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A Case Report on Antiphospholipid Syndrome

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ABSTRACT

Antiphospholipid syndrome is a systemic autoimmune disease characterized by the presence of antiphospholipid antibodies in association with thrombosis and/or obstetric complications. APS is also be presented with multiple organ involvement. We recently encountered a case of primary phospholipid syndrome in a young adult with newly diagnosed pulmonary embolism, immune thrombocytopenia and deep vein thrombosis of left leg. The diagnosis of APS was initially suspected with his prolonged APTT and elevated D-Dimmer values and later confirmed by triple positive antiphospholipid antibody profile and multiple organ involvement

Keywords: Antiphospholipid syndrome, Thrombosis, Autoimmune disease.

INTRODUCTION

Antiphospholipid syndrome is an autoimmune disease characterized by thrombosis affecting both venous and arterial circulation, that occurs in patients with persistent antiphospholipid antibodies.[1] The deep veins of lower limb and cerebral arteries are the most common sites of thrombus formation. Moreover, any tissue or organ can be affected.[2],[3] The presence of thrombosis involving multiple organs is known as catastrophic antiphospholipid syndrome.[4],[5] Obstetrical involvement is characterized by recurrent early miscarriages, fetal loss after 10th week of gestation, intrauterine growth restriction, or severe pre-eclampsia.[6] The non-thrombotic manifestations of antiphospholipid syndrome include valvular heart disease, antiphospholipid antibody related nephropathy, hemolytic anemia thrombocytopenia and cognitive dysfunction.[7] The antiphospholipid syndrome often associated with other systemic autoimmune diseases such as systemic lupus erythematosus (SLE), however it also occurs without other autoimmune manifestations known as primary antiphospholipid syndrome.[3] The classification criteria for the antiphospholipid syndrome emphasize the presence of specific autoantibodies as an essential component for the diagnosis. The autoantibodies include anti-β_2-glycoprotein (IgG, IgM) or anticardiolipin (IgG, IgM) or lupus anti-coagulant specific.[7]

CASE REPORT

A 27year old male patient presented with the complaints of shortness of breath, palpitation and sweating, and also presented with hemoptysis since 2 days. Detailed history revealed that patient has shortness of breath and double vision episode 1 month back. He was initially admitted in the MICU with the above mentioned complaints. On evaluation CT pulmonary angiogram suggested features of acute pulmonary embolism involving predominantly the right upper and lower lobar and segmental pulmonary arterial branches, with significant RV strain. Peripheral ground glass change with reticulation in the basal

segments of the lower lobe-CORADS III-IV (Mild extend of involvement) Moderate pericardial effusion. Laboratory investigations suggested peripheral smear with normocytic normochromic blood picture and thrombocytopenia with platelet count of 41,000/cmm. Markedly prolonged activated partial thromboplastin time of 68.5S was reported. Color doppler study of both lower limbs for DVT was performed which suggested echogenic thrombus in popliteal vein and the distal superficial femoral vein on left side. Echo cardiogram reports concluded dilated RA and RV, Severe pulmonary arterial hypertension, right ventricular dysfunction and mild pericardial effusion. Renal function test showed a creatine level of 1.4mg/dl which revealed AKI non oliguric. Ultra-sonogram of abdominal showed grade I fatty liver and liver function test presented slightly elevated AST and ALP levels. He was further diagnosed to have primary antiphospholipid syndrome with triple APLA- anti-β_2-glycoprotein (IgG). anticardiolipin (IgG) and lupus anti-coagulant positive, the diagnosis also involved pulmonary embolism, immune thrombocytopenia and Deep vein thrombosis. He was treated with STAT dose of solumedrol 125mcg, followed by oral prednisolone, anticoagulants, and vitamin supplements. Warfarin was administered and dose adjustments was done as per platelets and INR values. The patient symptomatically improved with the given treatment.

