Case Report

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Anaesthetic management of a case of alveolar proteinosis
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Abstract

Pulmonary alveolar proteinosis is a rare type of interstitial lung disease characterised by diffuse involvement and filling of distal air spaces by the amorphous, periodic-acid-schiff positive lipoproteinaceous material. Whole lung lavage is presently the standard mode of management of such cases. A 33-year-old male presented to our centre with cough and breathlessness on exertion. On evaluation he was found to have PAP. He underwent whole lung lavage (WLL) under general anaesthesia using one lung ventilation. We describe the anaesthetic consideration and the challenges faced in such a procedure.

Keywords: One Lung Ventilation, Pulmonary alveolar proteinosis, Lung lavage.

INTRODUCTION

Pulmonary alveolar proteinosis is a rare form of interstitial lung disease. The basic pathology lies in deposition of lipoproteinaceous material in the alveolar spaces.[1] Mechanical clearing of the alveolar deposits by large volume bronchoalveolar lavage is the standard procedure practiced now-a-days to improve such conditions.[2] Approach to such procedures has changed drastically over years but literatures are very few regarding the anaesthetic management of such conditions. Anaesthetic management of such procedure needs to consider interstitial lung disease, preoperative hypoxemia, one lung ventilation, implications of different positions and prolonged procedure.

CASE REPORT

A 33-year-old male presented to our centre with cough and scanty mucoid expectoration of 06 month duration and breathlessness on exertion of 3 months duration. At the time of presentation his effort tolerance was good (metabolic equivalent score, METS>4). Evaluation confirmed the diagnosis of PAP and finally the patient was subjected for WLL.

During preoperative check-up haemogram, liver and renal function tests were found within normal limits; 2D-echocardiogram showed left ventricular ejection fraction (LVEF) 60%; the arterial blood gas (ABG) sample while the patient is on room air showed pH = 7.39, PaO2 = 56.6, PaCO2 = 31.1, HCO3 =21.1, SaO2 89.8. The precise clearance of alveolar lipoprotein material was planned to perform in two sittings.

In the first sitting, the lavage of left lung was carried out under general anaesthesia with ventilating the right lung. One lung ventilation was maintained by a 41Fr (Lt) double-lumen tube (DLT) and the position of the tube was confirmed a 2.8 mm fiber-optic bronchoscope. Pt was made to lie in right-lateral position to lavage the non-dependant lung.

Lavage involved 1000 ml – 1200 ml aliquots of normal saline warmed to body temperature and rapidly instilled into the left lung using a wide bore perfusion tubing with a Y-shape connection (fig. 1) over 5-10 min. While the fluid was indwelling, chest percussions were performed to loosen the proteinaceous material. The saline was then drained from the lung by gravity. This step was repeated 10-12 times till the colour of drainage fluid became clear.

Throughout the procedure invasive blood pressure, central venous pressure, urine output, SaO2, capnography, naso-pharyngeal temperature, repeated ABG, serum electrolytes and fluid input-output...
were monitored meticulously. After 5-6 cycles of lavage patient had desaturation due to spillage of lavage fluid into the ventilated lung. It was managed with thorough suctioning and oxygenation.

At the end of the procedure DLT was replaced with standard 9.0 Portexuffed single lumen endotracheal tube and shifted to intensive care unit for postoperative ventilation. The procedure took 5 hours with total aminuted saline of 9L and effluent fluid of 7.5L. Patient remained intubated and ventilated for an additional period of 16 hours before extubation next morning. After extubation the ABG showed PaO₂ 61, SaO₂ 92% (Table 1); patient remained comfortable in room air with a respiratory rate of 14-16/min.

**Table 1: ABG analysis during whole lung lavage of left lung**

<table>
<thead>
<tr>
<th></th>
<th>pH</th>
<th>PaCO₂</th>
<th>PaO₂</th>
<th>HCO₃⁻</th>
<th>SaO₂</th>
<th>PEEP</th>
<th>FiO₂</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-procedure</td>
<td>7.41</td>
<td>33.5</td>
<td>63.0</td>
<td>25.0</td>
<td>91.0</td>
<td>10</td>
<td>0.7</td>
</tr>
<tr>
<td>End of procedure</td>
<td>7.29</td>
<td>53.4</td>
<td>59.4</td>
<td>24.8</td>
<td>90.0</td>
<td>10</td>
<td>0.7</td>
</tr>
<tr>
<td>Both lung ventilation</td>
<td>7.34</td>
<td>47.7</td>
<td>76.0</td>
<td>23.5</td>
<td>93.0</td>
<td>10</td>
<td>0.7</td>
</tr>
<tr>
<td>16 hr post ventilation</td>
<td>7.39</td>
<td>33.3</td>
<td>72.3</td>
<td>22.2</td>
<td>92.8</td>
<td>5</td>
<td>0.5</td>
</tr>
<tr>
<td>Post extubation</td>
<td>7.44</td>
<td>34.0</td>
<td>68.0</td>
<td>24.1</td>
<td>91.7</td>
<td>-</td>
<td>0.2</td>
</tr>
</tbody>
</table>

