Case Report ISSN (O):2395-2822; ISSN (P):2395-2814

Interstitial Lung Disease in Systemic Lupus Erythematosis: A case report.

Nikita Malhotra[1], Santwana Chandrakar[2], Varun Shetty[3]

[1]PG student, Department of Internal Medicine, DY Patil Medical College, Navi Mumbai.
[2]Professor, Department of Internal Medicine, DY Patil Medical College, Navi Mumbai.
[3]Associate Professor, Department of Internal Medicine, DY Patil Medical College, Navi Mumbai.

Received: September 2017
Accepted: September 2017

ABSTRACT

Introduction: Systemic lupus erythematosus (SLE) is a chronic inflammatory disease of unknown etiology that can affect virtually every organ in the body. Presentation of a patient can vary considerabaly. Many a times, pleurisy, coughing, and/or dyspnea are the first clues either to lung involvement or to SLE itself. In this case report we present a patient who was diagnosised with SLE along with pulmonary diagnosis of interstitial lung diasese (ILD). **Case report**: A 40 year old female presented with swelling of both lower extremities, facial puffiness for one month and dry cough for past 3 years. Examination revealed painful joints and bilateral fine crepts at the bases of lungs. Patient's ANA and anti-double stranded DNA indicated SLE. Pulmonary function test showed restrictive pattern of lung disease compatible with interstitial lung disease (ILD), which was supported by High resolution computed tomography thorax. 2D echocardiography revealed mild tricuspid rgurgitation and pulmonary artery systolic pressure of 80 mm of mercury. Based on the clinical and laboratory findings present in this patient, a final diagnosis of SLE with ILD and pulmonary hypertension was made. Treatment included immunosuppresive therapy, mycophenolate mofetil, pirfenidone and sildenafil. Mild improvement of respiratory symptoms was noted in the patient. **Conclusion**: Careful history taking, clinical examination and appropriate clinical investigations helped us in clinching the diagnosis. Further research is required to understand the mechanisms responsible for ILD in SLE patients and how these cases can be treated effectively.

Keywords: Diagnosis, immunotherapy, systemic lupus erythematosis, interstitial lung disease.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease of unknown etiology that can affect virtually every organ in the body. A prominent feature of the disease is the production of a large number of antinuclear antibodies. Epidemiological studies have shown that women are affected more frequently than men. Presentation of a patient can vary considerabaly. At some time during the course of the disease, many patients with systemic lupus erythematosus (SLE) may show signs of involvement of the lung, pulmonary vasculature, pleura, and/or diaphragm.[1] Many a times, pleurisy, coughing, and/or dyspnea are the first clues either to lung involvement or to SLE itself.[2] Even in some asymptomatic cases, abnormal pulmonary function tests (PFTs) and/ or chest radiographs may be detected in patients.[3] Patients with SLE and lung involvement must always be evaluated for infection, particularly due to bacteria or viruses.[4] In this case report we present a patient who presented to us with a long history of exertional dyspnea. Clinical, physical and laboratory examination pointed towards the diagnosis of SLE along with pulmonary diagnosis of interstitial lung diasese (ILD).

Name & Address of Corresponding Author
Dr. Nikita Malhotra,
PG student
Department of Internal Medicine,
DY Patil Medical College,
Navi Mumbai.

CASE REPORT

A 40 year old female presented with swelling of both lower extremities, facial puffiness for one month and decreased urination for ten days. Before the appearance of these symptoms, the patients had no such complaints. Patient's past medical history included dry cough for past 3 years, along with some exertional dyspnea. For past 8 months patient has been complaining of pain in large and small joints all over the body. Patient had no past history of orthopnoea, paroxysmal nocturnal, fever, rash, photosensitivity, oral ulcers or weight change. General examination of the patient revealed pallor and pitting edema in lower extremities.Vitals of the patient were within normal limits. On respiratory system examination, we found bilateral fine crepts,

mainly at the bases of lungs. Other systemic examinations were with in normal limits. Laboratory investigations revealed normal hemoglobin, total leucocyet count, platelet count, liver function tests, serum urea and creatinie levels [Table 1]. Patient's liver enzymes were with in normal range as well. Urinalysis revealed 3+ albumin and granular casts. 24 hour urinary protein excretion was 3.8 gm. Chest x ray was unremarkable for the patient. Laboratory tests for SLE were sent, which included ANA, which was 3+ (1:1000 dilution) and anti-double stranded DNA was 434 u/ml. Pulmonary function test showed restrictive pattern of lung disease compatible with interstitial lung disease [Table 2].

