Open Journal of
Clinical & Medical
Case Reports

Volume 3 (2017)
Issue 25

ISSN 2379-1039

Systemic lupus erythematosus with shortness of breath

***Geetha Wickrematilake, MBBS, MD, MRCP(UK)**

Consultant Rheumatologist, Rehabilitation Hospital, Digana, Sri Lanka
Email: gayaniw75@yahoo.com

Abstract

A 42-year-old lady with Systemic lupus erythematosus and secondary anti-phospholipid syndrome presented with repeated episodes of shortness of breath. Her lung auscultation was normal and chest Xray, ECG findings, and the wells score were normal, thus getting discharged from the emergency department several times. Her echocardiogram revealed moderate pulmonary hypertension and a CTPA revealed acute episodes of pulmonary emboli. She was anti-coagulated. The intensity of shortness of breath improved but did not completely disappear. Her repeated chest X-rays were compaired to see reduced lung volumes bilaterally. Her HRCT scan only revealed bilateral basal atelectasis but otherwise normal lung parenchyma and there was restriction in lung function tests. A diagnosis of shrinking lung syndrome with coexisting pulmonary emboli leading to pulmonary hypertension was made.

She was treated with methylprednisolone pulses and was started on oral steroids cyclophosphamide and theophylline. Hydroxychloroquine and warfarin were continued. Patient remains stable in lung function tests.

Keywords

anti-phospholipid syndrome; shrinking lung; pulmonary embolism; systemic lupus erythematosus

Introduction

Hoffbrand and Beck first described in 1965 in a case series about patients who developed "unexplained breathlessness" with progressively lowering lung volumes and restrictive ventilatory deficits [1]. This triad of elevated hemidiaphragms, normal lung parenchyma, and restrictive deficits on lung function testing in patients with systemic lupus are the universally described components of shrinking lung syndrome (SLS) [2].

This is an uncommon pulmonary manifestation of SLE, although the exact prevalence is not known. SLS and Pulmonary embolism(PE) in SLE with secondary APL syndrome have not been reported so far.

Case Report

A 42-year-old lady was referred to the Rheumatology clinic with bilateral knee joint effusions and all proximal interphalangeal joint swelling. Her rheumatoid factor was positive (250 iu/L) and the erythrocyte sedimentation rate (ESR) was also high- been 130mm/1st hour. She was already on methotrexate therapy started by a general physician for a diagnosis of Rheumatoid arthritis.

After few weeks, patient presented with a mild right sided upper motor neuron facial nerve palsy which improved within 24 hrs. Her blood sugar level was normal been 76mg/dl and lipids were not high. - Total cholesterol 201mg/dl and LDL -99 mg/dl. The blood pressure was 170/80Hgmm. Her renal ultrasound scan was normal and the inflammatory markers remained high. However, a week later patient was admitted to the medical ward with an episode of right upper motor neuron facial palsy, which this time had persisted for 72 days by the time she presented to the hospital. Her brain CT scan revealed an ischaemic infarct involving the left parietal region. At that time her blood pressure was elevated and the cardiac echocardiogram, Doppler neck arteries were normal with a negative antinuclear antibody (ANA) test. She was started on losartan and lipid lowering statins, and discharged.

Her inflammatory markers were repeatedly high (ESR 110mm, ESR 108mm) without any focus of infection or any significant joint involvement. Patient started complaining of excessive hair loss with a vague history of photosensitivity and her repeated ANA became positive in a speckled patternwith a titer of 1/320 and positive Ds-DNA levels. There was lymphocytopenia, (910/mm^3). Anti-cyclic citrullinated peptide (anti-CCP) antibody was negative and the screening for lupus anticoagulant was positive with a positive APTT, and DRVVT. The anticardiolipin antibodies IgG, IgM and Beta-2 glycoprotein 1were negative. Lupus anticoagulant test remained positive 12 weeks later.

Few months later patient started to complain central chest pains and shortness of breath. There was no history of fever, cough or haemoptysis. She had repeated visits to the hospital emergency department and was discharged after chest imaging, blood tests and ECG.

Clinical examination was not significant except for tachycardia of 110 beats per minute. Her wells score was 1.5 with a low risk for pulmonary embolism (PE). However, there was mild hypoxemia in blood gases, the dimer was elevated 0.6mg/L (<0.46mg/L) and the CTPA revealed bilateral multiple pulmonary emboli. She was anti-coagulated with heparin and later on converted to warfarin. The Echo cardiogram revealed moderate pulmonary hypertension and the leg venous Doppler was negative. Even a month later the patient did not significantly improve and the lung function tests revealed restrictive pattern, (FEV1/VC ratio was 110.5%) with normal parenchyma in HRCT except basal atelectasis (Figure 1).

