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Pericardial Effusion and Fibrotic Nonspecific Interstitial Pneumonia as Initial Manifestations of Primary Sjögren's Syndrome: A Case Report

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Abstract

Primary Sjogren's syndrome is a systemic autoimmune disease mainly characterized by lymphocytic involvement of exocrine glands, but extraglandular manifestations may occasionally dominate the clinical presentation. Pulmonary involvement is one of the most clinically relevant systemic manifestations, whereas pericardial involvement remains uncommon. We report the case of a 70-year-old woman with no toxic exposure who presented with a five-month history of progressive exertional dyspnea and a two-year history of subjective ocular dryness. Physical examination revealed bilateral basal crackles and a pericardial friction rub without signs of cardiac tamponade. Chest computed tomography showed a fibrotic nonspecific interstitial pneumonia pattern associated with bronchiectasis and a moderate pericardial effusion. Echocardiography confirmed a moderate circumferential pericardial effusion with preserved biventricular function and no hemodynamic compromise. Electrocardiography was normal and cardiac biomarkers were within normal limits. Immunological testing revealed positive antinuclear antibodies and anti-SSA antibodies. Ophthalmological assessment confirmed severe ocular dryness, while minor salivary gland biopsy was non-diagnostic. After exclusion of infectious, granulomatous, malignant and other systemic causes, primary Sjogren's syndrome with pulmonary involvement and probable pericarditis was retained. The patient improved under oral corticosteroids followed by mycophenolate mofetil as a steroid-sparing agent, with clinical, functional, radiological and echocardiographic improvement at one-year follow-up. This case highlights the importance of considering primary Sjogren's syndrome in patients with unexplained interstitial lung disease associated with pericardial effusion, even when sicca symptoms are limited.

Keywords: Primary Sjogren's syndrome; Interstitial lung disease; Nonspecific interstitial pneumonia; Pericardial effusion; Case Report.

Introduction

Primary Sjogren's syndrome is a chronic autoimmune disease characterized by lymphocytic infiltration of exocrine glands, classically causing ocular and oral dryness. However, it is now recognized as a systemic disease that may involve the lungs, kidneys, nervous system, vessels and, more rarely, the heart. Pulmonary disease is among the most frequent extraglandular manifestations and may include airway disease, bronchiectasis, interstitial lung disease and lymphoproliferative disorders [1-3].

Cardiac involvement is less commonly described in primary Sjogren's syndrome. Pericarditis or pericardial effusion may occur, but it is far less frequent than in other systemic autoimmune diseases such as systemic lupus erythematosus [4,5]. We report a case of primary Sjogren's syndrome revealed by fibrotic interstitial lung disease associated with moderate pericardial effusion. This case report was prepared in accordance with the CARE reporting guideline [6].

Case Presentation

A 70-year-old woman, housewife, with no history of smoking, passive smoking, tuberculosis, chronic heart disease, liver disease or kidney disease, was admitted for progressive exertional dyspnea evolving over five months. She also reported subjective ocular dryness for two years, without xerostomia or other systemic symptoms. There was no fever, cough, chest pain, weight loss or night sweats.

On admission, the patient was conscious, apyretic and hemodynamically stable. Oxygen saturation was 93% on room air, respiratory rate was 18 breaths/min, heart rate was 90 beats/min and blood pressure was 130/60 mmHg. Body mass index was 32.7 kg/m2. Chest examination revealed bilateral basal crackles. Cardiovascular examination found a pericardial friction rub, without jugular venous distension or peripheral edema. There was no parotid enlargement, palpable lymphadenopathy, skin lesion or arthritis.

Chest radiography showed bilateral basal alveolar-interstitial opacities associated with an enlarged cardiac silhouette (Figure 1). High-resolution chest computed tomography demonstrated bilateral cylindrical bronchiectasis and a fibrotic nonspecific interstitial pneumonia pattern, with ground-glass opacities, septal and non-septal thickening, subpleural reticulations, traction bronchiectasis and architectural distortion, without honeycombing. A moderate pericardial effusion was also present, without pleural effusion or mediastinal lymphadenopathy (Figure 2).

