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Case Report

A rare presentation of primary apla and pulmonary thrombolism

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ABSTRACT

Antiphospholipid syndrome (APS) is a state of hypercoagulation, causing thrombus in artery, vein and capillaries, with or without obstetrical events with the presence of autoantibodies, which includes lupus anticoagulant, anticardiolipin antibodies and anti-β2-glycoprotein 1 antibodies. It has variable clinical presentations such as vascular thrombosis and obstetrical complications, pulmonary thromboembolism (PTE), pulmonary hypertension, and acute respiratory distress syndrome. Hereby we presented a case of young male who came to our hospital with hemoptysis and isolated pulmonary embolism, with the background of APS which is rare. Hemoptysis and pulmonary thromboembolism both being fatal emergencies requiring completely different management methods.

Keywords: Pulmonary thromboembolism, Antiphospholipid syndrome, Hemoptysis

INTRODUCTION

Antiphospholipid syndrome (APS) is the state of hypercoagulation, which can be characterized by the presence of thrombosis with or without obstetrical events with the presence of autoantibodies, which includes lupus anticoagulant, anticardiolipin antibodies and anti-β2-glycoprotein 1 antibodies.

It has variable clinical presentations such as vascular thrombosis and obstetrical complications like fetal growth restriction, unexplained recurrent miscarriages, pre-eclampsia.[1] Various pulmonary manifestations associated with APS including pulmonary thromboembolism (PTE), pulmonary hypertension, and acute respiratory distress syndrome.[2] The incidence of thrombosis in Chinese APS patients was 75.4%, of which 40.1% were DVT, 23.8% were stroke, and 6.7% were PE.[3]

APS can be primary when there is no evidence of autoimmune disease, or it can be secondary due to autoimmune conditions like systemic lupus erythematosus (SLE) in 40% of the cases.[4] Certain risk factors increase the risk of antiphospholipid antibody associated thrombosis, such as coagulation factor mutations like HLA-DR7, DR4, DRw53, DQw7, and C4 null alleles and iinfections like borrelia burgdorferi, treponema, HIV, leptospira and drugs like procainamide, quinidine, chlorpromazine, phenytoin can induce antiphospholipid antibody production.[5,6]

Even normal individuals can have low levels of APLA. All the patients with APLA may not develop APLS. But the association is very strong between the presence of APLA and venous thrombosis, myocardial infarction, and ischemic stroke.[6]

Antibody profile, their type and titres, pre-existing comorbidities, may determine the development of APLS clinically. Positive lupus anticoagulant and high titres of anti-cardiolipin and anti-β2-glycoprotein 1 antibodies poses the highest risk for the development of APLS. In contrast, isolated or intermittent positivity or low titres of anti-cardiolipin or anti-beta-2-glycoprotein I antibodies pose a low risk.[7,8] Patients with SLE, association with risk factors of cardiovascular diseases, history of arterial

thrombosis are also at the increased risk for recurrent thrombosis. Here we are presenting a case of 28-year-old male with isolated PTE, a rare presentation of APS.

CASE REPORT

A 28-year-old male came to our hospital with complaints of right lower back pain for 6 days, which is of compressing type, without any radiation and no specific aggravating or relieving factors. Also, patient had history of cough for 5 days, which is acute in onset with no diurnal or seasonal variation associated with expectoration and hemoptysis for 5 days, mild (<50 ml/day), 8-10 episodes/ day 2-3 ml/episode.

Patient denies having other complaints like fever, weight loss, vomiting, evening rise of temperature, pain abdomen. History of pulmonary tuberculosis 20 years back, for which treatment was completed and repeat smear was found to be negative. Previous history of infection with COVID-19 pneumonia 6 months back and it was managed conservatively. Patient was an occasional smoker and social drinker. No significant family history and childhood history for bleeding.

On general examination, vitals were stable. On systemic examination, reduced air entry bilaterally with no added sounds. Complete blood count was normal. ESR and D-dimer was mildly elevated. Renal and liver function test, electrolytes and coagulation profile were within normal limits.

Sputum AFB was negative. Gram stain showed 10-25 pus cells, occasional gram-positive cocci in pairs, chains, clusters, occasional gram-negative bacilli. Culture sensitivity showed normal flora. Arterial blood gas, electrocardiogram and echocardiography was normal. Computed tomography of thorax showed fibroconsolidatory and fibroatelectatic changes in bilateral lower lobes, probably old infective sequelae (post Koch's).

