EJCRIM
European Journal
of Case Reports
in Internal Medicine

ISOLATED PULMONARY LIGHT CHAIN DEPOSITION DISEASE

Manogna Pendyala[1], Pranathi Bandarupalli[1], Purnoor Kaur[1], Vinod Khatri[2], Krishna Khatri[3]

[1] Internal Medicine, St Vincent Medical Center, Toledo, USA
[2] Pulmonary and Critical Care Medicine, St. Vincent Medical Center, Toledo, USA
[3] Internal medicine, BJ Medical College, Ahmedabad, India

Corresponding author's e-mail: manognapendyala97@gmail.com

Received: 17/12/2026
Accepted: 12/01/2026
Published: 26/01/2026

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: Signed informed consent for the publication of this case was obtained from the patient.
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How to cite this article: Pendyala M, Bandarupalli P, Kaur P, Khatri V, Khatri K. Isolated pulmonary light chain deposition disease. *EJCRIM* 2026;**13**:doi:10.12890/2026_006086

ABSTRACT

Background: Pulmonary involvement is a significant aspect of many autoimmune diseases. Different pulmonary manifestations of autoimmune diseases encompass a variety of conditions that impact the lungs, including interstitial lung disease, cystic lung disease, pulmonary hypertension, and pleural involvement. One rare manifestation is light-chain deposition disease. Our case report aims to highlight the condition's rarity and presentation variability.

Case presentation: A 55-year-old Caucasian female with a history of Sjögren's syndrome, papillary thyroid carcinoma status post thyroidectomy, and chronic leukopenia underwent a computed tomography (CT) of the chest for routine oncological surveillance. Imaging revealed an 8 mm left upper lobe nodule with bilateral bullous lung changes. A positron emission tomography/CT showed mildly ^{18}F-fluorodeoxyglucose-avid pulmonary nodules with large pneumatoceles, but no evidence of metastatic disease, which increased in size over the next two years, increasing the concern for progressive lung disease. Subsequently, a lung biopsy revealed monotypic kappa light-chain deposition with plasma cell infiltration, establishing the diagnosis of light-chain deposition disease. Despite the radiographic progression, the patient remained asymptomatic and stable. She was managed conservatively with serial paraproteinemia and image-based monitoring.

Conclusion: Pulmonary light chain deposition disease is an infrequent condition requiring a high index of suspicion for diagnosis. Imaging findings can help identify it, but biopsy remains the definitive diagnostic tool. Due to the lack of established management guidelines, treatment is personalized based on disease extent and organ involvement. Further research is needed to understand better this rare entity's prognosis and optimal treatment approaches.

KEYWORDS

Lung nodules, cysts, Sjogren's syndrome, light chains, metastasis

LEARNING POINTS

- Pulmonary light-chain deposition disease is a rare and under-recognized manifestation.
- Clinicians must have a high index of clinical suspicion while evaluating cystic lung disease, particularly in the background of an autoimmune disease.
- This case reinforces the importance of considering light chain deposition disease as a differential in evaluation of unexplained lung nodules or cysts.

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INTRODUCTION

Cystic lung diseases are a group of disorders characterized by multiple air-filled cysts in the lung[1,2]. The most encountered cystic lung diseases are lymphangioleiomyomatosis, pulmonary Langerhans cell histiocytosis, Birt-Hogg-Dube syndrome, and lymphoid interstitial pneumonia[1,2]. Light chain deposition disease (LCDD) is a rare systemic disorder characterized by monoclonal immunoglobulin light chain deposition in various tissues, which can present as cystic lung disease. Isolated pulmonary involvement is even less common[2]. The most common organ involved is the kidney. Although rare, a few cases of isolated pulmonary involvement in light-chain deposition disease have been reported. Our case report adds to this growing list, further highlighting the variability in presentation.

