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Case Report

Systemic lupus erythematous in men with interstitial lung pneumonia: a rare case report

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease with unknown etiology and very diverse clinical manifestations, disease course, and prognosis. It is most common in African-American women, the prevalence being 4 per 1000 females. SLE predominantly affects young women, most presenting between 20 and 40 years of age. Male SLE is rare, with only one male patient reported per nine females. Pulmonary involvement in SLE is various. SLE-associated interstitial lung disease (ILD), while rare, is a predictor of poor prognosis. We report a 50-year-old male patient with complaints of shortness of breath, previously diagnosed with systemic lupus erythematous by examination of the antinuclear antibody (ANA) profile. The patient underwent a computed tomography (CT) scan of the thorax and the results showed interstitial lung pneumonia. The patient was given azathioprine and methylprednisolone therapy for Systemic lupus erythematous and ceftriaxone and acetylcysteine for interstitial lung pneumonia.

Keywords: Systemic lupus erythematous, Men, Interstitial lung pneumonia

INTRODUCTION

Multisystem involvement characterizes systemic lupus erythematous, a complicated autoimmune disease. The presentation of systemic lupus erythematosus (SLE) is varied, encompassing a wide range of clinical manifestations from mild symptoms that go away on their own to severe organ involvement that presents a serious risk to life, such as kidney failure, lung involvement, and heart involvement. Based on clinical and test results, SLE is diagnosed.[1] The presence of antibodies against cytoplasmic and nuclear antigens is a characteristic of SLE. Additionally, patients with SLE may also have other autoantibodies, such as anti-phospholipid, anti-cardiolipin, anti-La, anti-Ro, and anti-Scl-70 antibodies, suggesting a broad correlation between SLE and other autoimmune diseases. The improved classification criteria used by the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) serve as the most advanced and precise criteria to date.[1,2]

There have been reports of varying SLE prevalence and incidence rates; these variations are primarily attributable to population variances. With a 9 to 1 female-to-male ratio, women of reproductive age are primarily affected by SLE. While it still remains twice as high as in men, women's risk does drop during menopause. Research has shown that men tend to have more severe cases of lupus, even though it is rare.[3]

SLE is a multisystem illness that manifests in various ways. Clinical characteristics might range from a relatively mild illness that only affects the skin to a severe, perhaps fatal illness that affects multiple organs. In SLE, every organ system may be affected. Serological abnormalities have been shown to develop several years before the

beginning of clinical lupus in several studies. Pre-clinical lupus is the term for a condition in which a patient may exhibit some clinical symptoms and serological abnormalities suggestive of SLE, but their symptoms still don't match the criteria for the disease. Research suggests that a sizable portion of these pre-clinical lupus patients, including those with undifferentiated connective tissue disease or incomplete lupus, may eventually develop clinical lupus and meet the requirements for systemic lupus later in life.[4-6]

Patients with SLE commonly develop pulmonary involvement; incidence ranges from 24% to 74%. However, only 2–4% of patients present with pulmonary implications as their initial SLE symptom. The prognosis and quality of life of patients are affected by pulmonary involvement in addition to other systemic signs. ILD is a pulmonary symptom of multiple connective tissue diseases (CTDs) that often result in significant morbidity and mortality.[7,8] The respiratory system, which includes the airways, parenchyma, vasculature, pleura, and respiratory muscles, is one of the many organs that can be affected by CTD. One of the pulmonary symptoms of CTD that can occur is interstitial lung disease (ILD), which can raise the morbidity and death rates of those who are impacted.[7]

ILD is not frequently found in SLE cases. Even though SLE is the most prevalent CTD in the general population, it is linked to long-term illness and only occurs in 1–15% of SLE patients. This explains why it might be difficult to detect ILD-related SLE because it is frequently confused with other pulmonary manifestations of the disease. We present a case of ILD in patients with SLE here.[9]

CASE REPORT

The patient is a 50-year-old man who was hospitalized for the first time at the Sanjiwani Hospital with a complaint of shortness of breath 1 week before entering the hospital which has worsened for the past 3 days. The patient also complained of a cough and fever 10 days ago. The patient was said to have been diagnosed with SLE 1 month ago and regularly takes medication for his autoimmune disease. Before being diagnosed, the patient often felt tired for the past 3 months, his face started to turn red in the shape of a butterfly, and he was sensitive to sunlight. The patient had no history of chronic disease and long-term drug consumption. The patient works in a hotel not far from where he lives. The patient does not smoke and has no family history of autoimmune disease.

The results of the patient's examination one month ago, showed that the ANA IF was 1: 100, with a positive ANA profile, PCNA, the patient regularly took the drugs azathioprine 1×50 mg, methylprednisolone 2×8 mg, hydroxychloroquine 1×200 mg.

The patient's blood pressure is 120/79 mmHg, the pulse is 84 x/minute, the temperature is 38 ^{o}C and the patient's respiratory rate is 30 x/minute. On physical examination, the sclerae were not icteric and the conjunctiva was anemic. There is a malar rash. No enlarged lymph nodes in the neck area. No elevation in jugular venous return. Chest examination found a vesicular breath sound, there was wheezing and crackles. The left heart border is normal and the heart sounds are normal S1 and S2, with no murmurs or gallops.no enlarged liver and spleen. Normal bowel sound. The extremities were warm and there was no pitting edema in the pretibial.

