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Shrinking Lung Syndrome in Systemic Lupus Erythematosus Department of Internal Medicine. La Rabta.University Hospital

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Abstract In systemic lupus erythematosus (SLE), the respiratory system is frequently compromised. One of its uncommon manifestations is the shrinking lung syndrome (SLS), characterized by unexplained dyspnea, a restrictive pattern on pulmonary function tests, and an elevated hemidiaphragm. The pathogenesis of the SLS remains unknown. We report three consecutive cases of women with systemic lupus erythematosus, aged 32, 33, and 35 years. Clinical features included dyspnea and chest pain. Onset of the SLS manifestations occurred in the absence of a SLE flare, in patients treated with low dose of corticosteroids. Diagnosis was based on clinical and laboratory patterns, particularly chest RMI showing basilar atelectasis, elevated diaphragms, and small lung volumes in two cases. Corticosteroid treatment induced improvement in the three cases and no relapse was noted. Clinical, laboratory, physiopathological, and therapeutic aspects of this syndrome are reviewed, particularly RMI findings, never discussed in previous studies.

Keywords: *systemic lupus erythematosus, shrinking lung syndrome, magnetic resonance imagery*

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1. Introduction

Shrinking Lung Syndrome (SLS) is a rare complication of systemic lupus erythematosus (SLE). The pathogenesis of SLS remains elusive. The diagnosis of this entity is difficult and is based on clinical arguments. The contribution of imaging and in particular magnetic resonance imaging (MRI) has not been discussed in earlier publications. Herein we report 3 cases and highlight the contribution of thoracic MRI in the diagnosis of SLS.

2. Clinical Cases

Case 1: A 32-year-old female patient with past history of SLE. She presented with dyspnea on exertion and bilateral pleuritic chest pain. The patient's disease course was characterized by recurrent arthritis involving the small joints of hands and feet, one episode of pleural effusion, photosensitivity, lymphopenia, positive antinuclear antibodies (ANA), positive anti DNA, anti Sm, and anticardiolipines antibodies. She had been treated with low-dose oral steroids and with hydroxychloroquine. On physical examination, the patient was febrile, tachypneic, and unable to assume the suspine position. Auscultation of the chest revealed bilaterally decreased breath sounds. ECG, echocardiography, and arterial blood gases were normal. Chest X-ray revealed left elevated hemi-diaphram and linear opacity of the right lower lobe. Spirometry showed restrictive syndrome. Thoracic CT revealed banded lobar atelectasis in inferior lobes bilaterally and in the lingula, and triangular retracted parenchymal condensation in the lower left lobe. Thoracic MRI revealed pleural and diaphragmatic thickness with an ascent of the diaphragmatic dome and mild bilateral pleural effusion. The diagnosis of SLS was retained. The patient received 40 mg/day (1 mg/kg/day) of oral prednisone with progressive tapering, 200 mg/day of hydroxychloroquine and 600 mg/day of oral theophylline. The outcome was favorable with improvement of the respiratory symptoms. One month later control thoracic MRI was normal. Three years later, the patient was still monitored without recurrence. She is receiving 5mg of oral prednisone and 200mg/day of hydroxychloroquine as maintenance treatment.

Case 2: A 33-year-female-patient was admitted in our department for prolonged fever. Physical examination showed 38°C fever, malar rash, myalgia, bilaterally decreased breath sounds in lower lobes. Chest X ray revealed bilateral pleural effusion. Routine laboratory tests showed leucopenia, lymphopenia, autoimmune hemolytic anemia, and biological inflammatory syndrome. ANA,

anti Sm, anti-histone, anti-ribosome antibodies were positive. The patient fulfilled the ACR criteria for the diagnosis of SLE and received oral prednisone and hydroxychloroquine. The outcome was favorable. Eight months later, she presented with pleuritic chest pain and tachypnea. We did not notice other symptoms related to active SLE. Chest radiography showed a band atelectasis at the left base (Figure 1). Chest CT revealed an ascent of the left diaphragmatic dome, and parenchymal retraction of the left lung. Chest MRI showed bilaterally basal atelectasis and left diaphragmatic dome ascent (Figure 2). The diagnosis of SLS was retained and the patient received oral prednisone at the dose of 60 mg (1mg/kg/day) with progressive tapering and 200 mg/day of hydroxychloroquine. The outcome was favorable with disappearance of pleuritic pain and polypnea. Control chest X ray and MRI were normal.

