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

V. Ortiz-Santamaria, A. Ponce, N. Busquets, N. Del Castillo, X. Surís.
Rheumatology Unit, Granollers General Hospital, Granollers, Barcelona, Spain

Background: Hypo or amypathic 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 amypathic 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
Gotttron 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.

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

V. Márquez Fernández¹, C. Soto¹, A. Robles¹, A. Noblejas¹, E. Martínez¹, G. Gúzman², C. Busca¹, F. Arnalich¹, J.J. Ríos¹. ¹Internal Medicine; ²Cardiology, Hospital Universitario la Paz, Madrid, Spain

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 following-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