

myopathy (IMNM,7) and 30 age-/sex-matched HC (25 females/5 males, mean age 54.9) without rheumatic/tumor diseases or manifest cardiovascular event were included. PM/DM patients fulfilled Bohan/Peter criteria for PM/DM. Anthropometric parameters and body composition were assessed (by densitometry-iDXA Lunar, and by bioelectric impedance-BIA2000-M), and physical activity was evaluated using Human Activity Profile (HAP) questionnaire. Routine biochemistry analysis was performed after 8 hours of fasting. Muscle involvement was evaluated by manual muscle test (MMT)-8. Data are presented as mean±SD.

Results: Compared to HC, patients with IIM had significantly increased body fat % as assessed by iDXA (BF%: 38.7±6.7 vs. 42.5±7.1%, $p=0.015$), but decreased lean body mass as assessed both by iDXA (LBM: 45.7±6.6 vs. 40.3±7.0 kg, $p=0.0005$) and BIA (LBM: 53.2±8.5 vs. 48.7±9.0 kg, $p=0.0295$), and increased ECM/BCM ratio (extracellular mass/body cell mass: 1.00±0.12 vs. 1.43±0.42, $p<0.0001$), which reflects worse muscle predispositions for physical exercise, aerobic fitness/performance, and also increases with deteriorating nutritional status. Compared to HC, IIM patients had significantly lower bone mineral density (BMD: 1.16±0.10 vs. 1.05±0.11 g/cm², $p=0.0010$), and were currently able to perform less energetically demanding physical activities according to HAP score (86.3±5.9 vs. 49.0±20.2, $p<0.0001$). Disease duration negatively correlated with BMD ($r=-0.392$, $p=0.004$) and LBM-BIA ($r=-0.272$, $p=0.047$). CRP was positively associated with BF% assessed both by DEXA ($r=0.276$, $p=0.035$) and BIA ($r=0.306$, $p=0.025$). MMT-8 score negatively correlated with ECM/BCM ratio ($r=-0.385$, $p=0.006$), and physical activity (HAP) negatively correlated with BF%_{DEXA} ($r=-0.292$, $p=0.032$).

Conclusions: Compared to healthy age-/sex-matched individuals we found significant negative changes in body composition of our IIM patients, which are associated with their disease duration, inflammatory status, skeletal muscle involvement, and physical activity, and could reflect their impaired nutritional status and predispositions for physical exercise, aerobic fitness and performance.

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AB0672 RISK FACTORS AND TREATMENT OF RECURRENT DERMATOMYOSITIS AND POLYMYOSITIS

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Background: Patients with polymyositis (PM) and dermatomyositis (DM) often have repeated exacerbations of myositis. Sometimes immunosuppressive agents are added to induce remission in steroid-resistant cases, with steroid reduction in maintenance therapy, and to prevent exacerbations. However, comparative clinical studies are difficult to conduct and the appropriate immunosuppressive agents are not known [1–2]. There are also no studies of the treatment in recurrent cases with repeated relapses.

Objectives: In this study, we defined myositis that relapses more than once as recurrent myositis and examined its risk factors and re-induction treatments.

Methods: Patients from our PM and DM database who were hospitalised between January 1991 and September 2016 were reviewed. We included only patients who had been followed for more than 1 year.

Results: There were 14 cases (13.1%) with recurrent myositis. The average observation period was 11.3 and 8.0 years in the relapsing and non-relapsing groups, respectively ($P=0.18$), and the average ages at onset were 46.2 and 55.0 years ($P=0.17$). The myositis relapse rate was significantly higher in the anti-aminocycl-tRNA synthetase (ARS) antibody-positive group than in the group with other antibodies [8/24 (33.3%) vs. 0/14 (0.0%) cases; $P=0.02$]. The myositis relapse rate was significantly lower in the group in which one more immunosuppressive agent was combined in the initial treatment than in the group without any [3/53 (5.7%) vs. 11/48 (22.9%) cases; $P=0.02$]. The myositis relapse rate did not differ significantly between DM and PM, men and women, the groups with or without malignancy, the groups with or without interstitial pneumonia (IP), and groups with or without the following initial findings: fever, joint pain, dysphagia, constipation, bedridden, muscle pain, distal muscle weakness, cutaneous symptoms (e.g., heliotrope rash, Gottron's sign, V signs, Raynaud's phenomenon, skin ulcers and necrosis, and itching sensation), cardiac complications, hypertension, diabetes, and smoking history. There was no significant difference in the serum creatine kinase and C-reactive protein between the relapse and no-relapse groups. Only 3 of 14 cases (21.4%) had successful re-remission induced and were on maintenance therapy. In two of these cases, two immunosuppressive agents other than glucocorticoid were added: tacrolimus and methotrexate in one and tacrolimus and mycophenolate mofetil in the other. The other 11 patients were treatment-resistant.

