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Clotting Conundrum: Unraveling Male Antiphospholipid Syndrome

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Abstract:
APS (Antiphospholipid syndrome) is categorized by thromboembolic events and pregnancy-related complications, accompanied by persistently higher levels of antiphospholipid antibodies. Clinical presentations of Antiphospholipid syndrome are highly variable, with potential involvement of any organ system, making it relevant across numerous medical and surgical specialties. We present a case of 38-year-old male having no prior co-morbidities reported a history of left lower limb pain as well as breathlessness for 2 days, with no history of immobilisation, recent surgery, smoking, or previous history of deep vein thrombosis. Lower limb Doppler ultrasound showed a thrombus in left popliteal vein. A CT pulmonary angiogram revealed thromboembolism in the right and left main pulmonary embolism. On thrombophilia workup, the patient has been found to be positive for Anti cardiolipin antibody IgM as well as Beta-2 glycoprotein IgG antibody. Patient was started on I.V heparin infusion. No other clinical or laboratory evidence for other systemic or autoimmune illnesses was identified. During the course of hospitalisation, the patient developed abrupt onset of abdominal pain. Contrast-enhanced CT scan of abdomen presented bilateral deep vein thrombosis extending into left renal vein along with inferior vena cava. Despite early initiation of treatment, our patient did not survive. This case report highlights the severe and rapidly progressive nature of antiphospholipid syndrome, underscoring the importance of early diagnosis and aggressive management to improve outcomes.
Keywords: *Antiphospholipid syndrome(APS), pulmonary embolism, anticardiolipin antibody, deep vein thrombosis, beta-2 glycoprotein antibody.*

INTRODUCTION:

APS (Antiphospholipid syndrome) is systemic autoimmune condition categorized by presence of antiphospholipid antibodies (aPL) that promote thrombosis along with pregnancy-related complications. It is significant cause of morbidity as well as mortality because of its association with arterial and venous thrombosis, along with recurrent fetal loss. Antiphospholipid syndrome could occur as a main disorder or as secondary disorder connected to autoimmune diseases, including SLE (systemic lupus erythematosus). The hallmark of antiphospholipid syndrome lies in persistent laboratory evidence of aPL, encompassing aCL (anticardiolipin), LAC (lupus anticoagulant), as well as anti-β2 glycoprotein I antibodies, detected at least twelve weeks apart. Clinically, antiphospholipid syndrome is diagnosed when patients have 1 or more thrombotic events (e.g., stroke, PE (pulmonary embolism), DVT(deep vein thrombosis)) or obstetric complications, including recurrent miscarriages or preeclampsia[1]

Management of antiphospholipid syndrome involves long-term oral anticoagulation, with aggressive management recommended for patients experiencing arterial events. In individuals with systemic lupus erythematosus, primary thromboprophylaxis is advised; additionally, it might also be beneficial in cases of purely obstetric antiphospholipid syndrome. Obstetric management relies on a multidisciplinary approach that combines high-risk medical and obstetric care, along with treatment involving aspirin as well as heparin. Antiphospholipid syndrome (APS) remains a diagnostic and therapeutic challenge due to its varied clinical manifestations and complications[2]. Here we present a young man who developed DVT extending to IVC (Inferior Vena Cava) and PE, later diagnosed to have Antiphospholipid syndrome.

CASE REPORT:

