Turk Toraks Derg 2014; 15: 33-5 DOI: 10.5152/ttd.2013.37

CASE REPORT / OLGU SUNUMU

Pulmonary Alveolar Microlithiasis and Preterm Delivery: A Case Report

Pulmoner Alveoler Mikrolitiyazis ve Preterm Doğum: Olgu Sunumu

Gülnur Erdem, Aslı Göktan, Fatih Erbay, Tamer Baysal

Department of Radiology, Faculty of Medicine, İnönü University, Malatya, Turkey

Abstract

Pulmonary alveolar microlithiasis (PAM) is a rare, chronic lung disease characterised by extensive intra-alveolar calcium and phosphorus deposition throughout the parenchyma of both lungs. The etiology and pathogenesis of the disease is unclear. In our case, PAM with pregnancy was presented; respiratory distress had been observed during the first and second trimesters. In the following weeks, premature activity developed and the foetus was not able to stay alive. This is the third case of PAM in pregnancy described in the literature. A 36 year-old female was referred to our emergency department complaining that she had suffered from dyspnoea on a couple of occasions since the first trimestry of her last pregnancy. On suspecting a pulmonary embolism, dynamic thorax multidetector computed tomography (MDCT) was performed after the delivery. MDCT revealed typical PAM findings. In PAM, radiological signs are not compatible with clinical symptoms; thus, the clinical symptoms are not as dramatic as imaging findings. However, it should be taken into consideration that this disease, with no effective treatment, may rarely progress into end stage pulmonary disease due to conditions which alter pulmonary functions, such as pregnancy.

KEY WORDS: Pulmonary alveolar microlithiasis, pregnancy, computed tomography

Özet

Pulmoner alveoler mikrolitiyazis (PAM), her iki akciğer parankiminde alveoler yaygın kalsiyum ve fosfor birikimi ile karakterize, nadir görülen bir hastalıktır. Etyoloji ve patogenezi tam olarak bilinmemektedir. Olgumuzda PAM'ın gebelik ile birlikteliği izlenmiş ve hastanın ilk ve ikinci trimesterinde solunum sıkıntısında artış gözlenmiştir. İlerleyen haftalarda durdurulamayan prematür eylem gerçekleşmiş ve fetus yaşatılamamıştır. Bu, literatürde bildirilen 3. PAM ve gebelik olgusudur. Otuz altı yaşında bayan hasta, gebeliğinin ilk trimesterinden itibaren efor dispnesi şikayeti ile birkaç kez acil servisimize başvurmuştur. Doğumdan sonra pulmoner emboli şüphesi ile dinamik toraks çok kesitli bilgisayarlı tomografi (ÇKBT) yapılmıştır. ÇKBT'de tipik PAM bulguları izlenmiştir. PAM'da, radyolojik bulgular klinik bulgular ile uyum göstermemektedir ve semptomlar görüntüleme bulguları kadar dramatik değildir. Bununla birlikte, efektif tedavisi olmayan hastalığın, gebelik benzeri pulmoner fonksiyonları zorlayıcı durumlarda bulgu verdiği ve nadirde olsa son dönem akciğer hastalığına ilerleyebileceği göz önünde bulundurulmalıdır.

ANAHTAR SÖZCÜKLER: Pulmoner alveoler mikrolitiyazis, gebelik, bilgisayarlı tomografi

Received/Geliş Tarihi: *10.01.2013* ***Accepted/Kabul Tarihi:*** *09.05.2013* ***Available Online Date/Çevrimiçi Yayın Tarihi:*** *14.06.2013*

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is a rare, chronic lung disease characterised by extensive intra-alveolar calcium and phosphorus deposition throughout the parenchyma of both lungs. Fifty percent of cases are familial and the disease follows an autosomal recessive model of inheritance. The etiology and pathogenesis of the disease is unclear [1]. Induction of microliths caused by increased and condensated alveolar mucous membranes due to declining mucosilier function is suggested in the pathogenesis [2]. The disease was first described by Harbitz in 1918 as 'extensive calcification of lungs'. The incidence of the disease is particularly high in Turkey (30% of the cases all over the world), Italy and the USA. The majority of cases have reached adulthood at first presentation. However childhood diseases are also reported. Although male predominance is known, the incidence in familial cases is similar in both sexes. A pulmonary alveolar chest X-ray is pathognomonic. Patients have poor or no symptoms though extensive involvement of both lungs [3,4].

