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Tofacitinib Treatment in Refractory Systemic Lupus Erythematosus with Shrinking Lung Syndrome: A Case-Based Review

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ABSTRACT

Shrinking Lung Syndrome (SLS) is a rare pulmonary complication of Systemic Lupus Erythematosus (SLE), which mainly manifests as dyspnea, pleural chest pain, and restrictive ventilation dysfunction, with or without an elevated hemi-diaphragm. Currently, there is no unified diagnostic criteria for SLS. SLS is a diagnosis of exclusion and can be diagnosed only in the absence of interstitial, alveolar and pulmonary vascular disease. The treatment is based on high-dose glucocorticoids and immunosuppressants. Here, we report a case of a 35-year-old woman who was diagnosed with SLE in 2008. Four years later, she was diagnosed with SLS due to chest pain, dyspnea, cough, and restrictive ventilatory dysfunction. Her condition improved after treatment with glucocorticoids and immunosuppressants but relapsed many times. She was treated with tofacitinib after relapse in May 2019, and then her condition improved and remains in remission with a four-year follow-up. We searched for and summarized 15 cases of patients with refractory SLE combined with SLS. They were all treated with biological agents. Tofacitinib is a new treatment choice for patients with refractory SLE with SLS.

Keywords: Tofacitinib; Systemic Lupus Erythematosus; Shrinking Lung Syndrome

Abbreviations: SLS: Shrinking Lung Syndrome; SLE: Systemic Lupus Erythematosus; FVC: Forced Vital Capacity; DNA: Anti-Double-Stranded; ANA: Anti-Nuclear Antibody; SM: Anti-Smith; TLC: Total Lung Capacity; DLCO: Diffusing Capacity of the Lungs for Carbon Monoxide; JAK: Janus kinase

Introduction

SLE is a chronic autoimmune connective tissue disease involving multiple systems. SLE occurs mainly in females of childbearing age and is characterized by the production of high level of autoantibodies. The lungs of approximately 30%-50% of lupus patients are affected in various ways [1], mainly pleural disease, lupus pneumonia, pulmonary hypertension, and pulmonary thromboembolism. SLS is a rare complication of SLE, and it is estimated to occur in 0.5% to 1.53% of all lupus patients and can occur at any point in the course of the disease [2,3]. The main clinical manifestations are dyspnea and pleural chest pain, and approximately half of the patients have active extra-thoracic disease at the time of diagnosis. The specific physiological and pathological mechanisms of SLS are still unclear [4]. At present, the relevant research on SLE combined with SLS is mainly based on case reports. The mortality rate after treatment is generally low, but the improvement of lung function is often limited. In Langenskiöld's research, it was found that only 20% of SLS patients recover a Normal Forced Vital Capacity (FVC), challenging the common belief that SLS has a good prognosis [5]. We report a case of SLE with SLS whose condition greatly improved and was able to reduce the dose of glucocorticoids after tofacitinib treatment. Through a literature review, we aimed to explore new treatments for SLS.

Case Presentation

A 35-year-old woman was admitted to the hospital with Raynaud's phenomenon, fever, cough, chest pain, and breathlessness. Since 2007, she began to have Raynaud's phenomenon of both of her hands. In January 2008, she had fever without an apparent cause (~40°C), accompanied by cough, shortness of breath, and chest pain. The examination revealed pleural effusion, Anti-Nuclear Antibody (ANA) positivity, Anti-Smith (Sm) antibody positivity, Anti-Double-Stranded DNA (dsDNA) antibody positivity, decreased complement, and slight proteinuria, and she was diagnosed with SLE. After treatment with prednisone 10 mg three times a day, the symptoms were relieved, and the proteinuria was resolved. When the prednisone dose was gradually reduced to 2.5 mg per day in October 2010, the symptoms recurred. She suffered a cough, chest pain, dyspnea, and fever with a Tmax of 39°C and was unable to lie on her right side. After treatment with prednisone 40 mg per day, HCQ 0.2 g twice a day and MMF 750mg twice a day, the symptoms were relieved. In July 2012, she experienced fever, cough, expectoration, chest pain and chest tightness. The chest pain worsened while lying on her right side and at the end of expiration. In addition, the thoracic expansion was less than 2.5 cm. Laboratory test parameters were as follows: C-reactive protein, 79.42 mg/L and erythrocyte sedimentation rate, 69 mm/h. Chest X-ray: there is no significant elevation of the diaphragm (Figure 1). Chest CT showed a slightly thickened right pleura (Figure 2).

