Successful staged operation for acute aortic dissection and chronic thromboembolic pulmonary hypertension

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INTRODUCTION

Pulmonary endarterectomy (PEA) is the potentially curative treatment for chronic thromboembolic pulmonary hypertension (CTEPH), but associated with relatively high in-hospital mortality. In the present report, we describe a patient with CTEPH who developed acute type A aortic dissection, which required emergency surgery for life-saving. We performed a staged procedure for acute type A aortic dissection and CTEPH, and instituted venoarterial-extracorporeal membrane oxygenation (VA-ECMO) after aortic surgery as a bridge to subsequent PEA. To our knowledge, this is the first report on successful treatment strategy for a case requiring complicated aortic surgery and PEA.

CASE REPORT

A 68-year-old woman with CTEPH was admitted to our hospital for a sudden onset of severe anterior chest pain. She underwent work-up examination for CTEPH 14 years ago and had been medically treated since then. At the time of diagnosis, a right heart catheterization showed a pulmonary artery pressure of 66/25 (mean 41) mmHg and a pulmonary vascular resistance of 527 dynes s cm\(^{-5}\). Despite no coagulation abnormality, she worsened pulmonary embolism while she stopped taking warfarin for teeth extraction and was treated with thrombolytic therapy 10 years ago. A chest computed tomography (CT) scan on admission showed acute type A aortic dissection, and the dilated bilateral pulmonary arteries with thrombus in the right lower lobe artery (Fig. 1A). A right heart catheterization with a Swan-Ganz catheter showed a pulmonary artery pressure of 77/25 (mean 49) mmHg and a pulmonary vascular resistance of 542 dynes s cm\(^{-5}\). We discussed with the patient and family the dismal prognosis without surgical treatment, which, however, carries very high risks, and decided to perform the operation. We planned a two-stage procedure consisting of emergent aortic replacement and subsequent PEA after the patient’s condition is stabilized with the aid of VA-ECMO. We first performed repair for acute type A aortic dissection under antegrade selective cerebral perfusion with moderate hypothermic circulatory arrest and instituted VA-ECMO. We excised the primary tear in the lesser curvature of the aortic arch and performed hemiarch replacement with reconstruction of the innominate artery. A prosthetic graft anastomosed to the right axillary artery was used for antegrade cerebral perfusion, and also used as an antegrade flow during VA-ECMO. In the immediate postoperative period, the patient developed cardiac tamponade due to persistent mediastinal bleeding, and required the evacuation of clots and haemostasis. Administration of heparin could not be initiated until postoperative day 4. After stabilization of the condition with ECMO support, we attempted weaning from ECMO on postoperative day 5. However, pulmonary artery pressure exceeded systemic artery pressure with decreasing ECMO flow: pulmonary artery pressure 71/39 mmHg and systemic artery pressure 58/19 mmHg. Pulmonary angiography showed de novo thrombus formation in the bilateral pulmonary arteries (Fig. 1B). On postoperative day 8, she underwent PEA under periods of deep hypothermic circulatory arrest and was successfully.
separated from cardiopulmonary bypass. The surgical specimens showed Jamieson classification type II organized thrombus and fresh thrombus (Fig. 2). She was extubated on postoperative day 3 and applied non-invasive positive pressure ventilation. Subsequent recovery was complicated with left phrenic nerve paralysis, which was treated by diaphragmatic plication, and pneumonia, but she gradually recovered from the respiratory complications with administration of antibiotics and prolonged rehabilitation. A right heart catheterization at 3 months after PEA showed improved pulmonary haemodynamics: a pulmonary artery pressure of 55/20 (mean 35) mmHg and a pulmonary vascular resistance of 465 dynes s cm\(^{-5}\). Four months after PEA, she was discharged from another rehabilitation hospital. One year after the surgery, her respiratory symptoms were significantly relieved with improvement of tricuspid pressure gradient from preoperative 74 to 46 mmHg, and CT scan showed no dilatation of dissected arteries.