CASE DESCRIPTION

A 55-year-old Caucasian woman with a history of Sjögren's syndrome, papillary thyroid cancer (s/p thyroidectomy in 2020), anaemia, and chronic leukopenia presented in 2019 with bilateral leg rash. Sjögren's syndrome was confirmed via anti-SSA/SSB antibodies, and hydroxychloroquine was initiated. Labs showed persistent hypergammaglobulinemia, elevated kappa light chains, and an abnormal kappa/lambda ratio, though serum and urine electrophoresis revealed no M-protein. Bone marrow biopsy demonstrated trilineage haematopoiesis without plasma cell dyscrasia or clonal proliferation; flow cytometry and fluorescence *in situ* hybridization were negative. In the absence of a plasma cell disorder, the cytopenias were attributed to immune-mediated suppression, and routine monitoring was recommended.

The patient remained clinically stable on hydroxychloroquine until 2021, when surveillance after her thyroidectomy showed an 8 mm nodule in the left upper lobe and bilateral bullous lung changes. A 2022 Positron emission tomography (PET)/Computed tomography (CT) indicated mildly ^{18}F-fluorodeoxyglucose-avid pulmonary nodules with large pneumatoceles but no metastatic disease. Over two years, serial CT scans revealed nodule enlargement and new subsolid and cyst-associated nodules *(Fig. 1)*, raising concerns about progressive lung pathology, although the patient remained asymptomatic. She eventually consented to a bronchoscopy with transbronchial biopsy, which showed monotypic kappa light chain deposition and plasma cell infiltration. Grocott methenamine silver, acid-fast bacillus, Congo red, and Movat stains were negative, excluding fungal, mycobacterial, and amyloid-related diseases, as well as other diseases. Mass spectrometry confirmed the presence of kappa light-chain peptides, indicating light-chain deposition disease in the lungs. Given no systemic involvement, conservative management was adopted, including biannual paraproteinemia monitoring, surveillance of absolute neutrophil count, and CT scans. Hydroxychloroquine was continued due to stable Sjögren's syndrome. The patient remained clinically and radiographically stable.

DISCUSSION

Multiple air-filled lucencies characterize cystic lung diseases, and patients with cystic lung disease may be asymptomatic, with incidental discovery on imaging obtained for other reasons[1]. In the case mentioned above, the differential diagnosis included:

- *Lymphoid interstitial pneumonia:* A rare lung manifestation of Sjögren's syndrome seen in women in their fifth decade, characterized by diffuse reticulonodular opacities and peribronchovascular