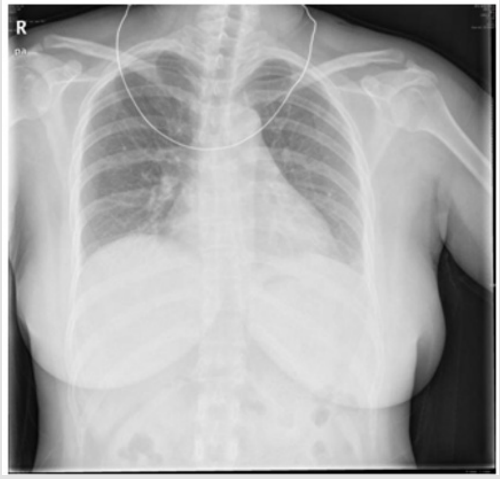

Figure 1: Chest X-ray of the patient (2012-09): No significant elevation of the diaphragm.

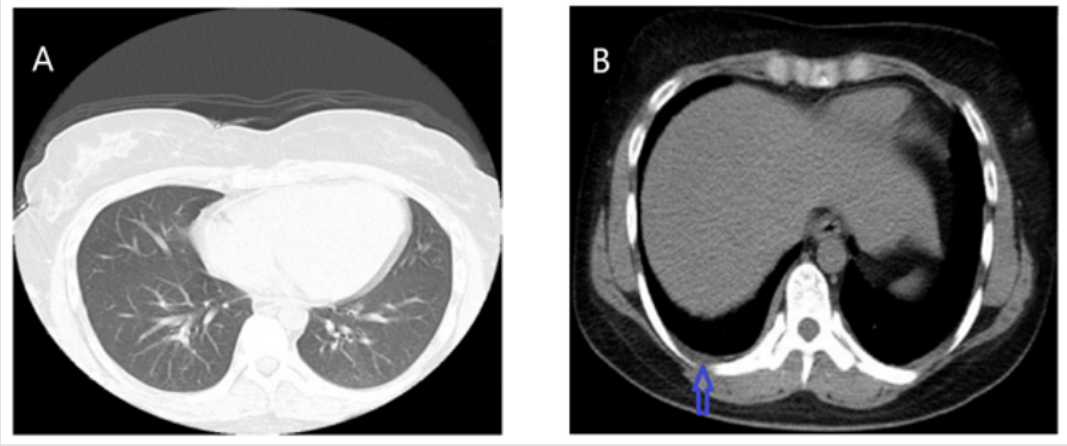

Figure 2: CT scan of the chest of the patient (2012-09).