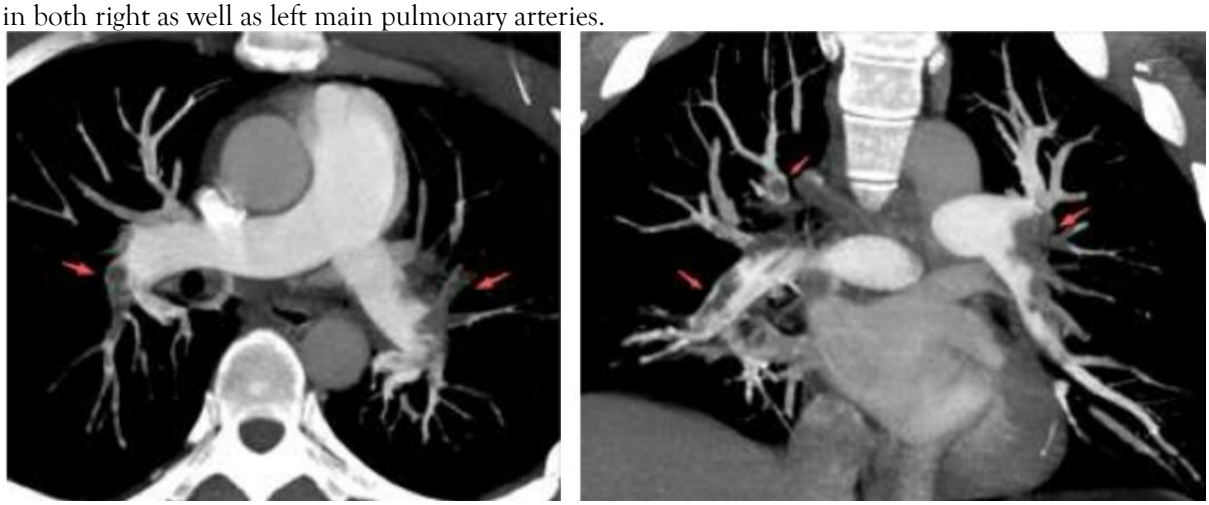
38-year-old male, who was a driver by occupation, having no prior co-morbidities, appeared to have left lower limb pain complaints for two days, as well as breathlessness for the past one day, sudden in onset, grade-3 MMRC. He denies any history of chest pain, palpitations, giddiness, or syncope. He denies smoking, prolonged immobilization, or any recent surgery. He denies any previous history of deep vein thrombosis.

On general examination, patient has been tachycardic with a heart rate 120 beats/min, initial BP-80/60mmHg, 130/80 on single ionotrope support(Inj. Noradrenaline), Oxygen saturation was 86% on room air, 96% on 4 liters oxygen. Systemic examination was uneventful. There was warmth and tenderness on the local examination of the left lower limb.

ECG showed Sinus tachycardia. Routine blood investigations were done. A complete blood count analysis showed Hb-16, total WBC count-11,500, and platelet-1.47 lakhs. Renal as well as liver parameters have been within normal limits. ESR has been within normal limits. CRP was elevated(64.1 mg/L). Peripheral smear showed mild thrombocytopenia.

The given clinical findings suggested a suspicion of DVT. Lower limb venous doppler ultrasound of left lower limb showed a thrombus in left popliteal vein. To rule out cardiac causes for breathlessness, a 2D echocardiogram was performed, which showed dilatation of the right atrium and right ventricle, TAPSE-1.3cm, suggesting the possibility of pulmonary embolism. The D-dimer was also elevated to 4200ng/ml.

CT pulmonary angiogram has been performed, which showed non-enhancing, non-calcific filling defect in both right as well as left main pulmonary arteries.



Figures 1 and 2: CT-Pulmonary Angiography - Filling defect (red arrow) noted in bifurcation pulmonary arteries and descending pulmonary arteries (Figure 1). Filling defects were noted in segmental and subsegmental branches of bilateral pulmonary arteries (Figure 2).

The patient was transferred to ICU on heparin infusion, oxygen support, and single inotrope support.

Intial Prothrombin time was 13.5sec, INR-1.1, aptt -78.6 seconds. Heparin was adjusted based on sixth-hourly apTT monitoring.

A thrombophilia workup was done to rule out the cause of this significant thromboembolic event. APLA profile was positive for serum cardiolipin IgM and beta-2 glycoprotein IgG and IgM. Serum homocysteine, protein-C, protein-S, factor-V Leiden genes, and anti-thrombin III have been within normal limits. Serum RA factor, anti-CCP, ANA IFA, ANA immunoblot, anti-ds DNA, C-ANCA P-ANCA, have been negative, ruling out other autoimmune diseases. Diagnosis of primary antiphospholipid syndrome has been established after multidisciplinary approach. Patient showed symptomatic improvement with heparin and was transitioned to oral rivaroxaban (10 mg twice daily).

