Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pulmonology

Unclassifiable Fibrotic Interstitial Lung Disease Revealing Late-Onset Systemic Lupus Erythematosus in an Elderly Patient

Hassan Nagueyeh[1*], Chynez Rachid[1], Mohamed Ijim[1], Oussama Fikri[1], Lamya Amro[1]

[1]Department of Pulmonology, Ar-Razi Hospital, Mohammed VI University Hospital Center, LRMS Laboratory, Faculty of Medicine and Pharmacy of Marrakech, Cadi Ayyad University, Marrakech, Morocco

DOI: https://doi.org/10.36347/sjmcr.2026.v14i04.058 | **Received:** 27.02.2026 | **Accepted:** 17.04.2026 | **Published:** 25.04.2026

***Corresponding author:** Hassan Nagueyeh
Department of Pulmonology, Ar-Razi Hospital, Mohammed VI University Hospital Center, LRMS Laboratory, Faculty of Medicine and Pharmacy of Marrakech, Cadi Ayyad University, Marrakech, Morocco

Abstract **Case Report**

Interstitial lung disease (ILD) is an uncommon manifestation of Systemic lupus erythematosus (SLE), occurring in only 1–4% of patients, and may rarely represent the initial presentation, particularly in elderly individuals. We report a 72-year-old man with progressive exertional dyspnea and chronic dry cough without systemic or rheumatologic features. High-resolution computed tomography demonstrated fibrotic interstitial pneumonia of idiopathic fibrotic pattern, characterized by reticulations, traction bronchiectasis, subpleural micronodules, and early architectural distortion. Serologic testing revealed positive antinuclear antibodies, high-titer anti–double-stranded DNA antibodies, and weak anti-Ro52 positivity, while minor salivary gland biopsy showed focal lymphocytic sialadenitis insufficient for Sjögren syndrome. Multidisciplinary evaluation established the diagnosis of late-onset SLE presenting predominantly with fibrotic ILD. This case highlights the diagnostic challenge of lung-dominant lupus, which may mimic idiopathic fibrotic pneumonias, and underscores the importance of radiologic-immunologic correlation for early diagnosis and appropriate management.

Keywords: Lupus, fibrotic ILD, late-onset SLE, HRCT, unclassifiablepathern.

INTRODUCTION

Pulmonary involvement in systemic lupus erythematosus encompasses a widespectrum, ranging from pleuritis to acute pneumonitis, shrinking-lung syndrome, pulmonary hypertension, and interstitial lung disease (ILD). Although ILD represents only 1–4% of pulmonary manifestations in SLE, it carries significant prognostic implications because of its potential to evolve toward fibrosis [1].

The diagnostic difficulty isheightened when ILD appears as the initial manifestation of SLE, especially in elderly patients in whomse rologic abnormalities maybe atypical. Radiologically, SLE-associated ILD frequently exhibits *unclassifiable or indeterminate patterns,* making distinction from idiopathic interstitial pneumoni as particularly challenging [2].

Late-onset SLE, defined as onsetafter 50 years of age, represents 10–20% of lupus cases and is associated with fewer systemic manifestations but a higher likelihood of pulmonary involvement [3]. This article describes a case of fibrotic ILD serving as the first manifestation of late-onset SLE, illustrating the crucial role of multidisciplinary evaluation and radiologic–immunologi cintegration.

CASE PRESENTATION

Clinical Presentation

A 72-year-old man with no previous pulmonary, autoimmune, or rheumatologic history presented with several months of progressive exertional dyspnea and a persistent dry cough. The symptoms developed gradually, without fever, chest pain, hemoptysis, photosensitivity, arthralgia, Raynaud phenomenon, or cutaneous lesions suggestive of connective tissue disease. On examination, fine bibasilar crackleswere audible, and oxygen saturation remained preservd at rest. No digital clubbing, synovitis, or extrathoracic signs of systemic disease were identified.

Radiologic Evaluation

High-resolution computed tomography demonstrated diffuse intralobular and interlobular reticulations, traction bronchiectasis, subpleural micronodules, and early architectural distortion. The distribution and combination of abnormalities did not correspond to usual interstitial pneumonia, non specific interstitial pneumonia, lymphoid interstitialpneumonia, or organizing pneumonia. No pleural effusion or mediastinal lymphadenopathy was observed. Overall, the imaging findings supported a diagnosis of *fibrotic interstitial pneumonia of indeterminate pattern,* a presentation frequently described in SLE-associated ILD [4].

