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Case Report Of Shrinking Lung Syndrome: A Rare Manifestation Of Systemic Lupus Erythematosus

[1]Dr. Anil Kumar Behera*, [1]Dr. H Guru Prasad [2]Dr. Navoday Gilla, [3]Dr. Shivaram Krishna Konda, [4]K. Divya, [5]Dr. Madhusudhan Reddy, [6]Dr. Sridhar, [7]Dr. Bharat, [8]Dr. Shailender, [9]Dr. Bharadwaj, [10]Dr. Athahullah, [11]Dr.Manaswini

[1*,4]Senior Resident, [1]Senior Consultant & Head of Department,
[2,3]Senior Consultant, [5,6,7,8]Junior Consultant, [9,10,11]Junior Resident,
Department of Internal Medicine, Care Hospitals, Banjara Hills – 500034, Hyderabad, India

***Corresponding Author:**
Anil Kumar Behera
Room No 216, Care Hospital, Banjara Hills, Hyderabad – 500034

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Abstract

Shrinking lung syndrome (SLS) is a rare pulmonary complication of Systemic Lupus Erythematous (SLE) it is characterized by progressive dyspnea, pleuritic chest pain, elevation of diaphragm and restricted diaphragmatic movement, decreased lung volume on imaging. Prevalence of SLS is estimated to be 0.5-1.1% among the patients with SLE. The pathophysiology is largely unclear, hypothesis has been suggested ranging from micro-atelectatic changes due to lack of surfactant and increased surface tension, diaphragmatic fibrosis and phrenic nerve palsy. A 50-year-old female came with the history of both leg swelling and diagnosed as bilateral lower limb cellulitis. Patient started having complaints of right-side pleuritic chest pain and mild shortness of breath. Patient was diagnosed to have SLE based on SLICC Criteria 2020. Ultrasound Diaphragm showed reduced diaphragmatic movement on the right side. Patient was diagnosed to have shrinking lung syndrome on basis of above evidence. Shrinking lung syndrome is rarely described in the medical literature with only 100 reported cases so far, giving estimated prevalence of <1. Majority of the cases are associated with dyspnea, decrease in lung volume with restrictive pattern and elevation of hemidiaphragm with reduced diaphragmatic movement. Shrinking lung syndrome is a rare manifestation of SLE. It remains under recognised and masquerades a diagnostic challenge. Patients with SLE with normal lung parenchyma, no evidence of pleural effusion and decrease in total lung capacity on PFT and elevated diaphragm with small lungs on imaging studies, reduced diaphragmatic movements on ultrasonography should alert the clinicians to raise a suspicion of SLS.

Keywords: NIL

Introduction

Shrinking lung syndrome (SLS) is a rare pulmonary complication of Systemic Lupus Erythematous (SLE) it is characterized by progressive dyspnea, pleuritic chest pain, elevation of diaphragm and restricted diaphragmatic movement, decreased lung volume on imaging [1]. Prevalence of SLS is estimated to be 0.5-1.1% among the patients with SLE [2]. The pathophysiology is largely unclear; however, hypothesis has been suggested ranging from micro-atelectatic changes due to lack of surfactant and increased surface tension, diaphragmatic fibrosis and phrenic nerve palsy [3]. There is no standardized treatment of SLS, though most of the patients are treated with medium to high dose of steroids, monoclonal antibodies to B lymphocyte antigen CD (cluster of differentiation) 20 are being studied as a possible treatment [4]. Despite lack of standardized and available therapy over all mortality seems to be low

Case report:

