International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Comprehensive Case Report on the Anaesthetic Management of Whole Lung Lavage in Severe Autoimmune Pulmonary Alveolar Proteinosis

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Abstract: ***This case report presents a comprehensive analysis of the anaesthetic management employed in the whole lung lavage (WLL) procedure for a 45 - year - old female diagnosed with severe autoimmune pulmonary alveolar proteinosis (PAP). Characterized by aberrant surfactant accumulation, PAP poses challenges to respiratory function, necessitating therapeutic interventions such as WLL. The anaesthetic strategy involved meticulous planning, lung isolation techniques, and the utilization of total intravenous anaesthesia (TIVA) to enhance procedural safety. Unique to this case was the strategic use of patient positioning during WLL, capitalizing on hypoxic pulmonary vasoconstriction (HPV) to optimize perfusion dynamics. Continuous monitoring, endobronchial suctioning, and judicious positive end - expiratory pressure (PEEP) application further ensured effective lung isolation. The post - procedural course, encompassing 16 - 18 hours of mechanical ventilation, a successful spontaneous breathing trial, and subsequent extubation, underscored the efficacy of the anaesthetic approach. This report contributes valuable insights into refining WLL techniques, emphasizing the integration of HPV and TIVA for optimal patient outcomes in severe autoimmune PAP cases.***

Keywords: whole lung lavage WLL, pulmonary alveolar proteinosis PAP, anaesthetic management, hypoxic pulmonary vasoconstriction HPV, total intravenous anaesthesia TIVA

1. Introduction

Pulmonary alveolar proteinosis (PAP) is a rare lung disorder characterized by an abnormal accumulation of surfactant - derived lipoprotein compounds within the alveoli of the lung (1). This accumulation interferes with the normal gas exchange and expansion of the lungs, ultimately leading to difficulty breathing and a predisposition to developing lung infections (1). The causes of PAP may be grouped into primary (autoimmune PAP, hereditary PAP), secondary (multiple diseases), and congenital (multiple diseases, usually genetic) causes, although the most common cause is a primary autoimmune condition inan individual (1).

The diagnosis of PAP is based on bronchoalveolar lavage, although characteristic x - ray and laboratory test abnormalities occur (2). Treatment involves whole - lung lavage and supportive care (3) (4). Whole - lung lavage is a procedure that involves washing out the lungs with saline solution while the patient is under general anaesthesia (3). The procedure is repeated until the lungs are clear of the accumulated material .

Whole lung lavage requires meticulous planning and knowledge of pulmonary pathophysiology within a multidisciplinary care team. Whole lung lavage may cause hemodynamic instability and hypoxemia, requiring careful anaesthetic management. Lung isolation is necessary to decrease cross - contamination of lungs with lavage washings (5). 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 (5).

2. Case Presentation

A 45 - year - old female presented with a 9 - month history of progressive exertional dyspnoea and non - productive cough. Patient is a known case of hypothyroidism and hypertension which were controlled with medications. On examination, patient was hypoxaemic at rest (SpO2 78%; on room air). High resolution computerized tomography (HRCT) scan of the thorax showed Diffuse ground glass opacities with interlobular septal thickening in bilateral lung field. Broncho – alveolar lavage (BAL) return was send for Cytological analysis, demonstrated lymphocyte in proteinaceous background. Lung biopsy showed dialated alveolar spaces filled with amorphous eosinophilic material, mild interstitial fibrosis and infiltration suggestive of chronic inflammatory infiltrate. A diagnosis of PAP was established based on Lung biopsy repeated BAL and HRCT findings.

