

Shrinking Lung Syndrome: A Case Report

Síndrome del pulmón encogido: a propósito de un caso

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ABSTRACT

Background: Shrinking lung syndrome (SLS) is a rare respiratory manifestation of systemic lupus erythematosus (SLE), characterized by dyspnea, diaphragmatic elevation, and a restrictive pattern on lung function testing, with no alternative pleuropulmonary disease accounting for these findings.

Objectives: To describe the case of a patient with recently diagnosed SLE who developed SLS, and to review the main pathophysiological, diagnostic, and therapeutic aspects.

Case report: We present the case of a 38-year-old woman with a history of smoking and hypertension who was recently diagnosed with SLE. She was evaluated with chest computed tomography, lung function testing (spirometry and lung volumes), and diaphragmatic ultrasound.

Results: The patient presented with a 5-month history of dyspnea, intermittent fever, polyarthralgia, and asthenia. Chest computed tomography revealed bilateral subsegmental atelectasis and elevation of the right hemidiaphragm. Lung function testing showed a severe restrictive pattern with markedly reduced diffusing capacity for carbon monoxide (DLCO), and diaphragmatic ultrasound confirmed bilateral diaphragmatic paralysis. Following a diagnosis of SLS, treatment with oral corticosteroids and immunosuppression with cyclophosphamide was initiated, with a favorable clinical response.

Conclusions: SLS should be suspected in patients with SLE who present with dyspnea and an unexplained restrictive pattern. Its diagnosis is one of exclusion and requires a high index of suspicion. Diaphragmatic ultrasound is a useful tool for its assessment, and treatment with corticosteroids and immunosuppression is effective in most cases.

Key words: shrinking lung syndrome, systemic lupus erythematosus, diaphragmatic dysfunction, diaphragmatic ultrasound, restrictive pulmonary disease

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RESUMEN

Introducción: El síndrome del pulmón encogido (SPE) es una manifestación respiratoria infrecuente del lupus eritematoso sistémico (LES), caracterizada por disnea, elevación diafragmática y un patrón restrictivo en las pruebas de función pulmonar, en ausencia de otra enfermedad pleuropulmonar que lo explique.

Objetivos: Describir el caso de una paciente con diagnóstico reciente de LES que desarrolló SPE, y revisar sus principales aspectos fisiopatológicos, diagnósticos y terapéuticos.

Presentación del caso: Se presenta el caso de una paciente de 38 años, con antecedentes de tabaquismo e hipertensión arterial, y diagnóstico reciente de LES, evaluada mediante tomografía de tórax, pruebas de función pulmonar (espirometría y volúmenes pulmonares) y ecografía diafragmática.

Resultados: La paciente presentó un cuadro de cinco meses de evolución con disnea, fiebre intermitente, poliartralgias y astenia. La tomografía de tórax evidenció atelectasias subsegmentarias bilaterales y elevación diafragmática derecha. Las pruebas de función pulmonar mostraron un patrón restrictivo grave con DLCO significativamente disminuida, y la ecografía diafragmática confirmó parálisis diafragmática bilateral. Con diagnóstico de SPE, se inició tratamiento con corticoides orales e inmunosupresión con ciclofosfamida, con buena respuesta clínica.

Conclusiones: El SPE debe sospecharse ante un cuadro de disnea con patrón restrictivo sin causa aparente en pacientes con LES. Su diagnóstico es de exclusión y requiere un alto índice de sospecha. La ecografía diafragmática es una herramienta útil para su evaluación, y el tratamiento con corticoides e inmunosupresión resulta eficaz en la mayoría de los casos.