A. The lung field is basically clear, no obvious atelectasis and interstitial changes;

B. The right pleura is slightly thickened, and there is no other obvious abnormality.

Pulmonary Function Test (PFT) parameters were as follows: forced expiratory volume in 1 second (FEV1)/Forced Vital Capacity (FVC), 86.76%; FVC, 1.2/36.7%; Total Lung Capacity (TLC), 2.55/53.4%; and Diffusing Capacity of the Lungs for Carbon Monoxide (DLCO), 1.08/57%. Echocardiography showed a pulmonary arterial pressure of 28 mmHg. After the exclusion of diseases such as pulmonary fibrosis, pulmonary hypertension, and pleural effusion, she was diagnosed with SLS. After treatment with methylprednisolone 40 mg per day, HCQ 0.2 g twice a day and AZA 100 mg per day, symptoms such as cough, chest tightness and chest pain improved. Upon PFT re-examination, FVC was 1.25/35% and TLC was 2.47/52%. Thereafter, symptoms recurred when prednisone was decreased to 15-20 mg per day. In May 2019, she was hospitalized with fever, chest pain, and dyspnea. Physical examination showed that breath sounds in both lungs were slightly lower, and dry rales were slightly detected in the lower lobe of the lungs. PFT parameters were as follows: FEV1, 54.7%; FVC, 58%; TLC, 60.7%; and DLCO, 53.9%. After treatment with prednisone 30 mg per day, tofacitinib 5 mg twice a day and mycophenolate mofetil 0.5 g twice a day for 2 weeks, her symptoms improved significantly. When prednisone was reduced to 10 mg per day in September 2019, she suffered chest pain and dyspnea again. Upon re-examination, her PFT parameters were as follows: FEV1, 47.7%; FVC, 46.2%; TLC, 52.6%; and DLCO, 41.8%. Tofacitinib was increased to 10 mg twice a day, and she improved again after 2 weeks of treatment. Thereafter, the prednisone was gradually reduced to 5 mg per day. In November 2020, her PFT parameters were as follows: FEV1, 63.8%; FVC, 61.9%; TLC, 61.3%; and DLCO, 43.4%. At present, she is in sustained remission, with a follow-up of four years.

Search Strategy

A comprehensive literature review was performed using electronic search platforms and databases, including PubMed, Web of Science and Medline, from inception to January 31, 2021, to look for cases of SLE with SLS. The following keywords and Mesh headings were used alone or in combination: “systemic lupus erythematosus”, “lupus”, “SLE”, “shrinking lung syndrome”, and “SLS”. We reviewed abstracts of related studies and selected appropriate articles. Two authors independently reviewed abstracts and retrieved articles based on the following criteria:

1. Articles that were case reports and case series of SLE combined with SLS with complete clinical data.
2. Articles with intractable cases treated with biological agents (glucocorticoids and immunosuppressive agents were ineffective or not applicable); and
3. Articles published in a peer-reviewed journal. Articles that did not meet the above inclusion criteria were excluded.

The following data were extracted from the articles: first author and year of publication, age, sex, clinical manifestations, autoantibodies, chest imaging, PFT parameters, reasons for treatment with biological agents, treatment, and outcome.

Literature Review

After a thorough literature review, we found 15 cases of SLE combined with SLS in which the patients were treated with biological agents, and all cases are summarized in Table 1. Among these 15 patients, there were 12 female patients and only 3 male patients. Except for the age of one male patient, which was not recorded, the average age of all patients was 36.3 years, reflecting that SLE with SLS is more common in young and middle-aged female patients. From the perspective of clinical symptoms, chest pain and dyspnea were the most common. Every patient had dyspnea, and chest pain was also seen in the vast majority of patients (11/15). Although the frequency was not high, cough (5/15) and fever (4/15) were also important clinical symptoms of SLS. For autoantibodies, excluding the 3 patients with missing data, the positive rate from highest to lowest was ANA (11/12), anti-ds-DNA antibody (10/12), anti-SSA antibody (5/12), anti-Ro antibody (3/12) and anti-Sm antibody (3/12), reflecting the high correlation between anti-SSA antibody and anti-Ro antibody in SLS. Regarding imaging manifestations, SLS patients mostly presented with diaphragm elevation (10/15), lung volume reduction (4/15), atelectasis (4/15) and pleural thickening (3/15), and diaphragm elevation was the most common imaging manifestation in SLS. Pulmonary function presented a restrictive pattern, showing decreased FEV1, FVC, TLC, and DLCO. FEV1 drops to 26% and FVC drops to 23% in the most severe patients, indicating that SLS patients can have extremely severe dyspnea.

Table 1: Summary of published cases of SLE with SLS in which biological agents were administered.

