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Internal Medicine

INITIAL PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS AS INTERSTITIAL LUNG DISEASE IN A MALE: A RARE CASE REPORT

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ABSTRACT Systemic lupus erythematosus (SLE) is a chronic autoimmune disease of unknown cause that can affect virtually any organ of the body. Immunologic abnormalities, especially the production of a number of antinuclear antibodies (ANA), are a prominent feature of the disease. Patients present with variable clinical features ranging from mild joint and skin involvement to life-threatening kidney, hematologic, or central nervous system involvement. The clinical heterogeneity of SLE and the lack of pathognomonic features or tests pose a diagnostic challenge for the clinician. To complicate matters, patients may present with only a few clinical features of SLE, which can resemble other autoimmune, infectious, or hematologic diseases. Here we report a case of 39-year-old male presenting with chronic pulmonary complaints and was diagnosed as SLE with CTD related ILD. At presentation, he did not had any classical symptoms of SLE and only when worked up for cause of diffuse parenchymal lung disease, he was diagnosed with SLE. He was started with systemic steroids after ruling out infection to which he responded well and was discharged.

KEYWORDS :

INTRODUCTION

Systemic lupus erythematosus is a multisystem autoimmune rheumatic disease characterized by the production of pathogenic autoantibodies. However, the disease pathogenesis is still poorly understood. The incidence of SLE complicated by interstitial lung disease (SLE-ILD) ranges from 4-10%, although some cohort studies have reported a higher prevalence. For example, in a Japanese cohort that included only hospitalized patients with SLE, 29% of patients were diagnosed with ILD. Most SLE-ILD cases are asymptomatic and are detected only incidentally using chest radiography or pulmonary functional testing. In another cohort of patients aged >50 years, 30% developed ILD, suggesting that the incidence of ILD may be higher in older patients with SLE, and hence, overall the incidence may have been underestimated (1). Existing data on ILD in Indian registries have variable findings, and the largest Indian registry on ILD available today was published in 2016, studying 1084 patients. The results showed hypersensitive pneumonitis in 47.3%, CTD-ILD in 13.9%, and idiopathic pulmonary fibrosis in 13.7%. SLE-ILD was seen in only 5 patients (out of 151 CTD-ILD patients) (2). The frequency of ILD in SLE is thus exceedingly rare, and a high proportion of SLE patients with ILD might have an underlying overlap syndrome, which always needs to be ruled out (3). SLE in males is uncommon, and SLE-ILD is rare, with its presentation as the initial symptom being exceedingly rare. Here, we report a case of a 39-year-old male with the initial presentation as SLE-ILD.

Case Report

Presentation

A 39-year-old male from West Bengal, with a background history of hypothyroidism, presented with the chief complaints of joint pain (large and small) on and off for 1 year; dry cough for 3-4 years, but continuous and worsened for 5-6 months; dyspnea on exertion for 5-7 days. He had no skin rash, oral ulcers, joint deformity, fever, alopecia, myalgia, or nail changes. He also reported no fever, headache, leg swelling, palpitation, gastrointestinal, or urinary tract symptoms. On examination in the ER, he was hypoxic, tachypneic, and tachycardiac. He was started on oxygen support, and ABG showed type 1 respiratory failure. 2D Echo showed LVEF of 60% with normal filling pressure, normal valvular function, no PAH, or RWMA. Chest X-ray showed predominant reticular opacities in the bilateral lung fields. Blood and urine cultures were sent, and he was started on broad-spectrum empirical IV antibiotics and admitted to MDCCU for further management.

Workup

CBC showed mild anemia, which was microcytic and hypochromic. LFT was within limits except for hypergammaglobulinemia. RFT, including spot urine PCR, was within limits. HRCT chest showed diffuse ground-glass changes with interstitial thickening and bronchiectasis, involving all lobes bilaterally, extensively affecting the lower lobes with bilateral mild pleural effusion, consistent with diffuse fibrotic lung disease of NSIP pattern. ESR (47 mm/hr) and CRP (88 mg/dl) were elevated. ANA by indirect immunofluorescence showed 4+ with a nuclear coarse speckled pattern, and dsDNA, ANCA were negative. ENA 15 antigen profile showed Anti-Sm and RNP positivity. RA and Anti-CCP were negative. Respiratory PCR panel and HIV (by Chemiluminescence immunoassay) were negative. BAL for Xpert MTB, pneumocystis carinii, aspergillus galactomannan, and nocardia were negative. C3 and C4 were within limits. Blood and urine cultures showed no growth after 5 days of incubation.

Diagnosis and Treatment

As per EULAR/ACR criteria 2019, this patient met two clinical criteria (joint involvement, pleural effusion) and one immunological criteria (anti-Sm positive) with a score of 17 (>10), and hence was diagnosed with systemic lupus erythematosus with probable SLE-ILD as NSIP pattern. He also did not meet the diagnostic criteria for any other connective tissue disorders, and MCTD/overlap syndrome was ruled out. With reasonable investigations, any underlying infection was ruled out, and he was started on systemic steroids. His oxygen requirement gradually decreased, and he was shifted to the ward after 4 days of ICU stay. He was mobilized in the ward and discharged after 10 days of inpatient care with minimal oxygen support required only during ambulation.

DISCUSSION

The intervening space lying between the alveolar epithelium and the pulmonary capillary endothelium is called the lung interstitium, and the disease processes involving it are called Interstitial Lung Diseases (ILD), also known as diffuse parenchymal lung diseases (DPLD). Etiological classification of ILD includes i) DPLD of known cause (drugs, environment, connective tissue disease), ii) idiopathic interstitial pneumonia, iii) granulomatous DPLD, and iv) other forms of DPLD (lymphangioleiomyomatosis, pulmonary Langerhans cell histiocytosis/histiocytosis X) (4).