During the hospital stay, the patient developed sudden-onset abdominal pain. Contrast-enhanced CT scan of abdomen presented non-enhancing, non-calcific, complete filling defect extending from the bilateral common femoral and external iliac arteries, extending to infra-renal, renal, and suprarenal IVC. After consultation with a vascular surgeon, thrombectomy was planned under high risk.

Despite high oxygen support, the patient's saturation deteriorated further. The patient went into sudden cardiac arrest and, despite aggressive resuscitation and multidisciplinary management, succumbed to his condition.

DISCUSSION:

APS(Antiphospholipid syndrome) is an autoimmune prothrombotic disorder characterized by persistent aPL presence, encompassing lupus anticoagulant, anticardiolipin, and anti-β2 glycoprotein I antibodies. It could occur as primary condition or be secondary condition, including SLE (systemic lupus erythematosus) [3]. Syndrome manifests clinically with venous or arterial thrombosis and/or obstetric complications, most commonly recurrent pregnancy loss. Venous thromboembolism, particularly DVT, is most frequent initial presentation of APS, and PE is a common complication along with life-threatening complications [4].

The pathogenesis of APS involves both cellular and humoral mechanisms. Antiphospholipid antibodies facilitate thrombosis by activation of endothelial cells, platelets, and monocytes, disrupting annexin A5 anticoagulant shield and inducing complement activation, thereby creating a hypercoagulable state [5]. Genetic and environmental factors, including infections, trauma, or surgical procedures, may act as a trigger for thrombotic events in predisposed individuals [6].

Diagnosis as per revised Sapporo criteria, which necessitate at least 1 clinical (pregnancy morbidity or vascular thrombosis) presence along with one laboratory criterion (persistent presence of aPL on two or more occasions, at least twelve weeks apart) [7]. In the presented case, the patient had significant thrombotic manifestations, including pulmonary embolism, DVT, along with inferior vena cava thrombosis. Diagnosis was confirmed by positive antiphospholipid antibody tests (IgM anticardiolipin as well as IgG anti-β2 glycoprotein I), meeting criteria for primary APS. Other prothrombotic and autoimmune conditions were ruled out.

Management of APS includes anticoagulation, with long-term warfarin treatment being the mainstay for patients with thrombotic events. DOACs (Direct oral anticoagulants), including rivaroxaban, are considered in some cases, although their efficacy in high-risk APS patients, particularly those with triple positivity, is controversial [8]. In acute settings, unfractionated heparin or low-molecular-weight heparin is preferred. Catastrophic variant of APS (CAPS), although not diagnosed here, should be considered when there is multiorgan thrombosis and rapid clinical deterioration. As per EULAR (European League Against Rheumatism), high-risk APS patients benefit from combination of low-dose aspirin and anticoagulation; immunosuppressive therapy is reserved for CAPS or coexisting autoimmune disorders [9].

This patient was appropriately managed with prompt initiation of intravenous heparin, inotropic and oxygen support, followed by a transition to oral rivaroxaban after stabilization. However, his sudden clinical deterioration with extensive IVC thrombosis despite anticoagulation highlights the aggressive nature of the thrombotic process in APS. In similar cases, early suspicion of APS and consideration of more intensive interventions like thrombectomy or thrombolysis may be warranted. Studies suggest that heparin remains superior to DOACs in avoiding recurrent thrombosis in high-risk APS patients [9].

This case underlines the need for early recognition of APS in young patients presenting with unprovoked thromboembolic events and the importance of timely laboratory testing and multidisciplinary care. Aggressive thrombotic involvement, even in the absence of comorbidities, can be fatal, as seen in this case.

CONCLUSION:

APS (Antiphospholipid syndrome) is rare but severe autoimmune prothrombotic disorder, particularly uncommon in males. This case highlights a fulminant presentation with extensive venous thrombosis as well as pulmonary embolism in previously healthy young man. Early diagnosis and prompt initiation of anticoagulation are crucial to improving outcomes. Despite appropriate management, the patient had a fatal outcome, underscoring the aggressive nature of APS in some cases. Clinicians must uphold a heightened level of suspicion for APS in young males exhibiting unprovoked thromboembolic events.

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