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STICKY BLOOD SYNDROME

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ABSTRACT

An increasingly important link between antiphospholipid antibodies and a clinical syndrome is becoming recognized worldwide. This syndrome, known as the APS or Sticky blood Syndrome, is a prothrombic disorder leading to both arterial and venous thrombosis and in pregnancy, recurrent abortion and pregnancy loss antiphospholipid syndrome is one of the most cost common acquired causes of hypercoagulability. The most common plasma protein target is beta 2 glycoprotein1. The three most important antiphospholipid antibodies are the lupus coagulant, anticardiolipin and beta 2 glycoprotein1. Unfortunately, it is a disease that is often under recognized and under diagnosed. This is probably because it can cause so many different problems many of which have other common causes. Early diagnosis is important to try to prevent serious complications.

Keywords: Hyper-coagulation, Beta 2 glycoprotein, Lupus coagulant

INTRODUCTION

The antiphospholipid syndrome (APS), first described in 1980's by Hughes, Harris, and Gharavi[(1)], is an acquired thrombophilic disorder in which autoantibodies are produced by a variety of phospholipids and phospholipid binding proteins. Together with the identified acquired thrombophilic states, APS is the most common and can be caused by lupus anticoagulant, anticardiolipin antibodies or other antiphospholipid antibodies. The antibodies in APS were originally thought to recognize that recognize certain phospholipids, fatty molecules that make up part of normal cell membranes, hence the name "anti-phospholipid antibodies". APS is called an autoimmune condition- when the body's normal defense begin to work against itself. It is now known that most of the autoantibodies in APS actually recognize certain blood proteins that bind to phospholipids, not the phospholipids themselves and the presence of antiphospholipid antibodies in the blood[(2)]. It is also known as Hughes syndrome, Sticky blood syndrome, Antiphospholipid syndrome.

There are two main types of APS

1. Primary APS: The individual has no known autoimmune disease present, other than APS. This type accounts for more than 50% of all cases.
2. Secondary APS: The individual had already been diagnosed with systemic lupus erythematosus (SLE) or another underlying immune disorder. More women than men have

this type of APS and other overlapping connective tissue diseases.

3. APS is called an autoimmune condition- when the body's normal defenses begin to work against itself.

ETIOLOGY

Antiphospholipid syndrome seems to be related to the presence of antiphospholipid antibodies in the blood, the exact cause is unknown. The immune system makes proteins called antibodies which are helpful and do jobs such as fighting infection. In APS, useful antibodies are formed and these attack normal substance called phospholipid (which is why the antibodies are called antiphospholipid). In most of the people, a certain event such as pregnancy or infection provides a trigger for this chain of events in APS.

- Infection: Infections like HIV, syphilis, Hepatitis C, Lyme disease to have a higher incidence of having antiphospholipid antibodies.
- Medications: The antibiotic amoxicillin, hydralazine take for high blood pressure, Quinidine is taken for regulation of heart rhythm, and an anti -seizure medication called phenytoin it may lead to higher risk for the development of APS.
- Genetics or Heredity: Research indicates that relatives of those with antiphospholipid syndrome have an increased likelihood of having the antibodies themselves, even though the disorder is not considered to be hereditary.

EPIDEMIOLOGY

The disease affects all age groups and the majority of patients are aged between 15 and 50 years. It is estimated that the incidence of APS is approximately 5 cases per 100,000 persons per year and the pre-valance is approximately 40-50 cases per 100,000 persons. Females seem to out-number males, though not to any extent and anti-phospholipid antibodies found more frequently in elderly persons and 30-40% patients with SLE, but only about 10% have APS approximately and half of the cases are not associated with rheumatic disease. APL syndrome is the cause of 14% of all strokes, 11% of myocardial infarction, 10% of deep vein thromboses, 6% of pregnancy morbidities and 9% of pregnancy losses[(2)].

CLINICAL PRESENTATION

1. VEIN THROMBOSIS	• Deep vein thrombosis: Example: leg or arm • Thrombosis in internal organs. Example: kidney, liver, lung, brain. • Thrombosis of skin vessels, skin ulcers, "Livedo" (bloatchiness of the skin)
2. ARTERY THROMBOSIS	• Brain- headaches (often migraines), weakness, slurred speech, strokes, seizures, memory loss. • Limb- pain, circulation problems, other organs- heart, Kidney, adrenal related.
3. PREGNANCY LOSS	• Beginning of pregnancy (miscarriage) or late (fetal death) • Periodic very early miscarriage may give rise to diagnosis of infertility[(3)]
4. THROMBOCYTOPENIA	• Low platelet count • Bruising 5-10%
5. STROKE/NEUROLOGICAL DISORDERS:	• Embolic stroke (caused by a blood clot that travels to the brain) • Cerebrovascular thrombosis (a blood clot that forms in the brain)[(4)] • Seizures, migraine, cognitive dysfunction.
6. RASHES OR SKIN CONDITIONS	• A rash like mottled or lacy net-like pattern • ulcers or sores most often on the legs and necrosis

