



Von – Willebrand factor, a possible marker for disease activity in vasculitis

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Background: Vasculitis is an inflammation of blood vessels. While in many cases markers of inflammation as c- reactive protein (crp) or erythrocytes sedimentation rate (ESR) may correlate with disease activity, in some cases inflammation markers are normal and evaluation of disease activity is based solely on clinical examination or imaging.

Von – Willebrand factor (vWF) is a large multimeric glycoprotein, present in blood plasma. It is produced constitutively as ultra-large VWF in endothelium and subendothelial connective tissue and is packaged into storage organelles, the Weibel–Palade bodies (WPBs) on the surface of the endothelium. Damaged endothelium releases vWF. Sporadic reports have suggested vWF to be elevated in vasculitis.

Aim: To evaluate vWF concentration in peripheral blood in patients with vasculitis as a marker of disease activity

Methods: Patients with systemic vasculitis according to Chapel hill consensus criteria were evaluated. Demographic characteristics and clinical manifestations were assessed by frontal interview, physical examination and by medical records. Disease activity was reported by BVAS score. VWF was recorded at diagnosis and where available after treatment. vWF was reported in % of predicted value and was adjusted to blood group (vWF level in patients with blood group o is normally reduced by 25% in compare to other blood groups)

Results: 25 patients with systemic vasculitis were compared to 15 healthy controls. There was no statistical difference between the two groups regarding age, 56 ± 17 years vs. 48 ± 16 years or gender 56% vs. 50% ($P=0.54$). 40% had ANCA associated vasculitis, 20% had Giant cell arteritis, 16% had polyarteritis nodosa, 8% had Takayasu and the rest had other vasculitides Time from diagnosis was $3 \pm 4.8.3$. Mean vWF was higher in patients with active vasculitis compared to patients in remission and to healthy controls, $212\% \pm 81$ vs. $159\% \pm 80$ vs. $117\% \pm 35$ $p < 0.001$. In 10 patients with active vasculitis, who had a follow up exam after treatment there was a significant decrease of vWF from $231\% \pm 61$ to $169\% \pm 53$ $p < 0.02$. There was a direct

correlation between vWF and BVAS score $r=0.31$, $p=0.04$. There was no statistically significant correlation between CRP or ESR to BVAS score, yet CRP correlated directly with vWF $r=0.32$ $p=0.07$. In 4 patients who were treated with IL-6 inhibitors, vWF was elevated in spite of normal CRP levels, while clinically vasculitis was active.

Conclusions: This study demonstrates that vWF may be a possible marker for disease activity in patients with vasculitis. As vWF reflects direct damage to endothelium and is not dependent on the IL-6 axis it is not confounded using IL-6 blockers, that are widely used in vasculitis these days. Further studies should be undertaken to validate these preliminary findings.