An acute multiorgan thrombotic disorder associated with antiphospholipid antibodies; two 'catastrophic' cases

Over the past decade the antiphospholipid syndrome (APS) was defined by the presence of antiphospholipid antibodies (aPL) and clinical manifestations including thrombosis, recurrent fetal loss, arterial and/or venous thrombosis, livedo reticularis, heart valve lesions, and renal involvement.1 Asherson et al first drew attention to a catastrophic variant of APS (CAPS) that is characterised by multiple widespread thromboses leading to multiple organ failure and often death.2 We describe two non-systemic lupus erythematosus (SLE) patients with a strikingly similar clinical presentation of CAPS and emphasise the difficulties in differentiating CAPS from other thrombotic angiopathies.

Case reports

CASE REPORT ONE

A 20 year old woman presented with transient hemichorea in 1993. Computed tomography of the brain was normal. From June 1995, episodes of hemichorea recurred together with severe frontal headaches. In November 1995 she was admitted to our hospital with rapid deterioration of vision and behavioural changes.

Physical examination showed subcutaneous, blood pressure 150/115 mm Hg, livedo reticularis, heart valve lesions, and renal involvement.1 Asherson et al first drew attention to a catastrophic variant of APS (CAPS) that is characterised by multiple widespread thromboses leading to multiple organ failure and often death.2 We describe two non-systemic lupus erythematosus (SLE) patients with a strikingly similar clinical presentation of CAPS and emphasise the difficulties in differentiating CAPS from other thrombotic angiopathies.

Laboratory findings included: platelet count 47 × 10^11 l, creatinine 176 µmol/l, fibrin degradation products 1.0 mg/l (normal <0.5), fibrinogen 4.0 g/l (normal range 2.0-4.0), haptoglobin 0.2 g/l (0.3-1.8), reticulocytes 62% (7-30), positive direct Coombs test, microscopic haematuria and proteinuria (3.9 g/day). Both Lupus anticoagulant (LAC; DRVTT; IL Test LAC;Cor-Gen and LAC-Test. Instrumentation Laboratory, Milan, Italy) and high titre IgG and IgM anticardiolipin antibodies (aCL) were present; ANA were negative. Echocardiography showed mitral regurgitation–stenosis and no vegetations. Brain magnetic resonance imaging showed multiple ischaemic infarctions. A skin biopsy specimen from a livedo-reticularis lesion showed thrombotic occlusions of arteries and venules with partial recanalisation.

Treatment consisted of high dose corticosteroids, plasma exchange, anticoagulation, cyclophosphamide (1000 mg intravenous), and platelet transfusions. After three weeks she gradually regained consciousness and renal