A complete blood count obtained hemoglobin 17.6 gr/Dl, leukocytes 9.790/ul, platelet 279.000/ul, MCV 99.9 fL, MCH 34.9 pg, neutrophils 8.42/86%, lymphocytes 0.92/9.4%, monocytes 0.43/9.4%, eosinophils 0.02/0.2%, and basophils 0.0/0.0%. Renal function tests included urea 20.6 mg/dl and creatinine 0.35 mg/dl. Examination of liver function AST 22 U/l and ALT 34 U/l. Examination of electrolytes sodium 129 mmol/l, potassium 3.9 mmol/l, and chloride 102 mmol/l. Arterial blood gas pH 7.391 mmHg, PO_2 94 mmHg, PCO_2 35.3 mmHg, HCO_3 21.5 mmol/l, be -4 mmol/l, $AaDO_2$ 539 mmHg. Examination of chest X-ray showed pneumonia and cardiomegaly. The result of MSCT scan thorax interstitial pneumonia and bronchiectasis.

The patient was treated for 5 days in the ICU because of severe shortness of breath and low blood pressure, requiring oxygen and blood pressure support medication. While in the ICU, an evaluation blood test was carried out, which obtained arterial blood gas pH 7.435 mmHg, PO_2 140 mmHg, PCO_2 43.2 mmHg, HCO_3 28.6 mmol/l, be 4 mmol/l, and $AaDO_2$ 121 mmHg.

During treatment in the ICU, the patient's autoimmune medication was temporarily postponed, and then after the condition began to improve, the autoimmune medication was continued. For pneumonia therapy, the patient was given the antibiotic ceftriaxone, acetylcysteine drip, nebulized salbutamol, and to prevent stress ulcers, the patient was given lansoprazole 2×30 mg and sucralfate 3×15 mg/ml, and paracetamol if fever.

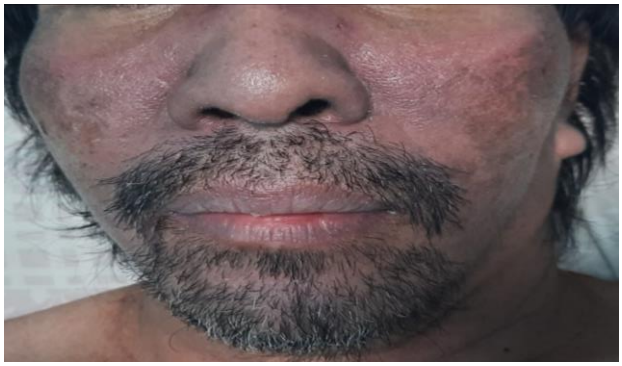

Figure 1: Malar rash on the patient face.

The patient's condition improved during treatment in the ICU and the patient was able to move to a regular treatment room after the sixth day and the autoimmune medication

was started again. After 7 days of antibiotic therapy, then had a complete blood count again with results of hemoglobin 14.3 gr/dl, leukocytes 5.190/ul, platelets 228.000/ul, MCV 103 fL, MCH 34.5 pg, neutrophils 32.7/62.9%, lymphocytes 1.58/30.4%, monocytes 0.33/6.5%, eosinophils 0.01/0.2%, and basophils 0.0/0.0%. Oxygen administration has begun to be tapered off and the patient's condition is starting to stabilize without medication support.

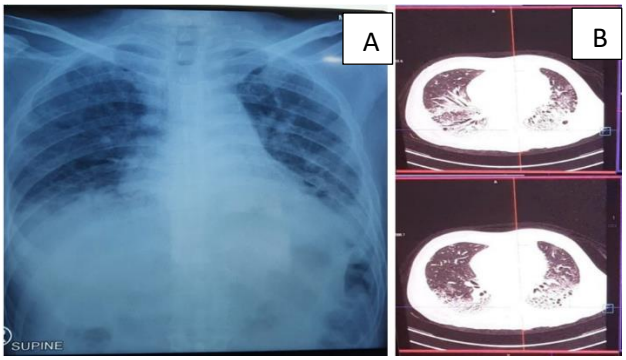

Figure 2: A) Chest X-ray of a pneumonia and B) CT scan of the thorax and results showed interstitial lung pneumonia.

