Case Report

Whole-lung Lavage in a Patient with Pulmonary Alveolar Proteinosis

Abstract

Pulmonary alveolar proteinosis (PAP) is a rare syndrome in which phospholipoproteinaceous matter accumulates in the alveoli leading to compromised gas exchange. Whole-lung lavage is considered the gold standard for severe autoimmune PAP and offers favorable long-term outcomes. In this case report, we describe the perioperative management and procedural specifics of a patient undergoing WLL for PAP in which an anesthesiologist serves as the proceduralist and a separate anesthesiologist provides anesthesia care for the patient.

Keywords: *Anesthesia, one-lung ventilation, pulmonary alveolar proteinosis, whole-lung lavage*

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Introduction

Pulmonary alveolar proteinosis (PAP) is a rare syndrome, with an estimated prevalence of one in two million people. In this syndrome, phospholipoproteinaceous matter accumulates in the alveoli leading to compromised gas exchange.[1] Patients often present with dyspnea and cough with bilateral alveolar opacification on radiographic imaging. Common treatment options include subcutaneous or inhaled granulocyte-macrophage colony-stimulating factor (GM-CSF) and whole-lung lavage (WLL). Although WLL is considered the gold standard for severe autoimmune PAP and offers favorable long-term outcomes, the potential for perioperative complications exists.[2] In this case report, we describe the anesthetic management of a patient undergoing WLL for PAP in which a separate anesthesiologist is the proceduralist as is customary at our institution.

Case Report

A 46-year-old man with autoimmune PAP diagnosed 9 years earlier by lung biopsy presented with progressive productive cough, shortness of breath, and increasing fatigability. Initial pulse oximetry saturation was 90% on room air. A computerized tomography scan demonstrated patchy ground-glass opacities and interlobular septal thickening, consistent with a history of PAP [Figure 1]. At the time of initial diagnosis, he underwent a trial of nebulized GM-CSF without significant improvement in his symptoms. He subsequently underwent WLL, which resulted in resolution of his symptoms for 8 years. Due to his progressive cough and shortness of breath as well as his poor response to GM-CSF previously, repeat WLL was recommended. Pulmonary function testing revealed mild restriction with forced expiratory volume in 1 s (FEV1) 3.13 L (83% of predicted), forced vital capacity (FVC) 3.84 L (82% of predicted), FEV1/FVC ratio 82%, and diffusing capacity of the lung for carbon monoxide (DLCO) 81% of predicted. Despite only mild impairment of pulmonary function testing parameters as had been similarly present before his initial WLL 9 years earlier, recurrence of his severe respiratory symptoms as well as reappearance of chest computerized tomography findings that had both completely resolved following the initial WLL prompted repeat WLL.

As is customary at our institution, the proceduralist performing the WLL was an attending anesthesiologist and intensivist. A respiratory therapist provided technical assistance with instillation and removal of lavage fluid. A second attending anesthesiologist, along with anesthesiology residents, delivered his anesthetic. General anesthesia was induced with fentanyl and propofol, and vecuronium was used for neuromuscular blockade. The patient's trachea was intubated uneventfully by direct laryngoscopy with a 37-Fr left-sided double-lumen endotracheal tube (DL-ETT)

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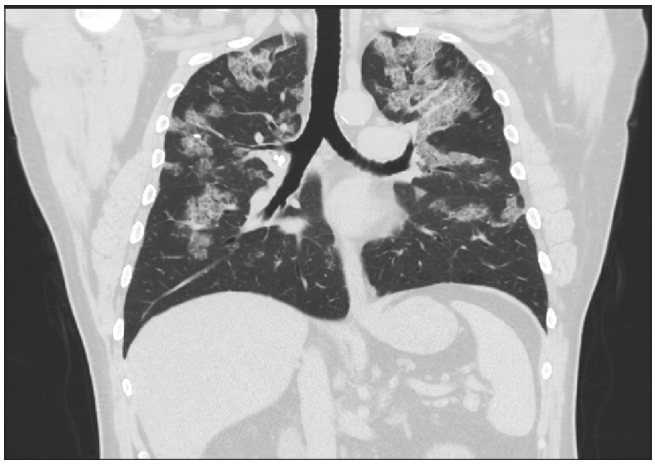

