Unsuccessful transjugular intrahepatic portosystemic shunt for a patient with right heart failure and portal hypertension

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Abstract

A 60-year-old women with a history of congenital pulmonary valve stenosis developed right heart failure, cardiac cirrhosis and end-stage renal disease requiring renal replacement therapy. Cirrhosis was complicated by portal hypertension, resulting in intractable gastro-intestinal bleedings despite optimal treatment with beta-blockers and endoscopic band ligation. Because of fears for worsening right heart failure, a decision for placement of a transjugular intrahepatic portosystemic shunt (TIPS) was initially turned down. However, as intractable bleeding problems persisted and caused heavy transfusion needs, TIPS was ultimately performed as a rescue procedure. Although TIPS successfully reduced the hepatic venous pressure gradient from 16 mmHg to 4 mmHg, portal pressure remained high at 14 mmHg because of persisting right heart failure with elevated central venous pressure. Hepatic encephalopathy soon developed after TIPS placement and culminated in multi-organ failure after another episode of gastro-intestinal bleeding. At this point, the family of the patient decided to withdraw care and the patient died subsequently. This case illustrates how important it is to diagnose and optimally treat right heart failure before cardiac cirrhosis with its impending complications emerges. Although TIPS may effectively treat complications of portal hypertension in the context of cirrhosis, persisting right heart failure may abrogate its beneficial effects. (Acta gastroenterol. belg., **2017**, 80, **63-66**).

Key-words : right heart failure; cardiac cirrhosis ; portal hypertension ; transjugular intrahepatic portosystemic shunt.

Introduction

Portal hypertension is an important complication of cirrhosis that causes congestive gastropathy and variceal bleedings in the gastro-intestinal system. Transjugular intrahepatic portosystemic shunt (TIPS) placement is normally a very effective strategy to lower portal pressure and reduce the gastro-intestinal bleeding risk (1). However, the evidence for TIPS is limited in the setting of extensive cardiac and renal comorbidities and there might be a concern for worsening right heart failure when venous return is increased (2). We describe the case of a woman with right heart failure due to pulmonary valve stenosis, who developed cirrhosis with portal hypertension and intractable gastro-intestinal bleeding problems, for which TIPS was ultimately performed.

Case report

A 60-year-old woman was admitted to the nephrology department with epigastric pain, hematemesis and lower leg oedema. Arterial blood pressure was 90/50 mmHg, which was inside the normal range for this patient. Laboratory results were remarkable for disturbed liver enzymes with a cholestatic pattern: alanine amino-transferase (ALT) and aspartate amino-transferase (AST) levels were slightly elevated at 28 U/L and 61 U/L, respectively, while alkalic phosphatase levels (388 U/L) and γ-glutamyltransferase levels (291 U/L) were strongly elevated. Serum albumin levels were 24 g/L and the international normalized ratio was normal at 1.02. Other laboratory findings, including total bilirubin (0.32 mg/dL), direct bilirubin (0.26 mg/dL), total platelet count (176,000 platelets per microliter), and white blood cell count (8,400 cells per microliter), were within the normal range.

The patient had an extensive history of cardiac and renal disease. At the age of 11 years, congenital pulmonary valve stenosis was diagnosed for which the patient underwent valvulotomy. Subsequently, ill-tolerated atrial fibrillation and flutter developed for which flutter ablation was attempted unsuccessfully. Because the patient remained symptomatic, atrioventricular node ablation with pacemaker placement was performed. Oral anticoagulation was started with fenprocoumon because of a CHA2DS2-Vasc score of 3 (woman, heart failure and peripheral vascular disease). At the age of 55 years, overt right heart failure developed and the patient continued to struggle with ascites and peripheral oedemas requiring multiple hospital admission. In addition, she also developed a problem of recurrent transudative pleural effusions of unclear aetiology. Echocardiography at that time showed severe pulmonary and tricuspid valve regurgitation with a moderately dilated right ventricle and right atrium. The left ventricle was slightly dilated with moderate mitral valve regurgitation. Right heart catheterization demonstrated pulmonary arterial pressures of 74/30 mmHg with a central venous pressure of 28 mmHg. A pulmonary homograft was placed when the patient was 57 years of age and concomitant tricuspid valve annuloplasty was performed. Despite a lower central venous pressure of 13 mmHg, the patient still continued to be readmitted

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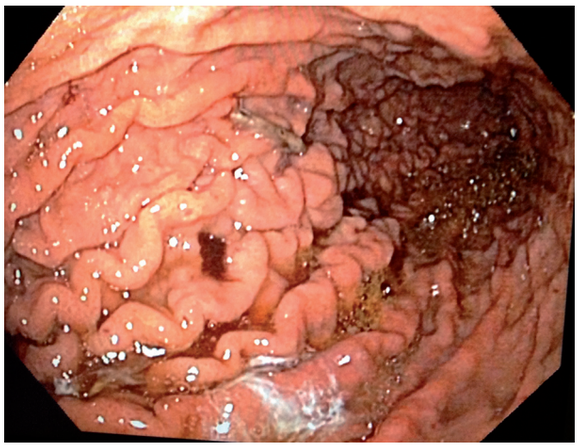

Fig. 1. — Gastroscopy imaging showing congestive mucosa with diffuse bleedings and few small erosions.

