Successful Pregnancy Outcome in a Patient with Antiphospholipid Syndrome : A Case Report

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Abstract

Antiphospholipid syndrome (APS) manifests clinically as recurrent venous or arterial thrombosis and/or fetal loss. Diagnosis requires a high index of suspicion during evaluation of women with recurrent pregnancy loss and vascular thrombosis. Low dose aspirin combined with heparin

can reduce morbidity and improve the pregnancy outcome. Here we report a case of a 22 year old lady having APS who presented with arthritis, recurrent miscarriages and venous thrombosis.

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Introduction

Antiphospholipid syndrome (APS) is a multisystem disorder diagnosed by the presence of lupus anticoagulant (LAC) and anticardiolipin (aCL) antibodies in association with venous and/or arterial thrombosis or pregnancy complications. These criteria were proposed in the Eighth International Symposium

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on Antiphospholipid Antibodies by the American Autoimmune Related Diseases Association. Sapporo criteria are considered acceptable for the diagnosis of APS which includes unexplained or repeated pregnancy loss at around 10 weeks of gestation and positive aCL IgM and IgG or LAC antibodies on at least 2 occasions 6 weeks apart. APS has been associated with events such as thrombosis, vascular injury and vasoconstriction, all of them are possible causes of abnormally reduced maternal-fetal interface blood flow. These patients are at high risk for intrauterine growth restriction, severe pregnancy-induced hypertension, prematurity and abruptio placentae. Treatment with aspirin and heparin improve the pregnancy outcome.

Case Report

A 22-year-old lady presented at the "Rheumatology Clinic" of BIRDEM with two year history of arthritis involving hand and wrist joints, photosensitive and pruritic rash over the face and recurrent oral ulcers. She did not have any history of fever, loss of hair, Raynauds phenomenon, oedema, convulsion or chest pain. Her past medical history included 3 spontaneous abortions between 6th and 9th weeks of gestation and one episode of left femoro-popliteal and external iliac vein thrombosis. Past medical record revealed that she had anaemia, splenomegaly, high ESR, positive ANA and normal renal function tests. She received treatment with various NSAIDs and prednisolone irregularly.

At the clinic she was found afebrile and anaemic. There was no rash, oral ulcer or lymphadenopathy. She had

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swelling and tenderness in MCP and PIP joints. Other examination findings were unremarkable.

Her Hb was 9gm/dl, normochromic, normocytic, normal total and differential WBC and platelet count, ESR was 86mm in1st hour, CRP was negative. Urine RME was normal as well as creatinine. ANA was positive but antids DNA, anti-sm antibodies and RF were negative. Antiphospholipid (aPL) antibody (done by ELISA) was positive. Abdominal ultrasound revealed normal findings.

So, the lady was diagnosed as having systemic lupus erythematosus (SLE) with APS. She was appropriately counseled regarding the disease, treatment, monitoring and prognosis specially pregnancy planning and outcome.

Treatment was started with hydroxychloroquine, aspirin and prednisolone. She was on regular follow up with rheumatologist and gynaecologist. About 4 months after the last flare, she conceived. There was no flare throughout the pregnancy and she delivered a male baby of 2.4 kg by Caesarean section at term. No perinatal complication occurred.

Discussion

About 5-15% of recurrent pregnancy loss are associated with APS³ and 21-56% of SLE patients have secondary APS.⁴ APS may affect any organ of the body and display a broad-spectrum of manifestations. These include deep venous thrombosis (31.7%), thrombocytopenia (21.9%) pulmonary embolism (9%), transient ischaemic attacks (7%), stroke (13.1%), myocardial infarction (2.8%), skin ulcers (3.9%) and rarely a catastrophic syndrome characterized by widespread vascular occlusion (0.8%).⁵ Most of these systemic features can be explained by vasculopathy and occlusion of small vessels due to platelet aggregation and subsequent thrombosis. In our case, the patient had arthritis, skin rash, oral ulcer, deep vein thrombosis, recurrent abortions which was highly suggestive of secondary APS.

The mechanism of vascular thrombosis and pregnancy loss in the APS are unknown but several theories have been postulated. The most popular one is level of annexin V. Annexin V is a phospholipid-binding protein with potent anticoagulant activity, levels of which are markedly reduced on placental villi of women with APS. Hypercoagulability in such women may therefore be due to the reduction of surface bound annexin V by aPL

antibodies. aPL antibodies reduce the levels of annexin V and accelerate the coagulation of plasma on cultured trophoblasts and endothelial cells. The reduction of annexinV levels on vascular cells may be an important mechanism of thrombosis and pregnancy loss in the APS.⁶

Patient with APS have high titres of aCL and LAC antibodies. The association of positive aPL and LAC antibodies with recurrent fetal loss is well established and varies between 4.6 to 50.7% (mean 15.5%) and 0 to 14% (mean 8.3%) respectively. The presence of aPL antibodies is also associated with placental insufficiency, fetal growth restriction, pre-eclampsia and preterm delivery.

Prevalence of APS varies according to the population assessed and the immunoassay used to detect aPL antibodies. The study published by Lockwood et al found a prevalence of 2.2% for aCL and 0.27% for LAC antibodies in low risk obstetrics patients. Our patient had positive aPL antibody.

With proper management, more than 70% of pregnant women with APS deliver a viable live infant. Present therapy for the thrombotic aspects of the APS-lupus complex is anticoagulation with heparin and low dose aspirin. Standard steroid and immunosuppressive therapy seem to have little effect on antibody levels. 10 Heparin is usually started in the early first trimester after confirmation of presence of a live embryo by ultrasonography. In one study only low dose aspirin was used for prevention of pregnancy loss, which was started one month before conception and continued throughout the pregnancy. Prior to therapy the rate of live-born babies was 6.1%, and after therapy it was 90.5%.¹⁰ Most investigators recommend preconceptional aspirin because of its beneficial effect on early stages of implantion.¹¹

We treated our case of secondary APS with hydroxychloroquine, corticosteroid, low dose aspirin. She conceived and delivered a healthy baby without any complication.

Conclusion

Routine screening of pregnant women for APS is not recommended as the prevalence is low. However, aCL and LAC antibodies must be checked in women presenting for evaluation of recurrent pregnancy loss.

Management of APS is aimed to improve maternal and fetal outcome. It can be achieved by preconceptional counseling, multidisciplinary approach and careful monitoring of pregnancy.

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