

Anaesthetic considerations for whole lung lavage for pulmonary alveolar proteinosis

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SUMMARY

Pulmonary alveolar proteinosis (PAP) is an uncommon lung disease characterized by excessive accumulation of pulmonary surfactant that usually requires treatment with whole-lung lavage. A 47-year-old female presented with history of dry cough and breathlessness for past 6 months. Chest radiograph demonstrated bilateral alveolar shadows and high resolution computerized tomography thorax showed crazy paving pattern. Broncho-alveolar lavage (BAL) and transbronchial lung biopsy confirmed a diagnosis of PAP. Due to worsening hypoxemia and respiratory failure, whole-lung lavage was planned and performed. Anaesthetic management involved integrated use of pre-oxygenation, complete lung isolation, one-lung ventilation with optimal positive end-expiratory pressure, vigilant use of positional manoeuvres, and use of recruitment manoeuvres for the lavaged lung. We have discussed valuable strategies for the anaesthetic management of patients undergoing this multifaceted procedure in a case of severe PAP.

Keywords: PAP, Whole lung lavage, anaesthesia, management

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INTRODUCTION

PAP is a rare lung disease characterized by the intra-alveolar accumulation of lipoproteinaceous surfactant.¹ The mainstay of treatment is whole lung lavage (WLL). Whole lung lavage is a challenging procedure for both the anaesthetists and the pulmonologists. Procedural course may be complicated by hemodynamic and oxygenation fluctuations. For inexperienced proceduralists, these intraprocedural events may be especially challenging to handle.

The post procedural course may also be unpredictable as the washed lungs are unable to function normally immediately following lavage and patients may require assisted post procedural mechanical ventilation especially when bilateral lung lavage is performed in the same sitting. We describe the anaesthetic management of WLL in a patient with Autoimmune PAP and elaborate the techniques to facilitate this procedure with minimization of risk of complications to the patient.

CASE PRESENTATION

A 47-year-old woman presented with a 6-month history of progressive exertional dyspnoea and non-productive cough. Patient was a lifetime non-smoker and had been diagnosed with hypothyroidism and essential hypertension which were controlled with medications. On examination, patient was hypoxaemic at rest (SpO₂ 87%;

PaO₂=61 mm Hg on room air). Posteroanterior chest radiograph demonstrated bilateral alveolar opacities in peri-hilar distribution. High resolution computerized tomography (HRCT) scan of the thorax showed bilateral ground glass opacities with interlobular septal thickening; suggestive of crazy paving pattern.

Flexible bronchoscopy was performed which showed normal airway anatomy and Broncho-alveolar lavage (BAL) return was milky. Cytological analysis of the BAL fluid demonstrated PAS positive-diastase resistant material. Transbronchial lung biopsy (TBLBx) revealed thickening of alveolar septae and alveoli filled with eosinophilic PAS-positive proteinaceous material. A diagnosis of PAP was established based on the BAL and TBLBx findings.

Anti GM-CSF antibody titres were suggestive of autoimmune PAP. Pulmonary function testing demonstrated a moderate restrictive ventilatory defect (FEV₁/FVC 78%; FEV₁ 63%; FVC 69%) and impaired carbon monoxide diffusion capacity (63% of predicted value). WLL was planned, under general anaesthesia.

In the operating room, Electrocardiography, pulse oximeter (SpO₂) and non-invasive blood pressure monitors were attached.

Case Report

Warming blanket (Bair Hugger™) and warm fluid were used to prevent hypothermia. Radial artery was cannulated for continuous arterial pressure monitoring and arterial blood gas (ABG) analysis. During pre-oxygenation, her SpO₂ improved to >92%. Anaesthesia was induced with 80mg propofol and 100microgram fentanyl and 60mg Rocuronium was administered to achieve muscle relaxation. After 3 minutes of ventilation, a 37 French, left-sided double-lumen endotracheal tube (DLT) was inserted and its position confirmed by a thin flexible bronchoscope (2.8 mm bronchoscope BF-XP160F; Olympus Corp, Tokyo, Japan). As the radiological involvement was greater on the right side, it was decided to lavage the right lung first followed by left lung.

