diagnosis, frequency of follow-ups and therapy of SSc in different age groups.

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THE IMBALANCE OF ACTIVITIES OF PURINE AND PYRIMIDINE METABOLISM ENZYMES IN RED BLOOD CELLS OF SYSTEMIC SCLERODERMA PATIENTS


Background: The systemic sclerosis (SSc) is a rare connective tissue disease of unknown cause. The key points in development of SSc are the increased number of high collagen-producing fibroblasts in the skin, endothelial dysfunction and immune activation. Recent studies report various metabolic disturbances in SSc patients. Purine and pyrimidine metabolic pathways are under the central processes of cellular life. The changes in activity of purine and pyrimidine metabolism enzymes in blood plasma and lysed lymphocytes depending on the SSc activity were described by us earlier [1]. At the same time, there are publications that confirm the relationship between changes in ADA activity in red blood cells and pronounced immune disorders [2].

Objectives: to characterize enzymatic patterns of the major purine and pyrimidine metabolic pathways enzymes in lysed red blood cells depending on the SSc activity.

Methods: 51 SSc patients and 30 healthy controls were enrolled in the study. The diagnosis was verified in accordance with the international standards (ACR/EULAR 2013). Disease activity was assessed in accordance with the national classification [3]. Adenosine deaminase (ADA; EC 3.5.4.4); adenosine kinase (AK; EC 2.7.1.20); guanylate kinase (GK; EC 2.7.4.8); dihydroorotate dehydrogenase (DODH; DC 1.3.1.14); IMP dehydrogenase (IMPDH; EC 1.1.1.205); purine nucleoside phosphorylase (PNP; EC 2.4.2.1); thymidine kinase (TK; EC 2.7.1.21); thymidine phosphorylase (TP; EC 2.4.2.4); uridine/thymidine dehydrogenase (UDH; EC 1.17.99.4); cytidine deaminase (CDA; EC 3.5.4.5) activities were measured in lysed red blood cells.

Results: Mean age of patients (Means: SD) was 42.8 ± 13.3 years, mean SSc duration was 7.9 ± 0.7 years. We revealed substantial changes in enzymatic activities related to both purine and pyrimidine metabolism in lysed red blood cells of SSc patients. The increased ADA (p < 0.001), AK (p < 0.001), IMPDH (p < 0.001), TK (p < 0.001), UDH (p < 0.001) activities and the decreased DODH (p < 0.001), PNP (p < 0.001) activities in lysed red blood cells were observed of SSc patients in comparison with healthy controls. AK, IMPDH, TK, UDH activities positively correlated with SSc activity. Negative correlations with SSc activity were revealed for ADA, CDA, DODH, GK, PNP, TP activities.

Conclusion: The progression of SSc goes with the imbalance of the purine and pyrimidine enzymes in a regular manner. Activity of the auto-immune inflammation is the factor that underlies the enzymatic pattern of purine and pyrimidine metabolism.

REFERENCES:

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ASSOCIATION OF EUROPEAN SCLERODERMA STUDY GROUP ACTIVITY INDEX (ESCSG-AI) WITH PROGRESSION PULMONARY ALTERATIONS BY HRCT IN PATIENTS WITH SYSTEMIC SCLEROSIS OVER A FIVE YEAR PERIOD

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Background: Systemic sclerosis (SSc) is a rare connective tissue disease with a heterogeneous clinical course. Interstitial lung disease (ILD) is a common manifestation of SSc and a leading cause of death. Patients with early active SSc are at great risk for progressive ILD.

Objectives: To assess the ESCSg-AI in patients with systemic sclerosis (SSc) and interstitial lung disease over a five year period.

Methods: It was a longitudinal study involving 77 pts with SSc-ILD (mean age was 46.2±13.4; 69% have limited subset of the disease; 93% were female). The mean duration of follow up was 58.9±11.4 months. Pts, were investigated with HRCT twice (at first visit and at the end of the study) and according the CT-changes were divided into 3 groups: the 1st group (16 pts) with improvement; 2nd group (39 pts) without any changes and 3rd group (22 pts) with worsening of fibrosis. Disease activity was assessed by the 2001 European Scleroderma Study Group Activity Index (ESCSG-AI).

Results: There were no significant differences between groups related to sex, frequency of diffuse form and duration disease. Mean dates of ESCSg-AI score of all pts and in the each groups in first visit and the end of follow up are present in table 1.

Table 1

<table>
<thead>
<tr>
<th>Groups</th>
<th>First visit</th>
<th>At the end of the study</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group 1 (n=16)</td>
<td>1.9 ± 1.8</td>
<td>1.7 ± 0.9</td>
<td>P &gt; 0.05</td>
</tr>
<tr>
<td>Group 2 (n=39)</td>
<td>2.0 ± 1.5</td>
<td>2.1 ± 1.3</td>
<td>P &gt; 0.05</td>
</tr>
<tr>
<td>Group 3 (n=22)</td>
<td>2.4 ± 1.5</td>
<td>3.25 ± 2.0</td>
<td>P &lt; 0.05</td>
</tr>
<tr>
<td>All pts (n=77)</td>
<td>2.2 ± 1.6</td>
<td>1.9 ± 1.8</td>
<td>P &gt; 0.05</td>
</tr>
</tbody>
</table>

After 5 years of follow up mean values of ESCSg-AI score increased significantly in group 3 and was more than 3, this actually means the activity of the disease. The mean values of ESCSg-AI score in group 3