SAT0491

CLINICAL FEATURES AND COMPLICATIONS IN A LARGE INTERNATIONAL COHORT OF ANTIMDA5 PATIENTS: A CHALLENGE FOR THE FUTURE

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Background: Anti-MDA5 antibodies are a known set of antibodies observed mainly in dermatomyositis, typically associated with cutaneous involvement, presence and often rapid progression (RP) of interstitial lung disease (ILD) and amyopathic muscle involvement. Despite the increased attention, described cohorts involves only a limited number of cases

Objectives: to define clinical characteristics of a large cohort of anti-MDA5 antibodies positive patients

Methods: Retrospective assessment of anti-MDA5 positive patients from centres referring to our group

Results: 82 anti-MDA5 positive cases (56 females) were collected. In median: onset age was 44 Y (IQR 22.57), diagnostic delay 4 Mo (IQR 1–10), follow-up 13 Mo (IQR 3–43). Fifty-three patients had ILD, that was RP in 24 cases (15 at ILD onset, 9 after ILD onset). Fifteen ILD patients were admitted in Intensive Care Unit (ICU): 4 were treated with Extracorporeal-Membrane-Oxygenation, 7 with mechanical-invasive and 2 with mechanical-non-invasive ventilation. Forty-nine patients had muscle involvement (39 symptomatic), 42 arthritis, 54 cutaneous involvement, 9 history of malignancy. Twelve patients died (ILD=7, ILD and sepsis=3, neoplasia=1, not specified=1), 7 of those in ICU

Conclusions: In our cohort, ILD was the most frequent finding. A RP of ILD was common, occurring also in ILD with symptomatic/chronic onset. The low rate of survival in ICU raised the problem of follow up and early treatment of ILD. In our cohort arthritis was common and muscle involvement mainly symptomatic. Finally, the high percentage of observed malignancies suggests a careful neoplastic screening and follow up

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PREVALENCE OF THE METABOLIC SYNDROME IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: The metabolic syndrome is an independent risk factor for ischaemic heart disease. Patients with systemic lupus erythematosus have an increased prevalence of the metabolic syndrome.¹ There are no controlled studies of the metabolic syndrome in patients with SSc.

Objectives: To compare the prevalence of the metabolic syndrome in patients with SSc and controls and to evaluate its relationship to activity and severity of disease.

Methods: Methods: 50 consecutive patients with SSc (45 female, median age 48; range 20–72) and 50 controls (patients with fibromyalgia) (45 female, median age 46; range 25–72) were studied. The prevalence of the metabolic syndrome was compared in patients and controls using the National Cholesterol Education Program Adult Treatment Panel III (NCEP-ATPIII): central obesity: waist. 102 cm in men and 88 cm in women;² hypertriglyceridaemia: >150 mg/dl;³ low HDL, 40 mg/dl in men and 50 mg/dl in women;⁴ high blood pressure: >130/85 mm Hg or use of drugs for high blood pressure; and⁵ high fasting glucose >110 mg/dl, and associations with activity and severity of the disease were examined.

Results: The metabolic syndrome was present in 24% of patients and in 30% of controls subjects (p>0.05). Among patients with SSc, the metabolic syndrome was significantly associated with higher value of ESR (ESR >30) (5/7 vs 4/34; p=0.027). Neither disease activity nor severity scores were associated with the metabolic syndrome

Conclusions: Patients with SSc have a lower prevalence of the NCEP-ATPIII-defined metabolic syndrome than controls. This result may help to justify the coronary prevalence in SSc patients similar to that of the general population.^{2–3}

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PLASMA LEVELS OF HSP90 ARE INCREASED IN INTERSTITIAL LUNG DISEASE AND SKIN FIBROSIS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Our previous study demonstrated that Heat shock protein 90 (Hsp90) is overexpressed in the skin of patients with systemic sclerosis (SSc), in cultured SSc fibroblasts and preclinical models of SSc in a TGF- β dependent manner. We showed that Hsp90 is a regulator of TGF- β signalling and its inhibition prevents the stimulatory effects of TGF- β on collagen synthesis and dermal fibrosis in three preclinical models of SSc¹.

Objectives: The aim of this study was to evaluate Hsp90 in the circulation of SSc patients and characterise its potential association with skin changes and SSc-related features.

Methods: A total of 91 patients (78 females; mean age 52.7; disease duration 6.0 years; diffuse cutaneous (dc)SSc/limited cutaneous (lc)SSc=38/53) who met the ACR/EULAR 2013 classification criteria for SSc and 85 age-/sex- matched healthy individuals were included. Plasma Hsp90 was measured by ELISA (eBio-science, Vienna, Austria). Data are presented as median (IQR, 25–75. percentile).

Results: Plasma Hsp90 levels were increased in SSc patients compared to healthy controls [12.5 (9.6–17.9) vs. 9.9 (7.9–12.6) ng/mL, p=0.001], but no difference between lcSSc and dcSSc was detected [13.1 (9.4–18.1) vs. 11.5 (9.5–17.5) ng/mL, p=0.316]. Hsp90 levels in all patients positively correlated with CRP