Whole Lung Lavage in a Patient with Pulmonary Alveolar Proteinosis: A Case Report

CHIH-YANG CHUNG¹, CHONG-KWAI CHEN², HUNG-PIN LIU², ANGIE CY HO³, MIN-WEN YUNG², AN-HSUN CHOU²

Pulmonary alveolar proteinosis (PAP) is a rare lung disease characterized by accumulation of protein and lipid material, called surfactant, within the air sacs or alveoli of the lungs with for unknown reasons. Because of the unknown pathogenesis, the most effective treatment for PAP is the mechanical removal of the proteinaceous material via whole-lung lavage (WLL) with isotonic saline as the lavage solution, and mechanical chest percussion during lavage. Serial arterial blood-gas determinations and measurement of lung compliance are done in during the intraoperative and postlavage periods. The major complications of WLL are hypoxia, circulatory disturbance and difficulty in positioning the double lumen endotracheal tube. An experienced team is needed in the operating room and a postoperative care facility is also required to reduce complications. Here we report a 44-year-old man with a history of diesel fuel inhalation, suffered from dyspnea on exertion, a productive cough, and low-grade fever. An open lung biopsy confirmed the diagnosis of PAP. The patient underwent WLL under general anesthesia. His clinical symptoms were improved and discharged with a relatively stable condition.

Key words: pulmonary alveolar proteinosis, whole lung lavage, one-lung ventilation

Introduction

Pulmonary alveolar proteinosis (PAP) is a rare lung disease. The pathohistology of PAP is associated with the filling of the alveoli by a proteinaceous material. The most effective therapy for PAP is whole-lung lavage (WLL) under one-lung anesthesia. Here we report a patient with PAP who underwent WLL under general anesthesia.

Case Report

A 44-year-old man with a history of diesel fuel inhalation, suffered from dyspnea on exertion, a productive cough, and low-grade fever for several days. An open lung biopsy confirmed the diagnosis of PAP. The chest radiograph showed increased infiltration over both lung fields. High resolution computed tomography of the both lungs showed diffuse ground glass attenuation with multifocal sparing bilaterally with thickening of the interlobular septa, intralobular interstitium and bilateral subpleural interstitium. The hemoglobin concentration, serum electrolytes, urea nitrogen and creatinine concentration were normal as was the electrocardiogram. Pulmonary function tests
revealed forced expiratory volume in 1 second (FEV₁) 2.0 L, forced vital capacity (FVC) 2.2 L and diffusing capacity of the lung for carbon monoxide 10.7 ml/mmHg/min with a restrictive ventilatory defect. Arterial blood gas analysis showed pH 7.464, PaCO₂ 37 mmHg, PaO₂ 72 mmHg, and SpO₂ (pulse oximetry) 92% while breathing 31% O₂ with a venturi mask. Bilateral WLL was done.

Anesthesia was induced with thiopental 5 mg/Kg, fentanyl 150 µg, and rocuronium 40 mg, via tracheal intubation with a 35 French left sided double lumen endotracheal tube (DLT). Sevoflurane 2-4% in 1 L O₂ was used to maintain anesthesia. The position of the DLT was checked by auscultation and was also confirmed by fiberoptic bronchoscopy. After good separation was assured, the lung compliance was measured in each lung separately and both lungs together was measured (Table 1). Compliance was determined by measuring the pressure with a preset tidal volume during the course of ventilation, and before and after the lavage procedure.

