

CLINICAL VIGNETTE

40 Year Old Female Presenting with ST Elevation Myocardial

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Introduction

Antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterized by the presence of arterial and/or venous thrombosis, recurrent fetal loss, thrombocytopenia, and elevated titers of antiphospholipid antibodies. Catastrophic APS or CAPS occurs in less than 1% of patients with APS¹. CAPS is defined by involvement of three or more organs, systems, or tissues, a rapid presentation (< 1 week), small vessel occlusion in at least one organ or tissue with platelet thrombi, and laboratory confirmation of antiphospholipid antibodies (either lupus anticoagulant or anticardiolipin antibodies) at least 6 weeks apart¹).

The most common causes of CAPS includes infections (35%), surgery (13%), neoplasia (8%), anticoagulation cessation (8%), and idiopathic (35%)¹. Establishing an effective treatment strategy has remained elusive given the relative rarity of the disease and the consequent inability to perform large-scale studies comparing various treatment regimens. The first-line therapy remains anticoagulation, and it has been shown to have a significant effect on reducing mortality (36.9% v. 77.8%, $p < 0.0001$)². Other strategies, such as plasma exchange, intravenous gamma globulins, high dose corticosteroids and other immunosuppressants have been used alone or in combination with variable success.

Survival with CAPS depends on the immediate recognition of the condition, reversal of any precipitating factors, and the initiation of aggressive therapy. Despite timely treatment, the mortality rate remains extremely high (30-48%)^{2,3}.

Case Report

A 40 year-old African American woman with a past medical history of hypothyroidism presented to the emergency department for acute-onset chest pain. Patient stated that the pain began suddenly thirty minutes prior to presentation, was substernal, rated 9/10, was pressure-like, and radiated to her left arm.

She was taking Levothyroxine 88mcg daily, but denied other prescription medications, tobacco, alcohol or illicit drugs. During the course of the brief interview, the patient became increasingly dyspneic and lethargic. An initial electrocardiogram (Figure 1) was performed and demonstrated an ectopic atrial rhythm at a rate of 96 beats per minute with 1-3mm ST segment elevations in the anterolateral leads and aVR, with right bundle and left anterior fascicular blocks.

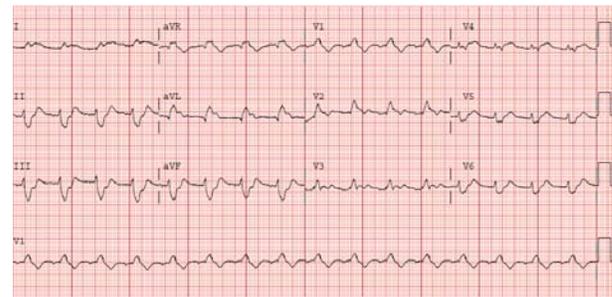


Figure 1. Patient's presenting ECG demonstrating an Anterior STEMI with right bundle branch block and left anterior fascicular block.

Physical Examination

The patient was diaphoretic with a temperature of 99° F, blood pressure 92/56 mmHg, respiratory rate 30 breaths per minute and oxygen saturation of 96% on a non-rebreathing facemask. PE was significant for jugular venous distention, S₃ gallop, a 3/6 holosystolic murmur at the apex which radiated into the axilla, diffuse rales throughout the posterior lung fields, and trace bilateral lower extremity pitting edema.

Emergency Room Course and Diagnostic Studies

In the emergency department, the patient received aspirin 325mg orally, 2 sprays of nitroglycerin 0.4mg sublingually and a 250ml bolus of normal saline intravenously.

