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Antiphospholipid syndrome (APS), also known as 'sticky blood syndrome' or Hughes syndrome, can cause blood clot in arteries and veins

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Abstract
Antiphospholipid syndrome (APS), is an autoimmune disorder, in which Antiphospholipid antibodies (Anticardiolipin antibodies and lupus anticoagulants) react against proteins that bind to anionic phospholipid which characterized by pregnancy complications (preeclampsia, still birth, miscarriage etc.), thrombosis, autoimmune thrombocytopenia, arthritis. ASPs are, treated by giving aspirin to inhibit platelet activation, and/or warfarin as an anticoagulant. Plasmapheresis includes as a treatment of choice for refractory cases.

Keywords: Antiphospholipid syndrome (APS), Hughes syndrome, autoimmune antibodies

Introduction
Antiphospholipid syndrome (APS) is an autoimmune condition characterized by a thrombotic event and/or pregnancy morbidity in the presence of persistently elevated antiphospholipid (aPL) antibody titers, which are most prevalent in patients with autoimmune (SLE), malignant, and infectious. Recent research shows that factors possibly integral to rheumatoid arthritis's increased morbidity and mortality related to aPL include oxidatively modified low-density lipoprotein antibodies, annexins, infectious agents, beta estradiol, homocysteine and specific gene polymorphisms. Moreover current scientific research addressing the pathophysiologic mechanisms and clinical implications of aPL in rheumatoid arthritis.

Definition
Antiphospholipid syndrome caused by Antiphospholipid antibodies (aPL) associated with variety of autoimmune, malignant, and infectious diseases, with or without the thrombotic or obstetric sequelae.

Epidemiology
Antiphospholipid antibodies (aPL) positively in
- 10 – 40% in SLE
- 20% approximately in rheumatoid arthritis
- 10% of 1st stroke patients, more in young upto 29%
- 20% in women with 3 or more consecutive fetal loses
- 14% in recurrent thromboembolic diseases

Risk Factors
- These include diabetes, hypertension or high blood pressure, hypercholesterolemia or high cholesterol, obesity, smoking, estrogen therapy for menopause or contraception, and any underlying systemic autoimmune disease.
- Having an autoimmune condition, such as systemic lupus erythematous or Sjogren's syndrome
- Having certain infectious diseases like syphilis, HIV/AIDS, hepatitis C or Lyme disease
- Taking certain medications, such as hydralazine for high blood pressure, the heart rhythm-regulating medication quinidine, the anti-seizure medication phenytoin (Dilantin) and the antibiotic amoxicillin
- Having a family history of antiphospholipid syndrome

Causes
Although, there are many risk factors in APLS, there is no specific cause for it,
It can be triggered by following factors
- Smoking
- Immobility (related, for instance, to the thrombosis seen after long-haul flights)
- the contraceptive pill
- Genetic factors – there may be a family history of clots, miscarriages, other autoimmune diseases such as lupus or thyroid problems.

Pathophysiology
The endothelium (inner most lining of blood vessels) has complex protective mechanisms to prevent thrombosis which gives an anticoagulant effect. antiphospholipid (aPL) antibodies can bind to beta2-glyco-protein I and prothrombin. Normally Beta2-glycoprotein I and prothrombin are proteins that bind to phospholipids on cell walls in the process of thrombus formation. The underlying trigger that results in the formation of antiphospholipid antibodies is idiopathic. Indeed, the presence of the antiphospholipid syndrome simply seems to tip the body's normal balance between thrombosis and anticoagulation in favor of thrombosis, it could be a reason for this condition named as 'sticky blood syndrome'.
Antiphospholipid syndrome (APS) is an autoimmune disease, which means that it's caused by immune system attacking healthy parts of the body instead of fighting infections. If patient has APS, the immune system produces harmful

antibodies called antiphospholipid antibodies (aPL) which attack proteins are linked to fats in the body. The most important of these proteins is called beta-2-glycoprotein I. When aPL stick to this protein they can interfere with blood cells. The cells change in such a way that the blood becomes sticky and more likely to clot inside the vessels.

Signs and Symptoms

- Most individuals with antiphospholipid antibodies do not have symptoms unless a complication occurs.
- APS have symptoms like rash, joint pain, migraine and tiredness even when they don't suffer thrombosis
- A thin, lacy, red-purple rash, livedo reticularis is common but does not cause symptoms.

Fig 1: Livedo reticularis in APS

- Patients with associated diseases such as SLE, arthritis will have the usual symptoms of that diseases.
- It can cause pregnancy complications, such as miscarriage and stillbirth.
- Pain, swelling, redness, or tenderness in leg or arm.
- Vein swelling and pain are severe or are accompanied by chest pain or shortness of breath. Blood clots in legs (DVT). The clots can travel to lungs (pulmonary embolism).
- Repeated miscarriages or stillbirths. Other complications of pregnancy include premature delivery and high blood pressure during pregnancy (preeclampsia).
- Stroke. A stroke can occur in a young person who has antiphospholipid syndrome but no known risk factors for cardiovascular diseases.
- Transitory ischemic attack (TIA). Similar to a stroke, a TIA usually lasts only a few minutes and causes no permanent damage.
- Neurological symptoms. Chronic headaches, including migraines; dementia and seizures are possible when a blood clot blocks blood flow to parts of your brain.
- Cardiovascular disease. Antiphospholipid syndrome can damage heart valves.
- Bleeding. Some people have a decrease in blood cells needed for clotting (platelets). If patients have this condition (thrombocytopenia), they might have few or no symptoms.
- However, if platelet count drops too low, might have episodes of bleeding, particularly from nose and gums. It can also bleed into skin, which will appear as patches of small red spots (petechiae). An unusually heavy menstrual period; vomit that is bright red or looks like coffee grounds; black, tarry stool or bright red stool; and unexplained abdominal pain.
- Blood clots in leg veins cause a condition known as deep vein thrombosis (DVT). Damage from blood clots in the organs, such as kidneys, lungs or brain, depends on the extent and location of the clot.