Transplantation can be performed in cases with end-stage lung disease. There is no effective medical treatment known [5].

In our case, PAM with pregnancy was presented. Respiratory distress had been observed during the first and second trimesters. In the following weeks, premature delivery activity was developed and foetus was not able to stay alive.

This study was presented at the 30th Annual National Radiology Congress, 4-9 November 2009, Antalya, Turkey.

Address for Correspondence / Yazışma Adresi: Gülnur Erdem, Department of Radiology, Faculty of Medicine, İnönü University, Malatya, Turkey Phone: +90 422 341 06 60 E-mail: gulnur.erdem@inonu.edu.tr

CASE PRESENTATION

A 36 year-old female was referred to our emergency department complaining from increasing dyspnoea which had occurred since the first trimester and had increased gradually through to the third trimester of her pregnancy. The patient had been diagnosed with PAM for five years; her medical history also revealed gestational diabetes mellitus and coronary artery disease for four years. She had no complaint of dyspnoea in her first pregnancy. There was no family inheritance, cough, fever or chest pain in her medical history. Although use of an oxygen concentrator was suggested because of PAM in early periods of her pregnancy, this was refused by the patient.

Since restrictive functional disorder and diminished lung diffusion capacity in pulmonary function tests as well as signs of hypoxy in her arterial blood gases had been detected in the first trimestry follow-up, the patient had been informed about the possibility of worsening of her complaints during the second and third trimestery of her pregnancy. The pregnancy was maintained on the demand of the patient and in the 31st week of gestation, after unstoppable preterm labour resulting in premature delivery with a caesarean section, the live male foetus weighing 1350 g was unable to stay alive. In the patient's final admission to hospital, the arterial blood gas analysis was as follows: pH: 7.34; pCO_2: 31.5 mm Hg; pO_2 32.4 mm Hg; bicarbonate 215 mEq/L; and oxygen saturation: 66.2%.

After parturition, because of worsening of dyspnoea, palpitation and findings of hypoxia, D-dimer and high resolution computed tomography (HRCT) were applied. The haemogram was normal and the D-dimer level was found to be increased (3.79 µg/mL). With pulmonary embolism suspected, a dynamic thorax investigation by 64-row multidetector computed tomography (MDCT) was performed after parturition. Marked calcification areas involving bilateral basal and subpleural regions of both lungs were seen; no thrombosis was detected in the pulmonary arteries on the mediastinal window (Figure 1-3). Since another possible differential diagnosis was infection, antibiotics (cefazoline 1x2 g) were given. On the second day of the antibiotics regimen, clinical symptoms were improved. A confirmed consent form was obtained from the patient for the procedure.

DISCUSSION

Pulmonary alveolar microlithiasis is a rare diffuse lung disease characterised by a slow course and occurs in both familial and sporadic forms. Familial cases predominantly affect females, whereas sporadic cases show a higher prevalence in males. The majority of cases have been reported in Europe. The number of cases which are reported in our country constitute the significant part of all cases reported in the medical literature [3,6].

The disease is mostly seen from birth up to 40 years of age and is usually diagnosed incidentally during radiological investigation of the chest for other reasons [6,7]. In early cases, diagnosis was primarily made at autopsy, whereas nowadays diagnostic investigations are made by radiological imaging (chest radiography, CT), bronchial lavage and transbronchial biopsy [6].

Most cases are asymptomatic at the time of diagnosis. At the late stages of disease, symptoms include a nonproductive cough and dyspnoea may develop [1]. Although the clinical course of the disease varies, it usually progresses slowly. It has three different phases: initial, evolution and stabilisation. While it remains static in some patients, it may progress into pulmonary fibrosis, respiratory failure and cor pulmonale in others [6,7].

