

RARE AND INTERESTING
CASES IN
**PULMONARY
MEDICINE**

ALI ATAYA
ELOISE HARMAN



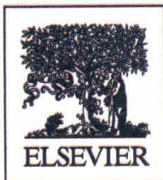
Rare and Interesting Cases in Pulmonary Medicine

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Dedication

**To my parents
To my wife
And to all my mentors and patients over the years**

– Ali Ataya

**To my family
My mentors Dr. Jay Block and Dr. William Bell
And to the patients who have taught me so much**

– Eloise Harman

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Preface

Patients with rare lung disorders are often misdiagnosed or diagnosed late in the course of their disease. This is frequently due to physicians not considering the diagnosis in the first place. However, interest in rare and orphan lung diseases has been increasing among healthcare providers, pharmaceutical companies, and patients.

The goal of this book is to provide an introduction to various rare lung disorders, with the hope that this may assist in the recognition, diagnosis, and treatment of these diseases as they are encountered in clinical practice. This book may also benefit physicians studying for their pulmonary board exams. Each case begins with a case study of a patient with one of these rare diseases followed by a rapid review of the disease or syndrome. This case-centered approach is expected to help the reader to recall the information if they encounter patients with one of these rare diseases.

Ultimately, we hope this book will contribute to improving diagnosis and care of patients with rare lung disorders.

Ali Ataya, MD
Eloise Harman, MD

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Case 1

A 60-year-old Caucasian female presents with progressive shortness of breath with exertion and a nonproductive cough for the last year and a half. She is a lifelong nonsmoker, has no significant past medical problems, and is not on any medications.

Examination of the heart and lungs is normal and there is no digital clubbing. A chest computed tomography scan revealed multiple small peripheral nodular opacities in the right upper and lower lobes as well as hilar and mediastinal adenopathy (Fig. 1.1). Endobronchial ultrasound with transbronchial needle aspiration of the mediastinal lymph nodes was performed. Histology is shown in Fig. 1.2. Further workup showed no other organ involvement of the disease.

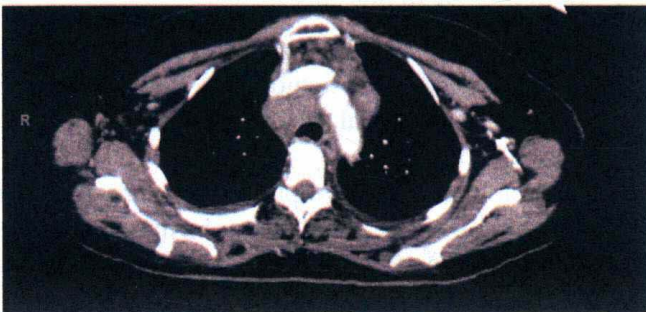


FIGURE 1.1 Chest computed tomography scan with contrast showing enlarged mediastinal 4R node.

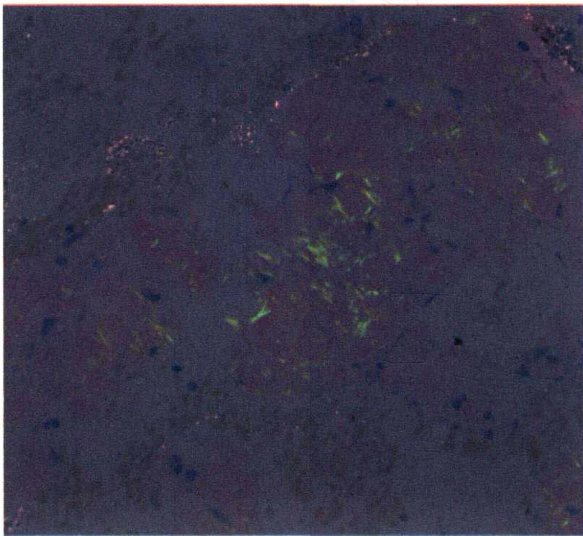


FIGURE 1.2 Histology showing clumps of amorphous material with Congo red stain under polarized light.

What is the diagnosis?

PULMONARY AMYLOIDOSIS

Amyloidosis is a systemic disease characterized by extracellular deposition of amyloid, which constitute insoluble β -pleated protein sheets, in different organs. Amyloidosis can be primary/idiopathic (AL type), or secondary/reactive (AA type). The secondary form may occur in the setting of an underlying malignancy, chronic inflammatory, or infectious disease, appear in the setting of chronic renal disease, or be heritable. Isolated pulmonary amyloidosis usually occurs in the setting of the idiopathic form of the disease. Isolated pulmonary amyloidosis is characterized by the occurrence of amyloidosis in the lungs without any systemic involvement.

Patients have nonspecific symptoms due to the diversity of its pulmonary manifestations and tissue biopsy is necessary to make the diagnosis. Isolated pulmonary amyloidosis comes in multiple forms:

1. Tracheobronchial amyloidosis: Most common form. Patients may present with cough, dyspnea, wheezing, or hemoptysis. Patients may have thickened trachea with stenosis. If proximal lesions are present, they may result in fixed upper airway obstruction.
2. Nodular form: Patients may be asymptomatic or present with a cough. A single nodule or multiple small nodular lesions may appear peripherally in the lower lobes. Amyloid nodules may be calcified and cavitate in 10% of cases.
3. Amyloid adenopathy: Amyloid is deposited in the hilar and mediastinal lymph nodes, usually bilaterally. This form of the disease rarely occurs alone or without systemic involvement.
4. Diffuse interstitial form: This is the rarest form of the disease. Amyloid gets deposited in the pulmonary interstitium between the alveoli and blood vessels, impairing gas transfer. Imaging will show a reticular or reticulonodular pattern that may present asymmetrically. Patients succumb to respiratory failure.

Tissue biopsy is the gold standard for diagnosis. Histology will show pink amorphous material that under polarized light will stain apple-green birefringence with Congo red stain.

There is no effective treatment for the disease. Patients with tracheobronchial involvement may undergo bronchoscopic treatment with Nd-YAG laser or clipping for obstructing lesions. For other forms external beam radiation and systemic immunosuppression have been used to halt progression.

This patient underwent further workup that showed no systemic involvement, including a bone marrow biopsy. She was diagnosed with nodular amyloid with hilar and mediastinal lymph node involvement and referred for systemic chemotherapy treatment.