AB0730

COMPARISON OF A SINGLE-CENTRE IDIOPATHIC INFLAMMATORY MYOPATHY COHORT FROM ARGENTINA WITH THE EUROMYOSITIS INTERNATIONAL REGISTRY

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Background: The idiopathic inflammatory myopathies (IIM) are rare systemic autoimmune diseases that affect the muscle and other organs. Traditionally, IIM encompasses polymyositis (PM) and dermatomyositis (DM), but progressively inclusion body myositis (IBM), Immune-mediated necrotising myopathy (IMNM), the antisynthetase syndrome (ASS) and connective tissue diseases-over myositis (CTD-OM) have been recognised within the IIM spectrum

Objectives: To compare the clinical characteristics and treatment in a IIM cohort from an Argentinian university hospital with the international IIM cohort EUROMYOSITIS

Methods: Descriptive, retrospective study. IIM patients defined by expert opinion followed in our centre between October 2007 and October 2017 were included. ASS was defined by the presence of arthritis, raynaud's phenomenon, mechanic hands, elevated CK, muscle weakness, interstitial lung disease and/or presence of antisynthetase antibodies and, as in EUROMYOSITIS, patients with IIM with positive antisynthetase antibodies were reclassified as ASS. CTD-OM was defined as patients with IIM fulfilling classification criteria for other CTD.

Demographic data, accumulated clinical features, time interval between disease onset and diagnosis, IIM subtype, treatment and presence of neoplasm were evaluated.

Ethnicity was defined using the same classification as in EUROMYOSITIS

Results: 58 patients were included: DM 24, PM 4, ASS 10, CTD-OM 20. 89.6% Hispanic, mean age 48.4±15.2 years, median time interval between disease onset and diagnosis 5 months (IQR 2–11 months), been higher in AAS (8.5 months, IQR 1.5–18.2 months). 6.89% (4/58) patients presented associated neoplasm, 3 with DM and 1 with CTD-OM. Table 1 shows the demographic and clinical features of our IIM cohort and EUROMYOSITIS. Table 2 shows treatments received in our cohort and EUROMYOSITIS.

Abstract AB0730 – Table 1. Demographic and clinical features

	DM		PM		ASS		CTD-OM	
	HCJSM (n=24)	EUROMYO (n=949)	HCJSM (n=4)	EUROMYO (n=813)	HCJSM (n=10)	EUROMYO (n=512)	HCJSM (n=20)	EUROMYO (n=358)
Gender % M/F	20 (71)	30 (76)	75 (25)	29 (71)	40 (40)	31 (48)	25 (75)	21 (79)
Mean Age at Onset, years (SD)	46.73 (15.29)	49 (15)	43 (13.95)	40 (15)	40.5 (13.89)	48 (15)	40.9 (15.98)	48 (15)
Mean Age at Diagnosis, years (SD)	46.74 (15.36)	51 (15)	44.75 (15.39)	52 (15)	41.9 (15.34)	49 (15)	41.35 (15.83)	48 (15)
Median interval between onset and diagnosis, months (IQR)	4 (1.7-5)	5 (2-11)	5.5 (3.25-65.75)	8 (3-19)	6.5 (1.5-19.25)	7 (3-13)	5 (2.25-9.75)	11 (4-24)
Muscle weakness (%)	75 (16)	52	100	98	60	50	95	94
Rayn (%)	91.6	100	0	0	60	44	40	32
Raynaud's phenomenon (%)	33.3	25	50	29	50	51	65	60
Periungual erythema (%)	20.88	52	0	6	10	32	15	33
Actinias (%)	33.3	20	50	20	60	50	40	40
Mechanic's hands (%)	16.6	22	0	0	20	38	5	16
Calcinosis (%)	29.16	7	50	1	50	3	30	10
Ulceration (%)	8.33	13	0	2	10	3	5	10
Interstitial Lung Disease (%)	16.6	21	25	17	90	71	50	32
Cardiac involvement (%)	16.6	9	0	9	20	11	10	12

Conclusions: DM was the most frequent IIM subtype in both cohorts. In our group, CTD-OM was second and ASS was third.

Muscle weakness was found less frequently in our DM and AAS than reported in EUROMYOSITIS. However, calcinosis was more frequent. This could be explained by our mostly Hispanic population and/or by frequent Systemic Sclerosis overlap in our patients. It's important to remark that the ethnic variety defined as Hispanic in EUROMYOSITIS has a complex composition in Latin America, due to interbreeding.

