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Pulmonary Alveolar Proteinosis *in Extremis*: The Case for Aggressive Whole Lung Lavage with Extracorporeal Membrane Oxygenation Support

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Pulmonary alveolar proteinosis (PAP) is a rare disorder in which lipoproteinaceous material is deposited in the alveoli, compromising gaseous exchange. We report the case of a 29-year-old female patient presenting with the most extreme case of PAP yet reported. She successfully managed by aggressive bilateral whole lung lavage (WLL) in a single sitting using extracorporeal membrane oxygenation (ECMO) support. Despite critical hypercarbia and ventilator-dependence for 12 days before lavage, the patient experienced rapid recovery of pulmonary function after WLL and ECMO could be discontinued on-table. Aggressive WLL with ECMO support can be safe and effective even in the most severe cases of PAP.

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Keywords. Pulmonary alveolar proteinosis (PAP); Extracorporeal membrane oxygenation (ECMO); Whole lung lavage (WLL)

Introduction

Pulmonary alveolar proteinosis (PAP) is a rare disease characterised by the deposition of lipoproteinaceous material in the alveoli, compromising pulmonary gas exchange. Whole lung lavage (WLL) is established as the palliative treatment of choice, but in those very rare cases where the patient already has critical respiratory failure extracorporeal membrane oxygenation (ECMO) support may be required during WLL.[1–9] Herein, we present the most extreme case of PAP-induced respiratory failure to our knowledge to be successfully managed by ECMO-supported bilateral WLL.

Case Report

A 29-year-old woman presented with a five-week history of productive cough, anorexia and malaise. She was a non-smoker with an unremarkable past medical history and no identifiable environmental risk factors. Her chest X-ray on admission showed diffuse pulmonary consolidations

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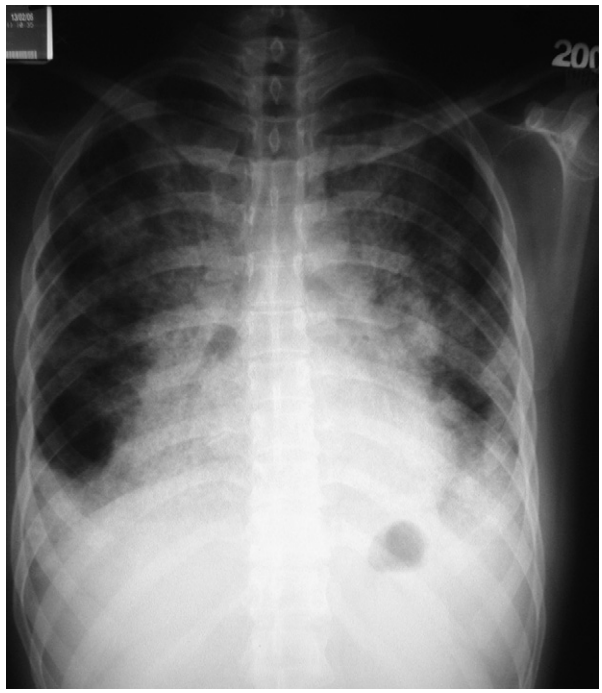

Figure 1. *Chest X-ray on admission showing bilateral patchy lung consolidations, predominantly in the parahilar regions and the middle and lower zones.*

(Fig. 1), and CT scanning confirmed patchy lung consolidations with interlobar septal thickening, giving a 'crazy paving' appearance suggestive of PAP (Fig. 2). She rapidly deteriorated after arrival, with arterial blood gas (ABG) measurements showing a pH of 7.28, pCO_2 of 60.6 mmHg, and pO_2 of 111.1 mmHg on a 100% O_2 rebreathing face mask. She was intubated and given mechanical ventilatory support in ICU. Fibre-optic bronchoscopy (FOB) confirmed large amounts of thick, white, mucoid sputum

