although objective and subjective symptoms or laboratory measurements are not sufficient to suggest liver cirrhosis. As such, patients without cirrhosis and with preserved liver function can develop difficult-to-treat chronic hepatic encephalopathy when the presence of a potential porto-systemic shunt is not considered (6).

Surgical techniques are effective in controlling chronic hepatic encephalopathy secondary to the presence of por-
to-systemic shunt, but are as well associated with high mortality rates. Hence, the current preference is to employ interventionist radiology techniques rather than surgical procedures. There is no consensus, but it is widely accepted that the embolization of a porto-systemic shunt should be performed when blood flow through the shunt is considerable (inverse flow), which is observed by doppler echography prior to embolization. Essentially, the technique used is based on metal micro-coils, although shunt occlusion can also be attained using a balloon or sclerosing solutions such as ethanol or ethanalamine olete. On occasions a combination of the two techniques can be used-injection of a sclerosing solution followed by the introduction of metal micro-coils into the shunt so as to achieve complete occlusion (8).

The closure of a porto-systemic shunt can encourage the development of ascites and the formation of esophageal-gastric varices. Hence, these patients need to be followed-up closely following the procedure. Other complications described in the literature are mild and include onset of fever within 24-48 hours after embolization, or development of small bilateral pleural effusion. The most debilitating adverse effects are the development of esophageal or gastric varices, which in some cases may manifest as massive bleeding (9). Hence, some authors propose performing partial splenic embolization simultaneously with the embolization of the porto-systemic shunt or immediately post-operatively. This additional treatment ensures that the increase in portal vein pressure is not as much elevated as when the shunt is the only embolized site (10,11). Given the general status of the patients described in our present communication we opted for close endoscopic follow-up, and we did not perform any additional splenic embolization. Neither of the two patients described developed esophageal-gastric varices during follow-up.

A recent long-term follow-up study (5 years) of 25 patients with recurrent chronic encephalopathy secondary to porto-systemic shunt divided the patients into two groups: 14 patients in whom the shunt was embolized using interventionist techniques followed by partial splenic embolization, and a second group of 11 patients in whom only porto-systemic embolization was performed. Ammonia values and extent of hepatic encephalopathy in the group of patients who underwent additional splenic embolization were significantly lower at 6 months, 9 months, 1 year, and 2 years following the procedure. The authors concluded that combined therapy benefited the patients with recurrent chronic encephalopathy secondary to the presence of a port-systemic shunt (12). Splenic embolization can improve the capacity for protein synthesis in the liver, and as such increase serum levels of albumin, and further reduce the risk of hemorrhage while improving thrombopenia (13).

In conclusion, recurrent chronic hepatic encephalopathy secondary to the presence of porto-systemic shunt may not only occur in cirrhotic but also in non-cirrhotic patients. In general, this type of hepatic encephalopathy responds poorly to conventional medical therapy. The treatment-of-choice in cases not satisfactorily controlled with medical treatment should be embolization of the shunt using minimum invasive techniques such as those based on interventionist radiology. Given the elevated risk of developing esophageal-gastric varices following the procedure, it is recommendable to perform a supplemental partial splenic embolization.

REFERENCES