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DESCRIPTIVE ANALYSIS AND STUDY OF CORRELATIONS IN A COHORT OF PATIENTS WITH INFLAMMATORY MYOPATHIES

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Background: Inflammatory myopathies (IM) are a group of rare diseases that involve muscle inflammation. Several types are defined with a wide range of different manifestations and prognosis.

Objectives: Describe the characteristics of the cohort of patients with IM in a tertiary hospital, in order to identify their demographic and clinical characteristics and try to find a correlation between them.

Methods: Retrospective observational study of patients with IM: dermatomyositis (DM), polymyositis, antisynthetase syndrome (ASS), necrotizing autoimmune myopathy and overlap syndrome (OS). Clinical, biological, neurophysio and histopathological data were collected. Statistical analysis was performed using SPSS 23 IBM®.

Results: 28 patients were included with a follow-up of 10.9 ± 9.8 years (y). According to the 2017 EULAR / ACR criteria for IM, 89.2% were classified as definitive MI, with an average score of 12.1 ± 3.2 . Age at diagnosis 47.3 ± 17.7 y; ratio 1.3; 78.6% Caucasian, 10.7% Asian and 10.7% Latino. 39.3% had DM, 3.6% hypomyopathic and 3.6% amyopathic DM, 28.6% OS and 17.9% ASS. Lung involvement was the most prevalent extramuscular manifestation (67.9%). Systemic sclerosis was the most frequent overlapping autoimmune disease (AD) (21.4%) and 2 patients (7.1%) overlapped more than 1 AD. In Table 1 are detailed the clinical characteristics of the patients, and in Figures 1 and 2, the autoantibody (aa) profile and treatments used. The incidence of neoplasm was 10.7% 10.3 ± 9.6 y after the diagnosis of myopathy (3 breast neoplasms, 1 colon and 1 cutaneous lymphoma), and of them, 66.7% had two synchronous neoplasms. No neoplasms were observed in the 2 anti-TIF1-γ+ patients. The subtype of IM in these patients was 1 OS anti-RNP+, 1 DM anti-PL-12+ and 1 ASS anti-OJ+. 17.9% smoked and 21.4% had taken statins at some point, without it being related to the start of the myopathic clinic. A capillaroscopy was performed in 67.9%, being pathological in 63.1%.

The positivity of anti-RNP ($p=0.01$) and steroid bolus ($p=0.039$) were correlated with a more severe disease, defined as a summation index composed of a series of manifestations (pulmonary hypertension (PH), ischemic heart disease, venous/arterial thrombosis, myo/pericarditis, interstitial lung

Table 1. Clinical characteristics.

Clinical manifestations	Frequency %
Presentation	
Acute	40.7
Subacute	22.2
Insidious	37
Muscular Weakness	82.1
Muscle Enzymes Elevation	85.7
Muscle Pain	67.9
Joint Manifestations (Mf)	67.9
Systemic Mf	67.9
Digestive Mf	46.4
Raynaud's Syndrome (RS)	53.6
Sclerodactyly	32
Digital ulcers (DU)	25
Calcinosis	10.7
ILD	67.9
Nonspecific Interstitial Pneumonia	63.2
Usual Interstitial Pneumonia	15.8
Organizing Pneumonia	10.5
Lymphocytic Interstitial Pneumonia	5.3
Undefined Pattern	5.3
PH	10.7
Serious Infections	17.9

Table 2. Correlations between clinical manifestations and aa.

Manifestations	Autoantibodies	p value
DU	Anti-MDA5	0.005
Sclerodactyly	Anti-RNP	0.011
Pericarditis	Anti-RNP	0.000
Myocarditis	Anti-RNP	0.005
Diabetes	Anti-RNP	0.027
RS	Anti-PM/Scl	0.033
Calcinosis	Anti-PM/Scl	0.027
Flexion Contractures	Anti-PM/Scl	0.027
Arrhythmias	Anti-PL-12	0.001

disease (ILD), severe infections, neoplasms or hospitalizations). Other statistically significant correlations between aa and clinical manifestations are detailed in Table 3, among which the anti-RNP+ with myopericarditis stands out. No correlation was found between the findings on capillaroscopy and the type of IM.

Conclusion: The most frequent subtype of IM was DM. 10.3% of the patients presented a neoplasm, all with different subtypes of myopathy and aa. The presence of anti-RNP+ correlated with greater severity of the disease and myopericarditis. Likewise, significant differences were found between the subtypes of aa and certain clinical manifestations. There is no correlation between findings on capillaroscopy and the type of IM.

Figure 1. Aa.

Figure 2. Treatment.

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AUTOMATIC QUANTIFICATION OF INTERSTITIAL LUNG DISEASE FROM CHEST COMPUTED TOMOGRAPHY IN SYSTEMIC SCLERODERMA

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