(Ref.) First author and year	Age, yrs/sex	Clinical manifestations	Auto-antibodies	Chest imaging	PFT	Reasons for using biological agents	Treatment	Outcome
Ciaffi et al. 2019 [6]	19/M	Pleuritic pain, Dyspnea, Cough	ANA,ds-DNA, LAC,U1RNP, RNP	Mild follicular bronchiolitis	FVC 65% FEV1 64% TLC 65% DLCO 69%	PFTs decline, conventional treatments failure	Belimumab 200 mg/wk	Restrictive defect improvement
Choudhury et al. 2020 [7]	51/F	Pleuritic pain, Dyspnea, Cough, Low-grade fever	NA	smaller lung Volume, Elevated Diaphragm	FVC 23% FEV1 26% TLC 38% DLCO 36%	symptoms decline, conventional treatments failure	Belimumab	Restrictive defect improvement
Reyes et al. 2016 [8]	44/F	dyspnea	NA	normal	Restrictive pattern	conventional treatments failure	Belimumab	Improvement
Duron [9] et al. 2016	62/F	Pleuritic pain, Dyspnea	ANA, ds-DNA, SSA, SSB	Elevated Diaphragm, Atelectasis, Pleural Thickening	TLC 46% DLCO 25% KCO 59%	conventional treatments failure	Rituximab	Improvement
Duron [9] et al. 2016	26/F	Pleuritic pain, Dyspnea	ANA, ds-DNA, RNP	Pleural Thickening, Reticulations	FVC 41% TLC 68% DLCO 34%	Class III glomerulonephritis and SLS	Rituximab	Improvement
Thordardottir et al. 2017 [10]	55/F	Pleuritic pain, Dyspnea, fever	ANA, ds-DNA, Ro ,SSA	Elevated Diaphragm, Atelectasis, Pleural effusion	FVC 36% FEV1 38% TLC 46% DLCO 44%	Difficulty in CS withdrawal Rituximab	Infliximab	deterioration
							Improvement	
Fleri et al. 2017 [11]	32/F	Pleuritic pain, Dyspnea, Cough	ANA, ds-DNA, Sm	smaller lung Volume, Elevated Diaphragm	FVC 57% FEV1 53% TLC 60% DLCO 63%	PFTs decline, conventional treatments failure	Rituximab	Severe allergic rash
Burns et al. 2014 [12]	13/F	Pleuritic pain, Dyspnea	ANA, ds-DNA, Sm,SSA	low lung volumes, left pleural effusion	FVC 31% TLC 32%	conventional treatments failure	Rituximab	Improvement
Toya et al. 2009 [13]	46/F	Pleuritic pain, Dyspnea	dsDNA, Ro, LAC	Elevated Diaphragm, Atelectasis, Pleural Thickening	FVC 57% FEV1 53% TLC 60% DLCO 63%	conventional treatments failure	Rituximab	Improvement

Dorval et al. 2020 [14]	NA/M	Dyspnea	NA	Elevated Diaphragm	Restrictive pattern	conventional treatments failure	Rituximab	Failure, developed anti-rituximab antibodies
Benham et al. 2010 [15]	27/F	Pleuritic pain, Dyspnea	ANA, ds-DNA	Elevated Diaphragm	FVC 35% FEV1 37% TLC 41% DLCO 62%	conventional treatments failure	Rituximab 375 mg/m2 q6w,Two doses	Improvement
Goswami et al. 2016 [16]	38/F	Dyspnea, Fever	ANA, Ro	Elevated Diaphragm	FVC 48% FEV1 46%	conventional treatments failure	Rituximab 1g q2w two doses	Improvement
Langenskiöld et al. 2012 [5]	28/F	Pleuritic pain, Dyspnea, Cough	ANA, SSA,SSB	Elevated Diaphragm	FVC 43% FEV1 38%	conventional treatments failure, rapidly respiratory deterioration	Rituximab 2g, 2 weeks apart every 6 months,2 cycles	Improvement
Peñacoba et al. 2014 [17]	57/F	Dyspnea	ANA, ds-DNA	Elevated Diaphragm, Atelectasis	FVC 43% FEV1 46% TLC 56% DLCO 55%	Difficulty in CS withdrawal	Rituximab 1g, 2 weeks apart every 6 months,2 years	Improvement
DeCoste et al. 2021 [18]	11/M	fever, pleuritic pain, dyspnea, cough	ANA, ds-DNA, Sm SSA	low lung volumes	FVC 29% FEV1 27% TLC 43%	conventional treatments failure	Rituximab 1 g, 2 doses, 2weeks apart	Improvement

