Abstract AB0632B Figure 1: View of the larynx on endoscopy showing the suspicious nodules of granulomatous inflammation

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Scleroderma, myositis and related syndromes

AB0633

FECAL CALPROTECTIN IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a connective tissue disease with heterogeneous symptoms and a non-predictable process. The most involved organ after the skin is the gastrointestinal tract (GI tract) and any part of it can be affected. The calprotectin level is increased in plasma, urine, sputum and feces when an inflammatory process is occurring with the presence of neutrophils which is measurable.

Objectives: So we decided to examine fecal calprotectin as a simple method to diagnose GI disorders and disease activity in SSc.

Methods: Totally 46 patients with SSc were invited by telephone, who have been referred to sayyad shirazi Hospital and the rheumatologists' offices in Gorgan and their information has been registered there. Seven patients did not enter our study because of having the items in exclusion criteria like Diabetes, a history of GI operation and having other connective tissue diseases at the same time. For the other 39 patients who was remained, a questionnaire about GI symptoms (UCLA SCTS 2.0) was filled in and then their skin was examined and the skin score was determined (the score more than 14 was considered as positive). Blood sample was taken from all patients to do tests as ESR, CRP, Magnesium, Albumin, Ferritin, Folic Acid, Vitamin B12 and Zinc and use them as a scale for malnutrition. Then the stool samples were collected to measure the calprotectin level. At last to determine GI disorders in a non-aggressive way, Barium Swallow (Esophagus malfunction) and CT Enterography (intestinal disorders) was done. The patients in our study according to the median of age which was 42, was divided in to two groups: young and middle aged. Due to the fact that FC level wasn't less than 50 µg/gr in any of our cases, the patients was divided in to two groups: the one with FC level less than 200 µg/gr and the one with FC level as 200 µg/gr or more than 200 µg/gr and the latter was considered as positive.

Results: As we didn't have any male patients, the correlation between FC level and sex was not verifiable. Increased levels of FC didn't have any correlation with age (P Value=0.79), disease type (P Value=0.59), small intestine wall thickening (P Value=0.10), esophageal dilatation (P Value=0.18), skin score (P Value=0.25), final GI score (P Value=0.30) and none of serum tests. One of the most important reasons, was low sample size. In our study just 5 cases had evidence of wall thickening in CT Enterography and all that 5 cases had increased levels of calprotectin so we see that if we have a larger sample size we may ensure the

connection between these two variables. Also as most of our patients take vitamin and mineral supplements, we couldn't find any signs of micronutrient deficiencies and any correlation with FC level as well.

Conclusion: Although there was no statistical correlation between FC level and variables, the results are in favor of FC's specificity for assessing intestinal wall thickening.

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AB0634

FINDINGS OF EXERCISE ULTRASOUND CARDIOGRAPHY IN PRE-PAH PATIENTS WITH SYSTEMIC SCLEROSIS ARE CORRELATED WITH EXTENT OF CAPILLARY LOSS DETECTED BY NAILFOLD VIDEOCAPILLAROSCOPY

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Background: Systemic sclerosis (SSc) is characterized by excessive fibrosis, injuries in vasculature, and production of autoantibodies (1). Patients with SSc are complicated with various organ involvements, which can give a great impact on their prognosis (2). Of these, pulmonary arterial hypertension (PAH) is one of leading causes of death and the determinant of prognosis in SSc (3). And early diagnosis is an important key to improve their prognosis. The nailfold videocapillaroscopy (NVC) is a non-invasive modality which can evaluate the status of microvasculature of patients with SSc. However, the significance of NVC in patients with "pre-clinical" PAH was unclear.

Objectives: To examine the significance of detection of microvascular injury by NVC in patients with pre-clinical PAH patients with SSc.