Her blood gases were normal and repeated chest X-rays when compared revealed significant lung volume loss (Figure 2, 3 and 4) qualifying for shrinking lung syndrome although ultrasound scan of the abdomen reveled satisfactory diaphragmatic movements.

She was initially pulsed with methyl prednisolone, and started on steroids (1mg/kg), cyclophosphamide. Her warfarin and hydroxychloroquine was continued. Theophylline was added together with beta 2 agonists, and now she remains clinically stable.

Discussion

The type of pulmonary impairment in SLE varies widely. Possibly the least common manifestation is the SLS. The pathogenesis of SLS is under debate. Various pathogenetic mechanisms including myositis of the diaphragm, phrenic nerve paresis, restrictive rib cage abnormality of unknown pathology, or pleural adhesions have been suggested [3].

SLS may complicate SLE at any time over its course, early as a few months to 24 years after disease onset [4]. This usually present with progressive dyspnoea initially with activity and later at rest. Pleuritic

chest pains are frequent and accompanies dyspnoea, whereas dry cough and fevers are rare [5]. Elevation of the diaphragm is a universal radiographic feature [6].

Hence, characteristic features of SLS are unexplained dyspnea, small lung volumes, elevation of the diaphragm, and restrictive lung function tests [7]. There is no definitive treatment although glucocorticoids are considered the first line therapy alone or in combination with immunosuppressive agents [8].

Some studies have demonstrated the efficacy of theophylline with the intent to increase diaphragmatic strength and contractility [9]. Non-responding SLS (neither to glucocorticoids nor immunosuppressants), have shown remarkable improvement after treatment with Rituximab [10].