After both lungs were ventilated 10 min with 100% O₂, the right lung was clamped for 5 min to allow oxygen absorption, with the left lung ventilated via the tracheal lumen of the tube. Lavage of the right lung was performed with the patient in a supine position. The right port of the DLT was connected to a tube, one limb of which was connected to a suction bottle on the floor, while the other was connected to a reservoir bag approximately 30 cm above the patient’s midaxillary line (Fig. 1). Warmed normal saline at 37°C was infused into the right lung at about 300 ml/min. Then the fluid was allowed to drain into a suction bottle. During the filling phase, the SpO₂ approached 99-100%, but decreased to 90-91%, and even 85-86%, as the fluid drained. Manual chest percussion and vibration were performed during all cycles of instillation and drainage. The amounts of instillation and drainage fluid were recorded in each cycle. Each cycle took about 20 minutes. After fifteen cycles, the lavage was terminated when the color of the drainage fluid had changed from thick creamy yellow to clear (Fig. 2). The right lung was instilled with 15800 ml of normal saline and 15500 ml of milky fluid was collected. After the lavage, as much fluid as possible was drained followed by thorough suctioning. High-volume ventilation with positive end-expiratory pressure (PEEP) was started until the compliance of the right lung improved. A tidal volume of 850 ml with a peak airway pressure of 28 cmH₂O and a plateau pressure of 22 cmH₂O was applied. As soon as the compliance of the right lung had improved, the patient’s SpO₂ was measured for ten to fifteen minutes with the left lung clamped and right lung ventilated. When the patient’s SpO₂ was sufficient to maintain oxygenation, left lung lavage was started. The duration from the end of right lung lavage to starting left lung lavage was more than one hour.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Intraoperative compliance</th>
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<tr>
<td>Ventilation</td>
<td>Baseline</td>
</tr>
<tr>
<td>Two lung ventilation Tidal volume (850 ml)</td>
<td>37 ml/cmH₂O</td>
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<tr>
<td>Left lung ventilation Tidal volume (530 ml)</td>
<td>27 ml/cmH₂O</td>
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<tr>
<td>Right lung ventilation Tidal volume (530 ml)</td>
<td>25 ml/cmH₂O</td>
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The left lung was instilled with 11500 ml of normal saline and 10500 ml of milky fluid was collected. Transient hypotension (blood pressure 86/55 mmHg) with bradycardia occurred while instilling lavage fluid into the left lung. The blood pressure returned to the normal range after reducing the amount of instilled fluid. Compliance was measured again (Table 1). Then the left lung was re-expended and a single lumen endotracheal tube was placed instead of the DLT. The patient was sent to the intensive care unit for mechanical ventilatory support and was extubated the next morning.

Chest radiography showed a marked decrease in infiltration (Fig. 3) and the arterial blood gas analysis revealed pH 7.380, PaCO₂ 46 mmHg and PaO₂ 109 mmHg breathing 31% O₂ via Venturi mask 24 hr after bilateral lung lavage. Follow-up pulmonary function tests after one month revealed FEV₁ 2.3 L, and FVC 2.6 L. His clinical symptoms such as dyspnea on exertion, productive cough had also improved.

**Discussion**

PAP is a rare lung disease, characterized by accumulation of phospholipoproteinaceous material in the alveolar spaces. This disorder was first described by Rosen et al in 1958[1]. Although the etiology of the disorder is not yet known, the phospholipoproteinaceous material is thought to be surfactant[2], and abnormal accumulation is due to failure of the clearance mechanism rather
than to enhanced formation\(^5\). The most effective treatment for PAP is mechanical removal of the proteinaceous material via WLL. Although there are no randomized controlled studies of this procedure, there is good evidence of efficacy with several studies showing improvement in exercise tolerance, pulmonary function, arterial oxygenation, and shunt fraction\(^4\). The indications for lung lavage include a PaO\(_2\) < 60 mmHg at rest or hypoxemic limitation of normal activity\(^5\). Therapeutic WLL was performed in our patient because of his poor tolerance of daily activity. Therapeutic WLL, first proposed by Ramirez, is performed under topical anesthesia\(^6\). However, general anesthesia can be used to avoid subjecting a conscious, seriously ill patient to the stress and discomfort of pulmonary lavage. More effective control of ventilation is achieved with general anesthesia and skeletal muscle paralysis\(^7,8\).

Complete separation of both lungs with a DLT is absolutely necessary to avoid the serious hazard of spillage during the lavage procedure\(^7\). After insertion of the DLT and checking its position, a volume ventilator that can deliver relatively high inflation pressure is required, because these patients have diseased and noncompliant lungs. Prelavage, a tidal volume of 15 ml/kg is needed for both lungs together and 10 ml/kg for each lung separately for noncompliant lungs\(^9\).

Most clinicians lavage the dependent lung with the patient in the lateral decubitus position (LDP), while others use the supine position\(^9\). The LDP with the lavaged lung dependent minimizes the possibility of accidental spillage of lavage fluid from the dependent lavaged lung to the nondependent ventilated lung. However, during periods of lavage fluid drainage, pulmonary blood flow, which is gravity-dependent, would preferentially perfuse the nonventilated dependent lung, and a right-to-left transpulmonary shunt would be maximal\(^9\). The LDP with the lavaged lung nondependent minimizes blood flow to the nonventilated lung, but, on the other hand, increases the possibility of accidental spillage of lavage fluid from the lavaged lung to the dependent ventilated lung. As a compromise, the supine position is used to balance the risk of aspiration with the risk of hypoxemia. In our patient, we preferred the supine position for this reason.

