EJCRIM
European Journal
of Case Reports
in Internal Medicine

SHRINKING LUNG SYNDROME WITH FAVOURABLE RESPONSE TO BELIMUMAB AND CORTICOSTEROIDS ALONE

Rémy Père[1], Anouk Le Goueff[2], Marina Nguenang[1]

[1] Department of Pneumology, Centre Hospitalier de Luxembourg, Luxembourg, Luxembourg
[2] Department of Immuno-Allergology, Centre Hospitalier de Luxembourg, Luxembourg, Luxembourg

Corresponding author's e-mail: remy.pere@ulb.be

Received: 05/07/2026
Accepted: 08/07/2026
Published: 16/10/2026

Conflicts of Interests: The Authors declare no conflict of interest.
Patient Consent: Written informed consent was obtained from the patient's parents for the publication of her clinical history and any accompanying images.
This article is licensed under a CC Attribution-NonCommercial-NoDerivatives 4.0 License

How to cite this article: Père R, Le Goueff A, Nguenang M. Shrinking lung syndrome with favourable response to belimumab and corticosteroids alone. EJCRIM 2026;13:doi:10.12890/2026_007315

ABSTRACT

Introduction: Shrinking lung syndrome (SLS) is a rare complication of systemic lupus erythematosus. It is characterized by dyspnoea and a restrictive ventilatory defect, often associated with pleuritic chest pain. Given its rarity, there is no standardized treatment. Most published case reports describe a favourable outcome with corticosteroids alone or in combination with other immunosuppressive agents, such as cyclophosphamide or rituximab. Only a few cases treated with belimumab have been reported.

Case description: We present the case of a 16-year-old female presenting with pleuritic chest pain, orthopnoea, and a restrictive ventilatory defect. Further investigations revealed positive antinuclear antibodies, a small left pleural effusion, and mild elevation of the left hemidiaphragm, findings consistent with SLS. Treatment with corticosteroids and belimumab was initiated, leading to significant clinical and biological improvement.

Conclusion: This case highlights the potential role of belimumab as a first-line treatment for SLS in combination with corticosteroids alone.

KEYWORDS

Shrinking lung syndrome, belimumab

LEARNING POINTS

- Shrinking lung syndrome is a rare condition with no standardized treatment.
- Most reported cases describe the use of corticosteroids in combination with azathioprine, mycophenolate mofetil or rituximab. Only a few cases have reported the use of belimumab.
- Belimumab may be considered as a first-line treatment option which could induce remission.

EFIM
European Federation of Internal Medicine

Publisher
smc media srl

INTRODUCTION

Shrinking lung syndrome (SLS) is a rare complication of some autoimmune diseases, most notably systemic lupus erythematosus (SLE). It is characterized by dyspnoea and a restrictive pulmonary syndrome that is often associated with pleural pain[1]. Given its rarity (<1% of SLE patients)[1], there are no validated criteria for establishing the diagnosis of SLS and no standardized treatment. Most case reports have shown a good prognosis with corticosteroids alone or in combination with other immunosuppressive drugs (such as cyclophosphamide or rituximab)[1].

Belimumab is an IgG1 λ monoclonal antibody that neutralizes B lymphocyte activating factor (BAFF) and is frequently used in cutaneous, articular or renal manifestations of SLE[2].

We report a case of SLS treated with belimumab, with resolution of symptoms and normalization of respiratory function.

CASE DESCRIPTION

A 16-year-old female presented to the pulmonology consultation with pleuritic thoracic pain, orthopnoea and dyspnoea on slightest exertion for one month. She had no fever. Her medical and family history was unremarkable, and she did not take any medication.

Her vitals were normal, cardiopulmonary auscultation was unremarkable. No arthralgia, no rash or no enlarged lymph nodes were identified.

Laboratory tests were ordered. Cardiac markers showed no elevation of troponins (<5 ng/l) but an increase of B-type natriuretic peptide (BNP) at 432 ng/l (<300 ng/l). A small inflammatory syndrome was noted, with C reactive protein (CRP) at 11.2 mg/dl (<5 mg/dl), lymphopenia at 0.8 g/l, a hypergammaglobulinemia at 27 g/l and a raised interferon signature at 12. Anti-nuclear antibodies (ANA) were positive at a titer greater than 1/1280, anti-double stranded deoxyribonucleic acid (dsDNA) at 1079.1 IU/ml (<27 IU/ml) and anti-nucleosomes at 193.4 U/mL (<18 U/ml). Complement was not activated. Viral serologies were negative. Proteinuria and haematuria were absent.

