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Stylianos T. Panopoulos, Maria G. Tektonidou

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CASE REPORT

Intracardiac thrombosis in a young female as first presentation of primary antiphospholipid syndrome: A case report

Stylianos T. Panopoulos, Maria G. Tektonidou

First Propaedeutic Department of Internal Medicine, Rheumatologic Unit, Laikon Hospital and Joint Academic Rheumatology Programme of School of Medicine, National and Kapodistrian University of Athens

ABSTRACT

Primary antiphospholipid syndrome (PAPS) is an entity characterized by spontaneous and recurrent abortion and recurrent vascular thromboses (arterial and venous). Intracardiac thrombosis is a rare but life-threatening complication of PAPS. Herein we describe a 21 year-old woman admitted to hospital due to left pleurodynia and shortness of breath with no history of thrombotic events. Helix chest tomography scan disclosed pulmonary embolism as well as a filling defect of the inferior vena cava, which in subsequent cardiac magnetic resonance (MRI) proved to be intracardiac thrombus. Laboratory tests showed triple positivity for antiphospholipid antibodies, renal involvement and thrombocytopenia; PAPS, possibly catastrophic, was diagnosed. The patient was treated with iv pulses of corticosteroids, cyclophosphamide, intravenous immunoglobulin and oral anticoagulation (INR levels between 2.5 and 3), improved gradually and was discharged after 15 days of hospitalization. At 6-month follow-up new cardiac MRI revealed complete resolution of the thrombus. Patients with APS that present with pulmonary embolism should be investigated for the possibility of intracardial thrombus. Indefinite anticoagulation treatment in these patients is warranted due to high recurrence rates.

Corresponding author:
Maria Tektonidou, MD, PhD
First Propaedeutic Department of Internal Medicine, Rheumatologic Unit
Laikon Hospital, School of Medicine, National and Kapodistrian University of Athens
75 Mikras Asias Str., 11527 Athens, Greece
Tel: 2107462710, Fax: 2107462710,
Email: mtektonidou@gmail.com

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CASE

A 21-year-old woman with no previous medical or family history of thrombotic disease or history of any drug abuse presented at the emergency room with left pleurodynia, shortness of breath and fever. On examination she was in respiratory distress, with tachypnea (20 breaths/min), blood pressure of 140/80 mmHg, tachycardia (120 beats/min) and body temperature 38.2°C. Chest auscultation revealed decreased breath sounds of the left lower lung field with no other abnormal findings from clinical examination. Arterial blood gas analysis showed pH: 7.413; pO_2: 61 mmHg; pCO_2: 31.4 mmHg and bicarbonate: 22.4 mmol/L. On electrocardiogram there was sinus tachycardia without significant ST-T segment change and on chest X-ray left pleural effusion was noted. Subsequent helix chest tomography scan disclosed pulmonary embolism as well as a filling defect of the inferior vena cava at its junction with the right atrium after contrast agent injection; a finding that was suspicious of intracardiac thrombus. However, transthoracic echocardiography did not confirm the presence of intracardiac mass or any other morphological abnormalities of the valves. Cardiac magnetic resonance (MRI) is not available in our hospital. Additionally, venous duplex examination of the legs showed normal blood flow with absence of cloats in any deep vein, rulling out the possibility of deep vein thrombosis (DVT).

Laboratory tests revealed highly elevated inflammatory markers, positive antinuclear antibody 1/640 (speckled pattern), a prolonged activated partial prothrombin time (APTT) of 61s, mild thrombocytopenia (80,000/μL) and active urine sediment indicative of renal involvement. Treatment with Enoxaparin 8000 iu bid and prednisolone iv 1 mg/kg/day was initiated with the clinical suspicion of systemic lupus erythematosus (SLE). Further immunological tests showed highly elevated IgG anticardiolipin (115 GPL) and IgG anti-β2-glycoprotein antibodies and positive lupus anticoagulant (LA), whilst anti-double stranded DNA and anti-Smith antibodies were tested negative. Complements levels C_3 and C_4 were within normal limits. The third day of her hospitalization, she developed liveloid vasculitis in both hands. The patient did not undergo renal biopsy due to high risk for new embolic event in case of temporary withdrawal of anticoagulation.

