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CASE REPORT

Decompensated porto-pulmonary hypertension in a cirrhotic patient with thrombosis of portocaval shunt

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Abstract

We report a case of decompensated porto-pulmonary hypertension closely associated with the development of intra-portocaval shunt thrombosis. A woman with Laennec's cirrhosis was hospitalized because of severe dyspnea and edema. She underwent surgical portocaval anastomosis ten years ago. Imaging studies showed massive intra-shunt thrombosis, portal hypertension, ascites, pleuro-pericardial effusions and enlargement of right cardiac cavities. Cardiac catheterization allowed to rule out coronary and left-sided heart abnormalities and led to the diagnosis of pre-capillary pulmonary hypertension. Antithrombotic treatment with low molecular weight heparin was instituted. The management also included ACE inhibitors, spironolactone, low-salt diet and lactulose. The patient was discharged and three months later we observed the disappearance of edema, ascites and pleuro-pericardial effusions, a marked body weight reduction and improved dyspnea and liver function tests. A possible link between the development of intra-shunt thrombosis and clinical decompensation in our patient was hypothesized. In fact, it has been demonstrated that the increased portal pressure, caused by occlusion of portosystemic shunt, reduces renal plasma flow and increases systemic endothelin-1 concentration. In our patient the disappearance of edematous state and improved dyspnea observed after recanalization of the shunt strongly support this hypothesis.

Key words: Porto-pulmonary hypertension; Porto-caval shunt; Thrombosis

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INTRODUCTION

Porto-pulmonary hypertension (PPHT) refers to the development of pulmonary arterial hypertension in the setting of portal hypertension with or without chronic liver disease and is defined by a mean pulmonary artery pressure > 25 mmHg in the presence of normal pulmo-capillary wedge pressure (< 15 mmHg)[1,2]. The pathogenesis of PPHT is under investigation, although histopathologic features are similar to those found in primary pulmonary hypertension[3]. A current study supports the hypothesis that pulmonary vasculature may be exposed to either cytokines or excess circulating vasoconstrictors, such as endothelin-1 (ET-1) produced by the diseased liver[4]. Although the vast majority of patients with PPHT are asymptomatic, dyspnea is the most frequent presenting symptom[5].

CASE REPORT

A 39-year-old obese woman with Laennec's cirrhosis (Child-Pugh B) was admitted to our hospital in January 2003 because of the recent onset of dyspnea (NYHA IV), dependent edema and abdominal pain. In 1991, she underwent surgical side to side portocaval shunt for refractory ascites and since then she has never complained of dyspnea or edema. Physical examination showed platypnea, arterial blood gas analysis showed hypoxaemia (pO2: 63 mmHg) with orthodeoxia (pO2: 49 mmHg). On admission, liver function tests were as follows: total bilirubin: 84.5 μmol/L, AST: 1.14 μkat/L, ALT: 0.55 μkat/L, GGT: 87 U/L, alkaline phosphatase: 4.2 nkat/L, serum albumin: 28 g/L, ammonia (as NH3): 88 μmol/L, prothrombin time 60%. Hepatic ultrasonography with Doppler imaging showed massive intra-shunt thrombosis, portal hypertension (18 mmHg) and mild ascites. No gastroesophageal varices were found by digestive endoscopy. D-dimer level was 1.4 mg/L and antithrombin activity was 43%. The presence of both peripheral venous thrombosis and recurrent microembolism was ruled out by Doppler ultrasonography and ventilation-perfusion lung scan. Computerized tomography confirmed recent intra-shunt thrombosis and showed also pleuro-pericardial effusions and ascites. Markers of autoimmunity were negative.

Right atrial and ventricular enlargement with severe

Table 1 Hemodinamic parameters during cardiac catheterization

Aortic pressure (mmHg)	103-76 (mean 85)
Right atrial pressure (mmHg)	11 (mean)
Right ventricular pressure (mmHg)	70-18 (mean 35)
Pulmonary arterial pressure (mmHg)	70-30 (mean 43)
Pulmonary capillary wedge pressure (mmHg)	10
Cardiac output (l/min)	5.43
Cardiac index (l/min per m^2)	2.83
Total pulmonary resistance (dyn•s/cm^5)	632
Pulmonary vascular resistance (dyn•s/cm^5)	488
Systemic vascular resistance (dyn•s/cm^5)	1088

tricuspidal regurgitation was also detected by echocardiography. Systolic pulmonary arterial pressure, as measured by Doppler analysis, was 50 mmHg. Cardiac catheterization showed no coronary and left-sided heart abnormalities. Hemodynamic parameters (Table 1) led to the diagnosis of moderate precapillary pulmonary hypertension. Antithrombotic treatment with low molecular weight heparin was instituted. The treatment also included ACE inhibitors, spironolactone, insulin, low-salt diet (2 gr pd) and lactulose. This management decreased the retention of sodium and water in the kidney from 12 mEq/d on admission to 186 mEq/d after 3 d of treatment, and improved edema, as demonstrated by natriuresis. The patient was discharged one month later with the above-mentioned prescription and, after three months of follow-up, physical examination showed the disappearance of edema, marked weight reduction (20 kg) and improved dyspnea (NYHA II). Liver function tests were as follows: total bilirubin: 15.3 μmol/L, AST: 0.56 μkat/L, ALT: 0.47 μkat/L, GGT: 36 U/L, alkaline phosphatase: 1.85 nkat/L, serum albumin: 30 g/L, ammonia (as NH3): 76 μmol/L, prothrombin time 67%. Abdominal ultrasonography showed recanalization of porto-caval shunt, disappearance of ascites and right pleural effusions. No pericardial effusions were found by ecocardiography. Antithrombin activity was 56%, D-dimer 0.2 mg/L. Orthotopic liver transplantation was excluded because of the high intra-operative mortality in patients with portopulmonary hypertension and the conflicting results reported in this condition[6]. Therefore, the patient was referred to a specialized center in order to start an appropriate vasodilatory therapy, which appears to be the only feasible treatment.

DISCUSSION

This case report suggests a possible link between the development of intra-shunt thrombosis and clinical decompensation in our patient. In fact, a recent study showed that increased portal pressure caused by occlusion of porto-systemic shunt reduces renal plasma flow, leading to renal sodium and water retention, and increases systemic ET-1 concentration[4]. In our patient, disappearance of edema and improved dyspnea, observed after recanalization of the shunt, strongly support this hypothesis, although further experience with similar cases is needed.

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