FRI0435

CLINICOPATHOLOGICAL CORRELATION IN INFLAMMATORY MYOSITIS: ANALYSIS OF THE REMICAM COHORT (REGISTRY OF INFLAMMATORY MYOSITIS FROM MADRID, SPAIN)

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Background: Associations between muscle pathology and clinical features in inflammatory myositis (IM) are not well defined

Objectives: to describe the pathological findings in REMICAM¹ muscle biopsies, and analyse their associations with clinical features

Methods: All patients with available biopsy were included. According to the score proposed by ENMC workshop², from muscle biopsy reports was extracted: inflammatory cells and location (endomysial, perimysial, perivascular), rimmed vacuola, fibre atrophy, necrosis, regeneration, and HLA expression. Descriptive analysis and association studies between histology and clinical features were performed (t and chi square tests, univariate logistic regression with OR)

Results: From 479 patients, 244 (51%) had available biopsy. Most frequent findings were: inflammatory infiltrates (75%): endomysial (44%), perimysial (49%) perivascular (26%); fibre atrophy (54%), perifascicular (27%); necrosis (55%) and regeneration (47%). HLA expression was + in 60/75 cases (80%). Endomysial infiltrate was associated with PM (OR 0.4; p<0.0001), cardiac involvement (OR 2.2; p=0.014), less arthritis (OR 0.5; p=0.01) and older age (51 vs 42 y; p<0.01); perimysial infiltrate and perifascicular atrophy with DM (OR 3.8; p<0.0001) and younger age (40 vs 48 y; p<0.05); necrosis with cardiac involvement (OR 2.1; p=0.035), neoplasia (OR 2.5; p=0.02) and no connective tissue disease (OR 0.4; p=0.01); and HLA with PM (OR 0.1; p=0.003)

Conclusions: most frequent histologic findings in IM muscle are inflammatory infiltrates, necrosis, HLA expression and perifascicular atrophy. Endomysial infiltration and HLA are characteristic of PM, and perimysial infiltration/perifascicular atrophy of DM. Endomysial infiltrates are associated with cardiac involvement. Necrosis is more frequent in neoplasia and less in connective tissue diseases, and is associated with cardiac involvement and older age. Our data suggest that muscle biopsy might help to identify those IM patients at higher risk of severe complications, as neoplasia or cardiac involvement

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FRI0436

COMPARISON OF THE 2017 EULAR/ACR CRITERIA WITH BOHAN AND PETER CRITERIA FOR THE CLASSIFICATION OF ADULT AND JUVENILE IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: The 2017 EULAR/ACR classification criteria provide a validated scoring system for the classification of both adult and juvenile idiopathic inflammatory myopathies (IIM) with reasonable sensitivity and specificity.

Objectives: To assess the performance of the 2017 EULAR/ACR criteria¹ in retrospective cohort of IIM and compare with Bohan and Peter criteria.^{2,3}

Methods: We conducted a retrospective study of all patients clinically diagnosed to have IIM in a tertiary care centre in the last ten years. Performance of both the criteria in the cohort was assessed and compared with clinical diagnosis.

Results: One hundred and seven patients were included in the study. Nine patients had juvenile onset. Clinical features are summarised in table 1. 80.4% of the patients were classified as probable/definite myositis using the Bohan and Peter criteria. 81.3% of the patients were classified as having probable/definite inflammatory myositis using the new criteria (score ≥5.5 without biopsy or ≥6.7 with biopsy). However the agreement between the two classification criteria was weak in our cohort (kappa=0.43). Complete details of muscle biopsy were available in 44 patients. In this subgroup 95.5% were classified by and Bohan and Peter and 84.1% by EULAR/ACR criteria. Both criteria performed poorly in patients without biopsy data and in polymyositis subset (table 2).