The disease is almost always diagnosed through radiological findings [2]. Although the radiological findings are diagnostic, many cases are mistaken for miliary tuberculosis, silicosis, berylliosis, sarcoidosis, haemosiderosis, fungal infections and carcinomatosis [8]. Radiologically, extensive micronodular opacities are seen in the basal and middle regions of both lungs in PAM. This typical X-ray appearance is described as a 'sandstorm lung'. Bullae and blebs can also be detected in the apexes [3]. CT confirms relatively symmetrical abnormalities. The calcifications are prominent in the peripheral, mediastinal and fissural subpleural regions. Each lobe appears surrounded by a fine dense outline like a 'stony lung'. Additionally, findings such as interlobular septal thickening, which can be seen in interstitial lung diseases, ground-glass appearance, peribronchovascular interstitial thickening, and parenchymal bands, may be observed in HRCT examination of the patients [4,6].

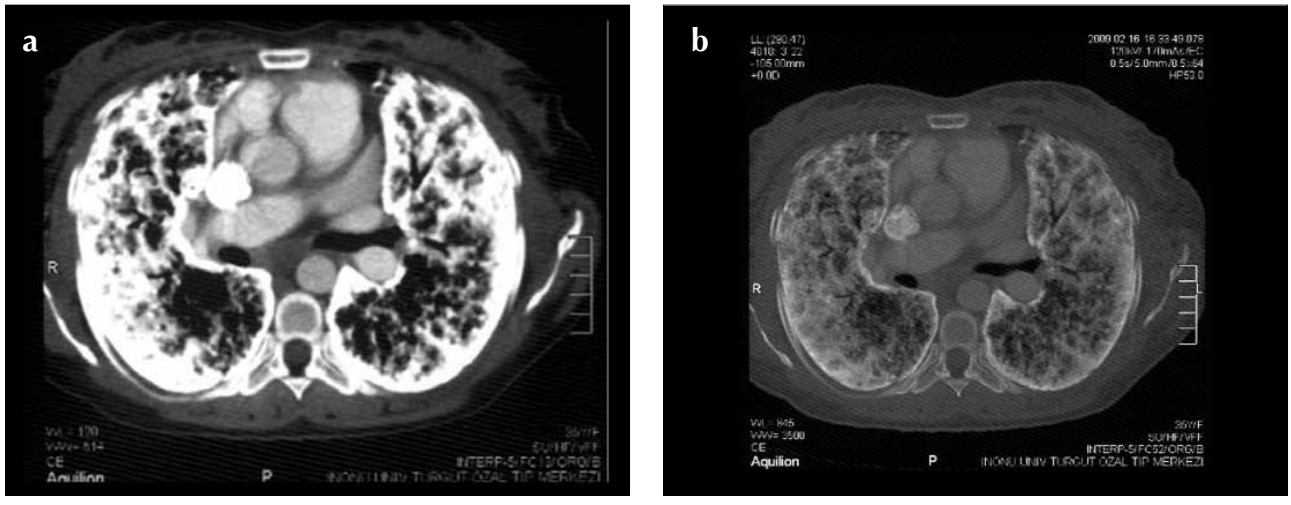

Figure 1. In the slice where main pulmonary arteries exist, diffuse alveolary density causing air bronchograms is seen (a). In the bone window, the density of the lung paranchima is similar to bone tissue (b)

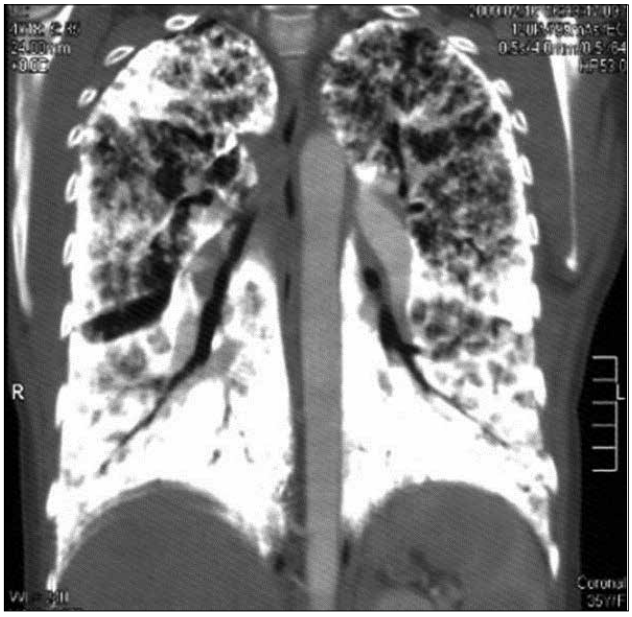

Figure 2. In coronal MDCT slices, microliths are predominantly seen in the basal regions of both lungs

