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Whole lung lavage in pulmonary alveolar proteinosis: anesthetic management and challenges

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Abstract

Background: Pulmonary alveolar proteinosis is a rare disorder characterized by alveolar obstruction secondary to the collection of lipoproteinaceous material in the alveoli leading to a spectrum of respiratory illness ranging from mild to severe respiratory failure. Whole lung lavage considered the gold standard for the treatment of PAP is performed under general anesthesia and presents a unique set of challenges to the anesthesiologist. The procedure involves manually removing the lipoproteinacious material by repeating aliquots of fluids and draining periodically after percussion. When done effectively it is quite effective and life-saving procedure.

Case presentation: A 39-year-old female presented with history of gradually progressive shortness of breath, diagnosed to have primary pulmonary alveolar proteinosis for which bilateral whole lung lavage was planned. During the lavage cycles, patient had episodes of desaturation to 60% during which the double lung ventilation was resumed. Her serial ABG revealed a deteriorating oxygenation. In view of increased oxygen requirements, the procedure was carried out in two stages. She made a complete recovery after the procedure.

Conclusions: A multidisciplinary team working with good communication, use of adequate back up plans, meticulous lung separation with double lumen tube, vigilant intraoperative monitoring, short-term postoperative ventilation go a long way in achieving adequate gas exchange in patients with respiratory failure due to pulmonary alveolar proteinosis.

Keywords: Pulmonary alveolar proteinosis, Whole lung lavage, Lipoproteinatious

Background

Pulmonary alveolar proteinosis (PAP) is a rare disorder characterized by alveolar obstruction secondary to the collection of lipoproteinaceous material in the alveoli leading to a spectrum of respiratory illness ranging from mild to severe respiratory failure (Rosen et al. 1958). Whole lung lavage (WLL) considered the gold standard for the treatment of PAP is performed under general anesthesia and presents a unique set of challenges to the anesthesiologist. The procedure involves manually removing the lipoproteinacious material by repeating aliquots of fluids and draining periodically after percussion. When done meticulously it is quite effective and life-saving procedure (Beccaria et al. 2004). There is desaturation during filling of lungs during WLL to functional residual capacity (FRC), that requires prompt management should the patient deteriorate. A safe conduct of WLL includes not only the pathophysiological understanding of the disease but also calls for a well-coordinated team work by the anesthesiologist, proceduralist, and physiotherapist. We report the successful conduct of a two stage whole lung lavage in a case of PAP with respiratory failure.

Case presentation

A 39-year-old female with no significant past history presented with history of gradually progressive shortness

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Fig. 1 Chest X-ray AP view, showing diffuse PAP

of breath and non-productive cough for 6 months. Based on the imaging studies and bronchoalveolar lavage (BAL) findings, she was diagnosed to have primary pulmonary alveolar proteinosis for which bilateral whole lung lavage was planned. On preoperative examination, the patient was on nasal prongs with oxygen at 4 l/min. Her respiratory rate was 30 breaths/min. She was tachycardic (120 beats/min) and chest auscultation revealed bilateral crepitation. Chest radiograph showed bilateral ill-defined opacities in lung parenchyma (Fig. 1). Preoperative HRCT chest revealed patchy areas of ground glassing with inter and intralobular septal thickening diffusely involving bilateral lung parenchyma. The examination of BAL was suggestive of PAP. Serum LDH levels were elevated. Routine lab investigations were normal. The preoperative ABG showed Po2: 60.5, Pco2 38.5, PH = 7.4, HCO3 = 25. Preoperative echocardiograph was normal. Pulmonary function test revealed a moderate restrictive pattern (FEV1/FVC79%; FEV1 62%; FVC 70%) . Written informed consent was taken from the patient. On the day of procedure, the patient was taken to the operating room on trolley with oxygen via nasal prongs at 4 l/min. All standard ASA monitors were attached and intravenous line secured with a 18-G iv cannula. After preoxygenation with 100% oxygen for 3 min, anesthesia was induced with 120 mg of propofol, 120 μg micrograms of fentanyl. To facilitate endotracheal intubation 6 mg of vecuronium was given. The trachea was intubated and the lung separation was ensured by placing left-sided 37 F double lumen endobronchial tube (DLT) into the left main stem bronchus. The correct position of DLT was confirmed with auscultation and fiberoptic bronchoscopy. The isolation of both the lungs was reconfirmed by ventilation of one lung and simultaneous checking for air leak by venting the non-ventilated lung tube orifice into the saline water seal cup while the ventilated lung was held at an airway pressure of 40–50 cm of water (Fig. 2). The absence of air bubbles confirmed the lung isolation. Pressure control ventilation with minimum PEEP of 6 and plateau pressure under 30 mmHg was set. Maintenance of anesthesia was done with sevoflurane, oxygen, top up doses of vecuronium, and fentanyl. A left-sided femoral arterial cannula was inserted for continuous hemodynamic monitoring. SPO2, capnography, invasive bp monitoring, urine output, nasopharyngeal temperature, and serial ABG monitoring were done during the procedure. Normothermia was ensured by using the warming blanket and prewarmed normal saline for lavage. Based on the chest radiograph findings and after discussion with the proceduralist, it was decided to proceed with the left lung lavage first followed by right if the patient tolerates the procedure well. We decided to proceed with supine position to prevent dislodgment of the tube. After initiation of right-side lung ventilation, seal was rechecked. A Y connector was attached from one side to the target lung tube and from the other side to the lavage fluid (prewarmed normal saline) and to the drainage fluid container. The clamps allowed the proceduralist to control the flow of fluid in and out of the lung. Five hundred milliliters of prewarmed normal saline was instilled into

