Case Report

Shrinking lung syndrome in systemic lupus erythematosus-scleroderma overlap

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ABSTRACT

Shrinking lung syndrome (SLS) is a infrequently reported manifestation of systemic lupus erythematosus (SLE). Reported prevalence of SLS is about 0.5% in SLE patients. Pathogenesis is not fully understood and different therapeutic modalities have been employed with variable results, as only 77 cases of SLS have been documented in literature. SLS in SLE-Scleroderma overlap has not been reported yet. We report a patient of SLE - scleroderma overlap presenting with dyspnea, intermittent orthopnea and pleuritic chest pain. Evaluation revealed elevated hemidiaphragms and severe restrictive defect. She was eventually diagnosed as a case of SLS. This case report is a reminder to the medical fraternity that SLS although a rare complication must be thought of in the special subset of patients of SLE having respiratory symptoms.

KEY WORDS: Scleroderma, scleroderma shrinking lung syndrome, systemic lupus erythematosus

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INTRODUCTION

Pleuropulmonary manifestations occur in 60-80% of patients with systemic lupus erythematosus (SLE), frequently in the form of pleuritis with or without pleural effusion, pneumonia, interstitial fibrosis, acute lupus pneumonitis and pulmonary hypertension.[1] Shrinking lung syndrome (SLS) is an uncommon pulmonary manifestation of SLE, although the exact prevalence is not known, as only 77 cases of SLS have been documented in the literature.[2] SLS in SLE-scleroderma overlap has not been reported so far. This condition might be unrecognized and the estimates of the prevalence may be inaccurate. The pathogenesis of SLS is not yet clear and different therapeutic approaches have been employed with variable success. We describe here a patient with SLE-scleroderma overlap presenting with pulmonary complaints and eventually being diagnosed as SLS.

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CASE REPORT

A 28-year-old woman, a resident of Maharashtra, India, presented in January 2013 with 2 years history of Raynaud's phenomenon, polyarthralgia, recurrent oral ulcers and diffuse hair loss from scalp. She continued to have these symptoms off and on since 2011; however, since last 2 months she had developed progressive darkening and tightening of skin (left more than right), restricted mouth opening and dysphagia especially to solids. On clinical examination she was having pallor, induration and hyperpigmentation of limbs, sclerodactyly with fixed flexion deformities in fingers of left hand, ulcers in buccal mucosa, diffuse alopecia, restricted mouth opening (3 cm) [Figure 1] and chest expansion of 2 cm. Her systemic examination was essentially normal.

Laboratory investigations [Table 1] showed anemia. Liver and kidney function tests were within normal range except for 24 hours urinary protein of 400 mg/day. Immunological work up revealed antinuclear antibody (ANA) and anti-ds DNA positive, anti-centromere antibody: 1.56 μ/ml (<3.0), SCL-70: 2.53 U/ml (<3.00), anti-Sm antibody: 97.94 IU/L (>15 positive), anticardiolipin antibodies (ACLA) IgG positive and anti-neutrophil cytoplasmic antibodies (ANCA) negative. A renal biopsy performed on her showed ANCA-negative pauci immune necrotizing glomerulonephritis.

She was thus diagnosed as a case of SLE-scleroderma overlap and started on oral prednisolone 40 mg OD, mycophenolate mofetil (MMF) 500 mg BD, hydroxychloroquine (HCQ) 200 mg BD. She was stabilized on this treatment and discharged.

In March 2013, while on treatment, she developed dyspnea on exertion with inability to walk for even 100 m on even ground, associated with intermittent episodes of orthopnea. She also complained of right sided chest pain which was pricking type exacerbated on deep inspiration with no history of cough or hemoptysis. Clinically she had tachycardia (pulse 110/min), tachypnea (respiratory rate 28/min, rapid shallow breathing) with normal saturation. Jugular venous pressure was not raised and there was no pedal edema. Chest examination showed bilaterally reduced lung volumes and chest expansion. Breath sounds were normal with no adventitious sounds. Second heart sound was not accentuated.

Chest radiograph showed elevation of both hemidiaphragms (right more than left) and bilaterally reduced lung volumes. Lung fields were normal [Figure 2] with high-resolution computed tomography (HRCT) of the chest being normal [Figure 3]. Ultrasonography of abdomen with sniff test revealed no infradiaphragmatic pathology or diaphragmatic palsy. Her 2D echocardiography was normal. Pulmonary function tests showed severe restrictive defect [Table 2]. She could not hold her breath for DLCO maneuver which, thus, could not be done.

