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# A Rare Case: Primary Pulmonary Amyloidosis

## Nadir Bir Olgu: Primer Pulmoner Amiloidoz

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**ABSTRACT**

Amyloidosis is a rare disorder characterized by the extracellular deposition of insoluble protein aggregates. There are two common types of amyloidosis: Amyloid A (AA) and amyloid light chain (AL). AA amyloidosis typically occurs secondary to chronic inflammatory processes such as tuberculosis and rheumatoid arthritis, with pulmonary involvement being a rare manifestation. AL amyloidosis, on the other hand, is often associated with conditions like multiple myeloma, gammopathies, and idiopathic primary amyloidosis. While kidney involvement is common, pulmonary involvement can also occur as part of systemic amyloidosis or, more rarely, in an isolated form. Pulmonary amyloidosis can present in three forms: nodular, diffuse, and tracheobronchial. This study presented a case of primary pulmonary amyloidosis in the nodular form, identified in a 69-year-old female patient.
**Keywords:** Nodular amyloidosis; amyloid; light chain; wedge.

**ÖZ**

Amiloidoz suda çözünmeyen proteinöz yapıların hücre dışında birikimi ile karakterize nadir görülen bir hastalıktır. Amiloid A (AA) ve amiloid hafif zincir (AL) olmak üzere iki yaygın amiloidoz tipi mevcuttur. AA tipi amiloidoz tüberküloz ve romatoid artrit gibi kronik inflamatuar süreçlere sekonder olarak ortaya çıkar ve pulmoner tutulum nadir bir bulgudur. Öte yandan, AL tipi amiloidoz ise sıklıkla multiple miyelom, gamopatiler ve idiyopatik olarak ortaya çıkan primer amiloidoz gibi durumlarla ilişkilidir. Böbrek tutulumu yaygın olmakla birlikte pulmoner tutulum sistemik amiloidozun bir parçası olarak veya daha nadiren izole bir formda da ortaya çıkabilir. Pulmoner amiloidoz nodüler, diffüz ve trakeobronşiyal olmak üzere üç formda ortaya çıkabilir. Bu çalışmada, 69 yaşında bir kadın hastada saptanan nodüler formdaki primer pulmoner amiloidoz vakası sunulmuştur.
**Anahtar kelimeler:** Nodüler amiloidoz; amiloid; hafif zincir; wedge.

## INTRODUCTION

Amyloidosis is a rare disease characterized by the extracellular accumulation of water-insoluble, proteinaceous substances called amyloid, first described by Virchow in 1854 (1,2). While amyloid deposits can occur in any organ, they most commonly affect the kidneys. There are two main types of amyloidosis: Amyloid A (AA) and amyloid light chain (AL). AA amyloidosis typically develops as a secondary complication of chronic inflammatory conditions, while AL amyloidosis is often idiopathic and represents primary amyloidosis. Pulmonary amyloidosis may manifest as part of primary systemic amyloidosis or as an isolated condition (1,3).

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Amyloid accumulation in the respiratory tract was first described by Lesser in 1877. Pulmonary amyloidosis can present in three forms: nodular, diffuse, and tracheobronchial. Localized forms are often seen as tracheobronchial disease (2,4).

In this report, a rare case of primary pulmonary amyloidosis, along with a review of the relevant literature, was presented.

## CASE REPORT

A 69-year-old female was admitted with progressively worsening dyspnea. The patient's physical examination findings revealed a dyspneic appearance, mild retractions in the intercostal muscles, and coarse crackles in the breath sounds. Peripheral non-invasive oxygen saturation without supplemental oxygen was measured at 92%. Her laboratory results revealed a white blood cell count of 10,500 /mm³, an eosinophil level of 3%, and a C-reactive protein (CRP) level of 35 mg/L. A chest X-ray showed bilateral, patchy consolidations with nodular features, leading to a presumptive diagnosis of pneumonia (Figure 1). Intravenous moxifloxacin was administered for 14 days however, despite partial clinical improvement, the patient's dyspnea persisted.