Conclusions: The presence of anti-ARS-antibody and initial treatment with glucocorticoid only (without any other immunosuppressive agent) were the only two risk factors for recurrent myositis; none of the other factors examined were significant. Recurrent myositis is often treatment-resistant; if one immunosuppressive agent cannot introduce remission, it is worth trying two agents.

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globulin infusions as treatment for dermatomyositis. *N Engl J Med.* 1993 Dec 30;329(27):1993–2000.

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AB0673 TESTING FOR ANTIBODIES SPECIFIC TO FIBRILLARIN (U3 RNP) SHOWS ADDED VALUE FOR DIAGNOSIS OF SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a heterogeneous and often systemic autoimmune disorder characterised by specific autoantibody subsets. The disease is associated with anti-nuclear antibodies (ANA) and the detection of each of the autoantibodies is useful during diagnosis. Most SSc patients are positive for anti-centromere or anti-Scl70 antibodies. The presence of more rare antibodies to, e.g. Fibrillarin (U3 RNP), RNA-polymerase III and PM-Scl are often indicative of a systemic or a limited skin involvement.

Objectives: In this in-house study the prevalence, and clinical sensitivity and /specificity of anti-Fibrillarin antibodies in a subset of patients diagnosed with SSc was investigated. The major aim was to find out whether Fibrillarin testing would identify patients which are negative for standard parameters and therefore add value for the diagnostic workup for patients with suspected SSc.

Methods: Sera from 187 SSc patients, 53 blood donors and 177 disease controls were analysed for the presence of antibodies to CENP, Scl-70s, PM-Scl, RNA Pol III and Fibrillarin using the ELIA platform (Thermo Fisher Scientific, Freiburg, Germany).

Results: In 149 out of the 187 patients one or more of the tested parameters were detected. Fibrillarin was identified in 7 out of the 149 patients including 6 solely positive for Fibrillarin. Overall sensitivity within this study is 79,68% with an overall specificity of 92,17%; for Fibrillarin sensitivity of 3,74% and specificity of 98,70% was shown within this cohort.

Conclusions: Increased diagnostic sensitivity was shown in this cohort of 187 patients with SSc by adding the analysis of antibodies against Fibrillarin. Testing allowed additional identification of 6 SSc patients which correlates to an increase of 3,74% in sensitivity.

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AB0674 YKL-40 IS THE BIOMARKER FOR THE PRESENCE OF INTERSTITIAL PNEUMONIA AND PULMONARY ARTERIAL HYPERTENSION IN SYSTEMIC SCLEROSIS

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Background: SSc is a refractory of connective tissue disease that causes fibrosis of the skin and various organs. Recently, it is attention to 2013 ACR/EULAR classification criteria because can diagnose early phase before causing skin involvement in SSc. Thus, we considered that YKL-40 which is known to be involved in inflammation, tissue fibrosis and remodeling, is useful marker for evaluating early diagnosis and complications in SSc. YKL-40 is a chitinase-like protein and expressed in the synovial membrane and cartilage fibroblast cells in Rheumatoid Arthritis. Several cases of relationship between SSc and YKL-40 have been reported overseas, but it is unclear. Our report is the first in Japan.

Objectives: We investigated serum YKL-40 levels and examined immunohistochemistry (IHC) with YKL-40 of cutaneous tissue in SSc patients. To clarify YKL-40 is useful biomarker in early diagnosis in the presence or absence of complications in Japanese SSc patients.