We reviewed all refractory cases in which biological agents were administered. Most of the patients were treated with biological agents because the symptoms and PFT parameters did not improve after treatment with glucocorticoids and immunosuppressants (12/15). Two patients experienced relapse after glucocorticoid reduction [5,6], and one patient was treated with rituximab due to grade III glomerulonephritis [7]. Rituximab was most widely used drug in the treatment of SLE with SLS (12/15); except one patient in whom rituximab efficacy was poor because she developed anti-rituximab antibody [8] and one who had severe allergic rash after rituximab use [9], the other patients (10/12) experienced a significant curative effect with rituximab. Belimumab successfully treated three patients with SLE with SLS [10-12], and no adverse reactions were reported. However, the determination of the clinical efficacy of belimumab in patients with SLE with SLS requires further observation due to the small sample size. Only one patient was treated with infliximab, and the efficacy was not satisfactory [5]; more cases are needed to verify the therapeutic effect of infliximab in the treatment of SLE with SLS.

Discussion

SLS is a rare complication of SLE. SLS is more prevalent in females (F/M ratio=17:1), and the average age of onset is 36±12 years [2]. There is often a long delay between the diagnosis of SLS and the diagnosis of SLE [13]. The main symptoms of SLS are dyspnea and pleural chest pain; some patients can also have fever, cough and other symptoms, and lung auscultation is often normal. Regarding autoantibodies, in addition to SLE-related antibodies such as ANA and anti-dsDNA, anti-RNP and anti-SSA/Ro antibodies have a higher detection rate in SLS patients [14]. Loïc Duron's study confirmed that radiographic or CT scans showed reduced lung volumes with elevated hemidiaphragms in 12 of 15 patients (80%), basal atelectasis in 8 of 15 (53%) patients, pleural thickening in 4 of 15 patients (27%), and pleural effusion in 2 of 15 patients (13%) [7]. The pulmonary function test mostly presents a restrictive pattern, which is manifested as a decrease in FEV1, FVC, TLC and DLCO [15]. Ultrasound may show abnormal movement of the diaphragm [16]. Currently, there are no unified diagnostic criteria for SLS, and the diagnosis is usually based

on typical symptoms, chest imaging and PFTs, excluding other causes of restrictive respiratory defects such as pulmonary fibrosis, obesity, and diaphragmatic paralysis [7].

The physiopathological mechanism of SLS is still unclear. Some hypotheses were proposed in previous studies. First, a group of researchers proposed that there is a deficiency in the surface film (a lipoprotein) lining the alveoli of the normal aerated lung, and the resultant excessive surface tension results in atelectasis and hyaline membrane formation.

This deficient surface film is thus liable to become involved in inflammatory processes in the alveolar walls and perivascular and peribronchial connective tissues and therefore may be responsible for the changes in the mechanical properties of the lungs in this condition [17]. Second, another group proposed that SLS is probably due to a dysfunction of the diaphragm rather than to primary intrapulmonary pathology. These authors believe that recurrent pleurisy and pleural adhesions may lead to the inability of the diaphragm to generate normal pressure, resulting in diaphragm weakness and restrictive ventilatory dysfunction [18]. A third group proposed that the peripheral neuropathy in SLE, which involves the deposition of immune complexes and ischemic lesions, can possibly lead to severe diaphragm palsy and diaphragm fibrosis. This may be the reason why SLS can show a restrictive ventilatory defect and decreased diaphragm strength [19]. Fourth, Henderson [14] believes that pleural inflammation triggers the inhibition of deep inspiration by neural reflexes and pain, resulting in chronic lung hypoinflation, which in predisposed patients leads to parenchymal remodeling, which decreases lung compliance. Impaired compliance worsens hypoinflation, initiating a positive feedback loop that helps to explain the gradual progression of SLS. This emphasizes the role of pleural inflammation in the pathogenesis of SLS. At present, there are no evidence-based guidelines for the treatment of shrinking lung syndrome. Generally, the treatment is based on high-dose glucocorticoids and immunosuppressants (azathioprine and cyclophosphamide are most commonly used).