Chest X-ray showed a small left pleural effusion with a small elevation of the left hemidiaphragm *(Fig. 1)*. Pulmonary function testing (PFT) showed a restrictive lung disease *(Table 1 and Fig. 3)*. Moreover, there was a 19% drop in forced vital capacity (FVC) when moving from sitting to supine position, which indicated diaphragmatic weakness. An electromyogram, sniff and carbon monoxide diffusion tests were within normal range

Cardiac ultrasound revealed a small pericardial effusion with no other abnormalities.

The work-up was completed by low-dose chest computed tomography (CT) scan *(Fig. 2)* which, apart from left basal parenchymal condensations, showed no parenchymal involvement. An arterial blood gas test was unremarkable. A 6-minute walk test revealed a saturation at 90%.

Viral pericarditis was suspected and empirical treatment

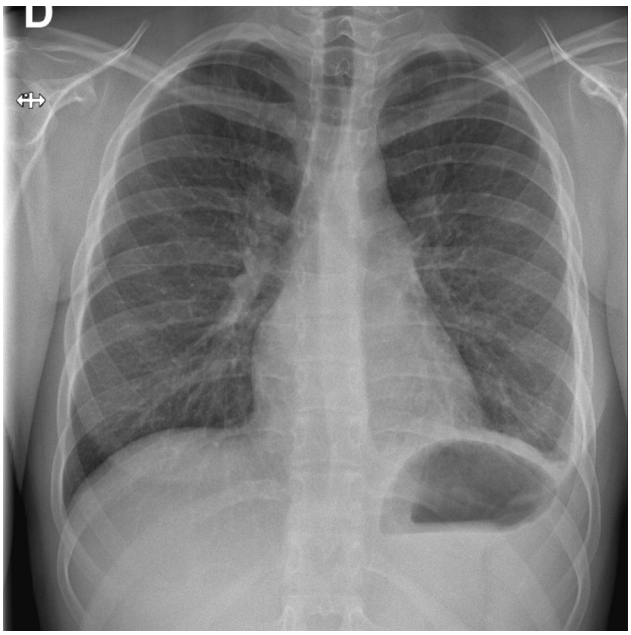


Figure 1. *Chest X-ray showing a left pleural effusion.*

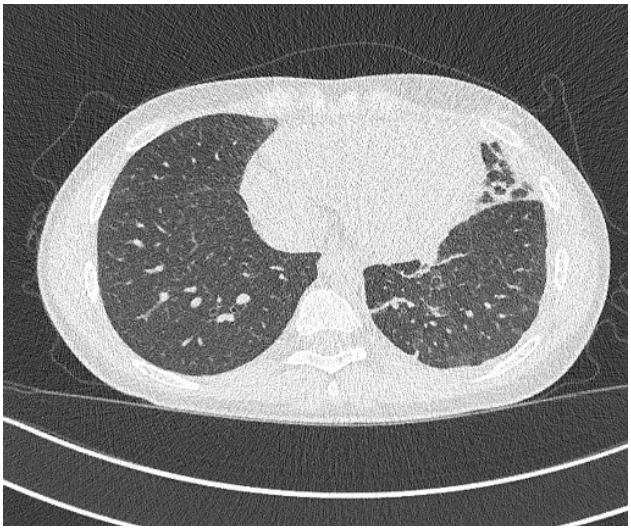


Figure 2. *Computed tomography scan of the thorax with pleural condensations.*

with colchicine (0.5 mg twice a day) and ibuprofen (400 mg three times a day) was started.

One month later, symptoms were persistent despite ibuprofen and colchicine. SLE was diagnosed (12 points according to the European League Against Rheumatism (EULAR)/ American College of Rheumatology (ACR) 2019 classification criteria) with probable SLS. Treatment with hydroxychloroquine (200 mg 2x/d) was added.