Methods: Between August 2014 and March 2016, we treated 57 SSc patients in our department. We excluded infection, malignant disease, and other active complication may be a factor that increases YKL-40 levels. The patients were divided into 4 groups depending on whether suffered IP or PAH, which can affect the prognosis. Group1 (n=30) did not suffer from either IP or PAH, Group2 (n=12) suffered from IP, Group3 (n=7) suffered from PAH, and Group4 (n=8) suffered from both IP and PAH. YKL-40 levels in 4 groups and a control group of healthy individuals (n=13) were measured by ELISA. And age percentile strata of YKL-40 were calculated because serum YKL-40 levels have reported to rise with age; YKL-40 age percentile = 100/(1 + (Serum YKL-40 levels⁻³) × (1.062²⁹⁹) × 5000). IHC with YKL-40 of skin biopsy tissue from patients and controls was performed to investigate YKL-40 expression.

Results: YKL-40 age percentile were elevated in Group1 compared to controls (42.0±24.5 vs 21.8±15.6), and were elevated due to complications (Group2, 3, 4: 61.8±21.4, 88.9±8.2, 94.4±8.7), with a tendency for PAH to cause greater elevation. There was no significant difference between Group3 and Group4, and

a significant difference was found between the other groups ($p < 0.05$). IHC with YKL-40 revealed staining of the vessels in the superficial dermis in SSc and normal skin remained unstained. The ROC curve for PAH showed an AUC=0.959, and the most valid cut-off levels of YKL-40 age percentile was 85.2 (sensitivity 86.7% and specificity 92.8%).

Conclusions: Our report is the first report of IHC with YKL-40 in SSc. IHC suggest that YKL-40 may be further elevated by angiogenesis caused by microcirculatory damage in SSc. Our results suggest that YKL-40 may be very useful biomarker for diagnosing SSc and complications associated with microvascular lesions.

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AB0675 ANTI-MDA5 POSITIVE DERMATOMYOSITIS: AN EMERGING ENTITY

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Background: Hypo or amyopathic dermatomyositis (DM) represent about 20% of the classic DM. Melanoma differentiation-associated gene 5 antibody or anti-MDA5 antibody, formerly known as anti-CADM140 antibody, has been reported in the last decade. This antibody is detected in more than 50% of patients with hypo or amyopathic DM and it has specific features such as the presence of ulcerative vascular lesions and rapid progression of the interstitial lung disease.

Objectives: To report anti-MDA5 positive DM cases diagnosed at a university county hospital, between 2012 and 2016.

Methods: Design: Retrospective descriptive study. Location: University county hospital. Reference area: ~350.000 inhabitants. Clinical manifestations, analytical results, diagnostic approach, treatment administered and follow-up have been exposed.

Results: We report four anti-MDA5 positive DM with a mean age at diagnosis of 54 years (35–77). Caucasian origin in 2 men and Maghreb origin in 2 women. They had no relevant medical history neither neoproliferative process was detected. ANA were negative and CK values were normal in all the cases. Nailfold capillaroscopy was characteristic of DM pattern. Case B did not present cutaneous manifestations and he was diagnosed as anti-MDA5 DM after his death.

Discussion: B and C cases showed abnormal heart rhythm that could be related to myocardial involvement, as suggested in a recent publication of anti-MDA5 DM (1). In case C, sinus tachycardia was resolved with the clinical response to treatment.

In the literature reviewed, anti-MDA5 positive DM has been reported as a dermatomyositis syndrome, but probably the clinical profile of these patients remains to be defined. In our cases, one of them did not present skin lesions and another case did not develop pulmonary involvement 18 months after the diagnosis.

Conclusions: Keep in mind anti-MDA5 antibody in patients with cutaneous manifestations of DM with vasculopathy lesions, minimal or absent muscular disease, heart rhythm abnormalities not explained by other causes and/or rapidly progressive pulmonary involvement.

Anti-MDA5 positive DM may have a poor response to treatment and fatal outcome. It remains to be seen if early diagnosis could improve the life expectancy of these patients.