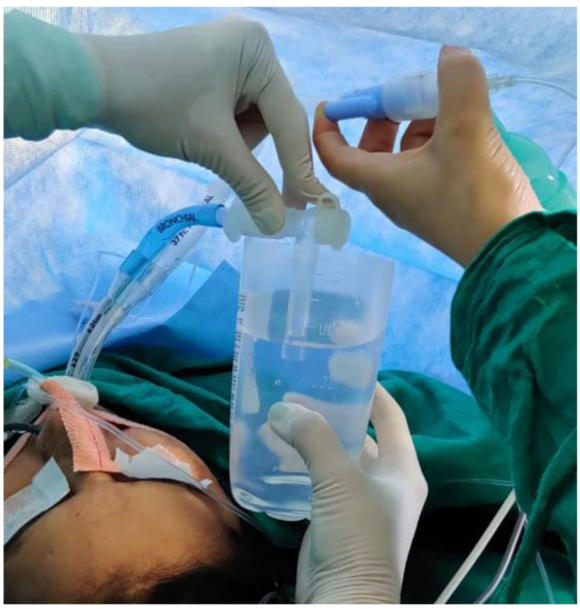

Fig. 2 Checking for adequate seal by water bubble method

Fig. 3 Lavage fluid for visual comparison

the lung under the gravitational force of saline column followed by manual chest percussion for 5 min. The saline was then drained to gravity after releasing the clamp on the Y connector. Each cycle took 5–10 min. The initial effluent was milky and total lavage volume of 10 L used to obtain a less turbid fluid effluent (Fig. 3). During the lavage cycles, patient had episodes of desaturation to 60% during which the double lung ventilation was resumed. Her serial ABG revealed a deteriorating oxygenation. In view of increased oxygen requirements proceduralist decided to abandon the procedure after one lung lavage only. At the end of procedure, two lung ventilations were started and recruitment maneuvers were applied to restore expansion of both the lungs. DLT was exchanged for a 7.5-mm single lumen endotracheal tube and the patient was shifted to intensive care unit for elective ventilation. The patient was extubated next day in ICU. Her fio2 requirements were reduced and she maintained oxygen saturation of 97% on room air. Her post extubation chest radiograph demonstrated radiological improvement. A day later, similar procedure was followed for right lung lavage. She was followed for a month and made a complete recovery.