Arterial blood gas analysis showed pH 7.53, pO_2 74.9, HCO_3 19.8, pCO_2 29.3. Brain natriuretic peptide (BNP) was normal. Based on these findings she was diagnosed as SLS and metered dose inhalers (with spacer) Formetrol + Budesonide (200 μg) 02 puffs BD were started along with tab Deriphylline (Theophyllin) 150 mg BD and oral prednisolone was continued. She responded to the treatment and her orthopnea improved after 2 days of therapy. Breathlessness gradually improved over next 5 days and the patient was eventually discharged.

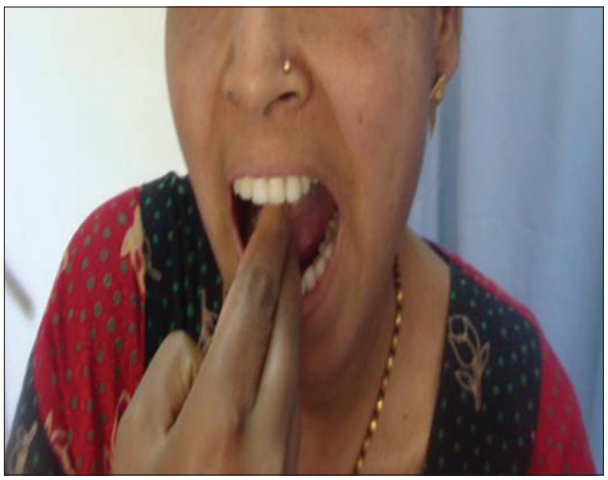

Figure 1: Reduced mouth opening (03 cm)

DISCUSSION

Pleuropulmonary involvement in SLE occurs in 60 to 80% of patients.[1-3]

SLS is a rare complication of SLE, with a reported prevalence of 0.5% of this overall population. First named in 1965 by Hoffbrand and Beck[4] SLS was used to describe a SLE patient who presented with dyspnea, radiological evidence of raised diaphragm, and a restrictive pattern of pulmonary function test. The precise pathogenetic mechanism underlying the SLS remains to be elucidated. Studies found no evidence of major parenchymal lung or pleural disease on thoracic CT scanning.[2] Pulmonary surfactant deficiency was initially thought to be the cause of alveolar microatelectasis and hyaline membrane formation. Later investigators found abnormalities in transdiaphragmatic pressures consistent with diaphragm dysfunction.[2,5,6]

Phrenic nerve involvement and myopathy were thought to be the cause of diaphragm dysfunction,[6,7] but this has not been established till date. The majority of neurophysiological studies yielded normal nerve conduction velocity that excluded the presence of a demyelinating neuropathy as the cause of diaphragmatic weakness. Pérez *et al*.[8] reported a case of SLS caused

Table 1: Investigations of the patient

Investigation	Value
Hb	9 gm/dl
TLC	7800/mm^3
Platelets	3.1 lakh/mm^3
Serum Bilirubin	0.8 mg/dl
AST	20 IU/dl
ALT	36 IU/dl
Creatinine	1 mg/dl
ESR	42 mm first hour
Urine RE	Normal
ANA	Positive
Anti-ds DNA	Positive
Anti centromere antibody	1.56 U/ml (normal<3.0)
SCL-70	2.53 U/ml (normal<3.00)
Anti Sm antibody	97.94 IU/L (>15 positive)
ACLA	IgG positive
ANCA	Negative
Anti-U1RNP	Negative

TLC: Total leucocyte count, AST: Aspartate transaminase, ALT: Alanine transaminase, ESR: Erythrocyte sedimentation rate, ANA: Antinuclear antibody, DNA: Deoxyribonucleic acid, ACLA: Anticardiolipin antibodies, ANCA: Anti-neutrophil cytoplasmic antibodies

Table 2: Pulmonary function tests

Values	Predicted	Actual	Percent predicted
TLC (L)	3.76	1.69	49.9
RV (L)	1.16	1.10	97.6
RV/TLC (%)	31.35	69.94	222.2
VC IN (L)	2.32	0.40	14.6
FEV 1 (L)	2.17	0.40	22.2
FEV 1/FVC (%)	63.67	97.37	116.4
PEF (L/s)	4.99	1.24	24.9

TLC: Total lung capacity, FEV: Forced expiratory volume, FVC: Forced vital capacity

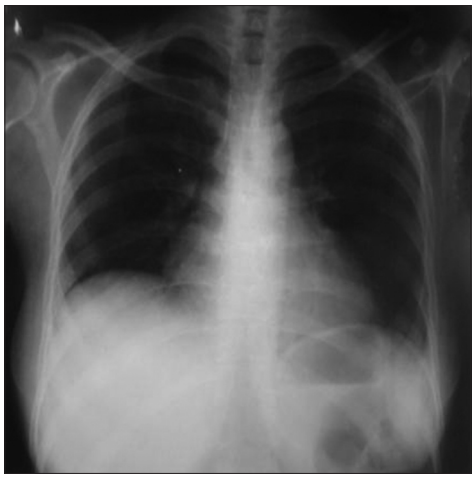

Figure 2: Chest radiograph showing elevation of both hemidiaphragms (right more than left) and bilaterally reduced lung volumes right hemidiaphragm

by lupus myopathy, who went into respiratory failure. The authors proposed that this may be due to infiltration of the chest wall muscles and the diaphragm by T lymphocytes (as they had demonstrated T cells in the deltoids sample). However, elevation of CPK, which is an indicator of myositis, has not been reported in patients with SLS.

