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CASE REPORT

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Left ventricular thrombus mimicking primary cardiac tumor in a patient with primary antiphospholipid syndrome and recurrent systemic embolism

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Abstract

Primary antiphospholipid syndrome (APS) is a well-defined entity characterized by spontaneous and recurrent abortion, thrombocytopenia and recurrent vascular thromboses (arterial and venous). Left ventricular thrombus mimicking primary cardiac tumor with recurrent systemic embolism has not been previously reported.

In this report we describe a 39 year-old man admitted to hospital presenting with left hemiparesis and a peripheral embolism. He had no history of thrombotic events. Transthoracic echocardiography showed a large, polypoid and mobile mass (4.0 × 1.2 cm) attached to the apex of the left ventricle, highly suggestive of primary cardiac tumor. The patient subsequently underwent open heart surgery. The histological examination showed an older thrombus and a fresh thrombus. Post-operative laboratory tests showed lupus anticoagulant activity, confirming the primary APS diagnosis. The patient initiated treatment with oral anticoagulation (INR levels between 2 and 3) and was discharged 29 days after surgery. At ten month follow-up, he was symptom-free with long-term anticoagulation therapy. No evidence of intracardiac mass recurrence on two-dimensional echocardiography was seen.

Intracardiac thrombus has been rarely reported as a complication of primary APS. Left ventricular mass mimicking primary cardiac tumor with recurrent systemic embolism has not been previously reported. Pre-operative investigations could not distinguish such a thrombus from a cardiac tumor and the diagnosis was made post-operatively. (Cardiol J 2009; 16, 6: 560–563)

Key words: left ventricular thrombus, systemic embolism, primary antiphospholipid syndrome, primary cardiac tumor, cardiovascular surgery

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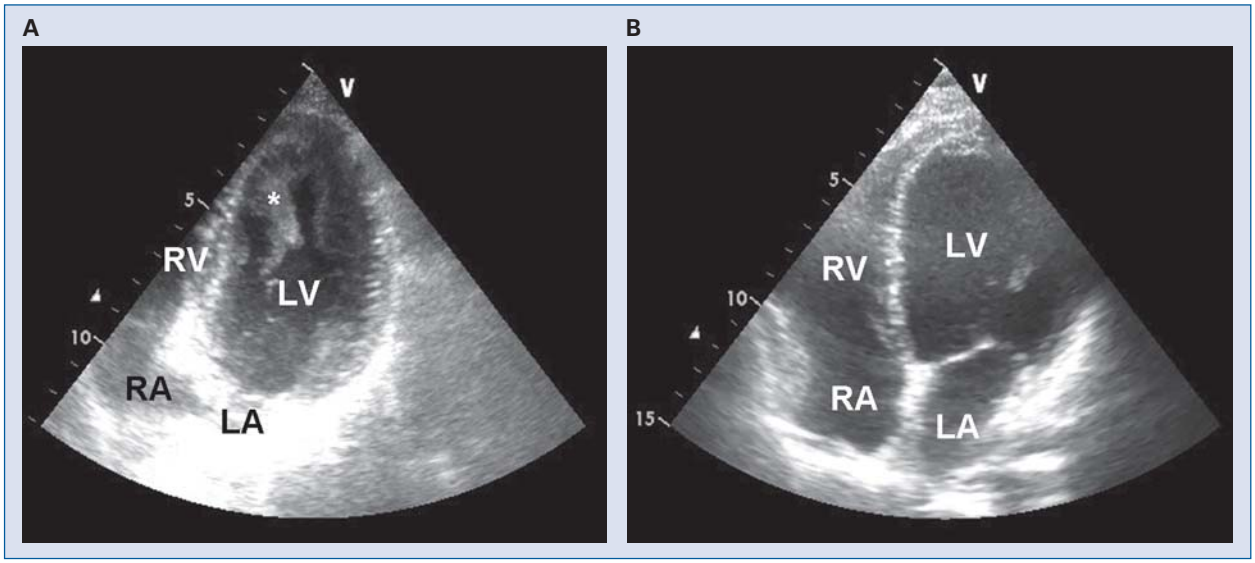

Figure 1. A. Transthoracic echocardiography shows a polypus mass attached to the apex of the left ventricle (LV), floating freely during cardiac cycle; **B.** No evidence of intracardiac mass recurrence at two-dimensional transthoracic echocardiography at ten month follow-up; RV — right ventricle; LA — left atrium; RA — right atrium.

Introduction

Antiphospholipid syndrome (APS) is characterized by spontaneous abortion, thrombocytopenia and recurrent vascular thromboses (arterial and venous) in association with medium to high titers of antiphospholipid (APL) antibodies or the demonstration of a positive lupus anticoagulant (LA) test. This disorder is referred to as primary APS when it occurs alone [1]. Less well known is the association between APL antibodies and primary intracardiac thrombosis.

