To the Editors:

Antiphospholipid syndrome in a man presenting with cold autoimmune haemolytic anaemia

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Ceylon Medical Journal 2015; **60**: 71-2

Antiphospholipid antibody syndrome (APS) is an auto antibody mediated acquired thrombophilic state characterised by arterial or venous thrombosis and several hematological manifestations. We report a middle aged man with progressive anaemia, dementia and a stroke who was found to have primary APS and cold auto-immune haemolytic anaemia (AIHA).

A 54-year old man presented with progressive dyspnoea and lethargy for two months. He gave a history of stroke six months before admission. On examination he was pale and icteric. No lymphadenopathy, malar rash, oral ulcers, livedo reticularis were noted. Cardiovascular system examination revealed a pansystolic murmur of mitral regurgitation. Firm, non-tender, moderate hepato-splenomegaly was detected in the abdomen. Ophthal-moscopy showed bilateral retinal hemorrhages. He had dementia with a mini mental score (MMSE) of 10/30 and left sided spastic hemiparesis. Investigations showed haemoglobin of 3.8 g/dl with normochromic, normocytic red cells, few polychromatic cells, marked auto agg-lutination suggestive of cold AIHA and thrombocytopenia. Reticulocyte count was 12%. Direct and indirect Coombs tests, pan reactive cold antibodies and direct antibody test with IgG and C3D specificities were positive. Trephine biopsy revealed hyper cellularity and predominant erythropoiesis with megaloblastic changes. ESR was 104 mm with a normal CRP level. Anti-nuclear antibody and antibodies to double stranded DNA were negative. Transthoracic echocar-diography revealed organised vegetations over the mitral valve. Three blood cultures were negative. Non contrast computed tomography (CT) showed multiple cerebral infarctions. Contrast enhanced CT scans of chest and abdomen did not reveal any mediastinal or para aortic lymphadenopathy. Mycoplasma serology was negative. Activated partial thromboplastin time was not prolonged. VDRL was positive with negative *Treponema pallidum* particle agglutination assay (TP-PA). Anti cardiolipin antibodies (AcL) were positive in IgG (129) and IgM (78.2) and remained positive after six weeks. The diagnosis of APS was made in the presence of one clinical (history of stroke and multi infarct dementia) and one laboratory criteria (positive AcL). Presence of cardiac vegetations with sterile blood cultures (Libman sacks endocarditis), retinal hemorrhages, thrombocytopenia and AIHA supported this diagnosis. The secondary causes for APS such as systemic lupus erythematousus (SLE) and cold AIHA (lymphoma, Mycoplasma infection) were excluded.

APS is a thrombophilic disorder mediated by autoantibodies against phospholipid binding plasma proteins [1]. Predominant features of APS include arterial or venous thrombosis, thrombocytopenia, coombs positive haemolytic anaemia, cardiac vegetations, seizures, multi-infarct dementia and migraine [2]. APS is often associated with SLE. APS without clinical or laboratory evidence of SLE is named as 'primary APS' [3]. This patient had several clinical and laboratory evidence of APS (multiple cerebral infarctions, positive aCL antibodies, cold AIHA, Libman-sacks endocarditis, retinal vessel thrombosis, thrombocytopenia and multi infarct dementia) in the absence of secondary cause (negative screening tests for SLE, lymphoma, Myco-plasma infection) [2, 4]. Coombs positive AIHA is rare in APS and cold type is even rarer and no similar cases are reported in the literature [3]. He was treated with oral prednisolone 1.5 mg/kg with improvement in haemo-globin level.

Conflicts of interest

We declare that there are no conflicts of interest.

References

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