SUCCESSFUL TREATMENT WITH PLASMAPHERESIS AND ANTI-CD20 THERAPY IN A PATIENT WITH MIXED CONNECTIVE TISSUE DISEASE

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Background: Mixed Connective Tissue Disease (MCTD) is characterized by a high titer of anti-U1-RNP antibodies and clinical features of systemic sclerosis, systemic lupus erythematosus and dermatomyositis.

Methods: We report the case of a 30 years old MCTD-affected woman, with Raynaud’s phenomenon, alopecia, myositis, arthralgia, ANA, anti-U1RNP and anti-Sm-RNP positivity, lymphopenia and hypergammaglobulinemia. Between 1997 and 2008 the patient was treated with steroids, cyclosporine A, hydroxychloroquine, and nifedipine. In 2008 cyclosporine A was suspended after an increase of blood pressure. Subsequently, a facial eruption appeared. A punch biopsy revealed changes characteristic of subacute cutaneous lupus erythematosus. Unfortunately, she failed to achieve a satisfactory response with hydroxychloroquine, methotrexate and azathioprine treatment. Mycophenolate mofetil was able to induce a skin improvement but it was stopped due to adverse reaction. Therefore, therapeutic plasma exchange (TPE) for four days every 4 weeks was employed for three cycles and the CD20 antibody rituximab (375 mg/m² weekly for 4 weeks) was added one month after the last TPE. Eight months later the patient received another cycle of TPE and rituximab.

Results: Following 3 TPE, there was a slight clinical improvement and the addition of rituximab has allowed to obtain a further clearing of facial eruption, while a complete skin healing was reached 8 weeks after the last rituximab infusion. Moreover, mitigation of Raynaud’s phenomenon, resolution of alopecia and increase of CD4+ T lymphocytes was observed.

Conclusions: The combination of TPE and rituximab should be considered as a valid therapeutic option for controlling disease in therapy resistant MCTD.