diagnosis of these patients are shown in Table 1. In the cluster analysis we could not identify clinical phenotypes, perhaps because of the small sample size. Only 50% of patients with myositis developed ILD. Regarding the final diagnosis, only 1 patient (5%) was diagnosed of scleromyositis. Besides detecting them in patients with SSc (39%) and idiopathic inflammatory myopathy (9%), anti-Ku Ab were detected in other SAD, the most frequent were systemic lupus erythematosus, rheumatoid arthritis (RA) and overlap syndrome of SSc + RA.

Table 1. Main clinical-analytical manifestations and final diagnosis of pacientes with anti-Ku Ab.

FINAL CLINICAL DIAGNOSIS

Scleromyositis: 1
Idiopathic inflammatory myopathy: 1
Systemic sclerosis (SSc): 6 (Pre-scleroderma: 3, limited SSc: 3).
Systemic lupus erythematosus: 2
Rheumatoid arthritis: 2
Overlap syndrome RA + limited SSc: 2
Primary Sjögren's syndrome: 1 (Secondary Sjögren's syndrome: 3)
Mixed connective tissue disease: 1
Polymyalgia rheumatica: 1
Undifferentiated connective tissue disease: 1
Acute hepatitis due to HEV: 1
Autoimmune thrombocytopenia (ITP): 1
Drug-induced fibrosing ILD: 1
Systemic graft versus host disease (GVHD) in a patient with acute lymphoblastic leukemia who received and autologous haematopoietic progenitor transplant: 1
Primary biliary cirrhosis: 1

CLINICAL MANIFESTATIONS (patients could have more than one):

Raynaud's phenomenon: 61% (14/23).
Inflammatory arthralgia/arthritis: 52% (12).
Lung involvement: 30.5% (7: NSIP 3, UIP 2, other patterns 2).
Serositis: 26% (6: pericarditis 4, pleuritis 1, pleuropericarditis 1).
Cardiac involvement: 26% (6: PHT by echocardiogram 3, myocarditis 2, arrhythmia 1).
Dry syndrome: 17% (4)
Myositis: 17% (4).
Esophageal involvement: 17% (4).
Autoimmune cytopenias: leucopenia: 17% (4) / thrombocytopenia: 13% (3).
Telangiectasias: 13% (3).
Photosensitivity: 13% (3).
Other: non-androgenic alopecia: 9% (2); sensory-motor polyneuropathy: 4.5% (1); Puffy hands: 4.5% (1); fever: 4.5% (1); lymphadenitis: 4.5% (1); cold sores: 4.5% (1), and retinal hemorrhage: 4.5% (1).

OTHER ASSOCIATED ANTIBODIES:

ANA: 91% (21/23)
anti Ro60/SSA Ab: 30.5% (7)
Anti Ro52 Ab: 30.5% (7)
Anti RNP Ab: 22% (5)
Anti-dsDNA: 17% (4)
Anti-La/SSB Ab: 17% (4)
Anticentromer Ab: 17% (4)
Anti Mi-2 Ab: 13% (3)
Other: anti Sm Ab: 9% (2); anticardiolipin Ab(IgG): 4.5% (1); PM/ScI: 4.5% (1); nucleosomes: 4.5% (1); ScI70: 4.5% (1); PL12: 4.5% (1); anti-U1-RNP: 4.5% (1) and NOR90: 4.5% (1).

Conclusion: Anti-Ku Ab are related to a great variety of SAD, without being a specific marker of any of them, nor being associated with any specific clinical manifestation. We couldn't confirm the existence of clinical phenotypes associated with the presence of these antibodies.

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AB0164

ACTIVITIES OF THE OXIDATIVE-RELATED ENZYMES IN SYSTEMIC SCLEROSIS

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Background: The oxidative-related enzymes are involved in the pathogenesis of various stages of systemic sclerosis (SSc). SSc is a chronic autoimmune disorder that is intimately associated with vascular damage and therefore with chronic perfusion/reperfusion and oxidative organ injury. Mesenchymal cell activation in SSc is now also considered to be mediated primarily through oxidative burst. Regulation of oxidative stress by specific enzymes including several purine metabolism enzymes is likely to play an important role in SSc progression.

Objectives: to characterize interrelationships among circulating xanthine oxidase (XO), xanthine dehydrogenase (XDH), superoxide dismutase (SOD) activities and SSc activity.