**Figure 1: Graphical representation of Lavage of Lt lung**

This patient underwent right lung lavage after 02 weeks with 12 L of normal saline with same anaesthetic management using a 39Fr (Rt) DLT. But this time we made the patient to lie down again in right-lateral position and lavaged the dependant lung (Rt).

**DISCUSSION**

Pulmonary alveolar proteinosis (PAP) is described as an orphan lung disease,[1] characterized by abnormal processing of surfactant due to macrophage dysfunction leading to deposition of amorphous material and cellular debris in the alveoli.[2,3] that finally results in impairment in gas exchange and variable severity of respiratory symptoms.[3] Epidemiological data is scarce; the annual incidence is 2 to 5 per million.[3] At the time of diagnosis, the median age is 40 years; most patients are men, and about three-quarters of them have a history of smoking.[1]

WLL is the standard therapy now practiced for PAP.[1] It’s the refined technique of mechanical clearing of the deposited lipoproteinaceous material from the alveoli. Ramirez started lung lavage in the name of ‘repeated segmental flooding’[4] in 1964 using up to 3 L of saline (with heparin or acetylcysteine) under local anesthesia. Subsequent development in the procedure routinely involves lavage of both the lungs under general anesthesia,[5] increased lavage volumes and addition of chest percussion.[6] Presently in some centers it is performed in the form of bilateral sequential lung lavage in the same session[7] or lavage of both the lungs, one after another in a week’s time.[1]

It is advised to lavage the more affected lung first for allowing better lung to provide gas exchange.[9] Left lung is lavaged first in our case because of equal involvement of both lungs so that the larger right lung was left to support gas exchange during one-lung ventilation.

We used an advanced OT table having facility for adjustment to achieve different positions improves the quality of WLL. Gaetane et al recommends use of an OT table having electrically adjusted positions for trendylenberg and reverse trendylenberg positions.[2] Though there are no recommendations, the lavage is conducted in lateral decubitus position to lavage dependent lung and to ventilate the nondependent lung. Beccaria et al ventilated the dependent lung and lavaged the nondependent one to give better ventilation perfusion ratio. Even lavage has been tried in prone as well by Andrew Perez and colleagues.[10] We performed the procedure in Rt lateral position to lavage the Lt lung first. But due to intra-op spillage in the first session, we maintained the same rt lateral position even for the lavage of Rt lung in the next session.

Infusion of large volumes of saline in lungs is associated with increased intrathoracic pressure, CVP. Pulmonary capillary wedge pressure; it has been found that ventricular filling is decreased due to impaired venous drainage[11], so it is important to monitor the patient with ASA standard monitors along with IBP & CVP monitoring. We managed to maintain
the physiologic variables within 20% of baseline and CVP within 8-12 cm H$_2$O.

Maintaining normothermia is one of the prime requisites of the procedure. We have used warming blankets and warm IV fluids and monitored the body temperature was monitored by a nasopharyngeal temperature monitoring probe.

At the end of the procedure, the patient can be extubated in the OT and then to be shifted to ICU for monitoring. We ventilated the patient in the post-operative period to rule out the complications of intra-operative spillage.

The potential complications of such elective procedure are hypoxia, pneumonia, sepsis, ARDS, pneumothorax and ischaemic complications of extremities; We encountered hypoxia because of intraoperative spillage of lavage fluid into the ventilated lung in the first session which was treated conservatively by administering F$_{2}$O$_{2}$ 1, continuing positive pressure ventilation and re-establishing the lung isolation. But hyperbaric oxygen therapy and sometimes use of extracorporeal membrane oxygenation to correct the oxygen saturation in such critically ill patients can be considered.

CONCLUSION

The procedure of Whole lung lavage needs proper lung isolation throughout the procedure. Strict vigilance and maintaining the vital parameters within the normal physiologic range is the key to uneventful outcome of such a prolonged procedure.

REFERENCES