Further evaluation suggested interstitial lung disease, and a computed tomography (CT) scan of the thorax revealed bilateral nodular consolidations, more pronounced on the right side (Figure 2). A tissue biopsy was planned for differential diagnosis, and a wedge resection was performed via mini-thoracotomy with general anesthesia, 3 weeks after medical treatment. Pathological evaluation revealed nodular areas stained with Congo red, which appeared green under polarized light, leading to the diagnosis of nodular amyloidosis (Figure 3).

The patient underwent biochemical tests to evaluate liver and kidney functions, specifically for systemic amyloidosis screening. No pathological findings were observed in the conducted tests. During the systemic screening for amyloidosis, ultrasound and CT did not reveal any involvement of other organs. The patient was diagnosed with primary pulmonary amyloidosis, and colchicine therapy was initiated. After six months of treatment, a clinical response was observed, although follow-up thoracic CT scans showed no significant radiological regression (Figure 4). Clinically, there was improvement in exertional capacity and reduced oxygen requirements, and the patient no longer required regular oxygen support during follow-up.

## DISCUSSION

Amyloidosis has an incidence of approximately 1 in 100,000 annually (4,5). AA amyloidosis typically occurs as a secondary complication of chronic inflammatory diseases, such as tuberculosis and rheumatoid arthritis, with pulmonary involvement being rare. In contrast, AL amyloidosis is more commonly associated with multiple myeloma, gammopathies, and idiopathic cases, representing primary amyloidosis. While primary systemic amyloidosis can affect all organs, primary localized amyloidosis specifically leads to pulmonary involvement (1,4). In a study by Hui et al. (6) involving 48 cases of pulmonary amyloidosis, 28 patients had the nodular form, 14 had tracheobronchial involvement, and 6 had diffuse disease.

Nodular pulmonary amyloidosis is often asymptomatic, while diffuse and tracheobronchial forms typically present with respiratory symptoms. In the tracheobronchial form, dyspnea and hemoptysis are common (1,2,4). Peng et al. (7) reported a case of a 59-year-old patient with calcifications and atelectasis around the bronchi on radiological evaluations. The patient presented with cough, dyspnea, stridor, hemoptysis, and fever. Our patient had the nodular