Palabras clave: síndrome del pulmón encogido, lupus eritematoso sistémico, disfunción diafragmática, ecografía diafragmática, enfermedad pulmonar restrictiva

INTRODUCTION

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease that can involve multiple components of the respiratory system, including the pulmonary parenchyma, pleura, pulmonary vasculature, airways, and respiratory muscles. Shrinking lung syndrome (SLS) is a rare manifestation primarily described in patients with SLE. However, it has also been reported in association with systemic sclerosis, Sjögren syndrome, and rheumatoid arthritis.¹⁻³

CASE REPORT

A 38-year-old patient with a history of tobacco use, systemic hypertension, and recently diagnosed with SLE presented with a 5-month history of dyspnea, intermittent fever, polyarthralgia, as-

thenia, and weakness. On admission, the patient's oxygen saturation was 90%, which corrected with a nasal cannula. A chest CT scan revealed bilateral subsegmental atelectasis associated with scattered cystic lesions and right diaphragmatic elevation. (Figures 1 and 2) Pulmonary function testing (spirometry and lung volumes) confirmed severe restriction and a significant decrease in DLCO, demonstrating severe gas transfer defect. The specific PFT values were as follows: FEV1/FVC 82.5 (LLN [lower limit of normal] 71.49), FEV1 1.21 (LLN 2.51), FVC 1.47 (LLN 3.06), TLC 2.94 (LLN 4.42), RV 1.43 (LLN 0.73), DLCO 9.89 (LLN 17.52), KCO 4.38 (LLN 3.49), VA 2.26 (LLN 4.46), MIP 69.52 (LLN 28.7), and MEP 47.14 (LLN 35.49). Diaphragmatic ultrasound subsequently demonstrated bilateral diaphragmatic paralysis. Based on the clinical presentation and imaging findings, a diagnosis of SLS was established. Oral corticosteroid therapy and immunosuppression with cyclophosphamide were initiated, resulting in a favorable response.

Figure 1. Chest computed tomography (coronal reconstruction) showing elevation of the right hemidiaphragm and the consequent loss of volume in the ipsilateral hemithorax.



Figure 2. Chest computed tomography (axial view, lung window) showing elevation of the right hemidiaphragm, bilateral subsegmental atelectasis, and small basal cystic lesions.



DISCUSSION

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease that can affect multiple components of the respiratory system. The most common pulmonary manifestations include pleuritis, lupus interstitial pneumonia, diffuse alveolar hemorrhage, and pulmonary hypertension, whereas shrinking lung syndrome (SLS) is an uncommon manifestation.¹⁻³

SLS was first described by Hoffbrand and Beck in 1965 and is characterized by dyspnea, pleuritic chest pain, elevation of the hemidiaphragm, and reduced lung volumes.⁴ Reported prevalence ranges from 0.5% to 1.5%, although estimates vary across published series. Borrell et al reported

a prevalence of 1.1% among patients with SLE.⁵ Similar to SLE, SLS predominantly affects women, with a female-to-male ratio of approximately 9:1.⁵

Although the precise pathophysiology remains uncertain and controversial, several hypotheses have been proposed. These include diaphragmatic dysfunction secondary to phrenic neuropathy, as well as paralysis resulting from vasculitis or immune complex deposition. However, most nerve conduction studies and electromyography findings are normal, suggesting that the impairment is primarily functional rather than a permanent electrical disruption.

Regarding diaphragmatic myopathies, the ultrasound has revealed reduced diaphragmatic thickness and strength, which correlates with a decrease in maximal inspiratory pressure (MIP). Some evidence suggests that lupus can cause primary muscle fibrosis or direct inflammatory myopathy of the diaphragm.⁶

Pleural involvement, including chronic pleuritis, mechanical restriction and pleural adhesions, can result in fibrosis that physically limits diaphragmatic excursion. Furthermore, pain induces a splinting effect, wherein protective muscle rigidity leads to disuse atrophy, thereby limiting lung expansion.^{6,7}

Regarding functional parenchymal abnormalities, microatelectasis and surfactant deficiency represent a traditional hypothesis. Inflammation is thought to impair surfactant production, thereby increasing surface tension and inducing diffuse microatelectasis that reduces lung parenchymal volume.^{6,7}

Mechanical abnormalities include reduced lung compliance and chest wall dysfunction. The most widely accepted hypothesis is that shrinking lung syndrome (SLS) results from chronic pleuritis with reflex inhibition of diaphragmatic motion, leading to hypoventilation, progressive loss of lung volume, and restrictive defect.^{8,9}

Within this framework, neural reflex inhibition is the most accepted mechanism. Pleural inflammation (pleuritis) activates sensory receptors that, via intercostal–phrenic reflex pathways, transmit inhibitory signals to the phrenic nerve, limiting diaphragmatic excursion to avoid pleuritic pain. This chronic hypoinflation progressively reduces lung compliance over time.