Diagnosis

- Serum anticardiolipin antibodies (aCL), anti-β2 -glycoprotein I antibo- dies (a-β2 -GPI), lupus anticoagulant (LAC), immunoglobulin M (IgM) rheumatoid factor (RF), and anticyclic citrullinated peptide antibodies (anti-CCP) were measured and disease activity can assessed.
- Laboratory criteria: Anticardiolipin antibody of the immunoglobulin G (IgG)/immunoglobulin M (IgM) isotype in medium/high titer (> 40 IgG phospholipid units [GPL], > 40 IgM phospholipid units [MPL], or > 99th percentile) on two or more occasions at least 12 weeks apart (measured by a beta2-GPI-dependent enzyme-linked immunosorbent assay).
- Prolonged phospholipid-dependent coagulation (e.g. activated partial thromboplastin time, Kaolin clotting time, dilute Russell viper venom test, dilute PT test).
- Failure to correct the prolonged coagulation time by a mix with platelet-poor plasma.
- Shortening or correction of the prolonged coagulation time with excess phospholipids.

Management

- Exclusion of other coagulopathies (e.g. factor VIII inhibitor, heparin) Management of thrombosis Often, treated by warfarin as an anticoagulant for the prevention of recurrent stroke and/or giving aspirin to inhibit platelet activation, showed that both are useful in patients with first ischemic stroke and a single positive aPL detection. After a first episode of thrombosis, patients with aPL antibodies have a higher risk of recurrent thrombosis than patients without the antibodies. The optimal regimen for arterial thrombosis is less clear.
- Single detection, low titers of anticardiolipin (aCL), isotype of aCL considered, lupus anticoagulant (LA) treatment not performed according to international recommendations
- To maintain the patient's INR between 2.0 and 3.0 the prophylactic treatment with warfarin is usually done in patients who have had no thrombotic symptoms.
- Patients with APS and a venous thromboembolic event should be treated with indefinite warfarin therapy to an INR of 2—3;
- patients with definite APS and arterial thrombosis and/or recurrent venous events should be treated with indefinite warfarin therapy to an INR > 3;
- The association of aspirin in the patients with recurrent thromboembolic events while on anticoagulant therapy is still a matter of some controversy and there are no consistent data to recommend it.
- Management of pregnancy Anticoagulation appears to prevent miscarriage in pregnant women. In pregnancy, low

molecular weight heparin and low-dose aspirin are used instead of warfarin because of warfarin's teratogenicity.
- Prior to pregnancy, a complete aPL-antibody profile should be performed. Pregnancy should be discouraged when patients have experienced a thrombotic event in the previous 6 months or in women with untreated hypertension or pulmonary hypertension. In women with APS visits every 2 weeks are recommended until mid-gestation and weekly thereafter, also monitoring of blood pressure and urine protein, and frequent ultrasounds to check fetal growth and well-being.
- Intravenous gamma globulin is an alternative treatment that has been proposed; however, the efficacy of this approach has not been demonstrated
- Plasmapheresis is used to remove aPL antibodies in an attempt to avoid spontaneous abortion in women with multiple previous miscarriage, but the usefulness of this procedure needs to be confirmed.
- During *in vitro* fertilization (IVF) procedures, as established at the 5th International Conference: Sex Hormones, Pregnancy and Rheumatic Disease (Florence, Italy, April 2007), the use of heparin is mandatory only in woman with classic APS undergoing IVF, while no consensus was obtained for heparin treatment in asymptomatic aPL-positive woman.

Nursing Interventions

Exercise

Although there aren't any specific exercises that can help with the condition, taking regular exercise will help keep you fit and keep your heart healthy.

Diet and nutrition

- Increasing the amount of essential fatty acids in diet, particularly omega-3 fatty acids found in oily fish, could help reduce risk of blood clots. As well as this, fish oils contain large amounts of vitamin A which can be harmful in pregnancy, so wouldn't recommend this in pregnancy.
- Eating a healthy, balanced diet is important for general health and may help prevent from developing blood clots, also keep a healthy weight.
- At present, no complementary therapies have been shown to help with APS.
- Avoid "bingeing" and crash diets.
- Limit intake of foods that are rich in vitamin K to one serving a day, for example, one cup of raw, or half a cup of cooked spinach, turnip greens, cucumber peel, broccoli, Brussels sprouts, green scallion, cabbage, and mustard greens. Avoid parsley, kale, seaweed, and green tea.

Reducing risk of getting clots

- Don't smoke – smoking will increase risk of clots.
- Limit the consuption of excessive amounts of alcohol.
- Think very carefully about what sort of contraception to use and discuss it with doctor, as some types of contraceptive pill increase the risk of clotting.
- hormone replacement therapy after the menopause may also increase risk of clots
- Another condition that can increase risk of blood clots (e.g. diabetes, high blood pressure or high cholesterol) should make sure that have regular checks to keep these factors under control.

Conclusions

The management of APS focuses on antithrombotic therapies and anticoagulation, however, there are many unanswered questions regarding the best practice for aPL-positive patients owing to difficulties in describing the wide clinical and serological spectrum of the syndrome. In addition to that, we need more studies to support over in it.

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