Shrinking Lung Syndrome in Systemic Lupus Erythematosus

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Shrinking lung syndrome (SLS) is an infrequently reported manifestation of systemic lupus erythematosus (SLE). Pathogenesis is not fully understood and different therapeutic modalities have been employed with variable results. We report a patient with SLS who responded subjectively and objectively to corticosteroids. Upon reduction of the dose she relapsed, but she responded again on increasing the dose. This case report shows that corticosteroids may improve dyspnea and lung function in shrinking lung syndrome.


Pleuropulmonary manifestations occur in more than 50% of patients with systemic lupus erythematosus (SLE), frequently in the form of pleuritis with or without pleural effusion, pneumonias, interstitial fibrosis, acute lupus pneumonitis and pulmonary hypertension.

Shrinking lung syndrome (SLS) is also not uncommon in SLE, occurring in 18 to 27% of patients. Yet, in a recent review found only 49 cases in the literature through a Medline search between 1965 and 1997. This suggests that this condition is unrecognized or that estimates of the prevalence are inaccurate. The pathogenesis of SLS it is not yet clear and different therapeutic approaches have been employed with variable success. Corticosteroids were reported to improve SLS by some authors, but not by others. We describe in this report a patient with SLS who responded to corticosteroid therapy but relapsed upon reduction of the dose. On increasing the dose, response was achieved again.

THE CASE

Thirty eight years-old Saudi woman was diagnosed to have SLE in early November 1998 on the basis of recurrent polyarthritis for 10 years, alopecia and strongly positive antinuclear antibodies and double stranded DNA. She was admitted for investigation of exertional dyspnea of 3 years duration, which became severe few weeks prior to admission, and was associated with orthopnea. It was not associated with cough and expectoration or chest pain. Chest examination revealed few basal crackles and reduced diaphragmatic excursion bilaterally. Examination of other systems was unremarkable, and in particular, there was no evidence of peripheral muscle weakness or cardiac abnormality.

Laboratory investigations showed normal full blood count, an elevated ESR (93 mm/hr), antinuclear antibody (1: 1280), double standard DNA (845), and a low serum complement (C3 0.59 [N = 0.7-1.3 gm/L], C4 0.11 [N 0.2 – 0.5 gm/L]). Urea, creatinine, serum electrolytes, creatinine kinase, liver function tests and bone profile were within normal range.

Chest radiograph (Fig. 1) showed reduced lung volume, high diaphragm and basal atelectasis. Computed tomography (CT) of the chest revealed peripheral band atelectasis at both lung bases with reduced lung volume and minimal pleural effusion. Fluoroscopy demonstrated diminished diaphragmatic movement bilaterally more on the right side. Initial pulmonary function tests showed severe restrictive defect (predicted values are shown in brackets): FVC 1.1 L (34%), FEV1 1.0 (38%), TLC 2.52 L (55%), diffusing capacity (DLCO) 9.5 ml/min/mmHg (58%) and a carbon monoxide coefficient (KCO) 4.8 ml/min/mmHg/L (118 %). Maximum inspiratory and expiratory

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pressures were 35 mmHg and 60 mmHg respectively. Arterial oxygen and carbon dioxide tensions were 83 and 36 mmHg respectively. Nerve conduction studies (done by stimulating the right and left phrenic nerves and obtaining recordings from the seventh and eighth intercostals spaces) showed normal results. Ventilation perfusion lungs scan and trans-thoracic echocardiogram were also normal.

Figure 1. The chest radiograph showing elevated hemidiaphragms, reduced and clear lung fields.

Diagnosis of shrinking lung syndrome was made and treated with prednisone 60 mg daily and salbutamol inhalers 200 µg every 6 hrs. Two weeks later, dyspnea and lung function improved significantly (Table 1).

