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Independent validation of the antiphospholipid score for the diagnosis of antiphospholipid syndrome

Savino Sciascia1,2, Maria Laura Bertolaccini1, Dario Roccatello2, Munther A Khamashta1,3

1Lupus Research Unit, The Rayne Institute, Division of Women’s Health, King’s College, London, UK
2Centro di Ricerche di Immunopatologia e Documentazione su Malattie Rare (CMID), Università di Torino, Torino, Italy
3Louise Coote Lupus Unit, Guy’s and St Thomas’ NHS Foundation Trust, St Thomas’ Hospital, London, UK

Antiphospholipid syndrome (APS) is a heterogeneous entity with a wide variation in clinical course and laboratory profile. It is accepted that the presence of antiphospholipid antibodies (aPL) confers a higher risk for both thrombosis and pregnancy morbidity, but quantifying such a risk is still a challenge. As a consequence, clinicians are unable to tailor the treatment according to the risk.

Recently, Otomo et al.1 developed and validated the so-called ‘antiphospholipid score’ (aPL-S) by testing multiple aPL and evaluating the aPL-S efficacy for the diagnosis of APS and predictive value for thrombosis. This score was shown to be a useful quantitative index for diagnosing APS and to be valuable as a predictive marker for thrombosis in autoimmune diseases.

In order to independently validate the aPL-S, we applied the proposed score system to a cohort of 211 consecutive patients who attended the Louise Coote Lupus Unit (St Thomas Hospital, London, UK). All the patients fulfilled the 1982 criteria for systemic lupus erythematosus (SLE).2 Overall, 81 patients fulfilled criteria for APS 1 and 73 patients had a history of thrombosis (48 arterial, 41 venous thrombosis). Out of 144 women who had ever been pregnant, 41 had a history of miscarriages and 34 a history of fetal death. To validate the aPL-S, we adapted the proposed score system using our inhouse cut-off values for aPL testing as previously reported or according to the current guidelines, as appropriate.3–8 aPL profile included anticardiolipin antibodies, lupus anticoagulant by partial thromboplastin time (aPTT – IL-test APTT-SP, Instrumentation Laboratory, Milan, Italy) and dilute Russell viper venom time, anti-β2glycoprotein-I antibody, and antibodies to phosphatidylserine–prothrombin complex.

aPL-S was calculated for each patient by adding together the points corresponding to the risk factors as described.1 Higher values of aPL-S were seen in patients who experienced thrombosis and/or pregnancy loss when compared with those without clinical events (median 17 (0–86) vs 4 (0–31), p<0.001). When analyzing clinical subgroups, patients who experienced thrombosis or pregnancy loss showed higher aPL-S compared with those without clinical events (median 18 (0–86) vs 4 (0–27), p<0.001 for thrombosis; 7 (0–69) vs 3 (0–29), p=0.029 for pregnancy loss) (figure 1). In our cohort, when the cut-off level for the aPL-S was defined as 30, as per the original study by Otomo et al.,1 the sensitivity and specificity of the aPL-S were 39% and 95%, respectively, compared with 37% and 96% shown in the Japanese cohort. The positive predictive value of an aPL-S ≥30 was 36%, whereas the negative predictive value was 91%.

We demonstrated that the aPL profile can be successfully quantified by the aPL-S in an independent cohort of SLE patients. The aPL-S correlated with a history of thrombosis or pregnancy loss in our cohort, suggesting that the aPL-S is a suitable quantitative marker of APS.

References


Figure 1

Antiphospholipid score (aPL-S) in systemic lupus erythematosus (SLE). The aPL-S was calculated according to Otomo et al. Data are shown as box plots, where each box represents the 25th to 75th percentiles: lines inside the box represent the median. The whiskers represent the 95% CI. Higher values of aPL-S were seen in patients who experienced thrombosis and/or pregnancy loss (PL) when compared with those without clinical events (p<0.001 by Mann–Whitney U test). When analysed separately, patients who experienced thrombosis or PL showed higher aPL-S when compared with those without clinical events.