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CASE REPORT

A Case Report on Antiphospholipid Antibody Syndrome with Chronic Pulmonary Embolism Secondary to Deep Vein Thrombosis and Thrombocytopenia

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Abstract: Antiphospholipid antibody syndrome (APS) is a multisystemic autoimmune disorder characterized by the persistent presence of antiphospholipid antibodies and a high risk of thrombotic events and adverse pregnancy outcomes. We report a case of a 42-year-old female patient with APS who developed chronic pulmonary embolism secondary to deep vein thrombosis (DVT) and thrombocytopenia. The patient presented with shortness of breath, cough with expectoration, black stools, and a history of DVT, abortion, and decreased platelet count. Physical examination revealed pallor, edema, and blackish discoloration of the right lower limb. Laboratory investigations showed anemia, thrombocytopenia, and an elevated reticulocyte count. Imaging studies, including ultrasonography and computed tomography (CT) scan, confirmed the presence of hepatic hemangioma and multiple pulmonary thrombi. The patient was diagnosed with APS with chronic pulmonary embolism secondary to DVT and thrombocytopenia. Treatment included anticoagulants (rivaroxaban and aspirin), diuretics (furosemide), antacids (pantoprazole), and symptomatic relief (cough syrup). This case highlights the importance of early recognition and prompt management of APS to prevent life-threatening thrombotic complications and adverse pregnancy outcomes.

Keywords: Antiphospholipid antibody syndrome; Deep Vein Thrombosis; Thrombocytopenia; Hepati hemangioma; Pulmonary thrombi.

1. Introduction

Antiphospholipid antibody syndrome (APS) is a systemic autoimmune disorder characterized by the persistent presence of circulating antiphospholipid antibodies (aPL) and a high risk of thrombotic events, such as deep vein thrombosis (DVT), pulmonary embolism (PE), and stroke, as well as adverse pregnancy outcomes, including recurrent miscarriages and stillbirths. [1] APS can be primary (occurring in the absence of any underlying autoimmune disease) or secondary (associated with other autoimmune disorders, such as systemic lupus erythematosus (SLE)). [2] The pathogenesis of APS is complex and not fully understood, but it is believed to involve the interaction of antiphospholipid antibodies with cellular components, leading to a pro-coagulant state and endothelial cell activation. The antiphospholipid antibodies implicated in APS include lupus anticoagulant (LA), anticardiolipin antibodies (aCL), and anti-β2-glycoprotein 1 antibodies (aβ2GP1). [3] These antibodies can interfere with the coagulation cascade, disrupt the anticoagulant properties of various proteins, and promote the activation of endothelial cells, platelets, and monocytes, ultimately leading to a thrombotic tendency. [4]

The clinical manifestations of APS are diverse and can involve multiple organ systems. Vascular thrombosis is a hallmark feature, with DVT and PE being the most common manifestations. Patients may also experience stroke, transient ischemic attack, or other arterial thrombotic events. In addition, APS can lead to pregnancy complications, such as recurrent miscarriages, intrauterine growth restriction, and preterm delivery. [5] The diagnosis of APS is based on both clinical and laboratory criteria. The clinical criteria include the presence of vascular thrombosis or pregnancy morbidity, while the laboratory criteria involve the detection of antiphospholipid antibodies (LA, aCL, or aβ2GP1) on two or more occasions at least 12 weeks apart.The management of APS is primarily focused on preventing and treating thrombotic events and managing pregnancy-related complications. Anticoagulant therapy, such as warfarin, heparin, or direct oral anticoagulants (DOACs), is the mainstay of treatment for the prevention and management of thrombotic events. In cases of obstetric APS, a combination of low-dose aspirin and heparin may be used to improve pregnancy outcomes. [6]

Despite advances in the understanding and management of APS, the condition remains a significant challenge, and complications can be life-threatening if not recognized and treated promptly. Early diagnosis and appropriate management are crucial to prevent

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adverse outcomes and improve the quality of life for patients with APS.[7] The objective of this work is to present a case report of a patient with APS who developed chronic pulmonary embolism secondary to DVT and thrombocytopenia, highlighting the importance of early recognition and prompt management of this complex autoimmune disorder.

2. Case Report

2.1. Presentation

A 42-year-old female patient was admitted to the general medicine ward with complaints of shortness of breath (grade 2 mMRC), aggravated by walking and physical activities, cough with mucoid expectoration, black stools, and amenorrhea for 4 months. The patient had a history of similar complaints, hospitalization due to decreased platelet count 3 years ago, and one abortion in the 8th month of gestation. Additionally, she had a history of DVT in the right leg 3 years ago, for which she was on rivaroxaban, coumarin, and deflazacort. On physical examination, the patient appeared pale, with edema and blackish discoloration in the right lower limb. Vital signs were within normal limits, except for a respiratory rate of 28 breaths per minute.

