

# Pulmonary Amyloidosis Mimicking Interstitial Lung Disease in a Patient With Polymyozitis

## İnterstisyel Akciğer Hastalığını Taklit Eden Pulmoner Amiloidoz

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### Summary

Primary pulmonary amyloidosis is a localized type of amyloidosis which usually presents with parenchymal nodules, diffuse alveolar damage, or submucosal deposits in the airways. Polymyozitis is an inflammatory autoimmune myopathy characterized by symmetric weakness of the limb girdle and anterior neck muscles. Nearly 80-90% of cases with polymyozitis are successfully treated with steroid and cyclophosphamide combination therapies. Polymyozitis also cause secondary interstitial lung disease and the radiological findings of these two diseases may not be easily discriminated from each other. *Türk J Phys Med Rehab* 2012;58:159-61.

**Key Words:** Polymyozitis, interstitial lung disease, primary pulmonary amyloidosis

### Özet

Primer pulmoner amiloidoz lokalize tip amiloidozlardan biri olup parankimal nodül, yaygın alveol hasarı veya hava yollarında submukozal birikimler ile kendini gösterebilir. Polimyozi otoimmün inflamatuvar bir myopati olup simetrik proksimal kas ve boyun kuvvetsizliği ile karakterizedir. Polimyozi hastaların %80-90 kadarı kortikosteroid ve siklofosamid kombinasyonları ile başarılı bir şekilde tedavi edilebilir. Polimyozi ise sekonder interstisyel akciğer hastalığına neden olur ve bu iki tablonun radyolojik ayrımı kolay olmayabilir. *Türk Fiz Tıp Rehab Derg* 2012;58:159-61.

**Anahtar Kelimeler:** Polimyozi, interstisyel akciğer hastalığı, primer pulmoner amiloidoz

### Introduction

Amyloid is composed of insoluble proteins and is easily deposited in various organs (1). In primary systemic amyloidosis, clonally expanded plasma cells that are located in the bone marrow produce monoclonal immunoglobulin (Ig) light chains, and these light chains accumulate in the extracellular matrix of many organs, and later cause organ dysfunction. Localized forms of amyloidosis are also present, which describes amyloidosis restricted to a specific organ. The lung is one of these organs and primary pulmonary amyloidosis (PPA) as a localized type of amyloidosis can present as parenchymal nodules, diffuse alveolar damage, or submucosal deposits in the

airways (2-5). Similar to the systemic disease, protein analysis of localized type of amyloidosis in the lung also reveal monoclonal Ig light chains (6-8).

Polymyozitis is an inflammatory autoimmune myopathy characterized by symmetric weakness of the limb girdle and anterior neck muscles. The thorax is frequently affected and this comes out in three forms: (a) hypoventilation and respiratory failure; (b) interstitial pneumonitis, (c) aspiration pneumonia (9). The frequency of radiographic parenchymal abnormalities is nearly 5% and is predominantly seen as basal reticular pattern which may become diffuse by time and progress to honeycombing (9).

Herein, we present a case of PPA radiologically mimicking interstitial lung disease in a patient with polymyozitis.

## Case

A 53-year-old man was admitted to our university hospital with generalized pain and muscle weakness. The first complaints began 15 years ago with proximal limb pain increasing with activity and weakness. When he was admitted for a lipoma excision 5 years later, his creatinin kinase (CK) level was found to be increased (CK: 3108 U/L) and referred to the department of internal medicine for further investigation. Laboratory findings were: CK: 3170 U/L erythrocyte sedimentation rate: 25 mm/h and C-reactive protein<1. Anti-nuclear antibody (ANA) and extractable nuclear antigens (ENA), especially anti-SS-A (Ro) were reported to be positive. As an initial work up for polymyositis, electromyography (EMG) and a muscle biopsy was performed. EMG was reported to be within normal limits, however, the muscle biopsy revealed inflammatory myopathy with mild muscle damage. He was also evaluated for secondary malignancies. Tumor markers and abdominal ultrasonography (USG) were unremarkable except for hepatomegaly (right lobe measured as 16.5cm). After the detection of the hilar prominence on a chest X-Ray, pulmonary high-resolution computed tomography (HRCT) was performed, which revealed a mass, 10 millimeters in diameter, with cystic nature, localized at anterior mediastinum apart from the thyroid gland without any calcification. With the possible diagnosis of thymoma, biopsy from the mass was decided; but the patient rejected the procedure. Therefore, he was discharged from the hospital with appropriate treatment, but lost to follow-up for 3 years, until he was readmitted to the hospital suffering from generalized pain, weakness and right wrist ache, unresponsive to several analgesics and myorelaxants. He was unable to lift his arm over his head. The muscle biopsy and the EMG were performed again. This time, EMG revealed inflammatory myopathy and the pathological diagnosis of the muscle biopsy diagnosis was compatible with myositis. Therefore, the patient was re-evaluated for polymyositis regarding to his clinical findings. Abdominal USG and CT were reported to be normal. The thoracic CT revealed the same mass and furthermore, verities compatible with interstitial lung disease (Figure 1). At the end, an open lung biopsy with thoracotomy for interstitial lung disease and thymectomy were performed, and the pathologic diagnosis was as micronodular type thymoma, surrounded by monoclonal B cell proliferation approved by molecular clonality studies and also amyloid accumulation in the lung, which was shown to be AL type of amyloid by mass spectrometry (Figure 2). Therefore, rectal biopsy was also done to search systemic amyloidosis but the result was negative for amyloidosis. After the operation, monthly intravenous pulse cyclophosphamide and mesna was ordered and his complaints disappeared after therapy. The patient do well with oral steroid and cyclophosphamide therapy for 5 years.