The patient was diagnosed with primary antiphospholipid syndrome (PAPS) with clinical evidence of ongoing

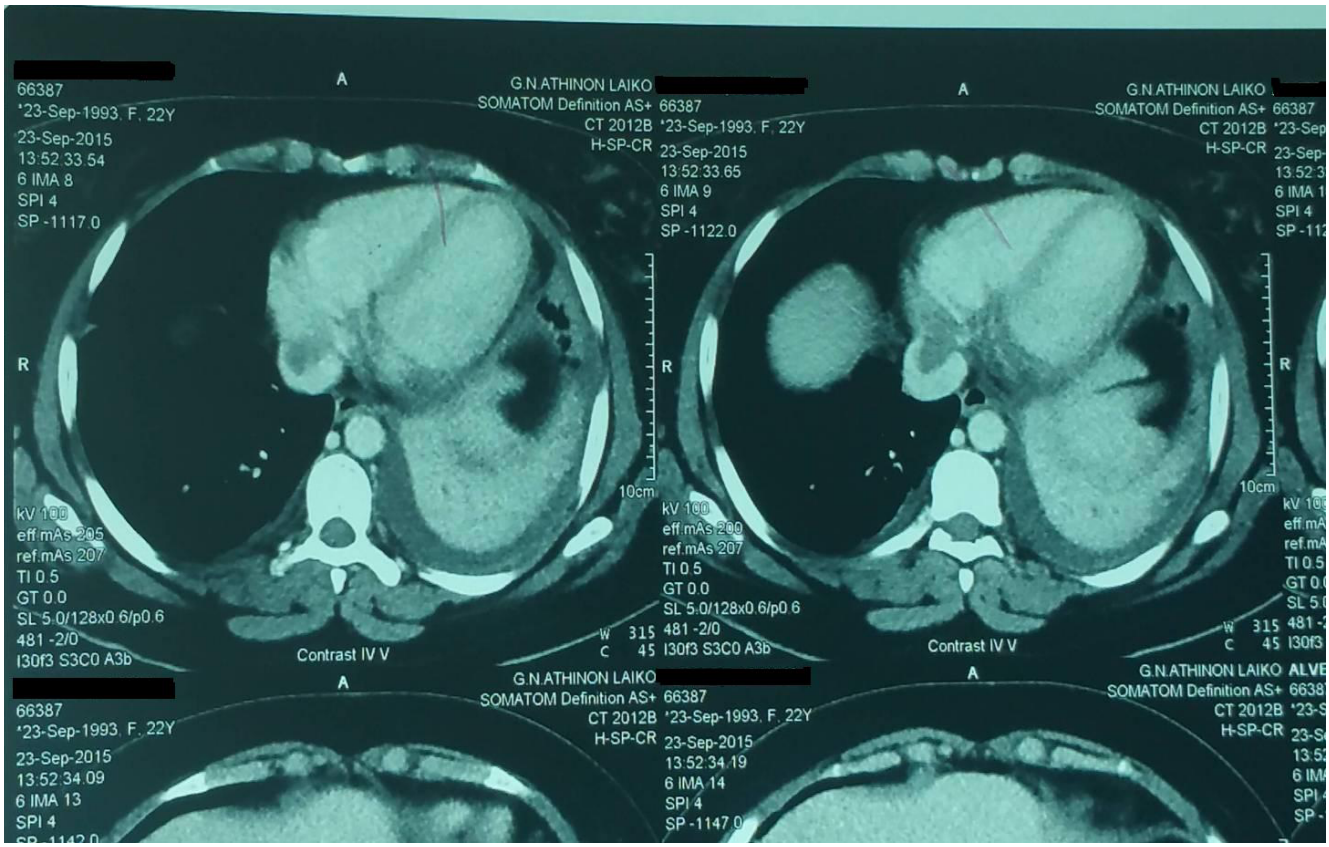

Figure 1. Computed tomography scan of the lungs shows a filling defect (arrow) in the inferior vena cava-right atrium, suspicious of intracardiac thrombosis

