Juan Marco Figueira Gonçalves,[a,*] Francisco Martínez Bugallo,[b] Ignacio García-Talavera,[a] Jesús Rodríguez González[a]

[a] *Servicio de Neumología y Cirugía Torácica, Hospital Universitario Nuestra Señora de la Candelaria, Santa Cruz de Tenerife, Spain*
[b] *Unidad de Genética, Servicio de Análisis Clínicos, Hospital Universitario Nuestra Señora de la Candelaria, Santa Cruz de Tenerife, Spain*

Corresponding author.
E-mail address: juanmarcofigueira@gmail.com
(J.M. Figueira Gonçalves).

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The Utility of Diaphragmatic Ultrasound in the Radiological Diagnosis of Systemic Lupus Erythymatosus Patients With Shrinking Lung Syndrome☆

Diagnóstico radiológico en el shrinking lung syndrome *en pacientes con lupus eritematoso sistémico. Utilidad de la ecografía diafragmática*

To the Editor,

We report the case of a 30-year-old woman, native of Bolivia, with no cardiovascular risk factors or toxic habits, diagnosed in December 2012 with systemic lupus erythematosus (SLE) and Sjögren's syndrome. She was receiving treatment with methotrexate, prednisone, and hydroxychloroquine.

She attended the systemic disease clinic in November 2015 due to dyspnea on moderate exertion, accompanied by orthopnea with no associated clinical evidence of infectious disease. Of note on examination were tachypnea and tachycardia with no tolerance to the decubitus position. She was hospitalized for further examinations and treatment.

Additional tests produced the following results:

- Clinical laboratory tests revealed anemia with leukopenia and mild thrombocytosis, and no other changes.
- Lung function tests showed a restrictive pattern and her maximum inspiratory and expiratory pressures were low, particularly maximum inspiratory pressure at 18.4% predicted value.
- Tests to detect heart disease were normal, including electrocardiogram, echocardiography, pro-BNP values, and enzymes for myocardial insult.

The following radiological tests were performed:

1. Chest radiograph showing elevation of both hemidiaphragms, with no other significant changes.
2. High-resolution computed tomography (HRCT), which showed laminar atelectasis in the right middle and lower lobe.
3. CT-angiogram revealed no evidence of acute or chronic pulmonary thromboembolism.
4. Initial ultrasonography of the chest and diaphragm showed limited diaphragmatic amplitude, both at rest and during deep breathing and voluntary sniff maneuvers, reduced inspiratory time and diaphragmatic cycle, and increased diaphragmatic contraction speed. Table 1 lists these parameters on admission and during the follow-up performed 3 months after discharge.

Based on these findings, shrinking lung syndrome in a patient with SLE and Sjögren's syndrome was diagnosed. Treatment began with theophylline, salbutamol, and high-dose prednisone. Clinical and ultrasound findings confirmed the patient's good progress, and she was discharged after 15 days of hospitalization.

Lung involvement in SLE is very common, and can occur in up to 60%–80% of cases,[1–3] often in the form of pleurisy with or without pleural effusion, pneumonia, interstitial fibrosis, acute lupus pneumonitis or pulmonary hypertension.[4]

A rarer, less common form of lung involvement in SLE is shrinking lung syndrome (SLS): less than 100 cases have been reported in the literature, and the prevalence among lupus patients is estimated to be less than 1%,[5] although an increasing number of authors are now claiming that this entity is underdiagnosed in mild cases.[6] The first authors to describe this syndrome were Hoffbrand and Beck in 1965,[7] in a study of 24 patients with lupus, 8 of whom developed unexplained dyspnea. They found that all patients had progressive reduction of lung volumes, and a restrictive ventilatory pattern on spirometry, associated with loss of ventilated lung volume on chest radiograph, leading the authors to propose the term "shrinking lung syndrome".

This syndrome is generally diagnosed 4 years after onset of SLE,[5] although cases have been published in which SLS was the first respiratory manifestation of the disease.[8–10]

The SLS triad is formed by elevated hemidiaphragms, dyspnea with normal lung parenchyma and restrictive pattern on spirometry.

Dyspnea with chest pain is the most frequent complaint among SLS patients, along with orthopnea and intolerance to the decubitus position.

The causes of the SLS are not entirely clear, although a number of hypotheses have been put forward; for example, the syndrome is caused by secondary microatelectasias due to a pulmonary surfactant deficiency, or it is caused by lupus myopathy due to T-cells infiltrating the diaphragm and the muscles of the chest wall.[11]

Diagnosis is derived from clinical suspicion, lung function tests showing a restrictive pattern, and radiological tests ruling out other diseases.

Treatment is not well established, but immunosuppressive drugs are the most widely used and prognosis is generally favorable.

Our case meets the criteria of SLS that have been described to date, since this was a patient diagnosed 3 years previously with SLE[5] who had Sjögren's syndrome with anti-Ro^{+} antibodies that are often associated with the presence of SLS. Our patient's clinical manifestations were typical, with dyspnea on exertion, chest pain, and orthopnea with intolerance to decubitus position, possibly related with weak respiratory muscles. To reach the presumptive diagnosis, other causes of dyspnea in lupus patients must be ruled out: the diagnosis of SLS is initially reached by exclusion of other entities.