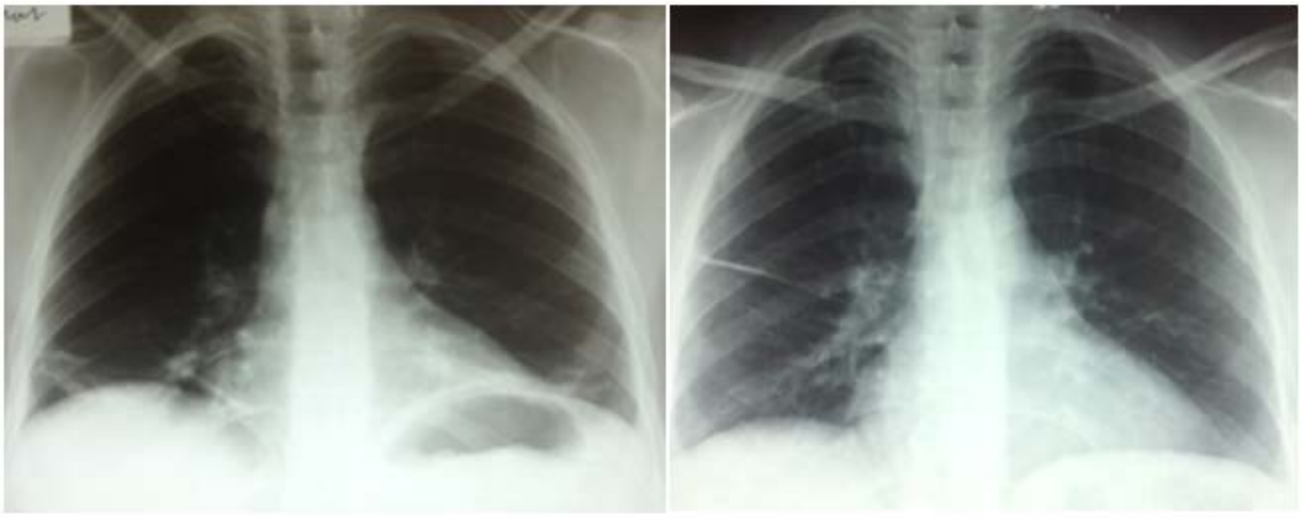

Figure 1. Posteroanterior chest radiographs of SLS (case no 2) before and treatment. The small lung volumes and elevated hemi diaphragms seen at presentation improved following therapy

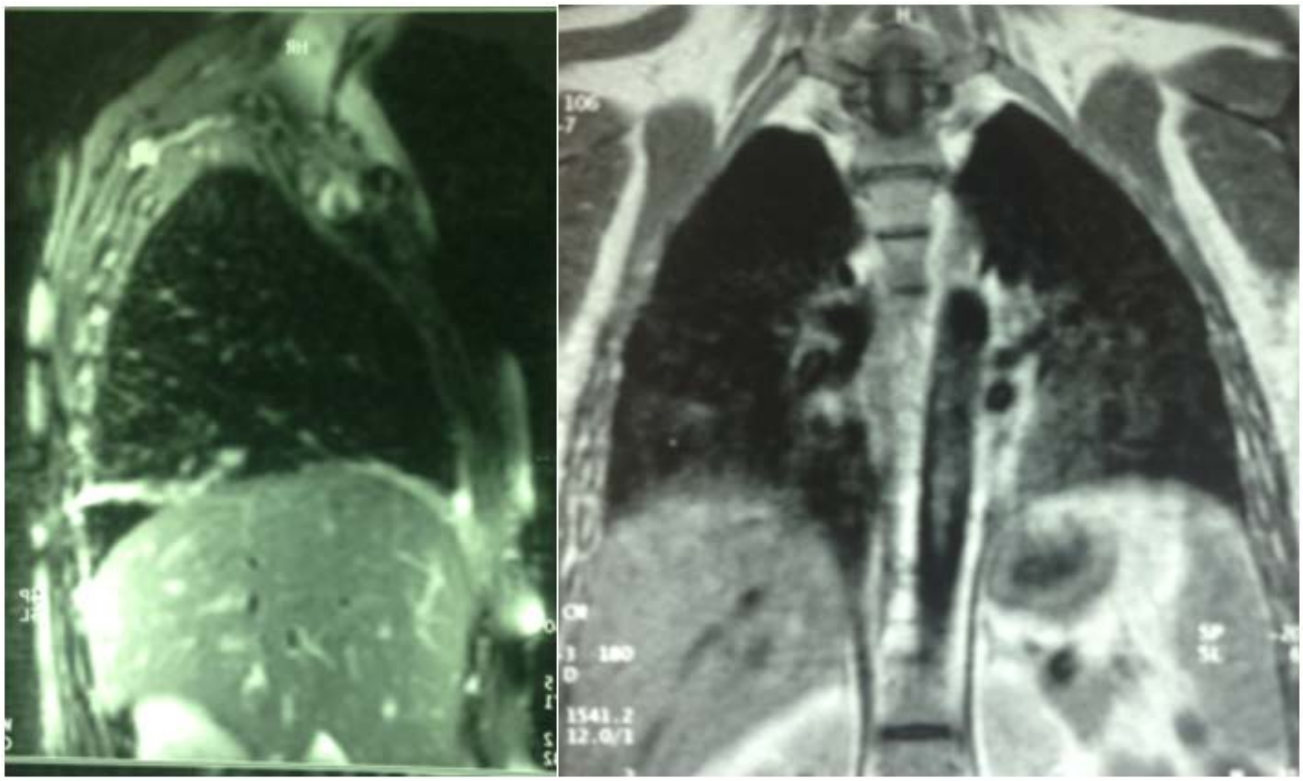

Figure 2. Chest MRI of SLS (case no2). Bilateral basal atelectasis and left diaphragmatic dome ascent

