Antiphospholipid antibody syndrome leading to massive pulmonary embolism and sudden death

Yoğun pulmoner emboli ve ani ölüme yol açan antifosfolipid antikor sendromu

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Antiphospholipid antibody syndrome is associated with venous and arterial thromboembolism. Coexistence of pulmonary embolism and intracardiac thrombus is rarely encountered. A 33-year-old male patient presented with severe dyspnea three months after surgery for acute arterial embolism. On physical examination, blood pressure was 80/60 mmHg and breath sounds were weaker in the lower zone of the left lung. Severe lower limb edema was noted. On cardiac auscultation, the third heart sound was elicited. Electrocardiography showed only a sinusal tachycardia. Transthoracic echocardiography revealed a huge thrombus in the right atrium and another thrombus in the main pulmonary artery. Hematological analysis showed a high titration of antiphospholipid antibodies. A diagnosis of massive pulmonary embolism was considered. During preparation for emergency operation, the patient developed cardiovascular collapse, which did not respond to cardiopulmonary resuscitation.

Key words: Antiphospholipid syndrome/complications; echocardiography; pulmonary embolism/etiology; venous thrombosis/etiology.

Antifosfolipid antikor sendromunda venöz ve arteryel tromboz görülebilir. Pulmoner emboli ve intrakardiyak trombüs birlikteliğine çok nadir rastlanır. Otuz üç yaşında bir erkek hasta, akut arteryel embolizm nedeniyle geçirdiği ameliyattan üç ay sonra ciddi nefes darlığı yakınmasıyla başvurdu. Fizik muayenede, kan basıncı 80/60 mmHg idi ve sol akciğer alt bölümünde solunum sesleri zayıftı. Alt ekstremitede ciddi ödem vardı. Kardiyak oskültasyonda üçüncü kalp sesi işitildi. Elektrokardiyografide sadece sinüs taşikardisi izlendi. Transtorasik ekokardiyografide, ilki çok büyük olmak üzere, sağ atriyumda ve ana pulmoner arterde trombüs görüldü. Hematolojik testlerde antifosfolipid antikor titreleri yüksek bulundu. Hastada yoğun pulmoner emboli olabileceği düşünüldü. Acil ameliyat için hazırlık sırasında kardiyovasküler kollaps gelişen hastada kardiyopulmoner resüssitasyon başarılı olmadı ve hasta kaybedildi.

Anahtar sözcükler: Antifosfolipid sendromu/komplikasyon; ekokardiyografi; pulmoner embolizm/etyoloji; venöz trombozis/etyoloji.

Antiphospholipid antibodies are associated with some connective tissue diseases such as systemic lupus erythematosus, and some infectious diseases such as syphilis and human immunodeficiency virus (HIV).[1,2] These antibodies are directed predominantly against negatively charged phospholipids. They play a role in thrombosis by an effect on platelet membranes, endothelial cells, and clotting proteins such as prothrombin, protein C, and protein S.

Antiphospholipid antibodies give rise to some clinical manifestations of thrombocytopenia, abortions, chorea, migraine, epilepsy, cutaneous symptoms, valvular heart disease, accelerated atheroma, and arterial and venous thromboses. The most common clinical presentation is deep venous thrombosis. The occurrence of pulmonary thromboembolism is approximately 9%. However, association of antiphospholipid antibody syndrome (APS) with a mobile intracardiac thrombus is an extremely rare finding.[3,4]

In this report, we presented a case in which APS resulted in sudden death due to massive pulmonary embolism.

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CASE REPORT

A 33-year-old male was admitted with dyspnea of increasing severity. He was in respiratory distress. He had a history of prior surgery for acute arterial embolism three months before in the department of cardiovascular surgery. On physical examination, breath sounds were weaker in the lower zone of the left lung and blood pressure was 80/60 mmHg. On cardiac auscultation, the third heart sound (gallop rhythm) was elicited. Severe lower limb edema was noted. Electrocardiography (ECG) showed only a sinusal tachycardia with no specific ECG changes. Transthoracic echocardiography revealed a huge thrombus in the right atrium (Fig. 1a). From the parasternal short-axis view, there was another thrombus in the main pulmonary artery (Fig. 1b). A diagnosis of massive pulmonary embolism was considered. Unfortunately, during arrangements for emergency operation, he developed cardiovascular collapse which did not respond to cardiopulmonary resuscitation.

Laboratory data were as follows: Serum urea and creatinine levels were high (68 mg/dl and 2.2 mg/dl, respectively), other biochemical parameters were in normal limits. There was no electrolyte imbalance. In hematological analysis, protein C, protein S, and fibrinogen were normal, but antiphospholipid antibodies were positive with high titration (IgG 90 U/ml, normal <15 U/ml; IgM 40 U/ml, normal <10 U/ml). Other rheumatologic markers (antinuclear antibodies, anti-double-stranded DNA antibodies, and anti-smooth muscle antibodies) were negative.

