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The Shrinking Lung Syndrome or Retracted Lung Syndrome During Systemic Lupus Erythematosus: About Two Cases

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Abstract **Case Report**

The shrinking lung syndrome represents an uncommon complication associated with systemic autoimmune diseases, primarily systemic lupus ery the matosus, but also Sjögren's syndrome and polymyositis. It is a condition that should be considered in any patient with an autoimmune disease presenting unexplained dyspnea. This syndrome is characterized by reduced lung volumes, elevation of the diaphragm, and restrictive physiology without significant parenchymal involvement. The article emphasizes the crucial role of thoracic CT in the diagnosis of the shrinking lung syndrome. Thoracic CT plays an essential role in identifying characteristic radiological signs such as pulmonary atelectasis and elevation of the diaphragmatic domes. These specific radiological features significantly contribute to confirming the diagnosis by excluding other potential causes of dyspnea, notably pulmonary embolism.

Keywords: shrinking lung syndrome, polymyositis, restrictive physiology, thoracic CT.

INTRODUCTION

Respiratory manifestations occur in 60 to 80% of systemic lupus erythematosus (SLE) patients. Shrinking Lung Syndrome (SLS) is a rare diaphragmatic involvement that affects 0.5 to 1% of lupus patients, with only 156 cases reported in the literature. It was first described in 1965 in 8 lupus patients but has sporadically been reported in other connective tissue diseases such as rheumatoid arthritis, scleroderma, and Sjögren's syndrome. SLS is characterized by unexplained dyspnea, elevation of diaphragmatic domes, and a restrictive ventilatory syndrome. We describe here two new cases of SLS in the context of lupus and provide a literature review, focusing particularly on the radiological characteristics of this rare association.

OBSERVATION

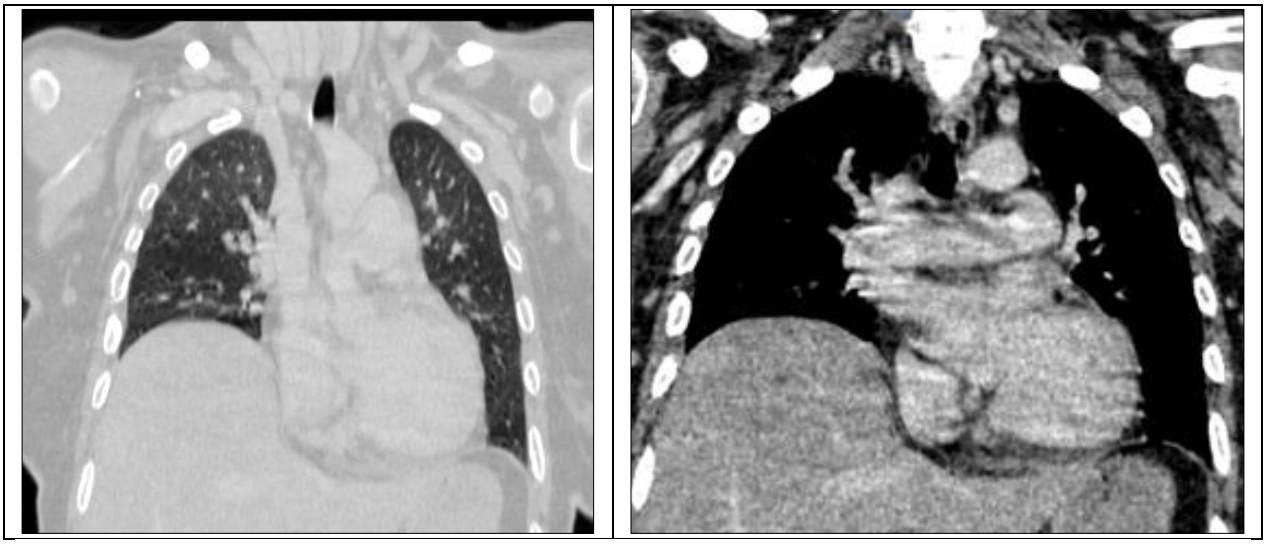
Case 1:

