von Willebrand factor antigen in giant cell arteritis

Sir: We were surprised at Dr Nordborg and colleagues' frequent negative appraisal of their findings in following von Willebrand factor (vWF) and plasminogen activator inhibitor levels in a large cohort of patients with giant cell arteritis. Although showing that vWF levels are not useful in monitoring clinical relapse, they did establish that vWF levels gradually fall to control values always 18–24 months, which correlates with the natural history of the disease in most patients. This is an important observation as several studies have concluded that vWF is raised but not useful in assessing clinical activity and have noted a decrease in vWF after successful corticosteroid treatment. 1–4

The logical conclusion from Dr Nordborg's data is that vWF levels reflect not only vascular injury but the activity of the fundamental disease process of giant cell arteritis. It also supports the argument that steroid treatment, although mitigating vascular injury progressing to thrombotic/embolic occlusion, has little effect on the underlying disease. In this study 'nine of 63 patients still receiving corticosteroids had a . . . vascular occlusive episode'. Moreover, the use of vWF as a marker obscures its pathogenic potential via its main function, the initiation of platelet adhesion. vWF release at the site of vascular injury is the vital first step in the thrombotic cascade, which even when it does not lead to occlusion will perpetuate vascular damage through elaboration of platelet-derived vasoactive substances and growth factors. There is a strong correlation between vWF levels and such growth factors after myocardial infarction. 5 Of further pathogenic relevance is the close relation between vWF release and expression of the endothelial cell polymorph adhesion molecule GMP-140, which occurs because both are stored in Weibel-Palade bodies. Thus the principal implication is that in relapsing cases or those unresponsive to steroids there should be a low threshold for using agents which could modulate or modify either vascular injury or the disease process and that vWF responses might provide clues to the most effective drugs and their optimal usage. von Willebrand factor responses may also help to identify new immunomodulators. Indeed, given that vWF release is Ca2+-dependent, it is tempting to speculate about the therapeutic effect of calcium channel blockers in giant cell arteritis and other vasculitides.
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