Figure 1: HRCT showing diffuse GGO with interlobular septal thickening noted in bilaterallung fields

Whole lung lavage was planned, under general anaesthesia. In the operating room, Electrocardiography, pulse oximeter (SpO2) and non - invasive blood pressuremonitors were attached and warm fluid were used to prevent hypothermia. During pre - oxygenation, patients SpO2 was 100%. Anaesthesia was induced with 120mg Propofol iv and 80 mcg fentanyl iv, 30mg atracurium iv was administered to achieve muscle relaxation. After 3 minutes of ventilation, a 37 Fr, left - sided double - lumen endotracheal tube (DLT) was inserted. The correct placement of the double - lumen endotracheal tube (DLT) was verified through auscultation and fiber opticbronchoscopy. The isolation of both lungs was reconfirmed by ventilating one lung while simultaneously checking for air leaks by venting the non - ventilated lung orifices into saline water placed in a kidney tray, maintaining the ventilated lung at an airway pressure of 40 to 45 cmH2O. The absence of air bubbles confirmed successful lung isolation.

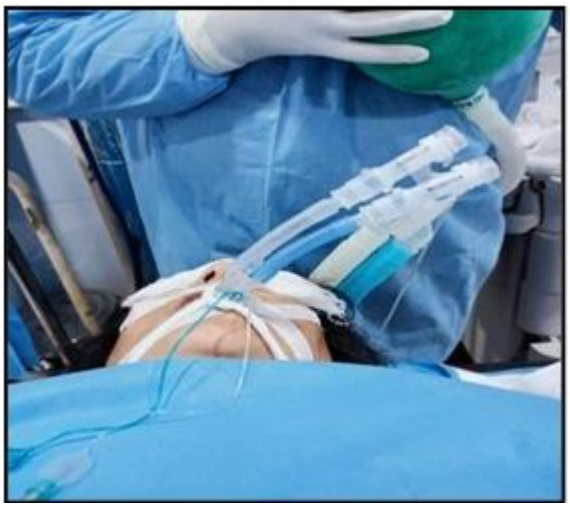

Figure 2: (L) Double Lumen Tube placed to isolate the two lungs prior to lavage

Volume control ventilation with minimal positive end - expiratory pressure (PEEP) of 6 and plateau pressure of 30 mmHg was instituted. Maintenance included oxygen, and propofol infusion.

As the radiological involvement was greater on the right side, it was decided to lavage the right lung. One - lung ventilation (OLV) of left lung was commenced and the pulmonology team performed repetitive cycles (13 cycles) of instillation of 1 L warmed 0.9% saline solution, followed by passive drainage under gravity. To achieveoptimal filling and drainage of all lung segments, a manual chest, percussion, positional manoeuvres were done and was drained under gravity. Normothermia was ensured with warming blankets, and warm normal saline was used for lavage. Throughout the lavage cycles, the patient experienced desaturation episodes down to 75%, prompting lung recruitment maneuvres that improved oxygen saturation to 88–94%. Airway pressures, 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 with subsequent sessions of lavage.

Figure 3: 13 cycles of lung lavage each instilled with 1L of 0.9% NS. sequence showing turbid to clear fluid from left to right

The procedure lasted approximately 3hoursand 30 minutes. A total lavage volume of 13L was used on the right side.20mg furosemide iv was injected to assist in the removal of excess fluid. Patients urine output was monitored intraoperatively with indwelling catheter, produced an output of~1200ml over the intra - op period. Finally, at the end of the procedure the DLT was replaced with 7.5mm single - lumen tube, two - lung ventilation was reinstated, and recruitment maneuvers were applied to restore the expansion of both lungs. Patient saturation was 99 % on FiO2 100%, patient remained hemodynamically stable and was transferred to the intensive care unit (ICU) for assisted mechanical ventilatory support. Over the course in ICU patient remained in Volume control ventilation with a PEEP of7 - 10 cmH2O. The patient was mechanically ventilated for 16 - 18 hours and was given a spontaneous breathing trialon the next day, which the patient tolerated. Patient was extubated 16 - 18 hours following the completion of the WLL procedure and required low flow oxygen for 2 days following which the patient was normoxaemic on room air. Serial chest radiographs revealed significant improvement.

