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AB0220

ALPHA- SMOOTH MUSCLE ACTIN EXPRESSION ON ENDOTHELIAL PROGENITORS CELLS OF SYSTEMIC SCLEROSIS PATIENTS: POSSIBLE ROLE IN THE ENDOTHELIAL-TO-MESENCHYMAL TRANSITION PROCESS

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Background: Endothelial-to-mesenchymal transition (EndoMT), a newly recognized type of cellular transdifferentiation, seems to be involved in Systemic Sclerosis (SSc) pathogenesis. In this process endothelial cells lose their specific markers, and acquire a mesenchymal phenotype, thus expressing cell products such as alpha smooth muscle actin (α -SMA) (1,2). Circulating endothelial progenitors cells (EPCs) derive from bone marrow stem cells and contribute to *de novo* vessels formation. Several studies, although with conflicting results, have shown that EPCs in the peripheral blood of patients with SSc are impaired in their number and function (3).

Objectives: to assess the expression of α -SMA, possibly associated with a pro-mesenchymal switch (EndoMT) of the circulating Early (CD34+KDR+CD 133+) and Late (CD34+KDR+) EPCs in the peripheral blood of SSc patients and of patients with Very Early Diagnosis of SSc (VEDOSS) compared with healthy controls (HC) using flow cytometry.

Methods: we enrolled 11 patients (7 SSc and 4 VEDOSS), classified according to the classification criteria for SSc (4) and for VEDOSS not fulfilling SSc criteria (5), and 10 HC. Phenotypic characterization was performed as previously described by Vasa et al. using a FACS Calibur (BD Immunocytometry Systems). EPCs number was expressed as a percentage of cells within the lymphocyte gate.

Results: we found a significantly higher percentage of α -SMA positive Early EPCs (CD34+KDR+CD 133+ α -SMA+) in all patients respect to HC (0,06% $\pm$ 0,03 vs 0,03% $\pm$ 0,01; $p=0,0149$) particularly in VEDOSS patients (0,07% $\pm$ 0,01 vs 0,03 $\pm$ 0,01 $p=0,008$). Moreover, in VEDOSS patients, also the percentage of Early EPCs (CD34+KDR+CD 133+), Late EPCs (CD34+KDR+) and α -SMA positive Late EPCs (CD34+KDR+ α -SMA+) were significantly higher than in HC (0,05% $\pm$ 0,01 vs 0,03% $\pm$ 0,01 $p=0,05$; 0,07% $\pm$ 0,01 vs 0,04% $\pm$ 0,02 $p=0,04$; 0,06% $\pm$ 0,01 vs 0,04% $\pm$ 0,02 $p=0,05$). Besides Early EPCs and α -SMA positive Early EPCs percentages seem to be significantly reduced in patients taking iloprost ($p=0,05$ and $p=0,01$ respectively), calcium channel blockers (CCB) ($p=0,05$ and $p=0,03$) and DMARDs ($p=0,017$ and $p=0,013$).

Conclusion: EndoMT seems to be involved in the pathogenesis of SSc and circulating EPCs seem to be impaired in number and function in SSc patients. In our study we found higher levels of EPCs, in particular α -SMA positive Early EPCs in both groups of patients (SSc and VEDOSS) respect to HC. Thus we can hypothesize a predominant pro-mesenchymal phenotype of this kind of EPCs. This could be considered the expression of the involvement of EPCs in EndoMT process and could better explain the controversial role of EPCs in SSc pathogenesis. Very interesting is the finding of a lower percentage of Early EPCs, and in particular of α -SMA positive Early EPCs, in those patients taking iloprost, CCB and DMARDs, suggesting a potential effect of these drugs on this subgroup of EPCs.

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AB0221

SEQUENCE OF CLINICAL SYMPTOMS ONSET AND ITS CORRELATION TO THE AUTOANTIBODIES PRESENCE IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Idiopathic inflammatory myopathies (IIM) is a heterogeneous group of autoimmune diseases with a broad spectrum of clinical presentations including musculoskeletal, respiratory, neurological, dermatological, gastrointestinal and cardiovascular symptoms. It is known that presence of autoantibodies is a predictor of disease course. However, there is very limited data summarizing long-term observations assessing, when the particular symptoms are expected to occur.

Objectives: To assess the development of the symptoms in IIM patients during the disease course with respect to their serology patterns.

Methods: Medical history of IIM patients treated in our clinic between 2008 and 2019 were reviewed. We excluded patients with: insufficient data for the first period of disease course, history of IIM <6 months, IIM patients who do not fulfill Peter & Bohan 1975 diagnostic criteria (less than 4 points). Finally 103 IIM patients medical history were included for this analysis. EUROLINE test containing 16 myositis specific or associated antibodies (MSAs, MAAs) was used to determine serology for all of those patients. Onsets of symptoms were noted only in case when symptoms were confirmed objectively. Mean time since onset (MTSO) in months was counted for each symptom.

Results: Numbers below are presenting MTSO in months for each symptom.

Anti-Jo1 antibodies positive patients (n:30) have presented: rash (0), mechanic hands (1.2), myocarditis (5), muscle weakness (10.3), Raynaud's phenomenon (RP) (10.4), arthritis (20.7), interstitial lung disease (ILD) (23.5), fever (29.1), Gottron's sign (31.3), muscle pain (46.2), dysphagia (53.4).

Anti-Mi-2 antibodies positive patients (n:12) have presented: RP (0), mechanic hands (0), rash (0.2), Gottron's sign (0.3), fever (2.5), muscle weakness (4.5), dysphagia (4.5), muscle pain (9), ILD (15.5).

Anti-PM-Scl antibodies positive patients (n:7) have presented: rash (1), fever (1), Gottron's sign (1.25), mechanic hands (4), ILD (4), RP (5), arthritis (10.2), muscle weakness (46.3), muscle pain (108.5).

Anti-SRP antibodies positive patients (n:5) have presented: muscle weakness, fever, arthritis and ILD symptoms simultaneously (0), then dysphagia (4) and muscle pain (10).

Anti-NXP2 antibodies positive patients (n:4) have presented: muscle weakness and pain simultaneously with dysphagia (0), ILD developed 4 months later.

Anti-MDA5 antibodies positive patients (n:2) have presented: arthritis (0), fever (0.5), muscle pain (2), rash (3).

Anti-Ro52 antibodies positive patients (n:12) have presented: RP (3), Gottron's sign (3.3), rash (3.5), arthritis (5), muscle weakness (6.2), muscle pain (11), myocarditis (11), ILD (12), mechanic hands (13), dysphagia (18.3).

MSAs and MAAs negative patients (n:21) have presented: fever (0), RP (0), dysphagia (0.3), muscle pain (0.4), muscle weakness (1.9), rash (20), arthritis (66).

Conclusion: Our study shows differences in disease course in IIM patients with each serology pattern adding new data on chronology of appearances of each symptom, which is important in predicting the future course of the disease and planning long-term management.

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