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Review

Shrinking lung syndrome associated with systemic lupus erythematosus: A multicenter collaborative study of 15 new cases and a review of the 155 cases in the literature focusing on treatment response and long-term outcomes☆

Loïc Duron [a,1], Fleur Cohen-Aubart [a,b,*,1], Elisabeth Diot [c], Raphaël Borie [d], Sébastien Abad [e], Christophe Richez [f], Christopher Banse [g], Olivier Vittecoq [g], David Saadoun [h], Julien Haroche [a,b], Zahir Amoura [a,b]

[a] AP-HP, Service de Médecine Interne 2, Centre National de Référence du Lupus Systémique, Syndrome des Anticorps Anti-phospholipides et Maladies Auto-immunes Systémiques Rares, Groupe Hospitalier Pitié Salpêtrière, Paris 75013, France
[b] Université Paris VI Pierre et Marie Curie, Sorbonnes Universités, Paris 75013, France
[c] Service de Médecine Interne B, Pôle Médecine Interne et Gériatrique, Pneumologie, CHRU de Tours–Hôpital Bretonneau, 37044 Tours Cedex 9, France
[d] Service de Pneumologie A, Groupe Hospitalier Bichat-Claude Bernard, APHP, Paris, France
[e] Service de Médecine Interne, Hôpital Avicenne, APHP, 93000 Bobigny, France
[f] Département de Rhumatologie, Hôpital Pellegrin, CHU de Bordeaux, Place Amélie-Raba-Léon, 33076 Bordeaux, France
[g] Service de Rhumatologie, CHU-Hôpitaux de Rouen, Inserm U905, CRB CIC 1404, IRIB, Université de Rouen, 76031 Rouen Cedex, France
[h] AP-HP, Service de Médecine Interne 1, Groupe Hospitalier Pitié-Salpêtrière, Paris, France

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ABSTRACT

Introduction: Shrinking lung syndrome (SLS) is a rare respiratory manifestation of systemic lupus erythematosus (SLE), characterized by dyspnea, chest pain, elevated hemidiaphragm and a restrictive pattern on pulmonary function tests. Here, we report 15 new observations of SLS during SLE and provide a systematic literature review. We studied the clinical, biological, functional and morphologic characteristics, the treatments used and their efficacy.

Methods: The inclusion criteria were all patients with SLE defined by the American College of Rheumatology criteria Hochberg (1997) , associated with a restrictive pattern on pulmonary function tests. The exclusion criteria were all differential diagnoses of restrictive patterns, including obesity and pulmonary fibrosis. The patients were recruited from local databases through chest physicians, rheumatologists and internists. The data for the literature review were extracted from the Medline database using "shrinking lung syndrome" and "lupus" as key words.

Results: All 15 new cases were women with a median age at SLS onset of 27 years old (range 17–67 years). All of them complained of dyspnea and all but one of chest pain. The antibodies were similar to those found in SLE, although the anti-SS-A was positive in 10 of 13 cases. Thoracic imaging showed elevated hemidiaphragm (12/15) and/or basal atelectasia (8/15). All of the patients had an isolated restrictive pattern on PFT, with a median decrease >50% of lung volume. All of the patients were treated, using corticosteroids (11/15), immunosuppressive drugs (8/15), beta-mimetics (2/15), physiotherapy (3/15) and/or colchicine (1/15). Improvement was described in 9 of 12 patients and stability in 3 of 12. We extracted 155 cases of SLE-associated SLS from the Medline database. The clinical, biological and functional parameters were similar to our cases. Clinical improvement was described in 48 of 52 cases (94%) and PFT improvement in 36 of 47 cases. Worsening occurred in 4 cases.

Conclusion: SLS is a rare SLE manifestation. Pain and parietal inflammation seem to play important pathogenic roles. Steroids and antalgics are the most commonly used therapies with good responses. There is no proof of efficacy with immunosuppressive drugs for this entity. Rituximab can be discussed after failure of corticosteroids, as well as antalgics, theophylline and beta-mimetics.

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* Corresponding author at: Service de Médecine Interne 2, Groupe Hospitalier Pitié-Salpêtrière, 47-83 Boulevard de l'Hôpital, 75651 Paris Cedex 13, France. Fax: +33 1 42 16 58 04.
E-mail address: fleur.cohen@psl.aphp.fr (F. Cohen-Aubart).
[1] Contributed equally to this work

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Contents

1. Introduction

Systemic lupus erythematosus (SLE) is a rare auto-immune disease principally affecting young women. It is characterized by several features, including arthritis, cutaneous and renal involvement, serositis, central nervous system disorders and hematologic abnormalities, associated with the presence of auto-antibodies targeting double-stranded DNA [2].

Respiratory involvement in SLE is common, affecting 60–80% of patients [3–5]. It consists mostly of pleuritis, alveolar hemorrhage, pulmonary embolism, pulmonary hypertension, acute pneumonitis, chronic interstitial lung disease and, less frequently, shrinking lung syndrome (SLS).

SLS is a rare although debilitating condition, involving up to 1% of SLE patients [6,7]. It was first described in 1965 by Hoffbrand and Beck, as a constellation of respiratory symptoms including dyspnea and pleuritic chest pain, associated with reduced lung volumes as demonstrated by elevated hemidiaphragms on chest radiography and a restrictive defect on pulmonary function tests (PFTs). SLS has been described in the spectrum of SLE, and it has been very rarely associated with other connective tissue diseases [8–12]. Differential diagnoses include restrictive respiratory defect due to pulmonary fibrosis, obesity, diaphragmatic palsy and central nervous system disorders.

There are no definite criteria for the diagnosis of SLS, and it usually relies on the association of reduced lung volumes and restrictive defects on PFTs, together with the exclusion of other causes. The physiopathology of SLS is not well understood, nor are its treatment response and long-term outcomes.

Here, we report 15 new patients with SLE-associated SLS, and we provide an exhaustive review of all cases of this rare association reported in the French and English literature, focusing on clinical, biological and radiological presentation, treatment response and long-term outcome.

2. Patients and methods

2.1. Patient selection and follow-up

We conducted a retrospective, multicenter study to identify and describe new cases of SLE-associated SLS. The identification of cases was achieved by interviewing French internal medicine physicians, chest physicians and rheumatologists.

Data were extracted retrospectively from medical records.

Patients were included in the study if they had

1. SLE defined by the American College of Rheumatology criteria [1]; and
2. A restrictive defect on PFT with total lung capacity (TLC) < 80% or forced vital capacity (FVC) < 80%.

They were excluded if there was an alternative cause of a restrictive pattern on PFT, including pulmonary fibrosis or obesity with body mass index (BMI) > 40 kg/m^2.

Demographic, clinical, biological and radiographic data were collected. Functional data, including FVC, TLC, forced expiratory volume in 1 s (FEV1), carbon monoxide diffusing capacity (DLCO) and carbon monoxide transfer coefficient (KCO), were analyzed.

All treatments received were noted, as well as the evolution of symptoms and PFT and imaging data.

2.2. Literature review

We searched through the National Library of Medicine's MEDLINE database for relevant literature using the key words shrinking lung syndrome. Sixty references were returned in the English, French, Portuguese ($n = 1$) and Spanish ($n = 1$) literature. We selected 155 patients from 58 articles published between 1965 and 2014 in the English and French literature.

Only patients with definite SLE were finally included in the study.

The demographic and clinical characteristics, diagnostic modalities and biological and functional results were collected, as well as the treatments received and outcomes.

3. Results

3.1. Reported cases

3.1.1. Patient characteristics

The clinical, biological and radiological findings of the new cases are reported in Table 1 and are summarized in Table 2.