Left ventricular giant thrombus mimicking a primary cardiac tumor with recurrent systemic embolism in a patient with primary antiphospholipid syndrome has not been previously reported.

Case report

In this report we describe a 39 year-old man admitted to hospital presenting with left hemiparesis and peripheral embolism. Physical examination revealed a blood pressure of 120/70 mm Hg, heart rate of 80 bpm and 16 breaths/min. Heart sounds were regular and distant. No murmurs or rubs were noted. Jugular venous pressure was normal. Lungs were clear, abdomen soft and no peripheral edema was found. Neurological examination revealed left hemiparesis. The chest X-ray film and a 12-lead electrocardiogram were unremarkable. Colour Doppler ultrasound and duplex scan of the lower limb ruled out deep venous thrombosis.

Two-dimensional transthoracic echocardiography revealed a normal left ventricular and left atrium dimensions, there were no wall motion abnormalities, heart valve abnormalities were not seen, with no evidence of vegetation. A large, polypoid and mobile mass (4.0 × 1.2 cm) was seen attached to the apex of the left ventricle, which suggested a primary cardiac tumor (Fig. 1A).

In view of these findings and the history of recurrent systemic embolism, the patient subsequently underwent open heart surgery. On cardiopulmonary bypass, the left ventricle was opened and a large mass was identified attached to the apex of the left ventricle. The gross examination of the operative specimen revealed multiple masses with irregular surfaces and the histological examination showed a partly older and a partly fresh thrombus (Fig. 2). The patient experienced no post-operative complications.

Post-operative laboratory tests showed LA activity without anticardiolipin antibodies, supporting the diagnosis of primary antiphospholipid syndrome [2]. The patient began treatment with oral anticoagulation, international normalized ratio (INR) levels 2–3. The patient was discharged from the hospital 29 days after surgery, on a regimen of warfarin (INR 2–3). At ten month follow-up, he was symptom-free with long-term anticoagulation therapy and no evidence of intracardiac mass recurrence on two-dimensional echocardiography was seen (Fig. 1B).

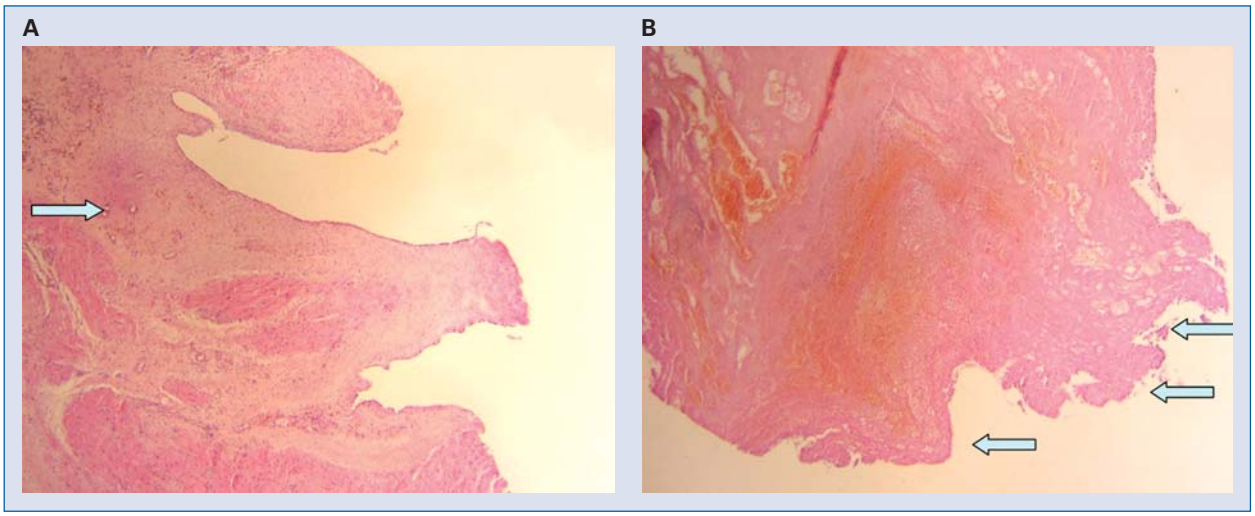

Figure 2. Histological examination shows an older thrombus with a fibrin network (**A**) and a fresh thrombus (**B**) with minimal vascular or perivascular inflammation (HE 10×).

Discussion

Antiphospholipid syndrome is defined according to the clinical and laboratory criteria. That means a history of thrombus associated with the presence in blood of lupus anticoagulant and anticardiolipin antibodies of inmunoblogulin G and/or immunoglobulin M isotype positive, in medium or high titers on two or more occasions at least 12 weeks apart [1].

The commonest manifestations of APS are deep vein thrombosis, pulmonary thromboembolism, stroke, myocardial infarction and spontaneous abortion.

