

Shrinking Lung Detected on Computed Tomography: Pictorial Essay of the Main Findings of the Image

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Abstract

Shrinking lung refers to a rare complication of systemic lupus erythematosus and is characterized by unexplained dyspnea, a restrictive pattern in lung function tests, and elevation of the diaphragmatic hemicupules. It is postulated to have a predilection for female involvement and occurs mainly during late stages of the disease. Chest X-rays usually show small, diaphragmatic lungs. Occasional basal atelectasis may be present. Chest tomography usually shows reduced lung volumes with diaphragmatic elevation, occasional basal atelectasis, without severe pulmonary or pleuropulmonary disease. Shrinking lung can cause significant morbidity and occasional mortality. There is no definitive therapy, while corticosteroids may decrease symptoms and improve lung function in some patients. The objective of this study was to describe the main imaging findings in Shrinking Lung, an important pulmonary alteration in lupus patients. We highlight the characteristics observed on radiography and computed tomography, with an emphasis on computed tomography. It is important that every radiologist is prepared to recognize these findings and understand the possible clinical repercussions.

Keywords

Chest Tomography, Pulmonary Diseases, Systemic Lupus Erythematosus, Shrinking Lung

1. Introduction

The diagnostic interpretation of the findings of Systemic Lupus Erythematosus requires that the examiner be familiar with the imaging spectra of this pathology. Shrinking lung is one of those findings and of importance for the course of the disease, because its early recognition may bring additional therapeutic substrates to the patient. Computed tomography of the chest is the reference method for the identification of these anomalies and their repercussions.

Shrinking lung syndrome (SLS) was first described as a complication of systemic lupus erythematosus (SLE) by Hoffbrand and Beck [1]. The term identifies a constellation of respiratory manifestations including dyspnea, reduced lung volumes and/or elevated hemidiaphragms on chest radiography (CXR), and a restrictive ventilatory defect assessed by pulmonary function tests (PFTs). Serial CXRs demonstrate steadily declining lung volumes as SLS patients become more dyspneic, rendering the impression that the lungs are vanishing. SLS is rare, with an estimated prevalence of <1% among patients with SLE, though several recent reports have suggested a higher prevalence [2] [3] [4]. The syndrome has been reported in other rheumatologic disorders including scleroderma, Sjögren's syndrome, rheumatoid arthritis, and undifferentiated connective tissue disorder [5] [6] [7] [8]. The pathogenesis of SLS remains unknown. Chest imaging shows no evidence of interstitial lung disease or vascular pathology, although chest pain and small pleural effusions are common [9]. While patients may become markedly dyspneic, mortality is rare.

2. Objective of the Study

The objective of this study was to describe the main imaging findings in Shrinking Lung, an important pulmonary alteration in lupus patients. We highlight the characteristics observed on radiography and computed tomography, with an emphasis on computed tomography. It is important that every radiologist is prepared to recognize these findings and understand the possible clinical repercussions.

3. Patients and Methods

The study selected patients with systemic lupus erythematosus in our general hospital. Patients were considered to have Shrinking lung if the following conditions were present: compatible clinical picture (progressive dyspnea of varying effort intensity with or without pleuritic chest pain); reduction of lung volume; and no evidence of parenchymal pulmonary disease or vascular pathology in the image (chest X-ray and high-resolution computed tomography).

4. Results

Three patients met the criteria for Shrinking lung (SLS) between 2017 and 2018 in our general hospital. Of the 3 patients with SLS, two are female (50 years of age and 20 years of age). The male patient is 52 years old. Patients were referred

to pulmonology and radiology for evaluation. All 3 patients are being followed up and have low morbidity. The sociodemographic and clinical characteristics of the patients selected in this study are summarized in **Table 1**.

5. Discussion

Systemic lupus erythematosus (SLE) is a diffuse connective tissue disease that presents innumerable clinical manifestations. Involvement of the respiratory tract may occur in more than 50% of patients at some stage of the disease, and the upper airways, pleura, parenchyma, and pulmonary vessels may be involved [10] (**Figure 1** and **Figure 2**).