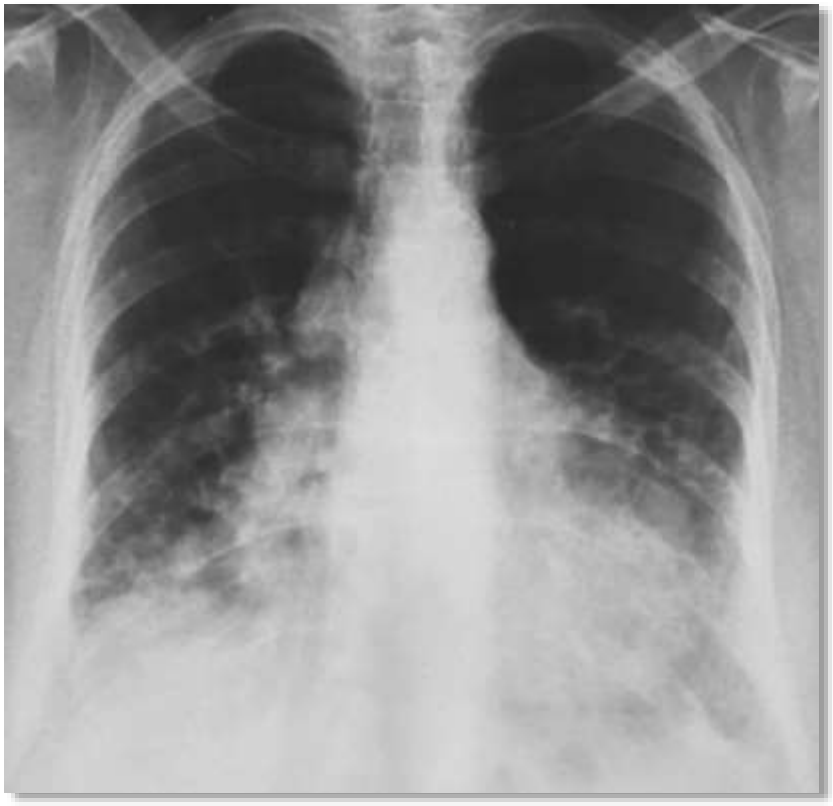

Figure 1: Chest radiograph at presentation showing bilateral basal alveolo-interstitial opacities, associated with prominent hila and an enlarged cardiac silhouette.