Methods: Patients with SSc who newly visited the outpatient clinic at Fujita Health University Hospital between August 2015 to March 2016 were consecutively involved. All the patients fulfilled the 1980 preliminary classification criteria for SSc (4). The diagnosis of PAH was based on the definition in 2015 ESC/ERS guideline. The NVC (Medcap[®]3.0, DS MEDICA) was used to detect microvascular change and average capillary density and the number of capillaries/view were calculated. Also, existence of giant capillaries, hemorrhage and patterns of capillary changes were also documented. Transthoracic ultrasound cardiography (UCG) was performed using iE33 with 5MHz sector probe S5-1 (Philips). Exercise loading was done with an ergometer (Lode). Ejection fraction, E/e', velocity of tricuspid regurgitation jet (TR jet), tricuspid annular plane systolic excursion (TAPSE) were examined in a supine position. Tricuspid regurgitation pressure gradient (TRPG) was calculated using velocity of TR jet based on Bernoulli principle. Exercise loading was started at 25 Watt, and then increased 25 Watt every 3 minutes and TRPG at maximum exercise was also calculated. Parameters of NVC were compared with those of UCG. Patients with "pre-clinical" PAH were defined as patients 1) asymptomatic, 2) who did not meet the definition of PH by right heart catheterization (RHC), 3) whose probability of PH is lower than "intermediate" by UCG, 4) who did not see any UCG signs for PH based on the guideline and 5) who did not have severe interstitial lung disease, especially% FVC > 70% and/or area of ILD based on CT scan was <20% of the entire lung field.

Results: Nineteen patients with SSc were newly visited. Male to female ratio was 4:15, mean age 63 ± 13 years, and mean disease duration was 138 ± 109 months. Ratio of limited skin type was 79%. 37% were with ILD and 11% were with PAH confirmed by RHC. There is no correlation between TRPG at rest and the density of capillaries. However, there is a significant, negative correlation between TRPG at exercise and the capillary density ($r = 0.57$, $P < 0.02$). Furthermore, the same result was observed even with seventeen "pre-PAH" cases ($r = 0.67$, $P < 0.04$).

Conclusion: Our study suggests that the finding of NVC, especially the extent of capillary loss, is correlated with disease process of PAH, even subclinically, and may predict the progression to PAH in SSc. Since NVC is non-invasive, NVC can detect earlier PAH candidates and contribute to the improvement of prognosis.

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AB0635

POTENTIAL BIOMARKERS OF SKIN CHANGES IN SYSTEMIC SCLEROSIS

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Background: Skin fibrosis is a hallmark of systemic sclerosis (SSc). There are no widely accepted biomarkers of skin involvement in this condition. Several serum or plasma markers have been studied in patients with SSc - monocyte chemoattractant protein-1 (MCP-1), chemokine (C-X-C motif) ligand 8 (CXCL8), interleukin-13 (IL-13), and some more recognized such as - platelet derived growth factor (PDGF), transforming growth factor-beta 1 (TGF-beta 1), epidermal growth factor (EGF) and basic fibroblast growth factor (bFGF).

Objectives: The aim of this study was to assess several circulating biomarkers which may be relevant to the fibrosing process and further to correlate the obtained data with clinical indicators specific for SSc skin involvement.

Methods: 59 SSc patients (M/F 9/50; mean age 52.1 years, mean disease duration 6.7 years, 36 patients with limited cutaneous SSc and 23 with diffuse cutaneous SSc. As a control group 36 healthy individuals matched to sex and age were examined. Serum concentrations of bFGF, granulocyte-colony stimulating factor (G-CSF), granulocyte-macrophage-colony stimulating factor (GM-CSF), MCP-1, PDGF, IL-8 and 13 were analysed using commercial multiplex kit. The following clinical examinations were performed: modified Rodnan skin score (mRSS), Hand Mobility in Scleroderma Test (assessing hand function) (HAMIS), Cochin Hand Function Scale (hand function) (CHFS), Delta Finger-to-Palm Distance (extension-flexion) (dFTP), Inter-lip Distance (inter-lip), Inter-incisor Distance (inter-incisor), and Mouth Handicap in Systemic Sclerosis Scale (mouth opening) (MHISS). For statistical evaluation Spearman's correlation coefficient was used.

