Combination of extracorporeal membrane oxygenation (ECMO) and pulmonary lavage in a patient with pulmonary alveolar proteinosis

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Abstract

We describe a rare case of pulmonary alveolar proteinosis in a young woman with dyspnea and progressive hypoxaemia due to the alveolar deposition of insoluble, surfactant-like material. Routine treatment includes whole-lung-lavage (WLL) using double-lumen-tubes for selective lavage of each lung. We performed three whole-lung-lavages and used veno–venous extracorporeal membrane oxygenation (v–vECMO) to support oxygenation during these procedures.

Keywords: Pulmonary alveolar proteinosis; Whole-lung-lavage; Extracorporeal membrane oxygenation

1. Introduction

Pulmonary alveolar proteinosis (PAP) is a rare disease, causing dyspnea and progressive hypoxaemia due to the deposition of insoluble, surfactant-like material in the alveoli of the lungs. Whole-lung-lavage (WLL) with isotonic saline seems to be the only consistently successful treatment best using double-lumen-tubes for selective lavage of each lung [1]. However, it may induce a temporary hypoxaemia with fatal consequences. To support oxygenation during lavage, we used veno–venous ECMO (v–vECMO) in a young woman for 6 days and report this case since, to our knowledge, only few authors used the same approach [2].

2. Patient, diagnosis and treatment

A 34-year-old woman (BMH-N29674), was transferred to our hospital to explain a progressively increasing dyspnea during the last 2 months. She smoked 20 cigarettes a day for 12 years. During the last 6 months, she underwent several upper airway infections. There were no environmental risk factors nor known allergies. At admission, she presented in a reduced general condition (weight 48 kg, height 158 cm) with oedemas of ankles and knees which had developed during the previous week. On chest auscultation, there were vesicular breathing sounds without rales. Blood-pressure was 110/80 mmHg, heart rate 100 bpm. Chest X-ray showed a normal heart size, but the lungs stroke with patchy confluent infiltrates and increased interstitial markings.

The immuno-serological examination was negative for both, allergic alveolitis and lung fibrosis. The cytology of bronchial lavage showed masses of acellular oval corpuscles of strong PAS-positive material and cell detritus. The lung biopsy showed a preserved alveolar structure filled with homogeneous eosinophilous material, partially granular and including few lamellar bodies. The histological examination of the bronchial lavage confirmed the diagnosis of PAP.

Under treatment with corticoids and antibiotics, neither clinical nor radiological findings improved. After the first bronchial lavage, the patient developed both, a respiratory insufficiency documented by increasing pulmonary infiltrates of the chest X-ray, and a low-cardiac-output-syndrome. Oxygenation deteriorated (PaO2 of 54 mmHg...
under oxygen 12 l/min) and the patient was intubated. Two hours later, the PaO₂ was 118 mmHg with a FiO₂ of 1.0.

To assure adequate oxygen supply during lavage, we implanted a v–vECMO. The oxygenation and cardiovascular situation improved rapidly. WLL was performed starting the day after cardiopulmonary stabilization (Figs. 1 and 2).

During the first WLL, we used saline as recommended [1–3]. We encountered severe cardiac arrhythmias that could not be explained by the moderate drop of oxygen tension. However, we found a considerable shift of potassium, a decrease of its plasma concentration from 5.8 to 4.0 mmol/l. The following WLL, we used Ringer’s solution, and still induced arrhythmias, but did not observe a change in the potassium level yet the core temperature of the patient dropped to 35.6°C. During the third WLL, and during further bronchial lavages, we prewarmed Ringer’s solution up to 37°C using a blood/infusion warmer (warmflow®) and faced no arrhythmias.

After three WLLs, the patient was easily weaned from ECMO, but developed septic temperature. *Candida, Pseudomonas* and oxacillin-resistant *Staphylococcus aureus* were found in blood- and tracheal-fluid cultures. A tracheotomy was performed 5 days after ECMO-explantation. The situation of the patient improved and initial signs of right cardiac insufficiency disappeared. Two weeks later, the patient was transferred to the ward. Transthoracic echocardiography was performed before discharge and revealed diffuse myocardial impairment with left ventricular dilatation. The myocardial biopsy showed unspecific myocardial injury concordant with a cardiomyopathy probably due to the infection. The patient was discharged home 7 weeks after the first bronchial lavage without any signs of adult respiratory distress syndrome (ARDS), nor had she disturbances of lung compliance.

The patient was readmitted twice with intervals of 6 and 8 weeks. She presented again with dyspnea, but uncomplicated WLL relieved her from shortness of breath. After short hospital stays, she was discharged home again.

3. Discussion

Hypoxaemia and infection are the most important factors influencing the outcome in pulmonary alveolar proteinosis [3–5]. WLL seems to be the only consistently successful treatment removing deposited material, as well as pathogenic organisms, from bronchial and alveolar spaces [5]. The problem is that this treatment may induce temporary hypoxaemia with fatal consequences. The risk of hypoxaemia is greatest during the draining of the lavage fluid from the lungs, allowing shunting of pulmonary blood flow through the non-ventilated lung. Using only WLL, a mortality rate of about 30% is reported [1,3,6].

The patient was in a disastrous situation after the first diagnostic and therapeutic bronchial lavage. Oxygenation had deteriorated rapidly, she fainted and presented a low-cardiac-output syndrome. Conventional ventilation did not result in substantially better oxygenation, hence, extracorporeal membrane oxygenation was indicated.

Two modes of ECMO are known: the arterio-venous and the veno-venous mode. The arterio-venous mode has been implemented several times in patients with PAP, mostly only for WLL with rapid weaning after the procedure [2,3,6–10]. We chose for the veno-venous mode for two reasons: first, the oxygen saturation of the venous blood is much higher, this may be important because the impaired organ – the lungs – are better supplied with oxygen, second, cannulation is easier and causes less risk of vascular injury and limb ischaemia.

WLL was recommended to be carried out with saline [1–3]. Our patient responded to saline with severe arrhythmias, which did not occur when prewarmed Ringer’s solution was used. It may be that the patient reacted more to saline since she had undergone a prolonged hypoxaemic period with impairment of the heart. On the other hand, the reduction of arrhythmias during the three WLL may also be due to the general recuperation. However, the authors are convinced that optimization of the lavage fluid contributes to an
improvement of outcome, at least in critically ill patients: lavage fluid should be Ringer’s solution warmed up to 37°C.

4. Conclusion

Our case-report shows that v–vECMO is a safe and easily applied method for adequate oxygenation during WLL in case of respiratory failure due to PAP. WLL can be improved by using 37°C warm Ringer’s solution in stead of saline.

References