A rare manifestation of this disease is Shrinking Lung, the so-called shrinking lung syndrome (SPE), which is characterized by dysfunction of the respiratory muscles, specifically the diaphragm, by mechanisms not yet well defined, leading to dyspnea [10] (**Figure 3(a)** and **Figure 3(b)**).

SPE is a rare clinical condition, classically described in patients with SLE [11], and rarely in patients with other diseases [8]. It is characterized by the appearance of dyspnea and ventilatory chest pain depending on the changes seen in the pulmonary function tests, which shows restrictive ventilatory disorder, sometimes severe, and radiological findings where there is evidence of unilateral or bilateral diaphragmatic elevation and absence/minimal parenchymal involvement [8] (**Figures 4(a)-(c)**).

The pathogenesis of SPE is controversial. There is a suggestion that there is primarily a dysfunction of the diaphragmatic muscles [12], a hypothesis not always accepted by other authors [9], diaphragmatic paralysis secondary to phrenic nerve injury, diffuse fibrosis of the diaphragm [13], or restriction of chest wall expansion by other factors [9].

Treatment of SPE is also not defined. In the cases described in the world literature, were used corticosteroids [14] [15], immunosuppressants [10], inhalational beta-agonists (because of the positive inotropic effect of this substance on diaphragmatic muscle receptors) [16], xanthines [17] and even digitalis (based on the argument of which diaphragm would respond to this drug in a similar way to heart muscle) [18].

Table 1. Sociodemographic and clinical characteristics of the patients selected in this study.

Age	Sex	Clinical symptoms	Imaging studies	Findings in the image
52 years old	Man	Clinically significant dyspnea	Computed Tomography (CT)	Elevation of diaphragmatic hemicupules; volume reduction of the lower lung fields; atelectatic alterations in the pulmonary peripheries
50 years old	Female	Progressive dyspnea and mild chest pain	Chest X-Ray and Computed Tomography (CT)	Elevation of the diaphragmatic hemicupules; volume reduction of the lower lung fields; with some laminar atelectasis at the pulmonary bases
20 years old	Female	Mild respiratory discomfort	Computed Tomography (CT)	Ground glass opacities, slight pulmonary volume reduction

