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(CASE REPORT)

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Diffuse interstitial pneumonitis as a revealing manifestation of systemic lupus erythematosus: A case report

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Abstract

Pulmonary fibrosis is a rare manifestation of systemic lupus erythematosus (SLE), occurring in 3 to 9% of cases, and is rarely the initial presenting symptom of the disease. Its pathophysiology remains poorly understood, and its clinical and radiological presentation resembles that of other connective tissue diseases. It is often associated with multi-organ involvement.

The article presents the case of a 64-year-old woman with no significant medical history, hospitalized for stage III dyspnea (according to Sadoul), dry cough, and fatigue, all evolving over six years. Her medical history revealed photosensitivity and inflammatory polyarthralgia of the large joints. Biological tests showed an inflammatory syndrome, leukopenia, and positive antinuclear and anti-DNA antibodies, which led to the diagnosis of systemic lupus erythematosus (SLE) based on the ACR criteria (photosensitivity, lymphopenia, positive antinuclear antibodies, and positive anti-DNA antibodies).

Imaging studies (chest X-ray and CT scan) revealed bilateral diffuse pulmonary fibrosis, with a honeycomb appearance and thickening of the septal lines. Pulmonary function tests showed a restrictive syndrome. The diagnosis of pulmonary fibrosis associated with SLE was confirmed. The patient was treated with corticosteroids and an immunosuppressant, with stable disease progression.

In summary, this case illustrates the complexity of diagnosing pulmonary fibrosis in the context of systemic lupus erythematosus, a rare but serious condition that requires a multi-faceted approach to management.

Keywords: Systemic Lupus Erythematosus; Interstitial Lung Involvement; Corticotherapy; Immunosuppressant; Common Interstitial Pneumonia

1. Introduction

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease of unknown etiology characterized by a multi-system connective tissue disorder that presents with variable clinical manifestations. These manifestations can include cutaneous, musculoskeletal, renal, neurological, hematological, and pulmonary involvement. The presence of interstitial lung disease (ILD) in SLE is relatively rare, and it is even rarer for ILD to be the sole initial manifestation of SLE without involvement of other organs. We describe here the case of a patient with SLE who presented with ILD as the only manifestation of the disease, without involvement of other organs.

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2. Case Report

A 64-year-old woman with no significant medical history was hospitalized for stage III dyspnea according to Sadoul, progressively developing over 6 years, associated with a dry cough and a sensation of chest tightness. The patient also reported extra-thoracic signs, including photosensitivity, xerophthalmia with xerostomia, and inflammatory polyarthralgias of the large joints. This was all occurring in the context of apyrexia and a decline in general condition. Saturation was at 97%, and bilateral basithoracic crepitations were noted during the clinical examination, primarily at the lung bases, along with digital clubbing. The chest X-ray (Figure A) revealed a bilateral interstitial syndrome consisting of diffuse bilateral reticular and micronodular opacities, along with free pleural effusions.