Three months later, there was no improvement. A new PFT showed a stable restrictive syndrome *(Table 1 ad Fig. 3)* and echocardiography showed persistent pericardial effusion. It was decided to initiate maintenance treatment with belimumab 600 mg (10 mg/kg) intravenously (IV) per month (with a 125 mg bolus of methylprednisolone during the first two months) and a rapidly decreasing regimen of oral methylprednisolone (started with 25 mg/day, with a steady dose of 7.5 mg/day after 2 months). After 2 administrations of belimumab, there was a normalization of

	FEV1		FVC		TLC		FEV1/FVC ratio (%)	
	Actual (l)	%Predicted	Actual (l)	%Predicted	Actual (l)	%Predicted	Actual	Predicted
First consultation	2.1	61	2.31	59	3.46	70	91.01	102
Control 1 month after	2.32	67	2.6	67	3.73	76	89.27	100
Colchicin and HCQ since 3 months	2.21	64	2.59	66	3.51	71	85.28	96
6 months after starting belimumab	2.97	86	3.37	86	4.47	90	88.05	99

Table 1. *Evolution of lung capacity over time.*
Abbreviations: FEV1, first second of forced expiration; FVC, forced vital capacity; TLC, total lung capacity, HCQ: hydroxychloroquine.

BNP (85 ng/l). Cardiac magnetic resonance imaging (MRI) showed enhanced gadolinium uptake in the pericardium and myocardium without pleural effusion.

Six months after starting belimumab, the symptoms disappeared completely and SLE biological activity decreased (anti-dsDNA to 144 IU/ml). A follow-up cardiac ultrasound showed no more pericardial effusion and BNP were normalized at 85 ng/l). This permitted a switch from IV to subcutaneous belimumab (200mg 1x/week) and a further reducing and eventually stopping methylprednisolone after 6 months. The patient underwent further PFT showing normalization of respiratory function *(Table 1 and Fig. 3)*. A new cardiac MRI one year after initiating belimumab showed no more enhanced uptake of the myocardium.

DISCUSSION

We diagnosed lupus based on the presence of ANA at a titer >1:80, anti-dsDNA and antinucleosome antibodies and pericardial effusions (12 points according to the 2019 EULAR/ACR criteria)[3].

SLS is a diagnosis of exclusion, there are no validated criteria. The diagnosis of SLS was made based on clinical features (progressive dyspnoea and pleuritic pain), diaphragmatic weakness, elevation of one hemidiaphragm, restrictive syndrome and the absence of parenchymal pathology (ruled out by chest CT scan)[4].

Usually, treatment for SLS is based on corticosteroids, non-invasive ventilation (NIV)[5] and respiratory rehabilitation[6]. Pulmonary sequelae may persist in up to 80% of patients[7].

Prednisone alone, from 20 mg to 1 mg/kg, sometimes preceded by boluses of methylprednisolone, is the first-line immunosuppressive treatment[8]. It may be combined with immunosuppressive agents such as cyclophosphamide, azathioprine, methotrexate and mycophenolate mofetil[1]. There was no prognostic difference between patients receiving prednisone alone and those receiving prednisone and another immunosuppressant. It should be noted that usually, when corticosteroids are ineffective in SLS, the same is true for cyclophosphamide, methotrexate and azathioprine[8].

Rituximab is also increasingly used as a treatment for SLS, in combination with corticosteroids in cases where the patient is refractory to corticosteroids alone[8].

Aerosols are also used: Theophylline has been described to increase diaphragm power, while beta-agonists can reduce diaphragm fatigability[1].

To our knowledge, only four case reports have described the use of belimumab in the treatment of

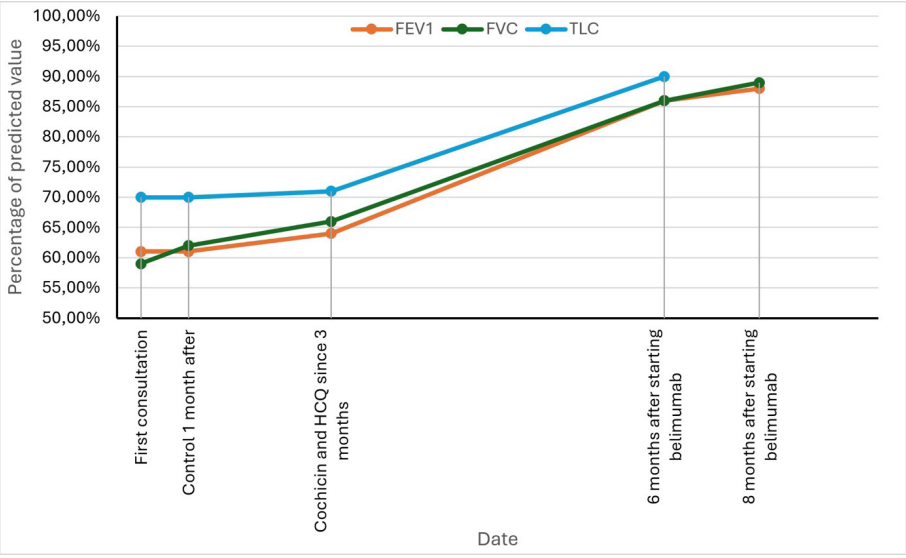


Figure 3. *Evolution of lung capacity (in percentage of predicted value) over time.*
Abbreviations: FEV1, first second of forced expiration; FVC, forced vital capacity; TLC, total lung capacity, HCQ: hydroxychloroquine.