DISCUSSION

Acute type A aortic dissection requires emergent surgery for life-saving, otherwise most of the patients die within a few days. Replacement of the ascending aorta with or without the aortic arch is a standard procedure, and antegrade selective cerebral perfusion is used as an adjunct to mild-to-moderate hypothermic circulatory arrest. PEA is performed under periods of deep hypothermic circulatory arrest, and other simple cardiac procedures can be performed simultaneously, usually during the warming period, with acceptable mortality and morbidity [1]. However, surgery for acute aortic dissection is a complicated procedure associated with high mortality of ~20% [2]. Therefore, a simultaneous procedure for acute type A dissection and CTEPH may have prohibitively high surgical risks. Our staged procedure would have been the only possible selection for the complicated situation accompanying the two serious pathologies.

We performed PEA a week after the hemiarch replacement, but optimal timing for subsequent PEA after the aortic surgery was uncertain. There were serious concerns about the effects of deep hypothermic circulatory arrest during PEA on the patient who underwent hemiarch replacement with antegrade cerebral perfusion and systemic circulatory arrest. We believed that stable organ function was required for subsequent PEA. On the contrary, delayed operation may increase the chance to accumulate clots in the pulmonary artery, which may complicate the procedure during PEA. Therefore, our initial plan was to perform the second operation as soon as there was recovery from the deleterious effects of the first operation. However, we had to wait for the second operation because of bleeding complications and renal dysfunction.

Usefulness of ECMO as a bridge to PEA in a case of preoperative severe hemodynamic compromise has been reported [3]. Although preoperative pulmonary hypertension was not so severe in the present case, we used ECMO as a bridge to subsequent PEA, because we considered that right heart failure due to pulmonary hypertension was inevitable after cardiac arrest during hemiarch replacement. An attempt to wean from ECMO after haemodynamic stability was achieved failed due to thrombus formation in the pulmonary arteries. Anticoagulation with heparin was not administered because there was persistent mediastinal bleeding, and blood flow to the right heart and lungs was diminished due to cardiac tamponade. These two factors may have been associated with thrombus formation. In addition, the history of thrombus formation in the pulmonary artery during cessation of warfarin in spite of no diagnosis of coagulation abnormality may suggest undiagnosed thrombotic tendency in the present case.

Although the patient needed ECMO for haemodynamic stability, ECMO after acute aortic dissection can cause serious complications, including malperfusion of the major organs due to deterioration of the dissection and compression of the true

Figure 1: (A) Enhanced computed tomography scan showing aortic dissection and dilated pulmonary trunk. (B) Pulmonary angiography showing central thrombus.

Figure 2: Pulmonary endarterectomy specimens.
lumen. However, antegrade flow via the right axillary artery has the potential advantage of preventing the morbidities over retrograde flow via the femoral artery. We used the right axillary artery as arterial cannulation for antegrade flow during VA-ECMO and excised the primary tear that existed in the aortic arch at the time of hemiarch replacement.

Successful separation from cardiopulmonary bypass after PEA indicated effectiveness of this procedure even for patients with severe haemodynamic compromise. A minority of the patients requires ECMO after PEA mainly due to persistent pulmonary hypertension and/or reperfusion lung oedema [4, 5]. The outcomes of ECMO were disappointing with quite high mortality of 43 and 70%, although these patients would probably die without ECMO [4, 5]. For the sake of the prevention of persistent pulmonary hypertension and/or reperfusion lung oedema, we attempted to control cardiac output to avoid pulmonary overflow at the time of weaning from cardiopulmonary bypass and applied non-invasive positive pressure ventilation after extubation.

In conclusion, our planned staged procedure was effective for a CTEPH patient requiring complex aortic surgery. VA-ECMO was useful as a bridge to subsequent PEA.

Conflict of interest: none declared.

REFERENCES