Respiratory Failure Due to Pulmonary Alveolar Microlithiasis: A Case Report

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Abstract

We presented a case of pulmonary alveolar microlithiasis (PAM) observed in the respiratory intensive care unit (RICU). PAM is a rare disease, characterized by the presence of calcified concretions in the alveolar spaces. Any respiratory infection may cause severe deterioration in the physiological condition, which can be evaluated as acute respiratory distress syndrome (ARDS). PAM was thought to be complicated by ARDS in this case and lung protective ventilation including low tidal volume and positive end-expiratory pressure (PEEP) was performed. We concluded that lung protective ventilation strategy is successful in PAM cases with ARDS.

Turkish Respiratory Journal, 2005;6:(2):105-108

Keywords: *pulmonary alveolar microlithiasis, ARDS, PEEP, lung protective ventilation*

Introduction

Pulmonary alveolar microlithiasis (PAM) is a rare, autosomal recessive chronic lung disease characterized by widespread deposition of intraalveolar calcospherites (1-3). Until the end of 2001, 424 cases of PAM were reported from all over the world, of which 225 were from Turkey (4,5).

Suggested possible etiologies for the disorder include an inherited metabolic abnormality in the lung involving the enzyme carbonic anhydrase, abnormalities in calcium and phosphorus metabolism, abnormalities in the immune system or in the anatomy and physiology of the lung (2,6).

Chest x-ray, high-resolution computed tomography, computed tomography, bronchoalveolar lavage, bone scintigraphy and transbronchial biopsy, all reveal characteristic patterns for PAM (1,7-12). The clinical condition of PAM patients goes through a slow and progressive deterioration resulting in respiratory failure at the end stage (7,13,14). Interstitial fibrosis can develop secondary to PAM, and is associated with poor prognosis (2,15-17).

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There is no specific treatment for alveolar microlithiasis. Cases of PAM should be observed carefully for treatable complications. There may be some fatal outcomes for most patients due to the complications produced by microlithiasis (2).

In spite of the fact that PAM is a rare disorder, it is observed quite frequently in Turkey. Also, we have not come across any reports of cases of PAM complicated by acute respiratory distress syndrome (ARDS). The aim of this report is to evaluate a PAM case with ARDS.

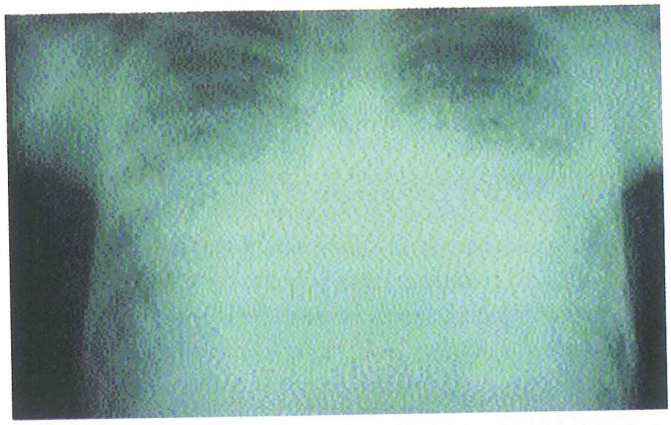

Figure 1. *Chest x-ray showing typical appearance of PAM. Bilateral extensive microcalcifereses mask pulmonary shadows and diaphragmatic borders. Fine calcifications can be seen in the upper zone of both lungs.*

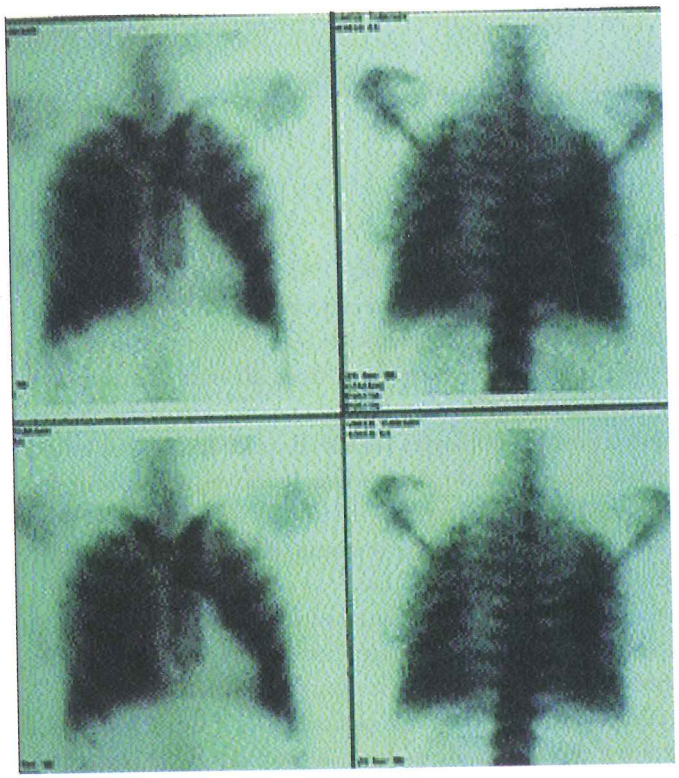

Figure 2. *Total body bone scintigraphy. Tc 99m accumulation in both lungs is typical for PAM diagnosis.*