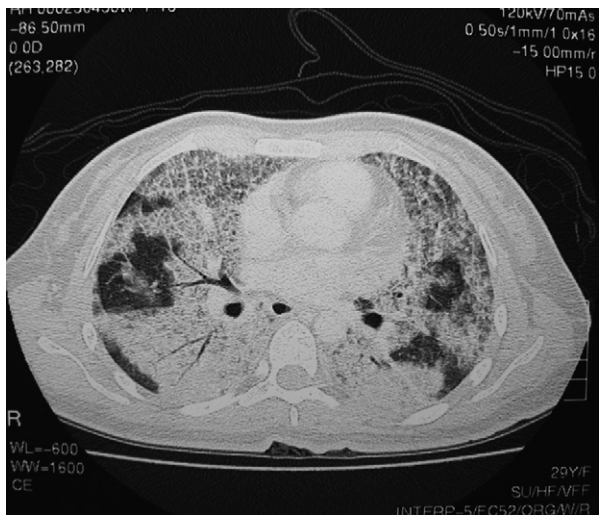

Figure 2. *High-resolution CT scan on admission showing diffuse consolidations and interlobar septal thickening giving a 'crazy paving' appearance suggestive of PAP.*

plugging the airways bilaterally. Analysis of the sputum showed characteristic periodic acid-Schiff (PAS)-positive granular material, consistent with the diagnosis of PAP. Microbiological investigations excluded any respiratory tract infections and the patient remained afebrile with normal leucocyte levels throughout the admission.

The patient was managed with repeated bronchoalveolar lavage (BAL) performed by FOB, and later by sequential bilateral WLL via a double-lumen endotracheal tube. Nonetheless, serial chest X-rays demonstrated progression of her bilateral lung consolidation. Twelve days after intubation, she further deteriorated with ABG measurements now showing a pH of 7.18, pCO_2 of 83.3 mmHg, and pO_2 of 48.2 mmHg on mechanical ventilation with an inspired oxygen fraction (FiO_2) of 1.0. Further attempts at WLL via FOB were precluded by the lack of any further pulmonary reserve, such that the patient suffered prohibitively severe hypoxaemia on instillation of lavage fluid. The Cardiothoracic Surgery Unit was therefore consulted for ECMO support.

The patient was brought to the operating theatre and veno-arterial ECMO was instituted. A 17F arterial cannula was placed to the left femoral artery and a 21F venous cannula placed via the right femoral vein to the right atrium. Sequential bilateral WLL using warm normal saline was performed via the double-lumen tube until the effluent lavage fluid was clear. Chest percussion therapy was maintained by a team of physiotherapists throughout the WLL procedure. The left lung received 19,500 ml of saline lavage with a return of 18,100 ml over a period of 198 min. The right lung then received 26,000 ml of saline lavage with a return of 24,720 ml over a period of 310 min. The total duration of ECMO support was 580 min.

Following completion of the lavage, the patient was immediately weaned from ECMO on the operating table without difficulty and the cannulae removed. The patient was returned to ICU where her ABG measurements now showed a pH of 7.37, pCO_2 of 45.1 mmHg, and pO_2 of 191.1 mmHg on mechanical ventilation with an FiO_2 of 1.0. Twelve hours after the lavage, the patient was weaned from positive pressure ventilatory support, and ABG measurements on 100% O_2 showed a pH of 7.44, pCO_2 of 41.1 mmHg, and pO_2 of 90.1 mmHg. The patient made an unremarkable recovery thereafter and no further ECMO-supported WLL was required. She was discharged home at one month after surgery following a program of respiratory rehabilitation.

Discussion

PAP is a rare disease first described by Rosen in 1958,[10] and whose exact aetiology is still not known. Thus far, there has been no conclusive evidence to show causal associations with smoking, inflammatory, auto-immune, infective or genetic factors. Some have theorised, however, that defective turnover of surfactant in the alveoli may be at least partly responsible.

The management of PAP is accepted to be palliative in virtually all cases. WLL has been established as the preferred palliative therapy to reduce dyspnea and improve