Emergent pulmonary thromboendarterectomy with percutaneous cardiopulmonary support system for chronic thromboembolic pulmonary hypertension

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Received 21 January 2003; received in revised form 3 June 2003; accepted 12 June 2003

Abstract

Pulmonary thromboendarterectomy offers a comparatively low surgical mortality rate with appropriate patient selection. However, the operative mortality in patients with high pulmonary vascular resistance or severe pulmonary hypertension and subsequent right ventricular failure is poor. We report an unusual case that survived an emergent pulmonary thromboendarterectomy for chronic thromboembolic pulmonary hypertension with right ventricular failure.

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Keywords: Chronic thromboembolic pulmonary hypertension; Pulmonary thromboendarterectomy; Right ventricular failure; Percutaneous cardiopulmonary support system

1. Introduction

Chronic thromboembolic pulmonary hypertension is a progressive and life-threatening disease that is resistant to medical treatment; its surgical cure can be provided only by pulmonary thromboendarterectomy or lung transplantation. Although the mortality rate of pulmonary thromboendarterectomy in the early period has been reported to be high [1], recent progress in technique as well as strategy has improved the outcome. Indeed, Jamieson reported a mortality of 6.4% [2]. However, patients with high pulmonary vascular resistance (PVR) or high pulmonary artery pressure (PAP) are at significantly increased operative risk [3]. We report a successful surgical treatment in a patient who underwent emergent pulmonary thromboendarterectomy for chronic thromboembolic pulmonary hypertension with right ventricular failure.

2. Case report

A 31-year-old male visited another hospital with a complaint of dyspnea. The computed tomographic scan showed thrombi in the pulmonary artery and the right femoral vein. The pulmonary angiogram demonstrated irregular and steeply narrowing arteries, while a lack of the distal branches suggested the possibility of distal disease. The mean PAP was 44 mmHg and the PVR was 287 dyne s cm$^{-5}$. The patient was diagnosed as having chronic thromboembolic pulmonary hypertension with antithrombin III deficiency. An inferior vena cava filter was placed and he was treated with anticoagulants. Thereafter, he gained 15 kg in 17 months and developed serious edema in the lower extremities, along with palpitation and orthopnea. He was again admitted, with a condition of New York Heart Association functional class IV. The chest X-ray showed remarkable cardiomegaly (Fig. 1), and echocardiography revealed right heart congestion with incompetent tricuspid valve. He was referred to us and treated with urokinase, amrinone, and digoxin. Although his symptoms improved on that occasion, ventricular arrhythmia appeared, following which dyspnea worsened, and he was taken to the ICU. He developed tachypnea and tachycardia with bigeminy, and the chest X-ray showed bilateral pleural effusion. Measured mean PAP was 62 mmHg and, assuming the pulmonary capillary wedge pressure (PCWP) to be 10 mmHg, the PVR was calculated...
as 2078 dyne s cm$^{-5}$. In consideration of limitation of medical treatment, an emergent pulmonary thromboendarterectomy was performed using cardiopulmonary bypass (CPB) under profound hypothermia with intermittent circulatory arrest. Tricuspid annuloplasty was performed at the same time in order to curtail postoperative right heart failure. Because the PAP overtook the systemic pressure after discontinuing CPB, the patient was returned to the ICU with a percutaneous cardiopulmonary support system (PCPS). The PVR was 425 dyne s cm$^{-5}$ assuming the PCWP to be 10 mmHg the following day. Eventually, PAP was decreased with inhalation of nitric oxide gas, and PCPS was discontinued on the second postoperative day. Pneumonia prolonged mechanical ventilation by as much as 38 days. The duration of the ICU stay was 45 days and, after discharge on the 89th postoperative day, he was treated with home oxygen therapy for 3 months. The chest X-ray showed improvement of cardiomegaly (Fig. 2) and the echocardiography demonstrated no tricuspid incompetence. PAP and PVR recovered to normal values without oxygen inhalation. Now the patient is in New York Heart Association functional class I without oxygen.

3. Comment

Pulmonary thromboendarterectomy is thought to be the primary treatment for chronic thromboembolic pulmonary hypertension and has recently offered a lower surgical mortality [2]. Better recognition by physicians and progress in diagnostic imaging have led to earlier referral for surgery while establishment of the principles of the operation have brought better results. However, high PVR and high PAP predict operative mortality. Hartz and his colleagues reported that operative mortality in the patients with a PVR of greater than 1100 dyne s cm$^{-5}$ was 41%, and in those with a mean PAP of greater than 50 mmHg it was 37% [3]. In our case, with an estimated PVR of 2078 dyne s cm$^{-5}$ and a measured mean PAP of 62 mmHg, the prognosis was thought to be very poor. Seventeen months previously these values had been 287 dyne s cm$^{-5}$ and 44 mmHg, under which terms they should have been reevaluated and the patient should have undergone pulmonary thromboendarterectomy earlier, even though there was a possibility of distal disease.

With regard to postoperative circulatory support, we failed to find any literature reporting patients who survived [3–5] except for Jamieson’s in which postoperative support to manage reperfusion injury was mentioned [6]. Furthermore, the operative mortality of emergent pulmonary thromboendarterectomy is very high [7]. We believe this is the first article that reports a case that survived an emergent pulmonary thromboendarterectomy with postoperative PCPS.

Why did our case survive? Engagement of the distal disease cannot explain the presence of normal PAP without oxygen after the operation. Although reperfusion injury was suggested by postoperative airway bleeding, reperfusion injury and preoperative right ventricular failure cannot account for a PAP so elevated that it overtook the systemic pressure. Nitric oxide gas inhalation was effective at reducing the PAP, and we believe there was medial disease of pulmonary artery. Prolonged pulmonary hypertension might have led to medial thickening, while intraoperative CPB, hypoxia during the periods of circulatory arrest, and hypothermia should have caused severe medial
constriction. Removal of proximal disease and the months of oxygen therapy might have improved medial thickening and eventually have reduced the PAP to normal. The combination of distal disease, medial disease, reperfusion injury, and preoperative right ventricular failure were responsible for extreme pulmonary hypertension, and their resolution for its subsequent improvement. Usually, tricuspid valve repair is not performed together with pulmonary thromboendarterectomy. However, in this case, the diameter of the annulus exceeded 7 cm and the regurgitation was so severe that we chose to repair it in order to minimize the likelihood or severity of postoperative right heart failure.

In conclusion, an emergent pulmonary thromboendarterectomy was able to alleviate chronic thromboembolic pulmonary hypertension with high PVR, high PAP, and subsequent right ventricular failure. PCPS was useful for overcoming severe temporary pulmonary hypertension. An accurate evaluation of the operative indication followed by timely and precise surgery is of utmost importance.

References