Anesthetic Considerations of Whole Lung Lavage in a Child with Pulmonary Alveolar Proteinosis: A Case Report

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Introduction:
Described in 1958 by Rosen, Pulmonary Alveolar Proteinosis (PAP) represents a rare disease involving accumulation of phospholipoprotein within alveoli and lung interstitium. More than 90% of all cases occur as a primary idiopathic disorder, but rarely the condition is secondary to other pathologies. Patient presentation is nonspecific, with symptoms of dyspnea on exertion and dry cough commonly reported. The vast majority of cases occur in the third or fourth decade of life. The gold standard for diagnosis is open lung biopsy. However, bronchoscopy with bronchoalveolar lavage and transbronchial biopsy is now being used. The most effective treatment for PAP is whole lung lavage (WLL) typically performed using double lumen tubes, since over 80% of reported cases occur in adults. A case of PAP in a child presenting for WLL and a review of the anesthetic challenges is submitted.

Case Report:
A 10 year old boy with a history of recurrent pneumonia, asthma, and worsening dyspnea on exertion presented from an outlying community hospital. Review of the documents accompanying him at transfer disclosed that bronchoscopy and bronchoalveolar lavage (BAL) were complicated by bronchospasm necessitating termination of the procedures. Therefore, a video assisted thoracic surgery (VATS) with lung biopsy was required for definitive diagnosis. Biopsy disclosed Pulmonary Alveolar Proteinosis. He was transferred to our institution for further management. Initial auscultation of the lungs revealed inspiratory and expiratory wheezing bilaterally. Baseline oxygen (O2) saturation was 88% on room air, greater than 90% with O2 by nasal cannula. Pulmonary function tests revealed a restrictive lung disease (FVC- 64%, FEV1-68%, ratio-93%).

The patient was scheduled for WLL under general anesthesia using one-lung ventilation. After informed consent was obtained, the patient was brought to the operating room. Since the patient had no intravenous (IV) access, an inhalation induction using Sevoflurane in 50% nitrous oxide and oxygen was performed once EKG, pulse oximetry, and BP cuff were in place. Induction was complicated by laryngospasm, relieved by intramuscular succinylcholine and followed by intravenous vecuronium. The patient could now easily be ventilated by mask. Direct laryngoscopy revealed a grade II view of the glottis. However, a 6.5 ID cuffed endotracheal tube (ETT) could not be passed beyond the vocal cords, and a 6.0 cuffed ETT was placed with no leak >40cm H2O. A left-sided 28 F double lumen endotracheal tube (DLT) was placed under fiberoptic guidance to replace the single lumen tube. Lavage was started through the bronchial lumen of the DLT on the left which was more affected as assessed by his chest x-ray. Five hundred milliliters of saline were instilled by gravity from approximately three feet above the patient. There was overspill of lavage fluid into the trachea and tube position was reconfirmed fiberoptically and radiographically. Subsequently, lavage was performed using smaller volumes and less hydrostatic pressure without further spillage into the ventilated lung. With every cycle of saline instillation, the patient’s saturation dropped to 75-90% on FiO2=1 requiring vigorous positive pressure ventilation. Oxygenation would recover with drainage of the instilled saline. After over 12 cycles of lavage, the recovered saline became increasingly clear from milky white layered to slightly transparent fluid. The left lung lavage was complete after 4.8 liters in and 4.7 liters out. Due to the difficult oxygenation, the decision was made to lavage the right on a subsequent day. Decadron 12 mg IV was given to minimize airway edema and the patient was extubated in the operating room. He immediately became tachypneic with a respiratory rate up to 60 breaths/min and hypoxic requiring 15 liters O2 by face mask to maintain saturations > 90% in the recovery room. Transfer to the Intensive Care Unit was necessary for 24 hours until he weaned to an FiO2 of 35% O2.
The patient returned to the operating room three days later for right lung lavage. Direct laryngoscopy was more difficult due to swelling with a grade IV laryngoscopy, requiring fiberoptic placement of a DLT. However, a left-sided 28 F DLT would not pass secondary to airway edema, but a left-sided 26 F DLT was successfully placed and confirmed in correct position. This time, with smaller lavage volumes, lower hydrostatic pressures for instillation, and much improved chest x-ray, the patient’s intraoperative oxygenation was easier to maintain with saturations of 93-99% on 100% O2. A total of 4.8 liters of saline was instilled with 4.7 liters drained from the right lung. He was extubated in the operating room, and taken to the recovery room without complications. He returned to the floor and discharged home the following day with marked improvement in dyspnea on exertion, but still required O2 by nasal cannula at night.

Discussion:
Pulmonary alveolar proteinosis is caused by a defect of alveolar macrophage function in the clearance of surfactant. Surfactant reduces the surface tension in the alveolar wall preventing alveolar collapse. Over the last decade, granulocyte-macrophage colony stimulating factor (GM-CSF) was found to play a critical role in surfactant homeostasis. In PAP, interruption of GM-CSF signaling in the macrophages impairs the breakdown of surfactant without impairing its uptake, resulting in accumulation of surfactant in alveoli.

Whole lung lavage is the best treatment that can be offered to patients with PAP. When it was first described in 1960 by Dr. Jose Ramirez-Rivera, the procedure was prolonged, distressing, and burdensome without sedation or anesthesia. The evolution of WLL now includes general anesthesia, increased lavage volumes, and completion of both lungs in the same treatment session. Anesthesia is complicated by the restrictive lung disease, hypoxemia, and the need for lung isolation. While the placement of a double lumen tube is routine in adults, it may be challenging in children. The use of WLL in neonates and young infants is less established because of the technical difficulties passing a large DLT as well as a bronchoscope to perform lavage. Lavage through a Foley catheter or bronchoscope placed into one mainstem while ventilating the other lung, cardiopulmonary bypass, extracorporal membrane oxygenation, or hyperbaric oxygen have been reported for children too small for double lumen tubes.

Our case presents an example of the lower age range for the use of a double lumen tube allowing WLL. Intraoperative overspill of lavage fluid and hypoxemia indicates the need to limit lavage volume and hydrostatic irrigation pressure to minimize the risk of spillage into the ventilated lung.

References:


