

Case Report

Tracheobronchial Amyloidosis: A Case Report

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Article History

Received: 26.02.2022

Accepted: 01.04.2022

Published: 12.04.2022

Journal homepage:

<https://www.easpublisher.com>

Quick Response Code



Abstract: Amyloidosis is a spectrum of diseases, in which various proteins which are a usual component of plasma are deposited as insoluble beta-pleated sheets extracellularly, disrupting the function of various organs. Amyloid light-chain amyloidosis occurs due to the deposition of proteins, derived from immunoglobulin light chains, routinely manifesting with multisystem involvement. Pulmonary involvement is seen in about 50% of cases [1]. Three common patterns of amyloidosis on computed tomography (CT) chest are tracheobronchial, nodular parenchymal, and diffuse alveolar septal variety. Here we report a case of amyloidosis involving tracheobronchial tree due to localized deposition of amyloid. Knowledge about pulmonary amyloidosis is important due to nonspecific findings in the CT chest and its poor prognosis.

Keywords: Amyloid light-chain amyloidosis, tracheobronchial amyloidosis, pulmonary amyloidosis, apple-green birefringence.

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INTRODUCTION

Amyloidosis is a group of disorders characterized by abnormal extracellular protein deposition, formed from various precursor proteins causing structural and functional disruption in various tissues. Most commonly the heart, liver, kidneys, and skin are affected. The patient presents with a wide range of clinical manifestations, depending on the site, amount, and type of protein deposited.

Primary amyloidosis is caused by plasma cell or lymphoplasmacytic neoplasm in which the monoclonal plasma cells secrete intact or fragments of abnormal immunoglobulin light chain that deposit in various tissues and form a beta-pleated sheet structure (amyloid light chain). Secondary amyloidosis or reactive systemic amyloidosis is characterized by the deposition of amyloid-associated (AA) protein formed by the incomplete breakdown of serum amyloid A, an acute phase reactant. Hence it is associated with chronic inflammatory conditions such as Rheumatoid arthritis, chron's disease, Familial Mediterranean fever, Hansen's disease, and tuberculosis. Both types are usually systemic in distribution. Localized amyloidosis is a condition in which amyloid infiltration is limited to a single organ in the absence of any systemic disease and plasma cell dyscrasia. Isolated tracheobronchial amyloidosis, however, is a rare disorder with amyloid

deposits limited specifically to tracheal and bronchial tissue. We report a case of tracheobronchial amyloidosis, which is often misdiagnosed as obstructive airway disease.

CASE HISTORY

A 65-year-old known diabetic female on regular treatment presented with increased dyspnoea, productive cough, and fever for 2 months' duration. She had a history of exertional dyspnoea for 12 years and history of recurrent lower respiratory tract infections for the last 3 years. No history of hemoptysis, loss of weight, loss of appetite, or no past history of tuberculosis or asthma. No family history of malignancy or connective tissue disorders. Clinical examination revealed crackles in the left anterior axillary area. Other systems were within normal limits. Sputum AFB was negative and Sputum cytology was negative for malignant cells. Chest X-ray and CT scan showed collapse and consolidation of the left middle and lower lobes. Fiber optic bronchoscopy was done in view of recurrent lower respiratory tract infections which showed multiple nodular lesions over the main carina, right and left bronchial tree. The right and left upper lobes were occluded with nodular lesions. She had no history of rheumatoid arthritis, tuberculosis, or other chronic ailments.

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Her complete blood count and serum calcium were within normal limits(8.8gm/dl). A skeletal survey was done which showed no focal punched out lytic lesions. Serum protein electrophoresis was done which was normal. She was treated symptomatically with nebulized bronchodilators and intravenous antibiotics.

Histopathology of the bronchial biopsy showed acellular homogenous eosinophilic material within the subepithelial region, and on Congo-red showed amyloid deposition and apple-green birefringence under polarized microscopy.