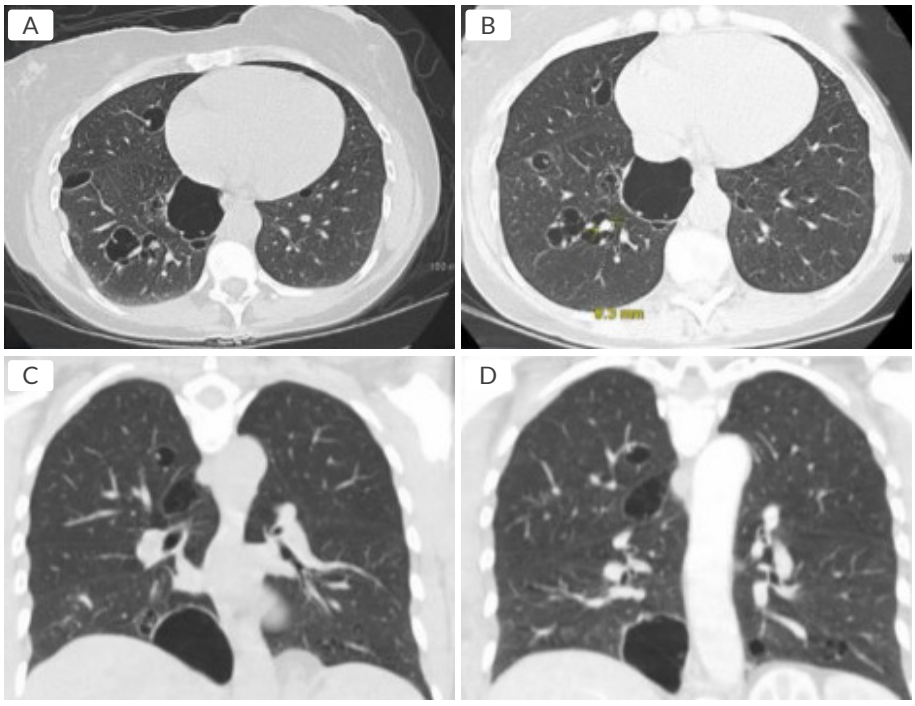


Figure 1. *CT images showing the progression of the disease showing the evolution of the lung cyst in the coronal view (A, B) and in sagittal view (C,D).*

thickening. Cysts vary in size, are predominantly located in the lower lung fields, and have a perivascular distribution[1,2].

- *Cystic metastasis:* With a history of thyroid malignancy, lung metastasis is a consideration. The lungs are the most common site of thyroid metastasis (43%). Although metastasis usually presents as nodules, it can also have cystic components/ lesions[3]. However, the PET/CT findings made a malignant process less likely.
- *Infections:* Certain pulmonary infections, like staphylococcal pneumonia and *Pneumocystis jirovecii*[4], can lead to cyst formation but typically present with acute symptoms, unlike our case, making infectious aetiology less likely.
- *Amyloidosis/LCDD:* Both conditions involve protein deposition, and biopsy is the definitive diagnostic method. In this case, amyloid-associated proteins were absent, confirming the diagnosis of LCDD. LCDD is a rare disease primarily affecting the kidneys, but it can also involve the lungs. It is characterized by abnormal protein deposition. Isolated pulmonary involvement is rare in LCDD[5,6].
- *Mucosa-associated lymphoid tissue lymphoma:* Given the underlying Sjögren's syndrome and the presence of lung nodules, a low-grade lymphoproliferative disorder was also a possibility. However, flow cytometry of the lung biopsy showed no aberrant clonal B-cell population, ruling out lymphoma.

Ultimately, lung biopsy confirmed pulmonary LCDD, a sporadic condition in the absence of systemic dysproteinaemia.

LCDD is a rare condition that mainly affects the kidneys and is associated with multiple myeloma or monoclonal gammopathy[7]. Pulmonary LCDD can present as nodules, cystic lesions, or bronchiectasis[8]. The radiologic manifestations of pulmonary LCDD vary. Radiologic manifestations of pulmonary LCDD are heterogeneous. In a case series by Sheard et al.[9], CT imaging of 9 patients showed nodules in most patients (8/9) and cysts in all (9/9). Nodule distribution was random in half of the evaluable cases (3/6), typically 2–5 in number (4/8) and measured approximately 10 mm in diameter. Cysts were likewise randomly distributed (6/9) with an average diameter of 10 mm[9]. Similarly, Gonzalez et al.[10] reported radiographic findings in four patients, noting that most exhibited both nodules and cysts (3/4). Nodules ranged from 1 to 5 in number (3/4) and measured 10–29 mm, while cysts also measured 10–29 mm in diameter[10]. In our case, she had multiple cysts predominantly in the lower lobes and six randomly distributed nodules.

The specific cause of isolated pulmonary LCDD has yet to be determined; however, there were very few cases with confirmed pulmonary B cell clones bearing restricted immunoglobulin sequences in the CDR3 regions, which suggests antigen-driven clonal expansion[8]. Diagnosis requires biopsy, and treatment varies; some asymptomatic patients are monitored, while others may receive chemotherapy or lung transplantation.

In conclusion, pulmonary LCDD is a rare disease requiring high clinical suspicion. While imaging aids detection, a biopsy confirms the diagnosis. Management varies based on disease extent, organ involvement, and functional impact. The scarcity of cases and absence of clear guidelines highlight the need for further research.

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