4. Couceiro A. Ética profesional y cuidados paliativos: en busca de la excelencia. In: Couceiro A, editor. Ética en cuidados paliativos. Madrid: Triacastela; 2004. p. 31-48.

Shrinking Lung Syndrome: A Rare Manifestation of Systemic Lupus Erythematosus

To the Editor: Systemic lupus erythematosus (SLE) is an autoimmune disease that may affect almost any organ or system. Pleuropulmonary signs are common and the most widely reported is pleuritis (with or without pleural effusion). A rare entity is the so-called shrinking lung syndrome or small lung. We report the case of a woman with SLE who was diagnosed with shrinking lung syndrome and who responded favorably to treatment with immunodepressant therapy.

The 38-year-old woman had been diagnosed with SLE a year earlier and was receiving treatment with hydroxychloroquine, indomethacin, and prednisone (5 mg daily). She was admitted to our ward complaining of dyspnea after minimal effort that had been progressing for 3 months, accompanied by pleuritic chest pain in the left side, but without fever, cough, or sputum. In the physical examination, the patient did not have fever and was eupneic at rest, with normal blood pressure and no tachycardia. She had a malar rash and monoarthritis of the index finger of the right hand. Cardiac and pulmonary auscultation and abdominal examination were all normal. The most relevant findings of the laboratory analyses were as follows: hemoglobin concentration, 12.5 g/dl; total white cell count, 2400 cells/µL; lymphocyte count, 900 cells/µL; erythrocyte sedimentation rate, 49 mm/h; C-reactive protein, 3.4 mg/L: C3, 108 mg/dL; C4, 13.2 of mg/dL; alanine aminotransferase, 53 U/L; aspartate aminotransferase, 44 U/L; and ferritin, 177.5 ng/mL. Tests for antinuclear, anti-DNA, anti-Sm, anti-Ro, and anti-La antibodies were positive, while the test for anticardiolipin antibodies was negative. Kidney function, ion, urine sediment, and muscle enzyme findings were normal. Arterial blood gas analysis breathing room air showed the following results: PaO_2, 78 mm Hg; $PaCO_2$, 32 mm Hg; bicarbonate, 23 mmol/L; and pH, 7.34. A chest radiograph revealed elevation of both hemidiaphragms and linear atelectasis in the right base. Computed tomography angiography was normal. Spirometry, carbon monoxide diffusing capacity, and determination of lung volumes revealed a severe restrictive pattern: forced vital capacity was 46% of the theoretical value (1.8 L), forced expiratory volume in 1 second, 53% (1.7 L), total lung capacity, 48% (2.6 L), and diffusing capacity of the lung for carbon monoxide, 56%, with a normal carbon monoxide transfer coefficient (94%).

After shrinking lung syndrome was diagnosed, treatment was initiated with daily intravenous bolus doses of 1 g of methylprednisolone for 3 days, followed by 0.5 mg/kg/d of prednisone for 4 weeks, tapered until treatment was withdrawn after 6 months. Azathioprine at doses

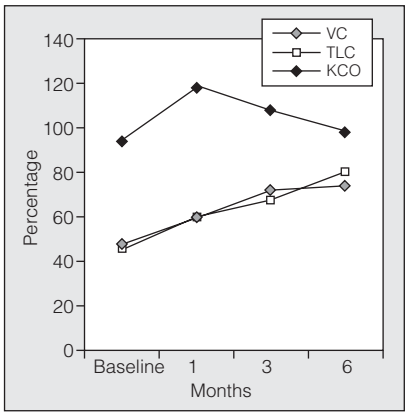

Figure. Changes in vital capacity (VC), total lung capacity (TLC), and carbon monoxide transfer coefficient (KCO) after initiation of treatment.

of 2 mg/kg/d was added to the treatment. Dyspnea and chest pain improved gradually during the following 4 weeks. Respiratory function tests were conducted at 1, 3, and 6 months of starting treatment, with gradual improvement of the respiratory function parameters (Figure). The chest radiograph became normal at 4 weeks.

Shrinking lung syndrome is a rare entity in patients with SLE and is distinguished by dyspnea, reduced lung capacity in lung function tests, and a raised diaphragm, with no abnormal radiographic findings in either lung.[1] Its etiology is debated, but in the light of lung function[2] and electrophysiology[3] findings, it seems reasonable to suppose that diaphragmatic dysfunction, whose origin is either myopathic or neuropathic due to phrenic nerve involvement, along with altered chest wall compliance[4] may jointly contribute to the development of shrinking lung syndrome to varying degrees. From a clinical perspective, it manifests as progressive dyspnea for weeks or months. Pleuritic chest pain is common, though it is not linked to any underlying pathogenetic mechanism, except for minimal pleural thickening in the computed tomography scan.[5] There is no standard treatment for this syndrome. The majority of published series describe the efficacy of different doses of corticosteroids (2060 mg daily).[5,6] Severe cases have been treated with regimens of corticosteroids in daily bolus doses (250-1000 mg) and immunosuppressants, such as cyclophosphamide or azathioprine, with varying results.[6] Other drugs, such as theophylline or β_2-agonists, have shown to be effective in isolated cases. In most of the cases published, ventilatory response to the treatment was incomplete or late (weeks or months).[5,6] The early, aggressive treatment of our patient might be the reason for the marked recovery in lung capacity and the normalization of radiographic alterations, circumstances rarely described in the medical literature. The long-term prognosis for shrinking lung syndrome is generally favorable, with improvement in dyspnea and chest pain, and functional improvement or stabilization of ventilation and lung diffusion capacity.[5] However, there may also be a worsening of respiratory function, requiring supplemental oxygen therapy and even endotracheal intubation and mechanical ventilation.

In conclusion, because shrinking lung syndrome is a rare entity whose prognosis is potentially limiting, the clinician should recognise and include it in the differential diagnosis of patients with SLE presenting with unexplained dyspnea and/or chest pain, in order to consider the onset or intensification of treatment with immunosuppressant drugs.

Rafael Vilaplana-García,
Antonio Javier Trujillo-Santos,
and Francisco Jesús Vera-Méndez
Servicio de Medicina Interna,
Hospital General Santa María del Rosell,
Cartagena, Murcia, Spain

1. Hoffbrand BI, Beck ER. Unexplained dyspnoea and shrinking lungs in systemic lupus erythematosus. Br Med J. 1965;1:1273-7.
2. Gibson CJ, Edmonds JP, Hughes GR. Diaphragm function and lung involvement in systemic lupus erythematosus. Am J Med. 1977;63:926-32.
3. Hardy K, Herry I, Attali V, Cadranel J, Similowski T. Bilateral phrenic paralysis in a patient with systemic lupus erythematosus. Chest. 2001;119:1274-7.
4. Laroche CM, Mulvey DA, Hawkins PN, Walport MJ, Strickland B, Moxham J, et al. Diaphragm strength in the shrinking lung syndrome of systemic lupus erythematosus. Q J Med. 1989;71:429-39.
5. Warrington KJ, Moder KG, Brutinel WM. The shrinking lungs syndrome in systemic lupus erythematosus. Mayo Clin Proc. 2000;75:467-72.
6. Oud KT, Bresser P, ten Berge RJ, Jonkers RE. The shrinking lung syndrome in systemic lupus erythematosus: improvement with corticosteroid therapy. Lupus. 2005;14:959-63.