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TREATMENT OF REFRACTORY DERMATOMYOSITIS WITH TOFACITINIB

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Background: Dermatomyositis (DM) is a systemic, autoimmune disease affecting the skin and proximal skeletal muscles. A subset of DM patients present with subclinical or resolved muscle involvement but continue to have skin disease. In these cases, first and second line treatments including glucocorticoids are sometimes insufficient for controlling the disease, necessitating escalation of treatment. Several recent studies have investigated the response of Tofacitinib, an oral Janus Kinase inhibitor approved for the treatment of rheumatoid arthritis, in DM patients and patients with inflammatory skin diseases.

Objectives: Due to the reported ability of JAK inhibitors to suppress type 1 interferon (IFN) signaling, which is suspected to be upregulated in DM, we evaluated the efficacy of treatment with Tofacitinib in four refractory DM patients.

Methods: Four patients with dermatomyositis without evidence of current muscle involvement began treatment with Tofacitinib 11 mg daily after they had failed or had adverse effects to first and second line immunosuppressive agents. Their medical records were reviewed at 0, 3, and 6 months, with improvement measured using the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) activity score. Throughout their treatment they were additionally monitored for improvement in markers of inflammation and the necessity for concomitant treatments. Patients were monitored for adverse effects to Tofacitinib treatment.

Results: All four patients within the case series showed significant improvement of their cutaneous disease activity (CDASI scores improved by 8-15 points) over the first 6 months, with three of the four having achieved minimal clinically improved difference of ≥ 5 point by three months. Based on the CDASI, three of the cases' disease classification changed from moderate-to-severe disease to mild disease. The last patient initially presented with mild disease. Other outcomes noted included improved pruritus in 3 patients and improvement of calcinosis in 1 patient. One patient was additionally able to stop concomitant treatment with prednisone and IVIG at 3 and 6 months, respectively. This patient had been on daily prednisone for 4 years. Only other patient on prednisone was on low dose of 3mg daily. The patients all had normal muscle exams prior to treatment with Tofacitinib. No worsening muscle involvement or adverse effects were noted with Tofacitinib use.

Conclusion: Tofacitinib is believed to play a role in the inhibition of IFN signaling pathways that are overactive in dermatomyositis. All four patients

Gender/ Age/ Ethnicity	DM Subtype / disease duration	Previous Treatment	Concomitant Treatment with TOF	CDASI Activity	CDASI Activity 3 months	CDASI Activity 6 months	Other Outcome
M/ 55/ Caucasian	Amyopathic DM; 5 years	AZA, HCQ, IVIG, MTX, MMF, Prednisone, RCI, RTX, TAC	HCQ	12	5	4	None
F/ 37/ Caucasian	Amyopathic DM; 7 years	AZA, HCQ, IVIG, MTX, MMF, Prednisone, RCI, TAC	Prednisone, IVIG, MTX	15	8	5	Pruritus and CRP improved
F/ 60/ Caucasian	Amyopathic DM; 4 years	AZA, MTX, Prednisone	Prednisone, MTX	17	15	5	Pruritus improved
F/ 47 African American	Classic DM; 5 years	AZA, colchicine, HCQ, IVIG, MTX, MMF, pamidro- nate Prednisone, RCI, RTX, TAC	IVIG, Colchicine	22	8	7	Pruritus and calcinosis improved

Abbreviations: CDASI, Cutaneous Dermatomyositis Disease Area and Severity Index; AZA, azathioprine; CRP, C-reactive protein; DM, Dermatomyositis; HCQ, hydroxychloroquine; IVIG, intravenous immunoglobulin; MTX, methotrexate; MMF, mycophenolate mofetil; RCI, repository-corticotropin injection; RTX, rituximab; TAC, tacrolimus; TOF, tofacitinib;

within this retrospective study showed significant improvement of cutaneous disease with Tofacitinib use.

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THE EFFECTS OF HYPERBARIC OXYGEN THERAPY TO QUALITY OF LIFE AND STATE OF MICROCIRCULATION IN PATIENTS WITH SYSTEMIC SCLEROSIS - A PILOT STUDY

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Background: Many treatments have been tried in therapy systemic sclerosis (SSc) patients but use of hyperbaric oxygen therapy (HBOT) is very limited.

Objectives: To assess the effects of HBOT to quality of life and state of microcirculation in SSc patients.

Methods: 18 female patients aged 29-68 years (mean 57 years) with limited SSc and digital or leg ulcers were included in this work. The HBOT protocol comprised 20 sessions 5 day/weekly, 60 min, 100% oxygen at 2.2 ATA. The treated patients were evaluated at baseline and after 20 HBOT sessions. Evaluation consisted of physical examination, capillaroscopy, pulmonary function tests, biochemical analyses, socio-demographic and clinimetric questionnaires: Systemic Sclerosis Questionnaire (SySQ) and Health Assessment Disability index Questionnaire (HAQ-DI).

Results: Mean value [before: after, mean (range)] for SySQ [15.5 (4-48) vs 9.0 (3-31)], HAQ-DI [0.60 (0-2.88) vs 0.35 (0 -1.75)], erythrocyte sedimentation rate [21 (4-42) vs 12 (3-27)], forced vital capacity (96.61±14.44% vs 115.94±16.69%), diffusing lung capacity of carbon monoxide (73.61±6.63% vs 87.33±9.30%) significantly improved after HBOT sessions (p<0.001). There was no significant changes in the total number of capillaries (325 vs 338, p=0.235), mean number of enlarged capillaries (21 vs 27, p=0.182), giant capillaries (14 vs 14, p=0.235) and ramified/bushy capillaries (14 vs 13, p=0.178) before and after HBOT. All patients had digital ulcers, and 5 patients had bilateral lesions (digital and leg ulcers). Mean size of ulceration before HBOT was 12x11mm, and after therapy was 4x4mm (p<0.001). Three patients had digital gangrene. Amputation was not require for any.

Conclusion: Our data confirm the efficacy of HBOT in treating SSc patients. Further studies are required to evaluate the protocol and to understand the duration of the clinical effect.

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