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A Case Report of Catastrophic Antiphospholipid Syndrome with Libman-Sacks Presenting as Interstitial Pneumonia

Chassidy Sumler Martin, MS, Rocco Cannistraro, MD
DeBusk College of Osteopathic Medicine, Lincoln Memorial University, Knoxville, TN 37932

Abstract

Introduction: Antiphospholipid syndrome (APS) is an autoimmune condition characterized by vascular thromboses and a positive antiphospholipid antibody. Catastrophic antiphospholipid syndrome (CAPS) is a rare disease that often results in death. CAPS is the most severe form of APS, which can develop in a short period of time and occurs in less than 1% of people with APS. CAPS involves multiple organs simultaneously with diffuse microvascular and macrovascular involvement.

Case Presentation: Here, we present a case of catastrophic antiphospholipid syndrome presenting as interstitial pneumonia that rapidly progressed to acute renal failure, acute ischemic cerebral infarcts, cardiac valvular vegetations, and heart failure.

Conclusion: This case report aims to bring awareness of prompt medical suspicion and treatment of CAPS in hopes of improving disease outcomes.

Introduction

Antiphospholipid syndrome (APS) is an autoimmune condition characterized by vascular thromboses and a positive antiphospholipid antibody. Catastrophic antiphospholipid syndrome (CAPS) is a rare disease that often results in death. CAPS is the most severe form of APS, which can develop in a short period of time and occurs in less than 1% of people with APS. CAPS involves multiple organs simultaneously with diffuse microvascular and macrovascular involvement. Females are more likely to be affected with CAPS than males, and the age range is broad. Early diagnosis and aggressive therapy are needed to lower morbidity and mortality.

Here we present a case of catastrophic antiphospholipid syndrome presenting as interstitial pneumonia that rapidly progressed to acute renal failure, acute ischemic cerebral infarcts, cardiac valvular vegetations, and heart failure.

Case Summary

A 42-year-old male presents to the emergency department complaining of shortness of breath that began one week prior. He stated that he had a syncopal episode and was not sure what caused the event. Upon arrival, the patient's O2 stats were 85%, heart rate was 145, and blood pressure was 94/69. A chest x-ray was performed and showed florid pulmonary edema and interstitial pneumonia. Lab values were drawn, and the patient had a pH of 7.5 with elevated neutrophils and lactic acid. A CT without contrast of the head was later done and showed an acute, subacute right MCA infarct, and neurology was consulted. After the patient was admitted, the first neurological exam showed no neurological deficits. Cardiology was then consulted, and a transthoracic echocardiogram showed mild concentric left ventricular hypertrophy, mild to moderate mitral regurgitation, and a large mobile mass attached to the atrial surface of the posterior leaflet measuring 3.6 cm x 1.6 cm. Sputum and blood cultures were sent, and the patient was started on ceftriaxone and azithromycin. Several blood cultures were taken, and the patient consistently showed negative cultures. Over the next few days, the patient began to decline rapidly. The patient was later intubated, and nephrology was consulted for rapid renal decline. On day eight, the patient had a BUN of 73, Creatinine of 2.93, Prothrombin of 40.4, a partial thrombin time of 71.7, WBC of 36.4, lactate dehydrogenase level of 530, and a Hemoglobin of 9.5. He also began to show evidence of hepatic dysfunction with worsening transaminases. His AST was 495, and his ALT was 304. After several more negative blood cultures, rheumatology was consulted. The patient tested positive for antiphospholipid antibodies and had a low level of C4. The patient was later diagnosed with the catastrophic antiphospholipid syndrome with Libman Sacks endocarditis. A repeat Transthoracic echocardiogram was performed and showed global systolic function with a left ventricular EF of 60%, severe mitral regurgitation, and mild tricuspid regurgitation. The patient was started on heparin for anticoagulation, plasma exchange daily for five days, pulse solumedrol 1mg IV daily x3 days, followed by solumedrol 1mg/kg/d. Approximately three weeks after being admitted, due to the patient's rapid renal decline, liver failure, and worsening coagulopathy, the patient succumbed to his illness

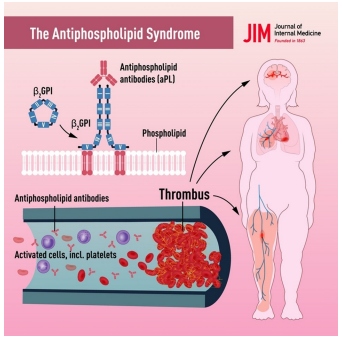

Figure 1 . Antiphospholipid Syndrome

Discussion

CAPS is a systemic autoimmune disease that is fatal in 20% of patients with APS. CAPS usually evolves rapidly and can cause multiple organ thromboses. Patients with systemic manifestations involving numerous organs that develop simultaneously over a short period of time should have a high suspicion of APS and possible CAPS. Aggressive therapy should be initiated in high suspicion of CAPS. Even with incomplete clinical information, the treatment should be given and revised based on changing clinic status. In 2018 a report showed that patients with triple therapy of anticoagulation, glucocorticoids, and plasma exchange were associated with higher survival rates. We hope that our case report will help better assess antiphospholipid-positive patients with multiple organ failures with the goal of preventing and reversing the fatal disease of CAPS.

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