Brief Communication

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

Tahereh Parsa[1], Ardeshir Tajbakhsh[2], Zargham Hossein Ahmadi [3], Behrooz Farzanegan[1], Alireza Jahangirifard[2*]

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 extracorporeal 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

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1. Tracheal Disease Research Center, National Research Institute of Tuberculosis and Lung Diseases (NRITLD), Shahid Beheshti University of Medical Sciences, Tehran, Iran
2. Anesthesiology Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran
3. Transplantation Research Center, National Research Institute of Tuberculosis and Lung Diseases (NRITLD), Shahid Beheshti University of Medical Sciences, Tehran, Iran

Corresponding Author:
Alireza Jahangirifard, MD; Anesthesiology Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran. Tel/Fax: (+98) 21-22432572; Email: omidjahangiri_55@yahoo.com; alirezajahangiri@sbmu.ac.ir
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Introduction

Pulmonary Alveolar Proteinosis (PAP) is a rare and complex disease due to the 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 the 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 the acquired type which has an auto-immune basis and accounted for 90% of cases (3). Surfactant homeostasis and molecular basis of PAP can be described in figure 1 (4).

The gold standard for the 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 have been reported (3).

Brief Report

A 28-year-old man with a history of cigarette and opium smoking was referred due to severe dyspnea and chest pain. At first, he was treated with a diagnosis of pneumonia but had not fully recovered.

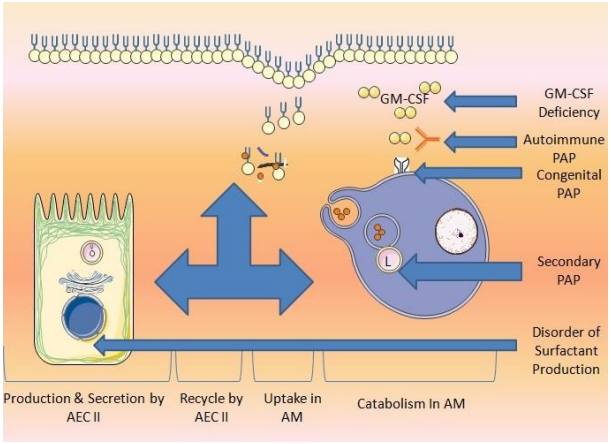

Fig. 1. Disorders of Surfactants Homeostasis.

On physical examination, his vital signs were as follows; Temperature 36°C; Respiratory Rate 25/min; Pulse Rate 110/min; Blood Pressure 95/69 mmHg; O2 Saturation 75% in room air and 92% with facial mask 6 lit/min. On lung auscultation fine crackle was heard at the bases of both lungs. In laboratory examination complete blood count and biochemical profile were within normal range and ABG was as follows; pH 7.36; pCO2 60; HCO3 36; pO2 40.

Further investigation revealed "Crazy Paving Pattern" in CT-scan. Therefore he went on bronchoscopy for bronchoalveolar lavage (BAL) and transbronchial lung biopsy (TBLB). After the procedure he was complicated with pneumothorax and became intubated and due to desaturation, he transferred to ICU. Due to low saturations under mechanical ventilation (63-85%) the process of weaning had failed and after 5 days Percutaneous Dilational Tracheostomy (PDT) was done. At this time the pathologic feature of TBLB was consistent with PAP therefore he was scheduled for WLL.

O2 saturation at the arrival to Operating Room was 71% with FiO2 100%. After the induction of anesthesia, a double lumen #39 was placed and checked with fiber optic bronchoscopy. An invasive Blood Pressure catheter and central vein catheter were 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 the rising of pCO2, we decided to perform a V-V ECMO.

For this, a return cannula was placed percutaneously in the right internal jugular vein and a 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) polymethyl pentene 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 the normal range. WLL continued with 17 lit of warm Normal Saline for the right lung and 13 lit for the left lung under ECMO without any complication (Figure 2). The patient was weaned from ECMO at the end of the 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.

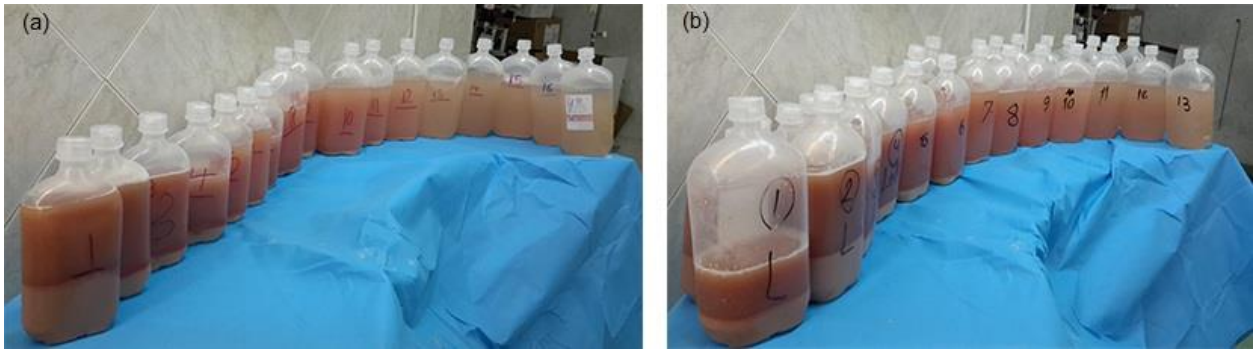

Fig. 2. Retrieved fluid from right (a) and left (b) lung.

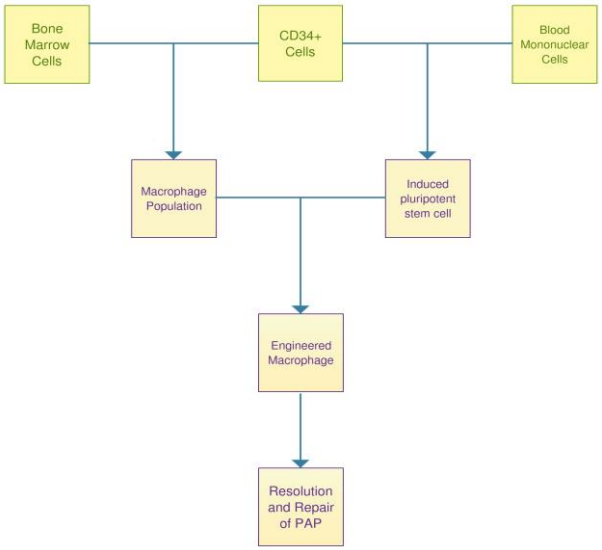

Fig. 3. Macrophage Transplantation for resolution of PAP.

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 histopatho-physiologic changes occurring in PAP were well studied. In the 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 autoantibodies in the serum of patients (7). Consequently, new treatments including inhalational and subcutaneous GM-CSF, Rituximab, and Plasmapheresis have 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 produces by numeral cells and binds to heterogeneous cell receptors (4). Ligation of these proteins forms a dodecahedral complex containing α, β, and β-associated Janus kinase 2 chains. Autophosphorylation of Janus kinases produces multiple signaling pathways. One of the major effects is in the transducer and activator of transcription-5 (STAT5) with effects on myeloid cells (4).

Conclusion

In this case report, several factors led to 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.

Acknowledgment

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Conflicts of Interest

The authors declare that there are no conflicts of interest.

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