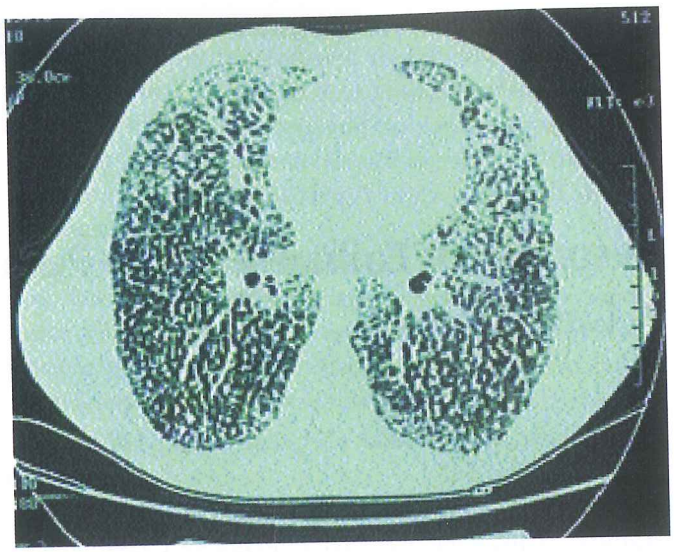

Figure 3. *Calcospherites in lung with high resolution of computed tomography*

Case Report

A 23-year-old male was admitted to the Emergency Unit with dyspnea.

The patient had presented to another hospital, two years ago, during his military service, with a history of exercise dyspnea. PAM was diagnosed on the basis of chest x-ray, 99m Technetium scanning (Figure 2) and high resolution computed tomography (Figure 3). One of his sisters had also been diagnosed as having PAM.

At this admission, the patient gave a history of progressively increasing cough and sputum. In the physical examination, peripheral cyanosis and finger clubbing were observed. Body temperature was 37.2°C. Arterial blood pressure was 110/70 mmHg, heart rate 125 min^{-1}, respiratory rate (RR) 25 min^{-1}, white blood cell count (WBC): 11 400/mm^3 and hematocrit 56%. Glasgow coma score was 15 at admission. Arterial blood gas analysis results were: pH: 7.415, PaO_2: 37 mmHg, $PaCO_2$: 33 mmHg and SaO_2: 72% while he was breathing O_2 6 l min^{-1} via nasal cannula. Chest x-ray findings were typical for PAM (Figure 1).

Results of the patient's pulmonary function tests performed two years ago and at this admission are presented in Table 1. The patient was transferred to the respiratory intensive care unit (RICU) and mechanical ventilation (MV) was initiated. PaO_2/FiO_2 ratio was 100, alveolar-arterial O_2 gradient was 270 and FiO_2 was 0.60. He was evaluated as ARDS and a lung protective ventilation strategy containing low tidal volume and PEEP was initiated. Acute Physiology, Age and Chronic Health Evaluation (APACHE) II score was calculated as 15 after his first day in RICU. Central venous pressure (CVP) was 12 mmHg, right ventricular pressure was 66/17 (mean 38) mmHg, pulmonary artery pressure (PAP) was 71/43 (mean 56) mmHg and pulmonary capillary wedge pressure (PCWP) was measured as 15 mmHg.

Table I. Results of pulmonary function tests		
Pulmonary function tests	At diagnosis of PAM	At present admission
FVC	2.48 l (55%)	1.75 l (36%)
FEV_1	1.98 l (51%)	1.50 l (37%)
FEV_1/FVC	80%	86%
DL_{CO}	18.6 ml min^{-1} $mmHg^{-1}$ (68%)	2.1 ml min^{-1} $mmHg^{-1}$ (19%)
DL_{CO}/VA	5.09 ml min^{-1} $mmHg^{-1}$ (120%)	0.70 ml min^{-1} $mmHg^{-1}$ (41%)
FVC: Forced vital capacity, FEV_1: Forced expiratory volume in 1 second, DL_{CO}: Diffusing capacity measured with carbon monoxide, VA: Alveolar volume		

The patient was mechanically ventilated with FiO_2 of 0.40, tidal volume of 300 ml. PEEP was approximately 10 cmH_2O and RR was 16 min^{-1} for 20 days in RICU. We tried to maintain the end-inspiratory plateau pressure below 35 cmH_2O.