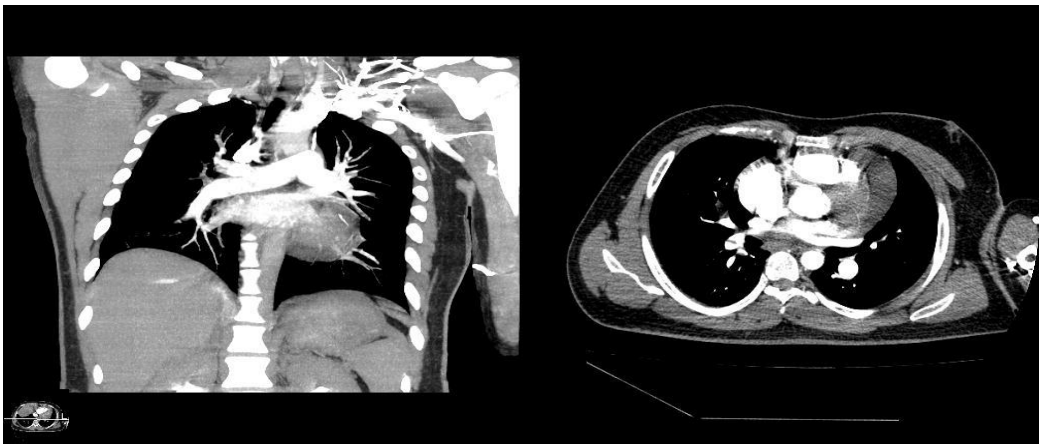

Fig1,2: Images representing pulmonary embolism

DISCUSSION

Antiphospholipid syndrome is prothrombotic syndrome which invade multiple organs with thrombosis and further cause organ damage. The damage caused by the thrombus can vary depending on the site of the thrombi. For instance, repeated small clots formed in the heart can cause heart valve thickening or damage, with the risk of releasing clots into the blood. Thrombosis in antiphospholipid syndrome can occur anywhere in the circulation and can affect any organ in the body.[8]

The production of these auto antibodies is likely triggered by an environmental factor, such as an infection occurring in an individual with genetic background make them more susceptible to the disease. APLA can be present in the blood stream for a long time, but thrombotic events results when other conditions favors clotting are present, such as prolonged inactivity, surgery or pregnancy.[1]

The major clinical manifestations of APS are the occurrence of thrombosis and pregnancy complications. Single or multiple thrombi in veins, arteries and microvasculature can occur and the time interval between these manifestations may vary from days to years. Therefore, the variability in the location of the thrombi results in wide spectrum of clinical

presentations involving many organ systems.[2],[5] Stroke is one of the common and severe neurological manifestations of APS.[9] Cardiac features associated with APS vary from Myocardial infarction, intracardiac thrombi, accelerated atherosclerosis, pulmonary hypertension, diastolic dysfunction, cardiomyopathy.[10] The prevalence of thrombocytopenia was found to be higher in individuals with APS. Pulmonary manifestations include pulmonary emboli and infarction.[2] Dermatological features include livedo reticularis which may be prolonged prognostic marker of severe disease. Other dermatological manifestation includes digital gangrene, skin ulcers, superficial skin necrosis, pseudo-vasculitis lesions and pyoderma gangrenosum-like lesions, characterized by deep, necrotic ulcers.[11] Renal manifestations associated with slow, occult onset of hematuria, proteinuria and renal insufficiency, or it may develop acutely and present with acute renal failure and hypertension.[12]

Primary thromboprophylaxis describes the prevention of thrombosis individuals without previous clots, whereas secondary thromboprophylaxis describes prevention of recurrent clots following a first thrombolytic event. The conventional management of cardiovascular risk factors by lifestyle changes plays a major role in primary thromboprophylaxis. Antiplatelet agents such as low-dose aspirin should be limited in high-risk individuals. Secondary thromboprophylaxis is based on anticoagulation mainly with vitamin k antagonists such as warfarin and heparin, although direct oral anticoagulant might have role as well.[13]

In patients with thrombotic APS hydroxychloroquine combined with vitamin k antagonists was not associated with recurrent thromboembolism, whereas individuals treated alone with vitamin k antagonists experienced a recurrent event of thrombosis.[14]

Acute management of patients with catastrophic ASP is based on anticoagulation, corticosteroids, plasma exchange and/or intravenous immuno-globulin administration.[5]

CONCLUSION

Antiphospholipid syndrome is a rare condition, which in the existence of additional thrombotic risk factors may lead to substantial morbidity and mortality. Appropriate diagnosis, its management and continued anticoagulation helps prevent future thrombotic events in people with antiphospholipid syndrome.

Conflict of Interest: None

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