This concerns a 38-year-old patient diagnosed with systemic lupus erythematosus and antiphospholipid syndrome for the past 4 years, who was lost to follow-up for the last 2 years. The patient presents with stage IV dyspnea accompanied by a decline in general health. This symptomatology is associated with thoracic pains characterized by oppression. Upon examination, the patient exhibits a respiratory rate of 27 cycles/min with a heart rate of 100 bpm. A COVID-19 PCR test was performed due to the circumstances of the SARS-CoV-2 pandemic, yielding a negative result. Biologically, the patient shows respiratory alkalosis on blood gas analysis, an inflammatory biological syndrome (CRP at 60), and a D-dimer level of 4220. Thoracic imaging reveals a healthy pulmonary parenchyma and a reduction in the right lung hemi-field, as evidenced by the elevation of the diaphragmatic dome and narrowing of homolateral intercostal spaces, confirmed during both inspiration and forced expiration (Figure 1 and 2). Thoracic CT angiography does not identify any pulmonary embolism. Respiratory functional exploration demonstrates a restrictive ventilatory disorder. The diagnosis of shrinking lung syndrome is established.

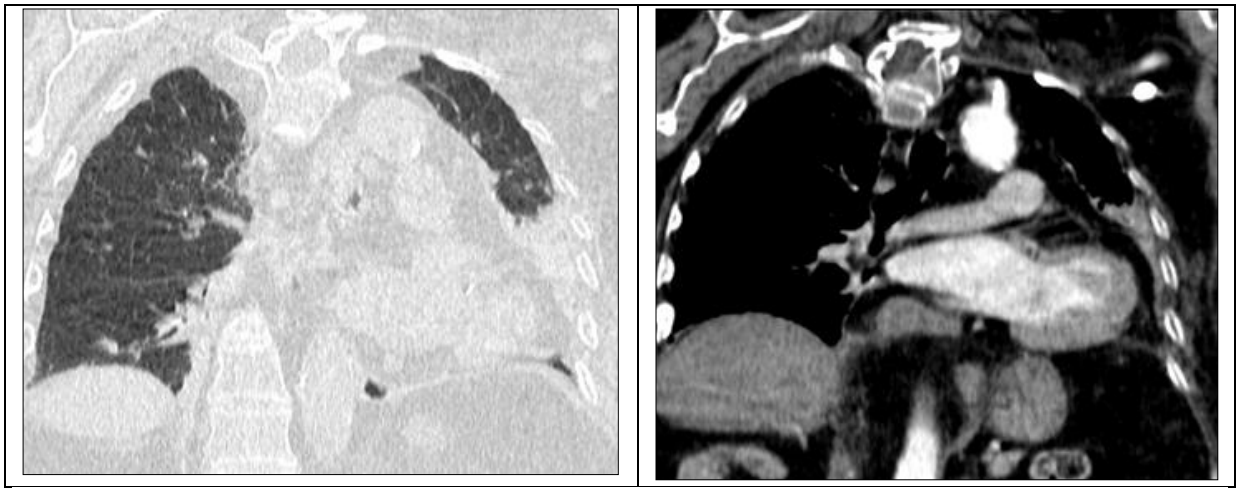
Case 2:

This concerns a 68-year-old patient, diagnosed with systemic lupus erythematosus since 2004 and receiving ongoing treatment. She is also being treated for deep vein thrombosis in the lower limbs with rivaroxaban. The patient was admitted with respiratory distress characterized by dyspnea initially upon exertion, which progressed to occurring at rest over the past 6 months. This symptomatology was associated with thoracic pains of an oppressive nature. Upon examination, she presented with a respiratory rate of 30 cycles/min and a heart rate of 105 bpm. Thoracic imaging (Figure 3 and 4) revealed focal pulmonary

atelectasis in the lingular, posterior, and left basolateral regions, displaying bronchogram and vasculogram features leading to a reduction in the left lung hemi-field with the attraction of mediastinal elements. Additionally, there was an elevation of the left diaphragmatic dome and narrowing of homolateral intercostal spaces. Thoracic CT angiography ruled out pulmonary embolism. Echocardiography did not show pericardial effusion, pulmonary arterial hypertension, or heart failure explaining her dyspnea. Spirometry indicated a restrictive ventilatory disorder. Capillaroscopy revealed right capillary dilation. The diagnosis of lupus with pulmonary involvement, specifically shrinking lung syndrome, was established. The patient was initiated on corticosteroid therapy at a dose of 0.5 mg/kg/day with a gradual taper, a calcium channel blocker, and hydroxychloroquine. The patient's condition improved, marked by a resolution of dyspnea and joint symptoms.