Our patient presented with classical triad of SLS namely, dyspnea, raised diaphragm and a restrictive pulmonary defect. Parenchymal lung disease was unlikely with clinical examination and normal chest X-ray (other than small lung volumes and raised hemidiaphragms) and HRCT scan. Cardiovascular cause was ruled out by an essentially normal echocardioy and BNP. Reduced total lung capacity with preserved residual volume and normal FEV1/FVC and absence of air trapping on radiology points towards an extrapulmonary/ neuro-muscular cause of restriction as is expected in SLS. This probably is the first reported case of SLS in SLE-scleroderma overlap.

She was already on steroids; theophylline and MDI of β-agonist + inhaled steroids were added to her prescription to which she responded subjectively with resolution of orthopnea. These drugs are thought to improve contractility by acting on beta receptors on the diaphragm. Since the pathogenesis is not fully understood, different therapeutic approaches were reported to treat SLS. However, no RCTs have been carried out or consensus reached regarding optimal therapy.

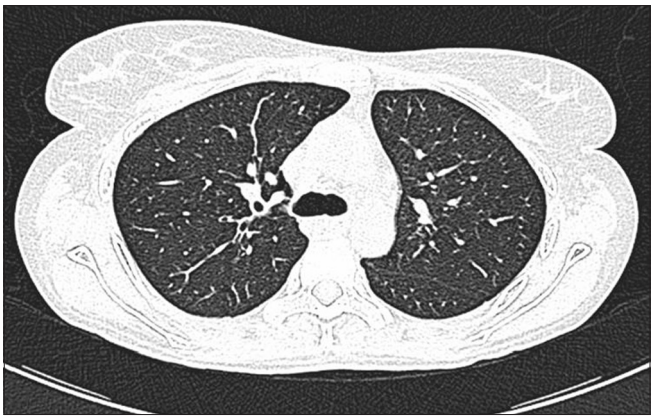

Figure 3: Normal HRCT scan

CONCLUSION

We have reported a case of SLS in a patient of SLE-Scleroderma overlap. Although this is a rare manifestation of the disease process but must be kept in hindsight in a patient of SLE who has dyspnea and orthopnea and is found to have normal chest examination and imaging with a restrictive pattern of PFT. The pathogenesis and optimal therapy for this condition has not yet been fully elucidated and continued research and possibly a RCT would be required to layout them out clearly.

REFERENCES

1. Fishback N, Koss MN. Pulmonary involvement in systemic lupus erythematosus. Curr Opin Pulm Med 1995;1:368-75.
2. Toya SP, Tzelepis GE. Association of the shrinking lung syndrome in systemic lupus erythematosus with pleurisy: A systematic review. Semin Arthritis Rheum 2009;39:30-7.
3. Karim MY, Miranda LC, Tench CM, Gordon PA, D'Cruz DP, Khamasthta MA, *et al*. Presentation and prognosis of the shrinking lung syndrome in systemic lupus erythematosus. Semin Artrhitis Rheum 2002;31:289-98.
4. Hoffbrand BI, Beck ER. "Unexplained" dyspnoea and shrinking lungs in systemic lupus erythematosus. Br Med J 1965;1:1273-7.
5. Warrington KJ, Moder KG, Brutinel WM. The shrinking lungs syndrome in systemic lupus erythematosus. Mayo Clin Proc 2000;75:467-72.
6. Wilcox PG, Stein HB, Clarke SD, Paré PD, Pardy RL. Phrenic nerve function in patients with diaphragmatic weakness and systemic lupus erythematosus. Chest 1988;93:352-8.
7. Gheita TA, Azkalany GS, El-Fishawy HS, Nour Eldin AM. Shrinking lung syndrome in systemic lupus erythematosus patients; clinical characteristics, disease activity and damage. Int J Rheum Dis 2011;14:361-8.
8. Pérez-de-Llano LA, Castro-Añón O, López MJ, Escalona E, Teijeira S, Sánchez-Andrade A. Shrinking lung syndrome caused by lupus myopathy. QJM 2011;104:259-62.

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