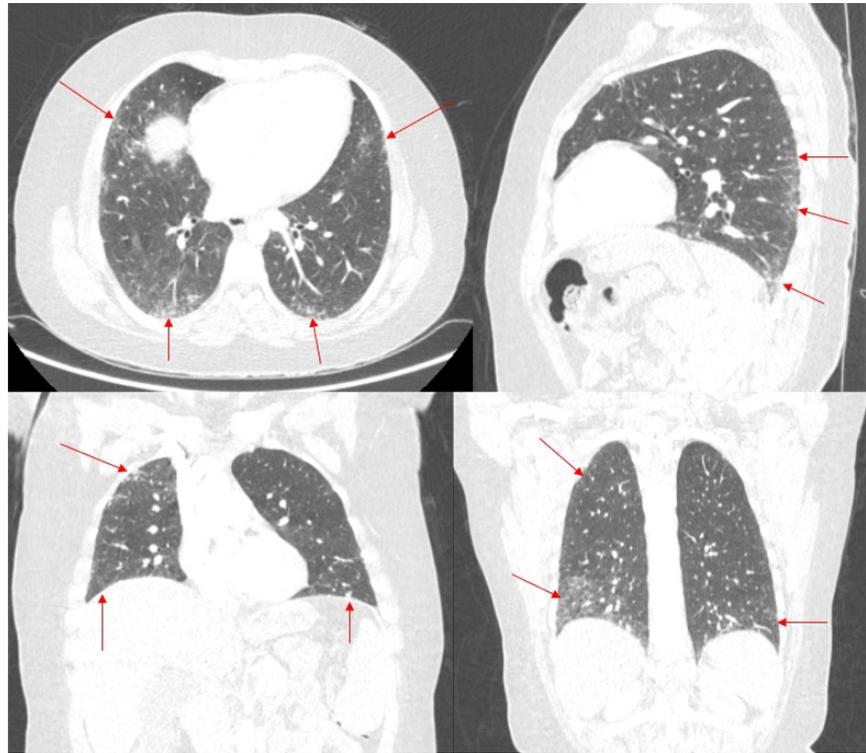


Figure 1. Young female patient with systemic lupus erythematosus diagnosed 2 years ago. Clinical signs of mild respiratory discomfort. Computed tomography in the axial, sagittal (superiorly) and coronal (inferiorly) sections, in the lung window. The images show peripheral ground glass opacities and bilateral basal predominance (red arrows), slightly more pronounced in the right lung. There is a slight pulmonary volume reduction, especially of the lower lobes.

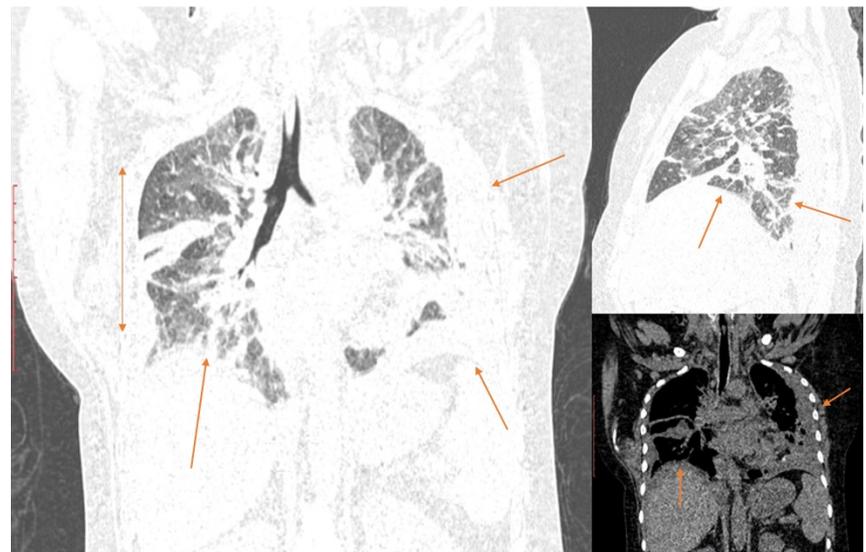
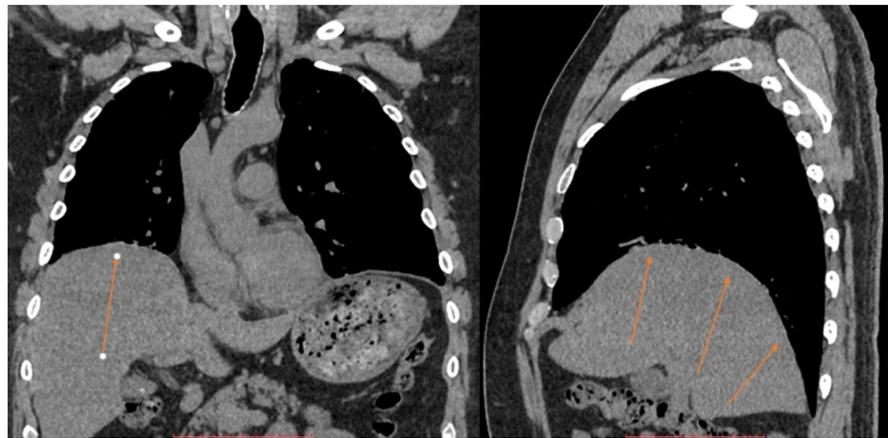
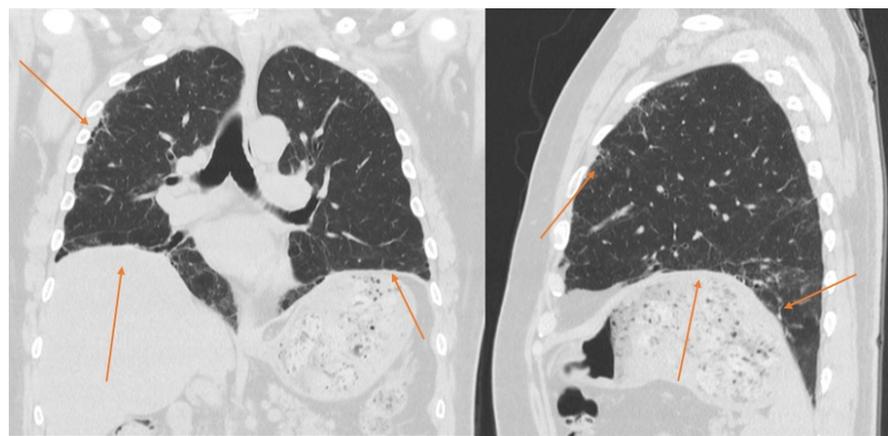