Abstract AB0675 – Table 1

| | A | B | C | D |
|-------------------------|------------------------|----------------------------------|--------------------------|----------------------------------|
| Sex | Female | Male | Female | Male |
| Age at disease onset | 43 | 77 | 35 | 62 |
| Fever | Yes | No | No | No |
| Heliotrope rash | Yes | No | Yes | No |
| Gottron papules | Yes | No | No | No |
| Skin ulceration | No | No | No | Yes |
| Joint disease (J) | No | No | RA like | Palindromic like |
| Raynaud's disease | No | No | No | No |
| Muscular disease (M) | No | No | Yes | No |
| Pulmonary disease (P) | Bronchiolitis | Interstitial lung disease | No | Interstitial lung disease |
| Cardiac disease (Card) | No | Arrhythmias | Sinus tachycardia | No |
| Aldolase (N<7) | Normal | NA | 11 | 14,6 |
| ESR/CRP mg/dL | 30/4,5 | 41/13,7 | 102/8,1 | 50/1,8 |
| Ferritin (30–400 ng/ml) | 1253 | NA | 225 | 2437 |
| Electromyography | NA | NA | Normal | NA |
| Muscle biopsy | NA | NA | Miopathy | NA |
| Skin biopsy | DM | NA | Nonspecific | DM |
| Treatment | CC + CPM + TACR + IVIG | CC | CC + MTX + HCQ | CC + CPM + TACR |
| Outcome (months) | Death | Death | Clinical response (18 m) | Alive (16 m) |

RA = Rheumatoid Arthritis; NA = Not Available; ANA = Antinuclear Antibodies; DM = Dermatomyositis; CC = Corticosteroids; CPM = Cyclophosphamide; TACR = Tacrolimus; IVIG = Intravenous Immunoglobulins; MTX = Methotrexate; HCQ = Hydroxychloroquine.

References:

- [1] *J Eur Acad Dermatol Venereol* 2014; 28: 1097–102.

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AB0676 RELATIONSHIP BETWEEN BIOLOGICAL BIOMARKERS AND CHANGES IN RIGHT VENTRICLE THAT PRECEDE PULMONARY HYPERTENSION IN PATIENTS WITH SYSTEMIC SCLEROSIS

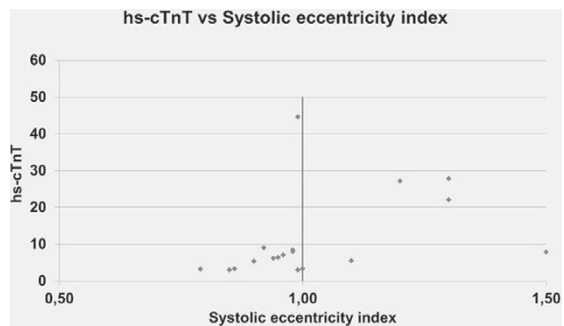
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Background: A higher prevalence of Pulmonary Hypertension (HP) has been described in patients with Systemic Sclerosis (ES) diagnosis, so this fact significantly overshadows the prognosis, since it leads to the failure of the right ventricle (RV). In this context, it could be very useful to identify ES patients who are at greater risk of developing PH in the future to make an early diagnosis of this complication, and prescribing an early therapy to improve their prognosis.

Objectives: The purpose of this study is to investigate if certain fibrosis, inflammation and vasculopathy biomarkers could be associated to early functional changes in RV, assessed by echocardiography and cardiac nuclear magnetic resonance (MRI) in ES patients.

Methods: A cross-sectional study was performed on a cohort of ES patients diagnosed according to the LeRoy modified criteria, who were being followed-up in the Systemic Autoimmune Diseases Unit of the Internal Medicine Service of La Paz University Hospital. Serum levels of inflammation (IL-13, IL-6, TNF alpha), vasculopathy (VEGF), fibrosis biological markers (endoglin, PDRFR, TGFB1, TGFB2, TGFB3) and cardiac biomarkers (NT-proBNP, High sensitivity T troponin) were determined as well as hemodynamic and cardiac morphological variables, were measured by transthoracic echocardiography (systolic and diastolic eccentricity index, E/E' ratio, E/A, pulmonary artery systolic pressure, RV thickness) and cardiac MRI (Mean pulmonary artery velocity). We investigated how many patients had these markers elevated and how many of them had hemodynamic and morphological cardiac altered measurements.

Results: Twenty patients with a diagnosis of ES (18 females/2 males) aged between 41 and 77 years, with a mean of 56 years, were included. A control group of 9 patients with similar demographic characteristics was included. ES subjects had higher levels of PDGFR (fibrosis marker) compared to the control group. In addition, in the group of ES patients, two statistically significant associations were observed: troponin T levels (hs-cTnT), endoglin and TGFB2 had higher levels in patients with a systolic eccentricity index >1; a negative correlation was evidenced between levels of NT-proBNP and the mean pulmonary artery velocity measurement (VELAP).



Conclusions: Hs-cTnT, Endoglin and TGF beta are biomarkers that could appear