7. AUTOIMMUNE DISEASE	• Systemic lupus erythematosus is most commonly associated with APS

PATHOPHYSIOLOGY

In APS homeostatic regulation of blood, coagulation is altered. One hypothesis postulates a defection in cellular apoptosis which uncovers membrane phospholipids to the binding of various plasma proteins such as beta 2 glycoprotein1. Once bound, a phospholipid-protein complex is formed and a neo-epitope is uncovered, which subsequently becomes a target of antibodies[(5)]. Recent studies suggest that oxidized beta 2 glycoprotein1 is able to bind and activate dendritic cells in a manner similar to the production of autoantibodies.

- Other proposed mechanisms for hypercoagulable effect of antiphospholipid antibodies include
- Production of antibodies against coagulation factors including prothrombin, protein C, protein S[(6)]
- Activation of platelets to increase endothelial adherence
- Activation of vascular endothelium, which, in turn, facilitates the binding of platelets and monocytes
- Reaction of antibodies to oxidized low-density lipoproteins, thus predisposing to atherosclerosis and myocardial infarction (MI)
- Complement activation has been increasingly recognized as a possible significant role in the pathogenesis of APS. Emerging evidence from murine models suggests that antiphospholipid mediated complement activation may be a primary event in pregnancy loss[(7)].

COMPLICATIONS

Depending on which organ is affected by a blood thrombi and how severe the obstruction of blood flow to that organ is, the untreated antiphospholipid syndrome can lead to permanent damage or death. Complications may include:

- Renal failure
- Stroke
- Cardiovascular problems
- Lung problems
- Pregnancy complications

DIAGNOSIS

Blood tests are used to make a diagnosis of antiphospholipid syndrome looking for at least one of these following

1. Lupus anticoagulant
2. Anti cardiolipin
3. Prolonged PTT
4. Beta 2 glycoprotein1

A diagnosis will be confirmed after the antibodies appear in your blood at least twice in tests that are taken at least 12 weeks apart

TREATMENT

The treatment plan for APS will be specific to the individual and their current health status with regards to the symptoms that they are experiencing. Ideally, any treatment regimen will be directed towards thinning (anti-coagulating) the blood to prevent clotting. Here are some medications (anticoagulants) used to treat APS and to prevent further blood clots

- Aspirin- low dose aspirin at a dose 75mg to 100mg a day will make the blood platelets less sticky which are an effective anti-coagulating agent[8]
- Heparin- low molecular weight heparin such as clexane useful as first line treatment in thrombosis prior to warfarin use which is also safe in pregnancy and only available in injection (parenteral) form[9].
- Warfarin: It is not advised in pregnancies and is an effective blood thinner[9].
- Clopidogrel: It thins the blood and prevents blood clot formation. Sometimes it is advised with warfarin or used alone.
- Antimalarial as Lupus treatment: Prednisolone- Are reserved for patients with very low platelet count or sometimes for women who experience a high number of early miscarriages.
- Hydroxychloroquine: Particularly effective in helping skin rashes, fatigue, aches and pains and used in treating autoimmune conditions. It also acts as mild anticoagulant and has also been found to reduce anticardiolipin antibodies
- Intravenous Immunoglobulin(IVIG): They neutralize a number of harmful antibodies. It is effective in patients with APS with low platelet counts
- Vitamin D: This supplement is advised as APS patients are found with vitamin D deficiency and it has additional advantages.
    - It is very important to speak with physician about any other medications or supplements which you are taking as they may cause the anticoagulant medications to be less than satisfactory or interact in a dangerous way with your anticoagulants

Things to reduce the chances of developing blood clots

- Don't smoke or use tobacco products
- Maintain cholesterol and triglyceride levels

RISK FACTORS

- Having a family member with Antiphospholipid syndrome
- Infections including syphilis, HIV/AIDS, hepatitis C, or Lyme disease
- Taking certain medications like hydralazine for high blood pressure, the heart rhythm controlling medication quinidine, the anti - seizure medication phenytoin and the antibiotic amoxicillin.
- The presence of an autoimmune condition such as systemic lupus erythematosus or Sjogren's disease.

CONCLUSION

While there is no cure for the anti-phospholipid syndrome, there are many successful ways to - manage and treat this condition. Clearly and regularly communicate with your physician if you are diagnosed with APS, be attentive of any new symptoms, maintaining good medical records with dates and descriptions of symptoms. This will help devise the optimum course of action and make necessary adjustments to get the best possible outcome as well as to help prevent any further complications.

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