One-lung ventilation (OLV) of left lung was commenced and the pulmonology team performed repetitive cycles of instillation of 1 L aliquots of warmed 0.9% saline solution from a height of 30cm above the patient, followed by passive drainage under gravity. To achieve optimal filling and drainage of all lung segments, a skilled physiotherapist performed manual chest vibration, percussion, positional manoeuvres and gravity drainage. Initially, WLL was started in the supine position and extended periods of desaturation were observed during the drainage phase. To rectify this, positional manoeuvres were used. During inflow, the lavaged lung was made dependent and the patient was positioned in the trendelenburg and reverse trendelenburg position intermittently to facilitate equal distribution of the fluid across the lung. Thereafter, the lavaged lung was made non-dependent to facilitate the outflow of the instilled fluid.

Airway pressure, respiratory system compliance, tidal volume end tidal carbon dioxide concentration, ABG and the net positive balance of the lavaged fluid (difference of the fluid instilled and drained) were monitored. Initially, milky fluid effluent was obtained which became clear later. The procedure lasted approximately 2 hours and a total lavage volume of 12L was used on the right side. After the procedure, asynchronous independent lung ventilation and recruitment manoeuvres were applied to the lavaged lung to restore its expansion.

In view of hemodynamic stability and adequate oxygenation, the right lung was isolated and ventilated and it was decided to WLL on the left lung also. Additional 12liters of fluid lavage was done on the left side in similar fashion. A total of 24 litres was used to lavage both the lungs and a total deficit of 2.6 litres was observed. Serial ABGs revealed a fall in haematocrit suggesting haemodilution so; 10mg furosemide was injected to assist in the removal of excess fluid.

Finally, the DLT was replaced with 8.0 mm single-lumen tube and the patient was transferred to the intensive care unit (ICU) for assisted mechanical ventilatory support. Respiratory system compliance was monitored and showed a gradual improvement from an immediate post-operative value of 21ml/cmH₂O to 51ml/cmH₂O (Figure 1).

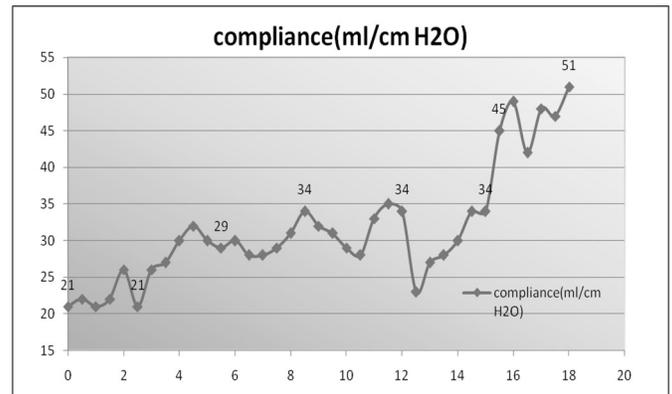


Figure 1 Changes in compliance over a period (hours) after whole lung lavage

Manual chest physiotherapy techniques were continued postoperatively for a few hours. The patient was mechanically ventilated for 24 hours and was given a spontaneous breathing trial on the next day, which the patient tolerated. Patient was extubated 28hours following the completion of the WLL procedure and required low flow oxygen for 2 days following which the patient was normoaxemic on room air.

Serial chest radiographs revealed significant improvement. On Subsequent follow up she has been asymptomatic without any radiologic progression on follow up for last 18 months.

DISCUSSION

We have described a safe and effective anaesthetic technique WLL in a patient with severe respiratory failure due to PAP. Autoimmune PAP is the most common variant of PAP (>90% cases). Presence of auto antibodies against pulmonary granulocyte-macrophage colony-stimulating factor (GM-CSF) results in dysfunction of alveolar macrophages, disruption of surfactant homeostasis and reduced surfactant clearance from alveoli.^{2,3} Investigations usually include radiographic bilateral patchy air-space infiltration, restrictive pulmonary function, impaired diffusion capacity and milky broncho-alveolar lavage (BAL) fluid rich in alveolar macrophages.^{4,5}

Case Report

The results of pulmonary function typically show a restrictive ventilatory defect, and severe reduction of the carbon monoxide diffusing capacity.⁶ In our patient the presentation, pathological, radiological, spirometry and BAL findings were consistent with a diagnosis of PAP.