Discussion

First described by Rosen Castleman and lie bow in 1958 (Rosen et al. 1958), pulmonary alveolar proteinosis (PAP) is a rare disorder, characterized by mechanical alveolar obstruction secondary to the collection of PAS (para aminosalicylic acid) positive, amorphous lipo proteinaceous material in the alveoli leading to mild to severe respiratory failure. Whole lung lavage is considered the gold standard for the treatment of PAP that involves sequential cycles of lavage through repeated filling and emptying of the affected lung to remove excess alveolar phospholipids (Beccaria et al. 2004). A safe conduct of whole lung lavage includes not only the pathophysiological understanding of the disease but also calls for a well-coordinated team work by the anesthesiologist, proceduralist, and physiotherapist. Whole lung lavage is generally indicated in patients with declining lung function, declining oxygenation, and radiographic worsening. Depending on the preoperative status of the patient a stand by ECMO or CPB can be planned. Anesthesia is usually induced with a combination of intravenous propofol, opioid, and neuromuscular blocker, although TIVA has also been used successfully in these cases. Besides the standard ASA monitoring, invasive BP, ABG monitoring is required. Temperature monitoring is important since patient is prone to develop hypothermia after lavage with more than 10 L of saline which is usually used in WLL cases. A serial ABG intraoperatively helps to monitor the oxygenation and ventilation and alerts the proceduralist for timely abandoning the procedure if required. An optimally placed double lumen tube with adequate seal is the backbone of this procedure. Ensuring the meticulous isolation of the lung using auscultation, flexible fiberoptic bronchoscope and saline water seal cup helps in avoiding inadvertent leakage of the lavage fluid into the ventilated lung .The presence of a bubbling gas in the lavage fluid or the presence of the lavage fluid in the ventilated lung, increased airway pressure, reduced lung compliance, and imbalance between the administered lavage volume and drained lavage volume should alert the anesthesiologist of the loss of lung isolation. Depending on the preoperative status of the patient either a sequential unilateral or a bilateral lung lavage can be planned in PAP. If a sequential lavage is planned and both the lungs are equally involved, the left lung lavage is preferably done first to support the gas exchange. The procedure can be done in supine, prone or lateral position with each position having its own pros

and cons. Prone and lateral decubitus position with lavaged lung up and reverse Trendelenburg position have been reported to result in better drainage of the fluid (Campo et al. 2016; Silva et al. 2014). The evidence for the benefit of one position over other is limited. Low oxygen saturation during WLL is not uncommon. Ventilation perfusion mismatch during lung lavage lead to hypoxemia. During the influx of the lavage fluid the perfusion of the non-ventilated lung is reduced by compression of pulmonary vasculature thereby reducing the shunt, while during the efflux of the fluid reperfusion of the non-ventilated lung increases the shunt leading to hypoxemia. Manual ventilation of the partially fluid filled lung, intermittent double lung ventilation, concomitant use of inhaled nitric oxide, ipsilateral pulmonary artery occlusion of the non-ventilated lung using pulmonary artery catheter, hyperbaric oxygen, extra corporal membrane oxygenation (ECMO) have been used for the management of refractory hypoxemia. We kept standby cardiopulmonary bypass, in case the patient did not respond to initial manouvres. Other complications associated with this procedure include pneumothorax, pleural effusion, hydropneumothorax. Manual percussion of the chest wall by a physiotherapist increases the clearance of the proteinaceous material. Hammom et al. (2014) demonstrated that the optical density of the recovered lavage fluid is greater from the patients who received manual percussion than those who received percussion using hand held devices or no percussion. The total volume of lavage volume used for WLL varies with an average of 15.4 ± 6.8 L per lung. A total volume of 250 ml/kg can be used on an average in adults and children. Depending on the intraoperative ABG findings, requirements of fio2, hemodynamics of the patient can be either electively ventilated or extubated at the end of the procedure.

Conclusions

A multidisciplinary team working with good communication, use of adequate back up plans, meticulous lung separation with DLT tube, appropriate ventilatory monitoring with adequate use of PEEP, vigilant intraoperative monitoring, short-term postoperative ventilation go a long way in achieving adequate gas exchange in patients with respiratory failure due to pulmonary alveolar proteinosis.

Abbreviations

PAP: Pulmonary alveolar proteinosis; WLL: Whole lung lavage; DLT: Double lumen tube; PEEP: Peek end expiratory pressure; ECMO: Extra corporal membrane oxygenation

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Authors' contributions

MM helped in the searching of the literature and helped in the methodology. PB helped in the editing and writing of the manuscript. Both MM and PB helped in the basic outline of the manuscript. The authors read and approved the final manuscript.

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Availability of data and materials

The data was extracted from the medical records file of the patient and the materials are being used which were available in our set up with valid reasons.

Declarations

Ethics approval and consent to participate

Ethical approval is not required for publication of isolated case reports.

Consent for publication

Written permission/consent for reproduction of images of the patient for the purpose of publication in an educational medical journal was obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

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