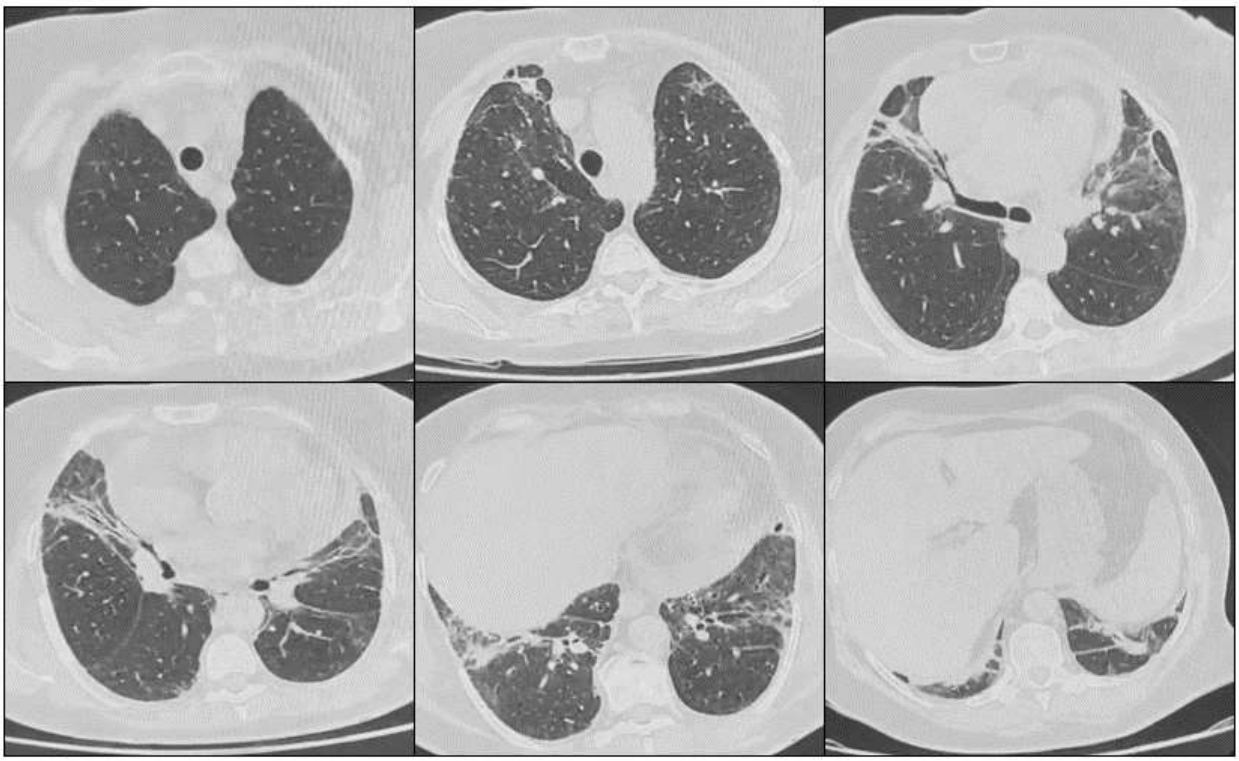

Figure 2: Initial high-resolution chest computed tomography showing bilateral cylindrical bronchiectasis and a fibrotic nonspecific interstitial pneumonia pattern, with ground-glass opacities, septal and non-septal thickening, subpleural reticulations, traction bronchiectasis and architectural distortion, associated with moderate pericardial effusion.

Transthoracic echocardiography confirmed a moderate circumferential pericardial effusion, predominantly posterior, without significant respiratory variation of transvalvular flows. Biventricular systolic function was preserved, with a left ventricular ejection fraction estimated at 62.5%. The inferior vena cava was non-dilated and compliant, and there were no indirect signs of pulmonary hypertension. Electrocardiography showed sinus rhythm without PR-segment depression, ST-segment elevation or other repolarization abnormalities, and cardiac biomarkers were within normal limits. These findings did not support concomitant myocardial involvement, although a normal electrocardiogram does not exclude pericardial inflammation [7]. Because the effusion was not safely accessible, pericardiocentesis was not performed, and close echocardiographic monitoring was chosen.

Laboratory investigations showed hemoglobin at 13.5 g/dL, white blood cell count at 7,620/microL, platelet count at 252,000/microL, erythrocyte sedimentation rate at 32 mm/h and C-reactive protein at 23.4 mg/L. Serum protein electrophoresis revealed polyclonal hypergammaglobulinemia. Renal function, urinalysis and 24-hour proteinuria were normal. Bronchoscopy showed diffuse bronchial inflammation with abundant mucoid secretions. Bronchial biopsies revealed only mild chronic nonspecific inflammation, without granuloma or malignancy. Microbiological studies, including routine cultures, GeneXpert MTB/RIF and acid-fast bacilli testing, were negative. Bronchoalveolar lavage showed marked lymphocytosis at 46% and neutrophilia at 22%.

Immunological testing showed positive antinuclear antibodies at a titer of 1:640 with a speckled pattern. Anti-SSA antibodies were positive, while rheumatoid factor, anti-cyclic citrullinated peptide antibodies and antineutrophil cytoplasmic antibodies were negative. Ophthalmological assessment confirmed severe

ocular dryness, with a reduced break-up time of 5 seconds and a Schirmer test of 4 mm in both eyes. Minor salivary gland biopsy showed chronic sialadenitis, Chisholm and Mason grade 2, with a focus score of 0. The diagnosis of primary Sjogren's syndrome was retained according to the 2016 ACR/EULAR classification criteria, based on anti-SSA positivity and an abnormal Schirmer test [1]. Alternative diagnoses were investigated and excluded, including tuberculosis, HIV and hepatitis C infections, sarcoidosis and amyloidosis. The etiological assessment of the pericardial effusion did not support heart failure, renal failure, hypothyroidism, infection, malignancy or another systemic autoimmune disease. The final diagnosis was primary Sjogren's syndrome with broncho-pulmonary involvement presenting as fibrotic nonspecific interstitial pneumonia and bronchiectasis, associated with probable Sjögren-related pericarditis revealed by a pericardial friction rub and moderate pericardial effusion.

Pulmonary function tests showed a probable restrictive ventilatory defect, with forced vital capacity at 65% of predicted. Arterial blood gas analysis showed pH 7.40, PaO2 75 mmHg and PaCO2 42 mmHg. The six-minute walk test showed desaturation to 89% at the third minute, with a walking distance corresponding to 78.5% of the predicted value.

After multidisciplinary discussion, oral corticosteroid therapy was initiated at 0.5 mg/kg/day, corresponding to prednisone 40 mg/day. A newly diagnosed type 2 diabetes was managed with metformin and close glycemic monitoring. After one month, dyspnea improved from mMRC grade II to grade I, and echocardiography showed marked regression of the pericardial effusion. Mycophenolate mofetil was then introduced progressively as a steroid-sparing agent.