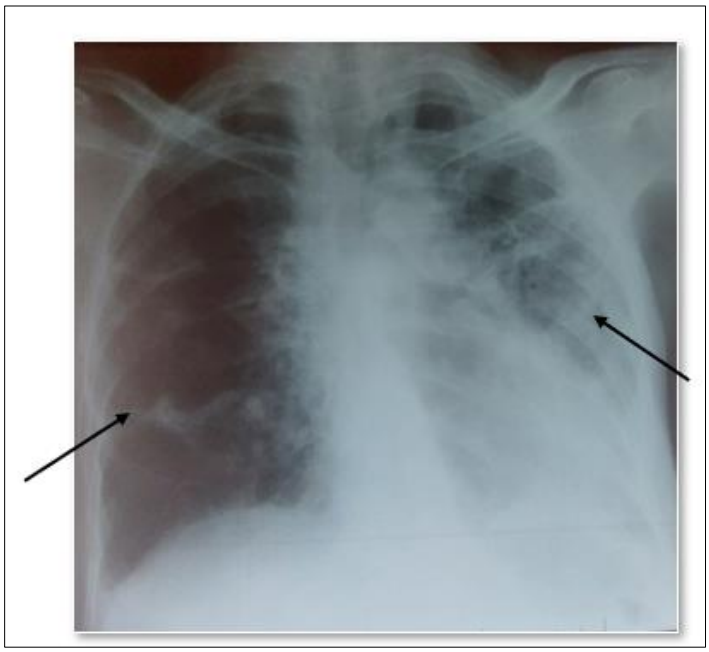

Figure 1 Chest X-ray: bilateral interstitial syndrome

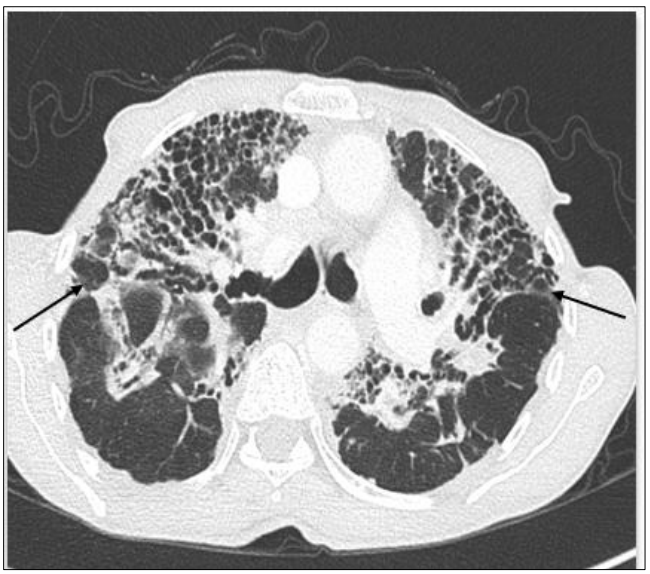

Figure 2 Axial chest CT scan: multiple microcystic images creating a honeycomb appearance with thickening of the septal lines, suggesting bilateral diffuse interstitial fibrosis

A high-resolution chest CT scan (Figure B) highlighted a honeycomb appearance visible in the middle lobe and the lingula, as well as in the left posterobasal region, with architectural distortion and traction bronchiectasis, bilateral thickening of the septal lines, and thickening of the subpleural fissure associated with a honeycomb appearance extending to the anterior portions of the upper lobes, along with non-systematized basal right ground-glass opacities creating a stable crazy paving appearance on both inspiration and expiration.

The biological assessment revealed an inflammatory syndrome (sedimentation rate of 82 in the first hour, CRP at 10 mg/dl, and leukopenia at 2000 cells/mm^3). The immunological evaluation showed positive antinuclear antibodies at a titer of 1/1600, along with positive anti-DNA antibodies. Bronchoscopy was performed, yielding staged bronchial biopsies on the right and left, which showed chronic non-specific inflammatory changes. Additionally, a bronchoalveolar lavage demonstrated inflammatory bronchial cytology with lymphocytic predominance and no signs of malignancy. A biopsy of the salivary glands revealed subacute and chronic non-specific sialadenitis grade II, with a normal Schirmer test. A cardiac ultrasound indicated moderate pulmonary arterial hypertension. The diagnosis of systemic lupus erythematosus was established based on the presence of 4 ACR criteria (photosensitivity, lymphopenia, positive antinuclear antibodies, and positive anti-DNA antibodies). Respiratory functional exploration revealed a restrictive syndrome associated with a diffusion impairment of CO at 40%. The patient was started on corticosteroid therapy at a dose of 1 mg/kg/day, combined with an immunosuppressant (Endoxan 1g per month for 6 months), with stable progression.

3. Discussion

Systemic lupus erythematosus (SLE) is one of the most common autoimmune diseases, primarily affecting young women of childbearing age. Pulmonary involvement is common in SLE and occurs in up to 50% of patients; however, in only 2 to 3% of cases, it is the initial manifestation. The prevalence and severity of diffuse infiltrative lung disease (DILD) appear to be lower in SLE than in other connective tissue diseases [1].

