Background: Idiopathic inflammatory myopathies are rare autoimmune diseases that affect between two and ten people per million.

Objectives: The aim of our work is to describe the epidemiological, clinical and paraclinical characteristics of autoimmune inflammatory myopathies as well as the therapeutic modalities.

Methods: This is a retrospective study involving 50 inflammatory myopathy patients admitted to Ibn Sina Hospital’s internal medicine department between January 2015 and June 2022.

Results: There were 50 patients, 37 women and 13 men (gender ratio M/F: 0.35). The mean age of onset of symptoms was 45.8 years old, the mean age of diagnosis was 47.6 years old (extremes: 23 and 80 years old) and the mean time between onset of symptoms and positive diagnosis was 14.9 months. The main reason for hospitalization was muscle impairment in 98% of our patients. It was dominated by muscle weakness with a mean score of 3/5 and myalgia. Proximal involvement was predominant. No facial musculature involvement was reported. At the time of diagnosis, dysphagia was present in 42% of cases, and no dysphonia was reported. Cutaneous signs were observed in 78% of cases with erythromelalgia type 22%, Gottron's papules 20% and maculopapule sign 16%. General signs were found in 56% of cases predominantly asthenia and weight loss, arthralgia was present in 34.7% of cases. 20% of patients had heart disease, myocardiitis type, pericarditis, and arrhythmia in 2 cases each, neurological disease was found in 16% of cases (pyramidal syndrome type in 5 (10%) cases and neurogenic disease in 3 (6%) cases). PID-type pulmonary disease was present in 24% of cases, with complications of pulmonary fibrosis in 4% of cases. Creatine phosphokinase was elevated in 76% of cases with a mean value of 2553 IU/L and extremes between 26 and 25000 IU/L. Fifteen patients at the time of diagnosis had normal CPK contrasting with clinical signs. LDH was increased in 73%, liver cytolysis in 73%, and biologic inflammatory syndrome in 51%. Electromyograms were performed in 80% of cases, showing a myogenic pattern in 82.5% of cases, associated with neurogenic involvement in 25% of cases. Muscle biopsy (performed in 75% of cases) confirmed the diagnosis in 27 patients (77.1%) and in 8 patients the biopsy was normal. Antinuclear antibodies were positive in 70% of the cases, anti-JO1 positive in 26% of the cases, 26.7% of the patients were anti-SSA positive, and 10% of the cases were anti-Mi2 positive. The diagnosis was dermatomyositis in 36% of cases, anti-synthetase syndrome in 20% of cases and cancer-associated myositis in 10% (5 patients), and overlapping myositis in 28% of cases. No cases of necrotizing myopathies or inclusion myopathies have been reported. The types of neoplasia revealed by idiopathic inflammatory myopathies in our study were: poorly differentiated carcinoma in 3 cases on lymph node biopsies, poorly differentiated adenocarcinoma of metastatic lung in 1 case and one case had Hodgkin's lymphoma. The treatment was based on corticosteroid therapy for all cases. Corticosteroid therapy was associated with an immunosuppressant in 58% of cases (methotrexate in 5 patients), cyclophosphamide in 20% of patients, azathioprine in 4 patients and 2 patients on MMF). In 32% of patients, IV immunoglobulin therapy was prescribed, and in 8%, Rituximab was prescribed. We found no deaths in our series, with 38% of cases going into complete remission.

Conclusion: The classification criteria for idiopathic inflammatory myopathies have been constantly changing. The diagnosis of inflammatory myopathy can currently be made in common practice when a patient's clinical, muscular histology, and serology support the diagnosis. The elimination of an associated cancer remains a challenge for the practitioner.

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AB0917

A RANDOMIZED CONTROLLED PROSPECTIVE SINGLE-CENTER FEASIBILITY STUDY OF RHEOPHERESIS FOR RAYNAUD'S SYNDROME AND DIGITAL ULCERS IN SYSTEMIC SCLEROSIS (RHEACT) - PRESENTATION OF FIRST INTERIM RESULTS

Keywords: Randomized control trial, Systemic sclerosis

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Background: Raynaud's phenomenon (RP) and digital ulcers (DU) are frequent manifestations of Systemic Sclerosis (SSc). There are very few available approved drugs with varying efficacy. Rheopheresis (RheoP) is an extracorporeal apheresis technique used to treat microcirculatory disorders by improving blood viscosity. The effect of RheoP in SSc is currently unknown, and here, we report the interim results of the first patients.

Objectives: To determine the efficacy of RheoP compared to Standard of Care (SoC) intravenous iloprost on RCS and DU healing.

Methods: A randomized controlled prospective single-center feasibility study of Rheopheresis for Raynaud's syndrome and Digital Ulcers in Systemic Sclerosis (RHEACT) aims to investigate the efficacy of RheoP on the Raynaud Condition Score (RCS) as the primary outcome measure after 16 weeks from baseline. A planned number of 30 patients will be randomized in a 1:1:1 ratio to one of two RheoP treatment groups or assigned to the SoC control group.

Results: We here report the results of the first seven patients. The patient assigned to the RheoP1 group had a baseline Raynaud Condition Score (RCS) of 8, which improved to 0 after 16 weeks (the primary endpoint). The two patients randomized to the RheoP2 had a baseline RCS of 8 and 6, respectively, which improved to 4 and stayed unchanged at 8 in the second patient. The four patients assigned to SoC had only minimal improvement. RheoP1 had a significantly lower RCS compared to RheoP2 and SoC at week 16 (**p<0.05). Furthermore, DU completely healed in patients with DU at baseline with either RheoP therapy and avoided amputation in one patient. Exemplary images are shown in figure 2 (A, baseline; B, at week 16).

Conclusion: RheoP improved the RCS and DU better than SoC (iloprost) treatment alone and avoided amputation in one patient.

Figure 1.

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AB0918

CLINICAL AND SEROLOGICAL FEATURES OF DERMATOMYOSITIS IN TUNISIA

Keywords: Myositis, Autoantibodies, Motor function