Figure 2. Young female patient with severe systemic lupus erythematosus. Clinical signs of acute respiratory failure. Computed tomography in the coronal and sagittal sections, in the lung and coronal window in the mediastinum/soft tissue window. Elevation of diaphragmatic hemicupules, volume reduction of lower lobes and signs of pulmonary congestion. Also, present bilateral pleural effusion (orange arrows).

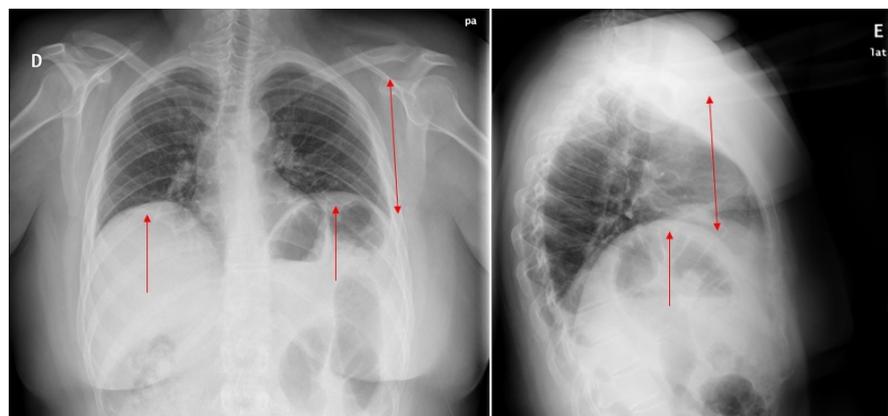


(a)



(b)

Figure 3. (a) A middle-aged male patient with systemic lupus erythematosus. Computed tomography in axial and sagittal sections in the mediastinum/soft tissue window. Evolving with clinically significant dyspnea. Elevation of the diaphragmatic hemicuples and volume reduction of the inferior pulmonary fields (orange arrows); (b) A middle-aged male patient with systemic lupus erythematosus. Evolving with clinically significant dyspnea. Computed tomography in axial and sagittal sections in the lung window. Elevation of diaphragmatic hemicuples, volume reduction of the lower lung fields, and atelectatic alterations in the pulmonary peripheries (orange arrows).



(a)