Figures

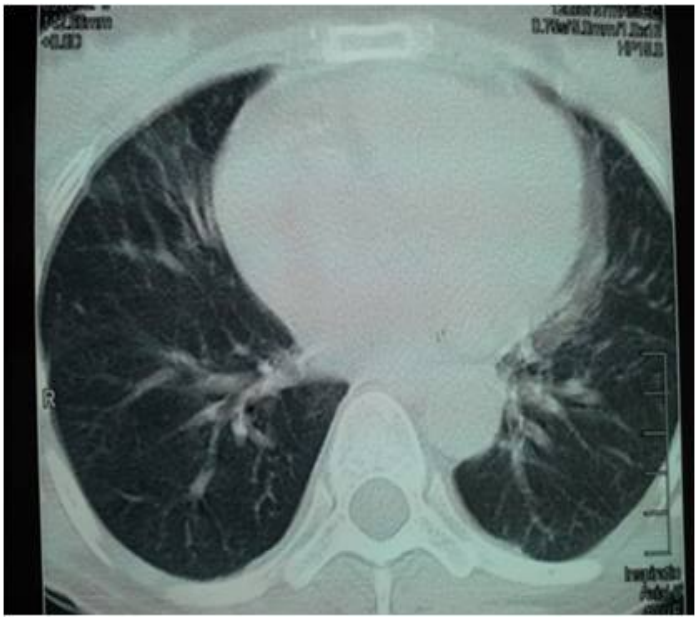

Figure 1: HRCT of the chest showing normal lung parenchyma

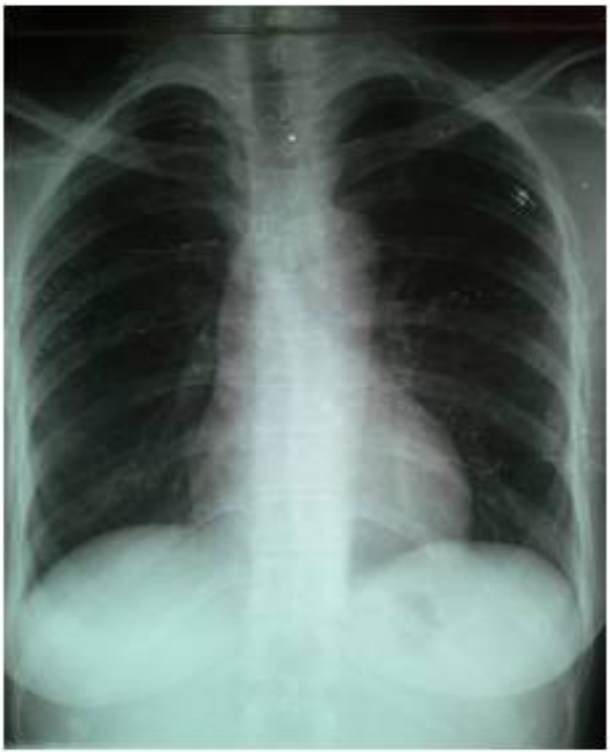

Figure 2: Chest X-ray at initial presentation, with normal lung volumes

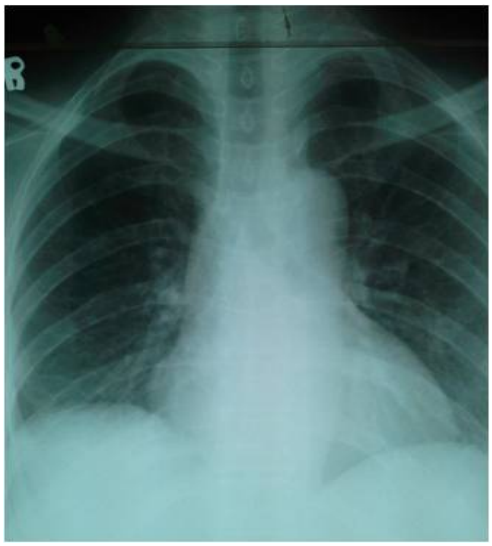

Figure 3: Chest X-ray showing reduction of lung volumes over one and half years' time with mild bilateral basal atelectasis

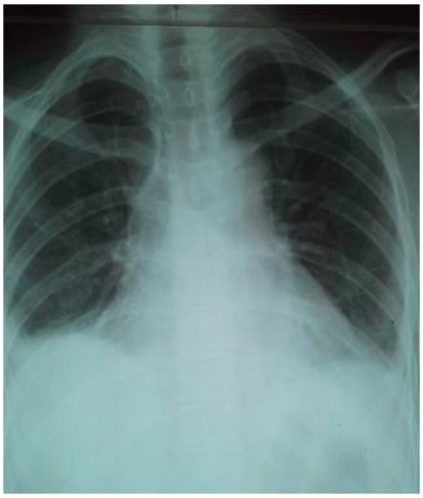

Figure 4: Chest X- ray showing reduction of lung volumes and mild effusion in the right lung-associated an episode of pulmonary emboli.

References

1. Hoffbrand BI, Beck ER. "Unexplained" dyspnoea and shrinking lungs in systemic lupus erythematosus. Br Med J. 1965;1:1273-1277.

2. Munoz ML, Gelber AC, Houston BA. Into Thin Air: Shrinking Lung Syndrome. Diagnostic dilemm. The American Journal of Medicine. August 2014Volume 127, Issue 8, Pages 711–713.

3. H.A Al-Raqum, S.S Uppal, M Al-Mutairy. Shrinking lung syndrome as a presenting manifestation of systemic lupuserythematosus in a female Kuwaiti. Clin Rheumatol 2006; 25:412-4.

4. Singh R, Huang W, Menon Y, Espinoza LR. Shrinking lungsyndrome in systemic lupus erythematosus and Sjogren'ssyndrome. J Clin Rheumatol 2002; 8: 340-5.

5. Warrington KJ, Moder KG, Brutinel WM. The Shrinking lungssyndrome in systemic lupus erythematous. Mayo Clin Proc 2000;75: 467-72.

6. Karim MY, Miranda LC, Tench CM, Gordon PA, D'cruz DP, Khamashta MA, et al. Presentation and prognosis of the Shrinking lung syndrome in systemic lupus erythematousus. Semin Arthritis Rheum 2002; 31: 289-98.

7. H.A Al-Raqum, S.S Uppal, M Al-Mutairy. Shrinking lung syndrome as a presenting manifestation of systemic lupusery thematosus in a female Kuwaiti. Clin Rheumatol 2006; 25:412-4.

8. Warrington KJ, Moder KG, Brutinel WM. The Shrinking lungs syndrome in systemic lupus erythematous. Mayo Clin Proc 2000;75: 467-72.

9. Van Veen S, Peeters AJ, Sterk PJ. The shrinking lung syndrome inSLE, treatment with theophylline. Clin Rheumatol 1993; 12: 4625.

10. Peñacoba Toribio P, Córica Albani ME, Mayos Pérez M, Rodriguez de la Serna A. Rituximab in the treatment of shrinking lung syndrome in systemic lupus erythematosus. Reumatol Clin2014; 10: 325-7.

Manuscript Information: Received: August 20, 2017; Accepted: December 08, 2017; Published: December 28, 2017

Authors Information: Geetha Wickrematilake, MBBS, MD, MRCP(UK)

Consultant Rheumatologist, Rehabilitation Hospital, Digana, Sri Lanka

Citation: Wickrematilake G. Systemic lupus erythematosus with shortness of breath. Open J Clin Med Case Rep. 2017; 1352.

Journal: Open Journal of Clinical and Medical Case Reports is an international, open access, peer reviewed Journal focusing exclusively on case reports covering all areas of clinical & medical sciences.

Visit the journal website at **www.jclinmedcasereports.com**
For reprints and other information, contact editorial office at **info@jclinmedcasereports.com**