The usual dose is between 0.5 and 1 mg/kg prednisolone daily and may be preceded by a short course of intravenous methylprednisolone depending on initial severity. For refractory cases that are not effectively treated with conventional therapy, biological agents are often chosen [11]. As mentioned above, the biological agents used to treat SLS documented in the literature are rituximab, belimumab, and infliximab, all in the form of case reports. There is still a lack of large sample randomized controlled studies to provide conclusive clinical evidence. Our patients mainly presented with chest pain, dyspnea, fever, and a restrictive pattern in PFTs. However, X-rays and chest CT showed no obvious abnormalities, such as diaphragm elevation or lung volume reduction, which may be related to active treatment at the early stage of the disease. Her symptoms were relieved after regular treatments, such as glucocorticoids and immunosuppressants, but relapsed many times after glucocorticoid reduction. In 2019, she relapsed again, and the effect of high-dose glucocorticoids and immunosuppressants (cyclophosphamide) was not satisfactory. Therefore, tofacitinib was chosen for treatment, which successfully alleviated the symptoms, enabling a reduction in the dose of glucocorticoids. A recent study found that the tumor necrosis factor alpha inducible protein 3 (TNFAIP3) gene is significantly related to the clinical phenotype of pleural inflammation in SLE [20]. TNFAIP3 encodes ubiquitin-modifying enzyme A20.

A20 is an inhibitor of the NF-κB signaling pathway. A lack of A20 can lead to an increase in the expression of downstream inflammatory cytokines such as TNF-α and IL-17 [21], which might be the reason why TNFAIP3 is obviously related to pleural inflammation in SLE. Tofacitinib is a Janus kinase (JAK) inhibitor that can downregulate the expression of interleukin, interferon, and other inflammatory factors by inhibiting the JAK-STAT signaling pathway [22]. We speculate that tofacitinib may reduce inflammatory factors and alleviate pleural inflammation, thereby exerting a therapeutic effect on SLE combined with SLS. SLE combined with SLS often occurs in women of child-bearing age, who often have fertility needs. Nevertheless, commonly used immunosuppressants such as cyclophosphamide and mycophenolate mofetil are teratogenic [23]. Our review also found that glucocorticoids and immunosuppressants are sometimes ineffective. Furthermore, there were also some patients who relapsed after glucocorticoid reduction [24]. Therefore, biological agents have essential value for the treatment of SLE combined with SLS. This case report is the first in which tofacitinib was successfully used to treat SLE combined with SLS, enabling glucocorticoid reduction [25-28]. Therefore, tofacitinib can provide clinicians with a new option when treating patients with refractory SLE combined with SLS.

Conclusion

Although extremely rare, it is important to recognize SLS as a possible cause of dyspnea and chest pain in a SLE patient and be aware that the complication may occur at the time of initial presentation of SLE [29]. Recent literature on biologicals use in SLS, and our case report suggests that additional therapy, including possible use of tofacitinib, should be considered in patients with SLS who have an incomplete response to initial immunosuppressive therapy [30]. However, data from randomized controlled trials are needed to evaluate the safety and clinical efficacy of tofacitinib in systemic lupus erythematosus with shrinking lung syndrome.

Authors' Contributions

Nan He and Bo Feng contributed equally to this paper, they were involved in the conception, design, data acquisition and write-up of the manuscript. Yongmei Han contributed to data acquisition, critical review of manuscript for important intellectual content and final approval of the manuscript. Mengjia Shen participated in the literature search and provided feedback on revisions.

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Informed Consent

Informed consent was obtained from the individual participant included in the study.

Conflict of Interest

The authors have declared no conflicts of interest.

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