ENDOTHEL INJURY IN ACUTE THROMBOTIC THROMBOCYTOPENIC PUPURA AND ITS CONNECTIONS WITH THE ACTIVATION OF THE COMPLEMENT SYSTEM

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Introduction: thrombotic microangiopathies (TMA) such as thrombotic thrombocytopenic pupura (TTP) are severe, episodic and rare diseases. In TTP, deficiency of the von Willenbrand cleaving protease (ADAMTS-13) as the main risk factor is well known, however, the mechanisms how the exact triggers exacerbate disease, are currently unknown. In previous studies our group showed the potential role of the neutrophil and complement activation in TTP. Although many studies described injury of the endothelium in TMA but there is no detailed information about its connections with innate immunity in TTP.

Our aim was to examine the role of the endothelial injury in TTP (acute, remission) in a clinical study. We also analyzed whether there is any connection between endothel injury and the activation of the neutrophil and complement activation.

Methods: in our study we enrolled 39 patients (33 female, mean age 43.7) and 57 healthy controls (30 female mean age 36.3). The diagnosis of the TTP was set up with low thrombocyte number (<150G/L), intravascular hemolysis and the presence of the fragmentocytes. We measured a precursor fragment of the ET-1 (Endothelin-1, CT-proET-1) as endothel injury marker with BRAHMS LIA, Kryptor system in plasma. We used non-parametric tests for statistical analysis (t-test, ANOVA and Spearman correlation coefficient).

Results: ET-1 levels were significantly higher in acute TTP (mean 63.4 pmol/L, SD 30.1) as compared to the control group (31.2, 14.8) and to the group of TTP patients in remission (45.9, 17.4; $p<0.0001$, ANOVA). There was a positive correlation between the ET-1 levels and levels of terminal pathway activation marker (SC5b-9). No further correlations were observed with other activation markers (such as C3a and Bb).

Discussion: our results indicate the presence of endothel injury in patients with acute TTP as compared to the TTP in remission and healthy controls and this activation is parallel to activation of the complement system. Complement and endothel activation could potentially contribute to the development of acute episodes in subjects with predisposition to TTP.

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