Table 1. Differential diagnosis of intracardiac masses

Cardiac Mass	T1 weighed sequence	T2 weighed sequence	LGE
Benign tumors			
Myxoma	**isointense**	**hyperintense**	**heterogenous**
Fibroma	**isointense**	**hypointense**	**increased enhancement**
Lipoma	**hyperintense**	**hyperintense**	**no uptake**
Rabdomyoma	**isointense**	**isointense**	**no/minimal uptake**
Malignant tumors			
Angiosarcoma	**heterogenous**	**heterogenous**	**heterogenous**
Lymphoma	**isointense**	**isointense**	**no/minimal uptake**
Rabdomyosarcoma	**isointense**	**hyperintense**	**homogenous**
Metastasis	**hypointense**	**hyperintense**	**heterogenous**
Thrombus	**hyperintense if acute (hypo- if chronic)**	**hyperintense if acute (hypo- if chronic)**	**no uptake**

catastrophic syndrome because of the development in less than one week period of pulmonary embolism with the potentially coexistent intracardiac thrombosis, renal involvement and livedoid vasculitis, in association with positive LA test and high levels of IgG anticardiolipin and anti-β2-glycoprotein antibodies. She was treated with 3 iv pulses of 1 gr methyl-prednisolone, 1 iv pulse of 1.2 gr cyclophosphamide (750 mg/m^2, patient's BSA: 1.5m^2) and intravenous immunoglobulin 400 mg/kg for 5 consecutive days. The patient improved gradually and was discharged after 15 days of hospitalization in good general condition. Her treatment included per os methylprednisolone 32 mg/day, mycophenolate mofetil 2gr/day and hydroxychloroquine 400 mg/day. The cardiac MRI in an outpatient diagnostic center showed a papillary mass (2x2cm), with hyperintense signal on T1- and hypointense signal on T2-weighted sequence and no enhancement after gadolinium injection, situated at the inferior vena cava at its junction with the right atrium, confirming the presence of intracardiac thrombus. At 6-month follow-up the patient was in perfect clinical condition without any symptoms, continued the same treatment (dosage of methylprednisone was tapered to 8 mg/day) and a new cardiac MRI that was performed revealed complete resolution of the thrombus.

DISCUSSION

APS has a variety of clinical manifestations with DVT being the most common as it develops in approximately 36-40% of patients, whilst pulmonary embolism is manifested in approximately 9% of APS patients.[1] Valvular abnormalities represent the most common cardiac manifestations ranging from 12 to 35% of cases.[1-2] Other heart manifestations include myocardial infarction, pulmonary hypertension, dilated myocardiopathy, coronary artery thrombosis and intracardial thrombosis. Intracardiac thrombosis is a rare but potentially life-threatening cardiac complication of APS as it can cause pulmonary and systemic embolic events. The differential diagnosis of intracardiac masses includes mainly benign or malignant tumors (most commonly myxoma), and thrombus (**Table 1**).[3] Right atrial thrombi can either originate from venous emboli that have become entrapped in the right heart or may develop in situ in the right atrium. In several cases, these thrombi are poorly visualised on trans-thoracic echocardiography (TTE), and a trans-oesophageal echocardiography (TEE) may be necessary for their detection. In general, TTE and TEE provide information only for the location and size of the intracardiac mass and further imaging by CT scan and, moreover, MRI is required in order to characterize the tissue composition and to differentiate between tumors and thrombus.[4]

The existing data on the management of intracardiac thrombosis in patients with APS are insufficient, as only sporadic cases with this rare manifestation have been reported.[3,5,6,7] In some of these cases including the one presented in this report, complete resolution of thrombi with anticoagulation alone has been described, but surgical intervention was in several cases required to remove the thrombus due to the great size of the mass and the high risk for recurrent systemic embolism. According to recommendations published in 2003 by a committee consensus for the treatment of cardiac disease in APS,[8] administration of intensive warfarin anticoagulation is always recommended in case of intracardiac thrombosis, while the decision for surgical intervention is individualised depending on the position and

size of the thrombus, the hemodynamic condition of the patient and the risk of recurrent events. The maintenance treatment for APS patients with thrombotic events, given the risk of recurrence,[9] requires lifelong anticoagulation treatment with warfarin (targeted to an international normalized ratio of 2.0–3.0). If thrombotic events recur, warfarin should be increased by means of high-intensity therapy. Alternatively, addition of anti-platelets to anticoagulation treatment can also be considered.[10]

In conclusion, the presence of intracardial thrombi is a rare but life-threatening complication of APS. Patients with APS that present with pulmonary embolism, especially if there is no proof of DVT, should be investigated for the possibility of intracardial thrombus. Indefinite anticoagulation treatment in these patients is warranted due to high recurrence rates.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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