The patient was consented and transported within 30 minutes to the cardiac catheterization laboratory for treatment of an anterior ST-elevation myocardial infarction (STEMI). Access was obtained in the right common femoral artery, and the opening aortic systolic blood pressure was measured as 72mmHg. The patient was started on a continuous dopamine infusion. Contrast angiography demonstrated a large thrombus occluding the left main artery proximal to the bifurcation of the left anterior descending (LAD) and circumflex arteries (Figure 2). TIMI II flow was noted distal to the obstructing thrombus. A Prowater wire was inserted into the left main artery to dislodge the thrombus and restore TIMI III flow (Figure 3). The patient remained hypotensive with a systolic blood pressure of 65mmHg. An intra-aortic balloon pump was placed via left femoral artery access. A pulmonary artery catheter was inserted into the left common femoral vein for hemodynamic monitoring.

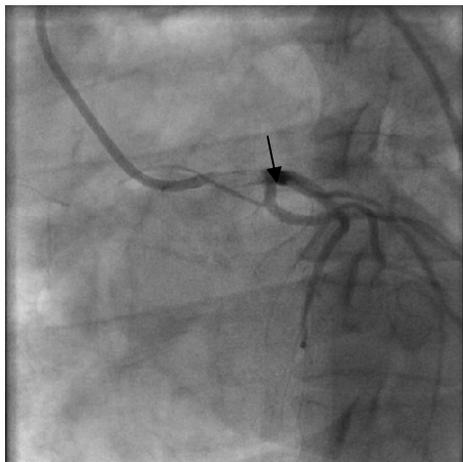


Figure 2. A left anterior oblique-cranial view during coronary angiography demonstrates a thrombus (arrow) in the proximal left main artery with distal TIMI II flow.



Figure 3. A right anterior oblique-cranial view during coronary angiography showing distal embolization of the thrombus with restoration of proximal flow with no evident arterial plaque.

The patient remained hypotensive despite maximal dose continuous dopamine infusion and the subsequent additions of norepinephrine and epinephrine infusions. Shortly thereafter, the patient's rhythm decompensated into ventricular fibrillation, requiring two rounds of countershock electrical cardioversion. The patient was successfully intubated, resuscitated and started on a lidocaine infusion. Repeat angiography demonstrated a widely patent left main artery with no evidence of ulceration or plaque. A small filling defect was noted in the apical LAD. The patient remained tenuous with a systolic blood pressure of 85 mmHg while on multiple pressors and she was transferred to the Cardiac Intensive Care Unit for further monitoring and treatment.

A portable chest radiograph is shown in Figure 4. Transthoracic echocardiogram showed an ejection fraction of 15-20%, with anterolateral and apical akinesis and moderate septal hypokinesis.



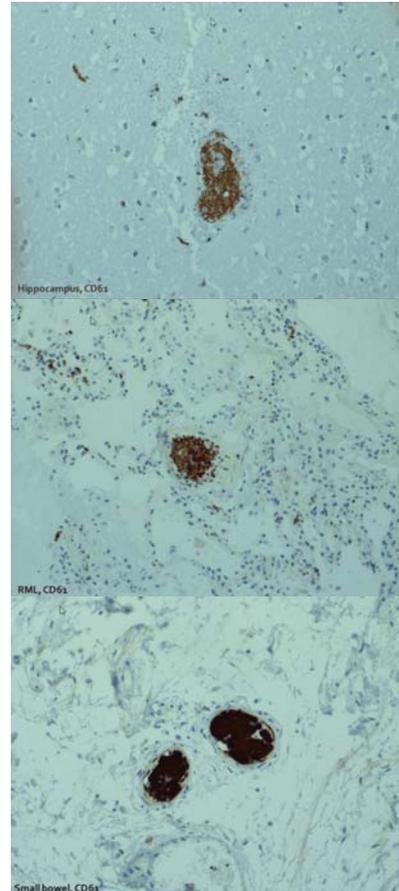
Figure 4. Portable chest radiography demonstrating opacification of the left lung field with an enlarged mediastinal silhouette.