In view of persistent hemoptysis, CECT with pulmonary angiogram which showed features of acute pulmonary thromboembolism with pulmonary infarcts in the posterior and medial basal segments of the right lower lobe and posterior basal segments of the left lower lobes. Bilateral lower limb venous Doppler showed no deep vein thrombosis changes.

Antinuclear antibody workup was negative. Antiphospholipid antibody workup showed positive for anticardiolipin IgM (5.58) and increased serum homocystein 24.5 (3.7-13.9). Factor V Leiden mutation-wild type (GG) was present.

Anti-β2-glycoprotein 1 antibodies (IgG and IgM) were negative. Our patient had presented with diagnostic as well as therapeutic challenge, who had ongoing hemoptysis and acute pulmonary embolism at the same time. Anticoagulation therapy to prevent thrombosis and procoagulant therapy for hemoptysis.

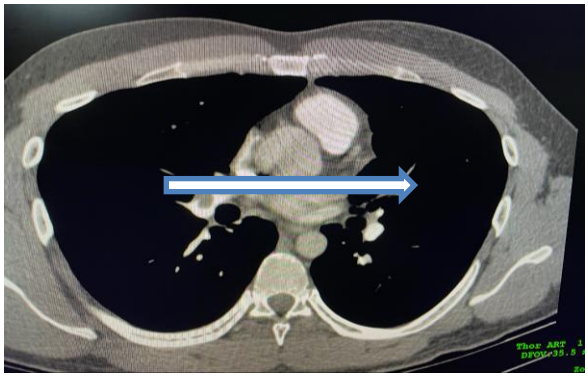

Figure 1: Pulmonary thromboembolism with pulmonary infarct.

DISCUSION

APS is an autoimmune disease which is more common in females than in males.[9] The diagnosis is mainly based on clinical and laboratory findings. Presence of anticardiolipin, anti-β2-glycoprotein 1 and antiphospholipid antibodies are essential for the diagnosis. Our patient was positive for anticardiolipin antibody IgM. Lupus anticoagulant is known to be associated with a higher risk for thrombotic events.[10] Patients presenting with pulmonary embolism can progress to pulmonary infarction in 10%-20% patients, which can result in hemoptysis in 5%-7% of patients.[11] As per revised Sapporo classification criteria, the diagnosis of APLA includes anticardiolipin antibody of IgG and/or IgM isotype in serum or plasma, present in medium or high titre on 2 or more occasions, at least 12 weeks apart, measured by a standardized enzyme-linked immunosorbent assay (ELISA).[12]

In case of patients with history of pulmonary Kochs, active tuberculosis and aspergilloma has to be ruled out in. Common conditions like bronchiectesis, lung abscess, tuberculosis and lung malignancies are the most common causes of massive haemoptysis.[13] The main cause of hemoptysis in pulmonary embolism is ischemic parenchymal necrosis.[14] Certain studies says that most of the cases with hemoptysis originate from the bronchial arteries, only <10% originate from pulmonary circulation and non-bronchial systemic arteries, which may lead to rupture and that manifests as hemoptysis.[15]

Our patient had presented with diagnostic as well as therapeutic challenge, who had ongoing hemoptysis and acute pulmonary embolism at the same time. Anticoagulation therapy to prevent thrombosis and procoagulant therapy for hemoptysis. First line treatment for hemoptysis is tranexamic acid, antifibrinolytic agent-an inhibitor of plasminogen activation. Tranexamic acid may reduce both the quantity and duration of hemoptysis and there is risk of short-term thromboembolic complications.[16] Hemoptysis was controlled with tranexemic acid and on further evaluation, patient was found to have pulmonary thrombo-embolism without any

respiratory discomfort. Management of choice for patients with pulmonary embolism is anticoagulation, which prevents both early mortality death and recurrence.[17] Unfractionated heparin (UFH) is given with monitoring of the activated partial thromboplastin time for anticoagulation when there is a risk of recurrent haemorrhage. In view of thrombo-embolism, patient was started on low molecular weight heparin, overlapped with vitamin K antagonist.

CONCLUSION

Isolated APS presenting in young and male population is rare. Initial presentation of PTE with hemoptysis remains a challenge in therapeutic aspects. Patient should continue vitamin K antagonist as a life-long anticoagulation therapy.

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