Thoracic CT scan: Figures 1 (parenchymal window in coronal section) and 2 (mediastinal window in coronal section): Reduction of the right lung field, as evidenced by the elevated diaphragm and narrowed ipsilateral intercostal spaces



Thoracic CT scan: Figure 1 (parenchymal window in coronal section) and Figure 2 (mediastinal window in coronal section): Focal areas of retractile pulmonary atelectasis associated with an elevation of the left diaphragmatic dome

DISCUSSION

The Shrinking Lung Syndrome (SLS) is still rarely described in medical literature, with only 100 reported cases so far, resulting in an estimated prevalence of <1% [5]. Some patients with systemic lupus erythematosus (SLE) may experience dyspnea without pleuroparenchymal or vascular involvement. It is important to consider a rare and less-known syndrome in such cases: the Shrinking Lung Syndrome, which corresponds to an involvement of respiratory muscles. SLS was initially described in the context of lupus and has later been reported in other autoimmune conditions such as Sjögren's syndrome, rheumatoid arthritis, or mixed connective tissue diseases [104].

It occurs within a period of 4 months to 24 years after the diagnosis of lupus but can also be contemporaneous. The prevalence of SLS is not well-known and challenging to estimate, but a study measuring transdiaphragmatic pressures found diaphragmatic dysfunction in 56% of systematically explored lupus patients [105].

Our patient had a long history of systemic lupus erythematosus (SLE) with multiple flare-ups presenting with shortness of breath and chest pain. These symptoms could be associated with common etiologies such as pulmonary embolism, pericarditis, or parenchymal lung disease. However, they could also have been the initial manifestations of her Shrinking Lung Syndrome (SLS), which is a rare condition [6, 7]. A case series by Ciaffi *et al*., highlights the challenges associated with diagnosing SLS due to its rarity and its nature as a diagnosis of exclusion [6]. Therefore, increased awareness and suspicion are crucial for early diagnosis and treatment. Early diagnosis and treatment can play a significant role in preventing disease progression and improving morbidity and mortality. The chest X-ray taken at the time of diagnosis showed an elevated right hemidiaphragm without clear evidence of parenchymal lung disease. The chest computed tomography (CT) scan also revealed normal lung parenchyma with no pleural effusion or signs of interstitial lung disease, except for band-like atelectasis at the lung bases, likely consequences of hypoventilation and diaphragmatic elevation.

Respiratory function tests reveal a restrictive syndrome with a decrease in mobilizable lung volumes. Carbon monoxide transfer is not impaired. Blood tests show no particular abnormalities, except for the reported association with the presence of anti-SSA antibodies [107]. The diagnosis is challenging because it is necessary to exclude other pleuroparenchymal and vascular involvements related to lupus. The absence of systematic review and research due to the rarity of reported cases continues to be a challenge for understanding the pathophysiology and developing evidence-based management for this condition. Several mechanisms have been proposed as possible causes, including respiratory myopathy, phrenic neuropathy, surfactant deficiency, and pleural adhesions [1-7]. Omdal *et al*., presented a case of bilateral elevation of hemidiaphragms and bibasilar atelectasis in a patient who initially experienced respiratory arrest. The patient was later diagnosed with systemic lupus erythematosus (SLE) and demonstrated phrenic neuropathy confirmed by electromyography and nerve conduction studies. They also reported diaphragmatic paralysis and myopathy in SLE patients. Similarly, in our patient, the elevation of her left hemidiaphragm could be caused by phrenic neuropathy and diaphragmatic myopathy due to poorly controlled SLE over an extended period.

Shrinking Lung Syndrome has a favorable prognosis. Most patients experience some improvement with appropriate immunosuppressive therapy. Clinical improvement is common, with the majority reporting symptomatic relief. There is stabilization or improvement in lung function testing in most cases [3-7]. Radiological improvement is less frequent, with 57% of cases in one study demonstrating improvement [15]. Complete recovery from clinical, functional, and radiographic abnormalities is rare There is no validated therapeutic strategy for Shrinking Lung Syndrome. Corticosteroids at a daily dosage of 30 to 60 mg prednisone equivalents are generally effective, leading to clinical improvement and improved lung volumes [115]. ß2-agonists and theophylline have been used anecdotally without rigorous evaluation. Immunosuppressants sparing corticosteroids such as azathioprine, methotrexate, or cyclophosphamide have been suggested by analogy to the treatment of myositis, but their efficacy has not been thoroughly assessed [116]. The same applies to respiratory physiotherapy and non-invasive ventilation.

CONCLUSION

The mode of presentation for lupus is rarely pulmonary. Lupus Shrinking Lung Syndrome is an extremely rare syndrome that primarily affects women and usually occurs during the course of the disease, being exceptionally revealing. Confirming it requires an extensive series of investigations to eliminate any causes of dyspnea.

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