DISCUSSION

The disease known as systemic lupus erythematosus, or SLE, is typically associated with fertile young women. Peak onset occurs in women between the ages of 20 and 30, whereas it is rarer in men, peaking around 45 to 60 years of age. Male SLE is rare, with only one male patient reported per nine females.[2] Men are more severely affected by the illness, and their relapses last less time. A virus infection, stress, or other variables that may excite your immune system and create an autoimmune reaction can combine with a genetic predisposition to cause lupus.[3,4]

In general, SLE in men has the same disease spectrum as in women, but with some variation. SLE manifests itself clinically in a very varied way. Arthralgia and arthritis are present in over 85% of individuals. Compared to women, men experience fewer joint and fibromyalgia symptoms. In addition to other constitutional symptoms, fever, exhaustion, headaches, and weight loss affect almost two-thirds of SLE patients. According to certain studies, SLE could be the reason for up to 50% of adult patients who come with a fever of unknown origin.[4,5,10] Peripheral neuropathy and vasculitis are more common in men. Because of antibodies against beta 2 glycoprotein 1, lupus anticoagulant, cardiolipin, and thrombin, thromboembolic illness affecting the arteries and/or veins might exacerbate SLE. Anticoagulants for lupus are more frequently detected in men.[12]

In 70% of cases, skin rashes and photosensitivity can be seen in SLE patients. The most common type, butterfly or malar rash, is less common in men. Discoid lupus is more common in men and may leave scars. Pleuritis, pulmonary thromboembolism, diffuse alveolar hemorrhage, and, less frequently, shrinking lung syndrome are the most common lung conditions in SLE patients. These different manifestations can be divided into three categories: lung parenchyma (acute pneumonitis or interstitial lung disease), pulmonary vascular involvement (pulmonary hypertension, pulmonary embolism, or vasculitis), and pleural (pleurisy or pleural effusion).[13,14]

It is unknown how SLE affects the lungs, however, the illness is known to have a high expression of type I interferon (IFN)-regulated genes. Furthermore, type 1 IFNs are essential for autoantibody synthesis and neutrophil production, which leads to immunological tolerance dysregulation in the lungs. Patients with SLE who have lung involvement also have greater levels of proinflammatory cytokines such as IFN-gamma, tumor necrosis factor-alfa (TNF-alfa), interleukin-6 (IL-6), IL-8, and IL-12.[15]

One study found that 2% to 4% of SLE patients appear to have ILD. Individuals with late-onset SLE have a higher risk of developing pulmonary events compared to younger individuals, according to a meta-analysis. One explanation could be that aging could accelerate lung immune senescence.[11] Laboratory abnormalities that suggest a potential pulmonary involvement associated with SLE include the presence of acute phase reactants in high titers, raised double-stranded DNA (dsDNA) antibody levels, and low complement levels. The proliferating-cell nuclear antigen (anti-PCNAs) laboratory autoantibodies in this patient were positive. Anti-PCNAs specifically target proteins (PCNA or cyclin) linked to the DNA polymerase enzyme. Regarding systemic lupus erythematosus, they are highly specific but have a low sensitivity.[11,16]

An overlap syndrome needs to be ruled out before the diagnosis of SLE-associated ILD may be made. This potential reason can result in an incorrect diagnosis. A high-resolution computed tomography (HR-CT) scan ought to be carried out after that. Clinical diagnosis of SLE-ILD typically involves ruling out other potential causes and employing imaging techniques like HRCT. The most common conditions found were ground glass opacities, lung consolidation, honeycombing appearance, and even traction bronchiectasis.

The differentiation of patients with idiopathic pulmonary fibrosis from those with CTD-related ILD may begin with the detection on thoracic CT of two novel distinct fibrosis patterns: the heterogenous lung destruction sign and island-like fibrosis. In patients, SLE was confirmed by examining the ANA IF and ana profile and confirming the diagnosis for ILD by carrying out an MSCT Thorax examination.[11,17,18] In this case, the diagnosis was made by examining the Ana Profile and MRCT scan and it was found that the patient had SLE with flare ILD.

In SLE, there is little management of ILD. The European League Against Rheumatism (EULAR) states that hydroxychloroquine should be the first-line treatment for all SLE patients. If required, glucocorticoids may also be added. To evaluate the safety and effectiveness of corticosteroids and immunosuppressant medications for the treatment of ILD owing to SLE.[11,12] More specifically, it is recommended to use azathioprine/MMF and corticosteroids as first-line treatment for mild and moderate forms; these medications can also be maintained as maintenance therapy.[11,13,14]

Patients with increasing and severe disease courses linked with fibrotic CTD-related ILD should be carefully evaluated for lung transplantation and antibiotic therapies. It is unknown yet whether ILD might be viewed as a risk factor for SLE patients's prognosis.[11,16]

The effectiveness of corticosteroids, despite their extensive usage in the treatment of ILD, is yet unknown. Tadaka et al reported that one patient saw a notable improvement following corticosteroid treatment. This is appropriate with this case report that patients' responses to corticosteroids was improved. Corticosteroids significantly improved the clinical condition of eleven SLE patients who were diagnosed with ILD in accordance Acharya et al. The patient was given azathioprine and methylprednisolone therapy for SLE and ceftriaxone and acetylcysteine for interstitial lung pneumonia. This is consistent with this patient's condition improving after administering antibiotic and corticosteroid therapy.[11,15]

CONCLUSION

ILD in SLE is a rare manifestation. Even though it is uncommon, ILD can potentially be detected as the first manifestation of lupus with CT imaging. This can lead to early initiation of immunosuppressive and corticosteroid therapies which help to reduce potential complications.

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