There was no growth in the culture of the purulent material aspirated via the endotracheal tube. Intravenous ampicillin-sulbactam 1 g three times a day was administered empirically. The patient's condition improved and the secretions became clear. On the seventh day of the mechanical ventilation body temperature was 38.5°C, leucocytosis (18 000/mm^3) and purulent sputum recurred. The condition was evaluated as ventilator induced pneumonia. The antibiotic regimen was changed to intravenous imipenem 500 mg four times a day and amicasin 1 g once a day. Following this treatment, the patient's condition improved. However, he experienced another attack with fever and purulent sputum on the 21st day of ventilation. He got a treatment of intravenous ciprofloksasin 400 mg twice a day and teicoplanin 300 mg once a day. It was not possible to wean the patient from mechanical ventilation. On the 26th day, a tracheotomy cannula was inserted.

The case was weaned on the 33rd day. The arterial blood gas analysis results at this time were as follows; pH: 7.40, PCO_2: 33 mmHg, PO_2: 88 mmHg and SaO_2 93% with O_2 6 l min^{-1} nasal O_2. CVP: 6 mmHg, right ventricular pressure: 61/28 (mean: 41) mmHg, PAP: 64/36 (mean: 32) mmHg, PCWP was 5 mmHg.

The tracheotomy cannula was removed on the following day and the patient was transferred to the ward.

Discussion

The chest x-ray has characteristic features in PAM. The radiological appearance and clinical course of the disease are relatively stable for several months and even years (11,18). However, any respiratory infection may cause severe deterioration of the physiological condition, which can be evaluated as ARDS. Several definitions of ARDS are given by different authors (19,20). ARDS can develop in a patient acutely after a precipitating event and previous chest x-rays are usually normal. However, ARDS may develop in a case that has a chronic disease such as PAM or Wegener's granulomatosis with specific pulmonary involvement.

The case presented here had criteria very compatible with ARDS criteria drawn by the American-European Consensus Conference in 1994. The American-European Consensus Conference members agreed that the diagnostic criteria for ARDS should include a) acute onset, b) bilateral chest radiographic infiltrates, c) a pulmonary artery occlusion pressure of ≤18 mmHg, or no evidence of left atrial hypertension; and d) impaired oxygenation regardless of the PEEP concentration, with a PaO_2/FiO_2 ratio ≤200 (21). According to this consensus criteria, our case of PAM was complicated by ARDS. The patient conformed to all ARDS criteria such as acute onset, bilateral pulmonary infiltrations and PCWP of 15 mmHg. In addition, PaO_2/FiO_2 ratio was 100 at admission. We believe that radiological features were not helpful for the diagnosis since the patient had already radiological signs of PAM at the onset of the ARDS.

The term ARDS, which denotes a condition rather than a disease with defined specific criteria, is mainly based on the gas exchange status of a patient. The radiological signs of PAM and ARDS may be confused. Bilateral infiltrations and cardiac shadows within normal limits are characteristic features of both conditions. Blood gas analyses and PCWP value are more important for the diagnosis and management of such cases. We managed this case as a case of ARDS, with lung protective ventilation strategy due to evidence of a history of PAM. The patient recovered with low tidal volume and PEEP application on mechanical ventilation.

The clinical spectrum of PAM ranges from a completely asymptomatic state to a state of progressive dyspnea, finger clubbing, prominent pulmonary arteries and diminished carbon monoxide diffusing capacity (11,15). Our case presents a severe course of PAM which had progressed to a very advanced stage at the time of admission. Deposition of intraalveolar calcospherites, and the fibrotic response to these deposits determine the course of the disease. Deteriorating gas exchange is the hallmark of progressive pulmonary fibrosis and the major determinant of development of cor pulmonale (17).

In the relevant literature, we have not encountered any case of PAM who had been mechanically ventilated except one patient who had been treated with nasal continuous positive airway pressure (nCPAP). In that report, Freiberg et al presented a 37-year-old man with respiratory failure and cor pulmonale. The diagnosis of PAM was made at age 20. These authors reported that nCPAP reduced the intrapulmonary shunt and allowed for correction of hypoxemia with a smaller oxygen flow rate (17).

Diffuse alveolar damage, rapid edema, atelectasis, inflammation, pulmonary fibrosis, capillary obliteration occur in patients with ARDS, followed by development of pulmonary hypertension. Severe hypoxemia is a hallmark for ARDS caused by the presence of an intrapulmonary shunt, or areas of very low ratios of ventilation/perfusion. This shunting is presumably due to edema and atelectasis. Asbaugh and associates originally reported that mechanical ventilation and PEEP improved gas exchange in ARDS patients. Later they also reported that PEEP prevented or reversed alveolar collapse by increasing the functional residual capacity (19). Lung protective ventilation strategy is the first therapy found to improve outcome in ARDS (22).

At an International Consensus Conference on Mechanical Ventilation it has been suggested that in patients with ARDS, plateau pressure during mechanical ventilation should ideally be maintained at less than 35 cmH_2O by decreasing tidal volume to as low as 5 ml kg^{-1} if necessary to avoid alveolar over distension (23).

We also concluded that lung protective ventilation strategy containing low tidal volume and PEEP administration was successful, as shown in our case with ARDS due to PAM.

Acknowledgement: The authors thank the Department of Chest Disease and Tuberculosis of Gülhane Military Medical Academy for the provision of previous data.

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