The most common lung conditions in systemic lupus erythematosus (SLE) are pleuritis, pulmonary thromboembolism, diffuse alveolar hemorrhage, and, less frequently, shrinking lung syndrome. These can be classified into pleural (pleurisy or pleural effusion), lung parenchyma (interstitial lung disease or acute pneumonitis), and pulmonary vascular involvement (pulmonary hypertension, pulmonary embolism, or vasculitis) (5). The pathogenesis of SLE-associated interstitial lung disease (SLE-ILD) is complex and not well understood, but cytokines play a crucial role. Elevated cytokine levels in SLE patients with lung involvement suggest they contribute to the development of pulmonary fibrosis (5). The role of autoantibody-induced epithelial cell injury triggering downstream profibrotic pathways leading to fibrosis was also reported (1). Anti-Ro52 and anti-U1RNP antibodies are linked to SLE-ILD, with studies showing that

CD4+ T cells in the bronchoalveolar lavage fluid of ILD patients promote a Th1-like inflammatory phenotype. Anti-U1RNP antibodies (positive in our patient) lead to enhanced CD4+ T cell activity, driving the abnormal immune response, exacerbating lung damage, and increasing the risk of ILD in SLE patients (1).

The onset of SLE-ILD is usually insidious and can be asymptomatic, with cough and dyspnea being the most common symptoms (5). To diagnose SLE-associated ILD, it is important to first rule out overlap syndromes that may cause misdiagnosis. Imaging, including HRCT, is key in confirming the diagnosis. Functional tests often show restrictive lung dysfunction and reduced DLCO. Early imaging may be normal, but later scans typically reveal bibasilar opacities, infiltrates, pleural involvement, or honeycombing. The most frequent pattern in SLE-ILD is NSIP, although findings are still debated (5). It appears that pulmonary manifestations occur a few years after CTD diagnosis; however, the exact duration is uncertain, and there are few case reports with SLE-ILD as an initial presentation (5), like our patient. Treatment primarily involves the treatment of the underlying disease, and options for SLE-ILD are limited (5). It is typically reserved for patients with a non-fibrosing radiological pattern showing progression, indicated by a decrease in FVC or DLCO of at least 10%. Patients with a progressive fibrosing phenotype, like UIP or fibrotic NSIP, are less likely to respond to immunosuppressive therapy but may benefit from antifibrotic treatment if immunosuppressants fail (6). For common radiological patterns, systemic glucocorticoids like prednisone (0.51 mg/kg/day) are initially recommended (6). For mild to moderate ILD, systemic steroid + mycophenolate or azathioprine + nintedanib may be used, though evidence is limited (5) (6). For severe or progressive ILD, treatment typically starts with high-dose glucocorticoids (1 g/day of intravenous methylprednisolone for 3 days) and cyclophosphamide or rituximab + nintedanib, transitioning to azathioprine or mycophenolate after 6-12 months (6). No specific treatment exists for fibrotic lung disease in SLE, but a clinical study of nintedanib showed reduced FVC decline in patients with progressive fibrosing ILDs. Nintedanib is given at 150 mg twice a day, 12 hours apart. Lung transplantation may be the last treatment option for selected patients with advanced fibrosing ILD (6).

Follow-up should be done every 36 months, including PFTs and assessing changes in symptoms or physical exam. For patients with normal DLCO, no dyspnea, and stable PFTs for over 3 years, PFTs can be reduced to every 2 years. A new HRCT may be needed if new respiratory symptoms develop or if there is a decrease in lung volumes or DLCO. In SLE, ILD typically progresses slowly and often stabilizes over time (5). The impact of ILD on prognosis in SLE patients is unclear and may also depend on the pattern of radiological findings. A pattern of UIP will probably lead to further progression and a possibly irreversible course (5) (6).

CONCLUSION

SLE-ILD remains a rare and under-recognized pulmonary manifestation of systemic lupus erythematosus, particularly as an initial presentation. This case underscores the importance of considering SLE-ILD in patients with unexplained interstitial lung disease, even in the absence of classical SLE features. Comprehensive immunological workup and exclusion of overlap syndromes are crucial for accurate diagnosis. Given the potential for progression, especially in fibrotic NSIP patterns, early initiation of immunosuppressive therapy may alter the disease trajectory. Further studies are needed to better define the natural history, prognostic markers, and optimal treatment strategies for SLE-ILD.

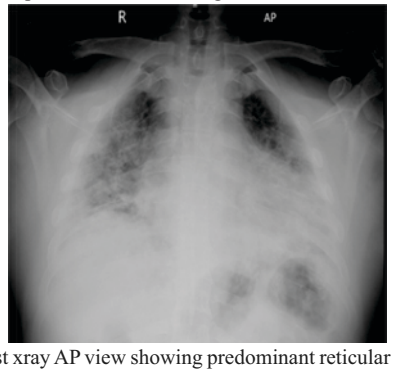

Fig 1: Chest xray AP view showing predominant reticular opacities in both lung fields

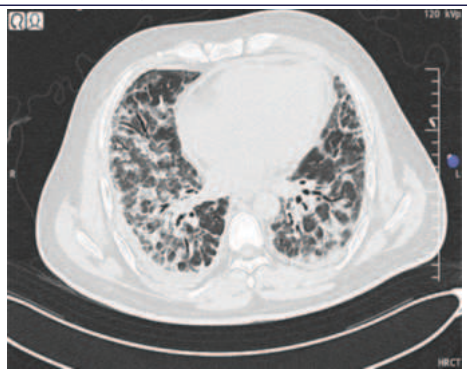

Fig 2: HRCTchest showing diffuse ground glass changes with Interstitial thickening and function bronchiectasis involving all lobes predominantly bilateral lower lobes

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