Regarding the diagnosis of APS, as Kaburaki et al (6) discuss in their letter in this issue of the journal, thrombocytopenia is too controversial to be included as one item of criteria and confusion in the definition of APS will follow.

Asherson defined APS patients without any underlying diseases as cases of primary APS (7), and when patients have some factors of SLE (less than 4 items of SLE criteria), he termed them as secondary APS to lupus-like disease. Here however, “lupus-like disease” should be clearly defined. Many individuals, including apparently healthy ones, carry one or two factors related to SLE criteria, but we usually do not classify them as patients with lupus-like disease. We suggest that use of “APS secondary to lupus-like disease” be avoided and that a new more appropriate criteria of APS and primary APS be designed.

This issue will be discussed in the APS criteria session at the 5th International Symposium on Antiphospholipid Antibody to be held October 6–9, 1998 in Sapporo, Japan.

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References


Thrombocytopenia in the Criteria for Anti-Phospholipid Antibody Syndrome

Dr. Funauchi’s reply:

To the Editor: Thrombocytopenia in the criteria of anti-phospholipid antibody syndrome (APS) poses problems of both diagnosis and differentiation (1, 2) as pointed out by Kaburaki et al (3). From the viewpoint of sensitivity, thrombocytopenia should be included in the criteria.

In our retrospective study (4), 7 out of 27 patients who had been once diagnosed as having idiopathic thrombocytopenic purpura (ITP) showed a prolonged activated partial thromboplastin time (aPTT) and positive anti-phospholipid antibodies (lupus anti-coagulant, β2 GPI-dependent anti-cardiolipin (aCL), or biologically false positivity for syphilis). However, only three of the above 7 patients later presented remarkable symptoms and signs of APS such as cerebral thrombosis or habitual abortion. During the clinical course, specific clinical findings of 2 of the rest of the patients were absent and those of another 2 patients (thrombophlebitis) were too mild to be suspectable of APS unless they were positive for serum anti-phospholipid antibodies. There are two possible reasons for the difficulty of the diagnosis of APS: 1) the existence of subclinical APS, and 2) the influence of the treatment of ITP with adreno-corticosteroid hormones. Therefore, the detection of the serum anti-phospholipid antibodies was thought essential for the screening of the APS.

Thrombocytopenia is sometimes observed in APS (5) and it has been regarded as one of the criteria in the diagnosis of APS (1, 6). As Atsumi et al describes (7), there are several mechanisms which cause thrombocytopenia in APS; consumption of the platelets at the site of thrombosis and destruction of the platelets by hemophagocytic systems. Since there may be cases without typical symptoms or signs of APS regardless of a positive test for the serum anti-phospholipid antibodies, it is thought important that thrombocytopenia is included in the criteria as a clue to find APS.

In our previous study (4), 5 out of 7 patients (71.4%), who were positive for any of the serum anti-phospholipid antibodies or had symptoms and signs of APS, had a positive test for serum aCL as determined by enzyme-linked immunosorbent assay (ELISA). Among the 5 patients with a positive test for aCL, 3 patients (60%) were positive for antinuclear antibody. This is in accordance with the data presented by Kaburaki et al (3). These patients might belong to the group of patients with the lupus-like disease described by Asherson and Cervera (8). It is known that systemic lupus erythematosus (SLE) is frequently complicated by both ITP and APS, and that ITP is also complicated by SLE. Furthermore, thrombocytopenia can occur in any of these 3 disorders. Since thrombocytopenia in the criteria apparently lowers the specificity of APS, discrimination of APS and ITP by detection of anti-phospholipid antibodies and analysis of the epitopes of the anti-platelet antibodies (9–12) is important to diagnose APS and subsequently to prevent the irreversible lesions such as infarction of various organs. In any case, a new criteria for APS is needed for the discrimination of these borderline diseases.

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