SLS[9-12]. In all reported cases, corticosteroids and other immunosuppressive agents were used as induction therapy, while belimumab was administered only as rescue or maintenance therapy.

In our case, monotherapy belimumab, combined with low- to moderate dose corticosteroids (0,5 mg/kg), led to rapid clinical and functional improvement, with normalization of respiratory function tests, laboratory biomarkers, cardiac MRI and symptoms. To our knowledge, this is the first case of SLS remission with corticosteroids and belimumab alone. Further studies are needed to prove the efficacy of belimumab monotherapy in SLS.

REFERENCES

1. Shin JI, Lee KH, Park S, Yang JW, Kim HJ, Song K, et al. Systemic Lupus Erythematosus and Lung Involvement: A Comprehensive Review. J Clin Med. 2022;11(22):6714. doi: 10.3390/jcm11226714.
2. Furie R, Rovin BH, Houssiau F, Malvar A, Teng YKO, Contreras G, et al. Two-Year, Randomized, Controlled Trial of Belimumab in Lupus Nephritis. N Engl J Med. 2020;383(12):1117-1128. doi: 10.1056/NEJMoa2001180.
3. Aringer M, Costenbader K, Daikh D, Brinks R, Mosca M, Ramsey-Goldman R, et al. 2019 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Systemic Lupus Erythematosus. Arthritis Rheumatol. 2019;71(9):1400-1412. doi: 10.1002/art.40930.
4. Carmier D, Marchand-Adam S, Diot P, Diot E. Atteinte respiratoire au cours du lupus érythémateux systémique [Respiratory involvement in systemic lupus erythematosus]. Rev Mal Respir. 2008;25(10):1289-303. French. doi: 10.1016/s0761-8425(08)75093-9.
5. Panchabhai TS, Bandyopadhyay D, Highland KB, Chaisson NF, Aboussouan LS. A 26-Year-Old Woman With Systemic Lupus Erythematosus Presenting With Orthopnea and Restrictive Lung Impairment. Chest. 2016;149(1):e29-33. doi: 10.1016/j.chest.2015.10.053.
6. Sari F, Oskay D, Tufan A. The effect of respiratory muscle training on respiratory muscle strength, diaphragm thickness/mobility, and exercise capacity in patients with systemic lupus erythematosus and associated shrinking lung syndrome. Lupus. 2024;33(3):289-292. doi: 10.1177/09612033241226755.
7. Borrell H, Narváez J, Alegre JJ, Castellví I, Mitjavila F, Aparicio M, et al. Shrinking lung syndrome in systemic lupus erythematosus: A case series and review of the literature. Medicine (Baltimore). 2016;95(33):e4626. doi: 10.1097/MD.0000000000004626.
8. Duron L, Cohen-Aubart F, Diot E, Borie R, Abad S, Richez C, et al. 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. Autoimmun Rev. 2016;15(10):994-1000. doi: 10.1016/j.autrev.2016.07.021.
9. Reyes F, Lazaro D. Systemic lupus erythematosus complicated by shrinking lung syndrome treated with belimumab: a case report. Chest. 2016;150(4 Suppl):300A. doi: 10.1016/j.chest.2016.08.313.
10. Esteves AC, Mendes B, Assunção H, Inês LS. Combination treatment with rituximab and belimumab in shrinking lung syndrome in systemic lupus erythematosus. Int J Rheum Dis. 2024;27(3):e15110. doi: 10.1111/1756-185X.15110.
11. Choudhury S, Ramos M, Anjum H, Ali M, Surani S. Shrinking Lung Syndrome: A Rare Manifestation of Systemic Lupus Erythematosus. Cureus. 2020;12(5):e8216. doi: 10.7759/cureus.8216.
12. DeCoste C, Mateos-Corral D, Lang B. Shrinking lung syndrome treated with rituximab in pediatric systemic lupus erythematosus: a case report and review of the literature. Pediatr Rheumatol Online J. 2021;19(1):7. doi: 10.1186/s12969-020-00491-0.