Our case had no symptoms during her first pregnancy in her younger ages. Pulmonary symptoms provoked by pregnancy and advancing age had developed. Worsening of the clinical symptoms were related to diabetes and advanced disease instead of thromboembolism. Restrictive type dysfunction in pulmonary function tests and inspiratory ralles in physical examination had been detected. After an asymptomatic period, an accompanying pregnancy had intensified the symptoms of the disease and the onset of preterm labour resulted in the loss of the premature infant. There have only been two cases of PAM in pregnancy described in the literature. The outcome of both cases was successful and the mothers had healthy newborns by caesarean section [9,10]. In our opinion, preterm delivery in our patient was related to diabetes mellitus and to the advanced stage of the disease.

On HRCT imaging of the case reported here, calcified nodules, septal thickening, pleural and sub-pleural calcification involving mainly bilateral basal and subpleural regions of both lungs were seen. Each hemithorax was surrounded by a fine dense outline. In PAM, radiological signs are not compatible with the clinical symptoms; thus, symptoms are not as dramatic as imaging findings. However, it has to be taken into consideration that this disease, with no effective treatment, may rarely progress into end stage pulmonary disease due to conditions which alter pulmonary function, such as pregnancy.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

Peer-review: Externally peer-reviewed.

Informed Consent: Did not obtained due to urgent admission.

Author Contributions: Concept - G.E., A.G., F.E., T.B.; Design - G.E., A.G., F.E., T.B.; Supervision - G.E., A.G., F.E., T.B.; Data Collection and/or Processing - G.E., A.G.; Analysis and/or Interpretation - G.E, A.G.; Literature Review - G.E., A.G.; Writer - G.E., A.G., F.E., T.B.; Critical Review - G.E., A.G., F.E., T.B.

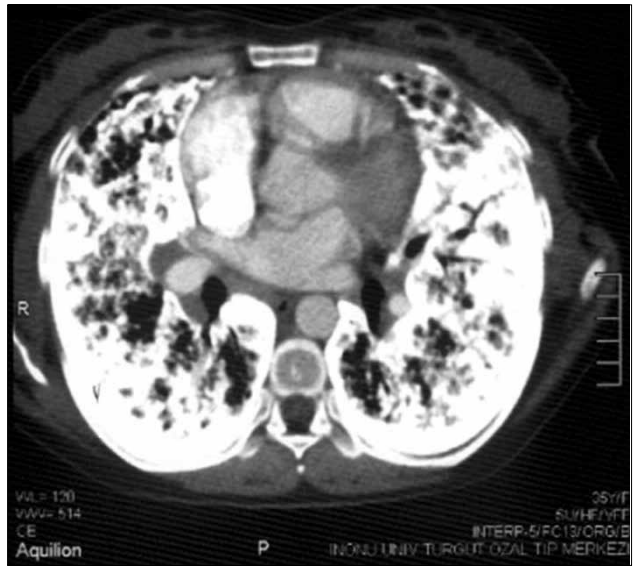

Figure 3. Slice through left atrium level shows density concentrated in the mediastinal and lateral subpleural regions

Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir

Finansal Destek: Yazarlar bu çalışma için finansal destek almadıklarını beyan etmişlerdir.

Hakem değerlendirmesi: Dış bağımsız.

Hasta Onamı: Acil başvuru nedeniyle alınmadı.

Yazar Katkıları: Fikir - G.E., A.G., F.E., T.B.; Tasarım - G.E., A.G., F.E., T.B.; Denetleme - G.E., A.G., F.E., T.B.; Veri toplanması ve/veya işlemesi - G.E., A.G.; Analiz ve/veya yorum - G.E, A.G.; Literatür taraması - G.E., A.G.; Yazıyı yazan - G.E., A.G., F.E., T.B.; Eleştirel İnceleme - G.E., A.G., F.E., T.B.

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