increased in both patient groups ($p<0.01$) and mDCs expressing CXCL8 only in lcSSc ($p<0.01$). SSc patients characterized by the presence or history of lung fibrosis, displayed a higher frequency of non-classical monocytes expressing CCL4 and CXCL10 in dcSSc patients as compared to those without this clinical manifestation ($p<0.01$ and $p<0.05$ respectively). Strikingly, the percentage of classical monocytes producing CXCL8 was augmented upon *in vitro* stimulation in lcSSc patients with lung fibrosis as compared to those without ($p<0.01$). No differences were found in the percentage of IL-6 producing cells.

Conclusions: These data point towards a role of activated non-classical monocytes and mDCs producing enhanced levels of proinflammatory cytokines in SSc, potentially contributing to lung fibrosis.

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Scleroderma, myositis and related syndromes

SAT0325 NAILFOLD CAPILLAROSCOPIC CHANGES IN PATIENTS WITH IDIOPATHIC AND SYSTEMIC SCLEROSIS-RELATED PULMONARY ARTERIAL IPERTENSION

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Background: Pulmonary arterial hypertension (PAH) represents one of the main clinical expression of the vascular changes in Systemic Sclerosis (SSc). Many clinical and experimental evidences suggest that lung microvascular changes play a role in the pathogenesis of idiopathic PAH (IPAH) also.

Objectives: The aim of this study is to investigate the presence of capillaroscopic abnormalities in patients with idiopathic PAH and to evaluate the differences in capillary naifold changes between patients with IPAH and SSc patients with and without PAH.

Methods: 37 subjects with SSc (of whom 17 with PAH), 21 subjects with IPAH and 20 healthy subjects were recruited. PAH was diagnosed by right heart chateterization. Periungual capillaroscopy was performed in all recruited subjects, considering the following parameters: loops length and width, capillary density, microhemorrhages, avascular areas, neoangiogenesis. To define the pattern of capillary changes in IPAH and healthy subjects a semiquantitative scoring (normal, minor abnormalities, major abnormalities) was used, whereas in SSc subjects the capillary changes were defined as early, active and late pattern.

Results: In all SSc subjects a capillaroscopic scleroderma pattern was found. Particularly, comparing SSc-PAH vs SSc-nonPAH we found an early pattern in 26,7% vs 50%, an active pattern in 66,6% vs 33,3% and a late pattern in a 6,6 vs 16,7% of subjects. None of IPAH subject presented a capillaroscopic scleroderma pattern, but interestingly in 36,4% of minor or major capillaroscopic changes were found. Analysing the single capillarocopic parameters, capillary density was lower in SSc subjects compared to the other groups; in SSc-PAH was lower than in SSc-nonPAH; in IPAH capillary density was lower compared to healthy control. Capillary width was higher in SSc patients compared to healthy and IPAH subjects, being higher in SSc-PAH compared to SSc-nonPAH. The number of megacapillaries, bushy capillaries and microhemorrages was significantly higher in patients with SSc-PAH compared to SSc-nonPAH patients. Interestingly, compared to healthy controls, the IPAH subjects presented a significantly lower capillary density and a significantly higher mean capillary width. Further, IPAH subjects presented a significant increase of number of microhemorrages and ectasic capillaries compared to healthy controls.

Conclusions: Microcirculation alterations, and particularly the reduction of capillary density and the increase od capillary width appears to be more severe in SSc subjects with PAH compared to SSc subjects without PAH. Capillaroscopic changes can be present in IPAH subjects also. These data support the hypothesis that in SSc peripheral microcirculation changes can be related to the entity of pulmonary microcirculation changes and that an altered vascular lung remodelling could play a role in IPAH also.

References:

[1] Cutolo M et al. The contribution ofcapillaroscopy to the differential diagnosis of connective autoimmune diseases. Best Pract Res Clin Rheumatol. 2007 Dec;21(6):1093–108.

[2] Riccieri V. Systemic sclerosis patients with and without pulmonary arterial hypertension: a nailfoldcapillaroscopy study. Rheumatology (Oxford). 2013 Aug;52(8):1525–8.

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SAT0326 THE ASSCOCIATION OF SERUM TYPE 1 INTERFERON ACTIVITY AND AUTOANTIBODIES IN INFLAMMATORY MYOSITIS

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Background: Recent reports had shown that most of clinically amyopathic dermatomyositis (CADM), which showed poor prognosis, was positive for anti-Melanoma differentiation-associated gene 5 (MDA5) antibody (Ab). It had been shown that patients not only with lupus but with dermatomyositis (DM) also showed increased type 1 interferon (IFN) signature. MDA5 acts as a cytosolic RNA sensor, which drives type 1 IFN production. These facts suggested that type 1 IFN might have some roles in anti-MDA5 Ab positive patients.