No difference was found in terms of most frequent treatments between both cohorts. However, use of IVIg was more frequent in our patients.

To our knowledge, this is the first comparative report of an argentinian single-centre IIM cohort and an international multi-centre cohort

Disclosure of Interest: None declared

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Abstract AB0730 – Table 2. Treatment

	DM		PM		ASS		CTD-OM	
	HCJSM (n=24)	EUROMYO (n=949)	HCJSM (n=4)	EUROMYO (n=813)	HCJSM (n=10)	EUROMYO (n=512)	HCJSM (n=20)	EUROMYO (n=358)
Steroids	95	98	50	100	90	98	85	96
Hydroxychloroquine	50	37	25	11	40	16	40	29
Methotrexate	37.5	69	50	76	20	60	25	72
Leflunomide	0	0	0	3	20	3	5	4
Azathioprine	33.3	44	25	52	60	60	35	51
Mycophenolate	0	20	25	20	30	31	10	29
Cyclophosphamide	8.33	15	0	14	20	39	10	18
IVIg	29.16	11	0	13	20	7	5	7

AB0731

TREATMENT ALGORITHMS FOR SYSTEMIC SCLEROSIS ACCORDING TO EXPERTS

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Background: Treatment for many aspects of systemic sclerosis (SSc) lacks agreement.

Objectives: To generate SSc treatment algorithms endorsed by high percentage of SSc experts.

Methods: Experts from the Scleroderma Clinical Trials Consortium and the Canadian Scleroderma Research group (n=170) were asked whether they agreed with SSc algorithms (from 2012¹). A further 2 consensus rounds refined agreement; 62 (36%), 54 and 48 experts completed surveys.

Results: For scleroderma renal crisis (SRC), 82% of the experts agreed (1st line ACEi, 2nd and 3rd adding: CCB or ARB). Pulmonary arterial hypertension (PAH) had 81% agreement. For mild PAH, PDE5i, then endothelin receptor antagonists plus PDE5i, then prostanooids; while for severe PAH prostanooids were first-line. Raynaud's phenomenon (RP) had 78% of agreement [mild (1st CCB, 2nd adding PDE5i, 3rd ARB or switching to another CCB, 4th prostanooids), severe (1st CCB, 2nd adding PDE5i, 3rd ERA, 4th prostanooids)]. Digital ulcer (DU) treatment had 69% agreement (1st CCB, 2nd PDE5i). Interstitial lung disease (ILD) had 65% agreement including induction (Mycophenolate mofetil (MMF) then intravenous cyclophosphamide then rituximab) and maintenance (1st line MMF). Skin involvement had 71% agreement. For a modified Rodnan skin score (mRSS) of 24 1st MTX, 2nd MMF; and for mRSS 32 1st MMF, 2nd MTX, 3rd intravenous cyclophosphamide (CYP), 4th hematopoietic stem cell transplantation. For inflammatory arthritis 79% agreed with 1st MTX, 2nd low dose glucocorticoids, 3rd hydroxy-chloroquine, 4th rituximab or tocilizumab. Cardiac and gastrointestinal algorithms had ≥75% agreement. The evolution of the agreement rates is shown in table 1:

Abstract AB0731 – Table 1

Algorithms for SSc treatment	Agreement 2012 ¹ (%)	Agreement 2017 (%)
Scleroderma renal crisis	69	82
Pulmonary arterial hypertension	45	81
Raynaud's phenomenon	66	78
Digital ulcers	58	69
Interstitial lung disease	64	65
Gastrointestinal involvement	NA	77
Skin involvement	56, 40, 36 [†]	71
Inflammatory arthritis	45	79
Cardiac involvement	NA	75

Agreement is the% who agreed to the algorithm.

SSc: Systemic Sclerosis; NA: Not applicable; [†]For modified Rodnan skin scores 10, 24 and 32, respectively

Conclusions: Total agreement for SSc algorithms was considerable. These SSc algorithms may guide treatment.

REFERENCE:

- Walker KM, Pope J. Treatment of Systemic Sclerosis Complications: What to Use When First-Line Treatment Fails-A Consensus of Systemic Sclerosis Experts. Semin Arthritis Rheum 2012;42:42–55.

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