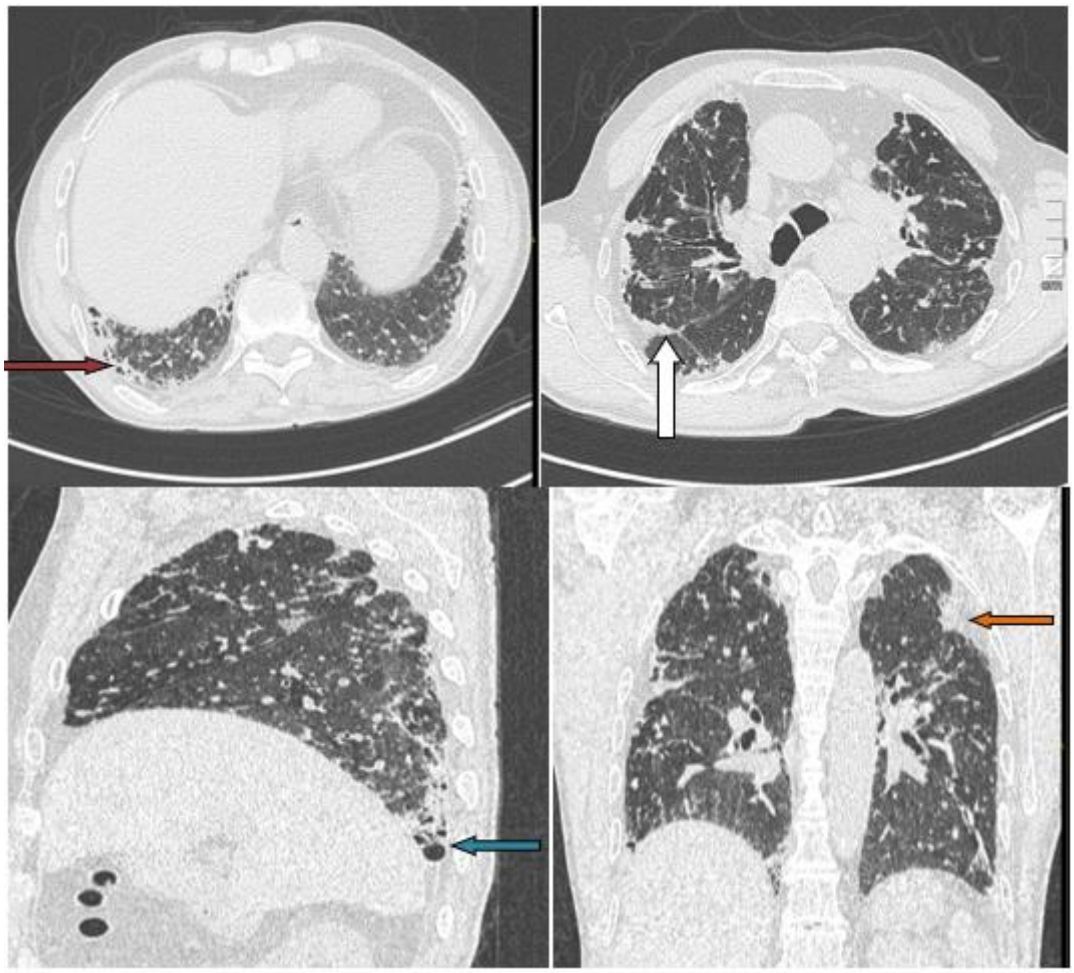

Figure 1: High-resolution CT of the chest in multiple planes demonstrating features of fibrotic interstitial lung disease associated with systemic lupus erythematosus

Laboratory and Immunologic Findings

Serologic testing revealed positive antinuclear antibodies and markedly elevated anti–double-stranded DNA antibodies,anticorps anti-nucleairs and rhumatoid facteur positif findings strongly suggestive of SLE. Weak Ro52 positivity was also present, although the remainder of the extractable nuclear antigen panel was negative. Inflammatory markers were mildly elevated, and renal, hepatic, and hematologic profiles were normal.

Because of mild sicca symptoms, a minor salivary gland biopsy was performed. Histopathologic analysis demonstrated focal lymphocytic sialadenitis (Chisholm grade II), which was insufficient to support a diagnosis of Sjögren syndrome. However, the presence of high-titer anti–double-stranded DNA antibodies and positive antinuclear antibodies provided strong immunologic evidence supporting systemic lupus erythematosus. In this context, and despite the absence of typical extrathoracic manifestations, the overall clinicoradiologic and immunologic profile was considered consistent with lupus-related interstitial lung disease.