Abstract FRI0436 – Table 1. Clinical features of patients with idiopathic inflammatory myositis

Age (at diagnosis)	38.73 years	Proximal Muscle weakness	104 (97.2%)
Sex (Female: male)	3.12:1	Dysphagia	33 (30.8%)
Raynauds	22 (20.6%)	ILD	10 (9.3%)
Arthralgia/arthritis	71 (66.4%)	Anti Jo1 positivity (n=91)	3 (2.8%)
ANA positivity	59 (55.1%)	Elevated muscle enzymes	94 (87.9%)
Clinical Diagnosis	52 (48.9%)	Cutaneous involvement (s/o DM)	65 (60.7%)
Dermatomyositis	2 (1.9%)	Heliotrope	35 (32.7%)
Amyopathic DM	20 (18.7%)	Gottron's papules	21 (19.6%)
Polymyositis	22 (20.6%)	Gottron's Sign	8 (7.5%)
Myositis Overlap Syndrome	8 (7.5%)	Other rashes	18 (16.8%)
Juvenile Dermatomyositis	1 (0.9%)		
Juvenile onset myositis	2 (1.9%)		
overlap			
Cancer associated myositis			

Abstract FRI0436 – Table 2. Classification using Bohan and Peter and 2017 EULAR/ACR criteria

	Bohan and Peter	EULAR/ACR
Total IIM (n=107)	86 (80.4%)	87 (81.3%)
With Biopsies (n=44)	42 (95.5%)	37 (84.1%)
Without Biopsies (n=63)	44 (69.8%)	50 (79.4%)
DM (n=67)	62 (92.5%)	61 (91.0%)
PM (n=40)	24 (60%)	26 (65%)

Conclusions: The new EULAR/ACR criteria did not perform better than Bohan and Peter criteria in our cohort. The new criteria are more time consuming and seem to be of limited utility in daily clinical practice.

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FRI0437

IS EXTENDED AUTO ANTIBODY PROFILING ASSOCIATED WITH THE PREVALENCE OF DISEASE COMPLICATIONS?

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Background: Auto antibodies play a prominent role in both classification and disease prognosis of systemic sclerosis (SSc). In several cohorts the association between auto antibodies and prognostic factors has been described. In recent years, new methods of antibody profiling have become available, and have shown an important prognostic role for anti-RNAPolymerase 3 antibodies (anti-RNAP3). However, it is uncertain whether other auto antibodies have associations with complications as well.

Objectives: The aim of this study is to determine prevalence of auto antibodies in a well characterised cohort of SSc and to evaluate the associations with complications.

Methods: A total of 319 patients from the Nijmegen SSc Cohort were consecutively included in this study. All patients fulfilled the ACR/EULAR 2013 classification criteria for SSc. Patients were subclassified as limited cutaneous SSc (LcSSc), diffuse cutaneous SSc (DcSSc) or SSc overlap syndrome according to Leroy and Medsger. Blood samples were collected at regular outpatient clinic visits and analysed using LIA. (SSc and ENA immunoblot, Euroimmun, Lubeck, Germany). Clinical data was collected prospectively.

Results: The Nijmegen cohort comprises mainly of Caucasians. The percentage of male patients and diffuse patients is higher compared to other cohorts¹ (table 1). There is a relative anticentromere antibody (ACA) dominance in our cohort and ACA antibodies are more common in patients with pulmonary arterial hypertension (PAH), which is consistent with previous studies.¹ However, in our cohort we

found a relatively high prevalence of anti-RO52 antibodies in patients with complications (figure 1).

Clinical/laboratory feature/ total	No. 319	%
Male gender	100	31%
LcSSc	182	57%
DcSSc	102	32%
Overlap	35	11%
PAH	36	11%
ILD	134	42%
Cardiac involvement	21	7%
Renal crisis	3	0.9%
ACA	90	28%
ATA	63	21%
Anti-RNAP3	14	4.4%
Anti-SSA/Ro52	30	9.4%
RP-11	22	6.9%
RNP-SM	22	6.9%
RP-155	21	6.5%
M2-3E	21	6.5%
M2	21	6.5%
PM-75	19	5.9%
PM-100	19	5.9%

Abstract FRI0437 – Table 1. Demographic and clinical characteristics

Abstract FRI0437 – Figure 1. Prevalence of auto antibodies in SSc with and without complications. Antibodies < 10% prevalent not shown. ILD interstitial lung disease; PAH pulmonary arterial hypertension.