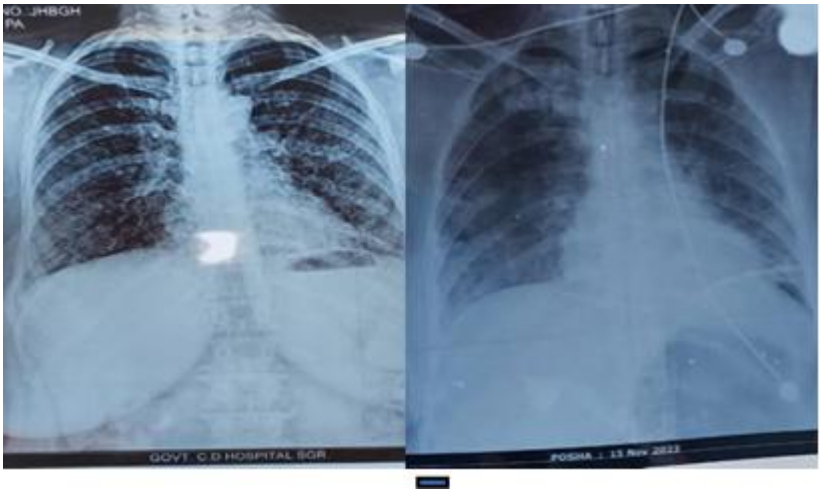

Figure 4: A Comparison between the pre op and post op CXR

3. Discussion

Anaesthetic management of whole lung lavage (WLL) in a patient with severe respiratory failure due to pulmonary alveolar proteinosis (PAP) demonstrates the pivotal role of careful planning and innovative strategies. PAP involves autoantibodies against pulmonary granulocyte - macrophage colony - stimulating factor (GM - CSF), resulting in dysfunctional alveolar macrophages and impaired surfactant clearance. Diagnostic criteria, including radiographic evidence, restrictive pulmonary function, and milky bronchoalveolar lavage (BAL) fluid, were consistent with established findings in PAP cases.

Lung separation under general anaesthesia, with lavage of the non - ventilated lung, remains the standard treatment for PAP, and the preference for a left - sided double - lumen tube (DLT) is justified by anatomical considerations. The most severely affected lung should be lavaged first. The oxygen uptake of each lung is determined separately and the most severely affected side is lavaged initially (6). Meticulous monitoring during WLL is crucial, focusing on airway pressure and tidal volume to detect and address fluid leakage into the ventilated lung. Strategies such as endobronchial suctioning, continuous monitoring of airway pressure, tidal volumes are essential components of maintaining lung isolation during the procedure.

Hypoxic pulmonary vasoconstriction (HPV), is a physiological response to hypoxia, wherein pulmonary vessels constrict in poorly ventilated areas to redirect blood flow to well - ventilated regions, thereby optimizing ventilation - perfusion matching. During WLL, the application of strategic patient positioning, such as placing the lavaged lung in a dependent position during the filling phase, leverages HPV to decrease perfusion to the lavaged lung and prevent severe hypoxemia. Simultaneously, TIVA is highlighted as a preferred anesthetic technique, reducing inhibition of HPV and improving hypoxemia. This combination of HPV and TIVA underscores a comprehensive approach to maintaining adequate oxygenation and mitigating complications during WLL procedures in patients with PAP (5).

Careful consideration of positive end - expiratory pressure (PEEP) dynamics during the drainage phase is crucial, with the case emphasizing the efficacy of modest PEEP in improving dependent lung compliance and oxygen delivery.

This case report contributes valuable insights to the evolving landscape of WLL techniques, emphasizing the importance of an individualized approach in managing PAP - related respiratory failure.

In summary, this comprehensive discussion highlights the successful application of anaesthetic techniques in WLL for autoimmune PAP but also introduces innovative strategies, shaping the future direction of individualized approaches in the management of severe respiratory failure.

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