Results: When compared with healthy controls serum concentrations of bFGF ($p<0.001$), G-CSF ($p<0.0001$), GM-CSF ($p<0.0001$), MCP-1 ($p<0.0001$) IL-8 ($p<0.0001$), and IL-13 ($p<0.001$) were significantly elevated in SSc cohort. PDGF levels were increased in SSc patients with only a lower significance ($p<0.01$). bFGF, G-CSF, MCP-1 and IL-8 levels correlated significantly ($p<0.05$) with mRSS and HAMIS. GM-CSF levels correlated with mRSS and HAMIS and there was only a trend for negative correlation with inter-incisor. The was no correlation of IL-13 and PDGF levels with the evaluated clinical data.

Conclusion: Our results have shown that G-CSF, GM-CSF and IL-8 play a substantial role in SSc fibrosing process. Potential biomarkers as bFGF, G-CSF, MCP-1 and IL-8 correlated with a few clinical indices of SSc skin involvement.

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AB0636

INFLUENCE OF CARDIOVASCULAR RISK FACTORS ON NAILFOLD VIDEOCAPILLAROSCOPY IN THE STUDY OF CONNECTIVE TISSUE DISEASES

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Background: Nailfold videocapillaroscopy is a non-invasive technique used to assess Raynaud syndrome. It is mainly used for the early diagnosis of connective tissue disorders (CTD) such as systemic sclerosis. There is some evidence that capillaroscopy findings may be altered by microcirculation abnormalities in patients with cardiovascular risk factors (CVRF).

Objectives: to analyze the influence of cardiovascular risk factors on nailfold capillaroscopy in patients with Raynaud or suspect of CTD.

Methods: An observational and descriptive study of consecutive patients that underwent a videocapillaroscopy examination for the study of Raynaud syndrome was conducted. A "Capiscope" model videocapillaroscope from Optilia was used, with a fixed magnification of 200x. Examination was made on 8 hand fingers, with 2 images per finger. The patients had to be at least 30 minutes in a fixed warm temperature room and without smoking 1 hour before the performance of the test. The following capillaroscopic parameters were considered: nailfold morphology, capillary loop enlargements, megacapillaries, microhaemorrhages, avascular areas and signs of neoangiogenesis. Demographic information (including age, gender and previous diagnosis) and cardiovascular risk factors (including arterial hypertension (HT), diabetes mellitus (DM), dyslipidemia (DL) and smoking habit), were collected. The influence of cardiovascular risk factors on nailfold capillaroscopy was analyzed, using univariate and multivariate logistic regression models, adjusted for possible confounders.

Results: Out of the 136 included patients, 91% were women. Mean age was 54.6 ± 18.7 years. Raynaud syndrome was reported in 83% patients, with a mean duration of 6.1 ± 5.7 years and 12% of the patients had a previous diagnosis of CTD, including systemic lupus erythematosus (5%), systemic sclerosis (4%), undifferentiated connective tissue disorder (2%) and mixed connective tissue disease (1%). Regarding CVRF, HT was observed in 25%, DM in 7%, DL in 23% and past or current smoking habit in 32%. Capillaroscopic findings were: loop enlargements (81%), megacapillaries (30%), microhaemorrhages (46%), signs of neoangiogenesis (71%) and avascular areas (20%).

Regarding the capillaroscopic pattern, 46% presented a normal or non-specific pattern; 31% a microangiopathy pattern and 23% a scleroderma pattern (of which 58% had an early or active scleroderma pattern and 42% a late scleroderma pattern). A new diagnosis of CTD was made in 24 patients (18% of the cohort). In the group of patients without CTD, HT was associated with microhaemorrhages ($p = 0.02$) and avascular