Case report

Extracorporeal membrane oxygenation as a bridge to lung transplantation in a patient with persistent severe porto-pulmonary arterial hypertension following liver transplantation

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Abstract

Idiopathic pulmonary artery hypertension (IPAH) is a progressive disease with a dismal prognosis and lung transplantation is often the only option for patients, who do not respond to pharmacological therapy. We report the use of an extracorporeal membrane oxygenation (ECMO) system in a 49-year-old woman with primary pulmonary hypertension, previously liver transplanted. The patient, listed for lung transplantation, developed respiratory and circulatory failure despite maximal pharmacological therapy and was successfully bridged to emergent bilateral lung transplantation with veno-arterial ECMO. Emergent veno-arterial ECMO was able to rescue the patient and bridge her to bilateral lung transplantation and should therefore be an option for patients with PAH and circulatory collapse.

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1. Introduction

Idiopathic pulmonary artery hypertension (IPAH) is a progressive disease with a dismal prognosis due to development of severe right-ventricular failure [1]. Lung transplantation is the only option for patients non-responsive to pharmacological therapy. Veno-venous extracorporeal membrane oxygenation (ECMO), substituting pulmonary gas exchange, can be used in patients with lung dysfunction but in patients with PAH, both circulatory and respiratory support may be necessary and veno-arterial ECMO is an option. Veno-arterial ECMO is also used in some centres to treat patients in cardiogenic shock [2,3]. We report a successful case of emergency use of veno-arterial ECMO in a patient with IPAH and circulatory collapse.

2. Case report

In January 2005, a 46-year-old woman with a medical history of ulcerous colitis, presented with fatigue and dyspnoea for 8 months. Examination revealed jaundice, ascites and pulmonary arterial hypertension (PAH), initially interpreted as secondary to liver disease and portal hypertension. Primary sclerosing cholangitis was diagnosed. Transthoracic echocardiography (TEE) revealed pulmonary artery pressures of 75 mmHg and normal right- and left-ventricular function. The patient was accepted for liver transplantation and treated with sildenafil and iloprost-tro-metanol (Ventavis®) during the waiting period. In May 2007, the patient underwent a successful liver transplantation but due to persistent PAH postoperatively, the treatment was changed to continuous infusion with prostacyclin (epoprostenol-Flolan®). The patient deteriorated in the following months, becoming oxygen dependent, and N-terminal prohormone brain natriuretic peptide (NTproBNP) increased from 200 to 20 000 indicating failing right-ventricular function, and was therefore evaluated and accepted for lung transplantation. The following data were registered: mean pulmonary artery 68 mmHg, cardiac output 3.5 l, mixed venous oxygen saturation (SvO₂) 57%, pulmonary artery wedge pressure 10 mmHg and maximal oxygen uptake $10.8 \text{ kg m}^{-1} \text{ min}^{-1}$. In the following week, the patient deteriorated rapidly with end-organ dysfunction and, despite maximal inotropic support, had cardiogenic shock. The decision was made to support the patient with veno-arterial ECMO. In local anaesthesia, using the Seldinger technique, the left subclavian artery was punctured percutaneously and a Biomedicus cannula 21 F was inserted. The right femoral artery was percutaneously cannulated with an Arrow® cannula 6 F distally to secure the peripheral circulation, and a Fem-Flex® (Baxter) 18 F was placed proximally. The
ECMO was adjusted to a flow of 4.2 l min\(^{-1}\) aiming to allow the heart to eject and the haemodynamic situation was stabilised instantly. The anticoagulation protocol in our institution is based on heparin infusion, and activated partial thromboplastin time (APTT) target level of 50–60 s depending on the flow and bleeding problems. Postoperatively, end-organ function recovered and the patient was awake and extubated. Twelve days later, a suitable donor became available and she underwent bilateral lung transplantation on ECMO via sternotomy. The transplantation was complicated because of oversized donor lungs resulting in a lower lobectomy on the right side and a partial resection of the left lower lobe. The ECMO system was terminated immediately after completion of the transplant surgery and right-ventricular function sufficiency, only supported with a moderate dose of milrinone (Corotrop\(^{®}\)). Postoperative haemostatic problems with a profound platelet dysfunction verified with multiplate analysis were repeatedly treated with platelets, packed red blood cells, plasma, fibrinogen and recombinant factor VII (NovoSeven\(^{®}\)) and surgical re-intervention. One week later, the patient was re-operated due to haemothorax and air leakage in the right lung at the lower lobe resection site. The broncho-pleural fistula was successfully closed with a vascularised pericardial patch. Henceforth, the clinical course was uneventful and the patient was discharged from the hospital, in good condition, 5 weeks after the lung transplantation.

At 2 years’ follow-up, the patient is doing fine but still proves to have reduced lung capacity, probably because of a dysfunctional right diaphragm. The cardiac function is normal.

3. Comments

The report above describes a very complicated clinical course in a patient with an uncommon variant of PAH where the life-saving support with veno-arterial ECMO resulted in complete recovery of end-organ function, and provided a bridge for the patient to bilateral lung transplantation. A emergent heart–lung transplantation was not considered as no organs would become available for both heart- and lung transplantations.

In our clinic, it has earlier been contraindicated to use ECMO in patients with PAH, mainly because of anticipated bleeding problems at the time of transplantation and also due to the shortage of donors. It can still be argued that donated lungs should be used in patients with a better chance to survive. On the other hand, patients with PAH are often young patients, who, in our experience, often expire after acceptance for transplantation during the waiting time.

Because veno-arterial ECMO completely bypasses and unloads both the lungs and the heart, it is a radical treatment of the ultimate problem with PAH, severe right-heart failure. However, with total bypass and minimal flow through the central circulatory bed, thrombus formation in the lungs or heart may occur and, therefore, it is important to adjust the flow to allow the heart to eject [4].

Veno-venous ECMO or right-ventricular assist device (RVAD) is an often insufficient treatment and does not solve the problem in this patient group [4].

4. Conclusion

In conclusion, a previously liver-transplanted woman with advanced IPAH developed circulatory collapse during the waiting time for donor lungs. Emergent veno-arterial ECMO was able to rescue the patient and bridge her to bilateral lung transplantation.

References