SLS reportedly originates from pleural inflam-

mation secondary to the underlying rheumatic disease. Activation of local neural reflexes, together with voluntary respiratory limitation due to pain, results in chronically reduced lung inflation.⁷ This process gradually impairs lung compliance through parenchymal changes that remain incompletely defined.

As the lung becomes less compliant, expansion becomes increasingly restricted, generating a progressive downward spiral of volume loss.^{7,10}

Clinically, the syndrome is characterized by dyspnea, which may or may not be accompanied by pleuritic chest pain. Less common manifestations include cough, fever, and orthopnea.^{4,5}

Chest radiography and high-resolution computed tomography (HRCT) can demonstrate elevation of the hemidiaphragm, with or without associated segmental or subsegmental atelectasis and pleural thickening.^{5,8,9} It should be noted that involvement may be bilateral in up to 70% of patients, according to Borrell et al.⁵ The HRCT is also useful to discard other structural processes of the lung parenchyma, the pleura, and pulmonary vasculature that may be present in patients with SLE, such as pleuritis and acute interstitial pneumonia.¹⁻³ Diaphragmatic ultrasound has emerged as an important diagnostic tool capable of identifying abnormalities in the diaphragmatic function. Both diaphragmatic excursion and thickening fraction have been shown to be useful ultrasound parameters for the assessment of diaphragmatic dysfunction or paralysis. However, as this technique is operator-dependent, findings should be interpreted in conjunction with the clinical and functional context.^{11,12}

Spirometry and lung volume measurements typically reveal a restrictive ventilatory pattern, characterized by reduced forced vital capacity (FVC) and total lung capacity (TLC), with a preserved FEV1/FVC ratio. In addition, diffusion capacity for carbon monoxide (DLCO) is frequently reduced; however, a normal DLCO does not exclude the diagnosis. Maximal inspiratory and expiratory pressures (MIP and MEP) may also be decreased and have shown to be useful in confirming respiratory muscle weakness.^{5, 8, 9, 13, 14}

SLS has been reported as the initial clinical

manifestation of SLE in approximately 10% of the cases, highlighting the importance of requesting an immunologic workup, particularly in patients with clinical suspicion and no prior diagnosis of autoimmune disease.

SLS is a diagnosis of exclusion, with high clinical suspicion in patients presenting with dyspnea, with or without pleuritic chest pain, associated with hemidiaphragm elevation and a restrictive pattern on lung function testing, in the absence of other pleuropulmonary condition.^{5, 8, 9, 11, 15}

Systemic corticosteroids are the treatment of choice for SLS. Although no standardized dosing regimen has been established, most clinical series report the use of moderate to high doses. Despite limited evidence, prednisone at 1 mg/kg/day appears to be a reasonable approach.¹⁵ Treatment duration is variable and should be individualized according to the patient's clinical response. Corticosteroid therapy may be combined with other immunosuppressive agents such as cyclophosphamide, mycophenolate, or azathioprine—particularly in cases involving severe respiratory impairment or a partial response to steroids, although controlled trials supporting this approach are currently unavailable.¹³ In refractory cases, benefits have been reported with rituximab, though data are primarily limited to case reports.^{16,17}

CONCLUSION

SLS is an uncommon manifestation of SLE that should be suspected in patients presenting with unexplained dyspnea and a restrictive ventilatory pattern. Early identification is crucial to prevent progressive respiratory impairment. Diagnosis is established by exclusion and requires a high index of suspicion supported by clinical, imaging (Figure 1), and pulmonary function findings. Diaphragmatic ultrasound represents a valuable tool for evaluating diaphragmatic dysfunction. While corticosteroid and immunosuppressive therapies have demonstrated efficacy in most cases, further studies are required to more precisely define optimal therapeutic strategies.

Conflict of interest

Authors have no conflicts of interest to declare. REFERENCES

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