A 50-year-old female, with past medical history of type 2 diabetes mellitus, chronic kidney disease, came with the history of both leg swelling and diagnosed as bilateral lower limb cellulitis. Patient started having complaints of right-side pleuritic chest pain and mild shortness of breath. Patient was started with oxygen supplementation and supportive measures. Laboratory investigations such as Hemogram showed mild leukocytosis, renal parameters showed mild increase in serum creatinine. Liver parameters, complete urine examination was normal. D-DIMER and Computed Tomography – Pulmonary Angiogram was normal, ruling out Pulmonary thrombo-embolism. Arterial blood gas analysis showed respiratory acidosis due to Type II respiratory failure. High resolution computed tomography revealed restrictive pattern of lungs. Anti-Nuclear Antibody profile was sent on suspicion of Systemic Lupus Erythematous, which came out positive. Patient was diagnosed to have SLE based on SLICC Criteria 2020. Ultrasound Diaphragm showed reduced diaphragmatic movement on the right side. Patient was diagnosed to have shrinking lung syndrome on basis of above evidence. Patient was started with IV steroids, Mycophenolate Mofetil, NIV support, and other supportive measures. Patient improved symptomatically and was discharged with advice of Pulmonary Function test on follow-up.

Discussion:

Shrinking lung syndrome is rarely described in the medical literature with only 100 reported cases so far, giving estimated prevalence of <1 [5]. Majority of the cases are associated with dyspnea, decrease in lung volume with restrictive pattern and elevation of hemidiaphragm with reduced diaphragmatic movement. The greatest challenge in the diagnosis is the absence of other pulmonary pathologies such as interstitial, alveolar or pulmonary disease [1,6]. Our patient had type II respiratory failure, with diagnosis of SLE, with confirmation of restrictive changes on lung CT, with reduced movement of diaphragm – which suggests the diagnosis of SLS. A case series by Ciaffi et al. highlights the challenges associated with diagnosis of SLS due to its rarity and because of it being diagnosis of exclusion [6]. Thus, increased awareness and suspicion is critical for early diagnosis and treatment. Early diagnosis and treatment can play an important role in preventing the disease progression and improving the morbidity and mortality. SLS is a rare complication of SLE with estimation of 0.5-1% of whole SLE population. Laroche et al. [7] reported a normal maximal transdiaphragmatic pressure during bilateral electric phrenic nerve simulation in all of 10 patients tested, suggesting a failure of diaphragm activation rather than intrinsic myopathy Phrenic nerve palsy was reported in 2 isolated cases of SLS [8,9], but no evidence of demyelinating phrenic neuropathy was found in all 9 subjects tested by Wilcox et al. [10]. As pleural involvement is common in SLE, it was suggested that SLS may be caused by pleural adhesions. However, SLS has not been observed in patients with pleural adhesions of other origin, and necropsy of 1 patient with SLS showed diaphragm fibrosis and atrophy without any inflammation or pleural adhesion [11]. More recently, the hypothesis of an inhibitory intercostal-phrenic and phrenic-to-phrenic reflex caused by pleural inflammation and pain has been suggested. Indeed, pleuritic pain is a prominent feature in 65% of patients with SLS and an inhibitory reflex could explain diaphragm dysfunction during spontaneous breathing and voluntary manoeuvres, in the absence of muscle weakness. Most authors currently believe that SLS is due to multiple pathogenic mechanisms.

Although the mortality of SLS appears to be low, it may lead to significant disability [12]. Some patients only experience improvement in symptoms but no change in lung function It has even been suggested that the perceived improvement could result more from the patient's adaptation to his or her limited functional capacity rather than to a real improvement.

Due to its rarity and uncertainty about its cause, the treatment of SLS is not standardized. Several reports describe the efficacy of variable doses of corticosteroids, either alone or combined with azathioprine, cyclophosphamide or methotrexate [13]. Anecdotic reports suggest a modest benefit of theophylline and β-agonists.

Conclusion:

Shrinking lung syndrome is a rare manifestation of SLE. It remains under recognised and masquerades a diagnostic challenge. Patients with SLE with normal lung parenchyma, no evidence of pleural effusion and decrease in total lung capacity on PFT and elevated

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diaphragm with small lungs on imaging studies, reduced diaphragmatic movements on ultrasonography should alert the clinicians to raise a suspicion of SLS.

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