In this respect, we would like to highlight the usefulness of ultrasonography of the diaphragm and the chest when SLS is suspected.[12–15] This is a relatively simple rapid and non-invasive examination that can be performed at the patient's bedside, and only minimal collaboration is needed to perform forced inspirations

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Table 1
Chest and Diaphragm Ultrasonography Values in our Patient Before and After Treatment.

M-Mode Ultrasonography		Normal	Pre-Treatment	Post-Treatment
Diaphragmatic excursion (cm)	Rest	1.6 ± 0.3	0.3 ± 0.1	0.6 ± 0.2
	Deep breathing	5.7 ± 1.0	1.4 ± 0.5	2.5 ± 0.6
	Voluntary sniff	2.6 ± 0.5	0.6 ± 0.2	1.1 ± 0.4
Speed of diaphragmatic contraction (cm/s)		1.3 ± 0.4	4.0 ± 0.5	2.3 ± 0.4
Inspiratory time (s)		1.6 ± 0.5	0.4 ± 0.2	1.1 ± 0.3
Diaphragmatic cycle length (s)		4.5 ± 1.1	1.6 ± 0.8	3.0 ± 1.0

when indicated. It provides real-time dynamic data on diaphragmatic function that cannot be obtained from other radiological tests, and as it involves no radiation, it is the procedure of choice in pregnant women and children. Because ultrasonography provides information on diaphragm mobility and excursion, and quantifies diaphragmatic contraction speed, duration of the breathing cycle and inspiratory time, we believe it can replace the fluoroscopic sniff test, the gold standard for evaluating diaphragm function. The objective parameters obtained with ultrasonography help establish the clinical diagnosis and assess the functional response of the diaphragm to treatment, and the study can also be used to explore the pulmonary parenchyma, yielding data suggestive of atelectasis or condensation, and the presence of serositis in the form of pleural or pericardial effusion, common in patients with lupus.

References

1. Gibson CJ, Edmonds JP, Hughes GR. Diaphragm function and lung involvement in systemic lupus erythematosus. Am J Med. 1977;63:926–32.
2. Orens JB, Martinez FJ, Lynch JP III. Pleuropulmonary manifestations of systemic lupus erythematosus. Rheum Dis Clin North Am. 1994;20:159–93.
3. Pena González A, Rodríguez Casal P, González Barcala FJ. Manifestaciones pulmonares del lupus eritematoso sistémico. Pneuma. 2010;6:109–16.
4. Keane MP, Lynch JP III. Pleuropulmonary manifestations of systemic lupus erythematosus. Thorax. 2000;55:159–66.
5. 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.
6. Allen D, Fischer A, Bshouty Z, Robinson DB, Peschken CA, Hitchon C, et al. Evaluating systemic lupus erythematosus patients for lung involvement. Lupus. 2012;21:1316–25.
7. Hoffbrand BI, Beck ER. Unexplained dyspnoea and shrinking lungs in systemic lupus erythematosus. Br Med J. 1965;1:1273–7.
8. Branger S, Schleinitz N, Gayet S, Veit V, Kaplanski G, Badier M, et al. Shrinking lung syndrome and systemic auto-immune disease. Rev Med Interne. 2004;25:83–90 [Article in French].
9. 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 erythematosus. Semin Arthritis Rheum. 2002;31:289–98.
10. Pillai S, Mehta J, Levin T, Muzumdar H, Nandalike K. Shrinking lung syndrome presenting as an initial pulmonary manifestation of SLE. Lupus. 2014;23:1201–3.
11. Martens J, Demedts M, Vanmeenen MT, Dequeker J. Respiratory muscle dysfunction in systemic lupus erythematosus. Chest. 1983;84:170–5.
12. Orozco-Levi M, Gayete A, Rodríguez C, Ramírez-Sarmiento A, Méndez R, Tous F, et al. Non-invasive functional evaluation of the reserve in fatigue and the diaphragm structure using transthoracic echography in B and M modes. Arch Bronconeumol. 2010;46:571–9.
13. Gallego Gómez MP, García Benedito P, Pereira Boo D, Sánchez Pérez M. La ecografía torácica en la enfermedad pleuro-pulmonar. Radiologia. 2014;56:52–60.
14. Yang PC, Luh KT, Chang DB, Yu CJ, Kuo SH, Wu HD. Ultrasonographic evaluation of pulmonary consolidation. Am Rev Respir Dis. 1992;146:757–62.
15. Lichtenstein D, Meziere G, Seitz J. The dynamic air bronchogram: a lung ultrasound sign of alveolar consolidation ruling out atelectasis. Chest. 2009;135:1421–5.

Laura Díaz Rubia,[a,*] José Luis Callejas Rubio,[b] José Luis Martín-Rodríguez[a]

[a] *Servicio de Radiodiagnóstico, Complejo Hospitalario Universitario de Granada, Granada, Spain*
[b] *Servicio de Medicina Interna, Complejo Hospitalario Universitario de Granada, Granada, Spain*

Corresponding author.
E-mail address: laurix_dr@yahoo.es (L. Díaz Rubia).

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