Laboratory investigations revealed anemia (hemoglobin 7.8 g/dL), thrombocytopenia (50,000 cells/cumm), elevated reticulocyte count (15%), and low total protein (3.2 g/dL). Imaging studies, including ultrasonography and computed tomography (CT) scan, showed hepatic hemangioma in the right lobe of the liver and multiple pulmonary thrombi in the right upper lobar artery, respectively. Based on the clinical presentation, laboratory findings, and imaging studies, the patient was diagnosed with APS with chronic pulmonary embolism secondary to DVT and thrombocytopenia [9, 10]

2.2. Treatment

The patient was treated with furosemide (20 mg IV twice daily) for edema management, pantoprazole (40 mg orally once daily) for acidity, aspirin (75 mg orally once daily) and rivaroxaban (20 mg orally once daily) for anticoagulation and prevention of blood clots, and a cough syrup containing chlorpheniramine and dextromethorphan hydrobromide (10 ml orally three times daily) for symptomatic relief.

2.3. Risk factors

Several risk factors have been identified for the development of APS and its associated complications. These include genetic predisposition, environmental triggers (such as infections or certain medications), and the presence of other autoimmune disorders like SLE. [8] In the case presented, the patient had a history of DVT and a previous abortion, which are known risk factors for APS. Additionally, the presence of thrombocytopenia and the development of chronic pulmonary embolism further increased the risk of adverse outcomes

2.4. Prevention strategies

Prevention of thrombotic events and adverse pregnancy outcomes is a crucial aspect of managing APS. Several strategies [9, 10] can be employed to reduce the risk of complications:

2.4.1. Anticoagulation therapy

Appropriate anticoagulation is the cornerstone of preventing and treating thrombotic events in APS. Warfarin, heparin, and direct oral anticoagulants (DOACs) like rivaroxaban are commonly used for this purpose.

2.4.2. Antiplatelets

Low-dose aspirin may be prescribed in combination with anticoagulants to further reduce the risk of thrombosis.

2.4.3. Pregnancy management

In patients with obstetric APS, close monitoring and treatment with low-dose aspirin and heparin may improve pregnancy outcomes and reduce the risk of complications like miscarriage, preterm delivery, and intrauterine growth restriction.

2.4.4. Lifestyle modifications

Maintaining a healthy lifestyle, including regular physical activity, a balanced diet, and smoking cessation, can contribute to the overall management of APS and reduce the risk of complications.

2.4.5. Patient education

Educating patients about the risks associated with APS, the importance of medication adherence, and the recognition of potential complications can empower them to actively participate in their care and seek medical attention promptly when needed.

2.4.6. Regular monitoring

Regular follow-up and monitoring of antiphospholipid antibody levels, coagulation parameters, and organ function can aid in the early detection of complications and guide appropriate treatment adjustments

3. Discussion

APS is a complex autoimmune disorder characterized by the presence of antiphospholipid antibodies (aPL), including lupus anticoagulant (LA), anticardiolipin antibodies (aCL), and anti-β2-glycoprotein 1 antibodies (aβ2GP1). The syndrome is associated with an increased risk of thrombotic events, such as DVT, PE, and stroke, as well as pregnancy complications like recurrent miscarriages and stillbirths. [11] In this case, the patient presented with symptoms of chronic pulmonary embolism, likely secondary to DVT, which is a known complication of APS. The laboratory findings of anemia, thrombocytopenia, and elevated reticulocyte count further supported the diagnosis of APS. Imaging studies confirmed the presence of pulmonary thrombi and hepatic hemangioma, which can be associated with APS. The treatment approach for APS involves anticoagulation to prevent and manage thrombotic events, as well as supportive care for symptom management. In this case, the patient was prescribed rivaroxaban and aspirin for anticoagulation, furosemide for edema management, pantoprazole for acidity, and a cough syrup for symptomatic relief. It is essential to note that APS can have severe consequences if left undiagnosed and untreated, including life-threatening thrombotic events and pregnancy complications. Early recognition and management of APS are crucial to prevent these complications and improve patient outcomes.

4. Conclusion

This case report highlights the importance of prompt diagnosis and management of APS, particularly in patients presenting with thrombotic events and pregnancy complications. Appropriate anticoagulation therapy and supportive care are crucial in the management of APS to prevent further complications and improve patient outcomes.

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