Antiphospholipid Syndrome Presenting as Intracardiac Thrombus With Pulmonary Embolism

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The presence of antiphospholipid antibodies is associated with arterial and venous thrombosis. A young female with initial presentation of dyspnea and cough that lasted for days is reported. A computed tomographic scan of her chest and echocardiography showed features of thrombus formation over the right atrium, complicated with pulmonary thromboembolism. Antiphospholipid syndrome was diagnosed according to elevated activated partial thromboplastin time, high serum titers of anticardiolipin antibody, and the presence of intracardiac thrombus with pulmonary embolism. This thrombus was subsequently removed successfully with surgical intervention, and the patient’s recovery was uneventful. (Circ J 2005; 69: 1290–1292)

Key Words: Activated partial thromboplastin time; Anticardiolipin antibody; Antiphospholipid syndrome; Intracardiac thrombus

The features of antiphospholipid syndrome (APS) are recurrent arterial and venous thrombosis, recurrent spontaneous abortion, and thrombocytopenia. Coexistence of intracardiac thrombus and pulmonary embolism in APS is relatively rare. We report a patient with this condition whose initial presentation of shortness of breath.

Case Report

A 22-year-old woman was diagnosed with atrial septal defect (secundum type) and underwent surgical repair in 1994. Her general condition was good after the surgery. She denied any intravenous drug abuse or tobacco use, and there was no known family history of thrombotic disease. Three days before the present admission, she had shortness of breath and a dry cough, and she presented at the emergency room for further evaluation. On examination, she was in respiratory distress, with tachypnea (27 breaths/min), blood pressure of 146/84 mmHg, tachycardia (133 beats/min), and body temperature measuring 36.8°C. The jugular vein was not engorged, and cardiac auscultation revealed a grade II/VI systolic murmur over the right lower sternal border. Chest auscultation showed mild fine crackles over both of the lower lung fields. Arterial blood gas analysis under oxygen 2 L/min revealed pH of 7.486; pO₂, 87 mmHg; pCO₂, 28.8 mmHg, and bicarbonate 21.3 mmol/L. There was sinus tachycardia without significant ST-T segment change on the electrocardiogram, and chest X-ray did not show any remarkable findings. However, transthoracic echocardiography showed a floating mobile mass adhered to the free wall of the right atrium by a slender stalk closely attached to the interatrial septum. This lesion was intermittently protruding into the right ventricle during diastole (Fig 1A,B). There was neither a morphological abnormality nor vegetative formation on the valves, although tricuspid regurgitation was observed. Subsequent chest computed

Fig 1. (A) Transthoracic echocardiogram and (B) transthoracic echocardiogram showing floating thrombus (arrow) in the right atrium (RA) and intermittently protruding into the right ventricle (RV). LV, left ventricle.
Intracardiac Thrombus in Antiphospholipid Syndrome

Antiphospholipid antibodies (aPL), namely LA and anticardiolipin, are a family of autoantibodies directed predominantly against negatively charged phospholipids. They were first identified in patients with syphilis and have been reported in as many as 40% of adult systemic lupus erythematosus (SLE) patients. However, aPL has also been associated with some other connective tissue diseases or infections, such as HIV. The principal clinical manifestations of APS include venous and arterial thrombosis, thrombocytopenia, and recurrent abortions. The serological diagnosis of APS is based on the elevated titer of LA or anticardiolipin antibodies, as in the present patient.

The clinical features of the APS are various, but the most common clinical presentation is DVT, which accounts for 32% of cases. The occurrence of pulmonary thromboembolism is approximately 9% in APS. The cardiac manifestations of APS included myocardial infarction, pericardial effusion, myocardial infarction, and coronary artery thrombosis, but the most common manifestation is valvular abnormalities, ranging from 11.6 to 32% of cases. However, the occurrence of a mobile intracardiac thrombus as initial presentation of APS is extremely rare; only those in the akinetic area of ventricle had been reported. In addition, there are some reports of thromboembolic events in patients with primary APS complicated by Libman-Sacks endocarditis. Libman-Sacks endocarditis might also occur in patient with SLE and APS. The mitral and tricuspid valves are most often affected with fibrinoid necrosis; mucoid degeneration; and subsequent development of small fibrous, sterile vegetations on either side of the valve leaflets. However, there was no histological evidence of Libman-Sacks endocarditis in the present patient. The mechanism of hypercoagulability in patients with aPL is presently unknown, and the clinical presentation of APS with intracardiac thrombosis is variable. Lim et al reviewed the clinical manifestations of 19 APS patients with intracardiac thrombi and the major clinical findings included moderate thrombocytopenia (<75 × 10^9/L) in 50% of the cases and underlying structural cardiac abnormalities in 20%. Monica et al reported that the detection of LA and IgG anticardiolipin antibodies at medium or high titers helps to identify APS patients at risk for thrombosis. Although the exact mechanism of intracardiac thrombus formation in APS is unclear, there were some reports of this rare presentation of APS. Julio and Carmen suggested that the endocardial surface might be an important site for thrombus formation in patients with circulating aPL, because these antibodies, in the presence of other hemostatic defects, will abolish the balance between thrombosis and fibrinolysis, and might change the endocardial surface factors so that clot formation is promoted. Coppock et al speculated that an abnormal intracardiac blood flow pattern might contribute to thrombosis; and Kaplan et al hypothesized that diffuse ventricular dysfunction might predispose to the formation of intracardiac thrombus.

Intracardiac thrombosis is a potentially life-threatening, but treatable manifestation of APS. Thrombus formation can cause pulmonary and systemic embolic events. Initial management usually requires surgical removal of the thrombus, which must be differentiated from intracardiac myxoma. The maintenance treatment for APS with thrombotic events is empirical according to the patient’s clinical situation and the size, shape, and location of the thrombus. Furthermore, 50% of APS patients are at risk of
developing recurrent embolic events so intensive anticoagulation treatment and immunosuppressive therapy in serious cases is recommended. Recently, a study comparing moderate- and high-intensity warfarin for patients with APS showed that high-intensity warfarin was not superior for thromboprophylaxis (hazard ratio = 3.1, p = 0.15). The absolute risk of recurrent thrombosis was low if warfarin therapy was targeted to an international normalized ratio of 2.0–3.0.

In conclusion, although it is uncommon, the possibility of coexistence of right heart thrombus should be considered in patients with APS presenting with pulmonary embolism. Early surgical removal of the thrombus and adequate maintenance anticoagulation therapy is needed for the prevention of recurrent intracardiac thrombotic events.

References