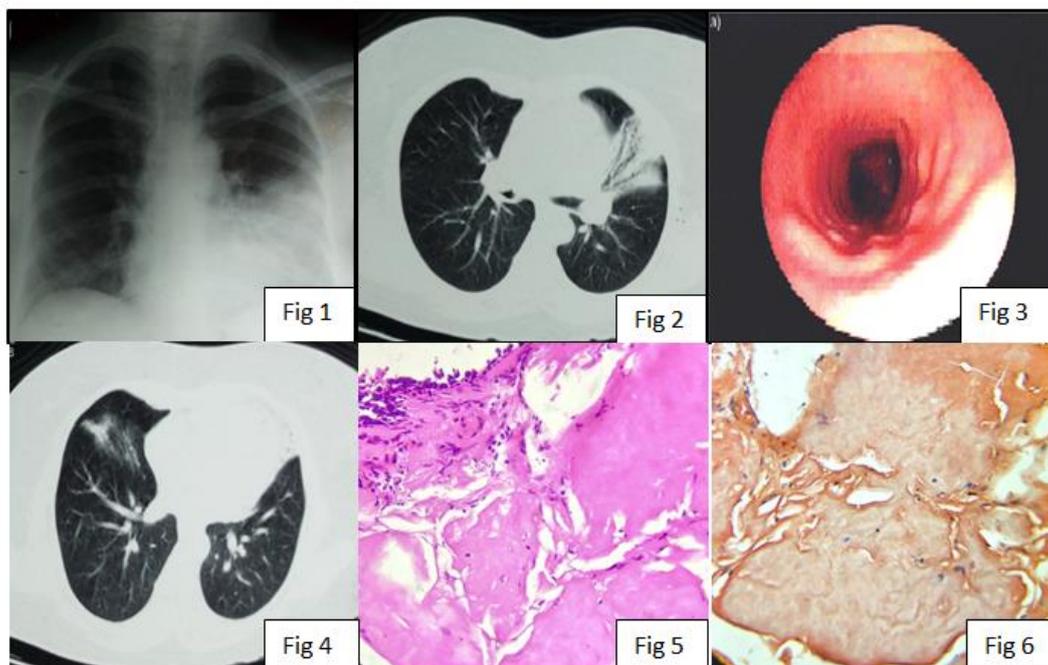


Fig 1 and 2: X-ray (1) and CT scan (2) showing left lower and middle lobe collapse and consolidation. Fig 3(bronchoscopy) & 4 (CT Scan) showing nodularities near carina and within the bronchial tree. Fig 5: Homogenous eosinophilic material deposition in the subepithelial region. Fig 6: Congo-red stain: amyloid deposition in the subepithelial region

DISCUSSION

Tracheobronchial amyloidosis usually occurs in the 5th to 6th decade and with an incidence of 6-10 cases/million/year⁽¹⁾. Most of the patients are usually asymptomatic. Other presentations which are common are wheeze, stridor, cough, and recurrent pneumonia. Pulmonary involvement includes three main patterns-tracheobronchial, nodular parenchymal, and diffuse parenchymal/diffuse alveolar septal pattern. The tracheobronchial pattern of involvement is the most common variety seen in 53% of cases of pulmonary amyloidosis [2]. HRCT chest shows localized nodules, circumferential thickening of the trachea, and rarely narrowing of the tracheobronchial lumen may be present which lead to recurrent infection, atelectasis, and bronchiectasis. In our case, the patient had cough and dyspnoea and recurrent LRTI infections. Radiological studies show a tracheobronchial pattern of involvement with distal collapse and consolidation of the left lower lobe of the lung.

Amyloidosis is classified into various subtypes as per WHO classification based on the type of fibrillar protein deposited. As many as 30 varies are described in human amyloidosis, the most important and common types being 1) AL amyloidosis (primary) due to immunoglobulin and light chain deposition 2) AA

amyloidosis/secondary amyloidosis due to serum AA protein deposition. 3) dialysis-related amyloidosis due to beta 2-microglobulin deposition 4) Heritable amyloidosis due to mutant transthyretin (pre albumin) deposition. 5) senile amyloidosis due to wild-type transthyretin deposition.

Screening for symptoms of systemic amyloidosis should be performed in any patient with confirmed TBA. Studies include electrocardiogram, echocardiogram, serum and urine protein electrophoresis, and creatinine level. Evaluation for multiple myeloma should be included in the workup. The cardiac, renal, and hepatic disease can range from asymptomatic to end-organ failure. Our patient was evaluated by hematology/oncology and the workup showed no evidence of systemic amyloidosis or any renal/cardiac/hepatic/bone involvement.

Diagnosis is made by tissue biopsy. With isolated TBA, a biopsy of lesions within the tracheobronchial airway is necessary for diagnosis. Histological findings characteristic of amyloidosis includes the classic “apple green” birefringence with congo-red staining and mass spectrometry can be used to further identify the specifically deposited protein

subunits. In the present case, the biopsy findings were confirmative.

Treatment of TBA remains a topic under investigation, with mixed results. Bronchoscopic recanalization with ND: YAG and CO2 laser, and mechanical resection have been documented to be successful in individual cases. Treatment with systemic glucocorticoids showed benefit in systemic amyloidosis.

The prognosis of patients with TBA is variable. Most of the literature consists of case reports or small case series. One study documented a 30% mortality rate from disease after 7-12 years [1], secondary to progressive obstructive disease while another study estimated approximately 30-50% 5-year survival [3]. Patients with systemic disease often have a much poorer prognosis. Our patient's symptoms were improved after the treatment.

CONCLUSION

Amyloidosis should be considered as one of the differentials along with another common diagnosis

such as malignancy, infection, and vasculitis when multisystem involvement is seen or when non-resolving pneumonia is encountered. Knowledge about pulmonary amyloidosis is important due to nonspecific findings in the CT chest. Although the prognosis of tracheobronchial amyloidosis is usually good, follow-up is crucial to act appropriately, if significant endobronchial occlusion occurs. We report a rare case of isolated tracheobronchial amyloidosis in an elderly lady.

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Cite This Article: Silpa S. Raj, Rejla R, Sankar S (2022). Tracheobronchial Amyloidosis: A Case Report. *East African Scholars J Med Sci*, 5(4), 109-111.