The commonest echocardiographic findings in patients with APS are either valve vegetations or thickening of the valve leaflets. These findings are more frequent than intracardiac thrombus. The literature reports only a few cases of intracardiac thrombosis [3, 4] associated with APS, observed in all cardiac chambers, provoking either pulmonary or systemic embolism. When intracardiac thrombosis occurs, the differential diagnosis of left ventricular mass includes tumor and thrombus.

Intravenous administration of the new generation contrast agents during a bedside echocardiographic study has been successfully used to distinguish between intracardial masses by evaluating their vascularity. Thrombus is considered an 'avascular' structure, so it does not reveal perfusion, unlike a malignant tumor which presents increased vascularity due to higher microvascular density [5].

The commonest primary cardiac tumor is myxoma. Five per cent of myxomas arise in the left ventricle. In our patient, without clinical history of APS, a left ventricular myxoma diagnosis was considered.

Left ventricular thrombi can develop in several situations. First, they may be associated with wall motion abnormalities, but our patient had normal regional left ventricular function.

The risk of intracardiac thrombosis and/or thromboembolism is increased in patients who show positive test results for LA activity. This was demonstrated in our patient, after surgery.

Venous thrombosis and pulmonary embolism are also more common in those with LA activity, while coronary, cerebrovascular, and peripheral arterial events are more likely in those with elevated levels of IgG or IgM anticardiolipin antibodies [6].

Patients who have primary APS can develop intracardiac thrombus, as did our patient. Pre-operative investigations cannot differentiate such a thrombus from a myxoma, and the diagnosis was made post-operatively.

It remains unknown whether prolonged heparin administration, thrombolysis, high-intensity anticoagulant with warfarin or a surgical excision is the best therapeutic approach.

Nonetheless, the great size of the mass and its free-floating appearance on two-dimensional echocardiography, the history of hemiparesis and the peripheral embolism, which seemed to place our patient at high risk for recurrent systemic embolism, led us to choose surgical excision. Moreover, the pathological features of an older thrombosis, which could constitute the nucleus for further fresh

thrombus deposition, further supported the surgical approach.

The mechanism of this intracardiac thrombus in patients with primary APS would be a hypercoagulability state mediated by monocyte tissue factor expression. This activity results in formation of a complex between tissue factor and coagulation factor VII on the cell surface, which activates factor IX and factor X. Factor X converts prothrombin to thrombin, the major serine protease leading to fibrin formation and platelet activation [7].

Some relapsing thrombosis has been reported despite long-term warfarin treatment. A follow-up by two-dimensional echocardiography should be recommended in patients with primary APS [8].

Conclusions

Intracardiac thrombus has been rarely reported as a sequel to antiphospholipid syndrome. Large, polypoid and mobile left ventricular thrombus mimicking myxoma with recurrent systemic embolism in a patient with primary antiphospholipid syndrome has not been previously reported. In our patient, the pre-operative investigations could not differentiate such a thrombus from a myxoma, and the diagnosis was made post-operatively.

Patients who have APS are at high risk of developing intracardiac thrombus. However, the thrombus can be successfully resected. To preclude thrombus formation, warfarin treatment is indicated in all patients with APS.

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References

1. Miyakis S, Lockshin MD, Atsumi T et al. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). J Thromb Haemost, 2006; 4: 295–306.
2. Galli M, Luciani D, Bertolini G, Barbui T. Lupus anticoagulants are stronger risk factors for thrombosis than anticardiolipin antibodies in the antiphospholipid syndrome: A systematic review of the literature. Blood, 2003; 101: 1827.
3. Cianciulli TF, Saccheri MC, Redruello HJ et al. Right atrial thrombus mimicking myxoma with pulmonary embolism in a patient with systemic lupus erythematosus and secondary antiphospholipid syndrome. Tex Heart Inst J, 2008; 35: 454–457.
4. Basso C, Bottio T, Rubino M et al. Antiphospholipid syndrome and right atrial mass. J Thorac Cardiovasc Surg, 2005; 130: 1462–1463.
5. Rallidis LS, Papadopoulos CC, Kelekis NL Kremastinos DTh. The role of myocardial contrast echocardiography to assess the origin of a mass in right cardiac cavities. Echocardiography, 2009; 26: 88–92.
6. Soltesz P, Veres K, Lakos G et al. Evaluation of clinical and laboratory features of antiphospholipid syndrome: A retrospective study of 637 patients. Lupus, 2003; 12: 302.
7. Amengual O, Atsumi T, Khamashta MA, Hughes GR. The role of the tissue factor pathway in the hypercoagulable state in patients with the antiphospholipid syndrome. Thromb Haemost, 1998; 79: 276–281.
8. Khamashta MA, Cuadrado MJ, Mujic F et al. The management of thrombosis in the antiphospholipid-antibody syndrome. N Engl J Med, 1995; 332: 993.