Pulmonary Alveolar Microlithiasis and Preterm Delivery: A Case Report

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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 trimester 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

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 condensed 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.

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 trimester follow-up, the patient had been informed about the
possibility of worsening of her complaints during the second and third trimester of her pregnancy. The pregnancy was main-
tained on the demand of the patient and in the 31st week of
gestation, after unstoppable preterm labour resulting in premi-
ture 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 fol-
lows: pH: 7.34; pCO₂: 31.5 mm Hg; pO₂: 32.4 mm Hg; bicar-
bonate 215 mEq/L; and oxygen saturation: 66.2%.

After parturition, because of worsening of dyspnoea, palpita-
tion and findings of hypoxia, D-dimer and high resolution
computed tomography (HRCT) were applied. The haemo-
gram was normal and the D-dimer level was found to be in-
creased (3.79 µg/mL). With pulmonary embolism suspected,
a dynamic thorax investigation by 64-row multidetector
computed tomography (MDCT) was performed after parturi-
tion. 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 diag-
nosis 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 dis-
ease characterised by a slow course and occurs in both
familial and sporadic forms. Familial cases predominantly
affect females, whereas sporadic cases show a higher preva-
ience in males. The majority of cases have been reported in
Europe. The number of cases which are reported in our coun-
try 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 trans-
bronchial 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, silico-
sis, berylliosis, sarcoidosis, haemorrhagia, fungal infections
and carcinomatosis [8]. Radiologically, extensive micronod-
ular 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 apices [3]. CT confirms relatively symmetrical abnor-
malities. 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 thick-
ening, 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].

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 clin-
cal symptoms were related to diabetes and advanced dis-
ease instead of thromboembolism. Restrictive type dysfunc-
tion in pulmonary function tests and inspiratory rales in
physical examination had been detected. After an asympto-
matic 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 litera-
ture. 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 dis-
ease.
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.

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