Concurrence of Pleural Effusion and Inferior Vena Cava Thrombus in Antiphospholipid Syndrome Complicated by Intra-Cardiac Thrombus

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Abstract

Introduction: Antiphospholipid syndrome (APS) associated with intra-cardiac thrombus and pulmonary embolism is infrequent. However, the presence of pleural effusion is extremely rare.

Case Presentation: A 42-year-old woman, admitted with palpitation, dyspnea and history of stroke and thrombocytopenia is described. Doppler sonography revealed deep vein thrombosis. Moreover, subsequent investigation confirmed the presence of right atrial thrombus, pulmonary arteries embolism and bilateral pleural effusion. Inferior vena cava thrombosis was confirmed during cardiac surgery and all thrombi were removed subsequently. Laboratory studies revealed the diagnosis of APS. The patient experienced deep vein thrombosis one year later and no evidence of recurrent disease was documented after three years follow up.

Conclusions: APS complicated by inferior vena cava thrombosis coexistence of pulmonary embolism and intra-cardiac thrombus is not previously reported. Moreover, presence of pleural effusion with these conditions describes a unique case of APS.

Keywords: Antiphospholipid Syndrome, Pleural Effusion, Pulmonary Embolism, Inferior Vena Cava, Intracardiac, Thrombosis

1. Introduction

Antiphospholipid syndrome (APS) is an autoimmune disease characterized by vascular thrombosis and/or pregnancy morbidity in association with elevated levels of anticardiolipin antibodies (aCL) and/or lupus anticoagulant (LAC) (1). Intra-cardiac thrombosis accompanied by pulmonary embolism in APS is infrequently described (2, 3). However, presence of inferior vena cava (IVC) thrombus and bilateral pleural effusion with these conditions is not previously reported.

The current paper presents an unusual case of APS complicated by pleural effusion and IVC thrombosis concomitant with right atrial thrombus and pulmonary embolism.

2. Case Presentation

A 42-year-old woman presented with palpitation and dyspnea. She was a known case of idiopathic thrombocytopenic purpura resistant to therapy who had undergone splenectomy three weeks before. On examination of her legs, Doppler sonography revealed that she had deep vein thrombosis. Transthoracic echocardiography showed free-floating masses in the right atrium accompanied by dilation of the right cardiac chambers and the pulmonary artery. Although severe tricuspid regurgitation was observed, there was no evidence of vegetation on the valves. Result of a computed tomography (CT) pulmonary angiography confirmed right atrial thrombi and pulmonary arteries embolism coexistence of bilateral pleural effusion and suspected IVC thrombosis (Figures 1A-C). The patient had experienced one month of headache because of the subdural hemorrhage (platelets, 30,000 k/uL) and also she explained a history of stroke 26 years ago. The patient was transferred to the operating room for emergency surgical treatment. Mid-sternotomy was performed under general anesthesia. There was a huge right atrium and the right ventricular was severely distended. Cannulation of the superior vena cava and aorta was performed and cardiopulmonary bypass was established subsequently. A longitudinal right atrial incision was made and large clots were extracted from the IVC and right atrium. After cannulation of the IVC, right ventricle was found free of clot. Through a separate incision in the main pulmonary artery, clots were removed from the right and left pulmonary arteries gently by Desjardin forceps and no clot remained within these arteries (Figure 1D). Repairs of the right atrium and main pulmonary artery were done. The patient was weaned from cardiopulmonary bypass with low dose inotropic medica-
tion. Closure of sternotomy was performed and then she was transferred to the intensive care unit in good hemodynamic state. The patient was discharged from hospital on warfarin therapy. On outpatient workup, laboratory studies revealed that this is a case of APS secondary to systemic lupus erythematosus (SLE) due to elevated aCL and LAC titers. The patient experienced deep vein thrombosis one year later but transthoracic echocardiography was normal. After three years follow up, no clinical evidence of recurrent disease was documented.

3. Discussion

Antiphospholipid antibodies such as aCL and LAC are a family of autoantibodies directed against phospholipid-protein complexes (4). The first identification of these antibodies was in the patients with syphilis (5). Love et al. reported aCL and LAC in about 40% of SLE patients by analysis of 29 published series (6). APS comprises clinical manifestations including venous and arterial thrombosis accompanied by medium to high levels of aCL and/or LAC, as in the present case (1). This disease can occur either as primary in healthy persons or secondary to other systemic autoimmune disorders, especially SLE.

Venous thromboembolism is the most common clinical feature of APS which occurs in more than 20% of patients. Thrombocytopenia, stroke and fetal loss are other frequent presentations of APS while pulmonary embolism is less common (about 9% of cases) (7, 8).

APS related pleural effusion is extremely rare; only those cases accompanied by congestive heart failure, pulmonary embolism or spreading erythema have been reported (9-11). In the current case, SLE was determined as the cause of pleural effusion.

Cardiac involvements in APS included valvular abnormalities (vegetations, irregular thickening and dysfunction), coronary artery disease, dilated cardiomyopathy, intra-cardiac thrombi and pulmonary hypertension. Although valvular heart disease is the commonest finding, which involves most frequently mitral valve, followed by the aortic and tricuspid valves, the presence of intra-cardiac thrombus is relatively rare (12).

Thrombus formation, as a source of pulmonary and systemic emboli, is described in any cardiac chambers, especially on the right side. Lim et al. found that the occurrence of pulmonary or systemic emboli is approximately 63% in APS patients with intra-cardiac thrombus. They also revealed clinical findings including moderate thrombocytopenia and underlying structural cardiac abnormalities in 50% and 20% of cases, respectively (13).

IVC thrombosis is an uncommon clinical expression of APS patients described with deep vein thrombosis (14), pulmonary hypertension (15) and intestinal perforation (16). However, it was not previously reported in APS patients complicated by pleural effusion concomitant with right atrial thrombosis and pulmonary embolism.

Treatment of the thrombotic events in APS is based on the clinical state of the patients and also the location, size and shape of the thrombi. Although anticoagulation alone is described to resolve thrombus completely, excisional biopsy to remove thrombi and rule out the diagnosis of cardiac myxoma is usually necessary (13).

In conclusion, APS conveys several hypercoagulable complications; however, intra-cardiac thrombus coexistence of pulmonary embolism and IVC thrombosis was not previously reported. Moreover, presence of pleural effusion with these conditions describes a unique case of APS. Multidisciplinary management in such patients precludes bleeding or thromboembolic events by playing an important role in maintaining the level of anticoagulation.

Footnote

Authors’ Contribution: Study concept and design: Jamshid Bagheri; acquisition of data: Jamshid Bagheri and Amin Bagheri; analysis and interpretation of data: Jamshid Bagheri; drafting of the manuscript: Amin Bagheri; critical revision of the manuscript for important intellectual content: Jamshid Bagheri and Amin Bagheri; administrative, technical, and material support: Jamshid Bagheri and Amin Bagheri; study supervision: Jamshid Bagheri.

References

It reveals the presence of two masses (arrows) in the A, right atrium and bilateral pleural effusion; B, Partial thrombosis in the right and left pulmonary arteries; C, suspicious thrombus in the IVC (arrow); D, Intraoperative findings. Multiple thrombi removed from pulmonary arteries, IVC and right atrium. RA, right atrium; RV, right ventricle.