Case Report

Removal of lung lavage fluid during whole-lung lavage using biphasic cuirass ventilation chest percussion in a patient with autoimmune pulmonary alveolar proteinosis

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ABSTRACT

Autoimmune pulmonary alveolar proteinosis (PAP) is a rare lung disease. Although recombinant human granulocyte macrophage colony-stimulating factor (GM-CSF) therapy has emerged as a new therapeutic modality, whole-lung lavage (WLL) with manual chest percussion has been a standard therapy in advanced cases. The application of biphasic cuirass ventilation (BCV) instead of chest percussion has rarely been reported. We describe the case of a patient with advanced PAP who recovered well in both lungs, without complication, after we performed WLL with BCV under anesthetic mechanical ventilation. Both radiographical chest findings and clinical symptoms were improved, and oxygen therapy was finally withdrawn. This case illustrates that BCV can enhance the effective removal of lavage fluid and is an alternative to manual percussion.

KEY WORDS: Biphasic cuirass ventilation, chest percussion, pulmonary alveolar proteinosis, whole-lung lavage

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INTRODUCTION

Pulmonary alveolar proteinosis (PAP) is a rare disease, characterized by accumulation of lipid-rich protein in the alveoli, causing respiratory failure.[1] Whole-lung lavage (WLL), used to clear the alveolar protein accumulation, is currently the standard treatment option for PAP.[2,3] However, some controversies remain about WLL procedures in terms of patient position, type of lavage fluid used, and chest percussion during this procedure.[4,5] In general, chest percussion, including manual or mechanical percussion, is considered useful for the removal of lung lavage fluid.[6] However, very few reports have addressed the use of biphasic cuirass ventilation (BCV) as a mechanical percussion method.[7] Here, we describe a case of autoimmune PAP treated with WLL in conjunction with BCV to enhance the effective removal of the lung lavage fluid.

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CASE REPORT

A 38-year-old Japanese man was referred to our hospital for productive cough and dyspnea on exertion that had gotten worse over the course of 1 week. The patient had a medical history of appendicitis and no family history of noteworthy medical conditions. In addition, he frequently engaged in house-painting and had been smoking one pack of cigarettes a day from 20 years. The patient's temperature was 37.6°C, his blood pressure was 176/114 mmHg, his pulse rate was 95/min, his respiratory rate was 20/min, and his SpO_2 was 88% at room air. Inspiratory crackles in bilateral lung fields and clubbing of the toes were observed.

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The laboratory data showed that elevated aspartate aminotransferase, alanine aminotransferase, and lactate dehydrogenase levels (80 IU/L, 52 IU/L, and 497 IU/L, respectively). KL-6 was markedly elevated to 27959 U/ml (normal range: <500 U/ml). Arterial blood gas analysis, under 4 L/min O_2 through nasal cannula, showed a pH of 7.52, PCO_2 of 27 mmHg, and PO_2 of 82 mmHg.

Chest X-ray showed diffused ground-glass opacity, bilaterally, and chest computed tomography (CT) revealed diffuse ground-glass opacity with a crazy-paving pattern [Figure 1a].

Bronchoscopy yielded bronchial alveolar lavage fluid that was muddy white in appearance, containing periodic acid Schiff-positive material in cytology. Transbronchial lung biopsy showed intra-alveolar eosinophilic mucin accompanying mild interstitial inflammation [Figure 2a]. Serum granulocyte macrophage colony-stimulating factor (GM-CSF) autoantibodies titer was elevated to 20.9 μg/ml (normal range: <1.0 μg/ml). Thus, the patient was diagnosed with autoimmune PAP.

WLL was required for hypoxemia and was performed on two separate occasions. Left lung lavage was performed on the 17th day of postadmission, and right lung lavage was performed on the 31th day. The patient underwent both procedures under general anesthesia, with one-lung ventilation by a double-lumen tube, in the intensive care unit. A lateral decubitus position (on the ventilated lung) was assumed to prevent severe hypoxemia during degassing. One liter of normal saline, warmed to 37°C, was infused into the target lung. BCV (RTX Respirator® Medivent Ltd, London, UK) was performed in clearance mode, at a frequency 300/min, with a negative pressure of 17 cmH2O, to induce vibrations to the chest wall during lavage [Figure 2b]. We managed elevated blood pressure during vibrations by infusing fentanyl and propofol. Lung lavage was repeated until drainage fluid became significantly clearer. Approximately 9520 ml and 15,500 ml of lavage fluid (of the total 9800 ml and 15,000 ml if infused saline) were recovered from the left and right lung, respectively (salvage rate of 97% and 103%, respectively). The patient was extubated on the next day after each WLL procedure.

