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.

KEY WORDS: Scleroderma, scleroderma shrinking lung syndrome, systemic lupus erythematosus
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 prickling 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, pO₂ 74.9, HCO₃ 19.8, pCO₂ 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.

**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.[5] 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/mm³       |
| Platelets           | 3.1 lakh/mm³   |
| 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) |
| ACA                 | IgG positive   |
| ANCA                | Negative       |
| Anti-U1RNP          | Negative       |

**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 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

---

**Figure 1:** Reduced mouth opening (03 cm)
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 echocardiog 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.

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, Khanashbta MA, et al. Presentation and prognosis of the shrinking lung syndrome in systemic lupus erythematosus. Semin Arthritis 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 KI, Modor KG, Brutinel WM. The shrinking lungs syndrome in systemic lupus erythematosus. Br Med J 1988;9:352-8.
6. Wilcox PG, Stein HB, Clarke SD, Paré PD, Pardy KL. Phrenic nerve function in patients with diaphragmatic weakness and systemic lupus erythematosus. Chest 1988;93:352-8.
7. Gheita TA, Azkalony 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.

How to cite this article: Guleria VS, Singh PK, Saxena P, Subramanian S. Shrinking lung syndrome in systemic lupus erythematosus-scleroderma overlap. Lung India 2014;31:407-9.

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