COACTION PUBLISHING

Diffuse pulmonary amyloidosis due to Familial Mediterranean Fever, a rare presentation

To the Editor,

Familial Mediterranean Fever (FMF) is a hereditary disease characterized by sporadic, paroxysmal attacks of fever and serosal inflammation described primarily in several ethnic groups originating from Mediterranean region. The most serious complication of FMF is chronic renal failure (CRF) developing due to secondary amyloidosis (1). Clinically evident lung involvement due to FMF associated amyloidosis is extremely rare and occurs in severe cases (2). On the other hand very fine amyloid deposits can be recognized in the peripheral alveolar septa in autopsies of FMF patients (1). Herein we present a case of diffuse pulmonary amyloidosis due to FMF in an asymptomatic patient referred to our clinic with a preliminary diagnosis of miliary tuberculosis.

The patient was a 31-year-old man who had a miliary pattern observed on a routine chest X-ray. He had a diagnosis of FMF since he was 10-years-old, had undergone a renal biopsy revealing amyloidosis when he was 19 and had started hemodialysis when he was 25. Unfortunately he stopped using colchicine at that time. Past medical history was unremarkable for tuberculosis or prior respiratory disease. Physical examination was normal. Laboratory analysis revealed a high erythrocyte sedimentation rate (65 mm/h, range: 0–20 mm/h), anemia (Hb: 9.3 g/dl, range: 12–14 g/dl), impaired renal function (BUN: 33 mg/dl, range: 0–20 mg/dl; creatinine: 5.1 mg/dl, range: 0.6–1.4 mg/dl), and hypoalbuminemia (3.1 g/dl, range: 3.5–5.5 g/dl). p-ANCA, c-ANCA and Anti-GBM antibody were all negative. Three sputum smears and cultures were negative for tuberculosis. Respiratory function tests revealed a restrictive flow pattern and a decreased diffusion capacity. Oxygenation was normal. High resolution computed tomography of the chest demonstrated diffuse reticulo-nodular pattern (Fig. 1). We retrospectively evaluated the prior routine chest x-rays and realized that the same radiological findings were also evident in the chest x-rays obtained within the previous year. We performed transbronchial lung biopsy from the right lower lobe and showed amyloid deposition within the alveolar septal interstitium and the walls of small blood vessels with green birefringence under polarized light after staining with congo-red. Amyloid deposition in the walls of vessels is seen in Fig. 2.

Pulmonary complications of chronic renal failure (CRF) include uremic pulmonary edema, pleural effusion, interstitial fibrosis, and increased risk for certain infections such as tuberculosis (3). Pulmonary amyloidosis is a rare presentation in a patient with CRF due to FMF. In the literature Erdem et al. (4) reported a diffuse pulmonary amyloidosis case mimicking interstitial lung disease. In that case there was an advanced picture of interstitial pulmonary involvement with fibrotic changes in addition to background emphysema. In the presented

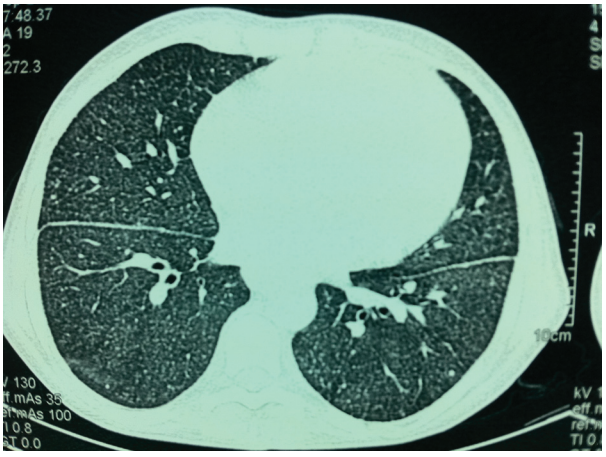

Fig. 1. High resolution computed tomography of the chest demonstrated diffuse reticulo-nodular pattern.

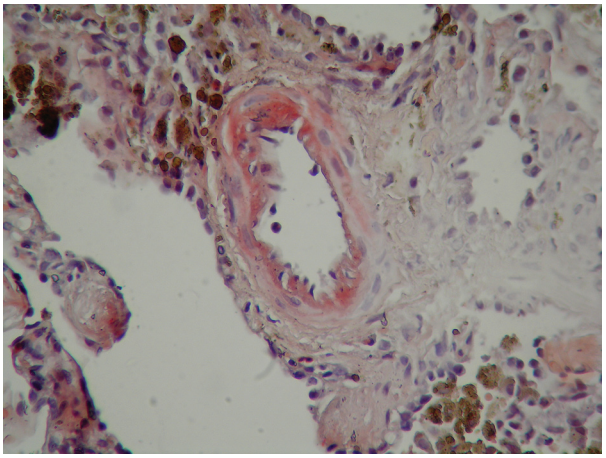

Fig. 2. Amyloid deposition in the walls of vessels (400 × Congo red).

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patient there were prominent diffuse reticulo-nodular pattern due to amyloidosis in a CRF patient due to FMF. Regular colchicine treatment is of great importance in reducing the development or progression of secondary amyloidosis (5). The discontinuation of colchicine therapy for a duration of 6 years might have caused diffuse pulmonary amyloidosis.

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Deniz Koksal, Neslihan Mutluay, Hulya Bayiz, Bahadir Berktas and Mine Berkoglu
Chest Diseases Clinic
Ataturk Chest Diseases and Chest Surgery Education and Research Hospital
Ankara, Turkey
Email: dckoksal@gmail.com

Funda Demirag
Department of Pathology
Ataturk Chest Diseases and Chest Surgery Education and Research Hospital
Ankara, Turkey

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