Figure 1: Coronal noncontrast computerized tomography scan demonstrating patchy ground-glass opacities and interlobular septal thickening compatible with a history of pulmonary alveolar proteinosis

with positioning confirmed with flexible bronchoscopy. A radial arterial line was placed after intubation for hemodynamic monitoring, and maintenance of anesthesia was accomplished through total intravenous anesthesia with dexmedetomidine, propofol, and remifentanil infusions. The patient was maintained in the supine position. Arterial blood gas samples were not obtained during the procedure due to use of pulse oximetry and end-tidal carbon dioxide monitoring. The right lung was lavaged with a total of 9.6 L of warm (37°C) sterile normal saline in one liter increments, with a return of 9.1 L. Pulse oximetry saturations on 100% oxygen changed from 97% to 96% during the time from before initiation to immediately following completion of the right WLL (nadir saturation 80% during WLL). Appropriate positioning of the DL-ETT was reconfirmed by flexible bronchoscope between WLL of the left and right lungs. The WLL procedure was then immediately repeated on the left lung with 9.4 L of lavage instillation and return of 9.3 L. Before initiation to immediately following completion left WLL, pulse oximetry saturations on 100% oxygen were unchanged at 96% (nadir saturation 84% during WLL). The total volume of lavage fluid used was based not upon a set required lavage fluid volume but instead on the progressive clinical improvement in fluid clarity from the first to the last saline aliquot in each lung [Figure 2].

Given the potential for transient hypoxemia following WLL, the decision was made to delay extubation and continue mechanical ventilation in the intensive care unit. At the completion of the procedure, the DL-ETT was uneventfully exchanged for a single-lumen ETT utilizing an airway exchange catheter. The patient was then transferred to the intensive care unit. He remained hemodynamically stable and sedated with propofol and on pressure support ventilation with 60% oxygen, pressure support of 5 cm H_2O, and positive end-expiratory pressure of 12 cm H_2O overnight. The following day, he was extubated and

Figure 2: Sequential lavage fluid from left (a) and right (b) lungs demonstrating improving fluid clarity

was dismissed home later that day with an oxygenation saturation of 96% on room air and resolution of his respiratory symptoms.

Discussion

WLL is considered the mainstay of treatment of severe autoimmune PAP, and multiple anesthetic techniques have been reported in a survey of centers performing the procedure.[3] In this case report, we describe a safe and effective method of anesthetic management in a PAP patient undergoing WLL.

Procedural details

A left-sided DL-ETT is typically preferred over a right-sided DL-ETT due to potential difficulty associated with right-sided DL-ETT movement during the procedure resulting in inadvertent leakage of lavage fluid to the contralateral lung and/or inadequate WLL of the right lung. After ensuring appropriate placement and securement of the left-sided DL-ETT, one-lung ventilation is initiated, and the DL-ETT lumen to the nonventilated lung is connected to lavage fluid tubing. The lavage fluid tubing is connected to additional inflow and outflow tubing through a Y-adapter. After clamping the outflow tubing, 37°C sterile normal saline is instilled through gravity in the nonventilated lung in 1 L increments. Meticulous attention to correct DL-ETT position and adequate lung isolation must be employed to ensure no leakage of lavage fluid to the contralateral ventilated lung, the latter of which may be indicated by the presence of bubbling gas through the lavage fluid or presence of lavage fluid in the ventilated lung. Following fluid instillation, the inflow tubing is clamped and the outflow tubing is placed on suction to facilitate fluid drainage. These steps are repeated until the exudate fluid clears, which in our case, was between 10 and 9 L of fluid for each lung. After completion of the unilateral WLL, recruitment maneuvers are utilized to re-expand the lavaged lung, and the WLL procedure is immediately repeated on the contralateral lung if oxygen saturations allow.