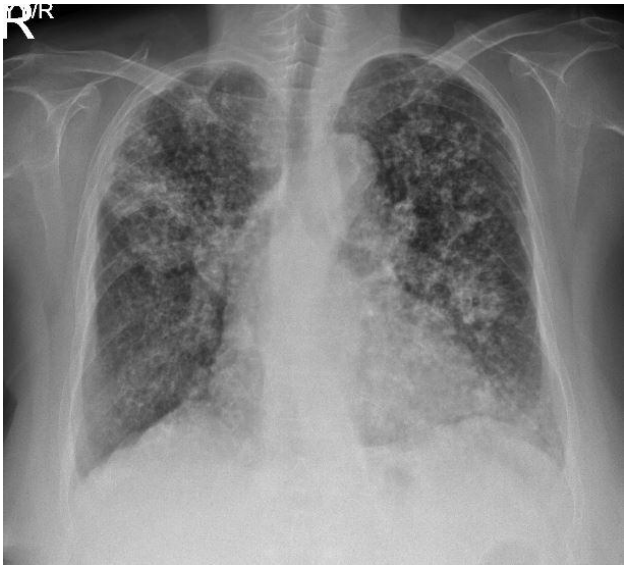

**Figure 1.** Amyloid deposition on chest X-ray

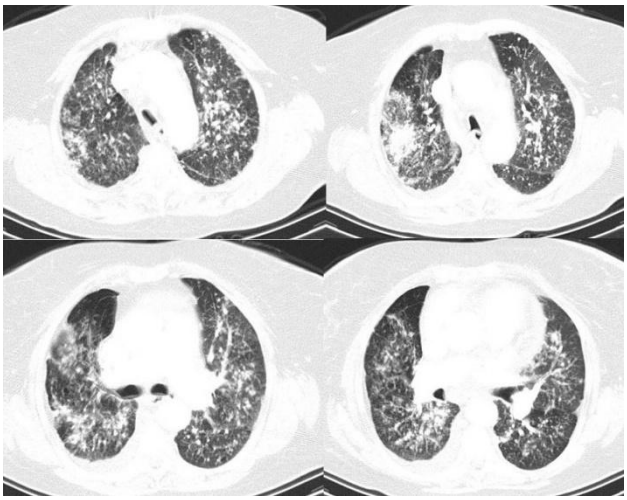

**Figure 2.** Chest computed tomography prior to surgery showing nodular amyloid deposition in the lungs

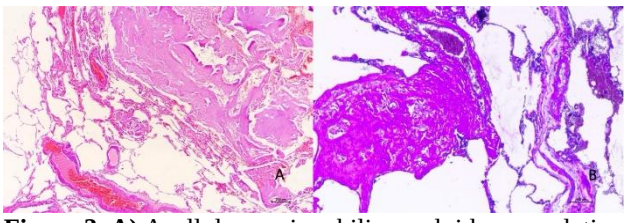

**Figure 3. A)** Acellular, eosinophilic amyloid accumulation in the alveolar septum, positively stained with Congo red on pathological examination (100x). **B)** Amyloid accumulation in the alveolar septum, stained positively with histochemical crystal violet (200x).

form, with dyspnea as the predominant symptom, and pulmonary function tests revealed a restrictive pattern.

The diagnosis of pulmonary amyloidosis is often made incidentally or based on respiratory symptoms, with chest X-rays and thoracic CT scans playing a crucial role. Imaging studies may show single or multiple nodular lesions in the nodular type, while the diffuse form presents with non-specific consolidations. Amyloid nodules can reach sizes of up to 15 cm and typically have lobulated contours. Tracheobronchial amyloid deposits can be detected via bronchoscopy (2,4,5). Amyloid nodules can radiologically mimic lung cancer. Oki et al. (8) presented a case of a 59-year-old female patient with an amyloid nodule located in the anterior upper lobe of the left lung, which radiologically mimicked lung cancer, and was diagnosed with an amyloid nodule in the pathological examination performed after wedge resection. In our case, thoracic CT scans revealed bilateral nodular lesions, consistent with findings reported in the literature.

Pathological evaluation is considered the gold standard for diagnosing amyloidosis. Congo red staining highlights amyloid deposits, which exhibit green birefringence under polarized light (2,4).

Although there is no definitive treatment for pulmonary amyloidosis, colchicine and steroid therapy have been used for both diffuse and nodular forms. Studies have shown symptomatic relief with colchicine, although no improvement in survival has been demonstrated. In tracheobronchial amyloidosis, bronchoscopic interventions, including laser therapy and stent placement, have been reported to yield successful outcomes (1,2,4). In our patient with nodular amyloidosis, colchicine therapy was initiated, resulting in symptomatic improvement, although no radiological response was observed.

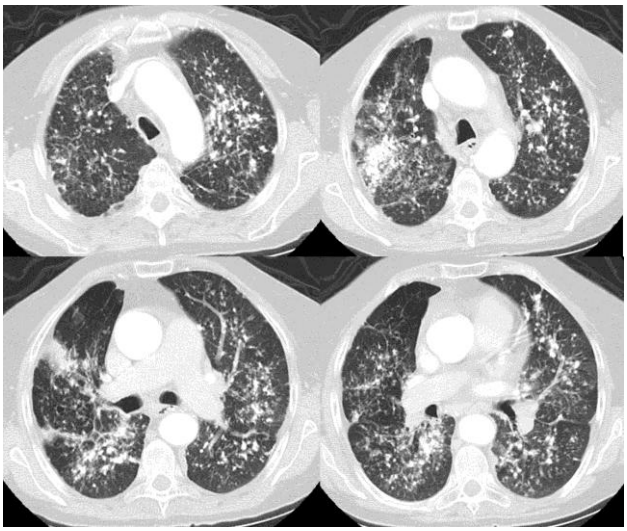

**Figure 4.** No significant change in amyloid deposition on follow-up chest computed tomography after treatment.

In conclusion, pulmonary amyloidosis should be considered in the differential diagnosis of diffuse or nodular parenchymal lesions in patients with interstitial lung disease.

**Informed Consent:** Written informed consent was obtained from the patient for publication and accompanying images.

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The first author, Mustafa Kuzucuoğlu, was working at Balıkesir University during the study period, and was appointed to a different city, which reflects his current place of employment.

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