To the Editors:

Antiphospholipid syndrome in a man presenting with cold autoimmune haemolytic anaemia

H M A Ediriweera¹, G G A Gayani¹, K D Pathirana², T P Weerarathna², M R Mohideen²

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. Ophthalmoscopy 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 echocardiography 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 haemoglobin level.

Conflicts of interest
We declare that there are no conflicts of interest.

References
To the Editors:

Successful management of a pregnancy complicated by essential thrombocythaemia with pegylated interferon

W M N D Jayasekara, S A S P Abeyratne, C Kulathilake, D Gunawardena, I S Wijesiriwardena

(Index words: essential thrombocythaemia, pregnancy, pegylated interferon alpha)

Ceylon Medical Journal 2015; 60: 72-73

Introduction

Essential thrombocythaemia (ET) is an acquired thrombophilic condition. Though it is more common in elderly females, about 20% are in the child-bearing age giving rise to the possibility of ET complicating pregnancy. In the few reported cases, majority were managed with Interferon-$\alpha$ (INF-$\alpha$), reflecting its efficacy and low toxicity in pregnancy.

We present a case report of ET complicating pregnancy which was successfully managed with pegylated interferon (PEG-IFN) $\alpha2a$, which has a longer half-life and less side effects.

Case report

The patient was a 36 year old woman, P 5C1 who was diagnosed with ET one year prior to the current pregnancy. She had a bad obstetric history with one first trimester miscarriage, one second trimester intrauterine death and one early neonatal death following premature delivery at 27 weeks due to pregnancy induced hypertension. She had a history of thrombosis (transverse sinus thrombosis) during the third pregnancy and was treated with therapeutic dose of low molecular weight heparin (LMWH). This was the only successful pregnancy which produced a healthy baby.

One year prior to the current pregnancy, she was investigated for thrombocytosis (platelet count was above 700,000/mm$^3$ in serial monitoring) and bone marrow trephine morphology was in favour of ET and JAK2 V617F mutation was positive. She was managed with hydroxyurea and low dose aspirin.

She was reviewed at 8 weeks of pregnancy and was off hydroxyurea during the preceding two weeks. The platelet count was 700,000/mm$^3$. Since she fell into the high risk category of ET, we treated her with Enoxaparin 40 mg/daily until 16 weeks of pregnancy and then 40 mg twice a day throughout pregnancy and aspirin 75 mg once a day [1]. As the platelet count was constantly around 600-700,000/mm$^3$, we treated her with PEG-INF-$\alpha2a$. The aim was to maintain the platelet count less than 400,000/mm$^3$ which we achieved with PEG-INF-$\alpha2a$ escalating dose of 45/90 $\mu$g/week throughout pregnancy. She delivered at 38 weeks of gestation by an elective caesarean section. PEG-INF-$\alpha2a$ was continued post-partum and she was allowed to breastfeed. Enoxaparin and aspirin were given for six weeks post partum, and low dose aspirin was continued thereafter. Cytoreductive therapy was changed from PEG-INF to hydroxyurea once she stopped breastfeeding.

Discussion

Essential thrombocythaemia is a sub category of myeloproliferative neoplasms (MPNs) characterised by a persistently elevated platelet count with associated