Multidisciplinary Assessment

A multidisciplinary ILD team reviewed the clinical, radiologic, serologic, and histopathologic data. The *indeterminate ILD pattern,* combined with *high-titer anti-dsDNA antibodies* and the absence of diagnostic features of Sjögren syndrome or other connective tissue diseases, supported the diagnosis of *late-onset systemic lupus erythematosus presenting primarily with fibrotic ILD*. Given the diagnosis of lupus-associated fibrotic

interstitial lung disease, immunosuppressive therapy was initiated with intravenous cyclophosphamide (Endoxan). The patient showed a favorable clinical response, with improvement in dyspnea and stabilization of respiratory symptoms. Follow-up assessment demonstrated no further progression of fibrotic changes, supporting a good therapeutic response.

DISCUSSION

Rarity and Diagnostic Difficulty

Interstitial lung disease is a rare but clinically significant manifestation of systemic lupus erythematosus, most often occurring in younger patients with well-established multisystem disease [1]. Its presentation as the first and predominant manifestation of SLE in elderly individuals is exceptional and markedly increases diagnostic uncertainty. The present case demonstrates how lupus-related ILD may precede extrathoracic features and mimic idiopathic forms, resulting in delays in appropriate diagnosis and management.

Radiologic Features and the Challenge of Indeterminate ILD

The imaging characteristics observed in this patient exemplify the radiologic heterogeneity of SLE-associated ILD. Instead of showing a typical UIP or NSIP pattern, the HRCT demonstrated acomplex mixture of reticulations, traction bronchiectasis, and subpleural micronodules without a distinct or classic distribution. Such indeterminate patterns are well recognized in connective tissue disease–associated ILD and highlight the need for expert radiologic interpretation, as misclassification as idiopathic pulmonary fibrosismayprofoundly affect therapeutic decisions [2,4]. The present findings support the literature notingthat up to half of SLE-related ILDs may be radiologically unclassifiable.

Immunologic Contribution to Diagnosis

Serologic evaluation was pivotal in refining the diagnosis. High-titer anti-dsDNA antibodies, anticorps anti-nucleairs and rhumatoid facteur positif represent one of the most specific markers of SLE and frequently correlate with organin volvement. Weak Ro52 positivity, although nonspecific, isincreasingly associated with fibrotic autoimmune interstitial lung disease [5]. The minor salivary gland biopsy, despite demonstrating focal lymphocytic infiltration, did not fulfill criteria for Sjögren syndrome, there by eliminating a key differential diagnosis. The convergence of radiologic complexity and immunologic specificity ultimately oriented the diagnosis toward lupus-associated ILD.

Late-Onset SLE: Clinical Specificities

Systemic lupus erythematosus in older adults differs from younger-onset forms in its subtler clinical manifestations, less aggressive serologic expression, and higher frequency of pulmonary involvement [6]. These features contribute to delayed recognition, particularly when ILD is the presentingsymptom. This case demonstrates the necessity of consideringlate-onset SLE in elderly patients with unexplained fibrotic ILD and compatible serology.

Therapeutic Implications

Distinguishing between lupus-associated ILD and idiopathic pulmonary fibrosisis essential because management strategies differ markedly. Autoimmune ILD typically responds to immunosuppressive therapy, where as antifibrotic agents are prioritized in IPF. Early identification of the autoimmune etiology is therefore crucial in optimizing treatment and potentially altering disease progression.

CONCLUSION

This case illustrates a rare presentation of late-onset systemic lupus erythematosus manifesting primarily as fibrotic interstitial lung disease. The combination of indeterminate HRCT findings and lupus-specific serology underscores the importance of multidisciplinary evaluation. Clinicians should remain vigilant for lung-dominant lupus in elderly patients presenting with unexplained fibrotic ILD, as early recognition carries important therapeutic implications.

REFERENCES

1. Toyoda Y, *et al.,* Clinicalfeatures of interstitialpneumoniaassociatedwithsystemic lupus erythematosus. *Respir Investig*. 2019 ;57 :435–443.
2. Ryerson CJ, *et al.,* Prevalence and prognosis of unclassifiableinterstitiallungdisease. *EurRespir J*. 2013 ;42 :750–757.
3. Bindroo MA, *et al.,* Late-OnsetSystemic Lupus Erythematosus: ClinicalFeatures and Outcomes. *Mediterr J Rheumatol*. 2023.
4. Enomoto N, *et al.,* Analysis of systemic lupus erythematosus-relatedinterstitialpneumonia. *SciRep*. 2019 ;9 :19502.
5. Matson SM, Dilling DF. Connective Tissue Disease–AssociatedInterstitial Lung Disease. *Clin Chest Med*. 2023 ;44 :53–68.
6. Jeleniewicz R, *et al.,* Clinicalpicture of late-onsetsystemic lupus erythematosus. *Lupus*. 2015.