DO WE KNOW ALL RISK FACTORS OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION?

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Background:
A late complication of pulmonary thromboembolism is chronic thromboembolic pulmonary hypertension (CTEPH) - characterized by chronic pulmonary occlusion and vascular remodeling. Current data suggest that CTEPH does not result from traditional, known thrombophilia or defective plasma fibrinolysis but antiphospholipid antibodies (APA) and the coagulation factor VIII have been found in a significant proportion of CTEPH patients in a majority of studies. Pathogenesis of CTEPH has not been completely elucidated and multiple risk factors predisposing to its development remain unexplained. The aim of our prospective study was to analyze well-expected hemostatic risk factors and potential hemostatic abnormalities in CTEPH population.
Materials and methods:
Following hemostatic parameters were investigated: platelet count, mean platelet volume (MVA) spontaneous platelet aggregation (SPA), von Willebrand factor antigen (vWF:Ag), von Willebrand factor activity (vWF:Ac), plasminogen activator inhibitor (PAI-1), fibrinogen, factor VIII (fVIII) and APA. Analyzed were 36 patients with confirmed CTEPH and compared with 32 healthy subjects as a control group.
Results:
In CTEPH compared to controls was observed: a significant decrease of platelet count [216±56 *vs* 261±60.10^9/L, P<0.01], higher MVA [11.2±0.9 *vs* 9.9±0.7fL, P<0.001], and higher SPA [10.9±4.3 *vs* 8.4±6.2%, P<0.05]. No changes of platelet aggregation induced by low concentrations of epinephrine (10/1/0.5µM) and ADP (2/1/0.05µM) in CTEPH were found against controls. These changes were accompanied by the increase of vWF:Ag [186.5±23 *vs* 124±36.4%, P<0,001], vWF:Ac [184.2±28,6 *vs* 122±37.3%, P<0.001], PAI-1 [2.8±1.6 *vs* 2±1.4U/mL, P=0.05], fibrinogen [3.8±0.8 *vs* 3.1±0.6g/L, P<0.001] and fVIII [158.2±30.7 *vs* 129.6±31.5%, P<0.001] level in plasma. Contrary to other studies, APA were not increased.
Conclusions:
In this study were analyzed multiple established hematologic risk factors in CTEPH patients, which are in agreement with current studies. Investigated were though also some extraordinarily risk factors like platelet abnormalities and vWF:Ac/Ag. Interestingly, the increased SPA was shown in patients with CTEPH, which could be a possible predictive risk marker and a novel way in the pathogenesis of CTEPH.