At one-year follow-up, the patient was asymptomatic under prednisone 7.5 mg/day and mycophenolate mofetil 2 g/day. ESSPRI was 0 and ESSDAI decreased from 11 to 5. Forced vital capacity improved from 65% to 95% of predicted. The six-minute walk test was completed without desaturation, with a walking distance corresponding to 95% of the predicted value. C-reactive protein normalized, serum protein electrophoresis returned to normal, and renal and hepatic tests remained normal. Follow-up chest computed tomography showed marked regression of bilateral interstitial abnormalities, mainly ground-glass opacities and septal thickening, without progression of fibrosis. Bronchiectasis and traction bronchiolectasis persisted. The marked functional improvement was consistent with regression of a substantial inflammatory and potentially reversible component under corticosteroids and mycophenolate mofetil, whereas the persistent bronchiectatic and traction-related changes represented residual structural lesions. There was no residual pleural or pericardial effusion (Figure 3).

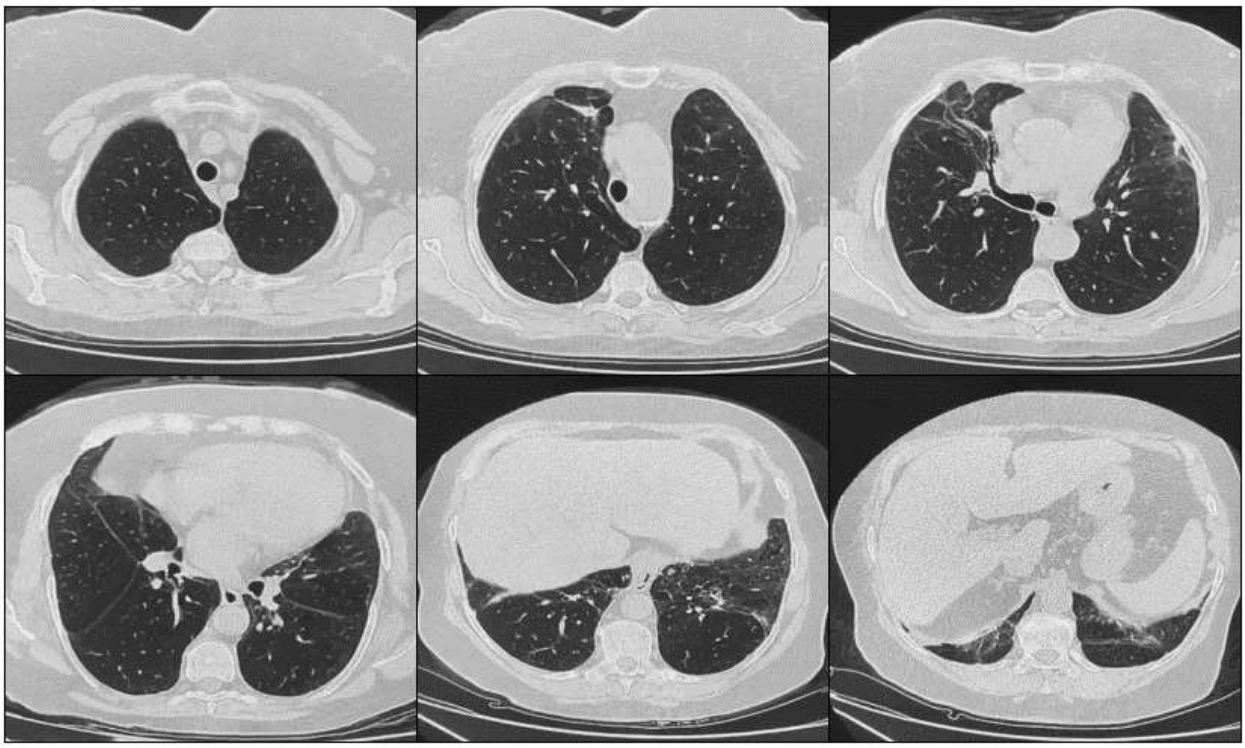

Figure 3: Follow-up high-resolution chest computed tomography after corticosteroid therapy and mycophenolate mofetil showing marked regression of bilateral interstitial abnormalities, particularly ground-glass opacities and septal thickening, without progression of fibrosis. Persistent bronchiectasis and traction bronchiolectasis are noted, with disappearance of the pericardial effusion.

Timeline

Table 1. Timeline of the clinical course according to the CARE framework.