Lung separation under general anaesthesia and lavage of the non-ventilated lung remain the standard treatment for PAP since first employed by Ramirez-Rivera.⁷ The postulated therapeutic rationale of WLL is the removal of pathological alveolar material and anti-GM-CSF auto antibodies.⁸ A left-sided DLT is preferred as the use of a right-sided tube tends to occlude the orifice of the right upper lobe bronchus.⁹ Furthermore, the shape of the cuff and the presence of the right upper lobe ventilation slit make an airtight seal difficult to accomplish.

Monitoring of airway pressure and tidal volume during one-lung ventilation is crucial to detect fluid leakage into the ventilated lung. An increase in airway pressure or decrease in tidal volume warrants caution as it may indicate a reduction in compliance of the ventilated lung and fluid leakage. Treatment involves endobronchial suctioning followed by effectual re-expansion of the flooded lung. In our patient airway pressure, the tidal volumes and lung compliance were monitored regularly to ensure adequate isolation of the ventilated lung. FOB guided inspection also confirmed that there was no flooding of the ventilated lung. As a safety measure suctioning was done to remove excess fluid that had remained in the lungs after adequate drainage maneuvers.

The major adverse effect from whole lung lavage is hypoxemia which can be minimized by increasing inspired oxygen concentration and managing shunt and V/Q matching during positional change during WLL.^{10,11} Leakage of fluid into the ventilated lung in cases with pre-existing respiratory failure increases the risk of hypoxia. So, adequate lung isolation is the key to success. During thoracic surgery, the operated lung is non-dependent and non-ventilated. The dependent lung is better perfused due to gravity and HPV but is poorly ventilated due to increase in chest wall compliance, resulting in hypoxia and V/Q mismatch. This can be frequently reduced by application PEEP to the ventilated lung.

However, in WLL we kept the lung being lavaged, dependent during filling phase and the position was reversed during the drainage phase. The hydrostatic pressure created by the column of saline above the patient decreased the perfusion to the lavaged lung.¹² This activated the hypoxic pulmonary vasoconstriction (HPV) and increased diversion of blood to the contra lateral ventilated lung. These prevented increases in the venous admixture and severe hypoxemia.¹¹

During the drainage phase, positioning the lavage lung up (ventilated lung dependent) decreased the shunt fraction by diverting the flow of blood flow (gravity assisted) towards the ventilated lung and improved oxygenation. Positioning the ventilated lung in the dependent position during drainage phase helped in improving oxygenation.¹¹ This judicious postural change helped in minimizing hypoxia and shunt during WLL.

So, the patient positioning should be carried out vigilantly. Lateral position with the ventilated lung in the non-dependent position during the filling phase and reversing the position during drainage phase minimized hypoxia and shunt fraction. However, it should be vigilantly done to prevent any flooding of the dependent ventilated lung with fluid.

During the drainage phase of WLL, an attempt to improve saturation by applying high PEEP increased pulmonary vascular resistance and increased hypoxia. Because a high PEEP probably diverted the blood flow to the non-ventilated lung, increased the shunt and lead to desaturation. In contrast, modest PEEP applied by us improved the compliance of the dependent lung and oxygen delivery.

This postural change to improve oxygenation during WLL has not been described earlier. Other strategies that can also be used for the management of hypoxemia during WLL include manual ventilation of partially fluid-filled lung, Hyperbaric oxygen, veno-venous extracorporeal membrane oxygenation¹⁵, intermittent double-lung ventilation, concomitant use of inhaled nitric oxide and almitrine, and pulmonary artery occlusion of the non-ventilated lung using a pulmonary artery catheter.^{13,14,15} At the end of procedure, the patient was shifted to intensive care unit for overnight controlled ventilation to recruit the alveoli.

CONCLUSION

We advocate multidisciplinary teamwork for the use of pre-oxygenation, complete lung separation with a left sided DLT, one-lung ventilation with PEEP, appropriate ventilatory monitoring, cautious use of positional manoeuvres and recruitment manoeuvres for the lavaged lung before single-lumen endotracheal tube exchange for short-term postoperative ventilation. Coordination and teamwork involving the respiratory physician, anaesthetist and the physiotherapist is essential for successful outcome in a patient for WLL.

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