

## Treatment of a Primary Pulmonary Alveolar Proteinosis Case With Severe Hypoxaemia by Using Segmental Lavage Technique

Pulmonary alveolar proteinosis (PAP) is a rare disease characterised by the accumulation of protein-rich liquid in the alveoli. Even though its pathogenesis is unknown, it is suggested that the accumulation of phospholipids-rich proteinaceous material, positively painted with periodic acid-Schiff (PAS), in the alveoli results from degeneration of the maturation or functions of alveolar macrophages and defects in the functions of granulocyte-macrophage-colony-stimulating factor (GM-CSF). Primary PAP is frequently seen in young males without preceding diseases.<sup>1</sup>

A 32-year-old male patient was admitted with dyspnoea and non-productive cough. It was understood that his dyspnoea had been worsening for about 1 year and there had been cyanosis on his fingers and lips during his daily activities.

In the physical examination of the case, dyspnoea, tachypnoea, peripheral and central cyanosis were detected. Breath count per minute was 30; rales were heard during inspiration bilaterally in the mid-lower zones, more apparent on the left side.

In the laboratory data, polycythemia [Haemoglobin (Hb): 18.2 gr/dL; Haematocrit (Hct): 52.2%]; in the blood gas analysis (breathing air): pH: 7.42; pCO<sub>2</sub>: 34 mm Hg; pO<sub>2</sub>: 47 mm Hg; HCO<sub>3</sub>: 22.5 mmol/L; oxygen saturation (Sat O<sub>2</sub>): 84% were determined. In the chest radiograph, consolidations were observed in mid-lower zones bilaterally. In high-resolution computed tomography (HRCT) of the patient, "ground-glass opacification" was detected in both lungs (Fig. 1). Bronchoalveolar lavage (BAL) from the right middle lobe and transbronchial biopsy (TBB) from the lateral segment of the right lower lobe was practised.

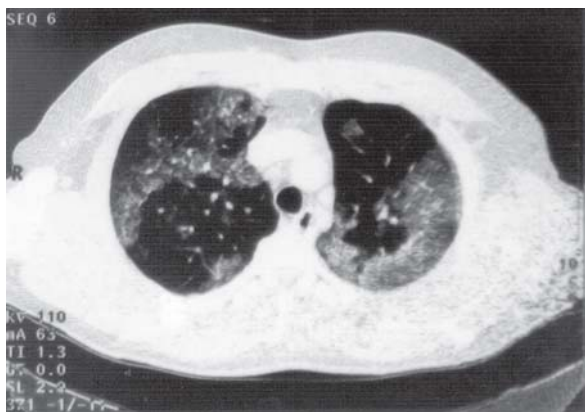


Fig. 1. Ground-glass opacification.

Pathologic result was concordant with PAP. No secondary reason was found in the patient who had been examined for preceding diseases and primary diagnosis was accepted as PAP.

BAL was firstly performed on the left lung of the patient under general anaesthesia. Since Sat O<sub>2</sub> was 80% before the procedure, instead of whole lung lavage (WLL) with double lumen tracheal tube, about 1 L to 1.5 L warmed saline solution in 50-cc injectors was given in and taken out of each segment in the left lung through number 9 tracheal tube in the supine position. In total, 10 L of warmed saline solution was instilled and 8.5 L was aspirated. The procedure was continued for about 4 hours until the lavage, which was milky white at first, turned fully clear. After bronchoscopy, Sat O<sub>2</sub> increased to 90%. After 4 hours of mechanic ventilation support in the intensive care unit, the patient was extubated and transported to the service. Ten days later, BAL was practised on the right lung under the same conditions and with the same methods. Again, by giving 1 L to 1.5 L to each segment, a total of 12 L was given and about 10.5 L was aspirated and the BAL procedure was completed in 3 hours. The patient, who needed less mechanic ventilation after the second bronchoscopy, had less breathing difficulty and cyanosis. The following were the laboratory data after the treatment; Hb: 16.1 gr/dL; Hct: 46%; in blood gas (breathing air): pH: 7.41; pCO<sub>2</sub>: 39.2 mmHg; pO<sub>2</sub>: 73 mm Hg; HCO<sub>3</sub>: 25 mmol/L; Sat O<sub>2</sub>: 94.7%. Improvement was observed in the chest radiography. In 1 year, his chest radiograph turned to normal and he had no dyspnoea even during exercise. He needed no further lavage but is still undergoing follow-up.

Primary pulmonary proteinosis has no specific treatment but improvement in pulmonary functions can be obtained with BAL.<sup>2</sup> This technique is practised in the form of BAL under general anaesthesia, with double lumen tracheal tube and in each time with about 1 L and in total, 10 L to 20 L of warmed saline solution. WLL has been shown to be successful in other studies.<sup>3</sup> However, in a study in Turkey,<sup>4</sup> in spite of clinical improvements, a whole radiological regression could not be obtained in a case where WLL was applied using double lumen tracheal tube under general anaesthesia. However, WLL has complications such as endotracheal granuloma, stenosis, pleural effusion, hydropneumothorax and empyema. Also, severe hypoxaemia restricts the applicability of the procedure. Thus, in our case, instead of WLL with double lumen

tracheal tube, about 1 L to 1.5 L of warmed saline solution in 50-cc injectors was given in and taken out of each segment with number 9 tracheal tube under general anaesthesia. In total, 22 L BAL was practised first on the left and, in a second session, on the right lung. The patient's general condition, blood gases and chest radiography improved dramatically.

Even though lobar and segmental lavage technique with fiber optic bronchoscopy (FOB) was defined before each session, it was applied to radiologically dominant lobe or segment; and not the whole lung.<sup>5</sup> Cheng et al<sup>5</sup> practised BAL on 3 cases with FOB with the support of local anaesthesia and nasal cannula. They preferred the lobes with the most serious changes in HRCT. Warm saline solution was given and taken back in 50-mL injectors. In each procedure, lavage was applied to 2 lobes in the same side. In about 2 hours, approximately 2 L was given. After 2 to 4 days, it was applied to the other side. The procedure was carried out 2 to 5 times. In 2 out of 3 patients with mild hypoxia, positive results were obtained. Hypoxaemia occurred during lavage, especially when the patient experienced a severe cough.

In this case, we applied segmental lavage technique to all lobes and segments in 1 hemithorax at a single session without any complication. We wished to discuss our severely hypoxic case from which a cure was obtained with segmental lavage technique, which was completed with FOB, yet without using double lumen tracheal tube as it would be a safer treatment compared with WLL.

Practising 10 L to 12 L BAL by giving to all lobes and segments in 1 lung at a single session in 50-cc injectors is

a different technique from those conducted so far. A very successful outcome was obtained in this severely hypoxic case without any complication. Therefore, we concluded that our method should be taken into consideration in the treatment of PAP.

#### REFERENCES

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