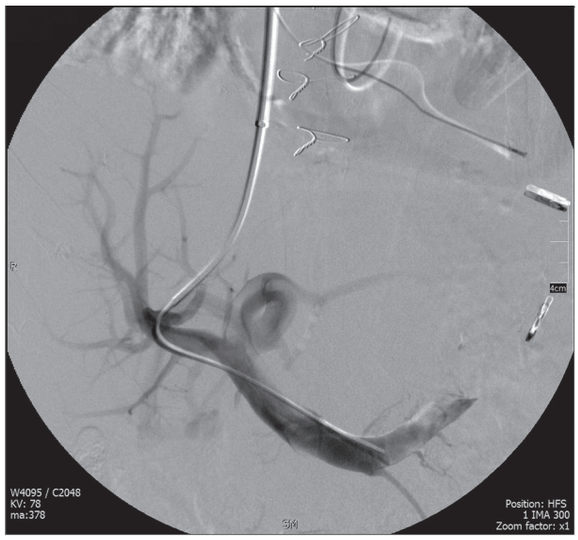

Fig. 2. — Radiographic image of transjugular intrahepatic portosystemic shunt placement.

multiple times for signs and symptoms of right heart failure. Persistent systemic venous congestion ultimately resulted in end-stage renal disease for which renal replacement therapy was started through intermittent haemodialysis. The case was complicated by recurrent severe gastro-intestinal bleedings, requiring frequent blood transfusions with up to 60 units of packed cells per year. Despite this in combination with intravenous iron substitution, haemoglobin levels varied between 5.3-8.8 g/dL. Because of these intractable bleeding problems, oral anticoagulation with fenprocoumon was stopped at the age of 59 years.

A gastroscopy showed an erosive gastropathy with a diffuse congestive, friable mucosa. (Fig. 1). As a first-line prevention of recurrent gastric bleedings in the setting of cirrhosis, propranolol was started at a dose of 5 mg orally twice daily (the patient did not tolerate higher dosing because of low blood pressure). In addition, endoscopic coagulation of a fundic angioma and clipping of a Dieulafoy lesion had been performed. These lesions were thought to be caused by hypertensive gastropathy. However, intractable bleedings remained. The hepatic venous pressure gradient was measured at 16 mmHg (27 mmHg-11 mmHg). Although, there was some reluctance because of fear for worsening right heart failure, a management decision was made to create a TIPS (Fig. 2). The procedure was uneventful. After TIPS placement, the hepatic venous pressure gradient decreased to 4 mmHg (14 mmHg-10 mmHg). Thus, although TIPS did lower the hepatic venous gradient considerably, portal pressure remained high because of the elevated central venous pressure.

After TIPS placement, the patient developed signs of hepatic encephalopathy and ammonia levels of 77 μmol/L were measured. Treatment with oral lactulose and enemas was started with little improvement. At that time, another episode of gastric bleeding recurred. Abdominal ultrasonography showed good patency of the TIPS with normal Doppler flow. Gastric bleeding from an antral ulcus was detected with endoscopy, despite gastric hyperaemia being visually decreased compared to the situation before TIPS. A biopsy was performed that showed an oedematous foveolar mucosa, suggesting mucosal ischemia. Bleeding was too diffuse for additional endoscopic ligation. No worsening liver function was found on laboratory results, with the international normalised ratio 1.01 at this time. Echocardiography showed no meaningful changes compared to the situation before TIPS placement. The right ventricle remained moderately dilated with poor longitudinal function and persistent severe tricuspid valve regurgitation. However, the general condition of the patient deteriorated and fever with bilateral pulmonary consolidations developed. Multi-organ failure emerged and at this point the family of the patient decided to withdraw care. The patient died eventually 43 days after the TIPS procedure.

Discussion

This case illustrates the complex management of a patient with right heart failure who developed cardiac cirrhosis with portal hypertension and its complications. It may serve as a reminder that the problem of right heart failure should be monitored closely and treated aggressively, even with consideration of orthotopic heart transplantation, before cirrhosis and/or end-stage renal disease develops. Our case is particularly insightful as TIPS was performed in the presence of ongoing right heart failure. No worsening right heart failure was observed immediately after the procedure, although portal pressures remained elevated because of persistently elevated right-sided cardiac filling pressures in the context of right heart failure.

Liver dysfunction secondary to heart failure results from two major pathophysiological mechanisms : [1] hypoperfusion caused by low cardiac output (i.e., forward failure) ; and [2] systemic congestion caused by elevated central venous pressure (i.e., backward failure). Liver dysfunction due to compromised arterial perfusion correlates with cardiac output, usually develops acute, and is associated with pronounced transaminase elevations (3). In contrast, the case described above is a clear example of liver dysfunction due to backward failure, ultimately leading to end-stage cirrhosis. This disease pattern primarily occurs in patients with right heart failure and elevated right-sided cardiac filling pressures. Tricuspid valve regurgitation is a major predisposing factor (4). It has been shown that liver dysfunction due to backward failure is characterized by a predominantly cholestatic pattern without elevation of transaminases (5). Both γ-glutamyltransferase and alkaline phosphatase levels have been shown to be independently associated with worse clinical outcome in this setting (6), and were markedly elevated in this case.