## Discussion

Primary pulmonary amyloidosis (PPA) is a rare disease of plasma cell origin, composes 1.1% of all amyloidosis cases (10). Localized type of amyloidosis in the lung can be seen in two forms: limited

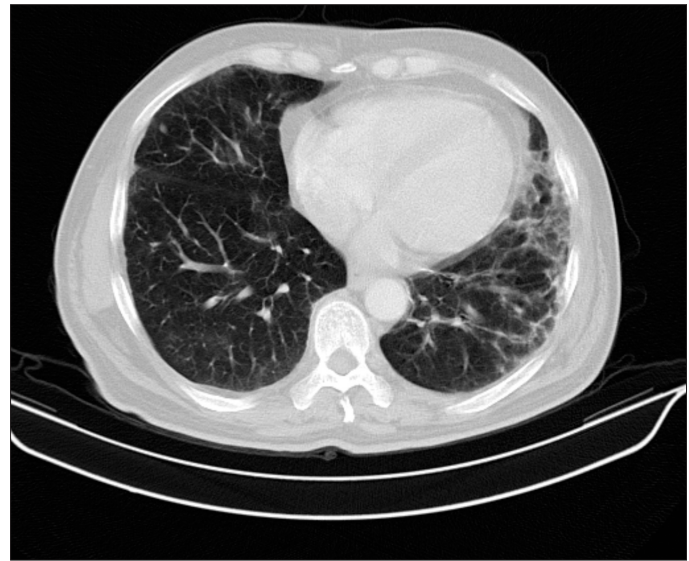


Figure 1. Peripheral predominant reticular abnormality with traction bronchiectasis and honeycombing. Also the vascularization of the lung is prominent with fissuritis on the right side. The pericardial fluid is a bit increased but with in normal limits.

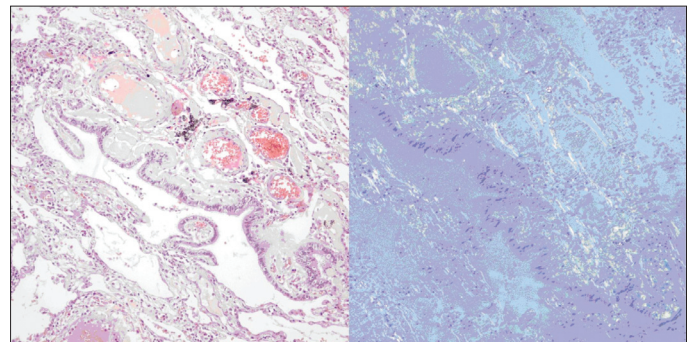


Figure 2. Lung biopsy revealed extracellular eosinophilic amorphous material around bronchioli. H&E;200x. Same area in Congo red stained section with apple-green birefringence under polarized light. Congo red;200x.

bronchopulmonary amyloidosis, which is usually AL type, may manifest itself as either tracheobronchial deposits or parenchymal nodules and sometimes masses in the lung paranchyme (2-5), diffuse interstitial pulmonary amyloidosis which clinically presents as recurrent pneumonias, cough, and bronchiectasis, is rare and usually associated with systemic AL type of amyloidosis and usually there are deposits in the alveolar-capillary gas exchange zone (10). Amyloidosis may also manifest itself as pulmonary hypertension, hilar and mediastinal lymphadenopathy or pleural involvement (1, 3, 4). AL type of amyloidosis may be associated also with pulmonary lymphoma (11). Occasionally, non-fibrillar, negatively stained with congo red, immunoglobulin deposits may occur, presenting as parenchymal pulmonary nodules or cysts (2, 12).

O' Regan et al. (10) evaluated their 3 patients with CT scan, pulmonary function tests and bronchoscopy. They pointed out bronchoscopy as the most useful diagnostic tool for PPA, but not for monitoring the disease progression.

Polymyositis is an inflammatory autoimmune myopathy that is characterized by symmetric weakness of the limb girdle and

anterior neck muscles. The incidence of polymyozitis is approximately 5-10 cases per million per year and two third of the patients are female (9). The thorax is frequently affected and this may be in three forms: (a) hypoventilation and respiratory failure due to involvement of the respiratory muscles, (b) interstitial pneumonitis, and (c) aspiration pneumonia due to pharyngeal muscle weakness. The last one is probably the most common pulmonary complication (9).

Interstitial lung disease in patients with polymyozitis or dermatomyozitis has a wide histological spectrum (9). Three major groups can be identified: bronchiolitis obliterans with organizing pneumonia (BOOP), usual or nonspecific interstitial pneumonia, and diffuse alveolar damage. The histologic appearance of interstitial lung disease in patients with polymyozitis and dermatomyozitis is a prognostic factor (13). Diffuse alveolar damage and usual interstitial pneumonia are poor prognostic subtypes, with only a 33% 5-years survival rate (13). However, BOOP and nonspecific interstitial pneumonia have better prognosis. The frequency of radiographic parenchymal abnormalities is nearly 5%, mostly symmetric, predominantly basal reticular pattern which may become diffuse by time and progress to honeycombing (9). Bilateral areas of consolidation corresponding histologically to diffuse alveolar damage or BOOP, generally develop in a 2- to 3-week period (14). Prominent interlobular septa, ground-glass attenuation, patchy consolidation, parenchymal bands, irregular peribronchovascular thickening, and subpleural lines may be the initial findings on HRCT (14).

In conclusion, radiological findings of PPA and interstitial lung disease secondary to polymyozitis may mimic each other, therefore, to exclude amyloidosis, transbronchial biopsy should be performed.

#### Conflict of Interest:

Authors reported no conflicts of interest.

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