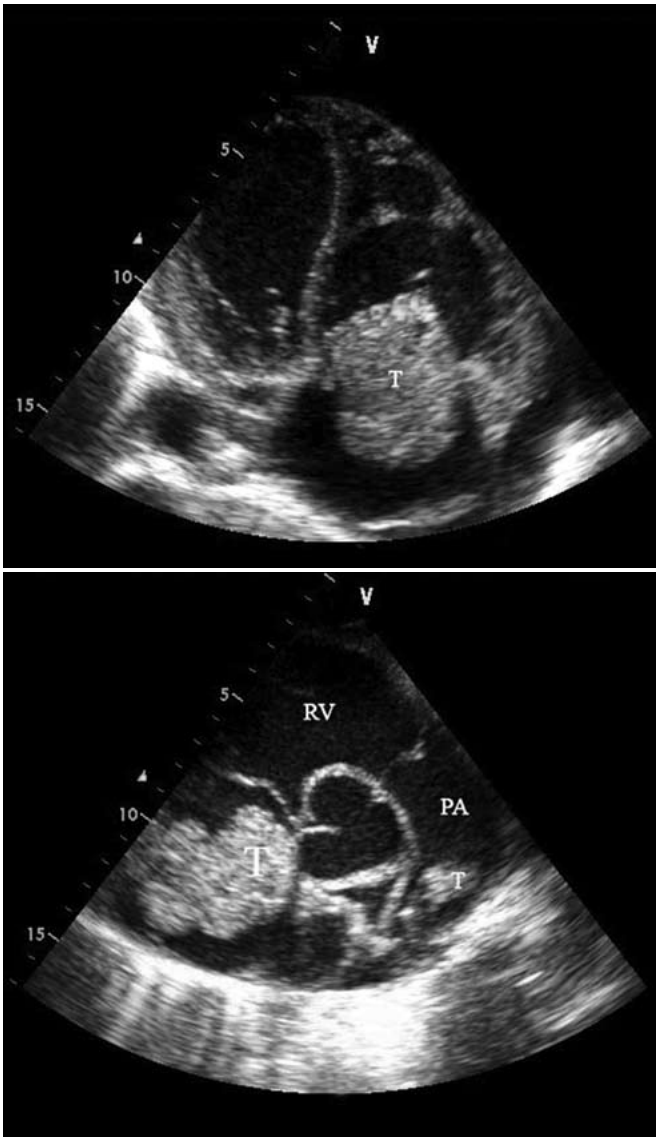

Figure 1. (A) The apical four-chamber view showing a huge thrombus in the right atrium. **(B)** The parasternal short-axis view showing both thrombi in the right atrium and the main pulmonary artery. RV: Right ventricle; PA: Pulmonary artery; T: Thrombus.

DISCUSSION

Massive pulmonary embolism is a catastrophic disease. Echocardiographic examination plays an important role in the diagnosis of pulmonary embolism. Generally, some indirect findings like right ventricular failure, right chamber dilatation, severe tricuspid regurgitation, and pulmonary hypertension are suggestive of pulmonary embolism. Demonstration of cardiac and/or pulmonary thrombus is extremely rare. Ye et al.[5] reported that the possibility of coexisting right heart thrombus should be considered in patients with APS presenting with pulmonary embolism.

Both venous and arterial thrombi are generally major findings in APS. In our case, the patient was operated on for an arterial thrombus three months before. Although there was a high suspicion of massive pulmonary embolism, we could not reach a definite diagnosis because the patient died before emergency operation and his family did not accept an autopsy. In a study of 19 APS patients with intracardiac thrombi, 50% of the cases had moderate thrombocytopenia (<75,000/μl) and 20% had underlying structural cardiac abnormalities.[6] In our case, platelet count was 98,000/μl and we did not determine any underlying cardiac abnormality.

The exact mechanism of intracardiac thrombus formation in APS is unclear. Circulating antiphospholipid antibodies, in the presence of other hemostatic defects, disrupt the balance between thrombosis and fibrinolysis, and might change the endocardial surface factors, contributing to clot formation.[6] In previous studies, it was speculated that an abnormal intracardiac blood flow pattern might contribute to

thrombosis,[7] diffuse ventricular dysfunction might predispose to the formation of intracardiac thrombus,[8] and rarely can an underlying abnormality be determined.[9] We demonstrated huge thrombi in the right atrium and main pulmonary artery by transthoracic echocardiography. In our case, apart from antiphospholipid antibodies, we did not determine any hemostatic defect, cardiac dysfunction, or an abnormal blood flow pattern causing thrombus formation. In our opinion, these features make our case original and rare.

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