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# AN UNUSUAL CASE OF REFRACTORY SYSTEMIC LUPUS ERYTHEMATOSUS/MIXED CONNECTIVE TISSUE DISEASE WITH CHYLOUS EFFUSION SUCCESSFULLY TREATED WITH SIROLIMUS

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**Objective:**

Sirolimus is an immunosuppressant used in patients with transplanted solid organs and lymphangioleiomyomatosis (LAM). Lately, sirolimus has also demonstrated the ability to reduce global disease activity of systemic lupus erythematosus (SLE) patients. Here, we present a patient diagnosed with refractory SLE/mixed connective tissue disease (MCTD) and chylous pleural effusion successfully treated with sirolimus.



The patient is a 33 year-old female who first presented in 2011 with swelling of small hands joints and knees, lower back pain, morning stiffness, Raynaud phenomenon and butterfly rash. After initial laboratory work-up, she was diagnosed with Sjogren syndrome and treated with prednisone and resochin. Her condition deteriorated in 2015 with appearance of bilateral chylous pleural effusions and ascites. The pathohistological finding of the lung and pleura biopsy (also revised in two international medical centers) ruled out LAM and changes were attributed to an underlying undetermined connective tissue disease now suspected to be SLE or MCTD. Described pulmonary arteries plexiform lesions were consistent with the clinical finding of newly developed moderate pulmonary arterial hypertension (PAH). During the next four years, she was treated with prednisone, resochine, and two pulmonary vasodilators - sildenafil and bosentan. While her cardiac function remained stable, ascites, hypoalbuminemia, legs oedema, pleural effusions and respiratory insufficiency worsened. Thus, in 2018, she received 11 cycles of low-dose cyclophosphamide pulses. Initially, her respiratory function stabilized, but then again worsened and cyclophosphamide was stopped. At the beginning of 2019, chest tube was placed and further worsened the production of chylous effusion and consequential severe hypoalbuminaemia. At that point, octerotide was introduced to therapy. This stabilized chylous production, increased albumin levels and enabled removal of the chest tube. Still, the patient required constant oxygen therapy (6L/min) at rest. In addition to monthly octerotide dose, further immunosuppression (mycophnolate mofetil) was introduced. The patient was stable but without significant improvement. Finally, in November 2019, sirolimus was added to the therapy regimen resulting in the reduction of pleural effusion and improvement of her functional class status. Today, after 9 months of therapy with sirolimus, mycophenolate mofetil and low doses of prednisone, she is respiratory sufficient at rest (and only requires 1L of oxygen on exertion), and her control CT scan showed significant improvement.

This off-label sirolimus application has resulted with significant reduction of disease activity and improved patient's quality of life. Still, more research is needed to explore sirolimus effects in patients with refractory SLE.

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