Conclusions: The distribution of the main auto-antibodies is comparable to other Caucasian cohorts.¹ Because the prevalence of anti-RNAP3 is relatively low, it is of limited value in our population. In contrast, the prevalence of anti-Ro52 was high in patients with the three major disease complications, but this antibody was also present in a number of patients who did not (yet) have these complications. Previous studies have shown ambiguous results concerning the relevance of this autoantibody.² Furthermore, RNP/SM and RNP-70 may be associated with cardiac involvement. To evaluate the prognostic value of anti-Ro52, RNP/SM and RNP-70 in SSc, a large prospective cohort study is necessary.

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FRI0438

HOSPITALISATION MORTALITY AND ASSOCIATED RISK FACTORS IN PATIENTS WITH POLYMYOSITIS AND DERMATOMYOSITIS: A RETROSPECTIVE CASE-CONTROL STUDY

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Background: Polymyositis and dermatomyositis (PM/DM) are systemic autoimmune diseases with multiple organ involvements that manifest as muscular and cutaneous disorders, interstitial lung disease (ILD) and malignancies. However, information concerning the outcomes and associated factors for PM/DM patients who are hospitalised is limited.

Objectives: We retrospectively reviewed the medical records of 982 PM/DM patients admitted to our centre from 2008 to 2014 and performed a case-control analysis to identify possible related risk factors for death among PM/DM patients in China.

Methods: We retrospectively reviewed the medical charts of PM/DM patients admitted to a Chinese tertiary referral hospital (Peking Union Medical College Hospital, PUMCH) from 2008 to 2014. The deceased group included 63 patients who had “deceased discharge” status or were confirmed to have died within two weeks of hospital discharge. The demographic data, clinical manifestations, and direct causes of death were analysed retrospectively. Medical records for 126 age- and sex-matched PM/DM patients were selected as controls from 982 inpatients successively admitted to the same centre during the same period. In addition to the comparison of clinical manifestations between the two groups, binary logistic regression was conducted to explore the risk factors related to PM/DM mortality.

Results: Over the past 6 years at PUMCH, the in-hospital mortality rate of PM/DM patients was 4.58%. The male gender and the elder patients had a high risk of death ($p=0.031$ and $p=0.001$ respectively). The three most frequent causes of death for PM/DM patients were pulmonary infection (35%), ILD exacerbation (21%) or both conditions (25%). Pulmonary infection ($p<0.001$, OR=5.63, 95% CI, 2.37–13.36), pneumomediastinum ($p=0.041$, OR=11.02, 95% CI, 1.10–110.54), Gottron’s papules ($p=0.010$, OR=3.24, 95% CI, 1.32–7.97), and elevated erythrocyte sedimentation rate (ESR) ($p=0.005$, OR=9.9, 95% CI 2.0–49.0) were independent risk factors for in-hospital mortality of PM/DM patients.

Conclusions: PM/DM patients continue to display high in-hospital mortality. Pulmonary infection is the strongest predictor of poor prognosis in PM/DM patients, followed by pneumomediastinum, Gottron’s papules, and elevated ESR.

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FRI0439

THE PROMISING ROLE OF LUNG MRI IN DETECTING SYSTEMIC SCLEROSIS (SSC)-RELATED INTERSTITIAL LUNG DISEASE (ILD)

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Background: Interstitial lung disease (ILD) is very frequent and highly disabling in patients with systemic sclerosis (SSc). Magnetic resonance imaging (MRI) is not routinely used for the evaluation of the lung, since MRI generally provides a less detailed view of the pulmonary parenchyma, as well as poorer spatial resolution, compared to the non-invasive gold-standard of high resolution computed tomography (HRCT). Thus, its usefulness in the evaluation of interstitial lung disease (ILD) is limited at present.

Objectives: To evaluate MRI signals in different pathological and non-pathological lung areas, and to establish their clinical and instrumental correlations.