Methods: The study was performed according to bioethical standards. 51 patients with verified SSc and 30 healthy controls were included in the study. The diagnosis was verified according to ACR/EULAR 2013 criteria. We assessed SSc activity in compliance with the original activity scale that is commonly used in Russia [Guseva N.G., 1993] and by the 2001 European Scleroderma Study Group Activity Index. XO (EC 1.17.3.2), XDH (EC 1.17.1.4), and SOD (EC 1.15.1.1) plasma activities were measured using spectrophotometric techniques as previously described [Dubinina E.E., 1986; Karpova O.V., 2006]. Results are expressed as mean±SD. The Mann-Whitney U test and Spearman's correlation coefficient were used for statistical analysis.

Results: Mean age of patients was 42.8±1.3 years, mean SSc duration was 7.9±0.7 years. Mean enzymatic activities in normal controls were 3.43±0.56 nmol/ml min (for XO), 5.19±0.71 nmol/ml min (for XDH), and 5.40±1.03 units (for SOD). The respective enzymatic activities in SSc group were 3.91±0.62 nmol/ml min, 7.10±0.71 nmol/ml min, and 7.10±2.19 units. All these mean activities were significantly higher in SSc patients comparing to healthy individuals (p<0.001). XO and XDH activities positively correlated with SSc activity (r=0.499, p<0.001, and r=0.741, p<0.001, respectively). The opposite but weaker trend was observed for SOD activity and SSc disease activity (r=-0.190, p=0.188).

Conclusion: SSc is characterized by an increase in the intensity of oxidative and antioxidant processes, more pronounced in high disease activity. A close relationship between function of prooxidant/antioxidant enzymes and some of the key SSc pathogenetic mechanisms, especially vascular disease and fibroblast activation, is widely considered. Overall increase of oxidative stress in patients with higher disease activity, as well as depletion of antioxidant capacity can be also linked with disturbance of purine metabolism through XO and XDH modulation. Pathogenetic influence of this imbalance can also be mediated through initial phase of neutrophil extracellular traps (NETs) formation, an eventual source of nucleoprotein containing autoepitopes.

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AB0165

THE ROLE OF CXCL4, CXCL8 AND GDF-15 IN SYSTEMIC SCLEROSIS

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Background: Systemic Sclerosis (SSc) is an autoimmune disease that can affect several organs and its mortality is fundamentally related to its pulmonary involvement.

It is mandatory to seek for biomarkers that help us with early diagnosis and that are also useful for predicting organic involvement, so that we can adjust the diagnostic and therapeutic approach.

Objectives: Our aim was to check if the presence of CXCL4, CXCL8 and GDF-15 is greater in the disease than in healthy population, and also their involvement in organic damage.

Methods: Observational and cross-sectional study, with a prospectively performed protocol, of patients diagnosed of SSc according to ACR/EULAR 2013 criteria. Demographic, clinical, analytical, activity (EUSTAR index), severity (Medsker scale and modified Rodnan index), health perception (SF36) and disability (HAQ and Cochin test) variables were collected. Moreover, Videocapillaroscopy (VCL) and Respiratory Function Test were made, as well High Resolution Lung Tomography and Echocardiography in order to describe pulmonary features. Serum levels of CXCL4, CXCL8 and GDF-15 were measured in SSc patients and in healthy controls.

Results: A total of 42 patients (95.4% women) were included, with an average age of 59.2 years. The median of years since diagnosis was 4, by 6 since the first non-Raynaud symptom. 20 patients were diagnosed with limited SSc, 20 patients diffusely and 2 patients with SSc without scleroderma. 42 healthy controls were also included.

We found significantly higher levels of GDF-15 in patients with SSc (P<0.001), without significant differences in CXCL4 and CXCL8 levels between patients with SSc and healthy controls.

The presence of GDF-15 was associated with diffuse SSc (P=0.009), pulmonary arterial hypertension (P=0.038), interstitial lung disease (P=0.004), decreased forced vital capacity (FVC), (P=0.002), high serum titres of antiScI70 (P=0.006), increased disease activity measured by EUSTAR index (P=0.001), as with capillary dilations in Capillaroscopy (P=0.015).

Moreover, we found an association between CXCL4 levels and the consumption of complement C3 fraction (P=0.008) and skin involvement (higher Rodnan modified score), (P = 0.001); not finding association with lung involvement or other features (spirometric or analytical changes, capillaroscopy or functional tests).