Case Report

Rituximab in the Treatment of Shrinking Lung Syndrome in Systemic Lupus Erythematosus

Patricia Peñacoba Toribio, a,⁎ María Emilia Córica Albani, b Mercedes Mayos Pérez, c Arturo Rodríguez de la Serna b

a Servicio de Neumología, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain
b Unidad de Reumatología, Servicio de Medicina Interna, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain
c Unidad de Trastornos Respiratorios del Sueño, Servicio de Neumología, Hospital de la Santa Creu i Sant Pau, Centro de Investigación Biomédica en Red en Enfermedades Respiratorias (Ciberehs), Universidad Autónoma de Barcelona, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain

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A B S T R A C T
Shrinking lung syndrome (SLS) is a rare manifestation of systemic lupus erythematosus. We report the case of a patient with non-responding SLS (neither to glucocorticoids nor immunosupressors), who showed remarkable improvement after the onset of treatment with rituximab. Although there is a little evidence, treatment with rituximab could be proposed in SLS when classical treatment fails.

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Rituximab en el tratamiento del síndrome del pulmón encogido del lupus eritematoso sistémico

R E S U M E N
El síndrome del pulmón encogido (SPE) es una manifestación poco frecuente del lupus eritematoso sistémico. Exponemos el caso de un paciente afectada SPE, refractario al tratamiento con glucocorticoides e inmunosupresores, que tras el inicio de tratamiento con rituximab presentó marcada mejora de los síntomas. Aunque la evidencia es escasa, el tratamiento con rituximab podría ser propuesto en el SPE cuando fracasa el tratamiento clásico.

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Introduction

Shrunken lung syndrome (SLS) is a rare manifestation of some autoimmune diseases such as systemic lupus erythematosus (SLE). It is characterized by a restrictive type ventilatory defect and should be suspected in patients with SLE presenting unexplained dyspnea. Treatment is not well established. Although the use of systemic corticosteroids as first-line treatment has been proposed in refractory cases, other immunosuppressive drugs, such as rituximab, have been suggested.

Clinical Case

The patient was a 57-year-old female followed by the Rheumatology Department since age 22, diagnosed with SLE in the context of fever, polyarticular pain, malar rash, and photosensitivity. She presented antinuclear antibodies 1/320 (normal <1/40), anti-native DNA 50.00 IU/ml (normal <15), IgG anticardiolipin 20.23 GPL/ml (normal 0.00–7.70), and positive anti-histone antibodies. She was initially treated with prednisone (15 g/day) and chloroquine (155 mg/day) with good response.

At age 29, due to an episode of focal segmental glomerulonephritis, treatment with azathioprine was added 50 mg/12 h, with complete remission.


⁎⁎ Corresponding author.

E-mail address: ppenacoba@santpau.cat (P. Peñacoba Toribio).

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The presenting symptoms are dyspnea and chest pain. Auscultation is usually normal.\textsuperscript{5,6}

Respiratory function shows a restrictive type of ventilatory defect with reduced lung volumes. Imaging tests show no evidence of pleural, parenchymal or vascular disease.\textsuperscript{1,2}

The cause of the SLS is not known.\textsuperscript{1–3} There are several hypotheses including the presence of atelectasis by the loss of surfactant,\textsuperscript{3,4} diaphragmatic myopathy, phrenic neuropathy, pleural adhesions, and inactivation of the diaphragm induced by pain.\textsuperscript{3} SLS has also been linked to anti-Ro antibody.\textsuperscript{2,3,7} None of these theories has been confirmed.

The long-term prognosis is generally good, with clinical stabilization or improvement.\textsuperscript{6} Deterioration appears in only 2\% of cases, requiring aggressive therapy\textsuperscript{8,9} or even mechanical ventilation.

There is no definitive treatment of SLS, although glucocorticoids are considered the first line therapy, alone or in combination with other immunosuppressive agents.\textsuperscript{1} In addition, theophylline and beta-2 agonists are used with the intention to increase diaphragmatic strength.\textsuperscript{1,3}

On the other hand, there has been much improvement both of pleuritic pain and of dyspnea in patients treated with rituximab. Rituximab is a chimeric mouse/human monoclonal antibody that specifically binds to the CD20 antigen expressed on pre-B and mature B lymphocytes. Although no prospective study of rituximab in SLS has been performed, isolated cases have been reported where refractory patients experience symptom improvement with increased exercise tolerance, and an improvement in respiratory function tests with increasing expiratory volume in the first second and forced vital capacity.\textsuperscript{3,10}

Conclusion

So far, the treatment of choice for SLS is corticosteroids alone or in combination. Refractory cases may benefit from treatment with rituximab.

Ethical Responsibilities

Protection of persons and animals. The authors state that no experiments were performed on persons or animals for this study.

Data confidentiality. The authors state that they have followed their workplace protocols regarding the publication of patient data and all patients included in the study have received enough information and have given their written informed consent to participate in the study.

Right to privacy and informed consent. The authors state that they have obtained informed consent from patients and/or subjects referred to in this article. This document is in the possession of the corresponding author.

Conflict of Interest

The authors declare no conflicts of interest.

References