Vasculitis and the antiphospholipid syndrome

Sir, In a recent letter, Handa et al. [1] mention that little information is available on the possible association of vasculitis and the antiphospholipid syndrome (APS). This would be true if a search were made only of papers that include both topics in their title, such as those included in their references. However, we have previously provided considerably more information in support of this association. Thus, in our original description of the primary APS we included patients who had episodes of vasculitis preceding occlusion of larger vessels [2] and, in our description of the antiphospholipid arterial vasculopathy, we proposed that the occurrence of vasculitis in our three patients could have triggered the reaction of antiphospholipid antibodies with the negatively charged phospholipids flipped to the outer leaflet of endothelial cells by damage to these cells [3]. We now know that beta-2-glycoprotein-1 readily binds to these phospholipids as they appear on the surface of endothelial cells, and antiphospholipid antibodies bind to it thereafter.

Our contributions to knowledge on the association of vasculitis with APS have been more than merely anecdotal. Thus, in our study of 667 patients with systemic lupus erythematosus, in which we proposed preliminary criteria for the classification of APS in this disease [4], we found a significant association of cutaneous vasculitis with definite as compared with negative APS, with an odds ratio of 2.0 and a 95% confidence interval of 1.2–3.5 ($P = 0.007$).

Also, in our study of vasculitis in systemic lupus erythematosus (SLE) [5], we found an association of clinical vasculitis with APS in lupus patients, an
association that became stronger when we considered only those patients in whom vasculitis had been proven. In patients with SLE with visceral vasculitis, but who had no cutaneous vasculitis, univariate logistic regression analysis showed that the visceral vasculitis was associated with vascular occlusions, particularly when they were arterial, with thrombocytopenia, with transient ischaemic attacks and with APS proper.

Recognizing that vasculitis does associate with APS is a necessary first step on the road towards the understanding of why and how it occurs.

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Accepted 24 January 2000

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