Rituximab is a chimeric monoclonal antibody against CD20 that depletes B-lymphocytes. There is increasing evidence for its use in Scleroderma ILD. Recently it has been reported as rescue therapy in patients with connective tissue disease related severe fibrotic lung disease who have failed conventional immunosuppression. It remains unclear which patients are most likely to benefit from this potent immunosuppressive treatment. We review here the experience of the Bristol Interstitial Lung Disease service in use of Rituximab in a subset of patients with myositis (Anti-synthetase syndrome and Dermatomyositis).

Methods We retrospectively reviewed the case notes of 10 patients with severe and progressive ILD despite immunosuppression with Cyclophosphamide and Mycophenolate Mofetil, who had received salvage treatment with Rituximab. Serial pulmonary function tests, 6 min walk distances and HRCT appearances (as assessed by a Thoracic radiologist) were compared in the year before and after Rixtuximab therapy. Changes in physiological variables compared to nadir at treatment were compared with paired-samples T-Test.

Results The average age of the patients was 49.8 (range 26.9–72.99), with 7/10 female. 4 patients had dermatomyositis, while 6 had Anti-Synthetase Syndrome (2 Anti-Jo1, 2 Anti-PL12, 1 Anti-PL7, 1 Anti-PM-Scl). There were complete lung function data available for 9 patients and 6MWD data for 6 patients.

CT appearances stabilised in all 9 patients with follow-up scans available, with significant improvement in 2 (1 after a second pulse of Rituximab).

FVC improved after treatment by an average of 9.2% (p = 0.023, 95% CI 1.67–17.67), with TLCO improving by an average of 6.1% (NS). Figure shows% change in FVC and TLCO leading to and after therapy, 6MWD remained stable.

There were no adverse events reported.

Summary Our experience adds to the growing evidence to support the use of Rituximab in severe CTD-ILD, and suggests that a subset of patients with myositis may show good therapeutic response.

REFERENCES


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