Laboratory studies were notable for an elevated white blood cell count of 12,500 with a left-shift (20% bandemia, 35% segmented neutrophils, 41% lymphocytes), and normal hemoglobin and platelet counts of 13.1 and 157,000, respectively. The patient had a positive lupus anticoagulant, with a weakly positive hexagonal phase confirmation test. PTT-LA screen (50 seconds) and DRVVT (45 seconds) were elevated. Activated protein C, C₃ and C₄ complement levels were low. Fibrinogen, D-dimer, and fibrinogen degradation products (FDP) were all elevated. Anticardiolipin and anti-nuclear antibodies were negative. The labs were trended and on hospital day 3, the patient's hemoglobin gradually decreased to 6.8 despite multiple transfusions and the platelet count dropped to 48,000.

During the course of her hospitalization, the patient developed renal failure, hepatic failure, and remained non-responsive. A neurologic exam performed on hospital day 3 noted that her pupils were fixed and dilated at 5mm. On hospital day 4, the patient had a PEA arrest and expired.

Diagnosis: Initial Presentation of Catastrophic Antiphospholipid Syndrome (CAPS) as an Anterior ST Elevation Myocardial Infarction (STEMI)

Autopsy: An autopsy was performed and demonstrated multiple intravascular CD61 positive platelet thrombi involving the vessels of the myocardium, brain, lung, small bowel, uterus, ovaries and thyroid (Figure 5).



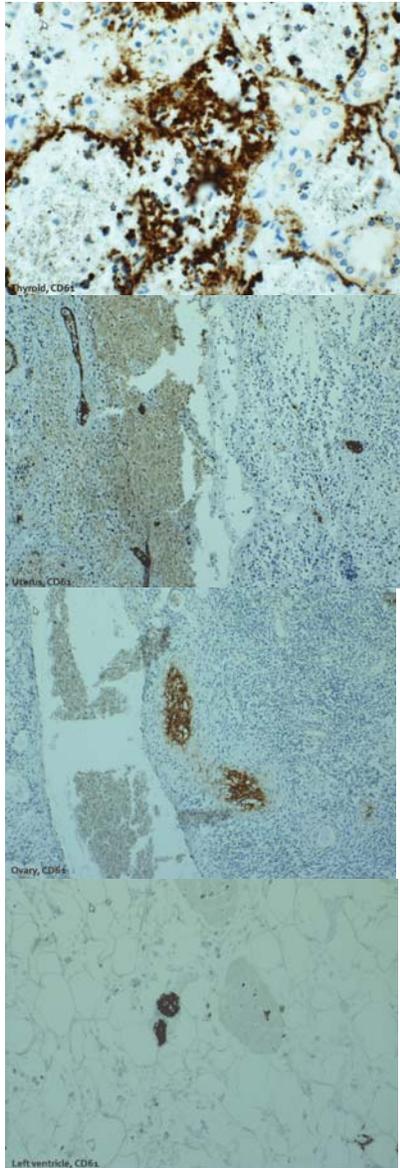


Figure 5. Histopathologic analysis exhibiting intravascular CD61 stain positive platelet thrombi in the vessels of brain, lung, small bowel, thyroid, uterus, ovary, and myocardium.

Based on the 2003 CAPS Guidelines¹, the patient met all four criteria for the diagnosis of Catastrophic Anti-Phospholipid Syndrome (CAPS), however, as the patient had no prior diagnosis of Antiphospholipid Syndrome (APS) and expired before a six week confirmatory test could be performed, this case is considered probable CAPS given the absence of the requisite confirmatory lupus anticoagulant serology after the initial positive test. The presence of CD61 positive platelets supports the diagnosis of CAPS.

Pearls:

- 1) Primary APS is more common in women than men and less than 1 % of APS patients develop CAPS.
- 2) Myocardial infarction is a very rare presentation in CAPS (< 1%) , however CAPS should be considered in young patients with myocardial infarction without coronary atherosclerosis⁴.
- 3) Due to the life threatening nature of CAPS the clinician needs to have high clinical suspicion to diagnosis it early. The diagnosis was not suspected in this case prior to autopsy.
- 4) First line therapy remains anticoagulation which has been shown to reduce mortality.
- 5) Even with prompt recognition and management, the mortality is extremely high (30-48%).

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Submitted on November 6, 2012