

Pulmonary Alveolar Proteinosis and Tracheal Stenosis after Exposure to Fire and Fire Extinguisher

Yangın ve Yangın Söndürücü Maruziyeti Sonrasında Gelişen Pulmoner Alveolar Proteinozis ve Trakeal Stenoz

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Abstract

Pulmonary alveolar proteinosis (PAP) is a rare disease characterized by the accumulation of acellular surfactant lipids and proteins within the alveoli. Idiopathic PAP accounts for about 90% of all patients. Secondary PAP factors include insecticides, inorganic dusts such as silica, hematological malignancies, Human Immunodeficiency Virus infection, leflunomide, toxic gas inhalation, various industrial exposures to aluminum- and metal oxide-containing dusts and clinical conditions. Herein, we present a case of pulmonary alveolar proteinosis and tracheal stenosis secondary to exposure to fire and fire extinguisher.

Key words: Pulmonary Alveolar Proteinosis, Fire extinguishing systems, Smoke Inhalation Injury.

Özet

Pulmoner alveoler proteinozis (PAP) nadir bir hastalıktır. PAP alveollere aselüler sürfaktan lipidlerin ve proteinlerin birikimi ile karakterizedir. İdiyopatik PAP bütün PAP olgularının %90'ını oluşturur. Sekonder PAP ise insektisitlerle, silika gibi inorganik tozlara, hematolojik malignitelere, insan immün yetmezlik virüs enfeksiyonuna, leflunomide, toksik gaz inhalasyonuna, alüminyum ve metal oksit içeren endüstriyel tozlara sekonder gelişebilir. Biz de yangın ve yangın söndürücü maruziyeti sonrası gelişen pulmoner alveolar proteinozis ve trakeal stenoz olgusu sunduk.

Anahtar Sözcükler: Pulmoner alveoler proteinozis, Yangın söndürme sistemleri, Duman inhalasyon hasarı.

Pulmonary alveolar proteinosis (PAP) was first described in 1958 (1). With an unknown etiology, it is a rare pulmonary disease characterized by the accumulation of acellular surfactant lipids and proteins within the alveoli. Irregularity in the clearance of surfactants from the alveoli due to impaired macrophage functions has been implicated in the pathophysiology of the disease (2). Pulmonary gas exchange is altered due to the accumulated material in the alveoli; shortness of breath develops; and defense mechanism of the lung is weakened. It is typically divided into three groups; congenital, idiopathic, and secondary. The etiolo-

gy of secondary PAP has been reported to be related with insecticides, inorganic dusts such as silica, hematological malignancies, HIV infection, and leflunomide (3).

While hydrofluorocarbon (HFC) used in the fire extinguisher is stable under normal conditions, it decomposes under high temperature, liberating the hydrofluoric acid (HFA). Kim et al. (4) reported describes PAP which developed after chronic, repeated exposure to fire extinguisher spray. The present paper also reports a similar case who developed PAP after being exposed to fire.

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CASE

A 41-year-old female patient was referred to us for the evaluation and treatment of airway obstruction. Her history revealed that she had no complaints until three months ago, when a fire started in the hotel she was working for. She had skin burn during this fire and she was also exposed to the smoke and fire extinguisher powder in the short term. The fire extinguisher system was the fire sprinkler system which contained HFC. She was treated in the intensive care unit for 40 days. Tracheostomy was performed because of shortness of breath and stridor on day 20 of the intensive care unit stay, and, then, she was intubated at a mechanic ventilator for a day. Reproduction did not occur in the bronchial lavage culture. Physical examination showed that she had second and third-degree burns on the 1/4 of the proximal back, shoulders, and upper extremities in a patchy pattern, and there were keloid tissues in these regions. She was tracheostomized, conscious, oriented, and cooperative with a spontaneous respiration. Auscultation revealed inspiratory rales in bilateral middle and lower zones. Other systems examinations were unremarkable. Her blood pressure: 94/74 mmHg; pulse rate, 75 bpm; and oxygen saturation from the finger tip: 99 % (with 3 L/min oxygen). Arterial blood gas analysis showed pH: 7.421, pO₂: 58.4 mmHg, and pCO₂: 40.6 mmHg (with 2 L/min oxygen). Chest X-ray showed a non-homogeneously increased density, more remarkably in the bilateral lower zones and middle right zone (Figure 1). A complete blood count analysis was normal, except Hb (10.4 g/dL). Biochemistry analysis results were normal, except LDH: 300 IU/L, C reactive protein (CRP) 63 mg/L, total protein: 4.9 g/dL, and albumin 2.8 g/dL. The indicators of hepatitis and collagen tissue were negative. Colistin-only-sensitive *Pseudomonas aeruginosa* was grown in the urine culture. No respiratory function test was performed, since she was tracheostomized. Thoracic computed tomography (CT) showed narrowing and irregularity of the tracheal lumen, bilateral patchy crazy-paving pattern in both lungs despite largely preserved areas (Figure 1). Rigid bronchoscopic examination showed that the lumen of the trachea was 95% obliterated by complex stenosis from the tracheostomy hole down to the carina, and there was a web-like stenosis in the left main bronchus 2 cm after the carina, narrowing the lumen by 80%. The narrowing in the trachea was dilated using a rigid tube. A fully patent lumen was achieved in the trachea, which was followed by cryotherapy in these areas. Repeated bronchoscopy was performed 14 days after the first one. A milk-like fluid was