Time point	Clinical events, investigations and interventions
Two years before admission	Onset of subjective ocular dryness without xerostomia or other systemic symptoms.
Five months before admission	Progressive exertional dyspnea developed and gradually worsened.
At admission	SpO2 was 93% on room air. Bilateral basal crackles and a pericardial friction rub were present. Chest radiography and high-resolution CT showed a fibrotic nonspecific interstitial pneumonia pattern, bronchiectasis and a moderate pericardial effusion.
Initial diagnostic assessment	Echocardiography confirmed a moderate circumferential pericardial effusion without tamponade or ventricular dysfunction. ECG was normal and cardiac biomarkers were within normal limits. ANA and anti-SSA antibodies were positive; severe ocular dryness was objectively confirmed, while minor salivary gland biopsy was non-diagnostic. Infectious, granulomatous, malignant, metabolic and other systemic causes were

Time point	Clinical events, investigations and interventions
	excluded. Primary Sjogren's syndrome with pulmonary involvement and probable Sjogren-related pericarditis was retained.
Treatment initiation	Prednisone 40 mg/day (0.5 mg/kg/day) was started after multidisciplinary discussion.
One-month assessment	Dyspnea improved from mMRC grade II to grade I, and echocardiography showed marked regression of the pericardial effusion. Mycophenolate mofetil was progressively introduced as a steroid-sparing agent.
One-year follow-up	The patient was asymptomatic under prednisone 7.5 mg/day and mycophenolate mofetil 2 g/day. FVC improved from 65% to 95% of predicted, exercise-induced desaturation resolved, inflammatory CT abnormalities markedly regressed and the pericardial effusion disappeared. Bronchiectasis and traction bronchiolectasis persisted as residual structural lesions.

Discussion

This case illustrates an uncommon presentation of primary Sjogren's syndrome in which respiratory and pericardial manifestations led to the diagnosis. Although sicca symptoms are the classical clinical hallmark, they may be mild, subjective or incomplete, particularly in patients whose extraglandular involvement is prominent. In our patient, ocular dryness was present but isolated, and the minor salivary gland biopsy was not diagnostic. The association of anti-SSA antibodies and objective ocular dryness nevertheless fulfilled the 2016 ACR/EULAR classification criteria [1].

Pulmonary involvement in primary Sjogren's syndrome is heterogeneous and includes both airway and interstitial abnormalities. Nonspecific interstitial pneumonia is among the most frequently reported radiological and histopathological patterns [2,3,8]. The CT pattern in our patient, combining ground-glass opacities, reticulations, traction bronchiectasis and absence of honeycombing, was compatible with fibrotic nonspecific interstitial pneumonia. Bronchiectasis may also be part of Sjogren-related airway disease and may coexist with interstitial lung disease. Recent longitudinal data also emphasize the heterogeneity of functional trajectories in Sjogren-associated interstitial lung disease, ranging from stability or improvement to progressive decline [9].

The main originality of this observation is the association with pericardial effusion. Cardiac manifestations are less commonly reported in primary Sjogren's syndrome than in other connective tissue diseases, and the published evidence remains largely based on case reports and small series [4,5]. In the present case, the pericardial friction rub and moderate effusion supported pericardial inflammation, while the absence of hemodynamic compromise, a normal electrocardiogram and normal cardiac biomarkers argued against significant myocardial involvement. Normal electrocardiographic and biomarker findings do not, however, exclude pericarditis [7]. The exclusion of alternative causes and the favorable response to corticosteroids supported probable autoimmune pericarditis related to primary Sjogren's syndrome. Because pericardial fluid analysis and cardiac magnetic resonance were not available, the causal attribution was considered probable rather than definitive.

The therapeutic approach was guided by the systemic activity and pulmonary involvement. Corticosteroids are commonly used for inflammatory interstitial lung disease associated with Sjogren's syndrome, while mycophenolate mofetil or azathioprine may be used as steroid-sparing agents in clinically significant or progressive disease [2,10]. The recent ERS/EULAR guideline for connective tissue disease-associated interstitial lung disease reinforces a multidisciplinary, individualized approach integrating symptoms, pulmonary function, imaging activity and disease progression [11]. In our patient, the striking improvement in forced vital capacity, exercise tolerance and ground-glass and septal abnormalities was interpreted as treatment response of a substantial inflammatory and reversible component. By contrast, the persistence of bronchiectasis and traction bronchiolectasis was consistent with residual irreversible structural damage.

This case also emphasizes the importance of long-term follow-up. Patients with primary Sjogren's syndrome, particularly those with systemic involvement and B-cell activation markers such as hypergammaglobulinemia, require surveillance for lymphoproliferative complications.

Conclusion

Primary Sjogren's syndrome should be considered in the etiological work-up of unexplained interstitial lung disease, even when sicca symptoms are mild or incomplete. The coexistence of interstitial lung disease and pericardial effusion should prompt a systematic search for autoimmune disease after exclusion of infectious, malignant, metabolic and granulomatous causes. In the absence of definitive pericardial sampling or tissue characterization, autoimmune pericarditis should be reported with appropriate diagnostic caution. Early multidisciplinary management may allow favorable clinical, functional, radiological and echocardiographic outcomes.

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