Pulmonary manifestations of systemic lupus erythematosus (SLE) include pleurisy, diffuse infiltrative lung diseases (DILD), pulmonary hypertension, shrinking lung syndrome, and alveolar hemorrhage. It is important to distinguish respiratory symptoms from infection, particularly in patients undergoing immunosuppressive treatment. Additionally, the risk of thromboembolic events is increased in individuals with antiphospholipid antibodies or lupus anticoagulant. The true prevalence of interstitial lung disease associated with chronic disseminated lupus erythematosus (SLE-ILD) is not well established, but prevalences of 3 to 9% have been reported in the literature. Non-specific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), organized pneumonia (formerly known as BOOP: bronchiolitis obliterans organizing pneumonia), lymphocytic interstitial pneumonia (LIP), follicular bronchiolitis, and nodular lymphoid hyperplasia have all been reported in association with SLE. Among these, NSIP appears to be the most frequently reported pattern in patients with SLE [2].

DILD in SLE typically follows a benign course with a subacute to chronic presentation. Although the clinical characteristics of SLE-DILD are typical across all patients, they may vary somewhat depending on the specific type of DILD. Patients usually present with an insidious onset of non-productive chronic cough, dyspnea, and reduced exercise tolerance, although some may be asymptomatic [3].

None of the serological markers of SLE, such as antinuclear antibodies and anti-double-stranded DNA, have shown a good correlation with the development of DILD [4]. Some reports have indicated the diagnostic utility of other serological markers, such as anti-extractable nuclear antigen antibodies (anti-Sm, anti-ribonucleoprotein), rheumatoid factor, anti-synthetase antibodies, and creatine kinase, for assessing overlap syndromes. Although tests indicating greater disease activity, such as C-reactive protein (CRP), are associated with SLE-DILD, these laboratory values are not useful for diagnosing DILD.

Abnormalities in pulmonary function tests are observed in the majority of SLE patients in some studies [5]. A decrease in carbon monoxide diffusion capacity (DLCO) as well as abnormal chest X-rays can be detected in asymptomatic patients [6]. Pulmonary abnormalities are not correlated with immune parameters. The presence of lupus is suggested by characteristic extrapulmonary and serological manifestations. In addition to restrictive abnormalities in pulmonary function tests (PFT), a decrease in DLCO and oxygen desaturation with exercise is observed.

HRCT can be extremely useful for diagnosis, and two patterns are frequently observed: ground-glass opacities (consistent with a biopsy showing a pattern of cellular infiltration indicative of NSIP) or a reticular pattern, usually with honeycombing (consistent with a biopsy showing a fibrotic pattern of usual interstitial pneumonia [UIP]). NSIP is the

most commonly observed histopathological pattern of DILD in SLE [7]. In addition to UIP and NSIP, other patterns observed include lymphocytic interstitial pneumonia and cryptogenic organizing pneumonia (BOOP).

Corticosteroids, either alone or in combination with an additional immunomodulator, are the cornerstone of treatment. However, patients with established pulmonary fibrosis are unlikely to benefit from immunosuppressive therapy. In cases of progressive or severe disease without signs of irreversibility (pulmonary fibrosis), high-dose corticosteroid therapy is initiated in conjunction with intravenous/oral cyclophosphamide, with a gradual transition to azathioprine or mycophenolate mofetil. Immunosuppressants such as azathioprine or mycophenolate mofetil may also be used in cases of corticosteroid ineffectiveness or dependency. [8,9] Antifibrotic agents such as Nintedanib and Pirfenidone are also indicated in the treatment of fibrotic lung diseases [10].

Interstitial lung disease, as a feature of SLE without any other significant systemic involvement, represents a unique presentation, and this case provides an example.

4. Conclusion

Patients with SLE can have varied presentations, making it challenging for clinicians to diagnose the condition. Additionally, many patients may exhibit interstitial lung disease without any overt symptoms of SLE, as seen in our case. A thorough history, clinical examination, and appropriate investigations aided in establishing the diagnosis. Further research is needed to understand the mechanisms underlying interstitial lung disease in patients with SLE and how these cases can be effectively treated.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

Ethical approval was obtained.

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