aspirated by bronchoalveolar lavage (BAL) from the right system. The web-like stenosis on the left was dilated by a scissor and a rigid tube (Figure 2a). A full patency was achieved in the lumen in the left main bronchus. The lavage fluid cytology was non-specific. A periodic acid Schiff staining was performed, which resulted in positive staining (Figure 2b). Based on these findings, the patient was diagnosed with multiple airway stenosis and pulmonary alveolar proteinosis due to the exposure to a fire or fire extinguisher powder. A therapeutic BAL was scheduled upon airway recovery. Unfortunately, the patient died from respiratory failure in the city, where the patient lived at six months of follow-up.

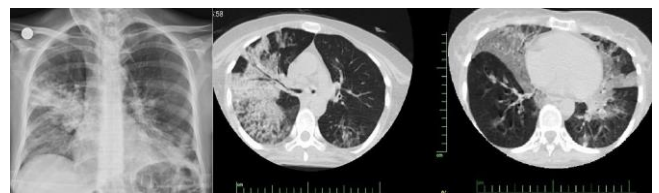


Figure 1: A crazy paving pattern on thoracic CT and chest X-ray

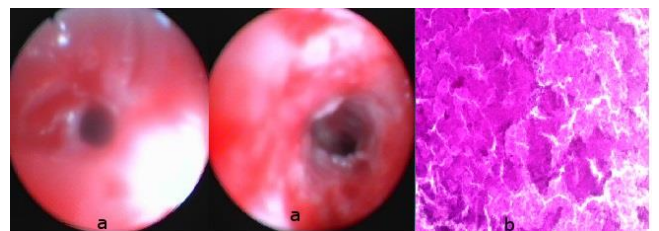


Figure 2a and b: Bronchoscopic view of before and after dilatation (a) and diffuse positivity in periodic acid Schiff staining (b)

DISCUSSION

Pulmonary alveolar proteinosis is a rare pulmonary disease. There is an accumulation of granular, acellular, and periodic acid Schiff-positive material within the alveoli. No interstitial inflammation and fibrosis are expected (5). Three different types of PAP have been described; congenital, primary and secondary. Idiopathic PAP accounts for about 90% of all patients, particularly more common in males and young adults (6).

Although pulmonary alveolar proteinosis is mainly idiopathic, patients with secondary PAP have also been identified. Secondary PAP factors include insecticides, inorganic dusts such as silica, hematological malignancies, HIV infection, leflunomide, toxic gas inhalation, exposure to silica, various industrial exposures to aluminum- and metal oxide-containing dusts and clinical conditions (6, 7). Known as FM 200 or HFC-227 1,1,1,2,3,3,3-heptafluoropropane is a hydrofluorocarbon used in the automatic fire extinguishing systems, and it is, in general, non-toxic under normal conditions (8). However, under

high temperatures, it decomposes to release hydrogen fluoride (9). Zierold et al. (8) reported patients who were exposed to hydrogen fluoride and developed acute respiratory failure under high temperatures. Similarly, Dote et al. (10) reported respiratory problems that were associated with exposure to the same agent. Although PAP case associated with fire was not reported, a case which may be associated with fire extinguisher was presented by Kim et al. (4). The authors also reported that the patient with PAP associated with chronic hydrogen fluoride inhalation (4). In the BAL or serum, anti-GM-CSF antibody measurement is useful for secondary PAP diagnosis and levels of anti-GM-CSF antibodies correlated with the severity of PAP (11). In one patient with PAP related to indium-tin oxide exposure, elevated levels of antibodies to GM-CSF were noted (12).

In the present case, secondary PAP was thought for exposure to fire and fire extinguishers. However, anti-GM-CSF antibody was unable to be investigated for BAL or serum. Our case had no previously known or newly identified concomitant disease, pulmonary infection or dust exposure during the diagnosis and treatment period. The only relevant history was her exposure to smoke and fire extinguishers during the fire. No other etiologic factor was found. Thus, we believe that she represents the second such case reported to date. The patient who was reported by Kim et al. (4) had little amount of exposure and chronic hydrogen fluoride inhalation during eight months. She also had a short-term and intensive exposure. It suggests that short-term exposure may be also associated with PAP. In conclusion, PAP should be considered in the differential diagnosis of respiratory complaints in individuals who were rescued from a fire and in case of exposure to various propellant gases containing hydrofluorocarbon accompanied with high temperatures. A milk-like appearance of the lavage fluid during bronchoscopy provides an important clue for PAP. The treatment of choice for stricture in the respiratory tract is mechanical dilation, while whole lung lavage is the main treatment for pulmonary alveolar proteinosis.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - M.Ç., E.T., K.A., M.A.Ö., E.Ç.; Planning and Design - M.Ç., E.T., K.A., M.A.Ö., E.Ç.; Supervision - M.Ç., E.T., K.A., M.A.Ö., E.Ç.; Funding -; Materials - M.Ç., E.T., M.A.Ö., E.Ç.; Data Collection and/or Pro-

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