Table 1. Clinical investigations in our patient

Investigation	**Value (range)**
Hemoglobin	9.4 gm% (12-15gm%)
Total Leucocyte count	3800/cumm (4000-11000)
Platelet count	1.8 L/cumm (1.5-4 L/cumm)
Erythrocyte sedimentation rate	100 mm/hr (0-20)
Bilirubin direct	0.2 U/L (0-1)
Bilirubin indirect	0.4 U/L (0-1)
Aspartate aminotransferase	14 U/L (12-32)
Alanine aminotransferase	22 U/L (4-36)
Prothrombin time (INR)	12 seconds (11-13.5)
Serum albumin	4.8 gm/dL (4-5.5)
Serum urea	37 mg% (15-50mg%)
Serum creatinine	0.9 mg% (0.2-0.8)
Urinalysis	
Casts	**Granular**
24 hour protein excretion	3.8 gm (less than 30 mg)
Anti-nuclear antibody	3+
Anti-double stranded DNA	434 U/mL

Table 2: Pulmonary investigations in the patient

Investigation	**Finding**
Pulmonary function test	
Total lung capacity	Decreased
Forced residual capacity	Decreased
Ratio of forced expiratory volume in one second & forced vital capacity	Increased
Imaging studies	
Chest x-ray	Unremarkable
2D echocardiography	Tricuspid regurgitation, pulmonary artery systolic pressure 80 mm Hg
High resolution CT	Diffuse bilateral ground glass opacification and interlobular septal thickening in basal lung fields

High resolution computed tomography thorax revealed diffuse bilateral ground glass opacification and interlobular septal thickening in basal lung fields, which were suggestive of interstitial lung disease [Figure 1]. Two-dimensional echocardiography revealed mild tricuspid rgurgitation, ejection fraction od 55% and pulmonary artery systolic pressure of 80 mm of mercury. Based on the clinical and laboratory findings present in this patient, a final diagnosis of SLE was made. Patient was started on immunosuppresive therapy and mycophenolate mofetil. For pulmonary arterial hypertension pirfenidone and sildenafil was started with follow up every month. Mild improvement of respiratory symptoms was noted in the patient. Because of the adverse effects assicated with long term systemic steoid therapy, we advised steroid sparing immunosuppresive therapy. Supportive care in the form of supplemental oxygen and vaccination against streptococcus pneumonia was advised.

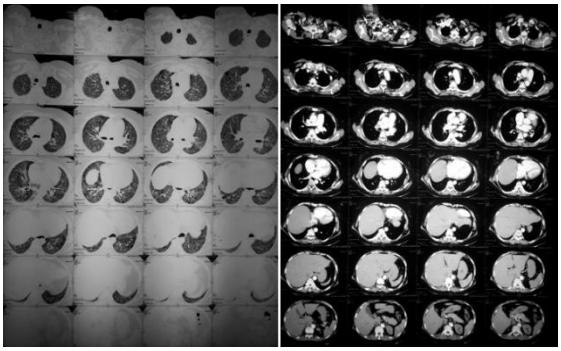

Figure 1: High resolution computed tomography of the patient.

DISCUSSION

Pulmonary manifestations of SLE include pleuritis, pneumonitis, ILD, pulmonary hypertension, shrinking lung syndrome, and alveolar hemorrhage. It is important to distinguish respiratory symptoms from infection, particularly in patients on immunosuppressive therapy. Moreover, the risk of thromboembolic involvement is increased in those with antiphospholipid antibodies or with lupus anticoagulant. True prevalence of chronic systemic lupus erythematosus-associated interstitial lung disease (SLE-ILD) is not known, but prevalences of 3 to 9% have been reported in the literature. Nonspecific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), organizing pneumonia (formerly called 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 common reported pattern in SLE patients,[6] while the endstage fibrotic pattern of usual interstitial pneumonia is relatively uncommon. Although the clinical characteristic sof SLE-ILD are typical in all patients, it may vary somewhat with the particular type of ILD. The patients usually present with an insidious onset of chronic nonproductive cough, dyspnea, and decreased exercise tolerance, but some may be asymptomatic.[7]