Case 3: A 35-year-old female patient with a 10-year history of cutaneo-articular SLE treated with low doses of oral steroids and hydroxychloroquine, presented with chest pain and fever. Chest X ray revealed elevation of the hemidiaphragm and linear atelectasis at the right base. Echocardiography and chest CT were normal. Spirometry revealed restrictive syndrome. For technical reasons, chest MRI could not be performed at the acute phase. The diagnosis of SLS was retained. The treatment included 50 mg (1mg/kg/day) of oral prednisone with progressive tapering and 200 mg/day of hydroxychloroquine. This led

to improvement of pain and control chest X ray showed normal lung volumes.

3. Discussion

« Shrinking Lung Syndrome » is a rare complication. Its prevalence is certainly underestimated because of lack of knowledge of various clinical and radiological presentations on one hand, and, on the other hand, the infeasibility in routine of diaphragm biopsy or electromyogram of phrenic muscle to establish the diagnosis of SLS [1]. To our best knowledge, about 105 cases have been reported in the literature [2,3,4]. SLS may complicate SLE at any time over its course, ranging from as early as a few months to 24 years after disease onset [5]. The mean time to onset of SLS after SLE diagnosis is 4.3 years and the mean age to diagnosis was 36 years [5]. Typically, it presents with progressive dyspnea, initially with activity and later at rest. Pleuritic chest pain frequently accompanies dyspnea, whereas dry cough and fever are rarely seen [6]. Elevation of the diaphragm is a universal radiographic finding [7]. Chest CT and lung V/Q scan show no evidence of parenchymal involvement or thromboembolism [7]. Pulmonary function tests are consistent with a restrictive defect [8]. SLS is characterized by unexplained dyspnea, small lung volumes, elevation of the diaphragm, and restrictive physiology [10].

The precise pathogenetic mechanism underlying the diaphragmatic dysfunction in SLS is unknown. Various pathogenetic mechanisms including myositis of the diaphragm, phrenic nerve paresis, restrictive rib cage abnormality of unknown pathology, or pleural adhesions have been evocated [10]. The volitional tests of diaphragmatic strength are also indicative of diaphragmatic weakness. The maximum transdiaphragmatic pressure, which represents the difference of gastric and esophageal pressure, is decreased during maximal inspiratory efforts. This invasive test can not be used in routine [1,11]. The contribution of MRI in the diagnosis of SLS was not discussed in previous publications. This imagery allows a range of arguments in favor of SLS. It highlights linear opacities corresponding to atelectasis at bases and rise of diaphragmatic dome. Altogether, these findings increase the specificity for a SLS. MRI can also eliminate other conditions such as pulmonary embolism, lupus-specific pneumonitis, or alveolar hemorrhage. It would be interesting to compare the chest MRI data of patients with SLS and other lupus patients with other respiratory manifestations to clarify more the contribution of MRI in the diagnosis of SLS. Besides, normal chest MRI control after steroids course in our patients can consolidate the diagnosis of SLS and the effectiveness of the prescribed treatment.

There is no definitive treatment of SLS, although glucocorticoids are considered the first line therapy, alone or in combination with other immunosuppressive agents such as azathioprine, cyclophosphamide, or methotrexate with variable succes [6,7]. Moreover, some studies have demonstrated the efficacy of theophylline (as was the case with our first observation) with the intent to increase diaphragmatic strengh and contractility [12,13]. Recently, a handful of cases of patient with non-responding SLS (neither to glucocorticoids nor immunosupresors), who showed remarkable improvement after the onset of treatment with rituximab were reported [3]. Since the pathogenesis is not fully understood, different therapeutic approaches were reported to treat SLS. However, no randomized controlled trials have been carried out or consensus reached regarding optimal therapy [2,3,14]. SLS remains a rare event in SLE, but its frequency is probably underestimated due to the lack of knowledge of its clinical and laboratory features and the absence of diagnostic criteria. The chest MRI may facilitate positive diagnosis. A descriptive and comparative study of chest MRI data of patients with SLS and other lupus patients with other lungs involvement is essential in order to identify and assess better the contribution of this review.

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