A Rare Cause of Acute Coronary Syndrome: Catastrophic Antiphospholipid Syndrome

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Abstract

The antiphospholipid syndrome (APS) is characterized by the occurrence of venous or arterial thrombosis or of specific pregnancy morbidity, in the presence of laboratory evidence of antiphospholipid antibodies. APS was rarely seen in our clinical practice as a cardiologist. In this case, a 26-year-old man was admitted with acute peripheral arterial occlusion and ST-segment elevation myocardial infarction. He was undergone aspiration thrombectomy and right aorta-popliteal bypass surgery. During medical treatment ischemic stroke was determined. Vascular thrombosis affecting more than two organs, distinguishes the case from others. It shows that we should keep in mind APS, when extensive vascular thrombosis are seen in the clinical practice.

Learning objective: Acute coronary syndromes seen in the clinical practice are caused by atherosclerosis more often than not. The antiphospholipid syndrome (APS) is one of a rare cause of acute coronary syndromes. This case is a brief review of the APS.

Keywords: Acute Coronary Syndrome (ACS); Antiphospholipid Syndrome (APS); Venous Thromboembolism (VTE); Acute peripheral arterial occlusion

Case Report

A 26-year-old man, who was diagnosed with left lower extremity deep venous thrombosis two weeks ago, was seen in the emergency department of our hospital because of right lower extremity pain. On presentation, he was not taking any anticoagulant therapy. He rated the pain at 10 on a scale of 0 to 10, with 10 indicating the most severe pain. There was neither sensory loss nor motor deficit. The blood pressure was 100/70 mmHg in the right arm and 95/65 mmHg in the left arm, and the other vital signs were normal. The lungs were clear. Although he didn’t describe any chest pain, the 12-lead electrocardiogram showed ST-segment elevation in leads II, III, aVF, V4 through V6 (Figure 1). The echocardiogram showed impaired inferior and inferolateral wall motion (Ascending aorta was 3.1 cm and there was no aortic regurgitation). 300 mg chewable acetylsalicylic acid and 600 mg clopidogrel were administered. The coronary angiography showed totally occluded right coronary artery with absent flow (Figure 2) and multiple filling defects suggesting intracoronary thrombus in left anterior descending artery (Figure 3). Prompt revascularization with aspiration thrombectomy, restored normal antegrade coronary perfusion. After aspiration thrombectomy, we performed right lower extremity angiogram and thrombus was seen in the femoral artery. And tirofiban and heparin infusions were administered because of massive thrombus burden. After the procedure >70% ST-segment resolution was obtained. After two hours, his lower extremity pain was intensified and sensory loss was determined. He was undergone aorta-popliteal bypass surgery. There was no hemodynamic compromise for 13 hours after aspiration thrombectomy. His urine output was more than 50 cc/h for fifteen hours. While continuing anticoagulant therapy (Heparin), stroke was suspected because of right upper extremity paresthesia at 14 hours after presentation. Cranial CT was performed, and hypodense lesion (28*15 mm) was determined. After two hours, his lower extremity pain was intensified and sensory loss was determined. He was undergone aorta-popliteal bypass surgery. There was no hemodynamic compromise for 13 hours after aspiration thrombectomy. His urine output was more than 50 cc/h for fifteen hours. While continuing anticoagulant therapy (Heparin), stroke was suspected because of right upper extremity paresthesia at 14 hours after presentation. Cranial CT was performed, and hypodense lesion (28*15 mm) was determined.

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mm) in the parietal lobe was determined compatible with infarction (Figure 4). Meanwhile the thoracoabdominal CT angiography was performed for pulmonary embolism, aortic dissection and there was neither thrombus formation nor dissection. And there was no thrombus formation in the left femoral artery. Despite of the fact that the next-of-kin denied permission. He had neither history of oral aphthous ulcers nor painful genital ulcerations. Due to suspicion of thrombophilia, the blood samples were drawn at presentation. The percentage of protein C and protein S levels were normal (The reference range 70-140%). Antithrombin level was 38 mg/dL (Reference range 17-39 mg/dL). Anti-nuclear antibody, HLA-B51 and anti-cardiolipin antibodies (ACA IgM 1.24 MPL U/mL, ACA IgG GPL U/mL, the reference range <15) were negative. His LAIgG (39.2 U/mL, the reference range: 0.1-15) and anti-β2 glycoprotein I IgG (145 U/mL, the reference range 0-100 U/mL) levels were elevated. Owing to hemodynamic instability; cranial magnetic resonance imaging couldn’t be performed. Ventricular fibrillation was determined and cardiopulmonary resuscitation was performed, but the patient was died 15 hours after admission. Autopsy wasn’t performed, on account of the fact that the next-of-kin denied permission.

**Discussion**

The antiphospholipid syndrome (APS) is characterized by the occurrence of venous or arterial thrombosis or of specific pregnancy morbidity, in the presence of laboratory evidence of antiphospholipid antibodies. APS occurs either as a primary condition or in the setting of an underlying disease, usually systemic lupus erythematosus (SLE). The Sapporo APS classification criteria were replaced by the Sydney criteria in 2006 [1]. Based on the most recent criteria, classification with APS requires one clinical and one (IgG and/or IgM anticardiolipin in moderate or high titer, antibodies to β2-glycoprotein I of IgG or IgM isotype at a titer >99th percentile, Lupus anticoagulant activity detected according to published guidelines) laboratory manifestation [1]. There are 3 distinct APS disease entities: primary (the absence of any comorbidity), secondary (when there is a pre-existing autoimmune condition, most frequently systemic lupus erythematosus, SLE), and catastrophic (when there is simultaneous multi-organ failure with small vessel occlusion). Clinical presentation of catastrophic form of APS appears as mixed thrombosis of arterial and venous circulation with varying degrees of thrombocytopenia, hemolytic anemia, deep vein thrombosis, transient ischemic attacks, pulmonary embolism, and spontaneous abortions [2]. The International Consensus Statement is commonly used for Catastrophic APS (CAPS) diagnosis [3]. Based on this statement, Definite CAPS diagnosis requires; vascular thrombosis in three or more organs or tissues and, development of manifestations simultaneously or in less than a week and, evidence of small vessel thrombosis in at least one organ or tissue and, laboratory confirmation of the presence of aPL [3]. Fortunately, antiphospholipid syndrome was rarely seen in our clinical practice as a cardiologist [4]. Patients with acute coronary syndrome can be treated by angioplasty or stenting, but in our patient none of them were not performed because he had normal coronary arteries with severe thrombus burden (Figure 2) and intervention would have led to more complications and recurrent stent thrombosis.

Assuming the patient had been discharged, his medication must have included not only dual antiplatelet therapy but also warfarin therapy. In spite of the coronary intervention and the aorta-popliteal bypass surgery, every so often undesired results are inevitable owing to extensive thrombus burden. To conclude, we should keep in mind antiphospholipid syndrome, when extensive vascular thrombosis is seen in the clinical practice.

**References**