Both lungs are ventilated with 100% oxygen for 10-15 min to wash out nitrogen. After that, one lung is excluded from the ventilatory circuit for another 10 to 15 min to allow absorption of oxygen, and then the collapsed lung is lavaged with isotonic saline at 37°C. The usual filling volume is 500-1000 ml. A nearly identical volume is then allowed to drain by gravity or suction into a bottle with the assistance of mechanical chest percussion. The filling and draining procedure are then repeated.
until the effluent becomes completely clear, usually after a lavage of 10-40 liters. After the effluent lavage fluid become clear, the procedure is terminated. The recovery procedure consists of repetitive periods of large tidal ventilations, vigorous suctioning and chest wall percussion to the lavage side and conventional two lung ventilation with PEEP. Immediately after the procedure, a chest radiograph should be obtained to rule out hydropneumothorax. Symptomatic improvement usually takes place within 24-48 hours. The other lung is lavaged 3-7 days after the first. Usually the most affected lung is lavaged first. However, repeat placement of a DLT may lead to endotracheal granuloma and stenosis. Difficulty in positioning the DLT due to tracheal stenosis may block lavage of the other lung. In our case, we decided to do bilateral WLL at the same time. However, there is a risk of hypoxemia because the compliance of the lavaged lung is reduced immediately after lavage. Before starting left lung lavage, we applied high-volume ventilation with PEEP to the right lung until the compliance had improved.

The major adverse effect of WLL is hypoxemia. Arterial oxygenation improves during the filling phase due to the increase in airway pressure and shunting of blood to the contralateral ventilated lung. An emptying the lung causes a decrease in airway pressure, and perfusion of the surfactant-filled alveoli creates a shunt in the lung and hence a fall in the PaO₂, so this phase should be as short as possible. Lavage of the lung also acts to remove a large amount of surfactant, leading to a stiff noncompliant lung. Ventilation for a few hours with PEEP until surfactant is replenished is mandatory to avoid respiratory failure secondary to atelectasis. In our cases, the lowest SpO₂ measurement was 85% during empty phase and it increased up to 99-100% during filling phase. The intervals between the end of the emptying phase and the next filling phase were as short as possible.

Hypotension can occur during lavage but does not appear to be a common problem. Invasive monitoring is unnecessary in most cases. Rogers et al. found that changes in cardiac output may be more related to anesthetic factors than the phase of lavage. Harrison et al. reported a case of hypotension associated with a mediastinum shift when a large volume was instilled into the lung. Other factors like hypovolemia and poor cardiac function should be noted before lavage. In our case, transient hypotension with bradycardia occurred while instilling lavage fluid into the left lung. The blood pressure returned to the normal range after the amount of instilled fluid was reduced.

PAP is a treatable disease with an excellent prognosis after adequate WLL. The technology, however, is complex and not widely available. It requires general anesthesia and an anesthesiologist who is experienced in placing the DLT and capable of frequently checking and adjusting the tube during the lavage procedure. Leakage of lavage fluid into the contralateral ventilated lung must be avoided. An experienced team is required in the operating room. A postoperative care facility is also needed.

References

5. Rogers RM, Levvin DC, Gray BA, et al.


肺泡蛋白質沉積症進行全肺部沖洗術之病例報告

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肺泡蛋白質沉積症是非常罕見的疾病，病理組織呈現肺泡中充滿蛋白質類的物質，到目前病理機轉
並不清楚。因為是未知病理機轉，對於這個疾病最有效也是最正確的治療方式則是全肺部沖洗術。治療
性全肺部沖洗術必須經由全身麻醉及置放雙口徑氣管內管，而這個治療方法會碰到問題則是低血氧，心
肺循環問題和置放雙口徑氣管內管技術問題。因此必須要有經驗的麻醉專科醫師實行，手術後也需要加
護病房照顧避免發生病發症。在此我們報告一位44歲男性，因吸入柴油，合併呼吸困難，咳嗽有痰，發
燒，診斷為肺泡蛋白質沉積症。在全身麻醉下病患接受全肺部沖洗術。沖洗後臨床症狀不但改善，肺功
能也明顯好轉。

關鍵詞：肺泡蛋白質沉積症，全肺部沖洗術，單側肺部換氣

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