None of the serologic markers for SLE like antinuclear antibody, anti-double stranded DNA has shown a good correlation with development of ILD.[8] Some reports have shown the diagnostic usefulness of other serologic markers, such as anti-extractable nuclear antigen antibodies (anti-Sm, anti-

ribonucleoprotein), rheumatoid factor, antisynthetase antibodies, and creatine kinase to evaluate for overlap syndromes. Although tests that indicate greater disease activity, like C-reactive protein (CRP), are associated with SLE-ILD, but these laboratory values are are not helpful in diagnosing ILD . Chest radiographs may reveal patterns of ILD, but high resolution computed tomography (HRCT) is the standard diagnostic imaging procedure for determining the presence of ILD and defining pattern of ILD, which may further correlate with a specific histopathologic diagnosis.

Due to the scarcity of cases reported in the literature and absence of controlled trials of treatment the optimal therapy for chronic SLE-ILD is not known. For most presentations of SLE-ILD systemic glucocorticoids are the first line agents, such as prednisone 0.5 to 1 mg/kg per day. In a case series of 14 patients with SLE-ILD, prednisone 60 mg a day followed by gradual tapering was effective in treating the disease.[9] It was shown that respiratory symptoms and diffusing capacity for carbon monoxide improved in majority of patients, while forced vital capacity was unchanged. Death of three patients was reported in the case series, two from progressive lung fibrosis and one due to bacterial pneumonia. In another series of 38 patients, there was no improvement in respiratory parameters with glucocorticoid treatment.[10] Therefore, conflicting evidence exists regarding the usefulness of systemic steroids in these patients. Although data in SLE patients are limited, steroid sparing agents like cyclophosphamide, azathioprine, mycophenolate, rituximab have been used in the past.[11]

CONCLUSION

Patients of SLE may have have varied presentations. It can be challenging for a clinician to diagnose SLE. Moreover, many patients can present with ILD without any overt symptoms of SLE, like in our case. Careful history taking, clinical examination and appropriate clinical investigations helped us in clinching the diagnosis. Further research is required to understand the mechanisms responsible for ILD in SLE patients and how these cases can be treated effectively.

REFERENCES

1. Kim JS, Lee KS, Koh EM, et al. Thoracic involvement of systemic lupus erythematosus: clinical, pathologic, and radiologic findings. J Comput Assist Tomogr 2000; 24:9.
2. Hellman DB, Kirsch CM, Whiting-O'Keefe Q, et al. Dyspnea in ambulatory patients with SLE: prevalence, severity, and correlation with incremental exercise testing. J Rheumatol 1995; 22:455.
3. Nakano M, Hasegawa H, Takada T, et al. Pulmonary diffusion capacity in patients with systemic lupus erythematosus. Respirology 2002; 7:45.
4. Rojas-Serrano J, Pedroza J, Regalado J, et al. High prevalence of infections in patients with systemic lupus erythematosus and pulmonary haemorrhage. Lupus 2008; 17:295.
5. Wiedemann HP, Matthay RA. Pulmonary manifestations of systemic lupus erythematosus. J Thorac Imaging 1992; 7:1.
6. Tansey D, Wells AU, Colby TV, et al. Variations in histological patterns of interstitial pneumonia between connective tissue disorders and their relationship to prognosis. Histopathology 2004; 44:585.
7. Pope J. An update in pulmonary hypertension in systemic lupus erythematosus - do we need to know about it? Lupus 2008; 17:274.
8. Yee CS, Hussein H, Skan J, et al. Association of damage with autoantibody profile, age, race, sex and disease duration in systemic lupus erythematosus. Rheumatology (Oxford) 2003; 42:276.
9. Weinrib L, Sharma OP, Quismorio FP Jr. A long-term study of interstitial lung disease in systemic lupus erythematosus. Semin Arthritis Rheum 1990; 20:48.
10. Nakano M, Hasegawa H, Takada T, et al. Pulmonary diffusion capacity in patients with systemic lupus erythematosus. Respirology 2002; 7:45.
11. Lim SW, Gillis D, Smith W, et al. Rituximab use in systemic lupus erythematosus pneumonitis and a review of current reports. Intern Med J 2006; 36:260

How to cite this article: Malhotra N, Chandrakar S, Shetty V. Interstitial Lung Disease in Systemic Lupus Erythematosis: A case report. Ann. Int. Med. Den. Res. 2017; 3(6): ME14-ME16.

Source of Support: Nil, **Conflict of Interest:** None declared