There is great variability among clinical practices in which lung is lavaged first. Some centers choose to universally lavage the left lung first due to lower lung volume compared to the right lung.[3] At our institution,

the lung with the most radiographic involvement of PAP is lavaged first. This approach allows for initial one-lung ventilation with the lung less involved with PAP, which we think may mitigate desaturations during the initial WLL. With the improvement in gas exchange following WLL of the "worse" lung, we think that this lung is then better suited for maintaining adequate oxygenation with one-lung ventilation while the other lung undergoes WLL. In addition, there is significant variation in the total amount of lavage fluid used across centers and even within centers among individual patients.[3,4] Similar to our case, other centers utilize visual inspection of fluid clarity as a clinical marker to determine adequate WLL in each lung.

Anesthetic management

Perioperative management for WLL has several potential anesthetic challenges. Given the need to isolate and lavage each lung at various time points in the procedure, we elect to administer a total intravenous anesthetic to ensure adequate and reliable continuous delivery of anesthesia. Perioperative complications reported in a prior survey of centers performing WLL most frequently included transient fever (18%) and hypoxemia (14%).[3] Not surprisingly, one-lung ventilation in patients with preexisting severe lung disease inherent with PAP can result in difficulty with oxygenation and/or ventilation. In addition, the potential for DL-ETT displacement places the patient at risk for lavage fluid flooding the ventilated lung with resulting hypoxemia. Although some institutions may rotate patients to the lateral position for this procedure, we find that drainage of lavage fluid is adequate and the risk of DL-ETT inadvertent movement is minimized by maintaining the patient in the supine position. Regardless of the patient's positioning, ensuring meticulous lung isolation with close monitoring of lung compliance and gas exchange is paramount to success of this procedure. Repeated lavage and drainage result in alterations in pulmonary physiology in the lung being intervened upon.[5] With lavage, pulmonary vasculature compression occurs by the lavage fluid, thereby reducing shunt to this nonventilated lung. During subsequent drainage, the nonventilated lung in turn experiences improves perfusion, resulting in increased shunt to this lung. Following completion of WLL of each lung and with reinitiation of two-lung ventilation, improvement of oxygenation is typically observed. However, the potential exists for postprocedural impairment of oxygenation due to residual lavage fluid and/or incompletely treated PAP. Finally, in patients with severe impairment in oxygenation who cannot tolerate traditional WLL as described above, the use of extracorporeal membrane oxygenation or a hyperbaric chamber could potentially allow for safe performance of WLL in these patients.

Anesthesiologist as proceduralist

The WLL practice at our institution utilizes an anesthesiologist as the proceduralist and another anesthesiologist to direct the anesthetic care of the patient. Both physician anesthesiologists' understanding of the involved cardiopulmonary pathophysiology as well as expertise in airway management and close communication allow for safe perioperative patient care.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Shah PL, Hansell D, Lawson PR, Reid KB, Morgan C. Pulmonary alveolar proteinosis: Clinical aspects and current concepts on pathogenesis. Thorax 2000;55:67-77.
2. Hadda V, Tiwari P, Madan K, Mohan A, Gupta N, Bharti SJ, *et al.* Pulmonary alveolar proteinosis: Experience from a tertiary care center and systematic review of Indian literature. Lung India 2016;33:626-34.
3. Campo I, Luisetti M, Griese M, Trapnell BC, Bonella F, Grutters J, *et al.* Whole lung lavage therapy for pulmonary alveolar proteinosis: A global survey of current practices and procedures. Orphanet J Rare Dis 2016;11:115.
4. Önemli CS, Çatal DA. Whole lung lavage in a pulmonary alveolar proteinosis patient with severe respiratory failure. Turk J Anaesthesiol Reanim 2016;44:111-5.
5. Webb ST, Evans AJ, Varley AJ, Klein AA. Anaesthesia for serial whole-lung lavage in a patient with severe pulmonary alveolar proteinosis: A case report. J Med Case Rep 2008;2:360.