Fatal pulmonary hypertension after distal splenorenal shunt in schistosomal portal hypertension

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INTRODUCTION

Mansonic schistosomiasis is the main cause of portal hypertension in Brazil. Hepatosplenic (HS) form is manifested by hepatomegaly mainly on the left hepatic lobe associated with large splenomegaly and bleeding due to esophageal varices with high mortality rates[1,2].

Pulmonary hypertension in the HS form of mansonic schistosomiasis is described in association with both the acute and chronic forms of the disease, with a prevalence of 5% and may be a serious complication in the evolution of the disease. It can also be the triggering factor for serious complications associated with any form of surgical approach[3,4].

Surgical treatment is indicated for patients with a history of bleeding due to esophageal varix rupture based on world previous experience with endoscopic treatment results[5]. At present, techniques vary in azigo-portal disconnection and splenectomy (APDS) and distal splenorenal shunt (DSRS) for surgical treatment of such patients. All patients submitted to DSRA were carefully evaluated pre-operatively with eletrocardiography, chest X-ray and transthoracic echocardiography to rule out pulmonary hypertension. We report two fatal cases of pulmonary hypertension arising after DSRS.

Case Reports

Case 1 A 33-year-old man was admitted to the Liver and Portal Hypertension Surgical Unit, Hospital das Clínicas, University of São Paulo Medical School for surgical treatment of portal hypertension due to hepatosplenic mansonic schistosomiasis after previous episode of bleeding esophageal varices. Ultrasound scan of the abdomen revealed discrete hepatomegaly, mainly on the left hepatic lobe associated with splenomegaly. Esophagogastroduodenoscopy revealed four large variceal vessels. The results found in routine laboratory tests were alanine amino transferase (ALT) of 29 U/L (normal: 10-30), aspartate amino transferase (AST) of 20 U/L (normal: 10-30), bilirubin of 0.8 mg/dL (normal: 0-2.1), gamma-glutamyl transferase of 86 U/L (normal: 7-32), and alkaline phosphatase of 241 U/L (normal: 32-104). The electrocardiogram, chest X-ray and echocardiography were within normal limits. The patient was submitted to DSRS. On the 4th postoperative day he developed progressive dyspnea and dyed 3 h later. The necropsy showed severe pulmonary hypertension, right ventricular dilatation and secondary myocardial infarction. The shunt was pervious.

Case 2 A 25-year-old woman was admitted to the Liver and Portal Hypertension Surgical Unit, Hospital das Clínicas, University of São Paulo Medical School for surgical treatment of portal hypertension due to hepatosplenic mansonic schistosomiasis after an episode of bleeding esophageal varices. Ultrasound scan of the abdomen revealed discrete hepatomegaly, mainly on the left hepatic lobe associated with splenomegaly. Esophagogastroduodenoscopy revealed two large variceal vessels. The results found in routine laboratory tests were alanine amino transferase (ALT) of 25 U/L (normal 7-45), aspartate amino transferase (AST) of 38 U/L (normal 7-45), bilirubin of 1.0 mg/dL (normal 0.2-1.0), gamma-glutamyl transferase of 31 U/L (normal 7-32), and alkaline phosphatase of 96 U/L (normal 32-104). The electrocardiogram, chest X-ray and echocardiography were within normal limits. The patient was submitted to DSRA. One the 7th postoperative day he developed progressive dyspnea and cardiogenic shock. A pulmonary artery catheter was inserted and showed high pulmonary mean artery pressure (64 mmHg) and a reduced cardiac index (CI = 1.2 L/min/m²). The patient died before the shunt could be occluded. The necropsy showed severe pulmonary hypertension, right ventricular dilatation and secondary myocardial infarction. The shunt was pervious.

DISCUSSION

The treatment of portal hypertension in hepatosplenic schistosomiasis is very different in cirrhotic patients mostly because hepatic function is well preserved in Manson’s disease. The surgical treatment has been considered the best alternative for schistosomatic patients with a history of bleeding from esophageal varix rupture because these patients had normal liver function and the only severe complication of the disease was digestive bleeding[2-6].

The two modalities of elective surgical treatment are selective shunt surgery (Warren procedure) or an esophagogastric devascularization procedure with splenectomy (EGDS). Shunt surgeries have been found to be effective for bleeding control, but were associated with postoperative encephalopathy and higher operative mortality rates when compared to devascularization procedures[2,6].

Although recognizable as a complication of shunt procedures, acute and fatal pulmonary hypertension have never been previously reported since Warren procedure for treatment of portal hypertension in mansonic schistosomiasis. Some groups in Brazil performed thousands of shunt surgeries for schistosomal portal hypertension, but have never described this important
Our group routinely performed a transthoracic echocardiography in order to evaluate pulmonary hypertension. The two patients had a normal pre-operative evaluation and were submitted to DSRS. The necropsy revealed typical findings of acute pulmonary hypertension, suggesting that sub-clinical elevated pulmonary artery pressure be present before surgery. The hyperflow determined by shunt surgery aggravated a previously unsuspected pulmonary hypertension, leading to death. This observation suggests that shunt surgeries should be employed very cautiously in young patients with portal hypertension due to hepatosplenic mansonic schistosomiasis and should be performed only after measurement of pulmonary artery pressure, if indicated.

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