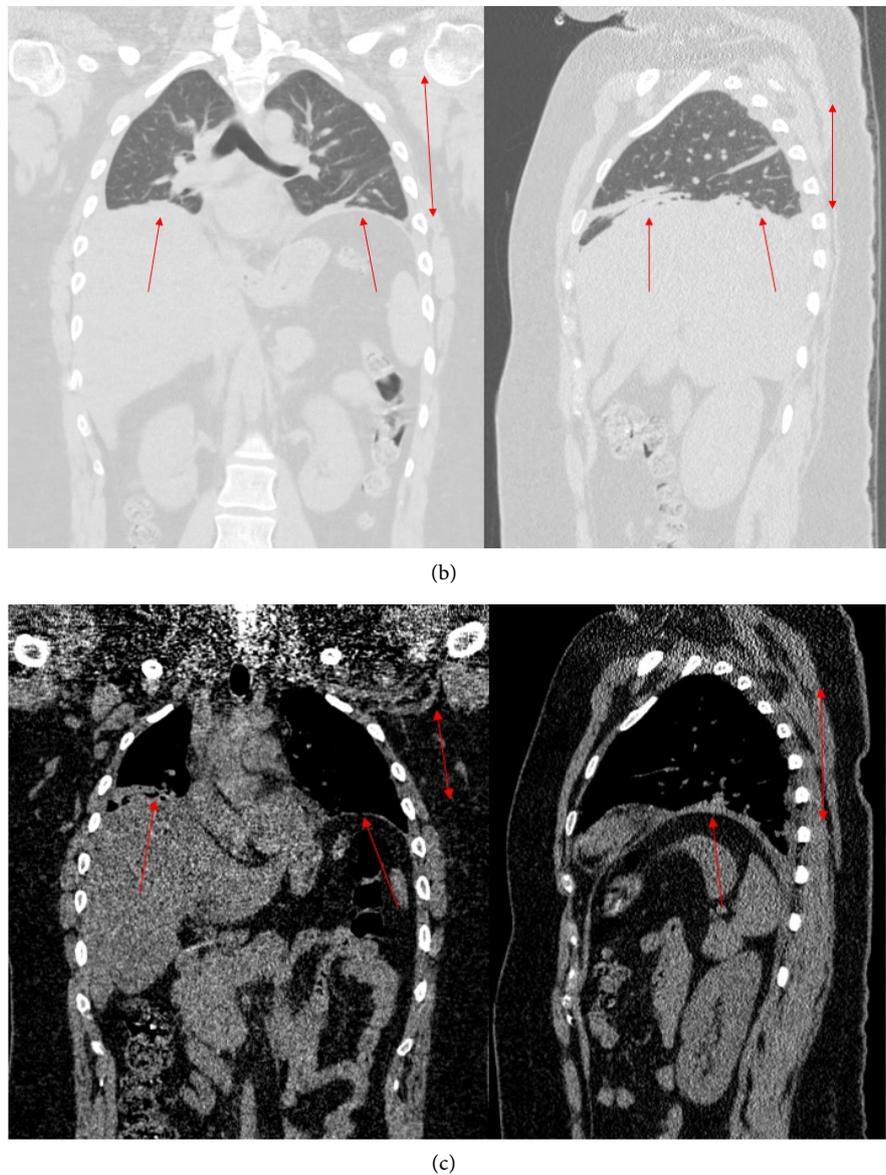


Figure 4. (a) Chest radiographs in PA and Profile of a middle-aged female patient with systemic lupus erythematosus. Clinical picture of progressive dyspnea and mild chest pain dependent. There is an elevation of the diaphragmatic hemicupules and a reduction in the volume of the lower pulmonary fields (red arrows); (b) Chest tomography in coronal and sagittal sections in the lung window of a middle-aged female patient with systemic lupus erythematosus. Clinical picture of progressive dyspnea and mild chest pain dependent. Imaging findings characterized by elevation of the diaphragmatic hemicupules and volume reduction of the lower lung fields, with some laminar atelectasis at the pulmonary bases. (Red arrows); (c) Chest tomography in coronal and sagittal sections in the mediastinal window of a middle-aged female patient with systemic lupus erythematosus. Clinical picture of progressive dyspnea and mild chest pain dependent. Imaging findings characterized by elevation of the diaphragmatic hemicupules and volume reduction of the lower lung fields, with some laminar atelectasis at the pulmonary bases. (Red arrows).

6. Conclusion

Although infrequent, shrinking lung should be included in the differential diag-

noses of lung changes seen in patients with systemic lupus erythematosus. Radiologists, pulmonologists and the medical community should be aware of this involvement, allowing better evaluation and better treatment of patients.

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