Objectives: We evaluated the association of serum type 1 IFN signature and autoantibodies in patients with inflammatory myositis, in particular anti-MDA5 Ab positive patients and anti-aminoacyl-tRNA synthetases (ARS) Ab positive patients.

Methods: Sera from 33 inflammatory myositis patients (13 DM, 10 PM and 10 CADM) were studied for type 1 IFN activity, using a functional reporter cell assay. Briefly WISH cells were incubated with serum containing media for 6 hours. Serum IFN signature scores of the incubated cells were evaluated by the sums of gene expressions of Mx1, IFIT3, IFI44L and IFI44 by real time PCR (Reference). Anti-MDA5 Ab and Anti-ARS Ab were measured by ELISA. We divided these patients into three groups, anti-MDA5 Ab positive group (MDA5 group), anti-ARS Ab positive group (ARS group) and double negative group (DN group). We included double positive patients into MDA5 group. The presence of interstitial lung disease (ILD) and the prognosis were also investigated.

Results: MDA5 group had 12 patients (8 CADM and 4 DM), ARS group had 8 patients (4 DM, 1 CADM and 3 PM), and DN group had 13 patients (5 DM, 1 CADM and 7 PM). 9 of MDA5 group, 5 of ARS group, and 1 of DN group were complicated with ILD. Serum IFN signature scores of MDA5 group were significant higher than those of ARS group and DN group (12.43 1.406, 2.407, p=0.0005). The most of ARS group showed low serum IFN signature activities. The overall survivals of ARS group were fairer than MDA5 group, but not significantly. The deceased cases of MDA5 group showed especially high serum IFN signature activities.

Conclusions: We characterized two major groups in inflammatory myositis patients. ARS group was characterized by low IFN signature with the susceptibility to DM and ILD. MDA5 group was characterized by high serum IFN signature with the high susceptibility to CADM. Our results suggest that these two entities may have different onset mechanisms, leading to different outcomes.

References:

[1] Niewold TB, Rivera TL, Buyon JP, Crow MK. Serum type I interferon activity is dependent on maternal diagnosis in anti-SSA/Ro-positive mothers of children with neonatal lupus. Arthritis Rheum. 2008 Feb;58(2):541–6.

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SAT0327 NUTRITIONAL STATUS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a chronic connective tissue disease characterized by involvement of multiple organs. Many clinical aspects, such as gastrointestinal involvement, mood disturbances, functional status, and inflammation, may lead to disease-related malnutrition [1]. The connection between inadequate nutritional status and systemic sclerosis is still not well established. It is important to identify the symptoms of malnutrition, because it is known as a predictor of poor clinical outcome [2].

Objectives: To assess nutritional status in patients with systemic sclerosis.

Methods: The study involved fifty-two patients with SSc (44 women and 8 men, mean age 54,3±11,7 year) who were diagnosed according to ACR/EULAR criteria. The assessment of nutritional status was determined by subjective global assessment (SGA), body mass index (BMI) and level of serum albumin. Appetite was assessed by simplified nutritional appetite questionnaire (SNAQ). In all patients hand grip strength and triceps skinfold were established. The C-reactive protein (CRP), lipid profile, and level of haemoglobin/lymphocytes were measured in serum.

Results: Inadequate nutritional status was diagnosed in 14 patients (26,9%) with SSc. According to SGA 11 (21,15%) patients had signs of mild malnutrition, while 41 (78,85%) were well-nourished. Considering BMI, 1 patient (1,92%) was underweight, 24 (46,15%) were eutrophic, 21 (40,38%) overweight and 6 (11,54%) obese. Significantly lower BMI had patients with inadequate nutritional status (23,17±4,47 vs. 25,98±3,34; p=0,009). Low level of serum albumin was detected in 5 patients (9,6%) with SSc. Level of serum albumin and appetite were significantly decreased in patients with inadequate nutritional status (p=0,009; p=0,003). No statistical differences were noticed in hand grip strength, triceps skinfold, lipid profile, levels of CRP and haemoglobin/lymphocytes.

Conclusions: Malnutrition in systemic sclerosis is still underestimated clinical issue. This study provides useful data about nutritional status of patients with