On subsequent follow up in the outpatient clinic, she remained stable and prednisone dose was gradually tapered. In April 2000, whilst on a dose of 10 mg alternating with 2.5 mg per day, she noted recurrence of severe dyspnea and orthopnea. Simultaneously, her pulmonary function tests showed significant deterioration and laboratory investigations showed a drop in serum complement (C₃ and C₄). Chest radiograph showed no new changes. Prednisone was increased to 20 mg daily. Her dyspnea improved and lung function came up to the base line values (Table 1), and she remained stable up to her last visit in December 2001.

Table I. Lung function parameters of the patient in relation to treatment with prednisone.

<table>
<thead>
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<th>1998</th>
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<td></td>
<td>14 Nov</td>
<td>27 Dec</td>
<td>7 Feb</td>
<td>17 April</td>
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<tr>
<td>SOB</td>
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<td>mild</td>
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<td>ESR</td>
<td>93</td>
<td>20</td>
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<td>FVC (L)</td>
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<td>FEV₁ (L)</td>
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<td>1.56</td>
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<tr>
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<td>58</td>
<td>55</td>
<td>66</td>
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<tr>
<td>KCO (% of predicted)</td>
<td>118</td>
<td>83</td>
<td>107</td>
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<td>90</td>
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<td>Compliment 3</td>
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<td>0.84</td>
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<td>0.59</td>
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<td>Compliment 4</td>
<td>0.11</td>
<td>0.24</td>
<td>-</td>
<td>0.12</td>
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<td>Prednisone (mg/day)</td>
<td>0</td>
<td>45</td>
<td>25</td>
<td>10 / 2.5 alternating</td>
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DISCUSSION
Small lung fields and elevated diaphragms were noted in association with SLE since 1954 and this was thought to be the result of parenchymal lung involvement. In 1965 Hoffbrand and Beck used the term SLS to describe a SLE patient who presented with dyspnea, radiological evidence of small lung fields and raised diaphragm, and a restrictive pattern of pulmonary function test.

The pathogenesis of this condition is still not entirely known. Proposed explanations include basal micro-atelectasis, recurrent diaphragmatic pleurisy leading to adhesions and fibrosis, myositis leading to respiratory muscle dysfunction and abnormal chest wall mechanics. More recently, Hardy et al. reported a patient in whom bilateral phrenic nerve paralysis was found to be the underlying mechanism.

Our patient presented with classical triad of SLS namely, dyspnea, raised diaphragm and a restrictive pulmonary defect. Parenchymal lung disease is unlikely with normal alveolar-arterial gradient and carbon monoxide diffusion coefficient. CT scan of the lungs showed only minor atelectatic changes that are likely to be secondary to the raised diaphragm. In addition, the low maximum inspiratory pressure and sluggish diaphragmatic movements on fluoroscopy are both consistent with diaphragmatic dysfunction. Nerve conduction studies were normal indicating that phrenic nerve paralysis is an unlikely mechanism for the diaphragmatic abnormality. She responded subjectively and objectively to steroids on presentation and after a relapse. A part from this relapse she had a stable course over the subsequent three years on a modest dose of steroids.

Since the pathogenesis is not fully understood, different therapeutic approaches were reported to treat SLS. Up to date we are not aware of any controlled trial, or a consensus regarding therapy. Several case reports concluded that corticosteroids were useful in achieving symptomatic as well as pulmonary function improvement. However, other authors reported failures. Van Veen et al. reported a patient who responded to theophylline after 6 weeks of unsuccessful steroid therapy. In addition, inhaled beta agonists were reported to help, and were used in our patient. These drugs are thought to improve contractility by acting on beta receptors on the diaphragm. However, our patient relapsed when corticosteroids were tapered despite being on the same dose of beta agonist, suggesting that it was not sufficient to sustain remission.

CONCLUSION

The patient showed both a subjective and objective response on two occasions, indicating that corticosteroids may be effective in SLS. The difference in reports regarding pathogenesis and in response to treatment suggests that more complex mechanisms are responsible for SLS, which require further investigation.

REFERENCES