Brief Communication

Management of Alveolar Proteinosis by Bronchopulmonary Lavage under Extra Corporeal Membrane Oxygenation (ECMO)

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Abstract

The gold standard of treating Pulmonary Alveolar Proteinosis (PAP) is bronchopulmonary lavage (BPL). We describe a rare case of BPD for PAP, who underwent extra corporeal membrane oxygenation (ECMO) due to hypoventilation in the setting of one lung ventilation. First, the clinical course of the patient is presented; furthermore, the biomolecular basis of PAP and new treatment approaches is discussed.

Keywords: Pulmonary Alveolar Proteinosis, Bronchopulmonary Lavage, ECMO


Introduction

Pulmonary Alveolar Proteinosis (PAP) is a rare and complex disease due to abnormal production of surfactant. Its annual incidence is around 0.36 per million population (1). PAP is more common in male smokers between 30 and 59 years old (2). Based on pathological aspect we can divide it into three main groups: 1) Congenital PAP, 2) Acquired PAP, 3) Secondary PAP (1).

The most common type is acquired type which has auto-immune basis and accounted for 90% of cases (3). Surfactant homeostasis and molecular basis of PAP can be described by figure 1 (4).

The gold standard for treatment of PAP is Whole Lung Lavage (WLL) (5). 60% of patients will fully recover after two WLL, 5% will require WLL every 6 months, and less than 10% will be unresponsive to our treatment (3). And the cause of death in this disease would be uncontrolled infection (72%), Respiratory Failure secondary to PAP (20%), and cases of cardiac arrest during WLL has been reported (3).

Brief Report

A 28-year-old man with history of cigarette and opium smoking was referred due to severe dyspnea and chest pain. At first, he was treated with diagnosis of pneumonia, but had not fully recovered. On physical examination, his vital signs were as follows; Temperature 36°C; Respiratory Rate
induction of anesthesia a double lumen #39 was placed and checked with fiber optic bronchoscopy. Invasive Blood Pressure catheter and central vein catheter was placed and cerebral oximetry was done. After one lung ventilation and starting of lavage on serial ABG we had respiratory acidosis with pCO2 of 105 and pO2 of 53. Due to rising of pCO2 we decided to perform a V-V ECMO.

For this, return cannula was placed percutaneous in the right internal jugular vein and drainage cannula was inserted in the right femoral vein. A 17 Fr. Bio-Medicus (Medtronic, Inc., Minneapolis, MN, USA) arterial cannulae was used as the return cannula and a 19 Fr. multi-port Bio-Medicus (Medtronic, Inc.Minneapolis, MN, USA) cannulae for the drainage cannula. ECMO circuits consisted of a QuadroxD (Maquet Cardiovascular, Wayne, NJ, USA) polymethylpentene oxygenator and a Rotaflow (Maquet Cardiovascular) centrifugal pump. Blood flow of 3-5 liter/min was utilized during the procedure. After ECMO initiation the blood gas values were within normal range. WLL continued with 17 lit of warm Normal Saline for right lung and 13 lit for left lung under ECMO without any complication (Figure 2). The patient was weaned from ECMO at the end of procedure and returned to the ICU.

He was transferred to ICU and after 29 hours he was weaned from ECMO with normal ABG afterward. Unfortunately 20 days after WLL he was expired from resistant pneumonitis.

Discussion

Dr. Benjamin Casteleman published the first report on PAP in 1953 and Dr. Jose Ramirez-Rivera...
performed the first WLL (1). Since then the pathohistophsiological changes occurring in PAP were well studied. In 1990s GM-CSF models of PAP had been published (6). As gene-knocked out mice died from a disease with characteristics of PAP, this link had been studied. Another consistent finding was extracting GM-CSF auto antibodies in serum of patients (7). Consequently new treatments including inhalational and subcutaneous GM-CSF, Rituximab and Plasmapheresis has been advised in various reports (1, 5, 6). On the other hand, another therapy has been emerged with promising results, in which macrophage transplantation is studied for the treatment of PAP (8) (Figure 3).

GM-CSF is a 23 kDa cytokine which produce by numeral cells and bind to heterogeneous cell receptors (4). Ligation of these proteins forms a dodecahedral complex containing α, β and β-associated Janus kinase 2 chains. Autophosphorilation of Janus kinases produce multiple signaling pathways. One of the major effects are in transducer and activator of transcription-5 (STAT5) with effects on myeloid cells (4).

**Conclusion**

In our case, there were several factors resulting in mortality. First, prior history of pneumonitis added to alveolar proteinosis made respiratory failure results in prolonged intubation and mechanical ventilation. Second, based on our experiments the bloody retrieved fluid is related to mortality and could be a sign of poor prognosis. Third, a new multi-drug resistant pneumonitis could cause patterns of ARDS and respiratory failure consequently.

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**Conflicts of Interest**

The authors declare that there are no conflicts of interest.

**References**