Portal hypertension frequently develops in cirrhosis and is defined as a hepatic venous pressure gradient ≥5 mmHg, but clinical severe complications such as variceal bleedings, ascites or encephalopathy, usually occur only when the gradient exceeds 10 mmHg (1,7). The causes of portal hypertension can be divided into pre-hepatic (i.e., portal vein thrombosis or extrinsic compression), intra-hepatic (i.e., liver diseases, mainly cirrhosis), or post-hepatic (i.e., right heart failure, pericarditis constrictive, veno-occusive disease or Budd-Chiari syndrome) causes. Portal hypertension is associated with severe complications of cirrhosis such as variceal bleedings, ascites, and encephalopathy (7). For prevention of recurrent variceal bleeding due to portal hypertension, the first-line treatment is a non-selective beta-blockers (e.g., propranolol or nadolol) are indicated as a first-line treatment. Non-selective beta-blockers are superior to cardio-selective beta1-blockers because they reduce splanchnic flow through their effects on beta^2-receptors (7). Indeed, the beta^2-receptor is ubiquitously present in the splanchnic circulation and leads to splanchnic vasodilation (8). In addition to beta-blocking agents, with nitrates often considered as an adjunctive therapy are often considered to reduce portal pressure (1). However, as our case illustrates, when blood pressure is low, pharmacological options to reduce recurrent gastric variceal bleedings are limited. In addition to optimal pharmacological therapy, endoscopic treatment can be used to treat and prevent bleeding. Endoscopic options consist of placement of elastic bands on variceal columns (variceal ligation), or to inject sclerosing agents (variceal sclerotherapy) or tissue adhesives (variceal obturation) (1). Oesophageal bleedings are usually treated with a combination of beta-blockers and endoscopic ligation, while gastric bleedings can be better controlled by sclerotherapy (9).In case of persistent bleeding problems despite optimal medical and endoscopic treatment, TIPS placement should be considered. TIPS is a connection through the liver parenchyma between the portal and hepatic veins and acts similar as a surgically placed porto-systemic shunt (8). It is effective in lowering the hepatic venous pressure gradient and reducing gastro-intestinal bleedings, by shunting a portion of the blood flow past the liver (9). This also explains the major complication of TIPS placement, which is worsening of hepatic encephalopathy. In a study encompassing 108 patients, Somberg et al. identified age >60 years, female gender, and low serum albumin levels as major risk factors for hepatic encephalopathy after TIPS (10). The patient described in this case report indeed had all of these risk factors. Yet, there was no other option to treat her intractable variceal gastric bleedings and additionally it was thought that the recurrent pleural effusions reflected hepatic hydrothorax, because they were out of proportion to the degree of left heart failure observed. Hepatic hydrothorax occurs in 5-10% of patients with end-stage liver disease and results in dyspnoea, hypoxia and infection (11). Hepatic hydrothorax is defined as a transudative pleural effusion, which predominantly occurs right-sided.

The exact underlying pathophysiological mechanism of hepatic hydrothorax is unknown, but it probably results from direct passage of fluid from the peritoneal to the pleural space via small diaphragmatic defects. When conservative management with dietary sodium restriction and diuretic therapy fails, TIPS placement should be considered because recurrent thoracentesis is associated with complications such as infection and bleeding. In this case with some degree of left ventricular dysfunction clearly present (presumably secondary to right ventricular pacing), the diagnosis of hepatic hydrothorax is however challenging.

Severe right heart failure with tricuspid valve regurgitation and elevated central venous pressure such as in this case is considered a relative contraindication for TIPS because systemic congestion can be transmitted to the portal circulation. This might result in insufficient lowering of the portal pressure, and when right heart failure worsens because of increased venous return, even worsening of portal hypertension may be observed (2). Because of concerns for worsening right heart failure, TIPS was delayed in this patient. When it was eventually performed as a salvage procedure, no worsening right heart failure was observed. Indeed, the central venous pressure before (11 mmHg) and after (10 mmHg) TIPS was similar and echocardiographic findings were virtually unchanged, while the hepatic venous pressure gradient decreased from 16mmHg to 4mmHg. Yet, portal hypertension did not disappear because central venous pressure was persistently elevated. Eventually, a catastrophical bleeding event occurred. In this case, the optimal time window to pursue aggressive treatment for right heart failure with consideration of orthotopic heart transplantation was missed. Orthotopic heart transplantation is the gold standard treatment for

refractory right heart failure, but obviously is limited to a minority of patients due to organ shortage. In this case, important contraindications such as end-stage renal disease and cirrhosis precluded consideration of this life-saving treatment. This may serve as a reminder that it is pivotal to carefully follow patients with right heart failure and consider orthotopic heart transplantation before cirrhosis occurs.

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