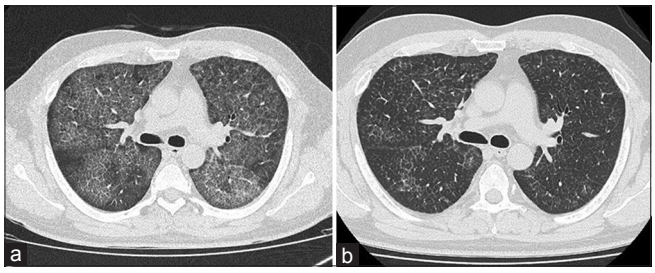

Figure 1: (a) A chest computed tomography scan upon admission revealed crazy-paving pattern. (b) A chest computed tomography showed diffuse ground-glass opacity was improved 4 months following procedure

There were no complications after either procedure. Dyspnea on exertion and productive cough were improved, and the patient was discharged on the day 40th postadmission, with home oxygen therapy. At a 4-month follow-up, the patient's O_2 saturation and PaO_2 improved to 96% and 71 mmHg under room air, respectively. Chest CT scans revealed improvements in diffuse ground-glass opacity [Figure 1b]. The patient could be successfully weaned from oxygen therapy.

DISCUSSION

The clinical course of PAP ranges from asymptomatic to progressive respiratory failure resulting in death.[8,9] Inoue *et al*. categorized the severity of PAP, according to PaO_2 or SpO_2 on room air.[10] The current indications of WLL are hypoxemia, for example, PaO_2 <70 mmHg or A-aDO2 >40 mmHg,[8] impaired lung function, and worsening radiographic findings.[4] We performed WLL due to hypoxemia. We should reflect on that introducing and setting up all WLL procedure-related equipment in the hospital took time due to rarity of PAP.

WLL requires an average of 15.4 ± 6.8 L normal saline/lung, and it is preferable to reduce the residual fluid in the lungs as much as possible.[4] Chest percussion, usually performed during each lavage cycle to facilitate emulsification of lipid-rich proteins in alveoli, can potentially help to minimize residual fluid following WLL. An analysis of bronchopulmonary lavage fluid optical density concluded that manual chest percussion is better than mechanical or no chest percussion.[6] However, to our knowledge, no studies to date have investigated the effects of manual or mechanical chest percussion on the rate and amount of lavage fluid removed after WLL although it has been reported that skin soreness was a complication of manual chest percussion.[5] As we have no experienced physiotherapist, we chose mechanical chest percussion.

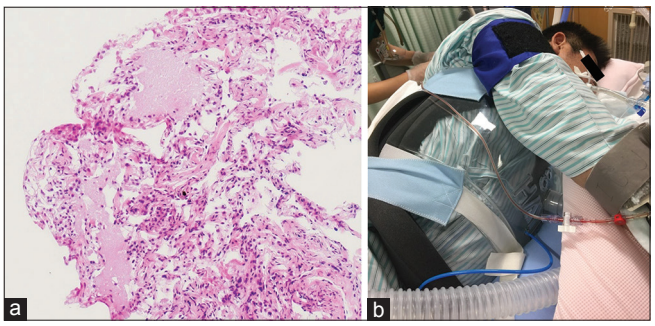

Figure 2: (a) Photomicrograph of the specimen obtained by transbronchial lung biopsy. Alveoli were filled with eosinophilic mucins accompanying mild interstitial inflammation while lung structure was preserved (hematoxylin and eosin, low-power field). (b) Patient fitted with biphasic cuirass ventilation (RTX Respirator® Medivent Ltd, London, UK) device during whole-lung lavage. Intubation was performed by double lumen tube for one-lung ventilation. Lateral decubitus position was assumed on the ventilated lung

We used a BCV to achieve mechanical chest percussion during WLL. BCV is a noninvasive respiratory support system involving a cuirass attached to the body. BCV supports ventilation at negative pressure in the inspiratory phase and positive pressure in the expiratory phase. BCV also has a clearance mode that induces mucus clearance by transmitting high-frequency vibrations to the chest wall, which facilitates mucus removal from the peripheral airway to central airway.[11] Several reports state that BCV had mitigated respiratory failure by removing mucus in this manner.[12,13] Therefore, BCV is sometimes used in Japan to remove mucus during WLL although its effectiveness has rarely been evaluated. Only Kato *et al*. reported that, in a case of PAP, that a combination of BCV, and the use of saline mixed with N-acetyl-L-cysteine was effective for removing surplus fluid.[7] As the risk of administrating N-acetyl-cysteine are unknown, we did not use it in the current case. In Kato *et al*.'s case, BCV was set at 600/min and negative pressure 17 cm H_2O.[7] To avoid elevated blood pressure, we set BCV to 300/min and used sedative and analgesic agents. Even with this reduced frequency, almost all lavage fluid was removed from the lungs. It is possible that a frequency of 300/min contributed to this outcome; however, the optimal degree of negative pressure and vibration frequency remains unknown. To evaluate its use in WLL, it will be necessary to monitor parameters such as the lavage fluid removal rate or the opacity of the drained fluid.

Khan *et al*. reported good clinical responses to some autoimmune PAP cases using recombinant human GM-CSF through the inhalation route.[14] We did not use this as the first choice, as WLL has been the standard therapy and human GM-CSF is at a clinical trial stage and is expensive without health insurance coverage in Japan. Should this patient relapse, human GM-CSF treatment would be the next step.

CONCLUSION

This case illustrates that BCV may be useful as a mechanical percussion instrument, for removing lavage fluid during WLL in patients with PAP. Although our findings help elucidate the potential benefits of BCV during WLL, further research is necessary to understand the implications of chest percussion during WLL in patients with PAP more fully.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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